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Case Report

Anaesthetic management of a case of Ebstein's anomaly undergoing a non cardiac surgery

Aakash Agarwal[1,*], Mohit Gupta[2], Nupur Gupta[3]

[1]*Dept. of Anaesthesiology, Sanar International Hospital, Gurugram, Haryana, India*
[2]*Dept. of Anaesthesiology, Marengo Asia Hospitals, Gurugram, Haryana, India*
[3]*Dept. of Critical Care Medicine, Artemis Hospitals, Gurugram, Haryana, India*

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ABSTRACT

Ebstein's anomaly is a rare congenital heart disease characterized by apical displacement of the Tricuspid valve leaflets leading to Tricuspid Regurgitation. Here we report the successful management of a case of a patient with Ebstein's anomaly who underwent Defunctioning Tranverse Colostomy. Given the complexity of Ebstein's anomaly and the potential for associated cardiac abnormalities, it is crucial to have a multidisciplinary team involved in the patient's care.

1. Introduction

Ebstein's anomaly is a rare congenital heart disease, originally described by Wilhelm Ebstein in 1866.[1] The disease is characterized by apical displacement of the tricuspid valve leaflet(septal and posterior) hinge points with normal position of the tricuspid valve orifice. This leads to partial atrialisation of the right ventricle and varying degrees of tricuspid valve insufficiency.[2] The management of a case of Ebstein's anomaly can be further complicated by associated heart conditions like cardiac dysrhythmias, pulmonary hypertension, intracardiac shunting.[3] Here we report the anesthetic management of a case of a patient with Ebstein's anomaly who underwent Defunctioning Tranverse Colostomy.

2. Case Report

A 44 years old male patient was admitted to our hospital with chief complaints of diffuse pain abdomen of recent duration. He had been operated for left sided Renal Cell Carcinoma a year back, and a current CT scan of the abdomen and pelvis revealed a large heterogeneously enhancing necrotic mass (17.5x17x12 cm) with air fluid level in left lumbar and hypochondrial region. Metastatic deposits were seen in the liver, mesentery and the omentum. The patient had no other comorbidities. It was decided to take up the patient for urgent defunctioning transverse colostomy in view of the bowel involvement by the mass.

A detailed preoperative anesthesia checkup was done. The Transthoracic 2D Echo findings revealed that the patient had a Dystrophic Tricuspid Valve, displaced Septal Tricuspid Leaflet, Severe eccentric TR, PASP=67mm Hg, severe PAH, dilated RV/RA, with a normal LV Systolic function. Based on Echo findings, a diagnosis of Ebstein anomaly was made. Preoperative investigations revealed Hb of 8.3 gm/dl, INR of 1.58; rest of the investigations were within normal limits.

In consultation with the cardiologist, the patient was started on Injection Lasix 20 mg IV BD preoperatively, to be continued till the morning of surgery. Injection Vitamin K was administered night before surgery and 1

* *Corresponding author*.
E-mail address: aakashagarwal06@gmail.com (A. Agarwal).

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Unit PRBC was given to the patient preoperatively. He was accepted for surgery under grade 4E American society of Anaesthesiology grading. A high-risk consent was obtained from the patient including consent for postoperative ICU stay and mechanical ventilation. Apart from these, routine preoperative orders were followed.

In the operating room, radial artery catheterization and intravenous (IV) cannulation was done under local anesthesia and monitoring instituted including Invasive Arterial Blood Pressure monitoring. For induction of anesthesia, he received IV Midazolam 1 mg, IV Fentanyl 2 mcg/kg, IV Etomidate 0.5 mg/kg, IV Rocuronium 1.2mg/kg. Maintenance of anaesthesia was achieved with Fentanyl 1mcg/kg hourly, Rocuronium boluses and Sevoflurane with oxygen-air mixture (50:50), and was mechanically ventilated. Central venous cannulation was done after induction for CVP monitoring intraoperatively. Normothermia was maintained throughout surgery, CVP was maintained between 10 to 12 cm of water. The surgical duration was around 2 hours.

The patient remained hemodynamically stable throughout surgery. He had a blood loss of around 100ml and Urine output during surgery was 200 ml. Total IV intake was 1400 ml. Following surgery, the patient was reversed and extubated. Recovery from anaesthesia was uneventful. During extubation, prophylactic IV Lignocaine (1mg/kg) was used to maintain hemodynamics. The patient was shifted to ICU for monitoring.

3. Discussion

In Ebstein's anomaly there is varying degree of impairment of right atrial and ventricular size and function. The goals of anaesthesia include maintenance of sinus rhythm, preload, afterload and to prevent any increase in right to left intracardiac shunt, if present.[4] Patients with Ebstein's anomaly are predisposed to occurrence of tachyarrythmias and paradoxical embolism. Left heart function can also be affected if there is severe RV dilatation leading to impaired filling and functioning of the LV.[5,6]

Our patient had severe right sided heart dysfunction with PAH with dilated RV and RA and a preserved LV function. We used IABP monitoring from the start to detect changes in hemodynamics early. CVP cannulation was done as our patient did not have any right to left intracardiac shunt. We used Etomidate for induction to avoid negative inotropic and vasodilatory effects of propofol. Fluid intake was strictly controlled to maintain CVP. Various cases of delayed induction by intravenous anaesthetics have been reported due to stasis of anaesthetic agents in enlarged right atrium, so careful dosing of intravenous agents is advised.[7] Tricuspid stenosis is generally not associated with Ebstein's anomaly and even if present is generally overshadowed by tricuspid regurgitation, so stenosis requires less consideration in terms of anesthetic management.[8] We followed the basic anesthetic considerations for a case of tricuspid regurgitation with PAH maintaining normal preload, normal to high heart rate, sinus rhythm and maintenance of afterload. All measures were taken to avoid any increase in pulmonary artery pressure like avoidance of nitrous oxide, avoiding high airway pressures, maintenance of pH, maintenance of body temperature and avoiding hypoxemia. Phosphodiesterase inhibitors milrinone and amrinone are generally the first-choice inotropes in view of their less significant alpha agonistic effects. Dobutamine was also kept standby in view of its pure beta agonistic action.

Supraventricular tachyarrhythmias are the most common arrhythmias seen in Ebstein's anomaly.[8] Adenosine and Amiodarone were kept standby for any such events. Postoperative management goals include the maintenance of hemodynamics, good analgesia and careful ICU monitoring as a standard of care.

4. Conclusion

The successful management of anesthesia in patients with Ebstein's anomaly requires a comprehensive understanding of the pathophysiology, a well-prepared anesthesia plan, and a multidisciplinary approach involving anesthesiologists, cardiologists, and cardiac surgeons. By carefully considering the unique challenges posed by this rare condition, healthcare providers can optimize patient outcomes. The anesthetic plan should be tailored to the individual patient, taking into consideration the severity of the anomaly, associated cardiac abnormalities, and the patient's overall health.

5. Source of Funding

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6. Conflict of Interest

None.

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Author biography

Aakash Agarwal, Consultant

Mohit Gupta, Senior Consultant

Nupur Gupta, Consultant

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