

Comments

Points in common, but different origins

Pulmonary hypertension is a complex, multidisciplinary disease for which the classification and guidelines for diagnostics and treatment were revised in 2009 (1, 2). The mean pressure in arteria pulmonalis is by definition ≥ 25 mm Hg at rest, determined by means of right-sided heart catheterisation. In the precapillary type of pulmonary hypertension (Groups 1, 3, 4, and 5) pulmonary artery wedge pressure is ≤ 15 mm Hg and reflects normal pressure in the left atrium. This is in contrast to post-capillary pulmonary hypertension with > 15 mm Hg (Group 2), reflecting increased filling pressure in the left side of the heart. The division into five groups is based on pathological, pathophysiological and therapeutic characteristics.

Group 1, *Pulmonary artery hypertension (PAH)*, is a disease that affects the small lung arteries, first in the form of vasoconstriction and subsequently through remodeling and structural changes that increase pulmonary vascular resistance. Small vessels are gradually obstructed, and the right ventricle encounters a resistance that over time causes reduced function and cardiac output. Without treatment the prognosis is very poor, and even though modern medication (endothelin receptor blockers, phosphodiesterase type 5 inhibitors and prostanoids) have improved life expectancy, there is no curative treatment (3). In addition to an idiopathic form there are also associated forms occurring in connective tissue disease, liver failure, HIV and congenital heart defect.

Group 2, *Pulmonary hypertension with left-sided heart disease*, occurs secondarily to failing systolic or diastolic function of the left ventricle and in connection with valve defects. Over time, increased filling pressure on the left side will be transmitted to the pulmonary vasculature with an increase in venous pressure, which in turn may be transmitted to the capillaries and the arterial side. Medicines specific to pulmonary arterial

hypertension are contraindicated with left-sided heart failure and may lower the patients' life expectancy (1, 2).

Group 3, *Pulmonary hypertension in lung diseases*, also appears to have a complex background, but is primarily conditioned by chronic hypoxaemia (4). This group is most often characterised by lower pressure and less haemodynamic disturbances than the two previous groups, also in the case of advanced disease. There is no documentation for measures other than chronic oxygen treatment (4).

Group 4, *Pulmonary hypertension with chronic thromboembolism*, is a disease that is potentially curable by means of thromboendarterectomy. Drugs specifically intended for pulmonary arterial hypertension have now shown definite beneficial effects in randomised trials (5), and no documented treatment option currently exists for those who are not candidates for surgery.

Group 5, *Pulmonary hypertension in cases with unclear and/or multifactorial mechanisms*. Pulmonary tumour thrombotic microangiopathy, which the patient in question had, is classified here together with haematological conditions such as splenectomy, systemic diseases such as sarcoidosis and metabolic disorders such as Gaucher's disease. This is an umbrella group conditioned by rare diseases and with no consensus regarding treatment. The case study refers to the many diagnostic aids and differential diagnostic considerations during evaluation, and convincing arguments are made for cross-disciplinary cooperation. A description is given of the fate of many patients in all five groups, despite treatment, with progressive right-sided heart failure and death. From a cardiological point of view only right-sided heart catheterisation (with aspiration of tumour cells in wedge pulmonary artery position), in addition to echocardiography, could have given an ante mortem diagnosis. Whereas a definite diagnosis is normally cru-

cial for correct treatment, in this case it would have been of uncertain value.

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