

## CASE REPORT

# Long-term Results After Open Mitral Commissurotomy for a One-Month-Old Infant With Mitral Stenosis

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**Abstract :** The strategy for an infant with congenital mitral stenosis should be determined by three important factors : left ventricular volume, the degree of the systemic outflow tract obstruction, and the type of mitral valve dysfunction. A successful staged biventricular repair in early infancy for a patient who had congenital mitral stenosis with short chordae, hypoplastic left ventricle and coarctation of the aorta, and the long-term results are described. There were the following important hemodynamic factors that led to the successful biventricular repair in the patient. Total systemic output was barely supplied through the hypoplastic left ventricle after closure of the ductus arteriosus on admission. The neonate underwent repair of coarctation of the aorta alone as the initial stage at 9 days after birth. Also, spontaneous closure of the foramen ovale following repair of coarctation of the aorta accelerated the progressive left ventricular growth. Open mitral commissurotomy with an interatrial fenestration using the modified Brawley's approach was performed for a 40-day-old infant. Good left ventricular growth and good mitral valve function have been observed for 18 years after open mitral commissurotomy. Appropriate early augmentation of left ventricular inflow through the mitral valve might be effective for growth of a hypoplastic left ventricle. *J. Med. Invest.* 64 : 187-191, February, 2017

**Keywords :** congenital mitral stenosis, open mitral commissurotomy, hypoplastic left ventricle, short chordae, long-term results

## BACKGROUND

Symptomatic mitral stenosis in early infancy is difficult to manage because of a dysplastic mitral valve apparatus and high prevalence of associated cardiac anomalies. Although the potentiality of biventricular repair should be considered as a definitive operation for surgical strategies in infants who have mitral stenosis without a diminutive ascending aorta, repair technique for mitral stenosis and simultaneous and staged associated operations have been performed (1, 2). A successful staged biventricular repair in early infancy for a patient who had congenital mitral stenosis with short chordae and hypoplastic left ventricle and the long-term results are described. Informed consent was obtained from the patient.

## CASE PRESENTATION

An 8-day-old female infant with tachypnea and retraction, weighing 2.7 kg, was referred to our institute for surgical repair. On admission, physical examination showed a gallop heart sound at the middle left sternal border, moderate hepatomegaly and systolic pressure gradient of about 10 mmHg between the upper and lower extremities. An echocardiographic diagnosis of severe mitral stenosis, severe tricuspid regurgitation, patent foramen ovale with left-to-right shunting, coarctation of the aorta and severe pulmonary hypertension without a patent ductus arteriosus was made on admission. The mitral valve had two papillary muscles at the normal position and showed ballooning with bilateral commissural fusion

and slight thickening. Annular diameter of the mitral valve was 6 mm (84% of normal) on admission. The patient had concordant atrio-ventricular and ventriculo-arterial connections, normal aortic valve and root (103% and 75% of normal, respectively), and no significant subaortic narrowing on admission. The left ventricular internal dimension in end-diastole from a parasternal short axis view was 6 mm (41% of normal) on admission, and we assessed the ventricle as hypoplastic left ventricle with left ventricular internal dimension in end-diastole of less than 70% of normal values (3) (Fig. 1).

It was difficult to determine on admission whether we should select a biventricular repair or a univentricular repair for the patient as a definitive repair. Because administration of prostaglandin E1 for recanalization of the ductus arteriosus was ineffective, the patient underwent repair of coarctation of the aorta alone as the initial stage at 9 days after birth.

The left ventricle of the patient developed after both the afterload reduction by the coarctation repair and the preload increase by the restrictive foramen ovale. The left ventricular-right ventricular-cross sectional area ratio in end-diastole from a parasternal short axis view changed from 0.23 on admission to 0.59. The patient's symptoms and hemodynamics improved temporarily after repair of coarctation of the aorta but deteriorated gradually due to severe mitral stenosis. From both the viewpoints and the geometrical change of the left ventricle after coarctation repair, we evaluated the left ventricle as being able to function as a systemic ventricle and performed open mitral commissurotomy (OMC) 40 days after birth.

