Outcomes of Reparative and Transplantation Strategies for Multilevel Left Heart Obstructions With Mitral Stenosis

Sunil P. Malhotra, MD, François Lacour-Gayet, MD, David N. Campbell, MD, Shelley Miyamoto, MD, David R. Clarke, MD, Marshall L. Dines, BA, D. Dunbar Ivy, MD, and Max B. Mitchell, MD

The Children's Hospital Heart Institute, Children's Hospital, and University of Colorado at Denver Health Sciences Center, Denver, Colorado

Background. **Conventional management for multilevel left heart obstructions and mitral stenosis (Shone's complex) involves multiple operations that carry additive risks. This study reviews our experience with reconstructive and transplantation approaches for Shone's complex.**

Methods. **Between 1987 and 2007, 43 patients with mitral stenosis and one or more left-sided obstructions were identified: supramitral ring (n = 13), subaortic stenosis (n = 25), aortic stenosis (n = 24), hypoplastic arch (n = 20), and coarctation (n = 38). Thirty patients underwent a staged reparative approach, including 27 mitral and 51 left ventricular outflow tract operations. Thirteen patients were referred for transplantation. Patients with severe hypoplasia of the left ventricle were excluded.**

Results. **There was one in-hospital death (2.5%) and six late deaths (14.2%). Actuarial 5- and 10-year survival for staged surgical and transplantation was 88% vs 61.3% and 83.1% vs 61.3% ($p = 0.035$). At a mean follow-up of 7.9 years, freedom from mitral reoperation was 83.3% and freedom from reoperation for subaortic stenosis was 78.0%. Wait-list mortality was 13.3% (2 of 13). Wait-list time exceeding 90 days was an incremental risk factor for death after transplantation ($p = 0.005$).**

Conclusions. **Despite the challenges of a reparative strategy for Shone's complex, favorable survival and durability outcomes can be expected. Heart transplantation, although avoiding the pitfalls of staged repair, confers increased risks from ongoing physiologic derangements due to uncorrected left heart inflow and outflow obstructions during the wait for donor heart availability.**

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Surgical management of congenital multilevel left-sided heart obstructive lesions with mitral involvement poses a formidable challenge. In 1963 Shone and colleagues [1] described a complex of lesions causing obstruction to left-sided inflow and outflow, including supramitral ring, parachute deformity of the mitral valve, subaortic obstruction, and aortic coarctation. In reality, these patients exist along a spectrum in which the presence and severity of these lesions varies. Not infrequently, other cardiovascular defects, including valvar aortic stenosis, aortic arch hypoplasia, and borderline left ventricular size contribute to the added complexity of this patient population.

Multiple operations during infancy and childhood are often necessary to address the left ventricular inflow and outflow lesions. Moreover, because of the complex pathology affecting the mitral subvalvar apparatus and hypoplasia of the mitral annulus, mitral valve-sparing procedures are often not possible or durable. Options for pediatric valve replacement are usually limited to mechanical prostheses; thus, the inherent risks of anticoagulation and thromboembolic complications must be accepted.

With the prospect of multiple operations on the inlet and outlet of the left ventricle, and the potential need for prosthetic valve replacement, cardiac transplantation may be an attractive option in selected patients with Shone's complex. Indeed, the presence of Shone's complex has been demonstrated to be a predictor of death for mitral valve replacement in children aged younger than 5 years [2]. Transplantation, however, is not without major drawbacks: Lengthy waiting times for an organ, acute and chronic rejection, and immunosuppression-related complications are variables that contribute to important morbidity and graft failure.

To gain an improved understanding of the optimal management strategies for patients with Shone's complex, we reviewed our experience with this patient population managed either by a staged repair (SR) or transplantation (TX) strategy. The review of the SR cohort focused on the cumulative effect on patient outcomes of both the multiple operations required and the morbidity

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Address correspondence to Dr Malhotra, Falk CVRB, Department of Cardiothoracic Surgery, Stanford University, 300 Pasteur Dr, Stanford, CA 94305; e-mail: spm@stanford.edu.

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Table 1. Spectrum of Left-Sided Heart Inflow Lesions

Pathology	SR	TX	Total
Structurally abnormal MV	30	13	43
Predominant mitral stenosis[a]	26	11	37
Predominant mitral regurgitation	4	2	6
Hammock or arcade MV	9	5	14
Parachute mitral valve	12	3	15
Supraannular mitral ring	11	2	13

[a] Gradient > 6 mm Hg.

