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Outcomes and reoperations after total correction of complete atrio-ventricular septal defect

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Abstract

Background: Surgical correction of complete atrio-ventricular septal defect (AVSD) achieves satisfactory results with low morbidity and mortality, but may require reoperation. Our recent operative results at mid-term were followed-up. **Methods:** From June 2000 to December 2007, 81 patients (Down syndrome; $n = 60$), median age 4.0 months (range 0.7–118.6) and weight 4.7 kg (range 2.2–33), underwent complete AVSD correction. Patch closure for the ventricular septal defect (VSD; $n = 69$) and atrial septal defect (ASD; $n = 42$) was performed with left atrio-ventricular valve (LAVV) cleft closure ($n = 76$) and right atrio-ventricular valve (RAVV) repair ($n = 57$). Mortality, morbidity, and indications for reoperation were retrospectively studied; the end point 'time to reoperation' was analyzed using Kaplan–Meier curves. Follow-up was complete except in two patients and spanned a median of 28 months (range 0.4–6.1 years). **Results:** In-hospital mortality was 3.7% ($n = 3$) and one late death occurred. Reoperation was required in 7/79 patients (8.9%) for LAVV insufficiency ($n = 4$), for a residual ASD ($n = 1$), for right atrio-ventricular valve insufficiency ($n = 1$), and for subaortic stenosis ($n = 1$). At last follow-up, no or only mild LAVV and RAVV insufficiency was present in 81.3% and 92.1% of patients, respectively, and 2/3 of patients were medication-free. Risk factors for reoperation were younger age (<3 months; $p = 0.001$) and lower weight (<4 kg; $p = 0.003$), and a trend towards less and later reoperations in Down syndrome ($p < 0.2$). **Conclusions:** Surgical correction of AVSD can be achieved with low mortality and need for reoperation, regardless of Down syndrome or not. Immediate postoperative moderate or more residual atrio-ventricular valve insufficiency will eventually require a reoperation, and could be anticipated in patients younger than 3 months and weighing <4 kg.

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Keywords: Atrio-ventricular septal defect; Surgery; Congenital heart disease

1. Introduction

Complete surgical correction of atrio-ventricular septal defect (AVSD) in infancy is a routine procedure, and early repair before the onset of irreversible pulmonary hypertension is mandatory to avoid immediate postoperative and long-term morbidity. Despite tremendous advances in technology, surgical technique, and understanding of the baseline pathophysiology and anatomy of this congenital heart defect, the results are satisfactory, but have not drastically improved in the past decade, as can be illustrated with mortality rates still ranging between 8.7 and 21.7% [1–5], and reoperation rates of 6.4–16.6% [2–4,6,7]. We review our recent experience with regards to mid-term results and

the rate of reoperation, and the risk factors or pitfalls influencing the latter.

2. Materials and methods

Permission to proceed with this study was granted by the institutional review board of our hospital. Retrospective chart review was performed for in-hospital data retrieval, and follow-up was obtained either by clinical and echocardiographic analysis in our own outpatient clinic, or by mail request of documents from referring pediatric cardiologists. Between June 2000 and December 2007, 81 consecutive patients (63% female) underwent primary surgical repair of complete AVSD in our unit. Patients with associated tetralogy of Fallot, single papillary muscle, unbalanced ventricles, and anomalous pulmonary venous connections were excluded. Two patients had prior coarctation repair through a left thoracotomy. One small-for-gestational-age newborn with

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Table 1
Patient demographics and results

Age	4.0 months (0.7 months to 9.9 years)
Weight	4.7 kg (2.2–33)
Down syndrome (<i>n</i>)	60
Hospital stay	17 days (8–142)
Complications	Chylothorax (<i>n</i> = 9; 11.1%) Pacemaker (<i>n</i> = 3; 3.7%) JET (<i>n</i> = 2; 2.5%) Temporary dialysis (<i>n</i> = 2; 2.5%) ECMO (<i>n</i> = 2; 2.5%)
Time to reoperation	145 days (6–510)
LAVV repair	4
RAVV repair	1
ASD closure	1
Subaortic stenosis resection	1
Mortality	<i>n</i> = 3; 3.7%

