CTEPH WEBINAR

MARION DELCROIX
UZ Leuven, Belgium

CTEPH – THE DISEASE

PH CLASSIFICATION

Group	Description
Group 1	Pulmonary arterial hypertension (PAH)
Group 2	Pulmonary hypertension (PH) due to left heart disease
Group 3	PH due to lung diseases and/or hypoxia
Group 4	Chronic thromboembolic PH (CTEPH) and …
Group 5	Unclear or multifactorial mechanisms

Humbert et al, ERJ and EHJ 2022

CTEPH DEFINITION

mPAP	>20 mmHg
V/Q scan	Abnormal with any mismatched perfusion defect
CT scan and/or pulmonary angiography	Abnormal with typical findings of CTEPH
Anticoagulation	At least 3 months

mPAP, mean pulmonary artery pressure.
Humbert et al, ERJ and EHJ 2022.

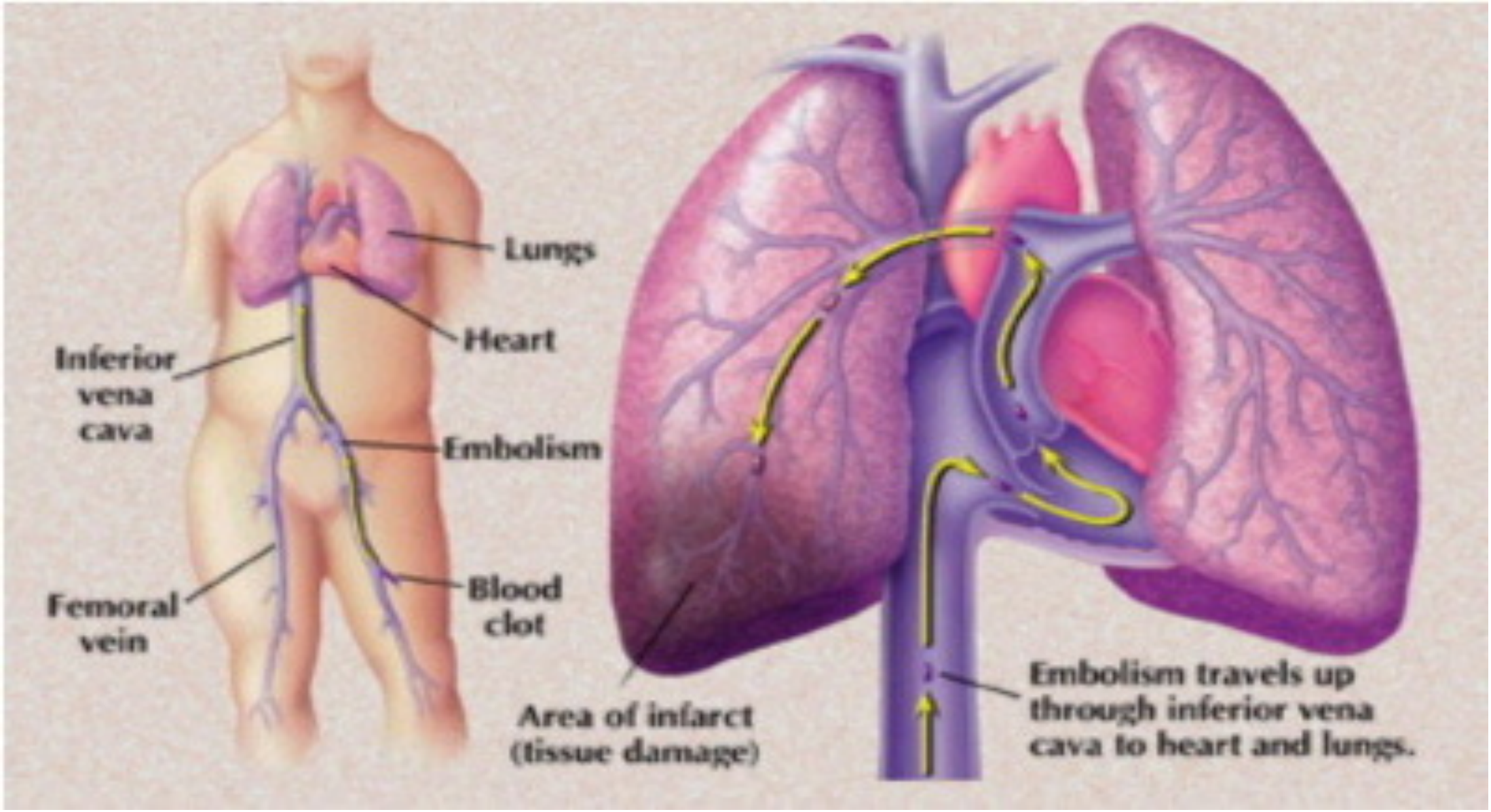
Chronic thrombo-embolic pulmonary disease (CTEPD) / Chronic thrombo-embolic pulmonary hypertension (CTEPH)

Chronic thromboembolic pulmonary disease (CTEPD) is the overarching term to characterize symptomatic patients who present with mismatched perfusion defects on V/Q lung scan and specific signs of chronic clots on CTPA or DSA, whether or not they have PH at rest.

For patients with PH a rest, the historical term CTEPH is still of use.

2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension
(*European Heart Journal*; 2022 – doi: 10.1093/eurheartj/ehac237 and *European Respiratory Journal*; 2022 – doi: 10.1183/13993003.00879-2022)

HISTORY OF PE IN 75% OF THE PATIENTS (MASSIVE OR REPEATED)



CTEPH IS DIFFERENT FROM PULMONARY EMBOLISM

Pulmonary Embolism

Embolectomy

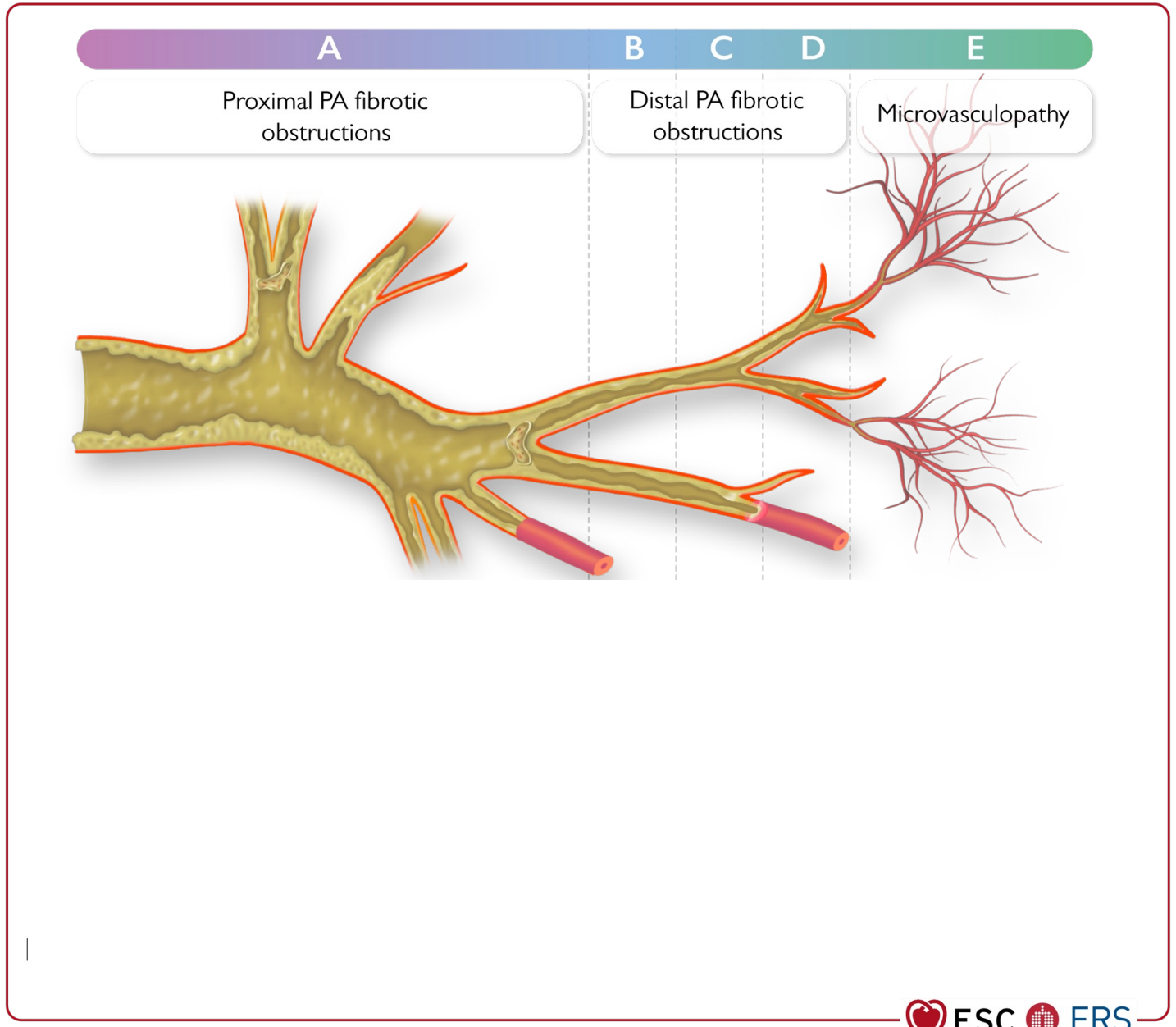
?

CTEPH

Pulmonary endarterectomy

Images, speaker's own.

Chronic thrombo-embolic pulmonary hypertension



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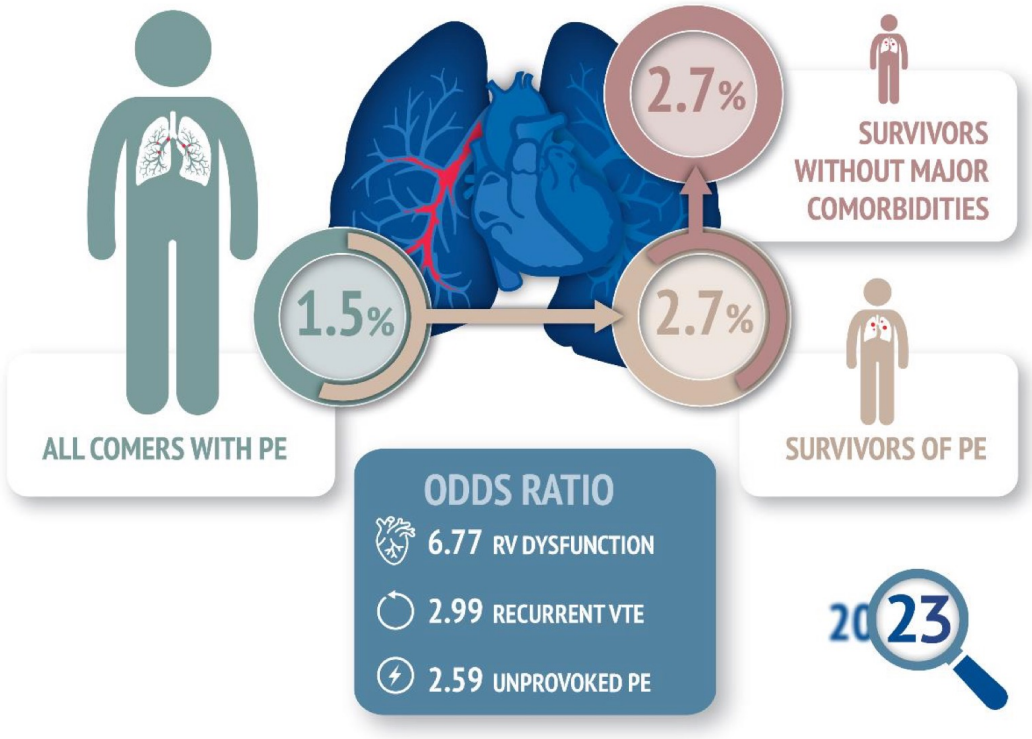
CTEPH AFTER ACUTE PE (updated meta-analysis)



Based on data from 30 studies in 10,249 PE patients

Luijten et al, *Eur Respir J* 2023; 62, 2300449

CTEPH MISDIAGNOSED AS PULMONARY EMBOLISM

- **Signs of PH already present in the majority of patients with PE later diagnosed with CTEPH (57%)**
  - Echocardiography: High PAP and dilated right heart
  - CT: Dilated right heart and signs of chronic vessel obstruction

Guerin L *et al. Thromb Haemost* 2014;112:598–605.

CTEPH IS RARE BUT UNDERDIAGNOSED

Observed incidence
of CTEPH

5-6 per million inh/year

Based on:

Registry data[4-6]

IPAH

2 per million inh/year

Estimated incidence
of new CTEPH

17 per million inh/year

Based on:

- CTEPH incidence post-PE = 3%[1]
- PE incidence = 1 per 1000 inh/year
  - 57% already have CTEPH[2], 43% not
  - 75% have a history of PE[3], 25% not
- 3% * 43% * 1000 * 100/75 = 17 per million

Delcroix M *et al. Ann Am Thorac Soc* 2016;13:S201–6. 1. Ende-Verhaar YM *et al. Eur Respir J* 2017;49:1601792. 2. Guerin L *et al. Thromb Haemost* 2014;112:598–605. 3. Pepke-Zaba et al, Circulation 2011. 4. Kramm *et al. Clin Res Cardiol* 2018. 5. UK National Audit of Pulmonary Hypertension 2012. 6. CTEPH registry, unpublished data. 7. Durrington et al, 10.1136/thorax-2022-BTSabstracts.99

CTEPH IS RARE BUT UNDERDIAGNOSED

Observed incidence
of CTEPH

5-6 per million inh/year

Based on:

Registry data[4-6]

Estimated incidence
of new CTEPH

17 per million inh/year

12.2/million/year in the Sheffield population[7]

Based on:

- CTEPH incidence post-PE = 3%[1]
- PE incidence = 1 per 1000 inh/year
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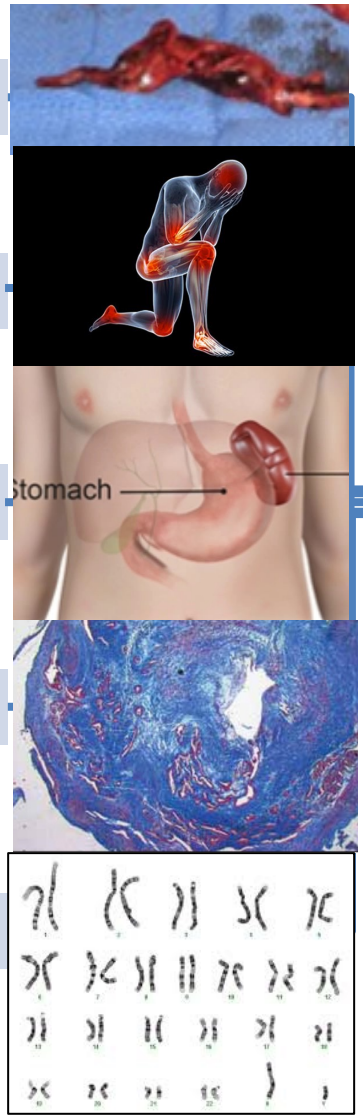
IPAH

2 per million inh/year

Delcroix M *et al. Ann Am Thorac Soc* 2016;13:S201–6. 1. Ende-Verhaar YM *et al. Eur Respir J* 2017;49:1601792. 2. Guerin L *et al. Thromb Haemost* 2014;112:598–605. 3. Pepke-Zaba et al, Circulation 2011. 4. Kramm *et al. Clin Res Cardiol* 2018. 5. UK National Audit of Pulmonary Hypertension 2012. 6. CTEPH registry, unpublished data. 7. Durrington et al, 10.1136/thorax-2022-BTSabstracts.99

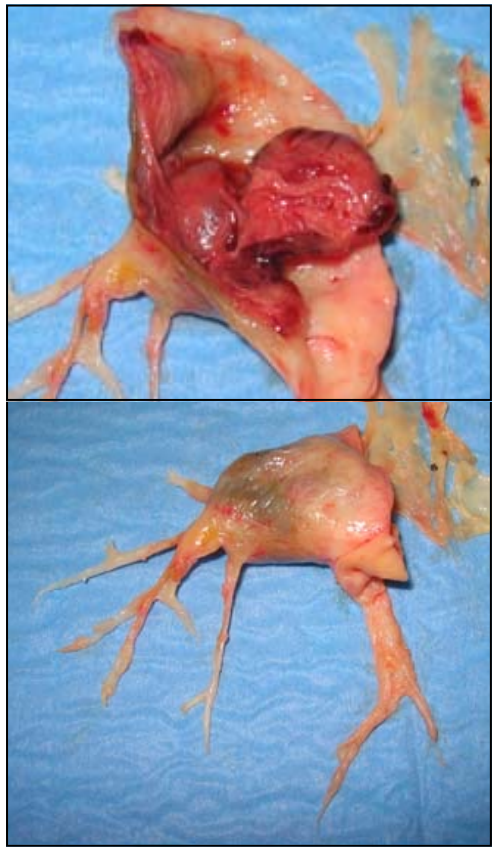
MECHANISMS OF UNRESOLVING PE

- Abnormal clot formation and dissolution
- Inflammation and Infection
- Removal of the spleen
- Deficient recanalisation by new vessels
- Genetic anomalies



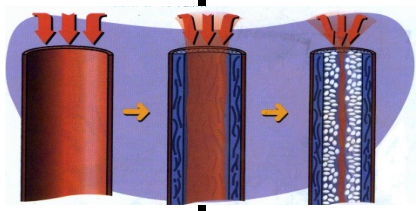
Unresolving pulmonary embolism

CTEPH MECHANISMS (distal)

