Hypertension Artérielle Pulmonaire

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ESC/ERS GUIDELINES

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

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

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Définition de l'hypertension artérielle pulmonaire

1. Pulmonary hypertension (PH) is a haemodynamic and pathophysiological condition defined as an increase in mean pulmonary arterial pressure ≥25 mmHg at rest as assessed by right heart catheterization (Table 3). PH can be found in multiple clinical conditions (Table 4).
2. Pulmonary arterial hypertension (PAH, group 1) is a clinical condition characterized by the presence of pre-capillary PH (Table 3) and pulmonary vascular resistance >3 Wood units, in the absence of other causes of pre-capillary PH such as PH due to lung diseases, chronic thromboembolic PH, or other rare diseases (Table 4). PAH includes different forms that share a similar clinical picture and virtually identical pathological changes of the lung microcirculation (Table 4).
3. There is no sufficient data to support the definition of 'PH on exercise'.

Définitions hémodynamiques
Classification de l'hypertension artérielle pulmonaire

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]	

Classification clinique de l'hypertension pulmonaire

1. Hypertension artérielle pulmonaire

- Idiopathiques
- Familiales
- Toxiques / Médicamenteuses
- Associées (maladies de système, VIH, **cardiopathies congénitales**…)

1'. Maladie veino-occlusive pulmonaire et/ou hémangiomatose capillaire pulmonaire

1''. Hypertension artérielle pulmonaire persistante du nouveau-né

2. Hypertension pulmonaire due une cardiopathie gauche

3. Hypertension pulmonaire due à une pathologie pulmonaire et/ou à une hypoxie

4. Hypertension pulmonaire chronique thromboembolique et autres obstructions artérielles pulmonaires

5. Hypertensions artérielles pulmonaires inexpliquées et/ou multifactorielles

Diagnostic de l'hypertension pulmonaire

- **Suspicion clinique**
- **Un ECG normal n'élimine pas le diagnostic**
- **Radiographie thoracique anormale dans 90% des cas**
- **Echocardiographie**
  - Toujours indiquée en cas de suspicion d'hypertension pulmonaire
  - Suspicion d'hypertension pulmonaire selon la vélocité de la régurgitation tricuspide et les autres constatations
  - Conséquences sur le ventricule droit
  - Recherche étiologique (cardiopathies gauches, valvulopathies, cardiopathies congénitales…)
- **Autres imageries: scintigraphie pulmonaire, scanner thoracique, IRM cardiaque…**
- **Cathétérisme cardiaque**
  - Indispensable au diagnostic
  - Test de réactivité vasculaire selon le diagnostic

Probabilité échocardiographique d'hypertension pulmonaire chez les patients symptomatiques

Peak tricuspid regurgitation velocity (m/s)	Presence of other echo 'PH signs'[a]	Echocardiographic probability of pulmonary hypertension
≤2.8 or not measurable	No	Low
≤2.8 or not measurable	Yes	Intermediate
2.9–3.4	No	
2.9–3.4	Yes	High
>3.4	Not required	

