A single-centre 37-year experience with reoperation after primary repair of atrioventricular septal defect†

Vladimir Sojak*, Marlotte Kooij, Aria Yazdanbakhsh, Dave R. Koolbergen, Eline F. Bruggemans and Mark G. Hazekamp

Department of Cardiothoracic Surgery, Leiden University Medical Center, Leiden, Netherlands

* Corresponding author. Department of Cardiothoracic Surgery, Leiden University Medical Center, Albinusdreef 2, 2333 ZA Leiden, Netherlands. Tel: +31-715262348; fax: +31-715248284; e-mail: v.sojak@lumc.nl (V. Sojak).

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Abstract

OBJECTIVES: To evaluate our experience with patients reoperated after primary repair of atrioventricular septal defect (AVSD) and identify predictors of poor outcome.

METHODS: Between 1976 and 2014, 69 patients were reoperated after primary repair of partial (n = 28), intermediate (n = 15) or complete (n = 26) AVSD.

RESULTS: Median age at first reoperation was 62.4 (range, 1.6–845) months, median interval to first reoperation was 22.3 (range, 0.2–598) months. Main indications for first reoperation included left atrioventricular valve (LAVV) pathology (66%), residual septal defect (19%) and left ventricle outflow tract obstruction (LVOTO; 4%). Procedures to address LAVV pathology included various valvuloplasties in 47 (77%) patients and valve replacement in 14 (23%) patients. A second, third, fourth and fifth reoperation was required in 54 (8). Procedures most commonly performed were LAVV replacement (LAVVR), LVOTO relief, pacemaker implantation and right atrioventricular valve repair. Freedom from reoperation after LAVV valvuloplasty (LAVVP) was 84 and 62% at 1 and 10 years, respectively. There were 10 early and 4 late deaths. Estimated overall survival at 1, 5 and 10 years was 87, 83 and 83%, respectively. Double orifice LAVV (DOLAVV) was a risk factor for in-hospital and overall mortality [odds ratio (OR) = 14.5; 95% confidence interval (CI) = 1.2–178.7; P = 0.037 and hazard ratio (HR) = 6.8; 95% CI = 1.5–31.7; P = 0.015, respectively]. Patients with LAVVP and LAVVR differed significantly in overall survival (P = 0.014). At the last follow-up (median, 9.8; range, 0–34 years), 84% survivors were in New York Heart Association Class I or II.

CONCLUSIONS: Many patients reoperated after primary AVSD repair needed surgical reintervention. LAVV pathology was the most common indication for reoperation. DOLAVV was a risk factor for mortality. Particular AVSD type did not appear to be a risk factor for mortality or LAVVP failure. There is some evidence for the close relationship between LAVV pathology and LVOTO in subjects undergoing reoperation after primary AVSD repair as some patients with initial LAVV problems needed LVOTO repair later on and vice versa.

Keywords: Atrioventricular septal defect · Valve repair · Valve replacement · Left ventricle outflow tract obstruction · Reoperation · Risk factors

INTRODUCTION

Atrioventricular septal defect (AVSD) represents a spectrum of congenital heart defects characterized by incomplete development of the septal tissue at atrial and/or ventricular levels along with abnormalities of atrioventricular valves (AVVs).

The results of surgical treatment of AVSD have improved significantly in the last 60 years [1, 2]. However, despite excellent survival, a significant number of patients need a surgical reintervention mainly because of severe left AVV (LAVV) regurgitation or left ventricular outflow tract obstruction (LVOTO) [3–5]. Recently, we published a 30-year experience with surgical correction of AVSD and long-term results of reoperation for LAVV regurgitation after correction of AVSD [6, 7]. The objective of our current study is to evaluate experience in patients undergoing reoperation after primary AVSD repair from all causes and to identify predictors of poor outcome.

PATIENTS AND METHODS

From January 1975 to April 2014, 457 consecutive patients received biventricular repair of AVSD at Centre for Congenital Heart Defects Amsterdam Leiden, Netherlands. The review of our surgical database has revealed 79 patients undergoing AVSD redo...
surgery at our institution. Inpatient and outpatient data were retrieved from individual medical records. Ten patients were lost to the follow-up. The complete data are available for 69 patients who enrolled in the current study. The study was approved with a waiver of consent by the local Ethics Committee because of its retrospective nature.

