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Background. This study was undertaken to evaluate long-term results of biventricular repairs for congenitally corrected transposition of the great arteries, and to analyze the risk factors that affect mortality and morbidity.

Methods. Between 1983 and 2009, 167 patients with congenitally corrected transposition of the great arteries underwent biventricular repairs. The physiologic repairs were performed in 123 patients, and anatomic repairs in 44. Average follow-up was 9.3 ± 6.6 years.

Results. Kaplan-Meier estimated survival was 83.3% ± 0.5% at 25 years in biventricular repair. In anatomic repair, left ventricular training and right ventricular dysfunction had negative impact on survival, but bidirectional cavopulmonary shunt had positive impact on survival. The reoperation-free ratio was 10.1% ± 7.8% at 22 years after physiologic repair, and 46.2% ± 12.4% at 15 years after anatomic repair ($p = 0.885$). Freedom from any arrhythmia was 49.6% ± 7.5% at 22 years after physiologic repair, and 60.8% ± 14.8% at 18 years after anatomic repair ($p = 0.458$). Freedom from systemic atrioventricular valve and ventricular dysfunction as well as tricuspid valve and right ventricular dysfunction was significantly higher in anatomic repair than in physiologic repair.

Conclusions. Long-term results of biventricular repair were satisfactory. Patients presenting with right ventricular dysfunction or need for left ventricular training represent a high-risk group of anatomic repair for which selection criteria are particularly important. Late functional outcomes of anatomic repair were excellent compared with physiologic repair. Anatomic repair is the procedure of choice for those patients if both ventricles are adequate or if surgical technique is modified with the help of additional a bidirectional cavopulmonary shunt.

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Congenitally corrected transposition of the great arteries (CCTGA) is characterized by atrioventricular (AV) and ventriculoarterial discordance [1]. The optimal surgical management of patients with CCTGA is still to be determined. The early results after physiologic repair are on the whole quite good; however, the results in the medium to long term are disappointing. A particular problem with physiologic repair seems to be a high incidence of late death caused by cardiac failure [2–5]. Concerns about the long-term function of the morphologic right ventricle (RV) and the systemic AV valve (tricuspid) have led to the concept of anatomic repair that incorporates the morphologically left ventricle (LV) and morphologically left AV valve (mitral) in the systemic circulation [6].

In 1990, Dr Ilbawi [6] reported success with anatomic repair for AV discordance as an alternative to physiologic repair. Ilbawi's contribution of using the LV in the systemic circulation expanded the options for these patients [7–9]. Advantages in terms of RV and tricuspid valve (TV) function combined with low operative morbidity and mortality, and the fact that the LV becomes the systemic ventricle, have resulted in the application of this procedure to almost all patients with CCTGA, despite its complexity [10–13].

So far, no study has analyzed the long-term results of biventricular repair including one-and-a-half ventricular repair for CCTGA. Because of the relative rarity of suitable candidates, we included data from four institutions. So, the present multicenter study seeks to evaluate the outcome of biventricular repair for CCTGA, and to analyze the factors that predict outcomes after biventricular repair. In particular, we wish to compare long-term outcomes after physiologic versus anatomic repair for CCTGA, and evaluate the impact of additional bidirectional cavopulmonary shunt on survival.

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Abbreviations and Acronyms

AV = atrioventricular
CCTGA = congenitally corrected transposition of the great arteries
LV = left ventricle
PA = pulmonary artery
PSVT = paroxysmal supraventricular tachycardia
PV = pulmonary ventricle
RV = right ventricle
TV = tricuspid valve
VSD = ventricular septal defect

Material and Methods

Institutional Review Board approval was obtained for the conduct of this retrospective study, and as individual patients were not identified, individual patient consent for the study was waived. Data collection was performed following strict guidelines to protect patient information. Between 1983 and 2009, a total of 167 patients (median age 4.1 years; range, 1 months to 61.9 years old) with CCTGA underwent biventricular repairs for CCTGA in four institutions. Medical records, preoperative and postoperative echocardiograph data, and cardiac catheterization data and operative notes were reviewed. Of the 167 patients, 141 were situs solitus and 26 were situs inversus; 135 patients had ventricular septal defect (VSD). Regarding pulmonary outflow tract, 41 patients had no pulmonary stenosis, 74 had pulmonary stenosis, and 52 pulmonary atresia. Fifty-four patients underwent preoperative Blalock-Taussig shunt.

