

# Barlow's mitral valve disease: results of conventional and minimally invasive repair approaches

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Barlow's valve is a clinically important form of degenerative mitral valve (MV) disease that is characterized by unique clinical, echocardiographic and pathological features. Successful and durable repair of Barlow's MV represents a clinical challenge for most cardiac surgeons. An armamentarium of different MV repair techniques may be required, resectional, neochordal or plicational techniques. Although conventional sternotomy remains the mainstay approach for MV surgery in the majority of cardiac surgery centers, minimally invasive surgery (MIS) is becoming increasingly accepted amongst patients, referring physicians and practicing cardiac surgeons. As surgical approaches, instrumentation and operative experience develop, select centers are now performing MIS MV surgery for nearly all MV patients. Although successful Barlow's MV repair is more complex than that for most degenerative pathologies, several centers have published relatively large series of MIS MV repair for Barlow's disease. In this review article, we highlight and compare the early and long-term results of conventional and minimally invasive approaches to Barlow's and bileaflet mitral prolapse disease. Recent studies from various large volume centers around the world have demonstrated equivalent safety and efficacy outcomes of the MIS approach compared to conventional sternotomy surgery. In addition, MIS MV surgery may allow patients to benefit from a cosmetically appealing incision, a faster recovery and a quicker return to normal activities. However, a definite learning curve has been demonstrated for MIS MV surgery. If a patient with Barlow's disease or other complex MV pathology desires to undergo MIS MV surgery, referral to a center and/or surgeon with extensive experience in MIS MV surgery is recommended.

**Keywords:** Barlow's disease; mitral valve repair; minimally invasive



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## Introduction

Fibroelastic deficiency and Barlow's disease are the two most common etiologies of degenerative mitral valve (MV) disease, often leading to significant mitral regurgitation (MR). Barlow's disease is characterized by pronounced annular dilatation, bileaflet prolapse and/or billowing, hooding, and the presence of thick, spongy leaflets due to excessive myxomatous tissue proliferation with or without calcification (1). Barlow's pathology constitutes a challenge

for surgeons performing MV repair. Achieving a durable surgical result may be a formidable task in these frequently young and otherwise healthy patients.

Minimally invasive (MIS) MV surgery has proven to be feasible and technically acceptable for a wide range of pathologies. Mounting data in the literature supports the hypothesis that MIS can provide at least equivalent results for surgical correction of MR with several associated clinical benefits (2-4). Although MIS should be predominantly reserved for non-complex pathologies (e.g., FED, isolated

P2 prolapse) during the initial part of the surgeon's learning curve (5), it has also been demonstrated that such techniques can be safely and effectively utilized for complex mitral pathologies (e.g., bileaflet prolapse and Barlow's disease) in high volume centers (3,4,6-10). The objective of this paper was to review the available results of MV repair in Barlow's disease and/or bileaflet MV prolapse via conventional versus minimally-invasive approaches.

### Established repair techniques in Barlow's disease

First described by John B. Barlow (11,12), Barlow's disease is characterized by excessive myxomatous tissue—the hallmark of this pathology—as well as annular dilatation, leaflet thickening, bileaflet prolapse, chordal lengthening and, not infrequently, valvular tissue calcification. Additionally, as pointed out by Hutchins (13), Barlow's valve is frequently associated with disjunction of the mitral annulus fibrosus. The resultant atrial displacement of the mitral leaflet attachment may lead to leaflet hypermobility and subsequent excessive mucoid degeneration. Histologically, Barlow's valve is characterized by myxoid infiltration, which destroys the 3-layer leaflet architecture, and collagen alterations (14).

MV repair in Barlow's disease is particularly challenging because of extensive bileaflet billowing, which makes it difficult for the surgeon to find a normal "reference point" and thereby complicates valve analysis and repair planning. Nevertheless, there is a variety of well described surgical techniques for successful repair of this challenging pathology (15). The one consistent element of various Barlow's repair techniques is the use of a large annuloplasty ring. As suggested by Adams *et al.* (16), the definition of a true Barlow's valve should be limited to those requiring a complete ring of size 36 mm or larger. Beyond this one consistent element, a wide variety of leaflet repair techniques have been successfully implemented including leaflet resection, neochord formation, plication and other techniques.

