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ORIGINAL ARTICLE

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What is the role of apical ventriculotomy in children and young adults with hypertrophic cardiomyopathy?

Alex J. Thompson MD[1] | Joseph A. Dearani MD[2] | Jonathan N. Johnson MD[1,3] | Hartzell V. Schaff MD[2] | Eric C. Towe MD[1] | Jared Palfreeman MD[1] | Philip L. Wackel MD[1] | Frank Cetta MD[1,3]

[1]Division of Pediatric Cardiology, Mayo Clinic, Rochester, Minnesota, USA

[2]Department of Cardiovascular Surgery, Mayo Clinic, Rochester, Minnesota, USA

[3]Department of Cardiovascular Diseases, Mayo Clinic, Rochester, Minnesota, USA

Correspondence
Jonathan Johnson, MD, Gonda 6-138-SW, Division of Pediatric Cardiology, Mayo Clinic, 200 First Street SW, Rochester, MN 55905.
Email: Johnson.Jonathan@mayo.edu

Abstract

Background: The transapical approach has been utilized in adult HCM patients with either midventricular obstruction or a small LV cavity; however, there are little data on its use in children.

Methods: We retrospectively reviewed all patients (age <21 years) with HCM who underwent a transapical myectomy from January 2002 to December 2016. Indication for surgery was midventricular obstruction in 19/23 (83%) and small LV cavity in 4 (17%). Preoperative symptoms included: dyspnea (96%), chest pain (65%), presyncope (61%), and syncope (35%). The mean age at the time of operation was 14 ± 4.0 years (range, 4–20).

Results: Overall, 23 patients (12 males) underwent transapical myectomy. A concomitant transaortic approach was performed in 16/19 (84%) with obstruction. The intraventricular gradient decreased from 71 mm Hg (IQR 44–92 mm Hg) preoperatively to 18 mm Hg (IQR 8–34 mm Hg, $P < .0001$) after myectomy. In patients with a small LV cavity, the mean left ventricular end diastolic dimension (LVEDD) increased from 40 ± 3 mm to 46 ± 3 mm ($P = .05$) after myectomy. There were no early deaths. Postoperative morbidity included complete heart block in 3 patients, 2 of which required pacemakers. Median follow up was 3.5 years (IQR 1.6–5.6). Symptoms improved in 95% of patients; the number of patients in NYHA class 3 or 4 heart failure decreased from 10/23 (43%) preoperatively to 3/23 (13%) postoperatively ($P < .0001$). Overall survival at 5 years postsurgery was 100%. Transplant-free survival was 91% and 87% at 1 and 5 years, respectively.

Conclusion: In children with HCM, transapical myectomy is an effective adjunct to a transaortic approach to abolish midventricular obstruction and it effectively increases LV stroke volume in patients with small LV cavities and nonobstructive HCM. It may be beneficial for these patients with significant symptoms and who have failed medical therapy as a treatment alternative to cardiac transplantation.

KEYWORDS
heart failure, hypertrophic cardiomyopathy, septal myectomy

1 | INTRODUCTION

Hypertrophic cardiomyopathy (HCM) occurs in 1/500 individuals in the general population and can cause significant morbidity and mortality.[1–3] Symptoms of HCM in children include dyspnea, decreased exercise tolerance, arrhythmias, sudden cardiac arrest, or death.[1,2,4,5] These symptoms often are caused by obstruction at the level of the left ventricular outflow tract (LVOT) or in the midventricular cavity. Symptoms may also arise due to insufficient left ventricle (LV) cavity size, which results in low stroke volume. Proper medical therapy is essential and when symptoms become severe or drug-refractory, surgical intervention is warranted.[6] Surgical management includes a

transaortic approach for basal LVOT obstruction, for which excellent outcomes have been reported previously in adult and pediatric patients.[7–10]

However, for a small subset of patients, the transaortic approach to myectomy is insufficient for adequate relief of midventricular obstruction because of poor exposure due to a small aortic annulus. In addition, a small number of patients will have heart failure-related symptoms due to "below normal" LV stroke volume. Apical myectomy to increase LV cavity size has been shown to be effective in adults.[11–14] However, minimal outcome data have been reported regarding the use of transapical myectomy in children. In this study, we have included the following: (1) the early and midterm results of transapical or combined transaortic and transapical myectomy for HCM in children in a subset of patients with advanced disease; (2) our operative technique for performing a transapical myectomy; (3) the need for advanced imaging studies for further quantification of important determinants of severity such as ventricular cavity size; and (4) a proposed algorithm to help guide management in this challenging patient population.

