Case Report

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Anaesthetic Management of a Pregnant Lady with Epstein Anomaly

Katta Dinesh Chandra Reddy[1], Chandramohan[2], N. Kiran[3], Ravi Madhusudhana[4]

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Abstract

Wilhelm Ebstein first described the clinical and anatomical features of an anomaly of the tricuspid valve in 1866, which occurs in 1 percent of congenital heart defects (1 in 110,000 of the general population). It is characterized by dysplastic abnormalities of both basal and free attachments of the tricuspid valve leaflets, with downward displacement and elongation of the septal and anterior cusp, with resulting tricuspid regurgitation. Congestive heart failure and sudden collapse are the most common causes of death

Case Report: A 21 year old primi with 37 wks gestation was referred to our hospital for complicated pregnancy management. She complained of occasional palpitations, and dyspnoea on severe exertion. She had no history of recurrent chest infections, cyanosis or heart failure in the past. On examination she had pansystolic and mid-diastolic murmurs with right parasternal thrill, along with widely split second heart sound. The ECG showed sinus rhythm with right bundle branch block. Chest X-ray showed enlarged right atrium with cardiomegaly, while echocardiogram showed the presence of moderately enlarged right atrium, tricuspid regurgitation with downwardly displaced tricuspid valves, confirming diagnosis of Ebstein's anomaly. Her blood reports were within normal limits. We kept ready all routine and emergency drugs and equipments.

Subsequently she was shifted into the OT, monitors were attached, and given antibiotics for bacterial endocarditis prophylaxis. A sitting epidural was tried at L3-4 interspace. INJ bupivacaine 0.5% 15cc given in graded doses of 5 cc every 5 minutes by monitoring hemodynamic response. Level achieved till T6. Thereafter left lateral tilt and oxygen were continuously administered to the patient. Intraoperatively her vitals were stable and duration of operation was 60 minutes and a female baby of birth weight 2.4kg was extracted. Postoperatively the patient vitals were stable.

Conclusion: In conclusion, women with Ebstein's anomaly may present with a multitude of problems and, should be considered as high risk and cared for in tertiary centres by a multidisciplinary team including obstetricians, cardiologists and obstetric anaesthetists in pregnancy and during delivery. Epidural anaesthesia in a fractionated or 'graded' manner provides a haemodynamically stable patient with adequate analgesia, and may be preferred to general anaesthesia in these patients.

Author's Affiliation: [1]Postgraduate Resident, [2]Assistant Professor, [3,4]Professor, Department of Anesthesiology, Sri Devaraj Urs Medical College, SDUAHER, Kolar 563101, Karnataka, India.

Corresponding Author: N. Kiran, Professor, Department of Anesthesiology, Sri Devaraj Urs Medical College, SDUAHER, Kolar 563101, Karnataka, India.

E-mail: dr.nkiran@gmail.com

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Key Messages: In presence of Ebstein's anomaly, there is impaired right ventricular size and function, further deteriorated by the increased blood volume and cardiac output during pregnancy. Increased right atrial pressure and volume both worsen tricuspid regurgitation. Raised catecholamine levels in pregnancy further predispose to arrythmias, particularly with maternal hypoxaemia and stress.

The basic principles for anaesthetizing a patient with cardiac disease are maintaining both afterload and preload along with a sinus rhythm. The advantages of epidural anaesthesia are minimal intravascular volume shift, decreased catecholamine levels, control of maternal hyperventilation and most importantly postoperative analgesia. Intrathecal anaesthesia may complicate a right to left cardiac shunt due to sudden decrease in sympathetic vascular resistance.

GA is associated with increased induction time with right to left shunts (increasing aspiration risk), increased catecholamine levels and an increased intrathoracic pressure which further increase intracardiac blood shunting. Large doses of synthetic oxytocin have marked vasodilating effects and administered cautiously, while methylergometrine and prostaglandines increase pulmonary vascular resistance and are generally avoided. Insertion of a centralline in patients of Ebstein's anomaly is technically difficult, and increases likelihood of arrhythmias, paradoxical emboli, and bacterial endocarditis.

