REVIEW

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# Pulmonary hypertension in pregnancy

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Nuswil Bernolian[1], Cindy Kesty[1,2*], Putri Mirani[1], Peby Maulina Lestari[1], Abarham Martadiansyah[1], Rizky Agustria[1]

[1]Division of Maternal-Fetal-Medicine Department of Obstetrics and Gynecology dr. Mohammad Hoesin General Hospital/ Faculty of Medicine Universitas Sriwijaya Palembang; [2]WHO/TDR Clinical Research Leadership Fellow at Infectious Diseases Data Observatory, University of Oxford.

*Correspondence:
Cindy Kesty;
Division of Maternal-Fetal-Medicine Department of Obstetrics and Gynecology dr. Mohammad Hoesin General Hospital/Faculty of Medicine Universitas Sriwijaya Palembang, WHO/TDR Clinical Research Leadership Fellow at Infectious Diseases Data Observatory, University of Oxford;
cindykestyjl18@gmail.com

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## ABSTRACT

Pulmonary hypertension (PH) is a persistent increase in mean pulmonary arterial pressure (mPAP) of at least 20–25 mm Hg during right cardiac catheterization. For every million patients, there were 97 PH cases. Women are more likely than men (1.7:1) to receive a diagnosis, with a mean age of 37 years. The classification, pathophysiology, mechanism, and management of postpartum hemorrhage (PH) are the main aims of this review study. Pulmonary artery hypertension (PAH), pulmonary hypertension (PH) associated with left heart disease, pulmonary hypoxia and/or lung illnesses, chronic thromboembolic PH, and PH with unknown multifactorial processes are the five categories into which PH is divided. Women, particularly those of reproductive age, make up about 80% of individuals with idiopathic PAH. Pregnancy-related PH is one of the long-standing heart conditions with a significant morbidity and mortality rate. Its estimated death rate ranges from 30.56%. Pregnancy is therefore not advised in PH patients. Treating people with PH requires early diagnosis and effective treatment. These patients have optimism because of the impending PH medications (phosphodiesterase type 5 inhibitors, nitric oxide, endothelin receptor antagonists, and calcium channel blockers) as well as the advancements in hemodynamic monitoring and intensive care in PH specialty facilities. Pregnant women with PH should be treated with a multidisciplinary approach, such as obstetricians, cardiologists, intensivists, and neonatologists.

**Keywords:** pulmonary hypertension, pregnancy, morbidity, mortality.
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## INTRODUCTION

Pulmonary hypertension (PH), a pulmonary vasculopathy, is defined as a steady rise in mean pulmonary arterial pressure (mPAP) of at least 20–25 mm Hg following right cardiac catheterization.[1–4] The hallmarks of pulmonary arterial hypertension (PAH), which underlies PH, include left ventricular filling pressure, pulmonary arterial wedge pressure (below than 15 mm Hg), and pulmonary vascular resistance (greater than three Wood units).[1,2]

AH is characterized by intimal fibrosis, pulmonary arterial occlusion brought on by increasing medial thickening, and a typical plexiform lesion that raises pulmonary vascular pressure.[5] For every million patients, there were 97 PH cases. Women are more likely than men (1.7:1) to receive a diagnosis, with a mean age of 37 years.[1,5–7] Many of the women who suffer from idiopathic PAH are of reproductive age, accounting for over 80% of the patients.[4] Congenital heart disease (CHD) was linked to 1 out of every 4 women with PH.[1,8] Unrepaired CHD and Eisenmenger syndrome, constitute the majority of cases and significantly contribute to the total illness burden of PH in developing countries, in contrast to PAH and valvular heart disease in developed countries.[8] Rarely seen but rapidly developing is idiopathic PAH (IPAH). If PH is poorly treated, the median survival age is just 2.8 years; if PH is adequately treated, it is 5–6 years.[8]

Pregnancy can be the first time that a female PAH patient is noticeable. Pregnancy was the first time for up to 16% of primary PAH diagnoses.[1,8] Pregnancy is not suggested for these patients because of their high mortality rate, which varied from 56% in 19th-century accounts to 25% to 30% in more recent ones.[8]

Pregnancy causes a 50% rise in clotting factors, except factors XI and XIII. Reductions in anticoagulant factors such as protein S and antithrombin III occur in addition to the cessation of fibrin breakdown.[9] This woman may be more susceptible to thromboembolic events as a result of their hypercoagulable status. PH is harmful when it occurs along with venous thromboembolism.[8] A family history of PAH, HIV, and portal hypertension are predisposing factors for PAH.[10] There has never been a thorough investigation of the predisposing factors linked to the prognosis of this uncommon illness.[5] Before the development of sophisticated pulmonary vascular-targeted medicines, PAH was associated with significant rates of morbidity and mortality during pregnancy, with death rates as high as 30–56%.[2,4,5] Its rate has declined to 5–25% in recent studies.[5] Pregnancy with PH is still a challenge due to its high mortality and morbidity. This review article aims to discuss the classification, pathophysiology, and mechanism of PH during pregnancy, management during pregnancy, delivery, and postpartum period.

