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CONGENITAL

Surgical Outcomes of Anatomic Repair in Congenitally Corrected Transposition of the Great Arteries: A 28-Year, Multicentre Dutch Experience

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Graphical abstract

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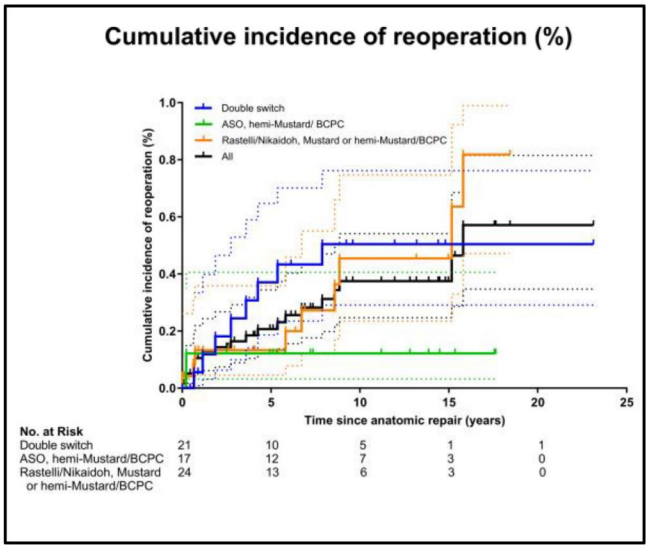
Summary

In a Dutch multicentre cohort of 62 ccTGA patients (1997-2025), anatomic repair was performed at a median age of 2.5 years. Following the early learning phase, hospital mortality decreased to 1.9%. At 20-year follow-up, survival exceeded 80% with preserved systemic left ventricular function. Reoperations (31%) and pacemakers (32%) were common, least after arterial switch combined with a hemi-Mustard and bidirectional Glenn.



Legend: ccTGA: congenitally corrected transposition of the great arteries

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Abstract

Objectives: Anatomic repair is the contemporary preferred management strategy for congenitally corrected transposition of the great arteries (ccTGA). Most outcome data derive from small single-centre series. The goal of this study was to review long-term outcomes of surgical treatment in a multicentre cohort of ccTGA patients in the Netherlands over a 28-year period.

Methods: All patients with ccTGA undergoing anatomic repair (double switch repair, arterial switch operation with hemi-Mustard/bidirectional Glenn, or Rastelli/Nikaidoh with Mustard or hemi-Mustard/bidirectional Glenn) between 1997 and 2025 were analysed retrospectively.

Results: Sixty-two patients underwent anatomic repair at a median age of 2.5 (IQR: 1.4-4.0) years. Overall hospital mortality was 6.5%, decreasing to 1.9% after 2006. At a median follow-up of 11.9 years, the 20-year survival exceeded 80%. Reoperation was required in 31%, most often for conduit replacement after the Rastelli/Nikaidoh procedure, baffle obstruction or neoaortic valve/root disease after a double switch procedure, and least often after the arterial switch with hemi-Mustard/bidirectional Glenn (11.7%). Percutaneous interventions were performed in 16%. Complete atrioventricular block requiring a pacemaker implant occurred in one-third, whereas late atrial arrhythmias were infrequent (<10%). At the latest follow-up, systemic left ventricular function was preserved in most patients, with only 2 patients showing moderate dysfunction.

Conclusions: After an initial learning curve, anatomic ccTGA repair exhibited low operative mortality, good long-term survival and preserved systemic left ventricular function in the majority of patients. Reintervention rates are considerable.

Keywords: congenitally corrected transposition of the great arteries; anatomic repair; double switch operation; arterial switch operation; pediatric cardiac surgery.

Introduction

Congenitally corrected transposition of the great arteries (ccTGA) accounts for approximately 0.5% of all congenital heart diseases.[1] ccTGA is characterized by atrioventricular and ventriculo-arterial discordance with the morphologic right ventricle (RV) supporting the systemic circulation and the tricuspid valve functioning as a systemic atrioventricular valve. Although ccTGA can present as an isolated lesion, it is often associated with additional anomalies, most commonly ventricular septal defect (VSD), left ventricular outflow tract obstruction, coarctation and Ebstein-like malformations of the tricuspid valve.[1] Coronary artery anomalies are also reported, though they occur less frequently than in complete transposition of the great arteries.

