Severe Pulmonary Arterial Hypertension in a 26-Week Pregnancy Complicated by Systemic Lupus Erythematosus

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Abstract

Pulmonary arterial hypertension is a rare condition characterized by vascular proliferation and remodeling of the pulmonary vessels. Despite improvements with modern management, pulmonary arterial hypertension continues to be associated with high rates of morbidity and mortality.i This risk increases further during pregnancy, as physiologic changes impose a significant threat to safety.ii This report describes the management of a pregnant patient with severe pulmonary arterial hypertension complicated by systemic lupus erythematosus. A 26-year-old G2P0101 at 26+1 weeks gestation with severe pulmonary arterial hypertension, systemic lupus erythematosus, and chronic hypoxemic respiratory failure, presented with progressive dyspnea and hypoxemia. She required immediate ICU admission. Echocardiogram estimated a right ventricular systolic pressure of 100-110 mmHg. A multidisciplinary team managed her with advanced vasodilator therapy, diuresis, oxygen supplementation. They planned for controlled preterm surgical delivery with regional anesthesia. This case highlights the unstable hemodynamic balance in pregnant patients with advanced pulmonary arterial hypertension complicated by autoimmune disease.

Background

Pulmonary arterial hypertension (PAH) is a serious, life-threatening condition characterized by endothelial dysfunction, smooth muscle hypertrophy, and intimal fibrosis. This ultimately leads to an increase in pulmonary vascular resistance (PVR) and right ventricular failure.[1] This case report describes PAH in the setting of pregnancy. Physiologic changes during pregnancy significantly worsen PAH due to increases in blood volume, cardiac output, and oxygen demand, along with decreases in functional residual capacity (FRC) and oxygen reserves. In a healthy pregnancy, the pulmonary vessels respond to these changes through vasodilation; however, in PAH, the pulmonary arteries are structurally remodeled, resulting in fixed vascular resistance and increased right ventricular (RV) afterload (Figure 1).[2] Hemodynamic management is challenging because these patients are highly preload dependent and tolerate a very limited physiologic range – excess volume worsens RV dilation and congestion, while reduced preload decreases cardiac output and can contribute to cardiovascular collapse. Due to these hemodynamic demands and associated maternal risk, pregnancy is strongly discouraged in women with PAH. It is grouped within the modified WHO Class IV cardiovascular risk category, indicating an extremely high likelihood of morbidity or mortality.[3] When pregnancy does continue, delivery is typically planned by cesarean section in a

controlled setting. Regional anesthesia is preferred to maintain spontaneous ventilation and avoid the increased PVR associated with general anesthesia. This approach can be achieved using spinal, epidural, or combined spinal-epidural technique. RV systolic pressure thresholds are commonly used to categorize PAH severity (Table 1), with pressures ≥60 mmHg indicating severe disease.[4]

Table 1. Pulmonary arterial hypertension severity classification.

PAH Severity	RV Systolic Pressure (mmHg)
Normal	<40
Mild	40-49
Moderate	50-59
Severe	≥60

The complexity of PAH increases when it occurs in the setting of systemic lupus erythematosus (SLE), which in this case was the likely underlying etiology contributing to vascular remodeling, pulmonary involvement, and thromboembolic risk. Pregnancy presented an additional physiologic strain and her decompensation at 26 weeks' gestation was driven in part by worsening ventilation-perfusion (V/Q) mismatch and declining gas exchange. Despite counseling regarding the markedly increased maternal mortality risk and strong recommendations to avoid continuing the pregnancy, she elected to proceed. This case highlights the challenges of managing maternal and fetal safety in the setting of severe PAH complicated by autoimmune disease.

Case Presentation

A 26-year-old G2P0101 woman at 26+1 weeks gestation was transferred to a high-risk obstetric ICU on August 8, 2025, for worsening respiratory distress and decreased fetal movement. Her medical history included severe PAH, SLE, chronic hypoxemic respiratory failure, hypothyroidism, cerebrovascular accident (CVA) and pulmonary embolism (PE). She had been maintained on advanced PAH therapy, including continuous IV treprostinil, sildenafil, diuretics, inhalers, and chronic steroid and immunosuppressives for SLE.[2,6]

This patient had a cardiology visit on July 15, 2025 (23 weeks EGA), where she reported worsening dyspnea, increasing oxygen requirements, tachycardia, abdominal pain, and fatigue. Chest CT angiography at that time revealed bilateral ground-glass opacities, likely related to either interstitial lung disease or pulmonary edema. Her transthoracic echocardiogram (TTE) findings are summarized in Table 2.

