Original Research Article

Outcomes following transaortic septal myectomy in hypertrophic cardiomyopathy

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Received: 15 February 2020
Revised: 04 March 2020
Accepted: 05 March 2020

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ABSTRACT

Background: The objective of the study was to evaluate effect of myectomy and its impact on survival for a period of one year and to identify the co-morbid conditions that would increase the risk of surgery.

Methods: The study was conducted on the patients admitted in a single unit of department of cardiothoracic surgery, NIMS, Hyderabad during the period of 2014 to 2018. The study was a retrospective observational study. 21 patients were enrolled in the study after approval from institute ethics committee. All the patients between 7 to 70 years who underwent septal myectomy were included in the study.

Results: Out of the 21 patients underwent modified Morrows myectomy 16 (76.2%) were male and 5 (23.8%) were female. The most common presenting symptom was dyspnea (81%), followed chest pain (76%), palpitations (62%) and syncope (38%). 5 (24%) patients had a family history of sudden cardiac death. Preoperative beta blockers were used by 15 (72%) patients. 11 patients had severe mitral regurgitation, out of which 8 patients underwent valve replacement and 3 underwent mitral valve repair. The mean preoperative left ventricular outflow tract obstruction gradient was 86.86 and the mean postoperative gradient was 23.47. 3 patients had implantable cardioverter defibrillator insertions. All patients had symptom relief.

Conclusions: Surgical treatment of hypertrophic cardiomyopathy through transaortic septal myectomy is safe and effective method to relieve left ventricular outflow tract obstruction. Mitral valve replacement can be done for cases with structural defect of mitral valve. Early detection and intervention in patients with family history of sudden cardiac death would reduce the risk of death and ensure long term survival.

Keywords: Hypertrophic cardiomyopathy, Morrows procedure, Sudden cardiac death

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is considered to be the most common familial heart disease.1 It is defined by increased left ventricular wall thickness (>15 mm in one or more LV myocardial segments), that cannot be explained by abnormal loading conditions.2 Although asymmetric septal hypertrophy is most common phenotype, there is significant heterogeneity in the degree and pattern of left ventricular hypertrophy.3

The clinical spectrum of HCM is complex and includes a variety of phenotypes, which leads to different types of manifestations. Although most of the patients are asymptomatic, a significant proportion of them (25%) will develop significant symptoms or HCM related death.4 The incidence of sudden cardiac death (SCD) is 1% per year in the HCM population.5 Therefore, the objectives of HCM diagnosis and management are to relieve symptoms, prevent disease progression and major cardiovascular complications and sudden cardiac death.
Although transcatheter septal ablation with ethanol relieves symptoms in most of the cases, the associated mortality and morbidity is higher than for surgery. Transaortic modified septal myectomy is considered to be a gold standard in treatment of hypertrophic cardiomyopathy.\(^6\)

Septal myectomy relieves left ventricular outflow tract obstruction and cardiac symptoms in both adults and children. In most of the cases of HCM associated with mitral regurgitation mitral valve replacement is not needed.

To evaluate effect of surgical myectomy and its impact on survival for a period of one year. And to identify the co-morbid conditions that would increase the surgical risk. Our objective was to perform an updated systematic review to compare the efficacy, mortality (short term) of surgical myectomy (SM).

**METHODS**

The subjects for the study were the cases admitted in a single unit of department of cardiothoracic surgery, NIMS, Hyderabad during the period of 2014 to 2018. The study was a retrospective observational study. The case was selected based on defined inclusion and exclusion criteria after approval from ethical committee of NIMS.

The data was collected from hospital database including admission record, operation theatre record, ICU charts, discharge records and follow-up notes. The statistical data was analyzed using SPSS for percentages, mean values, standard deviation, etc.

The inclusion criteria included all the patients between the ages of 7 to 70 years who underwent myectomy for HCM from 2014-18. The patients below 7 and above 70 years and those patients with hypertrophic cardiomyopathy who underwent valve replacement and not myectomy were excluded from the study. Further the patients whose left ventricular outflow tract (LVOT) gradients was less than 40 were also excluded from study. The indications for surgery included NYHA class III or more, severe gradients across the left ventricular outflow tract associated with severe mitral regurgitation and presence of other cardiac conditions requiring surgery.

