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VAGINAL DELIVERY AND MATERNAL SURVIVAL IN EISENMENGER SYNDROME COMPLICATED BY THIRD-TRIMESTER INTRAUTERINE FETAL DEMISE: A CASE REPORT

Dr Nidhi Jain — Junior Resident, Department of Obstetrics and Gynaecology, Grant Government Medical College and Sir JJ group of Hospitals, Mumbai,

Dr Sandeep Suhas Pophale* — Associate Professor, Department of Obstetrics and Gynaecology, Grant Government Medical College and Sir JJ group of Hospitals, Mumbai*Corresponding Author

ABSTRACT

Background: Eisenmenger syndrome (ES) — pulmonary arterial hypertension with reversed or bidirectional shunting through a congenital cardiac defect — is one of the strongest contraindications to pregnancy, carrying a reported maternal mortality of 20–50% and substantial fetal loss. Ventricular septal defect (VSD), when large and unrepaired, confers disproportionately higher maternal mortality than atrial-level shunts. Reports describing ES presenting with third-trimester intrauterine fetal demise (IUFD) followed by maternal survival remain limited.**Case Presentation:** A 27-year-old gravida 2 para 0 abortion 1 woman with a poorly compliant history of childhood congenital heart disease presented at approximately 29–30 weeks' gestation with reduced fetal movements. Ultrasonography confirmed IUFD in the setting of a known 14 mm perimembranous VSD. She was hypoxaemic (SpO_2 92–96%) but haemodynamically stable, with echocardiography confirming bidirectional shunting, right ventricular hypertrophy, and preserved left ventricular function — consistent with Eisenmenger physiology. A multidisciplinary team planned induction of labour with mechanical cervical ripening, central venous monitoring, and epidural analgesia. She delivered a macerated male stillbirth vaginally. The postpartum course was complicated by transient Type 1 respiratory failure, which resolved with supplemental oxygen, and she was discharged stable on cardiac medications with multidisciplinary follow-up. **Conclusion:** This case demonstrates that maternal survival is achievable in ES even after fetal loss with early recognition, invasive monitoring, avoidance of general anaesthesia, and multidisciplinary care, supporting vaginal delivery under regional analgesia as the preferred route and reinforcing the need for preconception counselling and reliable contraception in women with unrepaired cyanotic congenital heart disease.

KEYWORDS :

INTRODUCTION

Eisenmenger syndrome represents the end stage of long-standing left-to-right shunting through an unrepaired congenital cardiac defect, in which progressive pulmonary vascular remodelling raises pulmonary vascular resistance to or beyond systemic levels, reversing shunt flow and producing chronic cyanosis. It most commonly complicates ventricular septal defect, atrial septal defect, atrioventricular septal defect, or patent ductus arteriosus, and persists in settings with limited access to early paediatric cardiac diagnosis and correction.

Pregnancy imposes haemodynamic demands — increased blood volume, cardiac output, and heart rate, and falling systemic vascular resistance — that are poorly tolerated in ES. Falling systemic resistance worsens right-to-left shunting and hypoxaemia, while the fixed pulmonary vascular bed cannot accommodate augmented flow, predisposing to right heart failure, pulmonary hypertensive crisis, and sudden death. International guidelines classify ES as modified World Health Organization (mWHO) class IV and consider pregnancy contraindicated, recommending termination if conception occurs. Despite this, unplanned pregnancies continue to occur, particularly where congenital heart disease is undiagnosed or medication adherence is poor. We report a case of ES presenting with third-trimester IUFD, managed via a multidisciplinary pregnancy-heart-team model with maternal survival, and discuss it against current evidence and guidelines.

Case Presentation

A 27-year-old woman, married four years, gravida 2 para 0 abortion 1, presented to a tertiary teaching hospital in India as an emergency referral. She had childhood-onset congenital heart disease due to a perimembranous VSD, for which she had been prescribed a fixed-dose combination of sildenafil, a beta-blocker, and an aldosterone-antagonist diuretic for approximately one year, but reported poor compliance during the index pregnancy. She also had untreated hypothyroidism. No formal pre-pregnancy cardiac risk stratification or contraceptive counselling had been documented.

Her first pregnancy had ended in spontaneous abortion two years earlier. The index pregnancy was a spontaneous conception following ovulation induction; a first-trimester anomaly scan had been unremarkable, but she had been non-compliant with cardiac medication and antenatal supplementation throughout.

She presented with two days of reduced fetal movements. Outside ultrasonography had shown a single intrauterine gestation of approximately 29+6 weeks with absent fetal cardiac activity, confirming IUFD. On admission she was conscious and haemodynamically stable, afebrile, and without cyanosis or oedema; pulse was 104–116 bpm, blood pressure 106/70–120/80 mmHg, and SpO_2 92–96% on room air. A pansystolic murmur was audible. The uterus corresponded to 28–30 weeks with no fetal heart activity on hand-held Doppler; the cervical os was closed.

