# Bellerophon Therapeutics

Company Presentation | July 2020

Bellerophon THERAPEUTICS

# Forward Looking Statements

This presentation contains forward-looking statements that involve substantial risks and uncertainties. All statements, other than statements of historical facts, contained in this presentation, 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," "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. 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 due to a number of important factors, including risks and uncertainties relating to: INOpulse® not proving to be an effective treatment for COVID-19 or approved for marketing by the FDA, the timing and outcomes of our ongoing and expected clinical trials for our product candidates; our ability to successfully develop, commercialize and market any of our product candidates; our ability to obtain, maintain and enforce intellectual property rights; competition; our reliance on third parties; our ability to obtain necessary financing; and those risk factors discussed in the "Risk Factors" section and elsewhere in our most recent Form 10-K and other periodic filings we make with the SEC.

All forward-looking statements contained in this presentation reflect our current views with respect to future events. We assume no obligation, except as required by applicable law, to update any forward-looking statements publicly, or to update the reasons why actual results could differ materially from those anticipated in the forward-looking statements, even if new information becomes available in the future.

# Bellerophon Therapeutics (BLPH) Company Profile

## Clinical-Stage Biotherapeutics Company

- Focused on developing inhaled nitric oxide (iNO) based therapies for outpatient management of serious cardiopulmonary and infectious diseases
- Portable, lightweight delivery system (INOpulse®) allows for chronic home use
- Simplified regulatory approval pathway via existing nitric oxide NDA
- Company spun-off from Ikaria

## Novel Therapy Addressing Unmet Medical Needs

- Multiple late stage programs in pulmonary hypertension associated with underlying lung disease (WHO Group 3 & WHO Group 5)
- Novel targeted vasodilation provides potential for first approved therapy in intended indications
- Compelling anti-microbial and anti-viral properties
- Granted emergency expanded access followed by FDA clearance for and initiation of Phase 3 trial for the use of INOpulse in COVID-19

## Financial Summary

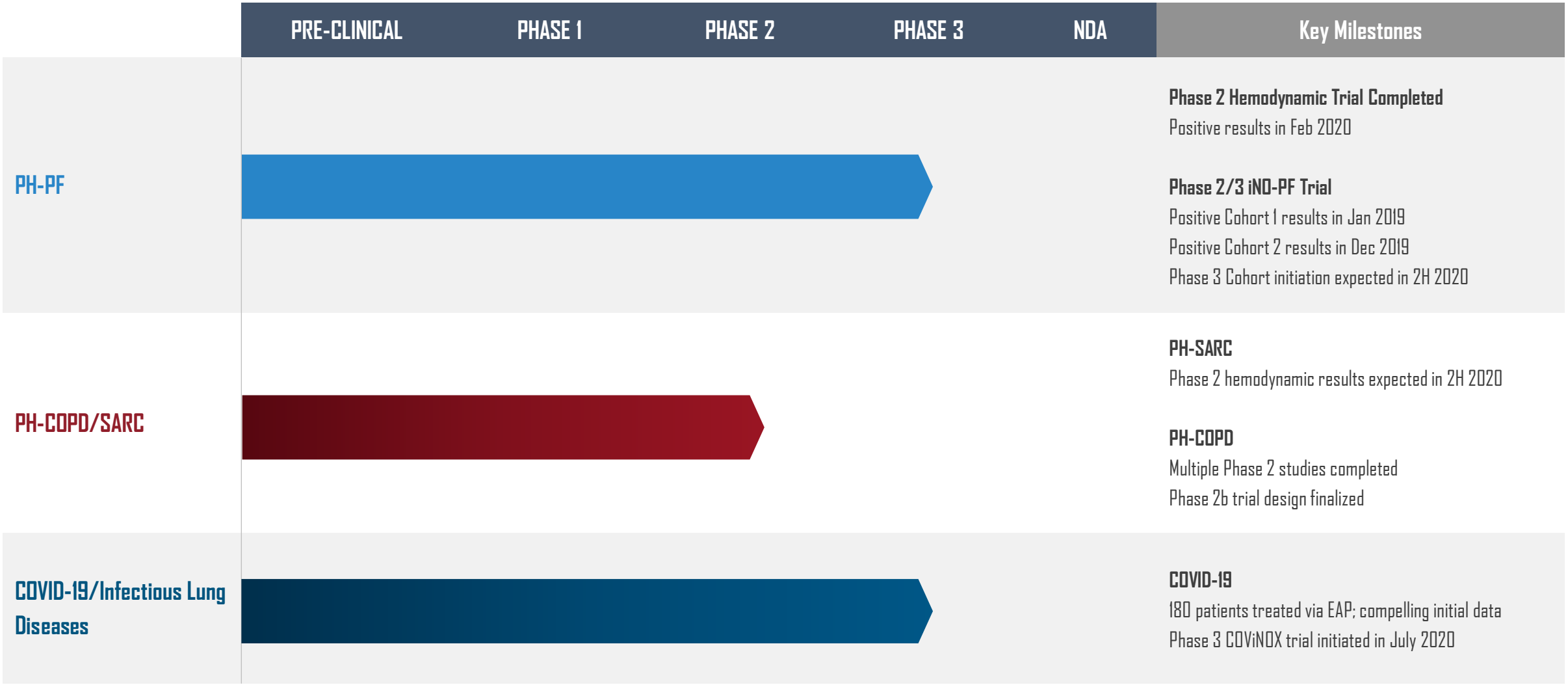
- Cash & Equivalents: $63.1[1,2]
- No Debt[1]
- Fully Diluted Shares Outstanding = 12.2 million

1. As of March 31, 2020
2. Includes $54.5M of net proceeds from public offerings and registered direct offerings that closed in Q2/2020

# Highly Experienced Leadership Team

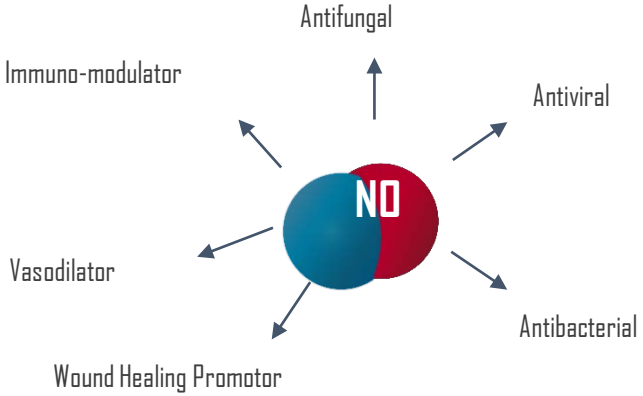
| | | |
|---|---|---|
| **Jonathan Peacock**<br>Chairman | **10 years** experience as CFO at Amgen and Novartis Pharma | AMGEN · McKinsey&Company · NOVARTIS · pwc |
| **Fabian Tenenbaum**<br>Chief Executive Officer | **15 years** of executive-level experience in finance, BD and operations | anterios · Unilever · SYNERON CANDELA |
| **Hunter Gillies, M.D.**<br>Chief Medical Officer | **20 years** experience in clinical research specializing in cardiometabolic and pulmonary vascular diseases | Pfizer · GILEAD · ACTELION |
| **Peter Fernandes**<br>Chief Regulatory & Safety Officer | **25 years** experience in global regulatory affairs specializing in respiratory products | IKARIA · NOVARTIS · Boehringer Ingelheim |
| **Assaf Korner**<br>Chief Financial Officer | **18 years** of financial experience in medical device and consumer product companies | SYNERON CANDELA · KPMG · Unilever |
| **Parag Shah, Ph.D.**<br>VP, Business Operations | **15 years** experience in pharmaceutical product development | IKARIA · Pfizer |
| **Amy Edmonds**<br>VP, Clinical Operations & Administration | **20 years** experience global clinical operations and training | IKARIA · Pfizer · Celgene |
| **Martin Dekker**<br>VP, Device Engineering & Manufacturing | **17 years** experience in new product development and launch | SPACELABS HEALTHCARE |

# Development Pipeline



# INOpulse Delivery System Overview

## Portable Delivery System Allows Chronic iNO Therapy

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INOpulse®

### Nitric Oxide

Well-established vasodilator approved for acute treatment of persistent pulmonary hypertension in hospitals

Broad spectrum antiviral that plays a key role in viral replication

### Portable pulsatile iNO delivery system

Proprietary algorithm ensures accurate dosing independent of patient's breath rate and tidal volume

Portable stand-alone system allows for out-patient treatment

### Novel drug-device combination therapy with multiple mechanisms of action

Targeted pulmonary vasodilation

Ventilation/Perfusion (V/Q) matching

Improved oxygen saturation

Antiviral potential

# INOpulse Delivery System

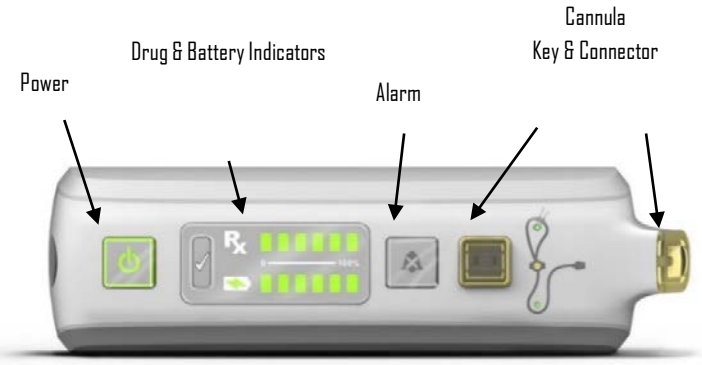
## Lightweight, Portable and User Friendly

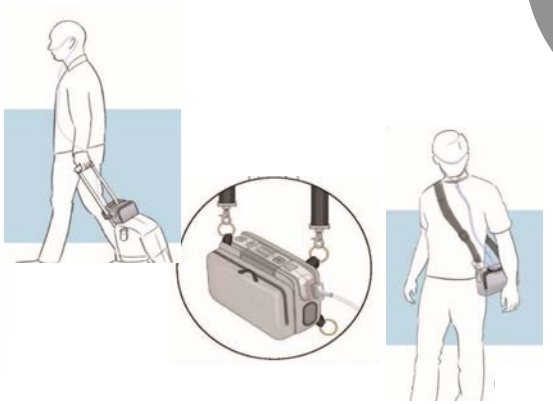
01 Swing engagement with drug cartridge

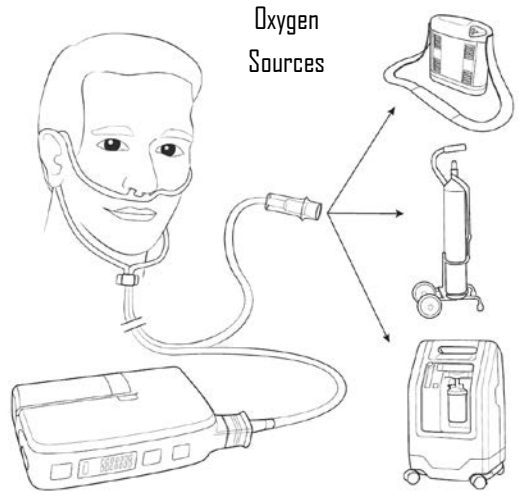
02 Intuitive and simple user interface

03 Tri-lumen cannula allows direct connection with oxygen

04 Lightweight portable design allows ease of transport





# Pulsatile Inhaled Nitric Oxide: Potential Treatment Option for COVID-19

## Nitric Oxide Plays a Key Role in Suppressing Viral Replication

- Naturally produced immune response that is upregulated by macrophages as a defense mechanism against pathogen infections

