Surgical management of infants with mitral valve stenosis or atresia without diminutive ascending aorta

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Abstract : The surgical strategy in infants with mitral valve stenosis or atresia without diminutive ascending aorta remains to be established, including the potential for biventricular repair as a definitive operation. Our surgical experience of six infants with mitral valve stenosis (4 patients) or atresia (2 patients) without diminutive ascending aorta was evaluated based on three important factors : left ventricular volume ; the nature of the systemic outflow obstruction ; and the type of mitral valve anomaly. Two patients with systemic outflow tract diameter less than 65% of normal underwent systemic outflow tract reconstruction, and the other patients with outflow tract diameter more than 68% of normal were able to maintain systemic circulation without repair. Only one patient with mitral valve stenosis without left ventricular outflow tract obstruction underwent a successful open mitral valvotomy as a biventricular repair after first-stage palliation. The left ventricle of the other patients did not grow after first-stage palliation. Due to progressive subaortic narrowing, pulmonary artery banding should be avoided in patients with mitral atresia due to absent atrioventricular connection who are future Fontan candidates. Most patients with this lesion can be expected to become candidates for safe Fontan-type repair. J. Med. Invest. 46 : 59-65, 1999

Key words **:** *congenital mitral stenosis, mitral atresia, left ventricular volume, systemic outflow tract obstruction, Fontan-type repair*

INTRODUCTION

Recently, advances in the surgical strategy for Fontan circulation in infants with the classical hypoplastic left heart syndrome characterized by a diminutive ascending aorta have been achieved based on the experience of Norwood, Bove, and their colleagues (1-4). However, the surgical strategy for infants with mitral valve stenosis or atresia without diminutive ascending aorta remains to be established, including the potential for biventricular repair as a definitive operation (5). The strategy should be determined based on three important factors : left ventricular volume, the nature of the systemic outflow obstruction, and the type of mitral valve anomaly. On the basis of this concept, we have retrospectively evaluated our surgical experience.

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PATIENTS AND METHODS

Between December 1986 and December 1994, 6 consecutive infants with mitral valve stenosis or atresia without diminutive ascending aorta and excessive pulmonary blood flow underwent surgical treatment at our institution (Table 1). Echocardiographic examinations were performed on all patients before the first-stage palliative surgery.

1) Left ventricular volume

Left ventricular volume was assessed by measurement of the left ventricular internal dimension and left ventricular-right ventricular cross-sectional area ratio at end-diastole from the parasternal short axis view. The left ventricular internal dimension at end-diastole was compared with normal values (6). For standardization of the latter, the ratio was measured in 7 healthy infants (age : 10-44 days [mean, 31 days], body weight : 2.5-4.4 kg [mean, 3.6 kg]). The normal value of the left ventricular-right ventricular cross-sectional area ratio was 0.96±0.14.

2) Nature of systemic outflow obstruction

Ventriculoarterial connections, the diameters of the aortic annulus and the aortic root, and subaortic narrowing were evaluated echocardiographically. The diameters of the aortic annulus and the aortic root were obtained at end-systole from the parasternal long axis view. The results were compared with normal values (6).

3) Type of mitral valve anomaly

The type of mitral valve anomaly was assessed based on the number and position of papillary muscles. The diameter of the mitral annulus was measured at end-diastole from the apical four-chamber view. The results were compared with normal values (6).

RESULTS

1) Left ventricular volume

All patients except for patient 4 could be evaluated before the first-stage of palliation (Table 2). The left ventricular internal dimension at end-diastole ranged from 4 to 12 mm (24% to 77% of normal). The left ventricular-right ventricular cross-sectional area ratio ranged from 0.05 to 0.42. The left ventricle of all patients except for patient 5 (left ventricular internal dimension=77% of normal value, left ventricular-right ventricular cross-sectional area ratio=0.42) was diagnosed as hypoplastic. We defined a hypoplastic left ventricle as one with a left ventricular internal dimension at end-diastole less than 70% of the normal value (6).

