

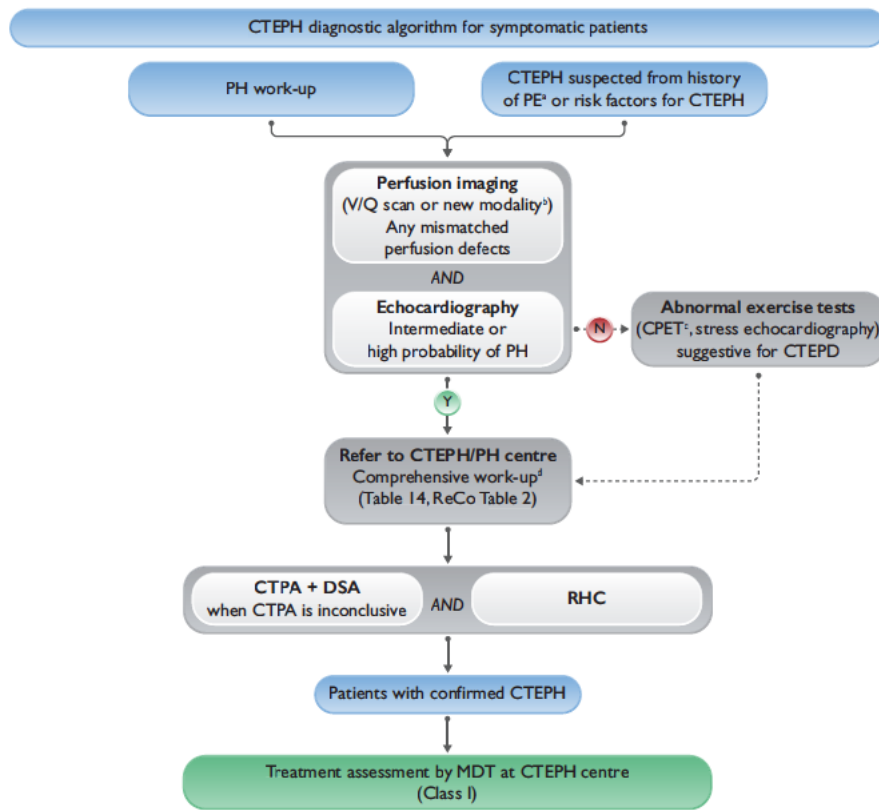


Figure 1. ESC 2022 GL on pulmonary hypertension. Diagnostic strategy in chronic thrombo-embolic pulmonary hypertension.

CPET, cardiopulmonary exercise test; CTEPD, chronic thromboembolic pulmonary disease; CTEPH, chronic thrombo-embolic pulmonary hypertension; CTPA, computed tomography pulmonary angiography; DECT, dual-energy computed tomography; DSA, digital subtraction angiography; MDT, multidisciplinary team; MRI, magnetic resonance imaging; N, no; PE, pulmonary embolism; PETCO₂, end-tidal partial pressure of carbon dioxide; PH, pulmonary hypertension; ReCo, recommendation; RHC, right heart catheterization; sPAP, systolic pulmonary arterial pressure; V/Q, ventilation/perfusion; VE/VCO₂, ventilatory equivalents for carbon dioxide; VO₂/HR, oxygen pulse; VO₂, oxygen uptake; Y, yes.

a CTEPH suspected from history of PE, including elevated sPAP on echocardiography and signs suggesting CTEPH on CTPA performed at the time of the acute PE.

b Alternative perfusion imaging techniques—such as iodine subtraction mapping, DECT, and MRI perfusion—are currently under evaluation.

c Typical pattern, including low PETCO₂, high VE/VCO₂, low VO₂/HR, and low peak VO₂.

d Comprehensive work-up after 3 months of therapeutic anticoagulation or sooner in unstable or rapidly deteriorating patients. Ideally, CTPA, DSA, and RHC are performed in CTEPH centres, but they are sometimes performed in PH centres, depending on the country and organization.

Table 1. ESC 2022 GL on Pulmonary Hypertension. Recommendations for chronic thrombo-embolic pulmonary hypertension and chronic thrombo-embolic pulmonary disease without pulmonary hypertension

CTEPH

Lifelong, therapeutic doses of anticoagulation in all patients with CTEPH	I C
Antiphospholipid syndrome testing in patients with CTEPH	I C
In patients with CTEPH and antiphospholipid syndrome, anticoagulation with VKAs	I C
All patients with CTEPH to be reviewed by a CTEPH team for the assessment of multimodality management	I C
PEA as the treatment of choice for patients with CTEPH and fibrotic obstructions within pulmonary arteries accessible by surgery	I B
BPA in patients who are technically inoperable or have residual PH after PEA and distal obstructions amenable to BPA	I B
Riociguat for symptomatic patients with inoperable CTEPH or persistent/recurrent PH after PEA	I B
Long-term follow-up after PEA and BPA, as well as for patients with CTEPH established on medical therapy	I C
A multimodality approach for patients with persistent PH after PEA and for patients with inoperable CTEPH	IIa C
Treprostinil s.c. in patients in WHO-FC III–IV who have inoperable CTEPH or persistent/recurrent PH after PEA	IIb B
Off-label use of drugs approved for PAH in symptomatic patients who have inoperable CTEPH	IIb B
In patients with inoperable CTEPH, a combination of sGC stimulator/PDE5i, ERA, or parenteral prostacyclin analogues	IIb C
BPA for technically operable patients with a high proportion of distal disease and an unfavourable risk:benefit ratio for PEA	IIb C

