RESEARCH ARTICLE

Cone repair had better short-term outcome than modified mayo repair in pediatric non-neonatal ebstein anomaly

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AIM: The study compared our results of two techniques of biventricular repair in pediatric patients with Ebstein anomaly.
BACKGROUND: Ebstein anomaly comprise a diversity of clinical presentations and hence a multitude of surgical techniques of repair. In a group of patients who deemed candidates for biventricular repair there are two common techniques of repair the simplest is Mayo clinic repair and the other is cone repair.
METHODS: A retrospective single center review of pediatric patients who underwent biventricular repair for Ebstein anomaly Between 2008 and 2018. Exclusion criteria were neonatal presentation, Tricuspid valve replacement and patients older than 16 years old as they managed in adult cardiac center. There were 17patients done by Mayo clinic technique and 14 patients by the recently introduced cone repair. The early outcome in both groups were evaluated regarding; the post-operative course, occurrence of low cardiac output, Reopening, Cardiac morbidity, ICU length of stay and presence of Significant Tricuspid regurgitation (defined as more than mild at 6 months in postoperative follow up echocardiography).
RESULTS: there were 31 patients enrolled in the study the mean follow up period was 24.0 ±9.38 months, range (12- 42 months), The study included 17 patients who received Mayo clinic repair, their median age was 36 months range (9 – 137) months and median weight was 13.5 kg range (7.5 -32) kg. while in Cone group repair the results were 47 months, range (19 -127) months and 15.5 kg range (11-30) kg respectively. There were no statistically significant differences among both groups regarding the incidence of postoperative cardiac morbidities, but the differences in ICU and postoperative Tricuspid regurgitation were statistically significant.
CONCLUSION: In spite of the diversity of available techniques for biventricular repair of pediatric Ebstein anomaly the Cone repair offer both anatomical and physiologic correction of those group of patients. The leaflet to leaflet interface and the shift up annular reconstruction alleviate the pathophysiologic alterations in Ebstein anomaly.

INTRODUCTION

Ebstein anomaly constitute nearly 1 % of all congenital heart defects, the presentation of those patients may appear in perinatal, neonatal, infantile, pediatric and adulthood. The clinical picture ranged from intractable cyanosis, shock and near arrest down to the asymptomatic adult patients [1,2]. Beyond the clinical picture the morphologic characters of the right heart differ between every patient [3]. As stated by Dr Dearani from Mayo clinic there is no black and white or all or none role in the management of Ebstein anomaly [4]. To tolerate this diversity surgeons over decades tried to allocate each subgroup to a tailored strategy [5]. A good procedure doesn't guarantee a good quality of life unless done in a tailored strategy [6]. So, in the group of patients who diagnosed as infantile Ebstein anomaly and grown up safely beyond the first few months of life the Biventricular pathway seems to be amenable [7]. There are multitude of techniques of biventricular repair of Ebstein the most common are 2 the Mayo clinic repair and the Cone repair [8-10]. Few who addressed the differences in both strategies in themanagement of non-neonatal Ebstein anomaly [11]. We will try in the current study to known the comparison between both technique in the biventricular management of non-neonatal pediatric Ebstein anomaly.

Patients and methods: a retrospective study conducted in 2018 to include patients operated at pediatric cardiac surgery unit between 2008 till 2017 who had Ebstein anomaly. Exclusion criteria were neonatal Ebstein, age above 16 years, Tricuspid valve replacement.

The operative records, ICU records and the outpatient's records were reviewed to obtain the most variables required in the study. The ethical committee at National Heart Institute approved the study protocol.

Indications for surgery included symptoms of right heart failure, cyanosis, shortness of breath, Progressive right ventricular enlargement, right ventricular dysfunction, or refractory atrial tachyarrhythmia or decreased exercise tolerance in children. The utilization of GOSE score and Carpentier's classification.

The preoperative data included the demographic data and the heart team decision for the surgical strategy. The operative records included anesthesia data, cardiopulmonary bypass data and the surgical procedure. The outpatient's records included the clinical data, follow up echocardiography and the follow up ECG.

