

Original Research Article

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Hypertrophic obstructive cardiomyopathy and its outcome following surgical myectomy: a retrospective study

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ABSTRACT

Background: Hypertrophic cardiomyopathy is highly heterogeneous with a diverse anatomy, pathophysiology, and clinical course. It is obstruction to left ventricular outflow that has become the major hallmark of the disease. Septal myectomy has been the gold standard treatment for the relief of left ventricular outflow tract obstruction and cardiac symptoms in both adults and children with obstructive hypertrophic cardiomyopathy. Objective of the study was to evaluate effect of Myectomy and its impact on survival for a period of one year.

Methods: The study design is a retrospective record based observational study. Data was retrieved from previous records both electronic as well as manual records of all the patients who underwent myectomy with or without concomitant procedures such as mitral valve replacement or aortic valve replacement or coronary artery bypass surgery during 2014 to 2018.

Results: Majority of the patients 11 (52.4%) in fourth decade i.e. 40-59 years age group. majority were males i.e. 16 (76.2%) and remaining 5 i.e. 23.8% were females. Male to female ratio was 3.2:1. Dyspnoea was present 81% and chest pain in 76.2%. Preoperative LVOT gradient was 86.86 ± 20.33 and post-operative gradient was 23.47 ± 20.49 .

Conclusions: Operative techniques have evolved from simple myotomy to the present method of extended septal myectomy which can be done in all adult cases of hypertrophic obstructive cardiomyopathy.

Keywords: Hypertrophic obstructive cardiomyopathy, Outcome, Surgical myectomy

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a genetic disorder of the heart muscle that is characterized by a small left ventricular cavity and marked hypertrophy of the myocardium with myocyte disarray.¹⁻⁴ HCM is caused primarily by mutations in sarcomere proteins and is inherited in an autosomal dominant manner. Thus, HCM is a disease of the myofilaments, whose alterations in structure and function underlie its pathology and pathophysiology, as described elsewhere in this Compendium.⁵ HCM is highly heterogeneous with a diverse anatomy, pathophysiology, and clinical course.⁶ It

is obstruction to left ventricular outflow that has become the major hallmark of the disease.^{3,4}

Although most of the patients are asymptomatic, a significant proportion of them (25%) will develop risk of arrhythmias and sudden cardiac death (SCD). Therefore, the objectives of HCM diagnosis and management are to relieve the patients' symptoms (chest pain, heart failure, syncope, palpitations, etc.), prevent disease progression and major cardiovascular complications and Sudden cardiac death.⁷ Septal myectomy has been the gold standard treatment for the relief of left ventricular outflow tract obstruction and cardiac symptoms in both

adults and children with obstructive hypertrophic cardiomyopathy. Abnormalities of the mitral valve and subvalvar mitral apparatus can be managed without the need for mitral valve replacement and other cardiac lesions can be repaired simultaneously.⁸

Clinical long-term observations of larger patient series and a comparison with conventional forms of therapy are necessary to determine the conclusive therapeutic significance. So, the study was conducted in order to perform an updated review to compare the efficacy, mortality (short term) of surgical myectomy (SM).

Objective of the study was to evaluate effect of myomectomy and its impact on survival for a period of one year.

METHODS

The study design is a retrospective observational study. Data was retrieved from previous records both electronic as well as manual records of all the patients who underwent myectomy with or without concomitant procedures such as mitral valve replacement or aortic valve replacement or coronary artery bypass surgery during 2014-2018.

The study was carried out at Sri Jayadeva Institute of Cardiovascular Sciences and Research. This database included the admission record, the patient's department record, operation theatre and perfusion record, ICU charts and discharge records and follow up notes. Inclusion criteria of patients include which contained epidemiological information (age, sex, occupation, and place), risk factor evaluation (smoking, alcohol, HTN, diabetes mellitus), information of clinical presentation (exertion dyspnoea, chest pain, syncope, mean functional NYHA class, heart failure symptoms, palpitations) and clinical signs from the institutional records. Pre-operative and post-operative ECG, 2D echo data (left ventricular outflow tract (LVOT) gradient, septal thickness, heart block, associated mitral valvular and sub valvular anomaly), arrhythmias, pacemaker requirement-temporary or permanent and medical history was considered.

Patients were followed up based on their medical records, telephonic interviews and outpatient visits.

