Sustained improvement after combined anterior mitral valve leaflet retention plasty and septal myectomy in preventing systolic anterior motion in hypertrophic obstructive cardiomyopathy in children

Eva Maria B. Delmo Walter, Henryk Siniawski, Roland Hetzer

Department of Cardiothoracic and Vascular Surgery, Deutsches Herzzentrum Berlin, Augustenburger Platz 1, 13353, Berlin, Germany

Received 4 September 2008; received in revised form 16 March 2009; accepted 23 March 2009; Available online 22 May 2009

Abstract

Objective: Anatomic alterations of the mitral valve such as increased mitral leaflet area, length and laxity, and anterior displacement of the papillary muscles in hypertrophic obstructive cardiomyopathy predispose patients to residual systolic anterior motion and persistence of outflow obstruction and mitral regurgitation after septal myectomy. We investigate the long-term results of combined anterior mitral leaflet retention plasty and septal myectomy in children with hypertrophic obstructive cardiomyopathy. Methods and results: Anterior mitral leaflet retention plasty and subaortic septal myectomy were performed in 12 children (mean age 10.8 ± 1.7 years) with hypertrophic obstructive cardiomyopathy. Mean preoperative left ventricular outflow tract pressure gradient was 49 ± 11 mmHg. After careful assessment of the mobility of the anterior leaflet and subvalvular apparatus, segments of the anterior leaflet nearest the trigones were sutured to the corresponding posterior annulus with polypropylene reinforced with untreated autologous pericardial pledgets. Intraoperative valve orifice measurement based on age-related valve leaflet and subvalvular apparatus, segments of the anterior leaflet nearest the trigones were sutured to the corresponding posterior annulus with polypropylene reinforced with untreated autologous pericardial pledgets. Intraoperative valve orifice measurement based on age-related valve leaflet and subvalvular apparatus, segments of the anterior leaflet nearest the trigones were sutured to the corresponding posterior annulus with polypropylene reinforced with untreated autologous pericardial pledgets. Intraoperative valve orifice measurement based on age-related valve leaflet and subvalvular apparatus, segments of the anterior leaflet nearest the trigones were sutured to the corresponding posterior annulus with polypropylene reinforced with untreated autologous pericardial pledgets. Intraoperative valve orifice measurement based on age-related valve leaflet and subvalvular apparatus, segments of the anterior leaflet nearest the trigones were sutured to the corresponding posterior annulus with polypropylene reinforced with untreated autologous pericardial pledgets. Intraoperative valve orifice measurement based on age-related valve leaflet and subvalvular apparatus, segments of the anterior leaflet nearest the trigones were sutured to the corresponding posterior annulus with polypropylene reinforced with untreated autologous pericardial pledgets. Mean follow-up is 11.85 ± 1.22 years. Mean left ventricular outflow tract pressure gradient was 6.2 ± 3.95 mmHg. No mortality, no repeat myectomy or repeat mitral valve repair or replacement, no mitral stenosis and no systolic anterior motion occurred. Conclusions: Long-term follow-up shows sustained absence of systolic anterior motion, attenuation of mitral regurgitation, sustained improvement in functional status, and reduction of outflow tract obstruction.

Keywords: Hypertrophic obstructive cardiomyopathy; Left ventricular outflow tract obstruction; Septal myectomy; Systolic anterior motion; Mitral valve repair

