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As confidentially submitted to the Securities and Exchange Commission on May 14, 2014

Registration No. 333-

UNITED STATES
SECURITIES AND EXCHANGE COMMISSION

Washington, D.C. 20549

FORM S-1

REGISTRATION STATEMENT
UNDER
THE SECURITIES ACT OF 1933

BELLEROPHON THERAPEUTICS LLC

(to be converted into Bellerophon Therapeutics, Inc.)
(Exact name of registrant as specified in its charter)

Delaware	**2834**	**36-4771642**
(State or other jurisdiction of incorporation or organization)	(Primary Standard Industrial Classification Code Number)	(I.R.S. Employer Identification No.)

Perryville III Corporate Park
53 Frontage Road, Suite 301
Hampton, New Jersey 08827
(908) 574-4770
(Address, including zip code, and telephone number, including area code, of registrant's principal executive offices)

Daniel Tassé
Chief Executive Officer
Bellerophon Therapeutics LLC
Perryville III Corporate Park
53 Frontage Road, Suite 301
Hampton, New Jersey 08827
(908) 574-4770
(Name, address, including zip code, and telephone number, including area code, of agent for service)

Copies to:

Lia Der Marderosian **Steven D. Singer** **Wilmer Cutler Pickering Hale and Dorr LLP** **60 State Street** **Boston, Massachusetts 02109** **Telephone: (617) 526-6000**	**Mitchell S. Bloom** **Thomas S. Levato** **Goodwin Procter LLP** **Exchange Place** **Boston, Massachusetts 02109** **Telephone: (617) 570-1000**

Approximate date of commencement of proposed sale to the public:
As soon as practicable after this Registration Statement is declared effective.

If any of the securities being registered on this form are to be offered on a delayed or continuous basis pursuant to Rule 415 under the Securities Act of 1933, check the following box. o

If this form is filed to register additional securities for an offering pursuant to Rule 462(b) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering. o

If this form is a post-effective amendment filed pursuant to Rule 462(c) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering. o

If this form is a post-effective amendment filed pursuant to Rule 462(d) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering. o

Indicate by check mark whether the registrant is a large accelerated filer, an accelerated filer, a non-accelerated filer, or a smaller reporting company. See the definitions of "large accelerated filer," "accelerated filer" and "smaller reporting company" in Rule 12b-2 of the Exchange Act.

Large accelerated filer o	Accelerated filer o	Non-accelerated filer ☒ (Do not check if a smaller reporting company)	Smaller reporting company o

CALCULATION OF REGISTRATION FEE

Title of Each Class of Securities To Be Registered	Proposed Maximum Aggregate Offering Price(1)	Amount of Registration Fee(2)
Common Stock, $0.01 par value per share		

(1) Estimated solely for the purpose of calculating the registration fee pursuant to Rule 457(o) under the Securities Act of 1933, as amended.

(2) Calculated pursuant to Rule 457(o) based on an estimate of the proposed maximum aggregate offering price.

The Registrant hereby amends this Registration Statement on such date or dates as may be necessary to delay its effective date until the Registrant shall file a further amendment which specifically states that this Registration Statement shall thereafter become effective in accordance with Section 8(a) of the Securities Act of 1933 or until the Registration Statement shall become effective on such date as the Commission, acting pursuant to said Section 8(a), may determine.

EXPLANATORY NOTE

Bellerophon Therapeutics LLC, the registrant whose name appears on the cover of this registration statement, is a Delaware limited liability company. Immediately prior to the effectiveness of this registration statement, Bellerophon Therapeutics LLC will be converted into a Delaware corporation, which we refer to as the Company Conversion, and renamed Bellerophon Therapeutics, Inc. Shares of the common stock, par value $0.01 per share, of Bellerophon Therapeutics, Inc. are being offered by the prospectus that forms a part of this registration statement. For convenience, except as context otherwise requires, all information included in the prospectus that forms a part of this registration statement is presented (i) giving effect to the Company Conversion and (ii) assuming the conversion of our non-voting shares into voting shares of our common stock.

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The information in this prospectus is not complete and may be changed. We may not sell these securities until the registration statement filed with the Securities and Exchange Commission is effective. This prospectus is not an offer to sell these securities and it is not soliciting an offer to buy these securities in any jurisdiction where the offer or sale is not permitted.

Subject to Completion, dated May 14, 2014

PRELIMINARY PROSPECTUS

Shares

Common Stock

We are offering shares of our common stock. This is our initial public offering and no public market currently exists for our shares. We anticipate that the initial public offering price of our common stock will be between $ and $ per share.

We intend to apply to list our common stock on The NASDAQ Global Market under the symbol "BLPH."

We are an emerging growth company as that term is used in the Jumpstart Our Business Startups Act of 2012, and as such, are subject to reduced public company disclosure standards. See "Prospectus Summary—Implications of Being an Emerging Growth Company."

Investing in our common stock involves a high degree of risk. Before buying any shares, you should carefully read the discussion of material risks of investing in our common stock in "Risk Factors" beginning on page 11 of this prospectus.

	Per Share	Total
Initial public offering price	$	$
Underwriting discount	$	$
Proceeds to us, before expenses	$	$

We have granted the underwriters an option to purchase up to additional shares of our common stock to cover over-allotments. The underwriters can exercise this option at any time within 30 days after the date of this prospectus.

Neither the Securities and Exchange Commission nor any state securities commission has approved or disapproved of these securities or passed on the adequacy or accuracy of this prospectus. Any representation to the contrary is a criminal offense.

The underwriters expect to deliver the shares of common stock to investors on or about , 2014.

Leerink Partners **Cowen and Company**

The date of this prospectus is , 2014.

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Engineering CAD illustrations of our (1) INOpulse® Mark2 device, (2) triple-lumen cannula and (3) nosepiece detail

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Neither we nor the underwriters have authorized anyone to provide you with information other than that contained in this prospectus or in any free writing prospectus prepared by or on behalf of us or to which we have referred you. We and the underwriters take no responsibility for, and can provide no assurance as to the reliability of, any other information that others may give to you. We are offering to sell, and seeking offers to buy, shares of our common stock only in jurisdictions where offers and sales are permitted. The information contained in this prospectus is accurate only as of the date of this prospectus, regardless of the time of delivery of this prospectus or any sale of our common stock. Our business, financial condition, results of operations and prospects may have changed since that date.

No action is being taken in any jurisdiction outside the United States to permit a public offering of our common stock or possession or distribution of this prospectus in that jurisdiction. Persons who come into possession of this prospectus in jurisdictions outside the United States are required to inform themselves about and to observe any restrictions as to this offering and the distribution of this prospectus applicable to that jurisdiction.

The trademarks, trade names and service marks appearing in this prospectus are the property of their respective owners.

PROSPECTUS SUMMARY

This summary highlights information contained elsewhere in this prospectus. This summary does not contain all of the information you should consider before investing in our common stock. You should read this entire prospectus carefully, especially the "Risk Factors" section and our financial statements and the related notes appearing at the end of this prospectus, before making an investment decision.

This prospectus relates to an offering of our common stock following certain transactions described herein that will occur prior to the effectiveness of the registration statement of which this prospectus is a part, which we refer to as the Company Conversion. As used in this prospectus, unless the context otherwise requires, references to the "Company," "Bellerophon," "we," "us" and "our" refer to (i) following the date of the Company Conversion discussed under the heading "Company Conversion," Bellerophon Therapeutics, Inc. and its consolidated subsidiaries, or any one or more of them as the context may require, and (ii) prior to the date of the Company Conversion, Bellerophon Therapeutics LLC and its subsidiaries, or any one or more of them as the context may require.

Company Overview

We are a clinical stage biotherapeutics company focused on developing innovative products at the intersection of drugs and devices that address significant unme medical needs in the treatment of cardiopulmonary and cardiac diseases. We are developing three product candidates. Two of our product candidates are based on our proprietary pulsatile nitric oxide delivery device, which we refer to as INOpulse®, and are in Phase 2 clinical trials—one for the treatment of pulmonary arterial hypertension, or PAH, and a second for the treatment of pulmonary hypertension associated with chronic obstructive pulmonary disease, or PH-COPD. Our third product candidate, bioabsorbable cardiac matrix, or BCM, is currently in a feasibility clinical trial (which is a CE mark registration trial in the European Union and is comparable to a Phase 2 clinical trial in U.S. drug development). We are developing BCM for the prevention of cardiac remodeling and subsequent congestive heart failure following acute myocardial infarction, or AMI, commonly known as a heart attack.

Our Product Candidates

Our product candidates are summarized in the table below. We have worldwide commercialization rights to our three product candidates.

Product Candidate	Primary Indication	Phase 1 / Pilot	Phase 2 / Feasibility	Phase 3 / Pivotal	Upcoming Milestone
INOpulse for PAH (IK-7001)	Improvement in functional capacity for patients with PAH				Trial completion expected by end of 2014
INOpulse for PH-COPD (IK-7002)	Reduction in mortality and morbidity in patients with PH-COPD on long-term oxygen therapy				Trial completion expected in mid-2014
BCM (IK-5001)	Prevention of cardiac remodeling and subsequent congestive heart failure following AMI				Trial completion expected in mid-2015

INOpulse

INOpulse is an extension of the technology that is used in hospitals to deliver continuous-flow inhaled nitric oxide. Use of inhaled nitric oxide is approved by the U.S. Food and Drug Administration, or the FDA, and certain other regulatory authorities to treat persistent pulmonary hypertension of the

newborn. Ikaria, Inc., or Ikaria, has marketed continuous-flow inhaled nitric oxide as INOmax for this indication since approval in 1999. In October 2013, Ikaria transferred the INOpulse program to us with exclusive worldwide rights to develop and commercialize INOpulse in PAH, PH-COPD and pulmonary hypertension associated with idiopathic pulmonary fibrosis, or PH-IPF, with no royalty obligations. Our INOpulse program is built on scientific and technical expertise developed for the therapeutic delivery of inhaled nitric oxide. From the inception of our business through December 31, 2013, $176.5 million was invested in the development o our product candidates, all of which was funded by Ikaria. In 2010, we filed an investigational new drug application, or IND, for INOpulse for the treatment of patien with PAH, which is a form of pulmonary hypertension that is closely related to persistent pulmonary hypertension of the newborn. In 2012, we filed a second IND for INOpulse for the treatment of patients with PH-COPD. These IND programs were included in the assets that were transferred to us by Ikaria.

Nitric oxide is naturally produced and released by portions of the blood vessels and results in smooth muscle relaxation. In particular, nitric oxide controls muscl tone in blood vessels and thus is an important factor in regulating blood pressure. As the muscles of the blood vessels relax, blood flow increases, helping the heart to deliver more blood to the body. When administered by inhalation, the action of nitric oxide has minimal effects on blood pressure outside of the lungs, an important safety consideration.

A limitation to the chronic use of inhaled nitric oxide is the lack of a safe and compact delivery system for outpatient use. We have designed INOpulse to be portable for use by ambulatory patients on a daily basis inside or outside their homes. INOpulse is designed to automatically adjust based on a patient's breathing pattern to deliver a constant and appropriate dose of the inhaled nitric oxide over time, independent of the patient's activity level, thus ensuring more consistent dosing in the alveoli of the lungs. In addition, our proprietary triple-lumen nasal cannula enables more accurate delivery of the dose to the patient with minimal infiltration of oxygen, which can have an undesirable reaction with inhaled nitric oxide. INOpulse is also compatible with many long-term oxygen therapy systems.

The ongoing INOpulse clinical trials are utilizing the first generation INOpulse DS device. We expect our future trials will use the next generation INOpulse Mark2 device, or the Mark2, which has approximately the same dimensions as a paperback book and weighs less than 2.5 pounds. The Mark2 has a simple user interface and a battery life of approximately 24 hours, which can be readily recharged in four hours, typically while the patient sleeps. The Mark2 has been well received by patients in the usability research we have conducted.

We have been issued patents with respect to the pulsed delivery of nitric oxide to ensure a consistent dose over time that expire as late as 2027 in the United State and as late as 2026 in certain other countries. We have also filed several patent applications for certain of the innovations included in the Mark2, and certain of the resulting patents, if issued, will expire in 2033.

INOpulse for PAH

We are developing our lead product candidate, INOpulse for the treatment of PAH, to address a significant and unmet medical need in an orphan disease. This product candidate represents a potential first-in-class therapy. PAH is characterized by abnormal constriction of the arteries in the lung, which increases the blood pressure in the lungs and results in abnormal strain on the heart's right ventricle, eventually leading to heart failure. If left untreated, primary PAH patients have a median survival of less than three years. While prevalence data varies widely, we estimate there are at least 35,000 patients currently treated for PAH in the United States and European Union. Moreover, because PAH is rare and causes varied symptoms, we believe there is significant under-diagnosis of the condition. There are several approved therapies for PAH, and we estimate, based on public product sale data, that 2012 combined global sales were over $4.0 billion. Despite treatment of PAH patients with these therapies,

PAH continues to be a life-threatening, progressive disorder with estimates of median survival ranging from three to five years.

We commenced a randomized, placebo-controlled Phase 2 clinical trial of INOpulse for PAH in April 2012. We plan to enroll at least 78 patients in this trial at 52 clinical sites in the United States and Canada. We expect to complete this trial by the end of 2014. After consultation with appropriate regulatory authorities, we plan to initiate a pivotal Phase 3 clinical trial program in the second half of 2015. The FDA has granted orphan drug designation to nitric oxide for the treatment of PAH.

INOpulse for PH-COPD

We are developing a second product candidate, INOpulse for the treatment of PH-COPD. COPD is a disease characterized by progressive and persistent airflow limitations. Patients with more severe COPD frequently have hypoxemia and are treated with long-term oxygen therapy. Despite treatment with oxygen, hypoxemia can progress and cause pulmonary hypertension. We estimate that there are approximately 700,000 PH-COPD patients in the United States alone. PH-COPD patients have a lower median life expectancy and a higher rate of hospitalization than COPD patients with similar respiratory disease but without pulmonary hypertension. Currently, the generally accepted treatments for PH-COPD are oxygen therapy, pulmonary rehabilitation and lung transplant.

We commenced a randomized, placebo-controlled, dose-confirmation Phase 2 clinical trial of INOpulse for PH-COPD in December 2012. We plan to enroll at least 159 patients in this trial at 43 clinical sites in the United States. We expect to complete this trial by mid-2014. After completion of this clinical trial and additiona discussions we expect to have with U.S. and EU regulatory authorities, we will assess our options for further development of INOpulse for PH-COPD, including potential partnerships.

BCM

We are developing a third product candidate, BCM, a medical device intended to prevent cardiac remodeling and subsequent congestive heart failure following an AMI. Cardiac remodeling is a structural alteration of the heart that results in reduced heart function and often leads to congestive heart failure. BCM is delivered during a minimally invasive, commonly performed cardiac procedure called a percutaneous coronary intervention procedure. BCM is a formulated sterile solution of sodium alginate and calcium gluconate designed to be administered as a liquid through the coronary artery. When administered following an AMI, BCM flows into damaged heart muscle where, in the presence of abnormally high extracellular calcium released by the damaged cells, it forms a protective hydrogel meshwork within the wall of the heart's left ventricle. In our pre-clinical animal studies, as calcium levels in the damaged area returned to normal, BCM dissolved and was excreted through normal kidney function. Based on pre-clinical animal studies, it appears that BCM can act as a flexible scaffold to provide physical support to the ventricle wall in the early stages of recovery following an AMI and prevent further structural damage while the heart muscle heals. In a 27-patient pilot trial conducted by BioLineRx Ltd., BCM was well tolerated. As a Class III device, BCM is eligible for development through the premarket approval, or PMA, regulatory pathway in the United States. We have an exclusive worldwide license to BCM from BioLineRx Ltd. and its subsidiary, or BioLine, including with respect to issued composition of matter patents on BCM that expire as late as 2029 in the United States, with a possible patent term extension to 2032 to 2034, depending on the timing of marketing approval and other factors, and 2024 in certain other countries.

Data from the American Heart Association and the European Association for Percutaneous Cardiovascular Interventions suggests that a total of approximately 2,000,000 patients suffer a heart attack in the United States and European Union each year, with at least 750,000 of these patients having a ST-segment elevated myocardial infarction, or STEMI. Of these STEMI patients,

approximately 75% in the United States and a weighted average across France, Germany, Italy and the United Kingdom of over 50% currently undergo a percutaneou coronary intervention procedure and could be candidates for BCM if they are at risk for remodeling. We are testing BCM in STEMI patients who have a percutaneous coronary intervention, specifically in those patients who meet our inclusion criteria for high risk for remodeling.

We initiated a feasibility clinical trial of BCM in December 2011 and enrolled the first patient in April 2012. This trial is a CE mark registration trial in the European Union and is comparable to a Phase 2 trial in U.S. drug development. We plan to enroll approximately 300 patients in this trial at up to approximately 90 clinical sites in Europe, Australia, North America and Israel. We expect to complete this trial by mid-2015. If the results of this trial are positive, we expect it would form the basis for our application for CE marking in the European Union. We also plan to initiate a pivotal trial to support a PMA submission for regulatory approval in the United States.

Our Strategy

Our goal is to become a leader in developing and commercializing innovative products at the intersection of drugs and devices that address significant unmet medical needs in the treatment of cardiopulmonary and cardiac diseases. The key elements of our strategy to achieve this goal include:

- *Advance the clinical development of INOpulse.* Our primary focus for INOpulse is for use in treating PAH patients. Assuming positive results from ou ongoing Phase 2 clinical trial in PAH, we intend to initiate a Phase 3 clinical trial program in the second half of 2015. For PH-COPD, assuming positiv results from our ongoing Phase 2 trial, we plan to evaluate our options for further development, including potential partnerships.

- *Advance the clinical development of BCM in the prevention of cardiac remodeling following AMI.* Assuming positive results from our ongoing feasibility clinical trial, we expect to file for CE marking in the European Union in the second half of 2015 and to initiate a pivotal trial to support a PMA submission seeking marketing approval in the United States.

- *Leverage our historical core competencies to expand our pipeline.* We have years of institutional experience in the use of inhaled nitric oxide in treating pulmonary hypertension and in the development of drug-device combination product candidates. We expect to develop INOpulse for treatmen of patients with PH-IPF. Our longer-term vision is to identify and opportunistically in-license innovative therapies that are at the intersection of drugs and devices and to develop and commercialize these product candidates.

- *Build commercial infrastructure in select markets.* As we near completion of the development of any product candidates, we expect to build a commercial infrastructure to enable us to market and sell our product candidates, if approved, using a specialty sales force in the United States. While we may partner with third parties to commercialize our product candidates in certain countries, we may also choose to establish commercialization capabilities in select countries outside the United States.

The Spin-Out

In October 2013, Ikaria completed an internal reorganization of certain assets and subsidiaries, in which it transferred to us exclusive worldwide rights, with no royalty obligations, to develop and commercialize INOpulse in PAH, PH-COPD and PH-IPF. Following the internal reorganization, in February 2014, Ikaria distributed all of our outstanding units to its stockholders through the payment of a special dividend on a pro rata basis based on each stockholder's ownership of Ikari capital stock. We refer to Ikaria's distribution of our outstanding units to its stockholders as the Spin-Out. Shortly after the Spin-Out, Ikaria was acquired by entities affiliated with Madison Dearborn Partners.

In connection with the Spin-Out, we entered into several agreements with Ikaria providing for, among other things, the provision of transition services, the cross license of certain intellectual property, commitments not to compete, the manufacture and supply of the INOpulse drug and device and certain employee matters.

As used in this prospectus, unless context otherwise requires, references to "Ikaria" refer to Ikaria, Inc. and its subsidiaries.

Risk Factors

Our business is subject to a number of risks of which you should be aware before making an investment decision. As a clinical stage biotherapeutics company, w may face inherent risks in our business and our industry generally. These risks are discussed more fully in the "Risk Factors" section of this prospectus immediately following this prospectus summary. These risks include, among others:

- We have incurred significant losses since inception. We expect to incur losses over the next several years and may never achieve or maintain profitability.
- Our very limited operating history as a stand-alone company may make it difficult for you to evaluate the success of our business to date and to assess our future viability. We currently rely on Ikaria for transition services and may be unable to make the changes necessary to operate as a stand-alone company.
- We will need substantial additional funding. Prior to the Spin-Out, we were funded by Ikaria. Going forward, if we are unable to raise capital when needed, we could be forced to delay, reduce or eliminate our product development programs or commercialization efforts.
- We are dependent on the success of our INOpulse and BCM product candidates and our ability to develop, obtain marketing approval for and successfully commercialize these product candidates. If we are unable to develop, obtain marketing approval for or successfully commercialize our product candidates, either alone or through a collaboration, or experience significant delays in doing so, our business could be materially harmed.
- We rely on Ikaria for our supply of nitric oxide for the clinical trials of our INOpulse product candidates. Ikaria is the sole supplier of nitric oxide. Ikaria's inability to continue manufacturing adequate supplies of nitric oxide, or its refusal to supply us with commercial quantities of nitric oxide on commercially reasonable terms, or at all, could result in a disruption in the supply of, or impair our ability to market, our INOpulse product candidates
- Clinical trials involve a lengthy and expensive process with an uncertain outcome. We may incur additional costs or experience delays in completing, ultimately be unable to complete, the development and commercialization of our product candidates.
- Our BCM product candidate is exclusively licensed from BioLine, and we may enter into additional agreements to in-license technology from third parties. If we fail to comply with our obligations under any such license agreements, we could lose rights that are important to our business. BioLine has raised concerns with respect to our performance under the terms of our license agreement, as well as certain alleged breaches under such agreement, and may bring suit against us.
- We may seek to enter into collaborations with third parties for the development and commercialization of our product candidates. If we fail to enter int such collaborations, or such collaborations are not successful, we may not be able to capitalize on the market potential of our product candidates.

- If we are unable to obtain and maintain patent protection for our technology and products or if the scope of the patent protection obtained is not sufficiently broad, our competitors could develop and commercialize technology and products similar or identical to ours, and our ability to successfully commercialize our technology and products may be impaired.

- We will be a "controlled company" within the meaning of the NASDAQ rules and, as a result, will rely on exemptions from certain corporate governance requirements that provide protection to stockholders of other companies. In addition, our principal stockholders own a significant percentage of our stock and will be able to exercise significant influence over matters subject to stockholder approval.

Corporate Structure and Company Conversion

We were incorporated under the laws of the State of Delaware on October 17, 2013 under the name Ikaria Development LLC. We changed our name to Bellerophon Therapeutics LLC on January 27, 2014. We currently have three wholly-owned subsidiaries: Bellerophon BCM LLC, a Delaware limited liability company; Bellerophon Pulse Technologies LLC, a Delaware limited liability company; and Bellerophon Services, Inc., a Delaware corporation.

Prior to the effectiveness of the registration statement of which this prospectus forms a part, we will complete transactions pursuant to which we will convert Bellerophon Therapeutics LLC from a Delaware limited liability company into a Delaware corporation named Bellerophon Therapeutics, Inc. See "Company Conversion."

Our Principal Equity Investors

Our principal stockholders are New Mountain Partners II (AIV-A), L.P., Allegheny New Mountain Partners, L.P. and New Mountain Affiliated Investors II, L.P., which we refer to collectively as the New Mountain Entities; IRDO Holding Corp., or ARCH; Venrock IK Holdings BT, Inc., or Venrock; Linde North America, Inc., an indirect wholly-owned subsidiary of Linde AG, or Linde; and 5AM-BT, Inc. and Aravis Venture I L.P., which we refer to collectively as the 5AM Entities. We refe to the New Mountain Entities, ARCH, Venrock, Linde and the 5AM Entities collectively as the Controlling Entities.

Following the completion of this offering, the Controlling Entities will own shares of our common stock, which collectively will represent approximately % of our outstanding common stock (or % of our outstanding common stock, if the underwriters exercise in full their option to purchase additional shares from us). The Controlling Entities are party to a voting agreement that will remain in effect following this offering pursuant to which such entities have the right to designate a total of six directors and have agreed to vote their shares for such directors. See "Certain Relationships and Related Person Transactions–Agreements with the Controlling Entities—Voting Agreement."

As a result of the voting agreement, we expect that, following the completion of this offering, the Controlling Entities will be deemed to hold their shares of our common stock as part of a group. In addition, upon completion of this offering, we anticipate that the Controlling Entities will continue to control a majority of our outstanding capital stock and will be able to elect a majority of our directors. As a result, we will be a "controlled company" under the rules established by The NASDAQ Global Market and will qualify for, and intend to rely on, the "controlled company" exception to the board of directors and certain committee composition requirements regarding independence under the rules of The NASDAQ Global Market.

In addition, following completion of this offering, the New Mountain Entities will continue to have approval rights over many corporate actions. See "Certain Relationships and Related Person Transactions—Stockholders Agreement."

Corporate Information

Our executive offices are located at Perryville III Corporate Park, 53 Frontage Road, Suite 301, Hampton, New Jersey 08827, and our telephone number is (908) 574-4770.

Implications of Being an Emerging Growth Company

We qualify as an "emerging growth company" as defined in the Jumpstart Our Business Startups Act of 2012, or the JOBS Act. As an emerging growth company we are permitted and intend to rely on exemptions from specified disclosure and other requirements that are applicable to other public companies that are not emergin growth companies. These exemptions include:

- being permitted to provide only two years of audited financial statements, in addition to any required unaudited interim financial statements, with correspondingly reduced "Management's Discussion and Analysis of Financial Condition and Results of Operations" disclosure;
- not being required to comply with the auditor attestation requirements in the assessment of our internal control over financial reporting;
- not being required to comply with any requirement that may be adopted by the Public Company Accounting Oversight Board regarding mandatory audit firm rotation or a supplement to the auditor's report providing additional information about the audit and the financial statements;
- reduced disclosure obligations regarding executive compensation; and
- exemptions from the requirements of holding a nonbinding advisory vote on executive compensation and shareholder approval of any golden parachut payments not previously approved.

We may take advantage of these exemptions for up to five years or such earlier time that we are no longer an emerging growth company. We would cease to be a emerging growth company if we have more than $1.0 billion in annual revenue, we have more than $700.0 million in market value of our stock held by non-affiliates or we issue more than $1.0 billion of non-convertible debt over a three-year period. We may choose to take advantage of some or all of the available exemptions. We have taken advantage of certain reduced reporting burdens in this prospectus. Accordingly, the information contained herein may be different than the information yo receive from other public companies in which you hold stock.

In addition, the JOBS Act provides that an emerging growth company can take advantage of an extended transition period for complying with new or revised accounting standards. This provision allows an emerging growth company to delay the adoption of certain accounting standards until those standards would otherwise apply to private companies. We have irrevocably elected not to avail ourselves of this exemption from new or revised accounting standards, and therefore, we will be subject to the same new or revised accounting standards as other public companies that are not emerging growth companies.

THE OFFERING

Common stock offered	shares
Common stock to be outstanding after this offering	shares
Over-allotment option	The underwriters have an option for a period of 30 days to purchase up to additional shares of our common stock to cover over-allotments.
Use of proceeds	We estimate that the net proceeds from this offering, after deducting underwriting discounts and commissions and estimated offering expenses payabl by us, will be approximately $ million, or approximately $ million if the underwriters exercise their over-allotment option to purchase additional shares from us in full, assuming an initial public offering price of $, the midpoint of the estimated price range set forth on the cover page of this prospectus. We intend to use the net proceeds from this offering, together with our cash, cash equivalents and restricted cash as of March 31, 2014, to fund through completion our ongoing Phase 2 clinical trials of INOpulse for PAH and INOpulse for PH-COPD and our ongoing feasibility trial of BCM, to fund our planned Phase 3 clinical trial program of INOpulse for PAH and for working capital and other general corporate purposes. See "Use of Proceeds" for more information.
Risk factors	You should read the "Risk Factors" section beginning on page 11 of this prospectus for a discussion of factors to consider carefully before deciding to invest in shares of our common stock.
Proposed NASDAQ Global Market symbol	"BLPH"

The number of shares of our common stock to be outstanding after this offering is based on 98,945,820 voting and non-voting shares of our common stock outstanding immediately prior to the closing of this offering and excludes:

- 7,744,480 shares of common stock issuable upon the exercise of stock options outstanding as of April 30, 2014 at a weighted average exercise price of $0.58 per share;
- additional shares of our common stock that will be available for future issuance under our 2014 equity incentive plan; and
- additional shares of our common stock that will be available for future issuance, as of the closing of this offering, under our public company stock incentive plan.

Unless otherwise indicated, all information in this prospectus:

- assumes no exercise of the outstanding options described above;
- assumes no exercise of the underwriters option to purchase additional shares;
- gives effect to the Company Conversion as described under "Company Conversion";
- assumes the conversion of all of the outstanding shares of our non-voting common stock into common stock upon the closing of this offering; and
- assumes the filing of our certificate of incorporation and the adoption of our bylaws upon the closing of this offering.

SUMMARY FINANCIAL INFORMATION

The following summary financial information as of and for the years ended December 31, 2013 and 2012 and for the period from August 26, 2009 (inception) to December 31, 2013 (as we are a development stage company) has been derived from our audited financial statements as of and for the years ended December 31, 201 and 2012 and for the period from August 26, 2009 (inception) to December 31, 2013 included elsewhere in this prospectus. The summary financial data below should be read in conjunction with our historical and pro forma financial statements and the related notes included elsewhere in this prospectus, as well as the "Selected Financial Information" and "Management's Discussion and Analysis of Financial Condition and Results of Operations" sections of this prospectus.

Our financial statements include allocations of costs from certain shared functions provided to us by Ikaria, including general corporate and shared services expenses. These allocations were made either based on specific identification or the proportionate percentage of employee time or headcount to the respective total Ikaria employee time or headcount, as applicable, and have been included in our financial statements.

The financial statements included in this prospectus may not necessarily reflect our financial position, results of operations and cash flows as if we had operated as a stand-alone company during all of the periods presented. Our historical results for any prior period are not necessarily indicative of results to be expected in any future period.

	Year Ended December 31,		Period from August 26, 2009 (inception) to December 31, 2013
(in thousands)	2013	2012	
Statement of Operations and Comprehensive Loss Information:			
Operating expenses:			
Research and development	$ 52,985	$ 38,727	$ 147,887
General and administrative	9,013	7,185	27,690
Other operating expense	—	315	938
Net loss and comprehensive loss	$ (61,998)	$ (46,227)	$ (176,515)

	As of December 31,		As of December 31, 2013	
(in thousands)	2013	2012	Pro Forma(1)	Pro Forma As Adjusted(2)
Balance Sheet Information:			(unaudited)	(unaudited)
Cash and cash equivalents	$ —	—	61,500	
Restricted cash(3)	—	—	18,500	
Working (deficit) capital	(12,440)	(10,892)	60,149	
Total assets	3,636	3,349	83,636	
Allocated portion of Ikaria special dividend bonus payable	4,273	2,865	—	
Other non-current liabilities	1,108	389	1,108	
Total invested (deficit) equity	(15,737)	(11,116)	70,375	

(1) The pro forma balance sheet data (i) gives effect to Ikaria's distribution of $80.0 million of cash to us and the payment of the allocated portion of the Ikaria special dividend bonus amount in respect of certain Ikaria dividend equivalent rights that are described in our financial statements included elsewhere in this prospectus and (ii) does not give effect to the Company Conversion.

(2) The pro forma as adjusted balance sheet data gives further effect to (i) the Company Conversion, (ii) our issuance and sale of shares our of common stock in this offering at an assumed initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us and (iii) the conversion of all of our outstanding non-voting common stock to voting common stock.

(3) Represents cash deposited into escrow to pay amounts owed under the transition services agreement with Ikaria.

A $1.00 increase (decrease) in the assumed initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, would increase (decrease) the pro forma as adjusted amount of each of cash and cash equivalents, working (deficit) capital, total assets and total invested (deficit) equity by approximately $ million, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. Each increase (decrease) of 1.0 million shares in the number of shares of common stock offered by us at the assumed initial public offering price would increase (decrease) the pro forma as adjusted amount of each of cash and cash equivalents, working (deficit) capital, total assets and total invested (deficit) equity by $ million, assuming an initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

RISK FACTORS

Investing in our common stock involves a high degree of risk. Before you decide to invest in our common stock, you should carefully consider the risks and uncertainties described below, together with the other information contained in this prospectus, including our financial statements and the related notes appearing at the end of this prospectus. If any of the following risks occur, our business, financial condition, results of operations and prospects could be materially and adversely affected. In these circumstances, the market price of our common stock could decline, and you might lose all or part of your investment.

Risks Related to Our Financial Position and Need for Additional Capital

We have incurred significant losses since inception. We expect to incur losses over the next several years and may never achieve or maintain profitability.

Since inception, we have incurred significant operating losses. Our net loss was approximately $62.0 million for the year ended December 31, 2013. As of December 31, 2013, we had a deficit accumulated during the development stage of $176.5 million. We do not know whether or when we will become profitable. We have not generated any revenues to date from product sales. We have not completed development of any product candidate and have devoted substantially all of our financial resources and efforts to research and development, including pre-clinical studies and clinical trials. We expect to continue to incur significant expenses and operating losses over the next several years. Our net losses may fluctuate significantly from quarter to quarter and year to year. Net losses and negative cash flows have had, and will continue to have, an adverse effect on our deficit and working capital. We anticipate that our expenses will increase substantially if and as we:

- continue our research and clinical development of our inhaled nitric oxide product candidates using our proprietary pulsatile technology, which we refer to as our INOpulse product candidates, for the treatment of pulmonary arterial hypertension, or PAH, and pulmonary hypertension associated with chronic obstructive pulmonary disease, or PH-COPD, and of bioabsorbable cardiac matrix, or BCM, our product candidate for the prevention of left ventricular remodeling following acute myocardial infarction, or AMI;
- identify, develop and/or in-license additional product candidates;
- seek regulatory approvals for any product candidates that successfully complete clinical trials;
- in the future, establish a manufacturing, sales, marketing and distribution infrastructure;
- maintain, expand and protect our intellectual property portfolio;
- add equipment and physical infrastructure to support our research and development;
- hire additional clinical, regulatory, quality control and scientific personnel; and
- add operational, financial and management information systems and personnel, including personnel to support our product development, any future commercialization efforts and our transition to a public company.

To become and remain profitable, we must succeed in developing and eventually commercializing products that generate significant revenue. We do not expect to generate significant revenue unless and until we are able to obtain marketing approval for, and successfully commercialize, one or more of our product candidates. This will require us to be successful in a range of challenging activities, including completing pre-clinical studies and clinical trials of our product candidates, discovering additional product candidates, obtaining regulatory approval for these product candidates, manufacturing, marketing and selling any products for which we may obtain regulatory approval, satisfying any post-marketing requirements and obtaining reimbursement for our products from private insurance or government payors. We are in the early stages of most of these activities and have not yet commenced other of these activities. We may never succeed in these activities and, even if we do, may never generate revenues that are significant enough to achieve profitability.

Because of the numerous risks and uncertainties associated with pharmaceutical product development, we are unable to accurately predict the timing or amount of increased expenses or when, or if, we will be able to achieve profitability. If we are required by the U.S. Food and Drug Administration, or the FDA, or the European Medicines Agency, or the EMA, to perform studies in addition to those currently expected, or if there are any delays in completing our clinical trials or the development of any of our product candidates, our expenses could increase.

Even if we do achieve profitability, we may not be able to sustain or increase profitability on a quarterly or annual basis. Our failure to become and remain profitable would depress the value of our company and could impair our ability to raise capital, expand our business, maintain our research and development efforts, diversify our product offerings or even continue our operations. A decline in the value of our company could cause you to lose all or part of your investment.

Our very limited operating history as a stand-alone company may make it difficult for you to evaluate the success of our business to date and to assess our future viability.

We were formed as a wholly-owned subsidiary of Ikaria, Inc., or Ikaria, in October 2013 and became a stand-alone company in February 2014 following our spin-out from Ikaria, which we refer to as the Spin-Out, and, as such, have a very limited operating history as a stand-alone company. Prior to the Spin-Out, Ikaria assisted us by providing financing and certain corporate functions. Following the Spin-Out, Ikaria has no obligation to provide assistance to us other than on an interim basis as provided for in the agreements we entered into in connection with the Spin-Out. See "Certain Relationships and Related Person Transactions—Relationship with Ikaria."

Our operations to date have been limited to organizing and staffing our company, developing and securing our technology, and undertaking pre-clinical studies and clinical trials of our product candidates. We have not yet demonstrated the ability to successfully operate as a stand-alone company or to complete development of any product candidates, obtain marketing approvals, manufacture a commercial scale product, or arrange for a third party to do so on our behalf, or conduct sales and marketing activities necessary for successful product commercialization. Consequently, any predictions you make about our future success or viability may not be as accurate as they could be if we had a longer operating history or a history of successfully developing and commercializing products.

Assuming we obtain marketing approval for any of our product candidates, we will need to transition from a company with a research and development focus to a company capable of supporting commercial activities or we will need to enter into strategic partnerships. We may encounter unforeseen expenses, difficulties, complications and delays and may not be successful in such a transition.

We will need substantial additional funding. If we are unable to raise capital when needed, we could be forced to delay, reduce or eliminate our product development programs or commercialization efforts.

We expect our expenses to increase in connection with our ongoing activities, particularly as we initiate additional clinical trials of our INOpulse and BCM product candidates and continue research and development and seek regulatory approval for these and potentially other product candidates. In addition, if we obtain regulatory approval for any of our product candidates, we expect to incur significant commercialization expenses related to product manufacturing, marketing, sales and distribution. In particular, the costs that may be required for the manufacture of any product candidate that receives marketing approval may be substantial. Furthermore, upon the closing of this offering, we expect to incur additional costs associated with operating as a public company. From the inception of our business through December 31, 2013, Ikaria invested $176.5 million in the development of our product candidates, but following the Spin-Out, we will no longer be funded by Ikaria. Accordingly, we will need to obtain substantial additional funding in connection with our continuing operations. If we are unable to raise capital when needed or on attractive terms, we could be forced to delay, reduce or eliminate our research and development programs or any future commercialization efforts.

We plan to use the net proceeds from this offering primarily to fund our ongoing research and development efforts. We will be required to expend significant funds in order to advance development of our INOpulse and BCM product candidates and any other potential product candidates. The net proceeds from this offering and our existing cash, cash equivalents and restricted cash will not be sufficient to fund all of the efforts that we plan to undertake, such as the development of our INOpulse for PH-COPD product candidate following completion of the ongoing Phase 2 clinical trial, or to fund completion of clinical development or commercialization of any of our product candidates. Accordingly, we will be required to obtain further funding through public or private equity offerings, debt financings, collaborations or licensing arrangements or other sources. Adequate additional funding may not be available to us on acceptable terms or at all. Our failure to raise capital as and when needed would have a negative impact on our financial condition and our ability to pursue our business strategy.

We believe that the net proceeds from this offering, together with our existing cash, cash equivalents and restricted cash as of March 31, 2014, will enable us to fund our operating expenses and capital expenditure requirements, as set forth below under "Use of Proceeds," for at least the next months. We have based this estimate on assumptions that may prove to be wrong, and we could use our capital resources sooner than we currently expect. Our future capital requirements will depend on many factors, including:

- the progress and results of our current and planned clinical trials of INOpulse and BCM;
- the costs, timing and outcome of regulatory review of our product candidates;
- the costs of operating as a stand-alone company;
- the cost and timing of future commercialization activities, including product manufacturing, marketing, sales and distribution, for any of our product candidates for which we receive marketing approval;
- the revenue, if any, received from commercial sales of any product candidates for which we receive marketing approval;
- our ability to establish and maintain strategic partnerships, licensing or other arrangements and the financial terms of such agreements;
- the costs and timing of preparing, filing and prosecuting patent applications, maintaining and enforcing our intellectual property rights and defending any intellectual property-related claims;
- the scope, progress, results and costs of discovery, pre-clinical development and clinical trials for any other product candidates;
- the extent to which we acquire or in-license other product candidates and technologies;
- our headcount growth and associated costs; and
- the costs of operating as a public company.

Identifying potential product candidates and conducting pre-clinical studies and clinical trials is a time-consuming, expensive and uncertain process that takes years to complete, and we may never generate the necessary data or results required to obtain regulatory approval and achieve product sales. In addition, our product candidates, if approved, may not achieve commercial success. Our commercial revenues, if any, will be derived from sales of products that we do not expect to be commercially available for several years, if at all. Accordingly, we will need to continue to rely on additional financing to achieve our business objectives. Adequate additional financing may not be available to us on acceptable terms, or at all. In addition, we may seek additional capital due to favorable market conditions or strategic considerations, even if we believe we have sufficient funds for our current or future operating plans.

Raising additional capital may cause dilution to our stockholders, including purchasers of our common stock in this offering, restrict our operations or require us to relinquish rights to technologies or product candidates.

Until such time, if ever, as we can generate substantial product revenues, we expect to finance our cash needs through a combination of public or private equity offerings, debt financings and/or license and development agreements with collaboration partners. We do not have any committed external source of funds. To the extent that we raise additional capital through the sale of equity or convertible debt securities, your ownership interest may be materially diluted, and the terms of such securities could include liquidation or other preferences that adversely affect your rights as a common stockholder. Debt financing and preferred equity financing, if available, may involve agreements that include restrictive covenants that limit our ability to take specified actions, such as incurring additional debt, making capital expenditures or declaring dividends.

If we raise funds through collaborations, strategic partnerships or marketing, distribution or licensing arrangements with third parties, we may have to relinquish valuable rights to our technologies, future revenue streams, research programs or product candidates or grant licenses on terms that may not be favorable to us. If we are unable to raise additional funds through equity or debt financings when needed, we may be required to delay, limit, reduce or terminate our product development or future commercialization efforts or grant rights to develop and market product candidates that we would otherwise prefer to develop and market ourselves.

Risks Related to Our Business and Industry

We may be unable to make, on a timely or cost-effective basis, the changes necessary to operate as a stand-alone company, and we may experience increased or unexpected costs after the Spin-Out or as a result of the Spin-Out.

We have historically operated as part of Ikaria's broader corporate organization, and Ikaria has assisted us by providing certain corporate functions. However, following the Spin-Out, Ikaria is contractually obligated to provide to us only those services specified in the transition services agreement, or the TSA, and the other agreements we entered into with Ikaria to govern our relationship following the Spin-Out. See "Certain Relationships and Related Person Transactions—Relationship with Ikaria" for a summary of these agreements. The TSA provides for certain services to be provided for 24 months from February 2014. We may be unable to replace in a timely manner or on comparable terms the services or other benefits that Ikaria previously provided to us that are not specified in the TSA or the other agreements. Also, upon the expiration of the terms of the required services under the TSA or other agreements, such services will be provided internally or by unaffiliated third parties, and we expect that in some instances, we will incur higher costs to obtain such services than we incurred under the terms of such agreements. Ultimately, we may be unable to replace in a timely manner or on comparable terms the services specified in such agreements. In addition, during the transitional services period, we will rely, in part, on the same executive team at Ikaria that also will continue to manage the business of Ikaria during such time, and there may be conflicting demands on their time, which could result in an inadequate level of attention to the demands of our business. If Ikaria does not continue to perform effectively the transition services and the other services that are called for under the TSA and other agreements, we may not be able to operate our business effectively and our business and financial condition could be adversely affected.

Prior to the Spin-Out, we utilized the executive management team and administrative resources of Ikaria. Many daily functions were performed by Ikaria, including those related to the preparation of our financial statements and the engagement of auditors to audit our financial statements, which have become our responsibility following the Spin-Out. We may need to acquire assets and resources in addition to those provided to us by Ikaria, and we may face difficulty in integrating newly acquired assets into our business. Additionally, as a stand-alone company, we no longer have access to Ikaria's financial resources. Instead, our ability to fund our capital needs will depend on our ongoing ability to

generate cash from operations, enter into partnering arrangements and to access capital markets, which are subject to general economic, financial, competitive, regulatory and other factors that are beyond our control. Our business, financial condition and results of operations could be harmed, possibly materially, if we have difficulty operating as a stand-alone company, fail to acquire necessary capital or assets that prove to be important to our operations, or are unable to enter into partnering or other business development arrangements.

We also anticipate that we will incur additional incremental expenses associated with being a stand-alone company. These incremental pretax expenses are estimated to be approximately $8.0 million to Ikaria under the TSA and additional expenses of $5.0 million for the year ending December 31, 2014.

Our historical and pro forma financial information is not necessarily representative of the results we would have achieved as a stand-alone company and may not be a reliable indicator of our future results.

The historical financial and pro forma financial information we have included in this prospectus may not reflect what our results of operations, financial position and cash flows would have been had we been a stand-alone company during the periods presented. This is primarily because:

- our historical financial information reflects allocations for services historically provided to us by Ikaria, which allocations may not reflect the costs we will incur for similar services in the future as a stand-alone company; and

- our historical financial information does not reflect changes that we expect to incur in the future as a result of our separation from Ikaria and from reduced economies of scale, including changes in the cost structure, personnel needs, financing and operations of our business.

In addition, the pro forma financial information included in this prospectus is based on the best information available, which in part includes a number of estimates and assumptions. These estimates and assumptions may prove not to be accurate, and accordingly, our pro forma financial information should not be assumed to be indicative of what our financial condition or results of operations actually would have been as a stand-alone company, nor to be a reliable indicator of what our financial condition or results of operations actually may be in the future.

Following this offering, we also will be responsible for the additional costs associated with being a public company, including costs related to corporate governance and having listed and registered securities. Therefore, our financial statements may not be indicative of our future performance as a stand-alone public company.

For additional information about our past financial performance and the basis of presentation of our financial statements, please see "Summary Financial Information," "Management's Discussion and Analysis of Financial Condition and Results of Operations" and our financial statements and the notes thereto included elsewhere in this prospectus.

The ownership by certain of our executive officers and directors of shares of common stock, options or other equity awards of Ikaria, as well as the continued roles of certain of our directors with Ikaria, may create, or may create the appearance of, conflicts of interest.

The ownership by certain of our executive officers and directors of shares of common stock, options or other equity awards of Ikaria may create, or may create the appearance of, conflicts of interest. Because of their current or former positions with Ikaria, certain of our executive officers and directors own shares of Ikaria common stock, options to purchase shares of Ikaria common stock or other equity awards. The individual holdings of common stock, options to purchase common stock or other equity awards of Ikaria, may be significant for some of these persons compared to such person's total assets. Ownership by certain of our executive officers and directors of common stock, options to purchase common stock or other equity awards of Ikaria creates, or may create the appearance of, conflicts of interest when these officers or directors are faced with decisions that could have different

implications for Ikaria than the decisions have for us. In addition, certain of our directors are currently in service for both our company as well as Ikaria, and we expect that following the consummation of this offering those directors will remain on the board of directors of both companies. The continued service or employment relationships at both companies creates, or may create the appearance of, conflicts of interest when these executive officers or directors are faced with decisions that could have different implications for Ikaria than the decisions have for us, such as the allocation of time and resources to the provision of transitional services to us by Ikaria pursuant to the TSA and other agreements.

We face substantial competition from other pharmaceutical, biotechnology and medical device companies and our operating results may suffer if we fail to compete effectively.

The pharmaceutical, biotechnology and medical device industries are highly competitive. There are many pharmaceutical, biotechnology and medical device companies, public and private universities and research organizations actively engaged in the research and development of products that may be similar to our product candidates. In addition, other companies are increasingly looking at the cardiopulmonary and cardiac disease market as a potential opportunity. Currently, there are 12 drugs approved for the treatment of PAH, within the following categories: prostacyclin analogs (including Flolan® (epoprostenol), which is marketed by GlaxoSmithKline, Tyvaso® (teprostinil), Orenitram® (treprostinil) and Remodulin® (teprostinil), which are marketed by United Therapeutics Corporation, and Ventavis® (iloprost) and Velentri® (epoprostenol), which are marketed by Actelion Pharmaceuticals, Inc., or Actelion), phosphodiesterase type-5 inhibitors (including Adcirca® (tadalafil), which is marketed by United Therapeutics Corporation, and Revatio® (sildenafil), which is marketed by Pfizer, Inc.), endothelin receptor antagonists (including Letairis® (ambrisentan), which is marketed by Gilead Sciences, Inc., and Opsumit® (macitentan) and Tracleer® (bosentan), which are marketed by Actelion) and a soluble guanylate cyclase stimulator (Adempas® (riociguat), which is marketed by Bayer Healthcare). One additional unapproved drug, Actelion's selexipag, is currently in Phase 3 clinical development. There are also other treatments in Phase 1 and Phase 2 clinical development, including other nitric oxide generation and delivery systems, including GeNO LLC's GeNOsyl™.

For PH-COPD, there are no therapies other than long-term oxygen therapy, pulmonary rehabilitation and lung transplant, and we are not aware of any therapies for PH-COPD in advanced clinical development.

There are no generally accepted products approved for structural support to prevent cardiac remodeling following an AMI. Other product candidates currently in clinical development include stem cell therapies to restore heart muscle cells following an AMI, with large Phase 3 trials expected to end in 2017 or 2018. While we do not believe BCM will directly compete with devices that are used to treat congestive heart failure, certain of such devices are currently in development and designed for administration during open heart surgery or by intra-thoracic injection. These include mesh restraining devices, for example HeartNet™; injectable biopolymers, for example Algisyl-LVR™; and implantable electro-stimulation devices, for example CardioFit™. In addition, volume reduction surgery or cardiac assist devices, or pumps, are sometimes used to treat patients with congestive heart failure.

Many of our competitors, either alone or through their strategic partners, have substantially greater name recognition and financial, technical, manufacturing, marketing and human resources than we do and significantly greater experience and infrastructure in the research and clinical development of medical products, obtaining FDA and other regulatory approvals of those products, and commercializing those products around the world. Additional mergers and acquisitions in the pharmaceutical, biotechnology and medical device industries may result in even more resources being concentrated in our competitors. Large pharmaceutical and medical device companies in particular have extensive expertise in pre-clinical and clinical testing and in obtaining regulatory approvals for medical products. In addition, academic institutions, government agencies, and other public and private organizations conducting research may seek patent protection with respect to potentially competitive

products or technologies. These organizations may also establish exclusive collaborative or licensing relationships with our competitors. Accordingly, our competitors may be more successful than we may be in obtaining approval for inhaled nitric oxide products and achieving widespread market acceptance. We anticipate that we will face intense and increasing competition as new products and technologies become available.

We will not be able to compete effectively unless we successfully:

- design, develop and commercialize products that are superior to other products in the market;
- attract qualified scientific, medical, sales and marketing, engineering and commercial personnel;
- obtain patent and/or other proprietary protection for our processes and product candidates; and
- obtain required regulatory approvals.

Risks Related to the Discovery, Development and Commercialization of Our Product Candidates

We are dependent on the success of our INOpulse and BCM product candidates and our ability to develop, obtain marketing approval for and successfully commercialize these product candidates. If we are unable to develop, obtain marketing approval for or successfully commercialize our product candidates, either alone or through a collaboration, or experience significant delays in doing so, our business could be materially harmed.

We currently have no products approved for sale and have invested a significant portion of our efforts and financial resources in the development of our INOpulse for PAH, INOpulse for PH-COPD and BCM product candidates. Our prospects are substantially dependent on our ability to develop, obtain marketing approval for and successfully commercialize these product candidates. The success of our product candidates will depend, among other things, on our ability to successfully complete clinical trials of each product candidate. The clinical trial process is uncertain, and failure of one or more clinical trials can occur at any stage of testing.

In addition to the successful completion of clinical trials, the success of our product candidates will also depend on several other factors, including the following:

- receipt of marketing approvals from the FDA or other applicable regulatory authorities;
- the performance of our future collaborators for one or more of our product candidates, if any;
- the extent of any required post-marketing approval commitments to applicable regulatory authorities;
- establishment of supply arrangements with third-party raw materials suppliers and manufacturers;
- establishment of arrangements with third-party manufacturers to obtain finished drug products that are appropriately packaged for sale;
- obtaining and maintaining patent, trade secret protection and regulatory exclusivity, both in the United States and internationally;
- protection of our rights in our intellectual property portfolio;
- launch of commercial sales if and when our product candidates are approved;
- a continued acceptable safety profile of our product candidates following any marketing approval;
- commercial acceptance, if and when approved, by patients, the medical community and third-party payors;
- establishing and maintaining pricing sufficient to realize a meaningful return on our investment; and
- competition with other products.

If we are unable to develop, receive marketing approval for, or successfully commercialize our product candidates, or experience delays as a result of any of these factors or otherwise, our business could be materially harmed.

We rely on Ikaria for our supply of nitric oxide for the clinical trials of our INOpulse product candidates. Ikaria is the sole supplier of nitric oxide. Ikaria's inability to continue manufacturing adequate supplies of nitric oxide, or its refusal to supply us with commercial quantities of nitric oxide on commercially reasonable terms, or at all, could result in a disruption in the supply of, or impair our ability to market, our INOpulse product candidates.

We have entered into a drug clinical supply agreement with Ikaria, pursuant to which Ikaria will manufacture and supply our requirements for nitric oxide for inhalation and corresponding placebo for use in clinical trials of our INOpulse product candidates. Ikaria manufactures pharmaceutical-grade nitric oxide at its facility in Port Allen, Louisiana, which is the only FDA-inspected site for manufacturing pharmaceutical-grade nitric oxide in the world. Ikaria's Port Allen facility is subject to the risks of a natural disaster or other business disruption. We maintain under controlled storage conditions a two- to three-month supply of clinical trial drug product, but there can be no assurance that we would be able to meet our requirements for our INOpulse product candidates if there were a catastrophic event or failure of Ikaria's manufacturing system. Because Ikaria's Port Allen facility is the only FDA-inspected site that can manufacture our INOpulse product candidates and because the manufacture of a pharmaceutical gas requires specialized equipment and expertise, there are few, if any, third-party manufacturers to which we could contract this work in a short period of time. Therefore, any disruption in Ikaria's Port Allen facility could materially and adversely affect supplies of our INOpulse product candidates and our ongoing and planned clinical trials. In addition, we do not currently have any arrangements with Ikaria to provide commercial quantities of nitric oxide. If we are unable to arrange for Ikaria to provide such quantities on commercially reasonable terms, or at all, we may not be able to successfully produce and market our INOpulse product candidates or may be delayed in doing so.

We have also entered into a device clinical supply agreement with Ikaria, pursuant to which Ikaria will manufacture and supply our requirements for nitric oxide delivery devices for use in our INOpulse clinical trials. This agreement will expire in February 2015. Ikaria's failure to perform effectively the services that are called for under the device clinical supply agreement could result in a disruption in the supply of, or impair our ability to develop, our INOpulse product candidates. Furthermore, if we are unable to negotiate a new agreement with Ikaria for the manufacture and supply of such delivery devices following the expiration of the device clinical supply agreement or fail to identify and contract with another third-party manufacturer who can meet such requirements, or fail to do so on commercially reasonable terms, our clinical trials and ability to commercialize our product candidates could be materially and adversely affected.

We rely on third-party suppliers and manufacturers to produce and deliver clinical drug supplies for our BCM product candidate and potentially other product candidates. Any failure by a third-party supplier or manufacturer to produce or deliver supplies for us may delay or impair our ability to complete our clinical trials.

We currently rely, and expect to continue to rely, on third parties for supply of the ingredients for our BCM product candidate. The suppliers are, and any future third-party suppliers with whom we enter into agreements may be, our sole suppliers of BCM or any of our other current or future product candidates for a significant period of time. These suppliers are commonly referred to as single-source suppliers. If our suppliers fail to deliver materials and provide services needed for the production of BCM or our other product candidates in a timely and sufficient manner, if they fail to comply with applicable regulations, or if we do not qualify alternate suppliers, clinical development or regulatory approval of our product candidates or commercialization of our products could be delayed, increasing

our costs to complete clinical development and to obtain regulatory approval, which could deprive us of potential additional product revenue.

Reliance on third-party manufacturers entails risks to which we would not be subject if we manufactured product candidates ourselves, including reliance on the third party for regulatory compliance and quality assurance, the possibility of breach of the manufacturing agreement by the third party because of factors beyond our control (including a failure to synthesize and manufacture our product candidates in accordance with our product specifications) and the possibility of termination or nonrenewal of the agreement by the third party, based on its own business priorities, at a time that is costly or damaging to us. In addition, the FDA and other regulatory authorities require that our product candidates be manufactured according to current Good Manufacturing Practices, or cGMP, and similar foreign standards. Any failure by our third-party manufacturers to comply with cGMP or failure to scale up manufacturing processes, including any failure to deliver sufficient quantities of product candidates in a timely manner, could lead to a delay in, or failure to obtain, regulatory approval of any of our product candidates. In addition, such failure could be the basis for action by the FDA to withdraw approvals for product candidates previously granted to us and for other regulatory action, including recall or seizure, fines, imposition of operating restrictions, total or partial suspension of production or injunctions.

We rely on our manufacturers to purchase the materials necessary to produce our product candidates for our clinical studies from third-party suppliers. There are a small number of suppliers for certain capital equipment and raw materials that are used to manufacture our product candidates. Such suppliers may not sell these raw materials to our manufacturers at the times we need them or on commercially reasonable terms. We do not have any control over the process or timing of the acquisition of these raw materials by our manufacturers. Any significant delay in the supply of a product candidate or the raw material components thereof for an ongoing clinical trial due to the need to replace a third-party manufacturer could considerably delay completion or increase the costs of our clinical studies, product testing and potential regulatory approval of our product candidates. If our manufacturers or we are unable to purchase these raw materials after regulatory approval has been obtained for our product candidates, the commercial launch of our product candidates would be delayed or there would be a shortage in supply, which would impair our ability to generate revenues from the sale of our product candidates.

We intend to rely on third parties to produce commercial supplies of any approved product candidates. Any failure by a third-party supplier or manufacturer to produce or deliver supplies for us may delay or impair our ability to commercialize our product candidates.

To date, our product candidates have been manufactured in small quantities for pre-clinical studies and clinical trials. If one or more of our product candidates are approved by the FDA or comparable regulatory authorities in other countries for commercial sale, we will need to manufacture such product candidate in larger quantities. We do not currently have any arrangements with Ikaria or another third-party manufacturer to provide commercial quantities of our product candidates. If we are unable to arrange for such a third-party manufacturing source, or fail to do so on commercially reasonable terms, we may not be able to successfully produce and market our product candidates or may be delayed in doing so.

If we successfully commercialize any of our product candidates, we may be required to establish or access large-scale commercial manufacturing capabilities. We do not own or operate manufacturing facilities for the production of clinical or commercial quantities of our product candidates, and we currently have no plans to build our own clinical or commercial scale manufacturing capabilities.

INOpulse is a sophisticated electro-mechanical device comprised of components that may fail or deteriorate over time or with improper use. If we experience problems with, failure of, or delays in obtaining any INOpulse components, our ability to supply our product candidates in our clinical trials would be adversely affected.

Because INOpulse is a sophisticated electro-mechanical device, the parts which comprise the device are subject to sudden failure or to wear and tear, which may result in decreased function or failure of those parts over time. Although we perform scheduled, preventive maintenance on our drug delivery system to limit device failures, and additional maintenance as needed whenever a user reports a device malfunction, components of our devices may fail. In addition, although we have designed INOpulse to be simple and easy to use and will provide user manuals and other training materials, users of INOpulse may use the devices improperly, which could cause the devices to fail or otherwise not work properly.

There are several components in INOpulse that are custom designed or assembled for us. We are dependent on a single company to supply us with some of these components. While we believe there are alternative suppliers from which we could purchase most of these components, there is a risk that a single-source supplier could fail to deliver adequate supply, or could suffer a business interruption that could affect our supply of these components.

We obtain some of the components for INOpulse through individual purchase orders executed on an as needed basis rather than pursuant to long-term supply agreements. Our business, financial condition or results of operations could be adversely affected if any of our principal third-party suppliers or manufacturers experience production problems, lack of capacity or transportation disruptions or otherwise cease producing such components.

Clinical trials involve a lengthy and expensive process with an uncertain outcome. We may incur additional costs or experience delays in completing, or ultimately be unable to complete, the development and commercialization of our product candidates.

We are currently conducting clinical trials of our INOpulse and BCM product candidates. The risk of failure of all of our product candidates is high. It is impossible to predict when or if any of our product candidates will prove effective or safe in humans or will receive regulatory approval. Before obtaining marketing approval from regulatory authorities for the sale of any product candidate, we must conduct extensive clinical trials to demonstrate the safety and efficacy of our product candidates in humans. Clinical testing is expensive, difficult to design and implement, can take many years to complete and is uncertain as to outcome. A failure of one or more clinical trials can occur at any stage of testing. The clinical development of our product candidates is susceptible to the risk of failure inherent at any stage of development, including failure to demonstrate efficacy in a clinical trial or across a broad population of patients, the occurrence of severe or medically or commercially unacceptable adverse events, failure to comply with protocols or applicable regulatory requirements and determination by the FDA or any comparable non-U.S. regulatory authority that a drug product is not approvable.

It is possible that even if one or more of our product candidates has a beneficial effect, that effect will not be detected during clinical evaluation as a result of one or more of a variety of factors, including the size, duration, design, measurements, conduct or analysis of our clinical trials. Conversely, as a result of the same factors, our clinical trials may indicate an apparent positive effect of a product candidate that is greater than the actual positive effect, if any. Similarly, in our clinical trials we may fail to detect toxicity of or intolerability caused by our product candidates, or mistakenly believe that our product candidates are toxic or not well tolerated when that is not in fact the case.

The outcome of pre-clinical studies and early clinical trials may not be predictive of the success of later clinical trials, and interim results of a clinical trial do not necessarily predict final results. Many companies in the biotechnology and pharmaceutical industries have suffered significant setbacks in late-stage clinical trials after achieving positive results in earlier development, and we cannot be certain that we will not face such setbacks.

The design of a clinical trial can determine whether its results will support approval of a product, and flaws in the design of a clinical trial may not become apparent until the clinical trial is well advanced or completed. We have limited experience in designing clinical trials and may be unable to design and execute a clinical trial to support marketing approval. In addition, pre-clinical and clinical data are often susceptible to varying interpretations and analyses. Many companies that believed their product candidates performed satisfactorily in pre-clinical studies and clinical trials have nonetheless failed to obtain marketing approval for the product candidates. Even if we believe that the results of clinical trials for our product candidates warrant marketing approval, the FDA or comparable non-U.S. regulatory authorities may disagree and may not grant marketing approval of our product candidates.

In some instances, there can be significant variability in safety or efficacy results between different clinical trials of the same product candidate due to numerous factors, including changes in trial procedures set forth in protocols, differences in the size and type of the patient populations, changes in and adherence to the clinical trial protocols and the rate of dropout among clinical trial participants. Any Phase 3 or other clinical trials that we may conduct may not demonstrate the efficacy and safety necessary to obtain regulatory approval to market our product candidates.

We are transitioning our INOpulse delivery system to a next generation device that has not been utilized in our ongoing Phase 2 clinical trials of our INOpulse product candidates. Failure by the FDA or other regulatory authority to support the transition and bridging strategy for our transition to the new device could increase our development costs and/or delay commencement of our planned Phase 3 clinical trials of our INOpulse product candidates.

Our ongoing Phase 2 clinical trials of our INOpulse product candidates utilize the first generation INOpulse DS device. We are near completion of a next generation INOpulse Mark2 device, or the Mark2, and we plan to transition our INOpulse delivery system from INOpulse DS to the Mark2 for any Phase 3 clinical trials of our INOpulse product candidates. We have developed a regulatory bridging strategy to show that the amount and timing of the inhaled nitric oxide delivered is similar across INOpulse device generations. We anticipate discussing data supporting this transition plan and bridging strategy with the FDA at an End-of-Phase 2 meeting that we expect will occur in early 2015. The FDA may not agree that our data support transition to this new device, in which case we may be required to provide additional data, perform a revised bridging assessment or repeat the Phase 2 clinical trial, any of which could increase our development costs and/or delay commencement of these Phase 3 clinical trials. In addition, even if the FDA accepts our transition plan and bridging strategy, use of the Mark2 in the Phase 3 clinical trial could produce results that are different than those we would expect based on the results from the Phase 2 clinical trial using the INOpulse DS device.

We intend to conduct, and may in the future conduct, clinical trials for certain of our product candidates at sites outside the United States, and the FDA may not accept data from trials conducted in such locations.

We have conducted, and may in the future choose to conduct, one or more of our clinical trials outside the United States. For example, our Phase 2 clinical trial of INOpulse for PAH includes sites in Canada and our clinical trial of BCM includes sites in Europe, Canada, Australia and Israel.

Although the FDA may accept data from clinical trials conducted outside the United States, acceptance of this data is subject to certain conditions imposed by the FDA. For example, the clinical trial must be well designed and conducted and performed by qualified investigators in accordance with good clinical practices, or GCP, in the case of drug trials, or the Declaration of Helsinki or the laws and regulations of the country in which the research is conducted, whichever affords greater protection to the human subjects, in the case of device trials. The trial population must also adequately represent the U.S. population, and the data must be applicable to the U.S. population and U.S. medical practice in ways that the FDA deems clinically meaningful. Generally, the patient population for any clinical trials conducted outside of the United States must be representative of the population for whom we

intend to seek approval in the United States. In addition, while these clinical trials are subject to the applicable local laws, FDA acceptance of the data will be dependent upon its determination that the trials also complied with all applicable U.S. laws and regulations. There can be no assurance that the FDA will accept data from trials conducted outside of the United States. If the FDA does not accept the data from our Phase 2 clinical trial of INOpulse for PAH in Canada or our clinical trial of BCM in Europe, Canada, Australia or Israel, or any future trial that we determine to conduct outside the United States, it would likely result in the need for additional trials, which would be costly and time-consuming and delay or permanently halt our development of INOpulse for PAH and BCM or any future product candidates.

In addition, the conduct of clinical trials outside the United States could have a significant impact on us. Risks inherent in conducting international clinical trials include:

- foreign regulatory requirements that could restrict or limit our ability to conduct our clinical trials;
- administrative burdens of conducting clinical trials under multiple foreign regulatory schema;
- foreign exchange fluctuations; and
- diminished protection of intellectual property in some countries.

If clinical trials of our product candidates fail to demonstrate safety and efficacy of our product candidates to the satisfaction of the FDA and comparable non-U.S. regulators, we may incur additional costs or experience delays in completing, or ultimately be unable to complete, the development and commercialization of these product candidates.

We are not permitted to commercialize, market, promote or sell any product candidate in the United States without obtaining marketing approval from the FDA. Comparable non-U.S. regulatory authorities, such as the EMA, impose similar restrictions. We may never receive such approvals. We must complete extensive pre-clinical studies and clinical trials to demonstrate the safety and efficacy of our product candidates in humans before we will be able to obtain these approvals.

Clinical testing is expensive, difficult to design and implement, can take many years to complete and is inherently uncertain as to outcome. We have not previously submitted a new drug application, or an NDA, to the FDA or similar drug approval filings to comparable non-U.S. regulatory authorities for any of our product candidates.

Any inability to successfully complete pre-clinical and clinical development could result in additional costs to us and impair our ability to generate revenues from product sales. In addition, if (1) we are required to conduct additional clinical trials or other testing of our product candidates beyond the trials and testing that we contemplate, (2) we are unable to successfully complete clinical trials of our product candidates or other testing, (3) the results of these trials or tests are unfavorable, uncertain or are only modestly favorable or (4) there are unacceptable safety concerns associated with our product candidates, we, in addition to incurring additional costs, may:

- be delayed in obtaining marketing approval for our product candidates;
- not obtain marketing approval at all;
- obtain approval for indications or patient populations that are not as broad as we intended or desired;
- obtain approval with labeling that includes significant use or distribution restrictions or significant safety warnings, including boxed warnings;

- be subject to additional post-marketing testing or other requirements; or
- be required to remove the product from the market after obtaining marketing approval.

For example, the FDA has asked us to conduct a study to test the environmental impact of using INOpulse at home. When inhaled nitric oxide is administered through INOpulse, a small portion of the nitric oxide will be exhaled or otherwise emitted and could react with oxygen in room air, forming nitrogen dioxide, which is an environmental pollutant. The study will measure the nitrogen dioxide in the room air with use of INOpulse under actual or simulated patient use conditions. If the FDA or other regulatory authority requires us to conduct additional testing or determines that an unacceptable amount of nitrogen dioxide is formed through the use of INOpulse, we may be required to alter the design of INOpulse, which may not be possible, and the clinical development timeline of our INOpulse product candidates may be delayed or prove to be more costly than we currently anticipate.

If we experience any of a number of possible unforeseen events in connection with clinical trials of our product candidates, potential marketing approval or commercialization of our product candidates could be delayed or prevented.

We may experience numerous unforeseen events during, or as a result of, clinical trials that could delay or prevent marketing approval of our product candidates, including:

- clinical trials of our product candidates may produce unfavorable or inconclusive results;
- we may decide, or regulators may require us, to conduct additional clinical trials or abandon product development programs;
- the number of patients required for clinical trials of our product candidates may be larger than we anticipate, patient enrollment in these clinical trials may be slower than we anticipate or participants may drop out of these clinical trials at a higher rate than we anticipate;
- our third-party contractors, including those manufacturing our product candidates or components or ingredients thereof or conducting clinical trials on our behalf, may fail to comply with regulatory requirements or meet their contractual obligations to us in a timely manner or at all;
- regulators or institutional review boards may not authorize us or our investigators to commence a clinical trial or conduct a clinical trial at a prospective trial site;
- we may experience delays in reaching or fail to reach agreement on acceptable clinical trial contracts or clinical trial protocols with prospective trial sites;
- patients who enroll in a clinical trial may misrepresent their eligibility to do so or may otherwise not comply with the clinical trial protocol, resulting in the need to withdraw such patients from the clinical trial, increase the needed enrollment size for the clinical trial or extend the clinical trial's duration;
- we may have to suspend or terminate clinical trials of our product candidates for various reasons, including a finding that the participants are being exposed to unacceptable health risks, undesirable side effects or other unexpected characteristics of a product candidate;
- regulators or institutional review boards may require that we or our investigators suspend or terminate clinical research for various reasons, including noncompliance with regulatory requirements or their respective standards of conduct, a finding that the participants are being exposed to unacceptable health risks, undesirable side effects or other unexpected characteristics of the product candidate or findings of undesirable effects caused by a chemically or mechanistically similar drug or drug candidate;

- the FDA or comparable non-U.S. regulatory authorities may disagree with our clinical trial design or our interpretation of data from pre-clinical studies and clinical trials;

- the FDA or comparable non-U.S. regulatory authorities may find regulatory non-compliance with the manufacturing processes or facilities of third-party manufacturers with which we enter into agreements for clinical and commercial supplies;

- the supply or quality of raw materials or manufactured product candidates or other materials necessary to conduct clinical trials of our product candidates may be insufficient, inadequate or not available at an acceptable cost, or we may experience interruptions in supply; and

- the approval policies or regulations of the FDA or comparable non-U.S. regulatory authorities may significantly change in a manner rendering our clinical data insufficient to obtain marketing approval.

Product development costs for us will increase if we experience delays in testing or pursuing marketing approvals and we may be required to obtain additional funds to complete clinical trials and prepare for possible commercialization of our product candidates. We do not know whether any pre-clinical studies or clinical trials will begin as planned, will need to be restructured or will be completed on schedule, or at all. Significant pre-clinical study or clinical trial delays also could shorten any periods during which we may have the exclusive right to commercialize our product candidates or allow our competitors to bring products to market before we do and impair our ability to successfully commercialize our product candidates and may harm our business and results of operations. In addition, many of the factors that cause, or lead to, clinical trial delays may ultimately lead to the denial of marketing approval of any of our product candidates.

If we experience delays or difficulties in the enrollment of patients in clinical trials, we may not achieve our clinical development on our anticipated timeline, or at all, and our receipt of necessary regulatory approvals could be delayed or prevented.

We may not be able to initiate or continue clinical trials for our INOpulse or BCM product candidates if we are unable to locate and enroll a sufficient number of eligible patients to participate in clinical trials. Patient enrollment is a significant factor in the timing of clinical trials, and is affected by many factors, including:

- the size and nature of the patient population;

- the severity of the disease under investigation;

- the proximity of patients to clinical sites;

- the eligibility criteria for the trial;

- the design of the clinical trial;

- limitations placed on enrollment by regulatory authorities;

- efforts to facilitate timely enrollment;

- competing clinical trials; and

- clinicians' and patients' perceptions as to the potential advantages and risks of the product candidate being studied in relation to other available therapies, including any new product candidates that may be approved for the indications we are investigating.

For example, we may experience difficulty enrolling our clinical trials, including, but not limited to, our clinical trials of INOpulse for the treatment of PAH, which is an orphan disease due to the small number of patients. Enrollment in PRESERVATION I is currently limited by the FDA to a maximum

of 40 patients in the United States and the Israeli Ministry of Health requires submission of additional safety data once 70 patients are enrolled in Israel. These limitations may hinder our ability to complete, or delay completion of, this clinical trial.

Our inability to enroll a sufficient number of patients for our clinical trials could result in significant delays or may require us to abandon one or more clinical trials altogether. Enrollment delays in our clinical trials may result in increased development costs for our product candidates, delay or halt the development of and approval processes for our product candidates and jeopardize our ability to achieve our clinical development timeline and goals, including the dates by which we will commence, complete and receive results from clinical trials. Enrollment delays may also delay or jeopardize our ability to commence sales and generate revenues from our product candidates. Any of the foregoing could cause the value of our company to decline and limit our ability to obtain additional financing, if needed.

We may not obtain orphan drug exclusivity, or we may not receive the full benefit of orphan drug exclusivity even if we obtain such exclusivity.

Regulatory authorities in some jurisdictions, including the United States and European Union, may designate drugs and biologics for relatively small patient populations as orphan drugs. Under the Orphan Drug Act, the FDA may designate a product as an orphan drug if it is a drug or biologic intended to treat a rare disease or condition, which is generally defined as a patient population of fewer than 200,000 individuals in the United States who have been diagnosed as having the disease or condition at the time of the submission of the request for orphan drug designation. The FDA has granted orphan drug designation for nitric oxide for the treatment of PAH. Under the Orphan Drug Act, the first company to receive FDA approval for inhaled nitric oxide in this indication will obtain seven years of marketing exclusivity, during which time the FDA may not approve another company's application for the use of inhaled nitric oxide in the same orphan indication.

Even though we have obtained orphan drug designation for nitric oxide to treat PAH, and even if we obtain orphan drug designation for our future product candidates or other indications, due to the uncertainties associated with developing pharmaceutical products, we may not be the first to obtain marketing approval for any particular orphan indication, or we may not obtain approval for an indication for which we have obtained orphan drug designation. Further, even if we obtain orphan drug exclusivity for a product candidate, that exclusivity may not protect the product effectively from competition because different drugs can be approved for the same condition. Even after an orphan drug is approved, the FDA can subsequently approve the same drug for the same condition if the FDA concludes that the later drug is safer, more effective or makes a major contribution to patient care. Orphan drug designation neither shortens the development time or regulatory review time of a drug, nor gives the drug any advantage in the regulatory review or approval process. Orphan drug exclusivity may be lost if the FDA determines that the request for designation was materially defective or if the manufacturer is unable to assure sufficient quantity of the product to meet the needs of patients with the rare disease or condition.

Serious adverse events or undesirable side effects or other unexpected properties of our product candidates may be identified during development that could delay or prevent the product candidate's marketing approval.

Serious adverse events or undesirable side effects caused by, or other unexpected properties of, our product candidates could cause us, an institutional review board or regulatory authorities to interrupt, delay or halt clinical trials of one or more of our product candidates and could result in a more restrictive label or the delay or denial of marketing approval by the FDA or comparable non-U.S. regulatory authorities. If any of our product candidates is associated with serious adverse events or undesirable side effects or has properties that are unexpected, we may need to abandon development or limit development of that product candidate to certain uses or subpopulations in which the

undesirable side effects or other characteristics are less prevalent, less severe or more acceptable from a risk-benefit perspective. Many drugs or devices that initially showed promise in clinical or earlier stage testing have later been found to cause undesirable or unexpected side effects that prevented further development of the drug or device.

For example, pending discussions with regulatory authorities, we may be required to conduct a drug-drug interaction study. We expect the FDA to require us primarily to study interactions with long-acting beta agonists, which is the only class of COPD drug that has been identified as having potential adverse cardiac side effects, to confirm that pulsed inhaled nitric oxide does not increase systemic bio-availability of inhaled beta agonists. If the results of such a study indicate increased bioavailability that we are not able to address to the satisfaction of the FDA, marketing approval of INOpulse for PH-COPD, if any, may be limited to patients who do not use long-acting beta agonists.

Additionally, our INOpulse product candidates are an extension of the technology that is used in hospitals to deliver inhaled nitric oxide to neonates with a form of pulmonary hypertension called persistent pulmonary hypertension of the newborn. Persistent pulmonary hypertension is an FDA-approved use of inhaled nitric oxide, which is currently marketed by Ikaria as INOmax. Because our INOpulse product candidates draw on the established efficacy and safety of INOmax, if any serious adverse events or undesirable side effects or other unexpected properties of INOmax or other inhaled nitric oxide delivery systems developed by Ikaria are identified, our INOpulse product candidates may be adversely affected and we may be required to interrupt, delay or halt clinical trials of one or both of our INOpulse product candidates.

We may not be successful in our efforts to identify or discover additional potential product candidates.

A significant portion of the research that we are conducting involves the development of innovative approaches to the pulsed delivery of nitric oxide. Our drug-device discovery efforts may not be successful in creating drugs or devices that have commercial value or therapeutic utility. Our research programs may initially show promise in creating potential product candidates, yet fail to yield viable product candidates for clinical development for a number of reasons, including that potential product candidates may, on further study, be shown to have harmful side effects or other characteristics that indicate that they are unlikely to be product candidates that will receive marketing approval and achieve market acceptance. Currently, we are dependent on Ikaria for our business development functions pursuant to the TSA and lack the capability to bring such functions in-house. If Ikaria does not perform such business development functions effectively, our business and prospects may be materially and adversely affected.

Our research programs to identify new product candidates will require substantial technical, financial and human resources. We may be unsuccessful in our efforts to identify new potential product candidates. In addition, we may focus our efforts and resources on one or more potential product candidates that ultimately prove to be unsuccessful.

Additionally, we are limited in the scope of potential product candidates that we can identify or discover due to non-competition agreements that we entered into with Ikaria. Pursuant to the these agreements, we and each of our subsidiaries agreed not to engage, anywhere in the world, in any manner, directly or indirectly, until the earlier of five years after the effective date of such non-competition agreement or the date on which Ikaria and all of its subsidiaries are no longer engaged in such business, in:

- the development, manufacture, commercialization, promotion, sale, import, export, servicing, repair, training, storage, distribution, transportation, licensing, or other handling or disposition of any product or service (including, without limitation, any product or service that utilizes, contains or includes nitric oxide for inhalation, a device intended to deliver nitric oxide or a service that delivers or supports the delivery of nitric oxide), bundled or unbundled, for or used

in connection with (a) the diagnosis, prevention or treatment, in both adult and/or pediatric populations, and whether in- or out-patient, of: (i) hypoxic respiratory failure associated with pulmonary hypertension, (ii) pulmonary hypertensive episodes and right heart failure associated with cardiovascular surgery, (iii) bronchopulmonary dysplasia, (iv) the management of ventilation-perfusion mismatch in acute lung injury, (v) the management of ventilation-perfusion mismatch in acute respiratory distress syndrome, (vi) the management of pulmonary hypertension episodes and right heart failure in congestive heart failure, (vii) pulmonary edema from high altitude sickness, (viii) the management of pulmonary hypertension episodes and right heart failure in pulmonary or cardiac surgery, (ix) the management of pulmonary hypertension episodes and right heart failure in organ transplant, (x) sickle cell vaso-occlusive crisis, (xi) hypoxia associated with pneumonia or (xii) ischemia-reperfusion injury or (b) the use of nitric oxide to treat or prevent conditions that are primarily managed in the hospital; or

- any and all development, manufacture, commercialization, promotion, sale, import, export, storage, distribution, transportation, licensing, or other handling or disposition of any terlipressin or any other product within the pressin family, (a) intended to treat (i) hepatorenal syndrome in any form, (ii) bleeding esophageal varices, or (iii) septic shock, or (b) for or in connection with the management of low blood pressure.

If we are unable to identify suitable additional compounds for pre-clinical and clinical development or at all, our ability to develop product candidates and obtain product revenues in future periods could be compromised, which could result in significant harm to our financial position and adversely impact our stock price.

Even if one of our product candidates receives marketing approval, it may fail to achieve the degree of market acceptance by physicians, patients, third-party payors and others in the medical community necessary for commercial success, and the market opportunity for the product candidate may be smaller than we estimate.

We have never commercialized a product. Even if one of our product candidates is approved by the appropriate regulatory authorities for marketing and sale, it may nonetheless fail to gain sufficient market acceptance by physicians, patients, third-party payors and others in the medical community. For example, physicians are often reluctant to switch their patients from existing therapies even when new and potentially more effective or convenient treatments enter the market. Further, patients often acclimate to the therapy that they are currently taking and do not want to switch unless their physicians recommend switching products or they are required to switch therapies due to lack of reimbursement for existing therapies.

Efforts to educate the medical community and third-party payors on the benefits of our product candidates may require significant resources and may not be successful. If any of our product candidates is approved but does not achieve an adequate level of market acceptance, we may not generate significant revenues and we may not become profitable. The degree of market acceptance of, and potential market opportunity for, our product candidates, if approved for commercial sale, will depend on a number of factors, including:

- the efficacy and safety of the product;
- the potential advantages of the product compared to alternative treatments;
- the prevalence and severity of any side effects;
- the clinical indications for which the product is approved;
- whether the product is designated under physician treatment guidelines as a first-line therapy or as a second- or third-line therapy;
- limitations or warnings, including distribution or use restrictions, contained in the product's approved labeling;

- our ability to offer the product for sale at competitive prices;
- our ability to establish and maintain pricing sufficient to realize a meaningful return on our investment;
- our ability to prevent use of our INOpulse for PH-COPD device by PAH patients due to expected pricing differences;
- the product's convenience and ease of administration compared to alternative treatments;
- the willingness of the target patient population to try, and of physicians to prescribe, the product;
- the strength of sales, marketing and distribution support;
- the approval of other new products for the same indications;
- changes in the standard of care for the targeted indications for the product;
- the timing of market introduction of our approved products as well as competitive products and other therapies;
- availability and amount of reimbursement from government payors, managed care plans and other third-party payors;
- adverse publicity about the product or favorable publicity about competitive products; and
- potential product liability claims.

The potential market opportunities for our product candidates are difficult to estimate precisely. Our estimates of the potential market opportunities, including our estimates with respect to pricing and reimbursement, are predicated on many assumptions, including industry knowledge and publications, third-party research reports and other surveys. While we believe that our internal assumptions are reasonable, these assumptions involve the exercise of significant judgment on the part of our management, are inherently uncertain and the reasonableness of these assumptions has not been assessed by an independent source. If any of the assumptions proves to be inaccurate, the actual markets for our product candidates could be smaller than our estimates of the potential market opportunities.

If any of our product candidates receives marketing approval and we, or others, later discover that the product is less effective than previously believed or causes undesirable side effects that were not previously identified, our ability to market the product could be compromised.

Clinical trials of our product candidates are conducted in carefully defined subsets of patients who have agreed to enter into clinical trials. Consequently, it is possible that our clinical trials may indicate an apparent positive effect of a product candidate that is greater than the actual positive effect, if any, or alternatively fail to identify undesirable side effects. If, following approval of a product candidate, we, or others, discover that the drug is less effective than previously believed or causes undesirable side effects that were not previously identified, any of the following undesirable events could occur:

- regulatory authorities may withdraw their approval of the product or seize the product;
- we may be required to recall the product or change the way the product is administered;
- additional restrictions may be imposed on the marketing of, or the manufacturing processes for, the particular product;
- we may be subject to fines, injunctions or the imposition of civil or criminal penalties;
- regulatory authorities may require the addition of labeling statements, such as a "black box" warning or a contraindication;

- we may be required to create a handout, sometimes referred to as a Medication Guide, outlining the risks of the previously unidentified side effects for distribution to patients;
- we could be sued and held liable for harm caused to patients;
- the product may become less competitive; and
- our reputation may suffer.

Any of these events could have a material and adverse effect on our operations and business and could adversely impact our stock price.

If we are unable to establish sales, marketing and distribution capabilities or enter into acceptable sales, marketing and distribution arrangements with third parties, we may not be successful in commercializing any product candidates that we develop, if and when those product candidates are approved.

We do not have a sales, marketing or distribution infrastructure and have limited experience in the sale, marketing or distribution of pharmaceutical products. To achieve commercial success for any approved product, we must either develop a sales and marketing organization or outsource these functions to third parties. We expect to build a commercial infrastructure to allow us to market and sell any approved product candidates using a specialty sales force in the United States, and we may choose to establish commercialization capabilities in select countries outside the United States. The development of sales, marketing and distribution capabilities will require substantial resources, will be time-consuming and could delay any product launch. We expect that we will commence the development of these capabilities prior to receiving approval of any of our product candidates. If the commercial launch of a product candidate for which we recruit a sales force and establish marketing and distribution capabilities is delayed or does not occur for any reason, we could have prematurely or unnecessarily incurred these commercialization costs. Such a delay may be costly, and our investment could be lost if we cannot retain or reposition our sales and marketing personnel. In addition, we may not be able to hire or retain a sales force in the United States that is sufficient in size or has adequate expertise in the medical markets that we plan to target. If we are unable to establish or retain a sales force and marketing and distribution capabilities, our operating results may be adversely affected. If a potential partner has development or commercialization expertise that we believe is particularly relevant to one of our product candidates, then we may seek to collaborate with that potential partner even if we believe we could otherwise develop and commercialize the product independently.

We may partner with third parties to commercialize our product candidates in certain countries outside the United States. As a result of entering into arrangements with third parties to perform sales, marketing and distribution services, our product revenues or the profitability of these product revenues may be lower, perhaps substantially lower, than if we were to directly market and sell products in those markets. Furthermore, we may be unsuccessful in entering into the necessary arrangements with third parties or may be unable to do so on terms that are favorable to us. In addition, we may have little or no control over such third parties, and any of them may fail to devote the necessary resources and attention to sell and market our product candidates effectively.

If we do not establish sales and marketing capabilities, either on our own or in collaboration with third parties, we will not be successful in commercializing any of our product candidates that receive marketing approval.

Even if we are able to commercialize any product candidate that we develop, the product may become subject to unfavorable pricing regulations, third-party payor reimbursement practices or healthcare reform initiatives that could harm our business.

The commercial success of our product candidates will depend substantially, both in the United States and abroad, on the extent to which the costs of our product candidates will be paid by health maintenance, managed care, pharmacy benefit and similar healthcare management organizations, or

reimbursed by government health administration authorities, private health coverage insurers and other third-party payors. If reimbursement is not available, or is available only to limited levels, we may not be able to successfully commercialize our product candidates. Even if coverage is provided, the approved reimbursement amount may not be high enough to allow us to establish and maintain pricing sufficient to realize a meaningful return on our investment.

There is significant uncertainty related to third-party payor coverage and reimbursement of newly approved drugs and devices. Marketing approvals, pricing and reimbursement for new drug and device products vary widely from country to country. Some countries require approval of the sale price of a drug or device before it can be marketed. In many countries, the pricing review period begins after marketing or product licensing approval is granted. In some non-U.S. markets, pricing remains subject to continuing governmental control even after initial approval is granted. As a result, we might obtain marketing approval for a product in a particular country, but then be subject to price regulations that delay commercial launch of the product, possibly for lengthy time periods, which may negatively impact the revenues we are able to generate from the sale of the product in that country. Adverse pricing limitations may hinder our ability to recoup our investment in one or more product candidates, even if our product candidates obtain marketing approval.

Our ability to commercialize our product candidates will depend in part on the extent to which coverage and reimbursement for these products and related treatments will be available from government health administration authorities, private health insurers and other organizations. Government authorities and third-party payors, such as private health insurers and health maintenance organizations, decide which medications they will cover and establish reimbursement levels. The healthcare industry is acutely focused on cost containment, both in the United States and elsewhere. Government authorities and third-party payors have attempted to control costs by limiting coverage and the amount of reimbursement for particular medications, which could affect our ability to sell our product candidates profitably. These payors may not view our products, if any, as cost-effective, and coverage and reimbursement may not be available to our customers, or may not be sufficient to allow our products, if any, to be marketed on a competitive basis. Cost-control initiatives could cause us to decrease the price we might establish for products, which could result in lower than anticipated product revenues. If the prices for our products, if any, decrease or if governmental and other third-party payors do not provide adequate coverage or reimbursement, our prospects for revenue and profitability will suffer. Approval of a product does not guarantee sufficient reimbursement to commercialize. For example, approval of CE marking for BCM in the European Union may be achieved with our ongoing feasibility trial but this data may not be sufficient to gain sufficient reimbursement for us to invest in commercialization activities.

There may also be delays in obtaining coverage and reimbursement for newly approved products, and coverage may be more limited than the indications for which the product is approved by the FDA or comparable non-U.S. regulatory authorities. Moreover, eligibility for reimbursement does not imply that any product will be paid for in all cases or at a rate that covers our costs, including research, development, manufacture, sale and distribution. Reimbursement rates may vary, by way of example, according to the use of the product and the clinical setting in which it is used. Reimbursement rates may also be based on reimbursement levels already set for lower cost products or may be incorporated into existing payments for other services.

In addition, increasingly, third-party payors are requiring higher levels of evidence of the benefits and clinical outcomes of new technologies and are challenging the prices charged. We cannot be sure that coverage will be available for any product candidate that we commercialize and, if available, that the reimbursement rates will be adequate. Further, the net reimbursement for drug products may be subject to additional reductions if there are changes to laws that presently restrict imports of drugs from countries where they may be sold at lower prices than in the United States. An inability to promptly obtain coverage and adequate payment rates from both government-funded and private payors for any our product candidates for which we obtain marketing approval could have a material adverse effect on our operating results, our ability to raise capital needed to commercialize products and our overall financial condition.

We anticipate that reimbursement of BCM will be based on the patient's diagnosis related group, or DRG, for patients who are covered by Medicare or Medicaid, or through similar reimbursement programs for patients to who are covered by private third-party payors. Within the DRG system, patients are classified by similar diagnoses, which are mapped from the International Statistical Classification of Diseases and Related Health Problems, or ICD, a medical classification list provided by the World Health Organization. The version of ICD that is currently in use with respect to DRG classifications is ICD-9. However, an updated version, ICD-10, has been adopted and we expect that DRG classifications will be required to be mapped against ICD-10 by October 2015. We believe that the DRG classifications will be mapped from ICD-10 rather than ICD-9 at the time we commercialize BCM, if ever, which would result in favorable reimbursement. However, if ICD-9 continues to be used for DRG classification mapping by hospitals or Medicare or Medicaid or other payors, or our expectations with respect to the applicable DRG classification prove incorrect, reimbursement for BCM may prove less favorable or inadequate. In addition, even if ICD-10 is adopted for reimbursement assessments, the mapping to the DRGs may change or the amount reimbursed for the DRGs may change, all of which could adversely affect the ability of our customers to gain sufficient reimbursement, which may affect the adoption of or price we can charge for BCM.

If the FDA or comparable non-U.S. regulatory authorities approve generic versions of any of our products that receive marketing approval, or such authorities do not grant our products appropriate periods of data exclusivity before approving generic versions of our products, the sales of our products could be adversely affected.

Once an NDA is approved, the product covered thereby becomes a "reference listed drug" in the FDA's publication, "Approved Drug Products with Therapeutic Equivalence Evaluations." Manufacturers may seek approval of generic versions of reference listed drugs through submission of abbreviated new drug applications, or ANDAs, in the United States, or through a similar process in foreign jurisdictions. In support of an ANDA, a generic manufacturer need not conduct clinical studies. Rather, the applicant generally must show that its product has the same active ingredient(s), dosage form, strength, route of administration and conditions of use or labeling as the reference listed drug and that the generic version is bioequivalent to the reference listed drug, meaning it is absorbed in the body at the same rate and to the same extent. Generic products may be significantly less costly to bring to market than the reference listed drug and companies that produce generic products are generally able to offer them at lower prices. Thus, following the introduction of a generic drug, a significant percentage of the sales of any branded product or reference listed drug may be typically lost to the generic product.

The FDA may not approve an ANDA for a generic product until any applicable period of non-patent exclusivity for the reference listed drug has expired. Manufacturers may seek to launch these generic products following the expiration of the applicable marketing exclusivity period, even if we still have patent protection for our product.

Competition that our products may face from generic versions of our products could materially and adversely impact our future revenue, profitability and cash flows and substantially limit our ability to obtain a return on the investments we have made in those product candidates.

Product liability lawsuits against us could divert our resources, cause us to incur substantial liabilities and limit commercialization of any products that we may develop.

We face an inherent risk of product liability claims as a result of the clinical testing of our product candidates despite obtaining appropriate informed consents from our clinical trial participants. We will face an even greater risk if we commercially sell any product that we may develop. For example, we may be sued if any product we develop allegedly causes injury or is found to be otherwise unsuitable during clinical testing, manufacturing, marketing or sale. Any such product liability claims may include allegations of defects in manufacturing, defects in design, a failure to warn of dangers inherent in the

product, negligence, strict liability or a breach of warranties. For example, improper use or failure of INOpulse may result in rebound pulmonary hypertension, which can be fatal in some patients. Rebound pulmonary hypertension may also occur if both the primary and back-up devices fail before we can replace them, if the built-in back-up with a device does not work properly or if the patient does not carry or have access to his or her back-up device. Claims could also be asserted under state consumer protection acts. If we cannot successfully defend ourselves against product liability claims, we may incur substantial liabilities or be required to limit commercialization of our product candidates. Regardless of the merits or eventual outcome, liability claims may result in:

- decreased demand for products that we may develop;
- injury to our reputation and significant negative media attention;
- withdrawal of clinical trial participants;
- significant costs to defend resulting litigation;
- substantial monetary awards to trial participants or patients;
- loss of revenue;
- reduced resources of our management to pursue our business strategy; and
- the inability to commercialize any products that we may develop.

Although we maintain general liability insurance of $1.0 million in the aggregate, umbrella insurance in the amount of $10.0 million in the aggregate and clinical trial liability insurance of $20.0 million in the aggregate, this insurance may not fully cover potential liabilities that we may incur. The cost of any product liability litigation or other proceeding, even if resolved in our favor, could be substantial. We will need to increase our insurance coverage if and when we begin the commercial sale of any product candidate that receives marketing approval. In addition, insurance coverage is becoming increasingly expensive. If we are unable to obtain or maintain sufficient insurance coverage at an acceptable cost or to otherwise protect against potential product liability claims, it could prevent or inhibit the development and commercial production and sale of our product candidates, which could adversely affect our business, financial condition, results of operations and prospects.

Risks Related to Our Dependence on Third Parties

We rely, and expect to continue to rely, on third parties to conduct our clinical trials, and those third parties may not perform satisfactorily, including failing to meet deadlines for the completion of such trials.

We currently rely on third-party clinical research organizations, or CROs, to conduct our clinical trials. We expect to continue to rely on third parties, such as CROs, clinical data management organizations, medical institutions and clinical investigators, to conduct our clinical trials. Our agreements with these third parties generally allow the third party to terminate the agreement at any time. If we are required to enter into alternative arrangements because of any such termination, the introduction of our product candidates to market could be delayed.

Our reliance on these third parties for research and development activities will reduce our control over these activities but will not relieve us of our responsibilities. For example, we design our clinical trials and will remain responsible for ensuring that each of our clinical trials is conducted in accordance with the general investigational plan and protocols for the trial. Moreover, the FDA requires us to comply with GCPs for conducting, recording and reporting the results of clinical trials to assure that data and reported results are credible and accurate and that the rights, integrity and confidentiality of trial participants are protected. Our reliance on third parties that we do not control does not relieve us of these responsibilities and requirements. We also are required to register ongoing clinical trials and post the results of completed clinical trials on a government-sponsored database, ClinicalTrials.gov, within specified timeframes. Failure to do so can result in fines, adverse publicity and civil and criminal sanctions.

Furthermore, these third parties may also have relationships with other entities, some of which may be our competitors. If these third parties do not successfully carry out their contractual duties, meet expected deadlines or conduct our clinical trials in accordance with regulatory requirements or our stated protocols, we will not be able to obtain, or may be delayed in obtaining, marketing approvals for our product candidates and will not be able to, or may be delayed in our efforts to, successfully commercialize our product candidates.

We also expect to rely on other third parties to store and distribute drug and device supplies for our clinical trials. Any performance failure on the part of our distributors could delay clinical development or marketing approval of our product candidates or commercialization of our products, producing additional losses and depriving us of potential product revenue.

Our BCM product candidate currently in development is exclusively licensed from BioLineRx Ltd., and we may enter into additional agreements to in-license technology from third parties. If BioLineRx Ltd. or other future licensors terminate the applicable license, or fail to maintain or enforce the underlying patents, our competitive position and market share will be harmed.

We have an exclusive worldwide license for our BCM product candidate, subject to certain retained rights of the licensor, from BioLineRx Ltd. and its subsidiary, who we collectively refer to as BioLine. Under the terms of the license agreement, we are obligated to use commercially reasonable efforts to develop and commercialize at least one product containing BCM. BioLine has the right to terminate its license agreement with us for an uncured material breach by us, upon which our exclusive license for BCM will terminate. We have recently been engaged in discussions with BioLine relating to our performance under the license agreement. BioLine has indicated to us that it believes that we have breached the license agreement in several ways, including, but not limited to, failure to use commercially reasonable efforts to develop BCM, failure to provide BioLine with material information concerning the development and commercialization plans for BCM and failure to notify BioLine in advance of material public disclosures regarding BCM. We and BioLine also disagree about the timing of a $12.5 million milestone payment that we would owe BioLine based upon progress in our BCM clinical development program. If we are unable to reach agreement with BioLine on these issues, BioLine could bring a lawsuit against us or seek to terminate the license agreement. Although we believe that we would have strong defenses in any litigation that could be brought by BioLine, if BioLine were to prevail in any such litigation, one of the potential remedies would be the termination of the license agreement and our consequent loss of rights to BCM. In addition, if BioLine were to prevail in any such litigation, or if we were required to pay the milestone in dispute sooner than we had planned, or if we were required to return BCM to BioLine, these events could have a material adverse effect on our business, results of operations, financial condition and/or liquidity.

We have also exclusively licensed our INOpulse product candidates, in certain indications and settings, and subject to certain retained rights of the licensor, from Ikaria. See "Certain Relationships and Related Person Transactions—Relationship with Ikaria" for a summary of our exclusive cross-license, technology transfer and regulatory matters agreement with Ikaria.

We may enter into additional license agreements as part of the development of our business in the future. Such licensors, if any, may be responsible for prosecution of certain patent applications and maintenance of certain patents. Such licensors may not successfully prosecute such patent applications or maintain such patents, which we have licensed and on which our business depends. Our licensors may fail to pursue litigation against third-party infringers, may fail to prove infringement, or may fail to defend against counterclaims of patent invalidity or unenforceability. If these in-licenses are terminated, or if the underlying patents fail to provide the intended market exclusivity, competitors would have the freedom to seek regulatory approval of, and to market, products identical to ours. This could have a material adverse effect on our competitive business position and our business prospects.

We may have received better terms from unaffiliated third parties than the terms we received in our agreements with Ikaria.

The agreements related to the Spin-Out, including the separation and distribution agreement, TSA, license agreement, drug clinical supply agreement, device clinical supply agreement, agreements not to compete and the other agreements, were negotiated in the context of our separation from Ikaria while we were still part of Ikaria and, accordingly, may not reflect terms that would have resulted from arm's-length negotiations among unaffiliated third parties. The terms of the agreements we negotiated in the context of our separation related to, among other things, allocation of assets, liabilities, rights, indemnifications and other obligations among Ikaria and us. We may have received better terms from third parties because third parties may have competed with each other to win our business. Some of our board members are also members of the Ikaria board. See "Certain Relationships and Related Person Transactions—Relationship with Ikaria."

Third parties may seek to hold us responsible for liabilities of Ikaria that we did not assume in our agreements.

In connection with our separation from Ikaria, Ikaria has generally agreed to retain all liabilities that did not historically arise from our business. Third parties may seek to hold us responsible for Ikaria's retained liabilities. Under our agreements with Ikaria, Ikaria has agreed to indemnify us for claims and losses relating to these retained liabilities. However, if those liabilities are significant and we are ultimately liable for them, we cannot assure you that we will be able to recover the full amount of our losses from Ikaria.

Any disputes that arise between us and Ikaria with respect to our past and ongoing relationships could harm our business operations.

Disputes may arise between Ikaria and us in a number of areas relating to our past and ongoing relationships, including:

- intellectual property, technology and business matters, including failure to make required technology transfers and failure to comply with non-compete provisions applicable to Ikaria and us;
- labor, tax, employee benefit, indemnification and other matters arising from our separation from Ikaria;
- distribution and supply obligations;
- employee retention and recruiting;
- business combinations involving us;
- the nature, quality and pricing of transitional services Ikaria has agreed to provide us; and
- business opportunities that may be attractive to both Ikaria and us.

We may not be able to resolve any potential conflicts, and even if we do, the resolution may be less favorable than if we were dealing with an unaffiliated party.

We may seek to enter into collaborations with third parties for the development and commercialization of our product candidates. If we fail to enter into such collaborations, or such collaborations are not successful, we may not be able to capitalize on the market potential of our product candidates.

We may seek third-party collaborators for development and commercialization of our product candidates. Our likely collaborators for any marketing, distribution, development, licensing or broader collaboration arrangements include large and mid-size pharmaceutical and medical device companies, regional and national biotechnology companies and pharmaceutical companies. We are not currently party to any such arrangement. However, if we do enter into any such arrangements with any third

parties in the future, we will likely have limited control over the amount and timing of resources that our collaborators dedicate to the development or commercialization of our product candidates. Our ability to generate revenues from these arrangements will depend on our collaborators' abilities to successfully perform the functions assigned to them in these arrangements.

Collaborations involving our product candidates would pose the following risks to us:

- collaborators have significant discretion in determining the efforts and resources that they will apply to these collaborations;
- collaborators may not pursue development and commercialization of our product candidates or may elect not to continue or renew development or commercialization programs based on clinical trial results, changes in the collaborators' strategic focus or available funding, or external factors such as an acquisition that diverts resources or creates competing priorities;
- collaborators may delay clinical trials, provide insufficient funding for a clinical trial program, stop a clinical trial or abandon a product candidate, repeat or conduct new clinical trials or require a new formulation of a product candidate for clinical testing;
- collaborators could independently develop, or develop with third parties, products that compete directly or indirectly with our products or product candidates if the collaborators believe that competitive products are more likely to be successfully developed or can be commercialized under terms that are more economically attractive than ours;
- collaborators with marketing and distribution rights to one or more of our products may not commit sufficient resources to the marketing and distribution of such product or products;
- collaborators may not properly maintain or defend our intellectual property rights or may use our proprietary information in such a way as to invite litigation that could jeopardize or invalidate our intellectual property or proprietary information or expose us to potential litigation;
- collaborators may infringe the intellectual property rights of third parties, which may expose us to litigation and potential liability;
- disputes may arise between the collaborators and us that result in the delay or termination of the research, development or commercialization of our products or product candidates or that result in costly litigation or arbitration that diverts management attention and resources; and
- collaborations may be terminated and, if terminated, may result in a need for additional capital to pursue further development or commercialization of the applicable product candidates.

Collaboration agreements may not lead to development or commercialization of product candidates in the most efficient manner or at all. If a collaborator of ours were to be involved in a business combination, the continued pursuit and emphasis on our product development or commercialization program could be delayed, diminished or terminated.

If we are not able to establish collaborations, we may have to alter our development and commercialization plans.

Our drug and device development programs and the potential commercialization of our product candidates will require substantial additional cash to fund expenses. For some of our product candidates, we may decide to collaborate with biotechnology and pharmaceutical companies for the development and potential commercialization of those product candidates.

We face significant competition in seeking appropriate collaborators. Whether we reach a definitive agreement for a collaboration will depend, among other things, upon our assessment of the collaborator's resources and expertise, the terms and conditions of the proposed collaboration and the proposed collaborator's evaluation of a number of factors. Those factors may include the design or results of clinical trials, the likelihood of approval by the FDA or similar regulatory authorities outside

the United States, the potential market for the subject product candidate, the costs and complexities of manufacturing and delivering such product candidate to patients, the potential of competing products, the existence of uncertainty with respect to our ownership of technology, which can exist if there is a challenge to such ownership without regard to the merits of the challenge and industry and market conditions generally. The collaborator may also consider alternative product candidates or technologies for similar indications that may be available to collaborate on and whether such a collaboration could be more attractive than the one with us for our product candidate. The terms of our current or future license agreements may restrict our ability to enter into agreements on certain terms with future collaborators. For example, our license agreement with Ikaria prohibits us from granting a sublicense under any of the intellectual property licensed to us under such license agreement to any of our affiliates or any third party, in each case, that directly or indirectly competes with the Ikaria nitric oxide business, and any future license agreements may contain similar restrictions. Collaborations are complex and time-consuming to negotiate and document. In addition, there have been a significant number of recent business combinations among large pharmaceutical companies that have resulted in a reduced number of potential future collaborators.

We may not be able to negotiate collaborations on a timely basis, on acceptable terms, or at all. If we are unable to do so, we may have to curtail the development of a product candidate, reduce or delay its development program or one or more of our other development programs, delay its potential commercialization or reduce the scope of any sales or marketing activities, or increase our expenditures and undertake development or commercialization activities at our own expense. If we elect to increase our expenditures to fund development or commercialization activities on our own, we may need to obtain additional capital, which may not be available to us on acceptable terms or at all. If we do not have sufficient funds, we may not be able to further develop our product candidates or bring them to market and generate product revenue.

Risks Related to Our Intellectual Property

If we are unable to obtain and maintain patent protection for our technology and products or if the scope of the patent protection obtained is not sufficiently broad, our competitors could develop and commercialize technology and products similar or identical to ours, and our ability to successfully commercialize our technology and products may be impaired.

Our success depends in large part on our ability to obtain and maintain patent protection in the United States and other countries with respect to our proprietary technology and products. We seek to protect our proprietary position by filing patent applications in the United States and abroad related to our novel technologies and product candidates.

The patent prosecution process is expensive and time-consuming, and we may not be able to file and prosecute all necessary or desirable patent applications at a reasonable cost or in a timely manner. It is also possible that we will fail to identify patentable aspects of our research and development output before it is too late to obtain patent protection. Moreover, in some circumstances, we do not have the right to control the preparation, filing and prosecution of patent applications, or to maintain the patents, covering technology that we license from third parties. Therefore, these patents and applications may not be prosecuted and enforced in a manner consistent with the best interests of our business.

The patent position of biotechnology and pharmaceutical companies generally is highly uncertain, involves complex legal and factual questions and has in recent years been the subject of much litigation. In addition, the laws of non-U.S. countries may not protect our rights to the same extent as the laws of the United States. For example, European patent law restricts the patentability of methods of treatment of the human body more than U.S. law does. Publications of discoveries in the scientific literature often lag behind the actual discoveries, and patent applications in the United States and other jurisdictions are typically not published until 18 months after filing, and in some cases not at all. Therefore, we

cannot know with certainty whether we were the first to make the inventions claimed in our owned or licensed patents or pending patent applications, or that we or our licensors were the first to file for patent protection of such inventions. As a result, the issuance, scope, validity, enforceability and commercial value of our patent rights are highly uncertain. Our pending and future patent applications may not issue as patents that protect our technology or products, in whole or in part, or which effectively prevent others from commercializing competitive technologies and products. Changes in either the patent laws or interpretation of the patent laws in the United States and other countries may diminish the value of our patents or narrow the scope of our patent protection.

Recent patent reform legislation could increase the uncertainties and costs surrounding the prosecution of our patent applications and the enforcement or defense of our owned or licensed issued patents. On September 16, 2011, the Leahy-Smith America Invents Act, or the Leahy-Smith Act, was signed into law. The Leahy-Smith Act includes a number of significant changes to U.S. patent law. The Leahy-Smith Act includes provisions that affect the way patent applications are prosecuted and affect patent litigation. The U.S. Patent and Trademark Office, or USPTO, recently developed new regulations and procedures to govern administration of the Leahy-Smith Act. Many of the substantive changes to patent law associated with the Leahy-Smith Act, and in particular, the first to file provisions, became effective on March 16, 2013. Accordingly, it is not clear what, if any, impact the Leahy-Smith Act will have on the operation of our business. However, the Leahy-Smith Act and its implementation could increase the uncertainties and costs surrounding the prosecution of our owned or licensed patent applications and the enforcement or defense of our owned or licensed issued patents, all of which could have a material adverse effect on our business and financial condition.

Moreover, we may be subject to third-party preissuance submissions of prior art to the USPTO, or become involved in opposition, derivation, reexamination, *inter partes* review, post-grant review or interference proceedings challenging our owned or licensed patent rights or the patent rights of others. An adverse determination in any such submission, proceeding or litigation could reduce the scope of, or invalidate, our patent rights, allow third parties to commercialize our technology or products and compete directly with us, without payment to us, or result in our inability to manufacture or commercialize products without infringing third-party patent rights. In addition, if the breadth or strength of protection provided by our patents and patent applications is threatened, it could dissuade companies from collaborating with us to license, develop or commercialize current or future product candidates.

Even if our owned and licensed patent applications issue as patents, they may not issue in a form that will provide us with any meaningful protection, prevent competitors from competing with us, or otherwise provide us with any competitive advantage. Our competitors may be able to circumvent our owned or licensed patents by developing similar or alternative technologies or products in a non-infringing manner. We may not receive patent term extension under the Drug Price Competition and Patent Term Restoration Act of 1984, referred to as the Hatch-Waxman Act, that we expect or our rights during the extension period may be more limited than the full scope of the patent, making it easier for our competitors to develop and market non-infringing technologies or products.

The issuance of a patent is not conclusive as to its inventorship, scope, validity or enforceability, and our owned and licensed patents may be challenged in courts or patent offices in the United States and abroad. Such challenges may result in loss of exclusivity or freedom to operate, or in patent claims being narrowed, invalidated or held unenforceable, in whole or in part, which could limit our ability to stop others from using or commercializing similar or identical technology and products, or limit the duration of the patent protection of our technology and products. Given the amount of time required for the development, testing and regulatory review of new product candidates, patents protecting such candidates might expire before or shortly after such candidates are commercialized. As a result, our owned and licensed patent portfolio may not provide us with sufficient rights to exclude others from commercializing products similar or identical to ours.

We may become involved in lawsuits to protect or enforce our patents or other intellectual property, which could be expensive, time consuming and unsuccessful.

Competitors may infringe our owned or licensed patents or other intellectual property. To counter infringement or unauthorized use, we may be required to file or participate in infringement claims, which can be expensive and time consuming. Any claims we or our licensors assert against perceived infringers could provoke these parties to assert counterclaims against us alleging that we infringe their patents. In addition, in a patent infringement proceeding, a court may decide that a patent of ours or our licensor is invalid or unenforceable, in whole or in part, construe the patent's claims narrowly or refuse to stop the other party from using the technology at issue on the grounds that our patents do not cover the technology in question. An adverse result in any litigation proceeding could put one or more of our owned or licensed patents at risk of being invalidated or interpreted narrowly.

If we fail to comply with our obligations under license agreements, we could lose rights that are important to our business. BioLine has raised concerns with respect to our performance under the terms of our license agreement and may bring suit against us.

We are party to a license agreement with BioLine relating to our BCM product candidate that imposes, and we may enter into additional license agreements that may impose, various diligence, milestone payment, royalty and other obligations on us. Under our existing license agreement with BioLine, we are obligated to pay royalties on the net sales of product candidates or related technologies to the extent they are covered by the agreement. We also have diligence and development obligations under this agreement. If we fail to comply with our obligations under current or future license agreements, our counterparties may have the right to terminate these agreements, in which event we might not be able to develop, manufacture or market any product that is covered by the agreement or face other penalties under the agreement. Such an occurrence could materially adversely affect the value of the product candidate being developed under any such agreement.

We have recently been engaged in discussions with BioLine relating to our performance under the license agreement. BioLine has indicated to us that it believes that we have breached the license agreement in several ways, including, but not limited to, failure to use commercially reasonable efforts to develop BCM, failure to provide BioLine with material information concerning the development and commercialization plans for BCM and failure to notify BioLine in advance of material public disclosures regarding BCM. We and BioLine also disagree about the timing of a $12.5 million milestone payment that we would owe BioLine based upon progress in our BCM clinical development program. If we are unable to reach agreement with BioLine on these issues, BioLine could bring a lawsuit against us or seek to terminate the license agreement. Although we believe that we would have strong defenses in any litigation that could be brought by BioLine, if BioLine were to prevail in any such litigation, one of the potential remedies would be the termination of the license agreement and our consequent loss of rights to BCM. In addition, if BioLine were to prevail in any such litigation, or if we were required to pay the milestone in dispute sooner than we had planned, or if we were required to return BCM to BioLine, these events could have a material adverse effect on our business, results of operations, financial condition and/or liquidity. Termination of our license agreement with BioLine, or any future license agreements we may enter into, or reduction or elimination of our rights under such agreements may result in our having to negotiate new or reinstated agreements with less favorable terms, or cause us to lose our rights under these agreements, including our rights to important intellectual property or technology.

Third parties may initiate legal proceedings alleging that we are infringing their intellectual property rights, the outcome of which would be uncertain and could have a material adverse effect on the success of our business.

Our commercial success depends upon our ability to develop, manufacture, market and sell our product candidates and use our proprietary technologies without infringing the proprietary rights of third parties. There is considerable intellectual property litigation in the pharmaceutical, biotechnology and medical device industries. We may become party to, or be threatened with, future adversarial proceedings or litigation regarding intellectual property rights with respect to our products and technology, including interference or derivation proceedings before the USPTO. Third parties may assert infringement claims against us based on existing patents or patents that may be granted in the future.

If we are found to infringe a third party's intellectual property rights, we could be required to obtain a license from such third party to continue developing and marketing our products and technology. However, we may not be able to obtain any required license on commercially reasonable terms or at all. Even if we were able to obtain a license, it could be non-exclusive, thereby giving our competitors access to the same technologies licensed to us. We could be forced, including by court order, to cease commercializing the infringing technology or product. In addition, we could be found liable for monetary damages, including treble damages and attorneys' fees if we are found to have willfully infringed a patent. A finding of infringement could prevent us from commercializing our product candidates or force us to cease some of our business operations, which could materially harm our business. Claims that we have misappropriated the confidential information or trade secrets of third parties could have a similar negative impact on our business.

We may be subject to claims by third parties asserting that we or our employees have misappropriated their intellectual property, or claiming ownership of what we regard as our own intellectual property.

Many of our employees were previously employed at other pharmaceutical, biotechnology or medical device companies, including our competitors or potential competitors. Although we try to ensure that our employees do not use the proprietary information or know-how of others in their work for us, we may be subject to claims that we or these employees have used or disclosed intellectual property, including trade secrets or other proprietary information, of any such employee's former employer. Litigation may be necessary to defend against these claims.

In addition, while it is our policy to require our employees and contractors who may be involved in the development of intellectual property to execute agreements assigning such intellectual property to us, we may be unsuccessful in timely obtaining such an agreement with each party who in fact develops intellectual property that we regard as our own. Even if timely obtained, such agreements may be breached, and we may be forced to bring claims against third parties, or defend claims they may bring against us, to determine the ownership of what we regard as our intellectual property.

If we fail in prosecuting or defending any such claims, we may lose valuable intellectual property rights or personnel, in addition to paying monetary damages. Even if we are successful in prosecuting or defending against such claims, litigation could result in substantial costs and be a distraction to management.

Intellectual property litigation could cause us to spend substantial resources and distract our personnel from their normal responsibilities.

Even if resolved in our favor, litigation or other legal proceedings relating to intellectual property claims may cause us to incur significant expenses, and could distract our technical and management personnel from their normal responsibilities. In addition, there could be public announcements of the results of hearings, motions or other interim proceedings or developments, and if securities analysts or

investors perceive these results to be negative, it could have a substantial adverse effect on the price of our common stock. Such litigation or proceedings could substantially increase our operating losses and reduce the resources available for development activities or any future sales, marketing or distribution activities. We may not have sufficient financial or other resources to conduct such litigation or proceedings adequately. Some of our competitors may be able to sustain the costs of such litigation or proceedings more effectively than we can because of their greater financial resources. Uncertainties resulting from the initiation and continuation of patent litigation or other proceedings could compromise our ability to compete in the marketplace.

If we are unable to protect the confidentiality of our trade secrets, our business and competitive position would be harmed.

In addition to seeking patents for some of our technology and product candidates, we also rely on trade secrets, including unpatented know-how, technology and other proprietary information, to maintain our competitive position. We seek to protect these trade secrets, in part, by entering into non-disclosure and confidentiality agreements with parties who have access to them, such as our employees, outside scientific collaborators, contract manufacturers, consultants, advisors and other third parties. We also enter into confidentiality and invention or patent assignment agreements with our employees and consultants. Despite these efforts, any of these parties may breach the agreements and disclose our proprietary information, including our trade secrets, and we may not be able to obtain adequate remedies for such breaches. Enforcing a claim that a party illegally disclosed or misappropriated a trade secret is difficult, expensive and time-consuming, and the outcome is unpredictable. Even if we are successful in prosecuting such claims, any remedy awarded may be insufficient to fully compensate us for the improper disclosure or misappropriation. In addition, some courts inside and outside the United States are less willing or unwilling to protect trade secrets. If any of our trade secrets were to be lawfully obtained or independently developed by a competitor, we would have no right to prevent them, or those to whom they communicate it, from using that technology or information to compete with us. If any of our trade secrets were to be disclosed to or independently developed by a competitor, our competitive position would be harmed.

Intellectual property rights do not necessarily address all potential threats to our competitive advantage.

The degree of future protection afforded by our intellectual property rights is uncertain because intellectual property rights have limitations and may not adequately protect our business, or permit us to maintain our competitive advantage. The following examples are illustrative:

- Others may be able to develop and commercialize treatments that are similar to our product candidates but that are not covered by the claims of the patents that we own or have exclusively licensed.
- We or our licensors might not have been the first to make the inventions covered by the issued patent or pending patent application that we own or have exclusively licensed.
- We or our licensors might not have been the first to file patent applications covering certain of our inventions.
- Others may independently develop similar or alternative technologies or duplicate any of our technologies without infringing our intellectual property rights.
- It is possible that our pending patent applications will not lead to issued patents.
- Issued patents that we own or have exclusively licensed may not provide us with any competitive advantages, or may be held invalid or unenforceable, as a result of legal challenges by our competitors.

- Our competitors might conduct research and development activities in countries where we do not have patent rights and then use the information learned from such activities to develop competitive products for sale in our major commercial markets.

- We may not develop additional proprietary technologies that are patentable.

- The patents of others may have an adverse effect on our business.

- Another party may be granted orphan exclusivity for an indication that we are seeking before us or may be granted orphan exclusivity for one of our products for another indication.

Risks Related to Regulatory Approval of Our Product Candidates and Other Legal Compliance Matters

Even if we complete the necessary clinical trials, the marketing approval process is expensive, time consuming and uncertain and may prevent us from obtaining approvals for the commercialization of some or all of our product candidates. If we are not able to obtain, or if there are delays in obtaining, required regulatory approvals, we will not be able to commercialize our product candidates, and our ability to generate revenue will be materially impaired.

Our product candidates and the activities associated with their development and commercialization, including their design, testing, manufacture, safety, efficacy, recordkeeping, labeling, storage, approval, advertising, promotion, sale and distribution, are subject to comprehensive regulation by the FDA and other regulatory agencies in the United States and by the EMA and similar regulatory authorities outside the United States. Failure to obtain marketing approval for a product candidate will prevent us from commercializing the product candidate. Our product candidates are in the early stages of development and are subject to the risks of failure inherent in drug and device development. We have not received approval to market any of our product candidates from regulatory authorities in any jurisdiction. We have only limited experience in conducting and managing the clinical trials, and in filing and supporting the applications necessary to gain marketing approvals and expect to rely on third-party CROs to assist us in this process. Securing marketing approval requires the submission of extensive pre-clinical and clinical data and supporting information to regulatory authorities for each therapeutic indication to establish the product candidate's safety and efficacy. Securing marketing approval also requires the submission of information about the product manufacturing process to, and inspection of manufacturing facilities by, the regulatory authorities. Our product candidates may not be effective, may be only moderately effective or may prove to have undesirable or unintended side effects, toxicities or other characteristics that may preclude our obtaining marketing approval or prevent or limit commercial use.

The process of obtaining marketing approvals, both in the United States and abroad, is expensive, may take many years if additional clinical trials are required, if approval is obtained at all, and can vary substantially based upon a variety of factors, including the type, complexity and novelty of the product candidates involved. Changes in marketing approval policies during the development period, changes in or the enactment of additional statutes or regulations, or changes in regulatory review for each submitted product application, may cause delays in the approval or rejection of an application. Regulatory authorities have substantial discretion in the approval process and may refuse to accept any application or may decide that our data are insufficient for approval and require additional pre-clinical, clinical or other studies. In addition, varying interpretations of the data obtained from pre-clinical and clinical testing could delay, limit or prevent marketing approval of a product candidate. Any marketing approval we ultimately obtain may be limited or subject to restrictions or post-approval commitments that render the approved product not commercially viable. If we experience delays in obtaining approval or if we fail to obtain approval of our product candidates, the commercial prospects for our product candidates may be harmed and our ability to generate revenues will be materially impaired.

Our failure to obtain marketing approval in foreign jurisdictions would prevent our product candidates from being marketed abroad, and any approval we are granted for our product candidates in the United States would not assure approval of product candidates in foreign jurisdictions.

In order to market and sell our products in the European Union and many other jurisdictions, we must obtain separate marketing approvals and comply with numerous and varying regulatory requirements. The approval procedure varies among countries and can involve additional testing. The time required to obtain approval may differ substantially from that required to obtain FDA approval. The regulatory approval process outside the United States generally includes all of the risks associated with obtaining FDA approval. In addition, in many countries outside the United States, it is required that the product be approved for reimbursement before the product can be approved for sale in that country. We may not obtain approvals from regulatory authorities outside the United States on a timely basis, if at all. Approval by the FDA does not ensure approval by regulatory authorities in other countries or jurisdictions, and approval by one regulatory authority outside the United States does not ensure approval by regulatory authorities in other countries or jurisdictions or by the FDA. We may not be able to file for marketing approvals and may not receive necessary approvals to commercialize our products in any market.

Even if we obtain marketing approval for our product candidates, the terms of approvals and ongoing regulation of our products may limit how we manufacture and market our products and compliance with such requirements may involve substantial resources, which could materially impair our ability to generate revenue.

Even if marketing approval of a product candidate is granted, an approved product and its manufacturer and marketer are subject to ongoing review and extensive regulation, including the requirement to implement a risk evaluation and mitigation strategy or to conduct costly post-marketing studies or clinical trials and surveillance to monitor the safety or efficacy of the product. We must also comply with requirements concerning advertising and promotion for any of our product candidates for which we obtain marketing approval. Promotional communications with respect to prescription drugs are subject to a variety of legal and regulatory restrictions and must be consistent with the information in the product's approved labeling. Thus, we will not be able to promote any products we develop for indications or uses for which they are not approved. In addition, manufacturers of approved products and those manufacturers' facilities are required to ensure that quality control and manufacturing procedures conform to cGMP, which include requirements relating to quality control and quality assurance as well as the corresponding maintenance of records and documentation and reporting requirements. We and our contract manufacturers could be subject to periodic unannounced inspections by the FDA and other regulatory authorities to monitor and ensure compliance with cGMP.

Accordingly, assuming we receive marketing approval for one or more of our product candidates, we and our contract manufacturers will continue to expend time, money and effort in all areas of regulatory compliance, including manufacturing, production, product surveillance and quality control. If we are not able to comply with post-approval regulatory requirements, we could have the marketing approvals for our products withdrawn by regulatory authorities and our ability to market any future products could be limited, which could adversely affect our ability to achieve or sustain profitability. Thus, the cost of compliance with post-approval regulations may have a negative effect on our operating results and financial condition.

Any product candidate for which we obtain marketing approval will be subject to strict enforcement of post-marketing requirements and we could be subject to substantial penalties, including withdrawal of our product from the market, if we fail to comply with all regulatory requirements or if we experience unanticipated problems with our products, when and if any of them are approved.

Any product candidate for which we obtain marketing approval, along with the manufacturing processes, post-approval clinical data, labeling, advertising and promotional activities for such product,

will be subject to continual requirements of and review by the FDA and other regulatory authorities. These requirements include, but are not limited to, restrictions governing promotion of an approved product, submissions of safety and other post-marketing information and reports, registration and listing requirements, cGMP requirements relating to manufacturing, quality control, quality assurance and corresponding maintenance of records and documents, and requirements regarding the distribution of samples to physicians and recordkeeping.

The FDA and other federal and state agencies, including the Department of Justice, closely regulate compliance with all requirements governing prescription drug and device products, including requirements pertaining to marketing and promotion of drugs and devices in accordance with the provisions of the approved labeling and manufacturing of products in accordance with cGMP requirements. Violations of such requirements may lead to investigations alleging violations of the Food, Drug, and Cosmetic Act and other statutes, including the False Claims Act and other federal and state health care fraud and abuse laws as well as state consumer protection laws. Our failure to comply with all regulatory requirements, and later discovery of previously unknown adverse events or other problems with our products, manufacturers or manufacturing processes, may yield various results, including:

- litigation involving patients taking our products;
- restrictions on such products, manufacturers or manufacturing processes;
- restrictions on the labeling or marketing of a product;
- restrictions on product distribution or use;
- requirements to conduct post-marketing studies or clinical trials;
- untitled or warning letters;
- withdrawal of the products from the market;
- refusal to approve pending applications or supplements to approved applications that we submit;
- recall of products;
- fines, restitution or disgorgement of profits or revenues;
- suspension or withdrawal of marketing approvals;
- damage to relationships with any potential collaborators;
- unfavorable press coverage and damage to our reputation;
- refusal to permit the import or export of our products;
- product seizure; or
- injunctions or the imposition of civil or criminal penalties.

Non-compliance by us or any future collaborator with regulatory requirements regarding safety monitoring or pharmacovigilance, and with requirements related to the development of products for the pediatric population, can also result in significant financial penalties. Similarly, failure to comply with regulatory requirements regarding the protection of personal information can also lead to significant penalties and sanctions.

Our relationships with customers and third-party payors will be subject to applicable anti-kickback, fraud and abuse and other healthcare laws and regulations, which could expose us to criminal sanctions, civil penalties, contractual damages, reputational harm and diminished profits and future earnings.

Healthcare providers, physicians and third-party payors will play a primary role in the recommendation and prescription of any product candidates for which we obtain marketing approval. Our future arrangements with third-party payors and customers may expose us to broadly applicable fraud and abuse and other healthcare laws and regulations that may constrain the business or financial arrangements and relationships through which we market, sell and distribute any products for which we obtain marketing approval. Restrictions under applicable federal and state healthcare laws and regulations, include the following:

- the federal Anti-Kickback Statute prohibits, among other things, persons from knowingly and willfully soliciting, offering, receiving or providing remuneration, directly or indirectly, in cash or in kind, to induce or reward, or in return for, either the referral of an individual for, or the purchase, order or recommendation of, any good or service, for which payment may be made under a federal healthcare program such as Medicare and Medicaid;

- the federal False Claims Act imposes criminal and civil penalties, including civil whistleblower or qui tam actions, against individuals or entities for, among other things, knowingly presenting, or causing to be presented false or fraudulent claims for payment by a federal government program, or making a false statement or record material to payment of a false claim or avoiding, decreasing or concealing an obligation to pay money to the federal government;

- the federal Health Insurance Portability and Accountability Act of 1996, or HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act, imposes criminal and civil liability for executing a scheme to defraud any healthcare benefit program and also imposes obligations, including mandatory contractual terms, with respect to safeguarding the privacy, security and transmission of individually identifiable health information;

- HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act and its implementing regulations, also imposes obligations, including mandatory contractual terms, with respect to safeguarding the privacy, security and transmission of individually identifiable health information;

- the federal false statements statute prohibits knowingly and willfully falsifying, concealing or covering up a material fact or making any materially false statement in connection with the delivery of or payment for healthcare benefits, items or services;

- the federal transparency requirements under the Patient Protection and Affordable Care Act, as amended by the Health Care and Education Reconciliation Act of 2010, or collectively the PPACA, requires applicable manufacturers of covered drugs, devices, biologics and medical supplies to report to the Department of Health and Human Services information related to payments and other transfers of value to physicians and teaching hospitals and physician ownership and investment interests; and

- analogous state laws and regulations such as state anti-kickback and false claims laws and analogous non-U.S. fraud and abuse laws and regulations, may apply to sales or marketing arrangements and claims involving healthcare items or services reimbursed by non-governmental third-party payors, including private insurers, and some state laws require pharmaceutical companies to comply with the pharmaceutical industry's voluntary compliance guidelines and the relevant compliance guidance promulgated by the federal government in addition to requiring drug manufacturers to report information related to payments to physicians and other health care providers or marketing expenditures. Some state laws require pharmaceutical companies to comply with the pharmaceutical industry's voluntary compliance guidelines and the relevant

compliance guidance promulgated by the federal government and may require drug manufacturers to report information related to payments and other transfers of value to physicians and other healthcare providers or marketing expenditures. State and non-U.S. laws also govern the privacy and security of health information in some circumstances, many of which differ from each other in significant ways and often are not preempted by HIPAA, thus complicating compliance efforts.

Efforts to ensure that our business arrangements with third parties will comply with applicable healthcare laws and regulations will involve substantial costs. It is possible that governmental authorities will conclude that our business practices may not comply with current or future statutes, regulations or case law involving applicable fraud and abuse or other healthcare laws and regulations. If our operations are found to be in violation of any of these laws or any other governmental regulations that may apply to us, we may be subject to significant civil, criminal and administrative penalties, damages, fines, imprisonment, exclusion of products from government funded healthcare programs, such as Medicare and Medicaid, and the curtailment or restructuring of our operations. If any of the physicians or other healthcare providers or entities with whom we expect to do business is found to be not in compliance with applicable laws, they may be subject to criminal, civil or administrative sanctions, including exclusions from government funded healthcare programs.

Laws and regulations governing any international operations we may have in the future may preclude us from developing, manufacturing and selling certain product candidates and products outside of the United States and require us to develop and implement costly compliance programs.

If we expand our operations outside of the United States, we must dedicate additional resources to comply with numerous laws and regulations in each jurisdiction in which we plan to operate. The Foreign Corrupt Practices Act, or the FCPA, prohibits any U.S. individual or business from paying, offering, authorizing payment or offering anything of value, directly or indirectly, to any foreign official, political party or candidate for the purpose of influencing any act or decision of such third party in order to assist the individual or business in obtaining or retaining business. The FCPA also obligates companies whose securities are listed in the United States to comply with certain accounting provisions requiring the company to maintain books and records that accurately and fairly reflect all transactions of the company, including international subsidiaries, and to devise and maintain an adequate system of internal accounting controls for international operations.

Compliance with the FCPA is expensive and difficult, particularly in countries in which corruption is a recognized problem. In addition, the FCPA presents particular challenges in the medical device industry, because, in many countries, hospitals are operated by the government, and doctors and other hospital employees are considered foreign officials. Certain payments to hospitals in connection with clinical trials and other work have been deemed to be improper payments to government officials and have led to FCPA enforcement actions.

Various laws, regulations and executive orders also restrict the use and dissemination outside of the United States, or the sharing with certain non-U.S. nationals, of information classified for national security purposes, as well as certain products and technical data relating to those products. If we expand our presence outside of the United States, it will require us to dedicate additional resources to comply with these laws, and these laws may preclude us from developing, manufacturing or selling certain product candidates and products outside of the United States, which could limit our growth potential and increase our development costs.

The failure to comply with laws governing international business practices may result in substantial civil and criminal penalties and suspension or debarment from government contracting. The Securities and Exchange Commission also may suspend or bar issuers from trading securities on U.S. exchanges for violations of the FCPA's accounting provisions.

If we fail to comply with environmental, health and safety laws and regulations, we could become subject to fines or penalties or incur costs that could harm our business.

Currently, we do not operate any research and development or production facilities, including laboratory, development or manufacturing facilities. However, if we decided to operate our own research and development and production facilities, we would be subject to numerous environmental, health and safety laws and regulations, including those governing laboratory procedures and the handling, use, storage, treatment and disposal of hazardous materials and wastes. Such operations may involve the use of hazardous and flammable materials, including chemicals and biological materials. Our operations may also produce hazardous waste products. Even if we contract with third parties for the disposal of these materials and wastes, we would not be able to eliminate the risk of contamination or injury from these materials. In the event of contamination or injury resulting from our use or disposal of hazardous materials, we could be held liable for any resulting damages, and any liability could exceed our resources. We also could incur significant costs associated with civil or criminal fines and penalties for failure to comply with such laws and regulations.

Although we would increase our level of workers' compensation insurance to cover us for costs and expenses we may incur due to injuries to our employees resulting from the use of hazardous materials, this insurance may not provide adequate coverage against potential liabilities. We do not expect to maintain insurance for environmental liability or toxic tort claims that may be asserted against us in connection with our possible future storage or disposal of biological, hazardous or radioactive materials.

In addition, we may incur substantial costs in order to comply with current or future environmental, health and safety laws and regulations. These current or future laws and regulations may impair our research, development or production efforts. Our failure to comply with these laws and regulations also may result in substantial fines, penalties or other sanctions.

Our INOpulse product candidates use lithium-ion battery cells, which have been observed to catch fire or vent smoke and flame, and these events may raise concerns about the batteries we use.

The battery pack used in our INOpulse product candidates makes use of lithium-ion cells. On rare occasions, lithium-ion cells can rapidly release the energy they contain by venting smoke and flames in a manner that can ignite nearby materials. Highly publicized incidents of laptop computers and cell phones bursting into flames have focused consumer attention on the safety of these cells. There can be no assurance that the battery packs we use would not fail, which could lead to property damage, personal injury or death, and may subject us to lawsuits. We may also have to recall our products, if any, which would be time consuming and expensive. Also, negative perceptions in the healthcare and patient communities regarding the suitability of lithium-ion cells for medical applications or any future incident involving lithium-ion cells could seriously harm our business, even in the absence of an incident involving us.

Risks Related to Employee Matters and Managing Growth

Our future success depends on our ability to retain key executives and to attract, retain and motivate qualified personnel.

Recruiting and retaining qualified scientific, clinical, manufacturing and sales and marketing personnel will also be critical to our success. Any of our employees may terminate their employment with us at any time. The loss of the services of our executive officers or other key employees could impede the achievement of our research, development and commercialization objectives and seriously harm our ability to successfully implement our business strategy. We do not maintain "key person" insurance for any of our executives or other employees. Furthermore, replacing executive officers and key employees may be difficult and may take an extended period of time because of the limited

number of individuals in our industry with the breadth of skills and experience required to successfully develop, gain regulatory approval of and commercialize products. Competition to hire from this limited pool is intense, and we may be unable to hire, train, retain or motivate these key personnel on acceptable terms given the competition among numerous pharmaceutical, biotechnology and medical device companies for similar personnel. We also experience competition for the hiring of scientific and clinical personnel from universities and research institutions. In addition, we rely on consultants and advisors, including scientific and clinical advisors, to assist us in formulating our research and development and commercialization strategy. Our consultants and advisors may be employed by employers other than us and may have commitments under consulting or advisory contracts with other entities that may limit their availability to us.

In addition, we currently have an interim chief executive officer. If we are successful in identifying and hiring a suitable chief executive officer, leadership transitions can be inherently difficult to manage and may cause some disruptions in our business.

Our employees may engage in misconduct or other improper activities, including noncompliance with regulatory standards and requirements and insider trading.

We are exposed to the risk of employee fraud or other misconduct. Misconduct by employees could include intentional failures to comply with FDA regulations, to provide accurate information to the FDA, to comply with federal and state healthcare fraud and abuse laws and regulations, to report financial information or data accurately, to disclose unauthorized activities to us or to comply with our Code of Business Conduct and Ethics. In particular, sales, marketing and business arrangements in the healthcare industry are subject to extensive laws and regulations intended to prevent fraud, kickbacks, false claims, inappropriate promotion, self-dealing and other abusive practices. These laws and regulations may restrict or prohibit a wide range of pricing, discounting, marketing and promotion, sales commission, customer incentive programs and other business arrangements. Employee misconduct could also involve the improper use of information obtained in the course of clinical trials, which could result in regulatory sanctions and serious harm to our reputation. The precautions we take to detect and prevent this activity may not be effective in controlling unknown or unmanaged risks or losses or in protecting us from governmental investigations or other actions or lawsuits stemming from a failure to be in compliance with such laws or regulations. If any such actions are instituted against us, and we are not successful in defending ourselves or asserting our rights, those actions could have a significant impact on our business, including the imposition of significant fines or other sanctions.

In addition, during the course of our operations, our directors, executives and employees may have access to material, non-public information regarding our business, our results of operations or potential transactions we are considering. We may not be able to prevent a director, executive or employee from violating our insider trading policies and trading in our common stock on the basis of, or while having access to, material, non-public information. If a director, executive or employee was to be investigated, or an action was to be brought against a director, executive or employee for insider trading, it could have a negative impact on our reputation and our stock price. Such a claim, with or without merit, could also result in substantial expenditures of time and money, and divert attention of our management team from other tasks important to the success of our business.

We expect to expand our development and regulatory capabilities and potentially implement sales, marketing and distribution capabilities, and as a result, we may encounter difficulties in managing our growth, which could disrupt our operations.

As of March 31, 2014, we had 55 full-time employees, of which 51 employees were engaged in research and development. We expect to experience significant growth in the number of our employees and the scope of our operations, particularly in the areas of development, regulatory affairs and, if any of our product candidates receives marketing approval, sales, marketing and distribution. To manage

our anticipated future growth, we must continue to implement and improve our managerial, operational and financial systems, expand our facilities and continue to recruit and train additional qualified personnel. Due to our limited financial resources and the limited experience of our management team in managing a company with such anticipated growth, we may not be able to effectively manage the expansion of our operations or recruit and train additional qualified personnel. The expansion of our operations may lead to significant costs and may divert our management and business development resources. Any inability to manage growth could delay the execution of our business plans or disrupt our operations.

Risks Related to This Offering and Ownership of Our Common Stock

You will not have the same protections available to other stockholders of NASDAQ-listed companies because we are a "controlled company" within the meaning of The NASDAQ Global Market's standards and, as a result, will qualify for, and will rely on, exemptions from several corporate governance requirements.

Certain of our stockholders, New Mountain Partners II (AIV-A), L.P., Allegheny New Mountain Partners, L.P. and New Mountain Affiliated Investors II, L.P., which we refer to collectively as the New Mountain Entities; IRDO Holding Corp., or ARCH; Venrock IK Holdings BT, Inc., or Venrock; Linde North America, Inc., an indirect wholly-owned subsidiary of Linde AG, or Linde; and 5AM-BT, Inc. and Aravis Venture I L.P., which we refer to collectively as the 5AM Entities, are parties to a voting agreement that will remain in effect following this offering and, as a result, will be deemed to hold their shares of our stock as part of a group. We refer to the New Mountain Entities, ARCH, Venrock, Linde and the 5AM Entities collectively as the Controlling Entities. Upon completion of this offering, the Controlling Entities are expected to control a majority of our outstanding capital stock and will be able to elect a majority of our directors. As a result, we will be a "controlled company" within the meaning of the rules governing companies with stock quoted on The NASDAQ Global Market. Under these rules, a company as to which an individual, a group or another company holds more than 50% of the voting power is considered a "controlled company" and can choose to be exempt from the following corporate governance requirements:

- a majority of the board of directors consist of independent directors;
- compensation of officers be determined or recommended to the board of directors by a majority of its independent directors or by a compensation committee that is composed entirely of independent directors; and
- director nominees be selected or recommended for election by a majority of the independent directors or by a nominating committee that is composed entirely of independent directors.

Following this offering, we intend to avail ourselves of these exemptions. Accordingly, you will not have the same protections afforded to stockholders of other companies that are subject to all of The NASDAQ Global Market corporate governance requirements as long as the Controlling Entities own a majority of our outstanding capital stock.

Our largest stockholders will continue to have substantial control over us after this offering and could limit your ability to influence the outcome of key transactions, including any change of control.

Upon the completion of this offering, we anticipate that our largest stockholders, the New Mountain Entities, will own, in the aggregate, approximately % of our outstanding common stock (% if the underwriters exercise their option to purchase additional shares in full). Following the completion of this offering, (i) the New Mountain Entities are entitled to elect (a) three directors, for so long as they beneficially own 15% or more of our outstanding common stock, (b) two directors, for so long as they beneficially own less than 15% but more than 5% of our outstanding common stock and (c) one director, for so long as they beneficially own less than 5% of our outstanding common stock but more than one share of our common stock and (ii) each of ARCH, Venrock and Linde is entitled to elect one director for so long as such holder owns 5% or more of our outstanding common stock. See "Certain Relationships and Related Person Transactions—Agreements with the Controlling Entities—Voting Agreement."

The New Mountain Entities will also retain the benefit of the rights conferred by the stockholders agreement. See "Certain Relationships and Related Person Transactions—Agreements with the Controlling Entities—Stockholders Agreement." As a result, the New Mountain Entities would be able to exert significant influence over matters requiring board approval, including the compensation and hiring and firing of our senior management, business combinations, issuance of shares of our capital stock, incurrence of debt, and payment of dividends, and their consent would be required for many matters requiring approval by our stockholders. These rights will terminate when the New Mountain Entities and their assignees beneficially own less than 15% of our outstanding capital stock. Upon completion of this offering, we anticipate that the New Mountain Entities together with our current stockholders will own, in the aggregate, approximately % of our outstanding common stock (% if the underwriters exercise their option to purchase additional shares in full).

The New Mountain Entities may have interests that differ from your interests, and they may vote in a way with which you disagree and that may be adverse to your interests. The concentration of ownership of our capital stock may have the effect of delaying, preventing or deterring a change of control of our company, could deprive our stockholders of an opportunity to receive a premium for their common stock as part of a sale of our company and may adversely affect the market price of our common stock.

We do not know whether a market will develop for our common stock or what the market price of our common stock will be and as a result it may be difficult for you to sell your shares of our common stock.

Before this offering, there was no public trading market for our common stock. If a market for our common stock does not develop or is not sustained, it may be difficult for you to sell your shares of common stock at an attractive price or at all. We cannot predict the prices at which our common stock will trade. It is possible that in one or more future periods our results of operations may be below the expectations of public market analysts and investors and, as a result of these and other factors, the price of our common stock may fall.

The market price of our common stock may be volatile, which could result in substantial losses for investors purchasing shares in this offering.

The initial public offering price for our common stock will be determined through negotiations with the underwriters. This initial public offering price may vary from the market price of our common stock after the offering. Some of the factors that may cause the market price of our common stock to fluctuate include:

- actual or anticipated results from and any delays in our clinical trials, including our ongoing clinical trials of our INOpulse and BCM product candidates, as well as results of regulatory input on our clinical trial programs and regulatory reviews relating to the approval of our product candidates;
- the results of our efforts to discover, develop, acquire or in-license additional product candidates or products;
- failure or discontinuation of any of our clinical development programs;
- the level of expenses related to any of our product candidates or clinical development programs;
- commencement or termination of any collaboration or licensing arrangement;
- disputes or other developments relating to proprietary rights, including patents, litigation matters and our ability to obtain patent protection for our technologies;
- announcements by us or our competitors of significant acquisitions, strategic partnerships, joint ventures and capital commitments;

- additions or departures of key scientific or management personnel;
- variations in our financial results or those of companies that are perceived to be similar to us;
- new products, product candidates or new uses for existing products introduced or announced by our competitors, and the timing of these introductions or announcements;
- results of clinical trials of product candidates of our competitors;
- general economic and market conditions and other factors that may be unrelated to our operating performance or the operating performance of our competitors, including changes in market valuations of similar companies;
- regulatory or legal developments in the United States and other countries;
- changes in the structure of healthcare payment systems;
- conditions or trends in the pharmaceutical, biotechnology and medical device industries;
- actual or anticipated changes in earnings estimates, development timelines or recommendations by securities analysts;
- announcement or expectation of additional financing efforts;
- sales of common stock by us or our stockholders in the future, as well as the overall trading volume of our common stock; and
- the other factors described in this "Risk Factors" section.

In addition, the stock market in general and the market for pharmaceutical, biotechnology and medical device companies in particular have experienced extreme price and volume fluctuations that have often been unrelated or disproportionate to the operating performance of those companies. These broad market and industry factors may seriously harm the market price of our common stock, regardless of our operating performance. In the past, following periods of volatility in companies' stock prices, securities class-action litigation has often been instituted against such companies. Such litigation, if instituted against us, could result in substantial costs and diversion of management's attention and resources, which could materially and adversely affect our business and financial condition.

A significant portion of our total outstanding shares may be sold into the public market in the near future, which could cause the market price of our common stock to drop significantly, even if our business is doing well.

Sales of a substantial number of shares of our common stock in the public market, or the perception in the market that the holders of a large number of shares intend to sell shares, could reduce the market price of our common stock. After this offering, we will have outstanding shares of common stock based on the number of shares outstanding as of April 30, 2014, assuming no exercise by the underwriters of their option to purchase additional shares. This includes the shares that we are selling in this offering, which may be resold in the public market immediately without restriction, unless purchased by our affiliates or existing stockholders. The remaining shares are currently restricted as a result of securities laws or lock-up agreements but will become eligible to be sold at various times after the offering. Moreover, after this offering, holders of an aggregate of 47,468,903 shares of our common stock will have rights, subject to specified conditions, to require us to file registration statements covering their shares or, along with holders of an additional 39,707,512 shares of our common stock, to include their shares in registration statements that we may file for ourselves or other stockholders. We also intend to register all shares of common stock that we may issue under our equity compensation plans. Once we register these shares, they can be freely sold in the public market upon issuance, subject to volume limitations applicable to affiliates and the lock-up agreements described in the "Underwriting" section of this prospectus.

You will incur immediate and substantial dilution as a result of this offering.

If you purchase common stock in this offering, you will incur immediate and substantial dilution of $ per share, representing the difference between the assumed initial public offering price of $ per share, the midpoint of the estimated price range set forth on the cover of this prospectus, and our pro forma net tangible book value per share after giving effect to this offering. Moreover, we issued options in the past to acquire common stock at prices significantly below the assumed initial public offering price. As of December 31, 2013, there were 7,744,480 shares of common stock subject to outstanding options with a weighted-average exercise price of $0.58 per share. To the extent that these outstanding options are ultimately exercised, you will incur further dilution.

We are an "emerging growth company," and the reduced disclosure requirements applicable to emerging growth companies may make our common stock less attractive to investors.

We are an "emerging growth company," as defined in the Jumpstart Our Business Startups Act of 2012, or the JOBS Act, and may remain an emerging growth company for up to five years. For so long as we remain an emerging growth company, we are permitted and intend to rely on exemptions from certain disclosure requirements that are applicable to other public companies that are not emerging growth companies. These exemptions include:

- providing only two years of audited financial statements, in addition to any required unaudited interim financial statements, with correspondingly reduced "Management's Discussion and Analysis of Financial Condition and Results of Operations" disclosure;
- not being required to comply with the auditor attestation requirements in the assessment of our internal control over financial reporting;
- not being required to comply with any requirement that may be adopted by the Public Company Accounting Oversight Board regarding mandatory audit firm rotation or a supplement to the auditor's report providing additional information about the audit and the financial statements;
- reduced disclosure obligations regarding executive compensation; and
- exemptions from the requirements of holding a nonbinding advisory vote on executive compensation and shareholder approval of any golden parachute payments not previously approved.

We may choose to take advantage of some, but not all, of the available exemptions. We have taken advantage of reduced reporting burdens in this prospectus. In particular, in this prospectus, we have provided only two years of audited financial statements and have not included all of the executive compensation related information that would be required if we were not an emerging growth company. We cannot predict whether investors will find our common stock less attractive if we rely on these exemptions. If some investors find our common stock less attractive as a result, there may be a less active trading market for our common stock and our stock price may be more volatile.

