



Liquidia R&D Day

Webcast presentation

October 28, 2025

Forward-looking statements

This presentation includes, and our response to questions may include, forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995 (“PSLRA”). All statements contained in this presentation other than statements of historical facts, including statements regarding our future results of operations and financial position, our strategic and financial initiatives, our business strategy and plans and our objectives for future operations, are forward looking statements. Such forward-looking statements, including statements regarding clinical trials, clinical studies and other clinical work (including the funding therefor, anticipated patient enrollment, safety data, study data, trial outcomes, timing or associated costs), regulatory applications and related submission contents and timelines, the timelines or outcomes related to patent litigation with United Therapeutics in the U.S. District Court for the District of Delaware and U.S. District Court for the Middle District of North Carolina, or other litigation between Liquidia and United Therapeutics or others, including rehearings or appeals of decisions in any such proceedings, the issuance of patents by the USPTO and our ability to execute on our strategic or financial initiatives, the potential for additional funding under the HCR Agreement, our anticipated use of net proceeds funded under the HCR Agreement, our estimates regarding future expenses, capital requirements and needs for additional financing, and potential revenue and profitability of YUTREPIA involve significant risks and uncertainties and actual results could differ materially from those expressed or implied herein. YUTREPIA’s approval and our launch of YUTREPIA remain subject to ongoing litigation in which United Therapeutics is seeking injunctive relief, which could block our ability to continue to sell YUTREPIA for one or both of PAH and PH-ILD. The words “anticipate,” “believe,” “continue,” “could,” “estimate,” “expect,” “intend,” “may,” “plan,” “potential,” “predict,” “project,” “should,” “target,” “would,” and similar expressions are intended to identify forward-looking statements. We have based these forward-looking statements largely on our current expectations and projections about future events and financial trends that we believe may affect our financial condition, results of operations, business strategy, short-term and long-term business operations and objectives and financial needs. These forward-looking statements are subject to a number of risks discussed in our filings with the U.S. Securities and Exchange Commission as well as a number of uncertainties and assumptions. Moreover, we operate in a very competitive and rapidly changing environment, and our industry has inherent risks. New risks emerge from time to time. It is not possible for our management to predict all risks, nor can we assess the impact of all factors on our business or the extent to which any factor, or combination of factors, may cause actual results to differ materially from those contained in any forward-looking statements we may make. In light of these risks, uncertainties and assumptions, the future events discussed in this presentation may not occur and actual results could differ materially and adversely from those anticipated or implied in the forward-looking statements. Although we believe that the expectations reflected in the forward-looking statements are reasonable, we cannot guarantee that future results, levels of activity, performance, achievements or events and circumstances reflected in the forward-looking statements will occur. We are under no duty to update any of these forward-looking statements after the date of this presentation to conform these statements to actual results or revised expectations, except as required by law. This presentation also contains estimates and other statistical data made by independent parties and by us relating to market size and growth and other data about our industry. This data involves a number of assumptions and limitations, and you are cautioned not to give undue weight to such estimates. In addition, projections, assumptions and estimates of our future performance and the future performance of the markets in which we operate are necessarily subject to a high degree of uncertainty and risk. This presentation includes long-term goals that are forward-looking, are subject to significant business, economic, regulatory and competitive uncertainties and contingencies, many of which are beyond our control and are based upon assumptions with respect to future decisions, which are subject to change. Actual results will vary, and those variations may be material. Nothing in this presentation should be regarded as a representation by any person that these goals will be achieved. We have no obligation under the PSLRA to update any forward-looking statements, and we undertake no duty to update our goals or to update or alter any forward-looking statements, whether as a result of new information, future events or otherwise.

Enhancing drug delivery to the lungs to **make every breath count**



Liquidia Corporation is a biopharmaceutical company **driven by science and compassion** to **revolutionize care** for patients with challenging **respiratory and vascular diseases** through precise, innovative therapies that **restore health and hope**

- **Improving drug delivery**
- **Using proprietary technologies**
- **Reducing burden of administration**
- **Helping patients breathe easier & live longer**

Ideal product profile for inhaled delivery

Therapeutic goal is to optimize each aspect

Targeted lung delivery



Reduces off-target toxicity from oral, IV/SC delivery

Portable



Convenience & ease-of-use to support compliance

Tolerable



Customizable and not dose limited

Titratable



Wide dose range to extend time on treatment

Dosing frequency



Easy and simple regimen

Yutrepia addresses the first four elements now

Therapeutic goal is to optimize each aspect

**Targeted
lung delivery**



**PRINT[®]
Technology**

Portable



**Low-effort
Trusted device**

Tolerable



**INSPIRE (PAH)
ASCENT (PH-ILD)**

Titratable



**Titratable in
PAH, PH-ILD**

**Dosing
frequency**



4x daily

YUTREPIA[™] (treprostinil) inhalation powder

L606 improves the inhaled product profile as the market expands

Therapeutic goal is to optimize each aspect

**Targeted
lung delivery**



**Liposomal
Technology**

Portable



**Rapid, breath-
actuated
nebulizer**

Tolerable



**Further
improved
tolerability**

Titratable



**Over 48-weeks
of treatment**

**Dosing
frequency**



**2x daily to
minimize peak
to trough
excursions**

YUTREPIA™ (treprostinil) inhalation powder

L606 (liposomal treprostinil inhalation solution)

Meet today's invited expert speakers

Key opinion leaders



UCLA
Health®

Dr. Richard Channick, MD

Saul Brandman Endowed Chair in Pulmonary Arterial Hypertension
Co-Director, Pulmonary Vascular Disease Program
Professor of Medicine
Pulmonary and Critical Care Division
David Geffen School of Medicine at UCLA



UCLA
Health®

Dr. Rajan Saggar, MD

Professor of Medicine
Director, Pulmonary Hypertension Program
Co-Director Pulmonary Vascular Disease Program
Lung & Heart-Lung Transplant and Pulmonary Hypertension Programs
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TGH Tampa General Hospital
USF
HEALTH

Dr. Ricardo Restrepo-Jaramillo

Associate Professor of Medicine, Morsani College of Medicine
Medical Director Center For Pulmonary Vascular Diseases
University of South Florida/
Tampa General Hospital

Today's outline

➤ **Our objectives**

➤ **Understanding PAH and PH-ILD today**

➤ **ASCENT study in PH-ILD (Week 24 data)**

➤ **L606, treprostinil liposomal inhalation suspension in U.S. (Week 48 data)**

➤ **Physician Roundtable Q&A**

Unmet Needs for Patients with PAH and PH-ILD

Dr. Richard Channick, MD

WHO clinical classification of PH

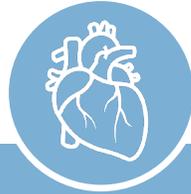


Group 1

PAH

- Idiopathic/heritable
- Associated conditions

Pressure in pulmonary arteries leads to vascular resistance, increased right ventricular pressure, right heart failure and death



Group 2

PH associated with left heart disease

- lpcPH
- CpcPH



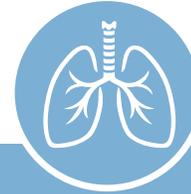
Group 3

PH associated with lung disease

- Non-severe PH
- Severe PH

Chronic hypoxia can cause **pulmonary vascular remodeling** and subsequent **elevation of pulmonary vascular resistance (PVR)**

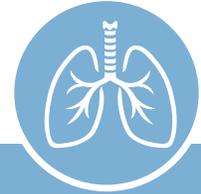
Ultimately leading to the development of PH within the course of ILD³



Group 4

PH associated with pulmonary artery obstructions

- CTEPH
- Other pulmonary



Group 5

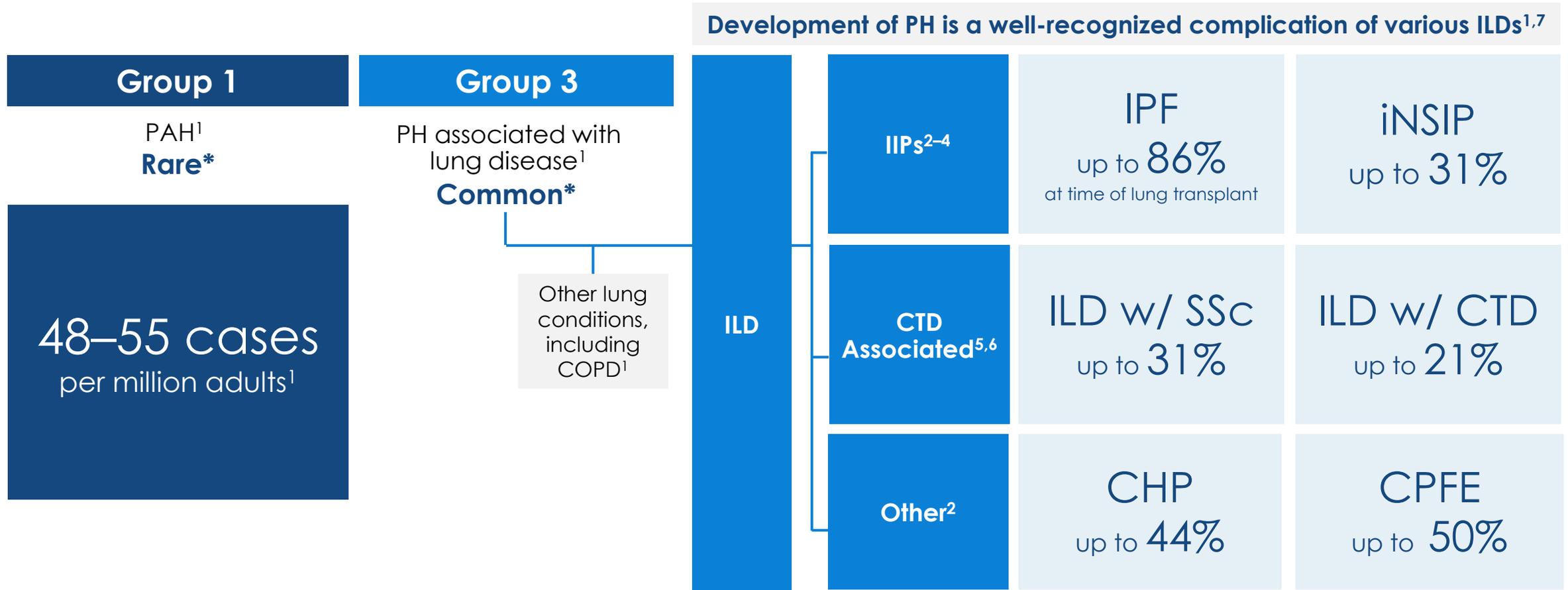
PH with unclear or multifactorial mechanisms

- Hematological disorders
- Systemic disorders

CpcPH, combined post- and pre-capillary pulmonary hypertension; CTEPH, chronic thromboembolic pulmonary hypertension; ILD, interstitial lung disease; lpcPH, isolated post-capillary pulmonary hypertension; PAH, pulmonary arterial hypertension; PH, pulmonary hypertension.

1. Humbert M et al. *Eur Heart J*. 2022;43(38):3618-3731. doi:10.1093/eurheartj/ehac237 2. Chang KY et al. *J Am Heart Assoc*. 2022;11(9):e024969. doi:10.1161/JAHA.121.024969 3. Kacprzak A et al. *Diagnostics*. 2023;13(14):2354. doi:10.3390/diagnostics13142354

Prevalence estimates may change with increased focus on treatment



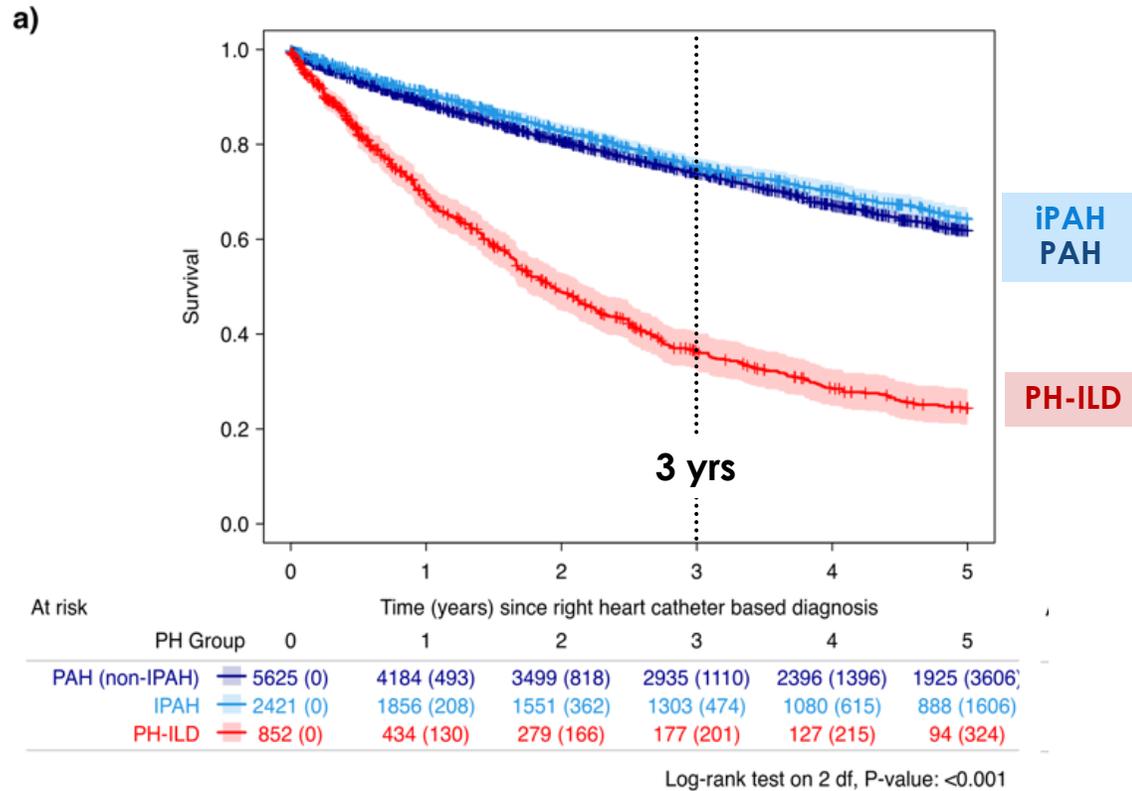
*Within pulmonary hypertension. CHP, chronic hypersensitivity pneumonitis; COPD, chronic obstructive pulmonary disease; CPFE, combined pulmonary fibrosis and emphysema; CTD, connective tissue disease; IIP, idiopathic interstitial pneumonia; iNSIP, idiopathic non-specific interstitial pneumonia; IPF, idiopathic pulmonary fibrosis.

