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Case Report

Management of Early Pregnancy in IPAH: Case Report and Review of Termination and Contraception Approaches

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Abstract

Background: Idiopathic Pulmonary Arterial Hypertension (IPAH) remains associated with substantial maternal morbidity and mortality during pregnancy, despite advances in targeted therapies. Current international guidelines strongly discourage pregnancy and advise that early termination be discussed when pregnancy occurs. This risk persists even in patients with apparently low-risk disease profiles.

Case Illustration: We report the case of a 30-year-old woman with low-risk IPAH, clinically stable on dual oral targeted therapy, who conceived after intentional discontinuation of contraception. Upon confirmation of pregnancy, endothelin receptor antagonist therapy was promptly discontinued due to teratogenic risk. At 8 weeks of gestation, the patient remained hemodynamically stable, with preserved Right Ventricular (RV) function and an estimated Systolic Pulmonary Artery Pressure (SPAP) of 60 mmHg. Following extensive counseling regarding the high maternal risk associated with pregnancy continuation, a multidisciplinary team and the patient jointly opted for early medical termination. Medical abortion with misoprostol was performed under continuous cardiopulmonary monitoring in a tertiary care center, resulting in complete uterine evacuation without maternal complications. Long-acting progestogen-only contraception was initiated post-procedure.

Conclusions: This case highlights that pregnancy in IPAH remains high risk, even in clinically stable patients receiving contemporary therapy. Early multidisciplinary evaluation, shared decision-making, and timely pregnancy termination are critical to optimizing maternal outcomes. Effective long-acting contraception is essential to prevent unintended high-risk pregnancies in this population.

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Introduction

Pulmonary Arterial Hypertension (PAH) is one of the few cardiovascular conditions in which pregnancy remains formally contraindicated due to persistently high maternal mortality.[1-2] Historically, maternal mortality rates in PAH were reported between 30% and 56%, including in Idiopathic Pulmonary Arterial Hypertension (IPAH) and PAH associated with congenital heart disease.[3-4] Although outcomes have improved in the modern management era, contemporary data continue to demonstrate maternal mortality rates ranging from 11% to 25%, with the highest risk occurring during the peripartum and early postpartum periods.[2,5-6]

The physiological adaptations of pregnancy impose a substantial hemodynamic burden, including increased plasma volume, reduced systemic vascular resistance, and a progressive rise in cardiac output. In PAH, the inability of the pulmonary circulation and right ventricle to accommodate these changes predisposes patients to Right Ventricular (RV) failure, arrhythmia, thromboembolism, and sudden cardiac death.[7-8] Consequently, international guidelines strongly discourage pregnancy in women with PAH; if pregnancy occurs, termination should be discussed, and early termination is advised when the disease is poorly controlled.[1-2]

Case Illustration

A 30-year-old woman with known IPAH presented at 8 weeks of amenorrhea for initial obstetric assessment. IPAH had been diagnosed two years earlier by right-heart catheterization demonstrating a mean pulmonary artery pressure of 35 mmHg, pulmonary vascular resistance of 5 Wood units, and normal pulmonary capillary wedge pressure, consistent with precapillary pulmonary hypertension. The patient was married and had no prior pregnancies or abortions. This was her first conception.

At diagnosis, she was classified as low risk, with World Health Organization (WHO) functional class II symptoms, preserved RV function, normal N-Terminal pro B-type Natriuretic Peptide (NT-proBNP) levels, and satisfactory six-minute walk distance. She remained clinically stable on dual oral therapy consisting of a phosphodiesterase-5 (PDE-5) inhibitor and an endothelin receptor antagonist.

Despite counseling regarding pregnancy risk, the patient intentionally discontinued contraception in order to conceive. Following confirmation of pregnancy, endothelin receptor antagonist therapy was promptly discontinued due to well-established teratogenicity.[1-2]

At 8 weeks' gestation, clinical examination revealed Blood Pressure (BP) of 110/70 mmHg, heart rate of 95 beats per minute, and oxygen saturation of 95% on room air, without signs of right heart failure. Transthoracic echocardiography demonstrated preserved RV size and systolic function, with an estimated Systolic Pulmonary Artery Pressure (SPAP) of approximately 60 mmHg. Obstetric ultrasound confirmed a viable intrauterine pregnancy.

