Journal of Cardiovascular Disease Research

ISSN: 0975-3583,0976-2833 VOL14, ISSUE 08, 2023

Congenitally Corrected Transposition of Great Arteries: A Review of Eight Cases of The Complex CHD

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Received Date: 20/07/2023 Acceptance Date: 28/08/2023

Abstract

Background: Congenitally corrected transposition of great arteries (cc-TGA) is a rare congenital heart disease (CHD) characterized by atrioventricular and ventriculoarterial discordance along with ventricular inversion. It constitutes less than 1% of all congenital heart diseases. Theoretically, there is no functional abnormality provided that, there are no associated intracardiac defects. But unfortunately, most cases are complicated by intracardiac defects, AV conduction disturbances or arrhythmias. The present study focuses on the various types of presentations and management of eight cases of cc-TGA at our institute over a period of five years. **Methods:** All patients diagnosed with cc-TGA between 2011 and 2015 were retrospectively analysed for various presentations and management. Patients with unfavourable anatomy that precluded any kind of surgical repair were excluded from the study. **Conclusion:** Our review study emphasizes the fact that optimal surgical management of patients with cc-TGA, with or without associated cardiac anomalies, need to be tailored according to the symptoms, clinical assessment, anatomy of the cardia, severity of pulmonary stenosis in association with a VSD, functional capacity of the systemic and pulmonary ventricles and systemic AV valve regurgitation status.

Key words: Congenitally corrected transposition of great arteries (cc-TGA), Atrioventricular discordance, Ventriculoarterial discordance

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Introduction

Congenitally corrected transposition of great arteries (cc-TGA) is a complex congenital cardiac condition involving discordant atrio-ventricular and ventriculo-arterial connections. The morphological right atrium is connected to the morphological left ventricle by mitral valve, and the left ventricle is connected to the pulmonary artery by the pulmonary valve. The morphological left atrium is connected to the morphological right ventricle by a tricuspid valve, and the right ventricle is connected to the aorta by the aortic valve. As a result of discordant connections at both levels, blood flows in a “congenitally corrected” physiological path despite the anatomical derangements.

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Etiology

As with almost all forms of congenital heart disease, the causes are thought to be multifactorial. Most of the clinical and surgical retrospective studies have reported a male predominance in corrected transposition.[1,2] A recent study suggested an autosomal recessive mechanism of transmission may be present in some families.[3] Interestingly, they found that transposition of the great arteries was the most common recurrent defect in families with congenitally corrected transposition suggesting a patho-genetic link between these two entities.

Relevant Anatomy

Knowing the position of the atrioventricular (AV) node in this defect is extremely important if injury to it during surgery is to be avoided. Because of the L-looping of the ventricles in this condition, the usual posterior position of the AV node is prevented from reaching the interventricular septum because of the malalignment of the atrial and inlet ventricular septa. An anterior node is present, either alone or in addition to the posterior node and it is located in the floor of the right atrial wall immediately anterolateral to the interatrial septum. This gives rise to an AV bundle that penetrates the fibrous annulus to make contact with the ventricular myocardium. It then passes anterior to the pulmonic annulus along the morphologic left ventricular side of the septum and subsequently courses anterior and superior to a perimembranous outlet ventricular septal defect (VSD).[4,5]

This anterior position of the AV node is more commonly reported in situs solitus and situs inversus, a posteriorly positioned AV node has been described.[6] However, other authors stress that either position of the AV node is possible with any situs.[7]

The coronary arteries arise from the aortic valve sinuses adjacent to each ventricle. The morphologic left coronary artery arises from the right-sided posterior sinus, and the morphologic right coronary artery arises from the left-sided posterior sinus. The course of the coronaries is always anterior to the pulmonary artery. The most common coronary artery abnormality is a single coronary artery that arises from the right-sided posterior sinus.[8]

Pathophysiology

Pathophysiology is determined by the presence and type of associated lesions. When no other defects are present, the path of the blood flow is physiologic; blood from the left atrium enters the right ventricle and is then directed into the aorta, and, on the right side, the deoxygenated blood from the vena cava enters the left ventricle. Because of the ventriculoarterial discordance, the deoxygenated blood is then directed into the pulmonary artery. Thus, the oxygen saturations in the heart chambers and in the great arteries are normal.

The most common anatomic associations include the presence of a ventricular septal defect (VSD), which may be observed in almost 80% of cases and the presence of pulmonary stenosis, which has been reported in approximately 50% of cases,[2] the presence of a VSD causes a systemic-to-pulmonary shunt; however, this is usually balanced because of the protective effect of coexisting pulmonic stenosis.

