A standardized repair-oriented strategy for mitral insufficiency in infants and children: Midterm functional outcomes and predictors of adverse events

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Objective: Surgical management of mitral regurgitation (MR) in children remains a challenge because of the heterogeneity of the anatomy, growth potential, and necessity to avoid valve replacement. Our objective was to assess the functional outcomes and prognostic factors of a standardized strategy of mitral valve (MV) repair for children with MR.

Methods: Consecutive patients aged <18 years who had undergone surgery for severe MR from 2001 to 2012 were studied retrospectively. The standardized repair strategy mainly included leaflet debridement, annuloplasty, and leaflet augmentation. Multivariable risk analyses for recurrent MR (grade > II), transmitral mean echocardiographic gradient >5 mm Hg, MV reoperation, replacement, and mortality were performed.

Results: A total of 106 patients were included (median age, 5.1 years; range, 11 days to 18 years). The mean follow-up period was 3.9 ± 3.2 years (range, 2 months to 11 years). The proportion of congenital and left heart obstruction-related (left ventricular outflow tract obstruction) etiology was 49% (n = 52) and 11% (n = 12), respectively. MV repair was performed primarily in 97% of the patients. The mortality, reoperation, replacement, and MR rate at the last follow-up visit was 4.5% (n = 5), 23% (n = 24), 5.5% (n = 6), and 17% (n = 18), respectively. Actuarial survival was $93\% \pm 2\%$ at 10 years. Freedom from MV replacement was $95\% \pm 2\%$ and $86\% \pm 7\%$ at 5 and 15 years, respectively. Native valve preservation was obtained in 85% of the infants and 94% beyond infancy. Independent predictors of recurrent MR, MV reoperation, and replacement included left ventricular outflow tract obstruction etiology (hazard ratio, 45; P = .004), associated preoperative mitral stenosis (hazard ratio, 21; P = .03), and young age (hazard ratio, 1.2; P = .04).

Conclusions: A standardized and reproducible MV repair strategy can achieve satisfactory functional results in infants and children with severe MR, allowing native valve preservation. The left ventricular outflow tract obstruction-related etiology was the main independent predictor of recurrent MR, MV reoperation, and MV replacement. (J Thorac Cardiovasc Surg 2014;148:1459-66)

Surgical management of mitral regurgitation (MR) in the pediatric population remains a challenge because of the heterogeneity of the lesions, requirement for growth potential, and necessity of avoiding mitral valve (MV) replacement. Various nonstandardized repair techniques have been reported in children with congenital MV disease, demonstrating the benefits of MV repair versus replacement.¹⁻¹¹ Nevertheless, these series have reported variable outcomes, related to the small size and heterogeneity of

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Copyright @ 2014 by The American Association for Thoracic Surgery http://dx.doi.org/10.1016/j.jtcvs.2014.02.057 the studied population and the variability of the surgical techniques performed at each center. Moreover, the predictors of adverse events in this surgically challenging pediatric population are still lacking in the published data, essentially because of the relative paucity of this condition and the subsequent limited number of patients previously reported in studies.

Our unit implemented a "nothing else but repair" strategy in which a standardized procedure on the MV was performed in all cases, ideally restoring durable, functional anatomy. The present study aimed at describing the features of this repair-oriented strategy, reporting the midterm functional outcomes, and determining the predictors of MV-related adverse events.

METHODS

Patients

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The echocardiograms, medical records, and operative reports for all patients aged <18 years who had been referred to our institution for surgical treatment of severe mitral insufficiency (MI) from 2001 to 2012 were retrospectively reviewed. A total of 106 patients were included. The median age at surgery was 5.1 years (range, 11 days to 18 years).

| Abbreviations and Acronyms | |
|--|------------------------|
| CI | = confidence interval |
| HR | = hazard ratio |
| LV | = left ventricular |
| LVOTO = left ventricular outflow tract obstruction | |
| MI | = mitral insufficiency |
| MR | = mitral regurgitation |
| MV | = mitral valve |
| | |

Of the 106 patients, 30 (25%) were <1 year and 41 (38.6%) were <2 years old (Figure 1). All patients presented with symptoms, including failure to thrive and intolerance to feeding in infants and young children, and New York Heart Association class III-IV in the older patients. The 4 most frequent etiologies of MR were congenital (n = 52; 49%), including the presence of a mitral leaflet cleft (n = 15; 14%), rheumatic (n = 13; 12%), related to left ventricular (LV) outflow tract obstruction (LVOTO; n = 12; 11%), and ischemic (n = 11; 10%; in 10, an anomalous left coronary artery arising from the pulmonary artery, and in 1, atresia of the left coronary ostium). Twelve patients (11%) had associated mitral stenosis, defined by a mean echocardiographic gradient of >10 mm Hg. Most patients had an associated lesion in addition to MR. Patients presenting with repaired atrioventricular septal defect were excluded. The patient and mitral valve anatomic and functional characteristics are listed in Table 1 and the functional classification in Table 2.

