# YUTREPIA® (treprostinil) Inhalation Powder for PH-ILD Patients

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Pulmonary hypertension associated with interstitial lung disease (PH-ILD) presents a significant clinical challenge, often leading to progressive dyspnea and reduced exercise capacity. Current therapeutic options are limited, underscoring the need for effective interventions. The introduction of YUTREPIA® (treprostinil) inhalation powder provides a prostacyclin treatment for this patient population, addressing a critical unmet need.1

**KEY TAKEAWAYS**

- The Pivot: YUTREPIA® (treprostinil) inhalation powder offers a new prostacyclin-based treatment for PH-ILD, expanding therapeutic options beyond existing pulmonary vasodilators.
- The Data: While specific trial data are not provided, treprostinil, as a prostacyclin analogue, is established in pulmonary arterial hypertension (PAH) and is now being applied to PH-ILD.
- The Action: Clinicians managing PH-ILD patients should consider YUTREPIA® as a potential treatment, particularly for those experiencing progressive symptoms despite conventional management.

Pulmonary hypertension associated with interstitial lung disease (PH-ILD) is a complex and progressive condition characterized by elevated pulmonary arterial pressures in the context of underlying ILD.1 This combination often leads to severe symptoms, including dyspnea, fatigue, and impaired functional capacity, significantly impacting patient quality of life and prognosis.2 The pathophysiology involves pulmonary vascular remodeling, inflammation, and fibrosis, distinct from idiopathic pulmonary arterial hypertension (PAH) but sharing some common pathways.3 Historically, treatment strategies for PH-ILD have been challenging, with many PAH-specific therapies showing limited or even detrimental effects in this population.4 The need for targeted therapies that can safely and effectively improve outcomes for PH-ILD patients remains substantial. The prevalence of PH-ILD varies depending on the underlying ILD type and diagnostic criteria, but it is estimated to affect a significant proportion of ILD patients, particularly those with more advanced disease.1 For instance, studies suggest that PH can be present in 30-50% of patients with idiopathic pulmonary fibrosis (IPF), a common and severe form of ILD.2 The presence of PH in ILD is an independent predictor of worse survival, underscoring the urgency for effective therapeutic interventions.3

## YUTREPIA® (treprostinil) Inhalation Powder

YUTREPIA® (treprostinil) inhalation powder represents a prostacyclin analogue delivered via a dry powder inhaler.5 Treprostinil, a synthetic analogue of prostacyclin (PGI2), exerts its therapeutic effects through direct vasodilation of the pulmonary and systemic arterial beds and inhibition of platelet aggregation.6 These actions contribute to

reducing pulmonary vascular resistance and improving cardiac output.7 The dry powder inhalation formulation offers a non-invasive delivery method, potentially improving patient adherence and convenience compared to intravenous or subcutaneous prostacyclin analogues.8 This method of delivery bypasses the need for complex infusion pumps or frequent injections, which can be burdensome for patients and limit their quality of life. The device design focuses on ease of use, aiming to ensure consistent drug delivery with minimal patient training.12

While specific trial data for YUTREPIA® in PH-ILD were not provided for this summary, the mechanism of action of treprostinil is well-established in the treatment of pulmonary arterial hypertension (PAH).9 In PAH, treprostinil has demonstrated improvements in exercise capacity, hemodynamics, and clinical worsening.10 The application of treprostinil to PH-ILD is based on the rationale that prostacyclin pathways are implicated in the vascular remodeling observed in PH-ILD, similar to PAH.11 The dry powder inhaler device is designed for ease of use, delivering a consistent dose of treprostinil.12 Patients typically self-administer the medication multiple times daily.13 The safety profile of inhaled treprostinil generally includes dose-dependent side effects such as cough, headache, jaw pain, and flushing, consistent with its vasodilatory properties.14 Careful titration is often required to optimize tolerability and efficacy.15 This titration process involves gradually increasing the dose over time, allowing patients to adjust to the vasodilatory effects and minimize adverse events. The typical starting dose and subsequent titration schedule would be determined by the treating physician based on individual patient response and tolerability. The target patient population for YUTREPIA® in PH-ILD includes adults diagnosed with PH-ILD who exhibit symptomatic disease despite conventional management. These patients often present with significant functional impairment and a reduced quality of life, making them candidates for therapies that can address the pulmonary vascular component of their disease. The non-invasive nature of the dry powder inhaler may be particularly beneficial for patients with advanced ILD who may have limited mobility or dexterity.8

The development and presentation of YUTREPIA® at ATS 2026 highlight an ongoing effort to expand therapeutic options for PH-ILD, a population with high unmet medical need.16 The availability of an inhaled prostacyclin analogue offers clinicians an additional tool in managing the complex vascular component of PH-ILD.17 Further research, including dedicated clinical trials in PH-ILD, will continue to refine the understanding of its precise role, optimal patient selection, and long-term outcomes in this specific patient group.18 Limitations in the current understanding of PH-ILD treatment include the heterogeneity of the disease, the lack of large-scale, randomized controlled trials specifically in PH-ILD, and the potential for differential responses based on the underlying ILD subtype. While treprostinil's mechanism of action is well-understood in PAH, its specific efficacy and safety profile in the context of the fibrotic lung environment of ILD require further investigation. The potential for drug-drug interactions with other medications commonly used in ILD patients also warrants careful consideration. The long-term impact of YUTREPIA® on disease progression, hospitalizations, and survival in PH-ILD patients will be critical areas for future study.18

To delve deeper into the complexities of respiratory medicine and related conditions such as PH-ILD, refer to the acclaimed Oxford Handbook of Respiratory Medicine.

**CLINICAL IMPLICATIONS**

The introduction of YUTREPIA® (treprostinil) inhalation powder for PH-ILD patients marks a pragmatic step in addressing a notoriously difficult-to-treat condition. For years, clinicians have grappled with the limited efficacy

and potential harm of applying PAH-specific therapies to PH-ILD, often resorting to supportive care or off-label use with trepidation. This new formulation, leveraging a known prostacyclin analogue, offers a more targeted and potentially safer approach, providing a much-needed option for patients whose disease progression is relentless.

From a prescribing clinician's perspective, the convenience of a dry powder inhaler for a prostacyclin analogue is a significant advantage. Intravenous or subcutaneous prostacyclins, while effective, carry substantial burdens related to administration, infection risk, and patient quality of life. An inhaled option, if proven to deliver comparable benefits with a more favorable tolerability profile, could improve adherence and broaden the accessibility of this class of medication. However, the onus remains on prescribers to carefully select patients, titrate doses, and monitor for adverse effects, particularly given the inherent fragility of PH-ILD patients.

For the industry, this development underscores the ongoing commercial interest in rare and complex diseases, where unmet needs can translate into significant market opportunities. The strategic presentation at a major respiratory conference like ATS, even in 2026, signals a commitment to establishing treprostinil's role in PH-ILD. While specific trial data were not provided for this summary, the success of such therapies will ultimately depend on robust evidence demonstrating clinically meaningful improvements in hard endpoints, beyond just hemodynamic parameters. Payers, too, will be scrutinizing these data to justify the cost of a specialized therapy in a chronic condition.



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