The subaortic position of the RV predisposes to progressive RV dysfunction and tricuspid regurgitation, both of which determine morbidity and mortality in the natural history of ccTGA and after physiologic repair. Recognition of the poor long-term prognosis of the systemic RV led to the development of anatomic repair strategies designed to restore the morphologic left ventricle (LV) to the aorta. These approaches, first described in the 1980s and more widely applied in the 1990s, include combinations of atrial and arterial switch procedures or Rastelli/Nikaidoh operations to address associated lesions.[2,3]

Although multiple centres have reported their experience with anatomic repair, most are single-centre series with modest sample sizes and variable follow-up.[4-10] As a result, the long-term preservation of systemic LV function, the burden of reoperations and the late incidence of arrhythmia after anatomic repair remain incompletely characterized. Furthermore, the optimal surgical approach continues to evolve, with recent adoption of the hemi-Mustard procedure combined with bidirectional cavopulmonary connection (BCPC) and either an arterial switch operation (ASO) or Rastelli/Nikaidoh procedures, aimed at reducing operative complexity and improving outcomes.[11] The present study reports long-term outcomes of anatomic ccTGA repair, based on a 28-year multicentre experience in the Netherlands.

Methods

Study population

Patients who underwent surgical treatment for ccTGA at Leiden, Utrecht and Groningen University Medical Centres between January 1997 and January 2025 were included in this study. Data on demographics, morphologic and operative details (including procedures prior to anatomical repair) and mid- and long-term outcomes were collected from hospital and outpatient records. The latest follow-up echocardiographic images were reviewed. The local committees for medical ethics at the respective university medical centres approved the study and waived the need for informed consent.

Outcomes

Primary end points included hospital and late mortality, surgical and percutaneous reinterventions and the LV ejection fraction (LVEF). Secondary end points included complete heart block and pacemaker implants, arrhythmias and echocardiographic parameters of right ventricular function as well as atrioventricular (AV) and (neo) aortic valve function. Hospital mortality was defined as death from any cause occurring during the same hospitalization in which anatomic repair was performed, even beyond 30 days. Late mortality was defined as death from any cause occurring after hospital discharge.

The LVEF was measured by echocardiography at the latest follow-up and categorized as normal (LVEF: ≥55%),

mildly reduced (LVEF: 45%-54%), moderately reduced (LVEF: 30%-44%) or severely reduced (LVEF: ≤30%). If objective data were missing, systolic function was assessed subjectively and categorized accordingly. The RV function was assessed subjectively and categorized as normal, mildly reduced, moderately reduced or severely reduced. Tricuspid, mitral and (neo)aortic valve regurgitation were assessed qualitatively and categorized as none/trivial, mild, moderate or severe.

Surgical strategy

Anatomic repair was performed by cardiothoracic surgeons from 2 centres across 3 institutions. The indications for anatomic repair were based on institutional protocol, but generally were similar, that is, patients with associated anomalies that necessitated surgical repair, progressive decline of systemic RV function, progressive tricuspid valve dysfunction and/or symptom development. Patients with isolated ccTGA without associated lesions and anatomical/physiologic anomalies of the tricuspid valve were managed conservatively and entered a lifelong follow-up program. Surgical strategies for anatomic repair included ASO combined with an atrial switch (Mustard or Senning; ie, double switch), ASO combined with a hemi-Mustard/BCPC, Rastelli or Nikaidoh procedure with an atrial switch (Mustard) and a Rastelli or Nikaidoh procedure with hemi-Mustard/BCPC.

In selected cases, the definitive surgical strategy was determined intraoperatively based on direct anatomic assessment. In patients with pulmonary atresia or severe pulmonary valve stenosis associated with an outlet-type VSD, a Rastelli procedure was performed, consisting of left ventricular-to-aortic tunnelling using a Gore-Tex baffle and right ventricular outflow tract reconstruction with an RV-to-pulmonary artery (PA) conduit (Contegra or homograft). Conversely, in the presence of a pulmonary LV outflow tract obstruction and an inlet or restrictive VSD, or straddling atrioventricular valves, an aortic root translocation (Nikaidoh) with biventricular outflow tract reconstruction was undertaken instead of a Rastelli repair. Right ventricle hypoplasia or anticipated RV volume loss after Rastelli or Nikaidoh procedures were indications for a hemi-Mustard/BCPC. Since 2015, one centre has adopted the hemi-Mustard/BCPC in combination with ASO or Rastelli/Nikaidoh as its primary approach, replacing the full atrial switch and favouring this strategy over the double switch. This shift was motivated by the fact that the hemi-Mustard procedure is technically less complex and generally faster to perform, particularly in patients with normal situs and dextrocardia or with situs inversus and levocardia. In addition, a bidirectional Glenn may offer advantages in the presence of greater-than-physiologic tricuspid regurgitation or when significant RV volume reduction is anticipated after a Rastelli procedure. A limitation of this approach is that a subsequent transvenous pacemaker implant is no longer feasible.