Risk Assessment at 8 Weeks' Gestation

Using the 2022 European Society of Cardiology/ European Respiratory Society (ESC/ERS) four-strata risk assessment model, the patient was classified as intermediate-low risk at presentation. Pre-pregnancy, she was low-risk, with all three formal variables at stratum 1 (WHO functional class II, NT-proBNP normal, six-minute walk distance >440 m). At 8 weeks' gestation, clinical assessment revealed WHO functional class II (stratum 1), a reduced six-minute walk distance of 400 m (stratum 2, down from >440 m pre-pregnancy), and NT-proBNP 350 pg/mL (stratum 2, mildly elevated), alongside an echocardiographic SPAP of 60 mmHg and preserved RV function (Tricuspid Annular Plane Systolic Excursion [TAPSE] 21 mm). Averaging the three formal risk variables [(1+2+2)/3 = 1.7, rounded to stratum 2] yielded an intermediate-low risk classification. This illustrates that apparently "low-risk" patients may shift to a higher risk category during early pregnancy, taking them outside the low-risk profile for which the guidelines recommend individual counseling and shared decision-making.[2]

Management and Pregnancy Termination

Given the formally contraindicated nature of pregnancy in IPAH and the unpredictable risk of rapid clinical deterioration, extensive counseling was conducted involving cardiology, pulmonary hypertension specialists, obstetrics, anesthesia, and intensive care teams.[1-2,7] The decision to proceed with early medical termination was made by a dedicated multidisciplinary team, with each member contributing a specific assessment relevant to IPAH management during pregnancy:

- **Pulmonary Hypertension Specialist**

  Assessed disease severity using ESC/ERS 2022 risk stratification (WHO functional class, NT-proBNP, echocardiographic parameters, prior right-heart catheterization data). Evaluated the anticipated hemodynamic impact of pregnancy continuation versus termination.

Determined which PAH therapies could be safely continued or required discontinuation (endothelin receptor antagonist stopped; PDE-5 inhibitor continued). Planned contingency escalation of PAH therapy in case of hemodynamic deterioration.

- **Cardiologist**

  Performed serial transthoracic echocardiography to assess RV size, systolic function (TAPSE), estimated SPAP, and presence of pericardial effusion. Monitored for early signs of RV failure. Provided real-time hemodynamic monitoring during the termination procedure and interpreted BP, heart rate, and oxygen saturation trends.

- **Obstetrician**

  Assessed gestational age by ultrasound, evaluated cervical status, and reviewed contraindications to medical versus surgical termination. Counseled the patient on the procedural details, expected duration, pain management, and signs of incomplete abortion or hemorrhage. Performed the misoprostol administration and confirmed complete uterine evacuation.

- **Anesthesiologist**

  Assessed airway status and risk of general anesthesia if emergency intervention became necessary. Evaluated the patient's tolerance for conscious sedation versus local anesthesia alone. Developed a contingency plan for pain-induced sympathetic activation, including low-dose opioids with careful respiratory monitoring. Prepared for potential need for vasopressor support if hypotension occurred.

- **Intensive care physician**

  Assessed the patient's baseline organ function and reserve. Planned the post-procedure monitoring setting (high-dependency unit versus intensive care unit). Established thresholds for transfer to intensive care (systolic BP <90 mmHg despite fluids, SpO_2 <90% on room air, arrhythmia, altered mental status). Coordinated with the cardiology team for escalation of PAH therapy if RV failure developed.

The team jointly reviewed the patient's clinical status, discussed the risks of pregnancy continuation versus termination, and supported the patient's informed decision to proceed with early medical termination.

