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# **Role of Septal Myectomy in Pediatric Hypertrophic Cardiomyopathy**

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## Abstract

Hypertrophic cardiomyopathy is a prevalent cause of sudden cardiac death among young people. This distinctive genetic condition can manifest at any age from infancy to adulthood. The primary features include unexplained left ventricular hypertrophy coupled with dynamic left ventricular outflow tract (LVOT) obstruction and a variable degree of mitral valve regurgitation. In certain cases, patients may also experience biventricular outflow tract obstruction. Surgical septal myectomy is the gold standard treatment for symptomatic children experiencing severe LVOT obstruction. This review focuses on surgical techniques for septal myectomy in pediatric patients and other adjunct procedures and summarizes the current outcomes.

**Keywords:** Hypertrophic cardiomyopathy, sudden cardiac death, left ventricular outflow tract obstruction, asymmetrical septal hypertrophy, left ventricular myectomy

## Introduction

Hypertrophic cardiomyopathy (HCM), formerly referred to as idiopathic hypertrophic subaortic stenosis, is a primary myocardial disorder with significant genetic component. This condition is marked by dynamic left ventricular outflow tract (LVOT) obstruction and asymmetrical hypertrophy of the interventricular septum (IVS). Although it is relatively rare in children compared with adults, HCM remains one of the leading causes of sudden cardiac death in younger populations. The clinical presentation varies widely from completely asymptomatic to exertional fatigue, chest pain, and/or dyspnea.

Medical management serves as the first-line approach for symptomatic individuals with LVOT obstruction<sup>(1)</sup>;



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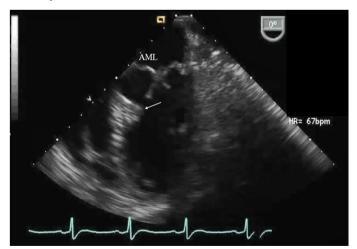
however, surgical septal myectomy should be considered for those who do not tolerate or respond adequately to medical therapies<sup>(2)</sup>.

## **Morphological Variants**

Various forms of septal hypertrophy exist within HCM, including basal (Figure 1), midventricular (Figure 2), and/ or apical variants (Figure 3). Diffuse hypertrophy may also occur<sup>(3)</sup>. Recognizing these patterns is crucial because they influence surgical decisions and approaches. For instance, the basal variant typically involves some degree of mitral valve regurgitation due to systolic anterior motion (SAM) affecting the anterior mitral valve leaflet (AML) (Figure 1). In apical cases, the left ventricular cavity tends to be considerably reduced without SAM.

## **Current Indications for Septal Myectomy**

Surgical septal reduction continues to be the gold standard in children and young adults with severe LVOT obstruction (LVOT gradient  $\geq$ 50 mmHg), intolerant to medical therapy, or who fail to respond to medications. In children, currently, there is no role for alcohol septal ablation or novel medical therapy that has been adopted recently in adults<sup>(4)</sup>.



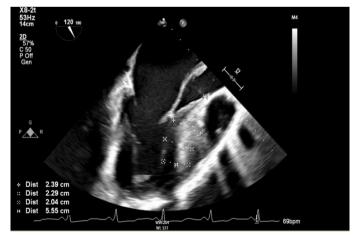
**Figure 1.** Transesophageal echocardiogram showing the characteristic features of hypertrophic cardiomyopathy with systolic anterior motion of the anterior mitral valve leaflet and basal septal hypertrophy (white arrow) *AML: Anterior mitral valve leaflet* 

Surgery should also be considered in the presence of other pathophysiological problems, such as intrinsic mitral valve (MV) disease, fixed subaortic obstruction, and midventricular and/or apical involvement.

## **Surgical Technique**

As previously noted, the specific pattern of septal hypertrophy dictates the surgical approach necessary to effectively eliminate LVOT obstruction. The procedure typically commences via standard median sternotomy with central cannulation of both the aorta and right atrium. Bicaval cannulation may be used in younger children to enhance their exposure.

