For Immediate Release

United Therapeutics Corporation Announces Full Enrollment of the *TETON-PPF* Study of Inhaled Treprostinil for the Treatment of Progressive Pulmonary Fibrosis

Approximately 200,000 patients in the United States have PPF, with limited treatment options

Topline data expected in the second half of 2027

SILVER SPRING, Md. & RESEARCH TRIANGLE PARK, N.C., August 17, 2026: United Therapeutics Corporation (Nasdaq: **UTHR**) announced full enrollment of its *TETON-PPF* study evaluating the use of Tyvaso® (treprostinil) inhalation solution (**nebulized Tyvaso**) for the treatment of progressive pulmonary fibrosis (**PPF**). The *TETON-PPF* study enrolled 754 patients.

"The rapid pace of enrollment underscores both the significant unmet need for new treatment options in PPF and the strong interest in nebulized Tyvaso's potential in addressing the burden of this disease, particularly following the highly successful results observed in our *TETON-1* and *TETON-2* studies in idiopathic pulmonary fibrosis (**IPF**)," said **Peter Smith, Pharm. D.**, Senior Vice President, Product Development at United Therapeutics and the lead for the global *TETON* program. "Due to the similarities in the mechanism of fibrosis between IPF and PPF, we are optimistic that anti-fibrotic therapies, such as inhaled treprostinil, will impact disease progression similarly in patients with these conditions. We are deeply grateful to the participants, investigators, and study teams whose commitment made this milestone possible."

If the *TETON-PPF* trial is successful, United Therapeutics intends to use the data from the study to support a supplemental New Drug Application (**sNDA**) to the U.S. Food and Drug Administration (**FDA**) to add PPF to the labeled indications for nebulized Tyvaso. Nebulized Tyvaso has not been approved by the FDA for PPF and remains investigational for this indication.

United Therapeutics is currently seeking priority review of an sNDA submitted to the FDA in June 2026 to add IPF to the labeled indications for nebulized Tyvaso based on data from the *TETON-1* and *TETON-2* studies. Data from these studies were published in the *New England Journal of Medicine*, available here and here, respectively.

About *TETON-PPF*

The *TETON-PPF* study is a 754-patient, multinational, randomized, double-blind, placebo-controlled phase 3 registration study to evaluate the safety and efficacy of nebulized Tyvaso in subjects with PPF over a 52-week period. The study reached full enrollment in August 2026 and topline data is expected in the second half of 2027.

Subjects were randomly allocated 1:1 to receive nebulized Tyvaso or placebo. All subjects will initiate nebulized Tyvaso or placebo at a dose of three breaths administered four times daily (**QID**) and will titrate to a target dosing regimen of 12 breaths QID. Study drug doses may be titrated up as tolerated, until the target dose or maximum clinically tolerated dose is achieved.

The primary endpoint of the study is the change in absolute forced vital capacity (**FVC**) from baseline to week 52. Secondary endpoints include: (1) time to first clinical worsening; (2) time to first acute

exacerbation of ILD; (3) overall survival at week 52; (4) change in percent predicted FVC from baseline to week 52; (5) change in the King's Brief Interstitial Lung Disease questionnaire score from baseline to week 52; and (6) change in diffusing capacity of the lungs for carbon monoxide from baseline to week 52.

Other data collected in the study will include the plasma N-terminal pro-brain natriuretic peptide concentration and supplemental oxygen use. Safety assessments include the development of adverse events, serious adverse events, vital signs, clinical laboratory parameters, and electrocardiogram parameters.

Subjects who complete the Week 52 visit may be offered the opportunity to enter an open-label extension study after completing the final study visit.

About PPF

PPF occurs in interstitial lung diseases (**ILD**) and exhibits progressive, self-sustaining fibrosis, and a similar disease course to IPF. PPF includes idiopathic interstitial pneumonias, autoimmune ILDs, chronic fibrosing hypersensitivity pneumonitis, and fibrotic ILDs related to environmental/occupational exposure. It is estimated that 13% to 40% of patients with these various ILDs will go on to develop PPF. Patients with PPF exhibit decreased lung function, poor quality of life, and increased mortality despite usual treatments for the underlying ILD. Estimates for median transplant free survival and overall survival are approximately 2.9 years and 3.7 years, respectively. While estimates vary, we believe the size of the U.S. PPF population is approximately 200,000 patients.

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 Cmax 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 Cmax 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).

About United Therapeutics

Founded by CEO Martine Rothblatt to discover a cure for her daughter's life-threatening rare disease, pulmonary arterial hypertension, United Therapeutics transforms the treatment of rare diseases and pioneers alternatives to expand the supply of transplantable organs. From our innovative therapies to our groundbreaking manufactured organs, we are bold and unconventional. We move quickly from scientific theory to practical technologies that can save lives. As a public benefit corporation, even our legal structure reflects our commitments. We serve patients, act with integrity, create long-term shareholder value, and operate with sustainable practices that protect the future we are working to build. Visit us at www.unither.com and follow us on LinkedIn, Facebook, and Instagram.

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 regarding: the potential timing and outcome of the *TETON-PPF* study; our plans to obtain FDA approval of sNDAs to add PPF and IPF to the labeled indications for nebulized Tyvaso; the potential benefits of nebulized Tyvaso for patients with PPF; and our goals of expanding the supply of transplantable organs, developing practical technologies that can save lives, creating long-term shareholder value, and operating with sustainable practices. 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 August 17, 2026, and assume no obligation to update or revise the information contained in this press release whether because of new information, future events or any other reason.

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