All patients underwent modified morrows procedure through transaortic subvalvular myectomy. The adequacy of resection was evaluated by direct inspection and digital palpation and by trans-esophageal echocardiography (TEE) after coming off bypass.

**RESULTS**

Modified morrows procedure was performed in 21 patients with a mean age of 41.6 years (range 7 to 65 years). There was a male preponderance of 76.2%.
observed in 54% (11) patients. Mitral regurgitation of mild to severe grades was present in 90% (19) of patients.

![Figure 4: Incidence of family history of SCD.](image)

Concomitant surgeries such as mitral valve replacement was done in 38% (8) patients and mitral valve repair was done in 14% (3) patients. All mitral valve replacements were done using a low profile, third generation, mechanical valve and mitral valve repair was done using teflon felt. One patient underwent coronary artery bypass surgery along with myectomy. One patient underwent aortic valve replacement as there was injury of the right coronary cusp during surgery. One female patient was on automatic implantable cardioverter defibrillator (AICD) on presentation to cardiothoracic surgery department. Two patients required PPI implantation post-operatively before discharge. One patient of 10 years old had post-operative ventricular septal defect (VSD) and was reoperated to close the VSD and this was one of the patients who needed a PPI implant for complete heart block post-operatively.

<table>
<thead>
<tr>
<th>MVR</th>
<th>Frequency</th>
<th>Percent</th>
<th>Cumulative percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>No MVR</td>
<td>10</td>
<td>47.6</td>
<td>47.6</td>
</tr>
<tr>
<td>MVR</td>
<td>8</td>
<td>38.1</td>
<td>38.1</td>
</tr>
<tr>
<td>MV repair</td>
<td>3</td>
<td>14.3</td>
<td>14.3</td>
</tr>
<tr>
<td>Total</td>
<td>21</td>
<td>100</td>
<td>100</td>
</tr>
</tbody>
</table>

Table 1: Frequency of mitral valve repair in the patients.

<table>
<thead>
<tr>
<th>LVOT gradients</th>
<th>N</th>
<th>Min</th>
<th>Max</th>
<th>Mean</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-operative</td>
<td>21</td>
<td>40</td>
<td>130</td>
<td>86.86</td>
<td>20.333</td>
</tr>
<tr>
<td>Post-operative</td>
<td>19</td>
<td>10</td>
<td>90</td>
<td>23.47</td>
<td>20.492</td>
</tr>
</tbody>
</table>

Table 2: LVOT gradients.

Early mortality was seen in two patients. One patient was a 7 years old boy suffering with mid-cavitary obstructive cardiomyopathy who had refractory ventricular tachycardia in the post-operative period. Another patient a 32 years old male died of multiorgan dysfunction secondary to sepsis as he underwent reoperation for severe mitral regurgitation in the post-operative period, for which mitral valve replacement was done on 4th post-operative day.

Echocardiographic studies done at the time of discharge revealed mean peak left ventricular outflow tract gradient of 23.4 (p<0.005 vs preoperatively). Systolic anterior mitral leaflet motion had resolved in all. Nineteen survivors were in NYHA I/II.

LVOT gradients were measured by two-dimensional echocardiography (2D echo) both preoperatively and at the time of discharge. LVOT gradient is an important determinant of cardiovascular morbidity and mortality in HCM patients.

Unexplained, exertion related syncope is a predictor of risk in all age groups, which was present in 38% of patients. Similar number of patients had hypertension. Family history of premature sudden death was present in 24% of patients. 71% of patients in the study were on beta blockers therapy. 11 patients had severe mitral regurgitation, out of which 8 patients underwent mitral valve replacement and 3 patients underwent mitral valve repair.

