



Cardiac Lecture #7: Pulmonary Hypertension in Pregnancy

May 2, 2023



Updates



- Next Maternal Webinar June 6th
Topic: OB Anesthesia and L&D Considerations | Erica Johnson, MD
- Q1 2022 HTN Data Submission – DUE NOW
- Q1 2023 Cardiac Data Submission – May 15th
- Sustainability Survey's

SIMULATION AND DRILLS FOR PATIENT SAFETY

OBSTETRIC IN-SITU DRILL PROGRAM MANUAL ▶



PRACTICING FOR PATIENTS SIMULATIONS PREPARATION CHECKLIST ▶



SAMPLE CASE SCENARIOS

- ☰ HYPERTENSION CASE SCENARIO 1
- ☰ HEMMORHAGE CASE SCENARIO 1
- ☰ HYPERTENSION CASE SCENARIO 2
- ☰ HEMMORHAGE CASE SCENARIO 2
- ☰ HYPERTENSION CASE SCENARIO 3
- ☰ HEMMORHAGE CASE SCENARIO 3
- ☰ HYPERTENSION SCENARIO TRAINING AIDS
- ☰ FETAL HEART RATE TONES TRAINING AIDS
- ☰ HEMORRHAGE SCENARIOS VISUAL AIDS
- ☰ ADDITIONAL HEMORRHAGE CASE SCENARIOS



SAMPLE CASE VIDEOS

- ▶ SEVERE HYPERTENSION CASE 1
- ▶ SEVERE HYPERTENSION CASE 2
- ▶ SEVERE HYPERTENSION CASE 3
- ▶ OBSTETRIC HEMORRHAGE - REQUIRING UTERINE TAMPONADE
- ▶ OBSTETRIC HEMORRHAGE - REQUIRING UTEROTONICS
- ▶ OBSTETRIC HEMORRHAGE WITH RETAINED PRODUCTS



TEAM REVIEW AND DEBRIEFING

- ☑ SEVERE HYPERTENSION FORM
- ☑ OBSTETRIC HEMORRHAGE FORM

TEAM BASED COMMUNICATION TRAINING

- ☑ TEAM STEPPS



PROTOCOL CHANGE FORM AND IMPLEMENTATION ACTION PLAN

- ☑ IN-SITU DRILLS FACILITY PROTOCOL CHANGE FORM
- ☑ PRACTICING FOR PATIENTS IMPLEMENTATION ACTION PLAN

PRACTICING FOR PATIENTS PRESENTATIONS

- ▶ PRACTICING FOR PATIENTS PRESENTATION FOR STAFF (PPT)
- ▶ PRACTICING FOR PATIENTS PRESENTATION FOR LEADERSHIP (PPT)



Key Driver Diagram: Maternal Cardiac Conditions

GOAL:

To reduce severe morbidity & mortality related to maternal cardiac conditions in Georgia.

SMART AIM:

By 02/6/2026, **National Wear Red Day**, to reduce harm related to existing and pregnancy related cardiac conditions through the 4th trimester by **20%**.

Key Drivers

Readiness: EVERY UNIT - Implementation of standard processes for optimal care of cardiac conditions in pregnancy and post-partum.

Recognition & Prevention: EVERY PATIENT - Screening and early diagnosis of cardiac conditions in pregnancy and post-partum.

Response: EVERY UNIT - Care management for every pregnant or postpartum woman with cardiac conditions in pregnancy and post-partum.

Reporting/System Learning: EVERY UNIT - Foster a culture of safety and improvement for care of women with cardiac conditions in pregnancy and post-partum.

Respectful, Equitable, and Supportive Care — EVERY UNIT/PROVIDER/TEAM MEMBER - Inclusion of the patient as part of the multidisciplinary care team.

INTERVENTIONS

- Train all obstetric care providers to perform a basic Cardiac Conditions Screen.
- Establish a protocol for rapid identification of potential pregnancy-related cardiac conditions in all practice settings to which pregnant and postpartum people may present.
- Develop a patient education plan based on the pregnant and postpartum person's risk of cardiac conditions.
- Establish a multidisciplinary "Pregnancy Heart Team" or consultants appropriate to their facility's designated Maternal Level of Care to design coordinated clinical pathways for people experiencing cardiac conditions in pregnancy and the postpartum period. S1**
- Establish coordination of appropriate consultation, co-management and/or transfer to appropriate level of maternal or newborn care.
- Develop trauma-informed protocols and training to address health care team member biases to enhance quality of care
- Develop and maintain a set of referral resources and communication pathways between obstetric providers, community-based organizations, and state and public health agencies to enhance quality of care. *

- Obtain a focused pregnancy and cardiac history in all care settings, including emergency department, urgent care, and primary care.
- In all care environments assess and document if a patient presenting is pregnant or has been pregnant within the past year. S2**
- Assess if escalating warning signs for an imminent cardiac event are present.
- Utilize standardized cardiac risk assessment tools to identify and stratify risk.
- Conduct a risk-appropriate work-up for cardiac conditions to establish diagnosis and implement the initial management plan.