Vacuum aspiration is generally preferred in IPAH due to its rapid completion and predictable hemodynamic profile. However, at our institution, same-day surgical termination was not available, and the patient expressed a strong preference for immediate intervention without delay. Additionally, the patient had a favorable cervical examination and wished to avoid any instrumentation. After multidisciplinary discussion, medical termination with misoprostol was elected, with continuous hemodynamic monitoring in a tertiary care setting.

Rationale for Misoprostol Regimen

Misoprostol was given vaginally at a reduced dose of 200 μg, repeated every 6 hours. The ESC guideline suggests a reduced misoprostol dose of 100 μg for medical termination up to 9 weeks of gestation in women with heart disease,[1] whereas the standard misoprostol-only regimen before 12 weeks is 800 μg given buccally, sublingually, or vaginally.[9]

Management of Potential Adverse Effects

The multidisciplinary team anticipated three potential hemodynamic challenges: (1) prostaglandin-induced vasodilation causing hypotension, (2) pain-induced sympathetic activation increasing heart rate and cardiac output, and (3) fluid shifts from uterine bleeding. To mitigate these risks, intravenous access was secured before misoprostol administration, fluid resuscitation was pre-planned with a 250 mL crystalloid bolus if systolic BP fell below 90 mmHg, and patient-controlled analgesia was available. No hypotension or significant pain requiring intervention occurred.

Uterine contractions developed within three hours, and complete expulsion of the products of conception occurred after ten hours without hemorrhage, hypotension, arrhythmia, or oxygen desaturation.

Monitoring Protocol and Hemodynamic Data

The patient was monitored in a high-dependency unit from 08:00, when misoprostol was started, with continuous electrocardiography, non-invasive BP measurement every 15 minutes during active uterine contractions and every 30 minutes thereafter, pulse oximetry, and hourly assessment of respiratory rate and symptoms. Transthoracic echocardiography was repeated 6 hours later, at 14:00 (Table 1).

No hypotension (systolic BP <90 mmHg), desaturation (SpO_2 <90%), or arrhythmia occurred during the 12-hour monitoring period.

Post-procedure recovery was uneventful. The patient was monitored for 12 hours and discharged in stable condition. Given her previous non-adherence to oral contraception, long-acting reversible contraception with a subcutaneous etonogestrel implant was initiated before discharge.[1,7]

Table 1. Hemodynamic parameters before and during medical termination.

Parameter	Pre-procedure	At 6 hours
Heart rate (bpm)	95	88
Systolic BP (mmHg)	110	108
Diastolic BP (mmHg)	70	68
Oxygen saturation (%)	95	96
Respiratory rate (/min)	18	16
NT-proBNP (pg/mL)	350	310
sPAP by echo (mmHg)	60	58
TAPSE (mm)	21	21

BP: Blood Pressure; NT-proBNP: N-Terminal pro B-type Natriuretic Peptide; sPAP: Systolic Pulmonary Artery Pressure; TAPSE: Tricuspid Annular Plane Systolic Excursion.

A male condom was also recommended as a second method; in PAH, two methods may be combined because of possible drug interactions with hormonal contraception, and dual contraception is indicated in women taking bosentan.[2,7]

Choice of Etonogestrel Implant Over Intrauterine Device

An Intrauterine Device (IUD) was considered but not selected for two reasons. First, IUD insertion involves cervical instrumentation, which carries a small risk of vasovagal reaction and pain-induced sympathetic activation that could theoretically affect pulmonary vascular resistance. Second, the patient expressed a preference for a subcutaneous implant to avoid any pelvic procedure following the termination. The etonogestrel implant offers comparable contraceptive efficacy with a favorable safety profile in PAH and was well tolerated.

Discussion

Pregnancy in women with IPAH remains associated with unacceptably high maternal morbidity and mortality, despite advances in targeted therapies and contemporary multidisciplinary care. This is a narrative review based on current ESC/ERS guidelines and selected published case series; no systematic search strategy was employed.