These findings are consistent with PAH-induced cor pulmonale. Her TTE results from the year prior (August 2024) reported a RV systolic pressure of 92 mmHg, demonstrating progressive PAH. These findings were exacerbated a year later during her pregnancy (Table 2).

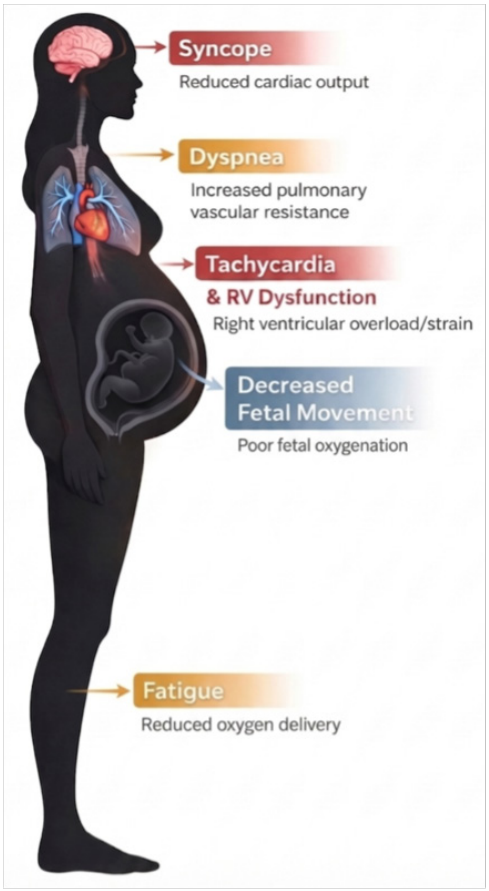

Figure 1. Clinical manifestations and pathophysiological mechanisms of PAH in pregnancy. Diagram illustrates key maternal and fetal manifestations associated with PAH during pregnancy, including syncope, dyspnea, tachycardia and RV dysfunction, fatigue, and decreased fetal movement.

Table 2. Echocardiogram results from August 6, 2025.

Parameter	Result
RV size	Severely dilated
RV systolic function	Severely reduced
RV systolic pressure	100-110 mmHg
Septum	D-shaped LV from septal flattening
RA	Moderately dilated
LV ejection fraction	~60%
IVC	Mildly dilated
Valvular findings	Severe tricuspid regurgitation

Investigation

On August 8, 2025 (26 weeks EGA), she presented to the emergency department in severe respiratory distress with shortness of breath and required 2-5 L/min supplemental oxygen to maintain adequate oxygenation. She was tachycardic and tachypneic but alert and cooperative. In the context of worsening RV dysfunction and ongoing clinical instability, the maternal fetal medicine team again counseled the patient that both maternal and fetal mortality risks were significantly elevated. At that point, the decision was made to optimize the patient for delivery, with continued multidisciplinary monitoring and hemodynamic stabilization.

During her hospitalization, the patient received continuous IV treprostinil for management of her PAH. Oxygen supplementation was maintained with a target saturation above 95%, and the patient was being closely monitored.
A cardiology consult on August 10, 2025, confirmed persistent symptoms consistent with advanced PAH with cor pulmonale. The patient continued IV treprostinil and inhaled nitric oxide 20 ppm to reduce preload. Her established treatment regimens for SLE, hypothyroidism, and history of PE were continued. There was no further diuresis at this point, as her volume status was managed with a goal central venous pressure (CVP) of 8-12 mmHg.

