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Cardiac Outcomes in Young Adult Survivors of the Arterial Switch Operation for Transposition of the Great Arteries

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Objectives We sought to determine cardiac outcomes in young adults with complete transposition of the great arteries (TGA) after the arterial switch operation (ASO).

Background Although cardiac outcomes in the pediatric population with TGA after ASO have been well described, outcomes in the adult population have not to our knowledge been studied.

Methods We determined late survival in all operative survivors with TGA after ASO performed before 1991 at our local pediatric referring hospital. In the subset of adults (n = 65) followed in our adult congenital cardiac clinic, we examined cardiac outcomes in adulthood.

Results Survival of the 132 infants discharged from hospital after ASO was 97% (70% confidence interval [CI]: 95.0% to 98.1%) at 20 years. In the 65 patients (mean age 21 ± 3 years, 62% male) followed at our institution, 17% (11 of 65) had at least 1 clinically significant cardiac lesion, including ventricular dysfunction, valvular dysfunction, or arrhythmias. Residual lesions were more common in those who had had cardiac reinterventions in childhood (odds ratio: 10.7, 95% CI: 2.1 to 55). In adulthood, 5 patients (8%) had arrhythmia requiring treatment and 7 patients (11%) required reinterventions (5 reoperations and 2 pacemaker implantations). Intervention for aortic valve regurgitation and aortic root dilation were not observed. Exercise capacity was reduced in most adults (82%) after ASO.

Conclusions Although most adults after ASO are well, and few have residual defects, there are subgroups, particularly those who needed further cardiac intervention in childhood, who are at higher risk for ventricular and valve dysfunction and arrhythmias. (J Am Coll Cardiol 2010;56:58–64) © 2010 by the American College of Cardiology Foundation

Complete transposition of the great arteries (TGA) is the most common cyanotic congenital heart disease presenting in the neonatal period (1,2). Without surgical intervention, survival is not expected (3). A number of different surgical approaches have been used over the years, the most contemporary of which is the arterial switch operation first performed by Dr. Adib Jatene in 1975 (4). Because of the current low operative mortality, excellent childhood survival, and the expectation that complications will be rare, the ASO is now the preferred surgical approach (5–10). Although this surgery is felt to be an improvement compared with the earlier atrial switch operation, late cardiac complications have been reported in children, including pulmonary artery stenosis, neo-aortic valve insufficiency, dilation of the aortic root (11), and coronary obstruction (12). Patients with ASO are now entering adult congenital cardiac clinics; however, cardiac outcomes in the adult population have not yet been well defined. Therefore, we sought to examine cardiac outcome in this early cohort of young adults with ASO.

Methods

After approval from the institutional ethical review board, we identified all 132 operative survivors with an ASO for TGA performed before 1991 at our local referring hospital, the Hospital for Sick Children, Toronto, Canada. Thus, all hospital survivors had the potential to be ≥18 years of

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age. At our institution, the first ASO was performed on August 1978. Complete details pertaining to underlying cardiac anatomy (including coronary artery pattern), operative techniques, and perioperative complications were obtained from the division of cardiac surgery database of the Hospital for Sick Children, Toronto, Canada. Survival data for all 132 operative survivors of an ASO for TGA was obtained by chart review or by contacting the referring physicians.

Of the 132 adults with ASO performed at the referring pediatric center, 4 had died (3 during childhood). Sixty-two patients were not followed at our clinic: 51 were followed at other Canadian adult congenital cardiac centers, 4 had moved abroad, and 7 were lost to follow-up. Detailed cardiac outcomes of the remaining 65 adults followed in our adult congenital cardiac clinic were examined. Routine care in our clinic includes an annual clinical evaluation with an electrocardiogram and transthoracic echocardiography. Thus, electrocardiographic and echocardiographic data at the time of last follow-up in the 65 patients seen in our institution was complete. Cardiopulmonary exercise testing and additional cardiac imaging such as myocardial perfusion imaging or magnetic resonance imaging are performed at the discretion of the treating physician. Demographics, medical and surgical histories, medication use, cardiac imaging, and adverse cardiac outcomes at the time of the last clinic visit were obtained by chart review. Cardiac morphology was classified as simple TGA (intact interventricular septum) or complex TGA (ventricular septum defect or a Taussig-Bing malformation).