Baseline haematological, renal, and liver investigations were unremarkable, with no evidence of infection or coagulopathy (Table 2). Electrocardiography showed sinus tachycardia with a right-axis, right-ventricular-strain pattern. Obstetric ultrasonography confirmed IUFD and additionally showed moderate maternal hepatosplenomegaly consistent with chronic hypoxaemic congestion. Transthoracic echocardiography demonstrated right ventricular hypertrophy, a large 14 mm perimembranous VSD with bidirectional shunting, mild tricuspid regurgitation, and preserved left ventricular ejection fraction (50%) — an appearance consistent with Eisenmenger physiology. The final diagnosis was a large perimembranous VSD complicated by Eisenmenger syndrome, presenting with third-trimester IUFD in a woman with untreated primary hypothyroidism.

Urgent cardiology, cardiothoracic surgery, and anaesthesia consultations were obtained. Surgery was deferred in favour of continued medical management, with elective catheter study planned as an outpatient. High-risk, poor-prognosis consent was obtained from the patient and family, an intensive/high-dependency bed and blood products were reserved, and four-hourly monitoring with fluid and salt restriction was instituted, with advice to avoid physical exertion.

A triple-lumen central venous catheter was inserted into the right internal jugular vein under ultrasound guidance for continuous central venous pressure monitoring. A lumbar epidural catheter was electively sited in anticipation of labour to provide titratable analgesia while avoiding the haemodynamic swings associated with general anaesthesia. Given the unfavourable, closed cervix, induction was undertaken by mechanical cervical ripening (transcervical Foley balloon catheter with intracervical saline instillation), avoiding pharmacological uterotonic priming agents. Continuous haemodynamic monitoring, supplemental oxygen as required, and epidural analgesia were maintained throughout induction. She progressed to adequate cervical dilatation without decompensation and delivered a macerated male stillbirth (1.8 kg) in vertex presentation, with an intact 500 g placenta. Active management of the third stage used intravenous oxytocin, avoiding ergometrine and prostaglandin analogues; 1 g intravenous tranexamic acid was given as an additional haemostatic measure. There was no postpartum haemorrhage.

Immediately postpartum she remained haemodynamically stable but developed Type 1 (hypoxaemic) respiratory failure, requiring supplemental oxygen (SpO_2 ~95% on 4 L/min) with high-dependency monitoring and serial arterial blood gases; her oxygen requirement was successfully weaned over the following days. Repeat echocardiography on postnatal day 1 confirmed unchanged findings. No surgical intervention was advised, and her pre-existing pulmonary vasodilator, beta-blocker, and diuretic regimen was continued; lactation suppression measures were instituted.

She was discharged approximately 10–11 days after admission, haemodynamically stable, on her cardiac regimen plus iron, multivitamins, and antibiotics, with structured multidisciplinary follow-up arranged in cardiology, cardiothoracic surgery, and obstetrics.

Table 1. Chronological summary of key clinical events

Timepoint	Event	Key findings / action
Day −2	Reduced fetal movements; outside USG	Single IUP ~29+6 weeks, absent fetal cardiac activity (IUFD); known 14 mm perimembranous VSD
Day 0	Admission to tertiary centre	SpO_2 92–96% RA, pansystolic murmur; repeat USG/echo confirm IUFD and Eisenmenger physiology
Day 0	Multidisciplinary referrals	Cardiology, CVTS, anaesthesia consulted; high-risk consent, HDU bed and blood products reserved
Day 0	Invasive monitoring & anaesthetic prep	USG-guided right internal jugular CVC sited; lumbar epidural catheter placed electively
Day 0–1	Induction of labour	Mechanical cervical ripening (Foley balloon + intracervical saline); continuous haemodynamic monitoring
Day 1	Vaginal delivery	Macerated male stillbirth (1.8 kg); placenta 500 g intact; oxytocin + tranexamic acid; no PPH
Day 1	Immediate postpartum	Transient Type 1 respiratory failure; SpO_2 ~95% on 4 L/min O_2; HDU monitoring instituted
Day 1–2	Postnatal cardiac reassessment	Repeat echo unchanged; cardiology/CVTS advise continued medical management, no surgery
Day 2–10	Clinical improvement	Oxygen weaned; lactation suppression instituted; condition stabilised
Day ~10–11	Discharge	Discharged haemodynamically stable on cardiac regimen; multidisciplinary follow-up arranged