## Nitric oxide has demonstrated potential benefit for SARS Coronavirus (SARS-CoV), which is 80% genetically similar to COVID-19

- NO reduces viral load and replication in SARS-CoV infected cells[1] and improves survival of SARS-CoV infected cells[2]
- Inhaled NO improves oxygen saturation (and less FiO2 required) and reduces the need for assisted ventilation in SARS-CoV patients[3]

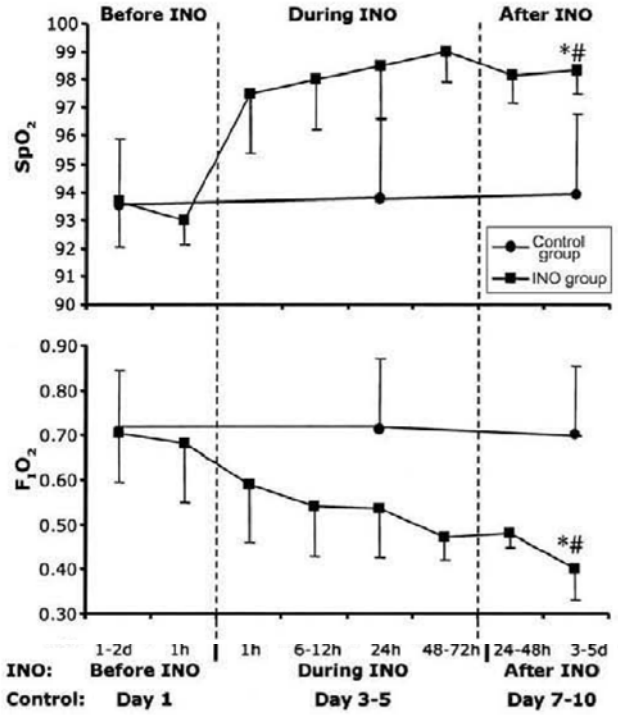
## INOpulse has potential to address a significant unmet need in the treatment of COVID-19 patients

- Overall hospital mortality is approximately 15 - 20%, increasing to 40% when admitted to the ICU[4]
- iNO has the potential to attenuate hypoxemia and reduce the need for assisted ventilation with the associated poor outcome and impact on hospital resources
- INOpulse delivery system is designed for outpatient use, which may be critical to preventing the further spread and alleviating the impact on hospitals and ICUs

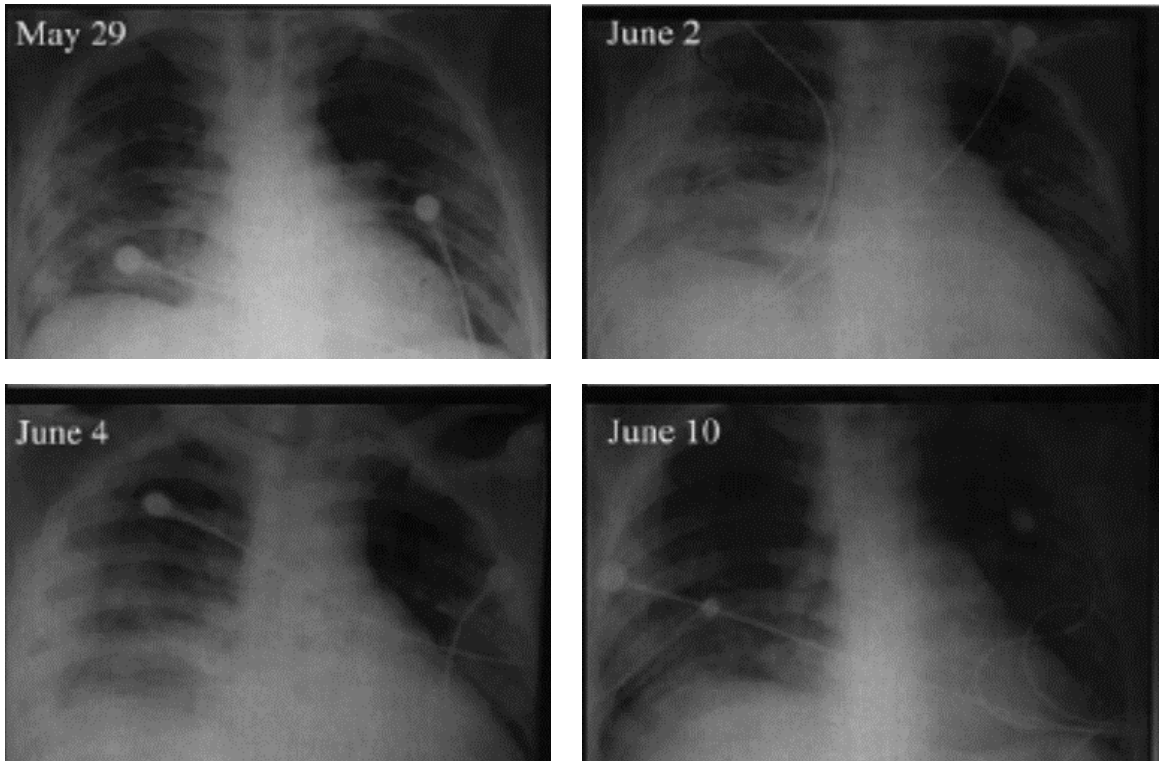

1. S. Akerstrom et al., J. of Virology, 79, 2005, 1966-1969.
2. E. Keyaerts et al., Int. J. Infect. Dis. 2004 Jul;8(4):223-6.
3. L. Chen et al., Clinical Infectious Disease, 39, 2004, 1531-1535
4. W. Joost Wiersinga et al., JAMA, online July 10, 2020.

# Inhaled NO Provides Benefit in Patients Infected with SARS-CoV

## iNO improves arterial oxygenation and reduces pneumonia infiltrates in SARS patients




Patients demonstrated improved arterial oxygenation and reduced need for ventilation support



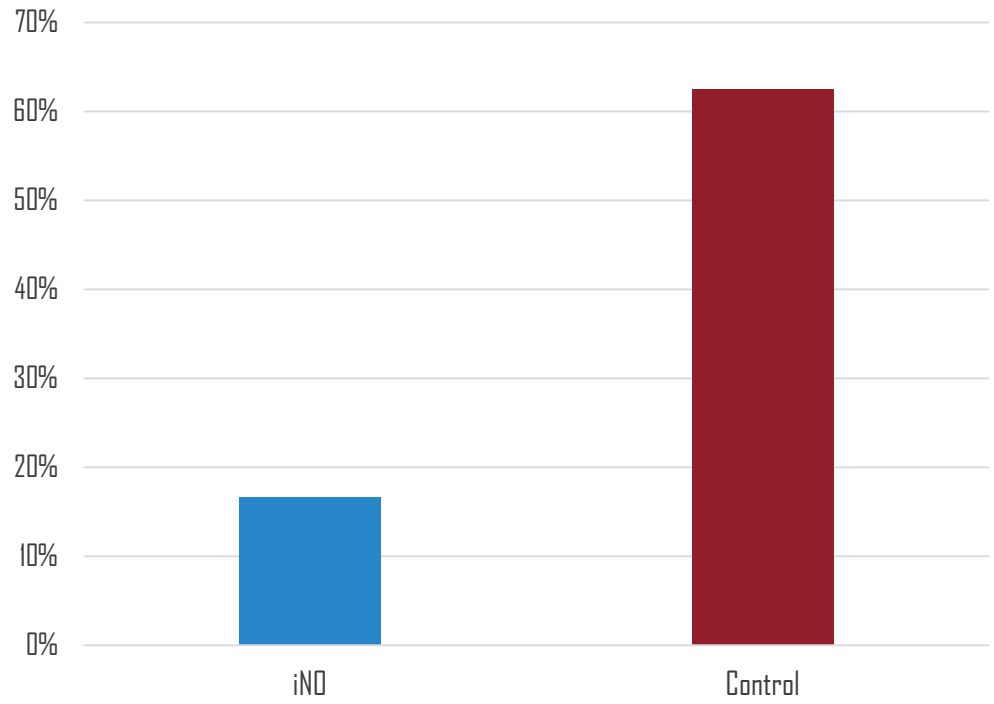

Progression of pneumonia (May 29 - June 2); Effect of iNO therapy demonstrating a decrease in the pneumonia infiltrates (June 4 - 10)

L. Chen et al., Clinical Infectious Disease, 39, 2004, 1531-1535

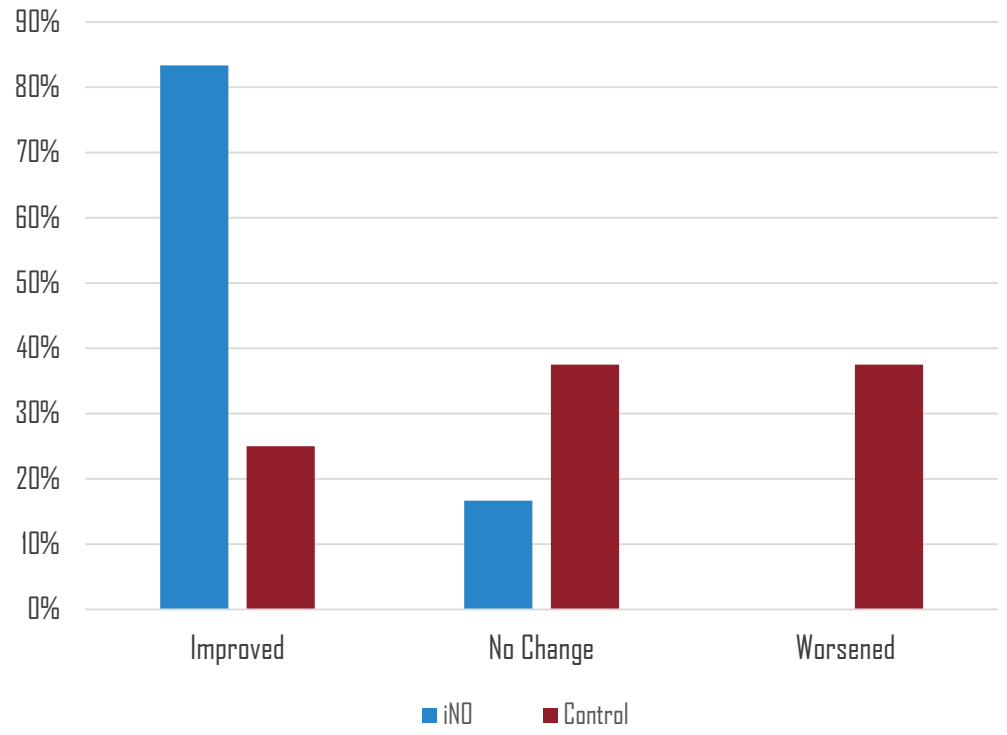
# Inhaled NO Provides Benefit in Patients Infected with SARS-CoV

