



Case Report

A case of Eisenmenger syndrome with obstetric complication

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Abstract

Background: Eisenmenger Syndrome is a serious complication of untreated congenital heart disease where long-standing hypertension causes reversal of blood flow, leading to hypoxemia. We report a 27-year-old female with a large atrial septal defect who later developed Eisenmenger syndrome with severe pulmonary hypertension. Her condition became complicated during pregnancy, which resulted in a missed abortion and medical termination. She later presented with cough and vomiting of dark brown material, and investigations showed gastric erosions. She was managed with supportive treatment, antibiotics, and acid-reducing therapy and improved clinically before discharge. This case highlights the importance of early diagnosis of congenital heart disease, careful monitoring in pregnancy, and multidisciplinary care to prevent serious complications.

Keywords: Eisenmenger syndrome, Hypoxemia

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1. Introduction

Eisenmenger Syndrome is a serious, late complication of untreated congenital heart disease characterized by the reversal of an initial left-to-right cardiac shunt into a right-to-left shunt due to the development of severe pulmonary hypertension. It most commonly occurs in atrial septal defect (ASD), ventricular septal defect (VSD), or patent ductus arteriosus (PDA). Over time, the persistent increase in pulmonary blood flow leads to progressive vascular remodeling of the pulmonary arteries, increasing pulmonary vascular resistance. When this resistance exceeds systemic vascular resistance, the direction of blood flow reverses, causing deoxygenated blood to enter systemic circulation, resulting in cyanosis.

Clinically, patients present with progressive dyspnea, fatigue, cyanosis, digital clubbing, and complications such as hemoptysis, arrhythmias, and thromboembolic events. On examination, features may include loud S2 sound, signs of right ventricular hypertrophy, and evidence of chronic hypoxia. Laboratory findings often show secondary polycythemia due to chronic hypoxemia.

Once Eisenmenger physiology develops, the condition is generally irreversible, and closure of the defect is contraindicated.

Management is mainly supportive and includes avoidance of dehydration, prevention of thromboembolism, and treatment with pulmonary vasodilators such as endothelin receptor antagonists, phosphodiesterase-5 inhibitors, and prostacyclin analogues. Pregnancy is strongly contraindicated due to high maternal mortality rates.

2. Case presentation

A 27-year-old female, previously asymptomatic until 2024, presented with breathlessness and episodes of hematoma and was diagnosed with congenital heart disease, specifically a large ostium secundum atrial septal defect (21 mm) with a left-to-right shunt. Echocardiography revealed dilated right atrium and right ventricle with adequate left ventricular function, corresponding to NYHA Class II status, along with moderate tricuspid regurgitation and severe pulmonary hypertension (90/36 mmHg, mean 55 mmHg), and she was a non-responder to nitric oxide.

Over time, her condition progressed, and by February 2026, repeat echocardiography demonstrated reversal of shunt (right-to-left), consistent with Eisenmenger Syndrome. She had no history of PCOS but had a pregnancy complicated by her cardiac condition, resulting in a missed abortion at nine weeks of gestation, for which she underwent medical termination of pregnancy.

During the same admission, she presented with cough with expectoration and vomiting of dark brown contents suggestive of altered blood. Laboratory investigations showed monocytosis (10.3%) with an absolute monocyte count of 1040 cells/mm³, while other parameters including liver function tests, renal function tests, electrolytes, and urine examination were within normal limits.