2 | METHODS

In this IRB-approved protocol, the Mayo Clinic Rochester surgical database was queried for all patients (<21 years of age) with HCM who had surgical myectomy that included a transapical approach between January 1, 2002 and December 1, 2016. There were 23 patients that met inclusion criteria and represent the patient cohort for this review. Six (26%) of these patients were evaluated for heart transplant prior to referral to our center. The electronic medical record of each patient was reviewed for demographic information, presenting symptoms, degree of hypertrophy, and surgical procedures performed. Data obtained included pre- and postoperative transthoracic echocardiogram (TTE) results, length of hospital stay, postoperative complications [arrhythmias, need for extracorporeal membrane oxygenation (ECMO) support, etc.] and the need for early reoperation (defined as a surgical procedure during initial hospital stay). A qualitative assessment was performed by our echocardiographers to assess degree of mitral regurgitation pre- and postoperatively. Follow-up data include current symptoms, New York Heart Association (NYHA) class and survival.

2.1 | Statistical analysis

Statistical analysis was performed using JMP Pro 10.0.0 (SAS, Cary, North Carolina). Data are reported as the mean ± standard deviation (*SD*). Matched pairs were used for continuous variables or chi-square likelihood ratios for nominal values were used to determine differences between subgroups. Significance was determined when the *P* value was ≤.05.

2.2 | Technique of apical myectomy

Standard aortic and right atrial cannulation is performed with aortic cross-clamping and cold blood antegrade cardioplegia. Making an incision 1.5 cm parallel and lateral to the left anterior descending coronary artery performs the apical ventriculotomy. The incision is usually approximately 1.5–2.0 cm in length and 2/3 of the incision is on the anterior wall of the left ventricle and 1/3 is on the posterior wall. Rake retractors are placed and the typical friction lesion between the point of contact between the hypertrophied papillary muscles and the hypertrophied ventricular septum is identified. A hook retractor is placed into the hypertrophic septal muscle at the level of the friction lesion and a septal resection is performed. The depth of the resection is determined by the overall thickness of the ventricular septum. After the septal resection has been accomplished, areas of the anterolateral and posterolateral free walls of the LV can also undergo muscle resection depending upon the degree of hypertrophy in these areas. Free wall resection is particularly important when the objective of myectomy is to increase LV cavity (ie, stroke volume) size). Hypertrophied papillary muscles are shaved along their sides. It is important to do very little myectomy at the edges of the ventriculotomy site so that ventriculotomy closure can be performed safely with an area of thickened muscle to avoid any potential aneurysm formation in the future. The ventriculotomy is closed with running monofilament suture in two layers over felt strips. Air is evacuated through the aortic root and the cross clamp is released in the usual manner. Post cardiopulmonary bypass transesophageal echo is performed to confirm absence of a midventricular gradient, or document increase in LV cavity size. In patients with obstruction, direct pressure measurements with exploring needles in the left ventricular cavity and ascending aorta are performed pre- and postmyectomy.

TABLE 1 Demographics and baseline data

	N = 23
Male/female	12/11
Age (years)	14 ± 4 (range, 4–20)
Weight (kg)	63.9 ± 19.6
Family history of HCM (%)	12/23 (52%)
Family history of SCD (%)	2/23 (9%)
Gene positive (%)	7/10 (70%)
Preoperative ICD (%)	11/23 (48%)
Preoperative RF ablation (%)	1/23 (4%)

Abbreviations: HCM, hypertrophic cardiomyopathy; SCD, sudden cardiac death; ICD, internal cardioverter defibrillator; RF, radiofrequency.