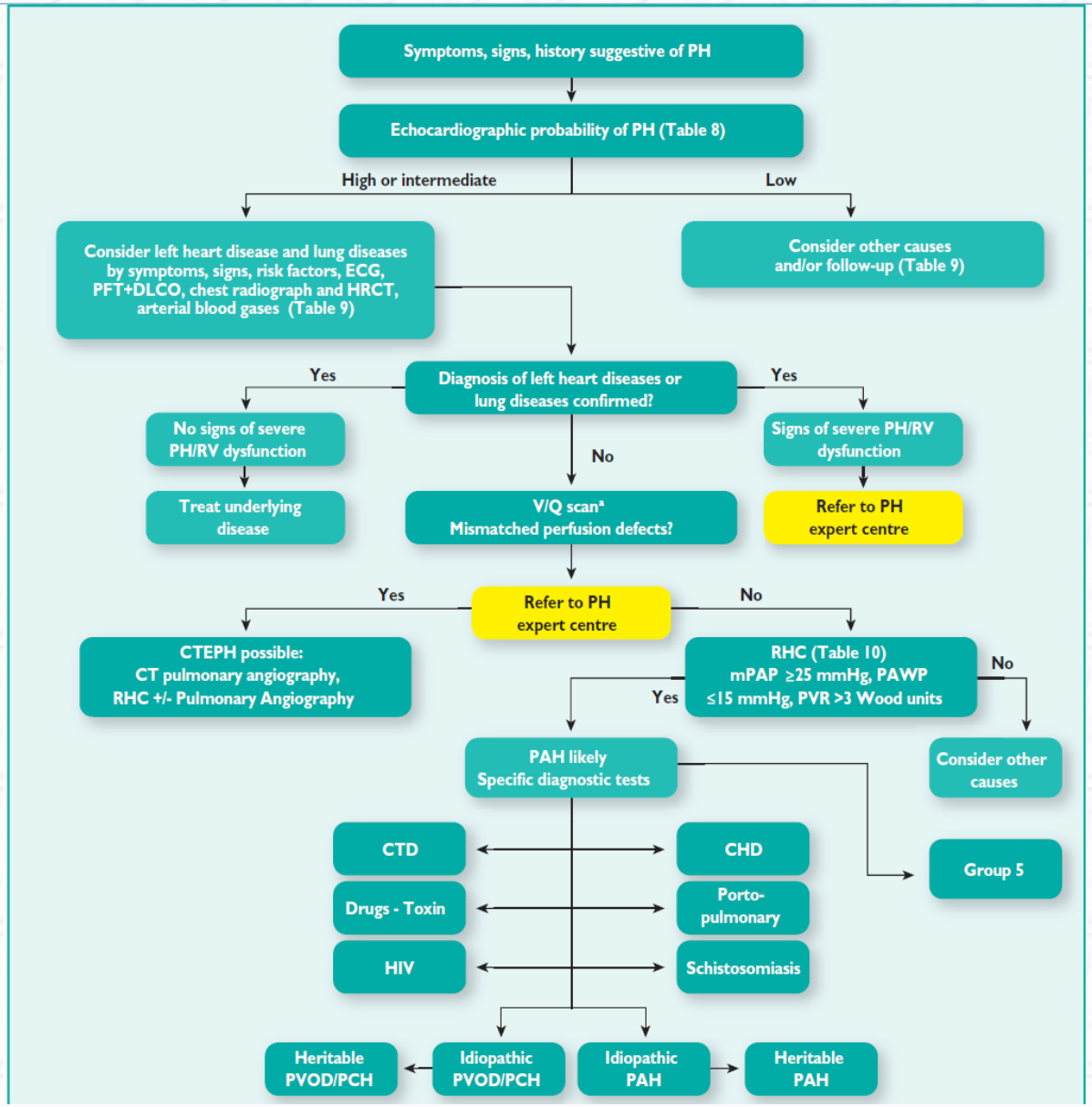
Autres signes échographiques :

A: The ventricles[a]	B: Pulmonary artery[a]	C: Inferior vena cava and right atrium[a]
Right ventricle/ left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systole) >18 cm^2
	PA diameter >25 mm.	

Diagnostic selon la probabilité d'hypertension pulmonaire chez les patients symptomatiques

Echocardiographic probability of PH	Without risk factors or associated condition for PAH or CTEPH[d]	Class[a]	Level[b]	With risk factors or associated conditions for PAH or CTEPH[c]	Class[a]	Level[b]	Ref[c]
Low	Alternative diagnosis should be considered	IIa	C	Echo follow-up should be considered	IIa	C	
Intermediate	Alternative diagnosis, echo follow-up, should be considered	IIa	C	Further assessment of PH including RHC should be considered[e]	IIa	B	45, 46
	Further investigation of PH may be considered[e]	IIb					
High	Further investigation of PH (including RHC[e]) is recommended	I	C	Further investigation of PH[e] including RHC is recommended	I	C	

Algorithme diagnostique de l'hypertension pulmonaire



Recommandations pour la stratégie diagnostique

Recommendations	Class[a]	Level[b]	Ref.[c]
Echocardiography is recommended as a first-line non-invasive diagnostic investigation in case of suspicion of PH	I	C	
Ventilation/perfusion or perfusion lung scan is recommended in patients with unexplained PH to exclude CTEPH	I	C	47
Contrast CT angiography of the PA is recommended in the workup of patients with CTEPH	I	C	93
Routine biochemistry, haematology, immunology, HIV testing and thyroid function tests are recommended in all patients with PAH to identify the specific associated condition	I	C	

