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RESEARCH ARTICLE

ANAESTHETIC MANAGEMENT OF A PATIENT WITH EBSTEIN'S ANOMALY POSTED FOR CEREBRAL ABSCESS DRAINAGE

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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 the right to left shunt. Severity can range from asymptomatic to severely debilitated. We have discussed a case of a 12-year-old female patient with Ebstein's anomaly for cerebral abscess drainage.

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Introduction:-

Ebstein's anomaly was first described by Wilhelm Ebstein.(1) It is a rare congenital cardiac anomaly represented by malformation of the tricuspid valve and right ventricle. It often results in pathological tricuspid regurgitation, right ventricular failure and arrhythmias. Common clinical features include exertional dyspnea, palpitations and cyanosis. Congestive cardiac failure and sudden collapse are the most common causes of death.(4) Cerebral abscess is also one of the common complications. This case report underlines the pathophysiology of Ebstein's anomaly and its anesthetic management for noncardiac surgery. It emphasizes the role of the interprofessional team in its management.

Case Presentation:

Our patient was a 12 year old female weighing 40 kgs. with complaints of headache and fever since 17-18 days. There was no history of seizures, fatigue or syncope. She was suspected to have cerebral abscess.

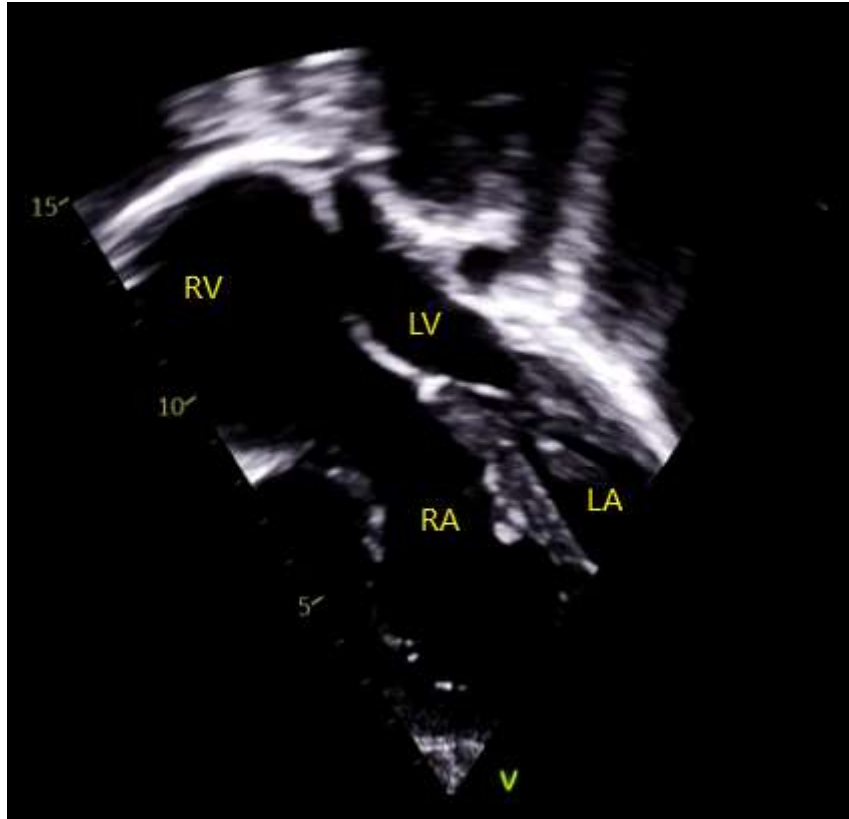
In her childhood, she had complaints of cyanosis. Her 2D echo at that time was suggestive of cyanotic congenital heart disease and hence, was advised corrective cardiac surgery; however she did not undergo any surgery. She did not have any physical developmental delay, had limited exercise tolerance and was subnormal in her mentation. There was no history of palpitations, cyanotic spells or recurrent chest infection.

During pre-anaesthetic evaluation, she was conscious and irritable. She had good exercise tolerance. The SpO₂ reading was 80-85% on room air and clubbing was present. On auscultation, patient had a grade 2 late systolic murmur in tricuspid area. No signs of heart failure were evident.