In addition, the JOBS Act also provides that an emerging growth company can take advantage of an extended transition period for complying with new or revised accounting standards. This allows an emerging growth company to delay the adoption of certain accounting standards until those standards would otherwise apply to private companies. We have irrevocably elected not to avail ourselves of this exemption from new or revised accounting standards and, therefore, we will be subject to the same new or revised accounting standards as other public companies that are not emerging growth companies.

We will incur increased costs as a result of operating as a public company, and our management will be required to devote substantial time to new compliance initiatives.

As a public company, and particularly after we are no longer an "emerging growth company," we will incur significant legal, accounting and other expenses that we did not incur as a private company. We expect that we will need to hire additional accounting, finance and other personnel in connection with our becoming, and our efforts to comply with the requirements of being, a public company, and our management and other personnel will need to devote a substantial amount of time towards maintaining compliance with these requirements. In addition, the Sarbanes-Oxley Act of 2002 and rules subsequently implemented by the Securities and Exchange Commission and NASDAQ have imposed various requirements on public companies, including establishment and maintenance of effective disclosure and financial controls and corporate governance practices. Our management and other personnel will need to devote a substantial amount of time to these compliance initiatives. Moreover, these rules and regulations will increase our legal and financial compliance costs and will make some activities more time-consuming and costly. For example, we expect that these rules and regulations may make it more difficult and more expensive for us to obtain director and officer liability insurance. Overall, we estimate that our incremental costs resulting from operating as a public company may be between $2.0 million and $4.0 million per year, which costs are in addition to our incremental costs resulting from operating as a stand-alone company.

Pursuant to Section 404 of the Sarbanes-Oxley Act of 2002, or Section 404, we will be required to furnish a report by our management on our internal control over financial reporting, including an attestation report on internal control over financial reporting issued by our independent registered public accounting firm. However, while we remain an emerging growth company, we will not be required to include an attestation report on internal control over financial reporting issued by our independent registered public accounting firm. We became a stand-alone company in February 2014 following the Spin-Out and, as such, have a very limited operating history. Accordingly, many of the internal controls over financial reporting have only recently been implemented and therefore have not been tested. To achieve compliance with Section 404 within the prescribed period, we will be engaged in a process to document and evaluate our internal control over financial reporting, which is both costly and challenging. In this regard, we will need to continue to dedicate internal resources, potentially engage outside consultants and adopt a detailed work plan to assess and document the adequacy of internal control over financial reporting, continue steps to improve control processes as appropriate, validate through testing that controls are functioning as documented and implement a continuous reporting and improvement process for internal control over financial reporting. Despite our efforts, there is a risk that neither we nor our independent registered public accounting firm will be able to conclude within the prescribed timeframe that our internal control over financial reporting is effective as required by Section 404. This could result in an adverse reaction in the financial markets due to a loss of confidence in the reliability of our financial statements.

We do not expect to pay any dividends for the foreseeable future. Investors in this offering may never obtain a return on their investment.

You should not rely on an investment in our common stock to provide dividend income. We do not anticipate that we will pay any dividends to holders of our common stock in the foreseeable future. Instead, we plan to retain any earnings to maintain and expand our existing operations. In addition, pursuant to our stockholders agreement, for as long as the New Mountain Entities and their assignees own at least 15% of our outstanding capital stock we may not pay or declare a dividend or distribution on any shares of our capital stock (other than dividends from a wholly-owned subsidiary to its parent company) without their approval. Accordingly, investors must rely on sales of their common stock after price appreciation, which may never occur, as the only way to realize any return on their investment. As a result, investors seeking cash dividends should not purchase our common stock.

We have broad discretion in the use of the net proceeds from this offering and may not use them effectively.

We cannot specify with certainty the particular uses of the net proceeds we will receive from this offering. Our management will have broad discretion in the application of the net proceeds, including for any of the purposes described in "Use of Proceeds." Accordingly, you will have to rely upon the judgment of our management with respect to the use of the proceeds, with only limited information concerning management's specific intentions. Our management may spend a portion or all of the net proceeds from this offering in ways that our stockholders may not desire or that may not yield a favorable return. The failure by our management to apply these funds effectively could harm our business. Pending their use, we may invest the net proceeds from this offering in a manner that does not produce income or that loses value.

Provisions in our certificate of incorporation, our bylaws or Delaware law might discourage, delay or prevent a change in control of our company or changes in our management and, therefore, depress the trading price of our common stock.

Provisions of our certificate of incorporation, our bylaws or Delaware law may discourage, delay or prevent a merger, acquisition or other change in control that stockholders may consider favorable, including transactions in which you might otherwise receive a premium for your shares of our common stock. These provisions may also prevent or frustrate attempts by our stockholders to change the composition of our board of directors or to replace or remove our management. These provisions include:

- limitations on the removal of directors;
- a classified board of directors so that not all members of our board are elected at one time;
- advance notice requirements for stockholder proposals and nominations;
- limitations on the ability of stockholders to call and bring business before special meetings and to take action by written consent in lieu of a meeting;
- limitations on the liability of, and the provision of indemnification to, our director and officers; and
- the ability of our board of directors to authorize the issuance of blank check preferred stock, which could be issued with voting, liquidation, dividend and other rights similar to our common stock.

In addition, we are subject to Section 203 of the Delaware General Corporation Law, which prohibits a publicly-held Delaware corporation from engaging in a business combination with an interested stockholder, generally a person which together with its affiliates owns, or within the last three years has owned, 15% of our voting stock, for a period of three years after the date of the transaction in which the person became an interested stockholder, unless the business combination is approved in a prescribed manner.

The existence of the foregoing provisions and anti-takeover measures could limit the price that investors might be willing to pay in the future for shares of our common stock. They could also deter potential acquirers of our company, thereby reducing the likelihood that you could receive a premium for your common stock in an acquisition.

If securities analysts do not publish research or reports about our business or if they publish negative evaluations of our stock, the price of our stock could decline.

The trading market for our common stock will rely in part on the research and reports that industry or financial analysts publish about us or our business. We do not currently have and may never obtain research coverage by industry or financial analysts. If no or few analysts commence coverage of us, the trading price of our stock would likely decrease. Even if we do obtain analyst coverage, if one or more of the analysts covering our business downgrade their evaluations of our stock, the price of our stock could decline. If one or more of these analysts cease to cover our stock, we could lose visibility in the market for our stock, which in turn could cause our stock price to decline.

SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS AND INDUSTRY DATA

This prospectus contains forward-looking statements that involve substantial risks and uncertainties. All statements, other than statements of historical facts, contained in this prospectus, including statements regarding our strategy, future operations, future financial position, future revenues, projected costs, prospects, plans and objectives of management, are forward-looking statements. The words "anticipate," "believe," "estimate," "expect," "intend," "may," "might," "plan," "predict," "project," "target," "potential," "will," "would," "could," "should," "continue" and similar expressions are intended to identify forward-looking statements, although not all forward-looking statements contain these identifying words.

The forward-looking statements in this prospectus include, among other things, statements about:

- the timing of the ongoing and expected clinical trials of our INOpulse and BCM product candidates, including statements regarding the timing of completion of the trials and the respective periods during which the results of the trials will become available;
- the timing of and our ability to obtain marketing approval of our product candidates, and the ability of our INOpulse and BCM product candidates to meet existing or future regulatory standards;
- our ability to operate, and the implementation of our business strategy, as a stand-alone company;
- our ability to comply with government laws and regulations;
- our commercialization, marketing and manufacturing capabilities and strategy;
- our estimates regarding the potential market opportunity for our product candidates;
- the timing of or our ability to enter into partnerships to market and commercialize our product candidates;
- the rate and degree of market acceptance of any product candidate for which we receive marketing approval;
- our intellectual property position;
- our expectations related to the use of proceeds from this offering;
- our estimates regarding expenses, future revenues, capital requirements and needs for additional funding and our ability to obtain additional funding;
- the success of competing treatments; and
- our competitive position.

We may not actually achieve the plans, intentions or expectations disclosed in our forward-looking statements, and you should not place undue reliance on our forward-looking statements. Actual results or events could differ materially from the plans, intentions and expectations disclosed in the forward-looking statements we make. We have included important factors in the cautionary statements included in this prospectus, particularly in the "Risk Factors" section, that we believe could cause actual results or events to differ materially from the forward-looking statements that we make. Our forward-looking statements do not reflect the potential impact of any future acquisitions, mergers, dispositions, joint ventures or investments we may make.

You should read this prospectus and the documents that we reference in this prospectus and have filed as exhibits to the registration statement of which this prospectus is a part completely and with the understanding that our actual future results may be materially different from what we expect. The forward-looking statements contained in this prospectus are made as of the date of this prospectus, and we do not assume any obligation to update any forward-looking statements, except as required by applicable law.

This prospectus includes statistical and other industry and market data that we obtained from industry publications and research, surveys and studies conducted by third parties. Industry publications and third-party research, surveys and studies generally indicate that their information has been obtained from sources believed to be reliable, although they do not guarantee the accuracy or completeness of such information. While we believe these industry publications and third-party research, surveys and studies are reliable, we have not independently verified such data.

USE OF PROCEEDS

We estimate that the net proceeds from our issuance and sale of shares of our common stock in this offering will be approximately $ million, assuming an initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. If the underwriters exercise in full their over-allotment option, we estimate that the net proceeds from this offering will be approximately $ million.

A $1.00 increase (decrease) in the assumed initial public offering price of $ per share would increase (decrease) the net proceeds to us from this offering by approximately $ million, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting estimated underwriting discounts and commissions. Each increase (decrease) of 1.0 million shares in the number of shares of common stock offered by us at the assumed initial public offering price would increase (decrease) the net proceeds to us for this offering by approximately $ million, assuming that the initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, remains the same and after deducting estimated underwriting discounts and commissions.

As of March 31, 2014, we had cash and cash equivalents of $58.4 million and restricted cash of $18.0 million. We currently estimate that we will use the net proceeds from this offering, together with our cash, cash equivalents and restricted cash, as follows:

- approximately $ to fund the Phase 3 clinical development of INOpulse for PAH;
- approximately $ to fund through completion our ongoing Phase 2 clinical trials of INOpulse for PAH and INOpulse for PH-COPD and our ongoing feasibility trial of BCM; and
- the remainder for working capital and other general corporate purposes.

This expected use of the net proceeds from this offering and our existing cash, cash equivalents and restricted cash represents our intentions based upon our current plans and business conditions. The amounts and timing of our actual expenditures may vary significantly depending on numerous factors, including the progress of our development and commercialization efforts, the status of and results from clinical trials, as well as any collaborations that we may enter into with third parties for our product candidates, and any unforeseen cash needs. As a result, our management will retain broad discretion over the allocation of the net proceeds from this offering. We have no current agreements, commitments or understandings for any material acquisitions or licenses of any products, businesses or technologies.

Based on our planned use of the net proceeds from this offering and our existing cash, cash equivalents and restricted cash described above, we estimate that such funds will be sufficient to enable us to fund our ongoing clinical trials of INOpulse for PAH, INOpulse for PH-COPD and BCM through completion, fund our planned Phase 3 clinical trial program of INOpulse for PAH and fund our operating expenses and capital requirements for at least the next months. We have based this estimate on assumptions that may prove to be wrong, and we could use our available capital resources sooner than we currently expect. We do not expect that the net proceeds from this offering and our existing cash, cash equivalents and restricted cash will be sufficient to enable us to fund the completion of development and commercialization of any of our product candidates.

Pending our use of the net proceeds from this offering, we intend to invest the net proceeds in short-term, interest-bearing, investment-grade securities.

DIVIDEND POLICY

We have never declared or paid cash dividends on our common stock. We currently intend to retain all of our future earnings, if any, to finance the growth and development of our business. We do not intend to pay cash dividends in respect of our common stock in the foreseeable future. In addition, pursuant to our stockholders agreement, for as long as the New Mountain Entities and their assignees own at least 15% of our outstanding capital stock we may not pay or declare a dividend or distribution on any shares of our capital stock (other than dividends from a wholly-owned subsidiary to its parent company) without their approval.

COMPANY CONVERSION

Prior to the effectiveness of the registration statement of which this prospectus forms a part, we will complete transactions pursuant to which we will convert Bellerophon Therapeutics LLC from a Delaware limited liability company into a Delaware corporation named Bellerophon Therapeutics, Inc., which we refer to as the Company Conversion. To consummate the Company Conversion, we will file a certificate of conversion with the Secretary of State of the State of Delaware. In connection with the Company Conversion:

- holders of our outstanding voting units will receive an equal number of shares of voting common stock for each unit held immediately prior to the Company Conversion;
- holders of our outstanding non-voting units will receive an equal number of shares of non-voting common stock for each non-voting unit held immediately prior to the Company Conversion; and
- options to purchase our non-voting units will become options to purchase non-voting shares of our common stock for each unit underlying such options immediately prior to the Company Conversion, at the same aggregate exercise price in effect prior to the Company Conversion.

Assuming the Company Conversion became effective as of April 30, 2014:

- 94,273,819 outstanding voting units of Bellerophon Therapeutics LLC would have converted into an aggregate of 94,273,819 shares of our voting common stock;
- 4,672,001 outstanding non-voting units of Bellerophon Therapeutics LLC would have converted into an aggregate of 4,672,001 shares of our non-voting common stock; and
- outstanding options to purchase 7,744,480 non-voting units of Bellerophon Therapeutics LLC would have become options to purchase an aggregate of 7,744,480 shares of our non-voting common stock, with exercise prices ranging from $0.02 to $1.43.

In connection with the Company Conversion, Bellerophon Therapeutics, Inc. will continue to hold all assets of Bellerophon Therapeutics LLC and will assume all of its liabilities and obligations. Bellerophon Therapeutics, Inc. will be governed by a certificate of incorporation filed with the Delaware Secretary of State and bylaws, the material portions of which are described in "Description of Capital Stock." On the effective date of the Company Conversion, the members of the board of directors of Bellerophon Therapeutics LLC will become members of the board of directors of Bellerophon Therapeutics, Inc. and the officers of Bellerophon Therapeutics LLC will become the officers of Bellerophon Therapeutics, Inc.

For the convenience of the reader, except as context otherwise requires, all information included in this prospectus is presented giving effect to the Company Conversion.

CAPITALIZATION

The following table sets forth our cash and cash equivalents, restricted cash and capitalization as of December 31, 2013:

- on an actual basis;
- on a pro forma basis (i) to give effect to Ikaria's distribution of $80.0 million of cash to us and the payment of the allocated portion of the Ikaria special dividend bonus amount in respect of certain Ikaria dividend equivalent rights that are described in our financial statements included elsewhere in this prospectus and (ii) without giving effect to the Company Conversion; and
- on a pro forma as adjusted basis to give effect to (i) the Company Conversion, (ii) our issuance and sale of shares of our common stock in this offering at an assumed initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us and (iii) the conversion of all of our outstanding non-voting common stock to voting common stock.

You should read this table together with our financial statements and the related notes appearing at the end of this prospectus and the "Management's Discussion and Analysis of Financial Condition and Results of Operations" section of this prospectus.

	December 31, 2013		
(in thousands)	**Actual**	**Pro Forma (unaudited)**	**Pro Forma As Adjusted (unaudited)**
Cash and cash equivalents	$ —	$ 61,500	
Restricted cash(1)	—	18,500	
Equity:			
Common stock, par value	—		
Additional paid-in capital	—		
Investment by Ikaria, net	160,778	246,890	
Deficit accumulated during the development stage	(176,515)	(176,515)	
Total invested (deficit) equity	(15,737)	70,375	
Total capitalization	$ (15,737)	$ 70,375	

(1) Represents cash deposited in escrow to pay amounts owed under the transition services agreement with Ikaria.

A $1.00 increase (decrease) in the assumed initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, would increase (decrease) each of cash and cash equivalents, additional paid-in capital, total invested (deficit) equity and total capitalization on a pro forma as adjusted basis by approximately $ million, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting estimated underwriting discounts and commissions. Each increase (decrease) of 1.0 million shares in the number of shares of common stock offered by us at the assumed initial public offering price would increase (decrease) each of cash and cash equivalents, additional paid-in capital, total invested (deficit) equity and total capitalization on a pro forma as adjusted basis by approximately $ million, assuming an initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, and after deducting estimated underwriting discounts and commissions.

DILUTION

If you invest in our common stock in this offering, your ownership interest will be diluted immediately to the extent of the difference between the public offering price per share of our common stock and the pro forma net tangible book value per share of our common stock after this offering.

Our historical net tangible book value as of March 31, 2014 was $ million, or $ per share of our common stock. Historical net tangible book value per share represents the amount of our total tangible assets less total liabilities, divided by the number of shares of our common stock outstanding, after giving effect to the Company Conversion.

After giving effect to our issuance and sale of shares of our common stock in this offering at an assumed initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us, our pro forma net tangible book value as of would have been $ million, or $ per share, in each case giving effect to the Company Conversion. This represents an immediate increase in pro forma net tangible book value per share of $ to existing stockholders and immediate dilution of $ in pro forma net tangible book value per share to new investors purchasing common stock in this offering. Dilution per share to new investors is determined by subtracting pro forma net tangible book value per share after this offering from the initial public offering price per share paid by new investors. The following table illustrates this dilution on a per share basis:

Assumed initial public offering price per share		$
Historical net tangible book value per share as of March 31, 2014	$	
Pro forma net tangible book value per share as of		
Increase in net tangible book value per share attributable to new investors		
Pro forma as adjusted net tangible book value per share after giving effect to this offering		
Dilution per share to new investors purchasing shares in this offering		$

A $1.00 increase (decrease) in the assumed initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, would increase (decrease) our pro forma net tangible book value by approximately $ or our pro forma net tangible book value per share by approximately $, and dilution per share to new investors by approximately $, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. Each increase (decrease) of 1.0 million shares in the number of shares of common stock offered by us at the assumed initial public offering price would increase (decrease) our pro forma net tangible book value by approximately $ or our pro forma net tangible book value per share by approximately $, and dilution per share to new investors by approximately $, assuming that the initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

If the underwriters exercise their over-allotment option or if any additional shares are issued in connection with outstanding options, you will experience further dilution.

The following table summarizes, on a pro forma basis as of , the total number of shares purchased from us, the total consideration paid, or to be paid, and the average price per share paid, or to be paid, by existing stockholders and by new investors in this offering at an assumed initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, before deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. As the table shows, new investors purchasing shares in this offering will pay an average price per share substantially higher than our existing stockholders paid.

	Shares Purchased		Total Consideration		Average Price Per Share
	Number	Percent	Amount	Percent	
Existing stockholders		%	$	%	$
New investors					
Total		100%		100%	

A $1.00 increase (decrease) in the assumed initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, would increase (decrease) the total consideration paid by new investors by $ million and increase (decrease) the percentage of total consideration paid by new investors by approximately %, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the same. Each increase (decrease) of 1.0 million shares in the number of shares of common stock offered by us at the assumed initial public offering price would increase (decrease) the total consideration paid by new investors by approximately $ million and increase (decrease) the percentage of total consideration paid by new investors by approximately %, assuming that the initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, remains the same.

If the underwriters exercise in full their over-allotment option, the following will occur:

- the percentage of shares of our common stock held by existing stockholders will decrease to approximately % of the total number of shares of our common stock outstanding after this offering; and
- the number of shares of our common stock held by new investors will increase to , or approximately % of the total number of shares of our common stock outstanding after this offering.

SELECTED FINANCIAL INFORMATION

The following selected financial information for the years ended December 31, 2013 and 2012 and for the period from August 26, 2009 (inception) to December 31, 2013 (as we are a development stage company) has been derived from our audited financial statements as of and for the years ended December 31, 2013 and 2012 and for the period from August 26, 2009 (inception) to December 31, 2013 included elsewhere in this prospectus. The selected financial data below should be read in conjunction with our historical and pro forma financial statements and the related notes included elsewhere in this prospectus, and the "Management's Discussion and Analysis of Financial Condition and Results of Operations" section of this prospectus.

Our financial statements include allocations of costs from certain shared functions provided to us by Ikaria, including general corporate and shared services expenses. These allocations were made either based on specific identification or the proportionate percentage of employee time or headcount to the respective total Ikaria employee time or headcount, as applicable, and have been included in our financial statements. Our historical results for any prior period are not necessarily indicative of results to be expected in any future period.

	Year Ended December 31,		Period from August 26, 2009 (inception) to December 31, 2013
(in thousands)	2013	2012	
Statement of Operations and Comprehensive Loss Information:			
Operating expenses:			
Research and development	$ 52,985	$ 38,727	$ 147,887
General and administrative	9,013	7,185	27,690
Other operating expense	—	315	938
Net loss and comprehensive loss	$ (61,998)	$ (46,227)	$ (176,515)

	As of December 31,		As of December 31, 2013	
(in thousands)	2013	2012	Pro Forma(1)	Pro Forma As Adjusted(2)
Balance Sheet Information:			(unaudited)	(unaudited)
Cash and cash equivalents	$ —	—	61,500	
Restricted cash(3)	—	—	18,500	
Working (deficit) capital	(12,440)	(10,892)	60,149	
Total assets	3,636	3,349	83,636	
Allocated portion of Ikaria special dividend bonus payable	4,273	2,865	—	
Other non-current liabilities	1,108	389	1,108	
Invested (deficit) equity	(15,737)	(11,116)	70,375	

(1) The pro forma balance sheet data (i) gives effect to Ikaria's distribution of $80.0 million of cash to us and the payment of the allocated portion of the Ikaria special dividend bonus amount in respect of certain Ikaria dividend equivalent rights that are described in our financial statements included elsewhere in this prospectus and (ii) does not give effect to the Company Conversion.

(2) The pro forma as adjusted balance sheet data gives further effect to (i) the Company Conversion, (ii) our issuance and sale of shares of our common stock in this offering at an assumed initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us and (iii) the conversion of all of our outstanding non-voting common stock to voting common stock.

(3) Represents cash deposited into escrow to pay amounts owed under the transition services agreement with Ikaria.

A $1.00 increase (decrease) in the assumed initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, would increase (decrease) the pro forma as adjusted amount of each of cash and cash equivalents, working (deficit) capital, total assets and invested (deficit) equity by approximately $ million, assuming that the number of shares offered by us, as set forth on the cover page of this prospectus, remains the

same and after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us. Each increase (decrease) of 1.0 million shares in the number of shares of common stock offered by us at the assumed initial public offering price would increase (decrease) the pro forma as adjusted amount of each of cash and cash equivalents, working (deficit) capital, total assets and total invested (deficit) equity by $ million, assuming an initial public offering price of $ per share, which is the midpoint of the estimated price range set forth on the cover page of this prospectus, after deducting estimated underwriting discounts and commissions and estimated offering expenses payable by us.

MANAGEMENT'S DISCUSSION AND ANALYSIS OF FINANCIAL CONDITION AND RESULTS OF OPERATIONS

You should read the following discussion and analysis of our financial condition and results of operations together with our financial statements and the related notes appearing in this prospectus. Some of the information contained in this discussion and analysis or set forth elsewhere in this prospectus, including information with respect to our plans and strategy for our business and related financing, includes forward-looking statements that involve risks and uncertainties. You should read the "Risk Factors" section of this prospectus for a discussion of important factors that could cause our actual results to differ materially from the results described in or implied by the forward-looking statements contained in the following discussion and analysis.

Overview

Business

We are a clinical stage biotherapeutics company focused on developing innovative products at the intersection of drugs and devices that address significant unmet medical needs in the treatment of cardiopulmonary and cardiac diseases. We are developing three product candidates. Two of our product candidates are based on our proprietary pulsatile nitric oxide delivery device, which we refer to as INOpulse, and are in Phase 2 clinical trials—one for the treatment of pulmonary arterial hypertension, or PAH, and a second for the treatment of pulmonary hypertension associated with chronic obstructive pulmonary disease, or PH-COPD. Our third product candidate, bioabsorbable cardiac matrix, or BCM, is currently in a feasibility clinical trial (which is a CE mark registration trial in the European Union and is comparable to a Phase 2 clinical trial in U.S. drug development). We are developing BCM for the prevention of cardiac remodeling and progression to congestive heart failure following acute myocardial infarction, commonly known as a heart attack.

We have developed a drug delivery system, INOpulse, that operates through the administration of nitric oxide as brief, controlled pulses through our proprietary triple-lumen nasal cannula that are timed to occur at the beginning of each breath. We have devoted significant time and resources to develop and optimize INOpulse. We have also incurred material costs to scale up manufacturing for BCM from pre-clinical studies to clinical trials.

We have devoted substantially all of our resources to our drug discovery and development efforts, including conducting clinical trials for our product candidates, protecting our intellectual property and the general and administrative support of these operations. To date, we have generated no revenue from product sales. We expect that it will be several years before we commercialize a product candidate, if ever.

Separation and Spin-Out from Ikaria

Prior to February 2014, we were a wholly-owned subsidiary of Ikaria. As part of an internal reorganization of Ikaria in October 2013, Ikaria transferred to us exclusive worldwide rights, with no royalty obligations, to develop and commercialize INOpulse in PAH, PH-COPD and pulmonary hypertension associated with idiopathic pulmonary fibrosis. Following the internal reorganization, in February 2014, Ikaria distributed all of our outstanding units to its stockholders through the payment of a special dividend to its stockholders on a pro rata basis based on each stockholder's ownership of Ikaria capital stock, which we refer to as the Spin-Out, and as a result we became a stand-alone company. For purposes of our financial statements, our inception date is August 26, 2009, which is the date that BCM was licensed to us by BioLineRx Ltd. and its subsidiary, or BioLine. Our operations since that date have included organization and staffing, business planning, in-licensing technology from research institutions, developing product candidates in clinical programs, evaluating potential future

product candidates, as well as undertaking pre-clinical studies and clinical trials of our product candidates.

We are in the process of developing and implementing plans to replace services currently provided to us by Ikaria under our transition services agreement, which we refer to as the TSA. These services include, among others, accounting and financial management support, human resources support, drug and drug safety services, biometrics support and manufacturing support. We expect the costs related to replacing the services currently provided by Ikaria will be approximately the same as the $772,000 per month that we are currently paying under the TSA. Although we believe our estimates are reasonable based on the information we have to date, certain significant components of our estimates are preliminary and subject to change.

Our historical financial statements included in this prospectus and discussed in this Management's Discussion and Analysis of Financial Condition and Results of Operations have been derived from the audited historical financial statements and accounting records of Ikaria for time periods presented. The financial information included in this prospectus, however, does not reflect what our financial position, results of operations and cash flows will be in the future or what our financial position, results of operations and cash flows would have been in the past had we been a public, stand-alone company during the periods presented.

Financial Position and Outlook

Since inception, we have never been profitable and have incurred significant operating losses. Our net losses were $62.0 million and $46.2 million for the years ended December 31, 2013 and 2012, respectively, and $176.5 million for our inception to date period, all of which was funded by Ikaria.

We expect to continue to incur significant expenses and operating losses for the foreseeable future as we continue the development and clinical trials of, and seek regulatory approval for, our product candidates. If we obtain regulatory approval for any of our product candidates, we expect to incur significant commercialization expenses. We do not currently have the infrastructure for the sale, marketing, manufacture and distribution of any products. To develop a commercial infrastructure, we will have to invest financial and management resources, some of which would have to be deployed prior to having any certainty of marketing approval.

We have entered into a license agreement with each of Ikaria and BioLine. In the future, we may enter into additional licensing and/or co-promotion agreements with strategic partners for the commercialization of our product candidates in the United States and other countries.

Following consummation of this offering, we expect to incur additional costs associated with operating as a public company. Unless and until we generate sufficient revenue to be profitable, we will seek to fund our operations primarily through public or private equity or debt financings or other means, which may include strategic partnerships with third parties in the United States or abroad in certain or all of our programs. Other additional financing may not be available to us on acceptable terms, or at all. Our failure to raise capital as and when needed could have a material adverse effect on our business, results of operations, financial condition, cash flows and future prospects.

Financial Operations Overview

Revenue

To date, we have not generated any revenue from product sales and do not expect to generate any revenue from product sales for the next several years, if ever. In the future, we may generate revenue from a combination of product sales, license fees, milestone payments and royalties from the sale of products developed under licenses of our intellectual property. Our ability to generate revenue and

become profitable depends primarily on our ability to successfully develop and commercialize or partner our INOpulse and/or BCM product candidates, each of which is currently in clinical development, as well as any product candidates we may advance in the future. We expect that any revenue we may generate will fluctuate from quarter to quarter as a result of the timing and amount of license fees, milestone and other payments, and the amount and timing of payments that we receive from the sale of our products, to the extent any are successfully commercialized. If we fail to complete the development of any of our three product candidates currently in Phase 2 clinical development or any future product candidates in a timely manner, or to obtain regulatory approval for such product candidates, our ability to generate future revenue, and our business, results of operations, financial condition and cash flows and future prospects would be materially adversely affected.

Research and Development Expenses

Research and development expenses consist of costs incurred in connection with the discovery and development of our product candidates.

In order to fairly present our historical information, certain departmental expenses from Ikaria have been allocated to us. The allocations were applied to us for the purpose of presenting our company as a stand-alone entity. Direct and indirect costs related to the INOpulse for PAH, INOpulse for PH-COPD and BCM clinical programs have been allocated to us. All allocations were based on actual costs incurred. For purposes of allocating non-project specific expenses, each departmental head provided information as to the percentage of employee time incurred on our behalf.

These expenses primarily consist of:

- employee-related expenses, including salary, benefits and allocated stock-based compensation expense;
- expenses incurred under agreements with contract research organizations, investigative sites that conduct our clinical trials and consultants that conduct a portion of our pre-clinical studies;
- expenses relating to vendors in connection with research and development activities;
- the cost of acquiring and manufacturing clinical trial materials;
- facilities, depreciation of fixed assets and other allocated expenses;
- lab supplies, reagents, active pharmaceutical ingredients and other direct and indirect costs in support of our pre-clinical and clinical activities;
- device development and drug manufacturing engineering;
- license fees related to in-licensed products and technology; and
- costs associated with non-clinical activities and regulatory approvals.

We expense research and development costs as incurred.

Conducting a significant amount of research and development is central to our business model. Product candidates in late stages of clinical development generally have higher development costs than those in earlier stages of clinical development primarily due to the increased size and duration of late-stage clinical trials. We plan to increase our research and development expenses for the foreseeable future as we seek to continue multiple clinical trials of our INOpulse and BCM product candidates and potentially advance earlier stage research and development projects.

INOpulse for PAH

We commenced a randomized, placebo-controlled Phase 2 clinical trial of INOpulse for PAH in April 2012. We plan to enroll at least 78 patients in this trial at 52 clinical sites in the United States and Canada. The goal of the trial is to determine the safety, tolerability and efficacy of two different doses of INOpulse for PAH. We expect to complete this trial by the end of 2014. As of April 30, 2014, we had enrolled 74 patients.

INOpulse for PH-COPD

We commenced a randomized, placebo-controlled, dose-confirmation Phase 2 clinical trial of INOpulse for PH-COPD in December 2012. We plan to enroll at least 159 patients in this trial at 43 clinical sites in the United States. We expect this trial to provide insight on any acute dose-dependent effects of inhaled nitric oxide in this patient population, which will inform dosing for the next phase of development. We expect to complete this trial by mid-2014. As of April 30, 2014, we had enrolled 150 patients.

BCM

We initiated a feasibility clinical trial of BCM, which we refer to as PRESERVATION I, in December 2011 and enrolled the first patient in April 2012. This trial is a CE mark registration trial for European Union regulatory purposes and is comparable to a Phase 2 clinical trial in U.S. drug development. We plan to enroll approximately 300 patients in this trial at up to approximately 90 sites in Europe, Australia, North America and Israel. We expect to complete this trial by mid-2015. As of April 30, 2014, we had enrolled 202 patients.

Research and Development Infrastructure

We invest in regulatory, quality, pharmacovigilance and program management activities, which are expensed as incurred. These activities primarily support our INOpulse for PAH, INOpulse for PH-COPD and BCM clinical development programs.

INOpulse Engineering and Manufacturing

The INOpulse device is configured to be highly portable and compatible with available modes of long-term oxygen therapy via nasal cannula delivery. Our ongoing clinical trials for PAH and PH-COPD are utilizing the first generation INOpulse DS device. We are near completion of a next generation INOpulse Mark2 device, or the Mark2, which we believe will significantly improve several characteristics of our INOpulse delivery system, but will require prototype manufacturing and bench top testing as well as verification and validation. We have also invested in design and engineering technology, through Ikaria, for the manufacture of our drug cartridges. We currently rely on Ikaria for manufacturing of our INOpulse devices and drug cartridges. Our current device manufacturing agreement with Ikaria terminates in February 2015. We plan to either negotiate a new agreement with Ikaria following termination of the device manufacturing agreement or enter into an arrangement with another third-party manufacturer. In addition to manufacturing our INOpulse delivery system, Ikaria is conducting substantial engineering and stability testing work with respect to the INOpulse devices on our behalf pursuant to the TSA.

In 2014, we plan to continue to invest in our three clinical development programs. For INOpulse for PAH, we expect to complete our ongoing Phase 2 clinical trial by the end of 2014. In addition, during 2014 we plan to continue to invest in the development of the Mark2. For INOpulse for PH-COPD, we plan to complete our ongoing Phase 2 dose-confirmation trial by mid-2014. For BCM, we plan to enroll the final patients in PRESERVATION I by the end of 2014.

It is difficult to determine with certainty the duration and completion costs of our current or future pre-clinical programs and clinical trials of our INOpulse and BCM product candidates, and any of our future product candidates we may advance, or if, when or to what extent we will generate revenue from the commercialization and sale of any of our product candidates that obtain regulatory approval. We may never succeed in achieving regulatory approval for any of our product candidates. The duration, costs and timing of clinical trials and development of our product candidates will depend on a variety of factors, including the uncertainties of future clinical trials and pre-clinical studies, uncertainties in clinical trial enrollment rate and significant and changing government regulation. In addition, the probability of success for each product candidate will depend on numerous factors, including competition, manufacturing capability and commercial viability. A change in the outcome of any of these variables with respect to the development of a product candidate could mean a significant change in the costs and timing associated with the development of that product candidate. For example, if the U.S. Food and Drug Administration or other regulatory authority were to require us to conduct clinical trials beyond those that we currently anticipate will be required for the completion of clinical development of a product candidate, or if we experience significant delays in enrollment in any of our clinical trials, we could be required to expend significant additional financial resources and time with respect to the development of that product candidate. We will determine which programs to pursue and how much to fund each program in response to the scientific and clinical success of each product candidate, as well as an assessment of each product candidate's commercial potential, including the likelihood of regulatory approval on a timely basis.

General and Administrative Expenses

General and administrative expenses consist principally of salaries and costs related to executive, finance, business development, marketing, legal and human resources functions, either through direct expenses or through the TSA. Other general and administrative expenses include patent filing, patent prosecution, professional fees for legal, insurance, consulting, information technology and auditing and tax services not otherwise included in research and development expenses.

We anticipate that our general and administrative expenses will increase in the future for the following reasons, among others:

- we expect to incur increased general and administrative expenses to support ourselves as a stand-alone company such as investing in a new general ledger system and new telecommunications services;
- we expect to incur, prior to the termination of the TSA, expenses in preparation for replacing services that are currently provided by Ikaria pursuant to the TSA, which will likely include dedicated accounting and human resources functions;
- we expect to incur increased general and administrative expenses to support our research and development activities, which we expect to expand as we continue to pursue the development of our product candidates;
- we expect our general and administrative expenses to increase as a result of increased payroll, expanded infrastructure, higher consulting, legal, accounting and investor relations costs, director compensation and director and officer insurance premiums associated with being a public company; and
- we may begin to incur expenses related to sales and marketing of our product candidates in anticipation of commercial launch before we receive regulatory approval of a product candidate.

Results of Operations

Comparison of Years Ended December 31, 2013 and 2012

The following table summarizes our results of operations for the years ended December 31, 2013 and 2012, together with the changes in these items in dollars and as a percentage.

(Dollar amounts in thousands)	Year Ended December 31, 2013	2012	$ Change	% Change
Research and development expenses:				
BCM	$ 17,266	$ 14,347	$ 2,919	20%
PAH	8,099	8,544	(445)	(5)
PH-COPD	8,420	1,767	6,653	377
Clinical programs	33,785	24,658	9,127	37
Research and development infrastructure	14,000	10,387	3,613	35
INOpulse engineering	5,200	3,682	1,518	41
Total research and development expenses	52,985	38,727	14,258	37
General and administrative	9,013	7,185	1,828	25
Other operating expenses	—	315	(315)	(100)
Total operating expenses	61,998	46,227	15,771	34
Net loss and comprehensive loss	$ (61,998)	$ (46,227)	$ (15,771)	34%

Total Operating Expenses. Total operating expenses for the year ended December 31, 2013 were $62.0 million compared to $46.2 million for the year ended December 31, 2012, an increase of $15.8 million, or 34%. This increase was primarily due to an increase in research and development expenses pertaining to our BCM and INOpulse for PH-COPD clinical programs, research and development infrastructure, INOpulse engineering and manufacturing, and general and administrative expenses.

Research and Development Expenses. Total research and development expenses for the year ended December 31, 2013 were $53.0 million compared to $38.7 million for the year ended December 31, 2012, an increase of $14.3 million, or 37%. Total research and development expenses consisted of the following:

- BCM research and development expenses for the year ended December 31, 2013 were $17.3 million compared to $14.3 million for the year ended December 31, 2012, an increase of $2.9 million, or 20%. The increase was primarily due to increased enrollment in PRESERVATION I to 120 patients in 2013 from 19 patients in 2012.

- PAH research and development expenses for the year ended December 31, 2013 were $8.1 million compared to $8.5 million for the year ended December 31, 2012, a decrease of $0.4 million, or 5%. The decrease was primarily due to a smaller number of devices being manufactured for our INOpulse for PAH trial in 2013 as compared to 2012, partially offset by increased patient enrollment in the Phase 2 clinical trial of INOpulse for PAH to 47 patients in 2013 from ten patients in 2012.

- PH-COPD research and development expenses for the year ended December 31, 2013 were $8.4 million compared to $1.8 million for the year ended December 31, 2012, an increase of $6.7 million, or 377%. The increase resulted from commencement of the first part of the Phase 2 clinical trial of INOpulse for PH-COPD in 2013.

- Research and development infrastructure expenses for the year ended December 31, 2013 were $14.0 million compared to $10.4 million for the year ended December 31, 2012, an increase of $3.6 million, or 35%. The increase was primarily due to a higher level of professional and consulting fees to support our INOpulse and BCM clinical programs, including those related to program risk analysis, regulatory, biometrics and drug and device safety in 2013.

- INOpulse engineering expenses for the year ended December 31, 2013 were $5.2 million compared to $3.7 million for the year ended December 31, 2012, an increase of $1.5 million, or 41%. The increase was primarily due to increased engineering activity related to the INOpulse devices in 2013.

General and Administrative Expenses. General and administrative expenses for the year ended December 31, 2013 were $9.0 million compared to $7.2 million for the year ended December 31, 2012, an increase of $1.8 million, or 25%. The increase was primarily due to allocated finance costs.

Other Operating Expenses. In 2012, we incurred a $0.3 million restructuring charge resulting from the closing of the research and development facility in Seattle, Washington, as we moved research and development operations to our facilities in North Brunswick, New Jersey.

Liquidity and Capital Resources

Since our inception, we have incurred net losses and negative cash flows from our operations. We incurred net losses of $62.0 million and $46.2 million for the years ended December 31, 2013 and 2012, respectively. Our operating activities used $57.2 million and $36.2 million of cash during the years ended December 31, 2013 and 2012, respectively, and $156.6 million for our inception to date period, all of which was funded by Ikaria. In addition, we had negative working capital of $12.4 million and no cash, cash equivalents, restricted cash or short-term investments as of December 31, 2013.

In connection with the Spin-Out, we received $80.0 million of cash from Ikaria. As of March 31, 2014, we had cash and cash equivalents of $58.4 million and restricted cash of $18.0 million, which are expected to be sufficient to satisfy our operating cash needs at least through December 31, 2014.

Cash Flows

The following table summarizes our cash flows for the years ended December 31, 2013 and 2012:

	Year Ended December 31,			
(Dollar amounts in thousands)	**2013**	**2012**	**$ Change**	**% Change**
Net cash (used in) provided by:				
Operating activities	$ (57,231)	$ (36,224)	$ (21,007)	58%
Investing activities	(727)	(3,478)	2,751	(79)
Financing activities	57,958	39,702	18,256	46
Change in cash and cash equivalents	$ —	$ —	$ —	

Net Cash Used in Operating Activities

Cash used in operating activities for the year ended December 31, 2013 was $57.2 million compared to $36.2 million for the year ended December 31, 2012, an increase of $21.0 million, or 58%. The increase was driven by an increase in clinical development expenses as a result of the increased activity in the INOpulse for PAH, INOpulse for PH-COPD and BCM clinical programs.

Net Cash Used in Investing Activities

Cash used in investing activities for the year ended December 31, 2013 was $0.7 million compared to $3.5 million for the year ended December 31, 2012, a decrease of $2.8 million, or 79%. The decrease in cash used in investing activities for the year ended December 31, 2013 compared to the year ended December 31, 2012 was primarily the result of a reduction in capital expenditures due to the timing of device investments to support our clinical trials.

Net Cash Provided by Financing Activities

Cash provided by financing activities for the year ended December 31, 2013 was $58.0 million compared to $39.7 million for the year ended December 31, 2012, an increase of $18.3 million, or 46%. The increase was primarily due to the increased net investment by Ikaria in 2013 that is discussed in Note 9 of the notes to our financial statements appearing elsewhere in this prospectus.

Plan of Operations and Future Funding Requirements

Our primary uses of capital are, and we expect will continue to be, compensation and related expenses, third-party clinical research and development services, contract manufacturing services, laboratory and related supplies, clinical costs, legal and other regulatory expenses and general overhead costs.

We expect that the net proceeds from this offering, together with our cash, cash equivalents and restricted cash as of March 31, 2014, will fund our operating expenses and capital expenditure requirements through , which we expect will enable us to fund our ongoing clinical trials of INOpulse for PAH, INOpulse for PH-COPD and BCM through completion and fund our planned Phase 3 clinical trial program of INOpulse for PAH. We have based these estimates on assumptions that may prove to be wrong, and we may exhaust our capital resources sooner than we expect. In addition, the process of testing product candidates in clinical trials is costly, and the timing of progress in clinical trials is uncertain. Because our product candidates are in Phase 2 development and the outcome of these efforts is uncertain, we cannot estimate the actual amounts necessary to successfully complete the development and commercialization of our product candidates or whether, or when, we may achieve profitability. Our future capital requirements will depend on many factors, including:

- the progress and results of our clinical trials of INOpulse for PAH, INOpulse for PH-COPD and BCM;
- our ability to manufacture sufficient supply of our product candidates and costs thereof;
- discussions with regulatory agencies regarding the design and conduct of our clinical trials and the costs, timing and outcome of regulatory review of our product candidates;
- the costs and timing of future commercialization activities, including product manufacturing, marketing, sales and distribution for any of our product candidates for which we receive marketing approval;
- the number and development requirements of any other product candidates we pursue;
- our ability to enter into collaborative agreements and achieve milestones under those agreements;
- the revenue, if any, received from commercial sales of our product candidates for which we receive marketing approval;
- the costs and timing of preparing, filing and prosecuting patent applications, maintaining and enforcing our intellectual property rights and defending any intellectual property-related claims;

- our expenses as a stand-alone company; and
- the extent to which we acquire or in-license other products and technologies.

Until such time, if ever, as we can generate substantial product revenues, we expect to finance our cash needs through a combination of equity and debt offerings, existing working capital and potential future collaboration arrangements. To the extent that we raise additional capital through the future sale of equity or debt, the ownership interest of our existing stockholders will be diluted, and the terms of such securities may include liquidation or other preferences that adversely affect the rights of our existing stockholders. If we raise additional funds through strategic partnerships in the future, we may have to relinquish valuable rights to our technologies, future revenue streams or product candidates or grant licenses on terms that may not be favorable to us. If we are unable to raise additional funds through equity or debt financings when needed, we may be required to delay, limit, reduce or terminate our product development or future commercialization efforts or grant rights to develop and market product candidates that we would otherwise prefer to develop and market ourselves.

Contractual Obligations and Commitments

The following is a summary of our long-term contractual cash obligations as of December 31, 2013 (in thousands):

	Payments Due by Period ($)				
Contractual Obligations	**Total**	**Less than 1 year**	**1 to 3 years**	**3 to 5 years**	**More than 5 years**
Operating Lease Obligations(1)	28	28	—	—	—

(1) Operating lease obligations reflect our obligation to make payments in connection with a lease for our operating facilities. The amounts in the table do not include (i) approximately $100,000 of milestone rent payable upon the closing of this offering or (ii) our rent obligation of $113,400 through March 15, 2015 under a lease that we signed subsequent to December 31, 2013.

Under the TSA, Ikaria provides certain administrative and other services to us for a period of 24 months following February 9, 2014, unless terminated earlier. Ikaria also provides us with the use of office space and research laboratory facilities at Ikaria's headquarters located in Hampton, New Jersey. In exchange for the services provided by Ikaria pursuant to the TSA, we pay to Ikaria a service fee in the amount of $772,000 per month and reimburse Ikaria for any out-of-pocket expenses, any taxes imposed on Ikaria in connection with the provision of services under the TSA and Ikaria's costs and expenses incurred in connection with the performance of any extraordinary services. The monthly service fee is payable by us regardless of the frequency or quantity of services actually utilized by us, and our obligation to pay such monthly service fee for 24 months will survive any early termination of the TSA. At the time of the Spin-Out, we deposited the sum of $18.5 million, representing the aggregate of the $772,000 monthly service fees payable by us under the TSA, in escrow to guarantee payment of the monthly service fees.

Milestone and royalty payments associated with our license agreement with BioLine have not been included in the above table of contractual obligations as we cannot reasonably estimate if or when they will occur. Under the terms of the license agreement, if we achieve certain clinical and regulatory events specified in the license agreement, we will be obligated to pay milestone payments to BioLine, which could total, in the aggregate, up to $115.5 million, and if we achieve certain commercialization targets specified in the license agreement, we will be obligated to pay additional milestone payments to BioLine, which could total, in the aggregate, up to $150.0 million. In addition, we will be obligated to pay BioLine a specified percentage of any upfront consideration we receive for sublicensing BCM, as well as royalties at a percentage in the low double digits below 20% on net sales, if any, of any

approved product containing BCM, subject to offsets for specified payments to third parties made in connection with such product.

BioLine has indicated to us that it believes that we have breached the license agreement in several ways, including, but not limited to, failure to use commercially reasonable efforts to develop BCM, failure to provide BioLine with material information concerning the development and commercialization plans for BCM and failure to notify BioLine in advance of material public disclosures regarding BCM. We and BioLine also disagree about the timing of a $12.5 million milestone payment that we would owe BioLine based upon progress in our BCM clinical development program. We believe we have complied with our obligations under the license agreement to use commercially reasonable efforts to develop BCM and are not currently in breach of our other obligations under the licence agreement. Although we have had multiple discussions with BioLine on these issues, and these discussions are continuing, we have not been able to resolve these outstanding issues. If we are unable to reach agreement with BioLine on these issues, BioLine could bring a lawsuit against us, although any claims relating to our alleged failure to use commercially reasonable efforts would first be subject to a non-binding 60-day mediation period with a third-party mediator. We believe that we would have strong defenses in any litigation that could be brought by BioLine. If BioLine were to prevail in any such litigation, one of the potential remedies would be the return of BCM to BioLine. In addition, if BioLine were to prevail in any such litigation, or if we were required to pay the milestone in dispute sooner than we had planned, or if we were required to return BCM to BioLine, these events could have a material adverse effect on our business, results of operations, financial condition and/or liquidity.

In the course of our normal business operations, we also enter into agreements with contract service providers and others to assist in the performance of our research and development and manufacturing activities. We can elect to discontinue the work under these contracts and purchase orders at any time with notice, and such contracts and purchase orders do not contain minimum purchase obligations.

Off-Balance Sheet Arrangements

We did not have during the periods presented, and we do not currently have, any off-balance sheet arrangements, as defined under applicable Securities and Exchange Commission rules.

Critical Accounting Policies and Significant Judgments and Estimates

Our management's discussion and analysis of our financial condition and results of operations is based on our financial statements, which have been prepared in accordance with accounting principles generally accepted in the United States. The preparation of these financial statements requires us to make estimates and judgments that affect the reported amounts of assets, liabilities, revenue and expenses and the disclosure of contingent assets and liabilities in our financial statements. On an ongoing basis, we evaluate our estimates and judgments, including those related to research and development expense, impairment of long-lived assets, stock-based compensation and income taxes. We base our estimates on historical experience, known trends and events and various other factors that we believe to be reasonable under the circumstances, the results of which form the basis for making judgments about the carrying values of assets and liabilities that are not readily apparent from other sources. Actual results may differ from these estimates under different assumptions or conditions.

While our significant accounting policies are described in Note 2 of the notes to our financial statements appearing elsewhere in this prospectus, we believe the following accounting policies to be most critical to the judgments and estimates used in the preparation of our financial statements.

Research and Development Expense

Research and development costs are expensed as incurred. These expenses include the costs of our proprietary research and development efforts, as well as costs incurred in connection with certain licensing arrangements. Upfront and milestone payments made to third parties in connection with research and development collaborations are expensed as incurred up to the point of regulatory approval. Payments made to third parties upon or subsequent to regulatory approval are capitalized and amortized over the remaining useful life of the related product. We also expense the cost of purchased technology and equipment in the period of purchase if we believe that the technology or equipment has not demonstrated technological feasibility and it does not have an alternative future use. Nonrefundable advance payments for goods or services that will be used or rendered for future research and development activities are deferred and are recognized as research and development expense as the related goods are delivered or the related services are performed.

As part of the process of preparing our financial statements, we are required to estimate our accrued research expenses. This process involves reviewing open contracts and purchase orders, communicating with applicable personnel to identify services that have been performed on our behalf and estimating the level of service performed and the associated cost incurred for the service when we have not yet been invoiced or otherwise notified of actual cost. The majority of our service providers invoice us monthly in arrears for services performed. We make estimates of our accrued research and development expenses as of each balance sheet date in our financial statements based on facts and circumstances known to us at that time. We periodically confirm the accuracy of our estimates with the service providers and make adjustments if necessary. Examples of estimated accrued research and development expenses include:

- fees paid to contract research organizations in connection with clinical trials;
- fees paid to investigative sites in connection with clinical trials;
- fees paid to contract manufacturers in connection with the production of clinical trial materials; and
- fees paid to vendors in connection with the pre-clinical development activities.

We base our expenses related to research and development and clinical trials on our estimates of the services received and efforts expended pursuant to contracts with multiple third parties, including research institutions and contract research organizations that conduct and manage clinical trials on our behalf. The financial terms of these agreements are subject to negotiation, vary from contract to contract and may result in uneven payment flows. Payments under some of these contracts depend on factors such as the successful enrollment of patients and the completion of clinical trial milestones. In accruing the research and development service fees, we consider the terms of each agreement, the time period over which the services will be performed and the level of effort required to complete the service. If the actual timing of the performance of the services or the level of effort varies from our estimate, we adjust the accrual accordingly. Although we do not expect our estimates to be materially different from amounts actually incurred, our understanding of the status and timing of services performed relative to the actual status and timing of services performed may vary and may result in us reporting amounts that are too high or too low in any particular period. Based on our level of accrued research and development expenses as of December 31, 2013, if our estimates are too high or too low by 5%, this may result in an adjustment to our accrued research and development expenses in future periods of approximately $450,000.

Impairment of Long-Lived Assets

Long-lived assets, such as property, plant and equipment, are reviewed for impairment whenever events or changes in circumstances indicate that the carrying amount of an asset may not be

recoverable. Recoverability of assets to be held and used is measured by a comparison of the carrying amount of an asset to estimated undiscounted expected future cash flows. If the carrying amount of an asset exceeds its estimated future cash flows, an impairment charge is recognized for the amount by which the carrying amount of the asset exceeds the fair value of the asset. Assets to be sold are no longer depreciated and are reclassified outside of property, plant and equipment at the lower of the carrying amount or fair value less costs to sell.

Stock-Based Compensation

We issue, and prior to the Spin-Out Ikaria issued, stock-based awards to employees and non-employees in the form of stock options and restricted stock units, or RSUs. The stock-based compensation expense recorded for the periods presented in our audited financial statements, included elsewhere in this prospectus, represents an allocation of Ikaria's stock-based compensation expense for employees and non-employees whose time was attributed to our business prior to the Spin-Out and, as a result, has been allocated to us for accounting purposes.

Ikaria applied the fair value recognition provisions of the Financial Accounting Standards Board, or FASB, Accounting Standards Codification, or ASC, Topic 718, Compensation-Stock Compensation, or ASC 718. ASC 718 requires all stock-based payments to employees, including grants of employee stock options and RSUs and modifications to existing stock options and RSUs, to be recognized in the statements of operations based on their fair values. Ikaria recognized and we will recognize the compensation expense of stock-based awards on a straight-line basis over the vesting period of the award for employees and non-employees. Compensation expense related to stock-based awards is subject to a number of estimates, including the estimated volatility and underlying fair value of our common stock, as well as the estimated life of the awards.

Ikaria estimated and we will estimate the fair value of its stock-based awards to employees and non-employees using the Black-Scholes-Merton option-pricing model, which requires the input of highly subjective assumptions, including (a) the fair value of the underlying stock, (b) the expected volatility of the underlying stock, (c) the expected term of the award, (d) the risk-free interest rate and (e) expected dividends. Due to the lack of a public market for the trading of Ikaria common stock and a lack of company-specific historical and implied volatility data, Ikaria based its estimate of expected volatility on the historical volatility of a group of similar companies that are publicly traded. For the volatility analyses, Ikaria selected companies with comparable characteristics to Ikaria, including enterprise value, risk profile and position within the industry, and with historical share price information sufficient to meet the expected life of the stock-based awards. Ikaria computed the historical volatility data using the daily closing prices for the selected companies' shares during the equivalent period of the calculated expected term of Ikaria's stock-based awards. We will also apply this process for purposes of our future stock-based compensation expense until a sufficient amount of historical information regarding the volatility of our own stock price becomes available. Because Ikaria had minimal historical information to develop expectations about future exercise patterns for its stock option grants, the expected term is based on an average of the expected term of options granted by Ikaria's publicly traded industry peers. The risk-free interest rates for periods within the expected life of the awards are based on the U.S. Treasury yield curve in effect during the period in which the awards were granted.

In addition, Ikaria was, and we will be, required to estimate forfeitures at the time of grant, and revise those estimates in subsequent periods if actual forfeitures differ from estimates. Ikaria used and we will also use historical data to estimate pre-vesting forfeitures and record stock-based compensation expense only for those awards that are expected to vest. To the extent that actual forfeitures differ from estimates, the difference is recorded as a cumulative adjustment in the period the estimates were revised. Stock-based compensation expense recognized in the financial statements is based on awards that are ultimately expected to vest.

For the periods presented, the weighted average grant date fair value of stock options granted to employees and directors of Ikaria and the weighted average assumptions used by Ikaria to estimate the grant date fair value of the options using the Black-Scholes-Merton option pricing model were:

	2013	2012
Weighted average grant date fair value	$ 1.95	$ 2.40
Valuation assumptions:		
Risk-free rate	0.90%	0.83%
Expected volatility	46.5%	47.6%
Expected term	5.00 yrs	5.00 yrs
Dividend yield	—	—

Ikaria has historically granted its stock options at exercise prices not less than the fair value of its common stock. Ikaria was a private company with no active public market for its common stock. Therefore, its board of directors periodically determined for financial reporting purposes the estimated fair value of its common stock using valuations performed in accordance with the guidance outlined in the AICPA Practice Aid, *Valuation of Privately Held Company Equity Securities Issued as Compensation*.

The compensation expense for the restricted stock units is based on the grant date fair value of the restricted stock unit, which was based on the fair value of the underlying stock.

The total stock-based compensation allocated to us by Ikaria for the years ended December 31, 2013 and 2012, was approximately $1.7 million and $1.5 million, respectively. Because these amounts relate to Ikaria stock-based awards, the amounts presented are not necessarily indicative of our future performance and do not necessarily reflect the stock-based compensation or compensation expense that we would have experienced as a stand-alone company for these periods.

In October 2011, Ikaria approved a special dividend plan, which provided for dividend equivalent rights for options, restricted stock units and other equity awards granted under its equity award plans. Pursuant to the special dividend plan, in the event that the Ikaria board declared a dividend, each employee of Ikaria who held equity awards was eligible to receive a cash payment equal to the amount of the dividend per share, multiplied by the number of equity awards outstanding. The payment was payable as of the declaration date for vested options. For unvested options and unvested restricted stock units, payment was due upon vesting. As of December 31, 2013, the allocated portion of the special dividend bonus payable was $6.1 million, of which $1.8 million was reflected in other current liabilities and $4.3 million was reflected in non-current liabilities. As of December 31, 2012, the allocated portion of the special dividend bonus payable was $3.6 million, of which $0.7 million was reflected in other current liabilities and $2.9 million was reflected in non-current liabilities.

Income Taxes

During the periods presented, we did not file separate tax returns, as we were included in the tax groupings of other Ikaria entities within the respective entity's tax jurisdiction. As such, the income tax provision included in our financial statements has been calculated using the separate return method, as if we filed a separate tax return in each of our respective tax jurisdictions.

For financial reporting purposes, we have historically recorded no tax expense or benefit due to our operating loss position. A valuation allowance has been established on net deferred tax assets because management believes that it is more likely than not that our net deferred tax assets will not be realized.

Deferred tax assets and liabilities are recognized for the estimated future tax consequences attributable to differences between the financial statement carrying amounts of existing assets and liabilities and their respective tax bases. Deferred tax assets and liabilities are measured using enacted

tax rates in effect for the year in which those temporary differences are expected to be recovered or settled.

We recognize the benefit of an uncertain tax position that we have taken or expect to take on income tax returns prepared under a separate return method if such tax position is more likely than not to be sustained on examination by the taxing authorities, based on the technical merits of the position. These tax benefits are measured based on the largest benefit that has a greater than 50% likelihood of being realized upon ultimate resolution. Unrecognized tax benefits related to net operating loss carryforwards or tax credit carryforwards are presented as a reduction to the related gross deferred tax asset. Unrecognized tax benefits for which a net operating loss carryforward or tax credit carryforward is not available are presented as a liability. A liability for unrecognized tax benefits is classified as non-current unless the liability is expected to be settled in cash within 12 months of the reporting date.

Certain deferred tax assets that arose as a result of Ikaria's past activities and resultant operating losses, such as federal and state net operating loss carryforwards, research and development credit carryforwards and acquired in-process research and development, do not constitute our assets and will continue to reside with Ikaria subsequent to the date of the Spin-Out.

Recently Adopted Accounting Standards

From time to time, new pronouncements are issued by the FASB or other standard setting bodies that may have an impact on our accounting and reporting. We believe that such recently issued accounting pronouncements and other authoritative guidance for which the effective date is in the future either will not have an impact on our accounting or reporting or that such impact will not be material to our financial position, results of operations and cash flows when implemented.

Quantitative and Qualitative Disclosure About Market Risk

We are exposed to market risk related to changes in interest rates. As of March 31, 2014, we had cash, cash equivalents and restricted cash of approximately $76.4 million, consisting primarily of demand deposits with U.S. banking institutions. Our primary exposure to market risk is interest rate sensitivity, which is affected by changes in the general level of U.S. interest rates, particularly because our investments are in cash and cash equivalents. Due to the short-term duration of our deposits and the low risk profile of our investments, an immediate 10% change in interest rates would not have a material effect on the fair market value of our deposits.

JOBS Act

We qualify as an "emerging growth company" as defined in the Jumpstart Our Business Startups Act of 2012, or the JOBS Act. As an emerging growth company, we may take advantage of specified reduced disclosure and other requirements that are otherwise applicable generally to public companies. These provisions include:

- only two years of audited financial statements, in addition to any required unaudited interim financial statements, with correspondingly reduced "Management's Discussion and Analysis of Financial Condition and Results of Operations" disclosure;
- reduced disclosure about our executive compensation arrangements;
- exemption from the non-binding advisory votes on executive compensation, including golden parachute arrangements; and
- exemption from the auditor attestation requirement in the assessment of our internal controls over financial reporting.

Generally, we may take advantage of these exemptions for up to five years or such earlier time that we are no longer an emerging growth company. We would cease to be an emerging growth company if we have more than $1.0 billion in annual revenue, we have more than $700.0 million in market value of our stock held by non-affiliates or we issue more than $1.0 billion of non-convertible debt over a three-year period. We may choose to take advantage of some, but not all, of the available exemptions. We have taken advantage of certain reduced reporting burdens in this prospectus. Accordingly, the information contained herein may be different than the information you receive from other public companies in which you hold stock.

In addition, the JOBS Act provides that an emerging growth company can take advantage of an extended transition period for complying with new or revised accounting standards. This allows an emerging growth company to delay the adoption of certain accounting standards until those standards would otherwise apply to private companies. We have irrevocably elected not to avail ourselves of this exemption from new or revised accounting standards and, therefore, we will be subject to the same new or revised accounting standards as other public companies that are not emerging growth companies.

BUSINESS

Overview

We are a clinical stage biotherapeutics company focused on developing innovative products at the intersection of drugs and devices that address significant unmet medical needs in the treatment of cardiopulmonary and cardiac diseases. We are developing three product candidates. Two of our product candidates are based on our proprietary pulsatile nitric oxide delivery device, which we refer to as INOpulse, and are in Phase 2 clinical trials—one for the treatment of pulmonary arterial hypertension, or PAH, and a second for the treatment of pulmonary hypertension associated with chronic obstructive pulmonary disease, or PH-COPD. Our third product candidate, bioabsorbable cardiac matrix, or BCM, is currently in a feasibility clinical trial (which is a CE mark registration trial in the European Union and is comparable to a Phase 2 clinical trial in U.S. drug development). We are developing BCM for the prevention of cardiac remodeling and subsequent congestive heart failure following acute myocardial infarction, or AMI, commonly known as a heart attack.

Our Product Candidates

Our product candidates are summarized in the table below. We have worldwide commercialization rights to our three product candidates.

Product Candidate	Primary Indication	Phase 1 / Pilot	Phase 2 / Feasibility	Phase 3 / Pivotal	Upcoming Milestone
INOpulse for PAH (IK-7001)	Improvement in functional capacity for patients with PAH				Trial completion expected by end of 2014
INOpulse for PH-COPD (IK-7002)	Reduction in mortality and morbidity in patients with PH-COPD on long-term oxygen therapy				Trial completion expected in mid-2014
BCM (IK-5001)	Prevention of cardiac remodeling and subsequent congestive heart failure following AMI				Trial completion expected in mid-2015

INOpulse

INOpulse is an extension of the technology that is used in hospitals to deliver continuous-flow inhaled nitric oxide. Use of inhaled nitric oxide is approved by the U.S. Food and Drug Administration, or the FDA, and certain other regulatory authorities to treat persistent pulmonary hypertension of the newborn. Ikaria, Inc., or Ikaria, has marketed continuous-flow inhaled nitric oxide as INOmax for this indication since approval in 1999. In October 2013, Ikaria transferred the INOpulse program to us with exclusive worldwide rights to develop and commercialize INOpulse in PAH, PH-COPD and pulmonary hypertension associated with idiopathic pulmonary fibrosis, or PH-IPF, with no royalty obligations. Our INOpulse program is built on scientific and technical expertise developed for the therapeutic delivery of inhaled nitric oxide. From the inception of our business through December 31, 2013, $176.5 million was invested in the development of our product candidates, all of which was funded by Ikaria. In 2010, we filed an investigational new drug application, or IND, for INOpulse for the treatment of patients with PAH, which is a form of pulmonary hypertension that is closely related to persistent pulmonary hypertension of the newborn. In 2012, we filed a second IND for INOpulse for the treatment of patients with PH-COPD. These IND programs were included in the assets that were transferred to us by Ikaria.

Nitric oxide is naturally produced and released by portions of the blood vessels and results in smooth muscle relaxation. In particular, nitric oxide controls muscle tone in blood vessels and thus is an important factor in regulating blood pressure. As the muscles of the blood vessels relax, blood flow

increases, helping the heart to deliver more blood to the body. When administered by inhalation, the action of nitric oxide has minimal effects on blood pressure outside of the lungs, an important safety consideration.

A limitation to the chronic use of inhaled nitric oxide is the lack of a safe and compact delivery system for outpatient use. We have designed INOpulse to be portable for use by ambulatory patients on a daily basis inside or outside their homes. INOpulse is designed to automatically adjust based on a patient's breathing pattern to deliver a constant and appropriate dose of the inhaled nitric oxide over time, independent of the patient's activity level, thus ensuring more consistent dosing in the alveoli of the lungs. In addition, our proprietary triple-lumen nasal cannula enables more accurate delivery of the dose to the patient with minimal infiltration of oxygen, which can have an undesirable reaction with inhaled nitric oxide. INOpulse is also compatible with many long-term oxygen therapy systems.

The ongoing INOpulse clinical trials are utilizing the first generation INOpulse DS device. We expect our future trials will use the next generation INOpulse Mark2 device, or the Mark2, which has approximately the same dimensions as a paperback book and weighs less than 2.5 pounds. The Mark2 has a simple user interface and a battery life of approximately 24 hours, which can be readily recharged in four hours, typically while the patient sleeps. The Mark2 has been well received by patients in the usability research we have conducted.

We have been issued patents with respect to the pulsed delivery of nitric oxide to ensure a consistent dose over time that expire as late as 2027 in the United States and as late as 2026 in certain other countries. We have also filed several patent applications for certain of the innovations included in the Mark2, and certain of the resulting patents, if issued, will expire in 2033.

INOpulse for PAH

We are developing our lead product candidate, INOpulse for the treatment of PAH, to address a significant and unmet medical need in an orphan disease. This product candidate represents a potential first-in-class therapy. PAH is characterized by abnormal constriction of the arteries in the lung, which increases the blood pressure in the lungs and results in abnormal strain on the heart's right ventricle, eventually leading to heart failure. If left untreated, primary PAH patients have a median survival of less than three years. While prevalence data varies widely, we estimate there are at least 35,000 patients currently treated for PAH in the United States and European Union. Moreover, because PAH is rare and causes varied symptoms, we believe there is significant under-diagnosis of the condition. There are several approved therapies for PAH, and we estimate, based on public product sale data, that, 2012 combined global sales were over $4.0 billion. Despite treatment of PAH patients with these therapies, PAH continues to be a life-threatening, progressive disorder with estimates of median survival ranging from three to five years.

We commenced a randomized, placebo-controlled Phase 2 clinical trial of INOpulse for PAH in April 2012. We plan to enroll at least 78 patients in this trial at 52 clinical sites in the United States and Canada. We expect to complete this trial by the end of 2014. After consultation with appropriate regulatory authorities, we plan to initiate a pivotal Phase 3 clinical trial program in the second half of 2015. The FDA has granted orphan drug designation to nitric oxide for the treatment of PAH.

INOpulse for PH-COPD

We are developing a second product candidate, INOpulse for the treatment of PH-COPD. COPD is a disease characterized by progressive and persistent airflow limitations. Patients with more severe COPD frequently have hypoxemia and are treated with long-term oxygen therapy. Despite treatment with oxygen, hypoxemia can progress and cause pulmonary hypertension. We estimate that there are approximately 700,000 PH-COPD patients in the United States alone. PH-COPD patients have a lower median life expectancy and a higher rate of hospitalization than COPD patients with similar respiratory

disease but without pulmonary hypertension. Currently, the generally accepted treatments for PH-COPD are oxygen therapy, pulmonary rehabilitation and lung transplant.

We commenced a randomized, placebo-controlled, dose-confirmation Phase 2 clinical trial of INOpulse for PH-COPD in December 2012. We plan to enroll at least 159 patients in this trial at 43 clinical sites in the United States. We expect to complete this trial by mid-2014. After completion of this clinical trial and additional discussions we expect to have with U.S. and EU regulatory authorities, we will assess our options for further development of INOpulse for PH-COPD, including potential partnerships.

BCM

We are developing a third product candidate, BCM, a medical device intended to prevent cardiac remodeling and impaired heart functioning following an AMI. Cardiac remodeling is a structural alteration of the heart that results in reduced heart function and often leads to congestive heart failure. BCM is delivered during a minimally invasive, commonly performed cardiac procedure called a percutaneous coronary intervention procedure. BCM is a formulated sterile solution of sodium alginate and calcium gluconate designed to be administered as a liquid through the coronary artery. When administered following an AMI, BCM flows into damaged heart muscle where, in the presence of abnormally high extracellular calcium released by the damaged cells, it forms a protective hydrogel meshwork within the heart wall. In our pre-clinical animal studies, as calcium levels in the damaged area returned to normal, BCM dissolved and was excreted through normal kidney function. Based on pre-clinical animal studies, it appears that BCM can act as a flexible scaffold to provide physical support to the ventricle wall in the early stages of recovery following an AMI and prevent further structural damage while the heart muscle heals. In a 27-patient pilot trial conducted by BioLineRx Ltd., BCM was well tolerated. As a Class III device, BCM is eligible for development through the premarket approval, or PMA, regulatory pathway in the United States. We have an exclusive worldwide license to BCM from BioLineRx Ltd. and its subsidiary, or BioLine, including with respect to issued composition of matter patents on BCM that expire as late as 2029 in the United States and 2024 in certain other countries. In the United States, the composition of matter patent may be extended to as late as 2032 to 2034, depending on the timing of marketing approval and other factors. A patent term extension may also be available in Israel and Japan.

Data from the American Heart Association and the European Association for Percutaneous Cardiovascular Interventions suggests that a total of approximately 2,000,000 patients suffer a heart attack in the United States and European Union each year, with at least 750,000 of these patients having a ST-segment elevated myocardial infarction, or STEMI. Of these STEMI patients, approximately 75% in the United States and a weighted average across France, Germany, Italy and the United Kingdom of over 50% currently undergo a percutaneous coronary intervention procedure and could be candidates for BCM if they are at risk for remodeling. We are testing BCM in STEMI patients who have a percutaneous coronary intervention, specifically in those patients who meet our inclusion criteria for high risk for remodeling.

We initiated a feasibility clinical trial of BCM in December 2011 and enrolled the first patient in April 2012. This trial is a CE mark registration trial in the European Union and is comparable to a Phase 2 trial in U.S. drug development. We plan to enroll approximately 300 patients in this trial at up to approximately 90 clinical sites in Europe, Australia, North America and Israel. We expect to complete this trial by mid-2015. If the results of this trial are positive, we expect it would form the basis for our application for CE marking in the European Union. We also plan to initiate a pivotal trial to support a PMA submission for regulatory approval in the United States.

Our Strategy

Our goal is to become a leader in developing and commercializing innovative products at the intersection of drugs and devices that address significant unmet medical needs in the treatment of cardiopulmonary and cardiac diseases. The key elements of our strategy to achieve this goal include:

- *Advance the clinical development of INOpulse.* Our primary focus for INOpulse is for use in treating PAH patients. Assuming positive results from our ongoing Phase 2 clinical trial in PAH, we intend to initiate a Phase 3 clinical trial program in the second half of 2015. For PH-COPD, assuming positive results from our ongoing Phase 2 trial, we plan to evaluate our options for further development, including potential partnerships.

- *Advance the clinical development of BCM in the prevention of cardiac remodeling following AMI.* Assuming positive results from our ongoing feasibility clinical trial, we expect to file for CE marking in the European Union in the second half of 2015 and to initiate a pivotal trial to support a PMA submission seeking marketing approval in the United States.

- *Leverage our historical core competencies to expand our pipeline.* We have years of institutional experience in the use of inhaled nitric oxide in treating pulmonary hypertension and in the development of drug-device combination product candidates. We expect to develop INOpulse for treatment of patients with PH-IPF. Our longer-term vision is to identify and opportunistically in-license innovative therapies that are at the intersection of drugs and devices and to develop and commercialize these product candidates.

- *Build commercial infrastructure in select markets.* As we near completion of the development of any product candidates, we expect to build a commercial infrastructure to enable us to market and sell our product candidates, if approved, using a specialty sales force in the United States. While we may partner with third parties to commercialize our product candidates in certain countries, we may also choose to establish commercialization capabilities in select countries outside the United States.

INOpulse Product Candidates

INOpulse Scientific Background

Nitric oxide is a naturally occurring molecule produced by many cells of the body. Researchers found that nitric oxide is produced and released by portions of the blood vessels and results in smooth muscle relaxation. In particular, nitric oxide controls muscle tone in blood vessels and thus is an important factor in regulating blood pressure. As the muscles of the blood vessels relax, blood flow increases, helping the heart to deliver more blood to the body. When administered by inhalation to patients with pulmonary hypertension, we expect inhaled nitric oxide to act in a similar manner to naturally produced nitric oxide. We believe the action of nitric oxide has minimal effects on blood pressure outside the lungs, an important safety concern.

The scientific journal Science named nitric oxide Molecule of the Year in 1992. Additionally, the three researchers who discovered the role of nitric oxide as a signaling molecule in the cardiovascular system earned the Nobel Prize for Physiology or Medicine in 1998.

In 1991, Dr. Warren Zapol and his associates at the Massachusetts General Hospital discovered that inhaling nitric oxide in gas form could reduce high blood pressure in the lungs, a condition known as pulmonary hypertension. Nitric oxide is a rapid and potent vasodilator, which means it quickly dilates, or widens, blood vessels. When inhaled, it quickly dilates blood vessels in the lungs, which reduces blood pressure in the lungs, strain on the right ventricle and shunting of de-oxygenated blood away from the lungs. Because more blood can flow through the lungs, blood levels of oxygen improve. In addition, inhaled nitric oxide improves the efficiency of oxygen delivery, and because it is a gas, it goes only to the portions of the lung that are ventilated, or receiving air flow, and increases blood flow only in these areas. Thus, inhaled nitric oxide improves ventilation-perfusion matching, an important

element of lung function involving the air that reaches the lungs, or ventilation, and the blood that reaches the lungs, or perfusion. Inhaled nitric oxide is quickly inactivated after contact with blood, and is selective for the lungs, meaning that it has minimal effects on blood pressure outside of the lungs, which is an important safety consideration.

Clinical trials supported the approval in 1999 of inhaled nitric oxide for the short-term treatment of persistent pulmonary hypertension of the newborn. Continuous-flow inhaled nitric oxide, which is administered to ventilated patients by a dedicated in-hospital device, is marketed by Ikaria and its commercialization partners worldwide as INOmax (INOflo in Japan).

INOpulse Drug Device Combination

INOpulse delivers brief, controlled pulses of nitric oxide, timed to occur at the beginning of each breath, that are inhaled by the patient through our proprietary triple-lumen nasal cannula. INOpulse is portable and therefore allows for treatment of ambulatory patients on a daily basis inside or outside their homes. INOpulse is designed to automatically adapt based on a patient's breathing pattern to deliver a constant dose of the drug over time, independent of the patient's activity level, thus ensuring predictable dosing in the alveoli of the lungs. We believe that INOpulse's targeted pulses of nitric oxide can deliver adequate drug to the muscles of the blood vessel, the site of action, using only 5% of the drug volume compared to continuous-flow delivery. In addition, the targeted pulsed delivery and built-in safety systems minimize the release of excess nitric oxide and its biproduct, nitrogen dioxide, into the patient's environment.

The INOpulse device is configured to be highly portable and compatible with available modes of long-term oxygen therapy via nasal cannula delivery. The ongoing clinical trials of INOpulse for PAH and PH-COPD are utilizing the first generation INOpulse DS device, which is derived from an older hospital-based system. While this device is portable and appropriate for use at home, to make INOpulse acceptable to a broader range of patients and to improve its usability, we are near completion of a next generation INOpulse Mark2 device, the Mark2. The Mark2 is approximately the size of a paperback book and weighs less than 2.5 pounds. It has a simple user interface and a battery life of approximately 24 hours, which can be readily recharged in four hours, typically while the patient sleeps. The Mark2 incorporates safety systems, proprietary software algorithms and our proprietary triple-lumen nasal cannula that allows the INOpulse device to deliver an accurate and consistent dose. Based on discussions with the FDA, we are required to show that the amount and timing of inhaled nitric oxide delivery is similar across INOpulse device generations. We have developed a regulatory bridging strategy to meet these requirements. We anticipate using the Mark2 in all of our future clinical trials.

The Mark2 is also designed to work with a broader range of oxygen therapy systems. We have filed several patent applications for certain innovations in our devices, and, if issued, certain of the resulting patents are expected to expire as late as 2033. The Mark2 has been well received by patients in the usability research we have conducted.

We are developing two similar, but importantly distinct, versions of our INOpulse device, one for use in patients with PAH and the other for use in patients with PH-COPD. Each device will be pre-programmed at the time of manufacturing to the dose setting specified for each indication. Since we expect that the dose for PAH will be as much as three-times higher than the dose for PH-COPD, the settings and the amount of drug used for each indication will differ substantially. In addition, the timing of dosing for the indication is different. The PH-COPD dose timing is targeted to a narrower window than for PAH. Further, since PAH patients have the potential for rebound pulmonary hypertension, which is a sudden and serious increase in pulmonary pressures that results from therapy withdrawal, patients with this condition are required to have a backup system. Accordingly, to ensure safety, we will be required to provide PAH patients with either a separate backup device or a device with a built-in pneumatic, or non-electrical, backup system. As part of our licensing terms with Ikaria,

we are required to lease and not to sell the device and also to maintain control and tracking of the indications for which each of the devices are used. We intend to meet these requirements by maintaining close monitoring of the use of the devices which is readily feasible through a planned remote data download and systems diagnostic feature. As a result of the differences between the two device versions and the required monitoring, patients will not be able to use either INOpulse device outside of the indication for which it is designed.

Introduction to Pulmonary Hypertension

Pulmonary hypertension is a disease characterized by constriction of the blood vessels in the lung, which causes blood pressure in the lung to rise and, in turn, increases the work required for the right ventricle of the heart to pump blood. The World Health Organization, or WHO, has endorsed a consensus classification that was updated most recently in 2013. The WHO classification has five broad pulmonary hypertension groups based on similarities in pathological and hemodynamic characteristics and therapeutic approaches. We are focusing development of INOpulse in Group 1 and Group 3 due to our view of the likelihood of success and the size and commercial viability of these markets. Group 1 pulmonary hypertension is called PAH and combines conditions with a range of causes, all of which have a characteristic pattern of vascular remodeling. PAH-specific therapy mainly targets patients in this Group. We expect that, because inhaled nitric oxide is a vasodilator, these patients will also benefit from INOpulse. Group 3 pulmonary hypertension consists of pulmonary hypertension associated with lung disease or hypoxemia, which is an abnormally low level of oxygen in the blood. This group includes patients with PH-COPD and PH-IPF, among others, and is the largest of the five pulmonary hypertension groups by prevalence.

INOpulse for Pulmonary Arterial Hypertension

We are developing our INOpulse for PAH product candidate to address a significant and unmet medical need in an orphan disease. This product candidate represents the development of a potential first-in-class therapy. Although current therapy for PAH provides some therapeutic benefit, there remains no cure, and approved therapies can have significant systemic side effects, such as hypotension and liver injury. INOpulse is designed to be a selective, short-acting pulmonary vasodilator and to act either additively or synergistically with most PAH medications to augment clinical benefit without adding significant risk to the adverse event profile of such medications.

Disease Background and Market Opportunity

PAH is a life-threatening, progressive disorder characterized by abnormally high blood pressure, or hypertension, in the pulmonary artery, the blood vessel that carries blood from the heart to the lungs. PAH occurs when most of the very small arteries, or arterioles, throughout the lungs narrow in diameter, which increases the resistance to blood flow through the lungs. To overcome the increased resistance, pressure increases in the pulmonary artery and the right ventricle, which is the heart chamber that pumps blood into the pulmonary artery. In addition, PAH may cause changes to the blood vessel lining that hinders the natural production of nitric oxide. Signs and symptoms of PAH occur when this increased pressure in the right ventricle cannot fully overcome the elevated resistance. The most common symptoms of PAH are shortness of breath during exertion and fainting spells. People with PAH may experience additional symptoms, particularly as the condition worsens, including dizziness, swelling of the ankles or legs, chest pain and a racing pulse.

If left untreated, primary PAH patients have an average survival of less than three years. There are a number of drugs approved for the treatment of PAH that work primarily by reducing pulmonary vascular resistance, which is the primary problem for these patients. However, despite treatment with these therapies, the mortality rate for PAH remains high, with estimates of median survival ranging from three to five years. Patients with PAH also report severe impairment of health-related quality of life, including poor general and emotional health and impaired physical functioning. These impairments

to health-related quality of life are comparable and sometimes more severe than those reported in patients with severely debilitating conditions such as spinal cord injury.

Since PAH is an orphan condition with poor diagnosis rates, published prevalence estimates for PAH vary widely. Based on epidemiological studies and current treatment rates, we estimate that there are at least 35,000 patients currently treated for PAH in the United States and European Union. The average age of PAH patients at diagnosis is approximately 50 years, and approximately 80% of PAH patients are female. PAH is often diagnosed late in the disease progression with approximately 73% of these patients already having progressed to WHO functional Class III or IV at the time of diagnosis. PAH is characterized by abnormal constriction of the arteries in the lung. There are three main mechanistic pathways affected by PAH, and PAH patients are generally treated with one or more of the four major classes of approved medications, which are prostacyclin analogs, phosphodiesterase type-5 inhibitors, endothelin receptor antagonists and a soluble guanylate cyclase stimulator. Approximately 45% of PAH patients are treated with more than one class of medication at a given time. Since hypoxemia can be a problem in these patients, it is generally treated with long-term oxygen therapy in accordance with broadly supported treatment guidelines in the United States and European Union.

We are testing INOpulse for PAH as an add-on therapy for use in patients whose disease is progressing and who use additional medications. If it is approved, INOpulse will be used in three partially overlapping treatment paradigms. The first would consist of patients with progressive disease who are on or considering treatment with prostacyclin or prostacyclin analogs. The second treatment paradigm would consist of patients who are being treated with oxygen therapy. Data from a U.S. and a French registry indicates that approximately 40% of patients are treated with oxygen at diagnosis for hypoxemia. While these patients may not be treated with INOpulse at diagnosis, we believe that their familiarity with and use of oxygen devices makes it more likely they will adopt INOpulse when prescribed. Data from our current trial supports our belief that these two groups of patients are likely users of INOpulse, as approximately 60% of the patients in the trial are on oxygen therapy and many others are treated or would be treated with prostacyclin or prostacyclin analogs. We believe that if INOpulse is shown to have clinical benefit when used only at night, a potential third paradigm would consist of use as an add-on therapy for PAH patient while they sleep.

A 2013 report by CVS Caremark Specialty Analytics provided examples of PAH medications with annual prices ranging from $96,000 to $162,000 per year in the United States. We expect that, if approved, the price of INOpulse will be in the range of other established PAH medications.

Scientific Rationale for Use of INOpulse for PAH

Since the discovery of the significant role of nitric oxide in vasodilation, there has been an expectation in the scientific community that inhaled nitric oxide could be an effective therapy for PAH. According to the Cleveland Clinic Center for Continuing Education section on Pulmonary Hypertension, exogenous administration of nitric oxide by inhalation is probably the most effective and specific therapy for PAH, but cost and technical complexity of delivering inhaled nitric oxide have limited its use to the hospital. Although not approved for the treatment of PAH, data from an in-hospital survey conducted by Ikaria showed an estimated 1,000 to 2,000 INOmax uses in PAH patients in the United States each year, indicating that physicians already use nitric oxide in some PAH patients. The difficulty in delivering inhaled nitric oxide outside of the hospital results from the size of the device and cylinder and the need for a specialized delivery system with built-in safety systems.

We are developing nitric oxide for treatment of PAH because nitric oxide is a proven vasodilator, and PAH is primarily a disease of high pulmonary vascular resistance. PAH is associated with impaired release of nitric oxide and thus we believe chronic administration of inhaled nitric oxide may be viewed as an adjunctive or replacement therapy in patients with PAH. The use of inhaled nitric oxide in PAH has been proposed since the role of nitric oxide in this disease was identified. This drug has been tested in limited investigational studies conducted at academic institutions, and published data from these studies indicate that administration of inhaled nitric oxide, in combination with other therapies, to patients with PAH decreases pulmonary vascular resistance, thereby reducing pulmonary pressure and improving clinical outcome with minimal systemic side effects.

When nitric oxide is delivered as a pulse at the beginning of inhalation, it travels to the alveoli where it diffuses rapidly across the alveolar capillary membrane into the adjacent smooth muscle of pulmonary vessels. This transport is similar to the natural transport of endogenous nitric oxide from the endothelial cells where it is produced to the smooth muscle cells where it relaxes the muscle and causes vasodilation of the pulmonary arteries.

We do not expect INOpulse to have systemic effects beyond the pulmonary vasculature. We believe this lack of systemic effects, combined with targeted delivery to the alveoli and the short half-life of nitric oxide, make INOpulse unlikely to have intolerable side effects, such as systemic hypotension or drug-drug interactions. Based on the mechanism of action, the nature of PAH and treatment protocols, we are developing INOpulse as an add-on or adjunctive therapy in patients with advancing disease, where we believe it has the highest commercial potential.

Clinical Development Program

INOpulse for PAH is designated as a drug-device combination by the FDA and is being evaluated through the Division of Cardiovascular and Renal Products of the Center for Drug Evaluation and Research with consultation from the Center for Devices and Radiological Health. For our IND for PAH, which we submitted in 2010, we performed animal studies in rats and sheep and referenced the INOmax NDA for additional supportive non-clinical information. Our pre-clinical studies and the referenced Ikaria studies demonstrated that nitric oxide is well tolerated and had activity in reducing pulmonary vascular resistance in these animals. The FDA has agreed that the non-clinical package is complete and adequate for supporting development and registration of inhaled nitric oxide and our INOpulse combination product candidate. In the European Union, since there is no formal drug-device designation, we expect the drug to be evaluated by the European Medicines Agency, or the EMA, with a reference in the label to the device and cannula, which will require a separate CE mark from a Notified Body.

We commenced a randomized, placebo-controlled Phase 2 clinical trial of INOpulse for PAH in April 2012 to determine the safety, tolerability and efficacy of two different doses of INOpulse for PAH. We plan to enroll at least 78 patients in this trial at 52 sites in the United States and Canada. The primary endpoint in this trial is a change in pulmonary vascular resistance at 16 weeks from baseline. Secondary endpoints include change in mean pulmonary arterial pressure and cardiac index as well as change in six-minute walk distance. We expect to complete this trial by the end of 2014. However, participants have the option to continue in Part B of the trial during which all participants will be exposed to inhaled nitric oxide either at low or high dose. Part B of the trial is expected to continue until approval of INOpulse for PAH or until clinical development of INOpulse for PAH is discontinued. Assuming successful results from this Phase 2 clinical trial, we plan to initiate a Phase 3 clinical trial program in the second half of 2015 after consultation with appropriate regulatory authorities. The design of the Phase 3 clinical program will depend on results from the ongoing trial and will be influenced by regulatory agency requirements relating to the size of the safety database and required statistical significance level. We expect to pursue approval of INOpulse for PAH as a selective pulmonary vasodilator for the chronic outpatient treatment of adults and children with WHO Group 1 PAH, and that the primary endpoint for any pivotal trial would be six-minute walk distance, which has been the standard approval endpoint for most PAH therapies.

INOpulse for PH-COPD

We are developing our INOpulse for PH-COPD product candidate to address a significant unmet medical need that is often overlooked in everyday clinical practice because of the lack of available therapy. Current therapies for COPD are targeted to relieve the symptoms and complications of the respiratory component of the disease. Unlike these therapies, INOpulse is directed at treating the

cardiovascular complications of PH-COPD. We believe PH-COPD patients on long-term oxygen therapy could use INOpulse in conjunction with their existing treatments.

Disease Background and Market Opportunity

COPD is a progressive disease caused by chronic inflammation and destruction of the airways and lung tissue. While COPD is primarily a respiratory disease, over time as the disease progresses, the chronic pulmonary restrictions and resulting deprivation of adequate oxygen, or hypoxia, can cause vasoconstriction in the pulmonary arterial bed. In addition, COPD patients can have deficiency in endogenous nitric oxide production in their lungs, which can worsen vasoconstriction. This pulmonary vasoconstriction puts pressure on the right side of the heart, making it less able to cope with stressors and potentially leading to progressive cardiac dilation, heart failure and death. This cardiovascular component of COPD is often overlooked despite pulmonologists' general awareness of the problem, in part because specific therapeutic modalities remain unavailable. While it is widely believed that the cardiovascular complications of COPD occur only in the advanced stage of the disease as a consequence of chronic hypoxemia, recent findings demonstrate an earlier involvement of the cardiovascular system in this disease.

According to a 2010 report by Data Monitor, over 1.4 million COPD patients in the United States were treated with long-term oxygen therapy. Based on academic studies, we estimate that 50% of COPD patients on long-term oxygen therapy in the United States have PH-COPD. Even though the degree of pulmonary hypertension in these patients is milder than in PAH patients, data published in literature suggests that even small elevations in mean pulmonary artery pressure in patients with advanced COPD can impact hospitalization, patient-assessed functional outcomes and mortality. Pulmonary hypertension is a well-known predictor of increased morbidity and mortality in COPD patients and is associated with poor quality of life, worse clinical outcomes and shorter survival time. PH-COPD patients had a four-year survival rate of approximately 50%. By contrast, COPD patients with similar pulmonary functions but without pulmonary hypertension had a four-year survival rate of 80%.

We expect INOpulse for PH-COPD, if approved, will be a treated as a specialty drug. Specialty drugs are typically high-cost medications, often ranging in price in the United States from approximately $15,000 to $50,000 per year, used to treat rare or complex conditions, requiring close clinical management and special handling and distributed through specialty pharmacies.

Scientific Rationale for Use of INOpulse for PH-COPD

The mechanism of action of inhaled nitric oxide in vasodilation at the alveolar smooth muscle in PH-COPD is similar to its action in PAH. Like endogenous pulmonary nitric oxide, inhaled nitric oxide works by selectively relaxing lung vascular smooth muscles, causing dilation of pulmonary blood vessels and consequently increased pulmonary blood flow. This reduces the elevated pulmonary artery pressure in patients with PH-COPD.

Pulsed inhaled nitric oxide is the only vasodilatory therapy for pulmonary hypertension that does not cause a fall in blood oxygen levels in COPD patients, as compared to other PAH drugs, which can worsen hypoxemia. This worsening of hypoxemia occurs because systemic agents for pulmonary hypertension, which are not targeted to specific ventilated alveoli, can cause indiscriminate pulmonary vasodilation, thereby lowering the average blood oxygenation levels. We believe that inhaled nitric oxide, as a locally active selective pulmonary vasodilator with minimal systemic effects, can drop pulmonary pressures, and when delivered with INOpulse as a targeted pulse to the well-ventilated alveoli, avoid this indiscriminate vasodilation and the consequent lowering of blood oxygen levels.

The targeted delivery of inhaled nitric oxide to specific alveoli is important because early trials with continuous-flow inhaled nitric oxide reduced pulmonary artery pressure but also resulted in

lowering of blood oxygen levels. It was postulated that this unwanted effect might be avoided by administering nitric oxide as a brief pulse at the beginning of each breath because well-ventilated alveoli open faster, and a brief early pulse would only reach these alveoli. As early as 1997, this concept was demonstrated by testing inhaled nitric oxide in PH-COPD patients during exercise, which allowed the dose to mimic pulse dosing. Recently, in a computational fluid-flow modeling study we conducted using high resolution computed tomography scans and computer simulations, data indicated that early pulsed delivery of nitric oxide could be directed specifically to the well-ventilated alveoli. Additional support for this hypothesis came from a small, open-label three-month trial, the results of which have been published, conducted at the University of Vienna in Austria, in which pulsed inhaled nitric oxide with long-term oxygen therapy reduced pulmonary pressures, lowered peripheral vascular resistance and increased cardiac output with no impact on blood oxygen levels in PH-COPD patients. Although the trial was conducted with a rudimentary pulsing device, this data indicates that pulsed delivery of inhaled nitric oxide was well-tolerated and showed a significant effect in lowering pulmonary pressures.

Clinical Development Program

INOpulse for PH-COPD is designated as a drug-device combination by the FDA and is being evaluated through the Division of Cardiovascular and Renal Products of the Center for Drug Evaluation and Research with consultation from the Division of Pulmonary, Allergy, and Rheumatology Products and the Center for Devices and Radiological Health. In our IND application for PH-COPD, we referenced the nonclinical information in our IND application for PAH. Additionally, we have performed computational modeling studies of pulsed nitric oxide delivery to the lung using the criteria established by the FDA in a recent draft guidance. The FDA has agreed that the non-clinical package is complete and adequate for supporting development and registration of inhaled nitric oxide and INOpulse combination product. In the European Union, since there is no formal drug-device designation, we expect the drug to be evaluated by the EMA with a reference in the label to the device and cannula, which will require a separate CE mark from a Notified Body.

We commenced a randomized, placebo-controlled, dose-confirmation Phase 2 clinical trial of INOpulse for PH-COPD. We plan to enroll at least 159 patients in this trial, testing doses between three and 75 microgram per kilogram ideal body weight per hour versus placebo, with a primary endpoint of change in pulmonary artery systolic pressure. Each patient in the trial is treated with one of five different doses of nitric oxide, or receives placebo, for approximately 20 minutes, during a single clinic visit, while being evaluated for cardiac changes using a non-invasive echocardiograph diagnostic test. We expect this trial to provide insight on any acute dose-dependent effects of inhaled nitric oxide in this patient population, which will inform our dose selection for the next phase of clinical development of INOpulse for PH-COPD. In addition, this trial is intended to validate an algorithm using non-invasive tests to identify at-risk patients that would allow us to avoid the need for invasive catheterization in these patients. We commenced this trial in December 2012 at 43 clinical sites in the United States. We expect to complete this trial by mid-2014.

Based on our interactions with the FDA, the primary endpoint to obtain approval has been defined as a substantial effect on the composite endpoint of all-cause mortality and all-cause hospitalizations. We believe, based on our market research, that proof of clinical benefit in these endpoints will be influential in driving physician behavior. While a clear association between the degree of pulmonary hypertension and mortality or morbidity in patients with COPD has been established, no proof is available that reduction of pulmonary artery pressure or pulmonary vascular resistance will lead to reduction in mortality or morbidity. To mitigate the risks inherent in this endpoint, we plan to use a staged approach to the development of INOpulse in this indication. This approach could include a trial of shorter duration and an adaptive design to validate the effect size of this therapy before we move into a Phase 3 pivotal trial. Given the risks and the size of the opportunity, after obtaining results

from the ongoing trial and additional discussions with U.S. and EU regulatory authorities, we will assess our options for further development of INOpulse for PH-COPD, including potentially partnering with other companies.

BCM for Prevention of Cardiac Remodeling following AMI

We are developing BCM through the medical device regulatory pathway to prevent congestive heart failure after AMI by preventing or reducing the structural alteration of damaged heart muscle caused by AMI, referred to as cardiac remodeling. Cardiac remodeling is a series of structural changes in the size and shape of the left ventricle after an AMI. These changes include thinning of the left ventricle wall at the infarction and the adjacent border zone, outward bulging of the infarcted region, hypertrophy of the non-infarcted portion of the left ventricle and dilation of the left ventricle chamber. Cardiac remodeling increases mechanical stresses on the left ventricular wall and reduces the efficiency of pumping blood leading to congestive heart failure.

BCM is intended to prevent cardiac remodeling by reducing the abnormal increase in ventricular wall stress and structural changes in the heart after an AMI. Once bloodflow has been re-established to the affected heart muscle of a patient following an AMI, a physician deploys BCM through the coronary artery related to the infarcted region of the left ventricle. BCM is designed to flow into the damaged heart muscle where it forms a flexible scaffold to enhance the mechanical strength of the heart muscle during recovery and repair, thereby preventing cardiac remodeling. We have an exclusive worldwide license to BCM under a license agreement we entered into with BioLine in August 2009.

Disease Background and Market Opportunity

An AMI is generally a sudden event resulting from a blockage of the one of the arteries supplying blood to the heart. This can cause the heart muscle to die or temporarily stop working. In some patients, particularly those with large areas of the heart affected by the AMI, the dead or stunned muscle in the infarcted area can start to degrade even if blood flow is subsequently restored.

Given recent advances in treating AMIs, patients do not typically die of the acute event, especially in developed countries with good hospital systems. Instead, post-AMI patients are at an increased risk of congestive heart failure that results from the loss of structural support where the tissue has died, leading to a change in the shape of the heart, or remodeling, excess blood being left in the heart after it beats and increased strain on the ventricular wall. These conditions can eventually cause the heart not to pump enough blood to the body, leading to congestive heart failure. A recent longitudinal study of the natural history of the disease showed that among patients 65 to 84 years old who suffered an AMI, nearly one-third developed congestive heart failure within three years. We are developing BCM to fill this unmet medical need by providing structural support of the heart muscle at a key point in the disease, with the goal of preventing cardiac remodeling and possibly progression to advanced stages of congestive heart failure.

According to hospital claims data and American Heart Association estimates, in 2014, the estimated incidence of AMI hospital admissions will be over 900,000 in the United States. There are two classifications of AMI, STEMI and non-STEMI. While both types of AMIs can cause significant damage to the heart, STEMIs tend to have more severe acute symptoms. We estimate that nearly one-third of AMI hospital admissions were for STEMI. Additionally, according to a published report, over one million people suffer from AMI and over half of whom suffer from STEMI annually in Europe. Patients who have had an AMI are at significant risk of cardiac remodeling and subsequent congestive heart failure, with approximately one-third of AMI patients progressing to congestive heart failure. In addition, the costs of treating the consequences of AMI can be substantial. The American Heart Association reported that the total cost of congestive heart failure in 2012 was approximately $30.0 billion in the United States, and we estimate that approximately 40% of these patients were treated for congestive heart failure following an AMI. Therefore, we believe a treatment that would help to prevent cardiac remodeling could generate significant medical cost savings in addition to improving the quality of life of these patients.

Scientific Rationale for Use of BCM in the Prevention of Cardiac Remodeling Following AMI

BCM is a clear, low-viscosity solution containing sodium alginate and calcium gluconate. Alginates, which are complex sugars obtained from seaweed, have been used extensively in the food industry as well as by the pharmaceutical and medical device industries. In medical devices, alginates have been used as wound dressings, as bone-void fillers and to create dental impressions. BCM's specific, patent-protected composition has been optimized to be partially cross-linked by calcium ions and to maintain a free-flowing liquid state for injection into the blood stream. However, when injected into the heart following an AMI when the blood vessels become leaky, this alginate flows into the damaged heart muscle where it comes into contact with the additional extra-cellular calcium that is released by the newly dead heart muscle cells, which causes additional cross-links within the alginate and turns it to gel meshwork with mechanical properties similar to the normal extracellular cardiac matrix. Based on data from animal studies, we believe these properties allow BCM to provide temporary structural support to the wall of the heart while it heals after an AMI.

Once deposited, BCM remains in the infarct zone for a few months. As the heart heals and the extracellular calcium levels return to normal, the crosslinks in the gel slowly degrade, and the alginate returns to liquid form and is excreted via the kidneys. In our pre-clinical animal studies of BCM, tissue sample analysis has shown that most of the alginate dissipates within three months and is no longer detectable in the heart or elsewhere in the body within six months after BCM injection. In an academic study published in the Journal of the American College of Cardiology, pigs were injected with either BCM or saline following an AMI. In this study, the pigs that received saline had approximately 44% greater enlargement in left ventricular chamber volume after 60 days compared to the pigs that received two milliliters of BCM. In another academic study conducted in dogs with AMI, deploying BCM at any time within one week of an AMI prevented cardiac remodeling compared to placebo.

Clinical Development Program

BCM is a Class III medical device that we are developing to prevent cardiac remodeling and subsequent congestive heart failure after AMI following successful re-opening of the blood vessels. The principal treatment for AMI is to re-establish blood flow in the blocked coronary artery at the earliest possible opportunity. This can be achieved by percutaneous coronary intervention, dissolving the blockage with medications or open heart surgery. BCM is designed to be deployed via a percutaneous coronary intervention into the previously blocked coronary artery after blood flow has been re-established.

BioLine completed a pilot clinical trial in Europe, in which BCM was safely administered to 27 patients within seven days following a moderate to large STEMI and percutaneous coronary intervention. This open-label trial was conducted in multiple centers in Germany and Belgium and included patients with a first AMI of substantial size. The primary purpose of this trial was to evaluate the safety of BCM deployment. In addition, some efficacy parameters could be observed as all patients suffered a STEMI and had serial echocardiography studies performed at one, three and six months. Based on historical comparison with the results of the study conducted by the REmodelage VEntriculaire study group, or the REVE study, we expected that patients in this pilot trial would have had an increase in left ventricular indices of cardiac remodeling similar to the worst quartile of patients in the REVE study. Instead, the echocardiograms of the patients showed preservation of left ventricular indices of cardiac remodeling similar to the lowest, or least sick, quartile in the REVE study.

We are developing BCM in the United States under an investigational device exemption, or IDE, and in consultation with a Notified Body in the European Union, which regulates the testing and use of devices. For our IDE application, we performed animal and in vitro studies and device effectiveness studies in pigs. Our pre-clinical studies demonstrated that BCM was well tolerated and showed activity

in reducing cardiac remodeling after AMI in pigs when deployed in either a dedicated percutaneous coronary intervention procedure or during an initial percutaneous coronary intervention procedure. The FDA has agreed that the non-clinical package is complete and adequate for supporting development and registration of BCM.

We are pursuing an indication for reduction or prevention of cardiac remodeling in which BCM is deployed during a dedicated second percutaneous coronary intervention procedure. We are conducting a clinical trial of BCM, which we refer to as PRESERVATION I. PRESERVATION I is a CE mark registration trial for EU regulatory purposes and a feasibility study (comparable to a Phase 2 trial in drug development) for U.S. regulatory purposes. This trial will enroll approximately 300 patients at up to approximately 90 sites in Europe, Australia, North America and Israel. In this double-blinded trial, subjects are randomized in a two-to-one ratio to BCM or placebo. The primary endpoint six months after device deployment is change in the anatomical measurement of left ventricular end-diastolic volume index by echocardiography. Secondary endpoints include the measurement of functional capacity of change in six-minute walk distance and the measurement of patient reported outcome as recorded on the quality of life tool of Kansas City Cardiomyopathy Questionnaire. In addition, as required by the trial protocol, we will follow all subjects to monitor safety for a period of five years after device deployment.

The FDA has granted us a conditional IDE for this study, which currently limits the number of patients we can enroll to 40 in the United States. Given the number of patients we expect to enroll in other countries, we believe we will complete this trial before we reach the limit. However, we are currently in discussions with the FDA to increase this limit in case we need additional patients in the United States. We expect to complete this trial by mid-2015. We also expect that if PRESERVATION I is successful, we will rely on the results to seek CE marking for BCM in the European Union.

Assuming positive results from PRESERVATION I, we plan to conduct a second, larger clinical trial to support approval in the United States through the PMA pathway. We met with the FDA to discuss U.S. regulatory requirements for a pivotal clinical trial. Based on discussions with the FDA Center for Devices and Radiological Health in May 2010, we expect that our pivotal trial will include approximately 1,000 patients, having a composite endpoint of anatomic measurements of left ventricular end-diastolic volume index, a patient outcomes measurement and a functional measure using either six-minute walk distance or a cardiopulmonary stress test.

We are testing BCM in patients with STEMIs since such patients tend to be more homogenous at presentation to the hospital, are often treated with a percutaneous coronary intervention and typically have a blockage responsible for AMI in only one vessel. Also, while there are several risk factors for congestive heart failure, one key measure is the size of the heart attack. Because we want to limit the number of patients required for our initial trial, we are initially testing BCM in an enriched, or sicker, population of patients with the largest STEMIs as identified by elevated cardiac enzymes and other parameters two to five days after the STEMI. Since our pre-clinical testing indicates that BCM acts equally whether given immediately after or within seven days of the AMI, we believe this trial design allows us to evaluate the BCM's effect in a more homogenous population with a likelihood of demonstrating clinical benefit. We expect that if this trial demonstrates that BCM is well tolerated and has a clinical benefit, we would investigate the safety of BCM injection during a primary percutaneous coronary intervention immediately after successful re-opening of the blood vessels, which would eliminate the need to administer BCM in a second, invasive procedure.

Relationship with Ikaria after the Spin-Out

The development of our product candidates was initiated under the leadership of our scientific and development team while at Ikaria. Ikaria's lead product, INOmax, is an inhaled nitric oxide product used for treatment of persistent pulmonary hypertension of the newborn. Our understanding of the

medical applications of nitric oxide and associated delivery devices, as well as our innovative approach to the pulsed delivery of nitric oxide, originated at Ikaria, and we in-licensed BCM while we were a part of Ikaria.

In October 2013, Ikaria completed an internal reorganization of certain assets and subsidiaries, in which it transferred to us exclusive worldwide rights, with no royalty obligations, to develop and commercialize INOpulse in PAH, PH-COPD and PH-IPF. Following the internal reorganization, in February 2014, Ikaria distributed all of our outstanding units to its stockholders through the payment of a special dividend to its stockholders on a pro rata basis based on each stockholder's pro rata ownership of Ikaria capital stock. We refer to Ikaria's distribution of our outstanding units to its stockholders as the Spin-Out. Shortly after the Spin-Out, Ikaria was acquired by entities affiliated with Madison Dearborn Partners. Ikaria retains the right to develop and commercialize inhaled nitric oxide products, including pulsed products, in all indications other than PAH, PH-COPD and PH-IPF.

In connection with the Spin-Out, we entered into several agreements with Ikaria providing for, among other things, the provision of transition services, the cross license of certain intellectual property, commitments not to compete, the manufacture and supply of the INOpulse drug and device and certain employee matters.

Transition Services Agreement

In February 2014, we entered into a transition services agreement with Ikaria, which we refer to as the TSA. Pursuant to the terms and conditions of the TSA, Ikaria has agreed to use commercially reasonable efforts to provide certain services to us, including human resources support, real estate support, information technology support, accounting and tax support, treasury support, financial planning and analysis support, purchasing support, management/executive services, legal services, quality services, regulatory services, drug and device safety services, business development support, biometrics support and manufacturing support. Ikaria is obligated, subject to the terms of the TSA (including the early termination provisions thereof and our obligation to use commercially reasonable efforts to provide the services for ourselves as soon as practicable), to provide such services until February 2016.

Ikaria has also agreed, on the terms and subject to the conditions of the TSA, to use commercially reasonable efforts to allow our employees to remain in Ikaria's Hampton, New Jersey facility for the continued operation of our business during the term of the TSA.

We are obligated to pay Ikaria a service fee in the amount of $772,000 per month and to reimburse Ikaria for any out-of-pocket expenses incurred in connection with its provisions of services under the TSA, any taxes imposed on Ikaria in connection with the performance or delivery of services under the TSA and any costs and expenses incurred by Ikaria in connection with the performance of any services that require resources outside of the existing resources of Ikaria or that otherwise interfere with the ordinary operations of Ikaria's business. This monthly service fee is be payable by us regardless of the frequency or quantity of services actually utilized by us under the TSA, and our obligation to pay such monthly service fee for 24 months will survive any early termination of the TSA. At the time we entered into the TSA, we also entered into an escrow agreement, pursuant to which we deposited $18.5 million, representing the aggregate amount of the monthly service fees payable by us under the TSA, into escrow to guarantee our payment of such fees to Ikaria. We are also obligated to pay any fees, costs, expenses or other amounts incurred by Ikaria to obtain the right to allow our employees to remain in the Hampton, New Jersey facility during the term of the TSA.

Exclusive Cross-License, Technology Transfer and Regulatory Matters Agreement

In February 2014, we entered into an exclusive cross-license, technology transfer and regulatory matters agreement with Ikaria. Pursuant to the terms of the license agreement, Ikaria granted to us a fully paid-up, non-royalty bearing, exclusive license under specified intellectual property rights controlled by Ikaria to engage in the development, manufacture and commercialization of nitric oxide, devices to deliver nitric oxide and related services for or in connection with out-patient, chronic treatment of patients with PAH, PH-COPD or PH-IPF, which we refer to collectively as the Bellerophon indications.

We have granted to Ikaria a fully paid-up, non-royalty-bearing, exclusive license under specified intellectual property rights that we control to engage in the development, manufacture and commercialization of products and services for or used in connection with the diagnosis, prevention or treatment, whether in- or out-patient, of certain conditions and diseases other than the Bellerophon indications specified in the license agreement and for the use of nitric oxide to treat or prevent conditions that are primarily managed in the hospital, which we refer to collectively as the Ikaria nitric oxide business.

We have agreed that, during the term of the license agreement, we will not, without the prior written consent of Ikaria, grant a sublicense under any of the intellectual property licensed to us under the license agreement to any of our affiliates or any third party, in either case that directly or indirectly competes with the Ikaria nitric oxide business. We have also agreed that we will include certain restrictions in our agreements with customers of our products to ensure that such products will only be used for the Bellerophon indications.

The license agreement will expire on a product-by-product basis for products for a specific Bellerophon indication at such time as we are no longer developing or commercializing any product for such indication. The license agreement may be terminated by either party in the event an act or order of a court or governmental authority prohibits either party from substantially performing under the license agreement. Either party may also terminate the license agreement in the event of an uncured material breach by the other party or in the event the other party is insolvent or in bankruptcy proceedings. Ikaria may also terminate the license agreement if we or any of our affiliates breach the agreements not to compete described below, or if we or any successor to our rights under the license agreement markets a generic nitric oxide product that is competitive with INOmax. Under certain circumstances, if the license agreement is terminated, the licenses granted to Ikaria by us will survive such termination.

Agreements Not to Compete

In September 2013, October 2013 and February 2014, we and each of our subsidiaries entered into an agreement not to compete with Ikaria. We refer to these agreements collectively as the agreements not to compete. Pursuant to the agreements not to compete, we and each of our subsidiaries agreed not to engage, anywhere in the world, in any manner, directly or indirectly, until the earlier of five years after the effective date of such agreement not to compete or the date on which Ikaria and all of its subsidiaries are no longer engaged in such business, in:

(1) the development, manufacture, commercialization, promotion, sale, import, export, servicing, repair, training, storage, distribution, transportation, licensing or other handling or disposition of any product or service (including, without limitation, any product or service that utilizes, contains or includes nitric oxide for inhalation, a device intended to deliver nitric oxide or a service that delivers or supports the delivery of nitric oxide), bundled or unbundled, for or used in connection with (a) the diagnosis, prevention or treatment, in both adult and/or pediatric populations, and whether in- or out-patient, of: (i) hypoxic respiratory failure associated with pulmonary hypertension, (ii) pulmonary hypertensive episodes and right heart failure associated with cardiovascular surgery, (iii) bronchopulmonary dysplasia, (iv) the management of ventilation-perfusion mismatch in acute lung injury, (v) the management of ventilation-perfusion mismatch in acute respiratory distress syndrome,

(vi) the management of pulmonary hypertension episodes and right heart failure in congestive heart failure, (vii) pulmonary edema from high altitude sickness, (viii) the management of pulmonary hypertension episodes and right heart failure in pulmonary or cardiac surgery, (ix) the management of pulmonary hypertension episodes and right heart failure in organ transplant, (x) sickle cell vaso-occlusive crisis, (xi) hypoxia associated with pneumonia or (xii) ischemia-reperfusion injury or (b) the use of nitric oxide to treat or prevent conditions that are primarily managed in the hospital; or

(2) any and all development, manufacture, commercialization, promotion, sale, import, export, storage, distribution, transportation, licensing, or other handling or disposition of any terlipressin or any other product within the pressin family, (a) intended to treat (i) hepatorenal syndrome in any form, (ii) bleeding esophageal varices, or (iii) septic shock, or (b) for or in connection with the management of low blood pressure.

The agreements not to compete expressly exclude the Bellerophon indications.

In February 2014, we also entered into drug and device clinical supply agreements and an employee matters agreement with Ikaria See "Manufacturing" below for a description of the drug and device clinical supply agreements and "Certain Relationships and Related Person Transactions" for a description of the employee matters agreement.

BioLine License Agreement

In August 2009, we entered into a license agreement with BioLineRx, Ltd. and BioLine Innovations Jerusalem L.P., under which we obtained an exclusive worldwide license to BCM. Under the license agreement, we are obligated to use commercially reasonable efforts to develop and commercialize at least one product containing BCM. We have established a joint development committee with BioLine to oversee the development of BCM.

We paid BioLine a $7.0 million upfront payment in 2009 and a $10.0 million milestone payment in 2010. Under the terms of the license agreement, if we achieve certain clinical and regulatory events specified in the license agreement, we will be obligated to pay milestone payments to BioLine that could total, in the aggregate, up to $115.5 million, and if we achieve certain commercialization targets specified in the license agreement, we will be obligated to pay additional milestone payments to BioLine that could total, in the aggregate, up to $150.0 million. In addition, we will be obligated to pay BioLine a specified percentage of any upfront consideration we receive for sublicensing BCM, as well as royalties at a percentage in the low double digits below 20% on net sales, if any, of any approved product containing BCM, subject to offsets for specified payments to third parties made in connection with BCM. Our obligation to pay BioLine royalties will expire on a product-by-product and country-by-country basis on the date on which BCM is no longer covered by a valid claim in the licensed patent rights in the given country.

BioLine has the option, exercisable under specified circumstances, to manufacture any product containing BCM for us pursuant to terms to be negotiated by the parties. If BioLine exercises this option, we would generally be obligated to purchase at least a specified percentage of our BCM requirements from BioLine at a price calculated using a pre-agreed methodology, and the parties would be required to establish a joint manufacturing committee to coordinate manufacturing efforts.

Except under specified circumstances, neither we, nor any other person that controls, is controlled by, or is under common control with us, may directly or indirectly acquire more than a specified percentage of the equity or debt securities of BioLine, or urge, induce, entice or solicit any other party to acquire such securities, without BioLine's consent.

We and BioLine have the right to terminate the license agreement for an uncured material breach by the other party. In addition, we have the right to terminate the license agreement if at any time we determine that further development of products containing BCM is not warranted.

BioLine has indicated to us that it believes that we have breached the license agreement in several ways, including, but not limited to, failure to use commercially reasonable efforts to develop BCM, failure to provide BioLine with material information concerning the development and commercialization plans

for BCM and failure to notify BioLine in advance of material public disclosures regarding BCM. We and BioLine also disagree about the timing of a $12.5 million milestone payment that we would owe BioLine based upon progress in our BCM clinical development program. We believe we have complied with our obligations under the license agreement to use commercially reasonable efforts to develop BCM and are not currently in breach of our other obligations under the license agreement. Although we have had multiple discussions with BioLine on these issues, and these discussions are continuing, we have not been able to resolve these outstanding issues. If we are unable to reach agreement with BioLine on these issues, BioLine could bring a lawsuit against us, although any claims relating to our alleged failure to use commercially reasonable efforts would first be subject to a non-binding 60-day mediation period with a third-party mediator. We believe that we would have strong defenses in any litigation that could be brought by BioLine. If BioLine were to prevail in any such litigation, one of the potential remedies would be the return of BCM to BioLine.

Manufacturing

Drug Products

In February 2014, we entered into a drug clinical supply agreement with Ikaria, or the drug supply agreement, pursuant to which Ikaria has agreed to use commercially reasonable efforts to manufacture and supply, and we have agreed to acquire from Ikaria, our requirements for nitric oxide for inhalation and corresponding placebo for use in our clinical programs for PH-COPD, PAH and PH-IPF. Pursuant to the drug supply agreement, we will pay Ikaria transfer pricing amounts equal to Ikaria's internal and external manufacturing cost plus 20%. Under the terms of the drug supply agreement, we have also granted Ikaria a right of first negotiation in the event that we desire to obtain supply of nitric oxide for inhalation and corresponding placebo (or any variant thereof or any version with different specifications) for commercial use. The drug supply agreement will expire on a product-by-product basis on the date we discontinue clinical development of such product. In addition, either party may terminate the drug supply agreement in the event of an uncured material breach by the other party.

Ikaria manufactures pharmaceutical-grade nitric oxide at its facility in Port Allen, Louisiana. This facility, which we believe is operated in compliance with current Good Manufacturing Practices, or cGMP, is the only FDA-inspected site for manufacturing pharmaceutical-grade nitric oxide in the world. The primary manufacturing activity at the site is the commercial production of INOmax and production of INOpulse. This production includes the chemical synthesis of high-purity nitric oxide, which is the active pharmaceutical ingredient in INOmax and INOpulse, and the filling of the gas cylinders in which the products are packaged. Ikaria has a back-up facility for cylinder filling in Coppell, Texas. In order to provide continuous drug supply under various adverse circumstances, we plan to establish the capability to fill gas cartridges for INOpulse at Ikaria's Coppell site, approximately at the time we commence Phase 3 clinical trials of INOpulse.

To support business outside of the United States, the Port Allen manufacturing facility has also successfully passed inspections by local agencies, the EMA, Health Canada; the Pharmaceutical and Medical Devices Agency, or PMDA, of Japan, and the Korean FDA, or KFDA. The EMA, the Health Protection Branch of Health Canada, PMDA and KFDA operate in a similar fashion to the FDA in that each requires submission of a dossier containing substantial evidence of safety and effectiveness prior to approval. These agencies' monitoring of safety in a post-marketing setting also is similar to that of the FDA.

The operations that Ikaria performs consist of two steps. The first step is to manufacture the concentrated drug product, which Ikaria conducts using the same processes that it uses to manufacture its own drug product. The second step is the filling operation in which the pre-mix product is mixed to the appropriate concentration and filled into the final cartridges that we use with INOpulse. As we have reduced the size and weight of INOpulse, we have also developed a smaller, more-concentrated drug cartridge for INOpulse. The filling process has been developed by Ikaria as a high-throughput batch fill process that leverages several technologies that Ikaria has developed, and we have licensed, to

fill smaller containers at a higher pressure and purity and at a significantly higher production rate than prior technology.

This manufacturing system is designed to be modular and can be expanded as needed. The current installed capacity within the Port Allen plant is sufficient to support our clinical program through the end of our planned Phase 3 clinical trials in INOpulse for PAH and COPD. In addition, the plant has the capacity to expand to meet additional demand. We have a license from Ikaria to use this fill process technology to work with additional companies, as needed, to produce the final cartridge. Commercial supply manufacturing can be supported with additional units installed at the Port Allen site or other regional locations, by Ikaria or other manufacturers, as determined by distribution requirements. For our clinical trials, Ikaria can supply and ship product from the Port Allen site and the current cartridges are expected to have a shelf life of at least one year. We are testing the finished product to potentially establish a shelf life of up to two years.

We currently outsource the manufacture of BCM for use in clinical trials. BCM is manufactured by our contract manufacturing organization under the terms of a manufacturing and supply agreement that expires in April 2017. The manufacturing and supply agreement will automatically renew every two years unless either party provides written notice of termination at least 24 months prior to expiration. BCM is composed of ultra-pure sodium alginate and calcium-D-gluconate. We purchase sodium alginate from FMC BioPolymer AS (doing business as NovaMatrix™) under the terms of a clinical supply agreement that expires in December 2018. We and FMC BioPolymer have agreed to negotiate a commercial supply agreement prior to the December 2018 expiration of the clinical supply agreement. Calcium-D-gluconate is a commodity item available from multiple suppliers. If BCM is approved for commercial sale, we will likely continue to outsource its manufacture to contract manufacturers.

Drug Delivery Systems

In February 2014, we entered into a device clinical supply agreement with Ikaria, or the device supply agreement, pursuant to which Ikaria will use commercially reasonable efforts to manufacture and supply our requirements for certain nitric oxide delivery devices specified in the device supply agreement for use in our clinical programs for PH-COPD and PAH. Pursuant to the device supply agreement, we will pay to Ikaria transfer pricing amounts equal to Ikaria's internal and external manufacturing cost plus 20%. The device supply agreement will expire on February 9, 2015. In addition, either party may terminate the device supply agreement in the event of an uncured material breach by the other party.

In 2008, Ikaria established a drug delivery system manufacturing facility in Madison, Wisconsin, which is responsible for the design, engineering, assembly, packaging and distribution of drug delivery systems, including INOpulse.

Competition

The biotechnology and pharmaceutical industries are highly competitive. There are many pharmaceutical companies, biotechnology companies, public and private universities and research organizations actively engaged in the research and development of products that may be similar to our products. In addition, other companies are increasingly looking at cardiac and cardiopulmonary indications as a potential opportunity. It is possible that the number of companies seeking to develop products and therapies for the treatment of unmet needs in our target markets will increase.

Our competitors, either alone or with their strategic partners, may have substantially greater financial, technical and human resources than we do and significantly greater experience in the discovery and development of product candidates, obtaining FDA and other regulatory approvals of products and the commercialization of those products. Accordingly, our competitors may be more successful than we may be in obtaining approval for therapies and achieving widespread market acceptance. We anticipate that we will face intense and increasing competition as new drugs and advanced technologies become available.

Currently, there are 12 drugs approved for the treatment of PAH, within the following categories: prostacyclin and prostacyclin analogs (including Flolan (epoprostenol), which is marketed by GlaxoSmithKline, Tyvaso (teprostinil), Orenitram (treprostinil) and Remodulin (teprostinil), which are marketed by United Therapeutics Corporation, and Ventavis (iloprost) and Veletri (epoprostenol), which are marketed by Actelion Pharmaceuticals, Inc., or Actelion), phosphodiesterase type-5 inhibitors (including Adcirca (tadalafil), which is marketed by United Therapeutics Corporation, and Revatio (sildenafil), which is marketed by Pfizer, Inc.), endothelin receptor antagonists (including Letairis (ambrisentan), which is marketed by Gilead Sciences, Inc., and Opsumit (macitentan) and Tracleer (bosentan), which are marketed by Actelion) and a soluble guanylate cyclase stimulator (Adempas (riociguat), which is marketed by Bayer Healthcare). One additional unapproved drug, Actelion's selexipag, is currently in Phase 3 clinical development. There are also other treatments in Phase 1 and Phase 2 clinical development, including other nitric oxide generation and delivery systems, including GeNO LLC's GeNOsyl.

For PH-COPD, there are no therapies other than long-term oxygen therapy and lung transplant, and we are not aware of any therapies for PH-COPD in advanced clinical development.

There are no generally accepted products approved for structural support to prevent cardiac remodeling following an AMI. Other product candidates that are currently in clinical development include stem cell therapies to restore heart muscle cells following an AMI, with large Phase 3 trials expected to end in 2018 or 2019. We do not expect BCM to compete with, or replace, current treatments for congestive heart failure following AMI, but instead believe it will become part of the treatment regimen used in conjunction with other therapies. In addition, because BCM can be delivered by a minimally invasive percutaneous coronary intervention procedure, we do not believe it will directly compete with devices that are used to treat congestive heart failure, some of which are currently in development, and designed for administration during open heart surgery or by intra-thoracic injection. These include mesh restraining devices, for example HeartNet; injectable biopolymers, for example Algisyl-LVR; and implantable electro-stimulation devices, for example, CardioFit. In addition, volume reduction surgery or cardiac assist devices, or pumps, are sometimes used to treat patients with congestive heart failure.

Patents and Proprietary Rights

We strive to protect the proprietary technologies that we believe are important to our business, including seeking and maintaining patent protection intended to protect, for example, our product candidates, related technologies and/or other aspects of the inventions that are important to our business. Our owned and licensed patents and patent applications cover patentable subject matter from composition of matter, methods of use, manufacturing processes for BCM and method of administration, devices and device components, critical safety features and design components with respect to INOpulse. However, patent protection is not available for the composition of matter of the active pharmaceutical ingredients in our INOpulse product candidates since nitric oxide is a naturally occurring molecule.

Actual protection afforded by a patent, which can vary from country to country, depends on the type of patent, the scope of its coverage and the availability of legal remedies in the country. We also rely on trade secrets and careful monitoring of our proprietary information to protect aspects of our business that are not amenable to, or that we do not consider appropriate for, patent protection.

We plan to continue to expand our intellectual property estate by filing patent applications directed to inventions which provide additional patent protection for our product offering, for instance, device enhancements, safety features and manufacturing processes. Our success will depend significantly on our ability to obtain and maintain patent and other proprietary protection for commercially important technology, inventions and know-how related to our business; defend and enforce our patents; maintain our licenses to use intellectual property owned by third parties; preserve the confidentiality of our trade secrets; and operate without infringing the valid and enforceable patents

and other proprietary rights of third parties. We also consider know-how, continuing technological innovation and in-licensing opportunities to develop, strengthen and maintain our proprietary positions.

A third party may hold intellectual property, including patent rights that are important or necessary to the development of our product candidates. It may be necessary for us to use the patented or proprietary technology of third parties to commercialize our product candidates, in which case we would be required to obtain a license from these third parties on commercially reasonable terms, or our business could be harmed, possibly materially. For example, if we want to expand the indications for which we could develop and commercialize INOpulse beyond PAH, PH-COPD and PH-IPF, we will need to obtain a license from Ikaria.

The patent positions of biotherapeutics companies like us are generally uncertain and involve complex legal, scientific and factual questions. In addition, the coverage claimed in a patent application can be significantly reduced before the patent is issued, and patent scope can be reinterpreted by the courts after issuance. Moreover, many jurisdictions permit third parties to challenge issued patents in administrative proceedings which may result in further narrowing or even cancellation of patent claims. Consequently, we do not know whether any of our product candidates will be protectable or remain protected by enforceable patents. We cannot predict whether the patent applications we are currently pursuing will issue as patents in any particular jurisdiction or whether the claims of any issued patents will provide sufficient protection from competitors. Any patents that we own or license may be challenged, narrowed, circumvented or invalidated by third parties.

Because patent applications in the United States and certain other jurisdictions are maintained in secrecy for 18 months or potentially even longer, and since publication of discoveries in the scientific or patent literature often lags behind actual discoveries, we cannot be certain of the priority of inventions covered by pending patent applications. Moreover, we may have to participate in interference proceedings declared by the U.S. Patent and Trademark Office, or USPTO, to determine priority of inventions for any patent applications filed with the USPTO on or before March 15, 2013. Likewise, derivation proceedings may also be declared for any patent filings filed after March 15, 2013.

The patents and patent applications that relate to our product candidates are described below.

INOpulse

We hold exclusive licenses from Ikaria to a broad portfolio of about a dozen families of issued and pending patent applications in both the United States and in certain other countries. Certain of these patent applications, if issued, will expire as late as 2033. These patent rights have been exclusively licensed for the treatment of patients with PAH, PH-COPD and PH-IPF and cover methods of delivery and the drug delivery device, as well as important safety features and the ornamental design of the drug delivery device.

Our primary basis for patent exclusivity is based on issued in-licensed patents directed to proprietary methods of administering pulsed inhaled nitric oxide, as well as a device for delivering the same. This patent family expires as late as 2027 in the United States and as late as 2026 in certain other countries.

We also hold a portfolio of additional in-licensed pending patent applications directed to novel nasal cannula features necessary for the safe and efficacious administration of pulsed nitric oxide. The patents issued based on these patent applications, if issued, will expire in 2033 in the United States and abroad.

Another in-licensed patent family relates to features of the drug delivery canister necessary for providing drug product for use with our proprietary pulsing drug delivery device. These patent applications, if issued, will expire in 2029. Several other patent families directed to device and safety features are pending as well as several ornamental design patent rights covering both the clinical and intended commercial device design.

In addition, the FDA has granted orphan drug designation for nitric oxide for the treatment of PAH, which could result in marketing exclusivity of seven years in the United States should this be the

first NDA approved for inhaled nitric oxide in this indication. The active ingredient, nitric oxide, was previously approved by the FDA as a drug in a separate clinical application. Accordingly, any related patent rights will not be eligible for a patent term extension under relevant provisions of the Hatch-Waxman Act.

BCM

Patent protection of BCM in the United States and elsewhere is provided by issued composition of matter and method of treatment patents, which we in-license from BioLine, that cover the intended commercial product. These issued patents are not limited to treatment of cardiac tissue, affording broad protection for the use of BCM in treating any damaged body tissue.

BCM will be regulated as a device and therefore data exclusivity will not be available. However, under the Hatch-Waxman Act, one issued U.S. patent covering the product will be eligible for patent term extension of up to five years to recover patent term lost during clinical trials. Accordingly, if the U.S. composition of matter patent that expires in 2029 is selected for this extension and a patent term extension is granted, certain rights under the patent may not expire until 2032 to 2034, depending on the timing of marketing approval and other factors. Corresponding issued patents may also be eligible for a patent term extension in certain other countries. We do not expect to get a patent term extension for composition of matter patents in Europe. These patents expire in May 2024. Patent term extensions may be available in other countries such as Japan and Israel.

Additional patents directed to methods of manufacturing, which we in-license, as well as patent applications relating to methods of manufacturing we developed and own, if issued, will expire, as late as 2032. Further, there is no abbreviated clinical trial pathway, such as an abbreviated new drug application, or ANDA, or a 505(b)(2) new drug application, for a device product approved via a PMA pathway.

Patent Term

The base term of a U.S. patent is 20 years from the filing date of the earliest-filed non-provisional patent application from which the patent claims priority. The term of a U.S. patent can be lengthened by patent term adjustment, which compensates the owner of the patent for administrative delays at the USPTO. In some cases, the term of a U.S. patent is shortened by terminal disclaimer that reduces its term to that of an earlier-expiring patent.

The term of a U.S. patent may be eligible for patent term extension under the Hatch-Waxman Act to account for at least some of the time the drug or device is under development and regulatory review after the patent is granted. With regard to a drug or device for which FDA approval is the first permitted marketing of the active ingredient, the Hatch-Waxman Act allows for extension of the term of one U.S. patent. Thus, patent term extension is not available for INOpulse since the active moiety is nitric oxide, which is already subject to an approved NDA. The extended patent term cannot exceed the shorter of five years beyond the non-extended expiration of the patent or 14 years from the date of the FDA approval of the drug or device. Some foreign jurisdictions have analogous patent term extension provisions that allow for extension of the term of a patent that covers a device approved by the applicable foreign regulatory agency. In the future, if and when BCM receives FDA approval, we expect to apply for a patent term extension on the patent covering BCM that we believe will provide the best exclusivity position if extended.

Trade Secrets

In addition to patents, we rely on trade secrets and know-how to develop and maintain our competitive position. We typically rely on trade secrets to protect aspects of our business that are not amenable to, or that we do not consider appropriate for, patent protection. For example, elements of the manufacture of our products are based on trade secrets and know-how that are not publicly disclosed. We protect trade secrets and know-how by establishing confidentiality agreements and invention assignment agreements with our employees, consultants, scientific advisors, contractors and

commercial partners. These agreements provide that all confidential information developed or made known during the course of an individual or entity's relationship with us must be kept confidential during and after the relationship. These agreements also provide that all inventions resulting from work performed for us or relating to our business and conceived or completed during the period of employment or assignment, as applicable, shall be our exclusive property. In addition, we take other appropriate precautions, such as physical and technological security measures, to guard against misappropriation of our proprietary technology by third parties.

Trademarks

We also seek trademark protection in the United States and in foreign jurisdictions where available and when appropriate. The symbol ™ indicates a common law trademark. Other service marks, trademarks and trade names appearing in this prospectus are the property of their respective owners.

Government Regulation

Government authorities in the United States, at the federal, state and local level, and in other countries and jurisdictions, including the European Union, extensively regulate, among other things, the research, development, testing, manufacture, quality control, clearance, approval, packaging, storage, recordkeeping, labeling, advertising, promotion, distribution, marketing, post-approval monitoring and reporting, and import and export of pharmaceutical products and medical devices. The processes for obtaining marketing approvals in the United States and in foreign countries and jurisdictions, along with subsequent compliance with applicable statutes and regulations and other regulatory authorities, require the expenditure of substantial time and financial resources.

Review and Approval of Drugs in the United States

In the United States, the FDA regulates drugs under the Food, Drug, and Cosmetic Act, or FDCA, and implementing regulations. The process of obtaining regulatory approvals and the subsequent compliance with appropriate federal, state, local and foreign statutes and regulations requires the expenditure of substantial time and financial resources. Failure to comply with the applicable U.S. requirements at any time during the product development process, approval process or after approval may subject an applicant and/or sponsor to a variety of administrative or judicial sanctions, including refusal by the FDA to approve pending applications, withdrawal of an approval, imposition of a clinical hold, issuance of warning letters and other types of letters, product recalls, product seizures, total or partial suspension of production or distribution, injunctions, fines, refusals of government contracts, restitution, disgorgement of profits, or civil or criminal investigations and penalties brought by the FDA and the Department of Justice or DOJ or other governmental entities.

Our product candidates must be approved by the FDA through the NDA process before they may be legally marketed in the United States. An applicant seeking approval to market and distribute a new drug product in the United States must typically undertake the following:

- completion of pre-clinical laboratory tests, animal studies and formulation studies in compliance with the FDA's good laboratory practice, or GLP, regulations;
- submission to the FDA of an IND, which must take effect before human clinical trials may begin;
- approval by an independent institutional review board, or IRB, representing each clinical site before each clinical trial may be initiated;
- performance of adequate and well-controlled human clinical trials in accordance with good clinical practices, or GCP, to establish the safety and efficacy of the proposed drug product for each indication;
- preparation and submission to the FDA of an NDA;
- review of the product by an FDA advisory committee, where appropriate or if applicable;

- satisfactory completion of one or more FDA inspections of the manufacturing facility or facilities at which the product, or components thereof, are produced to assess compliance with cGMP requirements and to assure that the facilities, methods and controls are adequate to preserve the product's identity, strength, quality and purity;

- payment of user fees and securing FDA approval of the NDA; and

- compliance with any post-approval requirements, including Risk Evaluation and Mitigation Strategies, or REMS, and post-approval studies required by the FDA.

Pre-Clinical Studies

Pre-clinical studies include laboratory evaluation of the purity and stability of the manufactured drug substance or active pharmaceutical ingredient and the formulated drug or drug product, as well as *in vitro* and animal studies to assess the safety and activity of the drug for initial testing in humans and to establish a rationale for therapeutic use. The conduct of pre-clinical studies is subject to federal regulations and requirements, including GLP regulations. The results of the pre-clinical tests, together with manufacturing information, analytical data, any available clinical data or literature and plans for clinical studies, among other things, are submitted to the FDA as part of an IND.

Companies usually must complete some long-term pre-clinical testing, such as animal tests of reproductive adverse events and carcinogenicity, and must also develop additional information about the chemistry and physical characteristics of the drug and finalize a process for manufacturing the drug in commercial quantities in accordance with cGMP requirements. The manufacturing process must be capable of consistently producing quality batches of the drug candidate and, among other things, the manufacturer must develop methods for testing the identity, strength, quality and purity of the final drug product. Additionally, appropriate packaging must be selected and tested and stability studies must be conducted to demonstrate that the drug candidate does not undergo unacceptable deterioration over its shelf life.

Human Clinical Studies in Support of an NDA

Clinical trials involve the administration of the investigational product to human subjects under the supervision of qualified investigators in accordance with GCP requirements, which include, among other things, the requirement that all research subjects provide their informed consent in writing before their participation in any clinical trial. Clinical trials are conducted under written study protocols detailing, among other things, the objectives of the study, the parameters to be used in monitoring safety and the effectiveness criteria to be evaluated. A protocol for each clinical trial and any subsequent protocol amendments must be submitted to the FDA as part of the IND. An IND automatically becomes effective 30 days after receipt by the FDA, unless before that time the FDA raises concerns or questions related to a proposed clinical trial and places the trial on clinical hold. In such a case, the IND sponsor and the FDA must resolve any outstanding concerns before the clinical trial can begin. Accordingly, submission of an IND may or may not result in the FDA allowing clinical trials to commence.

In addition, an IRB representing each institution participating in the clinical trial must review and approve the plan for any clinical trial before it commences at that institution, and the IRB must conduct continuing review and reapprove the study at least annually. The IRB must review and approve, among other things, the study protocol and informed consent information to be provided to study subjects. An IRB must operate in compliance with FDA regulations. Information about certain clinical trials must be submitted within specific timeframes to the National Institutes of Health for public dissemination on their ClinicalTrials.gov website.

A sponsor who wishes to conduct a clinical trial outside the United States may, but need not, obtain FDA authorization to conduct the clinical trial under an IND. If a foreign clinical trial is not conducted under an IND, the sponsor may submit data from the clinical trial to the FDA in support of an NDA or IND so long as the clinical trial is conducted in compliance with GCP, and the FDA is able to validate the data from the study through an onsite inspection if the agency deems it necessary.

Human clinical trials are typically conducted in three sequential phases, which may overlap or be combined:

- Phase 1: The drug is initially introduced into a small number of healthy human subjects or patients with the target disease (e.g. cancer) or condition and tested for safety, dosage tolerance, absorption, metabolism, distribution, excretion and, if possible, to gain an early indication of its effectiveness and to determine optimal dosage.

- Phase 2: The drug is administered to a limited patient population to identify possible adverse effects and safety risks, to preliminarily evaluate the efficacy of the product for specific targeted diseases and to determine dosage tolerance and optimal dosage.

- Phase 3: Phase 3 clinical trials are commonly referred to as "pivotal" studies, which typically denotes a study which presents the data that the FDA or other relevant regulatory agency will use to determine whether or not to approve a drug. In Phase 3 studies, the drug is administered to an expanded patient population, generally at geographically dispersed clinical trial sites, in well-controlled clinical trials to generate enough data to statistically evaluate the efficacy and safety of the product for approval, to establish the overall risk-benefit profile of the product, and to provide adequate information for the labeling of the product.

Progress reports detailing the results of the clinical trials must be submitted at least annually to the FDA and more frequently if serious adverse events occur. Phase 1, Phase 2 and Phase 3 clinical trials may not be completed successfully within any specified period, or at all. Furthermore, the FDA or the sponsor may suspend or terminate a clinical trial at any time on various grounds, including a finding that the research subjects are being exposed to an unacceptable health risk. Similarly, an IRB can suspend or terminate approval of a clinical trial at its institution, or an institution it represents, if the clinical trial is not being conducted in accordance with the IRB's requirements or if the drug has been associated with unexpected serious harm to patients. The FDA will typically inspect one or more clinical sites to assure compliance with GCP and the integrity of the clinical data submitted.

Section 505(b)(2) NDAs

NDAs for most new drug products are based on two full clinical studies which must contain substantial evidence of the safety and efficacy of the proposed new product. These applications are submitted under Section 505(b)(1) of the FDCA. The FDA is, however, authorized to approve an alternative type of NDA under Section 505(b)(2) of the FDCA. This type of application allows the applicant to rely, in part, on the FDA's previous findings of safety and efficacy for a similar product, or published literature. Specifically, Section 505(b)(2) applies to NDAs for a drug for which the investigations made to show whether or not the drug is safe for use and effective in use and relied upon by the applicant for approval of the application "were not conducted by or for the applicant and for which the applicant has not obtained a right of reference or use from the person by or for whom the investigations were conducted."

Thus, Section 505(b)(2) authorizes the FDA to approve an NDA based on safety and effectiveness data that were not developed by the applicant. NDAs filed under Section 505(b)(2) may provide an alternate and potentially more expeditious pathway to FDA approval for new or improved formulations or new uses of previously approved products. If the 505(b)(2) applicant can establish that reliance on the FDA's previous approval is scientifically appropriate, the applicant may eliminate the need to

conduct certain pre-clinical or clinical studies of the new product. The FDA may also require companies to perform additional studies or measurements to support the change from the approved product. The FDA may then approve the new drug candidate for all or some of the label indications for which the referenced product has been approved, as well as for any new indication sought by the Section 505(b)(2) applicant.

Submission of an NDA to the FDA

NDAs for most new drug products are based on two full clinical studies that must contain substantial evidence of the safety and efficacy of the proposed new product. Assuming successful completion of required clinical testing and other requirements, the results of the pre-clinical and clinical studies, together with detailed information relating to the product's chemistry, manufacture, controls and proposed labeling, among other things, are submitted to the FDA as part of an NDA requesting approval to market the drug product for one or more indications. Under federal law, the submission of most NDAs is additionally subject to an application user fee, currently exceeding $2.1 million, and the sponsor of an approved NDA is also subject to annual product and establishment user fees, currently exceeding $104,000 per product and $554,000 per establishment. These fees are typically increased annually.

The FDA conducts a preliminary review of an NDA within 60 days of its receipt and informs the sponsor by the 74th day after the FDA's receipt of the submission whether the application is sufficiently complete to permit substantive review. The FDA may request additional information rather than accept an NDA for filing. In this event, the application must be resubmitted with the additional information. The resubmitted application is also subject to review before the FDA accepts it for filing. Once the submission is accepted for filing, the FDA begins an in-depth substantive review. The FDA has agreed to specified performance goals in the review process of NDAs. Most such applications are meant to be reviewed within ten months from the date of filing, and most applications for "priority review" products are meant to be reviewed within six months of filing. The review process may be extended by the FDA for various reasons, including for three additional months to consider new information or clarification provided by the applicant to address an outstanding deficiency identified by the FDA following the original submission.

Before approving an NDA, the FDA typically will inspect the facility or facilities where the product is or will be manufactured. These pre-approval inspections cover all facilities associated with an NDA submission, including drug component manufacturing (such as Active Pharmaceutical Ingredients), finished drug product manufacturing, and control testing laboratories. The FDA will not approve an application unless it determines that the manufacturing processes and facilities are in compliance with cGMP requirements and adequate to assure consistent production of the product within required specifications. Additionally, before approving an NDA, the FDA will typically inspect one or more clinical sites to assure compliance with GCP.

The FDA may refer an application for a novel drug to an advisory committee or explain why such referral was not made. Typically, an advisory committee is a panel of independent experts, including clinicians and other scientific experts, that reviews, evaluates and provides a recommendation as to whether the application should be approved and under what conditions. The FDA is not bound by the recommendations of an advisory committee, but it considers such recommendations carefully when making decisions.

Accelerated Approval Pathway

The FDA may grant accelerated approval to a drug for a serious or life-threatening condition that provides meaningful therapeutic advantage to patients over existing treatments based upon a determination that the drug has an effect on a surrogate endpoint that is reasonably likely to predict

clinical benefit. The FDA may also grant accelerated approval for such a condition when the product has an effect on an intermediate clinical endpoint that can be measured earlier than an effect on irreversible morbidity or mortality, or IMM, and that is reasonably likely to predict an effect on irreversible morbidity or mortality or other clinical benefit, taking into account the severity, rarity, or prevalence of the condition and the availability or lack of alternative treatments. Drugs granted accelerated approval must meet the same statutory standards for safety and effectiveness as those granted traditional approval.

For the purposes of accelerated approval, a surrogate endpoint is a marker, such as a laboratory measurement, radiographic image, physical sign, or other measure that is thought to predict clinical benefit, but is not itself a measure of clinical benefit. Surrogate endpoints can often be measured more easily or more rapidly than clinical endpoints. An intermediate clinical endpoint is a measurement of a therapeutic effect that is considered reasonably likely to predict the clinical benefit of a drug, such as an effect on IMM. The FDA has limited experience with accelerated approvals based on intermediate clinical endpoints, but has indicated that such endpoints generally may support accelerated approval where the therapeutic effect measured by the endpoint is not itself a clinical benefit and basis for traditional approval, if there is a basis for concluding that the therapeutic effect is reasonably likely to predict the ultimate clinical benefit of a drug.

The accelerated approval pathway is most often used in settings in which the course of a disease is long and an extended period of time is required to measure the intended clinical benefit of a drug, even if the effect on the surrogate or intermediate clinical endpoint occurs rapidly. The accelerated approval pathway is usually contingent on a sponsor's agreement to conduct, in a diligent manner, additional post-approval confirmatory studies to verify and describe the drug's clinical benefit. As a result, a product candidate approved on this basis is subject to rigorous post-marketing compliance requirements, including the completion of Phase 4 or post-approval clinical trials to confirm the effect on the clinical endpoint. Failure to conduct required post-approval studies, or confirm a clinical benefit during post-marketing studies, would allow the FDA to withdraw the drug from the market on an expedited basis. All promotional materials for product candidates approved under accelerated regulations are subject to prior review by the FDA.

The FDA's Decision on an NDA

On the basis of the FDA's evaluation of the NDA and accompanying information, including the results of the inspection of the manufacturing facilities, the FDA may issue an approval letter or a complete response letter. An approval letter authorizes commercial marketing of the product with specific prescribing information for specific indications. A complete response letter generally outlines the deficiencies in the submission and may require substantial additional testing or information in order for the FDA to reconsider the application. If and when those deficiencies have been addressed to the FDA's satisfaction in a resubmission of the NDA, the FDA will issue an approval letter. The FDA has committed to reviewing such resubmissions in two or six months depending on the type of information included. Even with submission of this additional information, the FDA ultimately may decide that the application does not satisfy the regulatory criteria for approval.

If the FDA approves a product, it may limit the approved indications for use for the product, require that contraindications, warnings or precautions be included in the product labeling, require that post-approval studies, including Phase 4 clinical trials, be conducted to further assess the drug's safety after approval, require testing and surveillance programs to monitor the product after commercialization, or impose other conditions which can materially affect the potential market and profitability of the product. In addition, as a condition of approval, the FDA may require an applicant to develop a REMS. REMS use risk minimization strategies beyond the professional labeling to ensure that the benefits of the product outweigh the potential risks. To determine whether a REMS is needed, the FDA will consider the size of the population likely to use the product, seriousness of the disease,

expected benefit of the product, expected duration of treatment, seriousness of known or potential adverse events, and whether the product is a new molecular entity. REMS can include medication guides, physician communication plans for healthcare professionals, and elements to assure safe use (ETASU). ETASU may include, but are not limited to, special training or certification for prescribing or dispensing, dispensing only under certain circumstances, special monitoring, and the use of patient registries. The FDA may require a REMS before approval or post-approval if it becomes aware of a serious risk associated with use of the product. The requirement for a REMS can materially affect the potential market and profitability of a product.

The FDA may prevent or limit further marketing of a product based on the results of post-market studies or surveillance programs. After approval, many types of changes to the approved product, such as adding new indications, manufacturing changes and additional labeling claims, are subject to further testing requirements and FDA review and approval.

Post-Approval Requirements

Drugs manufactured or distributed pursuant to FDA approvals are subject to pervasive and continuing regulation by the FDA, including, among other things, requirements relating to recordkeeping, periodic reporting, product sampling and distribution, advertising and promotion and reporting of adverse experiences with the product. After approval, most changes to the approved product, such as adding new indications or other labeling claims, are subject to prior FDA review and approval. There also are continuing, annual user fee requirements for any marketed products and the establishments at which such products are manufactured, as well as new application fees for supplemental applications with clinical data.

In addition, drug manufacturers and other entities involved in the manufacture and distribution of approved drugs are required to register their establishments with the FDA and state agencies, and are subject to periodic unannounced inspections by the FDA and these state agencies for compliance with cGMP requirements. Changes to the manufacturing process are strictly regulated and often require prior FDA approval before being implemented. FDA regulations also require investigation and correction of any deviations from cGMP and impose reporting and documentation requirements upon the sponsor and any third-party manufacturers that the sponsor may decide to use. Accordingly, manufacturers must continue to expend time, money, and effort in the area of production and quality control to maintain cGMP compliance.

Once an approval is granted, the FDA may withdraw the approval if compliance with regulatory requirements and standards is not maintained or if problems occur after the product reaches the market. Later discovery of previously unknown problems with a product, including adverse events of unanticipated severity or frequency, or with manufacturing processes, or failure to comply with regulatory requirements, may result in revisions to the approved labeling to add new safety information; imposition of post-market studies or clinical trials to assess new safety risks; or imposition of distribution or other restrictions under a REMS program. Other potential consequences include, among other things:

- restrictions on the marketing or manufacturing of the product, complete withdrawal of the product from the market or product recalls;
- fines, warning letters or holds on post-approval clinical trials;
- refusal of the FDA to approve pending NDAs or supplements to approved NDAs, or suspension or revocation of product license approvals;
- product seizure or detention, or refusal to permit the import or export of products; or
- injunctions or the imposition of civil or criminal penalties.

