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Case Report

# Anaesthetic Management of Ebstein Anomaly in a Patient with Cleft Palate

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**ABSTRACT**

Ebstein anomaly is a challenge to the anaesthetist in terms of large right atrium, atrialised right ventricle and various valve abnormalities associated with it like Tricuspid Regurgitation(TR), Mitral Stenosis(MS), Mitral Regurgitation(MR).
In this case study, we report a 14-year-old female child presented with ebstein anomaly and cleft palate in our institution, SMS medical college & hospitals, Jaipur, Rajasthan. Cone's operation was planned but due to multiple fenestrations in tricuspid leaflet only tricuspid and mitral valves were replaced. Patient was extubated next day with uneventful recovery.

Ebstein Anomaly (EA)is an uncommon congenital anomaly that happens 1 in 20,000 live births and records for less than 1% of all congenital defects [1] yet addresses two-fifth of all congenital malformations of the tricuspid valve [2]. The anomaly is more common in twins and those with a family history of congenital heart disease and maternal benzodiazepenes exposure [3]. Associated anomalies are more frequent in patients with cleft lip and palate (32%) than in patients with cleft lip alone (11%) or patients with cleft palate alone (22%). The organs most commonly involved with associated anomalies in the order of decreasing incidence are eye, ear, heart, upper limb, lower limb, genitals, mandible, mental retardation, craniofacial clefts, skull, tongue, growth retardation, skin and hair. Cardiac anomalies are one of the most common congenital disorders associated in cleft lip and palate patients [4].
Tricuspid valve(TV) leaflet development occurs between 8 and 12 weeks of gestation. The posterior and the septal leaflets are formed by a process of delamination from the underlying myocardium [3]. The morphology includes malformation and displacement of the tricuspid valve leaflets towards the apex of the right ventricle. While the valve annulus remains in the normal position, the attachments of the septal and posterior leaflets are rotationally displaced from the atrioventricular rings, and the anterior leaflet is typically elongated and sail like. Apical displacement of the tricuspid valve creates an "atrialised" portion of the RV that has ventricular morphology but functionally serves as part of the atrium [1]. This results in diminished antegrade blood flow through the right heart, decreased functional RV size, progressive TV regurgitation and right heart dilation [3]. Clinical presentation varies significantly depending on the severity of disease and age at diagnosis. Neonates most commonly presents with cyanosis but may also exhibit signs of right heart failure or arrythmias whereas older children and adults typically present after evaluation for incidental murmur, tachyarrythmias, or right heart failure symptoms [3].

## Case Report

We report a case of Ebstein anomaly along with cleft lip, cleft palate, ectrodactyly and coexisting mitral stenosis (MS) who had undergone surgery for mitral valve and tricuspid valve replacement.

A 13-year-old female child presented to paediatrician with a history of breathlessness, chest pain and

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palpitations. She had undergone previous surgery for bilateral cleft lip at 6 months of age and now has cleft palate (Figure 1). She was also having congenital anomalies of lower limb ectrodactyly (Figure 2). A chest radiograph was advised which showed cardiomegaly-box shaped heart (Figure 3). Hence she was transferred to cardiothoracic surgery department for further treatment.

Preanaesthetic evaluation revealed that height and weight of the patient was 150cm and 30kg (BMI=13.3kg/m2) and has no pallor, cyanosis or jaundice. Airway examination showed complete cleft palate with mallampati grade 2. Systemic examination showed pansystolic murmur at left sternal border.

All blood investigations were within normal limits. ECG was done which showed right atrial enlargement, right axis deviation with an incomplete right bundle branch block and abnormal terminal QRS morphology (Figure 4). 2D ECHO showed grossly enlarged RA, atrialisation of RV cavity, upward displacement of tricuspid leaflet(30mm), severe TR, moderate to severe MR, Ebstein anomaly, IAS/IVS intact, pericardium normal, LVEF 62%. GORE Score (Great Ormond Street Echo score) or Celermajer index was calculated by a ratio of right atrium + atrialised RV by functional RV+ left atrium + left Ventricle volume) which was 1.23 i.e 3rd grade in this case. Tricuspid Annular Systolic Excursion (TAPSE) was 23mm.