Investigation & Treatment

On August 10, 2025, the anesthesiologist completed a pre-operative evaluation in anticipation of a high-risk cesarean section. The patient was classified as American Society of Anesthesiologists (ASA) class IV, which indicates there is severe systemic disease that poses a constant threat to life.[7] According to the Modified WHO Maternal Cardiovascular Risk Classification, severe PAH falls under Class IV, in which pregnancy is strongly contraindicated due to extremely high maternal mortality risk.[3] Given this patient's severe PAH, the anesthetic plan required very thoughtful consideration to avoid any physiologic triggers known to worsen PVR or impair RV function.[8,9]

General anesthesia presents significant risk in advanced PAH and was therefore avoided. Induction agents such as propofol and volatile anesthetics may cause abrupt decreases in systemic vascular resistance and blood pressure, reducing perfusion pressure to the RV.[8] In a hypertrophied and pressure-overloaded RV, systemic hypotension can rapidly lead to RV ischemia. Additionally, initiation of positive pressure ventilation increases intrathoracic pressure, reducing venous return and preload while increasing RV afterload, further compromising cardiac output.[9] Episodes of hypoxia, hypercarbia, inadequate analgesia, and acidosis can worsen pulmonary vasoconstriction.

Vaginal delivery was avoided because labor can produce rapid and unpredictable hemodynamic shifts. Pain, sympathetic activation, and the effects of uterine contractions may increase PVR and preload, while active pushing increases RV afterload. In patients with severe PAH and limited cardiac reserve, these changes substantially increase the risk of RV failure.[10]

For these reasons, a planned cesarean delivery using a controlled regional anesthetic technique was determined to be the safest approach. The patient underwent monitored anesthesia care (MAC) with neuraxial anesthesia for cesarean delivery and bilateral salpingectomy. She remained awake, breathing spontaneously throughout the procedure. A combined spinal epidural neuraxial block was performed at the L2-L3 level around 08:30. Mepivacaine 1.5% at a volume of 0.5 mL was given instead of hyperbaric bupivacaine for its shorter duration while balancing the risk of transient neurologic symptoms.[11] A T4 level block was then attained by epidural lidocaine 2% 3 mL every 5 minutes. The final dose of 2% lidocaine was 20 mL with no hemodynamic perturbations noted. No dedicated sedative agents were administered; instead, analgesia was provided with titrated doses of fentanyl to aid in discomfort while maintaining ventilation and cardiovascular stability. This approach reflects established principles of PAH management, including avoiding hypoxia, hypercarbia, hypotension, and abrupt increases in RV afterload.[9]

Outcome

Following delivery, the patient developed worsening anemia and an anterior abdominal wall hematoma requiring interventional radiology embolization of the left inferior epigastric artery. A repeat echocardiogram on September 3, 2025, demonstrated similar findings to the echocardiogram done during pregnancy – moderately to severely reduced RV systolic function and a severely elevated RV systolic pressure of 111 mmHg.

She was hospitalized on September 3-4, 2025, for syncopal episodes associated with tachycardia, dyspnea, and chest discomfort – likely related to her severe PAH, RV dysfunction, anemia, and the physiologic stress of recent delivery.[10] She was discharged with close outpatient cardiology follow-up. As of February 2026, the patient is stable and working to find an appropriate outpatient regime.

Discussion

This case highlights the complex relationship of pregnancy physiology, severe PAH, right ventricular failure, and autoimmune disease, all of which can contribute to increased maternal risk. Managing these overlapping conditions requires early recognition, coordinated multidisciplinary intervention, and careful planning.[1]

Pregnancy imposes substantial hemodynamic and respiratory stress on patients with severe PAH, particularly in the setting of RV failure. Blood volume increases 40-50%, systemic vascular resistance decreases, and cardiac output rises by up to 50%. Additionally, the enlarged uterus compresses the diaphragm, which reduces lung volume and increases susceptibility to hypoxemia. In a normal pregnancy, these changes are usually well tolerated; however, in PAH, the risk of heart failure significantly increases.[1,10] In this patient, pre-delivery oxygenation was impaired. This impairment was consistent with PAH physiology: reduced lung volumes, high RV afterload, and impaired gas exchange.[10] Following delivery, her oxygenation showed only modest improvement, and she continued to require cardiology-directed optimization and ongoing PAH therapy.