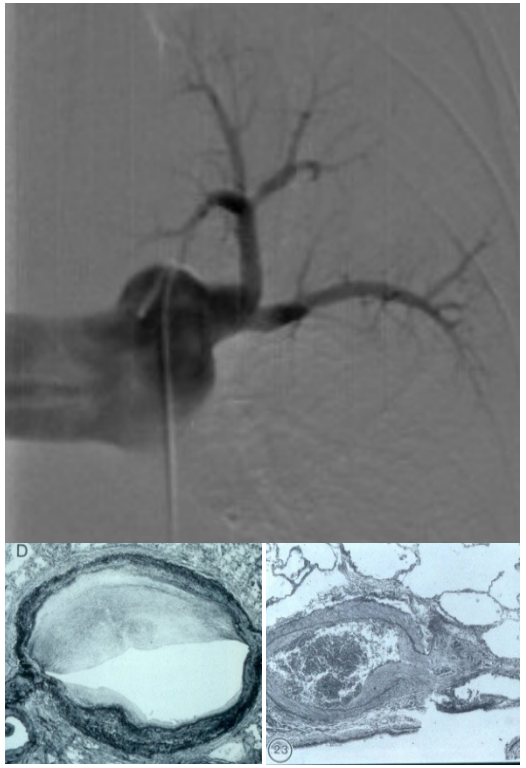


Unresolving pulmonary embolism (massive or repeated)

Exposure to high pressures/flow and vasoactive factors



Small vessel arteriopathy



Moser and Bloor, Chest 1993;103:685

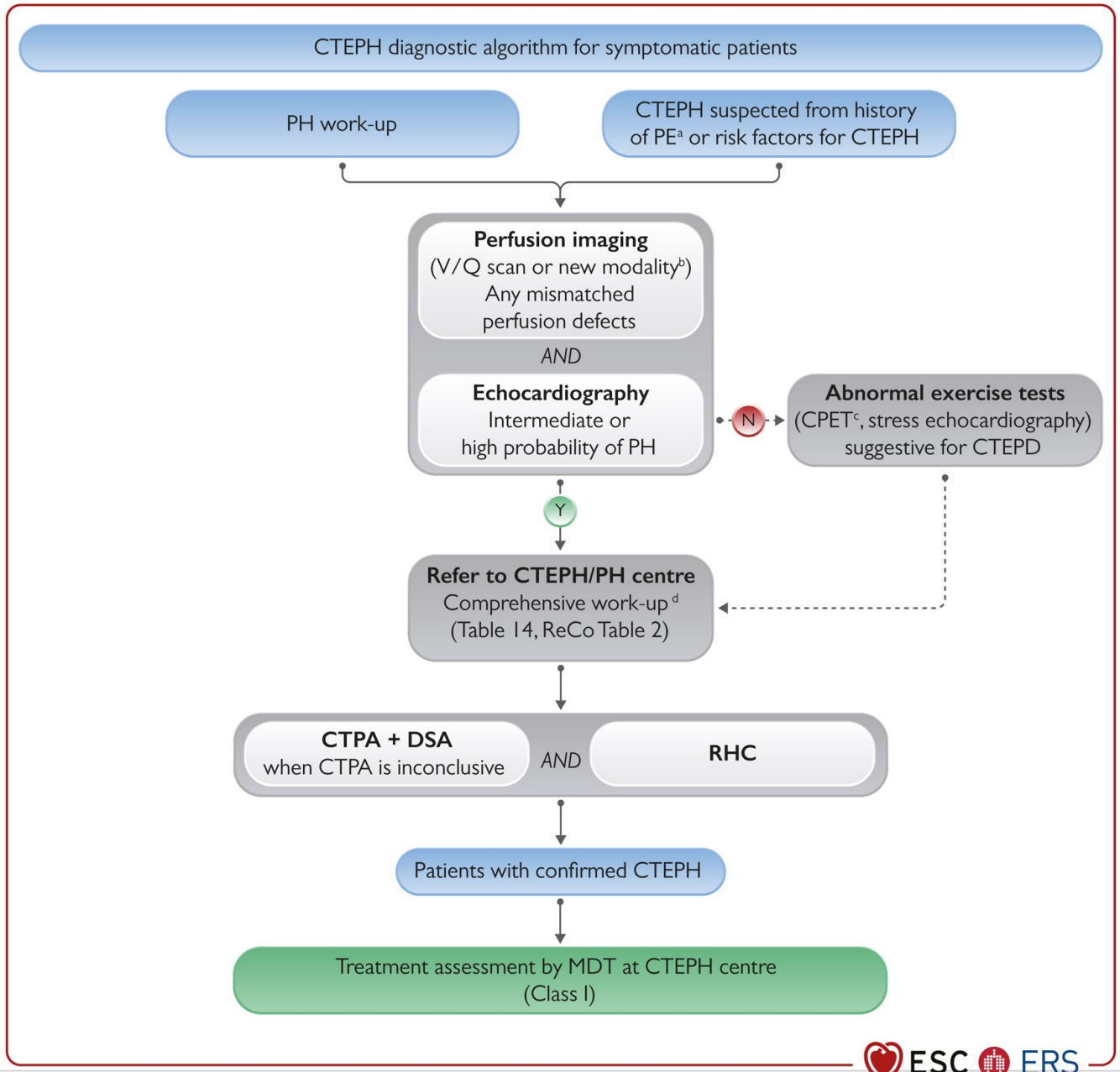
CTEPH – THE DIAGNOSIS

CTEPH SYMPTOMS AT DIAGNOSIS

Symptoms	CTEPH[1]
Number of patients	679
Symptoms to diagnosis (mo, median)	14.1
Dyspnea, %	**99.1**
Edema, %	**40.5**
Fatigue, %	**31.5**
Chest pain, %	15.3
Syncope, %	13.7
Dizziness, %	5.6
Hemoptysis, %	**4.4**

1. Pepke-Zaba J *et al. Circulation* 2011;124:1973–81.

Diagnostic strategy in chronic thrombo-embolic pulmonary hypertension



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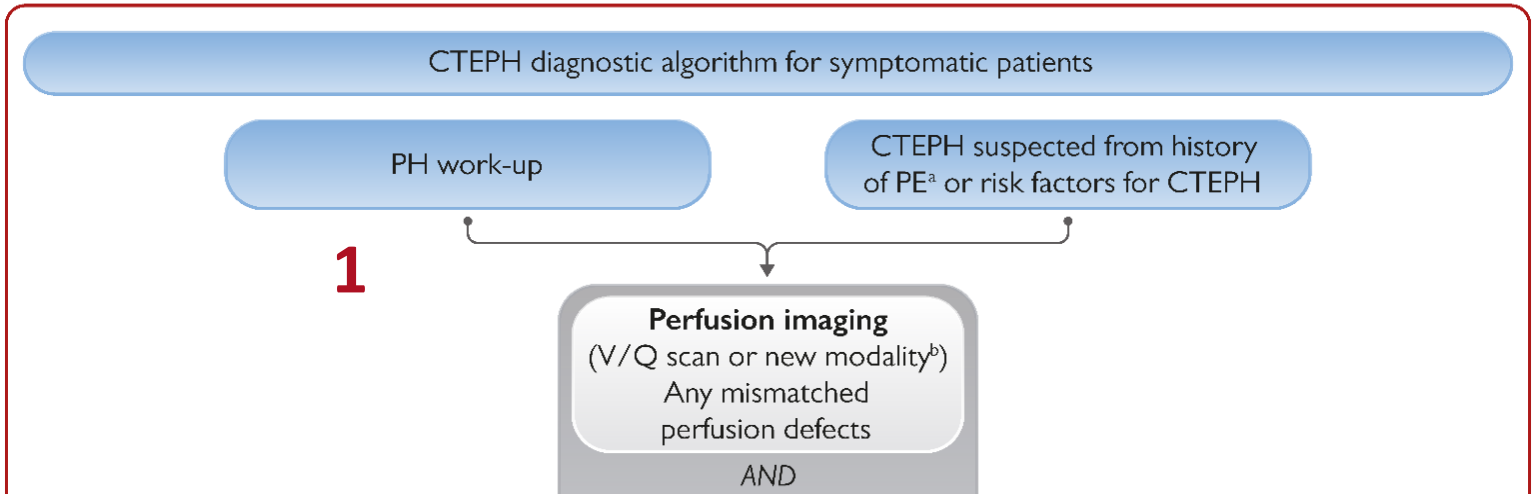
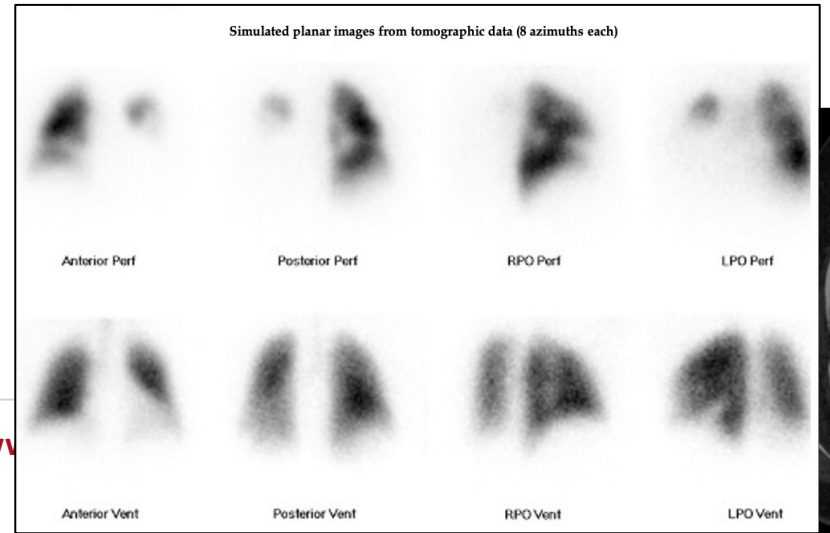
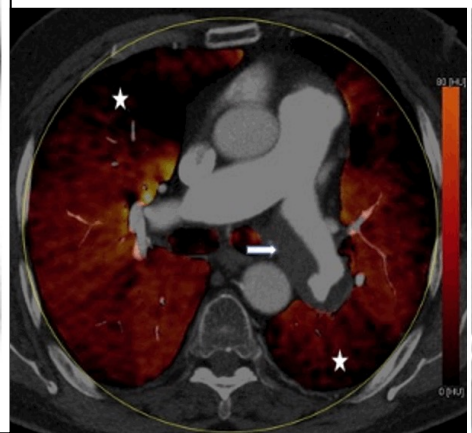
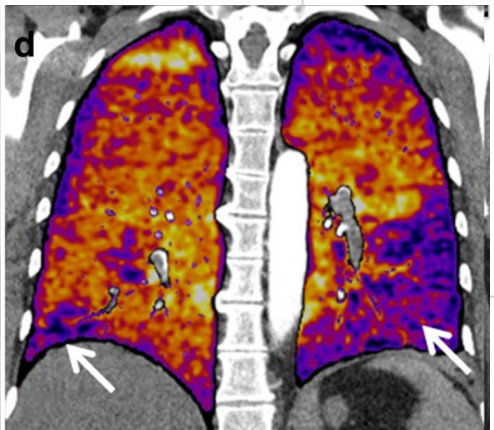
Diagnostic strategy in CTEPH



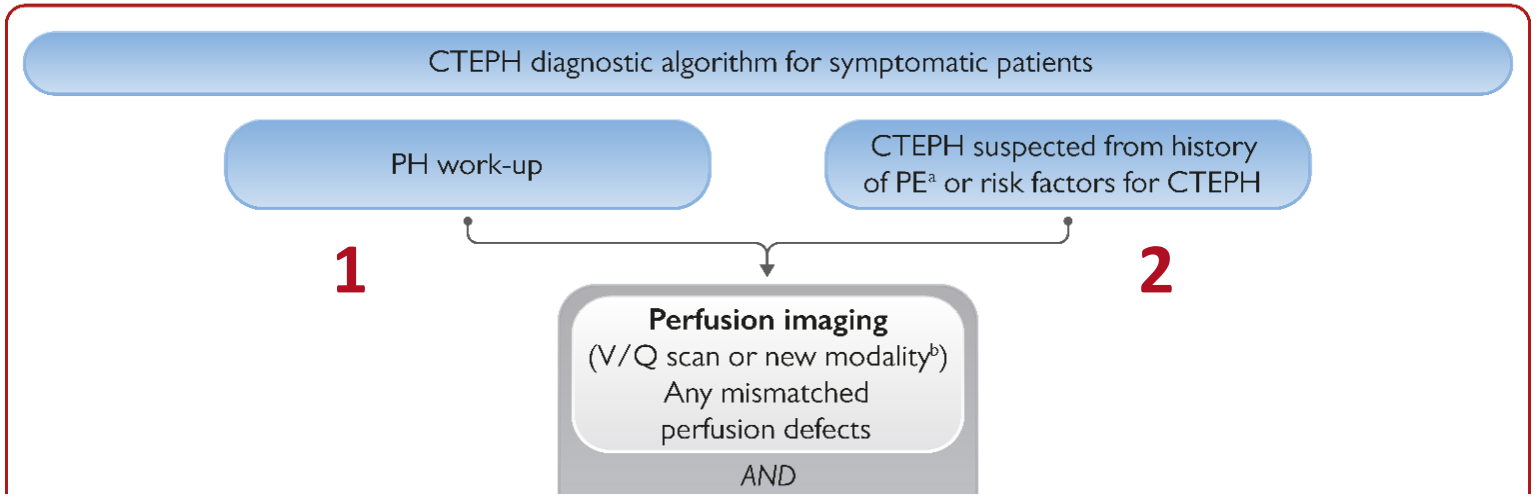
Recommendations	Class	Level
Imaging		
Ventilation/perfusion or perfusion lung scan is recommended in patients with unexplained PH to assess for CTEPH	I	C







Diagnostic strategy in CTEPH



Recommendations	Class	Level
CTEPH/CTEPD		
In patients with persistent or new-onset dyspnoea or exercise limitation following PE, further diagnostic evaluation to assess for CTEPH/CTEPD is recommended	I	C
For symptomatic patients with mismatched perfusion lung defects beyond 3 months of anticoagulation for acute PE, referral to a PH/CTEPH centre is recommended after considering the results of echocardiography, BNP/NT-proBNP, and/or CPET	I	C

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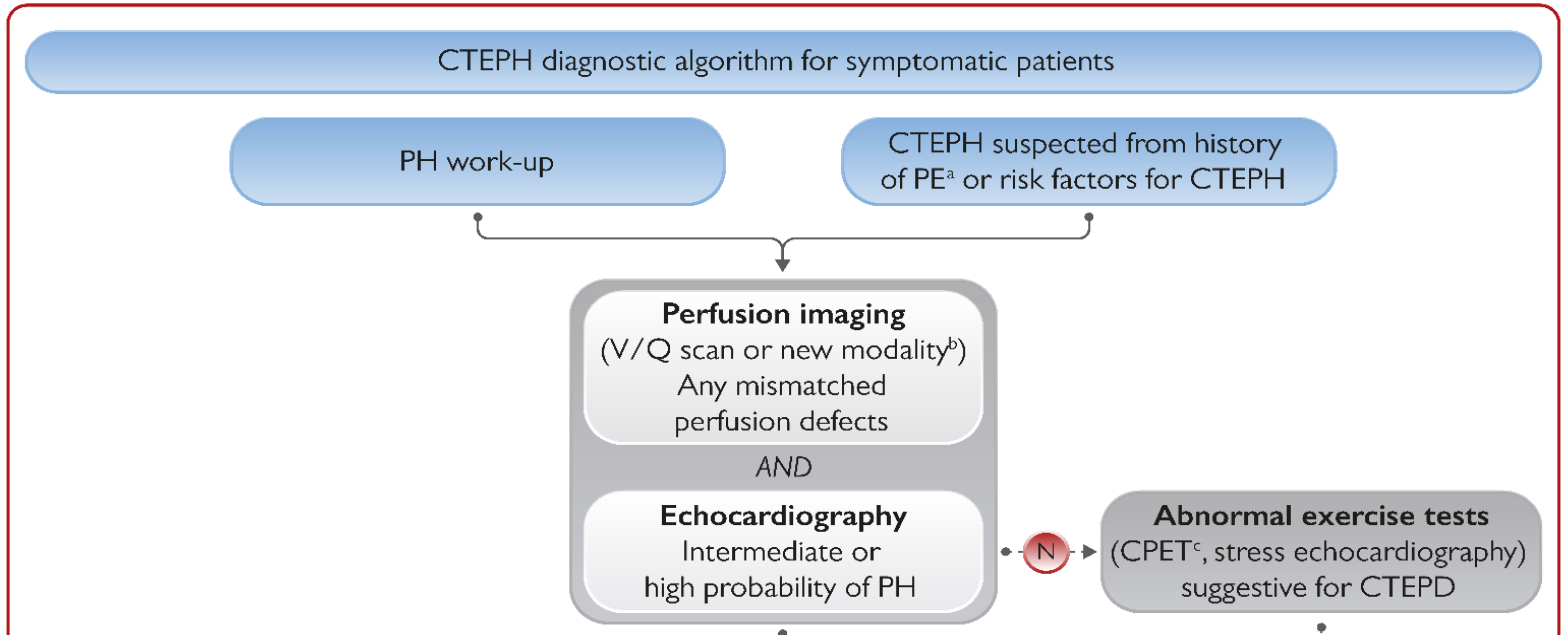

