Closure of the zone of apposition at correction of complete atrioventricular septal defect improves outcome

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Abstract

Objective: Outcome after correction of atrioventricular septal defect depends to a great deal on the postoperative function of the left atrioventricular valve. The related role of the zone of apposition (‘cleft’) has been debated: should it be closed (bileaflet repair) or should it be left untouched (trileaflet repair)? This study aims to answer the question by comparing the outcome of patients treated according to these two approaches. Methods: We reviewed all our patients who underwent repair of complete atrioventricular septal defect from 1984 to 1997 and selected those in whom the closure of the zone of apposition in principle would have been possible. Two groups with similar characteristics were constituted: group I (n = 63), where the zone of apposition was deliberately not closed as part of a trileaflet repair (postoperative open zone of apposition) and group II (n = 96), where it was electively closed as part of a bileaflet AV valve repair (closed zone of apposition). Since we changed from a trileaflet to a bileaflet repair in 1987, the two groups differ in terms of size and length of follow-up. Outcome was compared with regard to survival and freedom from reoperation for left atrioventricular valve incompetence. Late atrioventricular valve function was evaluated by Echo-Doppler. For statistical analysis, we used Chi-square or Fisher’s exact test, the Mann–Whitney test and the log-rank test for comparison of Kaplan–Meier curves. The difference was considered statistically significant with a \( P \)-value of 0.05 or less.

Results: Early mortality was 9.5\% (6/63) in group I and 3.1\% (3/96) in group II (\( P = 0.16 \)). Actuarial survival after 1, 4 and 8 years was 80.4, 68.4 and 64.8\%, respectively, for group I. Actuarial survival for group II was 94.7, 92.1 and 92.1\% (\( P = 0.0002 \)). Freedom from reoperation for left atrioventricular valve regurgitation was 90.2, 85.6 and 77.8\% for group I at the same time interval. It was a constant 97.9\% for group II (\( P = 0.0016 \)). At reoperation, left atrioventricular valve regurgitation was present through the open zone of apposition in 63\% of group I cases. The follow-up is 96\% (126/131) complete. An increase in degree of left atrioventricular valve incompetence was noted in 28\% (11/39) of group I cases and in 9\% (8/87) of group II cases (\( P = 0.0131 \)).

Conclusion: This study demonstrates the advantage of closing the zone of apposition (‘cleft’) as part of repair of complete atrioventricular septal defect. Survival, freedom from reoperation for left atrioventricular valve incompetence and over-all outcome were more favourable in patients of group II. The zone of apposition should be surgically addressed whenever the morphology of the left atrioventricular valve allows for closure without producing stenosis. © 2000 Elsevier Science B.V. All rights reserved.

Keywords: Atrioventricular septal defect; Surgery zone of apposition; Outcome; Cleft; Bileaflet procedure; Left AV valve incompetence

1. Introduction

Surgical management of patients with complete atrioventricular septal defect (AVSD) has advanced over the last 20 years from a two-staged approach with pulmonary artery banding and high-risk definitive repair, to successful complete primary correction in early infancy. Improved accuracy of preoperative diagnosis, better understanding of the surgical anatomy, progress of surgical technique, myocardial protection, and individualised postoperative care, including management of pulmonary hypertensive events, have contributed to increased survival and reduced rates of reoperation. The crucial task for the surgeon is to reconstruct the left atrioventricular (AV) valve without creating valve incompetence or stenosis. Regurgitation of the left AV valve remains the most important risk of postoperative morbidity and mortality.

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There is still no consensus concerning the nature of the area between the left superior and inferior bridging leaflet of the common AV valve. It is classically described as a ‘cleft’: a source of regurgitation, which has to be closed, thus resulting in a two leaflet ‘mitral’ valve. However, Rastelli [1] showed that suturing of a ‘cleft’ never produced a normal mitral valve. In 1979, Piccoli [2] and in 1982, Studer [3] described the ‘cleft’ as a true commissure and the left AV valve as a three-leaflet valve. On this basis Carpentier [4] promoted the concept of not closing the ‘cleft’, rendering the left AV valve essentially a three-leaflet valve. Recently, R.H. Anderson presented his concept of the common AV valve, in which the space between the facing surfaces of the left ventricular components of the bridging leaflets of the common AV valve is labelled zone of apposition (ZoA). Anderson enumerates important morphologic features that distinguish the zone of apposition from an isolated mitral cleft [5]. When the surgeon closes a cleft, he restores the anatomy of a normal anterior leaflet of the mitral valve. When he closes the zone of apposition, he does not produce a normal anterior leaflet of a mitral valve. Since the postoperative left AV valve in AVSD anomalies can never be a mitral valve, it would be misleading to use the term ‘cleft’ for the space between the facing surfaces of the left ventricular components of the bridging leaflets of the common AV valve. This space, on the other hand, is not a commissure either. A commissure of the mitral valve is defined as a breach in the leaflet skirt supported by a fan-shaped chord atop its prominent papillary muscle. It is a functional division between segments of leaflet tissue guarding either an atrioventricular or ventriculo-arterial junction. The area between the two bridging leaflets fulfills none of these criteria. Therefore, we have adopted the terminology of zone of apposition to stress the point that a cleft, a commissure and the zone of apposition are three separate, clearly discernible entities.

