

Congenital Mitral Valve Regurgitation in Adult Patients

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ABSTRACT

Objective: Congenital mitral valve regurgitation (MVR) is a rare disease found in adults. We report on our 5-year surgical experience with congenital MVR in adults.

Methods: We reviewed the data for 48 consecutive patients (26 men), aged >18 years (median, 42 years; range, 18-78 years) who underwent operations for severe congenital MVR between June 2005 and May 2010. Patients with atrioventricular septal defect were excluded.

Results: Congenital MVR was preoperatively diagnosed in 28 cases (58%). The lesions consisted of annular dilation (100%), valvular cleft (58%), prolapsed leaflet (40%), papillary muscle abnormality (5%), commissure fusion (2%), and leaflet deficiency (2%). Mitral valve repair was performed in 42 cases (88%) by means of Carpentier techniques. The other 6 patients underwent mitral valve replacement; one of these patients died of ventricular fibrillation 2 days after surgery. There were no other hospital deaths or late mortality. At the last follow-up (median, 38 months; range, 2-50 months), all 47 patients were in New York Heart Association functional class I or II. Echocardiography evaluations for the 42 patients who underwent the repairs revealed that 32 (76%) of the patients had no or trivial MVR and 10 patients (24%) had mild MVR. No patient underwent reoperation.

Conclusion: Congenital MVR is rare and often misdiagnosed in adults. Mitral valve repair is feasible in the majority of patients, with excellent immediate and medium-term results.

INTRODUCTION

Congenital mitral valve regurgitation (MVR) is a rare disease with a wide array of lesions that challenge surgical management. The disorder is seen primarily in infants and children, although there are many case reports of MVR in adult patients due to congenital mitral valve lesions [Tomita 1997; Hashimoto 2001; Shiran 2004; Zegdi 2005; Aramendi

2006; Mohammadi 2006]. Mitral valve repair can provide satisfactory long-term results [Chauvaud 1998; Ohno 1999] and is preferable to mitral valve replacement, which is associated with a poor prognosis in children [Beierlein 2007].

In this report, we describe our surgical experience of congenital MVR in adults over the last 5 years.

MATERIALS AND METHODS

Between June 2005 and May 2010, 48 consecutive patients (26 men) older than 18 years (median, 42 years; range, 18-78 years) underwent mitral valve surgery at our institution for congenital MVR. These 48 patients represented 5.1% of the adult patients who underwent valve surgery during the same period in our department. Excluded from the study were patients with atrioventricular septal defect. Two of the patients had previously undergone operations for ventricular septal defect, and 1 other patient had undergone surgery for aortic coarctation. Another patient had a history of operation for aortic valve stenosis.

Thirty-nine patients were in New York Heart Association (NYHA) functional class I or II, 6 patients were in NYHA class III, and the remaining 3 patients were in NYHA class IV. Forty-one patients were in sinus rhythm, and the other 7 patients presented with atrial fibrillation. Preoperative echocardiography evaluations revealed severe MVR in all instances. The left ventricular ejection fraction (LVEF) was >60% in 42 patients and <50% in 3 patients. Pulmonary hypertension (>50 mm Hg) was detected in 6 patients.

All of the operations were performed via a median sternotomy with the patient on cardiopulmonary bypass. Mild systemic hypothermia was used in all cases. Myocardial protection was carried out with antegrade cold blood crystalloid cardioplegia. Mitral valve exposure was achieved via a right atriotomy and interatrial septum incision or via a left atriotomy.

On the basis of the surgical analysis, the functional classification was type I (25 patients), type II (15 patients), type IIIa (1 patient), type IIIb (2 patients), type I plus type II (4 patients), or type I plus type IIIb (1 patient). Table 1 summarizes the operative findings for the mitral valve. All patients had >1 mitral valve lesion. Six patients had severe functional tricuspid valve regurgitation. No other cardiac lesions associated with the mitral valve disease were found.

Each mitral valve repair was assessed intraoperatively by transesophageal echocardiography and at discharge by

Received July 18, 2010; received in revised form September 21, 2010; accepted October 13, 2010.