The severity of AV valve regurgitation was graded as I (jet area/atrial area $< 10\%$); II (jet area/atrial area 10% to 20%); III (jet area/atrial area 20% to 33%); and IV (jet area/atrial area $> 33\%$), and valvar dysfunction was defined as regurgitation of grade II and more. Ventricular function was judged as good, mild, moderate, and severe dysfunction; and ventricular dysfunction was defined as mild to moderate dysfunction and worse. In physiologic repair, the mean grade of tricuspid regurgitation (TR) was 1.23 ± 1.40. The grade of TR was 0 in 56 patients, I in 22, I to II in 1, II in 18, II to III in 2, III in 9, and IV in 15. In anatomic repair, the mean grade of TR was 1.06 ± 1.10. The grade of TR was 0 in 19 patients, I in 7, I to II in 5, II in 6, II to III in 4, III in 2, and IV in 1. For physiologic repair, RV function was good in 94 patients, with mild dysfunction in 13, mild to moderate dysfunction in 9, and severe dysfunction in 7. For anatomic repair, RV function was good in 27 patients, with mild dysfunction in 7, mild to moderate dysfunction in 6, moderate dysfunction in 2, and severe dysfunction in 2.

For physiologic repair, preoperative rhythm was sinus in 113 patients, and for anatomic repair, it was sinus in 41. For physiologic repair, preoperative arrhythmia occurred in 10 patients. Complete AV block occurred in 4 patients (3 required intraoperative permanent pacemaker), second-degree AV block in 1, atrial fibrillation in 3, and intermittent paroxysmal supraventricular tachycardia (PSVT) in 2. For anatomic repair, preoperative arrhythmia occurred in 3 patients. Complete AV block occurred in 2 patients (requiring preoperative permanent pacemaker), and intermittent PSVT in 1.

The physiologic repairs consisted of atrial septal defect or VSD closure in 14 patients, TV surgery in 21, pulmonary ventricle (PV) to pulmonary artery (PA) conduit interposition in 54, relief of PV outflow tract obstruction in 26, and *réparation à l'étage ventriculaire* in 8. The anatomic repairs were double-switch operation (atrial switch plus arterial switch) in 10 patients, and atrial switch plus intraventricular rerouting (with or without extracardiac conduits) in 34 (see the Appendix). Five patients underwent physiologic one-and-a-half ventricular repair, and 14 underwent anatomic one-and-a-half ventricular repair.

Statistical analyses were performed using SPSS version 17.0 software (SPSS, Chicago, IL). Data are presented as mean $\pm$ SD or median with ranges, as appropriate. The χ^2 test was used to compare categorical variables, and the t tests for continuous variables. Estimated survival and freedom from events including reoperation, arrhythmia, AV valve regurgitation, and ventricular dysfunction were determined by the Kaplan-Meier method. Variables were evaluated using by the likelihood ratio test in the Cox proportional hazards regression model. Hazard ratios with 95% confidence intervals were constructed for the significant multivariable predictors. A p value of less than 0.05 was set as the level of statistical significance.

Results

Early mortality was 7.2%. Patients were followed for 9.4 ± 6.7 years (range, 0 to 24.9). Late mortality was 4.5%. The Kaplan-Meier survival rate was $92.8\% \pm 0.2\%$ at 1 year, $87.6\% \pm 2.8\%$ at 10 years, and $83.3\% \pm 0.5\%$ at 25 years in biventricular repair (Fig 1). The cause of mortality included low cardiac output syndrome in 6 patients, arrhythmia in 4, heart failure in 3, respiratory arrest in 1, disseminated intravascular coagulopathy in 1, ventricular dysfunction in 1, thromboembolism in 1, fungal endocarditis in 1, and mediastinitis in 1 (Table 1). Postoperative complications occurred in 53 patients (31.7%). After physiologic repair, complications occurred in 38 patients (30.9%), including arrhythmia in 23, TR in 1, acute respiratory distress syndrome in 1, and transient seizure in 1. After anatomic repair, complications occurred in 15 patients (34.1%), including arrhythmia in 5, low cardiac output syndrome in 2, neurologic complication in 2, atrial baffle stenosis in 2, and acute renal failure in 1 ($p = 0.70$).

