Guidelines for referring patients with pulmonary hypertension

What is pulmonary hypertension?
Pulmonary hypertension describes a diverse group of diseases which result in mean pulmonary artery pressure ≥ 25mmHg at rest. The underlying pathophysiology is that of progressive pulmonary vascular remodelling and subsequent right ventricular dysfunction and failure.

Current estimated incidence and prevalence
Whilst idiopathic pulmonary arterial hypertension (formerly primary pulmonary hypertension) remains an uncommon condition pulmonary hypertension associated with other conditions is not that rare. The recognition of pulmonary arterial hypertension is increasing (incidence currently estimated at 8 cases/ million/year, prevalence 20-50 cases /million). The availability of effective therapies and the increased recognition of pulmonary arterial hypertension in association with other conditions has resulted in the development of nationally designated specialist centres such as the Pulmonary Vascular Disease Unit (PVDU) at Papworth Hospital.

The incidence of chronic thromboembolic pulmonary hypertension is also greater than initially suggested (up to 4% of patients with pulmonary embolism after two years). Papworth Hospital has also been commissioned as the national referral centre for pulmonary endarterectomy.

Suggestive Symptoms

- Fatigue
- Exertional dyspnoea
- Syncope/presyncope
- Peripheral oedema
- Atypical chest pain
- Palpitations
- +/- Raynaud's phenomenon
- Systemic sclerosis/connective tissue disease

Associated Risk Factors

- Congenital heart disease
- Family history of IPAH
- Recurrent pulmonary embolism
- HIV infection
- Advanced liver disease
- Known history of venous thromboembolism

Who to refer
The PVDU encourages you to promptly refer any patient aged older than 16 whom you believe has:

- Unexplained pulmonary hypertension
- Pulmonary arterial hypertension (see causes above)
- Chronic thromboembolic pulmonary hypertension
- Miscellaneous causes of pulmonary hypertension

Patients with PH associated to left heart disease and/or lung disease are reviewed on an individual basis.

Echocardiography should be used as the primary screening modality, with pulmonary hypertension being suggested by:

- Estimated pulmonary artery systolic pressure of ≥ 40mmHg + RA pressure (Tricuspid regurgitant jet velocity of 2-8-3.4m/s)
- Evidence of significant right ventricular dilation or dysfunction
- **Absence of left ventricular dysfunction or significant mitral or aortic valve disease**

For patients with systemic sclerosis we recommend using the DETECT screening tool. A link to the online calculator can be found here http://www.detect-pah.com but Android and Apple apps are available for your phone. This is based on a multicentre multinational screening study (Coghlan et al Ann Rheum Dis 2014 Jul;73(7):1340-9).

Suggested investigations prior to referral:

- Chest radiograph
- Electrocardiograph
- Full lung function tests
- Echocardiography
- Pulse oximetry at rest +/- arterial blood gases
- Renal and liver function tests
- Autoantibody screen
- Consider ventilation/perfusion scan or CT pulmonary angiography

Please include copies of any radiological investigations and reports. Where possible we would prefer advanced imaging to be sent either via an electronic image link or on CD.

How to refer a new patient

Non urgent

Email: papworth.phreferrals@nhs.net

Urgent referral/case discussion

Pulmonary hypertension Consultant on-call - via hospital switchboard (01480 830541)

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