For Immediate Release
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UNITED THERAPEUTICS ANNOUNCES *INCREASE* DATA PRESENTATION AT AN AMERICAN THORACIC SOCIETY VIRTUAL SESSION

SILVER SPRING, Md. and RESEARCH TRIANGLE PARK, N.C., Wednesday, June 17, 2020: United Therapeutics Corporation (Nasdaq: UTHR) announced today that key data from the *INCREASE* study of Tyvaso® (treprostinil) Inhalation Solution in patients suffering from World Health Organization (WHO) Group 3 pulmonary hypertension associated with interstitial lung disease (PH-ILD) will be presented at a Breaking News Session hosted by the American Thoracic Society (ATS) on June 24, 2020.

The virtual presentation, entitled "Inhaled Treprostinil in Interstitial Lung Disease Associated Pulmonary Hypertension: The *INCREASE* Study" will be given by Dr. Steven D. Nathan, M.D., Director of the Advanced Lung Disease Program and Director of the Lung Transplant Program at Inova Fairfax Hospital. The presentation will take place on Wednesday, June 24, 2020 from 2:53 to 3:18 p.m., Eastern Daylight Time. Additional information about the ATS session, entitled "Breaking News: Clinical Trial Results in Pulmonary Medicine," including participant registration information, can be found on the ATS International Conference website at https://conference.thoracic.org/program/virtual-clinical-trials.php. Those interested in viewing the presentation should register free of charge in advance as a replay of the presentation will not be immediately available.

As previously announced on February 24, 2020, Tyvaso increased six-minute walk distance (6MWD) by 21 meters versus placebo (p=0.0043, Hodges-Lehmann estimate) after 16 weeks of treatment. Benefits of Tyvaso were observed across several key subgroups, including etiology of PH-ILD, disease severity, age, gender, baseline hemodynamics, and dose.

Significant improvements were also observed in each of the study's secondary endpoints, including reduction in the cardiac biomarker NT-proBNP, time to first clinical worsening event, change in peak 6MWD at Week 12, and change in trough 6MWD at week 15. Treatment with Tyvaso of up to 12 breaths per session, four times daily, in the *INCREASE* study was well tolerated and the safety profile was consistent with previous Tyvaso studies in pulmonary arterial hypertension and known prostacyclin-related adverse events (see the Important Safety Information below under "About Tyvaso").

United Therapeutics recently submitted a supplemental New Drug Application (sNDA) to the U.S. Food and Drug Administration seeking to revise the Tyvaso label to reflect the outcome of the *INCREASE* study.

About *INCREASE*

INCREASE was a phase III, multicenter, randomized, double-blinded, placebo-controlled, 16-week, parallel group study of Tyvaso in patients with pulmonary hypertension associated with interstitial lung disease. Enrollment into the study was completed in August 2019 with a total of 326 patients. Patients were randomized in a 1:1 Tyvaso (n=163) or placebo (n=163).

The primary endpoint was to evaluate the change in 6MWD measured at peak exposure from Baseline to Week 16.

Secondary objectives of the study included:

- Change in plasma concentration of N-terminal pro-brain natriuretic peptide (NT-proBNP) from Baseline to Week 16
- Time to clinical worsening calculated as the time from randomization until one of the following criteria are met:
    - Hospitalization due to a cardiopulmonary indication
    - Decrease in 6MWD >15% from Baseline directly related to disease under study, at two consecutive visits, and at least 24 hours apart
    - Death (all causes)
    - Lung transplantation
- Change in peak 6MWD from Baseline to Week 12
- Change in trough 6MWD from Baseline to Week 15

Exploratory objectives of the study evaluated changes in peak 6MWD at Week 4 and Week 8, changes in quality of life as measured by the St. George's Respiratory Questionnaire (SGRQ), and change in distance saturation product (DSP). Further exploratory analysis will also be performed on biomarkers and pharmacogenomics from this study.

Additional study results will be made available through scientific disclosure in upcoming peer-reviewed publications.

About TYVASO® (treprostinil) Inhalation Solution

For the treatment of pulmonary arterial hypertension (PAH; WHO Group 1) to improve exercise ability.

INDICATION

TYVASO (treprostinil) is a prostacyclin vasodilator indicated for the treatment of pulmonary arterial hypertension (PAH; WHO Group 1) to improve exercise ability.

Studies establishing effectiveness included predominately patients with NYHA Functional Class III symptoms and etiologies of idiopathic or heritable PAH (56%) or PAH associated with connective tissue diseases (33%).
The effects diminish over the minimum recommended dosing interval of 4 hours; treatment timing can be adjusted for planned activities.
While there are long-term data on use of treprostinil by other routes of administration, nearly all controlled clinical experience with inhaled treprostinil has been on a background of bosentan (an endothelin receptor antagonist) or sildenafil (a phosphodiesterase type 5 inhibitor). The controlled clinical experience was limited to 12 weeks in duration.

