

Drugs currently used for treatment of PAH

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Class	Medication	Mechanism	Route	Starting	Maintenance	Half-life	Company	Available
Standard	Digoxin	Inhibits Na ⁺ pump	PO/IV	0.0625-0.25 mg daily	Increase q2 wks for desired effect	36-48 hrs	GSK	Globally
Diuretic	Multiple	Renal-based	PO/IV	Varies	Titrate as needed	Varies	Generic	Globally
Anti-coagulation	Warfarin	Vitamin K antagonist	PO	Varies	Goal INR 2.0	40 hrs	Generic	Globally
Calcium channel blockers	Nifedipine XR	Block VDCC	PO	30mg daily	Titrate to max dose as tolerated	7 hrs	Pfizer	Globally
	Diltiazem XR	Block VDCC	PO/IV	120mg daily	Titrate to max dose as tolerated	5-10 hrs	Generic	Globally
	Amlodipine	Block L-type VDCC	PO	5mg daily	Titrate to max dose as tolerated	30-50 hrs	Generic	Globally
Nitric oxide	NO	Increases cGMP	Inh	Acute test-10-20ppm for 10 min		15-30 sec	INO Therapeutics	Globally
Adenosine	Adenosine	Via receptors A ₁ , A _{2A} , A _{2B} , A ₃	IV	Acute testing: 50 mg/kg/min	Titrate to symptoms or 350mcg/kg/min	<10 sec	Astellas Pharma US	Globally
Endothelin receptor antagonists (ERAs)	Bosentan	Block both ET _A and ET _B	PO	62.5mg b.i.d	Increase to 125 mg, b.i.d. after 4 wks	5 hrs	Actelion	Globally
	Ambrisentan	Block ET _A only	PO	5mg daily	Consider increase to 10 mg daily	9 hrs	Gilead	Globally
	Sitaxsentan	Blocks ET _A > ET _B	PO	100mg daily		10 hrs	Pfizer	
PDE inhibitors	Sildenafil	PDE5 Inhibition	PO	20mg 3 times daily		4 hrs	Pfizer	Globally
	Tadalafil	PDE5 Inhibition	PO	40mg daily		17.5 hrs	United Therapeutics	Globally
Prostacyclinalogues	Beraprost	PGI ₂ analogue	PO	20g, q.i.d.	Increase by 20g, q.i.d., as tolerated	35-40 min	United Therapeutics	Asia
	Iloprost	PGI ₂ analogue	Inh	2.5mg 6-9 times daily	Increase to 5 mg 6-9 times daily	20-30 min	Actelion	Globally
	Treprostinil	PGI ₂ analogue	Inh	6 mg, q.i.d.	Up to 18 mg, q.i.d.	4 hrs	Lung Rx, Inc	
Iloprost	PGI ₂ analogue	IV	1-2ng/kg/min		13 min	Schering Healthcare	not in USA	
	Epoprostenol	PGI ₂ analogue	IV	2ng/kg/min	Increase every 15 min until side effects occur	3-5 min	Actelion	Globally
	Treprostinil	PGI ₂ analogue	IV	1.25ng/kg/min	Increase <1.25 ng/kg/min q wk for 4 wks and after <2.5 ng/kg/min q wk	4-5 hrs	United Therapeutics	Globally
	Treprostinil	PGI ₂ analogue	SC	1.25ng/kg/min	Increase <1.25 ng/kg/min q wk for 4 wks and after <2.5 ng/kg/min q wk	4-5 hrs	United Therapeutics	Globally

Abbreviations: **PAH:** pulmonary arterial hypertension; **PO:** oral; **IV:** intravenous; **Inh:** inhalation; **PDE:** phosphodiesterase; **VDCC:** voltage-dependent calcium channels; **SC:** subcutaneous; **b.i.d.:** twice a day; **q.i.d.:** four times a day.

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