1. Humbert M et al. *Eur Heart J.* 2022;43(38):3618-3731. doi:10.1093/eurheartj/ehac237 2. Nikkho SM et al. *Pulm Circ.* 2022;12(3):e12127. doi:10.1002/pul2.12127 3. Parikh R et al. *Pulm Circ.* 2022;12(4):e12141. doi:10.1002/pul2.12141 4. Rahaghi FF et al. *Chest.* 2022;162(1):145–155. doi:10.1016/j.chest.2022.02.012 5. Young et al. *Arthritis Rheumatol.* 2017;71(8):1339–1349. doi:10.1002/art.40862 6. Hyltdgaard et al. *J Clin Med.* 2021;10(21):4830. doi:10.3390/jcm10214830 7. Kacprzak A et al. *Diagnostics.* 2023;13(14):2354. doi:10.3390/diagnostics13142354

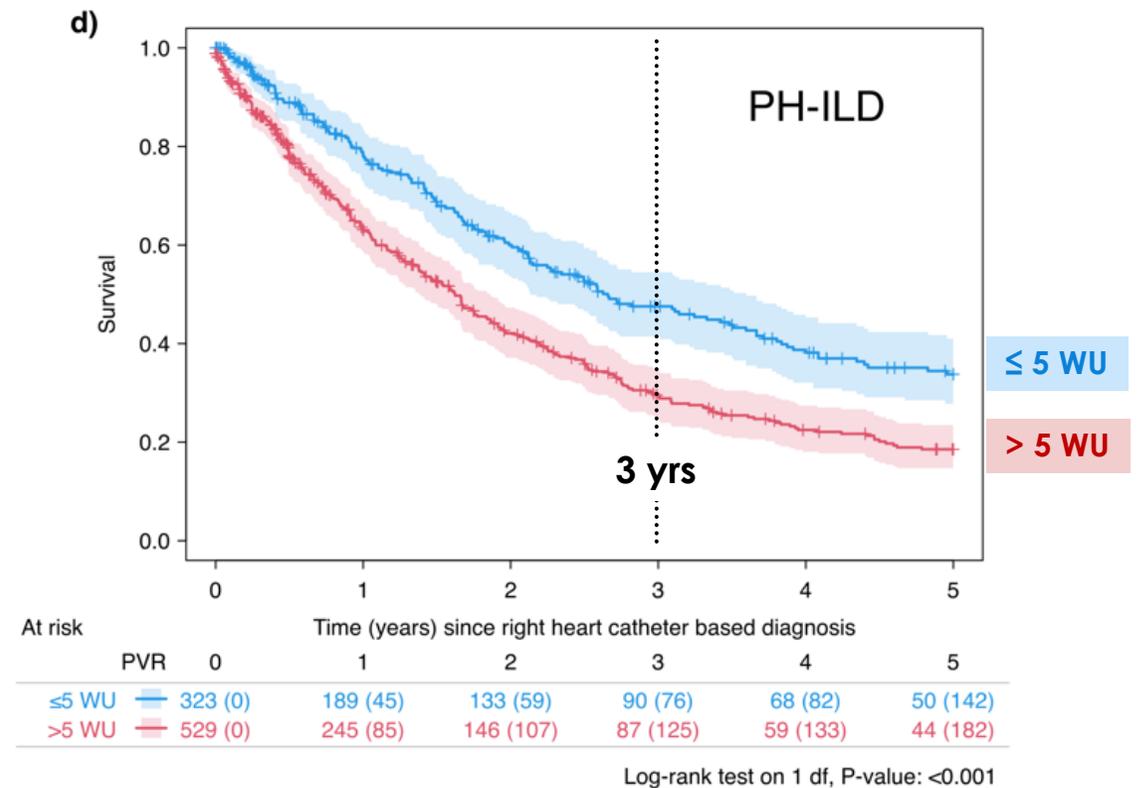
Recent real-world data reinforces poor outcomes in PAH & PH-ILD

PVRI GoDeep Registry¹

Risk of death is 2x higher in PH-ILD vs PAH



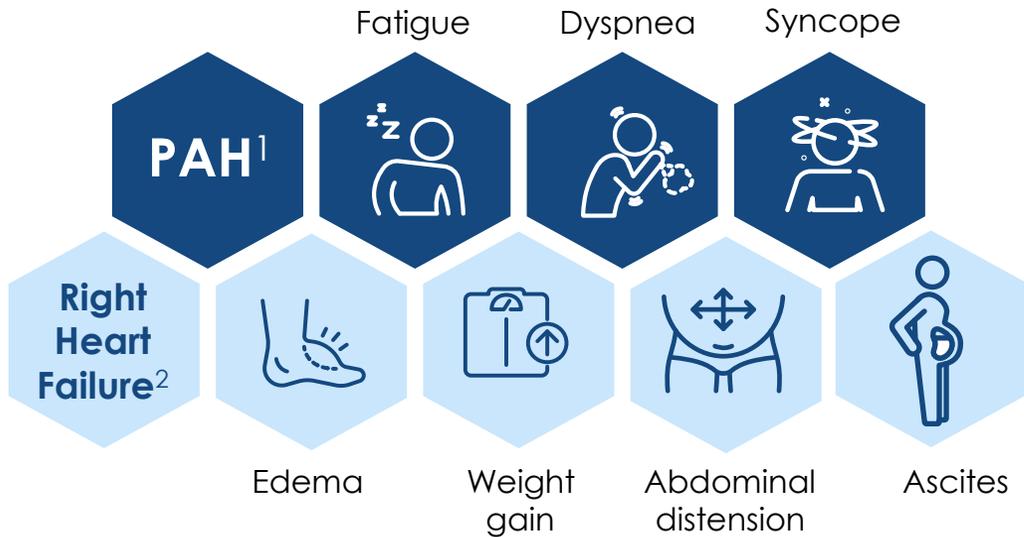
PVR is the strongest predictor of survival



Yogeswaran et al "Hemodynamics and PDE5i Treatment in PH-ILD" *Am J Respir Crit Care Med* Vol 211, Iss 10, pp 1855–1866, Oct 2025

Symptoms can be similar with different diagnosis

Early diagnosis and intervention may result in better clinical outcomes

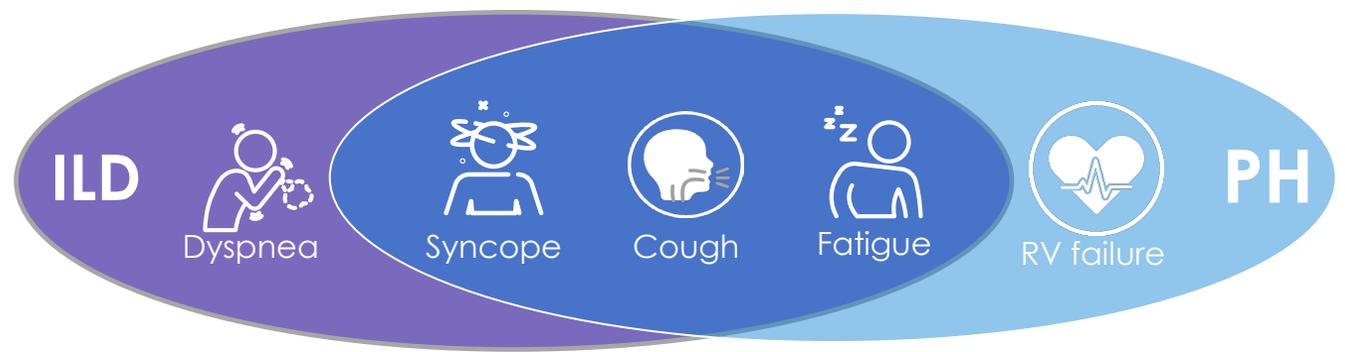


Detection: Assessment by Lung and Heart Specialist¹

- Echocardiography
- CPET

Confirmation: Comprehensive PH Workup by PH Center¹

- 6MWD
- Blood test
- Echocardiography or cMRI
- ABG or O₂ saturation
- Disease-specific HR-QoL
- CPET & RHC



Monitor for signs and symptoms disproportionate to ILD severity³

- Altered heart sounds
- Jugular venous distention
- Signs of right heart failure
- Ankle swelling/peripheral edema
- Hepatomegaly/ascites
- ILD requiring oxygen

Perform screening tests³

- Pulmonary function tests
- CT scan
- Oxygen saturation
- 6MWD
- Echocardiography

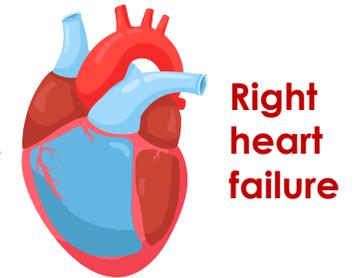
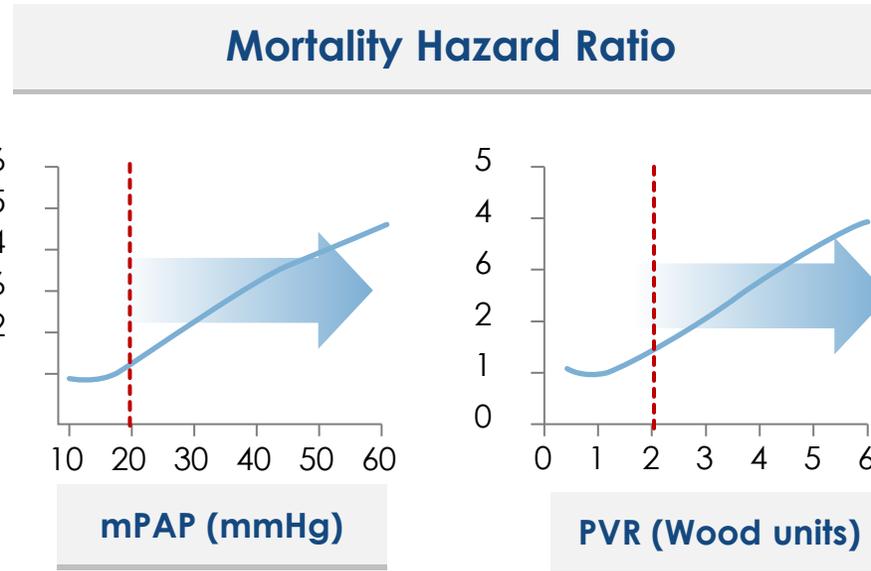
Confirm PH diagnosis³

- Right heart catheterization

1. Humbert M, et al. *Eur Heart J*. 2022;43(38):3618-3731. doi:10.1093/eurheartj/ehac237 2. Watson RD, et al. *BMJ*. 2000;320(7229):236-239. doi:10.1136/bmj.320.7229.236; 3. Rahaghi FF et al. *Chest*. 2022;162(1):145-155. doi:10.1016/j.chest.2022.02.012

Recently updated 7th WSPH hemodynamic definitions

PH	Pre-capillary PH
mPAP >20 mmHg	mPAP >20 mmHg PAWP ≤15 mmHg PVR >2 WU
	PH-ILD¹⁻⁴
	CLD with PH mPAP > 20 mmHg PVR ≥3 WU



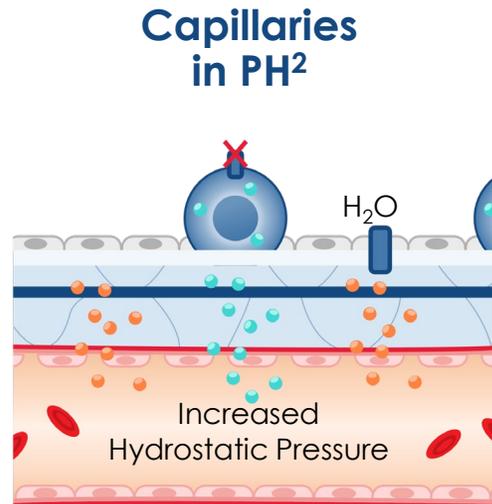
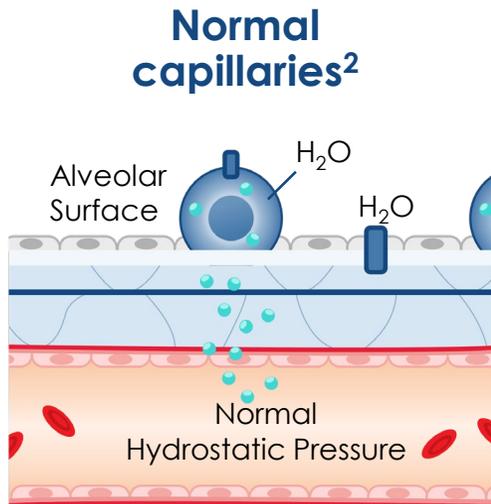
BLPH=borderline pulmonary hypertension; CpcPH=combined post-capillary and pre-capillary pulmonary hypertension; CI=cardiac index; CLD=chronic lung disease; ERS=European Respiratory Society; ESC=European Society of Cardiology; IpcPH=Isolated post-capillary Pulmonary Hypertension mPAP=mean pulmonary arterial pressure; MPH=mild pulmonary hypertension; PAWP=pulmonary arterial wedge pressure; PVR=pulmonary vascular resistance; SPH=severe pulmonary hypertension; WSPH=World Symposium on Pulmonary Hypertension; WU=wood units.

1. Humbert M et al. *Eur Heart J*. 2022;43(38):3618–3731. doi:10.1093/eurheartj/ehac237 2. Simonneau G et al. *Eur Respir J*. 2019;53:1801913. doi:10.1183/13993003.01913-2018 3. Kovacs G, Bartolome S, Denton CP, et al. Definition, classification and diagnosis of pulmonary hypertension. *Eur Respir J* 2024; 64: 2401324 [DOI: 10.1183/13993003.01324-2024]. 4. Modified from Shlobin OA. *Eur Respir J*. 2024;2401200. 5. Piccari L, et al. *Respiration*. 2022;101(8):717-727. doi: 10.1159/000524263

Prostacyclin pathway will remain critical to treatment

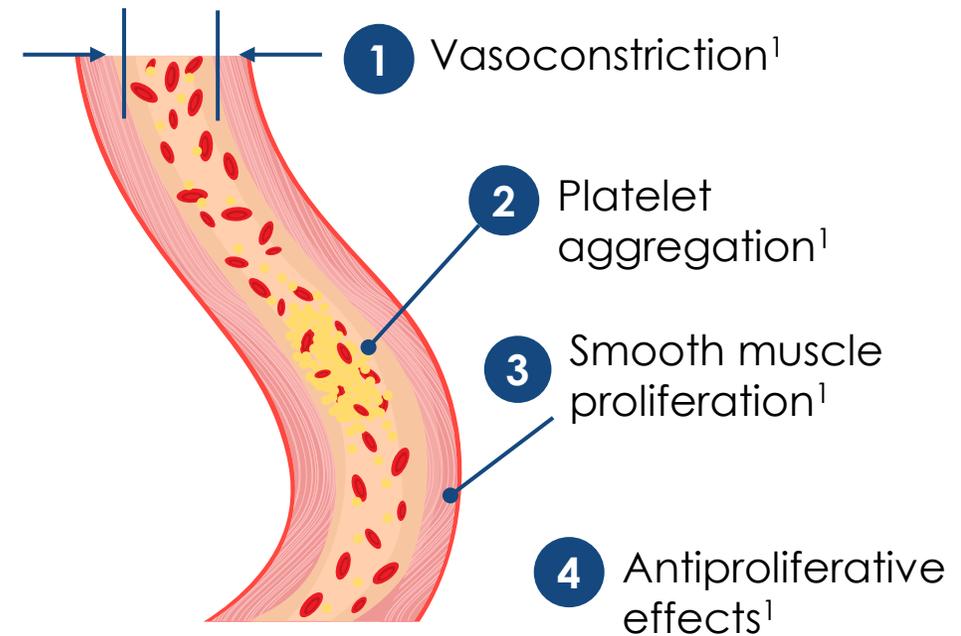
Targets the major pathologic changes that occur in PAH and PH-ILD

Patients with PAH have **decreased levels of endogenous prostacyclin**, a potent vasodilator and **anti-inflammatory agent**¹



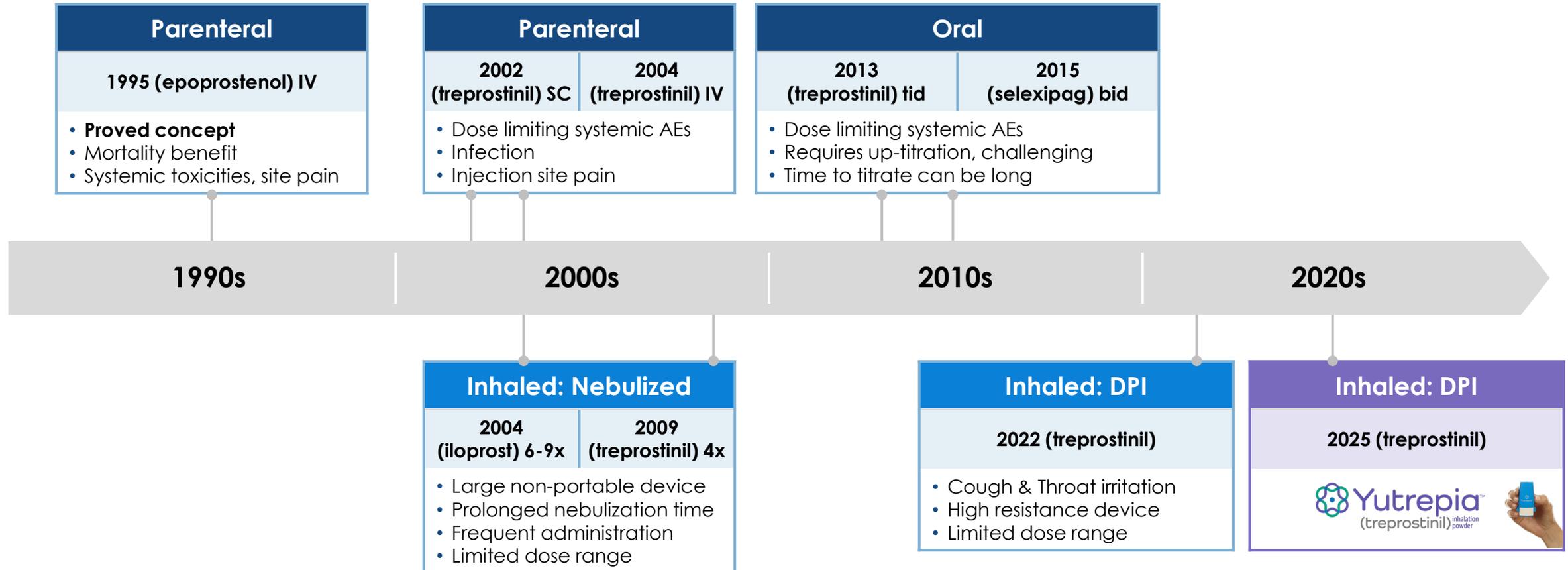
- Endothelial cell breaks
- Impaired permeability
- Protein loss

