



## Original Article

# Extended Myectomy for Hypertrophic Cardiomyopathy: Early Outcomes From a Nascent Centre of Excellence in Canada

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### ABSTRACT

Hypertrophic cardiomyopathy (HCM) is one of the commonest inherited cardiac abnormalities. The disorder is clinically and genetically heterogeneous and is characterized by left ventricular wall thickening that is not explained by abnormal loading conditions. HCM is estimated to affect between 1 in 200 and 1 in 500 people in the general population. In the majority of cases, HCM patients have a relatively benign course; however, if left untreated, this abnormality can lead to sudden cardiac death, especially in young adults and athletes. Therefore, early diagnosis is crucial to help implement the proper management for

### RÉSUMÉ

La cardiomyopathie hypertrophique (CMH) est l'une des plus fréquentes anomalies cardiaques héréditaires. L'anomalie qui est cliniquement et génétiquement hétérogène est caractérisée par l'épaississement de la paroi du ventricule gauche qui n'est pas expliqué par des conditions de charge anormales. On estime que la CMH touche entre une personne sur 200 et une personne sur 500 dans la population générale. Dans la plupart des cas de CMH, l'évolution de la maladie est relativement bénigne. Toutefois, si cette anomalie n'est pas traitée, elle peut mener à la mort subite d'origine

Hypertrophic cardiomyopathy (HCM) is one of the commonest inherited cardiac conditions, with an estimated gene prevalence of 1 in 200 individuals. Despite this prevalence, Canada has relatively few centres of excellence, which limits the availability of surgery for those with left ventricular outflow tract (LVOT) obstruction who might benefit from myectomy surgery. Ten years ago, for example, patients were still being referred from the west coast of Canada to the large surgical centre in Toronto, from which a single surgeon provided myectomy services for most of the Canadian population. However, this model is neither desirable nor sustainable long-term. Development of surgical HCM expertise in a more geographically distributed fashion is in the public-patient interest. However, patients may be rightly concerned about undergoing treatment in a centre that is in its nascent

phase. New HCM programs therefore have a duty to publicly report their outcomes *relatively soon after service initiation* so that their colleagues in the wider geographic area can refer their patients with confidence, and so patients can be reassured about seeking treatment at that location.

A centre of excellence for HCM was developed at the University of Ottawa Heart Institute at the start of 2018. This development occurred in response to a growing patient need and the arrival of several new staff members with significant prior experience in this field. A formal HCM clinic was established, and referrals were received from across the Champlain Local Health Integration Network (LHIN). This region covers a large area of Eastern Ontario up to the Quebec border, and the University of Ottawa Heart Institute is the quaternary provider for 14 referring hospitals. The referral catchment area also extends north into Nunavut, which is the largest and northernmost territory of Canada, and the principal home of the Inuit. The extreme tip of Nunavut contains the settlement of *Alert*, which is the northernmost permanently inhabited place in the world, at a little over 500 miles from the geographic North Pole. This paper describes our early experience with surgical myectomy performed in adult patients between January 2018 and December 2020.

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**Ethics Statement:** This study was reviewed and approved by the Ottawa Heart Institutional Review Board (20210238-01H). The requirement for individual patient consent was waived.

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See page 927 for disclosure information.

patients with HCM. In response to the growing need for more HCM centres of excellence in Canada, we developed one such centre at the University of Ottawa Heart Institute from the start of 2018. This centre will help in the early diagnosis and management of HCM patients, especially those with left ventricular outflow tract obstruction who might benefit from myectomy surgery. This paper describes our early experience with surgical myectomy in adult HCM patients between January 2018 and December 2020. We report the results of 27 patients with HCM who underwent myectomy surgery during the study period. All 27 patients survived to discharge, and all were still alive at 6 months postdischarge. Our experience highlights the crucial role that preoperative and perioperative imaging play in the management of this condition, in addition to the vital role of having a committed “heart team” of cardiologists, surgeons, and anesthesiologists.

## Material and Methods

This study was reviewed and approved by the Ottawa Heart Institutional Review Board (20210238-01H). The requirement for individual patient consent was waived.

