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CASE REPORT

Caesarean section using total intravenous anaesthesia in a patient with Ebstein's anomaly complicated by supraventricular tachycardia

A. J. R. Macfarlane, S. Moise, D. Smith

Department of Anaesthesia, Glasgow Royal Infirmary, Glasgow, UK

SUMMARY. Ebstein's anomaly is a rare congenital cardiac defect associated with both displacement and incompetence of the tricuspid valve. The condition is commonly complicated by supraventricular tachycardias. We describe the management of a patient with this condition undergoing caesarean section. Propofol and remifentanil total intravenous anaesthesia resulted in haemodynamic stability and delivery of a healthy baby who breathed spontaneously after two minutes.

Keywords: Anaesthesia obstetric; General anaesthesia; TIVA; Heart congenital defects; Arrhythmia; Ebstein's anomaly

INTRODUCTION

The number of parturients presenting with congenital heart disease (CHD) is increasing due to improved survival rates. Currently 85% of children with CHD can now expect to survive until adulthood, and therefore reproductive age, compared to 20% before the development of paediatric cardiac surgery.[1] Ebstein's anomaly accounts for less than 1% of all CHD[2] and consists of apical displacement of the septal and posterior leaflets of the tricuspid valve, commonly resulting in tricuspid regurgitation with a dilated right atrium. An inter-atrial communication is often present. Wolff-Parkinson-White syndrome, although not present in this case, is also commonly associated, predisposing to tachyarrhythmias. Evidence-based anaesthesia is difficult to practice in these situations. We could find no reports on the use of total intravenous anaesthesia for caesarean section in a patient with Ebstein's anomaly. We describe the use of propofol and remifentanil to anaesthetise a woman with Ebstein's anomaly, complicated by frequent supraventricular tachycardias (SVT), for caesarean section. The anaesthetic implications of Ebstein's anomaly and pregnancy are discussed.

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Correspondence to: Dr Drew Smith, Department of Anaesthesia, Glasgow Royal Infirmary, 84 Castle Street, Glasgow, G4 0SF UK.
Tel.: +0141 211 4620.
E-mail: ds.anaes.gri@btinternet.com.

CASE REPORT

A 27-year-old primiparous woman known to have Ebstein's anomaly was transferred urgently to our unit at 38 weeks' gestation for delivery. Her condition had been diagnosed at nine months of age but she had remained well until 20 years of age when she developed symptomatic SVTs. These were well controlled with sotalol, however, and she remained otherwise active (New York Heart Association class 1). She had no other past medical history.

Once pregnancy was diagnosed the β blocker was discontinued by her cardiologist because of the small risk of causing fetal abnormalities including growth restriction, fetal bradycardia and neonatal hypoglycaemia. Unfortunately from 26 weeks she suffered frequent SVTs, which despite taking verapamil (200 mg daily) occurred weekly. She was readmitted at 38 weeks to her local hospital with a prolonged, self terminating episode during which she was profoundly hypoxic (arterial oxygen saturation [SpO_2] 79% whilst breathing oxygen 15 L/min). There was also ultrasound evidence of increasing fetal growth restriction. In view of this and the increasing frequency and severity of maternal arrhythmias the woman was transferred to our hospital for delivery as cardiac surgical and intensive care services were available.

On arrival her heart rate was 88 beats/min, non invasive arterial blood pressure 136/92 mmHg, respiratory rate 16 breaths/min and SpO_2 79% breathing room air,

improving to 85% breathing oxygen 15 L/min. Examination revealed central cyanosis but no clubbing and lung fields were clear. Initial investigations confirmed an arterial PO_2 of 5.02 kPa breathing room air. Blood tests revealed a mild polycythaemia (Hb 15.3 g/dL) but otherwise full blood count, coagulation and biochemistry were normal. Resting electrocardiogram (ECG) demonstrated sinus rhythm with incomplete right bundle branch block but no pre-excitation. At this stage cardiotocography was satisfactory and fetal ultrasound confirmed growth restriction. During echocardiography a dilated right heart was visualised with an estimated pulmonary artery systolic pressure of 16 mmHg above central venous pressure (CVP). There was a right to left shunt during the Valsalva manoeuvre consistent with a patent foramen ovale (PFO). The septal leaflet of the tricuspid valve was apically displaced causing 2.2 cm of atrialisation of the right ventricle.