- Facility-wide standard protocols with checklists and escalation policies for management of **cardiac symptoms**.
- Facility-wide standard protocols with checklists and escalation policies for management of people **with known or suspected cardiac conditions**.
- Coordinate transitions of care including the discharge from the birthing facility to home and transition from postpartum care to ongoing primary and specialty care.
- Offer reproductive life planning discussions and resources, including access to a full range of contraceptive options in accordance with safe therapeutic regimens. *
- Provide patient education focused on general life-threatening postpartum complications and early warning signs, including instructions of who to notify if they have concerns, and time and date of a scheduled postpartum visit. S3**

- For pregnant and postpartum people at high risk for a cardiac event, establish a culture of multidisciplinary planning, admission huddles and post-event debriefs.
 - Perform multidisciplinary reviews of serious complications (e.g., ICU admissions for other than observation) to identify systems issues. S4**
 - Monitor outcomes and process data related to cardiac conditions, with disaggregation by race and ethnicity due to known disparities in rates of cardiac conditions experienced by Black and Indigenous pregnant and postpartum people.
- Process Measures – 1-5**

- Screen for structural and social drivers of health that might impact clinical recommendations or treatment plans and provide linkage to resources that align with the pregnant or postpartum person's health literacy, cultural needs, and language proficiency.
- Engage in open, transparent, and empathetic communication with pregnant and postpartum people and their identified support network to understand diagnoses, options, and treatment plans.
- Include each pregnant or postpartum person and their identified support network as respected members of and contributors to the multidisciplinary care team. *S5**



Joel T. Hardin, MD
Medical Director
Emory Adult Congenital Heart Center
Emory University School of Medicine



Georgia Perinatal Quality Collaborative

Pulmonary Hypertension and Pregnancy

Joel T. Hardin MD

Medical Director

Emory Adult Congenital Heart Center

Disclosures

- None

Agenda

- Clinical scenario presentation.
- Review of the 2022 guidelines for classification, evaluation and management of pulmonary hypertension.
- Recommendations pertaining to pregnancy and contraception for women with pulmonary hypertension.
- Clinical scenario questions and answers.

Clinical Scenario:

Congenital Heart Disease (CHD)

- 30-year-old woman with unrepaired large ventricular septal defect. On no medications. No known prior pregnancies.
- Has no symptoms at rest or with ordinary activity but has never been able to run or vigorously exercise without rapidly becoming short of breath. No history of chest pain, arrhythmia, syncope, or hemoptysis.
- BMI 22, BP 110/70 mmHg, SpO₂ 85% in room air, nailbeds are clubbed.
- She reports a positive home pregnancy test, and you subsequently confirm intrauterine pregnancy with estimated gestational age 10 weeks. *She wants your advice...*

2022 ESC/ERS Guidelines for Pulmonary Hypertension: Key Points

- Pulmonary hypertension (PH) is now defined by a mean pulmonary arterial pressure >20 mm Hg at rest.
- The definition of pulmonary arterial hypertension (PAH) also implies a pulmonary vascular resistance (PVR) >2 Wood Units and pulmonary arterial wedge pressure ≤ 15 mm Hg.
- The main diagnostic algorithm for PH has been simplified following a three-step approach, from suspicion by first-line physicians, detection by echocardiography, and confirmation with right heart catheterization in PH centers.

2022 ESC/ERS Guidelines for Pulmonary Hypertension: Key Points

- Women with PH who become pregnant or present during pregnancy with newly diagnosed PAH should be treated, whenever possible, in centers with a multidisciplinary team experienced in managing PH in pregnancy.
- It is recommended to stop endothelin receptor antagonists, riociguat, and selexipag because of potential or unknown teratogenicity. Despite limited evidence, calcium channel blockers, phosphodiesterase 5 (PDE5) inhibitors, and inhaled/IV/subcutaneous prostacyclin analogues are considered safe during pregnancy.

PH Nomenclature and Groups

Group 1

- Idiopathic **PAH**
- Hereditary PAH
- Drug induced PAH
- Connective tissue disease associated PAH (e.g. scleroderma)
- Congenital heart disease associated PAH

Group 2

- Due to **left heart disease**
- Risk factors for this type of PH include coronary artery disease, hypertension, diabetes, high cholesterol etc.)

Group 3

- PH due to **lung disease or hypoxia** (low oxygen)
- This can be caused by advanced lung disease including COPD, Interstitial lung diseases (e.g. IPF) or obstructive sleep apnoea

Group 4

- **Chronic thromboembolic pulmonary hypertension** (CTEPH) is the most common cause of Group 4 PH.