A Sternotomy for intra-cardiac repair and TV repair and MV replacement was planned. Before the day of surgery, she was given tab Alprazolam 0.5mg HS. On the day of surgery in preparation room, she was premedicated with inj Morphine 5mg and inj Phenargan 12.5mg IM 45min prior to induction. In the operation theatre, all monitors were attached. One peripheral intravenous line 20G was secured. Preoperatively, the heart rate was 100bpm, blood pressure 90/70 mmHg and Saturation 100% on room air. Ringer lactate drip was started @5ml/hr. Patient was premedicated with inj. midazolam 3mg i/v, inj fentanyl 100mcg i/v. She was preoxygenated with 100% oxygen by face mask for 3-5 min. Induction was done by inj etomidate 10mg i/v and inj rocuronium 30mg i/v. Positive pressure ventilation was done for 90 sec and under direct laryngoscopy, patient was intubated with 6.5 mm cuffed ET tube. Patient had cleft palate but we did not face any difficulty while larynoscopy and intubation. Bilateral air entry was checked and was equal, tube fixed at 19th mark. Tidal volume was kept at 10ml/kg and respiratory rate 16 breaths/min. However, during induction, ECG showed tachyarrythmias and rate was irregularly irregular, we injected inj Esmolol(0.5mg/kg) and inj magnesium 2gm was started in infusion. An arterial line was inserted in the right femoral artery for invasive BP monitoring and arterial blood gas analysis. An ABG was performed preoperatively which showed pH of 7.38, PaO2 464mmHg, PaCO2 32.5mmHg, H2CO3 20.4mmol/L, Na+ 139, K+ 3.74mmol/L, Cl- 109mmol/L,Hb 11.3g/dl, Hct33%. A triple lumen central line was inserted in right Internal jugular vein. Nasogastric tube was inserted. Nasopharyngeal temperature probe was placed in to monitor core body temperature. TEE probe was placed to confirm preop findings and also for intraoperative monitoring and management. Child was cathederised with 12F foley catheder to monitor urine output. Maintainance of anaesthesia by inj vecuronium 1mg i/v, inj midazolam .1mg i/v and inj fentanyl 10mcg iv every 30 min, with fresh gas flow of 100%oxygen @ 2l/min and sevoflurane @ 1-2%.