LAVV: left atrio-ventricular valve insufficiency; RAVV: right atrio-valve insufficiency; ASD: atrial septal defect; JET: junctional ectopic tachycardia; ECMO: extracorporeal membrane oxygenation.

severe cardiac failure underwent palliative pulmonary artery banding. Despite our policy to perform elective repair around 4 months of age, our consecutive series contains 13 patients older than 1 year who either had small restrictive ventricular septal defects (VSD), mild-to-moderate pulmonary stenosis, or were referred late. Preoperative pulmonary hypertension, defined as a systolic pulmonary pressure greater than half-systolic systemic pressure, was present in 6/21 patients with normal chromosomes (28.6%), and in 43/60 patients with Down syndrome (71.7%). Further patient demographics are given in Table 1.

Surgery was performed through median sternotomy, using full cardiopulmonary bypass with moderate hypothermia (26–32 °C), intermittent antegrade cold blood cardioplegia, and repair through a right atriotomy. Patch closure was performed for the atrial septal defect (ASD) in 51.9% of cases (*n* = 42), and for closure of the VSD in 85.2% of cases (*n* = 69). In the remaining patients, the atrial defects were small enough to be closed directly, as described by our senior author [8], or the ventricular defects were judged to be better closed directly to simplify repair without division of the common atrio-ventricular valves, as described by Nicholson et al. [9]. Concomitant repair of the left and right atrio-ventricular valves (RAVVs) was required in 76 (93.8%) and 57 (70.4%) instances, respectively. These included not only closure of the clefts in the atrio-ventricular valves, but also reduction de Vega or partial annuloplasty sutures, as required. The coronary sinus was systematically allowed to drain in the right atrium. Intra-operative transesophageal echocardiography (TEE) was routine in all cases, allowing for preoperative confirmation of the anatomy, as well as assessment of the repair at the end of surgery. In cases of large residual shunts with hemodynamic significance (>2 mm), or more than moderate atrio-ventricular valve insufficiency, as detected by TEE after weaning from cardiopulmonary bypass, revision of the repair was undertaken to correct residual lesions.

2.1. Statistical methods

Data are expressed as mean ± standard deviation or median (range), as appropriate. The end point 'time to

reoperation' has been analyzed using Kaplan–Meier curves, and the significance of its relationship with binary factors (Down/non-Down; weight <4 kg/>4 kg; age at surgery <3 months/>3 months) has been assessed using log rank tests. A *p* value smaller than 0.05 was considered significant.

3. Results

Follow-up was complete except for two patients living abroad, and spanned a median of 28 months (range 0.4 months to 6.1 years). Median cardiopulmonary bypass time was 140 min (range 58–270 min), and median aortic cross-clamp time was 82 min (range 32–189 min). Intra-operatively at the end of cardiopulmonary bypass, TEE disclosed a residual significant (>2 mm) intra-cardiac shunt in five instances, requiring a second run on cardiopulmonary bypass to close the residual defect. Also, more than moderate residual left atrio-ventricular valve (LAVV) insufficiency was detected by TEE in 13 cases, requiring a second run on bypass to handle either a residual cleft, or to achieve better leaflet coaptation with a partial annuloplasty. Postoperatively, liberal use of an open chest with delayed sternal closure on the intensive care unit was preferred, and was performed in nine patients, two of which needed an open chest for postoperative extracorporeal membrane oxygenation (ECMO) support. For patients with pulmonary hypertension and hemodynamic compromise upon weaning from cardiopulmonary bypass, liberal postoperative use of inhaled nitric oxide (maximum 20 ppm) was made on the intensive care unit until subsequent oxygen and ventilator weaning could be achieved (*n* = 24). The incidence of postoperative complications is given in Table 1. The duration of hospitalization ranged from 8 to 142 days (median 17), and did not vary significantly between Down (median 17 days; range 8–142) and non-Down patients (median 14; range 8–118).

