Pulmonary Hypertension with left heart disease

مؤتمر رابطة طب وجراحة الصدر طرطوس ٢٠٢٢
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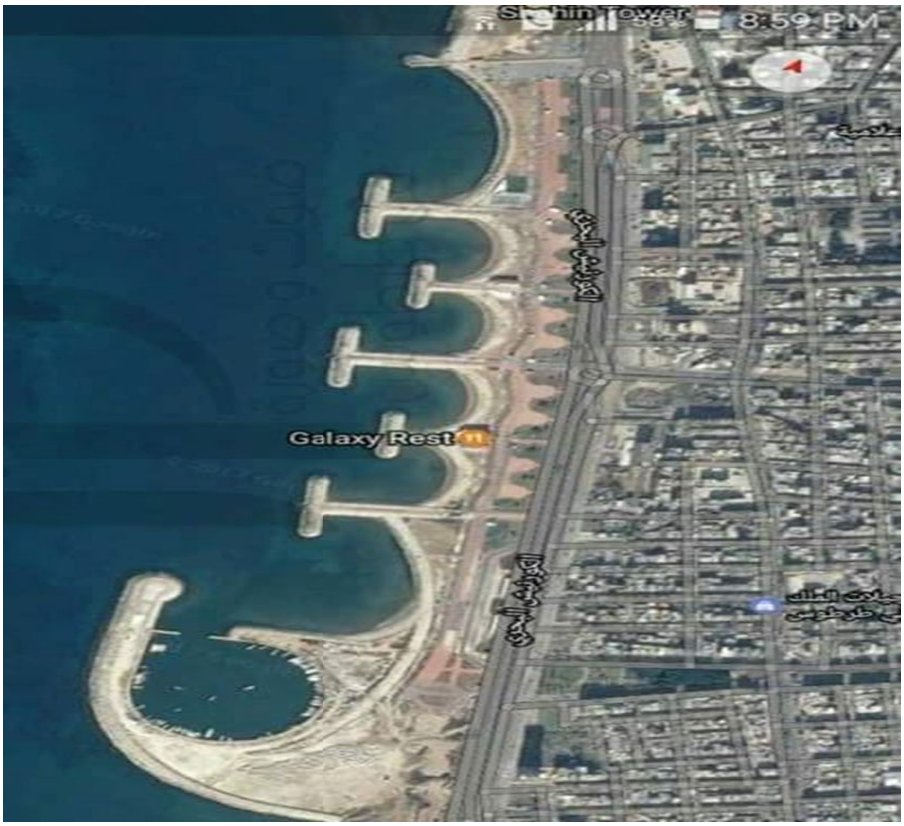
Pulmonary Hypertension with left heart disease

Causes & Diagnosis


Galaxy Rest

Definition

- PH is defined by mPAP >20mmHg
- Haemodynamic assessment by RHC
- mPAP = 2dPAP+sPAP / 3 mm Hg(RHC)
- m PAP=0.65*PASP +0.55 mm Hg (ECHO)
- PH is caused by left heart disease or parenchymal lung

Haemodynamic definitions of PH

Definition	Haemodynamic characteristics
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20 mmHg PAWP ≤15 mmHg PVR >2 WU
IpcPH	mPAP >20 mmHg PAWP >15 mmHg PVR ≤2 WU
CpcPH	mPAP >20 mmHg PAWP >15 mmHg PVR >2 WU
Exercise PH	mPAP/CO slope between rest and exercise >3 mmHg/L/min

Clinical classification

GROUP 1 Pulmonary arterial hypertension (PAH)

- 1.1 Idiopathic
  - 1.1.1 Non-responders at vasoreactivity testing
  - 1.1.2 Acute responders at vasoreactivity testing
- 1.2 Heritable[a]
- 1.3 Associated with drugs and toxins[a]
- 1.4 Associated with:
  - 1.4.1 Connective tissue disease
  - 1.4.2 HIV infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease
  - 1.4.5 Schistosomiasis
- 1.5 PAH with features of venous/capillary (PVOD/PCH) involvement
- 1.6 Persistent PH of the newborn

GROUP 2 PH associated with left heart disease

2.1 Heart failure:

2.1.1 with preserved ejection fraction

2.1.2 with reduced or mildly reduced ejection fraction[b]

2.2 Valvular heart disease

2.3 Congenital/acquired cardiovascular conditions leading to post-capillary PH

GROUP 3 PH associated with lung diseases and/or hypoxia

- 3.1 Obstructive lung disease or emphysema
- 3.2 Restrictive lung disease
- 3.3 Lung disease with mixed restrictive/obstructive pattern
- 3.4 Hypoventilation syndromes
- 3.5 Hypoxia without lung disease (e.g. high altitude)
- 3.6 Developmental lung disorders

GROUP 4 PH associated with pulmonary artery obstructions

- 4.1 Chronic thrombo-embolic PH
- 4.2 Other pulmonary artery obstructions[c]

GROUP 5 PH with unclear and/or multifactorial mechanisms

- 5.1 Haematological disorders[d]
- 5.2 Systemic disorders[e]
- 5.3 Metabolic disorders[f]
- 5.4 Chronic renal failure with or without haemodialysis
- 5.5 Pulmonary tumour thrombotic microangiopathy
- 5.6 Fibrosing mediastinitis

Overlap Between Hemodynamic and Clinical Classification of PH

DEFINITIONS	CHARACTERISTICS	PH CLINICAL GROUPS
Pre-capillary PH	mPAP >20 mm Hg PAWP ≤15 mm Hg PVR ≥3WU	PAH Lung disease Sleep-disordered breathing Miscellaneous causes
Isolated post-capillary PH (IpcPH)	mPAP >20 mm Hg PAWP >15 mm Hg PVR <3 WU	Left heart disease Miscellaneous causes
Combined pre- and post-capillary PH (CpcPH)	mPAP >20 mm Hg PAWP >15 mm Hg PVR ≥3 WU	Left heart disease Miscellaneous causes

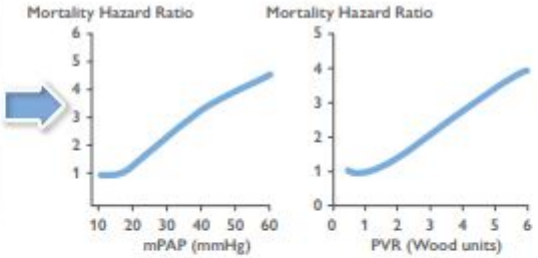
Pulmonary hypertension

Prevalence

1% Global population

Pulmonary congestion in post-capillary PH

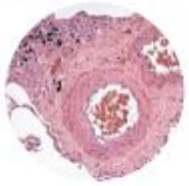
Pulmonary vascular disease / obstruction in pre-capillary PH



Right heart failure

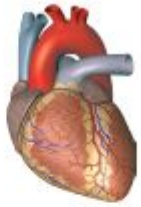
Clinical Classification

Pulmonary arterial hypertension (PAH)



- Idiopathic/heritable
- Associated conditions

PH associated with left heart disease

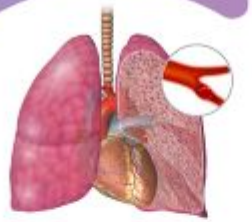


- IpcPH
- CpcPH

PH associated with lung disease

- Non-severe PH
- Severe PH

PH associated with pulmonary artery obstructions



- CTEPH
- Other pulmonary obstructions

PH with unclear and/or multifactorial mechanisms

?

- Haematologic disorders
- Systemic disorders

Prevalence

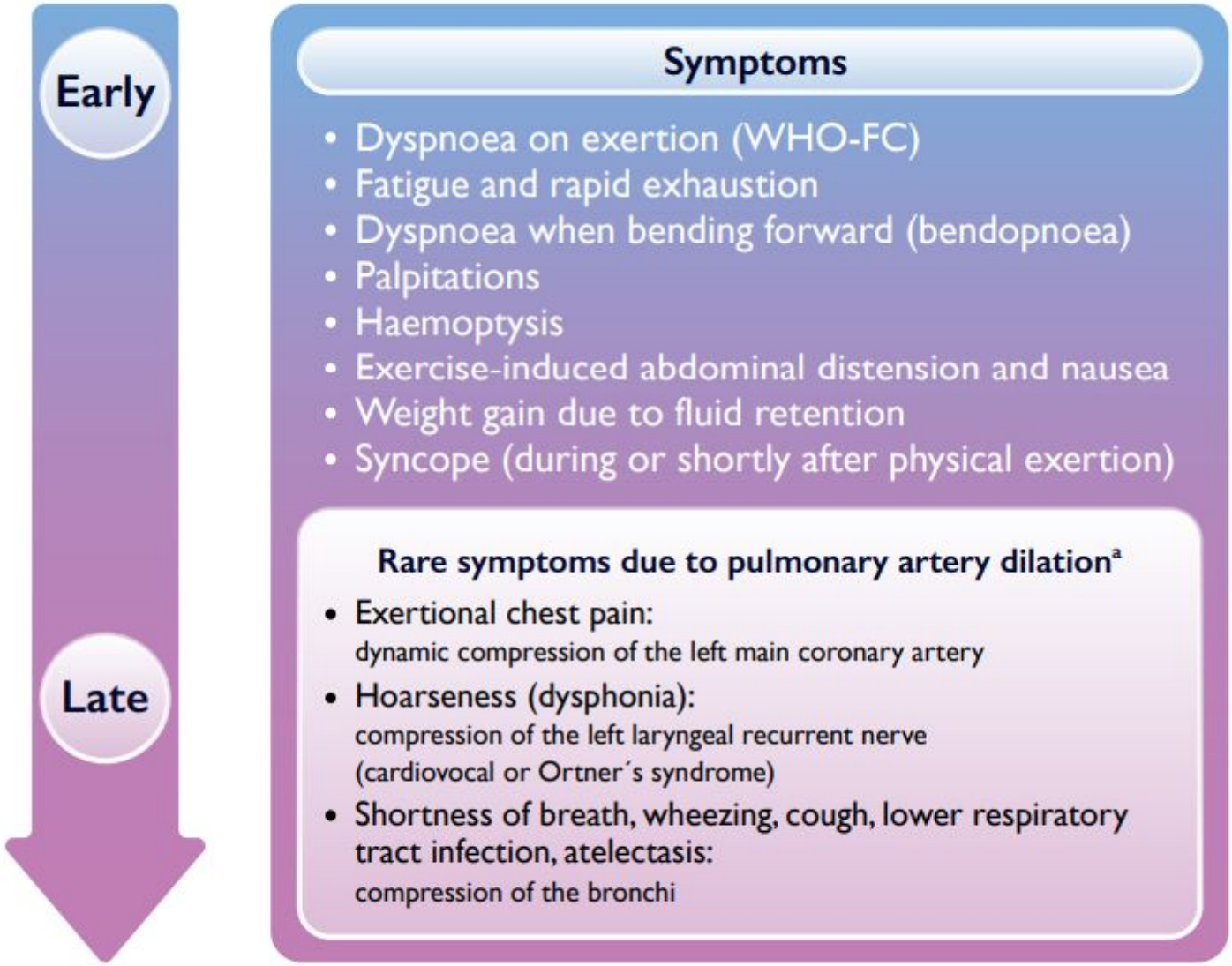
Rare	Very common	Common	Rare	Rare

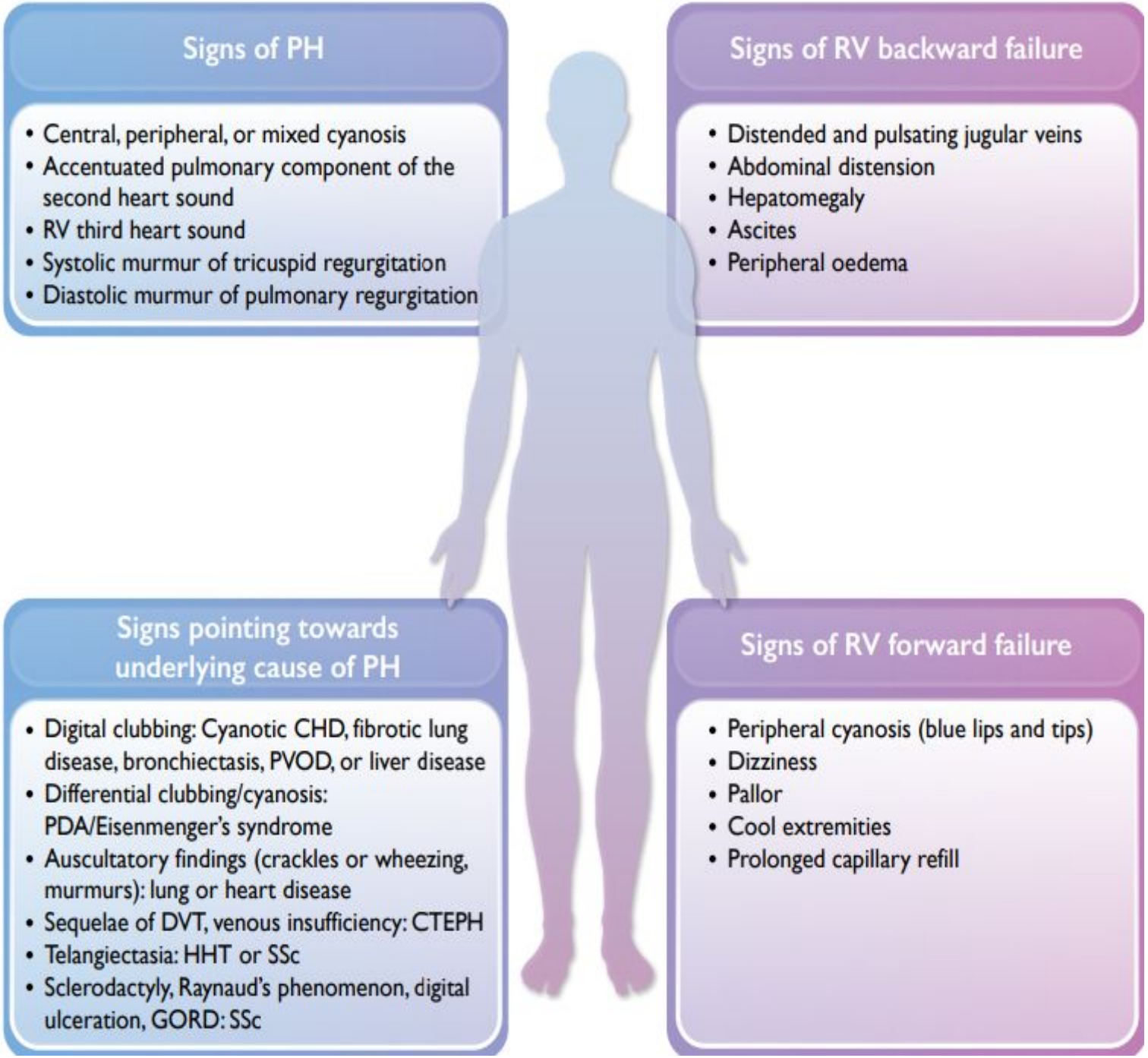
PH Diagnosis

PH Diagnosis

- Clinical presentation
- ECG
- C x ray
- Echocardiography
- Pulmonary functions test & ABG
- Ventilation \perfusion lung scan
- Contrast \non con CT scan digital subtraction angiography
- Cardiac MRI
- Blood tests and immunology
- Abdominal ultrasound
- Cardiopulmonary exercise testing
- Right heart catheterization, vasoreactivity, exercise, and fluid challenge

