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ASCENT TO WEEK 16

Safety and Exploratory Efficacy Data of LIQ861 Dry Powder Inhaled Treprostinil in PH-ILD Patients

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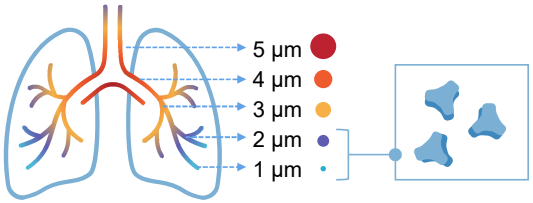
YUTREPIA™ (LIQ861) Engineered with PRINT® Technology

YUTREPIA™ (treprostinil) Inhalation Powder

Deep-lung Delivery[2,3]

PRINT® technology engineers monodispersed particles in the respirable range[3]



- ~1 µm sized trefoil shape[1]
- Free flowing[1,4]

Convenient Design[1]

Plastiape RS00 DPI is a portable, simple-to-use, robust, and low-effort inhaler[1,4,6]

Low Burden on Patients

Low inspiratory effort is required from the patient because formulation, particle size, and device improve aerosolization[1]

Device is not position-dependent[4]

PRINT® is a registered trademark of Liquidia Technologies, Inc.
PRINT® = Particle Replication in Nonwetting Templates.

1. Hill NS, et al. *Pulm Circ*. 2022;12(3):e12119. doi:10.1002/pul2.12119 2. Roscigno RF, et al. *Vascul Pharmacol*. 2021;138:106840. doi:10.1016/j.vph.2021.106840 3. Garcia A, et al. *J Drug Deliv*. 2012;2012:941243. doi:10.1155/2012/941243 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. Maynor B, et al. *Digit Respir Drug Deliv*. 2020;2:371-374. 6. YUTREPIA. Prescribing information. Liquidia Technologies, Inc; 2022.

Cohort A

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

Inclusion Criteria

WHO Group 3 PH-ILD, including CPFE

- Prostacyclin naïve
- Stable dose of ILD medications
- **18-75 years** of age; conditional 76-80
- **6MWD ≥125m**
- **FEV_1/FVC ≥70%**

		ILD LIMITED SUBSET (NO CPFE):
PVR	**≥3 WU**	**≥3 WU**
	OR	
PCWP	**≤15 mmHg**	**≤15 mmHg**
mPAP	**≥30 mmHg**	**≥21 - ≤29 mmHg**

Exclusion Criteria

WHO Groups 1, 2, 4, or 5

PAH oral therapy in **last 60 days** (PDE5-I)

Uncontrolled hypertension **160/100 mm Hg**

Chronic therapy new start **30 days**

> 8 L/min O_2 at baseline

Recent respiratory infection in **last 30 days**, ILD from SARS-CoV-2

Renal impairment eGRF **≤40 mL/min/1.73m²**

Initiated pulmonary rehab **within 12 weeks**

Acute PE **(90 days)**, TIA **(6 months)**

- On prostacyclin therapy
- History of prostacyclin intolerance
- Atrial septostomy
- History of left sided heart disease
- Hypersensitivity to IV contrast
- Pregnancy
- Sepsis

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)