### Data abstraction

An institutional surgical database was reviewed to identify all patients who underwent myectomy surgery within the included date range. Details of patient demographics, symptoms, investigations, surgical procedures, and 6-month outcomes were abstracted by 2 reviewers working in consensus. Patient comorbidity was recorded. Preoperative symptomatic status and 6-month outcomes were assessed by a cardiologist specializing in the care of HCM patients. This assessment included New York Heart Association (NYHA) status, and a crude quality-of-life score [“Most days how would you score the quality of your life due to symptoms out of 10, where 0 indicates symptoms so bad you would rather be dead and 10 indicates a perfect state of health?”]. EuroScore2 was calculated for each patient, using an online calculator (<http://www.euroscore.org/calc.html>).

### Echocardiography

Every patient underwent a comprehensive transthoracic echocardiogram including measurement of outflow tract velocities using continuous wave Doppler. Preoperative and postoperative septal thickness measurements were taken from perioperative echocardiographic studies. Peak echocardiogram gradients were recorded at rest, with the Valsalva maneuver, with exercise, and with intraoperative pharmacologic stimulation. Dynamic obstruction was defined as having a characteristic late-peaking velocity profile. Peak instantaneous gradient was calculated using the modified Bernoulli equation. Volumetric measurements were made according to the recommendations of the American Society of Echocardiography. Mitral regurgitation was graded visually as follows: none, trace, mild, mild-moderate, moderate, moderate-severe, severe, or torrential.

cardiaque, particulièrement chez les jeunes adultes et les athlètes. Par conséquent, le diagnostic précoce est crucial à la mise en œuvre d'une prise en charge adéquate des patients atteints de CMH. Pour répondre à la nécessité croissante d'un plus grand nombre de centres d'excellence sur la CMH au Canada, nous avons mis en place l'un de ces centres à l'Institut de cardiologie de l'Université d'Ottawa dès le début de 2018. Ce centre contribuera au diagnostic précoce et à la prise en charge des patients atteints de CMH, particulièrement des patients, dont la CMH est associée à une obstruction de la chambre de chasse du ventricule gauche, qui pourraient bénéficier d'une myectomie. Le présent article décrit nos premières expériences de myectomie chez les patients adultes atteints de CMH entre janvier 2018 et décembre 2020. Nous présentons les résultats de 27 patients atteints de CMH qui ont subi une myectomie durant la période étudiée. Les 27 patients ont survécu jusqu'à la sortie de l'hôpital, et étaient tous encore en vie six mois après. Notre expérience démontre le rôle crucial que joue l'imagerie en phase préopératoire et en phase périopératoire lors de la prise en charge de cette maladie, en plus du rôle essentiel de l'« équipe de cardiologie » dévouée qui est composée de cardiologues, de chirurgiens et d'anesthésistes.

## Other imaging

Most patients also had preoperative cardiac magnetic resonance imaging to aid with risk stratification and implantable cardioverter defibrillator consideration (standard risk factors plus percentage of myocardial scar from late gadolinium enhancement). After the first few cases, we realized that a better 3-dimensional appreciation of the leaflets and submitral apparatus was required for the surgeons to plan their operation. In many centres, this view is available only from transesophageal echocardiogram performed at the time of surgery. We felt that the surgical team should have a prior opportunity to understand the complexity of the intracardiac anatomy and the interplay of abnormal chordal structures, displaced papillary groups, and abnormally present and inserting muscle bundles—all of which may contribute to the mechanism of obstruction.

We therefore instituted a policy of preoperative contrast-enhanced cardiac computed tomography (CT) scanning. Subsequently, we used this tool earlier in the decision-making process to identify potentially suitable candidates for alcohol septal ablation. CT studies were performed with cardiac gating and acquired in fully retrospective mode. The studies were performed at 100 kV in order to reduce overall radiation dose. We did not modulate the tube current as, in our experience, doing so produces sufficient quantum mottle to make it difficult to fully appreciate delicate chordal structures across the range of cardiac motion.

Acquired data sets were post-processed using dedicated software (Syngo.via, Siemens Medical Solutions, Erlangen, Germany) to create multiplanar reformats in 2-chamber, 3-chamber, 4-chamber, and short-axis orientation. Measurements were taken of the maximal basal septal thickness and the cranio-caudal extent of resection estimated to be required to minimize the risk of residual obstruction. Anomalous chordal structures were identified wherever possible, and the potential contribution of other factors (anterior papillary muscle displacement and anomalous muscle bundles) was assessed.

