

Large Annuloplasty Rings Facilitate Mitral Valve Repair in Barlow's Disease

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Background. Excess leaflet tissue in Barlow's disease predisposes patients to left ventricular outflow tract obstruction and residual mitral regurgitation after mitral valve repair as a result of systolic anterior motion of the anterior mitral leaflet. In addition to conventional repair techniques such as sliding plasty and leaflet shortening, our approach in Barlow's disease has included the use of large remodeling annuloplasty rings (up to size 40 mm). We report our experience with the use of large rings in Barlow's disease.

Methods. From January 2002 to December 2005, 67 patients with Barlow's disease (46 men and 21 women; median age, 55 years; range, 22 to 85 years), mean ejection fraction 0.55 ± 0.08 , and grade 3+ or greater mitral regurgitation underwent mitral valve repair. All had Carpentier type II leaflet dysfunction, with anterior (n = 2), posterior (n = 41), or bileaflet (n = 24) prolapse. Predominant reconstructive techniques were posterior leaflet sliding plasty/plication (n = 65), anterior leaflet triangular resection (n = 16), and chordal

transfer (n = 25). Concomitant procedures included coronary artery bypass grafting surgery (n = 8), tricuspid valve repair (n = 20), aortic valve replacement (n = 3), and CryoMaze (n = 22).

Results. Mitral valve repair was successfully completed in all patients. Annuloplasty ring size was 36 mm (n = 17), 38 mm (n = 22), and 40 mm (n = 28). Predischarge transthoracic echocardiography showed absence of systolic anterior motion (n = 67), no or trace mitral regurgitation (n = 62), and mild mitral regurgitation (n = 5). There was no operative mortality.

Conclusions. Mitral valve repair can be predictably performed in Barlow's disease with excellent early outcomes. Large annuloplasty rings help minimize the risk of systolic anterior motion and are an important adjunct to established repair techniques in this patient cohort with large annular size and excess leaflet tissue.

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Barlow's disease is a common cause of mitral regurgitation (MR) and is characterized by myxoid degeneration with excessive leaflet tissue. Barlow's disease is a cause of MR, usually secondary to leaflet prolapse (Carpentier's type II dysfunction). The primary lesion causing prolapse is chordal elongation or rupture, and secondary lesions include annular dilatation, chordal thickening, papillary muscle calcification, and annular calcification. Although the "billowing posterior mitral valve syndrome" was first recognized as a cause of systolic murmurs in the 1960s by Barlow and Pocock [1], the surgical characterization of what is now known as Barlow's disease was largely undertaken in the 1970s by Carpentier and associates [2]. Excess leaflet tissue, the hallmark of Barlow's disease, poses a specific challenge during mitral valve repair because it predisposes to the development of systolic anterior motion (SAM) of the mitral valve. Systolic anterior

motion in this context occurs by means of two principal mechanisms—either the residual posterior leaflet is left too tall such that it displaces the anterior leaflet into the left ventricular outflow tract; or the annuloplasty ring is smaller than the surface area of the anterior leaflet, potentially forcing excess tissue into the outflow tract [3]. The former is dealt with effectively by Carpentier's sliding-plasty technique [4]. The latter requires that the annuloplasty ring is true-sized to the anterior leaflet, or that alternative procedures are undertaken to address excess anterior leaflet height [5]. Most Barlow valves are large, and if rings are being true-sized then one should expect a preponderance of large rings (36 mm or greater) in repair series that include Barlow valves. There are, however, little confirmatory data in the literature on the use of such large annuloplasty rings. Indeed many institutions do not stock these large ring sizes, and some annuloplasty rings are not manufactured in sizes above 36 mm. In this paper we present our recent experience with

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Dr Adams discloses that he has a financial relationship with Edwards Lifesciences.