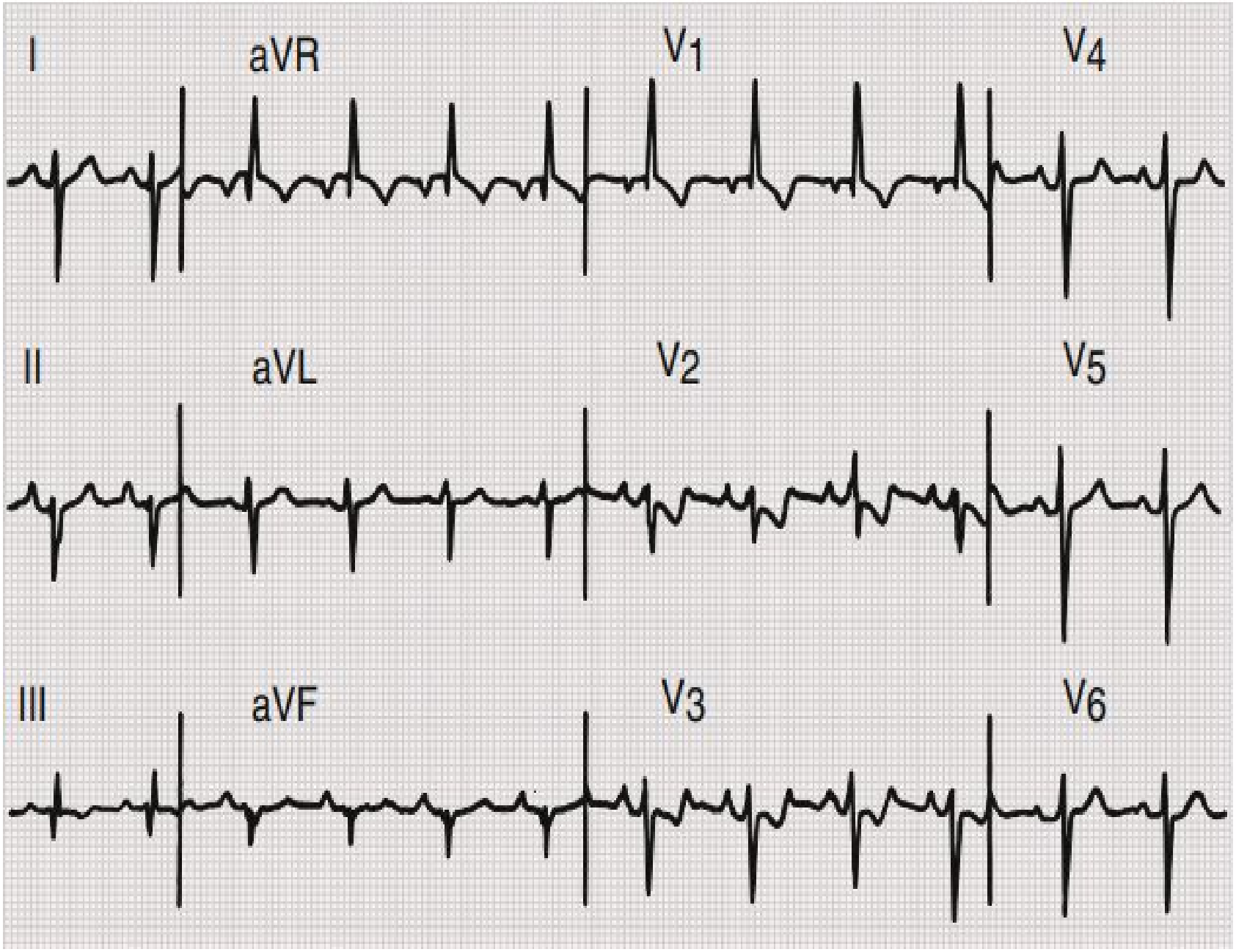
Clinical symptoms




Signs of PH
• Central, peripheral, or mixed cyanosis
• Accentuated pulmonary component of the second heart sound
• RV third heart sound
• Systolic murmur of tricuspid regurgitation
• Diastolic murmur of pulmonary regurgitation
Signs of RV backward failure
• Distended and pulsating jugular veins
• Abdominal distension
• Hepatomegaly
• Ascites
• Peripheral oedema
Signs pointing towards underlying cause of PH
• Digital clubbing: Cyanotic CHD, fibrotic lung disease, bronchiectasis, PVOD, or liver disease
• Differential clubbing/cyanosis: PDA/Eisenmenger's syndrome
• Auscultatory findings (crackles or wheezing, murmurs): lung or heart disease
• Sequelae of DVT, venous insufficiency: CTEPH
• Telangiectasia: HHT or SSc
• Sclerodactyly, Raynaud's phenomenon, digital ulceration, GORD: SSc
Signs of RV forward failure
• Peripheral cyanosis (blue lips and tips)
• Dizziness
• Pallor
• Cool extremities
• Prolonged capillary refill

ECG

- P pulmonale (P >0.25 mV in lead II)
- Right or sagittal axis deviation (QRS axis >90° or indeterminable)
- RV hypertrophy (R/S >1, with R >0.5 mV in V1; R in V1 + S in lead V5 >1 mV)
- Right bundle branch block—complete or incomplete (qR or rSR patterns in V1)
- RV strain pattern[a] (ST depression/T-wave inversion in the right pre-cordial V1–4 and inferior II, III, aVF leads)
- Prolonged QTc interval (unspecific)[b]

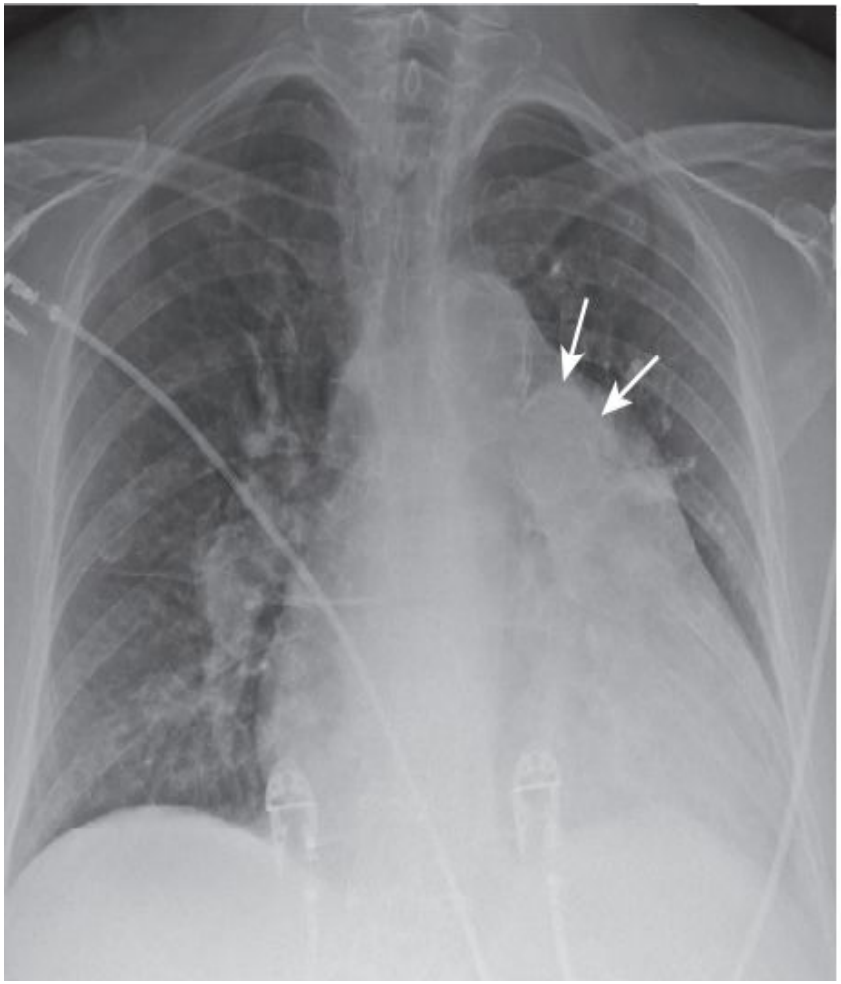

I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6

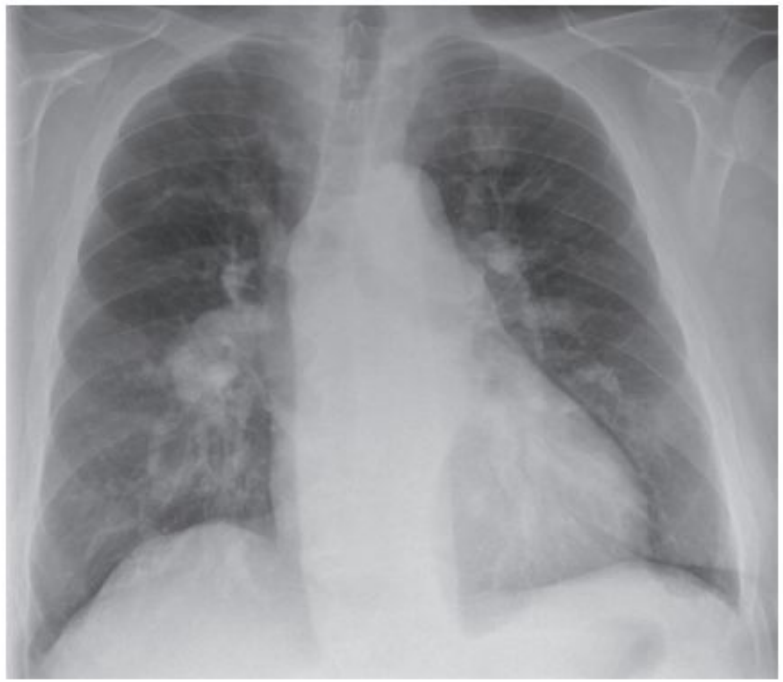
Chest roentgenogram

- The chest x- ray is abnormal in 90% of PAH
- Central pulmonary artery dilation
- Peripheral dearborization
- RA RV enlargement
- The presence of lung hyperinflation, or other features of primary lung disease,

