Incidence of dysrhythmias in congenitally corrected transposition of the great arteries

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SUMMARY: Kafalı G, Elsharshari H, Özer S, Çeliker A, Özme Ş, Demircin M. Incidence of dysrhythmias in congenitally corrected transposition of the great arteries. Turk J Pediatr 2002; 44: 219-223.

We reviewed hospital records of 45 children with corrected transposition of the great arteries (c-TGA) to determine the incidence and outcome of congenital and postoperative dysrhythmias seen in this congenital anomaly.

Our study comprised 45 patients (12 girls, 33 boys). The mean age of the patients at initial evaluation was 3.4 ± 3.7 years, and they were followed for a mean period of 3.5 ± 4 years.

Forty-three patients (95%) with c-TGA had associated intracardiac defects. Two patients (5%) did not have any cardiac defects. In 31 (69%) of the 45 patients, ventricular septal defect (VSD) was present, while the remaining 14 patients (31%) had intact ventricular septum. VSD repair was done in 17 of 31 patients.

Different types of dysrhythmias were detected in 19 of 45 patients. Six patients (13%) presented initially with congenital complete atrioventricular block (AVB) and five patients with postoperative complete ve AVB. Pacemaker implantation was required for 11 patients with complete AVB. In eight patients, ventricular and supraventricular ectopic beats, left bundle-branch block (LBBB) and first-degree AVB were determined but therapy was not required.

Twenty-five (58%) of 43 patients with intracardiac defects underwent different surgical procedures. Permanent pacemaker implantation was required for five patients (29%) after VSD repair (17 patients) due to postoperative complete AVB. The incidence of congenital AVB in 14 patients with intact ventricular septum was found to be high (29%) in comparison to the group with VSD (6%).

Patients diagnosed as c-TGA with or without cardiac defects should be followed carefully during their clinical course to identify and treat different types of dysrhythmias that can appear at any time.

Key words: corrected transposition, complete heart block, ventricular septal defect.

In congenitally corrected transposition of the great arteries (c-TGA), there is a discordant atrioventricular connection (right atrium to left ventricle and left atrium to right ventricle), and a discordant ventriculoarterial connection (left ventricle to pulmonary artery and right ventricle to aorta). Since it is physiologically a corrected transposition, the associated anomalies [ventricular septal defect (VSD), single ventricle, pulmonary stenosis (PS), left atrioventricular valve malformations] and conduction defects determine the prognosis.

Ventricular inversion frequently is associated with conduction disturbances ranging from first-degree atrioventricular block (AVB) to complete AVB. Complete AVB may appear spontaneously or during surgical treatment of associated defects. Approximately 10% of the patients present initially with complete AVB. First-degree or second-degree AVBs also have the tendency to progress to complete AVB with increasing age. In postoperative complete AVB, pacemaker therapy is always indicated. Congenital complete AVB may also not be well

tolerated; pacemaker therapy is recommended early after the recognition of complete AVB[1-3]. Atrial arrhythmias and Wolff-Parkinson-White syndrome are occasionally present[4-6].

In this study, we reviewed our patients with c-TGA with regard to associated intracardiac defects and their surgical management, types of dysrhythmias and follow-up results. We stress the importance of close follow-up in all patients with or without cardiac defects, to recognize and treat the different types of dysrhythmias which can appear at any time.

Material and Methods

In this study, 45 children (12 girls, 33 boys) diagnosed as c-TGA between January 1989 and December 1999 at Hacettepe Children's Hospital, Department of Pediatric Cardiology were retrospectively evaluated. The diagnosis of c-TGA was made in all patients by both two-dimensional and Doppler echocardiography, and then cardiac catheterization and biplane angiography were performed in 31 patients. All the patients were evaluated regarding dysrhythmia by 12-lead surface electrocardiogram and 24-hour ambulatory electrocardiographic monitoring at intervals of six to 12 months. The ages of the patients at the time of diagnosis ranged from 2 days to 11 years (mean 3.4 ± 3.7 years) and they were followed for a mean period of 3.5 ± 4 years (range 3 days to 12 years). In all patients, the following parameters were evaluated: a) cardiac defects and their surgical management, b) types of dysrhythmias, c) follow-up results of dysrhythmia.

