Medical Science

pISSN 2321–7359; eISSN 2321–7367

Fruitful case of Eisenmenger syndrome with ventricular septal defect in a case of pregnancy: A case report

Kamlesh Chaudhari[1], **Sanket Bakshi**[2], **Tanvi Chaurasia**[3]

To Cite:
Chaudhari K, Bakshi S, Chaurasia T. Fruitful case of Eisenmenger syndrome with ventricular septal defect in a case of pregnancy: A case report. *Medical Science* 2023; 27: e119ms2858.
doi: https://doi.org/10.54905/disssi/v27i132/e119ms2858

Authors' Affiliation:
[1]Professor, Department of Obstetrics and Gynaecology, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher education and research (Deemed to be University), Wardha, Maharashtra, India
Email: dr.kamleshchaudhari@gmail.com
[2]Intern, Datta Meghe Institute of Medical Sciences, Wardha, Maharashtra, India
Email: sanketbakshi25#gmail.com
[3]Junior Resident, Department of Obstetrics and Gynecology, Datta Meghe Institute of Medical Sciences, Wardha, Maharashtra, India
Email: tanvichaurasia001@gmail.com

Peer-Review History
Received: 20 January 2023
Reviewed & Revised: 24/January/2023 to 23/February/2023
Accepted: 24 February 2023
Published: 27 February 2023

Peer-review Method
External peer-review was done through double-blind method.

URL: https://www.discoveryjournals.org/medicalscience

ABSTRACT

Eisenmenger syndrome (ES) is an absolute contraindication to pregnancy. ES is distinguished by a congenital heart defect that causes a significant anatomical shunt. Hemodynamic forces cause a left-right shunt, resulting in severe pulmonary arterial hypertension (PAH). Because of increased pulmonary vascular resistance, the shunt will eventually become a right-to-left shunt, resulting in significant hypoxemia and cyanosis. The significant maternal morbidity in individuals with Eisenmenger's syndrome, including heart failure, dyspnoea, syncope and abrupt death, is a result of the circulatory alterations that take place during pregnancy. As a result, ES is considered to be completely contraindicated during pregnancy. Appropriate ES management entails assembling a multidisciplinary team to carefully and thoroughly monitor and manage the patient, Oxygen therapy, vigorous pulmonary vasodilator therapy, and care from a specialised multidisciplinary team may reduce mortality if a woman with ES decides to carry the pregnancy to term. We present a case of Eisenmenger syndrome with ventriclular septal defects with severe PAH in a 27-year-old woman who had a caesarean section at 35 weeks of pregnancy. She gave birth to a healthy baby girl. She was discharged on the seventh postoperative day with no complications.

Keywords: Pregnancy, Eisenmenger's syndrome, ventricular septal defect

1. INTRODUCTION

Eisenmenger's syndrome (ES) is the development of pulmonary arterial hypertension (Arvanitaki et al., 2020; Bhedodkar et al., 2022) with an atrioventricular or aortopulmonary shunt reversed, such as ventricular septal defects, atrial septal defects, patent ductus arteriosus, transposition of the great vessels or surgically created aortopulmonary connections. The size and location of the intracardiac defect affect the severity of ES development. Right ventricular (RV) failure, right ventricular hypertrophy, heart failure, dyspnoea, cyanosis, clubbing and arrhythmias are common consequences of ES. With a congenital cardiac defect incidence of only approximately 3% during pregnancy, ES is extremely uncommon. High maternal death rates of between 30 and 50 percent are seen in pregnancies with ES because of the

increased circulatory burden that occurs throughout pregnancy. Right ventricular failure, pulmonary hypertension crisis, arrhythmia and stroke are the main causes of death in ES. Uncertainty exists over the treatment of Eisenmenger syndrome during pregnancy and its effects on the mother and foetus. Around 30–50% of maternal deaths are caused by Eisenmenger syndrome and 65% are caused by Caesarean sections (Lopez et al., 2020). Due to its ability to lessen perioperative discomfort as well as pulmonary and systemic vascular resistances, epidural analgesia is favoured to caesarean delivery. The three leading causes of death are preeclampsia, thromboembolism and hypovolemia. Pregnancy in Eisenmenger syndrome is best avoided because it has a high rate of maternal mortality and a poor prognosis for the unborn child. Women who have a strong desire to become pregnant are more likely to seek a quick labour and a pain-free birth under an epidural block.