**Patient characteristics**

In the study cohort, 28 patients (40.6%) had partial AVSD (PAVSD), 15 patients (21.7%) had intermediate AVSD (IAVSD) and 26 patients (37.7%) had complete AVSD (CAVSD). There were 29 males and 40 females. The incidence of Down syndrome was 40.6%. The frequency of LAVV dysplasia and double orifice LAVV (DOLAVV) was 21.7 and 4.3%, respectively. Two of 3 patients with LAVV pathology, for first reoperation after primary AVSD repair were as follows: LAVV pathology (n = 61, 65.6%), residual septal defect (n = 18, 19.4%), LVOTO (n = 4, 4.3%), right AVV (RAVV) pathology (n = 4, 4.3%), cardiac arrhythmia requiring pacemaker (PM) implantation (n = 3, 3.2%), right ventricle outflow tract obstruction (RVOTO, n = 2, 2.2%) and endocarditis (n = 1, 1.1%) (Table 2).

At first reoperation, 47 patients (77%) and 14 patients (23%) who were reoperated on for LAVV pathology underwent LAVV valvuoplasty (LAVVP) and replacement (LAVVR), respectively. Residual septal defects (ASD = 7, VSD = 11) were closed in 17 patients. Four patients underwent RAWV repair. Four patients underwent LVOTO relief (membrane resection = 3, septal myectomy = 1). Three patients received PM. Two patients had RVOTO relief (muscle bundle resection + transannular patch). One patient with damaged LAVV due to endocarditis underwent LAVVR.

**Surgical technique**

All reoperations for residual structural defects were performed via median sternotomy using cardiopulmonary bypass under mild or moderate hypothermia. Myocardial protection was achieved by repeated administration of cold St Thomas crystalloid cardioplegia every 30 min. The choice of particular surgical technique was based on the indication for reoperation, intraoperative transoesophageal echocardiography and anatomical findings. In case of LAVV pathology, the valve was inspected for possible valvuloplasty. The LAVV ‘cleft’ dehiscence was addressed by direct

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**Table 1: Patient characteristics**

<table>
<thead>
<tr>
<th></th>
<th>PAVSD</th>
<th>IAVSD</th>
<th>CAVSD</th>
<th>Total</th>
<th>P-values</th>
</tr>
</thead>
<tbody>
<tr>
<td>n (%)</td>
<td>28 (40.6)</td>
<td>15 (21.7)</td>
<td>26 (37.7)</td>
<td>69 (100)</td>
<td></td>
</tr>
<tr>
<td>M/F</td>
<td>13/15</td>
<td>4/11</td>
<td>12/14</td>
<td>29/40</td>
<td>0.395</td>
</tr>
<tr>
<td>Down syndrome, n (%)</td>
<td>3 (10.7)</td>
<td>3 (20)</td>
<td>22 (84.6)</td>
<td>28 (40.6)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>LAVV dysplasia, n (%)</td>
<td>5 (17.9)</td>
<td>4 (26.7)</td>
<td>6 (23.1)</td>
<td>15 (21.7)</td>
<td>0.808</td>
</tr>
<tr>
<td>DOLAVV, n (%)</td>
<td>2 (7.1)</td>
<td>0 (0)</td>
<td>1 (3.8)</td>
<td>3 (4.3)</td>
<td>0.792</td>
</tr>
<tr>
<td>Heterotaxy, n (%)</td>
<td>5 (17.9)</td>
<td>3 (20)</td>
<td>0 (0)</td>
<td>8 (11.6)</td>
<td>0.042</td>
</tr>
<tr>
<td>Coarctation, n (%)</td>
<td>2 (7.1)</td>
<td>0 (0)</td>
<td>1 (3.8)</td>
<td>3 (4.3)</td>
<td>0.792</td>
</tr>
<tr>
<td>Tetralogy of Fallot, n (%)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>5 (19.2)</td>
<td>5 (7.2)</td>
<td>0.013</td>
</tr>
<tr>
<td>Age at first redo (median, range) in months</td>
<td>52.5 (1.6–830)</td>
<td>40.9 (2.3–293)</td>
<td>4.8 (1.8–904)</td>
<td>17.2 (1.6–830)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Time to first redo (median, range) in months</td>
<td>184.9 (1.6–845)</td>
<td>54.0 (3.1–631)</td>
<td>30.8 (2.2–353)</td>
<td>62.4 (1.6–845)</td>
<td>0.001</td>
</tr>
</tbody>
</table>

PAVSD: partial atrioventricular septal defect; IAVSD: intermediate atrioventricular septal defect; CAVSD: complete atrioventricular septal defect; M: males; F: females; LAVV: left atrioventricular valve; DOLAVV: double orifice left atrioventricular valve.