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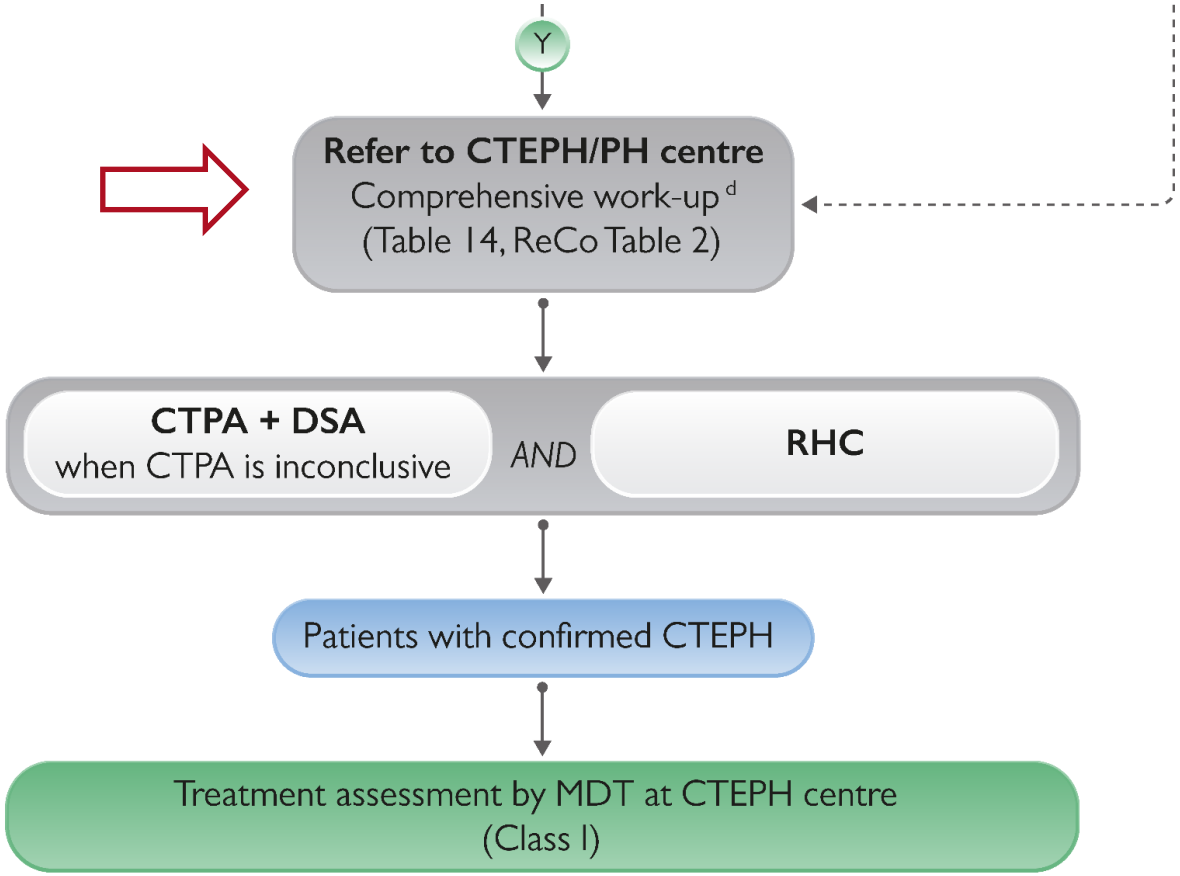

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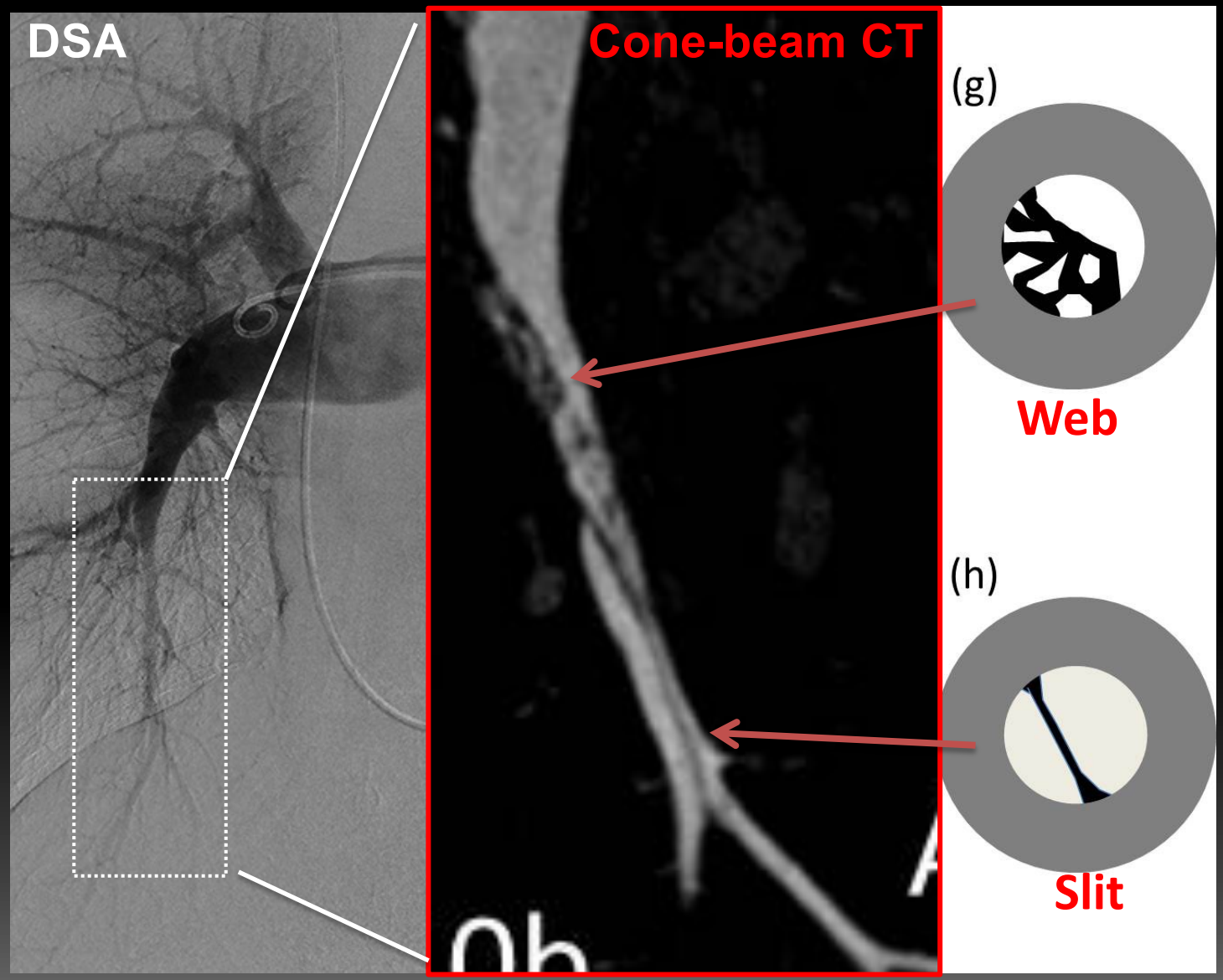
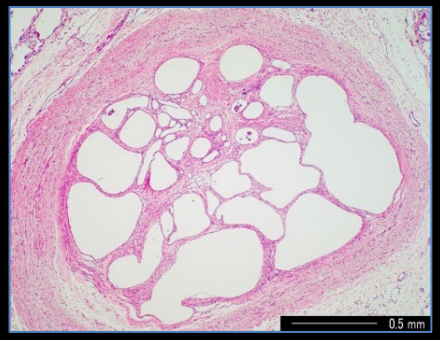
A TYPICAL CASE – CT SCAN

A TYPICAL CASE – PULMONARY ANGIOGRAPHY

Cone beam-CT for the detection of distal lesions (web & slit)





From Dr. Ueda, Dr. Ogo-Ohta
Department of Pathology,NCVC

D. Korn et al., Am J Pathol 1962, 40, 129-151.

Sugiyama M, Fukuda T, Ogo T et al. 2014 Jul;32(7):375-82 Jpn J Radiol

CTEPH: THE TREATMENT

CTEPH TREATMENT EVOLUTION

15 years ago

- Surgery

CTEPH TREATMENT EVOLUTION

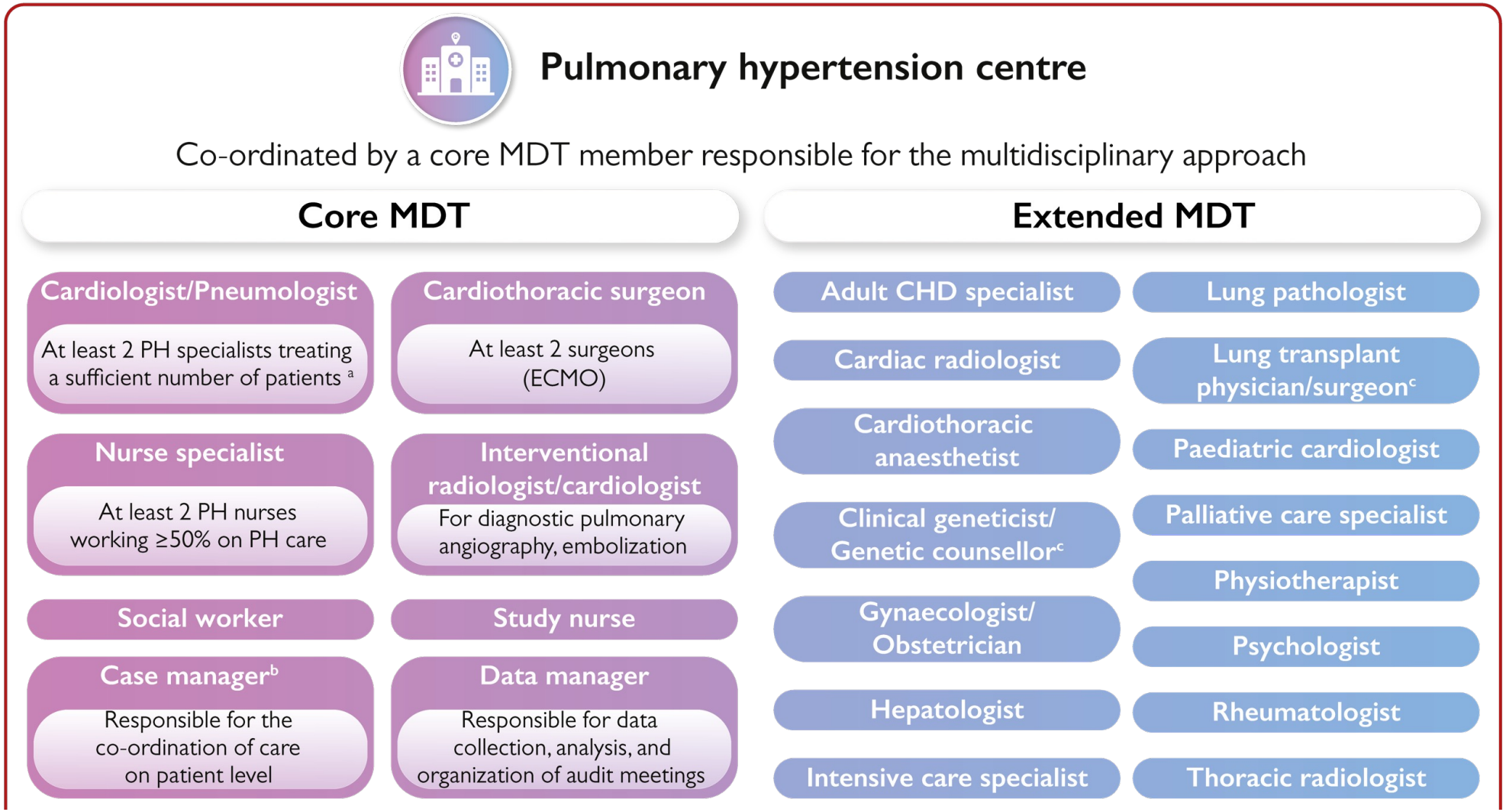
15 years ago

- Surgery

Today

- Surgery (PEA)
- Balloon dilatation (BPA, 2012)
- Medical therapy (2013)
- PEA+BPA same time
- PEA+BPA later
- Medical followed by BPA
- Medical followed by PEA

Pulmonary hypertension centre



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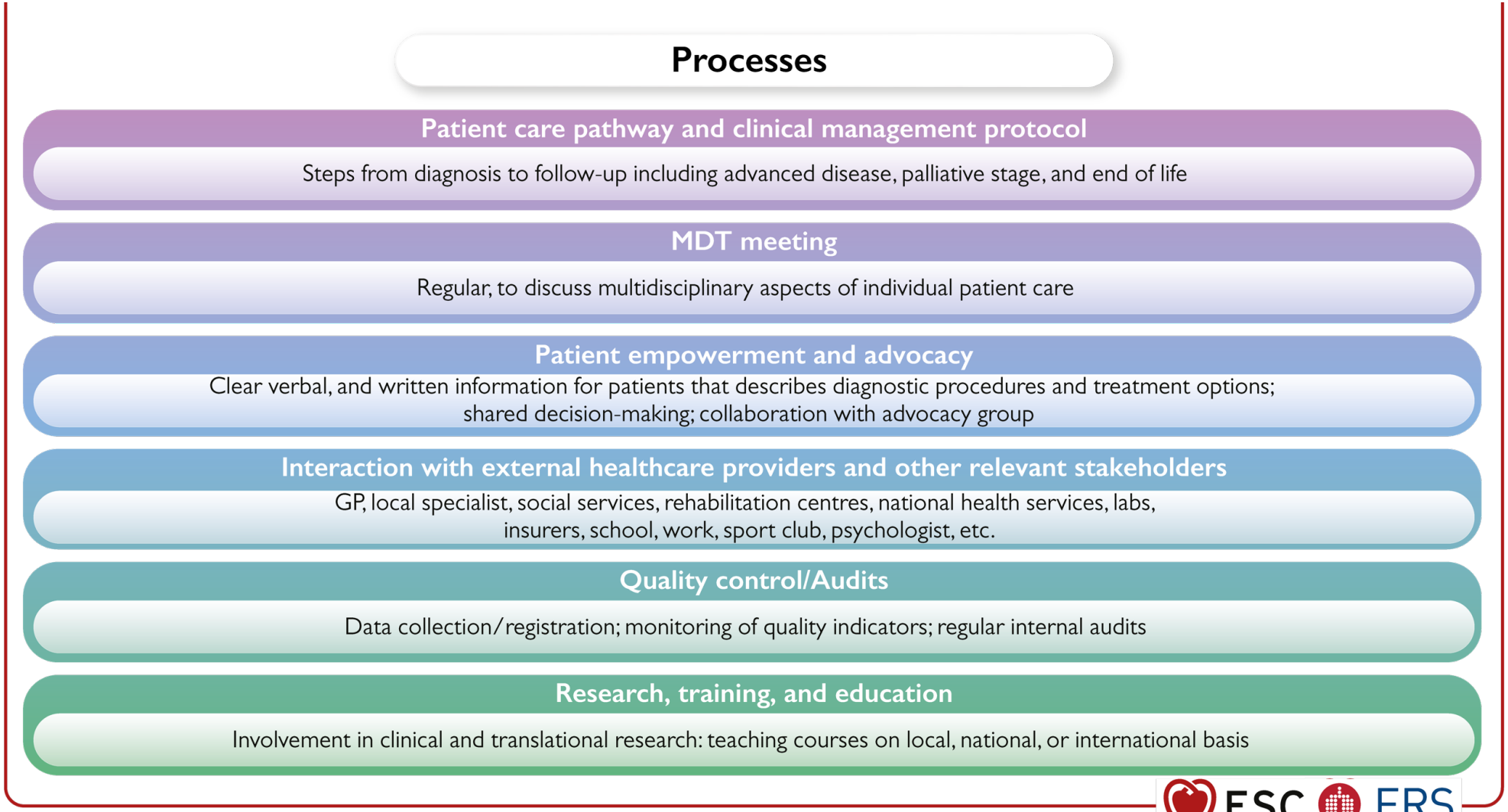
Pulmonary hypertension centre



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Pulmonary hypertension centre

- PH centres should provide care by a team of experts from different areas of medicine – including heart and lung professionals along with nurses, radiologists and social workers.
- Centres should have plans in place to make quick referrals to other services, such as transplant units or genetic counselling services. They should also maintain a patient registry to help with research. It is also recommended that they work closely with patient organisations.

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CTEPH centres

Chronic thrombo-embolic pulmonary hypertension team and experience criteria

- **To optimize patients' outcomes, CTEPH centres should fulfil criteria for a PH centre and have a CTEPH multidisciplinary team consisting of a PEA surgeon, BPA interventionist, PH specialist, and thoracic radiologist, trained in high-volume PEA and/or BPA centres.**
- **The team should meet regularly to review new referrals and posttreatment follow-up cases. Ideally, CTEPH centres should have PEA activities (>50/year)[1] and BPAs (>30 patients/year or >100 procedures/year)[2], as these figures have been associated with better outcome. The CTEPH centres should also manage medically treated patients. Based on regional requirements, these numbers may be adjusted for the country's population, ideally concentrating care and expertise in high-volume centres.**

1. Mayer et al, J Thorac Cardiovasc Surg 2011;141:702–710; 2. Brenot et al. Eur Respir J 2019;53:1802095

CONCLUSIONS

CTEPH is a rare complication of pulmonary embolism

- Why pulmonary embolism does not always resolve is still unknown
- Exact incidence and prevalence are unknown (5–17/10^6)
- CTEPH can be misdiagnosed as acute pulmonary embolism

Risk factors for CTEPH have been identified

- Confirm pulmonary embolism is at the origin of the disease
- Only a small percentage of the CTEPH population

CTEPH diagnosis

- Mandatory examinations are V/Q scan, echocardiography, catheterisation, CT-scan and pulmonary angiography

Management strategy of CTEPH

Xavier JAIS

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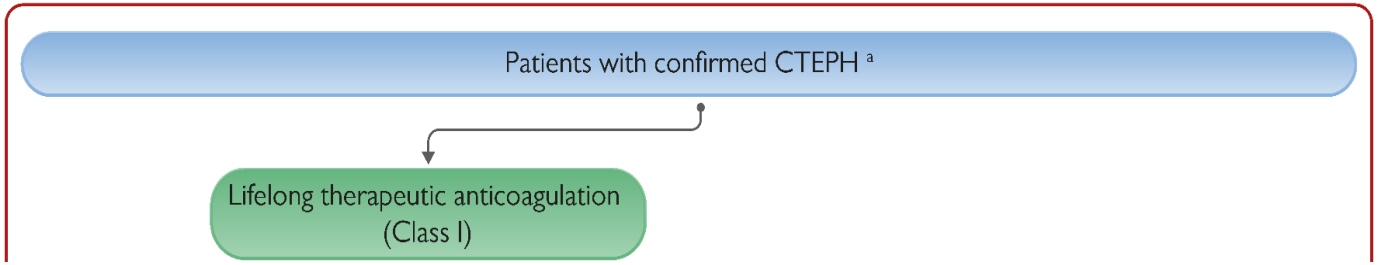
Le Kremlin-Bicêtre – France

Disclosure Statement of Financial Interest

- Financial relationships with relevant companies within the past 24 months:

Affiliation / Financial interest	Commercial Company
Grants/research support:	Acceleron, Janssen, Bayer HealthCare, MSD
Honoraria or consultation fees:	Janssen, MSD
Participation in a company sponsored bureau:	No
Stock shareholder:	No
Spouse / partner:	No
Other support / potential conflict of interest:	No