2. CASE DISCUSSION

A 27-year-old primigravida with 35 weeks gestational age (Figure 1) came to casualty with complains of dyspnoea and palpitations. Patient had no regular follow ups; she came to us in first trimester when medical termination of pregnancy was advised for which she denied and then she lost follow up and came at 35 weeks of gestation in casualty. There was no prior medical history of syncope, chest discomfort, giddiness or haemoptysis.

She appeared ill and pale on general examination (Table 1). She had dry mucus membranes and mild oral mucosa pallor. Her vital signs revealed that she had low blood pressure. The patient's oxygen saturation was low. She also showed clubbing, cyanosis and a low volume pulse but no symptoms of heart failure. A holosystolic murmur was detected by CVS examination along with a loud pulmonic component of the second heart sound on auscultation in the left lower sternal border. On respiratory examination, all lung fields had bilateral vesicular breath sounds.

On per abdomen examination uterus was 36 weeks size relaxed, cephalic foetal heart sound 146 beats per minute. On per vaginum examination os was closed uneffaced. A single viable foetus in cephalic presentation was revealed by abdominal ultrasound. The foetal weight was 2456g. The amniotic index was normal. The placenta was posterior and elevated. Doppler ultrasound revealed a normal umbilical artery Pulsatility index (Figure 2).

Laboratory tests revealed normal haemoglobin levels. The patient's haematocrit was of the normal range, his complete blood count was normal (CBC) (Table 2). Cardiomegaly with large pulmonary conus was seen on the Chest X-ray (Figure 3). Right axis deviation, right atrial and right ventricular hypertrophy, along with RV strain, were all visible on the ECG (Figure 4). VSD was seen on a 2D echo with eisenmengerization (Figure 5). She was identified as having 20 mm of mid-muscular non-restrictive VSD and severe PAH consistent with Eisenmenger syndrome.

Patient was planned for caesarean section. Under general anaesthesia patient was induced, a healthy baby 2.5 kg delivered. Intraoperatively haemostasis achieved. Patient withstood the procedure well (Figure 6). The patient and baby were vitally stable following the caesarean section and were immediately transferred to the cardiac surgery intensive care unit (CSICU) with no complications for monitoring. Restricted Fluids were administered to the patient to maintain pulmonary artery hypertension and diuretics were avoided to avoid a decline in PAH and the collapse of the inferior vena cava.

On postoperative day 1, she was using high-flow nasal oxygen and had an oxygen saturation of 92%, as well as pulmonary vasodilators. As a precaution, she was also taking anticoagulants, higher antibiotics were also given. Her uterus was well retracted. Her lochia was typical. Post operatively patient was vitally stable. She was discharged on postoperative day 7 with no further complications and was asked to follow up weekly (Figure 7). On discharge patient was given Tab bosentan, an endoethelial receptor antagonist for 7 days.

Table 1 General examination of the patient

Vital signs	Patient's result
Temperature	36.8 C
Heart rate	90/Min
Respiratory rate	22/Min
Blood pressure	100/60 Mm Hg
Oxygen saturation	92% At Room Air
Body mass index	25.3 Kg/M^2

Table 2 Laboratory test of the patient

Laboratory test	Patient results
Haemoglobin	12 GM%
Haematocrit	38%
Sodium	136 mmol/L
Potassium	4.5 mmol/L
Chloride	100 mmol/L
Urea	3.6 mmol/L
Creatnine	0.6
Estimated GFR	More than 60