Historical series reported maternal mortality rates ranging from 30% to 56%, largely driven by RV failure and thromboembolic complications.[3-4,7] Although outcomes have improved in the modern treatment era, recent observational data continue to demonstrate substantial maternal risk, with reported mortality rates between 11% and 25%.[2,5-6]

While the management approach in this case aligns with ESC/ERS guidelines, several practical lessons merit emphasis. First, detailed hemodynamic monitoring data during medical termination with misoprostol in a clinically stable IPAH patient are rarely reported; our case demonstrates that with appropriate precautions, medical termination can be completed without hemodynamic compromise. Second, the decision to pursue medical over surgical termination was driven by local unavailability of same-day vacuum aspiration, a real-world constraint not addressed in guideline documents. Third, this case illustrates the challenge of contraception adherence in young women with IPAH and the successful transition to long-acting reversible contraception following a pregnancy conceived against medical advice. These practical insights complement existing literature and may assist clinicians facing similar resource or adherence limitations.

Physiological cardiovascular adaptations during pregnancy, including increased plasma volume, reduced systemic vascular resistance, and elevated cardiac output, are poorly tolerated in PAH. These changes impose a critical burden on the right ventricle, which is often unable to augment output sufficiently, predisposing patients to progressive RV failure.[7] Importantly, maternal deterioration may occur abruptly and unpredictably, particularly during the late second trimester and early postpartum period, when hemodynamic stress is maximal.[2,5,7,10]

Risk stratification tools commonly used outside pregnancy do not reliably predict maternal outcomes during gestation, and women classified as low-risk before conception may nonetheless deteriorate during pregnancy, which carries unforeseeable risks and may accelerate disease progression. Consequently, current ESC/ERS guidelines advise that women with PAH be counseled against pregnancy at diagnosis and that early termination be advised for those who become pregnant with poorly controlled disease (an intermediate- or high-risk profile and signs of RV dysfunction); for women with well-controlled disease, a low-risk

profile, and near-normal resting hemodynamics, individual counseling and shared decision-making are recommended.[2]

Addressing the Possibility of Pregnancy Continuation in Stable IPAH

We acknowledge recent case reports and series documenting successful pregnancy outcomes in selected women with PAH, including well-controlled patients with preserved RV function.[5-6,11] Some experts advocate for individualized, case-by-case management rather than absolute contraindication. However, several considerations guided termination in this case. First, despite pre-pregnancy low-risk status, NT-proBNP elevation was already evident at 8 weeks, suggesting early hemodynamic changes that could progress unpredictably. Second, the patient was fully counseled regarding the 11–25% contemporary mortality risk and the possibility of close monitoring with continuation. After understanding both options, she independently elected termination, stating she was unwilling to accept the residual mortality risk. Third, pregnancy in PAH remains classified as contraindicated (modified WHO class IV) in the ESC pregnancy guidelines, and the ESC/ERS guidelines advise against it;[1-2] our practice adheres to these recommendations. We respect that other clinicians may adopt a different approach in similar cases, but the final decision must respect patient autonomy after thorough counseling. This case does not argue against pregnancy continuation in all women with PAH; rather, it illustrates that even "low-risk" patients may prefer termination when fully informed of residual risks.

Safety of Phosphodiesterase-5 Inhibitors during Pregnancy

Unlike endothelin receptor antagonists, PDE-5 inhibitors (sildenafil, tadalafil) are not associated with teratogenicity in human studies. While randomized trial data in pregnant women are limited, observational series and the ESC/ERS 2022 guidelines indicate that PDE-5 inhibitors may be continued during pregnancy when clinically indicated, as the maternal risk of uncontrolled PAH exceeds potential fetal risks.[1-2] In this case, the PDE-5 inhibitor was continued throughout the peritermination period without adverse effects.

Pregnancy Termination and Contraceptive Considerations

Early termination should be discussed when pregnancy occurs in patients with PAH and is advised when the disease is poorly controlled;[1-2] in contemporary series, planned terminations were completed without maternal complications.[5–8] Termination should ideally be performed during the first trimester in a tertiary care setting with expertise in pulmonary hypertension and high-risk obstetrics.[1-2]

Both medical and surgical methods are effective, with similar rates of major complications, but medical termination more often requires unanticipated operative evacuation (2.1% vs. 0.6%); surgical evacuation by vacuum aspiration is therefore

Table 2. Methods of early pregnancy termination in women with idiopathic pulmonary arterial hypertension: maternal risk, anesthesia, and key considerations.