1. Introduction

Hypertrophic obstructive cardiomyopathy (HOCM), a primary genetic myocardial disease [1] characterized by asymmetrically distributed left ventricular hypertrophy and left ventricular outflow tract (LVOT) obstruction is heterogeneous [2] in terms of natural history, clinical manifestations, hemodynamic characteristics, age of onset and risk for sudden death [3]. Variable degrees of LVOT obstruction induced by thickening of the interventricular septum as well as due to systolic anterior motion (SAM) of the anterior mitral valve leaflet are present. SAM is responsible for concomitant mitral regurgitation typically directed postero-laterally into the left atrium [4]. HOCM often presents with anatomic changes of the mitral valve such as increased mitral leaflet area, elongation and pronounced laxity, as well as anterior displacement of the papillary muscles [5–7]. These structural abnormalities are not corrected by septal myectomy alone, and thus patients develop propensity to a residual SAM, persistence of outflow obstruction and mitral regurgitation after septal myectomy leading to a suboptimal outcome [8,9]. The standard therapeutic intervention for children, is left ventricular septal myectomy, also known as Morrow procedure [1–3,10,11]. Several invasive therapeutic modalities such as percutaneous transluminal septal myocardial ablation (PTSMA) have been developed to diminish the LVOT obstruction by reduction of the interventricular septum width or thickness [4,12]. There have been few reports in surgical management of mitral valve in children with HOCM [3]. Mitral valve reconstruction techniques complementing septal myectomy to prevent postmitral repair SAM such as triangular resection of the anterior leaflet [13] sliding plasty of the posterior leaflet [14] edge-to-edge technique [15] anterior mitral leaflet extension with a pericardial patch [4,12,16] radical debridement and repositioning of the papillary muscles [2] as well as complete excision of secondary chordae of the anterior mitral leaflet...
may be useful. Mitral valve replacement is not desirable in children, because of its attendant morbidities as well as lack of suitable age-prosthetic sizes [17].

We performed anterior mitral leaflet retention plasty (ALRP) in combination with septal myectomy in children with HOCM and we investigated the long-term results of these combined procedures in 12 children.

2. Patients and methods

2.1. Patients

Between September 1994 and December 2008, 12 patients (mean age 10.8 ± 1.7 years, median 13.06 years, range 0.52—17 years) underwent surgery for HOCM. The diagnosis of HOCM was based on clinical evaluation and on echocardiography. In all these patients, the indications for operation were refractory symptoms despite optimal medical therapy consisting of β-blocking agents, calcium channel blockers, or both. Duration of treatment ranged from 2.8 months to 8.3 years. At the time of surgery, 3 (25%) patients were in New York Heart Association class II, 5 (41.6%) patients in class III, and 4 (33.3%) in class IV. These patients had a mean maximal preoperative LVOT pressure gradients of 96.83 ± 9.6 mmHg (median 84.5 mmHg, range 50—160 mmHg) and mean preoperative LVOT pressure gradient of 49 ± 11 mmHg (range 38—60 mmHg). All had associated moderate to severe mitral insufficiency, and one had associated ventricular septal defect. Eight patients had a history of ventricular arrhythmia. All patients had no previous surgery.

3. Surgical technique

3.1. Subaortic septal myectomy

Intraoperative transesophageal echocardiography (TEE) is performed after induction of general anesthesia, with particular attention to the cardiac anatomy, mitral valve morphology and function, and thickness of the ventricular septum. Exposure is gained through a median sternotomy and intracardiac pressures were measured simultaneously in the left ventricle and aorta. If the left ventricular outflow tract (LVOT) gradient is low (<30 mmHg) because of anesthesia, isoproterenol is administered or premature ventricular contractions are induced to determine the maximal gradient. Standard cardiopulmonary bypass with moderate hypothermia (28—32°C) was used and the left heart is vented. During aortic occlusion, myocardial protection, especially important because of the severe ventricular hypertrophy, was given through a generous infusion of antegrade cold crystalloid cardioplegia, followed by additional doses administered selectively onto the right and left coronary ostia every 15—20 min. An oblique aortotomy was carried rightward down to the non-coronary sinus and towards the aortic annulus. The aortic valve is inspected and the subvalvular region is exposed. Optimum visualization of the ventricular septum is facilitated by posterior displacement of the left ventricle. Aside from using Morrow technique, which used two parallel longitudinal incisions in the septum, we made parallel incisions into the septum directly opposite the anterior mitral leaflet. The resection of long blocks of septal myocardium from between the two incisions is started just below the aortic annulus of the right coronary sinus and the commissure between the right and the left coronary sinuses (Hetzer’s technique, Fig. 1A) [17]. Importantly, the incision is continued apically beyond the point of mitral—septal contact, marked by a fibrous band. This wide incision beneath the valve improves exposure of the important area toward the apex. Intraoperative pre-septal myectomy pressure gradient ranged from 40 to 105 mmHg (mean 60 ± 25 mmHg), and post-septal myectomy gradient ranged from 0 to 18 mmHg (mean 5 ± 6 mmHg).