Most of the resectional techniques described for Barlow's and bileaflet MV repair are well established and carry very good long-term results (17). One of the most traditional and well known of such techniques incorporates a complete resection of the middle scallop of the posterior mitral leaflet (PML) followed by a sliding or a folding plasty with the remaining lateral scallops. It might be supplemented furthermore with either a triangular resection of the

anterior leaflet (AML) (especially in the cases with a long, localized AML) or a correction of the AML prolapse with polytetrafluoroethylene (PTFE) chordae or loops (18,19). Secondary MV lesions, such as leaflet clefts and minor commissural prolapses, become apparent upon the water-sealing test. In such instances, the clefts are directly closed with Prolene or Cardionyl 5-0 sutures and the residual commissural prolapse can be corrected either by insertion of more artificial chords or by insertion of a vertical mattress stitch (also known as a "magic stitch"). When encountered, calcifications of the annulus should be removed as proposed by Carpentier (20). Although such resectional techniques are well established, they can be technically challenging to perform through a MIS approach.

Perrier's group first coined the term "respect rather than resect" to describe an alternative to traditional resection techniques (21). The goal of this approach is to correct MV prolapse without excision of leaflet tissue. This can be achieved for the PML with the use of PTFE chordae or Loops, with adjustment of their length so that the PML remains nearly vertical, posterior and parallel to the posterior wall of the left ventricle in the inflow region. This transforms the PML into a smooth, regular and vertical buttress against which the AML will come into apposition. The use of PTFE neochordae has also been described for correction of AML prolapse, with the Loop technique being particularly valuable for MIS surgery (3-10). The Loops to the AML are approximately 10 mm longer than those applied to the PML because of the increased AML mobility that is required to achieve MV competence. One can envision the AML acting like a "door" and the PML as a "doorframe" for the "respect" methods.

The double-orifice technique described by Alfieri's group (22) deserves special mention, as this technique drastically simplifies the repair of the Barlow's valve. In this approach, the edge-to-edge approximation of the middle scallop of the anterior and posterior leaflet allows the elimination of most of the mitral insufficiency, while the residual smaller regurgitant jets are effectively corrected by the association of a ring annuloplasty. One may argue that the Alfieri technique is counterintuitive for a pathology characterized by elongation of both the subvalvular apparatus and the leaflet height. Nevertheless, a properly placed edge-to-edge stitch that encompasses a large amount of leaflet tissue, in combination with a very large complete ring, can lead to a marked shortening of the height of both leaflets and a lowering of the coaptation point to within the left ventricle. Because of its technical simplicity and

**Table 1** Summary of the reported results of conventional and minimally invasive approaches for repair of Barlow's disease or bileaflet mitral prolapse

Author	Approach	Number of patients (etiology)	Follow-up	Freedom from > moderate or $\geq$ moderate MR
David <i>et al.</i> [2005] (24)	Sternotomy	250 (Bileaflet, incl. Barlow's)	12 years	67%
Flameng <i>et al.</i> [2008] (25)	Sternotomy	83 (Barlow's) and 265 (FED)	5 years; 10 years	82.2%; 64.9%
Jouan <i>et al.</i> [2012] (26)	Sternotomy	200 (Barlow's)	8 years	90.2%
Castillo <i>et al.</i> [2013] (27)	Sternotomy, lower hemi-sternotomy	110 (Barlow's) and 78 (FED)	1 year; 4 years; 7 years	100%; 93.7%; 90.3%
Lapenna <i>et al.</i> [2005] (23)	MIS	48 (Barlow's)	22.7 $\pm$ 10.6 months	99.9%
Speziale <i>et al.</i> [2011] (28)	Sternotomy vs. MIS (Randomized)	70 (Barlow's); 70 (Barlow's)	12.4 months	97% (P=0.9); 98% (P=0.9)

MR, Mitral regurgitation; MIS, Minimally invasive surgery; FED, Fibroelastic deficiency.

reproducibility, this technique can be easily applied through a MIS approach (23).

