

## Delayed Termination of Pregnancy in the Setting of Pulmonary Hypertension: A Case Report and Review of the Literature

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### Abstract

**Background:** Nonspecific interstitial pneumonia (NSIP) is a diffuse parenchymal lung disease typically present in young female patients. Pulmonary arterial hypertension (PAH) is a comorbidity of NSIP that can cause many complications during pregnancy due to the physiologic changes of increased estrogen and progesterone, increased metabolic rate, and high-volume states. In pregnant patients with PAH, early termination of pregnancy is recommended to avoid increased maternal risks and mortality.

**Case Report:** Our case involves a 31-year-old female patient with a history of NSIP on home oxygen therapy, diabetes, obesity, and hypertension who was previously evaluated for lung transplantation. She presented to our hospital as a transfer of care at 21.1 weeks with twin IUP from an outside hospital following a 5-week admission. She was initially admitted at 16.1 weeks due to worsening dyspnea on exertion associated with dry cough and orthopnea. She was counseled by pulmonology, cardiology, obstetrics, and maternal-fetal medicine regarding the option for pregnancy termination to improve maternal outcomes, but she declined. The transfer was placed to our institution for the availability of an in-house obstetrics service in the setting of increasing oxygen requirement at greater than 20 weeks. After arrival at our institution, her clinical status began to deteriorate significantly, with her oxygen requirement steadily growing until she was on a high-flow nasal cannula with a flow rate of 50 liters and FiO<sub>2</sub> of 0.9 with persistent tachycardia in the 130s. She was transferred to the ICU (Intensive Care Unit), where termination of pregnancy was strongly recommended as the only measure to save her life. The patient at first continued to decline termination with the goal of delivery after viability. She, however, continued to deteriorate with worsening dyspnea and orthopnea, and she elected for termination of pregnancy by dilation and evacuation (D&E) as a lifesaving measure at 22.0 weeks. This was performed without complication under epidural anesthesia. Her dyspnea and overall clinical condition rapidly improved following her D&E, and she was discharged from the hospital 6 days after her procedure on her baseline home oxygen therapy via nasal cannula. Unfortunately, she was readmitted to an outside hospital 1 month later, where she died secondary to healthcare-associated pneumonia, severe PAH secondary to her pulmonary fibrosis, and acute on chronic respiratory failure.

**Discussion:** In patients with high-risk illnesses such as PAH, preconception counseling should elucidate the high risks associated with pregnancy. Pregnant patients who present with PAH should be counseled on termination of pregnancy as a first-line treatment to decrease the risk of maternal death due to the high stakes of significant cardiac and pulmonary morbidity and mortality associated with PAH and pregnancy. Patients who decide to proceed with their pregnancy should be closely monitored for hypoxia, thrombolytic events, and intravascular volume shifts due to the physiologic changes of pregnancy. Due to the overturning of *Roe v. Wade* in the United States and variable state legislation regarding pregnancy termination, cases such as this illustrate the necessity for honest discussion with patients about the standard of care for pregnancies with critical comorbidities where termination of pregnancy is a lifesaving treatment. Clinicians should be able to have open, truthful, and frank discussions with patients regarding the termination of pregnancy without the fear of legal repercussions.

**Keywords:** *Roe v. Wade*, Nonspecific interstitial pneumonia (NSIP), Pulmonary Arterial Hyperplasia (PAH), Twin Pregnancy, Multidisciplinary, Termination of Pregnancy

### Introduction

Nonspecific interstitial pneumonia (NSIP) is a diffuse parenchymal lung disease that presents clinically in mid-adulthood with dyspnea, cough, and often fever and fatigue [1]. NSIP typically affects female patients of childbearing age, and pregnant women with NSIP are at considerable risk for developing severe pulmonary arterial

hypertension (PAH). PAH is a comorbidity of NSIP that may cause many complications during pregnancy due to the physiologic changes of increased blood volume, estrogen and progesterone, and metabolic rate. Right ventricular failure is a feared complication of PAH, especially in pregnancy, due to its high mortality. Avoidance of

pregnancy or early pregnancy termination is recommended in these patients to avoid the significantly increased risks of maternal morbidity and mortality [2,3]. In this case review, we discuss the incidence of NSIP and PAH in a patient with a twin pregnancy and how the delay in terminating her pregnancy resulted in high maternal risk and likely contributed to her death.

