Atypical forms of isolated partial atrioventricular septal defect increase the risk of initial valve replacement and reoperation

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Abstract

Objective: We consider the short- and long-term outcomes of the repair of the isolated partial atrioventricular (AV) septal defect to determine the role played by the atypical forms on the initial AV valve replacement and on the risk of reoperation. Methods: Two hundred and eight patients underwent an operation for this malformation between 1974 and 2001. Clinical and echocardiographic examinations were performed on all patients, the AV valve regurgitation was graded from 1 to 4 and a residual interatrial shunt was sought. Median age at the intervention was 5.8 years (3 months to 67 years). Results: Median follow-up time was 7.5 years (range 0:22.6 years). The cumulative 30-day, 5- and 20-year survival rates were 96.5, 95.4 and 94.6%, respectively. AV valve replacement was associated with a high mortality (P < 0.001). A reoperation was performed on 12 patients (5.7%) including six patients within less than a 30-day period, especially to repair residual AV valve regurgitation. We performed four AV valve repairs by annuloplasty and six AV valve replacements. Two patients who had initially undergone an AV valve replacement underwent a reoperation for valve thrombosis. The cumulative 30-day, 5- and 20-year rates of freedom from reoperation were 96.5, 93.6 and 83%, respectively. An atypical form was present in 24 patients (11.5%) and was a risk factor for initial AV valve replacement (P < 0.001) and for reoperation (P < 0.001). A complete AV block occurred in 13 patients (6.2%), all of them within a 30-day period. The AV valve replacement was a high risk factor for a complete AV block (P < 0.001). At the end of our study 180 patients (96%) were in NYHA I and 8 in NYHA II. Conclusions: The morbi-mortality of the isolated partial AV septal defect is primarily perioperative and is linked with the presence of an atypical form of the lesion. This atypical form was the main reason for reoperation for AV valve regurgitation. The AV valve replacement was associated with a high mortality and with the occurrence of complete AV block. Using a standardized technique, the AV septal defect can be repaired with excellent long-term clinical and echographic results.

Keywords: Atrioventricular valve; Complete atrioventricular block; Left atrioventricular valve repair; Ostium primum; Partial atrioventricular septal defect

1. Introduction

The partial atrioventricular (AV) septal defect associates an ostium primum interatrial communication (IAC) with a septal commissure. Presenting symptoms are low which explains the high incidence of late diagnosis between 1 and 2 years of age. After surgical correction, the usual complications are complete AV block explained by the proximity of the AV knot to the patch sutures on the IAC, and residual left AV insufficiency necessitating left AV valve replacement. The goal of our study was to evaluate, over a short- and a long-term period, 208 patients, all of whom had undergone an operation for isolated partial AV septal defect by the same surgical technique, in order to indicate the early and late risk factors of morbidity and mortality. We determine the role played by the atypical forms on the initial AV valve replacement, and on the risk of reoperation.

2. Patients and methods

2.1. Population

We retrospectively studied 208 consecutive patients who underwent an operation for isolated partial AV septal defect between 1974 and 2001. Mean age at the intervention was 12.2 ± 11.7 years and median age was 5.8 years (range from 3 months to 67 years, Fig. 1). All partial AV septal defects presenting with an associated cardiac pathology were excluded from the study. The sex ratio was 92 men to 116 women. Thirty-two patients had Down’s syndrome (15.4%). We distinguished two groups: 24 patients (11.5%) with an atypical form and the remaining 184 patients (88.5%) with a ‘typical’ form.
2.2. Preoperative clinical state

Eighty-five patients were in NYHA I (40.8%), 100 in NYHA II (48.1%), 11 in NYHA III (5.3%) and 12 in NYHA IV (5.8%). One hundred and twenty-eight patients did not have any treatments during the preoperative period and 80 had cardiac treatments (digitalics, diuretics, beta-blocking agents, inotropic agents). One hundred and ninety-three patients (92.8%) presented cardiomegaly on the preoperative thoracic X-ray. The electric preoperative data are presented in Table 1.

2.3. Preoperative haemodynamic and echographic data

One hundred and fifty-nine patients had a preoperative haemodynamic exploration (Table 2). The recent cases also had a preoperative echography establishing the diagnosis of partial AV septal defect and evaluating left AV insufficiency. Seventy-six patients (37%) had a left AV insufficiency grade of I/IV, 109 patients (52%) had a left AV insufficiency grade of II/IV, 17 patients (8%) had a left AV insufficiency grade of III/IV, and 6 patients (3%) did not present any preoperative left AV insufficiency.