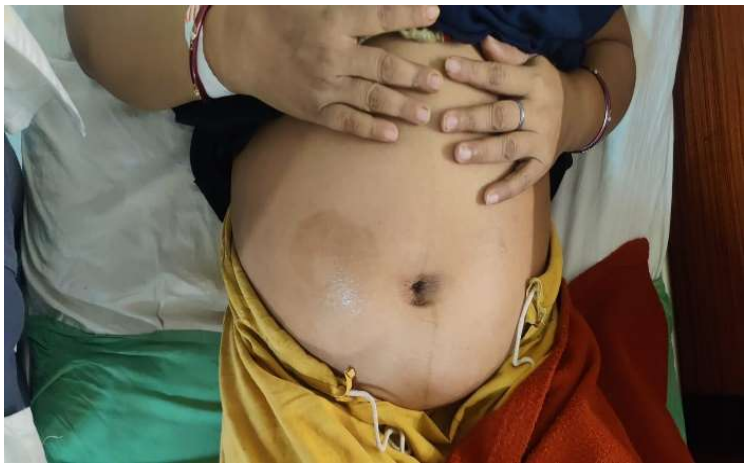

Figure 1 Primigravida with 35 weeks of gestational age

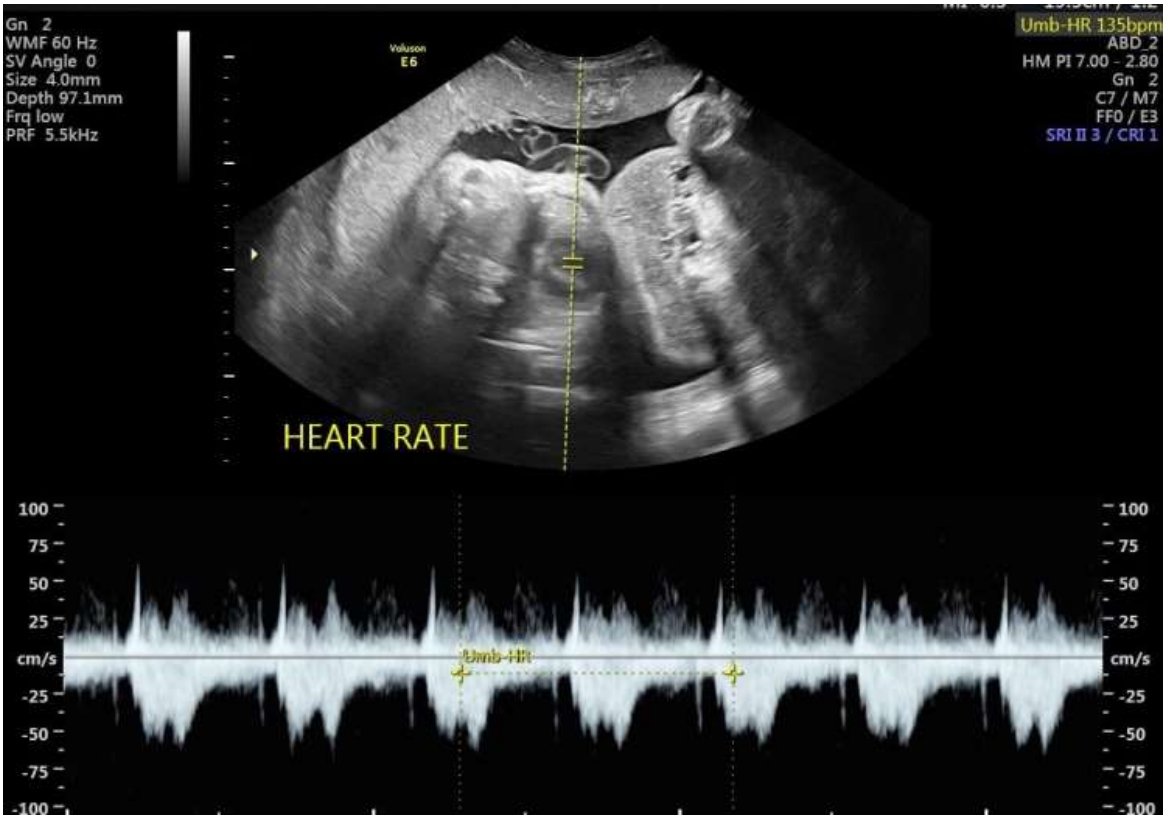

Figure 2 Umbilical artery Doppler ultrasound (normal)

