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Outcome of 121 Patients with Congenitally Corrected Transposition of the Great Arteries

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Abstract. Congenitally corrected transposition of the great arteries (ccTGA) is a rare disorder with reduced survival that is influenced by the presence of associated anomalies, tricuspid regurgitation (TR), and right ventricular (RV) function. The double switch procedure has been proposed as an aggressive surgical approach in selected patients. We sought to review our experience with conventional repair to determine if a change in surgical strategy was warranted. Clinical records of 121 patients with ccTGA and two adequate-sized ventricles were retrospectively reviewed. Median length of follow-up was 9.3 years; 5-, 10-, and 20-year survival rates were 92%, 91%, and 75%, respectively. Surgery was performed in 86 patients, including conventional biventricular repair in 47 patients. Risk factors for mortality by univariate analysis included age at biventricular repair ($p = 0.04$), complete atrioventricular (AV) canal defect ($p = 0.02$), dextrocardia ($p = 0.05$), moderate or severe TR ($p = 0.05$), and poor RV function ($p = 0.001$). By multivariate analysis, complete AV canal defect ($p = 0.006$) and poor RV function ($p = 0.002$) remained significant as risk factors for mortality. Risk factors for the development of significant TR included conventional biventricular repair ($p = 0.03$) and complete AV block ($p = 0.04$). Risk factors for progressive RV dysfunction included conventional biventricular repair ($p = 0.02$), complete AV block ($p = 0.001$), and moderate or severe TR ($p < 0.001$). This is the largest nonselected cohort of patients with ccTGA followed at a single center. Our results confirm that significant TR and poor RV function are risk factors for poor outcome and provide convincing evidence that patients undergoing conventional biventricular repair are at higher risk for deterioration of tricuspid valve and right ventricular function compared to palliated or unoperated patients. We support a move toward an alternative surgical approach (double switch procedure) in carefully selected patients.

Key words: Congenitally corrected transposition — Biventricular repair

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Atrioventricular and ventriculoarterial discordance, commonly known as congenitally corrected transposition of the great arteries (ccTGA), comprises <1% of all forms of congenital heart disease and represents a clinically heterogeneous patient population with frequently associated anomalies of ventricular septal defect (VSD), pulmonary stenosis (PS) or pulmonary atresia (PA), and Ebstein-like malformation of the tricuspid valve [2, 3, 6, 7, 20, 24]. The natural history is highly variable and is related to the presence and severity of these associated anomalies [16, 17, 20]. In general, long-term survival is possible [20]; however, in the presence of reduced systemic ventricular function and tricuspid regurgitation (TR), survival is significantly reduced [12]. Conventional biventricular repair, in which the morphologic right ventricle (RV) and tricuspid valve (TV) support the systemic circulation, has largely been reserved for symptomatic patients due to the unique location of the cardiac conduction tissue [2, 4], poor outcome of TV repair [26, 21], and concerns regarding earlier development of systemic ventricular dysfunction and TR following

Table 1. Patient characteristics (121 patients)

Male:female	80:41
Median age at diagnosis	1 month (28 months; prenatal–49 years)[a]
Asymptomatic ($n = 21$)	42 months (92 months; prenatal–49 years)
Congestive heart failure ($n = 44$)	1 month (23 months; prenatal–26.1 years)
Cyanotic ($n = 56$)	0.3 months (9.4 months; prenatal–8 years)
Median length of follow-up	9.3 years (10.9 years; 0.06 months–50.8 years)
Median age of survivors ($n = 101$)	12 years (13.3 years; 0.5 months–53 years)
Median age of death ($n = 20$)	13.2 years (11.3 years; 2 days–42.8 years)
Median age of transplant ($n = 5$)	30 years (32 years; 20–48 years)
Median age at intracardiac repair ($n = 47$)	4.9 years (8.2 years; 2 months–45.9 years)
Median age at first palliation ($n = 56$)	3 months (1.5 years; 1day–19.2 years)

[a] (Mean; range).