3.2. Anterior leaflet retention plasty (ALRP)

Access to the mitral valve is gained through a left atriotomy along the interatrial groove. After carefully assessing the morphology and mobility of the anterior mitral valve leaflets and the subvalvular apparatus, the segments of anterior mitral leaflet closest to the trigones are sutured to the corresponding posterior annulus wherein the polypropylene mattress sutures are pledged with untreated autologous pericardium. These sutures are passed through the coaptation line of the anterior leaflet and the corresponding posterior annulus trigone (Hetzer’s technique, Fig. 1B and C) [17]. Thus, the mobility of the anterior mitral valve leaflet is limited in its trigone-near segment, thereby unable to produce systolic anterior motion and mitral insufficiency (Fig. 1D and E) [17].

Fig. 1. (A) Hetzer’s septal myectomy (aortic view) opposite anterior mitral valve leaflet (dashed lines indicate myocardial septal incisions), (B) Hetzer’s ALRP for hypertrophic obstructive cardiomyopathy (HOCM) and systolic anterior motion (SAM), (C) Completed repair (atrial view), (D) mitral insufficiency in HOCM and SAM before repair and (E) redirection of mitral insufficiency following septal myectomy and ALRP.
In these series, laxity of the anterior mitral valve leaflets was present, but no distinct abnormalities of the chordae tendineae and papillary muscles were apparent. This repair was routinely guided by intraoperative TEE with particular attention to the septal anatomy and thickness, and mitral valve function, mobility as well as anatomy of the subvalvular apparatus.

After the septal myectomy and ALRP, the aortic and mitral valves were inspected to ensure that they have not been injured. After weaning the patient from cardiopulmonary bypass, pressures are remeasured in the left ventricle and aorta and TEE evaluation is repeated, with special attention to the width of the interventricular septum, the residual LVOT gradient, mitral valve regurgitation and SAM. If myectomy has been successful, there will be little or no residual gradient, and little or no SAM of the mitral valve. In general, we would resume cardiopulmonary bypass for resection, if the gradient were greater than 15—20 mmHg. Post-myectomy mitral insufficiency was reduced to a regurgitant fraction of 0—10%. There was no early or late mortality, or reoperation for repeat myectomy or repeat mitral valve repair or replacement.

Saline injection through the valves and intraoperative TEE were routinely performed to assess the adequacy of repair. Postoperative transthoracic echocardiography was carried out annually, or if clinically indicated on the basis of symptoms. The degree of mitral valve regurgitation was estimated by means of standard echocardiographic measurement techniques.

3.3. Follow-up

Follow-up data were provided by both the Department of Congenital Heart Disease/Pediatric Cardiology and Department of Clinical Studies, Deutsches Herzzentrum Berlin and written correspondence from the referring physicians, and/or telephone interviews with patients or families. No patients were lost to follow-up. The mean follow-up period was 11.85 ± 1.22 years. The end of the follow-up study was December 2008.

3.4. Statistical analysis

All data were analyzed with the SPSS 16.0 (SPSS Inc., Chicago IL, USA) software program. The data are expressed as absolute and percentage frequency values and continuous data as mean ± standard error of the mean, as appropriate. Freedom from reoperation was analyzed according to Kaplan—Meier estimates with 95% CI. Since there is no mortality, no Kaplan—Meier survival curve can be plotted.

