Predisposing Factors of Valve Regurgitation in Complete Atrioventricular Septal Defect

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Objectives. We sought to determine the intrinsic risk factors of valve regurgitation in complete atrioventricular septal defect.

Background. Progression of regurgitation varies in each case, although the structure of the common atrioventricular valve itself is a predisposing factor.

Methods. In 90 consecutive patients undergoing surgical repair, we evaluated the preoperative and postoperative regurgitation, valve morphology, age at surgery and associated anomalies. A regurgitation jet with a high velocity reaching the deep left atrial wall by echocardiography was estimated as marked regurgitation.

Results. None of the 40 patients with Rastelli type C and an undivided inferior bridging leaflet had preoperative regurgitation in the first year of life, and 12% of them (95% confidence intervals [CI]: 0% to 28%) showed regurgitation at the age of 2. Of the remaining 50 with Rastelli type A and/or a divided inferior leaflet, regurgitation was determined in 21% (95% CI: 6% to 35%) of those 1 year old and in 49% (95% CI: 29% to 69%) of those 2 years old (p < 0.01). All patients underwent corrective surgery using the double-patch technique, with the “cleft” being sutured adequately. Irrespective of the valve morphology, regurgitation remained in 52% (12 of 23) of those with preoperative regurgitation, whereas regurgitation developed postoperatively in 28% (16 of 58) of those without regurgitation (p < 0.001).

Conclusions. Those with Rastelli type C and an undivided inferior leaflet had a lesser degree of progression of preoperative regurgitation. However, regurgitation was likely to exist even after adequate repair once regurgitation had already advanced. Therefore, early primary repair before progression of the regurgitation may be the key to maintaining better competence of the atrioventricular valve.

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The results of surgical repair for atrioventricular septal defect with a common atrioventricular junction and orifice (the complete type) have improved markedly in the past few decades (1–4); however, valve regurgitation before and after surgery still remains an important issue (2–10). Because this malformation is characterized by not only a deficiency of the septum but also by an abnormal structure of the atrioventricular valve (11,12), there is an inherent problem of keeping the competence of the valve even after complete repair. Annular dilatation (10,13,14) and degeneration of the leaflets (6) have been considered to precipitate regurgitation, whereas little attention has been paid to the valve morphology itself (5,13). Since Rastelli et al. (15,16) classified this anomaly according to the configuration of the superior bridging leaflet, their classification has been widely used to delineate the structure of the atrioventricular valve. But one of the shortcomings of this is neglect of the different subtypes of the inferior bridging leaflet, which may have allowed us to overlook an inherent risk factor in the valve structure. The objective of our study, therefore, was to review the precise morphology of the atrioventricular valve in patients undergoing surgical repair and to clarify the predisposing factors of preoperative and postoperative regurgitation.

Methods

Patients. We studied a total of 90 consecutive patients with a complete atrioventricular septal defect undergoing corrective surgery between January 1982 and December 1997 in the Sakakibara Heart Institute in Tokyo. Cases with isomerism of the atrial appendages or a marked dominance of either ventricle were excluded from this study. The age at the time of complete repair ranged from 1 month to 13 years. There were 36 males and 54 females, and 78 patients (87%) had Down syndrome. Associated congenital heart abnormalities, other than secundum atrial septal defect and patent ductus arteriosus, were present in 15 patients (17%). Thirteen patients showed the morphologic features of tetralogy of Fallot, and the remaining 2 patients had coartation of the aorta. Palliative pulmonary artery banding was used as an initial step in 12 patients (13%). All 90 patients received cardiac catheterization within 2 months prior to the complete repair, which revealed severe pulmonary hypertension (greater than 80% of the
systemic blood pressure) and a large pulmonary to systemic flow ratio (greater than 2.0) in all except those with tetralogy of Fallot or those with a previous banding procedure.

**Measurements.** We evaluated the presence of regurgitation of the atrioventricular valve by echocardiography before and after surgical repair. A regurgitation jet with a high velocity reaching the deep left atrial wall in the echocardiogram was estimated as marked regurgitation (5,8,9). The precise morphology of the atrioventricular valve was inspected carefully during the operation. To determine predisposing factors of valve regurgitation and operative mortality, we evaluated the following variables: morphology of the superior and inferior bridging leaflets, age at corrective surgery, the presence of associated anomalies, previous pulmonary artery banding and Down syndrome.