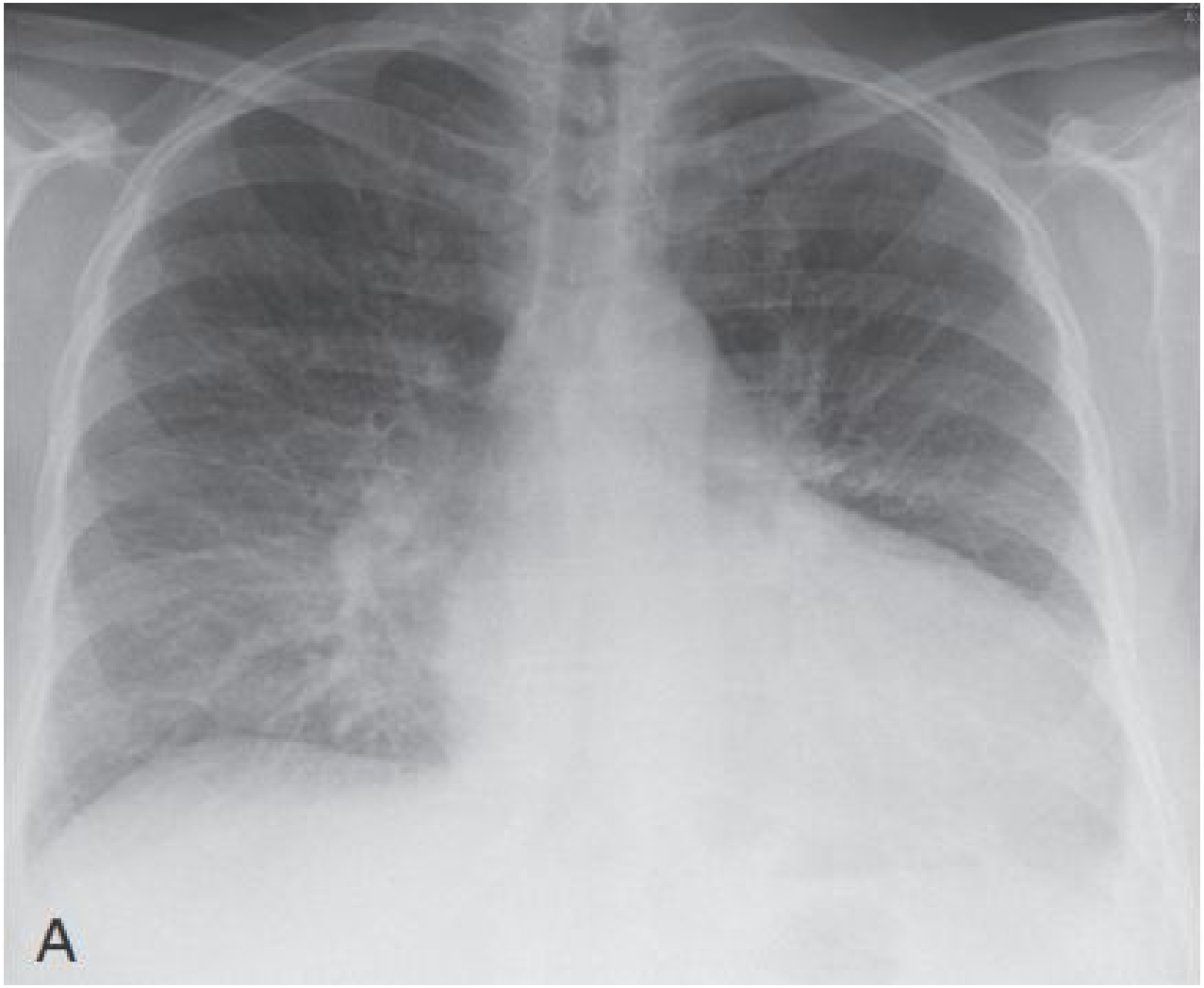
Signs of PH and concomitant abnormalities	Signs of left heart disease/ pulmonary congestion	Signs of lung disease
Right heart enlargement	Central air space opacification	Flattening of diaphragm (COPD/ emphysema)
PA enlargement (including aneurysmal dilatation)	Interlobular septal thickening 'Kerley B' lines	Hyperlucency (COPD/ emphysema)
Pruning of the peripheral vessels	Pleural effusions	Lung volume loss (fibrotic lung disease)
'Water-bottle' shape of cardiac silhouette[a]	Left atrial enlargement (including splayed carina) Left ventricular dilation	Reticular opacification (fibrotic lung disease)

Pre C PHT





Post C PHT

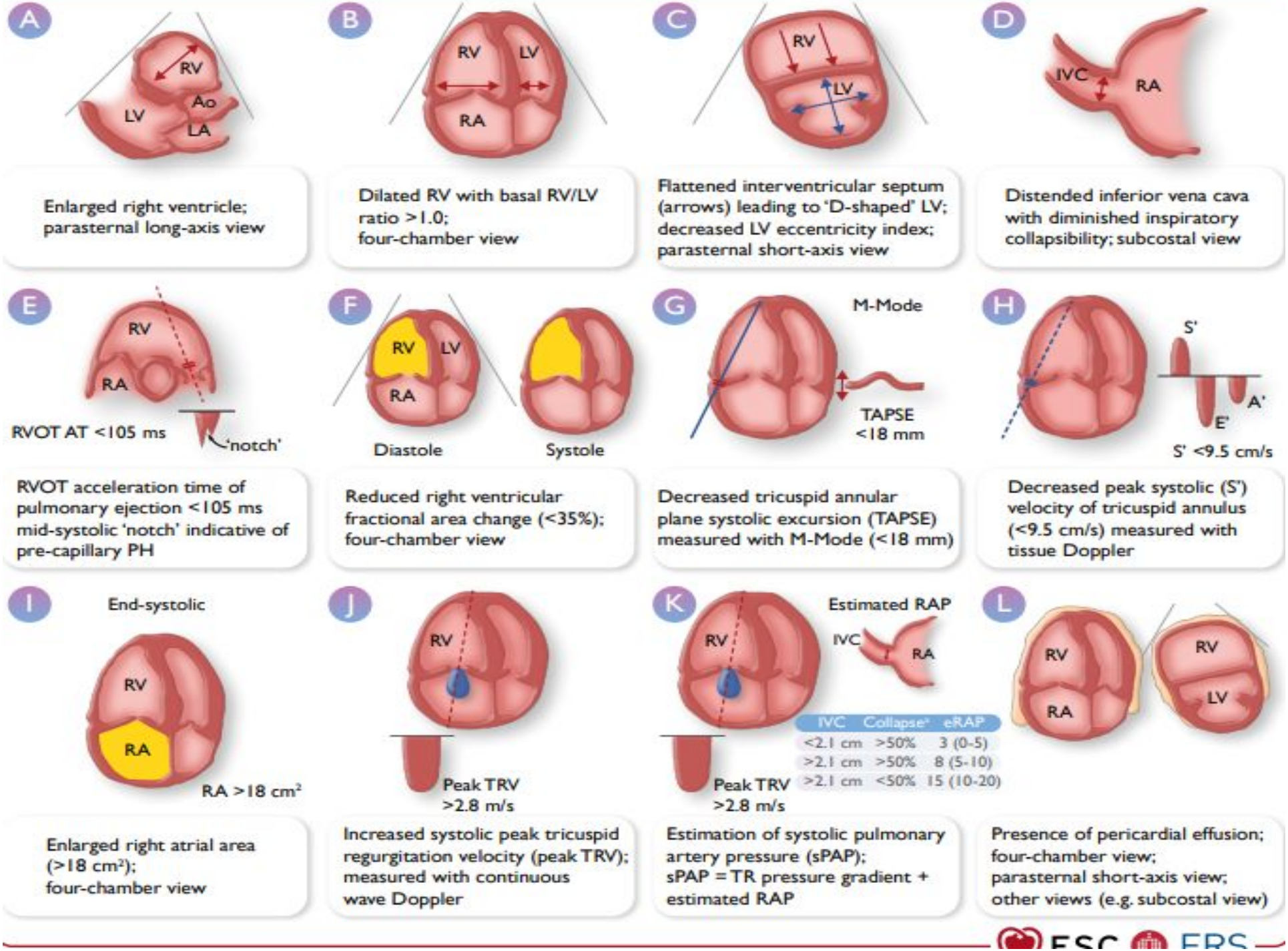


Echocardiography

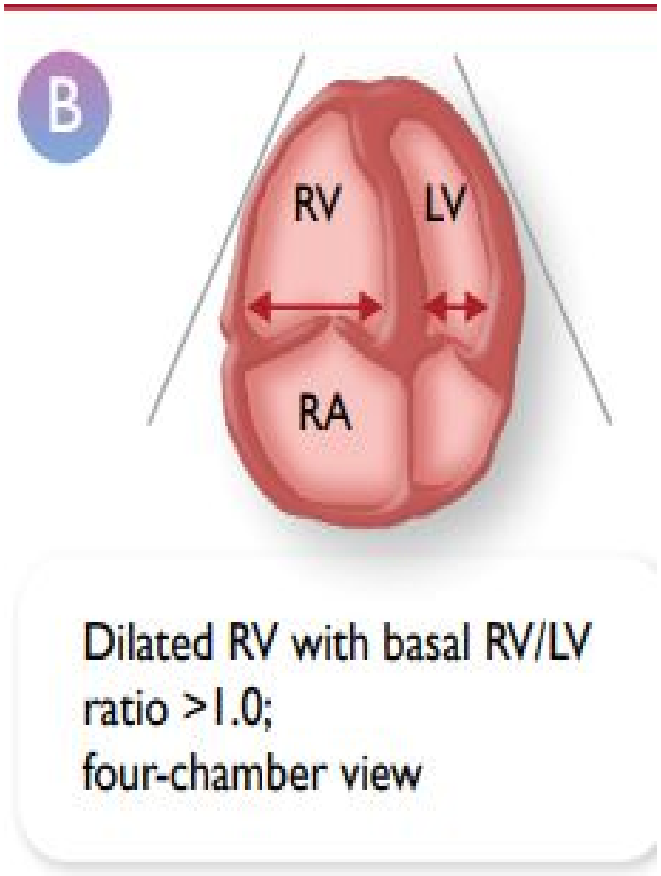
- Right and left heart morphology, RV and LV function, and valvular abnormalities,
- Gives estimates of haemodynamic parameters
- Detects the cause of suspected or confirmed PH, associated with LHD or CHD
- Agitated saline- enhanced echocardiography

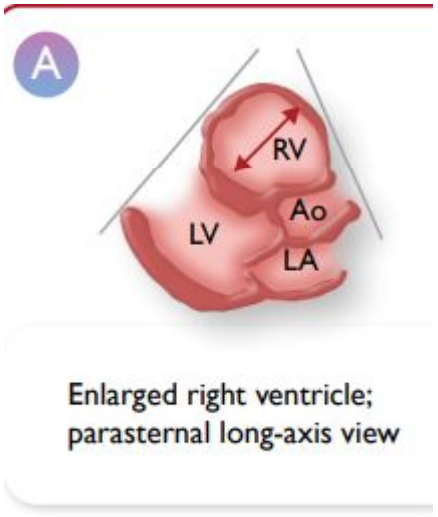
Echocardiography

- Disadvantage
- Can,t determine right atrial pressure, PVR,PAWP
- Wide chest anterior- posterior dimension
- COPD
- 1/3 PH (PASP is unmeasurable)
- Echocardiography alone is insufficient to confirm a diagnosis of PH, which requires RHC

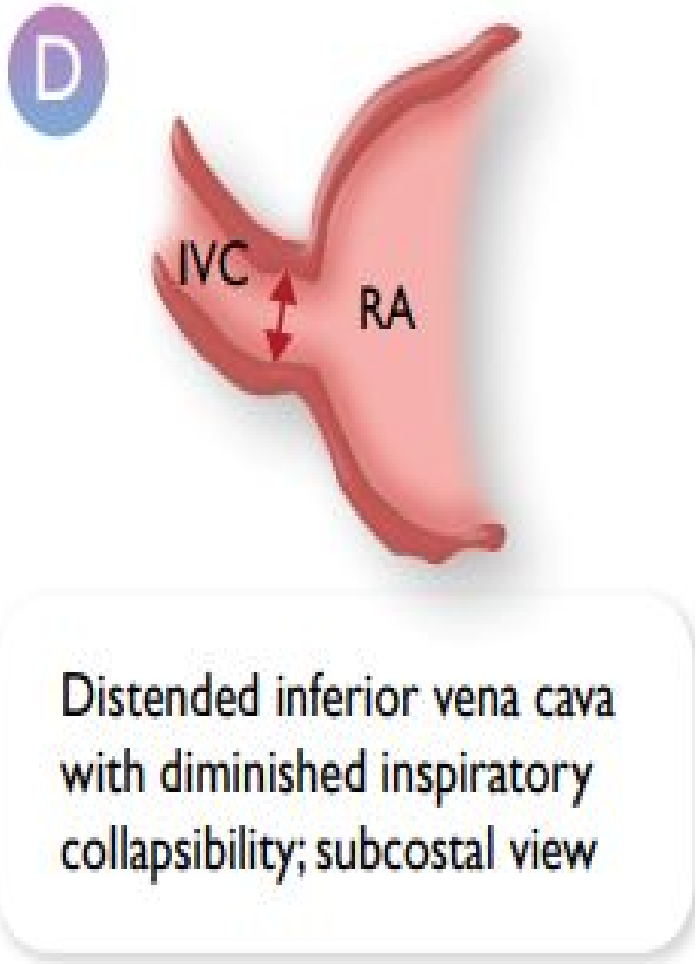

A
RV
Ao
LV
LA
Enlarged right ventricle; parasternal long-axis view
B
RV
LV
RA
Dilated RV with basal RV/LV ratio >1.0; four-chamber view
C
RV
LV
Flattened interventricular septum (arrows) leading to 'D-shaped' LV; decreased LV eccentricity index; parasternal short-axis view
D
IVC
RA
Distended inferior vena cava with diminished inspiratory collapsibility; subcostal view
E
RV
RA
RVOT AT <105 ms
'notch'
RVOT acceleration time of pulmonary ejection <105 ms mid-systolic 'notch' indicative of pre-capillary PH
F
RV
LV
RA
Diastole
Systole
Reduced right ventricular fractional area change (<35%); four-chamber view
G
M-Mode
TAPSE <18 mm
Decreased tricuspid annular plane systolic excursion (TAPSE) measured with M-Mode (<18 mm)
H
S'
A'
E'
S' <9.5 cm/s
Decreased peak systolic (S') velocity of tricuspid annulus (<9.5 cm/s) measured with tissue Doppler
I
End-systolic
RV
RA
RA >18 cm²
Enlarged right atrial area (>18 cm²); four-chamber view
J
RV
Peak TRV >2.8 m/s
Increased systolic peak tricuspid regurgitation velocity (peak TRV); measured with continuous wave Doppler
K
Estimated RAP
IVC
RA
RV
Peak TRV >2.8 m/s
IVC Collapse* eRAP
<2.1 cm >50% 3 (0-5)
>2.1 cm >50% 8 (5-10)
>2.1 cm <50% 15 (10-20)
Estimation of systolic pulmonary artery pressure (sPAP); sPAP = TR pressure gradient + estimated RAP
L
RV
RA
RV
LV
Presence of pericardial effusion; four-chamber view; parasternal short-axis view; other views (e.g. subcostal view)
ESC
ERS