Reoperations were required in 71 patients (45.8%). After physiologic repair, reoperations were required in 60 patients (50.9%), and after anatomic repair, in 11 (29.7%) (Fig 1). With physiologic repair, reoperations included conduit change in 33 patients, TV replacement in 22, permanent pacemaker implantation in 11, pulmonic valve replacement in 7, PA angioplasty in 7, TV repair in 5, relief of PV outflow tract obstruction in 4, mitral valve repair in 3, aortic valve repair in 2, mitral valve repair in 1, left ventricular outflow tract widening in 1, bidirectional cavopulmonary shunt in 1,

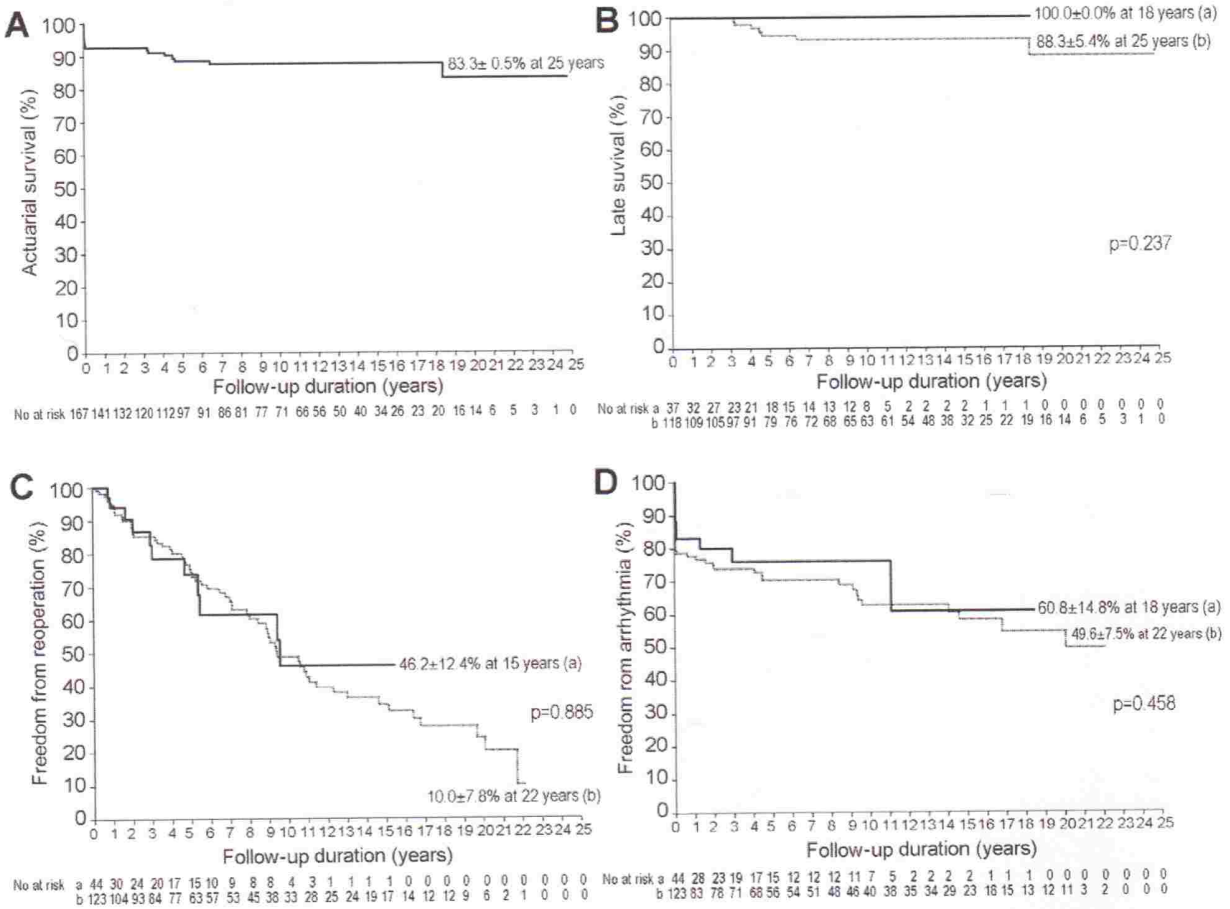

Fig 1. Kaplan-Meier curves for survival, reoperation, and arrhythmia. (A) The probability of survival, in years, for the whole group (n = 167). (B) Survival excluding hospital operative mortality. (C) Freedom from reoperation. (D) Freedom from arrhythmia. (a) Anatomic repair. (b) Physiologic repair.

Maze in 1, and implantable cardioverter defibrillator in 1. With anatomic repair, reoperations included conduit change in 10 patients, venous pathway widening in 5, subaortic membrane resection in 2, PA angioplasty in 2, left ventricular outflow tract widening in 2, pulmonic valve replacement in 1, TV replacement in 1, and permanent pacemaker implantation in 1 (Table 2).

