Clinical Reports

Anaesthesia for Caesarean section in a pre-eclamptic patient with Ebstein's anomaly

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Anaesthetic management for Caesarean section in a pre-eclamptic patient with Ebstein's anomaly and a right to left intracardiac shunt was accomplished under general anaesthesia. Air bubbles in the venous line were meticulously eliminated to prevent paradoxical air emboli. A rapid sequence induction using standard doses of thiopentone and succinylcholine did not result in loss of consciousness and muscle relaxation was inadequate for endotracheal intubation. This problem must be anticipated in patients with extremely enlarged right atria and the dose of induction agents should be increased. Carefully controlled epidural anaesthesia may be used if the cardiac reserve is adequate.

Key words

ANAESTHESIA: obstetric; GENETIC FACTORS:
Ebstein's anomaly; SURGERY: Caesarean section;
COMPLICATIONS: pre-eclampsia.

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Ebstein's anomaly occurs in one per cent of all congenital heart defects. The basic anomaly is a downward displacement and elongation of the septal cusp of the tricuspid valve thus "atrialising" the proximal part of the right ventricle which becomes thin walled and poorly contractile.

The severity of the disease depends upon the degree of stenosis of the tricuspid valve, presence of a patent foramen ovale and a right to left intracardiac shunt, pulmonary hypertension and cardiac dysrhythmias such as supraventricular tachycardia and Wolf-Parkinson-White syndrome.¹

Case report

A 23-yr-old female para 0 gravida 1 was transferred to our hospital at 33 weeks gestation with mild pre-eclampsia. She was known to have Ebstein's anomaly and complained of moderately decreased exercise tolerance and two pillow orthopnea since the 25th week of gestation. She denied chest pain, syncope, palpitations, or cyanotic spells.

On admission, her heart rate was 90 and regular, the blood pressure was 130/85 (17.3/11.3 kPa). The chest was clear. The heart sounds were normal but a loud tricuspid regurgitation murmur and a right parasternal heave were detected. There were no signs of right heart failure. The reflexes were normal. Laboratory examination showed a haemoglobin of $162 \text{ g} \cdot \text{L}^{-1}$ and normal coagulation, electrolytes, and uric acid levels. The ECG showed normal sinus rhythm with a right bundle branch block, and tall p waves in lead II. The chest x-ray showed a large globular heart with relatively avascular lung fields. A 2D echocardiogram

confirmed the Ebstein's deformity of the tricuspid valve and showed a markedly enlarged right atrium. There was also a significant right to left shunt at the atrial level. No pericardial effusion was noted. Arterial blood gases on room air were pH 7.66, $p\text{CO}_2$ 24 (3.2 kPa), and PO_2 54 (7.2 kPa). The PO_2 increased to 60 (8 kPa) on an FiO_2 of 0.35 by mask.

The patient's condition worsened in spite of bedrest and intravenous magnesium sulphate. The blood pressure rose to 150/100 (20/13.3 kPa) and she became hyperreflexic. It was therefore decided to deliver the baby by Caesarean section.

Preoperatively the patient received antibiotics as prophylaxis against bacterial endocarditis. The ECG, direct arterial blood pressure and urinary output were monitored. A large bore intravenous catheter was inserted to ensure access if blood transfusion was required. All intravascular catheters were freed from air to avoid paradoxical air emboli. The uterus was displaced to the left. The patient was preoxygenated for five minutes. During that time $5 \mu\text{g}\cdot\text{kg}^{-1}$ of fentanyl was given in an attempt to reduce the anticipated exaggerated hypertensive response to intubation that may be seen in severe pre-eclampsia. Then a rapid sequence induction using $4 \text{ mg}\cdot\text{kg}^{-1}$ of thiopentone and $1 \text{ mg}\cdot\text{kg}^{-1}$ of succinylcholine was given, but the patient was still awake and breathing spontaneously after 90 seconds. An additional $1 \text{ mg}\cdot\text{kg}^{-1}$ of thiopentone was given and this produced unconsciousness and apnoea. Tracheal intubation resulted in a vigorous cough and the blood pressure increased transiently from 150/90 (20/12 kPa) to 190/100 (25.3/13.3 kPa). The anaesthetic was maintained with 0.5–1 per cent halothane in an FiO_2 of 1.0 and small amounts ($0.3 \text{ mg}\cdot\text{kg}^{-1}$) of d-tubocurarine. Ventilation was controlled until peritoneal closure when the muscle relaxant was reversed with neostigmine and atropine and the patient was allowed to breathe spontaneously. She was extubated immediately postoperatively and monitored in the intensive care unit for 24 hours. The baby weighed 1210 g and had Apgar scores of five at one minute and eight at five minutes. Naloxone 0.02 mg was given to the baby at one minute of age.

Discussion

Patients with Ebstein's anomaly may be asymptomatic or severely disabled. In addition to significant

cardiac disease as indicated by functional impairment, marked cardiomegaly, probable pulmonary hypertension and an intracardiac right to left shunt, our patient also had pre-eclampsia.