4. Results

Demographic profiles of the 12 patients are shown in Table 1. Five (41.6%) and 4 (33.3%) had class III and IV symptoms, respectively, according to NYHA classification.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age at surgery, ±SD (range)</td>
<td>10.8 ± 1.7 years (0.52—17 years)</td>
</tr>
<tr>
<td>Sex (male/female)</td>
<td>9/3</td>
</tr>
<tr>
<td>NYHA Class, n (%)</td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>3 (25)</td>
</tr>
<tr>
<td>III</td>
<td>5 (41.6)</td>
</tr>
<tr>
<td>IV</td>
<td>4 (33.3)</td>
</tr>
<tr>
<td>Symptoms</td>
<td></td>
</tr>
<tr>
<td>Dyspnea</td>
<td>5 (41.6)</td>
</tr>
<tr>
<td>Angina</td>
<td>1 (8.3)</td>
</tr>
<tr>
<td>Syncope</td>
<td>6 (50)</td>
</tr>
<tr>
<td>Medical therapy</td>
<td></td>
</tr>
<tr>
<td>B-blocker</td>
<td>4</td>
</tr>
<tr>
<td>Calcium antagonist</td>
<td>5</td>
</tr>
<tr>
<td>Combination</td>
<td>3</td>
</tr>
<tr>
<td>Associated cardiac disease</td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>1 (8.3)</td>
</tr>
</tbody>
</table>

Table 1. Demographic profile of the 12 children with HOCM treated with septal myectomy and ALRP.

Mean preoperative interventricular septal thickness was 24.2 ± 3.1 (range 17—33 mm) and mean left ventricular posterior wall thickness was 25.6 ± 4 (range 18—27 mm). The preoperative mean maximal LVOT gradient was 96.83 ± 9.6 (range 50—160) mmHg. Most patients had moderate to severe mitral regurgitation (2.82 ± 0.18). All patients had typical marked SAM of the anterior mitral valve leaflet (mean grade 3.08 ± 0.61). Only 1 (8.3%) had an associated ventricular septal defect, in whom direct closure was performed during the septal myectomy and ALRP procedure.

4.1. Perioperative outcome

Preoperative, immediate postoperative as well as the latest follow-up echocardiographic data are presented in Table 2. Mean cardiopulmonary bypass time is 111.58 ± 16.69 (range 54—267) min. Mean ischemic time is 62.25 ± 7.9 (range 18—118) min. Post-myectomy mitral insufficiency was reduced to a regurgitant fraction of 0—10%. There was no early or late mortality, or reoperation for repeat myectomy or repeat mitral valve repair or replacement. Eight (66.6%) patients had concomitant epicardial pacemaker implantation for preoperatively diagnosed atrioventricular block II (n = 5), atrial fibrillation (n = 2) and supraventricular tachycardia (n = 1), unresponsive to medical treatment.

The mean follow-up period was 11.85 ± 1.22 years (range 9 months to 15 years). During the follow-up period, patients were either asymptomatic (33.3%) or had mild symptoms (50% and 16% in NYHA functional class I and II, respectively).

The number of cardiac drugs prescribed decreased significantly; before intervention, the number of cardiac drugs used was 1.25 ± 0.3 whereas after surgery, and during follow-up, it was significantly reduced. One of the eight patients with permanent cardiac pacemaker underwent replacement of the pacemaker generator system 12 years after its implantation.

In relation to the preoperative data, we found a mean reduction in septal thickness of 10.4 ± 0.5 mm after septal myectomy and further reduced by 1.9 ± 0.8. Moreover, the...
LVOT gradient as well as mitral regurgitation was significantly reduced after surgery and further reduced during the succeeding follow-up period. The most significant finding is that there is no SAM immediately postoperative and during the latest follow-up. During each of the latest patients’ follow-up, these results were sustained.