Management strategy of CTEPH



- Lifelong therapeutic anticoagulation is recommended for patients with CTEPH
- The goal of anticoagulation is to prevent the formation of new blood clots
- Vitamin K antagonists (VKAs) are the standard of care for CTEPH but non-vitamin K oral anticoagulants (NOACs) are increasingly being used [1,2]
- NOACs are less effective than VKAs in antiphospholipid syndrome (10% of CTEPH) [1,2]

1. Pengo V *et al. Blood 2018;* 2. Ordi-Ros J *et al. Ann Intern Med 2019.*
2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension
(*European Heart Journal*; 2022 – doi: 10.1093/eurheartj/ehac237 and *European Respiratory Journal*; 2022 – doi: 10.1183/13993003.00879-2022)

Recommendations for anticoagulation in CTEPH

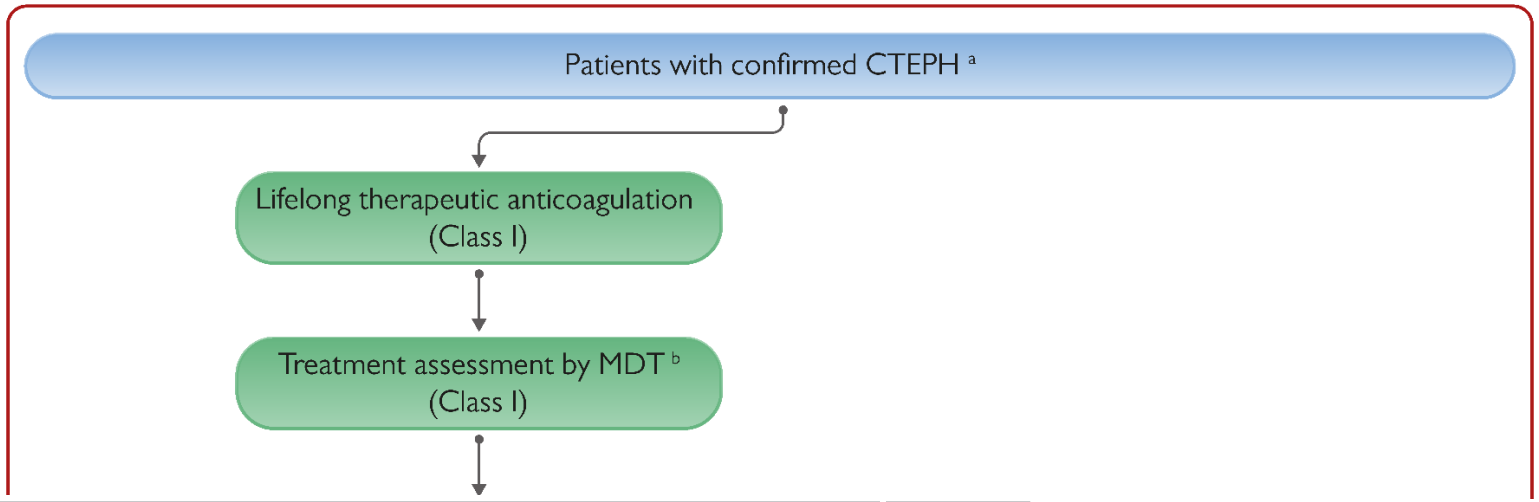
Guidelines 2022

ESC ERS

Recommendations	Class	Level
CTEPH		
Lifelong, therapeutic doses of anticoagulation are recommended in all patients with CTEPH	I	C
Antiphospholipid syndrome testing is recommended in patients with CTEPH	I	C
In patients with **CTEPH and antiphospholipid syndrome**, anticoagulation with **VKAs is recommended**	I	C

Humbert M, *et al. Eur Respir J* 2022; *in press*: 2200879; Humbert M, *et al. Eur Heart J* 2022; 43: 3618-3731.

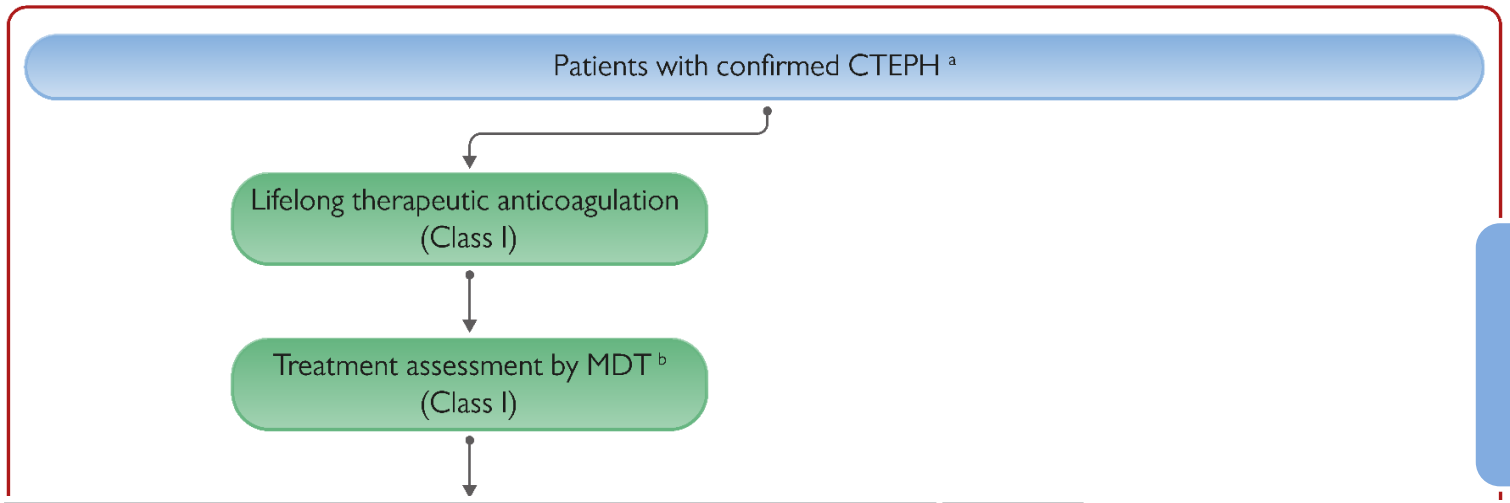
Management strategy of CTEPH



2022 Guidelines	Class
It is recommended that all patients with CTEPH are reviewed by a CTEPH team for the assessment of multi-modality management	I

Humbert M, *et al. Eur Respir J* 2022
Humbert M, *et al. Eur Heart J* 2022

Management strategy of CTEPH



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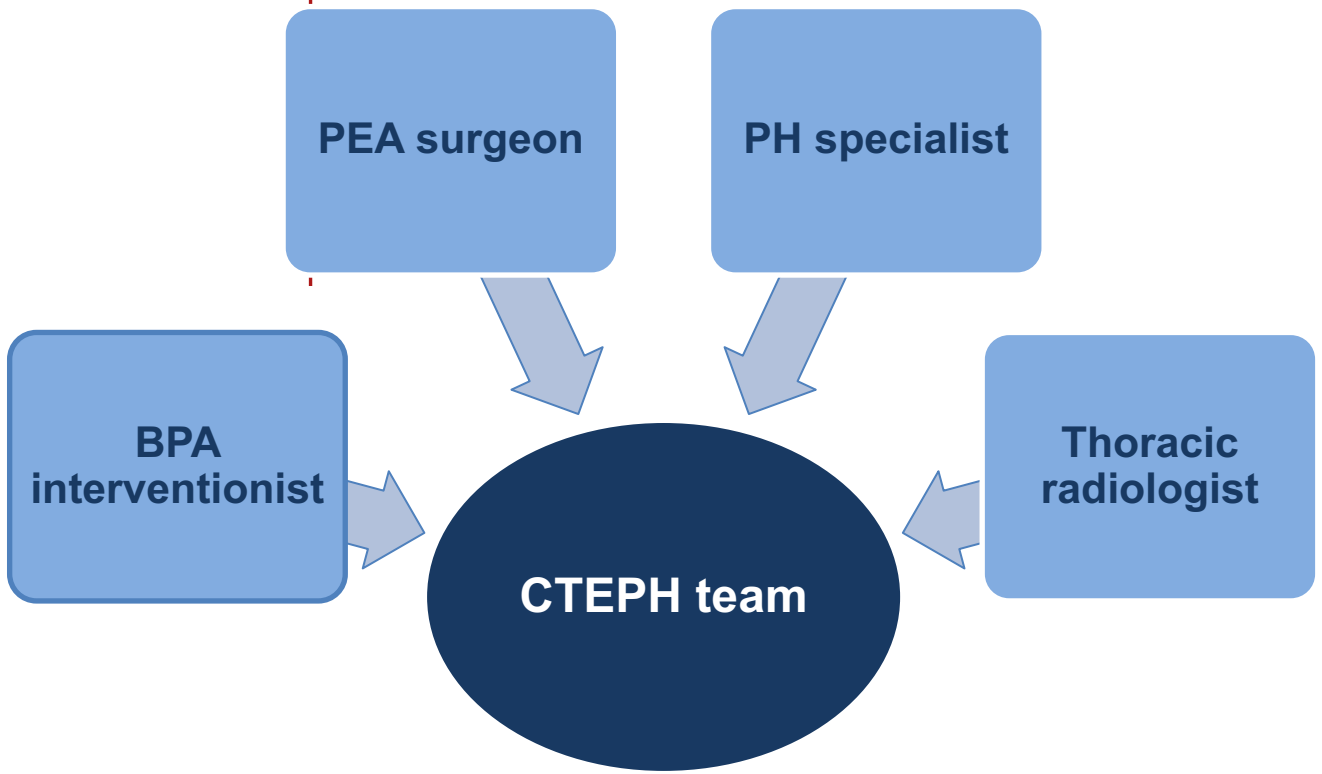
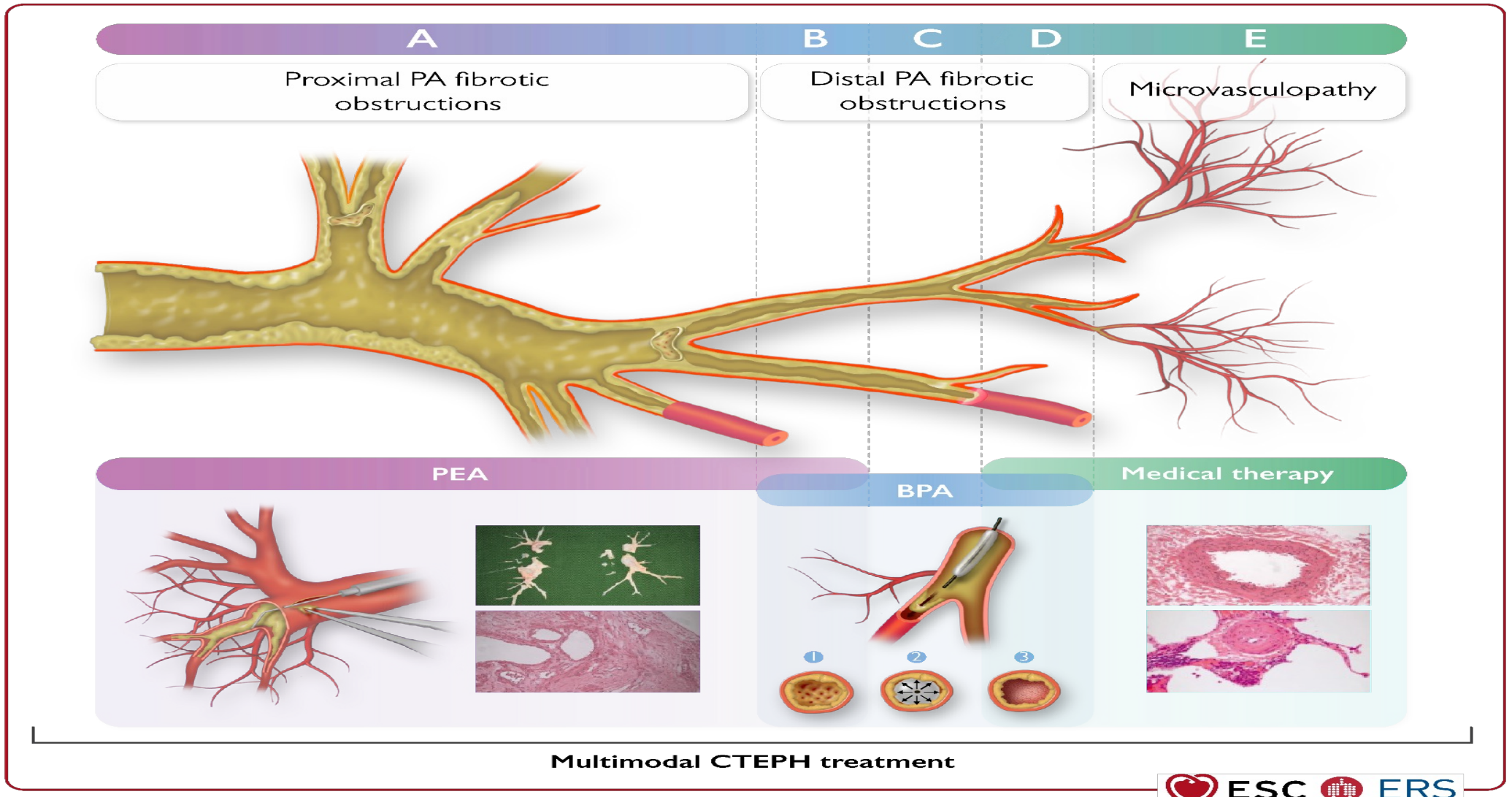


