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CASE REPORT

Caesarean section in Eisenmenger's syndrome: anaesthetic management with titrated epidural and nebulised alprostadil

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Pregnancy in patients with Eisenmenger's syndrome is associated with a high mortality. This article reports two cases of women with Eisenmenger's syndrome (secondary to two different primary cardiac defects) who presented with near-term pregnancies. Both the patients underwent successful elective Caesarean section with slowly titrated epidural anaesthesia. Nebulised prostaglandin E1(PGE1) analogue, alprostadil, administered immediately post-delivery resulted in a significant drop in systolic pulmonary artery pressures as measured from tricuspid regurgitant jet by transthoracic echocardiography. The postoperative period was uneventful in both patients. A slow induction of epidural anaesthesia can be a safe mode of anaesthesia for Caesarean section in pregnant patients with Eisenmenger's syndrome. Nebulised alprostadil intraoperatively or postoperatively in the intensive care unit (ICU) is readily available and a relatively cheap option as a selective pulmonary vasodilator in developing countries.

Keywords: alprostadil, Caesarean section, Eisenmenger's syndrome, epidural

Introduction

The management of the obstetric patient with Eisenmenger's syndrome remains a challenge,[1] with the mortality rate as high as 30–70%.[2] We report two cases of women with Eisenmenger's syndrome who underwent Caesarean section using similar anaesthetic techniques in a tertiary care centre.

Case history

Both patients were admitted with a history of long-standing shortness of breath. Neither woman was receiving any definitive treatment nor had any prior antenatal visit to our hospital. Patient consent was obtained prior to the procedure.

Patient 1

The first patient was a 32-year-old G1P0+0 of bodyweight 50 kg and height 155 cm (BMI = 20.8), with New York Heart Association (NYHA) class III classification, who presented at 35 weeks gestation with a diagnosis of patent ductus arteriosus (PDA). Positive findings on her physical examination were a parasternal heave, pansystolic murmur of tricuspid regurgitation (TR), short ejection systolic murmur in the pulmonary area, loud P2 with narrow split S2, bilateral vesicular breath sounds with mild right-sided basal crepitations and a baseline pulse oximeter oxygen saturation (SpO_2) of 90–94% on room air. She had a haematocrit of 42%. Her echocardiography report showed large (10 mm) PDA at the ampulla with severe pulmonary artery hypertension (PAH) with a pulmonary artery systolic pressure (PASP) of 100 mm Hg, bidirectional shunt and Eisenmengerisation, moderate TR, and right ventricular hypertrophy. She was optimised for two weeks with oral frusemide 40 mg once daily (OD), oral digoxin 0.25 mg OD, and oral sildenafil 25 mg twice a day (BID).

Patient 2

The second patient was a 25-year-old G1P0+0 of body weight 45 kg and height 150 cm (BMI = 20), with a NYHA class III classification, who presented at 36 weeks gestation. She had been recently diagnosed with a ventricular septal defect (VSD). Positive findings on her physical examination were a pansystolic murmur of TR, loud single S2 and a baseline SpO_2 of 90–91% on room air. She had a haematocrit of 31.5%. Her echocardiography report revealed a 6 mm perimembranous VSD with severe PAH, PASP = 103 mm Hg, bidirectional shunt and Eisenmengerisation, moderate TR, and mild aortic regurgitation. She was optimised for two weeks with oral sildenafil 25 mg BID.

Both patients underwent elective Caesarean section with a slowly titrated epidural to provide surgical anaesthesia. In the operation theatre, an intravenous infusion of Ringer's lactate was started and intravenous antibiotics were administered. An arterial line was inserted to allow continuous blood pressure (BP) monitoring, and central venous access was obtained. The shunt across the defect, together with PA systolic pressures, was monitored intraoperatively at regular intervals from TR jet analysis using transthoracic echocardiography (TTE) with Doppler measurements. The right radial artery was cannulated and the right internal jugular vein was catheterised. Supplemental oxygen was provided via facemask started at 6 litres per minute.

Baseline vital parameters were:

Patient 1: Blood pressure = 118/68 mm Hg, heart rate (HR) = 102 bpm, sinus rhythm, SpO_2 = 92%.

Patient 2: BP = 138/82 mm Hg, HR = 112 bpm, sinus rhythm, SpO_2 = 92% .

An epidural catheter was inserted in a sitting position at the level of the L2–L3 intervertebral space in patient 1 and at L3–4 in patient 2 using a standard loss-of-resistance technique and tested with 3 ml preservative-free 2% lignocaine with epinephrine. Titrated incremental doses of local anaesthetic were used to achieve a targeted level of sensory block at a height of T6 in both patients. Patient 1 received bupivacaine 0.5% 5 ml (25 mg) with fentanyl 50 µg initially and a repeat dose of 3 ml 0.5% (15 mg) bupivacaine after 15 min. Patient 2 was administered an initial dose of bupivacaine 5 ml 0.5% (25 mg) along with fentanyl 50 µg, and a repeat dose of 4 ml 0.5% (20 mg) bupivacaine after 15 min. Both patients achieved a Bromage score of 4 and remained haemodynamically stable with central venous infusions of milrinone (0.2–0.3 µg/kg/min) and phenylephrine (0.2–0.7 µg/kg/min). Healthy newborns with an Apgar score of 8 and 9 at 1 and 5 min respectively were delivered in both cases.

Immediately post-delivery, 5U oxytocin was infused very slowly with uterine massage resulting in a well-contracted uterus. Nebulisation with alprostadil 600 µg in 3 ml normal saline administered over 20 min resulted in a significant drop in PASP as measured from TR jet using TTE (100 to 57 mm Hg in patient 1 and 103 to 65 mm Hg in patient 2). Intraoperative TTE and Doppler monitoring also showed no increase in right to left shunt across the respective defect in both patients.

