HYPERTENSION ARTÉRIELLE PULMONAIRE

–

A 4 STEPS APPROACH FOR DIAGNOSIS AND TREATMENT

DR WIRTZ GIL

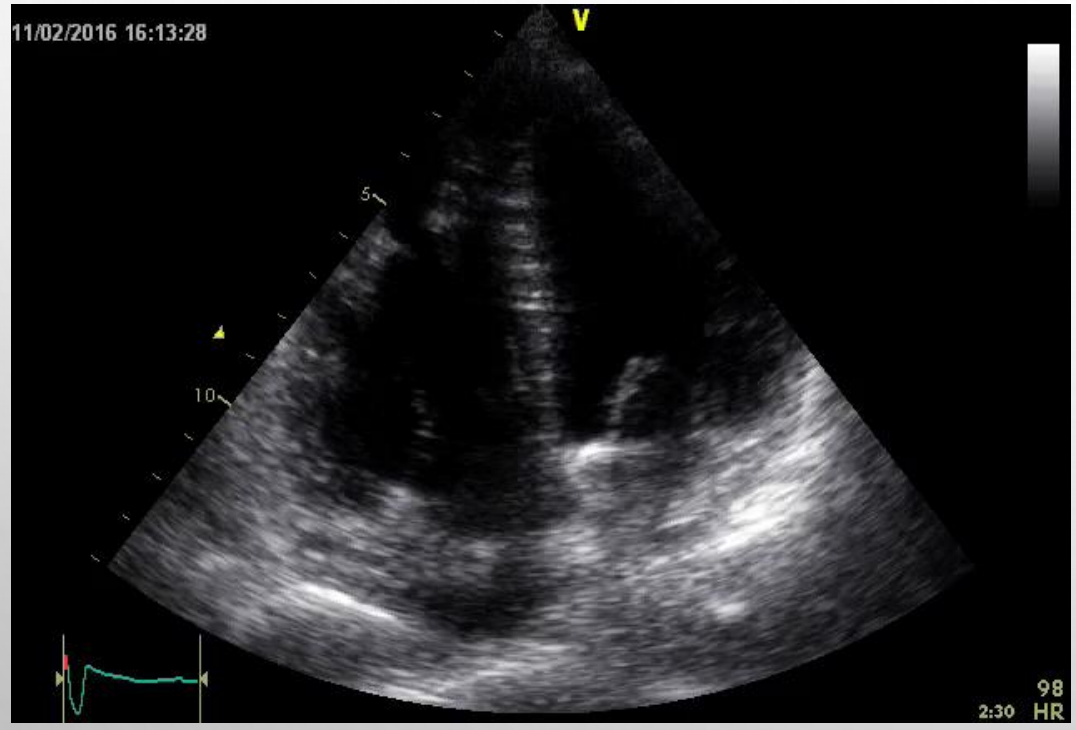
PRÉSIDENT DE LA SOCIÉTÉ LUXEMBOURGEOISE DE PNEUMOLOGIE