## CLASSIFICATION

According to etiology, PH was divided into five groups in the 2013 World Health Organization (WHO) updated classification: chronic thromboembolic PH, pulmonary artery hypertension, PH due to left heart disease, PH due to lung illnesses and/or hypoxia, and PH with unexplained multifactorial processes.[5–8] PVR ≥3 WU, PCWP ≤15 mmHg, and mPAP ≥20 mmHg would therefore be suggestive of pre-capillary PH and correspond clinically with groups 1, 3, and 4. Group 2 PH associated with left-sided cardiac dysfunction is characterized by elevated mPAP > 20 mmHg and high PCWP > 15 mmHg. It can also be differentiated into isolated post-capillary PH and mixed pre- and post-capillary PH based on PVR, where post-capillary PH is segregated using PVR <3 WU while pre- and post-capillary PH are blended using PVR ≥ 3 WU. Group 5 PH is a group of illnesses with unclear multifactorial etiology that might result in hemodynamic alterations or other PH signs. To guide the prognosis of patients with PH, both at the time of diagnosis and during follow-up, the World Health Organization (WHO) functional class (Table 1) is a useful tool.[8]

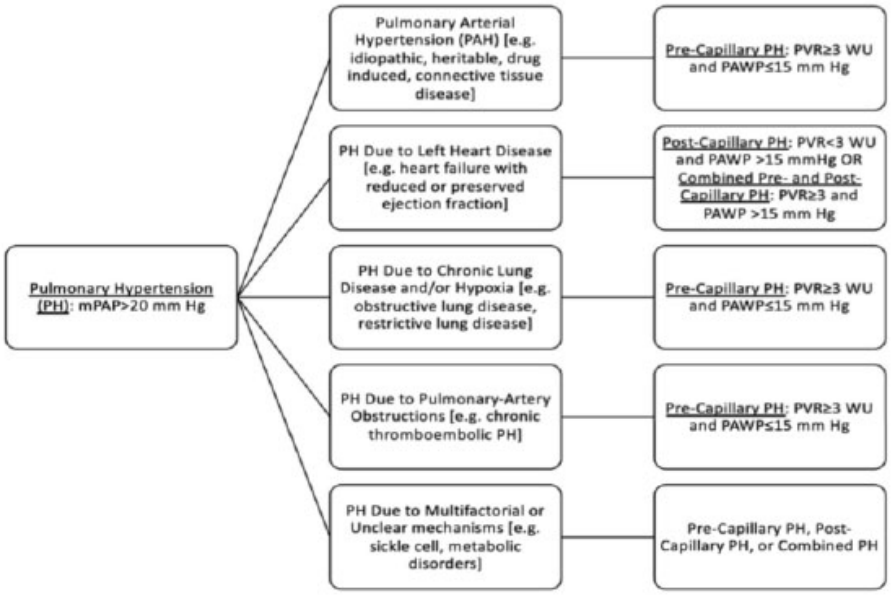


**Figure 1.** Classification of pulmonary hypertension. PAWP = pulmonary arterial wedge pressure. PVR = pulmonary vascular resistance. mPAP = mean pulmonary artery pressure. WU = Wood units.[6]

**Table 1. World Health Organization functional class description of patients with pulmonary hypertension[8]**

| Class | Description |
|---|---|
| I | No restrictions on routine physical activity; routine physical exercise does not result in exhaustion, dyspnea, chest discomfort, or presyncope |
| II | Mild physical activity restriction; minimal discomfort when at rest; nonetheless, regular activity increases dyspnea, exhaustion, chest pain, or presyncope |
| III | Marked restriction in activity; no discomfort when at rest, but greater dyspnea, exhaustion, chest pain, or presyncope when engaging in less physical activity than usual |
| IV | incapable of engaging in physical activity while at rest; symptoms exacerbated by nearly any physical exercise; may exhibit indications of RV failure |

## PATHOPHYSIOLOGY

Patients with PAH are more likely to have greater right-sided cardiac filling pressures due to their increased right ventricular afterload. Consequently, this limits the recruitment of cardiac stroke volume by increasing intravascular capacity and decreasing systemic vascular resistance.[2] The mother's hemodynamics change significantly throughout pregnancy. Reduced systemic vascular resistance, increased oxygen consumption, hypoxemia, pulmonary embolism, and deep vein thrombosis are risk factors. Hypercoagulable blood disorders are also a risk factor.[11]

Limitations in functional class and 6MWD that are unrelated to PAH or RV function may make these measurements harder to interpret as pregnancy progresses. Heart failure and cardiac strain can be objectively depicted by the biomarker BNP (B-type Natriuretic Peptide) or N-terminal-pro-BNPIn PAH, it has been demonstrated to link with both risk and prognosis. reducing the diameters of the left ventricle and left atrium reciprocally, increasing TR severity, and leftward interatrial and interventricular septal displacement are the outcomes of untreated right ventricular dilatation. An increase in cardiac output, a decrease in cardiac stroke volume, a rise in central venous pressure, and venous congestion in the liver and kidney may be the overall result.[2]