The FDA strictly regulates marketing, labeling, advertising and promotion of products that are placed on the market. Drugs may be promoted only for the approved indications and in accordance with the provisions of the approved label. The FDA and other agencies actively enforce the laws and regulations prohibiting the promotion of off-label uses, and a company that is found to have improperly promoted off-label uses may be subject to significant liability.

In addition, the distribution of prescription pharmaceutical products is subject to the Prescription Drug Marketing Act, or PDMA, which regulates the distribution of drugs and drug samples at the federal level, and sets minimum standards for the registration and regulation of drug distributors by the states. Both the PDMA and state laws limit the distribution of prescription pharmaceutical product samples and impose requirements to ensure accountability in distribution.

Abbreviated New Drug Applications for Generic Drugs

In 1984, with passage of the Hatch-Waxman Amendments to the FDCA, Congress authorized the FDA to approve generic drugs that are the same as drugs previously approved by the FDA under the NDA provisions of the statute. To obtain approval of a generic drug, an applicant must submit an abbreviated new drug application, or ANDA, to the agency. In support of such applications, a generic manufacturer may rely on the pre-clinical and clinical testing previously conducted for a drug product previously approved under an NDA, known as the reference listed drug, or RLD.

Specifically, in order for an ANDA to be approved, the FDA must find that the generic version is identical to the RLD with respect to the active ingredients, the route of administration, the dosage form, and the strength of the drug. At the same time, the FDA must also determine that the generic drug is "bioequivalent" to the innovator drug. Under the statute, a generic drug is bioequivalent to a RLD if the rate and extent of absorption of the drug do not show a significant difference from the rate and extent of absorption of the listed drug.

Upon approval of an ANDA, the FDA indicates whether the generic product is "therapeutically equivalent" to the RLD in its publication "Approved Drug Products with Therapeutic Equivalence Evaluations," also referred to as the "Orange Book." Physicians and pharmacists consider a therapeutic equivalent generic drug to be fully substitutable for the RLD. In addition, by operation of certain state laws and numerous health insurance programs, the FDA's designation of therapeutic equivalence often results in substitution of the generic drug without the knowledge or consent of either the prescribing physician or patient.

Under the Hatch-Waxman Amendments, the FDA may not approve an ANDA until any applicable period of non-patent exclusivity for the RLD has expired. The FDCA provides a period of five years of non-patent data exclusivity for a new drug containing a new chemical entity. In cases where such exclusivity has been granted, an ANDA may not be filed with the FDA until the expiration of five years unless the submission is accompanied by a Paragraph IV certification, in which case the applicant may submit its application four years following the original product approval. The FDCA also provides for a period of three years of exclusivity if the NDA includes reports of one or more new clinical investigations, other than bioavailability or bioequivalence studies, that were conducted by or for the applicant and are essential to the approval of the application. This three-year exclusivity period often protects changes to a previously approved drug product, such as a new dosage form, route of administration, combination or indication.

Hatch-Waxman Patent Certification and the 30-Month Stay

Upon approval of an NDA or a supplement thereto, NDA sponsors are required to list with the FDA each patent with claims that cover the applicant's product or an approved method of using the product. Each of the patents listed by the NDA sponsor is published in the Orange Book. When an

ANDA applicant files its application with the FDA, the applicant is required to certify to the FDA concerning any patents listed for the reference product in the Orange Book, except for patents covering methods of use for which the ANDA applicant is not seeking approval. To the extent that the Section 505(b)(2) applicant is relying on studies conducted for an already approved product, the applicant is required to certify to the FDA concerning any patents listed for the approved product in the Orange Book to the same extent that an ANDA applicant would.

Specifically, the applicant must certify with respect to each patent that:

- the required patent information has not been filed;
- the listed patent has expired;
- the listed patent has not expired, but will expire on a particular date and approval is sought after patent expiration; or
- the listed patent is invalid, unenforceable or will not be infringed by the new product.

A certification that the new product will not infringe the already approved product's listed patents or that such patents are invalid or unenforceable is called a Paragraph IV certification. If the applicant does not challenge the listed patents or indicate that it is not seeking approval of a patented method of use, the ANDA application will not be approved until all the listed patents claiming the referenced product have expired.

If the ANDA applicant or 505(b)(2) applicant has provided a Paragraph IV certification to the FDA, the applicant must also send notice of the Paragraph IV certification to the NDA and patent holders once the ANDA has been accepted for filing by the FDA. The NDA and patent holders may then initiate a patent infringement lawsuit in response to the notice of the Paragraph IV certification. The filing of a patent infringement lawsuit within 45 days after the receipt of a Paragraph IV certification automatically prevents the FDA from approving the ANDA until the earlier of 30 months after the receipt of the Paragraph IV notice, expiration of the patent, or a decision in the infringement case that is favorable to the ANDA applicant.

Orphan Designation and Exclusivity

Under the Orphan Drug Act, FDA may designate a drug product as an "orphan drug" if it is intended to treat a rare disease or condition (generally meaning that it affects fewer than 200,000 individuals in the United States, or more in cases in which there is no reasonable expectation that the cost of developing and making a drug product available in the United States for treatment of the disease or condition will be recovered from sales of the product). A company must request orphan product designation before submitting a NDA. If the request is granted, FDA will disclose the identity of the therapeutic agent and its potential use. Orphan product designation does not convey any advantage in or shorten the duration of the regulatory review and approval process.

If a product with orphan status receives the first FDA approval for the disease or condition for which it has such designation, the product will be entitled to orphan product exclusivity. Orphan product exclusivity means that FDA may not approve any other applications for the same product for the same indication for seven years, except in certain limited circumstances. Competitors may receive approval of different products for the indication for which the orphan product has exclusivity and may obtain approval for the same product but for a different indication. If a drug or drug product designated as an orphan product ultimately receives marketing approval for an indication broader than what was designated in its orphan product application, it may not be entitled to exclusivity.

Pediatric Studies and Exclusivity

Under the Pediatric Research Equity Act of 2003, a NDA or supplement thereto must contain data that are adequate to assess the safety and effectiveness of the drug product for the claimed indications in all relevant pediatric subpopulations, and to support dosing and administration for each pediatric subpopulation for which the product is safe and effective. With enactment of the Food and Drug Administration Safety and Innovation Act, or FDASIA, in 2012, sponsors must also submit pediatric study plans prior to the assessment data. Those plans must contain an outline of the proposed pediatric study or studies the applicant plans to conduct, including study objectives and design, any deferral or waiver requests, and other information required by regulation. The applicant, the FDA, and the FDA's internal review committee must then review the information submitted, consult with each other, and agree upon a final plan. The FDA or the applicant may request an amendment to the plan at any time.

The FDA may, on its own initiative or at the request of the applicant, grant deferrals for submission of some or all pediatric data until after approval of the product for use in adults, or full or partial waivers from the pediatric data requirements. Additional requirements and procedures relating to deferral requests and requests for extension of deferrals are contained in FDASIA.

Pediatric exclusivity is another type of non-patent marketing exclusivity in the United States and, if granted, provides for the attachment of an additional six months of marketing protection to the term of any existing regulatory exclusivity, including the non-patent exclusivity. This six-month exclusivity may be granted if an NDA sponsor submits pediatric data that fairly respond to a written request from the FDA for such data. The data do not need to show the product to be effective in the pediatric population studied; rather, if the clinical trial is deemed to fairly respond to the FDA's request, the additional protection is granted. If reports of requested pediatric studies are submitted to and accepted by the FDA within the statutory time limits, whatever statutory or regulatory periods of exclusivity or patent protection cover the product are extended by six months. This is not a patent term extension, but it effectively extends the regulatory period during which the FDA cannot approve another application.

Patent Term Restoration and Extension

A patent claiming a new drug product or medical device may be eligible for a limited patent term extension under the Hatch-Waxman Act, which permits a patent restoration of up to five years for patent term lost during product development and the FDA regulatory review. The restoration period granted on a patent covering a new drug product or a Class III medical device is typically one-half the time between the date a clinical investigation on human beings is begun and the submission date of an application for premarket approval of the product or medical device, plus the time between the submission date of an application for approval of the product or medical device and the ultimate approval date. Patent term restoration cannot be used to extend the remaining term of a patent past a total of 14 years from the product's approval date. Only one patent applicable to an approved drug product or medical device is eligible for the extension, and the application for the extension must be submitted prior to the expiration of the patent in question. A patent that covers multiple drugs or medical devices for which approval is sought can only be extended in connection with one of the approvals. The USPTO reviews and approves the application for any patent term extension or restoration in consultation with the FDA.

Review and Approval of Medical Devices in the United States

Medical devices in the United States are strictly regulated by the FDA. Under the FDCA a medical device is defined as an instrument, apparatus, implement, machine, contrivance, implant, *in vitro* reagent, or other similar or related article, including a component, part or accessory which is, among other things: intended for use in the diagnosis of disease or other conditions, or in the cure,

mitigation, treatment, or prevention of disease, in man or other animals; or intended to affect the structure or any function of the body of man or other animals, and which does not achieve its primary intended purposes through chemical action within or on the body of man or other animals and which is not dependent upon being metabolized for the achievement of any of its primary intended purposes. This definition provides a clear distinction between a medical device and other FDA regulated products such as drugs. If the primary intended use of the product is achieved through chemical action or by being metabolized by the body, the product is usually a drug. If not, it is generally a medical device.

Unless an exemption applies, a new medical device may not be marketed in the United States unless and until it has been cleared through filing of a 510(k) premarket notification, or 510(k), or approved by the FDA pursuant to a PMA. The information that must be submitted to the FDA in order to obtain clearance or approval to market a new medical device varies depending on how the medical device is classified by the FDA. Medical devices are classified into one of three classes on the basis of the controls deemed by the FDA to be necessary to reasonably ensure their safety and effectiveness.

Class I devices are those low risk devices for which reasonable assurance of safety and effectiveness can be provided by adherence to the FDA's general controls for medical devices, which include applicable portions of the FDA's Quality System Regulation, or QSR, facility registration and product listing, reporting of adverse medical events and malfunctions and appropriate, truthful and non-misleading labeling, advertising and promotional materials. Many Class I devices are exempt from premarket regulation; however, some Class I devices require premarket clearance by the FDA through the 510(k) premarket notification process.

Class II devices are moderate risk devices and are subject to the FDA's general controls, and any other special controls, such as performance standards, post-market surveillance, and FDA guidelines, deemed necessary by the FDA to provide reasonable assurance of the devices' safety and effectiveness. Premarket review and clearance by the FDA for Class II devices are accomplished through the 510(k) premarket notification procedure, although some Class II devices are exempt from the 510(k) requirements. Premarket notifications are subject to user fees, unless a specific exemption applies.

Class III devices are deemed by the FDA to pose the greatest risk, such as those for which reasonable assurance of the device's safety and effectiveness cannot be assured solely by the general controls and special controls described above and that are life-sustaining or life-supporting. A PMA application must provide valid scientific evidence, typically extensive pre-clinical and clinical trial data and information about the device and its components regarding, among other things, device design, manufacturing and labeling. PMA applications (and supplemental PMA applications) are subject to significantly higher user fees than are 510(k) premarket notifications.

Clinical Studies in Support of Development of a Medical Device

The types of clinical studies required for the development and approval of a medical device differ from those required for drug products. Clinical trials involving a drug product typically involve a sequential process of Phase 1, 2 and 3 clinical trials to test for the safety and efficacy of the product. The clinical development of a medical device, on the other hand, is often conducted in three different sequential phases, which may overlap or be combined. Those phases are a pilot study, which may also be referred to as an early feasibility study; a feasibility study; and a pivotal study.

- Pilot Study: A pilot study is a limited clinical investigation of a device early in development, typically before the device design has been finalized, for a specific indication. It may be used to evaluate the device design concept with respect to initial clinical safety and device functionality in a small number of subjects (generally fewer than ten initial subjects) when this information cannot practically be provided through additional nonclinical assessments or appropriate

nonclinical tests are unavailable. Information obtained from a pilot study may guide device modifications.

- Feasibility Study: A feasibility study is a clinical investigation that is commonly used to capture preliminary safety and effectiveness information on a near-final or final device design to adequately plan an appropriate pivotal study. Because the study of a near-final or final device design takes place later in development than a pilot study, the FDA has indicated that it expects to see more nonclinical (or prior clinical) data in a feasibility study IDE application. A feasibility study does not necessarily need to be preceded by a pilot study.

- Pivotal Study: A pivotal study is a clinical investigation designed to collect definitive evidence of the safety and effectiveness of a device for a specified intended use, typically in a statistically justified number of subjects. Evidence from one or more pivotal clinical studies generally serves as the primary basis for the determination of reasonable assurance of safety and effectiveness of the medical device of a PMA and FDA's overall benefit-risk determination. A pivotal study may or may not be preceded by a pilot study or feasibility study.

These three stages in the development of a medical device may be dependent on each other and conducting a thorough evaluation in one stage can make the next stage more straightforward. To determine which type of clinical study is appropriate to pursue, a manufacturer will consider several factors, such as the novelty of the device, the device's intended clinical use, the stability of the device design and the amount of test data available to support the IDE application. A pilot study is appropriate when device changes are expected and when, due to the novelty of the device or its intended use, a clinical study is expected to provide information that cannot be practically obtained through additional nonclinical assessments. A pilot study may also be appropriate even if a device or a prototype of the device has previously been used clinically for the intended clinical use. A feasibility study or a pivotal study may be more appropriate if the device design is near-final or final, respectively, depending on the amount of data available to justify the study.

510(k) Premarket Notification

To obtain 510(k) clearance, a manufacturer must submit a premarket notification demonstrating that the proposed device is "substantially equivalent" to a predicate device, which is a previously cleared 510(k) device or a pre-amendment device that was in commercial distribution before May 28, 1976, for which the FDA has not yet called for the submission of a PMA application. The FDA's 510(k) clearance pathway usually takes from three to 12 months from the date the application is submitted and filed with the FDA, but it can take significantly longer and clearance is never assured. The FDA has issued guidance documents meant to expedite review of a 510(k) and facilitate interactions between applicants and the agency. To demonstrate substantial equivalence, a manufacturer must show that the device has the same intended use as a predicate device and the same technological characteristics, or the same intended use and different technological characteristics and does not raise new questions of safety and effectiveness than the predicate device. Most 510(k)s do not require clinical data for clearance, but the FDA may request such data.

The FDA seeks to review and act on a 510(k) within 90 days of submission, but it may take longer if the agency finds that it requires more information to review the 510(k). If the FDA determines that the device is substantially equivalent to a predicate device, the subject device may be marketed. However, if the FDA concludes that a new device is not substantially equivalent to a predicate device, the new device will be classified in Class III and the manufacturer will be required to submit a PMA to market the product. Devices of a new type that the FDA has not previously classified based on risk are automatically classified into Class III by operation of section 513(f)(1) of the FDCA, regardless of the level of risk they pose. To avoid requiring PMA review of low- to moderate-risk devices classified in Class III by operation of law, Congress enacted section 513(f)(2) of the FDCA. This provision allows

the FDA to classify a low- to moderate-risk device not previously classified into Class I or II, a process known as the *de novo* process. A company may apply directly to the FDA for classification of its device as *de novo* or may submit a de novo petition within 30 days of receiving a not substantially equivalent determination.

Modifications to a 510(k)-cleared medical device may require the submission of another 510(k). Modifications to a 510(k)-cleared device frequently require the submission of a traditional 510(k), but modifications meeting certain conditions may be candidates for FDA review under a Special 510(k). If a device modification requires the submission of a 510(k), but the modification does not affect the intended use of the device or alter the fundamental technology of the device, then summary information that results from the design control process associated with the cleared device can serve as the basis for clearing the application. A Special 510(k) allows a manufacturer to declare conformance to design controls without providing new data. When the modification involves a change in material, the nature of the "new" material will determine whether a traditional or Special 510(k) is necessary.

Any modification to a 510(k)-cleared product that would constitute a major change in its intended use or any change that could significantly affect the safety or effectiveness of the device may, in some circumstances, requires the submission of a PMA, if the change raises complex or novel scientific issues or the product has a new intended use. A manufacturer may be required to submit extensive pre-clinical and clinical data depending on the nature of the changes.

The FDA requires every manufacturer to make the determination regarding the need for a new 510(k) submission in the first instance, but the FDA may review any manufacturer's decision. If the FDA disagrees with the manufacturer's determination and requires new 510(k) clearances or PMA approvals for modifications to previously cleared products for which the manufacturer concluded that new clearances or approvals are unnecessary, the manufacturer may be required to cease marketing or distribution of the products or to recall the modified product until it obtains clearance or approval, and the manufacturer may be subject to significant regulatory fines or penalties. In addition, the FDA is currently evaluating the 510(k) process and may make substantial changes to industry requirements.

Premarket Approval Application

The PMA process for approval to market a medical device is more complex, costly, and time consuming than the 510(k) clearance procedure. A PMA must be supported by extensive data, including technical information regarding device design and development, pre-clinical studies, clinical studies, manufacturing and controls information and labeling information, that demonstrates the safety and effectiveness of the device for its intended use. After a PMA is submitted, the FDA has 45 days to determine whether it is sufficiently complete to permit a substantive review. If the PMA is complete, the FDA will file the PMA. If the FDA accepts the application for filing, the agency will begin an in-depth substantive review of the application. By statute, the FDA has 180 days to review the application although, generally, review of the application often takes between one and three years, and may take significantly longer. If the FDA has questions, it will likely issue a first major deficiency letter within 150 days of filing. It may also refer the PMA to an FDA advisory panel for additional review, and will conduct a preapproval inspection of the manufacturing facility to ensure compliance with the QSR, either of which could extend the 180-day response target. In addition, the FDA may request additional information or request the performance of additional clinical trials in which case the PMA approval may be delayed while the trials are conducted and the data acquired are submitted in an amendment to the PMA. Even with additional trials, the FDA may not approve the PMA application.

If the FDA's evaluations of both the PMA and the manufacturing facilities are favorable, the FDA will either issue an approval letter authorizing commercial marketing or an approvable letter that usually contains a number of conditions that must be met in order to secure final approval. If the FDA's evaluations are not favorable, the FDA will deny approval of the PMA or issue a not approvable

letter. The PMA process, including the gathering of clinical and nonclinical data and the submission to and review by the FDA, can take several years, and the process can be expensive and uncertain. Moreover, even if the FDA approves a PMA, the FDA may approve the device with an indication that is narrower or more limited than originally sought. The FDA can impose post-approval conditions that it believes necessary to ensure the safety and effectiveness of the device, including, among other things, restrictions on labeling, promotion, sale and distribution. After approval of a PMA, a new PMA or PMA supplement may be required for a modification to the device, its labeling, or its manufacturing process. PMA supplements often require submission of the same type of information as an initial PMA application, except that the supplement is limited to information needed to support any changes from the device covered by the approved PMA application and may or may not require as extensive technical or clinical data or the convening of an advisory panel. The time for review of a PMA supplement may vary depending on the type of change, but it can be lengthy. In addition, in some cases the FDA might require additional clinical data.

Investigational Device Exemption

A clinical trial is typically required for a PMA and, in a small percentage of cases, the FDA may require a clinical study in support of a 510(k) submission. A manufacturer that wishes to conduct a clinical study involving the device is subject to the FDA's IDE regulation. The IDE regulation distinguishes between significant and nonsignificant risk device studies and the procedures for obtaining approval to begin the study differ accordingly. Also, some types of studies are exempt from the IDE regulations.

Significant risk devices are, among other things, devices that are substantially important in diagnosing, curing, mitigating, or treating disease or in preventing impairment to human health and present a potential for serious risk to the health, safety or welfare of a subject. Studies of devices that pose a significant risk require both FDA and an IRB approval prior to initiation of a clinical study. Nonsignificant risk devices are devices that do not pose a significant risk to the human subjects. A nonsignificant risk device study requires only IRB approval prior to initiation of a clinical study.

An IDE application must be supported by appropriate data, such as animal and laboratory testing results, showing that it is safe to test the device in humans and that the testing protocol is scientifically sound. An IDE application is considered approved 30 days after it has been received by the FDA, unless the FDA otherwise informs the sponsor prior to 30 calendar days from the date of receipt that the IDE is approved, approved with conditions, or disapproved. The FDA typically grants IDE approval for a specified number of subjects to be enrolled at specified study centers. The clinical trial must be conducted in accordance with applicable regulations, including but not limited to the FDA's IDE regulations and GCP. The investigators must obtain subject informed consent, rigorously follow the investigational plan and study protocol, control the disposition of investigational devices, and comply with all reporting and record keeping requirements. A clinical trial may be suspended or terminated by the FDA, the IRB or the sponsor at any time for various reasons, including a belief that the risks to the study participants outweigh the benefits of participation in the trial. Approval of an IDE does not bind the FDA to accept the results of the trial as sufficient to prove the product's safety and efficacy, even if the trial meets its intended success criteria.

Humanitarian Use Device

When a medical device is intended to treat or diagnose a disease or condition that affects or is manifested in fewer than 4,000 individuals in the United States per year, a manufacturer may seek approval through a humanitarian device exemption, or HDE, application to market its product as a humanitarian use device, or HUD. This pathway provides an incentive for the development of devices for the treatment or diagnosis of diseases affecting small populations and where a manufacturer's

research and development costs could exceed market return. Thus, the purpose of the HDE is to encourage device manufacturers to develop devices for rare conditions or diseases.

Prior to submitting the HDE application the device manufacturer must request HUD designation from the FDA's Office of Orphan Products Development. The FDA seeks to respond to the request within 45 days of submission. If granted, a manufacturer may file an HDE application for HUD approval.

An HDE application is similar to a PMA application but is exempt from the effectiveness requirements of a PMA. In submitting an HDE application a manufacturer is not required to include scientifically valid clinical investigation results demonstrating that the device is effective for its intended purpose. However, the application must contain sufficient information for the FDA to determine that the device does not pose an unreasonable or significant risk of illness or injury, and that the probable benefit to health outweighs the risk of injury or illness from its use, taking into account the probable risks and benefits of currently available devices or alternative forms of treatment. The manufacturer must also demonstrate that no comparable devices are available to treat or diagnose the disease or condition, and that the manufacturer could not otherwise bring the device to market. The FDA seeks to act on an HDE application within 75 days after accepting the HDE for filing.

If the FDA approves the HDE, the manufacturer may market the HUD. However, an HUD may only be used in facilities that have established an IRB to supervise clinical testing of devices and after an IRB has approved the use of the device to treat or diagnose the specific disease. HUDs are also subject to specific labeling requirements identifying the device as a HUD device and noting that although the device is authorized by the FDA, the effectiveness of the device for the specific indication has not been demonstrated. Moreover, a manufacturer cannot charge an amount for an HDE approved device that exceeds the costs of research and development, fabrication, and distribution.

Expedited Access PMA

The FDA has proposed a program to provide earlier access to high-risk medical devices that are intended to treat or diagnose patients with serious conditions whose medical needs are unmet by current technology. The Expedited Access Premarket Approval Application for Unmet Medical Needs for Life Threatening or Irreversibly Debilitating Diseases or Conditions program, or "Expedited Access PMA" or "EAP," allows for earlier and more interactive engagement with FDA staff. It also involves senior FDA management and a collaboratively developed plan for collecting scientific and clinical data to support approval—taken together, these features are meant to provide patients with earlier access to safe and effective medical devices by reducing the time associated with product development.

To be eligible for participation in the program, the medical device must be intended to treat or diagnose a life-threatening or irreversibly-debilitating disease or condition and represent one of the following:

- no approved alternative treatment exists;
- a breakthrough technology that provides a clinically meaningful advantage over existing technology;
- offers a significant, clinically meaningful advantage over existing approved alternatives; or
- availability of the device is in the patient's best interest.

The EAP must be accompanied by an acceptable data development plan that has been approved by the FDA. When utilizing the EAP program, the FDA will continue to apply the current approval standard of demonstrating a reasonable assurance of safety and efficacy.

Post-Marketing Restrictions and Enforcement

After a device is placed on the market, numerous regulatory requirements apply. These include, but are not limited to:

- submitting and updating establishment registration and device listings with the FDA;
- compliance with the QSR, which requires manufacturers to follow stringent design, testing, control, documentation, record maintenance, including maintenance of complaint and related investigation files, and other quality assurance controls during the manufacturing process;
- unannounced routine or for-cause device inspections by the FDA, which may include our suppliers' facilities; and
- labeling regulations, which prohibit the promotion of products for uncleared or unapproved or "off-label" uses and impose other restrictions on labeling; post-approval restrictions or conditions, including requirements to conduct post-market surveillance studies to establish continued safety data or tracking products through the chain of distribution to the patient level.

Under the FDA medical device reporting, or MDR, regulations, medical device manufacturers are required to report to the FDA information that a device has or may have caused or contributed to a death or serious injury or has malfunctioned in a way that would likely cause or contribute to death or serious injury if the malfunction of the device or a similar device of such manufacturer were to recur. The decision to file an MDR involves a judgment by the manufacturer. If the FDA disagrees with the manufacturer's determination, the FDA can take enforcement action.

Additionally, the FDA has the authority to require the recall of commercialized products in the event of material deficiencies or defects in design or manufacture. The authority to require a recall must be based on an FDA finding that there is reasonable probability that the device would cause serious adverse health consequences or death. Manufacturers may, under their own initiative, recall a product if any material deficiency in a device is found. The FDA requires that certain classifications of recalls be reported to the FDA within ten working days after the recall is initiated.

The failure to comply with applicable regulatory requirements can result in enforcement action by the FDA, which may include any of the following sanctions:

- untitled letters, warning letters, fines, injunctions or civil penalties;
- recalls, detentions or seizures of products;
- operating restrictions;
- delays in the introduction of products into the market;
- total or partial suspension of production;
- delay or refusal of the FDA or other regulators to grant 510(k) clearance or PMA approvals of new products;
- withdrawals of 510(k) clearance or PMA approvals; or
- in the most serious cases, criminal prosecution.

To ensure compliance with regulatory requirements, medical device manufacturers are subject to market surveillance and periodic, pre-scheduled and unannounced inspections by the FDA, and these inspections may include the manufacturing facilities of subcontractors.

Review and Approval of Combination Products in the United States

Certain products may be comprised of components that would normally be regulated under different types of regulatory authorities, and frequently by different Centers at the FDA. These products are known as combination products. Specifically, under regulations issued by the FDA, a combination product may be:

- a product comprised of two or more regulated components that are physically, chemically, or otherwise combined or mixed and produced as a single entity;
- two or more separate products packaged together in a single package or as a unit and comprised of drug and device products;
- a drug or device packaged separately that according to its investigational plan or proposed labeling is intended for use only with an approved individually specified drug or device where both are required to achieve the intended use, indication, or effect and where upon approval of the proposed product the labeling of the approved product would need to be changed, e.g., to reflect a change in intended use, dosage form, strength, route of administration, or significant change in dose; or
- any investigational drug or device packaged separately that according to its proposed labeling is for use only with another individually specified investigational drug, device, or biological product where both are required to achieve the intended use, indication, or effect.

Under the FDCA, the FDA is charged with assigning a center with primary jurisdiction, or a lead center, for review of a combination product. That determination is based on the "primary mode of action" of the combination product. Thus, if the primary mode of action of a device-drug combination product is attributable to the drug product, the FDA Center responsible for premarket review of the drug product would have primary jurisdiction for the combination product. The FDA has also established an Office of Combination Products to address issues surrounding combination products and provide more certainty to the regulatory review process. That office serves as a focal point for combination product issues for agency reviewers and industry. It is also responsible for developing guidance and regulations to clarify the regulation of combination products, and for assignment of the FDA center that has primary jurisdiction for review of combination products where the jurisdiction is unclear or in dispute.

Review and Approval of Drug Products in the European Union

In order to market any product outside of the United States, a company must also comply with numerous and varying regulatory requirements of other countries and jurisdictions regarding quality, safety and efficacy and governing, among other things, clinical trials, marketing authorization, commercial sales and distribution of drug products. Whether or not it obtains FDA approval for a product, the company would need to obtain the necessary approvals by the comparable foreign regulatory authorities before it can commence clinical trials or marketing of the product in those countries or jurisdictions. The approval process ultimately varies between countries and jurisdictions and can involve additional product testing and additional administrative review periods. The time required to obtain approval in other countries and jurisdictions might differ from and be longer than that required to obtain FDA approval. Regulatory approval in one country or jurisdiction does not ensure regulatory approval in another, but a failure or delay in obtaining regulatory approval in one country or jurisdiction may negatively impact the regulatory process in others.

Pursuant to the European Clinical Trials Directive, a system for the approval of clinical trials in the European Union has been implemented through national legislation of the member states. Under this system, an applicant must obtain approval from the competent national authority of a European Union member state in which the clinical trial is to be conducted. Furthermore, the applicant may only

start a clinical trial after a competent ethics committee has issued a favorable opinion. Clinical trial application must be accompanied by an investigational medicinal product dossier with supporting information prescribed by the European Clinical Trials Directive and corresponding national laws of the member states and further detailed in applicable guidance documents.

To obtain marketing approval of a drug under European Union regulatory systems, an applicant must submit a marketing authorization application, or MAA, either under a centralized or decentralized procedure.

The centralized procedure provides for the grant of a single marketing authorization by the European Commission that is valid for all European Union member states. The centralized procedure is compulsory for specific products, including for medicines produced by certain biotechnological processes, products designated as orphan medicinal products, advanced therapy products and products with a new active substance indicated for the treatment of certain diseases. For products with a new active substance indicated for the treatment of other diseases and products that are highly innovative or for which a centralized process is in the interest of patients, the centralized procedure may be optional.

Under the centralized procedure, the Committee for Medicinal Products for Human Use, or the CHMP, established at the EMA, is responsible for conducting the initial assessment of a drug. The CHMP is also responsible for several post-authorization and maintenance activities, such as the assessment of modifications or extensions to an existing marketing authorization. Under the centralized procedure in the European Union, the maximum timeframe for the evaluation of an MAA is 210 days, excluding clock stops, when additional information or written or oral explanation is to be provided by the applicant in response to questions of the CHMP. Accelerated evaluation might be granted by the CHMP in exceptional cases, when a medicinal product is of major interest from the point of view of public health and in particular from the viewpoint of therapeutic innovation. In this circumstance, the EMA ensures that the opinion of the CHMP is given within 150 days.

The decentralized procedure is available to applicants who wish to market a product in various European Union member states where such product has not received marketing approval in any European Union member states before. The decentralized procedure provides for approval by one or more other, or concerned, member states of an assessment of an application performed by one member state designated by the applicant, known as the reference member state. Under this procedure, an applicant submits an application based on identical dossiers and related materials, including a draft summary of product characteristics, and draft labeling and package leaflet, to the reference member state and concerned member states. The reference member state prepares a draft assessment report and drafts of the related materials within 210 days after receipt of a valid application. Within 90 days of receiving the reference member state's assessment report and related materials, each concerned member state must decide whether to approve the assessment report and related materials.

If a member state cannot approve the assessment report and related materials on the grounds of potential serious risk to public health, the disputed points are subject to a dispute resolution mechanism and may eventually be referred to the European Commission, whose decision is binding on all member states.

Review and Approval of Medical Devices in the European Union

The European Union has adopted numerous directives and standards regulating, among other things, the design, manufacture, clinical trials, labeling, approval and adverse event reporting for medical devices. In the European Union, medical devices must comply with the Essential Requirements in Annex I to the EU Medical Devices Directive (Council Directive 93/42/EEC), or the Essential Requirements. Compliance with these requirements is a prerequisite to be able to affix the CE mark of conformity to medical devices, without which they cannot be marketed or sold in the European Economic Area, or EEA, comprised of the European Union member states plus Norway, Iceland, and

Liechtenstein. Actual implementation of these directives, however, may vary on a country-by-country basis.

To demonstrate compliance with the Essential Requirements a manufacturer must undergo a conformity assessment procedure, which varies according to the type of medical device and its classification. Except for low risk medical devices, where the manufacturer can issue a CE Declaration of Conformity based on a self-assessment of the conformity of its products with the Essential Requirements, a conformity assessment procedure requires the intervention of a third-party organization designated by competent authorities of a European Union country to conduct conformity assessments, or a Notified Body. Notified Bodies are independent testing houses, laboratories, or product certifiers typically based within the European Union and authorized by the European member states to perform the required conformity assessment tasks, such as quality system audits and device compliance testing. The Notified Body would typically audit and examine the product's Technical File and the quality system for the manufacture, design and final inspection of the product before issuing a CE Certificate of Conformity demonstrating compliance with the relevant Essential Requirements.

Medical device manufacturers must carry out a clinical evaluation of their medical devices to demonstrate conformity with the relevant Essential Requirements. This clinical evaluation is part of the product's Technical File. A clinical evaluation includes an assessment of whether a medical device's performance is in accordance with its intended use, and that the known and foreseeable risks linked to the use of the device under normal conditions are minimized and acceptable when weighed against the benefits of its intended purpose. The clinical evaluation conducted by the manufacturer must also address any clinical claims, the adequacy of the device labeling and information (particularly claims, contraindications, precautions and warnings) and the suitability of related Instructions for Use. This assessment must be based on clinical data, which can be obtained from clinical studies conducted on the devices being assessed, scientific literature from similar devices whose equivalence with the assessed device can be demonstrated or both clinical studies and scientific literature.

With respect to implantable devices or devices classified as Class III in the European Union, the manufacturer must conduct clinical studies to obtain the required clinical data, unless relying on existing clinical data from similar devices can be justified. As part of the conformity assessment process, depending on the type of devices, the Notified Body will review the manufacturer's clinical evaluation process, assess the clinical evaluation data of a representative sample of the device's subcategory or generic group, or assess all the clinical evaluation data, verify the manufacturer's assessment of that data and assess the validity of the clinical evaluation report and the conclusions drawn by the manufacturer.

Even after a manufacturer receives a CE Certificate of Conformity enabling the CE mark on it products and the right to sell the products in the EEA countries, a Notified Body or a competent authority may require post-marketing studies of the products. Failure to comply with such requirements in a timely manner could result in the withdrawal of the CE Certificate of Conformity and the recall or withdrawal of the subject product from the European market.

A manufacturer must inform the Notified Body that carried out the conformity assessment of the medical devices of any planned substantial changes to the devices which could affect compliance with the Essential Requirements or the devices' intended purpose. The Notified Body will then assess the changes and verify whether they affect the product's conformity with the Essential Requirements or the conditions for the use of the devices. If the assessment is favorable, the Notified Body will issue a new CE Certificate of Conformity or an addendum to the existing CE Certificate of Conformity attesting compliance with the Essential Requirements. If it is not, the manufacturer may not be able to continue to market and sell the product in the EEA.

In the European Union, medical devices may be promoted only for the intended purpose for which the devices have been CE marked. Failure to comply with this requirement could lead to the

imposition of penalties by the competent authorities of the European Union Member States. The penalties could include warnings, orders to discontinue the promotion of the medical device, seizure of the promotional materials and fines. Promotional materials must also comply with various laws and codes of conduct developed by medical device industry bodies in the European Union governing promotional claims, comparative advertising, advertising of medical devices reimbursed by the national health insurance systems and advertising to the general public.

Additionally, all manufacturers placing medical devices in the market in the European Union are legally bound to report any serious or potentially serious incidents involving devices they produce or sell to the competent authority in whose jurisdiction the incident occurred. In the European Union, manufacturers must comply with the EU Medical Device Vigilance System. Under this system, incidents must be reported to the relevant authorities of the European Union countries, and manufacturers are required to take Field Safety Corrective Actions, or FSCAs, to reduce a risk of death or serious deterioration in the state of health associated with the use of a medical device that is already placed on the market. An incident is defined as any malfunction or deterioration in the characteristics and/or performance of a device, as well as any inadequacy in the labeling or the instructions for use which, directly or indirectly, might lead to or might have led to the death of a patient or user or of other persons or to a serious deterioration in their state of health. An FSCA may include the recall, modification, exchange, destruction or retrofitting of the device. FSCAs must be communicated by the manufacturer or its European Authorized Representative to its customers and to the end users of the device through Field Safety Notices. In September 2012, the European Commission adopted a proposal for a regulation which, if adopted, will change the way that most medical devices are regulated in the European Union, and may subject products to additional requirements.

Pharmaceutical Coverage, Pricing and Reimbursement

Significant uncertainty exists as to the coverage and reimbursement status of products approved by the FDA and other government authorities. Sales of products will depend, in part, on the extent to which the costs of the products will be covered by third-party payors, including government health programs in the United States such as Medicare and Medicaid, commercial health insurers and managed care organizations. The process for determining whether a payor will provide coverage for a product may be separate from the process for setting the price or reimbursement rate that the payor will pay for the product once coverage is approved. Third-party payors may limit coverage to specific products on an approved list, or formulary, which might not include all of the approved products for a particular indication. Additionally, the containment of healthcare costs has become a priority of federal and state governments, and the prices of drugs have been a focus in this effort. The U.S. government, state legislatures and foreign governments have shown significant interest in implementing cost-containment programs, including price controls, restrictions on reimbursement and requirements for substitution of generic products. Adoption of price controls and cost-containment measures, and adoption of more restrictive policies in jurisdictions with existing controls and measures, could further limit our net revenue and results.

In order to secure coverage and reimbursement for any product that might be approved for sale, a company may need to conduct expensive pharmacoeconomic studies in order to demonstrate the medical necessity and cost-effectiveness of the product, in addition to the costs required to obtain FDA or other comparable regulatory approvals. A payor's decision to provide coverage for a product does not imply that an adequate reimbursement rate will be approved. Third-party reimbursement may not be sufficient to maintain price levels high enough to realize an appropriate return on investment in product development.

In the European Union, pricing and reimbursement schemes vary widely from country to country. Some countries provide that drug products may be marketed only after a reimbursement price has been agreed. Some countries may require the completion of additional studies that compare the

cost-effectiveness of a particular product candidate to currently available therapies. For example, the European Union provides options for its member states to restrict the range of drug products for which their national health insurance systems provide reimbursement and to control the prices of medicinal products for human use. European Union member states may approve a specific price for a drug product or it may instead adopt a system of direct or indirect controls on the profitability of the company placing the drug product on the market. Other member states allow companies to fix their own prices for drug products, but monitor and control company profits. The downward pressure on health care costs in general, particularly prescription drugs, has become intense. As a result, increasingly high barriers are being erected to the entry of new products. In addition, in some countries, cross-border imports from low-priced markets exert competitive pressure that may reduce pricing within a country. Any country that has price controls or reimbursement limitations for drug products may not allow favorable reimbursement and pricing arrangements.

Healthcare Law and Regulation

Healthcare providers, physicians and third-party payors play a primary role in the recommendation and prescription of drug products that are granted marketing approval. Arrangements with third-party payors and customers are subject to broadly applicable fraud and abuse and other healthcare laws and regulations. Such restrictions under applicable federal and state healthcare laws and regulations, include the following:

- the federal Anti-Kickback Statute prohibits, among other things, persons from knowingly and willfully soliciting, offering, receiving or providing remuneration, directly or indirectly, in cash or in kind, to induce or reward either the referral of an individual for, or the purchase, order or recommendation of, any good or service, for which payment may be made, in whole or in part, under a federal healthcare program such as Medicare and Medicaid;

- the federal False Claims Act imposes civil penalties, and provides for civil whistleblower or qui tam actions, against individuals or entities for knowingly presenting, or causing to be presented, to the federal government, claims for payment that are false or fraudulent or making a false statement to avoid, decrease or conceal an obligation to pay money to the federal government;

- the federal Health Insurance Portability and Accountability Act of 1996, or HIPAA, imposes criminal and civil liability for executing a scheme to defraud any healthcare benefit program or making false statements relating to healthcare matters;

- HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act and its implementing regulations, also imposes obligations, including mandatory contractual terms, with respect to safeguarding the privacy, security and transmission of individually identifiable health information;

- the federal false statements statute prohibits knowingly and willfully falsifying, concealing or covering up a material fact or making any materially false statement in connection with the delivery of or payment for healthcare benefits, items or services;

- the federal transparency requirements under the Patient Protection and Affordable Care Act, as amended by the Health Care and Education Reconciliation Act of 2010, or collectively the PPACA will require applicable manufacturers of covered drugs, devices, drugs and medical supplies to report to the Department of Health and Human Services information related to payments and other transfers of value to physicians and teaching hospitals and physician ownership and investment interests; and

- analogous state and foreign laws and regulations, such as state anti-kickback and false claims laws, may apply to sales or marketing arrangements and claims involving healthcare items or services reimbursed by non-governmental third-party payors, including private insurers.

Some state laws require pharmaceutical companies to comply with the pharmaceutical industry's voluntary compliance guidelines and the relevant compliance guidance promulgated by the federal government in addition to requiring drug manufacturers to report information related to payments to physicians and other health care providers or marketing expenditures. State and foreign laws also govern the privacy and security of health information in some circumstances, many of which differ from each other in significant ways and often are not preempted by HIPAA, thus complicating compliance efforts.

Employees

As of March 31, 2014, we had 55 full-time employees, of which 51 employees were engaged in research and development, and four employees provided general and administrative support. Of our employees, 31 have earned advanced degrees. Our employees are not represented by a labor union or covered by a collective bargaining agreement.

Facilities

Our principal facilities consist of approximately 25,000 square feet of office space at Ikaria's headquarters located in Hampton, New Jersey and approximately 3,200 square feet of office space and research lab facilities at the Commercialization Center for Innovative Technologies located in North Brunswick, New Jersey. We have access to the office space at Ikaria's headquarters until February 2016, pursuant to the TSA. We lease the space in North Brunswick, New Jersey under a lease that expires in March 2015.

Legal Proceedings

We are not presently a party to any material litigation or regulatory proceeding and, except as described below, we are not aware of any pending or threatened litigation or regulatory proceeding against us that could have a material adverse effect on our business, operating results, financial condition or cash flows.

We have recently been engaged in discussions with BioLine relating to our performance under our license agreement. BioLine has indicated to us that it believes that we have breached the license agreement in several ways, including, but not limited to, failure to use commercially reasonable efforts to develop BCM, failure to provide BioLine with material information concerning the development and commercialization plans for BCM and failure to notify BioLine in advance of material public disclosures regarding BCM. We and BioLine also disagree about the timing of a $12.5 million milestone payment that we would owe BioLine based upon progress in our BCM clinical development program. Although we have had multiple discussions with BioLine on these issues, and these discussions are continuing, we have not been able to resolve these outstanding issues. If we are unable to reach agreement with BioLine on these issues, BioLine could bring a lawsuit against us, although any claims relating to our alleged failure to use commercially reasonable efforts would first be subject to a non-binding 60-day mediation period with a third-party mediator. We believe that we would have strong defenses in any litigation that could be brought by BioLine. If BioLine were to prevail in any such litigation, one of the potential remedies would be the return of BCM to BioLine. In addition, if BioLine were to prevail in any such litigation, or if we were required to pay the milestone in dispute sooner than we had planned, or if we were required to return BCM to BioLine, these events could have a material adverse effect on our business, results of operations, financial condition and/or liquidity.

MANAGEMENT

The following table sets forth the name, age and position of each of our executive officers and directors as of May 12, 2014.

Name	Age	Position
Daniel Tassé	54	Chief Executive Officer, President and Director
Manesh Naidu	44	Vice President and Chief Business Officer
Reinilde Heyrman, M.D.	53	Vice President, Chief Clinical Development Officer and Secretary
Martin Meglasson, Ph.D.	64	Vice President and Chief Scientific Officer
David Abrams	39	Treasurer
Aldo E. Belloni, Ph.D.	64	Director
Matthew Holt	37	Director
Andre V. Moura	32	Director
Robert T. Nelsen	51	Director
Howard Pien	56	Director
Adam B. Weinstein	35	Director

(1) Member of the Audit Committee

(2) Member of the Compensation Committee

(3) Member of the Nominating and Corporate Governance Committee

Daniel Tassé has served as our President and Chief Executive Officer and as a member of our board of directors since February 2014. Since January 2008, Mr. Tassé has served as President and Chief Executive Officer and as a member of the board of directors of Ikaria, a biotherapeutics company. Mr. Tassé was appointed chairman of Ikaria's board of directors in October 2009. From October 2004 to January 2008, Mr. Tassé served as General Manager of the Pharmaceuticals and Technologies Business Unit of Baxter International, Inc., a global diversified healthcare company. From July 2001 to October 2004, Mr. Tassé served as Vice President and Regional Director for Australasia at GlaxoSmithKline, a healthcare company. Mr. Tassé is a member of the Healthcare Leadership Council and a member of the board of directors of the Roundtable on Critical Care Policy. He also is a member of the board of directors and health section governing board of the Biotechnology Industry Organization, where he participates on the bioethics, regulatory environment and reimbursement committees. Additionally, Mr. Tassé is a member of the board of directors of the Pharmaceutical Research and Manufacturers Association of America, where he participates on the FDA and biomedical research committee. Mr. Tassé received a B.S. in biochemistry from the University of Montreal. As our President and Chief Executive Officer, Mr. Tassé provides a critical contribution to our board of directors as a result of his extensive track record of business building in the healthcare industry, his strong background within critical care, his global management experience, and his detailed knowledge of the pharmaceutical industry, our company, employees, client base and competitors.

Manesh Naidu has served as our Vice President and Chief Business Officer since February 2014. Mr. Naidu previously served as Vice President and General Manager of the INOpulse program of Ikaria from August 2011 to February 2014, and prior to that, he served as Senior Director, Marketing Strategy of Ikaria from May 2008 to August 2011. Prior to joining Ikaria, Mr. Naidu held several positions at Novartis Corporation and Pfizer Inc., both of which are pharmaceutical companies, from 2003 to 2008. He also worked at McKinsey & Company, a global management consulting firm, from 2001 to 2003. Mr. Naidu received an M.S. in chemical engineering from Oklahoma State University, a B.E. in chemical engineering and an M.S. in chemistry both from the Birla Institute of Technology and Science, and an M.B.A. from the Kellogg School of Management at Northwestern University.

Reinilde Heyrman, M.D. has served as our Vice President, Chief Clinical Development Officer and Secretary since February 2014. Prior to joining us, Dr. Heyrman served as Vice President, Chief Clinical Development Officer of Ikaria from March 2012 to February 2014. Dr. Heyrman held several positions at Daiichi Sankyo Pharma Development, a pharmaceutical company, from 2005 to March 2012, most recently as Vice President, Clinical Development. From 2001 to 2002 and 2002 to 2005, Dr. Heyrman served as Director Clinical Research and Senior Director Clinical Research, respectively, at Sankyo Pharma Development, a pharmaceutical company. Dr. Heyrman received an M.D. from the University of Antwerp, Belgium.

Martin Meglasson, Ph.D. has served as our Vice President and Chief Scientific Officer since February 2014. From July 2010 to February 2014, Dr. Meglasson served as Chief Scientific Officer of Ikaria. Prior to joining Ikaria, Dr. Meglasson served as Vice President, Head of Research and Development of Ligand Pharmaceuticals Incorporated, a biotechnology company, from February 2004 to July 2010. Dr. Meglasson received a B.S. in biology, an M.S. in physiology and a Ph.D. in pharmacology, each from the University of Houston.

David Abrams has been our Treasurer since February 2014, with responsibilities for treasury, financial planning and financial reporting. Prior to joining us, Mr. Abrams held various roles in strategic financial planning at Ikaria from October 2010 to February 2014 and at Johnson & Johnson, a healthcare products company, from May 2002 to October 2010. Mr. Abrams has previously held roles at Stern Stewart and Deutsche Bank. Mr. Abrams received a B.S. in economics from The Wharton School of Business of the University of Pennsylvania and a B.A. in history from the University of Pennsylvania.

Aldo E. Belloni, Ph.D. has served as a member of our board of directors since February 2014. Since 1980, Dr. Belloni has held several positions with Linde AG, a world leading supplier of industrial, process and speciality gases and engineering company, and its subsidiaries, and has served as a Member of the Executive Board of Linde AG since January 2000. Dr. Belloni received a Ph.D. in chemical engineering from the Milan Polytechnic Institute. We believe that Dr. Belloni is qualified to serve on our board of directors because of his extensive strategic and executive level experience, including as a member of the Executive Board of Linde AG.

Matthew Holt has served as a member of our board of directors since February 2014. Since 2001, Mr. Holt has been employed by New Mountain Capital, a private equity group, where he currently serves as a Managing Director. Prior to joining New Mountain Capital, Mr. Holt served in the mergers and acquisitions Group at Lehman Brothers, a financial services firm. Mr. Holt has served on the board of directors of Ikaria since March 2007. Mr. Holt received an A.B. in English and American literature and language from Harvard College. We believe that Mr. Holt is qualified to serve on our board of directors because of his financial expertise and his years of experience providing strategic advisory services across many industries.

Andre V. Moura has served as a member of our board of directors since February 2014. Mr. Moura joined New Mountain Capital in 2005, where he currently serves as a Director. Prior to joining New Mountain Capital, Mr. Moura was employed by McKinsey & Company, a global management consulting firm. Mr. Moura also serves on the board of directors of two privately held companies. Mr. Moura received an A.B. in computer science from Harvard College and an M.B.A. from Harvard Business School. We believe that Mr. Moura is qualified to serve on our board of directors because of his financial expertise and his years of experience providing strategic advisory services to diverse companies across multiple industries.

Robert T. Nelsen has served as a member of our board of directors since February 2014. Since 1986, Mr. Nelsen has served as a Co-Founder and Managing Director of ARCH Venture Partners, a venture capital firm focused on early-stage technology companies. Mr. Nelsen currently serves as a director of Agios Pharmaceuticals, Inc., Fate Therapeutics, Inc. and Kythera Biopharmaceuticals, Inc., each a

publicly traded biopharmaceutical company. Mr. Nelsen previously served as a director of Adolor Corporation, Array BioPharma Inc., Illumina, Inc., NeurogesX, Inc., Receptos, Inc. and Trubien Pharmaceuticals, Inc., each a biopharmaceutical company. Mr. Nelsen also serves on the board of several privately held companies, including Sage Therapeutics and Sapphire Energy Corporation. Mr. Nelsen received a B.S. from the University of Puget Sound, with majors in biology and economics, and an M.B.A. from the University of Chicago Graduate School of Business. We believe that Mr. Nelsen is qualified to serve on our board of directors because of his extensive experience with pharmaceutical companies, his financial expertise and his years of experience providing strategic and financial advisory services to pharmaceutical and biotechnology organizations, including evaluating business plans involving clinical trials.

Howard Pien has served as a member of our board of directors since February 2014. From May 2007 to September 2009, Mr. Pien served as the Chief Executive Officer of Medarex, Inc., a biotechnology company, or Medarex, until Medarex was acquired by Bristol-Myers Squibb. Mr. Pien served as the President and Chief Executive Officer and as a director of Chiron Corporation, a biopharmaceutical company, from April 2003 until Chiron Corporation's merger with Novartis AG in May 2006. Mr. Pien was chairman of the board of directors of each of Chiron Corporation and Medarex. From December 2000 to March 2003, Mr. Pien worked at GlaxoSmithKline, a healthcare company, where he held several positions in GlaxoSmithKline's worldwide pharmaceuticals business, including President, Pharmaceuticals International. Mr. Pien previously held key positions in SmithKline Beecham's pharmaceuticals business in the United States, the United Kingdom and North Asia, culminating in his tenure as President, Pharmaceuticals-North America. Prior to joining SmithKline Beecham, he worked for six years at Abbott Laboratories and for five years at Merck & Co. Currently, Mr. Pien serves on the boards of directors of the following companies: Vanda Pharmaceuticals Inc., a publicly traded drug development company; ImmunoGen, Inc., a publicly traded company engaged in anticancer therapeutic development; Juno Therapeutics, a development stage company focused on immunotherapy, and Sage Therapeutics, a development stage company focused on the central nervous system. He is also an advisor to Warburg Pincus, a private equity firm. Mr. Pien previously served on the boards of directors of ViroPharma Incorporated, a pharmaceutical development company, Talon Therapeutics, Inc., a biopharmaceutical company, and Medarex. Mr. Pien received a B.S. from the Massachusetts Institute of Technology and an M.B.A. from Carnegie-Mellon University. We believe that Mr. Pien is qualified to serve on our board of directors because of his extensive experience in the pharmaceutical industry, including his management expertise as a chief executive officer of pharmaceutical and biotechnology companies and his extensive corporate governance expertise as a director of private and public companies.

Adam B. Weinstein has served as a member of our board of directors since February 2014. He is a Managing Director of New Mountain Capital, LLC, and he joined that organization in 2005. At New Mountain, Mr. Weinstein serves as a Chief Financial Officer and is an Executive Vice President and is on the Board of Directors of New Mountain Finance Corporation, a publicly traded business development company. Prior to joining New Mountain, Mr. Weinstein held roles in the mergers and acquisitions and private equity investor services areas of Deloitte & Touche, LLP, in that firm's merger and acquisition and private equity investor services areas. Mr. Weinstein is a New York State Certified Public Accountant and received his B.S., summa cum laude, in accounting from Binghamton University. We believe that Mr. Weinstein is qualified to serve on our board of directors because of his financial and accounting expertise and valuable corporate governance experience.

Board Composition and Election of Directors

Our business and affairs are currently managed under our limited liability company board of directors, which consists of seven members. Upon the Company Conversion, the members of our

limited liability company board of directors will become our board of directors, and we refer to them as such.

Following the completion of this offering, we expect that, pursuant to a voting agreement, (i) the New Mountain Entities will be entitled to designate (a) three directors, for so long as they beneficially own 15% or more of our outstanding common stock, (b) two directors, for so long as they beneficially own less than 15% but more than 5% of our outstanding common stock and (c) one director, for so long as they beneficially own less than 5% of our outstanding common stock but more than one share of our common stock and (ii) each of ARCH, Venrock and Linde will be entitled to elect one director for so long as such holder beneficially owns 5% or more of our outstanding common stock. Messrs. Holt, Moura and Weinstein were designated as New Mountain directors by the New Mountain Entities, Mr. Nelsen was designated as an ARCH director by ARCH and Dr. Belloni was designated as a Linde director by Linde. Under the terms of the voting agreement, Venrock is entitled to designate a Venrock director to one of the open seats on our board of directors. We expect that Venrock will designate a director prior to the consummation of this offering. See "Certain Relationships and Related Person Transactions—Agreements with the Controlling Entities—Voting Agreement."

We will be deemed to be a "controlled company" under the rules established by The NASDAQ Global Market, and we will qualify for, and intend to rely on, the "controlled company" exception to the board of directors and committee composition requirements under the rules of The NASDAQ Global Market. Pursuant to this exception, we will be exempt from the rule that requires our board of directors to be comprised of a majority of "independent directors" and our compensation and nominating and corporate governance committees to be comprised solely of "independent directors," as defined under the rules of The NASDAQ Global Market. The "controlled company" exception does not modify the independence requirements for the audit committee, and we intend to comply with the requirements of the Sarbanes-Oxley Act and The NASDAQ Global Market rules, which require that our audit committee be composed of at least three members, each of whom will be independent within one year from the date of this prospectus.

Under applicable NASDAQ rules, a director will only qualify as an "independent director" if, in the opinion of our board of directors, that person does not have a relationship that would interfere with the exercise of independent judgment in carrying out the responsibilities of a director. Our board of directors intends to review its composition, the composition of its committees and the independence of each director according to the independence standards established by applicable SEC rules and NASDAQ rules. In making an independence determination, our board of directors will consider the relationships that each such non-employee director has with our company and all other facts and circumstances that our board of directors deems relevant in determining their independence, including the beneficial ownership of our capital stock by each non-employee director.

There are no family relationships among any of our directors or executive officers.

Following the closing of this offering, in accordance with the terms of our certificate of incorporation and bylaws that will become effective as of the closing of this offering, our board of directors will be divided into three classes, class I, class II and class III, with members of each class serving staggered three-year terms. Upon the closing of this offering, the members of the classes will be divided as follows:

- the class I directors are , and their term expires at our annual meeting of stockholders to be held in 2015;
- the class II directors are , and their term expires at our annual meeting of stockholders to be held in 2016; and
- the class III directors are , and their term expires at our annual meeting of stockholders to be held in 2017.

Upon the expiration of the term of a class of directors, directors in that class are eligible to be elected for a new three-year term at the annual meeting of stockholders in the year in which their term expires. Prior to the first time that the New Mountain Entities beneficially own less than 15% of our outstanding common stock, any director may be removed, with or without cause, by the affirmative vote of the holders of a majority of the outstanding shares of our common stock, provided that any director elected by the New Mountain Entities, ARCH, Venrock or Linde may only be removed without cause by the affirmative vote of a majority of the outstanding shares held by the applicable designating entity. After the New Mountain Entities beneficially own less than 15% of our outstanding common stock, directors may be removed only for cause and only by holders of at least 75% of the outstanding shares of our common stock, provided that any director elected by the New Mountain Entities, ARCH, Venrock or Linde may also be removed without cause by the affirmative vote of a majority of the outstanding shares held by the applicable designating entity. See "Description of Capital Stock—Delaware Anti-Takeover Law and Certain Charter and Bylaw Provisions—Staggered Board; Removal of Directors."

Board Committees

Our board of directors has established an audit committee, a compensation committee and a nominating and corporate governance committee, each of which will operate, as of the date of this prospectus, under a charter that has been approved by our board. The composition of each committee will be effective upon the closing of this offering. Pursuant to the terms of our stockholders agreement, until we are no longer a "controlled company" under the rules established by The NASDAQ Global Market, if any other committee is established, the members will include at least one director elected by the New Mountain Entities, if any, and either the director elected by ARCH or the director elected by Venrock, if any.

Audit Committee

The members of our audit committee are . chairs our audit committee. Our audit committee's responsibilities will include:

- appointing, approving the compensation of, and assessing the independence of our registered public accounting firm;
- overseeing the work of our independent registered public accounting firm, including through the receipt and consideration of reports from such firm;
- reviewing and discussing with management and our independent registered public accounting firm our annual and quarterly financial statements and related disclosures;
- monitoring our internal control over financial reporting, disclosure controls and procedures and code of business conduct and ethics;
- overseeing our internal audit function;
- overseeing our risk assessment and risk management policies;
- establishing policies regarding hiring employees from our independent registered public accounting firm and procedures for the receipt and retention of accounting related complaints and concerns;
- meeting independently with our internal auditing staff, our independent registered public accounting firm and management;

- reviewing and approving or ratifying any related person transactions; and
- preparing the audit committee report required by Securities and Exchange Commission, or SEC, rules.

All audit and non-audit services, other than *de minimis* non-audit services, to be provided to us by our independent registered public accounting firm must be approved in advance by our audit committee.

Our board of directors has determined that is an "audit committee financial expert" as defined in applicable SEC rules. We believe that the composition of our audit committee meets the requirements for independence under current NASDAQ and SEC rules and regulations.

Compensation Committee

The members of our compensation committee are . chairs our compensation committee. Pursuant to the terms of the stockholders agreement, until we are no longer a "controlled company" under the rules established by The NASDAQ Global Market, the chair of our compensation committee will be a director designated by the New Mountain Entities, if any, and the members of our compensation committee will include either a director designated by ARCH or Venrock, if any. See "Certain Relationships and Related Person Transactions—Agreements with the Controlling Entities—Stockholders Agreement." Our compensation committee's responsibilities will include:

- reviewing and approving, or making recommendations to our board with respect to, the compensation of our chief executive officer and our other executive officers;
- overseeing an evaluation of our senior executives;
- overseeing and administering our cash and equity incentive plans;
- reviewing and making recommendations to our board with respect to director compensation;
- reviewing and discussing annually with management our compensation disclosure required by SEC rules; and
- preparing the compensation committee report required by SEC rules.

Nominating and Corporate Governance Committee

The members of our nominating and corporate governance committee are . chairs our nominating and corporate governance committee. Pursuant to the terms of the stockholders agreement, until we are no longer a "controlled company" under the rules established by The NASDAQ Global Market, the members of our nominating and corporate governance committee will include the director designated by ARCH, if any, the director designated by Venrock, if any, and more than one director designated by the New Mountain Entities, if two or more directors designated by the New Mountain Entities are members of our board, or the sole director designated by the New Mountain Entities, if only one such director is a member of our board. See "Certain Relationships and Related Person Transactions—Agreements with the Controlling Entities—Stockholders Agreement." Our nominating and corporate governance committee's responsibilities will include:

- identifying individuals qualified to become members of our board;
- recommending to our board the persons to be nominated for election as directors and to each of our board's committees;
- reviewing and making recommendations to our board with respect to our board leadership structure;

- reviewing and making recommendations to our board with respect to management succession planning;
- developing and recommending to our board corporate governance principles; and
- overseeing a periodic evaluation of our board.

Compensation Committee Interlocks and Insider Participation

None of our executive officers serves as a member of the board of directors or compensation committee, or other committee serving an equivalent function, of any other entity that has one or more of its executive officers serving as a member of our board of directors or our compensation committee. None of the members of our compensation committee is, or has ever been, an officer or employee of our company.

EXECUTIVE COMPENSATION

Overview

We were formed on October 17, 2013 as a subsidiary of Ikaria and we became an independent, stand-alone operating company as a result of the Spin-Out on February 12, 2014. Because the costs and liabilities with respect to compensation of our employees for the fiscal year ended December 31, 2013 and for prior periods were paid by Ikaria on the basis of criteria and methodology not relevant to us and work performed with respect to businesses in addition to ours, we are not presenting compensation information for historical periods.

In preparing to become a stand-alone public company, we have begun a thorough review of all elements of our executive compensation program, including the function and design of our equity incentive programs. We have begun, and expect to continue in the coming months, to evaluate the need for revisions to our executive compensation program to ensure that our program is competitive with the companies with which we compete for executive talent and is appropriate for a public company. As we gain experience as a stand-alone, public company, we expect that the specific direction, emphasis and components of our executive compensation program will continue to evolve. Moving forward, our compensation committee will review and approve the compensation of our executive officers and oversee and administer our executive compensation programs and initiatives.

Compensation Components

This section describes the material elements of compensation of each of our executive officers: Daniel Tassé, Chief Executive Officer; Manesh Naidu, Vice President and Chief Business Officer; Reinilde Heyrman, Vice President, Chief Clinical Development Officer and Secretary; Martin Meglasson, Vice President and Chief Scientific Officer; and David Abrams, Treasurer. On the basis of the anticipated compensation of our executive officers for 2014, described in more detail below, we expect that our "named executive officers" will be Mr. Tassé, Dr. Heyrman and Dr. Meglasson.

Base Salary. We will pay our executive officers the following annualized base salaries for the year ending December 31, 2014: $0 to Mr. Tassé, $244,600 to Mr. Naidu, $433,500 to Dr. Heyrman, $363,000 to Dr. Meglasson and $179,500 to Mr. Abrams. Base salaries are used to recognize the experience, skills, knowledge and responsibilities required of all of our employees, including our executive officers. In determining the base salaries of our executive officers, we did not engage in any form of benchmarking in the determination of base salaries of our executive officers. Following the closing of this offering, our compensation committee will review the salaries of our executives annually at the beginning of each calendar year and recommend to our board of directors changes in salaries based primarily on changes in job responsibilities, experience, individual performance, and comparative market data.

Bonus Compensation. Our executive officers are expected to be eligible to receive an annual cash bonus award in accordance with the management incentive program then in effect with respect to such executive officer and based on an annualized target of base salary. Our executive officers are also expected to be eligible for performance-based annual bonus awards based on metrics to be determined by our board of directors, in consultation with the executive officer, and our board of directors will determine the extent to which the metrics have been satisfied and the amount of the annual bonus, if any. The performance-based bonuses are designed to motivate our employees to achieve annual goals based on our strategic, financial and operating performance objectives.

Long-Term Equity Based Incentive Awards. We believe that equity grants provide our executives with a strong link to our long-term performance, create an ownership culture and help to align the interests of our executives and our stockholders. In addition, we believe that equity grants with a time-based vesting feature promote executive retention because this feature incentivizes our executive

officers to remain in our employment during the vesting period. Accordingly, our compensation committee and board of directors periodically review the equity incentive compensation of our named executive officers and from time to time may grant additional equity incentive awards to them in the form of stock options.

Employment Agreements with Our Executive Officers

We have written employment agreements with Mr. Naidu, Dr. Heyrman and Dr. Meglasson. Each agreement provides for an employment term of one year, with the term automatically renewing for successive one-year terms, unless we or the applicable officer gives written notice of non-renewal at least 90 days prior to the renewal date. Each of these officers is subject to confidentiality, invention assignment, non-disclosure, non-competition and non-solicitation agreements.

In addition, following the end of each calendar year, each executive officer named below is eligible to receive an annual bonus based on the achievement of individual and company performance objectives, which will be determined by our board of directors in its sole discretion. The bonus is calculated as a percentage of the executive's annual base salary. As of the date of this prospectus, the target bonus percentage for each such executive officer is as follows: Mr. Naidu 35%; Dr. Heyrman 40%; and Dr. Meglasson 40%.

We do not have employment agreements with Mr. Tassé or Mr. Abrams.

Potential Payments Upon Termination or Change in Control

Each of Mr. Naidu, Dr. Heyrman and Dr. Meglasson are entitled to severance payments if his or her employment is terminated under specified circumstances.

Manesh Naidu. If we terminate Mr. Naidu's employment without cause, if Mr. Naidu terminates his employment with us for good reason (as defined in the employment agreement) or if Mr. Naidu terminates his employment at the end of a term following delivery by us of notice that we will not extend the term, Mr. Naidu is entitled to receive: a lump sum payment in an amount equal to earned but unpaid base salary and annual bonus and any accrued but unpaid vacation time; a payment equal to the sum of (i) Mr. Naidu's annual base salary and (ii) the greater of his applicable annual bonus target and the actual annual bonus actually paid to Mr. Naidu, determined on a monthly basis for a period of 12 months immediately following the date of termination; and continued coverage, at our expense, under our medical, dental and vision benefit plans for 12 months immediately following the date of termination.

In the event that we terminate Mr. Naidu's employment without cause, Mr. Naidu terminates his employment with us for good reason or Mr. Naidu terminates his employment at the end of a term following delivery by us of notice that we will not extend the term, in each case within 12 months of the occurrence of a change of control, any equity compensation granted to Mr. Naidu shall become fully vested as of the date of termination.

Dr. Reinilde Heyrman. If we terminate Dr. Heyrman's employment without cause, if Dr. Heyrman terminates her employment with us for good reason (as defined in the employment agreement) or if Dr. Heyrman terminates her employment at the end of a term following delivery by us of notice that we will not extend the term, Dr. Heyrman is entitled to receive: a lump sum payment in an amount equal to earned but unpaid base salary and annual bonus and any accrued but unpaid vacation time; a payment equal to the sum of (i) Dr. Heyrman's annual base salary and (ii) the greater of her applicable annual bonus target and the actual annual bonus actually paid to Dr. Heyrman, determined on a monthly basis for a period of 12 months immediately following the date of termination; and continued coverage, at our expense, under our medical, dental and vision benefit plans for 12 months immediately following the date of termination.

In the event that we terminate Dr. Heyrman's employment without cause, Dr. Heyrman terminates her employment with us for good reason, or Dr. Heyrman terminates her employment at the end of a term following delivery by us of notice that we will not extend the term, in each case within 12 months of the occurrence of a change of control, any equity compensation granted to Dr. Heyrman shall become fully vested as of the date of termination.

Dr. Martin Meglasson. If we terminate Dr. Meglasson's employment without cause, if Dr. Meglasson terminates his employment with us for good reason (as defined in the employment agreement) or if Dr. Meglasson terminates his employment at the end of a term following delivery by us of notice that we will not extend the term, Dr. Meglasson is entitled to receive: a lump sum payment in an amount equal to earned but unpaid base salary and annual bonus and any accrued but unpaid vacation time; a pro-rated portion of his annual bonus target for the year in which his employment terminates; payments equal to the sum of (i) Dr. Meglasson's annual base salary and (ii) the greater of his applicable annual bonus target and the actual annual bonus actually paid to Dr. Meglasson, determined on a monthly basis for a period of 12 months immediately following the date of termination; and continued coverage, at our expense, under our medical, dental and vision benefit plans for 12 months immediately following the date of termination.

In the event that we terminate Dr. Meglasson's employment without cause, Dr. Meglasson terminates his employment with us for good reason or Dr. Meglasson terminates his employment at the end of a term following delivery by us of notice that we will not extend the term, in each case within 18 months of the occurrence of a change of control, Dr. Meglasson is entitled to receive: a lump sum payment in an amount equal to 50% of the sum of (i) Dr. Meglasson's annual base salary and (ii) the greater of his target annual bonus and the actual annual bonus paid to Dr. Meglasson; an additional six months of continued coverage, at our expense, under our medical, dental and vision benefit plans; and the unvested portion of any equity compensation granted to Dr. Meglasson shall become immediately fully vested.

Retention Bonus Agreements with Our Executive Officers

On February 3, 2014, we delivered letters to each of Mr. Naidu, Dr. Heyrman, Dr. Meglasson and Mr. Abrams offering them a one-time "retention bonus" payment if Mr. Naidu, Dr. Heyrman, Dr. Meglasson or Mr. Abrams, respectively, remains an active employee of Bellerophon in good standing through December 19, 2014. The bonus would be payable within 30 days after December 19, 2014. The retention bonus payments payable are in the following amounts, less applicable taxes: $150,000 to Mr. Naidu, $150,000 to Dr. Heyrman, $150,000 to Dr. Meglasson and $15,000 to Mr. Abrams.

Equity Ownership of Our Executive Officers

In connection with the Spin-Out, Ikaria distributed our voting units to its stockholders through the payment of a special dividend to its stockholders on a pro rata basis based on each stockholder's ownership of Ikaria capital stock. Prior to the Spin-Out, we issued to certain employees and directors of ours and of our then-parent company, Ikaria, including certain of our executive officers, and certain accredited investors options to purchase the same number of our non-voting membership units as the number of shares of non-voting Ikaria stock subject to the Ikaria options then held by such employee, director or accredited investor at such time. Prior to the Spin-Out, we issued to certain employees and directors of ours or of Ikaria, including certain of our executive officers, and certain accredited investors the same number of restricted stock units in respect of our non-voting membership units, which we refer to as the Bellerophon RSUs, as the number of shares of non-voting Ikaria stock held by such employee, director or accredited investor at such time. We subsequently settled such Bellerophon RSUs by issuing and delivering non-voting units to the holders of Bellerophon RSUs.

The below table summarizes the outstanding equity ownership of our executive officers as of April 30, 2014, after giving effect to the Company Conversion and the conversion of non-voting shares into voting shares upon the closing of this offering.

Name	Shares	Shares Subject to Options
Daniel Tassé	1,301,400	1,200,000
Manesh Naidu	123,000	78,000
Reinilde Heyrman	100,000	—
Martin Meglasson	80,000	100,000
David Abrams	—	20,000
All executive officers as a group	1,604,400	1,398,000

Stock Option and Other Compensation Plans

The four equity incentive plans described in this section are (i) the assumed 2007 Ikaria stock option plan, which we refer to as the 2007 Ikaria plan, (ii) the assumed Ikaria 2010 long term incentive plan, which we refer to as the 2010 Ikaria plan, (iii) our 2014 equity incentive plan, which we refer to as the 2014 equity plan and (iv) our public company stock incentive plan, which we refer to as the public company incentive plan. Following the closing of this offering, we expect to grant awards to eligible participants under the 2014 equity plan and public company incentive plan.

Assumed 2007 Ikaria Plan

The 2007 Ikaria plan was adopted by Ikaria in March 2007 and we assumed the terms of the 2007 Ikaria plan in connection with the Spin-Out. Stock options granted under the 2007 Ikaria plan have a contractual life of ten years. Pursuant to the terms of the 2007 Ikaria plan, in the event of a liquidation or dissolution of our company, each outstanding option under the 2007 Ikaria plan will terminate immediately prior to the consummation of the action, unless the administrator determines otherwise. In the event of a merger or other reorganization event, each outstanding option will be assumed or an equivalent option or right will be substituted by the successor entity, unless such successor entity does not agree to assume the award or to substitute an equivalent option or right in which case such option will terminate upon the consummation of the merger or reorganization event.

Assumed 2010 Ikaria Plan

The 2010 Ikaria plan was adopted by Ikaria in February 2010 and amended and restated in May 2010, and and we assumed the terms of the 2010 Ikaria plan in connection with the Spin-Out. Pursuant to the terms of the 2010 Ikaria plan, upon our liquidation, dissolution, merger or consolidation, except as otherwise provided in an applicable option or award agreement, each option or award will be (i) treated as provided in the agreement related to the transaction, or (ii) if not so provided in such agreement, each holder of an option or award will be entitled to receive, in respect of each share subject to outstanding options or awards, the same number of stock, securities, cash, property or other consideration that he or she would have received had he or she exercised such options or awards prior to the transaction. The stock, securities, cash, property or other consideration shall remain subject to all of the conditions, restrictions and performance criteria which were applicable to the options and awards prior to any such transaction. If the consideration paid or distributed is not entirely shares of common stock of the acquiring or resulting corporation, the treatment of outstanding options and stock appreciation rights may include the cancellation of outstanding options and stock appreciation rights

upon consummation of the transaction as long as the holders of affected options and stock appreciation rights, at the election of the compensation committee, either:

- have been given a period of at least 15 days prior to the date of the consummation of the transaction to exercise the options or stock appreciation rights (whether or not they were otherwise exercisable); or

- are paid (in cash or cash equivalents) in respect of each share covered by the option or stock appreciation right being cancelled an amount equal to the excess, if any, of the per share price paid or distributed to stockholders in the transaction (the value of any non-cash consideration to be determined by the compensation committee in its sole discretion) over the exercise price of the option or stock appreciation right.

2014 Equity Incentive Plan

Our board of directors adopted and our stockholders approved, both in 2014, the 2014 equity plan. The 2014 equity plan is administered by our board of directors or by a committee appointed by our board of directors. The 2014 equity plan provides for the grant of options. Under the 2014 equity plan, there are shares of common stock available for the grant of options.

Our employees, officers, directors, consultants and advisors are eligible to receive awards under the 2014 equity plan. Subject to any limitation in the 2014 equity plan, our board of directors or any committee to which our board of directors has delegated authority will select the recipients of options and determine:

- the number of shares of common stock covered by options, the dates upon which those options become exercisable and the terms and conditions that apply to such options;

- the exercise price of options which may not be less than 100% of the fair market value of our common stock on the grant date;

- the duration of options, which may not be in excess of ten years;

- the methods of payment of the exercise price of options; and

- any amendments to the 2014 equity plan and/or any option agreement.

Our board of directors may exercise such powers and perform such acts as it deems necessary or expedient to promote the best interests of the Company which are not in conflict with the 2014 equity plan provisions.

Awards under the 2014 equity plan are subject to adjustment in the event of a split, reverse split, dividend, recapitalization, combination or reclassification of Company common stock, spin-off or other similar change in our capitalization, conversion of the Company into a corporation or other entity or event or any dividend or distribution to holders of our common stock other than an ordinary cash dividend.

Upon a merger or other reorganization event (as defined in the 2014 equity plan), our board of directors, may, in its sole discretion, take any one or more of the following actions pursuant to the 2014 equity plan, as to some or all outstanding options:

- provide that all outstanding options will be assumed, or substantially equivalent awards shall be substituted, by the acquiring or successor corporation or an affiliate thereof;

- upon written notice to a participant, provide that the participant's unvested and/or unexercised options will terminate immediately prior to the consummation of such transaction unless exercised by the participant;

- provide that outstanding options will become exercisable, realizable or deliverable, or restrictions applicable to an option will lapse, in whole or in part, prior to or upon the reorganization event;

- in the event of a reorganization event pursuant to which holders of shares of common stock will receive a cash payment for each share of common stock surrendered in the reorganization event, make or provide for a cash payment to the participants with respect to each option held by the participant equal to (1) the number of shares of common stock subject to the vested portion of the option, after giving effect to any acceleration of vesting that occurs upon or immediately prior to such reorganization event, multiplied by (2) the excess, if any, of the cash payment for each share of common stock surrendered in the reorganization event over the exercise price of such option and any applicable tax withholdings, in exchange for the termination of such option; and

- provide that, in connection with a liquidation or dissolution, options convert into the right to receive liquidation proceeds.

At any time, our board of directors may, in its sole discretion, provide that any award under the 2014 equity plan will become immediately exercisable in full or in part, free of some or all restrictions or conditions, or otherwise realizable in full or in part.

No option may be granted under the 2014 equity plan after , 2024. Our board of directors may amend, suspend or terminate the 2014 equity plan at any time, except that stockholder approval will be required to comply with applicable law or stock market requirements.

Public Company Stock Incentive Plan

We expect our board of directors to adopt and our stockholders to approve the public company incentive plan, which will become effective immediately prior to the closing of this offering. The public company incentive plan provides for the grant of incentive stock options, nonstatutory stock options, stock appreciation rights, restricted stock awards, restricted stock unit awards and other stock-based awards. Upon effectiveness of the public company incentive plan, the number of shares of our common stock that will be reserved for issuance under the public company incentive plan will be the sum of (1) plus (2) the number of shares (up to shares) equal to the sum of the number of shares of our common stock then available for issuance under the 2014 equity plan and the number of shares of our common stock subject to outstanding awards under the 2014 equity plan that expire, terminate or are otherwise surrendered, cancelled, forfeited or repurchased by us at their original issuance price pursuant to a contractual repurchase right plus (3) an annual increase, to be added on the first day of each fiscal year, beginning with the fiscal year ending December 31, and continuing until, and including, the fiscal year ending December 31, , equal to the lowest of shares of our common stock, % of the number of shares of our common stock outstanding on the first day of the fiscal year and an amount determined by our board of directors.

Our employees, officers, directors, consultants and advisors are eligible to receive awards under the public company incentive plan. However, incentive stock options may only be granted to our employees. The maximum number of shares of our common stock with respect to which awards may be granted to any participant under the public company incentive plan is per calendar year. For purposes of this limit on the maximum number of shares that may be awarded to any participant, the combination of an option in tandem with a stock appreciation right will be treated as a single award.

Pursuant to the terms of the public company incentive plan, our board of directors (or a committee delegated by our board of directors) administers the plan and, subject to any limitations in the plan, selects the recipients of awards and determines:

- the number of shares of our common stock covered by options and the dates upon which the options become exercisable;

- the type of options to be granted;
- the duration of options, which may not be in excess of ten years;
- the exercise price of options, which must be at least equal to the fair market value of our common stock on the date of grant; and
- the number of shares of our common stock subject to and the terms of any stock appreciation rights, restricted stock awards, restricted stock units or other stock-based awards and the terms and conditions of such awards, including conditions for repurchase, issue price and repurchase price (though the measurement price of stock appreciation rights must be at least equal to the fair market value of our common stock on the date of grant and the duration of such awards may not be in excess of ten years).

If our board of directors delegates authority to an executive officer to grant awards under the public company incentive plan, the executive officer has the power to make awards to all of our employees, except executive officers. Our board of directors will fix the terms of the awards to be granted by such executive officer, including the exercise price of such awards (which may include a formula by which the exercise will be determined), and the maximum number of shares subject to awards that such executive officer may make.

Upon a merger or other reorganization event, our board of directors may, in its sole discretion, take any one or more of the following actions pursuant to the public company incentive plan as to some or all outstanding awards other than restricted stock:

- provide that all outstanding awards shall be assumed, or substantially equivalent awards shall be substituted, by the acquiring or successor corporation (or an affiliate thereof);
- upon written notice to a participant, provide that all of the participant's unexercised awards will terminate immediately prior to the consummation of such reorganization event unless exercised by the participant;
- provide that outstanding awards shall become exercisable, realizable or deliverable, or restrictions applicable to an award shall lapse, in whole or in part, prior to or upon such reorganization event;
- in the event of a reorganization event pursuant to which holders of shares of our common stock will receive a cash payment for each share surrendered in the reorganization event, make or provide for a cash payment to the participants with respect to each award held by a participant equal to (1) the number of shares of our common stock subject to the vested portion of the award (after giving effect to any acceleration of vesting that occurs upon or immediately prior to such reorganization event) multiplied by (2) the excess, if any, of the cash payment for each share surrendered in the reorganization event over the exercise, measurement or purchase price of such award and any applicable tax withholdings, in exchange for the termination of such award; and/or
- provide that, in connection with a liquidation or dissolution, awards shall convert into the right to receive liquidation proceeds (if applicable, net of the exercise, measurement or purchase price thereof and any applicable tax withholdings).

Our board of directors does not need to take the same action with respect to all awards and may take different actions with respect to portions of the same award.

In the case of certain restricted stock units, no assumption or substitution is permitted, and the restricted stock units will instead be settled in accordance with the terms of the applicable restricted stock unit agreement.

Upon the occurrence of a reorganization event other than a liquidation or dissolution, the repurchase and other rights with respect to outstanding restricted stock awards will continue for the benefit of the successor company and will, unless the board of directors may otherwise determine, apply to the cash, securities or other property into which shares of our common stock are converted or exchanged pursuant to the reorganization event. Upon the occurrence of a reorganization event involving a liquidation or dissolution, all restrictions and conditions on each outstanding restricted stock award will automatically be deemed terminated or satisfied, unless otherwise provided in the agreement evidencing the restricted stock award.

At any time, our board of directors may, in its sole discretion, provide that any award under the public company incentive plan will become immediately exercisable in full or in part, free of some or all restrictions or conditions, or otherwise realizable in full or in part.

No award may be granted under the public company incentive plan on or after , 2024. Our board of directors may amend, suspend or terminate the public company incentive plan at any time, except that stockholder approval may be required to comply with applicable law or stock market requirements.

401(k) Retirement Plan

We maintain a 401(k) retirement plan that is intended to be a tax-qualified defined contribution plan under Section 401(k) of the Internal Revenue Code. In general, all of our employees are eligible to participate, beginning on the first day of the month following commencement of their employment. The 401(k) plan includes a salary deferral arrangement pursuant to which participants may elect to reduce their current compensation by up to the statutorily prescribed limit, equal to $17,500 in 2014, and have the amount of the reduction contributed to the 401(k) plan.

Limitations on Liability and Indemnification

Our certificate of incorporation, which will become effective upon the closing of this offering, limits the personal liability of directors for breach of fiduciary duty to the maximum extent permitted by the Delaware General Corporation Law and provides that no director will have personal liability to us or to our stockholders for monetary damages for breach of fiduciary duty or other duty as a director. However, these provisions do not eliminate or limit the liability of any of our directors:

- for any breach of the director's duty of loyalty to us or our stockholders;
- for acts or omissions not in good faith or that involve intentional misconduct or a knowing violation of law;
- for voting for or assenting to unlawful payments of dividends, stock repurchases or other distributions; or
- for any transaction from which the director derived an improper personal benefit.

Any amendment to or repeal of these provisions will not eliminate or reduce the effect of these provisions in respect of any act, omission or claim that occurred or arose prior to such amendment or repeal. If the Delaware General Corporation Law is amended to provide for further limitations on the personal liability of directors of corporations, then the personal liability of our directors will be further limited to the greatest extent permitted by the Delaware General Corporation Law.

In addition, our certificate of incorporation, which will become effective upon the closing of this offering, provides that we must indemnify our directors and officers and we must advance expenses, including attorneys' fees, to our directors and officers in connection with legal proceedings, subject to very limited exceptions.

We maintain a general liability insurance policy that covers certain liabilities of our directors and officers arising out of claims based on acts or omissions in their capacities as directors or officers. In addition, we have entered into indemnification agreements with our directors. These indemnification agreements may require us, among other things, to indemnify each such director for some expenses, including attorneys' fees, judgments, fines and settlement amounts incurred by him in any action or proceeding arising out of his service as one of our directors.

Certain of our non-employee directors may, through their relationships with their employers, be insured and/or indemnified against certain liabilities incurred in their capacity as members of our board of directors.

Rule 10b5-1 Sales Plans

Our directors and executive officers may adopt written plans, known as Rule 10b5-1 plans, in which they will contract with a broker to buy or sell shares of our common stock on a periodic basis. Under a Rule 10b5-1 plan, a broker executes trades pursuant to parameters established by the director or officer when entering into the plan, without further direction from the director or officer. The director or officer may amend or terminate the plan when not in possession of material, non-public information. In addition, our directors and executive officers may also buy or sell additional shares outside of a Rule 10b5-1 plan when they are not in possession of material, nonpublic information.

Director Compensation

Our board of directors intends to approve a compensation policy for our non-employee directors that will become effective upon the closing of this offering. This policy will be intended to provide a total compensation package that enables us to attract and retain qualified and experienced individuals to serve as directors and to align our directors' interests with those of our stockholders.

Prior to the closing of this offering, we are paying our independent director Howard Pien cash compensation in the amount of $80,000 per year.

CERTAIN RELATIONSHIPS AND RELATED PERSON TRANSACTIONS

Other than compensation arrangements, we describe below transactions and series of similar transactions during our last three fiscal years to which we were a party or will be a party, in which (i) the amounts involved exceeded or will exceed $120,000, and (ii) any of our directors, executive officers, or holders of more than 5% of our capital stock, or any member of the immediate family of the foregoing persons, had or will have a direct or indirect material interest. Compensation arrangements for our directors and executive officers are described elsewhere in this prospectus. We believe that all of these transactions were on terms as favorable as could have been obtained from unrelated third parties.

Indemnification Agreements

Our certificate of incorporation in effect upon the closing of this offering provides that we will indemnify our directors and officers to the fullest extent permitted by Delaware law. In addition, we intend to enter into indemnification agreements with each of our directors prior to the closing of this offering. See "Executive Compensation—Limitations on Liability and Indemnification" for additional information regarding these agreements.

Agreements with the Controlling Entities

Registration Rights

We are a party to a registration rights agreement with certain holders of our common stock, including our 5% stockholders and their affiliates and entities affiliated with our directors. The registration rights agreement provides these holders the right, following the completion of this offering, to demand that we file a registration statement or request that their shares be covered by a registration statement that we are otherwise filing. See "Description of Capital Stock—Registration Rights" for additional information regarding these registration rights.

Voting Agreement

We are party to a voting agreement with the Controlling Entities, which will remain in effect following this offering. The voting agreement provides that, among other things, each of the Controlling Entities is required to vote its shares to elect the designated individuals as members of our board of directors. In particular, following the completion of this offering, (i) the New Mountain Entities will be entitled to designate (a) three directors, for so long as they beneficially own 15% or more of our outstanding common stock, (b) two directors, for so long as they beneficially own less than 15% but more than 5% of our outstanding common stock and (c) one director, for so long as they beneficially own less than 5% of our outstanding common stock but more than one share of our common stock and (ii) each of ARCH, Venrock and Linde is entitled to designate one director for so long as such holder beneficially owns 5% or more of our outstanding common stock. If any member of our board of directors designated by the New Mountain Entities, ARCH, Venrock or Linde ceases to serve as a director for any reason, the resulting vacancy will be filled by a director nominated by the person or entity entitled to designate the director. In addition, each Controlling Entity has agreed to vote in favor of the removal of any designated director upon and only upon the written request of the person or entity entitled to designate such director. Each of the Controlled Entities has agreed (i) not to vote in favor of decreasing the size of our board of directors below the number of directors then in office or that would prevent any of the Controlled Entities from designating their respective directors and (ii) so long as the New Mountain Entities beneficially own more than 5% of our common stock, not to vote in favor of increasing the size of our board of directors without the prior written consent of the New Mountain Entities.

Pursuant to the terms of the voting agreement, following the completion of this offering and until we are no longer a "controlled company" under the rules established by The NASDAQ Global Market, (i) if any committee of our board of directors other than an audit committee, compensation committee or nominating and corporate governance committee is established, the members will include at least one director elected by the New Mountain Entities, if any, and either the director elected by ARCH or the director elected by Venrock, if any; (ii) the chair of our compensation committee will be a director elected by the New Mountain Entities, if any, and the members of our compensation committee will include either a director elected by ARCH or Venrock, if any; and (iii) the members of our nominating and corporate governance committee will include the director elected by ARCH, if any, the director elected by Venrock, if any, and more than one director elected by the New Mountain Entities, if two or more directors elected by the New Mountain Entities are members of our board, or the sole director elected by the New Mountain Entities, if only one such director is a member of our board.

Stockholders Agreement

We expect to enter into a stockholders agreement with the New Mountain Entities, which will remain in effect following this offering. The stockholders agreement will provide that, following the closing of the offering, we will be required to obtain the prior written approval of the New Mountain Entities to take certain actions, including, among other things, actions to:

- consolidate or merge into or with any other person, sell, lease or transfer all or substantially all of our assets or capital stock to another person or enter into any other similar business combination transaction, or effect a liquidation;
- authorize, issue, sell, offer for sale or solicit offers to buy any shares of our common stock or any convertible securities or any other equity or debt securities or rights to acquire any of our, or our subsidiaries' equity or debt securities, subject to certain exceptions, including among other things, the issuance under our stock incentive plan of grants that have been approved by our board of directors (or a board committee) and at least one director appointed by the New Mountain Entities;
- incur indebtedness or refinance any indebtedness;
- effect any stock dividend, stock split or other subdivision or combination of our capital stock or any other recapitalization;
- effect any redemption, retirement, purchase or other acquisition, directly or indirectly of our capital stock, subject to certain exceptions for repurchases from our employees, officers, directors, consultants or other persons performing services for us or our subsidiaries;
- hire or replace any of our chief executive officer, chief financial officer or our next two most senior executives (as determined by our board of directors), or materially amend the level or form of compensation or benefits payable to, or other compensation arrangements of, any such officer;
- pay or declare a dividend or distribution on any shares of our capital stock (other than dividends from a wholly-owned subsidiary to its parent company);
- amend, repeal or change any provision of our certificate of incorporation or bylaws; or
- agree or otherwise commit to do any of the foregoing (unless the commitment is conditioned on obtaining the approval of the New Mountain Entities).

These approval rights of the New Mountain Entities will terminate following the closing of the offering when the New Mountain Entities and their assignees beneficially own less than 15% of our outstanding common stock. Following this offering, we expect the New Mountain Entities to hold

% of our outstanding common stock (or % if the underwriters exercise in full their option to purchase additional shares from us).

Relationship with Ikaria

Prior to the Spin-Out on February 12, 2014, we were a wholly-owned subsidiary of Ikaria. See "Business—Relationship with Ikaria after the Spin-Out." Following the Spin-Out, Ikaria ceased to hold any of our equity interests and we became a stand-alone, independent operating entity.

Separation and Distribution Agreement

In connection with the Spin-Out, we and Ikaria entered into a separation and distribution agreement which sets forth the key provisions relating to the separation of our business from Ikaria's other businesses. The separation and distribution agreement described the assets and liabilities that remained with or were transferred to us and those that remained with or were transferred to Ikaria and the terms of Ikaria's distribution of all of our outstanding units to its stockholders. The separation and distribution agreement provides for a full and complete release and discharge of all liabilities between Ikaria and us, except as set forth in the agreement. We and Ikaria each agreed to indemnify, defend and hold harmless the other party and its subsidiaries, and each of their respective past and present directors, officers and employees, and each of their respective permitted successors and assigns, from any and all damages relating to, arising out of or resulting from, among other things, our business and certain additional specified liabilities or Ikaria's business and certain additional specified liabilities, as applicable. The separation and distribution agreement also provides that we and Ikaria will each use reasonable best efforts, including by cooperating with the other party, to, among other things, effect the transfer of any assets being transferred in connection with the Spin-Out that had not been transferred as of the date of the Spin-Out.

In connection with the Spin-Out, we and Ikaria have entered into other agreements that will govern various interim and ongoing relationships between us and Ikaria. These agreements, the material terms of which are summarized below, include:

- a transition services agreement;
- an exclusive cross-license, technology transfer, and regulatory matters agreement;
- an employee matters agreement;
- agreements not to compete; and
- drug and device supply agreements.

The principal agreements described below are filed as exhibits to the registration statement to which this prospectus forms a part, and the summaries of each of these agreements below set forth the terms of the agreements that we believe are material. These summaries are qualified in their entireties by reference to the full text of the applicable agreements, which are incorporated by reference into this prospectus.

Transition Services Agreement

In February 2014, we entered into the TSA. Pursuant to the terms and conditions of the TSA, Ikaria has agreed to use commercially reasonable efforts to provide certain services to us, including human resources support, real estate support, information technology support, accounting and tax support, treasury support, financial planning and analysis support, purchasing support, management/executive services, legal services, quality services, regulatory services, drug and device safety services, business development support, biometrics support and manufacturing support. Ikaria is obligated, subject to the terms of the TSA (including the early termination provisions thereof and our obligation

to use commercially reasonable efforts to provide the services for ourselves as soon as practicable), to provide such services until February 2016.

Ikaria has also agreed, on the terms and subject to the conditions of the TSA, to use commercially reasonable efforts to allow our employees to remain in Ikaria's Hampton, New Jersey facility for the continued operation of our business during the term of the TSA.

We are obligated to pay Ikaria a service fee in the amount of $772,000 per month and to reimburse Ikaria for any out-of-pocket expenses incurred in connection with its provisions of services under the TSA, any taxes imposed on Ikaria in connection with the performance or delivery of services under the TSA and any costs and expenses incurred by Ikaria in connection with the performance of any services that require resources outside of the existing resources of Ikaria or that otherwise interfere with the ordinary operations of Ikaria's business. This monthly service fee is payable by us regardless of the frequency or quantity of services actually utilized by us under the TSA, and our obligation to pay such monthly service fee for 24 months will survive any early termination of the TSA. We are also obligated to pay any fees, costs, expenses or other amounts incurred by Ikaria to obtain the right to allow our employees to remain in the Hampton, New Jersey facility during the term of the TSA. At the time of the Spin-Out, we deposited the sum of $18.5 million into escrow, representing the aggregate of the $772,000 monthly service fees payable by the Company under the TSA, to guarantee payment of the monthly service fees by the Company.

Exclusive Cross-License, Technology Transfer and Regulatory Matters Agreement

In February 2014, we entered into an exclusive cross-license, technology transfer and regulatory matters agreement with Ikaria. Pursuant to the terms of the license agreement, Ikaria granted to us a fully paid-up, non-royalty bearing, exclusive license under specified intellectual property rights controlled by Ikaria to engage in the development, manufacture and commercialization of nitric oxide, devices to deliver nitric oxide and related services for or in connection with out-patient, chronic treatment of patients with PAH, PH-COPD or PH-IPF, which we refer to collectively as the Bellerophon indications.

We have granted to Ikaria a fully paid-up, non-royalty-bearing, exclusive license under specified intellectual property rights that we control to engage in the development, manufacture and commercialization of products and services for or used in connection with the diagnosis, prevention or treatment, whether in- or out-patient, of certain conditions and diseases other than the Bellerophon indications specified in the license agreement and for the use of nitric oxide to treat or prevent conditions that are primarily managed in the hospital, which we refer to collectively as the Ikaria nitric oxide business.

We have agreed that, during the term of the license agreement, we will not, without the prior written consent of Ikaria, grant a sublicense under any of the intellectual property licensed to us under the license agreement to any of our affiliates or any third party, in either case, that directly or indirectly competes with the Ikaria nitric oxide business. We have also agreed that we will include certain restrictions in our agreements with customers of our products to ensure that such products will only be used for the Bellerophon indications.

The license agreement will expire on a product-by-product basis for products for a specific Bellerophon indication at such time as we are no longer developing or commercializing any product for such indication. The license agreement may be terminated by either party in the event an act or order of a court or governmental authority prohibits either party from substantially performing under the license agreement. Either party may also terminate the license agreement in the event of an uncured material breach by the other party or in the event the other party is insolvent or in bankruptcy proceedings. Ikaria may also terminate the license agreement if we or any of our affiliates breach the agreements not to compete described below, or if we or any successor to our rights under the license

agreement markets a generic nitric oxide product that is competitive with INOmax. Under certain circumstances, if the license agreement is terminated, the licenses granted to Ikaria by us will survive such termination.

Employee Matters Agreement

In February 2014, we entered into an employee matters agreement with Ikaria, pursuant to which the employment of certain Ikaria employees was transferred to us or our subsidiaries on the terms and conditions set forth therein. The employee matters agreement also sets forth the treatment of outstanding Ikaria stock options and RSUs in connection with the Spin-Out. We have agreed to assume and pay, perform, fulfill and discharge, in accordance with the terms of the employee matters agreement, all liabilities to or relating to such transferred employees. Effective as of the date of the Spin-Out, such transferred employees terminated participation in Ikaria's employee benefit plans, and we or our subsidiaries adopted employee benefit plans substantially similar to the following Ikaria plans: a 401(k) plan, a medical and dental plan, long-term disability, short-term disability, life and accidental death and dismemberment and flexible spending accounts, pursuant to the terms of the employee matters agreement.

Agreements Not to Compete

In September 2013, October 2013 and February 2014, we and each of our subsidiaries entered into an agreement not to compete with a subsidiary of Ikaria, which we refer to collectively as the agreements not to compete. Pursuant to the agreements not to compete, we and each of our subsidiaries agreed not to engage, anywhere in the world, in any manner, directly or indirectly, until the earlier of five years after the effective date of such agreement not to compete or the date on which Ikaria and all of its subsidiaries are no longer engaged in such business, in:

(1) the development, manufacture, commercialization, promotion, sale, import, export, servicing, repair, training, storage, distribution, transportation, licensing or other handling or disposition of any product or service (including, without limitation, any product or service that utilizes, contains or includes nitric oxide for inhalation, a device intended to deliver nitric oxide or a service that delivers or supports the delivery of nitric oxide), bundled or unbundled, for or used in connection with (a) the diagnosis, prevention or treatment, in both adult and/or pediatric populations, and whether in- or out-patient, of: (i) hypoxic respiratory failure associated with pulmonary hypertension, (ii) pulmonary hypertensive episodes and right heart failure associated with cardiovascular surgery, (iii) bronchopulmonary dysplasia, (iv) the management of ventilation-perfusion mismatch in acute lung injury, (v) the management of ventilation-perfusion mismatch in acute respiratory distress syndrome, (vi) the management of pulmonary hypertension episodes and right heart failure in congestive heart failure, (vii) pulmonary edema from high altitude sickness, (viii) the management of pulmonary hypertension episodes and right heart failure in pulmonary or cardiac surgery, (ix) the management of pulmonary hypertension episodes and right heart failure in organ transplant, (x) sickle cell vaso-occlusive crisis, (xi) hypoxia associated with pneumonia or (xii) ischemia-reperfusion injury or (b) the use of nitric oxide to treat or prevent conditions that are primarily managed in the hospital; or

(2) any and all development, manufacture, commercialization, promotion, sale, import, export, storage, distribution, transportation, licensing, or other handling or disposition of any terlipressin or any other product within the pressin family, (a) intended to treat (i) hepatorenal syndrome in any form, (ii) bleeding esophageal varices, or (iii) septic shock, or (b) for or in connection with the management of low blood pressure.

The agreements not to compete expressly exclude the Bellerophon indications.

Supply Agreements

Device Clinical Supply Agreement. In February 2014, we entered into the device supply agreement, pursuant to which Ikaria will use commercially reasonable efforts to manufacture and supply our requirements for certain nitric oxide delivery devices specified in the device supply agreement for use in our clinical programs for PH-COPD and PAH. Pursuant to the device supply agreement, we will pay to Ikaria transfer pricing amounts equal to Ikaria's internal and external manufacturing cost plus 20%. The device supply agreement will expire on February 9, 2015. In addition, either party may terminate the device supply agreement in the event of an uncured material breach by the other party.

Drug Clinical Supply Agreement. In February 2014, we entered into the drug supply agreement, pursuant to which Ikaria has agreed to use commercially reasonable efforts to manufacture and supply, and we have agreed to acquire from Ikaria, our requirements for nitric oxide for inhalation and corresponding placebo for use in our clinical programs for PH-COPD, PAH and PH-IPF. Pursuant to the drug supply agreement, we will pay Ikaria transfer pricing amounts equal to Ikaria's internal and external manufacturing cost plus 20%. Under the terms of the drug supply agreement, we have also granted Ikaria a right of first negotiation in the event that we desire to obtain supply of nitric oxide for inhalation and corresponding placebo (or any variant thereof or any version with different specifications) for commercial use. The drug supply agreement will expire on a product-by-product basis on the date we discontinue clinical development of such product. In addition, either party may terminate the drug supply agreement in the event of an uncured material breach by the other party.

Directors and Officers of Ikaria

Daniel Tassé, our President and Chief Executive Officer and a member of our board of directors, currently serves as President and Chief Executive Officer and is a member of the board of directors of Ikaria. Director Matthew Holt is a member of the board of directors of Ikaria.

Policies and Procedures for Related Person Transactions

Our board of directors has adopted written policies and procedures for the review of any transaction, arrangement or relationship in which we were or are to be a participant, the amount involved exceeds $120,000 and one of our executive officers, directors, director nominees or 5% stockholders, or their immediate family members, each of whom we refer to as a "related person," has a direct or indirect material interest.

If a related person proposes to enter into such a transaction, arrangement or relationship, which we refer to as a "related person transaction," the related person must report the proposed related person transaction to our . The policy calls for the proposed related person transaction to be reviewed and, if deemed appropriate, approved by our audit committee. Whenever practicable, the reporting, review and approval will occur prior to entry into the transaction. If advance review and approval is not practicable, the committee will review, and, in its discretion, may ratify the related person transaction. The policy also permits the chairman of the audit committee to review and, if deemed appropriate, approve proposed related person transactions that arise between committee meetings, subject to ratification by the committee at its next meeting. Any related person transactions that are ongoing in nature will be reviewed annually.

A related person transaction reviewed under the policy will be considered approved or ratified if it is authorized by the audit committee after full disclosure of the related person's interest in the transaction. As appropriate for the circumstances, the committee will review and consider:

- the related person's interest in the related person transaction;
- the approximate dollar value of the amount involved in the related person transaction;

- the approximate dollar value of the amount of the related person's interest in the transaction without regard to the amount of any profit or loss;
- whether the transaction was undertaken in the ordinary course of our business;
- whether the terms of the transaction are no less favorable to us than terms that could have been reached with an unrelated third party;
- the purpose of, and the potential benefits to us of, the transaction; and
- any other information regarding the related person transaction or the related person in the context of the proposed transaction that would be material to investors in light of the circumstances of the particular transaction.

The audit committee may approve or ratify the transaction only if the committee determines that, under all of the circumstances, the transaction is in our best interests. The committee may impose any conditions on the related person transaction that it deems appropriate.

In addition to the transactions that are excluded by the instructions to the SEC's related person transaction disclosure rule, our board of directors has determined that the following transactions do not create a material direct or indirect interest on behalf of related persons and, therefore, are not related person transactions for purposes of this policy:

- interests arising solely from the related person's position as an executive officer of another entity (whether or not the person is also a director of such entity) that is a participant in the transaction, where (a) the related person and all other related persons own in the aggregate less than a 10% equity interest in such entity, (b) the related person and his or her immediate family members are not involved in the negotiation of the terms of the transaction and do not receive any special benefits as a result of the transaction and (c) the amount involved in the transaction is less than the greater of $200,000 or 5% of the annual gross revenues of the company receiving payment under the transaction; and
- a transaction that is specifically contemplated by provisions of our charter or bylaws.

The policy provides that transactions involving compensation of executive officers shall be reviewed and approved by the compensation committee in the manner specified in its charter.

We did not have a written policy regarding the review and approval of related person transactions prior to this offering. Nevertheless, with respect to such transactions, it was our policy for our board of directors to consider the nature of and business reason for such transactions, how the terms of such transactions compared to those which might be obtained from unaffiliated third parties and whether such transactions were otherwise fair to and in the best interests of, or not contrary to, our best interests. In addition, all related person transactions required prior approval, or later ratification, by our board of directors.

PRINCIPAL STOCKHOLDERS

The following table sets forth information with respect to the beneficial ownership of our common stock as of April 30, 2014, after giving effect to the Company Conversion, by:

- each of our directors;
- each of our executive officers;
- all of our directors and executive officers as a group; and
- each person, or group of affiliated persons, who is known by us to beneficially own more than 5% of our common stock.

The column entitled "Percentage of Shares Beneficially Owned—Before Offering" is based on a total of 98,945,820 shares of our common stock outstanding as of April 30, 2014, after giving effect to the Company Conversion and assuming the conversion of our non-voting shares into voting shares of our common stock upon the closing of this offering. The column entitled "Percentage of Shares Beneficially Owned—After Offering" is based on shares of our common stock to be outstanding after this offering, including the shares of our common stock that we are selling in this offering, but not including any additional shares issuable upon exercise of outstanding stock options or upon exercise of the underwriters' option to purchase additional shares.

Beneficial ownership is determined in accordance with the rules and regulations of the SEC and includes voting or investment power with respect to our common stock. Shares of our common stock subject to options or warrants that are currently exercisable or exercisable within 60 days of April 30, 2014 are considered outstanding and beneficially owned by the person holding the options or warrants for the purpose of calculating the percentage ownership of that person but not for the purpose of calculating the percentage ownership of any other person. Except as otherwise noted, the persons and entities in this table have sole voting and investing power with respect to all of the shares of our common stock beneficially owned by them, subject to community property laws, where applicable. Except as otherwise set forth below, the address of the beneficial owner is c/o Bellerophon Therapeutics, Inc., Perryville III Corporate Park, 53 Frontage Road, Suite 301, Hampton, New Jersey 08827.

Following the completion of this offering, (i) the Controlling Entities will be entitled to designate six members of our board of directors, see "Description of Capital Stock," and (ii) the New Mountain Entities will continue to have approval rights over many corporate actions. For a description of the voting agreement and stockholders agreements and any other material relationships the stockholders have or have had with us, our predecessors or our affiliates or with the New Mountain Entities, see "Certain Relationships and Related Person Transactions." In addition, the Controlling Entities will be deemed to hold their shares of our stock as a group.

The table below represents the number of shares owned by each stockholder included in the table (but without taking into account that certain stockholders may be part of a group) to provide investors information concerning the economic ownership of each stockholder.

Name of Beneficial Owner	Shares Beneficially Owned	Percentage of Shares Beneficially Owned	
		Before Offering	After Offering
5% Stockholders			
New Mountain Entities(1)	47,468,903	47.97%	%
Linde(2)	15,918,769	16.09%	%
ARCH(3)	9,431,782	9.53%	%
Venrock(4)	9,400,532	9.50%	%
Executive Officers and Directors			
Daniel Tassé(5)	2,501,400	2.50%	%
Manesh Naidu(6)	201,000	*	%
Reinilde Heyrman	100,000	*	%
Martin Meglasson(7)	180,000	*	%
David Abrams(8)	20,000	*	%
Aldo E. Belloni, Ph.D.(9)	15,918,769	16.09%	%
Matthew S. Holt(10)	47,468,903	47.97%	%
Andre V. Moura	0	*	%
Robert Nelsen(11)	9,431,782	9.53%	%
Howard Pien(12)	102,500	*	%
Adam B. Weinstein(13)	47,468,903	47.97%	%
All executive officers and directors as a group (12 persons)(14)	75,924,354	75.59%	%

* Less than one percent.

(1) Consists of 3,389,078 shares held by Allegheny New Mountain Partners, L.P., 783,009 shares held by New Mountain Affiliated Investors II, L.P. and 43,296,816 shares held by New Mountain Partners II (AIV-A), L.P. The general partner of each of the New Mountain Entities is New Mountain Investments II, L.L.C. and the manager of each of the New Mountain Entities is New Mountain Capital L.L.C. Steven Klinsky is the managing member of New Mountain Investments II, LLC. Adam Weinstein, a member of our board of directors, is a member of New Mountain Investments II, L.L.C. Matthew Holt, a member of our board of directors, is a member of New Mountain Investments II, L.L.C. New Mountain Investments II, L.L.C. has decision-making power over the disposition and voting of shares of portfolio investments of each of the New Mountain Entities. New Mountain Capital, L.L.C. also has voting power over the shares of portfolio investments of the New Mountain Entities in its role as the investment advisor. New Mountain Capital, LLC is a wholly-owned subsidiary of New Mountain Capital Group, LLC. New Mountain Capital Group, LLC is 100% owned by Steven Klinsky. Since New Mountain Investments II, L.L.C. has decision-making power over the New Mountain Entities, Mr. Klinsky may be deemed to beneficially own the shares that the New Mountain Entities hold of record or may be deemed to beneficially own. Mr. Klinsky, Mr. Weinstein, Mr. Holt, New Mountain Investments II, L.L.C. and New Mountain Capital, L.L.C. disclaim beneficial ownership over the shares held by the New Mountain Entities, except to the extent of their pecuniary interest therein.

(2) Consists of 15,918,769 shares held by Linde North America, Inc., an indirect wholly-owned subsidiary of Linde AG. Aldo Belloni, a member of our board of directors, is a Member of the Executive Board of Linde AG. Dr. Belloni disclaims beneficial ownership of all shares held by Linde, except to the extent of his pecuniary interest therein, if any.

(3) Consists of 9,431,782 shares held by IRDO Holding Corp., or ARCH. ARCH Venture Fund VI, L.P., or ARCH VI, is the sole shareholder of ARCH. ARCH Venture Partners VI, L.P., or the GPLP, as the sole general partner of ARCH VI, may be deemed to beneficially own certain of the shares held of record by ARCH. The GPLP disclaims beneficial ownership of all shares held of record by ARCH in which the GPLP does not have an actual pecuniary interest. ARCH Venture Partners VI, LLC, or the GPLLC, as the sole general partner of the GPLP, may be deemed to beneficially own certain of the shares held of record by ARCH. The GPLLC disclaims beneficial ownership of all shares held of record by ARCH in which it does not have an actual pecuniary interest. Keith Crandell, Clinton Bybee and Robert Nelsen, a member of our board of directors, are the managing directors of the GPLLC and may be deemed to beneficially own certain of the shares held of record by ARCH. The managing directors disclaim beneficial ownership of all shares held of record by ARCH in which they do not have an actual pecuniary interest.

(4) Consists of 9,400,532 shares held by Venrock IK Holdings BT, Inc.

(5) Includes 1,200,000 shares of common stock issuable upon the exercise of options exercisable within 60 days after April 30, 2014.

(6) Includes 78,000 shares of common stock issuable upon the exercise of options exercisable within 60 days after April 30, 2014.

(7) Includes 100,000 shares of common stock issuable upon the exercise of options exercisable within 60 days after April 30, 2014.

(8) Includes 20,000 shares of common stock issuable upon the exercise of options exercisable within 60 days after April 30, 2014.

(9) Consists of 15,918,769 shares held by Linde North America, Inc., an indirect wholly-owned subsidiary of Linde AG. Aldo Belloni, a member of our board of directors, is a Member of the Executive Board of Linde AG. Dr. Belloni disclaims beneficial ownership of all shares held by Linde, except to the extent of his pecuniary interest therein, if any.

(10) Consists of 3,389,078 shares held by Allegheny New Mountain Partners, L.P., 783,009 shares held by New Mountain Affiliated Investors II, L.P. and 43,296,816 shares held by New Mountain Partners II (AIV-A), L.P. The general partner of each of the New Mountain Entities is New Mountain Investments II, L.L.C. and the manager of each of the New Mountain Entities is New Mountain Capital L.L.C. Matthew Holt, a member of our board of directors, is a member of New Mountain Investments II, L.L.C. New Mountain Investments II, L.L.C. has decision-making power over the disposition and voting of shares of portfolio investments of each of the New Mountain Entities. New Mountain Capital, L.L.C. also has voting power over the shares of portfolio investments of the New Mountain Entities in its role as the investment advisor. New Mountain Capital, LLC is a wholly-owned subsidiary of New Mountain Capital Group, LLC. Mr. Holt disclaims beneficial ownership over the shares held by the New Mountain Entities, except to the extent of his pecuniary interest therein.

(11) Consists of 9,431,782 shares held by IRDO Holding Corp., or ARCH. ARCH Venture Fund VI, L.P., or ARCH VI, is the sole shareholder of ARCH. ARCH Venture Partners VI, L.P., or the GPLP, as the sole general partner of ARCH VI, may be deemed to beneficially own certain of the shares held of record by ARCH. The GPLP disclaims beneficial ownership of all shares held of record by ARCH in which the GPLP does not have an actual pecuniary interest. ARCH Venture Partners VI, LLC, or the GPLLC, as the sole general partner of the GPLP, may be deemed to beneficially own certain of the shares held of record by ARCH. The GPLLC disclaims beneficial ownership of all shares held of record by ARCH in which it does not have an actual pecuniary interest. Robert Nelsen, a member of our board of directors, is a managing director of the GPLLC and may be deemed to beneficially own certain of the shares held of record by ARCH. Mr. Nelsen

disclaims beneficial ownership of all shares held of record by ARCH in which he does not have an actual pecuniary interest.

(12) Includes 102,500 shares of common stock issuable upon the exercise of options exercisable within 60 days after April 30, 2014.

(13) Consists of 3,389,078 shares held by Allegheny New Mountain Partners, L.P., 783,009 shares held by New Mountain Affiliated Investors II, L.P. and 43,296,816 shares held by New Mountain Partners II (AIV-A), L.P. The general partner of each of the New Mountain Entities is New Mountain Investments II, L.L.C. and the manager of each of the New Mountain Entities is New Mountain Capital L.L.C. Adam Weinstein, a member of our board of directors, is a member of New Mountain Investments II, L.L.C. New Mountain Investments II, L.L.C. has decision-making power over the disposition and voting of shares of portfolio investments of each of the New Mountain Entities. New Mountain Capital, L.L.C. also has voting power over the shares of portfolio investments of the New Mountain Entities in its role as the investment advisor. New Mountain Capital, LLC is a wholly-owned subsidiary of New Mountain Capital Group, LLC. Mr. Weinstein disclaims beneficial ownership over the shares held by the New Mountain Entities, except to the extent of his pecuniary interest therein.

(14) Includes 1,500,500 shares of common stock issuable upon the exercise of options exercisable within 60 days after April 30, 2014.

DESCRIPTION OF CAPITAL STOCK

General

The following description of our capital stock and provisions of our certificate of incorporation and bylaws are summaries and are qualified by reference to the certificate of incorporation and the bylaws that will be in effect upon the closing of this offering. We have filed copies of these documents with the SEC as exhibits to our registration statement of which this prospectus forms a part. The description of the capital stock reflects changes to our capital structure that will occur upon the closing of this offering.

Upon the closing of this offering, our authorized capital stock will consist of shares of our common stock, $0.01 par value per share, and 5,000,000 shares of our preferred stock, $0.01 par value per share, all of which preferred stock will be undesignated.

As of April 30, 2014, after giving effect to the Company Conversion, we had issued and outstanding:

- 98,945,820 shares of our voting and non-voting common stock held by 252 stockholders of record; and
- options to purchase 7,744,480 shares of our non-voting common stock, at a weighted average exercise price of $0.58 per share.

Common Stock

Holders of our common stock are entitled to one vote for each share held on all matters submitted to a vote of stockholders and do not have cumulative voting rights. Each election of directors by our stockholders will be determined by a plurality of the votes cast by the stockholders entitled to vote on the election. Holders of common stock are entitled to receive proportionately any dividends as may be declared by our board of directors, subject to any preferential dividend rights of outstanding preferred stock.

In the event of our liquidation or dissolution, the holders of our common stock are entitled to receive proportionately all assets available for distribution to stockholders after the payment of all debts and other liabilities and subject to the prior rights of any of our outstanding preferred stock. Holders of our common stock have no preemptive, subscription, redemption or conversion rights. The rights, preferences and privileges of holders of our common stock are subject to and may be adversely affected by the rights of the holders of shares of any series of our preferred stock that we may designate and issue in the future.

Preferred Stock

Under the terms of our certificate of incorporation that will become effective upon the closing of this offering, our board of directors is authorized to issue shares of preferred stock in one or more series without stockholder approval. Our board of directors has the discretion to determine the rights, preferences, privileges and restrictions, including voting rights, dividend rights, conversion rights, redemption privileges and liquidation preferences, of each series of preferred stock.

The purpose of authorizing our board of directors to issue preferred stock and determine its rights and preferences is to eliminate delays associated with a stockholder vote on specific issuances. The issuance of preferred stock, while providing flexibility in connection with possible acquisitions, future financings and other corporate purposes, could have the effect of making it more difficult for a third party to acquire, or could discourage a third party from seeking to acquire, a majority of our outstanding voting stock. Upon the closing of this offering, there will be no shares of preferred stock outstanding, and we have no present plans to issue any shares of preferred stock.

Options

As of April 30, 2014, after giving effect to the Company Conversion, we had outstanding options to purchase 7,744,480 shares of our common stock, at a weighted average exercise price of $0.58 per share.

Voting Agreement

We are party to a voting agreement with the Controlling Entities, which will remain in effect following this offering. The voting agreement provides that, among other things, each of the Controlling Entities is required to vote its shares to elect the designated individuals as members of our board of directors. In particular, following the completion of this offering, (i) the New Mountain Entities will be entitled to designate (a) three directors, for so long as they beneficially own 15% or more of our outstanding common stock, (b) two directors, for so long as they beneficially own less than 15% but more than 5% of our outstanding common stock and (c) one director, for so long as they beneficially own less than 5% of our outstanding common stock but more than one share of our common stock and (ii) each of ARCH, Venrock and Linde is entitled to designate one director for so long as such holder beneficially owns 5% or more of our outstanding common stock. If any member of our board of directors designated by the New Mountain Entities, ARCH, Venrock or Linde ceases to serve as a director for any reason, the resulting vacancy will be filled by a director nominated by the person or entity entitled to designate the director. In addition, each Controlling Entity has agreed to vote in favor of the removal of any designated director upon and only upon the written request of the person or entity entitled to designate such director. Each of the Controlled Entities has agreed (i) not to vote in favor of decreasing the size of our board of directors below the number of directors then in office or that would prevent any of the Controlled Entities from designating their respective directors and (ii) so long as the New Mountain Entities beneficially own more than 5% of our common stock, not to vote in favor of increasing the size of our board of directors without the prior written consent of the New Mountain Entities.

Pursuant to the terms of the voting agreement, following the completion of this offering and until we are no longer a "controlled company" under the rules established by The NASDAQ Global Market, (i) if any committee of our board of directors other than an audit committee, compensation committee or nominating and corporate governance committee is established, the members will include at least one director elected by the New Mountain Entities, if any, and either the director elected by ARCH or the director elected by Venrock, if any; (ii) the chair of our compensation committee will be a director elected by the New Mountain Entities, if any, and the members of our compensation committee will include either a director elected by ARCH or Venrock, if any; and (iii) the members of our nominating and corporate governance committee will include the director elected by ARCH, if any, the director elected by Venrock, if any, and more than one director elected by the New Mountain Entities, if two or more directors elected by the New Mountain Entities are members of our board, or the sole director elected by the New Mountain Entities, if only one such director is a member of our board.

Stockholders Agreement

We expect to enter into a stockholders agreement with the New Mountain Entities, which will remain in effect following this offering. The stockholders agreement will provide that, following the closing of the offering, we will be required to obtain the prior written approval of the New Mountain Entities to take certain actions, including, among other things, actions to:

- consolidate or merge into or with any other person, sell, lease or transfer all or substantially all of our assets or capital stock to another person or enter into any other similar business combination transaction, or effect a liquidation;

- authorize, issue, sell, offer for sale or solicit offers to buy any shares of our common stock or any convertible securities or any other equity or debt securities or rights to acquire any of our, or our subsidiaries' equity or debt securities, subject to certain exceptions, including among other things, the issuance under our stock incentive plan of grants that have been approved by our board of directors (or a board committee) and at least one director appointed by the New Mountain Entities;

- incur indebtedness or refinance any indebtedness;

- effect any stock dividend, stock split or other subdivision or combination of our capital stock or any other recapitalization;

- effect any redemption, retirement, purchase or other acquisition, directly or indirectly of our capital stock, subject to certain exceptions for repurchases from our employees, officers, directors, consultants or other persons performing services for us or our subsidiaries;

- hire or replace any of our chief executive officer, chief financial officer or our next two most senior executives (as determined by our board of directors), or materially amend the level or form of compensation or benefits payable to, or other compensation arrangements of, any such officer;

- pay or declare a dividend or distribution on any shares of our capital stock (other than dividends from a wholly-owned subsidiary to its parent company);

- amend, repeal or change any provision of our certificate of incorporation or bylaws; or

- agree or otherwise commit to do any of the foregoing (unless the commitment is conditioned on obtaining the approval of the New Mountain Entities).

These approval rights of the New Mountain Entities will terminate following the closing of the offering when the New Mountain Entities and their assignees beneficially own less than 15% of our outstanding common stock. Following this offering, we expect the New Mountain Entities to hold % of our outstanding common stock (or % if the underwriters exercise in full their option to purchase additional shares from us).

Delaware Anti-Takeover Law and Certain Charter and Bylaw Provisions

Delaware Law

We are subject to Section 203 of the Delaware General Corporation Law. Subject to certain exceptions, Section 203 prevents a publicly held Delaware corporation from engaging in a "business combination" with any "interested stockholder" for three years following the date that the person became an interested stockholder, unless either the interested stockholder attained such status with the approval of our board of directors, the business combination is approved by our board of directors and stockholders in a prescribed manner or the interested stockholder acquired at least 85% of our outstanding voting stock in the transaction in which it became an interested stockholder. A "business combination" includes, among other things, a merger or consolidation involving us and the "interested stockholder" and the sale of more than 10% of our assets. In general, an "interested stockholder" is any entity or person beneficially owning 15% or more of our outstanding voting stock and any entity or person affiliated with or controlling or controlled by such entity or person. The restrictions contained in Section 203 are not applicable to any of our existing stockholders that will own 15% or more of our outstanding voting stock upon the closing of this offering.

Staggered Board; Removal of Directors

Our certificate of incorporation and our bylaws divide our board of directors into three classes with staggered three-year terms. In addition, (i) prior to the first time that the New Mountain Entities beneficially own less than 15% of our outstanding common stock, any director may be removed, with or without cause, by the affirmative vote of the holders of a majority of the outstanding shares of our common stock, provided that any director elected by the New Mountain Entities, ARCH, Venrock or Linde may only be removed without cause by the affirmative vote of a majority of the outstanding shares held by the applicable designating entity and (ii) after the New Mountain Entities beneficially own less than 15% of our outstanding common stock, directors may be removed only for cause and only by holders of at least 75% of the outstanding shares of our common stock, provided that any director elected by the New Mountain Entities, ARCH, Venrock or Linde may also be removed without cause by the affirmative vote of a majority of the outstanding shares held by the applicable designating entity. Furthermore, our certificate of incorporation provides that the authorized number of directors may be changed only by the resolution of our board of directors. The classification of our board of directors and the limitations on the ability of our stockholders to remove directors, change the authorized number of directors and fill vacancies could make it more difficult for a third party to acquire, or discourage a third party from seeking to acquire, control of our company.

Stockholder Action; Special Meeting of Stockholders; Advance Notice Requirements for Stockholder Proposals and Director Nominations

Our certificate of incorporation and our bylaws provide that any action required or permitted to be taken by our stockholders at an annual meeting or special meeting of stockholders may only be taken if it is properly brought before such meeting and may not be taken by written action in lieu of a meeting. Our certificate of incorporation and our bylaws also provide that, except as otherwise required by law, special meetings of the stockholders can only be called by the chairman of our board of directors, our president or chief executive officer or our board of directors. In addition, our bylaws establish an advance notice procedure for stockholder proposals to be brought before an annual meeting of stockholders, including proposed nominations of candidates for election to our board of directors. Stockholders at an annual meeting may only consider proposals or nominations specified in the notice of meeting or brought before the meeting by or at the direction of our board of directors, or by a stockholder of record on the record date for the meeting, who is entitled to vote at the meeting and who has delivered timely written notice in proper form to our secretary of the stockholder's intention to bring such business before the meeting. These provisions could have the effect of delaying until the next stockholder meeting stockholder actions that are favored by the holders of a majority of our outstanding voting securities. These provisions also could discourage a third party from making a tender offer for our common stock, because even if it acquired a majority of our outstanding voting stock, it would be able to take action as a stockholder, such as electing new directors or approving a merger, only at a duly called stockholder meeting and not by written consent.

Super-Majority Voting

The Delaware General Corporation Law provides generally that the affirmative vote of a majority of the shares entitled to vote on any matter is required to amend a corporation's certificate of incorporation or bylaws, unless a corporation's certificate of incorporation or bylaws, as the case may be, requires a greater percentage. Our bylaws may be amended or repealed by a majority vote of our board of directors or the affirmative vote of the holders of at least 75% of the votes that all our stockholders would be entitled to cast in any annual election of directors. In addition, the affirmative vote of the holders of at least 75% of the votes that all our stockholders would be entitled to cast in any election of directors is required to amend or repeal or to adopt any provisions inconsistent with any of the provisions of our certificate of incorporation described above.

Registration Rights

We have entered into a registration rights agreement, dated as February 12, 2014, which we refer to as the registration rights agreement, with certain holders of our common stock, including our 5% stockholders and their affiliates and entities affiliated with our directors. Upon the completion of this offering, holders of a total of 86,176,415 shares of our common stock as of April 30, 2014, will have the right to require us to register these shares under the Securities Act of 1933, as amended, or the Securities Act, or to participate in future registrations of securities by us, under the circumstances described below. After registration pursuant to these rights, these shares will become freely tradable without restriction under the Securities Act.

Demand Registration Rights

At any time or from time to time, subject to specified limitations set forth in the registration rights agreement and to any lock-up period, the holders of 15% of the then outstanding shares having rights under the registration rights agreement, which we refer to as registrable shares, may at any time demand in writing that we register all or a portion of the registrable shares under the Securities Act if the total amount of registrable shares registered have an aggregate offering price of at least $10.0 million (based on the then current market price or fair value), unless the registration is of the balance of the registrable shares held by the holders. We are not obligated to file a registration statement pursuant to this provision on more than two occasions, and we are not obligated to file a registration statement pursuant to this provision within 90 days of the effective date of any other registration statement that we may file.

Form S-3 Registration Rights

In addition, at any time after we become eligible to file a registration statement on Form S-3, subject to specified limitations set forth in the registration rights agreement, the holders of registrable shares may demand in writing that we register on Form S-3 all or a portion of the registrable shares so long as the total amount of registrable shares being registered have an aggregate offering price net of selling expenses of at least $10.0 million (based on the then current market price), unless the registration is of the balance of the registrable shares held by the holders.

Incidental Registration Rights

If, at any time after the closing of this offering, we propose to file a registration statement under the Securities Act, other than pursuant to the demand registration rights described above, the holders of registrable shares will be entitled to notice of the registration and, subject to specified exceptions in the case of an underwritten offering, including market conditions, have the right to require us to register all or a portion of the registrable shares then held by them.

Underwritten Public Offering

In the event that any registration in which the holders of registrable shares participate pursuant to our registration rights agreement is an underwritten public offering, we agree to enter into an underwriting agreement containing customary representation and warranties and covenants, including without limitation customary provisions with respect to indemnification of the underwriters of such offering. Holders of registrable securities must agree to any such underwriting agreement as a condition to participation in the offering. If the total number of shares, including registrable shares, requested by holders to be included in such offering exceeds the largest number of shares to be sold (other than by us) that the underwriters believe can be sold in an orderly manner in such underwritten public offering, then we shall include shares in the offering pursuant to the priority guidelines set forth in the Registration Rights Agreement.

Expenses and Indemnification

Pursuant to the registration rights agreement, we are required to pay all registration expenses, including registration and filing fees, exchange listing fees, printing expenses and accounting fees and the fees and expenses of one counsel to represent the selling stockholders, other than any underwriting discounts and commissions, that are related to any demand or incidental registration described above. The registration rights agreement contains customary cross-indemnification provisions, pursuant to which we are obligated to indemnify the selling stockholders in the event of material misstatements or omissions in the registration statement attributable to us, and they are obligated to indemnify us for material misstatements or omissions in the registration statement attributable to them.

Transfer Agent and Registrar

The transfer agent and registrar for our common stock will be .

NASDAQ Global Market Listing

We intend to apply to have our common stock listed on The NASDAQ Global Market under the symbol "BLPH."

SHARES ELIGIBLE FOR FUTURE SALE

Prior to this offering, there has been no public market for our common stock, and a liquid trading market for our common stock may not develop or be sustained after this offering. Future sales of substantial amounts of our common stock in the public market, including shares issued upon exercise of outstanding options, or the anticipation of these sales, could adversely affect market prices prevailing from time to time and could impair our ability to raise capital through sales of equity securities.

Upon the closing of this offering, we will have outstanding shares of our common stock, after giving effect to the issuance of shares of our common stock in this offering, assuming no exercise by the underwriters of their option to purchase additional shares and no exercise of options or warrants outstanding as of .

Of the shares to be outstanding immediately after the closing of this offering, we expect that the shares to be sold in this offering will be freely tradable without restriction under the Securities Act unless purchased by our "affiliates," as that term is defined in Rule 144 under the Securities Act. The remaining shares of our common stock outstanding after this offering will be "restricted securities" under Rule 144, and we expect that substantially all of these restricted securities will be subject to the 180-day lock-up period under the lock-up agreements as described below. These restricted securities may be sold in the public market upon release or waiver of applicable lock-up agreements and only if registered or pursuant to an exemption from registration, such as Rule 144 or Rule 701 under the Securities Act.

Rule 144

In general, under Rule 144, beginning 90 days after the date of this prospectus, any person who is not our affiliate and has held their shares for at least six months, including the holding period of any prior owner other than one of our affiliates, may sell shares without restriction, subject to the availability of current public information about us. In addition, under Rule 144, any person who is not our affiliate and has not been our affiliate at any time during the preceding three months and has held their shares for at least one year, including the holding period of any prior owner other than one of our affiliates, would be entitled to sell an unlimited number of shares immediately upon the closing of this offering without regard to whether current public information about us is available.

Beginning 90 days after the date of this prospectus, a person who is our affiliate or who was our affiliate at any time during the preceding three months and who has beneficially owned restricted securities for at least six months, including the holding period of any prior owner other than one of our affiliates, is entitled to sell a number of shares within any three-month period that does not exceed the greater of:

- 1% of the number of shares of our common stock then outstanding, which will equal approximately shares immediately after this offering; and
- the average weekly trading volume of our common stock on The NASDAQ Global Market during the four calendar weeks preceding the filing of a Notice of Proposed Sale of Securities Pursuant to Rule 144 with respect to the sale.

Sales under Rule 144 by our affiliates are also subject to manner of sale provisions and notice requirements and to the availability of current public information about us.

Upon waiver or expiration of the 180-day lock-up period described below, approximately shares of our common stock will be eligible for sale under Rule 144, including shares eligible for resale immediately upon the closing of this offering as described above. We cannot estimate the number of shares of our common stock that our existing stockholders will elect to sell under Rule 144.

Rule 701

In general, under Rule 701 of the Securities Act, any of our employees, consultants or advisors, other than our affiliates, who purchased shares from us in connection with a qualified compensatory stock plan or other written agreement is eligible to resell these shares 90 days after the date of this prospectus in reliance on Rule 144, but without compliance with the holding period requirements of Rule 144 and without regard to the volume of such sales or the availability of public information about us. Subject to the 180-day lock-up period described below, approximately shares of our common stock will be eligible for sale in accordance with Rule 701.

Lock-Up Agreements

In connection with this offering, we, our directors, our executive officers and holders of our outstanding common stock, who collectively own % of our common stock, based on shares outstanding as of April 30, 2014, have each agreed to enter in lock-up agreements and will be subject to a lock-up period, meaning that we and our permitted transferees will not be permitted to sell any of the shares of our common stock for 180 days after the date of this prospectus, subject to certain exceptions, without the prior written consent of Leerink Partners LLC and Cowen and Company, LLC on behalf of the several underwriters. The lock-up restrictions and specified exceptions are described in more detail under "Underwriting."

Registration Rights

Upon the closing of this offering, the holders of shares of our common stock or their transferees will be entitled to various rights with respect to the registration of these shares under the Securities Act. Registration of these shares under the Securities Act would result in these shares becoming fully tradable without restriction under the Securities Act immediately upon the effectiveness of the registration, except for shares purchased by affiliates. See "Description of Capital Stock—Registration Rights" for additional information. Shares covered by a registration statement will be eligible for sale in the public market upon the expiration or release from the terms of lock-up agreements applicable to such shares.

Stock Options

As of April 30, 2014, we had outstanding options to purchase 7,744,480 shares of our common stock (as a result of the conversion of existing options to buy limited liability company units into options to buy shares of common stock pursuant to the Company Conversion), of which options to purchase 7,744,480 shares were vested. Following this offering, we intend to file one or more registration statements on Form S-8 under the Securities Act to register all of the shares of our common stock subject to outstanding options and options and other awards issuable pursuant to the public company incentive plan and our pre-IPO stock incentive plans. See "Executive Compensation—Stock Option and Other Compensation Plans" for additional information regarding these plans. Accordingly, shares of our common stock registered under the registration statements will be available for sale in the open market, subject to Rule 144 volume limitations applicable to affiliates, and subject to any vesting restrictions and lock-up agreements applicable to these shares.

MATERIAL U.S. FEDERAL TAX CONSIDERATIONS FOR NON-U.S. HOLDERS OF COMMON STOCK

The following is a general discussion of material U.S. federal income and estate tax considerations relating to ownership and disposition of shares of our common stock issued pursuant to this offering by a non-U.S. holder. For purposes of this discussion, the term "non-U.S. holder" means a beneficial owner of our common stock that is not, for U.S. federal income tax purposes:

- an individual who is a citizen or resident of the United States;
- a corporation, or other entity treated as a corporation for U.S. federal income tax purposes, created or organized in or under the laws of the United States or of any political subdivision of the United States;
- an estate the income of which is subject to U.S. federal income taxation regardless of its source; or
- a trust if (1) a U.S. court is able to exercise primary supervision over the administration of the trust and one or more U.S. persons have authority to control all substantial decisions of the trust or (2) the trust has a valid election to be treated as a U.S. person under applicable U.S. Treasury Regulations.

This discussion does not address the tax treatment of partnerships or other entities that are pass-through entities for U.S. federal income tax purposes or persons that hold their common stock through partnerships or other pass-through entities. A partner in a partnership or other pass-through entity that will hold our common stock should consult his, her or its own tax advisor regarding the tax consequences of acquiring, holding and disposing of our common stock through a partnership or other pass-through entity, as applicable.

This discussion is based on current provisions of the U.S. Internal Revenue Code of 1986, as amended, which we refer to as the Code, existing and proposed U.S. Treasury Regulations promulgated thereunder, current administrative rulings and judicial decisions, all as in effect as of the date of this prospectus and all of which are subject to change or to differing interpretation, possibly with retroactive effect. Any change could alter the tax consequences to non-U.S. holders described in this prospectus. There can be no assurance that the Internal Revenue Service, or the IRS, will not challenge one or more of the tax consequences described in this prospectus. We assume in this discussion that a non-U.S. holder holds shares of our common stock as a capital asset, generally property held for investment.

This discussion does not address all aspects of U.S. federal income and estate taxation that may be relevant to a particular non-U.S. holder in light of that non-U.S. holder's individual circumstances nor does it address any aspects of U.S. state, local or non-U.S. taxes, the alternative minimum tax, or the Medicare tax on net investment income. This discussion also does not consider any specific facts or circumstances that may apply to a non-U.S. holder and does not address the special tax rules applicable to particular non-U.S. holders, such as:

- insurance companies;
- tax-exempt organizations;
- financial institutions;
- brokers or dealers in securities;
- regulated investment companies;
- pension plans;

- controlled foreign corporations;
- passive foreign investment companies;
- persons that have a functional currency other than the U.S. dollar;
- owners deemed to sell our common stock under the constructive sale provisions of the Code;
- owners that hold our common stock as part of a straddle, hedge, conversion transaction, synthetic security or other integrated investment; and
- certain U.S. expatriates.

This discussion is for general information only and it is not tax advice. Accordingly, all prospective investors should consult their own tax advisors regarding the U.S. federal, state, local and non-U.S. tax consequences of the purchase, ownership and disposition of our common stock.

Distributions on Common Stock

As discussed in the "Dividend Policy" section of this prospectus, we do not expect to pay dividends in the foreseeable future. If we pay distributions on our common stock, those distributions generally will constitute dividends for U.S. federal income tax purposes to the extent paid from our current or accumulated earnings and profits, as determined under U.S. federal income tax principles. If a distribution exceeds our current and accumulated earnings and profits, the excess will be treated as a tax-free return of the non-U.S. holder's investment, up to such holder's tax basis in the common stock. Any remaining excess will be treated as capital gain, subject to the tax treatment described below under the heading "Gain on Sale, Exchange or Other Disposition of Common Stock." Any such distributions will also be subject to the discussion below under the section titled "Withholding and information reporting requirements—FATCA."

Dividends paid to a non-U.S. holder generally will be subject to withholding of U.S. federal income tax at a 30% rate or such lower rate as may be specified by an applicable income tax treaty between the United States and such holder's country of residence.

Dividends that are treated as effectively connected with a trade or business conducted by a non-U.S. holder within the United States, and, if an applicable income tax treaty so provides, that are attributable to a permanent establishment or a fixed base maintained by the non-U.S. holder within the United States, are generally exempt from the 30% withholding tax if the non-U.S. holder satisfies applicable certification and disclosure requirements. However, such U.S. effectively connected income, net of specified deductions and credits, is taxed at the same graduated U.S. federal income tax rates applicable to U.S. persons (as defined in the Code). Any U.S. effectively connected income received by a non-U.S. holder that is a corporation may also, under certain circumstances, be subject to an additional "branch profits tax" at a 30% rate or such lower rate as may be specified by an applicable income tax treaty between the United States and such holder's country of residence.

A non-U.S. holder of our common stock who claims the benefit of an applicable income tax treaty between the United States and such holder's country of residence generally will be required to provide a properly executed IRS Form W-8BEN or W-8BEN-E (or successor form) and satisfy applicable certification and other requirements. Non-U.S. holders are urged to consult their own tax advisors regarding their entitlement to benefits under a relevant income tax treaty.

A non-U.S. holder that is eligible for a reduced rate of U.S. withholding tax under an income tax treaty may obtain a refund or credit of any excess amounts withheld by timely filing an appropriate claim with the IRS.

Gain on Sale, Exchange or Other Disposition of Common Stock

A non-U.S. holder generally will not be subject to U.S. federal income tax on gain recognized on a sale, exchange or other disposition of shares of our common stock unless:

- the gain is effectively connected with the non-U.S. holder's conduct of a trade or business in the United States, and, if an applicable income tax treaty so provides, the gain is attributable to a permanent establishment or fixed-base maintained by the non-U.S. holder in the United States; in these cases, the non-U.S. holder will be taxed on a net income basis at the regular graduated rates and in the manner applicable to United States persons (as defined in the Code), and, if the non-U.S. holder is a foreign corporation, an additional branch profits tax at a rate of 30%, or a lower rate as may be specified by an applicable income tax treaty, may also apply;

- the non-U.S. holder is a non-resident alien individual who is present in the United States for 183 days or more in the taxable year of the disposition and certain other requirements are met, in which case the non-U.S. holder will be subject to a 30% tax (or such lower rate as may be specified by an applicable income tax treaty between the United States and such holder's country of residence) on the net gain derived from the disposition, which may be offset by certain U.S. source capital losses of the non-U.S. holder, if any (even though the individual is not considered a resident of the United States, provided that the non-U.S holder has timely filed U.S. federal income tax returns with respect to such losses); or

- we are or have been, at any time during the five-year period preceding such disposition (or the non-U.S. holder's holding period, if shorter) a "U.S. real property holding corporation" unless our common stock is regularly traded on an established securities market and the non-U.S. holder held no more than 5% of our outstanding common stock, directly or indirectly, during the shorter of the 5-year period ending on the date of the disposition or the period that the non-U.S. holder held our common stock. Generally, a corporation is a "U.S. real property holding corporation" if the fair market value of its "U.S. real property interests" equals or exceeds 50% of the sum of the fair market value of its worldwide real property interests plus its other assets used or held for use in a trade or business. We believe that we are not currently, and we do not anticipate becoming, a "U.S. real property holding corporation" for U.S. federal income tax purposes. No assurance can be provided that our common stock will be regularly traded on an established securities market for purposes of the rules described above.

Information Reporting and Backup Withholding Tax

We must report annually to the IRS and to each non-U.S. holder the gross amount of the distributions on our common stock paid to such holder and the tax withheld, if any, with respect to such distributions. Non-U.S. holders may have to comply with specific certification procedures to establish that the holder is not a U.S. person (as defined in the Code) in order to avoid backup withholding at the applicable rate, currently 28%, with respect to dividends on our common stock. Dividends paid to non-U.S. holders subject to withholding of U.S. federal income tax, as described above in "Distributions on Common Stock," generally will be exempt from U.S. backup withholding.

Information reporting and backup withholding generally will apply to the proceeds of a disposition of our common stock by a non-U.S. holder effected by or through the U.S. office of any broker, U.S. or foreign, unless the holder certifies its status as a non-U.S. holder and satisfies certain other requirements, or otherwise establishes an exemption. Generally, information reporting and backup withholding will not apply to a payment of disposition proceeds to a non-U.S. holder where the transaction is effected outside the United States through a non-U.S. office of a broker. However, for information reporting purposes, dispositions effected through a non-U.S. office of a broker with substantial U.S. ownership or operations generally will be treated in a manner similar to dispositions

effected through a U.S. office of a broker. Non-U.S. holders should consult their own tax advisors regarding the application of the information reporting and backup withholding rules to them.

Copies of information returns may be made available to the tax authorities of the country in which the non-U.S. holder resides or is incorporated under the provisions of a specific treaty or agreement.

Backup withholding is not an additional tax. Any amounts withheld under the backup withholding rules from a payment to a non-U.S. holder can be refunded or credited against the non-U.S. holder's U.S. federal income tax liability, if any, provided that an appropriate claim is timely filed with the IRS.

U.S. Federal Estate Tax

Shares of our common stock owned or treated as owned by an individual who is not a citizen or resident of the United States (as specially defined for U.S. federal estate tax purposes) at the time of death are considered U.S. situs assets and will be included in the individual's gross estate for U.S. federal estate tax purposes and, therefore, may be subject to U.S. federal estate tax, unless an applicable estate tax or other treaty provides otherwise.

Withholding and Information Reporting Requirements—FATCA

The Foreign Account Tax Compliance Act, or FATCA, generally imposes a U.S. federal withholding tax at a rate of 30% on payments of dividends on, and gross proceeds from the sale or other disposition of, our common stock if paid to a foreign entity unless (i) if the foreign entity is a "foreign financial institution," the foreign entity undertakes certain due diligence, reporting, withholding, and certain certification obligations, (ii) if the foreign entity is not a "foreign financial institution," the foreign entity identifies certain of its U.S. investors, if any, or (iii) the foreign entity is otherwise exempt under FATCA.

Under applicable U.S. Treasury Regulations, withholding under FATCA will only apply to payments of dividends on our common stock made after June 30, 2014 and to payments of gross proceeds from a sale or other disposition of our common stock made after December 31, 2016. Under certain circumstances, a non-US holder may be eligible for refunds or credits of the tax. An intergovernmental agreement between the United States and an applicable foreign country may modify the requirements described in this paragraph. Non-U.S. holders should consult their own tax advisors regarding the possible implications of this legislation on their investment in our common stock and the entities through which they hold our common stock, including, without limitation, the process and deadlines for meeting the applicable requirements to prevent the imposition of the 30% withholding tax under FATCA.

The preceding discussion of material U.S. federal tax considerations is for general information only. It is not tax advice. Prospective investors should consult their own tax advisors regarding the particular U.S. federal, state, local and non-U.S. tax consequences of purchasing, holding and disposing of our common stock, including the consequences of any proposed changes in applicable laws.

UNDERWRITING

Leerink Partners LLC and Cowen and Company, LLC are acting as representatives of the underwriters. Subject to the terms and conditions set forth in an underwriting agreement among us and the underwriters, we have agreed to sell to the underwriters, and each of the underwriters has agreed, severally and not jointly, to purchase from us, the number of shares of common stock set forth opposite its name below.

Underwriter	Number of Shares
Leerink Partners LLC	
Cowen and Company, LLC	
Total	

Subject to the terms and conditions set forth in the underwriting agreement, the underwriters have agreed, severally and not jointly, to purchase all of the shares sold under the underwriting agreement if any of these shares are purchased. If an underwriter defaults, the underwriting agreement provides that the purchase commitments of the non-defaulting underwriters may be increased or the underwriting agreement may be terminated.

We have agreed to indemnify the underwriters against certain liabilities, including liabilities under the Securities Act, or to contribute to payments the underwriters may be required to make in respect of those liabilities.

The underwriters are offering the shares, subject to prior sale, when, as and if issued to and accepted by them, subject to approval of legal matters by their counsel, including the validity of the shares, and other conditions contained in the underwriting agreement, such as the receipt by the underwriters of officers' certificates and legal opinions. The underwriters reserve the right to withdraw, cancel or modify offers to the public and to reject orders in whole or in part.

Commissions and Discounts

The representatives have advised us that the underwriters propose initially to offer the shares to the public at the public offering price set forth on the cover of this prospectus and to dealers at that price less a concession not in excess of $ per share. After the initial offering, the public offering price, concession or any other term of the offering may be changed.

The following table shows the public offering price, underwriting discount and proceeds before expenses to us. The information assumes either no exercise or full exercise by the underwriters of their option to purchase additional shares of our common stock.

	Per Share	Without Option	With Option
Public offering price	$	$	$
Underwriting discount	$	$	$
Proceeds, before expenses, to us	$	$	$

The expenses of the offering, not including the underwriting discount, are estimated at $ million and are payable by us.

Option to Purchase Additional Shares

We have granted an option to the underwriters, exercisable for 30 days after the date of this prospectus, to purchase up to additional shares at the public offering price, less the underwriting discount. If the underwriters exercise this option, each will be obligated, subject to conditions contained in the underwriting agreement, to purchase a number of additional shares proportionate to that underwriter's initial amount reflected in the above table.

No Sales of Similar Securities

We, our executive officers and directors and all of our other existing security holders have agreed not to sell or transfer any common stock or securities convertible into or exchangeable or exercisable for common stock, for 180 days after the date of this prospectus without first obtaining the written consent of the representatives. Specifically, we and these other persons have agreed, with certain limited exceptions, not to directly or indirectly:

- offer, pledge, sell or contract to sell any common stock;
- sell any option or contract to purchase any common stock;
- purchase any option or contract to sell any common stock;
- grant any option, right or warrant for the sale of any common stock;
- otherwise dispose of or transfer any common stock;
- request or demand that we file a registration statement related to the common stock; and
- enter into any swap or other agreement or any transaction that transfers, in whole or in part, the economic consequence of ownership of any common stock, whether any such swap, agreement or transaction is to be settled by delivery of shares or other securities, in cash or otherwise.

This lock-up provision applies to common stock and to securities convertible into or exchangeable or exercisable for common stock. It also applies to common stock owned now or acquired later by the person executing the agreement or for which the person executing the agreement later acquires the power of disposition.

NASDAQ Global Market Listing

We intend to apply to have our common stock listed on The NASDAQ Global Market under the symbol "BLPH."

Determination of Offering Price

Before this offering, there has been no public market for our common stock. The initial public offering price will be determined through negotiations between us and the representatives. In addition to prevailing market conditions, the factors to be considered in determining the initial public offering price are:

- the valuation multiples of publicly traded companies that the representatives believe to be comparable to us;
- our financial information;
- the history of, and the prospects for, our company and the industry in which we compete;
- an assessment of our management, its past and present operations, and the prospects for, and timing of, our future revenues;
- the present state of our development; and
- the above factors in relation to market values and various valuation measures of other companies engaged in activities similar to ours.

An active trading market for the shares may not develop. It is also possible that after the offering the shares will not trade in the public market at or above the initial public offering price.

The underwriters do not expect to sell more than 5% of the shares in the aggregate to accounts over which they exercise discretionary authority.

Price Stabilization, Short Positions and Penalty Bids

Until the distribution of the shares is completed, SEC rules may limit underwriters and selling group members from bidding for and purchasing our common stock. However, the representatives may engage in transactions that stabilize the price of the common stock, such as bids or purchases to peg, fix or maintain that price.