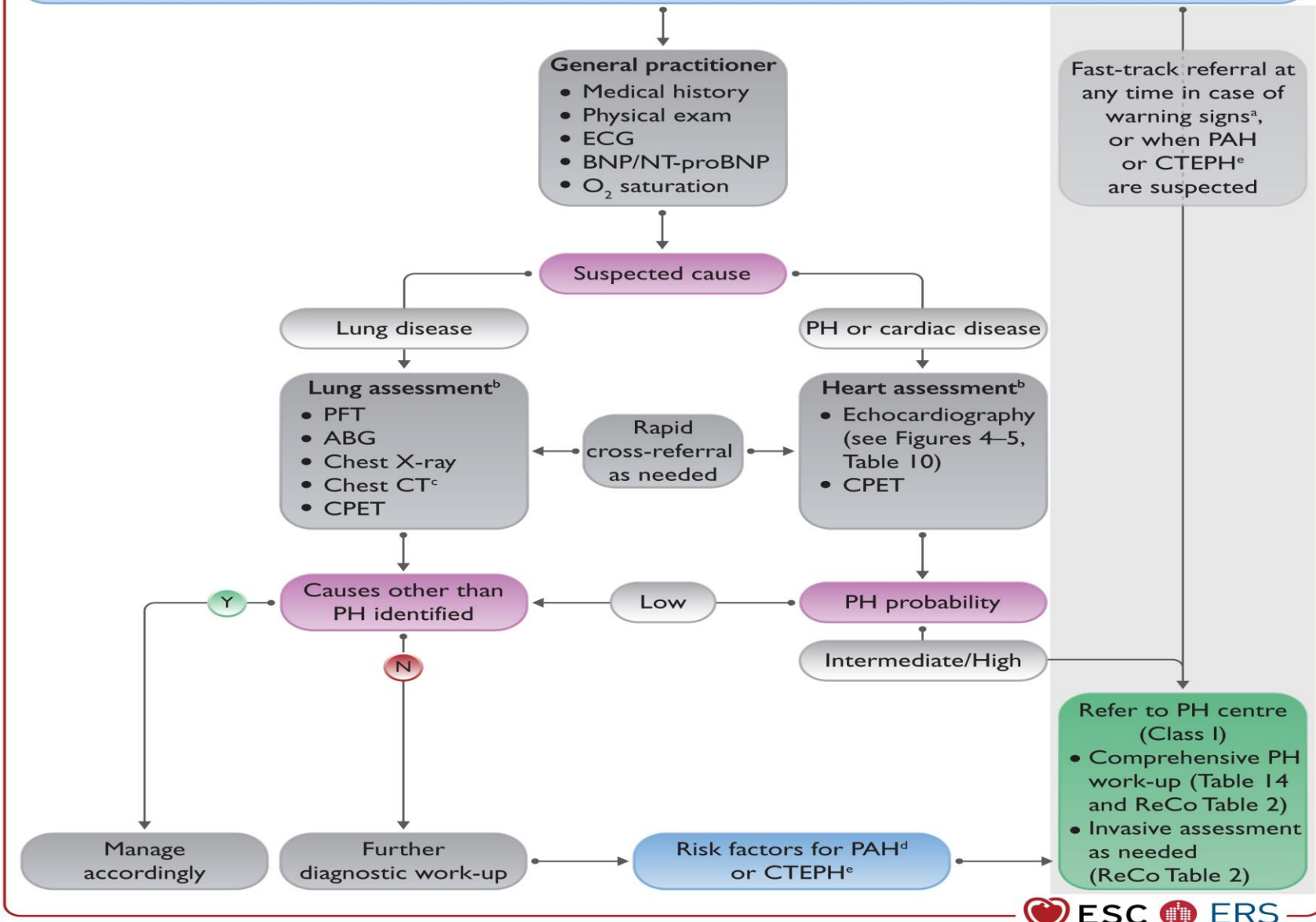
Group 5

- This is a '**miscellaneous**' group that includes causes of PH that are unclear or have multiple mechanisms such as sarcoidosis.

PH Epidemiology and Risk Factors

- All age groups are affected.
- PH prevalence of ~1% of the global population.
- Due to the presence of cardiac and pulmonary causes of PH, prevalence is higher in individuals aged >65 years.
- Left heart disease is the leading cause of PH.
- Lung disease, especially chronic obstructive pulmonary disease (COPD), is the second most common cause.

Diagnostic algorithm of patients with unexplained exertional dyspnoea and/or suspected 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)

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

Late

Table 15 World Health Organization classification of functional status of patients with pulmonary hypertension

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

PH, pulmonary hypertension; WHO-FC, World Health Organization functional class.

