USE OF A PROSTHESIS FOR REPAIR OF COMPLETE COMMON ATRIOVENTRICULAR CANAL

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From April, 1977 to November, 1979, 5 cases with common atrioventricular canal (Rastelli C-type) underwent surgery, in which the leaflets of both the mitral and tricuspid valves were extended using an auto-pericardium patch in order to control regurgitation. Their ages ranged from one year 3 months to 6 years (mean, 3 years 7 months). One case died on the 37th postoperative day from intracranial bleeding unrelated to the cardiac procedure. In the remaining 4 cases, postoperative examination showed an improvement, and their clinical courses which were followed from 2 years 7 months to 5 years 3 months, were favourable, with only minimal valvular regurgitation remaining in all cases.

Although the fate of the pericardium patch must be followed over a long period, this procedure appears to be effective for control of regurgitation in this disease.

IN persistent common atrioventricular canal, the surgical procedure is performed with utmost difficulty. Technical problems include the avoidance of the occurrence of conduction disturbances and the effective repair of the atrioventricular valves. The latter is more difficult to accomplish.

The atrioventricular valves have a wide variety of deformity not only in their cusps but also in their chordae tendineae and their papillary muscles. Because of the delicate and dynamic functions of these valves, the relationship between the shape and the function can hardly be

evaluated even during open heart surgery, and this is the primary factor which is responsible for the poor surgical results in this disease.¹⁻⁷

In the history of surgical procedure for complete persistent common atrioventricular canal, a dramatic improvement was seen in 1968 when Rastelli et al⁶ proposed an operation without using artificial valves, which is now widely used as the standard procedure for this disease.^{8,9} However, in severely malformed atrioventricular valves considerable regurgitation usually persists after the operation.

Recently, we have repaired 5 cases with Rastelli C-type by extending the leaflets of the atrioventricular valves using a flap of pericardium attached to the patches for septal defect closure. This new procedure was designed to minimize the postoperative regurgitation and appears to be effective for this purpose.

Key Words:

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TABLE I SUMMARY OF FIVE CASES: PREOPERATIVE DATA, FINDINGS AT SURGERY AND RESULTS

Case No.	Age, sex & body weight	Diagnosis	Physical findings	ECG findings	Hemodynamics	Operative findings	Results
1	1 year 3 months Female 6 kg	ECD (Rastelli-C) PDA Down syndrome	Cyanosis (+) Retardation of growth (+)	Axis -90° RVH LVH	Pp/Ps = 1.0 Shunt: L to R 62.2% R to L 17.7% Goose neck (+) MR: III	Rastelli-C Goor IIb VSD: 10mm in depth	good MR: minimal
2	4 years Male 14 kg	ECD (Rastelli-C) PS Down syndrome	Cyanosis (+) Retardation of growth (+)	Axis -120° A-V block (1°) RVH	PA pressure 19 mmHg RV inflow 105 mmHg Shunt: L to R 35.9% MR: +3	Pastelli-C Goor IIb VSD:	Chylothorax; died on the 37th day after surgery from cerebral bleeding; MR: minimal
3	5 years Female 15 kg	ECD (Rastelli-C) PH	Cyanosis (-) Retardation of growth (+)	Axis -80° PQ 0.20" IRBBB RVH LVH	Pp/Ps = 83/86 (mmHg) Shunt: L to R 52.2% R to L 23% Goose neck (+) MR: +2	Rastelli-C Goor IIb VSD: 8mm in depth Infundibular PS: abnormal muscle band	good MR: +1 PH: 45 mmHg
4	3 years Male 11 kg	ECD (Rastelli-C) PH Down syndrome	Cyanosis (+) Retardation of growth (+)	Axis -70° IRBBB PQ 0.14" RVH, LVH	Pp/Ps = 1.0 Shunt: L to R 55% R to L 22% Goose neck (+) MR: (-)	Rastelli-C VSD: 7mm in depth	good MR: minimal
5	6 years Female 15 kg	ECD (Rastelli-C) PDA PH PS (Fallot type)	Cyanosis (+) Retardation of growth (+)	Axis -160° PQ 0.17" RVH	Pp/Ps = 70/83 (mmHg) Shunt: L to R 63% R to L 33.8% Goose neck (+) MR: +2	Rastelli-C Infundibular PS (Fallot type)	good MR: minimal

Abbreviations: ECD = endocardial cushion defect; PDA = patent ductus arteriosus; PS = pulmonary stenosis; PH = pulmonary hypertension;
 ECG = electrocardiogram; RVH = right ventricular hypertrophy; LVH = left ventricular hypertrophy; IRBBB = incomplete
 right bundle branch block; Pp/Ps = pulmonary pressure/systemic pressure; MR = mitral regurgitation; VSD = ventricular septal
 defect

Fig.1. Operative procedure of valvuloseptoplasty. A: Teflon patch with the pericardium sutured to the muscular septum on the right ventricle. B: Anterior and posterior halves related to the mitral valve were approximated. Then, the pericardium patch was sutured to these halves so that valvuloplasty with a slightly larger septal cusp of the mitral valve can be performed. C: Atrial septal defect was closed using elastic Dacron patch. D: Finally, valvuloplasty for the septal leaflet of the tricuspid valve was performed.

