Surgical treatment of congenital mitral valve disease: Midterm results of a repair-oriented policy

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Objective: Management of congenital mitral valve disease is challenging because of a wide morphologic spectrum, frequent associated lesions, and small patient size. We evaluated the results of a repair-oriented policy.

Methods: All consecutive patients with congenital mitral valve disease who underwent surgery between 1996 and 2006 were studied retrospectively. Patients with atrioventricular canal, atrioventricular discordance, or ischemic regurgitation were excluded.

Results: During this period, 71 children (median age 2.9 years, range 3 days–20.8 years) underwent surgery. All but 1 underwent primary mitral valve repair. Twenty-two (30%) were younger than 12 months. Associated cardiac lesions were present in 45 children (63%) and were addressed concurrently in 35; previous cardiac procedures had been performed in 17 patients (24%). Mitral incompetence was predominant in 60 (85%) and stenosis in 11 (15%). During a median follow-up of 47.8 months (range 2–120 months), 14 patients underwent 17 mitral reinterventions: 14 repairs and 3 replacements. After 60 months, overall survival was $94\% \pm 2.8\%$; freedoms from reoperation and prosthesis implantation were $76\% \pm 5.6\%$ and $94\% \pm 3.6\%$, respectively. There were 4 deaths, and all survivors remain in New York Heart Association class I or II with moderate (6 patients) or less mitral dysfunction.

Conclusion: Surgical repair of the congenital mitral valve can be successfully performed with low mortality, satisfactory valvular function at midterm follow-up, and acceptable reoperation rate while obviating risks associated with valvular prostheses. Suboptimal primary repair was significant predictor for reoperation but re-repair was often successful.

In the past decade, the surgical approach to congenital mitral valve disease has significantly evolved as successive midterm and long-term series have been reported.¹⁻³ Pediatric patients can derive the same benefits from mitral valve repair as adults with regard to preservation of valvular tissue, subvalvular apparatus, and ventricular geometry, leading to optimal valve and ventricular function. Furthermore, avoidance of mechanical prostheses is especially desirable in young children, in whom annular growth should be fostered and who may have little physical space for the prosthesis in the heart.

After pediatric mitral valve replacement, mismatch between native annulus and mitral prosthesis has been shown to be a risk factor for both early and late death.⁴⁻⁶ The probability of mitral valve prosthesis re-replacement was demonstrated to be

Abbreviations and Acronyms

PAP = pulmonary arterial pressure
PTFE = polytetrafluoroethylene

inversely related to the absolute size of the prosthesis initially implanted.⁷ Finally, the cumulative risk generated by a life-long commitment to anticoagulation should be avoided whenever possible.

Diagnostic tools are evolving rapidly and allow superior anatomic diagnosis and monitoring of the surgical repair. The range of surgical techniques modified from adult surgery into pediatric practice or specially developed for pediatric patients is large and allows tailoring of the surgical techniques to anatomic requirements.

Congenital mitral valve disease is rare and frequently associated with other cardiac malformations. Because it is usually complex, intervention is ideally postponed to allow time for annular growth and tissue maturity.⁸ This is usually considered to be safe, because depressed systolic ventricular function has been shown to recover after successful mitral valve surgery in pediatric patients.^{9,10} Severe congestive cardiac failure refractory to maximal medical therapy, however, can result in surgery being undertaken in the first months of life.

In 1996, our unit implemented a strategy whereby mitral valve replacement if necessary is planned when the mitral valve annulus diameter allows it to be done with low early or long-term risk. Mitral valve repair in this context may be considered a palliative procedure designed to allow time for growth. This study reviews our 10-year experience in children undergoing this repair-oriented mitral valve strategy.

Materials and Methods

From January 1996 to March 2006, a total of 71 patients living in Australia underwent surgery at our institution for congenital mitral valve disease. Data were obtained from institutional databases, supplemented by medical records from referring cardiologists or general practitioners, from January 2006 to March 2006. This study was approved by our institutional human ethics committee. Median age at operation was 2.9 years (range 3 days–20.8 years); 22 patients (30%) were younger than 12 months. Median weight at operation was 15.0 kg (range 3.0–99.4 kg). Six patients were dependent on mechanical ventilation before the operation. Primary mitral valve repair was possible in all but 1 case. Seventeen patients had undergone 19 nonmitral cardiac interventions before mitral surgery. Associated cardiac lesions were present in 45 children (63%) and addressed concurrently in 35 when not previously treated.