PNEUMOLOGUE CENTRE HOSPITALIER DU LUXEMBOURG

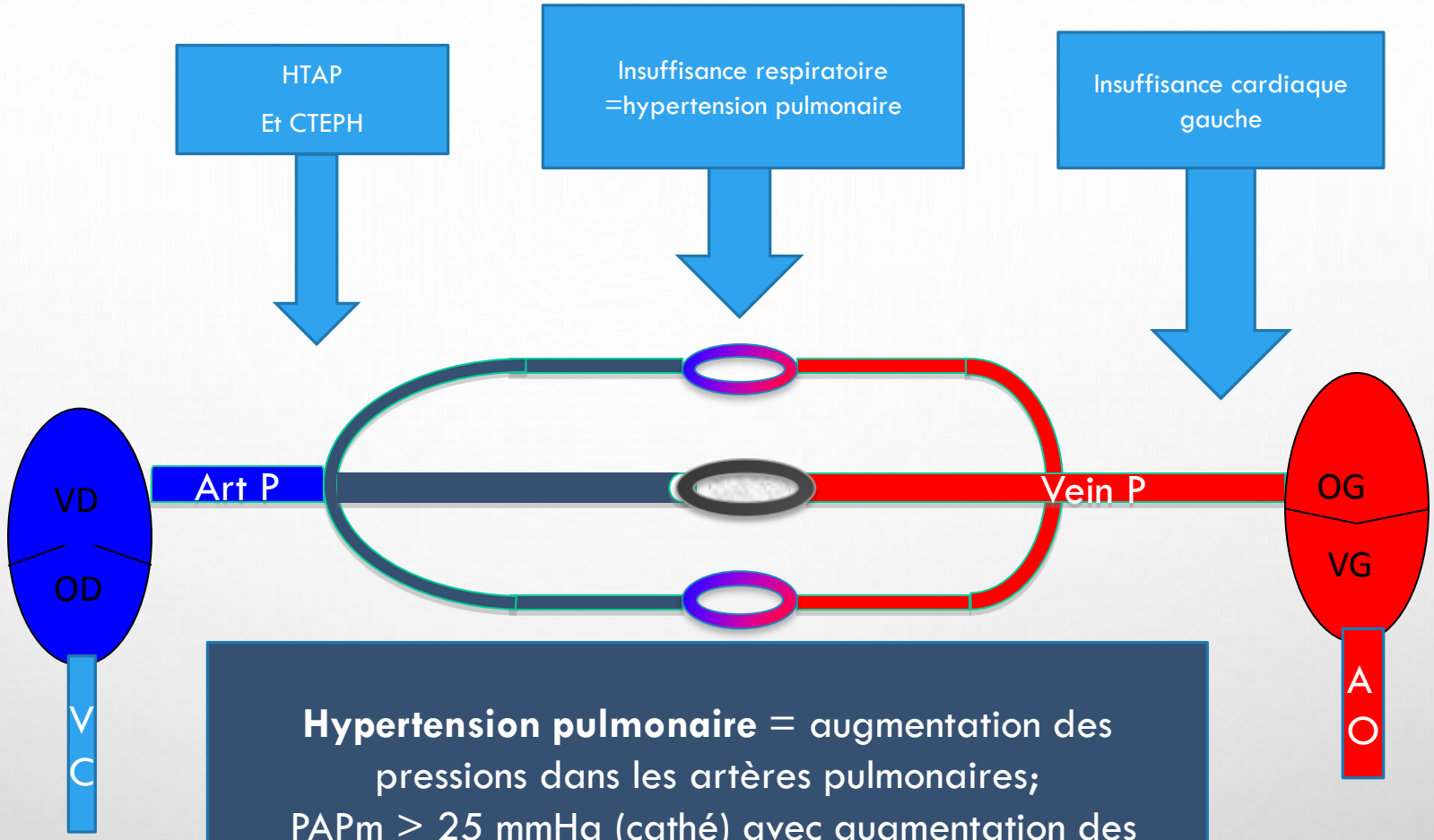
ALFORMEC
Association Luxembourgeoise pour la Formation Médicale Continue

Société Luxembourgeoise de Pneumologie

CAS CLINIQUE



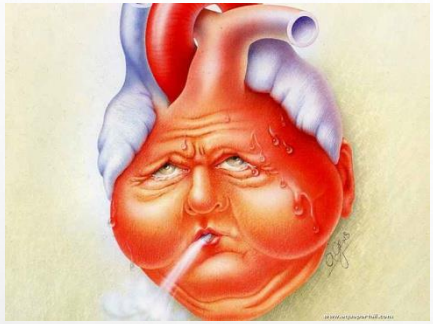
- PATIENT 52 ANS
- DYSPNÉE D'EFFORT CRESCENDO DEPUIS 2 ANS
- ATCD: HTA, TVP, PRISE D ANOREXIGÈNES, TABAGISME 35PA
- PAS DE TRAITEMENT
- ECHO COEUR: FEVG 50%, PAPS 65MMHG, DILATATION OREILLETTE DROITE



Hypertension pulmonaire = augmentation des pressions dans les artères pulmonaires;
PAPm > 25 mmHg (cathé) avec augmentation des résistances vasculaires pulmonaires

Pression dépend de la résistance dans les artérioles pulmonaire et du débit cardiaque droite

