호흡곤란 환자의 진단적 접근과 놓치지 말아야 할 폐혈관질환

이재승

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Definition of dyspnea

- a **subjective experience of breathing discomfort** that consists of qualitatively distinct sensations that vary in intensity
- the experience of dyspnea “derives from interactions among multiple **physiological, psychological, social, and environmental factors**, and may induce secondary physiological and behavioral responses

(ATS2011 statement)

Characteristic sensory descriptors

	Respiratory effort/work	Chest tightness	Air hunger
키워드	**힘들다, 버겁다, 지친다**	**답답하다, 조인다, 막힌다**	**부족하다, 안 들어온다, 질식**
한국어 표현	"숨 쉬는 게 너무 **힘들어요**..." "조금만 걸어도 숨 쉬는 게 **힘들어져요**." "가만히 있어도 숨 쉬는 게 **버거워요**." "숨쉬기 **지친다/피곤하다 ."**	"가슴이 너무 **답답해요**." "가슴이 **뻐근하게 답답해요**." "**쌕쌕거리면서** 가슴이 **답답해요**." "숨 쉴 때 가슴이 **조여요**." "가슴이 **꽉 막힌 느낌**이에요."	"공기가 **모자란 느낌**이에요." "숨 쉬는데도 **부족해요**." "숨이... **안 쉬어져요**." "숨이 **안 들어와요**." "아무리 쉬어도 **숨이 차지 않는 느낌이에요."** "**질식할 것 같은 느낌**이에요."

Respiratory effort/work

- **Respiratory muscle afferents project to the cerebral cortex, and subjects report sensations localized to respiratory muscles when the work of breathing is high.**
- Perceptions of work and effort probably arise through some **combination of respiratory muscle afferents and perceived cortical motor command or corollary discharge.**

Chest tightness

- “**Tightness” is commonly experienced during bronchoconstriction.**
- **Bronchoconstriction gives rise to both a sense of tightness and added physical work of breathing ;** however, blocking pulmonary afferents can diminish tightness.
- **Work/effort may be more related to increased respiratory motor output** needed to overcome airflow obstruction, whereas **tightness may be more specifically related to stimulation of airway receptors**.

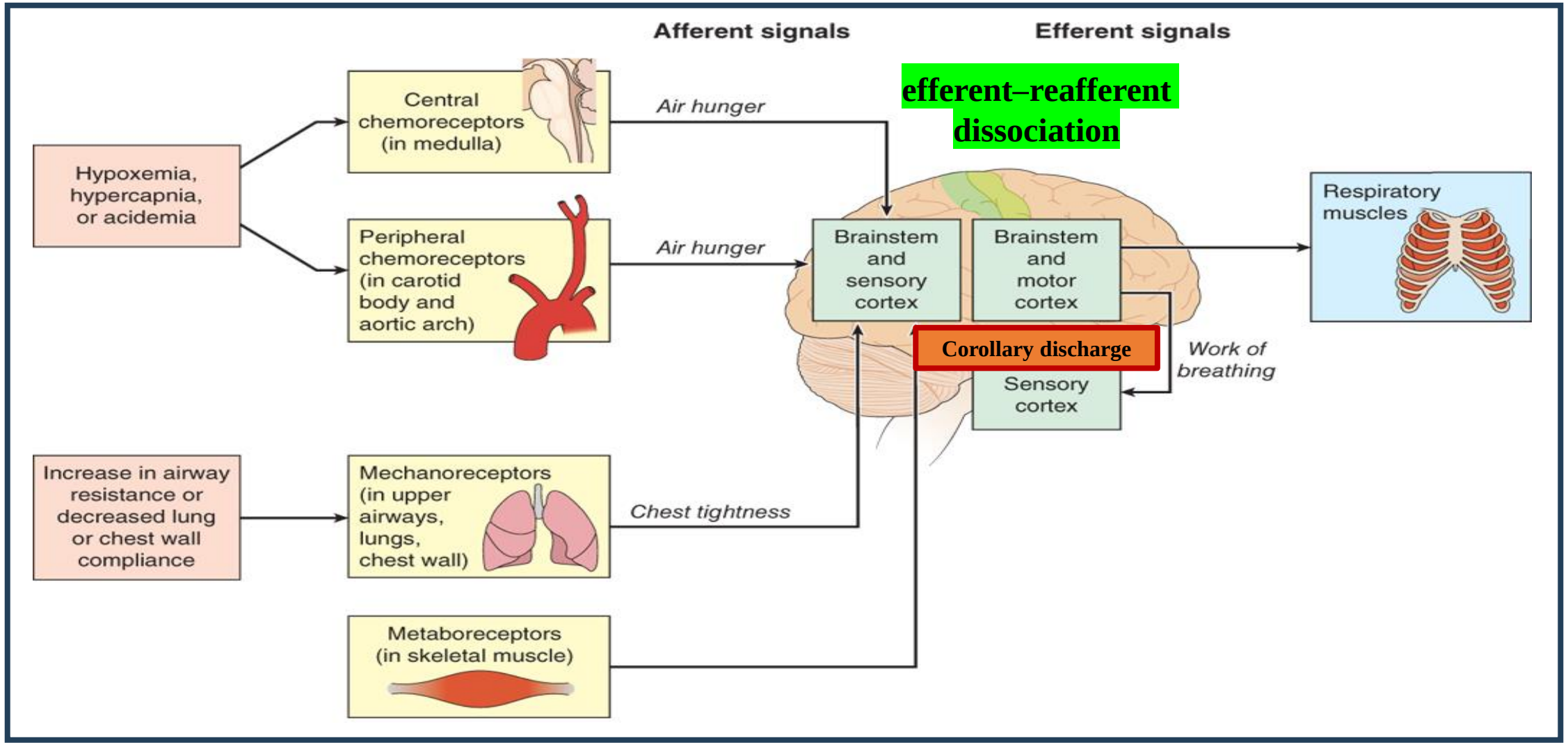
Air hunger/unsatisfied inspiration

- A perception of **not getting enough (or of needing more) air**, which has been variously labeled as air hunger, unsatisfied inspiration, or an unpleasant urge to breathe.
- **A state of imbalance develops between the motor drive to breathe, as sensed** via **corollary discharge**, and afferent feedback from mechanoreceptors of the respiratory system **(efferent–reafferent dissociation, neuro-mechanical uncoupling).**
- Air hunger/unsatisfied inspiration is intensified by stimuli that increase spontaneous ventilatory drive, such as **hypoxia, hypercapnia, acidosis**, and signals arising from **exercise-related drive.**

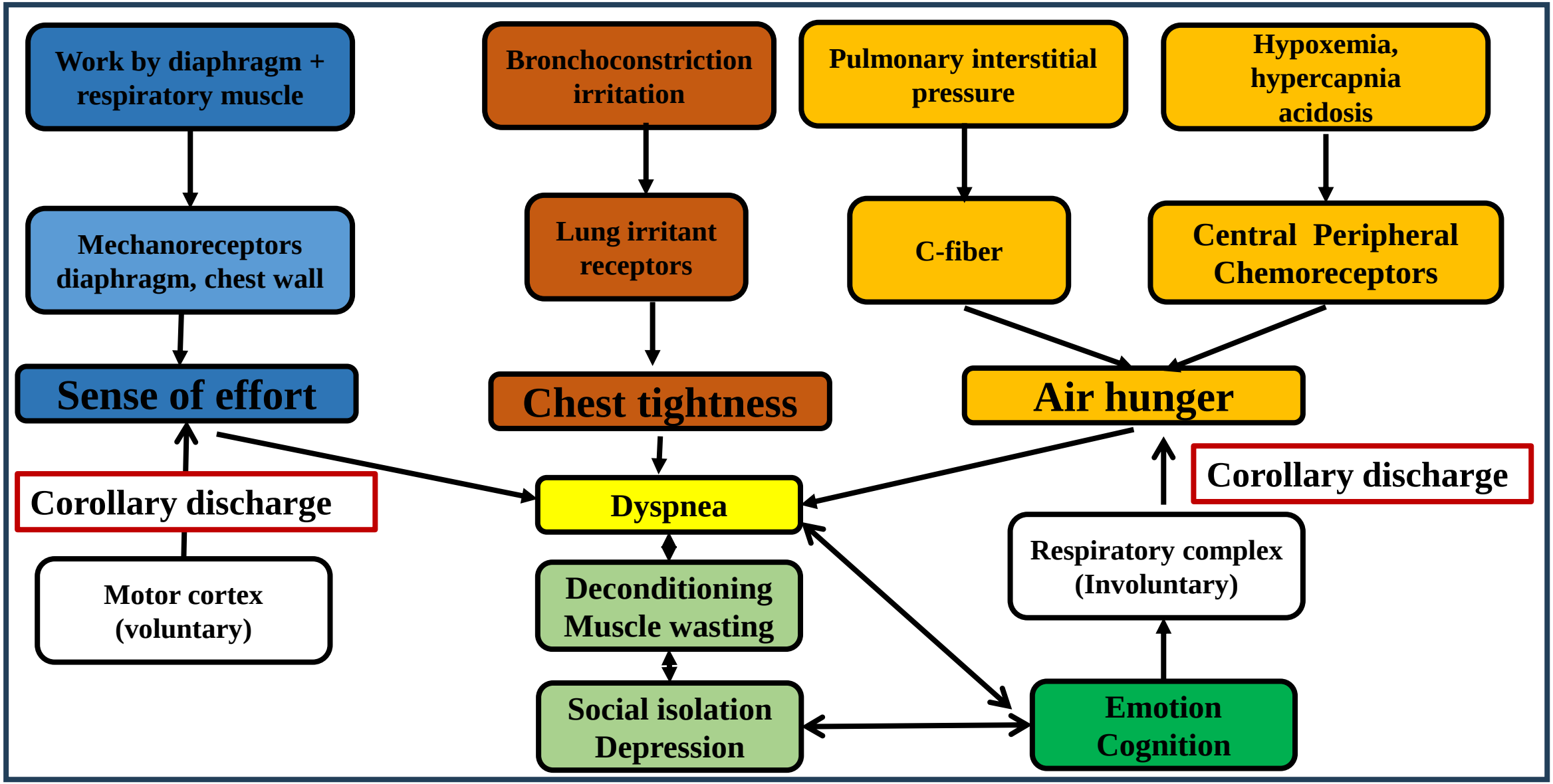
Afferent sources for respiratory sensation

Source of Sensation	Adequate Stimulus
Medullary respiratory corollary discharge	Drives to automatic breathing (hypercapnia, hypoxia, exercise)
Primary motor cortex corollary discharge	Voluntary respiratory drive
Limbic motor corollary discharge	Emotions
Carotid and aortic bodies	Hypercapnia, hypoxemia, acidosis
Medullary chemoreceptors	Hypercapnia
Airway C-fibers	Irritant substances
Upper airway “flow” receptors	Cooling of airway mucosa
Rapidly adapting pulmonary stretch receptors	Airway collapse, irritant substances, large fast (sudden) lung inflations/deflations
Slowly adapting pulmonary stretch receptors	Lung inflation
Pulmonary C-fibers (J-receptors)	Pulmonary vascular congestion
Vascular receptors (heart and lung)	Distention of vascular structures
Trigeminal skin receptors	Facial skin cooling
Chest wall joint and skin receptors	Tidal breathing motion
Muscle spindles in respiratory pump muscle	Muscle length change with breathing motion
Metaboreceptors in respiratory pump muscle	Metabolic activity of respiratory pump

Potential signaling pathways underlying the sensation of dyspnea



Mechanisms of dyspnea



Quality of dyspnea, physiology and disease states

Quality of Dyspnea	Physiology	Disease States
Air hunger, urge to breathe, need more air	Stimulation of respiratory controller via chemoreceptors, pulmonary receptors, vascular receptors	Pneumonia, pulmonary edema, pulmonary embolism, COPD with acute gas exchange abnormalities, asthma, pleural effusion, toxic inhalations
Chest tightness	Stimulation of pulmonary receptors	Asthma, pulmonary edema with bronchospasm, toxic inhalations with bronchospasm
Cannot get a deep breath	Stimulation of respiratory controller; dynamic hyperinflation	COPD, asthma
Increased work or effort to breathe	Mechanical load on the respiratory system; neuromuscular weakness	COPD, asthma, obesity, kyphoscoliosis, Guillain-Barré, myasthenia gravis
Breathing more	Increased ventilation; stimulation of metaboreceptors in muscles	Exercise, cardiovascular deconditioning

Respiratory sensations associated with various conditions

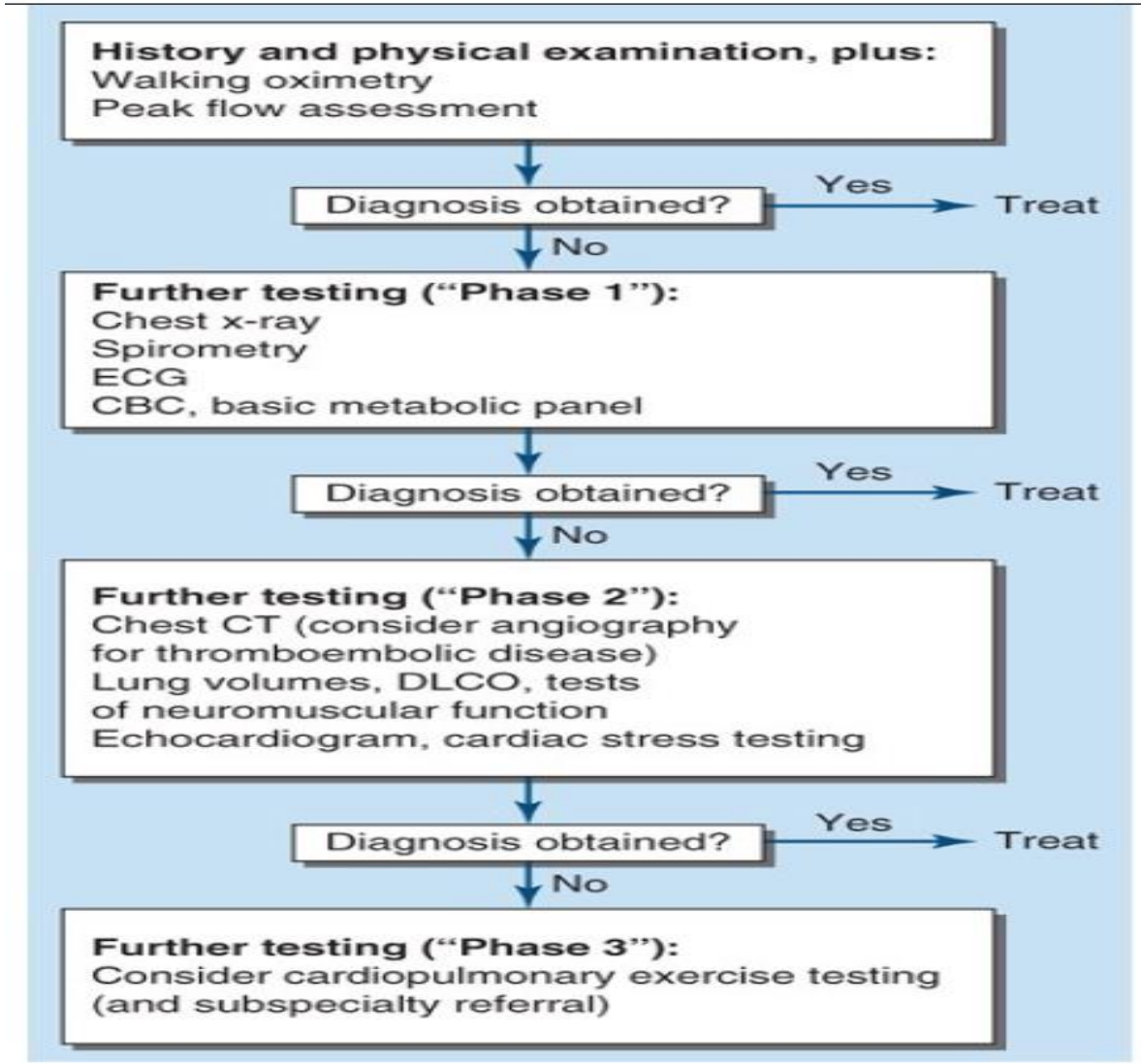
Sensation	COPD	Asthma	ILD	Neuro-muscular and chest wall disease	Congestive heart failure	Pulmonary vascular disease
Rapid breathing						
Incomplete exhalation						
Shallow breathing						
Increased effort/work						
Feeling of suffocation						
Air hunger						
Chest tightness						
Heavy breathing						

Manning HL, et al. N Engl J Med 1995;333:1547-1553

Summary of dyspnea qualities

- Although multiple distinguishable sensations varying in intensity are central to the definition of dyspnea, **these separate sensations seldom, if ever, occur in a pure or isolated fashion in the real world.**
- **Multiple uncomfortable sensations are often present in patients** and together produce the overall perception of dyspnea.

Approach to the patient with dyspnea

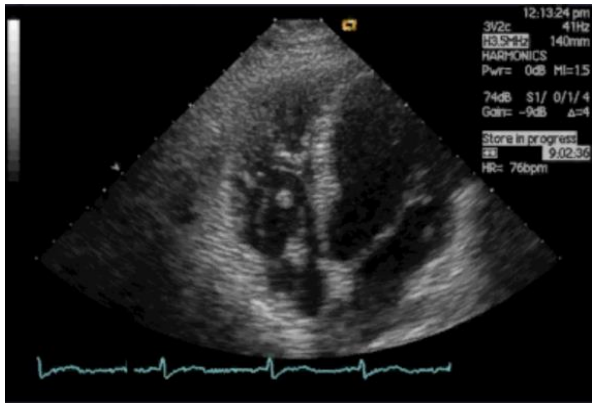


Walking oximetry is recommended as an initial test in the evaluation of dyspnea.

Differential diagnosis of acute dyspnea

Cardiac	Respiratory	Non-cardiac non-respiratory
(Substernal chest pain) CHF; HFrEF, HFpEF Acute coronary syndrome Arrhythmia Valve rupture Hypertensive crisis Tamponade	Upper airway obstruction (Stridor) Aspiration Anaphylaxis Epiglottitis Pneumothorax (absent breath sound) Bronchospasm (wheeze) Asthma COPD Infection (fever, cough, sputum) Pneumonia/TB Pulmonary embolism (pleuritic chest pain)	Metabolic acidosis Diabetes Renal failure Hyperventilation syndrome

Venous thromboembolism (VTE)



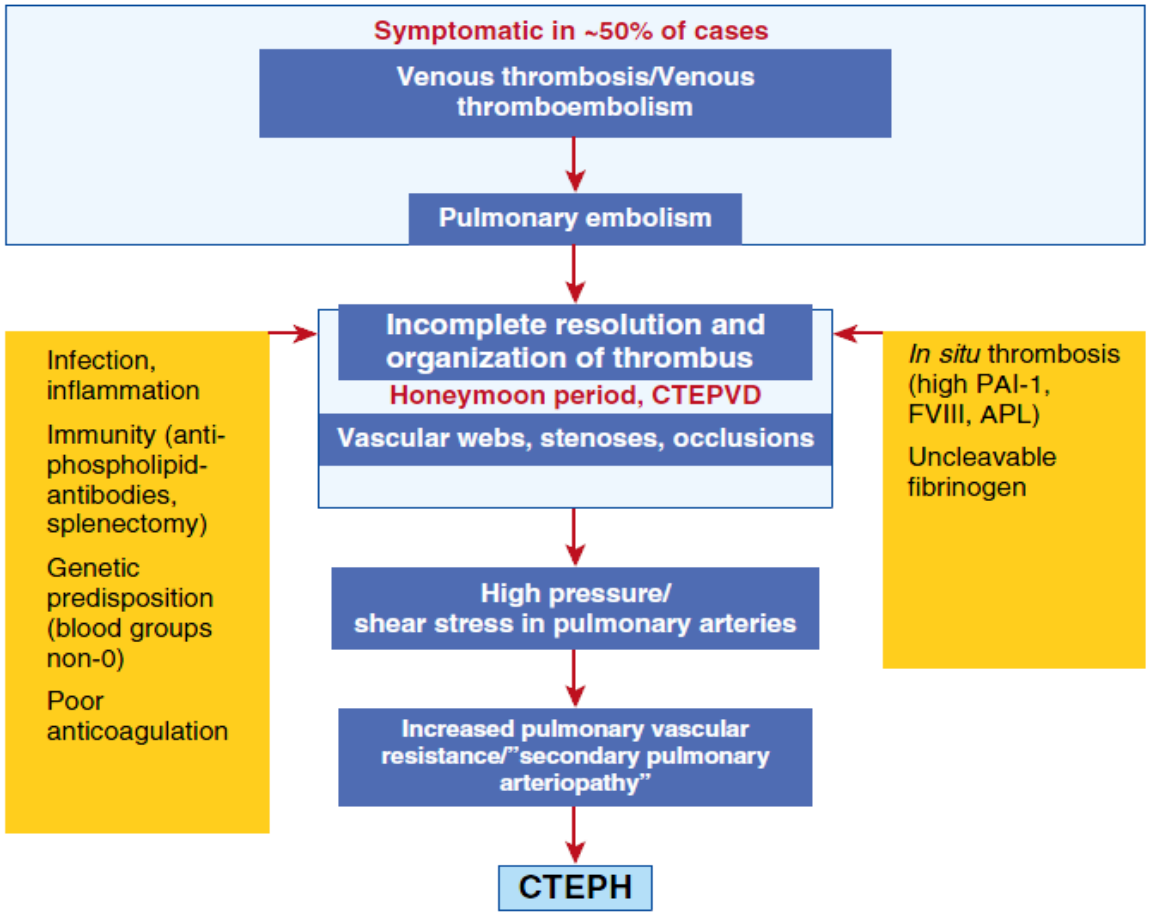
Deep vein Thrombosis (DVT)

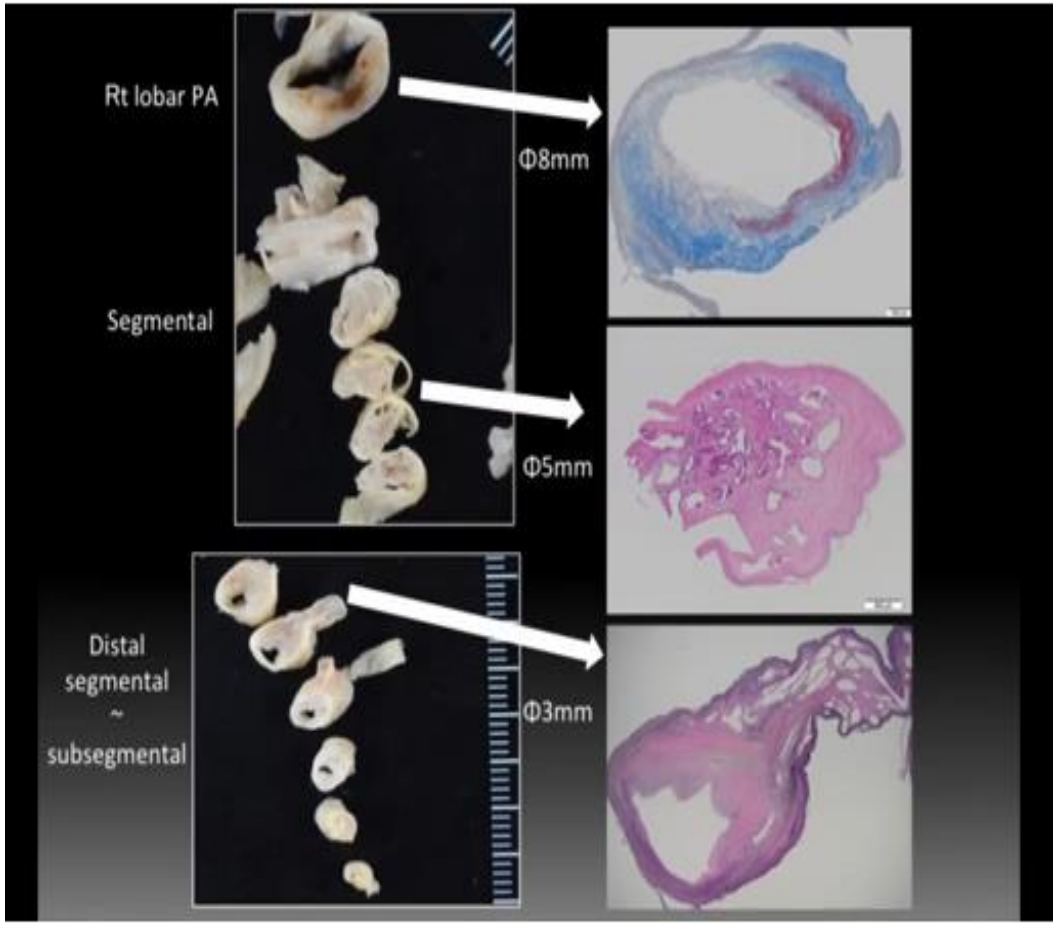
Embolus in transit

Acute pulmonary embolism (PE)

Chronic Thromboembolic Pulmonary hypertension (CTEPH)

Chronic thromboembolic pulmonary hypertension (CTEPH)