IMPORTANT SAFETY INFORMATION

WARNINGS AND PRECAUTIONS

- The efficacy of TYVASO has not been established in patients with significant underlying lung disease (such as asthma or chronic obstructive pulmonary disease). Patients with acute pulmonary infections should be carefully monitored to detect any worsening of lung disease and loss of drug effect
- TYVASO is a pulmonary and systemic vasodilator. In patients with low systemic arterial pressure, TYVASO may cause symptomatic hypotension
- Titrate slowly in patients with hepatic or renal insufficiency, as exposure to treprostinil may be increased in these patients
- TYVASO inhibits platelet aggregation and increases the risk of bleeding
- Co-administration of the cytochrome P450 (CYP) 2C8 enzyme inhibitor gemfibrozil may increase exposure to treprostinil. Co-administration of the CYP2C8 enzyme inducer rifampin may decrease exposure to treprostinil. Increased exposure is likely to increase adverse events, whereas decreased exposure is likely to reduce clinical effectiveness

DRUG INTERACTIONS/SPECIFIC POPULATIONS

- The concomitant use of TYVASO with diuretics, antihypertensives, or other vasodilators may increase the risk of symptomatic hypotension
- Co-administration of the CYP2C8 enzyme inhibitor gemfibrozil increases exposure to oral treprostinil. Co-administration of the CYP2C8 enzyme inducer rifampin decreases exposure to oral treprostinil. It is unclear if the safety and efficacy of treprostinil by the inhalation route are altered by inhibitors or inducers of CYP2C8
- Limited case reports of treprostinil use in pregnant women are insufficient to inform a drug-associated risk of adverse developmental outcomes. However, pulmonary arterial hypertension is associated with an increased risk of maternal and fetal mortality. There are no data on the presence of treprostinil in human milk, the effects on the breastfed infant, or the effects on milk production
- Safety and effectiveness in pediatric patients have not been established
- It is unknown if geriatric patients respond differently than younger patients. Caution should be used when selecting a dose for geriatric patients

ADVERSE REACTIONS

- The most common adverse reactions seen with TYVASO in ≥4% of PAH patients and more than 3% greater than placebo in the placebo-controlled clinical study were cough (54% vs 29%), headache (41% vs 23%), throat irritation/pharyngolaryngeal pain (25% vs 14%), nausea (19% vs 11%), flushing (15% vs <1%), and syncope (6% vs <1%). In addition, adverse reactions occurring in ≥10% of patients were dizziness and diarrhea

Please see the Full Prescribing Information, Patient Product Information, and the TD-100 and TD-300 TYVASO® Inhalation System Instructions for Use manuals.

For additional information about TYVASO, visit www.tyvaso.com or call 1-877-UNITHER (1-877-864-8437). For Consumer Important Safety Information, please see https://www.tyvaso.com/dtc/isi

About United Therapeutics

United Therapeutics Corporation focuses on the strength of a balanced, value-creating biotechnology model. We are confident in our future thanks to our fundamental attributes, namely our obsession with quality and innovation, the power of our brands, our entrepreneurial culture, and our bioinformatics leadership. We also believe that our determination to be responsible citizens – having a positive impact on patients, the environment, and society – will sustain our success in the long term.

Through our wholly owned subsidiary, Lung Biotechnology PBC, we are focused on addressing the acute national shortage of transplantable lungs and other organs with a variety of technologies that either delay the need for such organs or expand the supply. Lung Biotechnology is the first public benefit corporation subsidiary of a public biotechnology or pharmaceutical company.

Forward-looking Statements

Statements included in this press release that are not historical in nature are "forward-looking statements" within the meaning of the Private Securities Litigation Reform Act of 1995. Forward-looking statements include, among others, statements relating to our expectation that our recent submission of the *INCREASE* data to the FDA will lead to revised labeling for Tyvaso, our ability to create value and sustain our success in the long-term, and our efforts to develop technologies that either delay the need for transplantable organs or expand the supply of transplantable organs. These forward-looking statements are subject to certain risks and uncertainties, such as those described in our periodic reports filed with the Securities and Exchange Commission, that could cause actual results to differ materially from anticipated results. Consequently, such forward-looking statements are qualified by the cautionary statements, cautionary language and risk factors set forth in our periodic reports and documents filed with the Securities and Exchange Commission, including our most recent Annual Report on Form 10-K, Quarterly Reports on Form 10-Q, and Current Reports on Form 8-K. We claim the

protection of the safe harbor contained in the Private Securities Litigation Reform Act of 1995 for forward-looking statements. We are providing this information as of June 17, 2020 and assume no obligation to update or revise the information contained in this press release whether as a result of new information, future events, or any other reason.

TYVASO is a registered trademark of United Therapeutics Corporation.

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