

Emergency Caesarean Section of a Patient with Eisenmenger's Syndrome: A Tight-Rope Walk

Abstract

Successful management of a pregnant patient with complex congenital heart disease is a challenge for anesthesiologists, requiring thorough knowledge of the impact of pregnancy on the cardiac lesion. Hearing and speech impaired patients pose a barrier to effective communication between the patient and the doctors, thus increasing the anxiety and risk of complications. Here, we present a case of a hearing and speech impaired woman with the rare and dangerous Eisenmenger's syndrome, presenting for an emergency cesarean section (CS).

Keywords: *Communication barrier, Eisenmenger's syndrome, emergency caesarean section*

Introduction

Eisenmenger's Syndrome (ES), characterized by right to left or bidirectional shunting with severe pulmonary hypertension, is a rare complication of congenital heart disease during pregnancy, with a high maternal mortality of 30-50%.^[1] Women diagnosed with ES are advised to terminate their pregnancy. Successful management of ES in pregnancy is tricky and involves only a narrow margin of safety. Our challenges for this case were compounded by our inability to communicate directly with our patient due to her hearing and speech impairment.

Case

A 32-year-old, hearing and speech impaired third gravida, presented at 35 weeks of gestation, for lower segment cesarean section (CS), with non-progress of labor. We elicited her history by communicating with the patient's relatives, who in turn used sign language to communicate with her. Diagnosed with subaortic ventricular septal defect with a left-to-right shunt at 10 years of age, she had a spontaneous abortion 5 years ago and an emergency CS under general anesthesia for fetal distress 3 years ago. There was no history of cardiac failure, thrombotic events, or ES. She remained asymptomatic for the first two trimesters of this pregnancy but now complained

of dyspnoea on exertion grade III. On examination, she had grade II clubbing, pulse rate was 110 beats per minute, regular; blood pressure was 110/54 mm of Hg (right arm in lateral position), and her room air saturation was 94%. There was a loud P2 with pansystolic murmur over the left sternal border associated with thrill. She showed no signs of cardiac failure. Her electrocardiogram showed right ventricular hypertrophy. Echocardiography showed a large (24 mm), nonrestrictive, malaligned inlet VSD with bidirectional shunt, severe pulmonary stenosis (peak pressure 76 mm of Hg) with an ejection fraction of 65%. Her laboratory investigations were normal, with a hemoglobin of 11 g%. Her preoperative arterial blood gas showed a pH of 7.34, PaO₂ 60 mm of Hg, PaCO₂ 34 mm of Hg, and SaO₂ 92%.

With the use of sign language interpretation and pictures, we explained to both, our patient and her relatives, the prognosis and risks involved. The patient was offered a choice of anesthesia after explaining the risks and benefits of regional and general anesthesia. She chose to undergo epidural anesthesia. At the start of the procedure, our patient was anxious but cooperative. Hudson's mask was used to supplement oxygen at 6 L/min. Intravenous access was established with wide-bore peripheral cannulae on either hand and 14G peripherally inserted central catheter, in the right cubital vein. 500 ml Ringer's

**Gauri R
Gangakhedkar,
Rachana D
Chhabria,
Surabhee D Gour,
Yogalakshmi Palani**

*Department of Anaesthesiology,
Seth G.S. Medical College
and K.E.M. Hospital, Mumbai,
Maharashtra, India*

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Address for correspondence:

*Dr. Gauri R Gangakhedkar,
13/14, Chandangad
Apartments, Next to Rahul
Nagar, Near Karve Putala,
Kothrud - 411 038, Pune,
Maharashtra, India.
E-mail: gauri2903@gmail.com*

