



BIOPROSTHETIC VALVE REPLACEMENT IN EBSTEIN'S ANOMALY: OUR EXPERIENCE OF 5 YEARS

Cardiology

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ABSTRACT

Background: Tricuspid valve repair has become the main modality for treatment in Ebstein's anomaly and numerous methods of valve repair are now practised. However, Tricuspid valve replacement has still maintained its place in the management of Ebstein's anomaly. Also, it is the chief modality when the tricuspid valve cannot be repaired for any specific reason.

Materials and Methods: The aim of our study was to review our experience with bioprosthetic valve replacement in patients of Ebstein's anomaly. This was a retrospective study and we could find 12 patients who had undergone the same procedure over a period of 5 years. There was no early or late mortality in our study.

Conclusion: Tricuspid valve replacement with a bioprosthetic valve is a simple procedure with good post operative results in patients of Ebstein's anomaly.

KEYWORDS

Ebstein's, Tricuspid, Replacement, Bioprosthetic

Introduction

Ebstein's anomaly of the tricuspid valve is a rare congenital cardiac anomaly described in less than 1 percent of congenital heart diseases. [1] Description of this lesion was first done by Wilhelm Ebstein in 1866 and this disease came to be known as Ebstein's anomaly in 1927. Since the first description of this lesion, numerous advances have occurred regarding knowledge of its pathophysiology, etiology, echocardiographic features and its surgical management. Valvular reconstruction was first suggested by Hunter and Lillehei and further application and development of this procedure was done by Hardy and his colleagues. The first successful treatment of Ebstein's anomaly with a prosthetic valve insertion was done by Barnard and Schrine in 1963. [2] Today, most of the modalities of treatment of Ebstein's anomaly are directed towards preservation and repair of the tricuspid valve due to problems associated with prosthetic valves. However successful repair thrives on a good repair program and a good transesophageal echocardiography (TEE) backup. [3-6] As a result, bioprosthetic valve insertion at the tricuspid position is still practised today. The purpose of our study was to retrospectively assess our experience with bioprosthetic tricuspid valve replacement in Ebstein's anomaly patient.

Materials and Methods

This study was a retrospective study and we reviewed our data for the last 5 years from January 2015 to January 2020 and we could find 12 patients who had undergone bioprosthetic tricuspid valve replacement at our institution during the same period. We restricted our search to patients who had atrioventricular (AV) and ventriculo-arterial (VA) concordance and did not have any other additional structural anomaly. Surgery

The operative strategy for our patients was median sternotomy, aortobicaaval cannulation and moderate hypothermia (28 to 32 °C). Cardiac arrest was achieved by cold potassium rich del Nido cardioplegia with topical hypothermia. Repeat doses of cardioplegia, if required were given at 60 minutes of cross clamp time. Our plan was tricuspid valve replacement first followed by atrial septal defect (ASD) closure. After achieving cardiac arrest, the right atrium was opened through an incision parallel to the atrioventricular groove. We inspected to look for any other associated abnormalities or those missed by transthoracic echo. Any tricuspid valve tissue which would in future obstruct the new tricuspid valve orifice was excised. Pledged non absorbable mattress sutures were placed around the residual tricuspid annulus from the atrial to ventricular side such that the pledget stayed on the atrial side. This was done at the anterior and posterior leaflets. The suture line deviated away from the coronary sinus and towards the septum at the position of the septal tricuspid leaflet. Sutures were passed through the bioprosthetic valve and tied down. Plication of redundant right ventricle would be done along with suturing. After the valve was in place and its movements checked, the ASD was closed with a pericardial patch. Rewarming, weaning and termination of cardiopulmonary bypass followed.

Patients underwent a 2 D echo before discharge. During follow up the

patients had been undergoing a yearly 2d echo apart from the routine physical examination.

Results

Between January 2015 and December 2020, we could find 12 patients of Ebstein's anomaly who had undergone a tricuspid bioprosthetic valve insertion. Of these 8 patients were female and 4 patients were male. The mean age was 24.91 ± 6.28 years (range 16–35 yrs). The indications for surgery were NYHA class III- IV or increasing exercise intolerance in our patients.

Most of our patients (9 out of 12) were in NYHA class III while the remainder were NYHA class IV. 2 patients had cyanosis with an oxygen saturation of 84 and 82 percent respectively. 3 patients had atrial fibrillation. 1 patient had ventricular pre-excitation on electrocardiogram and underwent pre operative Radio Frequency Ablation by our cardiologists before being referred for surgery. Our pre operative data is summarised in table 1.

Table 1: Pre operative parameters

Pre operative characteristics	No of patients
Females	8 (66.66%)
Age	24.91 ± 6.28 yrs
NYHA class (mean)	3.6
Class 1	0
Class 2	0
Class III	9
Class IV	3
Cyanosis	2
Atrial Fibrillation	3
Ventricular Pre excitation	1
Cardio thoracic ratio	69.11 ± 5.1 %

Operative data

All patients underwent Bioprosthetic tricuspid valve replacement. Our mean cardiopulmonary bypass time was 121 ± 12.83 minutes and our aortic cross clamp time was 68 ± 4.88 minutes. We implanted Edwards Perimount 31 in 4 patients, St Jude Medical Epic 31 and Biocor 31 in 3 and 5 patients respectively. Our operative data is summarised in table 2.