2) Nature of systemic outflow obstruction

A concordant ventriculoarterial connection was found in 5 patients, and a transposed connection in one patient. The diameter of the aortic annulus at end-systole ranged from 4 to 7 mm (59% to 103% of normal) (Table 3). The aortic valvular movements of all patients were normal. The diameter of the aortic root at end-systole ranged from 3 to 6.6 mm (43% to 82% of normal). No significant subaortic narrowing before the first-stage palliative surgery was seen in any patient. Two patients were diagnosed with aortic stenosis of annular hypoplasia and the remaining 4 patients had normal systemic outflow tracts (we defined stenosis of the systemic outflow tract as a systemic outflow tract diameter at end-systole of less than 70% of normal values).

3) Type of mitral valve anomaly

Mitral valve atresia of an absent atrioventricular connection was detected in 2 patients (Table 4). The diameter of the mitral annulus in the other 4

Table 1. Summary of patients with mitral stenosis or atresia without diminutive ascending aorta.

Patient no.	Age (days)	Weight (kg)	Diagnosis
1	14	3.0	[S, D, D], MS, AS, hypoplastic LV, CoA
2	72	3.4	[S, D, D], MS, AS, hypoplastic LV, CoA
3	9	2.5	[S, D, D], MS, hypoplastic LV, CoA
4	117	4.2	[S, D, DR], MS, VSD, hypoplastic LV
5	18	2.9	[S, D, D], MA, muscular VSD, CoA
6	9	3.5	[S, D, D], MA, hypoplastic LV, muscular VSD, CoA

Abbreviations : [S, D, D]=[situs solitus, concordant loop, normal arterial relations] ; MS=mitral stenosis ; AS=aortic atenosis ; LV=left ventricle ; CoA=coarctation of the aorta ; MA=mitral atresia ; VSD=ventricular septal defect ; [S, D, DR]=[S, D, double outlet right ventricle]

Table 2. Left ventricular internal dimension and left ventricular-right ventricular-cross sectional area ratio at end-diastole.

Patient no.	LVDd mm	LVDd (% of normal)	Ratio
1	9	(57)	0.35
2	5	(30)	0.18
3	6	(41)	0.23
4	/	/	/
5	12	(77)	0.42
6	4	(24)	0.05

Only patient 4 could not be evaluated because of no precise short axis view before the first-stage of palliation. Abbreviations : LVDd=left ventricular internal dimension at end-diastole ; LVDd=9.369+0.002161×weight : (Weight-dependent equation [6])

Table 3. Nature of systemic outflow obstruction.

Patient no.	Aortic valve annular diameter		Aortic root diameter		Type of coarctation	Surgical procedure at first-stage of palliation
	mm	(% of normal)	mm	(% of normal)		
1	4.0	(59)	3.0	(43)	Preductal	PA to des. Ao conduit and PA banding
2	4.0	(57)	4.0	(55)	Juxtaductal	Norwood
3	6.3	(103)	4.9	(75)	Preductal	SFA
4	7.0	(90)	6.6	(82)	None	None
5	6.0	(92)	5.0	(72)	Preductal	SFA (first) Stansel (second)
6	6.0	(85)	5.0	(68)	Preductal	SFA (first) Stansel (second)

Abbreviations : PA=pulmonary artery ; des. Ao=descending aorta ; SFA : subclavian flap aortoplasty : annular diameter of aortic valve=3.648+0.000982×weight ; diameter of aortic root=4.221+0.000915×weight : (Weight-dependent equation [6])

patients ranged from 6 to 10 mm (56% to 85% of normal). Patients 1, 2, and 3 had two papillary muscles in the normal position, and patient 4 had one papillary muscle. The mitral valve of patients 1 and 2 showed normal valvular movement. Patients 1 and 2 were diagnosed with mitral valve stenosis of annular hypoplasia (we defined annular hypoplasia as a diameter of the mitral annulus at end-diastole of less than 70% of normal values). The mitral valve of patient 3 showed ballooning of leaflets despite a bilateral commissural fusion and slight thickening. Patient 3 was diagnosed with mitral stenosis with shortened chordae. Patient 4 was diagnosed with mitral valve stenosis with a parachute valve.

