Volume 4 Issue 1
www.escientificlibrary.com

ISSN: 2767-6560

ES Journal of Case Reports

Laparotomy in a patient with Ebsteins anomaly- from OT to ICU.

Case Report

DOI: 10.59152/ESJCR/1040

Abhishek Biswas[1*], Amit Kumar[2] and R.P.Doley[3]

[1]DNB, IDCCM Surgical ICU, Mohali, India
[2]MD, DNB, Additional director Fortis Hospital, Mohali, India
[3]MS, MCH, Sr. Consultant Fortis Hospital, Mohali, India

Received: March 29, 2023; **Accepted:** June 20, 2023; **Published:** June 26, 2023

***Corresponding author:** Abhishek Biswas, DNB, IDCCM Surgical ICU, Mohali, India

Introduction

Ebstein's anomaly (EA) is a rare cyanotic congenital heart disease, affecting 1/200,000 live births and accounts for <1% of congenital heart diseases.[1] First described by Wilhelm Ebstein in 1866 [2], the anomaly consists of downward displacement of the septal and posterior leaflet of tricuspid valve resulting in atrialization of the right ventricle and tricuspid regurgitation. The resulting functional impairment of right ventricle retard the forward flow of blood through right side of the heart, thereby causing a hypoplastic Right ventricle(RV), or RV outflow tract obstruction. This creates an accessory conduction pathways resulting in tachyarrythmias, pulmonary hypertension, intracardiac shunting. [3] Acute decompensated heart failure and sudden collapse are the most common cause of death.[4]

There is a wide spectrum of severity of disease with patients ranging from those who are asymptomatic, surgically treated or diagnosed incidentally later in adulthood. Although Ebstein's anomaly is not generally associated with other congenital defects, patients may occasionally require surgery for other comorbid conditions or for emergency surgery as happened in our patient.

We describe the management of a patient who was a known case of Ebstein's anomaly (repaired at 2009) posted for emergency laparotomy.

Keywords: Ebstein's anomaly; Anesthesia; Congenital heart disease; laparotomy.

Case report

Our patient was a 20 yrs old male, 75 kgs who came to TRIAGE with acute abdominal pain and fever for 2 days. CT scan abdomen showed acute appendicular perforation with peritonitis and pneumoperitoneum. Pre-anesthesia history included poor exercise tolerance, recurrent chest infections, cyanotic congenital heart disease for which he was operated in 2009(Ebsteins anomaly with ASD repair). Drug history revealed he was on regular treatment with Dytor, Clopidogrel and Aspirin since 2009.

On initial examination he was awake, oriented but hypotensive (90/40 MM of Hg), tachycardic (125/ min), tachypnic 32/min with oxygen saturation 78% with 6 lts oxygen. General examination showed clubbing in all limbs. Examination of cardiovascular system revealed the presence of a loud pansystolic murmur, best heard in the tricuspid area. Generalized abdominal tenderness with poor bowel sounds on abdominal examination.

Investigations showed haemoglobin of 17 gm%, tlc 17000, CRP250/PCT 2, all other investigations were normal (liver/ renal functions,blood sugar and serum electrolytes). Initial INR was 2.9 with normal platelets. Chest X-Ray showed enlarged cardiac shadow. ECG showed right axis deviation, right ventricular hypertrophy, right bundle branch block. Her ABG report showed severe metabolic acidosis with high lactates(Ph:7.1,lactate:5,bd -10,bicarb 12).Echocardiograply showed IVC diameter 1.2 cms fully collapsing in nature, severe tricuspid regurgitation, severe pulmonary arterial hypertension with complete atrialization of the right ventricle, displacement of the interventricular septum toward left with ejection fraction 50%. There was a small patent right to left shunt. Preoperative cardiac evaluation mentioned no signs of heart failure.

After securing two peripheral lines, 2 litres of crystalloids were given, Foleys catheter, arterial line and CVC line were placed. He received Meropenam with

Metrogyl pre operatively in view of high chances of septicemia after sending blood and urine cultures.

Our patient was classified as ASA 4E and general anaesthesia was planned. The anesthetic plan, risks, benefits with post surgery plan were discussed with patients parents and informed high risk consent was obtained.

He was shifted to surgery with vitals BP 100/70, SPO2 80%on 6 lts oxygen, pulse 100/m, urine 200 ml in last one hour. Premedication was given with Midazolam 1 mg, Paracetamol 1 gm, Fentanyl 100 mcg intravenous along with preoxygenation. Rapid sequence induction was done with Etomidate 10 mg, Suxamethonium 100 mg followed by Rocuronium 50 mg. Patient was intubated with 8.0 mm ID cuffed endotracheal tube. Incremental doses of Rocuronium, Fentanyl infusion, oxygen/air and Sevoflurane 1.5-2.5% on a circle system for controlled ventilation. Midline laparotomy findings were fecal peritonitis, large bowel perforation with gangrene and mesocolic thrombosis for which resection of large bowel segment, omentectomy with ileostomy was done. Peritoneal fluid was sent for culture.

