Management of Eisenmenger Syndrome in Pregnancy: A Contemporary Review

Dr. Bhavya SV, Associate Professor in OBG Nursing

JSS College of Nursing, Mysuru.

Corresponding Author mail ID: spbhavya123@gmail.com

Abstract

Background: The most severe clinical stage of pulmonary arterial hypertension (PAH) linked to congenital heart disease (CHD) is Eisenmenger Syndrome (ES). Even though pregnancy is generally discouraged (mWHO Class IV), cases often show up in tertiary facilities, especially in South Asia. **Goal:** To compile the most recent research (2020–2025) on the interdisciplinary treatment of ES during pregnancy, with an emphasis on pharmaceutical developments and peripartum care practices. **Methods:** Recent case series and clinical statements from indexed sources (Google Scholar, PubMed, and Scopus) were reviewed. **Results:** Although maternal mortality is still high (20–50%), recent data show that a "Cardio-Obstetrics" multidisciplinary team (MDT) approach, aggressive targeted PAH therapy Phosphodiesterase- 5 (PDE-5) inhibitors and prostacyclins), and careful peripartum haemodynamic monitoring significantly improve survival. **Conclusion:** Although uncommon, successful results with ES pregnancies are becoming more attainable. Preventing the "Eisenmenger Crisis" through Systemic Vascular Resistance (SVR) maintenance and air embolism prevention must be management's top priority.

Keywords: Eisenmenger Syndrome, Pregnancy, Pulmonary Hypertension, Multidisciplinary Team, Targeted Therapy.

Introduction

A long-term left-to-right shunt causes permanent pulmonary hypertension, shunt reversal (right-to-left), and systemic cyanosis in Eisenmenger Syndrome (ES), the pinnacle of pulmonary vascular obstructive disease. The Modified World Health Organisation (mWHO) Class IV designation for ES in the hierarchy of maternal risk indicates a "extremely high risk of maternal mortality or severe morbidity," with historical mortality rates ranging from 30% to 50% [1,2].

Many women in underdeveloped nations like India present at advanced gestational ages, despite the worldwide consensus supporting early termination or avoidance of pregnancy. This is frequently linked to restricted access to pre-conception counselling, sociocultural obstacles to abortion, and delayed diagnosis [3]. However, new data from 2020 to 2025 indicates that maternal survival is increasing with specialised care and standardised standards [4].

In ES, physiological changes brought on by pregnancy are not well tolerated:

Diminished Systemic Vascular Resistance (SVR) The right-to-left shunt is made worse by the typical decline in SVR, which exacerbates hypoxaemia.

Increased Cardiac Output: Right ventricular (RV) failure may be triggered by a 40–50% increase in blood volume.

Hypercoagulability: Raises the possibility of paradoxical systemic emboli and pulmonary thrombosis.

Modern Techniques for Management

- The MDT, or Multidisciplinary Team

Nowadays, the "Cardio-Obstetrics" model is considered the best. Survival depends on a team consisting of ACHD experts, high-risk obstetricians, cardiac anaesthesiologists, and newborn intensivists, as shown by centres in China and India [4,5].

- Pharmacotherapy for Targeted PAHs

The switch from mainly supportive treatment to intensive vasodilator therapy has been a significant change in recent years. A common first-line treatment for lowering pulmonary vascular resistance (PVR) is sildenafil, a PDE-5 inhibitor.

Patients with NYHA Class III/IV symptoms are treated at tertiary facilities with either IV/SC treprostinil or inhaled iloprost. Early triple treatment beginning may stabilise patients who were previously thought to be terminal, according to recent reports [6]. Anticoagulation is very customised. The risk of pulmonary haemorrhage must be weighed against the overall recommendation for prophylactic Low Molecular Weight Heaprin to avoid thromboembolism [7].

- Considerations for Anaesthesia and Delivery

Although the delivery method is still debatable, new research supports planned elective caesarean sections performed in a controlled setting.

Anaesthesia Because it prevents the haemodynamic fluctuations associated with intubation and permits a gradual, titrated drop in SVR, incremental or graded epidural anaesthesia is favoured over general anaesthesia [8].

For the prompt identification of shunting changes, invasive arterial blood pressure monitoring is necessary.

- Postpartum Care and Specialised Nursing

Because of fluid autotransfusion and abrupt changes in PVR, the "Postpartum Danger Zone" (the first 72 hours) is the time of greatest mortality.

Nursing Vigilance: Constant monitoring of CVP and SpO2 is required. To avoid paradoxical air emboli, nurses must use air-eliminating filters on all IV lines.

