



EBSTEIN'S ANOMALY – OUR EARLY AND MID-TERM RESULTS WITH CONE REPAIR.

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ABSTRACT

Introduction: Ebstein's anomaly is a rare disease, comprising less than 1% of all cases of congenital heart disease. Da Silva et al, in 2004, described cone reconstruction of tricuspid valve, which can restore the tricuspid valve anatomy. This repair technique has shown promising early and intermediate results. We present our experience with cone repair done over a period of 6 years at the Department of Cardiothoracic and Vascular Surgery, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi. **Material and Methods:** 7 patients of Ebstein's anomaly have been treated with cone repair in our hospital from 2017 to 2022. This was a retrospective study, where hospital records of all these patients were retrieved and studied. Patients were either traced from routine OPD follow-up or telephonically. Pre-operative, post-operative and follow-up parameters were analysed using various statistical tools. **Results:** Our study demonstrated good functional outcome after cone repair. There was significant improvement in tricuspid regurgitation ($p = 0.0003$), transannular plane systolic excursion ($p = 0.007$) and New York Heart Association functional class ($p = 0.004$). **Conclusion:** Cone repair for Ebstein's anomaly provides good short and mid-term outcome in experienced hands. The low morbidity and mortality associated with cone repair, in addition to improvement in RV function and TR makes it the procedure of choice for Ebstein's anomaly, irrespective of the anatomical type. However, long term studies with larger cohorts can provide better insight into the outcome of this novel procedure.

KEYWORDS : cardiac surgery, Ebstein's anomaly, cone repair, tricuspid ring annuloplasty, 1.5 ventricle repair

INTRODUCTION:

Ebstein's anomaly is a rare disease, comprising less than 1% of all cases of congenital heart disease¹. It is a congenital defect of the tricuspid valve in which origins of septal or posterior leaflets or both are displaced downwards into the right ventricle with variable degree of leaflet deformity. It is associated with right ventricle dysplasia with wide spectrum of severity^{1,2}. Normal development of heart includes the process of delamination, by which leaflets of the tricuspid valve split and detach from the myocardium¹. Ebstein's anomaly results from the failure of this process, thus resulting in adherence of septal and posterior leaflets to the underlying myocardium with downward displacement of the functional tricuspid annulus. This further leads to creation of atrialized ventricle – thin walled, non-contractile and dyskinetic part of right ventricle that lies above the functional annulus. Ebstein's anomaly is commonly associated with atrial septal defect or patent foramen ovale (80–90%), pulmonary valve stenosis or pulmonary atresia, ventricular septal defect (VSD), and patent ductus arteriosus (PDA)^{3,4}. Patients may present during the first week of life with breathlessness, cyanosis, severe cardiomegaly and often heart failure. However, many patients do not show any symptoms until later in childhood, or even adulthood. The symptoms include exertional dyspnea, fatigue, cyanosis (sometimes just exertional), and palpitations.

History of Ebstein's anomaly dates back to 1866, when Wilhelm Ebstein described this anomaly from an autopsy specimen of tricuspid valve. From palliation to corrective interventions, many surgical procedures have been described for Ebstein's anomaly. Da Silva et al.⁵, in 2004, described cone reconstruction of tricuspid valve, which can restore the tricuspid valve anatomy. This repair technique has shown promising early and intermediate results⁵.

We, hereby, present our experience with cone repair done over a period

of 6 years at the Department of Cardiothoracic and Vascular Surgery, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi.

MATERIAL AND METHODS:

From 2017 to 2022, 7 patients of Ebstein's anomaly have been treated at the department of cardiothoracic and vascular surgery, VMMC and Safdarjung Hospital, New Delhi. All these patients have undergone cone reconstruction of the tricuspid valve. This was a retrospective study, where hospital records of all these patients were retrieved and studied. Patients were either traced from routine OPD follow-up or telephonically from the mobile numbers provided in the admission papers. Transthoracic echocardiography was done in all patients on last follow-up and patients were clinically assessed using New York Heart Association (NYHA) functional class. All patients were followed-up till October 2022 and no patient was lost to follow-up.