Anatomic repairs were generally scheduled when the patient reached a weight of 8-12 kg. Pulmonary artery banding (PAB) was performed to achieve an LV pressure ≥ 90% of systemic pressure without inducing LV dilation or dysfunction. Rebanding was undertaken when the LV pressure decreased relative to systemic levels, indicating insufficient retraining and the need to delay the definitive procedure. Preparedness of the LV was typically reassessed after 9-12 months using echocardiography and cardiac catheterization. Criteria for LV readiness prior to anatomic repair followed the Stanford group's standards: LV pressure ≥ 90% of systemic pressure, LV ejection fraction > 55% and LV end-diastolic pressure < 12 mmHg.[11]

Statistical analysis

Clinical characteristics were presented as frequency (%) for categorical variables and as median (interquartile range [IQR]) for continuous non-normally distributed variables. Differences in proportions of categorical variables between anatomic repair subgroups were assessed using the Fisher-Freeman-Halton exact test. Because the analyses are primarily descriptive in nature, missing data were handled using complete-case reporting. No imputation was performed, and descriptive statistics reflect the number of available observations for each variable. Continuous non-parametric data were compared between subgroups using the Kruskal-Wallis test. Kaplan-Meier analyses were performed to estimate overall survival, and cumulative incidence functions were used for freedom from reoperation. For the estimation of follow-up time, the inverse Kaplan-Meier method was used; that is, patients were censored at the time of the event (death) and, for patients without an event, the last follow-up time was counted as an event. A median follow-up index was calculated; follow-up ended 28 August 2025. A *P*-value < .05 was considered statistically significant. Statistical analyses were performed using IBM SPSS Statistics 23.0.

Results

Patient characteristics

Sixty-two patients underwent anatomic repair, including a double switch procedure with or without VSD closure ($n = 21$), ASO with hemi-Mustard/BCPC ($n = 17$) and Rastelli or Nikaidoh procedures ($n = 24$; 17 Rastelli, 7 Nikaidoh) with either a Mustard atrial switch ($n = 5$) or a hemi-Mustard/BCPC ($n = 19$). Baseline characteristics are summarized in **Table 1**. Notable diagnostic features included true double outlet right ventricle in 7 patients and criss-cross heart in 2. Pulmonary atresia or (severe) pulmonary stenosis was present in 23 of 24 patients undergoing a Rastelli/Nikaidoh procedure, with dextrocardia being most common. Ebstein-like tricuspid valve anatomy was absent in the Rastelli/Nikaidoh group but was observed in 3 of 21 double switch cases (14%) and 9 of 17 ASO with hemi-Mustard/BCPC cases (53%). Moderate RV hypoplasia was noted in 2 patients in the latter group.

Forty-seven patients (76%) required cardiac operations prior to the anatomic correction (**Table 2**), including 15 with 2 or more procedures (2 required 3 procedures). Pulmonary artery banding, the most common intervention, was performed in 31 patients for LV training; 5 of them

Table 1. Baseline Characteristics of Patient Cohort

	Double switch procedure ($n = 21$)	ASO, hemi-Mustard, BCPC ($n = 17$)	Rastelli/Nikaidoh		*P*-value
			Mustard ($n = 5$)	hemi-Mustard, BCPC ($n = 19$)	
Male, *n* (%)	17 (81)	11 (65)	3-(60)	15 (79)	.539
Age at repair, years	2.4 [0.9-5.4]	2.5 [1.9-3.0]	2.3 [1.0-6.6]	2.9 [1.4-3.9]	.855
Visceral situs, *n* (%)					.878
Situs solitus	20 (95)	16 (94)	5 (100)	17 (89)	
Situs inversus	1 (5)	1 (6)	0	2 (11)	
Cardiac position, *n* (%)					<.001
Levocardia	18 (86)	13 (77)	2 (40)	4 (21)	
Mesocardia	2 (9)	4 (23)	0	5 (26)	
Dextrocardia	1 (5)	0	3 (60)	10 (53)	
Associated cardiac anomalies, *n* (%)					
DORV	2 (9)	0	0	5 (26)	.083
Criss-cross heart	1 (5)	0	0	1 (5)	1.000
RV hypoplasia	0	2 (12)	0	0	.228
Ventricular septal defect	13 (62)	7 (41)	5 (100)	19 (100)	<.001
MV cleft	2 (9)	1 (6)	0	1 (5)	1.000
Bilateral superior caval vein	2 (9)	0	0	2 (11)	.672
RVOT, *n* (%)					<.001
Pulmonary atresia	0	0	2 (40)	7 (37)	
Pulmonary stenosis[a]	0	0	2 (40)	12 (63)	
Subvalvular pulmonary stenosis[b]	3 (14)	2 (12)	1 (20)	0	
Tricuspid valve morphology, *n* (%)					<.001
No abnormality	17 (81)	4 (23)	5 (100)	18 (95)	
Ebstein-like TV	3 (14)	9 (53)	0	0	
TV dysplasia	1 (5)	1 (6)	0	0	
TV straddling	0	3 (18)	0	1 (5)	
Arch abnormality, *n* (%)					.059
Aortic coarctation[c]	4 (19)	1 (6)	1 (20)	0	
Hypoplastic aortic arch[d]	3 (14)	0	0	0	
Right aortic arch	0	0	1 (20)	2 (11)	
Coronary artery anatomy, *n* (%)[e]					.188
1R-2CxL	15 (71)	16 (94)	3 (75)	9 (60)	
1RCxL	2 (10)	0	0	4 (27)	
Miscellaneous	4 (19)	1 (6)	1 (25)	2 (13)	
Unknown	–	–	1	4	