Table 1. Patient Demographics

| Characteristics | N (%) |
|---------------------|------------------|
| Male gender | 46 (69) |
| Median age (y) | 55 (range 22–85) |
| Hypertension | 8 (12) |
| Diabetes | 3 (4) |
| Mean EF | 0.55 ± 0.08 |
| NYHA class | |
| I | 12 (18) |
| II | 35 (52) |
| III | 20 (30) |
| IV | 0 (0) |
| MR grade | |
| 3+ | 20 (30) |
| 4+ | 47 (70) |
| Atrial fibrillation | 22 (33) |
| Reoperation | 2 (3) |

EF = ejection fraction; MR = mitral regurgitation; NYHA = New York Heart Association.

mitral valve repair using large annuloplasty rings (between size 36 and 40 mm) in patients with Barlow's disease.

Material and Methods

Patients

All patients undergoing mitral valve surgery at our institution are entered into a prospective database. Institutional Review Board approval with a waiver of individual consent was obtained for this study. We retrospectively identified patients who underwent mitral valve surgery between January 2002 and December 2005 and had a surgical diagnosis of Barlow's disease. To be included in this series patients must have had an annuloplasty ring of 36 mm or greater and a primary indication for surgery of MR. Seventy-five patients underwent mitral valve surgery for Barlow's disease during the study period, all of whom underwent successful repair. There were no valve replacements undertaken for Barlow's disease during the study period. Eight patients had a *forme fruste* of the disease with excess, thickened tissue

but a valve size smaller than 36 mm and were excluded from this study, leaving 67 consecutive Barlow patients who had mitral valve repair using annuloplasty rings of 36 mm or greater. These 67 patients form the cohort for this analysis. Table 1 summarizes the preoperative characteristics. Twenty patients (30%) had 3+ MR (moderate to severe) whereas 47 patients (70%) had 4+ MR (severe).

Definitions

Barlow's disease was defined according to the lines of Carpentier and associates [2, 6, 7]. Usually patients with Barlow's disease have a long history (several years) of a murmur and do not have any alternative causative factors (such as Marfan's disease or endocarditis). On inspection of the valve, we used the following characteristics to define a valve as Barlow's (Fig 1A):

Billowing valve with excess tissue and thickened leaflets
Chords typically thickened and elongated
Large valve with severe annular dilatation

Chordal rupture, annular calcification, and papillary muscle calcification may be present.

Surgical Techniques

All procedures were performed by a single surgeon (D.H.A). Repairs were performed through a full sternotomy (N = 54, 80%) or minimally invasive approach (N = 13, 20%). Our minimally invasive approach was an 8- to 10-cm lower midline incision and lower hemisternotomy. Operations were performed using mild hypothermic cardiopulmonary bypass with antegrade and retrograde warm and cold blood cardioplegia and topical hypothermia used for myocardial protection. All valves were accessed through a left atriotomy in the interatrial groove. During valve analysis, lesions producing valve dysfunction(s) were identified as were the leaflet and annular dimensions. Valve repairs were performed according to the principles of Carpentier [4, 6, 8]. Our specific approach was to sequentially repair the posterior leaflet by resecting any prolapsing segment, reduce the posterior annular dimension, reconstruct the posterior leaflet, perform a true-sized annuloplasty using a complete semirigid remodeling ring, and then repair any residual prolapse of the anterior leaflet or commissures

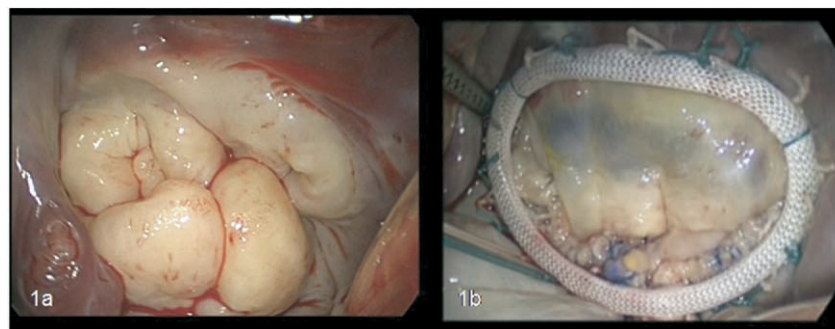


Fig 1. (A) Barlow's valve with excess thickened leaflet tissue and billowing of both leaflets. (B) Barlow valve repair after posterior leaflet sliding plasty and size 40-mm Physio-Ring annuloplasty.