Table 2: Operative and post operative data

Operative and post op parameters	
CPB time	121 ± 12.83 min
Aortic cross clamp time	68 ± 4.88 min
Valve size	31
ICU stay	3.08 ± 0.51 days
Post operative stay	7.91 ± 1.78 days
Hospital deaths	0

1 patient underwent immediate re exploration in view of excessive mediastinal drainage in the first 3 hours of surgery. The cause of the bleeding was an internal mammary bleeder. One patient developed

post operative wound infection and underwent wound debridement and re-suturing. This patient had a prolonged post operative stay of 13 days as compared to our mean stay of 7.91 ± 1.78 days for the other 11 patients. The ICU stay for our patients was 3.08 ± 0.51 days. There was no in-hospital mortality. All patients were in NYHA class I (10 out of 12) or NYHA class II (2 out of 12) at discharge. We did an echocardiographic evaluation for all patients before discharge which revealed good valve function, no intra or para valvular leak. These patients were put on Warfarin for the first 3 months after which we discontinued the anti-coagulant.

Follow up.

Although our follow up was just between 3 months and 5 years, all patients were healthy and following up regularly. There were no deaths during the follow up period. All patients were either in NYHA class I or II on follow up. There was no bioprosthetic valve degeneration. Few patients had complaints of occasional palpitations; however most of them were in sinus rhythm. There was no incidence of valve endocarditis during follow up or any thrombo embolic complications.

Discussion

Ebstein's anomaly is a rare congenital heart disorder occurring in 1 per 2,00,000 live births and accounts for <1% of all cases of congenital heart disease.[6] It is a malformation of the tricuspid valve and the right ventricle and is characterised by (1) adherence of the septal and posterior leaflets to the underlying myocardium (2) downward displacement of the functional annulus (3) dilatation of the atrialized part of the right ventricle (4) redundancy, fenestrations and tethering of the anterior leaflet and (5) dilatation of the right atrioventricular junction. [7,8] The cardinal symptoms in Ebstein's anomaly are cyanosis, right-sided heart failure, arrhythmias, and sudden cardiac death. The hemodynamic variations and clinical presentation depend on age at presentation, anatomic severity, hemodynamics, and degree of right-to-left interatrial shunting. [9] Children >10 years of age and adults often present with arrhythmias. Adults also present with progressive cyanosis, decreasing exercise tolerance, fatigue, or right-sided heart failure. In the presence of an interatrial communication, the risk of paradoxical embolization, brain abscess, and sudden death increases. Exercise tolerance is dependent on heart size and oxygen saturation.

The downward displacement of the septal leaflet of the tricuspid valve is associated with discontinuity of the central fibrous body and septal atrioventricular ring with direct muscular connections, thus creating a potential substrate for accessory atrioventricular connections and pre-excitation. From 6% to 36% of patients with Ebstein's anomaly have ≥ 1 accessory pathways and most accessory pathways are located around the orifice of the malformed tricuspid valve. In addition, wide QRS tachycardia over a septal accessory atrioventricular pathway, ventricular tachycardia, or flutter, as well as ectopic atrial tachycardia, atrial flutter, and atrial fibrillation, can occur. [6] We encountered 3 patients who had AF pre operatively and 1 who had classical ventricular pre excitation syndrome.

The optimum time and type of treatment of Ebstein's anomaly is still controversial and valve repair and valve replacement are the 2 available options. At many centres, valve repair is not practised today due to lack of expertise and equipment. At such centres, valve replacement remains the only available treatment. Also, in those valves which are difficult or are not amenable to repair, bioprosthetic tricuspid valve replacement is preferred. We had a favourable experience with bioprosthetic valve replacement in Ebstein's anomaly with no hospital deaths or on 5 year follow up. None of the patients had undergone structural valve degeneration in this study and the valve had served them well.

The surgical procedure remains very simple, just similar to any other valve replacement with the exception of sutures on the septal tricuspid leaflet where chances of injury to the AV node and conduction bundle are high. Our cross clamp and CPB times are just around an hour and two respectively, thus demonstrating the simplicity of the surgery. We have also used a variety of valves at the same position, however due to less number of patients; we did not do any comparison amongst those valves. Due to the annular dilatation that exists in these patients, it is relatively easy to insert large sized valves. All our patients were put valves of size 31 of different valve manufacturers.

was uneventful. Apart from atrial fibrillation and supra ventricular arrhythmias, most of the patients did not pose any challenges. Our patients were quickly relieved of their symptoms post surgery and after post operative care. This is evident from the fact that most of them reverted to NYHA class I or II before discharge and at follow up.

The choice of prosthesis at the tricuspid position remains a very interesting topic. There are reports of usage of bio prosthesis at the tricuspid position due to higher incidence of thromboembolic complications after mechanical valves. Bioprosthetic valves have shown favourable results at the tricuspid position as compared to other positions, especially in patients of Ebstein's anomaly. [10,11]

Our study was limited by the fact that we had less number of patients. Also we did not attempt to compare valve replacement with any tricuspid valve repair. Our maximum follow up duration was 5 years and although we did not observe any structural valve deterioration, we expect it to happen over a period of time. We will keep a close follow up of the same.

Conclusion

Tricuspid valve replacement using a bioprosthetic valve is an excellent modality for treatment in patients of Ebstein's anomaly with good in hospital results and results on short term follow up. The procedure is simple and can be replicated with good success. It has lasting effects on the patients' clinical status and can be performed with relative ease considering the very interesting clinical presentation and characteristics of Ebstein's anomaly.

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The ICU stay and post operative stay in our case, in nearly all patients