Anaesthetic management of emergency lower segment caesarean section in a patient with Ebstein's anomaly

Sir,

Ebstein's anomaly was described by Wilhelm Ebstein in 1866.[1] It is characterised by tricuspid regurgitation, due to dysplastic abnormalities of the tricuspid valve, an atrialised, thin walled and poorly contractile proximal part of the right ventricle and an enlarged right atrium.[2] Disease severity depends on the degree of valvular abnormality, presence of patent foramen ovale with intracardiac shunt, pulmonary hypertension, cardiac dysrhythmias and association with Wolff–Parkinson–White syndrome (up to 20% of patients).[2,3] Clinical presentation varies from congestive cardiac failure (CCF) in childhood to incidental diagnosis in adulthood.

We recently encountered a 22-year-old primigravida, with Ebstein's anomaly and patent foramen ovale, diagnosed at 12 weeks of conception. She presented to us at 37 weeks of gestation, for emergency lower segment caesarean section for breech in labour. She had dyspnoea on exertion (NYHA class II) and a history of 3 episodes of chest pain since conception, which resolved on their own. Auscultation revealed a pansystolic murmur in the tricuspid area. There were no signs of CCF. Her investigations were normal except that the electrocardiogram showed a Right Bundle Branch Block, Right Ventricular Hypertrophy, ST- segment depression in V2–V4 and biphasic T wave in V2–V5. 2D echo showed a moderate sized (18 mm) ostium secundum defect with left to right shunt, Ebstein's anomaly with severe tricuspid regurgitation, good biventricular function and no pulmonary hypertension.

Monitoring was done with electrocardiocardiography, pulse oximetry, noninvasive blood pressure monitoring, temperature and urine output. Oxygen supplementation was started with facemask at 6l/min. A central line was avoided to prevent supraventricular arrhythmias, and two 18G cannulae were introduced. Infective endocarditis prophylaxis and antiemetic prophylaxis were given. Care was taken during fluid administration to avoid air bubbles and to prevent paradoxical air embolism.

Epidural anaesthesia was given with an 18G catheter, at L3–L4 level, in sitting position. A sensory level of T6, with an inability to move legs, was achieved in 25 minutes with titrated doses of 2% lignocaine, up to a cumulative volume of 10 ml. Her haemodynamic parameters and respiration were assessed regularly. Aortocaval compression was avoided by placing a wedge under the right buttock. Uterine incision to delivery time was 50 seconds. Baby cried immediately after birth, with an Apgar score of 9/10 at 1 minute. Oxytocin 10 U was given by slow intravenous infusion. Only 500 ml of Ringer's lactate was transfused through the surgery. The patient remained haemodynamically stable, with saturation of 97–99%, in the intraoperative and postoperative period with good urine output. She was shifted to the intensive care unit for postoperative management. 5 ml of 0.125% bupivacaine was given 8 hourly, for 48 hours for pain relief.

The impaired right ventricular size and function due to Ebstein's anomaly deteriorates due to increased blood volume and cardiac output during pregnancy.[3] The increased right atrial pressure and volume worsens the tricuspid regurgitation. Increased catecholamine levels in pregnancy further predispose to arrhythmias, especially with maternal hypoxemia and stress. Presence of arrhythmias, cyanosis or pre-eclampsia is associated with increased maternal and foetal risk.[3] Circulating catecholamines, maternal haemodynamic instability and maternal hypoxemia in mothers with congenital heart disorders are associated with prematurity, low birth weight, congenital heart disorders (2–14%) and poor neonatal outcomes.[4] Fortunately, our patient had a normal baby, weighing 2.5 kg.

The goal of anaesthesia in these patients is to maintain sinus rhythm and both preload and afterload.[5] Subarachnoid blocks and general anaesthesia (GA) can potentially worsen the right to left cardiac shunt due to sudden decrease in systemic vascular resistance. Additionally, GA is associated with increased catecholamine levels and intrathoracic pressure which further increase intracardiac shunt.[4] Epidural anaesthesia is preferential as it causes minimal intravascular volume shift, decreased catecholamine levels, prevents maternal hyperventilation and most importantly provides postoperative analgesia.[3]

Oxytocin should be administered cautiously as large doses can cause marked vasodilation.

Methylergometrine and prostaglandins are avoided due to increase in pulmonary vascular resistance. Insertion of central line increases likelihood of arrhythmias, paradoxical emboli and bacterial endocarditis.[5] Pregnant women with Ebstein's anomaly cannot withstand either haemorrhage or excessive fluid resuscitation.

Our case adds to the limited literature on anaesthetic management of pregnancy in patients with congenital heart disease. In conclusion, a multidisciplinary management, better understanding of pathophysiology of cardiovascular diseases, their effect on pregnancy, thorough preparation and utmost vigilance result in successful outcomes.

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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