**Pulmonary Hypertension-***The Other High Blood Pressure*

Pulmonary arterial hypertension (PAH) is a rare disorder of the pulmonary vasculature which results in increasing resistance to blood flow through the lungs. It is defined as a mean pulmonary artery pressure greater than 25 mmHg at rest or greater than 30 mm of mercury with exercise. Because its symptoms mimic those of other diseases, and because standard testing in this situation is often unrevealing, the time from the onset of symptoms to diagnosis can reach up to two years. Unfortunately, pulmonary arterial hypertension is a life-threatening disease with a very high mortality if left untreated.

Formerly classified as either Primary or Secondary, the World Health Association (WHO) developed a new classification system in 1998, to aid in the appropriate diagnosis and treatment of pulmonary hypertension. (See illustration)

Symptoms typically include dyspnea and decreases in exercise capacity. EKG and CXR abnormalities, lower extremity edema, ascites, chest discomfort and syncope are late findings.

Severity of disease is graded based on the New York heart Association classes for congestive heart failure. Greater severity of symptoms correlates with greater mortality.

Diagnosis of PAH requires a high clinical suspicion. Cardiac echocardiography is the screening test of choice, and may reveal elevations in pulmonary artery systolic pressure, right sided chamber enlargement, and tricuspid regurgitation. It is also helpful in evaluating for left sided cardiac disease which may be contributing to the PH. Additional evaluation should include a thorough history and physical examination, laboratory screening for connective tissue diseases, thyroid disorders, blood dyscrasias and hypercoagulable states, VQ scanning, pulmonary function testing with measurement of diffusing capacity and oxygenation, screening for portal hypertension, EKG and CXR. Additionally, High Resolution CT of the chest may also be helpful to exclude pulmonary fibrosis and mediastinal abnormalities; a sleep study may identify patients with sleep disorders. An echo bubble study or trans-esophageal echo can non-invasively detect congenital cardiac abnormalities. Functional testing in the form of a 6 minute walk is helpful to determine the severity of the patient's disease and to follow the results of therapy. Prognostically, walking less than 350 m predicts a poorer long-term prognosis.

All patients with suspected pulmonary hypertension should undergo cardiac catheterization. Echocardiography alone is inadequate to diagnose pulmonary hypertension as it sometimes over or under estimates the severity of disease. Catheterization also provides additional prognostic data such as right atrial pressure and cardiac output which are essential in diagnosis and management. Catheterization is also used to exclude left-sided heart disease which, if present, could complicate or increase the risks associated with standard therapies for pulmonary hypertension. Pulmonary vasodilator testing should be performed in most patients. While vaso-reactivity is present in only 10 to 15% of patients, these patients often respond well to treatment with supra-therapeutic doses of calcium channel blockers. Vaso-reactivity testing should only be performed with the short acting, pulmonary selective agents epoprostenol, adenosine or nitric oxide. Catheterization in these patients should only be performed by clinicians experienced in the diagnosis and treatment of pulmonary hypertension.

Rapid advances in the understanding and treatment of pulmonary hypertension have occurred over the last several years. Epoprostenol (Flolan), the first available treatment for PAH, was approved as recently as 1995. While extremely expensive and complicated to use, its introduction has vastly improved the prognosis of patients with PAH. Because of it’s extremely duration of action, it must be infused continuously via an indwelling intravenous catheter. Two other prostanoid compounds are now available. Treprostinil (Remodulin) improves on Flolan with a longer duration of action, and is available for both intravenous and subcutaneous continuous infusion. Iloprost (Ventavis) is an inhaled prostanoid which is administered via nebulizer 6 - 8 times daily.

The endothelin receptor antagonists (ERA’s), bosentan (Tracleer) and ambrisentan (Letairis) as well as Sildenafil (Revatio) are oral therapies that improve symptoms and 6 minute walk distance. ERA’s are contraindicated in patients with hepatic disease and during pregnancy. Monthly hepatic enzyme monitoring is required. While patients with more severe symptoms, (Class 4 and some Class 3), should be treated with prostanoid therapy, those with milder therapies can often be treated successfully with orally. Very close follow-up and vigilance for progression of symptoms is essential, as most patients will progress, even with therapy.

For patients who fail medical therapy, lung transplantation should be considered. Dramatic improvements in symptoms, right heart function and survival are accomplished with lung transplantation; however long-term morbidity from obliterative bronchiolitis is substantial. Surgical thrombo-endarterectomy is a very effective therapy in patients with chronic thromboembolic pulmonary hypertension (CTEPH). This is available in only a few centers around the world.

PAH is a rare but increasingly recognized disease for which significant improvements in treatment have recently become available. Diagnosis requires a high index of suspicion, and diagnosis and treatment should be undertaken by physicians intimately familiar with the disease.

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