2.4. Surgical technique

All patients underwent an operation under cardiopulmonary bypass in moderate hypothermia (28–32 °C). The myocardial protection was always assured by an anterograde cold crystalloid cardioplegia. The surgical correction of the partial AV septal defect required the closure of the IAC (ostium primum) with a dacron patch leaving the drainage of the coronary sinus within the right atrium. Two hundred and two patients (97.1%) had a septal commissure repair with separated points. Twenty-four patients (11.5%) had an atypical form of the AV valve lesion and this lesion was associated with another valvular malformation. There were three accessory clefts (1.4%), six double-orifice left AV valves (2.9%), seven fenestrations of valve leaflets (3.4%), four malformed or malpositioned papillary muscles (1.9%), and four sequelae of previous bacterial endocarditis (1.9%). Of these 24 patients, six (25%) with irreparable valvular malformation, had a mechanical left AV valve replacement. Previously one patient underwent a left AV valve replacement with a Starr-Edwards tilting-disc valve, after which five other patients underwent a replacement with a St Jude bileaflet valve, the median size of the valve was 23 mm with a range from 17 to 29 mm. When a left AV valve annuloplasty was necessary because of a residual left AV insufficiency in the 30 postoperative days or in the long term, we used the implantation of a Carpentier ring. The median time for aortic clamping and cardiopulmonary bypass (CPB) were 33 min (range 10-78 min) and 50 min (range 13-110 min), respectively. The mean time for CPB in the group with atypical form was $61.1 \pm 29.3$ min compared with $50.2 \pm 11.6$ min in the group with typical form ($P=0.001$) and the mean time for aortic clamping in the group with atypical form was $38.6 \pm 13.3$ min compared with $32.7 \pm 8.7$ min in the group with typical form ($P=0.004$) (Table 3).

2.5. Follow-up

Patients were monitored from 1974 to 2002. In 2002, all patients were clinically examined, and underwent an electrocardiogram and trans-thoracic cardiac echography to assess residual interatrial shunt, left AV insufficiency and left ventricular function.

2.6. Statistical analysis

The description of continuous variables was expressed as a median (range minimal extreme to maximal extreme), or as a mean±SD. Categorical variables were presented as absolute numbers of patients and percentages. The statistical significance of the comparisons between two or several groups was tested using Pearson chi-square test and Fisher’s exact test, when necessary. Survival was calculated using a Kaplan-Meier method and compared with a log-rank test adjusted when necessary. A $P$ value of less than 0.05 was considered statistically significant. The analyses were carried out using SPSS software (version 11.5.1, SPSS, Inc.).

Table 1

<table>
<thead>
<tr>
<th>Preoperative electric data</th>
<th>Rhythm</th>
<th>Morphology</th>
<th>Conduction</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Sinus</td>
<td>Normal</td>
<td>10 (4.8%)</td>
</tr>
<tr>
<td></td>
<td>Junctional</td>
<td>Isolated LVH</td>
<td>10 (4.8%)</td>
</tr>
<tr>
<td></td>
<td>Ventricular (CAVB)</td>
<td>Isolated RVH</td>
<td>157 (75.5%)</td>
</tr>
<tr>
<td></td>
<td>AF</td>
<td>Biventricular hypertrophy</td>
<td>31 (14.9%)</td>
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<td></td>
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<td></td>
<td></td>
</tr>
</tbody>
</table>

CAVB, complete atroventricular block; AF, atrial fibrillation; LVH, left ventricular hypertrophy; RVH, right ventricular hypertrophy; RBBB, right bundle branch block; LBBB, left bundle branch block; AVB I, first degree atrioventricular block.
3. Results

3.1. Early results

3.1.1. Early mortality

Seven deaths occurred in the first postoperative month (3.4%) and none within the operative period. Two deaths were due to left cardiac insufficiencies in the immediate postoperative period (6 and 24 h). Both had initial left AV valve replacement. Four patients suffering from pulmonary hypertension died of right cardiac insufficiencies. Another patient died of complete AV block on the fourth postoperative day just before pacing.

3.1.2. Conduction complications

The postoperative conduction complications are reported in Fig. 2. The existence of a preoperative conduction disorder was not a risk factor for postoperative complete AV block ($P < 0.396$). Left AV valve replacement, either initial or secondary, was a major risk factor for complete AV block ($P < 0.001$).