Over time, more and more paediatric cardiac surgeons have adopted the concept of closing the zone of apposition. It now seems to prevail over the opinion that the commissure should be left untouched. Since 1981, both techniques have been used in our Department in a large number of cases by one surgeon. After studying our results with the trileaflet procedure, we changed our standard protocol for AVSD [6]. Since 1987, we progressively suture-closed the zone of apposition in all patients with balanced or left dominant AVSD. Today it seems timely to compare the outcome of patients treated according to the two different protocols in terms of survival, rate of reoperation and late function of the left AV valve.

2. Patients and methods

This study covers the period from January 1984 to January 1997. Clinical charts, cardiac catheterization data, echocardiographic reports and operation notes of all children operated on for complete atrioventricular septal defect were reviewed. Patients with an interventricular communication too small to be closed with a patch and those with an intermediate form of AVSD according to the definition by Wakai and Edwards [7] were excluded from the study as were all cases with parachute (n = 9) and double orifice (n = 13) left AV valve.

One hundred and fifty nine patients fulfilled the inclusion criteria. Median age at operation was 11 months (range 1–161 months), median weight 6.3 kg (range 2.6–42.7 kg). They fell into two groups. In group I patients (n = 63), the ZoA was left open. In group II patients (n = 96) it was closed either completely (n = 87) or partially (n = 9). Preoperative and perioperative data were analysed using Chi-square test or Fisher’s exact test for univariate analysis, and Mann–Whitney test to compare several variables (GraphPad Prism™ software). The difference was considered statistically significant with a P-value of 0.05 or less.

Table 1 summarises our findings. The two groups were comparable with regard to age and weight at operation, male and female ratio, association of Down’s syndrome, preoperative and perioperative surgical procedures, major associated cardiac lesions, incidence of pulmonary hypertension, distribution of Rastelli AVSD types, number of ventricular imbalance cases and duration of cardio-pulmonary bypass. For diagnosis of pulmonary hypertension, pulmonary pressure had to be greater than 50% of systemic mean blood pressure either measured during catheterization or estimated by echocardiography. The two groups however differ in terms of size (P < 0.0001), since we changed in 1987 from a trileaflet to a standard bileaflet repair. Moreover more patients in group II were operated on at the age of 7–12 months (P = 0.04). Prior to operation, there were more cases of moderate and severe incompetence of the left AV valve in group II (P = 0.004). The incompetence of the left AV valve was graded according to the echocardiographic findings when available, otherwise in conformity with the results of heart catheterization.

In all our patients, cardiac surgery was performed by the same surgeon during the period under review and varied little with respect to perioperative management. Cardiopulmonary solution (St. Thomas’ Hospital) was used in all patients. A two-patch technique was always employed to close the ventricular and atrial defects. In group II, the ZoA was closed simply by interrupted monofilament sutures. The zone of apposition usually was closed completely except in patients with small left AV valves where it was closed partially. Hegar dilator with the size of an age matched mitral valve diameter was used to measure the valve opening and to prevent postprocedural stenosis [6,8]. Since 1993, ultrafiltration using a modification of the technique of Naik et al. [9] was applied in all infants. Postoperative care was similar in both groups, especially with respect to management of pulmonary hypertensive episodes.

The follow-up interval for patients in group I was 12–198 months (median 107), in group II it was 10–126 months.
The current evaluation for the survivors took place in January 1998. Five patients (three in group I, two in group II) from abroad were lost to follow-up.

Follow-up evaluation, consisting of clinical examination and echocardiography, took place either in our Department (n=70) or in the referring centres (n=56) on the basis of an accepted guideline. Echocardiographic examinations focussed on the function of the left AV valve. The degree of regurgitation was estimated by measuring the maximum width of the jet at the base and the size of the valve annulus from two orthogonal planes. If the width was less than 1/3 of the valve annulus, it was considered as mild, if less than 2/3, as moderate, and if 2/3 or more, as severe. Kaplan–Meier curves for actuarial survival and freedom from reoperation were calculated using the GraphPad Prism™ software. The log-rank test was used to assess the statistical differences between the two groups, with determination of P-value. Where appropriate, means are given with ±standard deviation and percentages with 95% confidence limits (CL) in brackets.