There was no intra-operative mortality, but three cases of in-hospital mortality, all in children with Down syndrome (3.7%). One patient died 4 days after surgical repair, due to left ventricular failure, hemorrhagic lung edema, acute renal insufficiency, hemorrhagic ischemic enterocolitis, and ultimately multi-organ failure. Another required neonatal total repair at 1 month of age, with a fenestrated ASD and postoperative ECMO support. Postoperative severe right atrio-ventricular valve insufficiency warranted right-sided valve replacement with a 12 mm Contegra bovine jugular vein graft 6 days later, which allowed weaning from ECMO. The patient eventually died from sepsis and multi-organ failure on postoperative day 23. Finally, one patient underwent successful complete AVSD repair with a perfect result as documented on echocardiography, but developed an acute and severe pulmonary infection with respiratory syncytial virus on postoperative day 10 requiring respiratory ECMO support, which lasted 9 days until multi-organ failure and pulmonary superinfection led to ECMO discontinuation and the patient's demise. The one late death occurred 2 months after hospital discharge in a patient with Down syndrome, initially with very low birth weight and failure to thrive, an initial pulmonary artery banding followed by complete repair and resection of subaortic stenosis at the age of 3 months, with a stormy postoperative course due to interstitial lymphangiectasia of

the lungs with concomitant pulmonary hypertension. The infant died at home, probably subsequent to aspiration, while still on oxygen supplementation for interstitial lung disease and a naso-gastric feeding tube.

No or only mild LAVV insufficiency was noted in 81.3% of patients at last follow-up (61/75). In 14 patients there was moderate or severe valve dysfunction (18.7%), requiring reoperation in four cases. At reoperation, a combination of residual cleft and annular dilatation was encountered, and accordingly handled with residual cleft closure and/or reduction de Vega annuloplasty. After redo surgery for LAVV insufficiency, achieved without mortality, residual moderate ($n = 1$) and severe ($n = 1$) LAVV insufficiency were still present in two patients, indicating the difficulty in attempting repeat repair of a dysmorphic valve.

No or only mild RAVV regurgitation was present in 92.1% of patients at last follow-up (70/76), although 6.6% (5/76) had a deterioration of their RAVV function in time. Three of them underwent a reoperation for other indications, with concomitant RAVV repair in one case. The other two had no complications and were medication-free at last follow-up. One patient required reoperation specifically for right atrio-ventricular valve insufficiency (1/76; 1.3%), with valve replacement, as described above, and eventually expired.

Postoperative junctional ectopic tachycardia occurred in two patients (2.5%), successively treated with patient cooling and intravenous amiodarone. Third degree heart block requiring insertion of an epicardial pacemaker system occurred in three patients (3.7%), and was performed during the same hospitalization as the complete AVSD repair.

Altogether, reoperation was necessary in seven patients (7/79; 8.9%), and was performed at a median of 145 days after initial repair (range 6–510 days). Indications for redo surgery were LAVV insufficiency ($n = 4$), right atrio-ventricular valve insufficiency ($n = 1$), a residual ASD ($n = 1$), and subaortic stenosis ($n = 1$). More than half of the patients requiring a reoperation had Down syndrome ($n = 4$). As illustrated in Fig. 1, although the time to reoperation was shorter in non-Down patients, non-Down patients did not reach statistical significance with regards to the need for reoperation ($p < 0.2$). Both younger age (< 3 months) and

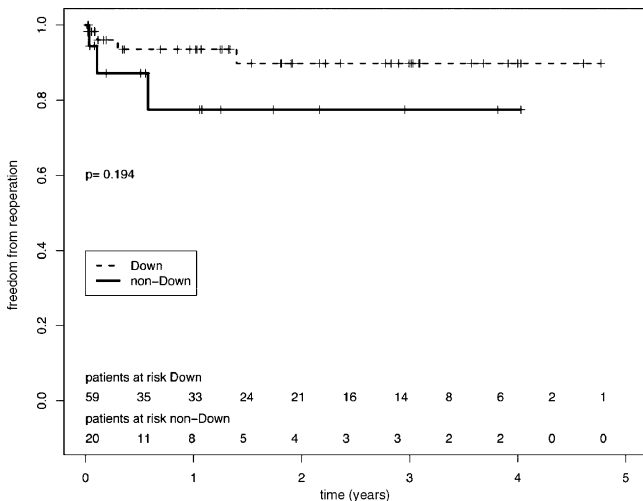


Fig. 1. Freedom from reoperation between Down and non-Down patients.