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**Table 2: Indications for first reoperation**

<table>
<thead>
<tr>
<th></th>
<th>PAVSD</th>
<th>IAVSD</th>
<th>CAVSD</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>LAVV pathology, n (%)</td>
<td>26 (74.3)</td>
<td>12 (60)</td>
<td>23 (57.5)</td>
<td>61 (65.6)</td>
</tr>
<tr>
<td>Residual septal defect, n (%)</td>
<td>5 (14.3)</td>
<td>3 (15)</td>
<td>10 (25)</td>
<td>18 (19.4)</td>
</tr>
<tr>
<td>LVOTO, n (%)</td>
<td>1 (2.9)</td>
<td>1 (5)</td>
<td>2 (5)</td>
<td>4 (4.3)</td>
</tr>
<tr>
<td>RAVV pathology, n (%)</td>
<td>2 (5.7)</td>
<td>1 (5)</td>
<td>1 (2.5)</td>
<td>4 (4.3)</td>
</tr>
<tr>
<td>Pacemaker implantation, n (%)</td>
<td>0 (0)</td>
<td>3 (15)</td>
<td>0 (0)</td>
<td>3 (3.2)</td>
</tr>
<tr>
<td>RVOTO, n (%)</td>
<td>0 (0)</td>
<td>2 (5)</td>
<td>2 (2.2)</td>
<td></td>
</tr>
<tr>
<td>Endocarditis, n (%)</td>
<td>1 (2.9)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (1.1)</td>
</tr>
<tr>
<td>Total, n (%)</td>
<td>35 (37.6)</td>
<td>20 (21.5)</td>
<td>38 (40.9)</td>
<td>93 (100)</td>
</tr>
</tbody>
</table>

PAVSD: partial atrioventricular septal defect; IAVSD: intermediate atrioventricular septal defect; CAVSD: complete atrioventricular septal defect; LAVV: left atrioventricular valve; RAVV: right atrioventricular valve; LVOTO: left ventricle outflow tract obstruction; RVOTO: right ventricle outflow tract obstruction.
closure, annular dilatation was managed by commissuroplasty and/or annuloplasty, leaflet prolapse was corrected by means of chordal shortening or by placing artificial chordae. The repair was considered successful in case of less than moderate residual LAVV regurgitation or stenosis (mean gradient < 5 mmHg) on intraoperative echocardiography. LAVVR using a mechanical prosthesis was performed only if valve pathology (dysplastic LAVV or defect due to endocarditis) was not amenable to repair. Residual septal defects were closed directly. RAVV regurgitation was managed by valvuloplasty (RAVP, commissuroplasty and/or annuloplasty) or replacement (RAVR), if necessary. LVOTO was relieved by membrane resection, myectomy or Ross-Konno operation. RVOTO was managed by pulmonary valve replacement (PVR). Aortic regurgitation was addressed initially by valvuloplasty (AVP) and subsequently by replacement (AVR) with a mechanical prosthesis. PM was implanted transventrally by valvuloplasty (AVP) and subsequently by replacement valve replacement (PVR). Aortic regurgitation was addressed initially by obstructive muscle bundle resection and transannular patch. Pulmonary regurgitation was managed by pulmonary valve replacement (PVR). Aortic regurgitation was addressed initially by valvuloplasty (AVP) and subsequently by replacement (AVR) with a mechanical prosthesis. PM was implanted transvenously or epicardially based on patient's weight and age.

Follow-up
The median follow-up for the study cohort was 9.8 (range, 0–34) years.

