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Case Report

Anaesthetic Management of Cesarean Section in a Patient with Large Atrial Septal Defect and Severe Pulmonary Hypertension: A Case Report

Shasankan Alagurajah*

Department of Medicine, Galle University, Galle, Sri Lanka

ABSTRACT

Pulmonary Hypertension (PH) in pregnancy is associated with high maternal mortality, particularly when combined with Atrial Septal Defect (ASD). We report a case of a 20-year-old primigravida with a large ostium secundum ASD and severe PH who underwent elective cesarean section at 36 weeks. Despite prior counseling, the patient continued pregnancy and experienced progression from moderate to severe PH with evolving bidirectional shunt. A multidisciplinary team approach was adopted, and graded epidural anesthesia was administered to minimize hemodynamic instability.

Keywords: Pulmonary hypertension; Atrial septal defect; Pregnancy; Cesarean section

INTRODUCTION

Pulmonary Hypertension (PH) in pregnancy carries significant maternal mortality risk of 30-56%. When complicated by Atrial Septal Defect (ASD), the risk increases further due to right ventricular volume overload and potential shunt reversal. Current guidelines recommend against pregnancy in such cases. We present the case of a 20-year-old woman with large ostium secundum ASD and severe pulmonary hypertension who underwent cesarean section [1,2].

CASE PRESENTATION

A 20-year-old primigravida was admitted at 15 weeks of gestation for specialized management of her pregnancy. She had been diagnosed with ASD and mild-to-moderate pulmonary hypertension at age 17 but had declined device closure. Despite medical advice for contraception and later pregnancy termination, she chose to continue the pregnancy [3].

The patient had a large ostium secundum ASD (30 mm) with initially mild-to-moderate pulmonary hypertension (TRPG 45 mmHg), which progressed to severe PH (TRPG>100 mmHg) during pregnancy. She was maintained on sildenafil 12.5 mg TDS and prophylactic enoxaparin 40 mg daily [4].

On examination, she weighed 39 kg with BMI 17.7 kg/m^2. She was mildly dyspneic with respiratory rate 18/min and SpO_2 94-96% on air. Cardiovascular examination revealed regular pulse (84/min), blood pressure 108/76 mmHg, parasternal heave, and ejection systolic murmur with wide fixed splitting of second heart sound. Serial echocardiograms showed progression from left-to-right shunt to balanced shunt with severe pulmonary hypertension [5-7].

Management

A multidisciplinary team including obstetricians, cardiologists, anesthesiologists, and neonatologists planned elective cesarean section at 36 weeks. Preoperative preparation included:

- ICU admission one day prior
- Central venous and arterial line placement
- Lumbar epidural catheter insertion at L4/L5
- Standard fasting protocols
- Antibiotic prophylaxis

RESULTS

Immediate postoperative course

The patient was immediately transferred to the ICU postoperatively for intensive monitoring. Continuous hemodynamic assessment was conducted using invasive arterial and central venous pressure lines to track vital parameters,

Correspondence to: Shasankan Alagurajah, Department of Medicine, Galle University, Galle, Sri Lanka, Tel: 770073271; E-mail: shasankan@gmail.com

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including blood pressure, heart rate, and oxygen saturation. Oxygen supplementation was initiated to maintain SpO_2 levels at or above 90%, with careful titration to avoid exacerbating pulmonary hypertension. Given the risk of fluid overload in the context of severe PH, fluid intake was tightly controlled and restricted, and the patient was closely monitored for signs of both volume depletion and congestion [8].

Epidural analgesia was maintained to manage postoperative pain and minimize the hemodynamic stress typically associated with narcotic analgesia. Deep Vein Thrombosis (DVT) prophylaxis was continued with enoxaparin, alongside regular monitoring for any signs of thrombotic complications, which are common in such high-risk patients. Additionally, the patient was kept on her preexisting medications, sildenafil and enoxaparin, to manage her pulmonary hypertension and prevent thromboembolic events.

Complications

On postoperative day 1, the patient exhibited significant clinical deterioration. Oxygen saturation dropped to 84% on room air, necessitating higher concentrations of supplemental oxygen. The patient's right ventricular dysfunction worsened, as evidenced by rising jugular venous distension and clinical signs of inadequate cardiac output, such as cold extremities and weak peripheral pulses. Cardiovascular imaging and echocardiography confirmed increasing signs of right ventricular strain, which prompted an escalation in pharmacological therapy.

The patient also developed bleeding complications, including a large hematoma, requiring blood transfusions. Despite aggressive fluid management and continued pharmacologic support, her clinical status remained precarious, and she was started on Bosentan (62.5 mg BD) to further reduce pulmonary vascular resistance [9].

On postoperative day 2, the patient experienced sudden hemodynamic instability, with a Mean Arterial Pressure (MAP) of 58 mmHg. This was followed by a critical change in her shunt physiology, with a reversal of flow from right-to-left, which contributed to profound systemic hypoxia and a further decline in cardiovascular function. Despite interventions with milrinone, vasopressin, and phenylephrine to stabilize her hemodynamics, the patient's condition continued to deteriorate.