repair [1, 9, 13, 17–19, 21, 28, 29]. Due to the disappointing results following conventional surgery, the double switch operation has been proposed as an alternative surgical strategy. The advantage of the double switch procedure is the return of the morphologic left ventricle and mitral valve to the systemic circulation by an atrial level switch and either an arterial switch or Rastelli operation [14, 15, 16, 22, 28]. Controversy exists regarding optimal patient selection and timing for performing a double switch, and there are also concerns due to late complications of the atrial switch [25, 27]. We have therefore been cautious to adopt this surgical approach in our patient population. In this study we sought to review our single center experience to determine if a change in surgical strategy was warranted.

Methods

Clinical records of all patients with ccTGA and two adequate-sized ventricles followed at Texas Children's Hospital from January 1952 to July 1999 were reviewed. Patients with double-outlet right ventricle and pulmonary atresia were included. Patients without follow-up within the previous 2 years were contacted to establish present clinical condition. Patient outcome measures were analyzed to the time of the last clinic visit. Patient survival was analyzed to the time of death or cardiac transplantation. Survival was not assumed past the last clinic visit. For comparison purposes, patients were divided into three physiologic groups at presentation: asymptomatic, congestive heart failure, or cyanotic [17].

Tricuspid Valve

The TV was defined as Ebstein-like if there was excessive inferior displacement of the attachments of the septal and posterior leaflets from the atrioventricular (AV) junction into the cavity of the RV as noted by echocardiography, surgical inspection, or autopsy [5]. The valve was defined as "dysplastic" if the leaflets were abnormally formed or thickened and as "straddling" if chordae crossed through a VSD with attachments into the contralateral ventricle. For comparison purposes, all valves defined as Ebstein-like, dysplastic, or straddling were considered abnormal. The degree of TR was graded by echocardiography or angiography as described elsewhere [11, 23]. TR of moderate or severe degree was considered significant.

Systemic Ventricular Function

Systemic ventricular function was graded subjectively as normal or mildly, moderately, or severely reduced. Patients with moderately or severely reduced function were defined as having poor function. In all but one case (postoperative double switch procedure), this was the morphologic RV.

Statistical Analysis

Survival analysis was performed using the Kaplan–Meier method. Cox regression was used for comparison of survival curves with adjustment for covariates. Chi-square analysis or Fishers exact test were used to determine the relationship between categorical variables. Student's t-test was used to compare groups with respect to continuous variables. A p value ≤ 0.05 was defined as statistically significant.

Results

From January 1952 to July 1999, 121 patients with ccTGA were followed at Texas Children's Hospital. Thirty patients had not been seen in the previous 2 years, with data analyzed to the time of the last clinic visit. Patient characteristics are shown in Table 1. Median age at diagnosis for the entire group was 1 month but varied within physiologic groups ($p = 0.001$). All but 9 patients had an associated lesion (Table 2) with VSD being the most common, followed by PS. Figure 1 shows the physiologic group of patients at presentation as well as the proportion of patients in each group with an abnormal TV. Asymptomatic patients had isolated ccTGA (9), a small VSD (4), mild PS (5), or coarctation of the aorta (1) or were hemodynamically well balanced with a VSD and PS (2). Patients with heart failure had a large VSD (18), VSD and mild PS (16), or a regurgitant TV (10). Patients with cyanosis had a

Table 2. Associated cardiac defects

Defect	No.	Defect	No.
Isolated ccTGA	9	Atrioventricular block	59
VSD	88	First degree	24
Pulmonary stenosis	61	Second degree	10
Pulmonary atresia	18	Third degree	25
ASD	44	Congenital	8
DORV	18	Postoperative	8
Dextrocardia	37	*Situs inversus totalis*	10
CoA, hypoplastic arch/isthmus/IAA	16	Mitral valve abnormalities	7
Complete atrioventricular canal	8	WPW, Long QT, concealed pathway	7
Tricuspid valve abnormalities	34	TAPVR/PAPVR	4
Ebstein	19	Single coronary artery	2
Dysplastic	7	Subaortic stenosis	4
Stradding	8	Discontinuous PAs	3

ASD, atrial septal defect; CoA, coarctation of the aorta; ccTGA, congenitally corrected transposition of the great arteries; DORV, double-outlet right ventricle; IAA, interrupted aortic arch; PA, pulmonary artery; PAPVR, partial anomalous pulmonary venous return; TAPVR, total anomalous pulmonary venous return; VSD, ventricular septal defect; WPW, Wolff–Parkinson–White syndrome.

