Long-term outcomes of reoperations following repair of partial atrioventricular septal defect

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Abstract

OBJECTIVES: Partial atrioventricular septal defect (pAVSD) is repaired with excellent long-term survival. However, up to 25% of patients require reoperations. This study reviews results of reoperation following pAVSD repair at a single institution.

METHODS: From 1975 to 2012, 40 patients (16%, 40/246) underwent reoperation following pAVSD repair at the study institution. The data were retrospectively reviewed.

RESULTS: The mean time to reoperation was 5.4 ± 5.8 years. The most common reoperations were left atrioventricular valve (LAVV) surgery (78%, 31/40) and resection of left ventricular outflow tract obstruction (20%, 8/40). The most common cause for LAVV surgery was regurgitation through the cleft (58%, 18/31), followed by central regurgitation (29%, 9/31). Most cases of LAVV regurgitation were treated by repair (77%, 24/31), rather than replacement (23%, 7/31). Since the introduction of a patch augmentation technique for LAVV repair in 1998, the rate of repair has increased from 54 to 94% ($P = 0.012$). The early mortality rate was 2.5% (1/40). The survival rate was 90% (95% CI: 76–96) at 10 years and 83% (95% CI: 60–94) at 20 years. The rate of freedom from further reoperation was 66% (95% CI: 46–80) at 10- and 20-year follow-up.

CONCLUSIONS: The most common cause for reoperation following pAVSD repair was LAVV regurgitation through the LAVV cleft. Reoperation is performed with survival comparable to that of primary pAVSD repair, yet the rate of further reoperations remains high. The patch augmentation technique for LAVVR has significantly increased the rate of successful LAVV repair.

Keywords: Congenital heart disease • Septal defects • Outcomes

INTRODUCTION

The spectrum of atrioventricular septal defect affects 3–5 per 10,000 live births, with partial atrioventricular septal defects (pAVSDs) constituting ~25% of these [1–4]. Current practice is to surgically correct pAVSD in the preschool years or earlier if heart failure develops. While early mortality is low and long-term survival excellent, up to 25% of these patients require reoperation at 30 years follow-up [5–13]. The most common causes of reoperation are left atrioventricular valve regurgitation (LAVR) and left ventricular outflow tract obstruction (LVOTO) [4, 14]. We performed a retrospective review of all reoperations performed following repair of pAVSD at a single institution.

PATIENTS AND METHODS

Patients

Between January 1975 and January 2012, 249 consecutive patients underwent surgical correction of pAVSD at the Royal Children’s Hospital (RCH), Melbourne. Of these, 40 underwent a cardiac reoperation and were included in the present study. The study was approved by the RCH Human Research Ethics Committee.

Data were obtained by retrospective review of patient records and follow-up was obtained by correspondence with the patient’s cardiologist or general practitioner. Patients were contacted by telephone and asked to complete a questionnaire if no current...
medical practitioner could be identified. For patients who were lost to follow-up, the Victorian Registry of Births, Deaths and Marriages was searched for death records.

Operative approach

Our approach to repair of pAVSD has previously been described in detail [13]. Decision to close the LAVV cleft is made intraoperatively based on a combination of direct inspection and echocardiographic assessment. In cases of trivial or less LAVVR, practice has changed over the study period; currently, it is routinely closed unless contraindicated.

Reoperations were performed via a median sternotomy on cardiopulmonary bypass with mild hypothermia. A single case was performed under hypothermic circulatory arrest.

Our institutional approach to LAVV regurgitation following repair of partial and complete AVSD has been previously described in detail [15, 16]. Patients are referred for reoperation when they have moderate or greater LAVVR; however, consideration is given to the age of the patient and the feasibility of repair on echocardiographic evaluation. In all cases, LAVV repair is preferred to LAVV replacement. Prior to 1998, repair was undertaken in a conventional manner including closure of the cleft between the superior and inferior bridging leaflets and annuloplasty, depending on the cause of the regurgitation as seen on echocardiography and intraoperatively. Following 1998, a patch augmentation technique was applied to those patients with normal papillary muscles. In this technique, the thickened edges of the superior and inferior bridging leaflets adjacent to the cleft are resected, leaving only pliable tissue. Subsequently, a thin strip of autologous pericardium treated with 0.625% glutaraldehyde is inserted into the defect, patching the area of the cleft and, thus, augmenting the leaflet. The free edge of the patch is then suspended either with Goretex neochordae or chordal sutures and annuloplasty, depending on the cause of the regurgitation as seen on echocardiography and intraoperatively.

For relief of LVOTO, all patients underwent myomectomy via an aortotomy. In cases where accessory LAVV tissue was seen to contribute, this was resected. In one of these cases, a left atriotomy was also performed to allow adequate resection of the accessory subvalvular tissue.