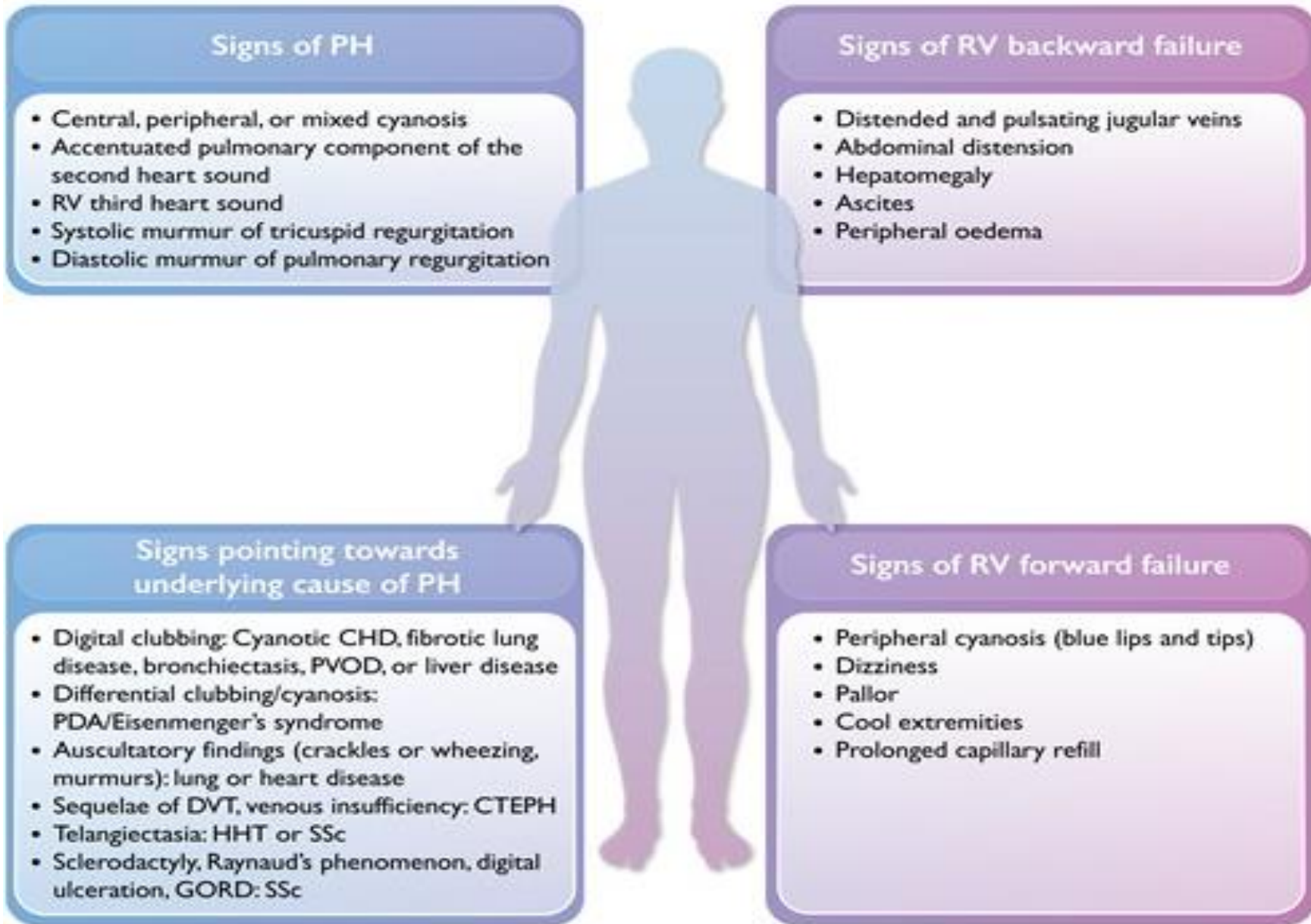


Table 8 Electrocardiogram abnormalities in patients with pulmonary hypertension

Typical ECG abnormalities in PH⁶⁶

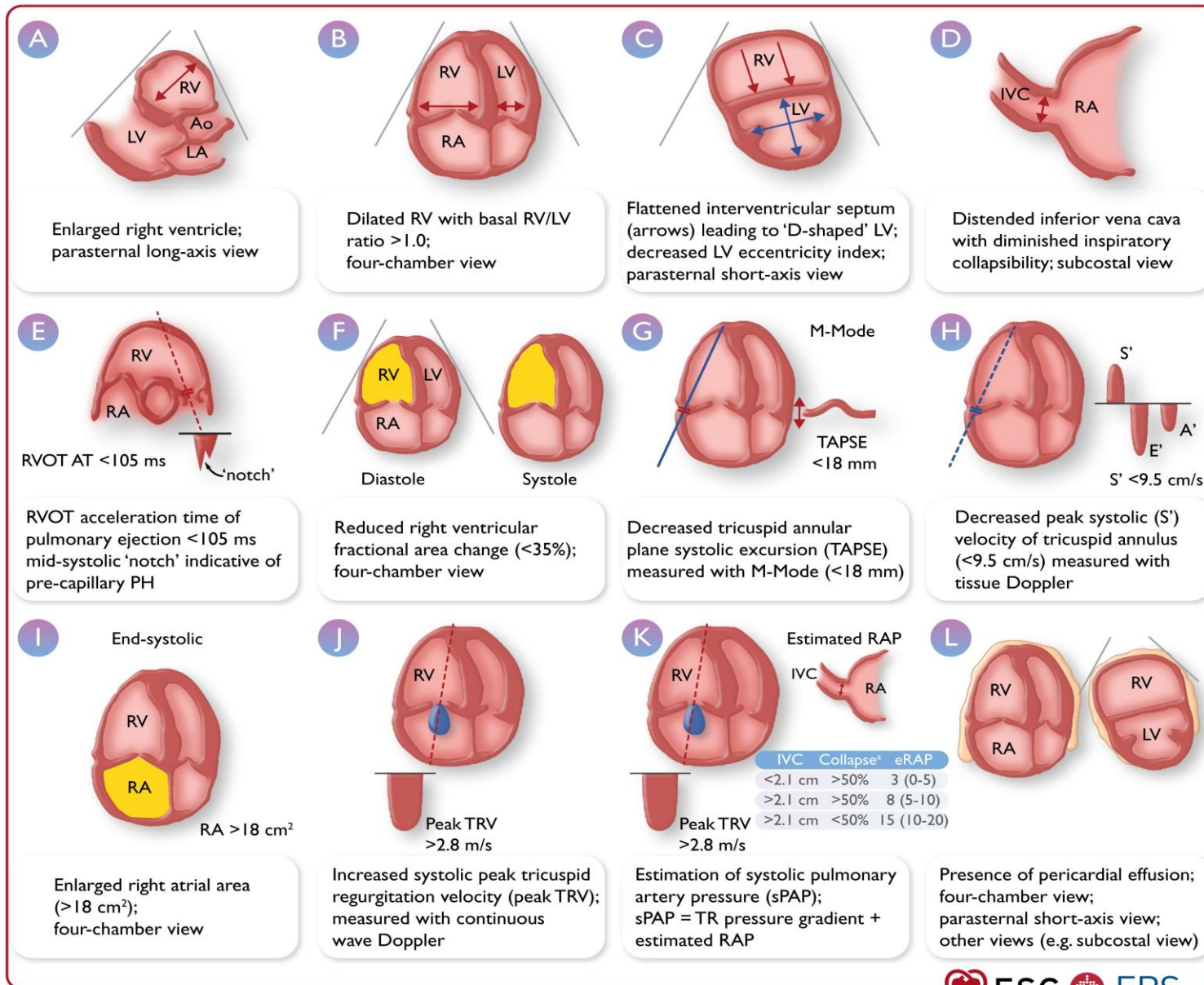
- P pulmonale (P >0.25 mV in lead II)
- Right or sagittal axis deviation (QRS axis >90° or indeterminable)
- RV hypertrophy (R/S >1, with R >0.5 mV in V1; R in V1 + S in lead V5 > 1 mV)
- Right bundle branch block—complete or incomplete (qR or rSR patterns in V1)
- RV strain pattern^a (ST depression/T-wave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