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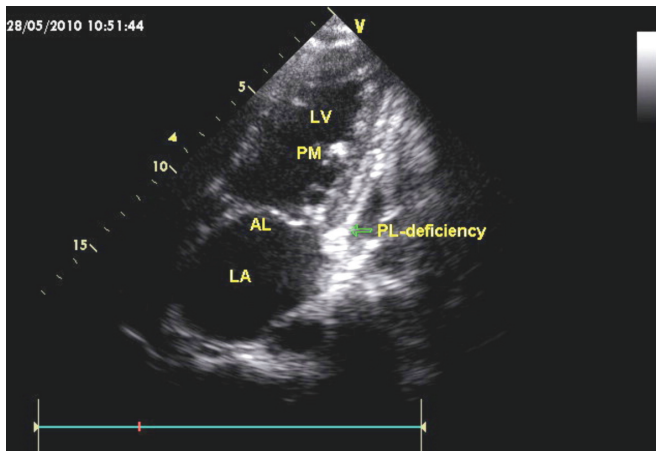


Figure 1. Transesophageal echocardiographic view of a hammock mitral valve. The papillary muscle (PM) is unique, overdeveloped, and displaced upwards. The posterior leaflet (PL) is deficient. LV indicates left ventricle; AL, anterior leaflet; LA, left atrium.

Table 1. Mitral Valve Lesions at the Time of Surgery

Lesion	Patients, n
Annular dilation	48 (100%)
Valvular cleft	28 (58%)
Anterior	26
Posterior	2
Valvular retraction	5 (10%)
Leaflet deficiency	1 (2%)
Leaflet prolapse	19 (40%)
Anterior	8
Posterior	5
Bivalvular	6
Commissure fusion	1 (2%)
Papillary muscle abnormality	3 (5%)
Papillary hypoplasia	2
Hammock valve	1

transthoracic echocardiography. Long-term follow-up data were obtained through contacts with the patients or their relatives.

Data are expressed as the median for continuous variables and as the percentage for categorical variables.

RESULTS

Congenital MVR was diagnosed preoperatively in 28 patients (58%) (Figures 1 and 2). In all other instances, congenital lesions were discovered intraoperatively.

Mitral valve repair was performed in 42 patients (88%). Details are presented in Table 2. The other 6 patients underwent mitral valve replacement. The median cross-clamping time was 59 minutes (range, 30–128 minutes), and the median

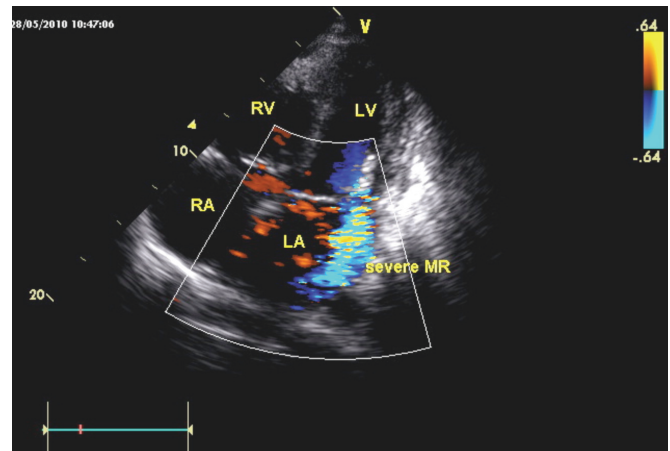


Figure 2. Color Doppler flow imaging of the patient in Figure 1. RV indicates right ventricle; LV, left ventricle; RA, right atrium; LA, left atrium; MR, mitral regurgitation.

Table 2. Repair Techniques in 42 Adult Patients with Congenital Mitral Regurgitation*

Repair Technique	Patients, n
Cleft suture	28 (67%)
Quadrangular resection of PL	6 (14%)
Without sliding plasty	4
With sliding plasty	2
Triangular resection of AL	3 (7%)
Chordal shortening	3 (7%)
Artificial chordae	2 (5%)
Edge to edge	3 (7%)
Commissuroplasty	1 (2%)
Prosthetic annuloplasty	40 (95%)

*PL indicates posterior leaflet; AL anterior leaflet.

bypass time was 91 minutes (range, 59–191 minutes). Concomitant procedures included aortic valve replacement in 1 case, tricuspid valvuloplasty in 6 patients, and left atrium folding in 12 patients.