Recommandations pour la stratégie diagnostique

Abdominal ultrasound is recommended for the screening of portal hypertension	I	C	67
Lung function test with DLCO is recommended in the initial evaluation of patients with PH	I	C	36
High-resolution CT should be considered in all patients with PH	IIa	C	94
Pulmonary angiography should be considered in the workup of patients with CTEPH	IIa	C	
Open or thoracoscopic lung biopsy is not recommended in patients with PAH	III	C	

Hypertension artérielle pulmonaire et cardiopathies congénitales

- **Pathologies hétérogènes**
- **5-10% des cardiopathies congénitales**
- **Présentation clinique**
  - Dyspnée, fatigue, syncope
  - Signes d'insuffisance cardiaque droite
  - Eisenmenger: cyanose
- **Echocardiographie**
  - Suspicion d'hypertension pulmonaire
  - Diagnostic de cardiopathie congénitale. Intérêt du scanner en cas de doute.
- **Cathétérisme cardiaque droit**
  - Evaluation de la PAP, résistances artérielles pulmonaires et systémiques, QP/QS
  - Pas d'indication d'évaluation de la réactivité vasculaire

Classification des hypertensions artérielles pulmonaires associées aux cardiopathies congénitales

1. Eisenmenger's syndrome

Includes all large intra- and extra-cardiac defects which begin as systemic-to-pulmonary shunts and progress with time to severe elevation of PVR and to reversal (pulmonary-to-systemic) or bidirectional shunting; cyanosis, secondary erythrocytosis, and multiple organ involvement are usually present.

2. PAH associated with prevalent systemic-to-pulmonary shunts

- Correctable[a]
- Non-correctable

Includes moderate to large defects; PVR is mildly to moderately increased, systemic-to-pulmonary shunting is still prevalent, whereas cyanosis at rest is not a feature.

3. PAH with small/coincidental defects[b]

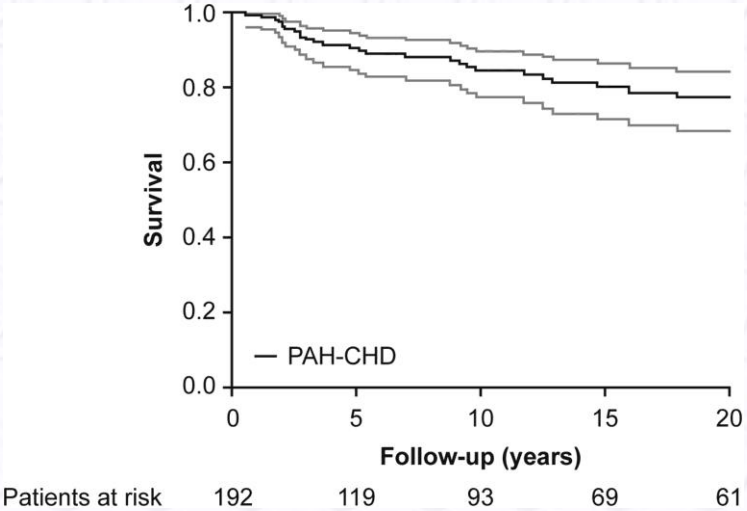
Marked elevation in PVR in the presence of small cardiac defects (usually ventricular septal defects <1 cm and atrial septal defects <2 cm of effective diameter assessed by echo), which themselves do not account for the development of elevated PVR; the clinical picture is very similar to idiopathic PAH. Closing the defects is contra-indicated.

4. PAH after defect correction

Congenital heart disease is repaired, but PAH either persists immediately after correction or recurs/develops months or years after correction in the absence of significant postoperative haemodynamic lesions.

Pronostic des hypertensions artérielles pulmonaires associées aux cardiopathies congénitales

- **192 pts avec cardiopathie congénitale et HTAP**
  - 90 syndromes d'Eisenmenger
  - 48 shunts systémiques-pulmonaires
  - 10 petites communications + HTAP
  - 44 HTAP après fermeture d'une communication
- **78% traités par des médicaments spécifiques à l'HTAP**
- **Survie tardive**
  - 85% à 10 ans
  - 77% à 20 ans