Abbreviations: DORV, double outlet right ventricle; MV, mitral valve; RV, right ventricle; RVOT, right ventricular outflow tract; TV, tricuspid valve.
[a]Bicuspid, fibrotic or small annulus.
[b]Aneurysmatic membranous septum.
[c]Coarctectomy performed.
[d]No arch repair needed.
[e]Leiden Convention coronary coding system.

required banding adjustment. The median age at PAB was 0.3 (IQR: 0.1-0.8) years, and the median time from the first PAB to the anatomic correction was 2.1 (IQR: 1.1-3.2) years.

Follow-up and mortality

The median age at anatomic repair was 2.5 (IQR: 1.4-4.0) years. Hospital mortality was 6.5% (4 patients): 2 after the double switch procedure (1 Senning; 1 Mustard), 1 after the Rastelli-Mustard and 1 after ASO with hemi-Mustard/BCPC. Notably, 3 of these 4 deaths occurred before 2006. Complete follow-up was available for all but 1 patient, who was lost to follow-up; the median follow-up index was 0.93 (IQR: 0.85-0.97). The median follow-up among hospital survivors was 11.9 (IQR: 5.1-16.1) years, with a maximum of 28.6 years. Five patients died late (8.1%) at a median interval of 1.2 (IQR: 0.3-14.1) years after the anatomic repair (at a median age of 3.4 [IQR: 1.7-22.2] years). Causes of death included cardiac failure due to severe neoaortic regurgitation in a resuscitation setting (age 1.7 years; 0.4 years after the double switch-Mustard); cardiac arrest during induction of anaesthesia for reoperation for baffle stenosis (age 3.4 years; 1.2 years after Nikaidoh-Mustard); progressive cardiac failure (age 1.6 years; 0.2 years [76 days] after Nikaidoh, hemi-Mustard/BCPC); cardiac failure secondary to influenza B pneumonia, unresponsive to ECMO support (age 20.3 years; 13.2 years after Rastelli with hemi-Mustard/BCPC); and sepsis (age 24.0 years; 15.0 years after Rastelli with hemi-Mustard/BCPC). The overall survival rate after anatomic repair was 88.6% at 5 years, 88.6% at 10 years, and 80.9% at 15 and 20 years. Survival curves did not significantly differ between subgroups ($P = .778$) (**Figure 1**).

Table 2. Number of Surgical and Percutaneous Procedures Prior To and After Anatomic Correction

	Double-switch procedure ($n = 21$)	ASO hemi-Mustard BCPC ($n = 17$)	Rastelli/Nikaidoh procedures ($n = 24$)
Surgical procedures prior to anatomical correction	*No. of patients (%)*		
Pulmonary artery banding (PAB)	12 (57)	13 (76)	0
PAB+atrioseptectomy	0	2 (12)	0
PAB+coarctectomy	3 (14)	1 (6)	0
mBTT-shunt (right, left or bilateral)	0	0	14 (58)
BCPC	0	0	3 (13)
Coarctectomy	1 (5)	0	1 (4)
Relief subaortic stenosis	1 (5)	0	0
Relief vascular sling (retrotracheal RSA)	0	1 (6)	0
None	6 (29)	1 (6)	8 (33)
	No. of patients (%)		
Concomitant procedures during anatomic correction			
Mitral valve repair	2 (10)	0	1 (4)
Mitral and tricuspid valve repair	0	3 (18)	0
Relief of aneurysmatic membranous septum in LVOT	1 (5)	1 (6)	0
Pulmonary valve commissurotomy	1 (5)	0	0
	No. of procedures/no. of patients (%)		
Reoperations after anatomic correction	*12/8*	*2/2*	*9/9*
Median follow-up time to reoperation, years	3.2 (1.3-5.1)	0.22	6.7 (0.7-12.0)
Reoperation procedures			
Relief obstruction pulmonary venous path	3 (14)	0	0
Relief obstruction systemic venous path	2 (10)	0	1 (4)
Coronary angioplasty	1 (5)	0	0
Replacement neoaortic valve	0	1 (6)	0
Valve sparing root replacement	1 (5)	1 (6)	0
RV-PA conduit implantation/replacement	0	0	6 (25)
Relief left ventricular outflow tract obstruction	0	0	1 (4)
Relief right ventricular outflow tract obstruction	2 (10)	0	0
Mitral valve repair	1 (5)	0	0
Closure residual ventricular septal defect	2 (10)	0	1 (4)
	No. of procedures/no. of patients		
Percutaneous interventions after anatomic correction	*2/2*	*2/2*	*7/6*
Balloon angioplasty unilateral PA branch stenosis	0	1 (6)	0
Stent unilateral PA branch stenosis	0	1 (6)	4 (17)
Stent superior caval vein	1 (5)	0	0
Stent distal RV-PA conduit	0	0	2 (8)
Percutaneous pulmonary valve implant	0	0	1 (4)
Percutaneous transluminal coronary angioplasty	1 (5)	0	0