All 23 patients (12 male; mean age 14 ± 4 years, range 4–20 years) underwent transapical myectomy. The mean patient weight was 63.9 ± 19.6 kg. Twelve (52%) patients had a family history of HCM, 2 (8%) had a family history of sudden cardiac death (SCD), and 7 of 10 (70%) who had genetic testing performed were positive for mutations associated with HCM (Table 1).

Symptoms were present in 22/23 (96%) patients. All 22 had dyspnea, 15/22 (65%) had chest pain, 14/22 (61%) had presyncope, and 8/22 (35%) had syncope. About 9 of 23 (39%) patients had an arrhythmia prior to surgical intervention with 7 of those patients having ventricular

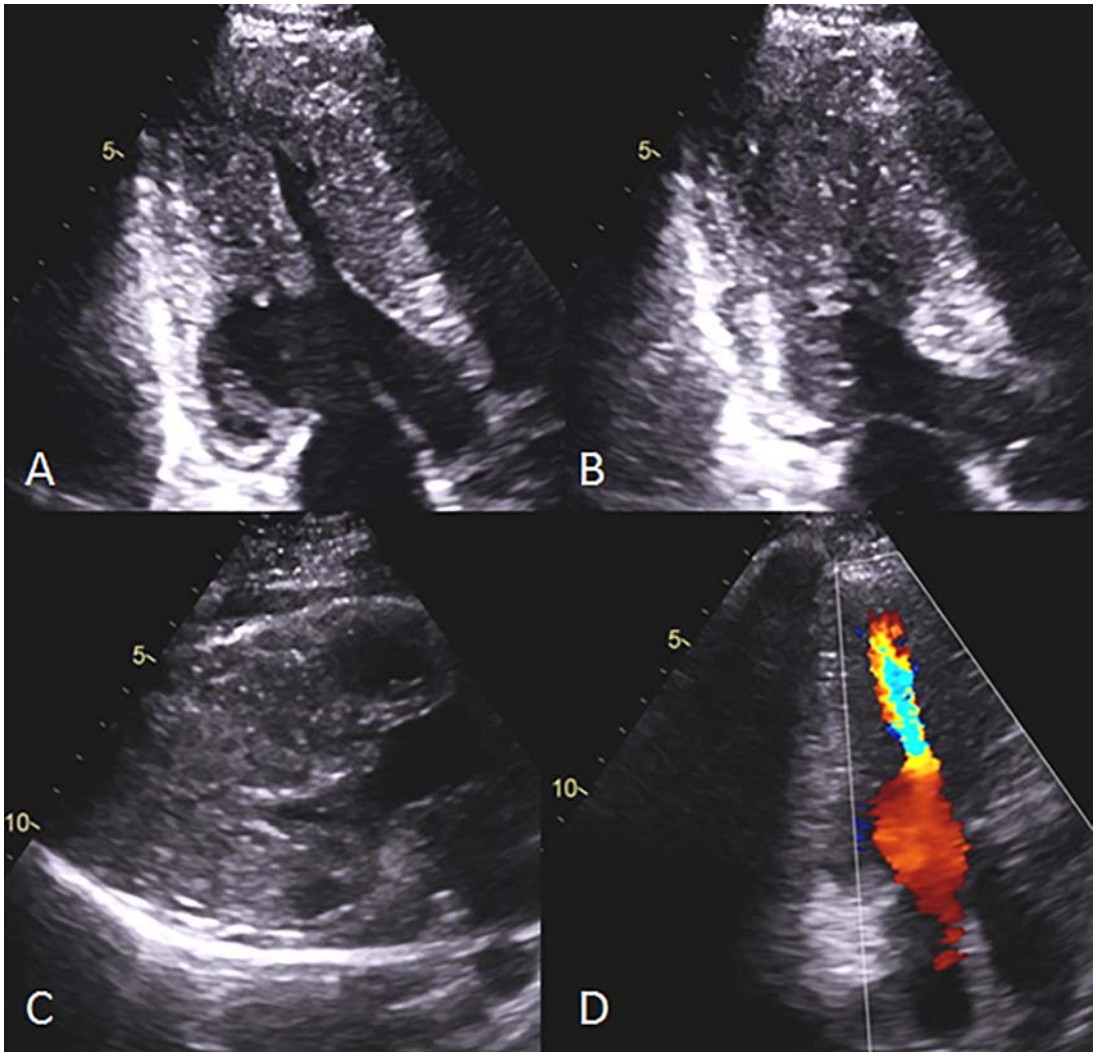

FIGURE 1 HCM with midventricular obstruction. (A) Apical long axis in diastole demonstrating severe circumferential apical hypertrophy. (B) Apical long axis in systole demonstrating absence of systolic anterior motion (SAM). (C) Parasternal long axis in systole showing cavity obliteration and absence of SAM. (D) Color flow aliasing at the midventricle consistent with midventricular obstruction without significant LVOT obstruction [Color figure can be viewed at wileyonlinelibrary.com]

tachycardia. Eleven of 23 (48%) patients had an internal cardioverter defibrillator (ICD) placed prior to myectomy. One patient (4%) had a successful preoperative radiofrequency ablation of an accessory conduction pathway for Wolff–Parkinson–White syndrome.

3 | RESULTS

3.1 | Midventricular obstruction group