DISCUSSION

Hypertrophic cardiomyopathy which occurs in 1:500 people in the general population, is relatively common cardiac disease. In India and china about 2 million people are affected.7

In our study the 52.4% of patients were in the age group of 40 to 59 years. Although the disease is usually seen in the third decade of life, majority of our patients presented in the fourth decade. In children peak incidence is in the second decade and symptomatic pediatric patients show a higher annual mortality than adults. This could be due to the technically difficult of operating through small aortic opening and lack technical expertise with paediatric transosophagal echocardiography for intra-operative evaluation. Three patients under the age of 14 years underwent myectomy, with preoperative mean LVOT gradients of 96.6 (range 80-110); post operatively mean LVOT gradients were 17 (range 15-38). In our study one death of a 7 years old boy occurred post-operative period due to intractable ventricular arrhythmias.

In a study conducted in 4111 subjects in general population by Maron the prevalence of HCM in men and women was found to be 0.26:0.09.8 This male preponderance was also noted in our study (M:F 76%:23.8%). This partly could be attributed to the inheritance pattern seen in men.

The family history of SCD reveals the existence of this disease in Indian subcontinent as a genetic disorder. In our study 23.8% gave a family history of SCD. In such
patients ICD implantation is recommended according to 2014 ESC guidelines.

In a study conducted by Karam and Lever in 78 patients found that hypertension can make the hypertrophy worse but is not the cause of HCM.6 Topol et al found severe concentric hypertrophy and heart failure in 21 patients >59 years age and termed it as hypertensive hypertrophic cardiomyopathy.10 The international data suggests hypertension as the risk factor of HCM, we had 38.1% of our patients with hypertension.

Although beta-adrenergic antagonists, verapamil, and disopyramide are the mainstays of medical therapy in HOCM with outflow tract obstruction.7 Beta-blockers are first-line therapy for the treatment of symptomatic patients with obstructive HCM. The bradycardic effect of beta-blockers results in an increase in LVEDV and a resultant decrease in LVOT gradient in patients with HOCM. Pre-operatively 71.4% of patients were on beta-blockers and were asymptomatic during admission or surgery.

The chest pain which occur both during rest and exercise and can also be precipitated by large meals or alcohol.11,12 The causes could be underlying coronary artery disease, tunneled left anterior descending artery, increased Left ventricular wall stress.13 In our study 76.2% of patients complained of chest pain although only one patient had coronary artery disease.

Unexplained syncope, a marker for sudden cardiac death occurs in 15-25% of patients with HOCM.14 A slightly higher proportion (38%) of patients in our study had history of syncope.

The causes of syncope in HCM include hypovolemia, complete heart block, sinus node dysfunction, sustained ventricular tachycardia, LVOT obstruction and abnormal vascular reflexes.10,15-18 There can be more than one reason why patients with HCM lose consciousness, including co-morbidities such as epilepsy and diabetes and diabetes was seen in 9.5% of patients in our study.19

Palpitations were present in 61.9% of patients in our study. A sustained episode of palpitation lasting for more than a few minutes is often caused by supraventricular arrhythmia which was present in only one of the patients preoperatively. In patients with frequent palpitations, 48 hours ambulatory electrocardiography should be performed.15 In our patient’s ambulatory ECG was not done.

Heart failure maybe due to left ventricular outflow (LVOT) obstruction, diastolic dysfunction with restrictive physiology, or both. LVOTO is defined as a peak instantaneous doppler LV outflow tract gradient of ≥30 mm Hg, but the threshold for invasive treatment is usually considered to be ≥50 mmHg. In our study the Peak preoperative mean LVOT gradient was 85.9 and Peak postoperative mean was 23.1 with a (p value of <0.005). Suggesting a good operative outcome. Pre-operatively 71.4% of patients were on beta-blockers and postoperatively these drugs were continued. In most patients, there is a life-long process of progressive and adverse cardiac remodelling, characterized by myocardial fibrosis and wall thinning.20

Transthoracic and doppler echocardiography evaluation during rest and valsalva manoeuvre are class IB indication.21,22 Evaluation of symptomatic patients with peak LVOT grad <50 mmHg to detect provokable LVOT obstruction and assessment of mitral regurgitation are also Class IB indications.23 The baseline findings of 21 patients with HOCM were mean peak LVOT gradient of 86.86 (SD-20.3), ejection fraction of 71.3 (SD-6.9), mean septal thickness of 24±3 (range 16-32). As an institute protocol - all patients underwent 2D echo at the time of discharge and the findings showed mean peak LVOT gradient of 23.47 (SD-20.4) suggestive of adequate resection of the muscle and a good overall outcome following surgery.