Abbreviations: ASO, arterial switch operation; BCPC, bidirectional cavopulmonary connection; LVOT, left ventricular outflow tract; mBTT, modified Blalock-Thomas-Taussig shunt; PA, pulmonary artery; PAB, pulmonary artery banding; RSA, right subclavian artery; RV, right ventricle.

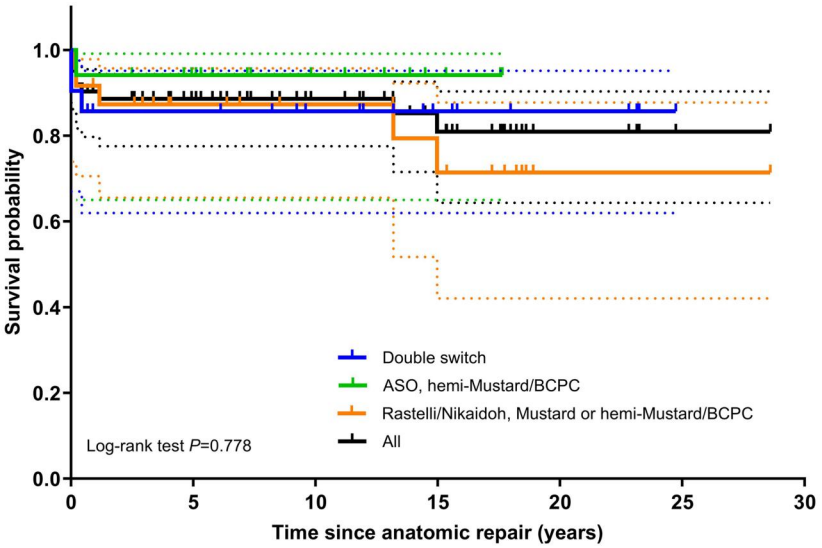
Reoperations and percutaneous interventions

Nineteen patients (30.6%) underwent a reoperation, with a total of 23 procedures, at a median of 3.6 (IQR: 0.7-7.9) years after anatomic repair (**Table 2**). Reoperation rates were 38.1% (8/21) after a double switch; 11.7% (2/17) after an ASO with a hemi-Mustard/BCPC and 37.5% (9/24) after the Rastelli/Nikaidoh operation (7/9 Rastelli with hemi-Mustard/BCPC, 1/9 Nikaidoh with hemi-Mustard/BCPC, 1/9 Rastelli-Senning) (**Figure 2**; **Table2**). In the double switch group, 5 of 8 reoperated patients had a baffle obstruction relieved (all following the Mustard atrial switch repair), and 1 required valve-sparing neoaortic root replacement (VSRR). In the ASO with the hemi-Mustard/BCPC group, reoperations (2/17) included neoaortic valve replacement (1) and VSRR (1). After the Rastelli/Nikaidoh repair, reoperations primarily involved RV-PA conduit replacement for conduit stenosis (6/9 reoperated patients).

Percutaneous interventions (11 procedures) were performed in 10 patients (16.1%) at a median of 5.7 (IQR: 0.8-9.7) years after anatomic repair. Intervention rates were 9.5% (2/21) after a double switch, 11.8% (2/17) after an ASO with a hemi-Mustard/BCPC and 25.0% (6/24) after a Rastelli/Nikaidoh repair (**Table 2**).