She subsequently developed a further episode of SVT which was treated by chemical cardioversion with intravenous adenosine (6, 12, 12 mg given). During this, arterial SpO_2 fell to 65% and ECG demonstrated an AV nodal re-entry tachycardia.

A multidisciplinary decision was made to proceed to caesarean section. This was to avoid shunting during an active second stage and also a possible emergency caesarean section or assisted delivery following induction of labour, both of which were more likely to be needed as the woman was nulliparous. The patient was fasted and premedicated with oral ranitidine 150 mg, 0.3 M sodium citrate 30 mL and verapamil 80 mg before transfer to cardiac theatre. Adenosine and DC cardioversion were ready for use in the event of further arrhythmias. With the patient in the left lateral position 14-gauge venous access was secured. In addition to standard monitoring (ECG and pulse oximetry) a 20-gauge radial arterial line and right internal jugular triple-lumen 10-cm line were placed, using a high approach with ultrasound guidance. Before induction of anaesthesia the heart rate was 68 beats/min, blood pressure 122/72 mmHg, SpO_2 99% and CVP 24 mmHg. A remifentanil infusion was started at 0.5 $\mu g\,kg^{-1}min^{-1}$. After three minutes of pre-oxygenation, a modified rapid sequence induction was then carried out by starting a target-controlled propofol infusion (Diprifusor^{TM}) set at 2 μg/mL. Cricoid pressure was applied following loss of consciousness and suxamethonium 100 mg administered. Direct laryngoscopy was straightforward (grade 1) and resulted in minimal haemodynamic response. Anaesthesia was maintained with a 50:50 oxygen-air mix, propofol at a target concentration of 2.5 μg/mL and a remifentanil infusion of between 0.05 and 0.15 $\mu g\,kg^{-1}min^{-1}$.

A live girl was delivered 8 min after induction of anaesthesia. Apgar scores were 8 and 9 at 1 and 5 min respectively. Hand ventilation was required for 2 min before the baby showed signs of spontaneous respiration. No opioid antagonism was administered. The baby was transferred to the special care baby unit and discharged after 24 h having required no further medical management.

Following delivery an intravenous oxytocin infusion of 20 units was given over 4 h. As the woman had developed a rash following previous penicillin exposure, ceftriaxone 1 g and metronidazole 500 mg were administered i.v. in accordance with the local obstetric protocol to reduce infection-related complications such as endometritis, wound infection and urinary tract infections.

The remainder of surgery was uneventful. Towards the end the remifentanil dose was reduced and morphine 25 mg administered i.v. Paracetamol 1 g was also given rectally. The heart rate had remained between 60 and 80 beats/min throughout and the blood pressure also varied by less than 20% of baseline with only one 0.5-mg bolus of metaraminol required. Estimated blood loss was 300 mL, and 400 mL of Hartmann's solution was administered i.v. during the procedure.

Following removal of the tracheal tube the patient was transferred to the cardiac intensive care unit for 24 h of monitoring. Regular oral paracetamol and i.v. morphine patient-controlled analgesia were used for postoperative pain relief. Enoxaparin 40 mg daily was prescribed. Although the dose of verapamil was increased to 240 mg daily postoperatively, the patient had one further episode of SVT four days later requiring chemical cardioversion. Otherwise the postoperative period was unremarkable and mother and baby were discharged after 6 days.

DISCUSSION

Ebstein's anomaly was first described in 1866[3] and is an uncommon but complex form of congenital cardiac disease with a broad pathology that can result in a variety of complications. The clinical course is variable, ranging from intrauterine death to survival into old age.[4] It is characterized by an abnormal septal tricuspid leaflet (and often also the posterior leaflet) that is apically displaced from the atrioventricular ring. This causes a portion of the right ventricle to be located at the atrial side of the tricuspid valve i.e. "atrialised". The displaced septal leaflet is associated with discontinuity of the central fibrous body, which causes a potential substrate for accessory pathways and pre-excitation. Pre-excitation and Wolff-Parkinson-White syndrome are more frequently associated with this anomaly (10%-29%) than any other congenital heart defect.[5] Supraventricular tachyarrhythmias therefore often occur and AV nodal re-entrant tachycardia and atrial flutter or fibrillation