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A STUDY ON CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

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ABSTRACT

Alms and objectives: Congenitally corrected transposition of the great arteries (CCTGA) is a congenital cardiac condition which includes discordance of atrioventricular and ventriculoarterial connections. It is a rare anomaly and accounts for less than 1% of congenital cardiac conditions, It can have multiple different co existent anomalies. Surgical decision making is challenging as CCTGA is a rare condition with diverse morphological malformations

Materials and methods. This is a retrospective twenty year institutional study where twenty patients with CCTGA underwent different surgical repairs using the conventional method. We have analysed our results based on in hospital morbidity, mortality, complications and ten-year late survival. We have compared our results with different studies of conventional repair and anatomical repair.

Results: Nine patients underwent ventricular septal defect (VSD)closure (459%). two patients had atrial septal defect closure (10%). One patient had pulmonary valvotomy (5%), Four patients underwent tricuspid valve replacement (TVR) (20%). Two patients underwent VSD closure plus pulmonary valvotomy (l0%). One patient underwent VSD closure plus left atrioventricular valve repair (5%). One patient underwent pulmonary valvotomy plus TVR (5%), Post-operative bradycardia was seen in one. patient of VSD closure group. Two patients of VSD closure group reported postoperative complete heart block. Early mortality was seen in one patient of tricuspid valve replacement group. Late mortality was seen in two patients-TVR group and VSD closure plus left atrioventricular valve repair group. Reoperation was seen in two patients. Average ten year survival was 85%. Two patients developed post-operative right ventricular dysfunction.

Conclusion: Surgical management of CCTGA by conventional repair provides good results comparable with anatomical repair with respect to post-operative morbidity, mortality and long

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term survival. However caution should be maintained in patients with preoperative severe TR. Early repair of tricuspid valve should be done ideally before the development of right ventricular dysfunction.

Keywords: Congenially corrected transposition of the et arteries (CCTGA), ventricular septal defect (VSD), Bradycardia, Right ventricular dysfunction.

INTRODUCTION

Congenially corrected transposition of the arteries (CCTGA) Cardiac condition which İncludes discordance atrioventricular (AV) and ventriculoarterial (VA) connections. It is thus unique condition which can have physiologically normal circulation. It is a rare cardiac defect that accounts for less than 1% of congenital cardiac conditions. It usually coexists with conditions like ventricular septal defect (VSD) which is most common, pulmonary stenosis(PS) atresia, atrial septal defect(ASD)and abnormalities of the tricuspid valve like tricuspid regurgitation(TR). Surgical repair is of utmost importance as long term survival in these patients is unlikely.

Surgical repair can be done by multiple methods. Conventional repair involves correction of coexistent malformations associated with CCTGA. Anatomical repair includes correction of discordant connections of CCTGA. These include double switch procedures. Single ventricular palliation is the third method.[1.2]

Surgical decision making is challenging as CCTGA İs a rare condition with diverse morphological malformations. Therefore, it is important to be aware-of them so that prompt management can be done when required. We have studied the management and outcomes of patients with CCTGA using conventional repair. Few studies have been conducted regarding the same. We have intended to bring forward the management of CCTGA by conventional repair in Indian scenario as very few studies have been done according to Indian perspective and we have compared our results with international studies. [3,4,5]

New surgical paradigms are being developed like Double switch surgery and root translocations which are technically very demanding, resource intense with high morbidity and mortality. However, their long-term results are considered to be better than conventional surgical strategies. We adopted conventional surgical strategies and have reviewed our results in the light of modern developments.

MATERIALS AND METHODS

The present study was a retrospective study conducted for twenty years from January 2001 to December 2021. 20 Patients who were found to have congenitally corrected transposition of great arteries in data records were included in the study.

Inclusion criteria: Patients diagnosed to have CCTGA who underwent different surgical procedures by conventional repair in the last twenty years.