The approach to the mitral valve is shown in Fig. 2. A usual incision was made in the left atrium between the interatrial sulcus and the right superior pulmonary vein and was extended to the connection of both right pulmonary veins. Right transverse atriotomy was made perpendicular to the interatrial sulcus, and the interatrial septum was incised from the interatrial sulcus to the atrial septal defect

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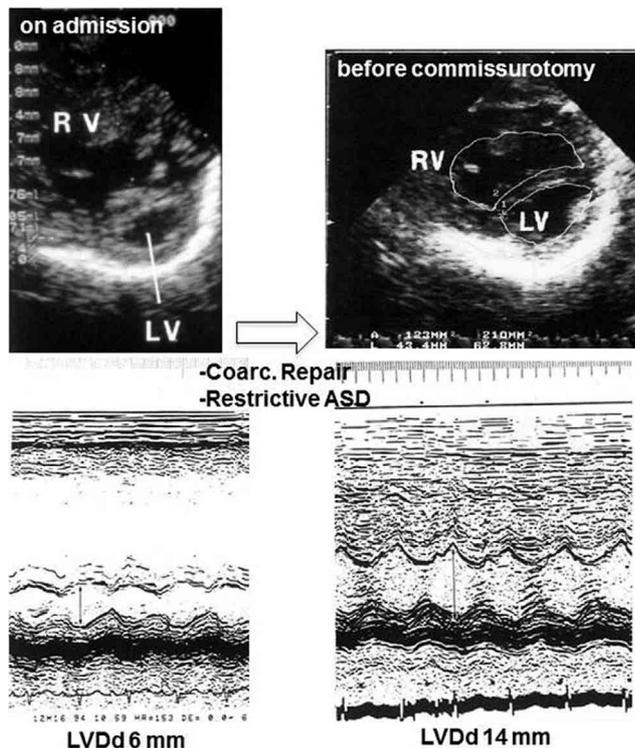


Fig. 1. Left ventricular volume was assessed by left ventricular internal dimension in end-diastole from a parasternal short axis view. The left ventricular internal dimension in end-diastole changed from 6 mm (41% of normal) on admission to 14 mm (95% of normal) before mitral valvotomy. Abbreviations: LVDd=left ventricular internal dimension in end-diastole.

created by the balloon atrial septostomy sequentially (4). As a result, good mitral valve exposure was obtained. Both mitral leaflets were thickened, anterolateral commissure was absent, posteromedial commissure was fused, and chordae attached to the anterolateral papillary muscle were extremely short. Leaflets were incised about 3 mm in the direction of the anterolateral region and 2 mm of posteromedial commissure. Chordotomy and papillotomy were not performed. After the commissurotomy, crystalloid cardioplegic solution was injected into the left ventricular cavity through the mitral valve, and the mitral valve showed mild regurgitation. To minimize the impact of rapid volume load on the hypoplastic left ventricle after the repair, the atrial septal defect was closed with a fenestrated polytetrafluoroethylene patch. The patient was easily weaned from cardiopulmonary bypass. Mean left atrial pressure was about 12 mm Hg in the intensive care unit. The patient was extubated 17 days after OMC. The left ventricle of the patient gradually developed to the left ventricular internal dimension in end-diastole of 16 mm at 5 months after commissurotomy.

The patient is now 18 years old and has been doing well. She has mild thickening of the mitral valve leaflets, trivial mitral regurgitation, peak velocity of E-wave of 183 cm/sec, peak velocity of A-wave of 78 cm/sec, pressure half time of 0.125 sec, and no shunt through the interatrial septum. The left ventricle now has normal growth with a left ventricular internal dimension of 44 mm in diastole (Fig. 3 and 4). The mitral annulus has developed to 23 mm in diameter (Fig. 5).