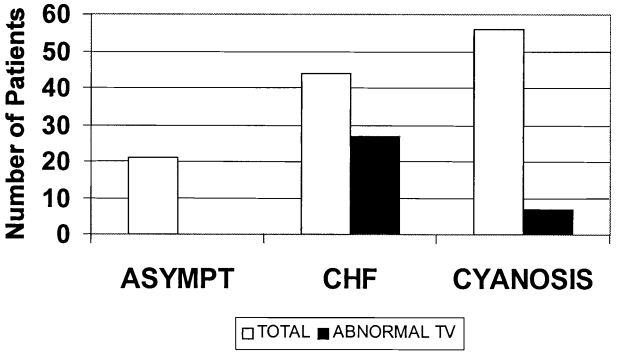


Fig. 1. Patient group according to presenting physiology and proportion of patients in each group with an abnormal tricuspid valve. *Asympt*, asymptomatic; *CHF*, congestive heart failure; *TV*, tricupid valve.

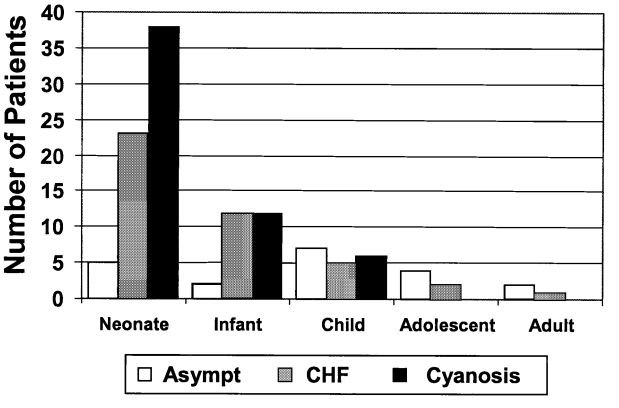
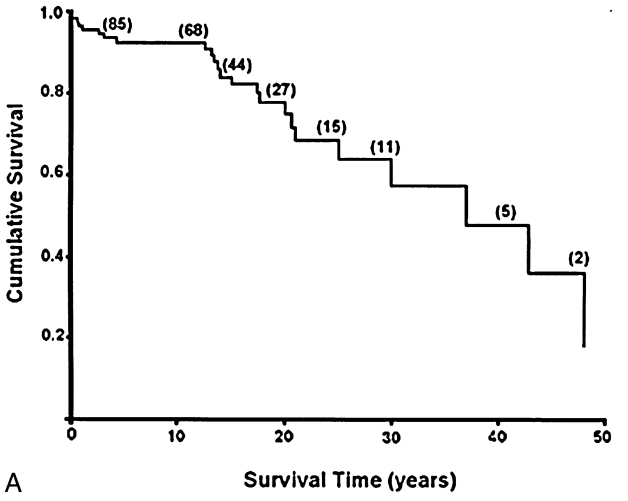


Fig. 2. Presenting features by age group. Age groups were defined as neonate (0–1 month), infant (1 month–2 years), child (2–10 years), adolescent (10–18 years), and adult (>18 years). *Asympt*, asymptomatic; *CHF*, congestive heart failure.