Statistical analysis

Data were analysed with STATA version 12 (Stata Corp, College Station, TX, USA). Unless stated to the contrary, continuous data were summarized as mean ± standard deviation. The time-dependent endpoints investigated were: mortality, second cardiac reoperation, second reoperation for LAVV regurgitation and second reoperation for LVOTO. For all endpoints, time was measured starting from the time of first reoperation following pAVSD repair. Kaplan–Meier analysis was used to estimate survival and freedom from reoperation. Statistical analysis of the impact of the patch augmentation on outcomes was performed as follows: the Fisher’s exact test was used to compare proportions of valves repaired prior to and after the introduction of the technique; conversely, a Cox proportional hazards test was used to compare freedom from reoperation between the patch augmentation technique and the traditional repair. The threshold for statistical significance was taken as P < 0.05.

RESULTS

Operative details

A total of 40 patients underwent reoperation, with demographic data presented in Table 1. The most common procedures were LAVV repair or replacement (31/40, 77.5%) and relief of LVOTO (10/40, 25%). Four patients underwent more than one concomitant procedure (4/40, 10%).

Details on LAVV surgery are presented in Table 2. The most common cause for reoperation on the LAVV was regurgitation through the cleft (18/31, 58%), followed by central regurgitation.
(9/31, 29%). Of the 18 cases of regurgitation through the cleft, 7 were due to rupture of the cleft closure, while the remaining 11 cases were due to a residual cleft (not closed, n = 6 and incomplete closure, n = 5). Of the cases of central regurgitation, 5 were due to annular dilatation, while the remaining 4 were due to dysplastic or deficient leaflets. In the majority of cases, the LAVV was repaired (24/31, 77.4%). In the remaining patients, the LAVV was replaced (7/31, 22.6%). Of the repairs, the patch augmentation technique was used in addition to this technique (2/13, 15%). Prior to 1998, 7 of 13 patients underwent repair (7/13, 53.8%) compared with 17 of 18 after the introduction of the technique (17/18, 94.4%), representing a significant increase in the proportion of valves successfully repaired (P = 0.012). The patch augmentation repair was introduced in 1998 [15].

Details on patients undergoing surgery for LVOTO are presented in Table 3. In all patients, a subaortic membrane was present (10/10, 100%), in 3 of these patients there was also accessory LAVV tissue (3/10, 30%). These cases were managed either with myomectomy in isolation or myomectomy with excision of redundant LAVV tissue. In 1 patient, the superior bridging leaflet was approached from the left atrium and detached to facilitate resection of fibrous tissue with subsequent reattachment.

The remaining 3 patients had residual atrial septal defect (n = 2) and residual VSD (n = 1) repairs.

**Survival**

There was a single death in the early postoperative period, giving an early mortality rate of 2.5% (1/40). This patient was a 3-week old boy, who presented in 1993 with coarctation of the aorta and pAVSD, with a severely dysplastic LAVV, but balanced ventricles and normal biventricular configuration. He initially underwent subclavian flap repair of the coarctation, but could not be weaned off the ventilator due to heart failure, and so was brought forward for pAVSD repair. Following pAVSD repair, multiple attempts to wean off inotropes resulted in severe pulmonary congestion. LAVV replacement with a 16-mm Carbomedics valve was performed, but the patient could not be weaned off bypass despite maximal inotropic support. A second attempt to replace the valve with a 10-mm pulmonary homograft was performed, despite an initial improvement in haemodynamics, bypass could not be weaned, and with rising pulmonary pressures despite maximal inotropic therapy, treatment was withdrawn.

Long-term survival is demonstrated in Fig. 1. The survival rate at 10 years was 90.2% (CI: 75.9–96.2%), while at 20 years it was 83.2% (CI: 60.2–93.6%). Of interest, all of the deaths occurred in patients whose first reoperation was on the LAVV.

**Reoperation**

Long-term freedom from a second reoperation is shown in Fig. 2. The rate of freedom from reoperation was 66% (CI: 46–80%) at both 10 and 20 years. Eleven patients underwent a further reoperation (11/40, 28%), while 5 patients underwent a third reoperation (5/40, 13%). Details of these procedures are provided in Table 4. There were 4 reoperations on the LAVV; 3 of these were in patients who had previous repairs and required late reoperation for severe LAVVR. At reoperation, the mechanisms of regurgitation were: annular dilatation (n = 1) deficient lateral leaflet in a patient with a single papillary muscle (n = 1) and LAVV endocarditis associated with rupture of a transposed chorda (n = 1). Of these 3 patients, 1 had undergone the patch augmentation technique for the initial LAVV repair.

There were no patients whose initial reoperation was for LVOTO who subsequently required relief of LVOTO and vice versa. There were no patients who underwent a fourth reoperation.