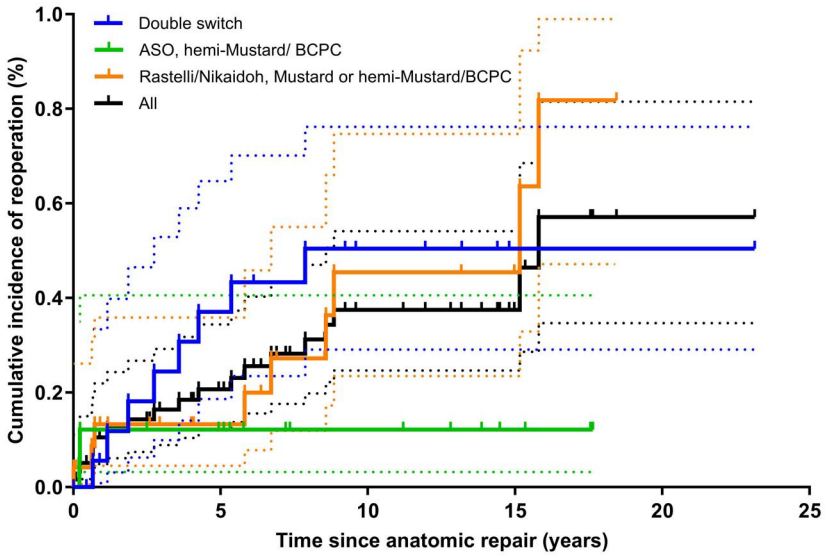
Arrhythmias, conduction disorders and cardiac devices

Of 62 patients, 20 (32.2%) developed complete AV block requiring a permanent pacemaker (PPM) implant. Three of these patients developed spontaneous AV block and underwent a PPM implant prior to anatomic repair at a median age of 25 days (IQR: 8-141 days). Ten patients required a PPM implant immediately following an anatomic correction, 4 at a later stage (at least >6 months post-repair) and 3 after a reoperation (surgical relief of



No. at Risk						
Double switch	21	16	12	7	4	0
ASO, hemi-Mustard, BCPC	17	13	7	3	0	0
Rastelli/Nikaidoh, Mustard or hemi-Mustard/BCPC	24	15	12	8	1	1

Figure 1. Kaplan-Meier Estimates of Overall Survival After Anatomic Repair.
Dotted lines denote 95% confidence intervals; tick marks denote censoring times.



No. at Risk					
Double switch	21	10	5	1	1
ASO, hemi-Mustard/BCPC	17	12	7	3	0
Rastelli/Nikaidoh, Mustard or hemi-Mustard/BCPC	24	13	6	3	0

Figure 2. Cumulative Incidence Curve of Reoperations.
Dotted lines denote 95% confidence intervals; tick marks denote censoring times.

LV outflow tract obstruction, VSRR and neoaortic valve replacement) (**Table S1**). Among patients requiring a PPM implant immediately following an anatomic correction, 5 were in the double switch group (5/21, 24%), none in the ASO with the hemi-Mustard/BCPC group and 5 in the combined Rastelli/Nikaidoh cohort (5/24, 21%). Within the Rastelli/Nikaidoh cohort, a PPM implant was more frequent after a Nikaidoh repair (3/7, 43%) than after a Rastelli repair (2/17, 12%). Four patients developed supraventricular tachyarrhythmias at a median age of 9.0 (IQR: 2.8-10.4) years (**Table S1**).

Ventricular performance and valve function

At the latest follow-up, a median LVEF was 50% (IQR: 48-54) after a double switch procedure, 53% (IQR: 49-55)

after an ASO with a hemi-Mustard/BCPC and 51% (IQR: 44-56) after the Rastelli/Nikaidoh procedures. Most patients demonstrated normal systemic LV function: 56% (10/18) of those following the double switch procedure, 88% (14/16) of those have following the ASO with hemi-Mustard/BCPC and 78% (14/18) of patients following the Rastelli/Nikaidoh procedures. Mild systolic dysfunction occurred in 39% (7/18) of patients following the double switch procedure, in 13% (2/16) of those following an ASO with a hemi-Mustard/BCPC and in 17% (3/18) of patients following the Rastelli/Nikaidoh. Moderate dysfunction was present in 2 patients: in 1 after the double switch procedure and in 1 after the Rastelli with the hemi-Mustard/BCPC repair (**Table S2**; **Figure S1**). The latter patient is currently listed for a heart transplant.

