

Case Series

Modified cone procedure along with bidirectional cavopulmonary shunt for Ebstein anomaly in adult patients

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Received: 20 June 2022

Revised: 02 July 2022

Accepted: 05 July 2022

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ABSTRACT

Ebstein anomaly is an uncommon congenital cardiac disease affecting the tricuspid valve and right ventricle. Da Silva pioneered the cone reconstruction of tricuspid valve for surgical repair of this anomaly in 2007. We describe our experience of 5 adult patients who underwent cone repair along with a bidirectional cavopulmonary shunt and tricuspid annuloplasty ring placement at our institute. Five adult patients with Ebstein anomaly with progressive disease and symptoms were taken up cone reconstruction from 2019 to 2021. Along with cone reconstruction of tricuspid valve, we routinely added an end to side superior vena cava to right pulmonary artery shunt and a tricuspid annuloplasty ring in the newly formed tricuspid annulus in all cases. All patients tolerated the procedure well and easily weaned off cardiopulmonary bypass. There were no complications regarding bidirectional cavopulmonary shunt and only one patient had a temporary heart block which was managed conservatively. This series shows routine addition of a cavopulmonary shunt and an annuloplasty ring improves outcomes and can be performed at other experienced cardiac surgery centres.

Keywords: Ebstein anomaly, Adult congenital heart disease, Modified cone reconstruction, Bidirectional cavopulmonary shunt, Tricuspid annuloplasty ring

INTRODUCTION

Ebstein anomaly is a rare congenital malformation affecting the tricuspid valve and right ventricle and accounts for less than 1% of all congenital heart diseases.¹ Wilhelm Ebstein, a German pathologist, first described this anomaly in 1866.² Asymptomatic patients with stable hemodynamic pass on to adulthood and are diagnosed later as disease progresses and require surgical treatment. After many unsuccessful attempts at repair, da Silva described the cone reconstruction technique.^{3,4} Modifications of this technique have been used by Dearani and others and it has become the preferred repair method for patients with Ebstein anomaly.⁵⁻⁹ Here we

described the surgical technique and the results of modified cone reconstruction along with bidirectional Glenn shunt used for the treatment of Ebstein anomaly in 5 adult patients at a tertiary care referral centre in North India.

CASE SERIES

All adult patients aged more than 18 years referred to us with Ebstein anomaly from 2019 to 2021 were included in the study. A total of 5 patients, all females underwent the corrective surgery after a detailed clinical examination, laboratory and echocardiographic evaluation. They presented with symptoms of dyspnoea

on exertion of New York heart association class II (4 patients), palpitation (all 5 patients), syncope (2 patients), fatigue (4 patients), cyanosis (2 patients) and new onset arrhythmias (1 patient). Demographic and other details are enlisted in (Table 1). One patient had preoperative diagnosis of supraventricular tachycardia and was

managed conservatively by cardiologist. Two patients above the age of 40 years underwent coronary angiograms and were ruled out of having atherosclerotic diseases. Written and informed consents were taken from the patients for publication of this series.

Table 1: Clinical profile of patients.

Case no.	Age years	Symptoms	Carpentier classification	Tricuspid regurgitation	Biventricular function	Cardiomegaly	Associated anomalies
1	31	Fatigue, dizziness, palpitation	B	Severe	Good	Present	Supraventricular tachycardia
2	18	Dyspnoea, fatigue, palpitation, syncope	B	Severe	Good	present	Atrial septal defect
3	50	Dyspnoea, fatigue, palpitation, syncope	C	Severe	Mild RV dysfunction	Present	None
4	50	Dyspnoea, palpitation	B	Severe	Good	Present	None
5	23	Fatigue, dyspnoea, palpitation	B	Severe	Mild RV dysfunction	Present	Atrial septal defect

Surgical technique

Cardiopulmonary bypass was instituted using aortobicaaval cannulation with high innominate cannulation. Procedure was started with a bidirectional cavopulmonary end to side anastomosis of superior vena cava to right pulmonary artery to reduce the work load of a dilated right ventricle in all cases after obtaining a favourable left atrium pressure measurement (<15 mm of Hg). Modified cone reconstruction of the tricuspid valve starts with meticulous surgical delamination of the viable leaflets (Figure 1).

The anterior leaflet detachment from annulus started from the most leftward point of its attachment to the annulus, leaving only a small segment attached and continues clockwise toward the posterior leaflet. While detaching care was taken to include a very small part of muscle at the tricuspid annulus, to provide sufficient tissue for future re-suturing to the annulus. All the muscular and fibrous attachments were freed except the chordae tendinea of the main papillary muscle attached to the free margin. The remnant small septal leaflet was also delaminated as much possible. After the septal leaflet has been delaminated, the mobilized leaflet complex was rotated clockwise and approximated to the leading edge of the anterior leaflet (Figure 2).



Figure 1: Delamination of tricuspid valve (thin arrow) and atrialised right ventricle (thick arrow), patent foramen ovale (star).



Figure 2: Plicated atrialised Right ventricle (thin arrow), lead in edge attached to free margin of septal leaflet with 360° rotation (thick arrow).

This creates a 360° cone of leaflet tissue at the true anatomic annulus. Atrialised portion of the right ventricle was then plicated with multiple horizontal mattress sutures safeguarding the right coronary artery and its branches. The newly formed true tricuspid annulus in then sized and reconstructed leaflet cone was attached to it. Care was taken to note the triangle of Koch and Vein of D as marker for not injuring the conduction system. Competency of the newly reconstructed valve was then tested with saline injection into the right ventricle (Figure 3).