### Repair outcomes in conventional approaches

In spite of challenges presented by MV repair in Barlow's disease, referral of these cases to selected centers has helped MV surgeons around the world to obtain excellent short- and long-term results for this difficult pathology. *Table 1* summarizes the outcomes of the largest series for MV repair in Barlow's disease or bileaflet prolapse via conventional sternotomy or MIS approaches. David and associates (24) reviewed their results in 701 patients with mitral prolapse, of which 250 patients were operated on for bileaflet prolapse including Barlow's disease. They report freedom from moderate or severe MR rates at 12 years of 80% $\pm$ 4% for posterior, 65% $\pm$ 8% for anterior, and 67% $\pm$ 6% for bileaflet prolapse.

Another series by Flameng *et al.* (25) describes a series of 348 patients who have undergone MV repair for degenerative MR. In this series comprising patients with Barlow's disease (n=83) and fibroelastic deficiency (n=265), freedom from recurrent MR (>2+) was 82.2% at five years and 64.9% at ten years for the entire group. However, recurrent MR was much more common in patients with Barlow's disease with a linearized recurrence rate of 6.0% per year (compared to 2.3% for fibroelastic deficiency).

Jouan *et al.* (26) described 200 patients with Barlow's

disease who underwent MV repair via a conventional sternotomy. A successful repair was achieved in 94.7% (179/189) of non-redo patients. Operative mortality was 1.5% (n=3) and the overall survival at eight years was 88.6% $\pm$ 3.1%. The freedom from MV reoperation at eight years was 95.3% $\pm$ 1.7% and the freedom from late recurrent MR (>2+) was 90.2% $\pm$ 3.1%.

Castillo, Adams *et al.* (27) described their results in 188 consecutive patients who underwent surgery for degenerative anterior or bileaflet mitral leaflet prolapse (Barlow's disease in 110 patients and fibroelastic deficiency in 78 patients). Freedom from more than moderate MR was 100% at one year, 93.7% $\pm$ 2.2% at four years, and 90.3% $\pm$ 3.7% at seven years. All procedures were performed through either a median sternotomy or a lower hemisternotomy approach.

### Repair outcomes in minimally invasive approaches

MIS MV surgery has slowly gained increasing acceptance within the cardiac surgical community and is experiencing a growing demand among patients and referring physicians. In Germany, over 40% of patients currently undergoing MV surgery are operated on with a MIS approach (29). MIS facilitates earlier resumption of normal activities, which is associated with improved cosmesis, and may reduce postoperative pain, blood loss, and hospital length of stay (4,7-10,30,31).

A MIS approach is defined as an operation utilizing a

chest wall incision other than a median sternotomy (32). Most commonly employed approaches include right mini-thoracotomy, right thoracic incisions for the robotically-assisted surgery and partial sternotomy. At the vast majority of centers employing a MIS approach to MV repair, including our own, the standard approach is via a small (5-7 cm in length) anterolateral mini-thoracotomy as described in detail elsewhere (2,18). Briefly, cardiopulmonary bypass (CPB) is instituted via femoral arterial and venous cannulation. The aorta is cross-clamped with a Chitwood clamp and myocardial protection is achieved with mild hypothermia (34 °C) and antegrade delivery of cardioplegia. The left atrium is then opened posterior to the interatrial groove and a left atrial retractor is used to expose the MV. The stepwise approach to the Barlow's valve repair is similar to that used during conventional sternotomy (15). Essentially, the entire spectrum of repair techniques can be utilized via a MIS approach including ring annuloplasty, leaflet resection with or without sliding annuloplasty, the "Loop" technique with premeasured Gore-Tex neochordae, chordal transfer, commissural plication, the edge-to-edge ("Alfieri") technique, cleft closure and calcium debridement. The edge-to-edge technique and the use of artificial chords (including the "Loop" technique) to correct leaflet prolapse are simpler and less technically challenging to perform, and therefore are more frequently utilized in MIS Barlow's operations. Several studies have reported on the results of MIS approaches to Barlow's MV repair. Lapenna *et al.* (23) described 48 such patients, all of whom underwent an "edge-to-edge" repair via a small right anterolateral thoracotomy and peripheral femoral vessel cannulation for CPB. MV repair was successful in 100% of patients. At a mean follow-up time of 22.7±10.6 months, no residual MR was detected on echocardiography in 33 (68.7%) patients and mild insufficiency was found in 15 (31.2%). At the time of follow-up, survival rate and freedom from reoperation was 100% and all patients were in NYHA class I.