In connection with the offering, the underwriters may purchase and sell our common stock in the open market. These transactions may include short sales, purchases on the open market to cover positions created by short sales and stabilizing transactions. Short sales involve the sale by the underwriters of a greater number of shares than they are required to purchase in the offering. "Covered" short sales are sales made in an amount not greater than the underwriters' option described above. The underwriters may close out any covered short position by either exercising their option or purchasing shares in the open market. In determining the source of shares to close out the covered short position, the underwriters will consider, among other things, the price of shares available for purchase in the open market as compared to the price at which they may purchase shares through the option granted to them. "Naked" short sales are sales in excess of such option. The underwriters must close out any naked short position by purchasing shares in the open market. A naked short position is more likely to be created if the underwriters are concerned that there may be downward pressure on the price of our common stock in the open market after pricing that could adversely affect investors who purchase in the offering. Stabilizing transactions consist of various bids for or purchases of shares of common stock made by the underwriters in the open market prior to the closing of the offering.

The underwriters may also impose a penalty bid. This occurs when a particular underwriter repays to the underwriters a portion of the underwriting discount received by it because the representatives have repurchased shares sold by or for the account of such underwriter in stabilizing or short covering transactions.

Similar to other purchase transactions, the underwriters' purchases to cover the syndicate short sales may have the effect of raising or maintaining the market price of our common stock or preventing or retarding a decline in the market price of our common stock. As a result, the price of our common stock may be higher than the price that might otherwise exist in the open market. The underwriters may conduct these transactions on The NASDAQ Global Market, in the over-the-counter market or otherwise.

Neither we nor any of the underwriters make any representation or prediction as to the direction or magnitude of any effect that the transactions described above may have on the price of our common stock. In addition, neither we nor any of the underwriters make any representation that the representatives will engage in these transactions or that these transactions, once commenced, will not be discontinued without notice.

Electronic Distribution

In connection with the offering, certain of the underwriters or securities dealers may distribute prospectuses by electronic means, such as e-mail.

Other Relationships

Some of the underwriters and their affiliates may in the future engage in investment banking and other commercial dealings in the ordinary course of business with us or our affiliates. They may in the future receive customary fees and commissions for these transactions.

In addition, in the ordinary course of their business activities, the underwriters and their affiliates may make or hold a broad array of investments and actively trade debt and equity securities (or related derivative securities) and financial instruments (including bank loans) for their own account and for the accounts of their customers. Such investments and securities activities may involve securities and/or

instruments of ours or our affiliates. The underwriters and their affiliates may also make investment recommendations and/or publish or express independent research views in respect of such securities or financial instruments and may hold, or recommend to clients that they acquire, long and/or short positions in such securities and instruments.

Notice to Prospective Investors in the European Economic Area

In relation to each Member State of the European Economic Area (each, a "Relevant Member State"), no offer of shares may be made to the public in that Relevant Member State other than:

A. to any legal entity which is a qualified investor as defined in the Prospectus Directive;

B. to fewer than 100 or, if the Relevant Member State has implemented the relevant provision of the 2010 PD Amending Directive, 150, natural or legal persons (other than qualified investors as defined in the Prospectus Directive), as permitted under the Prospectus Directive, subject to obtaining the prior consent of the representatives; or

C. in any other circumstances falling within Article 3(2) of the Prospectus Directive, provided that no such offer of shares shall require the Company or the representatives to publish a prospectus pursuant to Article 3 of the Prospectus Directive or supplement a prospectus pursuant to Article 16 of the Prospectus Directive.

Each person in a Relevant Member State who initially acquires any shares or to whom any offer is made will be deemed to have represented, acknowledged and agreed that it is a "qualified investor" within the meaning of the law in that Relevant Member State implementing Article 2(1)(e) of the Prospectus Directive. In the case of any shares being offered to a financial intermediary as that term is used in Article 3(2) of the Prospectus Directive, each such financial intermediary will be deemed to have represented, acknowledged and agreed that the shares acquired by it in the offer have not been acquired on a non-discretionary basis on behalf of, nor have they been acquired with a view to their offer or resale to, persons in circumstances which may give rise to an offer of any shares to the public other than their offer or resale in a Relevant Member State to qualified investors as so defined or in circumstances in which the prior consent of the representatives has been obtained to each such proposed offer or resale.

The Company, the representatives and their affiliates will rely upon the truth and accuracy of the foregoing representations, acknowledgements and agreements.

This prospectus has been prepared on the basis that any offer of shares in any Relevant Member State will be made pursuant to an exemption under the Prospectus Directive from the requirement to publish a prospectus for offers of shares. Accordingly, any person making or intending to make an offer in that Relevant Member State of shares which are the subject of the offering contemplated in this prospectus may only do so in circumstances in which no obligation arises for the Company or any of the underwriters to publish a prospectus pursuant to Article 3 of the Prospectus Directive in relation to such offer. Neither the Company nor the underwriters have authorized, nor do they authorize, the making of any offer of shares in circumstances in which an obligation arises for the Company or the underwriters to publish a prospectus for such offer.

For the purpose of the above provisions, the expression "an offer to the public" in relation to any shares in any Relevant Member State means the communication in any form and by any means of sufficient information on the terms of the offer and the shares to be offered so as to enable an investor to decide to purchase or subscribe the shares, as the same may be varied in the Relevant Member State by any measure implementing the Prospectus Directive in the Relevant Member State and the expression "Prospectus Directive" means Directive 2003/71/EC (including the 2010 PD Amending Directive, to the extent implemented in the Relevant Member States) and includes any relevant implementing measure in the Relevant Member State and the expression "2010 PD Amending Directive" means Directive 2010/73/EU.

LEGAL MATTERS

The validity of the shares of our common stock offered hereby is being passed upon for us by Wilmer Cutler Pickering Hale and Dorr LLP. Goodwin Procter LLP is acting as counsel for the underwriters in connection with this offering.

EXPERTS

The financial statements of Bellerophon Therapeutics LLC as of December 31, 2013 and 2012, and for the years ended December 31, 2013 and 2012 and for the period from August 26, 2009 (inception) to December 31, 2013, have been included herein and in the registration statement in reliance upon the report of KPMG LLP, independent registered public accounting firm, appearing elsewhere herein, and upon the authority of said firm as experts in accounting and auditing.

WHERE YOU CAN FIND MORE INFORMATION

We have filed with the SEC a registration statement on Form S-1 under the Securities Act with respect to the shares of our common stock we are offering to sell. This prospectus, which constitutes part of the registration statement, does not include all of the information contained in the registration statement and the exhibits, schedules and amendments to the registration statement. For further information with respect to us and our common stock, we refer you to the registration statement and to the exhibits and schedules to the registration statement. Statements contained in this prospectus about the contents of any contract, agreement or other document are not necessarily complete, and, in each instance, we refer you to the copy of the contract, agreement or other document filed as an exhibit to the registration statement. Each of these statements is qualified in all respects by this reference to such contract, agreement or other document.

You may read and copy the registration statement of which this prospectus is a part at the SEC's public reference room, which is located at 100 F Street, N.E., Room 1580, Washington, DC 20549. You can request copies of the registration statement by writing to the SEC and paying a fee for the copying cost. Please call the SEC at 1-800-SEC-0330 for more information about the operation of the SEC's public reference room. In addition, the SEC maintains an Internet website, which is located at www.sec.gov, that contains reports, proxy and information statements and other information regarding issuers that file electronically with the SEC. You may access the registration statement of which this prospectus is a part at the SEC's Internet website. Upon completion of this offering, we will be subject to the information reporting requirements of the Securities Exchange Act of 1934, as amended, or the Exchange Act, and we will file reports, proxy statements and other information with the SEC.

Upon closing of this offering, we will be subject to the informational and periodic reporting requirements of the Exchange Act. We will fulfill our obligations with respect to such requirements by filing periodic reports and other information with the SEC. We intend to furnish our stockholders with annual reports containing consolidated financial statements certified by an independent registered public accounting firm. We also maintain a website at . The information contained in, or which can be accessed through, our website does not constitute a part of this prospectus.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Index to Financial Statements

Report of Independent Registered Public Accounting Firm

The Board of Directors
Bellerophon Therapeutics LLC

We have audited the accompanying balance sheets of Bellerophon Therapeutics LLC, a development stage company (the "Company"), as of December 31, 2013 and 2012, and the related statements of operations and comprehensive loss, changes in invested equity (deficit), and cash flows for the years ended December 31, 2013 and 2012 and for the period from August 26, 2009 (inception) to December 31, 2013. These financial statements are the responsibility of the Company's management. Our responsibility is to express an opinion on these financial statements based on our audits.

We conducted our audits in accordance with the standards of the Public Company Accounting Oversight Board (United States). Those standards require that we plan and perform the audit to obtain reasonable assurance about whether the financial statements are free of material misstatement. An audit includes examining, on a test basis, evidence supporting the amounts and disclosures in the financial statements. An audit also includes assessing the accounting principles used and significant estimates made by management, as well as evaluating the overall financial statement presentation. We believe that our audits provide a reasonable basis for our opinion.

In our opinion, the financial statements referred to above present fairly, in all material respects, the financial position of Bellerophon Therapeutics LLC, a development stage company, as of December 31, 2013 and 2012, and the results of its operations and its cash flows for the years ended December 31, 2013 and 2012, and for the period from August 26, 2009 (inception) to December 31, 2013, in conformity with U.S. generally accepted accounting principles.

/s/ KPMG LLP

Short Hills, New Jersey
May 14, 2014

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Balance Sheets

(Amounts in thousands)

	Unaudited Pro Forma December 31, 2013 (Note 15)	December 31, 2013	December 31, 2012
Assets			
Current assets:			
Cash and cash equivalents	$ 61,500	$ —	$ —
Restricted cash	9,250	—	—
Prepaid expenses and other current assets	1,552	1,552	319
Total current assets	72,302	1,552	319
Restricted cash, non-current	9,250	—	—
Property, plant and equipment, net	2,084	2,084	1,703
Other assets	—	—	1,327
Total assets	$ 83,636	$ 3,636	$ 3,349
Liabilities and Invested Equity (Deficit)			
Current liabilities:			
Accounts payable	$ 1,368	$ 1,368	$ 1,487
Accrued research and development	7,591	7,591	6,634
Employee compensation and benefits	3,194	3,194	2,362
Other current liabilities	—	1,839	728
Total current liabilities	12,153	13,992	11,211
Allocated portion of Ikaria special dividend bonus payable	—	4,273	2,865
Other liabilities	1,108	1,108	389
Total liabilities	13,261	19,373	14,465
Commitments and contingencies (Note 13)			
Invested equity (deficit):			
Investment by Ikaria, Inc.	246,890	160,778	103,401
Deficit accumulated during the development stage	(176,515)	(176,515)	(114,517)
Total invested equity (deficit)	70,375	(15,737)	(11,116)
Total liabilities and invested (deficit)	$ 83,636	$ 3,636	$ 3,349

The accompanying notes are an integral part of these financial statements.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Statements of Operations and Comprehensive Loss

(Amounts in thousands)

	Year Ended December 31, 2013	Year Ended December 31, 2012	Period from August 26, 2009 (inception) to December 31, 2013
Operating expenses:			
Research and development	$ 52,985	$ 38,727	$ 147,887
General and administrative	9,013	7,185	27,690
Other operating expense	—	315	938
Total operating expenses	(61,998)	(46,227)	(176,515)
Income tax benefit (expense)	—	—	—
Net loss and comprehensive loss	$ (61,998)	$ (46,227)	$ (176,515)

The accompanying notes are an integral part of these financial statements.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Statements of Changes in Invested Equity (Deficit)

(Amounts in thousands)

	Investment by Ikaria, Inc.	Deficit Accumulated During the Development Stage	Total Invested Equity (Deficit)
Balance at August 26, 2009 (inception)	$ —	$ —	$ —
Net loss	—	(17,279)	(17,279)
Investment by Ikaria, Inc., net	7,282	—	7,282
Balance at December 31, 2009	7,282	(17,279)	(9,997)
Net loss	—	(13,581)	(13,581)
Investment by Ikaria, Inc., net	22,087	—	22,087
Balance at December 31, 2010	29,369	(30,860)	(1,491)
Net loss	—	(37,430)	(37,430)
Investment by Ikaria, Inc., net	36,459	—	36,459
Balance at December 31, 2011	65,828	(68,290)	(2,462)
Net loss	—	(46,227)	(46,227)
Investment by Ikaria, Inc., net	37,573	—	37,573
Balance at December 31, 2012	103,401	(114,517)	(11,116)
Net loss	—	(61,998)	(61,998)
Investment by Ikaria, Inc., net	57,377	—	57,377
Balance at December 31, 2013	$ 160,778	$ (176,515)	$ (15,737)

The accompanying notes are an integral part of these financial statements.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Statements of Cash Flows

(Amounts in thousands)

	Year Ended December 31, 2013	Year Ended December 31, 2012	Period from August 26, 2009 (inception) to December 31, 2013
Cash flows from operating activities:			
Net loss	$ (61,998)	$ (46,227)	$ (176,515)
Adjustments to reconcile net loss to net cash used in operating activities:			
Depreciation	429	85	585
Stock-based compensation	1,721	1,449	4,447
Loss on disposal of property, plant and equipment, net	—	2,840	2,841
Other items	149	315	674
Changes in operating assets and liabilities:			
Decrease (increase) in other current assets and other assets	94	(11)	(1,866)
Increase in accounts payable, accrued research and development, and other current liabilities	1,655	5,346	12,153
Increase (decrease) in other liabilities	719	(21)	1,108
Net cash used in operating activities	(57,231)	(36,224)	(156,573)
Cash flows from investing activities:			
Capital expenditures	(727)	(3,478)	(5,427)
Net cash used in investing activities	(727)	(3,478)	(5,427)
Cash flows from financing activities:			
Cash contributions from Ikaria, Inc., net	57,958	39,702	162,000
Net cash provided by financing activities	57,958	39,702	162,000
Net increase in cash and cash equivalents	—	—	—
Cash and cash equivalents, at beginning of year	—	—	—
Cash and cash equivalents, at end of year	$ —	$ —	$ —
Non-cash investing activities:			
Contribution of property, plant and equipment from Ikaria, Inc., net	83	—	83
Non-cash financing activities:			
Investment by Ikaria, Inc., net	$ (581)	$ (2,129)	$ (1,222)

The accompanying notes are an integral part of these financial statements.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Notes to Financial Statements

(1) Organization and Nature of the Business

Bellerophon Therapeutics LLC, or the Company, is a clinical stage biotherapeutics company focused on developing innovative products at the intersection of drugs and devices that address significant unmet needs in the treatment of cardiopulmonary and cardiac diseases. During the periods presented in these financial statements, the Company was the research and development operating segment of Ikaria, Inc., or Ikaria. As of December 31, 2013, the Company had two wholly-owned subsidiaries, Bellerophon Pulse Technologies LLC (formerly known as Ikaria Pulse Technologies LLC), a Delaware limited liability company, and Bellerophon BCM LLC (formerly known as Ikaria BCM LLC), a Delaware limited liability company. In January 2014, the Company formed a new wholly-owned subsidiary, Bellerophon Services, Inc., a Delaware corporation. The Company is conducting Phase 2 clinical trials of its inhaled nitric oxide product candidates using its proprietary pulsatile technology, which are referred to as the INOpulse product candidates, for the treatment of pulmonary arterial hypertension, or PAH, and pulmonary hypertension associated with chronic obstructive pulmonary disease, or PH-COPD. In addition, the Company is conducting a clinical trial of bioabsorbable cardiac matrix, or BCM, its product candidate for the prevention of left ventricular remodeling following acute myocardial infarction, or AMI, commonly known as a heart attack, which is intended to prevent progression to congestive heart failure.

The Company's business is subject to significant risks and uncertainties including, but not limited to:

- The risk that the Company will not achieve success in its research and development efforts, including clinical trials conducted by it or its potential collaborative partners.
- The expectation that the Company will experience operating losses for the next several years.
- Decisions by regulatory authorities regarding whether and when to approve the Company's regulatory applications as well as their decisions regarding labeling and other matters which could affect the commercial potential of the Company's products or product candidates.
- The risk that the Company will fail to obtain adequate financing to meet its future capital and financing needs.
- The risk that key personnel will leave the Company and/or that the Company will be unable to recruit and retain senior level officers to manage its business.

During the third quarter of 2013 in conjunction with Ikaria's financing activities, Ikaria began reporting financial information for two operating segments: its research and development business and its commercial business. During the fourth quarter of 2013, Ikaria completed an internal reorganization of the assets and subsidiaries of its two operating segments. In connection with the internal reorganization, Ikaria formed the Company as a new wholly-owned subsidiary and transferred the research and development-related assets related to INOpulse for PAH and INOpulse for PH-COPD to the Company and/or its subsidiaries. See Note 14 —*Equity Adjustments, Bellerophon Spin-Out and Merger*.

On December 24, 2013, Ikaria and Madison Dearborn Partners, or MDP, entered into an agreement and plan of merger, under which MDP would acquire a majority ownership position in Ikaria and existing shareholders retained a minority ownership position in Ikaria through certain merger transactions, or the Merger. The Merger was preceded by the pro rata distribution, or

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Notes to Financial Statements (Continued)

(1) Organization and Nature of the Business (Continued)

Spin-Out, of all of the outstanding units of the Company to existing Ikaria stockholders through a special dividend. The Merger and Spin-Out were completed on February 12, 2014. See Note 14—*Equity Adjustments, Bellerophon Spin-Out and Merger*.

(2) Summary of Significant Accounting Policies

As the Company has not yet earned any revenue from its operations, it considers itself a development stage company as defined under Financial Accounting Standards Board Accounting Standards Codification Topic 915, Development Stage Entities. Pursuant to Topic 915, the Company is required to provide certain additional disclosures regarding cumulative expenses, losses and cash flows, as well as other information, as applicable, from its date of inception. For purposes of the financial statements presented herein, the Company has used August 26, 2009, the date of the Company's license and commercialization agreement for BCM, as the effective inception date of the Company. There was no program-specific research and development activity conducted by Ikaria prior to August 26, 2009 included in the business of the Company.

(a) Basis of Presentation

The financial statements presented herein have been derived from the audited historical financial statements and accounting records of Ikaria. These financial statements include all costs that were directly attributable to the Company plus an allocated amount of Ikaria's general and administrative and certain research and development expenses.

Direct and indirect costs related to the Company for INOpulse for PAH, INOpulse for PH-COPD and BCM clinical programs have been allocated to the Company. All allocations were based on actual costs incurred. For purposes of allocating non-project specific expenses, each departmental head provided information as to the percentage of employee time incurred on behalf of the Company.

Allocations of general and administrative expenses by Ikaria to the Company include allocations of corporate management, finance, information technology, legal, human resources and other overhead expenses, based on an approximate pro-rata headcount of employees.

The Company's balance sheets include assets and liabilities that were specifically identified and those that were allocated by Ikaria to the Company based on an estimate of the benefit derived from the underlying asset or liability. Ikaria has historically used a centralized approach to cash management and financing of its operations. Cash funding for the Company has been accounted for as a capital contribution from Ikaria. See Note 3—*Liquidity*.

Management believes that the statements of operations include a reasonable allocation of costs and expenses incurred by Ikaria which benefited the Company. However, such amounts may not be indicative of the actual level of costs and expenses that would have been incurred by the Company if it had operated as an independent stand-alone company or of the costs and expenses expected to be incurred in the future. As such, the financial information herein may not necessarily reflect the financial position, results of operations and cash flows of the Company expected in the future or what it would have been had it been an independent stand-alone company during the periods presented.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Notes to Financial Statements (Continued)

(2) Summary of Significant Accounting Policies (Continued)

The financial statements have been prepared in accordance with generally accepted accounting principles in the United States, or GAAP. Intercompany transactions have been eliminated.

In addition to the allocation process outlined above, the preparation of financial statements in conformity with GAAP requires management to make estimates and assumptions that affect the amounts reported and disclosure of contingent assets and liabilities. Actual results could differ from those estimates.

(b) Cash and Cash Equivalents

The Company considers all highly liquid investments with an original maturity date of three months or less to be cash equivalents.

(c) Property, Plant and Equipment

Property, plant and equipment are recorded at acquisition cost, which for internally developed assets include labor, materials and overhead. Additions and improvements that increase the value or extend the life of an asset are capitalized. Repairs and maintenance costs are expensed as incurred.

Depreciation is computed using the straight-line method over the estimated useful lives described below:

Asset description	Estimated useful life (years)
Machinery, equipment and furniture	3 - 15

(d) Impairment of Long-Lived Assets

Long-lived assets, such as property, plant and equipment, are reviewed for impairment whenever events or changes in circumstances indicate that the carrying amount of an asset may not be recoverable. Recoverability of assets to be held and used is measured by a comparison of the carrying amount of an asset to estimated undiscounted expected future cash flows. If the carrying amount of an asset exceeds its estimated future cash flows, an impairment charge is recognized for the amount by which the carrying amount of the asset exceeds the fair value of the asset. Assets to be sold are no longer depreciated and are reclassified outside of property, plant and equipment at the lower of the carrying amount or fair value less costs to sell.

(e) Stock-Based Compensation

Stock-based compensation expense for the Company represents an allocation of Ikaria's stock-based compensation expense based on the allocation percentages of the Company's cost centers, which were determined based on specific identification or the proportionate percentage of employee time or headcount to the respective total Ikaria employee time or headcount.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Notes to Financial Statements (Continued)

(2) Summary of Significant Accounting Policies (Continued)

(f) Income Taxes

During the periods presented in these financial statements, the Company did not file separate tax returns as the Company was included in the tax groupings of other Ikaria entities within the respective entity's tax jurisdiction. As such, the income tax provision included in these financial statements has been calculated using the separate return method, as if the Company filed a separate tax return in each of its respective tax jurisdictions. The income tax provision included in these carve-out financial statements reflects Ikaria's status as a C-corporation. Subsequent to the Spin-Out, and prior to the conversion of the Company from a limited liability company to a corporation, the Company will be taxed as a partnership.

For financial reporting purposes, the Company has historically recorded no tax expense or benefit due to its operating loss position. A valuation allowance has been established on net deferred tax assets because management believes that it is more likely than not that the Company's net deferred tax assets will not be realized.

Deferred tax assets and liabilities are recognized for the estimated future tax consequences attributable to differences between the financial statement carrying amounts of existing assets and liabilities and their respective tax bases. Deferred tax assets and liabilities are measured using enacted tax rates in effect for the year in which those temporary differences are expected to be recovered or settled.

The Company recognizes the benefit of an uncertain tax position that it has taken or expects to take on income tax returns prepared under a separate return method if such tax position is more likely than not to be sustained on examination by the taxing authorities, based on the technical merits of the position. These tax benefits are measured based on the largest benefit that has a greater than 50% likelihood of being realized upon ultimate resolution. Unrecognized tax benefits related to net operating loss carryforwards or tax credit carryforwards are presented as a reduction to the related gross deferred tax assets. Unrecognized tax benefits for which a net operating loss carryforward or tax credit carryforward is not available are presented as a liability. A liability for unrecognized tax benefits is classified as non-current unless the liability is expected to be settled in cash within 12 months of the reporting date.

Certain deferred tax assets that arose as a result of Ikaria's past activities and resultant operating losses, such as federal and state net operating loss carryforwards, research and development credit carryforwards and acquired in-process research and development, do not constitute assets of the Company and continued to constitute assets of Ikaria subsequent to the date of the Spin-Out.

(g) Research and Development Expense

Research and development costs are expensed as incurred. These expenses include the costs of the Company's proprietary research and development efforts, as well as costs incurred in connection with certain licensing arrangements. Upfront and milestone payments made to third parties in connection with research and development collaborations are expensed as incurred up to the point of regulatory approval. Payments made to third parties upon or subsequent to regulatory approval are capitalized and amortized over the remaining useful life of the related product. The Company also expenses the cost of purchased technology and equipment in the period of purchase if it believes that the technology

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Notes to Financial Statements (Continued)

(2) Summary of Significant Accounting Policies (Continued)

or equipment has not demonstrated technological feasibility and it does not have an alternative future use. Nonrefundable advance payments for goods or services that will be used or rendered for future research and development activities are deferred and are recognized as research and development expense as the related goods are delivered or the related services are performed.

(h) Use of Estimates

The preparation of financial statements in conformity with accounting principles generally accepted in the United States requires management to make estimates and assumptions that affect the reported amounts of assets and liabilities and disclosure of contingent assets and liabilities at the date of the financial statements and the reported amounts of costs and expenses during the reporting period, including accrued research expenses, share-based compensation, income taxes and impairment of long-lived assets. Actual results could differ from those estimates.

(3) Liquidity

In the course of its development activities, the Company has sustained operating losses and expects such losses to continue over the next several years. The Company's ultimate success depends on the outcome of its research and development activities. From August 26, 2009 (inception) to December 31, 2013, the Company's net losses were $176.5 million. Management expects to incur additional losses in the future to conduct product research and development and recognizes the need to raise additional capital through the potential issuance of additional equity or borrowings or entering into strategic alliances with partner companies. However, if such financing is not available at adequate levels or strategic alliances with partner companies do not occur, the Company will need to reevaluate its plans.

In connection with the Spin-Out, $80.0 million of cash was distributed to the Company. This cash is expected to be sufficient to satisfy the Company's operating cash needs for at least 12 months from December 31, 2013. At the time of the Spin-Out, $18.5 million of the $80.0 million cash held by the Company was deposited in escrow to guarantee payment of the monthly services fees payable by the Company to Ikaria in exchange for the services to be provided by Ikaria pursuant to the transition services agreement during the 24 months following the Spin-Out. The escrowed cash will be classified as restricted cash in periods subsequent to the Spin-Out. See Note 11—*Related-Party Transactions*.

(4) Restructuring Charges

In December 2011, Ikaria announced the planned closure of its Seattle-based facility. Charges allocated to the Company included $0.6 million for facility lease obligations, which was recorded in other operating expense, $0.2 million for the impairment of fixed assets and $0.5 million for severance and related charges, which were recorded in research and development expense. Accrued severance and related charges were paid in 2012. Facility lease obligations extended through March 2014. In 2012, an additional $0.3 million charge was recorded for the impairment of fixed assets related to the closure of the Seattle-based facility, which was recorded in other operating expense in the Statement of Operations and Comprehensive Loss.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Notes to Financial Statements (Continued)

(5) Property, Plant and Equipment

Property, plant and equipment and accumulated depreciation either specifically identified or allocated to the Company by Ikaria consists of the following (in thousands):

	December 31,	
	2013	2012
Machinery, equipment and furniture	$ 2,943	$ 1,859
Less accumulated depreciation	(859)	(156)
	$ 2,084	$ 1,703

During 2013, Ikaria transferred to the Company gross fixed assets of $0.4 million with accumulated depreciation of $0.3 million in connection with the move of certain assets from Seattle to New Jersey.

(6) Other Current Liabilities

Other current liabilities either specifically identified or allocated to the Company by Ikaria consist of the following accrued expenses (in thousands):

	December 31,	
	2013	2012
Allocated current portion of Ikaria special dividend bonus payable	$ 1,839	$ 713
Other accrued liabilities	—	15
Total	$ 1,839	$ 728

See Note 8—*Share-Based Compensation*, for a discussion of the Ikaria special dividend bonus payable.

(7) Income Taxes

During the periods presented in these financial statements, the Company did not file separate income tax returns, as the Company was included in the tax groupings of other Ikaria entities within the respective entity's tax jurisdictions. As such, the income tax provisions included in these financial statements have been calculated using the separate return method, as if the Company filed a separate tax return in each of its respective tax jurisdictions, although no tax expense or benefit has been recorded due to its operating loss position. The Company has been treated as a C-corporation, based on Ikaria's tax status, for purposes of these financial statements.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Notes to Financial Statements (Continued)

(7) Income Taxes (Continued)

A reconciliation of the statutory federal income tax rate to the Company's effective tax rate for the years ended December 31, 2013 and 2012 is as follows:

	Year Ended December 31, 2013	Year Ended December 31, 2012
U.S. federal statutory rate	35.0%	35.0%
State and local taxes, net of federal tax effect	5.3%	5.2%
Research tax credits	5.0%	5.6%
Valuation allowance	(44.4)%	(44.6)%
Incentive stock options	(0.1)%	(0.2)%
Other	(0.8)%	(1.0)%
	0.0%	0.0%

Deferred taxes reflect the tax effects of the differences between the amounts recorded as assets and liabilities for financial reporting purposes and the comparable amounts recorded for income tax purposes. Significant components of the deferred tax assets (liabilities) at December 31, 2013 and 2012 are as follows (in thousands):

	December 31, 2013		December 31, 2012	
	Assets	(Liabilities)	Assets	(Liabilities)
Net operating loss carryforwards	$ 62,295	$ —	$ 38,510	$ —
Research tax credit carryforwards	9,615	—	5,511	—
Property, plant and equipment	—	(1,269)	—	(939)
Intangible assets	5,140	—	5,625	—
Accrued compensation	1,103	—	613	—
Other	28	—	141	—
Subtotal	78,181	(1,269)	50,400	(939)
Valuation allowance	(76,912)	—	(49,461)	—
Total deferred tax assets (liabilities)	$ 1,269	$ (1,269)	$ 939	$ (939)
Net deferred tax assets	$ 0		$ 0	

A valuation allowance is provided when it is more likely than not that some portion or all of the deferred tax assets will not be realized. As of December 31, 2013, management believed that it was more likely than not that the deferred tax assets would not be realized, based on future operations, consideration of tax strategies and the reversal of deferred tax liabilities. As of December 31, 2013 and 2012, the Company had gross deferred tax assets of $78.2 million and $50.4 million, respectively. The Company maintained a valuation allowance of $76.9 million and $49.5 million at December 31, 2013 and 2012, respectively.

As of December 31, 2013 and 2012, the Company had unrecognized tax benefits related to research tax credit carryforwards, which were reflected as an offset to the gross deferred tax asset.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Notes to Financial Statements (Continued)

(8) Share-Based Compensation

Ikaria's employees have received Ikaria share-based compensation awards, and therefore, the following disclosures pertain to share-based compensation and the Ikaria special dividend bonus payable that have been allocated to the Company related to Ikaria share-based awards. See Note 2(a)—*Summary of Significant Accounting Policies—Basis of Presentation*. Accordingly, the amounts presented are not necessarily indicative of future awards and do not necessarily reflect what the Company would have experienced as a stand-alone company for the periods presented.

Ikaria Special Dividend Plan

In October 2011, Ikaria approved dividend equivalent rights for options, restricted stock units, or RSUs, and other equity awards granted under its equity award plans. Pursuant to the special dividend plan, in the event that the Ikaria board declared a dividend, each employee of the Company who held equity awards was eligible to receive a cash payment equal to the amount of the dividend per share, multiplied by the number of equity awards outstanding. The payment was payable as of the declaration date for vested options. For unvested options and unvested RSUs, payment was due upon vesting. As of December 31, 2013, the allocated portion of the special dividend bonus payable was $6.1 million of which $1.8 million was reflected in other current liabilities and $4.3 million was reflected in non-current liabilities. As of December 31, 2012, the allocated portion of the special dividend bonus payable was $3.6 million of which $0.7 million was reflected in other current liabilities and $2.9 million was reflected in non-current liabilities.

Stock Options

Stock options are generally granted by Ikaria with an exercise price equal to the fair value of a share of non-voting common stock on the date of the grant. The fair value of the common stock has been determined by the board of directors after considering a broad range of factors, including, but not limited to, the rights, preferences and privileges of the redeemable convertible preferred stock relative to those of the Ikaria's common stock, Ikaria's operating and financial performance, the introduction of new products, the stage of development of Ikaria's product candidates and the likelihood of regulatory approval, Ikaria's revenue growth, the lack of an active public market for Ikaria's stock, industry information such as market growth and volume, the performance of similarly situated companies in Ikaria's industry, the execution of strategic and development agreements, and the likelihood of achieving a liquidity event, such as an initial public offering, given prevailing market conditions and the nature and history of Ikaria's business. Stock options have a contractual life of ten years and generally have a vesting term of four years. Ikaria issues previously unissued non-voting common stock upon the exercise of stock options.

Compensation expense for stock options granted to employees and directors is based on the estimated grant date fair value of options and is recognized by Ikaria over the requisite service period on a straight-line expense attribution method.

BELLEROPHON THERAPEUTICS LLC

(A Development Stage Company)

Notes to Financial Statements (Continued)

(8) Share-Based Compensation (Continued)

Valuation Assumptions for Stock Options

The weighted average grant date fair value of stock options granted to employees and directors of Ikaria and the weighted average assumptions used by Ikaria to estimate the grant date fair value of the options using the Black-Scholes-Merton option pricing model were:

	2013	2012
Weighted average grant date fair value	$1.95	$2.40
Valuation assumptions:		
Risk-free rate	0.90%	0.83%
Expected volatility	46.5%	47.6%
Expected term	5.00 yrs	5.00 yrs
Dividend yield	—	—

Because Ikaria is not publicly traded and has limited operating history, the expected volatility is based on the median historic volatility for publicly traded industry peers. In addition, Ikaria has minimal historical information to develop expectations about future exercise patterns for its stock option grants. As a result, the expected term is based on an average of the expected term of options granted by Ikaria's publicly traded industry peers. The risk-free interest rate is based on the implied yield on U.S. Treasury zero coupon bonds for periods commensurate with the expected term of the options. The dividend yield on Ikaria's common stock is zero which is consistent with offering dividend equivalent rights for vested options and RSUs. Prior to the dividend equivalent rights program, Ikaria did not intend to pay dividends at the time of grant or during the expected term of its stock options.

Restricted Stock Units

Ikaria has granted RSUs to employees that generally vest over a four-year period. RSUs granted prior to January 1, 2011 vested 25% annually. RSUs granted on and after January 1, 2011 vest 25% on the second and third anniversary of the date of grant and 50% on the fourth anniversary of the date of grant. Shares of Ikaria non-voting common stock are delivered to the employee upon vesting, subject to payment of applicable withholding taxes, which may be paid in cash or an equivalent amount of shares withheld. Compensation expense for all RSUs is based on the grant date fair value of the RSU issued, which is based on the fair value of non-voting common stock. Ikaria recognizes compensation expense for RSUs on a straight-line basis over the requisite service period.

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Notes to Financial Statements (Continued)

(8) Share-Based Compensation (Continued)

Stock-Based Compensation Expense, Net of Estimated Forfeitures

The following table summarizes allocated stock-based compensation expense by statement of operations line item for the years ended December 31, 2013 and 2012 and for the period from August 26, 2009 (inception) through December 31, 2013 (in thousands):

	2013	2012	Period from August 26, 2009 (inception) through December 31, 2013
Research and development	$ 1,120	$ 882	$ 2,795
General and administrative	601	567	1,652
Total expense	1,721	1,449	4,447
Tax benefit	(232)	(140)	(600)
Expense, net of tax benefit	$ 1,489	$ 1,309	$ 3,847

Long Term Incentive Plan

In August 2012, under Ikaria's Long-Term Incentive Plan, or LTIP, Ikaria granted cash settled awards to its employees. The awards vest over four years, 25% on the second and third anniversary of the grant and 50% on the fourth anniversary of the grant, and are expensed over the requisite service period on a straight-line expense attribution method. The award value is tied to Ikaria's stock price and is adjusted at each reporting period to estimated fair value. Upon vesting, the awards will be settled in cash. The Company recognized a de minimis amount of allocated expense in 2013 and 2012 and the period from August 26, 2009 (inception) through December 31, 2013 for the LTIP.

(9) Investment by Ikaria, Inc.

The Company's historical operating cash requirements have been provided by Ikaria. The balances in the investment by Ikaria account as of December 31, 2013 and 2012 of $160.8 million and $103.4 million, respectively, represent the investment by Ikaria in the Company, including cash funding as well as the impact of share-based compensation awards which increases equity and the Ikaria special dividend bonus payable allocated to the Company which decreases equity.

(10) Product Acquisitions and Other Agreements

The Company has entered, and will consider entering, into agreements to develop and commercialize product candidates, which may include research and development, marketing and selling, manufacturing and distribution agreements. These agreements often require milestone and royalty or profit share payments, contingent upon the occurrence of certain future events linked to the success of the asset in development, as well as expense reimbursements. Costs incurred pursuant to these agreements are reported in their respective expense line items in the Statements of Operations.

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Notes to Financial Statements (Continued)

(10) Product Acquisitions and Other Agreements (Continued)

BioLineRx Ltd.

On August 26, 2009, the Company entered into an agreement with BioLineRx Ltd. and BioLine Innovations Jerusalem L.P., which are referred to collectively as BioLine, under which the Company obtained the worldwide exclusive rights to BCM. The Company paid BioLine a $7.0 million upfront payment in 2009 and a $10.0 million milestone payment in 2010.

Under the terms of the license agreement, the Company is obligated to use commercially reasonable efforts to develop and commercialize at least one product containing BCM. Under the terms of the license agreement, if the Company achieves certain clinical and regulatory events specified in the license agreement, the Company will be obligated to pay milestone payments to BioLine that could total, in the aggregate, up to $115.5 million, and if the Company achieves certain commercialization targets specified in the license agreement, the Company will be obligated to pay additional milestone payments to BioLine that could total, in the aggregate, up to $150.0 million. In addition, the Company is obligated to pay BioLine a specified percentage of any upfront consideration it receives for sublicensing BCM, as well as royalties in the low double digits below 20% on net sales, if any, of any approved product containing BCM, subject to offsets for specified payments to third parties made in connection with BCM. The Company's obligation to pay BioLine royalties will expire on a product-by-product and country-by-country basis on the date on which BCM is no longer covered by a valid claim in the licensed patent rights in a given country.

BioLine has the option, exercisable under specified circumstances, to manufacture any product containing BCM for the Company pursuant to terms to be negotiated by the parties. If BioLine exercises this option, the Company would be obligated to purchase at least a specified percentage of its BCM requirements from BioLine at a price calculated using a pre-agreed methodology.

Except under specified circumstances, the Company may not directly or indirectly acquire more than a specified percentage of the equity or debt securities of BioLine, or urge, induce, entice or solicit any other party to acquire such securities.

The Company and BioLine have the right to terminate the license agreement for an uncured material breach by the other party. In addition, the Company has the right to terminate the license agreement if at any time the Company determines that further development of products containing BCM is not warranted. See Note 13—*Commitments and Contingencies*.

(11) Related-Party Transactions

In connection with the Spin-Out, in February 2014, the Company and Ikaria entered into a separation and distribution agreement which sets forth provisions relating to the separation of the Company's business from Ikaria's other businesses. The separation and distribution agreement described the assets and liabilities that remained with or were transferred to the Company and those that remained with or were transferred to Ikaria. The separation and distribution agreement provides for a full and complete release and discharge of all liabilities between Ikaria and the Company, except as expressly set forth in the agreement. The Company and Ikaria each agreed to indemnify, defend and hold harmless the other party and its subsidiaries, and each of their respective past and present directors, officers and employees, and each of their respective permitted successors and assigns, from any and all damages relating to, arising out of or resulting from, among other things, our business and certain additional specified liabilities or Ikaria's business and certain additional specified liabilities, as applicable.

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Notes to Financial Statements (Continued)

(11) Related-Party Transactions (Continued)

In February 2014, the Company entered into a cross-license, technology transfer and regulatory matters agreement with a subsidiary of Ikaria. Pursuant to the terms of the license agreement, Ikaria granted to the Company a fully paid-up, non-royalty-bearing, exclusive license under specified intellectual property rights controlled by Ikaria to engage in the development, manufacture and commercialization of nitric oxide, devices to deliver nitric oxide and related services for or in connection with out-patient, chronic treatment of patients who have PAH, PH-COPD or idiopathic pulmonary fibrosis, or PH-IPF. Pursuant to the terms of the license agreement, the Company granted Ikaria a fully paid-up, non-royalty-bearing, exclusive license under specified intellectual property rights that the Company controls to engage in the development, manufacture and commercialization of products and services for or used in connection with the diagnosis, prevention or treatment, whether in- or out-patient, of certain conditions and diseases other than PAH, PH-COPD or PH-IPF and for the use of nitric oxide to treat or prevent conditions that are primarily managed in the hospital. The Company agreed that, during the term of the license agreement, it will not, without the prior written consent of Ikaria, grant a sublicense under any of the intellectual property licensed to the Company under the license agreement to any of its affiliates or any third party, in either case, that directly or indirectly competes with Ikaria's nitric oxide business.

In September 2013, October 2013 and February 2014, the Company and each of its subsidiaries entered into an agreement not to compete with a subsidiary of Ikaria, or, collectively, the agreements not to compete. Pursuant to the agreements not to compete, the Company and each of its subsidiaries agreed not to engage, anywhere in the world, in any manner, directly or indirectly, until the earlier of five years after the effective date of such agreement not to compete or the date on which Ikaria and all of its subsidiaries are no longer engaged in such business, in:

(1) the development, manufacture, commercialization, promotion, sale, import, export, servicing, repair, training, storage, distribution, transportation, licensing, or other handling or disposition of any product or service (including, without limitation, any product or service that utilizes, contains or includes nitric oxide for inhalation, a device intended to deliver nitric oxide or a service that delivers or supports the delivery of nitric oxide), bundled or unbundled, for or used in connection with (a) the diagnosis, prevention, or treatment, in both adult and/or pediatric populations, and whether in- or out-patient, of: (i) hypoxic respiratory failure associated with pulmonary hypertension, (ii) pulmonary hypertensive episodes and right heart failure associated with cardiovascular surgery, (iii) bronchopulmonary dysplasia, (iv) the management of ventilation—perfusion mismatch in acute lung injury, (v) the management of ventilation—perfusion mismatch in acute respiratory distress syndrome, (vi) the management of pulmonary hypertension episodes and right heart failure in congestive heart failure, (vii) pulmonary edema from high altitude sickness, (viii) the management of pulmonary hypertension episodes and right heart failure in pulmonary or cardiac surgery, (ix) the management of pulmonary hypertension episodes and right heart failure in organ transplant, (x) sickle cell vaso-occlusive crisis, (xi) hypoxia associated with pneumonia, or (xii) ischemia-reperfusion injury, or (b) the use of nitric oxide to treat or prevent conditions that are primarily managed in the hospital; or

(2) any and all development, manufacture, commercialization, promotion, sale, import, export, storage, distribution, transportation, licensing, or other handling or disposition of any terlipressin or any other product within the pressin family, (a) intended to treat (i) hepatorenal

BELLEROPHON THERAPEUTICS LLC

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Notes to Financial Statements (Continued)

(11) Related-Party Transactions (Continued)

syndrome in any form (HRS), (ii) bleeding esophageal varices, or (iii) septic shock, or (b) for or in connection with the management of low blood pressure.

In February 2014, the Company and Ikaria and entered into a transition services agreement, pursuant to which Ikaria agreed to use commercially reasonable efforts to provide certain transition services to the Company for a two-year period, which services include management/executive, human resources, real estate, information technology, accounting, financial planning and analysis, legal, quality and regulatory support. Ikaria also has agreed to use reasonable efforts to provide the Company with the use of office space at Ikaria's headquarters in Hampton, New Jersey pursuant to the terms of the transition services agreement. In exchange for the services, beginning in February 2014, the Company is obligated to pay Ikaria monthly services fees in the amount of $772,000 plus out of pocket expenses and certain other expenses. At the time of the Spin-Out, the Company deposited the sum of $18.5 million, representing the aggregate of the $772,000 monthly service fees payable by the Company under the transition services agreement, in escrow to guarantee payment of the monthly services fees by the Company. The escrowed cash will be classified as restricted cash in periods subsequent to the Spin-Out.

In February 2014, the Company entered into drug supply and device supply agreements with a subsidiary of Ikaria. Under these agreements, Ikaria has agreed to use commercially reasonable efforts to supply inhaled nitric oxide and nitric oxide delivery devices for use in the Company's clinical trials, in each case at Ikaria's manufacturing cost plus a 20% mark-up, and in the case of the drug supply agreement, the Company has agreed to purchase its clinical supply of inhaled nitric oxide from Ikaria. The Company also granted Ikaria a right of first negotiation in the event that the Company desires to enter into a commercial supply agreement with a third party for supply of nitric oxide for inhalation.

In February 2014, the Company and Ikaria entered into an employee matters agreement, pursuant to which the employment of certain Ikaria employees was transferred to the Company or its subsidiaries on the terms and conditions set forth in the employee matters agreement. Under the employee matters agreement, the Company agreed to assume and pay, perform, fulfill and discharge, in accordance with the terms of the employee matters agreement, all liabilities to or relating to such transferred employees.

(12) Segments and Geographic Information

The Company operates in one reportable segment and solely within the United States. Accordingly, no segment or geographic information has been presented.

(13) Commitments and Contingencies

Legal Proceedings

The Company periodically becomes subject to legal proceedings and claims arising in connection with its business. The ultimate legal and financial liability of the Company in respect to all proceedings, claims and lawsuits, pending or threatened, cannot be estimated with any certainty.

BioLine has indicated to the Company that it believes that the Company has breached the license agreement in several ways, including, but not limited to, failure to use commercially reasonable efforts

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Notes to Financial Statements (Continued)

(13) Commitments and Contingencies (Continued)

to develop BCM, failure to provide BioLine with material information concerning the development and commercialization plans for BCM and failure to notify BioLine in advance of material public disclosures regarding BCM. The Company and BioLine also disagree about the timing of a $12.5 million milestone payment that the Company would owe BioLine based upon progress in the Company's BCM clinical development program. The Company believes it has complied with its obligations under the license agreement to use commercially reasonable efforts to develop BCM and is not currently in breach of its other obligations under the license agreement. Although the Company has had multiple discussions with BioLine on these issues, and these discussions are continuing, the Company has not been able to resolve these outstanding issues. If the Company is unable to reach agreement with BioLine on these issues, BioLine could bring a lawsuit against the Company, although any claims relating to the Company's alleged failure to use commercially reasonable efforts would first be subject to a non-binding 60-day mediation period with a third-party mediator. The Company believes that it would have strong defenses in any litigation that could be brought by BioLine. If BioLine were to prevail in any such litigation, one of the potential remedies would be the return of BCM to BioLine. No amounts have been accrued for this matter since no loss is probable as of December 31, 2013.

If BioLine were to prevail in any such litigation, or if the Company were required to pay the milestone in dispute sooner than planned, or if we were required to return BCM to BioLine, these events could have a material adverse effect on the Company's business, results of operations, financial condition and/or liquidity.

As of this report, other than as described above, there is no proceeding, claim or litigation, pending or threatened, that could, individually or in the aggregate, have a material adverse effect on the Company's business, operating results, financial condition and/or liquidity.

Operating Lease

The Company leases an operating facility located in North Brunswick, New Jersey under an operating lease arrangement. Future minimum rental commitments under the Company's non-cancellable operating lease in effect as of December 31, 2013 are as follows (in thousands):

2014	$ 28
Thereafter	—
Total	$ 28

The amounts in the table do not include (i) approximately $100,000 of milestone rent payable upon the closing of an initial public offering by the Company or (ii) our rent obligation of $113,400 through March 15, 2015, under a lease that the Company signed subsequent to December 31, 2013.

Rent expense, including direct and allocated expenses, is calculated on the straight-line basis and amounted to approximately $0.5 million for each of the years ended December 31, 2013 and 2012.

BELLEROPHON THERAPEUTICS LLC

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Notes to Financial Statements (Continued)

(14) Equity Adjustments, Bellerophon Spin-Out and Merger

Equity Adjustments

Stock Options

In February 2014, prior to the Spin-Out, each Ikaria stock option, other than options held by non-accredited investors who were also not employees of Ikaria, was adjusted such that it became an option to acquire the same number of shares of Ikaria non-voting common stock as were subject to the Ikaria stock option, or an Adjusted Ikaria Option, and an option to acquire the same number of non-voting limited liability company units of the Company as the number of shares of Ikaria non-voting common stock that were subject to the Ikaria stock option, or a Bellerophon Option. The exercise price of each option was adjusted by allocating the relative post Spin-Out estimated fair values of Ikaria and the Company in a ratio of 85% and 15%, respectively, reflecting the relative value of each entity. The expiration date of the options was not modified. In connection with the Spin-Out, each Adjusted Ikaria Option and each Bellerophon Option was fully accelerated.

Restricted Stock Units

In February 2014, prior to the Spin-Out, each Ikaria RSU was adjusted such that it became an RSU with respect to the same number of shares of Ikaria non-voting common stock as were subject to the Ikaria RSU, or an Adjusted Ikaria RSU, and an RSU with respect to the same number of non-voting limited liability company units of the Company as were subject to the Ikaria RSU. In connection with the Spin-Out, the vesting of each Adjusted Ikaria RSU and Bellerophon RSU was fully accelerated.

Bellerophon Spin-Out

On February 12, 2014, prior to the Merger, Ikaria distributed all of the Company's outstanding units to Ikaria's stockholders in a pro rata distribution through a special dividend. In the Spin-Out, each holder of Ikaria common stock received one voting limited liability company interest in the Company for each share of Ikaria common stock held. Following acceleration of the vesting of the Bellerophon RSUs, each Bellerophon RSU was settled through delivery of one non-voting limited liability company interest in the Company.

Merger

On February 12, 2014, through a series of merger subsidiary transactions, MDP acquired a majority ownership of Ikaria and Ikaria's existing shareholders retained a minority ownership position in Ikaria. In connection with the Merger, all amounts due under Ikaria's LTIP and special dividend bonus plan became vested and were settled in cash for $11.1 million and $34.7 million, respectively.

(15) Unaudited Pro Forma Balance Sheet

The unaudited pro forma balance sheet gives effect to the following transactions, as if each occurred on December 31, 2013:

i. Ikaria's distribution of $80.0 million of cash to the Company in connection with the Spin-Out, including $18.5 million of restricted cash for the guaranteed payment to Ikaria for services to

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Notes to Financial Statements (Continued)

(15) Unaudited Pro Forma Balance Sheet (Continued)

be provided in connection with the transition services agreement, with $9.25 million classified as current and $9.25 million classified as non-current; and

ii. Ikaria's payment of the Ikaria special dividend bonus amounts in connection with the Merger.

(16) Subsequent Events

The Company has evaluated events from the balance sheet date through May 14, 2014, the date at which the financial statements were available to be issued. There were no material subsequent events that required recognition or disclosure in these financial statements, except for the disclosures included in Note 1—*Organization and Nature of the Business*, Note 11—*Related-Party Transactions* and Note 14—*Equity Adjustments, Bellerophon Spin-Out and Merger*.

Shares

Common Stock

Leerink Partners **Cowen and Company**

, 2014

Until , (25 days after the date of this prospectus) all dealers that buy, sell or trade in our common stock, whether or not participating in this offering, may be required to deliver a prospectus. This is in addition to the dealers' obligation to deliver a prospectus when acting as underwriters and with respect to their unsold allotments or subscriptions.

Part II

INFORMATION NOT REQUIRED IN PROSPECTUS

Item 13. Other Expenses of Issuance and Distribution.

The following table sets forth the expenses to be incurred in connection with the offering described in this registration statement, other than underwriting discounts and commissions, all of which will be paid by us. All amounts are estimates except the Securities and Exchange Commission's registration fee and the Financial Industry Regulatory Authority, Inc., or FINRA, filing fee and The NASDAQ Global Market listing fee.

	Amount
Securities and Exchange Commission registration fee	*
FINRA filing fee	*
NASDAQ Global Market listing fee	*
Accountants' fees and expenses	*
Legal fees and expenses	*
Blue Sky fees and expenses	*
Transfer Agent's fees and expenses	*
Printing and engraving expenses	*
Miscellaneous fees and expenses	*
Total expenses	$ *

* To be filed by amendment.

Item 14. Indemnification of Directors and Officers.

Section 102 of the Delaware General Corporation Law permits a corporation to eliminate the personal liability of its directors or its stockholders for monetary damages for a breach of fiduciary duty as a director, except where the director breached his or her duty of loyalty, failed to act in good faith, engaged in intentional misconduct or knowingly violated a law, authorized the payment of a dividend or approved a stock repurchase in violation of Delaware corporate law or obtained an improper personal benefit. Our certificate of incorporation provides that no director shall be personally liable to us or our stockholders for monetary damages for any breach of fiduciary duty as a director, notwithstanding any provision of law imposing such liability, except to the extent that the Delaware General Corporation Law prohibits the elimination or limitation of liability of directors for breaches of fiduciary duty.

Section 145 of the General Corporation Law of the State of Delaware provides that a corporation has the power to indemnify a director, officer, employee, or agent of the corporation and certain other persons serving at the request of the corporation in related capacities against expenses (including attorneys' fees), judgments, fines and amounts paid in settlements actually and reasonably incurred by the person in connection with an action, suit or proceeding to which he or she is or is threatened to be made a party by reason of such position, if such person acted in good faith and in a manner he or she reasonably believed to be in or not opposed to the best interests of the corporation, and, in any criminal action or proceeding, had no reasonable cause to believe his or her conduct was unlawful, except that, in the case of actions brought by or in the right of the corporation, no indemnification shall be made with respect to any claim, issue or matter as to which such person shall have been adjudged to be liable to the corporation unless and only to the extent that the Court of Chancery or other adjudicating court determines that, despite the adjudication of liability but in view of all of the circumstances of the case, such person is fairly and reasonably entitled to indemnity for such expenses which the Court of Chancery or such other court shall deem proper.

Upon completion of this offering, our certificate of incorporation provides that we will indemnify each person who was or is a party or threatened to be made a party to any threatened, pending or completed action, suit or proceeding, whether civil, criminal, administrative or investigative (other than an action by or in the right of us) by reason of the fact that he or she is or was, or has agreed to become, our director or officer, or is or was serving, or has agreed to serve, at our request as a director, officer, partner, employee or trustee of, or in a similar capacity with, another corporation, partnership, joint venture, trust or other enterprise (all such persons being referred to as an "Indemnitee"), or by reason of any action alleged to have been taken or omitted in such capacity, against all expenses (including attorneys' fees), judgments, fines and amounts paid in settlement actually and reasonably incurred in connection with such action, suit or proceeding and any appeal therefrom, if such Indemnitee acted in good faith and in a manner he or she reasonably believed to be in, or not opposed to, our best interests, and, with respect to any criminal action or proceeding, he or she had no reasonable cause to believe his or her conduct was unlawful. Our certificate of incorporation also provides that we will indemnify any Indemnitee who was or is a party to an action or suit by or in the right of us to procure a judgment in our favor by reason of the fact that the Indemnitee is or was, or has agreed to become, our director or officer, or is or was serving, or has agreed to serve, at our request as a director, officer, partner, employee or trustee of, or in a similar capacity with, another corporation, partnership, joint venture, trust or other enterprise, or by reason of any action alleged to have been taken or omitted in such capacity, against all expenses (including attorneys' fees) and, to the extent permitted by law, amounts paid in settlement actually and reasonably incurred in connection with such action, suit or proceeding, and any appeal therefrom, if the Indemnitee acted in good faith and in a manner he or she reasonably believed to be in, or not opposed to, our best interests, except that no indemnification shall be made with respect to any claim, issue or matter as to which such person shall have been adjudged to be liable to us, unless a court determines that, despite such adjudication but in view of all of the circumstances, he or she is entitled to indemnification of such expenses. Notwithstanding the foregoing, to the extent that any Indemnitee has been successful, on the merits or otherwise, he or she will be indemnified by us against all expenses (including attorneys' fees) actually and reasonably incurred by him or her or on his or her behalf in connection therewith. If we do not assume the defense, expenses must be advanced to an Indemnitee under certain circumstances.

We intend to enter into indemnification agreements with our directors and executive officers prior to the closing of this offering. In general, these agreements will provide that we will indemnify the director or executive officer to the fullest extent permitted by law for claims arising in his or her capacity as a director or officer of our company or in connection with their service at our request for another corporation or entity. The indemnification agreements will also provide for procedures that will apply in the event that a director or executive officer makes a claim for indemnification and establish certain presumptions that are favorable to the director or executive officer.

We maintain a general liability insurance policy which covers certain liabilities of our directors and officers arising out of claims based on acts or omissions in their capacities as directors or officers.

The underwriting agreement we will enter into in connection with the offering of common stock being registered hereby provides that the underwriters will indemnify, under certain conditions, our directors and officers (as well as certain other persons) against certain liabilities arising in connection with such offering.

Item 15. Recent Sales of Unregistered Securities.

Except as set forth below, in the three years preceding the filing of this registration statement, we have not issued any securities that were not registered under the Securities Act of 1933, as amended, or the Securities Act.

On February 9, 2014, we, Ikaria, Inc., or Ikaria, and Ikaria Acquisition Inc. entered into a separation and distribution agreement which provided for and contained the key terms of our separation from Ikaria, which we refer to as the Spin-Out. Prior to the Spin-Out, we issued to certain employees and directors of ours or of our then-parent company, Ikaria, and certain accredited investors, options to purchase an aggregate of 7,744,480 of our non-voting membership units, at a weighted average exercise price of $0.58 per share.

Prior to the Spin-Out, we issued to certain employees and directors of ours or of Ikaria and certain accredited investors restricted stock units in respect of an aggregate of 4,672,001 of our non-voting membership units, which we refer to as the Bellerophon RSUs. We subsequently settled such Bellerophon RSUs by issuing and delivering an aggregate of 4,672,001 non-voting membership units to the holders of Bellerophon RSUs.

Each of the foregoing issuances was made by us in a transaction not involving a public offering pursuant to an exemption from the registration requirements of the Securities Act in reliance upon Section 4(a)(2) of the Securities Act or Rule 701 promulgated under Section 3(b) of the Securities Act. We did not pay or give, directly or indirectly, any commission or other remuneration, including underwriting discounts or commissions, in connection with any of the issuances of securities listed above, and no underwriters were involved in the foregoing issuances of securities. All recipients either received adequate information about the registrant or had access, through employment or other relationships, to such information.

Item 16. Exhibits and Financial Statement Schedules.

The exhibits to the registration statement are listed in the Exhibit Index attached hereto and incorporated by reference herein.

Item 17. Undertakings.

(a) The undersigned registrant hereby undertakes to provide to the underwriters at the closing specified in the underwriting agreement, certificates in such denominations and registered in such names as required by the underwriter to permit prompt delivery to each purchaser.

(b) Insofar as indemnification for liabilities arising under the Securities Act may be permitted to directors, officers and controlling persons of the registrant pursuant to the foregoing provisions, or otherwise, the registrant has been advised that in the opinion of the Securities and Exchange Commission such indemnification is against public policy as expressed in the Securities Act and is, therefore, unenforceable. In the event that a claim for indemnification against such liabilities (other than the payment by the registrant of expenses incurred or paid by a director, officer or controlling person of the registrant in the successful defense of any action, suit or proceeding) is asserted by such director, officer or controlling person in connection with the securities being registered, the registrant will, unless in the opinion of its counsel the matter has been settled by controlling precedent, submit to a court of appropriate jurisdiction the question whether such indemnification by it is against public policy as expressed in the Securities Act and will be governed by the final adjudication of such issue.

(c) The undersigned registrant hereby undertakes that:

(1) For purposes of determining any liability under the Securities Act, the information omitted from the form of prospectus filed as part of this registration statement in reliance upon Rule 430A and contained in a form of prospectus filed by the registrant pursuant to Rule 424(b)(1) or (4) or 497(h) under the Securities Act shall be deemed to be part of this registration statement as of the time it was declared effective.

(2) For the purpose of determining any liability under the Securities Act, each post-effective amendment that contains a form of prospectus shall be deemed to be a new registration statement relating to the securities offered therein, and the offering of such securities at that time shall be deemed to be the initial bona fide offering thereof.

SIGNATURES

Pursuant to the requirements of the Securities Act of 1933, the registrant has duly caused this registration statement to be signed on its behalf by the undersigned, thereunto duly authorized, in the Borough of Hampton, State of New Jersey, on this day of , 2014.

BELLEROPHON THERAPEUTICS LLC

By: _____

Daniel Tassé
Chief Executive Officer

SIGNATURES AND POWER OF ATTORNEY

We, the undersigned officers and directors of Bellerophon Therapeutics LLC, hereby severally constitute and appoint , and , and each of them singly (with full power to each of them to act alone), our true and lawful attorneys-in-fact and agents, with full power of substitution and resubstitution in each of them for him and in his name, place and stead, and in any and all capacities, to sign any and all amendments (including post-effective amendments) to this registration statement (or any other registration statement for the same offering that is to be effective upon filing pursuant to Rule 462(b) under the Securities Act of 1933), and to file the same, with all exhibits thereto and other documents in connection therewith, with the Securities and Exchange Commission, granting unto said attorneys-in-fact and agents, and each of them, full power and authority to do and perform each and every act and thing requisite or necessary to be done in and about the premises, as full to all intents and purposes as he might or could do in person, hereby ratifying and confirming all that said attorneys-in-fact and agents or any of them, or their or his substitute or substitutes, may lawfully do or cause to be done by virtue hereof.

Pursuant to the requirements of the Securities Act of 1933, this registration statement has been signed by the following persons in the capacities held on the dates indicated.

Signature	Title	Date
_____ Daniel Tassé	Chief Executive Officer and Director (principal executive officer)	
_____ David Abrams	Treasurer (principal financial and accounting officer)	
_____ Aldo E. Belloni, Ph.D.	Director	
_____ Matthew Holt	Director	

Signature	Title	Date
Andre V. Moura	Director	
Robert R. Nelsen	Director	
Howard Pien	Director	
Adam Weinstein	Director	

Exhibit Index

Exhibit Number	Description of Exhibit
1.1*	Underwriting Agreement
2.1*	Plan of Conversion
3.1*	Certificate of Incorporation of the Registrant
3.2*	Bylaws of the Registrant
3.3*	Form of Certificate of Incorporation of the Registrant (to be effective upon the closing of this offering)
3.4*	Form of Bylaws of the Registrant (to be effective upon the closing of this offering)
4.1*	Specimen Stock Certificate evidencing the shares of common stock
4.2*	Stockholders Agreement, dated , 2014
5.1*	Opinion of Wilmer Cutler Pickering Hale and Dorr LLP
10.1	Assumed 2007 Ikaria Stock Option Plan
10.2	Assumed 2010 Ikaria Equity Incentive Plan
10.3*	2014 Equity Incentive Plan
10.4*	Form of Option Agreement under 2014 Equity Incentive Plan
10.5*	Public Company Stock Incentive Plan
10.6*	Form of Incentive Stock Option Agreement under Public Company Stock Incentive Plan
10.7*	Form of Nonstatutory Stock Option Agreement under Public Company Stock Incentive Plan
10.8†	Amended and Restated License and Commercialization Agreement, dated as of August 26, 2009, among Ikaria Development Subsidiary One LLC, BioLineRx Ltd. and BioLine Innovations Jerusalem L.P., as amended by the Payment Date Extension Amendment and the Amendment to the Amended and Restated License and Commercialization Agreement effective as of April 21, 2010
10.9	Form of Agreement Not to Compete, entered into by Ikaria Acquisition LLC and each of the Registrant, Bellerophon BCM LLC, Bellerophon Pulse Technologies LLC and Bellerophon Services, Inc.
10.10†	Separation and Distribution Agreement, dated as of February 9, 2014, among the Registrant, Ikaria, Inc. and Ikaria Acquisition LLC
10.11†	Device Clinical Supply Agreement, dated as of February 9, 2014, between Bellerophon Pulse Technologies LLC and INO Therapeutics LLC
10.12†	Drug Clinical Supply Agreement, dated as of February 9, 2014, between Bellerophon Pulse Technologies LLC and INO Therapeutics LLC
10.13†	Employee Matters Agreement, dated as of February 9, 2014, between the Registrant and Ikaria, Inc.
10.14†	Exclusive Cross-License, Technology Transfer and Regulatory Matters Agreement, dated February 9, 2014, between Bellerophon Pulse Technologies LLC and INO Therapeutics LLC, as amended on March 27, 2014

Exhibit Number	Description of Exhibit
10.15†	Transition Services Agreement, dated as of February 9, 2014, between the Registrant and Ikaria, Inc.
10.16*	Registration Rights Agreement, dated as of February 12, 2014, among the Registrant, New Mountain Partners II (AIV-A), L.P., Allegheny New Mountain Partners, L.P., New Mountain Affiliated Investors II, L.P., IRDO Holding Corp., Venrock IK Holdings BT, Inc., Linde North Amerrica, Inc., 5AM-BT, Inc. and Aravis Venture I L.P.
10.17*	Voting Agreement, dated as of February 12, 2014, among the Registrant, New Mountain Partners II (AIV-A), L.P., Allegheny New Mountain Partners, L.P., New Mountain Affiliated Investors II, L.P., IRDO Holding Corp., Venrock IK Holdings BT, Inc., Linde North America, Inc., 5AM-BT, Inc. and Aravis Venture I L.P.
10.18*	Form of Indemnification Agreement
10.19*	Assumed Employment Agreement, dated January 4, 2012, between Manesh Naidu and Ikaria, Inc.
10.20*	Assumed Employment Agreement, dated August 10, 2008, between Martin Meglasson and Ikaria, Inc.
10.21*	Assumed Employment Agreement, dated March 26, 2012, between Reinilde Heyrman and Ikaria, Inc.
10.22	Form of Retention Bonus Letter for Executive Officers
21.1	Subsidiaries of the Registrant
23.1*	Consent of KPMG LLP independent registered public accounting firm
23.2*	Consent of Wilmer Cutler Pickering Hale and Dorr LLP (included in Exhibit 5.1)
24.1	Power of Attorney (included on signature page)

* To be filed by amendment.

† Confidential treatment requested as to portions of the exhibit. Confidential materials omitted and filed separately with the Securities and Exchange Commission.

Exhibit 10.1

ASSUMED IKARIA HOLDINGS, INC.

2007 STOCK OPTION PLAN

1. **Purposes of the Plan.** The purposes of this 2007 Stock Option Plan are to attract and retain the best available personnel for positions of substantial responsibility, to provide additional incentive to Employees and Consultants and to promote the success of the business of the Company and any Subsidiary, Parent or Affiliate. Options granted under the Plan may be Incentive Stock Options or Nonstatutory Stock Options, as determined by the Administrator at the time of grant of an option and subject to the applicable provisions of Section 422 of the Code and the regulations and interpretations promulgated thereunder.

2. **Definitions.** As used herein, the following definitions shall apply:

(a) **"Administrator"** means the Board or its Committee appointed pursuant to Section 4 of the Plan.

(b) **"Affiliate"** means an entity other than a Subsidiary (as defined below) which, together with the Company, is under common control of a third person or entity.

(c) **"Applicable Laws"** means the legal requirements relating to the administration of stock option and restricted stock purchase plans, including under applicable U.S. state corporate laws, U.S. federal and applicable state securities laws, other U.S. federal and state laws, the Code, any Stock Exchange rules or regulations and the applicable laws, rules and regulations of any other country or jurisdiction where Options are granted under the Plan, as such laws, rules, regulations and requirements shall be in place from time to time.

(d) **"Board"** means the Board of Directors of the Company.

(e) **"Cause"** for termination of a Participant's Continuous Service Status will exist if the Participant is terminated by the Company for any of the following reasons: (i) Participant's willful failure substantially to perform his or her duties and responsibilities to the Company or deliberate violation of a Company policy; (ii) Participant's commission of any act of fraud, embezzlement, dishonesty or any other willful misconduct that has caused or is reasonably expected to result in material injury to the Company; (iii) unauthorized use or disclosure by Participant of any proprietary information or trade secrets of the Company or any other party to whom the Participant owes an obligation of nondisclosure as a result of his or her relationship with the Company; or (iv) Participant's willful breach of any of his or her obligations under any written agreement or covenant with the Company. The determination as to whether a Participant is being terminated for Cause shall be made in good faith by the Company and shall be final and binding on the Participant. The foregoing definition does not in any way limit the Company's ability to terminate a Participant's employment or consulting relationship at any time as provided in Section 5(d) below, and the term "Company" will be interpreted to include any Subsidiary, Parent or Affiliate, as appropriate.

(f) **"Change of Control"** means (1) a sale of all or substantially all of the Company's assets, or (2) any merger, consolidation or other business combination transaction of the Company with or into another corporation, entity or person, other than a transaction in which

the holders of at least a majority of the shares of voting capital stock of the Company outstanding immediately prior to such transaction continue to hold (either by such shares remaining outstanding or by their being converted into shares of voting capital stock of the surviving entity) a majority of the total voting power represented by the shares of voting capital stock of the Company (or the surviving entity) outstanding immediately after such transaction, or (3) the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of capital stock of the Company; provided, however, that none of the following shall be considered a Change of Control: (i) a merger effected exclusively for the purpose of changing the domicile of the Company, (ii) an equity financing in which the Company is the surviving corporation, or (iii) a transaction in which the stockholders of the Company immediately prior to the transaction own 50% or more of the voting power of the surviving corporation immediately following the transaction. For avoidance of doubt, the term "Change of Control" does not include the Company's Series B Preferred Stock financing, the merger of a wholly owned subsidiary of the Company with Ikaria, Inc., or the acquisition of INO Therapeutics LLC by an Affiliate of the Company (collectively, the "Ikaria-INO Transactions").

(g) **"Code"** means the Internal Revenue Code of 1986, as amended.

(h) **"Committee"** means one or more committees or subcommittees of the Board appointed by the Board to administer the Plan in accordance with Section 4 below.

(i) Prior to an Initial Public Offering (as such term is defined in the Company's Amended and Restated Certificate of Incorporation as may be amended from time to time), the term **"Common Stock"** shall mean the Non-Voting Common Stock, par value $0.01 per share, of the Company. Upon and following an Initial Public Offering, the term **"Common Stock"** shall mean the Voting Common Stock, par value $0.01 per share, of the Company and all Options outstanding at the time of the Initial Public Offering (to the extent not previously exercised) shall automatically convert into options to purchase an equivalent number of shares of the Company's Voting Common Stock without any further action on the part of the Company or any Optionee.

(j) **"Company"** means Ikaria Holdings, Inc., a Delaware corporation.

(k) **"Consultant"** means any person, including an advisor, who is engaged by the Company or any Parent, Subsidiary or Affiliate to render services and is compensated for such services, and any director of the Company whether compensated for such services or not.

(l) **"Continuous Service Status"** means the absence of any interruption or termination of service as an Employee or Consultant. Continuous Service Status as an Employee or Consultant shall not be considered interrupted in the case of: (i) sick leave; (ii) military leave; (iii) any other leave of absence approved by the Administrator, provided that such leave is for a period of not more than ninety (90) days, unless reemployment upon the expiration of such leave is guaranteed by contract or statute, or unless provided otherwise pursuant to any policy of the Company or any Subsidiary, Parent or Affiliate thereof that may be adopted from time to time; or (iv) in the case of transfers between locations of the Company or between the Company, its

Parents, Subsidiaries, Affiliates or their respective successors. A change in status from an Employee to a Consultant or from a Consultant to an Employee will not constitute an interruption of Continuous Service Status.

(m) **"Corporate Transaction"** means a sale of all or substantially all of the Company's assets, or a merger, consolidation or other capital reorganization or business combination transaction of the Company with or into another corporation, entity or person, or the direct or indirect acquisition (including by way of a tender or exchange offer) by any person, or persons acting as a group, of beneficial ownership or a right to acquire beneficial ownership of shares representing a majority of the voting power of the then outstanding shares of capital stock of the Company.

(n) **"Director"** means a member of the Board.

(o) **"Employee"** means any person employed by the Company or any Parent, Subsidiary or Affiliate, with the status of employment determined based upon such factors as are deemed appropriate by the Administrator in its discretion, subject to any requirements of the Code or the Applicable Laws. The payment by the Company of a director's fee to a Director shall not be sufficient to constitute "employment" of such Director by the Company.

(p) **"Exchange Act"** means the Securities Exchange Act of 1934, as amended.

(q) **"Fair Market Value"** means, as of any date, the fair market value of the Common Stock, as determined by the Administrator in good faith on such basis as it deems appropriate and applied consistently with respect to Participants. Whenever possible, the determination of Fair Market Value shall be based upon the closing price for the Shares as reported in the Wall Street Journal for the applicable date.

(r) **"Incentive Stock Option"** means an Option intended to qualify as an incentive stock option within the meaning of Section 422 of the Code, as designated in the applicable Option Agreement.

(s) **"Listed Security"** means any security of the Company that is listed or approved for listing on a national securities exchange or designated or approved for designation as a national market system security on an interdealer quotation system by the National Association of Securities Dealers, Inc.

(t) **"Named Executive"** means any individual who, on the last day of the Company's fiscal year, is the chief executive officer of the Company (or is acting in such capacity) or among the four most highly compensated officers of the Company (other than the chief executive officer). Such officer status shall be determined pursuant to the executive compensation disclosure rules under the Exchange Act.

(u) **"Nonstatutory Stock Option"** means an Option not intended to qualify as an Incentive Stock Option, as designated in the applicable Option Agreement.

(v) **"Option"** means a stock option granted pursuant to the Plan.

(w) **"Option Agreement"** means a written document, the form(s) of which shall be approved from time to time by the Administrator, reflecting the terms of an Option granted under the Plan and includes any documents attached to or incorporated into such Option Agreement, including, but not limited to, a notice of stock option grant and a form of exercise notice.

(x) **"Option Exchange Program"** means a program approved by the Administrator whereby outstanding Options are exchanged for Options with a lower exercise price or are amended to decrease the exercise price as a result of a decline in the Fair Market Value of the Common Stock.

(y) **"Optioned Stock"** means the Common Stock subject to an Option,

(z) **"Optionee"** means an Employee or Consultant who receives an Option.

(aa) **"Parent"** means a "parent corporation," whether now or hereafter existing, as defined in Section 424(e) of the Code, or any successor provision.

(bb) **"Participant"** means any holder of one or more Options, or the Shares issuable or issued upon exercise of such Options, under the Plan.

(cc) **"Plan"** means this 2007 Stock Option Plan.

(dd) **"Reporting Person"** means an officer, Director, or greater than ten percent stockholder of the Company within the meaning of Rule 16a-2 under the Exchange Act, who is required to file reports pursuant to Rule 16a-3 under the Exchange Act.

(ee) **"Rule 16b-3"** means Rule 16b-3 promulgated under the Exchange Act, as amended from time to time, or any successor provision.

(ff) **"Share"** means a share of the Common Stock, as adjusted in accordance with Section 13 of the Plan.

(gg) **"Stock Exchange"** means any stock exchange or consolidated stock price reporting system on which prices for the Common Stock are quoted at any given time.

(hh) **"Subsidiary"** means a "subsidiary corporation," whether now or hereafter existing, as defined in Section 424(f) of the Code, or any successor provision.

(ii) **"Ten Percent Holder"** means a person who owns stock representing more than ten percent (10%) of the voting power of all classes of stock of the Company or any Parent or Subsidiary.

3. **Stock Subject to the Plan.** Subject to the provisions of Section 13 of the Plan, the maximum aggregate number of Shares that may be sold under the Plan is 8,058,834 Shares of Common Stock, of which number (i) 1,504,088 Shares relate to outstanding options originally granted under the Ikaria, Inc. 2004 Stock Option Plan (the "Existing Options"), which options were exchanged for options under this Plan in connection with consummation of the Ikaria-INO

Transactions, and (ii) the remaining 6,554,746 Shares do not relate to the Existing Options. The Shares may be authorized, but unissued, or reacquired Common Stock. If an award should expire or become unexercisable for any reason without having been exercised in full, or is surrendered pursuant to an Option Exchange Program, the unpurchased Shares that were subject thereto shall, unless the Plan shall have been terminated, become available for future grant under the Plan. In addition, any Shares of Common Stock which are retained by the Company upon exercise of an award in order to satisfy the exercise or purchase price for such award or any withholding taxes due with respect to such exercise or purchase shall be treated as not issued and shall continue to be available under the Plan. Shares issued under the Plan and later repurchased by the Company pursuant to any repurchase right which the Company may have shall not be available for future grant under the Plan.

4. **Administration of the Plan.**

(a) **General.** The Plan shall be administered by the Board or a Committee, or a combination thereof, as determined by the Board. The Plan may be administered by different administrative bodies with respect to different classes of Participants and, if permitted by the Applicable Laws, the Board may authorize one or more officers to make awards under the Plan.

(b) **Committee Composition.** If a Committee has been appointed pursuant to this Section 4, such Committee shall continue to serve in its designated capacity until otherwise directed by the Board. From time to time the Board may increase the size of any Committee and appoint additional members thereof, remove members (with or without cause) and appoint new members in substitution therefor, fill vacancies (however caused) and remove all members of a Committee and thereafter directly administer the Plan, all to the extent permitted by the Applicable Laws and, in the case of a Committee administering the Plan in accordance with the requirements of Rule 16b-3 or Section 162(m) of the Code, to the extent permitted or required by such provisions. The Committee shall in all events conform to any requirements of the Applicable Laws.

(c) **Powers of the Administrator.** Subject to the provisions of the Plan and in the case of a Committee, the specific duties delegated by the Board to such Committee, the Administrator shall have the authority, in its discretion:

(i) to determine the Fair Market Value of the Common Stock, in accordance with Section 2(q) of the Plan, provided that such determination shall be applied consistently with respect to Participants under the Plan;

(ii) to select the Employees and Consultants to whom Options may from time to time be granted;

(iii) to determine whether and to what extent Options are granted;

(iv) to determine the number of Shares of Common Stock to be covered by each award granted;

(v) to approve the form(s) of agreement(s) used under the Plan;

(vi) to determine the terms and conditions, not inconsistent with the terms of the Plan, of any award granted hereunder, which terms and conditions include but are not limited to the exercise or purchase price, the time or times when awards may be exercised (which may be based on performance criteria), any vesting acceleration or waiver of forfeiture restrictions, any pro rata adjustment to vesting as a result of a Participant's transitioning from full- to part-time service (or vice versa), and any restriction or limitation regarding any Option, Optioned Stock or restricted stock issued upon exercise of an Option, based in each case on such factors as the Administrator, in its sole discretion, shall determine;

(vii) to determine whether and under what circumstances an Option may be settled in cash under Section 10(c) instead of Common Stock;

(viii) to implement an Option Exchange Program on such terms and conditions as the Administrator in its discretion deems appropriate, provided that no amendment or adjustment to an Option that would materially and adversely affect the rights of any Optionee shall be made without the prior written consent of the Optionee;

(ix) to adjust the vesting of an Option held by an Employee or Consultant as a result of a change in the terms or conditions under which such person is providing services to the Company or any Subsidiary, Parent or Affiliate thereof;

(x) to construe and interpret the terms of the Plan and awards granted under the Plan, which constructions, interpretations and decisions shall be final and binding on all Participants; and

(xi) in order to fulfill the purposes of the Plan and without amending the Plan, to modify grants of Options to Participants who are foreign nationals or employed outside of the United States in order to recognize differences in local law, tax policies or customs.

5. **Eligibility.**

(a) **Recipients of Grants.** Nonstatutory Stock Options may be granted to Employees and Consultants. Incentive Stock Options may be granted only to Employees, provided that Employees of Affiliates shall not be eligible to receive Incentive Stock Options.

(b) **Type of Option.** Each Option shall be designated in the Option Agreement as either an Incentive Stock Option or a Nonstatutory Stock Option.

(c) **ISO $100,000 Limitation.** Notwithstanding any designation under Section 5(b), to the extent that the aggregate Fair Market Value of Shares with respect to which Options designated as Incentive Stock Options are exercisable for the first time by any Optionee during any calendar year (under all plans of the Company or any Parent or Subsidiary) exceeds $100,000, such excess Options shall be treated as Nonstatutory Stock Options. For purposes of this Section 5(c), Incentive Stock Options shall be taken into account in the order in which they were granted, and the Fair Market Value of the Shares subject to an Incentive Stock Option shall be determined as of the date of the grant of such Option.

(d) **No Employment Rights.** The Plan shall not confer upon any Participant any right with respect to continuation of an employment or consulting relationship with the Company or any Subsidiary, Parent or Affiliate thereof, nor shall it interfere in any way with such Participant's right or the right of the Company or any Subsidiary, Parent or Affiliate thereof to terminate the employment or consulting relationship at any time for any reason.

6. **Term of Plan.** The Plan shall become effective upon its adoption by the Board of Directors. It shall continue in effect for a term often (10) years unless sooner terminated under Section 15 of the Plan.

7. **Term of Option.** The term of each Option shall be the term stated in the Option Agreement; provided that the term shall be no more than ten years from the date of grant thereof or such shorter term as may be provided in the Option Agreement and provided further that, in the case of an Incentive Stock Option granted to a person who at the time of such grant is a Ten Percent Holder, the term of the Option shall be five years from the date of grant thereof or such shorter term as may be provided in the Option Agreement.

8. **[Reserved.]**

9. **Option Exercise Price and Consideration.**

(a) **Exercise Price.** The per Share exercise price for the Shares to be issued pursuant to exercise of an Option shall be such price as is determined by the Administrator and set forth in the Option Agreement, but shall be subject to the following:

(i) In the case of an Incentive Stock Option

(A) granted to an Employee who at the time of grant is a Ten Percent Holder, the per Share exercise price shall be no less than 110% of the Fair Market Value per Share on the date of grant; or

(B) granted to any other Employee, the per Share exercise price shall be no less than 100% of the Fair Market Value per Share on the date of grant.

(ii) In the case of a Nonstatutory Stock Option, the per share Exercise Price shall be such price as determined by the Administrator provided that if such eligible person is, at the time of the grant of such Option, a Named Executive of the Company, the per share Exercise Price shall be no less than 100% of the Fair Market Value on the date of grant if such Option is intended to qualify as performance-based compensation under Section 162(m) of the Code.

(iii) Notwithstanding the foregoing, Options may be granted with a per Share exercise price other than as required above pursuant to a merger or other corporate transaction.

(b) **Permissible Consideration.** The consideration to be paid for the Shares to be issued upon exercise of an Option, including the method of payment, shall be determined by the Administrator (and, in the case of an Incentive Stock Option, shall be determined at the

time of grant) and may consist entirely of (1) cash; (2) check; (3) subject to any requirements of the Applicable Laws (including without limitation Section 153 of the Delaware General Corporation Law), delivery of Optionee's promissory note having such recourse, interest, security and redemption provisions as the Administrator determines to be appropriate after taking into account the potential accounting consequences of permitting an Optionee to deliver a promissory note; (4) cancellation of indebtedness; (5) other Shares that have a Fair Market Value on the date of surrender equal to the aggregate exercise price of the Shares as to which the Option is exercised, provided that in the case of Shares acquired, directly or indirectly, from the Company, such Shares must have been owned by the Optionee for more than six months on the date of surrender (or such shorter or longer period of time as may be deemed necessary in the Administrator's sole discretion to avoid risk of the incurrence of adverse accounting charges by the Company); provided, however, that this clause (5) shall only apply to Options granted after March 28, 2007 if permitted by the Administrator at the time of exercise in its sole discretion; (6) if, as of the date of exercise of an Option the Company then is permitting employees to engage in a "same-day sale" cashless brokered exercise program involving one or more brokers, through such a program that complies with the Applicable Laws (including without limitation the requirements of Regulation T and other applicable regulations promulgated by the Federal Reserve Board) and that ensures prompt delivery to the Company of the amount required to pay the exercise price and any applicable withholding taxes; or (7) any combination of the foregoing methods of payment. In making its determination as to the type of consideration to accept, the Administrator shall consider if acceptance of such consideration maybe reasonably expected to benefit the Company and the Administrator may, in its sole discretion, refuse to accept a particular form of consideration at the time of any Option exercise.

10. **Exercise of Option.**

(a) **General.**

(i) **Exercisability.** Any Option granted hereunder shall be exercisable at such times and under such conditions as determined by the Administrator, consistent with the term of the Plan and reflected in the Option Agreement, including vesting requirements and/or performance criteria with respect to the Company and/or the Optionee.

(ii) **Leave of Absence.** The Administrator shall have the discretion to determine whether and to what extent the vesting of Options shall be tolled during any unpaid leave of absence; provided, however, that in the absence of such determination, vesting of Options shall be tolled during any such unpaid leave (unless otherwise required by the Applicable Laws). In the event of military leave, vesting shall toll during any unpaid portion of such

leave, provided that, upon a Participant's returning from military leave (under conditions that would entitle him or her to protection upon such return under the Uniform Services Employment and Reemployment Rights Act), he or she shall be given vesting credit with respect to Options to the same extent as would have applied had the Participant continued to provide services to the Company or any Subsidiary, Parent or Affiliate thereof throughout the leave on the same terms as he or she was providing services immediately prior to such leave.

(iii) **Minimum Exercise Requirements.** An Option may not be exercised for a fraction of a Share. The Administrator may require that an Option be exercised

as to a minimum number of Shares, provided that such requirement shall not prevent an Optionee from exercising the full number of Shares as to which the Option is then exercisable.

(iv) **Procedures for and Results of Exercise.** An Option shall be deemed exercised when written notice of such exercise has been given to the Company in accordance with the terms of the Option by the person entitled to exercise the Option and the Company has received full payment for the Shares with respect to which the Option is exercised. Full payment may, as authorized by the Administrator, consist of any consideration and method of payment allowable under Section 9(b) of the Plan, provided that the Administrator may, in its sole discretion, refuse to accept any form of consideration at the time of any Option exercise. If required by the Option Agreement, it shall be a condition precedent to the exercise of an Option that the Optionee (and such Optionee's spouse, if applicable) execute and deliver to the Company a counterpart signature page indicating such Optionee's agreement to be bound as a "Stockholder" by the Company's Common Stockholders Agreement attached to the Option Agreement or that is in use by the Company from time to time (the "Common Stockholders Agreement").

Exercise of an Option in any manner shall result in a decrease in the number of Shares that thereafter may be available, both for purposes of the Plan and for sale under the Option, by the number of Shares as to which the Option is exercised.

(v) **Rights as Stockholder.** Until the issuance of the Shares (as evidenced by the appropriate entry on the books of the Company or of a duly authorized transfer agent of the Company) and, if required by the Stock Option Agreement, a counterpart signature page to the Common Stockholders Agreement, no right to vote or receive dividends or any other rights as a stockholder shall exist with respect to the Optioned Stock, notwithstanding the exercise of the Option. No adjustment will be made for a dividend or other right for which the record date is prior to the date the stock certificate is issued, except as provided in Section 13 of the Plan.

(b) **Termination of Employment or Consulting Relationship.** Except as otherwise set forth in this Section 10(b), the Administrator shall establish and set forth in the applicable Option Agreement the terms and conditions upon which an Option shall remain exercisable, if at all, following termination of an Optionee's Continuous Service Status, which provisions may be waived or modified by the Administrator at any time. Unless the Administrator otherwise provides in the Option Agreement, to the extent that the Optionee is not vested in Optioned Stock at the date of termination of his or her Continuous Service Status, or if the Optionee (or other person entitled to exercise the Option) does not exercise the Option to the extent so entitled within the time specified in the Option Agreement or below (as applicable), the Option shall terminate and the Optioned Stock underlying the unexercised portion of the Option shall revert to the Plan. In no event may any Option be exercised after the expiration of the Option term as set forth in the Option Agreement (and subject to Section 7).

The following provisions (1) shall apply to the extent an Option Agreement does not specify the terms and conditions upon which an Option shall terminate upon termination of an Optionee's Continuous Service Status, and (2) establish the minimum post-termination exercise periods that may be set forth in an Option Agreement:

(i) **Termination other than Upon Disability or Death or for Cause.** In the event of termination of Optionee's Continuous Service Status other than under the circumstances set forth in subsections (ii) through (iv) below, such Optionee may exercise an Option for ninety (90) days following such termination to the extent the Optionee was vested in the Optioned Stock as of the date of such termination. No termination shall be deemed to occur and this Section 10(b)(i) shall not apply if (i) the Optionee is a Consultant who becomes an Employee, or (ii) the Optionee is an Employee who becomes a Consultant.

(ii) **Disability of Optionee.** In the event of termination of an Optionee's Continuous Service Status as a result of his or her disability (including a disability within the meaning of Section 22(e)(3) of the Code), such Optionee may exercise an Option at any time within six months following such termination to the extent the Optionee was vested in the Optioned Stock as of the date of such termination.

(iii) **Death of Optionee.** In the event of the death of an Optionee during the period of Continuous Service Status since the date of grant of the Option, or within ninety (90) days following termination of Optionee's Continuous Service Status, the Option may be exercised by Optionee's estate or by a person who acquired the right to exercise the Option by bequest or inheritance at any time within six months following the date of death, but only to the extent the Optionee was vested in the Optioned Stock as of the date of death or, if earlier, the date the Optionee's Continuous Service Status terminated.

(iv) **Termination for Cause.** In the event of termination of an Optionee's Continuous Service Status for Cause, any Option (including any exercisable portion thereof) held by such Optionee shall immediately terminate in its entirety upon first notification to the Optionee of termination of the Optionee's Continuous Service Status. If an Optionee's employment or consulting relationship with the Company or any Subsidiary, Parent or Affiliate thereof is suspended pending an investigation of whether the Optionee shall be terminated for Cause, all the Optionee's rights under any Option likewise shall be suspended during the investigation period and the Optionee shall have no right to exercise any Option. Nothing in this Section 10(b)(iv) shall in any way limit the Company's right to purchase unvested Shares issued upon exercise of an Option as set forth in the applicable Option Agreement.

(c) **Buyout Provisions.** The Administrator may at any time offer to buy out for a payment in cash or Shares an Option previously granted under the Plan based on such terms and conditions as the Administrator shall establish and communicate to the Optionee at the time that such offer is made.

11. **Taxes.**

(a) As a condition of the grant, vesting or exercise of an Option granted under the Plan, the Participant (or in the case of the Participant's death, the person exercising the Option) shall make such arrangements as the Administrator may require for the satisfaction of any applicable federal, state, local

or foreign withholding tax obligations that may arise in connection with such grant, vesting or exercise of the Option or the issuance of Shares. The Company shall not be required to issue any Shares under the Plan until such obligations are satisfied. If the Administrator allows the withholding or surrender of Shares to satisfy a

Participant's tax withholding obligations under this Section 11 (whether pursuant to Section11(c), (d) or (e), or otherwise), the Administrator shall not allow Shares to be withheld in an amount that exceeds the minimum statutory withholding rates for federal and state tax purposes, including payroll taxes.

(b) In the case of an Employee and in the absence of any other arrangement, the Employee shall be deemed to have directed the Company or a Subsidiary, Parent or Affiliate thereof, as applicable, to withhold or collect from his or her compensation an amount sufficient to satisfy such tax obligations from the next payroll payment otherwise payable after the date of an exercise of the Option.

(c) This Section 11(c) shall apply only after the date, if any, upon which the Common Stock becomes a Listed Security. In the case of Participant other than an Employee (or in the case of an Employee where the next payroll payment is not sufficient to satisfy such tax obligations, with respect to any remaining tax obligations), in the absence of any other arrangement and to the extent permitted under the Applicable Laws, the Participant shall be deemed to have elected to have the Company withhold from the Shares to be issued upon exercise of the Option that number of Shares having a Fair Market Value determined as of the applicable Tax Date (as defined below) equal to the amount required to be withheld; provided, however, that that this sentence shall only apply to Options granted after March 28, 2007 if permitted by the Administrator at the time of exercise in its sole discretion. For purposes of this Section 11, the Fair Market Value of the Shares to be withheld shall be determined on the date that the amount of tax to be withheld is to be determined under the Applicable Laws (the "Tax Date").

(d) If permitted by the Administrator, in its discretion, a Participant may satisfy his or her tax withholding obligations upon exercise of an Option by surrendering to the Company Shares that have a Fair Market Value determined as of the applicable Tax Date equal to the amount required to be withheld. In the case of shares previously acquired from the Company that are surrendered under this Section 11 (d), such Shares must have been owned by the Participant for more than six (6) months on the date of surrender (or such other period of time as is required for the Company to avoid adverse accounting charges).

(e) Any election or deemed election by a Participant to have Shares withheld to satisfy tax withholding obligations under Section 11(c) or (d) above shall be irrevocable as to the particular Shares as to which the election is made and shall be subject to the consent or disapproval of the Administrator. Any election by a Participant under Section 11(d) above must be made on or prior to the applicable Tax Date.

(f) In the event an election to have Shares withheld is made by a Participant and the Tax Date is deferred under Section 83 of the Code because no election is filed under Section 83(b) of the Code, the Participant shall receive the full number of Shares with respect to which the Option is exercised but such Participant shall be unconditionally obligated to tender back to the Company the proper number of Shares on the Tax Date.

12. **Non-Transferability of Options.**

(a) **General.** Except as set forth in this Section 12, Options may not be sold, pledged, assigned, hypothecated, transferred or disposed of in any manner other than by will or by the laws of descent or distribution. The designation of a beneficiary by an Optionee will not constitute a transfer. An Option may be exercised, during the lifetime of the holder of an Option, only by such holder or a transferee permitted by this Section 12.

(b) **Limited Transferability Rights.** Notwithstanding anything else in this Section 12, the Administrator may in its discretion grant Nonstatutory Stock Options that may be transferred by instrument to an inter vivos or testamentary trust in which the Options are to be passed to beneficiaries upon the death of the trustor (settlor) or by gift or pursuant to domestic relations orders to "Immediate Family Members" (as defined below) of the Optionee. "Immediate Family" means any child, stepchild, grandchild, parent, stepparent, grandparent, spouse, former spouse, sibling, niece, nephew, mother-in-law, father-in-law, son-in-law, daughter-in-law, brother-in-law, or sister-in-law (including adoptive relationships), a trust in which these persons have more than fifty percent of the beneficial interest, a foundation in which these persons (or the Optionee) control the management of assets, and any other entity in which these persons (or the Optionee) own more than fifty percent of the voting interests.

13. **Adjustments Upon Changes in Capitalization, Merger or Certain Other Transactions.**

(a) **Changes in Capitalization.** Subject to any action required under Applicable Laws by the stockholders of the Company, the number of Shares of Common Stock covered by each outstanding Option, and the number of Shares of Common Stock that have been authorized for issuance under the Plan but as to which no Options have yet been granted or that have been returned to the Plan upon cancellation or expiration of an Option, as well as the price per Share of Common Stock covered by each such outstanding Option, shall be proportionately adjusted for any increase or decrease in the number of issued Shares of Common Stock resulting from a stock split, reverse stock split, stock dividend, combination, recapitalization or reclassification of the Common Stock, or any other increase or decrease in the number of issued Shares of Common Stock effected without receipt of consideration by the Company; provided, however, that conversion of any convertible securities of the Company shall not be deemed to have been "effected without receipt of consideration." Such adjustment shall be made by the Administrator, whose determination in that respect shall be final, binding and conclusive. Except as expressly provided herein, no issuance by the Company of shares of stock of any class, or securities convertible into shares of stock of any class, shall affect, and no adjustment by reason thereof shall be made with respect to, the number or price of Shares of Common Stock subject to an Option. In the event of an "Initial Public Offering" (as such term is defined in the Company's Amended and Restated Certificate of Incorporation as may be amended from time to time) or any other conversion of all of the outstanding shares of the Company's Non-Voting Common Stock, all outstanding Options shall automatically convert into options to purchase an equivalent number of shares of the Company's Voting Common Stock without any further action on the part of the Company or any Optionee.

(b) **Dissolution or Liquidation.** In the event of the dissolution or liquidation of the Company, each Option will terminate immediately prior to the consummation of such action, unless otherwise determined by the Administrator.

(c) **Corporate Transaction.** In the event of a Corporate Transaction (including without limitation a Change of Control), each outstanding Option shall be assumed or an equivalent option or right shall be substituted by such successor corporation or a parent or subsidiary of such successor corporation (the "Successor Corporation"), unless the Successor Corporation does not agree to assume the award or to substitute an equivalent option or right, in which case such Option shall terminate upon the consummation of the transaction.

For purposes of this Section 13(c), an Option shall be considered assumed, without limitation, if, at the time of issuance of the stock or other consideration upon a Corporate Transaction or a Change of Control, as the case may be, each holder of an Option would be entitled to receive upon exercise of the award the same number and kind of shares of stock or the same amount of property, cash or securities as such holder would have been entitled to receive upon the occurrence of the transaction if the holder had been, immediately prior to such transaction, the holder of the number of Shares of Common Stock covered by the award at such time (after giving effect to any adjustments in the number of Shares covered by the Option as provided for in this Section 13); provided that if such consideration received in the transaction is not solely common stock of the Successor Corporation, the Administrator may, with the consent of the Successor Corporation, provide for the consideration to be received upon exercise of the award to be solely common stock of the Successor Corporation equal to the Fair Market Value of the per Share consideration received by holders of Common Stock in the transaction.

(d) **Certain Distributions.** In the event of any distribution to the Company's stockholders of securities of any other entity or other assets (other than dividends payable in cash or stock of the Company) without receipt of consideration by the Company, the Administrator may, in its discretion, appropriately adjust the price per Share of Common Stock covered by each outstanding Option to reflect the effect of such distribution.

14. **Time of Granting Options.** The date of grant of an Option shall, for all purposes, be the date on which the Administrator makes the determination granting such Option, or such other date as is determined by the Administrator, provided that in the case of any Incentive Stock Option, the grant date shall be the later of the date on which the Administrator makes the determination granting such Incentive Stock Option or the date of commencement of the Optionee's employment relationship with the Company or any Subsidiary, Parent or Affiliate thereof. Notice of the determination shall be given to each Employee or Consultant to whom an Option is so granted within a reasonable time after the date of such grant.

15. **Amendment and Termination of the Plan.**

(a) **Authority to Amend or Terminate.** The Board may at any time amend, alter, suspend or discontinue the Plan, but no amendment, alteration, suspension or discontinuation (other than an adjustment pursuant to Section 13 above) shall be made that would materially and adversely affect the rights of any Optionee under any outstanding grant, without his or her consent. In addition, to the extent necessary and desirable to comply with the

Applicable Laws, the Company shall obtain stockholder approval of any Plan amendment in such a manner and to such a degree as required.

(b) **Effect of Amendment or Termination.** Except as to amendments which the Administrator has the authority under the Plan to make unilaterally, no amendment or termination of the Plan shall materially and adversely affect Options already granted, unless mutually agreed otherwise between the Optionee and the Administrator, which agreement must be in writing and signed by the Optionee and the Company.

16. **Conditions Upon Issuance of Shares.** Notwithstanding any other provision of the Plan or any agreement entered into by the Company pursuant to the Plan, the Company shall not be obligated, and shall have no liability for failure, to issue or deliver any Shares under the Plan unless such issuance or delivery would comply with the Applicable Laws, with such compliance determined by the Company in consultation with its legal counsel. As a condition to the exercise of an Option, the Company may require the person exercising the award to represent and warrant at the time of any such exercise that the Shares are being purchased only for investment and without any present intention to sell or distribute such Shares if, in the opinion of counsel for the Company, such a representation is required by law. Shares issued upon exercise of Options granted prior to the date on which the Common Stock becomes a Listed Security shall be subject to a right of first refusal in favor of the Company pursuant to which the Participant will be required to offer Shares to the Company before selling or transferring them to any third party on such terms and subject to such conditions as is reflected in the applicable Option Agreement.

17. **Reservation of Shares.** The Company, during the term of this Plan, will at all times reserve and keep available such number of Shares as shall be sufficient to satisfy the requirements of the Plan.

18. **Agreements.** Options shall be evidenced by Option Agreements in such form(s) as the Administrator shall from time to time approve.

19. **Stockholder Approval.** If required by the Applicable Laws, continuance of the Plan shall be subject to approval by the stockholders of the Company within twelve (12) months before or after the date the Plan is adopted. Such stockholder approval shall be obtained in the manner and to the degree required under the Applicable Laws.

20. **Information and Documents to Optionees.** Prior to the date, if any, upon which the Common Stock becomes a Listed Security and if required by the Applicable Laws, the Company shall provide financial statements at least annually to each Optionee and to each individual who acquired Shares pursuant to the Plan, during the period such Optionee has one or more Options outstanding, and in the case of an individual who acquired Shares pursuant to the Plan, during the period such individual owns such Shares. The Company shall not be required to provide such information if the issuance of Options under the Plan is limited to key employees whose duties in connection with the Company or any Subsidiary, Parent or Affiliate thereof assure their access to equivalent information.

21. **Awards Granted to California Residents.** Options granted under the Plan on any date on which the Common Stock is not a Listed Security to persons resident in California shall be subject to the provisions set forth in Attachment A hereto. To the extent the provisions of the Plan conflict with the provisions set forth on Attachment A. the provisions on Attachment A shall govern the terms of such Options.

Attachment A
Provisions Applicable to Option Recipients

Resident in California

The following additional terms shall apply if required by the Applicable Laws to (i) Options granted on any date the Common Stock is not a Listed Security, and (ii) any Shares issued upon exercise of such Options on any date the Common Stock is not a Listed Security, which are granted or issued to persons resident in California as of the date of grant or issuance, as applicable (each such person, a "California recipient"):

1. In the case of an Option, whether an Incentive Stock Option or a Nonqualified Stock Option, that is granted to a California Recipient who, at the time of the grant of such Option, owns stock representing more than 10% of the total combined voting power of all classes of stock of the Company or any Parent or Subsidiary, the per Share exercise price shall be no less than 110% of the Fair Market Value on the grant date.

2. In the case of a Nonqualified Stock Option that is granted to any other California Recipient, the per Share exercise price shall be no less than 85% of the Fair Market Value per Share on the grant date.

3. With respect to an Option issued to any California Recipient who is not an Officer, Director or Consultant, such Option shall become exercisable, or any repurchase option in favor of the Company shall lapse, at the rate of at least 20% per year over five years from the grant date.

4. The following rules shall apply to an Option issued to any California Recipient or to stock issued to a California Recipient upon exercise of an Option, in the event of termination of the California Recipient's employment or services with the Company or any Subsidiary, Parent or Affiliate thereof:

(a) If such termination was for reasons other than death or disability, the California Recipient shall have at least 30 days after the date of such termination (but in no event later than the expiration of the term of such Option established by the Plan Administrator as of the grant date) to exercise such Option.

(b) If such termination was on account of the death or disability of the California Recipient, the holder of the Option may, but only within six months from the date of such termination (but in no event later than the expiration date of the term of such Option established by the Plan Administrator as of the grant date), exercise the Option to the extent the California Recipient was otherwise entitled to exercise it at the date of such termination. To the extent that the California Recipient was not entitled to exercise the Option at the date of termination, or if the holder does not exercise such Option to the extent so entitled within six months from the date of termination, the Option shall terminate and the Common Stock underlying the unexercised portion of the Option shall revert to the Plan.

(c) Section 10(b)(iv) of the Plan shall apply with equal effect to vested Shares acquired upon exercise of an Option granted prior to the date, if any, upon which the Common Stock becomes a Listed Security to a person other than an Officer, Director or Consultant, in that

the Company shall have the right to repurchase such Shares from the Participant upon the following terms: (A) the repurchase is made within 90 days of termination of the Participant's Continuous Service for Cause at the Fair Market Value of the Shares as of the date of termination, (B) consideration for the repurchase consists of cash or cancellation of purchase money indebtedness, and (C) the repurchase right terminates upon the effective date of the Company's initial public offering of its Common Stock. With respect to vested Shares issued upon exercise of an Option granted to any Officer, Director or Consultant, the Company's right to repurchase such Shares upon termination of the Participant's Continuous Service for Cause shall be made at the Participant's original cost for the Shares and shall be effected pursuant to such terms and conditions, and at such time, as the Administrator shall determine. Nothing in this Section 10(b)(iv) shall in any way limit the Company's right to purchase unvested Shares issued upon exercise of an Option as set forth in the applicable Option Agreement.

5. The Company shall provide financial statements at least annually to each California Recipient during the period such person has one or more Options outstanding, and in the case of an individual who acquired Shares pursuant to the Plan, during the period such individual owns such Shares. The Company shall not be required to provide such information if the issuance of awards under the Plan is limited to key employees whose duties in connection with the Company or any Subsidiary, Parent or Affiliate thereof assure their access to equivalent information.

6. Unless defined below or otherwise in this Attachment, Capitalized terms shall have the meanings set forth in the Plan. For purposes of this Attachment, "Officer" means a person who is an officer of the Company within the meaning of Section 16(a) of the Exchange Act and the rules and regulations promulgated thereunder.

Amendment No. 1 to the Ikaria Holdings, Inc.
2007 Stock Option Plan

WHEREAS, Ikaria Holdings, Inc. a Delaware corporation (the "Company"), has adopted the Ikaria Holdings, Inc. 2007 Stock Option Plan (the "Plan") for the purpose of granting options to purchase shares of common stock, par value $0.01 per share, of the Company (the "Common Stock") to the employees and consultants of the Company or its subsidiaries or affiliates;

WHEREAS, Section 15(a) of the Plan provides that the Board of Directors of the Company (the "Board") may amend the Plan, subject, in the case of an amendment that materially and adversely affects the rights of an optionee under any option granted prior to such amendment, to such optionee's consent;

WHEREAS, by resolution dated April 18, 2007, the Compensation Committee of the Board (the "Compensation Committee"), pursuant to authority granted to it by the Board, amended Section 2(e) of the Plan to provide that an option agreement may include a definition of "Cause" that differs from the definition included in the Plan; and

WHEREAS, by resolution dated April 18, 2007, the Compensation Committee, pursuant to authority granted to it by the Board, amended Section 9(b) of the Plan to provide that the consideration to be paid for the shares of Common Stock to be issued upon exercise of an option may be paid with the shares underlying such option.

NOW THEREFORE, the Plan is amended as follows:

1. Section 2(e) of the Plan is amended to insert the following sentence after the last sentence thereof:

"Notwithstanding the foregoing, an Option Agreement may include a definition of Cause that is different from the foregoing definition."

2. Section 9(b) of the Plan is amended and restated in its entirety to read as follows:

"Permissible Consideration. The consideration to be paid for the Shares to be issued upon exercise of an Option, including the method of payment, shall be determined by the Administrator (and, in the case of an Incentive Stock Option, shall be determined at the time of grant) and may consist entirely of (1) cash; (2) check; (3) subject to any requirements of the Applicable Laws (including without limitation Section 153 of the Delaware General Corporation Law), delivery of Optionee's promissory note having such recourse, interest, security and redemption provisions as the Administrator determines to be appropriate after taking into account the potential accounting consequences of permitting an Optionee to deliver a promissory note; (4) cancellation of indebtedness; (5) other Shares that have a Fair Market Value on the date of surrender equal to the aggregate exercise price of the Shares as to which the Option is exercised, provided that in the case of Shares acquired, directly or indirectly, from the Company, such Shares must have been owned by the Optionee for more than six months on the date of surrender (or such shorter or longer period of time as may be deemed necessary in the Administrator's sole discretion to avoid risk of the incurrence of adverse accounting

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charges by the Company); (6) Shares of Optioned Stock that have a Fair Market Value on the date of surrender equal to the aggregate exercise price of the Shares as to which the applicable Option is exercised; provided, however, that clauses (5) and (6) shall apply only to Options granted after March 28, 2007, if permitted by the Administrator at the time of exercise in its sole discretion; and provided, further, however, that clause (6) shall apply only to Nonstatutory Stock Options; (7) if, as of the date of exercise of an Option the Company then is permitting employees to engage in a "same-day sale" cashless brokered exercise program involving one or more brokers, through such a program that complies with the Applicable Laws (including without limitation the requirements of Regulation T and other applicable regulations promulgated by the Federal Reserve Board) and that ensures prompt delivery to the Company of the amount required to pay the exercise price and any applicable withholding taxes; or (8) any combination of the foregoing methods of payment. In making its determination as to the type of consideration to accept, the Administrator shall consider if acceptance of such consideration may be reasonably expected to benefit the Company and the Administrator may, in its sole discretion, refuse to accept a particular form of consideration at the time of any Option exercise.

IN WITNESS WHEREOF, this Amendment has been executed by the undersigned, thereunto duly authorized, effective as of May 14, 2007.

IKARIA HOLDINGS, INC.

/s/ Elizabeth A. Larkin
Name: Elizabeth A. Larkin
Title: Chief Financial Officer

Amendment No. 2 to the Ikaria Holdings, Inc.
2007 Stock Option Plan

WHEREAS, Ikaria Holdings, Inc. a Delaware corporation (the "Company"), has adopted the Ikaria Holdings, Inc. 2007 Stock Option Plan (the "Plan") for the purpose of granting options to purchase shares of common stock, par value $0.01 per share, of the Company (the "Common Stock") to the employees and consultants of the Company or its subsidiaries or affiliates;

WHEREAS, Section 15(a) of the Plan provides that the Board of Directors of the Company (the "Board") may amend the Plan, subject, in the case of an amendment that materially and adversely affects the rights of an optionee under any option granted prior to such amendment, to such optionee's consent; and

WHEREAS, by resolution dated January 21, 2008, the Board amended Section 9(b) of the Plan to clarify the authority of the Administrator (as such term is defined in the Plan) to determine of the type of consideration to be paid for the shares of Common Stock to be issued upon exercise of an option.

NOW THEREFORE, Section 9(b) of the Plan is amended and restated in its entirety to read as follows:

"Permissible Consideration. The consideration to be paid for the Shares to be issued upon exercise of an Option, including the method of payment, shall be determined by the Administrator in its sole discretion at the time of grant or, with respect to a Nonstatutory Stock Option, at that time or at any time thereafter and may consist entirely of (1) cash; (2) check; (3) subject to any requirements of the Applicable Laws (including without limitation Section 153 of the Delaware General Corporation Law), delivery of Optionee's promissory note having such recourse, interest, security and redemption provisions as the Administrator determines to be appropriate after taking into account the potential accounting consequences of permitting an Optionee to deliver a promissory note; (4) cancellation of indebtedness; (5) other Shares that have a Fair Market Value on the date of surrender equal to the aggregate exercise price of the Shares as to which the Option is exercised, provided that in the case of Shares acquired, directly or indirectly, from the Company, such Shares must have been owned by the Optionee for more than six months on the date of surrender (or such shorter or longer period of time as may be deemed necessary in the Administrator's sole discretion to avoid risk of the incurrence of adverse accounting charges by the Company); (6) Shares of Optioned Stock that have a Fair Market Value on the date of surrender equal to the aggregate exercise price of the Shares as to which the applicable Option is exercised; provided, however, that clauses (5) and (6) shall apply only to Options granted after March 28, 2007; and provided, further, however, that clause (6) shall apply only to Nonstatutory Stock Options; (7) if, as of the date of exercise of an Option the Company then is permitting employees to engage in a "same-day sale" cashless brokered exercise program involving one or more brokers, through such a program that complies with the Applicable Laws (including without limitation the requirements of Regulation T and other with applicable regulations promulgated by the Federal Reserve Board) and that ensures prompt delivery to the

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Company of the amount required to pay the exercise price and any applicable withholding taxes; or (8) any combination of the foregoing methods of payment. In making its determination as to the type of consideration to accept, the Administrator shall consider if acceptance of such consideration may be reasonably expected to benefit the Company and the Administrator may, in its sole discretion, refuse to accept a particular form of consideration at the time of any Option exercise."

IN WITNESS WHEREOF, this Amendment has been executed by the undersigned, thereunto duly authorized, effective as of January 21, 2008.

IKARIA HOLDINGS, INC.

/s/ Elizabeth A. Larkin
Name: Elizabeth A. Larkin
Title: Chief Financial Officer

Amendment No. 3 to the Ikaria Holdings, Inc.
2007 Stock Option Plan

WHEREAS, Ikaria Holdings, Inc. a Delaware corporation (the "Company"), has adopted the Ikaria Holdings, Inc. 2007 Stock Option Plan (the "Plan") for the purpose of granting options to purchase shares of common stock, par value $0.01 per share, of the Company (the "Common Stock") to the employees and consultants of the Company or its subsidiaries or affiliates;

WHEREAS, Section 15(a) of the Plan provides that the Board of Directors of the Company (the "Board") may amend the Plan, subject, in the case of an amendment that materially and adversely affects the rights of an optionee under any option granted prior to such amendment, to such optionee's consent; and

WHEREAS, by resolution dated May 4, 2010, the Board amended Section 9(b) of the Plan to clarify the authority of the Administrator (as such term is defined in the Plan) to determine of the type of consideration to be paid for the shares of Common Stock to be issued upon exercise of an option.

NOW THEREFORE, Section 9(b) of the Plan is amended and restated in its entirety to read as follows:

"**Permissible Consideration.** The consideration to be paid for the Shares to be issued upon exercise of an Option, including the method of payment, may consist entirely of (1) cash; (2) check; (3) subject to any requirements of the Applicable Laws (including without limitation Section 153 of the Delaware General Corporation Law), delivery of Optionee's promissory note having such recourse, interest, security and redemption provisions as the Administrator determines to be appropriate after taking into account the potential accounting consequences of permitting an Optionee to deliver a promissory note; (4) cancellation of indebtedness; (5) other Shares that have a Fair Market Value on the date of surrender equal to the aggregate exercise price of the Shares as to which the Option is exercised; (6) Shares of Optioned Stock that have a Fair Market Value on the date of surrender equal to the aggregate exercise price of the Shares as to which the applicable Option is exercised; provided, however, that clauses (5) and (6) shall apply only to Options granted after March 28, 2007; and provided, further, however, that clause (6) shall apply only to Nonstatutory Stock Options; (7) through a "same-day sale" cashless brokered exercise program involving one or more brokers that complies with the Applicable Laws (including without limitation the requirements of Regulation T and other applicable regulations promulgated by the Federal Reserve Board) and that ensures prompt delivery to the Company of the amount required to pay the exercise price and any applicable withholding taxes; or (8) any combination of the foregoing methods of payment. Notwithstanding the foregoing, (i) clauses (3) through (6) shall only apply prior to an Initial Public Offering (as such term is defined in the Company's Amended and Restated Certificate of Incorporation as may be amended from time to time) as determined by the Administrator at the time of grant or with respect to a

NonStatutory Stock Option, at any time thereafter, (ii) clause (7) shall only apply while the Company's Shares are publicly traded and (iii) the availability of any method of payment shall be subject to the terms and conditions of the Company's credit facilities as in effect from time to time."

IN WITNESS WHEREOF, this Amendment has been executed by the undersigned, thereunto duly authorized, effective as of May 4, 2010.

IKARIA HOLDINGS, INC.

/s/ Matthew M. Bennett
Name: Matthew M. Bennett
Title: Senior Vice President and Secretary

Amendment No. 4 to the Ikaria Holdings, Inc.
2007 Stock Option Plan

WHEREAS, Ikaria, Inc. (f/k/a Ikaria Holdings, Inc.), a Delaware corporation (the "Company"), has adopted the Ikaria Holdings, Inc. 2007 Stock Option Plan (the "Plan") for the purpose of granting options to purchase shares of common stock, par value $0.01 per share, of the Company (the "Common Stock") to the employees and consultants of the Company or its subsidiaries or affiliates;

WHEREAS, Section 15(a) of the Plan provides that the Board of Directors of the Company (the "Board") may amend the Plan, subject, in the case of an amendment that materially and adversely affects the rights of an optionee under any option granted prior to such amendment, to such optionee's consent; and

WHEREAS, the Company desires to amend Section 13(d) of the Plan to allow the plan administrator to make appropriate adjustments to the exercise price of certain outstanding options to reflect the effect of the extraordinary dividend declared on July 3, 2013.

NOW THEREFORE, Section 13(d) of the Plan is amended and restated in its entirety to read as follows:

Certain Distributions. In the event of any distribution to the Company's stockholders of securities of any other entity or other assets (other than dividends payable in cash or stock of the Company) without receipt of consideration by the Company, the Administrator may, in its discretion, appropriately adjust the price per Share of Common Stock covered by each outstanding Option to reflect the effect of such distribution. In connection with the extraordinary dividend declared by the Board on July 3, 2013, the Administrator may, in its discretion, appropriately adjust the price per Share of Common Stock to reflect the effect of such extraordinary dividend. The Administrator may make such adjustment(s) to any or all outstanding Options, and make different adjustments to different Options. In determining what adjustments, if any, to make, the Administrator may take into account any other actions that the Company may take with respect outstanding Options, including, but not limited to, any payments made or to be made to the holders of Options under any plan or arrangement of the Company.

[Signature Page Follows]

IN WITNESS WHEREOF, this Amendment has been executed by the undersigned, thereunto duly authorized, effective as of the date first set forth above.

IKARIA, INC.

/s/ James Briggs
Name: James Briggs
Title: Senior Vice President

Exhibit 10.2

ASSUMED AMENDED AND RESTATED
IKARIA HOLDINGS, INC.
2010 LONG TERM INCENTIVE PLAN

1. **Purpose.**

This Plan provides select employees, officers, consultants and directors of the Company and its Subsidiaries with an opportunity to receive long term incentive awards as set forth by the terms and conditions of this Plan. These awards have been designed to motivate recipients to achieve short term and longer term results for the Company, with an expectation that their expertise will contribute to the attainment of key business goals that will further benefit shareholders. Awards under this Plan include Incentive Stock Options, Nonqualified Stock Options, Stock Appreciation Rights, Restricted Stock, Restricted Stock Units, Dividend Equivalent Rights, Performance Units, Performance Share Units, Performance-Based Restricted Stock, Share Awards and Cash Incentive Awards (as each term is herein defined).

2. **Definitions.**

For purposes of the Plan, the following terms shall have the following meanings:

"Affiliate" means, with respect to any Person, any other Person which, directly or indirectly, controls, is controlled by or is under common control with, such Person.

"Agreement" means the written agreement between the Company and an Optionee or Grantee evidencing the grant of an Option or Award and setting forth the terms and conditions thereof.

"Award" means a Stock-Based Award or a Cash Incentive Award.

"Board" means the Board of Directors of the Company.

"Cash Incentive Award" means the right granted to an Eligible Individual to receive a payment of cash pursuant to Section 10.3.

"Change in Capitalization" means any increase or reduction in the number of Shares, or any change (including, but not limited to, in the case of a spin-off, extraordinary dividend or other extraordinary distribution in respect of Shares, a change in value) in the Shares or exchange of Shares for a different number or kind of shares or other securities of the Company or another corporation, by reason of a reclassification, recapitalization, merger, consolidation, reorganization, spin-off, split-up, issuance of warrants or rights or debentures, stock dividend, stock split or reverse stock split, cash dividend, property dividend, combination or exchange of shares, repurchase of shares, change in corporate structure or other, similar transaction or event.

"Code" means the Internal Revenue Code of 1986, as amended.

"Committee" means the Compensation Committee of the Board, or a subcommittee thereof, which shall administer the Plan and perform the functions set forth herein;

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provided, that, if there is no Compensation Committee of the Board, or if the Board determines that the Compensation Committee shall not be the Committee, then the Committee shall be the Board or such Directors as are appointed to the Committee by the Board.

"Company" means Ikaria Holdings, Inc., a Delaware corporation, and shall include any successor thereto by merger, consolidation, acquisition of substantially all the assets thereof, or otherwise.

"Director" means a director of the Company.

"Dividend Equivalent Right" means a right to receive all or some portion of the cash dividends that are or would be payable with respect to Shares.

"Division" means any of the operating units or divisions of the Company or any Subsidiary designated as a Division by the Committee.

"Eligible Individual" means any of the following individuals who is designated by the Committee as eligible to receive Options or Awards subject to the conditions set forth herein: (i) any director, officer or employee of the Company or a Subsidiary, (ii) any individual to whom the Company or a Subsidiary has extended a formal, written offer of employment or (iii) any consultant or advisor of the Company or a Subsidiary.

"Exchange Act" means the Securities Exchange Act of 1934, as amended.

"Fair Market Value" on any date means (i) the closing price at the end of normal market hours of the Shares on such date as quoted on the National Association of Securities Dealers Automated Quotation System (the "Nasdaq"), (ii) if the Shares are not quoted on the Nasdaq but are listed for trading on the New York Stock Exchange (the "NYSE") or other national securities exchange, the closing price at the close of the primary trading session of the Shares on such date on the NYSE (or, if the Shares are not listed for trading on the NYSE, on such other exchange) or (iii) if the Shares are not listed on the Nasdaq, the NYSE or other national exchange, or if there is no such closing price for such date on the Nasdaq, the NYSE or other national exchange, the fair market value of the Shares as determined in good faith by the Committee (and, if applicable, in accordance with Sections 409A and 422 of the Code).

"Grantee" means a person to whom an Award has been granted under the Plan.

"Incentive Stock Option" means an Option satisfying the requirements of Section 422 of the Code and designated by the Committee as an Incentive Stock Option.

"Initial Public Offering" shall have the meaning ascribed to such term in the Company's Amended and Restated Certificate of Incorporation as may be amended from time to time. An Initial Public Offering shall be deemed to have been consummated if and when the registration statement in connection with such public offering shall have been declared effective and the offering of securities thereunder shall have closed in accordance with the terms of the related underwriting agreement.

"Listed Security" means any security of the Company that is listed or approved for listing on a national securities exchange.

"Non-Employee Director" means a director of the Company who is a "non-employee director" within the meaning of Rule 16b-3 promulgated under the Exchange Act.

"Non-Voting Common Stock" means the non-voting common stock of the Company, par value $0.01 per share.

"Nonqualified Stock Option" means an Option which is not an Incentive Stock Option.

"Option" means a Nonqualified Stock Option or an Incentive Stock Option.

"Optionee" means a person to whom an Option has been granted under the Plan.

"Outside Director" means a director of the Company who is an "outside director" within the meaning of Section 162(m) of the Code and the regulations promulgated thereunder.

"Parent" means any corporation which is a parent corporation (within the meaning of Section 424(e) of the Code) with respect to the Company.

"Performance Awards" means Performance Units, Performance Share Units, Performance-Based Restricted Stock, Cash Incentive Awards or any or all of them.

"Performance-Based Compensation" means any Option or Award that is intended to constitute "performance based compensation" within the meaning of Section 162(m)(4)(C) of the Code and the regulations promulgated thereunder.

"Performance-Based Restricted Stock" means Shares of Restricted Stock issued or transferred to an Eligible Individual under Section 10.2.

"Performance Cycle" means a time period of not less than one year as specified by the Committee at the time Performance Awards or Cash Incentive Awards are granted during which the performance of the Company, a Subsidiary or a Division will be measured.

"Performance Objectives" has the meaning set forth in Section 10.4.

"Performance Share Units" means Performance Share Units granted to an Eligible Individual under Section 10.1.

"Performance Units" means Performance Units granted to an Eligible Individual under Section 10.1.

"Person" means an individual, a corporation, a partnership, a limited liability company, an association, a trust or any other entity or organization, including a government or political subdivision or an agency or instrumentality thereof.

"Plan" means the Amended and Restated Ikaria Holdings, Inc. 2010 Long Term Incentive Plan, as amended and restated from time to time.

"Recaptured Shares" has the meaning set forth in Section 4.2(b).

"Restricted Stock" means Shares issued or transferred to an Eligible Individual pursuant to Section 9.1.

"Restricted Stock Units" means rights granted to an Eligible Individual under Section 9.2 representing a number of hypothetical Shares.

"Securities Act" means the Securities Act of 1933, as amended.

"Share Award" means an Award of Shares granted pursuant to Section 11.

"Shareholder's Agreement" means the Shareholder's Agreement governing the rights, duties and obligations of certain present or former employees, officers, consultants and directors of the Company with respect to Shares issued pursuant to Options or Awards granted or sold to such persons, in such form as is in use by the Company from time to time.

"Shares" means (i) the Non-Voting Common Stock, prior to the consummation of an Initial Public Offering, and (ii) the Voting Common Stock, from and after the consummation of an Initial Public Offering.

"Stock Appreciation Right" means a right to receive all or some portion of the increase in the value of the Shares as provided in Section 7 hereof.

"Stock-Based Award" means a grant of Restricted Stock, Restricted Stock Units, a Stock Appreciation Right, a Performance Award, a Dividend Equivalent Right, a Share Award or any or all of them.

"Subsidiary" means (i) except as provided in subsection (ii) below, any corporation which is a subsidiary corporation within the meaning of Section 424(f) of the Code with respect to the Company, and (ii) in relation to the eligibility to receive Options or Awards other than Incentive Stock Options and

continued employment or service for purposes of Options and Awards (unless the Committee determines otherwise), any entity, whether or not incorporated, in which the Company directly or indirectly owns 50% or more of the outstanding equity or other ownership interests.

"Taxable Event" has the meaning set forth in Section 20.2.

"Ten-Percent Stockholder" means an Eligible Individual who, at the time an Incentive Stock Option is to be granted to him or her, owns (within the meaning of Section 422(b)(6) of the Code) stock possessing more than 10% of the total combined voting power of all classes of stock of the Company, or of a Parent or a Subsidiary.

"Transition Period" means the period beginning with the consummation of an Initial Public Offering and ending as of the earlier of (i) the date of the first annual meeting of

shareholders of the Company at which directors are to be elected that occurs after the close of the third calendar year following the calendar year in which the Initial Public Offering occurs and (ii) the expiration of the "reliance period" under Treasury Regulation Section 1.162-27(f)(2).

"Voting Common Stock" means the voting common stock of the Company, par value $0.01 per share.

"Withholding Taxes" has the meaning set forth in Section 20.2.

3. **Administration.**

3.1. The Plan shall be administered by the Committee, which shall hold meetings at such times as may be necessary for the proper administration of the Plan. Any decision or determination reduced to writing and signed by a majority of all of the members of the Committee shall be as fully effective as if made by a majority vote at a meeting duly called and held. The Committee shall consist of at least two Directors and may consist of the entire Board; provided, however, that from and after the consummation of an Initial Public Offering (a) if the Committee consists of less than the entire Board, then, with respect to any Option or Award granted to an Eligible Individual who is subject to Section 16 of the Exchange Act, the Committee shall consist of at least two Directors, each of whom shall be a Non-Employee Director, and (b) following the Transition Period, to the extent necessary for any Option or Award intended to qualify as Performance-Based Compensation to so qualify, the Committee shall consist of at least two Directors, each of whom shall be an Outside Director. For purposes of the preceding sentence, if one or more members of the Committee is not a Non-Employee Director and/or an Outside Director but recuses himself or herself or abstains from voting with respect to a particular action taken by the Committee, then the Committee, with respect to that action, shall be deemed to consist only of the members of the Committee who have not recused themselves or abstained from voting. A quorum shall consist of not fewer than two members of the Committee and a majority of a quorum may authorize any action. The Board may, in its sole discretion, permit the Chief Executive Officer of the Company to exercise the authority granted to the Committee pursuant to Sections 3.4(a) and (b) with respect to other Eligible Individuals (other than Eligible Individuals subject to Section 16 of the Exchange Act or receiving compensation subject to Section 162(m) of the Code), subject to such limitations as imposed by the Board in its discretion and subject to compliance with Section 157(c) of the Delaware General Corporation Law, as amended from time to time.

3.2. Board Reservation and Delegation. Except to the extent necessary for any Award or Option intended to qualify as Performance-Based Compensation to so qualify, the Board may, in its discretion, reserve to itself or exercise any or all of the authority and responsibility of the Committee hereunder and may also delegate to another committee of the Board any or all of the authority and responsibility of the Committee with respect to Awards or Options to Eligible Individuals who are not subject to Section 16(b) of the Exchange Act at the time any such delegated authority or responsibility is exercised. Such other committee may consist of one or more Directors who may, but need not be, officers or employees of the Company or any of its Subsidiaries. To the extent the Board has reserved to itself, or exercised, the authority and responsibility of the Committee, or delegated the authority and responsibility of

the Committee to such other committee, all references to the Committee in the Plan shall be to the Board or to such other committee.

3.3. No member of the Committee shall be liable for any action, failure to act, determination or interpretation made in good faith with respect to this Plan or any transaction hereunder. The Company hereby agrees to indemnify each member of the Committee for all costs and expenses and, to the extent permitted by applicable law, any liability incurred in connection with defending against, responding to, negotiating for the settlement of or otherwise dealing with any claim, cause of action or dispute of any kind arising in connection with any actions in administering this Plan or in authorizing or denying authorization to any transaction hereunder.

3.4. Subject to the express terms and conditions set forth herein, the Committee shall have the power from time to time to:

(a) determine those Eligible Individuals to whom Options shall be granted under the Plan and the number of such Options to be granted and to prescribe the terms and conditions (which need not be identical) of each such Option, including the exercise price per Share, the vesting schedule and the duration of each Option, and make any amendment or modification to any Option Agreement consistent with the terms of the Plan;

(b) select those Eligible Individuals to whom Stock-Based Awards and/or Cash Incentive Awards shall be granted under the Plan and to determine the number of Shares in respect of which each Stock-Based Award is granted, and the terms and conditions (which need not be identical) of each such Award, and make any amendment or modification to any Agreement consistent with the terms of the Plan;

(c) to construe and interpret the Plan and the Options and Awards granted hereunder and to establish, amend and revoke rules and regulations for the administration of the Plan, including, but not limited to, correcting any defect or supplying any omission, or reconciling any inconsistency in the Plan or in any Agreement, in the manner and to the extent it shall deem necessary or advisable, including so that the Plan and the operation of the Plan complies with Rule 16b-3 under the Exchange Act, Sections 162(m) and 409A of the Code (to the extent applicable) and other applicable law, and otherwise to make the Plan fully effective;

(d) to make such adjustments to the Options and Awards as are required by Section 13;

(e) to determine the duration and purposes for leaves of absence which may be granted to an Optionee or Grantee on an individual basis without constituting a termination of employment or service for purposes of the Plan;

(f) to exercise its discretion with respect to the powers and rights granted to it as set forth in the Plan; and

(g) generally, to exercise such powers and to perform such acts as are deemed necessary or advisable to promote the best interests of the Company with respect to the Plan.

All decisions and determinations by the Committee in the exercise of the foregoing powers shall be final, binding and conclusive upon the Company, each Subsidiary, the Optionees and Grantees, and all other persons having or claiming any interest under the Plan or any Option or Award granted hereunder.

3.5. Notwithstanding anything herein to the contrary, the Committee may determine the terms and conditions of Options and Awards and make such adjustments to the terms thereof and to the Plan as are necessary or advisable to fulfill the purposes of the Plan taking into account matters of local law or practice, including tax and securities laws of the individual States and of jurisdictions outside the United States. Any such adjustments to the Plan shall be evidenced by one or more written supplements to the Plan.

4. **Stock Subject to the Plan; Grant Limitations.**

4.1. Number of Shares Authorized for Issuance; Limitations on Options and Awards. Subject to any adjustment as provided in the Plan, the Shares to be issued under the Plan may be, in whole or in part, authorized but unissued Shares or issued Shares which shall have been reacquired by the Company and held by it as treasury shares. The aggregate number of Shares that may be made the subject of Awards or Options granted under the Plan shall initially be 2,793,062 (calculated as set forth in Section 4.2): provided, that, prior to the termination of the Plan in accordance with Section 16.1, that number shall be increased automatically on January 1st of each year commencing on January 1, 2011, in an amount such that the aggregate number of Shares that may be made the subject of Awards or Options granted under the Plan as of such January 1 shall be equal to the sum of (i) 3% of the total number of shares of Voting Common Stock ,Non-Voting Common Stock, Series A Preferred and Series B Preferred issued and outstanding on December 31st of the immediately preceding calendar year and (ii) the number of Recaptured Shares that have not previously been made the subject of Awards or Options hereunder; provided, that the Board may in its sole discretion reduce the amount of the increase in any particular year. In no event may more than 20 million Shares be made the subject of Incentive Stock Options under the Plan (calculated as set forth in Section 4.2).

4.2. Calculating Shares Available under the Plan.

(a) Upon the granting of an Option or an Award, the number of Shares available under Section 4.1 for the granting of further Options and Awards shall be reduced as follows:

(i) In connection with the granting of an Option or an Award (other than the granting of (A) Performance Units, (B) Cash Incentive Awards, (C) Dividend Equivalent Rights or (D) other Awards payable in cash), the number of Shares shall be reduced by the number of Shares in respect of which the Option or Award is granted or denominated,

with a corresponding adjustment if an Award is ultimately settled in whole or in part in a greater or lesser number of Shares.

(ii) In connection with the granting of a Performance Unit, the number of Shares shall initially be reduced by an amount equal to the quotient of (A) the dollar amount in which the Performance Unit is denominated divided by (B) the Fair Market Value of a Share on the date the Performance Unit is granted, with a corresponding adjustment if the Performance Unit is ultimately settled in whole or in part in a greater or lesser number of Shares.

(iii) In connection with the granting of a Dividend Equivalent or a Cash Incentive Award, the number of Shares available under Section 4.1 shall not be reduced; provided, however, that if Shares are issued in settlement of such an Award, the number of Shares available for the granting of further Options and Awards under Section 4.1 shall be reduced by the number of Shares so issued.

(iv) If any Option is exercised (A) by tendering Shares, either actually or by attestation, to the Company as full or partial payment of the exercise price or (B) by the Company reducing the number of Shares to be issued upon exercise of such Option in full or partial payment of the exercise price, the maximum number of Shares available under Section 4.1 shall be increased by the number of Shares so tendered or by the amount of such reduction.

(v) If Shares subject to any Option or Award are retained by the Company in satisfaction of any Withholding Taxes payable by an Optionee or Grantee, then the maximum number of Shares available under Section 4.1 shall be increased by the number of Shares so retained.

(b) Notwithstanding Section 4.2(a), in the event that an Award is granted that, pursuant to the terms of an Agreement, cannot be settled in Shares, the aggregate number of Shares that may be the subject of Options or Awards granted under the Plan shall not be reduced to reflect such Award. Whenever any outstanding Option or Award or portion thereof expires, is cancelled or forfeited, is settled in cash or is otherwise terminated for any reason without having been exercised or payment having been made in respect of the entire Option or Award, the Shares allocable to the expired, cancelled, forfeited, settled or otherwise terminated portion of the Option or Award ("Recaptured Shares") may again be the subject of Options or Awards granted hereunder. In addition, upon settlement of a Stock Appreciation Right in Shares, the excess of the number of Shares subject to the Stock Appreciation Right over the number of Shares issued in settlement of the Stock Appreciation Right may again be the subject of Options or Awards granted hereunder.

5. **Option Grants for Eligible Individuals.**

5.1. Authority of Committee. Subject to the provisions of the Plan, the Committee shall have full and final authority to select those Eligible Individuals who will receive Options, and the terms and conditions of the grant to such Eligible Individuals shall be set forth in an Agreement. Incentive Stock Options may be granted only to Eligible Individuals who are employees of the Company or a Subsidiary.

5.2. Exercise Price. The purchase price per Share under each Option shall be determined by the Committee and set forth in the Agreement but shall not be less than the Fair Market Value of a Share on the date of grant (110% of Fair Market Value in the case of an Incentive Stock Option granted to a Ten-Percent Stockholder).

5.3. Maximum Duration. Options shall not be exercisable after the expiration of ten years from the date of grant (five years in the case of an Incentive Stock Option granted to a Ten-Percent Stockholder); provided, however, that an Option (other than an Incentive Stock Option) may, upon the death of the Participant prior to the expiration of the Option, be exercised for such period following the date of the Participant's death as the Committee determines which may extend for up to one (1) year beyond ten (10) years from the date the Option is granted. The term of an Option shall be set forth in the Agreement evidencing such Option. To the extent permitted by applicable law, the Committee may, subsequent to the granting of any Option, extend the term thereof, but in no event shall the term as so extended exceed the maximum term provided for in the first sentence of this Section 5.3.

5.4. Vesting. Each Option shall become exercisable in such installments (which need not be equal) and at such times as may be designated by the Committee and set forth in the Agreement. To the extent not exercised, installments shall accumulate and be exercisable, in whole or in part, at any time after becoming exercisable, but not later than the date the Option expires. The Committee may accelerate the exercisability of any Option or portion thereof at any time.

5.5. Limitations on Incentive Stock Options. To the extent that the aggregate Fair Market Value (determined as of the date of the grant) of Shares with respect to which Incentive Stock Options granted under the Plan and "incentive stock options" (within the meaning of Section 422 of the Code) granted under all other plans of the Company or its Subsidiaries (in either case determined without regard to this Section 5.5) are exercisable by an Optionee for the first time during any calendar year exceeds $100,000, such Incentive Stock Options shall be treated as Nonqualified Stock Options. In applying the limitation in the preceding sentence in the case of multiple Option grants, Options which were intended to be Incentive Stock Options shall be treated as Nonqualified Stock Options according to the order in which they were granted such that the most recently granted Options are first treated as Nonqualified Stock Options.

5.6. Options Granted to California Residents. Options granted under the Plan on any date on which the Shares are not a Listed Security to persons resident in the State of California shall be subject to the provisions set forth in Attachment A hereto. To the extent the provisions of the Plan conflict with the provisions set forth on Attachment A, the provisions on Attachment A shall govern the terms of such Options.

6. **Terms and Conditions Applicable to All Options.**

6.1. No Sale or Transfer. Except to the extent permitted by the Committee with respect to Nonqualified Stock Options, no Option shall be transferable by the Optionee other than by will or by the laws of descent and distribution or, in the case of an Option other than an Incentive Stock Option, pursuant to a domestic relations order (within the meaning of

Rule 16a-12 promulgated under the Exchange Act), and an Option shall be exercisable during the lifetime of an Optionee only by the Optionee or his or her guardian or legal representative.

6.2. Manner of Exercise and Payment. The exercise of an Option shall be made by a written notice delivered in person or by mail to the Secretary of the Company at the Company's principal executive office, specifying the number of Shares to be exercised and, to the extent applicable, accompanied by payment therefor and otherwise in accordance with the Agreement pursuant to which the Option was granted, or in such other manner as prescribed by the Committee; provided, however, that Options may not be exercised by an Optionee for six months following a hardship distribution to the Optionee, to the extent such exercise is prohibited under Treasury Regulation Section 1.401(k)-1(d)(3)(iv)(E). The exercise price for any Shares purchased pursuant to the exercise of an Option shall be paid, in any of the following forms (or any combination thereof): (a) cash, (b) the transfer, either actually or by attestation, to the Company of Shares (if the Shares are then Listed Securities), such transfer to be upon such terms and conditions as determined by the Committee, (c) a reduction by the Company in the number of Shares (if the Shares are then Listed Securities) to be issued upon such exercise having a Fair Market Value on the date of exercise equal to the aggregate exercise price payable, (d) such other cashless exercise procedures (including through the use of a cashless exercise program with a registered broker-dealer approved by the Committee) or (e) a combination of the foregoing; provided, however, that (i) clauses (b) through (d) shall only apply prior to an Initial Public Offering (as such term is defined in the Company's Amended and Restated Certificate of Incorporation as may be amended from time to time) as determined by the Committee at the time of grant or with respect to a Nonqualified Stock Option, at any time thereafter, and (ii) the availability of any method of payment shall be subject to the terms and conditions of the Company's credit facilities as in effect from time to time. Any Shares transferred to the Company as payment of the exercise price under an Option shall be valued at their Fair Market Value on the day of exercise of such Option. If requested by the Committee, the Optionee shall deliver the Agreement evidencing the Option to the Secretary of the Company who shall endorse thereon a notation of such exercise and return such Agreement to the Optionee. No fractional Shares (or cash in lieu thereof) shall be issued upon exercise of an Option and the number of Shares that may be purchased upon exercise shall be rounded down to the nearest number of whole Shares. Notwithstanding anything in this Plan to the contrary, an Option may be exercised in accordance with the arrangements and procedures provided in this Section 6.2 only to the extent such arrangements or procedures comply with Section 13(k) of the Exchange Act and any other applicable laws, rules and regulations.

6.3. Rights of Optionees. No Optionee shall be deemed for any purpose to be the owner of any Shares subject to any Option unless and until (a) the Option shall have been exercised pursuant to the terms thereof, (b) the Company shall have issued and delivered Shares to the Optionee (such issuance and delivery to be subject to the Shareholder's Agreement, if applicable), (c) the Optionee's name shall have been entered as a stockholder of record on the books of the Company and (d) if required by the Committee, the Optionee shall have delivered a fully executed Shareholder's Agreement and stock power to the Company. Thereupon, the Optionee shall have full dividend and other ownership rights with respect to such Shares, including voting rights if and only if such Shares consist of Voting Common Stock, subject to such terms and conditions as may be set forth in the Agreement and, if applicable, the Shareholder's Agreement.

6.4. **Buyout Provisions.** The Committee may at any time offer to buy out for a payment in cash or Shares an Option previously granted under the Plan based on such terms and conditions as the Committee shall establish and communicate to the Optionee at the time that such offer is made.

7. **Stock Appreciation Rights.**

7.1. Grant of Stock Appreciation Rights. The Committee may in its discretion, either alone or in connection with the grant of an Option, grant Stock Appreciation Rights in accordance with the Plan, the terms and conditions of which shall be set forth in an Agreement. If granted in connection with an Option, a Stock Appreciation Right shall cover the same Shares covered by the Option (or such lesser number of Shares as the Committee may determine) and shall, except as provided in this Section 7, be subject to the same terms and conditions as the related Option.

7.2. Time of Grant. A Stock Appreciation Right may be granted (a) at any time if unrelated to an Option, or (b) if related to an Option, either at the time of grant or at any time thereafter during the term of the Option.

7.3. Stock Appreciation Right Granted in Connection With an Option.

(a) Exercise. A Stock Appreciation Right granted in connection with an Option shall be exercisable at such time or times and only to the extent that the related Options are exercisable and will not be transferable except to the extent the related Option is transferable. A Stock Appreciation Right granted in connection with an Incentive Stock Option shall be exercisable only if the Fair Market Value of a Share on the trading day immediately preceding the date of exercise exceeds the exercise price specified in the related Incentive Stock Option Agreement.

(b) Amount Payable. Upon the exercise of a Stock Appreciation Right related to an Option, the Grantee shall be entitled to receive an amount of cash or a number of Shares determined by multiplying (i) the excess of the Fair Market Value of a Share on the trading day immediately preceding the date of exercise of such Stock Appreciation Right over the per Share exercise price under the related Option, by (ii) the number of Shares as to which such Stock Appreciation Right is being exercised. Notwithstanding the foregoing, the Committee may limit in any manner the amount payable with respect to any Stock Appreciation Right by including such a limit in the Agreement evidencing the Stock Appreciation Right at the time it is granted.

(c) Treatment of Related Options and Stock Appreciation Rights Upon Exercise. Upon the exercise of a Stock Appreciation Right granted in connection with an Option, the Option shall be cancelled to the extent of the number of Shares as to which the Stock Appreciation Right is exercised, and upon the exercise of an Option granted in connection with a Stock Appreciation Right, the Stock Appreciation Right shall be cancelled to the extent of the number of Shares as to which the Option is exercised or surrendered.

7.4. Stock Appreciation Right Unrelated to an Option. The Committee may grant to Eligible Individuals Stock Appreciation Rights unrelated to Options. Stock

Appreciation Rights unrelated to Options shall contain such terms and conditions as to exercisability, vesting and duration as the Committee shall determine, but in no event shall they have a term of greater than ten years. Upon exercise of a Stock Appreciation Right unrelated to an Option, the Grantee shall be entitled to receive an amount of cash or a number of Shares determined by multiplying (a) the excess of the Fair Market Value of a Share on the date of exercise of such Stock Appreciation Right over the Fair Market Value of a Share on the date the Stock Appreciation Right was granted, by (b) the number of Shares as to which the Stock Appreciation Right is being exercised. Notwithstanding the foregoing, the Committee may limit in any manner the amount payable with respect to any Stock Appreciation Right by including such a limit in the Agreement evidencing the Stock Appreciation Right at the time it is granted.

7.5. No Sale or Transfer. The Grantee shall not sell, transfer, assign, exchange, pledge, encumber or otherwise dispose of a Stock Appreciation Right or any portion thereof.

7.6. Manner of Exercise. The exercise of Stock Appreciation Rights shall be made only by delivery of written notice to the Company. Such notice shall state that the Grantee is electing to exercise the Stock Appreciation Right, shall set forth the number of Shares in respect of which the Stock Appreciation Right is being exercised and shall be signed by the Grantee or, where applicable, by the Grantee's legal representative.

7.7. Form of Payment. Payment of the amount determined under Section 7.3(b) or 7.4 may be made in whole Shares in a number determined at their Fair Market Value on the trading day immediately preceding the date of exercise of the Stock Appreciation Right, or solely in cash, or in a combination of cash and Shares. Such form of payment shall be determined by the Committee and set forth in the Agreement evidencing the Stock Appreciation Right. If the Committee decides to make full payment in Shares and the amount payable results in a fractional Share, payment for the fractional Share will be made in cash.

8. **Dividend Equivalent Rights.**

Dividend Equivalent Rights may be granted to Eligible Individuals in tandem with an Option or Award or as a separate Award. The terms and conditions applicable to each Dividend Equivalent Right shall be specified in the Agreement under which the Dividend Equivalent Right is granted. Amounts payable in respect of Dividend Equivalent Rights may be payable currently or deferred until the lapsing of restrictions on such Dividend Equivalent Rights or until the vesting, exercise, payment, settlement or other lapse of restrictions on the Option or Award to which the Dividend Equivalent Rights relate. In the event that any portion of the amount payable in respect of Dividend Equivalent Rights is to be deferred, the Committee shall determine whether such amounts are to be held in cash or reinvested in Shares or deemed (notionally) to be reinvested in Shares. If amounts payable in respect of Dividend Equivalent Rights are to be held in cash, there may be credited at the end of each year (or portion thereof) interest on the amount of the account at the beginning of the year at a rate per annum as the Committee, in its discretion, may determine. Dividend Equivalent Rights may be settled in cash or Shares or a combination thereof, in a single installment or multiple installments, in each case as determined by the Committee.

9. **Restricted Stock; Restricted Stock Units.**

9.1. Restricted Stock. The Committee may grant Awards of Restricted Stock to Eligible Individuals, which shall be evidenced by an Agreement between the Company and the Grantee. Each Agreement shall contain such restrictions, terms and conditions as the Committee may, in its discretion, determine and (without limiting the generality of the foregoing) such Agreements may require that an appropriate legend be placed on Share certificates during the period of restriction. Awards of Restricted Stock shall be subject to the terms and provisions set forth below in this Section 9.

(a) Rights of Grantee. Shares of Restricted Stock granted pursuant to an Award hereunder shall be issued in the name of the Grantee as soon as reasonably practicable after the Award is granted provided that the Grantee has executed an Agreement evidencing the Award, the appropriate blank stock powers and, in the discretion of the Committee, a Shareholder's Agreement, an escrow agreement and any other documents which the Committee may require as a condition to the issuance of such Shares. If a Grantee shall fail to execute the Agreement evidencing a Restricted Stock Award and any other documents which the Committee may require, or otherwise indicate acceptance of the Restricted Stock Award in a manner prescribed by the Committee within the time period prescribed by the Committee at the time the Award is granted, the Award shall be null and void. At the discretion of the Committee, Shares issued in connection with a Restricted Stock Award shall be deposited together with the stock powers with the Company as escrow agent (or other escrow agent designated by the Committee). Except to the extent set forth in an Agreement and subject to the provisions of any applicable Shareholder's Agreement, upon delivery of the Shares to the escrow agent, the Grantee shall have all of the rights of a stockholder with respect to such Shares, including the right to receive all dividends or other distributions paid or made with respect to the Shares and the right to vote the Shares if and only if such Shares consist of Voting Common Stock.

(b) No Sale or Transfer. Until all restrictions upon the Shares of Restricted Stock awarded to a Grantee shall have lapsed in the manner set forth in Section 9(c), the Grantee shall not sell, transfer, assign, exchange, pledge, encumber or otherwise dispose of the Shares of Restricted Stock.

(c) Lapse of Restrictions. Restrictions upon Shares of Restricted Stock awarded hereunder shall lapse at such time or times and on such terms and conditions as the Committee may determine. The Agreement evidencing the Award shall set forth any such restrictions. Such restrictions may, in the discretion of the Committee, be contingent on future employment or services, the satisfaction of performance-related goals, or a combination of the foregoing.

(d) Treatment of Dividends. At the time an Award of Shares of Restricted Stock is granted, the Committee may, in its discretion, determine that the payment to the Grantee of dividends, or a specified portion thereof, declared or paid on such Shares by the Company shall be (a) deferred until the lapsing of the restrictions imposed upon such Shares and (b) held by the Company for the account of the Grantee until such time. In the event that dividends are to be deferred, the Committee shall determine whether such dividends are to be reinvested in Shares (which shall be held as additional Shares of Restricted Stock) or held in

cash. If deferred dividends are to be held in cash, there may be credited at the end of each year (or portion thereof) interest on the amount of the account at the beginning of the year at a rate per annum as the Committee, in its discretion, may determine. Payment of deferred dividends in respect of Shares of Restricted Stock (whether held in cash or as additional Shares of Restricted Stock), together with interest accrued thereon, if any, shall be made upon the lapsing of restrictions imposed on the Shares in respect of which the deferred dividends were paid, and any dividends deferred (together with any interest accrued thereon) in respect of any Shares of Restricted Stock shall be forfeited upon the forfeiture of such Shares.

