Surgery for biventricular obstruction in hypertrophic cardiomyopathy in children and young adults: technique and outcomes †

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Abstract

OBJECTIVES: Biventricular obstruction in hypertrophic cardiomyopathy (HCM) is uncommon, and some clinicians believe that, when symptoms are refractory to medical treatment, this severe form of HCM is best treated by transplantation. We describe our conventional surgical approach and outcomes to treat biventricular obstruction in HCM.

METHODS: From 1993 to 2013, we treated 11 symptomatic patients with biventricular outflow obstruction. Relief of left ventricular (LV) obstruction was obtained by performing a transaortic extended septal myectomy and/or a left apical ventriculotomy. Right ventricular outflow tract (RVOT) obstruction was relieved with patch enlargement in all patients and selective resection of muscle bundles.

RESULTS: The mean age at surgery was 13 years (2 months–28 years); of the total, 7 (63%) were males. All were symptomatic with shortness of breath, reduced exercise tolerance or failure to thrive. All patients had preserved biventricular systolic function and systolic anterior motion (SAM) of the mitral valve (9 patients had ≥ moderate mitral regurgitation). Preoperative RVOT and LV outflow tract gradients were 60 ± 18 and 78 ± 24 mmHg, respectively. There were no early deaths. Mitral regurgitation secondary to SAM resolved following LV myectomy. The median follow-up time was 4.6 years (maximum 16.3 years). Eight patients (72%) were in NYHA class I. There have been no late ventricular arrhythmias, sudden deaths, reoperations or heart transplantations at follow-up.

CONCLUSIONS: Biventricular obstruction is rare in HCM. Surgical relief of left- and right-sided obstruction can be achieved with good early outcomes. Symptoms are improved at intermediate-term follow-up and sudden death is rare.

Keywords: Hypertrophic cardiomyopathy • Outflow tract obstruction • Myectomy

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is the most prevalent inherited cardiomyopathy, leading to early death and congestive heart failure [1, 2]. Morphological abnormalities of the right ventricle resulting in obstruction are rare [3]. In patients with documented right ventricular outflow tract (RVOT) obstruction and HCM, there is usually associated subaortic left ventricular outflow tract (LVOT) obstruction [4, 5]. There are limited data on the incidence and natural history of biventricular obstruction in the setting of HCM. In addition, the literature is scarce with regard to the analysis of treatment approaches for this condition [6, 7]. The role of surgical repair versus heart transplantation continues to be the subject of debate. Our objective was to review our experience with conventional surgical options for patients with biventricular obstruction in HCM and examine outcomes.

PATIENTS AND METHODS

Between January 1993 and July 2013, 2283 patients underwent surgery for obstructive HCM at our institution. We retrospectively reviewed the operative notes of this cohort and identified only 11 patients who underwent surgical relief of biventricular obstruction. Patients had significant pressure gradients in both ventricles (≥ 50 mmHg peak to peak measured invasively), either at baseline or after provocation. The Mayo Clinic Institutional Review Board approved this study, and patients or parents gave

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informed consent to participate in clinical research. Data related to this population were retrieved from the medical record, written correspondence from referring physicians or patients themselves, questionnaires sent at 1, 3, 5, 10 and 15 years postoperatively, and the Social Security Death Index.

**Demographics**

The mean age at the time of operation was 13 years (range, 2 months–28 years; Table 1). There were 7 males (63%). All patients were symptomatic at the time of intervention. There were 3 patients who had a familial history of sudden death with documentation that suggested the presence of familial HCM (Table 2). In 3 patients, the diagnosis was made by physicians at our institution after previous medical assessments elsewhere. The remaining patients were directly referred for evaluation of surgical intervention. Two patients had previous operations involving the contralateral ventricle before the last operation, both of which were performed at our institution. Overall, 5 of 11 patients had features of Noonan or Leopard syndrome with characteristic skin lesions (Fig. 1).