Assessment of functional status was performed based on the last clinic visit. Functional status was classified according to the New York Heart Association functional classification. Reports of echocardiograms performed at the time of the last clinic visit were reviewed. All echocardiograms at our institution are read by experienced echocardiographers. Left ventricular (LV) ejection fraction (EF) is estimated using Simpson's method (13) and graded as normal (LVEF ≥55%) or mildly (LVEF 45% to 54%), moderately (LVEF 35% to 44%), or severely (LVEF <35%) dysfunctional. Right ventricular (RV) systolic function is assessed visually and graded as normal, mild, moderate, or severe. Regurgitant and stenotic lesions are defined and graded according to previously published guidelines (14). The diameter of the aortic root at the level of the sinus of Valsalva is measured, and diameters >36 mm were considered as dilated. Exercise capacity (peak oxygen consumption [MVo_2]) was obtained by bicycle from results of cardiopulmonary exercise testing. Not all exercise testing was performed at the time of the last clinical visit; however, exercise capacity as measured by maximal oxygen consumption was indexed for age and sex. The results of myocardial scintigraphy were classified as either normal or as perfusion defect at rest or with exercise.

The primary objective was to determine the clinical outcomes of adults (age ≥18 years) with TGA who had had an ASO (n = 65). Clinical outcomes of interest included: adverse cardiac events (cardiac death, myocardial infarction, heart failure, bradyarrhythmia, and/or tachyarrhythmia requiring treatment), functional capacity, ventricular function, neoaortic valve lesions or other valvular lesions, and aortic root diameter. On the basis of the clinical and echocardiographic data at last follow-up, we defined a cohort of patients with "clinically significant cardiac lesions." Clinically significant lesions were those cardiac lesions likely to have an impact on late outcomes, including moderate or greater systolic ventricular dysfunction (LVEF ≤45% and/or moderate or greater RV systolic dysfunction), moderate or greater valvular regurgitation or stenosis, and arrhythmias requiring therapy. We examined the prevalence and predictors of clinically significant cardiac lesions. We also examined the prevalence and predictors of the need for cardiac reinterventions in adulthood. Cardiac reinterventions included cardiac reoperations, catheter-based interventions, and pacemaker implantations.

Abbreviations and Acronyms

ASO = arterial switch operation
EF = ejection fraction
LV = left ventricle/ventricular
MVo_2 = peak oxygen consumption
RV = right ventricle/ventricular
TGA = transposition of the great arteries

Statistical analysis. SPSS software for Windows (version 17.0, SPSS Inc., Chicago, Illinois) was used for data analysis. Data are described as medians with ranges or means with SDs as appropriate. Comparisons of continuous or categorical variables were performed with Student *t*, chi-square, or Fisher exact tests. Survival was determined for all 132 hospital survivors of the ASO using a parametric survival curve. The prevalence of residual cardiac lesions at last follow-up and the prevalence of reintervention after age 18 years were calculated. A logistic regression model was used to determine univariate predictors of clinically significant cardiac lesions. A Cox regression analysis was used to determine the hazards for reinterventions in adulthood.

