

## THE CONE RECONSTRUCTION OF EBSTEIN ANOMALY: A CASE REPORT

Khuzaima Tariq<sup>1</sup>, Taimur Asif Ali<sup>2</sup>, Parveen Akhtar<sup>3</sup>, Arif ur Rehman<sup>4</sup>

Department of Adult Cardiac Surgery,  
National Institute of Cardiovascular  
Diseases (NICVD), Karachi - Pakistan

Address for Correspondence:

**Khuzaima Tariq**

Department of Adult Cardiac Surgery,  
National Institute of Cardiovascular  
Diseases (NICVD), Karachi - Pakistan

Emails: khuzaimatariq@gmail.com

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### Contribution

KT conceived the idea, planned the case report and helped in final draft. TAA and AUR helped with procedure and PA helped with CT report. All authors contributed significantly to submitted manuscript.

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### ABSTRACT

Ebstein anomaly is a rare congenital defect in which origins of variably deformed posterior and septal leaflets of tricuspid valve are displaced downwards into right ventricle (failure of delamination, with characteristic sail-like anterior leaflet. Rotational displacement of tricuspid valve divides the right ventricle into proximal (atrialized) and distal (ventricularized) portions. Due to marked dilation of true annulus and right ventricle, ebstein valves are usually regurgitant. Out of numerous repair techniques, Da Silva's Cone reconstruction is different in that it is closest to anatomical repair in which there is mobilization of anterior and posterior tricuspid leaflets from their anomalous attachments in the right ventricle, with clockwise rotation of the free edge and sutured to the septal border of anterior leaflet. The basic principle of this repair is to create a cone with its tip fixed at the right ventricular apex and the base sutured to the true tricuspid valve annulus level. We present a case of Ebstein anomaly in which Cone repair was done with uneventful recovery.

**Key Words:** Tricuspid valve; Ebstein anomaly; Cone procedure.

### INTRODUCTION

Ebstein anomaly (EA) is a rare congenital malformation of tricuspid valve (TV) and right ventricle (RV) that exhibits an infinite spectrum of malformation. Normal Tricuspid valve function depends on interactions among fibrous annulus, 3 leaflets (anterior, posterior, septal), papillary muscles, chordae tendinae and adjacent myocardium. In EA origins of variably deformed septal and posterior leaflets of TV are displaced into RV. Characteristically enlarged anterior leaflet is "sail like"<sup>1</sup>. Rotational displacement of TV divides RV into proximal (atrialized) and distal (ventricularized) portions<sup>2</sup>. There have been multiple surgical repair techniques proposed given the variable spectrum of anatomy in Ebstein's hearts including Danielson repair, Dearani technique, Sebening, Henley's approach, Carpentier repair etc. Out of numerous repair techniques, Da Silva's Cone reconstruction is different in that it is almost near to anatomical repair. Moreover it can be applied to wide variety of anatomic variations encountered in Ebsteins Anomaly. In this repair, there is mobilization of anterior and posterior tricuspid leaflets from their anomalous attachments in the right ventricle, with clockwise rotation of the free edge and suturing it to the septal border of anterior leaflet. The

basic principle of this repair is to create a cone with its tip fixed at the right ventricular apex and the base sutured to the true tricuspid valve annulus level.

## CASE PRESENTATION

We present a case of forty four years old male, presented in cardiology outpatient department with complaints of palpitations and shortness of breath NYHA II, on and off for past twenty years. He used to take medications for symptomatic relief. Lately he started having brief episodes of unconsciousness for past 2-3 years. On clinical exam a murmur was heard in right parasternal region. His laboratory values were normal. Chest x-ray showed cardiomegaly and rounded cardiac contour (Figure 1).

Transthoracic echocardiograph showed apical displacement of one of the three tricuspid leaflets, enlarged right atrium (RA) and RV with dysfunction and a large sized atrial septal defect (ASD). Transesophageal Echocardiograph yielded size of atrialisation to be 15 mm. Computed Tomography showed dilated RA with apical displacement of anterior and septal leaflet of TV (Figure 2).

With impression of EA this patient was referred for surgical management. Da Silva's Cone procedure was considered. After median sternotomy, standard cardiopulmonary bypass technique was instituted. TV approached through oblique right atriotomy. Intraoperative findings were enlarged RA, transposed atrioventricular groove towards RV, large right coronary artery, normal sized left ventricle, anterior and septal leaflet of tricuspid valve displaced, Large atrialised part of RV upto annulus superiorly, Large fenestrated ASD. The anomalous TV leaflet is mobilized from its abnormal attachments in the RV and the free edge is rotated

clockwise and sutured to the septal border of anterior leaflet. Plication of atrialised RV done with 2/0 pledgeted ethibond. ASD closed primarily. Patient smoothly weaned off cardiopulmonary bypass and had uneventful recovery in ICU. He remained event free during his hospital course and was discharged after two weeks.

Follow up: On follow up there was marked improvement in patient's clinical status. Echocardiograph showed well functioning Tricuspid Valve.