## THE MECHANISM OF PH AMONG PREGNANT WOMEN

Patients who suffer from pulmonary hypertension in mothers are susceptible to cardiac problems, such as heart failure and malignant arrhythmia. There may be a connection between cardiac risks and changes in cardiovascular physiology during pregnancy. Cardiac output rises from 50% to 70% of pre-pregnancy levels during pregnancy, as does blood volume. When a patient has pulmonary hypertension, their pulmonary vascular resistance is elevated. This results in an increased right ventricular afterload since they are unable to adjust to these changes. The detrimental cardiac conditions that could arise from this include heart failure, pulmonary hypertension crises, and impairment of cardiac function. Individuals with elevated pulmonary arterial pressure have a less favorable prognosis.[11]

A pregnant woman's pulmonary hypertension may have negative repercussions on the fetus. Fetal growth

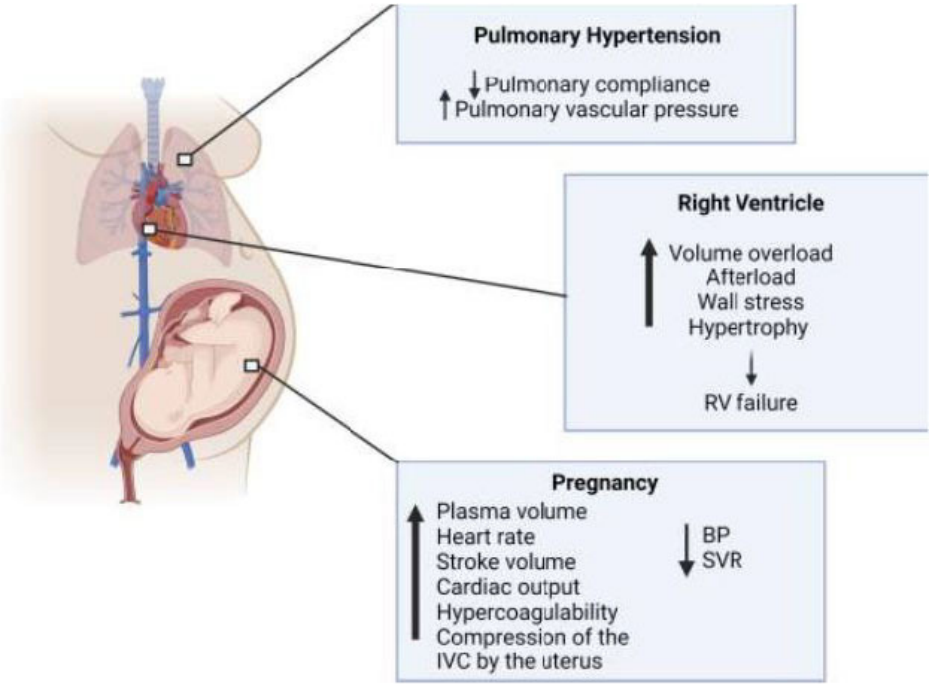


**Figure 2.** Mechanism of PH during pregnancy.[8]

and development during anoxic settings are affected by placental blood perfusion due to decreased blood oxygen levels. An embryo may suffer from this, die, or have its growth potential curtailed. Significant increases have been observed in the frequency of preterm deliveries, low birth weight newborns, and induced abortions. More patients in this group than in the mild group had moderate to severe pulmonary hypertension. They may be more likely to have low birth weights, small for gestational age babies, infant asphyxia, and neonatal deaths. There is still a significant probability of excellent delivery outcomes for women with mild pulmonary hypertension.[11]

Patients with premature heart disease (PH) often experience several physiological changes during pregnancy, something they could find hard to handle. Among the changes that could lead to right ventricular (RV) failure include increases in plasma volume, heart rate (HR), stroke volume, cardiac output, hypercoagulability, and compression of the inferior vena cava (IVC) by the uterus.[8]

Pregnancy causes a notable increase in pulmonary blood flow. On the other hand, PVR decreases due to pulmonary microvasculature recruitment and hormonal fluctuations, which leave PAP unaltered. Right ventricular (RV) failure is more likely in people with pulmonary vascular disease (PVD) due to the weakened compensatory mechanisms. Increased progesterone and estrogen levels are another side effect of pregnancy that causes vasodilation, which cause systemic vascular resistance (SVR) to decrease and diastolic blood pressure to drop dramatically. Because of the mechanical stress on the surrounding organs, there are further alterations in addition to the hormone changes. To reduce the venous return, for instance, the gravid uterus compresses the inferior vena cava while the patient is supine.[12]