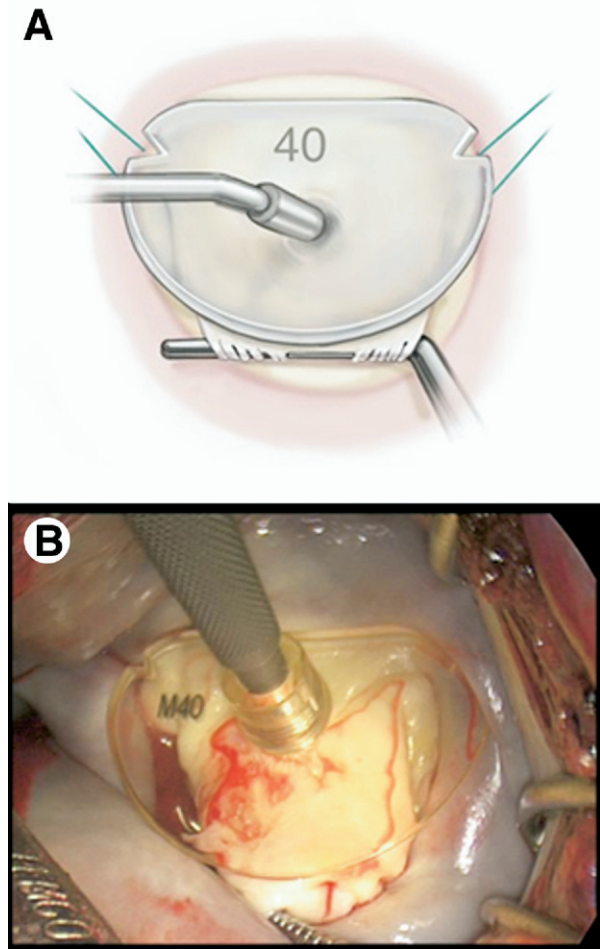


Fig 2. Correct leaflet sizing technique (A) and demonstration of a size 40-mm valve (B).

after inspecting the line of closure during saline testing. In the setting of excess posterior leaflet tissue, we performed a leaflet resection and a sliding leaflet-plasty to reduce the height of the posterior leaflet to 1 to 1.5 cm in all segments. Before leaflet plasty, we reduced the posterior annular circumference by placing horizontal plication sutures to relieve excess tension on the margin of the posterior leaflet. We corrected anterior leaflet prolapse primarily by chordal transfer and triangular resection. A Carpentier-Edwards (C-E) Physio Annuloplasty Ring, (Edwards LifeSciences, Irvine, CA) was used in all patients (Fig 1B) [8]. The ring size chosen was based on the true size of the anterior mitral leaflet. This was determined by using standard C-E mitral sizers to measure both the intercommissural distance and the anterior-posterior leaflet dimension, with a bias to choose the larger size if between sizes (Fig 2). Adjunctive procedures performed included tricuspid valve repair (n = 20; 30%), coronary artery bypass grafting surgery (n = 8; 12%), closure of patent foramen ovale (n = 8; 12%), aortic valve replacement (n = 3; 5%), and a modified maze procedure (n = 22, 33%). Intraoperative transesophageal echocardiography was performed in each patient after weaning

from cardiopulmonary bypass to detect left ventricular outflow tract obstruction owing to SAM and any residual MR.

Echocardiography

Before hospital discharge, two-dimensional and Doppler transthoracic echocardiography examination was performed in all patients using a 3.5-MHz transducer and commercially available echocardiographic systems. The presence of MR was assessed, and its severity was evaluated semiquantitatively using the regurgitant color jet area [9]. Regurgitation was classified as none (when no MR is present), trace (regurgitation that is barely detected), mild (jet area < 4 cm²), moderate (jet area > 4 and < 10 cm²) and severe (jet area > 10 cm²). All echocardiograms were retrospectively assessed in a core laboratory by a cardiologist (V.A.) blinded to the clinical outcome and to the initial interpretation of the echocardiogram.

Data Acquisition

At the time of mitral surgery, all valves were analyzed, and specific lesions and prolapsing segments were documented. All repair techniques were subsequently documented. These data were entered prospectively into a mitral valve database. Clinical outcome data were recorded prospectively in a general cardiac surgery database. Morbidity was defined according to guidelines of the New York State Department of Health [10]. Additional information was obtained by retrospective chart review as required.