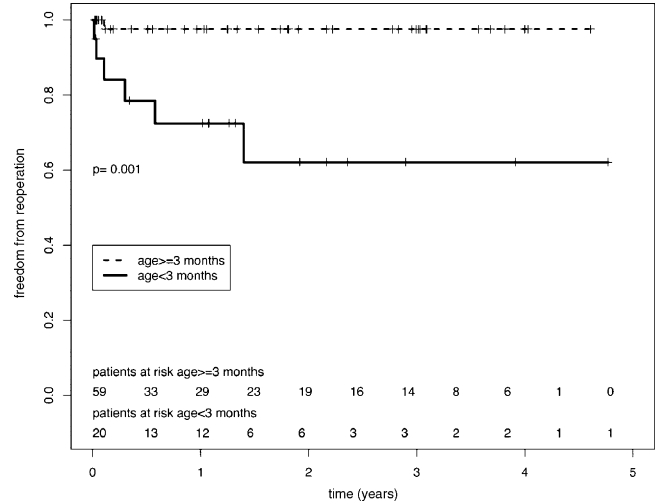


Fig. 2. Freedom from reoperation with regards to age at initial correction.

lighter weight (< 4 kg) were associated with a higher risk for earlier reoperation ($p = 0.001$ and $p = 0.003$, respectively), as can be seen in Figs. 2 and 3, respectively. There were no early or late complications after reoperation for LAVV.

Six small residual ASDs, of which two were intentional fenestrations, spontaneously closed after a median of 200 days. The intentional fenestrations were left in two patients with borderline smaller left ventricles, where two-ventricle repair was judged feasible. In two patients, small residual ASDs (< 2 mm) remained open during follow-up. In three patients, although no residual ASD was detected on post-operative echocardiography, a shunt at the atrial level was detected at last follow-up. The initial repair had been by direct ASD closure in all three cases, implicating perhaps excessive tension on the direct closure suture lines at primary repair, which may have dehisced subsequently. Only one of these patients required a reoperation for ASD closure owing to a hemodynamically significant left-to-right shunt.

Nine residual VSDs closed spontaneously after a median of 226 days. In four patients, an initial complete closure or a small residual VSD defect evolved into a larger residual shunt.

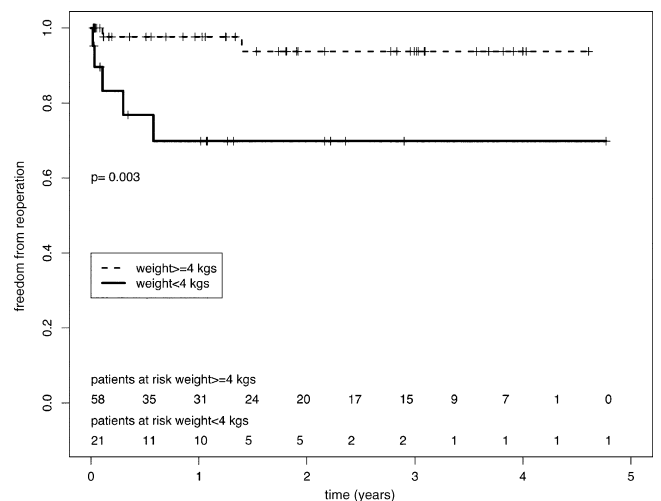


Fig. 3. Freedom from reoperation with regards to weight at initial correction.

Two of these patients required reoperation, not only for the VSD, but for residual LAVV insufficiency, which were concomitantly addressed during redo surgery.

Four patients (4/75; 5.3%) had evidence of turbulence in the left ventricular outflow tract after initial repair, which persisted until last follow-up. Only one of them required redo surgery for a subaortic stenosis gradient >50 mmHg, consisting of an uneventful resection of subaortic fibrous tissue, which has not recurred at last follow-up.

