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Mitral Valvuloplasty for Isolated Congenital Mitral Valve Regurgitation

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Summary. **Mitral valve regurgitation (MR) is uncommon as an isolated lesion in congenital heart disease. In 1995, we encountered two children with congenital isolated MR complicated with heart failure, and both underwent valvuloplastic surgery. Resection and/or plication of the redundant leaflets combined with annuloplasty for the dilated annulus successfully restored the valve competency. Traditionally prosthetic valve replacements have been performed for this condition; however, as a prosthetic valve inhibits annular growth in children, a gradual progression of mitral stenosis is the main problem later in life. Valvuloplasty allows the growth of the valve apparatus, and moreover, does not require postoperatively the troublesome life-long anticoagulation therapy. Valvuloplasty in congenital MR is enabled with a precise application of reconstructive techniques, and should be the procedure of choice for pediatric patients.**

Key words—congenital mitral regurgitation, mitral valvuloplasty, pediatric cardiac disease.

INTRODUCTION

Mitral valve regurgitation (MR) in congenital heart disease is usually associated with a left-to-right shunt anomaly such as a ventricular septal defect or atrioventricular canal. Isolated congenital MR is uncommon, and reports of its surgical repair during childhood are limited.[1-3] Although the valvuloplasty technique has been widely applied to patients with acquired MR in recent days,[4-6] a report of this type of surgery in congenital MR is rare.[7-9] In 1995 we encountered two pediatric patients with isolated congenital MR, who were successfully treated by valvuloplasty. We here report on the surgical details of the two cases.

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PATIENT 1

A girl at the age of 1 year and 10 months was admitted with a complaint of "failure to thrive". Although she had been found to have a loud cardiac murmur since birth with a diagnosis of congenital MR established by echocardiography, surgical treatment was deferred as a feasibility and the efficacy of the valvuloplastic surgery was questioned during early infancy. This time the patient was admitted due to severe and progressive heart failure. On admission, she was emaciated, with a body weight of 8.4 kg. A harsh systolic murmur (Levine 3/6) was audible at the apex, and the second heart sound was accentuated. The electrocardiogram (ECG) showed biventricular hypertrophy. The chest X-ray revealed cardiomegaly (cardiothoracic ratio 75%) and pulmonary congestion. The echocardiography showed an incompetent mitral valve due to poor coaptation of the leaflets. The cardiac catheterization showed pulmonary hypertension with a pulmonary artery pressure of 68/34 (mean 49) mmHg. The mean pulmonary capillary wedge pressure was 22 mmHg. The left ventriculogram demonstrated severe mitral regurgitation (grade 4/4), with a slight prolapse of the anterior leaflet (Fig. 1A). The operation was performed under hypothermic extracorporeal circulation. After cardiac standstill induced by cardioplegic solution, the left atrium was incised. The mitral valve was exposed and inspected. The anterior leaflet was

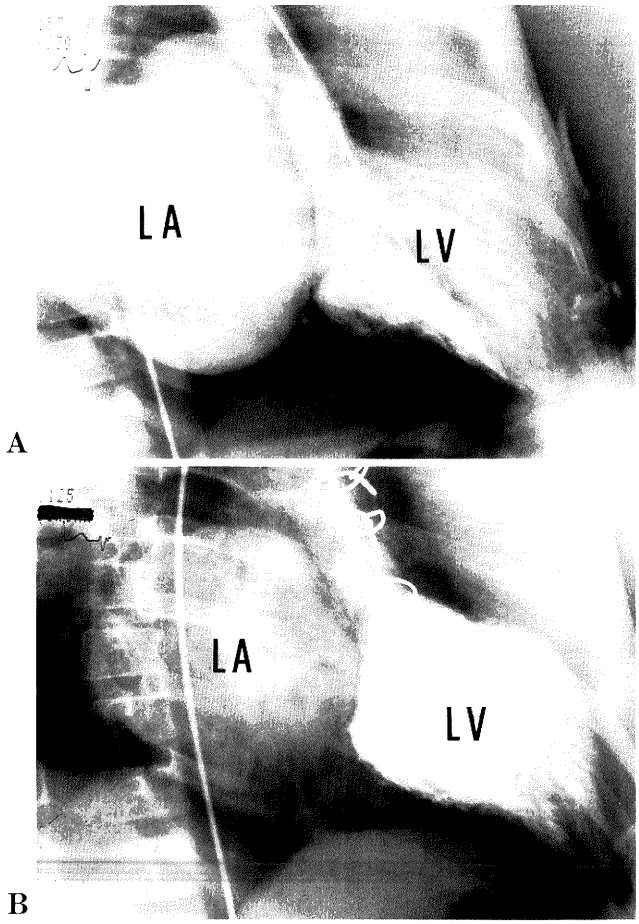

Fig. 1. The left ventriculogram of Patient 1 from the right anterior oblique view. LA; left atrium, LV; left ventricle. **A.** The preoperative state. Severe mitral regurgitation is observed. **B.** The postoperative state. The regurgitation is mild.