Patients with partial or complete atrioventricular canal, atrioventricular discordance, or ischemic mitral regurgitation from anomalous origin of the left coronary artery from the pulmonary artery were excluded. Patients with idiopathic dilated cardiomyopathy and functional mitral regurgitation were also excluded. Children

with cleft mitral valve were included, as were 3 patients undergoing staged univentricular pathways with normally sized left ventricle, hypoplastic right ventricle, and congenital mitral valve dysplasia. Genetic, chromosomal, or systemic syndromes were present in 18 patients (25%); these included Marfan syndrome, Shone syndrome, Barlow's disease, William syndrome, and Down syndrome.

Timing of Surgery

Indications for surgery varied according to the etiology and anatomy, the age of the patient, the size of the mitral valve annulus, and clinical status. Neonates and infants with severe mitral valve disease were only considered for operation if they had severe symptoms. In patients with an annulus larger than the smaller valve prostheses available (20 to 21 mm with cuff), no symptoms were required if the valve could be repaired simply without annuloplasty (cleft mitral valve); for more complex valves, symptoms were usually present at the time of surgery. Surgical indications for patients with predominant mitral stenosis were dictated by symptoms only. No specific threshold figure for either pulmonary arterial pressure (PAP) or transmitral gradient triggered a surgical indication if few or no symptoms were present.

Preoperative Evaluation

Preoperative valve function was assessed by transthoracic echocardiography according to the American Society of Echocardiography guidelines.¹¹ Mitral incompetence was severe in 37 patients, moderate in 19, and mild in 3, with associated severe stenosis in 1, moderate stenosis in 1, and mild stenosis in 4. In 1 patient, the cleft mitral leaflet was discovered during ventricular septal defect repair and closed concurrently. Mitral stenosis was severe (mean gradient >15 mm Hg) in 7 patients and moderate (mean gradient 10–15 mm Hg) in 4, with coexistent moderate mitral incompetence in 2 and mild incompetence in 2. All patients with predominant mitral stenosis had pulmonary hypertension; mean peak systolic PAP was 64 mm Hg (range 45–100 mm Hg).

Intraoperative echocardiography was used to assess mitral valve function before and after repair. In this era, transesophageal echocardiography was used in 62 cases (87%), with epicardial echocardiography used in the remainder. No patients underwent diagnostic catheter study.

Mitral Anatomic and Functional Classification

We classify the mitral valves according to three criteria: hemodynamic, functional, and anatomic. This is also in accordance with a standardized classification.¹² Hemodynamically, the valves may be considered to be predominantly regurgitant or stenotic. Mitral incompetence was predominant in 60 valves (85%), and stenosis predominated in 11 (15%). The functional classification was according to the Carpentier classification,¹³ with normal (type I), enhanced (type II), and restricted leaflet (type III) motion. From the anatomic point of view, we divided the congenital mitral valve anomalies into those with nondysplastic leaflets and those with dysplastic leaflets. Nondysplastic leaflet anatomy can occur with annular dilation, with or without elongation of the chordae or the papillary muscle. Such anomalies are usually found with significant volume loading of the left ventricle (large ventricular septal defect or large patent ductus arteriosus). In such cases, the papillary muscle may have a beige, ischemic appearance. A

common feature of mitral valves with dysplastic leaflets is a lack of valvular tissue, albeit variable in distribution. All three major anatomic types classically described by Carpentier and colleagues¹⁴ are found within this series: fusion of papillary muscle to commissure, arcade (hammock) mitral valve, and parachute mitral valve. In these three anatomic types, the hemodynamics can be either predominantly regurgitant, predominantly stenotic, or, rarely, both stenotic and regurgitant.

Mitral valve anomalies in descending order of frequency were as follows: nondysplastic leaflet with annular dilation ($n = 25$) with leaflet prolapse (anterior, posterior, or both, $n = 22$) or posterior leaflet restriction ($n = 3$), cleft anterior leaflet ($n = 24$), papillary muscle–commissural fusion ($n = 12$), parachute mitral valve ($n = 3$), hammock mitral valve ($n = 2$), accessory mitral valve tag or tissue ($n = 3$), hypoplasia of the posterior leaflet ($n = 1$), and atypical dysplasia ($n = 1$). Supravalvular mitral ring coexisted in 3 cases. When there was more than one anomaly, patients were grouped according to the anomaly considered the most significant.