The impact of a prior PAB on LV function at latest follow-up is summarized in **Table S3**. Among patients following the double switch procedure, 50% (5/10) with preserved LV function and 88% (7/8) with reduced LV function had prior PAB. For those in the double switch group with prior PAB (12/18), the median age at the initiation of PAB did not differ between survivors with reduced LV function ($n = 7$) and those with preserved LV function ($n = 5$): 0.11 years (IQR: 0.06-0.53) versus 0.25 years (IQR: 0.05-1.07), respectively. The median interval between PAB and anatomic repair was also comparable: 2.42 years (IQR: 0.43-5.50) for those with reduced LV function versus 4.56 years (IQR: 0.56-5.44) for those with preserved LV function. Among patients undergoing ASO with hemi-Mustard/BCPC, 50% had prior PAB. Of the survivors in this group, 88% (14/16) with preserved LV function had prior PAB, compared with 50% (1/2) of those with reduced LV function with comparable age at PAB and interval between PAB and anatomic repair.

Right ventricular function was normal in 83% (15/18) after a double switch procedure, 100% (16/16) after an ASO with the hemi-Mustard/BCPC and 63% (12/19) after the Rastelli/Nikaidoh procedures. Mild systolic dysfunction was observed in 9 patients: 17% (3/18) after the double switch procedure and 33% (6/18) after Rastelli/Nikaidoh (**Table S2**). (Neo)aortic valve function was generally good at the latest follow-up, with no, trivial or mild (neo)aortic regurgitation in ≥92% of patients. Among those with a neoaortic valve, more than mild regurgitation was observed in 3 patients (3/34, 9%): 2 with severe regurgitation (1 requiring a neoaortic valve replacement and 1 awaiting a replacement) and 1 with moderate regurgitation. Mitral and tricuspid valve function was also generally good. More than mild mitral regurgitation was observed in 2 patients (moderate regurgitation) following a Rastelli repair without concomitant mitral valve plasty (1 hemi-Mustard/BCPC; 1 Rastelli-Senning). More than mild tricuspid regurgitation occurred in 1 patient (moderate regurgitation) after the Nikaidoh operation with a hemi-Mustard/BCPC.

Discussion

This study represents one of the longest follow-ups of anatomic repair for ccTGA: It spans nearly 3 decades. Key findings include an overall hospital mortality of 6.5% (only 1.9% after 2006), good long-term survival and long-term preservation of systemic LV function in most patients. Reoperations were relatively common but manageable, with the lowest rates observed in patients undergoing ASO with hemi-Mustard/BCPC. Our findings add to the growing body of evidence that anatomic repair provides favourable long-term outcomes for patients with ccTGA.

Hospital mortality of 6.5% in our series compares favourably with results from earlier experiences. Initial reports from the 1990s and early 2000s documented perioperative mortality up to 14%.[12-15] Centres that introduced anatomic repair later reported similarly elevated early risk during their initial experience,[5,7] reflecting the steep learning curve associated with this complex procedure. In our cohort, 3 of the 4 early deaths occurred before 2006, underscoring this phenomenon. Over time, refinements in surgical technique, patient selection and perioperative management have reduced operative mortality to between 2% and 8%, as demonstrated in a recent large multicentre Society of Thoracic Surgeons database study and several recent meta-analyses.[1,16,17] Long-term survival in our cohort was good, consistent with outcomes from single-centre series[11,18-20] and a meta-analysis.[17] These data confirm that anatomic repair can be performed with relatively low operative risk and provides durable survival into adulthood.

Reintervention is common, reflecting the complexity of atrial and ventricular rerouting and the frequent need for conduits. In our cohort, one-third of patients required a reoperation and nearly one-fifth required percutaneous interventions. These numbers are in line with the 20%-40% reoperation rates reported internationally within a 10-15 year period.[4,19,20] The causes of reoperations vary by surgical strategy. Conduit replacement dominates after Rastelli and Nikaidoh procedures; baffle obstruction and neoaortic valve dysfunction complicate double switch repairs; (branch) PA stenosis often drives catheter-based treatment. Our findings mirror these patterns. Notably, patients undergoing an ASO with a hemi-Mustard/BCPC required fewer reoperations than those with double switch or Rastelli-type procedures. Similar advantages of 1.5-ventricle strategies with ASO and hemi-Mustard/BCPC have been reported, supporting this evolution in surgical strategy with reduced surgical morbidity and fewer reoperations.[9-11]

Conduction disorders are frequent in ccTGA. Complete AV block requiring a permanently implanted pacemaker occurred in one-third of our patients, paralleling the incidence from international reports ranging from 20% to 40%.[8,12,21,22] An atrioventricular block reflects both the intrinsic conduction abnormalities inherent in ccTGA and the additional risk introduced by septal interventions and atrial switch procedures. Although based on a limited number of cases in our series, our data suggest that in patients with ccTGA, normal situs and VSD with left ventricular outflow tract obstruction, the Nikaidoh procedure may be associated with a higher risk of complete AV block compared with the Rastelli procedure. This difference may be related to the transection of the infundibular septum required during the Nikaidoh procedure. The incidence of arrhythmias in our cohort was lower than in other series, with <10% developing supraventricular tachyarrhythmias. International groups report

higher rates of late atrial arrhythmias, particularly in double switch cohorts with extensive atrial surgery.[20,21] The relatively lower incidence in our patients may relate to the increased adoption of ASO with hemi-Mustard/BCPC, which reduces atrial manipulation.