VSD and either PS (40) or PA (16). The mode of presentation varied within patient age groups (Fig. 2), with neonates most commonly presenting with cyanosis and infants presenting equally with congestive heart failure (CHF) or cyanosis. There was a trend toward older patients presenting asymptomatically. Patients with an abnormal TV were more likely to present with CHF ($p = 0.006$) and more likely to develop significant TR ($p = 0.001$). Interestingly, these patients were not more likely to have earlier surgery ($p = 0.3$), nor were they statistically more likely to develop poor RV function ($p = 0.1$).

Outcome

During the follow-up period, there were 20 deaths (16.5%) and 5 cardiac transplants (4%) in 121 patients. Table 1 shows the median age of death or transplant. Causes of death included systemic (right) ventricular failure (6), postoperative (4), sudden death (3), sepsis (2), ventricular arrhythmia (1), complete AV block/acidosis (1), and unknown (3). For comparison purposes, cardiac transplant and death were considered to be the same outcome. The Kaplan–Meier estimate of survival of the entire group is shown in Fig. 3A. Five-, 10-, and 20-year survival rates were 92%, 91% and 75%, respectively. Of the 101 survivors, at last follow-up 74 patients were New York Heart Association (NYHA) class I, 15 were class II, 4 were class III, and 8 were class IV. Bacterial endocarditis occurred seven times in 5 patients; 1 additional patient suffered a cerebral abscess. There have been six successful pregnancies and one terminated pregnancy (for noncardiac indications).



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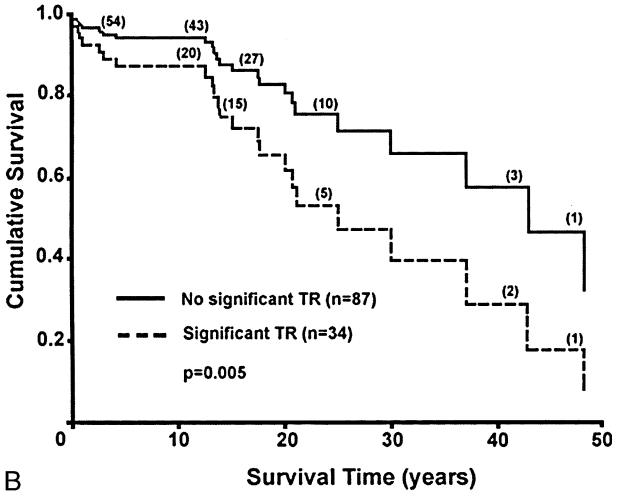

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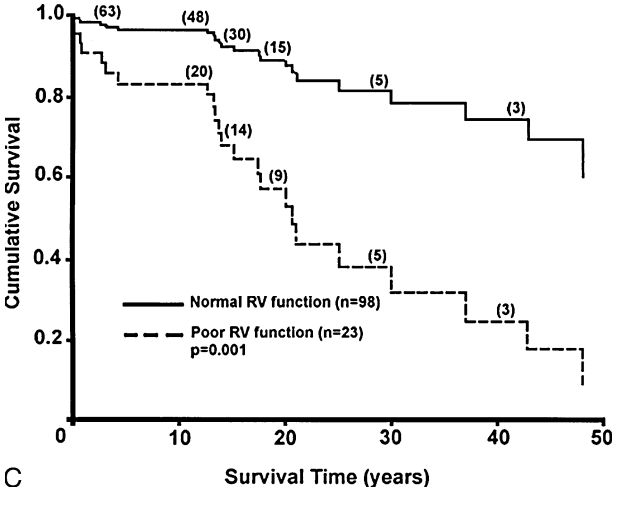

C

Thirty patients are currently managed with afterload-reducing agents.

Predictors of Outcome. Risk factors for mortality are shown in Table 3. Variables analyzed were age at diagnosis, physiologic group at presentation, the presence of associated lesions, TV abnormalities, AV block (naturally occurring and postoperative), the presence of a pacemaker, arrhythmias, type of surgery, age at surgery, year of surgery, preoperative pulmonary-to-systemic flow ratio, severity of pulmonary outflow tract obstruction, RV function, and TR. By multivariate analysis, complete AV canal defect ($p = 0.006$, RR = 7.3) and poor RV function ($p = 0.002$, RR = 5.9) remained significant as predictors of mortality. Figures 3B and 3C show actuarial survival curves for patients with significant TR and poor RV function, respectively.