## Case Report

A 31-year-old woman with twin intrauterine pregnancy (TIUP) at 16.1 weeks gestational age (GA) and a history of NSIP, pulmonary fibrosis, diabetes, obesity, and hypertension presented to an outside facility with complaints of dyspnea on exertion, dry cough, and orthopnea for three weeks. She was being evaluated for lung transplantation before her pregnancy was discovered but had been declined due to her weight. She was referred for pretransplant bariatric surgery. Her medications at the presentation time included azathioprine, prednisone, tiotropium bromide, and albuterol. She required 4 L of oxygen at baseline. Termination of pregnancy was discussed with her through pulmonology, cardiology, obstetrics (OB), and maternal-fetal medicine (MFM). She declined termination throughout her stay with the plan to consider it if her condition worsened and with the stated goal of delivering after fetal viability. She was transferred to our tertiary care center at 21.1 weeks GA due to advancing gestation, increasing oxygen requirement, and lack of in-house obstetric services at the outside facility. She was eventually admitted to the ICU for acute on chronic hypoxic respiratory failure secondary to her interstitial lung disease and PAH. Vital signs were notable for tachycardia and low arterial oxygen-to-f<sub>i</sub>O<sub>2</sub> ratios. Her chest x-ray showed significant bilateral perihilar and basal interstitial markings on admission. CT chest showed extensive scarring, interstitial opacities, and no evidence of pulmonary embolism. Her echocardiogram showed an average left ventricular ejection fraction with evidence of moderate to severe pulmonary hypertension, which was unchanged from one month prior. Initial management included atovaquone for pneumocystis jirovecii pneumonia prophylaxis, methylprednisolone, metoprolol, furosemide, benzonatate, and guaifenesin-dextromethorphan.

Due to sustained clinical deterioration and the risk of maternal death, a multidisciplinary team, made up of MFM, OB, critical care medicine, and pulmonology, recommended termination of pregnancy in an interdisciplinary family meeting. The patient at first continued to decline this intervention with the stated goal of delivery after viability. Her condition continued to worsen, and she elected to terminate on day four of admission at 21.6 weeks due to increasing dyspnea, tachycardia, and high flow oxygen requirement. After laminaria placement, on hospital day 5 at 22.0 weeks gestational age, she underwent dilation and evacuation (D&E) for termination of pregnancy and intrauterine device insertion, performed by MFM without complications. After termination, her dyspnea and tachycardia rapidly improved, and she was stable for transfer out of

the ICU approximately 24 hours after her procedure. She was discharged on postoperative day (POD) 6 after returning to baseline respiratory status. She was seen for a telehealth follow-up with pulmonology on POD 22.

On POD 25, the patient presented to an outside hospital with severe dyspnea. She was diagnosed with healthcare-associated pneumonia, further complicated by her baseline tenuous respiratory status and PAH. Despite aggressive management, her condition worsened, and she died on POD 32 of acute or chronic respiratory failure.

## Discussion

Pregnancy can significantly affect cardiopulmonary function. In the upper respiratory tract, estrogen changes can cause mucosal edema, hyperemia, capillary congestion, and fragility of the upper airway with decreased pharyngeal size [4]. In addition, pregnancy reduces systemic vascular resistance due to progesterone and estrogen-mediated vasodilation, leading to potential cardiac decompensation in susceptible patients [2].