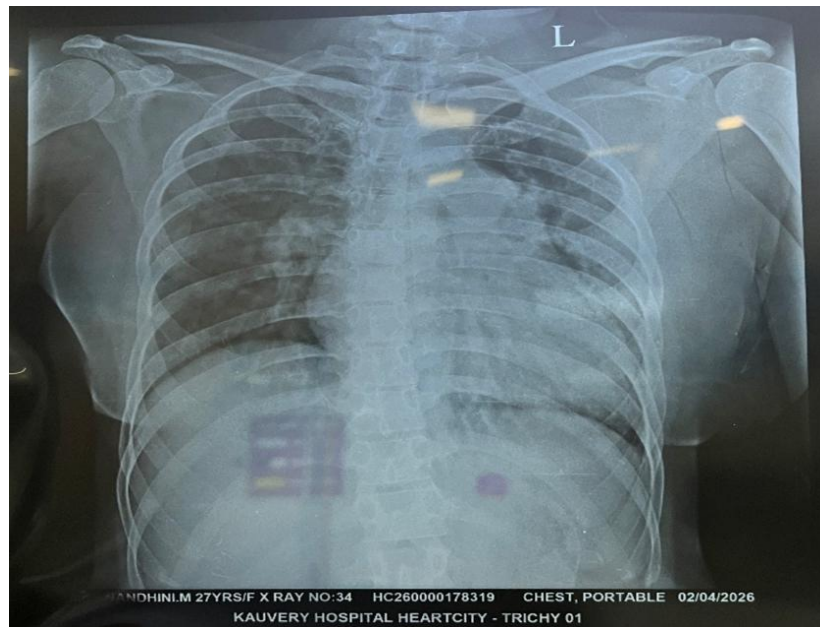


Fig (1): X-ray was normal.

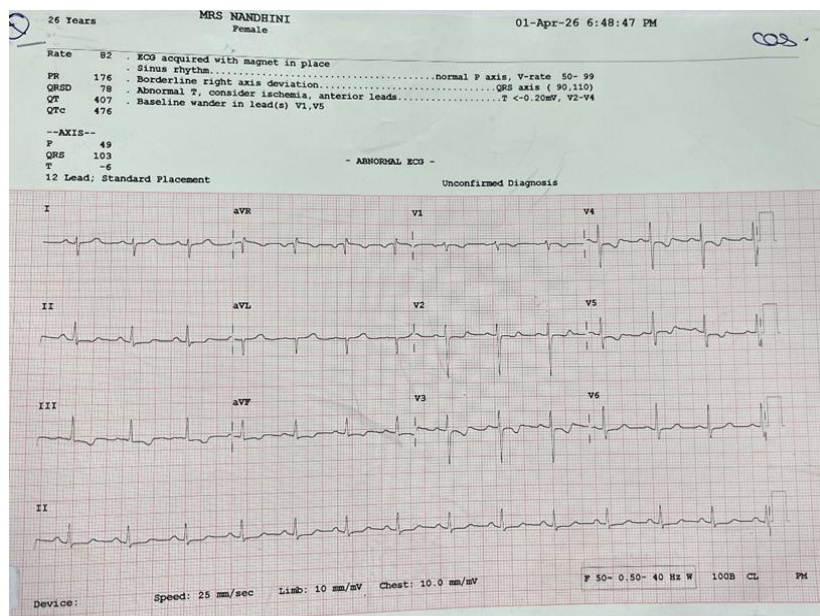


Fig (2): Echocardiography revealed abnormal T waves.

Echocardiography revealed RA, RV dilated, adequate RV and adequate biventricular systolic function, severe PAH, mild TR, large secundum 21mm ASD with Right to Left shunt.

She was managed with intravenous fluids, proton pump inhibitors, and intravenous antibiotics. Ultrasound abdomen suggested a hemorrhagic corpus luteum, and a urine pregnancy test was negative. Upper gastrointestinal endoscopy revealed gastric mucosal erosions, and a rapid urease test was performed. The patient was managed conservatively with supportive care, showed clinical improvement with no further episodes of vomiting, and was discharged in stable condition.

3. Complications

Patients commonly develop progressive cyanosis and digital clubbing due to long-standing right-to-left shunting. Chronic hypoxia leads to secondary polycythemia, which increases blood viscosity and predisposes to both thrombotic events and paradoxically bleeding tendencies due to platelet dysfunction and coagulation abnormalities. Hemoptysis is a serious and potentially life-threatening complication caused by rupture of dilated pulmonary vessels. Cardiac complications include right heart failure, arrhythmias, and sudden cardiac death. There is also an increased risk of infective endocarditis. Neurological complications such as brain abscess can occur due to paradoxical emboli. Renal dysfunction, gout (due to hyperuricemia), and gallstones are other systemic manifestations. In women, pregnancy carries a very high maternal mortality risk (30–50%) and is strongly contraindicated.