Prior to initiating cardiopulmonary bypass (CPB), we routinely assess the LVOT gradient through direct needle measurements taken from both the distal ascending aorta (Figure 4A) and the left ventricle via puncturing through the right ventricular free wall into the IVS (Figure 4B). Gradient assessment occurs at rest and with provocative maneuvers- most frequently through the induction of premature ventricular contractions (PVCs) using the Brockenbrough-Braunwald-Morrow maneuver-which serves to identify dynamic LVOT obstructions.



**Figure 2.** Preoperative transesophageal echocardiogram showing the characteristic pattern of hypertrophic cardiomyopathy in a patient with both basal and midventricular hypertrophy. Notice the significant septal thickness and how far it extends into the left ventricular cavity and below the aortic valve. The patient underwent both transaortic and transapical approaches





If PVC induction fails to reveal gradients satisfactorily, medications like isoproterenol may be used. Following CPB cessation, gradient measurements are repeated to ensure effective and complete myectomy and confirm the elimination of significant LVOT gradients (Figure 5B).

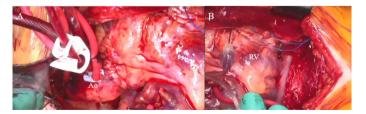
## **Surgical Approaches**

### Trans-aortic Approach

This method is particularly effective for basal variants of HCM<sup>(5,6)</sup>. After achieving cardioplegic arrest, a hockeystick incision lower than the typical aortotomy utilized for standard aortic valve replacement-extending toward the non-coronary sinus is performed (Figure 6). The first critical step is to thoroughly evaluate the LVOT and to



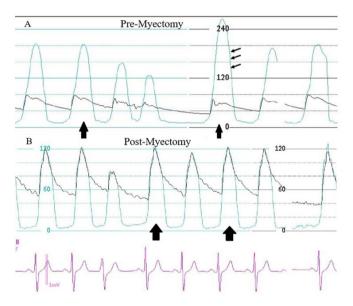
**Figure 3.** Near complete obliteration of the left ventricle apex is a feature of the apical variant of hypertrophic cardiomyopathy



**Figure 4.** Intraoperative images showing the direct assessment of the left ventricular outflow tract gradient before cardiopulmonary bypass. In image (**A**), a needle is inserted into the distal ascending aorta, while in image (**B**), a second needle is positioned within the left ventricular cavity, which is accessed indirectly through the right ventricle's free wall and the interventricular septum *Ao: Ascending aorta; RV: Right ventricle* 

identify any abnormalities within the mitral subvalvular apparatus, such as anomalous chordae or papillary muscles. This is essential because it can cause persistent or recurrent LVOT obstruction.

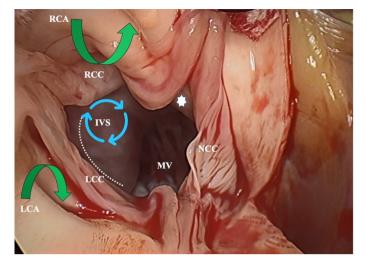
The initial incision starts just beneath the nadir of the right coronary cusp while considering some distance from its base-this precaution helps prevent future cusp prolapse with subsequent aortic regurgitation (Figure 7). The incision is then directed anticlockwise toward the commissure between the right and left coronary cusps. After initial resection widens the subaortic space, better access to the left ventricle cavity and deeper visualization of the interventricular septum are achieved. The area of resection is then widened until a satisfactory enlargement of the left ventricular cavity is achieved.



**Figure 5.** Intraoperative pressure tracing reveals: (A) before cardiopulmonary bypass initiation depicting the resting left ventricular outflow tract gradient as measured by direct needle pressures. The light blue line represents left ventricular pressure, whereas the black line indicates aortic pressure. The positive Brockenbrough-Braunwald-Morrow maneuver is indicated (via multiple thin black arrows) through induced premature ventricular pressure and a decrease in aortic pressure, thereby producing a significant left ventricular outflow tract gradient. (B) presents the tracing following complete septal myectomy, where there is no substantial gradient between the left ventricle and the aorta, indicating a negative Brockenbrough maneuver