Arrhythmias occurred in 49 patients (29.3%). Arrhythmias after physiologic repair occurred in 40 (32.5%), and after anatomic repair, in 9 (20.5%) (Fig 1). After physiologic repair, complete AV block occurred in 27 patients (including permanent pacemaker implantation in 23), atrial fibrillation in 9, PSVT in 7, atrial flutter in 3, sinus node dysfunction in 2, ventricular tachycardia in 1, and ventricular fibrillation in 1 (requiring implantable cardioverter defibrillator). After anatomic repair, complete AV block requiring permanent pacemaker implantation occurred in 3, atrial flutter in 5, PSVT in 2, ventricular tachycardia in 1, and first-degree AV block in 1. The freedom from tachyarrhythmia was 72.8% ± 7.9% at 23 years after physiologic repair, and 68.2% ± 16.1% at 18 years after anatomic repair ($p = 0.296$). The freedom from bradyarrhythmia was 69.3% ± 5.2% at 25 years after physiologic repair, and 83.8% ± 6.4% at 18 years after anatomic repair ($p = 0.301$). The incidence of complete AV block during follow-up after operation was significantly lower for anatomic repair group (6.8%) than for physiologic repair (22.0%; $p = 0.03$), and also tended to be lower for Rastelli-type anatomic repair (3.2%) than for double-switch repair (15.4%; $p = 0.14$) (Table 3).

Last follow-up echocardiography results after physiologic repair were available for 123 patients at a follow-up duration of 9.5 ± 7.0 years (range, 0 to 24). The grade of TR was 0 in 12 patients, I in 27, I to II in 15, II in 18, II to III in 6, III in 6, III to IV in 1, and IV in 17 (excluding TV replacement in 21). The mean grade of TR was 1.86 ± 1.24; the TR was significantly deteriorated as compared with the preoperative TR of 1.23 ± 1.40 ($p = 0.001$). The RV function was good in 52 patients, with mild dysfunction in 21, mild to moderate dysfunction in 27, moderate dysfunction in 7, and severe dysfunction in 16; the RV function was significantly deteriorated as compared with preoperative RV function ($p = 0.001$). Last follow-up echocardiography results after anatomic repair were

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Table 1. Risk Factors for Mortality in Biventricular Repair

	Univariable			Multivariable		
Variable	HR	95% CI	*p* Value	HR	95% CI	*p* Value
Biventricular repair						
Arrhythmia	0.683	0.246–1.898	0.464			
Pulmonary stenosis	1.601	0.642–3.992	0.313			
Pulmonary atresia	0.614	0.203–1.856	0.388			
Ventricular septal defect	0.848	0.281–2.557	0.770			
Preoperative PAB	4.878	1.402–16.971	0.013	5.203	1.440–18.801	0.012
Preoperative shunt	0.826	0.295–2.311	0.716			
BCPS	0.041	0.000–26.752	0.335	0.160	0.021–1.200	0.075
Tricuspid regurgitation	1.029	0.720–1.471	0.874			
RV dysfunction	1.331	0.832–2.128	0.233			
Physiologic repair	0.686	0.428–1.100	0.118			
CPB time	1.002	0.997–1.007	0.400			
Aortic cross-clamp time	1.006	1.000–1.013	0.055	1.006	1.000–1.013	0.048
Age	0.997	0.958–1.037	0.867			
Physiologic repair						
Arrhythmia	0.545	0.147–2.016	0.363			
Pulmonary stenosis	1.443	0.457–4.557	0.532			
Pulmonary atresia	0.890	0.240–3.302	0.862			
Ventricular septal defect	2.586	0.334–20.044	0.363			
Preoperative shunt	0.862	0.230–3.228	0.825			
BCPS	0.047	0.001–75920.957	0.675			
Tricuspid regurgitation	0.951	0.594–1.522	0.834			
RV dysfunction	0.988	0.472–2.068	0.974			
CPB time	0.995	0.981–1.009	0.487			
Aortic cross-clamp time	1.010	0.996–1.025	0.154			
Age	1.002	0.959–1.047	0.922			
Anatomic repair						
Arrhythmia	1.340	0.260–6.914	0.727			
Pulmonary stenosis	2.365	0.529–10.569	0.260			
Pulmonary atresia	0.230	0.028–1.913	0.174			
Ventricular septal defect	0.432	0.084–2.228	0.316			
Preoperative shunt	0.621	0.120–3.200	0.569			
Preoperative PAB	3.086	0.690–13.792	0.140			
LV training	5.348	1.195–23.933	0.028	5.750	1.283–25.757	0.022
Tricuspid regurgitation	1.282	0.682–2.411	0.441			
RV dysfunction	2.482	1.079–5.711	0.033	4.010	1.622–9.911	0.003
Age	1.043	0.900–1.210	0.574			
CPB time	1.001	0.993–1.009	0.779			
Aortic cross-clamp time	1.004	0.992–1.015	0.513			
Mustard/Senning	1.294	0.612–2.737	0.499			
ASO/Rastelli	1.348	0.638–2.850	0.434			
BCPS	0.285	0.065–1.247	0.028	0.150	0.020–0.985	0.050
Complete AV block	1.453	0.175–12.079	0.729			