1ST STEP: HYPERTENSION PULMONAIRE POST-CAPILLAIRE = DYSFONCTION CARDIAQUE GAUCHE

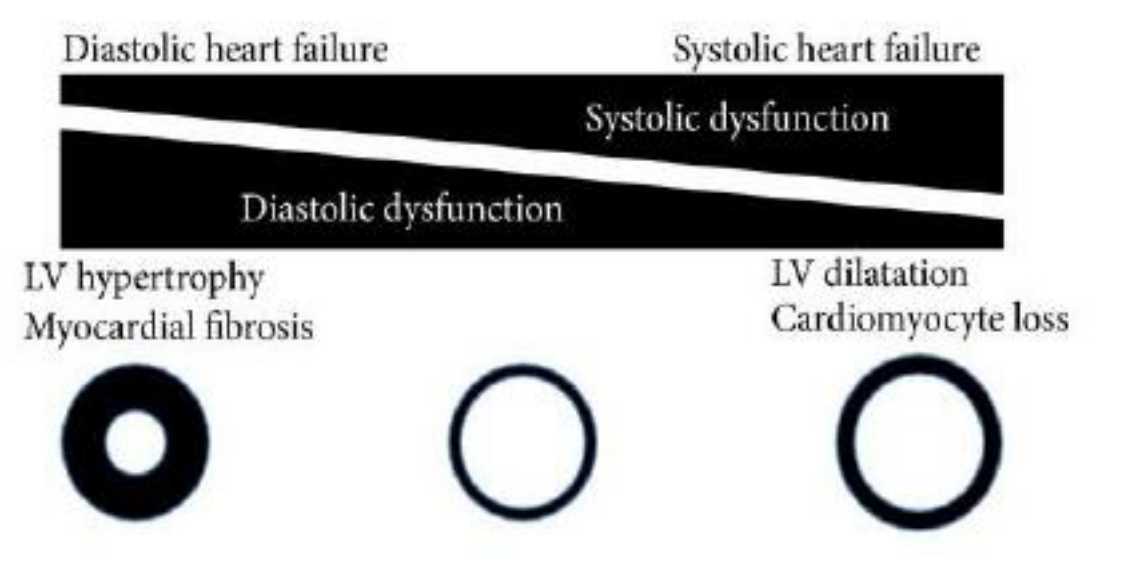


ECG+écho cœur+Rx thorax
Et discuter Holter TA, Holter ECG…

Clinical presentation	Echocardiography	Other features
Age >65 years	Structural left heart abnormality • Disease of left heart valves • LA enlargement (>4.2 cm) • Bowing of the IAS to the right • LV dysfunction • Concentric LV hypertrophy and/or increased LV mass	ECG • LVH and/or LAH • AF/Afib • LBBB • Presence of Q waves
Symptoms of left heart failure	Doppler indices of increased filling pressures • Increased E/e' • >Type 2–3 mitral flow abnormality	Other imaging • Kerley B lines • Pleural effusion • Pulmonary oedema • LA enlargement
Features of metabolic syndrome	Absence of • RV dysfunction • Mid systolic notching of the PA flow • Pericardial effusion	
History of heart disease (past or current)		
Persistent atrial fibrillation		

AF = atrial flutter; Afib = atrial fibrillation; ECG = electrocardiogram; IAS = inter-atrial septum; LA = left atrium; LAH = left atrial hypertrophy/dilatation; LBBB = left bundle branch block; LV = left ventricle; LVH = left ventricular hypertrophy; PA = pulmonary artery; RV = right ventricle.

APPROCHE THÉRAPEUTIQUE CARDIOPATHIE FEVG CONSERVÉE



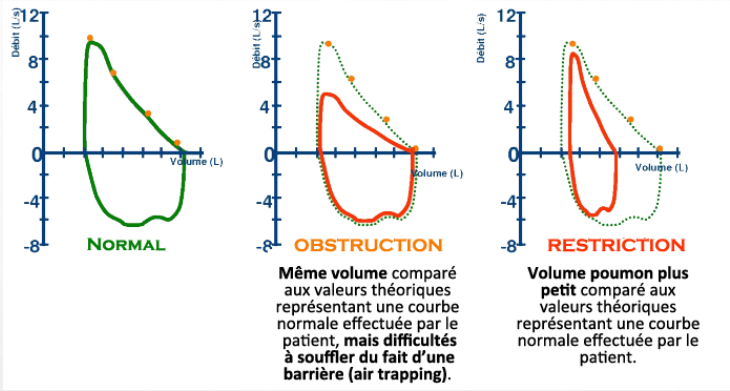
Équilibrer TA (Sartans?), diurétiques, aldactone, Traitement troubles du rythme, perte de poids, restriction sodée

Heart Failure with Preserved Ejection Fraction – Concept, Pathophysiology, Diagnosis and Challenges for Treatment

Lidija Veterovska Miljkovik[1*], Vera Spiroska[2]

2T STEP: INSUFFISANCE RESPIRATOIRE?

- EFR: MALADIE RESPIRATOIRE CHRONIQUE? BPCO? SYNDROME RESTRICTIF?