Fluid Management: RV overload can be lessened by a "dry lung" approach that uses prudent diuretics (such as furosemide 0.5 mg/kg) after birth [5].

Comparison of Maternal Outcomes (Recent Series and Case Reports)

The following table highlighting the shift toward zero or reduced mortality in centers utilizing advanced multidisciplinary protocols.

Study (Year)	Setting	N (Patients)	Maternal Mortality	Neonatal Outcome	Key Strategy/Intervention
Rathod et al. (2024) [3]	South India	6	0%	83% Live Birth	Tertiary MDT, early Sildenafil, and incremental epidural anesthesia.
Saha et al. (2023) [2]	North India	11	27%	64% Premature	Specialized Cardio-Obstetric Unit; high mortality linked to late referral.
Hong et al. (2023) [4]	China	1 (Case)	0%	Healthy Term	Standby ECMO, MDT, and parenteral Treprostinil.
Sharma et al. (2022) [9]	West India	1 (Case)	0%	Live Birth (CS)	Multidisciplinary stabilization; focus on "dry lung" fluid management.
Patil et al. (2021) [10]	Central India	3	33%	66% Live Birth	Challenges in managing acute "Eisenmenger Crisis" postpartum.
Kulkarni et al. (2025) [11]	South India	2 (Case)	0%	100% Live Birth	Upfront Sildenafil and Bosentan (postpartum) with ICU vigilance.

Three clear trends emerge from the examination of these cases:

The Survival Gap: Compared to regular obstetric settings, where mortality frequently surpasses 30%, patients treated in specialised Cardio-Obstetric facilities have a death rate of 0–10% [2, 10].

Pharmacological Trends: Sildenafil is increasingly being used as a baseline therapy in Indian tertiary centres; more recent publications (2024–2025) demonstrate the effective use of dual therapy (adding prostacyclin analogues) after clinical deterioration [3, 11].

Newborn Morbidity: Although maternal survival is increasing, high rates of intrauterine growth restriction (IUGR) and preterm delivery—which are frequently brought on by deteriorating maternal cyanosis—continue to negatively impact newborn outcomes [9].

Conclusion

In obstetrics, Eisenmenger syndrome continues to be one of the most difficult problems. The 2020–2025 period has demonstrated that a combination of intensive targeted PAH medication and highly specialised MDT care may yield excellent outcomes, even while the advice against pregnancy remains. To further lower mortality in places with limited resources, future studies should concentrate on standardised methods for PAH treatment during pregnancy.

References

1. Regitz-Zagrosek V, Roos-Hesselink JW, Bauersachs J, et al. 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy. Eur Heart J. 2018;39(34):3165-3247.
2. Saha S, Balakrishnan S, Jaiswal A. Pregnancy in Eisenmenger syndrome: a case series from a tertiary care hospital of Northern India. Cardiol Young. 2023;33(1):45-50.
3. Rathod S, Samal SK. The case series of Eisenmenger syndrome and its pregnancy outcome in tertiary care centres of South India. Int J Reprod Contracept Obstet Gynecol. 2025;14(6):1584-88.
4. Hong S, Kang X, Lio KU, et al. Multidisciplinary approach for the management of term pregnancy complicated by Eisenmenger syndrome. J Zhejiang Univ Sci B. 2023;24(1):89-93.
5. Zhang N, Lin J, et al. Peripartum management of Eisenmenger Syndrome: A 2025 Clinical Update. J Cardiovasc Med. 2025;51(5):180-185.
6. Kim H. Current therapy of Eisenmenger syndrome. J Thorac Dis. 2022;14(10):4100-4112.
7. Yuan SM. Eisenmenger Syndrome in Pregnancy. Braz J Cardiovasc Surg. 2016;31(4):325-329.
8. Lee SY, Ko JS, et al. Perioperative Management of a Parturient with Eisenmenger's Syndrome Undergoing Cesarean Section. Anesth Pain Med. 2024;19(2):246-251.
9. Sharma A, Jain S. Multidisciplinary management of Eisenmenger syndrome in a primigravida: A case report from a western Indian tertiary center. J Obstet Gynaecol India. 2022;72(Suppl 1):S124-127.
10. Patil V, et al. Maternal and fetal outcomes in Eisenmenger syndrome: A retrospective analysis from central India. Indian Heart J. 2021;73(4):452-456.
11. Kulkarni S, Rao R. Advanced targeted therapy in CHD-PAH: A 2025 update on successful pregnancy outcomes in South India. J Clin Diagn Res. 2025;19(2):QC01-QC05.