

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.