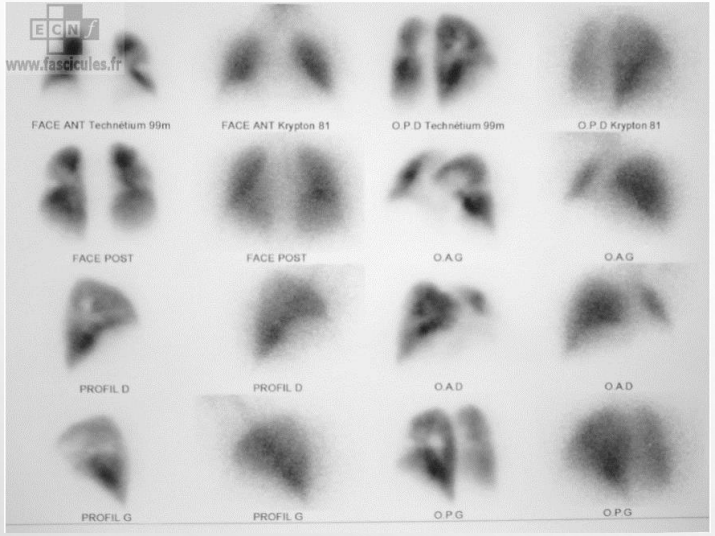


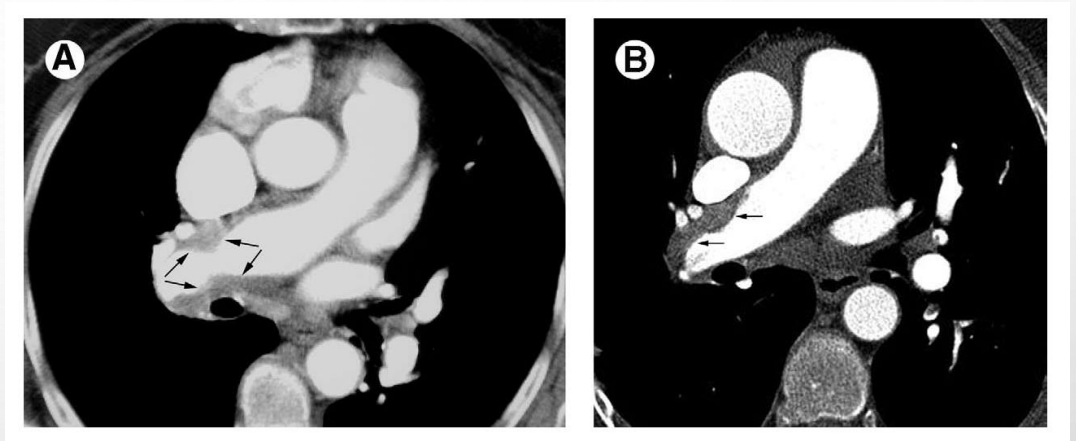
- TRANSFERT DU CO: TROUBLE DE LA DIFFUSION ISOLÉ AVEC PLÉTHYSMOGRAPHIE NORMALE EST EN FAVEUR D'UNE HTAP
- GDS: INSUFFISANCE RESPIRATOIRE?
- OXYMÉTRIE NOCTURNE:SYNDROME D'APNÉES DU SOMMEIL ?

APPROCHE THÉRAPEUTIQUE DE L'HYPERTENSION PULMONAIRE:

- TRAITEMENT DE LA MALADIE SOUS-JACENTE
- OXYGÉNOTHÉRAPIE CONTEXTE INSUFFISANCE RESPIRATOIRE SÉVÈRE
- PAS D'INDICATION TRAITEMENT SPÉCIFIQUE POUR HTAP

3 STEP: RECHERCHE HYPERTENSION PULMONAIRE THROMBOEMBOLIQUE CHRONIQUE





Anticoagulation 3mois

Persistance ???

Centre de compétence: Artériographie Pulmonaire + cathé droit

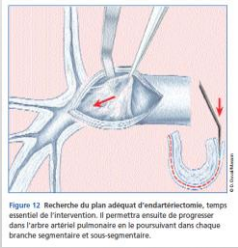


Figure 12 Recherche du plan adéquat d'endartériectomie, temps essentiel de l'intervention. Il permettra ensuite de progresser dans l'arbre artériel pulmonaire en le poursuivant dans chaque branche segmentaire et sous-segmentaire.