Table 4. Type of mitral valve anomaly.

Patient no.	Mitral valve			Surgical procedure
	Type of anomaly	Annular diameter		
		mm	(% of normal)	
1	Annular hypoplasia	8.0	(78)	None
2	Annular hypoplasia	6.0	(56)	None
3	Short chordae	8.0	(84)	Open mitral valvotomy
4	Parachute valve	10.0	(85)	None
5	Atresia	/	/	None
6	Atresia	/	/	None

Annular diameter of mitral valve=6.390+0.001277×weight : (Weight-dependent equation [6])

4) First-stage palliation

First-stage palliative surgery was performed between 9 and 117 days (mean, 40 days) after birth. Body weight at first-stage palliative surgery ranged from 2.5 to 4.2 kg (mean, 3.3 kg).

In the patients with aortic annulus or aortic root diameter less than 65% of the normal value, reconstruction of the systemic outflow tract was indicated based on our experience in the surgical treatment of infants with interrupted aortic arch, small ascending aorta, and ventricular septal defect separate from future definitive repair (Fig.1). Patients 1 and 2, who had aortic annulus and aortic root diameters less than 65% of normal values underwent placement of a main pulmonary artery to the descending aorta conduit (in the earliest patient in our series) or a modified Norwood operation. An aortic annulus and aortic root with a diameter greater than 68% of the normal value could maintain the systemic circulation without repair in the early postoperative period.

It was difficult to estimate on admission whether patient 3 would be a candidate for biventricular or Fontan-type repair, but the patient eventually underwent only repair of coarctation of the aorta. Patient 4,with a parachute mitral valve, double-outlet right ventricle, small muscular ventricular septal defect, and apparently significantly hypoplastic left ventricle, was judged to be a candidate for Fontan-type repair and underwent surgical atrial septal defect creation and pulmonary artery banding.

Patients 5 and 6, with mitral atresia of the absent atrioventricular connection, were originally diagnosed as candidates for Fontan-type repair and underwent repair of coarctation of the aorta, balloon atrial septostomy, and pulmonary artery banding.