Surgical Procedure:

All surgeries were performed via median sternotomy using conventional cardiopulmonary bypass and mild hypothermia after establishing all monitoring lines. Intraoperative trans-oesophageal echocardiography was performed before initiation of cardiopulmonary bypass. After establishment of cardiopulmonary bypass, superior and inferior cavae were looped, aorta cross clamped, antegrade root cardioplegia given and cardioplegic arrest achieved. Right atriotomy was done after snugging cavae, followed by the assessment of tricuspid valve. Anterior, posterior and septal leaflets were detached from the annulus and delamination of anterior leaflet done. The neo-tricuspid annulus was constructed by plicating the grossly dilated atrialised right ventricle, vertically, with interrupted 4-0 pledgeted prolene sutures and Kay's technique (plication of posterior tricuspid annulus)⁶. Anterior, posterior and septal leaflets sutured with neo-tricuspid annulus with 6-0 prolene to complete the cone reconstruction of the tricuspid valve. One patient with type-C Ebstein's

anomaly underwent bidirectional Glenn procedure in addition to cone repair (1.5 ventricle repair). A pericardial patch was used between septal and posterior leaflet to augment the valve in one case of deficient posterior and septal leaflets. Tricuspid annuloplasty ring was deployed in one case with grossly deformed annular anatomy. Patent foramen ovale (PFO) was not closed in one case. Atrial septal defects (ASD), were closed in all respective cases. This was followed by rewarming, right atrial closure, de-airing and weaning off from cardiopulmonary bypass. Post bypass trans-oesophageal echocardiography done to assess the status of repair, followed by sequential decannulation. Adequate hemostasis was achieved, drains placed and wound closure done in standard manner.

Statistical Analysis:

The analysis included profiling of patients on different demographic, clinical and laboratory parameters. Descriptive analysis of quantitative parameters were expressed as means and standard deviation. Categorical data were expressed as absolute number and percentage. Independent Student t-test and One-way ANNOVA were used for testing of mean between independent groups. A *p* < 0.05 was considered statistically significant. All analysis was done using SPSS software, version 24.0.

Results:

Table-1: Preoperative characteristics of the study group

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7
Age (years)	11	18	23	10	5	12	15
Gender	M	M	F	M	M	M	M
Height (cm)	135	162	148	130	104	145	180
Weight (Kg)	40	52	53	28	13	40	55
Body Surface Area (m2)	1.21	1.54	1.45	1.01	0.61	1.27	1.66
Carpentier Type	B	C	B	B	B	B	B
Associated Condition	PFO	Nil	ASD	Nil	ASD	ASD	ASD
Associated Arrhythmia	Nil	Nil	Nil	JET	AVRT	Nil	VT

Table-1 shows pre-operative characteristics of all the patients. Mean age of the patients was 13.4 ± 5 years (range: 5-23). Out of 7 patients, only 1 patient was female, rest were males. Average height of the patients was 143.4 ± 24 cm, average weight of patients was 40.1 ± 15 Kg and average body surface area was 1.25 ± 0.4 m2. 1 patient had Carpentier type-C Ebstein anomaly and rest of the patients had type B Ebstein's Anomaly. 4 patients had an associated secundum type ASD, 1 patient had PFO, and 2 patients had no other associated cardiac defect. 4 out of 7 patients had normal sinus rhythm, 1 patient had junctional tachycardia (managed medically), 1 patient had Wolff-Parkinson-White (WPW) Syndrome with atrio-ventricular re-entry tachycardia (AVRT) which was managed medically and 1 patient had recurrent monomorphic peri-tricuspid ventricular tachycardia for which pre-operative catheter ablation was done after EPS (electro-physiological study) and electroanatomic mapping. Average bypass duration was 136.7 ± 17 minutes and average cross-clamp time was 88 ± 25 minutes. Mean ICU stay of all patients was 6.7 ± 3 days. Mean post-operative hospital stay was 11.7 ± 7 days. Table-2 gives an overview of the intra-operative and post-operative variables.

Table-2: Intra-operative and immediate post-operative variables

Mean CPB duration	136.7 ± 17 minutes
Mean ACC duration	88 ± 25 minutes
Mean ICU stay	6.7 ± 3 days
Mean Hospital Stay	11.7 ± 7 days

Post-surgery, patients showed improvement in clinical and echocardiographic parameters. Patients showed significant improvement in NYHA functional class from pre-operative mean of 2.4 ± 0.5 to 1.3 ± 0.7 at the last follow-up (*p* = 0.03). Similarly, there was improvement in tricuspid regurgitation which was statistically significant (*p* < 0.001). Mean preoperative tricuspid regurgitation was 2.6 ± 0.8. Post-operative echocardiography, usually done during the ICU stay, showed a mean tricuspid regurgitation of 0.6 ± 0.5. Mean tricuspid regurgitation was 1.1 ± 0.9 at the last follow-up. TAPSE (trans-annular plane systolic excursion) was calculated before surgery and during the last follow-up. It also showed significant improvement (*p* = 0.007) from pre-operative TAPSE of 12.1 ± 2 to 14 ± 2 at the last follow-up. Table-3 shows the impact of surgery on functional and echocardiographic variables.