Surgical Techniques

Continuous cardiopulmonary bypass was performed with bicaval and ascending aortic cannulation at mild hypothermia of 32°C and pump flows of 150 to 200 mL/(kg · min). Intermittent antegrade cold blood cardioplegia was given every 20 to 30 minutes. Mean cardiopulmonary bypass and aortic crossclamp times were 152 ± 76 and 106 ± 54 minutes, respectively. Neither profound hypothermia nor circulatory arrest were used.

Through a midline sternotomy, access to the mitral valve was gained either by a left atriotomy in the interatrial groove ($n = 53$), transeptally ($n = 16$), or with a combined approach ($n = 2$). Exposure was optimized by cannulating the superior vena cava at a distance from the cavoatrial junction and the inferior vena cava adjacent to the cavoatrial junction. A self-retaining mitral valve retractor adapted to the size of the patient was used throughout. Visualization of the valve was further enhanced by mattress sutures in the posterior annulus and pulling the inferior vena cava more snugly up and to the left. The valve was then methodically inspected, and findings were integrated with the preoperative investigations, with care to note the following: presence of a supravalvular mitral ring; annular diameter; leaflet texture and size; number, distribution, and morphologic characteristics of chordae and papillary muscles; nature of commissural tissue; and, finally, the presence of any accessory mitral valve tissue or tag in the interchordal spaces. The valve orifice was measured with Hegar dilators before and after repair, and the value was compared to predicted normal values indexed to body surface area according to a modification of the sizes originally described by Kirklin and Barrat-Boyes.¹⁵ Surgical techniques were tailored to the anatomy and mechanism of dysfunction (Figures 1–3 and E1–E4). Annuloplasty was used in 34 patients (48%). Various other techniques were used and are reported in the Results section.

Follow-up

All patients underwent transthoracic echocardiographic examination before discharge from the hospital and then usually at 6-month intervals unless a change in clinical examination or status was noted. Concurrent follow-up for the 68 survivors was complete at a median of 47.8 months (range 2 months–120 months).

Figure 1. Polytetrafluoroethylene complete posterior annuloplasty. The arrow shows the site of interruption of the annuloplasty, if some growth of the posterior annulus is desired.

Statistical Analysis

Data are presented as mean $\pm$ SD or as median and range, unless otherwise specified. Proportions were expressed with continuity correction for the upper and lower limits. Primary end point of the statistical analysis was reoperation for mitral dysfunction. Risk factors for mitral reoperation were determined with logistic regression, and Cox proportional hazards analysis was performed for time to reoperation. All variables that achieved a P value less than .2 in the univariate analysis were included in a multivariate Cox regression model. Determinations of freedom from mitral reintervention and moderate or greater mitral regurgitation or stenosis were performed with the Kaplan–Meier method.

Results

Mortality, Morbidity, and Reoperations

There were 3 early deaths (4%). Primary mitral valve repair was performed in 70 patients. An 11-month-old patient with a large accessory valve tag that was responsible for left ventricular outflow tract obstruction and severe mitral regurgitation underwent mitral valve replacement after a failed attempt at repair and died on the first postoperative day of cerebral hemorrhage.

An 8-year-old girl with papillary muscle–commissural fusion with moderate regurgitation, severe left ventricular outflow tract obstruction, and Ohdo syndrome died on postoperative day 41 of ongoing sepsis. A 3-month-old boy with Marfan syndrome, severe mitral regurgitation, and severe left ventricular dysfunction died of low cardiac output on the day of his mitral valve repair. Seven patients underwent revision of the initial mitral valve repair in the same anesthetic session after transesophageal or epicardial echocardiography. During the follow-up period, 14 patients underwent 17 mitral valve reinterventions (14 repairs and 3 replacements). Indications for surgical intervention were increased mitral regurgitation ($n = 12$), repair rupture

Figure 2. Correction of functional type 2. **A,** Papillary muscle wedge resection. **B,** Polytetrafluoroethylene chordae. **A** indicates height of prolapse to be corrected.