Anaesthetic consideration

All patients were premedicated with midazolam 0.5 mg/kg orally 15 minutes before transfer to the operating room (OR). On arrival to the operating room 5-leads electrocardiogram (ECG) and pulse oximeter were connected to the patients and continuously displayed on the monitor (Drager Infinity Kappa, Danvers, USA). The patients received inhalational induction anaesthesia by sevoflurane 1% in 2 litre oxygen while inserting peripheral intravascular cannula and an arterial line if possible, then Induction of anaesthesia consisted of 2% sevoflurane in 80% oxygen, fentanyl 5 ug/kg and pancuronium 0.1 ug/kg to facilitate endotracheal intubation. After intubation the patients were connected to the anaesthesia machine (Drager Primus, Lubeck, Germany). Patients were ventilated with 60% inspired oxygen in air before cardiopulmonary (CPB) and 80% after bypass. Anaesthesia was maintained by 1% isoflurane in oxygen-air mixture and 1ug/kg fentanyl every 30 minutes before and after CPB, and during CPB with midazolam 0.1 mg/kg and 1% isoflurane.

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Surgical procedures

Through midline sternotomy, the pericardial cavity was entered after partial thymectomy. A large piece of pericardium was harvested and treated with 2% glutaraldehyde for 5minutes. After limiteddissection of the required structures, Heparinization, then the ascending aorta was cannulated and Both cava were cannulated selectively to obtain clear view in the right side of the heart After going on bypass, a state of moderate hypothermia is maintained during a state of cardioplegic arrest. The state of cardioplegic arrest was achieved by either timely infused doses of St Tomas cardioplegia solution before 2012 or a single dose of Custidiol after 2012. The dose of Custidiol is effective for three hours of continuous cardioplegic arrest.

Concepts of Cone repair

This technique was invented by Da Silva and colleagues, it entails surgical circumferential anterior tricuspid leaflet delamination from the underlying right ventricle. Then the Mobilized leaflets (anterior, diminutive inferior, and septal) were joined side to side to create a circumferential cone of leaflet tissue thus called 360-degreerepair (Figure 1,2). Some surgeons added Leaflet augmentation to increase leaflet height or increase the size of the TV annulus. When a linear attachment of the leading edge of the leaflet was present, Neochordaewere done or autologous neochordae werecreated by linear fenestrations. Internal RV plication was performed on smooth, nontrabeculated inferior RV wall endocardium [12,13].

Concepts of Mayo Clinic repair

The techniquescurrently used based on the design introduced by Gordon K. Danielson, Modifications of this technique included moving the base of the intact papillarymuscle on the RV free wall toward the ventricular septum atthe appropriate level with interrupted, pledgeted mattresssutures which referred as (Sebening sutures). the rightside of the anterior leaflet and annulus is approximated down to the ventricularseptum, thus improving proximity of the leading edge ofthe anterior leaflet with the ventricular septum. This repair was based on theconstruction of a functional "Monocusp" valve based on satisfactoryanatomy of the anterior leaflet. A purse string annuloplasty can be performed to further narrow the tricuspid annulus, especially in patients with extensive annular dilatation[14].

All patients had preoperative Electrophysiologic studies and accessory pathway was ablated in selected patients, we encountered a single case of refractory arrhythmias who required intraoperative intervention by radiofrequency ablation.

Echocardiographic Evaluations

All patients had preoperative and pre-discharge hospitalechocardiographic evaluations. Subsequent, late echocardiogramswere performed at our institution in 31 patientsat least every 6 months (rang 12-42 months) postoperatively. All studies were reviewed by onecardiologistto assess the degree of tricuspid regurgitation(TR), RV size, and systolic dysfunction. The degree of TR was assessed by standard techniques (width of vena contract, a density of Doppler waveform, and hepaticvenous flow changes). Tricuspid regurgitation was graded as 0, none; 1, trivial; 2, mild; 3, moderate; and 4, severe [2].

RESULTS

31 patients were included in the present study they constituted the patients who referred for biventricular pathway as deemed from preoperative evaluation and the heart team decision. The study included 17 patients who received Mayo clinic repair, their median age was 36 months range (9–137) months and median weight was 13.5 kg range (7.5 -32) kg. while in Cone group repair the results were 47 months, range (19-127) months and 15.5 kg range (11-30) kg respectively, there were no significant difference between both group regarding the Age and Weight as the P Values were 0.799 and 0.681 respectively. Demographic data analysis are shown in Table 1,2 and Figure 3 showing Gender distribution in both groups.