End-points and definitions
Study end-points included in-hospital, late and overall mortality; freedom from reoperation; freedom from poor outcome; and clinical status at the last follow-up. In-hospital mortality was defined as death before hospital discharge or within 30 days of any operation. Late mortality was defined as all-cause death beyond 30 days of any operation. Poor outcome was defined as death or need for subsequent reoperation after the first reoperation. Residual structural and functional defects were evaluated by echocardiography. Clinical status was evaluated by New York Heart Association (NYHA) functional class.

Statistical analysis
All statistical analyses were performed using the SPSS statistical software for Windows, version 20 (SPSS, Inc., Chicago, IL, USA). The categorical variables are presented by number and percentage. Since for all continuous variables, the Shapiro-Wilk test showed absence of normality, the continuous variables are expressed by the median and range. Differences in patient characteristics between the three AVSD groups were tested for categorical variables using the Pearson’s χ² test or Fisher’s exact test, where appropriate. For continuous variables, the Kruskal-Wallis test was used. Estimates of overall survival, freedom from LAVV reintervention and freedom from poor outcome were obtained by means of the Kaplan-Meier method, with the difference among subgroups (AVSD type, LAVVP vs LAVVR) being tested by the log-rank test. Univariable and multivariable (stepwise method) binary logistic regression analysis were used to investigate the relationship between potential risk factors and in-hospital mortality. Univariable and multivariable (stepwise method) Cox regression analysis were performed to examine risk factors for overall mortality and LAVV reintervention after LAVV repair. The predictor variables considered were age at primary repair, primary repair before or after 1999, gender, AVSD type, Down syndrome, LAVV dysplasia, presence of DOLAVV, heterotaxy, coarctation, tetralogy of Fallot, age at first reoperation, first reoperation before or after 1999, time to first reoperation and LAVVP vs LAVVR at first reoperation (only for overall mortality after LAVV surgery at first reoperation). Probabilities used for entrance and removal in the stepwise method were 0.05 and 0.10, respectively. A P-value of <0.05 (two-sided) was considered statistically significant.

RESULTS

Mortality
In-hospital mortality occurred in 10 patients (14.5%). Seven patients died after LAVV surgery (4 repairs, 3 replacements), 2 patients died after LVOTO relief (Ross-Konno procedure) and 1 patient died after RAVVP. The causes of death were congestive heart failure in 4 patients, sepsis in 4 patients and gastrointestinal problems in 2 patients. There was no significant difference in in-hospital mortality among PAVSD, LAVSV and CAVSD patients (25.0 vs 6.7 vs 7.7%; P = 0.122, Pearson’s χ² test). Results of univariable and multivariable binary regression analysis indicated the presence of DOLAVV as the only (independent) risk factor for in-hospital mortality [odds ratio (OR) = 14.5; 95% confidence interval (CI) = 1.2–178.7; P = 0.037] (Table 3).

Late deaths occurred in 4 patients (5.8%). Three patients died late after LAVV surgery (2 replacements, 1 repair) due to congestive heart and respiratory failure. One patient died late after right ventricle outflow tract surgery probably due to arrhythmia.

Overall survival
Estimated overall survival at 1, 5 and 10 years was 87, 83 and 83%, respectively. The log-rank test showed no statistically significant difference in overall survival among AVSD types (P = 0.525). Univariable and multivariable Cox regression analysis identified the presence of DOLAVV as the only (independent) risk factor for overall mortality [hazard ratio (HR) = 6.8; 95% CI = 1.5–31.7; P = 0.015]. In the subgroup of patients undergoing LAVV surgery at the first reoperation (n = 61), the univariable Cox regression analysis identified the presence of DOLAVV (HR = 6.7; 95% CI = 1.4–31.6; P = 0.017) and LAVVR at the first reoperation (HR, 3.8; 95% CI = 1.2–11.9; P = 0.022) as risk factors for overall mortality. In the multivariable regression analysis, both factors did not remain statistically significant probably due to small patient numbers. Yet, they still appeared to be potential independent predictors given the magnitude of the estimated effect size and relatively large CIs in this small patient sample (HR, 4.6; 95% CI = 0.9–23.3; P = 0.06 and HR, 3.2; 95% CI = 1.0–10.5; P = 0.05, respectively). Despite the difference in overall survival between LAVVP and LAVVR groups (P = 0.014, log-rank test) (Fig. 1), no statistically significant difference was found in poor outcome (freedom from death or subsequent reoperation) between both groups (Fig. 2).