(e) Delivery of Shares. Upon the lapse of the restrictions on Shares of Restricted Stock, the Company may maintain the shares in book-entry form; provided, however, that if the Grantee so requests, the Committee shall cause a stock certificate to be delivered to the Grantee with respect to such Shares, free of all restrictions hereunder; and provided, further, that, if Shares of Restricted Stock are subject to a Shareholder's Agreement upon the lapse of the restrictions hereunder, such Shares shall be maintained as provided in such Shareholder's Agreement.

9.2. **Restricted Stock Units.** The Committee may grant to Eligible Individuals Awards of Restricted Stock Units, which shall be evidenced by an Agreement. Each such Agreement shall contain such restrictions, terms and conditions as the Committee may, in its discretion, determine. Awards of Restricted Stock Units shall be subject to the terms and provisions set forth in this Section 9.2.

(a) Payment of Awards. Each Restricted Stock Unit shall represent the right of the Grantee to receive a payment upon vesting of the Restricted Stock Unit or on any later date specified by the Committee equal to the Fair Market Value of a Share as of the date the Restricted Stock Unit was granted, the vesting date or such other date as determined by the Committee at the time the Restricted Stock Unit was granted. The Committee may, at the time a Restricted Stock Unit is granted, provide a limitation on the amount payable in respect of each Restricted Stock Unit. The Committee may provide for the settlement of Restricted Stock Units in cash or with Shares having a Fair Market Value on the trading day immediately preceding the date of settlement equal to the payment to which the Participant has become entitled.

(b) No Sale or Transfer. The Grantee shall not sell, transfer, assign, exchange, pledge, encumber or otherwise dispose of an Award of Restricted Stock Units or any portion thereof.

10. **Performance Awards.**

10.1. Performance Units and Performance Share Units. The Committee, in its discretion, may grant Awards of Performance Units and/or Performance Share Units to Eligible Individuals, the terms and conditions of which shall be set forth in an Agreement.

(a) Performance Units. Performance Units shall be denominated in a specified dollar amount and, contingent upon the attainment of specified Performance Objectives within a specified Performance Cycle, represent the right to receive payment as provided in Sections 10.1(c) and (d) of the specified dollar amount or a percentage (which may be more than

100%) of the specified dollar amount depending on the level of Performance Objective attained; provided, however, that the Committee may at the time a Performance Unit is granted specify a maximum amount payable in respect of a vested Performance Unit. Each Agreement shall specify the dollar amount (or percentage thereof, if applicable) of Performance Units to which it relates, the Performance Objectives which must be satisfied in order for the Performance Units to vest and the Performance Cycle within which such Performance Objectives must be satisfied.

(b) Performance Share Units. Performance Share Units shall be denominated in Shares and, contingent upon the attainment of specified Performance Objectives within the Performance Cycle, each Performance Share Unit represents the right to receive payment as provided in Sections 10.1(c) and (d) of the Fair Market Value of a Share on the date the Performance Share was granted, the date the Performance Share Unit became vested or any other date specified by the Committee or a percentage (which may be more than 100%) of such amount depending on the level of Performance Objective attained; provided, however, that the Committee may at the time a Performance Share Unit is granted specify a maximum amount payable in respect of a vested

Performance Share Unit. Each Agreement shall specify the number of Performance Share Units to which it relates, the Performance Objectives which must be satisfied in order for the Performance Share Units to vest and the Performance Cycle within which such Performance Objectives must be satisfied.

(c) Vesting and Forfeiture. A Grantee shall become vested with respect to the Performance Share Units and Performance Units to the extent that the Performance Objectives set forth in the Agreement are satisfied for the Performance Cycle.

(d) Payment of Awards. Subject to Section 10.4(c), payment to Grantees in respect of vested Performance Share Units and Performance Units shall be made as soon as practicable after the last day of the Performance Cycle to which such Award relates or at such other time or times as the Committee may determine. Such payments may be made entirely in Shares valued at their Fair Market Value on the trading day immediately preceding the date of payment, entirely in cash or in such combination of Shares and cash as the Committee in its discretion shall determine at any time prior to such payment; provided, however, that if the Committee in its discretion determines to make such payment entirely or partially in Shares of Restricted Stock, the Committee shall determine the extent to which such payment will be in Shares of Restricted Stock and the terms of such Restricted Stock at the time the Award is granted.

10.2. Performance-Based Restricted Stock. The Committee, in its discretion, may grant Awards of Performance-Based Restricted Stock to Eligible Individuals, the terms and conditions of which shall be set forth in an Agreement between the Company and the Grantee. Each Agreement shall specify the number of Shares of Performance-Based Restricted Stock to which it relates, the Performance Objectives which must be satisfied in order for such Shares to vest and restrictions thereon to lapse, and the Performance Cycle within which such Performance Objectives must be satisfied, and may require that an appropriate legend be placed on Share certificates. Awards of Performance-Based Restricted Stock shall be subject to the following terms and provisions:

(a) Rights of Grantee. Shares of Performance-Based Restricted Stock granted pursuant to an Award hereunder shall be issued in the name of the Grantee as soon as reasonably practicable after the Award is granted provided that the Grantee has executed an Agreement evidencing the Award, the appropriate blank stock powers and, in the discretion of the Committee, a Shareholder's Agreement, an escrow agreement and any other documents which the Committee may require as a condition to the issuance of such Performance-Based Restricted Stock. If a Grantee shall fail to execute the Agreement evidencing an Award of Performance-Based Restricted Stock and any other documents which the Committee may require, or otherwise indicate acceptance of the Award of Performance-Based Restricted Stock in a manner prescribed by the Committee within the time period prescribed by the Committee at the time the Award is granted, the Award shall be null and void. At the discretion of the Committee, Shares issued in connection with an Award of Performance-Based Restricted Stock shall be deposited together with the stock powers with the Company as escrow agent (or other escrow agent designated by the Committee). Except as restricted by the terms of the Agreement and subject to the provisions of any applicable Shareholder's Agreement, upon delivery of the Shares to the escrow agent, the Grantee shall have, in the discretion of the Committee, all of the rights of a stockholder with respect to such Shares, including the right to receive all dividends or other distributions paid or made with respect to the Shares and the right to vote the Shares if and only if such Shares consist of Voting Common Stock.

(b) Lapse of Restrictions. To the extent that the Performance Objectives set forth in the Agreement are satisfied for the Performance Cycle, restrictions upon Performance-Based Restricted Stock awarded hereunder shall lapse at such time or times and on such terms, conditions and satisfaction of Performance Objectives as the Committee may, in its discretion, determine. In the event that the Performance Objectives set forth in the Agreement are not fully satisfied for the Performance Cycle, the Performance-Based Restricted Stock shall be forfeited to the extent set forth in the Agreement.

(c) Treatment of Dividends. At the time the Award of Performance-Based Restricted Stock is granted, the Committee may, in its discretion, determine that the payment to the Grantee of dividends, or a specified portion thereof, declared or paid on Shares represented by such Award which have been issued by the Company to the Grantee shall be (i) deferred until the lapsing of the restrictions imposed upon such Performance-Based Restricted Stock and (ii) held by the Company for the account of the Grantee until such time. In the event that dividends are to be deferred, the Committee shall determine whether such dividends are to be reinvested in shares of Stock (which shall be held as additional Shares of Performance-Based Restricted Stock) or held in cash. If deferred dividends are to be held in cash, there may be credited at the end of each year (or portion thereof) interest on the amount of the account at the beginning of the year at a rate per annum as the Committee, in its discretion, may determine. Payment of deferred dividends in respect of Shares of Performance-Based Restricted Stock (whether held in cash or as additional Shares of Performance-Based Restricted Stock), together with interest accrued thereon, if any, shall be made upon the lapsing of restrictions imposed on the Performance-Based Restricted Stock in respect of which the deferred dividends were paid, and any dividends deferred (together with any interest accrued thereon) in respect of any Shares of Performance-Based Restricted Stock shall be forfeited upon the forfeiture of such Shares.

(d) Delivery of Shares. Upon the lapse of restrictions on Shares of Performance-Based Restricted Stock awarded hereunder, the Company may maintain the Shares in book-entry form; provided, however, that if the Grantee so requests, the Committee shall cause a stock certificate to be delivered to the Grantee with respect to such Shares, free of all restrictions hereunder; and provided, further, that, if Shares of Performance-Based Restricted Stock are subject to a Shareholder's Agreement upon the lapse of the restrictions hereunder, such Shares shall be maintained as provided in such Shareholder's Agreement.

10.3. Cash Incentive Awards.

(a) Grants of Cash Incentive Awards. The Committee may, in its discretion, grant Cash Incentive Awards to Eligible Individuals, which shall represent the right, contingent upon the attainment of specified Performance Objectives within a specified Performance Cycle, to receive a payment of a specified dollar amount or a percentage (which may be more than 100%) of the specified dollar amount depending on the level of Performance Objective attained; provided, however, that the Committee may at the time a Cash Incentive Award is granted specify a maximum amount payable in respect of such Cash Incentive Award. The terms and conditions of each Cash Incentive Award shall be set forth in an Agreement between the Company and the Grantee. Each Agreement shall specify the dollar amount subject to such Award, the Performance Objectives which must be satisfied in order for the Award to be paid and the Performance Cycle within which such Performance Objectives must be satisfied. In the event that the Committee grants Cash Incentive Awards expressed as percentage interests in a bonus pool payment which is subject to the satisfaction of Performance Objectives, (i) the aggregate of all such percentage interests may not exceed 100% and (ii) the forfeiture or other reduction of the percentage interest of any Grantee in the bonus pool may not increase the amount of an Award paid to any other Grantee. The Committee may determine that different Performance Objectives are applicable to different Grantees with respect to a specified Performance Cycle.

(b) Payment of Awards. Each Cash Incentive Award to the extent earned shall be paid in a single lump sum cash payment or, if determined by the Committee at the time of grant, in a number of Shares with a Fair Market Value on the trading day immediately preceding the date of payment

equal to all or a portion of such cash amount (with any portion not paid in Shares to be paid in cash; provided, however, that if the Committee in its discretion determines to make such payment entirely or partially in Shares of Restricted Stock, the Committee shall determine the extent to which such payment will be in Shares of Restricted Stock and the terms of such Restricted Stock at the time the Award is granted) as soon as practicable following the Committee's certification described in Section 10.4(c) but in any event not later than the end of the fiscal year in which such certification is made.

10.4. Performance Objectives.

(a) Establishment. Performance Objectives for Performance Awards may be expressed in terms of (i) revenue, (ii) earnings per Share, (iii) net income per Share, (iv) Share price, (v) pre-tax profits, (vi) net earnings, (vii) net income, (viii) operating income, (ix)

cash flow, (x) earnings before interest, taxes, depreciation and amortization (EBITDA), (xi) sales, (xii) total stockholder return relative to assets, (xiii) total stockholder return relative to peers, (xiv) financial returns (including, without limitation, return on assets, return on equity and return on investment), (xv) cost reduction targets, (xvi) customer satisfaction, (xvii) customer growth, (xviii) employee satisfaction, (xviv) EBITDA margin, (xx) operating margin, (xxi) net margin, (xxii) gross margin, (xxiii) revenue growth, (xxiv) new contract win, (xxv) per-days sales outstanding, or (xxvi) any combination of the foregoing, or (xxvii) prior to the end of the Transition Period, such other criteria as the Committee may determine. Performance Objectives may be in respect of the performance of the Company, any of its Subsidiaries, any of its Divisions or any combination thereof. Performance Objectives may be absolute or relative (to prior performance of the Company or to the performance of one or more other entities or external indices) and may be expressed in terms of a progression within a specified range. The Performance Objectives with respect to a Performance Cycle shall be established in writing by the Committee by the earlier of (x) the date on which a quarter of the Performance Cycle has elapsed and (y) the date which is ninety days after the commencement of the Performance Cycle, and in any event while the performance relating to the Performance Objectives remain substantially uncertain.

(b) Effect of Certain Events. Unless otherwise provided by the Committee at the time the Performance Objectives in respect of a Performance Award are established, performance shall be adjusted to omit the effects of extraordinary items, gain or loss on the disposal of a business segment (other than provisions for operating losses or income during the phase-out period), unusual or infrequently occurring events and transactions that have been publicly disclosed and the cumulative effects of changes in accounting principles, all as determined in accordance with generally accepted accounting principles (to the extent applicable). In addition, at the time of the granting of a Performance Award or at any time thereafter, the Committee may provide for the manner in which performance will be measured against the Performance Objectives (or may adjust the Performance Objectives) to reflect the impact of specified corporate transactions (such as a stock split or stock dividend), special charges, and tax law changes; provided, that such provisions shall be permitted only to the extent permitted under Section 162(m) of the Code and the regulations promulgated thereunder without adversely affecting the treatment of any Performance Award as Performance-Based Compensation.

(c) Determination of Performance. Prior to the vesting, payment, settlement or lapsing of any restrictions with respect to any Performance Award that is intended to constitute Performance-Based Compensation made to a Grantee who is subject to Section 162(m) of the Code, the Committee shall certify in writing that the applicable Performance Objectives have been satisfied to the extent necessary for such Award to qualify as Performance Based Compensation. The Committee shall not be entitled to exercise any discretion otherwise authorized hereunder with respect to Awards intended to constitute Performance-Based Compensation made to a Grantee who is subject to Section 162(m) of the Code if the ability to exercise such discretion or the exercise of such discretion itself would cause the compensation attributable to such Awards to fail to qualify as Performance-Based Compensation. A Performance Award may be reduced in the discretion of the Committee at any time before payment or lapsing of restrictions.

10.5. No Sale or Transfer. The Grantee shall not sell, transfer, assign, exchange, pledge, encumber or otherwise dispose of an Award of Performance Units, Performance Share Units or Performance-Based Restricted Stock or Cash Incentive Awards or any portion thereof.

11. **Share Awards.**

11.1. Share Awards. The Committee may grant a Share Award to any Eligible Individual on such terms and conditions as the Committee may determine in its sole discretion. Share Awards may be made as additional compensation for services rendered by the Eligible Individual or may be in lieu of cash or other compensation to which the Eligible Individual is entitled from the Company. The Grantee shall not sell, transfer, assign, exchange, pledge, encumber or otherwise dispose of a Share Award or any portion thereof.

12. **Effect of a Termination of Employment or Service.**

The Agreement evidencing the grant of each Option and each Award shall set forth the terms and conditions applicable to such Option or Award upon a termination of the employment or service of the Optionee or Grantee by the Company, a Subsidiary or a Division (including a termination or change by reason of the sale of a Subsidiary or a Division). Unless set forth otherwise in an Agreement or as required by applicable law (including Section 409A of the Code), (1) the transfer of employment of an Optionee from the Company to a Subsidiary or from a Subsidiary to the Company (or among the Divisions of the Company or a Subsidiary) shall not constitute a termination of employment, and (2) if an individual is both an employee of the Company and/or a Subsidiary and a Director, his or her cessation of services in fewer than all such capacities shall not constitute a termination of his or her employment or services.

13. **Adjustment Upon Changes in Capitalization.**

13.1. In the event of a Change in Capitalization, the Committee shall make such adjustments, if any, as it determines are equitable and appropriate to (a) the maximum number and class of Shares or other stock or securities with respect to which Options or Awards may be granted under the Plan, (b) the maximum number and class of Shares or other stock or securities that may be issued upon exercise of Incentive Stock Options, (c) the maximum number and class of Shares or other stock or securities with respect to which Options or Awards may be granted to any Eligible Individual in any calendar year, (d) the number and class of Shares or other stock or securities which are subject to outstanding Options or Awards granted under the Plan and the exercise price therefor, if applicable, and (e) the Performance Objectives.

13.2. Any such adjustment in the Shares or other stock or securities (a) subject to outstanding Incentive Stock Options (including any adjustments in the exercise price) shall be made in such manner as not to constitute a modification as defined by Section 424(h)(3) of the Code and only to the extent permitted by Sections 422 and 424 of the Code or (b) subject to outstanding Awards that are subject to Section 409A of the Code shall be made only to the

extent permitted by Section 409A of the Code or (c) subject to outstanding Options or Awards that are intended to qualify as Performance-Based Compensation shall be made in such a manner as not to adversely affect the treatment of the Options or Awards as Performance-Based Compensation.

13.3. If, by reason of any such adjustment, a Grantee shall be entitled to, or an Optionee shall be entitled to exercise an Option, with respect to, new, additional or different shares of stock or securities of the Company or any other corporation, such new, additional or different shares shall thereupon be subject to all of the conditions, restrictions and performance criteria which were applicable to the Shares subject to the Award or Option, as the case may be, prior to such adjustment.

14. **Effect of Certain Transactions.**

14.1. Except as otherwise provided in an Agreement, in the event of (a) the liquidation or dissolution of the Company or (b) a merger or consolidation of the Company (a "Transaction"), the Plan and the Options and Awards issued hereunder shall continue in effect in accordance with their respective terms, except that following a Transaction either (i) each outstanding Option or Award shall be treated as provided for in the agreement entered into in connection with the Transaction or (ii) if not so provided in such agreement, each Optionee and Grantee shall be entitled to receive in respect of each Share subject to any outstanding Options or Awards, as the case may be, upon exercise of any Option or payment or transfer in respect of any Award, the same number and kind of stock, securities, cash, property or other consideration that each holder of a Share was entitled to receive in the Transaction in respect of a Share; provided, however, that, unless otherwise determined by the Committee, such stock, securities, cash, property or other consideration shall remain subject to all of the conditions, restrictions and performance criteria which were applicable to the Options and Awards prior to such Transaction. Without limiting the generality of the foregoing, the treatment of outstanding Options and Stock Appreciation Rights pursuant to this Section 14.1 in connection with a Transaction in which the consideration paid or distributed to the Company's stockholders is not entirely shares of common stock of the acquiring or resulting corporation may include the cancellation of outstanding Options and Stock Appreciation Rights upon consummation of the Transaction as long as, at the election of the Committee, (x) the holders of affected Options and Stock Appreciation Rights have been given a period of at least fifteen days prior to the date of the consummation of the Transaction to exercise the Options or Stock Appreciation Rights (whether or not they were otherwise exercisable) or (y) the holders of the affected Options and Stock Appreciation Rights are paid (in cash or cash equivalents) in respect of each Share covered by the Option or Stock Appreciation Right being cancelled an amount equal to the excess, if any, of the per share price paid or distributed to stockholders in the transaction (the value of any non-cash consideration to be determined by the Committee in its sole discretion) over the exercise price of the Option or Stock Appreciation Right. For avoidance of doubt, (1) the cancellation of Options and Stock Appreciation Rights pursuant to clause (y) of the preceding sentence may be effected notwithstanding anything to the contrary contained in this Plan or any Agreement and (2) if the amount determined pursuant to clause (y) of the preceding sentence is zero or less, the affected Option or Stock Appreciation Right may be cancelled without any payment therefor. The treatment of any Option or Award as provided in this Section 14.1 shall be conclusively presumed to be appropriate for purposes of Section 13.

14.2. Effective upon the consummation of an Initial Public Offering or any other conversion of all of the outstanding shares of the Company's Non-Voting Common Stock into Voting Common Stock, all outstanding Options shall automatically convert into Options to

purchase an equivalent number of shares of the Company's Voting Common Stock without any further action on the part of the Company or any Optionee.

14.3. The Committee may provide, in an Agreement or at any time thereafter, that Options or Awards may become vested and/or exercisable and may become free of restrictions, to the extent of all or any portion of such Option or Award, in the event of a change in ownership or effective control of the Company.

15. **Interpretation.**

15.1. Section 16 Compliance. The Plan is intended to comply with Rule 16b-3 promulgated under the Exchange Act and the Committee shall interpret and administer the provisions of the Plan or any Agreement in a manner consistent therewith. Any provisions inconsistent with such Rule shall be inoperative and shall not affect the validity of the Plan.

15.2. Section 162(m). Unless otherwise determined by the Committee at the time of grant, each Option, Stock Appreciation Right and Performance Award is intended to be Performance-Based Compensation. To the extent any provision of the Plan or any Agreement related to any Option, Stock Appreciation Right or Performance Award is inconsistent with Section 162(m) of the Code or the regulations promulgated thereunder, such provision shall be construed or deemed amended to the extent necessary to conform to such requirements, and no provision shall be deemed to confer upon the Committee discretion to increase the amount of compensation otherwise payable to an Eligible Individual in connection with any such Option or Award upon the attainment of the Performance Objectives.

15.3. Compliance With Section 409A. All Options and Awards granted under the plan are intended either not to be subject to Section 409A of the Code or, if subject to Section 409A of the Code, to be administered, operated and construed in compliance with Section 409A of the Code and any guidance issued thereunder. Notwithstanding this or any other provision of the Plan to the contrary, the Committee may amend the Plan or any Option or Award granted hereunder in any manner, or take any other action, that it determines, in its sole discretion, is necessary, appropriate or advisable to cause the Plan or any Option or Award granted hereunder to comply with Section 409A and any guidance issued thereunder. Any such action, once taken, shall be deemed to be effective from the earliest date necessary to avoid a violation of Section 409A and shall be final, binding and conclusive on all Eligible Individuals and other individuals having or claiming any right or interest under the Plan.

16. **Termination and Amendment of the Plan/Modification of Options and Awards.**

16.1. Plan Amendment or Termination. The Plan shall terminate on the day preceding the tenth anniversary of the date of its adoption by the Board, and no Option or Award may be granted thereafter. The Board may sooner terminate the Plan and the Board may at any time and from time to time amend, modify or suspend the Plan; provided, however, that:

(a) no such amendment, modification, suspension or termination shall impair or adversely alter any Options or Awards theretofore granted under the Plan, except with the consent of the Optionee or Grantee, nor shall any amendment, modification, suspension or

termination deprive any Optionee or Grantee of any Shares which he or she may have acquired through or as a result of the Plan; and

(b) to the extent necessary under any applicable law, regulation or exchange requirement, no amendment shall be effective unless approved by the stockholders of the Company in accordance with such applicable law, regulation or exchange requirement.

16.2. Repricing. The Board may, without stockholder approval, take any action under the Plan that constitutes a "repricing" within the meaning of the rules of the NASDAQ Stock Market.

16.3. Modification of Options and Awards. No modification of an Option or Award shall adversely alter or impair any rights or obligations under the Option or Award without the consent of the Optionee or Grantee, as the case may be; provided, however, that no action taken with respect to an Option or Award pursuant to Section 13 or 14.1 shall be deemed a modification that requires such consent of an Optionee or Grantee.

17. **Non-Exclusivity of the Plan.**

The adoption of the Plan by the Board shall not be construed as amending, modifying or rescinding any previously approved incentive arrangement or as creating any limitations on the power of the Board to adopt such other incentive arrangements as it may deem desirable, including, without limitation, the granting of stock options otherwise than under the Plan, and such arrangements may be either applicable generally or only in specific cases.

18. **Limitation of Liability.**

As illustrative of the limitations of liability of the Company, but not intended to be exhaustive thereof, nothing in the Plan shall be construed to:

(a) give any person any right to be granted an Option or Award other than at the sole discretion of the Committee;

(b) give any person any rights whatsoever with respect to Shares except as specifically provided in the Plan;

(c) limit in any way the right of the Company or any Subsidiary to terminate the employment or service of any person at any time; or

(d) be evidence of any agreement or understanding, expressed or implied, that the Company will employ any person at any particular rate of compensation or for any particular period of time.

19. **Regulations and Other Approvals; Governing Law.**

19.1. Except as to matters of federal law, the Plan and the rights of all persons claiming hereunder shall be construed and determined in accordance with the laws of the State of Delaware without giving effect to conflicts of laws principles thereof.

19.2. The obligation of the Company to sell or deliver Shares with respect to Options and Awards granted under the Plan shall be subject to all applicable laws, rules and regulations, including all applicable federal and state securities laws, and the obtaining of all such approvals by governmental agencies as may be deemed necessary or appropriate by the Committee.

19.3. The Board may make such changes as may be necessary or appropriate to comply with the rules and regulations of any government authority, or to obtain for Eligible Individuals granted Incentive Stock Options the tax benefits under the applicable provisions of the Code and regulations promulgated thereunder.

19.4. Each Option and Award is subject to the requirement that, if at any time the Committee determines, in its discretion, that the listing, registration or qualification of Shares issuable pursuant to the Plan is required by any securities exchange or under any state or federal law, or the consent or approval of any governmental regulatory body is necessary or desirable as a condition of, or in connection with, the grant of an Option or Award or the issuance of Shares, no Options or Awards shall be granted or payment made or Shares issued, in whole or in part, unless listing, registration, qualification, consent or approval has been effected or obtained free of any conditions as acceptable to the Committee.

19.5. Notwithstanding anything contained in the Plan or any Agreement to the contrary, in the event that the disposition of Shares acquired pursuant to the Plan is not covered by a then current registration statement under the Securities Act and is not otherwise exempt from such registration, such Shares shall be restricted against transfer to the extent required by the Securities Act and Rule 144 or other regulations thereunder. The Committee may require any individual receiving Shares pursuant to an Option or Award granted under the Plan, as a condition precedent to receipt of such Shares, to represent and warrant to the Company in writing that the Shares acquired by such individual are acquired without a view to any distribution thereof and will not be sold or transferred other than pursuant to an effective registration thereof under the Securities Act or pursuant to an exemption applicable under the Securities Act or the rules and regulations promulgated thereunder. The certificates evidencing any of such Shares shall be appropriately amended or have an appropriate legend placed thereon to reflect their status as restricted securities as aforesaid.

20. **Miscellaneous.**

20.1. Multiple Agreements. The terms of each Option or Award may differ from other Options or Awards granted under the Plan at the same time, or at some other time. The Committee may also grant more than one Option or Award to a given Eligible Individual during the term of the Plan, either in addition to, or in substitution for, one or more Options or Awards previously granted to that Eligible Individual.

20.2. Withholding of Taxes.

(a) At such times as an Optionee or Grantee recognizes taxable income in connection with the receipt of Shares or cash hereunder (a "Taxable Event"), the Optionee or Grantee shall pay to the Company an amount equal to the federal, state and local

income taxes and other amounts as may be required by law to be withheld by the Company in connection with the Taxable Event (the "Withholding Taxes") prior to the issuance, or release from escrow, of such Shares or the payment of such cash. The Company shall have the right to deduct from any payment of cash or delivery of Shares to an Optionee or Grantee an amount equal to the Withholding Taxes in satisfaction of the obligation to pay Withholding Taxes. The Committee may provide in an Agreement at the time of grant, or at any time thereafter, that the Optionee or Grantee, in satisfaction of the obligation to pay Withholding Taxes to the Company, may elect to have withheld a portion of the cash then payable to him or her or the Shares (if the Shares are then Listed Securities) then issuable to him or her, in either case having an aggregate Fair Market Value equal to the Withholding Taxes (but, in the case of withholding of Shares, only to the extent such withholding complies with Section 13(k) of the Exchange Act and other applicable laws, rules and regulations).

(b) If an Optionee makes a disposition, within the meaning of Section 424(c) of the Code and regulations promulgated thereunder, of any Share or Shares issued to such Optionee pursuant to the exercise of an Incentive Stock Option within the two-year period commencing on the day after the date of the grant or within the one-year period commencing on the day after the date of transfer of such Share or Shares to the Optionee pursuant to such exercise, the Optionee shall, within ten days of such disposition, notify the Company thereof, by delivery of written notice to the Company at its principal executive office.

20.3. Effective Date. The effective date of the Plan shall be the date of Board approval as listed on the first page of the Plan, subject only to the approval by the affirmative vote of the holders of a majority of the securities of the Company present, or represented, and entitled to vote at a meeting of stockholders duly held in accordance with the applicable laws of the State of Delaware within twelve months after the adoption of the Plan by the Board.

20.4. Post-Transition Period. Following the end of the Transition Period, any Option, Stock Appreciation Right or Performance Award granted under the Plan which is intended to be Performance-Based Compensation, shall be subject to the approval of the material terms of the Plan by the stockholders of the Company in accordance with Section 162(m) of the Code and the regulations promulgated thereunder.

20.5. Shareholder's Agreement. Unless the Committee determines otherwise, prior to the consummation of an Initial Public Offering, it shall be a condition to issuance of any Shares to any Grantee or Optionee pursuant to any Award or the exercise of any Option that the Grantee or Optionee, as applicable, shall have become a party to a Shareholder's Agreement which shall remain operative in accordance with its terms notwithstanding the lapse of any other restrictions on such Shares pursuant to the terms of the Plan or the relevant Agreement.

Attachment A
Provisions Applicable to Optionees
Resident in California

The following additional terms shall apply if required by applicable law to (i) Options or Awards that may result in the issuance of Shares granted on any date the Shares are not a Listed Security and (ii) any Shares issued pursuant to such Awards or upon exercise of such Options on any date the Shares are not a Listed Security, which are granted or issued to persons resident in California as of the date of grant or issuance, as applicable (each such person, a "California Recipient"):

1. Any Option granted to any California Recipient that is exercised before security holder approval is obtained or any Share that is issued pursuant to an Award to any California Recipient before security holder approval is obtained must be rescinded if security holder approval is not obtained in the manner described in Section 20.3 of the Plan. Such securities shall not be counted in determining whether such approval is obtained.

2. Notwithstanding anything in the Plan to the contrary, in the case of an Option granted to any California Recipient, the number of Shares covered by each outstanding Option, as well as the price per Share covered by each outstanding Option, and in the case of Shares allocated to a California Recipient pursuant to an Award, the number of Shares allocated, shall be proportionately adjusted in the event of a stock split, reverse stock split, stock dividend, combination, recapitalization, reclassification or other distribution of the Company's equity securities without receipt of consideration by the Company, of or on the Shares; provided, however, that conversion of any convertible securities of the Company shall not be deemed to have been "effected without receipt of consideration."

3. With respect to an Option granted to any California Recipient who is not an Officer, Director or Consultant, such Option shall become exercisable, or any repurchase option in favor of the Company shall lapse, at the rate of at least 20% per year over five years from the date of grant.

4. Notwithstanding anything in the Plan to the contrary, in the case of an Option granted to any California Recipient, Options shall not be exercisable after the expiration of 120 months from the date of grant (five years in the case of an Incentive Stock Option granted to a Ten-Percent Stockholder).

5. The following rules shall apply to an Option granted to any California Recipient or to stock issued to a California Recipient upon exercise of an Option, in the event of a termination of the California Recipient's employment or services with the Company other than for Cause (as defined in the applicable Agreement):

(a) If such termination was for reasons other than death or disability, the California Recipient shall have at least thirty days after the date of such termination (but in no event later than the expiration of the term of such Option established by the Committee as of the date of grant) to exercise such Option.

(b) If such termination was on account of the death or disability of the California Recipient, the holder of the Option may, but only within six months from the date of such termination (but in no event later than the expiration date of the term of such Option established by the Committee as of the date of grant), exercise the Option to the extent the California Recipient was otherwise entitled to exercise it at the date of such termination. To the extent that the California Recipient was not entitled to exercise the Option at the date of termination, or if the holder does not exercise such Option to the extent so entitled within six months from the date of termination, the Option shall terminate, and the Shares underlying the unexercised portion of the Option shall revert to the Plan.

6. To the extent required by applicable law, the Company shall provide financial statements at least annually to each California Recipient during the period such person has one or more Options or Awards outstanding, and in the case of an individual who acquired Shares pursuant to an Award or pursuant to the exercise of an Option under the Plan, during the period such individual owns such Shares. The Company shall not be required to provide such information if the issuance of Options and Awards under the Plan is limited to key employees whose duties in connection with the Company assure their access to equivalent information.

7. Unless defined below or otherwise in this Attachment, capitalized terms shall have the meanings set forth in the Plan. For purposes of this Attachment, (a) "Officer" means a person who is an officer of the Company within the meaning of Section 16(a) of the Exchange Act and the rules and regulations promulgated thereunder, and (b) "Consultant" means any person, including an advisor, who is engaged by the Company or a Subsidiary to render services and is compensated for such services, and any Director of the Company whether compensated for such services or not.

Amendment No. 1 to the Amended and Restated
Ikaria Holdings, Inc. 2010 Long Term Incentive Plan

WHEREAS, Ikaria, Inc. (formerly, Ikaria Holdings, Inc.) a Delaware corporation (the "Company"), has adopted the Amended and Restated Ikaria Holdings, Inc. 2010 Long Term Incentive Plan (the "Plan") for the purpose of granting long term incentive awards, including options to purchase shares of common stock, par value $0.01 per share, of the Company (the "Common Stock") to the employees and consultants of the Company or its subsidiaries or affiliates;

WHEREAS, Section 16.1 of the Plan provides that the Board of Directors of the Company (the "Board") may at any time and from time to time amend, modify or suspend the Plan.

NOW THEREFORE,

(a) All references to "Ikaria Holdings, Inc." in the Plan are hereby be deleted and replaced with "Ikaria, Inc."

(b) The definition of "Initial Public Offering" in Section 2 of the Plan is hereby amended and restated in its entirety to read as follows:

" "Initial Public Offering" means the first public offering of shares of Voting Common Stock and Non-Voting Common Stock or either of them. An Initial Public Offering shall be deemed to have been consummated if and when the registration statement in connection with such public offering shall have been declared effective and the offering of securities thereunder shall have closed in accordance with the terms of the related underwriting agreement."

(c) Section 4.1 of the Plan is hereby amended and restated in its entirety to read as follows:

"Number of Shares Authorized for Issuance; Limitations on Options and Awards. Subject to any adjustment as provided in the Plan, the Shares to be issued under the Plan may be, in whole or in part, authorized but unissued Shares or issued Shares which shall have been reacquired by the Company and held by it as treasury shares. The aggregate number of Shares that may be made the subject of Awards or Options granted under the Plan shall initially be 4,300,000 (calculated as set forth in Section 4.2): provided, that, prior to the termination of the Plan in accordance with Section 16.1, that number shall be increased automatically on January 1st of each year commencing on January 1, 2011, in an amount such that the aggregate number of Shares that may be made the subject

of Awards or Options granted under the Plan as of such January 1 shall be equal to the sum of (i) 3% of the total number of shares of Voting Common Stock ,Non-Voting Common Stock, Series A Preferred and Series B Preferred issued and outstanding on December 31st of the immediately preceding calendar year and (ii) the number of Recaptured Shares that have not previously been made the subject of Awards or Options hereunder; provided, that the Board may in its sole discretion reduce the amount of the increase in any particular year. In no event may more than 20 million Shares be made the subject of Incentive Stock Options under the Plan (calculated as set forth in Section 4.2)."

(d) The text "(as such term is defined in the Company's Amended and Restated Certificate of Incorporation as may be amended from time to time)" in the second sentence of Section 6.2 of the Plan is hereby deleted in its entirety.

[Remainder of page intentionally left blank]

IN WITNESS WHEREOF, this Amendment has been executed by the undersigned, thereunto duly authorized, effective as of July 30, 2010.

IKARIA, INC.

/s/ Matthew M. Bennett
Name: Matthew M. Bennett
Title: Senior Vice President and Secretary

AMENDMENT NO. 2 TO THE AMENDED AND RESTATED
IKARIA, INC. 2010 LONG TERM INCENTIVE PLAN

WHEREAS, Ikaria, Inc., a Delaware corporation (the "Company"), has adopted the Amended and Restated Ikaria, Inc. 2010 Long Term Incentive Plan, as amended (the "Plan") for the purpose of granting long term incentive awards, including options to purchase shares of common stock, par value $0.01 per share, of the Company (the "Common Stock") to the employees and consultants of the Company or its subsidiaries or affiliates;

WHEREAS, Section 16.1 of the Plan provides that the Board of Directors of the Company (the "Board") may at any time and from time to time amend, modify or suspend the Plan.

NOW THEREFORE, the second sentence of Section 4.1 of the 2010 Plan is hereby amended by deleting the text "The aggregate number of Shares that may be made the subject of Awards or Options granted under the Plan shall initially be 4,300,000 (calculated as set forth in Section 4.2)", and substituting the following text in lieu thereof: "The aggregate number of Shares that may be made the subject of Awards or Options granted under the Plan shall initially be 5,300,000 (calculated as set forth in Section 4.2)".

[Remainder of page intentionally left blank]

IN WITNESS WHEREOF, this Amendment has been executed by the undersigned, thereunto duly authorized, effective as of October 25, 2010.

IKARIA, INC.

/s/ Matthew M. Bennett
Name: Matthew M. Bennett
Title: Senior Vice President and Secretary

Amendment No. 3 to the Amended and Restated
Ikaria Holdings, Inc. 2010 Long Term Incentive Plan

WHEREAS, Ikaria, Inc. a Delaware corporation (the "Company"), has adopted the Amended and Restated Ikaria Holdings, Inc. 2010 Long Term Incentive Plan, as amended (the "Plan") for the purpose of granting long term incentive awards to the employees and consultants of the Company or its subsidiaries or affiliates;

WHEREAS, Section 16.1 of the Plan provides that the Board of Directors of the Company (the "Board") may at any time and from time to time amend, modify or suspend the Plan.

NOW THEREFORE, Section 4.1 of the Plan is hereby replaced in its entirety with the following:

> "4.1 Number of Shares Authorized for Issuance; Limitations on Options and Awards. Subject to any adjustment as provided in the Plan, the Shares to be issued under the Plan may be, in whole or in part, authorized but unissued Shares or issued Shares which shall have been reacquired by the Company and held by it as treasury shares. The aggregate number of Shares that may be made the subject of Awards or Options granted under the Plan as of September 22, 2011 shall be 9,500,000 (calculated as set forth in Section 4.2): provided, that, prior to the termination of the Plan in accordance with Section 16.1, that number shall be increased automatically on January 1st of each year commencing on January 1, 2012, in an amount such that the aggregate number of Shares that may be made the subject of Awards or Options granted under the Plan as of such January 1 shall be equal to the sum of (i) 3% of the total number of shares of Voting Common Stock, Non-Voting Common Stock, Series A Preferred and Series B Preferred issued and outstanding on December 31st of the immediately preceding calendar year and (ii) the number of Recaptured Shares that have not previously been made the subject of Awards or Options hereunder; provided, that the Board may in its sole discretion reduce the amount of the increase in any particular year. In no event may more than 20 million Shares be made the subject of Incentive Stock Options under the Plan (calculated as set forth in Section 4.2)."

[Remainder of page intentionally left blank]

IN WITNESS WHEREOF, this Amendment has been executed by the undersigned, thereunto duly authorized, effective as of October 7th, 2011.

IKARIA, INC.

/s/ Matthew M. Bennett
Name: Matthew M. Bennett
Title: Senior Vice President and Secretary

Exhibit 10.8

EXECUTION VERSION

Confidential Materials omitted and filed separately with the
Securities and Exchange Commission. Double asterisks denote omissions.

AMENDED AND RESTATED

LICENSE AND COMMERCIALIZATION AGREEMENT

BY AND AMONG

IKARIA DEVELOPMENT SUBSIDIARY ONE LLC

AND

BIOLINERX LTD.

AND

BIOLINE INNOVATIONS JERUSALEM L.P.

AUGUST 26, 2009

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AMENDED AND RESTATED
LICENSE AND COMMERCIALIZATION AGREEMENT

This Amended and Restated License and Commercialization Agreement (the "Agreement") is entered into this 26th day of August, 2009, by and among **Ikaria Development Subsidiary One LLC**, a Delaware limited liability company having a principal place of business at 6 State Route 173, Clinton, NJ 08809, USA ("Ikaria"), **BioLineRx Ltd.**, a corporation organized and existing under the laws of the State of Israel and having a principal place of business at 19 Hartum Street, P.O. Box 45158, Jerusalem 91450, Israel ("BioLineRx Ltd."), and **BioLine Innovations Jerusalem L.P.**, a limited partnership organized and existing under the laws of the State of Israel and having a principal place of business at 19 Hartum Street, P.O. Box 45158, Jerusalem 91450, Israel ("BioLine Innovations"; together with BioLineRx Ltd., "BioLineRx").

INTRODUCTION

WHEREAS, BioLineRx owns or controls certain intellectual property rights covering a liquid polymer composed of Sodium Alginate and Ca-D-Gluconate (designated by BioLineRx as "BL-1040");

WHEREAS, BioLineRx is currently developing the Product (as defined below) as a medical device for the direct treatment of cardiac tissue following acute myocardial infarction;

WHEREAS, BioLineRx is concluding the safety and clinical trials of the Product that were initiated by BioLineRx prior to the Effective Date (as defined below);

WHEREAS, BioLineRx desires to grant to Ikaria the worldwide exclusive rights to Develop, Manufacture, and Commercialize Products (as such capitalized terms are defined below); and

WHEREAS, Ikaria desires to obtain such exclusive rights in accordance with the terms and conditions of this Agreement.

NOW, THEREFORE, BioLineRx and Ikaria agree as follows:

Article I
Definitions; Interpretation

When used in this Agreement, each of the following capitalized terms has the meaning set forth in this Article I:

Section 1.1 "Affiliate" shall mean, with respect to a Party, any Person that controls, is controlled by, or is under common control with such Party. For purposes of this Section 1.1, "control" shall refer to (a) in the case of a Person that is a corporate entity, direct or indirect ownership of more than fifty percent (50%) of the stock, shares or membership units having the right to vote for the election of a majority of the directors of such Person, and (b) in the case of a Person that is an entity, but is not a corporate entity, the possession, directly or indirectly, of the

power to direct, or cause the direction of, the management or policies of such Person, whether through the ownership of voting securities, by contract or otherwise.

Section 1.2 "BGN License Agreement" shall mean that certain License Agreement, dated January 10, 2005, as amended, by and among BioLine Jerusalem L.P. and B.G. Negev Technologies and Applications Ltd. ("BGN") on behalf of Ben Gurion University.

Section 1.3 "BioLineRx Know-How" shall mean all Know-How that is (a) necessary or useful for the Development, Manufacture, or Commercialization of any Product and (b) either (i) is Controlled by BioLineRx as of the Effective Date or (ii) BioLineRx comes to Control during the term of this Agreement.

Section 1.4 "BioLineRx Patent Rights" shall mean Patent Rights that claim or disclose BioLineRx Know-How, including the Patent Rights listed in Exhibit B.

Section 1.5 "BioLineRx Intellectual Property" shall mean BioLineRx Patent Rights (including Patent Rights in the Sublicensed IP), and BioLineRx Know-How (including Know-How in the Sublicensed IP).

Section 1.6 "Business Day" shall mean a day that is not a Saturday, a Sunday or a day on which banking institutions in New York, New York, USA are authorized by law to remain closed.

Section 1.7 "Commercialization" or "Commercialize" shall mean any activities directed to marketing, promoting, distributing, importing, exporting, or selling a product.

Section 1.8 "Commercially Reasonable Efforts" shall mean the efforts, expertise and resources normally used by a Party to Develop, Manufacture and Commercialize a product owned by it or to which it has rights, which is of similar market potential at a similar stage in its development or product life, taking into account issues of safety and efficacy, product profile, difficulty in developing the product, competitiveness of the marketplace for the product, the proprietary position of the product, the regulatory structure involved, the availability and level of reimbursement for such treatment by Third Party payors or health insurance plans, the potential total profitability of the applicable product(s) marketed or to be marketed and other relevant factors affecting the cost, risk and timing of Development and the total potential reward to be obtained if a product is Commercialized. The Parties agree that Commercially Reasonable Efforts shall require a Party to expend efforts, expertise and resources that such Party would normally expend to Develop, use, Manufacture and Commercialize a product owned by it or to which it has rights, taking into account the foregoing factors.

Section 1.9 "Confidential Information" shall mean, with respect to a disclosing Party, all Know-How or other information (whether or not patentable) regarding such Party's technology, products, business information or objectives (whether disclosed before or after the Effective Date) that is of a confidential and proprietary nature, including reports and audits under Section 4.3, the Development Plan, the Commercialization Plan, the terms of this Agreement, and all proprietary tangible materials (and data and information associated therewith) of such Party. Notwithstanding the foregoing, Confidential Information shall not include Know-How or other information that:

(a) was rightfully known or used by the receiving Party or its Affiliates without an obligation of confidentiality prior to its date of disclosure to the receiving Party as demonstrated by contemporaneous written records; or

(b) either before or after the date of the disclosure to the receiving Party is lawfully disclosed to the receiving Party or its Affiliates by sources other than the disclosing Party rightfully in possession of such information and not bound by confidentiality obligations to the disclosing Party; or

(c) either before or after the date of the disclosure to the receiving Party or its Affiliates is or becomes published or otherwise is or becomes part of the public domain through no breach hereof on the part of the receiving Party or its Affiliates; or

(d) is independently developed by or for the receiving Party or its Affiliates without reference to or use of the Confidential Information of the disclosing Party as demonstrated by contemporaneous written records.

Section 1.10 "Control" shall mean the legal authority or right of a Party or its Affiliates to grant a license or sublicense of intellectual property rights to the other Party, or to provide tangible material to or otherwise disclose proprietary or trade secret information to such other Party, without breaching the terms of any agreement with a Third Party. For the avoidance of doubt, BioLineRx Controls the Sublicensed IP.

Section 1.11 "Cover" or "Covered" shall mean, with respect to a Patent Right and a product, that, in the absence of ownership of (with a retained right to exploit), or a license granted under, a Valid Claim included in such Patent Right, the Manufacture, Development, Commercialization, use, sale, import, or offer for sale, as applicable, of such product would infringe such Valid Claim in the country where such activity occurs.

Section 1.12 "Development" or "Develop" shall mean development activities, including test method development and stability testing, toxicology, formulation, optimization, quality assurance/quality control development, statistical analysis, clinical studies, regulatory affairs, product approval, and registration.

Section 1.13 "Development Term" shall mean the term of development of Products by Ikaria.

Section 1.14 "EU" shall mean the European Union and all the member states thereof, as it may be comprised from time to time.

Section 1.15 "EU Milestone Conditions" shall mean (a) satisfaction of all requirements for [**], (b) [**] set forth therein, **and** (c) [**].

Section 1.16 "Executive Officers" shall mean the Chief Executive Officer of Ikaria (or a senior executive officer of Ikaria designated by Ikaria) and the Chief Executive Officer of BioLineRx (or a senior executive officer of BioLineRx designated by BioLineRx).

Section 1.17 "FDA" shall mean the United States Food and Drug Administration or any successor agency thereof.

Section 1.18 "Field" shall mean any and all uses described or claimed in the BioLineRx Patent Rights.

Section 1.19 "First Commercial Sale" shall mean, with respect to a Product in a country, the first commercial sale of such Product by Ikaria, its Affiliates, distributors, agents or Licensees in such country. Sales for clinical trial purposes or compassionate or similar use shall not be considered to constitute a First Commercial Sale.

Section 1.20 Intentionally Omitted

Section 1.21 Intentionally Omitted

Section 1.22 Intentionally Omitted

Section 1.23 Intentionally Omitted.

Section 1.24 Intentionally Omitted."

Section 1.25 "Know-How" shall mean any tangible or intangible know-how, expertise, information, inventions, discoveries, documents and other works of authorship, copyrights, trade secrets, data, or materials, whether proprietary or not, including ideas, concepts, formulas, methods, procedures, designs, technologies, compositions, plans, applications, technical data, data generated in clinical trials, samples, chemical compounds and biological materials and all derivatives, modifications and improvements thereof.

Section 1.26 "Knowledge" shall mean, with respect to a Party, the Party's actual knowledge together with any knowledge of any of the Party's officers or director-level employees, that a Person in such party's position would be expected to obtain given the exercise of reasonably prudent scientific and business diligence in accordance with the standards of companies of such Party's size in such Party's industry.

Section 1.27 "Licensee" shall mean any Person to whom Ikaria licenses its rights under this Agreement in the manner provided in Section 2.1, including any Third Party contractors.

Section 1.28 "Manufacturing" or "Manufacture" shall mean any activities associated with the production, manufacture, supply, processing, filling, packaging, labeling, shipping, or storage of a product or any components thereof, including process and formulation development, process validation, stability testing, manufacturing scale-up, development and commercial manufacture and analytical development, product characterization, quality assurance and quality control development, testing, and release.

Section 1.29 "Net Sales" shall mean, with respect to a Product, the gross amounts billed by Ikaria, its Affiliates, or Licensees in respect of sales of such Product by Ikaria and its Affiliates or Licensees to unrelated Third Parties, in each case less the following deductions:

(a) Trade, cash, or quantity discounts (including amounts incurred in connection with government mandated rebate programs) actually allowed and taken with respect to such sales;

(b) Tariffs, duties, excises, sales taxes or other taxes imposed upon and paid with respect to the production, sale, delivery, or use of the Product (excluding national, state, or local taxes based on income);

(c) Amounts repaid or credited by reason of billing corrections, rejections, defects, recalls, or returns (due to spoilage, damage, expiration of useful life or otherwise) or because of chargebacks, refunds or retroactive price reductions and allowances for wastage replacement and bad debts;

(d) Portions of invoices sales amounts included in Net Sales in prior periods that are actually written off by Ikaria, its Affiliates, or licenses as uncollectible; and

(e) Postage, freight, shipping, insurance, and other transportation related charges incurred in shipping a Product to Third Parties.

Such amounts shall be determined from the books and records of Ikaria, its Affiliates, or Licensees, maintained in accordance with generally accepted accounting principles, consistently applied. For the avoidance of doubt, in no event will fines, penalties or other monetary damages assessed against Ikaria, its Affiliates or Licensees by any governmental authority for violation of any applicable law, result in an appropriate deduction to Net Sales.

If one or more Products is sold as part of a Combination Product (as defined below), the Net Sales from the Combination Product, for the purposes of determining royalty payments, shall be determined by multiplying the Net Sales (as determined above) of the Combination Product, during the applicable royalty reporting period, by the fraction, A/(A+B), where A is the average sale price of the Product(s) when sold separately in finished form and B is the average sale price of the other components included in the Combination Product when sold separately in finished form, in each case in the applicable country during the applicable royalty reporting period or, if sales of both the Product(s) and the other components did not occur in such country in such period, then in the most recent royalty reporting

period in which sales of both occurred. If such average sale price cannot be determined for both the Product(s) and all other components included in such Combination Product, Net Sales for the purposes of determining royalty payments shall be calculated by multiplying the Net Sales of the Combination Product by the fraction of C/(C+D) where C is the fair market value of the Product(s) and D is the fair market value of all other components included in the Combination Product. In such event, the Parties shall negotiate in good faith to arrive at a determination of the respective fair market values of the Product(s) and all other components included in the Combination Product. If the Parties are unable to agree on such determination within sixty (60) days, then such matter shall be resolved as provided in Article IX.

As used above, the term "Combination Product" means any therapeutic medical product that includes both (i) one or more Product(s) and (ii) other component(s).

Section 1.30 "On-Going Phase I/II Trial" shall mean that certain clinical trial of a Product that was initiated by BioLineRx prior to and that is ongoing as of the Effective Date, the protocol for which is attached hereto as Schedule 1.30.

Section 1.31 "Other On-Going Trials" shall mean those pre-clinical and CMC trials (other than the On-Going Phase I/II Trial) that were initiated by BioLineRx prior to, and that are ongoing as of, the Effective Date, descriptions of which are attached hereto as Schedule 1.31.

Section 1.32 "Party" shall mean BioLineRx or Ikaria; "Parties" shall mean BioLineRx and Ikaria.

Section 1.33 "Patent Rights" shall mean United States and foreign patents and patent applications (including provisional applications) and all substitutions, divisionals, continuations, continuations-in-part, reissuances, reexaminations, registrations, renewals, confirmations, supplementary protection certificates and extensions thereof.

Section 1.34 "Person" shall mean any natural person or any corporation, company, partnership, joint venture, firm, university, other entity, governmental authority, or subdivision thereof.

Section 1.35 "Pivotal Clinical Trial" shall mean a randomized, controlled clinical trial of a Product designed to demonstrate statistically significant clinical efficacy and safety in human patients (in conjunction with performance of a therapeutic procedure) pursuant to a clinical study agreed with the FDA, which trial the FDA accepts as a pivotal clinical trial necessary for Regulatory Approval of such Product. An outline of the structure of the initial Pivotal Clinical Trial is attached as Schedule 1.35.

Section 1.36 "Primary Indication" shall mean the diagnosis, prevention, mitigation, or treatment of injury to myocardial tissue via the administration of a Product to a human patient.

Section 1.37 "Product" shall mean a liquid polymer composed of Sodium Alginate and Ca-D-Gluconate (designated by BioLineRx as "BL-1040"), or any back-ups or second-generation polymers or polymer combinations thereof that is Developed under the Development Program.

Section 1.38 "Regulatory Approval" shall mean, with respect to a jurisdiction, the approval of the applicable Regulatory Authority required to market and sell a Product in such jurisdiction. For clarity, Regulatory Approval for a Product shall occur:

(a) in the United States, on the date when the FDA approves a Premarket Approval (PMA) application;

(b) in Europe, on the date when such Product may first be placed on the market as a medical device (as such terms are defined in Art. 1 Paragraphs 2(a) and (h) of Directive 93/42/EEC, as amended) bearing the CE marking according to Art. 17 of Directive 93/42/EEC, as amended, in any member state of the EU; and

(c) in Japan, on the date when the Ministry of Health approves a marketing authorization.

Section 1.39 "Regulatory Authority" shall mean any national (*e.g.*, the FDA), supra-national or other regulatory agency or governmental entity involved in the granting of Regulatory Approval for, or in the regulation of human clinical studies of, therapeutic medical devices.

Section 1.40 "Royalty Term" shall mean, with respect to a Product in a country of the Territory, the period of time commencing on the First Commercial Sale of such Product in such country and ending upon the earlier of (a) the expiration of the last-to-expire Valid Claim in the BioLineRx Patent Rights that Covers the sale or use of such Product in the Field in such country, or (b) the date of a judicial determination from which no appeal can be taken of invalidity of a set of claims in the BioLineRx Patent Rights that Cover the sale or use of such Product in the Field in such country and that are asserted through litigation (whether in an infringement action, a declaratory judgment action, or otherwise) to exclude a Third Party from selling or using a product in the Field in such country.

Section 1.41 "Sublicensed IP" shall mean that portion of the BioLineRx Intellectual Property licensed to BioLineRx pursuant to the BGN License Agreement.

Section 1.42 "Successful Completion" shall mean:

(a) with respect to the On-Going Phase I/II Trial, no treatment-related safety findings during the treatment period and the six (6) month follow up period, that were considered by the Independent Safety Monitoring Board for the On-Going Phase I/II Trial (in accordance with and subject to the Independent Safety Monitoring Board Charter attached hereto as Schedule 1.42(a)) to be of sufficient concern to discontinue the On-Going Phase I/II Trial;

(b) with respect to the Interim Analysis of the Pivotal Clinical Trial/Phase IIb Proof of Concept, safety and efficacy data from completion of all patients at the [**] follow up demonstrates more than a [**] probability of meeting pre-specified endpoints at [**] in the Pivotal Clinical Trial, and no apparent safety signal in the treatment group for the entire cohort at all times;

(c) with respect to the Pivotal Clinical Trial for the Primary Indication, safety and efficacy data from completion of all patients at the [**] follow up meets the primary endpoint and demonstrates a positive benefit-to-risk ratio to enable FDA submission; and

(d) with respect to all other clinical trials of a Product, that the JDC has determined that the final results of such clinical trial have achieved the success criteria established by the JDC with respect to such clinical trial.

Section 1.43 "Territory" shall mean the entire world.

Section 1.44 "Third Party" shall mean any Person other than a Party or any of its Affiliates or Licensees.

Section 1.45 "Valid Claim" shall mean a claim of any issued, unexpired patent that has not been revoked or held unenforceable or invalid by a decision of a court or governmental agency of competent jurisdiction from which no appeal can be taken, or with respect to which an appeal is not taken within the time allowed for appeal, and that has not been disclaimed or admitted to be invalid or unenforceable through reissue, reexamination, disclaimer, or otherwise.

Section 1.46 Additional Definitions. Each of the following terms is defined in the section of this Agreement indicated below:

Term	Section
"Agreement"	Preamble
"Bankruptcy Code"	Section 2.5
"BGN"	Section 1.2
"BioLineRx"	Preamble
"BL-1040"	Section 1.37
"Breaching Party"	Section 8.2
"Combination Product"	Section 1.29
"Commercialization Plan"	Section 3.7
"Competitive Infringement"	Section 5.3(a)
"Effective Date"	Section 2.1
"Existing Product Agreements"	Section 2.3
"Ikaria"	Preamble
"Development Plan"	Section 3.1
"Development Program"	Section 3.1
"Force Majeure Event"	Section 10.7
"Indemnified Party"	Section 10.1(c)
"Indemnifying Party"	Section 10.1(c)
"Invalidity Claim"	Section 5.3(d)
"Joint Development Committee" or "JDC"	Section 3.2
"Joint Manufacturing Committee" or "JMC"	Section 3.6(c)
"Lead Party"	Section 5.3(e)
"Losses"	Section 10.1(a)
"New Indication"	Section 2.4
"New Indication Invention"	Section 5.1(a)
"Non-Breaching Party"	Section 8.2
"OCS"	Section 2.1
"SEC"	Section 6.1
"Severed Clause"	Section 10.11
"Technology Exchange"	Section 3.5
"Technology Exchange Plan"	Section 3.5
"Third Party Payment"	Section 4.2(b)

Section 1.47 Interpretation. Whenever the context may require, any pronoun shall include the corresponding masculine, feminine, and neuter forms. The words "include", "includes" and "including" shall be deemed to be followed by the phrase "without limitation". The word "will" shall be construed to have the same meaning and effect as the word "shall". The word "or" shall be construed to have the same meaning and effect as "and/or". This Agreement has been prepared jointly with the assistance of counsel and shall not be strictly construed against either Party. The captions or headings of the sections or other subdivisions hereof are inserted only as a matter of convenience or for reference and shall have no effect on the meaning of the provisions hereof. Unless the context requires otherwise, (a) any definition of or reference to any agreement, instrument, or other document herein shall be construed as referring to such agreement, instrument, or other document as from time to time amended, supplemented, or otherwise modified (subject to any restrictions on such amendments, supplements, or modifications set forth herein or therein), (b) any reference to any laws herein shall be construed as referring to any law, statute, rule, regulation, ordinance, or other pronouncement having the effect of law of any federal, national, multinational, state, provincial, county, city, or other political subdivision, domestic or foreign, as they from time to time may be enacted, repealed, or amended, (c) any reference herein to any Person shall be construed to include the Person's successors and assigns, (d) the words "herein", "hereof", and "hereunder", and words of similar import, shall be construed to refer to this Agreement in its entirety and not to any particular provision hereof, (e) any reference herein to the words "mutually agree" or "mutual written agreement" shall not impose any obligation on either Party to agree to any terms relating thereto or to engage in discussions relating to such terms except as such Party may determine in such Party's sole discretion, and (f) all references herein to Articles, Sections, Exhibits, or Schedules shall be construed to refer to Articles, Sections, Exhibits, and Schedules of this Agreement.

Article II
Grant of Rights

Section 2.1 BioLineRx License Grant to Ikaria; Consent of OCS. Subject to the terms and conditions of this Agreement, including the consent of the Office of the Chief Scientist of the State of Israel ("OCS"), BioLineRx hereby grants to Ikaria the exclusive, royalty-bearing right and license in the Territory under the BioLineRx Intellectual Property (including, for clarity, a sublicense under the Sublicensed IP) to Develop, Manufacture and Commercialize Products for use in the Field. Subject to the consent of BioLineRx, which consent shall not be unreasonably withheld, conditioned or delayed, the foregoing license includes the right to grant sublicenses under the BioLineRx Intellectual Property, provided that, with respect to sublicenses granted under the Sublicensed IP, Ikaria shall (a) grant such sublicenses only for consideration and at arm's-length transactions, and (b) grant such sublicenses only pursuant to written agreements that contain such terms and conditions as may be required for Ikaria to comply with this Agreement. BioLineRx shall use its best efforts to obtain the written consent of the OCS to this Agreement within [**] days after August 26th, 2009, which consent must be in a form that is satisfactory to each Party. If the OCS has still not provided such consent during such [**] days, Ikaria shall have the right to require BioLineRx to continue to use best efforts to obtain such consent within the subsequent [**] day period. In addition, (i) Ikaria shall have the right to have a representative present at all interactions between BioLineRx's representatives and the OCS relating to such consent, (ii) BioLineRx shall (A) provide Ikaria with a reasonable opportunity to review and approve the request for consent submitted to the OCS and (B) keep Ikaria fully

informed as to the progress of such request for consent and shall consult with Ikaria in good faith with respect thereto, (iii) BioLineRx shall not engage in any activities or discussions with any Third Party relating to the subject matter of this Agreement, including pursuing any other transactions relating to the BioLineRx Intellectual Property, without Ikaria's consent, and (iv) Ikaria shall have the right, prior to the Effective Date, to unilaterally modify this Agreement to comply with the specific, formal, written requests of the OCS, provided that such modifications have no detrimental financial impact on BioLineRx under this Agreement. Notwithstanding BioLineRx's obligation to exercise best efforts to obtain the consent from the OCS as described above, BioLineRx shall not be required to (y) agree to any request by the OCS that would require BioLineRx to pay to the OCS an aggregate amount of more than $[**] or (z) obtain a consent based on the characterization of this Agreement as a "transfer of know-how outside of Israel" under Section 19B of the Israeli Law for the Encouragement of Industrial Research & Development, 1984. Notwithstanding anything herein to the contrary, subject to Section 8.6, the provisions of this Agreement other than this Section 2.1, Section 2.2, Article VII, Section 8.6 and Article X shall not be effective until such consent has been obtained and each Party has delivered the certificate set forth in Section 7.8 (the "Effective Date").

Section 2.2 Non-Competition. During the term of this Agreement, BioLineRx shall not, within the Territory, directly or indirectly (including through its Affiliates), conduct research or discovery activities, Develop, Manufacture (except as set forth in Section 3.6), Commercialize, or grant any rights or options or provide assistance to any Third Party to conduct research or discovery activities, Develop, Manufacture (except as set forth in Section 3.6) or Commercialize, (a) the Product or (b) any compound, substance, polymer, or product (whether pharmaceutical or device in nature) the method of action or effect of which is similar to any Product.

Section 2.3 Existing Product Agreements. BioLineRx hereby agrees that, upon the written request of Ikaria, BioLineRx shall assign to Ikaria each of the agreements listed in Schedule 2.3 attached hereto (the "Existing Product Agreements"), and all of its rights, title, and interest therein. BioLineRx shall cooperate with Ikaria, including by executing and recording documents, as may be necessary to effectuate such assignments and the exercise by Ikaria of its rights under the Existing Product Agreements.

Section 2.4 Intentionally Omitted.

Section 2.5 Section 365(n) of the Bankruptcy Code. All rights and licenses granted under or pursuant to any Section of this Agreement, including under this Article II and with respect to any BioLineRx Intellectual Property subject to Technology Exchange under Section 3.5, are rights to "intellectual property" (as defined in Section 101(35A) of Title 11 of the United States Code (such Title, the "Bankruptcy Code")). Each of Ikaria and BioLineRx hereby acknowledges "embodiments" of such intellectual property for purposes of Section 365(n) of the Bankruptcy Code shall include (a) copies of research data, (b) laboratory samples, (c) product samples, (d) formulas, (e) laboratory notes and notebooks, (f) data and results related to clinical studies, (g) regulatory filings and approvals, (h) rights of reference in respect of regulatory filings and approvals, (i) research data and results, and (j) marketing, advertising, and promotional materials, in each case, that relate to such intellectual property. Each Party shall retain and may fully exercise all of its rights and elections under the Bankruptcy Code or analogous legislation in any other jurisdiction. Upon the institution by or against BioLineRx of any assignment for the

benefit of creditors, composition, or any bankruptcy, reorganization, arrangement, insolvency, or similar proceedings under the laws of any jurisdiction, Ikaria shall further be entitled to a complete duplicate of, or complete access to, as appropriate, any such intellectual property (including embodiments thereof), and such intellectual property and embodiments, if not already in its possession, shall be promptly delivered to Ikaria, unless BioLineRx elects to continue, and continues, to perform all of its obligations under this Agreement.

Section 2.6 Retained Rights. Except as otherwise specifically provided for in this Agreement, each Party retains all rights and licenses to exploit its own intellectual property.

Article III
Development; Manufacturing; Commercialization

Section 3.1 General. Ikaria shall be solely responsible for conducting and funding all Development activities pursuant to the Development Plan, and shall have the sole right to Develop, Manufacture, and Commercialize Products in the Field in the Territory. Subject to its obligations under Section 3.8, Ikaria shall prepare a non-binding plan (the "Development Plan") for the Development of Product(s) (the "Development Program"). The Development Plan shall include an estimated budget setting forth Ikaria's anticipated development costs. Ikaria shall provide BioLineRx with a copy of its then-current Development Plan at least [**] per year, but no later than [**] days following the beginning of each year. The initial Development Plan is attached hereto as Schedule 3.1, which shall be non-binding, including any timelines or milestones that may be included therein. In addition, Ikaria shall, within [**] days after the Effective Date, provide BioLineRx with a revised draft protocol for the Interim Analysis of the Pivotal Clinical Trial/Phase IIb Proof of Concept and the Pivotal Clinical Trial, after taking into account any comments BioLineRx may wish to provide based on the initial draft of the protocol attached hereto as Schedule 1.35, that would include modifications designed to maximize the likelihood of obtaining reasonable reimbursement for one or more Products in any one or more of the following countries [**]. Upon the Successful Completion of the Interim Analysis of the Pivotal Clinical Trial/Phase IIb Proof of Concept, or, failing that, upon the Successful Completion of the Pivotal Clinical Trial, Ikaria shall, within [**] days thereafter, submit a formal written request for a reimbursement price for one or more Product(s) to the applicable governmental agency in one or more of the following countries: [**]

Section 3.2 Joint Development Committee.

(a) The Parties shall establish a Joint Development Committee (the "Joint Development Committee" or "JDC"), comprised of [**] representatives of Ikaria and [**] representatives of BioLineRx, to oversee the Development of Products. Each Party shall make its initial designation of its representatives not later than [**] days after the Effective Date. Each Party may change any one or more of its representatives to the Joint Development Committee at any time upon notice to the other Party.

(b) The JDC shall meet at least [**] during the Development Term or more or less frequently as the JDC may agree. The JDC may meet in person or by means of a telephone or video conference call. One meeting of the JDC per year shall be held in person at Ikaria's

headquarters in Clinton, NJ and one meeting of the JDC per year shall be held in person at BioLineRx's headquarters in Israel, provided, that the Parties' representatives may participate in person, via telephone, or video conference in their discretion. Each Party shall use reasonable efforts to cause its representatives to attend the meetings of the JDC. If a representative of a Party is unable to attend a meeting, such Party may designate an alternate to attend such meeting in place of the absent representative. Each Party shall bear its own costs with respect to its participation on the JDC. Prior to every meeting of the JDC, Ikaria will provide to the JDC detailed reports describing Ikaria's current clinical and development activities and plans.

(c) The JDC shall be the vehicle by which BioLineRx may offer insight and guidance to Ikaria with respect to (i) establishing the Development Plan setting forth the Development Program's objectives and the activities to be conducted, (ii) reviewing and updating the Development Plan from time to time, (iii) monitoring the progress and results of the Development Program, (iv) determining future Development Program activities, including Development activities relating to Manufacturing, to be conducted during the Development Term, and (v) establishing success criteria for the clinical trials (other than those for which success criteria are set forth in this Agreement), and determining whether the results of such clinical trials have achieved the applicable success criteria.

(d) The JDC shall only act unanimously, with each Party given one (1) vote regardless of the number of representatives. If, however, the JDC is unable to reach agreement with respect to any matter within [**] days, the matter shall be referred to the Parties' respective Executive Officers for resolution. If the Executive Officers are not able to resolve any such matter by consensus within [**] days following referral, Ikaria's Executive Officer shall have the right to decide the matter taking into account Ikaria's obligation to use Commercially Reasonable Efforts under Section 3.8.

Notwithstanding anything in this Section 3.2, neither Party shall have a unilateral right to resolve any dispute involving the breach or alleged breach of this Agreement, to amend or modify this Agreement or the Parties' respective rights and obligations hereunder or, except as expressly provided in this Section 3.2, any Development Plan or the Parties' respective rights and obligations thereunder.

Section 3.3 On-Going Trials. BioLineRx shall retain control of, bear all costs relating to the On-Going Phase I/II Trial and the Other On-Going Trials, and shall exercise Commercially Reasonable Efforts to continue and complete the On-Going Phase I/II Trial and the Other On-Going Trials, which shall be managed by BioLineRx. BioLineRx may modify the On-Going Phase I/II Trial and the Other On-Going Trials, including any changes to the protocols therefor, only with the prior written consent of Ikaria, which consent shall not be unreasonably withheld, conditioned or delayed.

Section 3.4 Regulatory Matters. Ikaria shall prepare and submit all filings with Regulatory Authorities relating to Products, which filings shall be in Ikaria's name, provided that Ikaria shall provide BioLineRx [**] days prior notice to enable BioLineRx to review and provide any comments on such submissions. With respect to regulatory matters concerning Products, BioLineRx shall cooperate with Ikaria in the preparation and support of each application for Regulatory Approval and shall provide Ikaria with such reasonable assistance as Ikaria may

request. For example, upon Ikaria's request, BioLineRx shall describe the materials in sufficient and reasonable detail as requested by Ikaria, the Manufacturing techniques and other appropriate characteristics of Products (and the components thereof), and provide Ikaria with such other information related to the Products, including materials, chemistry, Manufacturing, technical dossier and controls data, batch records, analytical and quality control, device master files (if applicable), data from the On-Going Phase I/II Trial or Other On-Going Trials, or other information as Ikaria may reasonably request.

Section 3.5 Technology Exchange.

(a) As soon as reasonably practicable after Ikaria's written request, BioLineRx shall complete the activities assigned to BioLineRx as set forth on the technology exchange plan attached hereto as Exhibit A (the "Technology Exchange Plan"), to effect the transfer to Ikaria (or Ikaria's designee(s)) of all embodiments of and information relating to BioLineRx Intellectual Property reasonably necessary for the exercise of Ikaria's rights under the license granted pursuant to Section 2.1, including the Manufacturing of Products ("Technology Exchange"). BioLineRx shall make available to Ikaria (or Ikaria's designee(s)) such number of technical personnel as may be set forth in the Technology Exchange Plan to answer any questions or provide instruction as reasonably requested by Ikaria (or Ikaria's designee(s)) concerning the items delivered pursuant to this Section 3.5, in connection with the Development, Manufacture and Commercialization of Products hereunder. Each Party shall bear its own costs with respect to the Technology Exchange.

(b) The Joint Development Committee shall be responsible for coordinating the technology exchange activities under the Technology Transfer Plan. Each Party shall cooperate with the other Party in such other Party's conduct of technology exchange activities under the Technology Exchange Plan.

(c) If Ikaria desires that BioLineRx provide technology exchange services beyond the scope of the Technology Exchange Plan, BioLineRx shall provide such services on terms to be agreed upon in good faith by the Parties. Notwithstanding the foregoing, BioLineRx shall provide Ikaria with reasonable access to BioLineRx's employees and consultants involved prior to the Effective Date and during the term of this Agreement with the Development of any Product.

Section 3.6 Manufacturing.

(a) Ikaria shall be solely responsible for the Manufacture of Products for Development or for Commercialization in the Field in the Territory, which Ikaria may conduct itself or through Affiliates or Licensees.

(b) BioLineRx Ltd. shall have the option (either directly or through an Affiliate), exercisable in its sole discretion no later than [**] months prior to the date on which Ikaria intends to file for Regulatory Approval in the U.S., to Manufacture Product pursuant to the terms of a supply agreement to be negotiated in good faith by the Parties, provided that (i) BioLineRx may exercise the foregoing option only to the extent that it has the demonstrated ability to manufacture the Product, including compliance with cGMP and all applicable laws and

regulations, including those of the FDA and EMEA, (ii) BioLineRx shall bear all expenses required to establish and qualify the BioLineRx manufacturing site, including the costs of scale-up batches, process validation batches and stability batches, (iii) BioLineRx shall not be entitled to assign such option or to utilize subcontract manufacturing, and (iv) neither Party shall have any obligation to enter into such agreement unless all of the terms and conditions thereof are acceptable to both Parties. If BioLineRx Ltd. exercises such option and the Parties enter into a supply agreement, (x) Ikaria shall be required to purchase no less than twenty percent (20%) of its requirements for the Product from BioLineRx, and (y) the per unit price for the Product shall be the [**], provided that the price shall not exceed [**] percent ([**]%) of the Net Sales price per unit of Product; provided, further, that if BioLineRx at any time shall fail to supply Product on time or such supply is otherwise disrupted, the minimum purchase requirement set forth in the preceding clause (x) shall no longer apply. Any clinical supply provided to Ikaria by BioLineRx would be provided at cost.

(c) The Parties will discuss the most efficient structure for the Manufacture and supply of Product for Development and Commercialization purposes. If the Parties determine that coordination in Manufacturing is appropriate, the Parties will establish a Joint Manufacturing Committee (the "Joint Manufacturing Committee" or "JMC") to coordinate Manufacturing efforts. If established, the JMC would be comprised of [**] representatives of Ikaria and [**] representatives of BioLineRx, to oversee the Manufacturing of Products. Each Party would make its initial designation of its representatives not later than [**] days after the Parties agreed to establish the JMC. Each Party shall designate as its representatives individuals who have the requisite experience and knowledge to discuss the Manufacturing of Products. Each Party would be permitted to change any one or more of its representatives to the JMC at any time upon notice to the other Party.

(d) The JMC would meet at least [**] or more or less frequently as the JMC may agree. The location of such meetings shall be as mutually agreed by the Parties. The JMC may also meet by means of a telephone or video conference call. Each Party shall use reasonable efforts to cause its representatives to attend the meetings of the JMC. If a representative of a Party is unable to attend a meeting, such Party may designate an alternate to attend such meeting in place of the absent representative. Each Party would bear its own costs with respect to its participation on the JMC.

(e) The JMC would only act unanimously. If, however, the JMC is unable to reach agreement with respect to any matter within [**] days, the matter shall be referred to the Parties' respective Executive Officers for resolution. If the Executive Officers are not able to resolve any such matter by consensus within [**] days following referral, Ikaria's Executive Officer shall have the right to decide the matter taking into account Ikaria's obligation to use Commercially Reasonable Efforts under Section 3.8.

Section 3.7 Commercialization. Ikaria shall be solely responsible for conducting, itself or through Affiliates or Licensees, the Commercialization of Products in the Field in the Territory, including (a) contracting with customers and booking sales, (b) setting the price and terms and conditions under which a Product may be sold to customers, and (c) handling of managed care accounts, and, subject to Section 1.29, Section 4.2(b), Section 5.2(d), Section 5.3(e) and Section 10.1(b), as between the Parties, Ikaria shall bear all costs associated therewith. Ikaria shall

produce and update from time to time a comprehensive Commercialization plan (the "Commercialization Plan"), which shall include plans for Commercializing Product in each major market in which Ikaria does not then have a presence. The Commercialization Plan shall include a preliminary timeline for the initial Commercialization of Products, which is intended as a planning and informational tool and shall not constitute a binding obligation on Ikaria, and shall be subject to adjustment by Ikaria from time to time, provided, that, Ikaria shall provide BioLineRx with prior written notice of any material proposed change to a timeline. The most recent preliminary Commercialization Plan is attached hereto as Schedule 3.7.

Section 3.8 Efforts. Ikaria shall use Commercially Reasonable Efforts, either itself or through Affiliates or Licensees, (a) to Develop at least one Product in the Territory and (b) to Commercialize at least one Product in the Territory.

Article IV
Financial Provisions

Section 4.1 Milestone Payments.

(a) Development and Regulatory Milestones. With respect to each of the following milestones, Ikaria shall pay BioLineRx the corresponding payment set forth below within [**] days after the achievement by Ikaria, its Affiliates or Licensees of such milestone:

MILESTONE			PAYMENT
1.	Effective Date		$ 7,000,000
2.	Successful Completion of On-Going Phase I/II Trial		$ 10,000,000
3.	[**]		$ [**]
	[**]	$ 12,500,000	
	[**]	$ [**]	
4.	[**]		$ [**]
5.	[**]		$ [**]

6.	[**]	$ [**]
Total Development and Regulatory Milestone Payments		132,500,000

(b) Commercialization Milestones. Ikaria shall pay each of the following milestone payments to BioLineRx within [**] days after the achievement of such milestone:

MILESTONE	PAYMENT
1. Annual Net Sales in Territory exceed $[**] in a Calendar Year	$ [**]
2. Annual Net Sales in Territory exceed $[**] in a Calendar Year	$ [**]
3. Annual Net Sales in Territory exceed $[**] in a Calendar Year	$ [**]

Each of the milestones set forth in Section 4.1(a) and Section 4.1(b) shall be paid only once regardless of the number of Products that achieve such milestone.

Section 4.2 Royalties on Net Sales of Products. During the Royalty Term applicable to each Product, and subject to adjustment as set forth in Section 4.2(b), Ikaria shall pay to BioLineRx royalties on a Product-by-Product basis, with the amount of such royalties calculated as a percentage of Net Sales in a calendar year for such Product as set forth below:

Net Sales	Royalty
Up to $[**]	[**]%
>$[**] to $[**]	[**]%
>$[**]	[**]%

(a) Royalties Payable Only Once. The obligation to pay royalties is imposed only once with respect to Net Sales of the same unit of a Product.

(b) Royalty Reductions for Third Party Payments. Ikaria shall use Commercially Reasonable Efforts to avoid any Third Party Payments. Ikaria shall provide BioLineRx written notice within [**] days of its receipt of any request or demand that Ikaria, its Affiliates or any Licensee obtain a license or immunity from suit from any Third Party in order for Ikaria, its Affiliates, or any Licensee to exercise or use the rights granted to Ikaria herein. If Ikaria is required to obtain a license or immunity from suit from any Third Party in order for Ikaria, its Affiliates, or any Licensee to exercise or use the rights granted to Ikaria herein, and Ikaria, its Affiliates, or any Licensee pays any Third Party any up-front fee, milestone, royalty, or other payment (each, a "Third Party Payment") in connection with such license or immunity from suit, Ikaria shall have the right to set off against any amounts payable to BioLineRx under this Article IV [**] percent ([**]%) of any Third Party Payments provided that in no event will the royalty paid to BioLineRx on Net Sales in the applicable country fall below [**] percent ([**]%). If the amount of Third Party Payments that Ikaria is entitled to set off exceeds the amount otherwise payable to BioLineRx at any given time, or is limited by the foregoing [**] percent ([**]%), Ikaria shall be entitled to carry over the excess for set off against amounts payable to BioLineRx in subsequent periods until Ikaria has been credited for the full amount it is entitled to set off. Prior to paying any Third Party Payment, the Parties shall obtain an analysis from their

respective counsel in respect of the validity of the claim of any Third Party seeking Third Party Payments. If the Parties are unable to agree on an assessment of the claim, the Parties shall jointly engage mutually acceptable independent patent counsel not regularly employed by either Party to assess such claims. Ikaria shall substitute the decision of such independent patent counsel for that of its own counsel with respect to deciding whether to obtain a license or immunity from suit from any Third Party in order for Ikaria, its Affiliates, or any Licensee to exercise or use the rights granted to Ikaria herein.

(c) Duration of Payments. The amounts payable to BioLineRx under Section 4.2 shall be paid on a Product-by-Product and country-by-country basis until the expiration of the Royalty Term for such Product in such country.

(d) Price Concessions. Ikaria shall not, and shall ensure that its Affiliates and Licensees do not, sell or distribute the Product at a discount (including in the form of government mandated rebates) (with or without consideration) in return substantially for (i) concessions or consideration received in transactions involving products or services other than the Product or (ii) concessions from any government or governmental authority relating to products or services other than the Product.

Section 4.3 Reports and Accounting.

(a) Reports; Payments. Ikaria shall deliver to BioLineRx, within [**] days after the end of each calendar quarter, reasonably detailed written accountings of Net Sales of Products that are subject to payment obligations to BioLineRx for such calendar quarter. Such quarterly reports shall indicate (i) gross sales and Net Sales on a country-by-country basis, (ii) the calculation of payment amounts owed to BioLineRx from such gross sales and Net Sales, and (iii) any amounts set off pursuant to Section 4.2(b) against payments owed to BioLineRx. When Ikaria delivers such accounting to BioLineRx, Ikaria shall also deliver all amounts due under Section 4.2 to BioLineRx for the calendar quarter. All payments shall be made by wire transfer to the account specified in Schedule 4.3(a).

(b) Audits by BioLineRx. Ikaria shall keep, and shall require its Affiliates and Licensees to keep, complete and accurate records of the most recent [**] years relating to gross sales and Net Sales and all information relevant under Section 4.1 and Section 4.2. For the sole purpose of verifying amounts payable to BioLineRx, BioLineRx shall have the right no more than [**] per calendar year, at BioLineRx's expense, to engage independent accountants to review such records in the location(s) where such records are maintained by Ikaria, its Affiliates, and its Licensees upon reasonable notice and during regular business hours. Prior to any review conducted pursuant to this Section 4.3(b), BioLineRx's accountants shall have entered into a written agreement with Ikaria limiting the use of such records to verification of the accuracy of payments due under this Agreement and prohibiting the disclosure of any information contained in such records to a Third Party and to BioLineRx for a purpose other than as set forth in this Section 4.3(b). The right to audit any royalty report or quarterly report or payment shall extend for [**] years from the end of the calendar year in which such royalty report or quarterly report was delivered or such payment made. Results of such

review shall be made available to Ikaria. If the review reflects an underpayment to BioLineRx, such underpayment shall be promptly remitted to BioLineRx. Likewise, if the review reflects an overpayment, Ikaria shall be entitled

to reduce any subsequent payments by the amount of the overpayment. If the underpayment to BioLineRx is equal to or greater than [**] percent ([**]%) of the amount that was otherwise due, BioLineRx shall be entitled to have Ikaria reimburse BioLineRx's reasonable out-of-pocket costs of such review.

Section 4.4 Currency Amounts. All dollar ($) amounts specified in this Agreement are United States Dollar amounts.

Section 4.5 Currency Exchange. With respect to sales of Products invoiced in U.S. Dollars and other amounts received or paid by Ikaria, its Affiliates or Licensees in U.S. Dollars, such amounts and the amounts payable hereunder shall be expressed in U.S. Dollars. With respect to sales of Products invoiced in a currency other than U.S. Dollars and other amounts received or paid by Ikaria, its Affiliates or Licensees in a currency other than U.S. Dollars, such amounts and the amounts payable hereunder shall be expressed in their U.S. Dollar equivalent calculated using the applicable rate of exchange reported by *The Wall Street Journal* (Eastern U.S. edition) on the last Business Day of the calendar quarter to which the report under Section 4.3(a) relates. All payments hereunder shall be made in U.S. Dollars.

Section 4.6 Tax Withholding. The Parties shall use all reasonable and legal efforts to reduce tax withholding on payments made to BioLineRx. The Parties agree to cooperate in good faith to provide one another with such documents and certifications as are reasonably necessary to enable Ikaria to minimize any withholding tax obligations. Ikaria shall promptly provide to BioLineRx documentation of the payment of any withholding taxes that are paid pursuant to this Section 4.6, including copies of receipts or other evidence reasonably required and sufficient to allow BioLineRx to document such tax withholdings adequately for purposes of claiming foreign tax credits and similar benefits.

Section 4.7 Upfront Payments Received Under Sublicenses. If Ikaria receives an upfront payment consideration under a sublicense granted to a Third Party under this Agreement, Ikaria shall pay to BioLineRx ten percent (10%) of any such payment within 30 days after actual receipt thereof from the Third Party.

Article V
Intellectual Property Ownership, Protection and Related Matters

Section 5.1 Ownership of Inventions.

(a) Intentionally Omitted.

(b) Intentionally Omitted.

(c) Inventorship. Questions of inventorship shall be resolved in accordance with United States patent laws. In the event of a dispute regarding inventorship, if the Parties are unable to resolve the dispute, the Parties shall jointly engage mutually acceptable independent patent counsel not regularly employed by either Party to resolve such dispute. The decision of such independent patent counsel shall be binding on the Parties with respect to the issue of inventorship.

(d) Further Actions and Assignments. Each Party shall take all further actions and execute all assignments requested by the other Party and reasonably necessary or desirable to vest in the other Party the ownership rights set forth in this Section 5.1.

Section 5.2 Prosecution and Maintenance of Patent Rights.

(a) Intentionally Omitted.

(b) BioLineRx Intellectual Property. Upon the Effective Date, Ikaria shall assume responsibility for the management of the preparation, filing prosecution and maintenance of any and all patent applications, including any interference proceedings related thereto, included in the BioLineRx Intellectual Property (including, for clarity, the Sublicensed IP, BioLineRx Patent Rights and patents and patent applications that claim or disclose BioLineRx Know-How).

(c) BioLineRx Step-in Right. If Ikaria, on a country-by-country basis, declines to file and prosecute, or elects not to take actions necessary to avoid abandonment of, any patent applications or maintain any patent in any country, in each case for which it has responsibility under Section 5.2(a) or Section 5.2(b), it shall give BioLineRx reasonable notice to this effect sufficiently in advance to permit BioLineRx to undertake such filing and prosecution without a loss of rights, and thereafter BioLineRx may, upon written notice to Ikaria, file and prosecute such patent applications and maintain such patents in such country. If BioLineRx files, prosecutes or maintains any such patent application or patent in such country and any resulting Valid Claim of BioLineRx Patent Rights constitutes the only BioLineRx Patent Rights Covering the Product in such country (*i.e.*, there are no other BioLineRx Patent Rights Covering the Product in such country), then [**].

If BioLineRx exercises the foregoing step-in right following the election by Ikaria to abandon all existing BioLineRx Patent Rights in a given country, Ikaria shall, within [**] days following BioLineRx's written request, notify BioLineRx in writing whether Ikaria intends to Commercialize a Product in the Field in such country. If Ikaria notifies BioLineRx that Ikaria has no intent to Commercialize a Product in the Field in such country, BioLineRx may, upon written notice to Ikaria within [**] days of receipt of Ikaria's notice of lack of intent, exercise a right to directly Commercialize a Product in the Field in such country. If BioLineRx provides Ikaria with such notice: [**].

(d) Costs and Expenses. Ikaria shall pay the costs and expenses of preparing, filing, prosecuting, and maintaining the Patent Rights covered by Section 5.2(a) or Section 5.2(b), [**].

(e) Cooperation Between Parties. Each Party agrees to cooperate with the other with respect to the preparation, filing, prosecution and maintenance of Patent Rights pursuant to this Section 5.2, including the execution of all such documents and instruments and the performance of such acts as may be reasonably necessary in order to permit the other Party to continue any preparation, filing, prosecution or maintenance of such Patent Rights, including Patent Rights that such Party has elected not to pursue, as provided for in subsections (a), (b) and (c) above. In addition, the filing, prosecuting and maintaining Party in subsections (a), (b) and (c) above shall promptly forward to the other Party copies of any substantive correspondence and actions prepared for or received from the U.S. Patent and Trademark Office or any foreign patent office that may materially affect the Patent Rights being prosecuted or maintained. The other Party's patent counsel may provide comments to the filing, prosecuting and maintaining Party. If any comments by the other Party's patent counsel are provided in sufficient time for the filing, prosecuting and maintaining Party to reflect such comments in its correspondence or response, and such comments are reasonably directed to maximizing the coverage of the claims of the Patent Rights being prosecuted or maintained, the filing, prosecuting and maintaining Party shall reflect such comments in its correspondence or response, if its patent counsel deems it prudent to do so.

(f) Coordination with BioLineRx pursuant to the Sublicensed IP. With respect to any Sublicensed IP which Ikaria is responsible for filing, prosecuting, and maintaining, Ikaria shall:

(i) consult with BioLineRx regarding the preparation, filing, and prosecution of all patent applications, and the maintenance of all patents, included within such Sublicensed IP, including the content, timing, and jurisdiction of the filing of such patent applications and their prosecution, and other details and overall global strategy pertaining to the procurement and maintenance of Patent Rights in such Sublicensed IP, and shall file, prosecute, and maintain all such Patent Rights through a law or patent attorney firm selected by Ikaria and approved by BioLineRx (and BioLineRx shall exercise its rights under the BGN License Agreement as may be necessary to obtain BGN's approval); and

(ii) provide BioLineRx with copies of all patent applications that claim or disclose such Sublicensed IP, and BioLineRx shall exercise its rights under the BGN License Agreement to ensure that BGN cooperates in a timely manner with Ikaria's efforts to register such Patent Rights, including by causing BGN to execute any documents as may be required for such purpose.

BioLineRx shall take all actions required to remain in compliance with the BGN License Agreement in connection with the foregoing.

Section 5.3 Third Party Infringement.

(a) Notice. Each Party shall promptly report in writing to the other Party during the term of this Agreement any (i) known or suspected infringement of any of the BioLineRx Patent

Rights or (ii) unauthorized use of any of the BioLineRx Know-How of which such Party becomes aware, including, in the case of either clause (i) or clause (ii) involving, or that may reasonably lead to, the Development, Manufacture, use or Commercialization of a product or product candidate that is or may be competitive with a Product in the Field ("Competitive Infringement"), and shall provide the other Party with all available evidence supporting such infringement, suspected infringement, unauthorized use or suspected unauthorized use.

(b) BioLineRx Intellectual Property; Step-in Rights.

(i) Ikaria shall have the first right, but not the obligation, to initiate a suit or take other appropriate action that either Party reasonably believes is required to protect BioLineRx Intellectual Property from Competitive Infringement . Ikaria shall give BioLineRx sufficient advance notice of its intent to file any such suit or take any such action, and the reasons therefor, and shall provide BioLineRx with an opportunity to make suggestions and comments regarding such suit or action. Thereafter, Ikaria shall keep BioLineRx informed, and shall from time to time consult with BioLineRx regarding the status of any such suit or action and shall provide BioLineRx with copies of all material documents (*i.e.*, complaints, answers, counterclaims, material motions, orders of the court, memoranda of law and legal briefs, interrogatory responses, depositions, material pre-trial filings, expert reports, affidavits filed in court, transcripts of hearings and trial testimony, trial exhibits and notices of appeal) filed in, or otherwise relating to, such suit or action. Any recovery obtained as a result of any proceeding pursuant to this subsection (b)(i), by settlement or otherwise, shall be applied in the following order of priority: (A) first, each Party shall be reimbursed, on a pro rata basis, for all costs incurred by such Party in connection with such suit; and (B) second, [**].

(ii) If Ikaria chooses not to initiate a suit or take other appropriate action under subsection (b)(i) above to protect BioLineRx Intellectual Property from Competitive Infringement, Ikaria will so notify BioLineRx of its intention, in which case BioLineRx shall have the right to initiate such suit or take such other appropriate action. BioLineRx shall give Ikaria sufficient advance notice of its intent to file any such suit or take any such action, and the reasons therefor, and shall provide Ikaria with an opportunity to make suggestions and comments regarding such suit or action. Thereafter, BioLineRx shall keep Ikaria informed, and shall from time to time consult with Ikaria regarding the status of any such suit or action and shall provide Ikaria with copies of all material documents (*i.e.*, complaints, answers, counterclaims, material motions, orders of the court, memoranda of law and legal briefs, interrogatory responses, depositions, material pre-trial filings, expert reports, affidavits filed in court, transcripts of hearings and trial testimony, trial exhibits and notices of appeal) filed in, or otherwise relating to, such suit or action. Any recovery obtained as a result of any proceeding pursuant to this subsection (b)(ii), by settlement or otherwise, shall be applied in the following order of priority: (A) first, each Party shall be reimbursed, on a pro rata basis, for all costs incurred by such Party in connection with such suit; and (B) second, any remainder shall be shared [**]% for BioLineRx and [**]% for Ikaria.

(iii) If BioLineRx chooses not to initiate a suit or take other appropriate action under subsection (b)(ii) above to protect Sublicensed IP from Competitive Infringement and

BGN exercises its rights under the BGN License Agreement to prosecute, prevent, or terminate such Competitive Infringement, any amount received by BioLineRx in connection therewith, whether by settlement or otherwise, [**].

(c) Claimed Infringement. If a Party becomes aware of any claim that the Development, Manufacture, or Commercialization of Products for use in the Field in the Territory infringes Patent Rights or any other intellectual property rights of any Third Party, such Party shall promptly notify the other Party. In any such instance, Ikaria shall have the exclusive right to settle such claim.

(d) Patent Invalidity Claim. If a Third Party at any time asserts a claim that any BioLineRx Patent Rights is invalid or otherwise unenforceable (an "Invalidity Claim"), whether (i) as a defense in an infringement action brought by Ikaria or BioLineRx pursuant to subsection (b) above, or (ii) in an action brought against Ikaria or BioLineRx referred to in subsection (c) above, or (iii) otherwise, the Parties shall cooperate with each other in preparing and formulating a response to such Invalidity Claim. Neither Party shall settle or compromise any Invalidity Claim without the consent of the other Party, which consent shall not be unreasonably withheld, conditioned or delayed.

(e) Conduct of Certain Actions; Costs. Ikaria shall have the sole and exclusive right to select counsel for any suit initiated by it referenced in subsection (b)(i) above or against it referenced in subsection (c) above, and BioLineRx shall have the sole and exclusive right to select counsel for any suit initiated by it referenced in subsection (b)(ii) above. If required under applicable law in order for a Party (the "Lead Party") to initiate or maintain such suit, the other Party shall join as a party to the suit. Such other Party shall offer reasonable assistance to the Lead Party in connection therewith at no charge to the Lead Party except for reimbursement of such other Party's reasonable out-of-pocket expenses incurred in rendering such assistance. The Lead Party shall assume and pay all of its own out-of-pocket costs incurred in connection with any litigation or proceedings referenced in the first sentence of this subsection (e), including the fees and expenses of the counsel selected by it. Subject to applicable law, the other Party shall have the right to participate and be represented in any such suit by its own counsel at its own expense.

(f) Coordination with BGN. With respect to any suit to protect Sublicensed IP from infringement for which Ikaria is the Lead Party, notwithstanding anything to the contrary in this Section 5.3:

(i) if required under applicable law in order for Ikaria to initiate or maintain such suit, BioLineRx shall (A) exercise its rights under the BGN License Agreement to cause BGN to join as a party to such suit, (B) exercise its rights under the BGN License Agreement to obtain BGN's approval of counsel selected by Ikaria to represent Ikaria and BGN in such suit, and (C) [**];

(ii) Ikaria shall not compromise or settle such suit without the prior written consent of BGN, which consent BioLineRx shall exercise its rights under the BGN License Agreement to obtain; and

(iii) any recovery obtained by Ikaria as a result of such suit, by settlement or otherwise, shall be applied in the following order of priority: (A) first, each Party shall be reimbursed, on a pro rata basis, for all costs incurred by such Party in connection with such suit (for clarity, BioLineRx shall be reimbursed for any costs of BGN paid by BioLineRx in accordance with clause (i)(C) above); (B) second, [**] percent ([**]%) of any remainder shall paid to BioLineRx for remittance to BGN as provided in Section 10.1.2 of the BGN License Agreement ; and (C) third, the remaining [**] percent ([**]%) shall be retained by Ikaria; [**].

Article VI
Confidentiality; Non-Solicitation; Standstill

Section 6.1 Confidential Information. Each Party agrees that all Confidential Information disclosed to it or its Affiliates by the other Party (a) shall not be used by the receiving Party or its Affiliates except to fulfill its obligations or exercise its rights under this Agreement, (b) shall be maintained in confidence by the receiving Party and its Affiliates, and (c) shall not be disclosed by the receiving Party or its Affiliates to any Third Party who is not a consultant of, or an advisor to, the receiving Party or its Affiliates without the prior written consent of the disclosing Party, which consent the disclosing Party may withhold in its sole discretion. Notwithstanding the foregoing, either Party may disclose Confidential Information of the other Party if such Party is required to make such disclosure by applicable law, regulation or legal process, including by Israeli securities laws, the rules or regulations of the United States Securities and Exchange Commission (the "SEC") or any similar regulatory agency in a country other than the United States or of any stock exchange, including the Tel Aviv Stock Exchange, in which event such Party shall provide prior notice of such intended disclosure to such other Party, if possible under the circumstances, and shall disclose only such Confidential Information of the other Party as is required to be disclosed. If this Agreement shall be included in any report, statement or other document filed by either Party or an Affiliate of either Party pursuant to the preceding sentence, such Party shall use, or shall cause its Affiliate, as the case may be, to use, reasonable efforts to obtain confidential treatment from the SEC, similar regulatory agency or stock exchange of any financial information or other information of a competitive or confidential nature, and shall include in such confidentiality request such provisions of this Agreement as may be reasonably requested by the other Party.

Section 6.2 Disclosures to Employees, Consultants, Advisors, Etc. Each Party agrees that it and its Affiliates shall provide Confidential Information received from the other Party only to the receiving Party's respective employees, consultants, advisors, Licensees and potential Licensees, and to the employees, consultants and advisors of the receiving Party's Affiliates, who have a need to know such Confidential Information to assist the receiving Party in fulfilling its obligations under this Agreement and only under conditions of confidentiality and non-use at least as stringent as the conditions imposed by this Agreement, provided that BioLineRx and Ikaria shall each remain responsible for any failure by its and its Affiliates' respective employees, consultants, advisors, Licensees and potential Licensees to treat such information and materials as required under Section 6.1. For clarity, (a) Ikaria is permitted to disclose Confidential Information to actual or potential Licensees, acquirors or financing sources; and (b) BioLineRx is permitted to disclose this Agreement and the Development Plan to BGN, solely to

the extent required under the BGN License Agreement; provided that any such disclosure subjects the receiving Third Party to conditions of confidentiality and non-use at least as stringent as the conditions imposed by this Agreement.

Section 6.3 Non-Solicitation. During the term of this Agreement and continuing for [**] months after the termination of this Agreement, neither Party shall directly or indirectly, for its own account or for the account of others, urge, induce, entice, or in any manner whatsoever solicit any employee directly involved in the activities conducted pursuant to this Agreement to leave the employment of the other Party or any of its Affiliates. For purposes of the foregoing, "urge", "induce", "entice" or "solicit" shall not be deemed to mean: (a) circumstances where an employee of a Party initiates contact with the other Party or any of its Affiliates with regard to possible employment; or (b) general solicitations of employment not specifically targeted at employees of a Party or any of its Affiliates, including responses to general advertisements.

Section 6.4 Standstill. Neither Ikaria nor any of its Affiliates shall directly or indirectly, for its own account or for the account of others, acquire more than [**] of the equity or debt securities of BioLineRx, or urge, induce, entice or solicit any Third Party to acquire the equity or debt securities of BioLineRx, in either case without the consent of BioLineRx, which may be withheld in its sole discretion. The obligations of Ikaria under this Section 6.4 shall terminate in the

event that (a) any Third Party initiates a tender or exchange offer, or otherwise publicly proposes or agrees to acquire, a majority of the equity or debt securities of BioLineRx (provided that the restrictions set forth in this Section 6.4 shall be reinstated in the event that such tender or exchange offer, or proposal, is terminated or withdrawn), (b) it is publicly disclosed that voting securities representing at least [**] of the total voting power of BioLineRx have been acquired by any one or more Third Parties, (c) BioLineRx publicly announces that it intends to seek a Third Party acquirer (provided that the restrictions set forth in this Section 6.4 shall be reinstated in the event that BioLineRx publicly announces that it no longer is seeking a Third Party acquirer and so notifies Ikaria in writing), (d) BioLineRx enters into any agreement to merge with, or sell or dispose of [**] or more of its assets or securities, or (e) this Agreement is terminated pursuant to Article VIII. BioLineRx shall provide Ikaria with prompt written notice of the occurrence of any of the foregoing events to the extent permitted under applicable law. For clarity, the acquisition by any employee benefit plan of Ikaria or its Affiliates in any diversified index, mutual or pension fund, which fund in turn holds BioLineRx securities, shall not be deemed a breach of this Section 6.4.

Section 6.5 Term. All obligations of confidentiality imposed under this Article VI shall survive until the date that is [**] years after the expiration or termination of this Agreement.

Section 6.6 Publicity. During the term of this Agreement, the content of any press release or public announcement relating to this Agreement or a Product shall be mutually approved by the Parties, except that (a) a Party may issue such press release or public announcement if the contents of such press release or public announcement have previously been made public other than through a breach of this Agreement by the issuing Party, (b) a Party may issue such a press release or public announcement if it is advised by counsel that such press release or public announcement is required by applicable law, regulation or legal process, including by Israeli securities laws, the rules or regulations of the SEC or any similar regulatory agency in a country

other than the United States or of any stock exchange, including the Tel Aviv Stock Exchange, and (c) Ikaria shall remain free to issue press releases and public announcements regarding the Development, Manufacturing, Commercialization and use of Products in the Field, provided that Ikaria shall provide BioLineRx with advance notice of at least [**] days prior to public disclosure of such releases and announcements or such shorter period as required to comply with any applicable law. In addition, BioLineRx shall reasonably implement any changes that Ikaria may recommend with respect to any filing to be made in accordance with the rules or regulations of the SEC or any similar regulatory agency in a country other than the United States or of any stock exchange, including the Tel Aviv Stock Exchange; provided that such Ikaria shall only have the right to comment upon portions of such filings that directly related to Ikaria or this Agreement. Nothing in the foregoing shall require BioLineRx to implement any change that Ikaria may recommend that is not consistent with the rules or regulations of the Israel Securities Authority, Tel Aviv Stock Exchange, the rules or regulations of the SEC, or any similar regulatory agency in a country other than the United States or Israel, as advised in writing by BioLineRx's legal counsel. BioLineRx's legal counsel will provide Ikaria confirmation of such advise.

Section 6.7 Publications. The results of the Development Program may be published by a Party as part of a scientific presentation or publication only after scientific review by and approval of the Joint Development Committee unless the other Party, acting reasonably, disapproves of the presentation or publication in writing within [**] days after receipt of the presentation or publication. Either Party may require that such Party's Confidential Information be redacted from such presentation or publication and may reasonably require that other information also be redacted. In addition, at the request of either Party, the date of submission for presentation or publication shall be delayed for a period of time sufficiently long to permit a Party to seek appropriate patent protection. Other than as provided for herein, BioLineRx shall not make any publication regarding any Product or containing any Confidential Information of Ikaria without the prior written consent of Ikaria. Notwithstanding the foregoing, to the extent necessary or appropriate as determined in Ikaria's discretion, Ikaria may disclose information otherwise covered by this Section 6.7 in documents filed with the SEC.

Article VII
Representations and Warranties

Section 7.1 Representations of Authority. BioLineRx and Ikaria each represents and warrants to the other Party that, except for the consent of the OCS, it has full corporate right, power and authority to enter into this Agreement and to perform its respective obligations under this Agreement and that it has the right to grant to the other Party the rights and licenses granted pursuant to this Agreement.

Section 7.2 Consents. BioLineRx and Ikaria each represents and warrants to the other Party that, except for the consent of the OCS, all necessary consents, approvals and authorizations of all government authorities and other Persons required to be obtained by it as of the date hereof in connection with the execution, delivery and performance of this Agreement have been obtained.

Section 7.3 No Conflict. BioLineRx and Ikaria each represents and warrants to the other Party that, notwithstanding anything to the contrary in this Agreement, except for the consent of the OCS, the execution and delivery of this Agreement, the performance of such Party's obligations in the conduct of the collaboration and the licenses and rights to be granted pursuant to this Agreement (a) do not conflict with or violate any requirement of applicable laws or regulations existing as of the date hereof and (b) do not conflict with, violate, breach or constitute a default under any contractual obligations of such Party or any of its Affiliates existing as of the date hereof.

Section 7.4 Enforceability. BioLineRx and Ikaria each represents and warrants to the other Party that this Agreement is a legal and valid obligation binding upon it and is enforceable against it in accordance with its terms.

Section 7.5 Additional BioLineRx Representations. BioLineRx represents and warrants to Ikaria that:

(a) BioLineRx has the right to grant the licenses granted to Ikaria on the terms set forth in this Agreement;

(b) BioLineRx is not engaged with any Third Party in any Development efforts directed to Products in the Field in the Territory other than with respect to the On-Going Phase I/II Trial, the Other On-Going Trials or the Existing Product Agreements;

(c) BioLineRx has provided Ikaria with true and complete copies of each of the Existing Product Agreements, each of which is in full force and effect in accordance with its terms as of the date hereof, and has obtained all consents necessary for the assignment to Ikaria of each of the Existing Product Agreements hereunder, and, following such assignment, Ikaria shall have the legal right to exercise all rights of BioLineRx that existed thereunder immediately prior to such assignment;

(d) to BioLineRx's Knowledge, the BioLineRx Patent Rights listed in Exhibit B are valid and enforceable and constitute all of the Patent Rights necessary or useful for Ikaria to fully exercise and enforce its rights hereunder;

(e) to BioLineRx's Knowledge, the BioLineRx Patent Rights are not being infringed and the BioLineRx Know-How is not being misappropriated by any Third Party;

(f) BioLineRx owns the entire right, title and interest in and to the BioLineRx Intellectual Property (other than the Sublicensed IP) free and clear of any liens, charges, claims and encumbrances, and no other Person has any claim of ownership or right to obtain compensation with respect to such BioLineRx Intellectual Property;

(g) to BioLineRx's Knowledge, the Products developed in the Development Program and the Development, Manufacture and Commercialization of such Products will not infringe or misappropriate any intellectual property rights not licensed to Ikaria hereunder; and

(h) BioLineRx has not received and has no Knowledge of any claim or demand of any Person pertaining to, or any proceeding which is pending or threatened that asserts, the

invalidity, misuse or unenforceability of the BioLineRx Patent Rights or that challenges BioLineRx's ownership of the BioLineRx Intellectual Property or that makes any adverse claim with respect thereto, and, to the Knowledge of BioLineRx, there is no basis for any such claim, demand or proceeding.

Section 7.6 BGN License Agreement. BioLineRx represents, warrants and covenants to Ikaria that:

(a) BioLineRx has provided Ikaria with a true and complete copy of the BGN License Agreement, which is in full force and effect in accordance with its terms as of the date hereof;

(b) BioLineRx shall obtain and provide to Ikaria within ten (10) days of execution of this Agreement a written statement from BGN certifying that the terms of this Agreement are consistent with those of the BGN License Agreement, including in the context of Section 13.4.1(c) thereof;

(c) BioLineRx has (i) achieved by its designated performance date each Milestone (as that term is defined in the BGN License Agreement) having a designated performance date on or before the date hereof, or obtained a waiver in respect thereof, and (ii) neither (A) committed any material breach of the its obligations under the BGN License Agreement nor (B) received any notice from BGN of any alleged material breach thereof by BioLineRx or of any Failure (as that term is defined therein);

(d) BioLineRx shall upon receipt by BioLineRx promptly provide Ikaria with a copy of any notice from BGN described in the foregoing clause (c)(ii)(B);

(e) BioLineRx shall not terminate, amend, supplement or otherwise modify the BGN License Agreement without Ikaria's prior written consent;

(f) the rights and obligations of BioLine Jerusalem L.P. under the BGN License Agreement have been assigned and delegated, or otherwise transferred, to BioLineRx;

(g) as between BioLineRx and Ikaria, BioLineRx shall be responsible for any and all payments to be made under the BGN License Agreement;

(h) in the event of any termination of the BGN License Agreement, BioLineRx shall, at Ikaria's request, provide all reasonable assistance to Ikaria in Ikaria's efforts to obtain from BGN an exclusive license to the Sublicensed IP, including through enforcement of the provisions of Sections 5.2.3 and 13.4.1(c) of the BGN License Agreement.

Section 7.7 Employee, Consultant and Advisor Legal Obligations. BioLineRx and Ikaria each represents and warrants that each of its and its Affiliates' employees, consultants and advisors who is or will be involved in performing any obligations hereunder has executed or will have executed an agreement or have an existing obligation under law requiring assignment to such Party of all intellectual property made during the course of and as the result of his, her or its association with such Party or such Affiliate, and obligating such employee, consultant or advisor to maintain the confidentiality of Confidential Information to the extent required under

Article VI. BioLineRx and Ikaria each represents and warrants that, to its Knowledge, none of its or its Affiliates' employees, consultants or advisors who is or will be involved in performing any obligations hereunder is, as a result of the nature of such obligations to be performed by the Parties, in violation of any covenant in any contract relating to non-disclosure of proprietary information, non-competition or non-solicitation.

Section 7.8 Accuracy of Representations and Warranties on Effective Date. The representations and warranties of each of the Parties set forth in the preceding sections of this Article VII remain true and accurate on and as of the Effective Date. Each Party shall promptly following receipt of acceptable consent from the OCS deliver to the other Party a certificate to such effect executed by its Chief Executive Officer.

Section 7.9 No Warranties. EXCEPT AS OTHERWISE EXPRESSLY SET FORTH IN THIS AGREEMENT, THE PARTIES MAKE NO REPRESENTATIONS AND EXTEND NO WARRANTIES OF ANY KIND, EITHER EXPRESS OR IMPLIED, INCLUDING THAT ANY PRODUCTS WILL BE ECONOMICALLY OR TECHNICALLY UTILIZABLE, THAT ANY SALES OF ANY PRODUCTS WILL OCCUR, THAT THE DEVELOPMENT PROGRAM ACTIVITIES WILL BE COMPLETED IN THE EXPECTED TIMEFRAME, OR THAT ANY PRODUCT WILL BE FREE OF ANY THIRD PARTY RIGHTS.

Article VIII
Term and Termination

Section 8.1 Term. The term of this Agreement shall begin on the Effective Date, may be terminated as set forth in this Article VIII, and shall expire on a Product-by-Product and country-by-country basis upon the date of expiration of the Royalty Term for such Product in such country, and shall expire in its entirety upon the last-to-expire Royalty Term, unless earlier terminated as set forth in this Article VIII.

Section 8.2 Termination for Material Breach. Upon any breach of a material provision of this Agreement by a Party (the "Breaching Party"), the other Party (the "Non-Breaching Party") may terminate this Agreement by providing ninety (90) days written notice to the Breaching Party specifying the material breach. The termination shall become effective at the end of the notice period unless the Breaching Party cures such breach during such notice period. Ikaria may terminate this Agreement pursuant to this Section 8.2 immediately upon any termination of the BGN License Agreement.

Section 8.3 Development-Related Termination. Ikaria shall have the right to terminate this Agreement upon sixty (60) days prior written notice, if Ikaria at any time determines, in its sole judgment, that the results of the Development Program do not warrant further Development of Products.

Section 8.4 Effect of Certain Terminations and Expiration.

(a) If this Agreement is terminated by Ikaria under Section 8.2:

(i) The licenses granted by BioLineRx to Ikaria under Section 2.1 and, notwithstanding any other provision in this Agreement to the contrary, Ikaria's obligations under Section 4.2, shall survive;

(ii) Section 2.2 shall survive until Ikaria is no longer obligated to pay royalties to BioLineRx under Section 4.2; and

(iii) Section 5.1 and Section 5.3 shall survive.

(b) If this Agreement is terminated by either BioLineRx under Section 8.2, or by Ikaria under Section 8.3, the licenses granted under Section 2.1 shall terminate as of the effective date of such termination; provided, however, that Ikaria, its Affiliates, and its Licensees shall be afforded a commercially reasonable period of time (but no less than [**] months) to sell off any then existing or in process stocks of the Products, subject to the terms and conditions of this Agreement, including the payment of royalties thereon.

(c) Upon any termination or expiration of this Agreement, each Party shall return to the other Party any tangible property owned by the other Party, including any books and records and Confidential Information, in accordance with the reasonable instructions given by the other Party, with any shipping costs to be borne by the other Party, provided, however, that a Party may retain a copy of any regulatory records it is required to maintain in accordance with applicable law.

Section 8.5 Survival. In the event of any expiration or termination of this Agreement, (a) all financial obligations under Article IV and Article V owed as of the effective date of such expiration or termination shall remain in effect, including such obligations that have accrued, but have not been invoiced, as of such effective date, and (b) the obligations set forth in Section 5.1, Article VI, Article IX and Article X, and all other terms, provisions, representations, rights and obligations contained in this Agreement that by their express terms survive expiration or termination of this Agreement (including Section 8.4 and this Section 8.5), shall survive and all other terms, provisions, representations, rights and obligations contained in this Agreement shall terminate.

Section 8.6 Termination Prior to Effective Date. Notwithstanding anything to the contrary in this Article VIII, Ikaria may terminate this Agreement prior to the Effective Date, with no liability to BioLineRx, if the OCS does not consent to the Agreement in a form reasonably satisfactory to both Parties within forty-five (45) days after the execution of this Agreement. The provisions of Article X (except for Section 10.1(a)) and this Section 8.6 shall survive such termination, and all other terms, provisions, representations, rights and obligations contained in this Agreement shall terminate.

Article IX
Dispute Resolution

Section 9.1 Negotiation. Any controversy, claim or dispute arising out of or relating to this Agreement shall be settled, if possible, through good faith negotiations between the Parties.

Section 9.2 Escalation. If the Parties are unable to settle any dispute after good faith negotiations pursuant to Section 9.1 after [**] days, such dispute (except for any matter that by its express terms shall be resolved as provided in this Agreement, including any matter arising under Section 3.2 or Section 3.6) shall be referred to the Executive Officers to be resolved by negotiation in good faith as soon as is practicable but in no event later than [**] days after referral.

Section 9.3 Mediation. Solely with respect to a dispute as to whether Ikaria has breached its obligations to use Commercially Reasonable Efforts as set forth in Section 3.8, if the Executive Officers are unable to settle such dispute after good faith negotiations pursuant to Section 9.2 within [**] days after referral to the Executive Officers, the Parties shall, within [**] days thereof, engage a mutually agreeable Third Party mediator on a non-binding basis to assist the Parties in determining whether such a breach has occurred. The Parties agree that they will participate in good faith in an effort to resolve the dispute in an informal, inexpensive and expeditious manner and that any mediator selected shall agree to render any judgments in a timely manner, but no later than [**] days after the mediator is selected. All expenses of the mediator will be shared equally by the Parties.

Section 9.4 Litigation. If the Executive Officers are unable to settle any dispute after good faith negotiations pursuant to Section 9.2 (other than a dispute as to whether Ikaria has breached its obligations to use Commercially Reasonable Efforts as set forth in Section 3.8) within [**] days after referral, or if the Parties continue to dispute whether Ikaria has breached its obligations to use Commercially Reasonable Efforts as set forth in Section 3.8 following mediation pursuant to Section 9.3, then either Party may seek resolution of the dispute (except for any matter that by its express terms shall be resolved as provided in this Agreement, including any matter arising under Section 3.2 or Section 3.6) through remedies available at law or in equity from any court of competent jurisdiction as set forth in Section 10.3.

Section 9.5 Equitable Relief. Each Party acknowledges and agrees that the other Party would be damaged irreparably if any of the provisions of Article II, Article V and Article VI are not performed in accordance with their specific terms or otherwise are breached. Accordingly, each Party agrees that the

other Party shall be entitled to an injunction or other equitable relief to prevent breaches of such provisions, to preserve status quo, and to enforce specifically such provisions in any action instituted in any court having jurisdiction over the Parties and the matter, in addition to any other remedy to which it may be entitled, at law or in equity.

Article X
Miscellaneous Provisions

Section 10.1 Indemnification.

(a) By Ikaria. Ikaria agrees to defend BioLineRx, its Affiliates and their respective directors, officers, employees and agents at Ikaria's cost and expense, and shall indemnify and hold harmless BioLineRx and its Affiliates and their respective directors, officers, employees and agents from and against any liabilities, losses, costs, damages, fees or expenses (collectively, "Losses") arising out of any Third Party claim to the extent relating to (i) any breach by Ikaria of any of its representations, warranties or obligations pursuant to this Agreement, or (ii) personal

injury, property damage, product liability or other damage resulting from the Development, Manufacture, use or Commercialization of a Product by Ikaria or its Affiliates or Licensees, excluding any claim for which BioLineRx indemnifies Ikaria under subsection (b) below.

(b) By BioLineRx. BioLineRx agrees to defend Ikaria, its Affiliates and their respective directors, officers, employees and agents at BioLineRx's cost and expense, and shall indemnify and hold harmless Ikaria and its Affiliates and their respective directors, officers, employees and agents from and against any Losses arising out of any Third Party claim to the extent relating to (i) any breach by BioLineRx of any of its representations, warranties or obligations pursuant to this Agreement, (ii) personal injury, property damage or other damage resulting from the conduct of the On-Going Phase I/II Trial or the Other On-Going Trials by or on behalf of BioLineRx or its Affiliates, (iii) the BGN Agreement, or (iv) any allegation that the practice of the BioLineRx Intellectual Property rights in the Development Program infringes or misappropriates any Third Party intellectual property rights, to the extent BioLineRx had Knowledge that such practice would infringe or misappropriate such Third Party intellectual property rights on or before the Effective Date.

(c) Claims for Indemnification. A Person entitled to indemnification under this Section 10.1 (an "Indemnified Party") shall give prompt written notification to the Party from whom indemnification is sought (the "Indemnifying Party") of the commencement of any action, suit or proceeding relating to a Third Party claim for which indemnification may be sought or, if earlier, upon the assertion of any such claim by a Third Party (it being understood and agreed, however, that the failure by an Indemnified Party to give notice of a Third Party claim as provided in this Section 10.1(c) shall not relieve the Indemnifying Party of its indemnification obligation under this Agreement except and only to the extent that such Indemnifying Party is actually damaged as a result of such failure to give notice). Within [**] days after delivery of such notification, the Indemnifying Party may, upon written notice thereof to the Indemnified Party, assume control of the defense of such action, suit, proceeding or claim with counsel reasonably satisfactory to the Indemnified Party. If the Indemnifying Party does not assume control of such defense, the Indemnified Party shall control such defense. The Party not controlling such defense may participate therein at its own expense. The Party controlling such defense shall keep the other Party advised of the status of such action, suit, proceeding or claim and the defense thereof and shall consider recommendations made by the other Party with respect thereto. The Indemnified Party shall not agree to any settlement of such action, suit, proceeding or claim without the prior written consent of the Indemnifying Party, which consent the Indemnifying Party shall not unreasonably withhold, condition or delay. The Indemnifying Party shall not agree, without the prior written consent of the Indemnified Party, which consent the Indemnified Party shall not unreasonably withhold, condition or delay, to any settlement of such action, suit, proceeding or claim or consent to any judgment in respect thereof that does not include a complete and unconditional release of the Indemnified Party from all liability with respect thereto or that imposes any liability or obligation on the Indemnified Party.

Section 10.2 Governing Law. This Agreement shall be construed and the respective rights of the Parties determined in accordance with the laws of the State of New York, USA (other than any principle of conflict or choice of laws that would cause the application of the laws of any other jurisdiction).

Section 10.3 Submission to Jurisdiction. Each Party (a) submits to the jurisdiction of any state or federal court sitting in the State of New York, USA in any action or proceeding arising out of or relating to this Agreement, (b) agrees that all claims in respect of such action or proceeding may be heard and determined in any such court, (c) waives any claim of inconvenient forum or other challenge to venue in such court, and (d) agrees not to bring any action or proceeding arising out of or relating to this Agreement in any other court, unless the state or federal courts sitting in the State of New York decline to exercise jurisdiction over any such action or proceeding or if those courts lack proper jurisdiction, then any action or proceeding arising out of or relating to this Agreement may be brought in any other U.S. court of competent jurisdiction. Each Party agrees to accept service of any summons, complaint or other initial pleading made in the manner provided for the giving of notices in Section 10.6, provided that nothing in this Section 10.3 shall affect the right of either Party to serve such summons, complaint or other initial pleading in any other manner permitted by law.

Section 10.4 Assignment. Ikaria may assign this Agreement or any right hereunder, or delegate any obligation hereunder, in its sole discretion, to (a) any Affiliate of Ikaria or (b) any entity acquiring all or substantially all of the assets of Ikaria Holdings, Inc. and its Affiliates. All other assignments by Ikaria, including (i) to any entity acquiring all or substantially all of the assets of Ikaria to which this Agreement relates or (ii) to any entity with which or into which Ikaria may consolidate or merge, are subject to BioLineRx's prior approval, which approval shall not be unreasonably withheld, conditioned or delayed. BioLineRx may assign its right to receive payments hereunder to a Third Party, in its sole discretion, but BioLineRx shall not otherwise be permitted to assign this Agreement, in whole or in part, without the prior written consent of Ikaria, which approval shall not be unreasonably withheld, conditioned or delayed. Any assignments in contravention of this Section 10.4 shall be null and void.

Section 10.5 Entire Agreement; Amendments. This Agreement constitutes the entire agreement between the Parties with respect to the subject matter hereof, and supersedes all previous arrangements between the Parties with respect to the subject matter hereof, whether written or oral, except for that certain Mutual Non Disclosure Agreement between the Parties dated February 25, 2009. Without limiting the generality of the foregoing, this Agreement hereby supersedes and replaces in its entirety the License and Commercialization Agreement by and among the parties dated as of July 5th, 2009. To the extent that any provision of this Agreement conflicts with any provisions of such Mutual Non Disclosure Agreement, the provision of this Agreement shall control. Except as set forth in Section 2.1(iv), any amendment or modification to this Agreement shall be made in writing signed by both Parties.

Section 10.6 Notices.

Notices to Ikaria shall be addressed to:

Ikaria Development Subsidiary One LLC
6 State Route 173
Clinton, NJ 08809, USA
Attention: Chief Executive Officer

with copy to:

Ikaria Holdings, Inc.
6 State Route 173
Clinton, NJ 08809, USA
Attention: General Counsel

Notices to BioLineRx Ltd. shall be addressed to:

BioLineRx Ltd.
19 Hartum Street
P.O. Box 45158
Jerusalem 91450, Israel
Attention: Chief Executive Officer

with copy to:

Arent Fox LLP
1050 Connecticut Avenue
Washington, DC 20036, USA
Attention: John Dwyer, Esq.

Notices to BioLine Innovations Jerusalem L.P. shall be addressed to:

BioLine Innovations Jerusalem L.P.
19 Hartum Street
P.O. Box 45158
Jerusalem 91450, Israel
Attention: Chief Executive Officer

with copy to:

Arent Fox LLP
1050 Connecticut Avenue
Washington, DC 20036, USA
Attention: John Dwyer, Esq.

Any Party may change its address by giving notice to the other Party in the manner herein provided. Any notice required or provided for by the terms of this Agreement shall be in writing and shall be (a) sent by registered or certified mail, return receipt requested, postage prepaid, (b) sent via a reputable international courier service, (c) sent by facsimile transmission, or (d) personally delivered, in each case properly addressed in accordance with the paragraph above. The effective date of notice shall be the actual date of receipt by the Party receiving the same.

Section 10.7 Force Majeure. No failure or omission by a Party in the performance of any obligation of this Agreement shall be deemed a breach of this Agreement or create any liability if the same shall arise from any cause or causes beyond the control of such Party, including the following: acts of God; fire; storm; flood; earthquake; accident; war; rebellion; insurrection; riot; and invasion (each such event, a "Force Majeure Event") and provided that such Party cures such

failure or omission resulting from one of the above causes as soon as is practicable after the occurrence of one or more of the above-mentioned causes.

Section 10.8 Independent Contractors. It is understood and agreed that the relationship between the Parties hereunder is that of independent contractors and that nothing in this Agreement shall be construed as authorization for either BioLineRx or Ikaria to act as agent for the other.

Section 10.9 Limitations of Liability. NEITHER PARTY SHALL BE LIABLE FOR ANY INDIRECT, INCIDENTAL, CONSEQUENTIAL, SPECIAL, EXEMPLARY OR PUNITIVE DAMAGES ARISING OUT OF THIS AGREEMENT OR THE EXERCISE OF ITS RIGHTS HEREUNDER, OR FOR LOST PROFITS ARISING FROM OR RELATING TO ANY BREACH OF THIS AGREEMENT, REGARDLESS OF ANY NOTICE OF SUCH DAMAGES. NOTHING IN THIS SECTION 10.9 IS INTENDED TO LIMIT OR RESTRICT (A) THE INDEMNIFICATION RIGHTS OR OBLIGATIONS OF EITHER PARTY WITH RESPECT TO THIRD PARTY CLAIMS; (B) ANY LOSSES, INCLUDING LOST PROFITS, ARISING FROM ANY (I) BREACH OF A PARTY'S OBLIGATIONS WITH RESPECT TO THE OTHER PARTY'S CONFIDENTIAL INFORMATION, (II) BREACH BY BIOLINERX OF THE EXCLUSIVE RIGHTS GRANTED IN SECTION 2.1 OR THE COVENANT CONTAINED IN SECTION 2.2, OR (III) USE OF ANY PATENT RIGHTS OR KNOW-HOW LICENSED HEREUNDER BEYOND THE SCOPE OF SUCH LICENSE; OR (C) ANY LOSSES ARISING AS A RESULT OF A PARTY'S FRAUD, GROSS NEGLIGENCE OR WILLFUL MISCONDUCT.

Section 10.10 No Implied Waivers; Rights Cumulative. No failure on the part of BioLineRx or Ikaria to exercise, and no delay in exercising, any right, power, remedy or privilege under this Agreement, or provided by statute or at law or in equity or otherwise, shall impair, prejudice or constitute a waiver of any such right, power, remedy or privilege or be construed as a waiver of any breach of this Agreement or as an acquiescence thereto, nor shall any single or partial exercise of any such right, power, remedy or privilege preclude any further or other exercise thereof or the exercise of any other right, power, remedy or privilege.

Section 10.11 Severability. If, under applicable law or regulation, any provision of this Agreement is invalid, incomplete or unenforceable, or otherwise directly or indirectly affects the validity of any other material provision(s) of this Agreement (such invalid, incomplete or unenforceable provision, a "Severed Clause"), this Agreement shall endure except for the Severed Clause. The Parties shall consult one another and use reasonable efforts to agree upon a valid, complete and enforceable provision that is a reasonable substitute for the Severed Clause in view of the intent of this Agreement.

Section 10.12 Execution in Counterparts; Facsimile Signatures. This Agreement may be executed in counterparts, each of which, when so executed and delivered, shall be deemed to be an original, and all of which, taken together, shall constitute one and the same instrument even if both Parties have not executed the same counterpart. Signatures provided by facsimile transmission shall be deemed to be original signatures.

REMAINDER OF PAGE LEFT EMPTY; NEXT PAGE IS THE SIGNATURE PAGE

IN WITNESS WHEREOF, the Parties have executed this License and Commercialization Agreement as of the Effective Date.

IKARIA DEVELOPMENT SUBSIDIARY ONE LLC

By: /s/ Matthew M. Bennett

Name: Matthew M. Bennett

Title: Senior Vice President

BIOLINERX LTD.

By: /s/ Morris Laster M.D.

Name: Morris Laster M.D.

Title: CEO

BIOLINE INNOVATIONS JERUSALEM L.P.
by its General Partner, BioLine Innovations Jerusalem, Ltd.

By: /s/ Morris Laster M.D.

Name: Morris Laster M.D.

Title: Director

SCHEDULE 1.30

PROTOCOL FOR ON-GOING PHASE I/II TRIAL

[*PROTOCOL IMMEDIATELY FOLLOWS*]

CLINICAL STUDY

Protocol No. BL-1040.01
Version 5.00 Incorporating Amendments 1, 2, 3 and 4
Safety and Feasibility
Final

A Phase I, multi-center, open label study designed to assess the safety and feasibility of the injectable BL-1040 implant to

provide scaffolding to infarcted myocardial tissue

BioLine Innovations Jerusalem

Confidentiality Statement

This document contains information that is the property of BioLine Innovations Jerusalem and therefore is provided to you in confidence for review by you, your staff, an applicable ethics committee/institutional review board and regulatory authorities. It is understood that this information will not be disclosed to others without written approval from BioLine innovations Jerusalem, except to the extent necessary to obtain informed consent from those persons to whom BL-1040 may be administered.

Annotated Protocol incorporating Amendment 1, Amendment 2, Amendment 3, and Amendment 4
01 December 2008

PROTOCOL NUMBER:	BL-1040.01 Safety and Feasibility
DATE OF PROTOCOL:	Final, **01 December** 2008 Version 2 incorporating Amendment 1, 07 August 2007 Version 3 incorporating Amendment 2, 03 December 2007 Version 4 incorporating Amendment 3, 17 April 2008 **Version 5 incorporating Amendment 4, 27 November 2008**
PROTOCOL TITLE:	A Phase I, multi-center, open label study designed to assess the safety and feasibility of the injectable BL-1040 implant to provide scaffolding to infarcted myocardial tissue
SPONSOR:	BioLine Innovations Jerusalem

Responsible study personnel:

Name:	Prof. Moshe Phillip, MD, Vice-President of Medical Affairs, Sr. Clinical Advisor
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Phone:	+972-2-548-9100
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Address:	Head, Neufeld Cardiac Research Institute. Tel-Aviv University Sheba Medical Center Tel-Hashomer, Israel 52621
Phone:	+972-3-534-8685, 972-3-530-2614
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e-mail:	leorj@post.tau.ac.il

CRO:	Venn Life Sciences AG
Address:	Elisabethenstrasse 23/3, CH- 4051 Basel
Phone:	+41 61 201 11 00Fax: +41 61 273 42 50

Authorized representative:	Voisin Consulting
Address:	3, rue des Longs Prés, 92100 Boulogne, France
Phone:	+33-1-41-31-8300
Fax:	+33-1 41-31-8309
e-mail:	voisin@voisinconsulting.com

Medical Monitor, US (ISMB support only)

Name:	Sanjay Machado, MD
Address:	Venn Life Sciences Group 7355 TransCanada Hwy Suite 200 Saint-Laurent, Quebec, Canada H4T 1T3
Phone:	+1 514.315.2992 ext 117
Fax:	+1 514.315.0995
e-mail:	sanjay.machado@vlsworldwide.com

Medical Monitor, Europe

Name:	Andrea Kempf-Mueller, MD
Address:	Venn Life Sciences AG Elisabethenstrasse 23/3, 4051 Basel, Switzerland
Phone:	+41 61 201 11 83
Fax:	+41 61 273 42 50
e-mail:	andrea.kempf-mueller@vlsworldwide.com

Investigator's Signature Page

INVESTIGATOR:

Name:

Address:

Phone:
Fax:
e-mail:

I, the undersigned, have reviewed this Protocol, including Appendices, and I will conduct the clinical study as described and will adhere to GCP/ICH and all the ethical and regulatory considerations stated. I have read and understood the contents of the Investigator Brochure.

Date/Place _____ Signature _____
(Name of Investigator)

Sponsor Signature Page

Sponsor:	**BioLine Innovations Jerusalem**
Address:	**19 Hartum St., POB 45158** **Jerusalem, Israel 91450**
Phone:	**+972-2-548-9100**
Fax:	**+972-2-548-9101**
e-mail:	**Info@biolineRx.com**

I have read the protocol and confirm that the protocol follows the current GCP guidelines.

Date/Place 27 Jan 2009 Signature /s/ Moshe Phillip
(Prof Moshe Phillip, VP of Medical Affairs, Sr. Clinical Advisor)

Date/Place 27 Jan 2009 Signature /s/ Shmuel Tuvia
(Shmuel Tuvia, PhD, Project Manager)

Date/Place 27 Jan 2009 Signature /s/ Moti Gal
(Moti Gal, Clinical Operations Manager)

Medical Advisor Signature Page

Name:	Prof Jonathan Leor, MD
Address:	Head, Neufeld Cardiac Research Institute.

Tel-Aviv University
Sheba Medical Center
Tel-Hashomer 52621
Israel
Phone: +972-3-534-8685
Fax: +972-3-5351139

I have read the protocol and confirm that the protocol follows the current GCP guidelines.

Date/Place 28/1/09 Signature /s/ Jonathan Leor
(Jonathan Leor, MD, Medical Advisor)

Synopsis

STUDY NUMBER	BL-1040.01
TITLE OF THE STUDY	A Phase I, multi-center, open label study designed to assess the safety and feasibility of the injectable BL-1040 implant to provide scaffolding to infarcted myocardial tissue
STUDY CENTER/COUNTRY	Approximately **10** centers in 3 countries: ~~Netherlands~~, Belgium, Germany, ~~Israel~~, possibly Switzerland
PLANNED STUDY PERIOD + CLINICAL PHASE	Q1 2008 to **Q1 2010** Phase I
INDICATION AND RATIONALE	Heart failure after myocardial infarction (MI) is often precipitated by early and progressive extracellular matrix degradation and pathological remodeling of the left ventricle (LV). In response to MI, a series of molecular, cellular and physiological responses are triggered, which can lead to early infarct expansion (infarct thinning), which may result in early ventricular rupture or aneurysm formation and the transition to heart failure. Late remodeling involves the left ventricle globally and is associated with time-dependent dilatation, and the distortion of ventricular shape. The failure to normalize increased wall stresses results in progressive dilatation, recruitment of border zone myocardium into the infarct, and deterioration in contractile function. Current anti-remodeling therapies are clearly limited, as many ventricles continue to enlarge and mortality and morbidity remain significantly high. Based on the mechanism of LV remodeling, it has been hypothesized that injection of biomaterials into the infarct could thicken the infarct, arrest infarct expansion, prevent LV dilatation and reduce wall stress that initiates progressive adverse LV remodeling. BL-1040 Myocardial Implant is a non-pharmacologic cross-linked alginate solution administered via intracoronary (IC) injection to infarcted tissue, forming a flexible, three-dimensional mechanical scaffold. BL-1040 Myocardial Implant presents a novel, safe and non-surgical therapy that directly addresses the stability and structural integrity of myocardial tissue while potentially preventing post infarction remodeling, primarily via limiting left ventricle dilation.
OBJECTIVES	· To evaluate the safety of the BL-1040 myocardial implant in patients after MI at high risk for LV remodeling and CHF. · To provide feasibility data in order to initiate and conduct a pivotal clinical study evaluating the safety and efficacy of the BL-1040 implant in patients following myocardial infarction.

ENDPOINTS	Primary safety endpoints
	Occurrence of all adverse events including but not limited to
	· All MIs
	· Cardiovascular hospitalization
	· Serious ventricular arrhythmias sustained:
	  · VT (symptomatic or sustained VT [duration longer than 30 seconds or 100 beats, or associated with hemodynamic collapse])
	  · VF
	  · symptomatic bradycardia, pauses of longer than 3.0 seconds, complete atrioventricular block, Mobitz II atrioventricular block
	· Symptomatic heart failure (NYHA criteria + physical examination OR hospitalization due to heart failure)
	· Renal failure
	· Stroke
	· Death

	Secondary safety endpoints · Change from baseline in LV dimensions (end-systolic volume index, end-diastolic volume index, left ventricular mass) · Change from baseline in regional (infarct related) and global wall motion score · Change from baseline in ejection fraction · Cardiac rupture · NT-proBNP
DESIGN	Multi-center, open label
PATIENTS	NUMBER Maximum 30

	MAIN INCLUSION CRITERIA	· Signed informed consent · 18 to 75 years of age, inclusive · Male or female · Negative pregnancy test for women of child-bearing potential, or surgically sterile, or post menopausal · Acute MI defined as: 1. Typical rise and gradual fall (troponin) or more rapid rise and fall (CK-MB) of biochemical markers of myocardial necrosis with at least one of the following: a) ischemic symptoms: b) development of pathologic Qwaves on the ECG: c) ECG changes indicative of ischemia (ST segment elevation or depression) 2. First anterior or inferolateral STEMI or Qwave MI (QMI Anterior: V1-V3 or V1-V4 or V1-V5 or V1-V6.QMI Inferior: L2, L3, AVF, or L2, L3, AVF+ V5, V6 or L2, L3, AVF+ V6-V9 [posterior leads]) 3. Regional wall motion score index (at least 4 out of 16 akinetic segments) · One or more of the following: · LVEF >20% and <45% measured and calculated by 2-dimensional measurement · Biomarkers: peak CK > 2000 IU · Infarct size > 25% as measured by MRI · Successful revascularization with PCI ~~with 1 stent only~~, within 7 days of the index MI (only safe and MRI compatible stents) · At time of application of study device, patient must have patent infarct related artery (IRA) and TIMI flow grade = 3
	MAIN EXCLUSION CRITERIA	· History of CHF, Class I to Class IV, as per NYHA criteria · History of prior LV dysfunction · At time of application of study device - Killip III-IV (pulmonary edema, cardiogenic shock - hypotension [systolic < 90 mmHg] and evidence of peripheral hypoperfusion [oliguria, cyanosis, sweating]) or HR > 100 bpm · Patient with pacemaker · Prior CABG · Prior MI · History of stroke · Significant valvular disease (moderate or severe) · Patient is a candidate for CABG or PCI on non-IRA · Patient is being considered for CRT within the next

		30 days · Renal insufficiency (eGFR < 60) · Chronic liver disease (> 3 times upper limit of normal) · Life expectancy < 12 months · Current participant in another clinical trial, or participation in another trial within the last 6 months · Any contraindication to coronary angiography, MRI or PCI procedures · Patient taking anti-coagulation medication prior to MI · Pregnant or lactating women; pregnancy confirmed by urine pregnancy test
STUDY DEVICE	ROUTE OF APPLICATION	Administered via intracoronary (IC) injection, using multiple commercially available devices
	DURATION AND FREQUENCY	2 mL of BL-1040 administered for no longer than **30** seconds
	FORMULATION	Calcium D-Gluconate (Gluconic acid hemicalcium salt) PRONOVA UP VLVG (Generic name: Sodium Alginate) Water for Injection USP/EP
SAFETY EVALUATIONS		
TIMING AND ASSESSMENTS PERFORMED		Screening · lst Coronary angiography, PCI and stent (as part of treatment of MI) · Physical examination · Vital signs · 12-lead ECG · Blood and urine sampling for laboratory safety parameters (biochemistry, hematology and urinalysis) · Total CK/CK MB · NT-proBNP · Mandatory echocardiography; MRI as an additional measurement is encouraged Telephone contact, 1 week post-procedure · Phone call to confirm status of patient discharged from the hospital Day 1 and during hospitalization · Physical examination daily during hospitalization · Vital signs daily during hospitalization

- 12-lead ECG prior to and after administration of BL-1040; daily during hospitalization
- 24 hour Holter monitor (after completion of 12-lead ECG)
- Blood and urine sampling for laboratory safety parameters (biochemistry, hematology and urinalysis), on Day 1 (only if not done within the previous 48 hours) and on day of discharge (only if not done within the previous 48 hours)
- Total CK/CK MB measured prior to, and 8, 16, 24 and 48 hours after administration of BL-1040
- NT-proBNP on Day 1 (only if not done within the previous 48 hours) and on day of discharge (only if not done within the previous 48 hours)
- continuous ECG during the procedure
- 2nd cardiac catheterization (for implantation of BL-1040)
- PTT or ACT measurements, during procedure only (prior to implantation of BL-1040 and prior to removal of sheath)

Follow-up visits (Days 30, 90 180 [End of Study]; Months 12, 24, 36, 48 and 60)
- Physical examination
- Vital signs
- 12-lead ECG
- 24 hour ambulatory Holter monitoring
- Blood and urine sampling for laboratory safety parameters (biochemistry, hematology and urinalysis)
- NT-proBNP (through Day 180 only)
- Mandatory echocardiography: MRI as an additional measurement is encouraged (MRI through Day 180 only)
- Minnesota Living with Heart Failure® questionnaire

AEs and SAEs will be collected throughout the study

PROCEDURE

Patient is admitted to the hospital as a result of an AMI. As part of the inclusion criteria for this study, the patient will undergo revascularization with PCI stent implantation. Within 7 days of the index MI, the patient will undergo an echocardiogram to determine LVEF. Although not mandatory, the patient will be encouraged to undergo an MRI as an additional assessment. If the patient satisfies inclusion/exclusion criteria, a 2nd cardiac catheterization will be performed to administer BL-1040 after revascularization but within 7 days of the index AMI. BL-1040 is applied via intracoronary injection through the infarct related artery. Patients discharged from the hospital will be contacted by phone on Day 8 for a safety follow-up. Follow-up examinations are scheduled for Day 30, Day 90 and Day 180 (End of Study) post-procedure. In addition, the patient will return to the hospital at Months 12, 24, 36, 48 and 60 for yearly follow-up assessments, as part of a long-term safety follow-up.

STATISTICAL METHODS

All data recorded will be presented in data listings and summary tables, as appropriate. Missing values will not be replaced.
No formal hypothesis testing will be performed.
All participants who received BL-1040 will be included in the safety analysis.
Any excluded cases will be documented together with the reason for exclusion.
All decisions on exclusions from the analysis will be finalized prior to database lock.
Continuous variables (age, height, weight) will be summarized using mean, median, standard deviation, minimum, maximum, and number of available observations. Qualitative variables will be summarized by counts and percentages.
An interim safety analysis will be performed after 5 patients have completed the Day 30 visit, on all data collected up to this timepoint.

Schedule of Events

Visits/Week	Hospitalization				Post discharge follow-up				
Study days	Screening (Day) (-7) to Day (-1)	Day 1 Day of application(1)	Daily during hospitalization(2)	Day of discharge	Telephone Contact Day 8 (± 1 day)	Day 30 (± 5 days)	Day 90 (± 5 days)	Day 180 (± 7 days) End of Study Visit	Follow-up Safety Visits (Months 12, 24, 36, 48 60, ± 30 days)
AMI	X								
Hospitalization	X	X	X	X	X				
Coronary angiography, PCI, stent(1)	X								
Informed consent	X								
Inclusion/exclusion criteria	X								
Pregnancy test	X								
Demography; medical history; concurrent illnesses	X								
Physical examination	X	X	X	X		X	X	X	X
Vital signs (temperature, arterial BP, weight)	X	X	X	X		X	X	X	X
12-lead ECG	X	X(4)	X	X		X	X	X	X
Laboratory safety parameters	X(5)	X(6)		X(6)		X	X	X	X
Total CK/CK MB	X	X(7)							
NT-proBNP	X	X(6)		X(6)		X	X	X	
Echocardiography/MRI	X					X	X	X	X
Continuous ECG monitoring		X(9)							
Cardiac catheterization; application of BL- 1040; coronary angiography		X							
PTT or ACT measurements		X(10)							
24-hour ambulatory Holter monitoring		X				X	X	X	X
Safety contact for discharged patients					X				
Minnesota Living with Heart Failure						X	X	X	X
Serious/Adverse events and concomitant medication	X	X	X	X	X	X	X	X	X

(1) Device to be administered within 7 days of AMI
(2) Patient must remain hospitalized for at least 48 hours after procedure.
(3) Done as treatment of AMI
(4) Prior to and after administration of BL-1040
(5) Troponin I or T to be measured at Screening only
(6) If not done within previous 48 hours
(7) Parameters to be assessed prior to, and 8, 16, 24 and 48 hours after administration of BL-1040
(8) Echocardiography to be done at each visit. MRIs are to be encouraged as an additional assessment through Day 180, but are contingent upon patient agreement. MRIs are not to be requested as part of the Follow-up Safety visits.
(9) Patient to be connected prior to implantation of BL-1040, and for the duration of the procedure
(10) Measured prior to implantation of BL-1040, and prior to removal of sheath

Table of Contents

List of Abbreviations

AE(s)	Adverse event(s)
ALT	Alanine transminase
AMI	Acute myocardial infarction
AST	Aspartate transaminase
BP	Blood pressure
bpm	Beats per minutes
BUN	Blood urea nitrogen
CABG	Coronary artery bypass graft
CHF	Chronic heart failure
CRF	Case Report Form
CRT	Cardiac Resynchronization Therapy
CV	Cardiovascular
ECG	Electrocardiogram
EF	Ejection fraction
eGFR	Estimated glomerular filtration rate
EOS	End of study
GCP	Good Clinical Practice
GGT	Gamma glutamyl transferase
GLP	Good Laboratory Practice
GMP	Good Manufacturing Practices
HPF	High power field
HR	Heart rate
IC	Intracoronary
ICH	International Conference on Harmonization
IRA	Infarct related artery
ISMB	Independent Safety Monitoring Board
LDH	Lactate dehydrogenase
LV	Left ventricle
LVEF	Left ventricular ejection fraction
MedDRA	Medical Dictionary for Regulatory Activities
mg	Milligram
MI	Myocardial infarction

min	Minute
mL	Milliliter
MRI	Magnetic resonance imaging
NCE	New chemical entity
NT-proBNP	N-terminal prohormone brain natnuretic peptide
NYHA	New York Heart Association
°C	Degrees centigrade
OTC	Over the Counter
PCI	Primary coronary intervention
QMI	Qwave myocardial infarction
SAE(s)	Serious Adverse Event(s)
SAS	Statistical Analysis System
STEMI	ST-segment elevation myocardial infarction
TIMI	Thrombolysis in Myocardial Infarction
VF	Ventricular fibrillation
VT	Ventricular tachycardia

1 Introduction

1.1 Background

1.1.1 Acute Myocardial Infarction- Definition

Acute myocardial infarction (AMI) is defined as death or necrosis of myocardial cells. It is a diagnosis at the end of the spectrum of myocardial ischemia or acute coronary syndromes. AMI occurs when myocardial ischemia exceeds a critical threshold and overwhelms myocardial cellular repair mechanisms that are designed to maintain normal cardiac function. Ischemia at this critical threshold level, when present for an extended time period, results in irreversible myocardial cell damage and cell death.

1.1.2 Infarction types and pathogenesis

Critical myocardial ischemia may arise as a result of increased myocardial metabolic requirement and/or reduction in the delivery of oxygen and nutrients to the myocardium through the coronary circulation, or both. An interruption in the supply of myocardial oxygen and nutrients occurs when blood flow to the myocardium is interrupted by occlusion of a coronary artery. Often, this event is caused by a thrombus superimposed on an ulcerated or unstable atherosclerotic plaque that left untreated for as little as a 20-40 minutes, can lead to irreversible cell damage and cell death. A high-grade (> 75%) permanent coronary artery stenosis due to atherosclerosis or a dynamic stenosis coupled with coronary vasospasm can also reduce the supply of oxygen and nutrients and be a factor involved in AMI. Additional cardiac valvular pathologies and low cardiac output states associated with a decreased aortic diastolic pressure, which is the prime component of coronary perfusion pressure, can also precipitate AMI.

1.1.3 Mechanisms of myocardial damage

The severity of an AMI is dependent on three factors: the level of the occlusion in the coronary artery, the length of time of the occlusion, and the presence or absence of collateral circulation. In general, the more proximal the coronary occlusion, there is a greater risk of an increased area of necrosis. The larger the AMI, the chance of death due to a mechanical complication or pump failure increases. In addition, the longer the time period of vessel occlusion, there is a greater chance of irreversible myocardial damage distal to the occlusion.

The death of myocardial cells first occurs in the area of myocardium that is most distal to the arterial blood supply, the endocardium. As the duration of the occlusion increases, the area of myocardial cell death enlarges, extending from the endocardium to the myocardium and ultimately to the epicardium. The area of myocardial cell death then spreads laterally to areas of watershed or collateral perfusion. The extent of myocardial cell death defines the magnitude of the AMI. If blood flow can be restored to at-risk myocardium, more heart muscle can be saved from irreversible damage or death. The ischemic zone will undergo inflammatory necrotic changes, and the myocardial tissue will eventually be completely replaced by fibrous infarct tissue. In the early stages after an AMI, the damage causes deterioration of cardiac muscle contractility and structural integrity. This results in thinning of the walls of the heart, which can have severe consequences including rupture at the site, expansion of the area of damage, and the formation of blood clots. After some weeks or months, this can evolve to dilatation of the heart, which further reduces its ability to pump blood efficiently, resulting in heart failure.

1.1.4 Treatment of AMI

The goal of treatment for AMI is early reperfusion by rapid revascularization of the occluded culprit coronary artery both by medical means to dissolve the clot with thrombolytics or by cardiac catheterization with primary coronary intervention (PCI) and deployment of stents to

maintain patency of the culprit coronary artery. However, while re-opening of the culprit coronary vessel can prevent the development of a large AMI and prevent further loss of viable myocardium, it does not affect myocardial tissue that has already undergone irreversible damage. An undeniable adverse outcome of AMI is progressive worsening of ventricular function that, if left unattended, culminates in the syndrome of congestive heart failure. To date, no treatment has been developed to reliably prevent the deterioration of ventricular function that follows a large AMI. Treatment options for AMI and for the resulting heart failure include medical management, heart transplantation. mechanical circulatory assist devices (left ventricular assist device, etc.), and surgical ventricular restoration, all of which have specific limitations.