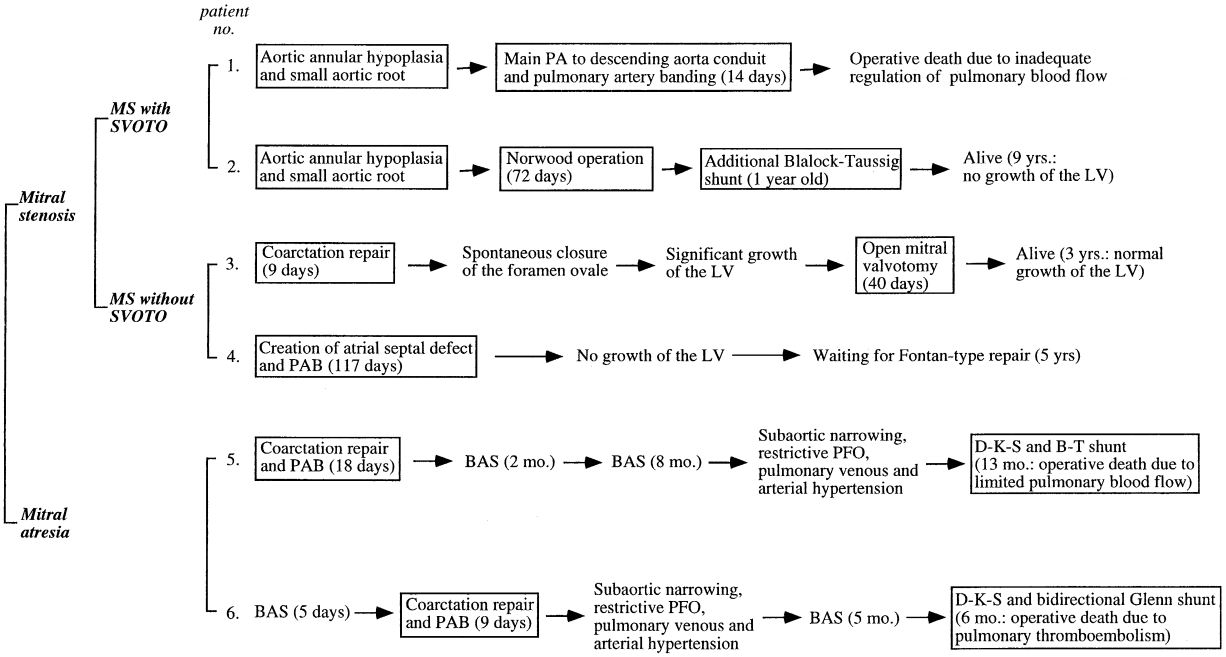

Fig. 1. Staged repairs and outcomes. Abbreviations : MS=mitral stenosis ; SVOTO=systemic ventricular outflow tract obstruction ; MA=mitral atresia ; PA=pulmonary artery ; PAB=pulmonary artery banding ; B-T shunt=Blalock-Taussig shunt ; LV=left ventricle ; BAS=balloon atrioseptostomy ; D-K-S=Damus-Kaye-Stansel ; PFO=patent foramen ovale.

5) Outcomes

After first-stage palliation the left ventricle did not grow due to insufficient preload to the left ventricle in all patients except for patient 3.

Patient 1 died because of inadequate regulation of the pulmonary blood flow after first-stage palliative surgery resulting in postoperative congestive heart failure. Patient 2, who survived the Norwood operation, is now 9 years old, and shows good hemodynamics. However, he did not undergo a Fontan-type operation because of other physical anomalies.

Patient 3 improved temporarily after repair of coarctation of the aorta but deteriorated gradually due to spontaneous closure of the foramen ovale. However, the left ventricle developed because of reduction of the afterload following repair of coarctation of the aorta and increased preload to the left ventricle following spontaneous closure of the foramen ovale. The left ventricular internal dimension at end-diastole in patient 3 increased from 6 mm (41% of normal) on admission to 14 mm (95% of normal) before mitral valvotomy (Fig. 2). The left ventricular-right ventricular cross-sectional area ratio in patient 3 also increased from 0.23 on admission to 0.59 before mitral valvotomy (Fig. 3). At that time we assessed the left ventricle as able to function as a systemic ventricle and performed a successful open mitral valvotomy 40 days after birth. However, cardiac catheterization at 26 months old showed residual mitral valve stenosis of the subvalvular lesion and moderate pulmonary artery hypertension.

Patient 4 had a good postoperative course. Cardiac catheterization at 3 years of age revealed low pulmonary vascular resistance by appropriate control of the pulmonary blood flow and normal right ventricular function. This patient is waiting for the Fontan-type operation.

Patients 5 and 6 required subsequent conversion to a Damus-Kaye-Stansel anastomosis, shunt operation, and creation of atrial septal defect due to progressive subaortic narrowing resulting from pulmonary artery banding in the first-stage palliative surgery and restrictive foramen ovale. However, patient 5 died on the first postoperative day because of limited pulmonary blood flow resulting from pulmonary venous and artery hypertension due to the delayed second-stage palliation (at 13 months of age). Patient 6 underwent a 4-mm modified Blalock-Taussig shunt between an innominate artery and the right pulmonary artery which caused excessive pulmonary blood flow and reduced systemic perfusion (at 6 months of age). The shunt was replaced with a bidirectional Glenn shunt. However, he died because of the pulmonary thromboembolism 6 days after the operation.