Preoperative and postoperative echocardiograms from one patient after septal myectomy and ALRP are shown in Figs. 2—5.

5. Discussion

Surgery in pediatric patients with HOCM was technically challenging because of the difficulty of exposure of the smaller structures. We developed an alternative surgical procedure concomitant with septal myectomy, which aims to reduce or even prevent the residual outflow obstruction due to SAM of the mitral valve. Transaortic septal myectomy presents special problems in young patients because of limited exposure through the aortic annulus. Likewise, special care must be taken to avoid injury to the delicate aortic cusps during retraction for exposure of the hypertrophied septum. Too deep a resection will create a ventricular septal defect [9,18] or even ventricular perforations, and the aortic valve is always at risk for injury from instruments passed through the valve and manipulated within the ventricle. Another special technical consideration in children relates to accurately judging the depth and length of the septal myectomy. Inadequate excision will leave residual LVOT obstruction and limit symptomatic improvement and survival. In our experience, adequate myectomy may be achieved in essentially all patients, but careful hemodynamic assessment after weaning from the cardiopulmonary bypass is obligatory for optimal results. Reinstitution of bypass and resection of additional septal muscle should be considered if the residual systolic peak gradient exceeds 20 mmHg. In the present series, there was an immediate 88% reduction in intraoperative LVOT gradient to a mean residual gradient of 6 mmHg after septal myectomy. The relief of LVOT obstruction was maintained in all 12 patients at latest follow-up; the mean maximum echocardiographic data

<table>
<thead>
<tr>
<th></th>
<th>Preoperative</th>
<th>Immediate postoperative</th>
<th>Latest follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Clinical data</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>NYHA class</td>
<td>1.08 ± 0.61</td>
<td>0.5 ± 0.4</td>
<td>0.27 ± 0.3</td>
</tr>
<tr>
<td>No. of drugs</td>
<td>1.25 ± 0.3</td>
<td>0.5 ± 0.4</td>
<td>0.48 ± 0.4</td>
</tr>
<tr>
<td><strong>Echocardiographic data</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IVS (mm)</td>
<td>24.2 ± 3.1</td>
<td>13.4 ± 2.6</td>
<td>11.5 ± 1.8</td>
</tr>
<tr>
<td>LV PW mean thickness (mm)</td>
<td>25.6 ± 4</td>
<td>15.5 ± 2</td>
<td>10.08 ± 2</td>
</tr>
<tr>
<td>LVOT mean pressure gradient (mmHg)</td>
<td>49 ± 11 (range 38—60)</td>
<td>6.2 ± 3.95 (range 0—19)</td>
<td>5 ± 6 (range 0—10)</td>
</tr>
<tr>
<td>LVOT maximum pressure gradient (mmHg)</td>
<td>96.83 ± 9.6 (range 50—160)</td>
<td>28.5 ± 5.2 (0—50)</td>
<td>12.78 ± 4.89 (range 0—37)</td>
</tr>
<tr>
<td>Mitral regurgitationa</td>
<td>2.82 ± 0.18</td>
<td>0.64 ± 0.15</td>
<td>0.8 ± 0.5</td>
</tr>
<tr>
<td>Systolic anterior motionb</td>
<td>3.08 ± 0.61</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>LA, mm</td>
<td>38.5 ± 5</td>
<td>37.3 ± 9</td>
<td>32.6 ± 8</td>
</tr>
</tbody>
</table>

a Graded based on a scale of 0—4.
b Graded based on a scale of 0—3.

Fig. 2. (A) Preoperative parasternal long axis view in patients with HOCM; S, septum; PW, posterior wall; white arrow indicate left ventricular outflow tract dimension (LVOT), yellow arrow indicate presence of SAM phenomenon. (B) The same patient after surgery; parasternal long axis view. White arrow shows LVOT dimension and yellow arrow indicate normal coaptation of anterior and posterior mitral leaflet (no SAM).