Surgery

Eighty-six patients underwent surgery with an overall operative mortality of 2.5%. Median total surgical follow-up was 7 years (mean 9 years; range 1 month to 47 years). Median follow-up after biventricular or Fontan repair was 4.8 years (mean 6.1 years; range 1 month to 25.8 years). Biventricular repair was performed in 47 patients and varied according to the patients' underlying anatomy (Table 4). In 27 patients no initial palliation was performed, 15 patients had a previous systemic-to-pulmonary artery shunt (multiple shunts in 6 patients), and 5 patients had a previous pulmonary artery band (rebanding in 1 patient). Twenty-six patients remain palliated with a systemic-to-pulmonary artery shunt (18) or pulmonary artery band (8). Two patients required coarctation of the aorta repair only, and 11 patients have undergone single ventricle palliation with either a bidirectional Glenn anastomosis (1) or Fontan (10).

Of the five cardiac transplant recipients, two patients had previous shunts; one had aortic valve and TV replacement, one had TV replacement and ASD and VSD closure, and one was previously unoperated. None of the eight patients with an AV canal defect underwent biventricular repair; three patients were shunted, two had a pulmonary artery

Fig. 3. Kaplan–Meier survival curves for all patients with congenitally corrected transposition of the great arteries (**A**), for patients with and without significant tricuspid regurgitation (defined as moderate or severe TR) (**B**), and for patients with and without poor right ventricular function (defined as moderate or severe reduction in RV function) (**C**). *RV*, right ventricle; *TR*, tricuspid regurgitation.

Table 3. Risk factors for mortality

	p value	RR	95% confidence interval
	Univariate analysis		
Age at biventricular repair	0.04	0.87	0.77–0.99
Complete atrioventricular canal defect	0.02	4.6	1.25–17.0
Dextrocardia	0.05	2.4	1.0–5.7
Moderate to severe TR	0.05	2.4	1.0–5.1
Moderate to severely decreased RV function	0.001	4.7	2.0–12.0
	Multivariate analysis		
Complete atrioventricular canal defect	0.006	7.3	5.9–8.8
Moderate to severely decreased RV function	0.002	5.9	4.8–7.0

RR, relative risk; RV, right ventricle; TR, tricuspid regurgitation.

Table 4. Surgical and transcatheter procedures (87 patients, January 1952 to July 1999)

Procedure	No.
Extracardiac/palliative procedures	
Systemic to pulmonary artery shunt (revisions)	39 (18)
Pulmonary artery band (rebanding)	17 (3)
Bidirectional Glenn anastomosis	5
CoA/arch repair (revision)	9 (2)
Intracardiac procedures	
VSD ± PS relief	35
VSD alone	9
VSD closure + LV–PA conduit	17
LV–PA conduit alone	2
VSD closure + valvotomy/sub-PS resection	7
TV repair (alone or combined with another procedure)	7
TV replacement (alone or combined with another procedure)	12
Atrial and arterial switch procedure	1
Fontan	10
Repeat TV replacement	3
Transplant	5
Cardiac catheterization procedure[a]	9
Other[b]	11

ASD, atrial septal defect; CoA, coarctation of the aorta; LV, left ventricle; PA, pulmonary artery; PS, pulmonary stenosis; TAPVR, total anomalous pulmonary venous return; TV, tricuspid valve; VSD, ventricular septal defect.

[a] ASD closure (3), PA stents (2), conduit dilatation (1), Fontan fenestration dilatation (1), Fontan conduit stent (1), pulmonary valve perforation and dilatation (1).

[b] TAPVR repair (4), Damus–Kaye–Stansel (1), conduit replacement (4), ASD closure (1), ventricular assist device (1).

band, and two had a Fontan. Four patients are currently awaiting transplantation and six are awaiting surgery within the next 6 months.

