

dose. Only available through restricted program; prescribers, pharmacies, patients must enroll. Hepatotoxicity; monitor LFTs. Contraindicated in pregnancy. If pulmonary edema occurs, consider veno-occlusive disease; if confirmed, discontinue therapy. Patients with LV dysfunction may develop significant fluid retention and worsening heart failure after initiating endothelin receptor antagonist therapy; monitor for signs and symptoms of fluid retention. Monitor hemoglobin. May reduce sperm count. Many drug interactions; see prescribing information. [Trade only: Tabs, unscored 62.5, 125 mg. Tab for oral susp 32 mg.] ▶L ♀X/X/X, May cause fetal injury and death. ▶ Do not breastfeed while taking bosentan. \$\$\$\$\$ ■

EPOPROSTENOL (Flolan, Veletri) **Pulmonary arterial hypertension:** Start 2 ng/kg/min IV infusion via central venous catheter. Adjust dose based on response. Avoid abrupt dose decreases or cessation. If pulmonary edema occurs, consider veno-occlusive disease; if confirmed, discontinue therapy. ▶Plasma ♀B ▶? \$\$\$\$

ILOPROST (Ventavis) **Pulmonary arterial hypertension:** Start 2.5 mcg/dose by inhalation (as delivered at mouthpiece); if well tolerated, increase to 5 mcg/dose by inhalation (as delivered at mouthpiece). Use 6 to 9 times a day (minimum of 2 h between doses) while awake. Only administer with I-neb AAD Systems. Avoid contact with skin/eyes or oral ingestion. Monitor vital signs when initiating therapy. Do not initiate therapy if SBP less than 85 mmHg. Discontinue therapy if pulmonary edema occurs; this may be sign of pulmonary venous hypertension. May induce bronchospasm. ▶L ♀C ▶? \$\$\$\$\$

MACITENTAN (Opsumit) **Pulmonary arterial hypertension:** 10 mg PO daily. Contraindicated with pregnancy. Only available through restricted program; prescribers, pharmacies, female patients must enroll. Monitor liver function test before initiating therapy and as clinically indicated thereafter. If pulmonary edema occurs, consider veno-occlusive disease; if confirmed, discontinue therapy. Patients with LV dysfunction may develop significant fluid retention and worsening heart failure after initiating endothelin receptor antagonist therapy; monitor for signs and symptoms of fluid retention. [Trade: Tabs 10 mg.] ▶L ♀X/X/X, May cause fetal injury and death. ▶ Do not breastfeed while taking macitentan. \$\$\$\$\$ ■

RIOCIGUAT (Adempas) **Pulmonary arterial hypotension:** Start 1 mg PO three times daily; max 2.5 mg three times daily. Contraindicated with pregnancy, nitrates, nitric oxide donors, PDE-5 inhibitors (e.g., sildenafil, tadalafil, vardenafil), nonspecific PDE inhibitors (e.g., dipyridamole, theophylline). Only available through restricted program; prescribers, pharmacies, female patients must enroll. [Trade: Tabs 0.5, 1, 1.5, 2, 2.5 mg.] ▶LK ♂ ♀X/X/X; Contraindicated. Discontinue if pregnancy detected. ▶ Do not breastfeed while taking this medication. \$\$\$\$\$ ■

SELEXIPAG (Uptravi) **Pulmonary arterial hypertension:** Start 200 mcg PO two times daily, increase by 200 mcg PO two times daily each week to highest tolerated dose, max 1600 mcg PO two times daily. With moderate hepatic impairment (Childs-Pugh class B): Start 200 mcg PO daily, increase by 200 mcg PO daily each week to highest tolerated dose, max 1600 mcg PO daily. Avoid with severe

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