Patients with PH may find it difficult to accept these changes. Low pressure and high compliance characterize the typical pulmonary circulation. The PH compliance decline is the cause of the incapacity to adjust to the increased pulmonary blood flow and cardiac output. RV dysfunction ultimately arises from an increase in end-diastolic volume and RV afterload. It is believed that the most important predictor of survival for those with PH is the fall in RV ejection fraction.[13]

All clotting factors except factors XI and XIII increase by 50% during pregnancy. Reductions in anticoagulant factors such as protein S and antithrombin III occur in addition to the cessation of fibrin breakdown.[9] This woman may be more susceptible to thromboembolic events as a result of their hypercoagulable status. PH is harmful when it occurs along with venous thromboembolism. Increased risk of pre-eclampsia has also been linked to PAH, which is linked to CHD and connective tissue illnesses.[14] RV diastolic dysfunction, a diminished strain pattern, and a slightly raised RV systolic pressure are all possible in pregnant women with pre-eclampsia. RV failure is more common in women with pre-eclampsia when there is PH, as the RV function is impaired.[15]

## COUNSELING GUIDELINES IN PREGNANT WOMEN WITH PULMONARY ARTERIAL HYPERTENSION

Women with PAH who are of childbearing age face significant hazards to both the mother and the fetus. It is therefore not advised that those women become pregnant. When PAH is diagnosed, the patient and her family should be notified of this. Every patient should also receive advice on appropriate contraceptive techniques. Although safe, barrier techniques might be unpredictable. Vasovagal responses during insertions of intrauterine copper devices (IUCDs) can have serious negative effects. Although they can result in a thromboembolic phenomenon, hormone-based treatments, such as progesterone-only pills, may be utilized. Anticoagulation does not completely prevent thrombotic episodes. For women who might want interim contraception, permanent techniques are not advised.[1] Moreover, women with PAH are advised against in vitro fertilization and ovarian hyperstimulation and venous thromboembolism (VTE) as potential risks associated with egg harvesting.[1]

The etiology of PAH is a significant factor to take into account because it influences treatment approaches. Pregnancy is not advised, for instance, if PAH is linked to connective tissue disease that is being treated with immunosuppressants.[1] Pregnancy is not advised in individuals with PAH.[2,5] The World Health Organization (WHO) cautions against pregnancy and categorizes PH as a class IV heart condition.[2]

## MANAGEMENT STRATEGIES

To optimize right heart function as much as feasible, PAH medication therapy was modified either before or during pregnancy. As a result, before birth, all patients attained low-risk PAH status. Advanced therapy modalities and the application of a multidisciplinary approach lead to improved survival rates in PAH and pregnancy. The lady should be advised to have a therapeutic abortion, preferably before 22 weeks of gestation, if she becomes pregnant because the rates are still too high. It has been demonstrated that delays longer than this increased mortality. Given the complexity of PAH in pregnancy, it is best treated in a PAH specialty facility with a multidisciplinary team that includes an intensivist, cardiologist, pulmonologist, anesthetist, geneticist, and obstetrician. Pregnant women who choose to carry their pregnancy to term should be closely observed, either through routine check-ups or voluntary hospital stays. In addition to the routine clinical assessment, there should be regular echocardiographic monitoring and fetal assessment for growth retardation. Because of the higher risk of hemodynamic problems and early labor, hospitalization in the second trimester is frequently necessary. Pregnant PAH patients do not have a set course of treatment. Patients should be consulted beforehand as management is often customized based on each patient's unique condition and the severity of their illness.[1]

The principal aim of PAH therapy is to adequately lower the PVR in order to return right ventricular size and function to a normal or near-normal range. According to new research by D'Alto et al., RV size was significantly reduced, and the RV systolic function was normalized, when the PVR was reduced by 60% or more from the baseline. These improvements in RV size and function are significantly connected with increases in submaximal exercise capacity, low-risk clinical status, and WHO functional class.[2]

Pregnancy and PAH management can be approached as pregnancy and PAH management (Figure 3).[1]