Table 3: Distribution according to clinical presentation.

Clinical features	Present		Absent	
	Frequency	Percentage	Frequency	Percentage
Chest pain	16	76.2	5	23.8
Palpitation	13	61.9	8	38.1
Syncope	8	38.1	13	61.9
Dyspnoea	17	81.0	4	19.0
Hypertension	8	38.1	13	61.9

Inclusion criteria

All the patients who underwent Myectomy for HOCM from 2014-2018, age between 7 to 70 yr and both males and females were included.

Exclusion criteria

Those patients of HOCM who underwent valve replacement and not myectomy, age below 7 yrs and above 70 yrs and those patients with LVOT gradient of less than 40 were excluded.

Statistical analysis

Authors analyzed data by using SPSS 21 version. Qualitative data was analyzed in terms of percentages.

RESULTS

Out of the 21 patients who got operated 19 were adults. Majority of the patients 11 (52.4%) in fourth decade i.e. 40-59 years age group.³ were pediatric patients (14.3%) under the age of 14 years. Least number of patients were from 60-80 years age group. i.e. 2 (9.5%) (Table 1).

In this study, majority were males i.e. 16 (76.2%) and remaining 5 i.e. 23.8% were females. Male to female ratio was 3.2:1 (Table 2).

Table 1: Distribution according to age.

Age group in years	Frequency	Percentage
0-19	3	14.3
20-39	5	23.8
40-59	11	52.4
60-80	2	9.5
Total	21	100

Table 2: Distribution according to age.

Gender	Frequency	Percentage
Male	16	76.2
Female	5	23.8
Total	21	100

Clinical presentation of the subjects revealed that in 17 patients (81%), dyspnoea was present. It is followed by chest pain in 16 i.e. 76.2%, palpitation in 13 (61.9%). (Table 3).

Pre-operative beta blocker was given in 71.4 patients. (Figure 1).

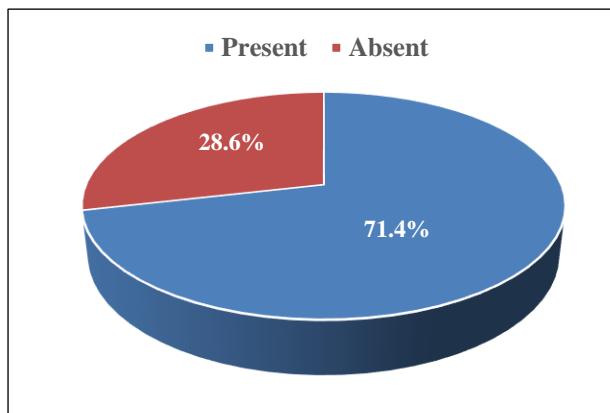


Figure 1: Distribution according to use of preoperative beta blockers.

Ten patients had severe MR and 8 patients underwent mitral valve replacement (MVR) and 3 patients underwent mitral valve repair. (Table 4).

Table 4: Distribution according to mitral valve replacement or repair.

Mitral valve treatment	Frequency	Percent
	No MVR	47.6
MVR	8	38.1
MV repair	3	14.3
Total	21	100

Table 5: Distribution according to LVOT grade.

LVOT grade	N	Mean	Std deviation
Pre-operative	21	86.86	20.33
Post-operative	19	23.47	20.49

LVOT gradients were measured by two-dimensional echocardiography (2D echo) both preoperatively and at the time of discharge. Preoperative LVOT gradient was 86.86 ± 20.33 and post-operative gradient was 23.47 ± 20.49 . LVOT gradient is an important determinant of cardiovascular morbidity and mortality in HOCM patients (Table 5).

DISCUSSION

Age

Hypertrophic cardiomyopathy can present at any age. In this study the 52.4% of patients were in the age group of 40 to 59 yrs. The age of onset of disease is in the third

decade of life, majority of patients presented to us in the fourth decade.¹⁰ In children peak incidence is in 2nd decade. Because of delay in phenotypic expression, HCM is not commonly recognized clinically in young children. Greater penetrance is seen in young males.¹⁰

Symptomatic pediatric patients show a higher annual mortality than adults. Surgery is technically difficult through the small aortic opening and lack technical expertise with pediatric TEE for intra-operative evaluation. Three patients under the age of 14 years underwent myectomy, with preoperative LVOT gradients were mean of 96.6 (range 80-110); post operatively LVOT gradients were with mean of 17 (range 15-38). As described in the literature that the younger age group is a poor prognostic factor for HCM. In this study one death of a 7-year-old boy occurred post-operative period due to intractable Ventricular arrhythmias.