Results

Valve Analysis

All patients had Carpentier type II dysfunction. Forty-one (61%) patients had isolated posterior leaflet prolapse, 24 (36%) bileaflet prolapse, and 2 (3%) isolated anterior leaflet prolapse. Segmental analysis revealed that when the posterior leaflet was involved, multiple segments were involved in 37 cases (57%), and when the anterior leaflet was involved, multiple segments were involved in 14 cases (54%). The P2 and A2 segments were the most

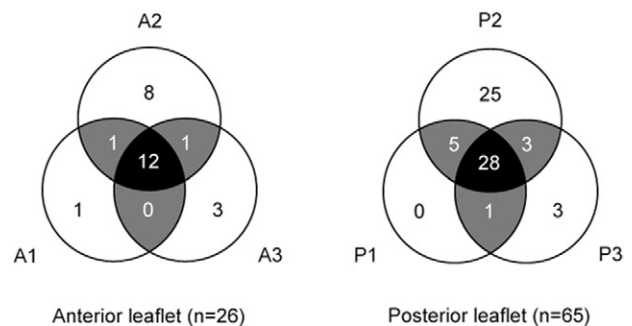


Fig 3. Segmental analysis of valve prolapse. Numbers in middle of large circles depict isolated prolapse of the labeled segment. Overlapping areas depict prolapse involving more than one segment.

Table 2. Surgical Procedures

| Procedure | N (%) |
|----------------------------------|---------|
| Annuloplasty ring size (mm) | |
| 36 | 17 (25) |
| 38 | 22 (33) |
| 40 | 28 (42) |
| Repair techniques | |
| Posterior leaflet resection | 65 (97) |
| Anterior leaflet resection | 16 (24) |
| Posterior leaflet sliding plasty | 56 (84) |
| Chordal transfer | 25 (37) |
| Anterior leaflet shortening | 2 (4) |
| Gore-Tex chordoplasty | 2 (3) |
| Papillary muscle shortening | 2 (3) |

commonly involved. Segmental analysis results are shown in Figure 3. The lesions present were annular dilatation, 67 (100%); chordal elongation, 57 (85%); chordal rupture, 26 (39%); annular calcification, 8 (12%); and papillary muscle calcification, 5 (7%).

Surgical Procedures

All patients underwent successful mitral valve repair documented by intraoperative transesophageal echocardiography. One patient in the series had SAM identified in the operating room. This patient had received a size 38 mm ring and had sliding plasty of the P2 and P3 segments. On the second time during cardiopulmonary bypass the P1 segment was shortened with complete resolution of SAM and no residual MR. The median cardiopulmonary bypass time was 199 minutes (interquartile range, 170 to 260 minutes) and cross-clamp time 168 minutes (interquartile range, 135 to 225 minutes). The valve repair techniques used are shown in Table 2.

Mortality and Morbidity

There was no in-hospital or 30-day mortality. Postoperative complications included respiratory failure requiring prolonged ventilation in 2 patients (3%), bleeding requiring reoperation in 1 patient (1%), and renal failure in 1 patient (1%). In addition, one deep sternal infection occurred (1%).

Echocardiographic Results

Predischarge transthoracic MR grade is shown in Table 3. Sixty-two patients (93%) had no or trace MR, whereas 5 patients (7%) had mild MR. There was no evidence of SAM in any patient. No significant change in mean ejection fraction was demonstrated.

Comment

Using Carpentier's techniques, with an emphasis on removing excess posterior leaflet tissue and respecting excess anterior leaflet tissue by the use of true-sized large annuloplasty rings (36 mm or greater), we have

shown that Barlow valves can be repaired with excellent early echocardiographic results including absence of significant residual MR and minimal risk of SAM. A review of the literature does not yield any similar series of large annuloplasty rings. Most published series on mitral valve repair for degenerative disease do not differentiate between Barlow and non-Barlow causes or do not give information on annuloplasty ring size. Thus it is difficult to know the true incidence of usage of large rings in patients with Barlow's disease. Given the paucity of data on this subject and the lack of large sizes (>36 mm) of some manufactured annuloplasty rings, we believe that the use of such large rings may be relatively infrequent. Most Barlow valves are, however, quite large and have excess tissue by definition. Given the distribution of ring size in our study with a mode of 40 mm, we would expect (assuming a natural distribution) that there are Barlow valves that are size 42 and 44 mm. Indeed we encountered two anterior leaflets with a true size greater than 40 mm (currently the largest manufactured ring size), which required shortening.