Univentricular Palliation. Eleven patients underwent single ventricle palliation with a Fontan (10) or bidirectional Glenn procedure due to straddling tricuspid valve (4) or mitral valve (4) or due to cardiologist/surgeon preference (3). Median age at follow-up was 16.2 years (range 3.8–23.3 years). There has been one death; a 2½-year-old male died of bacterial and fungal sepsis 6 months after fenestrated Fontan. The remaining patients are followed, with good ventricular function in 8, mildly decreased function in 1, and moderately decreased function in 1. Three patients have none/trivial TR, 6 have mild TR, and 1 patient has mild to moderate TR. Five patients are NYHA class I, 4 are class II, and 1 is class III. One patient is currently awaiting heart transplantation.

Tricuspid Valve Function

Biventricular repair was performed in 47 patients. Excluding 11 patients who underwent tricuspid valve repair or replacement prior to or at the time of biventricular repair, 19 of 36 patients (53%) had worsening TR to a moderate or severe degree following surgery. In contrast, of 39 patients who remained palliated (shunt/pulmonary artery band) or had a single ventricle repair, 7 (18%) had worsening TR, as did 8 of 35 patients (23%) who had no surgery ($p = 0.03$). There was a trend toward an increased frequency of abnormal tricuspid valves in the biventricular repair group (again excluding those with previous TV surgery) which did not reach statistical significance ($p = 0.06$). Median time to development of significant TR was 3.1 years after biventricular repair, 11.1 years after palliation/bidirectional Glenn/Fontan, and 13.5 years in patients requiring no surgery ($p = 0.05$).

Analysis of the entire group, comparing patients who developed worsening TR to those that did not, demonstrated that biventricular repair ($p = 0.03$) and complete AV block ($p = 0.04$) were the only factors that correlated with increasing TR. There was no correlation with an abnormal TV ($p = 0.7$),

preoperative physiologic group ($p = 0.7$), degree of PS preoperatively ($p = 0.15$), or systematic-to-pulmonary flow ratio ($p = 0.95$). There was a trend toward a correlation between younger age at biventricular repair and worsening TR which did not reach statistical significance ($p = 0.06$).

Right Ventricular Function

Excluding the 11 patients who underwent TV repair or replacement prior to or at the time of surgery, 12 of 36 patients undergoing biventricular repair (33%) had worsening RV function (moderate to severe dysfunction by echocardiography) following surgery compared to 5 of 39 patients (13%) who had palliation/bidirectional Glenn/Fontan and 3 of 35 patients (9%) who were unoperated ($p = 0.02$). Of the 20 patients with worsening RV function, 15 also had increasing TR. Predictors of worsening RV function included biventricular repair ($p = 0.02$), moderate or severe TR ($p < 0.001$), and complete AV block ($p = 0.001$).

Tricuspid valve repair was attempted in seven patients (Ebstein in two, dysplastic in one, and straddling in two). Three patients have had a good result, with reduction in the degree of TR and improving RV function. One patient has residual moderate TR and three patients have had subsequent valve replacement. Nine additional patients have had TV replacement (Ebstein in six and dysplastic in two). Two of these patients went on to cardiac transplant: one required a repeat TV replacement, and one required two further replacements.

Electrophysiologic Issues

Complete AV block was seen in 25 patients, including at birth in 8 patients' and as a postoperative complication in 8 patients. Atrial arrhythmias occurred in 22 patients and ventricular arrhythmias in 8 patients. Thirty patients (25%) required pacemaker implantation—20 for complete atrioventricular block (6 postoperative), 7 for advanced second-degree atrioventricular block (1 postoperative), 2 for sinus node dysfunction, and 1 for antitachycardia pacing. Of the 20 surviving patients with pacemakers, 16 are programmed in DDD mode and 4 in VVI mode. Pacemaker and generator revisions have been required in 16 patients.

