Short and Medium Term Outcomes of Surgery for Patients With Hypertrophic Obstructive Cardiomyopathy

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Background. We report one surgeon's experience of corrective surgery for hypertrophic obstructive cardiomyopathy (HOCM) over a 10-year span and comment on factors that influence longer term outcomes. Septal myectomy (SM) and adjunctive procedures, including shortening of the aorta, a novel technique in HOCM patients, are described.

Methods. Perioperative data were obtained by retrospective review of institutional surgical databases between 2001 and 2011. Review of most recent echocardiogram and clinical status by telephone interview was performed.

Results. A total of 211 patients underwent SM for HOCM. There was a bimodal age distribution related to sex; mean age for males and females was 46 ± 13 and 54 ± 14 years, respectively ($p < 0.001$). Functional New York Heart Association (NYHA) class improved significantly after surgery; 79% were in class III-IV preoperatively and 84% were in class I-II at follow-up ($p < 0.001$). Sixty percent had angina of Canadian Cardiovascular Society (CCS) grade III-IV preoperatively and 89% were in CCS I-II at follow-up ($p < 0.001$). There were significant improvements in resting left ventricular outflow tract gradient (64 ± 36 to 5 ± 5 mm Hg, $p < 0.001$), right ventricular systolic pressure (36 ± 7.3 to 32 ± 8 mm Hg, $p < 0.001$), left atrial size (4.6 ± 0.7 to 4.3 ± 0.6 cm, $p < 0.001$), and grade of mitral regurgitation (moderate to severe mitral regurgitation 28% to 3.5%, $p < 0.001$). In-hospital mortality was 0.5%, 1 year survival 98.6%, and 5-year survival 98.1%. Predictors of worse clinical outcomes were preoperative NYHA and CCS class III-IV ($p < 0.001$, $p = 0.05$), new onset atrial fibrillation ($p < 0.001$), and female sex ($p = 0.03$).

Conclusions. Septal myectomy in patients with obstructive HOCM offers excellent symptom relief and minimal operative risk.

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Hypertrophic cardiomyopathy is the most prevalent inherited cardiac disease, and is estimated to affect 0.2% of the population [1]. It is the most common cause of sudden death in young athletes [2]. Affected individuals may develop left ventricular outflow tract (LVOT) obstruction and have symptoms of exertional dyspnea, chest pain, and syncope [3]. The main causes of morbidity and mortality are sudden cardiac death, atrial and ventricular arrhythmias, and heart failure. The phenotypic expression is variable, ranging from minor to more severe life threatening forms.

Surgical correction of LVOT obstruction in symptomatic patients with hypertrophic obstructive cardiomyopathy (HOCM) is considered the gold standard of treatment for patients who are refractory to medical therapy and deemed suitable for surgery at experienced centers [3–5]. The operative technique has evolved from when Goodwin and colleagues [6] and Morrow and Brockenbrough [7] described the first myotomies, to more extensive resections involving the base, mid, and even the apex of the left ventricle [8]. In addition, corrections to the mitral valve (MV) and subvalvar apparatus have been reported, including plication of the anterior leaflet of the MV and release of tethering papillary muscles [9] and conversely, anterior leaflet augmentation [10].

We report the short and medium term clinical and echocardiographic outcomes of patients who underwent

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surgery for symptomatic HOCM between 2001 and 2011 by a single surgeon at a quaternary referral center. Septal myectomy (SM) was performed in all patients together with a range of additional surgical procedures described below, including shortening of the aorta, a novel technique in HOCM patients. In order to exclude the confounding effects of other cardiac lesions, we excluded patients with significant coexisting cardiac pathology (eg, coronary artery disease, structural mitral valve disease unrelated to systolic anterior motion of the mitral valve [SAM]) due to their influence on outcomes.

Patients and Methods

Identification of Study Cohort

Between January 1, 2001 and December 31, 2011, 359 patients underwent SM by 1 surgeon (A.R-E.) at Toronto General Hospital, University of Toronto. Of these, 148 had planned concomitant procedure(s) and were excluded from this study to avoid conflicting clinical outcomes. Fifty-seven had aortic valve procedures, 11 MV procedures, 78 coronary artery bypass grafting, and 2 had combined procedures, leaving 211 patients whose only cardiac disease was HOCM. Two patients had undergone SM at other institutions and 2 patients had previously undergone septal ethanol ablation (SEA).

