

Diagnostic tests

(Determining cause of PH)

| Test | PAH less likely than other causes |
|----------|-------------------------------------------------------------------------------------------------------|
| ECG | AF; LBBB; LAD; Q waves |
| PFT | FEV1 <60% predicted FVC < 70% predicted |
| Echo | LAA > 18cm ² ; LVEF < 50% LVH; EE' > 15; significant left sided valve disease |
| Blood | Scl70+; eGFR < 30ml/min |
| HRCT | Fibrosis > 20%; emphysema; |
| V/Q scan | Any perfusion defect – possible CTED (CTPA; Pulm Angio or MR angio) |
| Catheter | End expiratory wedge/LVEDP (off diurectics) > 15mmHg; Post fluid (500ml/5min) LVEDP/wedge > 18mmHg |

Any significant left heart or lung disease makes PAH a less likely diagnosis. AF – atrial fibrillation; LAD – left axis deviation; LAA – left atrial area; LVEF – left ventricular ejection fraction; CTED – chronic thromboembolic disease; LVEDP – left ventricular end diastolic pressure.

Treatment & Follow up

Treat according to ESC/ERS guidelines. Reassessment at least 3mo. after treatment change and annually

Vasodilator testing not useful in SSc avoid CCB

Anticoagulation not evidence based in SSc, avoid if any bleeding risk (e.g. iron deficiency; GAVE; angiodysplasia)

Encourage exercise within symptomatic limits

| Stable & satisfactory I/II if PAH limited | Parameter | Unstable & unsatisfactory III/IV if due to PAH |
|------------------------------------------------|-----------------|------------------------------------------------------------|
| ≤ 300m | FC | > 400m |
| Normal | 6MWD | > 2 x N |
| RAa falling; TAPSE > 2cm; No PC effusion | NTproBNP | RAa > 28cm ² ; TAPSE < 1.6cm; PC effusion |
| RA ≤ 8mmHg, CI > 2.7l/min/m ² | Echo | RA > 11mmHg, CI < 2l/min/m ² |
| No fall in O2 sats, KCO stable | Catheter | 5% fall in O2 sats off oxygen, 10% fall in KCO |
| | PVOD indicators | |

Modifications of ESC algorithm, FC 2 not always achievable, 415m = average 6MWD in SSc without PAH; normal RAa now known; higher CI required in SSc, PVOD more common in SSc.