Termination method	Anesthesia	Estimated maternal risk	ESC-aligned considerations
Ultrasound-guided vacuum aspiration	Local anesthesia ± minimal conscious sedation	Low	Preferred approach: similar effectiveness and major complications, but fewer unanticipated operative evacuations (0.6% vs. 2.1%); experienced center with on-site cardiac surgery.[1]
Medical abortion (misoprostol ± mifepristone)	None	Moderate to high	Can be considered up to 9 weeks of gestation, with a reduced misoprostol dose; more unanticipated operative evacuations (2.1% vs. 0.6%); experienced center with on-site cardiac surgery.[1]

ESC: European Society of Cardiology; PAH: Pulmonary Arterial Hypertension. Estimated maternal risk: authors' estimate for women with PAH.

favored in this high-risk group (Table 2).[1] Medical termination can be considered up to 9 weeks of gestation with a reduced misoprostol dose, and high-risk patients should be managed in an experienced center with on-site cardiac surgery.[1] In our patient, continuous cardiopulmonary monitoring was used because prostaglandin-induced vasodilation, pain, and fluid shifts could precipitate hemodynamic instability. General anesthesia should be avoided whenever possible, given its potential to exacerbate pulmonary vascular resistance and impair RV function.[4,7]

Following pregnancy termination, effective contraception is essential and should be addressed as a core component of PAH management. Long-acting reversible contraception, including progestogen-only implants or IUDs, has very low failure rates.[2,7] Estrogen-containing contraceptives are generally avoided because of the increased thromboembolic risk.[1,7] ESC/ERS guidelines emphasize that systematic contraceptive counseling should be provided to all women of childbearing potential with PAH to prevent unintended high-risk pregnancies.[1-2,7]

Preconception Counseling and Post-Event Changes

Before this pregnancy, the patient had received standardized counseling at IPAH diagnosis regarding pregnancy risks and the importance of reliable contraception. However, counseling was provided during a single outpatient visit without structured follow-up or documented patient understanding. After this pregnancy and its termination, our institution revised its counseling protocol for all women of childbearing age with PAH to include: (1) annual structured counseling sessions documented in the medical record, (2) provision of written educational materials, (3) involvement of a clinical psychologist for patients expressing desire for pregnancy despite risks, and (4) a dedicated family planning consultation with an obstetrician at each outpatient visit.

Management of Pregnancy When Continuation Is Chosen

Although early termination is generally advised, rare cases of pregnancy continuation may occur following informed patient refusal of termination. In such situations, management must be centralized in expert pulmonary hypertension centers with a strict multidisciplinary framework.[1-2,7]

Key management principles (adapted from the ESC and ESC/ERS guidelines[1-2]):

- Monitoring: Serial clinical assessment, echocardiography (RV size and function), and biomarker surveillance (NT-proBNP) are required throughout gestation, and supplemental oxygen should be given when indicated.[2,7]
- Medical therapy: Endothelin receptor antagonists are strictly contraindicated (teratogenicity) and must be discontinued immediately. PDE-5 inhibitors and prostacyclin analogs may be continued, as disease control is a key determinant of maternal survival.[1-2,5,8]
- Anticoagulation: Prophylactic anticoagulation with low-molecular-weight heparin may be considered during pregnancy and the postpartum period because of the increased thromboembolic risk.[2,7]
- Delivery planning: Elective delivery in a tertiary unit with locoregional anesthesia (preferred over general anesthesia) is recommended. No clear superiority of cesarean section over vaginal delivery has been demonstrated.[1,5,7]
- Postpartum period: The first weeks after delivery carry the highest maternal risk, requiring continuous monitoring and rapid escalation of PAH therapy.[3–5,7]
- Fetal outcomes: Preterm birth and small-for-gestational-age infants are common; in a meta-analysis, they occurred in 43% and 40% of pregnancies with moderate-to-severe pulmonary hypertension, respectively.[11]