Tricuspid valve anomalies, including dysplasia, straddling, or Ebstein-like malformation (with or without regurgitation) are also quite common and are reported in 14-56% of patients.[1,2] Tricuspid regurgitation in this setting of a systemic ventricle, which is the morphologic right ventricle, is much more ominous than it would be in an otherwise normal heart. Coarctation and interrupted aortic arch have also been frequently reported, but subvalvar and valvar aortic stenosis are quite uncommon.[2,10] Conduction abnormalities also are common. The reported incidence of complete atrioventricular (AV) block has ranged

ISSN: 0975-3583,0976-2833 VOL14, ISSUE 08, 2023

from 12-33%.[11,12] Spontaneous progression of AV block has been reported to occur at a rate of 2% per year.[11,13] Additional rhythm problems include Wolff-Parkinson-White syndrome, supraventricular tachycardia, atrial flutter, and atrial fibrillation.[1,12,14]

Presentation

Patients with isolated corrected transposition of the great arteries may present in adulthood because of abnormal radiography or ECG findings and may have no symptoms, at least for the first 3 or 4 decades of life.

In a study of 18 patients, Presbitero et al found that rhythm disturbances and tricuspid regurgitation were present more frequently after the third decade of life.[15] They found that this and impaired right ventricular function developed in 66% of patients older than 50 years, causing congestive cardiac failure.

A multi-institutional study confirmed that congestive cardiac failure is common in patients with or without associated cardiac defects.[16] By age 45 years, 67% of patients with associated anomalies and 25% of patients without associated anomalies were in congestive cardiac failure.

Another large study by Rutledge et al confirmed that survival rates are reduced in these patients.[17] They found poor right ventricular function and complete AV canal as risk factors for mortality. Risk factors for progressive right ventricular dysfunction included conventional biventricular repair, complete AV block, and severe tricuspid regurgitation.

Most patients who have associated anomalies present in infancy with a murmur or heart failure. Patients with bradycardia secondary to complete AV block can present at any age. Unless these patients have pulmonary atresia or severe pulmonic stenosis, cyanosis is not present.

Aim of the Study

This review study focuses on the presentation, evaluation, and management of eight cases of cc-TGA at our institute over a period of five years between 2011-2015.

ISSN: 0975-3583,0976-2833 VOL14, ISSUE 08, 2023

CASE REPORTS

Case No.	Presentation	Diagnosis	Operative findings	Procedure	Aim of the procedure	Case of death
1	• 14year old male • Diagnosed to have CHD at 8 years of age on routine examination. • Recurrent chest infection since infancy. • Cyanotic spells since 2 months of age. • Squatting episodes present. • Easy fatigability and squatting noticed since the time he was able to walk. • Delayed milestones.	CHD, SLL, cTGA, sub-pulmonic VSD, PS	• Right lower lobe hypoplastic. • Only RSPV and RMPV drain into LA. • Left pulmonary veins normal. • PVs - LA - TV - RV - Aorta (Aorta to the left and anterior) • RSVC, LSVC, IVC - RA - MV - LV - PA (PA to the right of aorta and posterior) • ASD (8 mm) • Sub-pulmonic VSD (10 mm)	Senning and Rastelli (double switch operation), RV to PA conduit using pulmonary homograft.	Anatomical correction and long term palliation.	-
2	• 16 year old female • Child to non-consanguineous parentage. • FTND • Diagnosed to have cardiac disease at the age of 12 years at a school health check-up camp. • Easy fatigability since the age of 2 years. • Dyspnoea on exertion – 3 year duration. • Cyanosis since infancy.	CHD, SLL, cTGA, sub-aortic VSD (bidirectional shunt), valvar PS, good sized PAs, mild systemic AV valve regurgitation.	• Right lower lobe hypoplastic. • Only RSPV and RMPV drain into LA. • Left pulmonary veins normal. • PVs - LA - TV - RV - Aorta (Aorta to the left and anterior) • SVC, IVC - RA - MV - LV - PA (PA to the right of aorta and posterior) • ASD (10 mm) • Sub-aortic VSD (25 mm) • Pulmonary valvar stenosis	VSD closure through aorta. Pulmonary valvotomy. Direct closure of ASD.	Long term palliation.	-