One patient (2.08%) died of ventricular fibrillation 2 days after an operation for mitral valve replacement. This case was complicated by a giant left ventricle (95 mm in diameter) and a poor left ventricular function (LVEF, 38%) preoperatively. Postoperative morbidities included 2 cases of low cardiac output syndrome, 1 case of reoperation for bleeding, and 1 case of hemodialysis for renal dysfunction.

At patient discharge, echocardiography evaluations revealed no cases of systolic anterior motion of the repaired mitral valve or prosthesis abnormality. Of the 42 patients who underwent mitral valvuloplasty, 36 (85.71%) had no or trivial regurgitation, and 6 patients (14.29%) had mild regurgitation. No mitral valve stenosis was found (mean trans-mitral

valve diastolic gradient, 2-5 mm Hg). Of the 47 surviving patients, 18 had an LVEF <50%.

Follow-up was completed for all 47 patients (median, 38 months; range, 2-50 months). There was no late mortality. All patients were in NYHA functional class I or II. The final echocardiography evaluation revealed no or trivial mitral regurgitation in 32 patients (76.19%) and mild regurgitation in 10 patients (23.81%). The mean transmitral diastolic gradient (for mitral valvuloplasty patients) was 3 mm Hg. The LVEF was <50% in 6 patients. No thromboembolic or hemorrhage episodes occurred.

DISCUSSION

Congenital MVR is a well-known but infrequent disease in the pediatric population. When sufficiently severe, mitral valve surgery is mandatory, owing to the poor prognosis of the disease, which can lead to left ventricular failure or pulmonary hypertension [Prifti 2002; Chauvaud 1998; Ohno 1999]. In adults, congenital MVR is rather rare. In our experience, it is present in 5.1% of all adult patients undergoing valve surgery. It is easily misdiagnosed, even when a mitral cleft has been surgically identified.

Carpentier et al [1976] proposed a classification of the lesions. These lesions may affect any level of the mitral valve apparatus. With the exception of parachute valve, all of the lesions were found in our adult population.

Isolated mitral valve cleft is usually diagnosed during childhood. Late diagnosis in adults has already been reported, including rare cases of bileaflet cleft [Mohammadi 2006]. In our series, cleft was the leading cause of MVR (58.33%). Clefts were found primarily in the anterior leaflet (75%); the other location was in the posterior leaflet involving P2 (3 patients) and P3 (4 patients).

Valvular prolapse is another important cause of MVR. In our series, it occurred in 39.58% of the cases. Distinguishing the congenital origin of valvular prolapse in adults from prolapse caused by degenerative conditions or endocarditis may be difficult in some instances. Careful analysis of the subvalvular apparatus may reveal specific pathologic findings that reveal a congenital origin. In our series, these findings included absent chordae (3 patients) and a malpositioned or malformed papillary muscle (4 patients).

Mitral valve repair for congenital MVR in adults has been described many times in the literature [Tomita 1997; Hashimoto 2001; Shiran 2004; Zegdi 2005; Aramendi 2006; Mohammadi 2006]. The feasibility of mitral valvuloplasty depends on 2 factors: the availability of a sufficient amount of leaflet tissue and the pliability of the tissue (ie, nonfibrotic or noncalcified). In congenital MVR, these 2 prerequisites are usually present, which explains the high rate of mitral valve repair. In our experience, 42 patients (87.50%) underwent mitral valvuloplasty, with excellent results observed in the postoperative period and at medium-term follow-up. The other 6 patients (12.50%) underwent mitral valve replacement because of a lack of the prerequisites for repair. Three of these patients were older than 70 years.

Prosthetic annuloplasty is considered an important additional procedure for valvular repair. It can also produce long-term stability. Of the 42 patients who underwent repair, all but 2 underwent prosthetic annuloplasty as a concomitant procedure. Proper sizing of the prosthetic ring depends on the size and the height of the anterior leaflet, owing to the fear of mitral valve stenosis or systolic anterior motion of the mitral valve [Carpentier 1983].

Compared with a pediatric series, the results of the follow-up of the adults after surgery were satisfactory. The 5-year survival rate in our series was 100%, in contrast to a 5-year survival rate of 87.2% in one pediatric series [Yoshimura 1999] and 93% in another [Chauvaud 1998]. Congenital MVR is a rare disease in adult patients. Mitral valve repair is feasible in the majority of these patients, with excellent immediate and medium-term results.

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