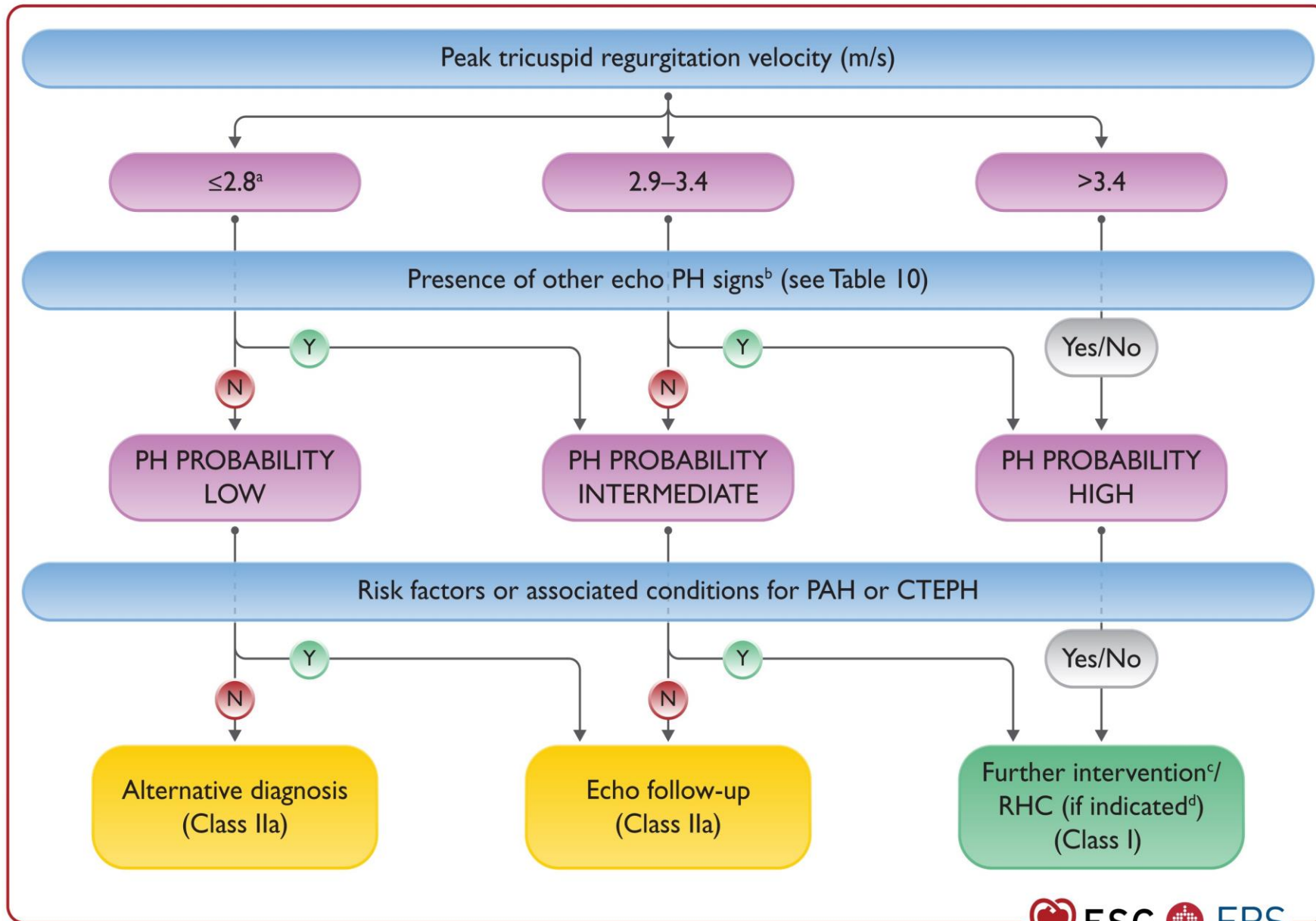
ECG, electrocardiogram; PH, pulmonary hypertension; QTc, corrected QT interval; RV, right ventricular.

Table 9 Radiographic signs of pulmonary hypertension and concomitant abnormalities

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)

COPD, chronic obstructive pulmonary disease; PA, pulmonary artery; PH, pulmonary hypertension.





PH Diagnostics: Additional Tests

- Blood tests: CBC, electrolytes, creatinine & calculated GFR, uric acid, ALT, AST, alk phos, GGT, bilirubin, serum iron, transferrin, ferritin, BNP or NT-proBNP, TSH.
- Serologies: Hepatitis viruses and HIV, ANA, anti-centromere antibodies, and anti-Ro.
- Genetic testing for PH associated mutations.
- Cardiopulmonary exercise testing.
- Cardiac catheterization (with vasoreactivity drug testing).
 - 500 mL IV saline over 5-10 min challenge during cath may reveal LV diastolic dysfunction in patients with baseline PAWP ≤ 15 mmHg

Table 18 Variables used to calculate the simplified four-strata risk-assessment tool

Determinants of prognosis	Low risk	Intermediate-low risk	Intermediate-high risk	High risk
Points assigned	1	2	3	4
WHO-FC	I or II ^a	-	III	IV
6MWD, m	>440	320–440	165–319	<165
BNP or NT-proBNP, ^a ng/L	<50 <300	50–199 300–649	200–800 650–1100	>800 >1100

6MWD, 6-minute walking distance; BNP, brain natriuretic peptide; NT-proBNP, N-terminal pro-brain natriuretic peptide; WHO-FC, World Health Organization functional class.