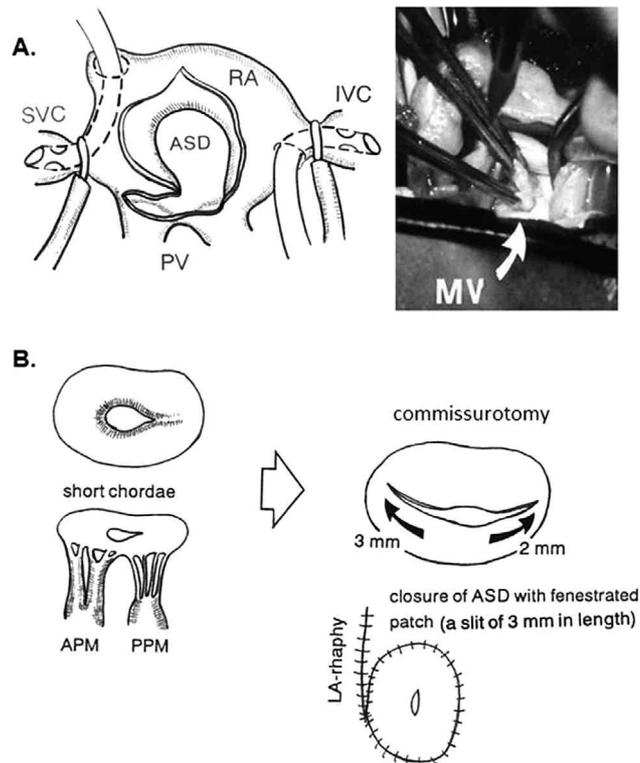


Fig. 2. Approach to the mitral valve and operative findings of the mitral valve and surgical procedure.

A) Approach to the mitral valve. An incision was made in the left atrium between the interatrial sulcus and the right superior pulmonary vein and was extended to the connection of both right pulmonary veins. Right atriotomy was made perpendicular to the interatrial sulcus, and the interatrial septum was incised from the interatrial sulcus to the atrial septal defect, which was created by balloon atrial septostomy 13 days before mitral valvotomy. Good mitral valve exposure was obtained on the right.

B) Operative findings of the mitral valve and surgical procedure. Both mitral leaflets were thickened, anterolateral commissure was absent, posteromedial commissure was fused, and chordae attached to the anterolateral papillary muscle are extremely short. Leaflets were incised about 3 mm in the direction of the anterolateral region and 2 mm of the posteromedial commissure. The atrial septal defect was closed with a polytetrafluoroethylene patch with a slit of 3 mm in length to prevent rapid volume load on the hypoplastic left ventricle. Abbreviations: SVC=superior vena cava; ASD=atrial septal defect; RA=right atrium; IVC=inferior vena cava; PV=pulmonary vein; MV=mitral valve; APM=anterior papillary muscle; PPM=posterior papillary muscle; LA=left atrium.

## DISCUSSION

Mitral stenosis without a diminutive ascending aorta has a broad spectrum because of several other anomalies. The strategy for an infant with congenital mitral stenosis should be determined by three important factors: 1) left ventricular volume, 2) the degree of the systemic outflow tract obstruction e.g.: ventriculo-arterial connection and the sizes of the ascending aorta, aortic annulus and subaortic region, and 3) the type of mitral valve dysfunction. In our experience concerning the second factor, the aortic valve and root with diameters of more than 65% of normal values seemed to maintain systemic circulation without renal failure despite the slow growth of the patient. However, in such patients, the first and third determinants were extremely problematic (5, 6).

Left ventricular volume was assessed by left ventricular internal dimension and left ventricular-right ventricular-cross sectional area ratio in end-diastole from a parasternal short axis view (3).