Prostacyclin mimetics have the potential to **improve blood flow to the lungs, decrease PAP, improve exercise capacity, and/or improve QoL**¹



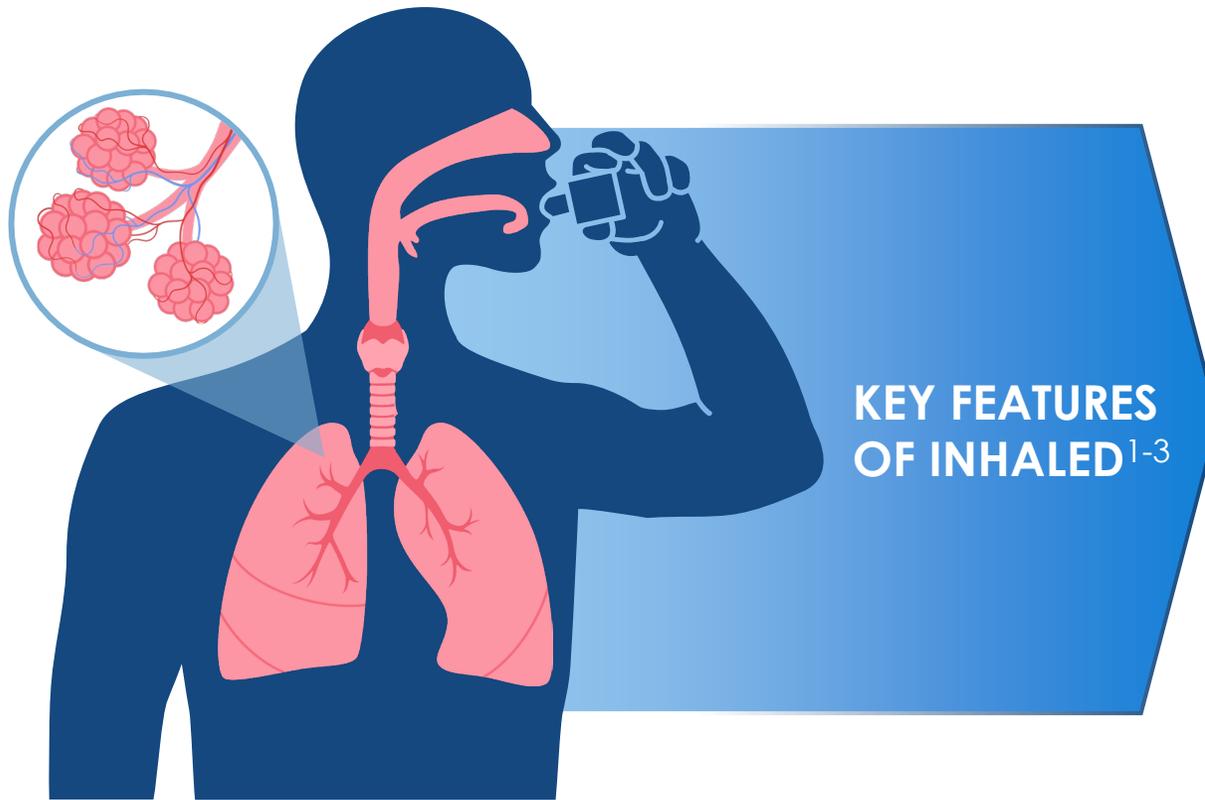
The last 30 years has been an effort in optimizing delivery

Inhaled has potential to maximize the balance of efficacy, tolerability and convenience



Increasing interest to optimize inhaled delivery given the benefits

Local delivery for local disease

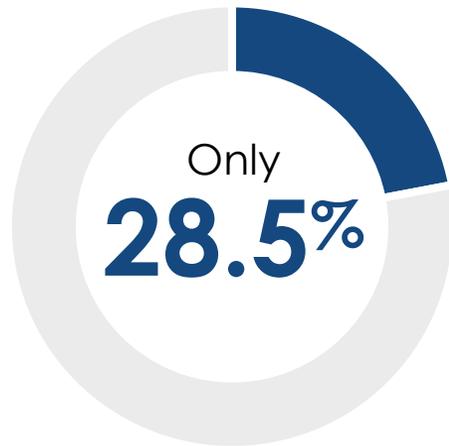


- **Reduced risk of systemic adverse effects**
- **Delivery of drug directly to lungs**
- **Enhanced pulmonary specificity**
- **Higher local drug conc. at lower dose**
- **Improved ventilation/perfusion matching**
- **May aid in patient compliance**

1. Berkenfeld K, et al. *AAPS PharmSciTech*. 2015;16(3):479-490. doi:10.1208/s12249-015-0317 2. Roscigno RF, et al. *Vascul Pharmacol*. 2021;138:106840. doi:10.1016/j.vph.2021.106840
3. Hill NS, et al. *Respir Care*. 2015;60(6):794-805. doi:10.4187/respcare.03927

Higher doses of inhaled treprostinil correlate to better outcomes

Retrospective analysis of 5,000 PAH patients (2009–18)*

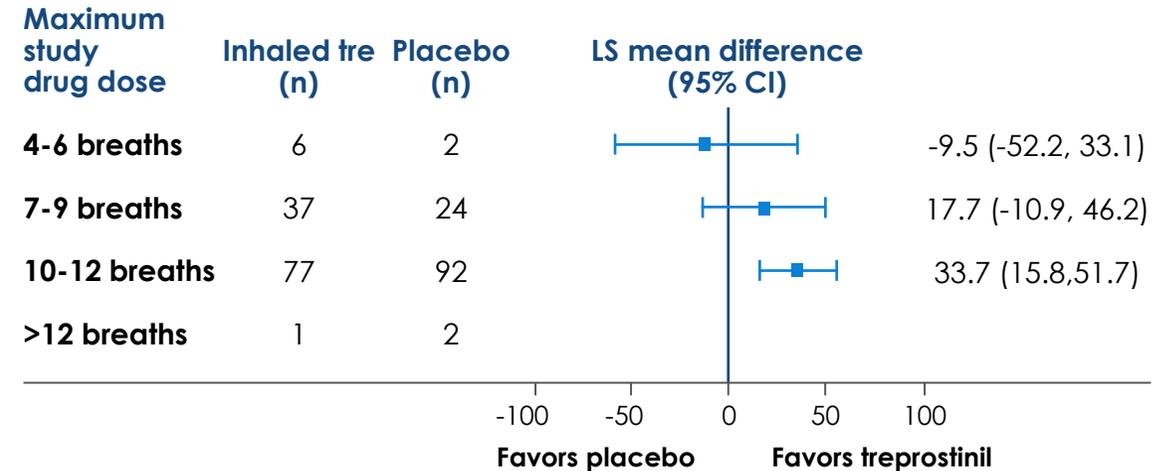


of patients were titrated >9 breaths QID

Patients with 12 or more breaths had better clinical outcomes

*These data are from a Phase 4, retrospective, real-world analysis of patients prescribed inhaled treprostinil solution from a specialty pharmacy database between September 2009 and June 2018. Of the 6709 patients who met all study eligibility criteria, a random sample of 5000 patients was selected for further analysis using simple random sampling with equal probability.
Shapiro S et al. *Pulm Circ.* 2021;11(4):20458940211052228. doi:10.1177/20458940211052228

Analyses of peak δ MWD at week 16 in PH-ILD^{1,2}



10 or more breaths of inhaled treprostinil had favorable improvements in δ MWD

6-minute walk distance (δ MWD); 1. Nathan et al, *CHEST Journal*, February 2023, Vol. 163, Issue 2, P398-406; 2. Supplement to: Waxman et al, *N Engl J Med* 2021;384:325-34

Real-world challenges with Tyvaso DPI in PH-ILD

Prostacyclin naïve patients showed worse tolerability and discontinued faster

Methods

- We prospectively gathered data on patients with PH-ILD who we initiated on treprostinil DPI (either naively or transition from inhaled treprostinil) to analyze safety and tolerability: BNP, 6-minute walk data, spirometry with DLCO, and RVSP and TAPSE on echocardiogram.
- Following transition, we recorded data obtained through routine standard-of-care testing at our center to ensure safety and tolerability in this patient population. (Table 2)
- This study was approved by the IRB at National Jewish Health.



Table 2: Discontinuation Rates of Tyvaso DPI

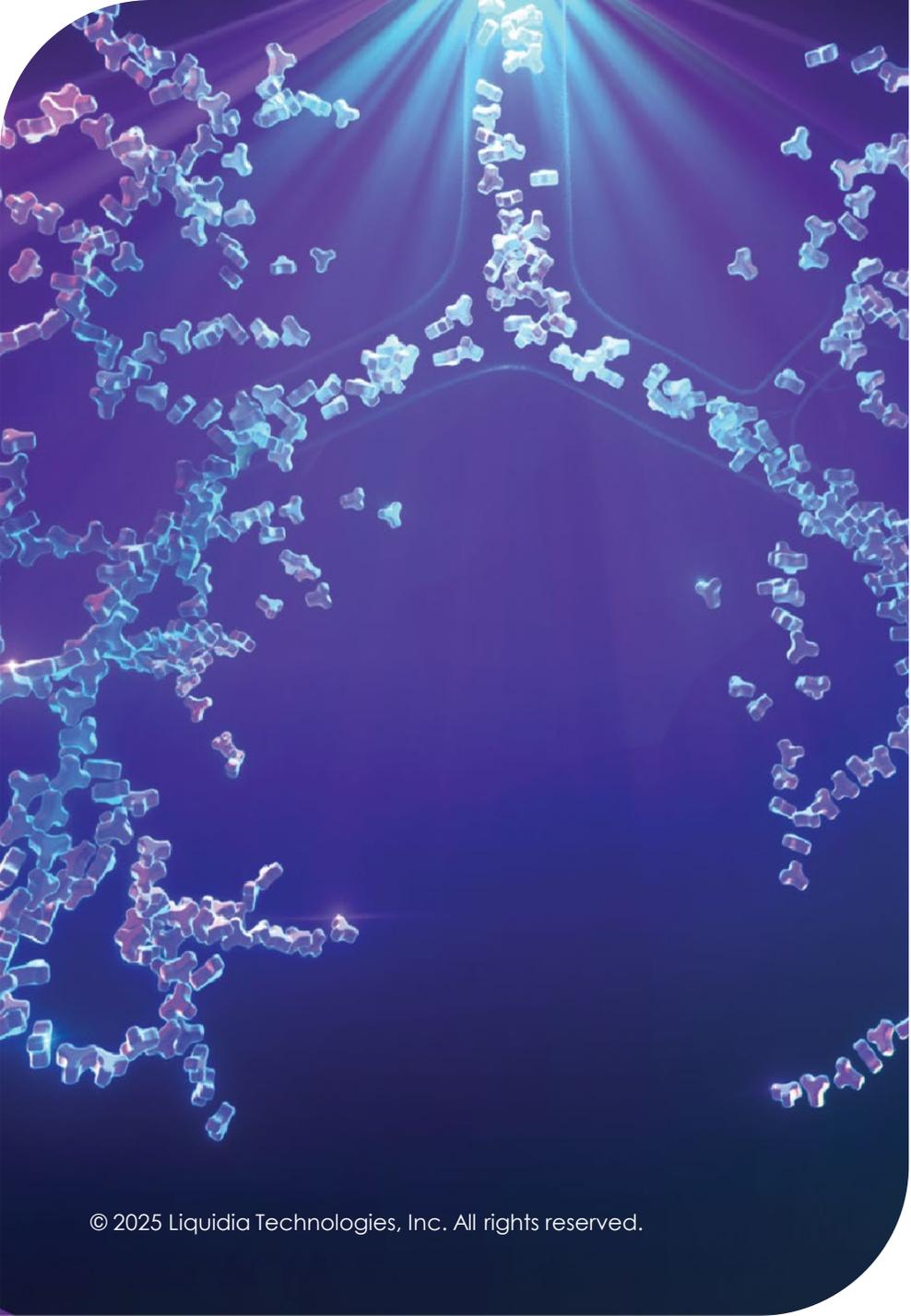
Naïve DPI Discontinuation Rate
26 started, 18 discontinued
11 transitioned to nebulizer 7 discontinued treatment completely
69% discontinuation rate
Discontinued due to cough (5), Hypotension (1), Clinical Worsening (9), Self-discontinuation (2), Death (1)
Transition DPI Discontinuation Rate
22 started, 11 discontinued
7 transitioned back to nebulizer 5 discontinued treatment completely
50% discontinuation rate
Discontinued due to cough (1), Hypotension (1), Clinical Worsening (6), Lung Transplant (2), Death (1)

Results

- 22 patients with PH-ILD were transitioned from the nebulized treprostinil to the treprostinil DPI at equivalent doses between July 2022 and April 2023. The most common form of ILD was CTD-associated ILD. (Table 1)
 - 50% of whom discontinued the treprostinil DPI
 - Average treatment duration on DPI prior to discontinuation was a mean of 195 days, median 223 days (min 13 - max 358)
- 26 patients with PH-ILD started the treprostinil DPI naively, titrating from 16 mcg to 64 mcg between July 2022 and April 2023.
 - 69% of whom discontinued the treprostinil DPI
 - Average treatment duration on DPI prior to discontinuation was a mean of 78 days prior to stopping, median 40 days (min 4 -max 171)
- The most common reasons for discontinuation were clinical worsening and cough
 - Clinical worsening was defined by our group as at least having one of the following: worsening testing upon follow up (PFT, Echo, 6MWT), reported increased shortness of breath, and/or increased oxygen requirement.

Rice et al, Tolerability and Efficacy of Treprostinil Dry-Powdered Inhaler in Patients with Pulmonary Hypertension Related to Fibrosing Interstitial Lung Disease at a Large Tertiary Referral Center, 2023 Pulmonary Hypertension Professional Network Symposium, September 28-30, 2023 [Poster]

Tyvaso® and Tyvaso® DPI are registered trademarks of United Therapeutics Corporation



 **Yutrepia™**
(treprostinil) inhalation powder



YUTREPIA is a dry-powder formulation of treprostinil enabled by PRINT® technology¹ designed for