The SpO_2 remained close to baseline throughout the surgery. TTE monitoring was continued postoperatively in the ICU. Both the patients remained haemodynamically stable postpartum. They were managed in the semi-recumbent position with oxygen therapy. Analgesia was provided with an epidural infusion of bupivacaine 0.0625% titrated to effect level of pain, and the first dose of prophylactic enoxaparin was administered in the evening. The epidural catheter was removed on postoperative day (POD) 1.

Phenylephrine and milrinone were slowly titrated down and stopped on POD 2 and 3. Both patients were transferred to the ward on POD 4 on oral paracetamol 500 mg 6 hourly, oral frusemide 40 mg daily, oral sildenafil 25 mg twice daily orally and subcutaneous enoxaparin 40 mg daily. Cardiology workup of both the neonates was normal. The patients were discharged from hospital subsequently on oral aspirin 75 mg daily, frusemide 40 mg daily, and sildenafil 25 mg twice daily. The women were advised to avoid strenuous activity and further pregnancy, and to receive regular cardiology follow-up.

Discussion

Eisenmenger's syndrome is defined as pulmonary hypertension with reversed or bidirectional shunt, associated with septal defects or patent ductus arteriosus. The provision of anaesthesia carries a high risk in these patients.[3] The goal of anaesthetic management is to maintain systemic vascular resistance (SVR) in order to prevent an increase in right to left shunt. Either regional or general anaesthesia may be used,[3,4] each with its own risks and benefits. During general anaesthesia, intermittent positive pressure ventilation causes a decrease in venous return and cardiac output and an increase in pulmonary artery pressure, which together produce an increase in right to left shunt.[3] However, general anaesthesia has been successfully used and is preferred in patients receiving antithrombotic drugs due to the increased risk of subdural haematoma following epidural anaesthesia.[5] Single-shot spinal epidural anaesthesia or combined-spinal epidural anaesthesia[6] leads to a sudden fall in SVR and increases right to left shunt.

Continuous spinal anaesthesia has been used successfully in a similar case.[7] Epidural anaesthesia has been safely used in these patients for elective Caesarean section. [8] We chose the technique of titrated epidural anaesthesia with incremental doses as the slow onset of the block allows compensation for the sympathectomy below the level of the block and results in relatively stable haemodynamics.

Intravenous (IV) milrinone infusion was titrated to manage the pulmonary hypertension[9] based on PA pressure measurements by transthoracic Doppler echocardiography. But as milrinone lacks pulmonary selectivity,[9] simultaneous phenylephrine infusion was used to combat its systemic vasodilatory effects, thereby preventing any sudden fall of blood pressure in the presence of the sympatholysis of neuraxial blockade. Following delivery, autotransfusion and aorto-caval decompression increases blood flow to the right side of the heart and may lead to pulmonary hypertensive crisis, right heart failure[10] and cardiac arrest. We attempted to reduce PA pressures further during this vulnerable period immediately after baby delivery by administering nebulised PGE1 analogue alprostadil.

Alprostadil (PGE1) is a selective pulmonary vasodilator that has been used to reduce shunting in ARDS and to improve pulmonary hypertension.[11] PGE1 has been used successfully during Caesarean section in Eisenmenger's syndrome.[12] Previous case reports and non-randomised studies suggest that inhalation of aerosolised PGE1 may improve life expectancy in intractable pulmonary hypertension.[13,14] Intravenous epoprostenol has been used successfully in a pregnant patient with Eisenmenger's syndrome.[10,15] The long-term neonatal effect is not well known with a reported case of neonatal death on the 11th day of life, although maternal pulmonary hypertension in pregnancy was well managed with IV epoprostenol.[10]

Aerosolised alprostadil is preferred to the intravenous route as the latter may lead to systemic side effects like systemic arterial hypotension, and postpartum maternal bleeding. Inhaled nitric oxide, although recommended, was not used because of non-availability in our centre. Inhaled nitric oxide use is not without side effects.[16] Pulmonary vasodilators including prostaglandins are not recommended during pregnancy because of possible teratogenicity and adverse effects on uterine circulation.[6]

The cost and availability of recommended selective pulmonary vasodilators (IV epoprostenol, IV/inhaled iloprost, IV/SC treprostinil, inhaled nitric oxide)[12] is a serious concern in our country. Alprostadil may be a cheaper alternative.

The efficacy and reliability of PA pressure measurement from TR jet by TTE with Doppler in the perioperative and ICU settings in adults has not been as extensively studied and validated as has been in done in neonates.[17] TR jet measurements do not always reflect pulmonary artery pressure and can significantly under- or over-estimate it in individual patients.[18]

We elected not to use a pulmonary artery catheter because of its inherent risks,[19] the restricted availability and the difficulty of inserting it in the correct position. Incorrect placement has the potential of tip advancement through the cardiac defect. Our

perioperative management was largely guided by monitoring of PASP from TR jet utilising noninvasive transthoracic Doppler echocardiography instead of an invasive PA catheter.

In conclusion, slow-titrated epidural anaesthesia can be a safe mode of anaesthesia for Caesarean section in pregnancy with Eisenmenger's syndrome. Alprostadil, intraoperatively or postoperatively in the ICU, is a readily available and cheaper option than other pulmonary vasodilators in developing countries. Perioperative PA pressure measurement from TR jet using TTE for guiding therapy can be a noninvasive and safer alternative to pulmonary artery catheterisation.

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