

Pulmonary Arterial Hypertension in Pregnancy

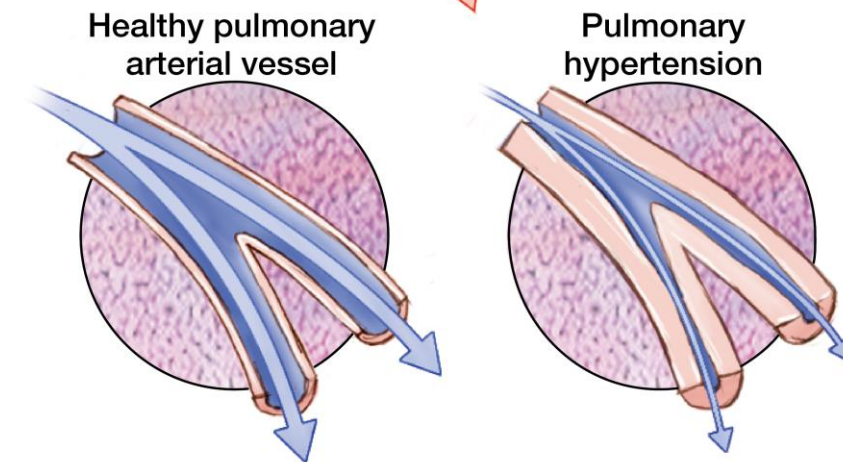
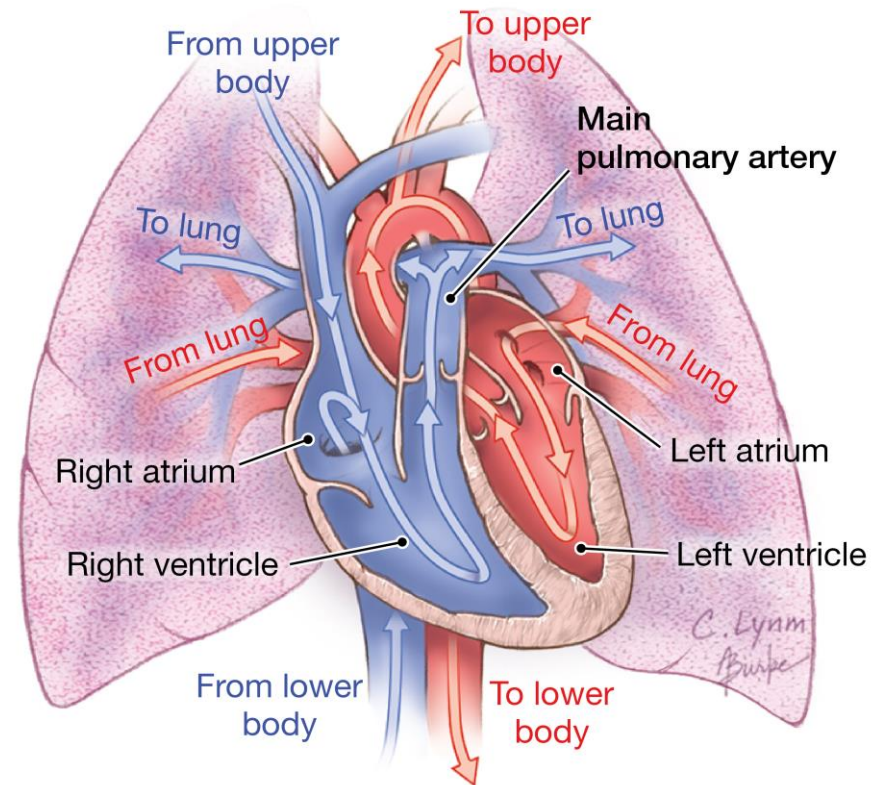
Brandon Jakubowski, MD

