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Anaesthetic management of a patient with eisenmenger's syndrome for caesarean section

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Abstract
Introduction: Eisenmenger's syndrome is a serious complication of congenital heart defects. It involves high pulmonary artery pressures and reversal of blood flow through a cardiac shunt (right-to-left), leading to significant cyanosis. In pregnancy, these patients are at high risk because normal pregnancy-related changes such as reduced systemic vascular resistance can worsen the right-to-left shunt and hypoxemia. Maternal mortality rates of 30-70% have been reported in such cases.
Case summary: We present the anaesthetic management of a 29-year-old primigravida with a large peri membranous ventricular septal defect (VSD) and Eisenmenger's syndrome at 37 weeks of gestation. She required an elective caesarean section due to severe foetal growth restriction (FGR) and oligohydramnios. After a multidisciplinary discussion, we administered a carefully titrated epidural anaesthetic to achieve a sensory block to T6. Invasive arterial monitoring was used to quickly detect and correct any drops in blood pressure. Postoperative analgesia was maintained with a bilateral transversus abdominis plane (TAP) block and a continuous low-dose epidural infusion of local anaesthetic. The patient's perioperative course was uneventful, and she recovered well.
Conclusion: This case underscores the importance of vigilant haemodynamic control, comprehensive monitoring, and adequate postoperative pain management in obstetric patients with Eisenmenger's syndrome. Suitable thromboprophylaxis and close coordination among obstetricians, cardiologists, and anaesthesiologists can help optimize both maternal and neonatal outcomes.

Keywords: Eisenmenger's syndrome, congenital heart disease, caesarean section, epidural anaesthesia, pregnancy, pulmonary hypertension

Introduction

Eisenmenger's syndrome develops from a structural heart defect (like a large VSD, atrial septal defect, or patent ductus arteriosus) that initially causes left-to-right shunting. Over time, the resulting increased pulmonary blood flow leads to changes in the lung vasculature and severe Pulmonary Arterial Hypertension (PAH). When pulmonary pressures exceed systemic pressures, the shunt reverses to a right-to-left pattern, causing systemic hypoxemia and cyanosis. This progression elevates the risk of complications and death, particularly in pregnant women.

During pregnancy, blood volume and cardiac output increase, while Systemic Vascular Resistance (SVR) decreases. In women with Eisenmenger's syndrome, a lower SVR can either lead to or worsen the right-to-left shunt, leading to further drops in blood oxygen levels. These dynamics not only threaten maternal health but also place the fetus at risk for conditions like fetal growth restriction, prematurity, and hypoxia-related complications. Maternal mortality rates in Eisenmenger's syndrome can range between 30% and 70%, highlighting its severity.

In addition, these patients often have a higher risk of thromboembolism because of secondary polycythemia (an increase in red blood cell mass) due to chronic hypoxemia. Clot formation in the venous system can become paradoxical emboli, passing from the right to the left side of the heart through the shunt. The postpartum period further raises the risk of thrombotic events because of the hypercoagulable state.

Obstetric management in such patients is challenging. While vaginal delivery may be possible in less severe cases, a planned caesarean section is often chosen in patients with advanced disease or when there are other obstetric indications. Careful anaesthetic planning is essential, with goals that include:

1. **Maintaining Systemic Vascular Resistance (SVR):** Low SVR could worsen the right-to-left shunt.

2. **Avoiding increased Pulmonary Vascular Resistance (PVR):** Factors like hypoxia, hypercarbia, acidosis, and certain drugs can elevate PVR.
3. **Ensuring adequate preload:** Both fluid overload and underfilling the right ventricle can be dangerous.
4. **Preventing arrhythmias:** Tachyarrhythmias reduce diastolic filling time and cardiac output.

Regional anaesthesia in Eisenmenger's syndrome is controversial due to concerns about sympathetic blockade causing a sudden drop in blood pressure and further increasing the right-to-left shunt. Nevertheless, small case series have reported safe outcomes with carefully titrated neuraxial techniques. General anaesthesia can also be risky because anaesthetic drugs and intubation can cause abrupt changes in haemodynamics. Therefore, a balanced approach with close monitoring is critical.

Multidisciplinary collaboration among obstetricians, anaesthesiologists, cardiologists, and neonatologists is vital. In the following case, the patient's stable condition, ongoing conservative management of her VSD, and the indication for caesarean section allowed for an epidural technique with incremental dosing, thorough monitoring, and a plan for prompt intervention if haemodynamic instability arose.