Of 45 patients with c-TGA, 43 (95%) had associated intracardiac defects (Table I). Two patients (5%) did not have any cardiac defect. One of these patients had dextrocardia and the other had a history of chest pain.

Twenty-five (58%) of 43 patients with intracardiac defects underwent different surgical procedures (Table II). The mean age of these patients at operation was 4.4 ± 4.2 years (range 2 months to 14 years). VSD was present in 31 (69%) of the 45 patients while 14 patients (31%) had intact ventricular septum. VSD repair was done in 17 (55%) of the 31 patients. Because of secondary pulmonary hypertension, pulmonary artery banding was also performed in five (2 patients had Ebstein's anomaly) of

Table I. Associated intracardiac lesions in 45 patients with corrected transposition of the great arteries

Intracardiac lesions	n	%
Ventricular septal defect	31	69
Left AV valve regurgitation	30	66
Pulmonary stenosis	14	31
Right AV valve regurgitation	8	17
Ebstein's anomaly	6	13
Atrial septal defect	5	11
Dextrocardia	5	11

AV: atrioventricular

Table II. Surgical procedures in 25 patients with corrected transposition of the great arteries

Surgical procedures	n
Ventricular septal defect repair	17
Left AV valve replacement	6
Atrial septal defect repair	5
Pulmonary artery banding	5
Pulmonary commissurotomy	3
Blalock-Taussig shunt	3
Left ventricle-pulmonary artery conduit	1

AV: atrioventricular

17 patients with VSD repair. Left AV valve replacement was performed in six (29%) (2 patients had Ebstein's anomaly) of 30 patients due to the left AV valve regurgitation. In 14 patients with PS, pulmonary commissurotomy (3 patients), Blalock-Taussig shunt (3 patients) and left ventricle-pulmonary artery conduit (1 patient) were done. Atrial septal defect repair was performed in five patients. All the patients were examined at intervals of six to 12 months for dysrhythmia.

Different types of dysrhythmias were determined in 19 of 45 patients (Table III). Ventricular and supraventricular ectopic beats, left bundle-branch block (LBBB) and first-degree AVB were determined in eight patients who had no complaint related to dysrhythmia and normal screening with 12-lead surface electrocardiogram. They were followed without requiring any treatment in their follow-up period.

Six patients had congenitally complete AVB when first seen. Three of them had a history of syncope and/or fatigue and one patient had fetal bradycardia. The ages of six patients (13%)

Table III. Dysrhythmias in 19 of 45 patients with corrected transposition of the great arteries c-TGA

Types of dysrythmia	Patient (n: 19)
First degree AVB + supraventricular ectopic beats	1
Supraventricular ectopic beats + left bundle-branch block	1
Left bundle-branch block	1
Supraventricular ectopic beats	2
Ventricular ectopic beats	3
Postoperative AVB	5
Congenital AVB	6

AVB: atrioventricular block

with congenitally complete AVB at the time of diagnosis ranged from 2 months to 11 years (mean 2.8 ± 4.2 years). Two of them had VSD and pulmonary hypertension for which pulmonary artery banding was performed and four had intact ventricular septum. One of the patients with intact ventricular septum had ASD which was repaired. Three other patients had left AV valve regurgitation, for which one had undergone valve replacement.

Postoperatively complete AVB developed in five (29%) of the 17 patients with VSD repair, and their mean age at the operation time was 4.9 ± 5.4 years (range 5 months to 14 years). Two of five patients had undergone both pulmonary commissurotomy procedure and VSD repair. Eleven patients with congenital (6 patients) and postoperative complete AV block (5 patients) underwent permanent pacemaker implantation.

The incidence of congenitally complete AVB in the patients with intact ventricular septum was found to be higher (29%) in comparison to the group with VSD (6%) (Table IV). The incidence of normal sinus rhythm among patients with VSD was found to be higher than among those with intact ventricular septum (94% and 71%, respectively).

Follow-up: An eight-year-old patient with the diagnosis of Eisenmenger syndrome as a result of VSD and pulmonary hypertension was followed with only drug therapy. A newborn patient with Ebstein's anomaly and VSD died of sepsis.