Exclusion Criteria: Patients of CCTGA who were operated elsewhere. Patients of CCTGA who were not operated.

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Prior to the commencement, the study was approved from the Ethical and Research Committee, Nizam's Institute of Medical Sciences, Hyderabad. As it was a retrospective study, no consent was required Waiver of consent was submitted to the Ethics committee.
Demographic data such as age, sex procedure details and post-operative details were obtained from case records. These findings were recorded on a predesigned proforma.

The patient was placed in a supine position for a standard median sternotomy. The anesthesiologist placed an arterial line (radial and femoral) for invasive monitoring of the patient's blood pressure(BP), a central line for venous access before induction of general anesthesia. General anesthesia administered and the patient was intubated.

The patient was prepped and draped, before surgical incision. Median Sternotomy was performed. The thymus was dissected and the pericardium was opened . Patient was heparinized. The patient's aorta superior vena cava and anterior vena cava were cannulated, and the tubing was attached to the cardiopulmonary bypass circuit (CPB) and cardiopulmonary bypass was Venous procedures were performed for different co existent anomalies.

RESULTS

The data obtained was analyzed and final results and observations were tabulated and interpreted

Table-1:Demographic distribution in present study.

Age range in years	**Number of patients**	**Percentages**
0-5	4	20%
6-10	4	20%
11-15	4	20%
16-20	1	5%
21-25	3	15%
26-30	0	0
31-35	0	0
36-40	2	10%
41-45	2	10%
Gender		
Males	13	65%
Females	7	35%
Symptoms		
Shortness of breath	18	90%
Bluish discoloration of skin	16	80%
Chest pain	1	5%
Upper respiratory tract infection(URTI)fever, cough	3	15%
palpitation	2	10%
Growth retardation	3	15%
Poor feeding	3	15%
Fatigue	12	60%
Signs		
Bradycardia	4	20%
Cyanosis	16	80%

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Hyperdynamic cardiac apex	20	100%
Loud second heart sound	5	25%
Heart murmurs- Pan systolic murmur Ejection systolic murmur	 17 3	 85% 15%
NYHA class		
I	1	5%
II	14	70%
III	4	20%
IV	1	5%

In our study most patients were in the age group range of 0-5 years (20%), 6- 10 years (20%) and l1-15 years (20%).Mean age was 784+/-14.15 years. Media age was 13.5 years, with range from I to 45 years. Most of the patients were male(65%) and only 35% were females.
Predominant symptoms were shortness of breath(90%), bluish discoloration of skin(80%) and fatigue(60%).

Hyperdynamic cardiac apex (100%) and majority of the patients had cyanosis(80%). 85% of patients had a pansystolic murmur.
Majority of the patients had NYHA II symptoms (70%). NYHA III symptoms were seen in 20% of patients.

Table-2: Preoperative findings in present study

Electrocardiogram	**Number of patients**	**Percentages**
Normal sinus rhythm	16	80%
1 st degree heart block	2	10%
2 nd degree heart block	1	5%
Complete heart block	1	5%
Q waves: reversal of precordial Q-wave pattern with deep Q waves in leads V2 and AVR, and QS complexed in leads V3 and AVF in right precordial leads.	2	10%
Coexistent cardiac defects		
VSD	9	45%
OS ASD	2	10%
Sever PS	1	5%
Left AV value	4	20%
Regurgitation	4	20%
VSD+PS	2	10%
VSD +Left AV Value regurgitation	1	5%
Sev PS +Left AV Value regurgitation	1	5%

Majority of the patients had normal sinus rhythm(80%). Co-existent VSD was present in 45% of patients.