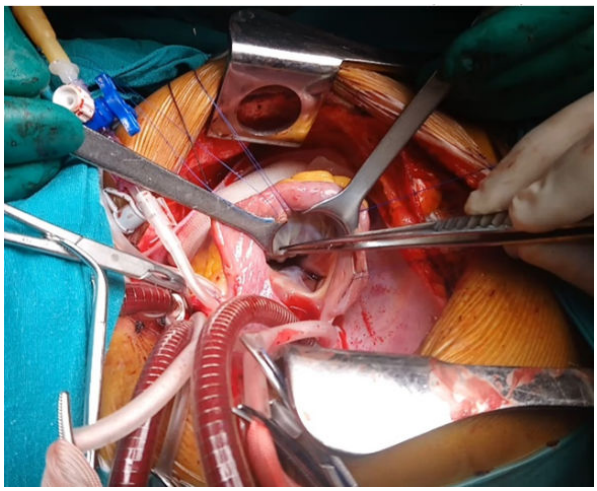

Figure 1) Intraoperative photo to demonstrate the morphology of the Tricuspid valve showing the adherent inferior and septal leaflets

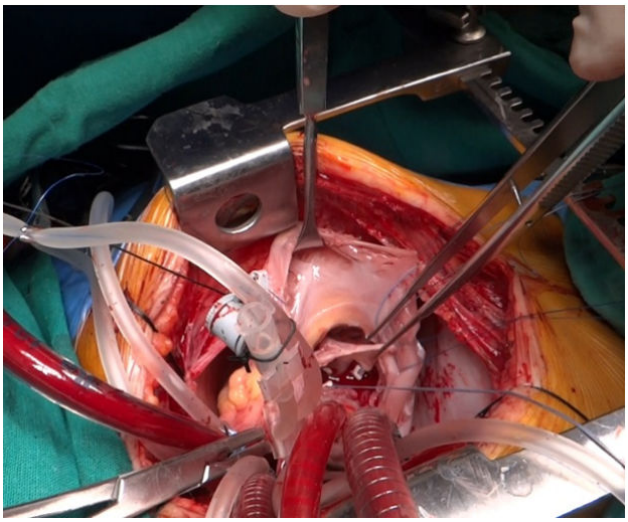

Figure 2) Intraoperative photo showing the separated anterior tricuspid leaflet and the Teflon pledge of the Sebening sutures

Table 1
Age and Weight comparison between groups

Group			Age	Wt
Cone	N	Valid	14	14
		Missing	0	0
		Median	36	13.5
		Range	128.5	24.5
		Minimum	9	7.5
		Maximum	137.5	32
Mayo	N	Valid	17	17
		Missing	0	0
		Median	47	15.5
		Range	108	19

	Minimum	19	11
	Maximum	127	30
Level of Significance	P Value	0.799	0.681

Table 2
Gender distribution on both groups

Group					
			Cone	**Mayo**	**Total**
Gender	**Female**	Count	3	4	7
		% within Group	21.40%	23.50%	22.60%
	Male	Count	11	13	24
		% within Group	78.60%	76.50%	77.40%
Total		Count	14	17	31
		% within Group	100.00%	100.00%	100.00%
P Value			0.889		

Each subscript letter denotes a subset of Group categories whose column proportions do not differ significantly from each other at the .05 level.