4. Discussion

Surgical mortality after AVSD repair varies tremendously and is institution-dependent, ranging from 3.0 to 21.7% [1–5]. Our mortality was 3.7%, which compares to these results, reflecting the overall improvements achieved by many centers in the more recent era. Postoperative morbidity and rate of reoperation also vary widely, depending on the degree of expertise at a given institution. Indications for reoperation include left or right AVV insufficiency, residual intracardiac shunts, and subaortic stenosis, whose rate in the literature varies between 6.4 and 16.6% [2–4,6,7], similar to that found in our series (10.5%). We found age younger than 3 months ($p = 0.001$) and weight less than 4 kg ($p = 0.003$) to be independent risk factors for reoperation. The goal of the study was to analyze our recent results with a standardized technique in a consecutive series of patients with relatively straightforward anatomical variants of AVSD. Therefore, concomitant anomalies with Fallot-type right outflow obstruction, single papillary muscle or unbalanced ventricles leading to single ventricle repair, or other complex anomalies were excluded in an effort to reduce confounding factors relevant to survival or reoperation.

4.1. Age and weight at repair

The optimal weight and age at operation has evolved over time, and is still controversial. Down syndrome is a risk factor for rapid progression of obstructive pulmonary vascular disease [10], which justifies the more aggressive approach towards earlier operative correction in these infants. Previously, early repair was considered as that up until 6 months of age [7,11,12]. Earlier repair with closure of shunts and the normalization of hemodynamics improves quality of life [12] and reduces operative mortality [13]. Currently, it is generally agreed that pulmonary hypertension develops earlier, will inevitably progress and worsen, and will negatively influence perioperative morbidity and perhaps mortality. Therefore, more centers including our own, are advocating earlier elective repair around 3–4 months of age. Although some concern has been raised as to the increased surgical mortality [3] when repair is performed before 6 months of age, this risk factor has been neutralized in the current era in most centers. In our experience, postoperative residual moderate or severe LAVV insufficiency was more frequent in patients when operated upon before 3 months of age. The mean age of the seven patients requiring reoperations was 2 months, and occurred in patients with prior intractable heart failure due to very large intracardiac shunts and/or pre-existing valve insufficiency. Both the increased fragility of valvar tissues

and the global cardiac dilatation from insufficiency forced us to operate on these patients at a younger age than the usual routine at our institution. Because of the potential increased risk for reoperation in the very young, we maintain our bias towards early complete repair around 3–4 months, in order to avoid the detrimental effects from ongoing pulmonary hypertension with increasing age, and the added postoperative morbidity/mortality there unto pertaining.

Studies by Günther and Prifti [3,14] revealed that weight less than 5 kg was a predictor for longer postoperative ventilatory assistance, longer intensive care stay, and need for reoperation, as compared to larger babies. Contrary to these findings, Al-Hay et al. [6] found no correlation between weight and need for reoperation. Our study shows that infants eventually requiring redo surgery had a lower median weight at first repair (3.5 kg), compared to the median weight of the total group (5.9 kg).

4.2. Down syndrome

In our experience, patients with trisomy 21 have a different atrio-ventricular valve apparatus than non-Down patients. In Down patients, valve tissue is more abundant, and allows for an easier reconstruction, leading to fewer reoperations for LAVV insufficiency. Confirming our impression, Al-Hay et al. [6] reported that dysplasia of the left atrio-ventricular valve is more common in non-Down patients, and thus is the main indication for reoperation in these patients, a finding also confirmed by others [2,15]. Along the same lines, Formigari et al. [16] argued a higher prevalence of recurrent/residual subaortic stenosis in non-Down patients, and a lower prevalence of reoperation in patients with Down syndrome. Similarly, Rizzoli et al. [17] described that Down syndrome was not an independent risk factor for operative mortality, and that they underwent fewer reoperations. Somewhat contrary to these findings, some have stated that Down syndrome is a potential risk factor for hospital and late death [13,18], as well as for reoperation [13]. We could not confirm this in our series, albeit our numbers may be too small to detect significance. Down patients presented more postoperative non-cardiac complications (16/60; 26.7%) than non-Down patients (2/21; 9.5%), although this did not prolong hospital length of stay, which was similar in both patient groups.

4.3. Left atrio-ventricular valve insufficiency and reoperation

LAVV insufficiency is the most common indication for reoperation in patients after complete AVSD repair [1,3,13,15,19,20], although the degree of insufficiency one may tolerate is not clear from the extant literature. Pozzi et al. [13] stated that preoperative valve incompetence was a significant risk factor for LAVV insufficiency after AVSD complete repair, and Günther [3] found that it was even a risk factor for operative mortality, although others [1,21] refute these findings. In one study, a lower reoperation rate for LAVV insufficiency was observed when using the two-patch technique [15], although we cannot confirm this finding.

