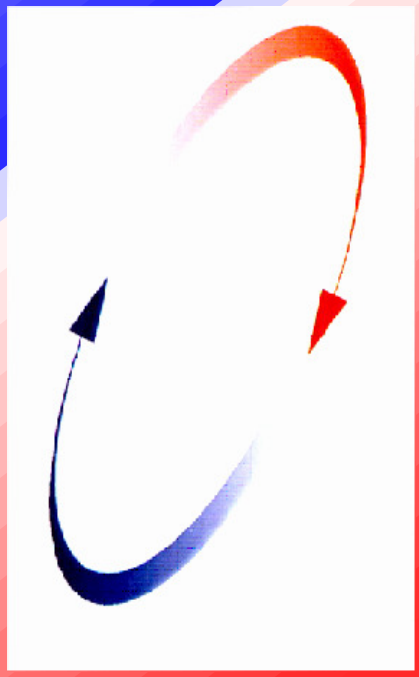
Scottish Pulmonary Vascular Unit

Referral Guidelines



Introduction

The Scottish Pulmonary Vascular Unit was established in 1999 with funding from the National Services Division of Scotland. The unit aims to provide evaluation, investigation and treatment for all patients with pulmonary arterial hypertension (PAH) in Scotland.

Pulmonary arterial hypertension is more common than previously recognised and new treatments have led to an improved prognosis. There are a variety of options for treatment including endothelin receptor antagonists, phosphodiesterase-5 inhibitors and nebulised, subcutaneous and intravenous prostaglandin analogues. Clinical trials of new agents are ongoing.

Specialist supervision of all patients with pulmonary arterial hypertension remains essential. Treatment is increasingly complex and expensive and requires careful ongoing scrutiny.

Whom to Refer

Confirmed / suspected:-

- Pulmonary arterial hypertension
  - Idiopathic
  - Connective tissue disease associated
  - Portopulmonary hypertension
  - Other associated conditions:- HIV, anorexigens, post-correction of congenital heart disease, sickle cell disease.
- Chronic thromboembolic pulmonary hypertension

Consider Referral

Pulmonary hypertension (PH) in:-

- Hypoxic lung disease (COPD, interstitial lung disease, sleep disordered breathing)
- Adult congenital heart disease

Treatment of pulmonary hypertension in these patient groups is not established. This is, however, an evolving field and referral for assessment should be considered if symptoms or echocardiogram pressures seem excessive (e.g. PASP on echocardiogram > 60mmHg) .

Patients with pulmonary hypertension secondary to left heart disease are not currently evaluated.

Suggested Assessment Prior to Referral

- We prefer all patients to have been seen by consultants in Cardiology or Respiratory prior to referral.
- All patients should have an ECG, chest X-ray, spirometry, transthoracic echocardiogram, V-Q scan and CTPA. We prefer to do cardiac catheterisation in Glasgow. Please do not delay referrals for extensive investigation if it is clear that PH is the dominant problem.
- Please include copies of reports (especially the echocardiogram) with the referral.

How to Refer

- We aim to see all patients within a month of referral and to start treatment where appropriate within two months. Please send or fax the referral to the address below.
- Patients can be admitted directly if clinic review is inappropriate. Please indicate the reason for this in the referral letter.
- For unwell patients with known or new PH, SPVU medical staff can be contacted for advice via radiopage or the hospital switchboard 24 hours/day and can arrange emergency transfer for assessment and treatment.

If there is doubt about whether to refer or assess further prior to referral, please contact SPVU medical staff to discuss.

Assessment and Follow-up at SPVU

- After clinic review patients will typically be admitted for full diagnostic assessment including cardiopulmonary exercise testing, cardiac MR and right heart catheter studies. Pulmonary angiography, vasodilator testing, transoesophageal echocardiogram and sleep studies will also be performed where indicated

- If PAH is diagnosed, a decision will be made during the admission regarding disease-targeted treatment.

- Patients will usually be followed up at 3 to 6 monthly intervals, sharing care with the referring consultant.

SPVU staff / contact details

Professor Andrew J Peacock
Director

Dr Martin K Johnson
Consultant Physician

Medical Staff: 24 hour advice Radiopage via switchboard: 0141 951 5000

Sr Agnes Crozier Clinical Nurse Specialist	0141 951 5621
Sr Alison Curran Clinical Nurse Specialist	0141 951 5623
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Screening for pulmonary hypertension

Pulmonary hypertension should be considered (and assessed with echocardiogram) in the differential diagnosis of breathlessness in patients with:-

- Connective tissue disease (especially systemic sclerosis)
- Past history of venous thromboembolism
- Portal hypertension
- HIV
- Hypoxic lung disease (where there is unexplained deterioration or symptoms are excessive)
- Sickle cell disease
- Family history of pulmonary hypertension

Screening is currently recommended for patients at risk for chronic thromboembolic pulmonary hypertension and in patients with connective tissue disease.

Screening for PAH in connective tissue disease

Echocardiogram & pulmonary function tests are recommended annually in patients with limited cutaneous systemic sclerosis or mixed connective tissue disease with U1 RNP antibodies.

Please consider referral if there is:-

- Tricuspid regurgitation (TR) velocity > 2.8 m/s
- TR velocity ≤ 2.8 m/s and the patient is symptomatic with no other explanation
- Strong clinical suspicion of pulmonary hypertension (symptoms / signs / reduced D_{LCO}) but inability to estimate PA pressure on echocardiogram

Screening for PH following PTE

Patients with previous venous thromboembolism who are breathless should have an echocardiogram. Patients who have had a submassive or massive PTE should be assessed by echocardiogram 6 to 12 weeks after the event..

Please consider referral if:-

- Convalescent TR velocity > 2.8 m/s
- Breathlessness on exertion with persistent abnormality on V-Q scan or CTPA despite adequate anticoagulation