Table-3: Comparison between pre-operative and post-operative variables

	Pre-operative	Post-operative	Follow-up	p
NYHA	2.4 ± 0.5	-	1.1 ± 0.4	0.004
TR	2.6 ± 0.8	0.6 ± 0.5	1.1 ± 0.9	0.0003
TAPSE	12.1 ± 2	-	14 ± 2	0.007

DISCUSSION:

Ebstein's anomaly of the tricuspid valve has been treated with various surgical techniques since 1958 7, 8. Despite high morbidity and mortality, valve replacement was the treatment of choice till 1970s 9. In 1988, Carpentier et al described anatomical classification of Ebstein's anomaly and described a new surgical technique, consisting of right ventricular reconstruction, repositioning of tricuspid valve at anatomical annulus and routine use of annuloplasty ring 10. Quagebeur et al followed Carpentier's technique, but without a prosthetic ring in younger population 11. Cone reconstruction of tricuspid valve was first described by da Silva in 2004 5. It has become the gold standard surgical treatment for Ebstein's anomaly, providing the best medium- and long-term results 12. Various modifications of cone repair have been performed due to variations in right ventricular atrialization and coronary anatomy with good results 13.

At Vardhman Mahavir Medical College and Safdarjung Hospital, cone repair program was started in 2017 and till October 2022, 7 patients of Ebstein's anomaly have successfully undergone cone repair. The aim of this study is to present our short and mid-term outcome of cone repair for this rare disease.

The period starting with the weaning off from cardiopulmonary bypass (CPB) to immediate post-operative period is considered very crucial after cone repair in view of high risk of right ventricular failure owing to non-competent tricuspid valve. Many groups have used bi-directional Glenn to off-load the right ventricle in patients facing right ventricular (RV) failure 14-16. Extracorporeal Membrane Oxygenation (ECMO) may also be used to bridge this critical period. Residual inter-atrial communication may help prevent RV failure as described by Attenhofer et al 17. In our series, one patient had a PFO, which was not closed owing to poor RV function. Rest of all the encountered defects were closed without any dire consequences.

Da Silva et al in his original article observed that his technique efficiently corrected tricuspid insufficiency and restored right ventricular morphology 5. Xiao Li et al observed in their study that cone repair restores the RV geometry and function. It also eliminates TR and enables growth of the annulus if prosthetic ring is not used 18. However, Cine N et al reported that tricuspid ring annuloplasty, when used to modify cone-type repairs, preserves valve coaptation and function in their early and mid-term results, and has beneficial effect on right ventricle remodelling, especially in adults and adolescent patients. In our series, ring annuloplasty was performed in an adult patient with grossly deformed annular anatomy. Many studies, including Da Silva et al 5, Xiao Li et al 18 and a large study of 235 cases by Holst KA et al 19 have described improvement in TR which was also reflected in our series. Although, all patients had mild or no TR in the immediate post-operative period, 1 patient showed deterioration of TR on follow-up. Exercise tolerance is one of the best indicators of success. In our series, increased exercise tolerance was observed in all the patients with all patients in NYHA functional class I or II. Similarly, RV function, as estimated by TAPSE, showed significant improvement in our series. Incidence of cardiac arrhythmias is high in Ebstein's anomaly after surgery. In our series, 1 patient developed AVRT which settled with medical therapy. Another patient developed Junctional Ectopic Tachycardia (JET), which again settled with medical therapy. As described earlier, 1 patient in our series had recurrent VT in pre-operative period. He underwent EPS, followed by mapping and catheter ablation but he again developed pulseless, polymorphic VT in the immediate post-operative period and required open cardiac massage for revival. Cardiology team was involved for the management of recurrent VT but patient settled with medical therapy only. 1 patient was re-explored on zero post-operative day in view of high drain output. Rest of the 3 patients had uneventful post-operative period. 2 patients in our series developed pericardial effusion during hospital stay and were managed with tube drainage.

CONCLUSION:

Cone repair for Ebstein's anomaly with certain modifications depending on intra-operative findings provides good short and mid-

term outcome in experienced hands. The low morbidity and mortality associated with cone repair, in addition to improvement in RV function and TR makes it the procedure of choice for Ebstein's anomaly, irrespective of the anatomical type. However, long term studies with larger cohorts can provide better insight into the outcome of this novel procedure.

Declarations:

Funding: None

Conflict of interest: None declared

Ethical approval: Not required

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