($n = 2$), severe stenosis ($n = 1$), severe valve-related hemolysis ($n = 1$), and secondary development of a supra-ventricular mitral ring ($n = 1$). Two patients required valve replacement with 21-mm mechanical prostheses in the anatomic position, and 1 patient received a 12-mm mitral homograft.

There was 1 late death of noncardiac sepsis at 6 months after the operation. At 60 months, actuarial survival was $94\% \pm 2.8\%$; actuarial freedoms from mitral reintervention and prosthesis implantation were $76\% \pm 5.6\%$ and $94\% \pm 3.6\%$, respectively.

Mitral Cleft

The group of patients with mitral cleft had the fewest and least severe symptoms; all had a normal preoperative PAP unless a large ventricular septal defect was present. Only 3 patients in this group required annuloplasty. One patient required a reoperation for secondary valve perforation. Two patients had moderate residual regurgitation at follow-up. All

other patients in this group remained free of symptoms at last follow-up.

Mitral Regurgitation With Etiology Other Than Cleft

There were 36 patients in the group with mitral regurgitation with exclusion of the cleft mitral valves. Only 11 patients had dysplastic leaflets: 2 hammock valves (no reoperation), 4 valves with papillary muscle–commissural fusion (1 reoperation with valve replacement and 1 early death), 1 parachute mitral valve (2 reoperations, with the second a valve replacement leading to a late death), 2 valves with accessory valve tissue (1 valve replacement with early death), 1 valve with agenesis of the posterior leaflet, and 1 complex mitral valve dysplasia in the context of tricuspid atresia (2 reoperations). Twenty-five patients had a nondysplastic anatomy with a functional type I and either an anterior type II ($n = 16$), a posterior type II ($n = 1$), or both anterior and posterior ($n = 5$). Three patients had coexisting type I and III posterior.

Figure 3. Correction of functional type 3. Detachment of posterior leaflet (1) facilitates papillary muscle mobilization (2). Patch enlargement allows larger annuloplasty (3).

This group had 6 reoperations and 1 early death (infant Marfan syndrome). At follow-up, among the 33 survivors, 5 patients have mild to moderate regurgitation and 11 patients have mild or less residual regurgitation.

Mitral Stenosis

All 11 patients with mitral stenosis had dysplastic leaflets. The median age was 7.3 months (range 3 days–14.5 years). This group had the most severe symptoms at the time of repair, with a mean peak systolic PAP of 64 mm Hg (range 45–100 mm Hg) and 5 patients with ventilator dependence. There were 8 patients with fusion of papillary muscle to commissure, 2 of whom had supramitral ring (1 reoperation for recurrent fusion). Three patients required 1 reoperation, and 1 required 3 reoperations. The latter patient eventually underwent replacement with a 21-mm mechanical prosthesis, whereas the annulus at the time of the first repair (at 7 months) was 11 mm. Two patients had a parachute mitral valve (1 reoperation for secondary supramitral ring); 1 patient had accessory mitral valve tag. No patient underwent annuloplasty, and there were no early or late deaths in this group. The recurrence of a supralvalvular mitral ring tends to support the acquired origin of this lesion as a result of turbulent flow in the mitral orifice. Ten patients have no symptoms at follow-up, and 2 patients have an elevated PAP (systolic values of 53 and 64 mm Hg, respectively). One of these 2 is without significant residual gradient; the other has residual moderate stenosis and regurgitation with good systolic function and should respond well to further valve replacement.

Annuloplasty

Among the 34 patients who underwent annuloplasty, there were no reoperations in the group with remodeling annuloplasty (0/9), 1 reoperation in the group with posterior band (1/5), 1 reoperation in the group with posterior polytetrafluoroethylene (PTFE) band (1/7) (Figure 1), 3 reoperations

in the group with divided PTFE band (3/5), no reoperations in the group with plication or compression of the annulus (0/4), and no reoperations in the group with annular compression with mattress sutures (0/3). There was 1 posterior annuloplasty with pericardium. In the group with mitral regurgitation of noncleft origin, among the 17 patients large enough to receive a commercially available device for annuloplasty or a continuous posterior PTFE annuloplasty, only 2 reoperations were required. On the other hand, the 18 patients who underwent either no annuloplasty or a custom annuloplasty to accommodate growth because of small size included 7 reoperations and 3 deaths ($P = .08$). Severely dysplastic mitral valves generated symptoms earlier in life and required a more difficult operations on smaller annuli than did less severe anatomic substrates. The statistical analysis failed to establish whether these patients were more susceptible to reoperation because of the anatomy, a less satisfactory annuloplasty, or both.