3.1.3. Left AV valve complications

Four left AV valve replacements and two left AV annuloplasties were subsequently performed because of a residual left AV insufficiency of grade III/IV. The rate of early reoperations was 2.9%. An atypical form of the AV lesion was a risk factor for initial AV replacement ($P < 0.001$) (Table 3).

3.2. Late results

3.2.1. Follow-up

Late follow-up was completed for 198 patients out of 208 (95.2%). Mean follow-up time was 8.2 $\pm$ 5.3 years, and the median of the follow-up time was 7.5 years (range 0-22.6 years). Ninety-six percent of the patients were in NYHA I and had a normal life and 4% of the patients were in NYHA II. In echographic assessment, left ventricular shortening index was 40 $\pm$ 7%, with 15 patients less than 30%.

3.2.2. Late mortality

Three late deaths occurred, the first due to right cardiac insufficiency (1 year in postoperative), the second, neurological coma (1 year in postoperative after haemorrhagic complications of an initial left AV valve replacement) and the third, coma due to a car accident (7 years in postoperative). The actuarial survival curve is represented in Fig. 3. The cumulated survival rates at 30 days, 1, 5, 10 and 20 years were 96.5, 95.9, 95.4, and 94.6%, respectively. An age less than or equal to 2 years was a significant risk factor of mortality with an univariate test ($P < 0.0175$) but was not a risk factor if related to the AV valve replacement with a bivariate test ($P < 0.1738$). The occurrence of a left AV valve replacement was a risk factor of mortality with a bivariate test adjusted for age ($P < 0.0402$). We did not observe any deaths after 1989, knowing that 78 patients of the 208 underwent an operation up to and including 1989 (38%). The date of the intervention was a risk factor for mortality in our series, with a higher risk before 1989 ($P < 0.001$).

3.2.3. Conduction complications

We did not observe any complete AV block after the 30-day postoperative period.

3.2.4. Left AV complication

In four patients, we observed a deterioration of the result with the appearance of a significant left AV insufficiency. These four patients had to undergo a second operation: two of them had a left AV valve replacement (after 1 year) and two had a left AV valve repair (after 12 and 13 years). Two other patients who had had an initial left AV valve replacement underwent a second operation for left AV valve replacement because of prosthesis thrombosis.

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Table 3

<table>
<thead>
<tr>
<th>Intraoperative and postoperative data in both atypical form group and typical form group</th>
<th>Atypical form ($n = 24$)</th>
<th>Typical form ($n = 184$)</th>
<th>$P$</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Intraoperative data</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean age at the operation (year)</td>
<td>7.9</td>
<td>12.7</td>
<td>NS</td>
</tr>
<tr>
<td>Mean time of aortic clamping (min)</td>
<td>38.6</td>
<td>32.7</td>
<td>$P &lt; 0.004$</td>
</tr>
<tr>
<td>Mean time of CPB (min)</td>
<td>61.1</td>
<td>50.2</td>
<td>$P &lt; 0.001$</td>
</tr>
<tr>
<td>Initial AV replacements</td>
<td>6</td>
<td>0</td>
<td>$P &lt; 0.001$</td>
</tr>
<tr>
<td><strong>Postoperative data</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean follow-up time (year)</td>
<td>7.5</td>
<td>8.3</td>
<td>NS</td>
</tr>
<tr>
<td>Deaths</td>
<td>3</td>
<td>7</td>
<td>NS</td>
</tr>
<tr>
<td>Early complete AV block</td>
<td>4</td>
<td>12</td>
<td>NS</td>
</tr>
<tr>
<td>Late AV reoperations</td>
<td>7</td>
<td>3</td>
<td>$P &lt; 0.001$</td>
</tr>
<tr>
<td>Late residual interatrial shunt</td>
<td>0</td>
<td>9</td>
<td>NS</td>
</tr>
</tbody>
</table>

CPB, cardiopulmonary bypass; AV: atrioventricular.
(first patient at 4 years after initial surgery and the second, 18 years). On long-term follow-up the state of the AV valve after correction showed: no insufficiency in 30 patients (15.2%), insufficiency grade 1 in 100 patients (50.5%), grade 2 in 47 patients (23.7%), grade 3 in 5 patients (2.5%) and 16 patients (8.1%) had a left AV replacement or an annuloplasty. An age ≤2 years was a risk factor for left AV valve replacement in first or second intention (P <0.001). An atypical form of the AV valve lesion was a risk factor for AV valve reoperation (P <0.001) (Table 3). The rate of reoperations was 3%. The free from reoperations curve is represented in Fig. 4. The cumulated rates of freedom from reoperation were at 30-days, 1, 5, 10 and 20 years, 96.5, 95.9, 93.6, 92.8 and 83%, respectively.