1.2 Rationale and justification

BL-1040 Myocardial Implant presents a novel, safe and non-surgical therapy that directly addresses the stability and structural integrity of myocardial tissue in this patient population. BL-1040 potentially prevents post infarction remodeling primarily via limiting left ventricle (LV) dilation, while the untreated patient LV will

continue to dilate or enlarge. BL-1040, by creating a scaffold, may stabilize the AMI and limit post AMI expansion manifested as LV dilation.

There are currently no other available medical and/or surgical interventions that directly address the stability and structural integrity of myocardial tissue damaged as a result of AMI. In the setting of an AMI, an inflammatory response triggers the degradation of the extracellular matrix, thus weakening of the collagen cross-link structure or structural “backbone” of the myocardium. Degradation of the extracellular matrix leads to infarct expansion manifested by myocardial wall thinning and often, aneurysmal dilation with subsequent ventricular enlargement. This process results in progressive LV remodeling and increased LV wall stress. The latter can increase myocardial oxygen consumption, a condition that the infarcted and/or failing LV can ill afford and one that can contribute to increased long-term mortality and morbidity.

LV dilation is the predominant cause for morbidity and mortality in congestive heart failure [2], demonstrated that patients with LV end systolic volume smaller than 95 mL showed a 94 % survival after 5 years while LV patients with LV end systolic volume greater than 130 mL showed a 52 % survival after 5 years. Both diastolic and systolic were the main predictors for mortality. Patients with end-stage ischemic heart failure presenting dilated LV with an akinetic/dyskinetic region over 35% and with left ventricular end systolic index >60 mL/m2 are offered LV reconstruction or surgical ventricular restoration (SVR) in order to reduce LV volume and to restore normal LV shape. Overall, in a large number of studies performed using SVR, there is strong evidence that SVR is safe and effective, showing significant reduction in mortality and readmission levels together with significant improvement in ejection fraction as well as in LV end systolic/diastolic index.

2 Study Objectives

The objectives of this study are:

- to evaluate the safety of the BL-1040 myocardial implant in patients after MI at high risk for LV remodeling and CHF, and
- to provide feasibility data in order to initiate and conduct a pivotal clinical study evaluating the safety and efficacy of the BL-1040 implant in patients following myocardial infarction.

3 Safety Endpoints

3.1 Primary endpoints

Primary safety endpoints include:

- occurrence of all adverse events including but not limited to
  - all MLs
  - cardiovascular hospitalization
  - serious ventricular arrhythmias sustained
    - VT (symptomatic or sustained VT [duration longer than 30 seconds or 100 beats, or associated with hemodynamic collapse])
    - VF
    - symptomatic bradycardia, pauses of longer than 3.0 seconds, complete atrioventricular block, Mobitz II atrioventricular block
  - symptomatic heart failure (NYHA criteria + physical examination OR hospitalization because of heart failure)
  - renal failure
  - stroke
  - death

3.2 Secondary endpoints

Secondary safety endpoints include:

- change from baseline in LV dimensions (end-systolic volume index, end-diastolic volume index, left ventricular mass)
- change from baseline in regional (infarct related) and global wall motion score
- change from baseline in ejection fraction
- cardiac rupture
- NT-proBNP

4 Investigational Plan

4.1 Summary of study design

This is an open label, multi-center, sequentially enrolled. Phase I study to assess the safety and feasibility of the injectable BL-1040 myocardial implant to provide scaffolding to infarcted myocardial tissue.

Patients who experience an MI will be admitted to the hospital. As part of the treatment for the MI, patients will undergo PCI and stent implantation. Patients will also undergo an echocardiography (and if they agree, an MRI) to determine the extent of damage to the infarct related artery (IRA). Patients who satisfy inclusion/exclusion criteria will be enrolled into the study. The BL-1040 myocardial implant will be injected into the IRA, distally to the implanted stent.

The first 2 patients will be sequentially enrolled. After both patients have completed Day 30 assessments, and after approval by the Independent Safety Monitoring Board (ISMB), the decision will be made to enroll 3 additional patients. After the ISMB reviews the Day 30 assessments of these patients, the decision will be

made to enroll a maximum of 25 additional patients. Details are provided in Sec. 4.2.

Both female and male patients must agree to use effective contraception (as agreed with the Investigator) for 6 months (180 days) after the procedure.

4.1.1 Estimated study duration

The study is planned to last from Q1 2008 to **Q1 2010**. The clinical study phase is **180** days for each patient. A long term safety follow-up will include visits at Months 12, 24, 36, 48, and 60. Patients will be consented for the entire 5 year period.

4.1.2 Number of Patients

The maximum number of patients enrolled in this study will be 30.

4.2 Sequential enrollment

The first 2 patients will be sequentially enrolled into the study. After the 1st patient has completed Day 30 assessments, the Independent Safety Monitoring Board (ISMB, Sec. 4.3) will review the patient's data through Day 30. The ISMB will then decide whether to give approval to enroll the 2nd patient. After the 2nd patient has completed Day 30 assessments, the ISMB will again review the data and provide approval for enrollment of the next 3 patients. After all 3 patients have completed Day 30 assessments, the ISMB will review the data from these patients and provide approval for opening enrollment to the balance of the patients (maximum of 25).

4.3 Responsibilities of the Independent Safety Monitoring Board

An Independent Safety Monitoring Board (ISMB) will be established prior to the start of the study to monitor the safety of BL-1040 during the conduct of the protocol. This ISMB will consist of physicians with expertise in cardiovascular disease, particularly in the area of coronary artery disease and with experience monitoring safety of drugs and/or devices for cardiovascular applications, and will have no participation in the trial in any other capacity.

The ISMB will ensure that this study meets the highest standards of patient safety. During the study the ISMB will have the following main responsibilities:

- review 30 day safety data patients from the first 2 sequentially enrolled patients to determine whether 3 additional patients may be enrolled: after reviewing the 30 day safety data from these 3 patients, will determine whether the balance of patients may be enrolled
- within 30 days of enrolment of each successive group of 5 patients receiving the device, will review all SAEs occurring to date and will recommend continuation, discontinuation, or modification of the procedure or protocol, based on a determination of whether the occurrence of serious, unexpected, or device-related adverse events (Sec. 7) might outweigh the potential benefit achievable with the device
- review emerging findings in patients and identify potential safety concerns with BL-1040
- will receive information, on an expedited basis, on all Serious Adverse Events (SAEs), clinically significant laboratory values/vital signs, ECG abnormalities and data from patients who decided to prematurely discontinue the study. All SAES that occur in the cath lab during or after the procedure to administer BL-1040 should be reviewed promptly by the ISMB. The ISMB will review this information and may decide to interrupt, alter, or terminate the trial
- will adjudicate whether or not an event is unexpected, based on a pre-specified list of expected SAEs within the study population.

4.3.1 Stopping Criteria

Given the uncontrolled nature of the study, and the small sample size, it is not practical to provide a quantitative stopping rule.

Moreover, given the severely ill nature of the patients who will be enrolled in the study (those with large myocardial infarction and substantial LV dysfunction), adverse cardiac outcomes, including fatal ones, are to be expected in this population, regardless of participation in the study.

The study will be stopped when any of the following occur:

1. Completion of the study
2. ISMB and sponsor judge that the study treatment appears to be unsafe for patients. The ISMB will make this assessment based not only upon the frequency of observed complications, but also upon the character and qualitative nature of the events. This determination will be made in the context of clinical judgement of experienced cardiologists regarding the expected outcome in this population of patients and whether observed outcomes differ substantively from the expectation. The committee reserves the right to stop the study after analysis of outcomes of sequential procedures. A decision to stop will be considered by the ISMB in the event of occurrence of severe, unusual or unexpected events.
3. The ISMB may consider putting the trial on hold or terminating it and will base it decision on weighing the balance between potential but hypothetical benefits and possible risks to the participants in the study.

4.4 Inclusion criteria

The inclusion criteria for this study are:

- voluntarily signed the informed consent form prior to the conduct of any study specific procedures
- male or female inpatients aged 18 to 75, inclusive

- negative pregnancy test for all women of child-bearing potential, or surgically sterilized (i.e. tubal ligation, hysterectomy) prior to Screening, or post-menopausal for at least 1 year
- acute MI defined as:
  - typical rise and gradual fall (troponin) or more rapid rise and fall (CK-MB) of biochemical markers of myocardial necrosis with at least one of the following: a) ischemic symptoms; b) development of pathologic Qwaves on the ECG; c) ECG changes indicative of ischemia (ST segment

elevation or depression)
    - first anterior or inferolateral STEMI or Qwave MI (QMI Anterior: V1-V3 or V1-V4 or V1-V5 or V1-V6.QMI Inferior: L2, L3, AVF, or L2, L3, AVF+ V5, V6 or L2, L3, AVF+ V6-V9 [posterior leads])
    - regional wall motion score index (at least 4 out of 16 akinetic segments)
- one or more of the following:
    - LVEF >20% and <45% measured and calculated by 2-dimensional measurement
    - Biomarkers: peak CK > 2000 IU
    - infarct size > 25% as measured by MRI
- successful revascularization with PCI within 7 days of the index MI (only safe and MRI compatible stents)
- at time of application of device patient must have patent infarct related artery (IRA) and TIMI flow grade = 3

4.5 Exclusion criteria

Exclusion criteria for this study are:

- history of CHF, Class I to Class IV, as per NYHA criteria
- history of prior LV dysfunction
- at time of application of study device - Killip III-IV (pulmonary edema, cardiogenic shock - hypotension (systolic < 90 mmHg) and evidence of peripheral hypoperfusion (oliguria, cyanosis, sweating) or HR > 100 bpm
- patient with pacemaker
- prior CABG
- prior MI
- history of stroke
- significant valvular disease (moderate or severe)
- patient is a candidate for CABG or PCI on non-IRA
- patient is being considered for CRT within the next 30 days
- renal insufficiency (eGFR < 60)
- chronic liver disease (> 3 times upper limit of normal)
- life expectancy < 12 months
- current participant in another clinical trial, or participation in another trial within the last 6 months
- any contraindication to coronary angiography, MRI or PCI procedures
- patient taking anti-coagulation medication prior to MI
- pregnant or lactating women; pregnancy confirmed by urine pregnancy test
- patients with a reasonable likelihood for non-compliance with the protocol
- any other reason that, in the Investigator’s opinion, prohibits the inclusion of the patient into the study

4.6 Withdrawal criteria during the study

Each patient has the right to withdraw from the trial at any time for any reason.

The Investigator must make at least 3 documented attempts to contact those patients who do not return for the scheduled follow-up visits. Attempts must be recorded in the patient’s file.

The Sponsor reserves the right to terminate the study at any time.

Upon withdrawal from the study any time after administration of study device, the patient will undergo the End of Study assessments (Section 6.2.1.5: Table 6.1).

Dropouts that occur after implantation of BL-1040 will not be replaced.

4.7 Treatment allocation

This is an open label study. All patients will be treated with BL-1040. Patient eligibility will be established prior to treatment with BL-1040.

If a patient discontinues from the study, the patient number will not be reused.

4.8 Method of blinding and unblinding

As this is an open label study, there will be no blinding or unblinding procedure.

5 Product Overview

5.1 BL-1040

BL-1040 myocardial implant is a non-pharmacologic, non-surgical, cross-linked alginate solution administered via intracoronary (IC) injection to infarcted tissue. BL-1040 completely disintegrates into its constituent polymers within approximately 90 days after deposition, and is excreted in the urine.

5.2 Formulation

The formulation of BL-1040 is shown in Table 5.1.

Table 5.1 Formulation of BL-1040

0.3% Calcium D-Gluconate (Gluconic acid hemicalcium salt)	Sigma, Dr. Paul Lohmann GmbH KG
1% PRONOVA UP VLVG Generic name: Sodium Alginate	FMC BioPolymer/ NovaMatrix
Water for Injection USP/EP	

5.3 Dosage and application

BL-1040 will be administered to the coronary vasculature using multiple commercially available devices. Table 5.2 provides a list of the commercially available components that will be required in order to delivery the BL-1040 implant.

Table 5.2 List of Commercially Available BL-1040 Delivery Devices

BL-1040 Implant Delivery Devices

1 Standard endovascular sheath (femoral or radial or brachial)
2 Standard coronary guiding catheter (example — Launcher, ref LA6AR10SH)
3 Guidewire 0.014 inch (example - Boston Scientific, ref. 383931-035J)
4 Torque device (example - Boston Scientific, ref. K903606))
5 Guidewire introducer (example Input Ref. 87311)
6 Microcatheter designed for coronary intravascular use such as multipurpose probing endovascular microcatheter. Example:(Boston Scientific Catalog number SCH 50058) or Transit microcatheter, (Cordis Endovascular Systems, MiMI Lakes, Fla.) or Renegase Hi-Flo microcatheter (Boston Scientific)
7. Disposable syringe, Intmed 5 mL sterile CE, ISO9001, ISO13488

Cardiac catheterization should be done according to the guidelines of the American College of Cardiology/Society for Cardiac Angiography and Interventions Clinical Expert Consensus Document on Cardiac Catheterization Laboratory Standards. All angiographies will be evaluated by a core laboratory. BL-1040 is delivered intra-coronary (IC) via a microcatheter that is intended for coronary intravascular use.

The timing of BL-1040 administration is within 7 days after the index MI. Two (2) mL of BL-1040 will be injected IC through the infarct related artery supplying the infarcted area. BL-1040 may not be mixed with any contrast medium.

All patients will be treated in the same manner.

Detailed instructions for the application of BL-1040 are provided in a separate Instruction Manual.

5.4 Labelling/Packaging

BL-1040 will be packed in a sterile cylindrical injection vial, type A glass. Vials are filled with sterile BL-1040 and sealed with a 20 mm rubber stopper, spun-on aluminum seal and a flip-off top.

All packages will be labeled according to the GMP guideline Volume 4, Annex 13 Manufacture of Investigational Medicinal Products (July 2003 Revision 1) [**1**] and local laws.

BL-1040 will be packed in labeled boxes, with at least the following information: study number, patient number, route of administration, storage guidelines, batch number, expiry date, instructions for administration, manufacturer name/code, and “Investigational use only”.

The Sponsor must notify the Site Investigator, who has the overall responsibility for the study device, of the anticipated date of arrival.

5.5 Storage

The Site Investigator is responsible for ensuring that BL-1040 is stored in a safe refrigerated location (2-8° C) with controlled access. At this temperature, BL-1040 has a shelf life of 3 months. The temperature must be monitored once daily, and recorded on a temperature log.

BL-104 must be removed from the refrigerator and kept at room temperature 30 minutes prior to administration.

5.6 Compliance

BL-1040 will be administered by the Investigator only, and will not be dispensed to the patient or any other personnel.

5.7 BL-1040 accountability

Under no circumstances is it permitted to use study supplies for any purposes other than those specified in the protocol.

The Investigator will be provided with forms to enable accurate recording of all investigational product at all times. The Investigator must sign a statement that he/she has received BL-1040 for the study. At any time the figures of supplied, used and remaining BL-1040 must match. At the end of the study, it must be possible to reconcile delivery records with those of used and unused stocks. Account must be given of any discrepancies.

At the end of the study, all unused BL-1040 supplies and empty containers must be returned to the Sponsor.

5.8 Concomitant medication

The following medications may only be administered as indicated:

- ceftriaxone may not be administered during the 48 hours immediately prior to the administration of BL-1040, and for the 48 hours immediately following administration of BL-1040
- calcium solutions may not be administered during the first week of the study

The introduction of any medication not allowed by the protocol at any point in the study will require a discussion between the Investigator and the Sponsor. If, in the opinion of the Investigator, it becomes necessary to administer any medication during the study, the

Investigator will determine the dose and time of intake, and document the medication(s) in the patient's CRF.

Patients must be instructed not to begin any new medication before consulting with the Investigator (unless required for emergency medical use). The patient must be instructed that this prohibition applies to over-the-counter products as well as prescription drugs.

All patients will receive optimal medical therapy according to the relevant, updated guidelines from the European Society of Cardiology [3,4,5]. Optimal therapy including aspirin, anticoagulation if indicated, angiotensin-converting-enzyme inhibition, beta-blockade, aldosterone antagonists, when appropriate, and lipid-lowering therapy, unless contraindicated. Clopidogrel therapy will be initiated before PCI and continued for 1 year after myocardial infarction [3].

6 Study Procedures

6.1 General study aspects

This is an open label, multi-center study to assess the safety and feasibility of the injectable BL-1040 myocardial implant to provide scaffolding to infarcted myocardium.

Patients will be admitted to the hospital for treatment of an acute myocardial infarction (AMI), to include angioplasty and implantation of a-stent/s. Within 7 days of successful revascularization, patients will undergo an echocardiogram for assessment of the extent of the changes to the heart, and to verify cardiac inclusion/exclusion criteria. MRIs are to be encouraged as an additional assessment, but are contingent upon the agreement of the patient. After the echocardiogram/MRI, but still within 7 days of the index AMI, patients will undergo a 2nd cardiac catheterization to administer BL-1040. Patients will remain hospitalized for at least 48 hours after the procedure.

The BL-1040 scaffold will be injected into one infarct related artery (IRA), distally to the implanted stent/s. Patients will undergo cardiac monitoring before, during and after the procedure: a 12-lead ECG will be done prior to and after administration of BL-1040; patients will be connected to a continuous ECG monitor and will have continuous hemodynamic measurements during the procedure; immediately after the completion of the 12-lead ECG, a Holter monitor will be placed and will remain connected for the following 24 hours.

Patients will undergo physical examinations, assessment of vital signs and an ECG daily during hospitalization; safety blood sampling will be done on the day of discharge.

Patients who have been discharged from the hospital will be contacted by phone on Day 8 to confirm the administration of any concomitant medications, general status of the patient, and any doctor visits since hospital discharge.

Patients will return for follow-up visits on Day 30, Day 90 and Day 180 (End of Study). Additional follow-up safety visits are planned for Months 12, 24, 36, 48 and 60. At each visit, patients will again undergo a physical examination with measurement of vital signs, ECG, blood sampling, echocardiography and completion of the Minnesota Living with Heart Failure questionnaire®. At each follow-up visit, the patients will be hooked up to a 24-hour ambulatory Holter monitor, which will be returned the following day. MRIs are to be encouraged through Day 180 as an additional assessment, but are contingent upon the agreement of the patient. MRIs are not to be requested as part of the long term safety visits.

Echocardiograms, ECGs, Holters, angiographies and MRIs, will be evaluated in a core laboratory.

The first 2 patients will be sequentially enrolled; if approved by the ISMB; 3 additional patients will be enrolled. After review and approval of the 30 day safety data from these 3 patients, the balance of patients may be enrolled. Details are provided in Sec. 4.2.

Both female and male patients must agree to use effective contraception (as agreed with the Investigator) for 6 months (180 days) after the procedure.

6.2 Outline of study procedures

All study procedures are outlined in the Schedule of Assessments below (Table 6.1). A more detailed description of the study procedures performed at each study stage/visit is given in the following sections.

Table 6.1 Schedule of Events

	Hospitalization				Post discharge follow-up				
Visits/Week Study days	Screening Day (-7) to Day (-1)	Day 1 Day of application(1)	Daily during hospitalization(2)	Day of discharge	Telephone Contact Day 8 (± 1 day)	Day 30 (± 5 days)	Day 90 (± 5 days)	Day 180 (± 7 days) End of Study Visit	Follow-up Safety Visits (Months 12, 24, 36, 48 60, ± 30 days)

AM1	X								
Hospitalization		X							
Coronary angiography, PCI, stent(1)	X								
Informed consent	X								
Inclusion/exclusion criteria	X								
Pregnancy test	X								
Demography medical history; concurrent illnesses	X								
Physical examination	X	X	X	X		X	X	X	X
Vital signs (temperature, arterial BP. weight)	X	X	X	X		X	X	X	X
12-lead ECG	X	X(4)	X	X		X	X	X	X
Laboratory safety parameters	X(5)	X(6)		X(6)		X	X	X	X
Total CK/CK MB	X	X(7)							
NT-proBNP	X	X(8)		X(6)		X	X	X	
Echocardiography/MRI	X					X	X	X	X
Continuous ECG monitoring		X(9)							
Cardiac catheterization; application of BL- 1040; coronary angiography		X							
PTT or ACT measurements		X(10)							
24-hour ambulatory Holler monitoring		X				X	X	X	X
Safety contact for discharged patients					X				
Minnesota Living with Heart Failure®						X	X	X	X
Serious/Adverse events and concomitant medication	X	X	X	X	X	X	X	X	X

(1) Device to be administered within 7 days of AMI
(2) Patient must remain hospitalized for at least 48 hours after procedure.
(3) Done as treatment of AMI
(4) Prior to and after administration of BL-1040
(5) Troponin I or T to be measured at Screening only
(6) If not done within previous 48 hours
(7) Parameters to be assessed prior to. and 8, 16, 24 and 48 hours after administration of BL-1040
(8) Echocardiography to be done at each visit. MRIs are to be encouraged as an additional assessment through Day 180, but are contingent upon patient agreement. MRIs are net to be requested as part of the Follow-up Safety visits.
(9) Patient to be connected prior to implantation of BL-1040, and for the duration of the procedure
(10) Measured prior to implantation of BL-1040, and prior to removal of sheath

6.2.1 Detailed description of study stages/visits

6.2.1.1 Screening, Day -7 to Day -1

Patients are admitted to the hospital for treatment of an AMI, prior to enrollment into the study. The treatment will include PCI with placement of a stent. After signing of Informed Consent, and prior to initiation of any study-related procedures, the following activities will be carried out:

- confirmation of inclusion/exclusion criteria
- negative pregnancy test for all women of child-bearing potential (as defined in Inclusion Criteria)
- demographics
- medical history
- physical examination
- vitals signs
- 12-lead ECG, in supine position
- blood and urine sampling for laboratory safety parameters (biochemistry, hematology and urinalysis)
- blood sampling for Total CK/CK MB
- blood sampling for NT-proBNP
- echocardiography
- MRI, if patient agrees
- concomitant medication record (all currently prescribed and over the counter medications must be recorded in the Case Report Form [CRF], with dose and reason for use)
- pre-device serious/adverse events

6.2.1.2 Day 1

BL-1040 must be implanted within 7 days of the index AMI; the day of implant will be considered Day 1 of the study. Prior to implantation, the following assessments will be carried out:

- physical examination
- vital signs
- 12-lead ECG

- blood and urine sampling for laboratory safety parameters (biochemistry [excluding troponin I or T], hematology, and urinalysis), if not done within the previous 48 hours
- Total CK/CK MB
- NT-proBNP, if not done within the previous 48 hours
- connection to continuous ECG monitoring

BL-1040 will be implanted in the infarcted tissue via the IRA, distally to the stent as outlined in the separate BL-1040 Instruction Manual. During the procedure the following assessments will be done:

- continuous ECG monitoring
- continuous hemodynamic measurements (arterial blood pressure)
- blood sampling for PTT or ACT, prior to implantation of BL-1040 and prior to removal of sheath

An additional coronary angiography will be done 3 minutes after implantation of the BL-1040, and will include an assessment of TIMI flow and myocardial blush.

The following assessments will be done after the procedure:

- urinalysis
- blood sampling at 8 hours, 16 hours and 24 hours after the procedure, for assessment of Total CK/CK MB
- 12-lead ECG
- connection to 24 hour Holtter monitor

Adverse events and concomitant medications will be monitored continuously during the procedure and recorded on the patient's CRF.

6.2.1.3 Daily during hospitalization

The patient must remain hospitalized for at least 48 hours after the procedure. The following assessments and procedures will be carried out during each day of hospitalization, including day of discharge:

- physical examination
- vital signs
- 12-lead ECG
- blood and urine sampling for laboratory safety parameters (biochemistry [excluding troponin I or T], hematology and urinalysis) on day of discharge and only if not done within the previous 48 hours
- NT-proBNP on day of discharge and only if not done within the previous 48 hours
- serious/adverse events
- concomitant medication

6.2.1.4 Telephone Contact, Day 8, ±1

Patients who have been discharged from the hospital will be contacted by phone 7 days after application of BL-1040. The patient should be asked the following questions:

1. How have you been feeling since your discharge? Have you had any chest pain or experienced any shortness of breath?
2. Did you call your doctor for any reason? If so, when, and for what reason? Did you go to the emergency room for any reason? If so, when and for what reason?
3. Are you taking any medications? If so, which ones?

The information collected from this phone call is to be recorded in the patient's CRF.

6.2.1.5 Day 30, Day 90 and Day 180 (End of Study)

The patient will return to the hospital for the following assessments and procedures on Day 30, Day 90 and Day 180. The visit on Day 180 will be considered the End of Study visit. If a patient is discontinued prior to Day 180 for any reason, the following assessments should be done at the time of discontinuation.

Assessments to be carried out include:

- physical examination:
- vital signs
- 12-lead ECG

- connection to 24-hour Holter monitor; to be returned on Day 31/Day 91/**Day 181**
- blood and urine sampling for laboratory safety parameters (biochemistry [excluding troponin I or T], hematology and urinalysis)
- NT-proBNP
- echocardiography
- MRI, if patient agrees
- completion of the Minnesota Living with Heart Failure® questionnaire
- serious/adverse events
- concomitant medication

6.2.1.6 Extended safety follow-up (Months 12, 24, 36, 48, 60 ±30 days)

Patients will return to the hospital yearly for completion of follow-up assessments.

Assessments are to include::

- physical examination
- vital signs
- 12-lead ECG
- connection to 24-hour Holter monitor; the patient is to be connected at the time of the follow-up visit, and the monitor is to be returned the following day
- blood and urine sampling for laboratory safety parameters (biochemistry [excluding troponin I or T], hematology and urinalysis)
- echocardiography
- completion of the Minnesota Living with Heart Failure® questionnaire
- completion of the following questions:
  - How have you been feeling since your last check up?
  - Have you been hospitalized for any reason? If so, when, and for what reason?
- serious/adverse events
- concomitant medication

6.3 Study evaluations and procedures

Safety will be evaluated by analyzing the results of physical examinations, laboratory examinations and cardiac assessments, as well as AEs (Section 7) and vital signs. Assessments will be carried out at the time points specified in Section 6.2, and as shown in Table 6.1.

All safety related investigations are to be performed by the Principal Investigator or a medically qualified designee, who is responsible for the overall treatment of the patient.

6.3.1 Safety

6.3.1.1 Physical examinations

Physical examinations will include height (Screening only), weight, and a general assessment of overall body systems (cardiovascular, respiratory).

6.3.1.2 Vital signs

The following vital signs will be assessed:

- pulse rate

- blood pressure (supine, systolic and diastolic)
- body temperature

The actual blood pressure and pulse rate should be recorded in the patient's CRF. Rounding of values is not allowed.

The following ranges will be used to define acceptable blood pressure:

- supine systolic blood pressure: 100 - 160 mmHg
- supine diastolic blood pressure: 60 - 95 mmHg
- supine pulse <100 bpm

Body temperature should be measured using the same methodology at each assessment, and should be measured in decimals.

6.3.1.3 ECGs

A standard supine 12-lead ECG shall be recorded. ECG morphology and ECG intervals (PR, RR, QRS, QT, and QTc) will be determined: QTc will be calculated using Bazett's formula.

Patients will be connected to a 24-hour ambulatory Holter monitor at each follow-up visit (Day 30, Day 90, Day 180).

Printouts/copies must be placed in the patient's chart, clearly labeled with the patient number, time, date, visit, and study number, and signed by the Investigator. A core laboratory will evaluate the results of both the ECG and Holter.

6.3.1.4 Echocardiograms

Echocardiograms will be performed and recorded according to specific criteria established for this study, and provided in a separate Echocardiogram Reference Manual. The same parameters will be measured at each assessment, throughout the study.

A core laboratory will evaluate echocardiograms.

The Principal Investigator, the Sponsor or the ISMB may review echocardiograms at any time if any safety concerns arise. Echocardiograms will be performed at the times indicated on the Schedule of Events and in Sec. 6.2 of the protocol.

6.3.1.5 MRIs

While the MRI is an optional procedure for cardiac assessment at Screening and all follow-up visits (Day 30, Day 90, Day 180/End of Study), patients should be encouraged to undergo the procedure at each relevant visit. Performance of the procedure is always contingent upon patient agreement.

MRIs will be performed according to specific criteria established for this study, and provided in a separate MRI Reference Manual. A core laboratory will evaluate MRIs.

The Principal Investigator, the Sponsor or the ISMB may review MRIs at any time if any safety concerns arise.

6.3.1.6 Clinical safety evaluations

Safety blood sampling

All laboratory samples will be processed at the local laboratory, except for NT-proBNP, which will be assessed at a core lab.

The Investigator must review the laboratory assessments (initialed and dated) within 24 hours after the receipt of those results. Out of range values will be interpreted by the Investigator with a comment of “not clinically significant” (NCS) or “clinically significant” (CS). Clinically significant abnormal laboratory values must be repeated on the appropriate clinical follow-up arranged by the Investigator and documented on the lab report until the lab value has stabilized or has returned to a clinically acceptable range (regardless of relationship to BL-1040). Any laboratory value that remains abnormal at the End of Study visit and is judged to be clinically significant will be followed according to accepted medical standards for up to 30 days or until resolution of the abnormality.

Approximately 15 mL safety blood samples will be collected at the time points indicated in Sec 6.2 and shown in Table 6.1. Analyses will include:

- biochemistry
  - total protein
  - albumin
  - total bilirubin
  - ALT
  - AST
  - GGT
  - LDH
  - alk phosphate
  - glucose
  - sodium
  - potassium
  - calcium
  - phosphate
  - urea/BUN
  - creatinine
  - PTT or ACT
  - troponin I or T (Screening only)

- hematology
  - red blood cell count
  - hemoglobin
  - hematocrit
  - mean cell hemoglobin
  - mean cell hemoglobin concentration
  - mean cell volume
  - white blood cell count and differential
  - platelet count

- cardiac biomarkers
  - Total CK/CK MB
  - NT-proBNP

- urinalysis

  - urine protein
  - urine glucose
  - urine blood
  - leukocytes
  - nitrites
  - urobilinogen
  - bilirubin
  - pH
  - specific gravity
  - ketones

If dipstick analysis reveals any pathological results, a full urine analysis will be conducted and the following should be checked:

1. Color
2. Appearance
3. Leukocytes + erythrocytes per HPF (High Power Field)
4. Squamos epithelial cells
5. Non squamos epithelial cells
6. Yeast in urine
7. Amorphous cells
8. Mucous in urine
9. Casts
10. Crystals

6.3.2 Core laboratories

Results of echocardiograms, ECGs, Holters, angiographies, and MRIs will be evaluated at Biomedical Systems:

Biomedical Systems
1945 Ch. de Wavre
B-1160 Brussels-Belgium
phone: +32 2 661 20 70
fax: +32 2 661 20 71
email: sjacobs@biomedsys.com

NT-proBNP samples will be assessed at the central laboratory at the University of Heidelberg:

Universitatsklinikum Heidelberg
Zentrallabor
Im Neuenheimer Feld 671
69120 Heidelberg, Germany
Tel.: 06221-56-8803
Fax: 06221-56-5205

6.4 Minnesota Living with Heart Failure® questionnaire

The Minnesota Living with Heart Failure® questionnaire (MLHQ) is a standardized and validated questionnaire designed to measure the effects of heart failure and treatments for heart failure on an individual's quality of life (ref. 6-8). The questionnaire measures the effects of symptoms, functional limitations, and psychological distress on the individual's life. These items are measured using a 6 point Likert scale (0-5) to indicate how much each of 21 items has affected their quality of life.

The scales will be administered by the Investigator or trained/designated personnel, in the local language.

7 Adverse and Serious Adverse Events

7.1 Adverse event definition

An adverse event (AE) is any untoward medical occurrence in a clinical trial patient who was administered a medicinal product and/or medical device and which does not necessarily have a causal relationship with this treatment. This includes any noxious, pathological or unintended change in anatomical, physiological or metabolic functions as indicated by physical signs, symptoms and/or laboratory detected changes occurring in any phase of the clinical study whether associated with the study drug/device and whether or not considered related to study intervention. This includes an exacerbation of pre-existing conditions or events, intercurrent illnesses, or drug/device interaction. Anticipated day-to-day fluctuations of pre-existing conditions that do not represent a clinically significant exacerbation need not be considered AEs. Discrete episodes of chronic conditions occurring during a study period should be reported as AEs in order to assess changes in frequency or severity.

AEs should be documented in terms of signs and symptoms observed by the Investigator or reported by the patient at each study visit. A medical diagnosis should be added.

Pre-existing conditions or signs and/or symptoms (including any which are not recognized at study entry but are recognized during the study period) present in a patient prior to the start of the study should be recorded in the Medical History form within the patient's CRF.

7.2 Recording adverse events

All non-serious AEs (serious or non-serious) will be recorded from the time of implantation of BL-1040 on Day 1 until the end of the active study period (Day 180); all serious AEs will be recorded from the time of implantation of BL-1040 until the end of the long term follow-up (Month 60). AEs are to be recorded on the appropriate AE pages in the patient's CRF: if the AE is serious, the appropriate box on the AE page of the CRF should also be ticked. Where possible, a diagnosis rather than a list of symptoms should be recorded. If a diagnosis has not been made then each symptom should be listed individually. The nature, time of onset and cessation, and any treatment provided shall be recorded.

According to "Medical Devices: Post Market Surveillance: Global Guidance for Adverse Event Reporting for Medical Devices — GHTF/SG2/N54R8: 2006, Study Group 2 Final Document'', typical adverse events for medical devices include but are not limited to:

- a malfunction or deterioration in the characteristics or performance
- an incorrect or out of specification test result

- an inaccuracy in the labeling, instructions for use and/or promotional materials. Inaccuracies include omissions and deficiencies. Omissions do not include the absence of information that should generally be known by the intended users.
- use error

All AEs (serious and non-serious) shall be reported as specified in this section of the Protocol and the expanded Medical Device Reporting Guidelines, which will be provided to all investigators prior to the start of the study.

7.3 Pre-device events

The Investigator will report any pre-device event directly observed or mentioned by the patient from the time of signing Informed Consent until the implantation of BL-1040 on Day 1. Pre-device events are reported in the CRF with at least the nature, the start date and the treatment (if applicable).

7.4 General adverse events

Information on any AE must be recorded when volunteered by the patient, observed by study personnel, or elicited by a non-leading question, such as “How are you feeling?”.

7.4.1 Assessment of severity of general adverse events

General events should be assessed according to the following scale:

- mild — the event is easily tolerated and does not interfere with usual activity; disappears without residual effects
- moderate — the event interferes with daily activity, but the patient is still able to function
- severe — the event is incapacitating and the patient is unable to work or complete usual activity; considered as unacceptable by the Investigator

7.4.2 Assessment of causality of adverse events

Every effort should be made by the Investigator to explain each AE, both serious and non-serious, and assess its causal relationship, if any, to implantation of BL-1040.

The relationship of BL-1040 to the event will be determined by how well the event can be understood in terms of one or more of the following

related	there is suspicion of a relationship between BL-1040 and AE (without determining the extent of probability); there are no other more likely causes and administration of BL-1040 is suspected to have contributed to the AE
possible	AE occurs within a reasonable time after the implantation of BL-1040 but can also be reasonably explained by other factors (as mentioned below)
unrelated	there is no suspicion that there is a relationship between BL-1040 and AE, there are other more likely causes and implantation of BL-1040 is not suspected to have contributed to the AE

Non-serious and serious AEs will be evaluated as two distinct types of events given their different medical nature. The Investigator will examine all events assessed as “serious” (Sec. 7.5.1) in order to determine, as far as possible, ALL contributing factors applicable to each serious AE.

Other possible contributors include:

- underlying disease
- Other medication
- protocol required procedure
- other (specify)

7.4.3 Follow-up of adverse events and assessment of outcome

All AEs will be followed to resolution (patient’s health has returned to baseline status or all variables have returned to normal); until an outcome has been reached; stabilization (Investigator does not expect any further improvement of worsening of the event); or the event is otherwise explained, regardless of whether the patient is still participating in the study. Where

Page appropriate, medical tests and examinations will be performed to document resolution of the event. All follow-up information will be recorded in the patient’s CRF until Day 180.

7.5 Serious Adverse Events

7.5.1 Definition of Serious Adverse Event (SAE)

A serious adverse event (SAE) is any untoward medical occurrence or effect that led to one of the following outcomes:

- death of a patient, user or other person
- serious injury of a patient, user or other person
  Serious injury (also known as serious deterioration in state of health) is either:
  - a life threatening illness or injury *

- permanent impairment of a body function or permanent damage to a body structure†
- a condition necessitating medical or surgical intervention to prevent permanent impairment of a body function or permanent damage to a body structure
  The term “permanent” means irreversible impairment or damage to a body structure or function, excluding minor impairment or damage
  Medical intervention is not in itself a serious injury. It is the reason that motivated the medical intervention that should be used to assess the reportability of an event.
- in-patient hospitalization‡ or prolongation of existing hospitalization
- an event that might lead to death or serious injury of a patient, user or other person if the event recurs (sometimes called a ‘‘near incident’’)

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*Life threatening: An AE is life threatening if the patient was at risk of death at the time of the event; it does not refer to an event which hypothetically might have caused death if it were more severe.

†Disabling/incapacitating: An AE is incapacitating or disabling if the event results in a substantial disruption of the patient’s ability to carry out normal life functions. This definition is not intended to include experiences of relatively minor medical significance such as headache, nausea, vomiting, diarrhea, influenza, or accidental trauma (e.g. sprained ankle).

‡Hospitalization: In general, hospitalization signifies that the patient has been detained (usually involving at least an overnight stay) at the hospital or emergency ward for treatment that would not have been appropriate in the physician’s office or out-patient setting.

Hospitalization for either elective surgery related to a pre-existing condition which did not increase in severity or frequency following initiation of the study or for routine clinical procedures¶ (including hospitalization for “social” reasons) that are not the result of an AE need not be considered as AEs and are therefore not SAEs. When in doubt as to whether ‘hospitalization’ occurred or was necessary, the AE should be considered serious.

¶Routine Clinical Procedure: procedure which may take place during the study period and should not interfere with the implantation of BL-1040 or any of the ongoing protocol specific procedures. If anything untoward is reported during an elective procedure, that occurrence must be reported as an AE, either ‘serious’ or non-serious according to the usual criteria.

For medical devices, typical serious adverse events include but are not limited to:

- use error (e.g. untrained user, incorrect route of administration) related to medical devices, which did result in death or serious injury
- damage to tissue or tissue function following administration of study device

- impairment of an organ or organ function following administration of study device
- interaction with concomitant treatment (other devices or drugs) that might lead to death or serious injury
- interaction with materials (e.g. catheters, stent), substances or gases entering into contact with the device during normal use that might lead to death or serious injury
- non-biocompatibility leading to serious irritation/allergy that results in in-patient hospitalization or prolongation of existing hospitalization

7.5.2 Pre-defined SAEs

For the purposes of this study, the following events will be defined as serious:

- re-infarction
- stroke or transient ischemic attack (TIA)
- acute heart failure (decompensation)

The occurrence of any of these events after implantation of BL-1040 will be considered an SAE; they are to be reported and followed up as specified in Sections 7.5.3 and 7.5.4.

7.5.3 Reporting serious adverse events

All Serious Adverse Events (SAEs) must be reported immediately by the Investigator without filtration, whether considered to be associated with BL-1040 and whether or not considered related to BL-1040. The Investigator must report SAEs within one calendar day of becoming aware of the event by telephone, fax or e-mail to the Study Contact for Reporting Serious Adverse Events as indicated below. This initial notification should include minimal, but sufficient information to permit identification of the reporter, the patient, study device, any medications administered, AEs, causality assessment and date of onset. The Investigator should not wait for additional information to fully document the event before providing notification. An acknowledgement letter will confirm the first notification. The report is then to be followed by submission of a completed SAE Report Form provided by Venn Life Sciences AG as soon as possible but at latest within 3 calendar days of the initial telephone/fax or e-mail report detailing relevant aspects of the AEs in question. All actions taken by the Investigator and the outcome of the event must also be reported immediately. For documentation of the SAE, any actions taken, outcome and follow-up reports, the SAE Report Forms are to be used. Where applicable, hospital case records and autopsy reports should be obtained.

Investigators must report SAEs to the appropriate ethics committee if requested by the committee and/or according to local legal requirements.

Study Contact for Reporting Serious Adverse Events.

Venn Life Sciences AG, Elisabethenstrasse 23/3, CH-4051 Basel

Fax:	00800 201 11 011
e-mail:	SAE@vlsworldwide.com
Tel:	+41 61 201 11 83

24/24 hour and 7/7 day availability

7.5.4 Follow-up of serious adverse events

All SAEs must be collected and documented until the end of the long term follow-up (Month 60), and followed up until the event either resolved, subsided, stabilized, disappeared or is otherwise

explained or the study patient is lost to follow-up. All follow-up activities must be reported, if necessary on one or more consecutive SAE report forms, in a timely manner. All fields with additional or changed information must be completed and the report form should be forwarded to the Study Contact for Reporting Serious Adverse Events as soon as possible but latest within 7 calendar days after receipt of the new information. Clinically significant laboratory abnormalities will be followed up until they have returned to normal, or a satisfactory explanation has been provided. Reports relative to the subsequent course of an AE noted for any patient must be submitted to Venn Life Sciences AG.

7.6 Treatment of adverse events

Treatment of any AE is at the sole discretion of the Investigator and according to current available best treatment. The applied measures should be recorded in the CRF of the patient.

7.7 Pregnancy

The Sponsor must be notified immediately of any pregnancy that occurs during the study. The SAE report form should be used to report the pregnancy, even though the pregnancy is not considered an SAE. Women who become pregnant during the study will be followed up until birth of the child. The health status of the newborn will be reported in the patient's CRF.

8 Data Evaluation and Statistics

In all analyses where a change from baseline is performed, baseline is defined as the last available value before device implantation.

8.1 Endpoints

The **primary** endpoints are occurrence of all adverse events including but not limited to:

- all MIs
- cardiovascular hospitalization
- serious ventricular arrhythmias sustained
  - VT (symptomatic or sustained VT [duration longer than 30 seconds or 100 beats, or associated with hemodynamic collapse]
  - VF
  - symptomatic bradycardia, pauses of longer than 3.0 seconds, complete atrioventricular block, Mobitz II atrioventricular block
- symptomatic heart failure (NYHA criteria + physical examination OR hospitalization due to heart failure)
- renal failure
- stroke
- death

Secondary Endpoints include the parameters:

- change from baseline in LV dimensions (end-systolic volume index, end-diastolic volume index, left ventricular mass)
- change from baseline in regional (infarct related) and global wall motion score
- change from baseline in ejection fraction
- cardiac rupture
- NT-proBNP

8.2 Estimated sample size

No formal sample size calculation was performed. Twenty patients followed up to Day 180 were deemed necessary to meet the objectives of this Phase I study. Taking into account drop-outs after the device implantation, thirty patients will be enrolled.

8.3 Planned methods of analysis

All data recorded will be presented in data listings and summary tables, as appropriate. Missing values will not be replaced. No formal hypothesis testing will be performed.

8.3.1 Analysis population

All participants who received the BL-1040 myocardial implant will be included in the safety analysis. Any excluded cases will be documented together with the reason for exclusion. All decisions on exclusions from the analysis will be finalized prior database lock.

8.3.2 Analysis of demographics

Continuous demographic variables (age, height, weight) will be summarized using mean, median, standard deviation, minimum, maximum, and number of available observations.

Qualitative demographic characteristics will be summarized by counts and percentages. Other patient characteristics (medical history, clinical findings, prior medications, inclusion/exclusion criteria) will only be listed.

8.3.3 Analysis of safety

AEs will be described in individual listings and frequency tables by system organ class and preferred terms (MedDRA version 10.0 or higher), regardless of relationship as well as for related AEs. The severity of AEs will also be tabulated.

Vital signs will be listed and changes from baseline and raw results will be summarized by means and standard deviations.

Laboratory test values will be presented by individual listings with flagging of values outside the normal ranges. Raw laboratory results and changes from baseline will be summarized by means and standard deviations.

12 lead ECG findings will be presented by listings and frequency tables, as appropriate. Continuous ECG data will be summarized using standard descriptive statistics.

The change from baseline in cardiac parameter (LV dimensions, wall motion score, ejection fraction) as well as the NT-proBNP data will be summarized using standard descriptive statistics.

8.4 Interim analysis

An interim safety analysis will be performed after 5 patients have completed the Day 30 visit, on all data collected up to this timepoint.

8.5 Final and follow-up reporting

The final clinical study report will be prepared based on data from Day 180, or End of Study, from the final patient. Thereafter, an annual safety report will be prepared after each yearly safety follow-up visit (Months 12, 24, 36, 48, 60).

8.6 Quality assurance

All data collected in the CRF will be double entered into a validated computerized clinical data management system (Clintrial). Laboratory values from the local lab will be entered into the CRF. Analysis of the data will only be performed after all queries have been resolved using an appropriate software for analysis (SAS 8.1).

9 Ethics and regulatory considerations

The study will be conducted according to Good Clinical Practice, the Declaration of Helsinki 2000 (Appendix A), and the rules and regulations of the European Union and Israel.

9.1 Informed Consent

The nature, purpose and potential risk of the study as well as the action of the BL-1040 myocardial implant will be explained to all patients both verbally and in writing. They will be given adequate time to consider the study before signing the consent form. Their questions will be actively encouraged. They will be informed that they may withdraw from the study at any time. This information is documented in the protocol and participants in the study will sign a consent form confirming that they have read and understood it; no study activities will take place until the consent form has been signed. They will also be given a Patient Information Sheet and copy of the consent form.

9.2 Authorities

The procedures laid out by the local regulatory authorities must be followed and all documents must be submitted to all concerned authorities, and where needed, approved before a clinical study may commence.

9.3 Protocol Amendments

There will be no alteration to the protocol without the express written approval of the Sponsor.

The local authorities or ethics committees must approve all major protocol amendments prior to implementation.

No protocol amendments should be adopted without prior written approval from the ethics committee except in the following cases:

- in order to eliminate immediate hazard to the patients,
- changes involving only logistical or administrative aspects of the trial. Then notification to the relevant authorities should be submitted.

In these cases, the implemented deviation or change should be submitted as soon as possible to the relevant authorities for review and approval.

No protocol deviations are anticipated. However, should any protocol deviations occur, the Principal Investigator must report the matter to the Sponsor as soon as reasonably practical. Details of the deviation and, if possible, the reason for its occurrence must be included in the study report.

Major modifications will need further approval, and will be submitted to the local authorities or ethics committees, according to local regulations, in the form of an Amendment. Minor administrative changes require only that the Chairman of the Ethics Committee be informed in writing without delay.

9.4 Patient confidentiality

Individual patient data obtained as a result of this study is considered confidential. A patient identification number will identify any patient data collected throughout the study only.

Data generated as a result of this study are to be available for inspection on request by all authorized Sponsor personnel, Venn Life Sciences AG personnel, audit personnel and regulatory authorities. The Informed Consent must clearly reflect this access.

9.5 Insurance

The compensation of the patient in the event of study related injuries will comply with the applicable obligatory requirements. Details will be included in the informed Consent.

9.6 Duration of the study

The active study phase for each patient is 180 days. Enrolment is expected to begin in Q1 2008; the study is expected to end **Q1 2010.**

10 Data Handling and Record Keeping

10.1 Documentation

Records must be retained for 15 years after study completion

10.2 Case Report Forms

The Investigator is responsible for maintaining adequate and accurate medical records from which accurate information will be transferred into the study database. Case Report Forms (CRFs) should be completed by the Investigator or delegated personnel.

CRFs will be provided for each patient. All data will be entered in black ink. Data/corrections entered will be signed or initialed by the study personnel undertaking that procedure. Overwriting data or use of liquid correcting fluid is not allowed. Detailed instructions are provided with the CRF.

10.3 Monitoring and quality control

To ensure compliance with relevant regulations, data generated by this study must be available for inspection upon request by representatives of BioLine Innovations Jerusalem, Venn Life Sciences AG(CRO), auditing personnel and relevant local regulatory authorities.

Regular on-site visits for monitoring of study activities and data recording will be scheduled. Formal reports of these visits will be generated and copies provided to relevant Sponsor and study personnel.

10.4 Publication policy

The results of the study are the property of the Sponsor. All manuscripts, abstracts or other modes of presentation arising from the results of the study must be reviewed and approved in writing by the Sponsor, in advance of submission. Co-authorship with any Sponsor personnel will be discussed and mutually agreed upon before submission of a manuscript to a publisher.

11 References

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6. Rector TS, Francis GS, Cohn JN. Patients' self-assessment of their congestive heart failure. Part 1 Patient perceived dysfunction and its poor correlation with maximal exercise tests. Heart Failure 1987, Oct/Nov; 192-196.
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Appendix A: Declaration of Helsinki

WORLD MEDICAL ASSOCIATION DECLARATION OF HELSINKI
Ethical Principles
for

Medical Research Involving Human Subjects

Adopted by the 18th WMA General Assembly
Helsinki, Finland, June 1964
and amended by the
29th WMA General Assembly, Tokyo, Japan, October 1975
35th WMA General Assembly, Venice, Italy, October 1983
41st WMA General Assembly, Hong Kong, September 1989
48th WMA General Assembly, Somerset West, Republic of South Africa, October 1996
and the
52nd WMA General Assembly, Edinburgh, Scotland, October 2000

Note of Clarification on Paragraph 29 added by the WMA General Assembly, Washington 2002
Note of Clarification on Paragraph 30 added by the WMA General Assembly, Tokyo 2004

A. INTRODUCTION

1. The World Medical Association has developed the Declaration of Helsinki as a statement of ethical principles to provide guidance to physicians and other participants in medical research involving human subjects. Medical research involving human subjects includes research on identifiable human material or identifiable data.

2. It is the duty of the physician to promote and safeguard the health of the people. The physician's knowledge and conscience are dedicated to the fulfillment of this duty.

3. The Declaration of Geneva of the World Medical Association binds the physician with the words, "The health of my patient will be my first consideration," and the International Code of Medical Ethics declares that, "A physician shall act only in the patient's interest when providing medical care which might have the effect of weakening the physical and mental condition of the patient."

4. Medical progress is based on research which ultimately must rest in part on experimentation involving human subjects.

5. In medical research on human subjects, considerations related to the well-being of the human subject should take precedence over the interests of science and society.

6. The primary purpose of medical research involving human subjects is to improve prophylactic, diagnostic and therapeutic procedures and the understanding of the aetiology and pathogenesis of disease. Even the best proven prophylactic, diagnostic, and

therapeutic methods must continuously be challenged through research for their effectiveness, efficiency, accessibility and quality.

7. In current medical practice and in medical research, most prophylactic, diagnostic and therapeutic procedures involve risks and burdens.

8. Medical research is subject to ethical standards that promote respect for all human beings and protect their health and rights. Some research populations are vulnerable and need special protection. The particular needs of the economically and medically disadvantaged must be recognised. Special attention is also required for those who cannot give or refuse consent for themselves, for those who may be subject to giving consent under duress, for those who will not benefit personally from the research and for those for whom the research is combined with care.

9. Research Investigators should be aware of the ethical, legal and regulatory requirements for research on human subjects in their own countries as well as applicable international requirements. No national ethical, legal or regulatory requirement should be allowed to reduce or eliminate any of the protections for human subjects set forth in this Declaration.

B. BASIC PRINCIPLES FOR ALL MEDICAL RESEARCH

10. It is the duty of the physician in medical research to protect the life, health, privacy, and dignity of the human subject.

11. Medical research involving human subjects must conform to generally accepted scientific principles, be based on a thorough knowledge of the scientific literature, other relevant sources of information, and on adequate laboratory and, where appropriate, animal experimentation.

12. Appropriate caution must be exercised in the conduct of research which may affect the environment, and the welfare of animals used for research must be respected.

13. The design and performance of each experimental procedure involving human subjects should be clearly formulated in an experimental protocol. This protocol should be submitted for consideration, comment, guidance, and where appropriate, approval to a specially appointed ethical review committee, which must be independent of the Investigator, the sponsor or any other kind of undue influence. This independent committee should be in conformity with the laws and regulations of the country in which the research experiment is performed. The committee has the right to monitor ongoing trials. The researcher has the obligation to provide monitoring information to the committee, especially any serious adverse events. The researcher should also submit to the committee, for review, information regarding funding, sponsors, institutional affiliations, other potential conflicts of interest and incentives for subjects.

14. The research protocol should always contain a statement of the ethical considerations involved and should indicate that there is compliance with the principles enunciated in this Declaration.

15. Medical research involving human subjects should be conducted only by scientifically qualified persons and under the supervision of a clinically competent medical person. The responsibility for the human subject must always rest with a medically qualified person and never rest on the subject of the research, even though the subject has given consent.

16. Every medical research project involving human subjects should be preceded by careful assessment of predictable risks and burdens in comparison with foreseeable benefits to the subject or to others. This does not preclude the participation of healthy volunteers in medical research. The design of all studies should be publicly available.

17. Physicians should abstain from engaging in research projects involving human subjects unless they are confident that the risks involved have been adequately assessed and can be satisfactorily managed. Physicians should cease any investigation if the risks are found to outweigh the potential benefits or if there is conclusive proof of positive and beneficial results.

18. Medical research involving human subjects should only be conducted if the importance of the objective outweighs the inherent risks and burdens to the subject. This is especially important when the human subjects are healthy volunteers.

19. Medical research is only justified if there is a reasonable likelihood that the populations in which the research is carried out stand to benefit from the results of the research.

20. The subjects must be volunteers and informed participants in the research project.

21. The right of research subjects to safeguard their integrity must always be respected. Every precaution should be taken to respect the privacy of the subject, the confidentiality of the patient's information and to minimise the impact of the study on the subject's physical and mental integrity and on the personality of the subject.

22. In any research on human beings, each potential subject must be adequately informed of the aims, methods, sources of funding, any possible conflicts of interest, institutional affiliations of the researcher, the anticipated benefits and potential risks of the study and the discomfort it may entail. The subject should be informed of the right to abstain from participation in the study or to withdraw consent to participate at any time without reprisal. After ensuring that the subject has understood the information, the physician should then obtain the subject's freely given informed consent, preferably in writing. If the consent cannot be obtained in writing, the non-written consent must be formally documented and witnessed.

23. When obtaining informed consent for the research project the physician should be particularly cautious if the subject is in a dependent relationship with the physician or may consent under duress. In that case the informed consent should be obtained by a well-informed physician who is not engaged in the investigation and who is completely independent of this relationship.

24. For a research subject who is legally incompetent, physically or mentally incapable of giving consent or is a legally incompetent minor, the Investigator must obtain informed consent from the legally authorised representative in accordance with applicable law. These groups should not be included in research unless the research is necessary to promote the health of the population represented and this research cannot instead be performed on legally competent persons.

25. When a subject deemed legally incompetent, such as a minor child, is able to give assent to decisions about participation in research, the Investigator must obtain that assent in addition to the consent of the legally authorised representative.

26. Research on individuals from whom it is not possible to obtain consent, including proxy or advance consent, should be done only if the physical/mental condition that prevents obtaining informed consent is a necessary characteristic of the research population. The specific reasons for involving research subjects with a condition that renders them unable to give informed consent should be stated in the experimental protocol for consideration and approval of the review committee. The protocol should state that consent to remain in the research should be obtained as soon as possible from the individual or a legally authorised surrogate.

27. Both authors and publishers have ethical obligations. In publication of the results of research, the Investigators are obliged to preserve the accuracy of the results. Negative as well as positive results should be published or otherwise publicly available. Sources of funding, institutional affiliations and any possible conflicts of interest should be declared in

the publication. Reports of experimentation not in accordance with the principles laid down in this Declaration should not be accepted for publication.

C. ADDITIONAL PRINCIPLES FOR MEDICAL RESEARCH COMBINED WITH MEDICAL CARE

28. The physician may combine medical research with medical care, only to the extent that the research is justified by its potential prophylactic, diagnostic or therapeutic value. When medical research is combined with medical care, additional standards apply to protect the patients who are research subjects.

29. The benefits, risks, burdens and effectiveness of a new method should be tested against those of the best current prophylactic, diagnostic, and therapeutic methods. This does not exclude the use of placebo, or no treatment, in studies where no proven prophylactic, diagnostic or therapeutic method exists.

30. At the conclusion of the study, every patient entered into the study should be assured of access to the best proven prophylactic, diagnostic and therapeutic methods identified by the study.

31. The physician should fully inform the patient which aspects of the care are related to the research. The refusal of a patient to participate in a study must never interfere with the patient-physician relationship.

32. In the treatment of a patient, where proven prophylactic, diagnostic and therapeutic methods do not exist or have been ineffective, the physician, with informed consent from the patient, must be free to use unproven or new prophylactic, diagnostic and therapeutic measures, if in the physician's judgement it

offers hope of saving life, re-establishing health or alleviating suffering. Where possible, these measures should be made the object of research, designed to evaluate their safety and efficacy. In all cases, new information should be recorded and, where appropriate, published. The other relevant guidelines of this Declaration should be followed.

§ § §

Appendix B: Minnesota Living with Heart Failure® questionnaire

LIVING WITH HEART FAILURE QUESTIONNAIRE

Instructions for Use

1. Patients should respond to the questionnaire prior to other assessments and interactions that may bias responses. You may tell the patient that you would like to get his or her opinion before doing other medical assessments.

2 Ample, uninterrupted time should be provided for the patient to complete the questionnaire.

3. The following instructions should be given to the patient each time the questionnaire is completed.

   a. Read the introductory paragraph at the top of the questionnaire to the patient.

   b. Read the first question to the patient - “Did your heart failure prevent you from living as you wanted during the past month by causing swelling in your ankles or legs”? Tell the patient. “If you did not have any ankle or leg swelling during the past month you should circle the zero after this question to indicate that swelling was not a problem during the past month”. Explain to the patient that if he or she did have swelling that was caused by a sprained ankle or some other cause that was definitely not related to heart failure he or she should also circle the zero. Tell the patient, “If you are not sure why you had the swelling or think it was related to your heart condition, then rate how much the swelling prevented you from doing things you wanted to do and from feeling the way you would like to feel”. In other words, how bothersome was the swelling? Show the patient how to use the 1 to 5 scale to indicate how much the swelling affected his or her life during the past month - from very little to very much.

4. Let the patient read and respond to the other questions. The entire questionnaire may be read directly to the patient if one is careful not to influence responses by verbal or physical cues.

5. Check to make sure the patient has responded to each question and that there is only one answer clearly marked for each question. If a patient elects not to answer a specific question(s) indicate so on the questionnaire.

6. Score the questionnaire by summating the responses to all 21 questions. In addition, physical (items 2, 3, 4, 5, 6, 7, 12 and 13) and emotional (items 17, 18, 19, 20, and 21) dimensions of the questionnaire have been identified by factor analysis, and may be examined to further characterize the effect of heart failure on a patient’s life.

LIVING WITH HEART FAILURE QUESTIONNAIRE

These questions concern how your heart failure (heart condition) has prevented you from living as you wanted during the last month. The items listed below describe different ways some people are affected. If you are sure an item does not apply to you or is not related to your heart failure then circle 0 (No) and go on to the next item. If an item does apply to you, then circle the number rating how much it prevented you from living as you wanted.

Did your heart failure prevent you from living as you wanted during the last month by:

		No	Very little				Very much
1.	Causing swelling in your ankles, legs. etc.?	0	1	2	3	4	5
2.	Making you sit or lie down to rest during the day?	0	1	2	3	4	5
3.	Making your walking about or climbing stairs difficult?	0	1	2	3	4	5
4.	Making your working around the house or yard difficult?	0	1	2	3	4	5
5.	Making your going places away from home difficult?	0	1	2	3	4	5
6.	Making your sleeping well at night difficult?	0	1	2	3	4	5
7.	Making your relating to or doing things with your friends or family difficult?	0	1	2	3	4	5
8.	Making your working to earn a living difficult?	0	1	2	3	4	5
9.	Making your recreational pastimes, sports or hobbies difficult?	0	1	2	3	4	5
10.	Making your sexual activities difficult?	0	1	2	3	4	5
11.	Making you eat less of the foods you like?	0	1	2	3	4	5
12.	Making you short of breath?	0	1	2	3	4	5
13.	Making you tired, fatigued, or low on energy?	0	1	2	3	4	5
14.	Making you stay in a hospital?	0	1	2	3	4	5
15.	Costing you money for medical care?	0	1	2	3	4	5
16.	Giving you side effects from medications?	0	1	2	3	4	5

17.	Making you feel you are a burden to your family or friends?	0	1	2	3	4	5
18.	Making you feel a loss of self-control in your life?	0	1	2	3	4	5
19.	Making you worry?	0	1	2	3	4	5
20.	Making it difficult for you to concentrate or remember things?	0	1	2	3	4	5
21.	Making you feel depressed'?	0	1	2	3	4	5

SCHEDULE 1.31

DESCRIPTIONS OF OTHER ON-GOING TRIALS

Name of Study	Estimated Duration	Estimated End Date
[**]	[**]	[**]
[**]	[**]	[**]
[**]	[**]	[**]

SCHEDULE 1.35

OUTLINE OF STRUCTURE FOR PIVOTAL CLINICAL TRIAL FOR PRIMARY INDICATION

(see Schedule 3.1)

SCHEDULE 1.42(a)

INDEPENDENT SAFETY MONITORING BOARD CHARTER

Independent Safety Monitoring Board

Charter

For

Bioline Innovations Jerusalem

Protocol No. BL-1040

A Phase I, multi-center, open label study designed to assess the safety and feasibility of the injectable BL-1040 implant to provide scaffolding to infarcted myocardial tissue

APPROVING OFFICIALS

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Moti Gal	Sponsor Contact Person		
Andrea Kempf-Müller, M.D	Drug Safety Officer		

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1. PROTOCOL BL-1040

A Phase I, multi-center, open label study designed to assess the safety and feasibility of the injectable BL-1040 implant to provide scaffolding to infarcted myocardial tissue.

Venn Life Sciences AG has been contracted by Bioline Innovations Jerusalem to provide services as the Contract Research Organization (CRO) for the trial.

2. SCOPE OF THE ISMB CHARTER

The International Independent Safety Monitoring Board (ISMB) was formed to monitor the safety of patients participating in this trial on an ongoing basis.

The ISMB will evaluate quality, accuracy and timeliness of data flow and assure confidentiality of data.

The ISMB will develop stopping rules for the termination of the study prior to the initiation.

Bioline Innovations Jerusalem will forward the charter to Regulatory Authorities, and/or Ethics Committees as necessary.

The objective of the ISMB Charter is to outline the specific purposes and functions of the ISMB. In addition, it describes the procedures for data abstraction and data delivery conventions to and from the ISMB members for review purposes.

3. COMPOSITION OF THE ISMB

The ISMB is composed of three members, three voting members including the Chairman. In addition a bio-statistician will consult the ISMB however will not attend as a voting member. The members are independent physicians in the field of cardiology and a bio-statistician experienced in evaluating safety data from cardiology clinical studies. Prof. Lincoff will serve as Chairman of the ISMB. All ISMB members have been approved by the sponsor, Bioline Innovations Jerusalem.

By signing the ISMB Charter, voting ISMB members verify that they do not have a vested interest in the outcome of the study, nor do they have a financial conflict of interest. ISMB members are not employees of Bioline Innovations Jerusalem have outside employment and will not be involved in patient recruitment or as investigators in the study.

The ISMB members are expected to serve until the study is completed. Should a member resign, the reason and effective date of resignation must be submitted in writing to Bioline Innovations Jerusalem and the ISMB Chairman. A replacement member will be sought by Bioline Innovations Jerusalem in consultation with the ISMB Chairman.

Except for the initial meeting of the ISMB where the background data on BL-1040 and the study design will be discussed by Bioline Innovations Jerusalem's' representatives, Bioline Innovations Jerusalem will not participate in the ISMB meetings unless requested by the ISMB.

ISMB Administration

From Venn Life Sciences AG, the ISMB Coordinator will arrange for the provision of the data and narratives required by the ISMB. Bioline Innovations Jerusalem will provide administrative, logistical and coordinating services to the ISMB.

ISMB Contacts & Consultants

The Chairman will be the representative of the ISMB who will be responsible for timely official communications between the ISMB and Bioline Innovations Jerusalem. The Chairman will provide leadership and oversee that the direction of ISMB meeting operations are in accordance with the ISMB charter.

From the sponsor, Bioline Innovations Jerusalem, an identified representative will serve as the primary contact person for the ISMB. The sponsor primary contact is named on the ISMB charter. This individual is not considered to be a member of the ISMB and will only attend open and final sessions of ISMB Data Review Meetings.

From Venn Life Sciences AG, the ISMB Coordinator will serve as the primary contact person for any questions the ISMB members have regarding the contents of the ISMB Data Reports. This individual is not considered to be a member of the ISMB and will

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only attend open and final sessions of ISMB Data Review Meetings. Additional individuals may also be invited to attend the open and final sessions of the ISMB Data Review meetings, as deemed appropriate.

The ISMB Chairman will ensure that ISMB contacts are ***not*** exposed to the ISMB review of the data until the ISMB has arrived at a conclusion. ISMB contacts may ***not*** be present during closed sessions, when the ISMB Data Report is reviewed, ISMB deliberations are made, ISMB recommendations are discussed and/or ISMB voting procedures are conducted.

4. ISMB ROLE & RESPONSIBILITIES

The ISMB is an independent expert advisory group commissioned and charged with the responsibility of evaluating accumulating data at regular intervals and ensuring the safety of the subjects enrolled in the study by monitoring cumulative safety data collected in the clinical program and providing recommendations to Bioline Innovations Jerusalem based on review of this data. The ISMB will contribute to efficient conduct of the trial by providing a fast review of emerging findings from the study. This ISMB will consist of physicians with expertise in cardiovascular disease, particularly in the area of coronary artery disease and with experience monitoring safety of drugs and/or devices for cardiovascular applications, and will have no participation in the trial in any other capacity.

These reviews in subsets of patients will have the objective of searching for signals of clinically important adverse safety findings that may be indicative of risk to currently enrolled patients as well as increased risk for future patients. In these reviews, the ISMB will assume a conservative approach in assessing safety.

The Chairman will be directly responsible for reporting the outcome of all ISMB meetings and be the primary contact for any emergency meetings, as appropriately convened. He will be a voting member of the ISMB. The Chairman will also be responsible for the preparation of the report and/or recommendations to Bioline Innovations Jerusalem.

The three voting members of the ISMB (along with the Chairman) will be responsible for evaluating the safety data and making recommendations on the continuation of the study as set out in the protocol. They may also make other pertinent safety recommendations for the conduct of the study. They will be guided by the ISMB Biostatistician's evaluation of the data, as required.

The bio-statistician will be involved in conducting any analysis that the ISMB recommends. The Bio-statistician will be responsible for designing and maintaining the safety database that the ISMB will use for its analysis. This database may differ from the database by Venn Life Sciences AG and, as such, is meant only for the use of the ISMB. The database will be created in such a way that it is reproducible and can be audited, if necessary. If the ISMB is considering a recommendation of premature termination of the study, the bio-statistician can contact Venn Life Sciences AG for additional data and/or for the performance of confirmatory analysis. The Bio-statistician can also arrange for the necessary ISMB communications to be documented and stored and only to be released after study completion.

The ISMB will ensure that this study meets the highest standards of patient safety. In their analysis of the data from the patients, the ISMB will be focused on determining if there is a signal of clinically significant pattern of change in safety parameters that may lead to termination of study. This may require the ISMB to perform/request additional data/analyses prior to making a decision.

The operating procedures of the ISMB are based on and are in compliance with guidance and definitions of the International Conference on Harmonization and the Food and Drug Administration. The ISMB will conduct all of its operations under the ICH Good Clinical Practices (GCP).

Specifically, the ISMB is authorized and charged to perform the following functions:

- review 30 day safety data patients from the first 2 sequentially enrolled patients to determine whether 3 additional patients may be enrolled; after reviewing the 30 day safety data from these 3 additional patients, will determine whether the rest of patients may be enrolled
- within 30 days of enrolment of each successive group of 5 patients receiving the device, will review all Serious and Severe Adverse Events occurring to date and will recommend continuation, discontinuation, or modification of the procedure or protocol, based on a determination of whether the occurrence of serious, unexpected, or device-related adverse events (Sec. 7 in protocol) might outweigh the potential benefit achievable with the device
- review emerging findings in patients and identify potential safety concerns with BL-1040
- will receive information, on an expedited basis, on all Serious and Severe Adverse Events, clinically significant laboratory values (as defined in the study safety plan), ECG abnormalities and vital signs that are associated with Serious and Severe Adverse Events, and data from patients who decided to withdraw from the study due to Serious and Severe Adverse Events. All Serious and Severe Adverse Events that occur in the catheter lab during the administration of BL-1040 or the hospitalization period after the procedure should be reviewed

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promptly by the ISMB. The ISMB will review this information and may decide to interrupt, alter, or terminate the trial.
- will adjudicate whether or not an event is unexpected, based on a pre-specified list of expected Serious and Severe Adverse Events as well as clinical judgment within the study population.

All ISMB members will review the safety data provided by the CRO. The members will reach their own individual decision on the relatedness and the potential hazard posed by the event. The ISMB will then collectively discuss the cases. In the event the majority opinion of the Board is that the events do not pose any significant risk then the ISMB will recommend continuing the trial as designed. However, if the Board decides that undue risk could accrue from continuation of the study as designed, the ISMB has the freedom to recommend appropriate changes to the study selection criteria, safety evaluations, etc. In addition, the CRO will provide datasets and listings capturing disposition, AEs, clinically significant Echocardiography, MRI, angiography, Holter, ECG vital signs/laboratory changes, once all patients complete study.

5. VENN LIFE SCIENCES AG ROLE & RESPONSIBILITIES

Venn Life Sciences AG will provide coordinating services for the study. The ISMB Coordinator will provide information, on an expedited basis, on all Serious and Severe Adverse Events, clinically significant laboratory values (as defined in the study safety plan, ECG abnormalities and vital signs that are associated with Serious and Severe Adverse Events as required, to the ISMB members. Venn Life Sciences AG will be charged with the following responsibilities:

- To identify a specific individual to interface with the ISMB.
- To provide all required information in advance of the meeting in a mutually agreeable format approved at the initial meeting of the ISMB.
- To provide a standard safety narrative for all patients who withdraw from the study due to Serious or Severe Adverse Events.
- To provide specific meeting issues in advance of the meeting.
- To keep the ISMB Chairman informed of any serious safety issues as the study progresses
- To inform each principal investigator of the ISMB recommendations, as required.
- To notify Bioline Innovations Jerusalem of any issues related to the ISMB which might negatively influence the study.

6. BIOLINE INNOVATIONS JERUSALEM'S RESPONSIBILITIES

Bioline Innovations Jerusalem will be responsible for the following:

- To make any necessary changes to the protocol recommended by the ISMB and approved by Bioline Innovations Jerusalem.

· To ensure that the ISMB is operating as needed for the purpose of the study.

7. ONGOING COMMUNICATIONS & NOTIFICATIONS

The ISMB Chairman will receive relevant information regarding serious adverse events and Early Terminations on an ongoing basis. The ISMB Chairman will determine whether further distribution of this material to the remaining voting ISMB members is necessary.

8. DATA REVIEW MEETINGS

ISMB Data Review meetings will be held in person or through teleconferences based on the volume of data to be reviewed. The ISMB Coordinator will establish the agenda for each ISMB Data Review meeting, with input from Bioline Innovations Jerusalem and the ISMB Chairman.

It is expected that there will be one initiation and at least three scheduled ISMB Data Review meetings. The initiation meeting will be held via face-to-face format, while the Data Review Meetings may be held via teleconference.

The first 2 patients will be sequentially enrolled into the study. After the 1st patient has completed Day 30 assessments, the Independent Safety Monitoring Board (ISMB, Sec. 4.3) will review the patient's data through Day 30 (first ISMB meeting). The ISMB will then decide whether to give approval to enroll the 2nd patient. After the 2nd patient has completed Day 30 assessments, the ISMB will again review the data and provide approval for enrollment of the next 3 patients (2nd ISMB meeting). After all 3 patients have completed Day 30 assessments, the ISMB will review the data from these patients and provide approval for opening enrollment to the rest of the patients (3rd meeting)

The ISMB may also elect to hold ad hoc meetings outside of the scheduled dates, if deemed necessary. For instance, as the ISMB Chairman will receive information regarding reported serious adverse events on a regular basis, ad-hoc ISMB meetings may also be held on a triggered basis (e.g. in response to a high number of safety events).

Voting

Input must be obtained from all three ISMB members, for voting purposes. The ISMB will strive for a consensus opinion regarding the data reviewed. If ISMB consensus is not possible, a majority vote will be required, to determine the final ISMB recommendation. If the ISMB vote does not result in a clear majority, the ISMB Chairman will assemble and present majority and dissenting opinions for all recommendations considered.

Meeting Minutes

ISMB Data Review meeting minutes will be divided by session and will reflect the attendance of voting ISMB members, the ISMB Coordinator, ISMB contacts and consultants and other individuals, as well as whether each individual attended in person or via teleconference.

Since all details of ISMB deliberations must be kept strictly confidential among members of the ISMB, portions of the ISMB Data Review meeting minutes must remain confidential until the completion of the final study analysis.

The ISMB Chairman will file all minutes from all sessions, centrally. Once the final study analysis is complete, the ISMB Chairman will forward the central file of all ISMB minutes for all sessions to Bioline Innovations Jerusalem for appropriate filing.

9. RECORDS RETENTION

The ISMB Chairman should maintain a record of all ISMB minutes until the investigation of the study device is discontinued. After this period, the ISMB Chairman will forward to the sponsor all records to the sponsor to determine if further retention and/or archiving is necessary.

Data Source and Content

10. ISMB COMMUNICATION OF FINAL CONCLUSIONS

The ISMB Chairman will contact Bioline Innovations Jerusalem within two working days after an ISMB meeting (via facsimile or telephone) to notify them of recommendations forthcoming from that meeting. Bioline Innovations Jerusalem will act upon these recommendations as appropriate, i.e., the final decision will rest with Bioline Innovations Jerusalem. Bioline Innovations Jerusalem's VP of Medical Affairs or designee will notify the project team and the CRO of the ISMB recommendations.

Bioline Innovations Jerusalem's VP of Medical Affairs will also write a memo to the files documenting the recommendations of the ISMB and convey to all investigators the decision to continue/discontinue the study.

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11. IMPLEMENTATION OF THE ISMB RECOMMENDATIONS

The decision to implement the recommendations of the ISMB will be made by Bioline Innovations Jerusalem. Bioline Innovations Jerusalem will notify the ISMB of the actual action taken, in response to all recommendations.

If the ISMB recommends early study termination or protocol modification and such action is not accepted or implemented, Bioline Innovations Jerusalem will address this decision with the ISMB in writing.

12. CONFIDENTIALITY

The ISMB will maintain a strictly confidential relationship to the study data. The ISMB will only reveal specific details and information associated with ISMB data review to appropriate parties, as specified by this ISMB Charter.

SCHEDULE 2.3

EXISTING PRODUCT AGREEMENTS

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SCHEDULE 3.1

INITIAL DEVELOPMENT PLAN

Project Boston Clinical Development Plan

Objective

This product is a unique concept, and will require a unique and sophisticated development plan to satisfy all stakeholders.

This product has been given a regulatory designation as a device (rather than drug). The objective of this development plan is to leverage that designation for a rapid and efficient regulatory approval, while providing adequate evidence for safety within the intended patient population.

Strategy

The strategy is to complete a minimal additional amount of preclinical safety in parallel with the clinical development program. [**].

The filing will be based on a [**]. We note that the current phase 2 study has no control group, and can give only general information about safety and tolerability, and no real information on efficacy in humans. For this reason the [**] will be designed with a 'vanguard' cohort of approximately [**] patients. Once the vanguard has completed 6 months of follow up, and interim analysis will be performed, assessing the study for 1) safety, 2) efficacy or futility and 3) performance of the endpoint. Specific, detailed and comprehensive criteria will be established to allow for stopping or continuation, or adjustments in sample size or inclusion criteria. The rules for the interim analysis will be agreed with regulatory authorities in advance of any unblinding, and appropriate adjustments will be made for type 1 error.

Following the interim analysis the number of participating centers will be increased to speed enrollment, and the study will continue to completion.

Endpoint and sample size

We will define [**] and then power the study to show at least a [**] with BL-1040 compared to placebo. This difference is clinically meaningful.

To give maximum power we want to define an endpoint that has a [**] after treatment, which would be reduced to [**]. We will design a [**] that ensures an event rate that is [**] in the control arm.

Failure could include [**] Any one of these events and the patient is [**]; none of these events and the patient is considered [**]. It is possible that other clinically relevant events may be added to the composite.

Next we will estimate how often each of these events will happen. [**].

Control Group Event Rate	Treatment Group Event Rate	Sample size per arm 90% power and type 1 error < 5%	Total
[**]	[**]	[**]	[**]
[**]	[**]	[**]	[**]
[**]	[**]	[**]	[**]

Although not required under device approval regulations, approximately [**] patients would be desirable for a safety database. If we assume that the placebo event rate will be approximately [**], we would estimate the sample size of the pivotal study to be approximately [**] patients, including the [**] patients in the vanguard cohort.

Budget

	2009	2010	2011	2012	2013	2014	2015	2016	TOTAL
[**]	[**]	[**]	[**]	[**]	[**]	[**]			
[**]						[**]			
[**]							[**]		
[**]							[**]	[**]	
[**]								[**]	
TOTAL	[**]	[**]	[**]	[**]	[**]	[**]	[**]	[**]	[**]

Phase III Study

Budget will assume [**] of [**] patients, with a primary endpoint at [**] major adverse cardiac outcomes at [**], and a safety follow up annually for [**].

Clinical:

Monitoring:		[**]
Per Patient total:	[**]	[**]
Pre Clinical	[**]	[**]
Total		[**]

Given that 15-20% of the total clinical costs are committed before the first patient is enrolled, we estimate that cost to decision point is approximately [**]. It may be possible to reduce cost to the

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decision point by [**], trading off for time-to-launch. This alternative scenario has not been modeled.

Cost by Year ($M)

[**]	[**]	[**]	[**]	[**]	[**]
[**]	[**]	[**]	[**]	[**]	[**]

[**] **Study**

Budget will assume a [**] (including ethnicity) of [**] patients. Study will start in [**] and end [**]

Cost by Year ($M)

[**]	[**]	[**]	[**]	[**]	[**]	[**]
[**]	[**]	[**]	[**]	[**]	[**]	[**]

Timeline

Phase III Study

Enrollment w/ [**] per site per month	**Part 1**	**Part 2**
Total Enrollment	[**]	[**]
Active Sites	[**]	[**]
Enrollment/Site/Month (on average)	[**]	[**]
Monthly Study Enrollment	[**]	[**]
Time to Enroll Patient per Part (months)	[**]	[**]
TOTAL ENROLLMENT TIME (months)	[**]	

Trial Task	**End Date**
Initiate Project	[**]
FPI	[**]
[**]	[**]
LPI	[**]
DB Lock	[**]
CSR	[**]
Submit PMA	[**]

Probability of success

Based on the available preclinical data it is not possible to come to a firm estimate of POS at this time. However, there is evidence of efficacy in preclinical models, and a consensus among experts that the mechanism is plausible. Given the existing data on the prior use of this class of compounds in humans, the likelihood of adequate safety and tolerability seems higher than would otherwise be possible at this stage, and given the device designation, the probability of clinical and regulatory success is likewise higher than it might otherwise be. Assuming the likelihood of adequate safety at [**] and the likelihood of adequate efficacy at [**], the overall POS to filing is in the range of [**].

SCHEDULE 3.7

PRELIMINARY COMMERCIALIZATION PLAN

Preface:

This document is prepared for the management of BioLineRx as a basis for discussion only, and is intended to be indicative of Ikaria's current intent with respect to global commercialization of BL-1040. Actual launch plans will continue to evolve over time, in accordance with the evolution of market dynamics, the global environment for cardiovascular drugs and devices, and the emerging product profile of BL-1040.

I. Situation Analysis

a. Unmet Medical Need

Each year cardiovascular disease (CVD) causes over 4.3 million deaths in Europe. CVD is estimated to cost the European Union (EU) economy €192 billion a year. The main forms of CVD are coronary heart disease (CHD) and stroke. Just under half of all deaths from CVD are from CHD. CV is also a large problem in Japan, and is emerging as a public health issue even in the developing countries.

Each year smoking kills over 1.2 million people in Europe (450,000 from CVD)). Dietary patterns across Europe are playing an increasing role in CVD. Levels of physical inactivity are high in many European countries and levels of obesity are increasing across Europe in both adults and children. Over 48 million adults in Europe have diabetes and the prevalence is increasing.

Estimates for population and cardiovascular statistics are presented in Table 1

Table 1

Country	Population (000,000)	Est. Annual non-fatal MI (000)	Interventional Cardiologist	Annual PCI Procedures
[**]	10.4	34.7	230	28
[**]	5.5	18.3	85	15
[**]	5.3	17.7	80	14
[**]	64.4	214.7	1,772	172
[**]	82.3	274.3	1,500	219
[**]	16.7	55.7	266	45
[**]	0.3	1.0	14	1
[**]	58.1	193.7	1,879	155
[**]	40.5	135.0	730	108
[**]	7.6	25.3	124	20
[**]	61.1	203.7	1,000	163
Total Europe	**352.2**	**1,174.0**	**7,682**	**939**
[**]	127.0	423.3	2,500	339
[**]	21	70	373	56
Grand Total	**479.2**	**1,597.3**	**10,182**	**1,278**

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b. Product

BL-1040, a novel, injectable, biodegradable polymer designed to be used in conjunction with Percutaneous coronary intervention (PCI) to provide mechanical scaffolding and reduce the risk of structural remodeling and heart failure in post-myocardial infarction (post-MI) patients, is currently in development and could be on the market as early as [**]. If successful, BL-1040 could be a breakthrough in the management of patients with cardiovascular disease and could represent a large commercial opportunity for Ikaria and **BioLineRx**.

c. Assessment of current level of CV practice

There is significant variability around the medical management of CHD across Europe. Theses groupings give a high level overview of the most common interventions:

Hospital admissions

Rates of admission for CVD vary considerably across Europe. In general, higher admission rates are found in Eastern European and Scandinavian countries. Similar geographical trends are seen for CHD.

Coronary revascularization and other procedures for CVD

While rates of revascularization vary widely across Europe, all countries have seen rates increase significantly since the 1990s. For example, since 1990 rates of PCI have increased fifteen-fold in Italy and twelve-fold in Finland. We expect that advances in medical technique and continued development of medical infrastructure around the world will drive continued growth in the coronary revascularization market.

Drugs

The use of drugs for secondary prevention in CHD patients varies considerably across populations, except in the case of anti-platelet drugs. Over 80% of patients took this form of drug (mostly aspirin). The use of beta blockers, lipid-lowering drugs and ACE inhibitors varies throughout the EU.

d. Pricing and reimbursement environment

The global market for cardiovascular drugs and devices is highly variable in terms of pricing and reimbursement climates.

Pricing

Pricing in the developed markets of western Europe tends to be similar to U.S. pricing, although prices can vary significantly by market, with Northern European markets having higher prices than southern European markets. By contrast, pricing in less developed markets (Eastern Europe, Latin America and the Far East) is highly variable, and will require careful study to ensure an appropriate price is selected in order to maximize penetration and profitability. A clear target product profile will be critical to assessment of pricing strategy in all markets.

Reference pricing is common practice in Europe, so timing of local launches must be carefully coordinated to ensure optimized pricing across the territory.

[**]

Reimbursement

With the exception of regulatory approval, reimbursement will be the single most important driver of commercial success.

The process by which products gain reimbursement can vary greatly from country to country, and may take a considerable amount of time. A recent study by IMS suggested that it was common for newly approved drugs to take between one and three years to gain widespread reimbursement coverage in the top 16 EU markets. Because most European countries operate centralized, government-financed health systems, it is not typical for patients to pay for treatments privately. In many countries where there is virtually no habit of citizens paying for their own healthcare, initiating selling activity without reimbursement would be virtually impossible, while inhabitants of some other countries may have no problem paying for healthcare out of their own disposable income.

Expected timing of reimbursement will, therefore, be a major driver of the timetable for building out sales infrastructure, and commencing selling activities. Ikaria will conduct extensive research between deal closing and launch to ensure that reimbursement conditions are clearly understood and that plans are in place to ensure broad and favorable access to major commercial markets.

II. Commercialization Plan

Product Positioning Strategy

Given the current expectations of the product profile, we aspire to — and expect that — BL-1040 will be positioned as the de facto standard for prevention of post-MI remodeling.

While this depends on the specific results of the clinical trials, the market conditions, including competitive scenario, and prevailing clinical practice standards, the goal will be to make BL-1040 use prevalent across a range of patient sub-groups that are at risk for remodeling. Specifically, the following patient groups will be addressed in the marketing plan:

- High-risk STEMI (includes patients with large myocardial Infarctions (MIs), anterior wall MIs and long lead time to PCI): [**]
- Other STEMI (includes all STEMI patients not considered of the highest risk): [**]
- NSTEMI (all patients who have an NSTEMI): [**]

In addition to the market development efforts listed above, the focus of marketing strategy will be on creating broad awareness of the significant long-term effects of remodeling as well as discussing the risks of myocardial damage and resulting negative consequences for all patients with MIs. In Europe, this will also require resetting of the current paradigm of treating non-primary PCI patients with medical therapy alone, and illustrating the benefits of treatment with a mechanical scaffolding device such as BL-1040.

Organization Size and Structure

As an experienced critical care company, Ikaria is committed to providing doctors and other medical professionals with a high level of customer service. Operating in a highly specialized, life-or-death environment Ikaria strives to match our customers own urgency and commitment to patient care.

To be successful in the area of post-MI care we anticipate creating an organization capable of delivering both the commercial and medical support desired by our target customer base. Ikaria intends to establish itself as the leader in critical care globally, and will use BL-1040 as the platform on which to establish its international presence. As such, we intend to build a robust but flexible organization with all the competencies necessary to achieve leadership of the field. Although BL-1040 will likely be Ikaria's first global product, we anticipate that our own internal pipeline candidates IK-1001 and Covox will not be far behind. The infrastructure envisioned by Ikaria and described in this document will therefore be sufficient to successfully commercialize all of Ikaria's present and future pipeline compounds.

Ikaria proposed to use a "hub and spoke" approach to commercializing BL-1040 in Europe—the "hub" being a European headquarters and the "spokes" representing local operating companies (LOCs) in major markets. The headquarters will provide overall strategic leadership and will spearhead European product development and commercial strategy, while local operating companies will be responsible for selling activity and local tactic implementation.

In addition to strategic marketing and leadership support, the European headquarters will be responsible for financial management and reporting of regional results, management of European regulatory affairs functions, development of a European clinical development program, development of effective key opinion leadership, development of compelling health economic data and development of HR strategies to maintain a strong and vibrant European organization.

The primary role of LOCs is to provide the necessary local sales and marketing efforts necessary to achieve financial objectives for BL-1040. In addition to the necessary commercial infrastructure, the local operating companies would also be staffed with the support functions essential to commercial success. This would include a small local finance team, medical affairs, regulatory affairs and human resource functions. The role of the local support staff is to implement strategic initiatives conceived at headquarters level, and support local initiatives as necessary. The medical affairs staff will be particularly important in supporting marketing in disseminating the full medical information on BL-1040 and the clinical specialists will also lead the training of physicians in using this product appropriately.

The LOC staffing level will be determined as a function of country population, disease prevalence and target doctor population. Sales Representatives will be recruited from companies with a depth of experience in cardiovascular drug and device sales to ensure we gain rapid

access to the necessary prescriber base. Representatives will be compensated through a blend of base salary and sales incentive bonus, according to Ikaria's existing sales force incentive plan. (See Table 2)

Table 2

Country	Population (000,000)	Est. Annual non-fatal MI (000)	Interventional Cardiologist	Annual PCI Procedures (000)	Sales Reps
[**]	10.4	34.7	230	28	[**]
[**]	5.5	18.3	85	15	[**]
[**]	5.3	17.7	80	14	[**]
[**]	64.4	214.7	1,772	172	[**]
[**]	82.3	274.3	1,500	219	[**]
[**]	16.7	55.7	266	45	[**]
[**]	0.3	1.0	14	1	[**]
[**]	58.1	193.7	1,879	155	[**]
[**]	40.5	135.0	730	108	[**]
[**]	7.6	25.3	124	20	[**]
[**]	61.1	203.7	1,000	163	[**]
Total Europe	**352.2**	**1,174.0**	**7,682**	**939**	[**]
[**]	127.0	423.3	2,500	339	[**]
[**]	21.0	70.0	373	56	[**]
Grand Total	**479.2**	**1,597.3**	**10,182**	**1,278**	[**]

NB: The number of sales reps anticipated to be needed in each market has been estimated as a function of [**]

Launch Timelines

To maximize the value of BL-1040 Ikaria intends to be ready to launch at the earliest possible opportunity. As described above, a key driver of launch readiness in any given market will be the ability to access reimbursement for BL-1040. Without appropriate reimbursement in place, attempting to launch BL-1040 would be at best un-productive, and at worst, damaging to the long-term perception of the product.

Ikaria proposes to immediately undertake a battery of research and analysis to understand the market-specific reimbursement environments across major target markets. Results of this research would guide future launch plans, and help inform the timing of key investments in people and infrastructure.

Development of Ikaria's ex-US presence will occur differently throughout the world:

1) Ikaria already has management structures in place in Canada, Japan and Australia. These budding organizations would be expanded in the near term to allow essential market preparation activities to begin as soon as possible. As the product profile of BL-1040 becomes clearer, and the expectations for launch timing crystallize, this existing in-country leadership infrastructure will be expanded to include all the local sales and medical affairs capability necessary to a successful launch.

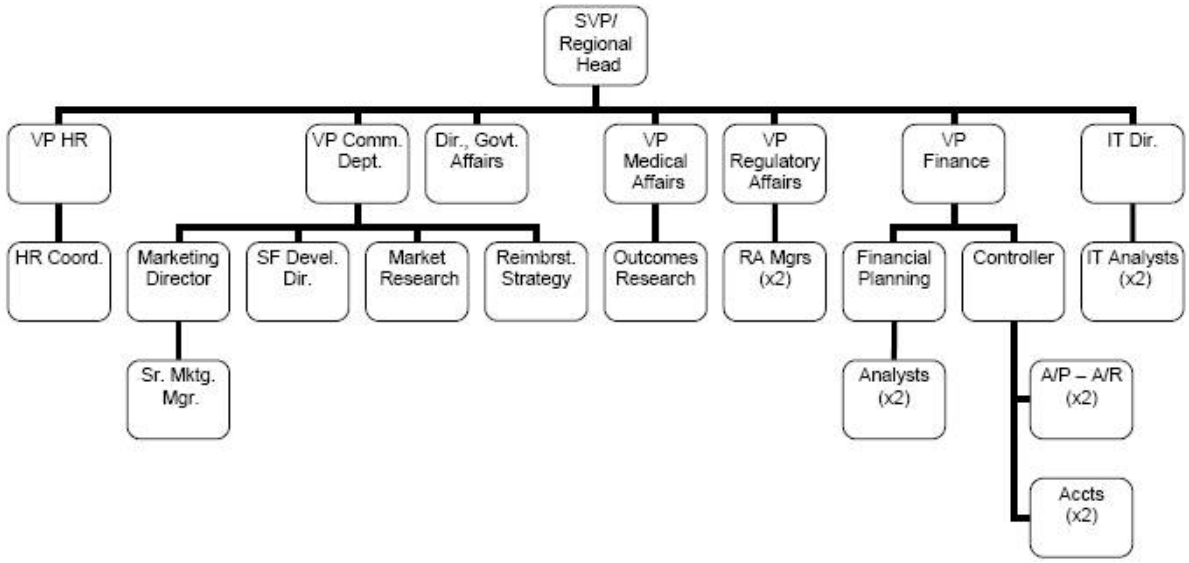
2) Establishment of a European Headquarters function would be a high priority. We anticipate filling key leadership positions as early as [**], so that high-level reimbursement, medical affairs and commercial strategic planning can commence. As a clearer view of the likely launch timeline for BL-1040 emerges, remaining HQ infrastructure will be built out to ensure a fully operational European headquarters well in advance of launch. In the event that a positive result emerges from the interim analysis and a decision is made to move up the commercial launch of the product, the development of the launch plans — including execution of reimbursement strategy and creation of marketing materials — will occur in parallel to the ramp up of the LOCs.

3) Additional, 2nd-tier markets will be evaluated in parallel with [**] commercial infrastructure development. Ikaria believes that there will be great potential for BL-1040 in markets such as [**], but will need more time to evaluate the optimal way to maximize sales in those territories.

[**]

Proposed European Structure

Headquarters



Human Resources

Human Resources will oversee European benefits programs, ensure compliance with local employment law, promote employee development and succession planning, and all functions necessary to building a world-class critical care business in Europe. The European HQ team will work closely with LOC country managers to ensure local employee needs are met and compliance with local laws is maintained. Local in-country contractors may be employed to deliver HR services at the local level.

Anticipated headcount: 2

Government Affairs

Appropriate reimbursement will be critical to the success of BL-1040. As described above, reimbursement can be highly variable across Europe. Development of a skilled government affairs capability within Ikaria Europe will be critical to our success, for BL-1040 as well as future Ikaria pipeline products.

Anticipated headcount: 1

Commercial Development

The European Commercial Development team is responsible for commercial strategy formulation across the European area, including both product and sales force strategy. The HQ marketing team will work closely with the Clinton, NJ-based marketing team to develop a cohesive global strategy suitable for implementation in European markets. The European team will have responsibility to ensure that brand strategies are implemented consistently across the area, and will perform market research to monitor performance and adjust strategy as appropriate. The team will also work in concert with country GMs and local marketing management to implement large-scale promotional and educations programs.

The European HQ team will also develop and implement European sales force strategies including development and maintenance of a customer relationship management system, sales skills training programs, and sales leadership development. The HQ team will work closely with LOC commercial management to ensure a top-class sales effort in each country.

Anticipated headcount: 5

Medical Affairs

Development of a strong base of key opinion leaders will be critical to the success of BL-1040. Cardiology is a fast moving, highly technical field, and for Ikaria to be a credible player we will need to make a significant commitment to supporting the medical community through education, research support, etc.

The European Medical Affairs team will take the lead in formulating strategy for the engagement of key opinion leaders in the formulation of brand development strategy, the development of brand champions and building high-level relationships between Ikaria and the medical community. The HQ Medical Affairs team will work closely with LOC Medical Affairs teams to align strategy across Europe and ensure a consistent medical approach.

The HQ Medical Affairs team will also be responsible for development of health outcome data to support cost-effectiveness arguments. The HQ team will work closely with LOC commercial teams to package health outcome data for effective presentation to in-country prescribers and reimbursement decision makers.

The HQ Medical Affairs team will also take responsibility for developing responses to requests for medical information about Ikaria products. The team will work with LOC Commercial and Medical Affairs teams to ensure a high level of customer support and satisfaction.

Anticipated headcount: 3

Regulatory Affairs

The European Regulatory Affairs (RA) team will lead all regulatory efforts on behalf of Ikaria's European operations. The HQ RA team will work closely with the Medical Affairs team to ensure development programs have maximal likelihood of success and that regulatory compliance is maintained at all times. The RA team will work in concert with in-country RA teams to execute on regulatory strategies and maintain product registrations with local authorities.

Anticipated headcount: 2

Finance

The European Finance team will support all local operating companies with financial reporting and planning functions as well as accounts payable and accounts receivable activities. The HQ team will consolidate European results and maintain a full European operating P&L. The HQ team will perform most of the finance functions on behalf of the European Area, with LOCs having minimal local requirement for finance headcount.

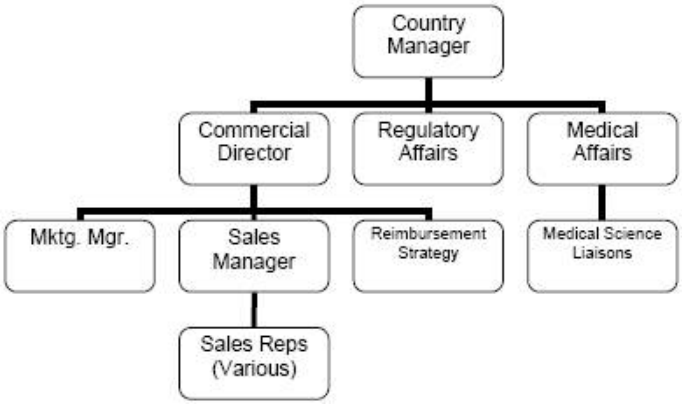
Anticipated headcount: 9

Information Technology

Ikaria's European IT requirements will be delivered by the European HQ team, with local support from 3rd-party contract services. The HQ team will liaise with Ikaria's corporate headquarters IT function in Clinton, NJ to ensure reliable systems functionality and robust customer support.

Anticipated headcount: 3

Local Operating Country (LOC) Structure



Human Resources

Human Resources support will be provided from HQ as described above. Specific local needs will be coordinated with HQ HR and delivered by local 3rd party providers

Anticipated headcount: None

Commercial Development

The LOC Commercial Development team is responsible for implementation of commercial strategy at the local level. The marketing team is responsible for implementation of European product strategy and for directing local tactical marketing in support of BL-1040. The LOC commercial director is also responsible for the development of a skilled critical care sales organization, including recruitment, training and management of reps and managers.

The number of sales reps required to promote BL-1040 will vary from country to country according to the market opportunity, the number of prescribing doctors, and the incidence of PCI procedures. (See Appendix A)

Anticipated headcount: Various

Medical Affairs

Maintenance of a strong relationships and robust medical affairs response capability will be essential for success at the local level. The LOC medical director will take responsibility for development of strong local relationships, coordination of company response to medical information requests. Clinical Specialists in each LOC will be responsible for training of physicians on use of product and for customer service.

Anticipated headcount: 1-2

Regulatory Affairs (RA)

The LOC RA team will work together with HQ RA teams to execute on regulatory strategies and maintain product registrations with local authorities.

Anticipated headcount: 1-2

Finance

The HQ team will perform most of the finance functions on behalf of the European Area, with LOCs having minimal local requirement for finance headcount.

Anticipated headcount: None

Information Technology

Ikaria's European IT requirements will be delivered by the European HQ team, with local support from 3rd-party contract services.

Anticipated headcount: None

SCHEDULE 4.3(a)

BIOLINERX WIRE TRANSFER INFORMATION

Bank Name: [**]

Bank Address: [**]

SWIFT Number: [**]

IBAN Number: [**]

Account Number: [**]

Account Name: [**]

EXHIBIT A

TECHNOLOGY EXCHANGE PLAN

Upon Ikaria's request, the following will be provided by BioLineRx to Ikaria or its designee:

7. All materials (original or copies as appropriate) in BioLineRx's possession and Control relating to Product, including documentation relating to Development and all regulatory filings, clinical information, and data and other documents relating to the On-Going Phase I/II Trial and the Other On-Going Trials.

8. Copies of all documents and available information in BioLineRx's possession and Control necessary for Manufacturing of Product at the time of technology exchange. These documents will include information necessary to assist Ikaria or its designee in setting up Manufacturing operations for such things as:

- raw material test methods, specifications, qualification and justification for use
- raw material vendor lists with part numbers
- analytical methods stated purpose, development, qualification and validation reports
- process development reports, laboratory notebooks and associated electronically stored data
- Manufacturing summary including
  - detailed process description with process schematics, operating parameters and target ranges, flow charts outlining critical process controls and steps, cartoons, verbal description including abbreviations, process scale, yield, and standard process instructions
  - in-process controls/tests and acceptance criteria including stated purpose of in-process tests
  - master batch record(s)
  - filling/packaging process
  - aseptic and process development and validation documents
  - facility and equipment requirements and design documents
  - descriptions of process equipment, including suppliers, part numbers, and historic invoices
  - product test methods, specifications and justification of specifications
  - product stability, test methods and qualification/validation reports, stability reports, shelf life recommendations

As available and agreed upon by the JDC at the time of a technology exchange, BioLineRx will provide requested technical manufacturing or engineering advice to Ikaria or its designee. Ikaria will ensure designee has necessary expertise in place to exchange the documentation and expertise in an orderly fashion.

EXHIBIT B

BIOLINERX PATENT RIGHTS

Family 1

INJECTABLE CROSS-LINKED POLYMER PREPARATIONS AND USES THEREOF

Country	Earliest Priority	Entry Date	Filing Date Application No.	Issue Date Patent No.	Status	Owner
[**]	[**]		[**]		[**]	[**]
[**]	[**]	[**]	[**]		[**]	[**]

[**]	[**]	[**]	[**]		[**]	[**]
[**]	[**]	[**]	[**]		[**]	[**]
[**]	[**]	[**]	[**]		[**]	[**]
[**]	[**]	[**]	[**]	[**]	[**]	[**]
[**]	[**]	[**]	[**]	[**]	[**]	[**]
[**]	[**]	[**]	[**]	[**]	[**]	[**]
[**]	[**]	[**]	[**]		[**]	[**]
[**]	[**]	[**]	[**]		[**]	[**]
[**]	[**]		[**]	[**]	[**]	[**]

Family 2

A METHOD OF TREATING MUSCLE TISSUES

Country	Earliest Priority	Entry Date	Filing Date Application No.	Issue Date Patent No.	Status	Owner
[**]	[**]		[**]	[**]	[**]	[**]

PAYMENT DATE EXTENSION AMENDMENT

Ikaria Development Subsidiary One LLC, a Delaware limited liability company having a principal place of business at 6 State Route 173, Clinton, NJ 08809, USA ("Ikaria"), BioLineRx Ltd., a corporation organized and existing under the laws of the State of Israel and having a principal place of business at 19 Hartum Street, P.O. Box 45158, Jerusalem 91450, Israel ("BioLineRx Ltd."), and BioLine Innovations Jerusalem L.P., a limited partnership organized and existing under the laws of the State of Israel and having a principal place of business at 19 Hartum Street, P.O. Box 45158, Jerusalem 91450, Israel ("BioLine Innovations"; together with BioLineRx Ltd., "BioLineRx") are party to an Amended and Restated License and Commercialization Agreement dated as of the 26th day of August, 2009 (the "Agreement"). Any defined terms used herein shall have them meaning ascribed thereto in the Agreement.

Pursuant to Section 4.1(a) the Agreement, Ikaria is required to make a milestone payment to BioLineRx of USD $10,000,000 upon the Successful Completion of the On-Going Phase I/II Trial (the "Second Milestone Payment") on or before [**]. BioLine and Ikaria are currently in discussions to determine whether Ikaria is required to withhold United States federal income taxes from the Second Milestone Payment. In order to enable the parties to complete those discussions, Ikaria and BioLine hereby agree that the due date for the Second Milestone Payment is hereby extended to [**].

Sections 10.2 ("*Governing Law*") and 10.3 ("*Submission to Jurisdiction*") of the Agreement are hereby incorporated herein by reference.

Acknowledged, Agreed, and Confirmed

/s/ Daniel Tassé
Daniel Tassé
Chief Executive Officer
Ikaria Development Subsidiary One LLC

/s/ Kinneret Savitsky
Kinneret Savitsky,
Chief Executive Officer
On behalf of, and as authorized representative of, both BioLineRx Ltd. and BioLine Innovations Jerusalem L.P.

AMENDMENT TO THE AMENDED AND RESTATED LICENSE AND COMMERCIALIZATION AGREEMENT

This Amendment (this "Amendment") is entered into this 21st day of April 2010 (the "Amendment Effective Date") by and between **Ikaria Development Subsidiary One LLC**, a Delaware limited liability company with a place of business at 6 Route 173, Clinton, NJ, 08809 USA ("Ikaria"), and **BiolineRx Ltd.**, a corporation organized and existing under the laws of the State of Israel and having a principal place of business at 19 Hartum Street, P.O. Box 45158, Jerusalem 91450, Israel ("BioLineRx Ltd."), and **BioLine Innovations Jerusalem L.P.**, a limited partnership organized and existing under the laws of the State of Israel and having a principal place of business at 19 Hartum Street, P.O. Box 45158 Jerusalem 91450, Israel ("BioLine Innovations"; together with BioLineRx Ltd., "BioLine Rx") . This Amendment amends the Amended and Restated License and Commercialization Agreement entered into by and between Ikaria and BioLineRx dated as of the 26th day of August 2009 (the "Agreement"). Any defined term used in this Amendment not expressly defined herein shall have the meaning ascribed thereto in the Agreement.

1. Modification of Payee. All payments to be made under the Agreement shall be made to BiolineRx Ltd. or any Third Party assignee of BioLineRx Ltd. permitted under Section 10.4 of the Agreement.

2. Modification of Assignment. The last two sentences of Section 10.4 of the Agreement are hereby amended and restated as follows:

> "BioLineRx Ltd. may assign its right to receive payments hereunder to a Third Party, in its sole discretion, provided that BioLineRx Ltd. provides Ikaria with prior written notice of the assignment and the name and address of the assignee. Any such Third Party assignee may not further assign the right to receive payments hereunder without providing Ikaria with prior written notice of the assignment and the name and address of the assignee. Ikaria shall maintain a written record of any such assignments. The parties intend that this Agreement shall be considered to be in "registered form" as defined in United States Treasury Regulations Section 5f.103-1(c). BiolineRx shall not otherwise be permitted to assign this Agreement, in whole or in part, without the prior written consent of Ikaria, which approval shall not be unreasonably withheld, conditioned, or delayed. Any assignment in contravention of this Section 10.4 shall be null and void."

3. Ratification of Agreement. Except as set forth in this Amendment, all of the other terms and conditions of the Agreement are hereby ratified and confirmed to be of full force and effect, and shall continue in full force and effect. This Amendment is hereby integrated into and made a part of the Agreement.

4. Counterparts. This Amendment may be executed in two or more counterparts, each of which shall be effective as of the Amendment Effective Date, and all of which shall constitute one and the same instrument. Each such counterpart shall be deemed an original, and it shall not be necessary in making proof of this Amendment to produce or account for more than one such counterpart.

5. Execution and Delivery. This Amendment shall be deemed executed by the parties when any one or more counterparts hereof, individually or taken together, bears the signatures of each of the parties hereto.

Acknowledged and Agreed to:

BIOLINERX LTD.

By: /s/ Kinneret L. Savitsky /s/ Philip Serlin
Signature

Kinneret L. Savitsky Philip Serlin
Printed Name

CEO CFO
Title

April 21, 2010

IKARIA DEVELOPMENT SUBSIDIARY ONE LLC

By: /s/ Matthew M. Bennett
Signature

Matthew M. Bennett
Printed Name

Vice President and Secretary
Title

April 21, 2010

BIOLINE INNOVATIONS JERUSALEM L.P., BY ITS GENERAL PARTNER BIOLINE INNOVATIONS JERUSALEM, LTD.

By: /s/ Kinneret L. Savitsky /s/ Philip Serlin
Signature

Kinneret L. Savitsky Philip Serlin
Printed Name

CEO CFO
Title

April 21, 2010

Exhibit 10.9

AGREEMENT NOT TO COMPETE

This AGREEMENT NOT TO COMPETE (this "Agreement") is made as of (the "Effective Date"), by and between IKARIA ACQUISITION INC., a Delaware corporation ("Ikaria"), and (" ").

BACKGROUND

A. Ikaria and certain other parties named therein are parties to an Amended and Restated First Lien Credit Agreement dated as of July 3, 2013 (the "First Lien Credit Agreement") and to a Second Lien Credit Agreement dated as of July 3, 2013 (the "Second Lien Credit Agreement" and, together with the first Lien Credit Agreement, the "Credit Agreements").

B. The Credit Agreements permit, under certain circumstances, for Ikaria and its Subsidiaries to engage in one or more R&D Business Asset Transfers.

C. Pursuant to Section 5.12(c) of the First Lien Credit Agreement and Section 5.12(c) of the Second Lien Credit Agreement, Ikaria is required to cause to enter into an R&D Business Subsidiary Noncompete Agreement (as defined in the Credit Agreements).

D. This Agreement is intended to satisfy Ikaria's aforementioned obligations under Section 5.12(c) of the First Lien Credit Agreement and Section 5.12(c) of the Second Lien Credit Agreement.

NOW, THEREFORE, for the purposes set forth above and in consideration of the mutual covenants contained herein, and other good and valuable consideration, the receipt and sufficiency of which are hereby acknowledged, and intending to be legally bound hereby, the parties hereto agree as follows:

1. Capitalized Terms: Definitions. All capitalized terms not defined in this Agreement shall have the same meanings ascribed to them in the Credit Agreements. In addition, the following terms shall have their respective meanings set forth below:

(a) "Controlling Representative" shall have the meaning assigned to such term in the Intercreditor Agreement dated as of July 3, 2013 (the "Intercreditor Agreement"), among Ikaria, Ikaria, Inc., the other Ikaria Companies party thereto, Credit Suisse AG, as collateral agent under each Credit Agreement, and the other representatives from time to time party thereto.

(b) "Ikaria Companies" shall mean Ikaria and its corporate parents, subsidiaries, and affiliates, and each of their respective assigns and successors (in each case whether now in existence or later formed, acquired, merged with or into, or otherwise, and regardless of the form of legal entity).

"Ikaria NO Business" shall mean the development, manufacture,

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commercialization, promotion, sale, import, export, servicing, repair, training, storage, distribution, transportation, licensing, or other handling or disposition of any product or service (including, without limitation, any product or service that utilizes, contains, or includes nitric oxide for inhalation, a device intended to deliver nitric oxide, or a service that delivers or supports the delivery of nitric oxide), bundled or unbundled, for or used in connection with (A) the diagnosis, prevention, or treatment, in both adult and/or pediatric populations, and whether in or out patient, of: (i) hypoxic respiratory failure associated with pulmonary hypertension, (ii) pulmonary hypertensive episodes and right heart failure associated with cardiovascular surgery, (iii) bronchopulmonary dysplasia, (iv) the management of ventilation – perfusion mismatch in acute lung injury, (v) the management of ventilation – perfusion mismatch in acute respiratory distress syndrome, (vi) the management of pulmonary hypertension episodes and right heart failure in congestive heart failure, (vii) pulmonary edema from high altitude sickness, (viii) the management of pulmonary hypertension episodes and right heart failure in pulmonary or cardiac surgery, (ix) the management of pulmonary hypertension episodes and right heart failure in organ transplant, (x) sickle cell vaso-occlusive crisis, (xi) hypoxia associated with pneumonia, (xii) ischemia-reperfusion injury, or (B) the use of nitric oxide to treat or prevent conditions that are primarily managed in the hospital. The "Ikaria NO Business" does not include the development, manufacture, commercialization, promotion, sale, import, export, servicing, repair, training, storage, distribution, transportation, licensing, or other handling or disposition of nitric oxide, a device intended to deliver nitric oxide, or a service that delivers or supports the delivery of nitric oxide for or in connection with the outpatient, chronic treatment of patients who have (1) pulmonary hypertension secondary to chronic obstructive pulmonary disease ("COPD") or idiopathic pulmonary fibrosis ("IPF") or (2) primary or idiopathic pulmonary arterial hypertension ("PAH"), in each case even if initiation of therapy occurs in a hospital setting or such treatment occurs as part of episodic treatment or hospitalization of patients with PAH, IPF, or COPD. In addition, the "Ikaria NO Business" does not include the use of a polymer comprised of Sodium Alginate and Calcium D-Gluconate currently referred to as Bellerophon BCM LLC f/k/a Ikaria Development Subsidiary One LLC's Bioabsorbable Cardiac Matrix (BCM) asset (or any back-ups or second-generation or other future polymers or polymer combinations thereof) in the management of acute myocardial infarction, or the prevention of left ventricular remodeling and the prevention of the progression to heart failure following acute myocardial infarction.

(c) "Terlipressin Business" shall mean any and all development, manufacture, commercialization, promotion, sale, import, export, storage, distribution, transportation, licensing, or other handling or disposition of any terlipressin or any other product within the pressin family, (a) intended to treat (i) hepatorenal syndrome in any form (HRS), (ii) bleeding esophageal varices, or (iii) septic shock, or (b) for or in connection with the management of low blood pressure.

2. Agreements Not to Compete.

(a) hereby agrees that it shall not, anywhere in the world, directly or indirectly, engage in the Ikaria NO Business in any manner, including, without limitation, as a partner, investor (other than as a passive investor in less than three percent (3%) of the outstanding capital stock of a publicly traded corporation), consultant, advisor, agent, representative, independent contractor, licensor, creditor, or otherwise, until the earlier of (i) five

years after the Effective Date of this Agreement or (ii) the date on which all Ikaria Companies are no longer engaged in the Ikaria NO Business (the "NO Noncompetition Period").

(b) hereby agrees that it shall not, anywhere in the world , directly or indirectly, engage in the Terlipressin Business in any manner, including, without limitation, as a partner, investor (other than as a passive investor in less than three percent (3%) of the outstanding capital stock of a publicly traded corporation), consultant, advisor, agent, representative, independent contractor, licensor, creditor, or otherwise, until the earlier of (i) five years after the Effective Date of this Agreement or (ii) the date on which all Ikaria Companies are no longer engaged in the Terlipressin Business (the "Terlipressin Noncompetition Period").

3. Acknowledgment. acknowledges that the restrictions contained in Section 2 are reasonable and properly required for the adequate protection of Ikaria's interest in the Ikaria NO Business and the Terlipressin Business. If any such restriction is deemed to be unreasonable by a court of competent jurisdiction, the parties shall submit to the reduction of such restrictions to such activities, geographical scope, or time period as such court shall deem reasonable.

4. Equitable Relief. acknowledges and agrees that Ikaria would be irreparably harmed by a breach of any of the provisions of Section 2, that Ikaria's remedies at law for such a breach would be inadequate and, in recognition of those facts, in the event of the breach or threatened breach by of the provisions of Section 2, it is agreed that, in addition to its remedies at law, lkaria shall be entitled to seek equitable relief in the form of specific performance, temporary restraining order, temporary or permanent injunction, or any other equitable remedy which may then be available, without posting any bond or other undertaking.

5. Waiver. No provision of this Agreement may be modified, waived, or discharged unless such waiver, modification, or discharge is agreed to in writing and signed by each party hereto, with the prior written consent of the Controlling Representative (not to be unreasonably withheld, delayed, or conditioned). No waiver by either party hereto at any time of any breach by the other party hereto of, or compliance with, any condition or provision of this Agreement to be performed by such other party shall be deemed a waiver of similar or dissimilar provisions or conditions at the same or at any prior or subsequent time.

6. Assignment. This Agreement and the rights and obligations hereunder shall be binding upon and inure to the benefit of the parties hereto, and their respective successors and assigns. Notwithstanding the foregoing, this Agreement shall not be assignable by , whether by operation of law or otherwise, without the express written consent of Ikaria. hereby acknowledges and consents to the assignment of this agreement by Ikaria, pursuant to each Guarantee and Collateral Agreement (as defined in each Credit Agreement), as security for the Obligations under each Credit Agreement.

7. Entire Agreement. This Agreement is intended to define the full extent of the legally enforceable undertakings and representations of the parties hereto, and no promise or representation, written or oral, which is not set forth explicitly in this Agreement is intended by either party to be legally binding.

8. Amendment. This Agreement may not be amended, supplemented or otherwise modified except by an instrument in writing that specifically refers to this Agreement, signed by both parties, with the prior written consent of the Controlling Representative (not to be unreasonably withheld, delayed, or conditioned).

9. Intended Third Party Beneficiaries. Each Collateral Agent (as defined in each Credit Agreement) and each corporate parent, subsidiary, and affiliate of Ikaria (whether now in existence or later formed, acquired, merged with or into, or otherwise, and regardless of the form of legal entity) shall be, and hereby is, an intended third party beneficiary of this Agreement (including, without limitation, the agreements by not to compete in the Ikaria NO Business and the Terlipressin Business, as set forth in Sections 2(a) and 2(b) above respectively) and (subject to the terms of each Guarantee and Collateral Agreement and the Intercreditor Agreement) shall have full right and authority to enforce the terms and conditions of this Agreement.

10. Termination of Certain Rights. Any and all rights of the Controlling Agent and each Collateral Agent under this Agreement shall immediately and automatically terminate upon the expiration or termination of the relevant Credit Agreement and the repayment in full of all Obligations due and owing under the relevant Credit Agreement.

11. Applicable Law; Jurisdiction. This Agreement shall be deemed to have been made in the State of New Jersey and its form, execution, validity, construction and effect shall be determined in accordance with the laws of the State of New Jersey, without giving effect to the principles of conflicts of law thereof.

12. Headings. The headings of the Sections of this Agreement are for convenience only and shall not control or affect the meaning or construction or limit the scope or intent of any of the provisions of this Agreement.

13. Counterparts. This Agreement may be executed in two or more counterparts, each of which shall be deemed an original, but which together shall constitute one and the same instrument. Any executed counterpart delivered by facsimile or other means of electronic transmission shall be deemed an original for all purposes.

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IN WITNESS WHEREOF, the parties hereto, intending to be legally bound, have caused this Agreement to be executed on the day and year first written above.

IKARIA ACQUISITION, INC.

By: _____

Name:

Title:

[ ]

By: _____
Name:
Title:

Exhibit 10.10

Execution Version

Confidential Materials omitted and filed separately with the
Securities and Exchange Commission. Double asterisks denote omissions.

SEPARATION AND DISTRIBUTION AGREEMENT

dated as of February 9, 2014

entered into by and among

IKARIA, INC.,

BELLEROPHON THERAPEUTICS LLC

and

IKARIA ACQUISITION INC.

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SEPARATION AND DISTRIBUTION AGREEMENT

This SEPARATION AND DISTRIBUTION AGREEMENT, dated as of February 9, 2014 (this "Agreement"), is entered into by and among Ikaria, Inc., a Delaware corporation ("Ikaria"), Ikaria Acquisition Inc., a Delaware corporation (together with its successor, Ikaria Acquisition LLC, a Delaware limited liability company "AcquisitionCo"), and Bellerophon Therapeutics LLC, a Delaware limited liability company ("R&DCo"). Each of Ikaria and R&DCo are sometimes referred to herein as a "Party," and Ikaria and R&DCo are sometimes referred to herein collectively as the "Parties." Capitalized terms used herein and not otherwise defined shall have the respective meanings assigned to them in Article 1 of this Agreement.

RECITALS

WHEREAS, Ikaria is a fully-integrated biotherapeutics company focused on developing and commercializing innovative therapeutics and interventions designed to meet the significant unmet medical needs of critically ill patients;

WHEREAS, the Board of Directors of Ikaria (the "Ikaria Board") has determined that it is appropriate, desirable and in the best interests of Ikaria and its stockholders to separate Ikaria into two independent companies, one for each of: (a) the Ikaria Business, which shall continue to be owned and conducted, directly or indirectly, in addition to any other line of business it may conduct, by Ikaria, and (b) the R&DCo Business, which shall be owned and conducted, directly or indirectly, by R&DCo (such separation, the "Separation");

WHEREAS, (a) the applicable Ikaria Group Members have transferred, or will transfer, all of their right, title and interest in and to the R&DCo Assets to the applicable R&DCo Group Members; (b) the applicable R&DCo Group Members have transferred, or will transfer, all of their right, title and interest in and to the Ikaria Assets to the applicable Ikaria Group Members; (c) the applicable Ikaria Group Members have assumed, or will assume, the Ikaria Liabilities; and (d) the applicable R&DCo Group Members have assumed, or will assume, the R&DCo Liabilities, in each case as more fully described in this Agreement and the Ancillary Documents;

WHEREAS, INO Therapeutics LLC, a Delaware limited liability company ("INO Therapeutics"), is entering into the R&D Cross-License Agreement with Bellerophon Pulse Technologies LLC (formerly known as Ikaria Pulse Technologies LLC), a Delaware limited liability company ("Pulse Technologies"), pursuant to which the parties thereto have granted to one another the rights and licenses set forth therein;

WHEREAS, AcquisitionCo is a direct Subsidiary of Ikaria, and INO Therapeutics is a direct Subsidiary of AcquisitionCo;

WHEREAS, for purposes of completing the Separation, (a) R&DCo has been formed as a direct Subsidiary of AcquisitionCo; (b) Pulse Technologies was formed initially as a direct Subsidiary of INO Therapeutics; (c) INO Therapeutics distributed to AcquisitionCo, by means of a special dividend, 100% of the outstanding membership interests of Pulse Technologies; (d) Ikaria Development Subsidiary One LLC, a Delaware limited liability company and a direct Subsidiary of AcquisitionCo, was initially renamed Ikaria BCM LLC and has been subsequently

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renamed Bellerophon BCM LLC ("Bellerophon BCM"); (e) AcquisitionCo contributed 100% of the membership interests of each of Pulse Technologies and Bellerophon BCM to R&DCo; and (f) R&DCo formed Bellerophon Services, Inc., a Delaware corporation, as its wholly owned subsidiary ("ServicesCo" and, together with Pulse Technologies and Bellerophon BCM, the "R&DCo Subsidiaries");

WHEREAS, on or prior to the date hereof AcquisitionCo will have distributed to Ikaria, by means of a special dividend, 1% of the outstanding R&DCo Voting Units (the "Initial Distribution");

WHEREAS, following the Initial Distribution but prior to the Merger Closing, all outstanding shares of Ikaria's preferred stock, par value $0.01 per share, and non-voting common stock, par value $0.01 per share, will convert into shares of Ikaria's voting common stock, par value $0.01 per share, pursuant to the terms of an amendment to be filed to Ikaria's Restated Certificate of Incorporation (the "Stock Conversion");

WHEREAS, following the Stock Conversion, Ikaria will distribute to holders of shares of Ikaria Capital Stock, by means of a special dividend, the R&DCo Voting Units received by Ikaria in the Initial Distribution, as more fully described in this Agreement and the Ancillary Documents (such distribution, the "1% Distribution");

WHEREAS, immediately following the 1% Distribution, AcquisitionCo will convert from a Delaware corporation into a Delaware limited liability company (the "LLC Conversion");

WHEREAS, following the LLC Conversion but prior to the Merger Closing, (a) AcquisitionCo will distribute to Ikaria, by means of a special dividend, the remaining R&DCo Voting Units held by AcquisitionCo and (b) immediately thereafter, Ikaria shall distribute to holders of shares of Ikaria Capital Stock, by means of a special dividend, such R&DCo Voting Units, as more fully described in this Agreement and the Ancillary Documents (the "99% Distribution" and, together with the 1% Distribution, the "Distribution");

WHEREAS, this Agreement is being entered into in connection with and prior to the consummation (the "Merger Closing") of the transactions contemplated by that certain Agreement and Plan of Merger, dated as of December 24, 2013 (as amended, the "Merger Agreement"), by and between Ikaria, Compound Holdings I, LLC, a Delaware limited liability company ("Purchaser Parent"), Compound Holdings II, Inc., a Delaware corporation and wholly owned Subsidiary of Purchaser Parent ("Purchaser"), Compound Merger Sub I, Inc., a Delaware corporation and wholly owned Subsidiary of Purchaser Parent, Compound Merger Sub II, Inc., a Delaware corporation and wholly owned Subsidiary of Purchaser, and New Mountain Partners II, L.P., a Delaware limited partnership, solely in its capacity as the stockholder representative; and

WHEREAS, it is appropriate and desirable to set forth certain transactions required to effect the Separation, the Distribution and certain other agreements that will govern certain matters relating to the Separation, the Distribution and the relationship of Ikaria, R&DCo and their respective Subsidiaries following the Separation and the Distribution.

AGREEMENT

NOW, THEREFORE, in consideration of the foregoing and the mutual covenants and agreements set forth below, and other good and valuable consideration, the receipt and adequacy of which is hereby acknowledged, the Parties hereby agree as follows:

ARTICLE 1
DEFINITIONS

The following terms, as used in this Agreement, have the following meanings:

"1% Distribution" has the meaning set forth in the Recitals to this Agreement.

"99% Distribution" has the meaning set forth in the Recitals to this Agreement.

"access" has the meaning set forth in Section 4.3(b).

"Accredited Investor" has the meaning ascribed thereto in Rule 501 under the Securities Act.

"AcquisitionCo" has the meaning set forth in the Recitals to this Agreement.

"Action" means any action, claim, demand, suit, arbitration, mediation, inquiry, audit, hearing, proceeding or investigation by or before any Governmental Authority.

"Affiliate" of any Person means a Person that controls, is controlled by, or is under common control with such Person. It is expressly agreed that, from and after the Distribution, solely for purposes of this Agreement (a) no member of the R&DCo Group shall be deemed to be an Affiliate of any member of the Ikaria Group and (b) no member of the Ikaria Group shall be deemed to be an Affiliate of any member of the R&DCo Group.

"Agreement" has the meaning set forth in the preamble to this Agreement.

"Ancillary Documents" means each of the Transition Services Agreement, the R&D Cross-License Agreement, the Manufacturing and Supply Agreements, the Employee Matters Agreement and the Non-Competition Agreements, including any exhibits, schedules, attachments, tables or other appendices

thereto.

"Assets" means assets, properties, claims and rights (including goodwill), wherever located (including in the possession of vendors or other third parties or elsewhere), of every kind, character and description, whether real, personal or mixed, tangible, intangible or contingent, in each case whether or not recorded or reflected or required to be recorded or reflected on the books and records or financial statements of any Person.

"BCM" (Bioabsorbable Cardiac Matrix) means a polymer comprised of Sodium Alginate and Calcium D-Gluconate (including that designated by Ikaria as "IK-5001" and by BioLineRx

as "BL-1040"), or any back-ups or second-generation or other future polymers or polymer combinations thereof, for any and all uses, including uses for the prevention of left ventricular remodeling after acute myocardial infarction or other indication.

"BCM Business" means the use of a polymer comprised of Sodium Alginate and Calcium D-Gluconate currently referred to as Bellerophon BCM LLC's Bioabsorbable Cardiac Matrix (BCM) asset (or any back-ups or second-generation or other future polymers or polymer combinations thereof) in the management of acute myocardial infarction, or the prevention of left ventricular remodeling and the prevention of the progression to heart failure following acute myocardial infarction.

"Bellerophon BCM" has the meaning set forth in the Recitals to this Agreement.

"Business Day" means a day other than a Saturday, a Sunday or a day on which banking institutions located in New York, New York are authorized or obligated by Law to close.

"Cash and Cash Equivalents" has the meaning set forth in the definition of "R&DCo Assets" set forth herein.

"Claim Notice" has the meaning set forth in Section 6.3(a).

"Claimed Amount" has the meaning set forth in Section 6.3(a).

"Consents" means any consents, waivers or approvals from, or notification requirements to, any third parties.

"Contracts" means any contract, agreement, lease, license, sales order, purchase order, instrument or other commitment that is binding on any Person or any part of its property under applicable Law.

"control" means the possession, directly or indirectly, of the power to direct or cause the direction of the management and policies of such entity, whether through ownership of voting securities or other interests, by contract or otherwise.

"Control Assessments and Audit" has the meaning set forth in Section 4.6(b).

"Controlling Party" has the meaning set forth in Section 6.3(d)(ii).

"COPD" means pulmonary hypertension secondary to chronic obstructive pulmonary disease.

"Copyright" has the meaning set forth in the definition of "Intellectual Property" contained herein.

"Damages" means all losses, claims, demands, damages, Liabilities, Taxes, judgments, dues, penalties, assessments, fines (civil, criminal or administrative), costs, liens, forfeitures, settlements, fees or expenses (including reasonable attorneys' fees and expenses and any other

expenses reasonably incurred in connection with investigating, prosecuting or defending a claim or Action), of any nature or kind, whether or not the same would properly be reflected on a balance sheet.

"Device Clinical Supply Agreement" means that certain Device Clinical Supply Agreement to be entered into by and between INO Therapeutics and Pulse Technologies on or prior to the Distribution Date, in the form attached hereto as Exhibit A.

"Distribution" has the meaning set forth in the Recitals to this Agreement.

"Distribution Date" means the date on which the 99% Distribution occurs.

"Drug Clinical Supply Agreement" means that certain Drug Clinical Supply Agreement to be entered into by and between INO Therapeutics and Pulse Technologies on or prior to the Distribution Date, in the form attached hereto as Exhibit B.

"Employee Matters Agreement" means that certain Employee Matters Agreement to be entered into by and between Ikaria and R&DCo on or prior to the Distribution Date, in the form attached hereto as Exhibit C.

"Environmental Law" means any Law or any Permit relating to health, safety, pollution or the environment (including ambient air, surface water, groundwater, land surface or subsurface strata) or to Releases or threatened Releases of any Hazardous Material, or otherwise relating to the manufacture, processing, distribution, use, treatment, storage, disposal, transport or handling of any Hazardous Material, including the Comprehensive Environmental Response, Compensation and Liability Act, the Superfund Amendments and Reauthorization Act and the Resource Conservation and Recovery Act and comparable provisions in state, local, foreign or international Law.

"Environmental Liabilities" means all Liabilities (including all removal, remediation, cleanup or monitoring costs, investigatory costs, response costs, natural resources damages, property damages, personal injury damages, judgment or other determination of Liability and indemnity, contribution or similar obligations and all costs and expenses, interest, fines, penalties or other monetary sanctions in connection therewith) relating to, arising out of or resulting from any (a) actual or alleged (i) compliance or noncompliance with any Environmental Law, (ii) generation, use, storage, manufacture, processing, handling, possession, management, treatment, transportation, distribution, emission, discharge or disposal of any Hazardous Material, or (iii) presence, Release or threatened Release of, or exposure to, any Hazardous Material (including any exposure of any current or former Employee (as defined in the Employee Matters Agreement) or any individual who is, or was, an independent contractor, temporary employee, temporary service worker, consultant, freelancer, agency employee, on-call worker, incidental worker, or non-payroll worker for any Ikaria Group Member or R&DCo Group Member, as the case may be, to Hazardous Materials, except for claims that arise under, or are covered or barred by, workers' compensation laws and/or workers' compensation, disability or other insurance providing medical care and/or compensation to injured workers) or (b) Contract pursuant to which Liability is assumed or imposed with respect to any of the foregoing.

"Escalation Notice" has the meaning set forth in Section 7.2(a).

"First Party" has the meaning set forth in Section 4.6(c).

"First Party's Auditors" has the meaning set forth in Section 4.6(c).

"GAAP" means U.S. generally accepted accounting principles, consistently applied.

"Governmental Authority" means any federal, state, local or foreign governmental or quasi-governmental entity or municipality or subdivision thereof or any authority, department, commission, board, bureau, agency, court, tribunal or instrumentality, or any applicable securities exchange or self-regulatory organization.

"Group" means either the Ikaria Group or the R&DCo Group, as the context requires.

"Group Member" means either an Ikaria Group Member or an R&DCo Group Member, as the context requires.

"Hazardous Material" means (a) any petroleum or petroleum products, radioactive materials, toxic mold, radon, asbestos or asbestos-containing materials in any form, lead-based paint, urea formaldehyde foam insulation, or polychlorinated biphenyls (PCBs); and (b) any chemicals, materials, substances, compounds, mixtures, products or byproducts, biological agents, living or genetically modified materials, pollutants, contaminants or wastes that are now or hereafter become defined or characterized as or included in the definition of "hazardous substances," "hazardous wastes," "hazardous materials," "extremely hazardous wastes," "restricted hazardous wastes," "special waste," "toxic substances," "pollutants," "contaminants," "toxic," "dangerous," "corrosive," "flammable," "reactive," "radioactive," or words of similar import, under any Environmental Law.

"Ikaria" has the meaning set forth in the preamble to this Agreement.

"Ikaria Assets" means:

(a) the Ikaria Names and all other Intellectual Property of Ikaria and its Subsidiaries;

(b) any and all Assets that are contemplated by this Agreement or any Ancillary Document (including any schedule or exhibit hereto or thereto) as Assets to be retained by an Ikaria Group Member;

(c) the capital stock and other equity interests of each of Ikaria's Subsidiaries other than R&DCo and the R&DCo Subsidiaries;

(d) all Contracts to which Ikaria or any of its Subsidiaries is a party or by which they or any of their respective Assets are bound and any rights or claims (whether accrued or contingent) of Ikaria or any of its Subsidiaries arising thereunder, other than this Agreement and the Ancillary Documents;

(e) the Ikaria Books and Records;

(f) all Tax credits, receivables, and loss carryforwards from Tax authorities of Ikaria and its Subsidiaries; and

(g) all other Assets of Ikaria and its Subsidiaries (treating for this purpose the R&DCo Group as Subsidiaries of Ikaria prior to the Distribution) that are not R&DCo Assets.

"Ikaria Board" has the meaning set forth in the Recitals to this Agreement.

"Ikaria Books and Records" means originals or true and complete copies thereof, including electronic copies (if available), of (a) minute books, corporate charters and bylaws or comparable constitutive documents, records of share issuances and related corporate records, of the Ikaria Group; (b) all books and records relating to (i) Ikaria Employees, (ii) the purchase of materials, supplies and services for the Ikaria Business, (iii) dealings with customers of the Ikaria Business and (iv) dealings with licensors to the Ikaria Business; and (c) all files relating to any Action the Liability with respect to which is an Ikaria Liability. Notwithstanding the foregoing, "Ikaria Books and Records" shall not include (A) any Tax Returns that relate solely to any R&DCo Group Member or (B) the R&DCo Books and Records.

"Ikaria Business" means the businesses or operations of the Ikaria Group other than the R&DCo Business.

"Ikaria Capital Stock" means any of the outstanding shares of capital stock of Ikaria, including shares of its (a) common stock, par value $0.01 per share, and (b) preferred stock, par value $0.01 per share, regardless of (i) whether such shares are designated "voting" or "non-voting" and (ii) which class such shares have been designated, in each case, pursuant to Ikaria's Restated Certificate of Incorporation, as amended.

"Ikaria Employee" has the meaning set forth in the Employee Matters Agreement.

"Ikaria Group" means Ikaria and each other Person that Ikaria now or hereafter controls (other than R&DCo or any R&DCo Group Member).

"Ikaria Group Member" means any Person now or hereafter included in the Ikaria Group.

"Ikaria Indemnified Parties" has the meaning set forth in Section 6.1.

"Ikaria Liabilities" means:

(a) any and all Liabilities that are expressly contemplated by this Agreement or any Ancillary Document (or any other schedule hereto or thereto) as Liabilities to be retained or assumed by an Ikaria Group Member, and all agreements and obligations of any Ikaria Group Member under this Agreement or any of the Ancillary Documents;

(b) any and all Liabilities relating to, arising out of or resulting from any Ikaria Assets;

(c) any and all Environmental Liabilities relating to, arising out of or resulting from:

(i) researching, developing, manufacturing, finishing, marketing, distributing, leasing, selling or other operations associated with the Ikaria Business and the Ikaria Assets to the extent conducted by Ikaria and its Subsidiaries, at any time at or prior to the date hereof in any such case whether occurring or arising before, on or after the date hereof; or

(ii) any real property that is an Ikaria Asset, whether occurring or arising before, on or after the date hereof.

(d) any and all Liabilities relating to, arising out of or resulting from any Indebtedness of any Ikaria Group Member (whether incurred prior to, or after the date hereof);

(e) any and all Liabilities of the Ikaria Group for Taxes that are imposed on the Ikaria Group (A) on or before the Distribution with respect to the operations of the R&DCo Group prior to the Distribution, and (B) with respect to the operations of the Ikaria Group, whether or not such Taxes are attributable to R&DCo Assets, including any Taxes incurred as the result of the Distribution; and

(f) any and all other Liabilities of Ikaria and its Subsidiaries that are not R&DCo Liabilities;

provided that, notwithstanding anything herein to the contrary, Ikaria Liabilities shall not include any R&DCo Liabilities.

"Ikaria Names" means the trade names specified on Schedule 1.1(a) (and any derivatives of any such trade name) and associated logos or Trademarks.

"Ikaria Stock Option" has the meaning set forth in the Employee Matters Agreement.

"Ikaria RSU" has the meaning set forth in the Employee Matters Agreement.

"Indebtedness" of any Person means (a) all obligations of such Person for borrowed money, (b) all obligations of such Person evidenced by bonds, debentures, notes or similar instruments, (c) all obligations of such Person upon which interest charges are customarily paid, (d) all obligations of such Person under conditional sale or other title retention agreements relating to property or assets purchased by such Person, (e) all obligations of such Person issued or assumed as the deferred purchase price of property or services, (f) all indebtedness of others secured by (or for which the holder of such indebtedness has an existing right, contingent or otherwise, to be secured by) any mortgage, lien, pledge, or other encumbrance on property owned or acquired by such Person, whether or not the obligations secured thereby have been assumed, (g) all guarantees by such Person of indebtedness of others, (h) all capital lease obligations of such Person and (i) all securities or other similar instruments convertible or exchangeable into any of the foregoing, but excluding daily cash overdrafts associated with routine cash operations.

"Indemnified Party" has the meaning set forth in Section 6.3(a).

"Indemnifying Party" has the meaning set forth in Section 6.3(a).

"Information" means information, whether or not patentable or copyrightable, in written, oral, electronic or other tangible or intangible form, stored in any medium, including studies, reports, records, books, contracts, instruments, surveys, discoveries, ideas, concepts, know-how, techniques, designs, specifications, drawings, blueprints, diagrams, models, prototypes, samples, flow charts, data, computer data, disks, diskettes, tapes, computer programs or other software, marketing plans, customer names, communications by or to attorneys (including attorney-client privileged communications), memos and other materials prepared by attorneys or under their direction (including attorney work product), and other technical, financial, employee or business information or data.

"Initial Distribution" has the meaning set forth in the Recitals to this Agreement.

"INO Therapeutics" has the meaning set forth in the Recitals to this Agreement.

"Intellectual Property" means all intellectual property throughout the world, including all U.S. and foreign (a) patents, invention disclosures, and all related continuations, continuations-in-part, divisionals, provisionals, renewals, reissues, re-examinations, additions, extensions (including all supplementary protection certificates), and all applications and registrations therefor ("Patent Rights"), (b) trademarks, service marks, names, corporate names, trade names, domain names, logos, slogans, trade dress, design rights, and other similar designations of source or origin and all applications and registrations therefor, together with the goodwill symbolized by any of the foregoing ("Trademarks"), (c) copyrights and copyrightable subject matter and all applications and registrations therefor ("Copyrights"), and (d) any and all trade secrets, confidential data and technical information, including practices, techniques, methods, processes,

inventions, developments, specifications, formulations, manufacturing processes, structures, chemical or biological manufacturing control data, analytical and quality control information and procedures, pharmacological, toxicological and clinical test data and results, stability data, studies and procedures and regulatory information ("Know-How").

"Internal Reorganization Contracts" means (i) that certain Contribution Agreement, dated as of October 17, 2013, by and between INO Therapeutics and Pulse Technologies; (ii) that certain Contribution Agreement, dated as of October 17, 2013, by and between AcquisitionCo and R&DCo; (iii) that certain Contribution Agreement, dated as of October 17, 2013, by and between AcquisitionCo and R&DCo; (iv) that certain Assignment and Assumption Agreement, dated as of October 17, 2013, by and between Ikaria and Bellerophon BCM; (v) that certain Assignment and Assumption Agreement, dated as of October 17, 2013, by and between Ikaria Therapeutics LLC and Bellerophon BCM; (vi) that certain Assignment and Assumption Agreement, dated as of October 17, 2013, by and between INO Therapeutics and Bellerophon BCM; and (vii) that certain Contribution Agreement, dated as of February 7, 2014, by and between Ikaria, INO Therapeutics and ServicesCo.

"IPF" shall mean idiopathic pulmonary fibrosis.

"Know-How" has the meaning set forth in the definition of "Intellectual Property".

"Law" means each applicable law, order, judgment, rule, code, statute, regulation, requirement, variance, decree, writ, injunction, award, ruling or ordinance of any Governmental Authority, including the common law.

"Liabilities" means debts, liabilities, guarantees, assurances, commitments and obligations of any nature or description, whether fixed, contingent or absolute, asserted or unasserted, matured or unmatured, liquidated or unliquidated, accrued or not accrued, known or unknown, due or to become due, whenever or however arising (including any of the foregoing arising out of (a) any Contract or tort based on negligence or strict liability or (b) any act or failure to act by any past or present Representative, whether or not such act or failure to act was within such Representative's authority), and whether or not the same would be required by GAAP to be reflected in financial statements or disclosed in the notes thereto.

"Lien" means any mortgage, security interest, pledge, lien, charge, claim, option, right to acquire, voting or other restriction, right-of-way, covenant, condition, easement, encroachment, restriction on transfer or other encumbrance of any nature whatsoever.

"LLC Conversion" has the meaning set forth in the Recitals to this Agreement.

"Manufacturing and Supply Agreements" means the Drug Clinical Supply Agreement and the Device Clinical Supply Agreement.

"Merger Agreement" has the meaning set forth in the Recitals to this Agreement.

"Non-Competition Agreements" means, collectively, the Agreement Not To Compete entered into by and between AcquisitionCo and Bellerophon BCM on September 20, 2013; the Agreement Not To Compete entered into by and between AcquisitionCo and Pulse Technologies on October 18, 2013; the Agreement Not To Compete entered into by and between AcquisitionCo and R&DCo on October 18, 2013; and the Agreement Not To Compete entered into by and between AcquisitionCo and ServicesCo on February 7, 2014.

"Non-controlling Party" has the meaning set forth in Section 6.3(d)(ii).

"Non-Party Affiliates" has the meaning set forth in Section 6.5.

"Other Party's Auditors" has the meaning set forth in Section 4.6(c).

"PAH" shall mean primary or idiopathic pulmonary arterial hypertension

"Party" has the meaning set forth in the preamble to this Agreement.

"Patent Rights" has the meaning set forth in the definition of "Intellectual Property" contained herein.

"Permit" means any license, permit, franchise, approval, consent, registration or authorization issued by any Governmental Authority.

"Person" means an individual, a partnership, a corporation, a limited liability company, an association, a joint stock company, a trust, a joint venture, an unincorporated organization or any Governmental Authority.

"Possessor" has the meaning set forth in Section 4.3(b).

"Post-Distribution Financing" means any public or private financing (or refinancing) undertaken by either Party or their respective Group Members, or a secondary offering of the securities of either Party or their respective Group Members by the stockholders of such Party or Group Members, following the Distribution Date, whether by means of a syndicated or unsyndicated commercial loan or a public or private offering of debt, equity or other securities.

"Pulse Technologies" has the meaning set forth in the Recitals to this Agreement.

"Purchaser Parent" has the meaning set forth in the Recitals to this Agreement.

"R&D Cross-License Agreement" means that certain Exclusive Cross-License, Technology Transfer, and Regulatory Matters Agreement to be entered into by and between INO Therapeutics and Pulse Technologies on or prior to the Distribution Date, in the form attached hereto as Exhibit D.

"R&DCo" has the meaning set forth in the preamble to this Agreement.

"R&DCo Assets" means:

(a) all Assets (i) that were transferred to R&DCo or its Subsidiaries pursuant to the Internal Reorganization Contracts and (ii) that are used solely and/or held for use solely in the BCM Business;

(b) the Intellectual Property described on Schedule 1.1(b);

(c) all R&DCo Books and Records;

(d) all accounting and other legal and business books, records, ledgers and files and all personnel records, in each case, whether printed, electronic, contained on storage media or written, or in any other form, in each case only to the extent solely relating to the R&DCo Business;

(e) all prepaid expenses, trade accounts and other accounts and notes receivable currently held by R&DCo that are not Ikaria Assets;

(f) all cash or cash equivalents, certificates of deposit, banker's acceptances and other investment securities of any form or maturity and all bank accounts, lock boxes and other deposit arrangements and all brokerage accounts currently held by R&DCo or its Subsidiaries, in each case net of outstanding or uncleared checks, drafts and wire transfers (collectively, "Cash and Cash Equivalents"); and

(g) all accessories that interface or interoperate with, or support the use of, the Assets that were transferred by INO Therapeutics to Pulse Technologies pursuant to the Contribution Agreement, dated October 17, 2013, by and between INO Therapeutics and Pulse Technologies.

"R&DCo Books and Records" means originals or true and complete copies thereof, including electronic copies (if available), of (a) all minute books, certificates of formation, limited liability company agreements, and bylaws or comparable constitutive documents, records of membership interest, unit or share issuances and related records of each member of the R&DCo Group, (b) all books and records exclusively relating to (i) Transferred Employees, (ii) the purchase of materials, supplies and services for the R&DCo Business and (iii) dealings with licensors to the R&DCo Business and (c) all files relating exclusively to any Action the Liability with respect to which is an R&DCo Liability. Notwithstanding the foregoing, "R&DCo Books and Records" shall not include any Tax Returns that relate solely to the Ikaria Group.

"R&DCo Business" means the development, manufacture, commercialization, promotion, sale, import, export, servicing, repair, training, storage, distribution, transportation, licensing or other handling or disposition of nitric oxide, a device intended to deliver nitric oxide, or a service that delivers or supports the delivery of nitric oxide for or in connection with the outpatient, chronic treatment of patients who have COPD, IPF, or PAH, in each case even if initiation of therapy occurs in a hospital setting or such treatment occurs as part of episodic treatment or hospitalization of patients with COPD, IPF or PAH. In addition, the term "R&DCo Business" shall also include the BCM Business.

"R&DCo Group" means R&DCo, the R&DCo Subsidiaries and each other Person that R&DCo controls.

"R&DCo Group Member" means any Person now or hereafter included in the R&DCo Group.

"R&DCo Indemnified Parties" has the meaning set forth in Section 6.2.

"R&DCo Liabilities" means:

(a) any and all Liabilities that are expressly contemplated by this Agreement or any Ancillary Document (or any other schedules hereto or thereto) as Liabilities to be retained or assumed by any R&DCo Group Member, and all agreements, obligations and Liabilities of any R&DCo Group Member under this Agreement, or any of the Ancillary Documents;

(b) any and all Liabilities relating to, arising out of or resulting from any R&DCo Assets;

(c) any and all Liabilities relating to, arising out of or resulting from the conduct and operation of the R&DCo Business, at any time prior to, on or after the date hereof (including any such Liability relating to, arising out of or resulting from any act or failure to act by any Representative of any Ikaria Group Member (whether or not such act or failure to act is or was within such Representative's authority)), in any such case whether occurring or arising before, on or after the date hereof;

(d) any and all Environmental Liabilities relating to, arising out of or resulting from the R&DCo Business or any R&DCo Assets, in any such case whether occurring or arising before, on or after the date hereof;

(e) any and all Liabilities that were assumed by R&DCo or its Subsidiaries pursuant to the Internal Reorganization Contracts;

(f) any and all Liabilities relating to, resulting from or arising out of any Action to the extent relating to the R&DCo Group;

(g) any and all Liabilities arising out of claims made by R&DCo's directors, officers, employees, agents, Subsidiaries or Affiliates against any member of the Ikaria Group or the R&DCo Group to the extent relating to the R&DCo Business; and

(h) any and all Liabilities of the R&DCo Group for Taxes that are imposed on the R&DCo Group with respect to the operations of the R&DCo Group after the Distribution.

"R&DCo LLC Agreement" means the Amended & Restated Limited Liability Company Agreement of Bellerophon Therapeutics LLC to be entered into on or about the date hereof (as amended, modified or supplemented from time to time).

"R&DCo Non-Voting Units" means the Units denominated as Non-Voting Units pursuant to the Amended and Restated Limited Liability Company Agreement of R&DCo.

"R&DCo Subsidiaries" has the meaning set forth in the Recitals to this Agreement.

"R&DCo Voting Units" means the Units denominated as Voting Units pursuant to the Amended and Restated Limited Liability Company Agreement of R&DCo.

"Record Date" means the close of business on the date to be determined by the Ikaria Board as the record date for determining the stockholders of Ikaria entitled to receive R&DCo Voting Units pursuant to the Distribution.