ASO = arterial switch operation; AV = atrioventricular; BCPS = bidirectional cavopulmonary shunt; CI = confidence interval; CPB = cardiopulmonary bypass; HR = hazard ratio; LV = left ventricle; PAB = pulmonary artery banding; RV = right ventricle.

available for 44 patients at a follow-up duration of 4.7 ± 4.8 years (range, 0 to 18.5). The grade of TR was 0 in 19 patients, I in 11, I to II in 6, II in 2, II to III in 3, and III in 3. The mean grade of TR was 0.92 ± 0.98. As compared with preoperative TR of 1.06 ± 1.10, no change was observed ($p = 0.367$). The RV function was good in 34 patients, with mild dysfunction in 5, and mild to moderate dysfunction in 5; the RV function was significantly improved as compared with preoperative RV function ($p = 0.002$). The grade of mitral regurgitation was 0 in 19 patients, I in 7, I to II in 8, II in 3, and III in 1. The mean grade of mitral regurgitation was 0.64 ± 0.81. The LV

Table 2. Risk Factors for Reoperation

	Univariable			Multivariable		
Variable	HR	95% CI	*p* Value	HR	95% CI	*p* Value
Pulmonary stenosis	0.941	0.589–1.501	0.798			
Pulmonary atresia	1.895	1.176–3.053	0.009	3.125	1.630–5.994	0.001
Ventricular septal defect	2.795	1.123–6.954	0.027			
Preoperative PAB	0.047	0.001–38.357	0.372			
Preoperative shunt	2.166	1.330–3.528	0.002			
Tricuspid regurgitation	0.818	0.663–1.009	0.261			
RV dysfunction	0.800	0.553–1.158	0.237			
Age	0.984	0.955–1.015	0.310			
Physiologic repair	1.024	0.739–1.420	0.885			
BCPS	1.661	0.740–3.732	0.219			
CPB time	1.000	0.997–1.003	0.874			
Aortic cross-clamp time	0.999	0.994–1.004	0.728			
Arrhythmia	1.152	0.715–1.857	0.561			

BCPS = bidirectional cavopulmonary shunt; CI = confidence interval; CPB = cardiopulmonary bypass; HR = hazard ratio; PAB = pulmonary artery banding; RV = right ventricle.

function was good in 35 patients, with mild dysfunction in 6, mild to moderate dysfunction in 1, moderate dysfunction in 1, and severe dysfunction in 1. Freedom from systemic AV valve and ventricular dysfunction as well as from TV and RV dysfunction were significantly higher after anatomic repair than after physiologic repair (Fig 2).

Follow-up data after physiologic repair were available for 111 patients at the follow-up duration of 10.6 ± 6.8 years (range, 0 to 24.9). The last follow-up rhythm was sinus in 81 patients (including intermittent PSVT in 3), pacing in 25 (including atrial fibrillation in 1), and atrial fibrillation in 5. Follow-up data after anatomic repair were available for 37 patients at the follow-up duration of 5.9 ± 4.6 years (range, 0.4 to 18.5). The last follow-up rhythm was sinus in 34 patients, and pacing in 3 (p = 0.017). After physiologic repair, 97 patients met the criteria for New York Heart Association functional class I, 11 met criteria for class II, and 3 met criteria for class III. After anatomic repair, 34 patients met the criteria for the New York Heart Association functional class I, and 2 who had moderate aortic regurgitation and LV dysfunction after the anatomic repair with arterial switch, met criteria for class II; 1 had neurologic complication (p = 0.46).

Comment

This study not only is one of the largest cohorts of biventricular repair in patients with CCTGA, but also analyzes long-term results of physiologic and anatomic repair, including one-and-a-half ventricular repair. For physiologic repair, our recent long-term survivals of 84.7% ± 5.4% at 25 years were good compared with the data previously reported [2–5]. The only previously identified risk factor for decreased long-term survival after physiologic repair was reported to be the presence of preoperative TR [14]. The poorest outcome was also reported among patients who required TV replacement either at the initial operation or later during follow-up [5]. Therefore, the surgical management should include consideration of earlier timing of TV replacement or a cardiac repair, which relieves the RV and TV of a systemic workload [15]. In our series, preoperative PA banding and cardiopulmonary bypass time were risk factors for overall mortality, but TR and RV dysfunction were not risk factors. The patients who required TV replacement had also good results, 4.8% overall mortality, because TV replacement was performed at the earliest sign of increasing symptoms or progressive systemic ventricular deterioration to preserve RV function [3].

