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Original Article:

Ebstein Anomaly: Single Centre Experience

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Abstract:

Ebstein anomaly, a congenital cardiac malformation affecting the tricuspid valve and right ventricle, presents with a spectrum of clinical features. This study reports on a series of surgeries performed to address Ebstein anomaly at GB Pant Hospital in Delhi. The mean age of patients at the time of surgery was 35 years, predominantly females. The most common symptoms were cyanosis, palpitations, and poor exercise tolerance. Surgical repair typically involved tricuspid valve reconstruction and atrial septal defect (ASD) closure, supported by bidirectional cavopulmonary shunt, with the goal of alleviating right ventricular dysfunction and tricuspid regurgitation.

Intricate procedures were undertaken, including delamination of tricuspid valve leaflets and plication of the atrialized right ventricle. The use of bidirectional cavopulmonary shunt played a significant role in improving outcomes. One patient required tricuspid valve replacement, and another had ASD closure without repair due to high surgical risk. Postoperative results were generally favorable, with the majority of patients experiencing minimal to mild tricuspid regurgitation.

The study emphasizes the importance of early intervention, particularly before significant right ventricular dysfunction occurs. The long learning curve and the need to protect vital structures during the procedure are highlighted. In summary, the combined approach of tricuspid valve reconstruction, ASD closure, and bidirectional cavopulmonary shunt is a promising strategy for managing Ebstein anomaly in adults.

Keywords: Ebstein anomaly, tricuspid valve repair, bidirectional cavopulmonary shunt, surgical outcomes.

Introduction

Ebstein anomaly is a congenital cardiac malformation primarily involving the tricuspid valve and right ventricle where tricuspid valve adheres to the underlying right ventricle myocardium. Failure of delamination leads to adherence of tricuspid leaflet to RV myocardium. The apical displacement of the valve divides RV into proximal atrialized and distal ventriculized portion. Right coronary artery denotes true location of of tricuspid annulus. Ebstein anomaly was first described by Wilhelm Ebstein in 1886 in a single autopsy specimen (1). Septal leaflet is most severely affected, posterior leaflet is nearly always affected. Anterior leaflet is enlarged and sail like. Septal and posterior are usually displaced with maximum displacement at the commissure between them. Thus functional tricuspid annulus is rotated apically and anteriorly.

Methodology:

CASE NO.	AGE/SEX	CARPENTIER CLASSIFIACTION	BIVENTRICULAR FUNCTION	ASSOCIATED ANOMALY	SURGERY PERFORMED
1	31/F	B	GOOD	SVT	CONES REPAIR+ BD Glenn
2	18/F	B	GOOD	ASD	CONES REPAIR+ BD Glenn
3	50/F	C	MILD RV DYSFUNCTION	NONE	CONES REPAIR+ BD Glenn
4	56/F	B	GOOD	NONE	CONES REPAIR+ BD Glenn
5	23/F	D	MILD RV DYSFUNCTION	ASD	TV replacement+ BD glenn
6	45/F	C		NONE	CONES REPAIR+ BD Glenn
7	24/F	B		NONE	ASD CLOSURE+ BD Glenn
8	22/F	D	MOD RV DYSFUNCTION	ASD	CONES REPAIR+ BD Glenn
9	46/M	D	GOOD	ASD	CONES REPAIR+ BD Glenn

Method

All patients undergoing surgery for ebstein anomaly at gb pant hospital were included in this series operated by two surgeons. Mean age of the patients at surgery was 35 yrs. Only one of the patient was male. Cyanosis, palpitation and poor exercise tolerence were the most common presentations, with cyanosis in 8 patients. Chest pain was present in 5 of the patients. Two patients had mild RV dyfunction and one had moderate dysfunction. Four patients had an associated ASD and one patient

had preoperative diagnosis of SVT which was managed medically. Three of patients underwent conventional coronary angiography which were normal.

Surgiacl Treatment

Cardiopulmonary bypass was initiated using bicaval canulation. Procedure was started with a bidirectional cavopulmonary end to side anastmosis of SVC to RPA to reduce workload of RV.

Right atrium was opened parallel to atrioventricular grove. Sump sucker was placed across atrial septum into left atrium. Tricuspid valve was examined, then anterior leaflet was detached from leftward point and continued towards posterior leaflet clockwise. All muscular and fibrous attachments were freed except the chordae tendinae from main papillary muscle attached to the free margin. Septal leaflet was also delaminated. Mobilized leaflet complex was rotated clockwise and approximated creating 360 degree cone.

Atrialized RV was plicated using multiple horizontal mattress sutures taking care of right coronary artery. Reconstructed leaflet cone was attached to newly formed tricuspid annulus. Tricuspid regurgitation was then tested with saline injection.