"Release" means any release, spill, emission, leaking, dumping, pumping, injection, pouring, deposit, disposal, discharge, dispersal, leaching or migration into the environment (including ambient air, surface water, groundwater, land surface or subsurface strata, soil and sediments).

"Representatives" means, with respect to any Person, any of such Person's directors, officers, employees, agents, consultants, advisors, accountants, attorneys or other representatives.

"Requestor" has the meaning set forth in Section 4.3(b).

"Required Disclosure" means a disclosure of Information that (a) a Party or one of its Group Members determines is required pursuant to applicable Law (including the rules and regulations of the SEC or any national securities exchange, including as the same may apply to the registration of any securities in connection with a proposed Post-Distribution Financing), (b)

is demanded or requested pursuant to any legal process or (c) is made in response to any demand or request by a Governmental Authority (including any such disclosure to the extent reasonably necessary to respond to any written request or official comment from a Governmental Authority, such as in connection with responding to (i) comment letters from SEC in connection with a Post-Distribution Financing or otherwise or (ii) requests for information from the U.S. Food and Drug Administration).

"Required R&DCo Financial Statements" shall mean (a) audited balance sheets of the R&DCo Business as of December 31, 2011 and December 31, 2012, and audited statements of operations, changes in invested equity (deficit) and cash flows for the R&DCo Business for each of the years ended December 31, 2011 and December 31, 2012 and for the period from August 26, 2009 (inception) to December 31, 2012, together with associated notes to those financial statements, in each case, prepared in accordance with GAAP, (b) an unaudited balance sheet as of September 30, 2013, and unaudited statements of operations, changes in invested equity (deficit) and cash flows for the R&DCo Business for each of the nine-month periods ended September 30, 2013 and 2012, together with associated notes to those unaudited financial statements, in each case, prepared in accordance with GAAP, and (c) reviews prepared by the R&DCo's independent auditor in respect of such independent auditor's performance of the procedures specified by the Public Company Accounting Oversight Board for a review of interim financial information as described in AU 722 with respect to the financial statements described in the foregoing clause (b).

"Restricted Information" has the meaning set forth in Section 4.3(b).

"Retention Period" has the meaning set forth in Section 4.4.

"SEC" means the United States Securities and Exchange Commission or any successor agency.

"Securities Act" means the Securities Act of 1933, as amended.

"Separation" has the meaning set forth in the Recitals to this Agreement.

"ServicesCo" has the meaning set forth in the Recitals to this Agreement.

"Stock Conversion" has the meaning set forth in the Recitals to this Agreement.

"Subsidiary" means with respect to any specified Person, any corporation or other legal entity of which such Person or any of its Subsidiaries controls or owns, directly or indirectly, more than 50% of the stock or other equity interest entitled to vote on the election of the members to the board of directors or similar governing body; provided, however, that unless the context otherwise requires, references to Subsidiaries of Ikaria shall not include R&DCo or any R&DCo Group Members.

"Tax" and "Taxes" means any and all taxes, charges, fees, duties, contributions, levies or other similar assessments or liabilities, including, without limitation, income, gross receipts, corporation, ad valorem, premium, value-added, net worth, capital stock, capital gains,

documentary, recapture, alternative or add-on minimum, disability, registration, recording, excise, real property, personal property, sales, use, license, lease, service, service use, transfer, withholding, employment, unemployment, insurance, social security, national insurance, business license, business organization, environmental, workers compensation, payroll, profits, severance, stamp, occupation, escheat, windfall profits, customs duties, franchise, estimated and other taxes of any kind whatsoever imposed by the United States of America or any state, local or foreign government, or any agency or political subdivision thereof, and any interest, fines, penalties, assessments or additions to tax imposed with respect to such items or any contest or dispute thereof.

"Tax Return" means any and all reports, returns (including information returns), declarations, or statements relating to Taxes, including any schedule or attachment thereto and any related or supporting workpapers or information with respect to any of the foregoing, including any amendment thereof filed with or submitted to any Governmental Entity in connection with the determination, assessment, collection or payment of Taxes or in connection with the administration, implementation or enforcement of or compliance with any legal requirement relating to any Tax, and including, for the avoidance of doubt, U.S. Department of the Treasury Form TD F 90-22.1.

"Third-Party Claim" has the meaning set forth in Section 6.3(d)(i).

"Trademark" has the meaning set forth in the definition of "Intellectual Property" contained herein.

"Transferred Employees" has the meaning set forth in the Employee Matters Agreement.

"Transition Services Agreement" means the Transition Services Agreement entered into by and between Ikaria and R&DCo, effective on or prior to the Distribution Date, to be in the form attached hereto as Exhibit E.

ARTICLE 2
ASSET TRANSFERS

Section 2.1 Transfer of Assets, Assumption of Liabilities.

(a) Except to the extent otherwise provided in this Agreement or any Ancillary Document and only to the extent not previously effectuated prior to the date hereof, Ikaria shall (and Ikaria shall cause each other Ikaria Group Member to) assign, transfer and convey to the applicable R&DCo Group Members, and R&DCo shall (and R&DCo shall cause each other R&DCo Group Member to) receive and accept from the Ikaria Group Members, all of the Ikaria Group Members' right, title and interest in and to the R&DCo Assets (with the R&DCo Assets relating to BCM being transferred to Bellerophon BCM, the R&DCo Assets relating to the employment of the Transferred Employees being transferred to ServicesCo and all other R&DCo Assets being transferred to Pulse Technologies). To the extent not previously effectuated, such assignments, transfers and conveyances shall be effective immediately prior to the Distribution, or at such other times as may be provided in each respective Ancillary

Document and shall be subject to the terms and conditions of this Agreement and any applicable Ancillary Document.

(b) Except to the extent otherwise provided in this Agreement or any Ancillary Document and only to the extent not previously effectuated prior to the date hereof, R&DCo shall (and R&DCo shall cause each other R&DCo Group Member to) assume and on a timely basis pay, perform, satisfy and discharge the R&DCo Liabilities in accordance with their respective terms (with Bellerophon BCM assuming the R&DCo Liabilities relating to BCM, ServicesCo assuming the R&DCo Liabilities relating to the Transferred Employees, and Pulse Technologies assuming all other R&DCo Liabilities). To the extent not previously effectuated prior to the date hereof, such assumptions of R&DCo Liabilities and Ikaria Liabilities shall be effective immediately prior to the Distribution or at such other times as may be provided in each respective Ancillary Document and shall be subject to the terms and conditions of this Agreement and any applicable Ancillary Document.

(c) For a period of seven years following the Distribution, if at any time or from time to time any Party (or Person in such Party's respective Group) shall receive or otherwise possess any Asset or Liability, as applicable, that is allocated to any other Person pursuant to this Agreement or any Ancillary Document, such Party shall use its reasonable best efforts to promptly transfer, or cause to be transferred, such Asset or Liability, as applicable, to the Person so entitled thereto or responsible for the assumption thereof; provided that R&DCo shall only be permitted to make claims under this Section 2.1(c) with respect to Cash and Cash Equivalents until the date that is 45 days after the Effective Time (as defined in the Merger Agreement), and if no such claims are made within such period, R&DCo shall have no further rights under this Section 2.1(c) with respect to Cash and Cash Equivalents following such date.

Section 2.2 Further Assurances. In furtherance of the transactions contemplated by Section 2.1, except to the extent otherwise provided in this Agreement or in any Ancillary Document, from and after the Distribution Date, the Parties shall execute and deliver, and they shall cause their respective Subsidiaries, as applicable, to execute and deliver: (i) to the extent not executed and delivered prior thereto, the Ancillary Documents; (ii) such bills of sale, membership interest or unit, powers, certificates of title, assignments of Contracts, subleases and other instruments of transfer, conveyance and assignment as, and to the extent, necessary or appropriate to evidence (A) the transfer, conveyance and assignment to the R&DCo Group Members of all of the Ikaria Group Members' right, title and interest in and to the R&DCo Assets and (B) the transfer, conveyance and assignment to the Ikaria Group Members of all of the R&DCo Group Members' right, title and interest in and to the Ikaria Assets; and (iii) such assumptions of Contracts and other instruments of assumption as, and to the extent, necessary or convenient to evidence the valid and effective assumption of (A) the R&DCo Liabilities by the R&DCo Group Members and (B) the Ikaria Liabilities by the Ikaria Group Members.

Section 2.3 Transfers Not Effected Prior to Distribution. The Parties acknowledge and agree that some of the transfers of R&DCo Assets contemplated by Section 2.1(a) may not be effected prior to the Distribution due to the inability of the Parties to take certain other actions necessary to effect such transfers prior to such time. To the extent any transfers of such R&DCo Assets contemplated by Section 2.1(a) have not been fully effected prior to the Distribution, each

of Ikaria and R&DCo shall cooperate and use its respective reasonable best efforts (and shall cause each of its applicable Group Members to use its respective reasonable best efforts) to take any other actions necessary to effect such transfers of such R&DCo Assets as promptly as practicable following the Distribution.

Section 2.4 Bank Accounts.

(a) It is intended that R&DCo and Ikaria will maintain separate bank accounts and separate cash management processes.

(b) As between Ikaria and R&DCo (and the members of their respective Groups), all payments made and reimbursements received after the date hereof by either Party (or member of its Group) that relate to a business, Asset or Liability of the other Party (or member of its Group) shall be held by such Party in trust for the use and benefit of the Party entitled thereto and, promptly upon receipt by such Party of any such payment or reimbursement, such Party shall pay over, or shall cause the applicable member of its Group to pay over to the other Party the amount of such payment or reimbursement without right of set-off.

Section 2.5 Novation of R&DCo Liabilities.

(a) R&DCo shall use its reasonable best efforts to obtain, or to cause to be obtained, as soon as practicable following the Distribution, any consent, substitution, approval, release or amendment requested by Ikaria required to novate or assign to the applicable Person in the R&DCo Group all obligations under agreements, leases, licenses and other obligations or Liabilities of any nature whatsoever that constitute R&DCo Liabilities, or to obtain in writing the unconditional release of all parties to such arrangements other than any Person in the R&DCo Group, so that, in any such case, the R&DCo Group Members will be solely responsible for such Liabilities; provided, however, that neither Ikaria nor R&DCo shall be obligated to pay any consideration therefor to any third party from whom such consents, approvals, substitutions, amendments and releases are requested; provided further, however, that any legal fees or other administrative costs associated with obtaining such consents, approvals, substitution, amendments and releases shall be borne by R&DCo.

(b) If R&DCo is unable to obtain, or to cause to be obtained, any such required consent, substitution, approval, release or amendment, the applicable Person in the Ikaria Group shall continue to be bound by such agreements, leases, licenses and other obligations that constitute R&DCo Liabilities and, unless not permitted by Law or the terms thereof, R&DCo shall cause a Person in the R&DCo Group, as agent or subcontractor for such Person in the Ikaria Group, to pay, perform and discharge fully all such obligations or other Liabilities of Ikaria or such other Person that constitute R&DCo Liabilities thereunder from and after the Distribution. R&DCo shall indemnify each Ikaria Indemnified Party and hold each of them harmless against any Liabilities arising in connection therewith. Ikaria shall, without further consideration, pay or remit, or cause to be paid or remitted, to R&DCo promptly all money, rights and other consideration received by it or any Person in the Ikaria Group in respect of such performance. If and when any such consent, approval, release, substitution or amendment shall be obtained or such agreement, lease, license or other rights or obligations shall

otherwise become assignable or able to be novated, Ikaria shall thereafter assign, or cause to be assigned, all its rights, obligations and other Liabilities thereunder or any rights or obligations of any Person in its Group to R&DCo (or to an R&DCo Group Member designated by R&DCo) without payment of further consideration, and R&DCo, without the payment of any further consideration, shall or shall cause its designee to, assume such rights and obligations.

Section 2.6 Novation of Ikaria Liabilities.

(a) Ikaria shall use its reasonable best efforts to obtain, or to cause to be obtained, as soon as practicable following the Distribution, any consent, substitution, approval, release or amendment requested by R&DCo required to novate or assign to the applicable Person in the Ikaria Group all obligations under agreements, leases, licenses and other obligations or Liabilities of any nature whatsoever that constitute Ikaria Liabilities, or to obtain in writing the unconditional release of all parties to such arrangements other than any Person in the Ikaria Group, so that, in any such case, the Ikaria Group Members will be solely responsible for such Liabilities; provided, however, that neither Ikaria nor R&DCo shall be obligated to pay any consideration therefor to any third party from whom such consents, approvals, substitutions, amendments and releases are requested; provided further, however, that any legal fees or other administrative costs associated with obtaining such consents, approvals, substitution, amendments and releases shall be borne by Ikaria.

(b) If Ikaria is unable to obtain, or to cause to be obtained, any such required consent, substitution, approval, release or amendment, the applicable Person in the R&DCo Group shall continue to be bound by such agreements, leases, licenses and other obligations that constitute Ikaria Liabilities and, unless not permitted by Law or the terms thereof, Ikaria shall cause a Person in the Ikaria Group, as agent or subcontractor for such Person in the R&DCo Group, to pay, perform and discharge fully all such obligations or other Liabilities of R&DCo or such other Person that constitute Ikaria Liabilities thereunder from and after the Distribution. Ikaria shall indemnify each R&DCo Indemnified Party and hold each of them harmless against any Liabilities arising in connection therewith. R&DCo shall, without further consideration, pay or remit, or cause to be paid or remitted, to Ikaria promptly all money, rights and other consideration received by it or any Person in the R&DCo Group in respect of such performance. If and when any such consent, approval, release, substitution or amendment shall be obtained or such agreement, lease, license or other rights or obligations shall otherwise become assignable or able to be novated, R&DCo shall thereafter assign, or cause to be assigned, all its rights, obligations and other Liabilities thereunder or any rights or obligations of any Person in its Group to Ikaria (or to an Ikaria Group Member designated by Ikaria) without payment of further consideration, and Ikaria, without the payment of any further consideration shall, or shall cause its designee to, assume such rights and obligations.

ARTICLE 3
THE DISTRIBUTION

Section 3.1 The Distribution.

(a) Ikaria shall effect the Distribution in accordance with the terms of the Merger Agreement. The Distribution shall involve the following steps:

(i) On or prior to the date hereof, AcquisitionCo shall have effected the Initial Distribution by distributing to Ikaria, by means of a special dividend, 1% of the outstanding R&DCo Voting Units.

(ii) Following the Initial Distribution but prior to the Merger Closing, Ikaria shall effect the Stock Conversion by causing the filing of a Certificate of Amendment to Ikaria's Restated Certificate of Incorporation, pursuant to which all outstanding shares of Ikaria's preferred stock, par value $0.01 per share, and non-voting common stock, par value $0.01 per share, shall convert into shares of Ikaria's voting common stock, par value $0.01 per share.

(iii) Following the Stock Conversion, Ikaria shall effect the 1% Distribution by distributing by means of a special dividend to the holders of outstanding shares of Ikaria Capital Stock as of the Record Date, the R&DCo Voting Units received by Ikaria in the Initial Distribution. Such special dividend shall be paid to the holders of Ikaria Capital Stock on a pro rata basis based on each such holder's pro rata ownership of Ikaria Capital Stock, with each such holder receiving 1% of one R&DCo Voting Unit for each share of Ikaria voting common stock held by such Ikaria stockholder on the Record Date.

(iv) Immediately following the 1% Distribution, AcquisitionCo shall effect the LLC Conversion pursuant to which AcquisitionCo shall convert from a Delaware corporation into a Delaware limited liability company.

(v) Following the LLC Conversion but prior to the Merger Closing, AcquisitionCo shall distribute to Ikaria, by means of a special dividend, the remaining R&DCo Voting Units held by AcquisitionCo.

(vi) Immediately thereafter, Ikaria shall effect the 99% Distribution by distributing by means of a special dividend to the holders of outstanding shares of Ikaria Capital Stock as of the Record Date, such R&DCo Voting Units. Such special dividend shall be paid to the holders of Ikaria Capital Stock on a pro rata basis based on each such holder's pro rata ownership of Ikaria Capital Stock, with each such holder receiving 99% of one R&DCo Voting Unit for each share of Ikaria voting common stock held by such Ikaria stockholder on the Record Date.

(b) In addition, in connection with the Distribution and prior to the Distribution Date:

(i) the exercise price of all outstanding Ikaria Stock Options will be adjusted downward to reflect the impact of the Distribution on the value of Ikaria, and options to purchase R&DCo Non-Voting Units will be distributed to holders of outstanding Ikaria Stock Options with exercise prices that are based on the value ascribed to R&DCo in the Distribution, all as set forth more fully in Article VII of the Employee Matters Agreement; provided that each holder of an outstanding Ikaria Stock Option that is neither an Accredited Investor nor an employee of Ikaria will receive, in lieu of each such options to purchase R&DCo Non-Voting

Units, an amount in cash equal to the difference between the fair market value of an R&DCo Non-Voting Unit (as determined by R&DCo) less the exercise price of the options to purchase R&DCo Non-Voting Units, less applicable withholding taxes, which R&DCo shall pay (or cause to be paid) to the holder promptly following completion of the Distribution; and

(ii) each holder of an Ikaria RSU shall receive an award consisting of the same number of restricted Non-Voting Units of R&DCo as were subject to the Ikaria RSU, as set forth more fully in Article VII of the Employee Matters Agreement.

(c) Subject to the terms of the Merger Agreement and with the prior written consent of Purchaser (not to be unreasonably withheld), Ikaria may, at any time and from time to time until the completion of the Distribution, modify or change the terms of the Distribution, including by accelerating or delaying the timing of the consummation of all or part of the Distribution. R&DCo shall cooperate with Ikaria in all respects to accomplish the Distribution and shall, at Ikaria's direction, promptly take any and all actions necessary or desirable to effect the Distribution. For the avoidance of doubt, if the Merger Agreement is terminated in accordance with its terms prior to the Closing as defined therein, Ikaria shall have the right not to complete all or any portion of the Distribution for any or no reason.

Section 3.2 Actions Prior to the Distribution. In connection with the Distribution, the Parties shall take the actions set forth in this Section 3.2.

(a) Ikaria and R&DCo shall prepare and mail, prior to any Distribution Date, to the holders of Ikaria Capital Stock, such information concerning R&DCo and the Distribution and such other matters as Ikaria reasonably determines is necessary or desirable.

(b) R&DCo shall use reasonable best efforts to take all such action as may be necessary or desirable under applicable federal and state securities and blue sky Laws of the United States (and any comparable Laws under any foreign jurisdictions) in connection with the Distribution.

Section 3.3 Certain Stockholder Matters.

(a) On or prior to the Distribution Date, Ikaria shall deliver to R&DCo true, correct and complete copies of the stock and transfer records reflecting the holders of Ikaria Capital Stock entitled to receive R&DCo Voting Units in connection with the Distribution. On the Distribution Date or as soon as reasonably practicable thereafter, Ikaria shall provide notice to each such holder or designated transferee(s) of such holder of the number of R&DCo Voting Units distributed to such holder in the Distribution. Ikaria and R&DCo shall cooperate with each other in connection with all aspects of the Distribution and all other matters relating to the issuance of the R&DCo Voting Units to be distributed to the holders of Ikaria Capital Stock in connection with the Distribution.

(b) Effective immediately upon the 1% Distribution, each Person described in Sections 3.1(a)(iii) and (vi) that acquires an R&DCo Voting Unit shall be admitted as a Member under the R&DCo LLC Agreement and shall be treated as a "Member" for all purposes thereunder.

(c) From and after the Distribution, R&DCo shall regard each Person described in Sections 3.1(a)(iii) and (vi) that is entitled to receive such R&DCo Voting Units as the record holder of R&DCo Voting Units received in accordance with the terms of the Distribution without requiring any action on the part of such Person, until such time as such Person shall have duly transferred the R&DCo Voting Units received in the Distribution in accordance with applicable Law or any other applicable restrictions including restrictions under the R&DCo LLC Agreement. R&DCo agrees that, until any transfer of such R&DCo Voting Units is validly effected, each such holder shall be entitled to all rights and be subject to all obligations as a "Member" under the R&DCo LLC Agreement, including the right to receive all distributions payable on, and exercise voting rights (if any) and all other rights and privileges with respect to, the R&DCo Voting Units then held by such holder in accordance with the R&DCo LLC Agreement.

(d) Ikaria shall be entitled to and shall deduct and withhold Taxes from the distribution of R&DCo Voting Units to holders of Ikaria Capital Stock pursuant to this Agreement as required by Law. Any amount that is so deducted and withheld shall be paid over to the applicable Governmental Authority, and any such amount so deducted, withheld and paid shall be deemed for purposes of this Agreement to have been distributed to the Person in respect of which such amount was so deducted and withheld.

ARTICLE 4
ACCESS TO INFORMATION; FINANCIAL STATEMENTS

Section 4.1 Restrictions on Disclosure of Information.

(a) Generally. Without limiting any rights or obligations under any other existing or future agreement between the Parties and/or any other members of their respective Group relating to confidentiality, for ten (10) years after the Distribution each Party shall, and each Party shall cause its respective Group Members and use its reasonable best efforts to cause its Representatives to, hold in confidence, with at least the same degree of care that applies to Ikaria's confidential and proprietary Information pursuant to policies in effect as of the Distribution Date, all confidential and proprietary Information concerning the other Group that is either in its possession as of the Distribution or furnished by the other Group or its respective Representatives at any time pursuant to this Agreement, any Ancillary Document or the transactions contemplated hereby or thereby. Notwithstanding the foregoing, each Party, its respective Group Members and its Representatives, may disclose such Information to the extent that such Party can demonstrate that such Information is or was (i) in the public domain other

than by the breach of this Agreement or by breach of any other agreement between or among the Parties and/or any of their respective Group Members relating to confidentiality or (ii) lawfully acquired from a third Person on a non-confidential basis or independently developed by, or on behalf of, such Party by Persons who do not have access to, or descriptions of, any such Information. Each Party shall maintain, and shall cause its respective Group Members and use its reasonable best efforts to cause its Representatives to maintain, policies and procedures, and develop such further policies and procedures as shall from time to time become necessary or appropriate, to ensure compliance with this Section 4.1. Nothing in this Article 4 shall be deemed

to limit INO Therapeutics' or Pulse Technologies' exercise of their respective rights under the R&DCo Cross License Agreement.

(b) Disclosure of Third-Person Information. R&DCo acknowledges that it and other R&DCo Group Members may have in its or their possession confidential or proprietary Information of third Persons that was received under confidentiality or non-disclosure agreement with such third Person while part of Ikaria. R&DCo shall, and R&DCo shall cause its respective Group Members and use its reasonable best efforts to cause its and their respective Representatives to, hold in confidence the confidential and proprietary Information of third Persons to which any R&DCo Group Member has access, in accordance with the terms of any agreements entered into prior to the Distribution between Ikaria Group Members (whether acting through, on behalf of, or in connection with, the R&DCo Business) and such third Persons.

Section 4.2 Protective Arrangements. If either Party or any of its respective Group Members or Representatives becomes required to or has received a request to make a Required Disclosure of any Information that it is otherwise obligated to hold in confidence pursuant to Section 4.1, such Party shall notify the other Party prior to disclosing or providing such Information and shall cooperate at the expense of such other Party in seeking any reasonable protective arrangements (including by seeking confidential treatment of such Information) requested by such other Party (it being understood that, with respect to the Required Disclosure of any Information of a third Person described in Section 4.1(b), Ikaria hereby authorizes the applicable R&DCo Group Members to notify the applicable third Person to satisfy any applicable review or consent requirements or to arrange for any appropriate protective arrangements). Subject to the foregoing, the Party that is (or whose Group Members are) required or requested to make such Required Disclosure may thereafter disclose or provide Information to the extent required or requested (as contemplated by the definition of "Required Disclosure"); provided that, upon request, such Party (or its Group Member) provides the other Party with a copy of the Information so disclosed in the format so disclosed. Notwithstanding the foregoing, a Party or its respective Group Members may make any public disclosure it believes in good faith is required by applicable securities laws, regulations or stock market rules, provided that such Party or Group Member uses reasonable efforts to advise the other Party of such disclosure and provide the other Party with a copy of the proposed disclosure prior to making the disclosure.

Section 4.3 Provision of Corporate Records; Access to Information.

(a) As soon as practicable following the date hereof, to the extent not effectuated prior to the date hereof and subject to the provisions of this Section 4.3(a), Ikaria and R&DCo shall transition (i) to R&DCo all R&DCo Books and Records in the possession of any Ikaria Group Member, and (ii) to Ikaria all Ikaria Books and Records in the possession of any R&DCo Group Member. The foregoing shall be limited by the following:

(i) Each Party may retain copies of books and records delivered to the other, subject to such Party's compliance with confidentiality obligations set forth in Section 4.1 with respect to the Information contained in such books and records.

(ii) Neither Party shall be required to deliver to the other books and records or portions thereof that are (A) the subject of a confidentiality agreement between such Party and a third Person that prohibits disclosure to the other Party (provided, however, that if requested by the other Party, such Party shall use reasonable best efforts to seek a waiver of or other relief from such confidentiality agreement, it being understood that no Party shall be obligated to pay any consideration (or otherwise incur any Liability or obligation) therefor to any third Person from whom any such waiver is sought (unless such Party is fully reimbursed or otherwise made whole by the requesting Party)), or (B) prohibited from disclosure under applicable Law.

(b) During the Retention Period, each Party shall cooperate with and afford, and shall cause its respective Group Members and use its reasonable best efforts to cause its Representatives to cooperate with and afford, to the other Party reasonable and timely access during normal business hours upon reasonable advance request to all Information (other than Information which is (i) protected from disclosure by the attorney-client privilege or work product doctrine, (ii) proprietary in nature, (iii) the subject of a confidentiality agreement between such Party and a third Person that prohibits disclosure to the other Party (provided, however, that if requested by the other Party, such Party shall use reasonable best efforts to seek a waiver of or other relief from such confidentiality agreement, it being understood that no Party shall be obligated to pay any consideration (or otherwise incur any Liability or obligation) therefor to any third Person from whom any such waiver is sought (unless such Party is fully reimbursed or otherwise made whole by the requesting Party)), or (iv) prohibited from disclosure under applicable Law (any such Information described in the foregoing clauses (i) through (iv), "Restricted Information")) owned by such Party or one of its Group Members or within such Party's or any of its respective Group Member's or Representative's possession which is created prior to the Distribution Date and which relates to the requesting Party's (the "Requestor") business, assets or liabilities, and such access is reasonably required by the Requestor (A) to comply with requirements imposed on the Requestor by any Law or Governmental Authority (including to make any Required Disclosure), (B) for use in preparation for or the prosecution or defense of any Action (except for any Action between the Parties or any of their respective Group Members), (C) to satisfy audit, accounting, Tax or similar requirements, (D) to obtain insurance, or (E) to comply with the Requestor's obligations under this Agreement or any Ancillary Document. As used in this Agreement, "access" shall mean the obligation of a Party in possession of Information (the "Possessor") requested by the Requestor to exert reasonable best efforts to locate all requested Information that is owned and/or possessed by Possessor or any respective Group Members or Representatives. The Possessor, at its own expense, shall conduct a diligent search designed to identify all requested Information and shall collect all such Information for inspection by the Requestor during normal business hours at the Possessor's place of business. Subject to such confidentiality and/or security obligations as the Possessor may reasonably deem necessary, the Requestor may have all requested Information duplicated at Requestor's expense. Alternatively, the Possessor may choose to deliver, at the Requestor's expense, all requested Information to the Requestor in the form requested by the Requestor. The Possessor shall notify the Requestor in writing at the time of delivery if such Information is to be returned to the Possessor. In such case, the Requestor shall return such Information when no longer needed to the Possessor at the Possessor's expense. In connection with providing Information pursuant to this Section 4.3, each Party shall, upon the request of the other Party and

upon reasonable advance notice, make available during normal business hours its respective employees (and those employees of its respective Group Members and Representatives, as applicable) to the extent that they are reasonably necessary to discuss and explain all requested Information with and to the Requestor.

Section 4.4 Record Retention. During the Retention Period, R&DCo shall, and R&DCo shall cause each of the other R&DCo Group Members to, adopt and comply with a record retention policy with respect to Information owned by or in the possession of the R&DCo Group and which is created prior to the Distribution Date that is no less stringent than Ikaria's record retention policy in effect as of the Distribution Date. During the Retention Period, Ikaria shall, and shall cause each of the other Ikaria Group Members to, maintain a record retention policy with respect to Information owned by or in the possession of the Ikaria Group and which is created prior to the Distribution Date that is no less stringent than Ikaria's record retention policy in effect as of the Distribution. Each Party shall, at its sole cost and expense, preserve and retain all Information in its respective possession or control that the other Party has the right to access pursuant to Section 4.3 or that it is required to preserve and retain in accordance with such record retention policy for a minimum of seven years or for any longer period as may be required by (a) any Governmental Authority, (b) any litigation matter, (c) applicable Law, or (d) any Ancillary Document (as applicable, the "Retention Period"). If either Party wishes to dispose of any Information that it is obligated to retain under this Section 4.4 prior to the expiration of the Retention Period, then that Party shall first provide 45 days' written notice to the other Party, and the other Party shall have the right, at its option but at the expense of the Party that desires to dispose of such Information, upon prior written notice within such 45-day period, to take possession of such Information within 90 days after the date of the notice provided pursuant to this Section 4.4. Written notice of intent to dispose of such Information shall include a description of the Information in detail sufficient to allow the other Party to reasonably assess its potential need to retain such materials.

Section 4.5 Production of Witnesses. After the Distribution, each Party shall use reasonable best efforts, and shall cause each of its respective Group Members to use reasonable best efforts, to make available to each other, upon written request, its past and present Representatives as witnesses to the extent that any such Representatives may reasonably be required (giving consideration to the business demands upon such Representatives) in connection with preparing for, defending or prosecuting any legal, administrative or other proceedings in which the requesting Party may from time to time be involved. The obligation to provide witnesses pursuant to this Section 4.5 is intended to be interpreted in a manner so as to facilitate cooperation and shall include the obligation to provide as witnesses inventors and other officers without regard to whether the witness or the employer of the witness could assert a possible business conflict. In connection with any matter contemplated by this Section 4.5 or any Information provided under Section 4.3 for use in a litigation, the Parties will enter into a mutually acceptable joint defense agreement so as to maintain to the extent practicable any applicable attorney-client privilege or work product immunity of any member of any Group.

Section 4.6 Financial Statements and Accounting.

(a) The Parties agree to use their reasonable best efforts to complete, with the assistance of the Parties' respective independent auditors, the Required R&DCo Financial Statements as promptly as practicable following the Distribution Date. For the avoidance of doubt, R&DCo (in consultation with its independent auditors) shall control the preparation and content of the Required R&DCo Financial Statements. In furtherance of the covenant set forth in the first sentence of this Section 4.6(a), Ikaria (i) shall and shall cause its Group Members and use its reasonable best efforts to cause its Representatives to provide access to all Information and (ii) shall use reasonable best efforts to cause its independent auditors to provide all assistance, in each case, as is reasonably required for purposes of completing the Required R&DCo Financial Statements in a timely manner.

(b) Without limiting the generality of Section 4.3 above, each Party agrees, on a timely basis, during normal business hours and upon reasonable advance request of the other Party, to provide to the other Party and its Representatives and/or to provide the other Party and its Representatives with access to, all Information reasonably required in connection with (A) the closing of the books and the preparation and audit of each Party's financial statements as of and for the year ended December 31, 2013, and (B) if and to the extent required under applicable Law: (I) each Party's management assessment of internal controls over financial reporting as of December 31, 2013, (II) the audit thereof by such Party's independent auditors and (III) each Party's management assessment of disclosure controls and procedures as of December 31, 2013 (the items described in the foregoing subclauses (I) through (III), the "Control Assessments and Audit"). Without limiting the generality of the foregoing, each Party will provide all required financial and other Information with respect to itself and its Group Members to its independent auditors within a sufficient and reasonable time and in reasonably sufficient detail to permit its independent auditors to take all steps and perform all reviews necessary to provide sufficient assistance to the other Party's independent auditors with respect to Information to be included or contained in the other Party's annual financial statements and to permit such other Party's independent auditors and management to complete the Control Assessments and Audit, if and to the extent required under applicable Law. Notwithstanding the foregoing, this Section 4.6(b) shall not require either Party to provide the other Party with any Restricted Information.

(c) Each Party (the "First Party") authorizes its respective independent auditors (the "First Party's Auditors") to make available to the other Party's independent auditors (the "Other Party's Auditors") both the personnel who performed, or are performing, the annual audit of the First Party for the year ended December 31, 2013 and work papers related to such annual audit, within a reasonable time prior to the First Party's Auditors' opinion date, so that the Other Party's Auditors are able to perform the procedures they consider necessary to take responsibility for the work of the First Party's Auditors as it relates to the Other Party's Auditors' report on the other Party's annual financial statements for the year ended December 31, 2013, all within sufficient time to enable the other Party to meet its timetable for the issuance and dissemination of such financial statements.

Section 4.7 Reimbursement. Unless otherwise provided in this Article 4, each Party providing access to Information or witnesses to the other Party pursuant to Sections 4.3, 4.4, 4.5

or 4.6(a) or (b) shall be entitled to receive from the receiving Party, upon the presentation of invoices therefor, payment for all reasonable costs and expenses (including allocated compensation, salary and overhead expenses) as may be reasonably incurred in providing such Information or witnesses. In furtherance of the foregoing, (a) R&DCo shall be liable for reimbursing Ikaria for the reasonable costs and expenses of Ikaria's Auditors incurred by Ikaria in connection with the preparation of the Required R&DCo Financial Statements and (b) Ikaria shall be liable for reimbursing R&DCo for the reasonable costs and expenses of R&DCo's Auditors incurred by R&DCo in connection with the assistance provided by the R&DCo Auditors pursuant to Section 4.6(c).

Section 4.8 Other Agreements Regarding Access to Information. The rights and obligations of the Parties under this Article 4 are subject to any specific limitations, qualifications or additional provisions on the sharing, exchange or confidential treatment of Information set forth in this Agreement or any Ancillary Document.

Section 4.9 Limitations of Liability. Except as otherwise provided in Article 6, no Person shall have any Liability in the event that any Information exchanged or provided pursuant to this Agreement is found to be inaccurate in the absence of willful misconduct by the Person providing such Information.

ARTICLE 5
ADDITIONAL COVENANTS AND OTHER MATTERS

Section 5.1 Further Assurances. Subject to the terms and conditions set forth elsewhere in this Agreement and the Ancillary Documents:

(a) each Party shall use reasonable best efforts to take, or cause to be taken, all actions and to do, or cause to be done, all things reasonably necessary, proper or advisable to consummate and make effective the Separation, the Distribution and the other transactions contemplated hereby and by the Ancillary Documents, including using reasonable best efforts to, and, except to the extent restricted by Law, cooperating with the other Party to (i) obtain all Consents from any Governmental Authority or other third party necessary or advisable in connection with the consummation of the Separation, the Distribution and the other transactions contemplated hereby or by any Ancillary Document, (ii) avoid any Action by any Governmental Authority in connection with the consummation of the Separation, the Distribution and the other transactions contemplated hereby or by any Ancillary Document, and (iii) defend any Actions, whether judicial or administrative, challenging this Agreement or the consummation of the transactions contemplated hereby, including seeking to have any stay or temporary restraining order entered by any court or other Governmental Authority vacated or reversed;

(b) at the request and expense of R&DCo, Ikaria shall execute and deliver, and shall cause applicable Ikaria Group Members to execute and deliver, to R&DCo and/or applicable R&DCo Group Members such other instruments of transfer, conveyance, assignment, substitution and confirmation and take such other actions as R&DCo may reasonably deem necessary or desirable in order to (i) transfer, convey and assign to R&DCo and the other R&DCo Group Members, as applicable, the R&DCo Assets, (ii) put R&DCo and the other

R&DCo Group Members, as applicable, in actual possession and operating control thereof and (iii) permit R&DCo and the other R&DCo Group Members, as applicable, to exercise all rights with respect thereto;

(c) at the request and expense of Ikaria, R&DCo shall execute and deliver, and shall cause applicable R&DCo Group Members to execute and deliver, to Ikaria and/or applicable Ikaria Group Members such other instruments of transfer, conveyance, assignment, substitution and confirmation and take such other actions as Ikaria may reasonably deem necessary or desirable in order to (i) transfer, convey and assign to Ikaria and the other Ikaria Group Members, as applicable, the Ikaria Assets, (ii) put Ikaria and the other Ikaria Group Members, as applicable, in actual possession and operating control thereof and (iii) permit Ikaria and the other Ikaria Group Members, as applicable, to exercise all rights with respect thereto;

(d) at the request and expense of Ikaria, R&DCo shall execute and deliver, and shall cause the applicable R&DCo Group Members to execute and deliver, to Ikaria and/or applicable Ikaria Group Members all instruments, assumptions, novations, undertakings, substitutions or other documents and take such other action as Ikaria may reasonably deem necessary or desirable in order to ensure that R&DCo and the other R&DCo Group Members fully and unconditionally assume and discharge the R&DCo Liabilities as contemplated under this Agreement, the Ancillary Documents or any document in connection herewith or therewith; and

(e) at the request and expense of R&DCo, Ikaria shall execute and deliver, and shall cause the applicable Ikaria Group Members to execute and deliver, to R&DCo and/or applicable R&DCo Group Members all instruments, assumptions, novations, undertakings, substitutions or other documents and take such other action as R&DCo may reasonably deem necessary or desirable in order to ensure that Ikaria and the other Ikaria Group Members fully and unconditionally assume and discharge the Ikaria Liabilities as contemplated under this Agreement, the Ancillary Documents or any document in connection herewith or therewith.

Section 5.2 Performance. Ikaria shall cause to be performed, and hereby guarantees the performance of, all actions, agreements and obligations set forth in this Agreement or in any Ancillary Document to be performed by any Ikaria Group Member. R&DCo shall cause to be performed, and hereby guarantees the performance of, all actions, agreements and obligations set forth in this Agreement or in any Ancillary Document to be performed by any R&DCo Group Member. Each Party further agrees that it shall cause its other Group Members not to take any action or fail to take any action inconsistent with such Party's obligations under this Agreement, any Ancillary Document or the transactions contemplated hereby or thereby.

Section 5.3 [Intentionally Omitted.]

Section 5.4 Insurance Matters. R&DCo acknowledges and agrees that from and after the Distribution Date (a) no Ikaria Group Member shall purchase or maintain, or cause to be purchased or maintained, any insurance policy for the protection of R&DCo or any R&DCo Group Member or any of their respective directors and officers, and (b) the R&DCo Group shall

purchase insurance coverage sufficient to protect its interests and to comply with the requirements of the R&D Cross-License Agreement.

Section 5.5 Signs; Use of Names. Except as otherwise contemplated by this Agreement or any Ancillary Document, prior to the Distribution Date, the Parties shall remove (or, if necessary, on an interim basis, cover up) any and all exterior and interior signs and identifiers on any R&DCo Asset that refer or pertain to any Ikaria Group Member or the Ikaria Business, or on any Ikaria Asset that refer or pertain to R&DCo or the R&DCo Business. After the Distribution, (a) no R&DCo Group Member shall use or display any Ikaria Name, or any variations thereof, or other Trademarks using any of such names or otherwise owned by or licensed to any Ikaria Group Member that have not been assigned or licensed to such R&DCo Group Member, and (b) no Ikaria Group Member shall use or display any Trademarks owned by or licensed to any R&DCo Group Member that have not been assigned or licensed to such Ikaria Group Member, without the prior written consent of the other Party; provided, that notwithstanding the foregoing, nothing contained in this Agreement shall prevent either Party from using the other's name in public filings with Governmental Authorities, materials intended for distribution to either Party's stockholders or members or any other communication in any medium that describes the relationship between the Parties or the respective businesses of the Parties, including materials distributed to employees relating to the transition of employee benefit plans and materials used in connection with any Post-Distribution Financing.

Section 5.6 Indemnification of R&DCo Officers and Directors. From the Distribution Date until the sixth anniversary of the Distribution Date, R&DCo shall fulfill and honor in all respects its obligations pursuant to its constitutive documents or other agreements providing for the indemnification, advancement of expenses and exculpation from liability of each person who is now, or has been at any time prior to the date of this Agreement, or who becomes prior to the Distribution Date, a director or officer of R&DCo or any of its Subsidiaries, but only to the extent not fully covered by insurance maintained and paid for by R&DCo, it being intended that an insurer who would otherwise be obligated to pay any such claim shall not be relieved of the responsibility with respect thereto or, solely by virtue of the indemnification provision hereof, have any subrogation rights with respect thereto.

ARTICLE 6
INDEMNIFICATION; MUTUAL RELEASE

Section 6.1 Indemnification by R&DCo Group. Subject to the provisions hereof, R&DCo shall, and shall cause each other entity in the R&DCo Group as of the Distribution to, jointly and severally, indemnify, defend and hold harmless each Ikaria Group Member, each of their respective past and present Affiliates, directors, officers, employees and other Representatives, and each of their respective permitted successors and assigns (collectively, the "Ikaria Indemnified Parties") from and against any and all Damages incurred or suffered by the Ikaria Indemnified Parties arising or resulting from the following, whether such Damages arise or accrue prior to, at or following the Distribution:

(a) the failure of R&DCo or any other R&DCo Group Member or any other Person to pay, perform or otherwise properly discharge any of the R&DCo Liabilities in accordance with their respective terms;

(b) the R&DCo Business or any R&DCo Liability;

(c) the matters set forth on Schedule 6.1(c); and

(d) any breach by R&DCo or any R&DCo Group Member of this Agreement or any Ancillary Document.

Notwithstanding anything to the contrary herein, in no event will any Ikaria Indemnified Party have the right to seek indemnification from any R&DCo Group Member with respect to any claim or demand against any Person in the Ikaria Group for satisfaction of any of the Ikaria Liabilities.

Section 6.2 Indemnification by Ikaria Group. Subject to the provisions hereof, Ikaria shall, and shall cause each other entity in the Ikaria Group as of the Distribution to, jointly and severally, indemnify, defend and hold harmless each R&DCo Group Member, each of their respective past and present directors, officers and employees, and each of their respective permitted successors and assigns (collectively, the "R&DCo Indemnified Parties") from and against any and all Damages incurred or suffered by the R&DCo Indemnified Parties arising or resulting from the following, whether such Damages arise or accrue prior to, at or following the Distribution:

(a) the failure of Ikaria or any other Ikaria Group Member or any other Person to pay, perform or otherwise properly discharge any of the Ikaria Liabilities in accordance with their respective terms;

(b) the Ikaria Business or any Ikaria Liability; and

(c) any breach by Ikaria or any Ikaria Group Member of this Agreement or any Ancillary Document.

Notwithstanding anything to the contrary herein, in no event will any R&DCo Indemnified Party have the right to seek indemnification from any Ikaria Group Member with respect to any claim or demand against any Person in the R&DCo Group for satisfaction of any of the R&DCo Liabilities.

Section 6.3 Claim Procedure.

(a) Claim Notice. A Person that seeks indemnity under this Article 6 (an "Indemnified Party") shall give written notice (a "Claim Notice") to the Party from whom indemnification is sought (an "Indemnifying Party"), whether the Damages sought arise from matters solely between the Parties or from Third-Party Claims. With respect to matters solely between the Parties, the Claim Notice must contain (i) a description and, to the extent known, estimated amount (the "Claimed Amount") of any Damages incurred or reasonably expected to

be incurred by the Indemnified Party, (ii) a reasonable explanation of the basis for the Claim Notice to the extent of facts then known by the Indemnified Party, and (iii) a demand for payment of those Damages. No delay or deficiency on the part of the Indemnified Party in so notifying the Indemnifying Party shall relieve the Indemnifying Party of any Liability or obligation hereunder except to the extent that the Indemnifying Party is prejudiced by such delay or deficiency or the amount of any associated Damages is increased by such delay or deficiency.

(b) Response to Notice of Claim. Within 30 days after delivery of a Claim Notice with respect to a matter solely between the Parties, the Indemnifying Party shall deliver to the Indemnified Party a written response in which the Indemnifying Party shall either: (i) agree that the Indemnified Party is entitled to receive all of the Claimed Amount and, in which case, the Indemnifying Party shall pay the Claimed Amount in accordance with a payment and distribution method reasonably acceptable to the Indemnified Party; or (ii) dispute that the Indemnified Party is entitled to receive all or any portion of the Claimed Amount.

(c) Contested Claims. In the event that the Indemnifying Party disputes the Claimed Amount, such dispute shall be resolved in accordance with Article 7. Upon ultimate resolution thereof, the Parties shall take such actions as are reasonably necessary to comply therewith.

(d) Third-Party Claims.

(i) In the event that the Indemnified Party receives notice or otherwise learns of the assertion by a Person who is not a member of either Group of any claim or the commencement of any Action (any such claim or Action, a "Third-Party Claim") with respect to which the Indemnifying Party may be obligated to provide indemnification under this Article 6, the Indemnified Party shall give written notification to the Indemnifying Party of the Third-Party

Claim. Such notification shall be given promptly after receipt by the Indemnified Party of notice of such Third-Party Claim, shall be accompanied by reasonable supporting documentation submitted by such third party (to the extent then in the possession of the Indemnified Party) and shall describe in reasonable detail (to the extent known by the Indemnified Party) the facts constituting the basis for such Third-Party Claim and the amount of the claimed Damages (to the extent they can be reasonably estimated by the Indemnified Party based on available information); provided, however, that no delay or deficiency on the part of the Indemnified Party in so notifying the Indemnifying Party shall relieve the Indemnifying Party of any Liability or obligation hereunder except to the extent that the Indemnifying Party is prejudiced by such delay or deficiency or to the extent the amount of any associated Damages is increased by such delay or deficiency. If, and for so long as, (A) the Indemnifying Party notifies the Indemnified Party as soon as practicable, but in no event later than 30 days, after delivery of such notification that the Indemnifying Party does not dispute the Indemnifying Party's obligation to indemnify hereunder and desires to defend the Indemnified Party against such Third-Party Claim, and (B) the Third-Party Claim (I) does not involve criminal liability or any admission of wrongdoing, (II) does not seek equitable relief or any other material non-monetary remedy against the Indemnified Party, (III) does not involve a claim which the Indemnified Party reasonably believes would have a material and adverse effect on the Indemnified Party's business or (IV) is not one in which the Indemnifying Party is also a party and, in the opinion of the Indemnified Party's outside counsel,

joint representation would be inappropriate or there may be legal defenses available to the Indemnified Party which are different from or additional to those available to the Indemnifying Party, then except as hereinafter provided, such Indemnifying Party shall have the right to defend against such Third-Party Claim by appropriate proceedings with legal counsel reasonably acceptable to the Indemnified Party, which proceedings shall be promptly settled or diligently prosecuted by the Indemnifying Party to a final conclusion. During any period in which the Indemnifying Party has not so assumed control of such defense, the Indemnified Party shall control such defense and is hereby authorized (but not obligated) prior to and during such period to file any motion, answer or other pleading and to take any other action which the Indemnified Party shall deem necessary or appropriate to protect the Indemnified Party's interests.

(ii) The Party not controlling such defense (the "Non-controlling Party") may participate therein at its own expense; provided, however, that if the Indemnifying Party assumes control of such defense and the Indemnified Party concludes, upon advice of counsel, that the Indemnifying Party and the Indemnified Party have conflicting interests or different defenses available with respect to such Third-Party Claim, the reasonable fees and expenses of one additional counsel to the Indemnified Party shall be considered "Damages" for purposes of this Agreement. The Party controlling such defense (the "Controlling Party") shall keep the Non-controlling Party reasonably advised of the status of such Third-Party Claim and the defense thereof and shall consider in good faith recommendations made by the Non-controlling Party with respect thereto. Subject to receiving a confidentiality undertaking from the Controlling Party and any redactions that the Non-controlling Party determines are advisable for purposes of maintaining privilege, the Non-controlling Party shall furnish the Controlling Party with such Information as it may have with respect to such Third-Party Claim (including copies of any summons, complaint or other pleading which may have been served on such Party and any written claim, demand, invoice, billing or other document evidencing or asserting the same) and shall otherwise cooperate with and assist the Controlling Party in the defense of such Third-Party Claim.

(iii) The Indemnifying Party shall not agree to any settlement of, or the entry of any judgment arising from, any such Third-Party Claim without the prior written consent of the Indemnified Party, which consent shall not be unreasonably withheld, conditioned or delayed; provided, however, that the consent of the Indemnified Party shall not be required if (A) such settlement involves only the payment of monetary damages and the Indemnifying Party agrees in writing to pay any amounts payable pursuant to such settlement or judgment, (B) such settlement or judgment includes a full, complete and unconditional release of the Indemnified Party and its Affiliates from further Liability and (C) such settlement involves no admission of wrongdoing by the Indemnified Party or its Affiliates. The Indemnified Party shall not agree to any settlement of, or the entry of any judgment arising from, any such Third-Party Claim without the prior written consent of the Indemnifying Party, which consent shall not be unreasonably withheld, conditioned or delayed.

Section 6.4 Survival; Limitations.

(a) All covenants and agreements of the Parties contained in this Agreement shall survive each of the Separation and the Distribution. The rights and obligations of Ikaria,

R&DCo and each of their respective Indemnified Parties under this Agreement shall survive the sale, assignment or other transfer by any Party of any Assets, businesses or Liabilities, or the change of form or change of control of any Party.

(b) The amount of any Damages for which indemnification is provided under this Agreement shall be net of any amounts actually recovered by the Indemnified Party from any third Person (including amounts actually recovered under insurance policies) with respect to such Damages (net of all costs and expenses, including collection costs and any retention amounts or increases in premiums). An insurer who would otherwise be obligated to pay any claim shall not be relieved of the responsibility with respect thereto or, solely by virtue of the indemnification provision hereof, have any subrogation rights with respect thereto.

(c) Unless otherwise required by applicable Law, the Parties agree that any indemnification payments made by one Party to another Party pursuant to this Agreement after the Distribution shall, to the extent permissible under applicable law, be treated for all Tax and financial accounting purposes as contributions or distributions, as appropriate, made immediately prior to the Distribution. If it is determined that the receipt or accrual of any indemnification payment is subject to Tax, such payment shall be increased so that the amount of such increased payment reduced by the amount of all Taxes payable with respect to the receipt thereof (but taking into account all correlative Tax deductions resulting from the payment of the applicable Damages that gave rise to such indemnification payment) shall equal the amount of the payment which the Party receiving such payment would otherwise be entitled to receive pursuant to this Agreement.

(d) Notwithstanding the joint and several indemnification obligations of each Group as set forth in Sections 6.1 and 6.2, the Parties agree that the indemnification obligation of any Ikaria Group Member or R&DCo Group Member, as applicable, for Damages shall be satisfied by a direct payment from Ikaria or R&DCo, as applicable, to the other Party irrespective of which Group Member is found liable for Damages.

(e) Notwithstanding anything to the contrary in this Agreement, to the extent the Employee Matters Agreement specifically provides indemnification with respect to certain employee-related R&DCo Liabilities, the Employee Matters Agreement shall govern with respect to that indemnification. To the extent indemnification is not provided in the Employee Matters Agreement, the indemnification terms of this Agreement shall govern.

(f) NOTWITHSTANDING ANY OTHER PROVISION OF THIS AGREEMENT OR ANY ANCILLARY DOCUMENT TO THE CONTRARY, IN NO EVENT SHALL EITHER PARTY OR ANY OF ITS GROUP MEMBERS BE LIABLE FOR ANY SPECIAL, INCIDENTAL, INDIRECT,

COLLATERAL, CONSEQUENTIAL OR PUNITIVE DAMAGES OR LOST PROFITS SUFFERED BY AN INDEMNIFIED PARTY, HOWEVER CAUSED AND ON ANY THEORY OF LIABILITY, IN CONNECTION WITH ANY DAMAGES ARISING HEREUNDER OR THEREUNDER; PROVIDED, HOWEVER, THAT TO THE EXTENT AN INDEMNIFIED PARTY IS REQUIRED TO PAY ANY SPECIAL, INCIDENTAL, INDIRECT, COLLATERAL, CONSEQUENTIAL OR PUNITIVE DAMAGES OR LOST PROFITS TO A PERSON WHO IS NOT A MEMBER OF EITHER GROUP IN

CONNECTION WITH A THIRD-PARTY CLAIM, OR INCURS ANY SPECIAL, INCIDENTAL, INDIRECT, COLLATERAL, CONSEQUENTIAL OR PUNITIVE DAMAGES OR LOST PROFITS AS A RESULT OF RESTRICTIONS OR LIMITATIONS IMPOSED ON AN INDEMNIFIED PARTY BY THE FDA AS A RESULT OF THE ACTS OR OMISSIONS OF (INCLUDING THE PROVISION OF ANY PRODUCT OR SERVICE BY) THE OTHER PARTY, SUCH DAMAGES SHALL CONSTITUTE DIRECT DAMAGES AND NOT SUBJECT TO THE LIMITATION SET FORTH IN THIS SECTION 6.4(f).

(g) Notwithstanding a Party's knowledge of any breach by the other Party of any representation, warranty, covenant or agreement contained in this Agreement or any Ancillary Document or any facts or circumstances potentially giving rise to a claim under this Agreement or any Ancillary Document (and regardless of how such Party shall have acquired such knowledge), such Party shall have the right to consummate the transactions provided for herein, and all of such Party's rights and remedies shall be preserved without regard to its knowledge of such information.

(i) Notwithstanding anything herein to the contrary, in no event shall any Indemnified Party be entitled to receive payment of Damages under this Agreement to the extent that such Indemnified Party has received payments in respect of such Damages pursuant to any rights to indemnification or otherwise under any Ancillary Documents.

Section 6.5 Non-Recourse. All claims or causes of action (whether in contract or in tort, in law or in equity) that may be based upon, arise out of or related to this Agreement or any Ancillary Document, or the negotiation, execution or performance of this Agreement or such Ancillary Document (including any representation or warranty made in connection with this Agreement or such Ancillary Document or as an inducement to enter into this Agreement or such Ancillary Document), may be made only against the Persons that are expressly identified as parties thereto. Except as provided for in Section 6.4, no Person who is not a named Party or is not named as a party to the applicable Ancillary Document, including any director, officer, employee, stockholder, member, Affiliate, agent or representative of any named Party or party to such Ancillary Document ("Non-Party Affiliates"), shall have any Liability (whether in contract or in tort, in law or in equity, or based upon any theory that seeks to impose Liability of an entity party against its owners or Affiliates) for any Liabilities arising under, in connection with or related to this Agreement or such Ancillary Document or for any claim based on, in respect of, or by reason of this Agreement or such Ancillary Document or its negotiation or execution; and each Party waives and releases all such Liabilities against any such Non-Party Affiliates. Non-Party Affiliates are expressly intended as third-party beneficiaries of this Section 6.5.

ARTICLE 7
DISPUTE RESOLUTION

Section 7.1 Disputes. Except as otherwise specifically provided in any Ancillary Document, the procedures for discussion, negotiation and mediation set forth in this Article 7 shall apply to all disputes, controversies or claims (whether arising in contract, tort or otherwise) that may arise out of or relate to, or arise under or in connection with this Agreement or the transactions contemplated hereby (including all actions taken in furtherance of the transactions

contemplated hereby on or prior to the date hereof), or the commercial or economic relationship of the parties relating hereto or thereto, between or among any Person in the Ikaria Group and any Person in the R&DCo Group.

Section 7.2 Escalation; Mediation.

(a) It is the intent of the Parties to use their respective reasonable best efforts to resolve expeditiously any dispute, controversy or claim between or among them with respect to the matters covered by this Agreement or any Ancillary Document that may arise from time to time on a mutually acceptable negotiated basis. In furtherance of the foregoing, any Party involved in a dispute, controversy or claim with respect to such matters (except any matters covered by the Transition Services Agreement) may deliver a notice (an "Escalation Notice") demanding an in person meeting involving representatives of the Parties at a senior level of management of the Parties (or if the Parties agree, of the appropriate strategic business unit or division within such entity). A copy of any such Escalation Notice shall be given to the General Counsel, or like officer or official, of each Party (which copy shall state that it is an Escalation Notice pursuant to this Agreement). Any agenda, location or procedures for such discussions or negotiations between the Parties may be established by the Parties from time to time; provided, however, that the Parties shall use their reasonable best efforts to meet within 30 days of the Escalation Notice.

(b) If the Parties are not able to resolve the dispute, controversy or claim through the escalation process referred to above, then the matter shall be referred to mediation. The Parties shall retain a mediator to aid the Parties in their discussions and negotiations by informally providing advice to the Parties. Any opinion expressed by the mediator shall be strictly advisory and shall not be binding on the Parties, nor shall any opinion expressed by the mediator be admissible in any other proceeding. The mediator may be chosen from a list of mediators previously selected by the Parties or by other agreement of the Parties. Costs of the mediation shall be borne equally by the Parties, except that each Party shall be responsible for its own expenses. Mediation shall be a prerequisite to the commencement of any Action by either Party against any member of the other Party's Group.

Section 7.3 Court Actions. In the event that any Party, after complying with the provisions set forth in Section 7.2 above, desires to commence an Action, such Party, subject to Section 8.2, may submit the dispute, controversy or claim (or such series of related disputes, controversies or claims) to any court of competent jurisdiction as set forth in Section 8.2.

ARTICLE 8
MISCELLANEOUS

Section 8.1 Governing Law. The internal Laws of the State of Delaware (without giving effect to any choice or conflict of law provision or rule, whether of the State of Delaware or any other jurisdiction, that would cause the application of Laws of any jurisdiction other than those of the State of Delaware) shall govern the construction, interpretation and other matters arising out of or in connection with this Agreement and, unless expressly provided therein, each

Ancillary Document, and each of the exhibits and schedules hereto and thereto (whether arising in contract, tort, equity or otherwise).

Section 8.2 Jurisdiction. If any dispute, controversy or claim arises out of or in connection with this Agreement or any Ancillary Document, except as expressly contemplated by any Ancillary Document, the Parties irrevocably (and the Parties shall cause each other member of their respective Group to irrevocably) (a) consent and submit to the exclusive jurisdiction of the Court of Chancery of the State of Delaware, New Castle County, or, if that court does not have jurisdiction, a federal court sitting in Wilmington, Delaware, (b) waive any objection to that choice of forum based on venue or to the effect that the forum is not convenient, and (c) WAIVE TO THE FULLEST EXTENT PERMITTED BY LAW ANY RIGHT TO TRIAL OR ADJUDICATION BY JURY. Either Party may make service on the other Party by sending or delivering a copy of the process to the other Party at the address and in the manner provided for the giving of notices in Section 8.3. Nothing in this Section 8.2, however, shall affect the right to serve legal process in any other manner permitted by Law.

Section 8.3 Notices. All notices and other communications under this Agreement or any Ancillary Document shall be in writing and shall be deemed duly delivered (a) four Business Days after being sent by registered or certified mail, return receipt requested, postage prepaid, (b) one Business Day after being sent for next Business Day delivery, fees prepaid, via a reputable nationwide overnight courier service, or (c) on the date of confirmation of receipt (or, the first Business Day following such receipt if the date of such receipt is not a Business Day) of transmission by facsimile, in each case to the intended recipient as set forth below.

If to Ikaria:

INO Therapeutics LLC
Perryville III Corporate Park
53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel

If to AcquisitionCo:

Ikaria Acquisition Inc.
Perryville III Corporate Park
53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel (or Chief Executive Officer if there is no General Counsel)

If to R&DCo:

Bellerophon Therapeutics LLC
Perryville III Corporate Park
53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel (or Chief Executive Officer if there is no General Counsel)

With a copy (which shall not constitute notice) to:

New Mountain Partners II, L.P.
c/o New Mountain Capital, L.L.C.
787 Seventh Avenue, 49th Floor
New York, New York
Attention: Matthew Holt
Email: MHolt@newmountaincapital.com

Any party may give any notice or other communication hereunder using any other means (including personal delivery, messenger service, ordinary mail or electronic mail), but no such notice or other communication shall be deemed to have been duly given unless and until it actually is received by the party for whom it is intended. Any party may change the address to which notices and other communications hereunder are to be delivered by giving the other parties notice in the manner herein set forth.

Section 8.4 Binding Effect and Assignment. This Agreement and each Ancillary Document binds and benefits the Parties and their respective permitted successors and assigns. Neither Party may assign any of its rights or delegate any of its obligations under this Agreement or any Ancillary Document without the written consent of the other Party and any assignment or attempted assignment in violation of the foregoing shall be null and void. Notwithstanding the preceding sentence, either Party may, upon written notice, assign this Agreement and the Ancillary Documents (other than the R&D Cross License, which can only be assigned in accordance with its terms) in connection with a merger transaction in which such Party is not the surviving entity or the sale of all or substantially all of its assets; provided that the surviving party or acquirer in such transaction agrees in writing to assume and be bound by all of such Party's obligations hereunder.

Section 8.5 Severability. Any term or provision of this Agreement or any Ancillary Document that is invalid or unenforceable in any situation in any jurisdiction shall not affect the validity or enforceability of the remaining terms and provisions hereof or thereof or the validity or enforceability of the offending term or provision in any other situation or in any other jurisdiction. If the final judgment of a court of competent jurisdiction declares that any term or provision hereof is invalid or unenforceable, the Parties agree that the court making such determination shall have the power to limit the term or provision, to delete specific words or phrases, or to replace any invalid or unenforceable term or provision with a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision, and this Agreement shall be enforceable as so modified. In the

event such court does not exercise the power granted to it in the prior sentence, the Parties agree to replace such invalid or unenforceable term or provision with a valid and enforceable term or provision that shall achieve, to the extent possible, the economic, business and other purposes of such invalid or unenforceable term.

Section 8.6 Specific Performance. The Parties agree that irreparable damage would occur in the event that any provision of this Agreement or any Ancillary Document were not performed in accordance with its specific terms or were otherwise breached. It is accordingly agreed that either Party shall be entitled to an injunction or injunctions to prevent breaches of this Agreement or any Ancillary Document and to enforce specifically the terms and provisions of this Agreement or any Ancillary Document, in each case without posting a bond or undertaking, this being in addition to any other remedy to which they are entitled at law or in equity. Each of the Parties agrees that it shall not oppose the granting of an injunction, specific performance and other equitable relief on the basis that (a) the Party seeking such remedy has an adequate remedy at law or (b) an award of specific performance is not an appropriate remedy for any reason at law or equity.

Section 8.7 Entire Agreement. This Agreement, together with the Ancillary Documents and each of the exhibits and schedules appended hereto and thereto, constitutes the final agreement between the Parties, and is the complete and exclusive statement of the Parties' agreement on the matters contained herein and therein. All prior and contemporaneous negotiations and agreements between the Parties with respect to the matters contained herein and therein are superseded by this Agreement and the Ancillary Documents, as applicable. In the event of any conflict between (a) any provision in this Agreement, on the one hand, and (b) any specific provision in any Ancillary Document, on the other hand, pertaining to the subject matter of such Ancillary Document, the specific provisions in such Ancillary Document shall control over the provisions in this Agreement, as applicable.

Section 8.8 No Third-Party Beneficiaries. Except for Purchaser, which from and after the Closing (as defined in the Merger Agreement) shall be a third party beneficiary of this Agreement, and except as expressly set forth in Section 6.5 (which may be enforced directly by any Non-Party Affiliate), neither this Agreement nor any Ancillary Document is intended, or shall be deemed, to confer any rights or remedies upon any Person other than the Parties and their respective Group Members, successors and permitted assigns, to create any agreement of employment with any Person or to otherwise create any third-party beneficiary hereto or thereto.

Section 8.9 Counterparts and Signature. This Agreement may be executed in two counterparts (including by facsimile or by an electronic scan delivered by electronic mail), each of which shall be deemed an original but both of which together shall be considered one and the same agreement and shall become effective when counterparts have been signed by each Party and delivered to the other Party, it being understood that both Parties need not sign the same counterpart.

Section 8.10 Expenses. Except as otherwise expressly provided in this Agreement or any Ancillary Document, following the Distribution, each Party will be responsible for its own

fees, costs and expenses incurred in connection with the transactions contemplated by this Agreement and the Ancillary Documents.

Section 8.11 Amendment. The Parties may amend this Agreement or any Ancillary Document only by a written agreement signed by the Party intended to be bound by the amendment and that identifies itself as an amendment to this Agreement or such Ancillary Document, as applicable.

Section 8.12 Waiver. The Parties may waive a provision of this Agreement or an Ancillary Document only by a writing signed by the Party intended to be bound by the waiver. A Party is not prevented from enforcing any right, remedy or condition in the Party's favor because of any failure or delay in exercising any right or remedy or in requiring satisfaction of any condition, except to the extent that the Party specifically waives the same in writing. A written waiver given for one matter or occasion is effective only in that instance and only for the purpose stated. A waiver once given is not to be construed as a waiver for any other matter or occasion. Any enumeration of a Party's rights and remedies in this Agreement or any Ancillary Document is not intended to be exclusive, and a Party's rights and remedies are intended to be cumulative to the extent permitted by Law and include any rights and remedies authorized in law or in equity.

Section 8.13 Authority; R&DCo Assets. Each of the Parties represents to the other on behalf of itself and each of its other Group Members that (a) it and its Group Members have the corporate or other requisite power and authority to execute, deliver and perform this Agreement and each of the Ancillary Documents to which it is a party, (b) the execution, delivery and performance of this Agreement and each of the Ancillary Documents to which they are a party by it and its Group Members have been duly authorized by all necessary corporate or other action, (c) it and its Group Members have duly and validly executed and delivered this Agreement and each of the Ancillary Documents to which they are a party, and (d) this Agreement and each of the Ancillary Documents to which they are a party is a valid and binding obligation, enforceable against it or its applicable Group Member in accordance with its terms, subject to applicable bankruptcy, insolvency, reorganization, moratorium or other similar Laws affecting creditors' rights generally and general equitable principles. R&DCo, on behalf of itself and each of its other Group Members, represents to Ikaria that (i) none of the R&DCo Assets are used in or necessary for the Ikaria Business except as otherwise set forth on Schedule 8.13(a) hereto, (ii) R&DCo, together with its Subsidiaries, has, and immediately following the Distribution will have, cash and cash equivalents in an amount equal to or greater than $80 million less (x) the amount deposited in escrow by R&DCo pursuant to the terms of the Transition Services Agreement and (y) $10,000 in respect of the aggregate cash payments made by or on behalf of R&DCo to holders of Ikaria Stock Options who immediately prior to the Distribution were neither employees of Ikaria nor Accredited Investors, pursuant to the terms of the Employee Matters Agreement (collectively, the "R&DCo Option Payments"), and (iii) the transactions contemplated by this Agreement do not require the consent, approval or authorization of any Person (before giving effect to the provisions of Section 2.3) except as set forth on Schedule 8.13(b).

Section 8.14 Construction of Agreement.

(a) Where this Agreement or any Ancillary Document states that a Party "will" or "shall" perform in some manner or otherwise act or omit to act, it means that the Party is legally obligated to do so in accordance with this Agreement or such Ancillary Document, as applicable.

(b) The captions, titles and headings, and table of contents, included in this Agreement and the Ancillary Documents are for convenience only, and do not affect this Agreement's or such Ancillary Documents' construction or interpretation. When a reference is made in this Agreement or any Ancillary Document to an Article or a Section, exhibit or schedule, such reference shall be to an Article or Section of, or an exhibit or schedule to, this Agreement or such Ancillary Document, as applicable, unless otherwise indicated.

(c) When used in this Agreement or any Ancillary Document, the words "including," "includes," or "include" shall be deemed to be followed by the phrase "without limitation."

(d) Any reference in this Agreement or any Ancillary Document to the singular includes the plural where appropriate. Any reference in this Agreement or any Ancillary Document to the masculine, feminine or neuter gender includes the other genders where appropriate. Any reference in this Agreement or any Ancillary Document to any Person includes such Person's successors and assigns but, if applicable, only if such successors and assigns are permitted by this Agreement. For purposes of this Agreement, after the Distribution the R&DCo Business shall be deemed to be the business of R&DCo and the R&DCo Group, and all references made in this Agreement to R&DCo as a party which operates as of a time following the Distribution, shall be deemed to refer to all R&DCo Group Members as a single party where appropriate.

(e) Any agreement, instrument or statute defined or referred to in this Agreement, any Ancillary Document or in any agreement or instrument that is referred to in this Agreement or any Ancillary Document means such agreement, instrument or statue as from time to time amended, modified or supplemented, including (in the case of agreements or instruments) by waiver or consent and (in the case of statutes) by succession or comparable successor statutes and references to all attachments thereto and instruments incorporated therein.

(f) Any reference to any federal, state, local, or foreign Law shall be deemed also to refer to all rules and regulations promulgated thereunder, unless the context requires otherwise.

(g) Unless otherwise expressly specified in an Ancillary Document, all references in this Agreement or any Ancillary Document to "dollars" or "$" means United States Dollars. If any payment required to be made hereunder is denominated in a currency other than United States Dollars, such payment shall be made in United States Dollars and the amount thereof shall be computed using the exchange rate published by the Wall Street Journal on the date of payment (or if the Wall Street Journal is not published on such date, the last date prior thereto on which the Wall Street Journal was published).

(h) Unless otherwise expressly provided, wherever the consent of a Party is required or permitted herein, such consent may be withheld in such Party's sole and absolute discretion.

Section 8.15 Termination. This Agreement may be terminated at any time after the Distribution, by the mutual written consent of Ikaria and R&DCo. In the event of a termination of this Agreement pursuant to the foregoing sentence, neither Party shall have any liability of any kind to the other Party under this Agreement, except for any breach of this Agreement that occurs prior to such termination.

Section 8.16 Insurance. R&DCo, at its expense, shall hold and maintain in full force and effect a policy or policies of appropriate insurance coverage as reasonably determined by R&DCo, such insurance coverage (i) to include at a minimum products liability coverage and clinical trial liability coverage and (ii) to provide coverage amounts as are reasonable for the activities to be conducted by R&DCo but in no event less than $[**] of combined coverage for the products liability coverage and clinical trial liability coverage. Each policy of insurance maintained by R&DCo shall name Ikaria as an additional insured. All insurance shall be on an occurrence basis, and not on a claims made basis, except to the extent that occurrence basis insurance is not available on commercially reasonable terms. Each insurance policy shall provide for a waiver of the insurer's subrogation rights against R&DCo, to the extent such a waiver is permitted by the insurer. At the request of Ikaria, R&DCo shall provide Ikaria with documentary evidence of compliance with its obligations under this Section including a copy of its current insurance certification and full policy details, and provide Ikaria with notice of any change to such policies within thirty (30) days from the effective date of such change. The obligations of R&DCo under this Section 8.16 shall terminate at such time as Ikaria is no longer manufacturing and supplying nitric oxide delivery devices or nitric oxide for inhalation and corresponding placebo to any R&DCo Group Member pursuant to the Manufacturing and Supply Agreements.

(This space intentionally left blank)

IN WITNESS WHEREOF, each of Ikaria, R&DCo and ServicesCo has caused this Agreement to be executed on its behalf by a duly authorized officer on the date first set forth above.

IKARIA, INC.
a Delaware corporation

By: /s/ Anastasios Konidaris

Name: Anastasios Konidaris

Title: Chief Financial Officer

BELLEROPHON THERAPEUTICS LLC
a Delaware limited liability company

By: /s/ Daniel Tassé

Name: Daniel Tassé

Title: Chief Executive Officer

IKARIA ACQUISITION INC.
a Delaware corporation

By: /s/ Matthew M. Bennett

Name: Matthew M. Bennett

Title: Vice President & Secretary

[Signature Page to Separation and Distribution Agreement]

Schedule 1.1(a)

Ikaria Names

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
	United States	5	85/915,731	04/26/2013	Not Applicable	Not Applicable	INO Therapeutics LLC
	United States	10	85/915,748	04/26/2013	Not Applicable	Not Applicable	INO Therapeutics LLC
DSIR	United States	10	85/070,932	06/24/2010	4,003,732	07/26/2011	INO Therapeutics LLC
IKARIA	United States	5, 9, 10, 37, 42, 44	77/334,544	11/20/2007	3,778,583	04/20/2010	Ikaria, Inc.
FLEXTRANET	United States	42	86/062,864	09/12/2013	Not Applicable	Not Applicable	INO Therapeutics LLC
IKARIA	United States	5, 9, 10, 37, 42, 44	77/334,547	11/20/2007	3,778,584	04/20/2010	Ikaria, Inc.
IKARIA ADVANCING CRITICAL CARE	United States	5, 9, 10, 37, 42, 44	77/334,549	11/20/2007	3,778,585	04/20/2010	Ikaria, Inc.
IKARIA	United States	5, 10, 42	77/257,398	08/16/2007	3,758,037	03/09/2010	Ikaria, Inc.
INOBlender	United States	10	79/026,694	04/04/2006	3,242,739	05/15/2007	INO Therapeutics LLC
INOCAL	United States	1	75/124,073	06/24/1996	2,109,970	10/28/1997	INO Therapeutics LLC

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
INOCAL CADDY	United States	12	85/791,544	11/30/2012	4,389,669	08/20/2013	INO Therapeutics LLC
INOCART	United States	12	85/791,541	11/30/2012	Not Applicable	Not Applicable	INO Therapeutics LLC
INOMAX	United States	5	75/342,495	08/18/1997	2,185,947	09/01/1998	INO Therapeutics LLC
INOMAX	United States	10	79/026,770	04/04/2006	3,280,185	08/14/2007	INO Therapeutics LLC
INOMAX DSIR	United States	10	85/071,349	06/25/2010	4,003,735	07/26/2011	INO Therapeutics LLC
INOMAX TOTAL CARE	United States	37, 39, 41	85/737,439	09/25/2012	4,389,579	08/20/2013	INO Therapeutics LLC
INOPULSE	United States	10	85/031,549	05/06/2010	3,893,703	12/21/2010	INO Therapeutics LLC
INOTHERAPY	United States	37, 39, 41	75/751,044	06/24/1999	2,678,500	01/21/2003	INO Therapeutics

							LLC
INOVENT	United States	10	75/124,072	06/24/1996	2,100,392	09/23/1997	INO Therapeutics LLC
NICU-PET	United States	16, 41	85/836,260	01/30/2013	4,410,417	10/01/2013	INO Therapeutics LLC
LUCASSIN	United States	5	79/006,270	10/19/2004	3,085,607	04/25/2006	Acorus Therapeutics Limited
ONSTORVIS	United States	5	85/876474 Intent-to-use	03/14/2013	Not Applicable	Not Applicable	Ikaria Therapeutics LLC
TERLIVAZ	United States	5	85/876,469 Intent-to-use	03/14/2013	Not Applicable	Not Applicable	Ikaria Therapeutics LLC
IKARIA	United States	5	78/980,137	04/22/2005	3,443,089	06/03/2008	Ikaria, Inc.
IKARIA	United States	10, 42	78/614,854	04/22/2005	3,450,879	06/17/2008	Ikaria, Inc.
IKARIA	United States	10, 42	77/257,378	08/16/2007	3,811,308	06/29/2010	Ikaria, Inc.
IKARIA	United States	10, 42	77/285,095	09/20/2007	3,811,324	06/29/2010	Ikaria, Inc.

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
COVOX	United States	5	78/863,912	04/18/2006	3,314,528	10/16/2007	INO Therapeutics LLC
IKARIA	Argentina	5	2895583	02/19/2009	2502255	05/04/2012	Ikaria Holdings Inc.
IKARIA	Argentina	10	2895584	02/19/2009	2,339,735	01/06/2010	Ikaria Holdings Inc.
I-N-O	Argentina	5	2012651	03/29/2007	2,201,350	12/12/2007	AGA Aktiebolag
INOBlender	Argentina	10	2662580	04/10/2006	2,163,772	06/12/2007	AGA Aktiebolag
INOFLO	Argentina	5	2223956	06/11/1999	2,052,090	11/07/2005	AGA Aktiebolag
INOMAX	Argentina	5	2209540	06/23/2010	2,446,383	06/13/2011	INO Therapeutics LLC
INOMAX	Argentina	10	2656178	03/13/2006	2,158,935	05/17/2007	AGA Aktiebolag
INOVENT	Argentina	10	2990875	06/11/2012	2,509,164	06/11/2012	INO Therapeutics LLC
INOCAL	Argentina	1	2035103	10/12/2007	2,237,520	07/08/2008	AGA Aktiebolag
IKARIA	Australia	5, 9, 10, 37, 42, 44	1279359	12/24/2008	1279359	11/01/2010	Akaria Inc.
I-N-O	Australia	5	649377	12/22/1994	649377	07/09/1996	INO Therapeutics LLC
INOBlender	Australia	9, 10, 11	1133965	04/04/2006	1133965	01/08/2007	INO Therapeutics LLC
INOCAL	Australia	1	710276	06/07/1996	710276	06/13/1997	INO Therapeutics LLC
INOMAX	Australia	5	788810	03/22/1999	788810	01/14/2000	INO Therapeutics LLC
INOMAX	Australia	10, 11, 42	1134019	04/04/2006	1134019	01/29/2007	INO Therapeutics LLC
INOTHERAPY	Australia	5, 10, 39, 41, 42	796442	06/04/1999	796442	08/25/2000	INO Therapeutics LLC
INOVENT	Australia	5	649372	12/22/1994	649372	08/13/1996	INO Therapeutics LLC

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
INOVENT	Australia	10	710275	06/07/1996	710275	09/11/1998	INO Therapeutics LLC
LUCASSIN	Australia	5	1362084	05/18/2010	13062084	01/10/2011	Ikaria Therapeutics LLC

INOblender	Bolivia	10	06001228	11/27/2006	016623-C	11/27/2006	AGA Aktiebolag
INOMAX	Bolivia	5	086728	08/26/2006	105147-C	08/26/2006	AGA Aktiebolag
INOMAX	Bolivia	10	06000847	10/09/2006	105897-C	10/09/2006	AGA Aktiebolag
INOTHERAPY	Bolivia	5	087904	08/01/2000	80368-C	08/01/2000	AGA Aktiebolag
INOTHERAPY	Bolivia	10	087906	06/06/2000	79014-C	06/06/2000	AGA Aktiebolag
INOTHERAPY	Bolivia	42	087910	06/06/2000	79018-C	06/06/2000	AGA Aktiebolag
INOTHERAPY	Brazil	10	821729209	06/21/1999	821729209	03/11/2003	
INOVENT	Brazil	5, 7, 8, 9, 10, 11, 12, 14, 21, 22, 37, 42	819273082	07/01/1996	819273082	02/23/1999	INO Therapeutics LLC
I-N-O	Brazil	5, 10, 42	818924136	12/11/1995	818924136	06/23/1998	
INOBLENDER	Brazil	10	828287090	04/10/2006	828287090	05/06/2008	AGA Aktiebolag
INOCAL	Brazil	1, 2, 3, 4, 5, 17, 21, 31, 35	819273074	07/01/1996	819273074	02/23/1999	AGA Aktiebolag
INOMAX	Brazil	5, 10, 42	821749005	06/29/1999	821749005	01/27/2009	AGA Aktiebolag
INOMAX	Brazil	10	828279225	03/29/2006	828279225	05/06/2008	AGA Aktiebolag
INOTHERAPY	Brazil	44	821729233	06/21/1999	821729233	03/11/2003	AGA Aktiebolag
IKARIA	Canada	5, 10, 37, 42	1444140	07/08/2009	TMA787272	01/14/2011	Ikaria Holdings Inc.

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
I-N-O	Canada	5	1086868	12/20/2000	TMA576774	03/03/2003	INO Therapeutics LLC
INOblender	Canada	10	1296888	04/06/2006	TMA738591	04/22/2009	INO Therapeutics LLC
INOCAL	Canada	5	1086867	12/20/2000	TMA576744	02/28/2003	INO Therapeutics LLC
INOFLO	Canada	5	1018335	06/09/1999	TMA562856	05/30/2002	INO Therapeutics LLC
INOMAX	Canada	5	1009727	03/24/1999	TMA621362	10/01/2004	INO Therapeutics LLC
INOMAX	Canada	10	1294778	03/22/2006	TMA738008	04/15/2009	INO Therapeutics LLC
INOPULSE	Canada	10	1110179	07/20/2001	TMA602882	02/20/2004	INO Therapeutics LLC
INOTHERAPY	Canada	5, 10, 37, 42	1018336	06/09/1999	TMA643009	06/27/2005	INO Therapeutics LLC
INOVENT	Canada	10	817096	07/05/1996	TMA540612	01/31/2001	INO Therapeutics LLC
ONSTORVIS	Canada	5	1622223	04/11/2013	Not Applicable	Not Applicable	Ikaria Therapeutics LLC
TERLIVAZ	Canada	5	1622222	04/11/2013	Not Applicable	Not Applicable	Ikaria Therapeutics LLC
IKARIA	Chile	5, 10	855480	02/19/2009	875699	02/11/2010	Ikaria Holdings Inc.
INOBLENDER	Chile	10	726119	04/11/2006	764847	08/14/2006	INO Therapetics LLC
INOMAX	Chile	5	954234	05/24/2011	924725	06/27/2011	INO Therapetics LLC
INOMAX	Chile	10	722926	03/14/2006	762143	07/10/2006	INO Therapetics LLC
INOTHERAPY	Chile	5, 10	889954	12/29/2009	874937	11/22/2009	INO Therapetics LLC
INOTHERAPY	Chile	41	889953	12/29/2009	874936	01/24/2010	INO Therapetics LLC

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
INOVENT	Chile	10	754714	12/11/2006	784521	03/24/2007	INO Therapetics LLC

INOBlender	China	9	G892785A	02/08/2006	G892785A	04/04/2006	AGA AB
INOBlender	China	10	G892785A	02/08/2006	G892785A	04/04/2006	AGA AB
INOBlender	China	11	G892785A	02/08/2006	G892785A	04/04/2006	AGA AB
INOMAX	China	10	G893015A	02/08/2006	G893015A	04/04/2006	AGA AB
INOMAX	China	11	G893015A	02/08/2006	G893015A	04/04/2006	AGA AB
INOMAX	China	42	G893015A	02/08/2006	G893015A	04/04/2006	AGA AB
INO	China	5	928660	03/29/1995	928660	01/14/2007	AGA Aktiebolag
INOMAX	China	5	928657	03/29/1995	928657	01/14/2007	AGA Aktiebolag
INOTHERAPY	China	1	1464135	06/11/1999	1464135	10/28/2010	AGA Aktiebolag
INOTHERAPY	China	5	1507781	06/11/1999	1507781	01/14/2011	AGA Aktiebolag
INOTHERAPY	China	39	1467378	06/11/1999	1467378	10/28/2010	AGA Aktiebolag
INOTHERAPY	China	41	1459508	06/11/1999	1459508	10/14/2010	AGA Aktiebolag
INOTHERAPY	China	42	1459493	06/11/1999	1459493	10/14/2010	AGA Aktiebolag
INOVENT	China	10	960071150	06/17/1996	1071982	08/07/1997	AGA Aktiebolag
I-N-O	Colombia	5	02054901	06/25/2002	300687	08/04/2005	AGA Aktiebolag
INOCAL	Colombia	1	03092357	10/16/2003	302673	08/16/2005	AGA Aktiebolag
INOFLO	Colombia	5	99036215	06/09/1999	226864	05/19/2000	AGA Aktiebolag
INOTHERAPY	Colombia	5	99036226	06/09/1999	225942	04/13/2000	AGA Aktiebolag
INOTHERAPY	Colombia	39	99036219	06/09/1999	225945	04/13/2000	AGA Aktiebolag
INOTHERAPY	Colombia	41	99036218	06/09/1999	225946	04/13/2000	AGA Aktiebolag
INOTHERAPY	Colombia	42	99036217	06/09/1999	225943	04/13/2000	AGA Aktiebolag

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
INOVENT	Colombia	10	96043125	08/14/1996	195231	03/19/1997	AGA Aktiebolag
IKARIA	Community Trademarks	5, 10, 42	004664652	10/21/2005	004664652	09/06/2006	Ikaria, Inc.
IIKARIA	Community Trademarks	5, 10, 42	005221593	07/26/2006	005221593	07/05/2007	Ikaria, Inc.
IKARIA	Community Trademarks	9, 37, 44	007590722	02/10/2009	007590722	09/09/2009	Ikaria Holdings Inc.
INO	Ecuador	5	63631	12/12/1995	2520	09/04/1997	INO Therapeutics LLC
INOblender	Ecuador	10	169881	04/17/2006	2181-07	03/21/2007	INO Therapeutics LLC
INOCAL	Ecuador	1	69161	06/05/1996	4941	12/10/1997	INO Therapeutics LLC
INOMAX	Ecuador	10	168716	03/15/2006	1604-07	02/16/2007	INO Therapeutics LLC
INOVENT	Ecuador	10	69160	06/05/1996	4940-97	12/10/1997	INO Therapeutics LLC
INOBLENDER	Hong Kong	10	300619975	04/13/2006	300619975	09/14/2006	INO Therapeutics LLC
INOCAL	Hong Kong	1	199802985	06/11/1996	199802985	03/30/1998	INO Therapeutics LLC
INOMAX	Hong Kong	5	200003618	03/23/1999	200003618	02/29/2000	INO Therapeutics LLC
INOMAX	Hong Kong	10	300601893	03/17/2006	300601893	08/10/2006	INO Therapeutics LLC
INOTHERAPY	Hong Kong	1	200004712	06/05/1999	200004712	03/23/2000	INO Therapeutics LLC
INOTHERAPY	Hong Kong	5	200004713	06/05/1999	200004713	03/23/2000	INO Therapeutics LLC
INOTHERAPY	Hong Kong	7	200101483	06/05/1999	200101483	02/09/2001	INO Therapeutics LLC

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
INOTHERAPY	Hong Kong	10	200004714	06/05/1999	200004714	03/23/2000	INO Therapeutics

							LLC
INOTHERAPY	Hong Kong	11	200101402	06/05/1999	200101402	02/08/2001	INO Therapeutics LLC
INOTHERAPY	Hong Kong	39	200105749	06/05/1999	200105749	05/17/2001	INO Therapeutics LLC
INOTHERAPY	Hong Kong	41	200106471	06/05/1999	200106471	06/01/2001	INO Therapeutics LLC
INOTHERAPY	Hong Kong	42	200106472	06/05/1999	200106472	06/01/2001	INO Therapeutics LLC
INOVENT	Hong Kong	5	199702525	03/11/1995	199702525	03/07/1997	The BOC Group Inc.
INOVENT	India	10	728393	06/24/1996	298112	06/24/1996	Ohmeda Inc.
I-N-O	India	5	658085	03/09/1995	714394	03/28/2008	AGA Aktiebolag
INOBLENDER	India	10	1447168	04/13/2006	748025	08/25/2008	AGA Aktiebolag
INOCAL	India	1	728392	06/24/1996	267071	07/15/2003	The BOC Group, Inc.
INOMAX	India	5	658089	03/09/1995	285498	01/06/2004	Ohmeda Inc.
INOMAX	India	10	1433164	03/15/2006	748006	08/25/2008	AGA Aktiebolag
INOTHERAPY	India	7	860253	06/09/1999	265158	06/13/2003	AGA Aktiebolag
IKARIA	Indonesia	5, 10	D002009006782	03/03/2009	IDM000271544	09/21/2010	Ikaria Holdings Inc.
INOREG	Indonesia	11	R002008007428	07/15/2009	IDM000213550	08/13/2009	
INOREG	Indonesia	11	R002008008669	10/14/2009	IDM000234196	01/25/2010	
INOTHERAPY	Indonesia	1	R002008007436	07/15/2009	IDM000213557	08/13/2009	
INOTHERAPY	Indonesia	5	R002008007429	07/15/2009	IDM000213551	08/13/2009	
INOTHERAPY	Indonesia	10	R002008007432	07/15/2009	IDM000213553	08/13/2009	
INOTHERAPY	Indonesia	11	R002008007431	07/15/2009	IDM000213552	08/13/2009	

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
INOTHERAPY	Indonesia	11	R002008008672	10/14/2009	IDM000234197	01/25/2010	
INOTHERAPY	Indonesia	39	V002008007435	07/15/2009	IDM000213556	08/13/2009	
INOTHERAPY	Indonesia	41	V002008007433	07/15/2009	IDM000213554	08/13/2009	
INOTHERAPY	Indonesia	44	V002008007434	07/15/2009	IDM000213555	08/13/2009	
I-N-O	Indonesia	5	R00200500549	03/22/20005	IDM000238514	03/02/2010	
INOMAX	Indonesia	5	R00200500550	03/22/2005	IDM000238515	03/02/2010	
IKARIA	Japan	5, 9, 10, 37, 42, 44	2009-052455	07/10/2009	5312137	03/26/2010	Ikaria Holdings Inc.
□□□□□□ (INOCAL in Katakana)	Japan	1	2003-037253	05/08/2003	4768539	04/30/2004	INO Therapeutics LLC
INOFLO	Japan	1, 5	H11-051919	06/11/1999	4455017	02/23/2001	INO Therapeutics LLC
INOMAX	Japan	5	H11-025043	03/19/1999	4359499	02/04/2000	INO Therapeutics LLC
□□□□□□□ (INOMAX in Katakana)	Japan	5	2003-037251	05/08/2003	4783465	07/02/2004	INO Therapeutics LLC
INOMETER	Japan	9, 10, 42, 44	2002-083154	10/01/2002	4710141	09/12/2003	INO Therapeutics LLC
INOTHERAPY	Japan	5, 10, 41, 42	H11-058119	06/28/1999	4478969	06/01/2001	INO Therapeutics LLC
□□□□□□□ (INOTHERAPY in Katakana)	Japan	5, 10, 44	2003-037252	05/08/2003	4765385	04/16/2004	INO Therapeutics LLC
□□□□□ (INOVENT in Katakana)	Japan	10	2001-045864	05/22/2001	4628986	12/13/2002	INO Therapeutics LLC
INOPULSE	Japan	10	2012-52253	06/28/2012	Not Applicable	Not Applicable	INO Therapeutics LLC
I-N-O	Malaysia	5	95003799	04/24/1995	95003799	04/24/1995	AGA Aktiebolag

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner

INOblender	Malaysia	10	06006346	04/19/2006	06006346	04/19/2006	INO Therapeutics LLC
INOCAL	Malaysia	1	97010861	08/06/1997	97010861	08/06/1997	AGA Aktiebolag
INOMAX	Malaysia	5	95003803	04/24/1995	95003803	04/24/1995	AGA Aktiebolag
INOMAX	Malaysia	10	06004524	03/22/2006	06004524	03/22/2006	AGA Aktiebolag
INOREG	Malaysia	7	99005103	06/14/1999	99005103	06/14/1999	AGA Aktiebolag
INOREG	Malaysia	11	99005106	06/14/1999	99005106	06/14/1999	AGA Aktiebolag
INOTHERAPY	Malaysia	1	99005108	06/14/1999	99005108	06/14/1999	AGA Aktiebolag
INOTHERAPY	Malaysia	5	99005109	06/14/1999	99005109	06/14/1999	AGA Aktiebolag
INOTHERAPY	Malaysia	7	99005248	06/16/1999	99005248	06/16/1999	AGA Aktiebolag
INOTHERAPY	Malaysia	10	99005105	06/14/1999	99005105	06/14/1999	AGA Aktiebolag
INOTHERAPY	Malaysia	11	99005113	06/14/1999	99005113	06/14/1999	AGA Aktiebolag
INOTHERAPY	Malaysia	39	99005112	06/14/1999	99005112	06/14/1999	AGA Aktiebolag
INOTHERAPY	Malaysia	41	99005111	06/14/1999	99005111	06/14/1999	AGA Aktiebolag
INOTHERAPY	Malaysia	44	99005110	06/14/1999	99005110	06/14/1999	AGA Aktiebolag
INOVENT	Malaysia	10	97010799	08/05/1997	97010799	08/05/1997	Datex-Ohmeda Inc.
INOFLO	Malaysia	1	99005107	06/14/1999	99005107	06/14/1999	AGA Aktiebolag
INOBLENDER	Mexico	10	777519	04/17/2006	932936	04/28/2006	AGA Aktiebolag
INOCAL	Mexico	1	795525	07/21/2006	980431	04/19/2007	AGA Aktiebolag
INOMAX	Mexico	5	368830	03/24/1999	660113	06/26/2000	AGA Aktiebolag
INOMAX	Mexico	10	771663	03/14/2006	929989	04/21/2006	AGA Aktiebolag
INOMETER	Mexico	9	636290	01/07/2004	824044	02/27/2004	AGA Aktiebolag
INOMETER	Mexico	10	636289	01/07/2004	824496	03/09/2004	AGA Aktiebolag
INOTHERAPY	Mexico	1	380022	06/22/1999	622110	08/31/1999	AGA Aktiebolag

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
INOTHERAPY	Mexico	5	380026	06/22/1999	620493	08/30/1999	AGA Aktiebolag
INOTHERAPY	Mexico	10	380021	06/22/1999	622109	08/31/1999	AGA Aktiebolag
INOTHERAPY	Mexico	11	380017	06/22/1999	622106	08/31/1999	AGA Aktiebolag
INOTHERAPY	Mexico	11	380020	06/22/1999	624904	09/27/1999	AGA Aktiebolag
INOTHERAPY	Mexico	39	380025	06/22/1999	647546	03/28/2000	AGA Aktiebolag
INOTHERAPY	Mexico	41	380016	06/22/1999	656311	05/30/2000	AGA Aktiebolag
INOTHERAPY	Mexico	42	380018	06/22/1999	622107	08/31/1999	AGA Aktiebolag
INOVENT	Mexico	10	795890	07/24/2006	969641	01/25/2007	Datex-Ohmeda Inc.
IKARIA	New Zealand	5, 10, 37, 42, 44	802093	02/04/1999	802093	08/12/2010	Ikaria Holdings Inc.
I-N-O	New Zealand	5	244337	12/22/1994	244337	01/24/1997	INO Therapeutics LLC
INOblender	New Zealand	10	746071	04/07/2006	746071	07/12/2007	INO Therapeutics LLC
INOCAL	New Zealand	1	263005	06/04/1996	263005	07/21/1997	INO Therapeutics LLC
INOMAX	New Zealand	10	744310	03/10/2006	744310	09/14/2006	INO Therapeutics LLC
INOREG	New Zealand	7	310701	06/04/1999	310701	12/07/2000	INO Therapeutics LLC
INOREG	New Zealand	11	310700	06/04/1999	310700	12/07/2000	INO Therapeutics LLC
INOTHERAPY	New Zealand	1	310704	06/04/1999	310704	12/07/2000	INO Therapeutics LLC
INOTHERAPY	New Zealand	5	310705	06/04/1999	310705	12/07/2000	INO Therapeutics LLC
INOTHERAPY	New Zealand	7	310706	06/04/1999	310706	12/07/2000	INO Therapeutics LLC
INOTHERAPY	New Zealand	10	310707	06/04/1999	310707	12/07/2000	INO Therapeutics LLC

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
INOTHERAPY	New Zealand	11	310708	06/04/1999	310708	12/07/2000	INO

							Therapeutics LLC
INOTHERAPY	New Zealand	39	310709	06/04/1999	310709	12/07/2000	INO Therapeutics LLC
INOTHERAPY	New Zealand	41	310710	06/04/1999	310710	12/07/2000	INO Therapeutics LLC
INOTHERAPY	New Zealand	42	310711	06/04/1999	310711	12/07/2000	INO Therapeutics LLC
INOVENT	New Zealand	10	263006	06/04/1996	263006	09/11/1997	INO Therapeutics LLC
INOblender	Pakistan	10	220555	04/07/2006	220555	11/01/2010	AGA Aktiebolag
INOCAL	Pakistan	1	167271	10/24/2000	167271	10/24/2007	AGA Aktiebolag
INOFLO	Pakistan	1	155649	06/10/1999	155649	Unknown	AGA Aktiebolag
INOMAX	Pakistan	10	219440	03/10/2006	219440	05/25/2011	AGA Aktiebolag
INOTHERAPY	Pakistan	1	155648	06/10/1999	155648	10/11/2005	AGA Aktiebolag
INOTHERAPY	Pakistan	11	155656	06/10/1999	155656	12/08/2005	AGA Aktiebolag
INOMAX	Pakistan	5	153912	Unknown	153912	Unknown	AGA Aktiebolag
I-N-O	Pakistan	5	167274	10/24/2000	Not Applicable	Not Applicable	AGA Aktiebolag
INOFLO	Pakistan	5	155650	06/10/1999	Not Applicable	Not Applicable	AGA Aktiebolag
INOTHERAPY	Pakistan	16	155909	06/25/1999	Not Applicable	Not Applicable	AGA Aktiebolag
I-N-O	Philippines	5	41995099402	03/31/1995	41995099402	07/30/2005	AGA Aktiebolag
INOblender	Philippines	10	42006003882	04/07/2006	42006003882	03/05/2007	AGA Aktiebolag
INOCAL	Philippines	1	42004007800	08/24/2004	42004007800	12/24/2007	AGA Aktiebolag
INOFLO	Philippines	1, 5	42002007546	09/05/2002	42002007546	05/09/2005	AGA Aktiebolag
INOMAX	Philippines	5	42006003069	03/17/2006	42006003069	11/13/2006	AGA Aktiebolag

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
INOREG	Philippines	7, 11	42002007548	09/05/2002	42002007548	03/20/2005	AGA Aktiebolag
INOTHERAPY	Philippines	1, 5, 7, 10, 11, 39, 41, 42	42002007547	09/05/2002	42002007547	03/20/2005	AGA Aktiebolag
INOMAX	Philippines	5	42009500140	3/26/2009	42009500140	8/13/2009	INO Therapeutics LLC
INOMAX	Philippines	10	42006002837	3/13/2006	42006002837	4/30/2007	AGA Aktiebolag
IKARIA	Singapore	5, 9, 10, 37, 42, 44	T09014281	02/12/2009	T09014281	05/21/2009	Ikaria Holdings Inc.
INOCAL	Singapore	1	T9605650C	06/05/1996	T9605650C	11/29/1999	INO Therapeutics LLC
INOMAX	Singapore	5	T9902879I	03/23/1999	T9902879I	09/14/2000	INO Therapeutics LLC
INOTHERAPY	Singapore	5	T9906031E	06/14/1999	T9906031E	09/22/2000	INO Therapeutics LLC
INOTHERAPY	Singapore	10	T9906033A	06/14/1999	T9906033A	11/01/2000	INO Therapeutics LLC
INOVENT	Singapore	10	T9605651A	06/05/1996	T9605651A	06/01/1999	INO Therapeutics LLC
INOTHERAPY	Singapore	11	T0525595H	12/20/2005	T0525595H	01/23/2007	AGA Aktiebolag
INOVENT	South Africa	10	1996/07268	06/03/1996	1996/07268	09/01/1999	
INOBLENDER	South Africa	10	2006/07894	04/07/2006	Not Applicable	Not Applicable	
IKARIA	Taiwan	5, 10	098004085	02/06/2009	01411355	05/16/2010	Akaria Holdings Inc.
INOblender	Taiwan	10	095017730	04/10/2006	01248877	02/01/2007	INO Therapeutics LLC
INOCAL	Taiwan	1	85026895	06/03/1996	00766655	07/16/1997	INO Therapeutics LLC
INOMAX	Taiwan	5	84009974	03/07/1995	00713867	04/16/1996	INO Therapeutics LLC

Mark	Country	Class(es)	Application	Application	Registration	Registered	Registered

			No.	Date	No.		Owner
INOMAX	Taiwan	10	95011908	03/13/1996	01234266	11/01/1996	INO Therapeutics LLC
INOTHERAPY	Taiwan	1	88029585	06/16/1999	00910823	11/01/2000	INO Therapeutics LLC
INOTHERAPY	Taiwan	5	88029584	06/16/1999	00908744	10/16/2000	INO Therapeutics LLC
INOTHERAPY	Taiwan	10	88029582	06/16/1999	00890901	05/01/2000	INO Therapeutics LLC
INOTHERAPY	Taiwan	11	88029581	06/16/1999	00907828	10/01/2000	INO Therapeutics LLC
INOTHERAPY	Taiwan	41	88029579	06/16/1999	00131154	10/16/2000	INO Therapeutics LLC
INOTHERAPY	Taiwan	42	88029578	06/16/1999	00137557	02/01/2001	INO Therapeutics LLC
INOVENT	Taiwan	5	84009979	03/07/1995	713868	04/16/1996	INO Therapeutics LLC
INOTHERAPY	Thailand	5	392056	07/08/1999	TM121076	07/08/1999	AGA AB
INOTHERAPY	Thailand	10	392058	07/08/1999	TM124636	07/08/1999	AGA AB
INOVENT	Thailand	10	312549	07/11/1996	TM60264	08/15/1996	AGA AB
INOblender	Thailand	10	623525	04/12/2006	TM251215	04/12/2006	AGA AB
INOMAX	Thailand	10	621585	03/24/2006	TM254899	03/24/2006	AGA AB
IKARIA	Uruguay	5, 10	399805	02/17/2009	399805	01/17/2012	Akaria Holdings Inc.
I-N-O	Uruguay	5	381551	06/08/2007	381551	08/04/1997	INO Therapeutics LLC
INOblender	Uruguay	10	370252	04/20/2006	370252	05/16/2007	INO Therapeutics LLC
INOCAL	Uruguay	1	392509	05/30/2008	392509	06/05/1998	INO Therapeutics LLC
INOMAX	Uruguay	5	429525	11/18/2011	Not Applicable	11/26/2001	AGA Aktiebolag

Mark	Country	Class(es)	Application No.	Application Date	Registration No.	Registered	Registered Owner
INOMAX	Uruguay	10	369364	03/10/2006	369364	03/28/2007	AGA Aktiebolag
INOTHERAPY	Uruguay	5, 10, 11, 39, 41, 44	419863	01/27/2011	419863	08/01/2000	INO Therapeutics LLC
INOVENT	Uruguay	10	287645	06/13/1996	287645	08/04/1997	INO Therapeutics LLC
INOVENT	Venezuela	10	2011-001901	02/07/2011	Not Applicable	Not Applicable	INO Therapeutics LLC
I-N-O	Venezuela	5	1995-020086	12/12/1995	P197278	05/09/1997	AGA Aktiebolag
INOBlender	Venezuela	10	2006-007649	04/07/2006	P282088	10/24/2007	AGA Aktiebolag
INOCAL	Venezuela	1	1996-008395	06/06/1996	P202514	01/23/1998	The BOC Group, Inc.
INOMAX	Venezuela	10	2006-005678	06/17/2006	P275561	11/30/2006	AGA Aktiebolag
INOTHERAPY	Venezuela	10	1999-010553	06/17/1999	P222135	07/07/2000	AGA Aktiebolag
INOTHERAPY	Venezuela	39	1999-010546	06/17/1999	S013332	07/07/2000	AGA Aktiebolag
INOTHERAPY	Venezuela	41	1999-010545	06/17/1999	S013331	07/07/2000	AGA Aktiebolag

Schedule 1.1(b)

R&DCo Intellectual Property

IKARIA REFERENCE NUMBER	TITLE	COUNTRY	STATUS	FILING DATE	APPLICATION NUMBER
[**]	[**]	[**]	[**]	[**]	[**]
[**]	[**]	[**]	[**]	[**]	[**]
[**]	[**]	[**]	[**]	[**]	[**]

[**] [**] [**] [**] [**] [**]

Schedule 6.1(c)

Indemnification Matters

The R&DCo Option Payments

Schedule 8.13(a)

R&DCo Assets Used in or Necessary for Ikaria Business

None.

Schedule 8.13(b)

Required Consents

None.

Exhibit A

Form of Device Clinical Supply Agreement

Incorporated by reference to Exhibit 10.11 to the Company's Registration Statement on Form S-1

Exhibit B

Form of Drug Clinical Supply Agreement

Incorporated by reference to Exhibit 10.12 to the Company's Registration Statement on Form S-1

Exhibit C

Form of Employee Matters Agreement

Incorporated by reference to Exhibit 10.13 to the Company's Registration Statement on Form S-1

Exhibit D

Form of R&D Cross License Agreement

Incorporated by reference to Exhibit 10.14 to the Company's Registration Statement on Form S-1

Exhibit E

Form of Transition Services Agreement

Incorporated by reference to Exhibit 10.15 to the Company's Registration Statement on Form S-1

Exhibit 10.11

Execution Version

Confidential Materials omitted and filed separately with the
Securities and Exchange Commission. Double asterisks denote omissions.

DEVICE CLINICAL SUPPLY AGREEMENT

This Device Clinical Supply Agreement (this "Agreement") is entered into as of February 9, 2014 (the "Effective Date") by and between by and between INO Therapeutics LLC, a Delaware limited liability company, with offices at Perryville III Corporate Park, 53 Frontage Road, Third Floor, Hampton, NJ 08827 d/b/a Ikaria ("Ikaria"), and Bellerophon Pulse Technologies LLC, a Delaware limited liability company, with offices at Perryville III Corporate Park, 53 Frontage Road, Third Floor, Hampton, NJ 08827 d/b/a Ikaria ("Pulse Technologies"). Ikaria and Pulse Technologies may be individually referred to as a "Party" and together as the "Parties."

WHEREAS, the Parties were formerly owned by a common parent company, Ikaria, Inc. ("Ikaria Parent Company");

WHEREAS, Pulse Technologies has, as part of certain spin-out transactions (the "Spin-Out"), ceased to be a direct or indirect subsidiary of Ikaria Parent Company, and is now therefore not owned by, or affiliated with, either Ikaria Parent Company or Ikaria;

WHEREAS, Pulse Technologies is engaged in the business of developing, manufacturing, and commercializing products for (a) pulmonary hypertension secondary to chronic obstructive pulmonary disease ("COPD") and (b) primary or idiopathic pulmonary arterial hypertension ("PAH") (collectively, the "Pulse Technologies Clinical Programs");

WHEREAS, prior to the Spin-Out, Ikaria manufactured the nitric oxide delivery devices listed in Exhibit A to this Agreement (the "Devices") used by Pulse Technologies as part of the Pulse Technologies Clinical Programs; and

WHEREAS, Pulse Technologies wishes Ikaria to continue on a short term basis to manufacture, and Ikaria wishes to continue to manufacture, the Devices for Pulse Technologies, all subject to and in accordance with the terms and conditions of this Agreement.

NOW THEREFORE, in consideration of the foregoing premises, which are incorporated into and made a part of this Agreement, and of the mutual covenants which are recited herein, the Parties agree as follows:

1. Definitions.

1.1 "Affiliate" means, with respect to a Party, any Person directly or indirectly controlling, controlled by or under common control with, such Party. For purposes of this definition only, "control" of a Person shall mean the ability, directly or indirectly, to direct the activities of the relevant Person, and with respect to corporate entities shall mean (a) ownership or direct control of fifty percent (50%) or more of the outstanding voting stock or other ownership interest of such Person, or (b) direct or indirect possession, of the power to elect or appoint fifty percent (50%) or more of the members of the governing body of such Person. Notwithstanding the foregoing or any direct or indirect control relationship that exists between them, Ikaria and Pulse Technologies shall be deemed not to be Affiliates of one another.

1.2 "COGS" means, as to Ikaria and its Affiliates, with respect to the Devices, the aggregate of internal and external costs of Ikaria and its Affiliates to manufacture such Devices, calculated as follows: (a) to the extent that Ikaria or its Affiliates performs all or any part of the manufacturing of such Devices, the actual direct material costs and direct labor costs for, plus manufacturing overhead reasonably allocable to, such manufacturing of such Devices (which may include the costs of audits, all directly incurred manufacturing variances, manufacturing administrative and facilities costs (including

depreciation)), all calculated in accordance with GAAP; and (b) to the extent that manufacturing of such Devices is performed by a Third Party, the costs paid to such Third Party for such activities and the reasonably allocated direct labor costs incurred by Ikaria or any of its Affiliates in managing and overseeing the Third Party relationship, determined in accordance with GAAP.

1.3 "Confidential Information" means information disclosed by a Party or its Affiliate (such Party referred to as the "Disclosing Party") to the other Party or its Affiliate (such Party referred to as the "Receiving Party"), which information relates either directly or indirectly to the business of the Disclosing Party, including information and data regarding the manufacture or use, pre-clinical or clinical data regarding, the status of research or development of any Device. Confidential Information of the Disclosing Party excludes any information that the Receiving Party can establish by written records: (a) was known by the Receiving Party prior to receipt from the Disclosing Party; (b) was disclosed to the Receiving Party by a Third Party having the right to do so; (c) was, or subsequently became, publicly known through no fault of the Receiving Party or its Affiliates; or (d) was concurrently or subsequently developed by personnel of the Receiving Party without having had access to the Disclosing Party's Confidential Information.

1.4 "COPD" shall have the meaning set forth in the recitals to this Agreement.

1.5 "Cross License" shall mean the Exclusive Cross-License, Technology Transfer, and Regulatory Matters Agreement by and between the Parties dated of even date herewith.

1.6 "Devices" shall have the meaning set forth in the recitals to this Agreement.

1.7 "Device IP" shall have the meaning set forth in Section 2.9.

1.8 "Effective Date" shall have meaning set forth in the preamble to this Agreement.

1.9 "Facility" means Ikaria's manufacturing facilities located in Madison, WI, or such other manufacturing site(s) specified by Ikaria from time to time.

1.10 "FDA" means the United States Food and Drug Administration or any successor organization.

1.11 "Federal Health Care Programs" shall have the meaning set forth in Section 5.4.

1.12 "Forecast" shall have the meaning set forth in Section 2.5.

1.13 "Ikaria Parent Company" shall have the meaning set forth in the recitals to this Agreement.

1.14 "Intellectual Property" means, collectively, patents, trademarks, copyrights (including to any software, whether in object code or source code form), trade secrets, know-how, and any other intellectual or proprietary property or rights.

1.15 "PAH" shall have the meaning set forth in the recitals to this Agreement.

1.16 "Person" means any individual, governmental authority, partnership, corporation, limited liability company, unincorporated organization or association, any trust or any other business entity.

1.17 "Pulse Technologies Clinical Programs" shall have the meaning set forth in the recitals to

this Agreement.

1.18 "Purchase Order" means a written purchase order issues by Pulse Technologies to Ikaria for the purchase of Devices.

1.19 "Quality Agreement" has the meaning set forth in Section 3.

1.20 "Regulatory Authority" means any competent governmental authority which regulates the manufacture, development or sale of any Device.

1.21 "Specification" means, with respect to each Device, the specifications in effect at Ikaria for that Device immediately prior to the Spin-Out.

1.22 "Term" has the meaning set forth in Section 8.1.

1.23 "Third Party" means any Person who is not a Party or an Affiliate of a Party.

2. Supply of Devices.

2.1 Obligations of Ikaria. During the Term of this Agreement, Ikaria will use commercially reasonable efforts to manufacture and supply Pulse Technologies' requirements for Devices for the Pulse Technologies Clinical Programs in accordance with the terms of this Agreement and the Quality Agreement. Pulse Technologies acknowledges and agrees that nothing in this Agreement shall require Ikaria to hire, obtain, or retain additional resources of any type (whether personnel, infrastructure, or otherwise), or to make capital expenditures of any kind, in order to manufacture and supply the Devices, nor shall anything in this Agreement require Ikaria to prioritize manufacturing and supplying the Devices to Pulse Technologies over performing similar services for its own benefit.

2.2 Obligations of Pulse Technologies. Pulse Technologies will provide Ikaria with such information and cooperation as may be necessary for the manufacture and supply of the Devices in accordance with this Agreement and the Quality Agreement.

2.3 Changes. Ikaria shall use commercial reasonable efforts to accommodate any reasonable changes to the Specifications for the Devices, it being understood and agreed that Pulse Technologies shall bear any and all costs associated with such changes. Pulse Technologies further acknowledges and agrees that no such requested change may be inconsistent with, or violative of, the terms and conditions of the Cross License, including the restricted abilities, attributes, capabilities, capacities, functions, and specifications set forth in Exhibit A to the Cross License.

2.4 Pricing. Pricing for the Devices shall be as set forth in Exhibit B to this Agreement, and shall be subject to the other terms and conditions set forth therein.

2.5 Forecasts. Within 10 business days after the Effective Date, Pulse Technologies shall provide to Ikaria a written forecast of all Devices which Pulse Technologies expects to order from Ikaria during the Term (the "Forecast"). Pulse Technologies shall update the Forecast in writing on a monthly basis. The Forecast shall constitute a non-binding, good faith estimate provided by Pulse Technologies solely to assist Ikaria in production planning, and shall not represent any purchase commitment by Pulse Technologies or a supply commitment by Ikaria.

2.6 Purchase Orders.

(a) During the Term, Pulse Technologies may place Purchase Orders with Ikaria. Acknowledging that the lead time for the Devices is estimated to be between four and six months, Pulse Technologies agrees to submit all Purchase Orders no later than six months after the Effective Date. Each Purchase Order shall specify the specific Devices ordered. Ikaria shall be deemed to have accepted a Purchase Order unless it objects within 30 days after receiving such Purchase Order. If Ikaria's believes it will be unable to manufacture the requested Devices (e.g., because of an inability to obtain required component parts), Ikaria shall promptly inform Pulse Technologies thereof. If a Purchase Order is accepted by, or deemed accepted by Ikaria under this Section 2.6(a), then Ikaria shall use commercially reasonable efforts to produce the quantity of Devices set forth in the Purchase Order.

(b) Each Purchase Order and any acknowledgment thereof shall be governed by the terms of this Agreement. If a Party uses forms or documents to place or accept Purchase Order that contain terms and conditions that are in addition to or contrary to those in this Agreement, the Parties agree and acknowledge that such forms or documents will be used for convenience only, and that no terms or conditions set forth therein, except with respect to quantity, shall be of any force or effect.

2.7 Delivery. Ikaria shall deliver the Devices to a carrier selected by Pulse Technologies. The Devices shall be made available EXW the Facility (Incoterms 2010). Title and risk of loss will pass to Pulse Technologies when the Devices are made available to the carrier selected by Pulse Technologies. Pulse Technologies is responsible for payment of all shipment costs, including any insurance necessary to guard against loss or damage during shipment.

2.8 Inspection and Acceptance of Devices.

(a) Pulse Technologies shall have 30 days from the date of receipt of each Device to inspect and reject acceptance by written notice to Ikaria; provided, however, that any such notice shall set forth Pulse Technologies' reasons for rejection in reasonable detail and provided, further, that Pulse Technologies may reject a Device only if Pulse Technologies believes that the Device in question does not conform in all material respects with the applicable Specifications. If Ikaria does not receive Pulse Technologies' written notice of rejection within such 30 day period, Pulse Technologies shall be deemed to have accepted such Device.

(b) If Pulse Technologies provides Ikaria with a timely notice of rejection under Section 2.8(a), Pulse Technologies shall return the rejected Device(s) to Ikaria at Ikaria's expense. Ikaria shall have 30 days following receipt of the rejected Devices in which to test such Devices. If Ikaria does not dispute a rejection, Ikaria shall rework or replace the rejected Device(s), at Ikaria's expense and such rework or replacement shall constitute Pulse Technologies' exclusive remedy and Ikaria's sole liability with respect to such rejection. If Ikaria disputes a rejection, Ikaria shall provide Pulse Technologies with written notice of such dispute within 30 days after receiving the returned Devices, and the Parties shall use commercially reasonable efforts to resolve the dispute amicably and promptly. If the Parties are unable to reach a resolution within 30 days after Pulse Technologies' notice of rejection, the returned Devices shall be submitted to an independent consultant mutually acceptable to the Parties, whose decision as to the conformity of such Devices with the applicable Specification shall be final and binding. The Party against whom the dispute is decided shall pay any charges for such consultant. If the consultant determines that the returned Devices did not conform to the Specification, Ikaria shall rework or replace the rejected Devices at no charge to Pulse Technologies, and such replacement shall constitute Pulse Technologies' exclusive remedy and Ikaria's sole liability with respect to such rejected Devices.

2.9 No Licenses. Pulse Technologies acknowledges and agrees that nothing in this Agreement grants, or shall be deemed or interpreted to grant, to Pulse Technologies any right, title, or interest in or to any Intellectual Property reflected, contained, or incorporated in, or practice under, as part

of or in the process of manufacturing and delivery of Devices (collectively, the "Device IP"). Pulse Technologies acknowledges and agrees that it is obtaining ownership to the physical Devices only under this Agreement, and not to or in any of the Device IP.

3. Quality Agreement. The Parties shall use reasonable efforts to negotiate and conclude a quality agreement ("Quality Agreement") within 60 days after the Effective Date. The Quality Agreement shall detail the division of responsibilities between the Parties regarding quality and regulatory controls and reporting concerning the Devices. In the case of a conflict between the Quality Agreement and this Agreement, the terms of this Agreement shall control unless such term in the Quality Agreement expressly references such conflict and the Parties intend to have the Quality Agreement control such provision.

4. Audit Rights. Pulse Technologies shall have the right to conduct reasonable audits and inspections of the Facility, Ikaria's manufacturing operations, and Ikaria's records relating to the manufacture of Devices under this Agreement. Ikaria shall reasonably cooperate with Pulse Technologies in conducting such audits and inspections.

5. Warranties.

5.1 General Warranties. Each Party warrants to the other Party that (a) it has the right and authority to enter into this Agreement and to carry out its obligations hereunder; (b) it is validly existing in the jurisdiction in which it is incorporated and is authorized to do business under the laws of each jurisdiction in which it engages in business activities; and (c) it is not aware of any legal, contractual or other restriction, limitation or condition that might adversely affect its ability to perform its obligations hereunder.

5.2 Warranties by Ikaria. Ikaria warrants to Pulse Technologies that each Device delivered hereunder shall conform in all material respects with its applicable Specifications. Pulse Technologies agrees that its exclusive remedies, and Ikaria's sole liabilities, with respect to any breach of the warranty set forth in this Section 5.2 are set forth in Section 2.8 of this Agreement.

5.3 Pass Through Warranties. Ikaria shall use commercially reasonable efforts to pass-through to Pulse Technologies the benefit of any warranties on component parts incorporated into the Devices to the extent Ikaria has the right to do so. Pulse Technologies acknowledges and agrees that this Section 5.3 does not provide Pulse Technologies with any additional rights or remedies vis-à-vis Ikaria beyond those stated in Section 5.

5.4 Debarment. Each party represents and warrants that, as of the Effective Date and throughout the Term, it (and each of its employees and agents) (a) is not currently excluded, debarred or otherwise ineligible to participate in the Federal health care programs as defined in 42 U.S.C. 1320a7b(f) (the "Federal Health Care Programs"); (b) has not been convicted of a criminal offense related to the provision of healthcare items or services but yet to be excluded, debarred or otherwise declared ineligible to participate in the Federal Health Care Programs; and (c) is not under investigation or otherwise aware of any circumstances which may result in it (or its agents, employees or any substitutes thereof performing any duties under this Agreement) being excluded from participation in the Federal Health Care Programs.

5.5 DISCLAIMER. EXCEPT AS EXPRESSLY PROVIDED HEREIN, NEITHER PARTY MAKES NOR RECEIVES ANY REPRESENTATION OR WARRANTY OF ANY KIND, EXPRESS, IMPLIED, STATUTORY OR OTHERWISE, OR ARISING FROM A COURSE OF DEALING OR USAGE OF TRADE PRACTICE, WITH REGARD TO THE DEVICES OR OTHERWISE UNDER

THIS AGREEMENT, INCLUDING THE IMPLIED WARRANTIES OF MERCHANTABILITY, NON-INFRINGEMENT, OR FITNESS FOR A PARTICULAR PURPOSE.

6. Indemnity. Pulse Technologies shall indemnify, defend and hold Ikaria, its Affiliates and their respective directors, officers, employees, agents, successors and assigns harmless from and against any damages, losses, judgments, claims, suits, actions, liabilities, costs and expenses (including, but not limited

to, reasonable attorneys' fees), as and when incurred, resulting from any Third Party claims or suits arising out of the ownership, use, handling, development, distribution, marketing, or sale of any Device.

7. Compliance.

7.1 Compliance with Laws. Each Party shall comply with all applicable laws and regulations governing the performance of such Party's obligations under this Agreement. Without limiting the foregoing, each Party shall comply with applicable US and other laws, rules and regulations that govern the import, export and re-export of the Devices, including the U.S. Export Administration Regulations, and will obtain any required export and import authorizations to perform its obligation hereunder.

7.2 Regulatory Filings. Pulse Technologies, at its expense, shall be solely responsible for the preparation, filing, and maintenance of all regulatory documents and all governmental permits, licenses and other approvals as may be necessary with respect to the formulation, marketing, distribution, sale, and use of each Device.

7.3 Permits. Ikaria at its expense shall be solely responsible for, and has the obligation to prepare, file, and maintain during the Term, all licenses, permits, and approvals as may be necessary with respect to the manufacture of Devices at the Facility.

8. Term and Termination.

8.1 Term. Unless otherwise terminated under this Section 8, this Agreement will commence as of the Effective Date and will continue for a period of twelve months (the "Term"); provided, however, that if a Purchase Order that has been accepted by Ikaria has not been fulfilled at the expiration of the Term, this Agreement shall continue to remain in effect for so long as necessary to complete the delivery of Devices under that Purchase Order.

8.2 Termination. This Agreement may be terminated by either Party upon 60 days written notice of the other Party's material breach of any provision of this Agreement; provided, however, that the breaching Party will have an opportunity to (a) cure the breach during the 60 day notice period, or (b) provide the non-breaching Party with a plan to remedy the breach within the 60 day notice period, and if so cured, no termination will be deemed to have occurred as long as the breaching Party diligently pursues the plan to remedy the breach and completes such plan in accordance with the time frame agreed to by the Parties (such time frame not to exceed an additional 60 days).

8.3 Effect of Termination. Termination or expiration of this Agreement shall not release either Party from any liability, right of action or other obligation which has arisen prior to such termination or expiration, including Ikaria's obligation to deliver to Pulse Technologies such quantity of Devices under any accepted Purchase Order to the effective date of termination or expiration, and Pulse Technologies' obligation to pay Ikaria the amount set forth in such Purchase Order. Notwithstanding any expiration or termination of this Agreement, the following provisions shall survive: Sections 2.6(b), 2.8(b), 2.9, 5.5, 6, 8.3, 9, 10, and 11, as well as Pulse Technologies' payment obligations under Exhibit B.

9. Confidentiality.

9.1 Non-Use and Non-Disclosure of Confidential Information. Each Receiving Party agrees that all Confidential Information of the Disclosing Party (a) shall not be used by the Receiving Party except to perform its obligations or exercise its rights under this Agreement, (b) shall be maintained in confidence by the Receiving Party, and (c) except as permitted by Section 9.2, shall not be disclosed by the Receiving Party to any Person without the prior written consent of the Disclosing Party.

9.2 Permitted Disclosures.

(a) The Receiving Party may provide the Disclosing Party's Confidential Information (i) to its Affiliates and to their employees, consultants, advisors, and contractors who have a need to know such Confidential Information for purposes of the Receiving Party exercising or granting licenses or sublicenses, (ii) in communications with existing or bona fide prospective acquirers, merger partners, lenders or investors, in each case of (i) and (ii), on a need to know basis and under appropriate confidentiality provisions substantially equivalent to those of this Agreement.

(b) The Receiving Party may provide the Disclosing Party's Confidential Information:

(i) to the Receiving Party's employees, consultants, advisors and contractors who have a need to know such Confidential Information and are bound by an obligation to maintain the confidentiality of the Disclosing Party's Confidential Information;

(ii) to patent offices or regulatory authorities in order to seek or obtain patent rights or approval to conduct clinical trials, or to gain regulatory approvals;

(iii) as reasonably required for development of Devices, in accordance with normal and customary commercial practice; or

(iv) if such disclosure is required by law (including by rules or regulations of any securities exchange) or to defend or prosecute litigation or arbitration; provided, that prior to such disclosure, to the extent permitted by law or such rules or regulations, the Receiving Party promptly notifies the Disclosing Party of such requirement and furnishes only that portion of the Disclosing Party's Confidential Information that the Receiving Party is legally required to furnish.

10. Limitations of Liability.

10.1 EXCEPT IN CONNECTION WITH PULSE TECHNOLOGIES' INDEMNIFICATION OBLIGATIONS UNDER SECTION 6, IN NO EVENT SHALL EITHER PARTY OR ITS AFFILIATES, BE LIABLE FOR ANY INDIRECT, INCIDENTAL, SPECIAL OR CONSEQUENTIAL DAMAGES (INCLUDING LOST PROFITS, LOST DATA, OR LOSS OF USE) ARISING OUT OF THIS AGREEMENT, REGARDLESS OF WHETHER SUCH DAMAGES ARE BASED ON TORT, WARRANTY, CONTRACT OR ANY OTHER LEGAL THEORY, EVEN IF ADVISED OF THE POSSIBILITY OF SUCH DAMAGES. THIS EXCLUSION IS INDEPENDENT OF ANY OTHER REMEDY SET FORTH IN THIS AGREEMENT.

10.2 TO THE FULLEST EXTENT PERMITTED BY LAW, IKARIA'S LIABILITY TO PULSE TECHNOLOGIES UNDER THIS AGREEMENT IS LIMITED TO THE AGGREGATE AMOUNTS PAID OR PAYABLE BY PULSE TECHNOLOGIES TO IKARIA IN RESPECT OF THE RELEVANT

PURCHASE ORDER FROM WHICH THE CLAIM AROSE.

11. Miscellaneous.

11.1 Notices. All notices required or permitted to be given under this Agreement must be in writing and delivered to the other Party as set forth below. Notices are validly given upon the earlier of confirmed receipt by the receiving Party or three business days after dispatch by a reputable courier or certified mail, return receipt requested. Either Party may change its designated contact and address for purposes of notice by giving notice to the other Party in accordance with these provisions.

If to Ikaria:

INO Therapeutics LLC
Perryville III Corporate Park
53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel

If to Pulse Technologies:

Bellerophon Pulse Technologies LLC
Perryville III Corporate Park
53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel

11.2 Escalated Dispute Resolution. Prior to pursuing legal remedies hereunder, the Parties' relationship managers agree to negotiate in good faith to resolve any disputes arising during performance of this Agreement. If such negotiations and meetings do not resolve the dispute within 10 business days after notice of the dispute, then a senior executive from each Party will meet within 10 days or as agreed between them to attempt to resolve such dispute. If the dispute is not resolved to the satisfaction of these executives within 10 days, then either Party may pursue all available legal remedies. Notwithstanding the foregoing, either Party may seek injunctive relief with respect to any disputed matter without following the dispute resolution procedure set forth above.

11.3 Force Majeure. Neither Party will be liable for any failure or delay in performance of its obligations under this Agreement to the extent such failure or delay is caused by any event beyond such Party's reasonable control, which may include fire, flood, explosion, unavailability of utilities or raw materials, labor difficulties, war, riot, act of God, export control regulation, or other laws or regulations, action or failure to act of any governmental authority, or any judgment, injunction or order of a court, administrative agency or regulatory authority having the effect of preventing or adversely affecting either Party's performance under this Agreement.

11.4 Independent Contractors. The relationship of the Parties established under this Agreement is that of independent contractors and neither Party is a partner, employee, agent or joint venturer of or with the other.

11.5 Assignment. Except as otherwise provided in this Section 11.5, neither this Agreement nor any part hereof may be assigned or transferred by either Party, whether by operation of law or otherwise, without the other Party's prior written consent. Notwithstanding the foregoing, either Party

may assign this Agreement, without the other Party's prior consent, in the event of a sale or transfer of the business as to which this Agreement relates, whether such sale or transfer occurs by merger, reorganization, asset and/or stock purchase, or by any other means, provided that the assignee agrees in writing to assume all of the assignor's obligations under this Agreement. Any assignment or purported assignment in violation hereof shall be void. This Agreement will be binding upon and inure to the benefit of the Parties and their permitted successors and assigns.

11.6 Headings; Construction; Interpretation. Headings used herein are for convenience only and shall not in any way affect the construction of or be taken into consideration in interpreting this Agreement. The terms of this Agreement represent the results of negotiations between the Parties and their representatives, each of which has been represented by counsel of its own choosing, and neither of which has acted under duress or compulsion, whether legal, economic or otherwise. Accordingly, the terms of this Agreement shall be interpreted and construed in accordance with their usual and customary meanings, and each of the Parties hereby waives the application in connection with the interpretation and construction of this Agreement of any rule of law to the effect that ambiguous or conflicting terms or provisions contained in this Agreement shall be interpreted or construed against the Party whose attorney prepared the executed draft or any earlier draft of this Agreement. Any reference in this Agreement to an Article, Section, subsection, paragraph, clause or Exhibit shall be deemed to be a reference to any Article, Section, subsection, paragraph, clause or Exhibit, of or to, as the case may be, this Agreement. Except where the context otherwise requires, (a) any definition of or reference to any agreement, instrument or other document refers to such agreement, instrument other document as from time to time amended, supplemented or otherwise modified (subject to any restrictions on such amendments, supplements or modifications set forth herein or therein); (b) any reference to any law refers to such law as from time to time enacted, repealed or amended; (c) the words "herein," "hereof" and "hereunder," and words of similar import, refer to this Agreement in its entirety and not to any particular provision hereof; (d) the words "include," "includes," "including," "exclude," "excludes," and "excluding," shall be deemed to be followed by the phrase "but not limited to," "without limitation" or words of similar import; and (e) all references in this Agreement to "days" will, unless otherwise specified herein, mean calendar days.

11.7 No Third Party Beneficiaries. No provisions of this Agreement are intended to confer or give, or will be construed to confer or give, to any person or entity other than Ikaria and Pulse Technologies any rights, remedies or other benefits under or by reason of this Agreement.

11.8 Severability. If any provision of this Agreement is determined by a court of competent jurisdiction to be invalid or unenforceable in any respect, such determination will not impair or affect the validity, legality or enforceability of the remaining provisions hereof, and each provision is hereby declared to be

separate, severable and distinct. To the extent that any such provision is found to be invalid, illegal or unenforceable, the Parties will negotiate in good faith to substitute for such provision, to the extent possible, a new provision that most nearly effects the Parties' original intent in entering into this Agreement or to provide an equitable adjustment in the event no such provision can be added. The other provisions of this Agreement will remain in full force and effect.

11.9 Entire Agreement. This Agreement constitutes the entire agreement between the Parties with respect to the subject matter hereof and supersedes all prior communications, representations or agreements, whether oral or written. No modifications, amendments, or waiver of any term, condition or provision of this Agreement will be binding on either Party unless in writing and signed by an authorized representative of each Party.

11.10 Governing Law. This Agreement shall be governed by and construed in accordance with the laws of the State of New Jersey, USA, without giving effect to any conflict of law provisions.

11.11 Counterparts. This Agreement may be executed in counterparts each of which, when executed and delivered, shall be original, but all such counterparts shall constitute one and the same document. The Parties agree that signatures transmitted via portable document format (PDF) shall be deemed originals until originals replace such copies.

IN WITNESS WHEREOF, each of the Parties has caused this Device Clinical Supply Agreement to be executed on its behalf by a duly authorized officer on the date first set forth above.

INO THERAPEUTICS LLC d/b/a IKARIA	BELLEROPHON PULSE TECHNOLOGIES LLC
By: /s/ Matthew M. Bennett	By: /s/ Daniel Tassé
Name: Matthew M. Bennett	Name: Daniel Tassé
Title: Vice President & Secretary	Title: Chief Executive Officer

EXHIBIT A
DEVICES

INOpulse DS for PAH

INOpulse DS-C for COPD

INOpulse Mark 1

CCM (Clinical Control Module)

EXHIBIT B
PRICING

Pricing for the Devices shall be as follows:

Device	Pricing
INOpulse DS for PAH	COGS plus [**]%
INOpulse DS-C for COPD	COGS plus [**]%
INOpulse Mark 1	COGS plus [**]%
CCM (Clinical Control Module)	COGS plus [**]%

Ikaria shall invoice Pulse Technologies as follows in respect of each Purchase Order accepted by Ikaria hereunder:

(a) promptly following acceptance of a Purchase Order by Ikaria, Ikaria shall invoice Pulse Technologies for estimated COGS for the Devices covered by the applicable Purchase Order; and

(b) upon completion of the Devices (i.e., the Devices are available for pickup by Pulse Technologies EXW Ikaria's Facilities as described in Section 2.7 of the Agreement), Ikaria shall invoice Pulse Technologies for (i) any actual COGS in excess of estimated COGS (or if actual COGS is less than estimated COGS, then Ikaria shall grant a credit in the appropriate amount), and (ii) the additional percentage fee specified in this Exhibit B.

Pulse Technologies shall promptly pay each invoice hereunder (but in all events, within 30 days after each such invoice has been issued by Ikaria), it being acknowledged and agreed by Pulse Technologies that (a) Ikaria shall not be required to take any further action with respect to the Devices under a Purchase Order unless and until Pulse Technologies has paid the applicable invoice in full, and (b) Ikaria shall not be required to release any Devices for pick-up until the applicable invoice has been paid in full.

All amounts not paid when due shall bear interest from the due date at the rate of the lesser of (a) [**] percent ([**]%) per month or (b) the maximum amount permitted by applicable law.

Exhibit 10.12

Execution Version

Confidential Materials omitted and filed separately with the
Securities and Exchange Commission. Double asterisks denote omissions.

DRUG CLINICAL SUPPLY AGREEMENT

This Drug Clinical Supply Agreement (this "Agreement") is entered into as of February 9, 2014 (the "Effective Date") by and between by and between INO Therapeutics LLC, a Delaware limited liability company, with offices at Perryville III Corporate Park, 53 Frontage Road, Third Floor, Hampton, NJ 08827 d/b/a Ikaria ("Ikaria"), and Bellerophon Pulse Technologies LLC, a Delaware limited liability company, with offices at Perryville III Corporate Park, 53 Frontage Road, Third Floor, Hampton, NJ 08827 d/b/a Ikaria ("Pulse Technologies"). Ikaria and Pulse Technologies may be individually referred to as a "Party" and together as the "Parties."

WHEREAS, the Parties were formerly owned by a common parent company, Ikaria, Inc. ("Ikaria Parent Company");

WHEREAS, Pulse Technologies has, as part of certain spin-out transactions (the "Spin-Out"), ceased to be a direct or indirect subsidiary of Ikaria Parent Company, and is now therefore not owned by, or affiliated with, either Ikaria Parent Company or Ikaria;

WHEREAS, Pulse Technologies is engaged in the business of developing, manufacturing, and commercializing products for (a) pulmonary hypertension secondary to chronic obstructive pulmonary disease ("COPD"), (b) primary or idiopathic pulmonary arterial hypertension ("PAH"), and pulmonary hypertension secondary to idiopathic pulmonary fibrosis ("IPF"; collectively, the "Pulse Technologies Clinical Programs");

WHEREAS, prior to the Spin-Out, Ikaria manufactured nitric oxide for inhalation (the "NO") and corresponding placebo ("Placebo"; collectively, "Product") for use by Pulse Technologies as part of the Pulse Technologies Clinical Programs; and

WHEREAS, Pulse Technologies wishes Ikaria to continue on a short term basis to manufacture and supply, and Ikaria wishes to continue to manufacture and supply, the Product for Pulse Technologies, all subject to and in accordance with the terms and conditions of this Agreement.

NOW THEREFORE, in consideration of the foregoing premises, which are incorporated into and made a part of this Agreement, and of the mutual covenants which are recited herein, the Parties agree as follows:

1. Definitions.

1.1 "Affiliate" means, with respect to a Party, any Person directly or indirectly controlling, controlled by or under common control with, such Party. For purposes of this definition only, "control" of a Person shall mean the ability, directly or indirectly, to direct the activities of the relevant Person, and with respect to corporate entities shall mean (a) ownership or direct control of fifty percent (50%) or more of the outstanding voting stock or other ownership interest of such Person, or (b) direct or indirect possession, of the power to elect or appoint fifty percent (50%) or more of the members of the governing body of such Person. Notwithstanding the foregoing or any direct or indirect control relationship that exists between them, Ikaria and Pulse Technologies shall be deemed not to be Affiliates of one another.

1.2 "Certificate of Analysis" means a document, signed by an authorized representative of Ikaria, describing (i) the Specifications for the applicable Product; (ii) the testing and methods applied to a batch or lot for such Product in order to verify compliance with the Specifications, and (iii) the results of such testing.

1.3 "cGMP" means the regulatory requirements for current good manufacturing practices promulgated by the FDA under the Food and Drug Act, including at 21 C.F.R. parts 210, 211 and 820. and under the Public Health Service Act, Biological Products, 21 C.F.R. parts 600 *et seq.*, as the same may be amended from time to time.

1.4 "COGS" means, as to Ikaria and its Affiliates, with respect to the Product, the aggregate of internal and external costs of Ikaria and its Affiliates to manufacture such Product, calculated as follows: (a) to the extent that Ikaria or its Affiliates performs all or any part of the manufacturing of such Product, the actual direct material costs and direct labor costs for, plus manufacturing overhead reasonably allocable to, such manufacturing of such Product (which may include the costs of audits, all directly incurred manufacturing variances, manufacturing administrative and facilities costs (including depreciation)), all calculated in accordance with GAAP; and (b) to the extent that manufacturing of such Product is performed by a Third Party, the costs paid to such Third Party for such activities and the reasonably allocated direct labor costs incurred by Ikaria or any of its Affiliates in managing and overseeing the Third Party relationship, determined in accordance with GAAP.

1.5 "Confidential Information" means information disclosed by a Party or its Affiliate (such Party referred to as the "Disclosing Party") to the other Party or its Affiliate (such Party referred to as the "Receiving Party"), which information relates either directly or indirectly to the business of the Disclosing Party, including information and data regarding the manufacture or use, pre-clinical or clinical data regarding, the status of research or development of the Product. Confidential Information of the Disclosing Party excludes any information that the Receiving Party can establish by written records: (a) was known by the Receiving Party prior to receipt from the Disclosing Party; (b) was disclosed to the Receiving Party by a Third Party having the right to do so; (c) was, or subsequently became, publicly known through no fault of the Receiving Party or its Affiliates; or (d) was concurrently or subsequently developed by personnel of the Receiving Party without having had access to the Disclosing Party's Confidential Information.

1.6 "COPD" shall have the meaning set forth in the recitals to this Agreement.

1.7 "Effective Date" shall have meaning set forth in the preamble to this Agreement.

1.8 "Facility" means Ikaria's manufacturing facilities located in Port Allen, LA, or such other manufacturing site(s) specified by Ikaria from time to time.

1.9 "FDA" means the United States Food and Drug Administration or any successor organization.

1.10 "Federal Health Care Programs" shall have the meaning set forth in Section 7.3.

1.11 "Forecast" shall have the meaning set forth in Section 2.4.

1.12 "Ikaria Parent Company" shall have the meaning set forth in the recitals to this Agreement.

1.13 "Intellectual Property" means, collectively, patents, trademarks, copyrights (including to any software, whether in object code or source code form), trade secrets, know-how, and any other intellectual or proprietary property or rights.

1.14 "IPF" shall have the meaning set forth in the recitals to this Agreement.

1.15 "NO" shall have the meaning set forth in the recitals to this Agreement.

1.16 "PAH" shall have the meaning set forth in the recitals to this Agreement.

1.17 "Person" means any individual, governmental authority, partnership, corporation, limited liability company, unincorporated organization or association, any trust or any other business entity.

1.18 "Placebo" shall have the meaning set forth in the recitals to this Agreement.

1.19 "Product" shall have the meaning set forth in the recitals to this Agreement.

1.20 "Product IP" shall have the meaning set forth in Section 2.9.

1.21 "Pulse Technologies Clinical Programs" shall have the meaning set forth in the recitals to this Agreement.

1.22 "Quality Agreement" has the meaning set forth in Section 4.1.

1.23 "Regulatory Authority" means any competent governmental authority which regulates the manufacture, development or sale of the Product.

1.24 "Specifications" means, with respect to the Product and Placebo, the specifications in effect for the Product immediately prior to the Spin-Out.

1.25 "Term" has the meaning set forth in Section 10.1.

1.26 "Third Party" means any Person who is not a Party or an Affiliate of a Party.

2. Supply of Product.

2.1 Obligations of Ikaria. During the Term of this Agreement, Ikaria shall use commercially reasonable efforts to manufacture and supply Pulse Technologies' requirements, and Pulse Technologies shall acquire from Ikaria its requirements, for the Product for the Pulse Technologies Clinical Programs in accordance with the terms of this Agreement and the Quality Agreement. Pulse Technologies acknowledges and agrees that nothing in this Agreement shall require Ikaria to hire, obtain, or retain additional resources of any type (whether personnel, infrastructure, or otherwise), or to make capital expenditures of any kind, in order to manufacture and supply the Product, nor shall anything in this Agreement require Ikaria to prioritize manufacturing and supplying the Product to Pulse Technologies over performing similar services for its own benefit.

2.2 Obligations of Pulse Technologies. Pulse Technologies shall provide Ikaria with such information and cooperation as may be necessary for the manufacture and supply of the Product in accordance with this Agreement and the Quality Agreement.

2.3 Commercial Supply. If Pulse Technologies desires to obtain supply of any Product (or any variant thereof or any version with different specifications) for commercial use, then prior to negotiating the terms of an agreement for such agreement, Pulse Technologies shall promptly notify Ikaria thereof in writing. Ikaria shall, within 60 days after receipt of such notice, indicate to Pulse Technologies in writing whether Ikaria or any of its Affiliates wishes to enter into such an agreement and, if Ikaria indicates that Ikaria or any of its Affiliates do wish to enter into such agreement, the Parties shall negotiate in good faith to enter into mutually agreeable terms pursuant to which Ikaria or any of its

Affiliates would enter into agreement with Pulse Technologies. In such negotiations, Ikaria may elect to supply [**]% of Pulse Technologies requirements for the Product(s) in question, or such lesser quantity as Ikaria may elect in its sole discretion. If either (a) Ikaria indicates it does not wish to pursue such agreement, (b) Ikaria fails to indicate its interest within such 30 day period or (c) Ikaria indicates it wishes to enter into such agreement but the Parties fail to reach agreement on the terms of such agreement or to execute a definitive agreement prior to 90 days after the date of Ikaria's indication of interest, then Pulse Technologies shall be free, without any further obligation to Ikaria under this Agreement with respect thereto, to enter into such an agreement with a Third Party; provided that, in the event clause (c) of this sentence is applicable, if Pulse Technologies proposes to enter such agreement with a Third Party on terms that are materially less favorable to Pulse Technologies than the terms last offered in writing to Pulse Technologies by Ikaria, then (i) Pulse Technologies shall, prior to entering into such agreement with such Third Party, offer such terms to Ikaria, (ii) Ikaria shall have 15 days after the date of receipt of such offer from Pulse Technologies to notify Pulse Technologies in writing of its acceptance of such offer and (iii) (A) if Ikaria so accepts, the Parties shall promptly enter into a definitive agreement for the commercial supply of such Product(s) on such terms, or (B) if Ikaria does not accept, then Pulse Technologies shall be free, without any further obligation to Ikaria under this Agreement with respect thereto, to enter into such commercial supply agreement with a Third Party; provided further that if Pulse Technologies does not enter into a definitive agreement for such commercial supply with a Third Party within 180 days after the expiration of Ikaria's under this Section 2.3, Ikaria's rights under this Section 2.3 shall be reinstated.

2.4 Forecasts; Committed Quantities.

(a) Commencing on the Effective Date and on the first business day of each calendar month thereafter, Pulse Technologies shall submit to Ikaria a written rolling forecast of the quantity of each Product which Pulse Technologies expects to order from Ikaria over the next 36 months (the "Forecast"). The Forecast shall constitute a non-binding, good faith estimate provided by Pulse Technologies solely to assist Ikaria in production planning, and shall not represent a purchase commitment by Pulse Technologies or a supply commitment by Ikaria; provided, however, that the first six months of each such forecast shall constitute a binding purchase order hereunder for the specific Product for the quantities specified for that six-month period.

(b) Each purchase of Product, and any acknowledgment thereof, shall be governed by the terms of this Agreement. If a Party uses forms or documents to place or accept a purchase order that contain terms and conditions that are in addition to or contrary to those in this Agreement, the Parties agree and acknowledge that such forms or documents will be used for convenience only, and that no terms or conditions set forth therein, except with respect to quantity, shall be of any force or effect.

2.5 Delivery. Ikaria shall deliver the Product to a carrier selected by Pulse Technologies. The Products shall be made available EXW the Facility (Incoterms 2010). Title and risk of loss will pass to Pulse Technologies when the Products are made available to the carrier selected by Pulse Technologies. Pulse Technologies is responsible for payment of all shipment costs, including any insurance necessary to guard against loss or damage during shipment.

2.6 Certificates. An appropriate Certificate of Analysis shall be provided with the shipment of each batch or lot of Product delivered to Pulse Technologies.

2.7 Shipping Instructions. Pulse Technologies will provide Ikaria with packaging and shipping instructions, including temperature requirements, temperature monitoring instructions and packaging specifications. Notwithstanding any other provision of this Agreement, Ikaria will not be

liable for any loss or damage caused by Ikaria's compliance with Pulse Technologies' packaging and shipping instructions or any loss or damage caused by Pulse Technologies' carrier.

2.8 Limited Use. Pulse Technologies acknowledges and agrees that the Products may only be used in the Pulse Technologies Clinical Programs (and, if Ikaria has exercised its right under Section 2.3 to supply Products for commercial use, then also for that limited commercial use) and for no other uses or purposes.

2.9 No Licenses. Pulse Technologies acknowledges and agrees that nothing in this Agreement grants, or shall be deemed or interpreted to grant, to Pulse Technologies any right, title, or interest in or to any Intellectual Property reflected, contained, or incorporated in, or practice under, as part of or in the process of manufacturing and delivery of the Product (collectively, the "Product IP").

2.10 Changes. Ikaria shall use commercial reasonable efforts to accommodate any reasonable changes to the Specifications for Product requested by Pulse Technologies, it being understood and agreed that Pulse Technologies shall bear any and all costs associated with such changes.

3. Price and Payment

3.1 Pricing. Pricing for the Product and Placebo shall be COGS plus [**] percent.

3.2 Payment. Ikaria shall invoice Pulse Technologies at the time of shipment of Product in accordance with this Agreement. Payment of an invoice is due the later of (a) thirty (30) days from the date of Pulse Technologies' receipt of invoice; or (b) delivery of Product to the carrier in accordance with Section 2.5.

3.3 Late Payments. All amounts not paid when due shall bear interest from the due date at the rate of the lesser of (a) [**] percent ([**]%) per month or (b) the maximum amount permitted by applicable law.

4. Quality.

4.1 Quality Agreement. The Parties shall use reasonable efforts to negotiate and conclude a quality agreement ("Quality Agreement") within 60 days after the Effective Date. The Quality Agreement shall detail the division of responsibilities between the Parties regarding quality and regulatory controls and reporting concerning the Product. In the case of a conflict between the Quality Agreement and this Agreement, the terms of this Agreement shall control unless such term in the Quality Agreement expressly references such conflict and the Parties intend to have the Quality Agreement control such provision.

4.2 Quality Assurance. Ikaria shall perform quality testing using assays agreed to by the Parties in order to assure that Product complies with the Specification, and shall retain samples of Product as required by applicable law. Ikaria shall maintain records, including batch or lot records, with respect to the quality testing.

5. Non-Conforming Product.

(a) Each batch or lot of Product delivered to Pulse Technologies hereunder shall be accompanied by a Certificate of Analysis. Pulse Technologies shall have 30 days from the date of receipt of Product to inspect and reject acceptance by written notice to Ikaria; provided, however, that any such notice shall set forth Pulse Technologies' reasons for rejection in reasonable detail and provided, further,

that Pulse Technologies may reject Product only if: (i) Pulse Technologies claims a material breach of Ikaria's representations and warranties in Section 7.2 of this Agreement with respect to such Product; or (ii) Ikaria has failed to deliver a Certificate of Analysis for such Product. If Ikaria does not receive Pulse Technologies' written notice of rejection within such 30 day period, Pulse Technologies shall be deemed to have accepted such Product.