Results

Baseline data of the 132 hospital survivors are shown in Table 1. In the early era of ASO, patients were more likely to undergo a palliative procedure before definitive ASO repair compared with the more recent era (63% palliation before 1987 vs. 10% thereafter, $p < 0.001$), and an ASO was performed at older age (504 days before 1987 vs. 6 days thereafter, $p < 0.001$). Survival of the 132 infants discharged from hospital after an ASO was 98% (70% CI: 96.2% to 98.6%) at 10 years and 97% (70% CI: 95.0% to 98.1%) at 20 years (Fig. 1). Three patients died in childhood, and 1 patient at age 18 years. Median age of death was 8 years (range 1 to 18 years). The reason of death was known in 3 patients (fungal endocarditis and myocardial infarction in two 1-year-old children, and sudden death in an 18-year-old patient not followed locally). The patient

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Table 1 Baseline Characteristics of the Survivors of an ASO Before 1991

	All Patients (n = 132)	Study Patients (n = 65)	Follow-Up Elsewhere (n = 67)	p Value
Male	89 (67%)	40 (62%)	49 (73%)	0.2
Era of ASO before 1987	40 (30%)	18 (28%)	22 (33%)	0.6
Palliative procedure prior to ASO	35 (27%)	15 (23%)	20 (30%)	0.4
Cardiac anatomy				0.3
TGA with IVS	73	36	37	
TGA with VSD	57	27	30	
Taussig-Bing	2	2	0	
Coronary pattern				0.06
Usual	98	42	56	
Single ostium	7	5	2	
CX from RCA	21	13	8	
LAD from RCA	3	3	0	
Inverted	1	0	1	
Other	2	2	0	
Median age at ASO, days (range)	8 (2–2,986)	6 (2–1,198)	13 (2–2,986)	0.02
Cardiopulmonary bypass time, min	158 ± 58	150 ± 54	166 ± 60	0.1
Aortic cross-clamp time, min	93 ± 21	93 ± 21	93 ± 21	1.0
Circulatory arrest time, min	29 ± 32	34 ± 34	24 ± 29	0.08
RV-PA conduit	10	7	3	0.3

ASO = arterial switch operation; CX = circumflex coronary artery; IVS = intact ventricular septum; LAD = left anterior descending coronary artery; RCA = right coronary artery; RV-PA = right ventricle to pulmonary artery; TGA = transposition of the great arteries; VSD = ventricular septum defect.

who died suddenly was born with an intact ventricular septum with no associated lesion. He had a normal coronary artery pattern and had a primary arterial switch operation at the age of 6 days. He was not followed in our institution, and details pertaining to his status prior to death were not available.

The subset of patients with regular clinical follow-up in our center (n = 65) did not differ statistically regarding sex, cardiac morphology, coronary artery pattern, and surgical data. However, the patients followed at our center were younger at the time of ASO (median 6 days vs. 13 days, p = 0.02). In the cohort followed at our center, 38% (25 of 65) had required cardiac reinterventions in childhood; 20 children had reoperations, and 13 children had catheter-based reinterventions. The most frequent reason for reoperation during childhood was relief for RV outflow tract obstruction and pulmonary arterioplasty for pulmonary artery stenosis (17 of 24 reoperations). Before the Lecompte modification was routinely performed (15), 50% (9 of 18) of the children needed reinterventions for RV outflow tract obstruction. Thereafter, the frequency decreased to 15% (7 of 47).

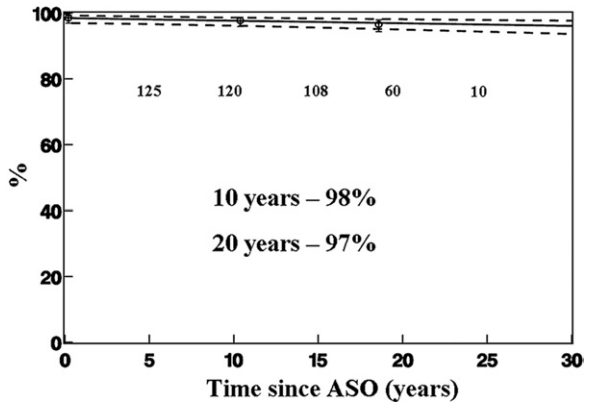


Figure 1 Survival Among 132 Operative Survivors of an ASO in Infancy

Each **circle** represents an actual death. **Solid line** represents survival for whole population. The **numbers** below the survival curve indicate the numbers of patients at risk. The **dashed lines** represent the 70% confidence interval. ASO = arterial switch operation.