Reoperations
Sixty-nine patients underwent 113 reoperations. A second reoperation was needed in 27 patients (39.1%) (LAVVR = 14, LVOTO relief = 6, LAVVP = 4, PM = 3). Twelve patients required a third reoperation (PM = 3, LAVVR = 3, LVOTO relief = 2, RAVVR = 2, RAVVP = 1, PVR = 1). A fourth reoperation was performed in 4
One patient underwent a fifth reoperation (AVR). The vast majority of subsequent reoperations were performed because of residual LAVV pathology (52%), LVOTO (23%) and arrhythmias (14%).

Left atrioventricular valve pathology

Out of 47 subjects receiving LAVVP at the first reoperation, 4 patients underwent valve repair and 13 patients had valve replacement at the second reoperation (Fig. 3). Among 4 patients with valve re-repair at the second reoperation, 1 patient had a valve replacement at the third reoperation. Four of 13 patients with valve replaced at the second reoperation needed subsequent valve re-replacement. At the last follow-up, 27 of 41 survivors (65.9%) of LAVVP at the first reoperation were prosthesis-free. Using a Kaplan–Meier analysis, freedom from LAVV reintervention (re-repair or replacement) at 1 and 10 years after LAVVP at the first reoperation was 84 and 62%, respectively (Fig. 4). In univariable analysis of the predictors of durability of LAVVP at the first reoperation, normal karyotype proved to be a potential predictor given the estimated effect size and large CIs in this relatively small patient group (HR = 2.5; 95% CI = 0.9–6.7; P = 0.078). In the subgroup of patients undergoing LAVVR at the first reoperation (n = 14), 1 patient needed valve re-replacement because of somatic outgrowth.

### Table 3: In-hospital and late mortality

<table>
<thead>
<tr>
<th>No.</th>
<th>Anatomy</th>
<th>Redo 1 indication</th>
<th>Redo 1 procedure</th>
<th>Redo 2 indication</th>
<th>Redo 2 procedure</th>
<th>Cause of death</th>
<th>Time from last redo</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>PAVSD</td>
<td>RAVR</td>
<td>RAVP</td>
<td></td>
<td></td>
<td>Sepsis</td>
<td>n/a</td>
</tr>
<tr>
<td>2</td>
<td>IAVSD</td>
<td>LAVR, RAVR, VSD</td>
<td>LAVP, RAVP, VSD closure</td>
<td>LAVR, RAVR</td>
<td>LAVVP, RAVP</td>
<td>Sepsis</td>
<td>n/a</td>
</tr>
<tr>
<td>3</td>
<td>PAVID</td>
<td>LAVR</td>
<td>RAVP</td>
<td></td>
<td></td>
<td>Sepsis</td>
<td>n/a</td>
</tr>
<tr>
<td>4</td>
<td>CAVID</td>
<td>LAVV</td>
<td>RAVP</td>
<td></td>
<td></td>
<td>GI bleeding, PH</td>
<td>n/a</td>
</tr>
<tr>
<td>5</td>
<td>PAVSD, DOAVV</td>
<td>LAVVR, LVOTO</td>
<td>LAVRRe, myectomy</td>
<td>LVOTO</td>
<td>Ross-Konno</td>
<td>CHF</td>
<td>n/a</td>
</tr>
<tr>
<td>6</td>
<td>PAVSD, DOAVV</td>
<td>LAVVR, LVOTO</td>
<td>LAVRRe</td>
<td>LVOTO</td>
<td>Ross-Konno</td>
<td>CHF</td>
<td>n/a</td>
</tr>
<tr>
<td>7</td>
<td>PAVSD, DOAVV</td>
<td>LAVVR, endocarditis</td>
<td>LAVRRe, RAVP</td>
<td></td>
<td></td>
<td>Sepsis</td>
<td>n/a</td>
</tr>
<tr>
<td>8</td>
<td>CAVID</td>
<td>LAVR</td>
<td>LAVRe</td>
<td></td>
<td></td>
<td>CHF</td>
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</tr>
<tr>
<td>9</td>
<td>PAVSD</td>
<td>LAVR</td>
<td>LAVRe</td>
<td></td>
<td></td>
<td>CHF</td>
<td>n/a</td>
</tr>
</tbody>
</table>