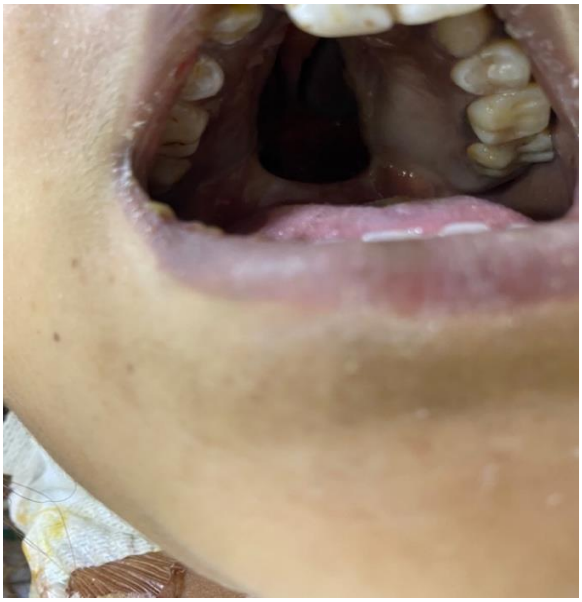

**Figure 1- Cleft palate**

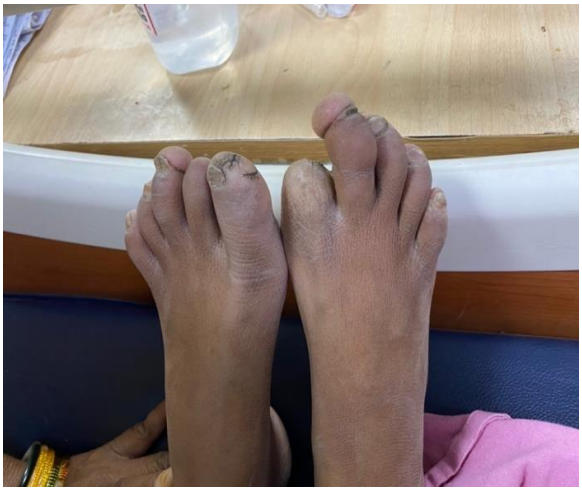

**Figure 2- Ectrodactyly**

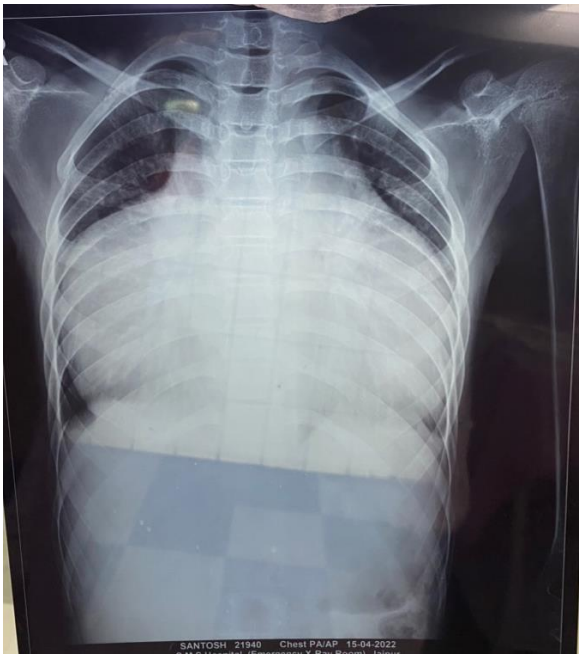


**Figure 3- Skiagram chest PA view**

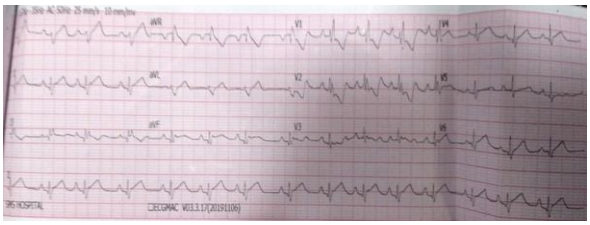

**Figure 4- ECG**

After skin incision midline sternotomy was performed. Inj heparin 120mg was given intravenously. Activated clotting time (ACT) was checked after 5 minutes which was more than 480 sec. Patient was taken on cardiopulmonary bypass machine and the ventilation was stopped. The CPB technique included use of membrane oxygenator (thymus nepro) and moderate hypothermia (28 – 300C). CPB flowrate were maintained between 2.2–2.8litre/meter2/min with mean pressure of 60-80 mm Hg and alpha stat blood gas management were used. Myocardial protection was achieved using anterograde Delnido blood cardioplegia. Meanwhile, serial ABGs and ACT was done by perfusionist team and correction was done accordingly. Intraoperative hydration and vitals were maintained. Cone's operation was tried but due to multiple fenestrations in tricuspid leaflet as there was severe tricuspid regurgitation, it was not possible. Therefore, tricuspid valve replacement (33 St judes mitral valve) and mitral valve replacement (St judes 25 size) was done. After completion of the valve replacement, patient came off bypass machine. Vitals were maintained using ionotropes, ventilation was resumed. Inj protamine sulphate 250mg was given intravenously after the test dose(ACT=130sec). Surgery completed and patient was shifted to cardiac ICU with ET tube in-situ with support of inj dobutamine(250mg in 50ml NS) @ 2ml/hr and inj epinephrine(2mg in 50ml NS)@ 4ml/hr. Shifting vitals-heart rate 120bpm, BP 110/59mmHg, SPO2 100% on IPPV. Total urine output was 2 litres during the surgery.

## Discussion

Several cardiac anomalies are associated with cleft lip and palate, including rare cardiac diseases like Fallot's tetralogy, Ebstein's anomaly, hypoplastic left heart syndrome, and pulmonary atresia [4]. Preoperative prophylaxis for infective endocarditis was started [5].