However, for anatomic repair, our series showed that the patients with RV dysfunction showed a worse survival rate than did the other patients. The reasons might include severe TR, especially associated with Ebstein anomaly, RV hypoplasia, high pulmonary vascular resistance, and heart failure. Our data also demonstrated that patients (n = 6; age range, 2.19 to 16.05 years) who complete LV training had deterioration in LV dysfunction at an earlier stage, resulting in more mortality compared with patients who did not require training. Actually, selecting the best procedure is difficult in these patients with an intact ventricle septum, severe TR, and RV dysfunction in old age, but in our early experience, LV training followed by anatomic repair was not decided under strict selection criteria. The durability of LV function after LV training has been reported to be low due to a high incidence of the trained LV failure after the anatomic repair [12, 16]. The LV conditioning time to which the ventricle was subjected to systemic pressures before the anatomic repair was also short [16] because the LV was subjected only to pulmonic pressures all through from birth to LV training. Longer follow-up will be needed for evaluation of deterioration of LV function late after LV training.

In this series, 5 patients with physiologic one-and-a-half ventricular repair had good results without mortal-

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Table 3. Risk Factors for Arrhythmia

	Univariable			Multivariable		
Variable	HR	95% CI	*p* Value	HR	95% CI	*p* Value
Overall arrhythmia						
Pulmonary stenosis	0.949	0.553–1.627	0.849			
Pulmonary atresia	0.527	0.271–1.023	0.058			
Ventricular septal defect	0.361	0.204–0.640	0.001			
Preoperative PAB	2.168	0.775–6.067	0.141			
Preoperative shunt	0.486	0.243–0.971	0.041			
Tricuspid regurgitation	1.417	1.171–1.715	0.001	1.550	1.211–1.983	0.001
RV dysfunction	1.507	1.135–2.000	0.005			
Age	1.030	1.014–1.046	0.001			
Physiologic repair	1.075	0.770–1.502	0.670			
BCPS	0.506	0.157–1.633	0.255			
Cardiopulmonary bypass time	1.001	0.998–1.005	0.421			
Aortic cross-clamp time	1.002	0.997–1.007	0.333			
Tachyarrhythmia						
Pulmonary stenosis	0.561	0.236–1.331	0.190			
Pulmonary atresia	1.013	0.415–2.469	0.978			
Ventricular septal defect	0.355	0.150–0.843	0.119			
Preoperative PAB	1.509	0.199–11.442	0.691			
Preoperative shunt	0.886	0.344–2.279	0.801			
Tricuspid regurgitation	1.633	1.228–2.172	0.001	2.232	1.434–3.472	0.001
RV dysfunction	1.930	1.329–2.801	0.001			
Age	1.042	1.019–1.066	0.001			
Physiologic repair	0.776	0.479–1.258	0.304			
BCPS	0.490	0.065–3.707	0.490			
Cardiopulmonary bypass time	1.004	1.000–1.008	0.047	1.007	1.003–1.012	0.002
Aortic cross-clamp time	1.007	1.001–1.014	0.029			
Bradyarrhythmia						
Pulmonary stenosis	1.118	0.581–2.151	0.739			
Pulmonary atresia	0.414	0.172–0.996	0.049			
VSD	0.401	0.200–0.805	0.010			
Preoperative PAB	2.226	0.677–7.320	0.188			
Preoperative shunt	0.416	0.173–1.003	0.051			
Tricuspid regurgitation	1.214	0.962–1.532	0.103			
RV dysfunction	1.244	0.868–1.785	0.234			
Age	1.022	1.002–1.043	0.028	1.022	1.001–1.043	0.041
Physiologic repair	1.252	0.806–1.943	0.317			
BCPS	0.480	0.115–2.008	0.315			
Cardiopulmonary bypass time	0.999	0.994–1.004	0.672			
Aortic cross-clamp time	0.999	0.992–1.006	0.763			

BCPS = bidirectional cavopulmonary shunt; CI = confidence interval; CPB = cardiopulmonary bypass; HR = hazard ratio; PAB = pulmonary artery banding; RV = right ventricle; VSD = ventricular septal defect.