Sex distribution

In the cardia study conducted in 4111 subjects in general population by Maron et al revealed the prevalence of HCM in men and women to be 0.26:0.09%.¹¹ This male preponderance was also noted in this study with 76% males and 23.8% females. This partly could be attributed to the inheritance pattern or deno-mutations seen in men. Women tend to present with more marked heart failure than men. There is no overall difference in mortality including sudden cardiac death between men and women.¹⁰

HCM with hypertension

Karam et al in a study conducted in 78 patients in Cleveland concluded that hypertension can make the hypertrophy worse but is not the cause of HCM.¹² Topolet et al severe concentric hypertrophy and heart failure in 21 patients >59 yrs age and termed it as hypertensive hypertrophic cardiomyopathy.¹³ The international data suggests hypertension as the risk factor of HCM. Authors had 38.1% of patients with hypertension.

Clinical symptoms

Clinical symptomatology of chest pain 76.2%, palpitations 61.9%, syncope 38.1%, dyspnea 81%, orthopnoea in 14.3% and fatigue in 100% of patients was present. Patients presenting to the surgical OPD were in NYHA class III or IV and post procedure 76.2% had NYHA class I and 14.3% of patients had NYHA class II symptomatology.

Chest pain

Many patients complain of chest pain at rest or on exertion. Pain may also be precipitated by large meals or alcohol.^{14,15} The causes of chest pain include myocardial ischemia due to microvascular dysfunction, increased LV

wall stress and LVOTO. Congenital coronary artery anomalies, including tunneled left anterior descending artery or atherosclerotic coronary artery disease, may also be responsible. Systolic compression of epicardial and intramural vessels is very common but is not usually of clinical importance. In this study 76.2% of patients complained of chest pain.

Surgery

Authors at Jayadeva Institute followed the transaortic approach for performing myectomy. The classical morrows procedure is not possible to perform in all cases and hence modified morrows procedure. The advantage of myectomy over septal ablation and medical therapy has been well established. Maron et al conducted a Multicenter registry study of ICDs implanted between 1986 and 2003 in 506 unrelated patients with HCM.¹⁶ ICD interventions appropriately terminated ventricular tachycardia or fibrillation in 103 patients (20%). Intervention rates were 10.6% per year for secondary prevention after cardiac arrest (5-year cumulative probability, 39% (SD, 5%)), and 3.6% per year for primary prevention (5-year probability, 17% (SD, 2%)).

Calvin et al conducted a study in which 32 patients underwent either MV repair or MVR.¹⁷ In HOCM, the associated MR is caused by SAM of valve leaflets and leakage is relieved after adequate myectomy for relief of LVOT obstruction. Structural abnormalities, primary leaflet prolapse contribute to MR in some patients. Residual LVOTO may be caused due to use of an annuloplasty band-predisposing to SAM and anterior displacement of hypertrophied LV with a standard left atriotomy may occur. Thus, some surgeons prefer to replace the valve. Cooley and associates were the first to perform MVR with prosthetic valve to relieve LVOTO and MR in patients with HOCM. In this study 38.1% of patients underwent mitral valve replacement and in 14.3% of patient's mitral repair was done along with myectomy. Authors at Jayadeva institute used a low-profile mechanical prosthesis and excise the anterior mitral leaflet routinely.

These results indicate that myectomy is an effective method for the treatment of HOCM.

CONCLUSION

Operative techniques have evolved from simple myotomy to the present method of extended septal myectomy which can be done in all adult cases of hypertrophic obstructive cardiomyopathy. Surgical treatment of HCM has become the gold standard of therapy with mortality <1%. Trans-aortic approach is safe and effective for the surgical treatment of HOCM. Symptom relief is best achieved by surgery. Early detection and intervention with echocardiography followed by surgery in patients with family history of sudden cardiac death would reduce the risk of death and ensure long term survival.

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Conflict of interest: None declared

Ethical approval: The study was approved by the Institutional Ethics Committee

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