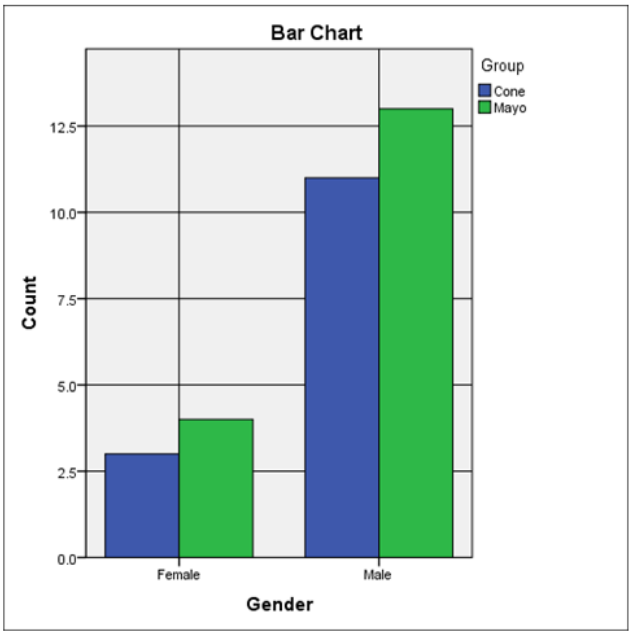

Figure 3) Bar chart of gender distribution in both groups

Postoperative outcome

In the ICU there were 2 patients (14.3%) in Cone group who developed post-operative dysrhythmias that required intervention vs 3 (17.6) in Mayo clinic repair group, P value: 0.800. these data are illustrated in Table 3 and presented in Figure 4.

Low cardiac output was present in 1 (7.1%) patient in Cone repair group vs 2(11.8%) patients in Mayo clinic group, P value: 0.664. overall morbidity was considered as 3 patients in Cone repair group vs 5 patients in Mayo clinic repair P value: 0.613. these data are illustrated in Table 4 and presented in Figure 5.

The median ICU stay in Cone repair group was 4 days, range (3-10) while in Mayo clinic repair group was 5 days, range (3-13). There was a significant difference between both groups regarding the ICU stay as the P value was 0.04. as shown in Table 5 and Figure 6.

All cases of morbidity are analyzed and presented in Table 6 and Figure 7.

There was one early mortality in cone repair (7.1%) and 2 (11.8%) in Mayo clinic repair and the P value was 0.665, these data are illustrated in Table 7 and Figure 8.

Tricuspid valve evaluation: the outpatients records revealed significant tricuspid regurgitation ≥ moderate on time table in 2 patients who had cone repair and 4 patients in Mayo clinic repair, P value: 0.045, these data are shown in Table 8 and 9 and presented in Figure 9.

Table 3
Incidence of Dysthymias during ICU stay in both groups

			Group		**Total**
			Cone	**Mayo**	
DysRhythm	No	Count	12	14	26
		% within Group	85.7%	82.4%	83.9%
	Yes	Count	2	3	5
		% within Group	14.3%	17.6%	16.1%
Total		Count	14	17	31
		% within Group	100.0%	100.0%	100.0%
P Value			0.800		

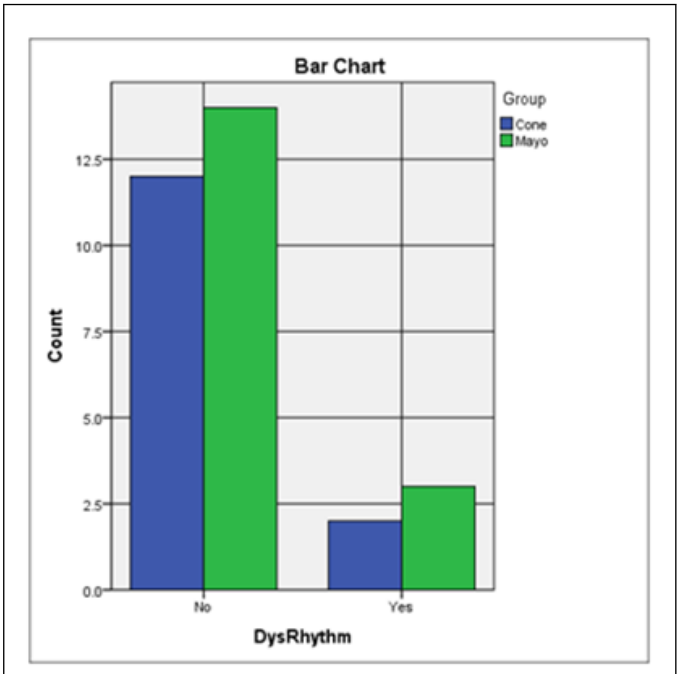

Figure 4) Bar chart showing incidence of Dysthymias at the ICU in both groups

Table 4
Incidence of low cardiac output during ICU stay in both groups

	Group		**Total**
	Cone	**Mayo**	

Low_CO	**No**	Count	13	15	28
		% within Group	92.9%	88.2%	90.3%
	Yes	Count	1	2	3
		% within Group	7.1%	11.8%	9.7%
Total		Count	14	17	31
		% within Group	100.0%	100.0%	100.0%
P Value			0.664		