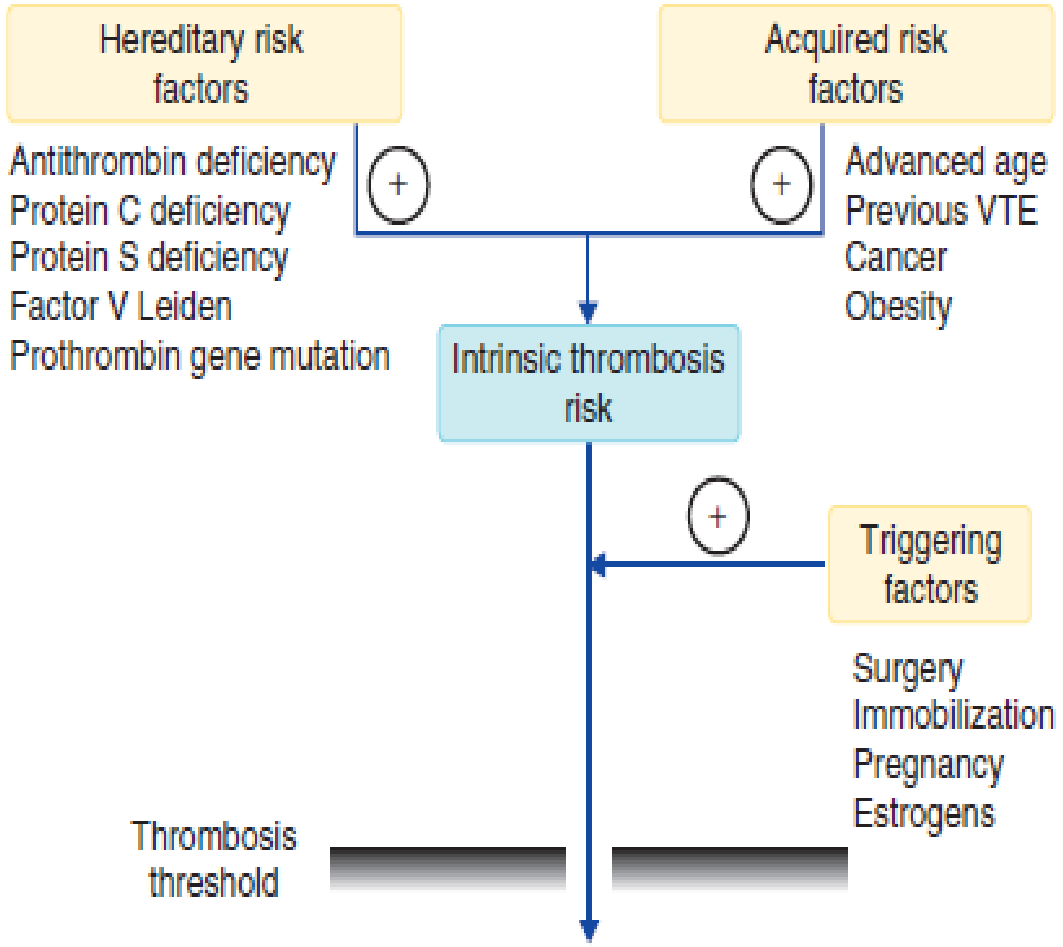


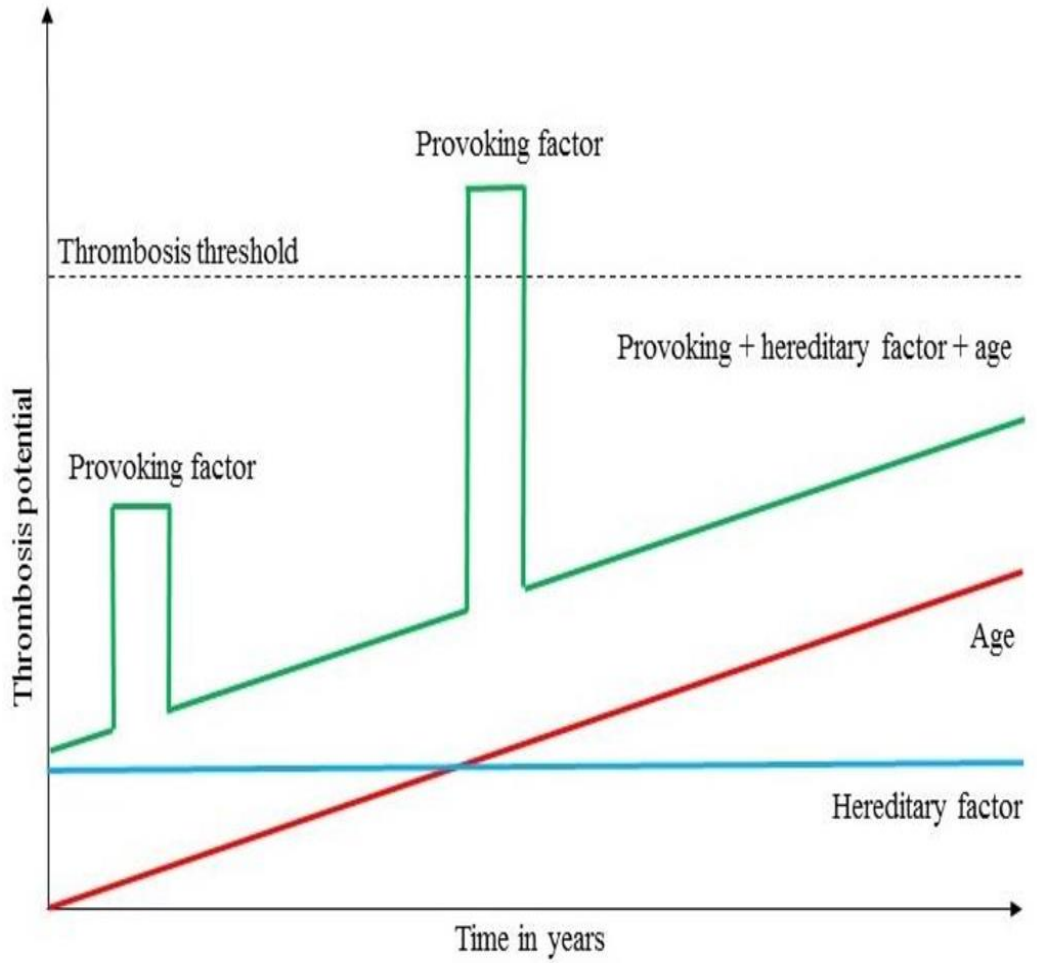


Risk factors of VTE

	Transient or reversible	Persistent
Major	• **Major surgery** (General anesthesia >30 mins) • **Major trauma** with fractures • **Hospitalization** (≥3 days) with while confined to hospital bed	• **Active cancer** (Metastatic or undergoing treatment) • **Antiphospholipid Syndrome** (APS) • **Chronic paralysis** or limb paresis
Minor	• Minor surgery (GA <30 mins) • Pregnancy or postpartum period • Hormonal therapy (Oral contraceptives, HRT) • Long-distance travel (e.g., >8 hours) • Minor leg injury with reduced mobility	• Congestive heart failure • Obesity (BMI >30 kg/m²) • **Inherited thrombophilia** • Inflammatory Bowel Disease (IBD)

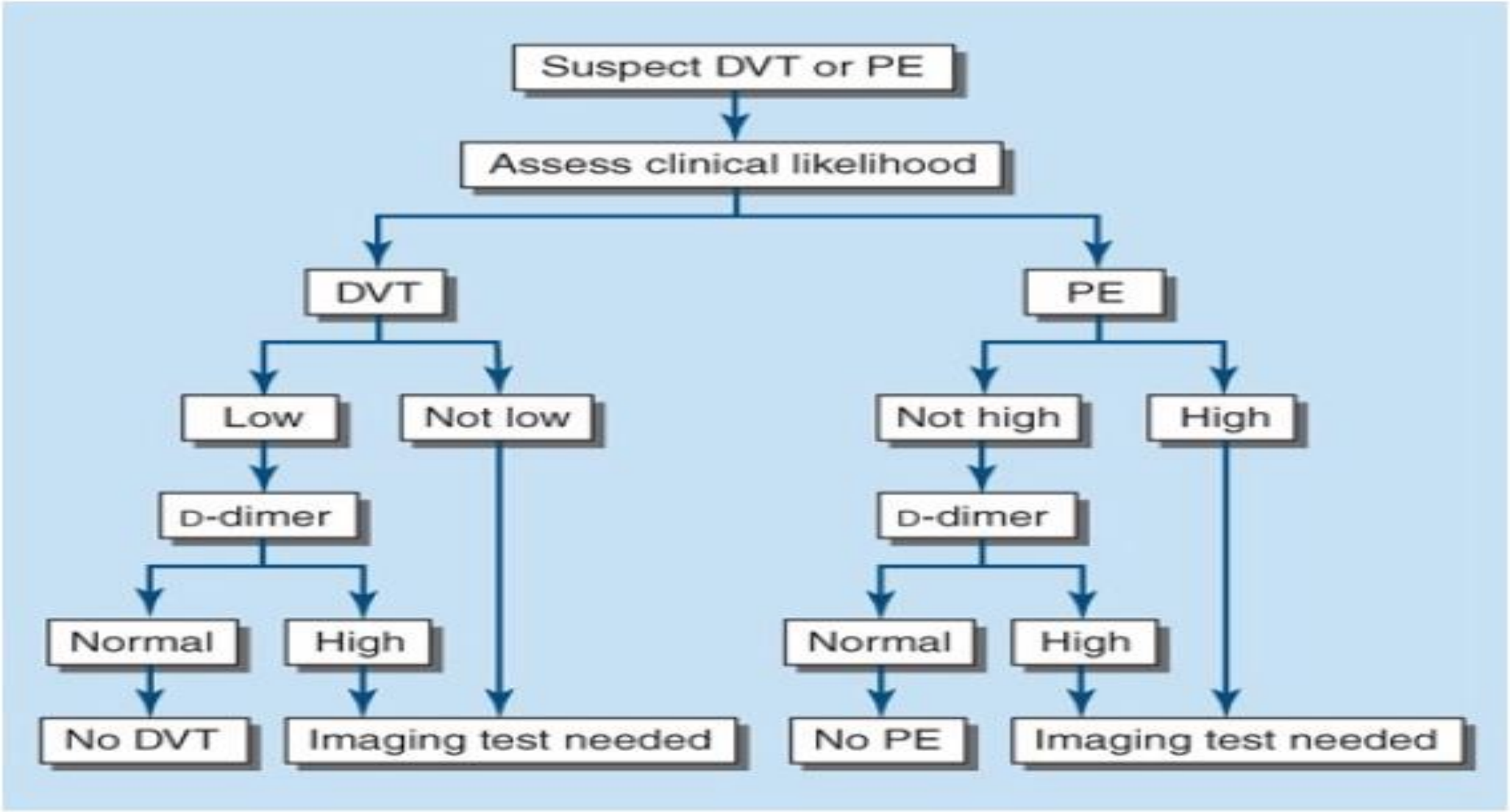
Thrombosis potential model





Rosendaal, F.R. Lancet 1999; 353: 1167–1173

Diagnosis of DVT or PE



Clinical presentations of acute PE

Feature	PE confirmed (*n* = 1880)	PE not confirmed (*n* = 528)
Dyspnoea	50%	51%
Pleuritic chest pain	39%	28%
Cough	23%	23%
Substernal chest pain	15%	17%
Fever	10%	10%
Haemoptysis	8%	4%
Syncope	6%	6%
Unilateral leg pain	6%	5%
Signs of DVT (unilateral extremity swelling)	24%	18%

Clinical prediction rules for PE: Revised Geneva score

Variable	Points
Predisposing factors	
Age > 65years	1
Previous DVT or PE	3
Surgery or fracture within 1 month	2
Active malignancy	2
Symptoms	
Unilateral lower limb pain	3
Hemoptysis	2
Clinical signs	
HR 75-94/min	3
HR ≥ 95/min	5
Pain on lower limb deep vein at palpation and unilateral edema	4

Low 0-3 (≒ 10%) Intermediate 4-10 (≒ 30%) High >10 (≒65%)

Clinical prediction rules for PE: Wells' score

Items	Clinical decision rule points	
	Original version	Simplified version
Previous PE or DVT	1.5	1
Heart rate >100 b.p.m	1.5	1
Surgery or immobilization within the past 4 weeks	1.5	1
Hemoptysis	1	1
Active cancer	1	1
Clinical signs of DVT	3	1
Alternative diagnosis less likely than PE	3	1
Clinical probability		
Three-level score		
Low(~10%)	0~1	N/A
Intermediate(30%)	2~6	N/A
High(65%)	≥7	N/A
Two-level score		
PE unlikely(~12%)	0~4	0~1
PE likely(30%)	≥5	≥2

b.p.m. = beats per minute; DVT = deep vein thrombosis; N/A = not applicable; PE = pulmonary embolism.

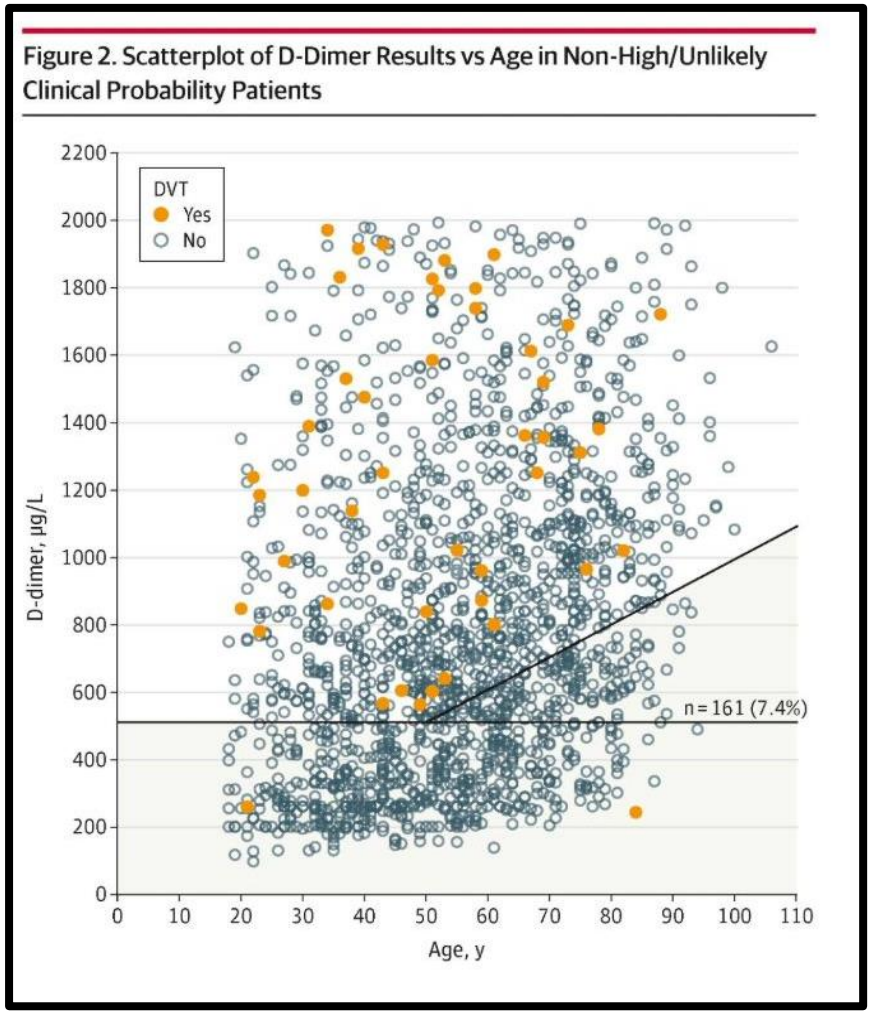
D-dimer assays

- **Sensitivity depend on method:**
  - **ELISA (sensitivity-95%)**
  - Latex agglutination (less sensitive, fast, cheap)
  - Whole blood agglutination
  - **FEU(Fibrinogen Equivalent Units; 500ug/L)**
  - **DDU(D-dimer Units; 250ug/L)**
- **Specificity 30-70%**

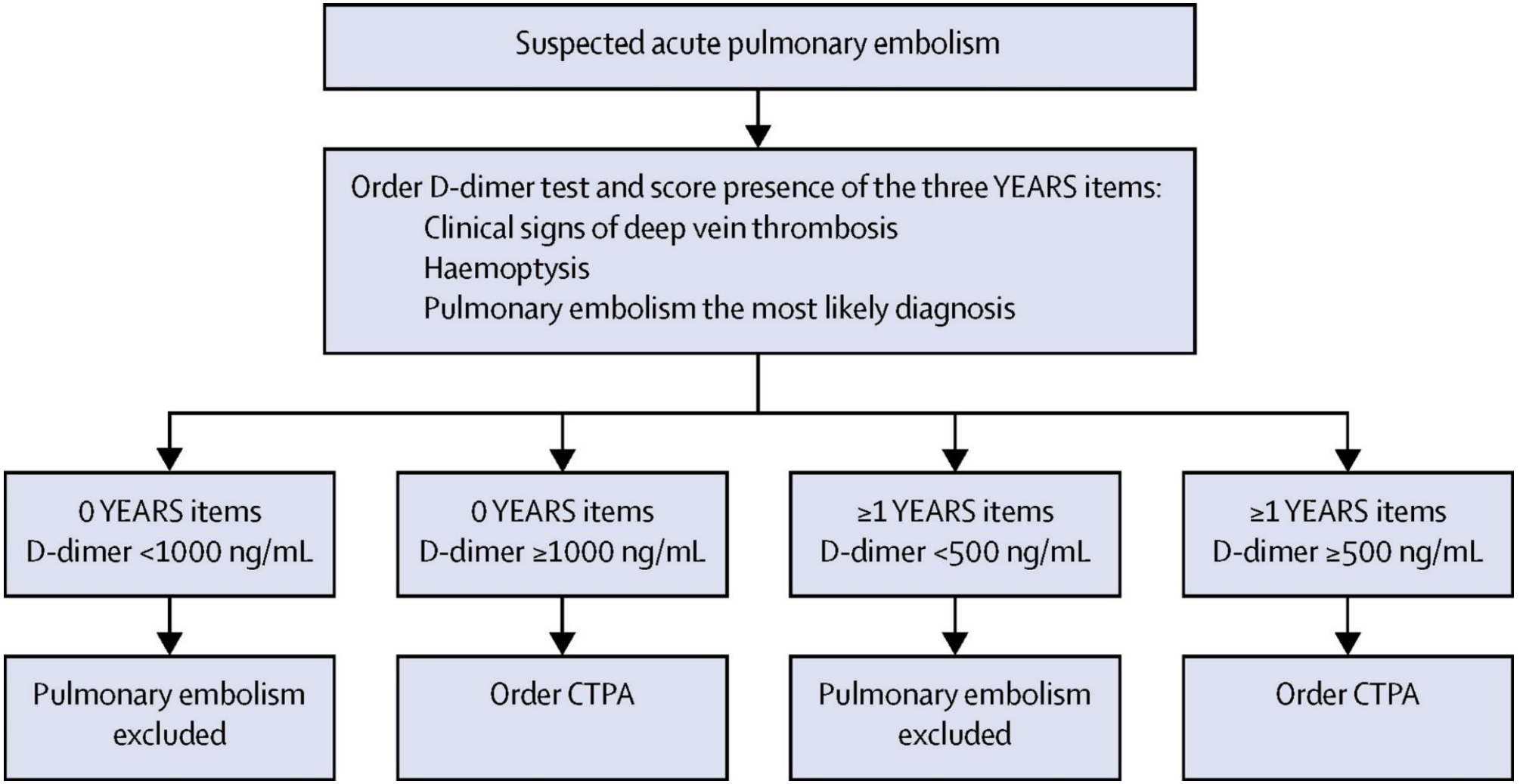
  (cancer, inflammation, bleeding, surgery, trauma and necrosis, etc)

➢ **Age-adjusted D-dimer**

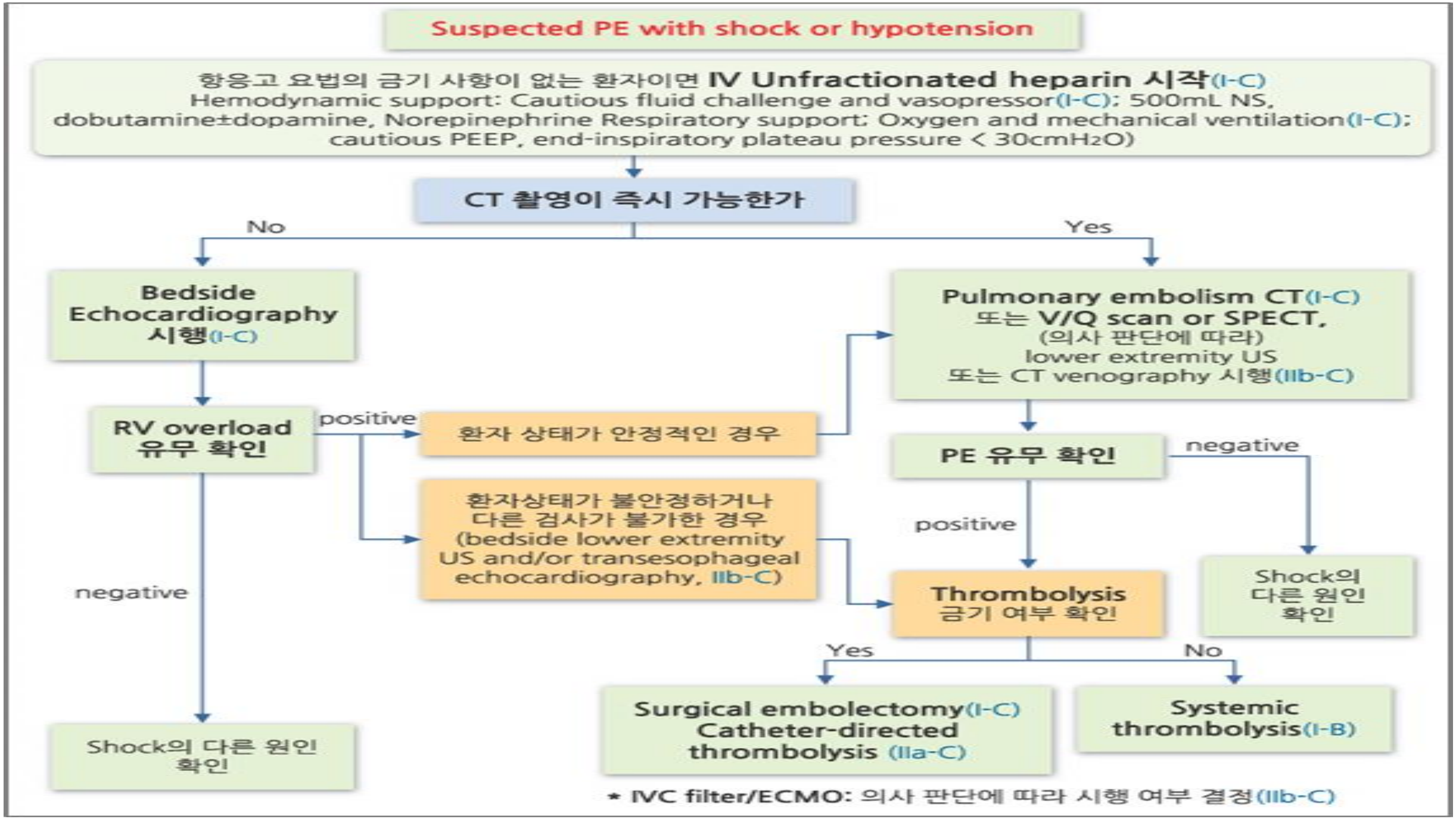
Age *10 ug/L (> 50 years)



The YEARS algorithm for acute PE



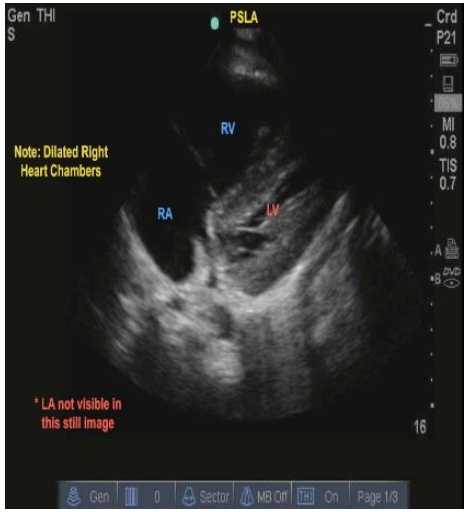
The Lancet 2017 390289-297DOI: (10.1016/S0140-6736(17)30885-1)

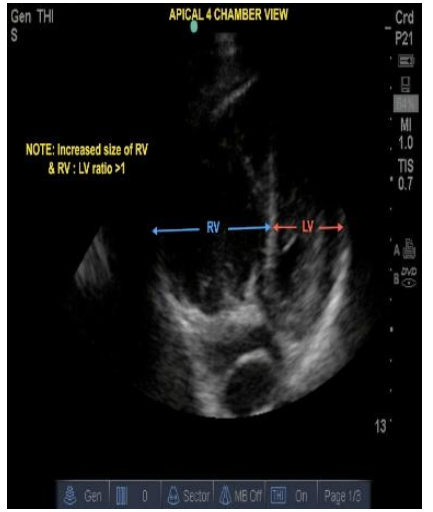

Suspected PE with shock or hypotension
항응고 요법의 금기 사항이 없는 환자이면 IV Unfractionated heparin 시작(I-C)
Hemodynamic support: Cautious fluid challenge and vasopressor(I-C): 500mL NS, dobutamine±dopamine, Norepinephrine Respiratory support: Oxygen and mechanical ventilation(I-C); cautious PEEP, end-inspiratory plateau pressure < 30cmH2O)
CT 촬영이 즉시 가능한가
No
Yes
Bedside Echocardiography 시행(I-C)
RV overload 유무 확인
positive
환자 상태가 안정적인 경우
환자상태가 불안정하거나 다른 검사가 불가한 경우 (bedside lower extremity US and/or transesophageal echocardiography, IIb-C)
negative
Shock의 다른 원인 확인
Pulmonary embolism CT(I-C) 또는 V/Q scan or SPECT, (의사 판단에 따라) lower extremity US 또는 CT venography 시행(IIb-C)
PE 유무 확인
negative
positive
Shock의 다른 원인 확인
Thrombolysis 금기 여부 확인
Yes
No
Surgical embolectomy(I-C) Catheter-directed thrombolysis (IIa-C)
Systemic thrombolysis(I-B)
★ IVC filter/ECMO: 의사 판단에 따라 시행 여부 결정(IIb-C)

Echocardiography

- **RV overload**

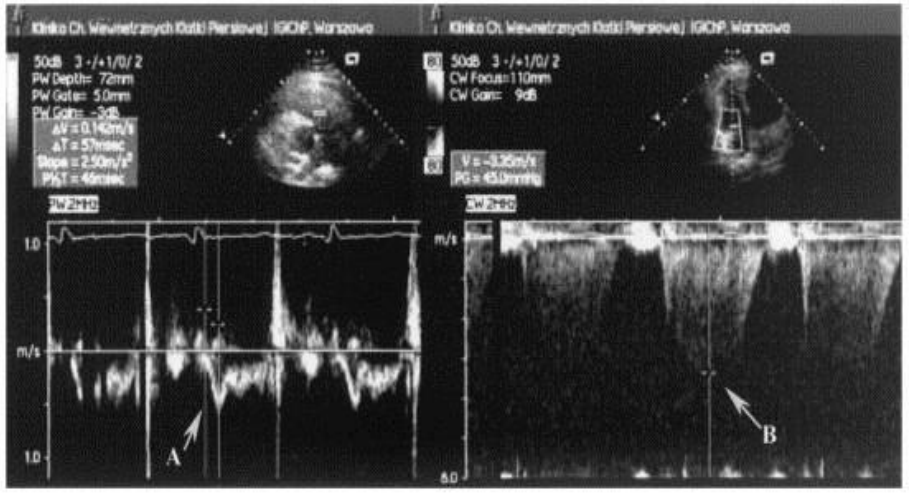
1) right side cardiac thrombus
2) RV diastolic dimension > 30mm or a RV/LV ratio> 1
(3) systolic fattening of interventricular septum
4) Acceleration time <90ms or TVPG > 30mmHg in absence of RV hypertrophy





- **McConnell sign**

➔ normokinesia or hyperkinesia of the apical segment of RV wall despite hypokinesia and/or akinesia of the remaining part of the RV free wall