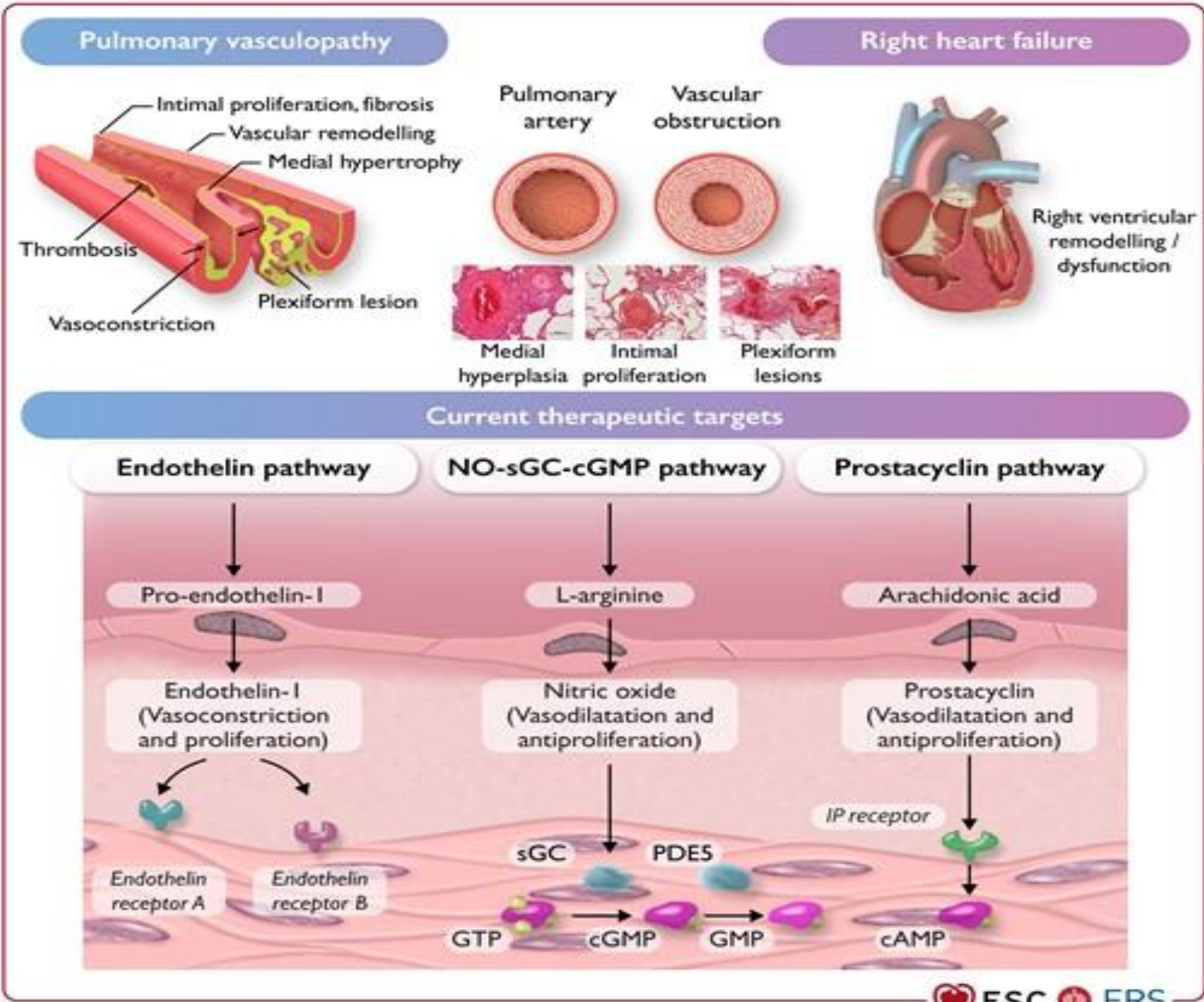


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Starting and Target Dosing of PAH Medications

Calcium channel blockers

Amlodipine	5 mg o.d.	15–30 mg o.d. ^a
Diltiazem	60 mg b.i.d. ^b	120–360 mg b.i.d. ^b
Felodipine	5 mg o.d.	15–30 mg o.d. ^a
Nifedipine	10 mg t.i.d.	20–60 mg b.i.d. or t.i.d.

Endothelin receptor antagonists (oral administration)

Ambrisentan	5 mg o.d.	10 mg o.d.
Bosentan	62.5 mg b.i.d.	125 mg b.i.d.
Macitentan	10 mg o.d.	10 mg o.d.

Phosphodiesterase 5 inhibitors (oral administration)

Sildenafil	20 mg t.i.d.	20 mg t.i.d. ^c
Tadalafil	20 or 40 mg o.d.	40 mg o.d.