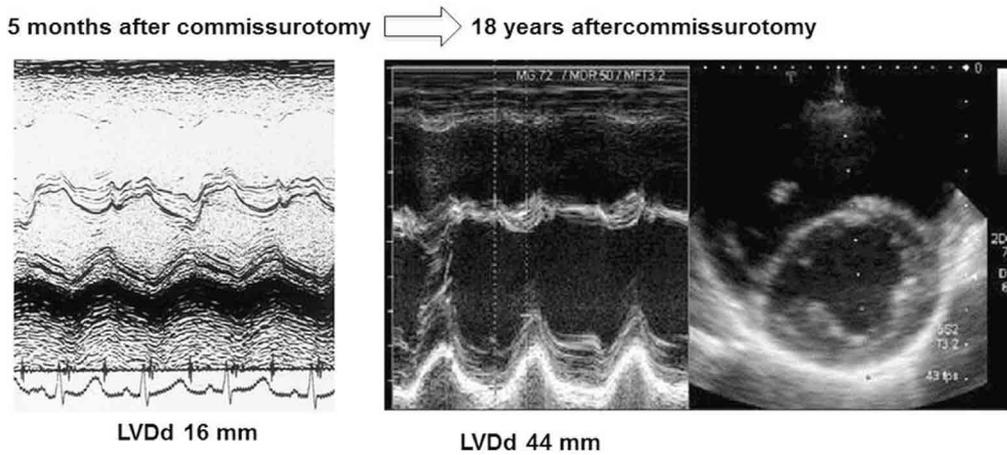


Fig. 3. Growth of the left ventricle. The left ventricular internal dimension in end-diastole changed from 16 mm at 5 months after commissurotomy to 44 mm at 18 years after commissurotomy. Abbreviations : LVDd=left ventricular internal dimension in end-diastole.

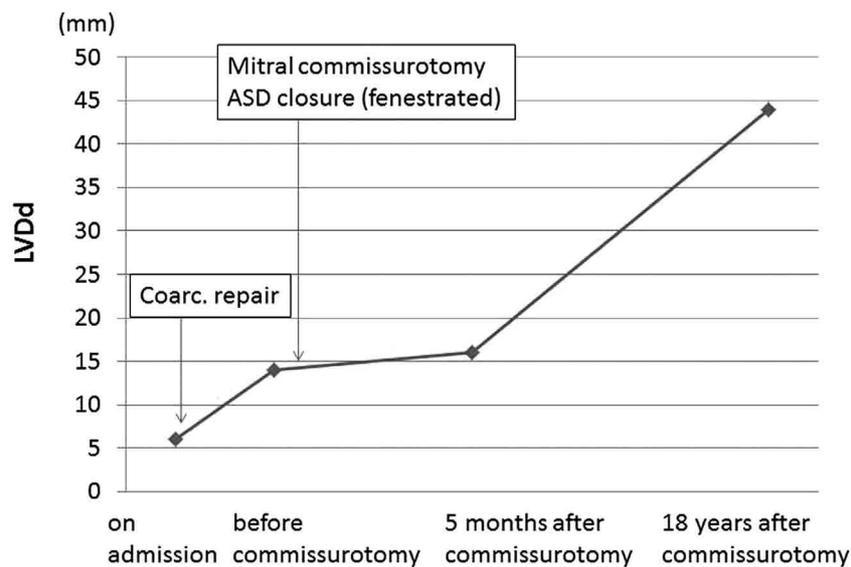


Fig. 4. Growth of the left ventricle as assessed by the left ventricular internal dimension in end-diastole is shown.

However, because of volume and pressure load on the right ventricle derived from the interatrial left-to-right shunt and mitral stenosis, the right ventricle in patients with congenital mitral stenosis is usually enlarged and the left ventricular-right ventricular-cross sectional area ratio is considerably small. To estimate the function of the left ventricle against the volume load after repair of the mitral valve, it is important to evaluate the change in not the left ventricular-right ventricular-cross sectional area ratio but the left ventricular internal dimension in end-diastole. From that viewpoint, we define a hypoplastic left ventricle as one with an internal dimension in end-diastole of less than 70% of normal values. The left ventricle of the patient seemed to have developed to a normalized ventricle after initial palliation. We consider that there were the following important hemodynamic factors that led to the successful biventricular repair in the patient. First, total systemic output was barely supplied through the hypoplastic left ventricle after closure of the ductus arteriosus on admission. Also, spontaneous closure of the foramen ovale following repair of coarctation of the aorta accelerated the progressive left ventricular growth.

