

Limited Distribution Notice for Opsumit, Tracleer, Uptravi, Veletri, Ventavis, and Zavesca

Notice to 340B Covered Entities

June 2020

This notice provides information to 340B covered entities regarding how to acquire Opsumit (macitentan), Tracleer (bosentan), Uptravi (selexipag), Veletri (epoprostenol), Ventavis (iloprost), and Zavesca (miglustat) at 340B ceiling prices. Actelion products are primarily dispensed through two specialty pharmacies which support the vast majority of Actelion patients.

Opsumit is an endothelin receptor antagonist (ERA) indicated for the treatment of pulmonary arterial hypertension (PAH, WHO Group I) to reduce the risks of disease progression and hospitalization for PAH. Effectiveness was established in a long-term study in PAH patients with predominantly WHO Functional Class II-III symptoms treated for an average of 2 years. Patients had idiopathic and heritable PAH (57%), PAH caused by connective tissue disorders (31%), and PAH caused by congenital heart disease with repaired shunts (8%). For all female patients, OPSUMIT is available only through a restricted program called the OPSUMIT Risk Evaluation and Mitigation Strategy (REMS) Program. Please see full prescribing information for OPSUMIT, including **BOXED WARNING**, at Opsumithcp.com

Tracleer is an ERA indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1):

- in adults to improve exercise ability and to decrease clinical worsening. Studies establishing effectiveness included predominantly patients with WHO Functional Class II-IV symptoms and etiologies of idiopathic or heritable PAH (60%), PAH associated with connective tissue diseases (21%), and PAH associated with congenital heart disease with left-to-right shunts (18%).
- in pediatric patients aged 3 years and older with idiopathic or congenital PAH to improve pulmonary vascular resistance (PVR), which is expected to result in an improvement in exercise ability.

Because of the risks of hepatotoxicity and birth defects, Tracleer is available only through a restricted program called the Bosentan REMS Program. Please see full prescribing information for Tracleer, including **BOXED WARNING**, at Tracleer.com/hcp.

Uptravi is a prostacyclin receptor agonist indicated for the treatment of pulmonary arterial hypertension (PAH, WHO Group I) to delay disease progression and reduce the risk of hospitalization for PAH. Effectiveness was established in a long-term study in PAH patients with WHO Functional Class II-III symptoms. Patients had idiopathic and heritable PAH (58%), PAH associated with connective tissue disease (29%), PAH associated with congenital heart disease with repaired shunts (10%). Please see full prescribing information for Uptravi at Uptravihcp.com.

Veletri is a prostanoid vasodilator indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to improve exercise capacity. Studies establishing effectiveness included predominantly patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH or PAH associated with connective tissue diseases. Please see full prescribing information for Veletri at Veletri.com.

Ventavis is a prostacyclin mimetic indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to improve a composite endpoint consisting of exercise tolerance, symptoms (NYHA Class), and lack of deterioration. Studies establishing effectiveness included predominately patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH (65%) or PAH associated with connective tissue diseases (23%). Please see full prescribing information for Ventavis at 4Ventavis.com.

Zavesca is a glucosylceramide synthase inhibitor indicated as monotherapy for the treatment of adult patients with mild to moderate type 1 Gaucher disease for whom enzyme replacement therapy is not a therapeutic option (e.g. due to allergy, hypersensitivity, or poor venous access). Please see full prescribing information for Zavesca at Zavesca.com.

340B covered entities can access 340B ceiling prices for these products through contract pharmacy arrangements with Accredo Specialty Pharmacy. The Accredo point of contact is Gene McCabe who can be reached at (615) 943-7500 or gene.mccabe@accredo.com. Orders must be placed with CuraScript Specialty Distribution by contacting Karen Caldwell at (407) 474-6298 or KCaldwell2@Curascript.com. CuraScript will facilitate bill to/ship to replenishment orders for the products at the 340B ceiling price. 340B covered entities will be charged the 340B ceiling price, and product will be shipped to Accredo Specialty Pharmacy.

Actelion is committed to compliance with the 340B program. If you have questions regarding access to 340B ceiling prices for Opsumit, Tracleer, Uptravi, Veletri, Ventavis, and Zavesca, please contact John O'Neil with Johnson & Johnson Health Care Systems at (732) 562-3770.