He received 2 litres of crystalloids, 4 FFP (INR 2.9) with urine output 500 ml intraoperatively. He was shifted to ICU on mechanical ventilation under sedation, with high ionotrope /vasopressor infusions (nor epinephrine/ vasopressin) along with intravenous fluids. Patient was gradually tapered off ionotropes /vasopressors with cautious intravenous fluid approach under guidance of arterial blood gas, echocardiography, urine output and lung ultrasound in ICU. Early parenteral nutrition along with albumin was started for nutrition. On mechanical ventilation his Etco2 was between 32-35 with variable Spo2 between 75-85% on Fio2 45% which was due to the patent ASD leak and shunt as confirmed by echocardiography.

His vitals remained stable on day 2 of surgery and he was subsequently extubated after spontaneous trial. While blood and urine remained sterile, peritoneal fluid showed growth of both Enterococcus Faecium and E.coli XDR (Carbapenamase resistant) for which he was put on injection Tiecoplanin and Colistin. Central line and arterial lines were taken out early to avoid infective endocarditis. As his platelets remained below 70000 till day 5 with INR ranging from 2.9(DAY 1) to 2.6 (DAY 3) we avoided anti coagulants but early mobilization and DVT pump in both legs were applied. Postsurgery pain was managed with Fentanyl and Paracetamol intravenously. He remained afebrile with stable vitals, oriented and pain free.

On day 3 of surgery he had a sudden onset of generalized tonic clonic seizure and he was eventually put on mechanical ventilation due to rapidly deteriorating GCS. MRI brain was done which showed acute infarcts in bilateral parietal lobes. On high suspicion of cardio pulmonary embolic source we did CT pulmonary angiogram and D-dimer which came out to be normal. Anti epilieptics, anti platelets and anticoagulants were started immediately. His GCS remained below 8/15 in the next two days and early tracheostomy was done on day 6 in view of prolonged mechanical ventilation. All his fresh cultures became sterile by day 12 and he was gradually weaned off antibiotics. His family decided to shift the patient to other hospital (GCS E3VTM5) for further supportive care and rehabilitation. Subsequently, we lost the patient follow-up.

Discussion

Anesthetic management for surgical correction of Ebsetin's anomaly has been described but there is paucity of case reports of noncardiac surgery and its post operative ICU management 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 .There is risk of paradoxical emboli, risk of systemic complications including stroke or brain abscess, pooling of blood in the right atrium, tachyarrhythmias, potential for increased pulmonary vascular resistance (PVR), and right ventricular dysfunction .[5]Our patient had moderate patent right to left shunt due to ASD leak with moderate dyspnea and clubbing (Hb 17, Hct 52) which suggested chronic hypoxia.

Basic principles of anaesthetia and postoperative ICU management in a patient of Ebstein's anomaly are to maintain preload, afterload, avoidance of tachycardia, maintainance of sinus rhythm, prevention of increased right to left shunting causing hypoxia. Hypoxia may also precipitate if there is decrease in systemic vascular resistance, increase in pulmonary vascular resistance, with increased intrathroacic pressure leading to impaired right ventricle filling.

Etomidate and fentanyl were used as induction agent to mantain hemodynamic stability. Rocuronium is a cardio stable muscle relaxant which is preferred over others. End-tidal CO2 was kept on lower side to prevent pulmonary vascular hypertension and hence right to left shunt. We accepted saturation around 75 to 80 as it was his base line level. As excess fluid administration can lead to an increase in right to left shunt and hypoxemia, we made every effort to prevent any excess fluid administration in intra operative and postoperative period with regular ultrasound of lungs, blood gas with lactates, arterial pulse pressure variations and urine output.

There is always high prevalence of embolism in patient of right to left shunt ASD as happened in our patient resulting in embolic stroke. We applied mechanical DVT pump with early mobilization for DVT prophylaxis. As his

coagulation parameters were already deranged we avoided anticoagulants/antiplatelets initially. After ischemic stroke we started antiplatelets/anticoagulants and kept strict monitoring of any bleeding.

Conclusion

Ebsteins anomaly posesses a complex challenge during intra operative and post operative period. We managed septic shock in such patient with judicious use of fluids with echo/usg guidance, ionotropes, vasopressors, and antibiotics. Paradoxical embolism is a known complication which we encountered as ischemic stroke in our patient. High risk cardiac patients like those with Ebstein's anomaly require coordinated and concerted effort between the surgeon, anesthesiologist and intensivist to ensure optimal anesthesia plan and post surgery care.

References

1. Attenhofer Jost CH, Connolly HM, Dearani JA, Edwards WD, Danielson GK. Ebstein's anomaly. Circulation 2007;115:277 85.

2. 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.

3. Misa VS, Pan PH (2007) Evidence based case report for analgesic and anaesthetic management of parturient with Ebstein's anomly and WolffParkinson- White syndrome. Int J Obstet Anesth 16: 7

4. Saxena KN, Mandal A, Wadhwa B (2018) Anesthetic Management in a Patient with Ebstein Anomaly. J anesthesiol pain res 1: 105.

5. Anesthetic Care During Posterior Spinal Fusion in an Adolescent With Ebstein's Anomaly Kenneth Phia, e , David P. Martinb, c , Allan Beebed, J Med Cases. 2020;11(3):68-72