Statistical Analysis

According to univariate analysis, only age younger than 1 year ($P = .042$) and residual valvular dysfunction of moderate or greater stenosis or regurgitation at hospital discharge ($P = .007$) were significant risk factors for reoperation. Multiple logistic regression isolated the residual valvular dysfunction as sole predictor for reoperation ($P = .07$).

Discussion

Surgical Strategy

Congenital malformations of the mitral valve are rare, complex, and frequently associated with other cardiac malformations. Treatment options include mitral valve repair and replacement with mechanical prostheses; bioprostheses are contraindicated in children because of accelerated and premature degeneration.

Figure 4. Kaplan–Meier curves ($n = 71$) showing freedoms from death (solid line), reoperation (Reop., thick dashed line), and prosthesis (thin dashed line) in patients undergoing mitral valve surgery.

Oversized mechanical valves in the annular or supra-annular position are associated with native annulus–prosthesis mismatch, with a strong negative impact on survival⁴; however, recent series in small patients with implantation in the annular position have reported good long-term results. This is especially true in centers where the management of pediatric warfarin therapy is optimal. Atrioventricular valve development is still ongoing in the first months of postnatal life,⁸ so surgical intervention should be deferred as long as the patient's condition can be managed medically to increase the chances of a satisfactory and robust repair. Although good valve function is frequently achieved, if this proves infeasible, the principal aim is then to improve the hemodynamic status and postpone the implantation of a mitral valve prosthesis until annular growth has occurred. Our surgical strategy derives from these concepts and is illustrated by the cases of 2 patients who later successfully received 21-mm mechanical prostheses. Furthermore, where initial repair was suboptimal, interim palliation could be achieved before successful further repair.

We were surprised to find that congenital mitral valve stenosis was associated with a better outcome than dysplastic mitral regurgitation in this series. This finding is probably explained by earlier surgical intervention than in the past, when established damage to the pulmonary circulation adversely affected outcome, and by an enhanced medical armamentarium for the treatment of pulmonary hypertension in the intensive care unit. As is borne out in this series, repair of most valves is possible in patients older than 1 year.¹⁷ As the tolerance of anticoagulation in children steadily improves,¹⁸ however, a lower threshold than ours for mitral valve replacement can be supported.

Classification

Usually, regurgitant and stenotic mitral valves are classified as distinct entities and presented separately. In congenital mitral anomalies, however, mostly because of the presence of the dysplastic leaflet group, the anatomy over-

laps the functional groups and repair strategies can be identical.

Technique

Diverse techniques were used in this series.¹⁶ In most cases, several techniques were used simultaneously, including annuloplasty. Reoperations to repair rupture were indicated for only 2 patients younger than 12 months. In both cases, further repair was successfully achieved. In most instances, rupture of the initial repair could not be demonstrated at reoperation; instead, it is likely that the initial repair itself was unsatisfactory or that the annulus progressively enlarged. In that respect, residual regurgitation after repair in a very small annulus can be considered beneficial in promoting accelerated growth of the annulus.

Cleft Mitral Valve

Two patients in the group with cleft mitral valve had long-term moderate regurgitation. This is explained by the secondary lesion caused by the chronic regurgitation before surgery. The secondary lesions profoundly altered the pliability and elasticity of the leaflet tissue, significantly compromising the chances of a satisfactory result. Because this surgery has virtually no risk and can provide perfect results, it is essential that patients with cleft mitral valve be referred as soon as their regurgitation is greater than moderate. At that stage, patients are usually free of symptoms.¹⁹

Patch Augmentation of Leaflet

Patch augmentation^{20,21} is an invaluable tool for the repair of congenital valves. In dysplastic leaflets, it allows compensation for the lack of native valvular tissue; in small valves, it is used to increase the area of the leaflet tissue to permit a larger annuloplasty. The glutaraldehyde-treated autologous pericardium is the only material that can match the flexibility of the pediatric valvular tissue. To increase the flexibility, we have started treating the pericardium for a shorter time (4 minutes).²² With respect to the lesser result obtained with custom

annuloplasty to achieve growth in the smaller patient, we continue to advocate its use in this challenging subgroup in lieu of a more satisfactory alternative.