Fig. 3. Preoperative color Doppler picture of parasternal long axis view. Yellow arrow indicates SAM and white arrow demonstrates severe mitral regurgitation as a consequence of SAM. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)
graphic maximum pressure gradient of 12.78 ± 4.89 mmHg was reduced by 87% from the preoperative mean maximal gradient of 96.83 ± 9.6 mmHg. There was no incidence of recurrent LVOT obstruction with our described technique of septal myectomy.

Besides the myocardium, the anatomy of the mitral valve apparatus also frequently appears anomalous adding to the obstruction component. Normally the mitral valve leaflets coaptate below the outflow tract as they are pulled posteriorly by the papillary muscles, where they do not impede the outward-directed blood flow. Known elements that may disrupt this coaptation mechanism are valve elongation and anterior displacement of the papillary muscles, which predispose to systolic anterior motion. Consequently, outflow tract obstruction and mitral regurgitation may persist after successful surgical myectomy. To counteract residual SAM, several authors [19–21] reported mitral valve replacement as an alternative procedure in patients regarded as less suitable candidates for septal myectomy. In our institution, mitral valve reconstruction is the preferred technique for any kind of mitral valve disease in infants, children and adolescents, in the face of the complete lack of a prosthesis suitable for this age group, aside from avoiding the need of valve replacement with all its drawbacks [17]. While we agree that the mitral valvular dysfunction is an integral factor in producing outflow obstruction, replacement of the valve is not essential either for reduction of the outflow gradient or alleviation of mitral insufficiency.

In this present study, we report our collective cases of 12 consecutive patients with HOCM associated with moderate to severe mitral valve insufficiency caused by systolic anterior motion. Our preferred approach was transaortic septal myectomy to address the left ventricular outflow tract obstruction (LVOTO), with concomitant anterior leaflet reten-

plasty performed through a left atriotomy. These combined procedures effectively abolished the LVOTO, SAM, and mitral insufficiency was absent or mild in 97% of patients. Clinical and hemodynamic parameters including functional class, reduction of the LVOT gradient, and attenuation of mitral regurgitation and SAM showed sustained improvement during long-term follow-up. In all patients, treated with this technique, SAM was virtually non-existent. It is important to realize that the severity of mitral regurgitation is related to the presence of SAM [22]. It is very important to evaluate the presence of abnormalities of the subvalvular apparatus in HOCM as well, since failure to recognize and treat it may be fatal or lead to incomplete relief of obstruction [23]. Although these are absent in this series, the most important of these anomalies is anomalous papillary muscle insertion directly onto the anterior mitral leaflet, extensive fusion of papillary muscles with the ventricular septum or left ventricular free wall, abnormal chordae tendineae (false chords) that attach to the ventricular septum or free wall, and accessory papillary muscles, all of which may tether the mitral leaflets towards the septum and produced LVOTO. This would contraindicate anterior leaflet retention plasty.

We address the prolapsing segment or unsupported anterior leaflet by avoiding the use of an annuloplasty ring. Anterior leaflet retention plasty combined with septal myectomy is excellent to restrict mitral valve motion and to allow for more complete relief of subaortic obstruction and mitral insufficiency, and avoiding SAM. Symptomatic improvement of the 12 patients was gratifying and all improved by at least one functional class, and remain improved on each patient’s latest follow-up. There has been no occurrence of mitral stenosis following ALRP in this series.

We regard ALRP as a safe and excellent addition to septal myectomy because there was not even a single complication that occurred. In addition, SAM was completely abolished.