redundant, and the posterior leaflet was hypoplastic with a limited area of the clear zone. Annular dilatation and chordal shortening beneath the antero-lateral commissure were observed. There was no cleft in either leaflet. The valve competence and the leaflet motion were assessed by cold saline injection into the left ventricle. There was a massive regurgitation from the orifice because each coaptation zone of the leaflets stood at a different level. The reparative procedure consisted of plication of the redundant anterior leaflet and commissure annuloplasty, as described by Kay et al. (Fig. 2).[10] At the end of the procedure, the saline injection test indicated complete disappearance of the regurgitation. The patient tolerated the surgery well, and had a smooth postoperative course. She was discharged from the hospital 3 weeks after the surgery with substantial symptomatic improvement. The cardiac catheterization 3 months after the surgery revealed the disappearance of the pulmonary hypertension. The pulmonary artery pressure was 28/10 (mean 18) mmHg, and the mean pulmonary capillary wedge pressure was 9 mmHg. The left ventriculogram revealed mild residual regurgitation (Fig. 1B). Now she is showing normal development without any complaints.

PATIENT 2

A 13-year-old boy, who had been hospitalized for relapsing autoimmune hemolytic anemia, was referred to the cardiac surgery department because of progressive heart failure. In 1994, the patient started to complain of shortness of breath and hypersweating. He had no history of rheumatic fever. Although the patient had been known to have a cardiac murmur since early childhood, no close cardic examination was performed until March, 1995. On physical examination, he had a slightly obese constitution due to steroid therapy. His body weight was 35 kg, the blood pressure was 110/70 mmHg, and the heart rate was 90/min. A loud systolic murmur was audible at the apex. The ECG showed a sinus rhythm with left ventricular hypertrophy. The chest X-ray revealed pulmonary congestion and marked cardiomegaly with a cardiothoracic ratio of 71%. The echocardiography indicated severe mitral valve regurgitation with anterior leaflet prolapse. The flow velocity at the valve was 4.6 m/sec. The left ventricular contraction was reduced to 30% of ejection fraction. On cardiac catheterization, the pulmonary artery pressure was 27/17 mmHg, the mean pulmonary capillary wedge pressure was 13 mmHg with a v-wave of 16 mmIlg, and the left ventricular pressure was 94/14 mmHg. The left ventriculography demonstrated grade 4/4 mitral rugurgitation. Because of the development of cardiac symptoms, surgery was indicated. Under hypothermic extracorporeal circulation, the heart was arrested by cardioplegic solution. The mitral valve was visualized through the left atriotomy. On inspection of the valve tissue, both anterior and posterior leaflets were redundant. Anterior leaflet prolapse with chordal elongations was also observed. The closure of the leaflets was incomplete, and by saline injection into the left ventricle, a massive regurgitation was recognized on the entire line of the coaptation zone of the leaflets. Therefore the reconstructive procedure consisted of the partial excision of the redundant tissue in both leaflets, and annuloplasty using a Carpentier ring (Fig. 3). Care was taken to make each coaptation zone of the

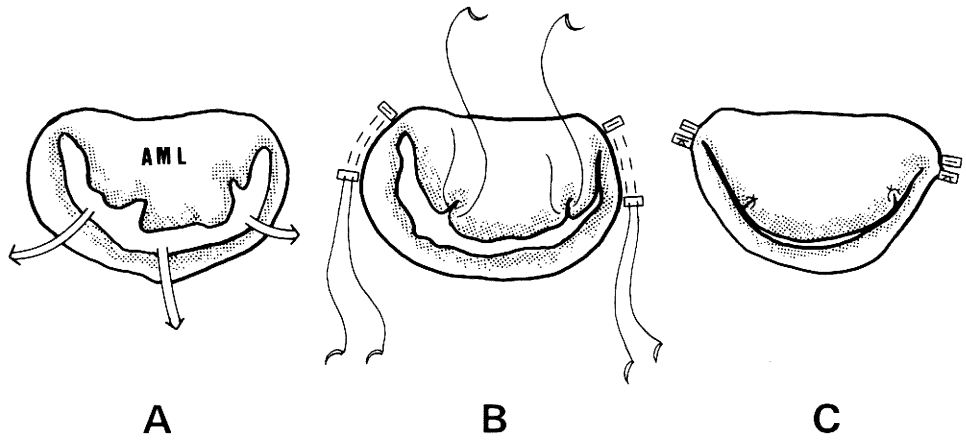

Fig. 2. The operative findings and the reparative procedure for Patient 1. **A.** A large amount of regurgitation *(arrows)* comes from the entire orifice. Redundant anterior leaflet, hypolastic posterior leaflet, annular dilatation, and chordal shortening beneath antero-lateral commissure are observed. AML; Anterior mitral leaflet. **B.** Plication of the anterior leaflet at two points and commissural annuloplasty on both sides were performed. **C.** The coaptation zone of both leaflets meets well, resulting in a competent valve.