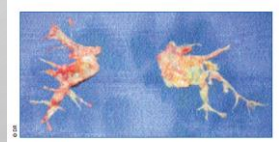
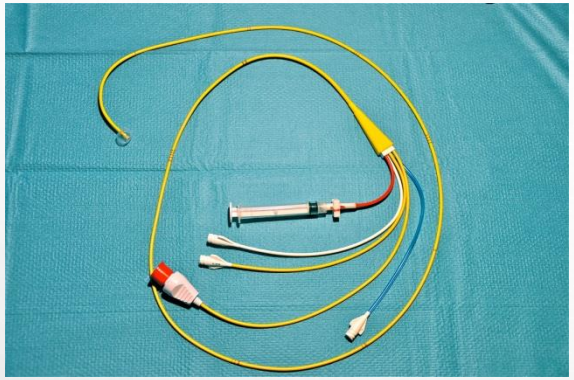
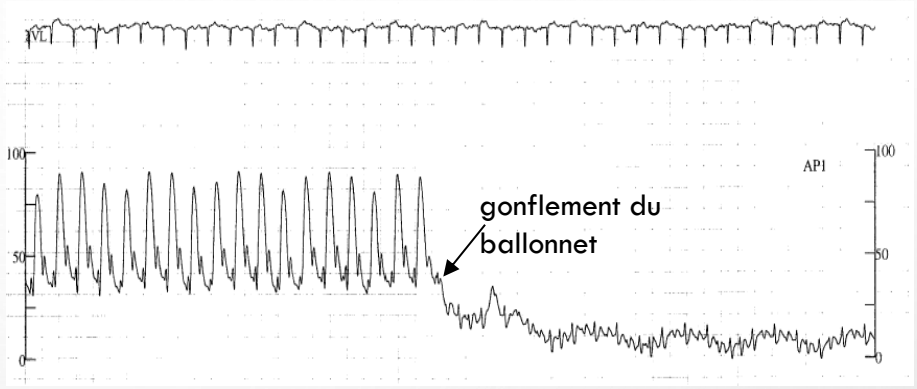


Figure 13b Séquestre témoignant d'une désobstruction très distale alors que les troncs des artères