Two patients who had undergone VSD repair died suddenly on the postoperative first day because of probable dysrhythmia. One of the patients had also undergone left ventricle-pulmonary artery conduit procedure.

At the time of last follow-up, 41 of 45 patients were in New York Heart Association Functional Class I - Class II.

Discussion

Ventricular inversion with L-transposition of the great arteries occurs in 1.4% of congenital cardiac defects[2]. The most commonly associated intracardiac defects are VSD (50-80%), PS (45-53%) and left atrioventricular valve regurgitation (18-90%)[1-3,7-9]. In our study, we found incidence of VSD as 69%, incidence of PS as 31% and incidence of left AV valve regurgitation as 66%.

In c-TGA, usually the posterior AV node fails to connect to His' bundle. An accessory anterior AV node is present, anteriorly adjacent to the pulmonary and mitral annuli. This node gives rise to the bundle of His, which penetrates the mitral annulus and then passes into the anterior aspect of the roof of the pulmonary outflow tract and reaches the ventricular septum. In the specimens with intact septum. His' bundle is

Table IV. Classification of 11 patients with congenitally complete AVB according to VSD

	n	Sinus rhythm n (%)	Congenital AVB n (%)
VSD	31	29 (94%)	2 (6%)
Intact ventricular septum	14	10 (71%)	4 (29%)
Total	45	39 (87%)	6 (13%)

AVB: atrioventricular block; VSD: Ventricular septal defect

anterior to the membranous septum and intervenes between the pulmonary outflow tract and left-sided ventricle. In the specimens with defects, the bundle is related to the right side of the anterior rim of the defect and bifurcates to give rise to inverted bundle branches[10-11]. Avoidance of these areas in surgical repair of VSDS and correction of subvalvar pulmonary stenosis in such cases should reduce the risk of traumatic heart block.

Furthermore, as a result of developing fibrosis in this precariously positioned AV bundle, congenital AV conduction abnormalities may occur with increasing age. Huhta et al.[12] reported that the risk of complete AVB increased linearly with age at a range of approximately 2% per year. The incidence of complete AVB in the median 10 year follow-up period is as high as 30%[12,13]. The patients reported previously had been followed from childhood to late adulthood. Congenital complete AVB in our series was 13% in the median 3.5 years (range 3 days to 12 years). The incidence of congenital complete AVB in patients with intact ventricular septum was higher than in those with VSD (29% and 6%, respectively). Similar results were also reported by Huhta et al.[12] and Daliento et al[13]. These results may be due to the longer life expectancy in these patients than in those with complicated c-TGA or to the greater mechanical stress exerted on the conduction system when the ventricular septum is intact[10,12,13].

The incidence of postoperative complete AVB in 17 patients who had VSD repair was 29% (5 patients). Two of five patients with VSD repair also underwent pulmonary commissurotomy. Previous studies have reported incidence of postoperative complete AVB as 24-26% in patients with VSD repair[12,14]. Furthermore, McGrath et al.14 observed that older age at repair was a risk factor for the development of heart block in patients with ventriculoarterial connections other than discordant ventricouloarterial connection, in which atrioventricular valve chordae may straddle or attach to the edge of the VSD. This may be due to a morphologic variability in the location of the AV node and His' bundle that makes patients with c-TGA less likely to develop complete heart block.

In congenitally c-TGA, first-and second-degree AV block may precede complete AV block, as histologic study has revealed that fibrosis of the His bundle is a typical finding of acquired complete AV block[1,12,13]. Therefore, first-and second-degree blocks in patients with c-TGA must be followed carefully. One patient had first-degree AV block and was followed up without progressing to complete AV block.

In congenitally c-TGA, atrial arrhythmias and Wolff-Parkinson-White syndrome are occasionally present[4,6]. We observed ventricular and supraventricular ectopic beats in seven of 19 patients with dysrhythmia. Ventricular ectopic beats were fewer than 10 to 20 per hour, and were evaluated as infrequent[15]; no treatment was necessary for them.

We conclude that patients with c-TGA are at increased risk of developing complete AV block throughout their lives. Careful follow-up of these patients may be benefical in preventing poor prognosis resulting from dysrhythmia. Indication for the implantation of a permanent pacemaker should be considered in symptomatic children because of the high incidence of sudden cardiac deaths.

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