**Analysis of data.** The multiple logistic regression model was used to estimate presenting features as risk factors of valve regurgitation before surgery. The Kaplan–Meier curve was constructed to determine actuarial freedom from preoperative regurgitation, and the generalized Wilcoxon test was performed for the statistical analysis between the groups. The relationship between preoperative and postoperative regurgitation was assessed by the McNemar test, and the chi-square test was used to compare the incidence between the noncontinuous groups. A p value of <0.05 was considered statistically significant.

**Results**

**Preoperative valve regurgitation (Table 1).** Of the 90 patients in the overall group, 26 (29%) showed marked regurgitation before surgery. In 51 patients undergoing surgical repair at an age of less than 1 year old, 5 (10%) had marked preoperative regurgitation, whereas 54% (21 of 39) of those aged 1 year or more at surgery had regurgitation (p < 0.001). As for the morphology of the inferior bridging leaflet, 68 (76%) patients had an undivided morphology, and the remaining 22 (24%) had the leaflet partitioned by a slit or raphe (the divided morphology). Marked regurgitation was determined preoperatively in 21% (14 of 68) of those with an undivided inferior leaflet, whereas 55% (12 of 22) of those with a divided morphology showed regurgitation (p < 0.01). None of the factors, such as the morphology of the superior bridging leaflet, presence of associated anomalies, previous pulmonary artery banding or Down syndrome, was related to the incidence of preoperative regurgitation.

Forty patients (44%) had a free-floating superior bridging leaflet (Rastelli type C) with an undivided inferior bridging leaflet, and 8 (9%) had type C morphology with a divided inferior leaflet. Rastelli type A morphology with an undivided inferior leaflet was observed in 28 patients (31%), and the remaining 14 (16%) showed type A with a divided inferior leaflet. The actuarial freedom from preoperative valve regurgitation was almost the same among the groups, except for those with type C and an undivided inferior leaflet (Fig. 1). In this group, it was 100% at 1 year, 94% (95% confidence intervals [CI], 82% to 100%) at 1.5 years and 88% (95% CI: 72% to 100%) at 2 years. In those with the other valve morphologies, on the contrary, the actuarial freedom from regurgitation was 79% (95% CI: 65% to 94%) at 1 year, 75%
Operative mortality. We performed corrective surgery in all cases with the double-patch technique, and both bridging leaflets and their supporting chords were never cut to place a patch. The so-called cleft (the zone of apposition between the bridging leaflets) was sutured adequately to keep the competence of the valve. There were nine operative deaths among the overall group of 90 patients (10%). Five died from refractory pulmonary hypertension and the remaining four died as a result of low cardiac output secondary to myocardial failure. As for their preoperative condition, three patients had suffered intractable heart failure and the other two had tetralogy of Fallot. The operative mortality decreased significantly over the period of this study from 21% (6 of 29) before 1990 to 5% (3 of 61) since then (p < 0.01). Two patients died late, 3 and 10 years after the operation, of recurrent pneumonia and cerebral infarction. Accordingly, deaths from valve regurgitation were not an important issue in this series.

Postoperative valve regurgitation. In the 81 patients who survived the operation, the follow-up period ranged from 4 months to 15 years (median 6.3 years). Marked regurgitation of the left atrioventricular valve was determined in 28 patients (35%) after surgical repair. Irrespective of the valve morphology, regurgitation remained after the operation in 52% (12 of 23) of those with preoperative regurgitation. On the other hand, marked regurgitation developed after surgery in 28% (16 of 58) of those without preoperative regurgitation (p < 0.001, Fig. 3). Seven patients underwent reoperation for the advance of regurgitation, from 1 month to 7 years after corrective surgery. The regurgitation in those cases was caused mainly by the remnant “cleft” with annular dilatation in four cases, laceration of the leaflet in two and a thick and hardly mobile superior bridging leaflet in the remaining one who subsequently received valve replacement. Of the overall 79 patients followed regularly, 76 (96%) were in New York Heart Association class I and the remaining 3 (4%) were in class II undergoing pharmacologic treatment. The incidence of asymptomatic regurgitation detected by echocardiography decreased to 15% (5 of 33) in the last 4 years.