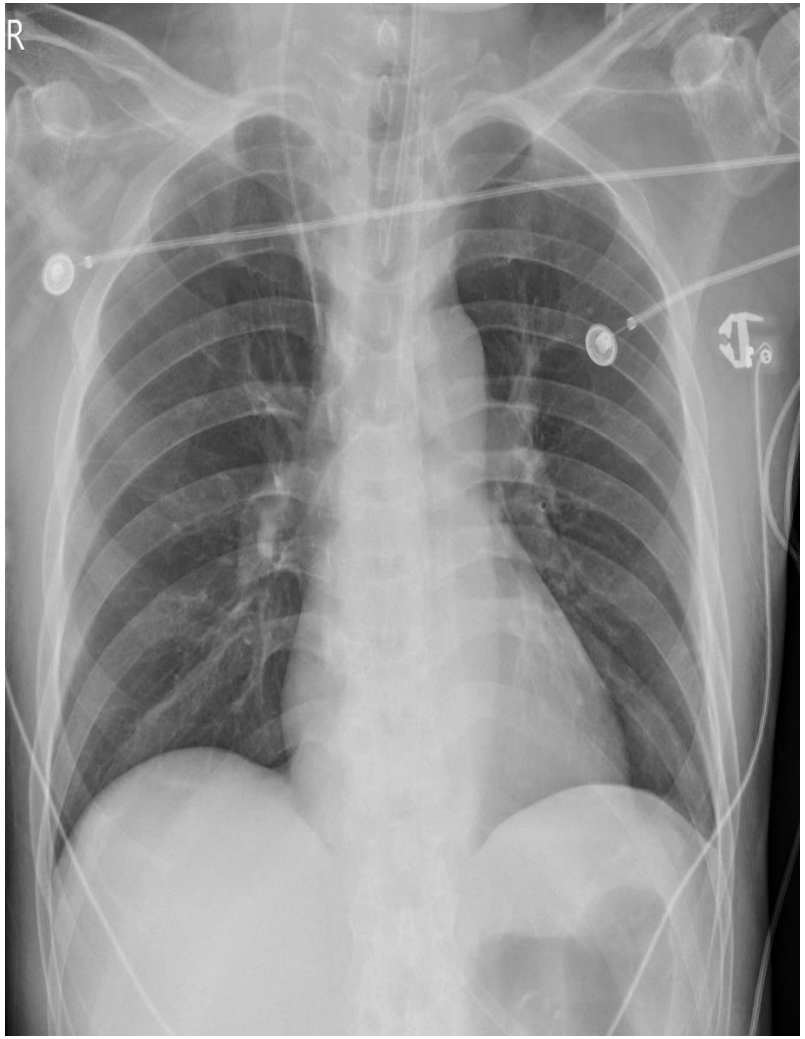
- **The 60/60 sign**

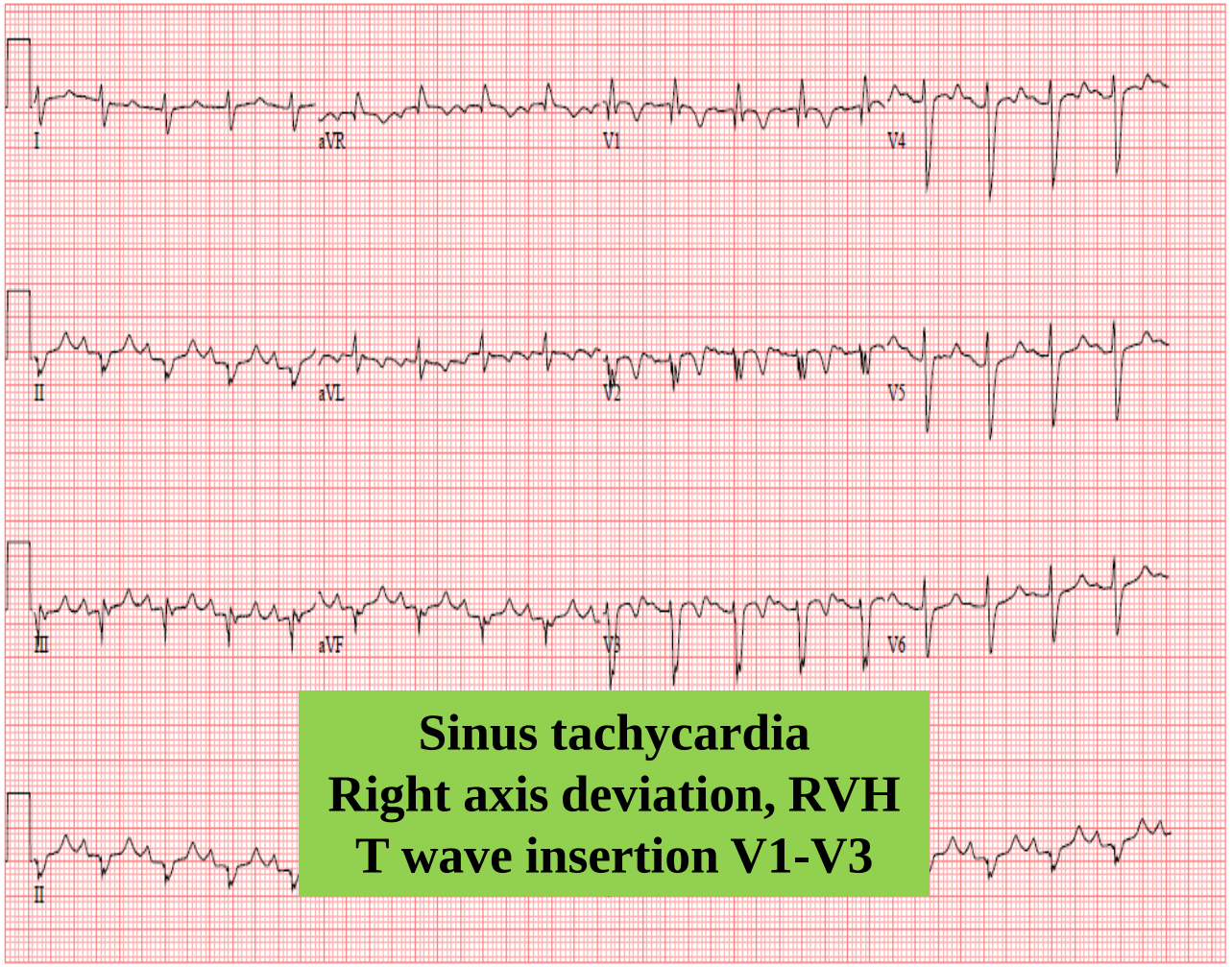
-> RV acceleration time of ≤60 ms
in presence of TVPG ≤60 mm Hg

60/M **Previous healthy**

Dyspnea onset: 1 hours ago

He visited ER with oxygen supplement





60/M Previous healthy

Dyspnea onset: 1 hours ago

He visited ER with oxygen supplement

CBC 18700-18.2-215k,

PT 13.6s (1.22 INR) aPTT 27.1 s

BUN/Cr 24/1.80mg/dL Protein/albumin 7.1/4.2 g/dL

AST/ALT 27/36IU/L CRP 5.37mg/dL

Na/K/Cl/CO2 139/4.6/103/9.3 nmol/L

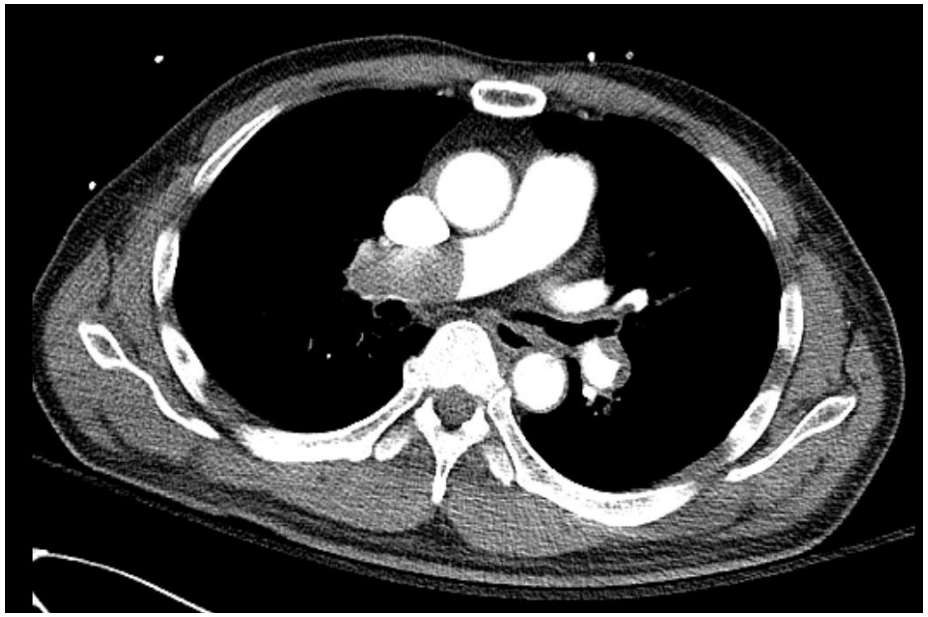
BNP 351 pg/mL

CK-MB 12.1 ng/mL (0-5) Tn-I 0.18 ng/mL (<1.5)

D-dimer 8.05 ug/mL (<0.5)

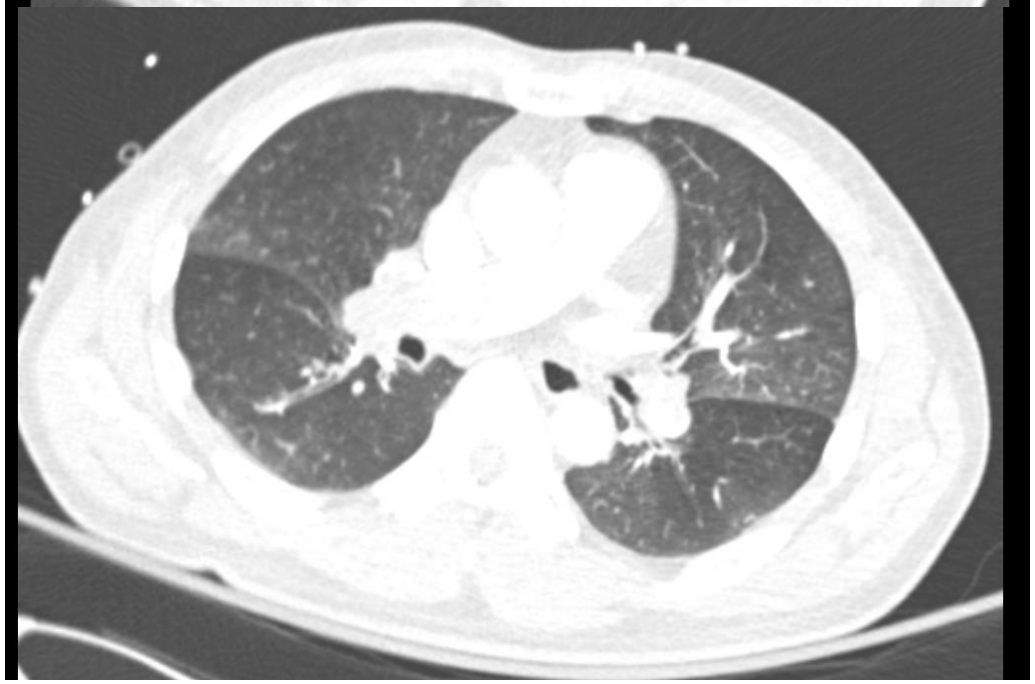
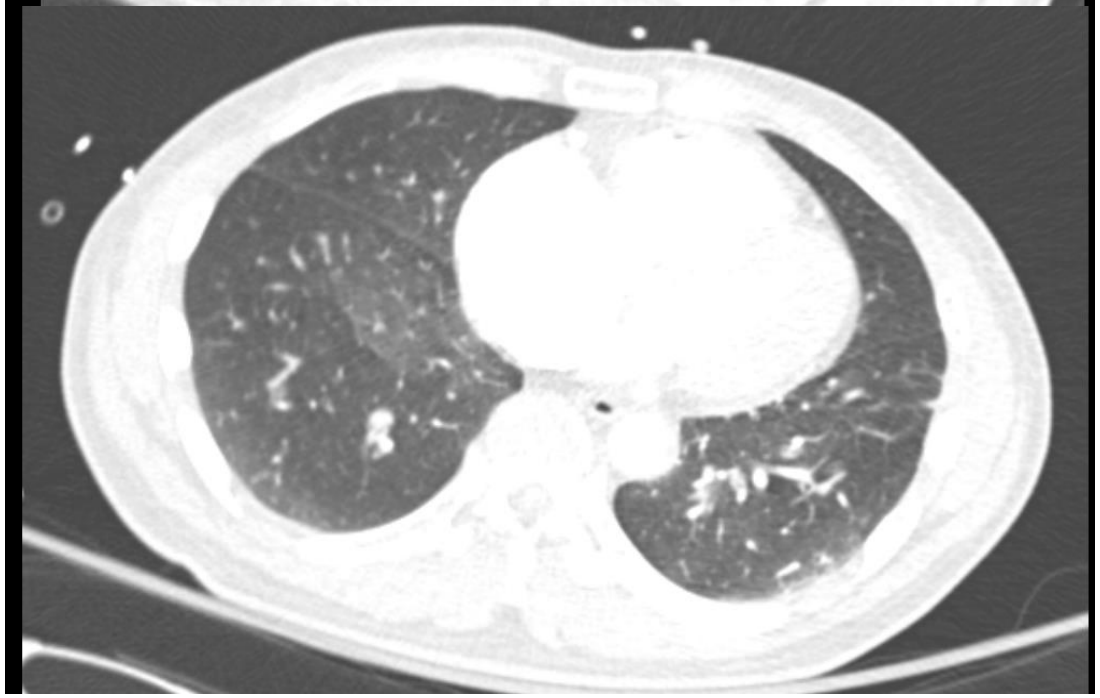
ABGA 7.29-25 mmHg-59.0 mmHg-12.0 mmEq/L (BE -12.2), SaO2 87% (RM 15 L/m)

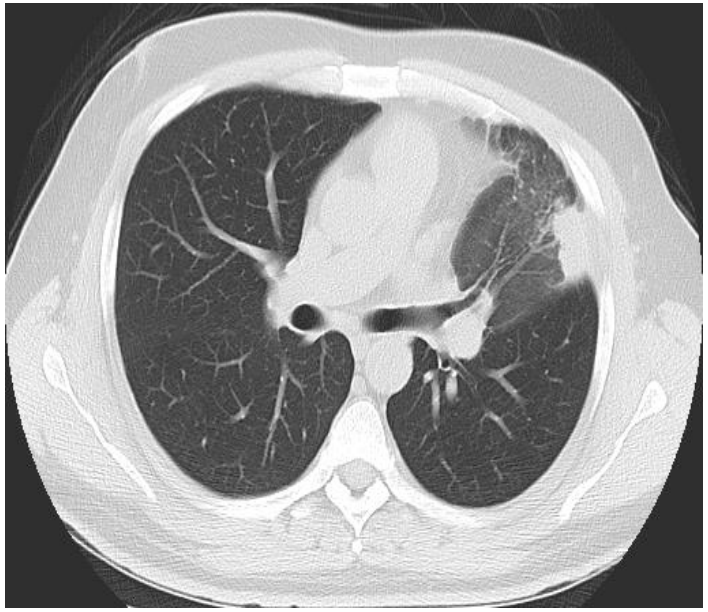
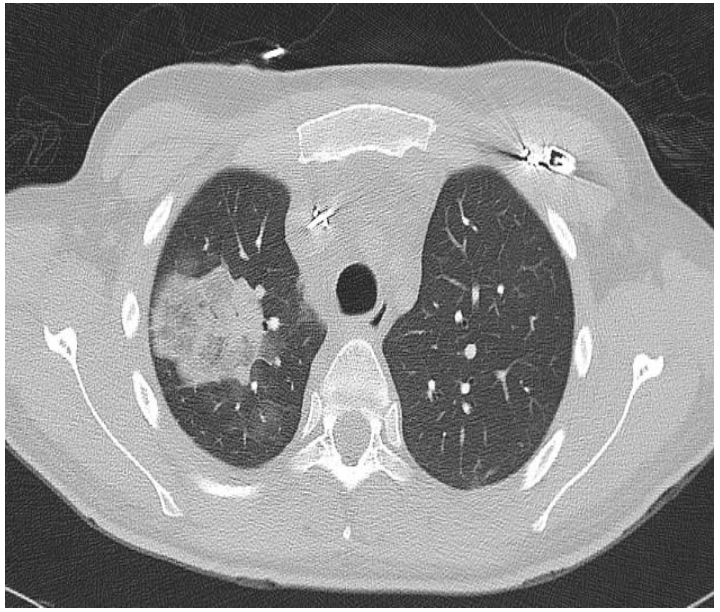
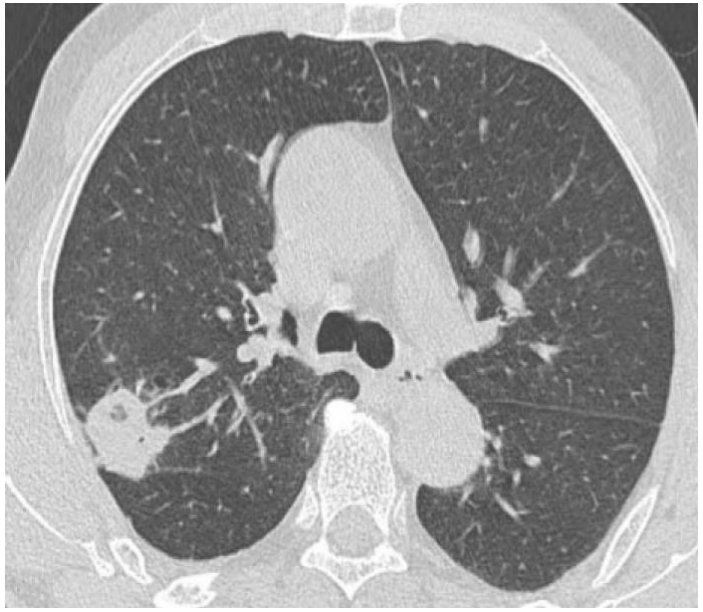
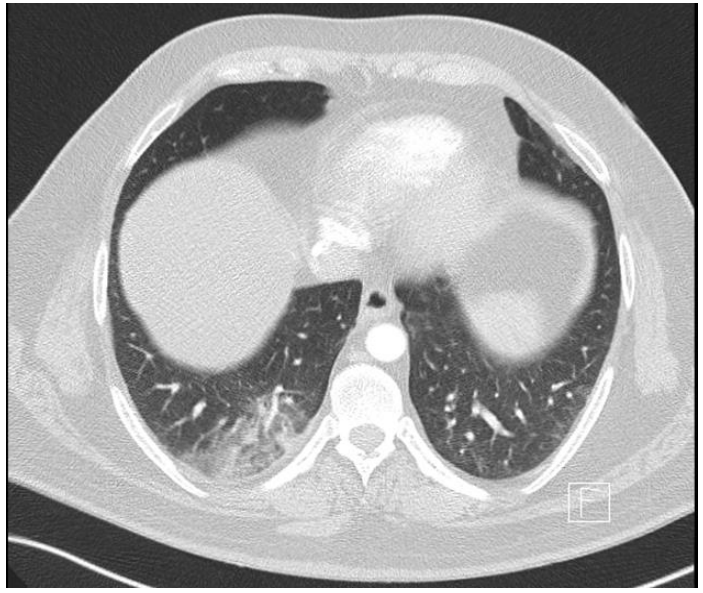
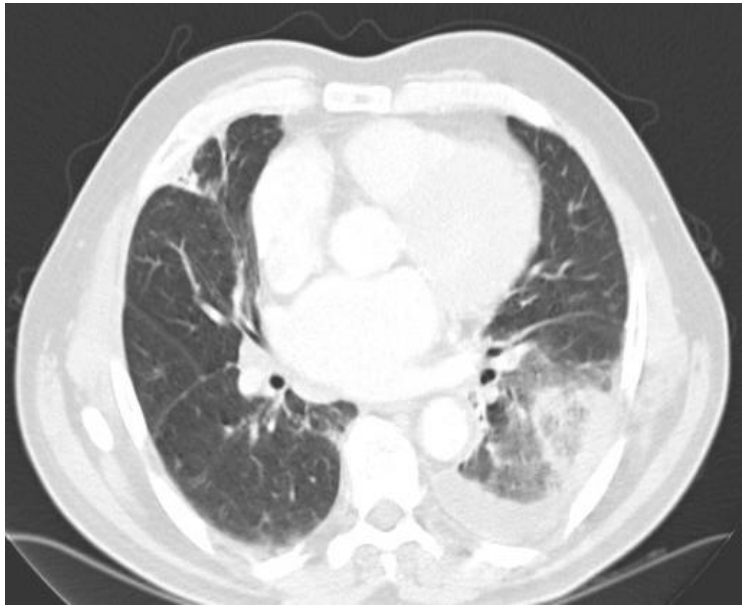
Lactic acid 4.9 mmol/L

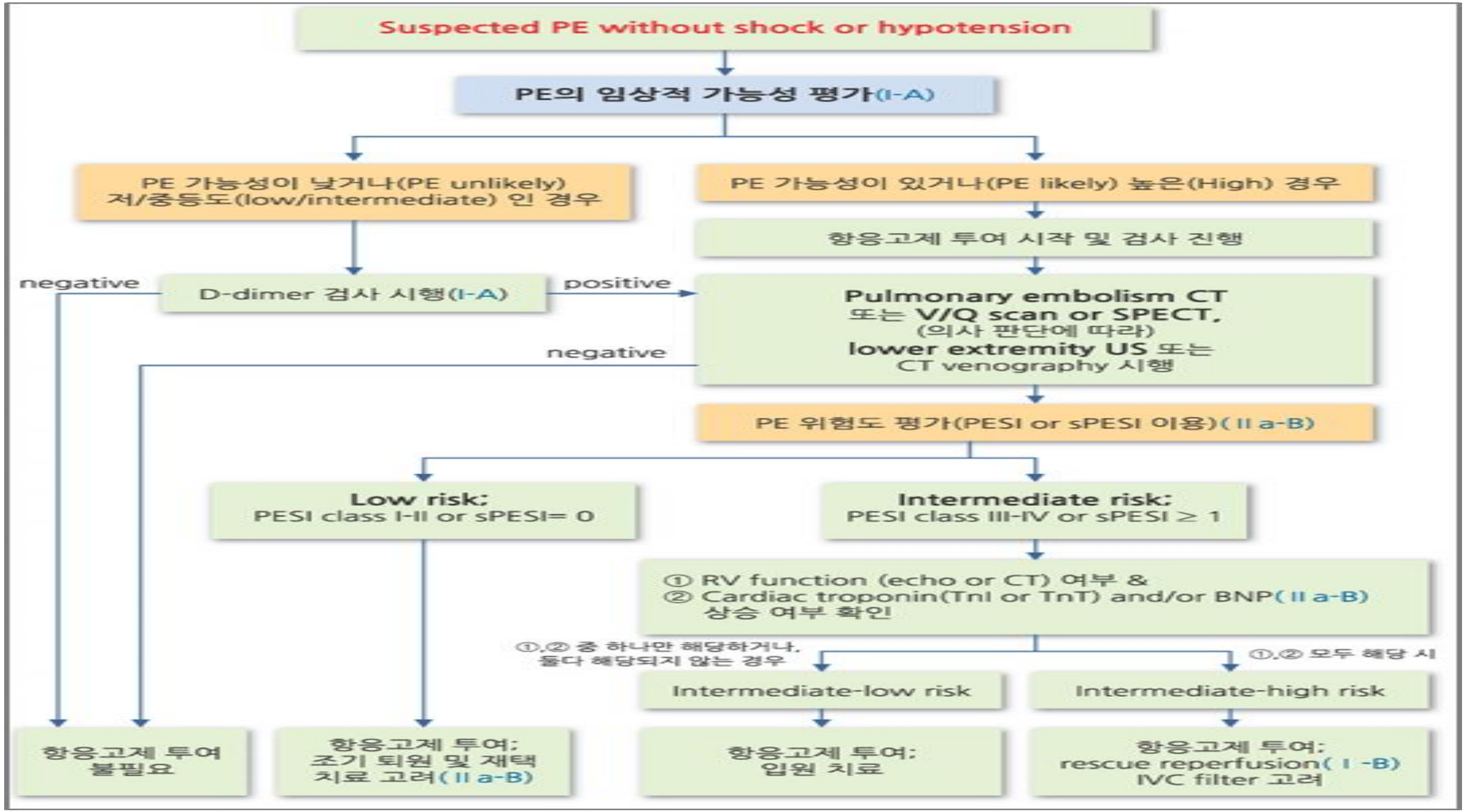


11:16	RM 15L/m ER 내원
11:32	Portable echo 시행
11:38	Intubation
12:30	BP 54/37 mmHg → NE 0.04 ug/kg/min → 0.32 ug/kg/min → N/S 500 cc
13:23	Heparin 5000 IV bolus → IV continuous infusion
13:52	BP 74/49 PR 107 SpO2 98% NE 0.04ug/kg/min
14:18	Chest CT and lower extremity venography
15:10	t-PA 100mg IV for 2 hr
15:52	VF, PEA → CPR, defibrillation
18:30	VA ECMO insertion
23:40	Emergency embolectomy
POD #8	Discharge



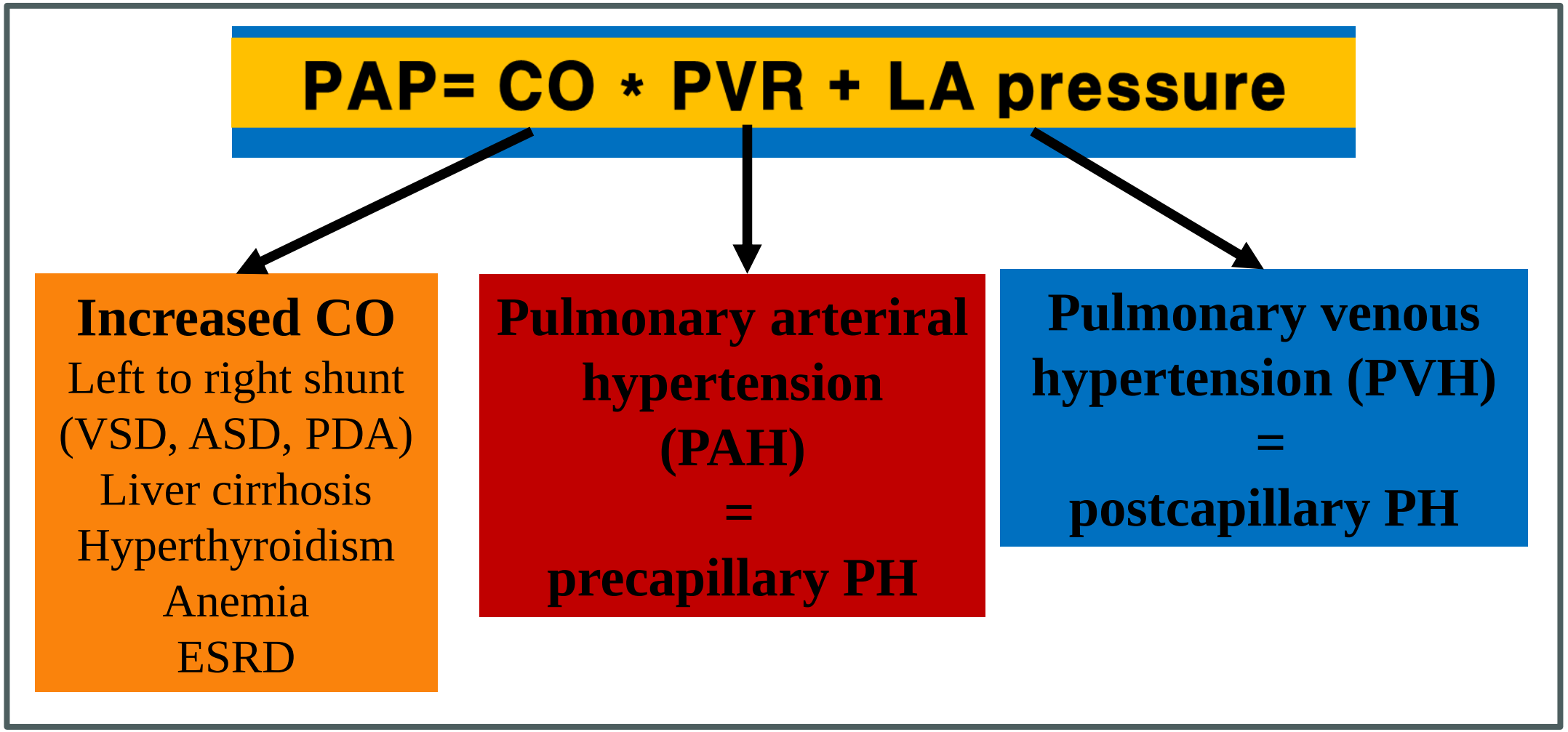








Suspected PE without shock or hypotension
PE의 임상적 가능성 평가(I-A)
PE 가능성이 낮거나(PE unlikely)
저/중등도(low/intermediate) 인 경우
PE 가능성이 있거나(PE likely) 높은(High) 경우
항응고제 투여 시작 및 검사 진행
negative
D-dimer 검사 시행(I-A)
positive
Pulmonary embolism CT
또는 V/Q scan or SPECT,
(의사 판단에 따라)
lower extremity US 또는
CT venography 시행
negative
PE 위험도 평가(PESI or sPESI 이용)(II a-B)
Low risk;
PESI class I-II or sPESI= 0
Intermediate risk;
PESI class III-IV or sPESI ≥ 1
① RV function (echo or CT) 여부 &
② Cardiac troponin(TnI or TnT) and/or BNP(II a-B)
상승 여부 확인
①,② 중 하나만 해당하거나,
둘다 해당되지 않는 경우
①,② 모두 해당 시
Intermediate-low risk
Intermediate-high risk
항응고제 투여
불필요
항응고제 투여;
조기 퇴원 및 재택
치료 고려(II a-B)
항응고제 투여;
입원 치료
항응고제 투여;
rescue reperfusion(I -B)
IVC filter 고려

Anticoagulation in VTE treatment

	Initial (0-21 days)	Long-term (~3 months)	Extension (>3 months)
Conventional	IV or SC UFH or SC LMWH SC fondaparinux	VKA with PT monitoring	
Mono-therapy	Rivaroxaban 15mg bid (3weeks)	Rivaroxaban 20mg qd	Rivaroxaban 10mg qd
	Apixaban 10mg bid (1 week)	Apixaban 5mg bid	Apixaban 2.5mg bid
Switch Therapy	IV UFH or SC LMWH (1 week)	Dabigatran 150mg bid	
		Edoxaban 60mg qd (30mg qd if Ccr 30-50 ml/m or body weight < 60kg)	

Hemodynamic profiles in PH



Hemodynamic criteria of PH

	Haemodynamic characteristics
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20 mmHg PAWP ⩽15 mmHg PVR >2 WU
Isolated post-capillary PH (ipcPH)	mPAP >20 mmHg PAWP >15 mmHg PVR ⩽2 WU
Combined post- and pre-capillary PH (cpcPH)	mPAP >20 mmHg PAWP >15 mmHg PVR >2 WU
Exercise PH	mPAP/CO slope >3 mmHg/L/min between rest and exercise

mPAP: mean pulmonary arterial pressure; PAWP: pulmonary arterial wedge pressure; PVR: pulmonary vascular resistance; WU: Wood Units; CO: cardiac output.