3. Results

3.1. Survival

Nine of the 159 patients died within 30 days of operation with an early mortality of 5.7% (95%-CL: 2.6–10.4) in the whole series, six in group I (9.5%) and three in group II (3.1%). Early mortality did not statistically differ whether
the zone of apposition was closed or not (P = 0.15). Table 2 details the main causes of death.

Nineteen patients of the 150 early survivors died late, considerably more in group I with the ZoA open (15/57) than in group II (4/93) where it was closed. The difference in the actuarial survival between both groups is statistically significant (P = 0.0002). In children with closed ZoA, the risk of late death stabilises quickly and approaches 0 after 16 months, with survival at 92.1% (95%-CL: 86.3–97.9). When the ZoA is left open, survival rate continues to decrease over several years with an actuarial survival of 64.8% (95%-CL: 52.4–77.2) from the 79th postoperative month on (Fig. 1). Pulmonary vascular disease constitutes the principal cause of late death (Table 2). Remarkably, all 42 patients with major associated cardiac anomalies have survived and are still alive.

3.2. Reoperation

Thirteen out of the 16 patients who had to be reoperated (81%, 95%-CL: 54–96) underwent reintervention for severe regurgitation of the left AV valve (Table 3). Fig. 2 gives the time related freedom from re-repair for this indication. The risk of re-repair is significantly higher in patients where the ZoA of the left AV valve was left open (P = 0.0016). When the ZoA is not closed, it takes 7 years for this hazard to stabilise at 22.2%. In patients with surgical closure of the ZoA, the risk is constant at 2.1% after the first postoperative month.

At reoperation, the regurgitation was found to be predominantly through the open zone of apposition in seven of the 11 cases of group I (63%). In three other cases it was due to suture-dehiscence of the anterior or posterior leaflet from the patch. In the remaining patient, the cause was a prolapse of the lateral leaflet. On seven occasions, the valve could be repaired. The correction essentially consisted of closure of the ZoA. Annuloplasty and refixation of the patch were performed when indicated. In four children the valve had to be replaced by a mechanical prosthesis. Four reoperated patients in group I died, three of them after valve replacement, one in the early postoperative period. In one case, valve replacement was complicated by complete heart block that necessitated pacemaker implantation. One re-intervention was performed in another hospital.

In group II, reintervention for severe left AV valve incompetence was necessary in two cases early after the initial repair on the 15th and 16th postoperative day, respectively. The suture approximating the ZoA was broken in one patient with several valve chordae ruptured. In the other patient, the anterior leaflet had dehisced from the patch and infarction of the left ventricle was suspected. Both patients died, the first late after valve repair, the second early after valve replacement.

3.3. Late left AV valve function

In 98 of the 126 surviving patients (78%, 95%-CL: 70–85), including reoperation survivors, the left AV valve was
The purpose of this study was to compare the outcome of two different surgical approaches for repair of the complete form of AVSD: either to leave the zone of apposition (
\textit{‘left’) untouched, or to close it as part of AVSD repair. We categorised our patients with repair of complete AVSD according to these two approaches into group I and group II. The two groups were similar for the usual factors reported to influence early survival [6,10] such as young age, major associated cardiac anomalies, Down’s syndrome and Rastelli AVSD types. Patients with parachute and double (accessory) orifice left AV valve were excluded, since the reconstruction of the left AV valve is different under these circumstances: the zone of apposition has often to be left untouched to avoid creating a postoperative stenotic valve [3,11,12]. Moreover, their outcome is said to be less favourable than in patients with the ‘usual’ valve morphology of the left portion of the common AV valve [3,11,13,14].

Our study includes two different time frames. Until October 1987, we employed the trileaflet technique, leaving the ZoA untouched. We thereafter moved to the bileaflet repair, closing surgically the ZoA. During this transitional period of 14 months, some patients with no preoperative AV valve regurgitation still had the trileaflet repair. After 1988, all patients—whether they had or had not AV valve incompetence—had bileaflet correction with closure of the ZoA. This progressive change explains the 11 ‘overlapping’ patients in group I and the 2 in group II (see Table 1: year of operation). We chose to include all those 13 ‘transitional’ patients in the study, as they met selection criteria; their exclusion would have reduced the size of group I in particular, thereby decreasing the number of events and increasing the chances of statistical errors. The inclusion of our ‘transitional’ patients has—on the other hand—not statistically altered outcome. When comparing the outcome of the 11 ‘overlapping’ cases of group I with the remainders of their group in a statistical model, early (P = 0.58) and late (P = 0.46) mortality are not different and rate of reoperation for left AV valve incompetence is the same (P = 1). As a consequence of the different time frames, the follow-up for patients with an open ZoA is longer. This difference was statistically neutralised by applying the log-rank test for comparison of actuarial survival rates and freedom from reoperation.