Nineteen of 23 (83%) patients exhibited midventricular obstruction. An example of a patient with apical variant HCM and midventricular obstruction without LVOT obstruction is shown in Figure 1. Transapical myectomy was performed in all cases for resection of the obstruction with a concomitant transaortic approach in 16/19 (84%). The mean septal thickness measured by echocardiography was 29.6 ± 8.0 mm preoperatively, and it was reduced to 20.4 ± 6.4 mm postoperatively ($P < .01$). Reduction of the maximal intra-ventricular gradient (either midventricular or LVOT) was successful in 18/19 (95%) patients. Maximal instantaneous gradient was reduced from 71 mm Hg (median; IQR 44–92 mm Hg) to 18 mm Hg (IQR 9–34 mm Hg, $P < .01$). Ejection fraction (EF) decreased from 72% to 63% ($P < .01$, Table 2), but was still within normal limits. Preoperative cardiac magnetic resonance (CMR) was performed within three months of surgery in two patients and showed an

TABLE 2 Midventricular obstruction group: pre- and postsurgical results

N = 19	Presurgical	Postsurgical	*P*
Maximum septal thickness (mm)	30 ± 8	20 ± 6	<.0001
Intraventricular gradient (mm Hg)	68.9 ± 27.0	22 ± 21	<.0001
Abbreviation: LVEF (%)	72	63	.0004

Abbreviation: LVEF, left ventricular ejection fraction.

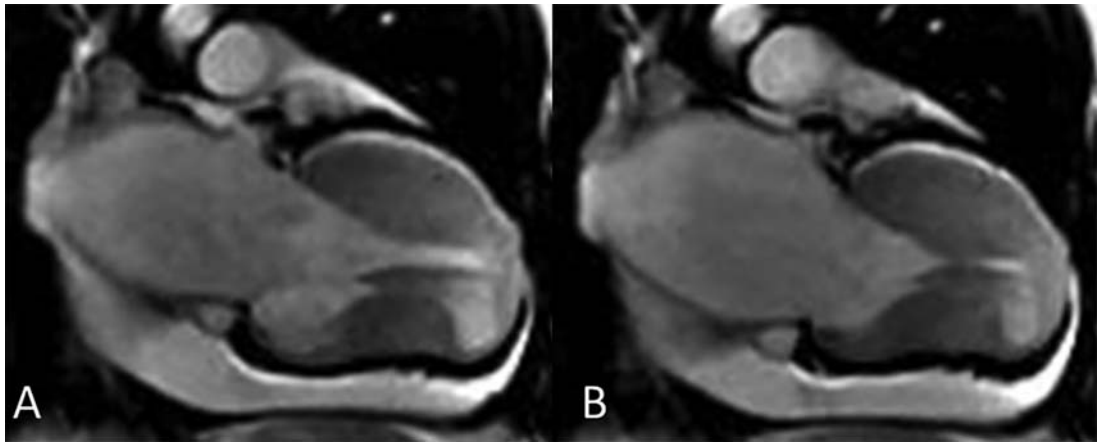

FIGURE 2 HCM with small LV cavity and systolic obliteration. Cardiac magnetic resonance imaging demonstrating: (A) small left ventricular cavity in diastole (LVEDVI 41 mL/m^2) and (B) cavity obliteration in systole (LVESVI 16 mL/m^2)