Cardiac status in adulthood. The mean age at last follow-up was 21 ± 3 years (median 21 years, range 18 to 33 years, 10 patients were ≥25 years of age). Seven patients (11%) were not in sinus rhythm (junctional rhythm in 5 patients, atrial or ventricular paced rhythms in 2 patients). Cardiac death, heart failure, or myocardial infarction were not observed in the 65 adult patients followed in our institution. Five patients (8%) had a history of arrhythmia in adulthood (median age 20 years, range 19 to 27 years). Atrial flutter requiring ablation and cardioversion occurred in 3 adults, 2 of whom had RV to pulmonary artery nonvalved conduits. One adult had sick sinus syndrome with syncope and required a pacemaker implantation, and the other adult had LV systolic dysfunction (LVEF 35%) and recurrent nonsustained ventricular tachycardia and therefore was treated with amiodarone. Tachyarrhythmia in the absence of RV to pulmonary artery conduits and ventricular dysfunction was not observed.

All adults were in New York Heart Association functional class I. In a subset of 45 adults who had a cardiopulmonary

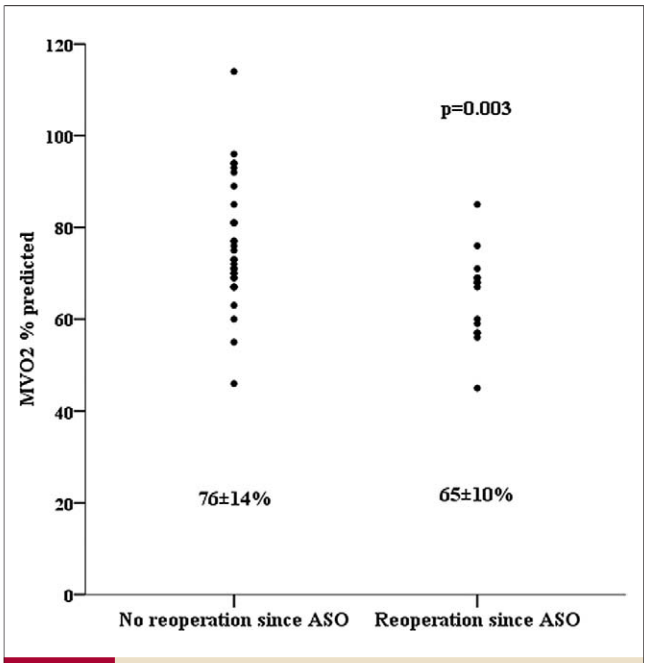


Figure 2 Oxygen Consumption in Adults With an ASO

Percentage predicted peak oxygen consumption (MVo_2) (indexed to age and sex) in patients with and without reoperation after the arterial switch operation (ASO).

exercise test, the mean MVo_2 was 73 ± 14% (range 45% to 114%) of the predicted value for age and sex. The mean age at the time of exercise testing was 19 ± 4 years. Overall, 82% (37 of 45) had abnormal exercise capacity <80% of predicted MVo_2. Adults with simple TGA had higher predicted values compared with those with complex TGA (77 ± 13% vs. 68 ± 13%, p = 0.03), and adults with a reoperations in childhood had lower predicted values compared with those without (65 ± 10% vs. 76 ± 14%, p = 0.003) (Fig. 2). Patients who did not perform a cardiopulmonary exercise test (n = 20) were less likely to have an unusual coronary pattern compared with those who performed an exercise test.

Overall, LV systolic function was normal (LVEF 60%, range 35% to 79%). The LV systolic function was mildly or moderately impaired in 14% of adult patients (9 of 65). There were no adults with severe LV systolic dysfunction. Adults with cardiac reoperations in childhood had worse LV systolic function compared with those without (LVEF 58 ± 8% vs. 63 ± 7%, p = 0.04). Ejection fraction was not statistically different between the patients with simple TGA compared with those with complex TGA.