Perioperative transesophageal echocardiography is recommended in patients undergoing septal myectomy to guide the surgical strategy and to detect adequacy of resection and is a class IC indication.24-31 TOE has been used in most (16 adult) of our patients.

Though cardiac MRI, late gadolinium enhancement studies, nuclear imaging and cardiac CT has been recommended. Due to the lack of resources and financial non-viability these investigations were not performed.

Surgical myectomy is the standard treatment for patients of HCM who are symptomatic with an LVOT gradient >30 mmHg and patients refractory to maximal medical therapy.

Surgical exposure of the septum can be obtained through the aorta, the left ventricle, left atrium, or right ventricle.32-35 In our institute, the transaortic approach for performing myectomy is followed.

Gradient reduction results from surgical basal septal thinning with enlargement of the LVOT area (and redirection of forward flow with loss of the drag and venturi effects on the mitral valve).36,37 Thus, SAM of the mitral valve and mitral-septal contact is abolished. Mitral regurgitation decreases without the need for additional mitral valve surgery.37 The most common reason for residual gradients across LVOT is incomplete extension of septectomy towards the base of papillary muscle. We had one patient with a peak gradient of 130 preoperatively and postoperative gradient was 90. The reason for such gradient was midcavitary obstruction and lack of TEE intraoperatively. Nevertheless, patient improved symptomatically at the time of discharge and was asked to follow up for further management with ICD.

International Surgery Journal | April 2020 | Vol 7 | Issue 4 | Page 1273
Intra-operative TEE is routinely used now in our institute for all adult patients.

In a multicenter registry study of ICDs implanted between 1986 and 2003 in 506 unrelated patients HCM Maron, Spirito et al found that ICD interventions appropriately terminated ventricular tachycardia or fibrillation in 103 patients (20%), suggesting that ICD implantation would reduce the mortality rates due to HCM.69 In our study one patient was on TPI and 2 patients had PPI before they presented to us for myectomy. All 3 patients required PPI post procedure and later had ICD insertions by cardiologists in accordance with international guidelines especially for patients with family history of SCD. Till date the affordability of an ICD in India is an issue.

Wan et al conducted a study in which 32 patients underwent either mitral valve repair or mitral valve replacement.60 In HOCM, the associated mitral regurgitation is caused by systolic anterior motion (SAM) of mitral valve leaflets and regurgitation is relieved after adequate myectomy for relief of LVOT obstruction. Residual LVOT obstruction may be due to use of an annuloplasty band - predisposing to SAM and anterior displacement of hypertrophied LV with a standard left atriotomy. Therefore, some surgeons prefer to replace the valve instead of repair by annuloplasty, due to the fear of SAM. In our study 38.1% of patients underwent Mitral valve replacement and in 14.3% of patient’s Mitral repair was done along with myectomy. In our institute we use a low-profile mechanical prosthesis and excise the anterior mitral leaflet.

Our results indicate that myectomy is an effective method for the treatment of HOCM. The change in functional class from III or IV to I or II in all cases and the decrease in LOVT gradient acutely with near disappearance of symptomatology and patient satisfaction establishes myectomy as the main stay of management in HOCM.

The limitation of our study was that it was a retrospective observational study of only one centre and the patient volume was low. The genetic analyses, MRI and other advanced investigation were not done due to the financial constraints as recommended in guidelines.

CONCLUSION

Surgical treatment of HCM through transaortic septal myectomy is safe and effective method to relieve left ventricular outflow tract obstruction is a gold standard therapy with mortality of <1%. Mitral valve replacement can be done for cases with structural defect of mitral valve- myxomatous valve. Intra-operative transoesophageal echo is a mandatory tool which would ensure adequate resection and reduce the risk of reoperations. Early detection and surgery in patients with family history of sudden cardiac death would reduce the risk of death and ensure long term survival.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the NIMS Hospital Ethics Committee, Hyderabad

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