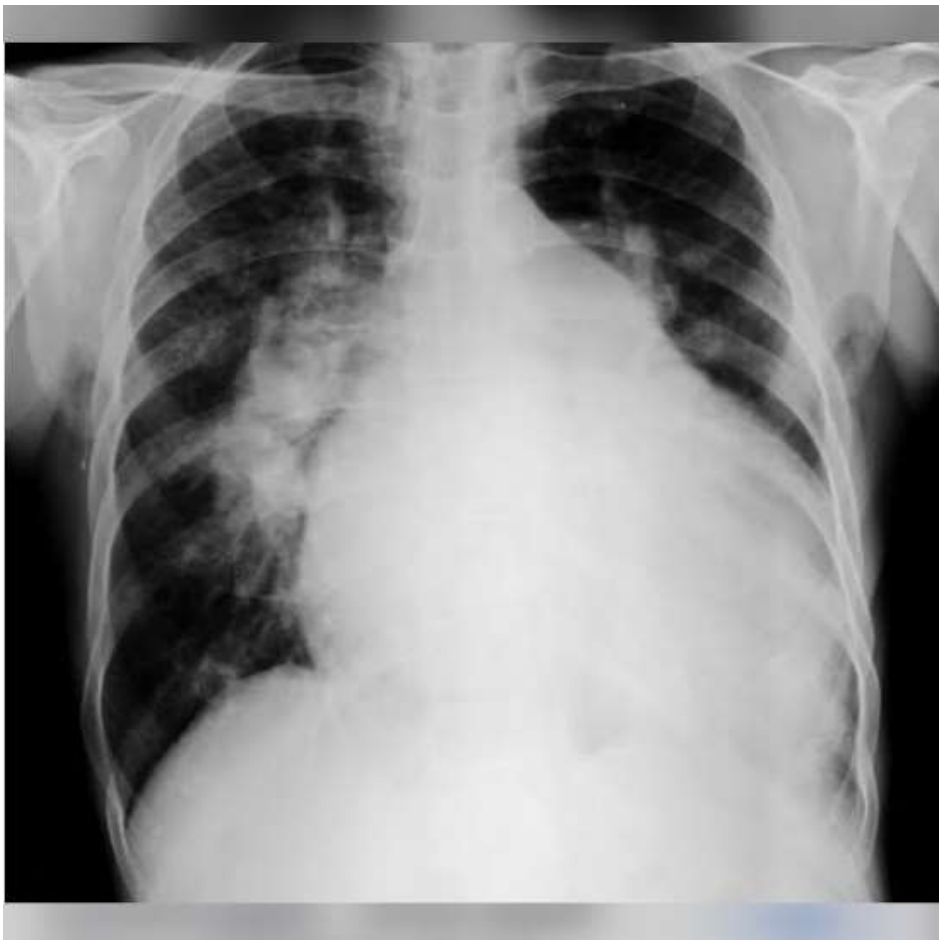

Figure 3 Cardiomegaly with large pulmonary conus on Chest X-ray

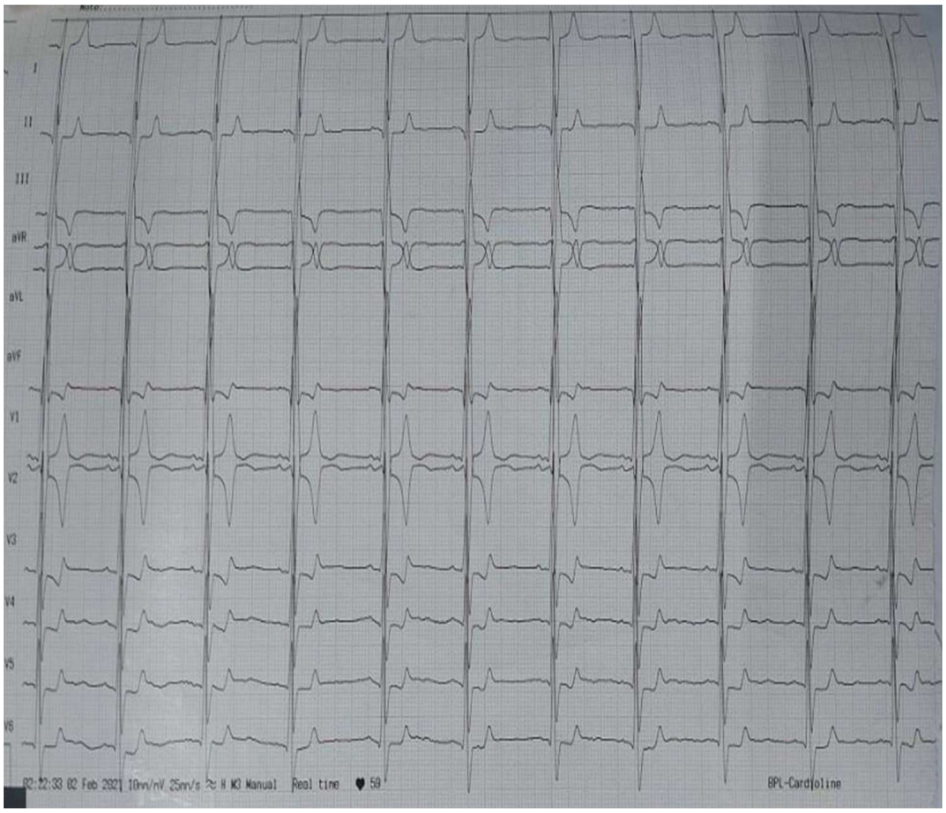

Figure 4 Right axis deviation, right atrial and right ventricular hypertrophy, along with RV strain on ECG