Aortic valve regurgitation of any degree was present in 52% of adults (34 of 65). None of the patients had moderate or greater aortic valve regurgitation. Aortic root dilation (sinus of Valsalva >36 mm) was identified in 31% (20 of 65) of the adult patients (median 34 mm, range 25 to 48 mm). Aortic root dilation was more common in adults with pulmonary arterial banding before arterial switch surgery (67% vs. 28%, p = 0.02).

Myocardial scintigraphy was performed in a subset of 23 adults. A perfusion defect with exercise was present in 5 adults. In 4 of those, the size of perfusion defect was small to moderate; in the remaining patient, the perfusion defect was moderate to large. Of those 5 patients, 1 had atypical chest pain and underwent subsequent angiogram showing no evidence of coronary artery disease. The remaining 4 patients were asymptomatic, and no angiogram was performed. No therapy was started in those 5 patients.

Significant cardiac lesions and reintervention in adulthood. One or more significant cardiac lesions were present in 17% (11 of 65) of adults. Structural lesions included: moderate LV systolic dysfunction (n = 1), moderate RV systolic dysfunction (n = 3), severe pulmonary valve regurgitation (n = 2), severe tricuspid regurgitation (n = 1), and moderate LV outflow tract obstruction due to a subvalvular membrane (n = 1). As mentioned previously, 5 adults had arrhythmia that required treatment. Univariate predictors of these clinically significant lesions observed at the last follow-up are listed in Table 2. Adults with surgical palliation prior to ASO were more likely to have clinically significant residual cardiac findings compared with adults with a primary ASO in infancy (40% vs. 10%, p = 0.01). Similarly, adults who have had reinterventions in child-

Table 2 Predictors of Significant Residual Cardiac Lesions in Adulthood After ASO

	Clinically Significant Lesion (n = 11)	No Clinically Significant Lesion (n = 54)	Odds Ratio (95% CI)	p Value
Female	7 (64%)	18 (33%)	3.5 (0.9–13.5)	0.07
Complex TGA	8 (73%)	21 (39%)	4.2 (1.0–17.6)	0.05
Unusual coronary pattern	6 (55%)	17 (31%)	2.6 (0.7–9.8)	0.2
Era of ASO before 1987	4 (36%)	14 (26%)	1.6 (0.4–6.4)	0.5
Palliation prior to ASO	6 (55%)	9 (17%)	6.0 (1.5–24.0)	0.01
Age of ASO <14 days	5 (45%)	13 (24%)	2.6 (0.7–10.0)	0.2
Cardiopulmonary bypass time >180 min	6 (55%)	11 (20%)	4.7 (1.2–18.3)	0.03
RV-PA conduit	2 (18%)	5 (9%)	2.2 (0.4–13.0)	0.4
Overall reinterventions in childhood	9 (82%)	16 (30%)	10.7 (2.1–55)	0.005
Catheter interventions in childhood	5 (45%)	8 (15%)	4.8 (1.2–19.5)	0.03
Reoperations in childhood	8 (73%)	12 (22%)	9.3 (2.1–40.8)	0.003

CI = confidence interval; other abbreviations as in Table 1.

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Table 3 Reinterventions in Adulthood After ASO

Sex	Anatomy	ASO	Anastomosis	Reoperation in Childhood	Reoperation in Adulthood
F	Complex	1980	RV-PA conduit	None	Pulmonary arterioplasty, sternal wire removal
F	Complex	1980	RV-PA conduit	Conduit replacement	PVR, pulmonary arterioplasty, RVOT resection
F	Complex	1981	RV-PA conduit	Pulmonary arterioplasty	Coronary artery revision, conduit replacement
M	Complex	1986	RV-PA conduit	RVOT repair, pulmonary arterioplasty	Pacemaker replacement
F	Complex	1987	Lecompte	Mitral valve repair/replacement/PM	Mitral valve replacement
M	Complex	1989	Lecompte	Pulmonary arterioplasty	Pulmonary arterioplasty
M	Simple	1990	Lecompte	None	Pacemaker implant

PM = pacemaker; PVR = pulmonary valve replacement; RVOT = right ventricular outflow tract; other abbreviations as in Table 1.

hood were more likely to have clinically significant cardiac lesions compared with those who had not had required cardiac reinterventions in childhood (36% vs. 5%, p = 0.002).

