A comprehensive review of idiopathic pulmonary arterial hypertension: Epidemiology, pathogenesis and management.

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Abstract

Idiopathic Pulmonary Arterial Hypertension (IPAH) is a rare and life-threatening disease that affects the pulmonary vasculature. It is characterized by increased pulmonary artery pressure and resistance, leading to right heart failure and death. The exact cause of IPAH is unknown, but it is thought to be a complex interplay between genetic, environmental, and epigenetic factors. The management of IPAH involves a multidisciplinary approach, including pharmacotherapy, oxygen therapy, and supportive care. In this mini-review, we provide a comprehensive overview of the epidemiology, pathogenesis, and management of IPAH.

Keywords: Idiopathic pulmonary arterial hypertension, Disease, Pulmonary vasculature.

Introduction

Idiopathic Pulmonary Arterial Hypertension (IPAH) is a rare and life-threatening disease that affects the pulmonary vasculature. It is characterized by increased pulmonary artery pressure and resistance, leading to right heart failure and death [1]. The exact cause of IPAH is unknown, but it is thought to be a complex interplay between genetic, environmental, and epigenetic factors.

IPAH is a rare disease, with an estimated prevalence of 5-15 cases per million adults worldwide. It is more common in females than males and typically affects individuals between the ages of 20 and 50 years [2]. IPAH can occur in isolation or as part of other conditions, such as connective tissue disorders or congenital heart disease.

The pathogenesis of IPAH involves several interconnected pathways, including endothelial dysfunction, smooth muscle cell proliferation, and inflammation. These pathways lead to the remodelling of the pulmonary arteries, resulting in increased resistance and pressure [3].

Genetic factors are thought to play a significant role in the pathogenesis of IPAH. Mutations in several genes, including BMPR2, ALK1, ENG, SMAD9, and CAV1, have been identified in patients with IPAH. These genes are involved in the regulation of vascular homeostasis and the response to injury [4].

Environmental factors, such as exposure to toxins and infections, may also contribute to the development of IPAH. For example, exposure to drugs such as fenfluramine and dexfenfluramine has been associated with an increased risk of IPAH.

The management of IPAH involves a multidisciplinary approach, including pharmacotherapy, oxygen therapy, and supportive care. The goal of treatment is to improve symptoms, quality of life, and survival [5].

Several classes of medications are used to treat IPAH, including prostacyclin analogs, endothelin receptor antagonists, and phosphodiesterase type 5 inhibitors. These medications act on different pathways involved in the pathogenesis of IPAH and can improve pulmonary artery pressure, exercise capacity, and survival.

Oxygen therapy can improve symptoms and quality of life in patients with IPAH who have hypoxemia. It can also reduce the risk of right heart failure and improve survival [6].

Supportive care includes measures such as diuretics, anticoagulants, and vaccination against influenza and pneumococcal infections. These measures can reduce the risk of complications and improve outcomes in patients with IPAH.

Lung transplantation may be considered in patients with advanced IPAH who are refractory to medical therapy. It can improve survival and quality of life in selected patients.

Conclusion

IPAH is a rare and life-threatening disease that affects the pulmonary vasculature. The exact cause of IPAH is unknown, but it is thought to be a complex interplay between genetic, environmental, and epigenetic factors. The management of IPAH involves a multidisciplinary approach, including pharmacotherapy, oxygen therapy, and supportive care. Early diagnosis and treatment are essential to improve outcomes in patients with IPAH.

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