ISSN: 0975-3583,0976-2833 VOL14, ISSUE 08, 2023

3	• 2 year old male • Child to second degree consanguineous parentage. • FTND • Diagnosed to have cardiac disease at 1 year 3 months while evaluating for recurrent chest infections and was advised surgery. • Admitted with failure and treated medically for a month and was on ventilator for some time. • Cyanosis noticed since 1 month of age.	CHD, SS, cTGA, dextrocardia, Anomalous systemic venous connection (LSVC to common pulmonary venous chamber), Cardiac TAPVC, OS-ASD.	• Aorta anterior and to left and arising from RV. • MPA posterior and to the right, arising from LV. • Good sized branch Pas. • RA in midline and posterior; recognized by IVC connection. • LA anteriorly located with its typical appendage. • All pulmonary veins drain to common posterior chamber adjacent to RA and to the left (common chamber draining to RA). • LSVC drains to common pulmonary venous chamber in its most cephalad position.	Venous Switch Operation LSVC blood redirected to RA by creating a tunnel between common chamber to RA (margins of ASD); hence common chamber blood automatically drains only to LA; Pericardial patch augmentation of LA and common venous chamber done.	Anatomical correction not technically feasible and hence procedure is aimed at long term palliation.	Low cardiac output state due to biventricular failure status post open heart surgery in a case of CHD, SS, Dextrocardia, cTGA, Anomalous systemic venous connection (LSVC to common pulmonary venous chamber), Cardiac TAPVC, OS-ASD.
4	• 6 year old male • Dyspnoea on exertion since 2 months of age. • Cyanosis since 2 months of age. • No cyanotic spells or feeding difficulty. • No developmental delay.	CHD, SLL, cTGA, Large sub-pulmonic VSD, Infundibular PS, OS-ASD.	Large VSD, not routable to aorta.	Initial plan was double switch v/s Senning and Rastelli • Large VSD, not routable to aorta and not able to close through aorta	Anatomy not suitable for double switch or Senning and Rastelli and hence the procedure was aimed at long term palliation.	-

ISSN: 0975-3583,0976-2833 VOL14, ISSUE 08, 2023

				as it was remotely located. Hence, closed through PA. • Epicardial pacing done in view of post procedural CHB. • Permanent pacemaker implantation done 1 week later.		
5	• 6 year old male • First born child to second degree consanguineous parentage. • FTND • Palpitations noted since birth. • Evaluated and found to have cardiac disease.	CHD, SS, Dextrocardia, cTGA, sub-aortic VSD, Bicuspid PV with commissural fusion, valvar PS, sub-pulmonic membrane, LSVC.	• Aorta anterior and arising from morphological RV. • Pulmonary artery posterior and arising from morphological LV. • Pulmonary valve leaflets were thickened with commissural fusion.	Pulmonary valve commissurotomy, sub-pulmonic membrane excision, VSD closure through aorta.	Closure of the ventricular level shunt; relief of LVOTO and long term palliation.	-
6	• 17 year old male • First born child to non-	CHD, SLL, Dextrocardia,	• Aorta anterior, PA posterior. • mLV was posterior to mRV.	Bidirectional Glenn shunt done	Long term palliative	-

ISSN: 0975-3583,0976-2833 VOL14, ISSUE 08, 2023

	consanguineous parentage. • FTND • Diagnosed to have cardiac disease at 3 months of age while evaluating for cyanosis. • Cyanotic spells present.	cTGA, DORV, VSD, PS, LSVC.	• mRV formed the entire anterior surface of the cardiac mass. • PA annulus was hypoplastic. • RPA arid LPA of adequate size.	off CPB after studying the anatomy. PAP – 15/9 mmHg (Mean – 13 mmHg).	surgeries possible in this case were: • VSD closure + RVOT resection; not done as there was hypoplastic PA. • VSD closure + mLV – PA conduit; technically not possible in view of extreme posterior position of mLV. • Intraventricular tunneling of LV to aorta + Rastelli – not done as mRV to PA homograft would be compressed by the sternum. Bidirectional Glenn shunt is	

