

### 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: 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.



## iNO-PF Cohort 2 Top-Line Results

Statistically and clinically significant placebo corrected improvement of 14 minutes/day in MVPA

Consistent and meaningful benefit in overall activity

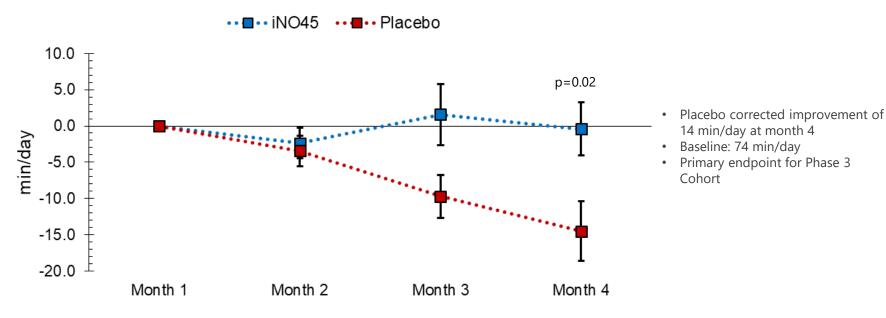
• Clinically meaningful benefit in SGRQ and UCSD demonstrating improvement in quality of life and dyspnea

Pulsed inhaled NO was safe and well-tolerated



### Statistically Significant Benefit Over Placebo Demonstrated in MVPA

#### Normalized MVPA - Change from baseline

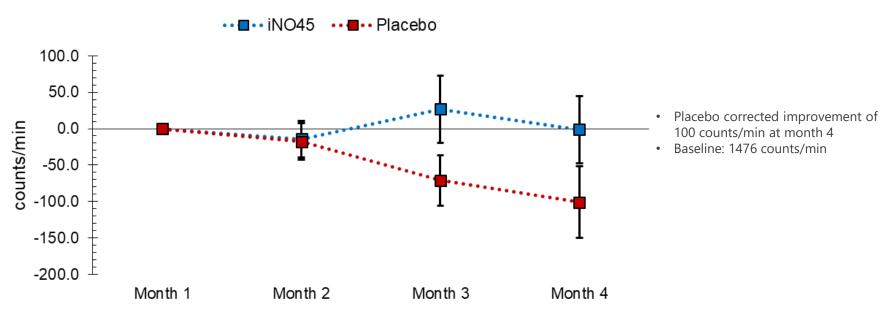


- MVPA = moderate to vigorous physical activity
- Data points and error bars = mean and standard error
- Cohort 2: n=44; randomized 2:1



## Consistent and Meaningful Benefit Exhibited in Overall Activity

### Overall Activity - Change from baseline

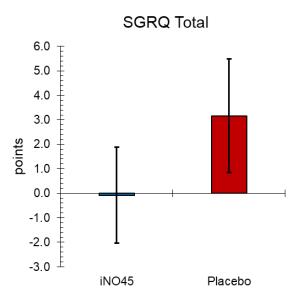


- Data points and error bars = mean and standard error
- Cohort 2: n=44; randomized 2:1

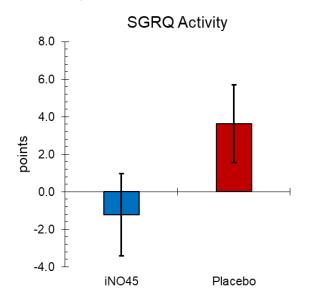


# St. George's Respiratory Questionnaire (SGRQ) Indicates QOL Benefit in Multiple Measures

Increased score indicative of worsening



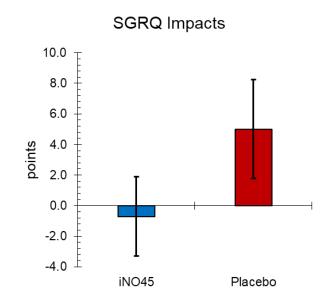
- Placebo corrected improvement of 3 points
- · Measures health status and quality of life



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



- Change from baseline at month 4
- Cohort 2: n=44; randomized 2:1

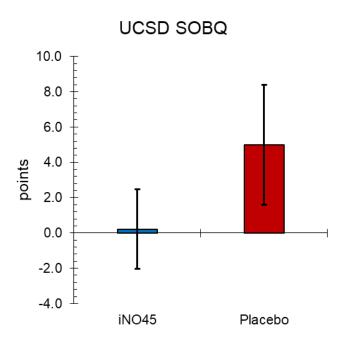


- Placebo corrected improvement of 6 points
- Measures psychological and social impact of the disease



### 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
- Cohort 2: n=44; randomized 2:1



# Safety Summary

- Pulsed Inhaled Nitric Oxide was welltolerated in Cohort 2
  - Incidence of AEs and SAEs was low in both active and placebo groups
  - AEs were generally non-serious with no observable trends
  - All SAEs were reported as unrelated to the study drug

	iNO 45 n=30	Placebo n=14
Total Adverse Events Reported	26 (0.87/subject)	9 (0.64/subject)
Total Serious Adverse Events Reported	5 (0.17/subject)	7 (0.50/subject)
Subjects with Serious Adverse Events	3 (10%)	3 (21.4%)
Deaths	0	0



## Next Steps in PH-ILD

• Results from Cohort 2 support continuing program into Phase 3 with MVPA as primary endpoint

Phase 3 initiation expected in 1Q2020 upon selection of dose and confirmation of trial size

 Company entered into \$10M convertible financing facility with existing institutional investors, extending cash runway into 2021





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