Pacemaker placement was associated with poor RV function ($p = 0.001$) and significant TR during the course of follow-up ($p = 0.04$), although there was no correlation with significant TR at the last follow-up ($p = 0.15$). There was no association between the presence of a pacemaker and NYHA class.

Discussion

This study represents the largest nonselected cohort of patients with ccTGA followed at a single center. In our 121 patients, associated defects were present in accordance with previously reported cohorts [9, 12, 17, 28]. Our study confirms that long-term survival in these patients is reduced and is influenced by the presence of associated anomalies as well as the development of significant TR and RV dysfunction as previously reported [12, 17, 21]. Complete AV canal defect, a predictor of mortality in our group, has not been reported in previous studies. The median age of death in our population was 13.2 years (mean 15.6 years), which is somewhat younger than in other series [7, 9, 28], likely reflecting the fact that our study reports the youngest median age at diagnosis for a cohort with ccTGA.

Systemic Ventricular Function

The ability of the RV to sustain systemic cardiac output over time is a primary concern in long-term follow-up of patients with ccTGA. In previous series, RV failure accounts for up to 50% of deaths [6, 7]. Several investigators have evaluated ventricular function in ccTGA patients with conflicting results. Using quantitative angiography in 19 patients with ccTGA, Graham et al. [10], concluded that RV function was maintained in childhood (<10 years) but RV dysfunction became more common after 17 years of age, suggesting an inability of the RV to sustain systemic output over a normal lifetime. Connelly et al. [7], using radionuclide angiography in 25 adults with ccTGA, found a significant reduction in exercise performance and systemic ventricular function compared to those of normal adults as well as an inability of the systemic RV to increase ejection fraction in response to exercise. In contrast, Dimas et al. [8], using quantitative angiography and echocardiography, showed that RV ejection fraction did not significantly change over 10 years of follow-up in 18 adults with ccTGA.

Theoretical factors contributing to deteriorating RV function include volume overload (such as from significant TR or a large VSD), conduction abnormalities and arrhythmias, systemic afterload, and surgical considerations (including surgical AV block and potential worsening of RV function and TR induced by surgery). In our series, complete AV block

(postoperative or acquired), biventricular repair, and moderate or severe TR all correlated with worsening RV function over time. Although previous studies have shown an association between significant TR, RV volume overload, and complete AV block with deteriorating RV function [21, 23], the current study provides convincing evidence that patients undergoing a conventional biventricular repair are statistically more likely to develop poor RV function compared to patients who remain palliated, have a single ventricle repair, or require no surgery. Sano et al. [23] reported worsening RV function following biventricular repair in 12 of 22 patients; however, there was no comparison made with patients who remained palliated or were unoperated. In that study, the preoperative pulmonary-to-systemic-flow ratio was significantly higher, as was the presence of significant TR in those patients who developed RV dysfunction postoperatively. Voskuil et al. [28] reported a trend toward an increased incidence of deterioration of RV function following biventricular repair compared to patients who were unoperated or were palliated. In a recent multi-institutional study of 182 adult patients with ccTGA, RV function was found to deteriorate over time. By 45 years of age, 56% of patients with significant associated lesions and 32% of patients without significant associated lesions had this complication. Risk factors for RV dysfunction in that series included history of any open-heart surgery, TR, tricuspid valve surgery, significant arrhythmia, and pacemaker therapy [9].

Our results, combined with these reports, clearly show that patients undergoing conventional biventricular repair in which the morphologic RV supports the systemic circulation are at increased risk for developing worsening RV function over time.

Tricuspid Regurgitation

Long-term follow-up of patients with ccTGA has shown that significant TR develops over time, regardless of whether there are associated TV structural abnormalities [6, 17, 21]. Several studies have suggested that TV function can be affected following intracardiac repair. Implicated factors include RV dysfunction following cardiopulmonary bypass leading to tricuspid annular dilation [17], annular distortion caused by VSD closure [19], changes in interventricular septal configuration and annular dilatation after relief of left ventricular outflow tract obstruction [18, 23], poor tolerance of the TV to systemic pressure after VSD closure, and postsurgical complete AV block [1, 23, 26, 29].

