Brief Note

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Archives of Medicine
ISSN 1989-5216

2021
Vol.13 No.8:39

Brief Note on Pulmonary Hypertension

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Citation: Zeytun D (2021) Brief Note on Pulmonary Hypertension. Arch Med Vol. 13 No. 8: 39

Received: August 20, 2021; Accepted: August 25, 2021; Published: August 30, 2021

Brief Note

Pulmonary hypertension is a fatal disease of multiple etiologies that is estimated to affect over 100 million people worldwide. The disease is defined hemodynamically as a mean pulmonary artery pressure ≥ 25 mmHg at rest. Pulmonary hypertension is a type of high blood pressure that affects the arteries in your lungs and the right side of your heart. The pulmonary circulation is responsible for carrying deoxygenated blood from the heart to the lungs and returning oxygenated blood back to the heart for delivery to the systemic circulation. The pulmonary circulation has a central role in cardiopulmonary gas exchange and oxygen transport. In one sort of pulmonary hypertension, called Pulmonary Arterial Hypertension (PAH), blood vessels in your lungs are narrowed, congested or damaged. The damage slows blood flow through your lungs, and blood pressure in the lung arteries rises. Your heart must work harder to pump blood through your lungs. The extra effort eventually causes your heart muscle to develop weak and fail.

There are three types of pulmonary arterial hypertension based on the origin of the cause: idiopathic, heritable, and drug and toxin-induced. Pulmonary hypertension was previously divided into primary and secondary categories; primary pulmonary hypertension described an idiopathic hypertensive vasculopathy exclusively affecting the pulmonary circulation. Whereas secondary pulmonary hypertension was associated with a causal underlying disease process. The histological appearance of lung tissue in each of these conditions is similar and consists of intimal fibrosis, increased medial thickness, pulmonary arteriolar occlusion and plexiform lesions.

Recent advances in clinical recognition, classification, and understanding of the underlying pathological processes in pulmonary hypertension have led to improved diagnostic testing and therapeutic options for patients. Clinically, these pulmonary vascular changes initially present as nonspecific symptoms, including unexplained dyspnea on exertion, fatigue, chest pain, and syncope.

Symptoms

The signs and symptoms of pulmonary hypertension develop slowly. You may not notice them for months or even years. Symptoms get worse as the disease progresses.

Pulmonary hypertension symptoms include:

- Shortness of breath (dyspnea), initially while exercising and eventually while at rest
- Fatigue
- Dizziness or fainting spells (syncope)
- Chest pressure or pain
- Swelling (edema) in your ankles, legs and eventually in your abdomen (ascites)
- Bluish color to your lips and skin (cyanosis)
- Racing pulse or heart palpitations

Pulmonary hypertension caused by chronic blood clots

- Chronic blood clots in the lungs (pulmonary emboli)
- Other clotting disorders

Pulmonary hypertension triggered by other health conditions

- Metabolic disorders, including glycogen storage disease
- Inflammatory disorders such as sarcoidosis and vasculitis
- Tumors pressing against pulmonary arteries
- Kidney disease
- Blood disorders, including polycythemia vera and essential thrombocythemia

Risk factors

Growing older can raise your risk of developing pulmonary hypertension. The condition is more often detected in people ages 30 to 60. However, idiopathic PAH is more common in younger adults.

Other things that can raise your risk of pulmonary hypertension include:

A family history of the condition

- Blood-clotting disorders or a family history of blood clots in the lungs
- Being overweight
- Use of Selective Serotonin Reuptake Inhibitors (SSRIs), used to treat depression and anxiety
- Genetic disorders, including congenital heart disease
- Living at a high altitude
- Use of certain weight-loss drugs
- Use of illegal drugs such as cocaine
- Exposure to asbestos

Pulmonary hypertension due to lung diseases and/or hypoxia

- Chronic obstructive pulmonary disease
- Interstitial lung disease
- Chronic exposure to high altitude
- Other pulmonary diseases with mixed restrictive and obstructive pattern
- Sleep-disordered breathing
- Developmental lung diseases
- Alveolar hypoventilation disorders

Diagnostics

The diagnostic algorithm for Pulmonary Arterial Hypertension (PAH), Electrocardiographic (ECG) signs of the right heart compromise contain right axis deviation, right ventricular hypertrophy and peaked p waves. However, the ECG lacks sufficient diagnostic accuracy to serve as a screening tool for the detection of PAH. The chest x ray may reveal abnormal anatomic features due to Pulmonary Arterial Hypertension (PAH) and its potential causes, it is usually normal in those without symptoms. The most useful and accessible imaging modality for diagnostic purposes is echocardiography.

Many drugs and toxins are known to induce or be associated with development of PAH and are categorized by strength of evidence in the newest guidelines. However, the accurate mechanisms of most of these compounds remain to be elucidated. Those with definite associated with PAH development include certain anorexigens (e.g. dexfenfluramine, aminorex, fenfluramine), benfluorex, toxic rapeseed oil, and Selective Serotonin Reuptake Inhibitors (SSRIs).

Identifying cause of pulmonary hypertension is essential in order to give specific treatment for etiology of pulmonary hypertension and prevent its further complication. While not all pulmonary hypertension can be prevented, you can take steps to prevent it by making healthy lifestyle changes and managing high blood pressure, coronary heart disease, chronic liver disease, and chronic lung disease from tobacco use.