Usually patients benefit from mild premedication to avoid anxiety induced tachycardia; however concomitant oversedation should be avoided to prevent an increase in PVR [1]. The anaesthetic agents used should not cause significant chronotropic action [6]. Supraventricular tachyarrhythmias and ventricular tachyarrhythmias can occur during induction. In our case we injected inj esmolol .5mg/kg bolus to treat it. so ECG should be carefully monitored [1]. Defibrillators must be ready prior to induction of anaesthesia to end arrhythmias that may be associated with hemodynamic instability. Intravenous beta blockers and amiodarone, may help restore sinus rhythms of clinically stable pre-excited tachycardia. Intravenous adenosine can be used for narrow QRS complex tachycardia [6]. There are several case reports of delayed intravenous induction by pooling of anaesthetics in the large right atrium [1]. Therefore, careful dosing and patience are required to avoid overdose of anaesthetics. The large right atrium also acts as a reservoir to delay the release of drug, prolonging the duration of action [1]. This delay can also result in the delivery of much larger doses of the drug [6].

Patients with fixed cardiac output are very sensitive to the myocardial depressant effect of intravenous induction agents [7]. We have induced our patient with inj Etomidate to avoid decrease in cardiac output as this patient had associated severe mitral stenosis. The hypercapnia can reduce the myocardial contractility and lower the threshold for arrhythmias. In our patient, we maintained the EtCO2 at 28 -32 mmHg by adjusting the ventilator settings to minimize the possibility of hypercapnia [5].

After intubation, access to the artery and central veins was made. TEE helps identify the anatomy of the tricuspid valve, the mechanism of TR, and evaluate biventricular function. After the initiation of the bypass, the surgeon examined the tricuspid valve to determine the feasibility and type of valve repair [1]. It also provides real time information on RV and LV function, TR or stenosis degree and also helps to properly deair the heart before stopping CPB. Immediately after bypass, the relevance of repaired tricuspid valve leaflet function, paravalvular leakage and prosthetic mitral valve function can be assessed. TEE also helps optimize preload, assess

postoperative RV and LV function and guides the use of inotropes [6].

The hemodynamic effects of EA are defined by the TV's functional condition, as well as RV and/or LV function impairment. Increased afterload is poorly tolerated in these patients due to inadequate preload reserve and reduced LV systolic function, which may be exacerbated by the presence of concomitant MS. As a result, any rise in afterload should be avoided at all costs. TR causes RA and RV volume overload, resulting in RA dilation and modest RV functional impairment. When Ebstein's disease is combined with MS, problems such as atrial fibrillation and heart failure arise early. Elevated LA pressure is also transmitted to pulmonary venous system, which lead to early onset of pulmonary arterial hypertension (PAH). PAH causes worsening of TR and early RV failure. Inotropic support and RV afterload reduction are helpful in the immediate post bypass period. Pulmonary vascular resistance should be minimized, and nitric oxide may be indicated in patients with residual TR and depressed RV function. In our patient, we started inj nitroglycerine (1mcg/kg/min) and inj milrinone (0.5mcg/kg/min) and inj adrenaline (0.1mcg/kg/min) infusion. We did not encounter any difficulty in coming off bypass intraoperatively as well as in post operative period.

By avoiding an increase in PVR, we were able to keep RV function. Acidemia, hypoxia, and hypercarbia, which are all reversible causes of elevated PVR, were avoided. In patients with severe pulmonary hypertension, agents that lower PVR, such as nitric oxide, may be useful. Using a pure beta-agonistic inotropic drug, such as dobutamine, can improve PVR, but it also has the potential to cause cardiac arrhythmia, therefore it should be used with caution [6].

To reduce blood loss and the need for transfusions, the anti-fibrinolytic medication traxenamic acid was administered after correct heparin reversal with protamine (1.5 times the total heparin used). To avoid arrythmia exacerbation, electrolytes and acid base gases were closely monitored and rectified as needed [1].

## Conclusion

Thorough preoperative evaluation, judicious intraoperative management and monitored post operative vigilance is mandatory for safe outcome of EA patients.

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