Eleven percent (7 of 65) of adults had reinterventions in adulthood: 5 reoperations and 2 pacemaker implantations (Table 3). There were no interventions for coronary stenosis, neo-aortic valve regurgitation, or aortic root dilation. Reinterventions were performed at a median age of 20 years (range 18 to 27 years). Right ventricular to pulmonary artery conduit reoperations (replacement in 2 patients, and patch repair in 1 patient) accounted for reoperations in 3 patients. In the subgroup of patients who had their ASO after the routinely performed Lecompte modification, reintervention for pulmonary artery stenosis after age 18 years occurred in only 1 adult patient (2%). Univariate predictors of reinterventions are shown in Table 4.

At their last clinic visit, 77% (50 of 65) of the adults after ASO did not have any clinically significant cardiac lesion nor had they required a reintervention in adulthood.

Discussion

Early mortality, survival into childhood, and cardiac morbidity in children with TGA after ASO have been described by several groups (5,7,8). In contrast, our study begins to define the emerging adult cohort after ASO. The mean age of our patients was 21 ± 3 years. Generally, this early cohort of adults after ASO continues to do well with excellent long-term survival after hospital discharge. Most of the adults (77%) do not have clinically significant cardiac lesions nor have they required reinterventions in early adulthood. In our study, adult survivors who had a primary ASO (i.e., were not palliated) and who had no childhood reinterventions are doing exceptionally well. In contrast, those adults with a RV to pulmonary artery conduit are at highest risk for having a cardiac reintervention in adulthood. Adults with surgical palliation before the ASO and those with reintervention in childhood are more likely to have significant cardiac lesions as adults. In the current era of ASO, RV to pulmonary artery conduits are very rarely used, and ASO is usually performed without prior palliation. Therefore, it is anticipated that there will be fewer clinically significant lesions in the upcoming group of young adults with ASO.

Functional status of the adult ASO survivor. A notable finding in our study was that most young adults do not have normal MVO_2. In contrast, 1 study in children (mean age 9 years) showed that MVO_2 was normal or low-normal relative to age- and sex-predicted values (16). The discrepancy may be explained by the nature of our early ASO cohort, who often required reintervention in childhood for RV outflow tract obstruction. RV outflow tract obstruction has a known adverse effect on exercise capacity (17). Abnormal coronary blood flow reserve, seen in children after ASO (18), might also contribute to our findings of reduced MVO_2 in young adult patients. In our study,

Table 4 Predictors of Cardiac Intervention in Adulthood After ASO

	Reintervention (n = 7)	No Reintervention (n = 58)	Hazard Ratio (95% CI)	p Value
Female	4 (57%)	21 (36%)	1.8 (0.4–8.4)	0.4
Complex TGA	6 (86%)	23 (40%)	6.4 (0.8–54.2)	0.08
Unusual coronary pattern	4 (40%)	19 (33%)	0.8 (0.2–4.0)	0.8
Era of ASO before 1987	4 (57%)	14 (24%)	2.0 (0.4–10.3)	0.4
Palliation prior to ASO	3 (43%)	12 (21%)	1.6 (0.3–7.5)	0.6
Age of ASO <14 days	5 (71%)	13 (22%)	4.3 (0.8–23.6)	0.09
Cardiopulmonary bypass time >180 min	3 (43%)	14 (24%)	1.4 (0.3–6.8)	0.7
RV-PA conduit	4 (57%)	3 (5%)	9.1 (1.9–44.2)	0.006
Catheter intervention in childhood	2 (29%)	11 (19%)	1.7 (0.3–9.4)	0.5
Reoperation in childhood	5 (71%)	15 (26%)	4.2 (0.8–22.3)	0.09

Abbreviations as in Tables 1 and 2.

patients who underwent a cardiopulmonary exercise test were more likely to have an unusual coronary pattern compared with those with no exercise testing. This finding requires further observation in the long-term follow-up.