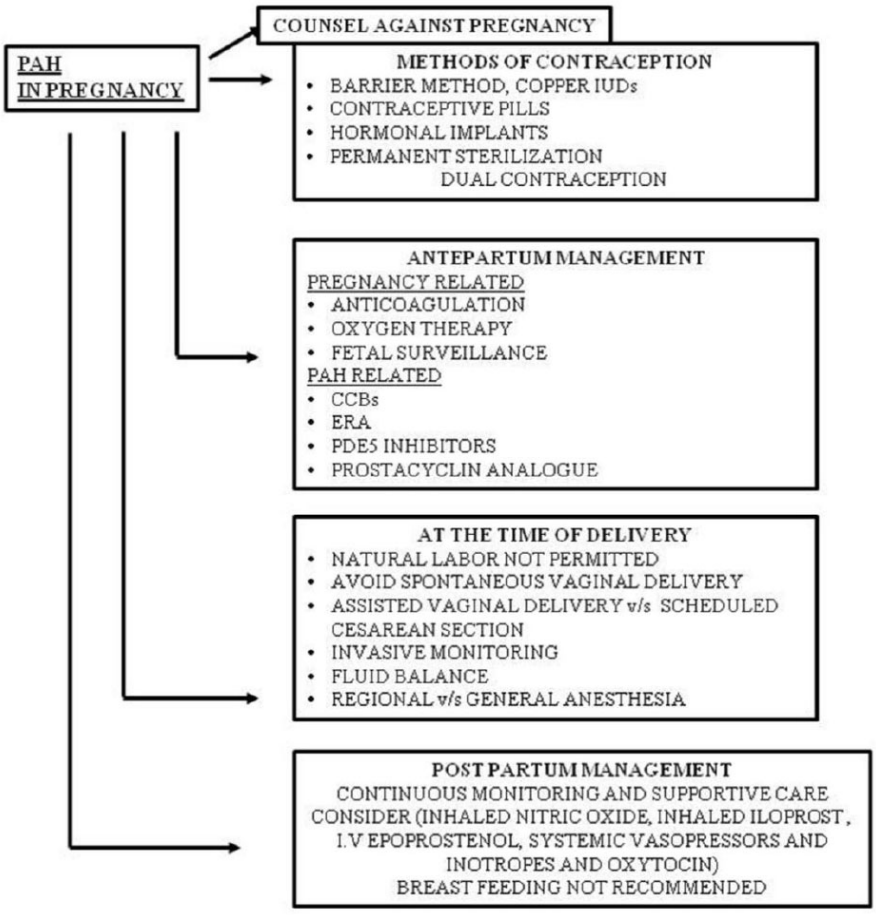


**Figure 3.** Management of PAH with pregnancy. PDE5, phosphodiesterase 5; ERA, endothelin receptor antagonist; PAH, pulmonary arterial hypertension; CCB, calcium channel blocker.[1]

### Pulmonary Arterial Hypertension (PAH) Specific Therapy

Recommendations from the European Society of Cardiology and the European Respiratory Society state that women with PAH who choose to continue their pregnancy should get treatment with medications specific to their disease, with the exception of ERAs. An increasing amount of research is showing that pregnancy can be successfully treated with PAH-specific medications, such as CCBs, phosphodiesterase type-5 inhibitors, and prostacyclin and its analogs. Nevertheless, no comparative trials of various PAH treatment regimens or controlled research have been reported to date.[16]

Pregnancy-related CCBs may be beneficial for certain PAH women, according to reports. Eight women who used CCBs and had straightforward pregnancies were found in a prospective registry of PAH patients, for instance. Additionally, there are multiple case reports detailing the successful use of intravenous (i.v.) epoprostenol in women with PAH during pregnancy and delivery, whether by cesarean section or spontaneous vaginal delivery. Additionally, pregnant women with PAH have had the benefit of inhaled iloprost. Sildenafil, a phosphodiesterase type-5 inhibitor, has been successfully used to treat pregnant PAH women in two case reports that have been published.[16]

## INFLUENCE OF PREGNANCY ON MANAGEMENT STRATEGIES

Pregnancy causes hypercoagulability since thrombotic arteriopathy is a major feature of PAH and pregnancy, which may alter the patient's course and outcome. In individuals with heritable PAH, secondary cause PAH, and IPAH, anticoagulation therapy is initiated. According to earlier research, 52–68% of women receive anticoagulant medication on average. As

anticoagulants, vitamin K antagonists are contraindicated because they induce fetal hemorrhage, spontaneous abortion, central nervous system malformation, and fetal craniofacial deformities during the first trimester. Due to its teratogenic properties during placental passage, warfarin is not recommended. Subcutaneous delivery of enoxaparin and low-molecular-weight heparin (LMWH) at a dose of 1 mg/kg is advised. Heparin serves as the mainstay of care for acute venous thromboembolic episodes during pregnancy because it does not cross the placental barrier.[1]

In addition to being symptoms of right-sided heart failure, increased blood volume and fluid retention happen during pregnancy. Therefore, it's critical to control peripheral edema. It is advised that patients avoid lying on their backs since this can put pressure on the IVC. Edema can be treated with diuretics, with torsemide and furosemide being the best options. Because of its antiandrogenic properties, spironolactone is not recommended.[1] Diuretics cause the placenta's blood flow to decrease, so they should be used carefully. When blood gas measurements suggest that a patient is hypoxic, oxygen therapy is administered.[1]

Regular monitoring at a facility where PAH patients are treated is recommended for patients who are adamant about carrying on with their pregnancy. Occasionally, admission choices could be taken into account till the birth. In addition to a clinical assessment, routine echocardiograms, and fetal well-being evaluations should be part of close observation. The possibility of hemodynamic compromise and early labor may cause hospitalization during the second trimester. When a high-risk patient has a crisis, prompt evaluation for lung transplantation may be necessary.[1]