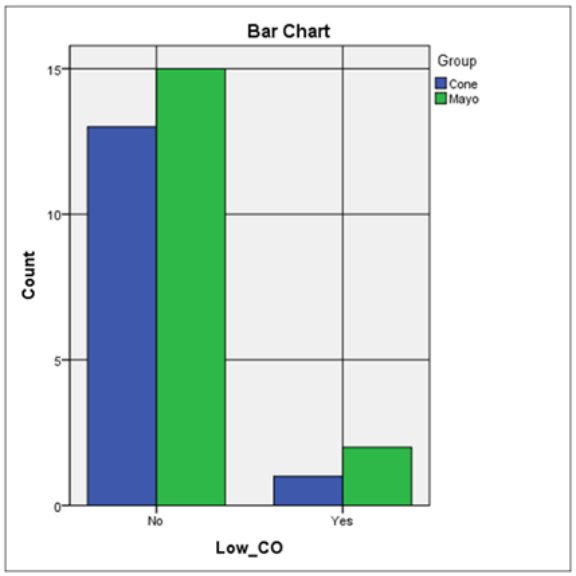

Figure 5) Bar chart showing incidence of low cardiac output at the ICU in both groups

Table 5
Descriptive statistics of the ICU stay in both groups

			ICU stay days
Cone	N	Valid	14
		Missing	0
	Mean	4.50	
	Median	4.00	
	Range	7	
	Minimum	3	
	Maximum	10	
Mayo	N	Valid	17
		Missing	0
	Mean	6.47	
	Median	5.00	
	Range	10	
	Minimum	3	
	Maximum	13	

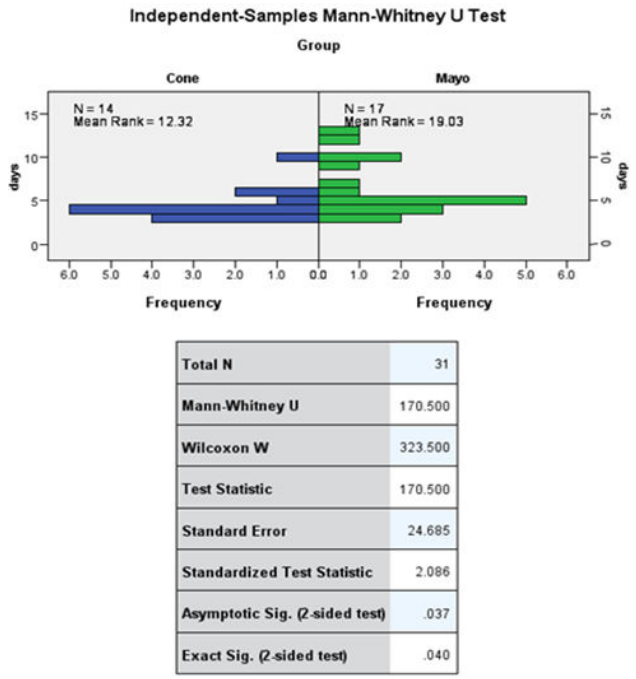

Figure 6) Bar chart showing statistical analysis of the ICU stay (days) in both groups

Table 6
Incidence of Morbidity during ICU stay in both groups

			Group		Total
			Cone	**Mayo**	
Morbidity	No	Count	11	12	23
		% within Group	78.6%	70.6%	74.2%
	Yes	Count	3	5	8
		% within Group	21.4%	29.4%	25.8%
Total		Count	14	17	31
		% within Group	100.0%	100.0%	100.0%