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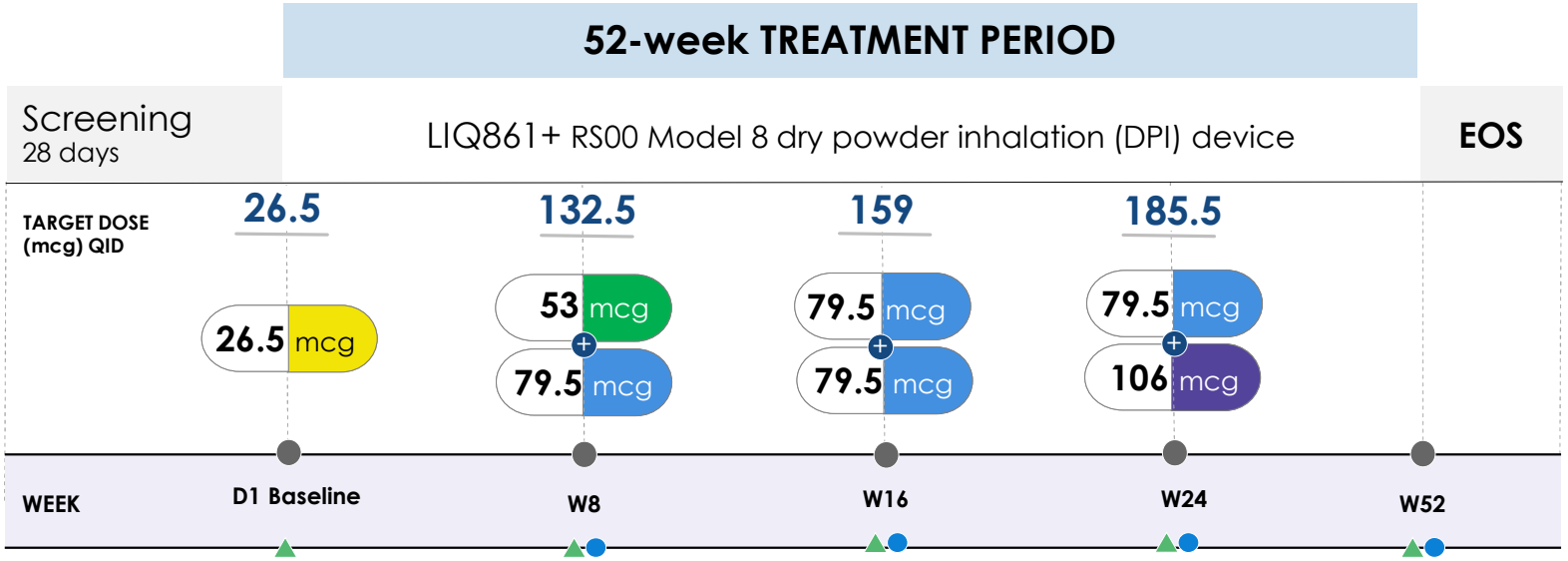
Cohort A

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



▲ **PRIMARY SAFETY ENDPOINTS**

- **Incidence of treatment-emergent drug- or device-related AEs and serious AEs**
- Changes from study drug initiation to week 24 in clinical laboratory and vital signs