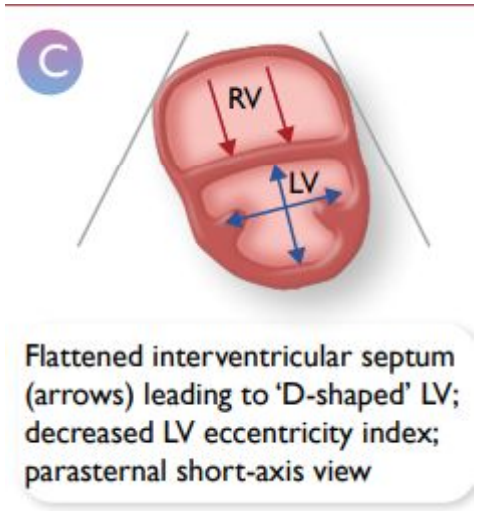
Transthoracic echocardiographic parameters



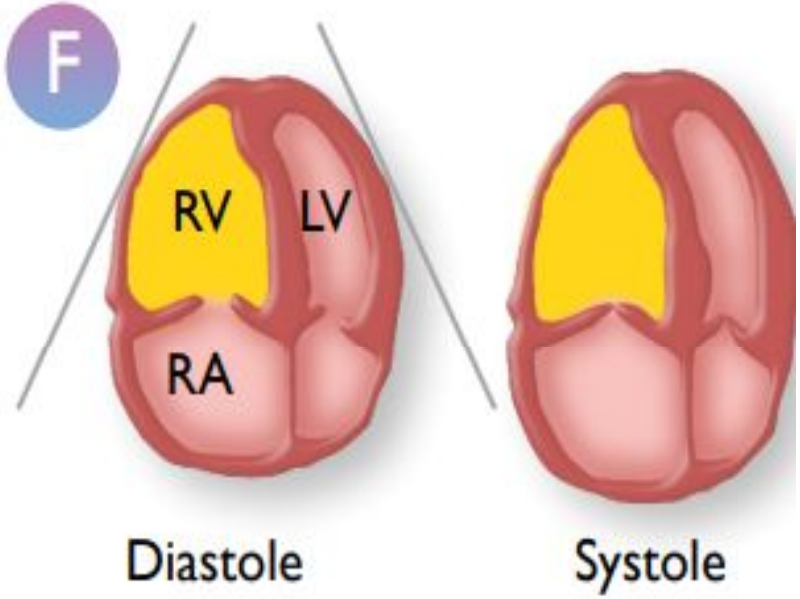


Transthoracic echocardiographic parameters

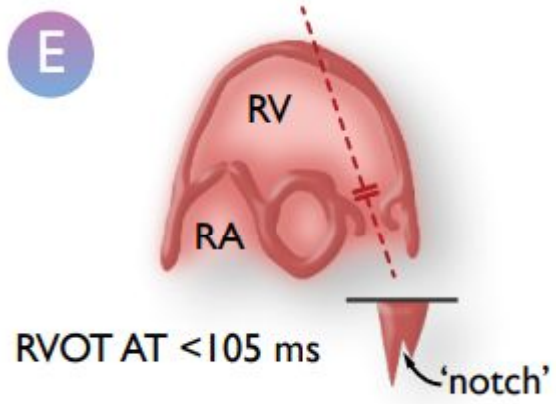




Transthoracic echocardiographic parameters



Reduced right ventricular fractional area change (<35%); four-chamber view



RVOT acceleration time of pulmonary ejection <105 ms mid-systolic 'notch' indicative of pre-capillary PH

mPAP= 79- (0.45*PAcT)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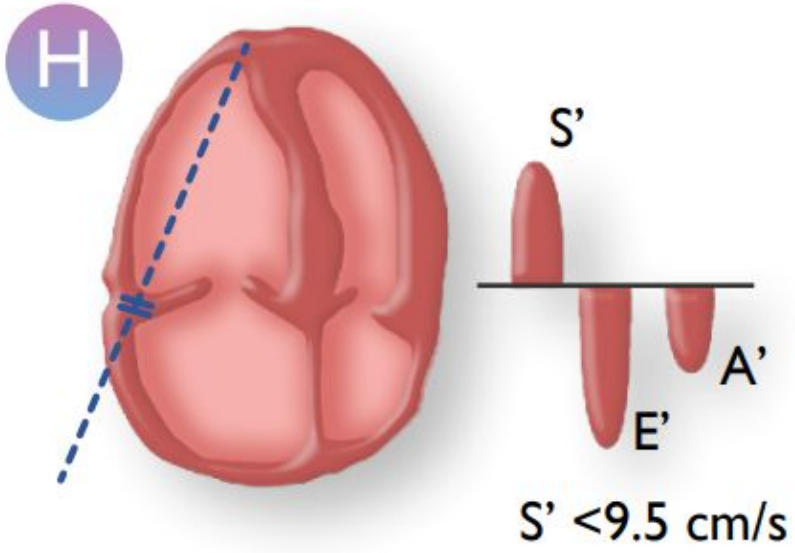
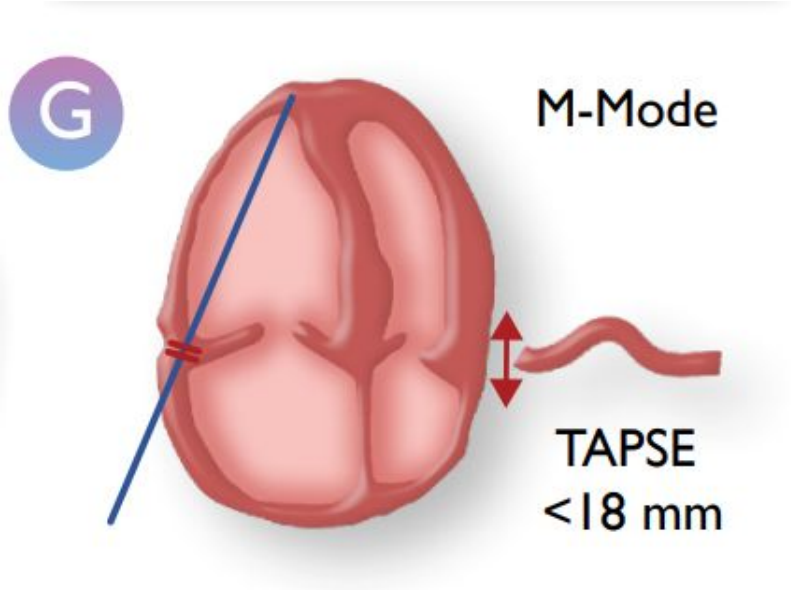
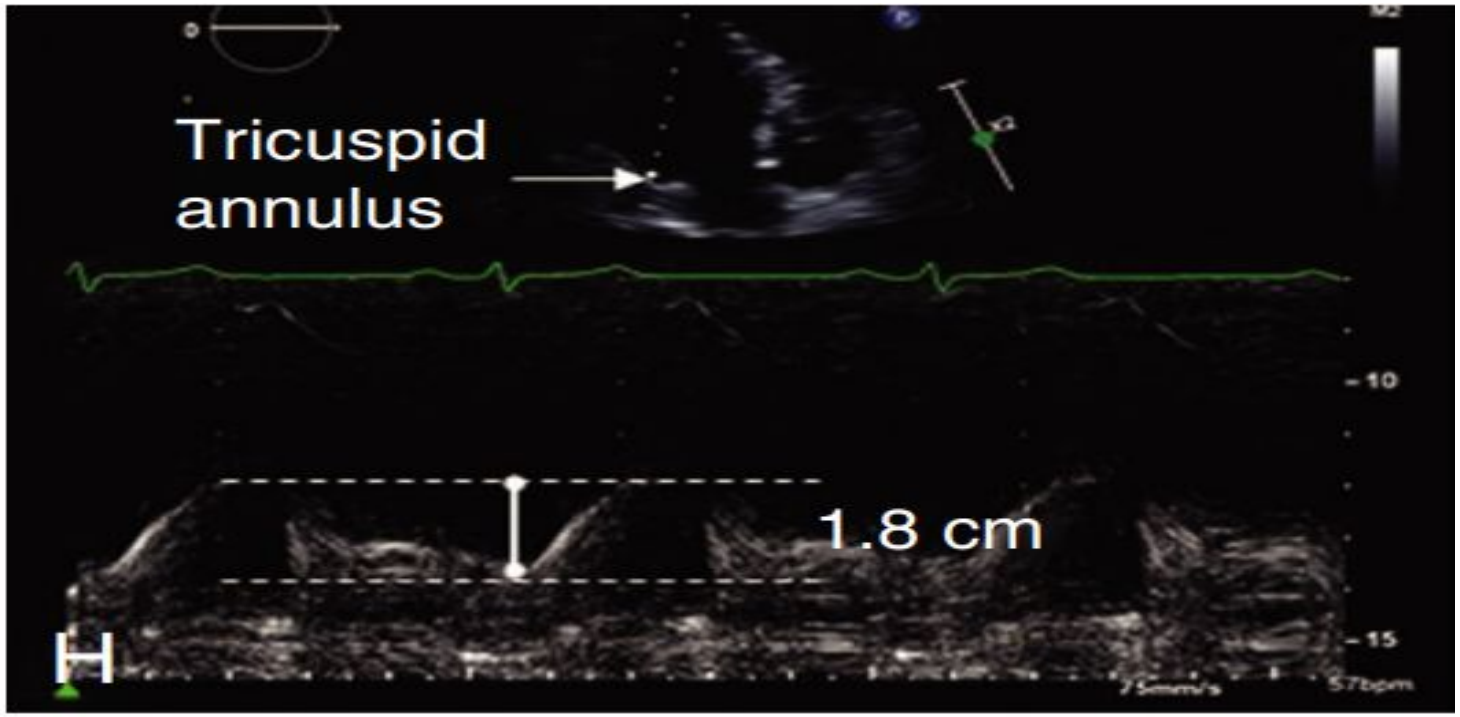
Transthoracic echocardiographic parameters

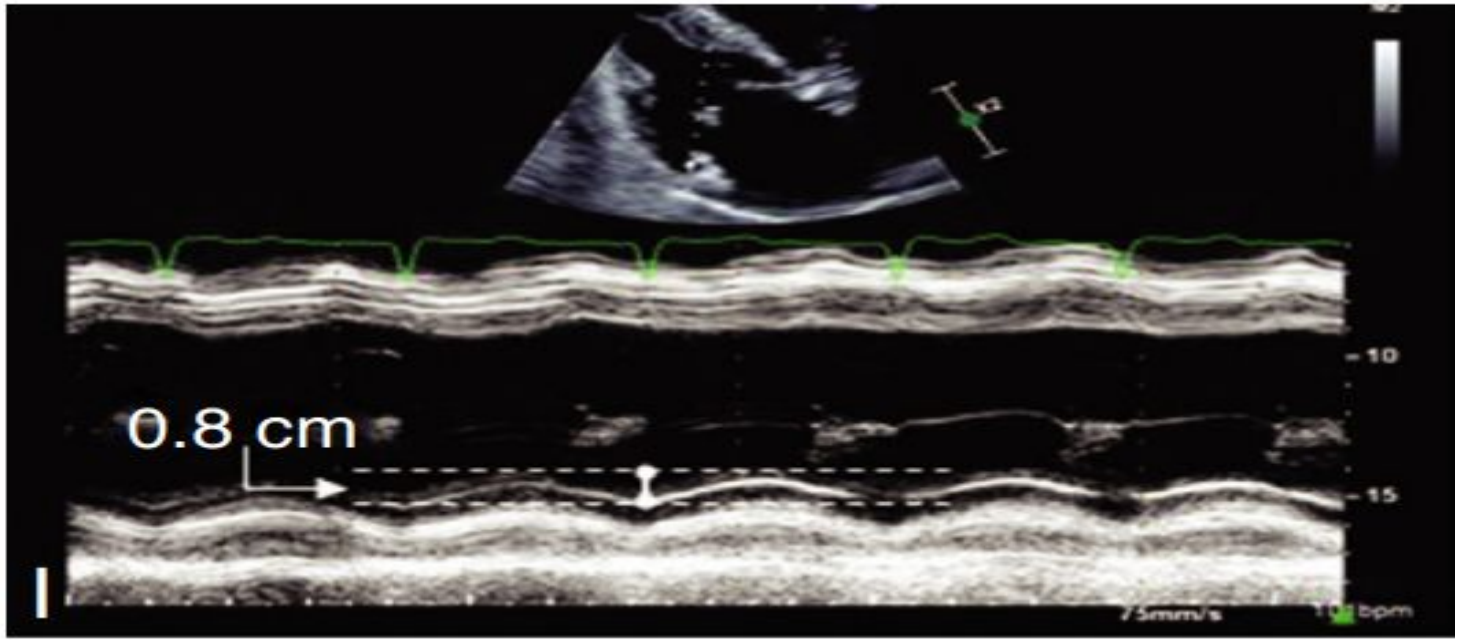


Decreased peak systolic (S') velocity of tricuspid annulus (<9.5 cm/s) measured with tissue Doppler