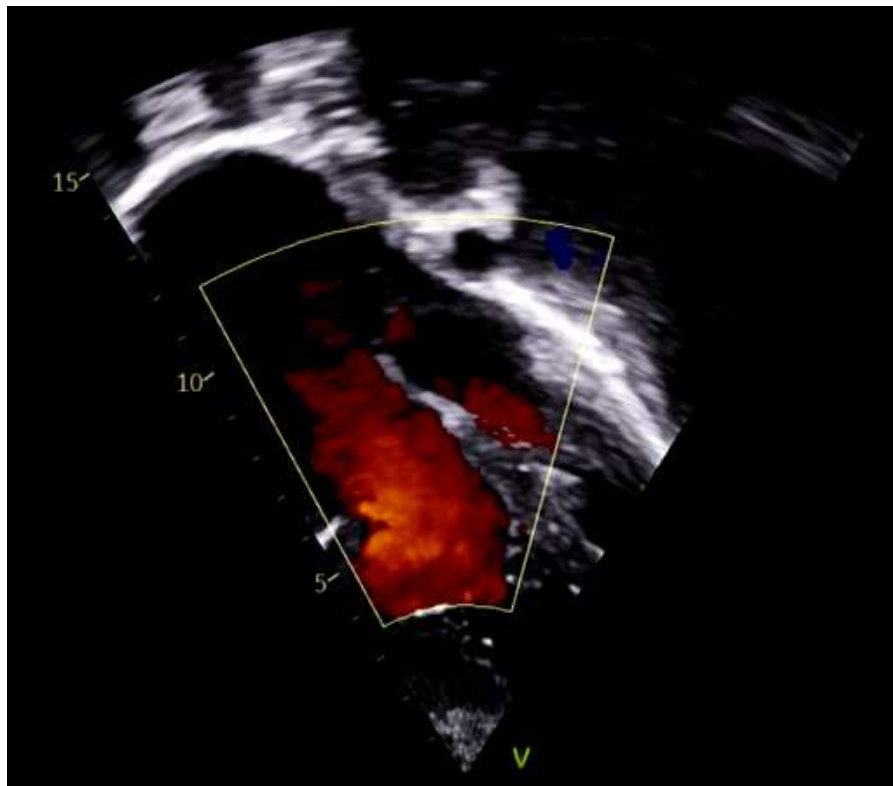
Her investigations showed haemoglobin of 17.7 mg/dl, other blood investigations were unremarkable. Chest X-Ray showed cardiomegaly, ECG showed normal sinus rhythm. 2D Echo demonstrated Ebstein's anomaly of the tricuspid valve, redundant and hypoplastic septal leaflet of tricuspid valve, non coaptation of anterior and septal leaflet with almost free tricuspid regurgitation, and moderate secundum ASD with bidirectional shunt.

