



For Immediate Release

United Therapeutics Corporation to Review Data from the Successful *TETON-2* Pivotal Study of Nebulized Tyvaso® in Idiopathic Pulmonary Fibrosis

SILVER SPRING, Md. & RESEARCH TRIANGLE PARK, N.C., September 19, 2025: United Therapeutics Corporation (Nasdaq: **UTHR**) announced today that it will host a webcast to review data from the successful *TETON-2* pivotal study evaluating the use of nebulized Tyvaso® (treprostinil) Inhalation Solution for the treatment of idiopathic pulmonary fibrosis (**IPF**) on Sunday, September 28, 2025, at 12:30 p.m. Eastern Time.

During the webcast, Steven D. Nathan, M.D., Schar Chair, Advanced Lung Disease and Lung Transplant Program at Inova Fairfax Hospital and Chair of the *TETON* Steering Committee, will review data that were presented earlier in the day at the European Respiratory Society Congress. He will be joined by Leigh Peterson, Ph.D., Executive Vice President, Product Development and Xenotransplantation at United Therapeutics, Peter Smith, Pharm.D., Senior Vice President, Product Development at United Therapeutics and the lead for the global *TETON* program, and CQ Deng, Ph.D., M.D., Senior Vice President, Biostatistics, Statistical Programming and Data Management at United Therapeutics.

United Therapeutics previously [announced](#) that the *TETON-2* study met its primary efficacy endpoint of demonstrating improvement in absolute forced vital capacity (**FVC**) relative to placebo, including that statistically significant improvements relative to placebo were also observed in most secondary endpoints.

The webcast will be accessible via United Therapeutics' website at <https://ir.unither.com/events-and-presentations>. A rebroadcast of the webcast will be available for one year and can be accessed at the same location.

About *TETON-2*

The *TETON-2* study ([NCT05255991](#)) was a 597-patient, multicenter, randomized, double-blind, placebo-controlled phase 3 registration study evaluating the safety and efficacy of nebulized Tyvaso in subjects with IPF over a 52-week period at sites in Argentina, Australia, Belgium, Chile, Denmark, France, Germany, Israel, Italy, Mexico, the Netherlands, New Zealand, Peru, South Korea, Spain, and Taiwan. The study achieved full enrollment in July 2024.

Subjects were randomly assigned 1:1 to receive nebulized Tyvaso or placebo, stratified by IPF background therapy use. All subjects initiated nebulized Tyvaso or placebo at a dose of three breaths administered four times daily (**QID**) and titrated to a target dosing regimen of 12 breaths QID. Study drug doses were titrated up as tolerated, until the target dose or maximum clinically tolerated dose was achieved.

The primary endpoint of the study was the change in absolute FVC from baseline to week 52. Secondary endpoints included: (1) time to clinical worsening; (2) time to first acute exacerbation of IPF; (3) overall survival at week 52; (4) change in percent predicted FVC from baseline to week 52; (5) change in the K-BILD questionnaire from baseline to week 52; and (6) change in DLCO from baseline to week 52.

Safety assessments included the development of adverse events, serious adverse events, vital signs, clinical laboratory parameters, and electrocardiogram parameters.

Eligible subjects completing the *TETON-2* study could enroll in the *TETON-OLE* study ([NCT04905693](#)), an ongoing open-label extension study to evaluate the long-term safety and tolerability of nebulized Tyvaso in subjects with fibrotic lung disease.

About IPF

Idiopathic pulmonary fibrosis, or IPF, is a scarring disease of the lungs of an unknown (idiopathic) cause and is the most common of the idiopathic interstitial pneumonias. IPF is characterized by the progressive loss of the ability of the lungs to transfer oxygen into the blood, ultimately resulting in respiratory failure and death. While the precise causes of IPF remain unknown, IPF rarely presents before age 50 and can be associated with cigarette smoking and certain genetic dispositions. In addition, some evidence suggests that gastroesophageal reflux (acid reflux, or heartburn), certain viral infections, air pollution, and workplace exposures may be risk factors for IPF. [According to recent research](#), IPF is estimated to affect between 0.33 and 4.51 people per 10,000 persons worldwide. Further, United Therapeutics estimates there are over 100,000 IPF patients in the United States.

About Tyvaso® (treprostinil) Inhalation Solution

INDICATION

TYVASO (treprostinil) Inhalation Solution is a prostacyclin mimetic indicated for the treatment of:

- Pulmonary arterial hypertension (PAH; WHO Group 1) to improve exercise ability. Studies with TYVASO establishing effectiveness predominately included 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 clinical experience with inhaled treprostinil has been on a background of an endothelin receptor antagonist (ERA) and/or a phosphodiesterase type 5 (PDE-5) inhibitor. The controlled clinical experience with TYVASO was limited to 12 weeks in duration.

