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SURGICAL REPAIR OF EBSTEIN'S ANOMALY- 5 YEARS' EXPERIENCE OF A TERTIARY CARE CENTRE

Cardiothoracis

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ABSTRACT

Aims: The aim of this study is to review our experience with Ebstein's's Anomaly Repair in a tertiary care center over 5 years. **Materials and methods:** This is a retrograde observational study. All patients who underwent Cone Repair from May 2019 to April 2024 were included in the study. **Results:** Total 6 patients underwent Ebstein's repair. The median age of the patients was 15 years with Male: Female ratio 2:3. 3 patients (50%) had Carpentiers type A and 3 patients had Carpentiers type B (50%) of Ebstein's anomaly. Cones repair was done in 5 patients and Danielson's repair in 1 patient. BD Glenn shunt was added in 3 patients. Patients median age at the time of surgery was 11.5 years with 5 paediatric cases and 1 adult case. Mean hospital stay was 6.3±1.2 days. Early re-exploration in 1 patient and mortality in 1 patient (16.7%) **Conclusion:** Ebstein's anomaly is a rare congenital anomaly. Most patients are managed by Cones Repair. The overall post operative outcomes are good especially in paediatric patients.

KEYWORDS

Ebstein's anomaly, surgical repair of Ebstein's anomaly, Cone repair, congenital heart disease.

INTRODUCTION

Ebstein's anomaly is a rare congenital heart defect that affects the structure and function of the tricuspid valve, which separates the right atrium from the right ventricle. It accounts for 0.5% of all congenital heart defects and 0.005% of all live births.[1,2] This condition is characterized by abnormal development of the tricuspid valve, leading to displacement of the valve leaflets into the right ventricle. As a result, the right ventricle becomes partially incorporated into the right atrium, causing a spectrum of physiological abnormalities that can vary widely in severity.

First described by Wilhelm Ebstein's with the autopsy findings of abnormal tricuspid valves in 1866. The patient, Joseph Prescher, was a 19-year-old worker who presented with cyanosis, dyspnoea, palpitations, cardiomegaly, and distended jugular veins.[3] While the exact cause of Ebstein's anomaly is often unknown, it is believed to involve a combination of genetic and environmental factors during foetal development.

In 1993, Da Silva et al.[4] proposed the cone repair. This technique uses principles of bringing the TV leaflets to the true tricuspid annulus level and longitudinal plication of the atrialized RV with advantage of leaflet-to-leaflet coaptation and restoration of RV geometry and function avoiding a prosthetic ring, thus enabling growth and flexibility of the tricuspid annulus[4]

We hereby share a single centre experience of cone repair in Ebstein's Anomaly patient over a period of 5 years

MATERIALS AND METHODS:

Study design

This is a retrospective observational study of 5 consecutive patients with echocardiography confirmed diagnosis of Ebstein's Anomaly and one patient with incidental intraoperative finding of Ebstein's Anomaly presenting to a tertiary care centre who underwent Ebstein's repair from May 2019 to April 2024.

Data collection and Statistical analysis

The patients' demographics, peri-operative data, post-operative course and outcomes were retrospectively obtained from chart review and collected in the electronic database. Data were analysed using SPSS 23.0

Surgical techniques

After placement of interventional monitoring lines under general anaesthesia median sternotomy done for all patients. Right vertical pericardiotomy, pericardial reflection over SVC dissected up to junction of innominate vein with SVC. MPA looped. RPA dissected and looped. Azygous dissected and left patent. Systemic heparinization, Aortobicaval cannulation, cardiopulmonary bypass initiated. Aorta cross clamped. Cold Blood Cardioplegia + Adenosine given through aortic root. Topical Hypothermia and total cardiopulmonary bypass achieved, SVC and IVC snugged. RA opened and findings noted. LV vented through PFO. Anterior, posterior and septal Tricuspid Leaflet mobilized & delaminated. Horizontal plication of atrialized ventricle done (Carpentier). Tricuspid leaflets mobilised & sutured to true annulus. If required Tricuspid annuloplasty done using tricuspid ring. TV tested with saline insufflations for leaflet coaptation. If spo2 is not maintaining Glenn shunt can be done for which PDA dissected out and ligated. SVC divided between clamps. Proximal end of SVC oversewn with 6-0 prolene continuous suture. Arteriotomy done in the superior wall of RPA. SVC anastomosed with RPA in end to side fashion with 7-0 prolene. MPA left intact. Glenn Pressure measured. RA closed with 5-0 Prolene. Aortic root vented, heart deaired, Aortic cross clamp removed. CPB weaned off using inotrope. Heparin reversed with Protamine. Hemostasis achieved. Sternum closed with ethibomd no 2 after putting drains.

RESULTS

Patient demographics

From May 2019 to Apr 2023, 6 patients with a diagnosis of Ebstein's Anomaly were referred to our CTVS department. Out of 6 patients, 2 were male and 4 were female with median age was 11.5 years. 5 Patients belonged to pediatric age group and 1 patient was adult of age 45years.

Diagnosis was made preoperatively in 5 patients on echocardiography. One patient was incidentally diagnosed intraoperatively while operating for VSD + PS. Three patients had associated ASD, one patient had VSD and one patient with preop diagnosis of VSD+PS was incidentally found to have Ebstein's anomaly.