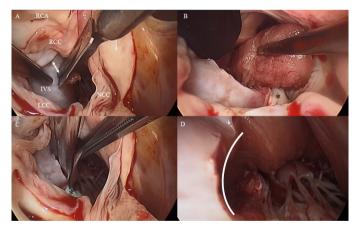






**Figure 6.** The anatomical references for the left ventricular outflow tract were identified following aortotomy and before septal myectomy. The interventricular septum can be observed along with the membranous septum (indicated by an asterisk), which should be preserved during resection. Additionally, the mitral valve is located deep within the left ventricle

RCA: Right coronary artery ostium, RCC: Right coronary cusp, LCA: Left coronary artery ostium, LCC: Left coronary cusp, NCC: Non-coronary cusp; MV: Mitral valve, IVS: Interventricular septum



**Figure 7.** The steps of the extended left ventricular septal myectomy via the trans-aortic approach are shown through these operative images. (A) the initial incision starts below the nadir of the right coronary cusp and a few millimeters away from the base of the cusp (double head arrow), then it goes toward the left/right coronary commissure, (B) the initial resected part of the septum is grasped by the forceps. Notice the very close relationship of the anterior mitral valve leaflet (asterisk), (C) The resection is completed by the scissors and (D) the area of the initial resected in the marked difference in the thickness of the septum in this area compared with the non-resected portion

RCA: Right coronary artery ostium, RCC: Right coronary cusp, LCC: Left coronary cusp, NCC: Non-coronary cusp, IVS: interventricular septum

Additional maneuvers that may enhance exposure include placing a sponge stick against the right ventricular free wall to enhance the visibility of the interventricular septum. This "extended myectomy" when compared with the initial "Morrow" technique, is characterized by its three-dimensional extension that ensures complete elimination of the gradient<sup>(7)</sup>. Once satisfactory resection is achieved, the aortotomy is closed using a two-layer running suture, followed by a standard de-airing process and removal of the aortic cross-clamp.

## Word of Caution for Children Undergoing Septal Myectomy

It can be challenging in children to perform transaortic myectomy due to the need to navigate through a small aortic root. The surgeon must exercise extreme caution when stretching the root/aortic valve during resection to avoid injuring the cusps or future aortic regurgitation.

Protecting the aortic valve during resection is paramount, and these are a few tips that can help facilitate the procedure:

Placement of the left ventricular vent through the left atrium ensures a dry field and better visualization and minimizes the need for suction through the aortic valve.

Placing a 5/0 or 6/0 polypropylene suture at the base of the cusp into the aortic sinus helps to retract the cusp away from the surgeon's view without the need to add additional instruments through the small aortic root (Figure 8A).

The use of an aortic valve leaflet retractor can protect the cusp until the initial resection is completed, and the surgeon has adequate visualization of the left ventricular cavity (Figure 8B).

After the initial resection, a small retractor can be placed through the created "trough" below the aortic valve to protect the cusp and facilitate retraction (Figure 8C).

### Trans-Apical Myectomy

The transapical approach is particularly useful for the midventricular and apical variants of HCM. In these cases, achieving complete resection through the trans-aortic





approach is challenging, if not impossible, especially in patients with small aortic annulus. The technical aspects of this approach have been described in detail elsewhere<sup>(8)</sup>.

Briefly, after cardioplegic arrest, the incision for this approach is placed 1 cm lateral to the left anterior descending coronary artery (Figure 9A) and is more

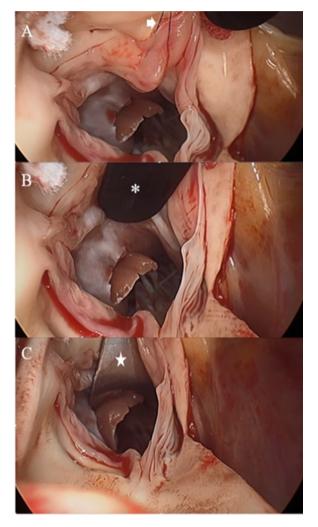


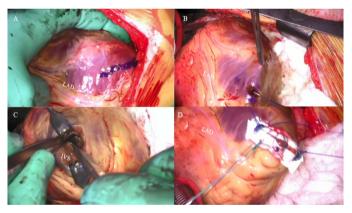
Figure 8. Intraoperative photos showing the few tips to protect the aortic valve during myectomy: (A) 5/0 or 6/0 polypropylene stitch (white arrow head) can be placed at the very base of the cusp and into the corresponding sinus, which when retracted can elevate the cusp and improve visualization, (B) a malleable retractor (asterisk) can be used to protect the right coronary cusp, but it is important to avoid over stretching and forceful retraction of the underlying cusp; and (C) once the initial trough is created, placing a low profile retractor with a small lip (white star) under the valve can help expose and protect the cusp at the same time positioned on the left ventricular free wall anteriorly rather than on the actual apex (Figure 9B).