**Diagnosis**

The diagnosis of HCM was made when findings of LV hypertrophy without dilatation were present on two-dimensional transthoracic echocardiography and could not be attributed to any other systemic or cardiac disease [8]. Outflow gradients were measured preoperatively by Doppler echocardiography and were confirmed intraoperatively by direct measurements with needles in the left ventricle and aorta, or right ventricle and pulmonary artery [9]. In cases where invasive tests had a resting gradient <20 mmHg, provocative manoeuvres (induction of premature ventricular contraction or isoproterenol administration) were performed as previously described [10].

The diagnosis of RV obstruction was made with preoperative transthoracic or intraoperative transoesophageal echocardiography or by gradient measurements using exploratory needles at the time of surgery. Relief of obstruction was assessed after separation from cardiopulmonary bypass using simultaneous direct pressure measurements with needles in the ventricles and great vessels.

**Surgical approach**

Median sternotomy with extracorporeal circulation via aortic and right atrial or bicaval cannulation, and aortic occlusion with cold blood antegrade cardioplegia were utilized. Left-sided subaortic dynamic outflow tract obstruction was relieved with a transaortic extended septal myectomy [11]. Anomalous muscular and fibrous attachments of the papillary muscles were incised down to the base of the papillary muscle(s) in addition to the myectomy to relieve obstruction. When midventricular obstruction could not be safely reached through the aortic root due to a small aortic annular size, a limited apical left ventriculotomy was performed [11] and septal and free wall resection was undertaken from below. Visualization from the transapical approach allowed a safe and extensive myectomy of the midventricular region, with the ability to assess papillary muscle hypertrophy and the presence of contact lesions at this level.

The RV obstruction was approached via a limited subpulmonary longitudinal ventriculotomy. The anatomy was examined by retracting the edges of the right ventriculotomy. In the event that there was infundibular hypertrophy narrowing the RVOT at the subpulmonary valve level, muscle bundles were resected to provide appropriate widening. Special attention was given to the presence of the subvalvar tricuspid valve apparatus attachments to the hypertrophic septum. The significant variability and evidence of numerous tricuspid septal chordal attachments