P Value	.613

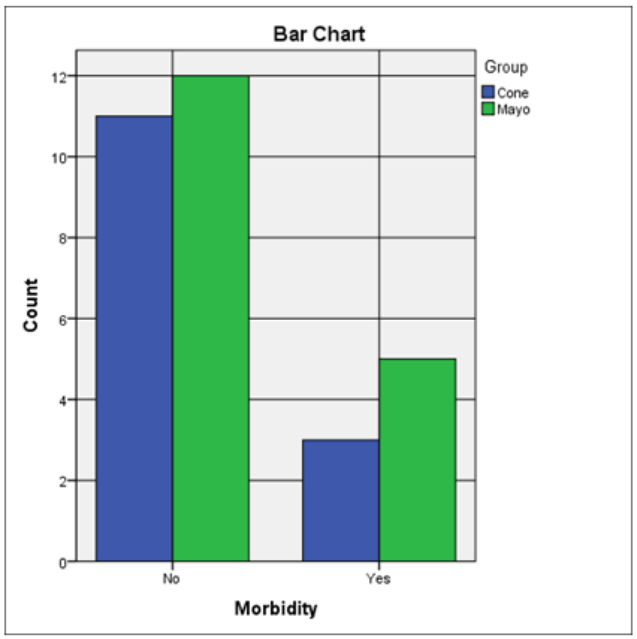

Figure 7) Bar chart showing incidence of Morbidity postoperative course in both groups

Table 7
Incidence of early Mortality in both groups

			Group		Total
			Cone	Mayo	
Mortality	No	Count	13	15	28
		% within Group	92.9%	88.2%	90.3%
	Yes	Count	1	2	3
		% within Group	7.1%	11.8%	9.7%
Total		Count	14	17	31
		% within Group	100.0%	100.0%	100.0%

P Value	.665

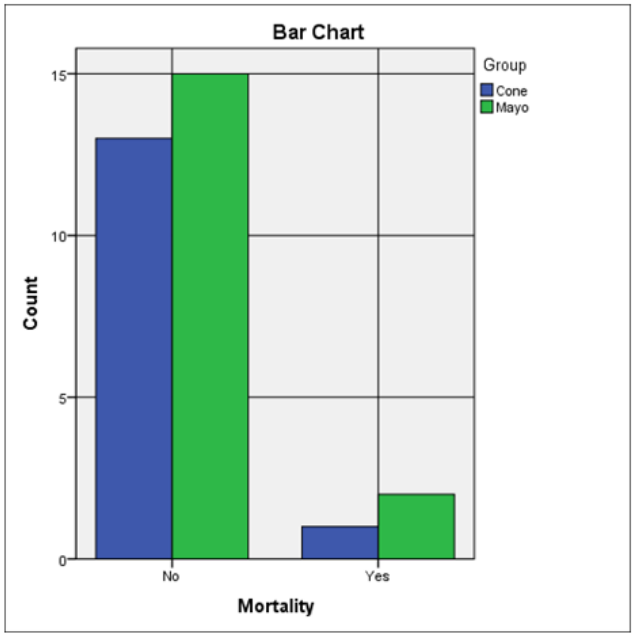

Figure 8) Bar chart showing incidence of early postoperative Mortality in both groups

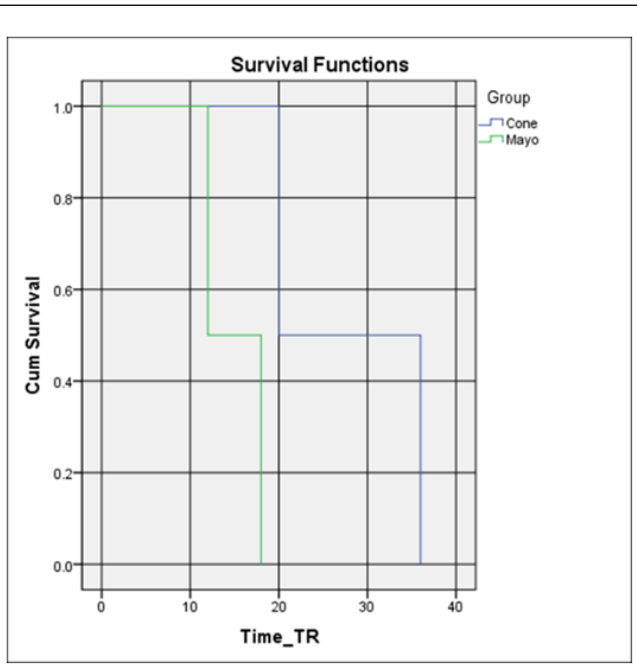

Figure 9) Kaplan-Meier for freedom of significant TR in both groups

Table 8
Statistical analysis of freedom from significant TR in the studied group

Overall Comparisons			
	Chi-Square	df	Sig.
Log Rank (Mantel-Cox)	4.032	1	.045

Test of equality of survival distributions for the different levels of Group.