Ventricular dysfunction. For patients with complete TGA and atrial switch repairs, the morphologic right ventricle supports the systemic circulation and progressive dysfunction is a major concern and impacts long-term outcome (19–24). It is less likely to be an issue after an ASO, since the LV is the subaortic ventricle, and no evidence of early time-related deterioration of function has been reported (25,26). Good LV systolic function was reported in older children (27), and in the absence of coronary artery abnormalities, severe LV systolic dysfunction is not expected. As demonstrated in our study, patients with a cardiac reoperation in childhood are at higher risk for LV systolic dysfunction. Although we found that 14% of adults had some degree of ventricular dysfunction, with fewer childhood reinterventions expected, ventricular function is likely to be preserved in the upcoming generation of adults after ASO. We did not examine diastolic function.

Neo-aortic valve regurgitation and aortic root dilation. Progression of neo-aortic valve regurgitation is reported in childhood after ASO (28). Similarly, aortic root dilation can occur after ASO, but may not to be progressive later in childhood (11). In our cohort, none of the adults had significant aortic valve regurgitation at last follow-up or needed replacement of the neo-aortic valve or the aortic root. Further follow-up is needed to evaluate whether aortic valve regurgitation and aortic root dilation will progress in later adulthood, specifically when these patients start to develop acquired adult onset disease such as arterial hypertension.

Coronary complications. Although coronary events have been reported in 7% of childhood ASO survivors (12), we did not observe coronary events in our cohort. However, coronary disease may be present, but not recognized clinically. In children after ASO, a proportion of patients with ASO had reversible myocardial perfusion defects and mild wall motion abnormalities (29). With time, problems related to abnormal coronary reserve may become manifest. Our study was not able to address these issues.

Reinterventions in adulthood. Cardiac reinterventions after age 18 years, when they occur, are usually for relief of RV outflow tract obstruction and pulmonary artery stenosis. In childhood, pulmonary artery stenosis was the most common reason for late reoperation after an ASO (8,10). However, with improvement in arterial switch surgeries, this complication has been almost eliminated (30). The high rate of reinterventions for RV outflow tract obstruction and/or pulmonary artery stenosis in childhood in our cohort reflects the large proportion of survivors who had an ASO prior to the routinely performed Lecompte modification and in whom RV to pulmonary artery connection was frequently maintained by a conduit. Not surprisingly, patients with a RV to pulmonary artery conduit have the highest prevalence of cardiac reinterventions as adults.

Study limitations. The short follow-up in adulthood is the main limitation of the present study. As this cohort ages, longer follow-up will become available, and cardiac outcomes may differ. In the interim, this study highlights the group of adults at highest risk for cardiac complications and the need for continued surveillance. Only the subset of ASO survivors followed at our institution had detailed follow-up for residual cardiac lesions and reinterventions. However, the group not seen at our clinic had baseline characteristics that differed little from our cohort. Another limitation is the retrospective nature of the study. Because of the small numbers and the restricted age range, serial electrocardiographic and echocardiographic measures over time were not studied. A multivariate analysis could not be performed to determine predictors for cardiac lesions and reinterventions because of the relatively low event rates. Finally, assessment of coronary arteries and coronary perfusion defects was not routinely performed in our institution as appropriate protocols are lacking. The significance of abnormal coronary perfusion is unclear, and the potential for coronary complications remains a concern as this population continues to age.

Conclusions

Our study highlights the overall good outcomes in this early cohort of young adults after ASO. Nevertheless, there are subgroups, particularly those who needed further intervention in childhood, who are at higher risk for ventricular and valve dysfunction and arrhythmias. This study stresses the importance of lifelong surveillance of patients with an ASO for TGA in specialized adult congenital heart disease centers.

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