3.2.5. Other complications

The echography showed a residual interatrial shunt in nine patients (4.5%). None of them led to reoperations. No patient presented with severe haemolysis syndrome. We observed neither left AV stenosis, subaortic stenosis nor bacterial endocarditis during the postoperative period. The results summary after surgical correction of the partial AV septal defect is represented in Table 4.

4. Discussion

The aim of the present study is to evaluate the early and late results of the surgical correction of partial AV septal defect and to underscore the factors of predicative morbidity and mortality. The results show that major mortality and morbidity mainly occurred at the hospital and depended on both the complete AV block occurrence and the necessity for a left AV valve replacement. The atypical forms of the AV valve lesion increase the necessity for an initial AV valve replacement and the necessity for a reoperation. The long-term follow-up showed excellent results in terms of left AV valve and septal repair, associated with a low rate of late complications.

4.1. Population

The interest of our study focuses on the fact that most of the main studies about partial AV septal defect [1–4] incorporated patients suffering from other associated cardiac anomalies such as aortic coarctation, pulmonary stenosis, ostium secundum IAC, double superior vena cava, anomalies of the venous return, or hypoplasia of the left ventricle. This is the first and largest series reporting of patients with an isolated partial AV septal defect and a large age-range. Some authors have only studied patients who were more than 40 years [5] or less than 5 [6]. We preferred to include all patients of all ages to analyze the possible risk factors associated with age for morbidity or mortality. We observed a high proportion of patients with Down’s

Table 4

<table>
<thead>
<tr>
<th>Complications</th>
<th>Early results</th>
<th>%</th>
<th>Late results</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>AF</td>
<td>3</td>
<td>1.4</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>CAVB</td>
<td>16</td>
<td>7.7</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Initial left AVVR</td>
<td>6</td>
<td>2.9</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Secondary left AVVR</td>
<td>4</td>
<td>1.9</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Secondary plastics</td>
<td>2</td>
<td>1</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Left AVVR redux</td>
<td>0</td>
<td>0</td>
<td>9</td>
<td>4.6</td>
</tr>
<tr>
<td>Residual IAC</td>
<td>0</td>
<td>0</td>
<td>9</td>
<td>4.6</td>
</tr>
<tr>
<td>Death</td>
<td>7</td>
<td>3.4</td>
<td>3</td>
<td>1.5</td>
</tr>
<tr>
<td>Drop out</td>
<td>0</td>
<td>0</td>
<td>10</td>
<td>5</td>
</tr>
</tbody>
</table>

AF, atrial fibrillation; CAVB, complete atrioventricular block; AVVR, atrioventricular valve replacement; IAC, interatrial communication.
syndrome (15.4%) compared to other authors [7-9], which could largely explain the good results obtained on the left atrioventricular valve correction. This population is less at risk for reoperation for residual left atrioventricular insufficiency because of a more favorable morphology for correction [10,11].

4.2. Surgical technique

Most of the authors [1,7,12] report several surgical procedures (suture or not of the septal commissure, annuloplasty, prosthetic or pericardial patch on the IAC) motivated by their own experience. All our patients were operated on using the same surgical technique (systematic closure of the septal commissure, dacron™ patch on the IAC, drainage of the coronary sinus in the right atrium) over a long period. We observed a smaller proportion of patients (11.5%) with atypical forms, compared to the high proportion (20%) observed by Abbruzzese. Although an atypical form of the AV valve lesion was a risk factor for initial AV valve replacement, we preferred to repair the valve as much as possible to avoid the need for replacement. Indeed the AV valve replacement is difficult in this specific anatomy and is a high risk factor of mortality. But in six patients with an atypical form we observed six impressive and unusual lesions (2.9%), four of whom were suspected to be sequelae of previous bacterial endocarditis (retracted posterior valve associated with shortened chordal apparatus) and the remaining two were malformed papillary muscles. In these six cases the lesion was irreparable and we decided to practice a left AV valve replacement. This high proportion of initial AV valve replacement in the group of atypical form explained easily the significant increase of mean time for aortic clamping and CPB in this group comparing with the group with typical form.