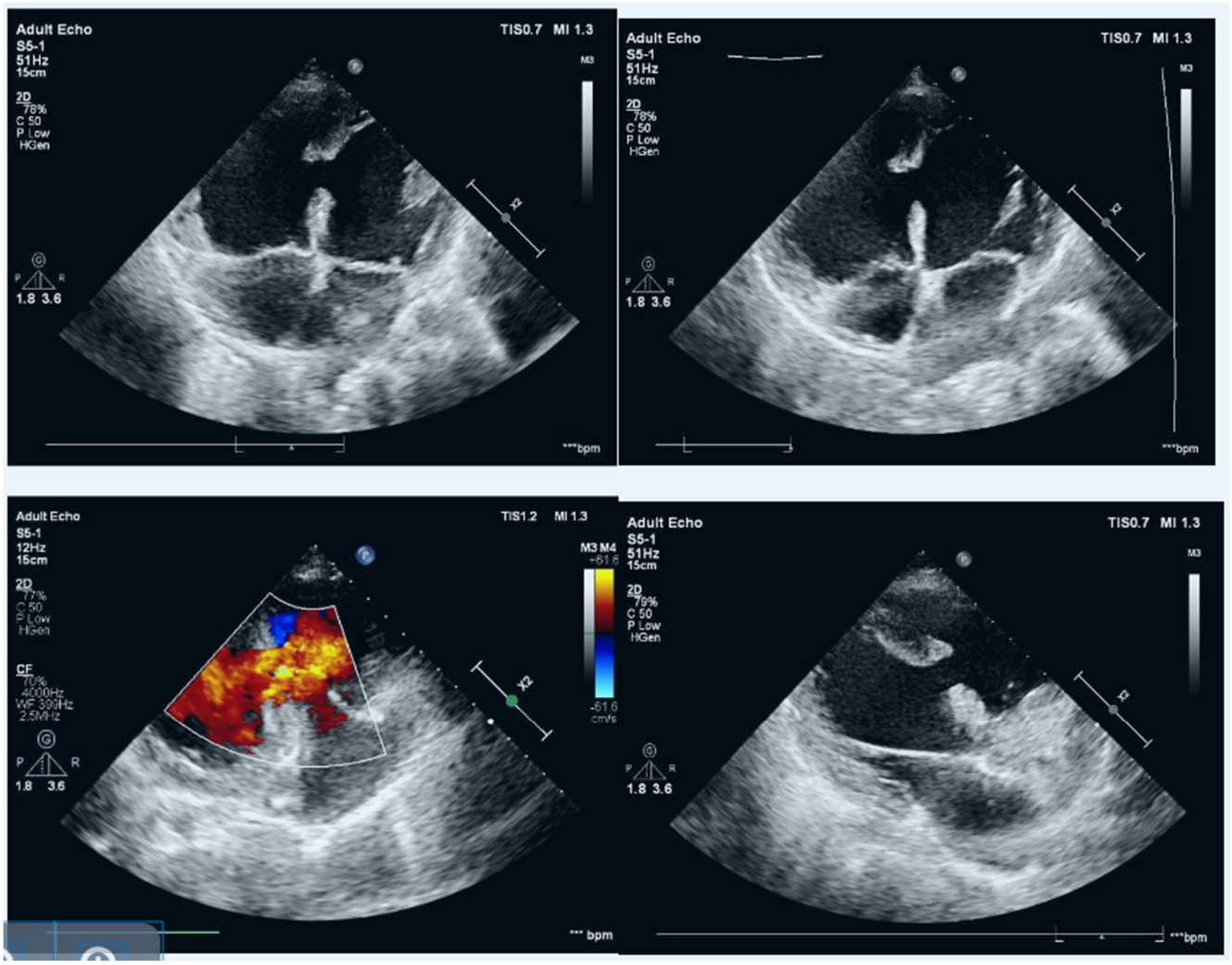

Figure 5 VSD was seen on a 2D echo with eisenmengerization

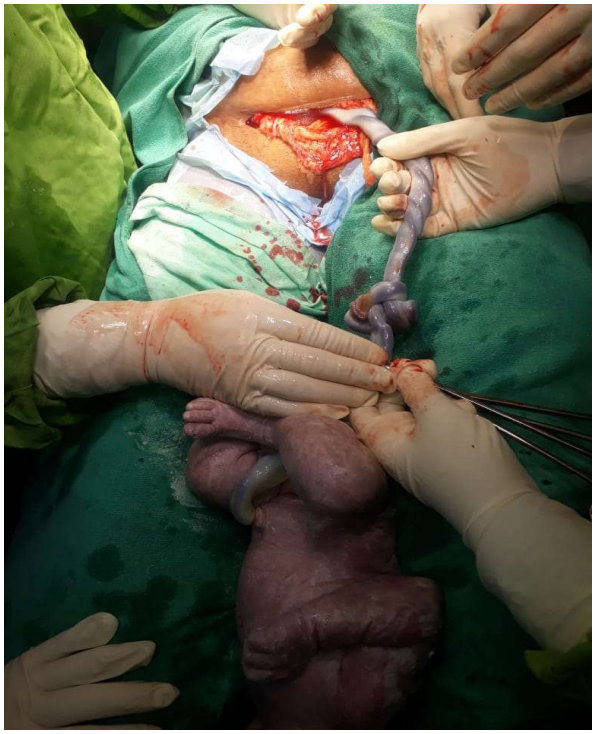

Figure 6 Patient underwent caesarean section under general anesthesia

Figure 7 Mother with healthy baby discharged on POD-7

3. DISCUSSION

Eisenmenger syndrome is a disorder that develops as a result of congenital cardiac abnormalities such as VSD, PDA and ASD (Burkett, 2020). Chronic left to right shunts increases pulmonary artery blood flow, which causes pulmonary arterial hypertension to gradually become hyperkinetic and shunt reversal to culminate in cyanosis and clubbing (Horstkotte et al., 2003). Patients with Eisenmenger syndrome are more susceptible to hemodynamic alterations, such as a decrease in systemic vascular resistance (SVR) and an increase in right-to-left shunting, which may lead to circulatory collapse brought on by anaesthesia or surgery. Excessive bleeding, postoperative arrhythmia, deep vein thrombosis and paradoxical emboli are additional risks related to surgery. According to a study, maternal mortality in pregnant Eisenmenger syndrome patients was 40% and foetal mortality was 8%. Seventy percent of deaths typically occur on postpartum days 2-30 or at the time of birth, according to another study, which found 34% mortality related with vaginal delivery and 75% mortality associated with Caesarean section. During pregnancy, the normal blood volume can rise by up to 50% and when the uterus contracts during labour, the blood volume continues to rise. The hypoxia worsens in Eisenmenger syndrome patients when the right to left shunt increases and the systemic resistance decreases. People with Eisenmenger syndrome should avoid getting pregnant since it is a pro-thrombotic state that can induce pulmonary embolism and sudden cardiac death. These patients appeared to have significant rates of maternal and foetal mortality, according