ISSN: 0975-3583,0976-2833 VOL14, ISSUE 08, 2023

					the only possible palliative surgery in this situation if PAP is <15 mmHg.	
7	• 3 year old female • First born child to non-consanguineous parentage. • Delivery by C-section; cried immediately after birth. • Dyspnoea on exertion and easy fatigability since 2 months (restricted playing activities). • No cyanosis/squatting episodes. • No h/o recurrent LRTI.	CHD, SI, Mesocardia, cTGA, large sub-pulmonic VSD (bidirectional shunt), infundibular and valvar PS.	• ILD (van Praagh) • IVC in midline • RA to left and LA to right • RA-MV-LV-PA • LA - TV - RV - Aorta • Aorta rightward and anterior; PA posterior and leftward to aorta • Pulmonary valvar and annular stenosis • Good sized PAs • Sub-arterial VSD (10 mm) • Large PFO (8 mm) As RV was more anterior due to mesocardia, routing of homograft from RV to PA was not possible without compression or kink; hence decided to VSD closure + LVOT repair rather than double switch.	VSD closure through aorta. Infundibular resection through RA. Pulmonary valvotomy + infundibulotomy (trans RA and mitral valve). Post procedural CHB was managed by epicardial pacing.	Long term palliation and relief of symptoms.	-
8	• 32 year old male • Fourth born child to second degree consanguineous parentage. • FTND	CHD, SI, IDD, Mesocardia, cTGA, mid muscular and inlet VSD	• IDD; PV-LA-TV-RV-Aorta • Aorta to the right • SVC, IVC, Hepatic veins - RA - Mitral valve - LV - PA • PA to the left of aorta • 15 mm sub-pulmonic VSD	VSD closure through RA; LV to PA pulmonary homograft conduit. Postoperative	Surgery for long term palliation and relief of symptoms.	-

ISSN: 0975-3583,0976-2833 VOL14, ISSUE 08, 2023

	• Diagnosed to have cardiac disease at the age of 3 months while evaluating for chest infection. • Easy fatigability since childhood. • Dyspnoea on exertion since childhood.	(bidirectional shunt), Severe valvar PS, interrupted IVC with azygos continuation, second degree heart block.	• 15 mm posterior muscular VSD • Pulmonary valvar stenosis • MPA, LPA, RPA – good sized • 5 mm PFO	CHB managed by pacing.		

ISSN: 0975-3583,0976-2833 VOL14, ISSUE 08, 2023

Discussion

c-TGA is a complex congenital heart defect characterized by atrioventricular and ventriculo-arterial discordance, and in which mRV cannot support the systemic circulation because it becomes overloaded. Patients need early intervention to avoid progressive dilatation of the mRV and aggravation of TR, which could lead to a vicious circle and eventually result in RV failure or severe TR. The optimal management for these patients remains controversial. Conventional repair, still leaving the mRV in the systemic circulation, comprises VSD repair, tricuspid valve replacement or plasty and pulmonary valvuloplasty. Conservative management, such as these, is still associated with a high incidence of late progressive mRV dysfunction and TR. Anatomical correction may be superior to conventional repair in terms of higher survival rate and better living quality.

Interest has recently been expressed in a more anatomic repair that permits the morphological LV to function in the systemic circulation. This strategy would have the theoretical advantage of relieving the hemodynamic burden on the RV and tricuspid valve, potentially improving surgical results and longevity. Anatomic repair can be accomplished by using a venous switch procedure (either the Mustard or Senning technique), and, in those with a normal LV outflow tract, the arterial switch operation. For patients with a VSD, a patch can be inserted to tunnel the LV flow into the aorta, and the morphological RV is connected to the pulmonary artery via a conduit (Rastelli technique). Before anatomic repair, the LV needs to be "prepared" to function as a systemic ventricle, and therefore, in the absence of pulmonary stenosis that would have "trained" the LV to hypertrophy and function at a higher pressure than a venous ventricle, pulmonary banding may be performed. Concomitant tricuspid valve surgery can also be performed if necessary.

Given the complexity of these operations, however, the early mortality appeared encouraging in high volume and experienced centres, but several reinterventions had to be performed. Patients with the Rastelli procedure may need a repeat conduit or aortic valve replacement. Percutaneous balloon dilatations for baffle obstruction or pulmonary stenosis may also be needed.

In addition, atrial arrhythmias are common after venous baffle procedures and increase with longer follow-up. Whether older patients are suitable candidates for these procedures remains to be determined because the idea that the LV can be "trained" after childhood remains controversial. Some authors have reported failure of the LV after pulmonary banding even before adolescence. Long-term follow-up will be necessary to determine whether the various surgical management strategies confer a survival advantage over the natural history and to determine the preferred surgical procedure for a given situation.

Conclusion

Our review study emphasizes the fact that optimal surgical management of patients with cc-TGA, with or without associated cardiac anomalies, need to be tailored according to the symptoms, clinical assessment, anatomy of the cardia, severity of pulmonary stenosis in association with a VSD, functional capacity of the systemic and pulmonary ventricles and systemic AV valve regurgitation status. The postoperative cases are being followed up regularly for assessment of ventricular function, systemic AV valve status and conduction system of the heart. The scope for further management during postoperative follow up, if indicated and feasible, will be reviewed timely.

ISSN: 0975-3583,0976-2833 VOL14, ISSUE 08, 2023

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