Preoperative left AV valve incompetence was found more frequently in group II. This fact may be attributed to a bias in methodology since there were fewer preoperative echocardiographic data in group I (15) than in group II (65). The more sensitive echocardiographic techniques that were

### Table 2
Main causes of death after repair of complete AVSD without (group I) and with closure (group II) of the zone of apposition

<table>
<thead>
<tr>
<th>Cause</th>
<th>Group I (n₁ = 63)</th>
<th>Group II (n₂ = 96)</th>
<th>P-value (log-rank-test)</th>
</tr>
</thead>
<tbody>
<tr>
<td>30 Day mortality</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
<td>4</td>
<td>0</td>
<td>0.15</td>
</tr>
<tr>
<td>Cardiogenic shock</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Ventricular tachycardia</td>
<td>1</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Sepsis</td>
<td>0</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Brain death</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Death after reoperationa</td>
<td>1/6</td>
<td>1/3</td>
<td></td>
</tr>
</tbody>
</table>

#### Table 3
Indication for reoperation after repair of complete atrioventricular septal defect

<table>
<thead>
<tr>
<th>Indication</th>
<th>All patients (n = 159)</th>
<th>Group I (n₁ = 63)</th>
<th>Group II (n₂ = 96)</th>
<th>P-value (log-rank-test)</th>
</tr>
</thead>
<tbody>
<tr>
<td>No.</td>
<td>% of n</td>
<td>No.</td>
<td>% of n₁</td>
<td>No.</td>
</tr>
<tr>
<td>Left AV valve regurgitation</td>
<td>13 8.2</td>
<td>11 17.5</td>
<td>2 2.1</td>
<td></td>
</tr>
<tr>
<td>Ventricular patch dehiscence</td>
<td>1 0.6</td>
<td>1 1.6</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Atrial patch dehiscence</td>
<td>2 1.2</td>
<td>1 1.6</td>
<td>1 1.0</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>16 10.0</td>
<td>13 20.6</td>
<td>3 3.1</td>
<td></td>
</tr>
</tbody>
</table>
available only after we had changed our standard protocol are likely to have increased the detection of left AV valve regurgitation. But since outcome in group II was not worse than in group I but, in fact, better, it seems safe to assume that this potential bias did not influence our results. Several authors [14–16] failed to detect any relationship between the degree of preoperative and postoperative atrioventricular regurgitation.

Earlier or later date of operation seems to have had little influence on the results in this series. Although one would expect that increasing surgical experience over time and continual improvement of postoperative management favours group II patients, the 30-day mortality is not statistically different between the two groups. This absence of statistical difference might be due to the small number of events in relation to the size of the two groups.

If patients with double orifice or parachute left AV valve are taken into account, early mortality is significantly higher in group I [17]. This is due to the fact that in these patients an individualised approach is necessary where the zone of apposition cannot be closed completely in most of the cases because suture-closure would lead to a postoperative stenotic AV valve. Since the scope of this study was to evaluate the role of the ZoA in patients with otherwise normal left AVSD-AV valve morphology, we decided to exclude patients with left AV valve anomalies.

There were more patients aged 7–12 months in group II than in group I (univariate analysis). But since no difference exists between both groups with respect to the number of cases operated on during the first year of life and since cases of pulmonary hypertension were equally distributed, we presume that this fact is negligible. However, our policy in the last years has been to perform primary repair earlier in infancy.

To grade the regurgitation of the left AV valve at follow-up, we decided to use a semiquantitative colour Doppler method (measuring the maximum width of the jet and the size of the valve annulus in two orthogonal planes) which is a modification of the colour Doppler assessment described by Helmcke [18]. This method proved to be sufficiently sensitive and specific in the identification of left AV valve

Table 4
Left AV valve function at follow-up after repair of complete atrioventricular septal defect

<table>
<thead>
<tr>
<th>Function</th>
<th>All patients</th>
<th>Group I</th>
<th>Group II</th>
</tr>
</thead>
<tbody>
<tr>
<td>No/mild incompetence</td>
<td>98</td>
<td>25</td>
<td>73</td>
</tr>
<tr>
<td>Mild incompetence with mild stenosis</td>
<td>8</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>Moderate incompetence with mild stenosis</td>
<td>3</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Moderate incompetence</td>
<td>12</td>
<td>6</td>
<td>6</td>
</tr>
<tr>
<td>Severe incompetence</td>
<td>4</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Not assessable*</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>126</td>
<td>39</td>
<td>87</td>
</tr>
</tbody>
</table>

* Survivor after left AV valve replacement.
regurgitation and sufficiently accurate in estimating its severity. We are aware of the fact that there are several other echocardiographic methods to assess the severity of left AV valve incompetence with similar reliability.