Clinical classification of PH

PULMONARY HYPERTENSION

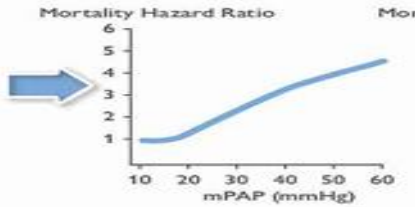
Prevalence

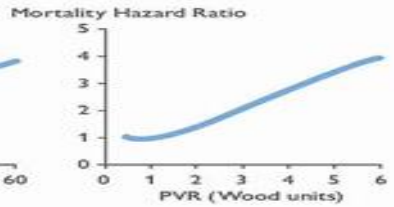
1%

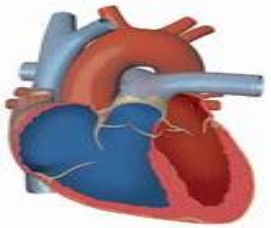
Global population

Pulmonary congestion in post-capillary PH

Pulmonary vascular disease / obstruction in pre-capillary PH

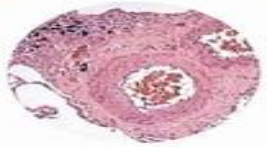
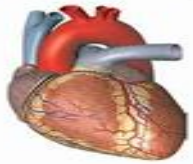






Right heart failure

CLINICAL CLASSIFICATION

Pulmonary arterial hypertension (PAH)	PH associated with left heart disease	PH associated with lung disease	PH associated with pulmonary artery obstructions	PH with unclear and/or multifactorial mechanisms
				?
• Idiopathic/heritable • Associated conditions	• IpcPH • CpcPH	• Non-severe PH • Severe PH	• CTEPH • Other pulmonary obstructions	• Haematologic disorders • Systemic disorders

PREVALENCE

Rare	Very common	Common	Rare	Rare
				

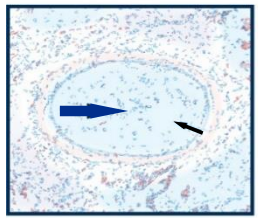
Classification of PH

Group 1 **Pulmonary arterial**	**1.1 Idiopathic** **1.1.1 Long-term responders to calcium channel blockers**
	1.5 PAH with features of venous/capillary (PVOD/PCH) involvement
	1.6 Persistent PH of the newborn

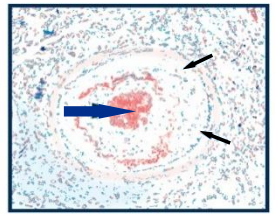
Group 2 **PH associated with left heart disease**	**2.1 Heart failure:** **2.1.1 with preserved ejection fraction** **2.1.2with reduced or mildly reduced ejection fraction** **2.1.3 cardiomyopathies with specific aetiologies** **2.2 Valvular heart disease:** **2.2.1 aortic valve disease**
Group 3 **PH associated with lung diseases and/or hypoxia**	**3.1 COPD and/or emphysema** **3.2 Interstitial lung disease** **3.3 Combined pulmonary fibrosis and emphysema** **3.4 Other parenchymal lung diseases+** **3.5 Nonparenchymal restrictive diseases:** **3.5.1 hypoventilation syndromes** **3.5.2 pneumonectomy** **3.6 Hypoxia without lung disease (e.g. high altitude)** **3.7 Developmental lung diseases**
pulmonary artery obstructions	**obstructions**
Group 5 **PH with unclear and /or multifactorial mechanisms**	5.1Haematological disorders 5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis and neurofibromatosistype1 5.3Metabolicdisorders 5.4Chronic renal failure with or without haemodialysis 5.5 Pulmonary tumour thrombotic microangiopathy 5.6Fibrosin gmediastinitis 5.7Complex congenita lheartdisease

Vascular changes in IPAH

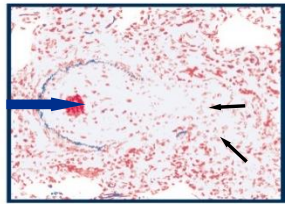
특발성폐동맥고혈압은 직경 1mm 이하의 폐동맥에서 발생



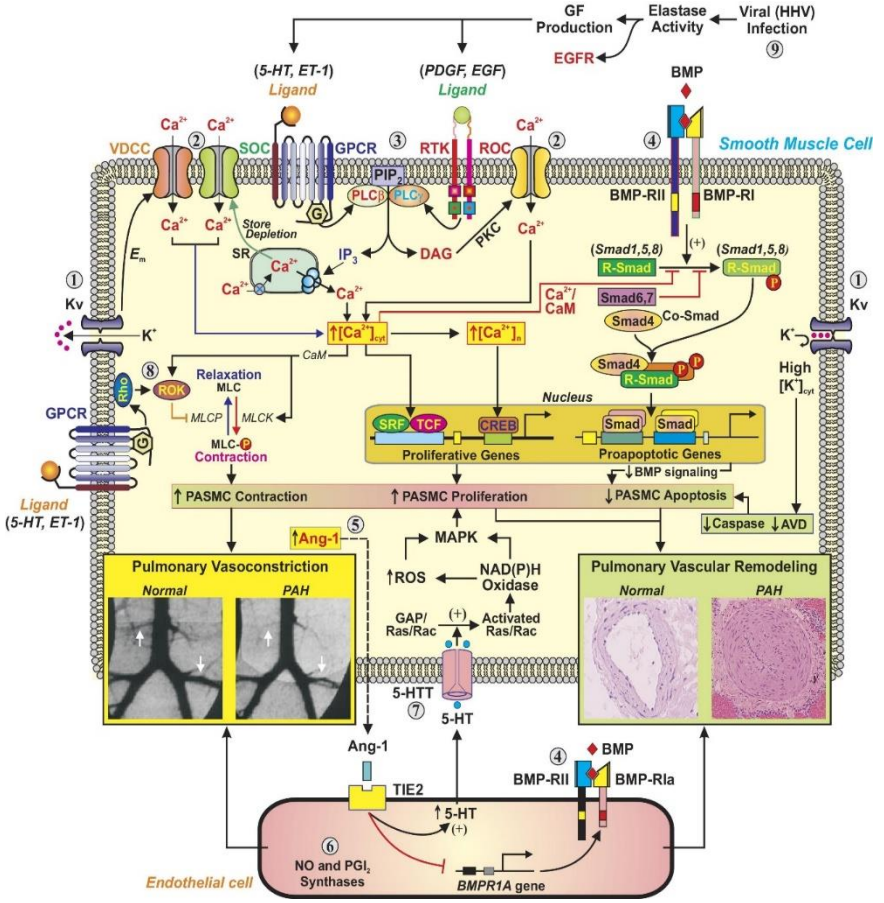
Intimal proliferation



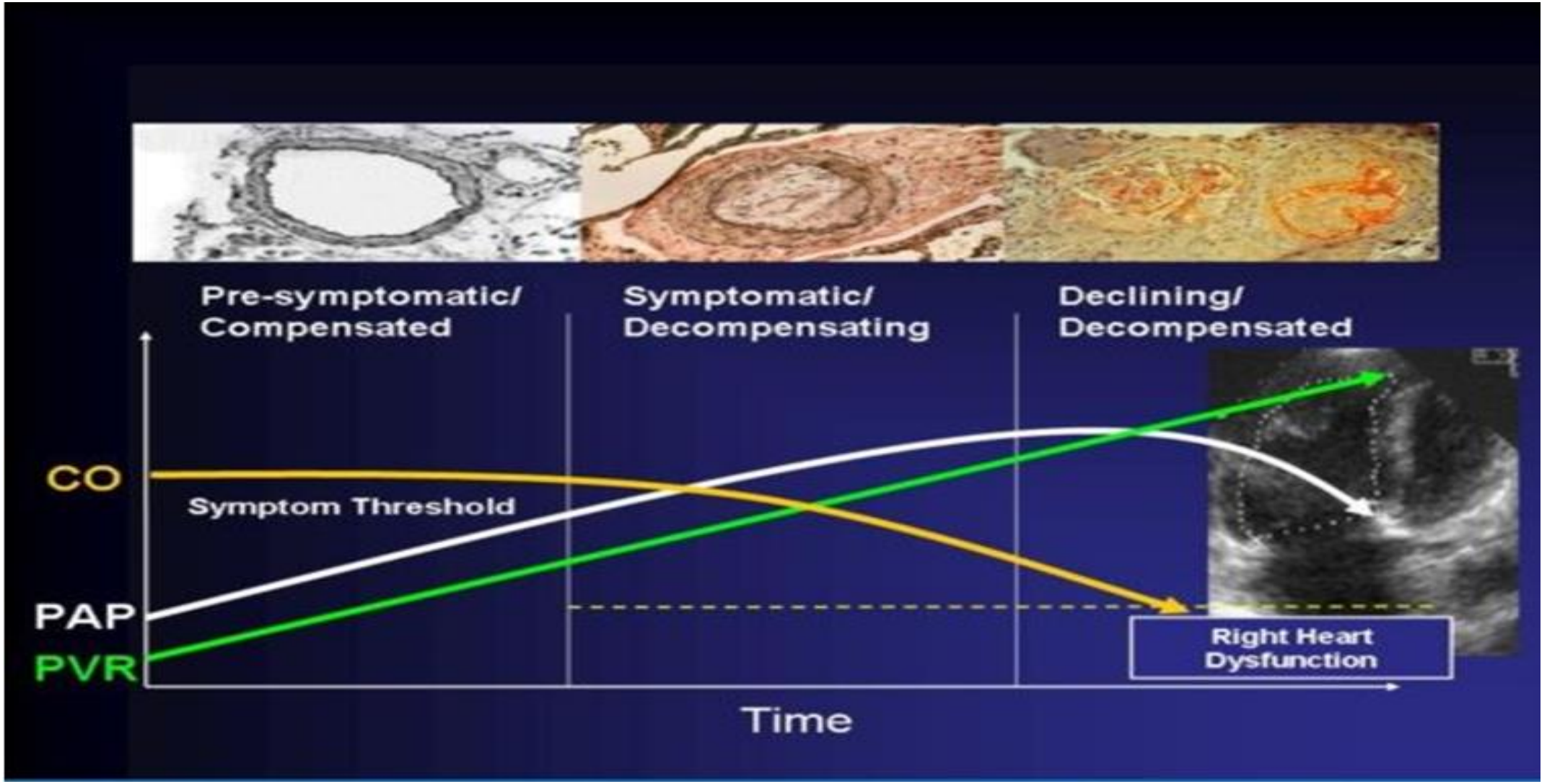
Medial hypertrophy



Plexiform lesions



Evolution of PAH



Symptoms in patients with PH

Early

Symptoms

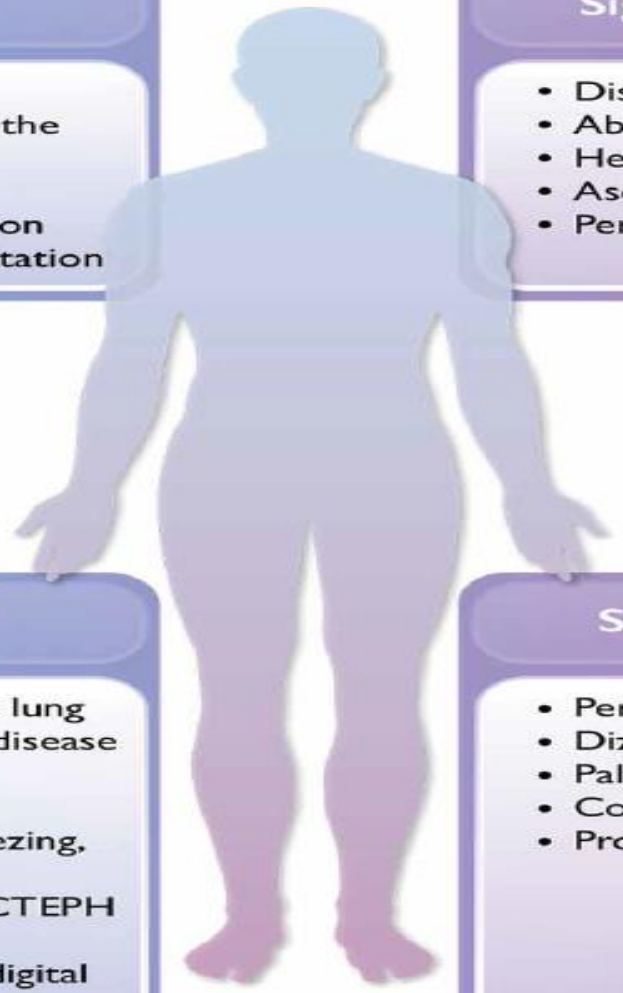
- Dyspnoea on exertion (WHO-FC)
- Fatigue and rapid exhaustion
- Dyspnoea when bending forward (bendopnoea)
- Palpitations
- Haemoptysis
- Exercise-induced abdominal distension and nausea
- Weight gain due to fluid retention
- Syncope (during or shortly after physical exertion)

Late

Rare symptoms due to pulmonary artery dilation[a]

- Exertional chest pain:
  dynamic compression of the left main coronary artery
- Hoarseness (dysphonia):
  compression of the left laryngeal recurrent nerve
  (cardiovocal or Ortner´s syndrome)
- Shortness of breath, wheezing, cough, lower respiratory tract infection, atelectasis:
  compression of the bronchi

Clinical signs in patients with PH



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

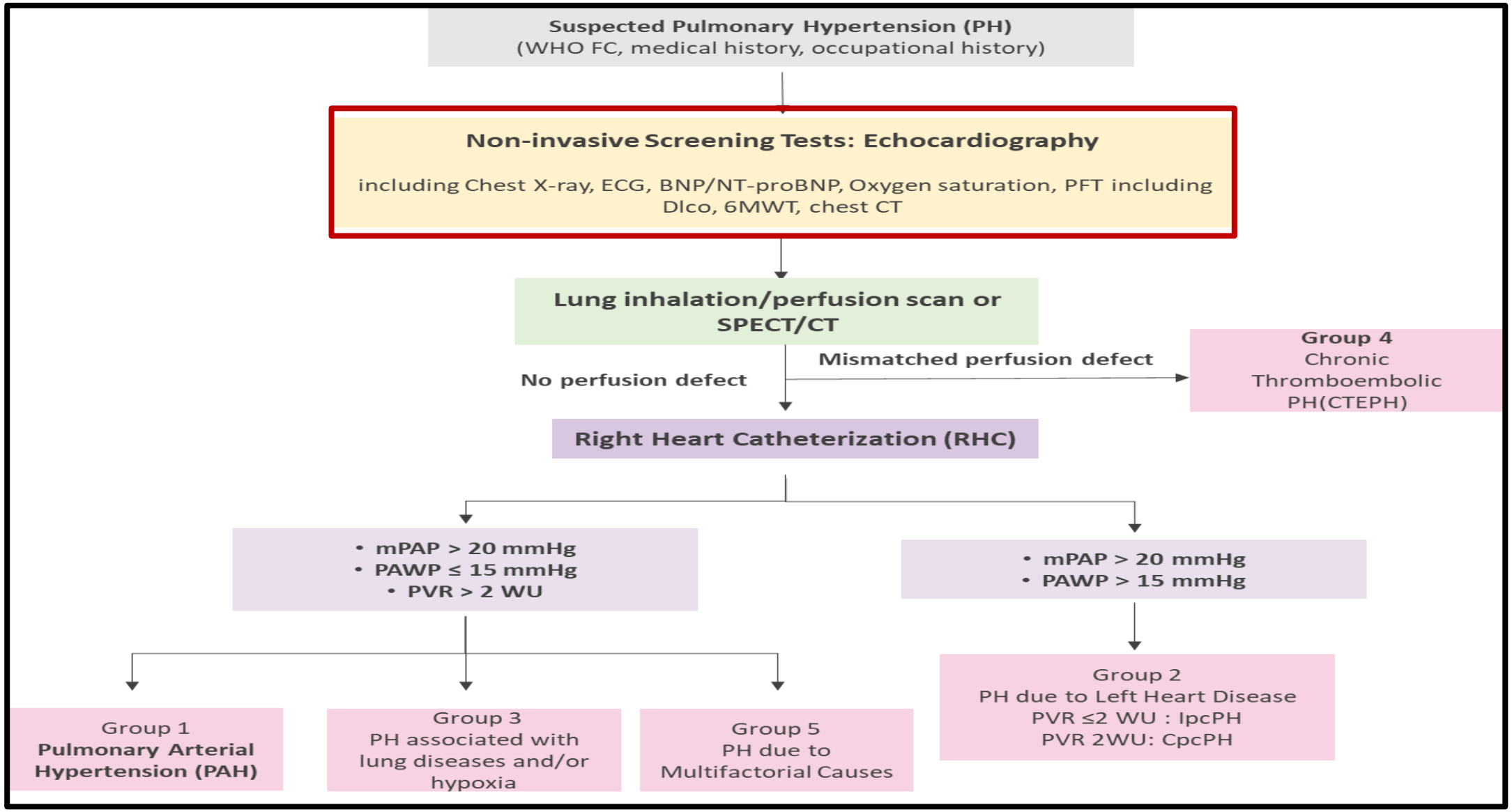
Signs pointing towards underlying cause of PH

- 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

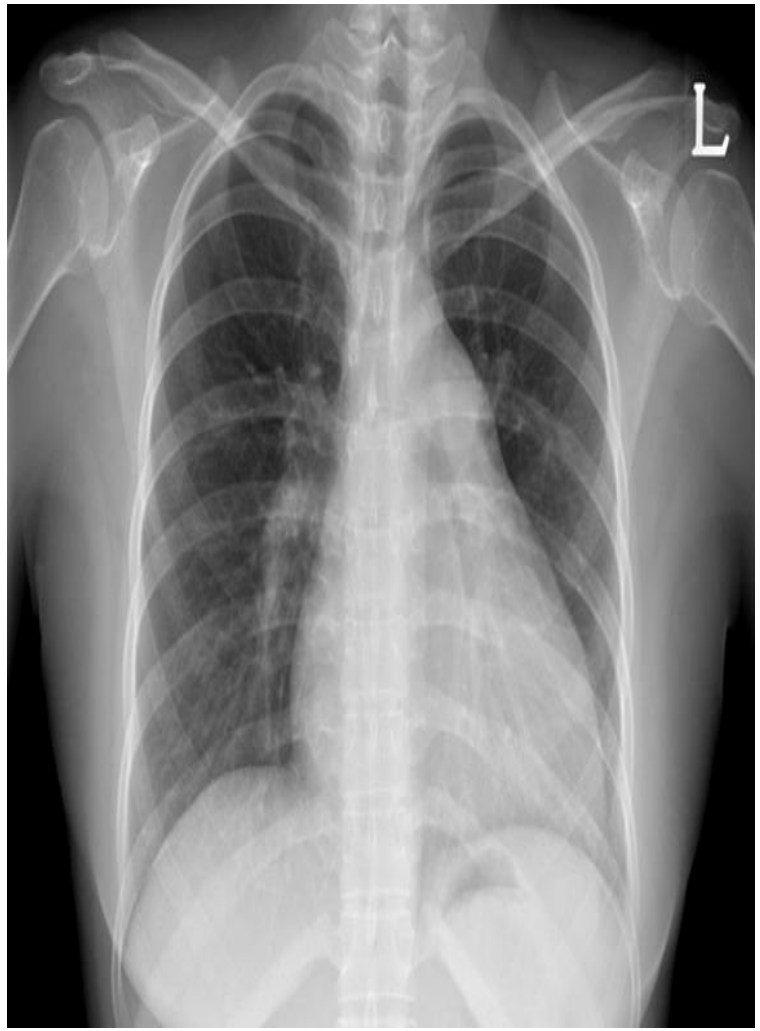
- Peripheral cyanosis (blue lips and tips)
- Dizziness
- Pallor
- Cool extremities
- Prolonged capillary refill

Diagnostic algorithm of PH

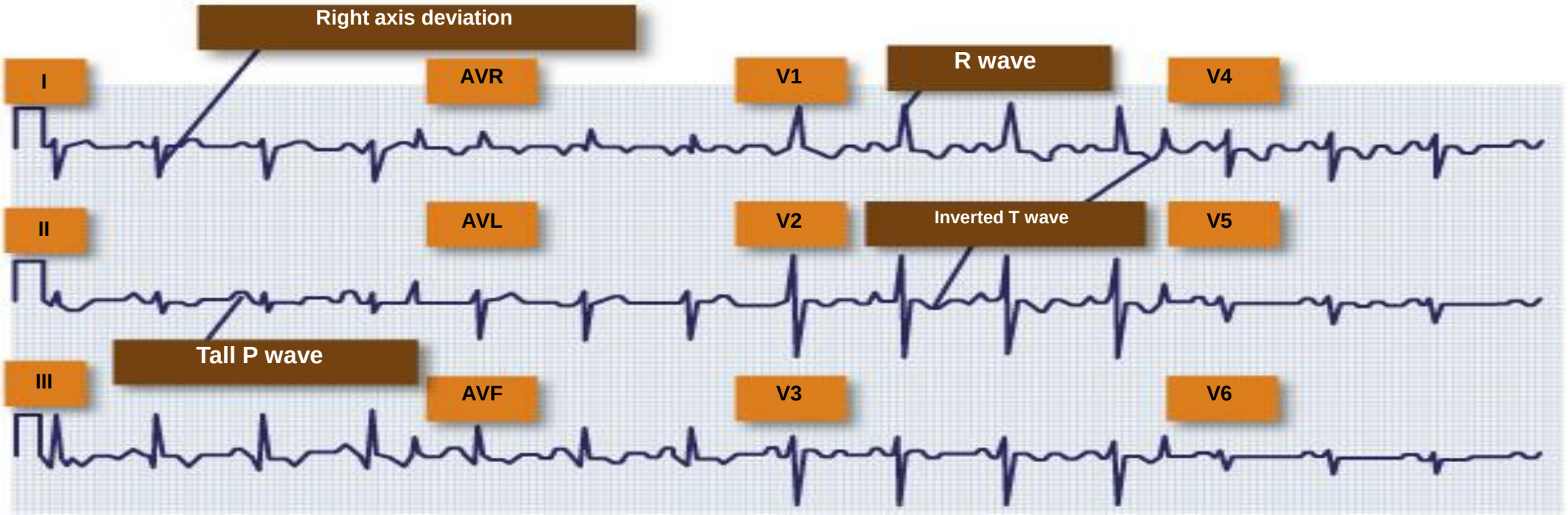


Radiologic signs of PH

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)	Reticular opacification (fibrotic lung disease)
	Left ventricular dilation	