## Multidisciplinary team review

The acquired imaging information (echocardiogram, cardiac magnetic resonance exam, and cardiac CT) was reviewed with the surgical and anesthetic team (when available) the night before the planned date of surgery. This process lasted 20-30 minutes for each patient. No more than 2 patients were discussed at any one time so that the risk of confusing patient details was minimized. The aim was that the surgeons should leave the meeting with a clear understanding of the anatomy, any particular challenges, and a well-thought-out surgical strategy ahead of time.

## Intraoperative pharmacologic stimulation

Isoproterenol infusion was increased at 3-minute intervals, from 1 mg/min to 4 mg/min until the patient's heart rate was greater than 150 beats per minute. Challenge was performed in the operating room prior to the commencement of myectomy and again immediately prior to closing the chest. The protocol involved a combination of inotropic stimulation with intravenous isoproterenol infusion<sup>1</sup> combined with lowering of systemic blood pressure to between 60 and 80 mm Hg using 1 of 2 methods, as follows: (i) infusion of nitroglycerin; or (ii) transient occlusion of the inferior vena cava. Measurements of intraoperative LVOT gradient were made by transesophageal echocardiogram (TEE) performed from the deep gastric view in order to maximally align the transducer parallel to the LVOT.<sup>2</sup>

## Surgery

All surgeries were performed through a transaortic approach by 2 experienced surgeons. The heart was arrested with antegrade cardioplegia, and a transverse aortotomy was used in all cases. Three retention 4-0 Prolene sutures (Ethicon, Johnson and Johnson, NJ) were used to expose the root and the LVOT. Specially designed long malleable retractors were used to aid in the exposure.

Each patient underwent a 3-stage procedure, as follows: (i) extended septal myectomy to significantly debulk the thickened septum; (ii) resection of abnormal secondary chordae inserting onto the body of the anterior leaflet of the mitral valve; (iii) mobilization and realignment of the anterolateral papillary muscle group toward the posterior medial group when indicated. This last maneuver was done using 1 or 2 pledgeted 4-0 Gore-Tex sutures placed through the head of the posterior medial and anterolateral papillary muscle groups and tied, thus realigning the papillary muscle anatomy toward the posterior aspect of the left ventricle away from the LVOT.

Copious irrigation was used to clear all the debris. Closure of the aorta in a double-layer fashion was then completed. De-airing procedures were undertaken until TEE demonstrated no intracavitary air. At this point, all patients came off cardiopulmonary bypass without complications. Next, resting and dynamic testing with isoproterenol and nitroglycerin were again repeated. Resting and provoked gradients for each patient were documented in the operative note, in addition to the presence or absence of systolic anterior motion of the mitral valve and the degree of associated mitral insufficiency. No evidence of residual systolic anterior motion of the mitral valve was noted on TEE in any of the cases; in addition, no significant resting or provoked gradients were observed.

**Table 1. Preoperative patient variables**

Total cohort size n = 27	
Age, y	56 (18–76)
Male sex	12 (44)
BMI, kg/m <sup>2</sup>	30 (19–41)
Morbidly obese	1 (3.7)
Obese	11 (40.7)
Overweight	11 (40.7)
Normal range	4 (14.8)
Underweight	0 (0.0)
Comorbidities	
Diabetes	4 (14.8)
Hypertension	15 (55.6)
Dyslipidemia	16 (59.3)
Coronary artery disease	15 (55.6)
Obesity (BMI ≥ 30)	12 (44.4)
Obstructive sleep apnea	1 (3.7)
Atrial fibrillation at baseline	4 (14.8)
Chronic kidney disease	3 (11.1)
Smoker	4 (14.8)
Former smoker	7 (26.0)
NYHA class	
1	2 (7.4)
2	2 (7.4)
3	17 (63.0)
4	6 (22.2)
Family history of HCM	6 (22.2)
History of:	
Syncope	6 (22.2)
Angina	11 (40.7)
Dyspnea	24 (88.9)
Any atrial arrhythmia	6 (22.2)
Ventricular tachycardia	5 (18.5)
Prior myectomy	1 (3.7)
Attempted prior septal ethanol ablation	2 (7.4)
Medication use	
Beta-blocker	19 (70.4)
Diltiazem	2 (7.4)
Verapamil	1 (3.7)
Disopyramide	9 (33.3)
Amiodarone	0 (0.0)
Statin	11 (40.7)

Values are number (percent) for discrete variables, and median (range) for continuous variables.