Outcome Data and Patient Selection

Contemporary series suggest that favorable outcomes are more often reported in women with well-controlled disease, preserved RV function, and near-normal hemodynamics under therapy, including vasoreactive patients treated with calcium channel blockers.[2,6] Conversely, maternal deaths and emergency cardiopulmonary transplantation have been reported predominantly in women with severe or poorly controlled PAH.[5-6]

Continuation of pregnancy in PAH should be considered exceptional, undertaken only after exhaustive counseling, and managed exclusively in experienced expert centers.[1-2]

Lessons From This Case (Directly Derived from Patient Course):

- Medical termination with misoprostol can be completed without hemodynamic compromise in a clinically stable IPAH patient under continuous hemodynamic monitoring when surgical termination is not

immediately available.
- Despite pre-pregnancy low-risk classification, NT-proBNP elevation occurred by 8 weeks, illustrating the rapid hemodynamic impact of early gestation.
- Contraception non-adherence is a real-world challenge that requires transition to long-acting reversible methods.

General Recommendations from the Literature (ESC/ERS Guidelines and Published Series):

- Vacuum aspiration remains the preferred termination method, as medical termination more often requires unanticipated operative evacuation.
- Women with PAH should be counseled against pregnancy at diagnosis; those with poorly controlled disease (an intermediate- or high-risk profile with RV dysfunction) who become pregnant should be advised to undergo early termination.
- Highly effective contraception, such as long-acting reversible methods, is essential for all women with PAH of childbearing age.

Conclusion

Pregnancy in IPAH carries persistently high and unpredictable maternal risk; even patients who begin pregnancy with a low-risk profile while receiving contemporary targeted therapy can move to a higher risk category within weeks, at which point early termination should be discussed.[1-2] This case illustrates that favorable baseline functional status does not reliably translate into safe pregnancy outcomes. Early multidisciplinary evaluation, guideline-directed counseling, and timely first-trimester pregnancy termination are critical to optimizing maternal prognosis.[1-2,6–8] Effective long-acting contraception should be systematically implemented as an integral component of care to prevent unintended high-risk pregnancies in women with PAH.[1-2,7]

List of Abbreviations

BP	Blood Pressure
ERS	European Respiratory Society
ESC	European Society of Cardiology
IPAH	Idiopathic Pulmonary Arterial Hypertension
IUD	Intrauterine Device
NT-proBNP	N-Terminal pro B-type Natriuretic Peptide
PAH	Pulmonary Arterial Hypertension
PDE-5	Phosphodiesterase type 5
RV	Right Ventricular
SPAP	Systolic Pulmonary Artery Pressure
TAPSE	Tricuspid Annular Plane Systolic Excursion
WHO	World Health Organization

Ethical Clearance

Written informed consent for publication of this case report was obtained from the patient. Institutional policies on case publication were followed. The patient was informed that her anonymity would be protected. No identifiable patient information is included in this manuscript. This case report did not require institutional review board approval as it describes routine clinical care.

Publication Approval

All authors consent to the publication of this manuscript.

Authors' Contributions

AO: Conceptualization, direct patient care, data collection, and drafting the initial manuscript; KA: Conceptualization, direct patient care, data collection, and drafting the initial manuscript; IA: Data interpretation and critical revision of the manuscript; AB: Supervision and critical revision of the manuscript; JK: Supervision and critical revision of the manuscript for important intellectual content; MABH: Overall supervision, critical revision, and final approval of the version to be published. All authors have read and approved the final manuscript and meet the ICMJE criteria for authorship.

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Conflict of Interest

The authors have no conflict of interest to declare.

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The authors confirm that no generative AI or AI-assisted technologies (such as large language models, or automated writing assistants) were used at any stage of the manuscript preparation. Also , it was not used to generate or alter the scientific content, analyses, or conclusions presented in this manuscript.

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