pulmonaires étaient reperméabilisés.

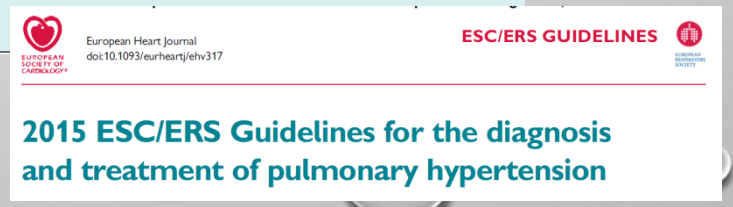
2eme ligne: Riociguat
Angioplastie artères pulmonaires

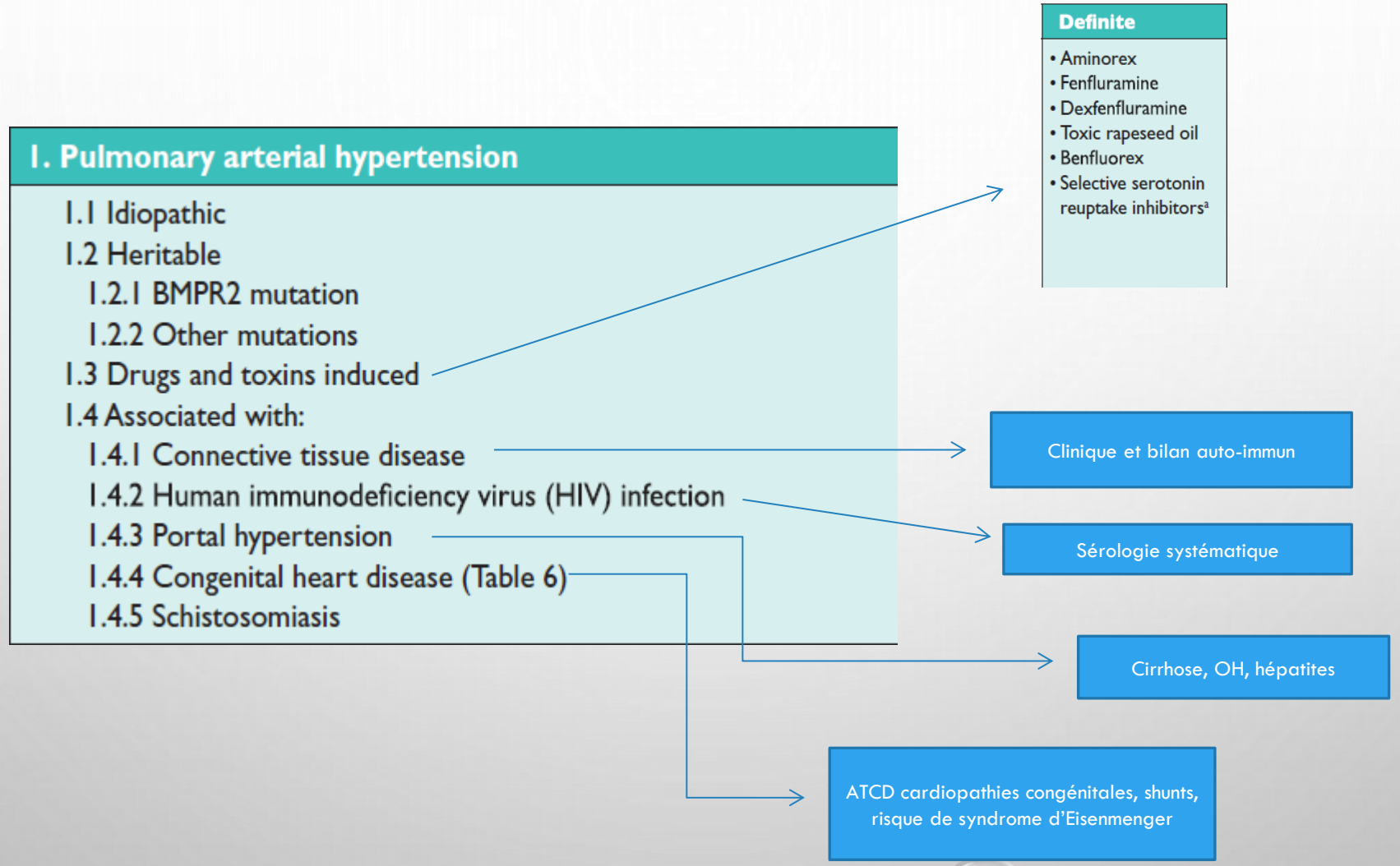
POURSUITE BILAN PAR UN CATHÉTERISME DU CŒUR DROIT