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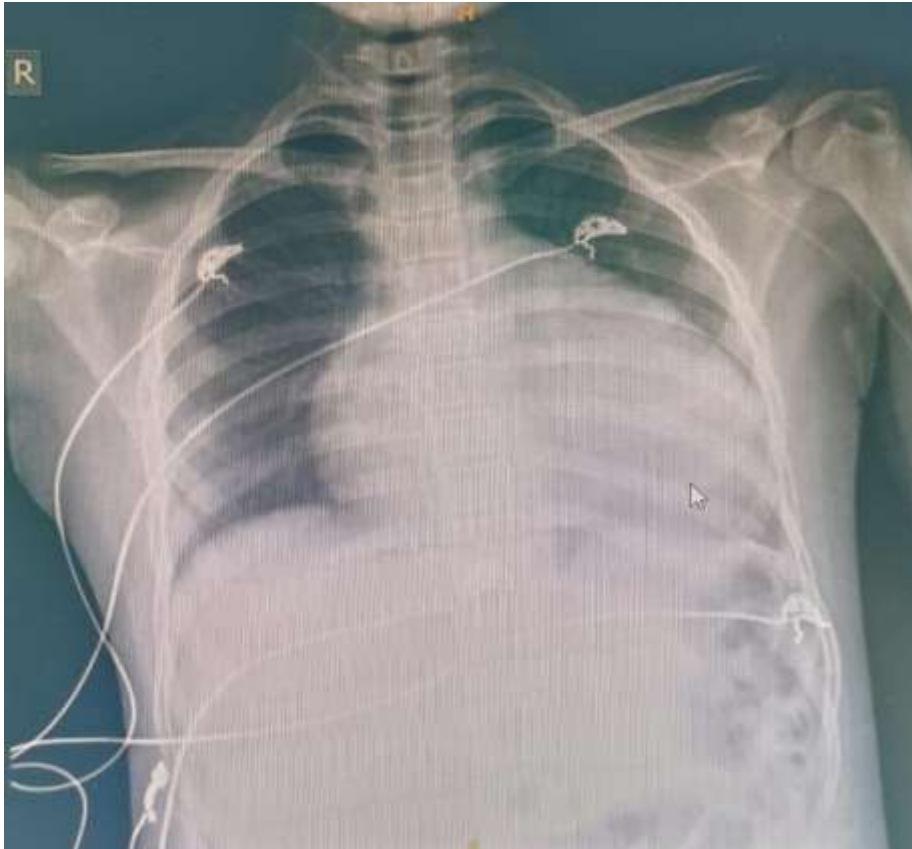
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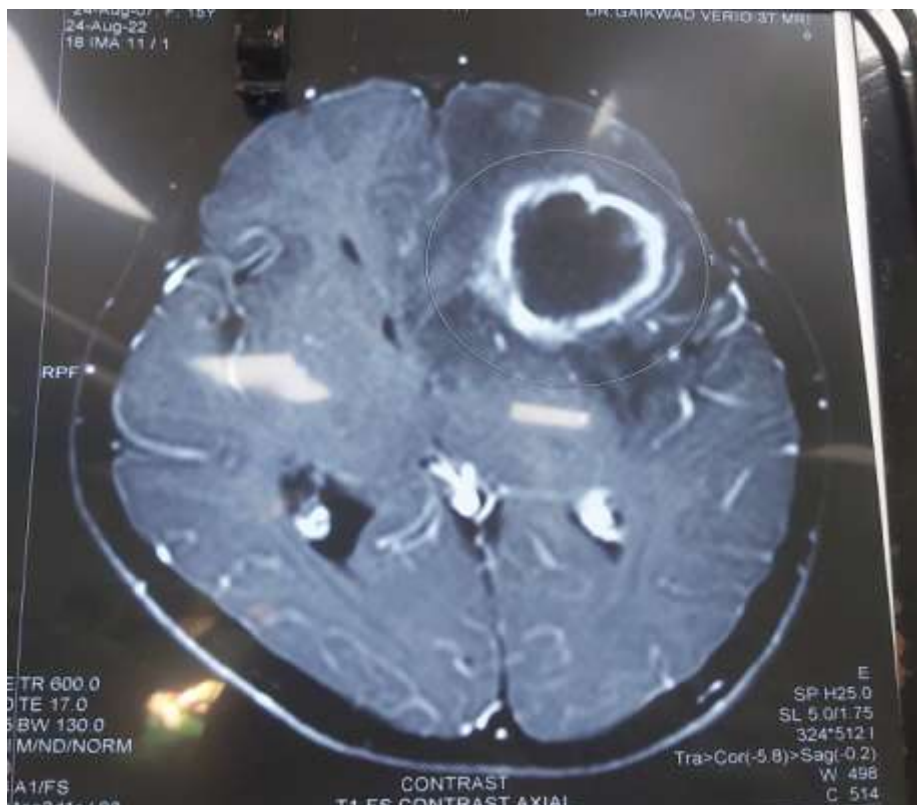
2D Echo showing Right ventricle enlarged as compared to left ventricle, atrialized Right ventricle- right heart enlargement, with atrial septal defect (ASD).



2D Echo view with Doppler showing tricuspid regurgitation



Chest X-Ray showing cardiomegaly.



CT scan of the brain showed 3.6x3.4 cm hypodense lesion with significant perilesional oedema in left frontal region, causing mass effect due to compression. MRI brain was suggestive of cerebral abscess, likely bacterial in origin, possibly tubercular.

A cardiology opinion was sought, advised high risk for general anesthesia, to watch for arrhythmias, keep antiarrhythmic drugs ready, and to shift the patient to PICU and ventilatory support if required. She was not initiated on any medications for cardiac optimization.