Enhanced deep-lung delivery¹⁻³



Ease of use with a low-effort device^{1,4-6}



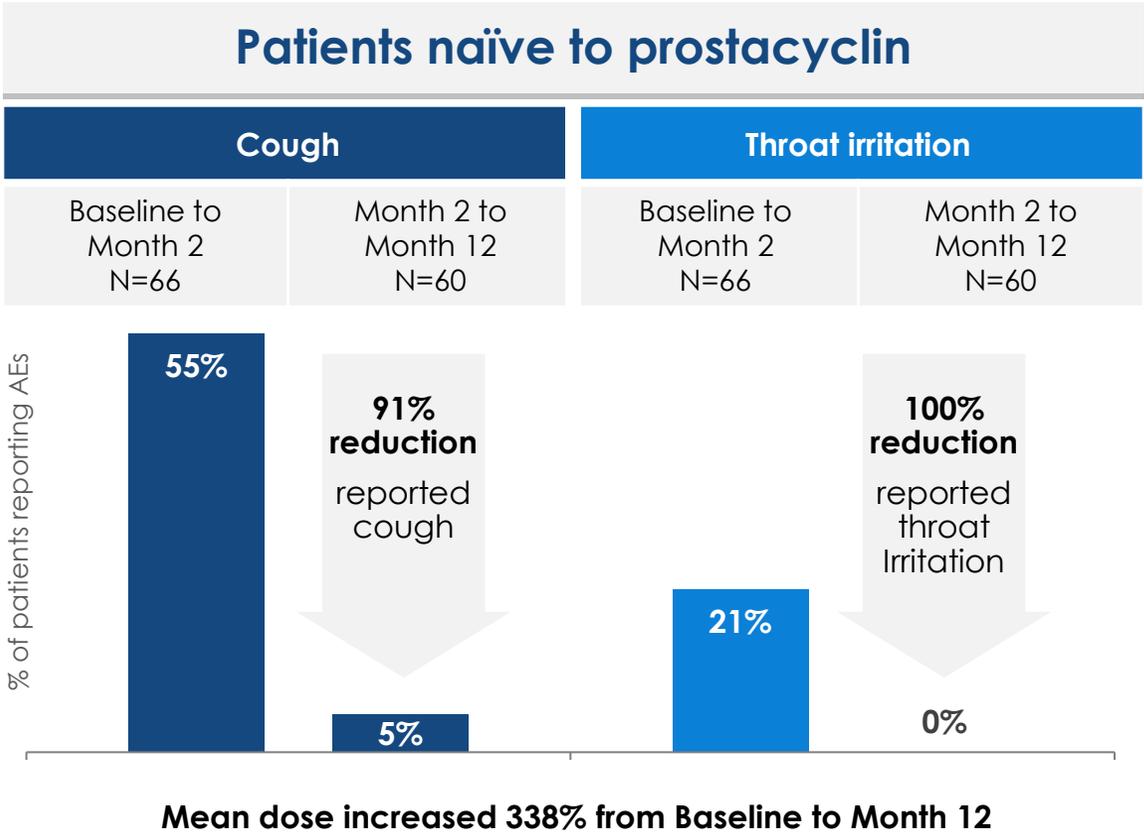
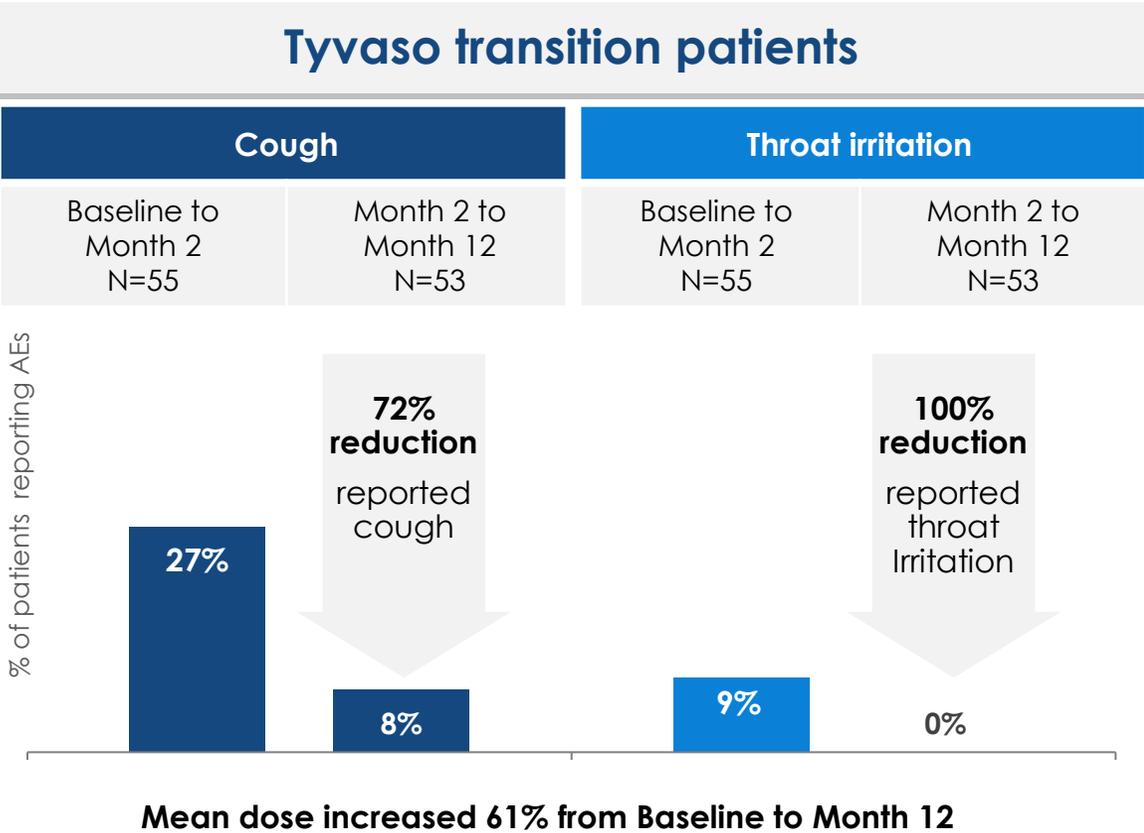
Titration to higher therapeutic doses^{1,7}

PRINT® is a registered trademark of Liquidia Technologies, Inc.

1. Hill NS et al. *Pulm Circ.* 2022;12(3):e12119. doi:10.1002/pul2.12119 **2.** Garcia A et al. *J Drug Deliv.* 2012;2012:941243. doi:10.1155/2012/941243 **3.** Roscigno RF et al. *Vascul Pharmacol.* 2021;138:106840. doi:10.1016/j.vph.2021.106840 **4.** Patel S et al. Robustness of YUTREPIA, a dry-powder inhaled formulation of treprostinil, in patient misuse scenarios. Poster presented at: CHEST 2022 Annual Meeting; October 16-19, 2022; Nashville, TN. **5.** Price D et al. *Multidiscip Respir Med.* 2015;10:36. doi:10.1186/s40248-015-0033-0 **6.** National Health Service Sunderland. Sunderland COPD Inhaler Guide. National Health Service; 2020. **7.** YUTREPIA. Prescribing information. Liquidia Technologies, Inc; 2024.

INSPIRE study reinforced tolerability profile of Yutrepia over time

Important related TEAEs decreased from Month 2 to Month 12^{1,2, 3}

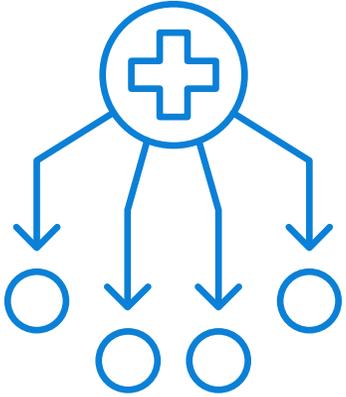


1. Yutrepia Prescribing Information, [Table 2](#): Adverse Reactions Occurring in ≥ 4% of Patients in the INSPIRE Study; 2. Hill et al, "Safety and Tolerability of LIQ861 in Pulmonary Arterial Hypertension (PAH): Results From INSPIRE Study" [Poster](#), ATS 2022; 3. Liquidia Data on File

Standard of care is rapidly changing with new options

Real world experience

Major themes of a changing landscape



- **Dose matters in PAH and PHLD**
- **Inhaled therapy to avoid or delay the need for parenteral**
- **Facilitating transition off parenteral with addition of sotatercept**
- **Inhaled therapy allowing physicians to start prostacyclin earlier**
- **New delivery methods are allowing higher doses of treprostinil**
- **Transitioning from oral prostacyclin pathway drugs to inhaled**

ASCENT Study in PH-ILD @ Week 24

YUTREPIA™ (treprostinil) inhalation powder

Dr. Rajan Sagggar, MD

ASCENT Cohort A

Open-label, Multicenter Study to Evaluate Safety and Tolerability of LIQ861 in Patients with Newly Diagnosed PH-ILD

PATIENTS

- N=54
- Age 18-80 years

KEY INCLUSION CRITERIA

- mPA \geq 30 mmHg & PVR \geq 3 WU or
- mPA \geq 21 mmHg & PVR \geq 3 WU*
- Baseline 6MWD \geq 125m
- CT Chest consistent with ILD or CPFE
- FEV₁/FVC \geq 70%

Week	52-week TREATMENT PERIOD				
	LIQ861+ RS00 Model 8 Dry Powder Inhalation (DPI) Device				
Screening 28 days	D1 Baseline	Week 8	Week 16	Week 24	Week 52
Target Dose (mcg) QID	26.5	132.5	159	185.5	EOS

PRIMARY SAFETY ENDPOINTS

- Incidence of treatment-emergent drug- or device-related AEs and serious AEs

EXPLORATORY ENDPOINTS

- Echocardiogram
- Dyspnea-12, Emphasis 10, simplified cough score
- 6MWD & cardiac effect
- CT CHEST

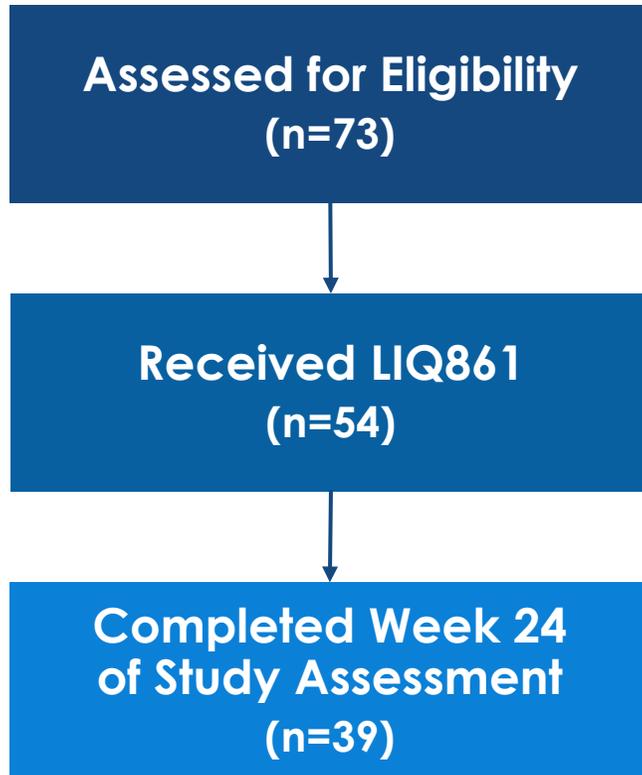
6MWD=6-Minute Walk Distance; CE=cardiac effect; CT=computed tomography; CPFE=combined pulmonary fibrosis and emphysema; FEV₁/FVC=forced expiratory volume in 1 second to forced vital capacity; ILD=interstitial lung disease; WU=wood units; PH-ILD=pulmonary hypertension associated with interstitial lung disease. Reference: LTI-401 Protocol, p.20-22.

*Limited subset of patients

ASCENT (NCT06129240)

Most PH-ILD patients remained on treatment at Week 24

ASCENT: Week 24



Patients	@ Week 8		@ Week 16		@ Week 24	
Completed n (%)	53	(98.1)	43	(79.6)	39	(72.2)
Missed Study visit	-	-	1	(1.9)	-	-
Discontinued	1	(1.9)	10	(18.5)	15	(27.8)
Physician Decision			1	(1.9)	1	(1.9)
Withdrawal of Patient			2	(3.7)	2	(3.7)
Protocol Violation			1	(1.9)	3	(5.6)
Lung Transplant*			3	(5.6)	3	(5.6)
Adverse Event	1	(1.9)	3	(5.6)	6	(11.1)
	• lung neoplasm		• lung neoplasm • chronic pancreatitis • coronavirus		• lung neoplasm (2) • bronchitis (1) • chronic pancreatitis (1) • coronavirus (1) • sudden cardiac death (1)	

*Lung transplant listing was approved upon enrollment per agreement with Sponsor; transplantation was expected to occur ~ 3 months pending donor availability; Liquidia Data on File.

Baseline demographics

ASCENT: Week 24

Naïve PH-ILD	N=54
Age, mean \pm SD, y	68.5 \pm 8.9
Sex, Female (%)	28 \pm 51.9%
Duration of PH Diagnosis, y	0.5 \pm 0.8
Duration of ILD Diagnosis, y	5.1 \pm 5.7
<i>ILD Subtypes (%)</i>	
IIPs	26 (48.1%)
Autoimmune ILDs	19 (35.2%)
HP	1 (1.9%)
Other ILDs	3 (5.6%)
CPFE	5 (9.3%)
<i># Background antifibrotics, n (%)</i>	
Nintedanib	19 (35.2%)
Pirfenidone	4 (7.4%)
<i>Background PH Drugs, n (%): PDE5i</i>	7 (13%)
Peak Inspiratory Flow Rate (L/min)	Mean = 90.6 \pm 22.3 Median = 90.0 Range = 39-120

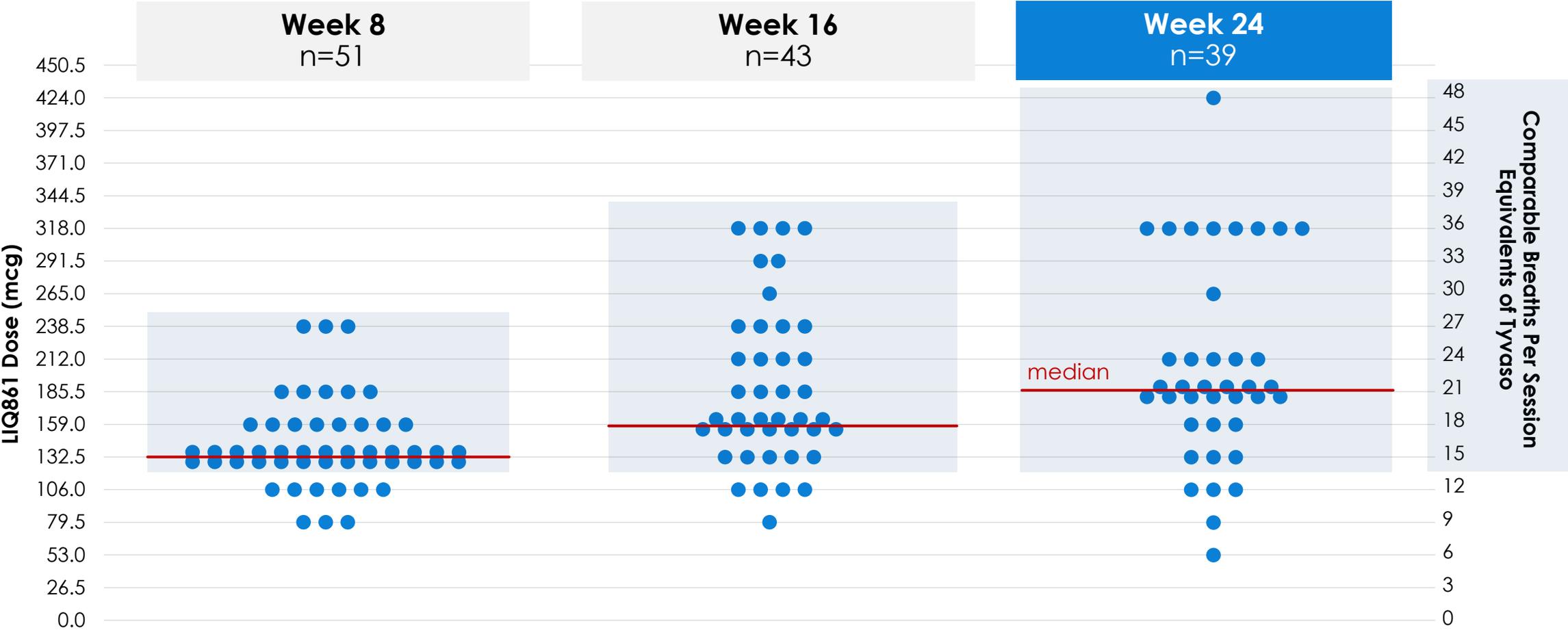
Hemodynamics	Mean \pm SD
mPAP (mmHg)	33.4 \pm 8.4
PCWP (mmHg)	8.6 \pm 3.3
Cardiac Output (L/min)	4.5 \pm 0.9
PVR (WU)	6.0 \pm 2.9

Pulmonary Function Test	Mean \pm SD
FVC, L	2.07 \pm 0.767
FVC (% predicted)	65.9 \pm 20.7
FEV1/FVC	79.6 \pm 17.1
DLCO (% predicted)	36.2 \pm 13.9

Clinical Characteristic	Mean \pm SD
δ MWD (Meters), \pm SD	298.1 \pm 80.3
NTPro-BNP (pg/ml) [GM=210.5]	611.0 \pm 1246.0
Dyspnea-12	11.7 \pm 6.8
EmPHasis-10	24.6 \pm 9.7
Simplified Cough Score	1.3 \pm 0.8

At each time point, more than 80% patients receive ≥ 132.5 mcg

ASCENT: Week 24



Liquidia Data on File; at Week 16, one patient discontinued dosing on clinic visit, which is recorded as 0 mcg

No significant changes in tolerability as dose was titrated

ASCENT: Week 24 (n=54)

Treatment-related TEAEs n (%)	Cumulative TEAEs		
	Week 8	Week 16	Week 24
Median Dose (mcg)	132.5	159	185.5
Cough	23 (42.6)	26 (48.1)	26 (48.1)
Headache	7 (13.0)	10 (18.5)	10 (18.5)
Oropharyngeal pain	3 (5.6)	4 (7.4)	4 (7.4)
Fatigue	2 (3.7)	4 (7.4)	4 (7.4)
Throat Irritation	2 (3.7)	4 (7.4)	4 (7.4)
Diarrhea	2 (3.7)	3 (5.6)	3 (5.6)
Dry Throat	3 (5.6)	3 (5.6)	3 (5.6)

Cough Severity

- Mild n=24 (44.4)
- Moderate n=2 (3.7)

No discontinuations due to cough at Week 24

• No treatment-related SAEs

- Treatment related AE's predominantly mild to moderate

Treatment-emergent adverse event.(TEAE); AE reported if > 5%; severe TEAEs (n=2): respiratory tract irritation (1); hypoxia (1)