Annuloplasty

The function of annuloplasty is to stabilize the repair and to match the area of the leaflet tissue to the cross-sectional area of the mitral orifice in systole. It should be considered mandatory in all repairs for mitral valve incompetence, with the exception of some isolated type I variants with no annular dilation, such as cleft mitral valve. Repair of mitral valve incompetence without annuloplasty often results in recurrence,¹⁷ as supported by a trend identified in our series ($P = .08$). We anticipate that absorbable annuloplasty techniques may be helpful in managing children with very small annuli.

Several techniques of annuloplasty were used in this series. We used a remodeling rigid ring each time an adult-sized device could be inserted (30 mm for female patients and 32 mm for male). For intermediate annuli, we used a posterior annuloplasty with continuous PTFE band. In very small annuli, we used a row of compression mattress sutures from one commissure to the other. We favor this technique rather than the divided PTFE band.

Predictors of Outcome

The Kaplan–Meier freedom from reoperation curve (Figure 4) shows that 50% of reoperations took place within 2 to 3 months after primary repair, suggesting that the initial repair may have been suboptimal. A feature of the current literature concerning congenital mitral valve repair is a lack of predictors of adverse events, a lack that may be inherent in the small series and heterogeneous morphologic subtypes. Postrepair results of intraoperative transesophageal echocardiography were not a statistically significant predictor for future reintervention, which is perhaps not surprising given errors of underestimation during general anesthesia relative to postoperative transthoracic echocardiography in awake patients.²³

Conclusions

This series shows that it is possible to delay and often avoid altogether mitral valve replacement for congenital mitral valve disease. Early mitral valve repair saves the lives of patients with severe symptoms, particularly those with mitral stenosis. Early repair can be performed with low mortality and with an acceptable reoperation rate while obviating risks associated with valvular prostheses. When definitive repair cannot be realized, effective palliation can be achieved to permit growth and subsequent implantation of larger prostheses in anatomic position. Suboptimal primary repair is a significant predictor for reoperation, but re-repair is most often successful. In patients with mitral regurgitation, the annuloplasty of small annuli remains a difficult technical issue.

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Discussion

Dr John J. Lamberti (*San Diego, Calif*). I compliment Dr Oppido for his excellent presentation. Oppido and colleagues have analyzed their experience in repairing anatomic mitral valves. Seventy of 71 patients underwent successful valve repair, with a relatively small percentage returning for early re-repair. In general, I agree with their indications for surgery and many of the technical details described in the article.

Oppido and colleagues favor exposure through a conventional left atriotomy. I prefer the transeptal approach, and I have a very low threshold for extending my incision into the dome of the left atrium, thereby creating optimal exposure of the valve, especially in small patients. Intraoperative valve testing requires that the valve be undistorted while static testing is taking place.

Dr Oppido didn't mention in his presentation, but transesophageal echocardiography was used in about 87% of their cases. We rely heavily on the postrepair transesophageal echocardiography in determining whether our repair is adequate. In complex repairs, we recommend performing a preliminary transesophageal echocardiogram after recovery of the myocardium has occurred but not necessarily before full rewarming has been completed. In about 10% of patients, we may go back to bypass and make adjustments to the repair. This approach allows fine-tuning of the repair. We always place a left atrial pressure monitoring catheter before separation from bypass. The postrepair analysis must be performed under optimal loading conditions if we are to make inferences about the quality and durability of the repair.

Most of the early reoperations in Oppido and colleagues' series occurred in patients younger than 1 year. Only 87% of the patients underwent transesophageal echocardiography at the time of the first repair. In our experience, analysis of the repair in the operating room is very important in predicting short- and long-term outcomes. When a patient is too small for a transesophageal echocardiographic probe, we use epicardial echocardiography to analyze the repair. We assume that the quality of the repair will never be better than as seen on the immediate postrepair transesophageal echocardiogram. The predischarge transesophageal echocardiogram often shows a little worse regurgitation. We integrate our operative findings with those of the postrepair transesophageal echocardiogram, and on a few occasions we've been able to modify the repair to improve function.