BMI, body mass index; HCM, hypertrophic cardiomyopathy; NYHA, New York Heart Association.

Closure of the patient's chest was then performed in the customary manner. Temporary epicardial pacing wires were routinely placed. Patients were then taken to the intensive care unit in stable condition.

## Postoperative care

The physician most responsible for the patient, once the patient was out of the intensive care unit, remained the operating surgeon(s); however, the referring cardiologist was kept fully informed of the postoperative course. He was involved with any peri-operative management problems, including failure to progress due to anemia, fluid retention, or post-operative heart block and/or pacing issues. Diuretic therapy was titrated according to not only chest X-ray appearances, but also N-terminal pro-brain natriuretic peptide levels, which were expected to be below preoperative levels at the time of discharge. All patients underwent a pre-discharge echocardiogram to exclude significant pericardial effusion, ventricular septal defect, and residual outflow tract obstruction.

**Table 2. Echocardiographic findings**

Basal septal thickness preoperatively, mm	17 (11–25)	
Left atrial volume index, mL/m <sup>2</sup>	41 (21.7–93.6)	
LVEF ≥ 60% at baseline	27 (100)	
Maximum LVOT gradient		
Resting	56 (2–174)	
Valsalva	79 (10–180)	
Exercise	121 (13–324)	
Maximum provoked* gradient		
Premyectomy	111 (30–300) <sup>†</sup>	
Postmyectomy	12 (1–41) <sup>†</sup>	
<b>Maximum provoked* mitral regurgitation intraoperatively</b>	<b>Premyectomy</b>	<b>Postmyectomy</b>
None	0	1
Trace	3	14
Mild	2	10
Mild-moderate	1	2
Moderate	2	0
Moderate-severe	1	0
Severe	15	0
Torrential	3	0

Values are median (range).

LVEF, left ventricular ejection fraction; LVOT, left ventricular outflow tract obstruction.

\*“Provoked” implies intraoperative challenge according to isoproterenol protocol (see text).

<sup>†</sup>Significant difference between pre/post provoked gradients,  $P < 0.001$ .

## Statistical analysis

Data were cleaned and analyzed using Stata 16.0 (Stata-Corp, College Station, TX). Descriptive statistics were used to examine the central tendency and spread of the data. Normality of data was checked using the Shapiro-Wilk test. The Wilcoxon signed-rank test was used to compare change in NYHA class and quality-of-life scores before vs after myectomy. The threshold for significance was set at  $P < 0.05$ .

## Results

A total of 27 patients, 44% male, underwent myectomy surgery during the study period (Table 1). The median age of the patients was 56 years. All patients underwent a trial of medical therapy, including beta blockade and disopyramide, for at least 3-6 months prior to surgery. A total of 23 patients were NYHA class 3 or class 4 with limiting breathlessness on exertion. Two patients had previously undergone unsuccessful alcohol septal ablation, and a third patient had had remote myectomy a decade earlier, with a more recent recurrence of symptoms. Comorbidities were at significant levels, including obesity (44%), dyslipidemia (59%), and hypertension (56%). A smaller proportion had atrial fibrillation (15%) or chronic kidney disease (11%). Just over 40% were current or ex-smokers. The mean **European System for Cardiac Operative Risk Evaluation (EuroSCORE)2** was  $3.9 \pm 2.6$ .

## Preoperative imaging

Median septal thickness preoperatively was 17 mm, with a maximum of 25 mm; however, 12 of 27 patients (44%) had a preoperative thickness of 15 mm or less. The median peak LVOT gradient at rest was 56 mm Hg; the median Valsalva gradient was 79 mm Hg; and the median exercise gradient was 120 mm Hg.