## iNO reduces need for mechanical ventilation post-treatment and improves chest x-ray

### Need For Ventilatory Support Post-Treatment



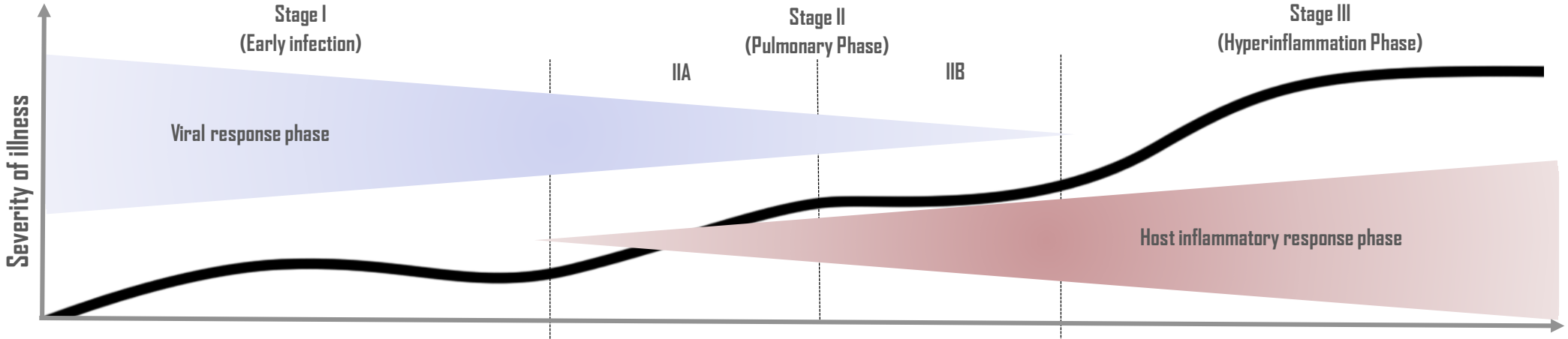

### Chest X-Ray




L. Chen et al., Clinical Infectious Disease, 39, 2004, 1531-1535

# COVID-19 Treatment Paradigm

## Inhaled Nitric Oxide: Potential Therapy for Stage II Patients




| | | | |
|---|---|---|---|
| **Clinical Symptoms** | Mild constitutional symptoms<br>Fever > 99.6ºF<br>Dry Cough, diarrhea, headache | Shortness of Breath<br>Hypoxia (PaO2/FiO2≤300mmHg) | ARDS<br>SIRS/Shock<br>Cardiac Failure |
| **Clinical Signs** | Lymphopenia, increased prothrombin time, increased D-Dimer and LDH (mild) | Abnormal chest imaging<br>Transaminitis<br>Low-normal procalcitonin | Elevated inflammatory markers (CRP, LDH, IL-6, D-dimer, ferritin)<br>Troponin, NT-proBNP elevation |
| **Potential Therapies** | INOpulse  | | |
| | Remdesivir, chloroquine, hydroxychloroquine, convalescent plasma transfusions | | |
| | Reduce immunosuppression | Corticosteroids, human immunoglobulin, IL-6 inhibitors, IL-2 inhibitors, JAK inhibitors | |

Image recreated from H. Siddiqi et al., COVID-19 Illness in Native and Immunosuppressed States: A Clinical-Therapeutic Staging Proposal, J. of Heart and Lung Transplantation, 2020, DOI: 10.1016/j.healun.2020.03.012

# Next Steps in INOpulse for the Treatment of COVID-19

## FDA granted emergency expanded access for pulsatile inhaled nitric oxide for treating COVID-19

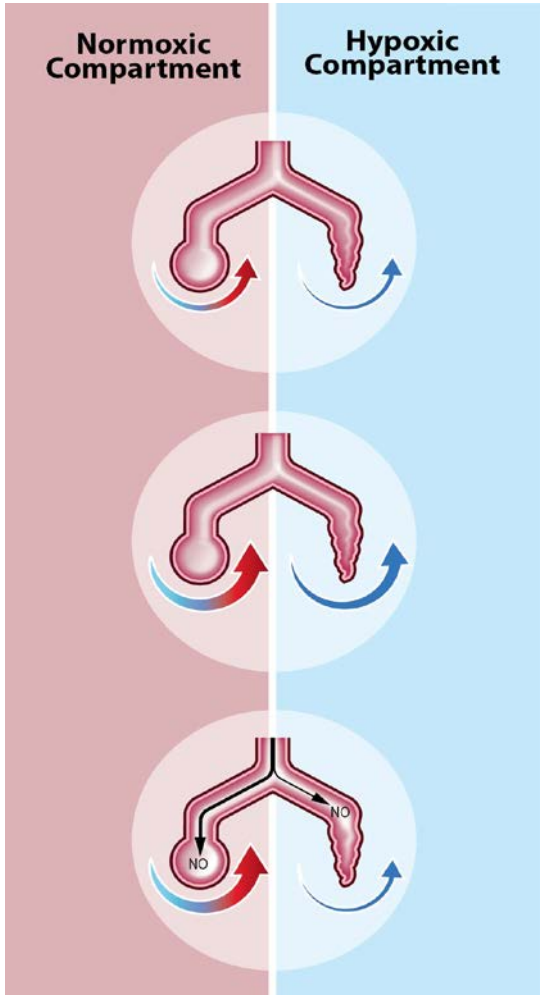
- 180 patients treated at 18 hospitals across the US under emergency expanded access
- **Preliminary data demonstrated recovery rate of 73.0% and mortality rate of 6.3% at day 14 from treatment initiation**
- INOpulse was well-tolerated with no safety concerns related to the therapy

## COViNOX: Phase 3 randomized, placebo-controlled study initiated in July 2020

COViNOX is an adaptive study utilizing a mortality/morbidity endpoint and designed to serve as a registrational trial for approval

- **Study Population:** Up to 500 patients diagnosed with COVID-19 who require supplemental oxygen
- **Primary Endpoint:** Proportion of subjects that had respiratory failure or mortality
- **Study Funding:** Applied for federal funding via BARDA and NIH

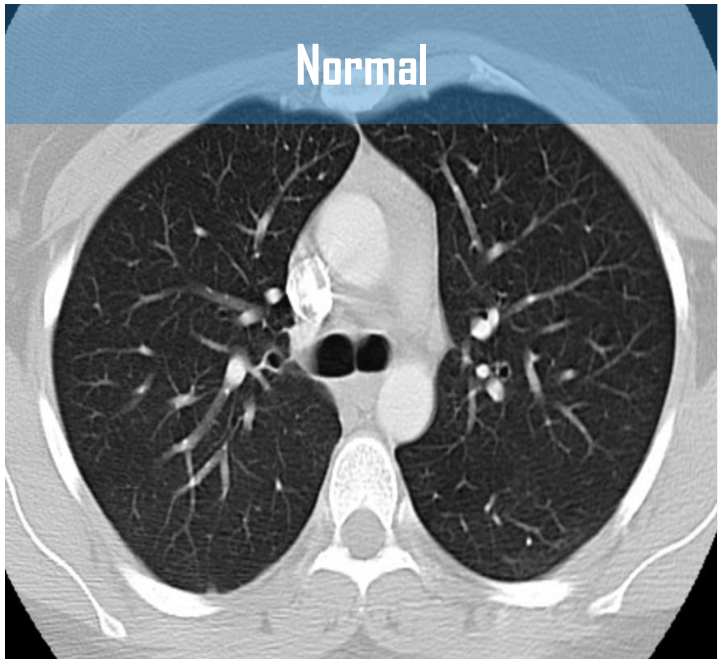
# INOpulse Provides a Unique and Differentiating Mechanism of Action

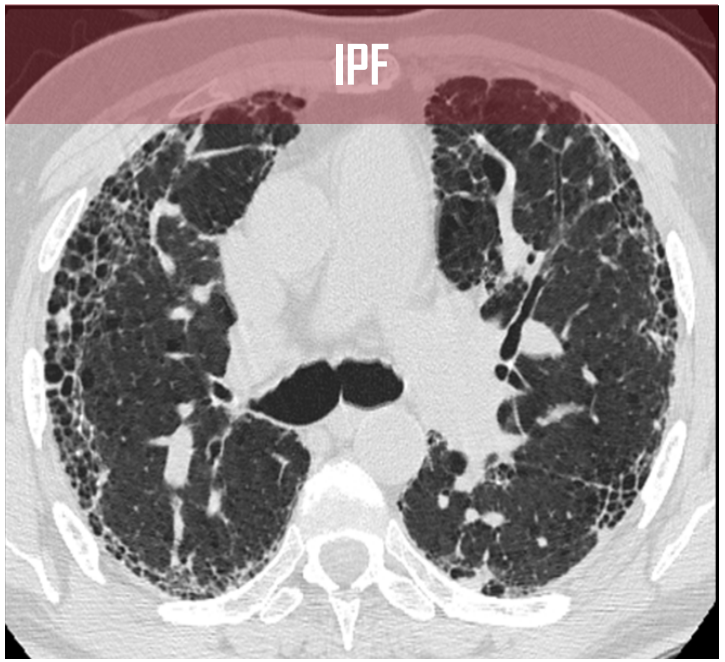



| | |
|---|---|
| **Baseline** | Hypoxic pulmonary vasoconstriction prevents oxygen desaturation |
| **Systemic Vasodilators** | Systemic vasodilators can reverse hypoxic vasoconstriction leading to ventilation/perfusion (V/Q) mismatch and arterial 02 desaturation |
| **INOpulse** | Providing iNO early in the inspiratory phase allows for targeted vasodilation of only the well ventilated alveoli thereby preventing V/Q mismatch and 02 desaturation |