## DELIVERY MANAGEMENT IN PULMONARY ARTERIAL HYPERTENSION

Another point of contention in PAH is delivery timing and method. Some authors suggest giving birth at approximately 34 weeks gestation since the physiological changes peak at the end of pregnancy and the cardiovascular system finds it more difficult to adjust. Patients with mild PAH who are stable and do not worsen during pregnancy can have a baby between 34 and 37 weeks gestation.[3,5]

Pregnancy should never occur spontaneously for women with PAH; instead, the delivery should always be scheduled and take place in a controlled setting. The literature contains conflicting views about the best delivery method, which is either an assisted vaginal delivery with assistance or a scheduled cesarean section. Less blood is lost (500 mL) during a typical vaginal delivery, blood volume and other hemodynamic factors change less, thromboembolic risk is reduced, and infections are decreased. However, there are drawbacks as well, such as pain and stress during the delivery process, elevated sympathetic activity that raises blood pressure, and a rate that strains the right ventricle. There's also hypercapnia, hypoxia, and acidosis brought on by childbirth. It's necessary to regularly check the intra-arterial blood pressure (BP), central venous pressure (CVP), pulse oximetry, and ECG**.** That's why an intensive care unit (ICU) setting is appropriate for construction. Decrease the pressure effect on the IVC by performing it ideally in the left lateral position. Because it constricts the pulmonary vasculature, nitrous oxide (N2O) should be avoided.[1]

According to new research, PAH may not be a total contraindication for vaginal delivery in pregnant women if pulmonary arterial pressure is appropriately controlled and the second stage of labor is shortened. Compared to the moderate to severe group, the mild group had a vaginal delivery rate that was three times higher. Consequently, vaginal deliveries were chosen and successfully performed by pregnant women with mild pulmonary hypertension.[11]

The delivery environment is more controlled during a cesarean section, on the other hand. When everything is perfect, it is done voluntarily. Anesthesia effects and a large postoperative fluid shift are its drawbacks, but it avoids protracted labor. The goal of scheduling cesarean sections is to keep the mother from going into spontaneous labor, usually between 32 to 36 weeks of gestation.[1]

According to a statement released by the Pulmonary Vascular Research Institute, elective CD is recommended between weeks 34 and 36 of pregnancy.[4,5] Nonetheless, vaginal delivery has also received support.[4] In addition to causing discomfort, vaginal birth raises intrathoracic pressures, which may reduce venous return.[5] According to recent studies, vaginal deliveries are preferred over C-sections because the latter are linked to harmful hemodynamic effects from pushing that can result in acidosis, hypercapnia, or hypoxia, as well as an increase in CO of 34% at full cervical dilation and an increase in venous return during protracted labor.[3] Options for delivery modes are addressed in a multidisciplinary manner at our institution. Vaginal birth is normally favored, but during vaginal delivery, vasovagal reactions and Valsalva maneuvers might reduce venous return, which can quickly worsen hemodynamic conditions.[4] The European Society of Cardiology's most recent guidelines state that elective cesarean or vaginal delivery carries a lower risk than intrapartum or emergency CS. General anesthesia, however, is typically necessary for patients with severe right heart failure, particularly for those with unstable hemodynamics.[5,11]

PAH should be well-regulated regardless of the dosing method. Although no trials have shown an improvement in outcomes, several facilities begin intravenous (IV) epoprostenol therapy just before delivery. There has been postpartum worsening and mortality even with well-controlled PAH before delivery. As a result, some hospitals keep administering IV epoprostenol even after delivery. One More woman will likely wish to take on the danger of having children as PAH therapy improves and more patients have low-risk prognoses.[3]

## ANESTHESIA

To avoid discomfort, acidosis, hypoxemia, hypercapnia, and PVR, which raise pulmonary pressures, anesthesia is used. It should only be administered by a qualified anesthetist to avoid uterine contractions and an increase in CO and pulmonary pressure. Comparing local and general anesthesia.[1]

**Table 2. Strengths and weaknesses of delivery methods for patients with PAH undergoing childbirth[6]**

| Delivery Method | Strengths | Weaknesses |
|---|---|---|
| **Vaginal Delivery** | • Non-surgical (lower the risk of postoperative complications and lessen the perioperative risk of intubation)<br>• Choice for altered Pushing valsava to prevent vasovagal reaction<br>• Assistance with second-stage delivery (either vacuum extraction or forceps lift-out) can lower the risk associated with valsava. | • Possibility of inadequately managed pain (which could trigger a vasovagal spiral and release catecholamines)<br>• Increased heart rate during vigorous labor, which can overpower RV<br>• Risk of hypotension or adverse drug effects in the event of a scheduled induction. |
| **Cesarean Delivery** | • Regional anesthesia is an option<br>• A controlled environment<br>• Avoiding prolonged labor<br>• Guidelines advocate this approach for patients with PAH. | • Both the likelihood of surgical problems (such the possibility of ileus) and perioperative hazards (like fluid shifts)<br>• Infection risk at the surgery site<br>• The dangers of intubation in the event that general anesthesia is needed |