Progesterone directly stimulates the medulla to increase respiratory drive while increased metabolic rate and carbon dioxide production increase minute ventilation. Maternal PaCO<sub>2</sub> decreases, and maternal PaO<sub>2</sub> increases, which facilitates the transfer of oxygen from the maternal to fetal circulation. Maternal hemoglobin increases, increasing oxygen-carrying capacity, but overall hemoglobin is reduced by dilutional anemia. Increased cardiac output is required to maintain oxygenation in the setting of higher plasma volume and increased metabolic demands of the fetus and pregnant state. [2,4,5]. The physiologic changes of pregnancy can ultimately lead to right-sided heart failure in susceptible patients through multiple mechanisms. Plasma volume progressively increases through several physiological processes, including systemic vasodilation, activating the renin-angiotensin-aldosterone system, resulting in fluid and sodium retention [2]. Cardiac changes include ventricular dilation and mild tricuspid regurgitation due to increased plasma volume. Increased cardiac output is seen due to the rise in stroke volume and heart rate during pregnancy [2].

These physiologic changes of pregnancy in the setting of NSIP can result in the induction or worsening of pulmonary arterial hypertension (PAH). Contraceptive counseling is recommended for all patients with pregnancy potential, and preconception counseling is crucial in patients with PAH who are considering pregnancy [3]. If patients with NSIP become pregnant, termination should be strongly considered for maternal benefit, significantly when PAH compounds the disease process because of the high maternal mortality [1,3]. Even if the pregnancy results in a third-trimester delivery, maternal mortality is increased postpartum due to acute right heart failure from uterine autotransfusion and volume after delivery [3]. While both medical and surgical termination of pregnancy are safe in low-risk populations, surgical termination is typically preferred by patients [6,7]. In patients with critical conditions, surgical termination of

pregnancy is recommended via D&E since it can be accomplished quickly with local or regional anesthesia and, if needed, minimal anxiolysis or sedation. This avoids the cardiac workload of prolonged uterine contractions with labor induction [8].

This patient's preexisting NSIP led to an increased risk for PAH during pregnancy. The development and worsening of her PAH led to right heart failure, and termination was recommended. The patient first declined termination. She, however, continued to deteriorate with worsening dyspnea and orthopnea, and she agreed to termination via dilation and evacuation. The late decision to terminate her pregnancy led to further respiratory decompensation and increased her risk for complications. After the pregnancy ended, this patient died due to a nosocomial infection complicated by multiple respiratory comorbidities. The toll of pregnancy on her cardiorespiratory system with her comorbidities contributed to her demise.

On June 24, 2022, the United States Supreme Court overruled *Roe v. Wade*, a federal legislation that included the right to decide whether to continue a pregnancy. The overturning of this federal legislation significantly impacts the care of patients with critical comorbidities in pregnancy. The inability to proactively terminate high-risk pregnancies in some states and the fear clinicians have of undergoing legal proceedings that may affect their career and personal freedom significantly complicate the ability of patients to

receive potentially lifesaving care. Additionally, states continue to create legislation defining abortion rights, ranging from complete abortion bans to the preservation of *Roe v. Wade* at the state level. This increases state economies' costs, with the Institute for Women's Policy Research estimating that a lack of access to abortion costs states an average of \$105 billion in 2020 [9]. The risk of medicolegal repercussions to health care providers in states where abortion legislation is vague continues to have unseen and severely detrimental effects on maternal care and mortality.

In patients with critical illnesses exacerbated by pregnancy, such as the patient discussed in this case study, termination is recommended due to the high risk of maternal mortality. Due to the overturning of *Roe v. Wade* in the United States and variable state legislation regarding pregnancy termination, cases such as this illustrate the necessity of more significant research being required to clearly define the standard of care for patients with critical comorbidities in pregnancy when termination of pregnancy provides the lowest risk outcome. Ultimately, we believe patients like ours will have better results if clinicians act in the best interest of their patients without fear of federal legal ramifications. We hope this case report continues to generate discussion on this topic to minimize the adverse outcomes of delayed maternal lifesaving termination of high-risk pregnancies.

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