Analyses of preoperative demographic, clinical, and echocardiographic data, operative and pathology reports, immediate postoperative and latest echocardiogram with clinical follow-up by telephone consultation, and review of hospital records were carried out. The study had full approval from the University Health Network Research Ethics Board. Data collection was started June 2012 and completed July 2013.

Preoperative Clinical Assessment

All patients were evaluated by a consultant cardiologist in the Hypertrophic Cardiomyopathy Clinic at Toronto General Hospital. Patients referred for surgery had unacceptable symptoms attributable to LVOT obstruction despite a trial of medical therapy. The rationale for performing SM rather than SEA was based on the decision of the consulting cardiologist, taking into account the patient's clinical profile, comorbidities, and patient preference [4]. In addition to transthoracic echocardiography all patients had preoperative coronary angiography.

Intraoperative Assessment and Surgical Technique

Intraoperative transesophageal echocardiography was performed in all patients to measure the interventricular septum (IVS), the distance from the base of the right coronary cusp of the aortic valve to the point of contact of the anterior mitral leaflet with the septum (SAM-septal contact point), and the mechanism of mitral regurgitation. The adequacy of the resection, the resolution of SAM, and the LVOT gradient were assessed on completion.

Briefly, the surgical technique was as follows: a transverse aortotomy distal to the sinotubular junction was performed to prevent distortion of the sinuses of Valsalva and aortic valve. A Ross retractor was used to display the muscular septum. The myectomy was commenced 2 to 4 mm below the aortic annulus adjacent to the middle of the right coronary cusp to the border of the anterolateral papillary muscle. The borders were created by making judicious incisions toward the apex, the distance and depth defined by the preceding echocardiographic findings. The myectomy was completed by making a succession of 2-mm deep incisions parallel to the curve of the aortic annulus, pulling the specimen away from the septum, creating a degree of "under-cutting." In this way a specimen of suitable thickness was developed from the base of the septum and cannot be achieved with a simple, straight cut made toward the apex. The excised muscle was measured, noting the endocardial friction lesion from SAM septal contact, and ensuring the resection extends 1 cm or more apically. Palpating the septum confirmed the creation of a smooth trench extending to the mid or apical septum. Any steps or ridges were carefully excised and debris evacuated.

The following additional procedures were performed: (1) Division of left anterior descending artery muscle bridge (LADMB) in all patients with coronary angiographic evidence of dynamic vessel obstruction (99 patients); (2) 2 to 4 mm plication of the anterior leaflet of the MV when it exhibited abnormal lengthening and redundancy contributing to SAM (2 patients); (3) resection of papillary muscles crossing the LVOT when present; (4) pulmonary vein isolation using radiofrequency ablation or cryotherapy and left atrial appendage ligation was considered in all cases of chronic atrial fibrillation (AF); (5) Aortic transection and shortening was performed in patients with severe sigmoid septum that was accentuated by an elongated aorta, as evidenced by a decreased angle between the aorta and plane of the left ventricular cavity on echocardiography (approaching 90 degrees) and a characteristic steep or reversed angle take-off of the right coronary artery from the aorta seen at coronary angiography. The aorta was transected 1 cm above the sinotubular junction. A 1.5 to 2 cm circumferential segment of ascending aorta was resected. It was reapproximated with a 15 to 30 angle rotation to correct the unfolded aorta; and (6) patent foramen ovale and atrial septal defects were routinely closed.

Statistical Analysis

Descriptive statistics are provided as the proportion, mean with standard deviation, or median with range, as appropriate. We used the paired and unpaired t tests to compare continuous variables where appropriate, and the χ^2 and Fisher exact tests to compare categoric data. Paired ordinal variables were compared with the McNemar χ^2 test where applicable. Data with greater than 10% of entries missing were not considered. If the percentage of missing data was less than 10% for a given variable, missing values were estimated by informative or mean imputation as appropriate. A p value less than 0.05 was considered significant. Calculations were performed