Table 2. Summary of key investigations

Investigation	Result	Interpretation
Haemoglobin / WBC / platelets	12.8 g/dL / 10.7 $\times 10^3/\mu L$ / 243 $\times 10^3/\mu L$	Normal; no leukocytosis or thrombocytopenia
Renal & liver function	Creatinine 0.6 mg/dL; SGOT/SGPT 23/18 IU/L	Normal
Electrolytes / viral markers	Na^+ 137, K^+ 4.0 mEq/L; HIV/HBsAg/HCV/VDRL non-reactive	Normal; no coagulopathy or infection documented
Thyroid-stimulating hormone	Elevated (exact value not legible)	Known untreated hypothyroidism
Electrocardiogram	Sinus tachycardia; right-axis, RV-strain pattern	Consistent with chronic pulmonary hypertension
Transthoracic echocardiography	14 mm perimembranous VSD, bidirectional shunt, RVH, mild TR, LVEF 50%	Eisenmenger physiology; unchanged on postnatal day 1
Obstetric ultrasonography	IUP ~29–30 weeks, absent fetal cardiac activity	Confirms IUFD
Maternal abdominal ultrasonography	Moderate hepatomegaly, mild splenomegaly	Chronic hypoxaemia-related congestion
Right heart catheterisation	Not performed during admission	Advised as outpatient study by CVTS

DISCUSSION

Eisenmenger physiology results from chronic pulmonary vascular remodelling that renders pulmonary vascular resistance fixed and eventually suprasystemic, producing bidirectional or reversed shunting, cyanosis, and secondary erythrocytosis. Pregnancy's 30–50% rise in blood volume and cardiac output, combined with falling systemic vascular resistance, increases right-to-left shunting and worsens maternal hypoxaemia and fetal oxygen delivery — the physiological basis for the poor outcomes observed in this population.

Fetal loss in ES is multifactorial, reflecting chronic hypoxaemia-driven placental insufficiency, erythrocytosis-related hyperviscosity impairing uteroplacental perfusion,

and frequent co-occurrence of pre-eclampsia and growth restriction (1,2). In this case, IUFD was the sentinel presentation of previously under-monitored maternal disease, a pattern described elsewhere in cyanotic congenital heart disease (3), underscoring the need to consider maternal cardiac status whenever a woman with known or suspected structural heart disease presents with reduced fetal movements, even if she appears clinically stable.

Guideline positions on delivery mode have shifted. The 2018 ESC guidelines listed ES among lesions where elective caesarean could be considered, but registry data and the 2025 ESC update favour vaginal delivery as the default for most cardiac disease, reserving caesarean for obstetric indications or specific severe cardiac criteria, since caesarean carries greater blood loss, thromboembolic risk, and abrupt haemodynamic shifts without demonstrated mortality benefit (4,5,6). Once IUFD has occurred and fetal compromise from labour is no longer a consideration, this evidence favours vaginal birth even more strongly. Mechanical rather than pharmacological cervical ripening avoided uterotonic haemodynamic effects, and incremental epidural analgesia — rather than general anaesthesia, which carries disproportionate mortality via abrupt falls in systemic vascular resistance and positive-pressure ventilation effects on right ventricular afterload — provided titratable analgesia while blunting the catecholamine response to labour (7,8,9). Central venous pressure monitoring balanced the competing risks of hypovolaemia, which reduces right ventricular preload, and volume overload, which can precipitate right heart failure; pulmonary artery catheterisation was avoided given its excess procedural risk in this population. Oxytocin, rather than ergometrine or prostaglandin F2 analogues, was used for third-stage management to avoid acute pulmonary arterial pressure elevation.

Care was coordinated through a pregnancy heart team spanning obstetrics, cardiology, cardiothoracic surgery, and anaesthesia/critical care, consistent with international cardio-obstetric guideline recommendations for high-risk cardiac pregnancy (4,5). This case is notable for the near-absence of classical poor prognostic markers and for achieving vaginal delivery with maternal survival despite the pregnancy already having reached its most adverse fetal outcome, suggesting that individualised risk stratification — rather than reflexive caesarean delivery — may be appropriate in selected patients with ES.

Because most ES-related deaths occur postpartum, as autotransfusion from uterine involution acutely increases venous return and cardiac workload, high-dependency monitoring was continued well beyond delivery; the patient's transient Type 1 respiratory failure, successfully managed with supplemental oxygen, exemplifies this vulnerable window (10). This case reinforces the need for structured preconception counselling and reliable contraception for women with congenital heart disease, ideally through dedicated transition clinics, and for protocolised multidisciplinary pathways when pregnancy nonetheless occurs.

CONCLUSION

This case demonstrates that maternal survival is achievable in Eisenmenger syndrome complicated by third-trimester intrauterine fetal demise through early multidisciplinary engagement, invasive haemodynamic monitoring, mechanical induction, epidural-based analgesia, vaginal delivery, and vigilant postpartum surveillance. It reinforces the international consensus that Eisenmenger syndrome constitutes a very-high-risk (mWHO class IV) contraindication to pregnancy and highlights the ongoing need for structured preconception counselling, reliable contraception, and transition-of-care pathways for women with congenital heart disease, particularly in resource-limited settings where the underlying lesion may not be optimally characterised before conception. Clinicians should maintain a high index of suspicion for underlying cardiac disease whenever a pregnant woman presents with reduced fetal movements or unexplained hypoxaemia, as timely multidisciplinary escalation remains the single most modifiable determinant of maternal outcome.

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