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Table-3: Types of procedures performed

Types of procedures	**Number of patients**	**Percentages**	**Mean pump time (mins)**	**SD**
VSD closure	9	45%	108	8.631
ASD closure	2	10%	93	7.071
Pulmonary valvotomy	1	5%	116	
Tricuspid valve replacement	4	20%	221.25	16.091
VSD closure-**pulmonary valvotomy**	2	10%	112.5	7.778
VSD closure-**left AV value repair**	1	5%	156	
Pulmonary valvotomy-TVR	1	5%	111	

Average pump time in VSD closure group was 108 minutes, in ASD closure group was 93 mins, in pulmonary valvotomy group was 116 mins. One patient developed bradycardia post VSD closure. Patient was kept on AV sequential epicardial pacing for 2 days post operatively. 2 patients who underwent VSD closure had complete heart block. One of the 2 patients had complete heart block. One of the 2 patients had complete heart block preoperatively also needed permanent pacemaker implantation post operatively. Other patient had transient complete heart block and recovered spontaneously.

Table-4: Total duration of ventilation

Types of procedures	**Number of patients**	**Total ventilation duration**		
		Upto 24 hrs	**24-48 hrs**	**>48hrs**
VSD closure	9	7	2	-
ASD closure	2	2	-	-
Pulmonary valvotomy	1	1	-	-
Tricuspid valve replacement	4	3	-	-
VSD closure-pulmonary valvotomy	2	2	-	-
VSD closure-left AV value repair	1	1	-	-
Pulmonary valvotomy-TVR	1	1	-	-

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17 patients 85% were ventilated for less than/equal to 24 hrs. 2 patients with VSD closure were ventilated for 24-48 hrs. One patient who underwent tricuspid valve replacement was ventilated for>48 hrs. due to low cardiac output syndrome and hypotension. Patients remained intubated for 144 hrs. Patients expired on the 6 th post-operative day.

Table-5: Total Intensive care unit (ICU) stay

Types of procedures	Number of patients	Total Intensive care unit (ICU) stay		
		Upto 3 days	3-5 days	>5days
VSD closure	9	6	3	-
ASD closure	2	2	-	-
Pulmonary valvotomy	1	1	-	-
Tricuspid valve replacement	4	2	1	1
VSD closure-pulmonary valvotomy	2	2	-	-
VSD closure-left AV value repair	1	1	-	-
Pulmonary valvotomy-TVR	1	-	1	-

Majority of the patients stayed in the ICU for 3 days. One patient stayed in ICU for 6 days. Patient underwent tricuspid valve replacement and developed low cardiac output postoperatively. Patient underwent re-exploration and expired due to low cardiac output.

One patient who underwent tricuspid valve replacement developed transient atrial fibrillation after surgery. Patient was administered Inj. Amidarone infusion and recovered spontaneously on post-operative day 2.

One patient who underwent VSD closure developed moderate tricuspid valve dysfunction . TV dysfunction after the operation was defined as the need for TVR or TV function that deteriorated after the operation to moderate or severe incompetence. Patient eventually developed sever TR and needed tricuspid valve regurgitation at 8 months after surgery.

One patient who underwent tricuspid valve replacement developed low cardiac output . Patient underwent reexploration and expired on the 6 th post-operative day.

One patient in VSD closure group developed CHB within 1-5 years postoperatively and eventually required a permanent implantation. One patient of VSD closure group developed sever TR 8 months after surgery and required TVR . One patient of VSD closure+left AV valve repair group developed sever TR at 6 years postoperatively and required TVR.

Average 10 years survival of available patients was 85% . Time related survival of patients was studied after surgery till 2021 December. The lowest 5 year and 10 year survival rates were seen in the TVR group(50%). Five year survival of patient of VSD closure were operated in

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2018. Hence average 6-10 year survival study for this groups was not feasible. 5 year survival of ASD closure patients was 100% . One patient of ASD closure was operated in 2018. Hence 10 year survival study was not feasible. 10 year survival of pulmonary valvotomy group and VSD closure +pulmonary valvotomy group was 100% each. 5 year survival of patient who underwent pulmonary valvotomy +TVR was 100%

Two patients who underwent TVR and RV dysfunction preoperatively and did not survive. 2 patients developed RV dysfunction post operatively . Among them one patient had undergone VSD closure and developed RV dysfunction and sever TR 8 months postoperatively . Patient eventually needed TVR . One patient who underwent VSD closure +left Av valve repair developed RV dysfunction and sever TR six years after surgery and underwent TVR eventually and expired.

