

Therapeutic Management

Although no cure is known, several therapies have shown promise in slowing the progression of the disease and improving quality of life. In general, situations that may exacerbate the disease and cause hypoxia, such as exercise and high altitudes, are avoided. Supplemental oxygen, especially at night while sleeping, is commonly used to relieve hypoxia. Patients are at risk for thromboembolic events leading to pulmonary emboli, so anticoagulation with warfarin (Coumadin) is often prescribed.

A number of new drug therapies have been used in this patient population and have promise in improving quality of life and survival. Several studies and newer approaches in treatment emphasize combined therapy that targets each of the major pathways of the disease process rather than monotherapy approaches.

Vasodilator therapy (which relaxes vascular smooth muscle and reduces pulmonary artery pressure) can prolong survival of patients with PAH. Oral calcium channel blockers have been successful in some children. For patients who are nonresponders in vasodilator testing, a new oral drug, bosentan (an endothelin-receptor antagonist), is now available that reduces pulmonary artery pressure and resistance and is safe and well tolerated in children ([Barst, Ivy, Dingemans, et al, 2003](#)). It has been used in combination with IV prostacyclin.

Lung transplantation may be another treatment option for those with severe disease. Patients with pulmonary hypertension have had a higher mortality rate than after lung transplantation than other lung transplant patients. The management of PAH continues to evolve as new information is learned and new combination therapies are tested and evaluated.

Quality Patient Outcomes: Hypertension

- Underlying cause of hypertension identified
- Blood pressure (BP) control maintained
- Dietary practices and lifestyle changes effectively used to control hypertension