4.3. Early results

We did not observe any operative mortality and very low perioperative mortality (3.4%). This rate might be explained by the exclusion of the patients suffering from other cardiac anomalies. Hypoplasia of the left ventricle for example, is usually linked to a high operative and perioperative mortality [4]. Baufreton [3] has reported a 13% perioperative mortality mainly due to severe infections. We did not observe any dramatic infections in our patients. Overall mortality has evolved over time. We did not notice any deaths after 1989, which might be explained by the progress in medical pre and postoperative resuscitation. King [2] has observed the same phenomenon since the 1980. Like Baufreton [3], we did not find any age-death relationship, indeed we noticed a risk factor with an age $\leq$ 2 years for the outcome of death, but it was linked with the AV valve replacement, unlike King [2] who noted statistical differences for those aged 4 or less. The need for a left AV valve replacement, especially initial left AV valve replacement, was also related to overmortality and morbidity, given that the partial AV septal defect morphology is not favorable to implantation of a left AV valve prosthesis [13]. Left AV valve replacement lengthened the duration of the cardiopulmonary bypass and of cross-clamping, with subsequent higher morbidity and mortality rates.

The 13 observed complete AV blocks were diagnosed within the first 2 weeks, increasing the need for a permanent pacemaker (5.8%). This rate is higher than that recorded in other sources [2,7]. This proportion cannot be explained by our surgical technique. For some authors [14], leaving the coronary sinus drainage in the left atrium decreases the conduction complications, whereas Stuart did not observe any complete AV block while leaving the coronary sinus within the right atrium as we did [15]. For this reason, we did not modify our surgical technique. Currently there is no consensus on this specific surgical consideration. In our series the rate of preoperative conductive trouble is important and could be responsible for the postoperative complete AV block, but it was not demonstrated by statistical analysis, whereas a left AV valve replacement, either initial or secondary, was an important risk factor of complete AV block.

4.4. Late results

The late mortality was low (1.5%) with 95% of survival after 25 years. El-Nadjawi [7] has reported 87% of survivors after 20 years. Ninety-six percent of our patients are in NYHA I with excellent left ventricular functions. No complete AV block occurred past the 30-days postoperative period, confirming that conduction complications are linked to the surgical repair. The frequency of late reoperation is also acceptable (3%). Reoperations were necessary for serious left AV insufficiency in four patients and for thrombosis of left AV prosthesis in two. Abruzzese reported 18% of patients showing moderate to serious late left AV insufficiency, including 10% of patients who needed a reoperation, bearing in mind that within the series he did not suture the septal commissure in 31.1% of his patients [16]. Our results encourage a systematic suturing of the septal commissure [15]. Michelon also found that the reoperation rate for residual left AV insufficiency decreased with a bivalvar, instead of a trivulvar, approach to the left AV valve [17]. We found that an age less than or equal to 2 years at the time of the operation was a risk factor for left AV valve replacement in first or second intention. Additionally, Giamberti has also reported a high rate of reoperations (24%) for residual left AV insufficiency within a subgroup of children but for an age less than 1 year which he explains by the complexity of the left AV valve repair at this age [4]. We also maintain the operative presence of important lesions (large septal commissure, defect of the valvular tissue) as a predictive factor of AV valve replacement in first or second intention [13], but the most predictive factor was the presence of an atypical form of the AV valve lesion. Our low rate of reoperation might also be explained by the fact that we did not observe any subaortic stenosis unlike other authors [2,7], related in our opinion to the fact that we did not systematically use an annuloplasty in first intention. The weak rate of late secondary operation is also due to the absence of reoperation for a residual IAC unlike other authors [2,3,7], related, in our opinion, to the correction of the IAC with a patch instead of simple stitches [8]. Despite the use of prosthetic equipment for the IAC closure, we did not notice any serious haemolysis. Thus, the use of
a pericardial patch as some authors recommend, is questionable [15].

5. Conclusion

The morbimortality of partial AV septal defect is first and foremost surgical and perioperative. It is mainly linked to the anatomical shape of the partial AV septal defect, especially to the presence of an atypical form of the AV valve lesion, with the necessity of a left AV valve replacement and with the patient’s age. Once the perioperative period is over, the long-term prognosis is excellent; thanks to a standardized surgical technique including a systematic closure of the septal commissure, entailing minimal rates of reoperation for residual left AV insufficiency. Atypical forms of partial AV septal defect was a major risk factor for initial AV valve replacement and reoperation.

References