(b) If Pulse Technologies provides Ikaria with a timely notice of rejection as set forth in Section 5(a), Pulse Technologies shall return the rejected Product to Ikaria at Ikaria's expense. Ikaria shall have 30 days following receipt of the rejected Product in which to test such Product. If Ikaria does not dispute a rejection, Ikaria shall rework or replace the rejected Product, at Ikaria's expense and such rework or replacement shall constitute Pulse Technologies' exclusive remedy and Ikaria's sole liability with respect to such rejection. If Ikaria disputes a rejection, Ikaria shall provide Pulse Technologies with written notice of such dispute within 30 days after receiving the returned Product, and the Parties shall use commercially reasonable efforts to resolve the dispute amicably and promptly. If the Parties are unable to reach a resolution within 30 days after Pulse Technologies' notice of rejection, the returned Product shall be submitted to an independent laboratory or consultant mutually acceptable to the Parties, whose decision as to the conformity of such Product with the applicable Specification shall be final and binding. The Party against whom the dispute is decided shall pay any charges for such laboratory or consultant. If the laboratory or consultant determines that the returned Product did not conform to the Specification, Ikaria shall rework or replace the rejected Product at no charge to Pulse Technologies, and such replacement shall constitute Pulse Technologies' exclusive remedy and Ikaria's sole liability with respect to such rejected Product.

6. Audit Rights. Pulse Technologies shall have the right to conduct reasonable audits and inspections of the Facility, Ikaria's manufacturing operations, and Ikaria's records relating to the manufacture of Product under this Agreement. Ikaria shall reasonably cooperate with Pulse Technologies in conducting such audits and inspections.

7. Warranties.

7.1 General Warranties. Each Party warrants to the other Party that (a) it has the right and authority to enter into this Agreement and to carry out its obligations hereunder; (b) it is validly existing in the jurisdiction in which it is incorporated and is authorized to do business under the laws of each jurisdiction in which it engages in business activities; and (c) it is not aware of any legal, contractual or other restriction, limitation or condition that might adversely affect its ability to perform its obligations hereunder.

7.2 Warranties by Ikaria. Ikaria warrants to Pulse Technologies that the Product delivered hereunder shall (a) conform in all material respects with its applicable Specifications and (b) be manufactured in compliance applicable law (including applicable cGMPs). Pulse Technologies agrees that its exclusive remedies, and Ikaria's sole liabilities, with respect to any breach of the warranty set forth in this Section 7.2 are set forth in Section 5(b) of this Agreement.

7.3 Debarment. Each party represents and warrants that, as of the Effective Date and throughout the Term, it (and each of its employees and agents) (a) is not currently excluded, debarred or otherwise ineligible to participate in the Federal health care programs as defined in 42 U.S.C. 1320a7b(f) (the "Federal Health Care Programs"); (b) has not been convicted of a criminal offense related to the provision of healthcare items or services but yet to be excluded, debarred or otherwise declared ineligible to participate in the Federal Health Care Programs; and (c) is not under investigation or otherwise aware of any circumstances which may result in it (or its agents, employees or any substitutes thereof performing any duties under this Agreement) being excluded from participation in the Federal Health

Care Programs.

7.4 DISCLAIMER. EXCEPT AS EXPRESSLY PROVIDED HEREIN, NEITHER PARTY MAKES NOR RECEIVES ANY REPRESENTATION OR WARRANTY OF ANY KIND, EXPRESS, IMPLIED, STATUTORY OR OTHERWISE, OR ARISING FROM A COURSE OF DEALING OR USAGE OF TRADE PRACTICE, WITH REGARD TO THE PRODUCT OR OTHERWISE UNDER THIS AGREEMENT, INCLUDING THE IMPLIED WARRANTIES OF MERCHANTABILITY, NON-INFRINGEMENT, OR FITNESS FOR A PARTICULAR PURPOSE.

8. Indemnity. Pulse Technologies shall indemnify, defend and hold Ikaria, its Affiliates and their respective directors, officers, employees, agents, successors and assigns harmless from and against any damages, losses, judgments, claims, suits, actions, liabilities, costs and expenses (including, but not limited to, reasonable attorneys' fees), as and when incurred, resulting from any Third Party claims or suits arising out of the ownership, use, handling, development, distribution, marketing, or sale of any Product.

9. Compliance.

9.1 Compliance with Laws. Each Party shall comply with all applicable laws and regulations governing the performance of such Party's obligations under this Agreement. Without limiting the foregoing, each Party shall comply with applicable US and other laws, rules and regulations that govern the import, export and re-export of the Product, including the U.S. Export Administration Regulations, and will obtain any required export and import authorizations to perform its obligation hereunder.

9.2 Regulatory Filings. Pulse Technologies, at its expense, shall be solely responsible for the preparation, filing, and maintenance of all regulatory documents and all governmental permits, licenses and other approvals as may be necessary with respect to the formulation, marketing, distribution, sale, and use of the Product.

9.3 Permits. Ikaria at its expense shall be solely responsible for, and has the obligation to prepare, file, and maintain during the Term, all licenses, permits, and approvals as may be necessary with respect to the manufacture of Product at the Facility.

10. Term and Termination.

10.1 Term. Unless earlier terminated under Section 10.2, this Agreement shall commence as of the Effective Date, and unless earlier terminated pursuant to Section 10.2, shall expire on a Product by Product basis as follows (the "Term"):

(a) With respect to Product for the Pulse Technologies Clinical Program for COPD, at the point in time that Pulse Technologies is no longer actively and continuously engaged that Pulse Technologies Clinical Program;

(b) With respect to Product for the Pulse Technologies Clinical Program for PAH, at the point in time that Pulse Technologies is no longer actively and continuously engaged that Pulse Technologies Clinical Program; and

(c) With respect to Product for the Pulse Technologies Clinical Program for IPF, at the point in time that Pulse Technologies is no longer actively and continuously engaged that Pulse Technologies Clinical Program.

10.2 Termination. This Agreement may be terminated by either Party upon 60 days written notice of the other Party's material breach of any provision of this Agreement; provided, however, that the breaching Party will have an opportunity to (a) cure the breach during the 60 day notice period, or (b) provide the non-breaching Party with a plan to remedy the breach within the 60 day notice period, and if so cured, no termination will be deemed to have occurred as long as the breaching Party diligently pursues the plan to remedy the breach and completes such plan in accordance with the time frame agreed to by the Parties (such time frame not to exceed an additional 60 days). In addition, Ikaria may terminate this Agreement for any reason or no reason by giving 30 days' notice to Pulse Technologies.

10.3 Effect of Termination. Termination or expiration of this Agreement shall not release either Party from any liability, right of action or other obligation which has arisen prior to such termination or expiration, including Ikaria's obligation to deliver to Pulse Technologies such quantity of Product under any accepted purchase order to the effective date of termination or expiration, and Pulse Technologies' obligation to pay Ikaria the amount set forth in such purchase order (except in the case of a material breach hereof by Pulse Technologies, in which case Ikaria may elect not to supply and manufactured but not yet delivered Product). Notwithstanding any expiration or termination of this Agreement, the following provisions shall survive: Sections 2.3, 2.8, 2.9, 5, 7.4, 8, 10.3, 11.1, 12 and 13.

11. Confidentiality.

11.1 Non-Use and Non-Disclosure of Confidential Information. Each Receiving Party agrees that all Confidential Information of the Disclosing Party (a) shall not be used by the Receiving Party except to perform its obligations or exercise its rights under this Agreement, (b) shall be maintained in confidence by the Receiving Party, and (c) except as permitted by Section 11.2, shall not be disclosed by the Receiving Party to any Person without the prior written consent of the Disclosing Party.

11.2 Permitted Disclosures.

(a) The Receiving Party may provide the Disclosing Party's Confidential Information (i) to its Affiliates and to their employees, consultants, advisors, and contractors who have a need to know such Confidential Information for purposes of the Receiving Party exercising or granting licenses or sublicenses, (ii) in communications with existing or bona fide prospective acquirers, merger partners, lenders or investors, in each case of (i) and (ii), on a need to know basis and under appropriate confidentiality provisions substantially equivalent to those of this Agreement.

(b) The Receiving Party may provide the Disclosing Party's Confidential Information:

(i) to the Receiving Party's employees, consultants, advisors and contractors who have a need to know such Confidential Information and are bound by an obligation to maintain the confidentiality of the Disclosing Party's Confidential Information;

(ii) to patent offices or regulatory authorities in order to seek or obtain patent rights or approval to conduct clinical trials, or to gain regulatory approvals;

(iii) as reasonably required for development of the Product, in accordance with normal and customary commercial practice; or

(iv) if such disclosure is required by law (including by rules or regulations of any securities exchange) or to defend or prosecute litigation or arbitration; provided, that prior to such disclosure, to the extent permitted by law or such rules or regulations, the Receiving Party promptly

notifies the Disclosing Party of such requirement and furnishes only that portion of the Disclosing Party's Confidential Information that the Receiving Party is legally required to furnish.

12. Limitations of Liability.

12.1 EXCEPT IN CONNECTION WITH PULSE TECHNOLOGIES' INDEMNIFICATION OBLIGATIONS UNDER SECTION 8, IN NO EVENT SHALL EITHER PARTY OR ITS AFFILIATES, BE LIABLE FOR ANY INDIRECT, INCIDENTAL, SPECIAL OR CONSEQUENTIAL DAMAGES (INCLUDING LOST PROFITS, LOST DATA, OR LOSS OF USE) ARISING OUT OF THIS AGREEMENT, REGARDLESS OF WHETHER SUCH DAMAGES ARE BASED ON TORT, WARRANTY, CONTRACT OR ANY OTHER LEGAL THEORY, EVEN IF ADVISED OF THE POSSIBILITY OF SUCH DAMAGES. THIS EXCLUSION IS INDEPENDENT OF ANY OTHER REMEDY SET FORTH IN THIS AGREEMENT.

12.2 TO THE FULLEST EXTENT PERMITTED BY LAW, IKARIA'S LIABILITY TO PULSE TECHNOLOGIES UNDER THIS AGREEMENT IS LIMITED TO THE AGGREGATE AMOUNTS PAID OR PAYABLE BY PULSE TECHNOLOGIES TO IKARIA IN RESPECT OF THE RELEVANT PURCHASE ORDER FROM WHICH THE CLAIM AROSE.

13. Miscellaneous.

13.1 Notices. All notices required or permitted to be given under this Agreement must be in writing and delivered to the other Party as set forth below. Notices are validly given upon the earlier of confirmed receipt by the receiving Party or three business days after dispatch by a reputable courier or certified mail, return receipt requested. Either Party may change its designated contact and address for purposes of notice by giving notice to the other Party in accordance with these provisions.

If to Ikaria:

INO Therapeutics LLC
Perryville III Corporate Park
53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel

If to Pulse Technologies:

Bellerophon Pulse Technologies LLC
Perryville III Corporate Park
53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel

13.2 Escalated Dispute Resolution. Prior to pursuing legal remedies hereunder, the Parties' relationship managers agree to negotiate in good faith to resolve any disputes arising during performance of this Agreement. If such negotiations and meetings do not resolve the dispute within 10 business days after notice of the dispute, then a senior executive from each Party will meet within 10 days or as agreed between them to attempt to resolve such dispute. If the dispute is not resolved to the satisfaction of these

executives within 10 days, then either Party may pursue all available legal remedies. Notwithstanding the foregoing, either Party may seek injunctive relief with respect to any disputed matter without following the dispute resolution procedure set forth above.

13.3 Force Majeure. Neither Party will be liable for any failure or delay in performance of its obligations under this Agreement to the extent such failure or delay is caused by any event beyond such Party's reasonable control, which may include fire, flood, explosion, unavailability of utilities or raw materials, labor difficulties, war, riot, act of God, export control regulation, or other laws or regulations, action or failure to act of any governmental authority, or any judgment, injunction or order of a court, administrative agency or regulatory authority having the effect of preventing or adversely affecting either Party's performance under this Agreement.

13.4 Independent Contractors. The relationship of the Parties established under this Agreement is that of independent contractors and neither Party is a partner, employee, agent or joint venturer of or with the other.

13.5 Assignment. Except as otherwise provided in this Section 13.5, neither this Agreement nor any part hereof may be assigned or transferred by either Party, whether by operation of law or otherwise, without the other Party's prior written consent. Notwithstanding the foregoing, either Party may assign this Agreement, without the other Party's prior consent, in the event of a sale or transfer of the business as to which this Agreement relates, whether such sale or transfer occurs by merger, reorganization, asset and/or stock purchase, or by any other means, provided that the assignee agrees in writing to assume all of the assignor's obligations under this Agreement. Any assignment or purported assignment in violation hereof shall be void. This Agreement will be binding upon and inure to the benefit of the Parties and their permitted successors and assigns.

13.6 Headings; Construction; Interpretation. Headings used herein are for convenience only and shall not in any way affect the construction of or be taken into consideration in interpreting this Agreement. The terms of this Agreement represent the results of negotiations between the Parties and their representatives, each of which has been represented by counsel of its own choosing, and neither of which has acted under duress or compulsion, whether legal, economic or otherwise. Accordingly, the terms of this Agreement shall be interpreted and construed in accordance with their usual and customary meanings, and each of the Parties hereby waives the application in connection with the interpretation and construction of this Agreement of any rule of law to the effect that ambiguous or conflicting terms or provisions contained in this Agreement shall be interpreted or construed against the Party whose attorney prepared the executed draft or any earlier draft of this Agreement. Any reference in this Agreement to an Article, Section, subsection, paragraph, clause or Exhibit shall be deemed to be a reference to any Article, Section, subsection, paragraph, clause or Exhibit, of or to, as the case may be, this Agreement. Except where the context otherwise requires, (a) any definition of or reference to any agreement, instrument or other document refers to such agreement, instrument other document as from time to time amended, supplemented or otherwise modified (subject to any restrictions on such amendments, supplements or modifications set forth herein or therein); (b) any reference to any law refers to such law as from time to time enacted, repealed or amended; (c) the words "herein," "hereof" and "hereunder," and words of similar import, refer to this Agreement in its entirety and not to any particular provision hereof; (d) the words "include," "includes," "including," "exclude," "excludes," and "excluding," shall be deemed to be followed by the phrase "but not limited to," "without limitation" or words of similar import; and (e) all references in this Agreement to "days" will, unless otherwise specified herein, mean calendar days.

13.7 No Third Party Beneficiaries. No provisions of this Agreement are intended to confer or give, or will be construed to confer or give, to any person or entity other than Ikaria and Pulse Technologies any rights, remedies or other benefits under or by reason of this Agreement.

13.8 Severability. If any provision of this Agreement is determined by a court of competent jurisdiction to be invalid or unenforceable in any respect, such determination will not impair or affect the validity, legality or enforceability of the remaining provisions hereof, and each provision is hereby declared to be separate, severable and distinct. To the extent that any such provision is found to be invalid, illegal or unenforceable, the Parties will negotiate in good faith to substitute for such provision, to the extent possible, a new provision that most nearly effects the Parties' original intent in entering into this Agreement or to provide an equitable adjustment in the event no such provision can be added. The other provisions of this Agreement will remain in full force and effect.

13.9 Entire Agreement. This Agreement constitutes the entire agreement between the Parties with respect to the subject matter hereof and supersedes all prior communications, representations or agreements, whether oral or written. No modifications, amendments, or waiver of any term, condition or provision of this Agreement will be binding on either Party unless in writing and signed by an authorized representative of each Party.

13.10 Governing Law. This Agreement shall be governed by and construed in accordance with the laws of the State of New Jersey, USA, without giving effect to any conflict of law provisions.

13.11 Counterparts. This Agreement may be executed in counterparts each of which, when executed and delivered, shall be original, but all such counterparts shall constitute one and the same document. The Parties agree that signatures transmitted via portable document format (PDF) shall be deemed originals until originals replace such copies.

IN WITNESS WHEREOF, each of the Parties has caused this Drug Clinical Supply Agreement to be executed on its behalf by a duly authorized officer on the date first set forth above.

INO THERAPEUTICS LLC d/b/a IKARIA	BELLEROPHON PULSE TECHNOLOGIES LLC
By: /s/ Matthew M. Bennett	By: /s/ Daniel Tassé
Name: Matthew M. Bennett	Name: Daniel Tassé
Title: Vice President & Secretary	Title: Chief Executive Officer

[Signature Page to Drug Clinical Supply Agreement]

Exhibit 10.13

Execution Version

Confidential Materials omitted and filed separately with the
Securities and Exchange Commission. Double asterisks denote omissions.

EMPLOYEE MATTERS AGREEMENT

by and between

IKARIA, INC.

and

BELLEROPHON THERAPEUTICS LLC

dated as of February 9, 2014

Employee Matters Agreement

This Employee Matters Agreement, dated as of February 9, 2014, is made and entered by and among Ikaria, Inc., a Delaware Corporation ("Ikaria"), and Bellerophon Therapeutics LLC, a Delaware limited liability company ("R&DCo"). Ikaria and R&DCo are sometimes referred to herein individually as a "Party" and together as the "Parties." Capitalized terms used herein and not otherwise defined shall have the meanings assigned to them by the Separation and Distribution Agreement, dated as of the date hereof, between the Parties (the "Separation and Distribution Agreement").

Recitals

WHEREAS, the Board of Directors of Ikaria has determined that it is appropriate, desirable and in the best interests of Ikaria and its stockholders to separate Ikaria into two independent companies, one for each of: (a) the Ikaria Business, which shall continue to be owned and conducted, directly or indirectly, in addition to any other line of business it may conduct, by Ikaria, and (b) the R&DCo Business, which shall be owned and conducted, directly or indirectly, by R&DCo (such separation, the "Separation");

WHEREAS, pursuant to the terms of, and as described more fully in, the Separation and Distribution Agreement, the Parties will engage in a series of transactions culminating in the distribution to holders of shares of Ikaria Capital Stock, by means of one or more special dividends, of, in the aggregate, 100% of the R&DCo Voting Units (the "Distribution"); and

WHEREAS, in furtherance of the foregoing, the Parties have agreed to enter into this Agreement for purposes of (a) addressing the treatment of Employees who provide services to the R&DCo Business, the transfer of their employment to Bellerophon Services, Inc., a wholly owned subsidiary of R&DCo ("ServicesCo"), and their participation in employee benefit plans and programs following the Distribution; (b) addressing the effect of the Distribution on certain equity awards held by such Employees; and (c) allocating assets, liabilities, rights and responsibilities with respect to employee compensation and benefits and other employment matters as a result of the Separation.

NOW, THEREFORE, in consideration of the mutual promises contained herein and in the Separation and Distribution Agreement, the Parties agree as follows:

ARTICLE I.

DEFINITIONS

Section 1.1. Definitions

"Acquisition Agreement" means that certain Agreement and Plan of Merger, dated as of December 24, 2013, by and among Ikaria, Compound Holdings I, LLC, Compound Holdings II, Inc., Compound Merger Sub I, Inc. Compound Merger Sub II, Inc. and, solely in its capacity as Stockholder Representative, New Mountain Partners II, L.P.

"Agreement" means this Employee Matters Agreement, together with the exhibits, schedules, appendices, and annexes hereto.

"COBRA" has the meaning ascribed to it in Section 5.3.

"Code" means the United States Internal Revenue Code of 1986, as amended.

"Combined Group Employee" means any Employee, or following the Distribution, any Ikaria Employee or R&DCo Employee.

"CommercialCo FMV" means the fair market value of Ikaria as of immediately following the Distribution, which fair market value shall be determined by the Ikaria Board of Directors based on the aggregate fair market value of the merger consideration payable to the Ikaria securityholders (including but not limited to stockholders, option holders and restricted stock unit holders) pursuant to the Acquisition Agreement.

"Delayed Transfer Employee" has the meaning ascribed to it in Section 3.1(b).

"Distribution" has the meaning ascribed to it in the Recitals to this Agreement.

"Employee" means any individual who, prior to the Distribution, is or was employed by any member of the Ikaria Group, including any Transferred Employee and any Delayed Transfer Employee.

"Ikaria" has the meaning set forth in the preamble to this Agreement.

"Ikaria 401(k) Plan" means the Ikaria, Inc. 401(k) Plan as in effect or as it may be amended from time to time.

"Ikaria Acquisition" means the acquisition of Ikaria pursuant to the Acquisition Agreement.

"Ikaria Equity Award" means an equity award under an Ikaria Equity Plan after the adjustment provided in Section 7.1, including the Post-Distribution Ikaria Options and the Post-Distribution Ikaria RSUs.

"Ikaria Equity Plans" mean the Ikaria Holding, Inc. 2007 Stock Option Plan and the Amended and Restated Ikaria Holdings, Inc. 2010 Long Term Incentive Plan.

"Ikaria Employee" means an individual who immediately following the Distribution remains employed by any member of the Ikaria Group. Such an individual shall remain an Ikaria Employee through his or her Transfer Date, if applicable, or other termination of employment with Ikaria. "Ikaria Employee" shall also include, where applicable, the beneficiaries and dependents of an individual described in the first sentence of this definition.

"Ikaria Group" means Ikaria, Inc. and each of its subsidiaries (other than R&DCo or any R&DCo Group Member).

"Ikaria Plan" means any Plan maintained or sponsored by any member of the Ikaria Group.

"Ikaria Stock Option" means an option to acquire Ikaria non-voting common stock issued under an Ikaria Equity Plan.

"Ikaria RSU" mean a restricted stock unit representing the right to receive one share of Ikaria non-voting common stock issued under an Ikaria Equity Plan.

"Ikaria Welfare Plans" has the meaning ascribed to it in Section 5.1(a).

"Individual Agreement" means an individual employment contract or other similar agreement that specifically pertains to any Transferred Employee or Delayed Transfer Employee.

"Outstanding Equity Award" means an equity award under an Ikaria Equity Plan outstanding as of the day prior to the Distribution Date.

"Party" or "Parties" has the meaning ascribed to it in the preamble to this Agreement.

"Plan" means any plan, policy, program, payroll practice, on-going arrangement, contract, trust, insurance policy or other agreement or funding vehicle, whether written or unwritten, providing compensation or benefits to Employees, or former Employees of any member of the Ikaria Group or R&DCo, as the case may be, in respect to their services for the Ikaria Business or the R&DCo Business.

"Post-Distribution Ikaria Option" has the meaning ascribed to it in Section 7.1(a).

"Post-Distribution Ikaria RSU" has the meaning ascribed to it in Section 7.1(b).

"Post-Distribution R&DCo Option" has the meaning ascribed to it in Section 7.1(a).

"Post-Distribution R&DCo RSU" has the meaning ascribed to it in Section 7.1(b).

"R&DCo" has the meaning ascribed to it in the preamble to this Agreement.

"R&DCo 401(k) Plan" has the meaning ascribed to it in Section 6.1.

"R&DCo Employee" means (a) each Transferred Employee and (b) effective upon the Transfer Date, each Delayed Transferred Employee. "R&DCo Employee" shall also include, where applicable, the beneficiaries and dependents of an individual described in the first sentence of this definition. Following the Distribution, all R&DCo Employees shall initially be employees of ServicesCo.

"R&DCo Equity Award" means an equity award under the R&DCo Equity Plan issued in connection with the adjustment provided in Section 7.1, including the Post-Distribution R&DCo Options and the Post-Distribution R&DCo RSUs.

"R&DCo Equity Plan" means each Ikaria Equity Plan, to the extent assumed by R&DCo in connection with the Distribution and pursuant to this Agreement.

"R&DCo FMV" means the fair market value of R&DCo equity as of immediately following the Distribution to be determined based on an independent third party valuation.

"R&DCo Plan" means any Replacement Plan, each R&DCo Equity Plan and any other Plan established and adopted by R&DCo for the benefit of any R&DCo Employee.

"Replacement Plans" has the meaning ascribed to it in Section 4.1.

"Separation" has the meaning ascribed to it in the Recitals to this Agreement.

"ServicesCo" has the meaning ascribed to it in the Recitals to this Agreement.

"Transfer Date" has the meaning ascribed to it in Section 3.1(b).

"Transferred Employee" has the meaning ascribed to it in Section 3.1(a).

ARTICLE II.

RELATIONSHIP AT AND FOLLOWING DISTRIBUTION

Section 2.1. Employees, Employee Liabilities Generally.

Except as provided in this Section 2.1, each of Ikaria and R&DCo shall be responsible for the Liabilities arising with respect to the employment by any member of the Ikaria Group or the R&DCo Group, as the case may be, of (a) each person in its employment immediately following the Distribution, and (b) each person whose last employment with any member of the Ikaria Group or the R&DCo Group was performing services for the Ikaria Business or the R&DCo Business, respectively. Notwithstanding the foregoing:

(i) to the extent that compensation is payable to any Combined Group Employee in the form of or with respect to an Ikaria Equity Award, such compensation shall be payable by Ikaria and to the extent that compensation is payable to any Combined Group Employee in the form of or with respect to an R&DCo Equity Award, such compensation shall be payable by R&DCo; and

(ii) to the extent that any benefit or compensation (including any benefit or compensation otherwise described in subclause (i)) is payable to, or any other Liability in respect of any such Combined Group Employee is expressly allocated to, Ikaria or R&DCo pursuant to the terms of this Agreement, such specific allocation shall control.

Section 2.2. Employees and Benefits Generally; Possibility of Transition Services.

As of the date hereof, all Employees actively engaged on a regular basis in the R&DCo Business are Employees of members of the Ikaria Group and such Employees participate in the Ikaria Plans. Following the Distribution Date, pursuant to this Agreement, R&DCo will establish its own Plans and the participation of R&DCo Employees in the Ikaria Plans will cease as of the Distribution Date or as soon as practicable thereafter (or Transfer Date, as applicable).

ARTICLE III.

TRANSFERRED EMPLOYEES; ASSUMPTION OF LIABILITIES

Section 3.1. Transferred Employees.

(a) General. The employment of, and all rights and obligations under any employment agreement with, any Employee identified on Schedule A hereto and any other Employee that the Parties agree to add to Schedule A after the date hereof (the

"Transferred Employees") will be transferred to ServicesCo immediately prior to the Distribution.

(b) Delayed Transfer Employees. In the event that Ikaria and R&DCo agree to transfer the employment of any other Employee of any member of the Ikaria Group to ServicesCo in connection with the Distribution, but following the Distribution Date (each, a "Delayed Transfer Employee"), then effective as of the date the employment of such Delayed Transfer Employee is transferred or such other date as may otherwise be agreed in writing by the Parties (the "Transfer Date"), R&DCo shall assume all Liabilities of the type and nature that would have been assumed by R&DCo pursuant to Section 3.2 had such Delayed Transfer Employee been a Transferred Employee as of the Distribution Date, including all Liabilities arising during the period beginning on the Distribution Date and ending on the Transfer Date that arose in the ordinary course of business with respect to such Delayed Transfer Employee.

Section 3.2. Assumption of Liabilities. Except as otherwise expressly provided for in this Agreement, as of the Distribution Date (or, if later, the Transfer Date), R&DCo shall assume and agree to pay, perform, fulfill and discharge, in accordance with their respective terms, all Liabilities to or relating to Transferred Employees, Delayed Transfer Employees, and former Employees who primarily performed services for the R&DCo Business, to the extent relating to, arising out of, or resulting from employment with any member of the Ikaria Group on or prior to the Distribution Date (or, as applicable, the Transfer Date), including, without limitation, (i) Liabilities in respect of the reduction in force described on Exhibit E to the Acquisition Agreement and any other severance, bonus, benefits or other compensation obligations to or in respect of Transferred Employees, Delayed Transfer Employees, and former Employees who primarily performed services for the R&DCo Business (including the individuals listed in either schedule to that certain letter from Ikaria to Compound Holdings II, Inc. dated as of December 24, 2013), whether arising before or after the closing of the Ikaria Acquisition, (ii) the retention bonus pool to be distributed by Ikaria as described on Exhibit E to the Acquisition Agreement and (iii) all rights and obligations under an employment agreement with such Employees (and all such employment agreements will be assigned to ServicesCo).

Section 3.3. General Principles.

(a) Non-Termination of Employment or Benefits. Except as otherwise expressly provided herein or as otherwise required by applicable law, no provision of this Agreement or the Separation and Distribution Agreement shall be construed to create any right, or accelerate any entitlement, to any compensation or benefit whatsoever on the part of any Employee. Without limiting the generality of the foregoing, except as may be explicitly provided in an Individual Agreement, neither the Distribution nor the transfer of employment shall cause any Employee to be deemed to have incurred a termination of

employment or create any entitlement to any severance benefits or the commencement of any other benefits under any Ikaria Plan or any Individual Agreement.

(b) No Right to Continued Employment. Nothing contained in this Agreement shall confer any right to continued employment on any Employee. Except as specifically provided otherwise herein, this Agreement shall not limit the ability of either Ikaria or R&DCo, as applicable, to change the position, compensation or benefits of any of its or its subsidiary's Employees for a performance-related, business or any other reason or require any such entity to continue the employment of any such Employee for any particular period of time; provided that R&DCo shall bear all Liability associated with any such termination of employment or modification of terms and conditions of employment with respect to any R&DCo Employee and former Employees who primarily performed services for the R&DCo Business; and provided further that R&DCo covenants that its intent is to cause ServicesCo to continue to employ all of the R&DCo Employees pursuant to substantially similar terms and conditions as they are employed by Ikaria as of the date hereof and with such terms and conditions that are required under applicable law.

Section 3.4. Reimbursement.

(a) By R&DCo. From time to time after the Distribution, R&DCo shall reimburse Ikaria promptly, and in no event more than fifteen business days after delivery by Ikaria of an invoice therefore containing reasonable substantiating documentation of such costs and expenses, for the cost of any obligations or Liabilities that Ikaria is required to pay or otherwise satisfy, that are, or that pursuant to this Agreement have become, the responsibility of R&DCo.

(b) By Ikaria. From time to time after the Distribution, Ikaria shall reimburse R&DCo promptly, and in no event more than fifteen business days after delivery by R&DCo of an invoice therefore containing reasonable substantiating documentation of such costs and expenses, for the cost of any obligations or Liabilities that R&DCo is required to pay or otherwise satisfy, that are, or that pursuant to this Agreement have become, the responsibility of Ikaria.

ARTICLE IV.

R&DCO PLANS

Section 4.1. Establishment of R&DCo Plans. ServicesCo shall adopt, effective as of the Distribution Date or as soon as practicable thereafter, employee benefit plans that shall substantially replicate, to the extent commercially feasible, the following Ikaria Plans: a 401(k) plan, a medical and dental plan, Long-Term Disability, Short-Term Disability, Life and Accidental Death and Dismemberment, and Flexible Spending

Accounts (the "Replacement Plans"). ServicesCo shall become the plan sponsor of, and, from and after the date of adoption of each Plan, shall have sole responsibility for each Replacement Plan.

Section 4.2. Terms of Participation by R&DCo Employees.

(a) Right and Entitlements. Except as otherwise agreed to by the Parties in accordance with Section 2.2 and the Transition Services Agreement, each R&DCo Employee shall terminate participation in the Ikaria Plans on the Distribution Date (or Transfer Date, as the case may be). Subject to the express terms and conditions of this Agreement, each of the Replacement Plans shall be, with respect to R&DCo Employees who are participants in such Plans, the successors in interest to and shall recognize rights and entitlements under the corresponding Ikaria Plans in effect as of the Distribution Date in which such R&DCo Employees participated prior to the Distribution Date (or Transfer Date, as the case may be). The Parties agree that R&DCo Employees are not entitled to receive duplicative benefits for the same periods of service from the Ikaria Plans and the R&DCo Plans.

(b) Service and Other Factors Determining Benefits. With respect to R&DCo Employees, each R&DCo Plan shall provide that all service, all compensation, and all other factors affecting benefit determinations that, as of the Distribution Date or Transfer Date, were recognized under the corresponding Ikaria Plan (for periods immediately before the Distribution Date or Transfer Date, as applicable) shall receive full recognition, credit, and validity and be taken into account under such R&DCo Plan to the same extent that such service, compensation and other factors were taken into account under the corresponding Ikaria Plan, as though arising under such R&DCo Plan (or in the case of an R&DCo Plan that is not a Replacement Plan as if such individual had been employed by ServicesCo since his or her date of hire with any member of the Ikaria Group). Notwithstanding the immediately preceding sentence, in no event shall the crediting of service or any other action taken pursuant to the immediately preceding sentence result in the duplication of benefits for any Employee under any Ikaria Plan and any R&DCo Plan.

(c) Power to Amend. Notwithstanding the foregoing provisions of this Section 4.2 and except as specifically set forth herein, nothing in this Agreement shall preclude R&DCo from amending, merging, modifying, terminating, eliminating, reducing, or otherwise altering in any respect any R&DCo Plan, any benefit under any Plan or any trust, insurance policy or funding vehicle related to any R&DCo Plan.

ARTICLE V.

HEALTH AND WELFARE

Section 5.1. Assumption of Health and Welfare Plans.

(a) Cessation of Coverage in Ikaria Plans. Ikaria maintains health and welfare plans for the benefit of eligible Employees (the "Ikaria Welfare Plans"). On the Distribution Date (or Transfer Date, as applicable) or such later date as may be agreed to by the Parties pursuant to Section 2.2 hereof or the Transition Services Agreement, each person who is an R&DCo Employee on such date shall cease to be covered under the Ikaria Welfare Plans. Notwithstanding the immediately preceding sentence, with respect to any R&DCo Employee who would not be eligible for coverage under any R&DCo Plan because such person is not actively at work on the Distribution Date (or Transfer Date) due to any medical, sickness, accident leave or any other approved leave of absence, Ikaria shall

administer claims for such persons on behalf of R&DCo as though the relevant Ikaria Welfare Plan had continued to be applicable (at the cost and expense of R&DCo) until the earliest to occur of (i) the date such person first resumes active employment with ServicesCo; (ii) the date such person ceases to be an Employee of ServicesCo (including, without limitation, by reason of not returning to work at the expiration of any approved leave); (iii) the date on which such approved leave of absence expires without such person returning to active employment or terminating employment; and (iv) the date, if any, more than six months following the Distribution Date (or Transfer Date) that Ikaria shall choose, in its discretion, to cease to provide such continued coverage upon not less than 90 days advance written notice to R&DCo and the affected R&DCo Employees.

(b) Claims Arising Prior to Distribution Date. On or after the Distribution Date, but subject to the obligations of R&DCo pursuant to Section 2.2, Ikaria and the Ikaria Welfare Plans shall remain responsible for all Liabilities in respect of or relating to R&DCo Employees relating to claims or expenses incurred on or prior to the Distribution Date (or Transfer Date, as applicable). For purposes of the foregoing sentence, to the extent that an eligible beneficiary under any such Ikaria Welfare Plan commences a hospital confinement on or prior to the Distribution Date (or Transfer Date) that continues after the Distribution Date (or Transfer Date), to the extent consistent with the applicable insurance policy under the Ikaria Welfare Plan, all expenses related to such hospitalization (including any related services that are incurred during the period of the same continuous hospital confinement) shall be considered part of the claim incurred on or prior to the Distribution Date (or Transfer Date). All other claims shall be deemed incurred on the date the actual expense is incurred.

(c) No Transfer of Assets Pertaining to Welfare Plans. Nothing in this Agreement shall require Ikaria or any Ikaria Welfare Plan to transfer assets or reserves with respect to the Ikaria Welfare Plans to R&DCo.

Section 5.2. Terms of Participation in the R&DCo Welfare Plans. R&DCo shall cause the applicable R&DCo Plans to (a) waive all limitations as to preexisting

conditions, exclusions, service conditions and waiting period limitations, and any evidence of insurability requirements applicable to any such R&DCo Employees other than such limitations, exclusions, and conditions that were in effect with respect to R&DCo Employees as of the Distribution Date (or Transfer Date), in each case under the corresponding Ikaria Welfare Plan and (b) honor any deductibles, out-of-pocket maximums and co-payments incurred by R&DCo Employees under the corresponding Ikaria Welfare Plan in satisfying the applicable deductibles, out-of-pocket expenses or co-payments under such Ikaria Welfare Plan for the calendar year in which the Distribution Date (or Transfer Date) occurs.

Section 5.3. COBRA. R&DCo shall be responsible for liability associated with the continuation coverage requirements for "group health plans" under Title X of the Consolidated Omnibus Budget Reconciliation Act of 1985, as amended ("COBRA"), with respect to any R&DCo Employee and any of his or her dependents having rights derived from such R&DCo Employee with respect to qualified events incurred during any period commencing immediately following Distribution Date (or Transfer Date, as applicable). Ikaria shall be responsible for liability associated with COBRA with respect to any Employee and any of his or her dependents having rights derived from such Employee where such individual has incurred an initial qualifying event prior to or through the Distribution Date (or Transfer Date), including any qualifying event incurred in connection with the transactions contemplated by this Agreement or the Separation and Distribution Agreement.

Section 5.4. Workers' Compensation Claims. Effective on the Distribution Date, R&DCo shall assume responsibility for all Liabilities for R&DCo Employees and any former Employees who primarily performed services for the R&DCo Business related to any and all workers' compensation claims and coverage, whether arising under any law of any state, territory, or possession of the U.S. or the District of Columbia and whether arising before or after the Distribution Date. For the avoidance of doubt, Ikaria shall be fully responsible for the administration of all such claims made prior to Distribution Date, but R&DCo shall reimburse and otherwise fully indemnify Ikaria for all Liabilities related to such claims in respect of such R&DCo Employees and any former Employees who primarily performed services for the R&DCo Business, including (a) the costs of administering the plans, programs or arrangements under which any such Liabilities have accrued or otherwise arisen, (b) paying benefits and settlements and (c) establishing reserves, in each case as determined by Ikaria or its designate. With respect to any claim for worker's compensation or similar benefits by an R&DCo Employee or any former Employees who primarily performed services for the R&DCo Business made after the Distribution Date, R&DCo shall be solely responsible for such claim and for complying with all applicable laws with respect thereto.

Section 5.5. Flexible Spending Accounts. As soon as reasonably practicable following the date that ServicesCo adopts a flexible spending account Replacement Plan,

Ikaria will transfer the R&DCo Employees' health-care and dependent-care spending account balances (determined as of such date (the "Flex Plan Transfer Date")) under Ikaria's flexible spending account plans to ServicesCo's flexible spending account plan and the net aggregate flexible spending account balances for R&DCo Employees, positive or negative, will be offset from the next request for reimbursement by the Parties under this Agreement. On and after the Flex Plan Transfer Date, ServicesCo's flexible spending account plan shall be responsible for reimbursement of eligible health-care and dependent-care expenses incurred during the 2014 calendar year by R&DCo Employees and their respective eligible spouses and dependents (to the extent such expenses have no previously been reimbursed under Ikaria's flexible spending account plan).

ARTICLE VI.

401(K) PLANS

Section 6.1. Establishment of the R&DCo 401(k) Plan. R&DCo shall be responsible for taking or causing to be taken all necessary, reasonable, and appropriate action to establish, maintain, and administer a Replacement Plan intended to qualify under Section 401(a) of the Code having a cash or deferred feature qualified under Section 401(k) of the Code (the "R&DCo 401(k) Plan") as soon as reasonably practicable following the Distribution Date. For the avoidance of doubt, nothing in this Section 6.1 shall be construed to require R&DCo to maintain any particular investment option under such plan or to provide a company match, or a company match at the same level as in the Ikaria 401(k) Plan.

Section 6.2. Plan-to-Plan Transfer; Rollover. The Parties shall cooperate in effecting a plan-to-plan transfer of assets and liabilities from the Ikaria 401(k) Plan to the R&DCo 401(k) Plan. In the event the Parties determine jointly that such a transfer is not practicable, the R&DCo 401(k) Plan shall accept direct rollovers of the R&DCo Employees' balances in the Ikaria 401(k) Plan, including loan rollovers to the extent administratively feasible.

Section 6.3. Employer Contributions. To the extent that Ikaria contributes additional employer contributions following the Distribution Date (or Transfer Date, as applicable) to the Ikaria 401(k) Plan on behalf of R&DCo Employees or former Employees who primarily performed services for the R&DCo Business, R&DCo shall promptly reimburse Ikaria for the cost of such employer contributions that are allocated to such Employees' accounts.

ARTICLE VII.

EQUITY BASED AND OTHER LONG-TERM INCENTIVE AWARDS

Section 7.1. Assumption of Equity Incentive Plans; General Treatment of Outstanding Awards.

(a) Treatment of Outstanding Ikaria Stock Options. Prior to and in connection with the Distribution, each outstanding Ikaria Stock Option, whether vested or unvested, and each Ikaria Equity Plan itself, insofar as it relates to outstanding Ikaria Stock Options, shall be adjusted so that each Ikaria Stock Option shall become (i) an option to acquire, on the same terms and conditions as were applicable to such Ikaria Stock Option prior to the Distribution, an option to acquire the same number of shares of Ikaria non-voting common stock as were subject to the Ikaria Stock Option, at a price per share (rounded up to the nearest whole cent) equal to the product of (x) the quotient of (A) the CommercialCo FMV and (B) the sum of the (I) CommercialCo FMV and (II) the R&DCo FMV and (y) the exercise price per share of such Ikaria Stock Option (such options, the "Post-Distribution Ikaria Options"); and (ii) an option to acquire, on the same terms and conditions as were applicable to such Ikaria Stock Option prior to the Distribution, an option to acquire the same number of R&DCo non-voting common units as were subject to the Ikaria Stock Option, at a price per unit (rounded up to the nearest whole cent) equal to the product of (x) the quotient of (A) the R&DCo FMV and (B) the sum of (I) the CommercialCo FMV and (II) the R&DCo FMV and (y) the exercise price per share of such Ikaria Stock Option (such options the "Post-Distribution R&DCo Options"); provided, however, that Ikaria Stock Options held by holders who are not Accredited Investors or employees of Ikaria will also be adjusted pursuant to this Section 7.1(a), but the holders thereof will receive, in lieu of each such Post-Distribution R&DCo Option, an amount in cash equal to the product of (1) the difference between the fair market value of an R&DCo non-voting common unit (as determined by R&DCo) less the exercise price of the Post-Distribution R&DCo Option such holder would have been granted, multiplied by (2) the number of R&DCo non-voting common units that would have been subject to such R&DCo Option, less applicable withholding taxes, which cash amount R&DCo shall pay (or cause to be paid) to the holder promptly following completion of the Distribution. Solely to the extent that Ikaria accelerates in full the vesting of the Post-Distribution Ikaria Options in connection with the Ikaria Acquisition, then R&DCo hereby undertakes to accelerate in full the vesting of the Post-Distribution R&DCo Options.

(b) Treatment of Outstanding Ikaria RSUs. Prior to and in connection with the Distribution, each outstanding Ikaria RSU, and each Ikaria Equity Plan itself, insofar as it relates to outstanding Ikaria RSUs, shall be adjusted so that each Ikaria RSU shall become (i) a restricted stock unit, on the same terms and conditions as were applicable to such Ikaria RSU prior to the Distribution, with respect to the same number of shares of Ikaria non-voting common stock as were subject to the Ikaria RSU (such restricted stock units, the "Post-Distribution Ikaria RSUs"); and (ii) a restricted unit, on the same terms and conditions as were applicable to such Ikaria RSU prior to the Distribution, with respect to the same number of R&DCo non-voting common units as were subject to the

Ikaria RSU (such restricted units, the "Post-Distribution R&DCo RSUs"). Solely to the extent that Ikaria accelerates in full the vesting of the Post-Distribution Ikaria RSUs in connection with the Ikaria Acquisition, then R&DCo hereby undertakes to accelerate in full the vesting of the Post-Distribution R&DCo RSUs and to settle such Post-Distribution R&DCo RSUs by the delivery of one R&DCo non-voting common unit with respect to each such Post-Distribution R&DCo RSU; provided, however, that a portion of the Post-Distribution R&DCo RSUs may be accelerated prior to the closing of the Ikaria Acquisition in the discretion of the Board of Directors of R&DCo.

(c) Ikaria Equity Award Actions. Ikaria shall continue the Ikaria Equity Plans to the extent necessary to govern the Ikaria Equity Awards following the adjustment contemplated by this Section and shall continue to take all corporate action necessary to reserve a sufficient number of shares of Ikaria non-voting common stock for delivery upon exercise of the Post-Distribution Ikaria Options issued pursuant to this Section and the settlement of the Post-Distribution Ikaria RSUs issued pursuant to this Section; provided, however, that (i) nothing herein shall limit Ikaria's rights with respect to the disposition of an Ikaria Equity Award in accordance with the Ikaria Acquisition and (ii) the provisions of this Section 7.1(c) shall cease to apply following the closing of the Ikaria Acquisition.

(d) R&DCo Equity Award Actions. R&DCo shall assume each Ikaria Equity Plan solely to the extent necessary to govern the R&DCo Equity Awards and such plans shall be renamed as R&DCo Equity Plans. R&DCo shall take all corporate action necessary to reserve for issuance a sufficient number of R&DCo non-voting common units for delivery upon exercise of the Post-Distribution R&DCo Options assumed in accordance with this Section and the settlement of the Post-Distribution R&DCo RSUs assumed in accordance with this Section. For the avoidance of doubt and without limitation of Section 2.1(i), R&DCo shall be responsible for all Liabilities with respect to the R&DCo Equity Awards that remain outstanding following the Distribution, including (a) all income, payroll, or other tax reporting related to income of R&DCo Employees and Ikaria Employees from any R&DCo Equity Awards and (b) remitting the applicable tax withholdings for such income to each applicable taxing authority. R&DCo shall provide written notice to Ikaria in the event that any Ikaria Employee exercises an R&DCo Equity Award, which notice shall be provided within thirty (30) days following R&DCo's receipt of notice of such exercise.

ARTICLE VIII.

INCENTIVE PLANS

Section 8.1. Incentive Plans. R&DCo shall be responsible for all Liabilities relating to R&DCo Employees in respect of any short term incentive plan related to their services for the R&DCo Business, including that, with respect to any Transferred

Employee (or Delayed Transfer Employee or any former Employee who primarily performed services for the R&DCo Business), R&DCo shall be responsible for, or shall reimburse Ikaria for, the payment of any portion of any incentive payment payable to any Transferred Employee (or Delayed Transfer Employee or any former Employee who primarily performed services for the R&DCo Business) related to service performed in the applicable performance period through the Distribution Date (or, if applicable, the Transfer Date).

ARTICLE IX.

EXECUTIVE AGREEMENTS

[Reserved.]

ARTICLE X.

GENERAL AND ADMINISTRATIVE

Section 10.1. Sharing of Information. Subject to any consents required or any other restrictions imposed by law, each Party shall each provide to any other Party and its agents and vendors all information that such other Party may reasonably request to enable the requesting party to administer efficiently and accurately each of its Plans and to determine the scope of, and to fulfill, its obligations under this Agreement. Ikaria shall provide R&DCo or its designees, on a timely basis, such information including, without limitation, dates of termination, length of service and last known addresses, and other assistance as it or they shall reasonably request from time to time to administer its on-going obligations under this Agreement. Any information shared or exchanged pursuant to this Agreement shall be kept confidential by the Parties and used only for and to the extent necessary to establish, maintain and administer the plans, programs and agreements as contemplated by this Agreement.

Section 10.2. Cooperation. Each of the Parties hereto will use its commercially reasonable efforts to promptly take, or cause to be taken, any and all actions and to do, or cause to be done, any and all things necessary, proper and advisable (including, without limitation, any actions required under applicable laws and regulations) to fulfill their respective duties and obligations contemplated by this Agreement. The actions described in the immediately preceding sentence shall include, without limitation, adopting plans or plan amendments and the payment of compensation due to any Employee, Ikaria Employee or R&DCo Employee. Each of the Parties hereto shall cooperate fully on any issue relating to the duties and obligations contemplated by this Agreement for which the

other Party seeks a determination letter or any other filing, consent, or approval with respect to governmental authorities.

Section 10.3. Consent of Third Parties. If any provision of this Agreement is dependent on the consent of any third party (such as a vendor or insurer) and such consent is withheld, the Parties shall use their reasonable best efforts to implement the applicable provisions of this Agreement to the full extent practicable. If any provision of this Agreement cannot be implemented due to the failure of such third party to consent, the Parties shall negotiate in good faith to implement the provision in a mutually satisfactory manner.

Section 10.4. No Third Party Beneficiaries.

(a) Except as provided in Section 10.4(d), nothing in this Agreement shall confer upon any person (or any beneficiary thereof) any rights under or with respect to any plan, program or arrangement described in or contemplated by this Agreement and each person (and any beneficiary thereof) shall be entitled to look only to the express terms of any such plan, program or arrangement for his or her rights thereunder.

(b) Nothing in this Agreement shall create any right of any Person to object or to refuse to assent to R&DCo's assumption of, succession to or creation of any Individual Agreement, or other agreement or plan, program or arrangement relating to employment, employment separation, severance or employee benefits, nor shall this Agreement be construed as recognizing that any such rights exist.

(c) Nothing in this Agreement shall amend or shall be construed to amend any plan, program or arrangement described in or contemplated by this Agreement or to alter or limit R&DCo's or any member of the Ikaria Group's ability to amend, modify or terminate any particular benefit plan, program or agreement.

(d) Except for Purchaser, which shall be a third party beneficiary of this Agreement, nothing in this Agreement is intended, or shall be deemed, to confer any rights or remedies upon any Person other than the Parties and their respective Group Members, successors and permitted assigns, to create any agreement of employment with any Person or to otherwise create any third-party beneficiary hereto or thereto. This Agreement may only be amended with Purchaser's consent (which shall not be unreasonably withheld).

Section 10.5. Notices. All notices and other communications under this Agreement shall be in writing and shall be deemed duly delivered (a) four Business Days after being sent by registered or certified mail, return receipt requested, postage prepaid, (b) one Business Day after being sent for next Business Day delivery, fees prepaid, via a reputable nationwide overnight courier service, or (c) on the date of confirmation of

receipt (or, the first Business Day following such receipt if the date of such receipt is not a Business Day) of transmission by facsimile, in each case to the intended recipient as set forth below.

If to Ikaria:

INO Therapeutics LLC
Perryville III Corporate Park
53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel

If to R&DCo:

Bellerophon Therapeutics LLC
Perryville III Corporate Park

53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel (or Chief Executive Officer if there is no General Counsel)

Either Party may give any notice or other communication hereunder using any other means (including personal delivery, messenger service, ordinary mail or electronic mail), but no such notice or other communication shall be deemed to have been duly given unless and until it actually is received by the Party for whom it is intended. Either Party may change the address to which notices and other communications hereunder are to be delivered by giving the other Party notice in the manner herein set forth.

Section 10.6. Governing Law. The internal Laws of the State of Delaware (without giving effect to any choice or conflict of law provision or rule, whether of the State of Delaware or any other jurisdiction, that would cause the application of Laws of any jurisdiction other than those of the State of Delaware) shall govern the construction, interpretation and other matters arising out of or in connection with this Agreement and each of the exhibits and schedules hereto and thereto (whether arising in contract, tort, equity or otherwise).

Section 10.7. Jurisdiction. If any dispute, controversy or claim arises out of or in connection with this Agreement, the Parties irrevocably (a) consent and submit to the exclusive jurisdiction of the Court of Chancery of the State of Delaware, New Castle County, or, if that court does not have jurisdiction, a federal court sitting in Wilmington,

Delaware, (b) waive any objection to that choice of forum based on venue or to the effect that the forum is not convenient, and (c) WAIVE TO THE FULLEST EXTENT PERMITTED BY LAW ANY RIGHT TO TRIAL OR ADJUDICATION BY JURY. Either Party may make service on the other Party by sending or delivering a copy of the process to the other Party at the address and in the manner provided for the giving of notices in Section 10.5. Nothing in this Section 10.7, however, shall affect the right to serve legal process in any other manner permitted by Law.

Section 10.8 Binding Effect and Assignment. This Agreement binds and benefits the Parties and their respective permitted successors and assigns. Neither Party may assign any of its rights or delegate any of its obligations under this Agreement without the written consent of the other Party and any assignment or attempted assignment in violation of the foregoing shall be null and void. Notwithstanding the preceding sentence, either Party may, upon written notice, assign this Agreement in connection with a merger transaction in which such Party is not the surviving entity or the sale of all or substantially all of its assets; provided that the surviving party or acquirer in such transaction agrees in writing to assume and be bound by all of such Party's obligations hereunder.

Section 10.9 Severability. Any term or provision of this Agreement that is invalid or unenforceable in any situation in any jurisdiction shall not affect the validity or enforceability of the remaining terms and provisions hereof or thereof or the validity or enforceability of the offending term or provision in any other situation or in any other jurisdiction. If the final judgment of a court of competent jurisdiction declares that any term or provision hereof is invalid or unenforceable, the Parties agree that the court making such determination shall have the power to limit the term or provision, to delete specific words or phrases, or to replace any invalid or unenforceable term or provision with a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision, and this Agreement shall be enforceable as so modified. In the event such court does not exercise the power granted to it in the prior sentence, the Parties agree to replace such invalid or unenforceable term or provision with a valid and enforceable term or provision that shall achieve, to the extent possible, the economic, business and other purposes of such invalid or unenforceable term.

10.10 Specific Performance. The Parties agree that irreparable damage would occur in the event that any provision of this Agreement were not performed in accordance with its specific terms or were otherwise breached. It is accordingly agreed that either Party shall be entitled to an injunction or injunctions to prevent breaches of this Agreement and to enforce specifically the terms and provisions of this Agreement in each case without posting a bond or undertaking, this being in addition to any other remedy to which they are entitled at law or in equity. Each of the Parties agrees that it shall not oppose the granting of an injunction, specific performance and other equitable relief on

the basis that (a) the Party seeking such remedy has an adequate remedy at law or (b) an award of specific performance is not an appropriate remedy for any reason at law or equity.

Section 10.11 Entire Agreement. This Agreement, together with the Separation and Distribution Agreement and the other Ancillary Documents and each of the exhibits and schedules appended hereto and thereto, constitutes the final agreement between the Parties, and is the complete and exclusive statement of the Parties' agreement on the matters contained herein and therein. All prior and contemporaneous negotiations and agreements between the Parties with respect to the matters contained herein and therein are superseded by this Agreement, the Separation and Distribution Agreement and the other Ancillary Documents, as applicable. In the event of any conflict between (a) any provision in this Agreement, on the one hand, and (b) any specific provision in the Separation and Distribution Agreement, on the other hand, pertaining to the subject matter of this Agreement, the specific provisions in this Agreement shall control over the provisions in the Separation and Distribution Agreement, as applicable.

Section 10.12 Amendment. The Parties may amend this Agreement only by a written agreement signed by both Parties and that identifies itself as an amendment to this Agreement.

Section 10.13 Termination. This Agreement may be terminated (a) at any time after the Distribution, by the mutual written consent of Ikaria and R&DCo; or (b) at any time prior to the Distribution by (and in the sole discretion of) Ikaria without the approval of R&DCo. In the event of a termination of this Agreement pursuant to the foregoing sentence, neither Party shall have any liability of any kind to the other Party under this Agreement, except for any breach of this Agreement that occurs prior to such termination.

[*Signature Pages Follow*]

IN WITNESS WHEREOF, each of the Parties has caused this Agreement to be executed on its behalf by a duly authorized officer on the date first set forth above.

IKARIA, INC.
a Delaware corporation

By: /s/ James Briggs
Name: James Briggs
Title: Senior Vice President,
Human Resources

BELLEROPHON THERAPEUTICS LLC
a Delaware limited liability company

By: /s/ Daniel Tassé
Name: Daniel Tassé
Title: Chief Executive Officer

[Signature Page to Employee Matters Agreement]

SCHEDULE A TO EMPLOYEE MATTERS AGREEMENT
TRANSFERRED EMPLOYEES

Confidential Materials omitted and filed separately with the Securities and Exchange Commission. A total of two pages were omitted. [**]

Exhibit 10.14

Execution Version

Confidential Materials omitted and filed separately with the
Securities and Exchange Commission. Double asterisks denote omissions.

**EXCLUSIVE CROSS-LICENSE, TECHNOLOGY TRANSFER,
AND REGULATORY MATTERS AGREEMENT**

THIS EXCLUSIVE CROSS-LICENSE, TECHNOLOGY TRANSFER, AND REGULATORY MATTERS AGREEMENT (the **"Agreement"**), is dated February 9, 2014 (the **"Effective Date"**) by and between INO Therapeutics LLC, a Delaware limited liability company, d/b/a Ikaria, with offices at Perryville III Corporate Park, 53 Frontage Road, Third Floor, Hampton, NJ 08827 (**"Ikaria"**), and Bellerophon Pulse Technologies LLC, a Delaware limited liability company, with offices at Perryville III Corporate Park, 53 Frontage Road, Third Floor, Hampton, NJ 08827 (**"Pulse Technologies"**). Ikaria and Pulse Technologies may be individually referred to as a **"Party"** and together as the **"Parties**.**"**

WHEREAS, immediately prior to the Effective Date, Ikaria and Pulse Technologies were indirect subsidiaries of Ikaria, Inc., a Delaware corporation (**"Ikaria Parent Company"**);

WHEREAS, Ikaria was, prior to the Effective Date, engaged in the Ikaria NO Business (as defined herein) and the R&D Business (as defined herein);

WHEREAS, Pulse Technologies has been organized to pursue the development and commercialization of products and services for the R&D Business;

WHEREAS, pursuant to that certain Separation and Distribution Agreement between Ikaria, Inc. and Bellerophon Therapeutics LLC (the **"Separation Agreement"**), dated as of the Effective Date, the parties to the Separation Agreement will effectuate spin-out transactions pursuant to which Pulse Technologies will cease to be a direct or indirect subsidiary of Ikaria Parent Company (the **"Spin-Out"**); and

WHEREAS, in connection with the Spin-Out, the Parties desire to grant one another the rights and licenses set forth in this Agreement, subject to and in accordance with the terms and conditions of this Agreement.

NOW, THEREFORE, in consideration of the mutual covenants contained herein, and for other good and valuable consideration, the receipt and sufficiency of which are hereby acknowledged, the Parties hereto agree as follows:

1. **Definitions and Interpretation.**

1.1. The foregoing preamble and Annexes hereto form an integral part of this Agreement.

1.2. In this Agreement the terms below shall bear the respective meanings assigned to them below and other capitalized terms shall bear the respective meanings assigned to them in their parenthetical definition, unless specifically stated otherwise:

1.2.1. **"Affiliate"** shall mean, with respect to a Party, any Person directly or indirectly controlling, controlled by or under common control with, such Party (in each case whether now in existence or later formed, acquired,

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merged with or into, or otherwise, and regardless of the form of legal entity). For purposes of this definition only, "control" of a Person shall mean the ability, directly or indirectly, to direct the activities of the relevant Person, and with respect to corporate entities shall mean (a) ownership or direct control of fifty percent (50%) or more of the outstanding voting stock or other ownership interest of such Person, or (b) direct or indirect possession, of the power to elect or appoint fifty percent (50%) or more of the members of the governing body of such Person. Notwithstanding the foregoing or any direct or indirect control relationship that exists between them, Ikaria and Pulse Technologies shall be deemed not to be Affiliates of one another.

1.2.2. **"Agreement"** shall have the meaning set forth in the Preamble.

1.2.3. **"Bankruptcy Code"** shall mean United States Bankruptcy Code (Title 11, U.S. Code), as amended.

1.2.4. **"Confidential Information"** shall mean information disclosed by, or on behalf of, a Party or its Affiliate (such Party referred to as the **"Disclosing Party"**) to the other Party or its Affiliate (such Party referred to as the **"Receiving Party"**), which information relates either directly or indirectly to the business or activities of the Disclosing Party, including information and data regarding the composition, formulation, manufacture or use, pre-clinical or clinical data regarding, the status of research or development of, or information regarding Patent Rights relating to, any R&D Product or Grant-Back Field Product.

Confidential Information of the Disclosing Party excludes any information that the Receiving Party can establish by written records:

(a) was known by the Receiving Party prior to receipt from the Disclosing Party;

(b) was disclosed to the Receiving Party by a Third Party having the right to do so without an obligation of confidentiality to the Disclosing Party; or

(c) was, or subsequently became, publicly known through no fault of the Receiving Party or its Affiliates.

1.2.5. **"Control"** shall mean, with respect to any IP of a Party possession of the right to grant the licenses or sublicenses as provided herein without violating the terms of any agreement or other arrangement with any Third Party. Notwithstanding the foregoing, IP of a Party that is licensed or otherwise acquired from a Third Party and would otherwise be considered to be under the Control of a Party shall not be deemed to be under the Control of such Party if the application of such definition in the context of any license grants or sublicenses under this Agreement

would require the granting Party to make additional payments or royalties to a Third Party in connection with such license or sublicense grants, unless the other Party agrees to pay the additional payments or royalties to the Third Party.

1.2.6. **"COPD"** shall mean pulmonary hypertension secondary to chronic obstructive pulmonary disease.

1.2.7. "**Copyright**" shall mean copyrights and copyrightable subject matter and all applications and registrations therefor.

1.2.8. **"Core R&D Patents"** shall mean (a) the following Patents: [**], (b) any substitutions, divisionals, continuations, continuations-in-part, provisional applications, reissues, renewals, registrations, confirmations, re-examinations, extensions, supplementary protection certificates and the like of any such patents or patent applications, and (c) foreign counterparts of any of the foregoing.

1.2.9. **"Disclosing Party"** shall have the meaning set forth in Section 1.2.4.

1.2.10. **"Effective Date"** shall have the meaning set forth in the preamble to this Agreement.

1.2.11. **"Grant-Back IP"** shall mean all the IP rights Controlled by Pulse Technologies either as of the Effective Date or as of any time thereafter during the Term that are or may be necessary or useful to engage in the Ikaria NO Business; provided, however, that (a) if Pulse Technologies is acquired by any Third Party (whether by merger, stock sale, asset sale, or otherwise) after the Effective Date, the Grant-Back IP shall not include any IP of the acquiror or its Affiliates; and (b) if Pulse Technologies acquires any Third Party (whether by merger, stock sale, asset sale, or otherwise) after the Effective Date, the Grant-Back IP shall not include any IP rights Controlled by the acquired party.

1.2.12. **"Grant-Back Field Products"** shall mean any product or service offered to a customer as part of the Ikaria NO Business (either as of the Effective Date or at any time thereafter) and that in anyway uses, incorporates, reflects, is based upon, or relies upon any Grant-Back IP.

1.2.13. **"Grant-Back Patent Rights"** shall mean Patent Rights included in the Grant-Back IP, including the R&DCo Intellectual Property as defined in the Separation Agreement.

1.2.14. **"HF Patents"** shall mean the Patents Rights claiming priority to [**].

1.2.15. **"Ikaria"** shall have the meaning set forth in the preamble to this Agreement.

1.2.16. **"Ikaria Competitor"** shall mean any Affiliate of Pulse Technologies or any Third Party that is in any way anywhere in the world, directly or indirectly, engaged in the Ikaria NO Business.

1.2.17. **"Ikaria Content"** shall have the meaning set forth in Section 2.6.

1.2.18. **"Ikaria Licensed Items"** shall have the meaning set forth in Section 2.6.

1.2.19. **"Ikaria Marks"** shall have the meaning set forth in Section 2.6.

1.2.20. **"Ikaria NO Business"** shall mean the business of the development, manufacture, commercialization, promotion, sale, import, export, servicing, repair, training, storage, distribution, transportation, licensing, or other handling or disposition of any product or service (including any product or service that utilizes, contains, or includes nitric oxide for inhalation, a device intended to deliver nitric oxide, or a service that delivers or supports the delivery of nitric oxide), bundled or unbundled, for or used in connection with (a) the diagnosis, prevention, or treatment, in both adult and/or pediatric populations, and whether in or out patient, of: (i) hypoxic respiratory failure associated with pulmonary hypertension, (ii) pulmonary hypertensive episodes and right heart failure associated with cardiovascular surgery, (iii) bronchopulmonary dysplasia, (iv) the management of ventilation — perfusion mismatch in acute lung injury, (v) the management of ventilation — perfusion mismatch in acute respiratory distress syndrome, (vi) the management of pulmonary hypertension episodes and right heart failure in congestive heart failure, (vii) pulmonary edema from high altitude sickness, (viii) the management of pulmonary hypertension episodes and right heart failure in pulmonary or cardiac surgery, (ix) the management of pulmonary hypertension episodes and right heart failure in organ transplant, (x) sickle cell vaso-occlusive crisis, (xi) hypoxia associated with pneumonia, or (xii) ischemia-reperfusion injury; or (b) the use of nitric oxide to treat or prevent conditions that are primarily managed in the hospital.

1.2.21. **"Ikaria Parent Company"** shall have the meaning set forth in the recitals to this Agreement.

1.2.22. **"IP"** shall mean Patent Rights, Copyrights and Know-How.

1.2.23. **"IPF"** shall mean pulmonary hypertension secondary to idiopathic pulmonary fibrosis.

1.2.24. **"Know-How"** shall mean any and all trade secrets, confidential data and technical information, including practices, techniques, methods, processes, inventions, developments, specifications, formulations, manufacturing processes, structures, chemical or biological

manufacturing control data, analytical and quality control information and procedures, pharmacological, toxicological and clinical test data and results, stability data, studies and procedures and regulatory information.

1.2.25. **"PAH"** shall mean primary or idiopathic pulmonary arterial hypertension.

1.2.26. **"Party"** or **"Parties"** shall have the meaning set forth in the preamble to this Agreement.

1.2.27. **"Patent Rights"** shall mean (a) all patents (including design patents) and patent applications in any country or supranational jurisdiction in the Territory, (b) any substitutions, divisionals, continuations, continuations-in-part, provisional applications, reissues, renewals, registrations, confirmations, re-examinations, extensions, supplementary protection certificates and the like of any such patents or patent applications, and (c) foreign counterparts of any of the foregoing.

1.2.28. **"Person"** shall mean any individual, partnership, joint venture, limited liability company, corporation, firm, trust, association, unincorporated organization, governmental authority or agency, or any other entity not specifically listed herein.

1.2.29. **"R&D Business"** shall mean the business of the development, manufacture, commercialization, promotion, sale, import, export, servicing, repair, training, storage, distribution, transportation, licensing, or other handling or disposition of (a) nitric oxide, (b) a device intended to deliver nitric oxide, or (c) a service that delivers or supports the delivery of nitric oxide; in each case, solely for or in connection with the outpatient, chronic treatment of patients who have COPD, IPF or PAH and even if initiation of therapy occurs in a hospital setting or such treatment occurs as part of episodic treatment or hospitalization of patients with COPD, IPF or PAH.

1.2.30. **"R&D NO Delivery Devices"** shall mean versions (including any existing designs and prototypes) of pulse administering nitric oxide devices (including the "INOpulse" device) for the treatment of COPD and PAH.

1.2.31. **"R&D IP"** shall mean all the IP rights Controlled by Ikaria either as of the Effective Date or as of any time thereafter that are or may be necessary or useful to engage in the R&D Business, including Know-How disclosed by Ikaria to Pulse Technologies pursuant to Sections 7.1.2 and 7.2, but excluding, for the avoidance of doubt, the Ikaria Licensed Items; provided, however, that (a) if Ikaria is acquired by any Third Party (whether by merger, stock sale, asset sale, or otherwise) after the Effective Date, the R&D IP shall not include any IP of the

acquiror or its Affiliates; and (b) if Ikaria acquires any Third Party (whether by merger, stock sale, asset sale, or otherwise) after the Effective Date, the R&D IP shall not include any IP rights Controlled by the acquired party.

1.2.32. **"R&D Patent Rights"** shall mean Patent Rights included in the R&D IP (which includes the Core R&D Patents).

1.2.33. **"R&D Product"** shall mean (a) any product or service offered to a customer as part of the R&D Business (either as of the Effective Date or at any time thereafter) and that in any way uses, incorporates, reflects, is based upon, or relies upon any R&D IP, or (b) any R&D NO Delivery Device.

1.2.34. **"R&D Product Approval"** shall mean any applications, approvals, clearances, or other government authorizations of any type of or for an R&D Product.

1.2.35. **"R&D Product Customer"** shall mean any end user of an R&D Product.

1.2.36. **"Receiving Party"** shall have the meaning set forth in Section 1.2.4.

1.2.37. **"Sublicense"** shall mean any right granted or license given by a Party and/or its Sublicensees to any other Person, permitting the exercise by such other Person of rights or licenses granted to such Party in Section 2.1 or 2.2, as applicable; and the term "**Sublicensee**" shall be construed accordingly.

1.2.38. **"Sublicensee"** shall have the meaning set forth in Section 1.2.37.

1.2.39. **"Term"** shall have the meaning set forth in Section 11.1.

1.2.40. **"Territory"** shall mean worldwide.

1.2.41. **"Third Party"** shall mean any Person other than Ikaria, Pulse Technologies and their respective Affiliates.

1.3. In this Agreement, words importing the singular shall include the plural and vice-versa and words importing any gender shall include all other genders.

1.4. The words "including" and "includes" mean including, without limiting the generality of any description preceding such terms.

1.5. In the event of any discrepancy between the terms of this Agreement and any of the annexes hereto, the terms of this Agreement shall prevail.

1.6. Article, section, paragraph and annex headings shall not affect the interpretation of this Agreement.

2. **License Grants.**

2.1. Subject to the terms and conditions of this Agreement, Ikaria hereby grants to Pulse Technologies and any other current or future direct subsidiaries of Pulse Technologies a sole and exclusive (even as to Ikaria, except that Ikaria retains the right to practice such IP as necessary to perform contract manufacturing services for Pulse Technologies as specified in manufacturing and/or supply agreements between the Parties), fully paid-up, non-royalty-bearing right and license, during the Term, under the R&D IP, to engage in the R&D Business in the Territory, and, subject to Section 3, to grant Sublicenses under any such rights to Third Parties or its Affiliates.

2.2. Subject to the terms and conditions of this Agreement, Pulse Technologies hereby grants to Ikaria and any other current or future direct subsidiaries of Ikaria Parent Company a sole and exclusive (even as to Pulse Technologies), fully paid-up, non-royalty-bearing, perpetual (except as otherwise set forth in Section 11.5) right and license, under the Grant-Back IP, to engage in the Ikaria NO Business in the Territory, and to grant Sublicenses under the Grant-Back IP to Third Parties or its Affiliates. Pulse Technologies shall ensure that it at all times Controls any IP invented, developed, authored, or otherwise created by it or its Affiliates or Sublicensees for or in respect of the R&D Business such that it would be "Grant-Back IP" for purposes of this Section 2.2.

2.3. Except as expressly provided in Sections 2.1 and 2.2, as between the Parties, all rights in and to the IP Controlled by each Party shall be retained by such Party.

2.4. Upon either Party's reasonable request, and at such reasonable times as may be agreed upon by the Parties, the Parties shall meet (in person or be telephone conference) to discuss and identify to the other Party any new IP Controlled by each Party that falls within the scope of the respective license grant to other Party.

2.5. In exercising the license rights granted by Ikaria to Pulse Technologies under Section 2.1, Pulse Technologies agrees that it will not, either directly or indirectly (whether through a Subsidiary, a Sublicensee, or otherwise), modify, develop, or manufacture, or commercialize (or permit to be modified, developed, manufactured, or commercialized) any long term pulse administering nitric oxide device for COPD or PAH that has any of the abilities, attributes, capabilities, capacities, functions, or specifications set forth in Exhibit A to this Agreement.

2.6. Except as set forth in this Section 2.6, Pulse Technologies acknowledges and agrees that Ikaria does not grant to Pulse Technologies any right, title, or interest, or any license rights of any type, in or to any trademark or service mark of Ikaria or its Affiliates. Notwithstanding the foregoing, and subject to the subsections of this Section 2.6, Ikaria hereby grants to Pulse Technologies a non-exclusive, non-

transferable, license to use such trademarks, logos, and other marks owned by Ikaria relating to the R&D Products as Ikaria may from time to time specify (collectively, the **"Ikaria Marks"**) and such photographs, graphics, designs, descriptions, and other works of authorship as Ikaria may from time to time specify (collectively, the **"Ikaria Content"**; together with the Ikaria Marks, the **"Ikaria Licensed Items"**) solely to the extent necessary for use in connection with clinical trials on an R&D Product ongoing as of the Effective Date in the same form and manner as the Ikaria Marks are used as of the Effective Date. Pulse Technologies agrees to use the Ikaria Licensed Items in accordance with the terms and conditions of this Agreement and with good trademark and copyright practices, including by protecting the value of the goodwill associated with the Ikaria Licensed Items. Each initial use of the Ikaria Licensed Items by Pulse Technologies shall be subject to Ikaria's prior written approval. Pulse Technologies shall not modify or create any derivative works of any Ikaria Licensed Item. In addition, all use of the Ikaria Licensed Items shall be subject to the following:

2.6.1. Pulse Technologies acknowledges the value and goodwill associated with the Ikaria Marks and agrees that the nature and quality of all uses relating to the R&D Products that use any of the Ikaria Licensed Items shall conform to standards set by Ikaria, and be under the control of, Ikaria. Pulse Technologies shall comply with any guidelines regarding the use of the Ikaria Licensed Items that Ikaria may from time to time provide to Pulse Technologies in writing. Pulse Technologies shall use appropriate trademark, Copyright, or other symbols wherever appropriate and as directed by Ikaria.

2.6.2. Pulse Technologies acknowledges that, as between the Parties, Ikaria owns all right, title, and interest in the Ikaria Licensed Items worldwide and the goodwill associated with the same. All goodwill created by Pulse Technologies' use of the Ikaria Licensed Items shall inure to the benefit of Ikaria or its applicable Affiliate. Pulse Technologies hereby assigns to Ikaria all rights it may acquire by operation of law or otherwise in the Ikaria Licensed Items, including all applications or registrations therefor, along with the goodwill associated therewith. Pulse Technologies agrees to execute and deliver to Ikaria all documents necessary to protect and/or register the Ikaria Licensed Items. Except as provided in this Section 2.6, nothing in this Agreement shall be construed to grant to Pulse Technologies any right, title, interest, or license in or to the Ikaria Licensed Items or any of Ikaria's other names, logos, trademarks, trade dress, service marks, designs, marks, domain names, or IP. Pulse Technologies agrees not to contest the validity of, by act or omission jeopardize, or take any action inconsistent with, Ikaria's rights or goodwill in any Ikaria Licensed Item in any country, including by attempted registration of any Ikaria Licensed Item, or, in the case of the Licensed Marks, use or attempt registration of any confusingly similar mark or domain name.

2.6.3. Pulse Technologies shall promptly notify Ikaria of any known, threatened, or suspected infringement, imitation, or unauthorized use of the Ikaria Licensed Items by any third party. Ikaria, in its sole discretion, shall determine what action, if any, should be taken in response to any infringement, imitation, or unauthorized use of the Ikaria Licensed Items. Pulse Technologies shall take no action to enforce any rights in the Ikaria Licensed Items against any third party without the prior approval of Ikaria, which Ikaria may withhold in its sole discretion. Pulse Technologies shall use best efforts to cooperate with Ikaria's efforts in connection with enforcing its rights in the Ikaria Licensed Items, at Ikaria's expense, including making personnel available to testify and providing relevant documentation and information. Pulse Technologies agrees to become a co-party to litigation if Ikaria deems it advisable.

2.7. All rights and licenses granted under or pursuant to any section of this Agreement, including the licenses granted under this Article 2, are and will otherwise be deemed to be for purposes of Section 365(n) of the Bankruptcy Code, licenses of rights to "intellectual property" as defined in Section 101(35A) of the Bankruptcy Code. Each Party will retain and may fully exercise all of its respective rights and elections under the Bankruptcy Code. Each Party agrees that the other Party, as licensee of such rights under this Agreement, will retain and may fully exercise all

of its rights and elections under the Bankruptcy Code or any other provisions of applicable law outside the United States that provide similar protection for "intellectual property."

2.8. Pulse Technologies acknowledges and agrees that the limitations and restrictions set forth in this Agreement (including those set forth in Sections 2.5, 3.1, and 4) are reasonable and properly required for the adequate protection of Ikaria's interest in the Ikaria NO Business. If any such restriction is deemed to be unreasonable by a court of competent jurisdiction, the Parties shall submit to the reduction of such restrictions to such activities, geographical scope, or time period as such court shall deem reasonable.

2.9. Pulse Technologies shall promptly notify Ikaria if at any point Pulse Technologies is not actively and continuously engaged in the development or commercialization (either directly or through one or more Sublicensees) of an R&D Product for each of the following: (a) COPD, (b) PAH, or (c) IPF. Upon request from Ikaria from time to time, Pulse Technologies shall confirm in writing (within 10 business days after Pulse Technologies' receipt of such request from Ikaria) whether Pulse Technologies is engaged in such development or commercialization.

2.10. **NOTWITHSTANDING ANYTHING TO THE CONTRARY CONTAINED IN THIS AGREEMENT, NEITHER PARTY MAKES ANY REPRESENTATIONS OR EXTENDS ANY WARRANTIES OF ANY KIND, EXPRESS OR IMPLIED, AS TO ANY IP LICENSED HEREUNDER, INCLUDING ANY REPRESENTATIONS OR**

WARRANTIES OF MERCHANTABILITY, TITLE, FITNESS FOR A PARTICULAR PURPOSE, NONINFRINGEMENT, OR VALIDITY OF PATENT CLAIMS.

3. **Sublicensing**

3.1. Pulse Technologies hereby agrees that during the Term it shall not (without Ikaria's prior written consent, which Ikaria may withhold in its sole discretion) grant any Sublicenses under the R&D IP to an Ikaria Competitor.

3.2. All Sublicenses granted by Pulse Technologies of rights to practice under or otherwise use or have the benefit of any R&D IP shall be pursuant to written agreements. All such Sublicenses shall impose obligations, responsibilities and standards upon a Sublicensee that, in all material respects, are not less than those imposed on Pulse Technologies hereunder. Further, each Sublicense shall include a provision prohibiting the Sublicensee from further sublicensing any of the rights granted by Pulse Technologies. Each Sublicense shall specifically reference this Agreement and shall name Ikaria as an intended third party beneficiary of such Sublicense with the right to enforce the terms thereof against the Sublicensee directly and for its own benefit. Ikaria shall have the right to review any proposed Sublicense to ensure that it complies with the requirements of this Agreement; provided, however, that Pulse Technologies may only redact from the proposed Sublicense the amounts of payments to be made by either party thereunder. Pulse Technologies shall not enter into any Sublicense without having first obtained Ikaria's written confirmation that the proposed Sublicense meets the requirements of this Agreement, including the terms and conditions of this Section 3.2, which shall not be unreasonably withheld or delayed. As soon as reasonably practicable after execution of a Sublicense hereunder, Pulse Technologies shall provide a true and complete copy thereof to Ikaria, provided, however, that Pulse Technologies may redact only the amounts of payments to be made by either party thereunder.

4. **Agreements with Customers**

4.1. Pulse Technologies shall ensure that all R&D Products are used solely within the scope of the R&D Business. All provision of R&D Products to R&D Product Customers shall be made under an appropriate written agreement. Each such agreement shall include (a) a restriction requiring that such customer use the applicable R&D Product only for a use within the scope of the R&D Business, (b) a mechanism for Pulse Technologies to audit and confirm that such restriction is complied with, and (c) a termination right permitting Pulse Technology to terminate that agreement in the event such restriction has been violated. In the event of any such violation, Pulse Technologies shall ensure that such violation is promptly remedied, and if it is not, Pulse Technologies shall terminate the customer agreement in question. Use of an R&D Product by a Sublicensee or an R&D Product Customer outside of the scope of the R&D Business shall be deemed to be a material breach by Pulse Technologies of this Agreement.

4.2. Pulse Technologies agrees to provide (whether doing so directly or through a Subsidiary, Sublicensee, or otherwise) the delivery device portion of R&D Products to R&D Product Customers on a loaned or leased-basis only, and therefore agrees not to transfer title to the delivery device portion of any R&D Product to any R&D Product Customer.

5. **Certain Patent Prosecution and Maintenance Expenses.**

Pulse Technologies shall pay to Ikaria (a) [**] percent ([**]%) of the reasonable and documented out-of-pocket costs and expenses incurred by Ikaria during the Term for the prosecution and maintenance of the Core R&D Patents and (b) [**] percent ([**]%) of the reasonable and documented out-of-pocket costs and expenses incurred by Ikaria during the Term for the prosecution and maintenance of all other R&D Patent Rights. Pulse Technologies shall pay such amounts to Ikaria within thirty (30) days after Pulse Technologies' receipt of Ikaria's invoices therefor.

6. **Records and Audits.**

During the Term and for two (2) years thereafter, Pulse Technologies shall maintain (and shall require each Sublicensee to maintain) documentation and records sufficient to demonstrate its compliance with the requirements of this Agreement. Upon reasonable notice from Ikaria, Pulse Technologies shall provide (and shall require its Sublicensees to provide) to Ikaria or its agents with access to Pulse Technologies' (and its Sublicensees') premises during normal business hours to examine or copy all records requested by Ikaria or otherwise relevant to determine whether Pulse Technologies (and each Sublicensee) is in compliance with the requirements of this Agreement. Without limiting the generality of the foregoing, Ikaria shall have the right to review any and all (a) Sublicenses granted by Pulse Technologies to any R&D IP or Grant-Back IP and (b) agreements with R&D Product Customers.

7. **Technology Transfer.**

7.1. Technology Transfer Regarding Drug Product.

7.1.1. Ikaria and Pulse Technologies each acknowledge that they are entering into a separate Drug Clinical Supply Agreement on or prior to the Effective Date pursuant to which Ikaria will supply to Pulse Technologies 100% of Pulse Technologies' requirements for clinical supply of inhaled nitric oxide for use in Pulse Technologies' clinical trials in respect of R&D Products. Ikaria and Pulse Technologies each acknowledge that until such time as Ikaria is no longer providing 100% of Pulse Technologies' requirements for clinical supply of nitric oxide for use in Pulse Technologies' clinical trials in respect of R&D Products (or, as the case may be in the future, commercial supply), there is no need for Ikaria to disclose to Pulse Technologies any methods, processes, techniques, know-how, technology, data, or other information relating to or supporting the manufacture of nitric oxide for R&D

Product, other than as necessary to support Pulse Technologies' pursuant of R&D Product Approvals.

7.1.2. If there is a time when Ikaria no longer provides 100% of Pulse Technologies' requirements for clinical supply of nitric oxide for use in Pulse Technologies' clinical trials in respect of R&D Products (or, as the case may be, commercial supply), and at that point in time this Agreement remains in full force and effect, Ikaria shall, on Pulse Technologies' written request and at such times and in such manners and formats as are reasonably acceptable to both Parties, use commercially reasonable efforts to disclose to Pulse Technologies (or its designated Sublicensee) the methods, processes, techniques, know-how, technology, data, and other information used by Ikaria at that point in time to produce nitric oxide for Pulse Technologies for R&D Products (**"NO Know-How"**). Pulse Technologies acknowledges and agrees that nothing in this Agreement shall require Ikaria to transfer to Pulse Technologies or any designated Sublicensee any physical items used to, or in support of the, manufacture of nitric oxide (including production columns or testing equipment) or to transfer any personnel, it being understood and agreed that the NO Know-How disclosure provided for in this Section 7.1.2 consists solely of information in documentary form. Ikaria and Pulse Technologies agree to use commercially reasonable efforts to complete the disclosure of NO Know-How described in this Section 7.1.2 within 12 months after such disclosure is commenced.

7.2. Technology Transfer regarding R&D NO Delivery Devices. Ikaria and Pulse Technologies each acknowledge that they are entering into a separate Device Clinical Supply Agreement on or prior to the Effective Date pursuant to which Ikaria will supply to Pulse Technologies on a short term basis with 100% of Pulse Technologies' requirements for the supply of R&D NO Delivery Devices for use in Pulse Technologies' clinical trials in respect of R&D Products. In order to achieve an efficient and expeditious transition of manufacturing of R&D NO Delivery Devices from Ikaria to Pulse Technologies (or its designated Sublicensee), Ikaria shall, at such times and in such manners and formats as are reasonably acceptable to both Parties, use commercially reasonable efforts to disclose to Pulse Technologies (or its designated Sublicensee) the methods, processes, techniques, know-how, technology, data, and other information used by Ikaria as of the Effective Date to manufacture the R&D NO Delivery Devices (**"Device Know-How"**). Pulse Technologies acknowledges and agrees that nothing in this Agreement shall require Ikaria to transfer to Pulse Technologies or any designated Sublicensee any physical items used to, or in support of the, manufacture of R&D NO Delivery Device or to transfer any personnel, it being understood and agreed that the Device Know-How disclosure provided for in this Section 7.2 consists solely of information in documentary form. Ikaria and Pulse Technologies agree to use commercially reasonable efforts to complete the disclosure of Device Know-How described in this Section 7.2 within 12 months after the Effective Date.

8. **Regulatory Filings.**

8.1. COPD. Ikaria shall transfer to Pulse Technologies (and Pulse Technologies shall accept such transfer) [**] within 30 days after the Effective Date.

8.2. PAH. Ikaria shall transfer to Pulse Technologies (and Pulse Technologies shall accept such transfer) [**] within 30 days after the Effective Date.

8.3. Right of Reference. Pulse Technologies hereby grants Ikaria, Ikaria Parent Company, and any other subsidiaries of Ikaria Parent Company the right to reference any and all R&D Product Approvals for R&D Products (including [**]), at no cost to Ikaria, whether such R&D Product Approvals are held by Pulse Technologies, an Affiliate of Pulse Technologies, or a Sublicensee of Pulse Technologies, for any and all purposes relating to the Ikaria NO Business. Pulse Technologies shall have the right to reference [**] solely for purposes of pursuing R&D Product Approvals for R&D Products within the R&D Business. Upon request, each Party shall provide the other Party access to and copies of any regulatory filings or supporting materials covered by this Section 8.3 for purposes of exercising its rights granted by this Section 8.3. Further, each Party hereby agrees to provide, and to cause its Affiliates and Sublicensees to provide, the other Party with a letter of consent to permit such referencing, which letter may be provided by the receiving Party or its Affiliates to the applicable governmental authorities for purposes of exercising its rights or performing its obligations hereunder. Each party shall otherwise reasonably cooperate with the other to affect such referencing.

9. **Intellectual Property Rights.**

9.1. Prosecution and Maintenance of Patent Rights.

9.1.1. Initial Right. As between the Parties, (a) Ikaria shall have the initial right, but not the obligation, to file, prosecute, and maintain all R&D Patent Rights in the Territory, at Pulse Technologies' expenses to the extent set forth in Section 5 and otherwise at Ikaria's expense, and (b) Pulse Technologies shall have the initial right, but not the obligation, to file, prosecute, and maintain the Grant-Back Patent Rights in the Territory, at Pulse Technologies' expense.

9.1.2. Prosecution and Maintenance. With respect to the rights described in Section 9.1.1, upon a request by either Party, the Parties will discuss and consider in good faith filing separate Patent Rights for claims that cover R&D Products specifically or generically and claims that cover Grant-Back Field Products specifically or generically.

9.1.3. Ikaria Step-In Right. If Pulse Technologies declines to file, prosecute, or maintain any Grant-Back Patent Right, elects to allow any Grant-Back Patent Right to lapse in any country, elects to abandon any Grant-Back Patent Right before all appeals within the respective patent office

or other applicable government authority have been exhausted or to abandon any Grant-Back Patent Right, then:

(a) Pulse Technologies shall provide Ikaria with reasonable notice of such decision (no less than 30 days prior to any abandonment or loss of rights with respect to such Patent Right) so as to permit Ikaria to decide whether to file, prosecute, or maintain such Patent Right and to take any necessary action.

(b) Ikaria may, at Ikaria's sole cost and expense, assume control of the filing, prosecution, and/or maintenance of such Patent Right in the name of the owner(s) of such Patent Right; provided, at the request of Pulse Technologies, Ikaria shall provide Pulse Technologies, on a timely basis so that Pulse Technologies can review and comment prior to filing or response, with drafts of all patent applications and other material submissions to, and copies of all office actions and other communications from, any patent authorities pertaining to the Grant-Back Patent Rights, and Ikaria shall consider in good faith all timely comments provided by Pulse Technologies. Notwithstanding the foregoing, in the event that Pulse Technologies reasonably determines that Ikaria's filing, prosecution and/or maintenance of any Patent Right could materially impair Pulse Technologies' business operations or IP rights, Pulse Technologies may notify Ikaria of the same, in which case Ikaria shall not undertake or shall cease any such filing, prosecution and/or maintenance.

(c) Pulse Technologies shall, at Ikaria's expense and reasonable request, assist and cooperate in the filing, prosecution, and maintenance of such Grant-Back Patent Right.

9.1.4. Pulse Technologies Step-In Right for Core R&D Patents. If Ikaria declines to file, prosecute, or maintain any Core R&D Patent, elects to allow any Core R&D Patent to lapse in any country, elects to abandon any Core R&D Patent before all appeals within the respective patent office or other applicable government authority have been exhausted or to abandon any Core R&D Patent, then:

(a) Ikaria shall provide Pulse Technologies with reasonable notice of such decision (no less than 30 days prior to any abandonment or loss of rights with respect to such Core R&D Patent) so as to permit Pulse Technologies to decide whether to file, prosecute, or maintain such Core R&D Patent and to take any necessary action.

(b) Pulse Technologies may, at Pulse Technologies' sole cost and expense, assume control of the filing, prosecution, and/or

maintenance of such Core R&D Patent in the name of Ikaria; provided, at the request of Ikaria, Pulse Technologies shall provide Ikaria, on a timely basis so that Ikaria can review and comment prior to filing or response, with drafts of all patent applications and other material submissions to, and copies of all office actions and other communications from, any patent authorities pertaining to the R&D Patent Rights, and Pulse Technologies shall consider in good faith all timely comments provided by Ikaria. Notwithstanding the foregoing, in the event that Ikaria reasonably determines that Pulse Technologies' filing, prosecution and/or maintenance of any Patent Right could materially impair Ikaria's business operations or IP rights, Ikaria may notify Pulse Technologies of the same, in which case Pulse Technologies shall not undertake or shall cease any such filing, prosecution and/or maintenance.

(c) Ikaria shall, at Pulse Technologies' expense and reasonable request, assist and cooperate in the filing, prosecution, and maintenance of such Core R&D Patent.

9.2. Enforcement of Patent Rights.

9.2.1. Notice. Pulse Technologies shall, as soon as reasonably practicable but in any event within 30 days, provide Ikaria with written notice reasonably detailing any known or alleged infringement by a Third Party of the R&D Patent Rights hereunder, and shall notify Ikaria of any "patent certification" filed in the United States under 21 U.S.C. §355(b)(2) or 21 U.S.C. §355(j)(2) or similar provisions in other jurisdictions, and of any declaratory judgment, opposition, or similar action alleging the invalidity, unenforceability or non-infringement of the R&D Patent Rights licensed hereunder, in each case, as soon as reasonably practicable but in any event within seven (7) days.

9.2.2. Infringement.

(a) With respect to any actual or suspected infringement of the R&D Patent Rights by a Third Party, Ikaria shall have the initial right, but not the obligation, to initiate a legal action against such Third Party with respect to such infringement, at Ikaria's expense.

(b) With respect to any actual or suspected infringement of the Grant-Back Patent Rights by a Third Party making, using, or selling a product that is or may be, in Ikaria's reasonable judgment, competitive with a Grant-Back Field Product, Ikaria shall have the initial right, but not the obligation, to initiate a legal action against such Third Party with respect to such infringement, at Ikaria's expense.

(c) If, after receiving a written request from Pulse Technologies to take such action, Ikaria declines to timely take any action (which, with respect to any "patent certification" filed in the United States under 21 U.S.C. §355(b)(2) or 21 U.S.C. §355(j)(2) or similar provisions in other jurisdictions, shall mean that Ikaria has not provided Pulse Technologies with written notification within 30 days that Ikaria intends to bring an infringement action within the statutorily set 45 day period) with respect to any infringement of any Core R&D Patents or Grant-Back Patent Rights by a Third Party making, using, or selling a product that is or may be, in Pulse Technologies' reasonable judgment, competitive with an R&D Product, Pulse Technologies shall have the right to initiate a legal action against such Third Party with respect to such competitive infringement, at Pulse Technologies' expense. Ikaria may, but, subject to Section 9.3, shall not be required to, join in such action as a party and may be represented by counsel of its choice, at Ikaria's expense. In the case of any action taken under this Section 9.2.2(c), Pulse Technologies shall provide Ikaria, on a timely basis so that Ikaria can review and comment prior to filing or response, with drafts of all pleadings and shall consider in good faith all timely comments provided by Ikaria. Notwithstanding the foregoing, in the event that Ikaria reasonably determines that Pulse Technologies' pursuit of any action with respect to infringement of any Core R&D Patents could materially impair Ikaria's business operations or IP rights, Ikaria may notify Pulse Technologies of the same, in which case Pulse Technologies shall not undertake or shall cease any such action, provided that Ikaria shall reasonably consider any adverse effect of such restriction on Pulse Technologies' business before imposing such restriction.

(d) Pulse Technologies may, but, subject to Section 9.3, shall not be required, to join in any action by Ikaria pursuant to Section 9.2.2(a) or 9.2.2(b) as a party, and may be represented by counsel of its choice, at Pulse Technologies' request and expense. In the case of any action taken under Section 9.2.2(a) or 9.2.2(b), Ikaria shall provide Pulse Technologies, on a timely basis so that Pulse Technologies can review and comment prior to filing or response, with drafts of all pleadings and shall consider in good faith all timely comments provided by Pulse Technologies. Notwithstanding the foregoing, in the event that Pulse Technologies reasonably determines that Ikaria's pursuit of any action with respect to infringement of any Grant-Back Patent Rights could materially impair Pulse Technologies' business operations or IP rights, Pulse Technologies may notify Ikaria of the same, in which case Ikaria shall not undertake or shall cease

any such action, provided that Pulse Technologies shall reasonably consider any adverse effect of such restriction on Ikaria's business before imposing such restriction.

(e) At the reasonable request and expense of the Party initiating the legal action pursuant to this Section 9.2.2, the other Party shall provide reasonable assistance to the initiating Party in connection with such action, at the initiating Party's sole cost and expense.

(f) Without the prior written consent of the Party Controlling the Patent Rights asserted pursuant to this Section 9.2.2, such consent not to be unreasonably withheld of delayed, the initiating Party shall not enter into any settlement admitting the invalidity of, or otherwise impairing the Controlling Party rights under this Agreement in, such asserted Patent Rights.

(g) Any recoveries resulting from an action asserted pursuant to this Section 9.2.2 shall be applied as follows:

(i) First, to reimburse the non-initiating Party for all out-of-pocket costs in connection with such proceeding; and

(ii) Second, the remainder of the recovery shall be retained by the initiating Party.

(h) Notwithstanding any other term or condition of this Agreement, and for the avoidance of doubt, Ikaria does and shall retain the sole exclusive right to initiate a legal action with respect to any actual or suspected infringement of any of the R&D Patent Rights (including the HF Patents) other than the Core R&D Patents, which shall be subject to the terms of this Section 9.2.2.

9.3. Other Actions. Except as provided in Section 9.2, as between the Parties, the Controlling Party shall have the sole right to protect its IP rights licensed hereunder from any actual or suspected infringement or misappropriation or any challenges to the validity or enforceability of such IP rights licensed hereunder. In any legal action so brought by the non-owning Party, the owning Party shall join in such action as a party at the non-owning Party's request and expense in the event that an adverse party asserts, the court rules or other laws provide, or the non-owning Party determines in good faith, that a court would lack jurisdiction or the non-owning Party would lack standing based on the owning Party's absence as a party in such suit; but control of such action shall remain with the non-owning Party. At the Controlling Party's reasonable request and expense, the other Party shall provide reasonable assistance to the Controlling Party in connection with such action. Any recoveries resulting from such an action shall allocated in the same manner as set forth in Section 9.2.2(g) for recoveries made in actions initiated under this Section 9.3.

9.4. No Challenge. Pulse Technologies shall not, and will cause or ensure that its Affiliates shall not, challenge, or voluntarily assist any third party in challenging, the enforcement or validity of the R&D IP or Ikaria Licensed Items (including in any proceeding before the United States Patent and Trademark Office or the patent or trademark offices of any other jurisdiction). Without limiting any other rights or remedies that Ikaria may have, at law or equity, in the event that Pulse Technologies or its Affiliate challenges or supports a challenge to the enforcement or validity of the R&D IP or Ikaria Licensed Items, Ikaria shall have the right, to the extent permitted by law, to terminate this Agreement immediately and at Ikaria's sole discretion upon written notice to Pulse Technologies. Ikaria shall not, and will cause or ensure that its Affiliates shall not, challenge, or voluntarily assist any third party in challenging, the enforcement or validity of the Grant-Back IP (including in any proceeding before the United States Patent and Trademark Office or the patent or trademark offices of any other jurisdiction). Without limiting any other rights or remedies that Pulse Technologies may have, at law or equity, in the event that Ikaria or its Affiliate challenges or supports a challenge to the enforcement or validity of the Grant-Back IP, Pulse Technologies shall have the right, to the extent permitted by law, to terminate this Agreement immediately and at Pulse Technologies' sole discretion upon written notice to Ikaria.

10. **CONFIDENTIAL INFORMATION**

10.1. Non-Use and Non-Disclosure of Confidential Information. Each Receiving Party agrees that during the Term and for a period of five (5) years thereafter, all Confidential Information of the Disclosing Party (a) shall not be used by the Receiving Party except to perform its obligations or exercise its rights under this Agreement, (b) shall be maintained in confidence by the Receiving Party, and (c) except as permitted by Section 10.2.1 and 10.2.2, shall not be disclosed by the Receiving Party to any Person without the prior written consent of the Disclosing Party; provided, however, that trade secrets shall remain subject to the requirements of this Section 10 for so long as they maintain their status as trade secrets under applicable law.

10.2. Permitted Disclosures.

10.2.1. The Receiving Party may provide the Disclosing Party's Confidential Information (a) to its Affiliates, Sublicensees and potential Sublicensees and to their employees, consultants, advisors and contractors who have a need to know such Confidential Information for purposes of the Receiving Party exercising or granting licenses or Sublicenses as permitted herein, (b) in communications with existing or bona fide prospective acquirers, merger partners, lenders or investors, in each case of (a) and (b), on a need to know basis and under appropriate confidentiality provisions substantially equivalent to those of this Agreement.

10.2.2. The Receiving Party may provide the Disclosing Party's Confidential Information:

(a) to the Receiving Party's employees, consultants, advisors and contractors who have a need to know such Confidential Information and are bound by an obligation to maintain the confidentiality of the Disclosing Party's Confidential Information;

(b) to patent offices or regulatory authorities in order to seek or obtain Patent Rights or approval to conduct clinical trials, or to gain regulatory approvals;

(c) as reasonably required for development and commercialization of R&D Products or Grant-Back Field Products, as applicable, in accordance with customary commercial practice (e.g., consistent with customary public disclosures regarding scientific and technical information relating to marketed drug products and medical devices); and

(d) if such disclosure is required by law (including by rules or regulations of any securities exchange) or to defend or prosecute litigation or arbitration; provided, that prior to such disclosure, to the extent permitted by law (including by subpoena or other governmental order) or such rules or regulations, the Receiving Party promptly notifies the Disclosing Party of such requirement and furnishes only that portion of the Disclosing Party's Confidential Information that the Receiving Party is legally required to furnish.

11. **Term and Termination.**

11.1. Term.

11.1.1. R&D Products. This Agreement shall commence on the Effective Date, and unless earlier terminated pursuant to this Section 11, shall terminate on an R&D Product by R&D Product basis as follows (the "**Term**"):

(a) with respect to R&D Products for COPD, this Agreement (including the license granted under Section 2.1) shall terminate at the point in time when Pulse Technologies is no longer actively and continuously engaged in the development or commercialization (either directly or through one or more Sublicensees) of an R&D Product for COPD;

(b) with respect to R&D Products for PAH, this Agreement (including the license granted under Section 2.1) shall terminate at the point in time when Pulse Technologies is no longer actively and continuously engaged

in the development or commercialization (either directly or through one or more Sublicensees) of an R&D Product for PAH; and

(c) with respect to R&D Products for IPF (including the license granted under Section 2.1), this Agreement shall terminate at the point in time when Pulse Technologies is no longer actively and continuously engaged in the development or commercialization (either directly or through one or more Sublicensees) of an R&D Product for IPF.

11.1.2. Patent Rights. The applicable license granted under Section 2.1 and Section 2.2 for each Patent Right shall, if not terminated earlier, expire upon expiration of such Patent Right.

11.2. Termination. This Agreement may be terminated immediately by notice at any time during the Term (subject to any applicable cure period):

11.2.1. By either Party, if a court of competent jurisdiction or governmental authority, regulatory or administrative agency or commission shall have enacted any law, statute, rule, or regulation or issued any final and non-appealable order or decree that permanently restrains, enjoins, or otherwise prohibits either Party from performing or substantially performing under this Agreement;

11.2.2. By either Party upon any voluntary or involuntary bankruptcy or insolvency of the other Party, or if any action or proceeding is instituted against the other Party relating to any of the foregoing and said action or other proceeding is not dismissed within 60 days after institution thereof; and

11.2.3. By either Party if the other Party shall have breached or failed to comply with any material term or condition required to be performed or complied with by such other Party, and such breach or failure is not cured within 30 days after written notice thereof by the terminating Party; or

11.2.4. By Ikaria, if Pulse Technologies or any of its Affiliates materially breaches the provisions of the Agreement Not to Compete between Ikaria Acquisition, Inc. and Pulse Technologies, dated October 18, 2013, the Agreement Not to Compete between Ikaria Acquisition, Inc. and Bellerophon BCM LLC (f/k/a Ikaria Development Subsidiary One LLC), dated September 20, 2013, or the Agreement Not to Compete between Ikaria Acquisition, Inc. and Bellerophon Therapeutics LLC (f/k/a Ikaria Development LLC), dated October 18, 2013 and fails to cure such breach in all material respects within 30 days after written notice thereof by Ikaria (the **"Non-Compete Agreements"**).

11.2.5. By Ikaria, if (a) Pulse Technologies or any Person that is a successor in interest to Pulse Technologies' rights under this Agreement (whether by

assignment of this Agreement or otherwise) markets a generic nitric oxide product that is competitive with Ikaria's INOMAX product in any country in which Ikaria, Ikaria Parent Company or any other subsidiary of Ikaria Parent Company markets such INOMAX product and (b) Pulse Technologies or such successor, as applicable, does not cease such marketing of a competing product in such country within 30 days after notice from Ikaria of its intent to terminate this Agreement pursuant to this Section 11.2.5.

11.3. Material Obligations. For purposes of Section 11.2.3, each of the following Sections or subsections shall be deemed a "material term or condition" of this Agreement without any further obligation of either Party to prove materiality: Sections 2.1, 2.2, 2.5, 2.6, 2.9, 3, 4, 8, 9.4, 10, 12, and 15.

11.4. Upon Termination.

11.4.1. R&D Product Approvals.

(a) Upon termination of this Agreement with respect to R&D Products for COPD, Pulse Technologies shall, upon written request from Ikaria, transfer to Ikaria (or to another party designated by Ikaria) any and all R&D Product Approvals relating thereto (including [**] and any NDA granted thereon).

(b) Upon termination of this Agreement with respect to R&D Products for PAH, Pulse Technologies shall, upon written request from Ikaria, transfer to Ikaria (or to another party designated by Ikaria) any and all R&D Product Approvals relating thereto (including [**] and any NDA granted thereon).

(c) Upon termination of this Agreement with respect to R&D Products for IPF, Pulse Technologies shall, upon written request from Ikaria, transfer to Ikaria (or to another party designated by Ikaria) any and all R&D Product Approvals relating thereto (including any IND therefor and any NDA granted thereon).

(d) Pulse Technologies shall reasonably cooperate with Ikaria to effectuate any of the transfers referenced in 11.4(a)-(c).

11.5. Survival. The following Sections shall survive any expiration or earlier termination of this Agreement: 1, 2.2, 2.3, 2.7, 2.10, 6, 9.4, 10, 11.4, 11.5, 12.1, 14, 16, 17, 18, 19, 20, 21, 22, 23, 24 and 25. Notwithstanding the foregoing, the license granted to Ikaria by Pulse Technologies under Section 2.2 shall not survive termination of this Agreement by Pulse Technologies pursuant to the final sentence of Section 9.4 or by Pulse Technologies pursuant to Section 11.2.3 based on a material uncured breach of this Agreement by Ikaria. Expiration or earlier termination of this Agreement for any reason shall be without prejudice to any rights that shall have accrued to the benefit of any Party prior to such termination,

relinquishment, or expiration, and any and all damages or remedies arising from any breach hereunder.

12. **Disclaimer of Warranties.**

12.1. Disclaimer of Warranties. Other than as expressly set forth in this Agreement, neither Party makes any other representations and warranties, express or implied, under this Agreement, including, regarding merchantability, fitness for a particular purpose, title, infringement of third party intellectual property rights, or validity or enforceability of intellectual property.

13. **Amendments.**

The Parties may amend this Agreement only by a written agreement signed by the Party intended to be bound by the amendment and that identifies itself as an amendment to this Agreement.

14. **Severability.**

Any term or provision of this Agreement that is invalid or unenforceable in any situation in any jurisdiction shall not affect the validity or enforceability of the remaining terms and provisions hereof or thereof or the validity or enforceability of the offending term or provision in any other situation or in any other jurisdiction. If the final judgment of a court of competent jurisdiction declares that any term or provision hereof is invalid or unenforceable, the Parties agree that the court making such determination shall have the power to limit the term or provision, to delete specific words or phrases, or to replace any invalid or unenforceable term or provision with a term or provision that is valid and enforceable and that comes closest to expressing the intention of the invalid or unenforceable term or provision, and this Agreement shall be enforceable as so modified. In the event such court does not exercise the power granted to it in the prior sentence, the Parties agree to replace such invalid or unenforceable term or provision with a valid and enforceable term or provision that shall achieve, to the extent possible, the economic, business and other purposes of such invalid or unenforceable term.

15. **Assignment; Change in Control.**

15.1. Except with Ikaria's prior written consent (which may be withheld in its sole discretion), Pulse Technologies shall not assign or transfer this Agreement or any of its rights, or delegate any of its duties or obligations, under this Agreement, whether voluntarily, by merger or operation of law or otherwise, and whether directly or indirectly, to an Ikaria Competitor. A change in control (as defined below) of Pulse Technologies to an Ikaria Competitor shall be deemed to be a prohibited assignment under this Section 15.1. For the purposes of this Section 15.1, a **"change in control"** shall mean the sale or transfer of a majority of the share capital or membership interests (or right to direct the operations) of Pulse Technologies, or the sale or transfer of a substantial portion of its business or assets or a similar type of transaction.

15.2. Ikaria may assign or transfer this Agreement, in whole or part, to any of its Affiliates or to the successor in interest to all or substantially all of its assets (whether voluntarily, by merger or operation of law or otherwise). For the purposes of this Section 15.2, a change of control of Ikaria, Ikaria Parent Company, or any of their respective subsidiaries shall not be deemed to require the consent of Pulse Technologies.

15.3. Each Party shall require any assignee or transferee to assume this Agreement in writing, and that it will faithfully and fully comply with the terms and conditions of this Agreement (including the scope of license restrictions set forth in Sections 2.1 and 2.2, as the case may be).

15.4. This Agreement and the rights and obligations hereunder shall be binding upon and inure to the benefit of the Parties hereto, and their respective successors and permitted assigns.

15.5. In the event of Pulse Technologies' bankruptcy under Chapter 11 of Title 11 of the U.S. Code, Pulse Technologies and the trustee shall not have the right to assume, or assume and assign, this Agreement to the debtor in possession or the entity emerging from bankruptcy or to a Third Party. Any assignment in violation of this Agreement shall be null and void.

16. **Equitable Relief.**

Pulse Technologies acknowledges and agrees that Ikaria and its Affiliates would be irreparably harmed by a breach of any of Section 2.1, 2.2, 2.5, 2.6, 4, 10 or 15, that Ikaria's remedies at law for such a breach would be inadequate and, in recognition of those facts, in the event of the breach or threatened breach by Pulse Technologies of any of its Sublicensees of any of Section 2.1, 2.2, 2.5, 2.6, 4, 10 or 15, it is agreed that, in addition to its remedies at law, Ikaria shall be entitled to seek, and Pulse Technologies hereby consents to Ikaria's seeking, equitable relief in the form of specific performance, temporary restraining order, temporary or permanent injunction, or any other equitable remedy which may then be available, without posting any bond or other undertaking.

17. **Entire Agreement.**

This Agreement and its exhibits, together with the Separation Agreement and the Non-Compete Agreements, constitutes the final agreement between the Parties, and is the complete and exclusive statement of the Parties' agreement on the matters contained herein and therein. All prior and contemporaneous negotiations and agreements between the Parties with respect to the matters contained herein and therein are superseded by this Agreement the Separation Agreement and the Non-Compete Agreements, as applicable. Nothing in this Agreement shall be deemed to limit Pulse Technologies' obligations under the Agreement Not to Compete between Ikaria Acquisition Inc. and Pulse Technologies dated October 18, 2013.

18. **Waiver.**

The Parties may waive a provision of this Agreement only by a writing signed by the Party intended to be bound by the waiver. A Party is not prevented from enforcing any right, remedy or condition in the Party's favor because of any failure or delay in exercising any right or remedy or in requiring satisfaction of any condition, except to the extent that the Party specifically waives the same in writing. A written waiver given for one matter or occasion is effective only in that instance and only for the purpose stated. A waiver once given is not to be construed as a waiver for any other matter or occasion. Any enumeration of a Party's rights and remedies in this Agreement is not intended to be exclusive, and a Party's rights and remedies are intended to be cumulative to the extent permitted by law and include any rights and remedies authorized in law or in equity.

19. **Further Assurances.**

Each Party agrees to execute, acknowledge and deliver such further documents and instruments and do any other acts, from time to time, as may be reasonably necessary, to effectuate the purposes of this Agreement.

20. **Third Parties.**

None of the provisions of this Agreement shall be enforceable by any Person who is not a Party to this Agreement.

21. **Notices.**

All notices and other communications under this Agreement shall be in writing and shall be deemed duly delivered (a) four Business Days after being sent by registered or certified mail, return receipt requested, postage prepaid, (b) one Business Day after being sent for next Business Day delivery, fees prepaid, via a reputable nationwide overnight courier service, or (c) on the date of confirmation of receipt (or, the first Business Day following such receipt if the date of such receipt is not a Business Day) of transmission by facsimile, in each case to the intended recipient as set forth below.

If to Ikaria:

INO Therapeutics LLC

Perryville III Corporate Park
53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel

If to Pulse Technologies:

Perryville III Corporate Park

53 Frontage Road, Third Floor
P. O. Box 9001
Hampton, NJ 08827
Attention: General Counsel

Either Party may give any notice or other communication hereunder using any other means (including personal delivery, messenger service, ordinary mail or electronic mail), but no such notice or other communication shall be deemed to have been duly given unless and until it actually is received by the Party for whom it is intended. Either Party may change the address to which notices and other communications hereunder are to be delivered by giving the other Party notice in the manner herein set forth.

22. **Governing Law.**

The internal Laws of the State of Delaware (without giving effect to any choice or conflict of law provision or rule, whether of the State of Delaware or any other jurisdiction, that would cause the application of Laws of any jurisdiction other than those of the State of Delaware) shall govern the construction, interpretation and other matters arising out of or in connection with this Agreement (whether arising in contract, tort, equity or otherwise).

23. **Jurisdiction.**

If any dispute, controversy or claim arises out of or in connection with this Agreement, the Parties irrevocably (and the Parties shall cause each other member of their respective Group to irrevocably) (a) consent and submit to the exclusive jurisdiction of the Court of Chancery of the State of Delaware, New Castle County, or, if that court does not have jurisdiction, a federal court sitting in Wilmington, Delaware, (b) waive any objection to that choice of forum based on venue or to the effect that the forum is not convenient, and (c) WAIVE TO THE FULLEST EXTENT PERMITTED BY LAW ANY RIGHT TO TRIAL OR ADJUDICATION BY JURY. Either Party may make service on the other Party by sending or delivering a copy of the process to the other Party at the address and in the manner provided for the giving of notices in Section 21. Nothing in this Section 23, however, shall affect the right to serve legal process in any other manner permitted by Law.

24. **Exclusion of Consequential Damages.**

EXCEPT FOR CLAIMS OF A THIRD PARTY ARISING FROM THE OTHER PARTY'S GROSS NEGLIGENCE OR WILLFUL MISCONDUCT, NEITHER PARTY, NOR ANY OF THEIR RESPECTIVE AFFILIATES OR SUBLICENSEES, WILL BE LIABLE TO THE OTHER PARTY TO THIS AGREEMENT, ITS AFFILIATES OR SUBLICENSEES FOR ANY INDIRECT, INCIDENTAL, CONSEQUENTIAL, SPECIAL OR PUNITIVE DAMAGES OR LOST PROFITS OR ROYALTIES, LOST DATA OR COST OF PROCUREMENT OF SUBSTITUTE GOODS OR SERVICES, WHETHER LIABILITY IS ASSERTED IN CONTRACT, TORT, INDEMNITY OR CONTRIBUTION, AND IRRESPECTIVE OF WHETHER THAT PARTY OR ANY

REPRESENTATIVE OF THAT PARTY HAS BEEN ADVISED OF, OR OTHERWISE MIGHT HAVE ANTICIPATED THE POSSIBILITY OF, ANY SUCH LOSS OR DAMAGE.

25. **Interpretation.**

Both Parties have had the opportunity to have this Agreement reviewed by an attorney; therefore, neither this Agreement nor any provision hereof shall be construed against the drafter of this Agreement.

26. **Counterparts.**

This Agreement may be executed in two counterparts (including by facsimile or by an electronic scan delivered by electronic mail), each of which shall be deemed an original but both of which together shall be considered one and the same agreement and shall become effective when counterparts have been signed by each Party and delivered to the other Party, it being understood that both Parties need not sign the same counterpart.

IN WITNESS WHEREOF, each Party has caused this Agreement to be executed by its duly authorized representatives effective as of the Effective Date.

INO Therapeutics LLC		**Bellerophon Pulse Technologies LLC**	
By:	/s/ Matthew M. Bennett	By:	/s/ Daniel Tassé
Name:	Matthew M. Bennett	*Name*:	Daniel Tassé

Title:	Vice President & Secretary	*Title*:	Chief Executive Officer

[Signature Page to Exclusive Cross-License, Technology Transfer, and Regulatory Matters Agreement]

Exhibit A

Restricted Abilities, Attributes, Capabilities, Capacities, Functions, and Specifications for R&D NO Delivery Devices

[**]

FIRST AMENDMENT TO EXCLUSIVE CROSS-LICENSE, TECHNOLOGY TRANSFER, AND REGULATORY MATTERS AGREEMENT

THIS FIRST AMENDMENT TO EXCLUSIVE CROSS-LICENSE, TECHNOLOGY TRANSFER, AND REGULATORY MATTERS AGREEMENT (this "Amendment") is entered into the later of the dates in the signature block below (the "Amendment Effective Date") by and between INO Therapeutics LLC, d/b/a Ikaria, a Delaware limited liability company having a place of business at Perryville III Corporate Park, 53 Frontage Road, Third Floor, Hampton, NJ 08827 ("Ikaria"), and Bellerophon Pulse Technologies LLC, a Delaware limited liability company, with offices at Perryville III Corporate Park, 53 Frontage Road, Third Floor, Hampton, NJ 08827 ("Pulse Technologies"). Ikaria and Pulse Technologies may be individually referred to as a "Party" and together as the "Parties."

WHEREAS, Ikaria and Pulse Technologies entered into that certain Exclusive Cross-License, Technology Transfer, and Regulatory Matters Agreement dated as of February 9, 2014 (the "Agreement");

WHEREAS, Ikaria and Pulse Technologies now wish to amend certain provisions of the Agreement.

NOW, THEREFORE, in consideration of the mutual covenants contained herein, and for other good and valuable consideration, the receipt and sufficiency of which are hereby acknowledged, the Parties hereto agree as follows:

1. Modification of Section 1.2.34. Section 1.2.34 of the Agreement is hereby amended and restated in its entirety as follows:

1.2.34. "**R&D Product Approval**" shall mean any applications, approvals, clearances, or other government authorizations, foreign or domestic, of any type of or for an R&D Product. With respect to an R&D Product for COPD, the term "R&D Product Approval" shall include the COPD Filings. With respect to an R&D Product for PAH, the term "R&D Product Approval shall include the PAH Filings.

2. New Definitions. The following new subsections shall be added to Section 1.2 of the Agreement:

1.2.42 "**COPD Filings**" shall mean [**].

1.2.43 "**PAH Filings**" shall mean [**].

3. Modification of Section 8. Section 8 of the Agreement is hereby amended and restated in its entirety as set forth below:

8. **Regulatory Filings.**

8.1 COPD. Ikaria shall transfer to Pulse Technologies (and Pulse Technologies shall accept such transfer) the COPD Filings within 120 days after the Effective Date.

8.2. PAH. Ikaria shall transfer to Pulse Technologies (and Pulse Technologies shall accept such transfer) the PAH Filings within 120 days after the Effective Date.

8.3. Right of Reference. Pulse Technologies hereby grants Ikaria, Ikaria Parent Company, and any other subsidiaries of Ikaria Parent Company the right to reference any and all R&D Product Approvals for R&D Products (including the COPD Filings and the PAH Filings), at no cost to Ikaria, whether such R&D Product Approvals are held by Pulse Technologies, an Affiliate of Pulse Technologies, or a Sublicensee of

Pulse Technologies, for any and all purposes relating to the Ikaria NO Business. Pulse Technologies shall have the right to reference Ikaria's [**] and Ikaria's [**] solely for purposes of pursuing R&D Product Approvals for R&D Products within the R&D Business. Upon request, each Party shall provide the other Party access to and copies of any regulatory filings or supporting materials covered by this Section 8.3 for purposes of exercising its rights granted by this Section 8.3. Further, each Party hereby agrees to provide, and to cause its Affiliates and Sublicensees to provide, the other Party with a letter of consent to permit such referencing, which letter may be provided by the receiving Party or its Affiliates to the applicable governmental authorities for purposes of exercising its rights or performing its obligations hereunder. Each party shall otherwise reasonably cooperate with the other to affect such referencing.

4. Ratification of Agreement. Except as set forth in Sections 1 through 3 of this Amendment, all of the other terms and conditions of the Agreement are hereby ratified and confirmed to be of full force and effect, and shall continue in full force and effect. This Amendment is hereby integrated into and made a part of the Agreement.

5. Counterparts. This Amendment may be executed in two counterparts, each of which shall be effective as of the Amendment Effective Date, and all of which shall constitute one and the same instrument. Each such counterpart shall be deemed an original, and it shall not be necessary in making proof of this Amendment to produce or account for more than one such counterpart.

6. Execution and Delivery. This Amendment shall be deemed executed by the parties when any one or more counterparts hereof, individually or taken together, bears the signatures of each of the parties hereto.

IN WITNESS WHEREOF, each Party has caused this Amendment to be executed by its duly authorized representatives effective as of the Amendment Effective Date.

INO THERAPEUTICS LLC d/b/a IKARIA	BELLEROPHON PULSE TECHNOLOGIES LLC
By: /s/ William Scheinler	By: /s/ Manesh Naidu
Name: William Scheinler	Name: Manesh Naidu
Title: Assistant Secretary	Title: Vice President
3/27/2014	3/27/2014

Exhibit 10.15

Execution Version

Confidential Materials omitted and filed separately with the
Securities and Exchange Commission. Double asterisks denote omissions.

TRANSITION SERVICES AGREEMENT

THIS TRANSITION SERVICES AGREEMENT (the "Agreement") is made as of February 9, 2014, by and between Bellerophon Therapeutics LLC, a Delaware limited liability company ("R&DCo"), and Ikaria, Inc., a Delaware corporation ("Ikaria"). In this Agreement, each of R&DCo and Ikaria are sometimes referred to individually as a "Party" and, collectively, as the "Parties."

WHEREAS, R&DCo and Ikaria are parties to a Separation and Distribution Agreement of even date herewith (the "Separation Agreement"), which sets forth the terms upon which Ikaria will be separated into two independent companies, one for each of (a) the Ikaria Business (such term and each other capitalized term used but not defined herein to have the meanings given to such terms in the Separation Agreement), which shall continue to be owned and conducted, directly or indirectly, by Ikaria, and (b) the R&DCo Business, which shall be owned and conducted, directly or indirectly, by R&DCo;

WHEREAS, R&DCo and the other R&DCo Group Members have requested that Ikaria provide temporary, transition services while the R&DCo Group Members are working to provide those services for themselves (either directly or by retaining third party providers); and

WHEREAS, Ikaria is willing to provide those services on a limited time basis on an as requested, as available basis, all on the terms and conditions set forth herein;

NOW, THEREFORE, in consideration of the foregoing and the mutual covenants and agreements set forth below, and other good and valuable consideration, the receipt and adequacy of which is hereby acknowledged, the Parties hereby agree as follows:

1. SERVICES

1.1 General. During the term of this Agreement, Ikaria (or another Ikaria Group Member) shall use commercially reasonable efforts to provide, or cause such Ikaria Group Member to use commercially reasonable efforts to provide, the services (individually, a "Service" and collectively, the "Services") set forth in Schedule A attached hereto to R&DCo (or another R&DCo Group Member). R&DCo acknowledges and agrees that nothing in this Agreement shall require Ikaria or any Ikaria Group Member to hire, obtain, or retain additional resources of any type (whether personnel, infrastructure, or otherwise) to provide the Services, nor shall anything in this Agreement require Ikaria (or any Ikaria Group Member) to prioritize providing Services to R&DCo (or another R&DCo Group Member) over performing similar services for its own benefit in support of the Ikaria Business.

1.2 Level of Services. Subject to Section 1.1, the Services shall be provided to R&DCo (or another R&DCo Group Member) in a manner substantially similar in scope, quality, and nature to those provided to, or provided on behalf of, the R&DCo Business prior to the date hereof.

1.3 Cooperation. Each Party shall cause its employees and the employees of its respective Group Members to reasonably cooperate with employees of the other Party and such other Party's Group Members to the extent required for effective delivery of the Services. In addition, each Party shall name a point of contact who shall be responsible for the day-to-day implementation of this Agreement (each such person, a "Service Coordinator"), including attempted resolution of any issues that may arise during the performance of any of Party's obligations hereunder pursuant to the dispute resolution provisions referenced in Section 8.1.

1.4 Third Party Services. Ikaria shall have the right to engage the services of independent contractors to deliver some or all of the Services, or assist Ikaria in the delivery of Services, contemplated under this Agreement. If Ikaria utilizes independent contractors to deliver or assist Ikaria in the delivery of any Services, Ikaria will impose on such third parties the confidentiality obligations specified in this Agreement and will use commercially reasonable efforts to supervise the performance of such third parties to ensure that the Services meet the requirements of this Agreement.

1.5 Access.

(a) R&DCo shall, and shall cause the other R&DCo Group Members to, permit the Representatives of Ikaria to have access (during normal business hours upon reasonable advance notice and in a manner so as not to interfere with the conduct of the R&DCo Business) to the information, personnel, equipment, office and storage space and Systems (as defined in Section 1.5(b) below) required for Ikaria to provide the Services. Notwithstanding the foregoing, neither R&DCo nor any other R&DCo Group Member shall be obligated to provide any information, documents or access to any Person other than Ikaria or another Ikaria Group Member unless Ikaria is responsible for the use and disclosure of any information obtained by such Person from R&DCo or such other R&DCo Group Member, and such Person is subject to confidentiality obligations with Ikaria consistent with Article 4 of the Separation Agreement. Further, neither R&DCo nor any other R&DCo Group Member shall be obligated to provide (i) any Restricted Information, (ii) any information or access that would result in the disclosure of any information of R&DCo or any of its Affiliates unrelated to the Services (and R&DCo and the R&DCo Group Members shall be permitted to redact any such information from any materials provided to Ikaria or its Representatives) or (iii) any consolidated, combined, affiliated, or unitary Tax return that includes R&DCo or any of its Affiliates or any Tax-related work papers. Notwithstanding the foregoing, in the event that R&DCo or another R&DCo Group Member elects not to provide information, documents or access to Ikaria or its Representatives in accordance with this Section 1.5(a), Ikaria shall not be obligated to provide any Service to R&DCo or such other R&DCo Group Member that cannot reasonably be provided without such information, documents or access. Ikaria shall cause all of its Representatives, when on the premises of R&DCo or another R&DCo Group Member or when given access to any information, personnel, equipment,

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office and storage space and Systems, to conform to the policies and procedures of R&DCo or such other R&DCo Group Member concerning health, safety and security which are made known to Ikaria in advance in writing.

(b) System Security. If either Party is given access to the other Party's (or the other Party's Group Members') computer system(s), facilities, networks (including voice or data networks), software, or other information technology assets (collectively, "Systems") in connection with performance or transition of the Services, such Party shall comply with all security regulations and other policies and procedures reasonably required by the other Party (or such other Party's Group Members) from time to time which are made known to such Party in advance in writing ("Regulations"), and will not intentionally tamper with, compromise or circumvent any security, privacy or audit measures that are employed by the other Party (or such other Party's Group Members) and which are made known to such Party in advance in writing. The Representatives of the Party being granted access to the other Party's (or such other Party's Group Members') Systems may be required to execute a reasonable, separate system access agreement for individuals who are to have access to such Systems. The Party being granted such access shall ensure that only those users who are specifically authorized by the other Party (or such other Party's Group Members) to gain access to the other Party's (or such other Party's Group Members') Systems as necessary to utilize or provide the Services, as applicable, gain such access. Each Party shall be responsible for all acts and omissions of its Representatives. If at any time a Party determines that any Representative of either Party (or a Party's Group Members) has sought to circumvent or has circumvented the other Party's (or the other Party's Group Members') Regulations or other security, privacy or audit measures or that an unauthorized person has accessed or may access the other Party's (or such other Party's Group Members') Systems or a person has engaged in activities that may lead to the unauthorized access, destruction or alteration or loss of data, information or software, the determining Party shall promptly notify the other and the other Party shall have the right to immediately terminate any such person's access to such Party's (or such Party's Group Members') Systems.

1.6 Independent Contractor. For all purposes hereof, each Party shall at all times act as an independent contractor and shall have no authority to represent the other Party or any of the other Party's Group Members in any way or otherwise be deemed an agent, lawyer, employee, representative, joint venturer or fiduciary of such other Party or such other Party's Group Members, nor shall this Agreement or the transactions contemplated hereby be deemed to create any joint venture between the Parties or any of their respective Group Members. Each Party shall not declare or represent to any third party that such Party shall have any power or authority to negotiate or conclude any agreement, or to make any representation or to give any undertaking on behalf of the other Party or any of the other Party's Group Members in any way whatsoever.

1.7 Changes in Law. If either Party becomes aware of a change in applicable Law affecting compliance of the Services with such Law, such Party shall provide notice to the other Party and the Parties shall discuss in good faith (including, if necessary, discussion between each Party's legal counsel) any necessary modifications to the Services to achieve compliance. The Parties agree to use commercially reasonable efforts to identify a work-around solution that enables Ikaria to perform the Services in compliance with such modified Law. If the Parties cannot agree on any such work-around, then the Parties agree to use commercially reasonable efforts to (i) modify the applicable Services to comply with such changes in applicable Law and (ii) agree on the extent (if any) to which all or a portion of the fees and expenses of Ikaria arising from such modifications shall be borne by R&DCo; provided that Ikaria shall not be required to continue to provide the applicable Service that violates applicable Law as a result of such a change in such law or regulation, nor to modify such Service, except to the extent a modification would not result in Ikaria being required to incur any material out-of-pocket expenses. If compliance with applicable Law would result in Ikaria being required to incur any material out-of-pocket expenses, Ikaria shall not be required to continue to provide the applicable Service unless and until R&DCo and Ikaria agree on whether all or any portion of such fees and expenses shall be borne by R&DCo and, if R&DCo and Ikaria agree on the amount (if any) to be borne by R&DCo, Ikaria shall promptly implement the modifications necessary to comply with such changes in applicable Law.

1.8 Additional Services. If requested by R&DCo, Ikaria may provide services in addition to the Services to R&DCo or another R&DCo Group Member. The scope of any such services, as well as the prices and other terms applicable to such services, shall be as agreed in writing by R&DCo and Ikaria.

2. PAYMENTS

2.1 Services Pricing.

(a) In consideration of making the Services available to R&DCo and the other R&DCo Group Members under this Agreement, R&DCo shall pay Ikaria $772,000.00 per month (the "Service Cost"). The Parties agree that any pricing information set forth on Schedule A is for information purposes only and shall not affect the amount to be paid by R&DCo to Ikaria hereunder, and R&DCo acknowledges that the fees set forth in this Section 2.1(a) are due to Ikaria regardless of the frequency or quantity of Services actually utilized by R&DCo or the other R&DCo Group Members under this Agreement, and that all such fees are non-refundable. In addition, R&DCo shall promptly reimburse Ikaria for any and all out-of-pocket expenses incurred in connection with the provision of Services hereunder, and if performing the Services requires resources outside of the existing resources of Ikaria or otherwise interferes with the ordinary operations of the Ikaria Business (in either case, the "Extraordinary Services"), then R&DCo shall pay, in addition to the Service Cost, the costs and expenses incurred by Ikaria in connection with performing such Extraordinary Services.

(b) Concurrently with the execution of this Agreement, the Parties are entering into an escrow agreement with a third party escrow agent (the "Escrow Agreement"). Pursuant to the terms of the Escrow Agreement, concurrently with the execution of this Agreement R&DCo shall deposit $18,528,000 into escrow, and Ikaria shall have the right to withdraw the monthly Service Cost from such escrow on a monthly basis in arrears.

2.2 Invoicing and Payment. Within 30 days following the end of each calendar month during the term hereof, Ikaria shall provide to R&DCo an invoice setting forth any out-of-pocket expenses, Tax costs and any expenses related to the provision of Extraordinary Services incurred by Ikaria hereunder during such month (each, an "Invoice"). Each Invoice shall contain a brief description of such out-of-pocket expenses, Tax costs and/or Extraordinary Services expenses, including a listing of any third party charges included therein. R&DCo shall pay all amounts due under each Invoice no later than 30 days following receipt of an Invoice. Any Invoices not paid when due shall bear interest from the due date at the rate of the lesser of (a) [**] percent ([**]%) per month or (b) the maximum amount permitted by applicable law. R&DCo agrees to pay on demand all costs of collection, including reasonable attorneys' fees, incurred by Ikaria in collecting any such Invoice.

2.3 Taxes. Any federal, state, municipal, or other U.S. or foreign government taxes, duties, excises, tariffs, fees, assessments or levies now or hereinafter imposed on the performance or delivery of Services or direct costs (other than income taxes imposed on Ikaria) shall be paid by R&DCo to Ikaria in addition to the other fees payable pursuant to this Article 2 (the "Tax Costs"). For the avoidance of doubt, liability for the payment and remittance of any taxes, duties, excises, tariffs, fees, assessments or levies imposed with respect to the performance or delivery of Services or direct costs hereunder shall be the responsibility of Ikaria.

2.4 Records. Ikaria and R&DCo shall, and each shall cause its respective Group Members to, keep such full and adequate records as are necessary to determine the charges to be assessed pursuant to this Section 2, and shall have reasonable access to such records and any other records or other information relevant to the provision of Services hereunder in accordance with the provisions of Article 4 of the Separation Agreement (including, for the avoidance of doubt, the access to Information covenant set forth in Section 4.3 of the Separation Agreement, regardless of whether such records were created prior to the Distribution Date).

3. HAMPTON FACILITY

Subject to the provisions hereof and Schedule B attached hereto, Ikaria shall use commercially reasonable efforts to obtain the right to allow the employees of the R&DCo Group Members to remain in Ikaria's Hampton, New Jersey facility (the "Facility") for the continued operation of the R&DCo Business. R&DCo shall be solely responsible for, and shall promptly pay, any fees, costs, expenses, or other amounts (including any additional security deposit) incurred by Ikaria to obtain such right or required on a one-time or ongoing basis in respect of use of the Facility by any R&DCo Group Member.

4. SOFTWARE LICENSES AND OTHER CONSENTS

Except as provided in the Separation Agreement or the R&D Cross-License Agreement, Ikaria and its Group Members shall not be required to transfer or assign to R&DCo or another R&DCo Group Member any assets, including third-party software licenses, data, data subscriptions, or any software or hardware or other technology assets owned by Ikaria or any of its Group Members in connection with the provision of the Services, and no licenses, express or implied, are granted hereunder unless expressly set forth herein. Notwithstanding the foregoing, Ikaria shall use commercially reasonable efforts to obtain any waivers, permits, consents, licenses or sublicenses required for the provision of the Services to R&DCo or another R&DCo Group Member under the terms of any third-party software license, data subscription or other agreement necessary to provide such Services (each, a "Consent"); provided that, notwithstanding any other provision of this Agreement, Ikaria shall have no obligation to provide that part of the Service hereunder if it is unable, after using commercially reasonable efforts, to (i) obtain any Consent therefor, or (ii) provide that part of the Service or procure the provision of an equivalent service to R&DCo or another R&DCo Group Member, as applicable. Any and all out-of-pocket costs and expenses incurred by Ikaria associated with obtaining or soliciting Consents (including, (a) fees and other out-of-pocket expenses incurred by Ikaria in connection with obtaining or soliciting the consent of any third party vendors and (b) in the event any Consent is not obtained, out-of-pocket costs and expenses incurred by Ikaria, using commercially reasonable efforts, in connection with providing an alternate method of delivering any Service) shall be paid by R&DCo. Notwithstanding the foregoing, in the event a Consent is required but not obtained, the Parties agree to use commercially reasonable efforts to identify a work-around solution that enables Ikaria to perform the Services without such Consent; provided that the foregoing shall continue to apply if the Parties cannot agree on any such work-around. At Ikaria's reasonable request and at R&DCo's cost and expense, R&DCo shall, and shall cause the other R&DCo Group Members to, cooperate with and assist Ikaria in obtaining or soliciting any Consent hereunder.

5. CONFIDENTIALITY

Confidential Information. For purposes of this Agreement, "Confidential Information" shall mean all information disclosed by either Party or its respective Group Members to the other in connection with this Agreement, whether orally, visually, in writing or in any other tangible

form, and includes, but is not limited to, economic, scientific, technical, product and business data, business plans, and the like. Confidential Information shall be treated as "Information" and shall be subject to the provisions of Article 4 of the Separation Agreement (regardless of the date on which such information was created); provided that the obligation to keep such information confidential pursuant to Section 4.1 of the Separation Agreement shall continue for five (5) years after the termination of this Agreement in accordance with its terms.

6. INDEMNIFICATION

6.1 Indemnification by R&DCo Group. R&DCo shall, and shall cause each other R&DCo Group Member receiving Services hereunder to, indemnify and hold harmless each Ikaria Indemnified Party from and against all Damages incurred by such Ikaria Indemnified Party arising from the provision of Services by Ikaria or any other Ikaria Group Member hereunder, except as set forth in Section 6.2.

6.2 Indemnification by Ikaria Group. Ikaria shall, and shall cause each other Ikaria Group Member providing Services hereunder to, indemnify and hold harmless each R&DCo Indemnified Party from and against all Damages incurred by such R&DCo Indemnified Party arising from gross negligence or willful misconduct by Ikaria or any other Ikaria Group Member or any of Ikaria's or such Ikaria Group Member's employees in providing Services hereunder, except to the extent that such employees were acting in accordance with specific written instructions from R&DCo or any other R&DCo Group Member.

6.3 Procedures for Third Party Claims. The Parties shall follow the applicable procedures set forth in Section 6.3(d) of the Separation Agreement with respect to any indemnified claims.

6.4 Limitations of Liability.

(a) THE LIABILITY OF THE IKARIA GROUP MEMBERS IN CONNECTION WITH THE PERFORMANCE, DELIVERY OR PROVISION OF ANY SERVICE OR OTHERWISE UNDER THIS AGREEMENT SHALL BE LIMITED TO A SUM EQUAL TO THE TOTAL SERVICE COST PAID HEREUNDER TO THE IKARIA GROUP MEMBERS.

(b) NOTWITHSTANDING ANY OTHER PROVISION OF THIS AGREEMENT TO THE CONTRARY, IN NO EVENT SHALL EITHER PARTY OR ANY OF ITS GROUP MEMBERS BE LIABLE FOR ANY SPECIAL, INCIDENTAL, INDIRECT, COLLATERAL, CONSEQUENTIAL OR PUNITIVE DAMAGES OR LOST PROFITS SUFFERED BY AN INDEMNIFIED PARTY, HOWEVER CAUSED AND ON ANY THEORY OF LIABILITY, IN CONNECTION WITH ANY DAMAGES ARISING HEREUNDER OR THEREUNDER; PROVIDED, HOWEVER, THAT TO THE EXTENT AN INDEMNIFIED PARTY IS REQUIRED TO PAY ANY SPECIAL, INCIDENTAL, INDIRECT, COLLATERAL, CONSEQUENTIAL OR PUNITIVE DAMAGES OR LOST

PROFITS TO A PERSON WHO IS NOT A MEMBER OF EITHER GROUP IN CONNECTION WITH A THIRD-PARTY CLAIM, SUCH DAMAGES SHALL CONSTITUTE DIRECT DAMAGES AND NOT SUBJECT TO THE LIMITATION SET FORTH IN THIS SECTION 6.4(b).

(c) THE SERVICES ARE PROVIDED "AS IS" AND, TO THE FULLEST EXTENT OF THE LAW, PROVIDED WITHOUT WARRANTIES, CLAIMS OR REPRESENTATIONS MADE BY IKARIA, EITHER EXPRESS, IMPLIED, OR STATUTORY, INCLUDING, BUT NOT LIMITED TO, WARRANTIES OF QUALITY, PERFORMANCE, NON-INFRINGEMENT, MERCHANTABILITY, OR FITNESS FOR A PARTICULAR PURPOSE, NOR ARE THERE ANY WARRANTIES CREATED BY COURSE OF DEALING, COURSE OF PERFORMANCE, OR TRADE USAGE.

(d) Nothing contained in this Agreement shall limit or alter (i) the obligation of either Party to indemnify the other Party pursuant to the Separation Agreement or any other Ancillary Document or (ii) the right of either Party to make a claim pursuant to the Separation Agreement or any other Ancillary Document; provided, that no Party shall obtain duplicative recoveries.

7. TERM AND TERMINATION

7.1 Term. Unless earlier terminated in accordance with Section 7.2 below, this Agreement shall be in effect until the second anniversary of the date hereof. Notwithstanding the foregoing, R&DCo shall, and shall cause the other R&DCo Group Members to, use commercially reasonable efforts to provide the Services for itself and the other R&DCo Group Members as soon as practicable (whether by hiring additional employees, retaining third party service providers, or otherwise).

7.2 Termination. This Agreement may be terminated by either Party if the other Party (the "Defaulting Party") has materially breached its obligations under this Agreement and if the Defaulting Party has not cured such default within thirty (30) days following the date on which the other Party (the "Notifying Party") has given written notice specifying the facts constituting the default. Notwithstanding the foregoing sentence, this Agreement shall not be terminated due to a default by the Defaulting Party if such default is directly attributable to a breach of this Agreement by the Notifying Party.

7.3 Effect of Termination. Upon termination of this Agreement for any reason, all rights and obligations of the Parties under this Agreement shall cease and be of no further force or effect, except that the provisions of Section 1.6, the first sentence of Section 4, Section 5 and Section 6 of this Agreement, and R&DCo's obligation to pay the Service Cost and any amounts pursuant to Section 2.1 or Section 2.3, shall survive any such termination or expiration.

7.4 Further Actions. Following any termination of this Agreement, Ikaria shall cooperate in good faith with the R&DCo Group Members to transfer applicable records and take all other actions reasonably requested by the R&DCo Group Members to enable the R&DCo Group Members to make alternative arrangements for the provision of services substantially consistent with the Services provided pursuant to this Agreement.

8. GENERAL

8.1 Dispute Resolution. The dispute resolution procedures set forth in Article 7 of the Separation Agreement shall apply to all disputes, controversies or claims that may arise out of or relate to, or arise under or in connection with this Agreement or the transactions contemplated hereby.

8.2 Miscellaneous. The provisions of Sections 8.1, 8.2, 8.3, 8.4, 8.5, 8.6, 8.7, 8.8, 8.9, 8.10, 8.11, 8.12, 8.13 and 8.14 of the Separation Agreement shall apply *mutatis mutandis* to this Agreement, as if set forth in this Agreement in full.

8.3 Force Majeure. Ikaria shall not bear any responsibility or liability for any Damages arising out of any delay, inability to perform or interruption of its performance of its obligations under this Agreement due to any acts or omissions of the other party hereto or for events beyond its reasonable control including, without limitation, acts of God, acts of governmental authorities, acts of the public enemy or due to war, riot, flood, civil commotion, insurrection, labor difficulty, severe or adverse weather conditions, lack of or shortage of electrical power, malfunctions of equipment or software programs, or any other cause beyond the reasonable control of such party (each, a "Force Majeure Event"); provided, that Ikaria (a) as soon as reasonably practical following the occurrence of a Force Majeure Event, gives written notice to R&DCo of such event, including a description of the circumstances preventing its performance and of its plans and efforts to implement a work-around, and (b) uses reasonable best efforts to resume or restore performance as expeditiously as possible. The obligations of Ikaria seeking to be excused shall then be tolled for the duration of the Force Majeure Event to the extent that the Force Majeure Event prevents it from performing its obligations hereunder. R&DCo shall have no obligation to pay any fees or other amounts to Ikaria with respect to any Services that Ikaria is unable to provide hereunder for so long as Ikaria is unable to provide such Services in compliance with this Agreement.

[*Remainder of page intentionally left blank.*]

IN WITNESS WHEREOF, each of the Parties has caused this Transition Services Agreement to be executed on its behalf by a duly authorized officer on the date first set forth above.

IKARIA, INC., a Delaware corporation	BELLEROPHON THERAPEUTICS LLC, a Delaware limited liability company
By: /s/ Anastasios Konidaris	By: /s/ Daniel Tassé
Name: Anastasios Konidaris	Name: Daniel Tassé
Title: Chief Financial Officer	Title: Chief Executive Officer

SCHEDULE A

SERVICE	BILLING TYPE & RATE	END DATE (Subject to Section 7.1 of Agreement)
Human Resources Support [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Real Estate Support · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Information Technology Support · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Accounting & Tax Support · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Treasury Support [**] ***FP&A Support*** [**] ***Purchasing Support*** · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Management/Executive Services · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Legal Services · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Sub-Total Corporate Services	**Fixed fee for length of agreement of $ [**]per month + out of pocket expenses**	
Quality Services · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Regulatory Services · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Drug and Device Safety Services · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Business Development [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Biometrics · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Manufacturing · [**]	Fixed fee for length of agreement of: $[**] per month + out of pocket expenses	24 months from Effective Date
Sub-Total R&D Group & Manufacturing Support	**Fixed fee for length of agreement of $[**] per month + out of pocket expenses**	
Total	**Fixed fee for length of agreement of $772K per month + out of pocket expenses**	

SCHEDULE B

FACILITY PROVISIONS

1. **Right to Use the Facility.**

1.1 **Facility**. R&DCo Group Members shall have the right to use the Facility, subject to the provisions of this Agreement, including Section 3 and this Schedule B, and shall comply in all material respects with all obligations of the tenant under that certain Lease Agreement by and between Crown Perryville, LLC and INO Therapeutics LLC, dated July 9, 2008, as amended by the Lease Assignment and Assumption Agreement between INO Therapeutics LLC and Ikaria, dated October 24, 2010, as amended by the Amendment to Lease Agreement between Crown Perryville, LLC and Ikaria, dated October 24, 2010, and the Subordination, Non-Disturbance, and Attornment Agreement by and between Ikaria, Crown Perryville, LLC, and TD Bank, N.A., dated March 17, 2011 (the "Lease") as such obligations relate to the Facility and use by the R&DCo Group Members of the Common Areas (as defined below), including, without limitation, the obligation to maintain insurance.

1.2 **Common Areas**. The right of the R&DCo Group Members to use the Facility shall include a non-exclusive right to use such common areas as may exist with respect to the Facility to the extent such right has been granted to Ikaria under the Lease, which may include circulation corridors, stairwells, lobbies, library, cafeteria, clinic, restrooms and conference rooms, if any, parking areas and sidewalks, if any (collectively referred to as the "Common Areas").

2. **Use.**

2.1 **Changes to Facility.** R&DCo shall not, and shall cause the other R&DCo Group Members not to, make any alterations or improvements to the Facility without the prior written consent of Ikaria, which consent shall not be unreasonably withheld, conditioned or delayed. Failure of Ikaria's landlord to consent to or approve the alterations or improvements, where required, or non-compliance of alterations or improvements with the Lease, shall be a reasonable grounds for Ikaria to withhold consent under this Section.

2.2 **Ikaria Right to Alter.** Ikaria reserves the right, at any time, and from time to time, to make alterations, additions, repairs or improvements to or in any part of the premises adjoining the Facility, provided that any such alterations shall be conducted in a manner and at such times as shall not unreasonably affect use of the Facility by the R&DCo Group Members.

2.3 **Use of Facility.** R&DCo and the other R&DCo Group Members may use the Facility for the conduct of the R&DCo Business, but may not make any other use of the Facility without the prior written consent of Ikaria, which consent shall not be unreasonably withheld, conditioned or delayed.

3. **Maintenance; Compliance with Laws.**

3.1 **Maintenance.** R&DCo shall not, and shall cause the other R&DCo Group Members not to, cause or permit any damage to the Facility and shall maintain the Facility in a clean, safe and sanitary condition, reasonable wear and tear and damage caused by any casualty excluded and otherwise in accordance with the terms of the Lease. R&DCo shall not, and shall cause the other R&DCo Group Members not to, permit or suffer any injury, waste or nuisance in or to the Facility.

3.2 **Compliance With Laws.** R&DCo shall, and shall cause the other R&DCo Group Members to, comply with all applicable Laws relating to the R&DCo Group's use or occupation of the Facility, including any Environmental Laws and the New Jersey Industrial Site Recovery Act (N.J.S.A. 13:1K-6 et seq.) ("ISRA") and shall be responsible for any environmental liabilities relating to, arising out of, or resulting from the R&DCo Group Members' use or occupation of the Facility.

4. **Utilities and Services.**

4.1 **Utilities and Services.** Ikaria shall use reasonable efforts to cause the landlord under the Lease to furnish to or for the benefit of the Facility the utilities and services that the landlord is obligated to provide under the Lease.

5. **Cancellation of Lease.**

5.1 **Cancellation of Lease**. In the event of the cancellation or termination of the Lease for any reason whatsoever or of the involuntary surrender of the Lease by operation of law prior to the expiration date of this Agreement, the rights of the R&DCo Group Members under this Agreement as to the Facility and Common Areas shall terminate.

Exhibit 10.22

Bellerophon Services Inc.

February 3, 2014

[ ]

Dear ,

At Bellerophon Services Inc. ("Bellerophon"), we recognize our success is the result of the hard work and unique expertise of professionals like you. Retaining the right people is essential to our ongoing ability to achieve our business objectives and succeed in the long-term. The following retention bonus is being offered to you in recognition of both your past contributions and our anticipation of future achievements. The terms of your retention bonus are as follow:

1) You will be entitled to receive a one-time payment of $, less applicable taxes and other withholding if you remain an active employee of Bellerophon in good standing on December 19, 2014 (the "Bonus")

2) The Bonus would be paid within 30 days after December 19, 2014.

3) You continue to meet or exceed any performance criteria established for your position.

By signing and returning this letter, you agree to hold all information relating to the Bonus (including, without limitation, the existence of this letter) in strict confidence, and to not disclose any information relating hereto to any party other than your financial or legal advisors.

You acknowledge and agree that you will be, and will remain, an employee at-will of Bellerophon, and that nothing in this letter is intended to be or create an obligation of Bellerophon to employ you for any specific period of time.

Our offer to provide the Bonus is contingent upon you signing and returning this letter. You will not be eligible to receive the Bonus unless you have done so.

We appreciate the commitment you have made to ensure the patients and healthcare providers that may someday rely on our products can do so with confidence. I thank you in advance for your ongoing service.

Sincerely,

The Leadership Team
Bellerophon Services Inc.

Employee Signature	Date

Cc: File
Human Resources

Exhibit 21.1

SUBSIDIARIES OF
BELLEROPHON THERAPEUTICS LLC

NAME OF SUBSIDIARY	JURISDICTION OF ORGANIZATION
Bellerophon Services, Inc.	Delaware
Bellerophon Pulse Technologies LLC	Delaware
Bellerophon BCM LLC	Delaware