**Table 3. Modes of anesthesia and delivery[1]**

| | Number | Mode of delivery | Anesthesia administration | Maternal mortality | Fetal mortality |
|---|---|---|---|---|---|
| **Bedard et al** | 73 IPAH<br>29 CHD<br>29 OPH | Vaginal 7–30%<br>CS 70–93% | Regional 28–67% | 17–33% | 10–13% |
| **Kiely et al** | 10 | CS | Regional | 10% | 0 |
| **Jais et al** | 20 | Vaginal 5%<br>CS 95% | Regional 80%<br>General 20% | 20% | 10% |
| **Duarte et al** | 12 | CS 100% | Regional 66%<br>General 25% | 16,7% | 0 |
| **Mongale et al** | 19 | Vaginal 42%<br>CS 58% | Regional 50%<br>General 50% | | |

Abbreviations: IPAH, idiopathic pulmonary arterial hypertension; CS, cesarean section; CHD, congenital heart disease OPH, other pulmonary hypertension.

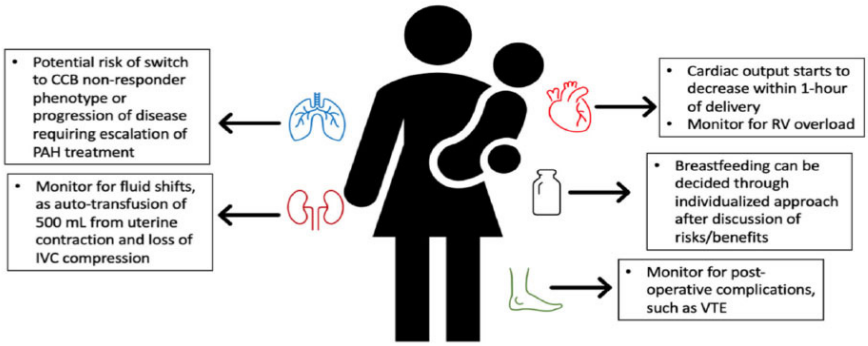


**Figure 4.** Postpartum changes in the patient with PAH.[6]

According to widespread consensus statements and clinical experience, epidural anesthesia is preferable to general anesthesia where practicable. Spinal anesthesia is generally not advised due to the risk of abrupt hypotension; however, the progressive titration of anesthetic to avoid hemodynamic fluctuations can be accomplished with an epidural or combined spinal-epidural anesthesia. In order to lessen acute peripheral vasodilation, a local anesthetic is administered in the epidural under moderate titration; however, opioid medications are usually the only ones used for spinal anesthesia. While general anesthesia has been used with success, there is a higher chance of hemodynamic instability and possible deterioration. Elevated intrathoracic and pulmonary artery pressure are the outcome of positive pressure breathing and tracheal intubation during general anesthesia. This lowers venous return, alters systemic hemodynamics, and raises the heart's workload, ultimately resulting in permanent heart failure.[9]

Sun X et al. (2018) found that only patients with an aborted fetus underwent vaginal delivery and that all patients with a gestational age of more than 24 weeks underwent cesarean procedure due to the preterm nature of all deliveries. On the ideal delivery method, there is, however, a dearth of solid information. The results of their study indicate that the death rate following a cesarean section performed under epidural/spinal anesthesia is 0.6 instead of 3/16 when general anesthesia is used. This difference in mortality may be attributable to the reduced impact of regional anesthesia on hemodynamics. For pregnancies complicated by PH, cesarean birth under regional anesthetic is advised. Patients can pass the delivery phase more effectively and safely thanks to CS, which is another factor.[5,9]

While there is a higher risk of bleeding as compared to general anesthesia, the advantages of regional anesthetic include a lesser impact on pulmonary resistance and cardiac contractions (Table 4).[1]

## POSTPARTUM CHANGES

After giving birth, all women experience a variety of physiological changes in the postpartum period, some of which may be harmful to patients with PAH (Figure 4).

## POSTNATAL CARE

Following delivery, all patients should remain under the care of pulmonary hypertension experts for at least 72 hours. This monitoring should include daily BNP testing, serial JVP (jugular venous pressure) assessments, and periodic checks for the onset of right ventricular congestion. The first month following delivery is when pregnant women with PAH die the most. Due to the contracting uterus's pressure on the abdominal aorta, which results in auto-transfusion and an increase in peripheral vascular resistance, there is a spike in blood pressure during delivery, both systolic and diastolic.[1,7] Additionally, there is a transient increase in venous return due to the gravid uterus relieving caval compression, It increases the chance of right-sided cardiac failure and hemodynamic stress. Since there is always a risk of pulmonary embolism after birth, ongoing patient monitoring and supportive care are essential to preventing right-sided cardiac failure. To treat postpartum women with PAH, medications such as inotropes, oxytocin, IV epoprostenol, inhaled NO, and inhaled iloprost are used. However, because of its tendency to induce reflex tachycardia and hypotension, oxytocin should be taken cautiously.[1]