predicted predictable septal resection on the right side. The relief of right-sided obstruction was accomplished with free wall resection at the level of the ventriculotomy and placement of an RVOT patch. If there were no chordal attachments at this level, a limited right septal myectomy would be performed. The RVOT incision was closed with a bovine pericardial or Dacron patch. Figure 2A illustrates the surgical approach to the RVOT obstruction, and Fig. 2B shows the corresponding intraoperative echocardiographic images before and after the obstruction is relieved.

**Statistical analysis**

Continuous data are expressed as mean value ± standard deviation or median and range as appropriate. Categorical and nominal data are expressed as percentages.

**RESULTS**

Table 2 summarizes individual clinical data, anatomical features and surgical procedures. The mean age at symptom onset was 8

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**Table 1: Patient characteristics**

<table>
<thead>
<tr>
<th>Variable</th>
<th>n = 11</th>
</tr>
</thead>
<tbody>
<tr>
<td>Demographic and operative data</td>
<td></td>
</tr>
<tr>
<td>Age at surgery (years)</td>
<td>13.1 ± 11.2</td>
</tr>
<tr>
<td>Gender (male)</td>
<td>7 (63%)</td>
</tr>
<tr>
<td>Maximal LVOT gradient (mmHg)</td>
<td>78 ± 24</td>
</tr>
<tr>
<td>RVOT gradient (mmHg)</td>
<td>60 ± 18</td>
</tr>
<tr>
<td>Cardiopulmonary bypass (min)</td>
<td>111 ± 61</td>
</tr>
<tr>
<td>Aortic cross-clamp (min)</td>
<td>70 ± 30</td>
</tr>
<tr>
<td>Grams of tissue removed (g/m²)</td>
<td>4.5 ± 2.1</td>
</tr>
<tr>
<td>Maximal residual LVOT gradient (mmHg)</td>
<td>6 ± 8</td>
</tr>
<tr>
<td>Residual RVOT gradient (mmHg)</td>
<td>3 ± 6</td>
</tr>
<tr>
<td>Postoperative beta-blockers or calcium channel blockers</td>
<td>11 (100%)</td>
</tr>
<tr>
<td>Preoperative echocardiographic data LVEF (%)</td>
<td>68 ± 11</td>
</tr>
<tr>
<td>Maximal septal thickness (mm/m²)</td>
<td>19.1 ± 7.1</td>
</tr>
<tr>
<td>Posterior wall thickness (mm/m²)</td>
<td>10.4 ± 7.1</td>
</tr>
<tr>
<td>LVESD (mm/m²)</td>
<td>19 ± 7.1</td>
</tr>
<tr>
<td>LVEDD (mm/m²)</td>
<td>33.1 ± 13.5</td>
</tr>
<tr>
<td>RV enlargement</td>
<td>1 (9.1%)</td>
</tr>
<tr>
<td>Severe left atrial enlargement</td>
<td>11 (100%)</td>
</tr>
<tr>
<td>Severe right atrial enlargement</td>
<td>11 (100%)</td>
</tr>
<tr>
<td>LV mass index (g/m²)</td>
<td>173 ± 132</td>
</tr>
<tr>
<td>LA volume index (ml/m²)</td>
<td>44 ± 19</td>
</tr>
<tr>
<td>Preoperative diastolic dysfunction (grades 0–4)</td>
<td>1.8 ± 1</td>
</tr>
</tbody>
</table>