Assistant Professor

Pulmonary Critical Care
Medicine

Assistant Director Pulmonary
Vascular Disease Program

UT Southwestern Medical
Center



Learning Objective S

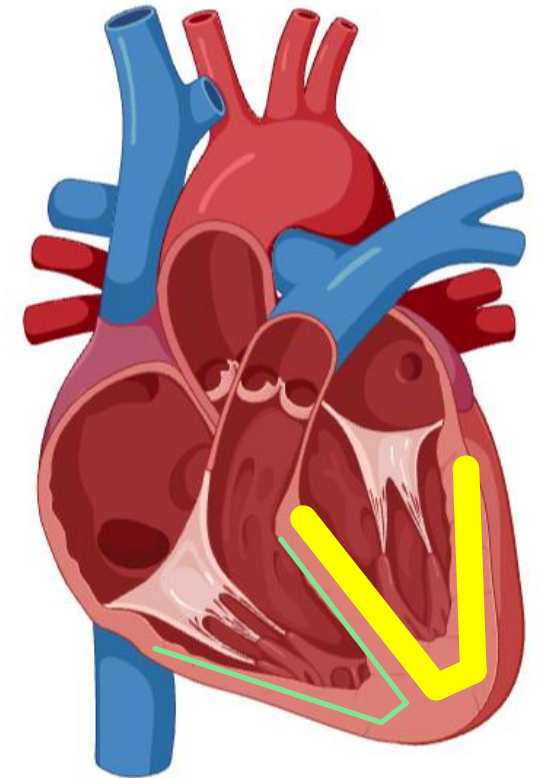
1. Define pulmonary arterial hypertension (PAH) & understand the epidemiology
2. Presentation & work-up of PAH in pregnancy
3. Review the maternal PAH risk
4. Treatment of PAH in pregnancy
5. Peri-partum considerations
6. Importance of contraception in PAH

Definition

Pulmonary
Lungs

Hypertens
ion
Elevated
BP

Systemic Circulation	Pulmonary Circulation
Normal systemic BP = 120/80	Normal pulmonary BP = 25/10
Supports entire cardiac output = ~6L/min	
Arteries are thick-walled, have a higher proportion of smooth muscle	Thinner walls, with less smooth muscle and elastic tissue
Hypertensive system	
"Silent killer"	Insidious, non-specific symptoms
CVA, MI, CKD, Left-heart failure	Right-heart failure
	Requires an index of suspicion



Clinical Presentation & Work-Up

Symptoms of Pregnancy	Symptoms of PAH
	Nausea
	Early satiety
	Dyspnea on exertion
	Fatigue
	Weight gain
	Fluid retention

Symptoms/signs of concern

- Dyspnea out of proportion to gestational age or rapidly worsening dyspnea symptoms
- History of associated conditions
- Any degree of hypoxemia
- Exertional near-syncope or syncope
- Chest pain with exertion
- Signs of right-heart failure
- Dysphonia

PAH suspected

TTE

RHC

Epidemiology

PAH affects
15 to ~60
people per 1 million
individuals

~1000
NEW CASES



~1000 new cases are diagnosed in the United States every year

PAH is more commonly diagnosed in women (70%-80% of cases)



Average age: 35 years

Rare
disease



Young
females

Maternal Outcomes

	Country	Study type	# pts	PAH	C-section	Post-partum death	Neonatal outcomes	Misc
Selected data on pregnancy outcomes in pts with PH published 2017-2026	Meng 2017	USA*	49	30 PAH 7 CTEPH	22/41 (54%)	8/49 (16%)	5 terminations 1 IUFD No neonatal death	ECMO used in 6 pts, all of whom died
	Lim 2019	South Korea	9	7 CHD-PAH 2 IPAH/HPAH	5/10 (50%)	0/10 (0%)	10/10 (100%) alive	
	Kamp 2011	Germany	13	9 IPAH 2 CTD-PAH 2 CHD-PAH	17/17 (100%)	0/17 (0%)	18/18 (100%) alive	1 pt required ECMO → lung transplant
	Corbach 2021	Switzerland	5	3 IPAH 1 CTD-PAH 1 Schistosomiasis	7/7 (100%)	0/5 (0%)	7/7 (100%) alive	
	Marshall 2022	USA	70	12 PAH	38/70 (58%)	0/70 (0%)	1 stillbirth No neonatal death	
	Vaidya 2022	USA	6	3 IPAH 2 CHD-PAH 1 CTD-PAH	4/7 (57%) 3 VAVD	0/6 (0%)	7/7 (100%) alive	
	Boyers 2022	Australia	6	6 UPAH	2/7 (29%)	0/6 (0%)	2 abortions No neonatal	