● **EXPLORATORY ENDPOINTS**

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

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ASCENT Site Map

University of Utah
Salt Lake City, UT

National Jewish
Denver, CO

University of Minnesota
Rochester, MN

Ascension Medical Group
Indianapolis, IN

University of Rochester
Rochester, NY

Summit Health
Bend, Oregon

University of Michigan
Ann Arbor, MI

Tufts Medical Center
Boston, MA

UC Davis
Davis, CA

Hartford Hospital
Hartford, Connecticut

UCSF
San Francisco, CA

NYU Langone Medical Center
New York City, New York

Medical Research Group
Fresno, CA

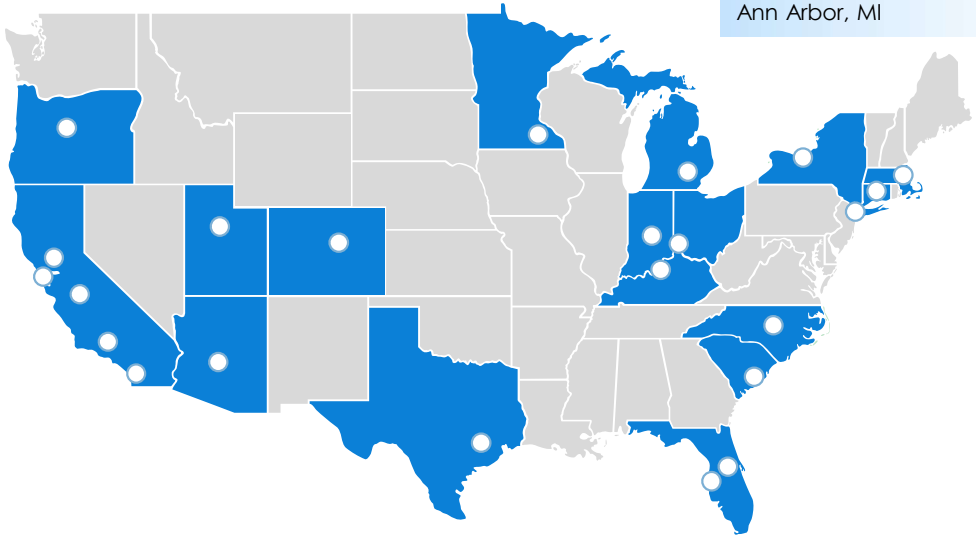
University of Cincinnati
Cincinnati, OH

UCLA
Los Angeles, CA

Norton Pulm Specialists
Louisville, KY

UCSD
San Diego, CA

University of North Carolina
Chapel Hill, NC



AZ Pulmonary Specialists
Scottsdale, AZ

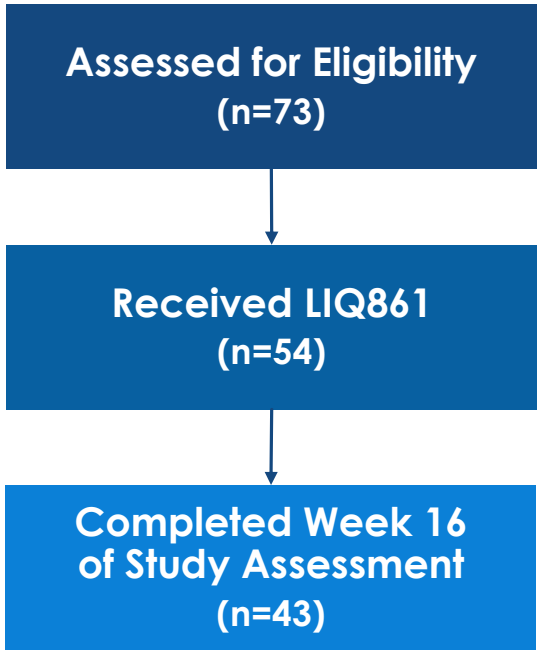
Houston Methodist
Houston, TX

Tampa General Hospital
Tampa, FL

Advent Health Medical Group
Orlando, FL

MUSC
Charleston, SC

Patient Disposition



Patients Completed Week 16 Visit, n (%)			
Completed	**43**	**(79.6)**	
Missed Study visit	**1**	**(1.9)**	
Discontinued	**10**	**(18.5)**	
Physician Decision	1	(1.9)	
Withdrawal of Patient	2	(3.7)	
Protocol Violation	1	(1.9)	
Adverse Event	3	(5.6)	chronic pancreatitis, coronavirus, lung neoplasm
Other	3	(5.6)	lung transplant

*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

Naïve PH-ILD	N=54
Age, mean ±SD, y	**68.5** ± 8.9
Sex, Female (%)	**28** ± 51.9%
Duration of PH Diagnosis, y	**0.5** ± 0.8
Duration of ILD Diagnosis, y	**5.1** ± 5.7
ILD Subtypes (%)	
IIPs	**26** (48.1%)
Autoimmune ILDs	**19** (35.2%)
HP	**1** (1.9%)
Other ILDs	**3** (5.6%)
CPFE	**5** (9.3%)
# Background antifibrotics, n (%)	
Nintedanib	**19** (35.2%)
Pirfenidone	**4** (7.4%)
Background PH Drugs, n (%): **PDE5i**	**7** (13%)