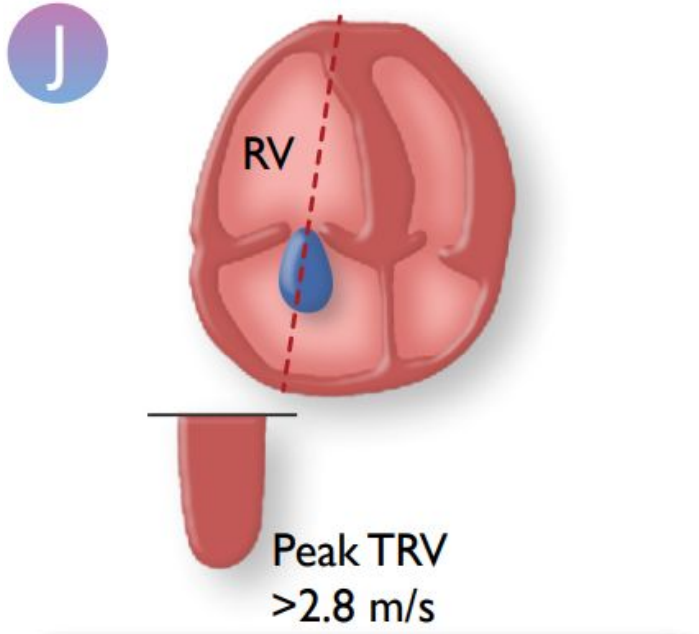


Decreased tricuspid annular plane systolic excursion (TAPSE) measured with M-Mode (<18 mm)

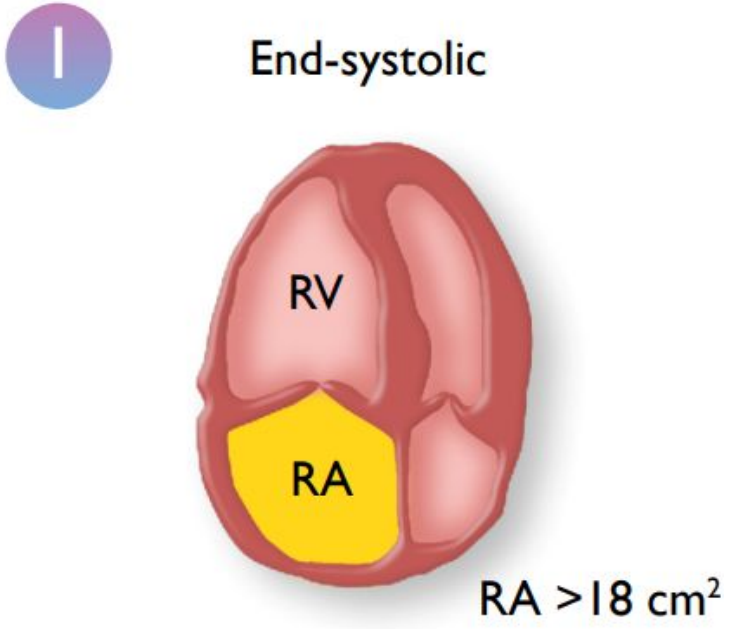

Tricuspid
annulus
1.8 cm
H
-10
-15


0.8 cm
I
-10
-15

Transthoracic echocardiographic parameters

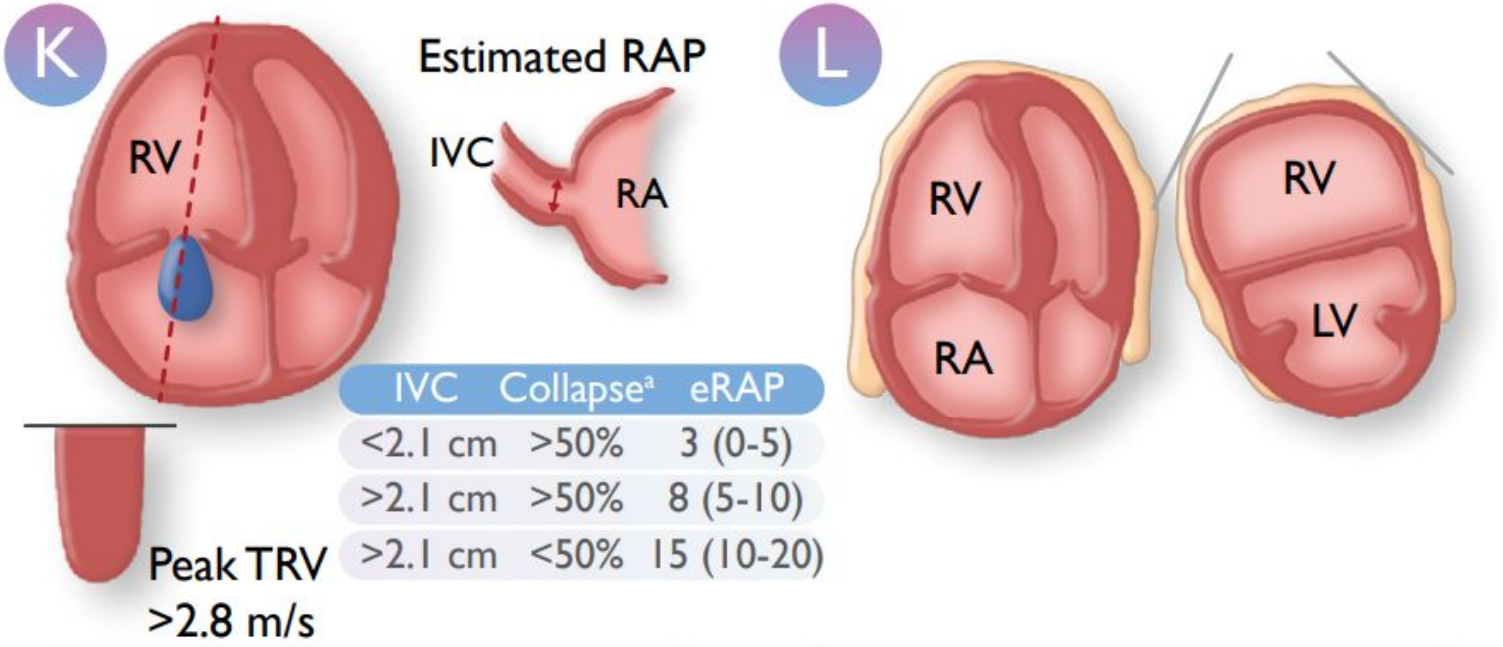


Increased systolic peak tricuspid regurgitation velocity (peak TRV); measured with continuous wave Doppler



Enlarged right atrial area (>18 cm^2); four-chamber view

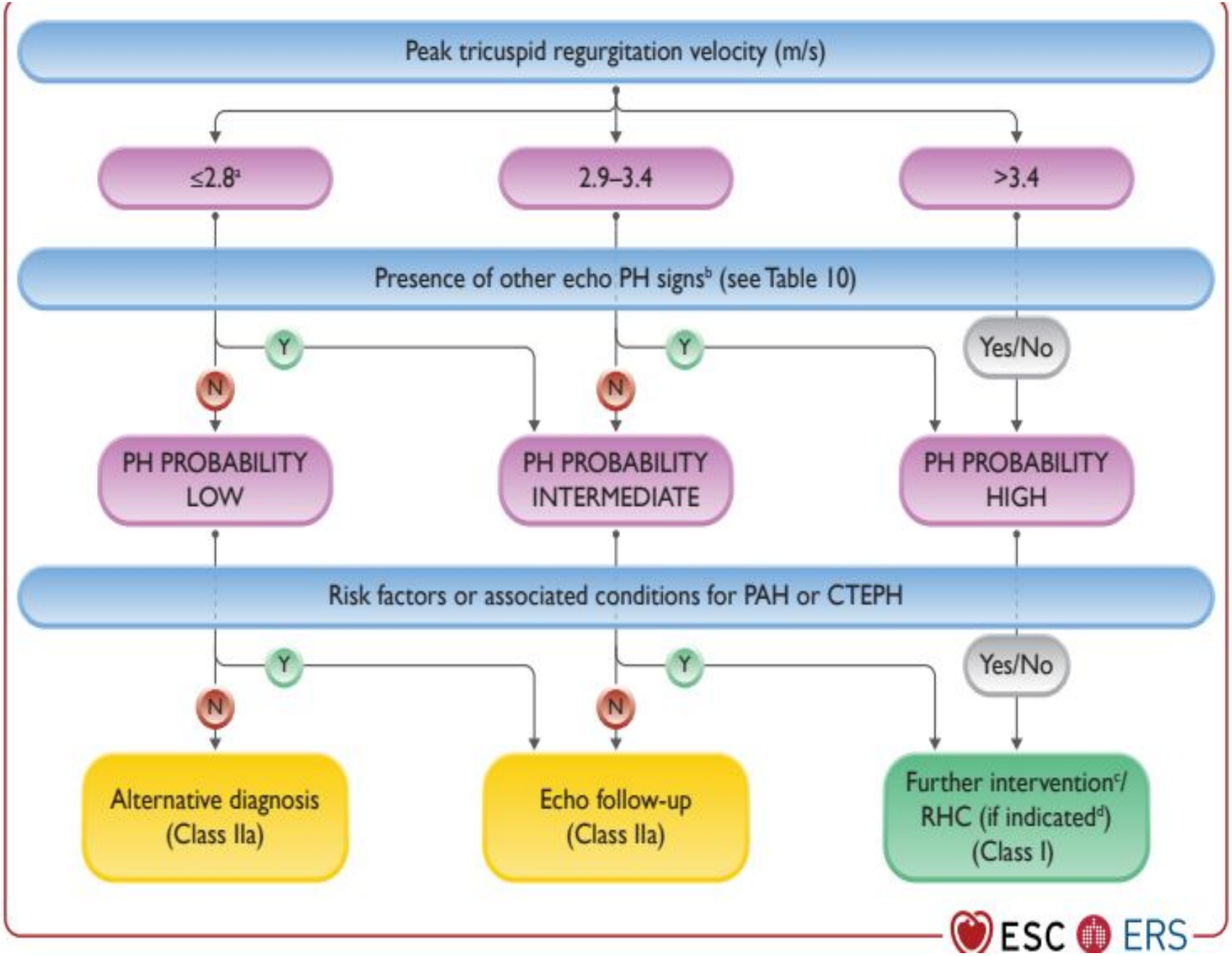
Transthoracic echocardiographic parameters



IVC	Collapse[a]	eRAP
<2.1 cm	>50%	3 (0-5)
>2.1 cm	>50%	8 (5-10)
>2.1 cm	<50%	15 (10-20)

Estimation of systolic pulmonary artery pressure (sPAP);
sPAP = TR pressure gradient + estimated RAP

Presence of pericardial effusion;
four-chamber view;
parasternal short-axis view;
other views (e.g. subcostal view)


Peak tricuspid regurgitation velocity (m/s)
≤2.8[a]
2.9–3.4
>3.4
Presence of other echo PH signs[b] (see Table 10)
Y
N
Y
N
Yes/No
PH PROBABILITY LOW
PH PROBABILITY INTERMEDIATE
PH PROBABILITY HIGH
Risk factors or associated conditions for PAH or CTEPH
Y
N
Y
N
Yes/No
Alternative diagnosis (Class IIa)
Echo follow-up (Class IIa)
Further intervention[c]/ RHC (if indicated[d]) (Class I)
ESC
ERS