ity. It was particularly applicable in situations where the atrial switch with the Rastelli procedure is not possible in CCTGA with pulmonary stenosis because of the position of the VSD. It was also beneficial in the patients with hypoplasia or malfunction of the LV because of unloading the LV by reducing flow through the LV to PA connection. The anatomic one-and-a-half type of ventricular repair is very useful particularly in hypoplasia or malfunction of the RV because of unloading the RV. It carries also no risk of superior vena cava obstruction, with less risk of obstruction to the pulmonary venous return from increasing the intra-atrial space, less risk of arrhythmias from reducing the intra-atrial suture lines, and reduced ischemic time. The widest and most reliable application of partial biventricular repair is likely to be for patients with hypoplasia or functional compromise of the right-side heart complex, or both [17, 18]. The presence of a straddling AV valve or an inlet VSD with no outlet extension would make it difficult to tunnel the VSD to the aorta without compromising the volume of the RV because of the size of the baffle as well as TV function. In these circumstances, the anatomic one-and-a-half type of

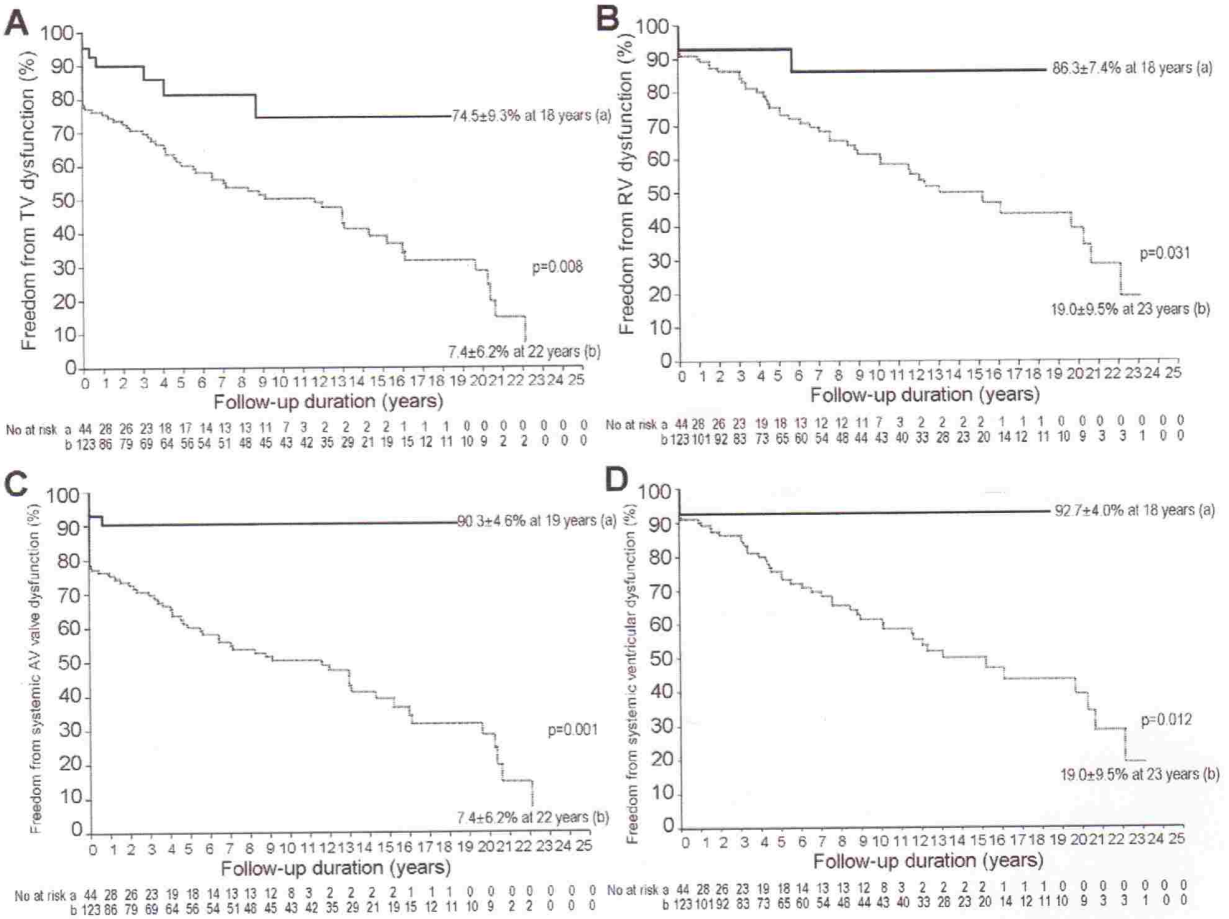

Fig 2. Freedom from atrioventricular valve and ventricular dysfunction. (A) Freedom from tricuspid valve (TV) dysfunction. (B) Freedom from right ventricle (RV) dysfunction. (C) Freedom from systemic atrioventricular (AV) valve dysfunction. (D) Freedom from systemic ventricular dysfunction. Atrioventricular valve dysfunction was defined as regurgitation grade of 2 and more; ventricular dysfunction was defined as mild to moderate dysfunction and worse. (a) Anatomic repair. (b) Physiologic repair.

ventricular repair was applied, resulting in a positive impact on survival, as demonstrated by this study.