Registry Data

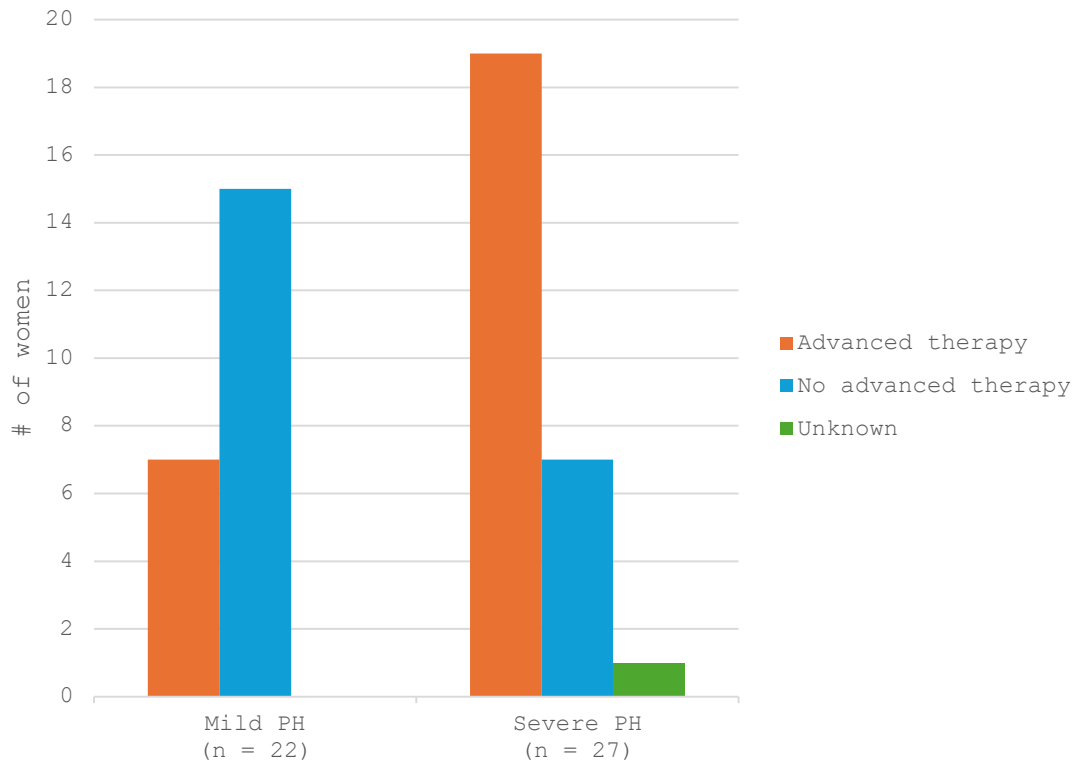
NIS: 1-2/10,000 patients hospitalized for delivery