5.1. Mechanism of ALRP

In HOCM, the abnormal motion of the anterior mitral valve leaflet in systole plays a key role in creating the outflow obstruction. The pull and push mechanism of the leaflets due to either increased mitral leaflet length and laxity allow the valve to protrude in the outflow tract. Accordingly, inward displacement of the papillary muscles towards each other results in chordal slack in the central leaflet portion and consequently SAM in the central portion of the valve [6]. To counteract these forces, the segments of anterior mitral leaflet closest to the trigones are sutured to the correspond-

Fig. 4. (A) Preoperative echocardiogram: short axis with narrow LVOT obstructed by subvalvular apparatus (yellow arrow), white arrow indicates septum thickness (30 mm). (B) The same projection in the same patient after successful surgery. It shows widely opened outflow tract (yellow arrow) and septum (white arrow) after resection with maximum 11 mm in this area. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)

Fig. 5. (A) Postoperative echocardiogram of five chamber view with CW Doppler registration. (B) Indicate small mean gradient (8 mmHg) across LVOT.
coaptation line of the anterior leaflet and the corresponding posterior annulus trigone. Hence, the mobility of the anterior mitral valve leaflet becomes limited in its segment near the trigone, unable to produce systolic anterior motion and mitral insufficiency. Likewise the normal anatomy of the mitral valve is maximally restored. By suturing the segments of anterior mitral leaflet closest to the trigones to the corresponding posterior annulus, we restrict the buckling part of the anterior leaflet, restoring the normal mobility. This decreases the length of the leaflet which may cause a lateral shift of the chordae attaching prolapsed portion of the leaflet, which in turn stretches the chordae, erects them, and enhances leaflet coaptation.

Decreasing the laxity of the anterior mitral leaflet could also add positively to the maintenance of valve tethering once the streams of flow are normalized after septal myotomy, wherein the flow straightens, hitting the mitral valve onto the anterior surface and pressing the anterior leaflets towards the left atrium. The force that directs the enlarged valve posteriorly is linear to the leaflet area exposed to that flow. In fact, the retained anterior leaflet appears tightened in systole, is directed toward the left atrium, and SAM and mitral regurgitation are abolished. This approach is designed to avoid mitral valve replacement, and hemodynamic results are encouraging. The selection criteria for retention plasty included not only a relatively thick ventricular septum, presence of an abnormal mitral valve apparatus and prolapse of anterior leaflet, but also a persistent obstruction after myectomy. In the present study, we did not treat previously operated patients. Also, we could not compare ALRP to the results with valve replacement, because we generally perform mitral valve repair and reconstruction in children [17] especially in those with HOCM.

6. Conclusions

ALRP in combination with septal myotomy is an effective and safe treatment for children and adolescents with HOCM. Long-term follow-up shows sustained absence of systolic anterior motion, attenuation of mitral regurgitation, sustained improvement in functional status, and absence or reduction of outflow tract obstruction. Because the present technique offers a broadening of the surgical possibilities, we believe that ALRP could become the preferred choice in persistent LVOT obstruction and SAM.

Acknowledgment

We thank Anne Gale, medical editor, for assistance with this article. We also appreciate the assistance of Astrid Benhennour, Christine Detschades, Daniela Moeske-Scholz, Heike Schultz, Julia Stein, Karla Weber, Helge Haselbach and Thomas Farr.

References

Appendix A. Conference discussion

Dr Sertac Çiçek (Istanbul, Turkey): Hypertrophic cardiomyopathy is usually not recognized until adulthood, and presentation in childhood is associated with high mortality and morbidity. Most of the patients referred for surgery have severe obstruction of the left ventricular outflow tract that is associated with systolic anterior motion. Systolic anterior motion, which was once believed to be resulting from the Venturi effect, increases the severity of the obstruction and causes mitral regurgitation. Though, the picture is more complicated than this and mitral valve apparatus abnormalities are common, including elongated and enlarged mitral leaflets as stated by the authors.

Surgical myectomy alone successfully relieves obstruction and resolves mitral regurgitation in the great majority of patients. The impact of the mitral valve abnormalities on surgical decision-making and surgical technique is poorly defined because of small cohort size, focus on individual abnormalities, and focus on autopsy series.