**DISCUSSION**

It has been well documented that complete repair of pAVSD is associated with a low early mortality rate in the range of 1.2–5% in recent studies [5–13]. However, there is a relatively high requirement for reoperation, up to 25% at 30 years of follow-up [5, 7, 13]. This requirement for reoperation represents a target for potential reduction in morbidity and improvement in the quality of life in patients with pAVSD, and prompted the current review. Previously, there has only been a single review evaluating outcomes following reoperation for pAVSD reported in the literature [14], while several groups have reviewed outcomes of combined cohorts with partial, transitional and complete AVSD undergoing reoperation [15–22].

The indication for reoperation in our cohort was predominantly LAVV regurgitation or LVOT obstruction, which is consistent with previous reports [5, 7, 14]. At our institution, we have previously identified risk factors for reoperation on the LAVV to be preoperative congestive heart failure and moderate or greater LAVVR on discharge echocardiogram [13]. In examining LAVV regurgitation, the majority of cases were due to regurgitation through the cleft, and it is interesting to note that almost half of these cases were due to rupture of an initial cleft repair, with the remaining cases being due to a residual cleft. This finding emphasizes the importance of secure cleft closure at initial correction of a pAVSD.

The majority of patients with LAVVR in our study underwent repair (77.4%). This is a higher rate of repair when compared with Stulak et al. [14] who described a LAVV repair rate of 52% in their cohort of 92 patients. However, our results are consistent with other groups who have analysed LAVV reoperation on partial and complete AVSD patients and recommended repair over replacement [18–22].

A recent paper by our group described the patch augmentation technique in patients undergoing reoperation for LAVVR following either complete or partial AVSD [15]. It demonstrated that there was improved freedom from reoperation in the patch augmentation group. In the present study, there was no statistically significant difference in freedom from reoperation between the groups, likely due to the small number of patients. However, the rate of

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**Table 3: LVOTO reoperation details**

<table>
<thead>
<tr>
<th>Aetiology of LVOTO</th>
<th>n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fibromuscular subaortic membrane</td>
<td>10 (100.0)</td>
</tr>
<tr>
<td>Accessory LAVV tissue</td>
<td>3 (30.0)</td>
</tr>
<tr>
<td>LVOTO techniques</td>
<td></td>
</tr>
<tr>
<td>Myomectomy</td>
<td>10 (100.0)</td>
</tr>
<tr>
<td>Resection accessory LAVV tissue</td>
<td>3 (30.0)</td>
</tr>
</tbody>
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LVOTO: left ventricular outflow tract obstruction; LAVV: left atrioventricular valve.
A successful repair has significantly increased, since the introduction of this technique in 1998, from 54 to 94%. This may reflect the fact that this technique increases the range of valve pathologies that can be repaired.

LVOTO requiring surgery occurs in up to 10% of patients at 10 years and is multifactorial, related to intrinsic narrowing and elongation of the LVOT, malalignment of the aorta and anomalous insertion of LAVV chordae [23]. In our series, the indication for reoperation for LVOTO was mostly a fibromuscular subaortic membrane, with additional contribution of a redundant LAVV tension apparatus in approximately one-third of patients.

We have demonstrated an early postoperative mortality rate of 2.5%, which is consistent with the early mortality reported for initial pAVSD repair, and less than a previously reported result for reoperation following pAVSD repair of 5.2% [14]. Long-term survival at 20 years of follow-up was 83%, similar to the long-term survival following initial repair of pAVSD [5–7]. All deaths occurred in patients who underwent LAVV surgery; however, this may simply reflect the fact that it is by far the most common procedure.

The requirement for further reoperation in our cohort was relatively high with a 10-year freedom rate from a second reoperation of 66%. Interestingly, all reoperations occurred in the first 10 years and the rate remained unchanged at 20 years. This is a higher proportion of reoperations than seen in the group reviewed by Stulak et al. [14], who reported 10-year freedom rate from reoperation of 83%. It is, however, in line with other groups who had a similar rate of LAVV repair to our cohort [18–22]. The pattern of further reoperation is similar to that of the initial reoperation, dominated by procedures on the LAVV and LVOT. Interestingly, patients retained their pattern of pathology, with no patient who had LAVV reoperation subsequently requiring relief of LVOTO, and vice versa.

The time to reoperation in our cohort was 5.8 years, shorter than described by Stulak et al. [14]. This may represent a more aggressive strategy of reintervention on the LAVV. However, it is similar to that reported by Sojak et al. [24], who argued that early repair of the LAVV may prevent dysplastic changes which render it unrepairable. The fact that they achieved a similar rate of repair to our group would seem to support this contention.

CONCLUSION

The most common cause for reoperation following pAVSD repair was LAVV regurgitation through the LAVV cleft. Reoperation is performed with survival comparable to that of primary pAVSD repair.
yet the rate of further reoperations remains high. The patch augmentation technique for LAVVR has significantly increased the rate of successful LAVV repair.

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**REFERENCES**