Table 9
Cox Regression analysis of significant TR in the studied group

Group			N	Percent
Cone	Cases available in analysis	Event	2	14.3%
		Censored	0	0.0%
		Total	2	14.3%
	Cases dropped	Cases with freedom from Sig_TR	12	85.7%
		Total	12	85.7%
	Total	14	100.0 %	
Mayo	Cases available in analysis	Event	4	23.5%
		Censored	0	0.0%
		Total	4	23.5%
	Cases dropped	Cases with freedom from Sig_TR	13	76.5%
		Total	13	76.5%
	Total	17	100.0 %	

DISCUSSION

The biventricular strategy in the management of Ebstein anomaly is related mainly to the ability of the anatomical RV to manipulate the pulmonary circulation efficiently which may not be affected by the morphology of the tricuspid valve [15-17].

In the original technique of Mayo clinic repair the plication of the atrialized part was in transverse manner so the base become nearer to the apex, Monocusp orientation of the valve as the posterior annulus had placation [18]. The currently approved Mayo clinic added 2 important modifications the 1st was Sebening, s sutures and the 2nd was longitudinal approximation of the right component of the anterior annulus to meet the ventricular septum. Boston, et al in 2006 reported the long term follow up after Ebstein repair in children Mean age was 7.1 ± 3.9 years (range, 5 months to 12 years) the early mortality was 5.8%, the early incidence of moderate tricuspid regurgitation was 24% and 6% was moderately severe. They recommended repair over replacement in the setting of optimum anterior leaflet morphology, even for late failure of tricuspid repair they noticed that the bioprosthetic valve replacement in the tricuspid position have better outcome than any other position in the heart due to low gradient in the tricuspid position. The most important finding was absence of significant tricuspid stenosis even after 25 years of successful repair [19]. These results are comparable to our results as we got 23% incidence of significant TR and we had mortality incidence of 11%. Another group from Deutsches Herzzentrum Berlin operated on a group of patients with multiple age groups and applied multitude of techniques in tricuspid repair including Mayo clinic and Carpentier's repairs, they got lower mortality incidence (2.9%) which may be attributed to the higher mean age in their study[16].

Jose Pedro da Silva and his colleagues introduced the Cone repair and the mid-term results of their series were reported in 2007, 40 patients were operated for repair of Ebstein anomaly their mean age 16.76± 12.27 range (1-49) the early mortality was 2.5% and the results of tricuspid repair revealed grade 2 (5/40) , grade 3 (3/40) and no significant tricuspid stenosis[12]. In 2012 they reported the 100 patient's series they had 3% incidence of hospital mortality, 4% mid-term mortality and significant improvement in tricuspid competence[20].

Kimberly A. Holst et al operated 134 children who were diagnosed as Ebstein anomaly and got Cone repair, mean age in this study was 9.5 yrs. (range 4.4 – 13.2 years), no early mortality in their series. In this series Antiarrhythmic procedures were added to nearly 15% of patients and more modifications to the Cone repair techniques were added like; Leaflet augmentation, Neochordae and Annuloplasty band. Overall survival at 6 years was 98% they concluded that Cone repair is safe, but the learning curve is significant.Sustained reduction in TR and favorable changes in RV size and area at follow-up suggest that cone repair may have an advantageous impact on RV remodeling those findings make Cone repair better than Mayo Clinic repair even in their experience[14,21].

In our series of Cone repair and Mayo Clinic repair the reported Antiarrhythmic procedures were less as nearly all patients had preoperative electrophysiologic study only 1 patient in Mayo repair who had refractory arrhythmias that cleared by right atrial Maze procedure. we did not encounter heart block in this patient in the follow up after this limited Maze.

In our study we noticed better tricuspid valve competence, better postoperative hemodynamic profile in the group of patients who obtained Cone repair rather than Mayo clinic repair, these findings consistent with findings of Shu-Chien Huang et al, Kimberly A. Holst et al and Joseph A. Dearani et al.

LIMITATIONS

This rare anomaly did not permit us to enroll a good number of patients, this small number and the wide spectrum of morphologic features added to the difficulty in both techniques. The learning curve of the Con repair is slow that require a large number and longer follow up periods.

CONCLUSION

Ebstein anomaly is a spectrum between white and black, there is no all or none rule in the management of this anomaly, Anatomical repair is represented by the Cone repair, which is a demanding technique less reproducible than other techniques but carries the best outcome.

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