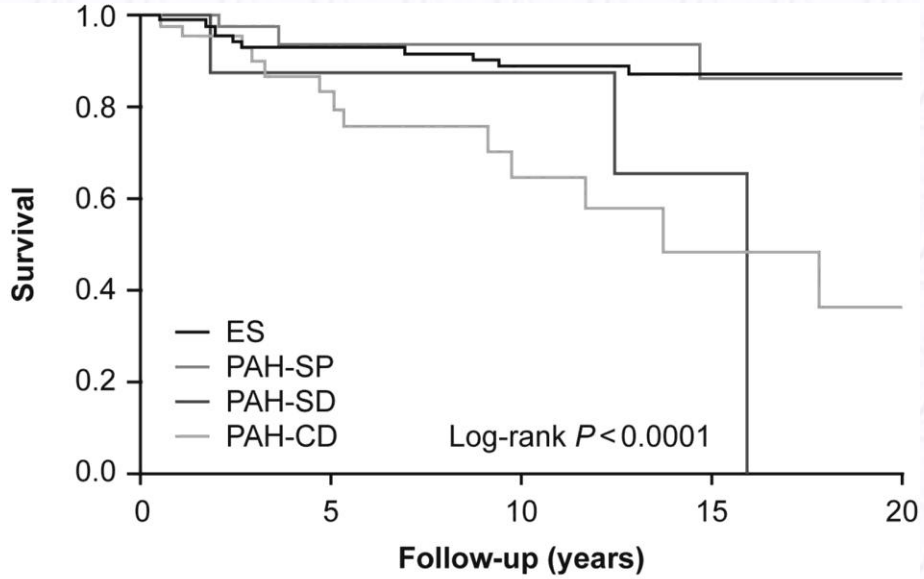
- **Groupe témoin de 278 pts avec HTAP idiopathique**
  - Survie: 46% à 10 ans, 38% à 15 ans

(Manes et al. Eur Heart J 2014;35:716–24)

Pronostic des hypertensions artérielles pulmonaires associées aux cardiopathies congénitales

Survie à 20 ans :

- Eisenmenger 87%
- Shunts syst.-pulmonaires 86%
- Petites communications *(15 ans: 66%)*
- Après fermeture 36%



Patients at risk					
ES	90	71	59	52	48
PAH-SP	48	22	18	11	10
PAH-SD	10	4	4	2	0
PAH-CD	44	22	12	4	3

(Manes et al. Eur Heart J 2014;35:716–24)

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Recommandations pour le traitement des HTAP associées aux cardiopathies congénitales

Recommendations	Class[a]	Level[b]	Ref.[c]
Bosentan is recommended in WHO-FC III patients with Eisenmenger syndrome	I	B	200,322
Other ERAs, PDE-5is and prostanoids should be considered in patients with Eisenmenger syndrome	IIa	C	223,314, 323,324
In the absence of significant haemoptysis, oral anticoagulant treatment may be considered in patients with PA thrombosis or signs of heart failure	IIb	C	
The use of supplemental O_2 therapy should be considered in cases in which it produces a consistent increase in arterial O_2 saturation and reduces symptoms	IIa	C	179

Recommandations pour le traitement des HTAP associées aux cardiopathies congénitales

If symptoms of hyperviscosity are present, phlebotomy with isovolumic replacement should be considered, usually when the haematocrit is >65%	IIa	C	183
The use of supplemental iron treatment may be considered in patients with low ferritin plasma levels	IIb	C	184
Combination drug therapy may be considered in patients with Eisenmenger syndrome	IIb	C	207,314
The use of CCBs is not recommended in patients with Eisenmenger syndrome	III	C	189

Recommandations pour la correction des cardiopathies congénitales avec shunt systémique-pulmonaire

Recommendations			Class[a]	Level[b]	Ref.[c]
PVRi (WU • m^2)	PVR (WU)	Correctable[d]			
<4	<2.3	Yes	IIa	C	317
>8	>4.6	No	IIa	C	317
4–8	2.3–4.6	Individual patient evaluation in tertiary centres	IIa	C	317

Conclusion

- **Hétérogeneïté de l'hypertension pulmonaire**
- **Nécessité :**
  - D'un diagnostic précis basé sur le cathétérisme cardiaque
  - De l'analyse du contexte clinique et de l'imagerie pour une classification appropriée (type, étiologie)
  - Du recours à l'algorithme diagnostique
- **HTAP associés aux cardiopathies congénitales**
  - Hétérogeneïté
  - Diagnostic de la cardiopathie congénitale sous-jacente
    - Différences de pronostic
    - Impact thérapeutique
    - Indications limitées de fermeture des communications, seulement chez des patients sélectionnés avec des shunts systémiques-pulmonaires prédominants
  - Meilleur pronostic du syndrome d'Eisenmenger que de l'HTAP idiopathique