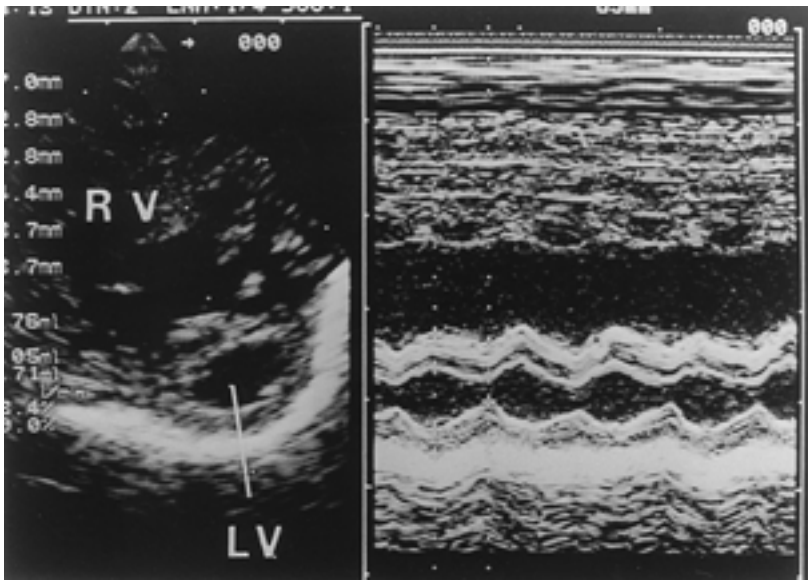

Fig. 2. The left ventricular internal dimension at end-diastole of patient 3 was 6 mm at admission, and the left ventricular-right ventricular-cross sectional area ratio was 0.23.

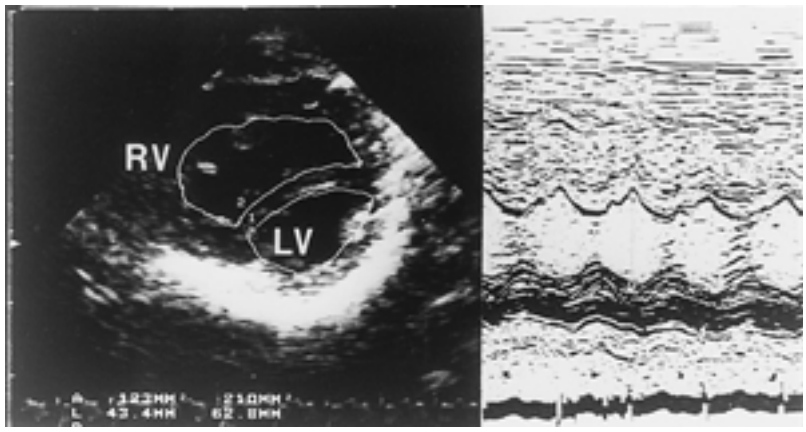

Fig. 3. The left ventricular internal dimension at end-diastole of patient 3 was 14 mm before open mitral valvotomy, and the left ventricular-right ventricular cross-sectional area ratio was 0.59. The left ventricle developed after reduction of the afterload following repair of coarctation of the aorta and increased preload to the left ventricle following the spontaneous closure of the foramen ovale. Abbreviations : RV=right ventricle ; LV=left ventricle.

DISCUSSION

Although the surgical strategy for patients with mitral valve stenosis or atresia without diminutive ascending aorta should be determined based on three factors, it seems unusual for them to become candidates for future biventricular repair. Therefore, most patients are expected to be candidates for safe Fontan-type repair.

1) Fontan-type repair

A hypoplastic aortic valve is defined as having a diameter of less than 5 or 6 mm by echocardiographic assessment (5, 7), and a hypoplastic aortic root as having a diameter of less than 6 mm by the same method (8). In our patient series, a systemic outflow tract with a diameter greater than 68% of normal was capable of maintaining the systemic circulation without repair, with sufficient feeding and weight gain. We experienced that one patient from another series with an interrupted aortic arch, ventricular septal defect, and aortic root with a diameter of 64% of normal values showed low cardiac output syndrome after complete repair and died. Generally speaking, in infants with body weight of 3 kg, an aortic annulus diameter of 4.3 mm and aortic root diameter of 4.5 mm are 65% of normal values and correspond to about 42% of the normal cross-sectional area of the aortic annulus and aortic root. We think that an aortic annulus or aortic root with a diameter of less than 65% of normal might be a reasonable criterion for performing the Norwood procedure as a first-stage palliative surgery.