Right heart catheterization

The gold standard diagnostic test

1- RHC

2-Vasoreactivetity

3- Exercise

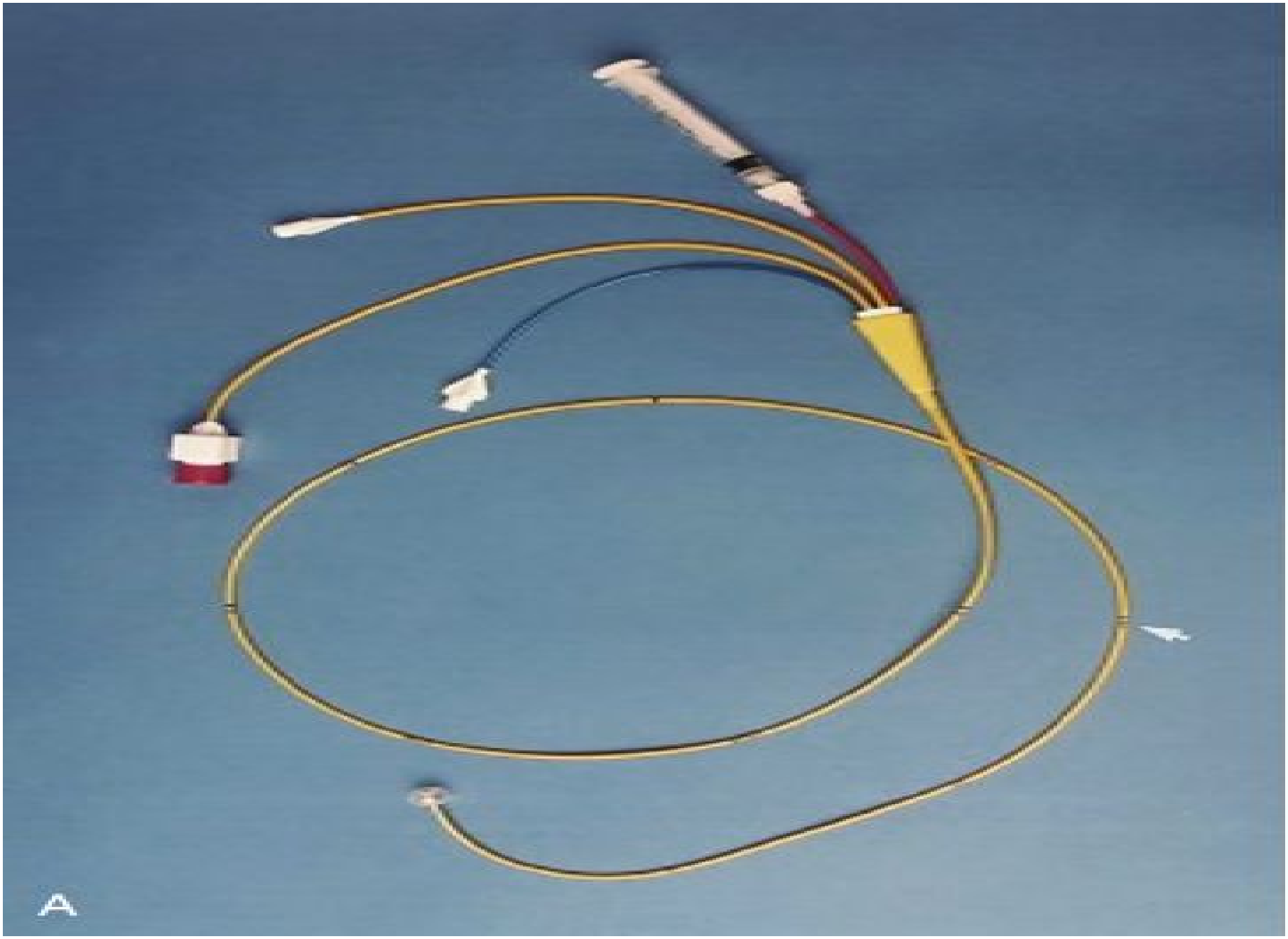
4-Fluid challenge

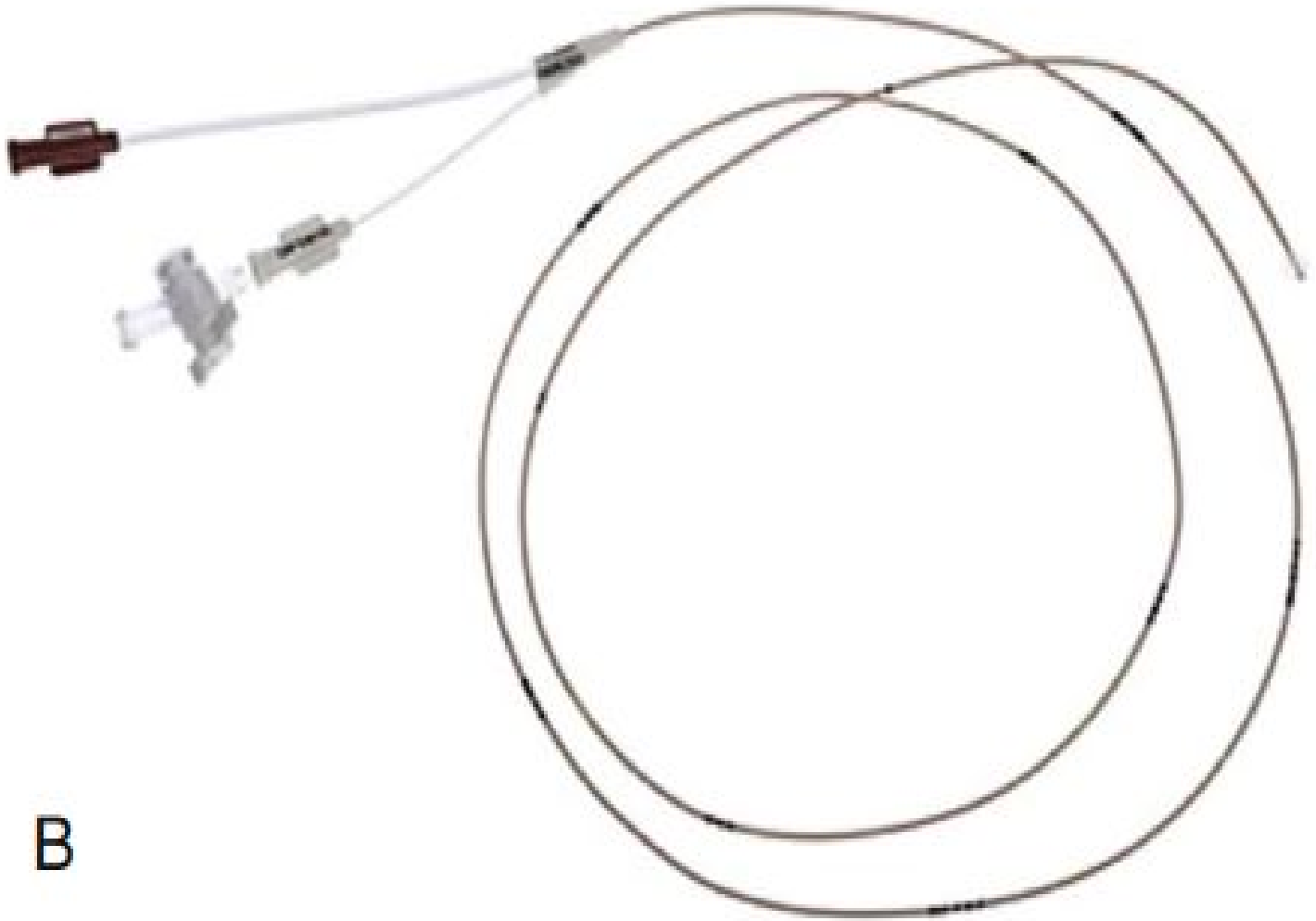
Catheters

- pulmonary balloon- tipped flotation catheters

  with multiple lumens for pressure recording and a thermistor sensor (Swan- Ganz)

- Single-lumen balloon wedge catheters, larger caliber, less catheter whip artifact
- Easy to advance into the right atrium, the right ventricle, and on to the pulmonary artery and PCW position
- pressure tracings alone, or with fluoroscopy





B

Right heart catheterization

- the gold standard for diagnosing and classifying PH.
- haemodynamic
- assessment of heart or Lung transplantation .
- evaluating congenital cardiac shunts.

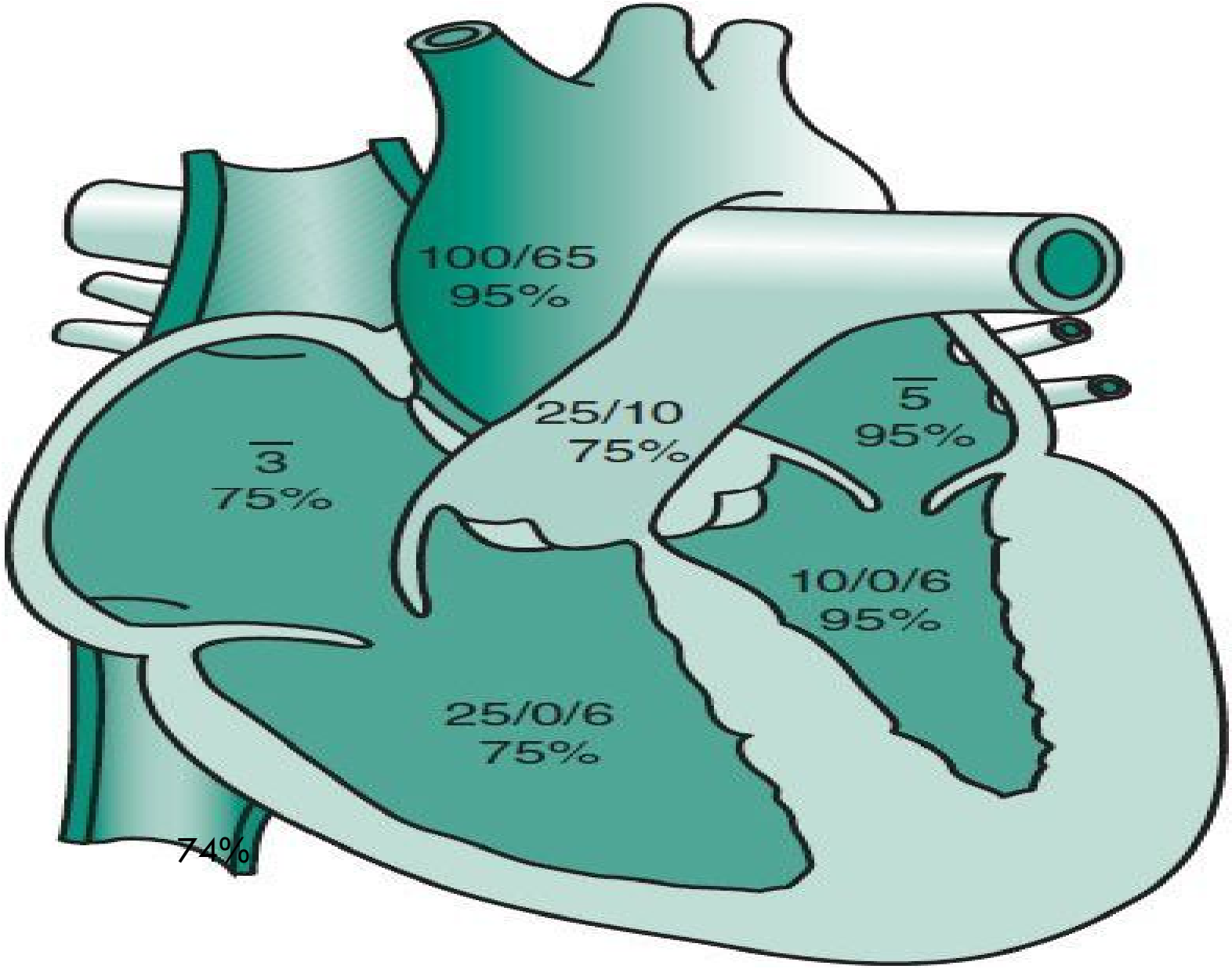
- Serious adverse events (1.1%) and procedure-related mortality (0.055%) .
- A known thrombus or tumor in the RV or RA.
- recently implanted (1 month) pacemaker, mechanical right heart valve, TriClip
- acute infection

Right heart catheterization

- SaO2analysis in different cardiac compartments
- intravascular and intracardiac pressure measurement
- CO assessment, and PAWP measurement ,PVR

RHC

- A complete assessment of cardiopulmonary haemodynamics.
- Cardiac output (CO) should be assessed by the direct Fick method or thermodilution
- pressure measurements should be performed at end expiration(without breath-holding manoeuvre).


100/65
95%
25/10
75%
$\overline{3}$
75%
$\overline{5}$
95%
10/0/6
95%
25/0/6
75%
74%

O2 LEFT- TO- RIGHT/RIGHT –TO- LIFT

- Pa sat >80% = L to R shunt
- Aa sat < 93% = R to L shunt
- PA > SVC 8% =L to R
- An inter- atrial shunt (Low, mid, and high RA)
- An inter-ventricular shunt (RV inflow tract, apex, and outflow tract)

Notes

- Any increase > 5%
- A small L to R shunt might be missed
- The catheter should always be directed away from the coronary sinus (the lowest Sat)
- oxygen saturation in the IVC >SVC
- Flamm's formula

$$Mv\ O2 = = (3 \times SVC\ O2 + IVC\ O2)/4$$

Haemodynamic measures obtained during RHC

Measured variables	Normal value
Right atrial pressure, mean (RAP)	2–6 mmHg
Pulmonary artery pressure, systolic (sPAP)	15–30 mmHg
Pulmonary artery pressure, diastolic (dPAP)	4–12 mmHg
Pulmonary artery pressure, mean (mPAP)	8–20 mmHg
Pulmonary arterial wedge pressure, mean (PAWP)	≤15 mmHg
Cardiac output (CO)	4–8 L/min
Mixed venous oxygen saturation (SvO_2)[a]	65–80%
Arterial oxygen saturation (SaO_2)	95–100%
Systemic blood pressure	120/80 mmHg

Calculated parameters	
Pulmonary vascular resistance (PVR)[b]	0.3–2.0 WU
Pulmonary vascular resistance index (PVRI)	3–3.5 WU·m^2
Total pulmonary resistance (TPR)[c]	<3 WU
Cardiac index (CI)	2.5–4.0 L/min·m^2
Stroke volume (SV)	60–100 mL
Stroke volume index (SVI)	33–47 mL/m^2
Pulmonary arterial compliance (PAC)[d]	>2.3 mL/mmHg

Cardiac Output Measurements

- Thermodilution Method (Td)
- Fick Method
- The indirect Fick method is considered to be less reliable
- Td+ Fick Method is superior to fick alone

Thermodilution Method

- injection of a saline bolus cooler than blood temperature

-The faster the circulation (cardiac output) the quicker the neutralization of the temperature change