## BREASTFEEDING

Due to the secretion of pulmonary vasodilators or PAH medication in breast milk, breastfeeding is not advised.[1,7] Furthermore, prolactin is detrimental to the heart. Therefore, careful, long-term monitoring is advised. Some medicines, such sildenafil and calcium channel blockers, are regarded to have a low risk of gravely injuring neonates even though they pass via breast milk. Following a review of the advantages and disadvantages, decisions about breastfeeding can be made on an individual basis.[6] Preterm care units should be the place where PAH moms' newborns are cared for because they are smaller than term babies.[1]

## CONTRACEPTIVE COUNSELLING CONSIDERATIONS IN PATIENTS WITH PAH

A comprehensive assessment of each person's contraception risks (such as bleeding profile, thromboembolism risk, adherence, and side effect profile), access reversibility preferences, barriers to contraception, and reproductive goals should be conducted in order to inform patient counseling.[6]

Before starting medicine, preconception counseling can be given to prevent unintentional teratogenic exposure before early pregnancy detection. Preconception counseling may also cover options for fertility preservation, such as cryopreservation of ovaries or sperm, prior to initiating teratogenic medicines. The hazards associated with general anesthesia during oocyte retrieval, however, might restrict this technique.[6]

Counseling for permanent, irreversible contraception consists of patients with serious, incurable diseases who are frequently advised to use permanent contraception. The best method for reducing procedure risks is hysteroscopic sterilization. Tubal ligation may be treated with a mini-laparotomy, but general anesthesia carries some perioperative hazards. For tubal ligation, a laparoscopic method is often less preferable due to the risks involved with the treatment.[6]

Hormone contraceptive counselling consists of using estrogen-containing contraceptives (oral, transvaginal, ring, or patch), as well as injectable progestin, increases the risk of venous thromboembolism, which can be fatal in those with compromised or compromised renal function. Since estrogen-containing contraceptives pose an unacceptable risk of developing pulmonary vascular disease and PAH, they are classified as contraceptives in WHO Class IV risk categories. In light of this, patients with PAH are typically advised against using these choices. On the other hand, when used in conjunction with anticoagulation, low-dose estrogen-containing alternatives have been explored.[6]

Oral progestin-only substitutes are not as likely to induce thrombosis, but they are generally not recommended because their ability to prevent pregnancy depends on stricter adherence. Barrier techniques are safe, but because of their greater failure rate (about 18–28% after one year), they are not usually advised. Barrier techniques are occasionally advised in conjunction with progesterone treatments to address the diminished efficacy of monotherapy. Oral contraceptive tablets and progestogen implants are less effective when taken alone as contraceptive options because of the endothelin receptor antagonist bosentan.[6]

Progestin-only intrauterine devices, also referred to as Long-Acting Reversible Contraceptive (LARC) (IUDs) are often the most recommended treatment for women with cardiovascular disease due to their effectiveness, durability, and minimal procedural risk of side events. IUD implantation carries a potential risk of vasovagal reaction, hence it is important to limit discomfort. For patients with serious diseases, inpatient monitoring during placement may be an option.[6]

## COMPLICATIONS

Although there is a greater risk for mothers and newborns, the prognosis has improved recently due to the availability of new PAH treatments. Since PAH is categorized as a class IV heart condition by the World Health Organization (WHO), pregnancy is not advised for those who have it. Newborn death (0.7%), preterm delivery (21.7%), fetal growth restriction (19%), miscarriage (5.6%), and fetal loss (2%), are examples of fetal problems. Pregnancy can happen in PAH despite these dangers.[2,17]

## CONCLUSION

The high maternal and fetal mortality rate associated with pulmonary arterial hypertension, a deadly form of pulmonary vascular disease and afterload-dependent right heart failure, has made pregnancy outcomes related to these conditions a major obstetrical concern. There is a correlation between maternal mortality and the NYHA (New York Heart Association) classification, PH severity, and delayed identification of

PH. Patients with PAH can still become pregnant, nevertheless. In such cases, it is advised that the pregnancy be ended. Yet, pregnancies can be successful, particularly in low-risk women, thanks to newly developed intensive care modules in PAH specialist centers and forthcoming PAH-specific treatments. Therefore, for this subgroup of patients, a multidisciplinary strategy should be used. When undergoing a cesarean section, regional anesthesia is preferable to a general anesthetic. To better analyze PH in pregnancy, more research evaluating it using various study designs and methodologies is required.

## DISCLOSURES

### Funding

There is no funding for this study.

### Conflict of Interest

The authors declare that there is no conflict of interest.

### Author Contribution

NB, PM, and CK are involved in concepting and designing the manuscript. PML and CK collect and filter the references. AB, CK, and RA revise and finalize this manuscript. All authors prepare the manuscript and agree for this final version of the manuscript to be submitted to this journal.

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