Liquidia Data on File

Mean daytime cough scores remained stable as dose was titrated

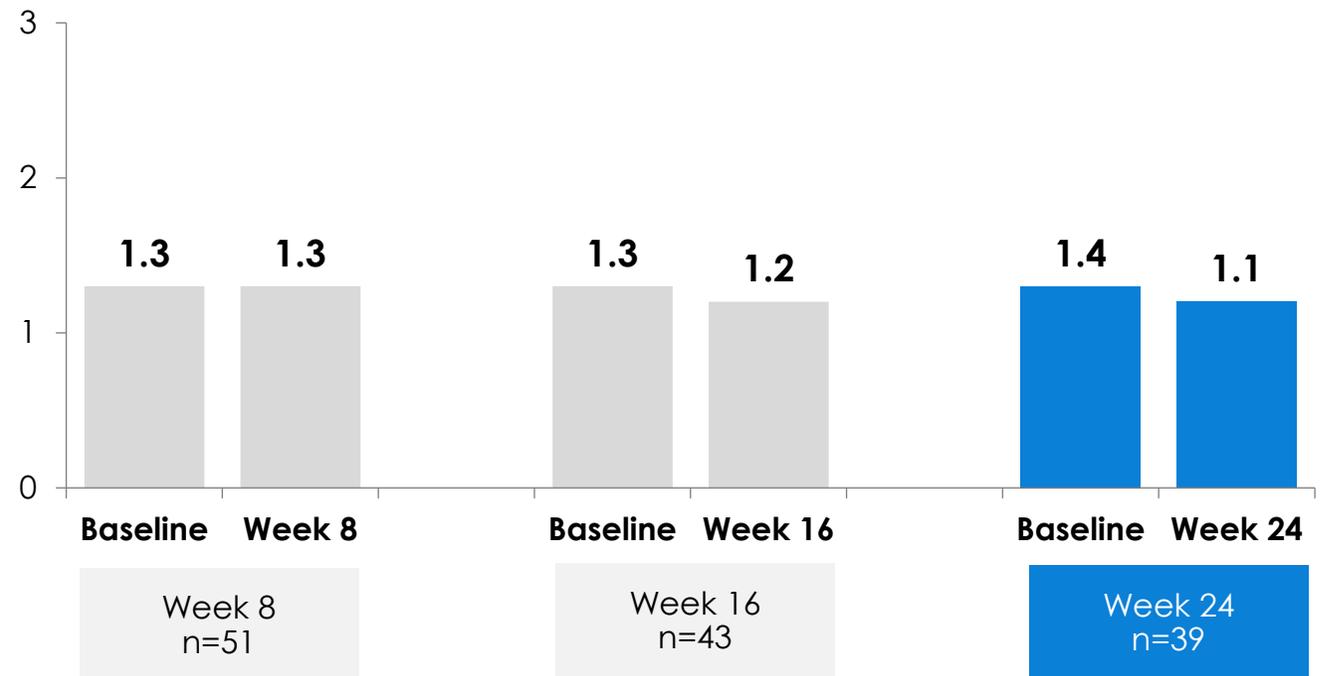
ASCENT: Week 24

Score	Daytime Cough
0	No cough
1	Transient cough occasionally during the daytime
2	Frequent cough mildly affecting daily life
3	Frequent cough severely affecting daily life

Instructions: The patient should circle the score that best describes their cough over the past two weeks

Cough Scores from Baseline to Week 8, Week 16, Week 24

Mean Cough Score



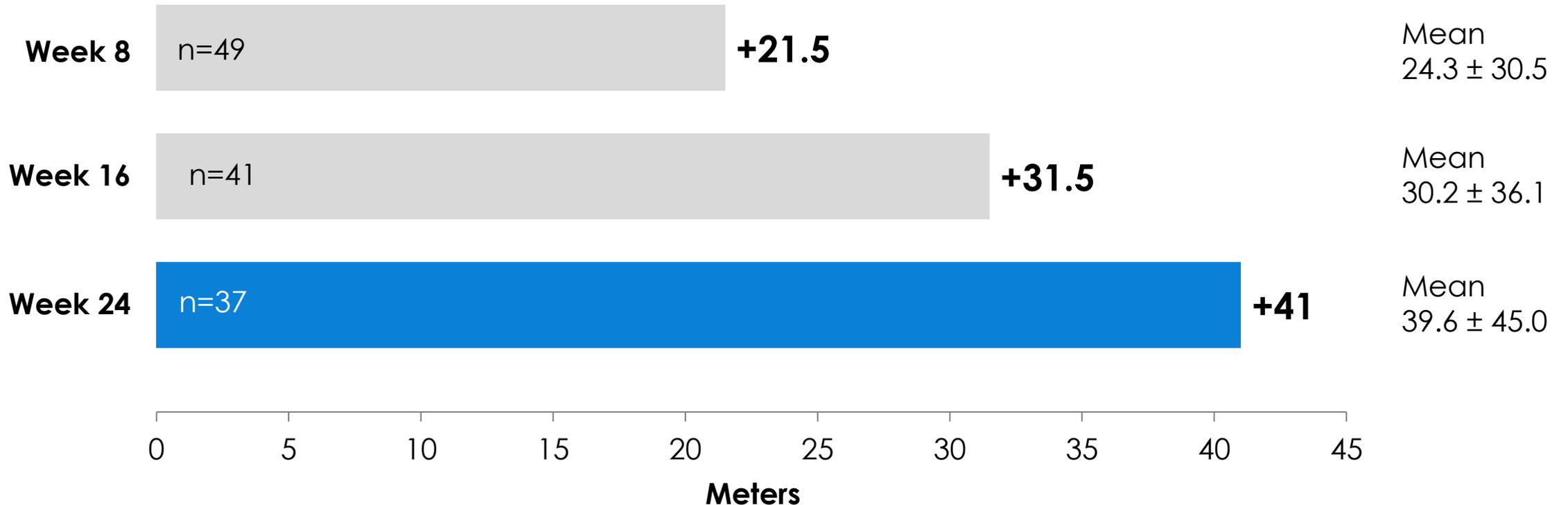
Wang Z, Wang M, Wen S, Yu L, Xu X. Types and applications of cough-related questionnaires. *J Thorac Dis.* 2019 Oct;11(10):4379-4388.

Liquidia data on file

Δ6MWD from baseline continued to increase

ASCENT: Week 24

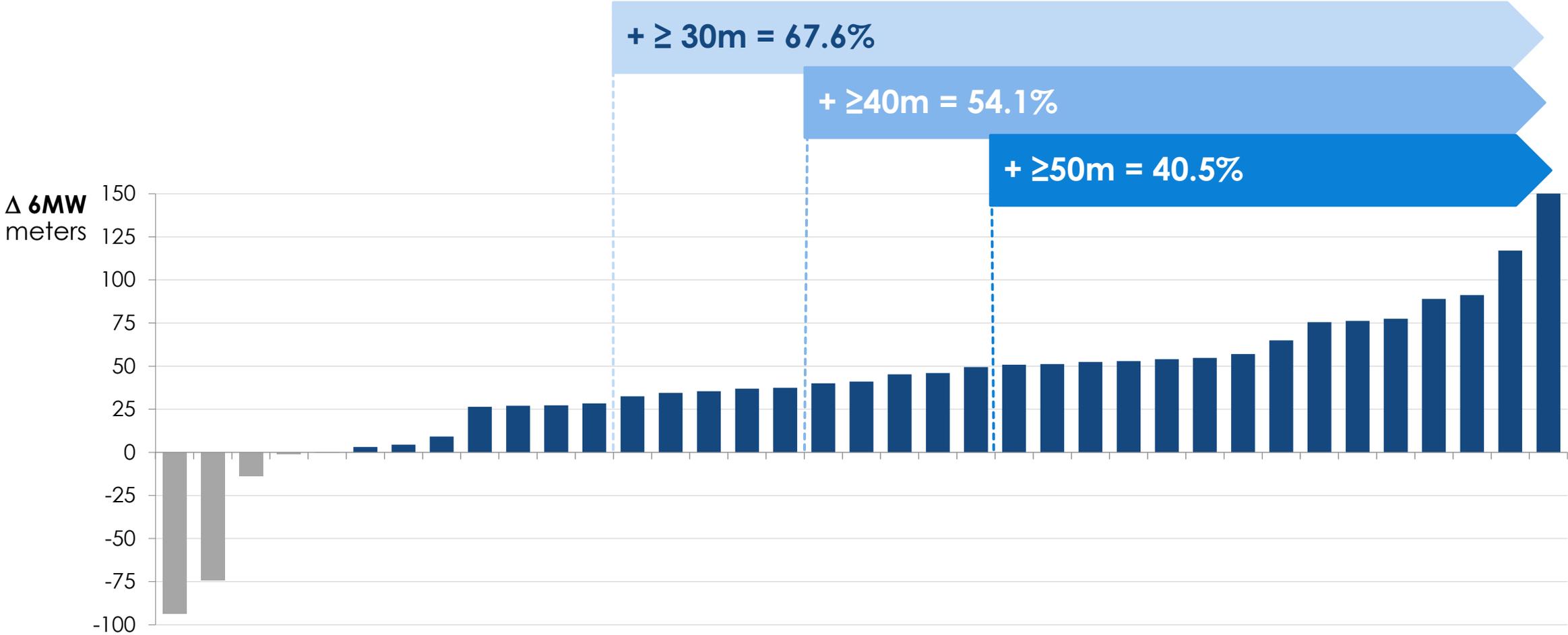
Median change from baseline in 6MWD



Mean ± Standard Deviation, Liquidia data on file

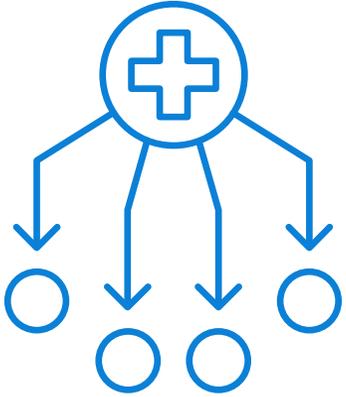
Most patients improved $\Delta 6MWD$ by at least 40 meters (54%)

ASCENT: Week 24 by patient



Summary of ASCENT at Week 24

Thoughts on PH-ILD at UCLA



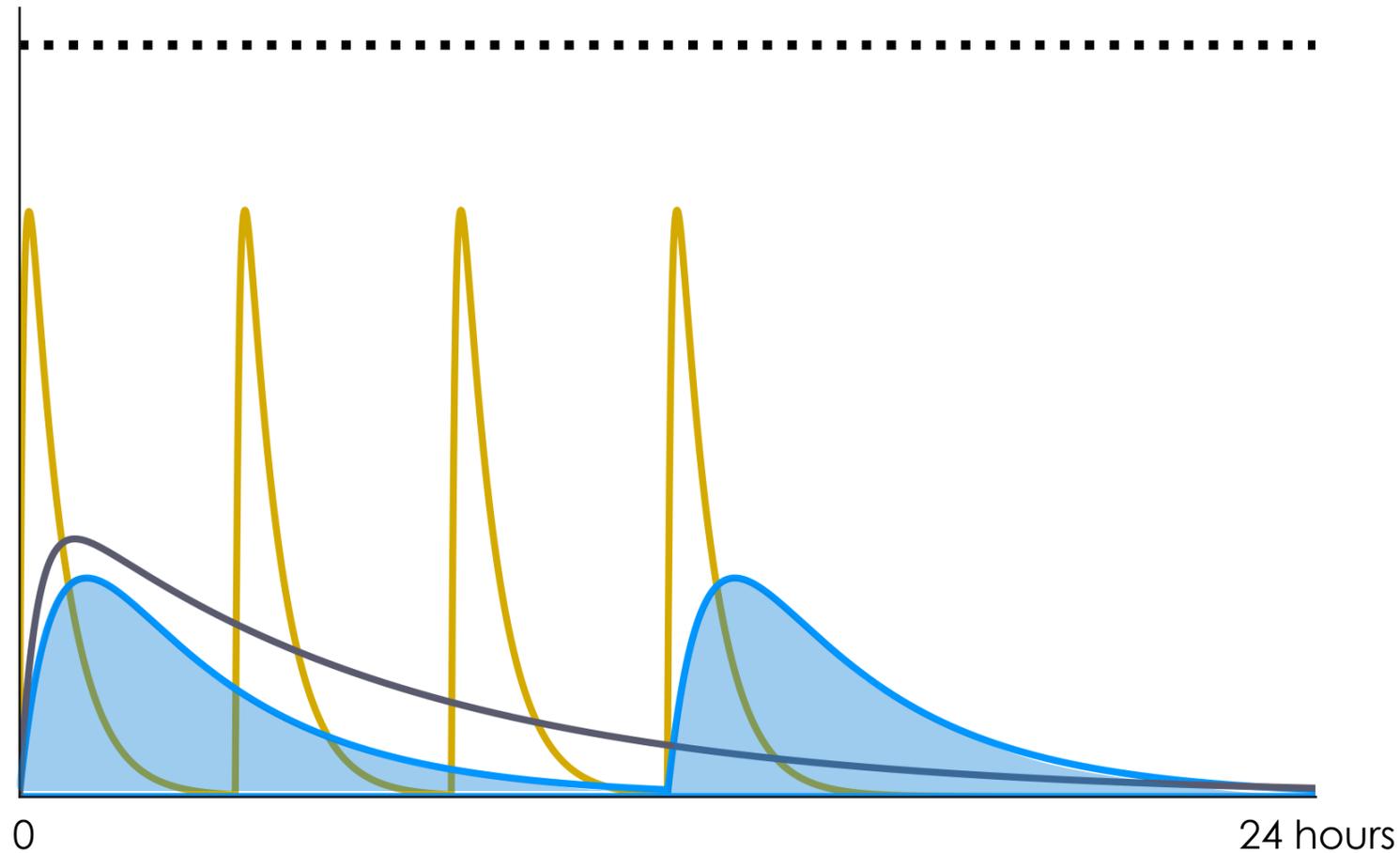
- **Patients continue to titrate to higher doses**
- **No meaningful change in tolerability**
- **Continued improvement in $\Delta 6MWD$ to +41 meters**

L606 (treprostinil liposomal inhalation suspension)

Dr. Rajeev Saggarr

Classic formulation challenge to optimize exposure over 24 hours

Illustrative example



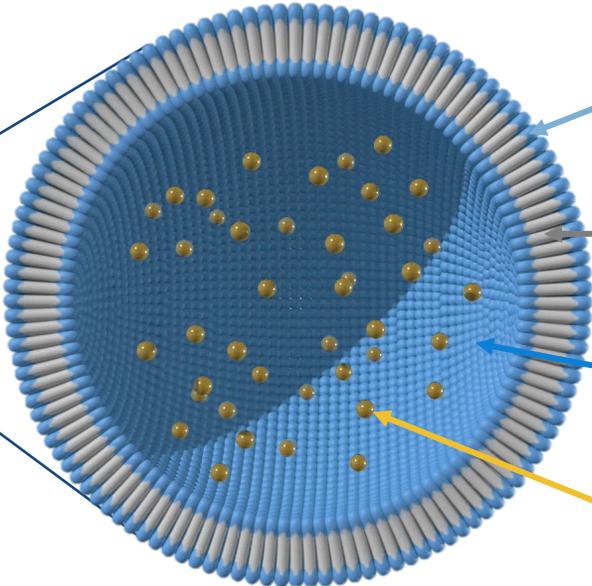
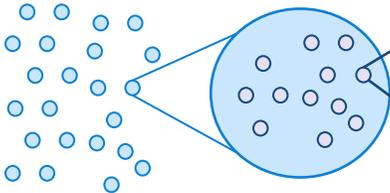
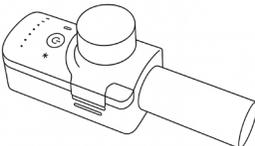
Introduction to L606 Liposome technology