# Pulmonary Fibrosis (PH-PF)

## A Significant Unmet Medical Need

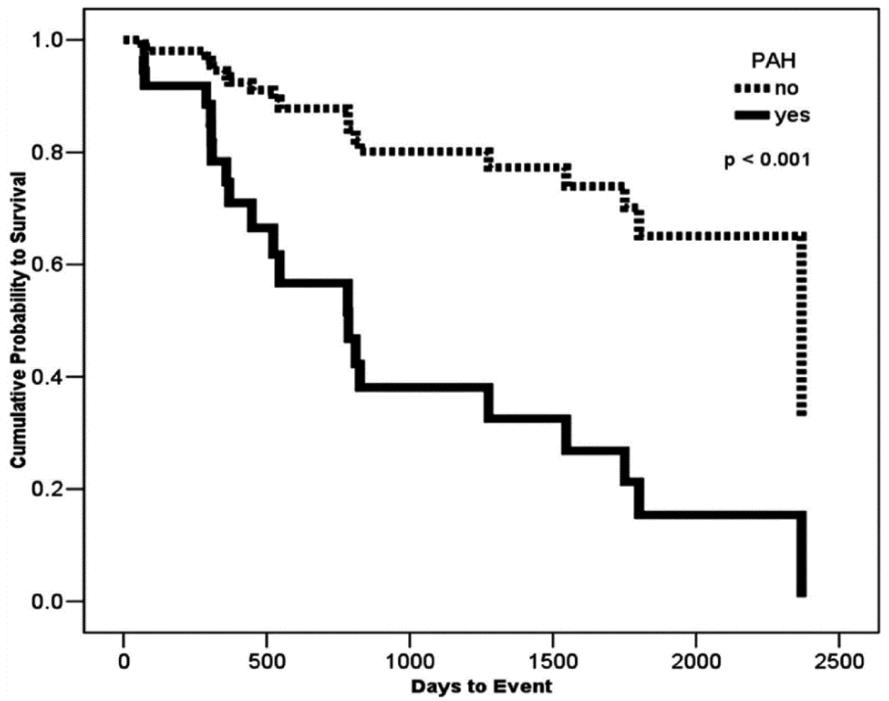



- Pulmonary Fibrosis (PF) is a broad category of diffuse lung diseases characterized by variable amounts of inflammation and fibrosis
- Idiopathic Pulmonary Fibrosis (IPF) is the largest and most serious of these diseases
- Patients with PF have thickening and scarring of the air sacs in the lungs, and often require supplemental oxygen to maintain adequate oxygen saturation

# PH-PF Significantly Reduces Survival

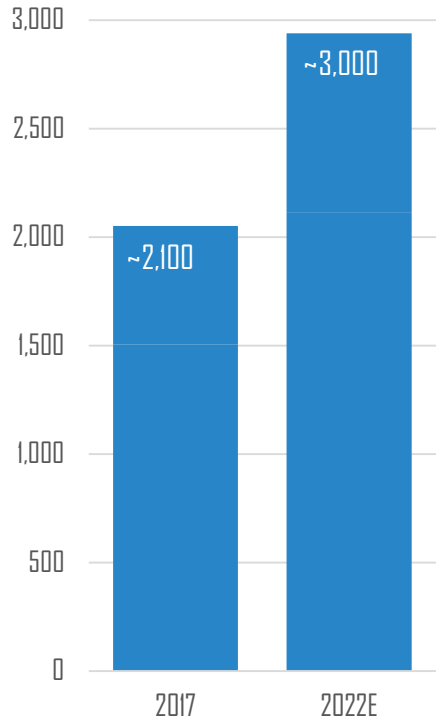
## Pulmonary Hypertension as Predictor of Survival in IPF

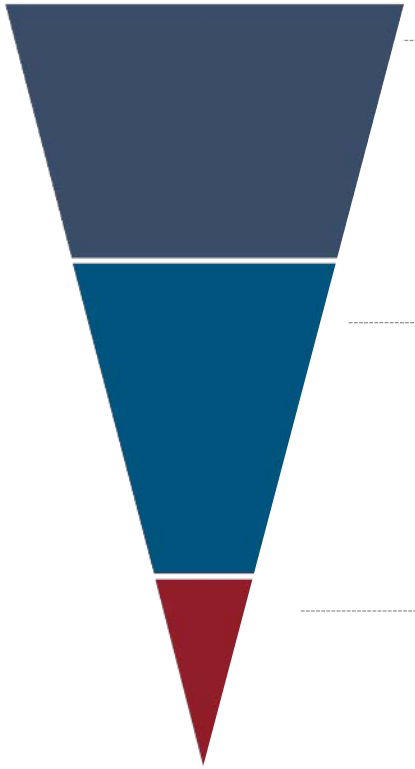



- Approximately 40% of IPF patients exhibit symptoms of pulmonary hypertension at rest, including elevated pulmonary pressures
- Prognosis and survival are significantly worse for patients with pulmonary hypertension
- PH-IPF associated with a 3-fold increase in risk of death compared to IPF alone
- **No approved therapy for treating PH in these patients**
- INOpulse has the potential to provide targeted vasodilation while avoiding concerns of V/Q mismatch which have prevented current PAH systemic vasodilators to be approved for this unmet medical need

Rivera-Lebron, Advances in Pulmonary Hypertension, 2013

# PH-PF Market Opportunity in the US



**PF Patient Population: ~226,000 in US**

Majority of patients >80% exhibit PH on exercise and can be eligible for INOpulse

~44% are IPF (~100,000 in US)

**PH-PF: ~84,000 in US**

Estimated patient population with PH at rest (35-40%)

Represents intermediate/high risk of PH by echo

**Estimated Penetration: ~24,000 in US Based on:**

LTOT penetration of 70-75%

Physician adoption rate of 35-60%

**$2B+ US Market Opportunity**

KOL feedback supports high unmet medical need in PH-PF

Pricing assessment supported by current IPF and PAH therapies priced at $100-200k/year

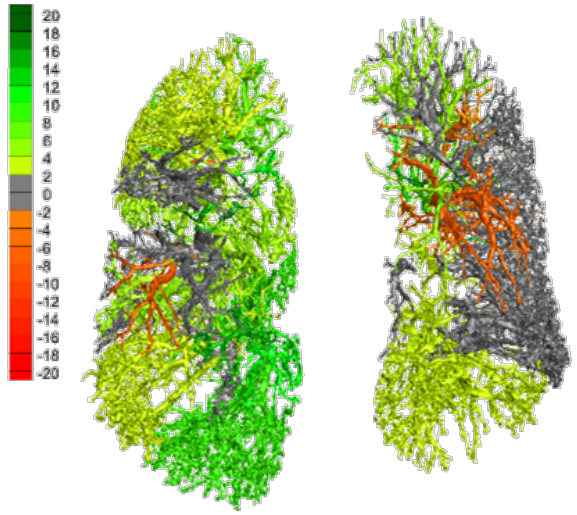
Source: LEK report

# Phase 2: INOpulse Demonstrates Targeted Vasodilation in PH-PF

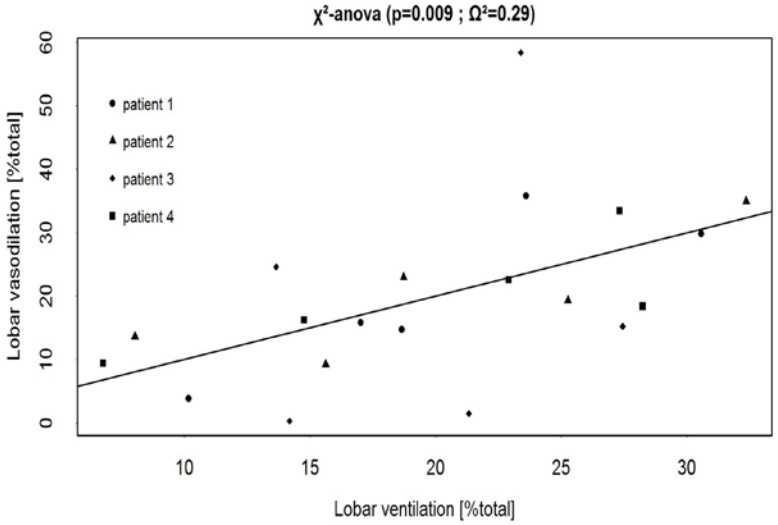
## Acute Phase Data Showed Immediate Benefit of iNO on Vasodilation and Hemodynamics

- Significant correlation between ventilation and vasodilation, demonstrating selective vasodilation to better ventilated areas of the lung (p=0.009)
- Consistent and clinically meaningful reduction of 14% in systolic pulmonary arterial pressure (sPAP)
- Clinically meaningful improvement oxygen desaturation of 28.5% and SpO2 nadir of 5.5%

### Targeted Vasodilation




### Ventilation/Vasodilation Matching

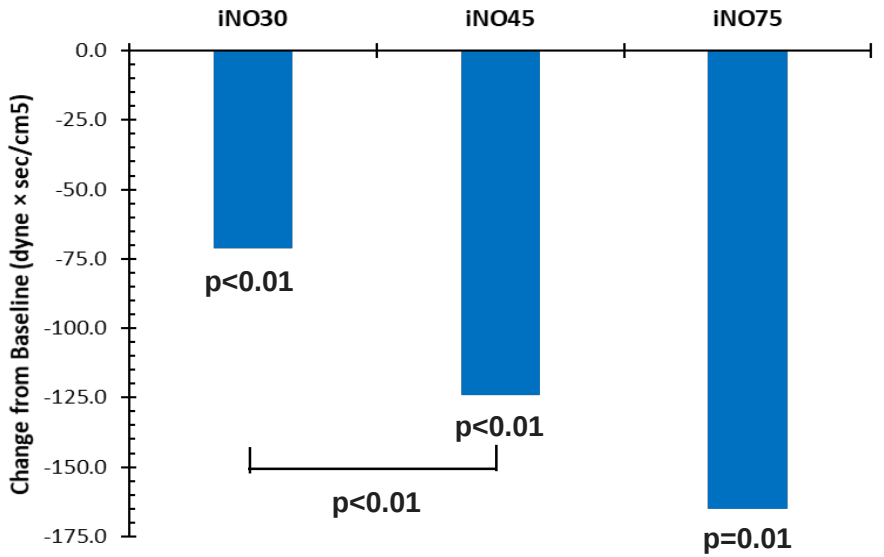


# Phase 2: INOpulse Demonstrates Acute Hemodynamic Benefit in PH-PF

## Clinically and statistically meaningful cardiopulmonary improvement on iNO30 w/ cont. benefit on dose escalation

Pulmonary hemodynamics measured via right heart catheterization at baseline and following each sequential iNO dose

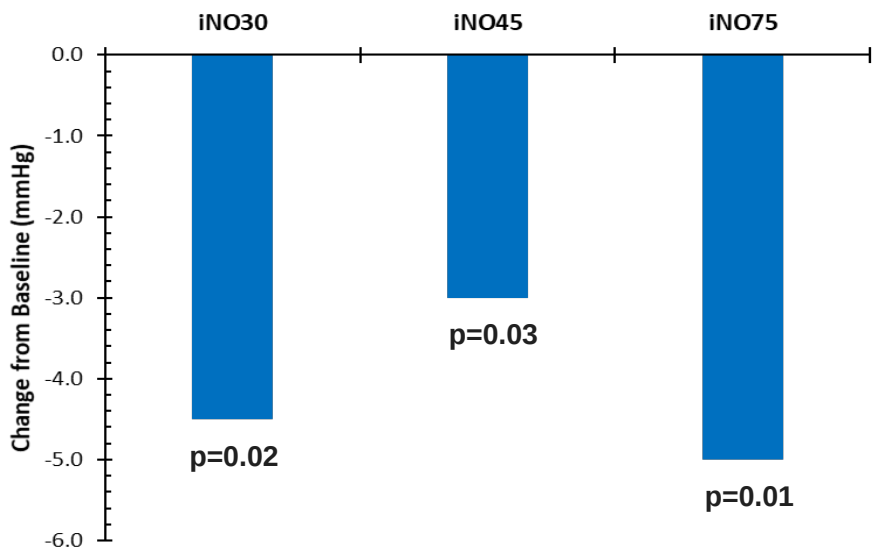
### Pulmonary Vascular Resistance (PVR)