Preservation of systemic LV function is the primary goal of anatomic repair. In our study, most patients demonstrated normal or near-normal systolic LV function at long-term follow-up, with only 2 patients showing moderate dysfunction. These results are consistent with outcomes from other centres.[8,9,11,18] PAB performed for LV training prior to anatomic repair has been implicated in the development of late LV dysfunction, particularly if PAB was performed late, was too tight or remained for too long[6]; however, these associations have not been reproduced at the group level in large series.[11,18,19] In the present study, prior PAB was undertaken in most patients undergoing a double switch (14/21) or an ASO with hemi-Mustard/BCPC (17/18). No association between PAB and late LV dysfunction was observed across the cohort, likely reflecting early PAB and anatomic repair within the first 3 years of life; however, adverse effects in individual patients cannot be excluded. In contrast, systemic RV dysfunction and progressive tricuspid regurgitation are well recognized sequelae after physiologic repair.[17,23] Tricuspid regurgitation is particularly associated with the development of RV failure in this setting. In our cohort treated with anatomic repair, RV function was generally preserved, especially in the ASO with hemi-Mustard/BCPC group. Atrioventricular valve function was also favourable: tricuspid regurgitation remained limited, and mitral regurgitation was rare, consistent with the protective effect of restoring the ventricles to their natural roles.[8,9,18,22]

The systemic neoaortic root remains vulnerable to progressive dilatation and valve dysfunction after a double switch procedure or an ASO with a hemi-Mustard/BCPC. Reoperation rates for neoaortic valve and/or root pathology in ccTGA double switch cohorts range from 5% to 10%.[18-20] In our study, 3 of 19 reoperated patients underwent neoaortic procedures (1 valve replacement, 2 VSRRs), which aligns with published experiences. One patient with severe neoaortic regurgitation died in the first year after anatomic repair. Prior PAB has been linked to increased risk of subsequent root dilatation and valve dysfunction in both d-TGA and ccTGA cohorts, although our numbers were too small to study this association. Although the hemi-Mustard/BCPC approach has potential benefits, including reduced baffle- and sinus node-related complications and technical simplicity,[11] there are currently no reports or 4-dimensional flow magnetic resonance imaging studies evaluating whether a full atrial switch procedure provides superior haemodynamics.

Limitations

This study has several limitations inherent in its retrospective design. Operative strategies have evolved over the nearly 3-decade study period, introducing a potential era effect due to progressive refinements in operative techniques, perioperative management and patient selection. Stratified analyses were not performed because the limited cohort size precluded meaningful statistical comparison across time periods. Secondly, long-term outcomes of the one-and-a-half repair beyond the second postoperative decade remain unknown, and functional capacity data for our cohort were not available, limiting conclusions regarding lifelong benefits compared with conventional anatomical repair procedures. Lastly, although the cohort is large for this rare anomaly, it remains modest in absolute terms, reflecting the low prevalence of ccTGA. Consequently, the small sample size inherently limits the statistical power, increasing the risk of type II error; observed differences between groups should therefore be interpreted with caution and considered hypothesis-generating rather than definitive.

Conclusions

Anatomic repair of ccTGA can be performed with low operative mortality, good long-term survival and preservation of systemic LV function in the majority of patients. Reinterventions remain common, particularly after the double switch and the Rastelli/Nikaidoh procedures, but occur less frequently following an ASO with a hemi-Mustard/BCPC. These findings are consistent with a growing body of international literature and support anatomic repair as the optimal treatment strategy for most ccTGA (at least in those with associated lesions), provided the operations are performed in specialized centres. Lifelong multidisciplinary follow-up remains essential to manage the complications of this condition.

Author contributions

Roel van der Palen (Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Validation, Visualisation, Writing—original draft, Writing—review & editing), Kirolos Jacob (Conceptualization, Data curation, Investigation, Methodology, Writing—original draft, Writing—review & editing), Irene Kuipers (Data curation, Investigation, Writing—review & editing), Enza Sarlemijn (Data curation, Writing—review & editing), Sara Arrigoni (Data curation, Writing—review & editing), Felix Haas and Paul Schoof (Investigation, Supervision, Writing—review & editing), Mark Hazekamp (Conceptualization, Investigation, Supervision, Validation, Writing—review & editing).

Conflicts of interest

None declared.

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Data availability

Derived data supporting the findings of the study are available from the corresponding author on request.

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