In the true apical variant of HCM, the left ventricular apex is completely obliterated with hypertrophied muscles, and in these cases, initial resection should be limited to the area of the IVS (Figure 9C) to avoid injury to the subvalvular apparatus of the MV, which is usually displaced apically in HCM cases.

After the identification of the left ventricular cavity, additional resection is performed to further enlarge the cavity. The apex is then closed in two layers supported by two strips of Teflon felt (Figure 9D).

### Trans-Mitral Myectomy

This approach requires detachment of the anterior leaflet of the MV to expose the IVS, and once resection is completed, patch augmentation of the AML is usually required<sup>(9)</sup>. One has to be familiar with the anatomy and



**Figure 9.** Intraoperative images showing the technique of transapical myectomy: **(A)** the apical incision is performed 1 cm lateral to the left anterior descending coronary artery, **(B)** notice the thin edges of the opened apex (asterisk) in this patient reflect the presence of a small aneurysm at the apex, **(C)** the perspective through the exposed left ventricular apex reveals the resected muscle specimen and interventricular septum. It is crucial for the surgeon to remain on the interventricular septum side throughout the resection process until they can clearly identify the papillary muscles associated with the mitral valve, thereby preventing any accidental damage to the subvalvular structures of the mitral valve and **(D)** Final closure of the apical incision, which is performed using a two-layer closure supported by Teflon felt strips

LAD: Left anterior descending coronary artery, IVS: Interventricular septum





visualization of the IVS through the MV, as it differs from the commonly used trans-aortic approach. This approach may be particularly useful in children and patients with a small aortic root in which resection and exposure via the transaortic approach are limited. A third potential benefit of this approach is that it allows the treatment of MV pathology and septal myectomy via one incision.

## Adjunct Procedures for Left Ventricular Myectomy

## Resection of Anomalous Papillary Muscles/Cordae

As described above, failure to recognize and address these MV abnormalities may result in persistent or recurrent LVOT obstruction<sup>(10,11)</sup>. In addition, these abnormalities can be difficult to detect on preoperative echocardiography; thus, the initial assessment of the LVOT is key. It is important to distinguish the anomalous from the true papillary muscle of the MV, with the major difference being that the former attaches to the body of the leaflet with no chordal attachment and can therefore be safely excised. Anomalous chordae are easily distinguished from the primary chordal structure of the MV because they insert into the septum and are not attached to the free leading edge of the leaflet. These limitations limit the AML mobility, leading to SAM. It is important to remember that no chordal attachment should occur between the MV apparatus and the IVS.

## Unroofing of Associated Myocardial Bridging

Identification of myocardial bridging (MB) in patients with HCM can be challenging, and identifying those requiring unroofing can be even more challenging. There are no clear guidelines or recommendations regarding the best management approach for these patients.

Following septal myectomy, the mid-term results of different treatment modalities for MB in patients with HCM were reported by Wang et al.<sup>(12)</sup>. There were 823 patients included, with 31 events identified (mortality in 24 and non-fatal myocardial infarction in 7) by the authors. The authors concluded that surgery for MB is advantageous

when performed at the time of septal myectomy. Most patients, especially those presenting with chest pain in the setting of HCM, undergo coronary angiography. If a MB is identified, haemodynamic assessment of the bridge is done to determine if it is clinically significant, which will assist in decision-making for a concurrent unroofing procedure. Our technique of MB unroofing is performed via CPB and on the arrested heart in order not to inadvertently damage the coronary artery<sup>(13)</sup>. Using sharp and electrocautery dissection, we unroof the entire bridged segment (Figure 10). It is important to note that the bridged segment of the coronary artery is fragile, and unroofing should be meticulously performed.