Echocardiographic Evaluation

All 106 patients had undergone an echocardiographic examination before surgery, during surgery (control transesophageal echocardiography), at discharge from the hospital, and annually during the follow-up visits. Preoperative MV function was assessed by transthoracic echocardiography according to the European Association of Echocardiography guidelines.¹² In brief, the echocardiographic examinations were used to assess the MV anatomy and function (ie, annulus, leaflet, subvalvar apparatus), quantified the MR (ie, size of the left cavities using M mode, reverse flow in the pulmonary veins using pulsed Doppler, and proximal isovelocity surface area), and evaluate the consequences of MR (ie, LV ejection fraction using Simpson's method, systolic pulmonary artery pressure calculated from the tricuspid regurgitation flow velocity, mean pulmonary artery pressure calculated from the pulmonary regurgitation flow velocity). A multimodality approach combining all these parameters was used to separate MR into 4 classes from I to IV. Preoperatively, 54 patients (51%) had moderate to severe MR (grade III) and 44 (42%) had severe (grade IV) MR.

Surgical Technique

After median sternotomy and pericardotomy, cardiopulmonary bypass was established using standard aortic and bicaval cannulation, antegrade intermittent warm blood cardioplegia, and normothermia. A patch of pericardium was prepared free from adipose and extrapleural tissue and treated in glutaraldehyde for 7 minutes in the primary procedures.

After dissection of Sondergaard's groove, a left atriotomy was performed. Annular stay stitches were placed on both trigones and posterior part of the annulus to expose the MV. Direct and systematic inspection of the MV apparatus (ie, papillary muscles, chordae, leaflets, and annulus) using nerve hooks allowed us to confirm the echocardiographic findings and completely assess the valve pathologic condition and MR mechanism and etiology.

A 3-step standardized repair strategy was then applied, leading to surgical interventions according to the MR mechanism and etiology and

MV anatomy. The final objective of this standardized repair strategy was to restore the functional MV anatomy to as close as possible to the normal functional anatomy.

Step A. The first step consisted of subvalvular apparatus rehabilitation, mainly to increase the leaflet mobility through extensive leaflet debridement. This step, involving both leaflets and using nerve hooks to expose the lesions, required

- 1. Searching for and cutting all useless and restrictive secondary chordea tendinae
- 2. Splitting the papillary muscles in the case of shortened leaflets or "papillary muscle–leaflet fusion"
- 3. In the case of leaflet prolapse, shortening the papillary muscle, directly or by intraventricular repositioning, chordae transfer, and/or the use of artificial PTFE chordae or resection of redundant anterior leaflet tissue associated with repositioning of a native chordae coming from the anterior or posterior leaflet

Twenty patients (19%) benefitted from the latter technique. The treatment of posterior leaflet prolapse was performed by quadrangular resection of the posterior leaflet associated with sliding plasty of each hemileaflet (n = 9; 8%). Step A was required in more than two thirds of the patients (n = 75; 71%).

Step B. Step B consisted of plication and reinforcement of the mitral annulus. The 3 tools were used to meet this aim were as follows:

- 1. Commissural and posterior annuloplasty stitches, in younger patients and larger patients, in the case of insignificant annular dilatation. This procedure was used in 15 patients (14%).
- 2. Posterior annuloplasty, using a Gore-Tex Accuseal interrupted strip (W. L. Gore & Associates, Inc, Flagstaff, Ariz), in the case of severe annular dilatation in growing children. This strip should be sutured to the posterior annulus using mattress sutures, extending a few millimeters over each trigon. Stitches placed on the annulus will unequally plicate different segments of the posterior annulus, especially at the level of each trigone and the level of P3 in LVOTO-related and ischemic MR. The Accuseal strip should be cut in a semicircular fashion, with a length corresponding to approximately two thirds of the Rowlatt table-based indexed annulus size. Additional resorbable polydioxanone annuloplasty sutures to allow further growth of the annulus at a distance from the surgery has also been occasionally used. Posterior annuloplasty was used in 50 patients (47%).
- 3. Complete annuloplasty, using a prosthetic ring, in the case of severe dilatation when a \geq 28 mm prosthetic ring was suitable. The ring should

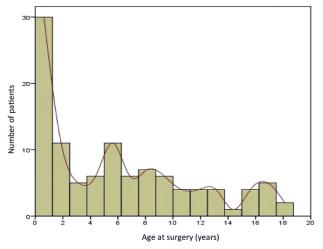


FIGURE 1. Age at surgery stratified by the number of patients.

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