indexed LV end systolic volume (LVESVI) of 20.5 ± 6.4 mL/m^2 (Normal range: 16–40 mL[15]) and indexed LV end diastolic volume (LVEDVI) of 73.5 ± 46.0 mL/m^2 (Normal range: 56–104[15]) The indexed LV stroke volume (LVSVI) was 53.5 ± 40.3 mL/m^2 (Normal range: 36–68 mL[15]) Three other patients had CMR done 1–3 years prior to myectomy. Mean LVESVI was 16.5 ± 2.1 mL/m^2, LVEDVI was 68.1 ± 7.4 mL/m^2, and LVSVI was 50.0 ± 4.4 mL/m^2.

3.2 | Small LV cavity group

Four patients did not have midventricular obstruction but had a transapical myectomy because of a small LV cavity size; the objective was to increase LV cavity size and increase stroke volume. An example of a small LV cavity and cavitary obliteration in systole is shown in Figure 2. One of these patients had a concomitant transaortic approach. In these four patients, the preoperative mean left ventricular end systolic dimension (LVESD) and left ventricular end diastolic dimension (LVEDD) was 22 ± 5 mm and 40 ± 3 mm, respectively. Postoperatively, LVESD was increased to 32 ± 2 mm ($P < .02$), LVEDD was increased to 46 ± 3 mm ($P = .05$) and LVEF did not change significantly. Preoperative CMR was performed in two patients that showed a mean LVESVI of 19.5 ± 9.2 mL/m^2 (RR: 16–40 mL[15]) and indexed LVEDVI of 62.6 ± 6.5 mL/m^2 (RR: 56–104[15]). Mean LVSVI was 43.1 ± 2.7 mL/m^2 (RR: 36–68 mL[15]).

3.3 | Early results

Three patients had additional procedures performed at the time of myectomy that are listed in Table 3. Two patients required ECMO support for failure to separate from cardiopulmonary bypass; one had midventricular obstruction and one had a small LV cavity. The patient with midventricular obstruction had a mitral valve injury and underwent successful mitral valve repair at separation from ECMO support. The other patient with a small LV cavity had worsening heart failure secondary to diastolic dysfunction and underwent cardiac transplantation for failure to separate from ECMO support on postoperative day 17. Mean cardiopulmonary bypass time was 75.7 ± 44.8 min with a mean aortic cross clamp time of 56 ± 26 min.

There were no early deaths. Other morbidity included prolonged pleural effusions requiring thoracentesis in two patients. Three patients developed complete heart block after surgery, two of which required pacemaker implantation. In one patient who underwent pacemaker implantation, she was no longer pacemaker dependent two months after surgery. These three patients with complete heart block had additional procedures at the time of transapical myectomy; one patient had concomitant transaortic myectomy, subaortic membrane resection, and infundibular muscle resection and the other patient had a small iatrogenic VSD that required closure. Mitral valve regurgitation was significantly improved overall ($P < .01$) and is shown in Table 4.

3.4 | Late results

Late follow-up was available in 22/23 patients with a median time of 3.5 years (maximum, 8.9 years). The patient who had cardiac transplantation was alive at follow-up but was not included in this analysis. Symptom improvement following surgery occurred in 20/21 patients (95%). At latest follow up, reported symptoms included dyspnea in 9/21 (43%), chest pain in 6/21 (29%), presyncope in 6/21 (29%), and syncope in 1/21 (5%) (Figure 3). NYHA class is shown in Figure 4. Preoperative NYHA class was ≥ III in 13/23 patients (57%) and decreased to functional class I in 7/23 (30%, $P < .01$) at most recent follow-up.