Case report

Patient profile and history

A 29-year-old primigravida (G1P0) at 37 weeks of gestation was referred to our centre with severe Fetal Growth Restriction (FGR) and oligohydramnios. She had a known history of a sub-pulmonic VSD since childhood, managed non-surgically. Her prenatal follow-up was irregular, but she remained mostly asymptomatic until late pregnancy.

Over the previous few days, she noted increased fatigue, palpitations, and mild breathlessness. Clinical examination revealed:

- **Blood pressure:** 110/70 mmHg
- **Heart rate:** 120 beats per minute
- **Pulse oximetry on room air:** ~92 to 94%
- Loud P2 on auscultation, with a systolic murmur in the pulmonary area

Investigations

1. **Complete blood count:** Mildly elevated haematocrit (45%), haemoglobin 9.8 g/dL, platelet count 150,000/mm^3.
2. **Serum biochemistry:** Within normal limits.
3. **Arterial blood gas (Room air):** pH 7.40, $PaCO_2$ 36 mmHg, PaO_2 60 mmHg, SaO_2 90%.
4. **Electrocardiogram (ECG):** Right axis deviation and signs of right ventricular hypertrophy.
5. **Transthoracic echocardiogram:** Large perimembranous VSD with bidirectional shunting, severe pulmonary arterial hypertension (estimated systolic pressure >80 mmHg), and preserved left ventricular function (~60% ejection fraction).

A multidisciplinary discussion was held involving obstetrics, cardiology, anaesthesia, and neonatology. Given the situation of severe FGR, oligohydramnios, and high cardiovascular risk of labor, the team decided on an elective caesarean section.

Anaesthetic plan

We chose a controlled epidural anaesthetic with slow, incremental dosing to avoid a sudden drop in SVR. The plan included:

1. **Invasive arterial monitoring:** A radial arterial line for continuous blood pressure measurement.
2. **Epidural catheter insertion:** At the L2-L3 interspace for carefully titrated local anaesthetic.
3. **Local anaesthetics:** Ropivacaine 0.75% in incremental boluses to achieve a sensory block to T6.
4. **Vasopressor support:** Phenylephrine or mephentermine boluses if systolic blood pressure fell below 110 mmHg.
5. **Oxygen supplementation:** To maintain SpO_2 above 92%.
6. **Readiness for general anaesthesia:** If the patient showed signs of instability or if the epidural block was inadequate.

Intraoperative course

After standard monitors were attached (ECG, pulse oximetry), the patient was positioned with a left lateral tilt to reduce aortocaval compression. Following a negative test dose (2% plain lignocaine), incremental boluses of 0.75% ropivacaine were given until a T6 sensory block was reached. This slow administration helped maintain stable haemodynamics.

- **Blood pressure and heart rate:** Systolic blood pressure was maintained between 100-120 mmHg with occasional 50-100 µg phenylephrine boluses. Heart rate stayed between 90-110 beats per minute.
- **Oxygen saturation:** Ranged between 92-94% on 4 L/min nasal oxygen.
- **Estimated blood loss:** ~500 mL, managed with balanced crystalloid solutions and transfusion of one pint of blood.

A baby girl weighing 1.6 kg was delivered with Apgar scores of 7 at 1 minute and 9 at 5 minutes. After clamping the umbilical cord, oxytocin (10 IU) was started at a slow infusion rate to avoid sudden blood pressure fluctuations.