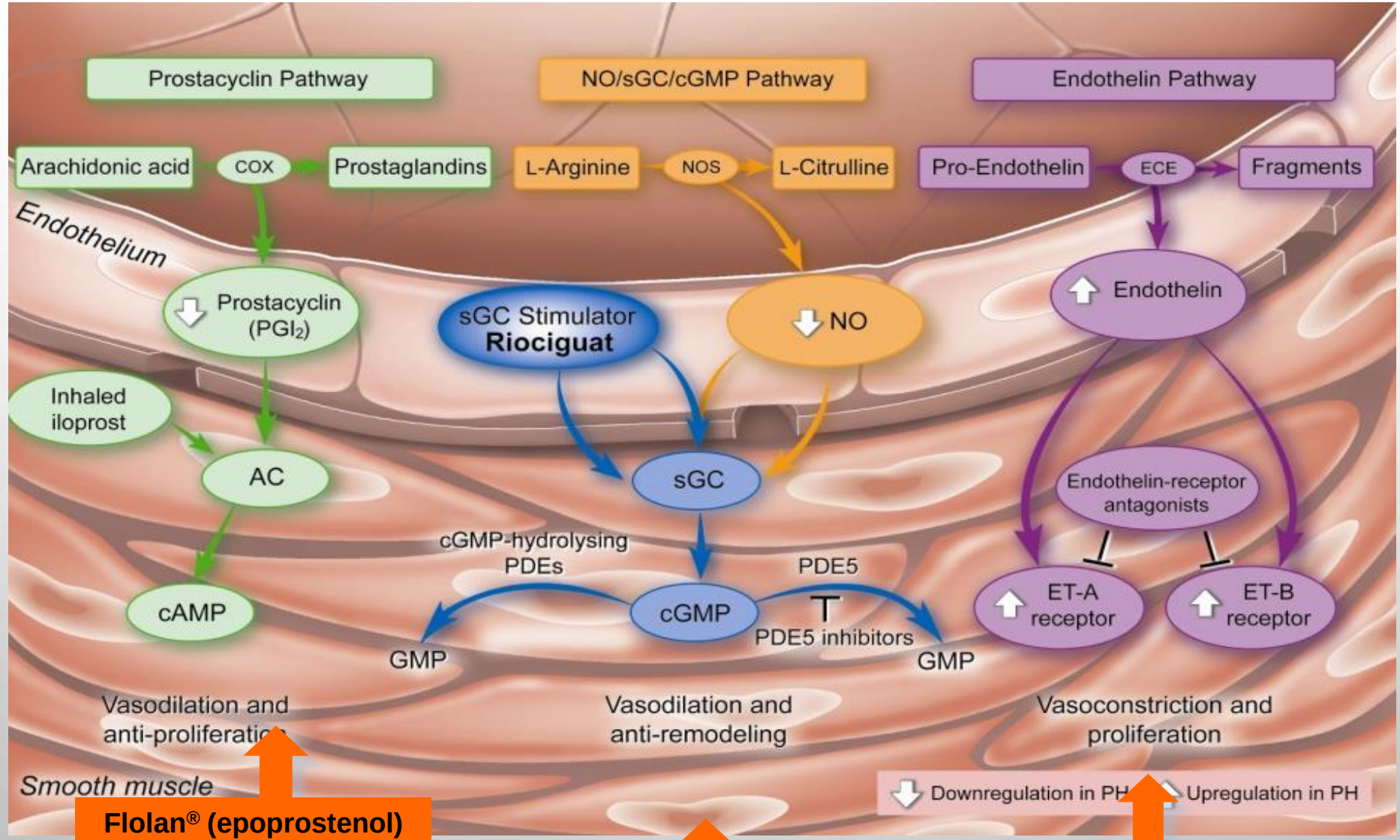


Definition	Characteristics[a]	Clinical group(s)[b]
PH	PAPm ≥25 mmHg	All
Pre-capillary PH	PAPm ≥25 mmHg PAWP ≤15 mmHg	1. Pulmonary arterial hypertension 3. PH due to lung diseases 4. Chronic thromboembolic PH 5. PH with unclear and/or multifactorial mechanisms
Post-capillary PH	PAPm ≥25 mmHg PAWP >15 mmHg	2. PH due to left heart disease 5. PH with unclear and/or multifactorial mechanisms
Isolated post-capillary PH (Ipc-PH)	DPG <7 mmHg and/or PVR ≤3 WU[c]	
Combined post-capillary and pre-capillary PH (Cpc-PH)	DPG ≥7 mmHg and/or PVR >3 WU[c]	




1. Pulmonary arterial hypertension
1.1 Idiopathic
1.2 Heritable
1.2.1 BMPR2 mutation
1.2.2 Other mutations
1.3 Drugs and toxins induced
1.4 Associated with:
1.4.1 Connective tissue disease
1.4.2 Human immunodeficiency virus (HIV) infection
1.4.3 Portal hypertension
1.4.4 Congenital heart disease (Table 6)
1.4.5 Schistosomiasis
Definite
• Aminorex
• Fenfluramine
• Dexfenfluramine
• Toxic rapeseed oil
• Benfluorex
• Selective serotonin reuptake inhibitors[a]
Clinique et bilan auto-immun
Sérologie systématique
Cirrhose, OH, hépatites
ATCD cardiopathies congénitales, shunts, risque de syndrome d'Eisenmenger