**Table 3. Summary of surgical interventions**

Surgical procedure	Number of patients
Extended myectomy	27
Detachment of anomalous cords	22
Papillary muscle realignment	17
Papillary muscle resection	8
Direct intervention on mitral valve leaflets	1
Cox-Maze surgery for atrial fibrillation	1
Aortic valve replacement	3
Mitral valve replacement	1

## Perioperative imaging

Median peak gradient under anesthesia was 111 mm Hg at the start of surgery and had decreased to 12 mm Hg by the end of surgery. Prior to resection commencing, 21 of 27 patients (78%) had moderate or worse posteriorly directed mitral regurgitation, and associated systolic anterior motion, with isoproterenol provocation. No patients (0%) were in this category when isoproterenol stimulation was repeated at the end of surgery (Table 2). The mean cardiopulmonary bypass time was 129 minutes ( $\pm 31$ ). The mean cross-clamp time was 106 minutes ( $\pm 26$ ). Procedural details are summarized in Table 3 (and at individual patient level in Supplemental Table S1).

## Outcomes at 30 days

All 27 patients survived to discharge, and all were still alive at 6 months postdischarge. The median length of stay was 7 days (range: 4-26). The median LVOT gradient at discharge was 0 mm Hg (interquartile range: 0-7.5). Perioperative complications are listed in Table 4.

Two patients (7.4%) were in complete heart block at the end of surgery and required dual-chamber pacemaker implantation. One of these patients (3.7%) was also found to have a small peri-membranous ventricular septal defect prior to discharge, which was left untreated. Three patients (11.1%) had minor superficial sternal wound infections, which responded promptly to antibiotics and appropriate wound care. One patient had an unexplained seizure in the first few postoperative days; no cause for this was determined—CT brain scan was normal, with no recurrence at either 30 days or 6 months. One patient with baseline mild chronic renal impairment suffered acute worsening, with creatinine level rising from 99 preoperatively, peaking at 258 postoperatively, and then dropping toward normal by discharge. One patient suffered thrombosis of an internal jugular vein secondary to central line placement; this responded to anticoagulant therapy. One patient had resistant heart failure post-surgery; this condition seemed to be due to significant diastolic dysfunction that had not been well appreciated prior to the operation. This condition responded incompletely to diuretic therapy. This patient's course also was complicated by atrial fibrillation requiring cardioversion, as well as a pericardial collection that necessitated return to the operating room for drainage.

NYHA class had improved significantly by 6 months, with a median improvement of 2 classes ( $P < 0.001$ ). Patient-scored quality of life also improved significantly, from a median of 4 of 10 preoperatively to 8 of 10 at

**Table 4. Surgical outcomes at 30 days and 6 months**

Patient ID No.	Length of stay, d	30-d MACE	Discharge LVOT gradient, mm Hg	NYHA preop	NYHA at 6 months postop
1	7	N	N	3	3
2	8	SWI	8	3	2
3	7	CHB	N	3	1
4	11	N	N	4	2
5	5	N	N	1	1
6	21	N	N	2	1
7	5	N	N	3	1
8	7	N	N	4	1
9	9	Seizure	N	3	1
10	7	SWI	N	3	1
11	6	N	21	3	1
12	10	AKI	N	3	1
13	6	N	17	4	1
14	9	RIJV thrombosis	N	3	1
15	9	SWI	N	1	1
16	6	N	N	4	2
17	5	N	14	3	1
18	5	N	N	2	1
19	6	N	7	3	1
20	5	N	16	4	1
21	4	N	N	3	1
22	8	N	N	3	1
23	6	N	13	3	1
24	6	N	N	3	1
25	26	AF requiring cardioversion; pericardial collection requiring open drainage	N	3	3
26	7	N	19	3	1
27	8	CHB VSD	N	4	1

AF, atrial fibrillation; AKI, acute kidney injury; CHB, complete heart block; ID, identification; LVOT, left ventricular outflow tract; MACE, major adverse cardiovascular events; N, none; NYHA, New York Heart Association; preop, preoperatively; postop, postoperatively; RIJV, right internal jugular vein; SWI, sternal wound infection; VSD, ventricular septal defect.

6-month follow-up ( $P < 0.001$ ). For 2 patients, the NYHA class had not improved at 6 months post-surgery. Both cases had NYHA class 3 symptoms prior to surgery. The first was a woman who also underwent concomitant aortic valve surgery; the operation appeared successful at the time, and subsequent extensive investigation not only excluded any significant gradient but also failed to find an explanation for her persistent symptoms of fatigue and breathlessness. The second patient who failed to improve had a prolonged inpatient stay complicated by diastolic heart failure and arrhythmia; he also had no evidence of any residual outflow tract obstruction, but over subsequent months, he developed a rise in pulmonary artery pressure, suggesting very poor diastolic ventricular compliance, and is currently being evaluated for advanced heart failure therapies. One patient underwent surgery with a NYHA class of 1; the procedure was done following an episode of resuscitated cardiac arrest on the basis that high outflow gradients are a recognized risk factor for sudden cardiac death.