Values are shown as mean ± standard deviation or n (%).

LA: left atrium; LV: left ventricle; LVESD: left ventricular end-systolic dimension; LVEDD: left ventricular end-diastolic dimension; LV: left ventricular ejection fraction; LVOT: left ventricular outflow tract; RVOT: right ventricular outflow tract.
<table>
<thead>
<tr>
<th>#</th>
<th>Age</th>
<th>Gender</th>
<th>BSA (m²)</th>
<th>Age</th>
<th>Medical attention</th>
<th>Onset of symptoms</th>
<th>Family history HCM/SD</th>
<th>Syndrome</th>
<th>Redo LV pattern of obstruction</th>
<th>LV myectomy</th>
<th>RV pattern of obstruction</th>
<th>RV surgery</th>
<th>Additional procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10 years</td>
<td>Female</td>
<td>0.85</td>
<td>3 months</td>
<td>Murmur</td>
<td>8 years</td>
<td>No/no</td>
<td>No</td>
<td>Yes</td>
<td>Subaortic MidV APM</td>
<td>TAortic (8 years) TaPical APM resection</td>
<td>Septal Infundibular Free wall</td>
<td>Infundibular resection</td>
</tr>
<tr>
<td>2</td>
<td>23 years</td>
<td>Female</td>
<td>1.93</td>
<td>Birth</td>
<td>Murmur</td>
<td>5 years</td>
<td>No/no</td>
<td>No</td>
<td>Subaortic MidV</td>
<td>TAortic TaPical</td>
<td>Septal</td>
<td>RVOT patch</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>2 months</td>
<td>Male</td>
<td>0.56</td>
<td>Birth</td>
<td>Murmur</td>
<td>2 months</td>
<td>No/no</td>
<td>Noonan</td>
<td>Subaortic MidV</td>
<td>TAortic TaPical</td>
<td>Septal Infundibular</td>
<td>RVOT-transannular patch</td>
<td>PFO closure</td>
</tr>
<tr>
<td>4</td>
<td>13 years</td>
<td>Female</td>
<td>1.49</td>
<td>5 years</td>
<td>Murmur</td>
<td>Birth</td>
<td>Possible/yes</td>
<td>No</td>
<td>Subaortic MidV</td>
<td>TAortic TaPical</td>
<td>Septal Infundibular</td>
<td>RVOT patch</td>
<td>Aortic valve repair</td>
</tr>
<tr>
<td>5</td>
<td>4 years</td>
<td>Male</td>
<td>0.68</td>
<td>Birth</td>
<td>Respiratory distress</td>
<td>Birth</td>
<td>No/no</td>
<td>Noonan</td>
<td>Subaortic MidV APM</td>
<td>TAortic TaPical</td>
<td>Septal Infundibular</td>
<td>RVOT-transannular patch</td>
<td>Muscular VSD direct closure</td>
</tr>
<tr>
<td>6</td>
<td>27 years</td>
<td>Female</td>
<td>1.4</td>
<td>2 years</td>
<td>Murmur</td>
<td>2 years</td>
<td>No/no</td>
<td>Atypical Leopard</td>
<td>Yes</td>
<td>Subaortic</td>
<td>TAortic</td>
<td>Septal Infundibular (2 years) RVOT patch (2 years) Valvotomy (2 years)</td>
<td>Pulmonary valve repair</td>
</tr>
<tr>
<td>7</td>
<td>1 year</td>
<td>Male</td>
<td>0.37</td>
<td>5 months</td>
<td>Respiratory distress</td>
<td>5 months</td>
<td>No/no</td>
<td>No</td>
<td>Subaortic APM</td>
<td>TAortic APM resection</td>
<td>Septal Infundibular Free wall</td>
<td>Infundibular resection RVOT patch</td>
<td>Mitral valve replacement</td>
</tr>
<tr>
<td>8</td>
<td>6 months</td>
<td>Male</td>
<td>0.74</td>
<td>Birth</td>
<td>Murmur</td>
<td>6 months</td>
<td>Possible/yes</td>
<td>Leopard</td>
<td>Subaortic MidV</td>
<td>TAortic</td>
<td>Septal Infundibular</td>
<td>RVOT patch</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>28 years</td>
<td>Male</td>
<td>1.8</td>
<td>8 years</td>
<td>Murmur</td>
<td>26 years</td>
<td>No/no</td>
<td>No</td>
<td>Subaortic MidV APM</td>
<td>TAortic APM resection</td>
<td>Septal Free wall</td>
<td>Septal resection RVOT patch</td>
<td>Cor triatrium repair</td>
</tr>
<tr>
<td>10</td>
<td>26 years</td>
<td>Male</td>
<td>2.4</td>
<td>6 years</td>
<td>Familial screening</td>
<td>26 years</td>
<td>Yes/no</td>
<td>No</td>
<td>Subaortic MidV</td>
<td>Subaortic</td>
<td>TAortic</td>
<td>Septal</td>
<td>Septal resection RVOT patch</td>
</tr>
<tr>
<td>11</td>
<td>11 years</td>
<td>Male</td>
<td>1.1</td>
<td>4 months</td>
<td>Murmur</td>
<td>8 years</td>
<td>Possible/yes</td>
<td>Leopard</td>
<td>Subaortic MidV APM</td>
<td>TAortic APM resection</td>
<td>Septal Free wall</td>
<td>Septal resection Infundibular resection RVOT patch</td>
<td>Epicardial ICD implant</td>
</tr>
</tbody>
</table>