ROPAC: ~5% of maternal CV disease

Maternal mortality is high - ~5-30%

Maternal Outcomes

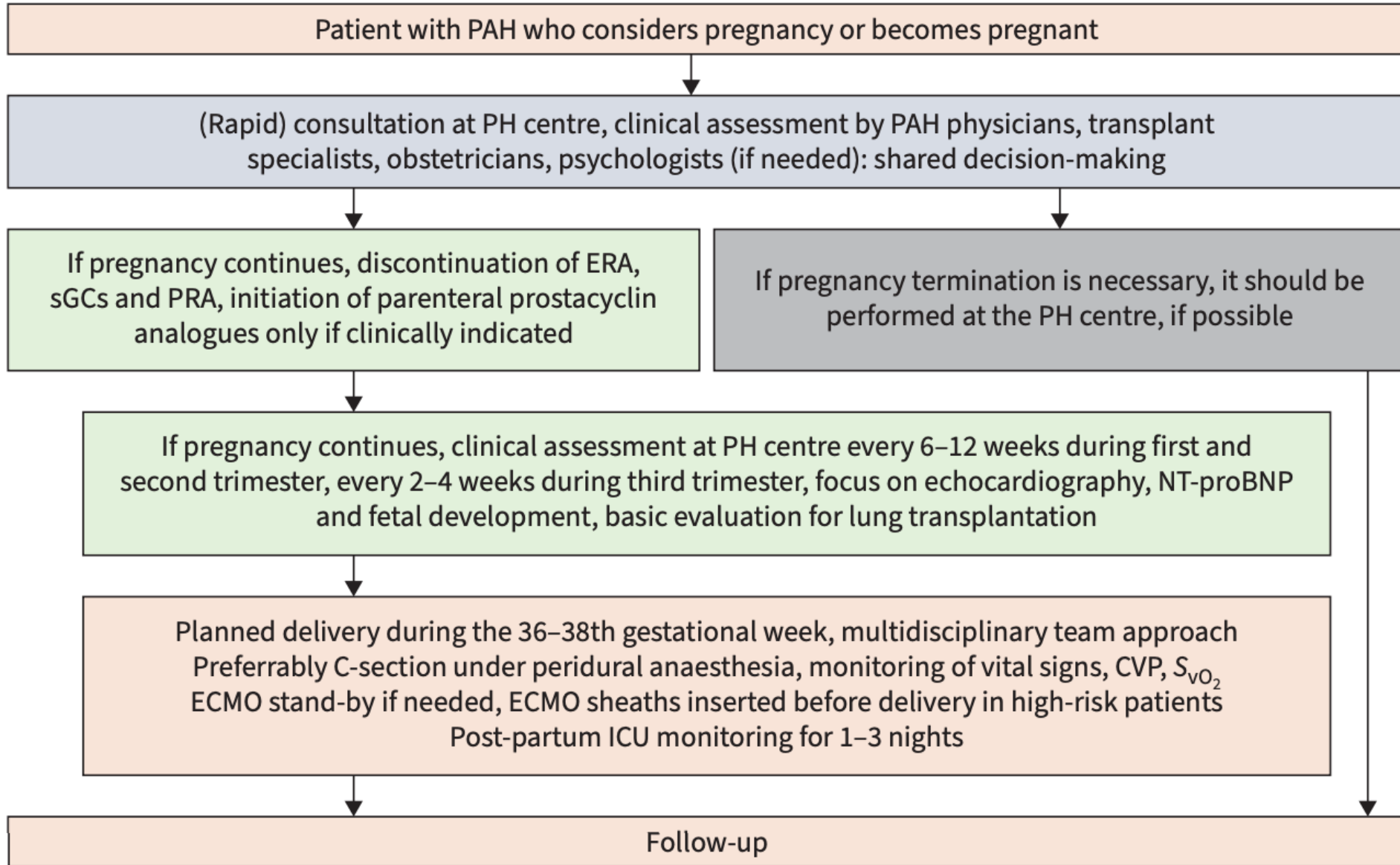
	Country	Study type	# pts	PAH	C-section	Post-partum death	Neonatal outcomes	Misc
Meng 2017	USA*		49	30 PAH 7 CTEPH	22/41 (54%)	8/49 (16%)	5 terminations 1 IUFD No neonatal death	ECMO used in 6 pts, all of whom died



Trimester	Plan
1 st	Monthly visits
2 nd	Biweekly visits
3 rd	Weekly visits
Admission	If RV failure suspected

Delivery method	Neuraxial analgesia
25 vaginal	22/25
22 cesarean	20/22

Pre-term
8 mild
19 severe



PAH Management

Mechanism of action	Medication	Potentially Teratogenic	Route of Administration
Nitric oxide enhancers	Sildenafil	<input type="checkbox"/>	Oral
	Tadalafil	<input type="checkbox"/>	Oral
	Riociguat	<input checked="" type="checkbox"/>	Oral
Endothelin receptor antagonists	Ambrisentan	<input checked="" type="checkbox"/>	Oral
	Macitentan	<input checked="" type="checkbox"/>	Oral
	Bosentan	<input checked="" type="checkbox"/>	Oral
Activin signaling inhibitors	Sotatercept	<input checked="" type="checkbox"/>	Subq
	Selexipag	<input checked="" type="checkbox"/>	Oral
Prostacyclin pathway agents	Treprostinil	<input type="checkbox"/>	Oral IV Subq Inh
	Epoprostenol	<input type="checkbox"/>	IV

Increasing efficacy & Increasing teratogenicity

PAH treatment armamentarium is littered with medications that are potentially teratogenic

Significant AEs are associated with most potent of PAH therapies

Drug delivery mechanisms are high-risk

- Not ideal to have to start in an "emergent" setting

Prepartum

PH, MFM, lung transplant, anesthesiology coordination

Care plan discussions (delivery method, location, anesthetic approach)

Bi-monthly → weekly outpt visits with surveillance imaging/screening



Peripartum

Lines: arterial monitoring & CVC/PICC
Consider PAC

ECMO anticipation?
Place 5F sheaths prior to labor

Oxytocin may raise PVR and PAP, administer slowly and consider inotrope
Terbutaline is safe