4.2. Survival

Surgical repair of complete AVSD can be accomplished with acceptable early and late mortality. Early mortality in this series was 5.7% and corresponds well to the range of 2–16% that has been reported in the literature, [3,10,14,15,19,20]. Persistent high postoperative pulmonary arterial pressure is a well-known risk factor for early mortality [19]. Remarkably, four of the six patients in group I who died early died of persistent pulmonary hypertension whereas none of the three patients in group II who died early had pulmonary hypertension. Three of the four patients were older than 1 year at the time of repair. Our findings substantiate the widely held view that early intervention is a crucial factor for successful surgical repair and prevention of the development of irreversible pulmonary vascular changes.

Group I had a longer follow-up time than group II. To be able to make use of all the data available and to compare the resulting Kaplan–Maier curves, we applied the log-rank test. Although closure of the ZoA did not significantly reduce the rate of death in the first 30 postoperative days, it improved late survival considerably. This reduction in late mortality has been demonstrated for 9-years of follow-up of group II. It seems very likely that the improvement will be sustained, considering the stabilisation of the survival curves. Contrary to the less favourable results reported by others [3,14,21], the subset of patients with coexisting major cardiac anomalies did remarkably well in both groups of our series.

4.3. Reoperation

Regurgitation of the left AV valve is reported to lead to reoperation in 6–20% of patients after repair of complete AVSD [14–16,22]. Other indications such as residual shunts and patch dehiscence are infrequent [10,15,23]. Our per- operative analysis showed in most patients that the incompetence was due to an unsutured or reopened ZoA, sometimes in combination with additional anomalies, and that there was no chordal support in this area. Similar observations were mentioned in various publications [3,15,24,25].

Several authors have shown that the failure to achieve a competent left AV valve at AVSD correction is the most important risk factor associated with mortality and reoperation [3,14,15,21,22]. This is confirmed by our study. We were able to demonstrate that there were more postoperative cases with an incompetent AV valve in group I and that their outcome was worse than for group II patients.

During reoperation, every effort was made to preserve the native valve. We rather accepted to retain a mild to moderate incompetent postoperative valve than to replace it. Koboda and Jonas [26] have indicated that valve replacement in patients with AVSD (complete or partial), either during primary repair or at reoperation, was associated with elevated early mortality (22%), and carried a high risk of creating complete heart block. We confirm their observation: four out of five of our patients who underwent valve replacement eventually died. The only complete heart block in this series occurred after valve replacement.

4.4. Left AV valve function at follow-up

Over the course of time, the function of the reconstructed left AV valve is not necessarily stable; a moderately incompetent valve after reconstruction may improve, an initially competent left AV valve may deteriorate. Usually, mild regurgitation is stable and severely compromised left AV valve function is rare [20]. In our series, in a majority of valves where the ZoA was left untouched, valve function deteriorated significantly more than in those with a closed ZoA and early after AVSD correction. The reintervention included closure of the ZoA whenever possible, which in turn protected valvular function in the survivors: hence the similarity of valve function in both groups at follow-up echocardiography.

The main goal of left AV valve reconstruction in patients with complete AVSD is to create a competent valve without creating valve stenosis. Stenosis of the left AV valve after surgical reconstruction was rare among patients with a closed ZoA. This could be explained by the fact that during reconstruction an aged-related minimal normal mitral valve diameter was used as a guide during ZoA closure to prevent valve stenosis.

5. Conclusion

This study unequivocally demonstrates that patients with complete atrioventricular septal defect benefit from closing the ZoA, which is the gap between the facing surfaces of the left component of the bridging leaflets of the common AV valve. Suture-closure of the ZoA (‘cleft’) reduces the risk of developing severe regurgitation of the left AV valve as well as its deterioration, thereby decreasing the rate of reoperation. The procedure significantly improves survival and overall outcome. We conclude that, whenever the anatomy and size of the left AV valve allows closure of the ZoA during complete repair of a common AVSD, it should be closed.

References