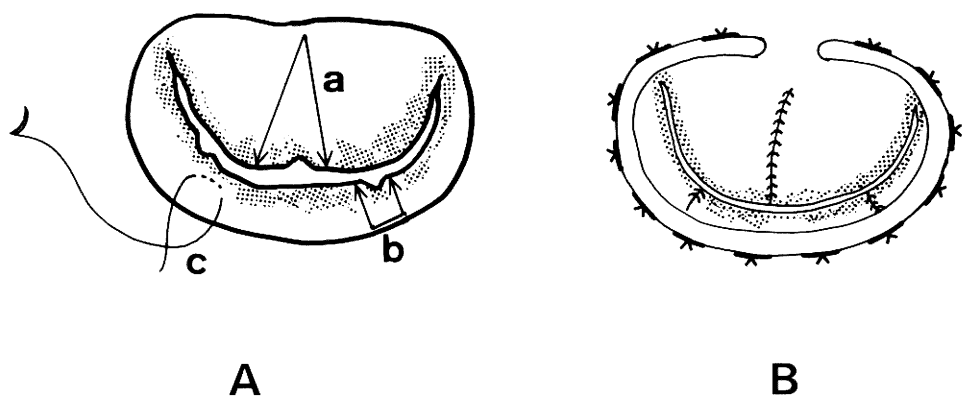

Fig. 3. The reparative procedure for Patient 2. **A.** A triangular resection at the middle portion of the anterior leaflet (a), and a quadrangular resection at the postero-medial side of the posterior leaflet (b) were performed. Plication of the posterior leaflet at the antero-lateral side (c) was added to make the coaptation zones meet at the same horizontal level. **B.** Carpentier annuloplasty ring (size 28 mm) was sutured to the annulus. Complete closure of the leaflets was obtained.

leaflets meet at the same horizontal level. After a 28 mm Carpentier ring was fixed to the annulus, no regurgitation was observed. The patient was weaned from the extracorporeal circulation, and the postoperative course was uneventful. Anticoagulation therapy by warfarin administration was maintained for 4 months after surgery, and then discontinued. In the fifth postoperative month, the patient was well without any heart murmur. The chest X-ray showed an improvement of pulmonary congestion, and the cardiothoracic ratio decreased to 61%. The echocardiography demonstrated the disappearance of mitral regurgitation, and the left ventricular ejection fraction was increased to 39%.

DISCUSSION

For young active patients, prosthetic valve replacement poses many problems including later stenosis,[11-14] and life-long anticoagulation with its attendant thromboembolic versus hemorrhagic risks is not desirable. Therefore valvuloplastic surgery should be the procedure of choice for congenital MR, but reports of such surgery are limited. In 1962, Creech et al. first reported a 2-year-old patient with congenital MR with a cleft in the posterior leaflet which was repaired by suturing the cleft.[1] In clinical settings, congenital MR is most often encountered in coexistence with other intracardiac anomalies such as ventricular septal defect, atrial septal defect, or atrio-ventricular canal. In these instances, the pathological change of the mitral valve is usually a cleft formation in either leaflet, and closure of the cleft sufficiently restores valve competence in most cases.[2] When congenital MR is caused by anomalies other than clefts, the reparative procedure is not simple and the operative technique varies, owing to the underlying pathology of the mitral valve. In adult cases, many reparative techniques have been described,[4-6,10] and essentially these techniques can be used in congenital MR in pediatric patients. However, as the size of the mitral valve is much smaller and the growth of valve apparatus has to be anticipated, surgeons must be cautious in applying these techniques in young children. In Patient 1, the repair could be accomplished by plication of the anterior leaflet combined with commissure annuloplasty. In Patient 2, partial resection and suture of both leaflets combined with ring-annuloplasty successfully established the valve competence. Usually in pediatric patients, ring-annuloplasty is not employed because of the limitation of annular growth. However, in this particular case, the annulus was dilated and the size of the anterior leaflet indicated a 28 mm ring, which was sufficient even for an adult patient, and therefore a 28 mm Carpentier ring was used to reinforce the annuloplasty. In both cases, the basic concept of the repair was to attain a precise coaptation of leaflets at the same horizontal level by reducing the redundancy of the leaflets and shortening the dilated posterior annulus. With the experience of these two cases, the feasibility and adequacy of an application of reparative technique in young children was confirmed. In order to reserve the potential for valvular growth, children with congenital MR should be repaired by valvuloplasty whenever possible.

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