Methylergonovine and Carboprost should be avoided if possible (raise PVR, cause bronchospasm, etc)



Postpartum

ICU monitoring for 24-72hrs

Recognize early RV failure, monitor volume status, liberal use of inotropes

Effects of Pregnancy/Delivery

Pregnancy

- Fixed pulmonary vascular cannot accommodate \uparrow CO \rightarrow RHF
- \downarrow SVR \rightarrow \downarrow coronary perfusion pressure \rightarrow RHF
- \uparrow blood volume \rightarrow \uparrow RV preload in a pressure-overloaded system
- VQ mismatch due to \downarrow FRC
- \uparrow Oxygen demand/consumption
- Hypercoagulability

Delivery

- Labor-associated auto-transfusion
- Preload changes (relief of IVC compression)
- Neurohormonal stress \rightarrow precipitate arrhythmias

Management Considerations

Minimize PVR

- Administer supplemental O₂
- Avoid over-sedation
- Avoid hypercapnia
- Maintain effective neuraxial analgesia

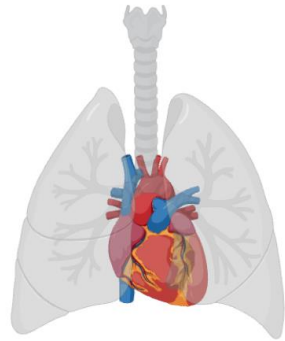
Maintain adequate blood volume and venous return

- Strict monitoring of fluid balance
- Early recognition of hypertensive disorders of pregnancy
- Early recognition and aggressive response to hemorrhage

Avoid myocardial depressants

- No beta-blockers
- Monitor arrhythmia
- Arterial BP monitoring
- Careful titration of local anesthetic
- Titrate oxytocin carefully
- Invasive PAC may be needed

Key Point



PAH
managemen
t

Coordination
of care
Planning

Preconception
Counseling

Guideline/ Society	Stance on Pregnancy	Language
ESC/ERS	Strongly discouraged	"Pregnancy in PAH associated with very high maternal morbidity and mortality"
ESC	Contraindicated	"PAH classified as WHO class IV"
PVRI	Advised against	"High maternal mortality persists despite modern therapy"
ISHLT	Generally contraindicated	"Severe PAH viewed as incompatible with safe pregnancy"
SMFM	Strongly discouraged; termination should be discussed	"PAH associated with extreme maternal risk, especially peripartum and postpartum"
ACOG	Avoid pregnancy in high-risk cardiac disease	"PAH considered among highest-risk cardiac conditions"
WHO		"discouraged"

Preconception/Contraceptive Counseling

- Discuss reproductive goals → short-term vs long-term planning (LARCs)
- Review morbidity/mortality and neonatal outcomes with pt
- Review teratogenic medications
- Urge patient of timely notification to clinic for unintended pregnancy

MOC Question

A 29yo G1P0 presents to her obstetrician's office at 24 weeks. She is reporting progressive dyspnea; notably, she has difficulty climbing two flights of stairs and is feeling more fatigued than she expected for pregnancy. Her pregnancy has been uncomplicated. Her vitals are notable for a HR of 98, BP of 104/64, RR of 20, and SpO2 of 95% on room air. She has a gravid abdomen appropriate for gestational age and trace lower extremity edema. On review of systems, she otherwise endorses morning nausea, early satiety, several episodes of near-syncope, ankle swelling, and weight gain.

Her obstetrician is concerned about possible PAH in pregnancy. Which of the following symptoms is the most concerning for PAH pulmonary arterial hypertension in pregnancy?

- A. Dyspnea with exertion
- B. Exertional near-syncope
- C. Lower extremity edema
- D. Morning nausea

Nursing Pearls for PAH

Early
recognition
of PAH

Vitals are
vital in PAH

IV fluids are
not our
friend

Protect the
(IV) line!

PAH patients
deteriorate
shockingly
quickly

Take Home Points

1. PAH is a hemodynamically defined disease that is initially suspected based on symptomatology and abnormal echocardiographic findings
2. PAH commonly affects a population with child-bearing potential
3. Maternal outcomes with PAH and pregnancy remain suboptimal
4. All pregnant patients with PAH should be managed by a multidisciplinary team of PAH specialists, MFMs, and obstetrical anesthesiologists
5. Close monitoring throughout all trimesters is essential for good outcomes
6. Preconception counseling is a critical piece of routine PAH care