The use of true-sized large rings was integral to our approach to avoid SAM and residual MR after mitral valve repair for Barlow's disease. Some workers have advocated reducing the anterior leaflet height [5], performing an edge-to-edge repair [11], using incomplete flexible bands in preference to complete annuloplasty rings [12, 13], or even avoiding annuloplasty rings altogether [14] as a means of reducing the incidence of SAM. Routine resection of the anterior leaflet changes the geometric relationship between the anterior leaflet height and the intercommissural distance, which forms the basis for the concept of remodeling annuloplasty; we prefer to respect this relationship, except in the unusual circumstance of a giant anterior leaflet (>40 mm). Although the edge-to-edge technique has been reported as a means of preventing SAM, the long-term durability of this technique in degenerative mitral valve repair is unknown. Systolic anterior motion has been described both with the use of flexible posterior bands [15] and even without the use of annuloplasty rings [16, 17], such that ring choice or use alone is not the key to avoiding SAM.

A possible explanation for the paucity of data on large rings is that a significant proportion of patients with large Barlow valves may be undergoing mitral valve replacement rather than repair, as these large Barlow valves are among the most complex valves to repair and are also those at greatest risk of SAM.

Table 3. Predischarge Echocardiographic Mitral Regurgitation Grade

| Grade | n (%) |
|---------------|----------|
| None or trace | 62 (93%) |
| Mild | 5 (7%) |
| Moderate | 0 (0%) |
| Severe | 0 (0%) |

Although much is written on its prevention, SAM remains a concern in contemporary degenerative mitral valve repair [15], and such concerns about complexity of repair and SAM likely remain a limiting factor in the uptake of mitral valve repair [18]. We believe large remodeling rings have played a role in our ability to achieve a 100% repair rate for Barlow's disease without postoperative SAM or significant residual MR.

Strengths and Limitations

A unique strength of our study lies in the prospective documentation of valve analysis and repair procedures, and our uniform application of surgical technique [19]. All valve classifications were done at the time of surgery and not based on retrospective chart review. Likewise, we did not rely on echocardiographic reports for postoperative MR grading, but had the studies re-reviewed in a systematic core manner. Our series is one of the most comprehensive surgical documentations of Barlow's disease in contemporary literature. We were necessarily limited by the absence of a control group, ie, we do not know what our experience would have been without the use of large true-sized rings. We were unable to identify any suitable historic, concurrent, or literature controls. Most published series do not define specific cause of degenerative disease and also do not document ring size, preventing useful comparison. As our series is relatively recent, and our study objective was to examine early outcomes (SAM and residual MR), we do not have any mid-term outcomes data. The absence of residual MR (no patients in our series had more than mild residual MR) is, however, encouraging as this is a strong predictor of excellent long-term durability after mitral valve repair [20, 21].

Conclusions

Although we advocate repair for all degenerative valves [22], we acknowledge that Barlow mitral valve repairs using Carpentier's techniques are technically sophisticated and lengthy procedures. In our experience, more than 25% of repairs require cardiopulmonary bypass times in excess of 4 hours, and a majority of cases included prolapse of multiple leaflet segments that required correction. However, as we achieved a 100% success rate (no significant residual MR), with minimal morbidity and no mortality, we believe such complicated mitral valve repairs are worthwhile and are preferable to valve replacement.

As mitral valve repair extends to completely asymptomatic patients (18% in our series), we must aim to guarantee a successful repair in every circumstance, regardless of cause, valve size, and valve lesions; we found the use of true-sized large remodeling annuloplasty rings a useful adjunct to achieving this goal in our series.