PAVSD: partial atrioventricular septal defect; RAHR: right atrioventricular valve regurgitation; RAVP: right atrioventricular valvuloplasty; n/a: not applicable; IAVSD: intermediate atrioventricular septal defect; LAVR: left atrioventricular valve regurgitation; VSD: ventricular septal defect; LAVRRe: left atrioventricular valve replacement; LAVVP: left atrioventricular valvuloplasty; GI: gastrointestinal; PH: pulmonary hypertension; CAVID: complete atrioventricular septal defect; DOAVV: double orifice left atrioventricular valve; CHF: congestive heart failure; TOF: tetralogy of Fallot; RVOTO: right ventricle outflow tract obstruction; TAP: transannular patch; PR: pulmonary regurgitation; PVR: pulmonary valve replacement; RF: respiratory failure.
Left ventricle outflow tract obstruction

At the first reoperation, 4 patients underwent LVOTO relief. Three of them had initially membrane resection followed by redo membrane resection in 1 patient. One patient who had undergone septal myectomy required a Ross-Konno procedure later. At the second reoperation, 4 additional patients (2 after LAVVP and 2 after LAVVR at the first reoperation) had developed LVOTO and they underwent membrane resection (n = 3) or Ross-Konno procedure (n = 1). One patient after membrane resection needed redo membrane resection. One patient required membrane resection at the third and fourth reoperation after previous LAVVP followed by LAVVR. One patient needed a Ross-Konno procedure at the fourth reoperation after previous septal defect closure, LAVVR and PM implantation (Fig. 5).

Pacemaker

Three patients required PM implantation (1 for sick sinus syndrome and 2 for complete heart block) at the first reoperation. Three additional patients with postoperative complete heart block had PM implanted at the second reoperation (LAVVR = 2, LAVVP = 1). Three patients with postoperative complete heart block received PM at the third reoperation (LAVVR = 2, Ross-Konno = 1).

Functional status

Forty-six survivors (83.6%) were in NYHA Class I or II at the last follow-up. In the subgroup of 27 patients with preserved native LVAVR, echocardiography showed mild and moderate LVAV regurgitation in 20 and 7 patients, respectively. In the subgroup of patients undergoing LVOTO relief, all survivors had no or mild LVOTO on echocardiography. In the subgroup of patients with preserved RAVV, 48 of 53 (90.6%) patients had no more than mild-to-moderate RAVV regurgitation on echocardiography.

DISCUSSION

The outcomes of surgical treatment of AVSD have markedly improved over the last decades. In spite of substantially reduced
mortality, however, the need for reoperations and long-term morbidity remain an issue in this patient population [2, 6–10].

While most studies on reoperations after primary AVSD repair were focused either on a specific AVSD type or on a primary cause for reoperation (LAVV regurgitation or LVOTO), we analysed the entire cohort of patients undergoing reoperation from all causes after initial AVSD repair. We believe this can highlight the burden of the disease in terms of associated mortality and morbidity.

In-hospital mortality rate in the entire study cohort was 14.5%. This figure is similar to that reported by Pontailler et al. [2], yet higher than those reported by other authors [1, 6, 10]. Nevertheless, it reflects in-hospital deaths after any reoperation including high-risk operations, such as the Ross-Konno procedure. We found DOLAVV to be an independent risk factor for in-hospital and overall mortality. Two of 3 patients with DOLAVV had also had pseudoparachute LAVV, and they died after the Ross-Konno procedure in an attempt to address associated complex LVOTO. Poor outcome in AVSD patients with DOLAVV was also reported previously [2, 7]. Particular AVSD type did not appear to be a risk factor for mortality.

Estimated overall survival at 1, 5 and 10 years was 87, 83 and 83%, respectively. While some authors did not find any statistically significant difference in survival between replacement and repair subjects [1, 3], others including our own group [2, 7, 10] observed worse survival in LAVVR patients. This may reflect in-hospital deaths after any reoperation including high-risk operations, such as the Ross-Konno procedure. We found DOLAVV to be an independent risk factor for in-hospital and overall mortality. Two of 3 patients with DOLAVV had also had pseudoparachute LAVV, and they died after the Ross-Konno procedure in an attempt to address associated complex LVOTO. Poor outcome in AVSD patients with DOLAVV was also reported previously [2, 7]. Particular AVSD type did not appear to be a risk factor for mortality.