This patient's course also emphasizes the importance of avoiding factors such as hypoxia, hypercarbia, pain, and excessive sympathetic stimulation. Excess sedation during the procedure was minimized to prevent respiratory depression and subsequent increase in PVR. General anesthesia was deferred given the high risk of sudden right ventricular collapse and the difficulty of maintaining stable PVR in severe PAH.[8,9] A combined spinal-epidural technique was chosen because it provide reliable neuraxial spread while allowing the team to slowly titrate the epidural dose. This helped limit the initial sympathectomy and maintain more stable hemodynamics during delivery.[12]

Because of the high mortality rate associated with PAH, pregnancy is generally discouraged. Patients that do become pregnant require a complex multidisciplinary approach and skilled physicians to manage the physiologic burden associated with the condition. The team usually consists of aternal etal edicine, ardiology,

nesthesiology, ynecology, ICU, and heart failure specialists.[8] Daily reassessments allowed the team to identify early signs of deterioration, such as rising oxygen needs, declining saturations, and worsening dyspnea. Continuous communication between anesthesiology and cardiology was necessary to identify and maintain the most effective methods for preserving hemodynamic stability.[9]

In patients with severe PAH, the timing of delivery is balanced between maternal deterioration, fetal maturity, and the risk of surgical intervention. In this case, early delivery planning was initiated once maternal decompensation became apparent, even though the fetus was premature.

Early planning allowed optimization of PAH-specific therapy, as well as a carefully controlled anesthetic strategy. Delaying delivery for fetal maturity alone can be dangerous if maternal decompensation is occurringhe safest time to deliver is when maternal physiology is stable.[1]

Despite an initially controlled peripartum course, the patient developed acute blood loss anemia from postoperative hematomas and later required rehospitalization for syncopal episodes. These events underscore the need for close surveillance and early follow-up inadvanced PAH, as postpartum decompensation is well recognized due to fluid shifts and the vulnerability of RV-dependent circulation.[10]

Conclusion

Severe pulmonary arterial hypertension remains one of the most high-risk encounters in maternal fetal medicine. This case illustrates how the physiological stressors of pregnancy can lead to life-threatening RV failure in patients with pre-existing PAH, especially when other comorbid conditions are present.[1]

PAH in pregnancy is classified as WHO group IV, the method of delivery becomes a crucial aspect of maternal safety, with planned cesarean section in a controlled setting often favored to avoid the hemodynamic shifts of labor. The choice of anesthesia is equally as important. A combined spinal-epidural technique provides a reliable neuraxial block while allowing slow titration to limit abrupt sympathectomy, whereas a single-shot spinal carries a higher risk of sudden hypotension in this population.[12]

Early recognition, continuous inpatient monitoring, aggressive medical therapy, and multidisciplinary planning are essential to optimizing outcomes.[9] Clinical compliance and structured follow-up remain critical, as significant maternal risk persists into the postpartum period due to ongoing hemodynamic and immunologic instability.[5,6]

Conflict of interest: The author(s) declare no conflict of interest. Funding: No funding source for this research submission.

References

1. Dominoni M, et al. Arch Gynecol Obstet. 2024;310(6):2839-2852.
2. Taichman DB, et al. Chest. 2014;146(2):449-475.
3. Regitz-Zagrosek V, et al. Eur Heart J. 2011;32(24):3147-3197.
4. Strange G, et al. J Am Coll Cardiol. 2019;73(21):2660-2672.
5. Abu-Raya B, et al. Front Immunol. 2020;11:575197.
6. Duarte AG, et al. Chest. 2013;143(5):1330-1336.
7. Doyle DJ, Garmon EH. StatPearls. 2025.
8. Rajagopal S, et al. Circulation. 2023;147(17).
9. Minai OA, et al. Chest. 2013;144(1):329-340.
10. Martin SR, Edwards A. Obstet Gynecol. 2019;134(5):974-987.
11. Tan H, et al. Adv Ther. 2022;39(5):2151-2164.
12. Roofthooft E, et al. Best Pract Res Clin Anaesthesiol. 2023;37(2):189-198.