

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 cystogastomy 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

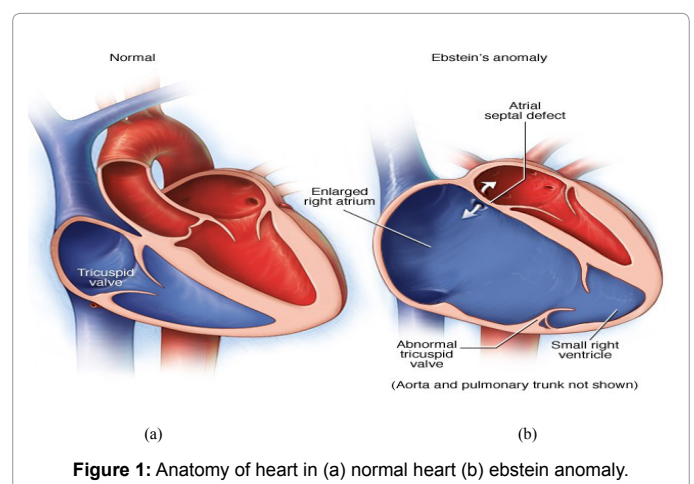
Ebstein's anomaly is a rare complex congenital heart disorder. First described by Wilhelm Ebstein in 1866 [1], it is a malformation of the tricuspid valve and right ventricle. The leaflets of tricuspid valve are displaced apically and there is atrialization of right ventricle (Figure 1). The resulting 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 cystogastrotomy. Pre-anesthesia history included reduced exercise tolerance which indicated probably a cardio-respiratory etiology. However she gave no history of palpitation, syncope, cyanotic spells or recurrent chest infections.

On examination, she was found to be an asthenic lady. Her pulse was regular (HR: 94/min), she was normotensive (BP: 118/68 mm Hg), Jugular venous pressure was not raised and there was no pedal edema, pallor or cyanosis. Airway examination was normal. Examination of cardiovascular system revealed the presence of a loud pansystolic murmur, best heard in the tricuspid area. Liver was not palpable and there were no other signs of heart failure.

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, pO₂: 62 mmHg, pCO₂: 34 mmHg, O₂ saturation: 94%, base excess: -6.1 mmol/L. Echocardiography revealed Ebstein's anomaly, septal leaflet displaced apically by 4.4 cms, right atrium hugely dilated, small residual right ventricle, ostium secundum 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% xylocard 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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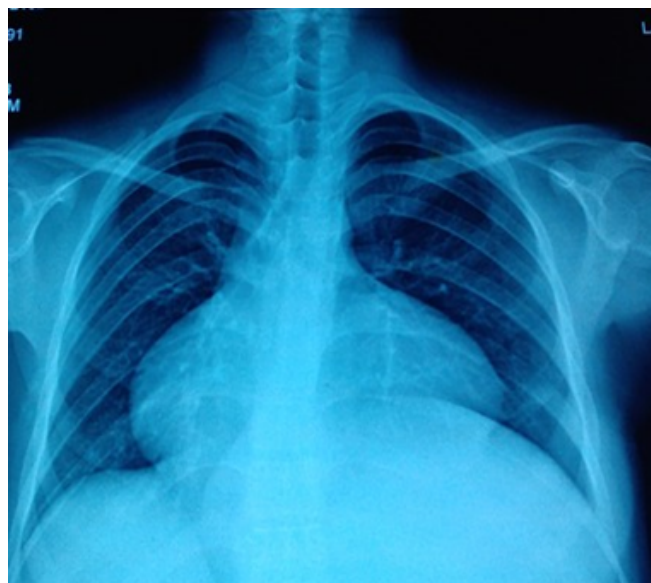


Figure 2: Chest x-ray showing cardiac anomaly.