Humbert M, *et al. Eur Respir J* 2022
Humbert M, *et al. Eur Heart J* 2022

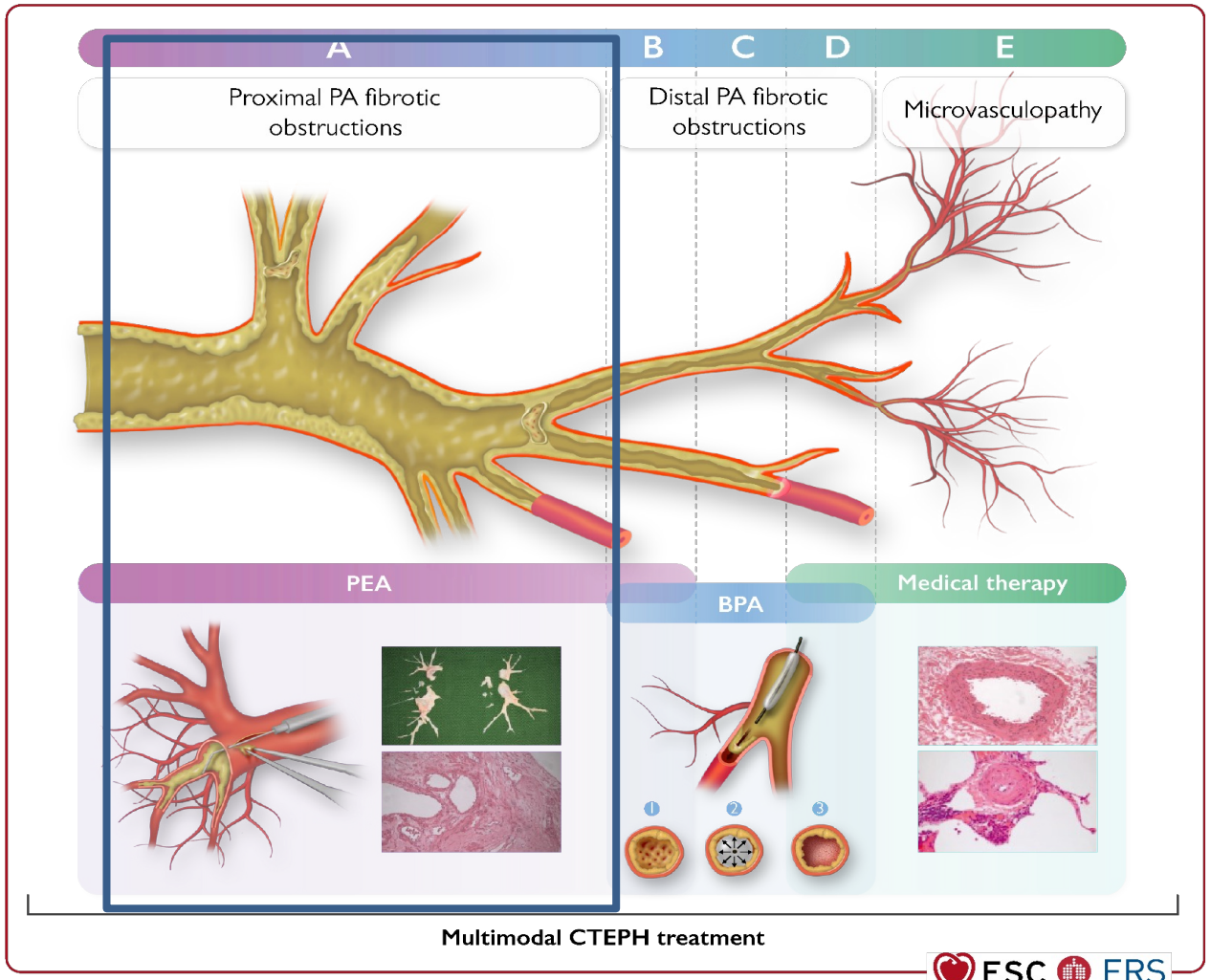
Rationale for multimodal therapy

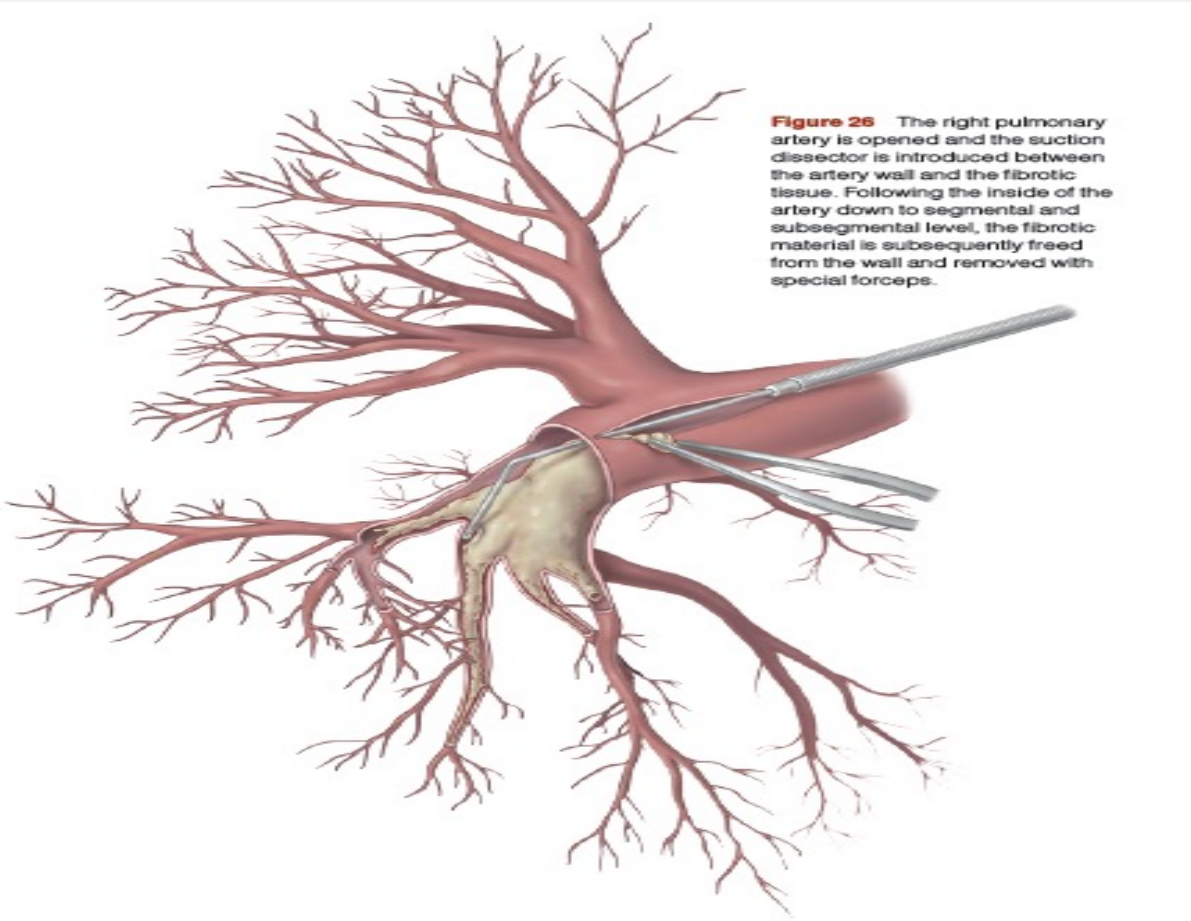


PEA: pulmonary endarterectomy; BPA: balloon pulmonary angioplasty

Humbert M, *et al. Eur Respir J* 2022
Humbert M, *et al. Eur Heart J* 2022

Pulmonary endarterectomy (PEA)





2022 Guidelines	Class
PEA is recommended as the treatment of choice for patients with CTEPH and fibrotic obstructions within pulmonary arteries accessible by surgery	I

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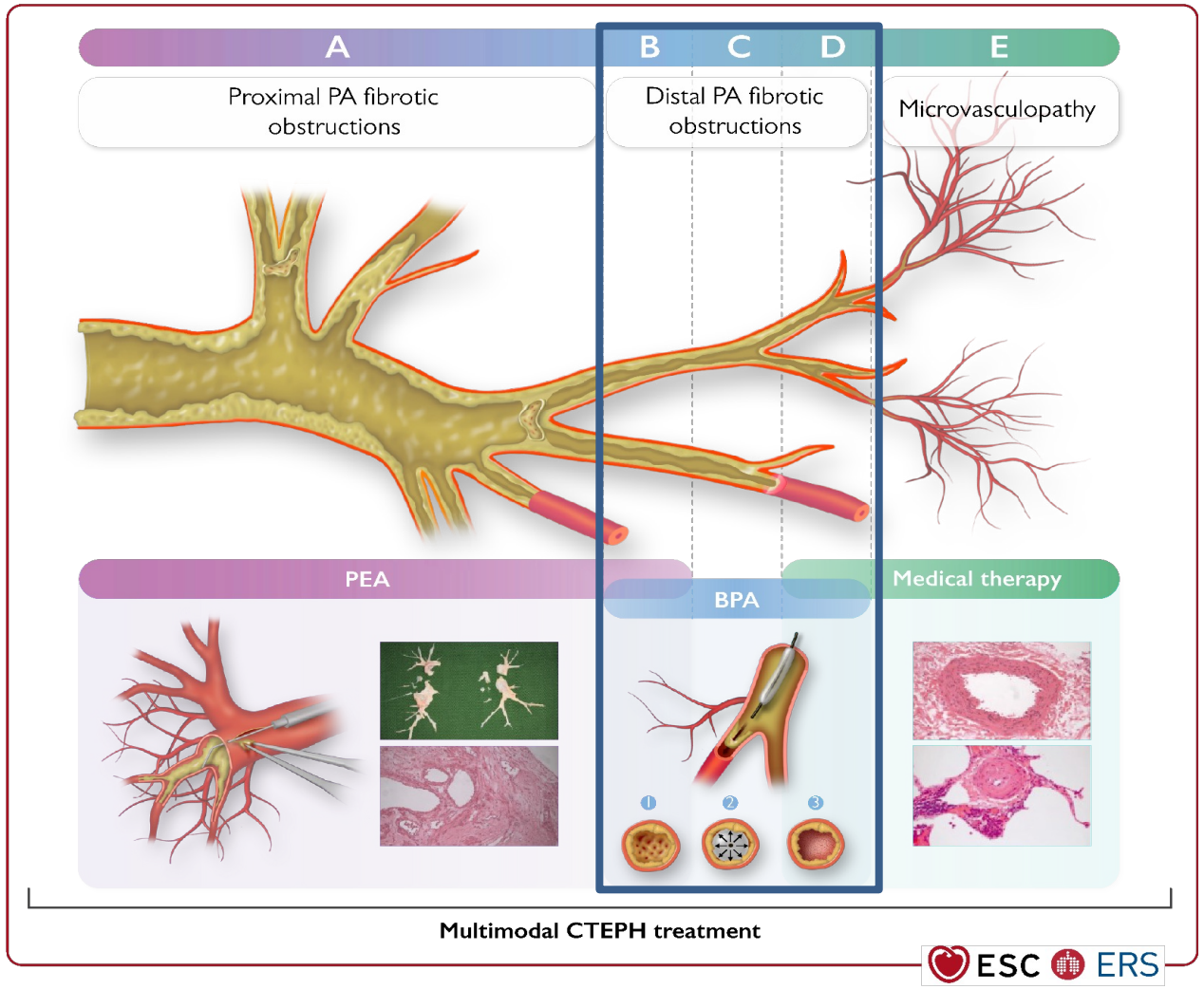
Balloon pulmonary angioplasty (BPA)



BPA is recommended in patients who are technically inoperable

I

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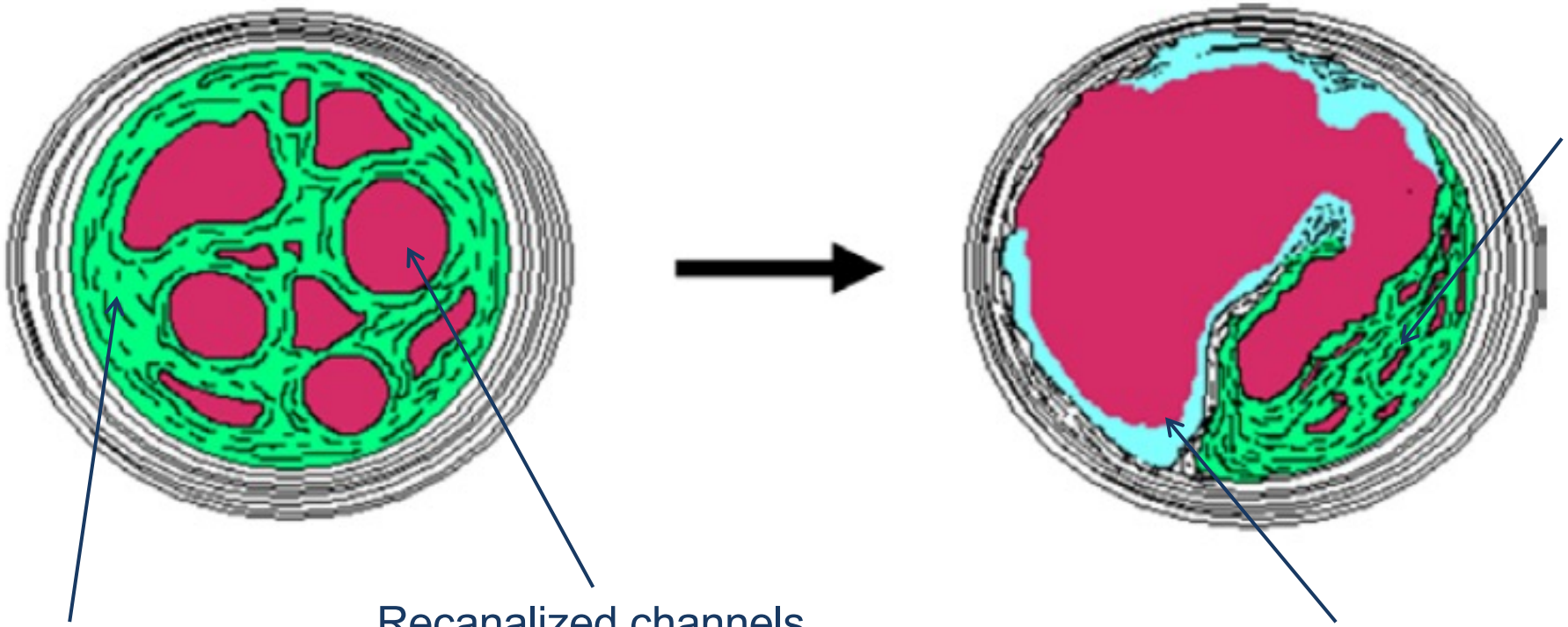
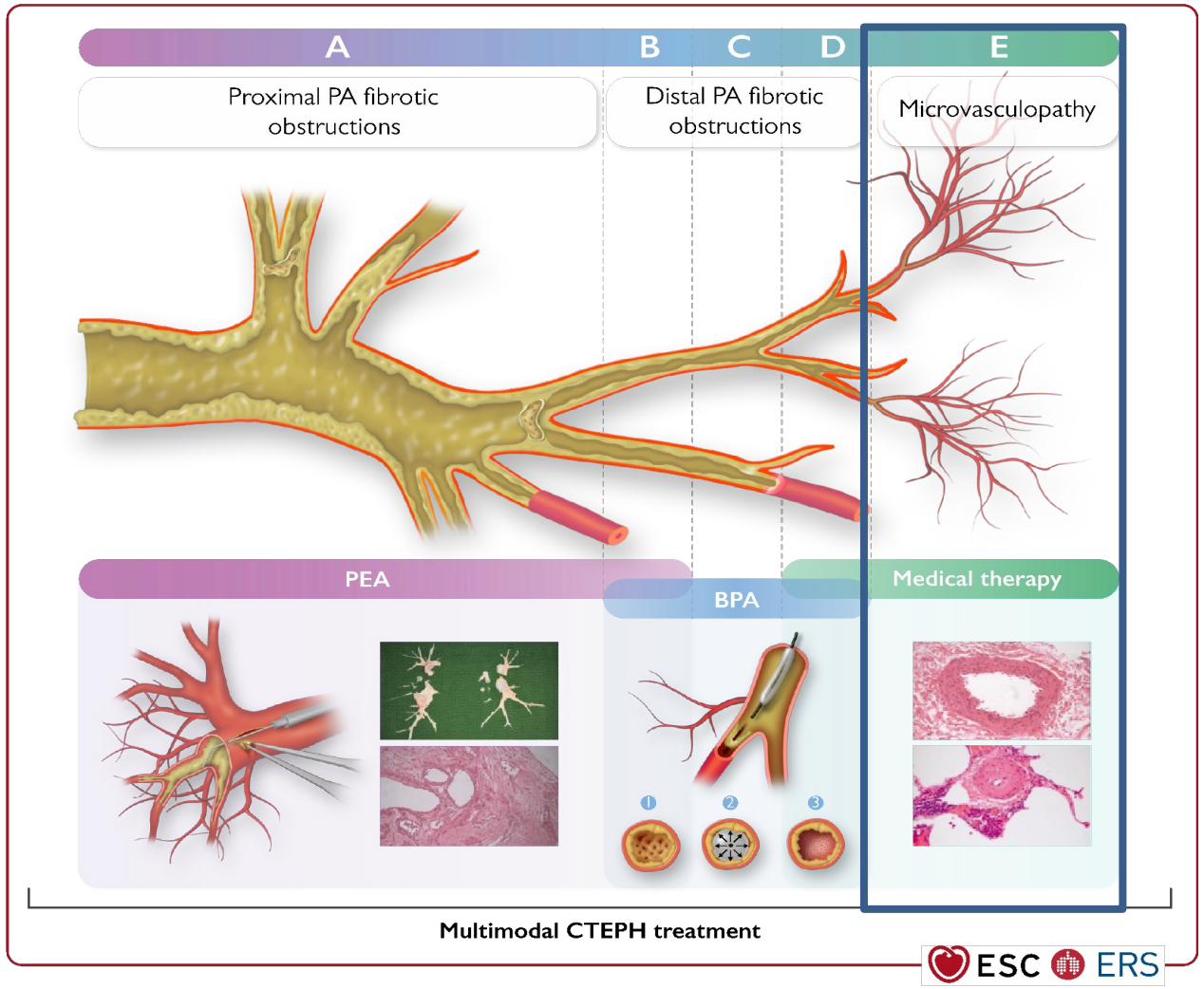
Principles of BPA

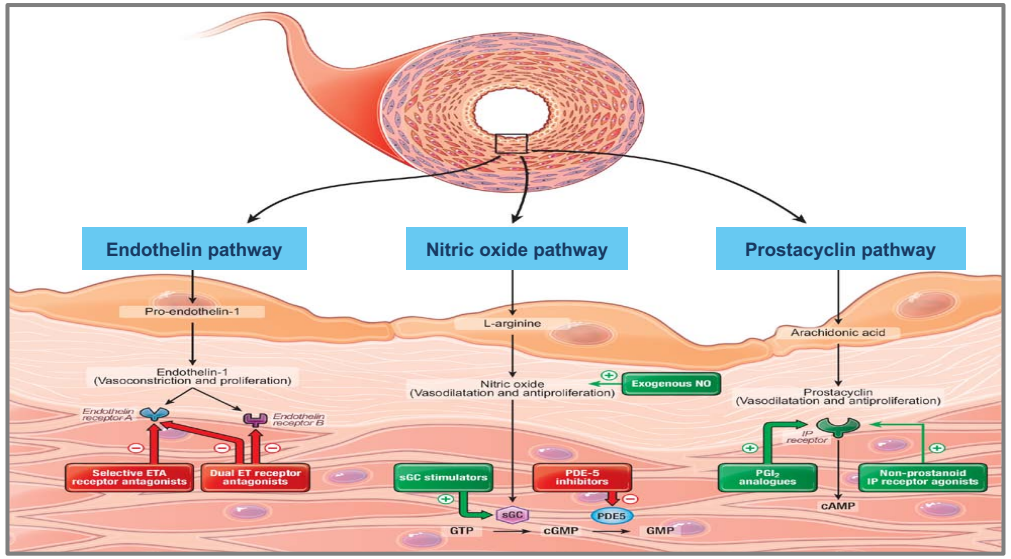
Before angioplasty

After angioplasty



Medical therapy





Endothelin pathway

Endothelin receptor antagonists

- Ambrisentan
- Bosentan[1]
- Macitentan[4]

Nitric oxide pathway

PDE5 inhibitors

- Sildenafil[2]
- Tadalafil

sGC stimulators

- Riociguat[3]

Prostacyclin pathway

Prostanoids

- Epoprostenol iv
- Treprostinil sc[5]

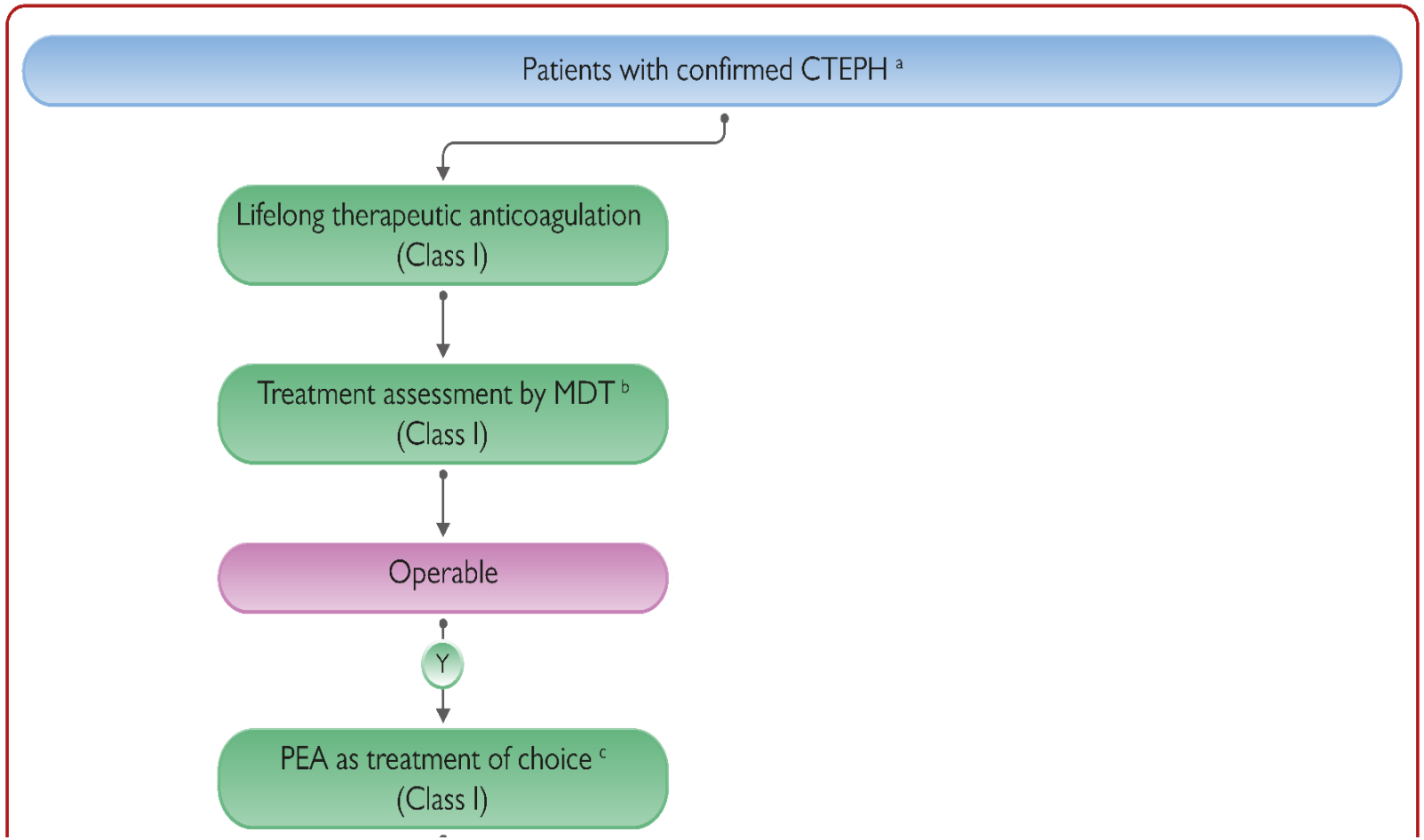
Non-prostanoid IP receptor agonist

- Selexipag[6]

