

# PULMONARY HYPERTENSION

<b>SETTING</b>	Trustwide
<b>FOR STAFF</b>	Medical and Nursing Staff
<b>PATIENTS</b>	All adult and paediatric patients with Pulmonary Arterial Hypertension.

**These guidelines are for the investigation and management of children and adults with unexplained pulmonary arterial hypertension. This will exclude those with high left atrial pressure (usually due to left heart disease), high pulmonary blood flow due to congenital heart disease and known chronic lung disease.**

**The definition of Pulmonary Arterial Hypertension is**

- **Mean Pulmonary artery pressure >25mmHg**
- **Mean left atrial pressure <15mmHg**
- **Pulmonary vascular resistance > 3 Wood units x BSA (M<sup>2</sup>)**
  
- **Screening can be with echocardiogram and peak TR >2.7m/s (with no outflow tract obstruction)**

## History

- Presenting History
  - Unexplained breathlessness
  - Non-wheezy “asthma”
  - Fainting episodes, syncope or near-miss syncope
  - Downs syndrome with or without upper airway obstruction
- Past History
  - Preterm delivery
  - Cyanosis at birth
  - Chronic lung disease or oxygen dependency
  - Drug history – any illegal or slimming drugs or St Johns’ Wort
- Family History
  - Sudden unexpected death in relatives at less than 40 years

## Examination

- Full respiratory examination
- Cardiac examination including blood pressure
  - Apex position
  - Left parasternal heave (right ventricular hypertrophy)
  - Loud P2
  - Hepatomegaly
- Oxygen saturations, right hand and feet in infants (feet within 3% of hand sats)
- Peak Flow

Investigations		
Blood tests	FBC + Film	If necessary
	Procoagulation screen	'Pulmonary Hypertension' label
	Liver FT	
	Virology screen	Hepatitis and HIV
	Autoimmune profile	ANCAs, complements
	DNA storage	(Consent required)
Radiography	Chest X-Ray	
	CT scan	For interstitial lung disease
Pulmonary Function Tests	FVC, FEV1, PEFr	
	V/Q scan	
Cardiology	Echocardiography	Full PH protocol
	Right Heart Catheter	Pulmonary vascular resistance study

### Outcome

If a child or adult is thought to have Pulmonary Hypertension, then please discuss the case with Professor Tulloh (contact below). The investigations above are to help identify the cause. Medication for appropriate cases is only available through the National Pulmonary Hypertension Service at the PH clinics.

### RELATED DOCUMENTS

European society of Cardiology Guidelines  
<http://www.escardio.org/guidelines-surveys/esc-guidelines/GuidelinesDocuments/guidelines-PH-FT.pdf>

### SAFETY

**Do not give large doses of beta blockers for CT or MRI scan to patients with Pulmonary Hypertension**

### QUERIES

Contact Professor Rob Tulloh (Ext 28176 (sec 28856) / Mobile via switch) – or the on call Adult Congenital Heart Disease team (adults) or Paediatric Cardiologist (bleep 2424, children).