Baseline PVR: 583 dyne×sec/cm$^5$

Average reduction of 125 dyne×sec/cm$^5$

### mean Pulmonary Arterial Pressure (mPAP)




Baseline mPAP: 34.7 mmHg

Average reduction of 4 mmHg

- p-value based on Wilcoxon Rank Test
- Bar graphs are median values at each assessment for all available subjects
- Oxygen saturation remained stable and iNO was well-tolerated with no safety concerns across doses
- Data based on 9 PH-PF subjects

# Continuous Physical Activity Monitoring (Actigraphy) Allows Objective Assessment of Daily Physical Activity

## Continuous Monitoring of Physical Activity

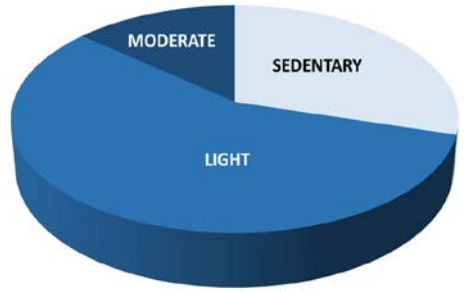
Subject wears actigraphy monitor on non-dominant arm

Monitor continuously measures movement in acceleration units

## Movement is Categorized into Activity Intensity Levels

| Activity Intensity | Example activities |
|---|---|
| **Sedentary**<br>(<100 counts)<br>(< 1.5 METs) | • Lying<br>• Sitting<br>• Computer work |
| **Light**<br>(100 -1951 counts)<br>(1.6 - 3.0 METs) | • Getting dressed<br>• Bathing/showering<br>• Light house cleaning |
| **Moderate**<br>(1952-5724 counts)<br>(3.1 - 6.0 METs) | • Walking<br>• Ascending/descending stairs<br>• Housework/yardwork |
| **Vigorous**<br>(>5724 counts)<br>(> 6.0 METs) | • Slow/fast running<br>• Intense sports |

## Provides Profile of Daily Activity




Subjects spend 74 minutes per day in moderate physical activity

Subjects are unable to achieve vigorous activity levels

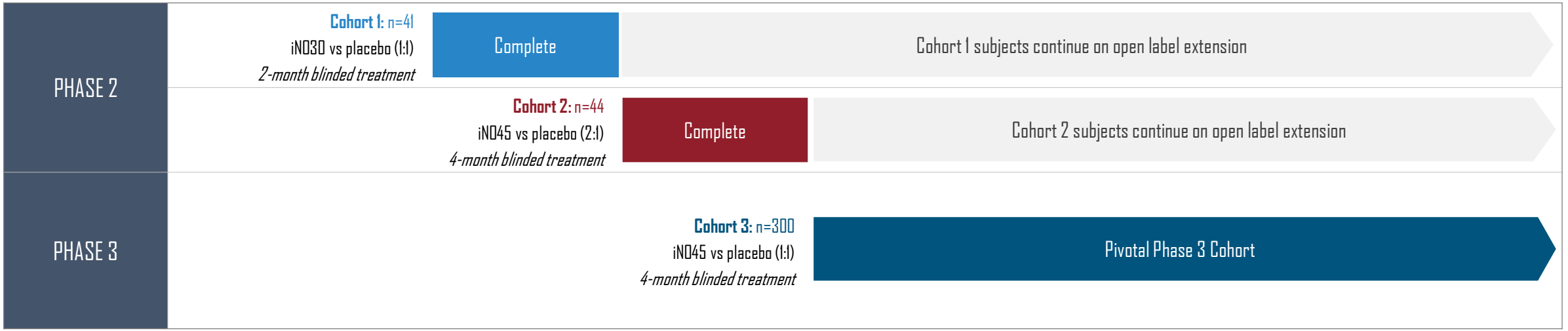
MVPA is the sum of moderate and vigorous activity

# Phase 2/3 (iNO-PF) Study Allows Immediate Transition into Pivotal Phase 3 Cohort

## FDA agreement on Cohort 3 to serve as pivotal Phase 3 study with MVPA as primary endpoint

Double-blind placebo-controlled study will assess subjects with pulmonary fibrosis at low or intermediate/high risk of associated pulmonary hypertension

- Phase 2b Complete: Cohorts 1 and 2 demonstrate statistically significant improvement in MVPA supported by other activity parameters and patient reported outcomes (Cohort 2)
- Phase 3: Pivotal Cohort 3 planned to be initiated in 2H 2020 with MVPA as primary endpoint and iNO45 dose



# Patient Demographics

| | Cohort 1 (n=41) | | Cohort 2 (n=44) | |
|---|---|---|---|---|
| | iNO 30<br>n=23 | Placebo<br>n=18 | iNO 45<br>n=30 | Placebo<br>n=14 |
| Age – years<br>(mean, SD) | 68.6 (6.45) | 65.8 (13.73) | 68.9 (9.95) | 71.4 (5.14) |
| Male<br>(n, %) | 16 (69.6%) | 13 (72.2%) | 15 (50.0%) | 10 (71.4%) |
| Intermediate to High Probability of PH<br>(n, %) | 15 (65.2%) | 14 (77.8%) | 18 (60.0%) | 9 (64.3%) |
| Baseline DLCO – % predicted<br>(mean, SD) | 30.7 (11.4) | 30.4 (10.2) | 35.7 (14.2) | 35.3 (7.9) |
| Baseline FVC – % predicted<br>(mean, SD) | 56.3 (10.2) | 59.9 (18.4) | 59.7 (15.9) | 60.5 (15.1) |

# Top-Line Results

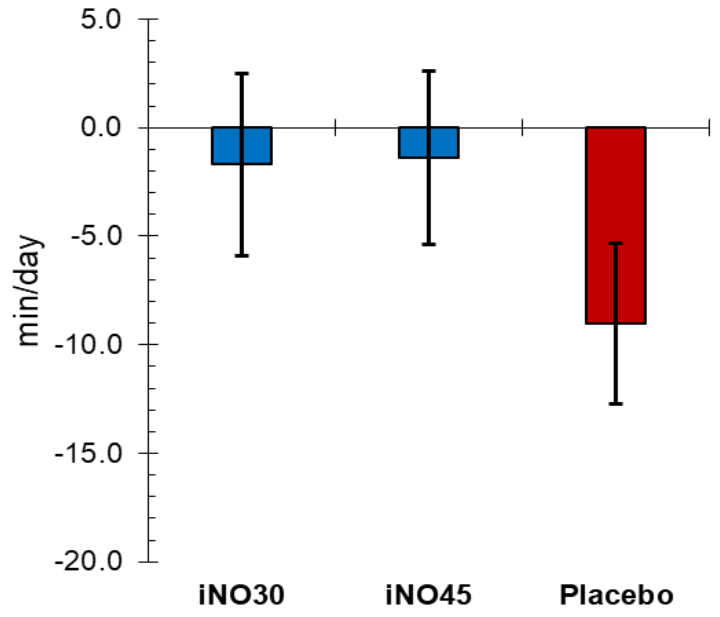
## Statistically and clinically significant benefit in MVPA (moderate to vigorous physical activity)

- Subjects on INOpulse (iNO30 and iNO45) maintain activity levels while subjects on placebo deteriorate across activity parameters
- Subjects on iNO45 maintain PRO scores (UCSD & SGRQ) while subjects on placebo deteriorate consistent with changes in activity levels
- Physical activity levels maintained during open label extension with subjects transitioning from placebo to active demonstrating reversal from decline to maintenance
- INOpulse targeted delivery maintains oxygen saturation during exercise
- INOpulse was safe and well-tolerated

# iNO45 Demonstrates Statistically and Clinically Meaningful Benefit in MVPA

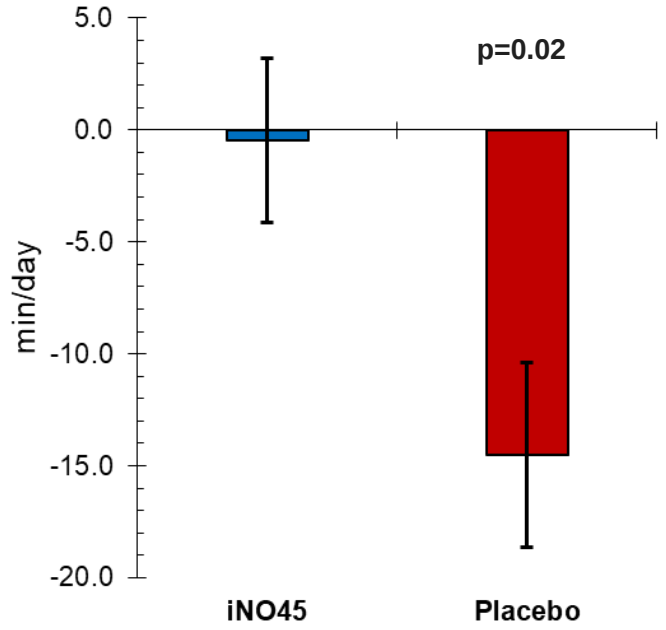
## Subjects on iNO maintain activity levels while subjects on placebo deteriorate

### MVPA Month 2




Subjects on INOpulse demonstrate 7.5 min/day (~10%) benefit over placebo after 2 months of treatment

### MVPA Month 4



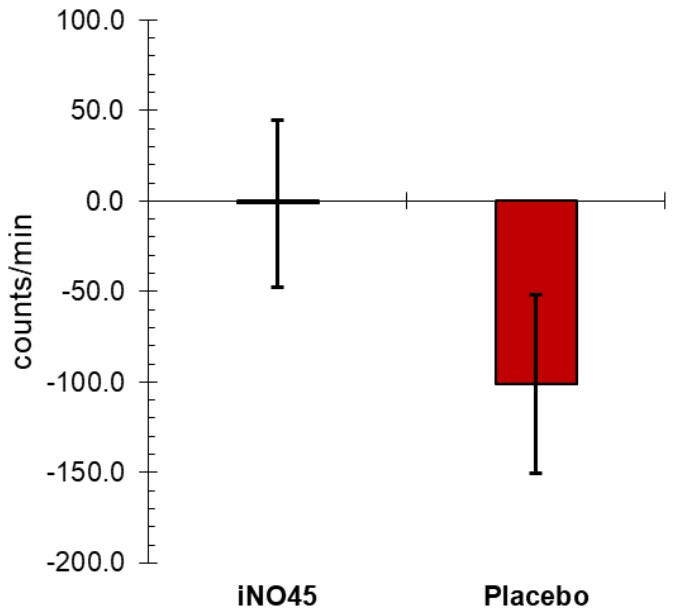

Benefit increases to 14 min/day (~20%) after 4 months of treatment

- Data points and error bars = mean and standard error
- Month 2 analysis based on pooled placebo from Cohorts 1 and 2