1. Jaïs X *et al. J Am Coll Cardiol* 2008; 2. Suntharalingam J *et al. Chest* 2008; 3. Ghofrani HA *et al. NEJM* 2013; 4. Ghofrani HA *et al. Lancet Respit Med* 2017; 5. Sadushi-Kolici R, *et al. Lancet Respir Med* 2018; Ogo T, *et al. Eur Respir J 2021.*

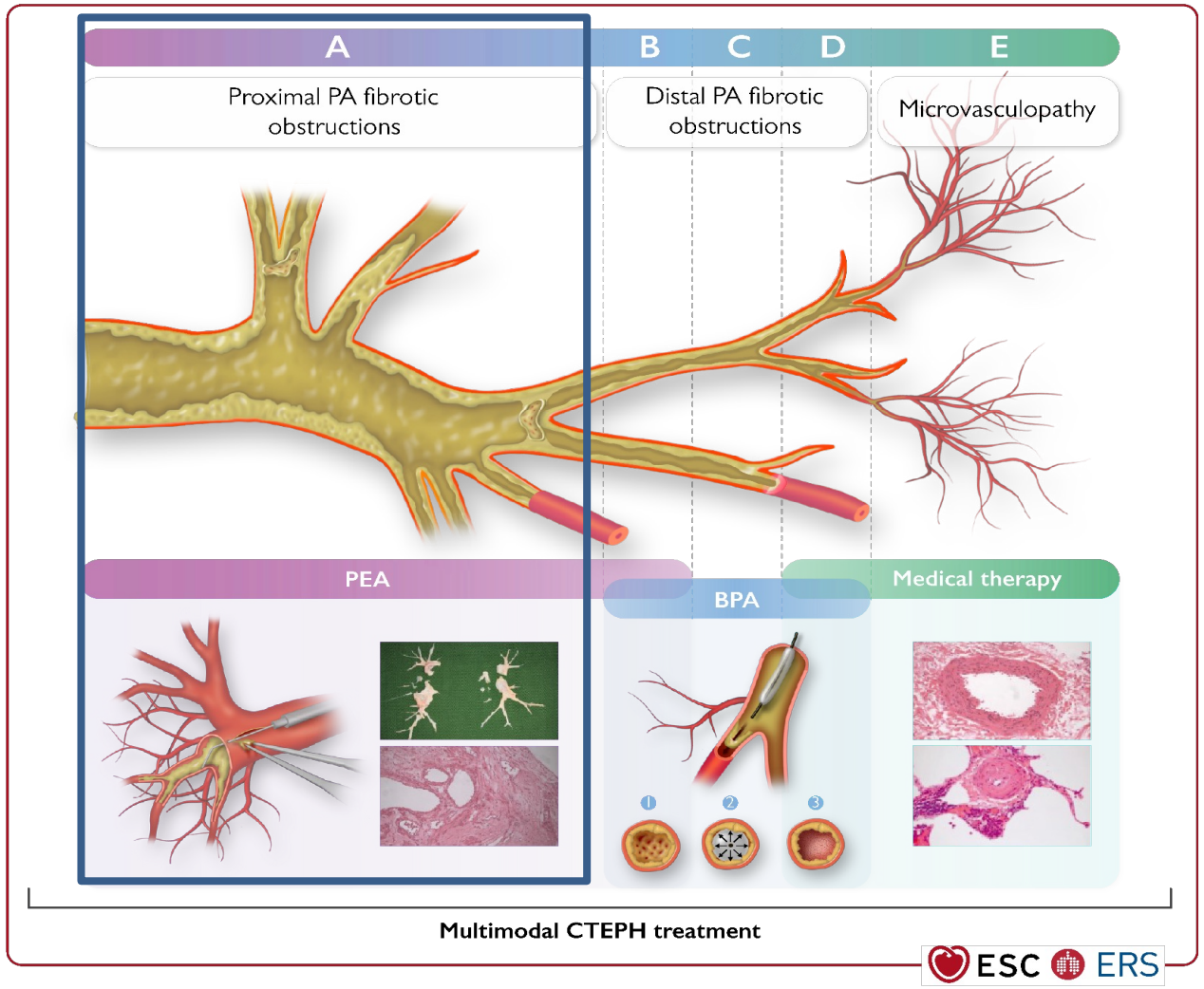
2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

Management strategy of CTEPH



Humbert M, *et al. Eur Respir J* 2022
Humbert M, *et al. Eur Heart J* 2022

Pulmonary endarterectomy (PEA)



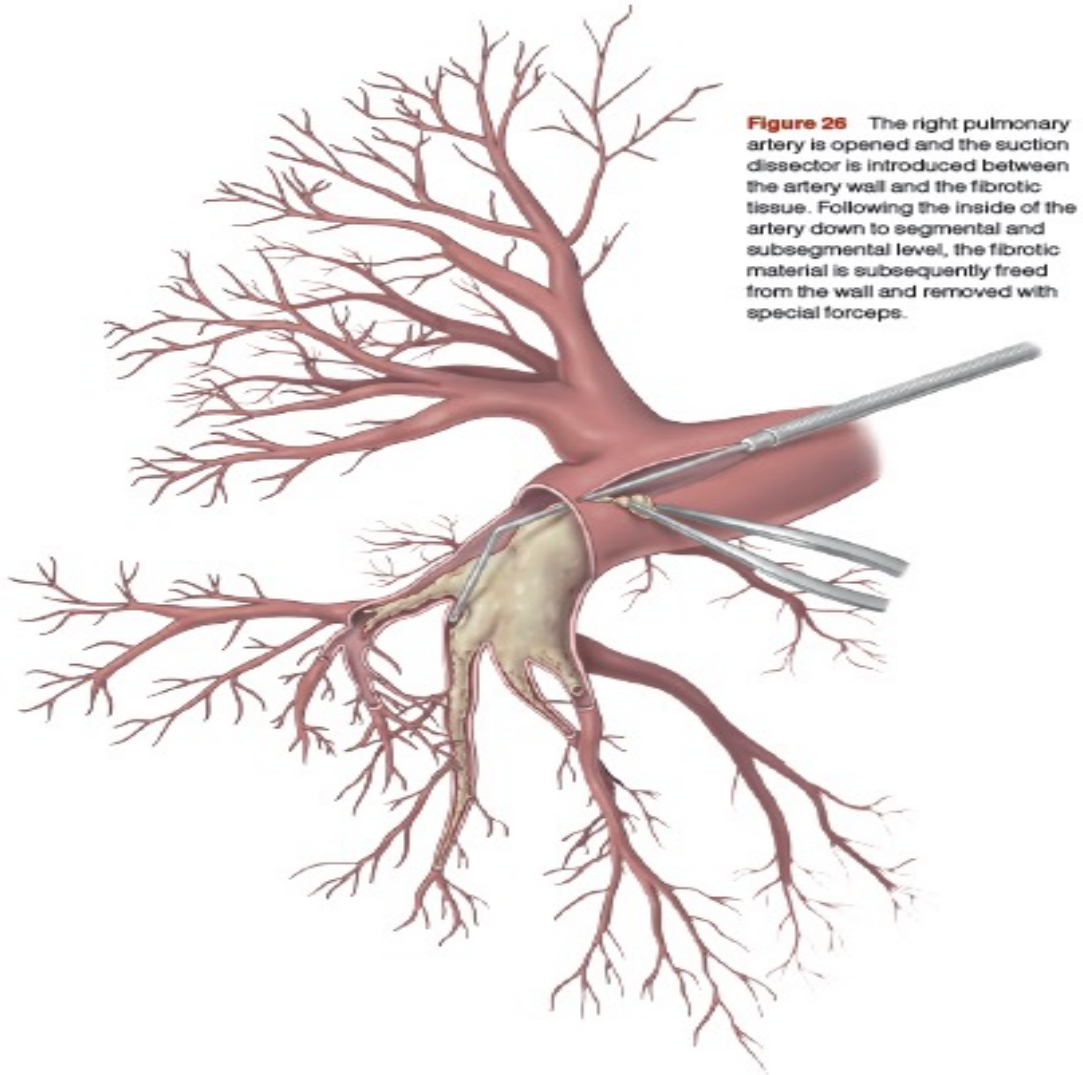


Figure 26 The right pulmonary artery is opened and the suction dissector is introduced between the artery wall and the fibrotic tissue. Following the inside of the artery down to segmental and subsegmental level, the fibrotic material is subsequently freed from the wall and removed with special forceps.

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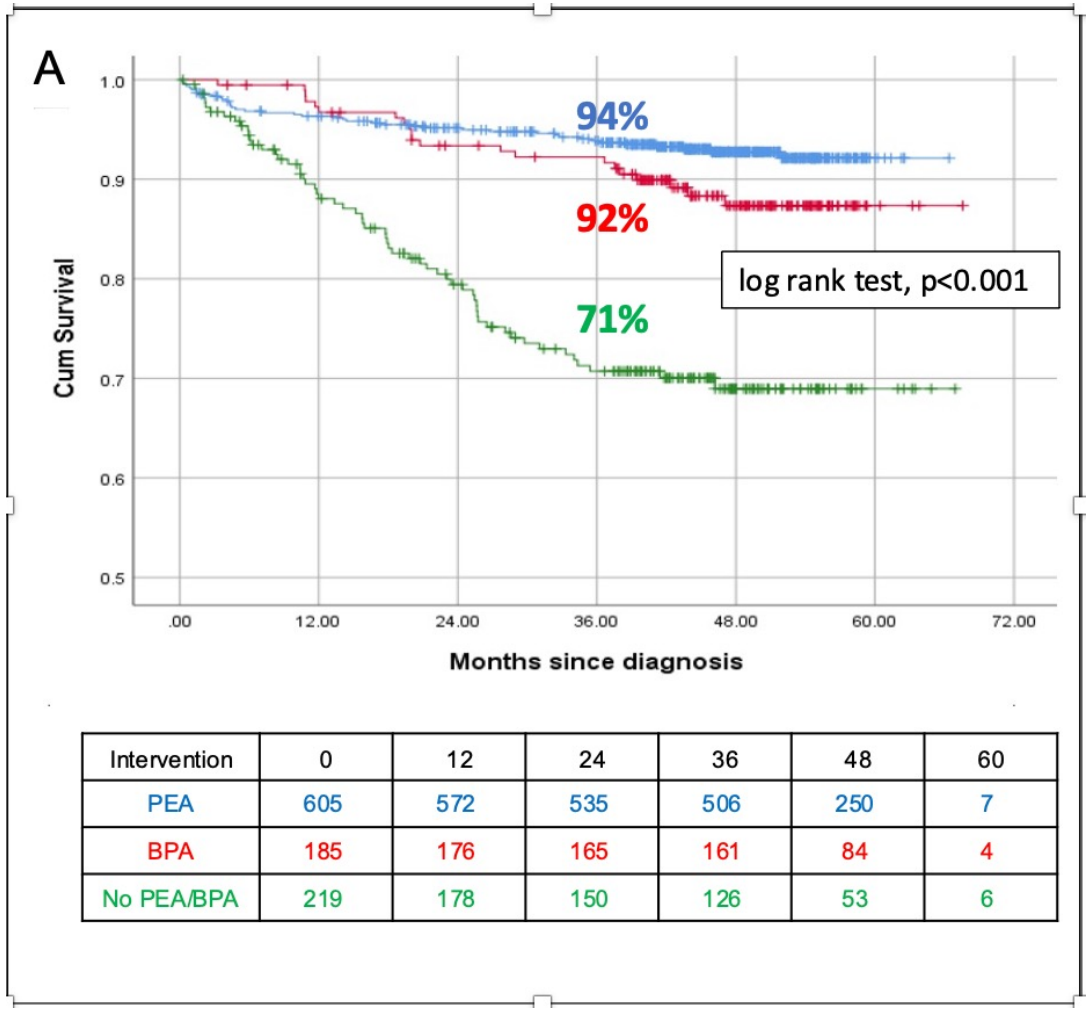
Outcome after PEA

- PEA improves symptoms, exercise capacity, quality of life
- PEA leads to a decrease in pulmonary pressures and resistance

Survival at 3 years:

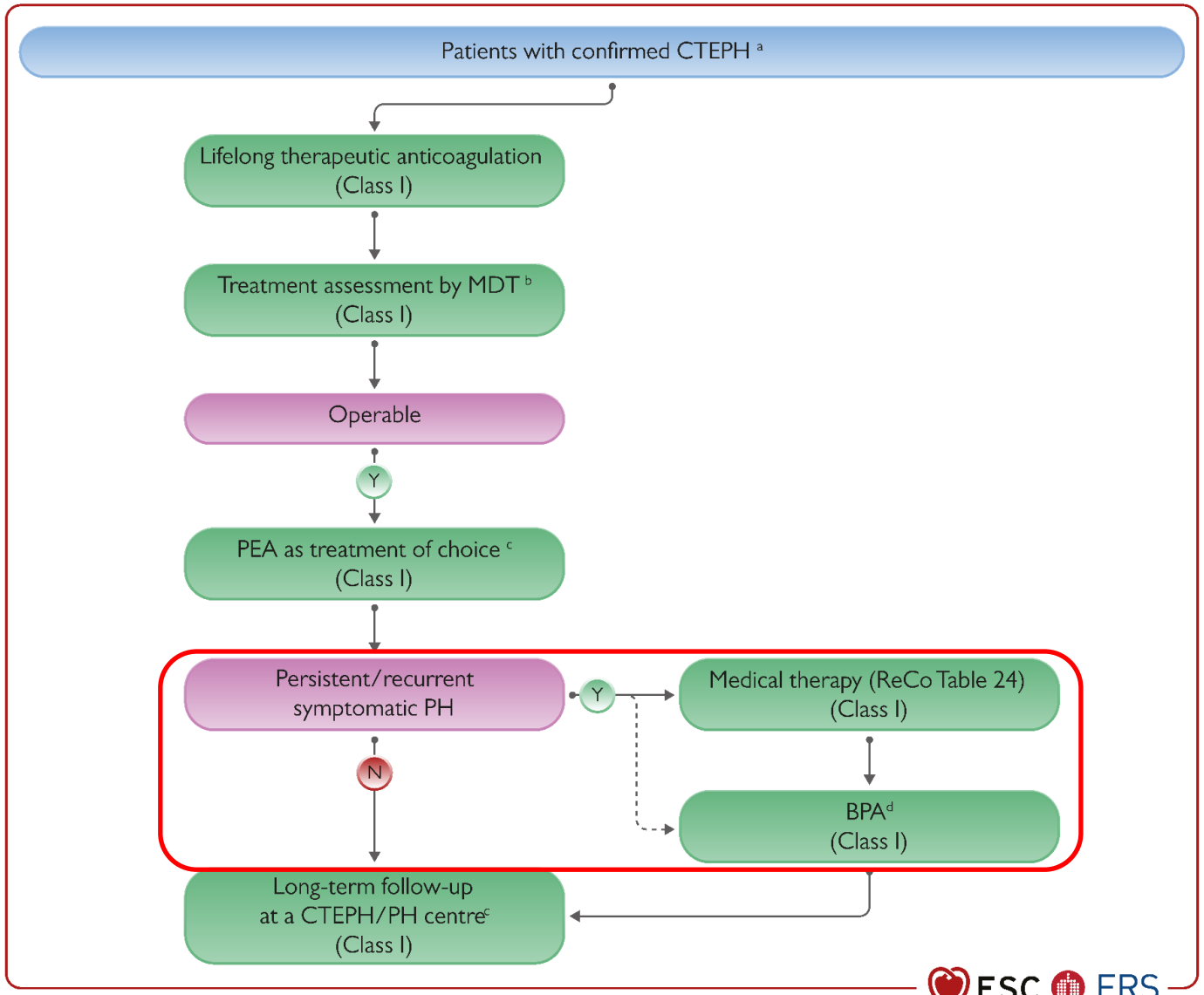
Operated patients= 94%



Intervention	0	12	24	36	48	60
PEA	605	572	535	506	250	7
BPA	185	176	165	161	84	4
No PEA/BPA	219	178	150	126	53	6

Delcroix M, et al. *ERS* 2021

Management strategy of operable CTEPH



Humbert M, *et al. Eur Respir J* 2022
Humbert M, *et al. Eur Heart J* 2022

World Health Organization (WHO) classification of functional status of patients with pulmonary hypertension

Class	Description
WHO-FC I	Patients with PH but without resulting limitation of physical activity. Ordinary physical activity does not cause undue dyspnoea or fatigue, chest pain, or near syncope
WHO-FC II	Patients with PH resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity causes undue dyspnoea or fatigue, chest pain, or near syncope
WHO-FC III	Patients with PH resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity causes undue dyspnoea or fatigue, chest pain, or near syncope.
WHO-FC IV	Patients with PH with an inability to carry out any physical activity without symptoms. These patients manifest signs of right heart failure. Dyspnoea and/or fatigue may even be present at rest. Discomfort is increased by any physical activity

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(*European Heart Journal*; 2022 – doi: 10.1093/eurheartj/ehac237 and *European Respiratory Journal*; 2022 – doi: 10.1183/13993003.00879-2022)

Which patients should be offered medical treatment and/or BPA after surgery?