On echocardiography in three patients TR was mild, moderate TR was in three patients and severe TR was present in 1 patient. On evaluation

for Severity of RV dysfunction four patients had mild, 1 had moderate and 1 had severe RV dysfunction preoperatively. (Table 1)

Table 1: Preoperative findings

Variables	Total , n=6(%)
Male: female	1:2
Median age (years)	20.5±14.9
Type of associated anomaly ASD VSD VSD+ PS	 3 (50%) 1 (16.7%) 1 (16.7%)
Severity of TR Mild Moderate Severe	 1 (16.7%) 3 (50%) 2 (33.3%)
Severity of RV systolic dysfunction Mild Moderate Severe	 4 (66.7%) 1 (16.7%) 1 (16.7%)

Intra operative and postoperative findings

Cone repair was done in 1 patient, cone repair with BD Glenn was done in 3 patients, cone repair with VSD closure was done in 1 patient and 1 patient underwent trans RA VSD closure with RVOT resection with Danielson's technique for Ebstein's anomaly. Tricuspid annuloplasty using tricuspid ring was done in 3 patients.

Mean CPB time was 242 ± 88.7 minutes. Mean aortic cross clamp time was 166.5±59.2 minutes. Postoperative TEE revealed no TR in 1 patient, moderate TR in 1 patient and mild TR in 4 patients.2 patients had normal sinus rhythm post operatively and 3 patients had paced rhythm. 1 patient had normal sinus rhythm initially but on POD 4 patient started having ventricular arrythmias. The patient was reexplored in view of recurrent ventricular fibrillation and right ventricular systolic dysfunction and was put on ECMO. No patient underwent permanent pacemaker implantation. Median intubation time was 18 hours. 1 patient who was re-explored and put on ECMO expired on POD 6. Other than this there was no early in hospital mortality. The mean duration of the hospital stay was 6.3±1.2 days.

Table 2: Intraoperative and post operative findings

Variables	Total, n=6(%)
Surgeries Cone repair Cone repair +BD Glenn shunt Cone repair +VSD closure Trans RA VSD closure +RVOT resection + Danielson's repair	 1(16.7%) 3(50%) 1(16.7%) 1 (16.7%)
Tricuspid annuloplasty	3(50%)
Mean CPB time	242±88.7 minutes
Mean ACC time	166.5±59.2 minutes
Post- operative TR No Mild Moderate Severe	 1(16.7%) 4(66.7%) 1(16.7%) 0
Rexploration	1(16.7%)
Median intubation time	18 hours
Mean hospital stay	6.3±1.2 days
Mortality	1(16.7%)

DISCUSSION

Ebstein's anomaly of the tricuspid valve is a cardiac malformation characterized by downward displacement of the septal and inferior tricuspid valve (TV) leaflets, redundant anterior leaflets with a sail-like morphology, dilation of the true right atrioventricular annulus, TV regurgitation, and dilation of the right atrium and ventricle. This malformation is thought to be due to defects in process of "delamination from the underlying myocardium" [5] during development of the tricuspid valve. There is wide variety of anatomic and pathophysiologic presentations of Ebstein's anomaly which resulted in development of different methods of surgical repair.

The clinical presentation of Ebstein's anomaly can range from asymptomatic cases discovered incidentally to severe cases presenting with significant cardiovascular compromise shortly after birth. Common symptoms include cyanosis (blue discoloration of the skin), shortness of breath, fatigue, palpitations, and heart murmurs. The severity of symptoms largely depends on the degree of tricuspid valve displacement and associated cardiac abnormalities.

Surgical treatment of Ebstein's anomaly was firstly reported by Hunter and Lillehei in 1958 but initial reconstructions were associated with high incidence of tricuspid dysfunction and tricuspid valve replacement.[6] In 1988, Carpentier et al,[7] described a technique in which longitudinal plication of RV and returning of TV to the correct level with reinforcement with a prosthetic ring was done. Danielson et al.[8] proposed a transverse plication of the atrialized RV, posterior tricuspid annuloplasty, and right reduction atrioplasty. Da Silva et al. developed a surgical technique in 1993, where cone reconstruction of tricuspid valve is done. In this repair, also sometimes known as cone repair, most of the anatomic TV defects that occurred during embryologic development are undone and a cone-like structure is created from the available leaflet tissue resulting in almost normal TV anatomy[4]

In our study, 5 patients underwent cones repair and one patient underwent Danielson procedure. BD Glenn was done in 3 patients in view of moderate RV dysfunction in one patient, falling post CPB spo2 in 1 patient and post operative ventricular arrythmias with RV dysfunction in 1 patient. Preoperatively three patients had mild TR, moderate TR was in three patients and severe TR was present in 1 patient. This improved in immediate postoperative transoesophageal echocardiography to no TR in 1 patient, moderate TR in 1 patient and mild TR in 4 patients

Median intubation time in our study was 18 hours. 1 patient was re-explored and put on ECMO in view of severe right ventricular dysfunction but later expired on POD 6. Other than this there was no early in hospital mortality.

The limitations of this study are its retrospective design and limited sample size. Larger sample size and more detailed analyses are required to support our results.

Total mortality in the current study was 16.7% in the first 30 postoperative days, and there were no late deaths. In a cohort of 539 patients, Brown et al.[9] reported total mortality of 29% in a 20-year follow-up

CONCLUSION

Ebstein's anomaly is a rare congenital anomaly. Most patients are managed by Cones Repair. The overall post operative outcomes are good especially in paediatric patients.

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