During pregnancy, blood volume increases by 35 per cent by the middle of the third trimester. Cardiac output rises by about 40 per cent from pre-pregnant levels.² If pre-eclampsia and hypertension occurs, there is a marked rise in systemic vascular resistance and a low wedge pressure and cardiac output. Treatment with a vasodilator and fluids increases the cardiac output to normal pregnant values.³ Because of these changes, patients with severe cardiac disease may develop congestive heart failure as the demands of the foetal-placental unit for blood flow increase during gestation or if toxemia develops. After delivery of the baby, and contraction of the uterus, cardiac output again rises as blood is transfused from the uterus into the circulation.⁴ Thus the parturient is at risk for congestive heart failure for several hours post-delivery.

Ebstein's anomaly of the tricuspid valve may be accompanied by a significant right to left intracardiac shunt if the foramen ovale is patent. With an increase in systemic blood pressure, the shunt would be expected to decrease unless the right atrial pressure remains higher than left atrial pressure. This may occur because of the abnormal compliance of the right atrium as in this condition. If there is systemic hypotension secondary to drug effects or aortocaval compression, the left ventricular end diastolic pressure and therefore the left atrial pressure would decrease. This would increase the right to left shunt.

Invasive monitoring may be necessary to evaluate the intraoperative cardiac status in patients with severe heart disease. However, central venous pressure monitoring or the insertion of a pulmonary artery catheter may be technically difficult in patients with Ebstein's anomaly and lead to complications such as tachyarrhythmias, paradoxical emboli and stroke, and bacterial endocarditis. Non-invasive measurement oxygen saturation is a helpful monitor for patients with right to left intracardiac shunts.

During pregnancy, gastric emptying is delayed and often incomplete because of hormonal and mechanical factors. Because of this, patients in the last trimester must be treated as though they have a

full stomach even if they have been fasting for several hours. This implies that if general anaesthesia is chosen, a rapid sequence induction, cricoid pressure and endotracheal intubation is the method of choice for induction. Patients who have Ebstein's anomaly may exhibit prolonged induction times, increasing the risk of aspiration of gastric contents. Prolonged induction occurs because intravenous medications pool in the markedly enlarged right atrium.⁵ The bolus effect of thiopentone on the central nervous system would decrease, leading to a larger dose requirement. In addition less succinylcholine would be available at the myoneural junction because of its prolonged contact with circulating pseudocholinesterase.

Epidural anaesthesia has been successfully used for Caesarean section in a patient with Ebstein's anomaly. A two catheter technique was used to limit the dose of local anaesthetic and confine the level of the block to T₆. The patient was "prehydrated" with 500 cc's of Ringers' lactate and her legs were bound with crepe bandages to prevent hypotension.⁶

Intrathecal anaesthesia is contraindicated in patients with right to left intracardiac shunts. Profound hypotension may occur because of the rapid loss of sympathetic tone of the lower body. This would severely exacerbate maternal hypoxemia.

General anaesthesia has several advantages over the extradural technique in patient's with severe heart disease. Hypotension is usually avoided and although this complication can be treated with fluids and vasopressors, both of these may result in further instability in patients with Ebstein's anomaly. Excessive administration of fluid may lead to congestive heart failure or may increase right atrial pressure sufficiently to cause an increased right to left shunt and hypoxaemia. Ephedrine is the vasopressor of choice in obstetrics because it does not cause uterine artery vasoconstriction which may compromise foetal oxygenation.⁷ However, ephedrine is a potent beta adrenergic agonist which may precipitate supraventricular tachyarrhythmias in patients with Ebstein's anomaly. Metaraminol may be required for these patients in spite of potential uterine artery vaso-constriction seen with its use.⁷ General anaesthesia with endotracheal intubation reliably controls oxygen delivery to the patient – an FiO₂ of 1.0 can be given through the endotracheal

tube. If pulmonary congestion occurs, positive end expiratory pressure can be added to improve oxygenation.

In summary, both epidural and general anaesthesia can be used in patients with Ebstein's anomaly who require Caesarean section. Epidural anaesthesia, when carefully induced, has the advantages of reducing the risk of pulmonary aspiration of gastric contents and avoiding hypertension immediately after endotracheal intubation. If the patient has severe cardiac disease and rapidly worsening pre-eclampsia, an epidural anaesthetic may cause profound hypotension that is difficult to treat. General anaesthesia seldom causes hypotension and endotracheal intubation enables the anaesthetist to control oxygen delivery. However, patients with Ebstein's anomaly may require larger doses of induction agents and a prolonged induction time should be anticipated.

References

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Résumé

La conduite d'une anesthésie générale pour une césarienne chez une patiente pré-éclampsique avec une anomalie d'Ebstein et un shunt intra-cardiaque droit-gauche est décrite. Les bulles gazeuses dans les lignes veineuses ont été méticuleusement éliminées afin de prévenir une embolie gazeuse paradoxale. Une séquence rapide d'induction utilisant les doses standards de thiopentone et de succinylcholine était incapable d'assurer une perte de conscience et une relaxation musculaire adéquate pour l'intubation endotrachéale. Ce problème doit être anticipé chez les patients présentant une oreillette droite extrêmement élargie et les doses d'agents d'induction doivent être augmentées. Une anesthésie épidurale soigneusement contrôlée peut être utilisée si la réserve cardiaque est adéquate.