#: patient number; HCM: hypertrophic cardiomyopathy; LV: left ventricular; APM: abnormal papillary muscle; BSA: body surface area; MidV: midventricular; PFO: patent foramen ovale; PS: pulmonary stenosis; RVOT: right ventricular outflow tract; SD: sudden death; TAortic: transaortic; TaPical: transapical; ICD: internal cardioverter-defibrillator.
years (range, birth to 26 years). One patient presented with symp-
toms at birth consisting of respiratory distress requiring admission
to a neonatal intensive care unit. In 8 patients (72%), the diagnosis
was made after identification of a systolic heart murmur led to
further testing. Another patient was diagnosed at 5 months of age
when he required hospitalization for respiratory distress. One
patient was diagnosed during familial screening for HCM.

Surgery was performed electively in all cases. All patients had
symptoms of congestive heart failure, and 8 patients (72%) were in
an advanced NYHA class (III/IV) at the time of operation. Two
(18%) had associated angina and 2 other patients had a history of
syncope. There were no patients who had preoperative sudden
death. All patients were receiving appropriate medical therapy
consisting of beta-blockers (8 patients), calcium channel blockers
(1 patient) or both (2 patients). Despite the presence of significant
RV obstruction, none of these patients had symptomatic right
heart failure.

Two patients had an internal cardioverter-defibrillator (ICD)
placed before surgery as prevention of sudden death. Each of
these patients had multiple HCM-related risk factors for sudden
death; 1 had prior syncope, maximal septal thickness of 33 mm
and blood pressure drop during exercise, while the other had
prior syncope, a septal thickness of 30 mm and delayed enhance-
ment on cardiac MR imaging. There were no episodes of sudden
death or defibrillator discharges before or after surgery. One addi-
tional patient had an ICD implanted postoperatively for primary
prevention of sudden death; this patient had a positive family
history of HCM with sudden death, delayed myocardial enhance-
ment on cardiac MR imaging and drop in blood pressure during
exercise.

Obstructive lesions

The mechanisms of obstruction were reviewed using operative
notes and echocardiographic images. The universal presence of
significant diffuse septal hypertrophy contributed to biventricular
obstruction in the entire cohort. Figure 3 illustrates biventricular
obstruction and the surgical approaches used. In this series, all

Figure 1: Typical pigmented skin lesions associated with Leopard syndrome.

Figure 2: Left panel: Surgical approach to right ventricular outflow tract (RVOT) obstruction. RVOT limited longitudinal incision below the pulmonary valve. Dotted
lines depict areas of muscle resection involving the free wall, subvalvular muscle bundles; and limited septal myectomy based on the implantation of the tricuspid ap-
paratus. The ventriculotomy is uniformly closed with a patch. Right panel: Transoesophageal echocardiography performed at the time of surgical intervention focusing
on the right ventricle. Image (A) is a short-axis still frame during early systole demonstrating the typical segmental RV hypertrophy seen in our patient population,
with hypertrophy of the free wall and septum (asterisk). RA: right atrium; RV: right ventricle; LA: left atrium; PV: pulmonary valve; LVOT: left ventricular outflow tract.
Image (B) demonstrates the dynamic narrowing that occurs just below the RVOT in late systole (black arrow). This narrowing is exemplified using colour Doppler
imaging, demonstrating the lack of RV obstruction in early systole (C) with subsequent severe dynamic obstruction (black arrow) in late systole (D). Note the prominent
colour signal in the LVOT as well (black dashed arrow), as this patient had concurrent LVOT obstruction. After myectomy and RVOT patch enlargement, the RV colour
flows are laminar without evidence of significant residual obstruction in early (E) and late (F) systole.
patients had concomitant RV and LV obstruction, leading to significant outflow tract gradients (≥50 mmHg). In all patients, there was a significant right-sided septal hypertrophy bulging into the RVOT as an isolated cause of obstruction, or in association with septal muscle bundles or free wall hypertrophy. The level of obstruction in the RVOT usually was at the subpulmonary region at a distance that would allow enlargement of the outflow tract with a patch without compromising the pulmonary annulus. Importantly, 6 of these patients had tricuspid valve chordal or muscle attachments to the hypertrophied septum that precluded septal resection at the time of myectomy. In addition, 3 syndromic patients also had lesions involving the pulmonary valve complex.

LVOT obstruction occurred at the subaortic region alone in 3 patients, and the remainder had extensive septal hypertrophy involving the subaortic region and extending to the midventricular level. The presence of abnormal papillary muscles contributing to obstruction in the left side was common (5 patients).

After surgery and at follow-up, residual gradients in both sides were negligible: RVOT gradient 3 mmHg and LV/LVOT gradient 6 mmHg.

Valve abnormalities

Ten patients had systolic anterior motion (SAM)-mediated mitral regurgitation (9 patients had moderate mitral regurgitation) and 1 had severe mitral valve dysplasia. This last patient required mitral valve replacement during infancy at the time of a prior myectomy. Resolution of SAM-mediated mitral regurgitation to mild or less occurred in all patients after septal myectomy alone; there were no interventions on the mitral valve in any of these patients.

Abnormalities of the pulmonary valve were observed in 3 patients and they were associated with different clinical syndromes. Two patients had Noonan syndrome and required transannular RVOT patch enlargement to treat associated pulmonary stenosis. One patient with atypical Leopard syndrome had required a pulmonary valvotomy at age 2 when she underwent a subpulmonary patch enlargement and muscle bundle resection for obstruction related to HCM.