TABLE 3 Concomitant procedures performed at time of myectomy

Patient	Concomitant procedures
1	MV replacement (mechanical), TV repair (annuloplasty), MV cyst excision, ASD closure
2	MV repair, transannular RVOT reconstruction, subaortic stenosis resection, anomalous MV chordae resection
3	Aortic valve repair

Abbreviations: MV, mitral valve; TV, tricuspid valve; ASD, atrial septal defect; RVOT, right ventricular outflow tract.

TABLE 4 Degree of mitral regurgitation: pre- and postmyectomy

Degree of MR	Presurgical	Postsurgical
None to trivial	10	15
Mild	5	6
Moderate	7	1
Severe	1	1

Abbreviation: MR, mitral regurgitation.

Survival at 1 and 5 years was 100%. Transplant-free survival was 91% at 1 and 87% at 5 years.

4 | DISCUSSION

This is the first report addressing the role of apical left ventriculotomy in children with HCM. Patients with HCM who have midventricular obstruction may have a worse long-term outcome than those with basilar subaortic obstruction.[16] It has also been noted that the pathogenesis of the midventricular obstruction may be different than that of basilar subaortic stenosis, with the former being caused by decreased cavity size and narrowing at the level of the papillary muscles as opposed to systolic anterior motion (SAM) of the mitral valve.[17] Additionally, there is a subset of patients with HCM and no LVOT or midventricular obstruction that exhibit similar debilitating heart failure symptoms. These patients often have severe hypertrophy in the apical portion of the ventricle with "below normal" LV cavity dimensions and symptoms that are due to the reduced LV cavity size resulting in low stroke volume. This combination of problems results in elevated left atrial pressure and inadequate diastolic filling. It is common for these patients to be evaluated for heart transplantation because of severe diastolic dysfunction.

In our practice patients undergo a thorough imaging and hemodynamic evaluation. We specifically look for obstruction in the LVOT or in the midventricle. Provocative maneuvers during TTE include Valsalva or amyl nitrite. The presence of SAM strongly suggests or confirms basal LVOT obstruction. Importantly, SAM is typically absent in the setting of midventricular obstruction because the obstruction is below the mitral leaflet and chordae and is at or below the papillary muscles. If there is a suggestion of midventricular obstruction but it is not demonstrated on TTE, then, in older children and young adults, we perform left heart hemodynamic catheterization with isoproterenol provocation. In patients with total LV cavity obliteration (particular at the apex) without obstruction, MR imaging is performed to calculate LV cavity dimensions in systole and diastole, as well as stroke volume, to determine whether they are in the normal range. It is difficult to accurately assess this using echocardiography as 2D volumetric measurements are inaccurate and often underestimate cavity size and stroke volume measurement is subject to error and depends greatly on aortic annulus diameter measurement. MR also allows for added visualization of the anatomy, function, and size of the ventricles and a quantitative measure of fibrosis of the myocardium. If MR imaging cannot be performed (eg, presence of an ICD, etc.), then left heart catheterization in older children and young adults with documentation of left-sided pressures are performed to determine whether or not the LV cavity size is below normal.

Apical myectomy is recommended when there is a maximum instantaneous gradient at rest or with provocation that exceeds 50 mm Hg; the objective of the procedure is to abolish the gradient and it is expected that heart failure symptom improve. In patients without obstruction, apical myectomy is recommended when the calculated LV dimensions are below normal, specifically; the LVEDV, LVESV, and SV are below normal. The objective of the procedure is to normalize stroke volume. In this patient group, we have seen improvement in heart failure symptoms; however, the degree of improvement may be less dramatic and less predictable that those with obstruction. Importantly, this group of patients is more likely to have progressive diastolic dysfunction and require transplantation in the future. Apical myectomy is viewed as a "bridge to transplant" or an alternative to transplant for these very high risk patients.

