Anesthetic Management in a Patient with Ebstein Anomaly

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Abstract

Ebstein’s anomaly is a rare complex congenital heart disorder with malformation of the tricuspid valve and right ventricle associated with right to left shunt. The disease severity can range from asymptomatic patients to severe debilitated disease. We discuss the anesthetic management of a young female aged 22 years with Ebstein’s anomaly who successfully underwent cystogastrostomy under general anesthesia. Anesthesia in these patients is fraught with complications and high degree of morbidity and mortality.

Keywords: Ebstein’s anomaly; Right to left shunt; Anesthetic management

Introduction

Ebstein’s anomaly is a rare complex congenital heart disorder. First described by Wilhelm Ebstein in 1866 [1], it is a malformation of the tricuspid valve and right ventricle. The leaflets of tricuspid valve are displaced apically and there is atrialization of right ventricle (Figure 1). The resulting functional impairment of right ventricle and regurgitation of tricuspid valve retard forward flow of blood through the right side of the heart, thereby decreasing the volume of ejected blood [2]. Associated heart disease in Ebstein’s anomaly such as pulmonary hypertension, intracardiac shunting, cardiac dysrhythmias [3] have further effect on physiology. There is a wide spectrum of severity of disease with patients ranging from those who are asymptomatic to those who are debilitated [4]. Congestive heart failure and sudden collapse are the most common causes of death.

Case Presentation

Our patient was a 22 years old female weighing 35 kgs presenting with complaint of pain abdomen for 6 months. CT scan revealed pancreatic pseudocyst of size 162 ×149 mm in axial scan and 210 mm in craniocaudal extent which required cystogastrostomy. Pre-anesthesia investigations showed haemoglobin of 14.3 gm%, normal blood counts, normal liver/renal functions, normal blood sugar and normal serum electrolytes. Chest X-Ray showed enlarged cardiac silhouette (Figure 2). ECG showed right axis deviation, right ventricular hypertrophy, right bundle branch block, poor progression of R wave and T wave inversion in leads II, III and aVF (Figure 3). Her ABG report showed pH: 7.358, P0₂: 62 mmHg, PCO₂: 34 mmHg, O₂ saturation: 94%, base excess: -6.1 mmol/L. Echocardiography revealed Ebstein’s anomaly, septal lealet displaced apically by 4.4 cms, right atrium hugely dilated, small residual right ventricle, ostium secondum type atrial septal defect of size 2.4 cms with bidirectional shunt and ejection fraction of 60%. Preoperative cardiac evaluation mentioned no signs of heart failure and she was not advised any medication to improve cardiac function. After explaining the risks involved with surgery and anaesthetic management, written informed consent was obtained. Epidural block and general endotracheal anesthesia was planned. Defibrillator and all the emergency drugs were kept ready. ASA standard monitoring was used and an intravenous 18G cannula and urinary catheter were inserted for monitoring of urine output.

Under universal precautions, 18 G epidural catheter was placed in T7-T8 interspace and checked with test dose of 3 ml of 2% lignocaine with 1:200000 adrenaline. Pre oxygenation was performed for 5 minutes. Premedication was with midazolam 1 mg IV and fentanyl 100 mcg IV. The patient was induced with etomidate 5 mg IV. Muscle relaxation was achieved with rocuronium 30 mg IV. 3 ml of 2% xilocard IV was given 90 seconds before intubation to blunt the sympathetic response to intubation. Patient was intubated with a 7.0 mm ID cuffed endotracheal tube. Incremental doses of rocuronium, oxygen and nitrous oxide in 1:1 ratio and sevoflurane 1.5-2.5% on a circle system were used for controlled ventilation.

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Figure 1: Anatomy of heart in (a) normal heart (b) ebstein anomaly.
right to left shunting which may occur if there is decrease in systemic vascular resistance or increase in pulmonary vascular resistance or with increased intrathoracic pressure and avoidance of tachycardia as this leads to impaired right ventricle filling.

Etomidate was used as induction agent to maintain hemodynamic stability. Fentanyl also provides adequate hemodynamic stability. Vecuronium and rocuronium are cardio stable muscle relaxants preferred over pancuronium and atracurium [6]. Increase in arterial hypoxemia can occur due to increase in magnitude of right to left intracardiac shunt. Maintenance of right ventricular function and avoidance of an increase in pulmonary vascular resistance is needed. End-tidal CO$_2$ was kept on lower side to prevent pulmonary vascular hypertension and hence right to left shunt.

We used segmental thoracic epidural analgesia in a graded manner and were able to block T4-T12 dermatomes without any significant hemodynamic changes [7]. Phenylephrine was kept standby for use in case of reversal of shunt occurred intraoperatively. The advantages of epidural analgesia are decreased intraoperative anaesthetic requirements with minimal changes in systemic vascular resistance and heart rate, prevention of splinting of diaphragm during reversal and most importantly postoperative analgesia.

These patients are predisposed to development of supraventricular tachydysrhythmias [8]. Factors which are known to precipitate arrhythmias e.g. light plane of anesthesia, fluid or acid base disturbance, hypoxia and hypercapnia were avoided. Defibrillation may be required to terminate any possible arrhythmias. Measured IV fluids with careful titration were given to prevent right heart failure [9]. Post operatively, good pain relief, O$_2$ by mask and monitoring in ICU ensured minimal complications.

**Conclusion**

High risk cardiac patients like those with Ebstein's anomaly require coordinated and concerted effort between the surgeon, anesthesiologist and cardiologist to ensure optimal anesthesia plan and delivery with minimal complications and good perioperative outcome.

**Reference**