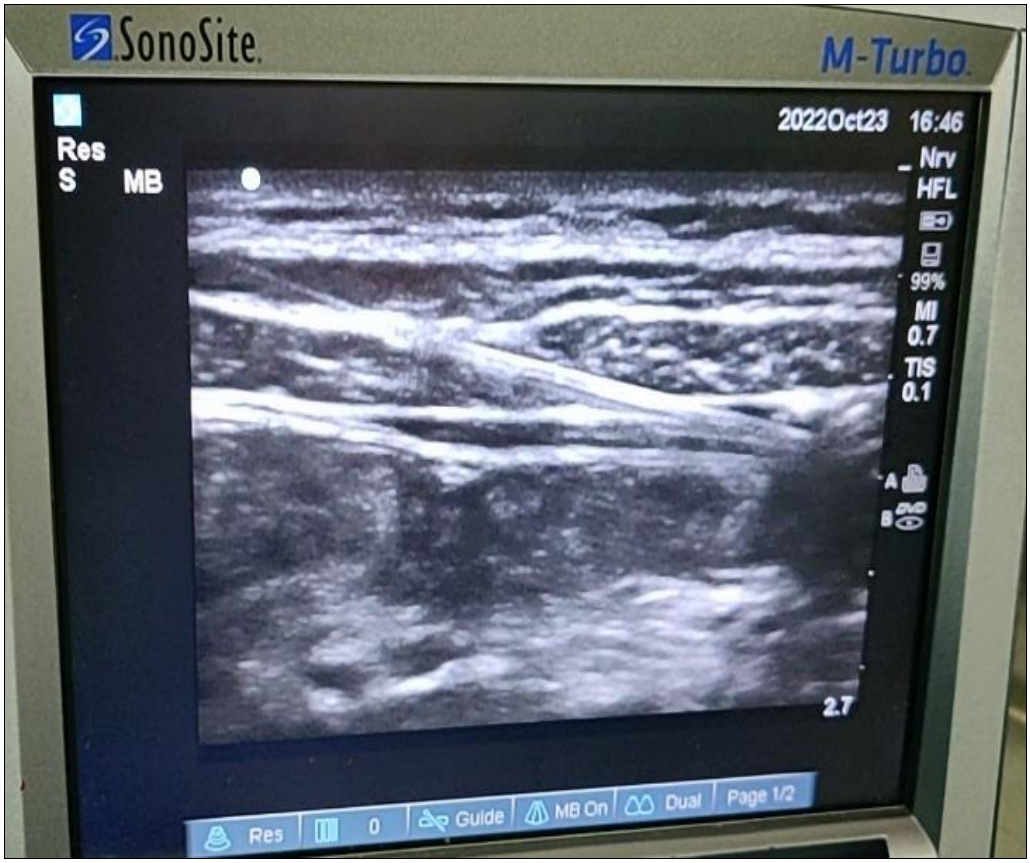
For postoperative pain control, a bilateral transversus abdominis plane (TAP) block was performed under ultrasound guidance with 0.25% ropivacaine (20 mL on each side). A continuous epidural infusion of 0.2% ropivacaine at 3-4 mL/hour provided additional analgesia.

Immediate postoperative management

The patient was monitored in a High-Dependency Unit (HDU) for 48 hours. Thromboprophylaxis with Low-Molecular-Weight Heparin (LMWH) at 40 mg subcutaneously once daily began 12 hours post-surgery. Vital signs, central venous pressure (if indicated), and oxygen saturation were continuously observed. She remained haemodynamically stable, pain scores were low (2-3/10 on a numerical rating scale), and there were no arrhythmias.