Patients with midventricular obstruction that is not related to systolic anterior motion (SAM) or patients with small LV cavities that are not amenable to medical therapy may be best served by surgical intervention, notably surgical myectomy.[3,6,10] However, surgical myectomy via a transaortic approach alone often is insufficient in patients with midventricular gradients or small apical cavities. These patients are much different than the "garden variety" HCM with basilar obstruction, which can be relieved by a septal myectomy with transaortic approach alone. Those patients have been shown to have excellent outcomes with significant reduction of LVOT gradient to single digits, a low complication rate, and very low mortality.[11,18]

The Mayo Clinic group has described the surgical technique for enlarging the LV cavity with a transapical myectomy for patients with "below normal" LV cavity dimensions approach (ie, low stroke volume).[12,14] Schaff et al reviewed a cohort of 44 adult patients with apical hypertrophic cardiomyopathy who had a transapical approach to enlarge the LV cavity.[14] The majority of these patients were also evaluated for transplantation since significant diastolic dysfunction was present. They showed a significant increase in LVEDVI, an increase in stroke volume measured by cardiac catheterization and a decrease in LV end diastolic pressure. Improvement of symptoms occurred in 41/42 (98%) and 23/42 (74%) of patients were NYHA class I or II at latest follow-up. Survival was 81% at 5 years. We have since learned from

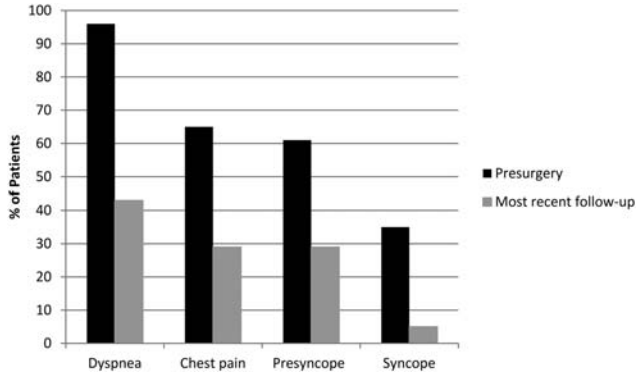

FIGURE 3 Preoperative symptoms and at most recent follow-up

the adult population with nonobstructive HCM that the benefit of LV cavity enlargement is only effective when the LV cavity dimensions and stroke volume are below the normal range. Specifically, increasing LV cavity size from normal to "above normal" is an ineffective strategy.

Our current patient cohort differs from earlier reports by Kunkala and Schaff in that it is the first to focus on children and young adult patients. Intraventricular gradients were effectively reduced in 18/19 patients who initially presented with obstruction. In addition, LVEDD was increased in those with small LV cavities. However, in our patient with nonobstructive HCM (small LV cavity) that required ECMO support postoperatively and ultimately underwent transplantation, his LV cavity dimensions were small but were in the normal range. In patients with follow-up beyond 30 days, all but one had an improvement in symptoms and NYHA class was improved. Our algorithm for the surgical management of obstructive and nonobstructive HCM is shown in Figure 5.

There were no early deaths in this series, but there was some morbidity. Complete heart block occurred in three patients, two of which required pacemaker implantation. One of the patients who required a pacemaker was found to have return of AV nodal conduction without pacemaker dependence two months later. Of the two patients who had continued pacemaker dependence, both had a combined transaortic and transventricular myectomy. Two patients required ECMO following surgery; one of those patients had an iatrogenic mitral valve injury that underwent successful repair at the time of separation from ECMO support; she was subsequently discharged to home. Another child with worsening heart failure was supported on ECMO and underwent cardiac transplant on POD 17. Other potential complications of a transapical approach have been outlined in the literature and include papillary muscle or mitral valve injury, apical aneurysm and ventricular arrhythmias.[12] While one of our patients had mitral valve injury, we have not seen apical aneurysm or an increase in ventricular arrhythmias in our adult population undergoing apical myectomy or in this pediatric group during the intermediate follow-up period. Two patients with nonobstructive HCM who had initial improvement in symptoms eventually developed worsening heart failure secondary to severe diastolic dysfunction and subsequently underwent heart transplantation for restrictive physiology at 7 months and 4 years, respectively.