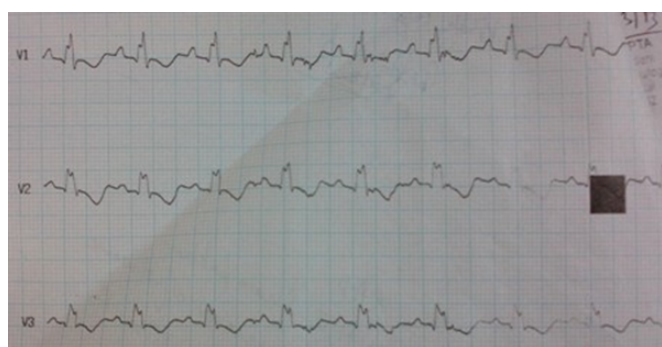


Figure 3: ECG showing right axis deviation and right bundle branch block.

Total of 10 ml of 0.25% bupivacaine and 1 mg morphine was given epidurally in incremental doses after negative aspiration. 1000 mg paracetamol IV was given as part of multimodal analgesia. A total of 1 L of ringer's lactate was used in the intraoperative period. Our patient was hemodynamically stable throughout the operation. The end-tidal CO₂ remained between 28 and 32 mmHg and SpO₂ from 94 to 88%. The total duration of surgery was around 90 minutes. At the end of surgery, reversal of neuromuscular blockade was achieved with neostigmine 2.5 mg and glycopyrrolate 0.4 mg. The recovery from anesthesia and extubation were uneventful and the patient was shifted to ICU for observation for 24 hours.

Discussion

Anesthetic management for surgical correction of Ebstein's anomaly has been described but there is paucity of case reports of noncardiac surgery in these patients. The severity of Ebstein's anomaly can be described as anatomically mild, moderate, or severe based on echocardiographic appearance of displacement and tethering of the leaflets and the degree of right ventricular dilatation [2]. Our patient had moderate/severe Ebstein's anomaly. Basic principles of anesthetic management [5] in a patient of Ebstein's anomaly are to maintain preload and afterload and maintain sinus rhythm, to prevent increased

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 CO₂ 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 tachyarrhythmias [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, O₂ 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

1. Mann RJ, Lie JT (1979) The life story of Wilhelm Ebstein (1836-1912) and his almost overlooked description of a congenital heart disease. *Mayo Clin Proc* 54: 197-204.
2. Attenhofer Jost CH, Connolly HM, Dearani JA, Edwards WD, Danielson GK (2007) Ebstein's anomaly. *Circulation* 115: 277-285.
3. Misa VS, Pan PH (2007) Evidence based case report for analgesic and anaesthetic management of parturient with Ebstein's anomaly and Wolff-Parkinson-White syndrome. *Int J Obstet Anesth* 16: 77-81.
4. Groves ER, Groves JB (1995) Epidural analgesia for labour in a patient with Ebstein's anomaly. *Can J Anaesth* 42: 77-79.
5. Rathna R, Tejesh CA, Manjunath AC, Mathew KT (2008) Anesthesia for incidental surgery in a patient with Ebstein's anomaly. *SAARC J Anesth* 1: 85-87.
6. Franco SA, Hines RL (2014) Congenital heart disease. In: Hines RL, Marshall K (eds.). *Stoelting's Anesthesia & Co-Existing Disease* (Second South Asia edition). Reed Elsevier India Private Limited, Delhi, India.
7. Chatterjee S, Sengupta I, Mandal R, Sarkar R, Chakraborty P (2008) Anaesthetic Management of Caesarean Section in A Patient with Ebstein's Anomaly. *Indian J Anaesth* 52: 321-323.
8. Macfarlane AJ, Moise S, Smith D (2007) Caesarean section using total intravenous anaesthesia in a patient with Ebstein's anomaly complicated by supraventricular tachycardia. *Int J Obstet Anesth* 16: 155-159.
9. Choudhuri AH, Uppal R, Khaitan M (2012) Laparoscopic cholecystectomy in a patient with Ebstein's anomaly: Anesthetic considerations. *Saudi J Anaesth* 6: 301-302.