Speziale *et al.* (28) randomized patients with Barlow's disease to undergo conventional open repair via median sternotomy or MIS repair. In both groups (each group comprised of 70 patients), MV repair was performed using Gore-Tex neo-chordae for chordal reimplantation on both leaflets. MV repair was successful in 98.5% of MIS patients and 100% of median sternotomy patients. Although operative and CPB times were longer in MIS patients, no differences in safety or efficacy outcomes were observed between the two groups, including freedom

from reoperation (100% in each group) and freedom from moderate or worse MR (98% *vs.* 97%) at 1-year follow-up.

We have also recently examined our outcomes for MIS surgery in patients with Barlow's disease (33). A total of 145 Barlow's patients underwent MIS MV surgery at our institution over an 11-year period. MV repair was successfully performed in 95% of patients using Loop neochordae in 72%, PML resection in 30%, Alfieri stitch in 18%, commissural plication in 9%, chordal transfer in 9% and AML resection in 7% of patients. The perioperative rates of morbidity and mortality were very low. Long-term follow-up revealed a ten-year survival rate of 88%, while ten-year freedom from MV reoperation or recurrent (>2+) MR amongst survivors was 90% and 88%, respectively. Such results confirm that Barlow's pathology can be successfully treated with MIS surgery using a variety of surgical techniques.

## Conclusions

Growing evidence demonstrates that the results of the MIS approach for MV surgery are at least as good as those achieved with conventional surgery, with several clinical advantages over a full sternotomy approach. One critique of MIS surgery has been its suitability to treat complex MV pathology, such as Barlow's disease. We herein describe very good early and long-term results for the MIS approach in Barlow's patients operated on in select, large volume centers. The fact that these case series come from centers with a large MIS MV repair experience should not be overlooked, since we and others have clearly shown a learning curve effect associated with MIS surgery (5). Complex MV pathologies such as Barlow's valve should not be attempted during the early portion of the MIS learning curve.

Patients with severe MR due to Barlow's disease are often asymptomatic and may be reluctant to undergo a median sternotomy. The small right anterolateral incision is not only cosmetically appealing, but also allows patients to return faster to an active life. A multitude of MV repair techniques can be employed via the MIS approach, although the edge-to-edge and neochordae/Loop technique are technically easier through a small incision.

Prêtre (34) once noted that "one should never lose sight of our cardinal priority: the cosmetic must be achieved primarily on the heart, not on the skin". Undoubtedly, the success rate and durability of the MV repair should be the first priority in patients with MR. However, the evolution

of technology, the increasingly widespread adoption of MIS valve surgery, the dissipation of knowledge within the cardiac surgery community, and the increase in the cardiac surgeon's armamentarium of MV repair techniques have all made it possible to achieve excellent results through a MIS approach, even in patients with complex mitral pathology. We must also acknowledge the fact that the rapid advancements in transcatheter valve techniques are likely to result in an even larger proportion of patients requesting MIS surgery in the foreseeable future.

### Summary

MIS MV repair is feasible in patients with Barlow's disease. Given the previously demonstrated clinical benefits of MIS surgery, this approach may be strongly considered for patients presenting with Barlow's disease. However, the MIS approach is associated with a definite learning curve and, therefore, patients with complex MV pathology, including Barlow's, should be referred to centers with established experience in complex MIS MV repair.

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