In patients with mitral valve stenosis after the Norwood-type operation, the left ventricle appears not to grow due to insufficient preload. Therefore, to perform an optimal Fontan-type operation by achieving low pulmonary vascular resistance and prevention of myocardial hypertrophy, it is important to proceed swiftly with a second-stage hemi-Fontan procedure (2).

An unusual patient with mitral atresia of the imperforate membrane might be better corrected by resection of the imperforate membrane and the insertion of a prosthetic valve in the mitral position. However, in most patients with mitral valve atresia of the absent atrioventricular connection, normally related great arteries, and muscular ventricular septal defect, the left ventricle appears to function as the outlet chamber of the main right ventricle through a muscular ventricular septal defect, and not to grow even if it is not hypoplastic. We think that patients with such complex lesions should be treated from the start in anticipation of a future Fontan-type repair. Therefore, pulmonary artery banding, which causes subaortic narrowing and reduction of myocardial compliance, should be avoided as first-stage palliation for patients with

such complex lesions (9, 10). A palliative arterial switch operation (11, 12) or modified Norwood/Damus-Kaye-Stansel procedure are advocated as first-stage palliative surgery for neonates with such complex lesions, which have the potential to result in systemic ventricular outflow tract obstruction (7, 13-16). If pulmonary artery banding is performed reluctantly, such detrimental long-term effects should be monitored closely and be relieved by the above procedures or hemi-Fontan operation/bidirectional Glenn shunt as early as possible.

2) Biventricular repair

Only a patient with mitral valve stenosis without the need for systemic outflow tract reconstruction should undergo a biventricular repair attempt.

Some mitotic activity in rat cardiac myocytes was reported to persist during early neonatal life (17). Although this has not been definitely proven in humans, myocyte mitotic activity and the potential for hyperplasia allegedly persist for 3 to 6 months after birth (18). According to previous reports, only patients with a normal mitral valve who underwent a Norwood-type operation as a first-stage palliation due to subaortic obstruction have successfully undergone a Rastelli-type biventricular procedure as a definitive repair (5). Early improvement of forward flow through the mitral valve might result in hypoplastic left ventricle growth, as seen in the left ventricle of our patient 3.

However, it is difficult to estimate the future growth potential of the left ventricle associated with congenital mitral valve stenosis before first-stage palliative surgery in neonates. Due to the volume and pressure load on the right ventricle derived from interatrial left-to-right shunt and mitral valve stenosis, the right ventricle is usually enlarged and the left ventricular-right ventricular cross-sectional area ratio is reduced. To estimate the function of the left ventricle against the volume load after repair of the mitral valve, it is important to closely follow the changes in the left ventricular internal dimension in end-diastole.

Mitral valve repair is a useful palliative procedure which may provide symptomatic relief and allow growth of the child in the current absence of a satisfactory prosthetic valve substitute. However, valvotomy for most infants obviously does not result in complete reduction of the mitral valve gradient, because the obstruction of flow results mainly not from commissural fusion but from obliteration of the interchordal spaces. An innovative left ventricular apical approach for repair of subvalvular stenosis was reported by Barbero-Marcial and his coleagues (19). Although we used a modified Brawley approach (20) because of the slight subvalvular lesion in patient 3, additional papillotomy would be needed to remove the residual subvalvular stenosis and to restore adequate leaflet motion.

Our experience does not establish a definitive surgical strategy for treatment of these lesions because the number of patients was too few. However, to achieve either Fontan-type repair for most patients or biventricular repair for rare patients as a definitive operation, suitable first-stage palliation and additional interventions in early infancy should be performed based on changes in the three determinants of left ventricular volume, nature of the systemic outflow obstruction, and type of mitral valve anomaly.

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