CTEPD without PH

In patients with CTEPD without PH, long-term anticoagulant therapy on an individual basis ^a	IIa C
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PEA or BPA in selected symptomatic patients with CTEPD without PH	IIa C
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GRADE

<i>Quality of evidence</i>	<i>Strength of recommendation</i>
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In patients with CTEPH who are candidates for BPA, medical therapy should be considered prior to the intervention	Very low	Conditional	IIa B
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BPA, balloon pulmonary angioplasty; CTEPD, chronic thrombo-embolic pulmonary disease; CTEPH, chronic thrombo-embolic pulmonary hypertension; ERA, endothelin receptor antagonist; PAH, pulmonary arterial hypertension; PDE5i, phosphodiesterase 5 inhibitor; PEA, pulmonary endarterectomy; PE, pulmonary embolism; PH, pulmonary hypertension; s.c., subcutaneous; sGC, soluble guanylate cyclase; VKA, vitamin K antagonist; WHO-FC, World Health Organization functional class.

a Long-term anticoagulant therapy is recommended when the risk of PE recurrence is intermediate or high, or when there is no history of venous thrombo-embolism.

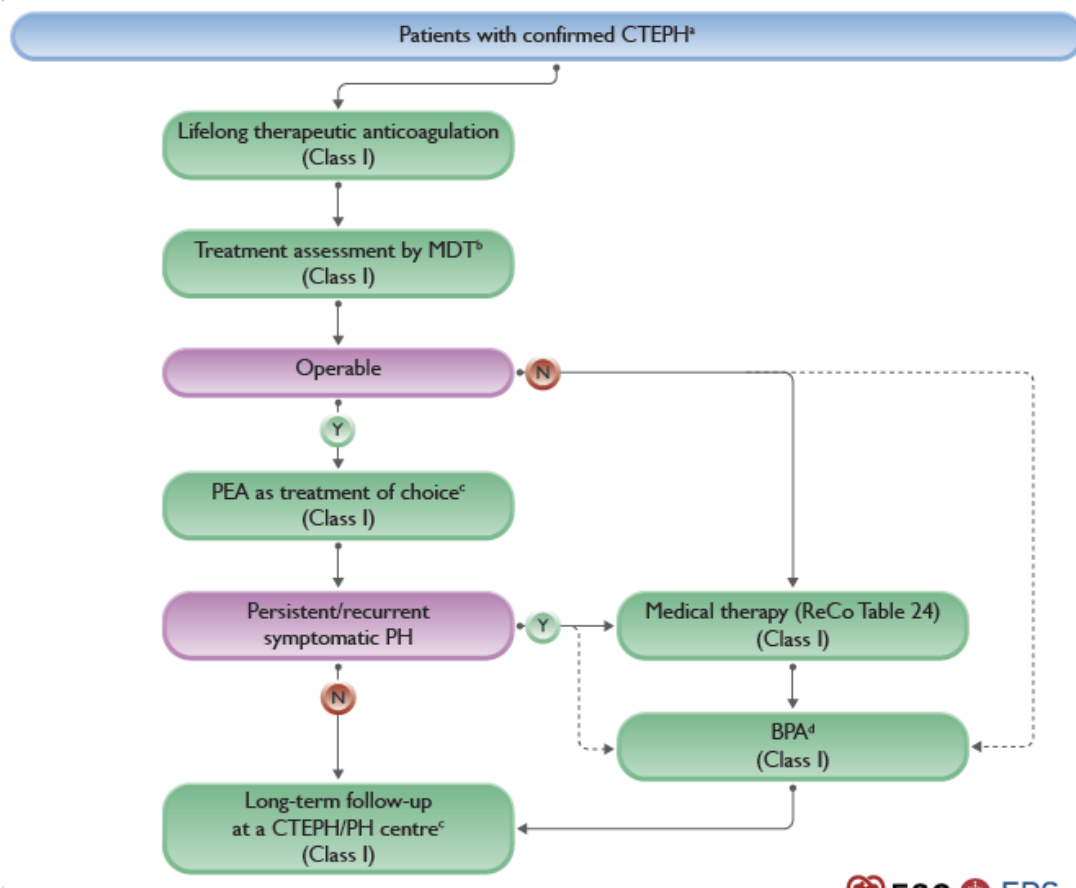


Figure 2. ESC 2022 GL on pulmonary hypertension. Management strategy in chronic thrombo-embolic pulmonary hypertension.