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DISCUSSION

DR R. MORTON BOLMAN (Boston, MA): Nice paper, Dr Adams. I would like to ask two questions. One is, this is a complicated problem for all of us, but how do you decide how much anterior leaflet prolapse you can correct with these large rings and how do you decide which ones need direct approaches to the leaflet? And my second question is, why do you use a circumferential ring in this set of patients? I think most people would feel comfortable using a posterior annuloplasty band in this subset of patients. Thank you.

DR ADAMS: The dysfunction of prolapse of the anterior leaflet is not treated by an annuloplasty alone. If there is prolapse of the free margin, a technique such as triangular resection of the leaflet, chordal transfer or Gore-Tex chordoplasty should be performed. The size of the ring is not based on anterior leaflet dysfunction. Ring size is based on the surface area of the anterior leaflet, to ensure effective remodeling.

Your second question related to the choice of a complete ring versus a partial reduction band for Barlow's patients at risk for systolic anterior motion. A review of the literature shows all rings have been associated with systolic anterior motion in the setting of excess leaflet tissue, and in fact, there are just as many reports of SAM with flexible bands as there are with remodeling rings. Indeed some papers have reported SAM without the use of an annuloplasty ring. So the type of ring does not appear to influence the risk of SAM. We believe in complete remodeling rings for Barlow patients because we think the annulus is involved circumferentially in the disease process.

DR EDWARD B. SAVAGE (St. Louis, MO): One of the effects of the ring is not only to support the repair but also to reduce the size of the annulus and restore some of the sphericity of the ventricle, and I wondered if you compared the ventricular volumes of these patients to what would be considered normal for that patient's body surface area?

DR ADAMS: That is a good question. We did not look at ventricular volumes. I can tell you the ejection fractions were unchanged. I think you probably do change the base of the heart to some degree with the use of remodeling rings.

DR JOSÉ LUIS POMAR (Barcelona, Spain): Congratulations for the paper. I have two questions. One, do you think people or surgeons are not properly sizing the ring they are going to implant? Because it is true that you can avoid the problem of SAM with a bigger ring, but most of the time probably they never use more than a 36, and if they do it properly, they should use a 40 mm ring probably. And the other question is whether you think in this particular patient with Barlow's there is also a

dilatation of the intertrigonal distance or it is just because the valve is big itself?

DR ADAMS: Can you repeat your second question?

DR POMAR: Do you think there is an enlargement of the intertrigonal distance in those patients like has been published for some of the cardiomyopathies?

DR ADAMS: José, I would like to answer the second question first. I am not aware of any study that has focused on the intertrigonal distance in patients with Barlow's syndrome, but I agree with Dr Carpentier that it is likely the annulus may be involved circumferentially in the degenerative process. Some patients with excess tissue, however, actually have an increase in the antero-posterior dimension of the anterior leaflet versus the intercommisural distance, and in that case we use a smaller Carpentier Classic ring and bend it to increase the antero-posterior dimension.

It is not often clear from the literature how surgeons size the valve at the time of ring selection, or for that matter how prevalent the use of larger annuloplasty rings actually is. I agree with you that true sizing is fundamental to avoid SAM.

DR SARY F. ARANKI (Boston, MA): David, I have one question for you. For those surgeons who do an excellent leaflet repair and the largest size they have is 36 and they get SAM, what do you advise them to do?

DR ADAMS: Sary, the first thing I would advise them is to put a 38 and a 40 mm ring on the shelf, especially if you are going to operate on young patients with a history consistent with Barlow's disease. If I had implanted a size 36 ring and had SAM, I would make a horizontal incision in the anterior leaflet about 4 mm from the annulus and simply sew it back together. That will shorten the anterior leaflet about 4 mm. If I needed to shorten it more, I would resect an elliptical piece of tissue and add it to the 4 mm, to shorten the anterior leaflet to the desired length.

DR SOON J. PARK (San Francisco, CA): What was your incidence of SAM before you adopted this technique? What was your incidence of SAM before?

DR ADAMS: I am not sure how to answer that. I was influenced by Carpentier's techniques early in my career. Never say never, but thus far I have found shortening the posterior leaflet height to less than 1.5 cm and implanting a true sized remodeling ring an effective strategy to avoid SAM in this subgroup of patients with excess tissue.