Some mitotic activity in cardiac myocytes of a rat was reported to persist during early neonatal life (7). Although it is not definitely proven in humans, myocyte mitotic activity and the potential for hyperplasia allegedly persist for 3 to 6 months after birth (8). According to the clinical experience in a previous study, in a patient with a normal mitral valve who underwent a Norwood-type operation as a first-stage palliation because of subaortic obstruction, biventricular repair using a Rastelli-type repair was successfully performed as a definitive repair (9). We think that early improvement of forward flow through the mitral valve might be effective for growth of a hypoplastic left ventricle. However, the opposite procedure allowing mitral annular and left ventricular growth by closure of the mitral orifice without a prosthetic patch and creation of a muscular ventricular septal defect has been reported (10).

A precise diagnosis of mitral valve malfunction should be obtained preoperatively by echocardiography. In our patient, we selected the modified Brawley's approach in considering the slight subvalvular lesion (4). This technique can be extremely useful in patients with a slight subvalvular lesion. However, valvotomy for

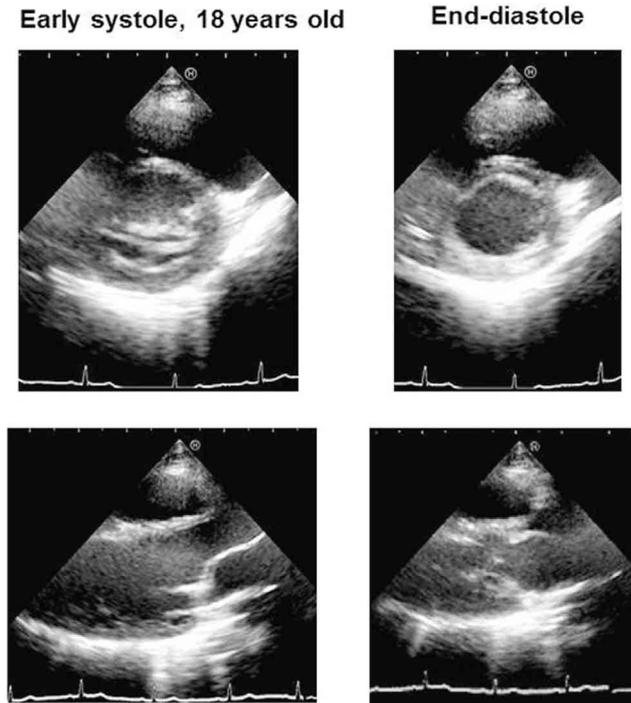


Fig. 5. Mitral valve function at 18 years after commissurotomy. Echocardiography from a parasternal short axis view shows mild mitral valve stenosis and trivial regurgitation with good leaflets motion.

most infants obviously does not result in complete reduction in the mitral valve gradient because the obstruction of flow is mainly due to obliteration of the interchordal spaces and not to commissural fusion. Fortunately our patient had mitral valvular lesion which was thickened but flexible leaflet, and mild subvalvular lesion. So all we have to do was commissurotomy of anterolateral and posteromedial sides. An innovative left ventricular apical approach for repair of subvalvular stenosis was reported by Barbero-Martial and co-workers (1). Although we are afraid of the impairment of left ventricular function despite a small ventriculotomy at the apical region with a length of no more than 2 cm, we would like to perform this technique in selected patients. In any case, mitral valve repair is a useful palliative procedure that may result in symptomatic relief, prevent pulmonary edema, and allow growth of the child. For a patient with aortic stenosis, severe involvement of the valve may require eventual valve replacement.

The smallest available mechanical prosthesis is 16 mm in diameter. A smaller bioprosthesis could be excised from a valved conduit in which a smaller prosthesis is used. Because of a small mitral annulus, supra-annular placement of a prosthetic valve had been used in special cases in infancy. However, a hazardous postoperative management for thrombosis and the necessity of formidable reoperations remained. Generally, palliation as suggested for hypoplastic left heart syndrome, i.e., atrial septectomy, Damus-Kaye-Stansel procedure, and shunt, would abandon the use of the left ventricle as a systemic ventricle and commit the patient to a Fontan procedure as the definitive operation. If the mitral valve in our patient had shown severe regurgitation after valvotomy, the incised line of the mitral valve would have been oversewn and palliation as suggested for hypoplastic left heart syndrome would have been performed because of several anticipated complications after supra-mitral annular placement of a prosthetic valve.