**Discussion**

Surgical mortality of complete atrioventricular septal defect has improved dramatically in recent years (1–4). Regurgitation of the left atrioventricular valve, however, still remains a problem and reoperation is indispensable in some cases (1–3,7,9,13). A common atrioventricular valve postoperatively never creates structurally normal mitral and tricuspid valves, but it is possible to maintain better competence of the new valves by eliminating the predisposing factors of regurgitation.

**Predisposing factors of preoperative regurgitation.** There are several reports that preoperative valve regurgitation progresses as patients become older (6,10,13,14). Yusu et al. (6) reported that degeneration of the atrioventricular valve increases with age. Michielon et al. (10) also described annular dilatation as an important factor for regurgitation. In our series, not only was an older age at operation an independent risk factor for preoperative regurgitation, but also the mor-
dilatation, degenerative changes and dislocation of the whole valve (6,10,13,14), it will be difficult to stop the regurgitation completely even with adequate repair and annuloplasty (2,10,14).

As described previously, the first 6 months of life should be the golden time for definitive repair (6,7,10,13,14,18) because not only preoperative regurgitation, but also pulmonary vascular obstructive disease hardly develops at this period (19,20). We have been performing corrective surgery routinely as soon as the diagnosis was completed, and blood transfusion is seldom necessary when patients weighed more than 4.0 kg. The long-term prognosis remains to be clarified; however, none of these cases had symptomatic regurgitation or was required to continue medical treatment. The incidence of regurgitation estimated by echocardiography also markedly decreased.

For a patient with complex anomalies in addition to an atrioventricular septal defect, the time of surgical repair varies in each case. In those with tetralogy of Fallot, for example, the size of the pulmonary artery is also an important factor for successful repair. Fortunately, almost always these cases had type C morphology of the superior bridging leaflet (21) and about 70% of them had an undivided inferior bridging leaflet. We might be able to wait for the growth of the pulmonary artery for a few years without much fear of progression of regurgitation in some cases of this combination. Hypoplasia of the left ventricle or an abnormal structure of the left atrioventricular valve, such as dual orifices and a small mural leaflet, are other reasons for postponing surgical repair. Those patients often received pulmonary artery banding as an initial step, although it remains to be established whether the banding procedure delays the development of valve regurgitation (3,9,18).

**Study limitations.** Our improved surgical technique and perioperative care over the period of this study may have biased the intrinsic risk factors of the common atrioventricular valve and the recent decrease in the postoperative regurgitation. We evaluated the presence of valve regurgitation by echocardiography, but this estimation may not always correlate with symptomatic regurgitation because of its hypersensitivity. Another important issue is that extrinsic factors other than the age at surgery, such as infection and cyanosis, may also influence the progression of preoperative regurgitation.

**Conclusions.** The development of valve regurgitation varied with the morphology of the common atrioventricular valve in patients with complete atrioventricular septal defect. Those with Rastelli type C and an undivided inferior bridging leaflet had a lesser degree of progression of preoperative regurgitation than did those with the other valve morphologies. However, regurgitation was likely to remain even after adequate repair in patients with complete atrioventricular septal defect. Those with Rastelli type C and an undivided inferior bridging leaflet had a lesser degree of progression of preoperative regurgitation than did those with the other valve morphologies. Therefore, primary repair in the first 6 months of life, before the development of annular dilatation and degenerative changes of the valve, may be the key to maintaining better competence of the atrioventricular valve.

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**Figure 4.** In those with Rastelli type A and/or a divided inferior leaflet, the coaptation zone between both bridging leaflets is triangular or diamond shaped (painted in black). On the other hand, those with Rastelli type C and an undivided inferior leaflet, showing a lesser degree of progression of preoperative regurgitation, do not have such a structure.
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