In our series, excluding patients who had TV repair or replacement, 53% of patients undergoing biventricular repair developed moderate or severe TR compared to 18% who had palliation or single ventricle repair and 23% who had no surgery. Patients undergoing biventricular repair developed significant TR sooner than the other groups as well. Predictors of worsening TR in our series included complete AV block and biventricular repair. Patients underwent repair either via a right atriotomy or a left ventriculotomy if a conduit was required, and therefore direct surgical damage to the TV is unlikely to be responsible for worsening TR. Despite complete AV block being a predictor of worsening TR, there was a low occurrence of surgical AV block in our series (8 cases). Our results are in contrast to those of Prieto et al. [21], who reported increasing TR in 5 of 19 patients following open-heart surgery compared to 10 of 19 non-open-heart surgery cases. Only the presence of a morphologically abnormal TV was significantly associated with the development of significant TR in their study. However, Sano et al. [23] reported worsening TR (moderate to severe degree) in 16 of 24 patients following conventional biventricular repair and Voskuil et al. [28] reported increasing TR in 16 of 32 patients following biventricular repair compared to 11 of 36 patients who did not require surgery or were palliated.

Attempts at TV repair were largely unsuccessful in the majority of patients in our study (three of seven patients have progressed to tricuspid valve replacement and one patient continues to be followed with moderate TR). We support consideration for the double switch procedure in patients with an abnormal, regurgitant TV as opposed to attempting repair or replacement alone or in conjunction with a conventional biventricular repair.

Fontan Palliation

The Fontan operation has been advocated as an alternative surgical strategy for patients with ccTGA, especially in the presence of straddling AV valves or ventricular hypoplasia. In our series, there has been a reasonable mortality rate and excellent long-term functional outcome with the majority of survivors being in NYHA class I or II. Ventricular function appears to be relatively well maintained over time as well. Our generally favorable results following the Fontan operation support consideration of this surgical approach in patients not suitable for biventricular repair.

Double Switch Operation

Anatomical repair of ccTGA (atrial switch via baffling of the venous return combined with arterial switch),

the so-called double switch operation, resulting in a systemic left ventricle, has been performed with increasing frequency with encouraging early results. Perioperative mortality has been comparable to that of conventional biventricular repair and short-term follow-up studies have reported significant reductions in TR with improved RV volume and function [14, 15, 16, 22, 28]. Although long-term studies are lacking, the double switch procedure appears promising, especially in patients with an abnormal TV and significant TR. Because of these encouraging results, some centers have advocated this procedure as the treatment of choice for selected patients with ccTGA. Since July 1999, we have performed this operation in selected patients with favorable short-term results (not included here due to insufficient follow-up).

Study Limitations

Texas Children's Hospital is a tertiary care facility with a large referral base. There is likely to be an underestimation of either asymptomatic patients with no associated cardiac anomalies or patients who are hemodynamically "well balanced" that may be followed at other centers. Although both adult and pediatric patients are followed at our center, the relatively young patient population in this series, compared to other reports, suggests that adult patients are underrepresented. In addition, the heterogeneous nature of this lesion warrants caution in extrapolating our experience to adult survivors with this lesion. However, given the propensity for worsening TR and RV function with time, our finding of deterioration in RV function and TR at a relatively young age is not likely to be overestimated and would only be more evident with the inclusion of more adult patients. The degree of RV dysfunction was graded by transthoracic echocardiography. Although the echocardiography assessment of RV function is not ideal, it is the most widely used method and the quality of images was generally adequate in our patient population.

In conclusion, survival in patients with ccTGA is largely determined by the presence of associated cardiac anomalies. The presence of significant TR and RV dysfunction consistently predicts mortality. In our series, conventional biventricular repair was significantly associated with progressive TR and RV dysfunction. We believe that our results support an alternative surgical approach (i.e., double switch procedure) for repair of associated lesions in patients with ccTGA.

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