# iNO45 Demonstrates Consistent and Meaningful Benefit in Overall Activity

## Subjects on iNO maintain activity levels while subjects on placebo deteriorate

### Overall Activity Month 4

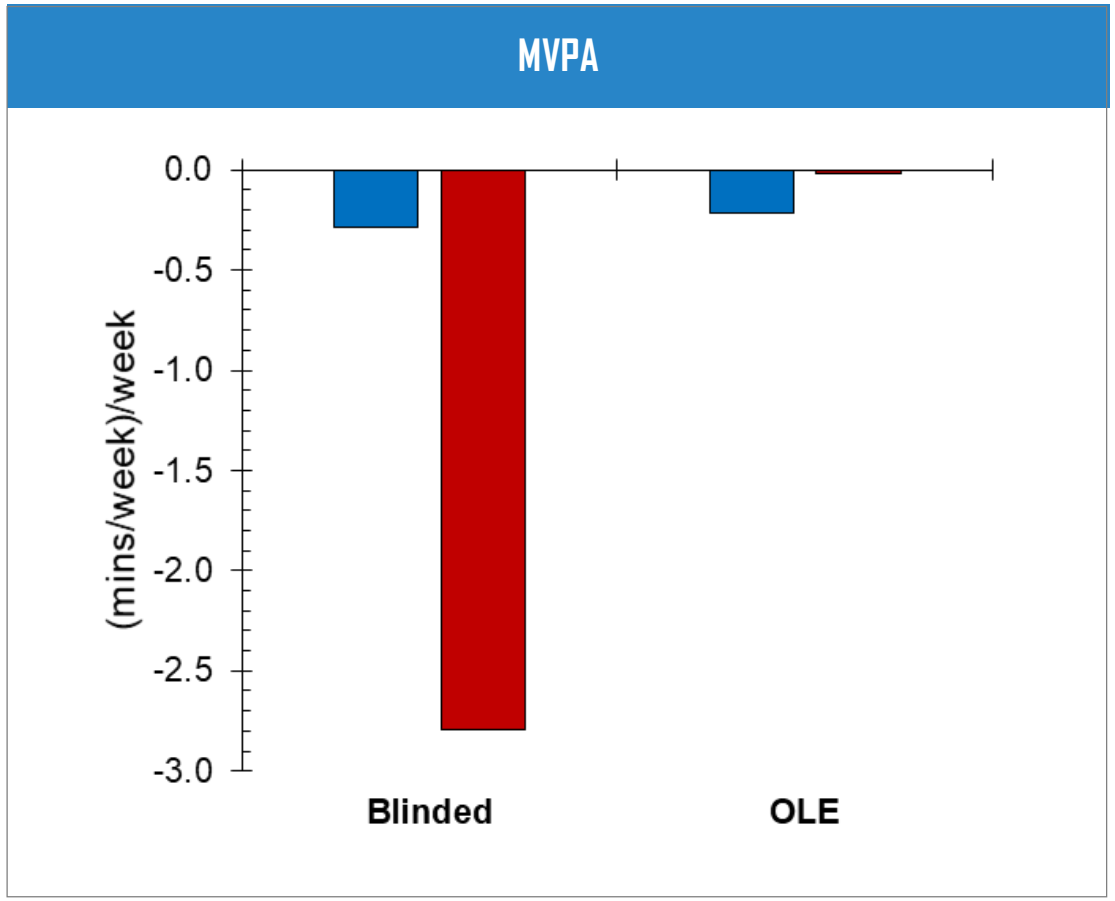


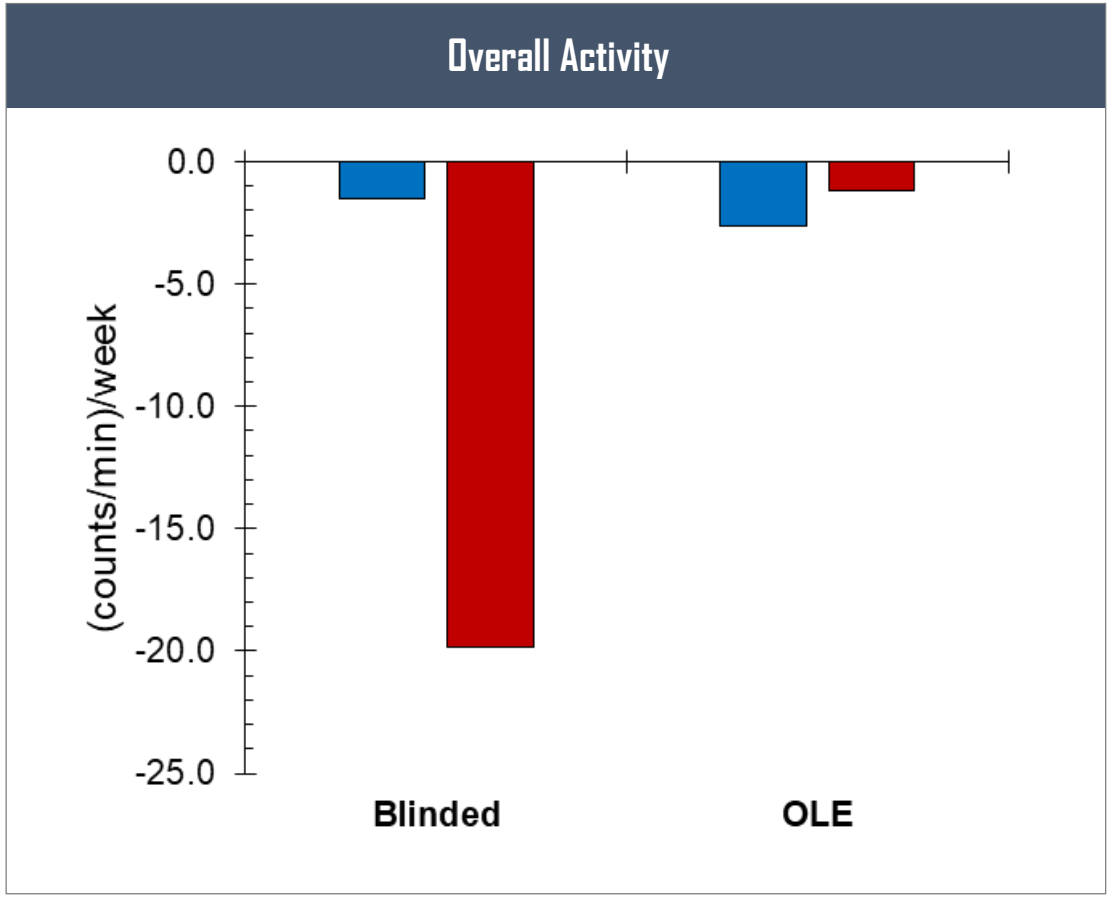

Subjects on INOpulse demonstrate ~100 counts/min (~7%) benefit over placebo after 4 months of treatment

• Data points and error bars = mean and standard error

# Continued Benefit for Subjects on Open Label Extension (OLE)

## Subjects transitioning from placebo to active demonstrate trend reversal from deterioration to maintenance

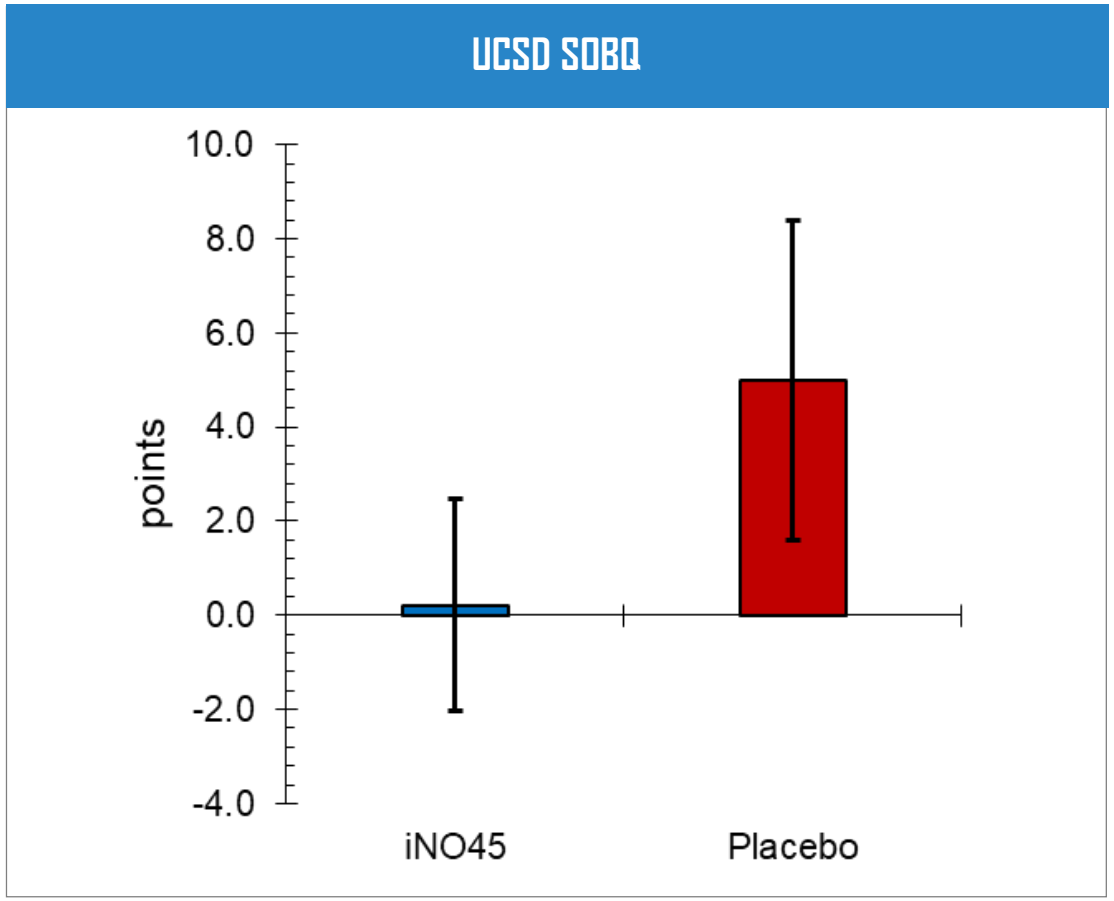



- MVPA = moderate to vigorous physical activity
- Change is based on Cohort I subjects with available data during blinded treatment and open label treatment (average 27 weeks of OLE)