### Discussion

We present initial results of the first 27 patients who underwent myectomy surgery at our centre, following the institution of a formal HCM program. We demonstrate that even nascent programs can have excellent surgical results if

close attention is given to appropriate perioperative assessment. This success was achieved even though complex cases, often with relatively thin septa, were overrepresented in our series. We experienced no deaths within the first 30 days after surgery, and at 6 months postoperatively. These results are important because surgical myectomy experience in Canada is geographically restricted, and many patients therefore have only the option of alcohol septal ablation, despite surgical septal myectomy being the primary recommended treatment strategy in HCM guidelines.<sup>3</sup>

The first myotomy was reported in Britain by Dr W.P. Cleland.<sup>4</sup> Myectomy surgery has since emerged as the reference standard for the relief of LVOT obstruction due to HCM.<sup>5</sup> In expert centres, the mortality rate is under 1%, based on data on more than 3500 cases operated on in the 7 largest centres for HCM in North America.<sup>6</sup> Data from the National Inpatient Sample 1998-2011 in the US, however, suggested a mortality rate of up to 6%, perhaps reflecting lower case volumes and lower expertise levels in many centres (Supplemental Table S2).<sup>7,8</sup> As with all surgery, optimum outcomes with HCM are unlikely to occur with infrequent exposure. Concern is focused on not only mortality but also morbidity. Non-expert centres have higher rates of both mitral valve replacement and complete heart block than do high-volume centres. In a 2017 publication calling for greater training of myectomy surgeons, the authors included a table of

“Major active myectomy surgeons” from across the globe, defined as surgeons who had performed more than 100 myectomies.<sup>9</sup> That table contained the names of only 11 men, aged mostly in their 50s or 60s. The authors’ conclusion is hard to disagree with—that the inadequate number of expert myectomy surgeons “represents a significant clinical issue for patients with HCM and a major impediment to optimal management of the disease.” Mentorship for surgeons outside the Western world is particularly important.<sup>10</sup> Even allowing for sufficient surgical skill, acquiring the expertise to decide which patients are best left to nonsurgical management can be challenging for the HCM surgeon who is at an early stage in her/his experience. Poor case selection makes for poor outcomes, and we believe our approach (described below) minimized this risk and contributed to our excellent outcomes.

Myectomy programs have been described as starting “with surgeons experienced with HCM and the myectomy operation,”<sup>9,11</sup> However, both cart and horse are necessary for a successful trip. A successful myectomy program takes root in the presence of at least one cardiologist with interest and expertise in treating the condition. A correct decision to refer a patient for a myectomy necessitates a comprehensive assessment to demonstrate not only the presence of dynamic obstruction but also its mechanism and, crucially, the level (or levels) at which it occurs. This last point is not trivial, because if the bulk of the obstruction is mid-ventricular, a myectomy procedure will not provide symptomatic relief. In our program, we comprehensively phenotype each HCM patient to understand how they can be best helped. Surgical referral is then made on the basis of the response to the following 2 principal questions: (i) do you agree that the majority of symptoms result from LVOT obstruction?; and (ii) is surgical relief possible at acceptable risk? Once accepted for surgery, every patient is formally presented at HCM rounds, and all the clinical and imaging data are reviewed. This review takes place 1-2 days prior to the operation so the details of the anatomy will be fresh in the surgeon’s mind. The meeting includes at minimum the cardiologist and at least 1 of the 2 surgeons (usually both); we also welcome the anesthetist rostered for the case to attend, as this individual also will be performing the TEE assessment of perioperative gradients.