4. Surgical / Interventional Options

Once Eisenmenger physiology is established, definitive surgical correction of the original cardiac defect (e.g., ASD/VSD closure) is contraindicated, because the shunt acts as a “pressure relief valve” for the right heart—closing it can precipitate acute right ventricular failure and death.

However, in selected cases, the following options may be considered:

- Heart–lung transplantation: This is the most definitive treatment, especially in advanced disease with severe pulmonary vascular damage.
- Lung transplantation with intracardiac defect repair: Bilateral lung transplantation combined with closure of the defect (e.g., ASD/VSD) is an alternative in suitable candidates.
- Atrial septostomy (palliative procedure): In rare cases, this may be performed to decompress the right heart and improve symptoms, though its role in established Eisenmenger syndrome is limited.

5. Importance of maintaining a registry

It helps doctors and hospitals to:

- Identify patients early before it becomes severe
- Track on how patients are doing over time
- Improve treatment based on real-life outcomes
- Recognize high-risk situations like pregnancy and surgery
- Plan better healthcare services and resources

6. Discussion

In this case, the patient had a large ostium secundum ASD diagnosed in adulthood, which progressed to severe pulmonary hypertension and eventual reversal of shunt from left-to-right to right-to-left circulation, confirming Eisenmenger physiology. Compared to typical cases, this patient had several high-risk features including early development of severe pulmonary hypertension (mean PA pressure 55 mmHg) and non-responsiveness to nitric oxide, indicating advanced pulmonary vascular disease. Similar to reported literature, she presented with progressive dyspnea and later developed systemic complications due to chronic hypoxemia.

A key distinguishing feature in this case is the pregnancy-related complication, which is well documented as extremely high risk in Eisenmenger syndrome. Pregnancy likely worsened hemodynamic instability, contributing to miscarriage and subsequent clinical deterioration. This aligns with existing evidence that maternal and fetal outcomes are poor due to fixed pulmonary hypertension and inability to adapt to increased circulatory demands.

Another notable aspect is the gastrointestinal presentation with vomiting of dark brown content and gastric erosions, which may be related to hypoxia-induced mucosal ischemia, stress-related gastritis, or drug-related effects during hospitalization. Such extra-cardiac manifestations are increasingly recognized in advanced Eisenmenger syndrome due to chronic systemic hypoxemia and multi-organ involvement.

Unlike early diagnosed congenital heart disease cases where surgical correction can be curative, this patient had already progressed to irreversible pulmonary vascular disease, making defect closure contraindicated. Management therefore remained supportive, focusing on symptom control, infection prevention, and stabilization.

In comparison with standard outcomes, this case highlights the late diagnosis, rapid progression, and multi-system involvement seen in resource-limited or delayed referral settings. It emphasizes the importance of early detection of congenital heart disease, timely surgical correction before pulmonary hypertension becomes irreversible, and strict avoidance of pregnancy in established Eisenmenger syndrome.

7. Conclusion

Eisenmenger Syndrome is a preventable but irreversible complication of uncorrected congenital heart disease that significantly worsens prognosis once established. This case highlights the progression of an undiagnosed atrial septal defect to advanced pulmonary hypertension with shunt reversal, complicated further by pregnancy and multi-system involvement. Early detection and timely surgical correction of congenital heart defects are crucial to prevent development of Eisenmenger physiology. Once established, management is mainly supportive, focusing on symptom control, prevention of complications, and multidisciplinary team approach with clinical pharmacists play a vital role in improving safety and outcomes in Eisenmenger syndrome by ensuring rational drug use, preventing medication-related complications, and supporting long-term disease management through patient counseling and continuous monitoring. This case also reinforces the importance of strict pregnancy avoidance and close monitoring in affected patients to reduce maternal and fetal risk.