

Anesthetic management of a parturient with Eisenmenger's syndrome: A case report

Anshul Singh¹, Nishant Patel²

From ¹Assistant Professor, Department of Anaesthesiology, University College of Medical Sciences and Guru Tegh Bahadur Hospital, ²Assistant Professor, Department of Anaesthesiology, All India Institute of Medical Sciences, New Delhi, India

ABSTRACT

Eisenmenger's syndrome results from certain uncorrected congenital abnormalities of the heart facilitating left to right shunt and chronic volume overload of the pulmonary vasculature, leading to irreversible changes in the pulmonary vasculature. Pregnancy in the patients of this syndrome is associated with a high risk of cardiovascular decompensation, thromboembolic complications, and sudden cardiac death. We present the case of a 25-year-old pregnant woman with Eisenmenger's syndrome who delivered through the vaginal route at 37 weeks of gestation, under labor epidural anesthesia. A healthy male baby was delivered within 40 min of drug deposition in the epidural space. The postpartum period was essentially uneventful with successful maternal and neonatal outcomes.

Key words: Cesarean section, Eisenmenger's syndrome, Pregnancy, Vaginal delivery

Eisenmenger's syndrome results from certain uncorrected congenital abnormalities of the heart facilitating left to right shunt and chronic volume overload of the pulmonary vasculature, leading to irreversible changes in the pulmonary vasculature. As a result, higher pulmonary arterial pressures develop, eventually leading to bidirectional shunt through the defect. This results in deoxygenated blood being shunted to the systemic circulation, progressive hypoxemia, and heart failure. Pregnancy in the patients of this syndrome is associated with a high risk of cardiovascular decompensation, thromboembolic complications, and sudden cardiac death [1]. Therefore, it is recommended that pregnancy should be avoided, or followed by early termination in these patients [2,3]. However, highly desirable patients, who reach the end of pregnancy, need multidisciplinary care and management by a team consisting of experienced specialists from anesthesiology, obstetrics, pulmonary medicine, cardiology, neonatology, and critical care medicine.

We present a rare case of a 25-year-old pregnant woman with Eisenmenger's syndrome who delivered through the vaginal route at 37 weeks of gestation, under labor epidural anesthesia. The patient was managed successfully under labor epidural analgesia throughout the postpartum period as well, with an essentially uneventful stay in the ICU.

CASE REPORT

A 25-year-old pregnant woman with Eisenmenger's syndrome was admitted for observation and delivery at a period of gestation of 34 weeks. Her medical treatment constituted tablet Lasilactone 20 mg BD. At 37 weeks of gestation, a trial of normal vaginal delivery was planned.


The American Society of Anesthesiology (ASA) mandated monitors were attached and the left radial artery was cannulated. Her blood pressure was 113/74 mm Hg, pulse rate was 85 per min, and oxygen saturation was 92–93% on room air. Two-dimensional echocardiography findings revealed a large patent ductus arteriosus (PDA) with the bidirectional flow, with severe pulmonary artery hypertension and end-diastolic gradient of 90 mmHg. Her chest radiography findings revealed cardiomegaly.

She was administered epidural anesthesia after the onset of the first stage of labor. Inj. Syntocin infusion was started after the onset of the first stage. Under aseptic precautions, after obtaining loss of resistance (LOR) through an 18 G Tuohy needle, a catheter was inserted. 3 ml of 0.125% bupivacaine with 2 micrograms/ml of fentanyl was administered. The patient became comfortable and a male baby was delivered after 40 min of giving the single bolus. Apgar scores at both 1 and 5 min were 10.

Post-delivery, the patient remained hemodynamically stable with blood pressure in the range of 100–120/50–70 mm Hg, oxygen saturation of 92–93% on room air, and heart rate in the

Correspondence to: Dr. Nishant Patel, Department of Anesthesiology, All India Institute of Medical Sciences, New Delhi - 110029, India. E-mail: pateldrnishant@gmail.com

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range of 80–85 beats per minute. Labor epidural was removed 24 h after delivery after giving two boluses constituting injection morphine sulfate 2 g at an interval of 12 h. Puerperium was essentially uneventful with a stable hemodynamic profile.

In the postpartum period, the patient was restarted on diuretics. 2D ECHO following 48 h was associated with the same pulmonary artery pressure and flow across the PDA. The patient was discharged on postoperative day 7 after an uneventful postoperative course.

DISCUSSION

Congenital heart defects that may result in Eisenmenger's syndrome include ventricular septal defect (VSD), atrioventricular septal defect (AVSD), or atrioventricular canal defect (AVCD), PDA, atrial septal defect (ASD), and D-transposition of the great vessels. Uncorrected defects facilitate left to right shunt, leading to chronic volume overload and shear stress to the pulmonary vasculature. This leads to irreversible changes in the pulmonary arterioles in the form of thickening and chronic pressure overload. Persistent pulmonary over circulation gives rise to raised pulmonary artery pressure, and pulmonary artery hypertension (PAH). When the pulmonary arterial pressure is too high, bidirectional shunt or reversal of shunt ensues, which is the hallmark of Eisenmenger's syndrome. In Eisenmenger's syndrome, deoxygenated blood is shunted to the systemic circulation, leading to progressive hypoxemia, dyspnea, severe cyanosis, and progressive right ventricular failure.