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Overall survival did not statistically differ among particular AVSD types.

In accordance with other reports from the literature [1, 2], reoperation rate was high with 39.1 and 17.3% patients requiring second and third redo procedure, respectively. This is substantially higher than after primary AVSD repair [4, 6, 9, 13], and it may suggest selection of subjects with worse anatomical variants compared with the initial AVSD cohort. The indications for subsequent reoperations included mainly LAVV regurgitation, LVOTO and cardiac arrhythmias.

Surgical treatment of recurrent LAVV regurgitation can be challenging in a paediatric population. It is obvious that LAVVP is superior to LAVVR because of preserved somatic growth and absence of haemorrhagic, thromboembolic and infectious complications. However, durability remains the Achilles heel of LAVVP particularly in patients with complex LAVV pathology, such as LAVV dysplasia, DOLAVV and pseudoparachute LAVV [8, 10, 13, 14]. More than three-fourth of our patients received LAVVP at the first reoperation for residual LAVV pathology. Nevertheless, a significant number of them had to undergo subsequent valve re-repair or replacement with 66% remaining prosthesis-free at the last follow-up. Similar figures were published by Malhotra et al. [1] and Pontailler et al. [2]. Freedom from LAVV reintervention at 1 and 10 years after LAVVP at the first reoperation was 84 and 62%, respectively. This is in agreement with previously published data [1, 10]. We found the absence of Down syndrome as a potential risk predictor for LAVV failure in this small patient sample. This has also been reported by other authors [2, 3]. For patients with complex LAVV pathologies, LAVVR may be the only alternative.

Despite any hard evidence, we firmly believe that reoperations for residual LAVV pathology can be prevented to some extent.
Symptomatic infants in particular those with AVSD should be operated on primarily at 1–3 months of age to prevent development of LAVV dysplasia and annular dilatation. Routine LAVV ‘cleft’ closure, and commissural and/or annular plication to reduce and stabilize the LAVV annulus should be performed whenever possible at primary AVSD repair. Perioperative transesophageal echocardiography should be used standardly to minimize the incidence of residual postoperative LAVV pathology. Finally, adequate postoperative surveillance and aggressive timing of reoperation might prevent progressive changes in LAVV structure and function, which renders the valve unrepairable. On the other hand, patients with severely dysplastic LAVVs will remain at higher risk for redo irrespective of the above measures.

LVOTO is the second most common indication for reoperation after primary AVSD repair [5, 15]. Myers et al. [16] suggest that LVOTO usually is complex, and more structures including LAVV are often involved. Furthermore, it has a tendency to progression and recurrence. Accordingly, 2 of our patients with DOLAVV developed complex LVOTO and required the Ross-Konno procedure. Fifty and 33% of the patients undergoing LVOTO repair at the first and second reoperation needed subsequent LVOTO relief, respectively. A small sample size precluded meaningful statistical analysis of risk factors.

The fate of patients undergoing reoperation after primary AVSD repair may suggest close relationship between LAVV pathology and LVOTO as some patients with initial LAVV problems needed LVOTO repair later on and vice versa. This is in agreement with previous reports in the literature [3, 16].

Six patients (8.7%) required PM implantation for complete heart block after LAVVR (66%), LAVVP (17%) and LVOTO relief (17%). This is in line with data reported by other authors [1, 10].

STUDY LIMITATIONS

This is a retrospective study including a small number of patients with all inherent limitations.

CONCLUSION

The current study showed that overall mortality and reoperation rate in patients undergoing reoperation was higher than after primary AVSD repair. DOLAVV was an independent risk factor for in-hospital mortality and overall survival. Residual LAVV pathology and LVOTO were the most common indications for subsequent reoperations. Particular AVSD type did not appear to be a risk factor for mortality or LAVVP failure. There is some evidence for the close relationship between LAVV pathology and LVOTO in subjects undergoing reoperation after primary AVSD repair as some patients with initial LAVV problems needed LVOTO repair later on and vice versa.

Conflict of interest: none declared.

REFERENCES


APPENDIX. CONFERENCE DISCUSSION

Dr O. Ghez (London, UK): This is a very important analysis of the long-term results of AVSD repair and you showed very clearly that reoperations are frequent in about one-sixth of the patients operated, and that the mortality and morbidity are very significant.