- Pulmonary hypertension associated with interstitial lung disease (PH-ILD; WHO Group 3) to improve exercise ability. The study with TYVASO establishing effectiveness predominately included patients with etiologies of idiopathic interstitial pneumonia (IIP) (45%) inclusive of idiopathic pulmonary fibrosis (IPF), combined pulmonary fibrosis and emphysema (CPFE) (25%), and WHO Group 3 connective tissue disease (22%).

IMPORTANT SAFETY INFORMATION

WARNINGS AND PRECAUTIONS

TYVASO is a pulmonary and systemic vasodilator. In patients with low systemic arterial pressure, TYVASO may produce symptomatic hypotension.

TYVASO inhibits platelet aggregation and increases the risk of bleeding.

Co-administration of a cytochrome P450 (CYP) 2C8 enzyme inhibitor (e.g., gemfibrozil) may increase exposure (both C_{max} and AUC) to treprostinil. Co-administration of a CYP2C8 enzyme inducer (e.g., rifampin) may decrease exposure to treprostinil. Increased exposure is likely to increase adverse events associated with treprostinil administration, whereas decreased exposure is likely to reduce clinical effectiveness.

Like other inhaled prostaglandins, TYVASO may cause acute bronchospasm. Patients with asthma or chronic obstructive pulmonary disease (COPD), or other bronchial hyperreactivity, are at increased risk for bronchospasm. Ensure that such patients are treated optimally for reactive airway disease prior to and during treatment with TYVASO.

DRUG INTERACTIONS/SPECIFIC POPULATIONS

The concomitant use of TYVASO with diuretics, antihypertensives, or other vasodilators may increase the risk of symptomatic hypotension.

Human pharmacokinetic studies with an oral formulation of treprostinil (treprostinil diolamine) indicated that co-administration of the cytochrome P450 (CYP) 2C8 enzyme inhibitor, gemfibrozil, increases exposure (both C_{max} and AUC) to treprostinil. Co-administration of the CYP2C8 enzyme inducer, rifampin, decreases exposure to 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.

Across clinical studies used to establish the effectiveness of TYVASO in patients with PAH and PH-ILD, 268 (47.8%) patients aged 65 years and over were enrolled. The treatment effects and safety profile observed in geriatric patients were similar to younger patients. In general, dose selection for an elderly patient should be cautious, reflecting the greater frequency of hepatic, renal, or cardiac dysfunction, and of concomitant diseases or other drug therapy.

ADVERSE REACTIONS

Pulmonary Arterial Hypertension (WHO Group 1)

In a 12-week, placebo-controlled study (TRIUMPH I) of 235 patients with PAH (WHO Group 1 and nearly all NYHA Functional Class III), the most common adverse reactions seen with TYVASO in ≥4% of PAH patients and more than 3% greater than placebo 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 ≥4% of patients were dizziness and diarrhea.

Pulmonary Hypertension Associated with ILD (WHO Group 3)

In a 16-week, placebo-controlled study (INCREASE) of 326 patients with PH-ILD (WHO Group 3), adverse reactions with TYVASO were similar to the experience in studies of PAH.

Please see Full Prescribing Information for TYVASO, the TD-300 TYVASO® Inhalation System Instructions for Use manual, and additional information at www.TYVASOHCP.com or call 1 844 UNITHER (1-844-864-8437).

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United Therapeutics: Enabling Inspiration

At United Therapeutics, our vision and mission are one. We use our enthusiasm, creativity, and persistence to innovate for the unmet medical needs of our patients and to benefit our other stakeholders. We are bold and unconventional. We have fun, we do good. We are the first publicly-traded biotech or pharmaceutical company to take the form of a public benefit corporation (**PBC**). Our public benefit purpose is to provide a brighter future for patients through (a) the development of novel pharmaceutical therapies; and (b) technologies that expand the availability of transplantable organs.

You can learn more about what it means to be a PBC here: unither.com/pbc.

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, our efforts to innovate for the unmet medical needs of our patients, to benefit our other stakeholders, and to pursue our public benefit purpose of developing novel pharmaceutical therapies and technologies that expand the availability 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 September 19, 2025, 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.

For Further Information Contact:

Investors

<https://ir.unither.com/contact-ir>

Media

mrteam@unither.com