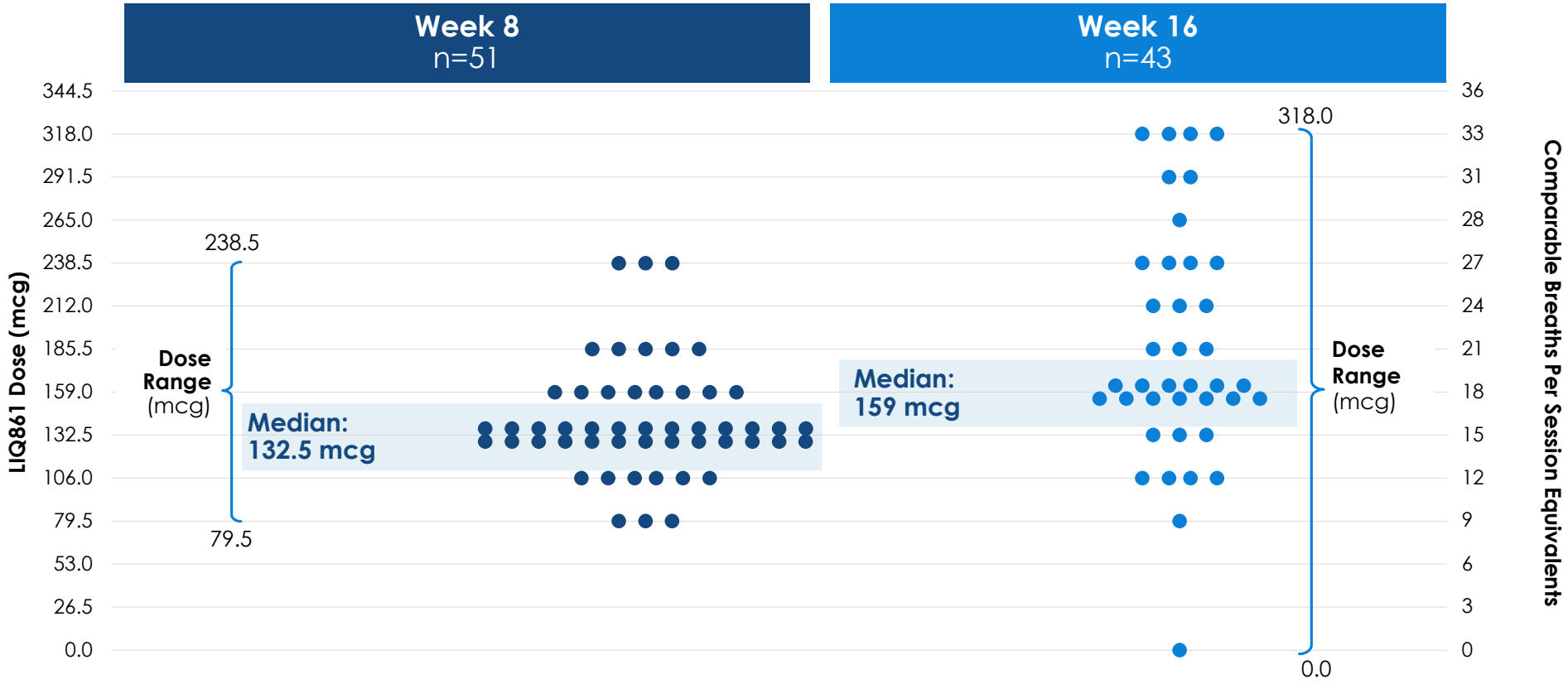
Peak Inspiratory Flow Rate (L/min)	**Mean = 90.6** ± 22.3 Median = 90.0 Range = 39-120

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

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

Clinical Characteristic	Mean ±SD
6MWD (Meters), ±SD	**298.1** ± 80.3
NTPro-BNP (pg/ml)	**611.0** ± 1246.0 [GM=210.5]
Dyspnea-12	**11.7** ± 6.8
EmPHasis-10	**24.6** ± 9.7
Simplified Cough Score	**1.3** ± 0.8

LIQ861 Dosing QID



Liquidia Data on File

ASCENT Safety: PH-ILD Week 16

Treatment-related TEAEs	ASCENT Naïve n=54 (n %)
Cough	26 (48.1)
Headache	10 (18.5)
Oropharyngeal pain	4 (7.4)
Fatigue	4 (7.4)
Diarrhea	3 (5.6)
Throat Irritation	3 (5.6)
Dry Throat	3 (5.6)