## DISCUSSION

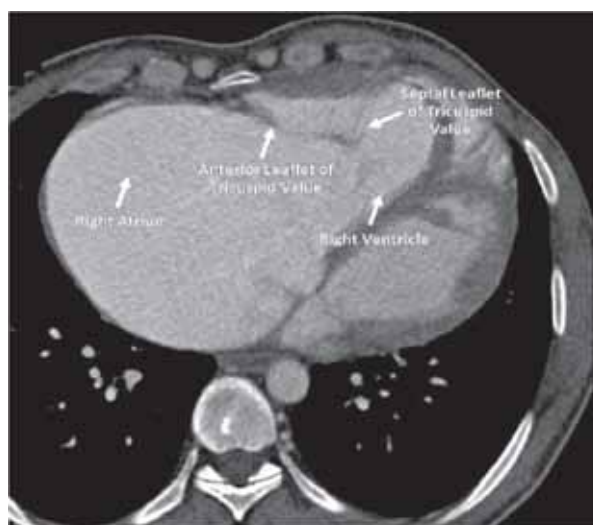
EA is uncommon with a prevalence of 0.5% having equal incidence in both sexes. The primary pathophysiologic features that predominate are RV abnormalities, TV malformation and accessory conduction pathways. Both origin of TV from the atrioventricular (AV) ring and its chordal attachments within RV are malpositioned, with malformed leaflets.<sup>3,4</sup> The septal leaflet appears always to be affected, the posterior leaflet nearly always, and anterior leaflet seldom. Rotational displacement of valve divides RV into proximal (atrialized) and distal (ventricularized) portions (Figure 3). An ASD is present in 80 to 95% of patients.<sup>5</sup>

Over time multiple repair techniques like Danielson, Dearani, Sebening, Henley's approach have been proposed, Cone reconstruction described by Dr. da Silva et al has revolutionized the surgical approach to these patients. It bears similarities to Carpentier's technique but differs in extent of mobilization and rotation of leaflet apparatus.<sup>6,7</sup> In this procedure, posterior and anterior leaflets are detached from annulus as a single unit, after mobilization from their anomalous attachments in RV, this unit is rotated in a

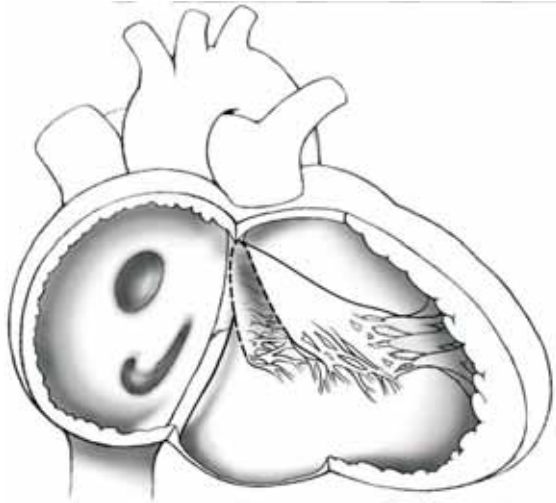
**Figure 1: Chest X-Ray Showing Cardiomegaly and Rounded Cardiac Contour**



**Figure 2: Computed Tomography Showed Dilated Right Atrium with Apical Displacement of Anterior and Septal Leaflet of Tricuspid Valve**



**Figure 3: Preoperative Demonstration of the Displaced Tricuspid Valve and Atrialized Right Ventricle in Ebstein's Anomaly**



clockwise fashion and sutured to septal border of anterior leaflet. The anatomic annulus is plicated with sutures at its true annulus (atrioventricular junction) level so the hinge point of valve is now in a normal anatomic location. The end result of cone reconstruction includes 360° of tricuspid leaflet tissue surrounding the true tricuspid annulus at right atrioventricular junction (Figure 4).

This repair incorporates a longitudinal plication of atrialized portion so any areas of RV dyskinesia are eliminated. ASD closure is done in a valved fashion.

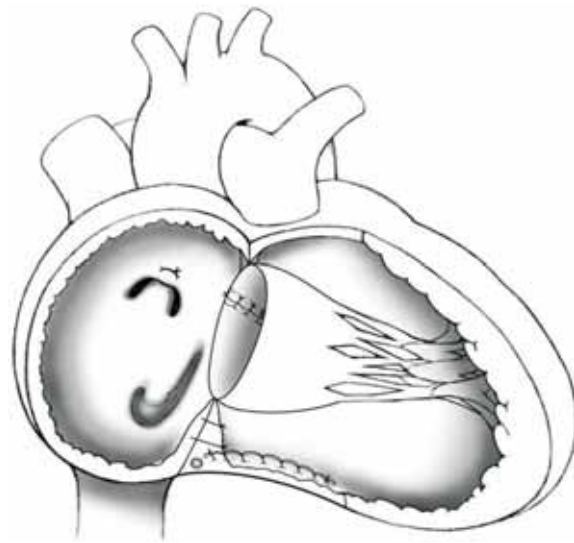
## CONCLUSION

EA is a rare congenital cardiac malformation with a vast spectrum of anatomic and pathophysiological presentation. The significance of extensive mobilization of displaced and malformed tricuspid leaflets to allow better leaflet-to-leaflet coaptation and right ventricular functional area restoration, precluding conventional valve replacements after cone reconstruction is highlighted. Early referral for surgical evaluation prior to deleterious effects of chronic right ventricular volume overload is recommended.

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**Figure 4: The Completed Cone Reconstruction of the Tricuspid Valve for Ebstein's**



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