DISCUSSION

Optimal surgical management of CCTGA remains controversial . There are multiple methods in the surgical management of CCTGA and we have described our experience by performing conventional repair. In present study , patients in the age range of 4 years and 45 years . Most patients were in the age group range of 0-5 years (20%, 6-10 years (20%) and 11-15 years (20%). Among adult patients , majority belonged to the age range of 21-25 years(15%). In a study by Shin' 'oka et al[6], median age of patients with CCTGA undergoing, surgery was 8.3 years and age range was 2 months to 47 years. In a study by Hraska' et al[11], median age at the time of operation was 4.1 years. In a study by Bogers et al[7] 32 patients with CCTGA underwent different study by Sano et al [13], median age of surgery was 4 years and 9 months. The results of all these studies were comparable with the present study. the present study, majority of adults undergoing surgery were in the age range 21-25 years (15%). In the present study, majority of adults undergoing surgery were in the age range of 21-25 years(15%). This was comparable with a study by Talwar et al[8], where patients in the age range of l6 years- 41 years underwent surgery. Median age 25 years.

The present study, majority of patients were males(65%)while only 35% of patients were females, In a study by Hong Hsu et al.[9] 56 patients underwent different surgical repairs. Among them 15 patients underwent conventional repair. ln our study conventional repair group, male: female ratio was l0:5 which was comparable repair. In study by Graham Jr et al[10], in a study group of CCTGA patients with associated lesions with 132 patients, the number of male patients (63%) dominated the study group.

In our study, predominant symptoms were shortness of breath, bluish discoloration of skin seen in 80% of patients . Fatigue was present in 60% of patients. Other symptoms were repeated upper respiratory tract infections(15%), poor feeding(l5%) and growth failure(15%), palpitations(10%) and chest pain(5%)

Majority of the patients had NYHA II symptoms (70%) NYHA III symptoms were seen in 20% off patients. All the he patients in our study had a hyperdynamic cardiac apex(100%)and majority of the patients had cyanosis (80%). 85% of patients had a pansystolic murmurs, 15% had an ejection systolic murmur, 25% of patients had a loud second heart sound 20% of patients had bradycardia.

Many a times, patients of CCTGA might be asymptomatic and they might be diagnosed incidentally on chest x ray and electrocardiogram. Frequently patients have shortness of breath

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and cyanosis . RV dysfunction or tricuspid valve abnormalities could contribute to them. VSD or obstruction of left ventricle outflow tract may also contribute to them.

Majority of the patients in the present study had coexistent VSD(40%) .Left AV valve regurgitation was present in 20% of patients. In a study by Hraska et al[11], out of 123 patients , majority of the patients who underwent surgery for CCTGA had coexistent VSD (94 patients). 17 patients had coexistent tricuspid valve regurgitation. Kang Hong Hsu and colleagues[9] reported CCTGA with VSD in 12 cases out of 15 cases undergoing surgical repair by conventional methods. Significant TR was present in 6 cases. Sever PS was present in 11 cases. J Horer and colleagues[12] reported a study of 56 patients undergoing different surgical techniques for CCTGA . Among them VSD was present in 48 patients TR was present in 20 patients.