Outcome 3-6 months post-PEA surgery (UK cohort, 880 consecutive patients)

- 85% of patients in FC I or II
- mPAP ≤ 20 mmHg: 28% of patients; 21-24 mmHg: 21%; **≥ 25 mmHg: 51%**

Time point of measurement

- 3-6 months after surgery

Haemodynamic criteria

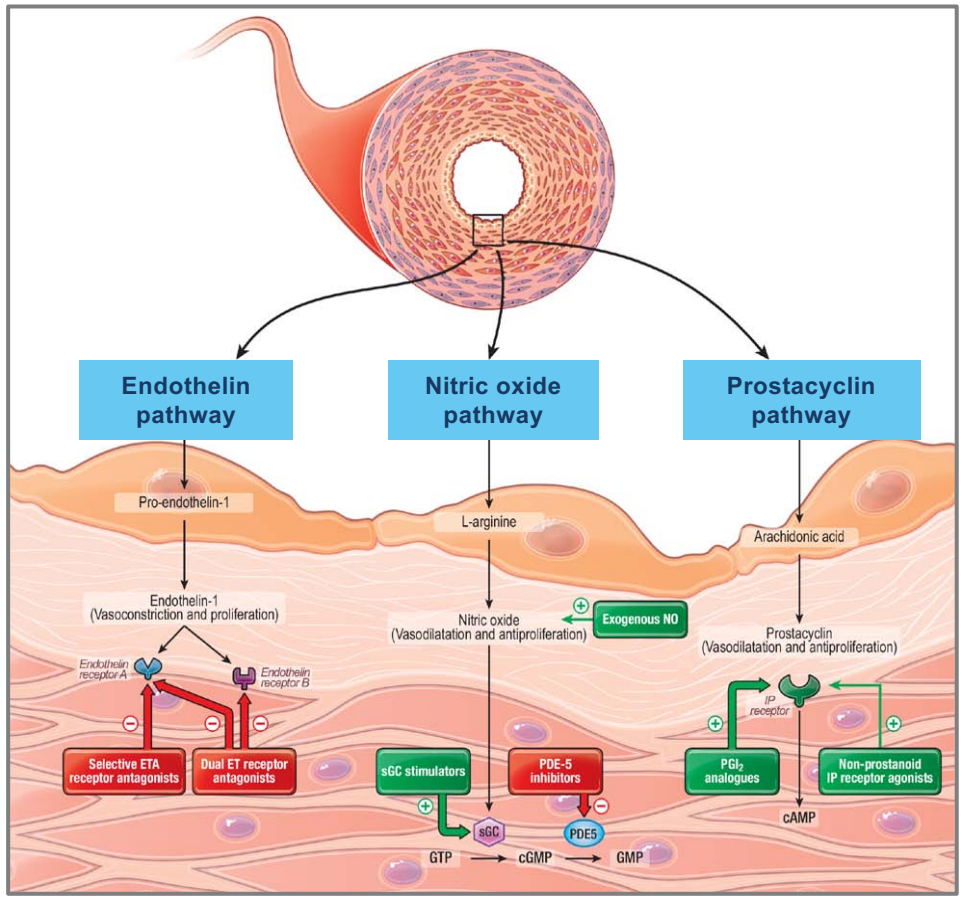
- mPAP ≥ 30 mmHg correlated with the initiation of PH-targeted therapy post-PEA
- **mPAP ≥ 38 mmHg and PVR ≥ 425 dyn.s.cm^{-5}** ***correlated with worse long-term survival***

Recommendation

- Re-assessment after surgery and long-term follow-up important,
- **Consider add-on therapy (medical/BPA) if symptoms (NYHA FC II-IV) and mPAP > 25 mmHg, PAWP ≤ 15 mmHg, PVR > 3 UW.**

Cannon JE *et al, Circulation* 2016

Medical therapy in CTEPH



Endothelin pathway

Endothelin receptor antagonists

- Ambrisentan
- Bosentan[1]
- Macitentan[4]

Nitric oxide pathway

PDE5 inhibitors

- Sildenafil[2]
- Tadalafil

sGC stimulators

- Riociguat[3]

Prostacyclin pathway

Prostanoids

- Epoprostenol iv
- Treprostinil sc[5]

Non-prostanoid IP receptor agonist

- Selexipag[6]

2022 Guidelines	Class
Riociguat is recommended for symptomatic patients with persistent/recurrent PH after PEA	I
Treprostinil s.c. may be considered in patients in WHO-FC III–IV who have persistent/recurrent PH after PEA	IIb

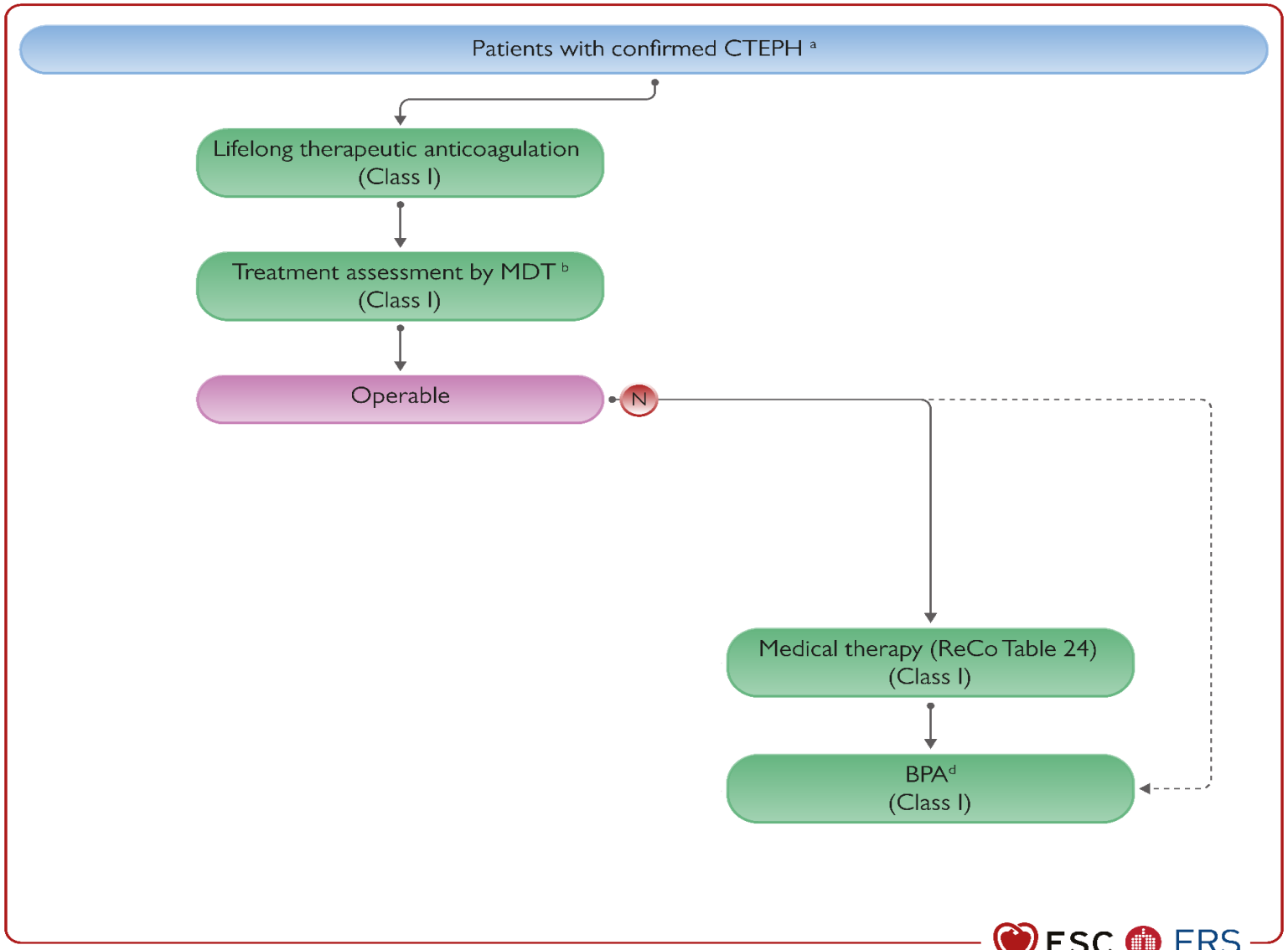
1. Jaïs X *et al. J Am Coll Cardiol* 2008; 2. Suntharalingam J *et al. Chest* 2008; 3. Ghofrani HA *et al. NEJM* 2013; 4. Ghofrani HA *et al. Lancet Respit Med* 2017; 5. Sadushi-Kolici R, *et al. Lancet Respir Med* 2018; 6. Ogo T, *et al. Eur Respir J 2021;* Humbert M, *et al. Eur Respir J* 2022.

Multimodal approach combining medical treatment and BPA in patients with persistent symptomatic PH after surgery

Author	Year	Patients (n)	Time from PEA to BPA (months)	Medical treatment before BPA	Before BPA PVR (dyn.s.cm^{-5})	After BPA PVR (dyn.s.cm^{-5})	Treatment effect
Shimura	2015	9	49.2 (2.7-7.9)	?	1250 (624-1512)	448 (280-520)	-64%
Yanaka	2018	10	7.3±2.3	20%	386±42	242±39	-37%
Araszkiewicz	2019	15	28.1±25.8	100%	552±185	344±124	-38%
Ito	2021	25	1-12	44%	392±160	296±40	-25%
Kirkby	2023	20	47±39.3	100%	532±172	454±182	-15%

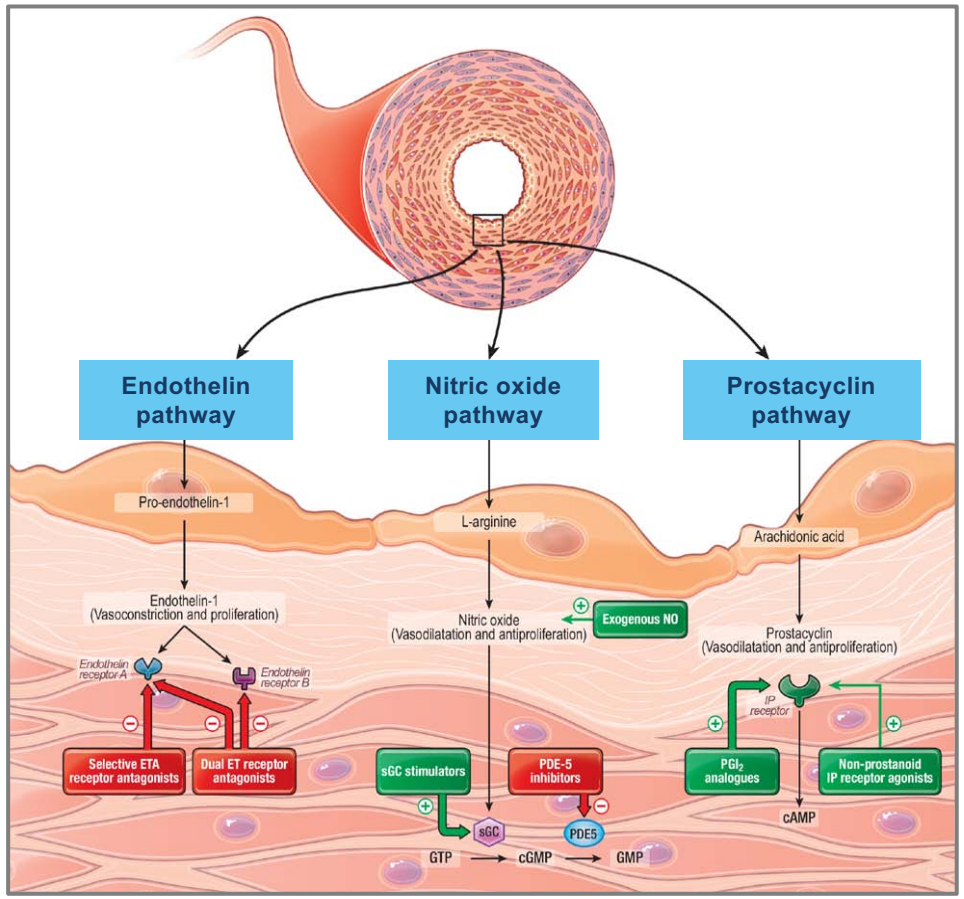
Shimura N *et al. Int J Cardiol* 2015; Yanaka K *et al. J Am Heart Assoc* 2018; Araszkiewicz A *et al. Int J Cardiol* 2019; Ito R *et al. Int J Cardiol* 2021; Kirkby LC *et al.* Pulm Circ 2023.

Management strategy of inoperable CTEPH



Humbert M, *et al. Eur Respir J* 2022; Humbert M, *et al. Eur Heart J* 2022.

Medical therapy in CTEPH



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Prostacyclin pathway

Prostanoids

- Epoprostenol iv
- Treprostinil sc[5]

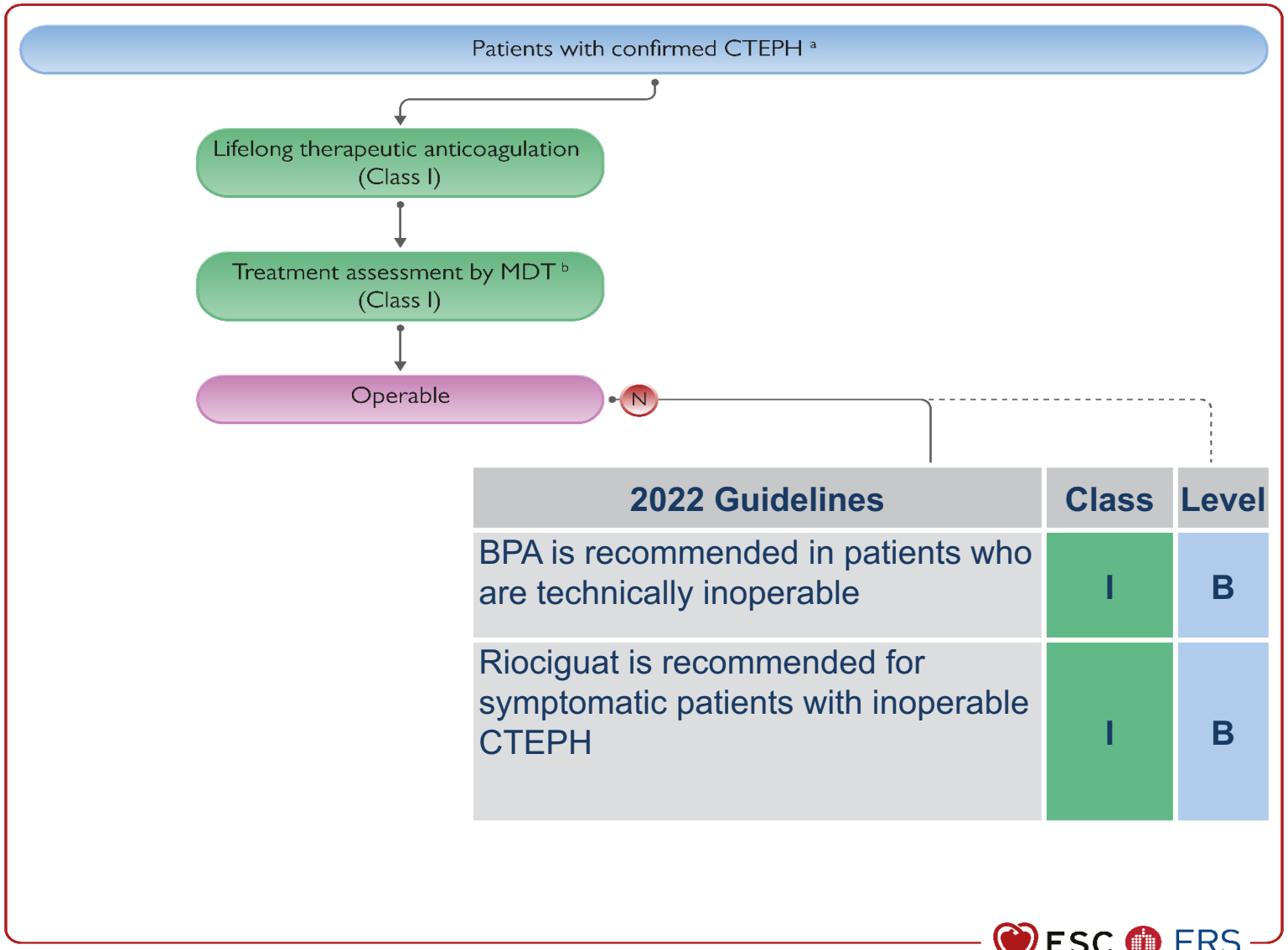
Non-prostanoid IP receptor agonist

- Selexipag[6]

2022 Guidelines	Class	Level
Riociguat is recommended for symptomatic patients with inoperable CTEPH	I	B
Treprostinil s,c, may be considered in patients in WHO-FC III-IV who have inoperable CTEPH	IIb	B
Off-label use of drugs approved for PAH may be considered in symptomatic patients who have inoperable CTEPH	IIb	B
In patients with inoperable CTEPH, a combination of sGc stimulator/PDE-5i, ERA, or prostacyclin analogues may be considered	IIb	C

1. Jaïs X *et al. J Am Coll Cardiol* 2008; 2. Suntharalingam J *et al. Chest* 2008; 3. Ghofrani HA *et al. NEJM* 2013; 4. Ghofrani HA *et al. Lancet Respit Med* 2017; 5. Sadushi-Kolici R, *et al. Lancet Respir Med* 2018; 6. Ogo T, *et al. Eur Respir J 2021;* Humbert M, *et al. Eur Respir J* 2022.