Outcome

By postoperative day 3, she was ambulating, vitally stable, and her surgical wound had no complications. She continued to breastfeed without any issues. A repeat echocardiogram showed no significant changes in right ventricular pressures or any new wall motion abnormalities compared to preoperative findings. She was discharged on postoperative day 7 with advice for close cardiology follow-up, extended LMWH prophylaxis for at least 4-6 weeks, and a future assessment for possible surgical or catheter-based VSD repair.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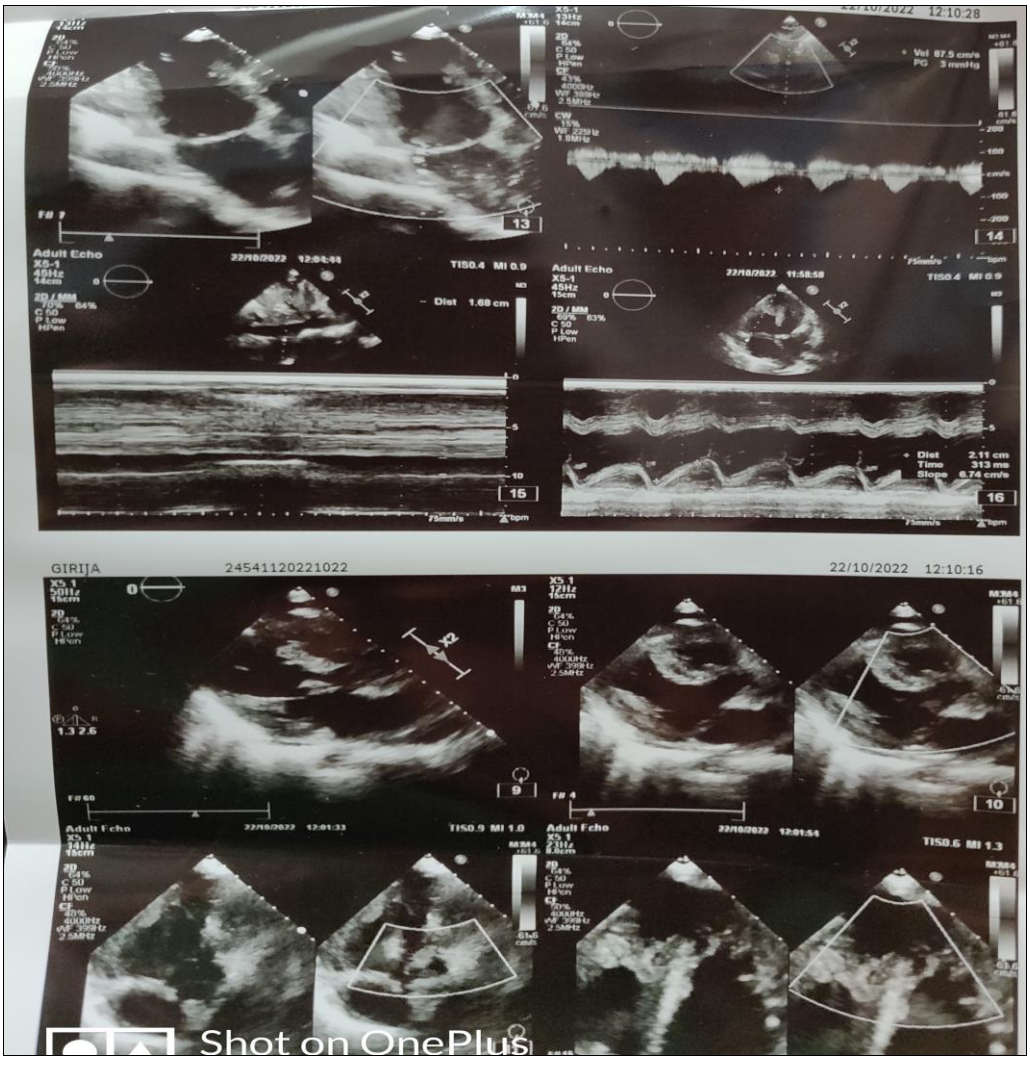
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Discussion

Eisenmenger's syndrome involves severe pulmonary arterial hypertension and reversal of flow through a congenital cardiac defect, creating a right-to-left shunt. During pregnancy, normal changes such as increased blood volume, decreased systemic vascular resistance (SVR), and elevated cardiac output can magnify this shunt and worsen hypoxemia, endangering both mother and fetus. Key anaesthetic goals include maintaining adequate SVR (to limit right-to-left flow), avoiding triggers that raise pulmonary vascular resistance (like hypoxia and hypercarbia), ensuring stable preload, and preventing tachycardia and arrhythmias that can lower cardiac output. Although regional anaesthesia risks a drop in SVR, a carefully titrated epidural can avert sudden hypotension; if general anaesthesia is chosen, induction must be handled gently to minimize cardiovascular instability. Close monitoring with invasive arterial lines allows early detection and correction of small changes in blood pressure. Thromboprophylaxis is critical due to the heightened risk of venous and paradoxical emboli in the setting of chronic hypoxemia, especially in the hypercoagulable postpartum state. Adequate postoperative analgesia using techniques such as transversus abdominis plane (TAP) blocks or low-concentration epidural infusions helps prevent pain-induced sympathetic surges and encourages early mobilization. Most important is a coordinated, multidisciplinary approach involving obstetricians, cardiologists, anaesthesiologists, and neonatologists, which is central to achieving optimal outcomes in these high-risk patients.

Conclusion

Caesarean section in a patient with Eisenmenger's syndrome carries very high risks because of severe pulmonary hypertension, right-to-left shunting, and normal pregnancy-related changes that can worsen the condition. Despite these challenges, a carefully planned approach using slow, incremental epidural anaesthesia, vigilant haemodynamic monitoring, prudent fluid management, and prompt vasopressor support can yield successful outcomes for both mother and baby.

Thromboprophylaxis is vital to prevent potentially fatal thromboembolic events. Adequate postoperative pain control using multimodal techniques (e.g., epidural infusion, TAP blocks) helps prevent large haemodynamic swings. This case highlights the importance of meticulous planning and coordinated teamwork among specialists. With such measures, it is possible to achieve a good maternal recovery and a healthy neonatal outcome, even in a condition as high-risk as Eisenmenger's syndrome.

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