key points

SELECTED BY

Dr Peter Savill
Former GP, Community Cardiology Specialist, Southampton and Mid Hants, UK

Pulmonary hypertension (PH) is a haemodynamic state where the mean pulmonary artery pressure measured at cardiac catheterisation is ≥ 25 mmHg. Precapillary PH arises from increased resistance to blood flow in the pulmonary arterioles and postcapillary PH from elevated left atrial pressure. In postcapillary PH the cause is left heart disease whereas precapillary PH may be caused by any other form of PH. Patients develop symptoms only when the disease is advanced. The symptoms at clinical presentation are non specific. Adults almost universally present with breathlessness and they may also complain of exercise-induced dizziness or syncope (an ominous sign) and angina pectoris.

An echocardiogram is the best investigation to ascertain the probability of PH. This is estimated from the peak velocity of the tricuspid regurgitation jet using continuous wave Doppler and integrated with other echo signs of PH. Based on the probability of PH, appropriate referral can be made for further investigation. Echo may also identify a cardiac cause for PH.

An ECG and chest radiograph will be normal in 10% of patients at presentation. When abnormal, the ECG may show right axis deviation, right bundle branch block, right atrial enlargement and right ventricular hypertrophy. The chest radiograph may show pulmonary artery and cardiac enlargement. Pulmonary function tests are important to look for airways disease and parenchymal lung disease. Blood tests including a full blood count, renal, liver, thyroid function, autoantibodies and HIV may reveal a cause for PH and its differential diagnosis. Further imaging and invasive haemodynamic investigations in hospital are mandatory to confirm diagnosis.

Patients who are suspected of having PH and who have already had an echo showing an intermediate or high probability of PH should be referred directly to the UK National Pulmonary Hypertension Service at one of seven designated adult centres. Patients who have not had an echo should be referred to cardiology or respiratory medicine.

For patients with idiopathic, heritable or drug-induced PAH, a vasoreactivity study carried out at cardiac catheterisation determines their suitability for high-dose calcium channel blocker treatment. For patients with a negative vasoreactivity study or another cause of PAH, specialist PH drug therapies have been developed. There are now 11 licensed vasodilator drugs. For most patients, initial dual combination therapy is recommended. Failure to achieve low-risk status indicates the need for a third drug. If triple therapy is inadequate then bilateral sequential lung transplantation should be considered in eligible patients. General measures include avoiding pregnancy which carries a high maternal mortality, vaccination against influenza and pneumococcal pneumonia, diuretic management of fluid retention, and psychological and social support.