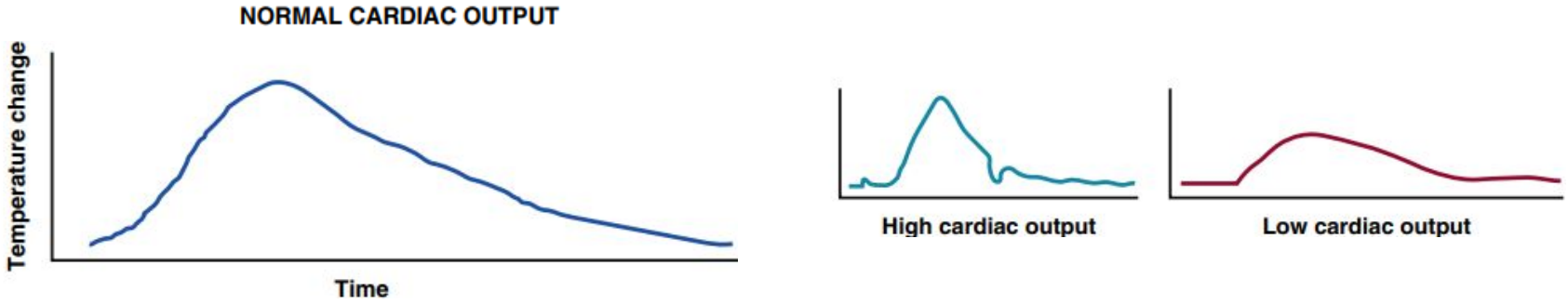
- easy to use

- less accurate in tricuspid regurgitation

-pulmonic regurgitation intracardiac shunt

- low cardiac output

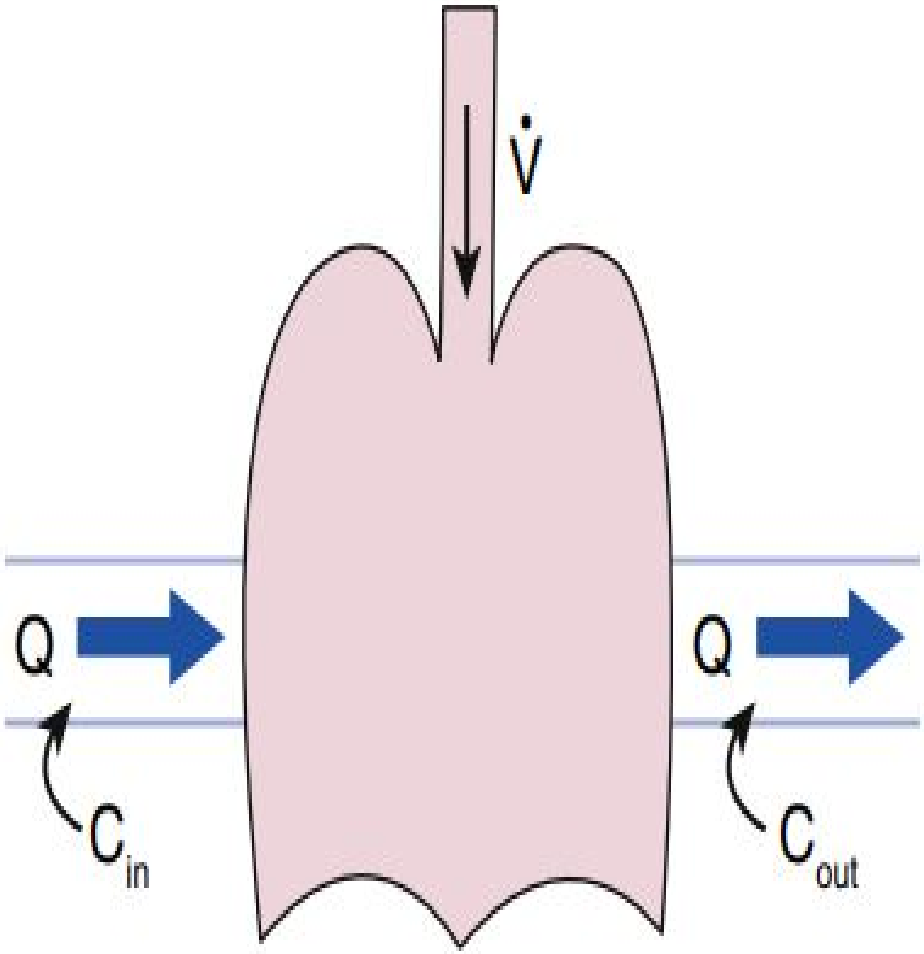
- irregular rhythms.


NORMAL CARDIAC OUTPUT
Temperature change
Time
High cardiac output
Low cardiac output

Fick method

Fick method

- The pulmonary blood flow equals the systemic blood flow PBF = SBF
- The same number of red blood cells (RBCs) that enter the lung must leave the lung
- The difference in the concentration of oxygen between arterial and venous blood and the rate of oxygen uptake in the lung



Rate of indicator out = rate in + rate added

$$Q \times C_{out} = Q \times C_{in} + \dot{V}$$

$$Q = \frac{\dot{V}}{(C_{out} - C_{in})}$$

When O_2 is used as indicator :

$$Q = \frac{\dot{V}O_2}{CaO_2 - C\bar{v}O_2}$$

Fick method

- $$CO = \frac{o2\ consumption}{(Sao2 - Svo2) * 1.36 * Hb * 10}$$
- O2 c =3ml O2 /kg

- $$PVR = \frac{mPAP - PCWP}{CO}$$
- m PAP=2dPAP+sPAP / 3 mm Hg

Blood Flow

- PBF = VO2 ÷ [(PvO2 – Pa O2) × Hb × 1.36 × 10]
- SBF =VO2 ÷ [(SaO2 – Mv O2) × Hb × 1.36 ×10]
- PBF = SBF
- Qp/Qs =$(Sao2-Mvo2)/(Pvo2-Pao2)$
- Flamm's formula :

  Mv O2= (3 × SVC O2 + IVC O2)/4

Fick method

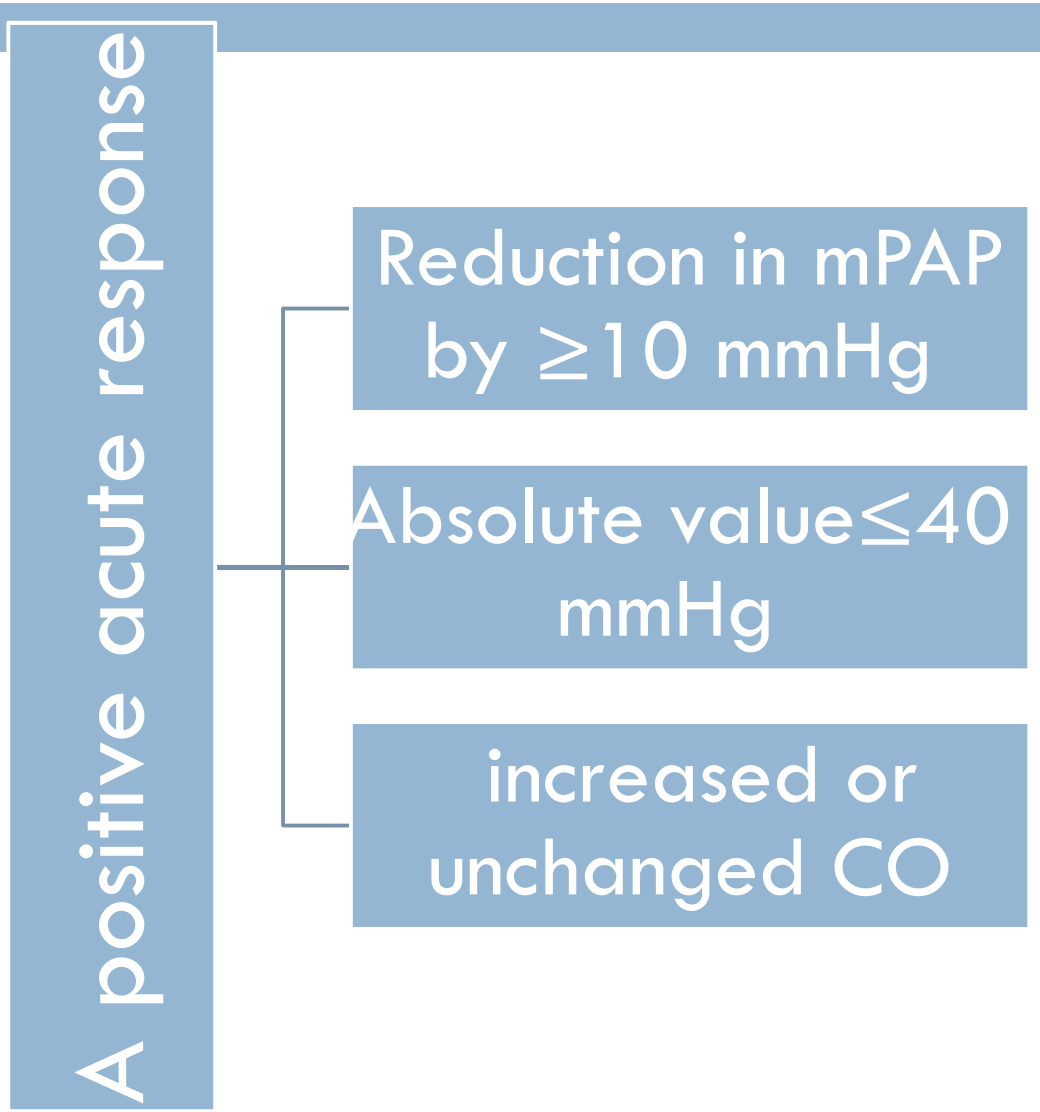
- Shunt fraction $Qp/Qs = \frac{Sao2 - Svo2}{Pvo2 - Pao2}$
- A ratio <1.5 indicates a small left- to- right shunt
- A ratio of 1.5 to 2.0 a moderate- sized shunt
- A ratio of >2.0 a large left to right
- A ratio <1.0 indicates a net right to left shunt

Vasoreactivity testing

- IPAH, HPAH, or DPAH
- Candidates for treatment with high-dose CCB
- CHD…. Defect closure
- PH-LHD…..Heart transplantation.
- Inhaled nitric oxide or Iloprost .
- IV epoprostenol. Adenosine.

Compound	Route	Half-life	Dosage	Duration
Nitric oxide[129]	inh	15–30 s	10–20 p.p.m.	5–10 min[a]
Iloprost[130,131]	inh	30 min	5–10 μg[b]	10–15 min[c]
Epoprostenol[129]	i.v.	3 min	2–12 ng/kg/min	10 min[d]

Results


A positive acute response
Reduction in mPAP by ≥10 mmHg
Absolute value≤40 mmHg
increased or unchanged CO

Recommendations for vasoreactivity testing

Vasoreactivity testing		
Vasoreactivity testing is recommended in patients with I/H/DPAH to detect those who can be treated with high doses of a CCB[129,146]	I	B
It is recommended that vasoreactivity testing is performed at PH centres	I	C
It is recommended to consider a positive response to vasoreactivity testing by a reduction in mPAP ≥10 mmHg to reach an absolute value of mPAP ≤40 mmHg with an increased or unchanged CO[c129]	I	C
Inhaled nitric oxide, inhaled iloprost, or i.v. epoprostenol are recommended for performing vasoreactivity testing[129–132]	I	C
Vasoreactivity testing, for identifying candidates for CCB therapy, is not recommended in patients with PAH other than I/H/DPAH, and in PH groups 2, 3, 4, and 5[124,129]	III	C

Fluid challenge

- HFpEF PAWP ≤15 mmHg .
- rapid infusion over(5–10 min) of 500 mL NS.
- PAWP to ≥18 mmHg suggestive of HFpEF.
- There are insufficient data on the haemodynamic response to fluid challenge in patients with PAH.
- Passive leg raise

Exercise right heart catheterization

- Unexplained dyspnea and normal resting haemodynamic.
- Detect early PVD or left heart dysfunction
- prognostic and functional information in patients at risk of PAH and CTEPH
- exercise RHC may be combined with CPET
- All parameters at rest and peak exercise
- The mPAP/CO and PAWP/CO slopes

Exercise right heart catheterization

- mPAP/CO slope >3 (PAWP/CO slope <2) = PVD
- PAWP/CO slope >2 =LHD (MR, HFpEF)

1-A PAWP > 25 mmHg during supine exercise has been recommended for diagnosing HFpEF

2- lung disease intrathorasic pressure (RAP mPAP)

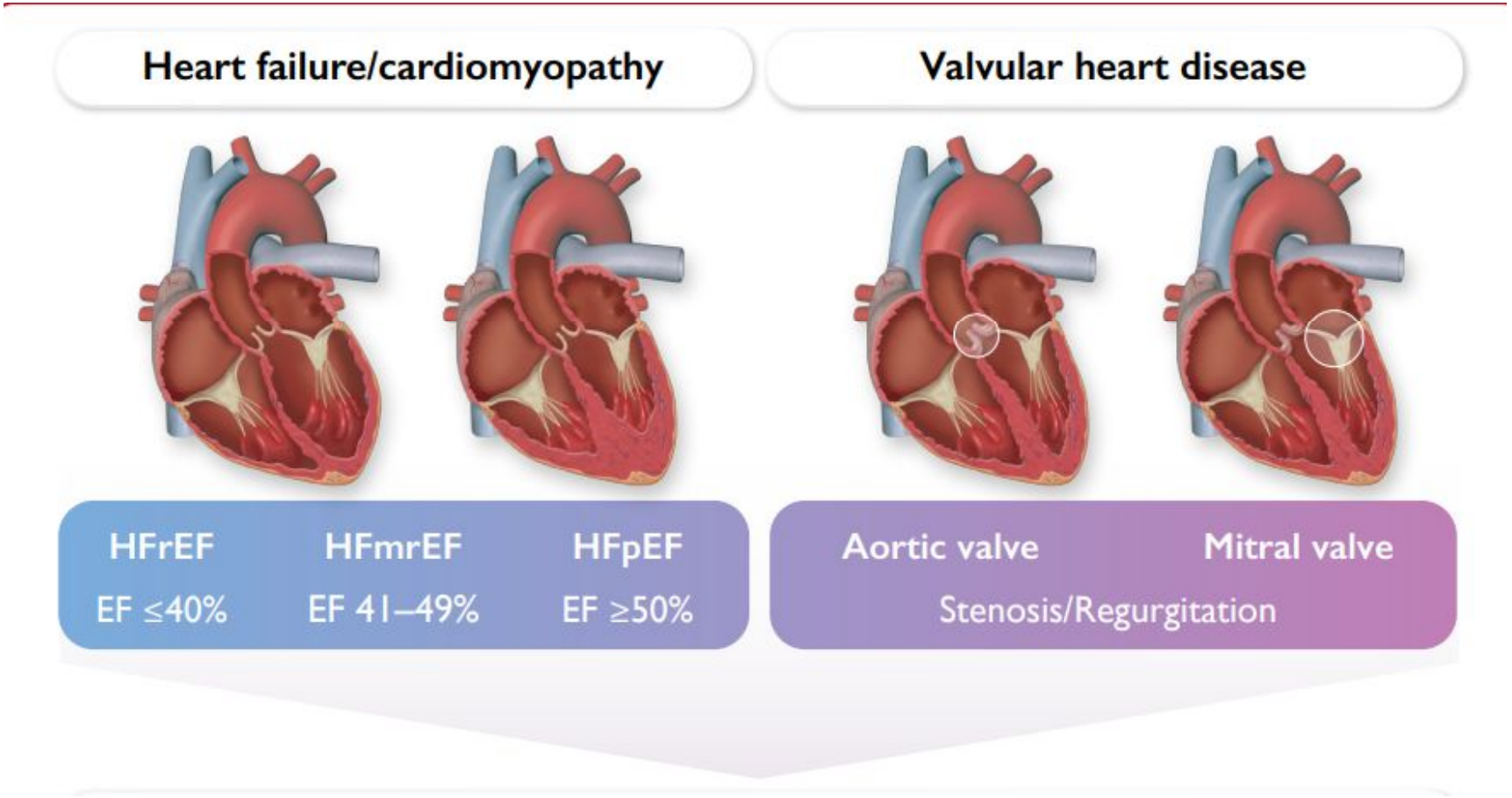
Recommendations for right heart catheterization