Early and late outcomes

There was no early mortality. Two patients required permanent pacing for a complete heart block. One patient required aortic valve repair due to new-onset aortic regurgitation due to stretching of the aortic valve at the same time of myectomy. Three patients required inotropic support early after surgery, mild doses of epinephrine or milrinone. Three patients required mechanical ventilation over 24 h. One patient required delayed chest closure to ensure haemodynamic stability. There were no other early complications. The mean intensive care unit stay was 2.8 days (median 1 day) and the mean in-hospital length of stay 7.1 days (median 6 days).

One patient was lost at follow-up; however, there were no late deaths in the remaining cohort. All patients were maintained on beta-blocker therapy.

At the 1-year follow-up after intervention, 10 patients were in NYHA functional class I and 1 patient was in functional class II. At the last follow-up (median follow-up 4.6 years, maximum 16.3 years), 8 patients (72%) were in functional class I. The patient lost after 5 years of follow-up had class III symptoms at the last evaluation.

DISCUSSION

RV wall thickening may be more common than previously understood in the HCM population, and correlation between RV and LV thicknesses in patients with HCM has been reported [3]. The presence of combined RV and LV obstruction can lead to severe diastolic dysfunction with restrictive physiology. The response to myectomy in these patients is uncertain, and some clinicians refer highly symptomatic patients with HCM and biventricular obstruction for early transplantation. The literature clearly demonstrates that transplantation is rarely required in patients with dynamic left-sided obstruction following elimination of the gradient with myectomy [12].

The anatomical aspects of RV involvement in HCM were initially described by Maron et al. [7]. The authors reported thickening of the parietal and septal limbs of the trabecula septomarginalis in 4 patients, and all 5 patients had significant RV free wall hypertrophy. Maron et al. suggested that, in contrast to the dynamic lesions occurring in the left ventricle associated with SAM, obstruction in the RV was due to static and fixed impediment to RV outflow. In addition, other authors [13–15] have described the projection of the hypertrophied RV septum into the RV cavity as a contributing factor to outflow obstruction. The presence of SAM of the tricuspid valve leading to dynamic subpulmonic stenosis in HCM has also been described [16].

Patients from this cohort may differ from other hypertrophic obstructive cardiomyopathy patients with only LV/LVOT obstruction.
obstruction in the sense that they may have presented with refractory symptoms earlier in life requiring surgical intervention at younger ages. In our series, the prevalence of biventricular obstruction accounts for less than 0.5% of our surgical experience with HCM, suggesting that the prevalence of this condition is low. However, it is not uncommon for mild-to-moderate right-sided obstruction to be overlooked and the prevalence of right-sided pathology to be underestimated by the dominant dynamic outflow tract obstruction present on the left side. Diagnosis of associated RV obstruction in patients with HCM and LVOT obstruction may be difficult because the relatively limited spatial resolution of echocardiography (two-dimensional) restricts the ability to assess RV anatomy.

Our population of patients with significant biventricular obstruction is young and severely symptomatic, conferring a challenging clinical scenario. It has been reported that septal myectomy in younger children is more complex due to difficulty in exposure of the subaortic septum through a small aorta compromising apical extension of the myectomy with an increased risk of aortic or mitral valve iatrogenic injury. Our experience with left-sided lesions in HCM has led our group to pursue myectomy in almost all patients with HCM and obstruction. Indeed, late survival of HCM patients following relief of LVOT obstruction parallels that of an age-matched population.

These results and accumulating clinical experience have led us to be more aggressive with conventional surgery for biventricular obstruction, despite the concern about advanced diastolic abnormalities that may be more likely present in this group. In our series, echocardiographic LV diastolic assessment was available for 7 patients. While not statistically significant due to the small number of patients, the mean degree of diastolic dysfunction did show a trend towards improvement after myectomy. This is consistent with prior studies that demonstrated improvement in diastolic parameters after relief of LV outflow obstruction.