Restricting leaflet motion by retention plasty is an interesting concept and simple technique that could be reproduced easily and seems effective. However, obstruction due to systolic anterior motion mainly originates from the A2 component of the anterior leaflet which could easily be seen by the location of the septal mitral contact lesion. Plicating anterior mitral leaflet through the aortic valves makes more sense if this is the case. If A2 component is not included, one may surmise that myectomy alone would suffice to restore the left ventricular outflow tract geometry and relieve systolic anterior motion. It is very difficult to conceptualize any beneficial effect of an intervention that is directed to A1 and A3 when the culprit is obviously A2 component which is left untouched by the present technique.

The second point I would like to make is, although preoperative arrhythmias are common — you have not presented the data here, but you have sent me the full manuscript including this data — preoperative complete heart block is extremely rare association and 5 patients in this series have reported to have pacemakers implanted concomitantly. Add into this, 2 patients with atrial fibrillation and 1 patient with ventricular arrhythmia, 8 patients out of 12 had a pacemaker placed concomitantly. This is completely different from our practice and, to my knowledge, there are only very few cases of this combination reported.

Finally, although the late mortality is reduced by surgical myectomy, sudden deaths are not entirely prevented and it is important to identify the patients who are at high risk. Patients with history of cardiac arrest pose special risk and placement of an implantable cardioverter defibrillator is highly recommended for secondary prevention of sudden death. You have at least one such patient.

In the light of foregoing, I have three questions.

1. How did you study these patients preoperatively and was the complete heart block presenting symptom?
2. Could you speculate about etiology and mechanisms of complete heart block, incidence of complete heart block on this series, and what is your approach with atrial fibrillation and hypertrophic cardiomyopathy? Do you use ablation?
3. What are the current recommendations of your group regarding concomitant implantable cardioverter defibrillator implantation for this group of patients?

Dr Delmo Walter: With regards to patients where we concomitantly implanted pacemaker, those were patients who were diagnosed preoperatively with AV block, and/or atrial fibrillation. During surgery, we took the opportunity to implant the needed pacemaker. Because these are all children and not adults, we don’t like to subject them to another operation.

And then on your question about the ICD, yes, of course, we’ve been implanting such on our patients, as previously stated, even before the AICD implantation in children has become known. I think I should write that also in my manuscript.

Dr A. Corno (Liverpool, United Kingdom): The quantification and degree of mitral valve regurgitation is largely dependent on several variables, particularly on the mismatch between the observer, his/her experience and the echocardiographic technique, and the situation of the patient at the moment of mitral valve evaluation in terms of preload, afterload and contractility. These parameters are extremely variable immediately postoperative period. What kind of criteria did you define to avoid this possible mistake of comparison between preoperative and postoperative mitral valve function?

Dr Delmo Walter: Only one highly experienced echocardiographer was doing all echocardiographic studies preoperatively, intraoperatively and postoperatively, and doing follow-up, on all our study patients. So there is a uniform criteria and parameters that he used, that of Helmcke et.al. However, assessment of regurgitation by color echocardiography is in principle the same for experienced and inexperienced observer. Preload and afterload conditions should be standardized, because regurgitation volume is sensitive to these changes. The awareness of the investigator to these limitations enables him to make precise estimation of regurgitant lesion. It should be mentioned that the concept of color jet propagation is that the very small jet means slight regurgitation and no jet means no regurgitation, which is very simple and true.

Dr J. Fragata (Lisbon, Portugal): May I just ask you a very small question. We know that SAM is a functional issue and anatomical substrate, and anomalies of insertion of the papillary muscles have been referred in the subset. It makes sense to attack the mitral valve itself. But have you done anything about the positioning or resection of muscle, of the papillary muscles, nearby the apex of the heart?

Dr Delmo Walter: Luckily we did not find any abnormality of the papillary muscles. The papillary muscles are only anteriorly displaced, hence, by retaining the anterior leaflet then we streamline the entire valve as well.

Dr Fragata: On the mitral side.