Recommendations	Class[a]	Level[b]
Right heart catheterization		
It is recommended that RHC is performed to confirm the diagnosis of PH (especially PAH or CTEPH) and to support treatment decisions[25,26]	I	B
In patients with suspected or known PH, it is recommended that RHC is performed in experienced centres[125]	I	C
It is recommended that RHC comprises a complete set of haemodynamics and is performed following standardized protocols[25,26,145]	I	C

Pulmonary hypertension associated with left heart disease

- Left-sided valvular heart disease
- Cardiomyopathy
- Ischemic heart disease
- HFpEF HFrEF HFmrEF
- Congenital heart disease (L to R) shunt
- Eisenmenger syndrome (R to L) shunt

Pulmonary hypertension associated with left heart disease



Definition

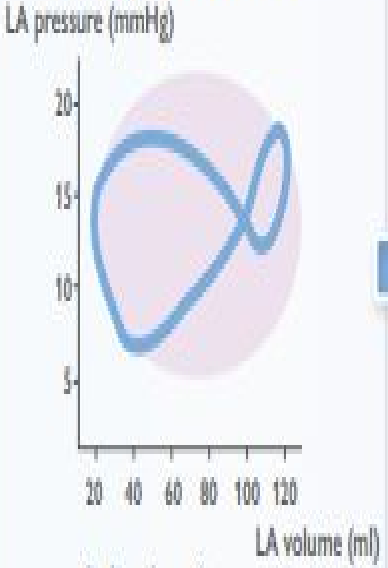
- mPAP >20 mmHg and a PAWP >15 mmHg.
  - IpcPH is defined by PVR ≤2 WU
  - CpcPH is defined by PVR >2 WU
- PH- LHD 65–80% of cases
- 40–72% in patients with HFrEF
- 36–83% in those with HFpEF

Pathophysiology

- An initial passive increase in LV filling pressures and backward transmission into the pulmonary circulation
- PA endothelial dysfunction (vasoconstriction)
- vascular remodeling (both venules and/or arterioles)
- RV dilatation/dysfunction and functional TR
- altered RV–PA coupling

Variable degree of pulmonary congestion, vasoconstriction, vascular remodelling

LA pressure ↑

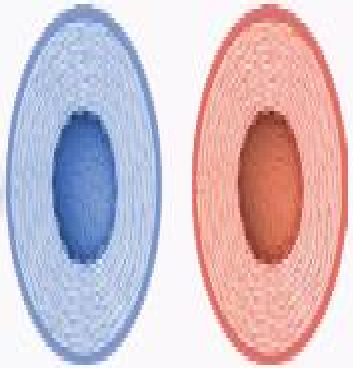


LA dysfunction

Congestion

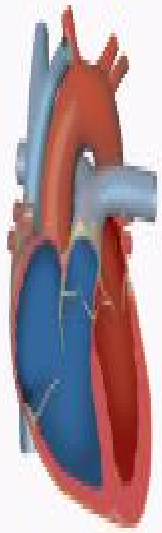
Vasoconstriction

Remodelling

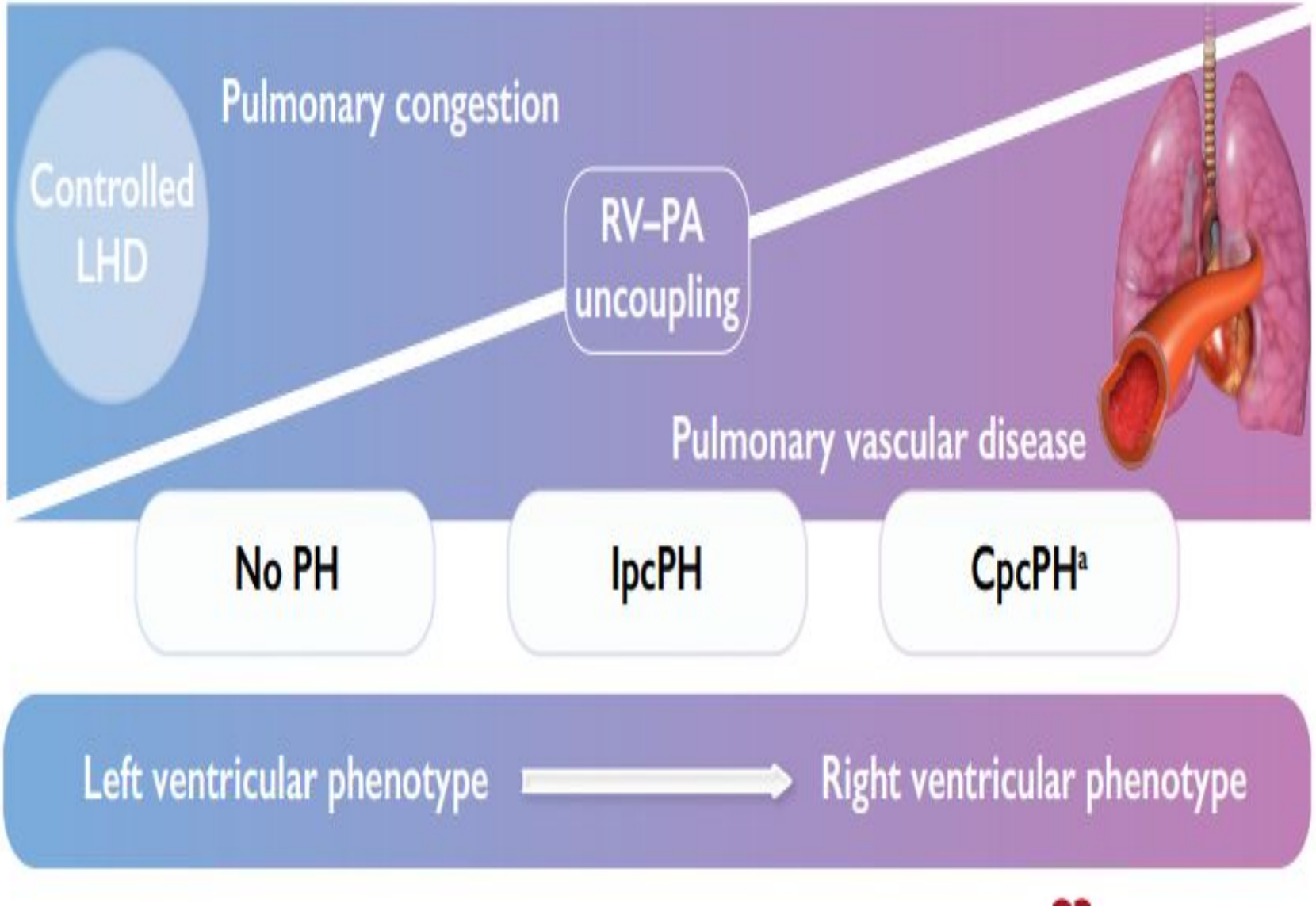


Venous *Arteriolar*

Right heart strain



RV dysfunction


Controlled LHD
Pulmonary congestion
RV–PA uncoupling
Pulmonary vascular disease
No PH
IpcPH
CpcPH[a]
Left ventricular phenotype
Right ventricular phenotype

Prognosis

- LHD increase PHT –PVR worse outcome
- a PVR ≥2.2 WU increase mortality
- CpcPH risk of mortality increase with PVR
- a PVR > 5 WU (HFrEF HFpEF VHD)
- Elevated PVR associated with decreased survival

(patients undergoing interventions for correcting valvular heart disease,heart transplantation , LVAD

PH RV dysfunction high mortality

Heart Failure and Cardiomyopathy

- Ischemic
- infiltrative
- Hypertensive
- substance abuse cardiomyopathy

Heart Failure and Cardiomyopathy

HFrEF

- The prevalence of PH in HFrEF is 30% to 50%

  (PASP cut off >45 mmHg) (mPAP >30mmHg)

  mPAP = 0.65 × PASP + 0.55 mm Hg.

- PH is present in 62% to 77% of HFrEF patients

- PASP (5mmHg) increase mortality by 6-8 %

- mPAP (5mmHg) increase mortality 85% in myocarditis

Pathophysiology

- CpcPH vs. IpcPH and elevated PVR increased RV afterload . Dysfunction RV
- RV dysfunction is predictors of mortality
- Other causes

  CTEPH

  PAH

  COPD

  Sleep Apnoea

Heart Failure and Cardiomyopathy

HFpEF

- 80% of HFpEF patients have PH(PASP >35 mm Hg)
- 30% mortality risk increase per 10 mm Hg PASP
- 50 % mPAP > 25 mm Hg
- 50 % HOCM have PH
- HFpEF, where RV dysfunction , but not LV

Valvular Heart Disease

- Aortic valve disease
  - stenosis AS
  - Regurgitation AR
- Mitral valve disease
  - Stenosis MS
  - Regurgitation MR

Mitral valve disease

Mitral stenosis MS

- PH > 50% of patients MS
- Sever PH >60 mm Hg

- restenosis following mitral balloon valvuloplasty

-decreased 3- year survival following valvotomy

- virtually all patients with severe MS

Mitral valve disease

Mitral regurgitation MR

1 - Most patients with degenerative or functional MR

2- positive association between MR grade and PASP

3- The average 5- year survival rate among primary MR is 25% less with PH /without PH

Aortic Stenosis

- 30% and 36% of asymptomatic AS | mild PH
- 20 % sever PH
- 60% have PH

PH with adult congenital heart

- PH in adults with CHD has a negative impact on the natural course of CHD and worsens clinical status and overall outcome
- ASD VSD PDA

PH with adult congenital heart

- 3–7% of patients with adult CHD
- Female
- underlying lesion
- age and age at defect closure
- 3% after correcting a simple cardiac defect

Eisenmenger syndrome

- Advanced form of adult CHD-associated PAH
- Shunt R to L

1- multiorgan effects of chronic hypoxaemia

2-cyanosis

3-haematological (secondary erythrocytosis and thrombocytopenia)

Diagnosis

- Medical history, physical examination
- Imaging (especially echocardiography)
- Right heart catheterization ((Qp/Qs)
- Direct Fick method
- PVR
- Pulmonary vascular resistance may be overestimated due to erythrocytosis

Clinical classification of PHT associated with congenital heart disease

(1) Eisenmenger syndrome

Includes all large intra- and extracardiac defects that begin as systemic-to-pulmonary shunts and progress to severely elevated PVR and to reverse (pulmonary-to-systemic) or bidirectional shunting. Cyanosis, secondary erythrocytosis, and multiple organ involvement are usually present. Closing the defects is contraindicated.

(2) PAH associated with prevalent systemic-to-pulmonary shunts

- Correctable[a]
- Non-correctable

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

(3) PAH [illegible] defects[b]

(3) PAH with small/coincidental[b] defects

Markedly elevated PVR in the presence of cardiac defects considered haemodynamically non-significant (usually ventricular septal defects <1 cm and atrial septal defects <2 cm of effective diameter assessed by echocardiography), which themselves do not account for the development of elevated PVR. The clinical picture is very similar to IPAH. Closing the defects is contraindicated.

(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, post-operative, haemodynamic lesions.

New but Strange

١-جديد
٢-لاعلاقة له بالكورونا او اي فيروس او باكتريا مرضية معروفة
٣- السيدات غالبا فوق ال ٢٠ سنة
٤- كامل النسيج الرئوي(خاصة القصبات القصيبات) (الاسناخ)
٥- تتهم دور الازياء بنقل العدوى
٦- لا علاج.....لا لقاحات
٧- ينصح الابتعاد...غض النظر(يفضل عدم الحملقة)
٨- Paparazzi
٩- تحذير اسري (+++١٨)


حصري مؤتمر طب وجراحة الصدر
طرطوس ٢٠٢٢


حصري مؤتمر طب
وجراحة الصدر
طرطوس ٢٠٢٢

Thank to your attention