

Sheffield Teaching Hospitals: Pulmonary Hypertension

Information for Medical Staff 31/03/2014

Common Problems

1. Right heart failure

Patients with significant PH can commonly develop right heart failure. Right (and left) ventricular function and cardiac output can be improved by off-loading the right ventricle with effect diuresis. Patients may often require hospital admission if an increase in oral diuretics is not successful. We aim to off-load 1 litre of fluid (1kg of weight) per day while closely monitoring renal function. Patients often require intravenous frusemide administered bd or via a continuous infusion with K+ sparing diuretics and sometimes the addition of alternate/daily thiazide.

2. Arrhythmias

Patients with PAH are especially prone to developing atrial flutter due to dilatation of their right atrium. The intrinsic atrial rate can often be slower than usual and atrial flutter with 2:1 block may produce a ventricular rate of ~120. An ECG should therefore be obtained in every deteriorated patient and closely examined for the presence of atrial flutter. Management of atrial flutter may consist of digoxin and amiodarone, DC cardioversion or referral for electrophysiology.

3. Hickman line infection

Patients receiving intravenous iloprost via an indwelling line have a risk of developing a line infection of 0.4 - 0.6 / 1000 days. Patients often note increased breathlessness and worsened exercise capacity. Line infections are generally not associated with exit site infections and patients may not give classical symptoms of infection. CRP is a very useful screening tool. If suspected then peripheral and central cultures should be taken and iv antibiotics covering gram-positive and gram-negatives commenced ([see link to Local Guidelines](#)).

4. Iloprost side effects

Iloprost is associated with several side effects: headache, jaw pain, vomiting, diarrhoea, leg pain, flushing. These are especially problematic when patients commence therapy or have recently increased their dose. Side effects generally settle spontaneously but if severe a reduction in dose may be indicated – please contact us to discuss. Renal function should be monitored if diarrhoea and/or vomiting are particularly bad or persists.

5. LFT monitoring

Bosentan is associated with a ~5% risk of elevated transaminases although all endothelial receptor antagonists require LFT monitoring. Monthly LFT monitoring is performed via a system of sending filled blood tubes back to Sheffield in pre-addressed and stamped packaging. If abnormal LFTs (especially AST or ALT >3x upper limit of normal) are detected then please contact us and ask the patient to temporarily stop their endothelin receptor antagonist.

6. Patient deterioration

The most common causes of deterioration in PAH are worsening disease with subsequent right heart failure, arrhythmia and sepsis. Pulmonary embolism also may require exclusion.