Nanosized liposomes are delivered in micron sizes aerosol droplets

Vibrating Mesh nebulizer

Aerosol Droplet ~4 μm contain liposomes

A single liposome is between 100-140nm



Phospholipids

Bilayer lipid membrane

Inner Aqueous Compartment

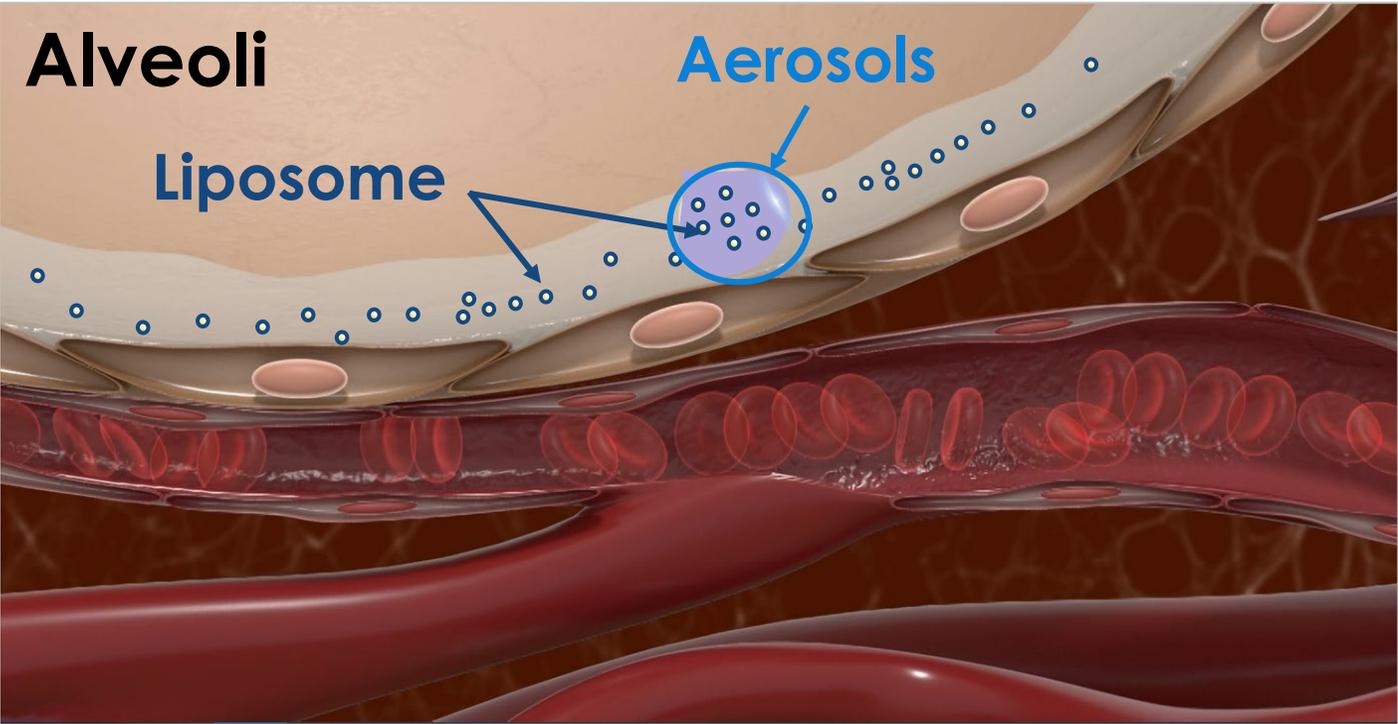
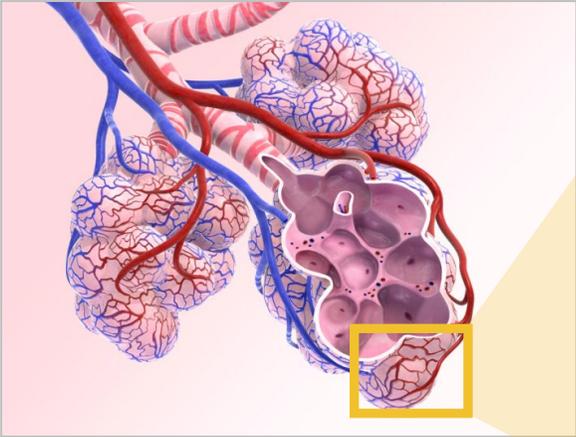
Treprostinil

Pulses to form aerosol droplets

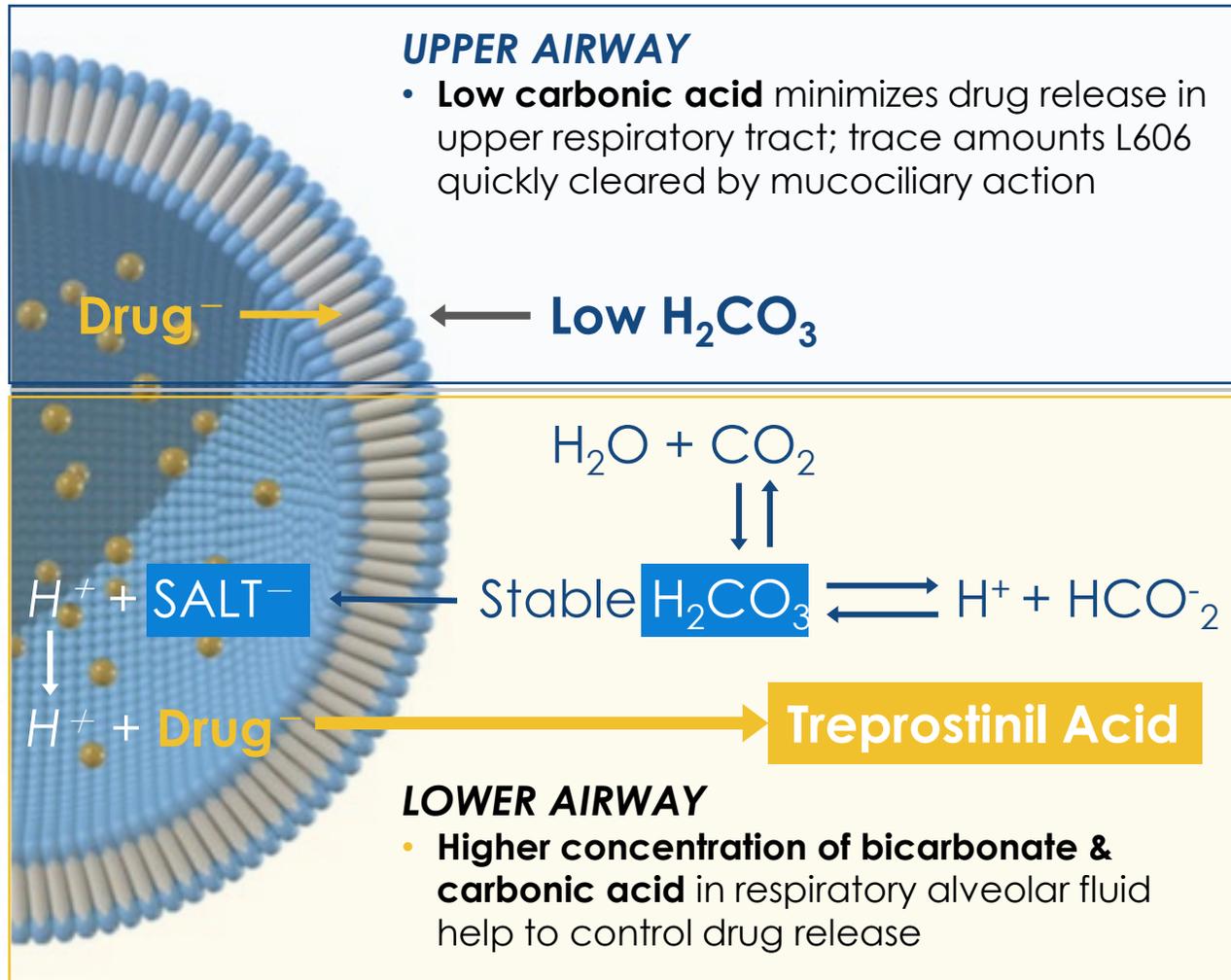
Aerosol Droplets Single droplet contains liposomes

Liposome-containing aerosols are deposited in lung periphery

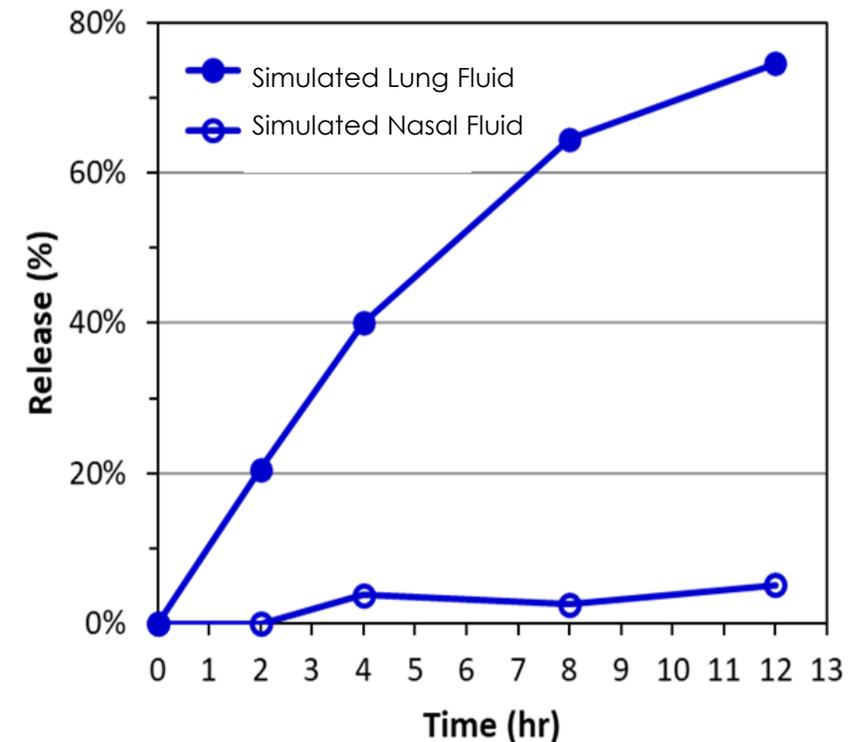
Pulmonary pharmacokinetics of inhaled L606 liposomes



Liposomes extended drug release with salt-sensitive process



● Controlled release rate in lower airway



○ Minimal release rate in upper airway

FOX™ Vibrating Mesh Nebulizer

The FOX device is a handheld, breath-activated, battery-powered inhalation system that delivers nebulized liquid drugs with high performance using a vibrating mesh technology.

The FOX device is suitable for the delivery of small molecules and biologics formulated as solutions or nanosuspensions.



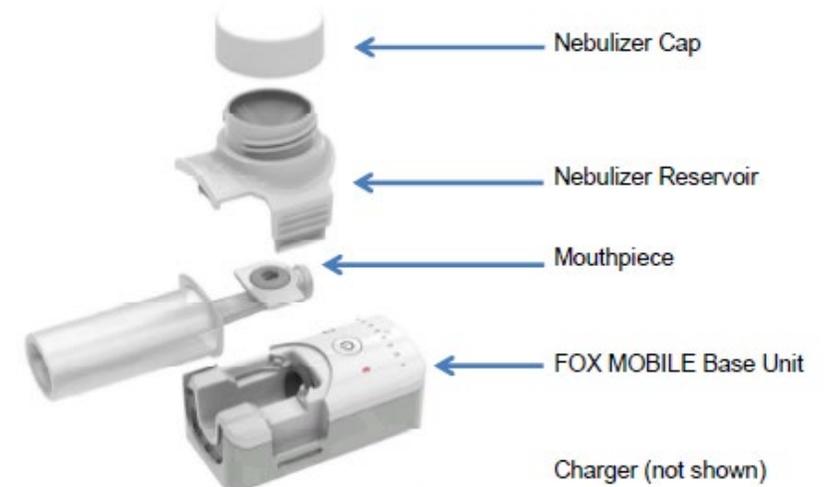
L606 can be rapidly administered in about 1 minute

Highly portable vibrating mesh nebulizer delivers L606 doses of less than 500µL

Older technology



FOX Mobile uses breath-activated technology



L606 drug is supplied in 6 different dose strengths in disposable ampoules that are used in combinations across a wide dose range

Tyvaso® is registered trademark of United Therapeutics Corporation

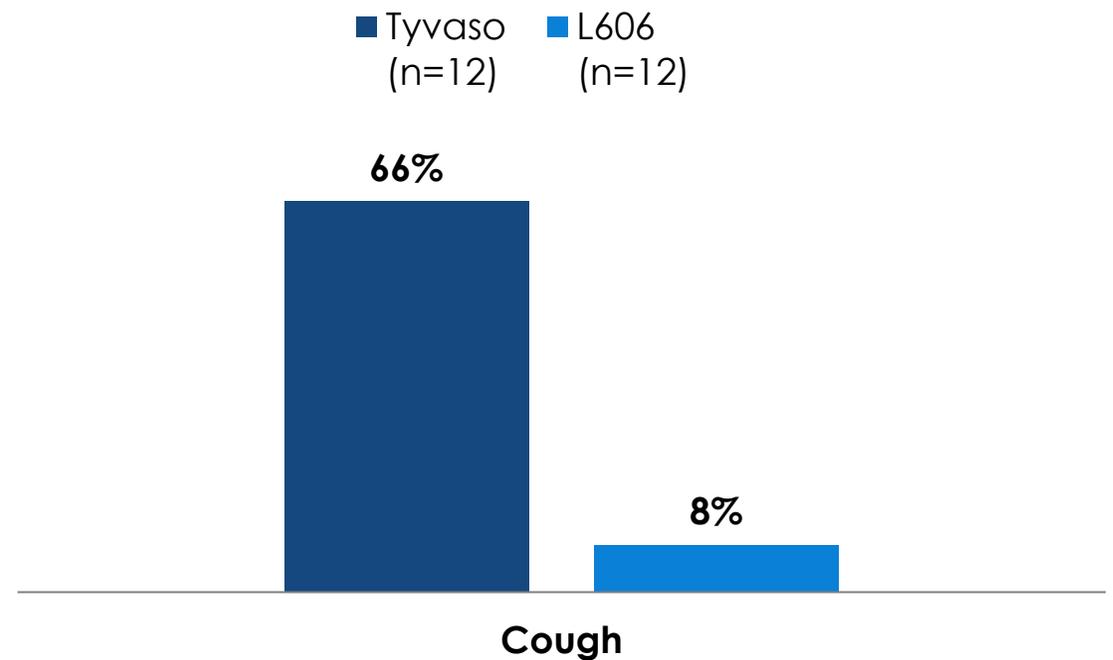
Phase 1 cross-over study supports similar PK, better tolerability

Subjects were healthy volunteers

7.3-fold lower C_{max} with similar AUC

PK Parameters ¹		Tyvaso n = 12	L606 n = 12
Dose	µg	54	51
T _{max}	h (median)	0.18	1.25
C _{max}	pg/mL	1090 (38.4)	140 (24.0)
AUC _{inf}	h*pg/mL	1040 (27.5)	1050 (18.3)
T _{1/2}	h	0.445 (14.6)	4.81 (29.2)
CL/F	L/h	52.0 (27.5)	48.8 (18.3)

Lower cough incidence with L606 in SAD study



*Geometric mean (geometric CV%)