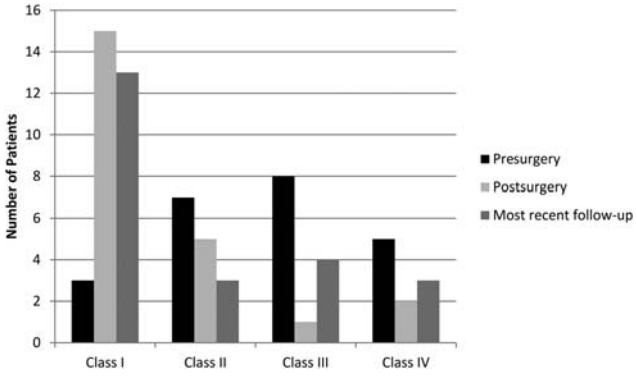

FIGURE 4 Preoperative New York Heart Association class and at most recent follow-up

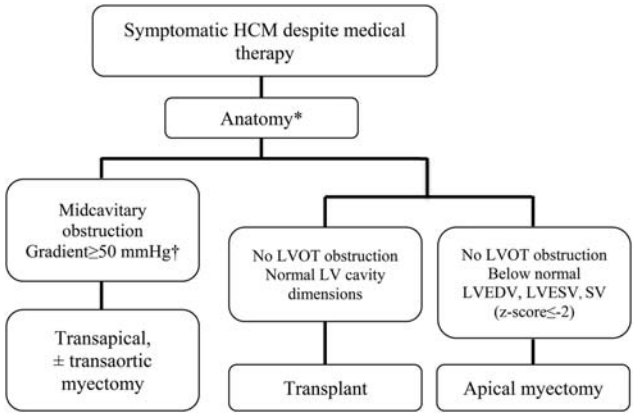

FIGURE 5 Symptomatic HCM despite medical therapy. Abbreviations: HCM, hypertrophic cardiomyopathy; LVOT, left ventricular outflow tract; LV, left ventricle; LVEDV, left ventricular end diastolic volume; LVESV, left ventricular end systolic volume; SV, stroke volume. *Subset of HCM patients with either obstruction at or below the papillary muscle level and difficult to reach by standard transaortic approach OR small LV cavity requiring enlargement. †Echo derived maximum instantaneous gradient

4.1 | Limitations

This overall patient series is small, but this is a subset of young patients with debilitating symptoms and difficult to treat HCM with a poor prognosis. There were very few patients with nonobstructive HCM undergoing LV cavity enlargement procedures, so caution should be advised when considering this procedure as an alternative to cardiac transplantation. In patients without MRI prior to surgery, echo derived end diastolic dimension, and stroke volume z-scores were used to determine if LV cavity was small. These echo parameters have inherent measurement and reproducibility pitfalls and are often different than MRI-derived values. Follow-up was relatively short (3.5 years), so the potential downside of a LV apical scar may not be long enough to reveal late ventricular arrhythmias or apical aneurysm formation.

5 | CONCLUSION

Transapical myectomy for HCM can be performed safely in children. It is an effective adjunct to a transaortic approach to abolish midventricular obstruction and it effectively increases LV stroke volume in patients with small LV cavities and nonobstructive HCM. It may be beneficial for these patients with significant symptoms and who have failed medical therapy as a treatment alternative to cardiac transplantation. Heart failure symptoms are improved in the majority of patients. Cardiac MR and catheterization are helpful in further delineating patients who may have improvement with surgical intervention, especially patients with apical hypertrophy.

AUTHOR CONTRIBUTIONS

Data analysis/interpretation, statistics, drafting article: Alex Thompson

Concept/design, drafting article, critical revision, approval: Joseph Dearani

Concept/design, critical revision, approval: Jonathan Johnson, Hartzel Schaff, Frank Cetta

Data analysis/interpretation, drafting article: Eric Towe, Jared Palfreeman

Critical revision: Phil Wackel

ORCID

Alex J. Thompson http://orcid.org/0000-0003-1553-4307

Jonathan N. Johnson http://orcid.org/0000-0001-7850-0636

Philip L. Wackel http://orcid.org/0000-0003-4560-2753

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