

Table 5. Current Approved Pharmacologic Therapies for Pulmonary Arterial Hypertension

<b>Name of Drug</b>	<b>Class of Drug</b>	<b>Route</b>	<b>Dose</b>	<b>Common Side Effects</b>	<b>Comments</b>
Epoprostenol (Flolan)	Prostanoid	IV	Start 1-2 ng/kg/min and titrate following side effect and PAH symptoms. Dosing must be individualized. Optimal dose for chronic therapy 25-40 ng/kg/min	Prostanoid side effects*	Central tunneled catheter needed  Long term data available  Effective in advanced PAH  Therapy complicated. Recommended patients be referred to PAH centers
Treprostinil (Remodulin)	Prostanoid	SC	Start 1.25 – 2.5 ng/kg/min and titrate following side effect and PAH symptoms	Prostanoid side effects*  Injection site pain and erythema	Site pain effects majority of patients  Experienced centers have reported successful outcome in managing patients with site pain  Long term survival data available
Treprostinil (Remodulin)	Prostanoid	IV	Start 2 ng/kg/min and	Prostanoid side effects*	More convenient for chronic infusion

			titrate following side effect and PAH symptoms.	Leg pain more common than epoprostenol	than epoprostenol  Revised recommendations for central tunneled catheter care
Treprostinil (Tyvaso)	Prostanoid	Inhaled	4 times a day	Prostanoid side effects* though generally less; also cough	Selective delivery of prostacyclin to lungs  Start 3 breaths QID; can increase up to 9-12 breaths QID as tolerated  Well tolerated as combination treatment with oral therapies
Iloprost (Ventavis)	Prostanoid	Inhaled	20 mcg, 6-9 inhalations a day	Prostanoid side effects* though generally less; also cough	Selective delivery of prostacyclin to lungs  Compliance can be an issue with need for frequent treatments  Well tolerated as combination treatment with oral therapies
Bosentan (Tracleer)	ERA	Oral	62.5 mg BID x 4 weeks then 125 mg BID if LFT normal	Headache, dizziness, edema	Need LFTs checked monthly  Most effective in non-FC IV patients  Contraindicated with cyclosporine and Glyburide  Decreases effectiveness of oral hormonal contraceptives  Drug interaction with sildenafil  Long-term

					observed survival data available
Ambrisentan (Letairis)	ERA	Oral	5 mg QD or 10 mg QD	Peripheral edema, nasal congestion, sinusitis	More reported incidence of edema in elderly  Decreases effectiveness of oral hormonal contraceptives  No drug interaction observed in combined treatment with sildenafil
Sildenafil (Revatio)	PDE5-inhibitor	Oral	20 mg TID	Epistaxis, Headache, flushing, diarrhea	Contraindicated with nitrates  Some patients may need up titration of dose
Tadalafil (Adcirca)	PDE5-inhibitor	Oral	40 mg QD	Headache, myalgia, flushing	Contraindicated with nitrates

\*Side effects related to prostacyclin: jaw pain, diarrhea, flushing, headache, nausea  
ERA: Endothelin Receptor Antagonists; PDE-5 Inhibitor: Phosphodiesterase-5 Inhibitor