Pregnancy imposes a hemodynamic burden to fulfill metabolic demands of growing fetus and mother, which leads to rapid cardiac decompensation and sudden death in this patient population. The physiological volume expansion of pregnancy, in the presence of PAH and decreased systemic vascular resistance further exaggerates the right to left shunt that worsens the hypoxia, setting a vicious cycle of further increases in the PAH and right ventricle overload. The strained right ventricle is further strained due to decreased oxygen supply on account of worsening hypoxia. This mechanism explains the high maternal mortality rate of 30–70% during pregnancy in Eisenmenger's syndrome [4]. Cardiovascular load peaks at around 32 weeks of gestation, and most of the patients with Eisenmenger's syndrome present with the highest risk of morbidity and mortality from uncontrollable heart failure at this stage [1].

The neonatal outcomes are also guarded with a high risk of spontaneous abortion, intrauterine growth restriction (IUGR), small for gestational age (SGA), preterm birth, low birth weight, and congenital cardiac malformations [5]. In our case, a healthy male baby was delivered with none of the above-mentioned complications.

Patients who are desirous of continuing pregnancy should be advised bed rest and administered oxygen therapy. Eisenmenger's syndrome parturient can present for cesarean section, labor analgesia, or post-delivery care in the ICU. Indications for delivery include deterioration of the maternal condition, the onset of labor, or fetal distress. As such, delivery by vaginal route is not contraindicated in Eisenmenger's syndrome.

Regarding the anesthetic technique of choice, both general anesthesia and regional anesthesia have been suggested [6]. General anesthesia is associated with a prominent control of hemodynamics and stability of systemic vascular resistance (SVR). However, on the downside, its use is associated with aspiration risk, encountering a difficult airway and intubation response, if the patient has associated pregnancy-induced hypertension. There is also a theoretical concern of surge in PVR due to catecholamine release during laryngoscopy, intubation, and surgical stimulation which might exacerbate right to left shunt. Positive pulmonary ventilation has the risk of further reducing venous return and systemic blood pressure, further increasing right to left shunt. Many authors have successfully used slowly titrated epidural anesthesia and single-shot spinal anesthesia in these patients. In a review of 103 participants with Eisenmenger's syndrome, Martin *et al.* demonstrated the safety of regional anesthesia, suggesting its use in this patient population [7]. Apart from avoidance of the aforementioned drawbacks, avoidance of myocardial depression is another noteworthy benefit associated with RA. T6 level of the sensory blockade should be targeted while administering RA in this patient population, to avoid the bradycardia associated with cardiac myo-accelerator fibers blockade [8].

Subarachnoid block can lead to precipitous fall in SVR, which can lead to an exaggerated right to left shunt and circulatory collapse. Minicucci *et al.* administered single-shot spinal anesthesia using Levobupivacaine in an Eisenmenger's syndrome parturient with subsequent administration of Inj. Ketamine and Inj. Phenylephrine [9]. Sudden drop of SVR after a single-shot spinal block is a concern that has been addressed by the use of vasopressors such as norepinephrine and phenylephrine [9,10].

The advantage of slow titration of anesthesia according to the level and thus, avoidance of profound reduction in SVR makes epidural anesthesia a preferred technique of choice over the subarachnoid block. Graded boluses constituting local anesthetic and opioids, would cause an even less prominent fall in SVR, and therefore no effect on the right to left shunt. However, unreliability and the longer onset of blocks are the recognized disadvantages.

Labor epidural was planned to provide analgesia and blunt the catecholamine surge during labor. The labor duration was shortened and the baby was delivered within 40 min of the first shot of the epidural bolus. Thus, the choice of general versus epidural-spinal anesthesia should be made after considering the patient's unique physiology and with consultation with cardiologists, obstetricians, neonatologists, and cardiac anesthesiologists. Our anesthetic goals for intraoperative management included maintaining the ratio of systemic vascular resistance and pulmonary vascular resistance, by preventing sudden fall of SVR and increase in PVR. To minimize the attempts, we utilized ultrasonography for securing an arterial line, since these patients are at high risk of thromboembolism. Perioperative fluid therapy was strictly titrated, as overzealous fluids would put a strain on the already strained right ventricle. At the same time, adequate hydration was ensured in order to prevent the associated thromboembolic complications. After

the delivery of the baby, inj. Syntocinon was administered as an infusion, to avoid rapid fall in SVR [11,12].

The patient was monitored in an intensive care unit in the postpartum period since this period carries the highest risk of heart failure, thromboembolic complications, and sudden death. Initiation of early thromboprophylaxis, ensuring adequate analgesia, and oxygen therapy was the cornerstones of our post-operative management.

CONCLUSION

Anesthesiologists play a crucial role in the management of a pregnant Eisenmenger's syndrome patient, be it during delivery or intensive care management. A carefully tailored anesthesia approach based on a thorough understanding of the physiology of Eisenmenger's syndrome during pregnancy, oxygen therapy, early thromboprophylaxis, and vigilant monitoring are the keys to successful management.

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