Management strategy of inoperable CTEPH



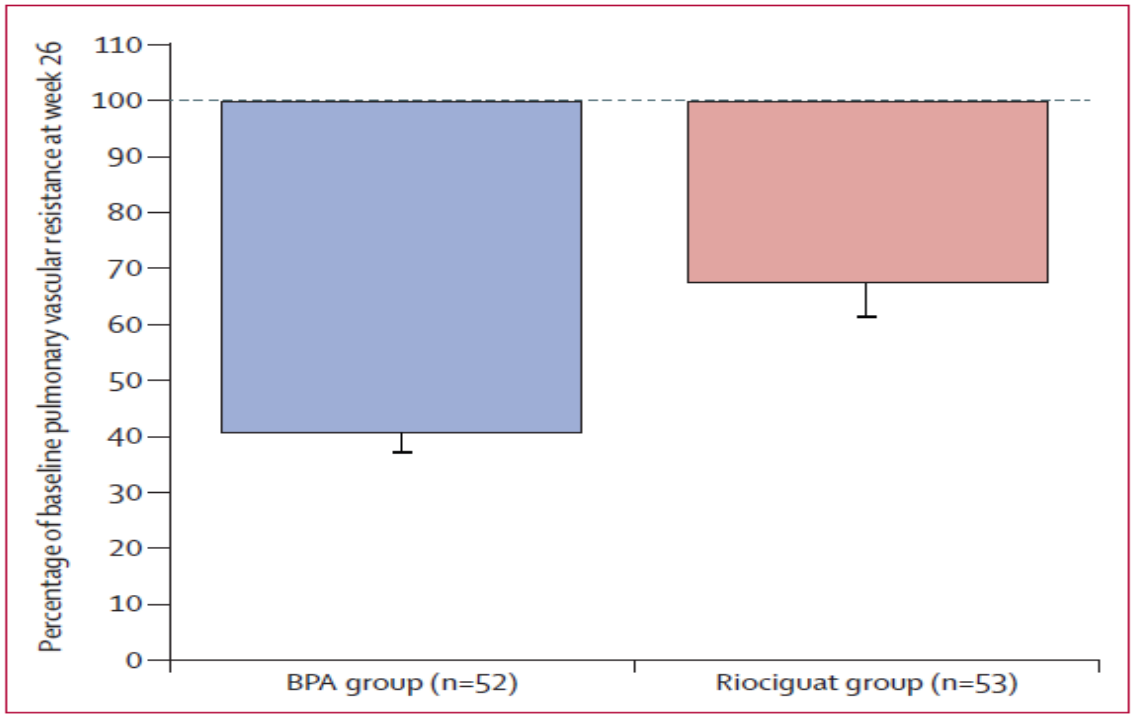
2022 Guidelines	Class	Level
BPA is recommended in patients who are technically inoperable	I	B
Riociguat is recommended for symptomatic patients with inoperable CTEPH	I	B

ESC ERS

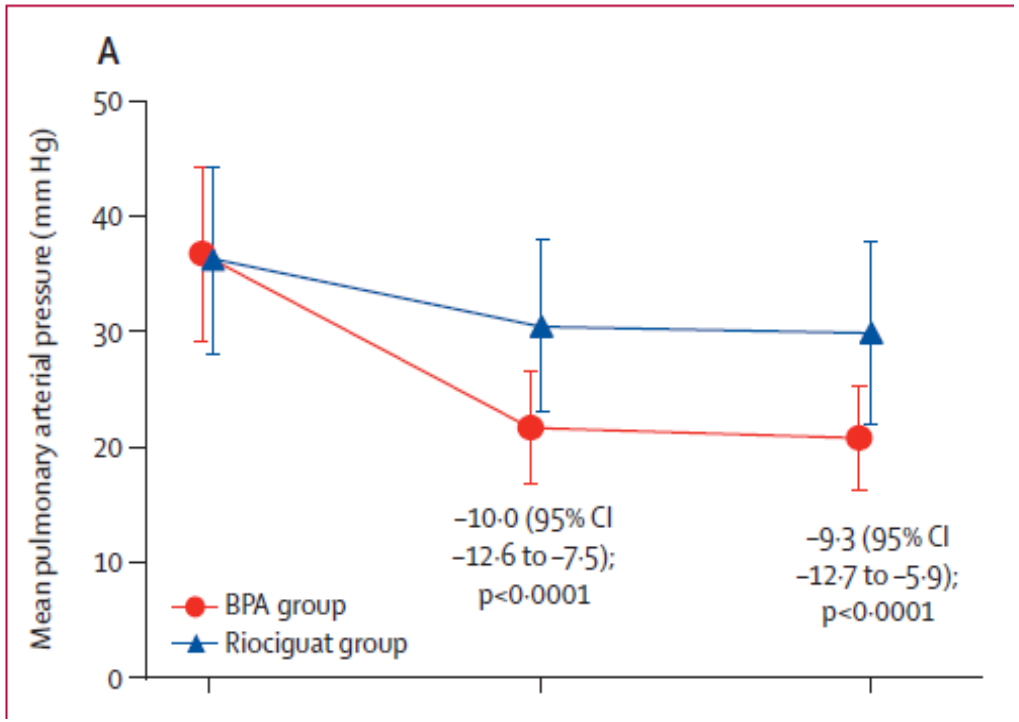
Humbert M, *et al. Eur Respir J* 2022; Humbert M, *et al. Eur Heart J* 2022.

BPA vs Riociguat inoperable CTEPH

RACE study



MR BPA study



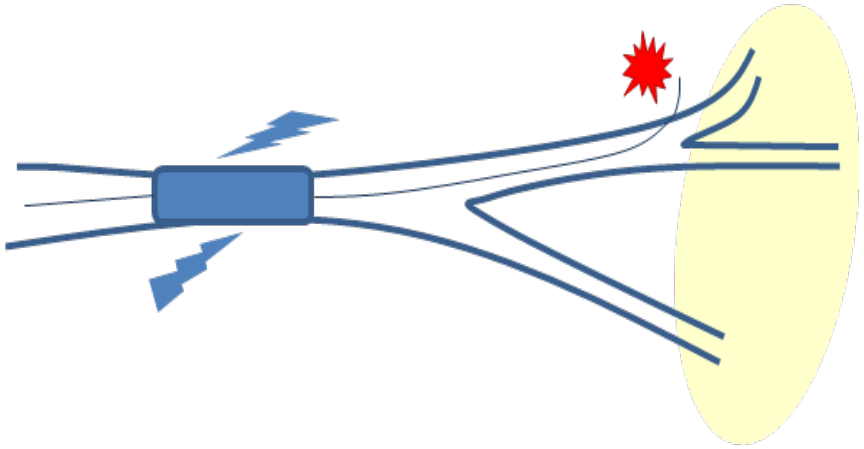
- Treatment-related adverse events were more common with BPA than with riociguat

Jaïs X *et al. Lancet Respir Med 2022.*

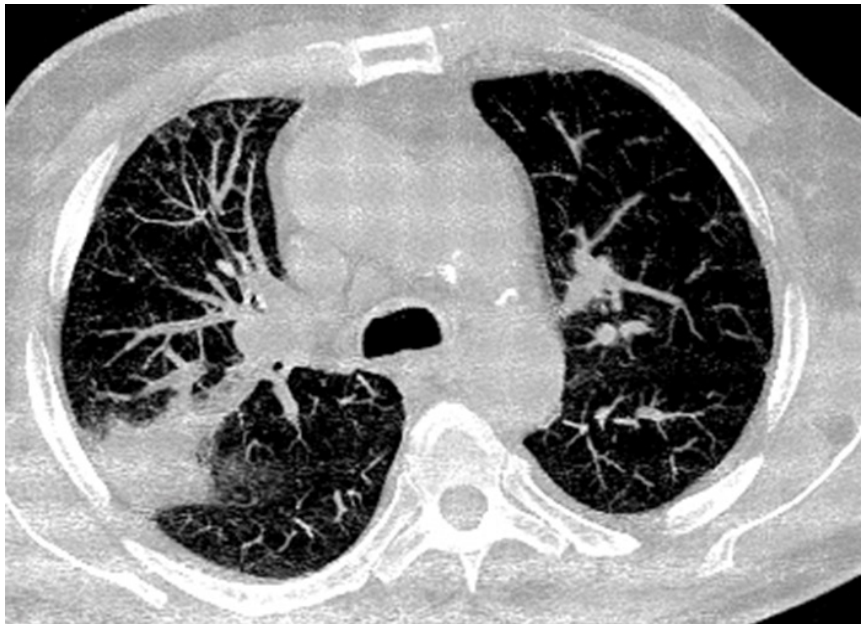
Kawakami T *et al. Lancet Respir Med 2022.*

Complications related to BPA

- **Vascular injury**
  - Pulmonay artery perforation
  - Pulmonary artery rupture



- **Severity of hemodynamics predicts BPA-related complications**
  - High pulmonary pressures and resistance are associated with higher risk of BPA-related complications



1. *Ejiri K et al. Circ Cardiovasc Interv 2018; 2.* Jaïs X *et al. Lancet Respir Med 2022; 3.* Wiedenroth CB *et al. J Heart Lung Transplant 2022.*

Safety of BPA can be improved by pre-treatment with riociguat

Impact of riociguat prior to BPA on BPA-related complications

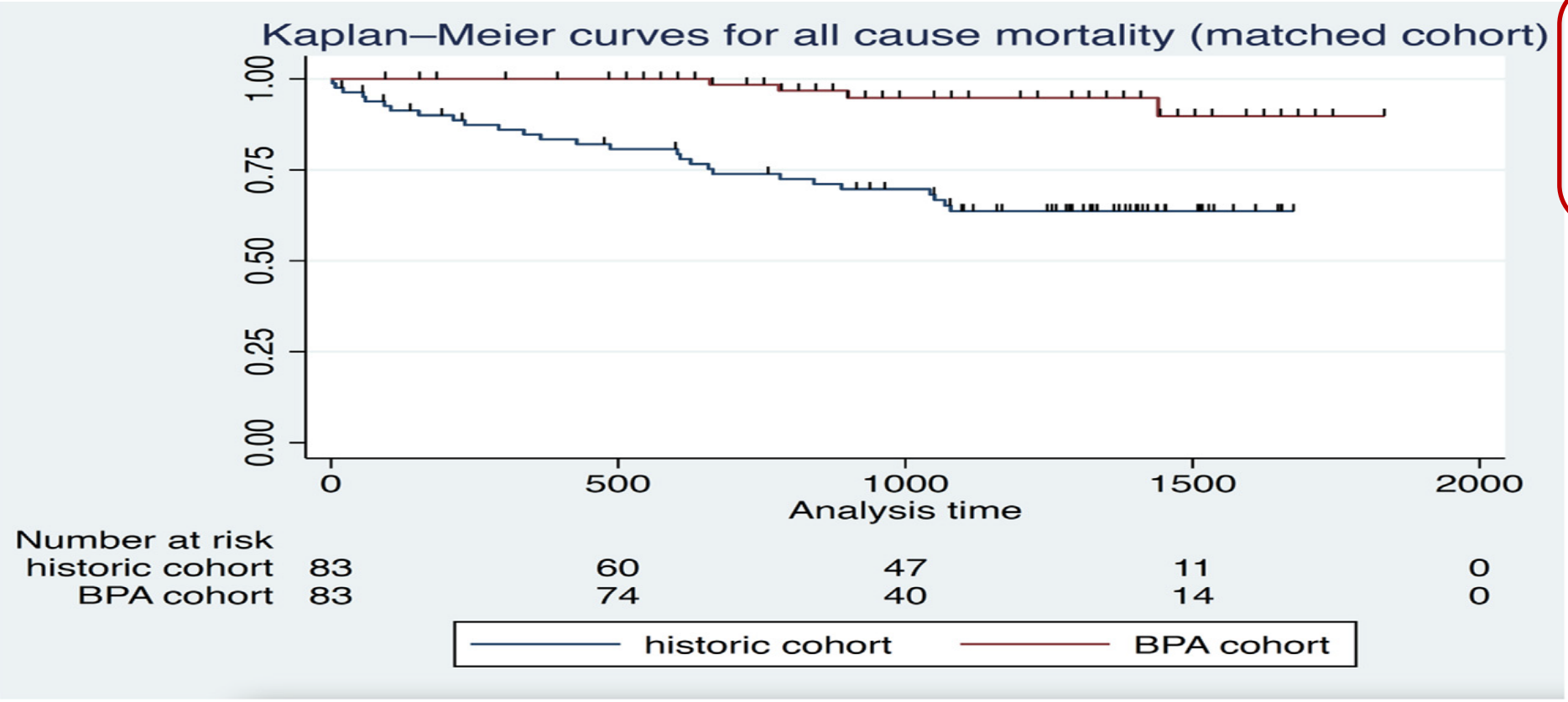
	First-line Riociguat then BPA (N = 36)	First-line BPA (N = 52)	P value
Patients with ≥1 SAE related to BPA– n (%)	5 (14)	22 (42)	0.0045
Patients with ≥1 severe BPA procedure-related complication– n (%)	3 (8)	18 (35)	0.0045

	GRADE			
Recommendations	Quality of evidence	Strength of recommendation	Class	Level
In patients with CTEPH who are candidates for BPA, medical therapy should be considered prior to the intervention	Very low	Conditional	IIa	B

Jaïs X *et al. Lancet Respir Med 2022.*

Outcome after combination of riociguat and BPA in inoperable CTEPH

- Riociguat and BPA improve symptoms, exercise capacity, quality of life
- Riociguat and BPA lead to a decrease in pulmonary pressures and resistance



Survival at 3 years:

Rio and BPA cohort= 93%

Historical cohort= 69%

Wiedenroth CB *et al. J Heart Lung Transplant 2023.*

Multimodal approach, treatment goals and follow-up

2022 Guidelines	Class
A multi-modality approach should be considered for patients with persistent PH after PEA and for patients with inoperable CTEPH	IIa
Long-term follow-up is recommended after PEA and BPA, as well as for patients with CTEPH established on medical therapy	I

- No consensus on what is the therapeutic goal after BPA/PEA or medical therapy
- Most experts accept achieving a good FC (WHO FC I-II) and/or normalization or near normalization of haemodynamics at rest, obtained 3-6 months post-procedure, and improvement in quality of life as a treatment goal.
- Follow-up should include RHC 3-6 months after intervention (PEA or last BPA)
- After successful treatment, yearly non-invasive follow-up (echo and evaluation of exercise capacity)

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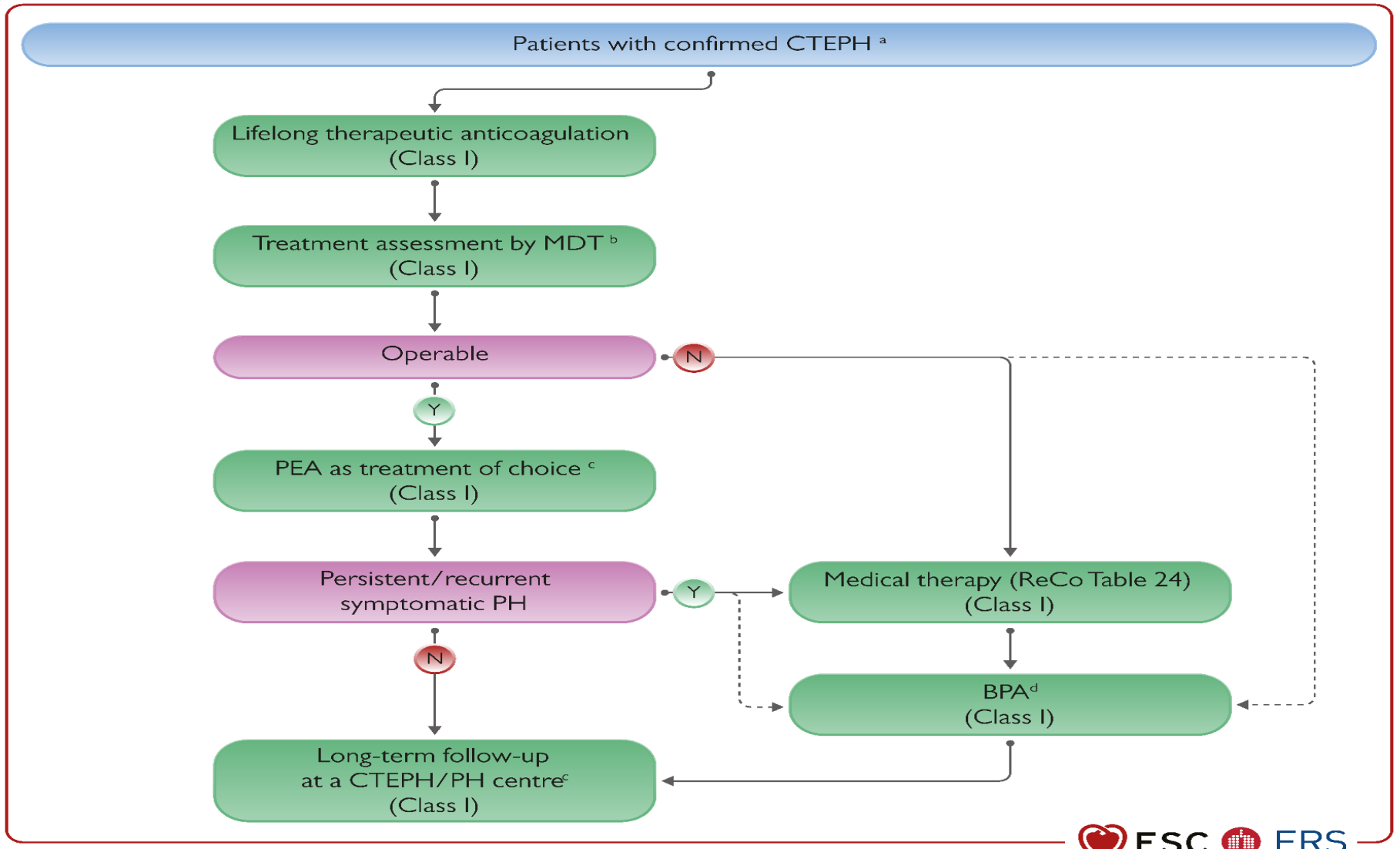
Management strategy of CTEPH in 2023



Humbert M, *et al. Eur Respir J* 2022; Humbert M, *et al. Eur Heart J* 2022.

Thank you for your attention!