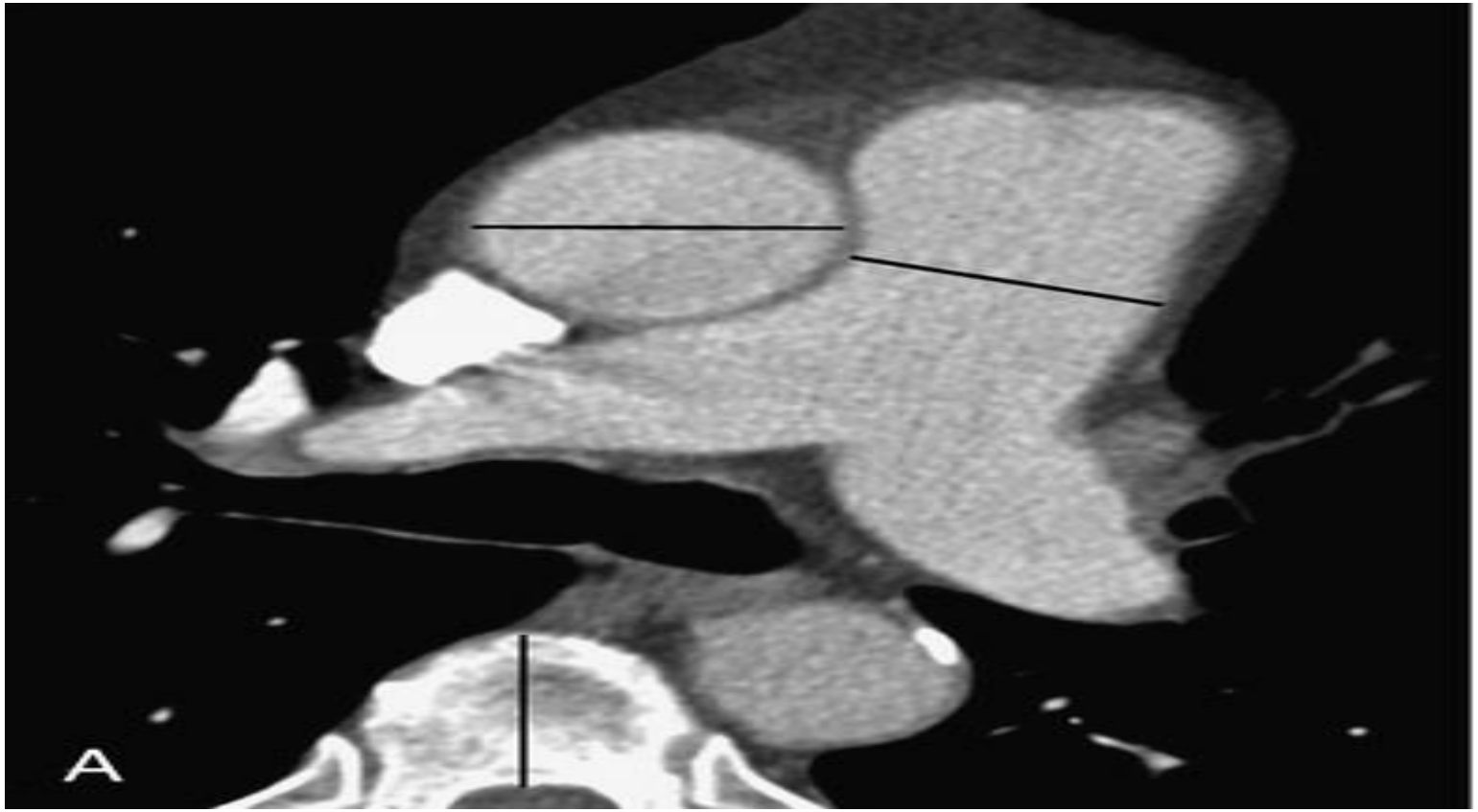


ECG in patients with PH



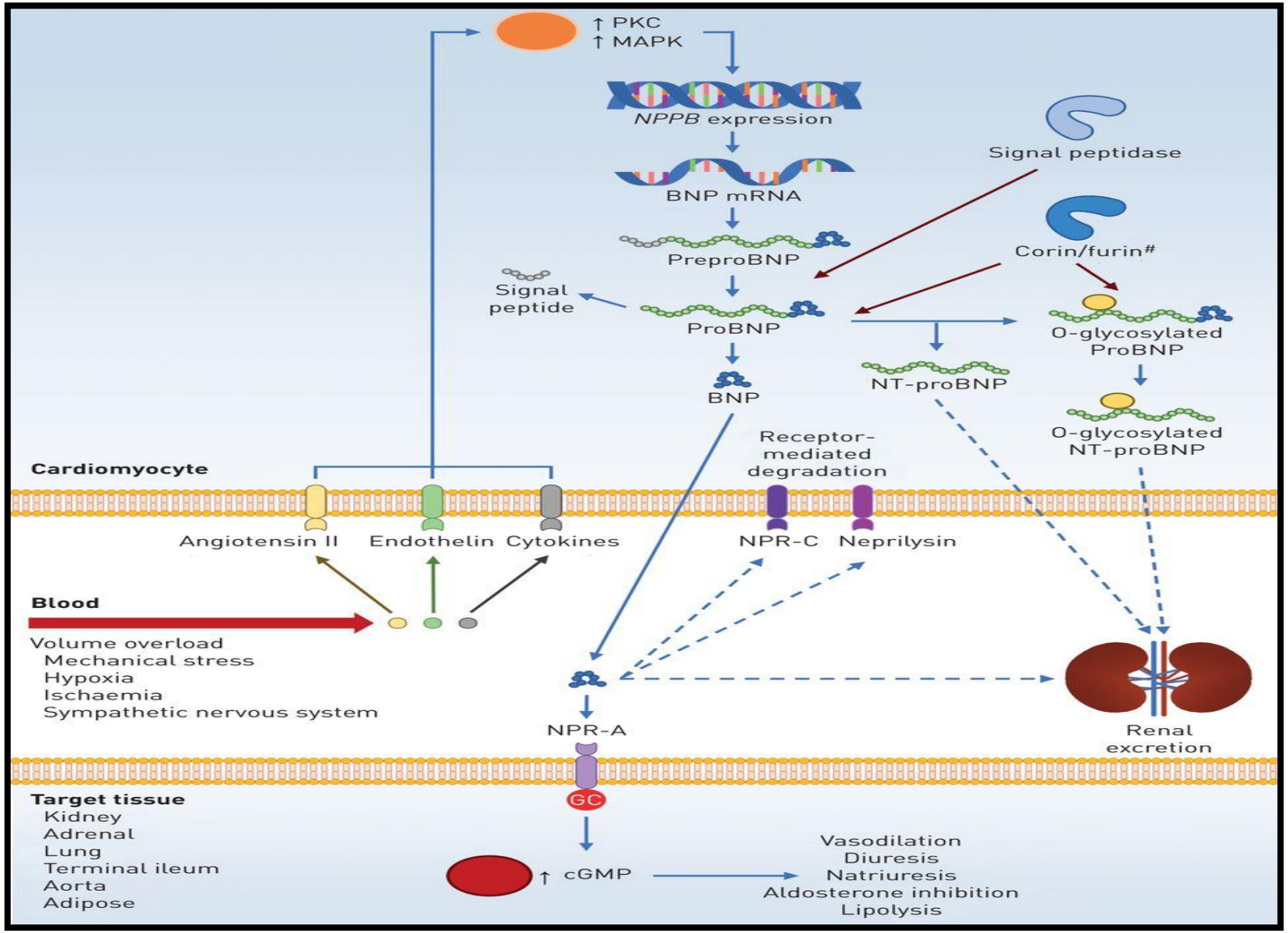
- P pulmonale (P>0.25 mV in lead II)
- Right or sagittal axis deviation (QRS axis >90° or indeterminable)
- RV hypertrophy (R/S >1, with R >0.5 mV in V1; R in V1 + S in lead V5 >1 mV)
- Right bundle branch block—complete or incomplete (qR or rSR patterns in V1)
- RV strain pattern[a] (ST depression/T-wave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

Chest CT for detecting PH



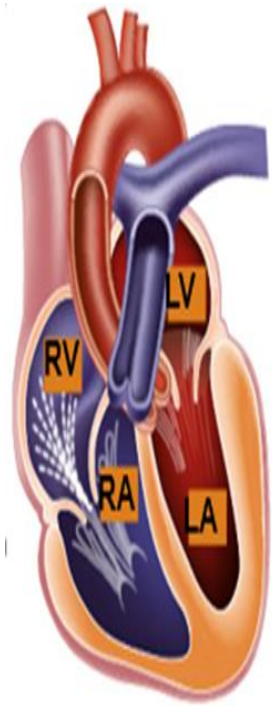
The ratio of the main pulmonary artery to ascending aorta diameter could predict PH , with a ratio >1 (range 0.9–1.1).

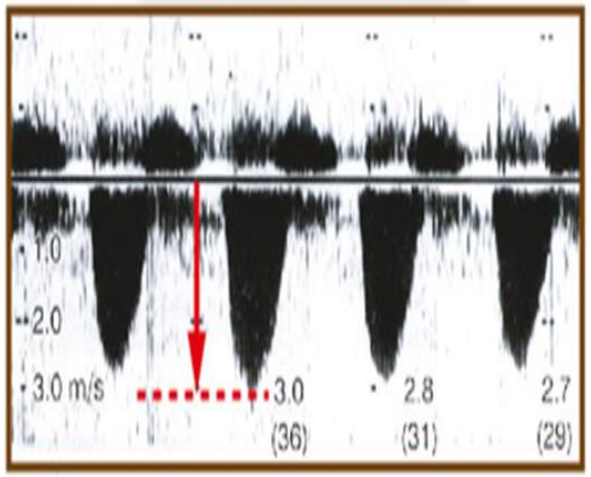
BNP/NT-pro BNP



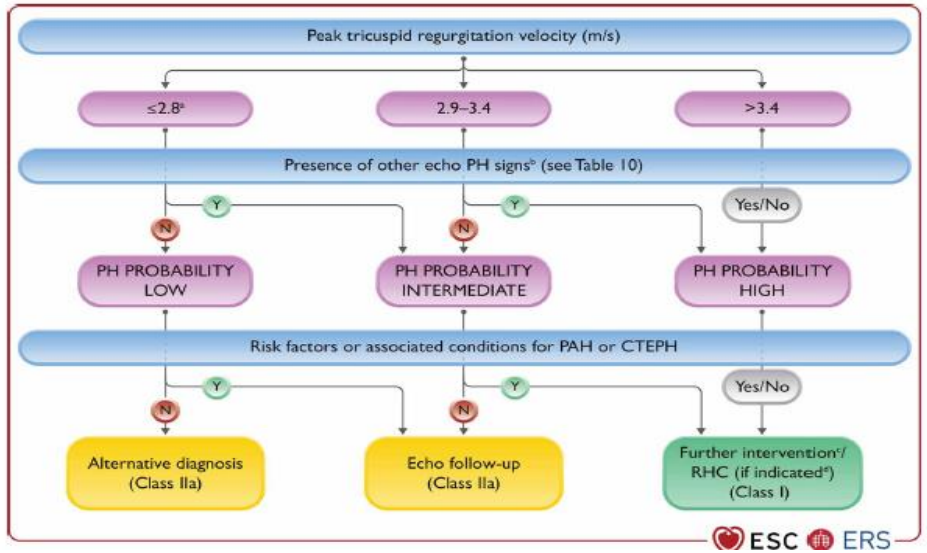
Lewis RA, et al. Eur Respir Rev. 2020;29(156):200009.

Echocardiographic probability of PH



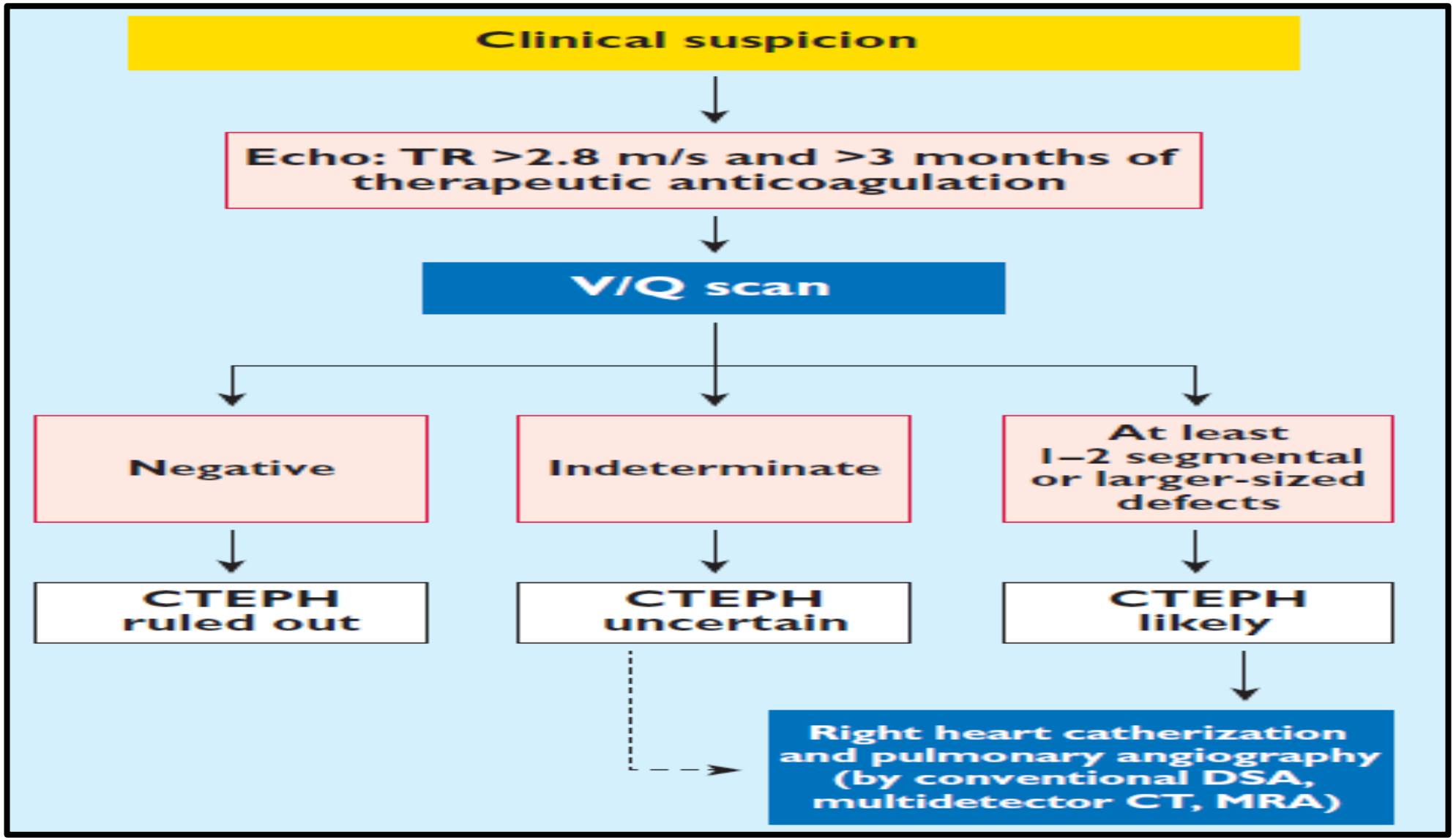


Syst PAP = right ventricular systolic pressure
(in absence of pulmonary outflow obstruction)
$RVSP = 4v^2 + RAP^*$



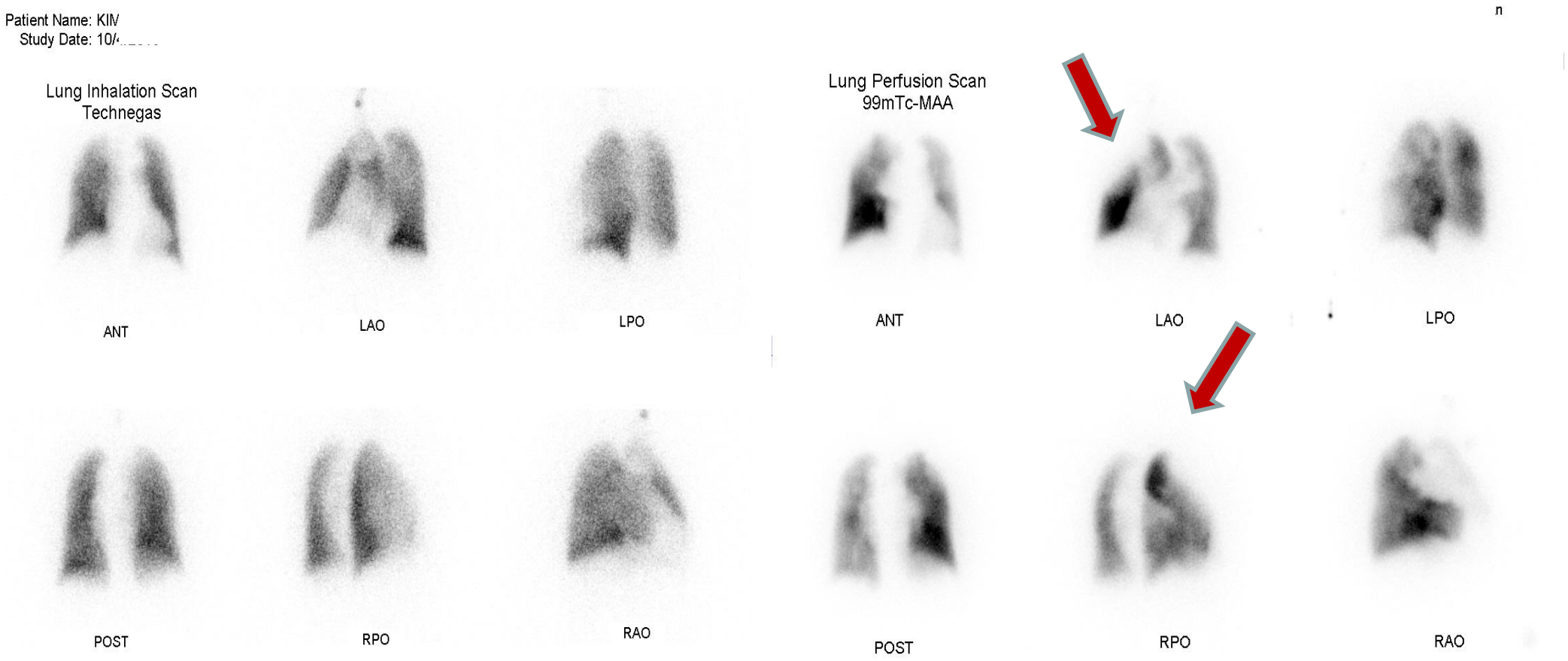
A: The ventricles[a]	B: Pulmonary artery[a]	C: Inferior vena cava and right atrium[a]
Right ventricle/ left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<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 algorithm of CTEPH

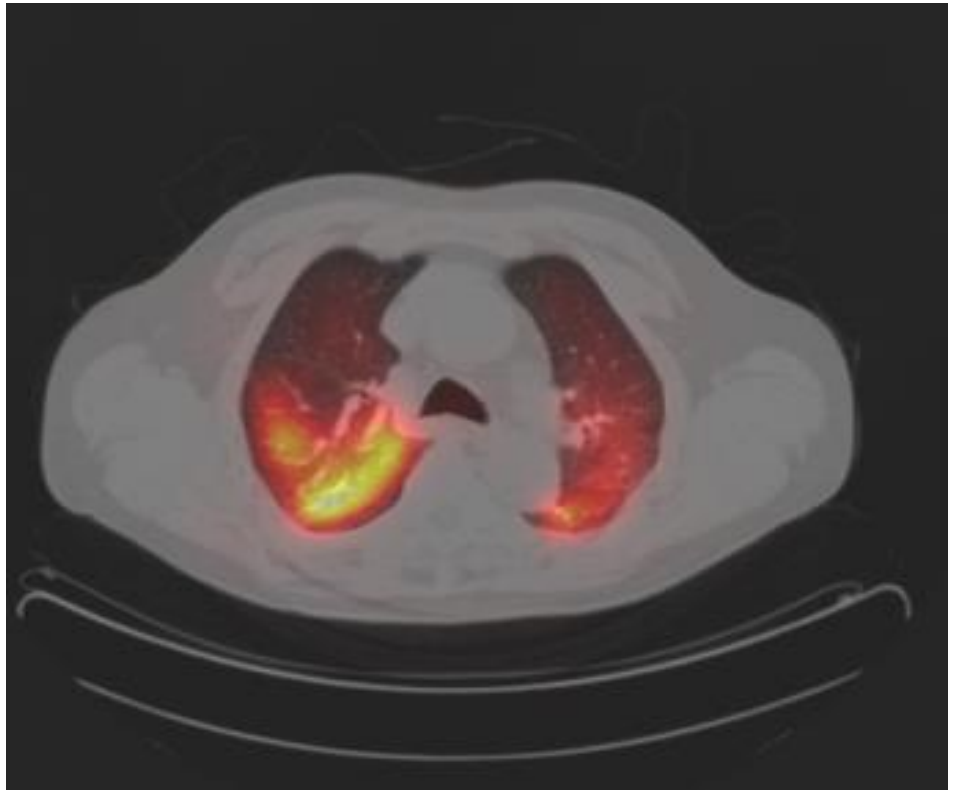


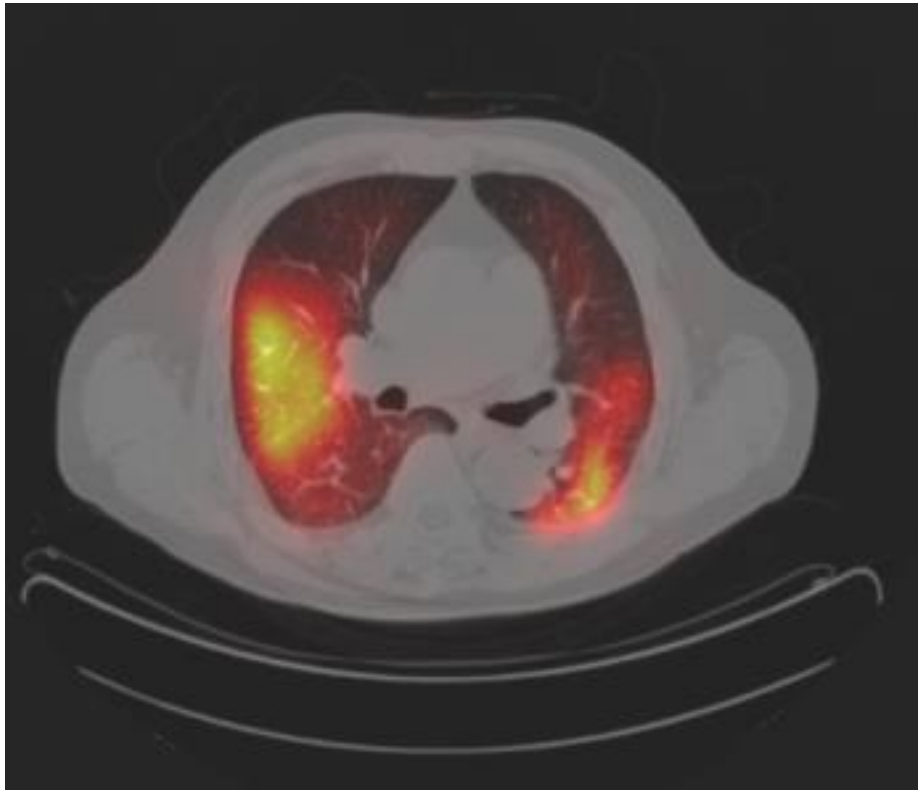
Lung inhalation/perfusion scan

만성혈전색전증에 의한 폐고혈압 감별을 위한 필수 검사



Fusion SPECT/CT





Right heart catheterization

필수 검사

1. 폐고혈압 확진 및 분류

2. 치료 방법 및 약제 선택

3. 폐고혈압 중증도 평가

RHC in patients with PH

- The following variables must be recorded during RHC

  Right atrial pressure

  RV pressure

  PAP (systolic, diastolic, mean)

  PAWP (if inaccurate

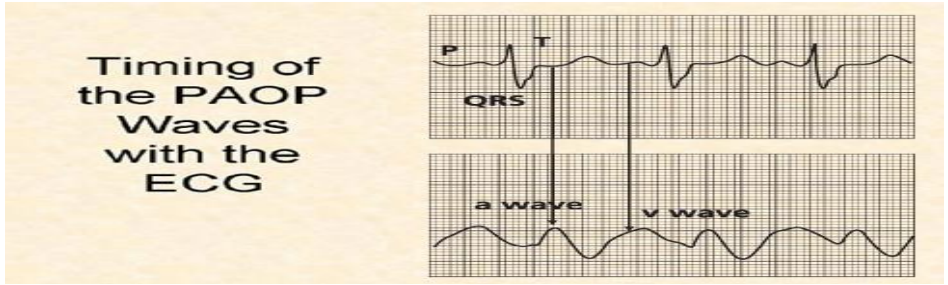
  -> left heart cath. -> measure LVEDP)

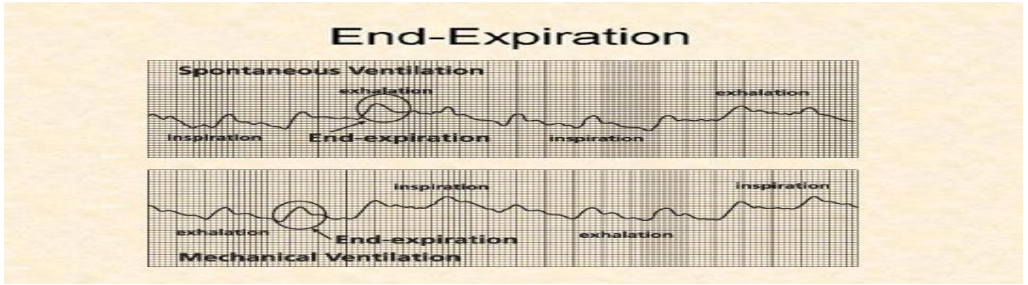
  CO – Thermo-dilution method, Fick method

  Mixed venous oxygen saturation (ScvO2)

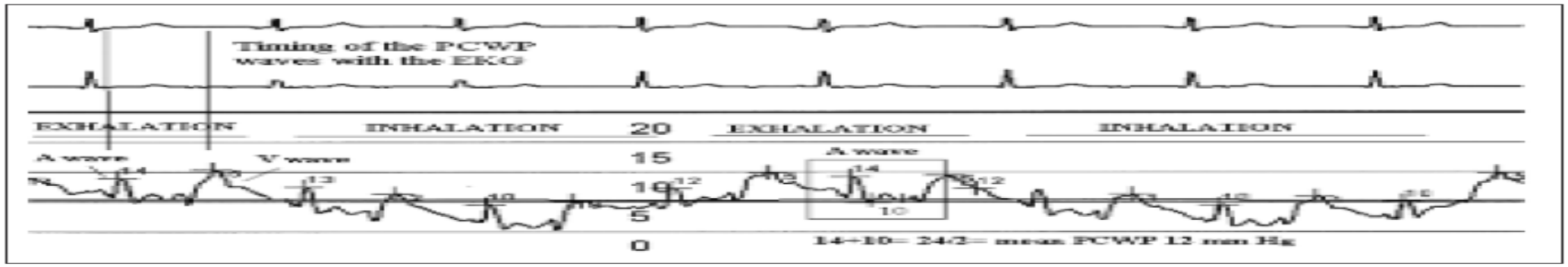
Measurement of PAWP

1) **Identification of the A wave during exhalation**
(right before the beginning of the pressure decline)





2) **Average the top and bottom values of the A wave**



Measurement of cardiac output

❖Fick method

$$CO\ (L/m) = \frac{\text{oxygen consumption (mL } O_2\text{/m)}}{\text{arteriovenous oxygen difference (mL of } O_2\text{/L)}}$$

- **Direct Fick method: gold standard method**

- **Indirect Fick method: inaccurate estimate of O_2 consumption**

❖Thermo-dilution method

- At least 3 measurements within 10% of each other
- limitations in severe TR and congenital heart defect
- **must be performed by trained personel**

25/F IPAH

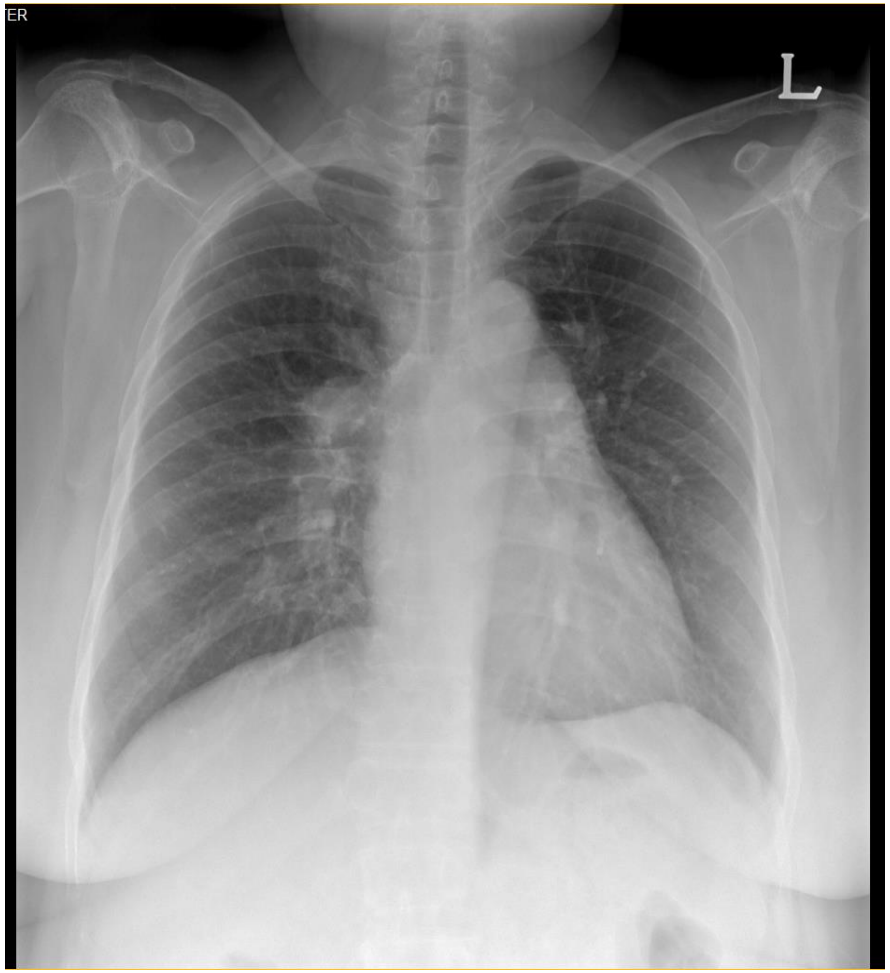
C. C : recurrent syncope, 10 times

P. M. Hx. : none

Height 155 weight 92 BMI 38

BNP 329 pg/ml

6MWT 190m (SpO2 94% -> 94%)