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lactate was administered, slowly through the duration of the surgery. Care was taken to avoid accidental intravenous injection of air bubbles and prevent paradoxical air embolism. After establishing electrocardiography, noninvasive blood pressure monitoring, and pulse oximetry, a 16 G epidural catheter was inserted at T₁₀₋₁₁ level, in sitting position. 8cc of 2% injection lignocaine and 8cc of 0.5% injection bupivacaine with 25 µg injection fentanyl were given in graded aliquots to achieve a level of T₆ over 20 min, keeping a strict watch over her hemodynamics. Aortocaval compression was avoided by placing a wedge under the right buttock. The patient delivered a male child, who cried at birth and had no congenital anomalies. Slow intravenous injection oxytocin 10U infusion was given. After baby delivery, the mean arterial pressures transiently dropped to 50 mm of Hg, responded to an intravenous bolus of 100 µg phenylephrine. There were no episodes of desaturation. After her surgery, the patient was shifted to the intensive care unit for further monitoring. Postoperative epidural analgesia was achieved with 8 hourly top-ups of 6 cc of 0.125% bupivacaine, administered slowly. She was discharged on postoperative day 7.

Discussion

Around 11% of patients with left-to-right shunts develop ES, with a history of symptom-free childhood and acclimatization to lower exercise tolerance. Over time, the shunting causes a complex pathobiological process resulting in structural abnormalities and histological changes in the pulmonary vasculature. The progressive pulmonary obstructive lesion, overall decreases in pulmonary vasculature heralds ES. ES sets in the third decade of life, with an increase in pulmonary vascular resistance (PVR) over systemic vascular resistance (SVR), ultimately leading to shunt reversal.^[2] This process was probably hastened in our patient who faced the additional triggers of increased blood volume, cardiac output in pregnancy along with the stress of labor.

The precipitous decrease in circulating blood volume during a CS can be fatal, increasing the incidence of maternal mortality to 65%.^[1] Vaginal delivery with a shortened second stage of labor is much more desirable.

The anesthetic goal is to maintain cardiac output, SVR and reduce PVR, to minimize the shunt reversal.^[3] General and spinal anesthesia can decrease SVR causing an increase in shunt and hypoxemia. Epidural anesthesia decreases PVR and SVR by sympathetic block, reduces catecholamine levels, decreases tachycardia, myocardial oxygen consumption, and reduces the right-to-left shunting, making it safe for use.^[1] Oxygen, a pulmonary vasodilator, decreases the right-to-left shunt thus improving oxygenation. Large doses of oxytocin have marked vasodilating effect and should be administered cautiously. Methylergometrine and prostaglandins are avoided due to increased PVR.

The period of maximum mortality extends from delivery to the first post-partum week, necessitating intensive postoperative monitoring.^[1]

Dealing with a hearing and speech impaired patient can intensify the challenge while dealing with high-risk obstetric patients. In a multicentre study, Bartlett *et al.* found that patients with communication problems were three times more likely to experience a preventable adverse event than other patients.^[4] Trained interpreters are preferred to converse with hearing impaired patients as relatives do not possess medical interpretation skills and are too emotionally involved to remain objective.^[5] While the choice of epidural anesthesia by our patient was the safest option for her, it imposed an additional challenge, since her co-operation was required for the procedure. Since trained interpreters were unavailable, we had to rely on the patients' relatives to communicate with and counsel her. Neuraxial anesthesia can be explained using videos or flash cards.^[6] Good intraoperative communication with the patient is necessary to assess sensory level of blockade and allay anxiety. Poor assessment and management of perioperative pain can have profound effects on patients leading to anxiety, sleep disturbances, irritability, aggression, and unwanted stress.^[7] This would prove particularly hazardous in our patient, where it could potentially trigger cardiac decompensation. The need for the use of flash cards, interpretation for communication, increases the time required to obtain the requisite consent, thus putting a strain on the anesthesiologists during medical emergencies. We were fortunate to have had been involved in the preoperative period when there was no pressing maternal or fetal emergency, allowing us to successfully communicate, explain the prognosis, risks involved, allay her anxiety, and ensure her cooperation. Through the surgery, we continued to reassure our patient by holding her hand and providing positive nonverbal cues.

Conclusion

The challenge of managing complex congenital cardiac diseases in obstetric practice is augmented when risks and management strategies cannot be communicated. However, our case report highlights that with extensive planning, interdisciplinary cooperation and active efforts to overcome communication barriers, it is possible to allow the patient to understand her disease, maintain her autonomy, and make an informed choice regarding the plan of anesthesia, all while successfully managing her disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal.

The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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