MATERIALS

Cases are listed in Table I. They were 2 males and 3 females, ranging in age from 1 year 3 months to 6 years 1 month with a mean age of 3 years 7 months. All except Case 3 had cyanosis. Their hemoglobin ranged from 13.4 to 14.6 g/dl (mean 14.02), their hematocrit 40 to 44.2% (mean 42.2) and their red blood cell count 4.6 to 7.1 million/mm³ (mean 5.5). Retardation of growth was observed in all cases. Three had Down syndrome. Chromosomal test revealed a 47 and XX + G in Case 1, a 46 + XX in Case 3 and a 21 trisomy in Case 5.

Cardiac catheterization and angiography were performed in all cases. High pulmonary arterial pressures nearly equal to the levels of systemic pressure were found in Cases 1, 3 and 4. In Case 2 infundibular stenosis with a systolic pressure gradient of 86 mmHg was revealed, and in Case 5 septal and parietal band hypertrophy, just like

tetralogy of Fallot, was noticed in the right ventricle. Goose neck sign was found in all cases.

SURGERY AND RESULTS

Operative Procedures

Findings during surgery confirmed that all cases belonged to type C of Rastelli's classification.

Circulatory arrest with deep hypothermia was applied in Case 1 and moderate hypothermia and topical cooling with a cardioplegic solution was used in the remaining 4 cases.

Valvuloseptoplasty was done essentially in the same manner. In the first 2 cases, however, a pericardial patch was used not only for valvular extension but also with the fabric patch for septal closures. In the remaining 3 cases a pericardial patch was used only for the extension of the leaflets.

Case 1: Following a median sternotomy, the

Fig.2. Modified filling material for the valvuloseptoplasty. Pericardium patch was used only for extension of the leaflets.

ductus arteriosus of 3 mm in diameter and 8 mm in length was ligated. Then, prior to extracorporeal circulation, 2 pieces of the pericardium were prepared for later use as a "filling material (prosthesis)" (Fig. 1).

Observation of the inside of the heart revealed no lobulation of the common anterior cusp, and this case was diagnosed as persistent common A-V canal of the Rastelli C type.

Lobulation of the common posterior cusp, corresponding to Goor IIb,¹⁰ was observed. The interventricular defect was 2.5 cm wide and 1 cm deep. The coronary sinus was present in the right atrium.

At first, the common anterior cusp was divided into the left and right halves parallel with the ventricular septum, while the common posterior cusp was cut into halves along the lobulation. To close the ventricular septal defect the first patch was sutured to the muscular septum on the right ventricular side using mattress sutures with spaghetti pledget.

Stitches in the posterior half were made shallowly, as low as possible in the right ventricle below the upper margin of the septum in order

to prevent the occurrence of conduction system damage.

The height of the patch for closing the septal defect was decided from the cusp height estimated in systole. Next, the free edges of the halved common anterior and posterior leaflets relating to the mitral valve were sutured together and the prepared pericardium was sutured in place in order to construct a larger anterior leaflet of the mitral valve. Mitral valve function was evaluated by injecting a saline solution into the left ventricle through the left ventricular vent, and regurgitation was observed to be minimal.

Then, the atrial septal defect was closed using a second patch. Stitches were carefully placed to avoid impairing the conduction system, which runs around the opening of the coronary sinus.

Finally, valvuloplasty for the septal leaflet of the tricuspid valve was performed. Sinus rhythm was restored when the cardiac beat resumed, but precautionary pacemaker wires were sutured before the chest was closed.

Case 2: Surgery was performed under a moderate hypothermic extracorporeal circulation of 28°C with topical cooling. Intracardiac

Fig.3. Chest roentgenogram of Case 1. Before (A) and after surgery (B).

procedures were the same as Case 1. Abnormally hypertrophied septal and parietal muscle bands, which had induced infundibular stenosis, were resected at the level of crista supraventricularis.

Case 3 and Case 4: The operation were done in almost the same manner as in the previous cases, but the pericardial patch was used only for valvular extension, as shown in Fig. 2.

Case 5: Surgery was done under a moderate hypothermia of 28°C with topical cooling. The infundibular stenosis consisted mainly of a hypertrophied parietal band similar to that in Fallot's tetralogy, and overriding of the aorta was also distinctive. The procedure of valvuloseptoplasty was the same as in Cases 3 and 4 except that the overriding required special consideration in closing the ventricular septal defect. Infundibulectomy was done by making another incision in the right ventricle.

Postoperative Course

All cases had smooth postoperative course except Case 2 who had bilateral chylothorax and died from intracranial bleeding on the 37th postoperative day. The remaining 4 patients are living and performing normal activities, although minimal mitral regurgitation has remained.