# iNO45: UCSD Shortness of Breath Questionnaire (SOBQ) Indicates Benefit in Dyspnea

## Increased score indicative of worsening



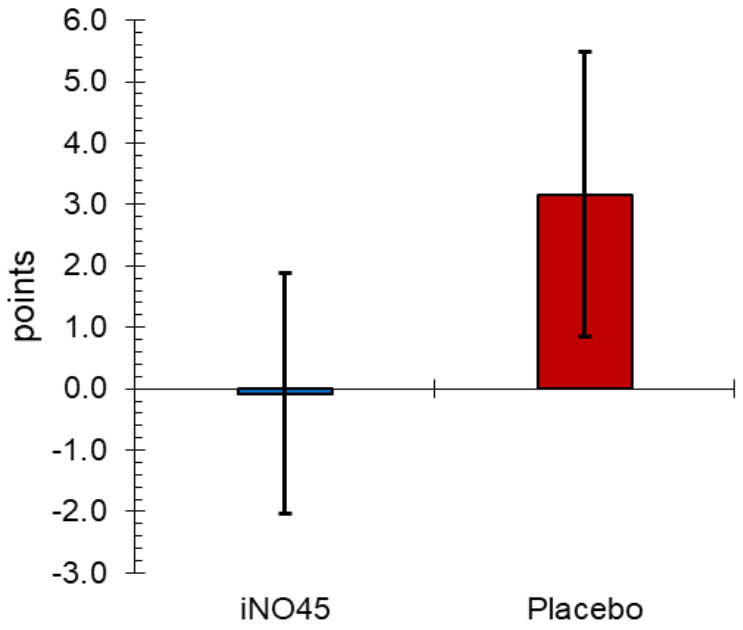

- Placebo corrected improvement of 5 points
- Measures shortness of breath while patients perform daily physical activity

- Data points and error bars = mean and standard error
- Change from baseline at month 4

# iNO45: St. George's Respiratory Questionnaire (SGRQ) Indicates QOL Benefit in Multiple Measures

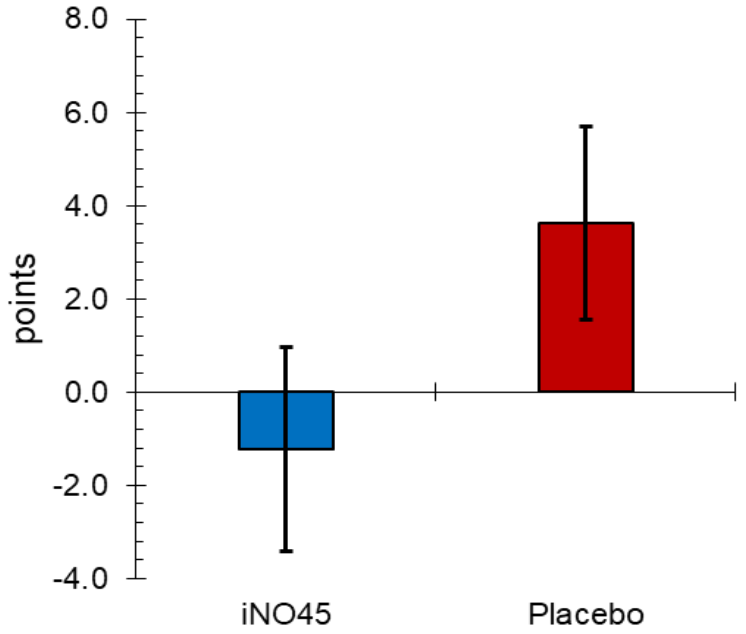
## Increased score indicative of worsening

### SGRQ Total



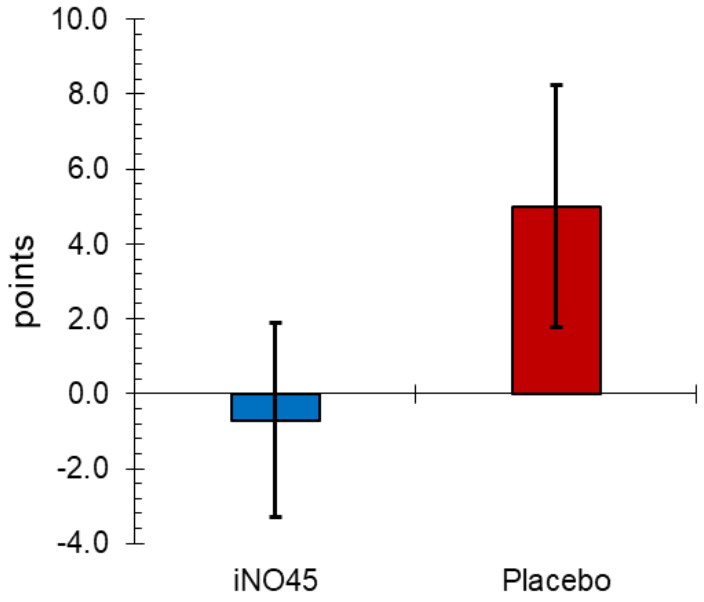

- Placebo corrected improvement of 3 points
- Measures health status and quality of life

### SGRQ Activity




- Placebo corrected improvement of 5 points
- Measures disturbances to patients' daily physical activity

### SGRQ Impacts




- Placebo corrected improvement of 6 points
- Measures psychological and social impact of the disease

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- Data points and error bars = mean and standard error
- Change from baseline at month 4

# Safety Summary

## iNO was well-tolerated in Cohort 1 and Cohort 2

- Incidence of SAEs was low in both active and placebo groups and all reported as unrelated to study drug
- AEs were balanced and generally non-serious with no observable trends

| | Cohort 1 (8 weeks) | | Cohort 2 (16 weeks) | |
|---|---|---|---|---|
| | iNO 30<br>n=23 | Placebo<br>n=18 | iNO 45<br>n=30 | Placebo<br>n=14 |
| Subjects with Adverse Events | 19 (82.6%) | 15 (83.3%) | 26 (86.7%) | 9 (64.3%) |
| Subjects with Serious Adverse Events | 2 (8.7%) | 2 (11.1%) | 3 (10.0%) | 3 (21.4%) |
| Total Serious Adverse Events Reported | 3 (0.13/subject) | 4 (0.22/subject) | 4 (0.14/subject) | 7 (0.5/subject) |
| Deaths | 1 (4.3%) | 0 | 0 | 0 |

# Next Steps in PH-PF

## FDA Agreement on iNO-PF Phase 2/3 Study Design

- MVPA as measured by actigraphy is primary endpoint for Phase 3
- Cohort 3 of ongoing iNO-PF study serves as the pivotal Phase 3 study for approval

## Cohort 3 Pivotal Phase 3 Study

- iNO45 planned to be progressed into Phase 3 based on results from Cohorts 1 and 2
- Primary endpoint is change in MVPA as measured via actigraphy
- Double-blind placebo-controlled study will assess subjects with pulmonary fibrosis at low or intermediate / high risk of associated pulmonary hypertension
- Planned enrollment of 300 subjects (150 per arm) powered for primary endpoint of MVPA as well as multiple secondary endpoints
- 4-month blinded treatment period
- Study to be initiated in 2H 2020

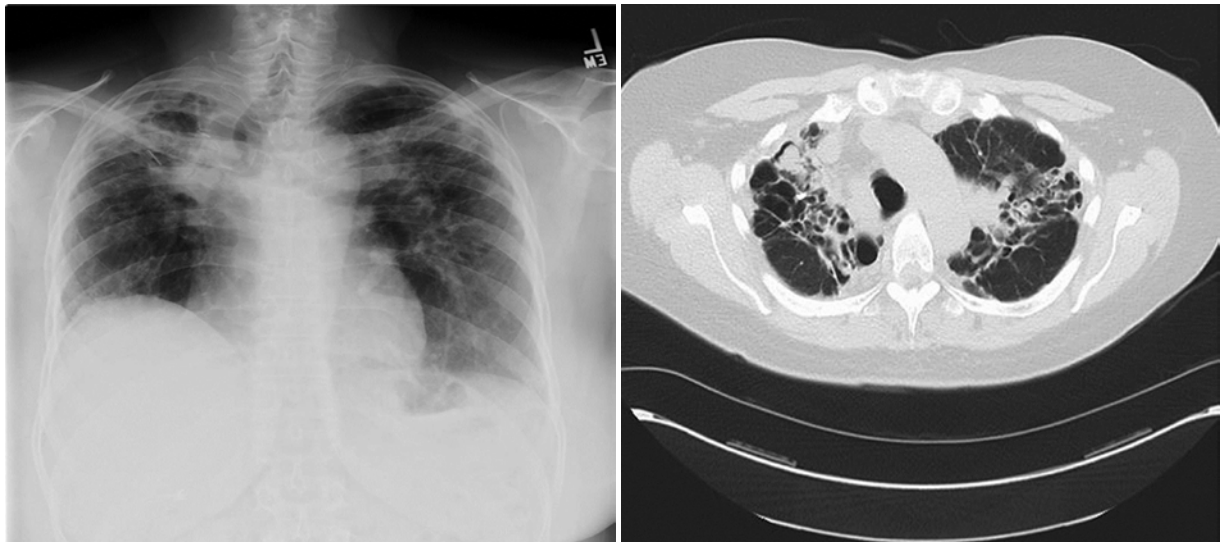
# Pulmonary Hypertension associated with Sarcoidosis (PH-Sarc)

## An Orphan Unmet Medical Need

Sarcoidosis is characterized by the growth of inflammatory cells (granulomas) most commonly in the lungs or lymphatic tissues

Prevalence of sarcoidosis is estimated at 200,000 in the US with up to 30% with associated pulmonary hypertension

**Chest x-ray & CT of patient with PH-Sarc**



**Patients with associated PH have significantly reduced survival**

| | 1-year survival | 3-year survival | 5-year survival |
|---|---|---|---|
| PH-Sarc | 84% | 74% | 59% |
| Sarc | 100% | 96% | 96% |

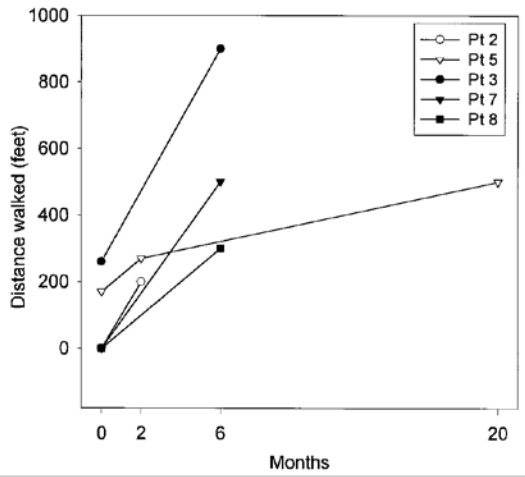
# INOpulse MoA has the Potential to Provide Benefit to PH-Sarc Patients

## Inhaled nitric oxide has been shown to improve hemodynamics and exercise capacity in PH-Sarc

**Acute Hemodynamic Benefit on iNO**

| Parameter | % Change |
|---|---|
| mPAP | -18 ± 4 |
| PVR | -31 ± 5 |
| CO | 12 ± 4 |

**Chronic Benefit on Exercise Capacity**




Systemic vasodilators exacerbate hypoxic vasoconstriction and cause hypoxemia

**No approved therapy for treating PH in these patients**

Phase 2 study designed to verify hemodynamic effect of INOpulse in PH-Sarc

- Acute dose escalation study with right heart catheterization
- Primary endpoint: change in mean PAP, PCWP, cardiac output and PVR
- Study results expected in 2H 2020