Risks for reoperation were pulmonary atresia, as indicated by multivariate analysis, which was caused by the need for conduit change or pulmonary valve replacement. After physiologic repair, the prevalence of systemic AV valve failure is likely to result in a considerably higher incidence of reoperation in the longer term among patients who have undergone a physiologic approach [2, 4, 19]. The most frequent early complication after surgical correction of CCTGA is complete heart block [2, 4, 20]. The incidence of complete heart block tends to be lower after anatomic repair than after physiologic repair [2, 4, 20–22]. In this study, the incidence of postoperative complete AV block was also lower after anatomic repair than after physiologic repair (6.8% versus 22.0%, $p = 0.03$). In the Rastelli-type anatomic repair, the closure of VSD through the right ventriculotomy allows a more ready accessibility to the RV aspect of the septum than the other approaches, and that is important in avoiding the conduction system [6, 23]. In our patients, the Rastelli-type anatomic repair (3.2%) was associated with the lowest incidence of postoperative heart block compared with the anatomic repair with arterial switch (15.4%) and the physiologic repair (22.0%) [22]. The pulmonary atresia, VSD, and preoperative shunt had protective effect on the bradyarrhythmia, which was related to the fact that in CCTGA with pulmonary atresia and VSD, the physiologic repair with PV-PA conduit interposition had less complete AV block than relief of PV outflow tract obstruction without conduit; and the Rastelli-type anatomic repair also had the lowest incidence of complete AV block, but old age was a risk factor for the bradyarrhythmia. Although a long-term complication after anatomic repair is the development of surgically induced arrhythmias by suture line and suture load, our anatomic repair group tended to have similar tachyarrhythmic events even though atrial switch was added, but they had fewer bradyarrhythmic events such as complete AV block.

In our series, the long-term follow-up data demon-

strate that funtional status was good in both biventricular repairs, but freedom from systemic AV valve and ventricular dysfunction as well as from TV and RV dysfunction were significantly higher after anatomic repair than after physiologic repair. Late mortality was 5.9% after physiologic repair, but 0% after anatomic repair. Our data showed that with the restoration of the LV to the systemic circulation by the anatomic repair, the incidence of systemic AV valve regurgitation and ventricular failure was reduced, and good functional status was achieved without long-term mortality.

The study is limited by its retrospective design, which might include bias as to the selection of surgical methods. The 25-year period of surgical experience might have been associated with changes in selection and management of patients undergoing the biventricular repair that could influence long-term results. The earliest candidates for anatomic repair were often the worst. They were often selected by virtue of being unsuitable for the physiologic repairs owing to the presence of various risk factors and contraindications. A well-designed, large, multicenter cohort study will be the only practical solution in resolving the optimal choice of surgical procedures for these patients.

In conclusion, long-term results of biventricular repair were satisfactory. Patients presenting with RV dysfunction or need for LV training represent a high-risk group of anatomic repair patients for whom selection criteria are particularly important. Our study demonstrates the superiority of anatomic repair compared with physiologic repair in systemic AV valve and ventricular function, although the incidence of reoperation or arrhythmia was not different. Anatomic repair is the procedure of choice for those patients if both ventricles are adequate or if surgical technique is modified with the help of an additional bidirectional cavopulmonary shunt.

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Appendix

Type of Operation

Diagnosis	Physiologic Repair		Anatomic Repair	
	Type of Operation	No.	Type of Operation	No.
No pulmonary stenosis				
VSD or ASD	VSD or ASD closure	14	Senning and ASO	6
			Mustard and ASO	4
Tricuspid regurgitation	Tricuspid valve replacement	21		
Pulmonary stenosis or atresia				
VSD or ASD	PV-PA conduit interposition	54	Senning and Rastelli	15
	Relief of PVOT obstruction	26	Mustard and Rastelli	14
	REV	8	Senning and ASO	2
			Mustard and ASO	1
			Senning and REV	1
			Mustard and REV	1
Total		123		44

ASD = atrial septal defect; ASO = arterial switch operation; PA = pulmonary artery; PV = pulmonary ventricle; PVOT = pulmonary ventricular outflow tract; REV = réparation à l'étage ventriculaire; VSD = ventricular septal defect.

Outcomes of Biventricular Repair for Congenitally Corrected Transposition of the Great Arteries

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