A SJM ring of adequate size was used for annuloplasty. ASD was closed using native pericardium. TEE was done intaroperative (Fig. 1) routinely to ensure outcome of repair after weaning from bypass. One patient had difficulty in delamination with small anterior leaflet which was inadequate for cones repair for that tricuspid was replaced by SJM size 21 metallic valve

In one patient with carpentier classification D and on intraop TEE GOSE score of 2.1, repair and plication was not feasible. On saline injection there was mild to moderate TR, only ASD was closed with creation of patent foramen ovale. Patient had moderate TR on TEE after weaning and patient had an uneventful postoperative course. External plication was done in one patient with high risk of injury to right coronary, result was satisfying.

All patients tolerated procedure well, the mean cardiopulmonary bypass time was 300 minutes and aortic cross clamp was around 180 minutes. Postoperative TEE showed 5 patients had minimal to mild tricuspid regurgitation. 2 patients had moderate TR. one patient expired on day 4 due to neurological complication (meningitis). one patient had AV block was managed with temporary pacing.

Discussion

Tricuspid valve repair with ASD closure is prefered correction. Most repairs convert tricuspid valve into monoleaflet using anterior leaflet to establish valve competence (2). Danielson and colleagues plicated atrialized portion of ventricle, narrows the tricuspid orifice resulting in monoleaflet (3). Modification of Danielson used ring for annuloplasty (4). In Carpentier repair anterior leaflet is mobilised and detached from anulus. Atrialized ventricle is pliacted vertically at right angles to direction used by Danielson. Da silva and collegues purposed cone repair where anterior and posterior leaflets are detached from anulus as single unit(5). Mobilized from there anomalous attachment and rotated clockwise fashion to suture to septal border of anterior leaflet. Rotated leaflets are are attached to true anatomical anulus.

General agreement is that operation should be advised before signifiacant RV dysfunction. Right ventricular dysfunction is important preoperative parameter as evident as ebstein anomaly is more than

valve abnoramlity in that RV has underlying myopathy (6). Other imporatant predictors were cynosis GOSE score and carpentier score.

Ebstein anomaly is associated with various other abnormalities like ASD, VSD, RVOT obstruction, PDA Coarctation of aorta, acessory conduction pathway (wolf parkinson white WPW syndrome) etc6. Common presentation in infant is cynosis, heartfailure. In adults it presents as arrythmia decreased exercise tolerernce fatigue or right side failure. Idiaction of surgery includes deteriorating exercise capacity, progressive cardiomegaly, cynosis, progressive RV dilatation, appperance or progression of arrythmia and paradoxical embolism.

Plication was done in all pateints except one where ASD alone was closed without repiar. Plication helps improving systolic function by reducing size of nonfunctional RV. It also improves LV function by decrasing compression and septal bowing. The major disadvantage being potential risk of injury to RCA worsening RV function and incidence of arrythmia. No injury to RCA was seen in any case of our patients.

Bidirectional cavopulmonary shunt was done in all patients to create one and half ventricular repair, it can also aleviate incidence of tricuspid stenosis which may be due to reducing size of tricuspid anulus but risk of facial swelling, pulmonary AV fistulas and compromise of access for pace maker leads. Most of patients referred to our institute were having sever

tricuspid regurgitation, right ventricle dysfunction. None of our patients had complication from BD shunt.

Conclusion

Our early experince with cones reonstruction with routine bidirectional cavopulmonary shunt with tricuspid annuloplasty in adult is satisfactory. However due to long learning curve, protection of conduction bundle and RCA during plication are key factors for successful repair of ebsteins anomaly. Adding bidirectional cavopulmonary shunt is good option to alleviate RV dysfunction and TR Valve replacement and ASD closure are also option which can be used in extremes of ebstein anomaly.

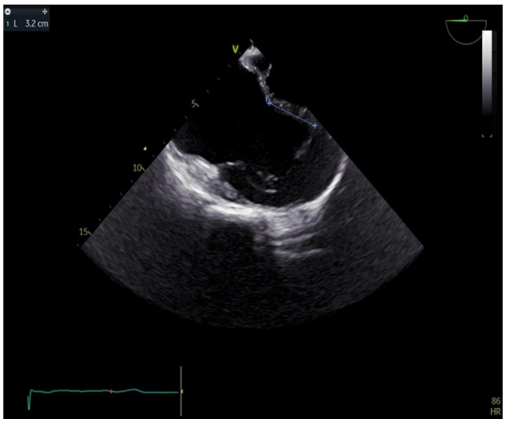

Figure 1: Intra-op transesophageal echocardiography showing dealamination of septal leaflet with apical displacement

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