We agree with Oppido and colleagues that a very eclectic approach must be taken to repair the unusual pathology encountered in patients born with congenital abnormalities of the mitral valve. Oppido and colleagues have demonstrated that most abnormal mitral valves can be salvaged. In the future, preoperative 3-dimensional echocardiography could be useful in planning repair strategies. We don't have that available at our institution.

The goal of this surgery, as clearly stated by Oppido and colleagues, is not cure but postponement of mitral valve replacement as long as possible. I have several questions.

First, Dr Oppido, did you use epicardial echocardiography in those patients for whom transesophageal echocardiography was not available?

Dr Oppido. Yes. We strongly believe, as you do, that transesophageal echocardiography is the criterion standard for intraoperative evaluation before repair, to inform an appropriate repair, and to check the immediate result of the repair. As for the patients who didn't undergo transesophageal echocardiography, they did undergo epicardial echocardiography.

Dr Lamberti. Was any valve revised in the operating room on the basis of the postrepair transesophageal or epicardial echocardiogram?

Dr Oppido. Yes, we revised the repair in approximately 10% of the patients on the basis of what we saw on the transesophageal echocardiogram.

Dr Lamberti. So actually, all the patients had some form of echocardiography in the operating room?

Dr Oppido. Yes.

Dr Lamberti. In reading the article, that wasn't clear, and I was wondering whether the early failures correlated with patients who did not have an echocardiogram before leaving the operating room.

Dr Oppido. All the patients underwent either transesophageal echocardiography (which was done in nearly all patients, 87%) or epicardial echocardiography.

Dr Lamberti. Thank you. It's an excellent article.

Dr Rodolfo Neirotti (*Cambridge, Mass*). I agree that annuloplasty techniques involving a ring can be a problem in the pediatric population. Although rigid and flexible rings meet the needs of adults, they do not allow room for growth of the native annulus when implanted in children. I think that biodegradable rings will address this problem. Atrioventricular valve repair with this new technology is feasible, with good early and midterm results.

Dr Oppido. In this series, we didn't have any experience with absorbable annuloplasty devices, but we anticipate that they may be helpful in selected cases.

Dr Alain F. Carpentier (*Paris, France*). Dr Oppido, I congratulate you. When comparing this surgery to what I did in the past, it's obviously a tremendous improvement. The mortality has been reduced significantly, and also you followed the guidelines. Obviously, there has been a lot of progress made. I do have two questions.

First, you mentioned the need to have a sort of annuloplasty allowing the growth of the annulus, which is true for babies and people younger than 10 years but not for people older than 10 years. So do you have a different strategy, depending on the age, and particularly regarding using remodeling annuloplasty whenever possible, when you don't expect the need for growth of the annulus? That's my first question; I have another brief question afterward.

Dr Oppido. We used several techniques for annuloplasty, and we strongly believe that every patient with mitral regurgitation and annular dilation requires annuloplasty. So in the smaller patients it should be feasible to use the sort of interrupted posterior annuloplasty with three-times-folded PTFE. After being positioned in the posterior annulus, this band is cut in one or more points to allow room for growth. Some other techniques, such as interrupted mattress suture to compress the posterior annulus, were also done in the smaller patients.

Dr Carpentier. No, my question is specifically, do you have another strategy for older patients who can accommodate remodeling annuloplasty, that is to say, a complete ring? I noticed that you have

patients as old as 20 years, or at least beyond 15 years. Of course, it's a strategy more for an adult than for a young baby.

Dr Oppido. Yes, of course, in adult patients we implant an adult-sized commercially available complete ring. We did it in 9 patients in this series: posterior band in 5 patients and posterior complete annuloplasty with PTFE in 7 patients.

Dr Carpentier. For my second question, how often have you been using leaflet enlargement with pericardial patching?

Dr Oppido. Posterior leaflet was enlarged in 10 patients; anterior leaflet was enlarged in 4 patients.

Dr Carpentier. And you haven't seen any calcification of the patch?

Dr Oppido. None at all.

Dr Carpentier. Thank you.

Dr Carlos Troconis (Miami, Fla). In your article, you did note that the mitral valve was replaced in 4 cases, 1 early and 3 late. What type of prosthesis did you use, mechanical or bioprosthesis? What size and technique? What criteria did you chose?

