

Overview of treatment of PAH

Recommendation	Level of Evidence	Net Benefit
<p><i>Calcium channel blockers (CCBs)</i> Acute responders to vasoreactivity testing, defined as fall in mean PAP ≥ 10 mmHg to ≤ 40 mmHg with increased or unchanged cardiac output, and absence of right heart failure may be treated with oral CCBs, with careful reassessment:</p> <ol style="list-style-type: none"> 1. Idiopathic PAH 2. Secondary PAH from underlying conditions 	<p>low expert opinion</p>	<p>substantial intermediate</p>
<p><i>Sildenafil</i> Patients who are not candidates or in whom CCB therapy has failed can initiate long-term therapy for the following functional class:</p> <ol style="list-style-type: none"> 1. II or III 2. IV 	<p>good low</p>	<p>substantial indeterminate</p>
<p><i>Intravenous epoprostenol</i> Patients who are not candidates or in whom CCB therapy has failed can initiate long-term therapy for the following functional class:</p> <ol style="list-style-type: none"> 1. III or IV 	<p>good</p>	<p>substantial</p>
<p><i>Treprostinil</i> Patients who are not candidates or in whom CCB therapy has failed can initiate long-term therapy for the following functional class:</p> <ol style="list-style-type: none"> 1. II <ol style="list-style-type: none"> a. Subcutaneous or intravenous 2. III or IV <ol style="list-style-type: none"> a. Subcutaneous b. Intravenous 	<p>low fair low</p>	<p>small/weak intermediate intermediate</p>
<p><i>Inhaled iloprost</i> Patients who are not candidates or in whom CCB therapy has failed can initiate long-term therapy for the following functional class:</p> <ol style="list-style-type: none"> 1. III 2. IV 	<p>good fair</p>	<p>intermediate intermediate</p>
<p><i>Bosentan</i> Patients who are not candidates or in whom CCB therapy has failed can initiate long-term therapy for the following functional class:</p> <ol style="list-style-type: none"> 1. III 2. IV 	<p>good fair</p>	<p>substantial intermediate</p>

Strength of Recommendation Scale: A, strong; B, moderate; C, weak; D, negative; I, inconclusive; E/X, expert opinion only/conditional