In our study majority of the patients underwent VSD closure (45%) . 20% of the patients underwent tricuspid valve replacement . 10% of patients underwent ASD closure. 5% of patients underwent pulmonary valvotomy, VSD closure +left AV valve repair and pulmonary valvotomy +tricuspid valve replacement each. In a study by Hraska et al[11] different surgical procedure were carried out in 123 patients. Among them 96 patients underwent conventional repair and majority of these patients(76) underwent VSD surgery and TV valve surgeries (14). Total pump time was taken as an outcome variable in our study. Average pump time in VSD closure group was 108 minutes , in ASD closure was 93 mins, in pulmonary valvotomy group was 116 minutes. TVR group had an average pump time of 221.25 minutes , in VSD closure+pulmonary valvotomy group was 1125 minutes, VSD closure +left AV valve repair was 156 minutes and n pulmonary valvotomy +TVR group was 111 minutes. P value was 0.001 which was statically significant,

In the present study one patient developed bradycardia post VSD closure. Patient was kept on AV sequential epicardial pacing for 2 days post operatively. Isoprenaline infusion was given for 2 days. Patient achieved normal sinus rhythm on post-operative day 3. 2 patients who underwent VSD closure had complete heat block . one of the 2 patients had complete heart block preoperatively also and needed permanent pacemaker implantation postoperatively. Other patient had transient complete heart block and recovered spontaneously.

In a study by Hraska[11] 113 patients of CCTGA underwent different surgical procedures. Among them 109 patients who did not have CHB preoperatively and 31 among those developed perioperatively CHB(28%).

Hong hsu and colleagues[9] conducted a study where different surgical procedures were performed for CCTGA. Group I had 15 patients who underwent conventional repair and group II had 18 patients who underwent anatomic repair. 2 patients from both groups had CHB who required permanent pacemaker implantation. 2 patients in group II had transient CHB after surgery which recovered spontaneously.

In a study by Sano et al[13] , 28 patients underwent conventional repair for CCTGA. 9 patients developed perioperative CHB(5 among those had CHB preoperatively also).Hiramatsu et al[14[performed double switch operations in90 patients. Group I has 72 patients who underwent trial switch with intraventricular rerouting . Group II had 18 patients with atrial switch plus arterial switch operations. Among whom 81 patients survived postoperatively. In the long term follow-up , atrial tachycardia was seen in 14 , ventricular tachycardia in three,

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arterial fibrillation in three, atrioventricular block in eight and sick sinus syndrome in six in group I and atrioventricular block in 2 patients in group II. The freedom from arrhythmia was 57.1% in group I vs 78.6% in group II.

These results are comparable with our study, Presence of perioperative CHB is one of the important features in CCTGA and may account for one of the reasons for low survival rate in CCTGA. Prior knowledge of the conduction system in CCTGA may help in reducing the risk of postoperative heart block. This may however not always be feasible since there may be an acquired abnormal progressive change in the conduction system. Studies reported surgical techniques to avoid injury to the Conduction system during surgical intervention of CCTGA. These include VSD closure to the left aspect of the interventricular septum through a right atrial right atrioventricular valve exposure. Talwar et al[8] reported no complications in patients undergoing anatomical and biventricular (BV) repair

The present study, 17 patients (85%) were ventilated for less than/equal to 24 hours. 2 patients with VSD closure were ventilated for 24-48 hours. One patient who underwent tricuspid valve replacement was ventilated for > 48 hours due to low cardiae output syndrome and hypotension. Patient remained intubated for 144 hours. Patient expired on the sixth postoperative day. Mean duration of ventilation was 38 hours. This is slightly higher than the present study. Reason Could be increased complexity of anatomic repair as compared to conventional repair. In study by Tabib et al[15], out of 300 patients undergoing congenital heart Surgery, 70% patients were extubated within 24 hours of surgery. In a study by Polito et al. [16] 11% of pediatric patients undergoing cardiac surgery remained ventilated for more than seven days. Risk factors like delayed sternal closure sepsis, reintubation and high vasoactive inotropic score were associated with prolonged ventilation>72 hours. According to Alrddadi et al[17] prolonged mechanical ventilation (defined as >72 hours) post congenital heart surgery was associated with high morbidity mortality and use of hospital resources. Early extubation shortens the intensive care unit complications and improved outcomes.