Dr Oppido. The main goal of our policy, first of all, is to implant a prosthesis only in the intra-annular position, so we tend to avoid any different kind of implant, such as supra-annular and so on. So we implanted in this series, as a primary replacement, 19-mm mechanical prostheses. And in the follow-up, during the reoperations, we implanted two 21-mm mechanical prostheses and a homograft valve as the fourth replacement. And it is interesting that we could implant the two 21-mm prostheses in patients who were operated on early in life; the annulus at the beginning, at the first operation, was 11 or 12 mm. That was a good result, I think.

Pedro J. del Nido (Boston, Mass). I also congratulate you. This is a follow-up to your previous study on recurrent mitral regurgitation. Now you are presenting more longitudinal information, and the results seem to hold up over the years.

My question relates to an area in which we've become much more interested, congenital mitral stenosis, particularly in the infant. It's a very different disease than regurgitation. Most of the problem appears to be at the leaflet level. Images like the one that you showed, with apparent short chordae, are in fact deceptive. In fact, if you put a light behind those leaflets, you see that the chords are much, much longer; they're simply covered by a layer of what looks like endocardial tissue that actually mats those chords together, and you can strip this tissue off the chords. Van Praag demonstrated this more than 20 years ago.

My question relates to your management of mitral stenosis. It appears that your approach is primarily that of leaflet augmentation or leaflet replacement, with pericardium or whatever available tissue you have. Have you looked into leaflet thinning and removal of

the endocardial layer of tissue that actually creates much of the stenosis? My concern with the replacement approach is that it still leaves a funnellike mitral valve, and attempts at commissurotomy are just going to lead to regurgitation. So might a different approach be more applicable?

Dr Oppido. Thank you for your question. Yes, of course, I showed just a few of the techniques we used. And especially in patients with mitral stenosis and papillary muscle–commissural fusion, we used several techniques, such as removing the excessive tissue between the chordae, chordal fenestration (and when the stenosis was at the papillary muscle level, papillary muscle fenestration), and when there was stenosis at the commissural level, commissurotomy. Of course, these were also integrated with the posterior leaflet enlargement.

Dr del Nido. Can you comment about the durability of that approach? In other words, how many of those patients required reoperation?

Dr Oppido. You're talking about what patients?

Dr del Nido. Patients with congenital mitral stenosis.

Dr Oppido. In this series, we had 11 patients with congenital mitral stenosis. All are alive. Only 1 patient has moderate dysfunction, and 1 patient has required replacement.

Dr Giovanni Stellin (Padua, Italy). I congratulate you on your results. A similar experience from our group was presented 3 days ago at the meeting of the World Society for Pediatric and Congenital Heart Surgery. We have managed to reconstruct the whole spectrum of the mitral valves disease.

You showed an image of a mitral valve malformation that you have defined as papillary muscle–commissural fusion (Image shown in Brizard,¹⁶ figure 124-3). According to Carpentier's classification, what you have shown is not papillary muscle–commissural fusion; it appears to me more like the typical hammock valve or perhaps a mitral arcade, a malformation defined by two big papillary muscles and small chordae. I therefore ask, how do you define a hammock valve or a mitral arcade?

Dr Oppido. Of course, there is a wide, continuous spectrum among those dysplastic valves, especially in the stenotic group; however, what we define as hammock mitral valve is a mitral valve with an extremely dysplastic subvalvular apparatus in which it is not even possible to recognize papillary muscle in the tensor apparatus, or with some dysmorphic papillary muscle displaced toward the base of the heart, just behind the posterior leaflet, pulling both leaflets toward the posterior part of the heart. That's how we define hammock valve; what we showed is for us papillary muscle–commissural fusion, or short chordae syndrome.

Figure E1. Correction of functional type 1. Two types of divided posterior annuloplasty.

Figure E2. Correction of functional type 2. A, Chordal shortening. A represents height of prolapse to be corrected. B, Chordal transfer. Only secondary chordae should be used.

Figure E3. Correction of functional type 3. **A,** Patch enlargement of anterior leaflet to address lack of leaflet tissue. **B,** Splitting of papillary muscle in conjunction with removal of excess extravascular tissue.

Figure E4. Resection of supra-avalvular ring.