Figure 3: Completed cone reconstruction with competent tricuspid valve.

It was then followed by reinforcement with tricuspid annuloplasty ring (St Jude Medical, Tailor series) of adequate size. Atrial septal defect if present was closed with native pericardium with a 4 mm surgical patent foramen ovale creation to provide post operative decompression of any residual tricuspid regurgitation. Right atrial reduction-plasty was then done if required. Transesophageal echocardiography was done to ensure outcome of the repair after coming out of cardiopulmonary bypass (Figure 4 and 5).



Figure 4: Pre-operative trans esophageal echocardiography showing “sail like” anterior leaflet.



Figure 5: Post-operative trans esophageal echocardiography showing good coaptation of new tricuspid valve.

All patients tolerated the procedure well and were able to wean off cardiopulmonary bypass with ease. The mean cardiopulmonary bypass time was 300±41 min whereas aortic cross-clamp was 185±27 min. Post operative transesophageal echocardiography showed 4 patients with no to mild and 1 with moderate tricuspid regurgitation. All patent foramen ovale had a left to right shunt. One out of the five patients developed an atrioventricular block in the post operative period and was managed with temporary pacing. None of the patients had significant facial swelling of engorged head and neck veins, right ventricular dysfunction or pleural effusion. All had uneventful recovery and were discharged with stable hemodynamics. In follow up period, longest being 1 year, all patients were asymptomatic with no signs of arrhythmias, right heart failure, tricuspid stenosis. Two patients were found to have mild tricuspid regurgitation on postoperative echocardiography and 3 patients having no shunt flow across foramen ovale.

DISCUSSION

Ebstein anomaly was first described by Dr. Wilhelm Ebstein (1836-1912), a German physician, in 1866 in a 19-year-old labourer who died of cyanotic heart disease.² Ebstein malformation of the tricuspid valve is characterized by failure of delamination of the septal, inferior, and anterior leaflets of the tricuspid valve with subsequent adherence of the leaflets to the underlying myocardium; apical displacement of the functional tricuspid annulus (septal>inferior>anterior); dilation of the atrialised portion of the right ventricle; anterior leaflet fenestrations, redundancy or tethering and dilation of the true anatomic tricuspid annulus.¹⁰ This defect is associated with various other cardiac abnormalities like Atrial septal defect, Ventricular septal defect, right ventricular outflow tract obstruction, Patent ductus arteriosus, Coarctation of the aorta, accessory conduction pathways (Wolff-Parkinson White WPW syndrome) etc.¹¹ The most common presentation varies with age at

presentation including cyanosis, heart failure in infants, an incidental murmur in children. In adults it presents as arrhythmia, decreased exercise tolerance, fatigue or right-sided heart failure.¹²

Asymptomatic patients or symptomatic patients with functional classes I and II are managed conservatively. Indication for surgical intervention include deteriorating exercise capacity, NYHA class III, IV; progressive cardiomegaly on chest X-ray (cardiothoracic ratio >0.65), cyanosis (oxygen saturation <90%), progressive right ventricular dilation or reduction of systolic function, appearance or progression of arrhythmias and paradoxical embolism.¹³ surgical treatment is aimed at tricuspid valve repair or replacement, plication of atrialised right ventricle, closure of associated atrial or ventricular septal defects, reduction of right atrium, removing any right ventricle outflow obstruction and anti-arrhythmic procedures if indicated. Da Silva revolutionised the repair of tricuspid valve with cone reconstruction due to its close similarity to native anatomy.³ Dearani et al added modifications in the procedure for the neocuspidisation of tricuspid valve.⁵⁻⁹

In our series we routinely added a bidirectional cavopulmonary shunt to the procedure to decompress the right ventricle and new tricuspid valve with good results irrespective of presence of right ventricular dysfunction. Though adding this shunt has its own disadvantages like facial swelling, veno-venous collaterals, pulmonary artery venous fistulas and more importantly loss of access through internal jugular vein for future cardiac catheterisation and pacemaker implantation, we did not encounter any of these complications in any of our patients. In a series from Mayo clinic about 30% of patients required a bidirectional cavopulmonary shunt.¹⁴ We also routinely used complete annuloplasty ring for the new true tricuspid annulus which prevents from future annular dilatation while protecting the conduction system. Only one of our 5 patients developed early post operative heart block which was managed conservatively. Overall results published by da Silva in 2007 and later by Anderson et al in 2013 had good outcomes post cone reconstruction with few complications.^{3,14}

CONCLUSION

Our case series shows initial promising results of cone reconstruction with routine bidirectional cavopulmonary shunt with tricuspid annuloplasty ring implantation in adult patients. Though a bidirectional cavopulmonary shunts are not mandatory with additional risks, we performed them routinely without associated complications. Tricuspid annuloplasty ring added a stability to newly formed annulus. However, mastering the technique has long learning curve. protection of the right coronary system while plicating the atrialised right ventricle and conduction system to prevent any heart blocks are the key to successful outcome.

ACKNOWLEDGEMENTS

Rakesh Sharma, Satyajit Samal, Rahul Banik was actively involved in the management of the cases and collection of data and prepared the manuscript. Saket Agarwal, trained at Cleveland Clinic, Ohio, USA in congenital cardiac surgery and Sayyed Ehtesham Hussain Naqvi were the chief surgeons of the cases. Mohammad Abid Geelani reviewed the manuscript and supervised the series.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Sharma R, Samal S, Banik R, Aggarwal S, Naqvi SEH, Geelani MA. Modified cone procedure along with bidirectional cavopulmonary shunt for ebstein anomaly in adult patients. *Int Surg J* 2022;9:1470-4.