14-FEB-1998 (25 yr)
Female Asian

Room:CV
Loc:112

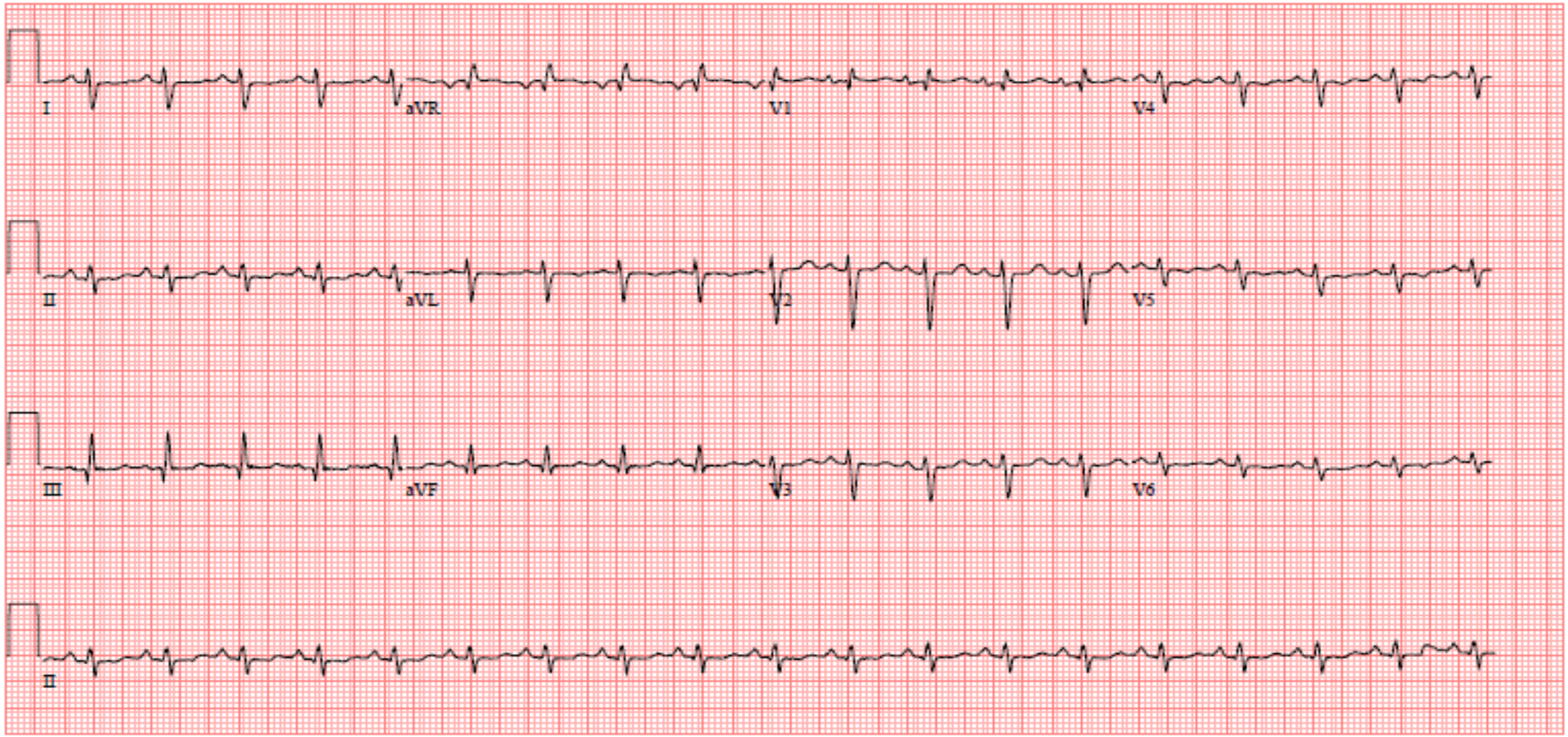
Vent. rate	113	BPM
PR interval	148	ms
QRS duration	94	ms
QT/QTc	346/474	ms
P-R-T axes	45 143	94

Sinus tachycardia
Right axis deviation
Incomplete right bundle branch block
Possible Right ventricular hypertrophy
Nonspecific T wave abnormality
Abnormal ECG
No previous ECGs available

Technician: JHS
Test ind:

Referred by: CV, PARK JIN SEON

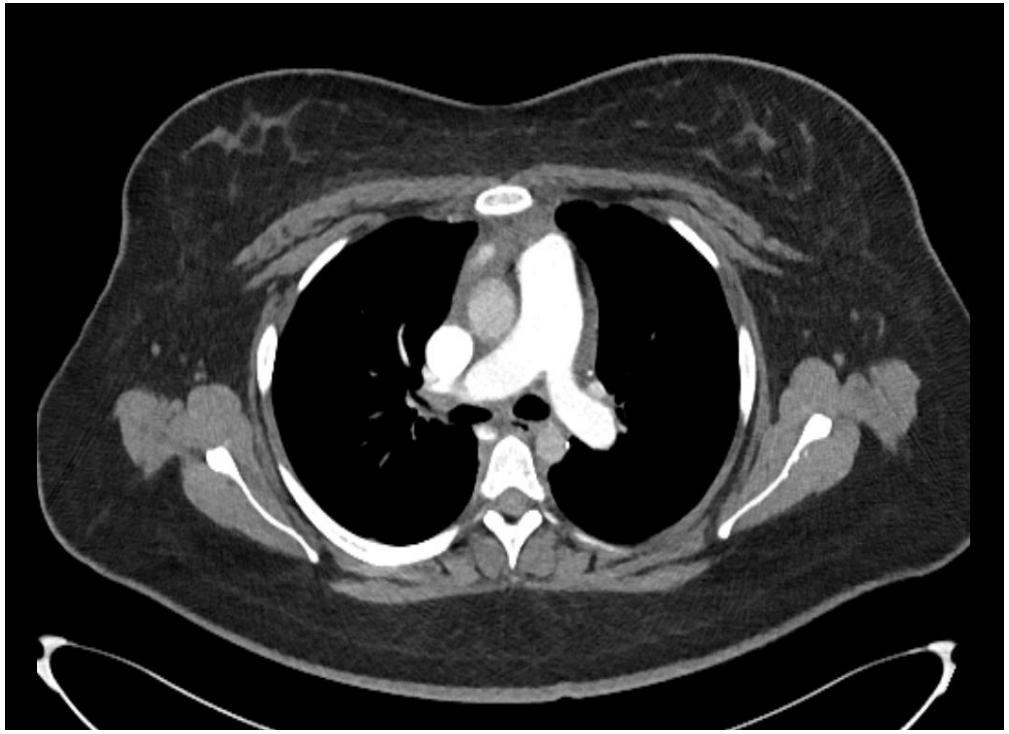
Unconfirmed

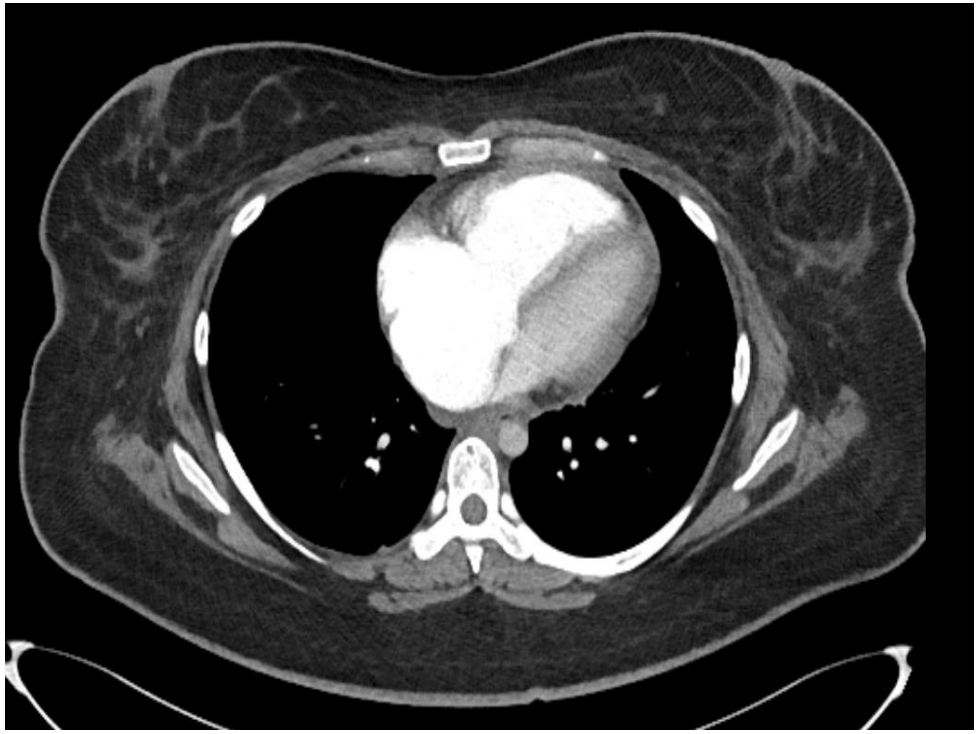


25mm/s 10mm/mV 150Hz 9.0.9 12SL 241 CID: 112

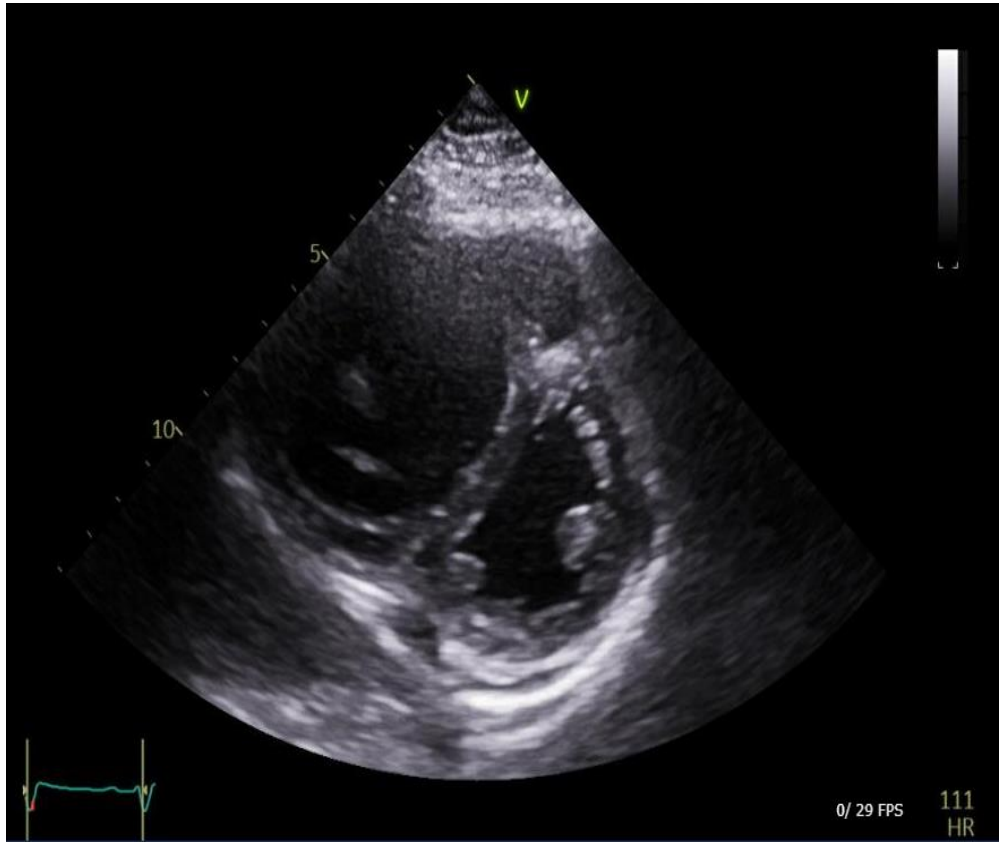
SID: _ EID: EDT: ORDER: 23217655

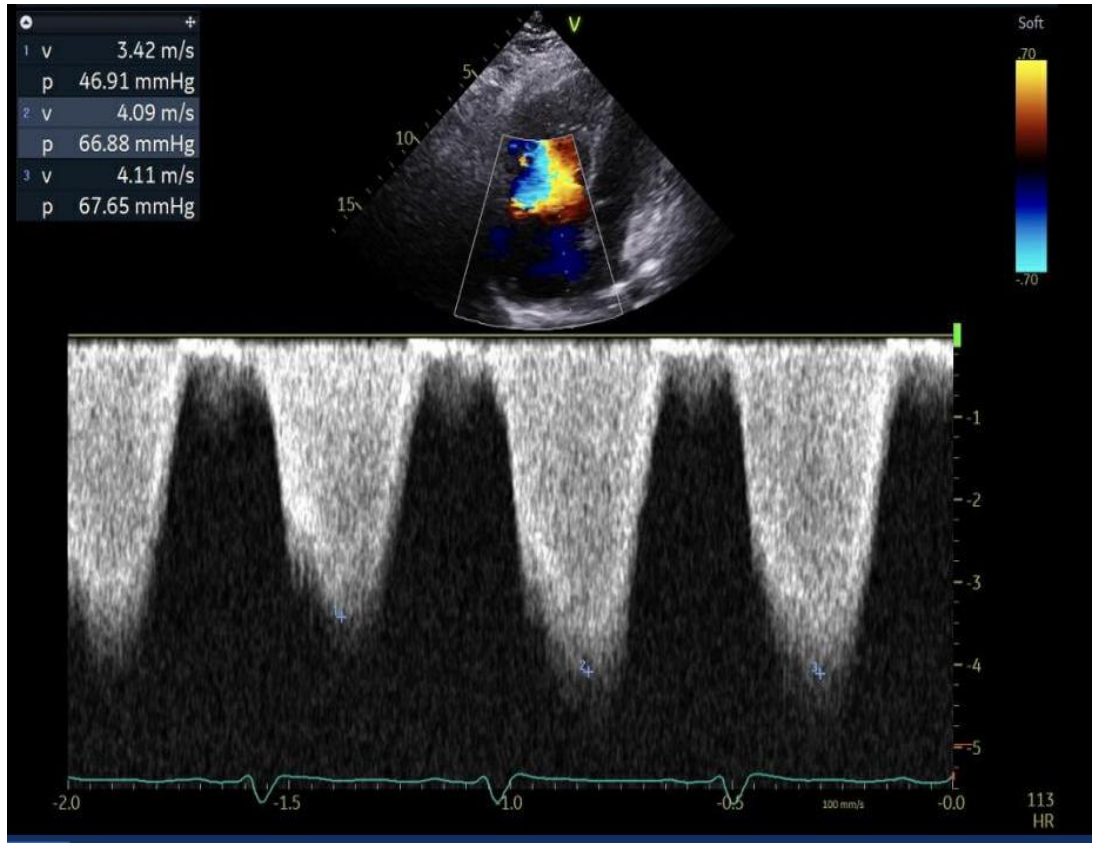
25/F IPAH





25/F IPAH





Severe functional TR with severe resting PH and RV dysfunction

Baseline and f/u after triple combination (SQ treprostinil, macitentan, sildenafil)

	2023.12	2024.09
WHO function class	**IV**	**III**
BNP (pg/ml)	**329**	**61**
6MWT (m)	**190**	**482**
mRAP (mmHg)	**25**	**13**
Mean PAP (mmHg)	**61**	**57**
ScvO2 (%)	52	65
CO (L/m)	**2.5**	**4.7**
PVR (WU)	**18.4**	**9.4**

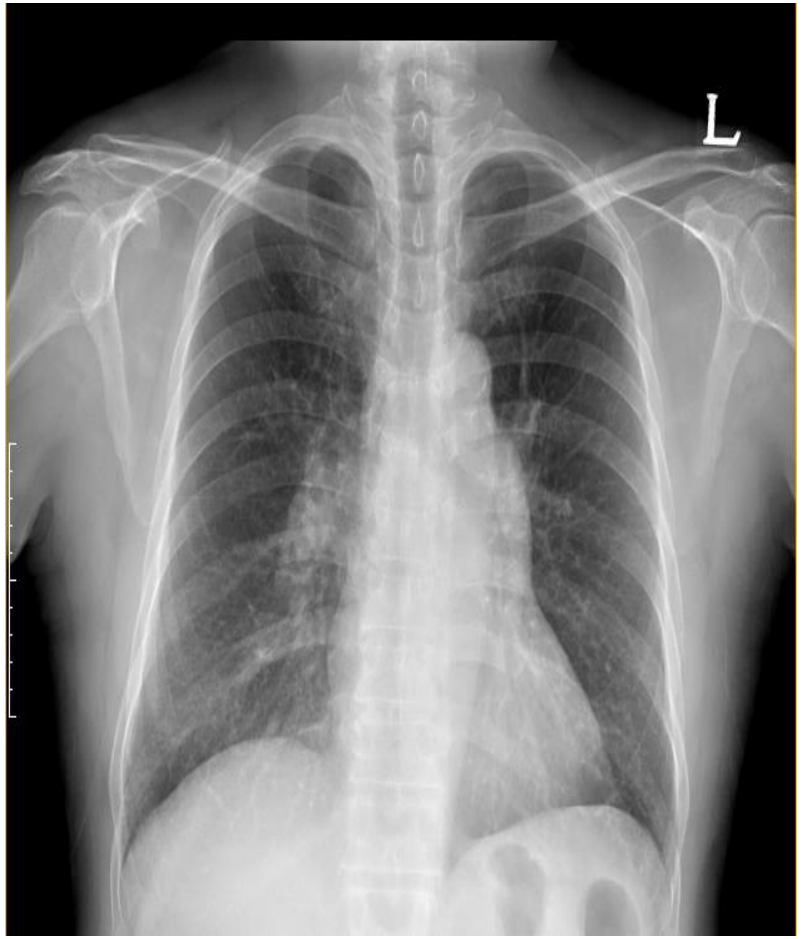
66 male patient

C. C : Dyspnea (Fc III)

P. M. Hx. :

Myocardial infarction (2001)

45 pack-years ex-smoker



ABGA

pO_2 52 mmHg **pCO_2 34 mmHg**

pH 7.39 **HCO 21 mmEq**

66 male patient

SPIROMETRY

		(GK0015)		(GK0022) Bronchodilator		
	Pred.	Meas.	%Pred.	Meas.	%Pred.	%Chg.
FVC	4.03	3.36	83	3.27	81	-3
FEV1	2.89	2.16	75	2.20	76	2
FEV_1 /FVC	73	64	88	68		
$FEF_{25\_75\%}$	2.50	0.82	33	1.06	42	29
PEF	6.89	7.75	112	8.35	121	8

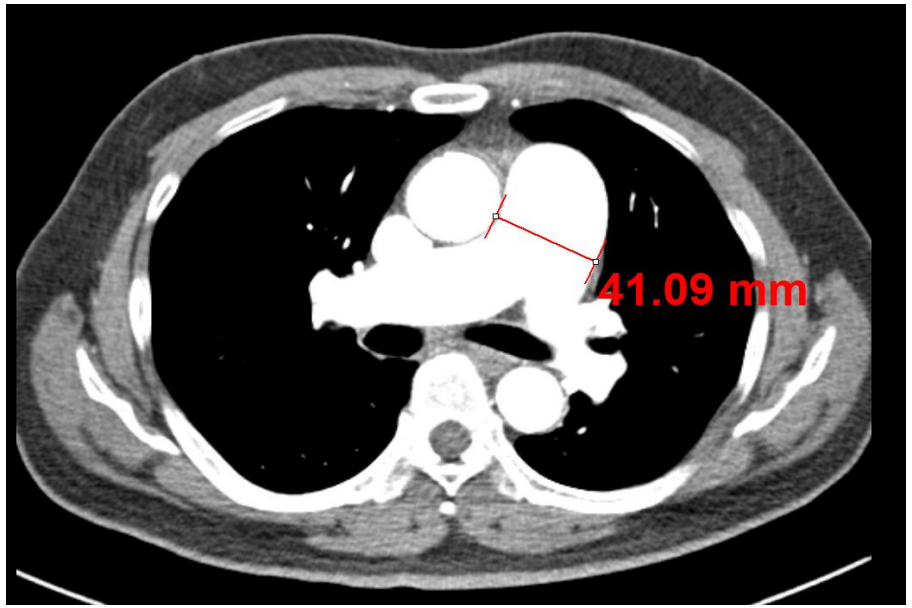
DIFFUSING CAPACITY

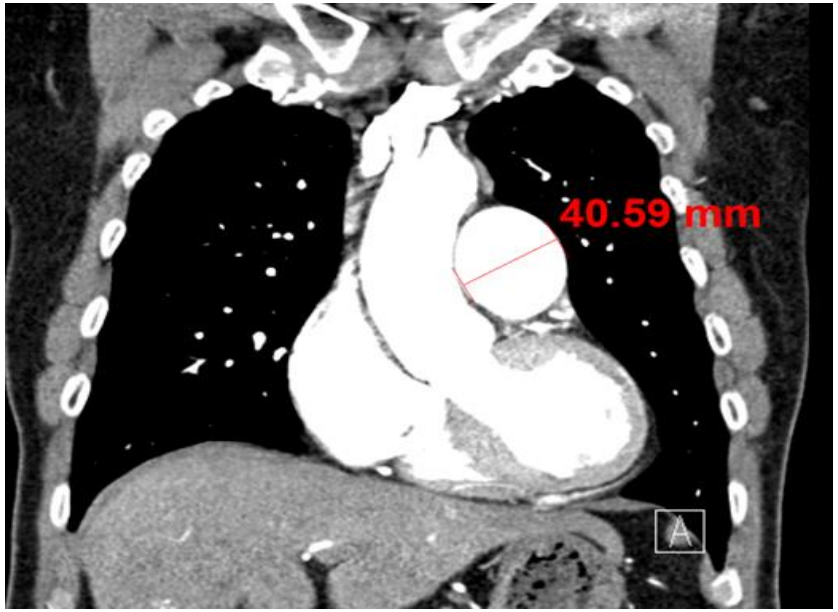
	Pred.	Meas.	%Pred.
DL	19.2	6.8	35
VA(TLC)		3.80	
DL/VA	3.87	1.79	46
Corrected	19.2	6.3	33
Hb	Date 2013.06.24 17.7 g/dl		

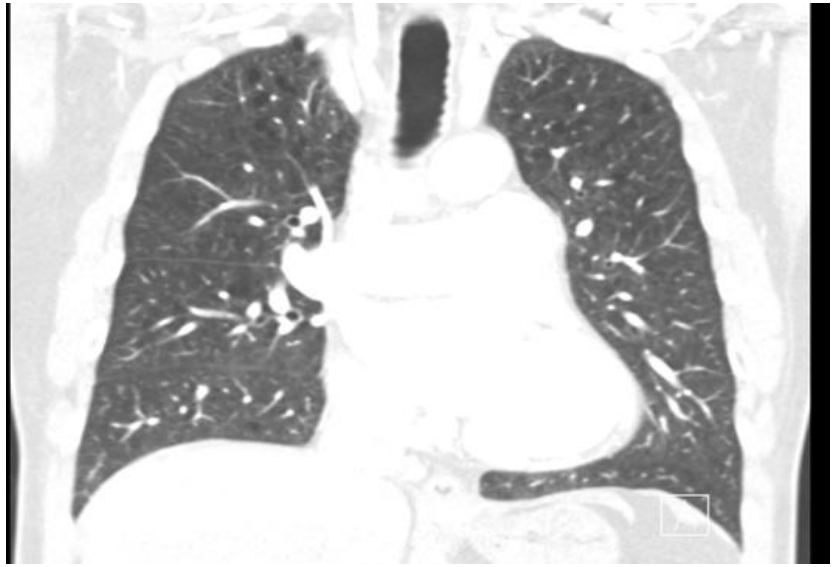
6 minute walk test

TEST(I) O2 LPM		
Pulse	SaO_2	DISTANCE
63	93	
65	93	
70	90	
90	88	
103	85	
107	83	
110	83	
112	82	
115	82	
115	82	
116	81	
117	82	
117	82	368

