key points

SELECTED BY

Dr Peter Saul
GP, Wrexham and Associate GP Dean for North Wales

Pulmonary hypertension (PH) is defined as an increase in mean pulmonary arterial pressure ≥ 25 mmHg at rest as assessed invasively by right heart catheterisation. It can affect patients at any age and presents with non-specific symptoms. Accurate diagnosis is important as while PH is a potentially lethal disease it is treatable. Identification of the cause of PH is crucial to ensure that the patient receives appropriate management. The most common causes are heart and lung disease and specialist drug therapies for PH are contraindicated in these patients. However, PH is also associated with a diverse group of diseases.

The most common symptom of PH is breathlessness on exertion. Syncope may be the presenting symptom especially in children. Other PH symptoms include: angina pectoris, palpitations, dry cough, exertional nausea and vomiting. Lack of response to conventional treatment for dyspnoea should alert the GP to search for another cause of the patient's symptoms, one of which could be PH.

Syncope is one of the most serious manifestations of clinical deterioration in PH patients. On clinical examination the most common clinical signs are: presence of right ventricular heave, loud pulmonary component of the second heart sound and a pansystolic murmur of tricuspid regurgitation. In the presence of heart failure, clinical examination may also reveal elevated jugular venous pressure, ascites and peripheral oedema.

Patients should have an ECG, a chest radiograph and a full blood count, renal and liver function as well as thyroid function tests. When PH is suspected, referral to the local cardiology or respiratory department is recommended. This referral should be urgent if the patient is syncopal, has rapidly progressing symptoms or is in heart failure. The investigation of choice for detecting elevated pulmonary pressures is the echocardiogram. If the echocardiogram shows signs of PH then the patient should be referred to a designated PH centre if pulmonary arterial hypertension (PAH) or chronic thromboembolic pulmonary hypertension (CTEPH) are suspected or if PH is of uncertain cause.

In patients with CTEPH the mainstay of treatment is warfarin. Most patients with PAH are also treated with warfarin. PAH drugs are prescribed by the PH team but GPs should be familiar with these drugs in order to recognise their possible side effects and avoid interactions with other medications. PAH drugs should never be stopped or downtitrated without discussion with the PH team. PAH patients are susceptible to developing pneumonia, which may not be accompanied by fever, with an increased mortality risk. Vaccination against influenza and pneumococcal pneumonia is recommended.