BPA, balloon pulmonary angioplasty; CTEPD, chronic thrombo-embolic pulmonary disease; CTEPH, chronic thrombo-embolic pulmonary hypertension; MDT, multidisciplinary team; N, no; PAH, pulmonary arterial hypertension; PEA, pulmonary endarterectomy; PH, pulmonary hypertension; PVR, pulmonary vascular resistance; ReCo, recommendation; WU, Wood units; Y, yes.

a Selected symptomatic patients with CTEPD without PH can also be treated by PEA and BPA.

b MDT meeting can be virtual.

c Treatment assessment may differ, depending on the level of expertise in PEA and BPA.

d For inoperable patients with PVR ≥ 4 WU, medical therapy should be considered prior to BPA; there are limited data on BPA as first-line therapy.

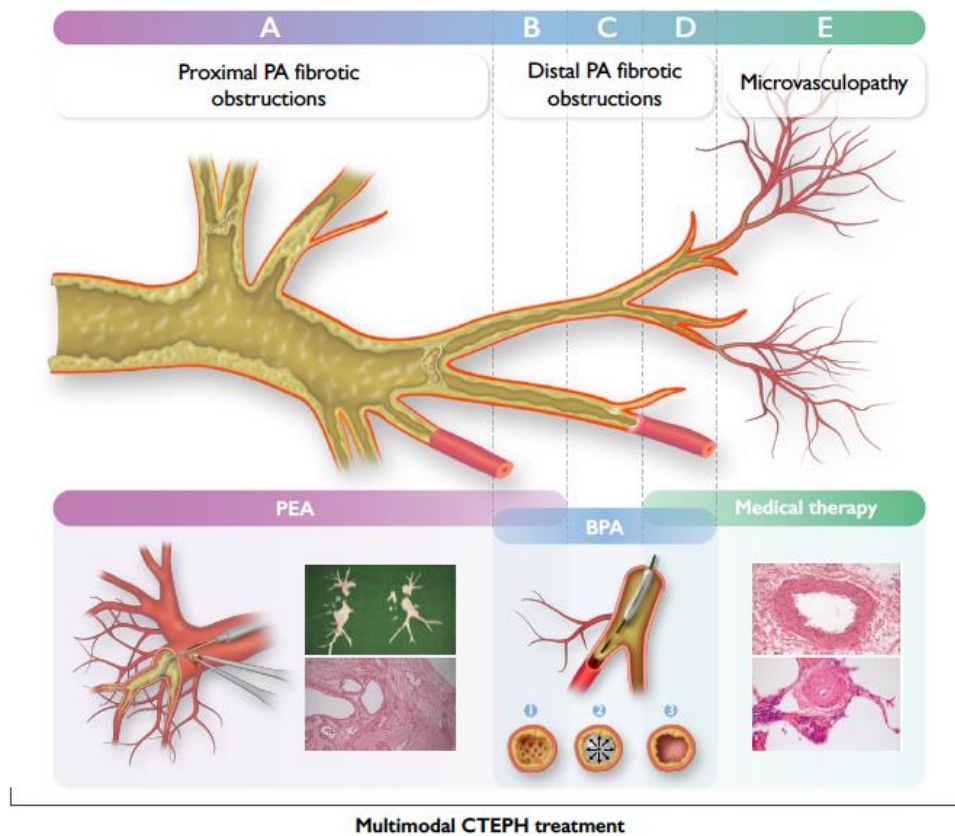


Figure 3. ESC 2022 GL on pulmonary hypertension. Overlap in treatments/multimodality approaches in chronic thrombo-embolic pulmonary hypertension.

BPA, balloon pulmonary angioplasty; CTEPH, chronic thrombo-embolic pulmonary hypertension; PA, pulmonary artery; PEA, pulmonary endarterectomy. Top panels: (A) Proximal PA fibrotic obstructions (vessel diameter 10–40 mm). (B) Distal segmental and subsegmental PA fibrotic obstruction potentially suitable for both PEA and BPA interventions (vessel diameter 2–10 mm). (C) Distal subsegmental PA fibrotic obstructions form a web-lesion in a subsegmental branch of the PA suitable for BPA interventions (vessel diameter 0.5–5 mm). (D) Distal subsegmental PA fibrotic obstructions form web-like lesions, which might be accompanied by microvasculopathy (vessel diameter ,0.5 mm). (E) Microvasculopathy (vessel diameter ,0.05 mm) treated with medical therapy. Bottom panels: (A) bottom left: PEA; vessel diameter (0.2–3 cm). The right PA is opened and the suction dissector is introduced between the artery wall and fibrosis. Following the inside of the artery down to segmental and subsegmental levels, the fibrotic material is subsequently freed from the wall and removed with forceps. (A) bottom right: PEA specimen with ‘tails’ to subsegmental branches of the PA; cross-section of partially organized and permeabilized thrombotic lesion of the large PA dissected during PEA. (B, C, D) The wire is introduced between the fibrotic material (1), then the balloon is inflated, leading to a rupture of the web (2). Fibrotic material is connected to the vessel wall (3). (E) Small muscular PA displaying eccentric intimal fibrosis involving intimal thickening and proliferation—target for medical therapies.