Early extubation and early weaning of children from mechanical ventilation after CHD surgery lead to rapid ambulation, improvement of the cardiopulmonary function, and decreased 1CU and hospital stays. In the present study majority of the patients (85%) achieved early extubation (<72 hours) after surgery.

Majority of the patients (70%) stayed in the ICU for three days. One patient stayed in ICU for six days. Patient underwent tricuspid valve replacement and developed low cardiac output postoperatively. Patient underwent re exploration and expired due to low cardiac output. Man-shik Shim reported the median stay in the ICU to be 5 days (range 4-35) among 15 survivors after anatomic repair for CCTGA. This is slightly higher than the present study. Prolonged 1CU stay was due to complications like prolonged pleural effusion, transient arrhythmia, CHB, severe pulmonary edema and aspiration pneumonia. Prolonged stay in the ICU is associated with high hospital mortality and considerable decrease in late survival.

Lubiszewska B et al[18], defined ICU stay upto 3 days as standard ICU hospitalization after pediatric cardiac surgery.

Preoperative risk factors for ICU stay >3 days include neonatal age, myocardial dysfunction (LVEF<25%). Intraoperative risk factors are raised complexity of operation, increased cardiopulmonary bypass time(more than 60 minutes), raised aortic cross clamp time and

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increases hypothermic circulatory arrest time. Post-operative risk factors include low cardiac output syndrome , important arrhythmia, sepsis, acute kidney injury and intensive postoperative bleeding. According to Wheeler et al[19] factors independently associated with a prolonged postoperative stay in the cardiac intensive care unit included prematurity, need for inotropic support prior to surgery, difficulty feeding, evidence of systemic capillary leak on postoperative day 2, and postoperative infectious complications.

One patient developed low cardiac output after tricuspid valve replacement. Patient underwent re exploration and expired on the sixth postoperative day. Patient were followed up till December 2021. On patient who underwent tricuspid valve replacement developed congestive cardiac failure and expired three years after surgery. One patient who underwent VSD closure +left AV valve repair developed tricuspid valve regurgitation six years post-surgery and underwent tricuspid valve replacement and expired. In a study by Hong Hsu et al [9] 15 patients underwent conventional repair 18 patients underwent anatomic repair and 23 patients underwent single ventricle palliation. Collectively there were 6 in hospital deaths. Reasons include severe heart failure in 1 patient of conventional repair group and 3 patients of anatomic repair group and one patient with sever sepsis with multiple organ failure in each group. No mortality was reported in single ventricle palliation group. Late mortality was seen in 7 patients due to sever heart failure (4 patients) and sepsis in 3 patients. Sano et al[13] conducted study of conventional repair in 28 patients, ln hospital mortality was 1 (4%). There were 3 late mortalities.

The above studies are comparable with the present study. Hong Hsu and colleagues reported severe TR in five patients in conventional repair after surgery. No TR was reported in anatomic and single ventricle palliation group. This study differed from the present study probably due to the low sample size of our study. According to Mongeon et al[20], post-operative RV function after TVR can be predicted from preoperative RV function. For best results, TVR should be considered at an early age, before the systemic RV EF falls below 40% and the sub pulmonary ventricular systolic pressure rises above 50 mm Hg. The present study agrees with the above study. Unfavorable outcomes were seen in 2 patients requiring TVR. The other 2 patients who underwent TVR did not show TV dysfunction postoperatively. Two patients who survived after TVR had good RV EF preoperatively and were operated early. Patients who did not survive after TVR reported depressed RV function preoperatively. In a study by Hraska et al[11], TV dysfunction occurred it 36(42%) of 86 patients with available data who underwent different procedures. TVR had worse outcomes compared to other surgeries. Causes of TV regurgitation include annular dilatation and high chances of morphologic TV abnormalities. Post-operative RV dysfunction contributes to TR.