INTRODUCTION

Wilhelm Ebstein first described the clinical and anatomical features of an anomaly of the tricuspid valve in 1866, which occurs in 1 percent of congenital heart defects (1 in 110,000 of the general population). It is characterized by dysplastic abnormalities of both basal and free attachments of the tricuspid valve leaflets, with downward displacement and elongation of the septal and anterior cusp, with resulting tricuspid regurgitation. Thus the proximal part of the right ventricle is 'atrialised', becoming thin walled and poorly contractile, along with an enlarged right atrium. Disease severity depends upon the degree of valvular abnormality, presence of a patent foramen ovale with intracardiac shunting, pulmonary hypertension, ventricular and supraventricular tachycardias and particularly association with Wolf-Parkinson-White syndrome (up to 20% of patients). Congestive heart failure and sudden collapse are the most common causes of death.

CASE REPORT

A 21 year old womanprimi with 37 wks gestation was referred to our hospital for complicated pregnancy management. She complained of occasional palpitations, and dyspnoea on severe exertion. She had no history of recurrent chest infections, cyanosis or heart failure in the past. On examination she had pansystolic (increasing with inspiration) and mid-diastolic murmurs with right parasternal thrill, along with widely split second heart sound. The ECG showed sinus rhythm with right bundle branch block. Chest X-ray showed enlarged right atrium with cardiomegaly, while echocardiogram showed the presence of moderately enlarged right atrium, tricuspid regurgitation with downwardly displaced tricuspid valves, confirming diagnosis of Ebstein's anomaly. There was no atrial septal defect or pulmonary hypertension. Her blood reports were within normal limits. We kept ready all routine and emergency drugs and equipments.

Subsequently she was shifted into the OT, monitors (ECG, pulse oximeter and NIBP) were attached, and given antibiotics for bacterial endocarditis prophylaxis. A sitting epidural was tried at L3-4 interspace using a 18 G Tuohy needle and was sited at L3-4 interspace. INJ bupivacaine 0.5% 15cc given in graded doses of 5cc every 5 minutes by monitoring hemodynamic response. Level achieved till T6. Thereafter left lateral tilt and oxygen were continuously administered to the patient. Intraoperatively her vitals were stable and duration of operation was 60 minutes and a female baby of birth weight 2.4kg was extracted. Postoperatively the patient vitals were stable.

DISCUSSION

In presence of Ebstein's anomaly, there is impaired right ventricular size and function, further

deteriorated by the increased blood volume and cardiac output during pregnancy.[1] Increased right atrial pressure and volume both worsen tricuspid regurgitation. Raised catecholamine levels in pregnancy further predispose to arrythmias, particularly with maternal hypoxaemia and stress.[2]

Arrythmias in Ebstein's anomaly with pregnancy have been treated with various antiarrythmics and in resistant cases cardioversion[2], without affecting the fetus. Haemorrhage along with excessive fluid resuscitation is generally poorly tolerated. Women with Ebstein's anomaly tolerate pregnancy well with good fetal outcomes. The presences of arrythmias, cyanosis or preeclampsia are associated with increased maternal and fetal risk.[3] The combined effect of increased circulating catecholamine, maternal heamodynamic instability and maternal hypoxaemia in mothers with congenital heart disorders is associated with prematurity, low birth weight and poor neonatal outcomes[4,5] and, 2-14% of these babies have congenital heart disorders. The female baby of our patient with Ebstein's anomaly was normal.

The basic principles for anaesthetizing a patient with cardiac disease are maintaining both afterload and preload along with a sinus rhythm.[5] The advantages of epidural anaesthesia are minimal intravascular volume shift, decreased catecholamine levels, control of maternal hyperventilation and most importantly postoperative analgesia.[4,5] Intrathecal anaesthesia may complicate a right to left cardiac shunt due to sudden decrease in sympathetic vascular resistance.

GA is associated with increased induction time with right to left shunts (increasing aspiration risk), increased catecholamine levels and an increased intrathoracic pressure which further increase intracardiac blood shunting.[6,7] Large doses of synthetic oxytocin have marked vasodilating effects and administered cautiously, while methylergometrine and prostaglandines increase pulmonary vascular resistance and are generally avoided. Insertion of a centralline in patients of Ebstein's anomaly is technically difficult, and increases likelihood of arrhythmias, paradoxical emboli, and bacterial endocarditis.[8]

CONCLUSION

In conclusion, women with Ebstein's anomaly may present with a multitude of problems and, should be considered as high risk and cared for in tertiary centres by a multidisciplinary team including obstetricians, cardiologists and obstetric anaesthetists in pregnancy and during delivery. Epidural anaesthesia in a fractionated or 'graded' manner provides a haemodynamically stable patient with adequate analgesia, and may be preferred to general anaesthesia in these patients.

Conflict of Interest: Nil

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