Do you think that obtaining an absolutely perfect result in these patients at the first operation might decrease the risk for reoperation? And in your experience has the use of intraoperative echo made a difference?

Dr Sojak: Well, the presented data reflect a 37-year study period. So of course, our surgical policy and technique have evolved over the study period. Now, we tend to repair the patients at the time of primary repair, much earlier. Thus, we think that the ideal time for operation is between the first and third month of age.

Secondly, we have found that leaving the cleft open during the primary repair is a risk factor for redo. So now we routinely close the cleft unless the patient has a complex left AV valve pathology.

In addition, we routinely use echo and we’ve been using it since 1998. This measure coincided very well with the improved outcomes in our patient group. We found that the surgical era before 1998 was a risk factor for death or reoperation. Therefore, we think it should be a routine part of the management. We don’t actually mind going back on the heart-lung machine, and try to repair the valve better if we are not satisfied with the original result. Ideally...
we’d be happy to have a residual left AV valve regurgitation that is less than moderate and we would accept a gradient across the left AV valve of less than 5 mm Hg.

Dr Ghez: Regarding the reoperations, are you very aggressive in terms of timing of reoperation? What are your criteria?

Dr Sojak: Certainly, that’s a good point. Because our previous study and those of other centres have clearly shown that the longer you wait for reoperation, the more you are running the risk of the patient developing left AV valve dysplasia or annular dilatation which are much more difficult to manage. In our previous study we noted that many patients with left AV valve dysplasia had to have their valve replaced, which is a significant risk factor of death. Of course, with newer techniques of patch augmentation or annuloplasty, the left AV valve replacement could have been probably avoided. But again, if we have an asymptomatic patient, a patient who is having more than grade 2 left AV valve regurgitation we would not hesitate too long and go right away with the reoperation.

Dr Ghez: A significant number of these patients are lost to follow-up in your manuscript. This is a common problem in congenital heart disease, obviously. What measures do you have in place in your centre to follow these patients in the long term and in particular for the transition to adult congenital care?

Dr Sojak: We have acknowledged the problem of having a significant proportion of patients lost to follow-up. Therefore, in close collaboration with our GUCH cardiologists and paediatric cardiologists, we have established at least in our referring area a network of outpatient clinics where paediatric cardiologists closely cooperate with GUCH cardiologists in order not to lose too many patients. In this way we can assure that the patients will stay under the control.

In terms of patients who were operated much earlier, we started a massive media campaign in order to recruit them and get them under cardiologist control.

Dr S. Cicek (Istanbul, Turkey): This is one of the largest and longest follow-up series with complete AV canal defect. Do you have any data regarding the surgical technique at primary operation? Is it a standard technique or has it evolved over the years? If so, have you observed any changes regarding the reoperation rate?

Dr Sojak: Actually, the surgical technique has been pretty stable in the last 20–25 years. The only difference was the cleft was left initially open in patients. In the last 20 years our common policy was to close the cleft whenever possible.

Dr Cicek: Your primary technique was single patch repair, or double patch?

Dr Sojak: Well, the single patch technique was used in the very first 3 or 4 years of the study period. The remaining patients operated on after 1979 received a two-patch technique.

Dr M. Danton (Glasgow, UK): In managing left ventricular outflow tract obstruction, 2 patients underwent Ross-Konno procedure and, if I am correct, both patients died after surgery?

Dr Sojak: Well, in the total patient cohort there were 3 patients receiving a Ross-Konno operation. Unfortunately, two of them with partial AVSD and double orifice left AV valve, developed complex LVOTO later and died after Ross-Konno procedure.

Dr Danton: The Ross-Konno operation is normally quite a good option for multilevel LVOT obstruction. Is it not a good option in this particular anatomy, AVSD, or how do you explain that?

Dr Sojak: Well, these patients had a small and dysplastic aortic valve not amenable to repair. Otherwise, they would just receive a myectomy or a modified Konno procedure.

Dr Danton: Yes, but did they have a particularly poor outcome compared to patients undergoing a Ross-Konno for other conditions? You have a high mortality of Ross-Konno in this group. Is that attributed to the anatomy of AVSD?

Dr Sojak: Not other than mentioned before.