Moderate to severe LAVV insufficiency was present in 12/81 (14.8%) of our patients, although only four of these patients

with moderate ($n = 3$) and severe ($n = 1$) LAVV insufficiency required reoperation. Despite systematic intra-operative TEE assessment of repair and the tremendous adjunct it represents with regards to intra-operative understanding of the mitral valve apparatus, this echocardiographic diagnostic tool has its shortcomings, and underestimates the degree of actual LAVV insufficiency [22]. Our policy is to accept only mild residual MI, and to go back on cardiopulmonary bypass to attempt re-repair of anything more severe. However, we and others [22] have increasingly noticed that residual LAVV insufficiency is difficult to quantify intra-operatively using TEE, with a frustrating discrepancy observed between immediate post-bypass TEE and the transthoracic echocardiography performed days to weeks later. Accordingly, we have documented initial satisfactory repairs of the LAVV intra-operatively, only to observe a deterioration of LAVV insufficiency after weeks or months, some of which even required reoperation. This is mirrored by the majority (4/6) of our patients who initially had a fully uneventful and uncomplicated postoperative course and who eventually went on to require a reoperation. An uneventful postoperative course is therefore not a surrogate for a reoperation-free future, and minimal residual or recurring lesions may potentially evolve and worsen. None of these patients had postoperative endocarditis to explain an infectious dehiscence of suture lines in the cleft or patch material.

Reconstruction of recurrent or residual LAVV insufficiency at reoperation is difficult, as illustrated by two patients in our series who maintained moderate and severe valve insufficiency even after attempted redo reconstruction. Therefore, every effort should be made at the primary complete correction to avoid residual LAVV insufficiency, with the hope that it will not recur or worsen with patient growth. Poirier et al. [7] have described a successful technique to deal with this difficult issue, with satisfactory results.

4.4. Residual defects

There are few representative studies about residual atrial and ventricular septal defects. In our study, after primary surgical repair until last follow-up, 88.9% (72/81) of patients had no residual septal defects. In their series of 158 patients, Backer et al. [15] described no patients requiring a reoperation for residual shunts. We found only 1/81 patient (1.2%) who required a reoperation for a significant residual ASD. Nine small residual VSDs closed spontaneously after a median of 226 days. Of the four patients with a residual VSD >2 mm, two underwent a reoperation for concomitant shunt closure with the primary surgical indication of LAVV valve repair.

Residual or recurrent subaortic stenosis is a potential critical lesion requiring close follow-up, and eventual reintervention [4,23]. We found subaortic stenosis in four patients, in whom one required a second operation. We surgically address this lesion when the peak gradient reaches 50 mmHg, or with any evidence of aortic valve insufficiency detected by echocardiography, so as to avoid progressive left ventricular hypertrophy and further aortic valve damage.

4.5. Arrhythmia and heart block

Our incidences of postoperative JET (2.5%) and complete heart block requiring a pacemaker (3.7%) are similar to those

found in the literature [1–6], illustrating the standardized surgical technique and improvements in intensive care management one expects in centers experienced with patients congenital heart disease.

5. Conclusion

Complete elective repair of AVSD around 3–4 months of age, or semi-urgently with a weight >4 kg in the neonatal period, can be achieved with low mortality rates. The aim is to restore normal hemodynamics, and avoid the consequences of progressive pulmonary hypertension, without increasing the risk for reoperation. In the vast majority of cases, small residual intracardiac shunts (<2 mm) will spontaneously close in time, and seldom require reintervention. Intra-operatively, TEE may underestimate mild to moderate MI, which can deteriorate in time, despite an uneventful initial postoperative course. Improvements in intra-operative assessment of repair with regards to the left atrio-ventricular valve are mandatory, in order to reduce the incidence of reoperations in the future. Moderate or more severe residual LAVV insufficiency is the most frequent indication for reoperation, and is more prevalent in patients with normal chromosomes, as compared to patients with Down syndrome.

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