General anesthesia was planned for the patient with endotracheal intubation. The risks associated with surgery and anaesthesia were explained to the patient's parents and a detailed written informed consent was taken. A defibrillator and all emergency drugs were kept ready in operating room. Antibiotic prophylaxis was given with Inj. Meropenem 1.6gm and Inj. Linezolid 400 mg in the pre-operative period. The patient was taken in the operation theatre and ASA standard monitoring was attached. The patient was preoxygenated with 100% oxygen for 3 minutes. Inj. Midazolam 1 mg was given as a premedication in aliquots. The patient was induced 100mcg of Inj. Fentanyl (2.5 mcg/kg). The muscle relaxation was achieved with Inj. Rocuronium 40 mg (1mg/kg). Inj. 2% Xylocard 2 ml was given 90 seconds before intubation to blunt the sympathetic response. She was intubated with a size 6.0 cuffed ETT. The patient was maintained on Oxygen:Air mixture with FiO₂ 50% and Sevoflurane (MAC 1) on closed circuit with controlled ventilation. Under all aseptic precautions left radial artery was cannulated with 20G arterial cannula and right sided femoral vein was cannulated with double lumen central venous catheter (5.5 Fr) with extra vigilance about not allowing air escaping into the circulation. IJV was not cannulated keeping in mind the lateral position with head turn given for surgery.

Inj. Rocuronium was given in intermittent doses for muscle relaxation. Inj. Paracetamol 1 gm and Inj. Diclofenac 75 mg were given as part of multimodal analgesia. Ringer Lactate solution was given as IV fluid of choice intraoperatively.

The patient was haemodynamically stable throughout the surgery. The heart rate was maintained between 66-90 beats per min. BP was maintained between 100-120/60-80 mmHg. The oxygen saturation of the patient remained between 88-92% and EtCO₂ was maintained between 30-34 mmHg. The duration of surgery was around 2 hours 45 minutes. At the end of the surgery, the neuromuscular blockade was reversed with Inj. Neostigmine 0.005 mg/kg and Inj. Glycopyrrolate 0.008 mg/kg. The extubation and the recovery of the patient from anesthesia was smooth. The patient was shifted to pediatric ICU for observation and further care. The patient was monitored for any adverse events like hypoxia, arrhythmias and hypotension in the paediatric ICU. She was later discharged from the hospital in stable condition.

Discussion:-

Ebstein's anomaly is a rare congenital anomaly concerning the tricuspid valve and the right ventricle. It arises when there is an displacement of the tricuspid valve toward apex with tethering of attachments of the leaflets. The tricuspid valve and the right ventricle are both dysplastic. It results in impairment in the function of the right ventricle. As a result, there is a decrease in the volume of blood ejected. Due to the presence of right to left shunt, these patients are more at risk for paradoxical embolization and cerebral abscess formation.

It is classified into four grades of increasing severity based on echocardiographic measurement of the ratio of the combined area of the right atrium and atrialized right ventricle to that of the functional right ventricle and left heart in a four-chamber view at end-diastole.(5) In addition, it may be associated with conduction system aberrations due to compression of the atrioventricular node by the septal malformation and disturbance of the atrioventricular node connection.

The goals in anaesthetic management of a patient with Ebstein's anomaly are to maintain sinus rhythm, avoid an increase in preload and afterload. An event of decrease in systemic vascular resistance or increase in pulmonary vascular resistance, leads to a right to left shunt. Tachycardia leads to impairment in right ventricular filling (6). These patients are particularly predisposed to development of supraventricular arrhythmias. Factors known to predispose arrhythmias viz. light plane of anesthesia, fluid or acid base imbalance, hypercapnia and hypoxia are to be avoided. Defibrillation may be required to treat any possible arrhythmias. Measured IV fluids with prudent titration is given to prevent right heart failure.

Inj.Fentanyl 100 mcg was given as induction dose initially for the patient to minimise haemodynamic instability during induction. Later incremental doses of 10mcg was given to maintain analgesia and hemodynamic. A total dose of 150 mcg.was used till the end of surgery. Inj.Rocuronium was the muscle relaxant of choice for its cardiostabilty. EtCO₂ was maintained between 30-34 mmHg to reduce the chance of developing right to left shunt due to increased pulmonary vascular resistance. Arterial line and central venous line were inserted to monitor for sudden changes in blood pressure and better intravenous access to emergency drugs in case of eventuality. Nitrous oxide was not used in maintaining anaesthesia to prevent rise in pulmonary pressures. Adequate analgesia was provided with Inj. Paracetamol and Inj Diclofenac to prevent tachycardia caused due to pain.

Since, these patients are prone to the development of life -threatening arrhythmias (7) hypoxia, hypercapnia, hypovolemia are to be avoided during the surgery. Optimum analgesia and maintainance of adequate depth of anaesthesia is critical to avoid arrhythmias.

Conclusion:-

Patients with Ebstein's anomaly are high risk for surgery and anaesthesia. A co-ordinated team effort among surgeon, anaesthesiologist, cardiologist and intensive care specialist is required to ensure uneventful surgery and satisfying recovery of the patient.

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