One reason for our successful outcomes at 6 months is the routine use of intraoperative provocative pharmacologic challenge to assess first the baseline level of obstruction and then also the adequacy of surgical intervention before closing the chest, an approach described at the Mayo Clinic<sup>1</sup> and subsequently adopted by the Boston group.<sup>12</sup> Provocative testing is vital, as both the degree of myocardial contractility and left ventricular unloading may be affected by anesthesia, resulting in lower resting gradients that are unreliable as a measure of the true physiology that will be experienced when the patient is awake and active.<sup>2</sup> Utilizing isoproterenol challenge, we were able to come off bypass with a median provoked gradient of only 12 mm Hg (from a median starting provoked gradient under anesthesia of more than 100 mm Hg). However, given that the mechanism of breathlessness is not due to simply outflow tract obstruction but also dynamic mitral regurgitation, we were careful to ensure that the latter had been dealt with adequately. This approach is reflected in the frequency with which papillary

muscle relocation was performed alongside resection of abnormal secondary mitral valve chordae in our series. As a result, of the 21 patients who had moderate or severe mitral regurgitation preoperatively, no patient had this degree of regurgitation postoperatively. These interventions can be challenging and come at some risk to the stability of the mitral apparatus. However, although 18 patients in our series had severe or torrential mitral regurgitation at preoperative provocation, no such cases were seen at the end of surgery.

Two of our patients previously had undergone aborted attempts at alcohol septal ablation prior to the formal institution of our HCM service. Currently, we reserve this technique for those who are unwilling or too infirm to undergo open septal reduction therapy. The original septal myectomy technique involved resection of only the proximal bulge of the septum, but because this addresses only one part of the obstructive mechanism, relief of obstruction is only partial.<sup>13,14</sup> Extended septal myectomy, which extends to the mid-septum, is our preference, and the values we provide for the extent of the incisions made underscore the extent of resection necessary. This approach also provides opportunity to address many of the issues involving the mitral and sub-mitral apparatus. In one Chinese series, mitral subvalvular abnormalities were found much more commonly in non-responders compared to responders.<sup>15</sup> Other groups have seen similarly impressive reductions in gradient with this approach.<sup>16</sup> Note that although most of our patients underwent procedures to address chordal anomalies, direct intervention on the mitral valve leaflets themselves very often was not required. This result is consistent with the experience from the Mayo group with a very large number patients, that isolated extended myectomy alone eliminated significant mitral regurgitation in more than 90%.<sup>17</sup> Indeed, as the accompanying editorial from the Toronto group put it: “Less is more than enough.”<sup>18</sup>

Several of our patients did not appear to gain symptomatic benefit despite apparent abolition of outflow gradient. Although disappointing, this phenomenon is common to most myectomy series and may have several explanations. In some cases, persistent obstruction remains at mid-cavity level that cannot be addressed even with a deep extended myectomy. The only option for such patients is a combined approach through both aortotomy and left ventricular apex.<sup>19-22</sup> A transapical approach of this kind is a highly skilled operation offered by only a handful of centres around the world, and it is more effective at reducing gradients than alcohol septal ablation targeted at the mid-cavity level,<sup>23</sup> despite initial reports of success with the latter approach.<sup>24</sup> Persistent symptoms of breathlessness also may reflect intrinsic abnormalities of diastolic relaxation, an energy-dependent process known to be impaired in HCM.<sup>25-27</sup> Severity of diastolic dysfunction is more difficult to assess than systolic impairment. Increasingly, we have been using bicycle exercise right heart catheterization prior to surgery to identify a subgroup of patients in whom a desultory change in outflow tract gradient from a low baseline, coupled with a rapid increase in left atrial pressure, makes them unlikely to gain benefit from myectomy. Finally, the experience of most HCM practitioners has been that patients with multiple other comorbidities tend to have a less satisfactory reduction

in symptoms than patients with HCM alone.<sup>28</sup> This finding speaks to the importance of deep phenotyping of all patients, to maximize the risk/reward ratio from surgery.

## Conclusion

Surgical myectomy has been shown to be a safe and effective procedure for symptomatic obstructive outflow tract obstruction, conferring a likelihood of survival similar to that of the general population.<sup>29</sup> We have demonstrated that a possible approach is for a committed “heart team” of cardiologists, surgeons, and anesthesiologists to develop a surgical myectomy centre in a relatively short amount of time and produce outcomes that are not only acceptable but comparable to those in the largest and longest-established centres in the world. Meticulous attention to detail is required from the team at every step from preoperative assessment to discharge. Fellowship training opportunities for both cardiologists and surgeons are required to expand myectomy surgery availability to a greater proportion of HCM patients around the world.

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### Supplementary Material

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