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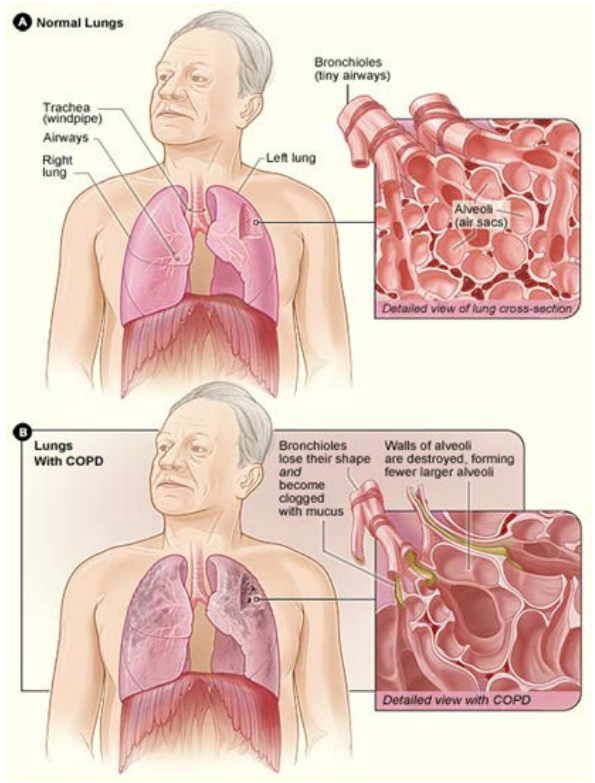
Preston et al., Chest, 2001

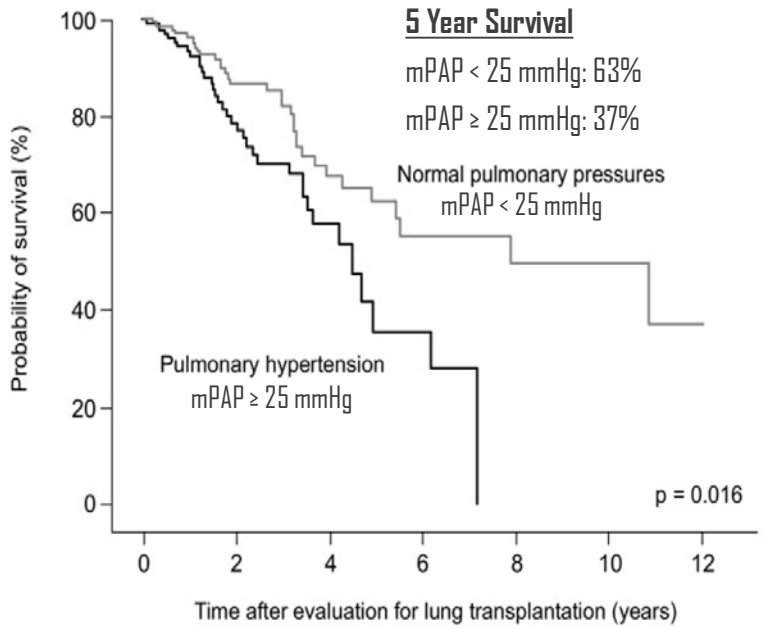
# Pulmonary Hypertension associated with Chronic Obstructive Pulmonary Disease (PH-COPD) Represents Large Unmet Medical Need

## COPD is a group of lung diseases characterized by progressive airflow obstruction and chronic airway inflammation




- Typically associated with smoking or exposure to other pollutants
- Obstruction of the bronchioles and alveoli reduces the ability to get oxygen and ultimately leads to hypoxemia
- Hypoxemia and inflammation contribute to development pulmonary hypertension

## Pulmonary hypertension predicts survival in COPD



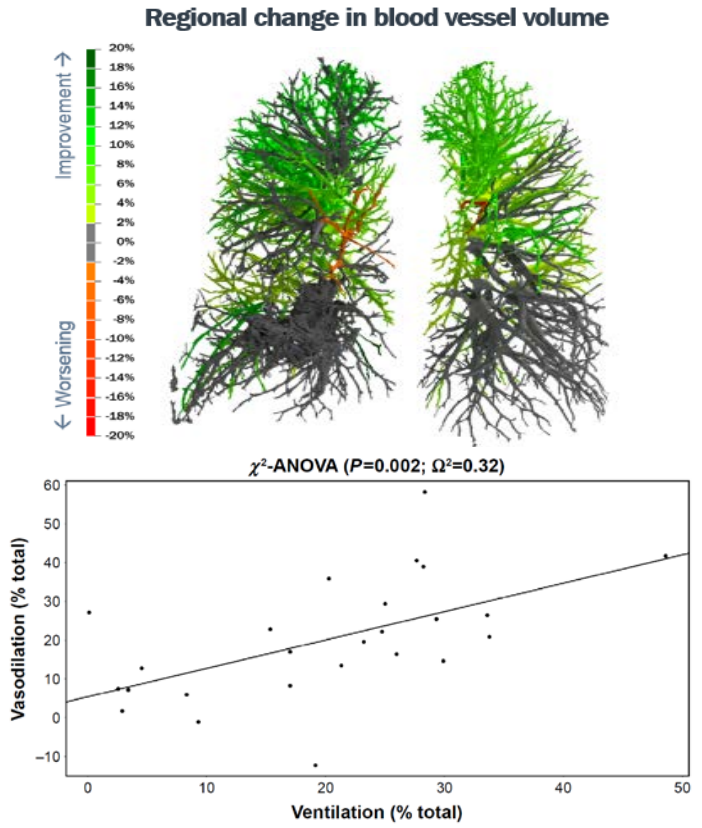

Survival years

**No approved therapy for treating PH in these patients**

Andersen KH et al. Prevalence, Predictors and Survival in Pulmonary Hypertension Related to End-stage Chronic Obstructive Pulmonary Disease. Journal of Heart and Lung Transplantation 2012; 31: 373-380.

# INOpulse Provides Targeted Delivery and Improves Hemodynamics and Exercise Capacity in PH-COPD Patients

## INOpulse targets delivery to well ventilated pulmonary vessels

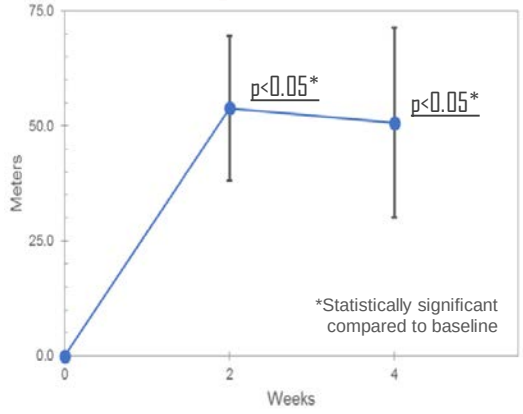



## Chronic treatment with INOpulse provides statistically significant improvement in 6MWD and hemodynamics

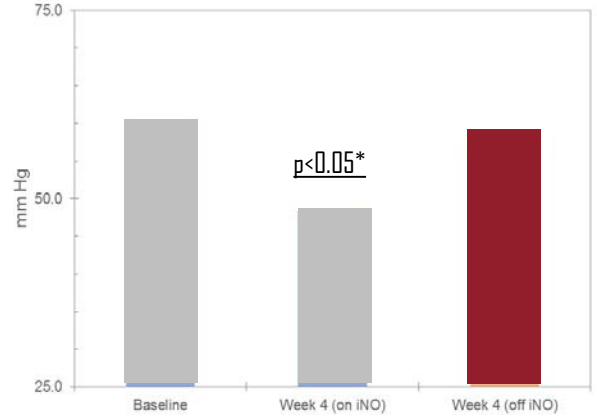
### Subjects completing 4 weeks on iNO 30 demonstrated:

- Statistically significant increase in 6MWD at 2 weeks and 4 weeks (+50.7m)
- Statistically and clinically significant decrease in sPAP at 4 weeks (-12.0 mmHg; 19.9% reduction)
- sPAP increased to near baseline upon stopping treatment with iNO

### Change in 6MWD




### sPAP




Haijan et al., Int J of COPD, 2016

# INOpulse Intellectual Property Protection

| Patent | Status | Expiration | Description |
|---|---|---|---|
| Method of NO administration | US/EU: Issued<br>Other Territories: Issued/Pending | Jan 2027 | Covers consistent delivery of prescribed dose independent of respiratory rate |
| Breath Skipping & Pulse Volume Variation | US: Issued | Sept 2025 | Covers skipping breaths or modifying pulse volume to ensure consistent dose independent of respiratory rate |
| Method of Administering High Concentration NO | US/EU: Pending<br>Other Territories: Pending | Mar 2033 | Limits delivery rate of high concentration iNO to prevent safety concerns |
| Optimized Pulse Shape | US: Pending | Oct 2035 | Covers key parameters of pulse shape |
| INOpulse Design | US: Issued | Apr 2028 | Covers design of the INOpulse device |
| Tip Purge | US: Issued<br>EU/Other Territories: Pending | Apr 2033 | Covers the use tip purge to ensure purity of iNO within the cannula |
| Triple-Lumen Cannula | US/ EU: Issued<br>Other Territories: Pending | Dec 2033 | Covers accurate dose delivery and reduced $NO_2$ formation |
| Index valve | US/EU: Issued<br>Other Territories: Issued/Pending | May 2029 | Ensures other cartridges cannot be used with INOpulse |

Orphan Drug designation for IPF provides **exclusivity for 7 years (US) and up to 10 years (EU)**

Multiple provisional patent applications filed from 2017-2019 that can **extend patent coverage into 2039**

# Financial Summary

| | Amount (in millions) |
|---|---|
| Cash and Cash Equivalents | $63.1[1,2] |
| Restricted Cash | $0.4[1] |
| Debt | $0[1] |
| Fully Diluted Shares Outstanding | 12.2 |

1. As of March 31, 2020
2. Includes $54.5M of net proceeds from public offerings and registered direct offerings that closed in Q2/2020

# Investment Highlights

## Established iNO Therapeutic Benefit

- Approved for acute treatment of persistent pulmonary hypertension in neonates
- Positive results from multiple Phase 2 studies support INOpulse MoA and benefit

## Advanced Clinical Stage Product

INOpulse technology focused on several large unmet orphan indications

| PH-PF | PH-SARC / PH-COPD | COVID-19 / Infectious Lung Diseases |
|---|---|---|
| Successful Phase 2 studies in PH-IPF | PH-Sarc: Phase 2 results expected in 2H 2020 | 180 patients treated with INOpulse under expanded access |
| Positive results for Phase 2/3 study in Cohorts 1 and 2 | PH-COPD: Successful Phase 2 study completed | FDA cleared Phase 3 placebo-controlled registrational study |
| Pivotal Phase 3 cohort to initiate in 2H 2020 with FDA agreement on primary endpoint | PH-COPD: Phase 2b study design finalized in agreement w/ FDA | Phase 3 (COViNOX) study initiated in July 2020 |

## Proprietary INOpulse Technology

Strong IP protection on core programs through 2033 and ability to extend coverage into 2039

# Investor Contacts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