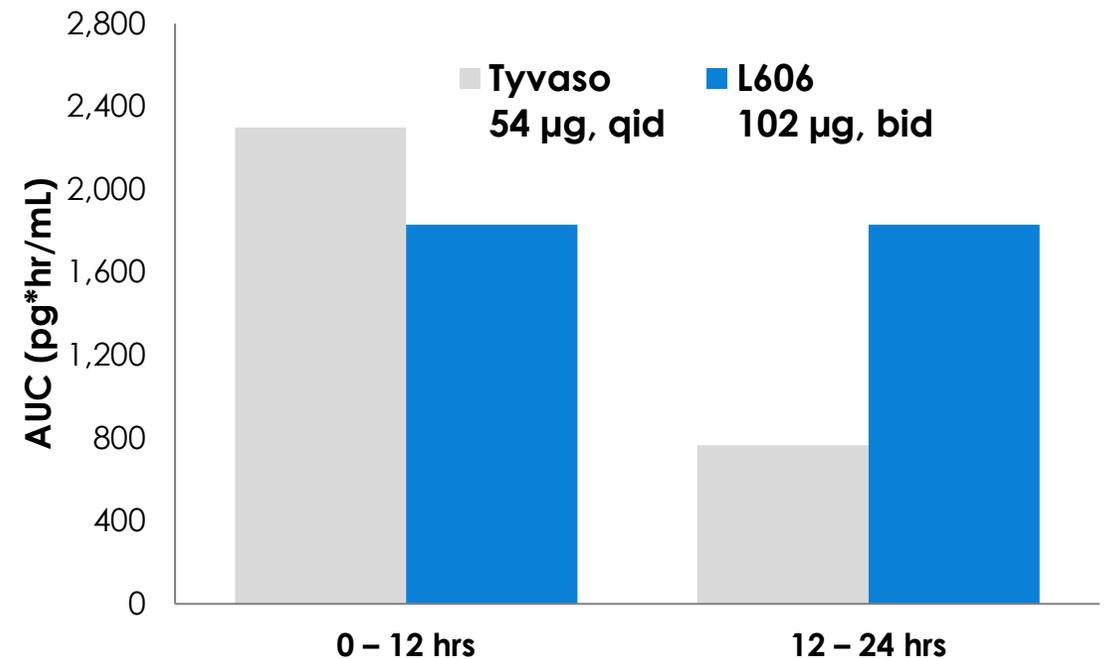
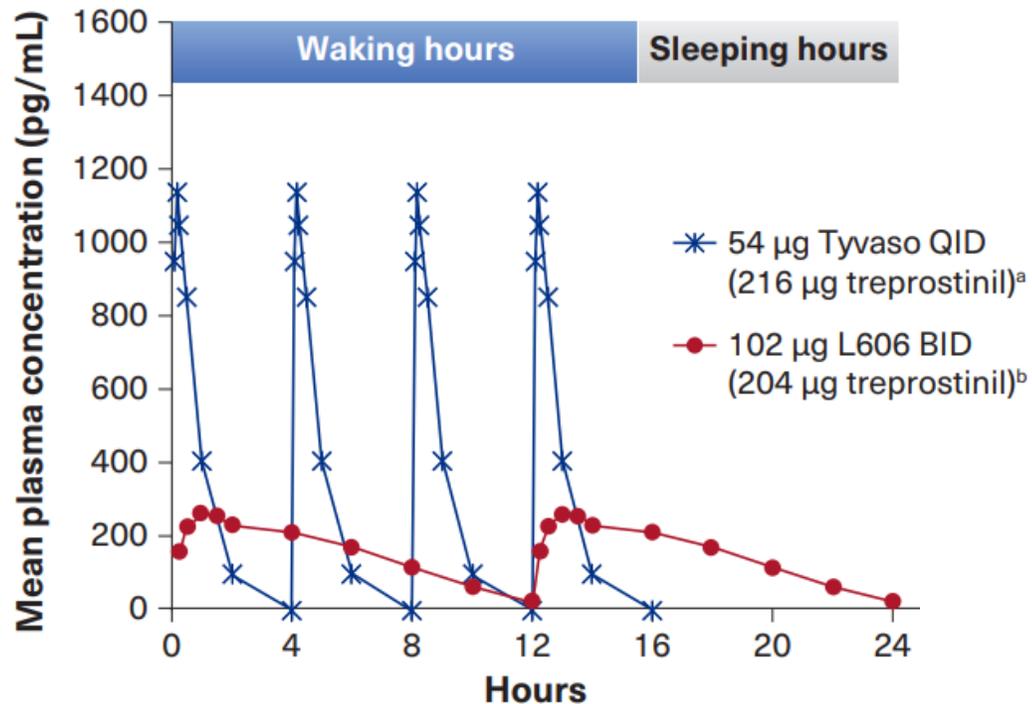
1. Phase 1 (Part B) as published in Tully et al. Clinical Pharmacokinetics of an Extended-Release Formulation of Inhaled Liposomal Treprostinil (L606) to Reduce Dosing Frequency [POSTER]. Pulmonary Vascular Research Institute (PVRI) 2024 Annual Congress; 2024 Feb 2, London.

L606 provides more consistent exposure over 24 hours with bid dosing

Provides therapeutic levels during sleeping hours

Modeled AUC of daily dosing (24hrs) is similar

L606 offers more consistent exposure



Tully et al. Clinical Pharmacokinetics of an Extended-Release Formulation of Inhaled Liposomal Treprostinil (L606) to Reduce Dosing Frequency, Pulmonary Vascular Research Institute (PVRI) 2024 Annual Congress; 2024 Feb 2, London.

Clinical Data from U.S. Open-Label Study

L606 (treprostinil liposomal inhalation suspension)

Dr. Ricardo Restrepo-Jaramillo

Phase 3 study evaluating safety & tolerability in PAH or PH-ILD patients

Initiated by Pharmosa in 2021 and amended and led by Liquidia since July 2023

PATIENTS

- N=28
- Age 18-80 years

KEY INCLUSION CRITERIA

Cohort A (Transitions):

- PAH and PH-ILD
- Stable on Tyvaso

Cohort B (Naïve):

- PAH
- Naïve to prostacyclin

	48-week TREATMENT PERIOD L606 + Vibrating mesh nebulizer				
Week	Week 2,4,8	Week 12	Week 24	Week 36	Week 48
Endpoints	●	● ▲	● ▲	● ▲	● ▲

● PRIMARY SAFETY ENDPOINTS

- Incidence of treatment-emergent drug- or device-related AEs and serious AEs

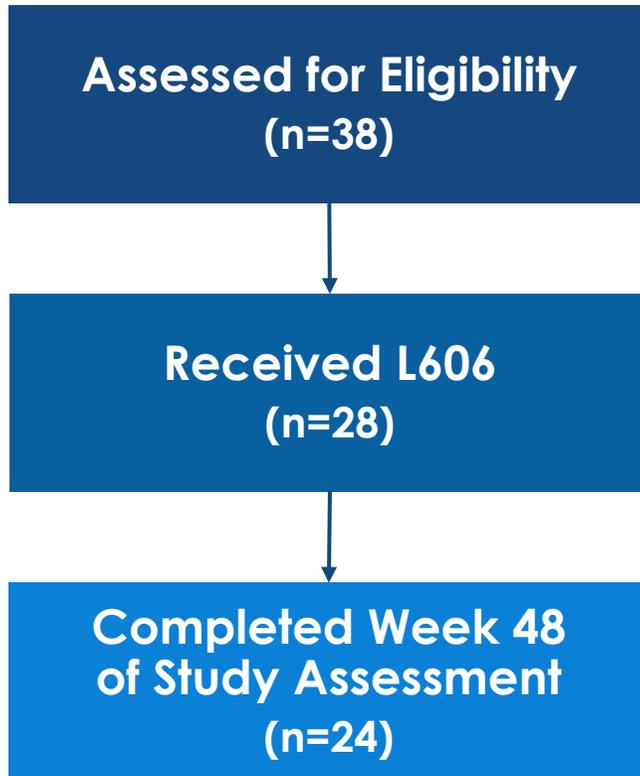
▲ EXPLORATORY ENDPOINTS

- Change in peak and trough 6MWD
- Treatment Satisfaction Questionnaire for Medication (TSQM) transition patients only

Link to [clinicaltrials.gov](https://clinicaltrials.gov/ct2/show/study/NCT04691154): [NCT04691154](https://clinicaltrials.gov/ct2/show/study/NCT04691154)

Most patients continued treatment with L606

L606 Open-Label (U.S.): Week 48



Patients Completed Week 48 Visit, n (%)

Completed	24 (85.7)
Discontinued	4 (14.3)
Protocol Violation	1
Adverse Event	3

- Chest discomfort (related, moderate)
- Dyspnea (possibly related, mild)
- Respiratory failure, transplant (not related, severe)

Participant demographics show naïve patients earlier in disease

L606 Open-Label (U.S.): Week 48

		Cohort A Tyvaso Transition			Cohort B PCY Naïve	Overall
		PAH (N=18)	PH-ILD (N=5)	Total (N=23)	PAH (N=5)	(N=28)
Age , years median (min, max)		64.5 (21, 75)	66 (56, 74)	65 (21, 75)	57 (29, 60)	61 (21, 75)
Female , n (%)		15 (88.9)	1 (20.0)	16 (69.6)	5 (100)	21 (75.0)
White , n (%)		15 (83.3)	4 (80.0)	19 (82.6)	3 (60.0)	22 (78.6)
BMI , kg/m ² median (min, max)		27.3 (22.3, 39.7)	32.6 (27.3, 36.8)	28.3 (22.3, 39.7)	26.1 (21.7, 33.6)	27.4 (21.7, 39.7)
Duration of Dx , yr median (min, max)		7.8 (0.6, 21.2)	3.9 (1.2, 9.9)	6.95 (0.6, 21.2)	1.05 (0.4, 15.5)	6.57 (0.4, 21.2)
No. PAH Rx , n (%)	1	-	1 (20.0)	1 (4.3)	3 (60.0)	4 (14.3)
	2	1 (5.6)	4 (80.0)*	5 (21.7)	2 (40.0)	7 (25.0)
	3	17 (94.4)	-	17 (73.9)	-	17 (60.7)
NYHA Class n (%)	II	11 (61.1)	2 (40.0)	13 (56.5)	2 (50.0)	15 (55.6)
	III	7 (38.9)	3 (60.0)	10 (43.5)	2 (50.0)	12 (44.4)

*PH-ILD Patients on 2 PAH medicines included inhaled treprostinil & PDE5; body mass index (BMI)

Baseline disease characteristics show stable population of patients

L606 Open-Label (U.S.): Week 48

	Cohort A Tyvaso Transition			Cohort B PCY Naïve	Overall
	PAH (N=18)	PH-ILD (N=5)	Total (N=23)	PAH (N=5)	(N=28)
6MWD , meters* median (min, max)	390.5 (219.7, 525)	396 (231.7, 446)	396 (219.7, 525)	395 (240, 490)	395.5 (219.7, 525)
NT-proBNP , pg/mL median (min, max)	176 (41, 1194)	152 (33, 452)	168 (33.3, 1194)	206.5 (45, 946)	168 (33, 1194)

*6MWD is comparable to other contemporaneous PAH studies at ~400m

Six Minute Walk Distance (6MWD), N-terminal pro B-type natriuretic peptide (NT-proBNP), forced expiratory volume (FEV), forced vital capacity (FVC)

Source: Liquidia Data on File

Well tolerated with no treatment related SAEs or dose modifications

L606 Open-Label (U.S.): Week 48

	Cohort A Tyvaso Transition			Cohort B PCY Naïve	Overall
	PAH (N=18)	PH-ILD (N=5)	Total (N=23)	PAH (N=5)	(N=28)
Any TEAE	15 (83.3)	5 (100)	20 (87.0)	5 (100)	25 (89.3)
Treatment Related TEAE	5 (33.3)	2 (40.0)	8 (34.8)	2 (40.0)	10 (35.7)
Serious TEAE (SAE)	3 (16.7)	2 (40.0)	5 (21.7)	1 (20.0)	6 (21.4)
Treatment Related SAE	-	-	-	-	-
Treatment related TEAE Led to Dose Reduction	1 (5.6)		1 (4.3)	-	1 (3.6)
Treatment Discontinuation	1 (5.6)	1 (20.0)	2 (8.7)	1 (20.0)	3 (10.7)
Death	-	-	-	-	-

Six Minute Walk Distance (6MWD), forced vital capacity (FVC), forced expiratory volume (FEV), N-terminal pro B-type natriuretic peptide (NT-proBNP)
Liquidia data on file

Most patients titrated to doses comparable to >12 bps Tyvaso QID

L606 Open-Label (U.S.): Week 48

Week 12 (n=26)

L606 Doses

360mcg

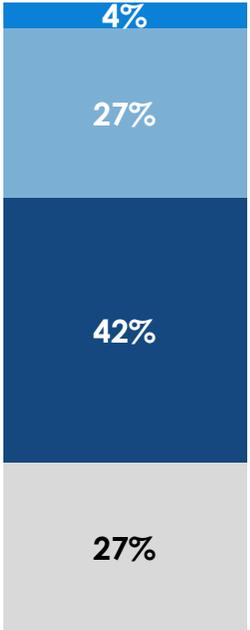


**Median
169 mcg**

comparable
to ~13 bps



42mcg



L606 comparable
dose equivalents to
Tyvaso bps QID

- 25+ bps
- 19-24 bps
- 13-18 bps
- 5-12 bps

Week 12

Week 48 (n=24)

L606 Doses

378mcg

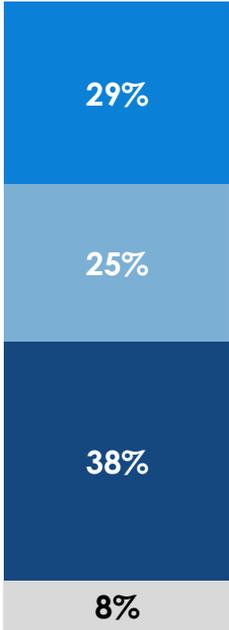


**Median
229 mcg**

comparable
to ~19 bps



42 mcg



L606 comparable
dose equivalents to
Tyvaso bps QID

- 25+ bps
- 19-24 bps
- 13-18 bps
- 5-12 bps

Week 48

Breaths per session (bps) QID, Liquidia data on file

L606 is very tolerable with 4 (14%) patients reporting related cough

L606 Open-Label (U.S.): Week 48

Most Common TEAEs Reported > 10 %	TEAE		Related TEAE	
	%	n	%	n
Cough	32.1	9	14.3	4
Dyspnea	28.6	8	3.6	1
Fatigue	21.4	6	3.6	1
Dizziness	21.4	6	3.6	1
Nausea	10.7	3	3.6	1
Pruritis	10.7	3	3.6	1
COVID 19	17.9	5	-	-
Edema	14.3	4	-	-
Nasopharyngitis	10.7	3	-	-
Pneumonia	10.7	3	-	-
Upper Respiratory Tract Infection	10.7	3	-	-
NT-pro-BNP increased	10.7	3	-	-
Arthralgia	10.7	3	-	-
Back Pain	10.7	3	-	-
Hypotension	10.7	3	-	-

Cough Severity	Related TEAE	
	%	n
Mild	14.3	4
Moderate	-	-
Leading to DC	-	-

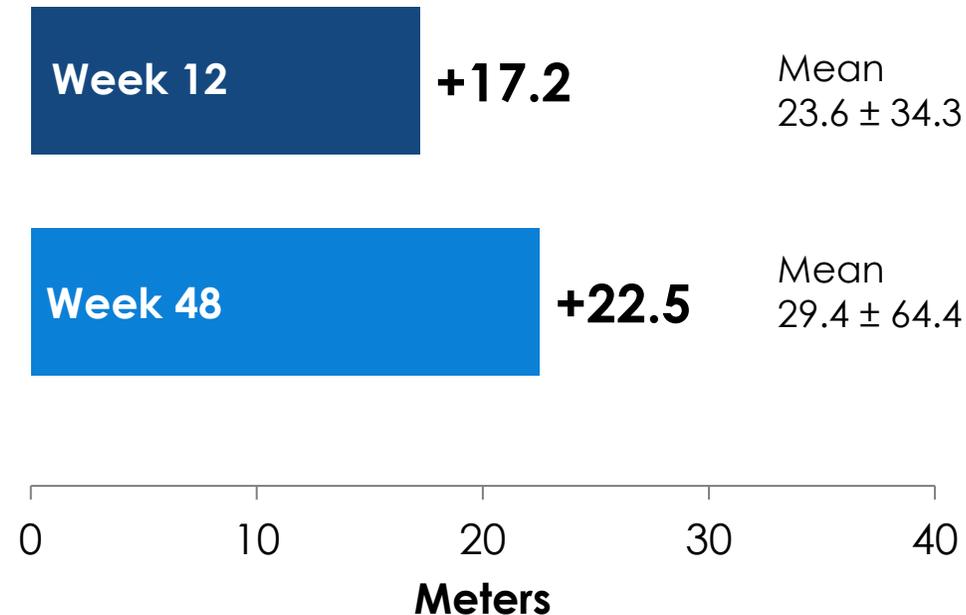
Liquidia data on file

Most patients maintained or improved 6MWD over time

L606 Open-Label (U.S.): Week 48

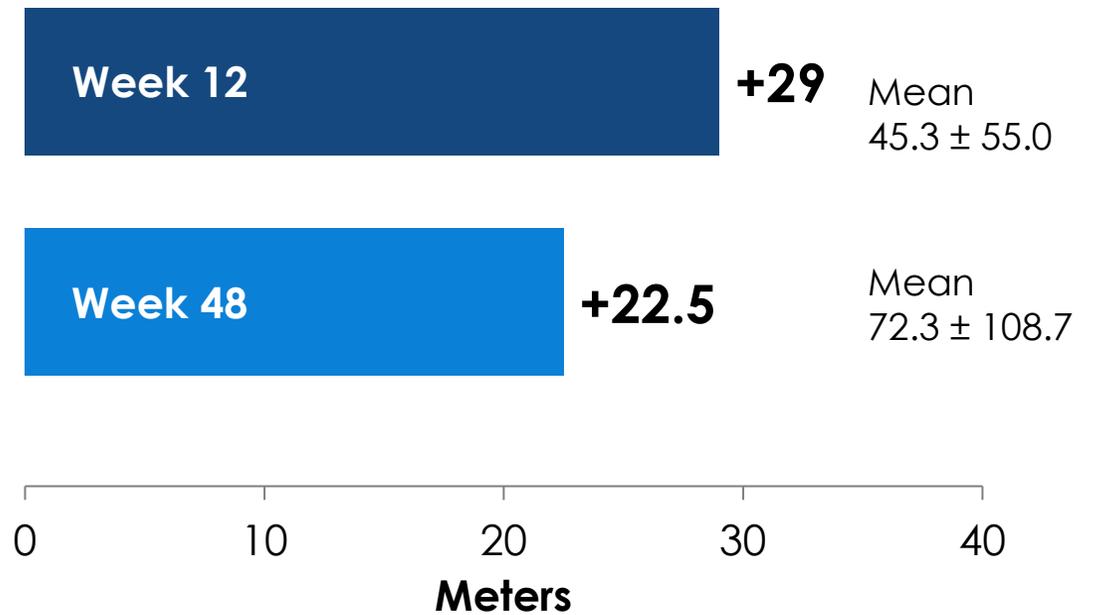
All participants (N=24)