Case 1: For 3 days after surgery, controlled respiration and administration of isoproterenol were needed, but thereafter, her recovery was smooth. She was discharged 2 months after surgery, when her chest roentgenogram showed a

decrease in the cardiothoracic ratio from 66% (before surgery) to 63%, as shown in Fig. 3. Her postoperative electrocardiogram disclosed a right bundle branch block, but no change was seen in the P-Q interval.

As shown in Fig. 4, her phonocardiogram showed only a faint systolic murmur and the accentuated pulmonary component of the second heart sound, suggesting residual pulmonary hypertension. As seen in Fig. 5, her echocardiogram showed an intact ventricular septum with bright patch echoes. Sufficient amplitude and a good diastolic descent rate of the mitral valve were seen and the amplitude of the tricuspid valve was well maintained.

Case 2: The clinical course was satisfactory immediately after surgery. On the first postoperative day the intratracheal tube could be removed. An electrocardiogram showed sinus rhythm. Only an ejection murmur attributable to residual pulmonary stenosis was heard.

A chest roentgenogram on the 3rd postoperative day disclosed bilateral hydrothorax. The fluid was identified as chyle and the patient was treated conservatively with dietary control and chest drainage. On the 33rd day after surgery the chyle flow stopped, but the patient died on the 37th postoperative day due to serious hemolytic jaundice and acute renal insufficiency complicated by intracranial bleeding.

Case 3: The postoperative course was smooth without any complication. An angio-

Fig.4. Postoperative phonocardiogram of Case 1. A slight systolic murmur and an accentuated pulmonic component of the 2nd heart sound is noticed.

gram after surgery showed only minimal mitral regurgitation.

Case 4: An episode of bradycardia with a heart rate of 50 per minute occurred on the 21st postoperative day, but was successfully treated medically.

A grade 2/6 apical systolic murmur persisted.

Case 5: Postoperatively, right heart failure occurred and intratracheal tube could not be removed until the 8th postoperative day. An artificial pacemaker was required for 2 weeks after surgery because of episodes of bradycardia. After 3 weeks, however, the patient had recovered satisfactorily and no significant cardiac murmur was present.

DISCUSSION

Although a remarkable improvement in surgical procedures has been attained as compared with those in earlier reports^{2,8,9} atrioventricular regurgitation remains to a significant degree

after surgery. The difficulty in surgical procedure for persistent common A-V canal may be attributable to its complicated morphology, especially to the extreme malformation of the atrioventricular valves.

The septal cusp of the tricuspid valve and the anterior leaflet of the mitral valve are defective in the central portion of the valve.

In the method of Rastelli, the ventricular and atrial septal defects are closed by a single patch to which the leaflets are sutured. By this method the originally hypoplastic septal cusp should decrease in size further, causing the inadequate prevention of regurgitation (Fig. 6).

In this regard, an improvement has been made by Shirotani, Kawai et al.¹¹ They inserted common cusps of the atrioventricular valves in between the patches, closing the ventricular septal defect and the atrial septal defect, without cutting the common cusps (Fig. 6).

In our valvuloplasty, as in the method of Rastelli, the height of the atrioventricular valve

Fig.5. Postoperative echocardiogram of Case 1. Arrows show the interventricular patch. p(IAS) = patch of interatrial septum; TV = tricuspid valve; MV = mitral valve; PLVW = posterior wall of the left ventricle

Fig.6. Methods of repair of complete form of persistent atrioventricular canal. Comparison of Rastelli's, Shirokani's and the present method. RA = right atrium; LA = left atrium; RV = right ventricle; LV = left ventricle

in the ventricular septal defect was adjusted when the flap of the pericardium was sutured to the patch. The size of the patch was determined according to the height of the valve during ventricular systole. The atrioventricular valves are thus enlarged so as to control regurgitation. In other words, by using pericardium, slightly larger than the margin for suture, valvuloplasty could be accomplished and the hypoplastic septal cusps could be enlarged (Fig. 6).

Adaptation of the valves actually should be made by the formation of the valve margin not as a point but as a face with convexity towards the atrium. Therefore, regurgitation does not seem

to be increased even if the septal cusp is enlarged a little too much.

The present method can be designated as a valvuloseptoplasty with "endocardial cushion prosthesis". This idea was reported independently by Kawashima et al.¹² in 1975. They used Dacron patch for valvular extension. Naito et al.¹³ have reported one case with valvular stenosis after the use of Dacron patch for valvular extension and 2 cases with acceptable results using pieces of auto-pericardium.

The procedure of giving a greater share of the common leaflets to the mitral valve than to the tricuspid valve has a high incidence of significant tricuspid regurgitation, which is often followed by a low cardiac output.

The present procedure can be characterized as simple and easily standardized, with stable hemodynamics in the early postoperative stage.

We have some concern about the possible shrinkage of the pericardium. However, as the septal cusp maintains its continuity with auto-tissue, growth of the septal cusp may be expected, although long-term follow-up is necessary before any conclusion can be drawn.

As for the timing of the operation, in cases without pulmonary stenosis the operation should be performed under 2 years of age, similar to cases with isolated ventricular septal defect associated with severe pulmonary hypertension.^{8, 15, 16}

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