In response to worsening cardiac function, an emergency re-exploration was planned to address potential causes of the decompensation, including the possibility of surgical site bleeding or tamponade. Unfortunately, during the induction of anesthesia for this procedure, the patient suffered a cardiac arrest. Despite prolonged efforts at resuscitation, including Advanced Cardiac Life Support (ACLS), the patient did not regain spontaneous circulation and ultimately succumbed to right ventricular failure and cardiovascular collapse on the second postoperative day (Figure 1).

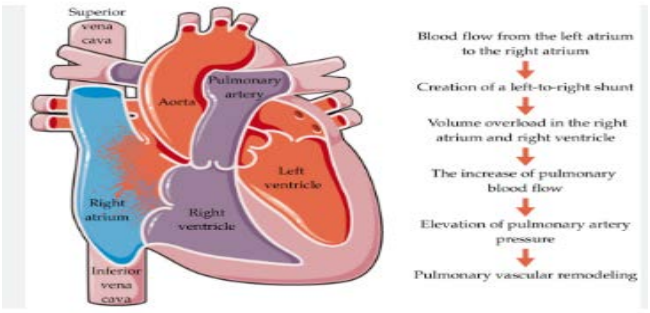


Figure 1: Atrial septal defects: From embryology to pediatric pulmonary hypertension.

DISCUSSION

Pulmonary Hypertension (PH) in pregnancy remains one of the most challenging clinical scenarios in modern obstetrics and cardiology, with reported maternal mortality rates ranging from 30–56%. When associated with Atrial Septal Defect (ASD), particularly large defects such as ostium secundum ASD, the risk is further amplified due to progressive Right Ventricular (RV) volume overload and the potential for shunt reversal. This case exemplifies the catastrophic trajectory that can occur despite adherence to contemporary management strategies.

Physiologically, pregnancy induces a 30-50% increase in blood volume and cardiac output, which significantly exacerbates pre-existing pulmonary vascular disease. In patients with ASD, this increased preload initially augments left-to-right shunting. However, as pulmonary vascular resistance rises especially in the context of worsening Pulmonary Hypertension the shunt may become bidirectional or reverse (right-to-left), leading to systemic hypoxemia and cyanosis. In this patient, the transition from a left-to-right to a balanced shunt, and eventually to right-to-left shunting, reflects the development of Eisenmenger-like physiology, which carries a particularly poor prognosis.

The use of pulmonary vasodilators such as Sildenafil and later Bosentan is consistent with current therapeutic approaches aimed at reducing pulmonary vascular resistance. However, their efficacy in pregnancy is limited, particularly in advanced disease. Additionally, the hypercoagulable state of pregnancy necessitates thromboprophylaxis, as was appropriately administered with enoxaparin. Despite these interventions, the progression to severe PH (TRPG>100 mmHg) indicates irreversible pulmonary vascular remodeling.

The decision to proceed with elective cesarean section at 36 weeks was appropriate, as planned delivery under controlled conditions is generally preferred in such high-risk cases. A multidisciplinary team approach, including intensive care monitoring, invasive hemodynamic assessment, and epidural anesthesia, aligns with best practice recommendations. Epidural anesthesia is often favored over general anesthesia due to its ability to provide gradual hemodynamic changes and avoid abrupt increases in pulmonary vascular resistance. Nevertheless, even with meticulous planning, the peripartum and immediate

postpartum periods remain the most critical, as sudden shifts in preload, afterload, and vascular tone can precipitate acute RV failure.

In this case, the rapid postoperative deterioration highlights several key pathophysiological mechanisms. The onset of hypoxemia and worsening RV dysfunction on postoperative day 1 suggests decompensation triggered by fluid shifts and increased pulmonary vascular resistance. The subsequent development of hemodynamic instability and shunt reversal on day 2 indicates severe RV failure with inability to maintain forward pulmonary blood flow. The requirement for inotropes and vasopressors such as milrinone, vasopressin, and phenylephrine reflects attempts to optimize cardiac output and systemic vascular resistance; however, these measures are often insufficient once decompensation reaches an advanced stage.

The occurrence of cardiac arrest during induction for re-exploration further underscores the fragility of these patients. Induction of anesthesia, even when carefully performed, can lead to sudden reductions in systemic vascular resistance and myocardial depression, both of which are poorly tolerated in severe PH. This highlights the importance of extreme caution during any procedural intervention in such patients.

This case strongly reinforces existing guideline recommendations that pregnancy is contraindicated in women with severe PH, particularly when associated with congenital heart disease such as ASD. Preconception counseling plays a pivotal role in preventing such high-risk pregnancies. Women diagnosed with significant PH should be clearly informed about the substantial maternal mortality risk, and effective contraception should be emphasized. In cases where pregnancy occurs, early termination should be discussed as a life-saving option.

CONCLUSION

This case reinforces that pregnancy in the setting of severe pulmonary hypertension and ASD carries extremely high mortality risk. Despite optimal multidisciplinary care and appropriate anesthetic management, such cases can have fatal outcomes. The importance of pre-pregnancy counseling, early termination options, and strict contraception cannot be overemphasized for women with significant pulmonary hypertension.

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