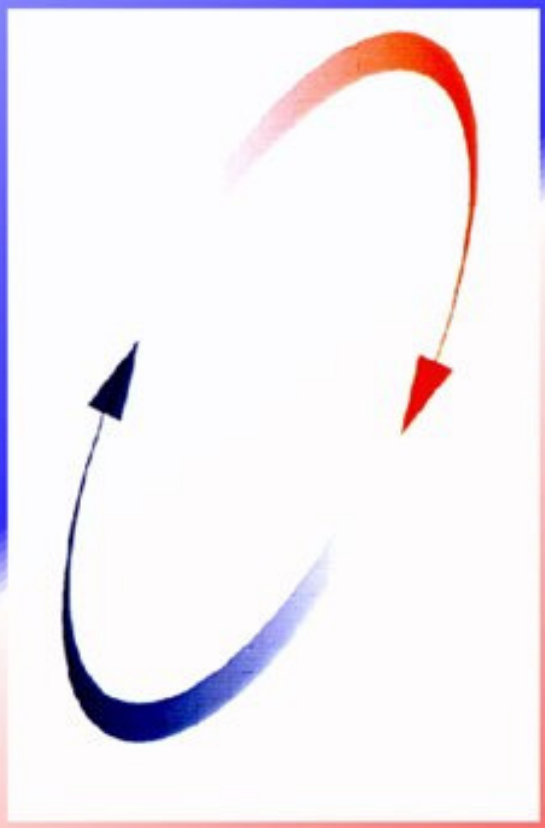
Scottish Pulmonary Vascular Unit

Referral Guidelines



Referral Guidelines

Introduction

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

Pulmonary 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 high dose calcium channel blockers, endothelin antagonists (bosentan, sitaxsentan, ambrisentan), PDE-5 inhibitors (sildenafil, tadalafil) and nebulised, subcutaneous and intravenous prostaglandin analogues. Clinical trials of new agents are ongoing.

Specialist supervision of all patients with pulmonary hypertension remains necessary: treatment is increasingly complex and expensive and requires careful ongoing scrutiny.

Who to Refer

Confirmed / suspected:-

- Idiopathic pulmonary arterial hypertension
- Connective tissue disease associated PAH
- Chronic thromboembolic pulmonary hypertension
- Portopulmonary hypertension
- Other associated PAH: HIV; anorexigens; post-correction right-left shunt; sarcoid; sickle cell disease.

Consider Referral

Pulmonary hypertension in :-

- Hypoxic lung disease (COPD, interstitial lung disease, sleep disordered breathing)
- Cardiac disease (R-L cardiac shunt / Eisenmenger's)

Treatment of PH in these patient groups is not established and evaluation is not usually undertaken. This is, however, an evolving field and referral for assessment should be considered if symptoms or echo pressures seem excessive (PASP >60mmHg) or there are other PH associated conditions.

Patients with pulmonary hypertension secondary to left heart disease are not currently evaluated, unless there are other PH associated conditions.

Referral Guidelines

Suggested Assessment Prior to Referral

- We prefer if all patients have been seen by consultants in Cardiology or Respiratory
- All patients should have an ECG, chest X-ray, spirometry, transthoracic echocardiogram
- Other investigations (V/Q, CT chest) may be indicated to assess for associated disorders etc. These are routinely performed during patients assessment at the SPVU: in particular, we prefer to do cardiac catheterisation here. Please do not delay referrals for extensive investigation if it is clear that PH is the dominant problem
- Please include copies of reports / radiographs with the referral

How to Refer

- Routine referrals are seen at monthly new patient clinic: please send written or faxed referral to SPVU secretaries
- Patients can be admitted directly if clinic review is inappropriate: please indicate on referral letter
- Unwell patients with known or new PH: SPVU medical staff can be contacted for advice 24 hours / day and can arrange transfer for assessment / treatment

If there is any 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 cardiorespiratory work-up comprising:- transthoracic echo; V/Q scan; HRCT chest / CTPA; cardiac MRI; exercise testing; right heart catheter studies. Pulmonary angiography, vasodilator testing, transoesophageal echo and sleep studies will also be performed, where indicated.
- If PAH is diagnosed decision will be made during this admission regarding disease-targeted treatment: typically this will be initiated on a further day-case admission.
- Patients will usually be followed up at 3-6 monthly intervals, sharing care with the referring consultant.

Referral Guidelines

Screening for pulmonary hypertension

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

- Connective tissue disease (esp limited cutaneous 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 & in patients with connective tissue disease

Screening for PH in connective tissue disease

Echocardiography & pulmonary function tests are recommended annually.

Please refer for assessment if:-

- PA systolic pressure (PASP) is >40mmHg
- PASP is 30-40mmHg and patient is symptomatic with no other explanation
- Reduced gas transfer (DIco <50% predicted) with no evidence of lung disease on HRCT
- Strong clinical suspicion of pulmonary hypertension (symptoms & clinical signs) with inability to estimate PA pressure on echo

Screening for PH following PTE

Most patients who have had a submassive or massive PTE should be assessed at 6 months with an echocardiogram. Patients with recurrent events or evidence of persistent clot on imaging studies are at particular risk of developing pulmonary hypertension.

Please refer for assessment if:-

- Convalescent PA systolic pressure (PASP) is >40mmHg
- Persisting symptoms with evidence of persistent clot on V/Q scan &/or CTPA despite adequate anticoagulation

Referral Guidelines

SPVU staff / contact details

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