No discontinuations due to cough at 16 weeks

Cough
24 (92.3%): mild
2 (7.7%): moderate

1 (1.9%) severe respiratory tract irritation

No treatment related SAEs

AE reported if ≥ 5%
TEAE=treatment-emergent adverse event.
Liquidia Data on File

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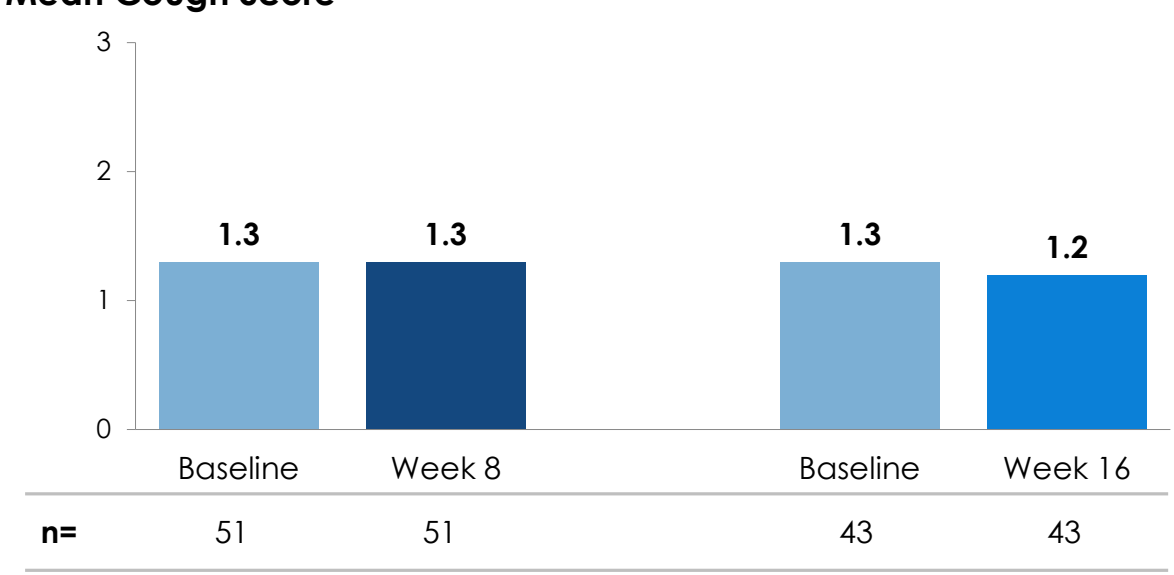
LIQ816 Simplified Cough Score

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

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

Mean Daytime Cough Scores Remained **Stable from Baseline Through Week 8 And 16** with LIQ861 Treatment



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.
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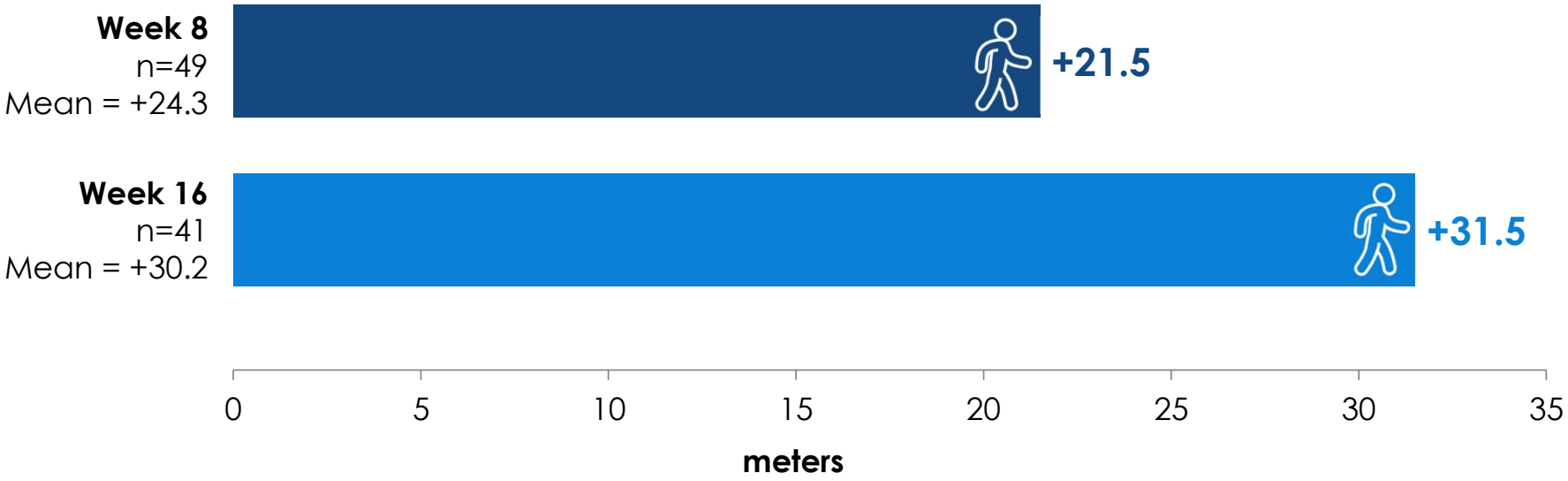
Median Change from Baseline in 6MWD in meters