Our patient is now 18 years old and has been doing well. Surgical techniques allowing relief of mitral stenosis have been very well

established during 18 years. This has resulted from advances in the understanding of mitral valve pathology with advanced preoperative imaging using two and three dimensional echocardiography which has provided more precise delineation of valve pathology (11). And some procedures of mitral valve repair are selected depending on the mitral valve pathology that is in supra-annular, valvular and subvalvular lesion (12).

The new guideline for neonates with congenital mitral stenosis still have not established because of its wide spectrum of malformations, a high prevalence of associated lesions and the relatively limited experience in each institution (5, 6). However, suitable initial palliation and additional repairs for patients with congenital mitral stenosis according to the present three important determinants might be able to achieve not only univentricular repair but also biventricular repair as a definitive operation. The increased potentiality of the left ventricle after staged operations in early infancy might be very encouraging to direct cardiac surgeons to biventricular repair.

## CONSENT

Written informed consent was obtained from the patient for publication of this Case Report and any accompanying images.

## COMPETING INTERESTS

The authors declare that they have no competing interests.

## AUTHORS' CONTRIBUTIONS

TK described and designed the article. TK edited the article. HF and HS supervised the edition of the manuscript. Other remaining co-authors collected the data and discussed the content of the manuscript. All authors read and approved the final manuscript.

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## REFERENCES

1. Barbero-Marcial M, Riso A, De Albuquerque AT, Atik E, Jatene A : Left ventricular apical approach for the surgical treatment of congenital mitral stenosis. *J Thorac Cardiovasc Surg* 106 : 105-110, 1993
2. Moore P, Adatia I, Spevak PJ, Keane JF, Perry SB, Castaneda AR, Lock JE : Severe congenital mitral stenosis in infants. *Circulation* 89 : 2099-2106, 1994
3. Imai T, Satomi G, Yasukochi S, Harada Y, Takeuchi T, Gotoh H, Sakamoto T : Normal values for cardiac and great arterial dimensions in premature infants by cross-sectional echocardiography. *Cardiol Young* 5 : 319-325, 1995
4. Brawley RK : Improved exposure of the mitral valve in patients with a small left atrium. *Ann Thorac Surg* 29 : 179-81, 1980
5. Han RK, Gurofsky RC, Lee KJ, Dipchand AI, Williams WG,

- Smallhorn JF, McCrindle BW : Outcome and growth potential of left heart structures after neonatal intervention for aortic valve stenosis. *J Am Coll Cardiol* 50 : 2406-2414, 2007
6. Schwartz ML, Gauvreau K, Geva T : Predictors of outcome of biventricular repair in infants with multiple left heart obstructive lesions. *Circulation* 104 : 682-687, 2001
  7. Anversa P, Olivetti G, Loud AV : Morphometric study of early postnatal development in the left and right ventricular myocardium of the rat. I. Hypertrophy, hyperplasia, and binucleation of myocytes. *Circ Res* 46 : 495-501, 1980
  8. Brown AL : Morphologic factors in cardiac hypertrophy. In : Alpert NR, eds. *Cardiac Hypertrophy*. Academic Press, New York, 1971, pp.11-18
  9. Kanter KR, Miller BE, Cuadrado AG, Vincent RN : Successful application of the Norwood procedure for infants without hypoplastic left heart syndrome. *Ann Thorac Surg* 59 : 301-304, 1995
  10. Westerman GR, VanDevanter SH, Norton JB Jr, Readinger RI : Congenital mitral valve stenosis in infancy : a different approach to a difficult problem. *J Thorac Cardiovasc Surg* 94 : 305-307, 1987
  11. Baird C, Marx GR, Borisuk M, Emani S, del Nido PJ : Review of congenital mitral valve stenosis : Analysis, repair techniques and outcomes. *Cardiovasc Eng Technol* 6 : 167-173, 2015
  12. del Nido PJ, Baird C : Congenital mitral valve stenosis : Anatomic variants and surgical reconstruction. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann* 15 : 69-74, 2012