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Pulmonary Hypertension

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Pulmonary hypertension (PH) is a disorder in which the pressure in the pulmonary arteries is above normal.[1] People with PH develop a markedly decreased exercise tolerance and quality of life. PH is a severe and life-threatening disease that can lead to heart failure and death.[2]

Prevalence of Pulmonary Hypertension

PH affects people worldwide and encompasses multiple disease subtypes.[3] Pulmonary arterial hypertension (PAH) affects an estimated 30-50 people per million but this is considered to be only a small portion of the overall PH population.[4] If left untreated, the median survival is only 2.8 years following diagnosis.[5]

Signs, Symptoms and Diagnosis

Early PH is often asymptomatic. Unfortunately, by the time symptoms appear, disease progression is usually well advanced. The most common symptoms of PH include shortness of breath with physical exercise (exertional dyspnea), fatigue, dizziness and fainting, all of which are worsened by exertion.

As PH signs and symptoms are non-specific and the disease is rare, diagnosis is often delayed. While echocardiogram, electrocardiogram, exercise testing (e.g. six-minute walk test), certain blood markers (e.g. cardiac troponin T and/or B-type natriuretic peptide) and other tests can be helpful, a definitive diagnosis requires inserting a special pressure-sensing catheter into the right side of the heart.

Medical Treatment for Pulmonary Hypertension

PH encompasses multiple disease subtypes, but currently existing treatments are indicated solely for pulmonary arterial hypertension (PAH), which accounts for only a small portion of overall PH patients.[6] Fortunately, the management of PAH has evolved significantly over the past decade.[7] For example, Bayer's Ventavis® (iloprost), the only approved inhaled PAH treatment worldwide, has had a positive impact on exercise capacity, functional class, hemodynamics and health-related quality of life (HRQoL) of patients.[8] However, despite these improvements, the prognosis for PH patients remains poor underscoring the need for new treatments.

Types of Pulmonary Hypertension

According to the World Health Organization (WHO) classification, there are five different types of PH, based on the underlying causes of PH, which are as follows:[3]

1. Pulmonary arterial hypertension (PAH)
   - 1.1. Idiopathic (IPAH) – Cause unknown
   - 1.2. Familial (FPAH) – Genetic background
   - 1.3. Associated with (APAH) – Collagen vascular disease, congenital systemic-to-pulmonary shunts, portal hypertension, HIV infection, drugs and toxins, etc.
   - 1.4. Associated with significant venous or capillary involvement – Pulmonary veno-occlusive disease (PVOD), pulmonary capillary hemangiomatosis (PCH)
   - 1.5. Persistent pulmonary hypertension of the newborn (PPHN)

2. Pulmonary hypertension with left heart disease
   Left-sided ventricular or atrial heart disease, left-sided valvular heart disease

3. Pulmonary hypertension associated with lung disease and/or hypoxemia
   Chronic obstructive pulmonary disease (COPD), interstitial lung disease, sleep-disordered breathing, alveolar hypo-ventilation disorders, chronic exposure to high altitude, developmental abnormalities

4. Pulmonary hypertension due to chronic thrombotic and/or embolic disease (CTEPH)
   Thromboembolic obstruction of the proximal/distal pulmonary arteries; non-thrombotic pulmonary embolism (tumor, parasites, foreign material)

5. Miscellaneous
   Sarcoidosis, histiocytosis X, lymphangiomatosis, compression of pulmonary vessels (adenopathy, tumor, fibrosing mediastinitis, etc.)

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