ASCENT LIQ861



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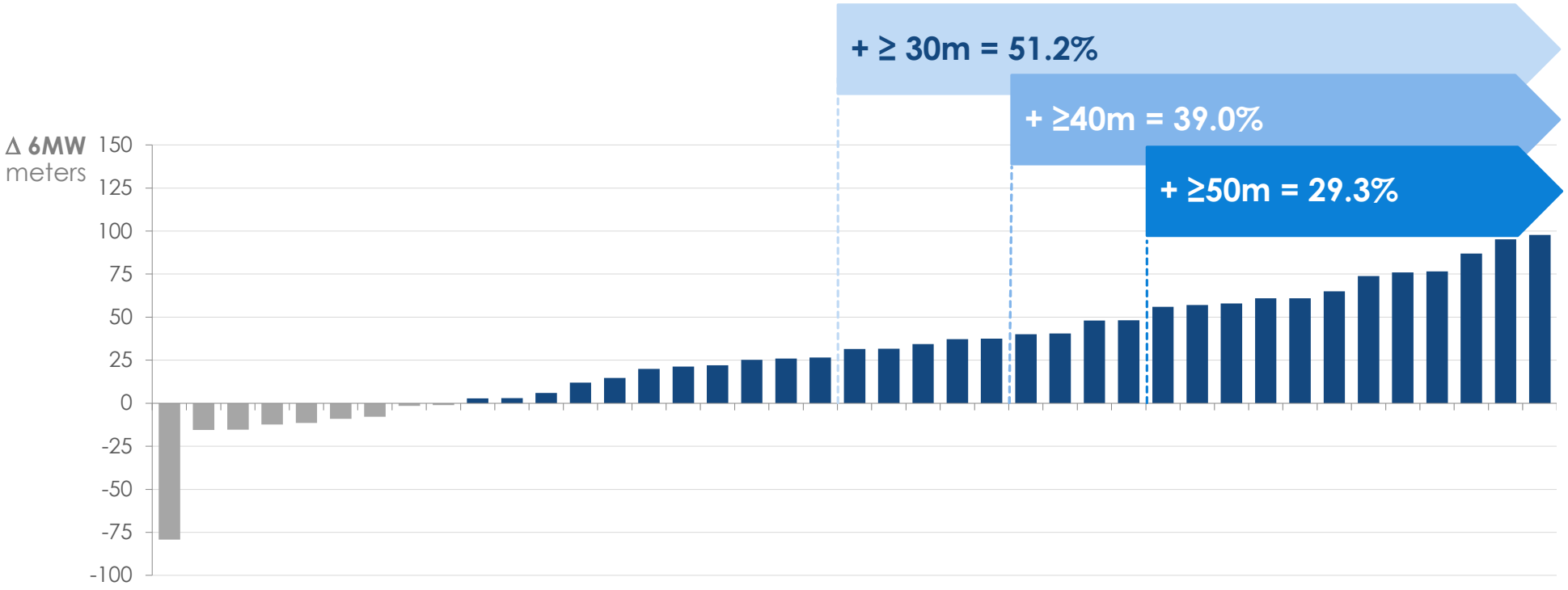
Week 16: Δ6MWD by Individual Patient

78% of patients maintained or improved from baseline



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ASCENT LIQ861 – Summary

1. The treatment-related TEAEs observed are consistent with the known safety profile of inhaled treprostinil

2. The median dose at Week 8 was 132.5mcg QID (15 nebulized breath equivalent), increasing to 159mcg QID (18 nebulized breath equivalent) by Week 16

3. 92.3% of treatment related cough was mild. No worsening in patient-perceived cough score was observed despite higher LIQ861 dosing

4. By Week 16, median improvement in 6MWD was +31.5 meters above baseline. 29.3% improved by +50 meters or more.