to several investigations. Currently, stronger vasodilators include endothelial receptor blockers such bosentan, ambrisentan and macitentan (Pesto et al., 2016). The patient must cease using these medications before becoming pregnant because they are all teratogenic (category X) drugs. A patient should receive oxygen therapy and diuretics if they show indicators of heart failure. The ideal birthing mode is vaginal and the appropriate pain management is provided by intravenous analgesics or low-dose epidural anaesthesia.

4. CONCLUSION

It is essential for these patients' pregnancy and delivery if the underlying heart defect was identified and treated in early childhood. Pregnancy in ES is still linked to high rates of maternal and foetal morbidity and mortality, especially when compounded by severe PAH. Pregnancy termination in the first trimester is advised and effective preconception counselling is a must. When pregnancy continues into the third trimester, risks considerably rise. The ES patient with PAH should be closely watched and managed in a

tertiary facility with the help of obstetricians, cardiologists, anaesthesiologists, paediatricians and intensivists. There is no set method for managing ES throughout pregnancy and the individualization of care for every patient appears to be crucial for excellent perinatal results.

Acknowledgement
We thank all the participants who have contributed in this study.

Informed Consent
Informed Consent was obtained from the patient.

Author's contribution
All the authors contributed equally to the case report.

Funding
This study has not received any external funding.

Conflict of interest
The authors declare that there is no conflict of interests.

Data and materials availability
All data sets collected during this study are available upon reasonable request from the corresponding author.

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