MV = mitral valve; SR = staged repair; TX = transplantation.

associated with prosthetic valve complications. For the TX group, the morbidity of waiting time for organ availability and transplant-related complications were specifically analyzed to accurately assess outcomes of transplantation for Shone's complex.

Patients and Methods

During a 20-year period, between June 1987 and May 2007, 43 consecutive patients presented to Children's Hospital, Denver, with mitral stenosis and multiple levels of left heart obstruction. Patient records, including operative reports, diagnostic reports, inpatient records, and outpatient clinic visit notes were reviewed in accordance with an approved protocol from the Colorado Multiple Institutional Review Board. Individual patient or parental consent was waived because of the retrospective nature of the study. Patients requiring a single-ventricle management pathway were excluded. Echocardiography was used to characterize the cardiovascular anatomy in all patients. In patients who underwent transplantation, additional anatomic data were obtained from pathologic examination of the explanted heart.

Patient Groups

The SR group comprised 30 patients (17 boys, 13 girls) with mitral stenosis and multiple left-sided obstructive lesions who underwent a staged reparative approach. In 27 patients (90%), the initial operation was performed when they were aged younger than 1 year.

The TX group comprised 13 patients (7 boys, 6 girls) with Shone's complex who were listed for transplantation. The median age at first operation was 22 days (range, 2 to 76 days). Of these, 11 (85%) survived to transplantation. Median age at transplantation was 5 months (range, 1 to 20 months).

Table 2. Distribution of Left-Sided Outflow Obstructive Lesions

Diagnosis	SR	TX
Coarctation	26	12
Subaortic obstruction	17	8
Valvar aortic stenosis	16	8
Hypoplastic aortic arch	14	6
Supravalvar aortic stenosis	2	1

SR = staged repair; TX = transplantation.

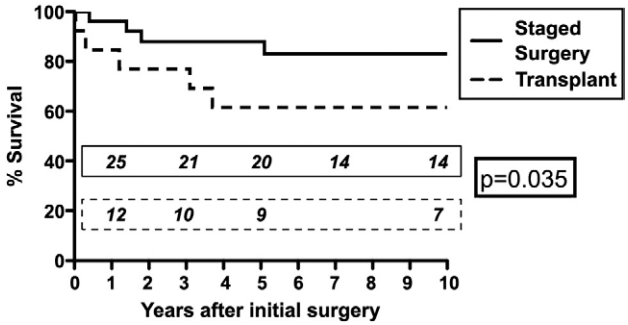

Fig 1. Actuarial survival at 1, 5, and 10 years is shown for staged repair (solid line) and transplantation (dashed line) approaches. For the transplant group, survival data are based on patients who underwent heart transplantation. Patients at risk for each group are listed.

Mitral Valve Morphology

The specific mitral lesions are summarized in Table 1. All patients had structural abnormalities of the mitral valve. A mean gradient across the left ventricular inflow greater than 6 mm Hg was present in 37 children (86%). Parachute deformity of the mitral subvalvar apparatus was present in 15 (35%), and a supravalvar mitral ring was present in 13 (30%). Severe deformities of the subvalvar apparatus such as the hammock or arcade malformations, as described by Uva and colleagues [3] with a combination of papillary muscle hypertrophy and foreshortened chordae, were found in 14 patients (33%).

Left-Sided Outflow Obstructions

Left-sided heart obstructive lesions are reported in Table 2. Coarctation of the aorta was the most prevalent left-sided obstructive lesion and was present in 38 of 43 patients (88%). Hypoplasia of the aortic arch was present in 20 (47%) with coarctation. Significant subaortic stenosis affected 25 patients (58%). Important valvar aortic stenosis developed in 24 patients (56%), supravalvar stenosis occurred in 3 (7%), and an associated ventricular septal defect was present in 20 (47%).

Surgical Procedures

In the SR group, 30 patients underwent a total of 72 operations. There were 31 left ventricular inflow procedures in 22 patients. Twenty-seven patients underwent 56 procedures to address outflow obstructive lesions.

The median age at first operation was 6.5 days (range, 1 day to 1.6 years). Coarctation repair was the initial operative procedure in 24 of 30 patients (80%). Aortic arch hypoplasia was addressed in 5 patients undergoing coarctation repair. Other procedures performed at time of coarctation repair included pulmonary artery banding (PAB) in 4 and aortic valvotomy in 2. Other procedures performed at the initial presentation include subaortic