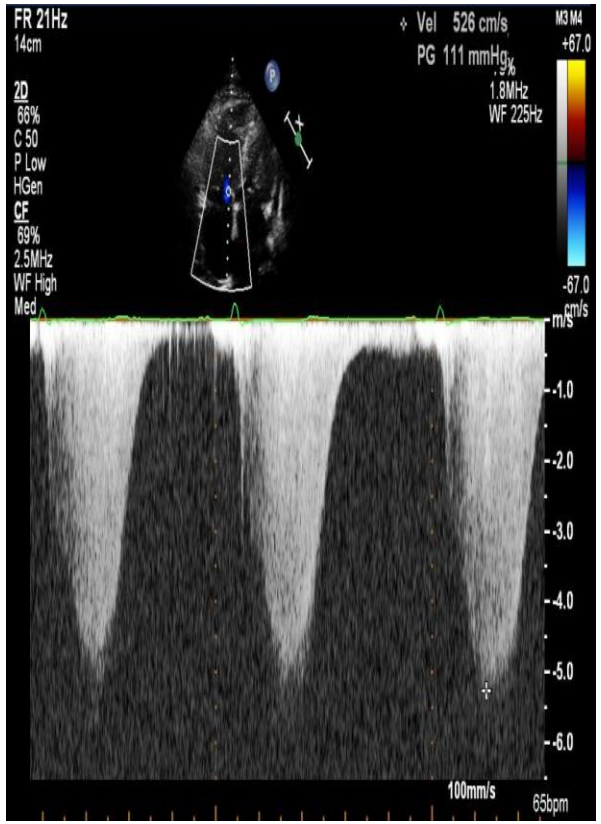
66 male patient







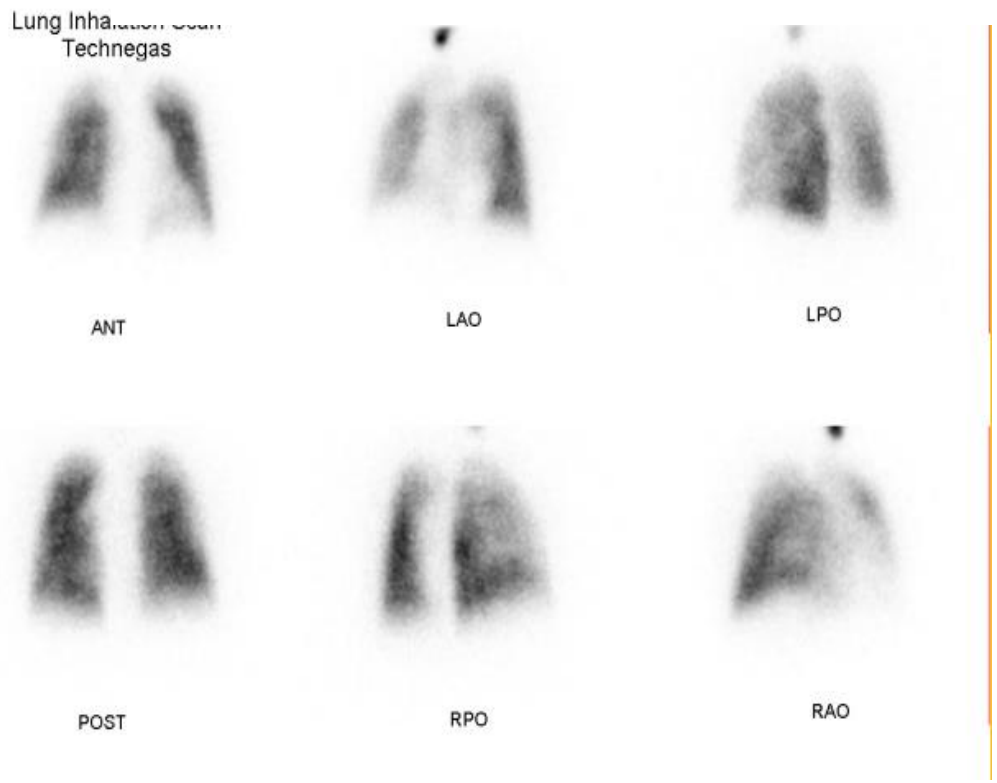
66 male patient

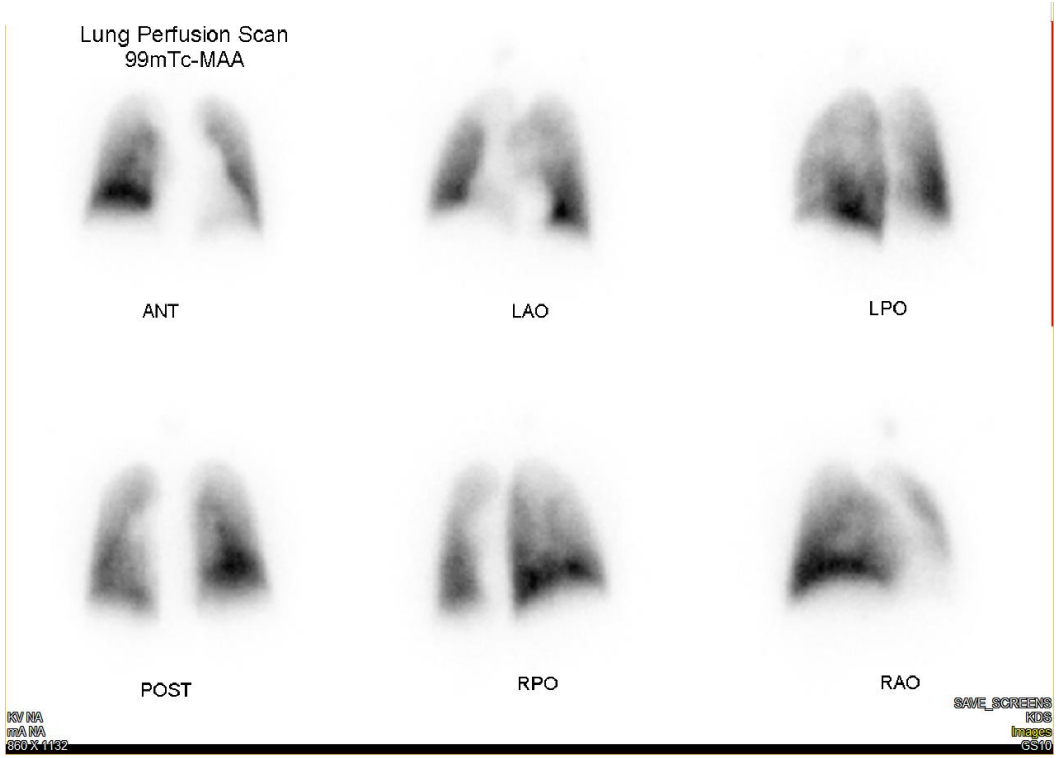


Newly developed severe resting pulmonary hypertension with RV dysfunction

TR Vmax 5.3 m/s PG(RV-RA) 111 mmHg

66 male patient





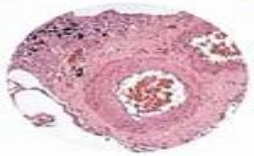
No significant V/Q mismatching lesion

Right heart catheterization

Parameter	
Mean RAP, mmHg	4
Mean PAP, mmHg	49
PAWP (mmHg)	11
CO (L/m)	4.0
PVR (WUs)	9.5
ScVO2	63%

Therapeutic approach of PH

Clinical Classification

Pulmonary arterial hypertension (PAH)	PH associated with left heart disease	PH associated with lung disease	PH associated with pulmonary artery obstructions	PH with unclear and/or multifactorial mechanisms
				
• Idiopathic/heritable • Associated conditions	• IpcPH • CpcPH	• Non-severe PH • Severe PH	• CTEPH • Other pulmonary obstructions	• Haematologic disorders • Systemic disorders

Prevalence

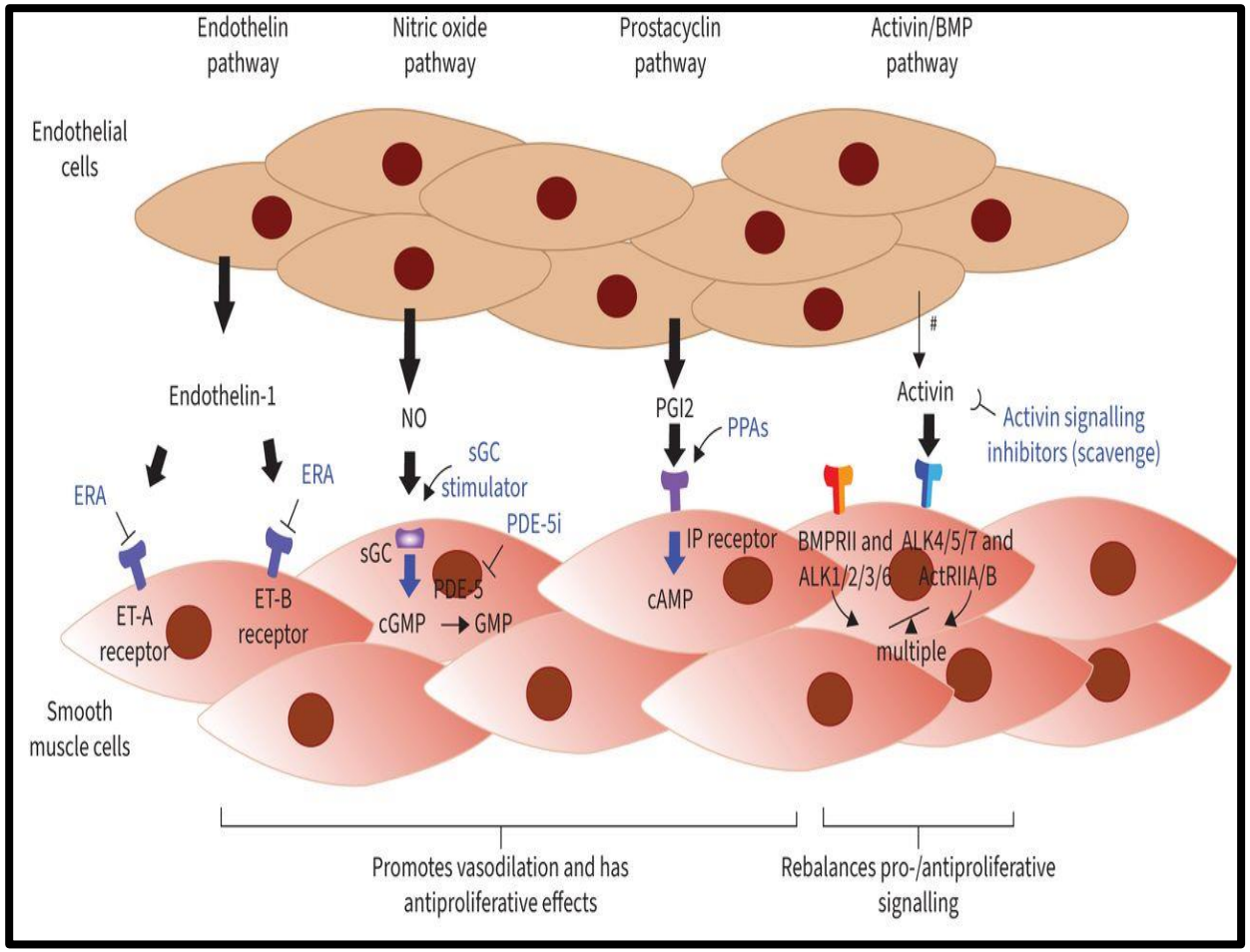
Rare	Very common	Common	Rare	Rare
				

Therapeutic Strategies

Medical therapy • PAH drugs • CCB in responders **Lung transplantation**	**IpcPH:** • Treatment of LHD[a] **CpcPH:** • Treatment of LHD[a] • Potentially: PAH drugs (trials)	**PH-lung disease:** • Optimized care of underlying lung disease **Severe PH:** • Potentially: PAH drugs (trials)	**Surgical therapy:** • PEA **Interventional:** • BPA **Medical therapy:** • PH drugs	**Optimized treatment of underlying disease** • Potentially: PAH drugs (trials)

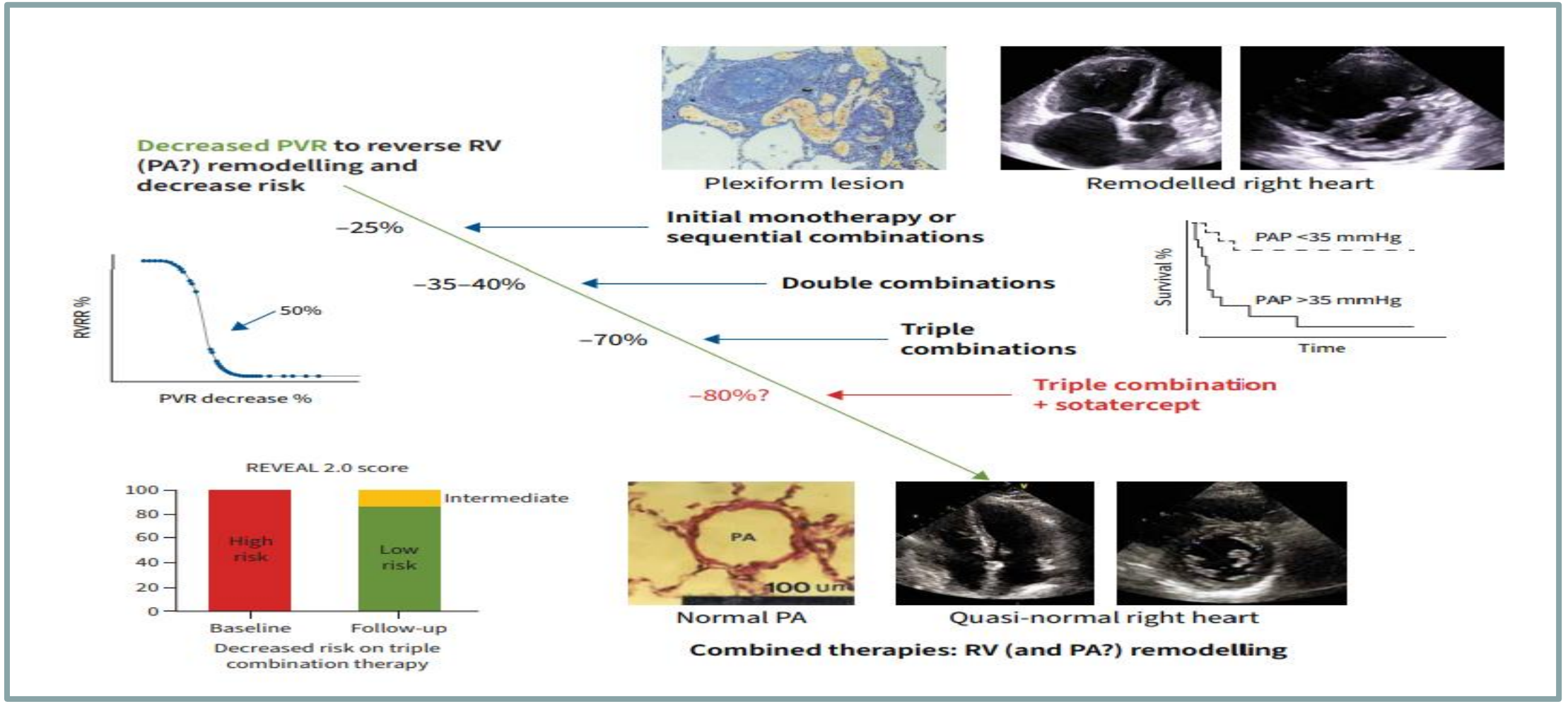
ESC ERS

PAH target drugs



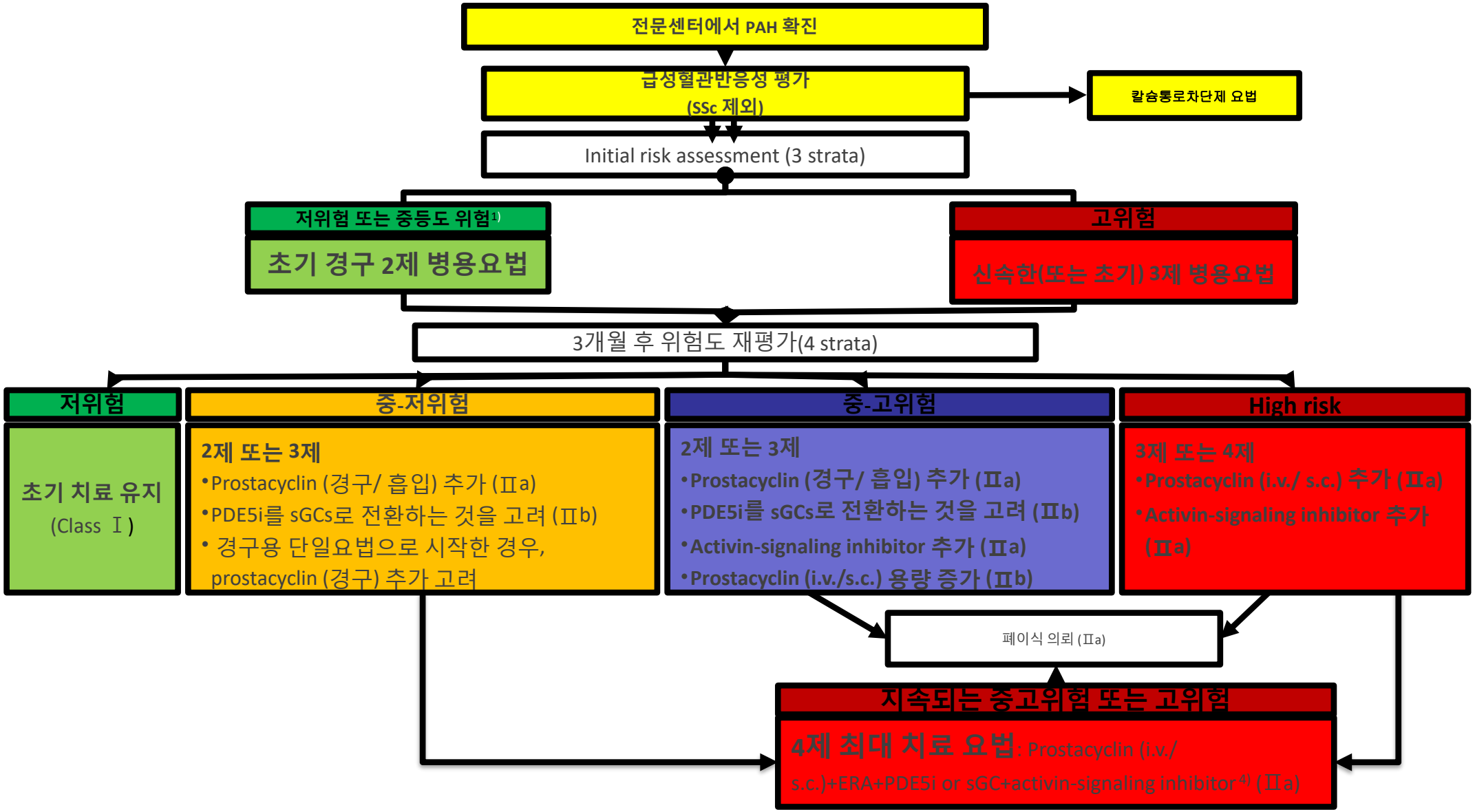
Endothelin receptor antagonist	Bosentan
	Ambrisentan
	Macitentan
PDE-5 inhibitor	Sildenafil
	Tadalafil
sGC stimulator	Riociguat
Prostacyclin analogues	Epoprostenol
	Iloprost
	Treprostinil
Selective IP receptor agonist	Selexipag
Activin signaling inhibitor	Sotatercept

Combination treatment in PAH



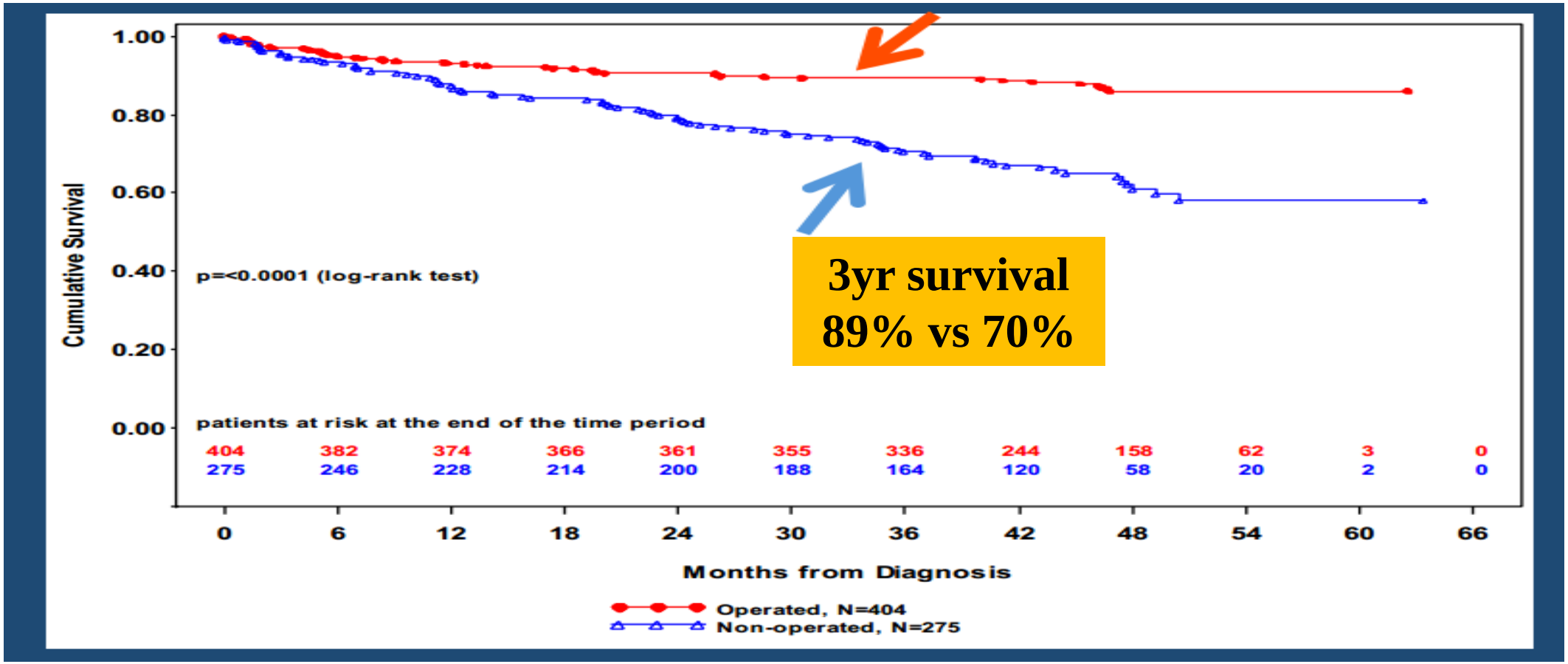
Rubin LJ, Naeije R.Eur Respir J 2023; 61: 2201972.

Treatment algorithm for PAH



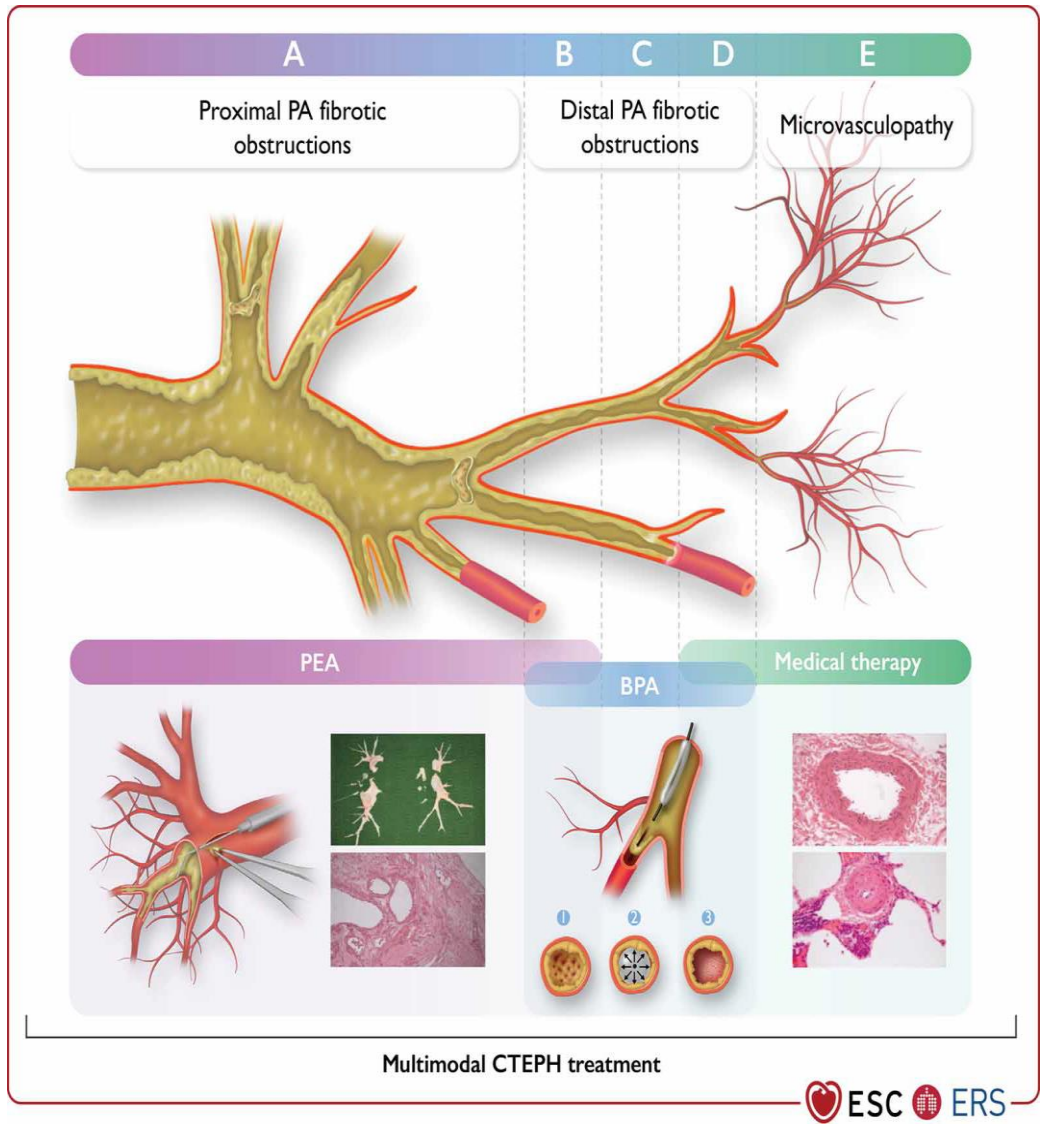
CTEPH: Surgical PH

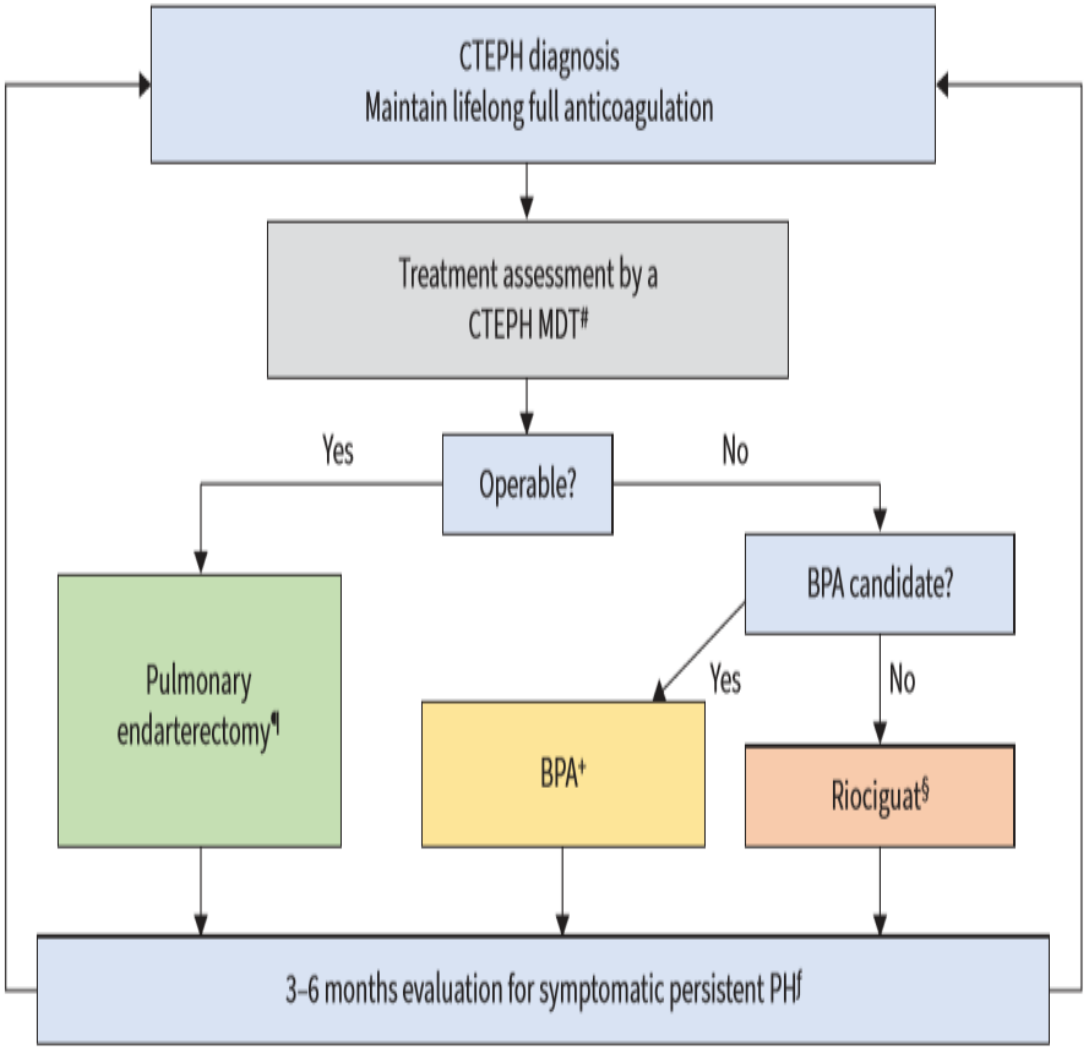
Results From an International Prospective Registry