In the present study one patient in VSD closure group developed CHB within 1-5 years postoperatively eventually required permanent pacemaker implantation. One Patient of VSD closure group developed severe TR 8 months after surgery and required TVR. One patient of VSD closure left AV valve + developed severe TR 8 months after surgery and required TVR. One patient of VSD closure + left AV valve repair group developed severe TR at 6 years postoperatively and required TVR.

Hiramatsu and colleagues [14]conducted a study of patients of CCTGA who underwent the double-switch operation, which comprised of atrial switch plus intraventricular rerouting in 72 patients (Group-I) and an atrial switch plus arterial switch in 18 patients(group-2). They

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reported 11 reoperations(10 in group-1 and1 in group 2). They reported 17 reinterventions in group I.

Horer and colleagues[12] reported lower freedom of operation in anatomic repair as compared to conventional repair. Shin'oka and colleagues[6] reported freedom from reoperation of 64.2% in conventional group and 76.6 % in anatomic group. Ther1 was no statistical difference between the two group.

Average 10 year survival of available patients was 85%. Time related survival of patients was studied after surgery till December 2021. The lowest 5 year and 10 year survival rates were seen in TVR group (50%). Five year survival of patients of VSD was l00%. 2 patients who underwent VSD closure were operated in 2018. Hence average 6-10 year survival study for this group was not feasible. Five year survival of ASD closure patients was I00%. One closure was operated in 2018. Hence ten year survival was not feasible. Ten year survival of pulmonary valvotomy group was 100% each. Five year survival of patient who underwent VSD closure + left AV valve repair was 100%. This patient developed tricuspid valve regurgitation six years post-surgery and underwent colleagues survival valve replacement and expired. Ten year survival of the patient who pulmonary valvotomy + TVR was 100%.
Sano et al[13] reported a 10 year survival after conventional repair as 83%.

Survival was not significantly different between patients who underwent anatomic or "classic repair or univentricular palliation 83.3 +/- 15.2%, 79.7% +/- 6.9%, 90.9 +/- 8.7% at 10 years, respectively according to Horer and colleagues. According to Shin' oka et al [6] no statistical difference between the anatomic and conventional repair groups was observed in regard to l0 year time related survival.

Two patients who underwent TVR had RV dysfunction preoperatively and did not survive . Two patients developed RV dysfunction postoperatively. Among them one patient had undergone VSD closure and developed RV dysfunction and severe TR 8 months post operatively. Patient eventually needed TVR. One patient who underwent VSD closure left Av valve repair developed RV dysfunction and severe TR six years surgery and underwent TVR eventually and expired.

Cause tor RV dysfunction postoperatively in the present study can probably be development of severe TR. A combination of depressed RV ejection function with severe TR suggests that the systemic RV is functioning with afterload with afterload mismatch with elevated afterload due to an inappropriate compensation against of severe volume overload with or without impaired myocardial contractility. 18 patients had good RV function preoperatively and freedom from postoperative RV function among them was 88.88% in the present study. In study by Hong Hsu et al[9], out of 13 survivors of conventional group 5 patients developed systemic dysfunction. Out of 13 survivors of anatomical repair group, 5 patients developed systemic dysfunction. Left ventricular dysfunction remains a risk after anatomic repair. Due to incidence of eft LV dysfunction, residual mitral and aortic valve regurgitation and arrhythmia, anatomical repair cannot be labelled as an ideal management of CCTGA.

CONCLUSION

We have studied and analyzed patients of CCTGA who underwent surgical management of associated cardiac defects by conventional repair and have assessed these patients regarding intraoperative and postoperative complications, mortality, time related survival and

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postoperative RV dysfunction. Surgical management of CCTGA by conventional repair provides good comparable results with anatomical repair with respect to post-operative morbidity, mortality and long term time related survival. 3) However caution should be maintained in patients with preoperative severe TR. & Early repair of TV valve should be done before the development of RV dysfunction. This study had a small sample size, these findings need further evaluation in a bigger study.

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