From a clinical standpoint, it is important to point out the relatively high incidence of syndromic features (5/11 patients) with cutaneous manifestations in the form of pigmented cutaneous lesions ( Noonan and Leopard syndromes). In isolated reports, the rate of prevalence of HCM in patients with Leopard syndrome has been up to 71% . Although not proved, this association of melanocytic lesions may suggest a particular genetic relationship between this feature in HCM patients and possible RV involvement. Although Noonan and Leopard patients have genetically and histologically different diseases, their haemodynamic and clinical behaviours have not been different from the rest of patients in this series.

Another important issue is the natural history of this group of patients regarding the incidence of sudden death. None of these patients presented with sudden death or significant arrhythmias before or after surgery. This should be considered and interpreted with caution, with this being a young population with significant ventricular hypertrophy and familial occurrence of sudden death. The use of protective devices in young populations is not free of difficulty. There is a trend towards improvement after myectomy. This is consistent with prior studies that demonstrated improvement in diastolic parameters after relief of LV outflow obstruction.

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In our surgical experience, the presence of the hypertrophic septum bulging in the RVOT and contributing to obstruction has been thought to be a consistent pathological finding when obstruction is present at the RV level. Other factors contributing to obstruction have been hypertrophy of the RV free wall and infundibular hypertrophy. We have not appreciated dynamic RVOT obstruction with abnormal motion of the tricuspid valve, although whenever provocative manoeuvres to elucidate gradients were done, the study focused on the LVOT. We noted that, in 6 patients, there were variable tricuspid valve attachments along the hypertrophied septum that precluded septal resection without disrupting chordal support to the septal leaflet.

Limited data are available on the surgical management of this condition and there is no standardized technique. Borisov recently presented his experience, addressing the same topic in a cohort of 7 patients using a single limited RV longitudinal incision. Our surgical strategy differs significantly from his method—we advocate a biventricular cavity approach to relieve obstruction from both the LVOT and the RVOT. As mentioned previously, a significant proportion of these patients had attachments of the tricuspid valve chordae to the bulging septum precluding myectomy at this level without compromising valve function. We have found it necessary to use a subpulmonic patch to enlarge the RVOT to prevent residual and recurrent obstruction. We believe that left-sided lesions require extended LV septal myectomy to eliminate the gradient on the left side. We do not believe that right-sided relief of obstruction addresses or treats subaortic or midventricular obstruction on the left side.

Our study has focused on the complete treatment and marked reduction or elimination of biventricular obstruction in HCM. Dramatic reduction in postoperative gradients and improvement in symptoms were demonstrated despite some degree of diastolic dysfunction. Direct relief of outflow tract obstruction can be achieved with low morbidity and good intermediate- to long-term results. Conventional surgery should be offered in the setting of biventricular obstruction in HCM as it may provide significant symptomatic improvement. Despite achieving such results, we consider this a higher-risk population undergoing myectomy and thus intervention should only be offered by surgeons with expertise in HCM.

**Limitations of the study**

The patient numbers in this study are small and, while the follow-up is complete, it is short for this diagnosis since progression of diastolic dysfunction frequently occurs over decades. We recognize that identification of these 11 patients retrospectively by means of reviewing operative notes is subject to selection bias, although to our knowledge this is the longest series reported undergoing surgical correction of this rare condition. While transplantation and sudden death have been avoided in the intermediate term, longer follow-up is required. As, in our institution, patients with significant outflow tract obstruction are referred for surgery, we are unable to compare these patients with a controlled group with biventricular obstruction managed medically. Thus, conclusions regarding usefulness of this approach to delay or avoid cardiac transplantation cannot be drawn from this small population. This series reviews a single-centre experience in HCM surgery encompassing 20 years. The detection of this condition and surgical management has increased during the last decade, possibly due to a more extensive assessment of right-sided lesions. Thus, the true incidence of this disease remains unknown as our practice may be biased large number of complex referrals.
CONCLUSIONS

Concomitant biventricular obstruction is a rare condition in an HCM referral centre. Surgery addressing both right- and left-sided obstructive lesions offers symptomatic relief with low perioperative morbidity. Detailed analysis of the RVOT at the time of HCM evaluation should be routinely performed.

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