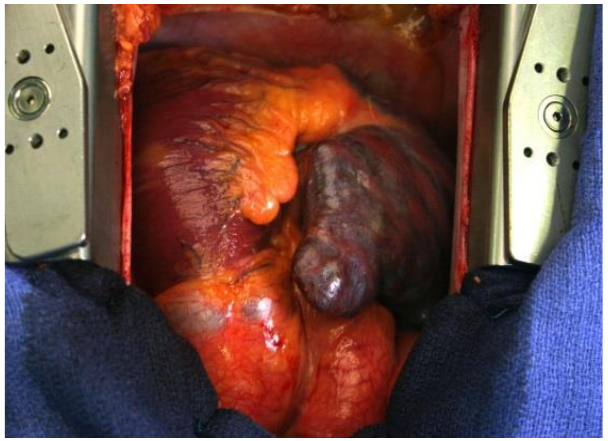
Delcroix M, et al. Circulation. 2016;133:859–871

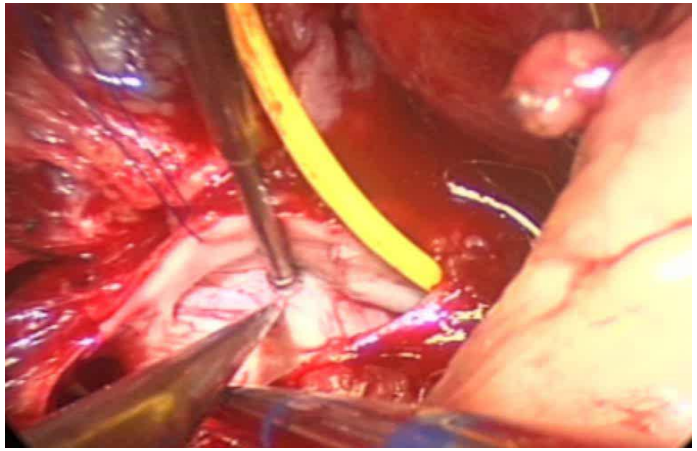
The management options for CTEPH

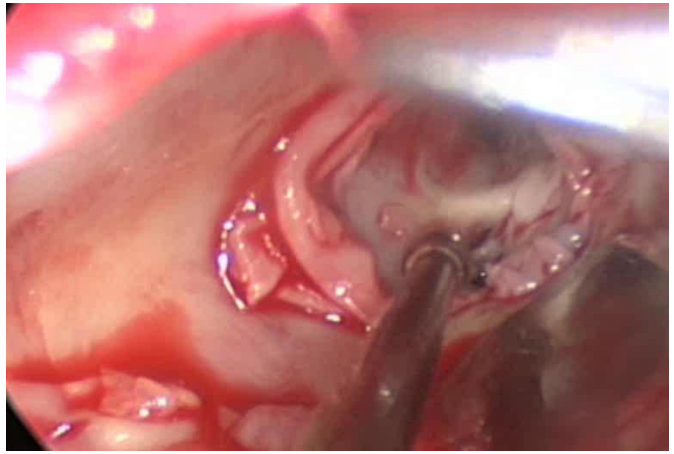




Pulmonary endarterectomy (PEA)







- 정중 흉골절개술
- 인공심폐기 통한 체외순환
- 저체온 (20° C)
- 삼장 정지 (20 분)
- 폐동맥의 혈전 제거 및 내막 박리

Case. (64/M) CTEPH

Chief complaint : dyspnea (WHO II)

Present illness

2005 syncope -> acute PE with DVT

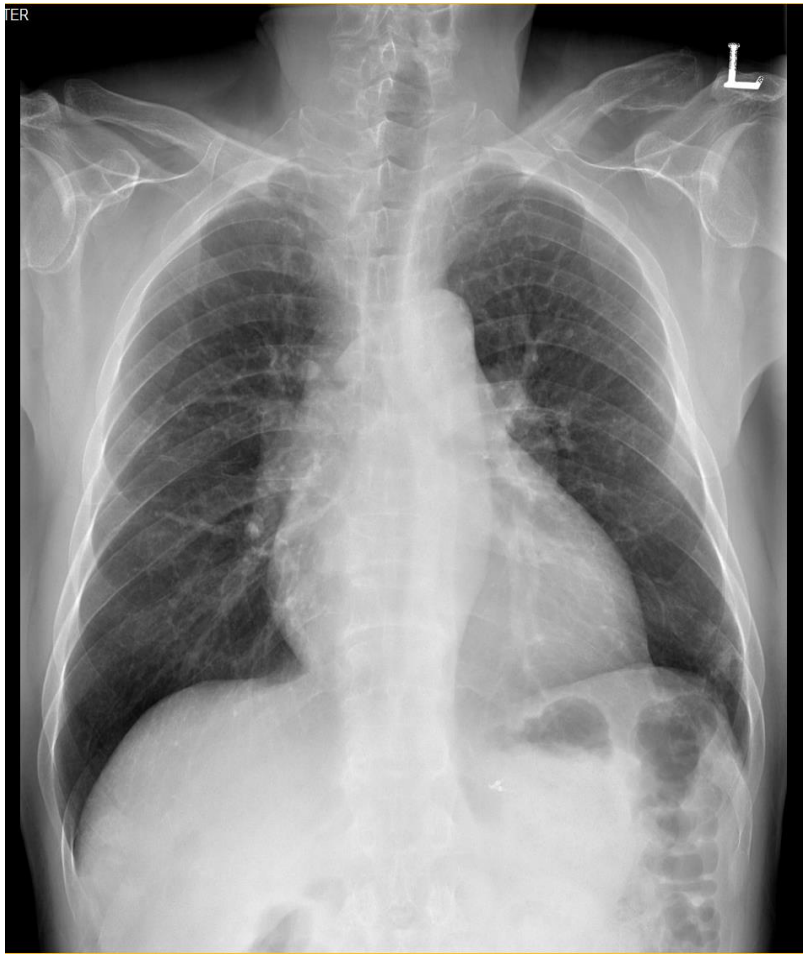
-> thrombolysis and warfarin.

2012CTEPH-> sildenafil

2014 Stomach cancer -> referred to AMC

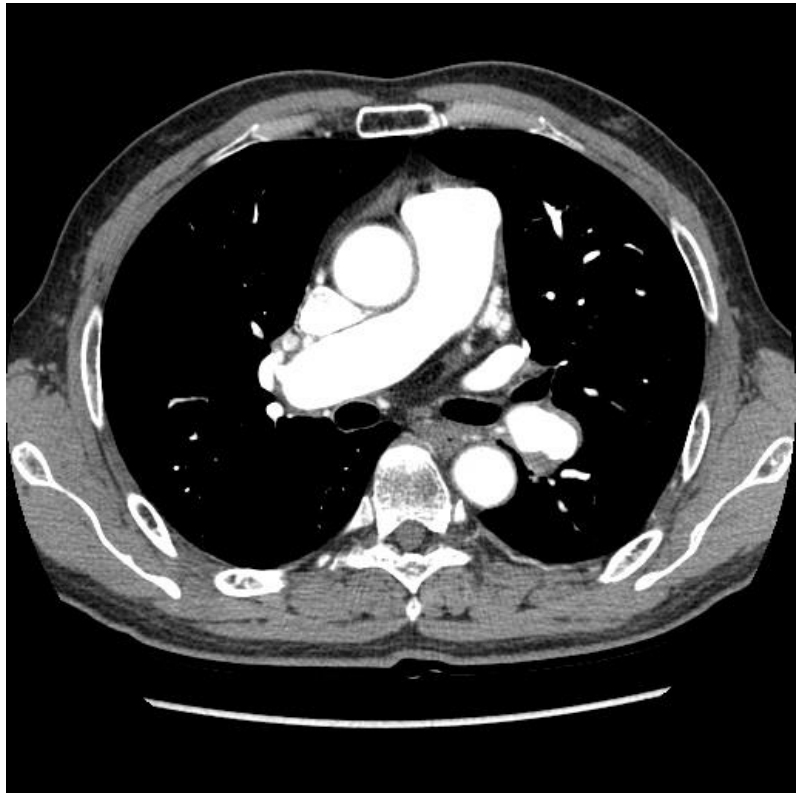
BNP 192 pg/ML

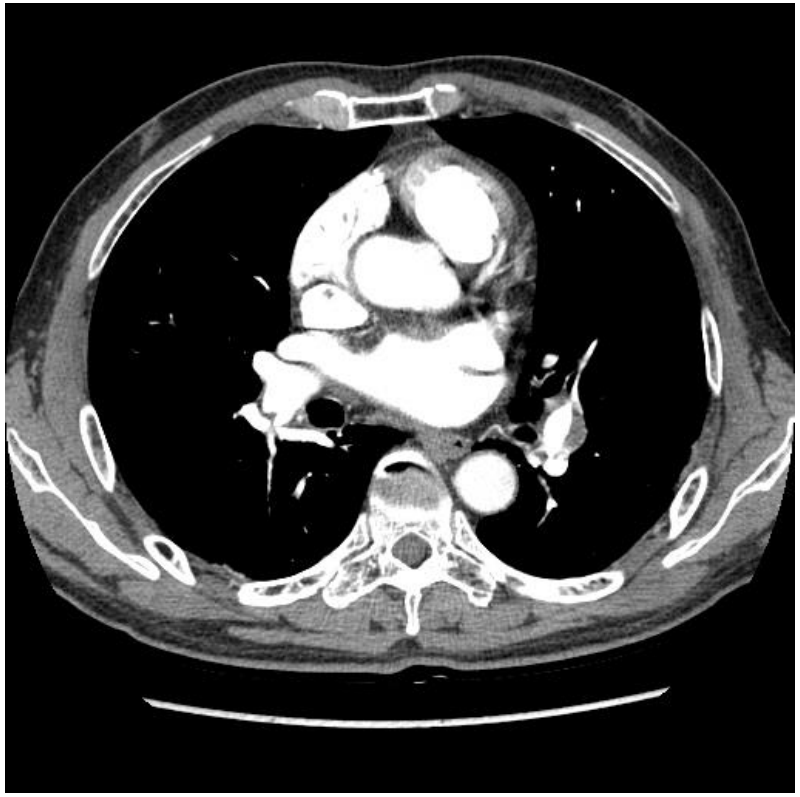
6MWT 438m (SaO_2 97 -> 92%)



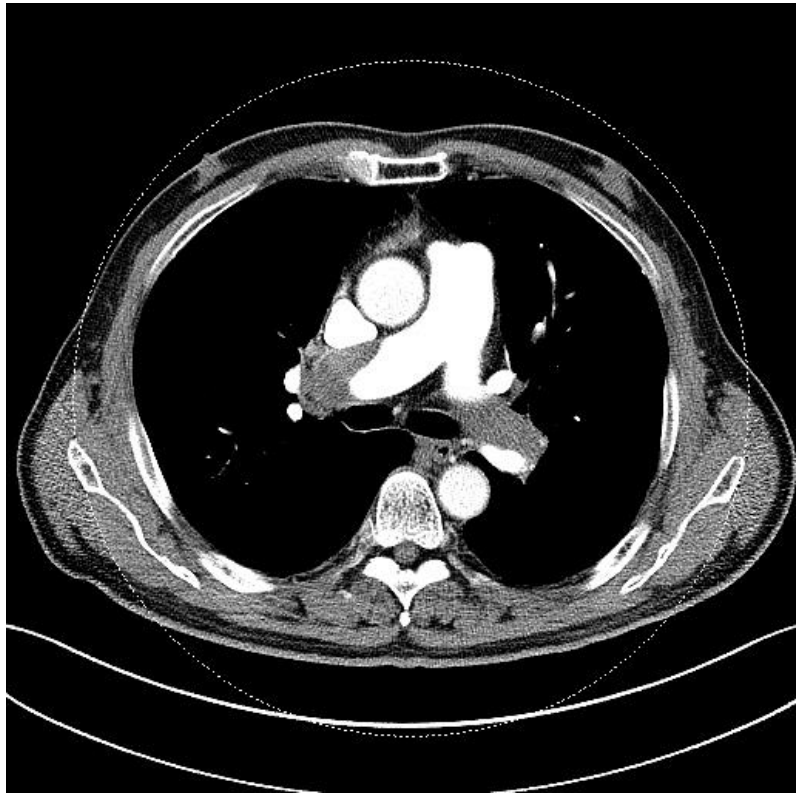
Case; Chest CT (9 years ago)

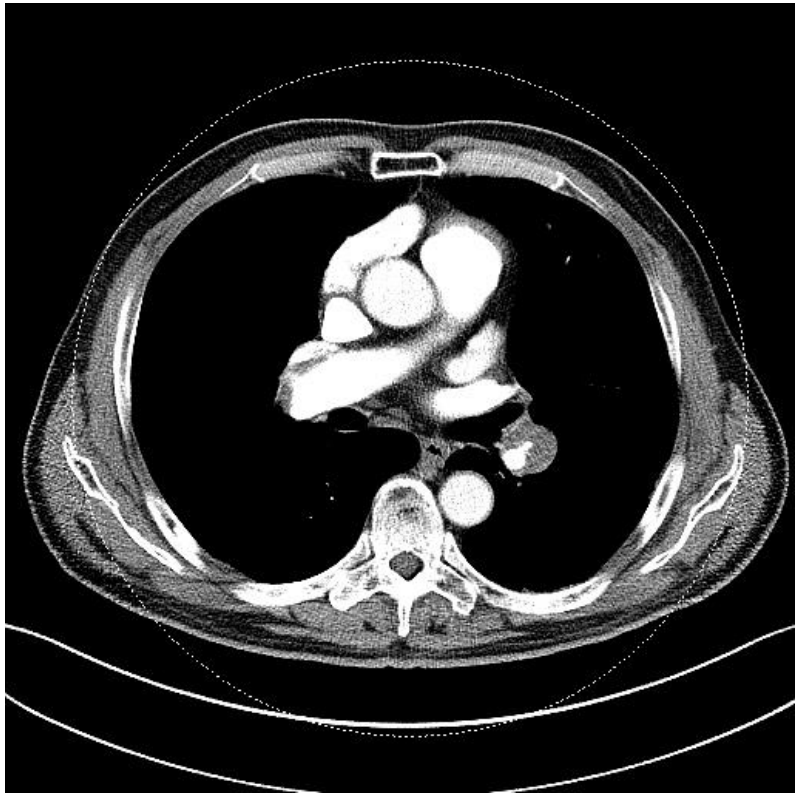
Case; Chest CT (after thrombolysis)



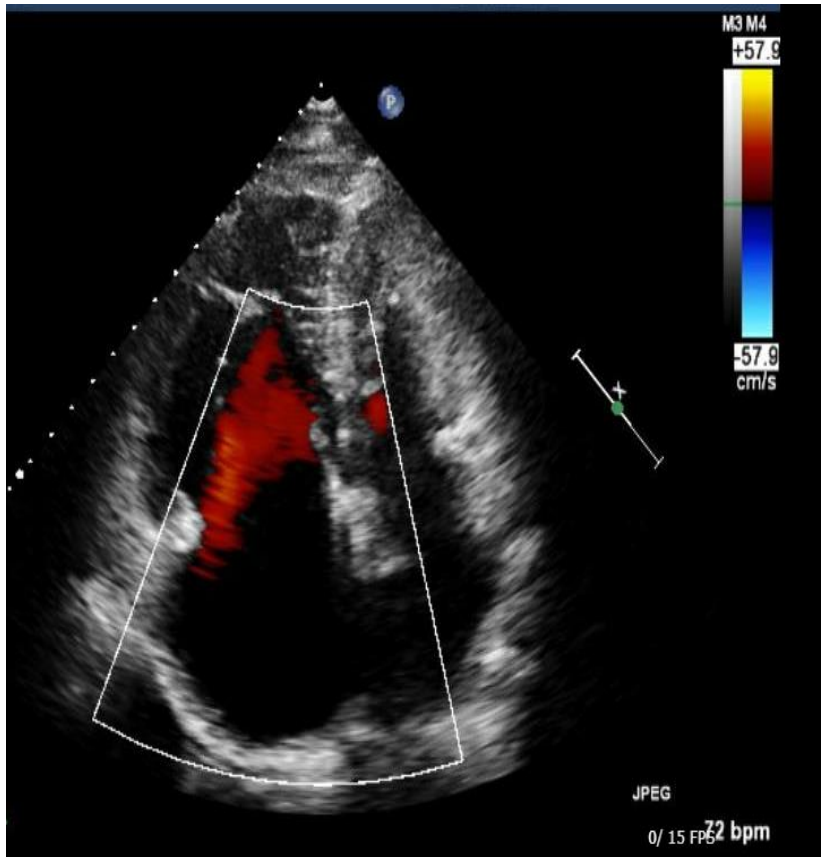


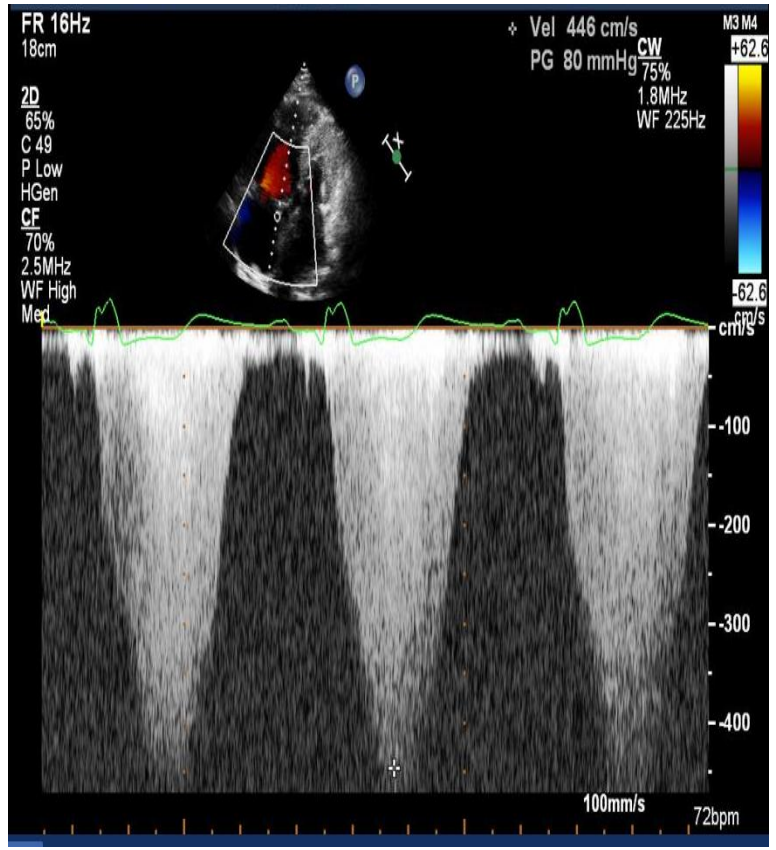
Case; Chest CT(1 week ago)





Severe PH and RV dysfunction

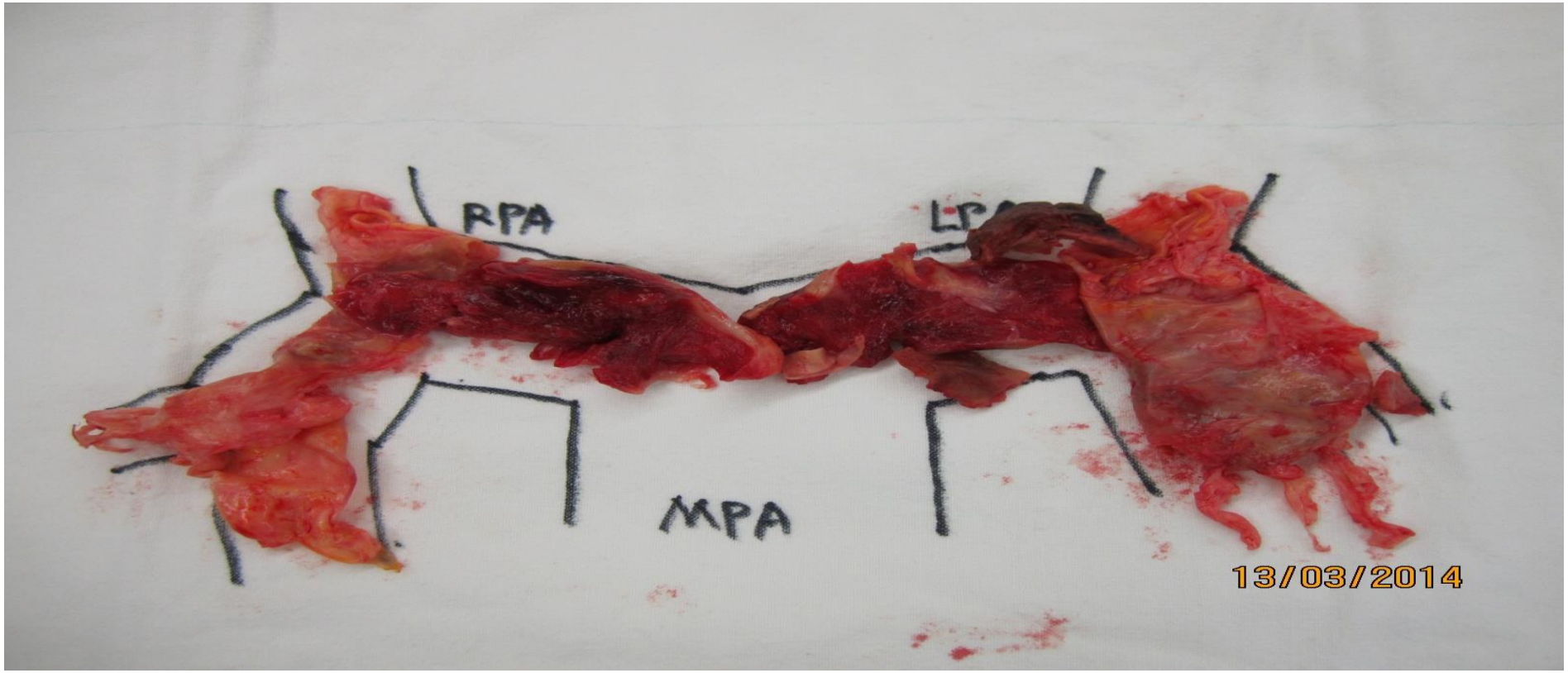




Right heart catheterization

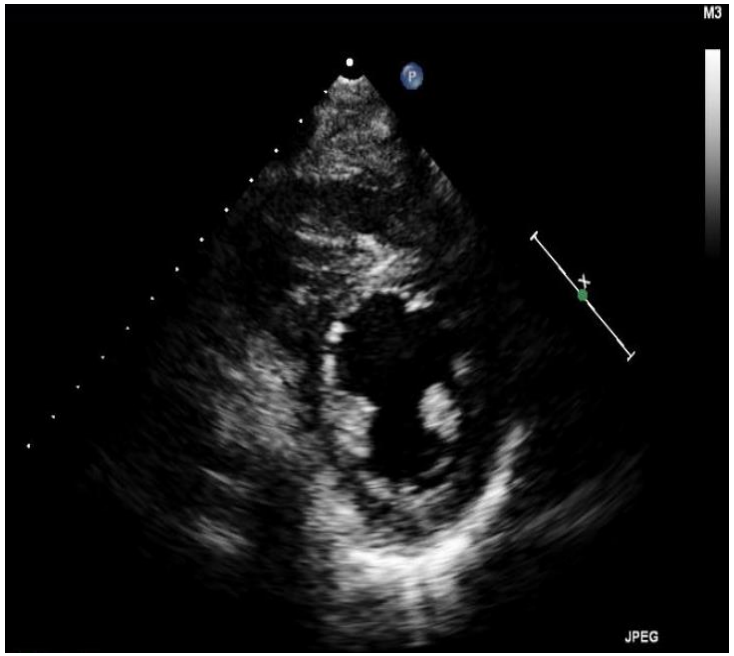
Parameter	
Mean RAP, mmHg	9
Mean PAP, mmHg	42
PAWP (mmHg)	10
CO (L/m)	2.79
PVR (WUs)	11.4
ScVO2	62%

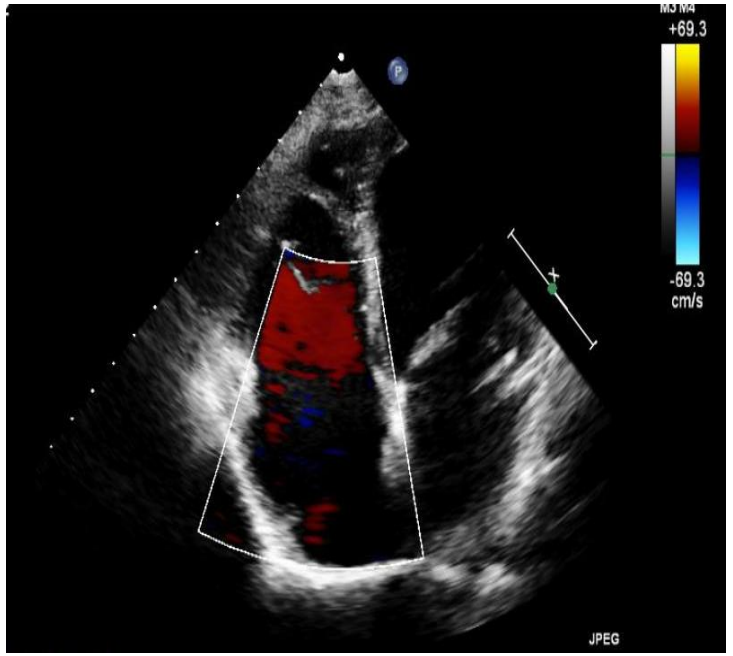
Pulmonary endarterectomy (PEA)

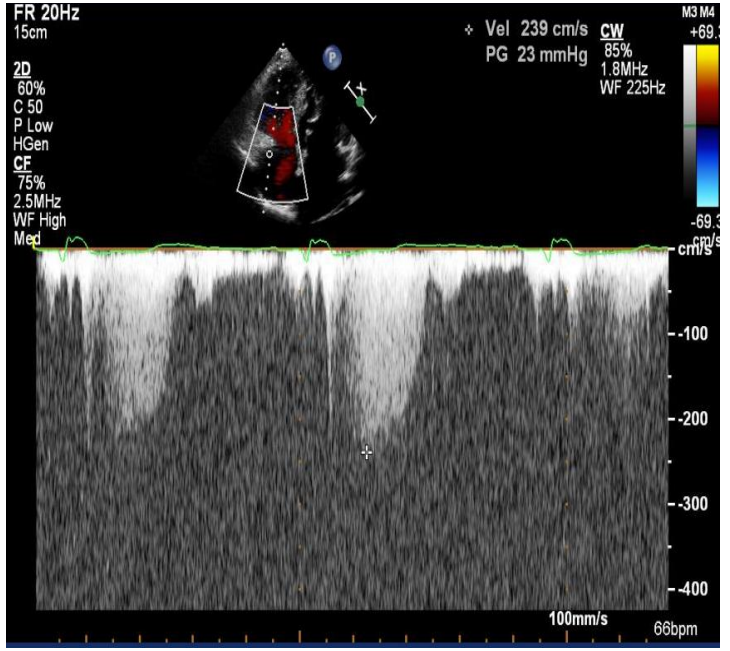


1-year after PEA

WHO Fc I
BNP 90 pg/ML
6MWT 553 m (SaO_2 100 -> 97%)







No PH and normal RV function

Take home messages

- **Acute PE**

  → Strongly suspect when symptoms are unexplained by routine imaging or ECG, especially in the presence of relevant risk factors.

- **PH**

  → Consider in patients with exertional dyspnea or hypoxemia that is disproportionate to findings on chest imaging and PFTs.