4 VOIES THÉRAPEUTIQUES

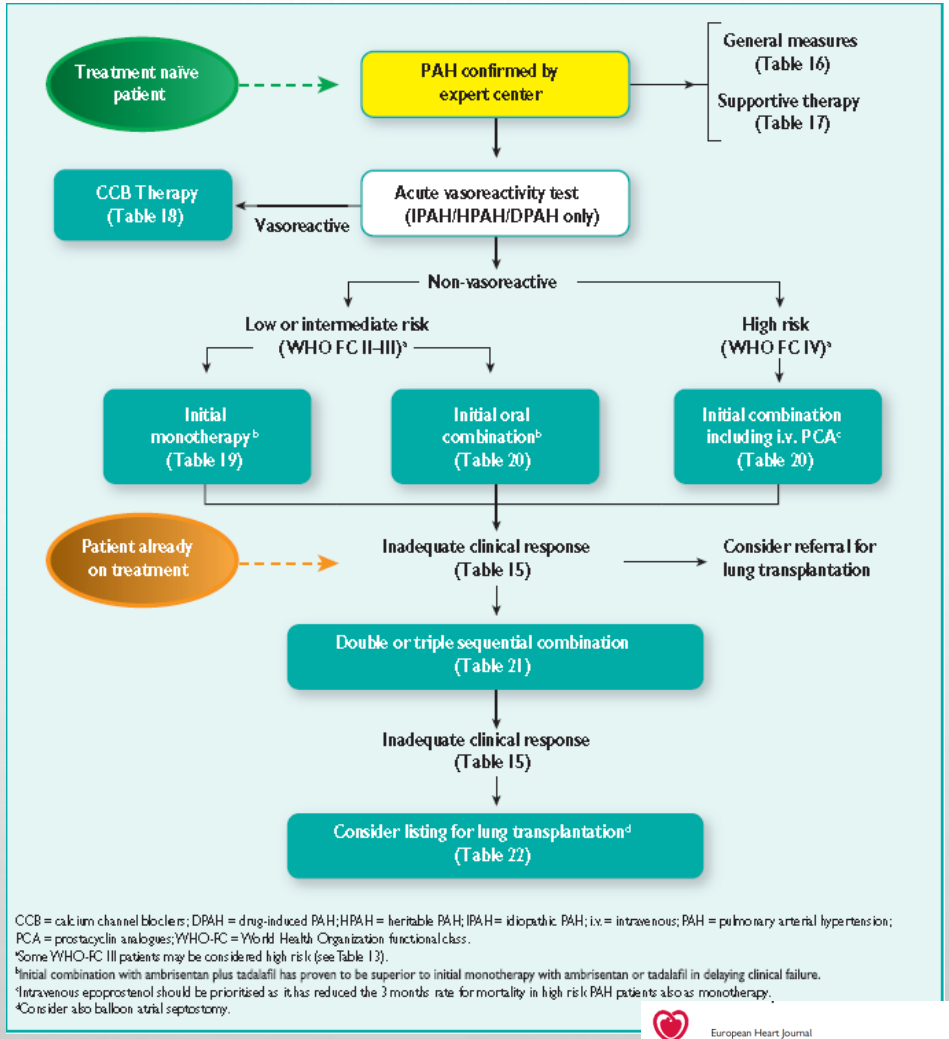


Flolan® (epoprostenol)
Remodulin (treprostenil)
Ventavis® (iloprost)
Uptravi ® (selixipag)

Adcirca® (tadalafil)
Revatio® (sildenafil)

Tracleer ® (bosentan)
Volibris® (ambrisentan)
Opsumit (macitentan)

Approche thérapeutique



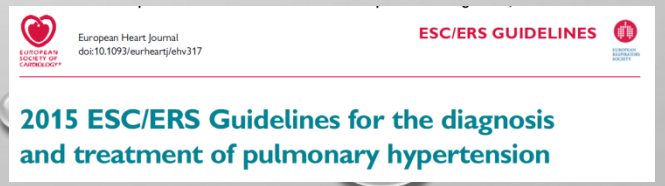
CCB = calcium channel blockers; DPAH = drug-induced PAH; HPAH = heritable PAH; IPAH = idiopathic PAH; i.v. = intravenous; PAH = pulmonary arterial hypertension; PCA = prostacyclin analogues; WHO-FC = World Health Organization functional class.
[a]Some WHO-FC III patients may be considered high risk (see Table 13).
[b]Initial combination with ambrisentan plus tadalafil has proven to be superior to initial monotherapy with ambrisentan or tadalafil in delaying clinical failure.
[c]Intravenous epoprostenol should be prioritised as it has reduced the 3 months rate for mortality in high risk PAH patients also as monotherapy.
[d]Consider also balloon atrial septostomy.

European Heart Journal
doi:10.1093/eurheartj/ehv317
ESC/ERS GUIDELINES

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

ÉVALUATION GRAVITÉ HTAP CLASSE I

Determinants of prognosis[a] (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope[b]	Repeated syncope[c]
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
Cardiopulmonary exercise testing	Peak VO_2 >15 ml/min/kg (>65% pred.) VE/VCO_2 slope <36	Peak VO_2 11–15 ml/min/kg (35–65% pred.) VE/VCO_2 slope 36–44.9	Peak VO_2 <11 ml/min/kg (<35% pred.) VE/VCO_2 ≥45
NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/ml	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
Imaging (echocardiography, CMR imaging)	RA area <18 cm^2 No pericardial effusion	RA area 18–26 cm^2 No or minimal, pericardial effusion	RA area >26 cm^2 Pericardial effusion
Haemodynamics	RAP <8 mmHg CI ≥2.5 l/min/m^2 SvO_2 >65%	RAP 8–14 mmHg CI 2.0–2.4 l/min/m^2 SvO_2 60–65%	RAP >14 mmHg CI <2.0 l/min/m^2 SvO_2 <60%



TAKE HOME MESSAGES

- 1ERE ETAPE: CARDIOPATHIE DYSFONCTION DIASTOLIQUE
- 2EME ETAPE: INSUFFISANCE RESPIRATOIRE
- 3EME ETAPE: HYPERTENSION PULMONAIRE THROMBOEMBOLIQUE CHRONIQUE
- 4EME ETAPE: HTAP CLASSE I
  - CATHÉ DROIT INCONTOURNABLE
  - 4 CLASSES THÉRAPEUTIQUES: PD5I, INHIBITEURS DE L'ENDOTHELINE, RIOCIGUAT, PROSTACYCLINES
  - ESCALADE THÉRAPEUTIQUE RAPIDE, PRIVILIGÉGIER BITHÉRAPIES

Importance du bilan, prise en charge et suivi pluridisciplinaire!