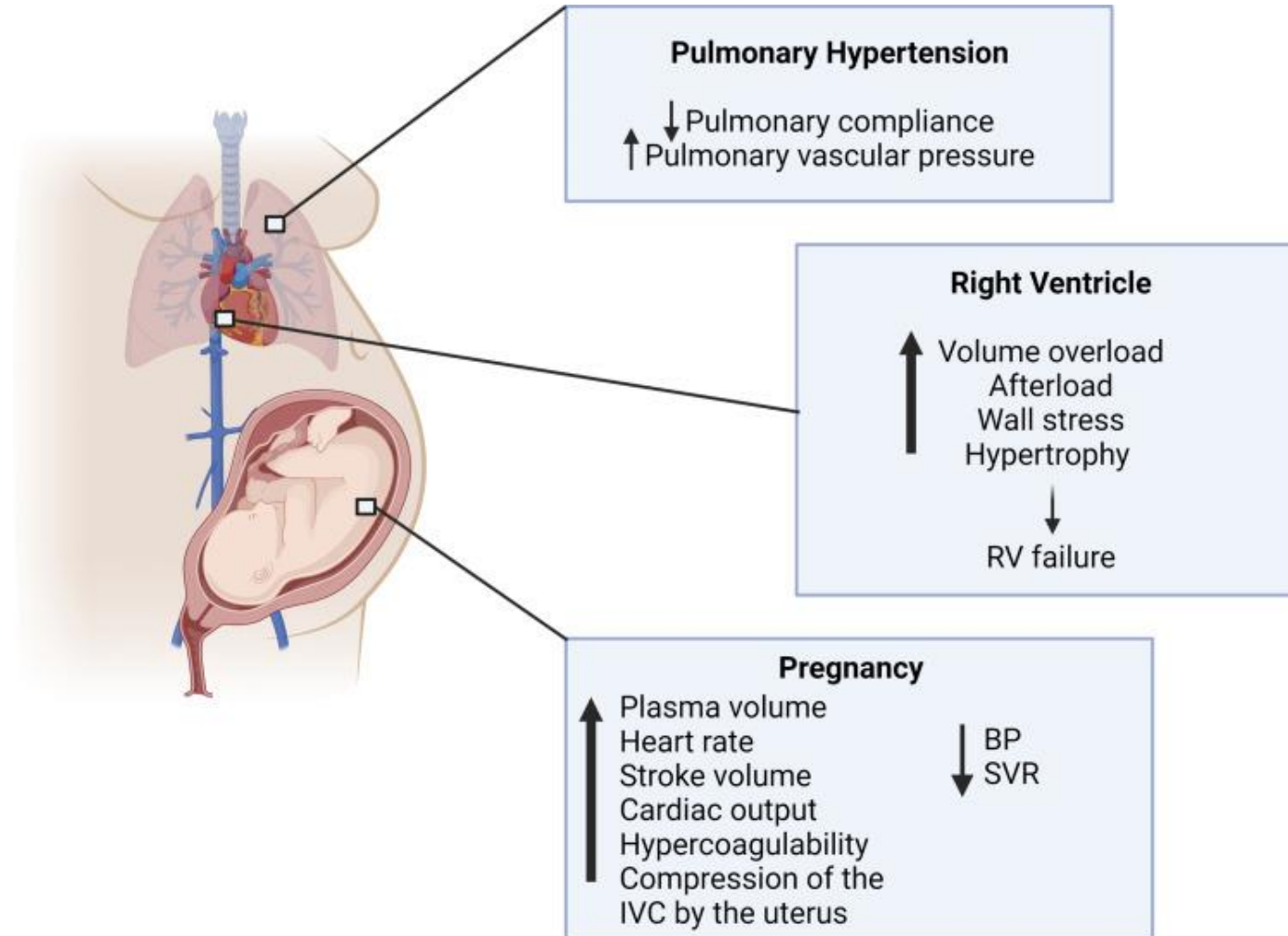
Prostacyclin analogues (oral administration)

Beraprost sodium	20 µg t.i.d.	Maximum tolerated dose up to 40 µg t.i.d.
Beraprost extended release	60 µg b.i.d.	Maximum tolerated dose up to 180 µg b.i.d.
Treprostinil	0.25 mg b.i.d. or 0.125 mg t.i.d.	Maximum tolerated dose

Prostacyclin receptor agonist (oral administration)

Selexipag	200 µg b.i.d.	Maximum tolerated dose up to 1600 µg b.i.d.
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Pregnancy and PH



Historical Perspective

- Historically, pregnancy in women with PAH and other forms of severe PH has been associated with maternal mortality rates of up to 56% and neonatal mortality rates of up to 13%.
- With improved treatment of PAH and new approaches to managing women during pregnancy and the peri-partum period, maternal mortality has declined but remains high, ranging 11-25%.
- Pregnancy remains associated with unforeseeable risks, deterioration can occur at any time during or after pregnancy, and pregnancy may accelerate PAH progression.
- Previous guidelines have recommended that patients with PAH should avoid pregnancy.

Current Recommendations

- Women with poorly controlled disease, indicated by an intermediate- or high-risk profile and signs of RV dysfunction, are at high risk of adverse outcomes; in the event of pregnancy, they should be carefully counselled, and early termination should be advised.
- For patients with well-controlled disease, a low-risk profile, and normal or near-normal resting hemodynamics who consider becoming pregnant, individual counselling and shared decision-making are recommended.

Pharmacological Management

- PH specific therapies during pregnancy:
 - Calcium channel blockers are generally well-tolerated during pregnancy and are not associated with a teratogenic risk.
 - Prostacyclins and PDE5i can be considered during pregnancy
 - Women who are chronically on medications with potential teratogenic effects prior to pregnancy are advised to switch to a different class before attempting pregnancy.
- Anticoagulation during pregnancy:
 - Low molecular weight heparin or unfractionated heparin is recommended for idiopathic and hereditary PAH, as well as chronic thromboembolic pulmonary hypertension (CTEPH).
 - Low-dose aspirin might also be considered to lower the risk of pre-eclampsia.