Median change from baseline in 6MWD at peak



Cohort B: PCY Naïve (N=4)

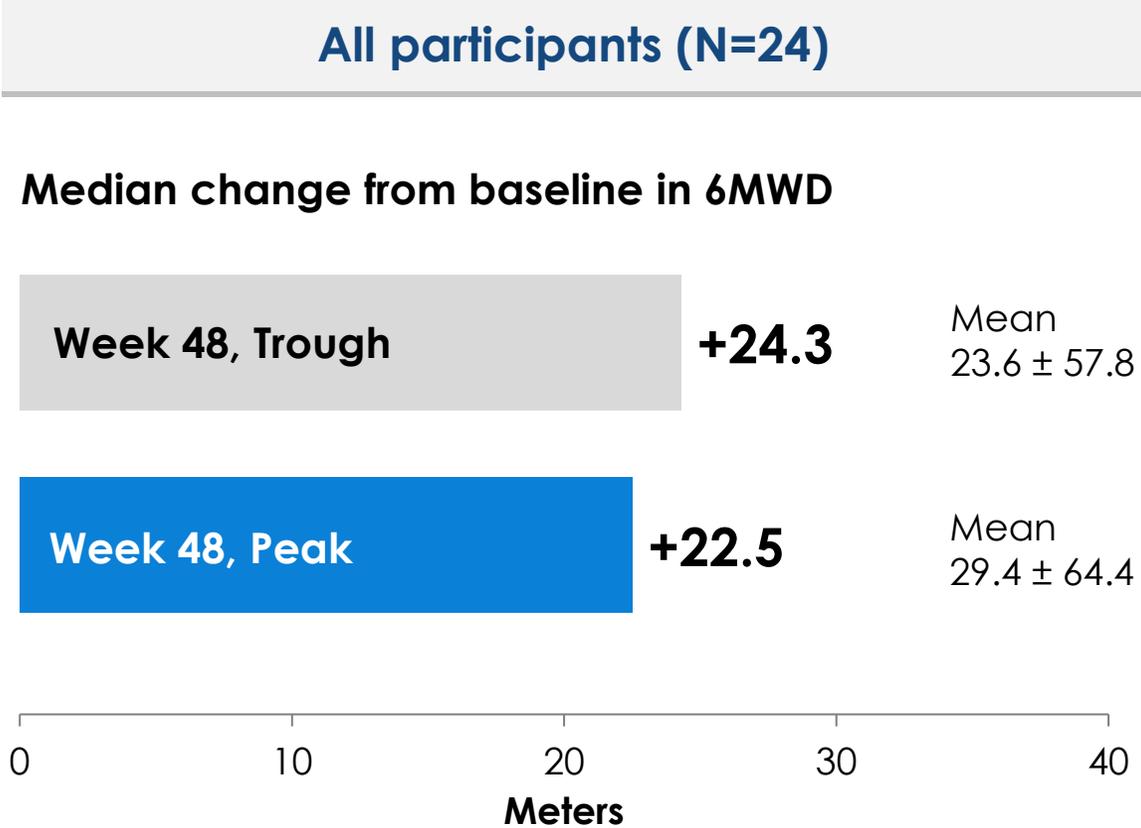
Median change from baseline in 6MWD at peak



Mean ± Standard Deviation, Liquidia data on file

Observed minimal variability between peak and trough measures

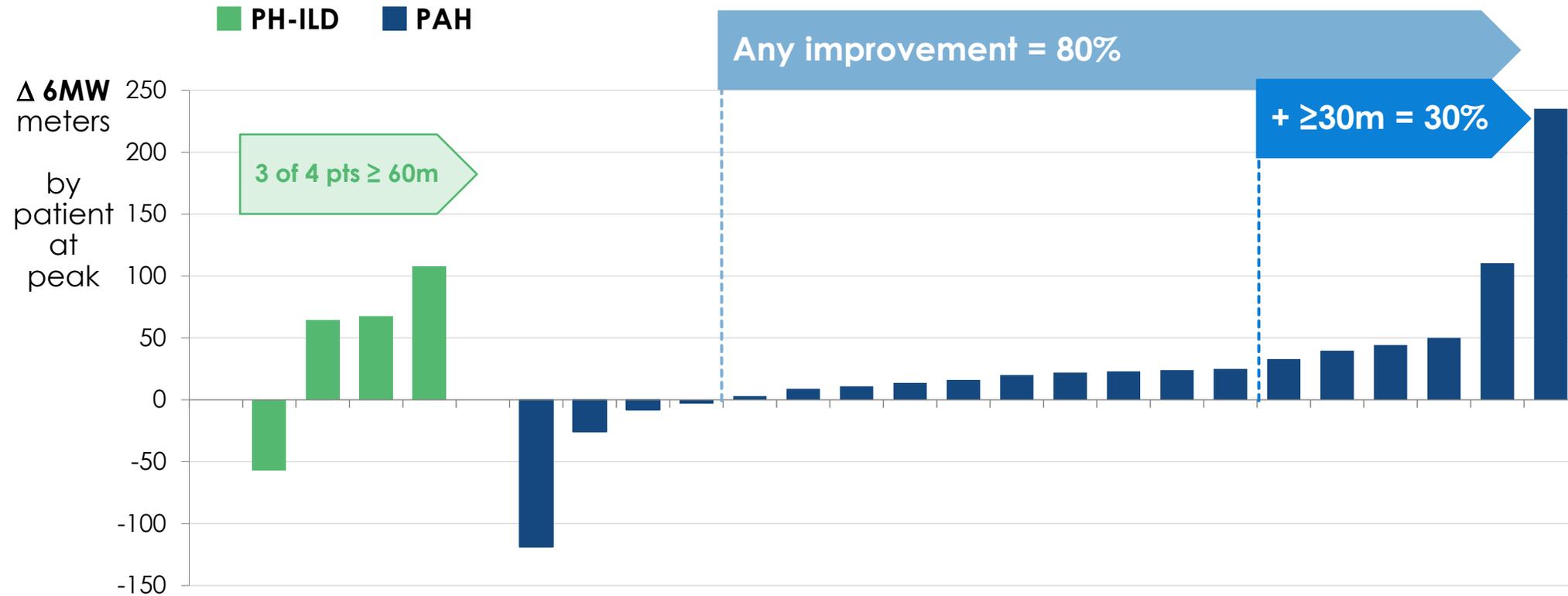
L606 Open-Label (U.S.): Week 48



Trough 6MWD conducted in morning of visit; Peak 6MWD conducted within 90-120 minutes after administering clinically observed dose
Mean ± Standard Deviation, Liquidia data on file

Most patients maintained or improved $\Delta 6MWD$ at Week 48

L606 Open-Label (U.S.): Week 48

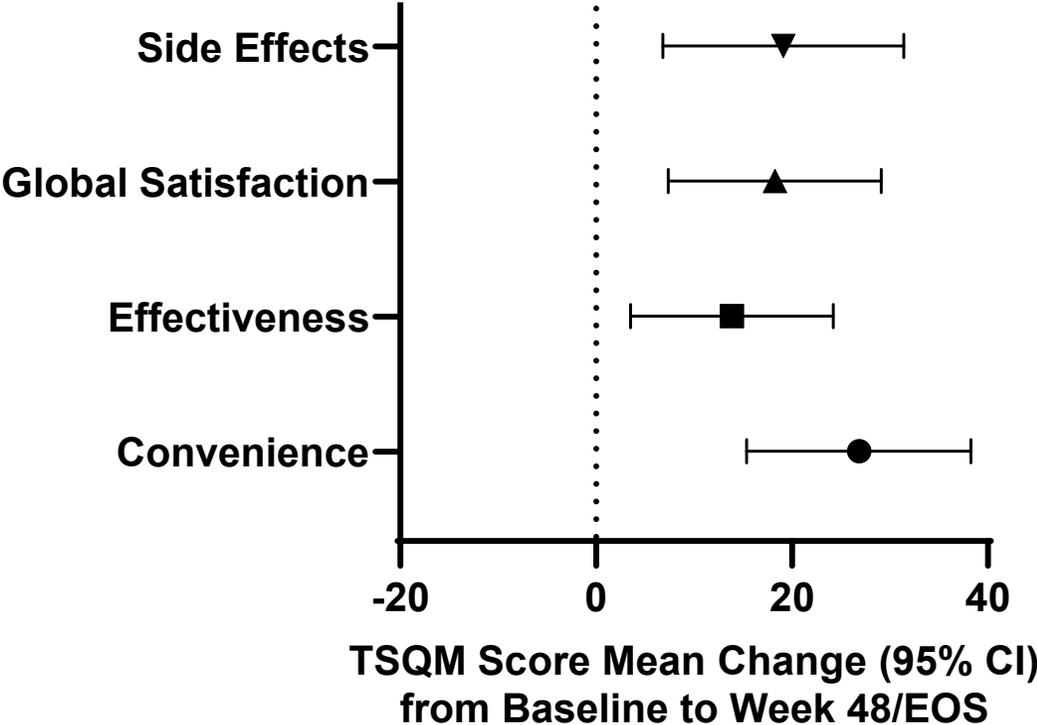


Liquidia Data on File; PAH patient (-118m) had knee surgery prior to conducting the 6MWD at Week 48
Liquidia data on file

Mean TSQM improved across all dimensions measured at 95% CI

L606 Open-Label (U.S.): Week 48

Cohort A: Tyvaso Transitions

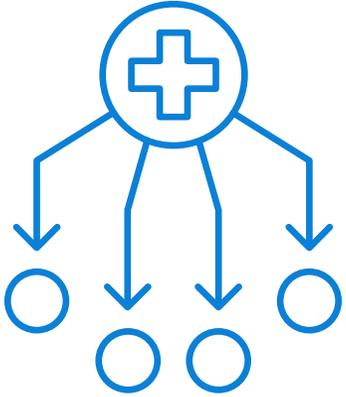


Scores range from 0 to 100, with higher scores indicating greater satisfaction.

Treatment Satisfaction Questionnaire for Medication (TSQM), Scores range from 0 to 100, with higher scores indicating greater satisfaction
Liquidia data on file

Summary of L606 results

Thoughts on inhaled and sustained release



- **L606 was well-tolerated in patients over 48 weeks**
- **Only 4 patients reports mild related cough over 48 weeks (14%)**
- **Most patients maintained or improved in 6MWD**
- **Observed minimal variability between peak and trough measures**

Re-Spire Pivotal Study

L606 (treprostinil liposomal inhalation suspension)

Dr. Rajeev Saggar

Seeking PAH & PH-ILD indication for L606 with one pivotal trial

Three data sets required per FDA and EMA feedback

 **Completed** **Comparable bioavailability**
to Tyvaso[®] in Phase 1¹

 **Ongoing** **Open-label safety study**
in U.S. of PAH & PH-ILD²

 **Planned** **Randomized placebo-controlled**
for efficacy in PH-ILD

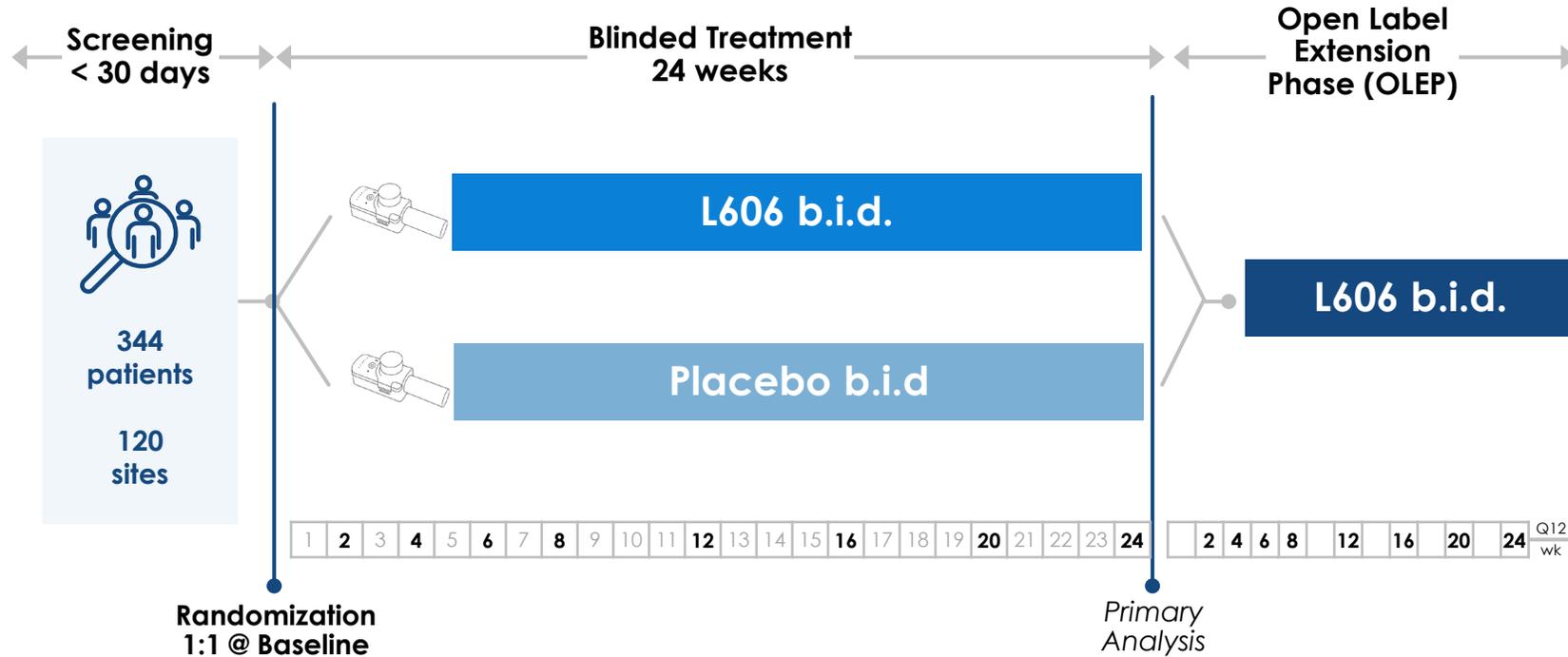
**Plan to initiate
sites globally**



Powered by Bing

1. Phase 1 study [NCT04041648](#); 2. Open label PAH & PH-ILD study [NCT04691154](#)

Phase 3, multi-center, randomized (1:1), double-blind, placebo-controlled, parallel group study



Primary Endpoint

- Change from BL in peak 6MWD (wk 16)

Secondary Endpoint

- Change from BL in peak 6MWD (wk 24)
- Change from BL in trough 6MWD (wk 16)
- TTCW from randomization

Baseline (BL), Six Meter Walk Distance (6MWD), Time to Clinical Worsening (TTCW), Quality of Life (QOL), Clinical Worsening Event (CWE), Combined Pulmonary Fibrosis and Emphysema (CPFE)

Key enrollment eligibility criteria

Any fibrotic ILD including Combined Pulmonary Fibrosis Emphysema



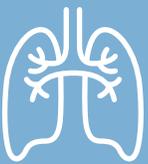
ILD diagnosed on institutional HRCT Chest

- HRCT Chest to be performed within 12-months of screening
- If HRCT Chest > 12-months a scan be performed during screening



Eligibility based on central read from two independent thoracic radiologists

- Consistent with ILD – evidence of diffuse parenchymal disease
- Evidence of “fibrosis”
- Total Lung Emphysema must be $\leq 15\%$



Allow patients on approved anti-fibrotic treatment (pirfenidone/nintedanib)

- Started at least 30-days prior to screening & stable dose with intent to continue throughout study
- This could also include possible new drugs approved for IPF/fibrotic ILD during the study e.g., PDE4-inhibitor

Q&A Discussion

Q&A session



Dr. Roger Jeffs
Chief Executive Officer



Dr. Richard Channick, MD



Dr. Rajeev Saggur
Chief Medical Officer



Dr. Rajan Saggur, MD



**Dr. Ricardo Restrepo-
Jaramillo**

Drivers of future value for Liquidia's programs

Portfolio built on optimizing inhaled delivery

Exposure
drives
Efficacy

Tolerability
drives
Durability

Convenience
drives
Compliance

Thank you for joining us today!