LEARNING THE ROPES: PULMONARY HYPERTENSION

Introduction

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Pulmonary hypertension is defined as an invasively measured mean pulmonary arterial pressure (mPAP) of more than 20-25 mm Hg at rest (normal values are 12 ± 2 mm Hg) or more than 30 mm Hg during exercise. Non-invasive measurements by echocardiography provide estimated systolic pulmonary arterial pressures (sPAP), and are considered normal if below 35-40 mm Hg. Pulmonary hypertension may appear spontaneously, with no known underlying disease, or it can occur in connection with other conditions. While pulmonary arterial hypertension (previously known as primary pulmonary hypertension) has a relatively low prevalence (approximately 10-15 cases per 1 000 000 people), connections to other diseases such as pulmonary thromboembolism, chronic lung diseases including fibrosis, hypertensive heart disease, and chronic heart failure are more frequent. The most common cause of pulmonary hypertension, however, is chronic obstructive pulmonary disease (COPD).

Patients (mostly women) with pulmonary hypertension have an insidious onset of exertional dyspnoea and their condition often comes to attention of the doctor late in the course of the illness or when symptoms and signs of right ventricular heart failure develop.

Since pulmonary hypertension is a condition caused by a heterogeneous group of diseases, treatment should be directed at its primary cause(s). Patients who have symptoms resulting in a limitation of physical activity and those who have symptoms at rest used to have a poor prognosis, with a short (2-3 years) survival from the time of diagnosis. With modern therapy the prognosis has improved. The cause of death is usually acute right ventricular heart failure or sudden death.

Below, we have invited a group of distinguished authors to discuss pulmonary hypertension with a focus on genetics, pathophysiology, treatment (now and in the years to come) and clinic. Have a good read of four excellent papers on a topic many of us know little about!

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