Antepartum Care Coordination

- Referral to specialized PH facilities with extracorporeal membrane oxygenation (ECMO) and transplant capabilities.
- Multidisciplinary teams involving maternal fetal medicine, neonatal intensive care, pulmonology, cardiology, obstetric and cardiac anesthesiologists, critical care medicine, pharmacy, and the ECMO team are recommended.
- Elective delivery is often recommended around 34–36 weeks of pregnancy, but this practice is geographically variable.

Labor and Delivery

- Use “bubble trap” 0.2-micron air filters for IV lines to reduce risk of paradoxical air embolism in patients who have or may potentially have right-to-left intracardiac shunts.
- Assisted vaginal delivery is considered safe in some patients with PH associated with CHD, and for those in functional class I or II.
- If cesarean section is considered, epidural anesthesia is preferred due to the negative effects of spinal and general anesthesia on myocardial function and increased PVR.
- Routine use of pulmonary artery monitoring catheters is not generally recommended, but careful monitoring of vital signs, ECG, and pulse oximetry in an intensive care setting is prudent.

Post-Partum Care

- For prevention of postpartum hemorrhage, slow and low-dose oxytocin intravenous infusion or intrauterine injection is recommended when hemostasis is necessary. Routine dosing may precipitate a pulmonary hypertensive crisis with marked systemic hypotension.
- Ergometrine or prostaglandins are contraindicated given their vasoconstrictive effects on the pulmonary vasculature.
- The first 24–36 hours of the post-partum period is a critical time, carrying the highest risk of maternal morbidity and mortality.

Post-Partum Care

- Anticoagulation is usually preferred during the first postpartum week in PH patients, especially if they are confined to bed and non-ambulant. Those already on anticoagulation should resume therapy once hemostasis is achieved.
- If appropriate, medications with potential teratogenic effects such as endothelin-receptor antagonists may be re-introduced promptly after delivery.
- Breastfeeding while on sildenafil, treprostinil and bosentan has been anecdotally reported, however; strong safety data are lacking.

Contraception in Women with PH

- Many forms of contraception, including oral contraceptives, are highly effective.
- In patients treated with bosentan, reduced efficacy of hormonal contraceptives should be carefully considered.
- Using hormonal implants or an intrauterine device are alternative options with low failure rates.
- Surgical options may be considered but are associated with peri-operative risks.
- Emergency post-coital hormonal contraception is safe in PH.

Clinical Scenario:

Congenital Heart Disease (CHD)

- 30-year-old woman with unrepaired large ventricular septal defect. On no medications. No known prior pregnancies.
- Has no symptoms at rest or with ordinary activity but has never been able to run or vigorously exercise without rapidly becoming short of breath. No history of arrhythmia, syncope, or hemoptysis.
- BMI 22, BP 110/70 mmHg, SpO₂ 85% in room air, nailbeds are clubbed.
- She reports a positive home pregnancy test, and you subsequently confirm intrauterine pregnancy with estimated gestational age 10 weeks. *She wants your advice...*

Question #1

- What tools would your expert PH referral center multidisciplinary team use to assess her current cardiovascular adaptation to 1st trimester of pregnancy and help inform her prognosis?

Question #2

- She has the following test results:
 - ECG shows sinus rhythm.
 - Echocardiogram shows normal left ventricular ejection fraction, dilated and hypertrophied right ventricle with normal systolic function, large VSD with right-to-left shunting.
 - Hgb 19, MCV 90, plts 175,000, HIV negative, creatinine 0.6, BNP 30.
 - 6 min walk distance 510 meters.
 - Right heart catheterization confirms systemic level pulmonary artery pressures and no significant vasoreactivity to pulmonary vasodilator drug trial.
- How would you advise her regarding continuation of pregnancy?

Question #3

- You correctly told her that her pregnancy is very high-risk and can result in life-changing complications, acceleration / worsening of her heart disease even after pregnancy and could have a mortality risk as high as 25%. She carefully considers your advice and decides to move forward with pregnancy. What is your multidisciplinary team going to recommend next?

Question #4

- She follows the advice of your multidisciplinary team. Consultants in MFM, neonatology, adult congenital heart disease, obstetric anesthesia, critical care medicine, and cardiothoracic surgery have met with her. She accepts your recommendation for aspirin 81 mg chewable once daily. A fetal echocardiogram at 22 wk gestation is normal. At 36 wk she reports new onset dyspnea at rest, she has lower extremity edema but is not hypertensive. SpO₂ is now lower at 80% in room air. BNP is higher at 180. You decide it is time to deliver the baby. What mode of delivery, obstetric anesthesia choice, and post-partum observation plan do you advise?

Outcome

- You rightly assess her condition to be worsening at 36 wk gestation and all members of the expert multidisciplinary team agree. Although assisted vaginal delivery option was given consideration, practical aspects of bringing together all contingency support services at your center favors a planned cesarean section delivery instead. An epidural anesthetic is administered, and all IV lines are protected from air embolisms with bubble filters. Her baby is delivered free of apparent health problems and admitted to NICU. She recovers for the first 2 nights after delivery in your intensive care unit with no significant obstetrical bleeding and no new complaints. Her adult congenital cardiology consultant offers to buy every team member a drink!

Thank you! Do good things!

Joel T. Hardin MD

joel.hardin@emory.edu

(404) 778-5545 office

(404) 778-5035 FAX