## Left Ventricular Apical Aneurysm

Apical and pseudoaneurysms may occur in HCM. They are usually thin walls with varying sizes and can appear dyskinetic or akinetic. It occurs particularly in those with apical and midventricular variants (15-30% of patients)<sup>(14)</sup>. Identification of these aneurysms is not always straightforward. In an analysis of 1332 patients with apical HCM, the diagnosis of concomitant apical aneurysms on echocardiography was missed in 64.5%<sup>(15)</sup>.

Surgical resection of these aneurysms is recommended at the time of septal myectomy to minimize the risks of ventricular arrhythmias, heart failure, and/or sudden cardiac death, which have been reported in these cases<sup>(16)</sup>.



**Figure 10.** Intraoperative photograph of repeat sternotomy with unroofing of a long segment of the left anterior descending coronary artery in a patient with hypertrophic cardiomyopathy who underwent previous septal myectomy. The diagnosis of myocardial bridge in this patient was missed during his first surgery





## Placement of Epicardial Internal Cardioverter Defibrillators

In the pediatric population, and due to the limitation of transvenous implantation of internal cardioverter defibrillators (ICD), it is not unusual to place an epicardial system at the end of the septal myectomy procedure (Figure 11). The indications include those with risk factors for sudden cardiac death, history of sustained ventricular tachycardia/fibrillation, and/or septal thickness  $\geq$ 3 cm<sup>(17)</sup>. Another option in children is placement of a subcutaneous system<sup>(18)</sup>.

## Right Ventricular Myectomy

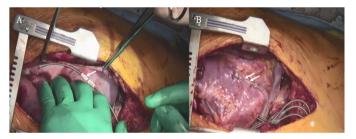
A specific subset of patients with HCM, particularly those linked to genetic conditions such as Noonan syndrome, may exhibit biventricular outflow tract obstruction. Recognizing this condition is crucial during assessment, particularly when considering left-sided surgical myectomy.

The procedure for septal myectomy on the right side differs from that on the left; notably, due to the presence of septal attachment of the tricuspid valve on the right side. Therefore, caution must be exercised when shaving the IVS on the right side to prevent damage to both the conduction tissue and the tricuspid valve apparatus. Typically, this approach is achieved via an infundibular incision, followed by patch augmentation of the right ventricle outflow (Figure 12) tract<sup>(19)</sup>.

#### **Outcomes of Septal Myectomy in A Children**

Children diagnosed with HCM often exhibit symptoms that resemble those seen in adults. These symptoms predominantly originate from a combination of diastolic dysfunction and notable mitral regurgitation. The initial signs of sudden death occur more frequently in children than in adults<sup>(20)</sup>.

The surgical procedure is technically more complex for pediatric patients because of various anatomical challenges, particularly those related to the small size of the aortic annulus and the LVOT. A study conducted at the Mayo Clinic examined 127 patients aged 2-21 years who underwent septal myectomy. The findings indicated no early mortality; the most prevalent additional procedures included resection of accessory papillary



**Figure 11.** For pediatric patients requiring an internal cardioverter defibrillator, the defibrillator implantation is typically performed postmyectomy. During this process, an epicardial system is installed, with the defibrillator coil (indicated by the white arrow in Figure A) affixed to the pericardium beneath the phrenic nerve. Concurrently, multiple sensing epicardial leads (depicted by white arrows in Figure B) are attached to the epicardial surface of the right ventricle, while the device itself is positioned in the epigastric region behind the rectus abdominis muscle (noted by asterisk)

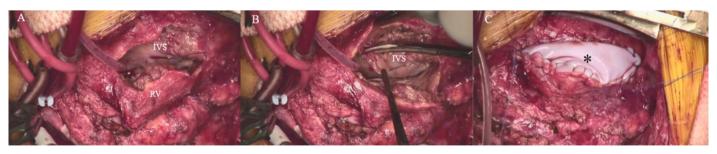


Figure 12. Intraoperative photographs showing the technique of right ventricular myectomy, which is performed through an incision in the right ventricular outflow tract (A) Limited resection is performed on the right side of the septum (B), followed by patch augmentation (asterisk) of the outflow tract (C)

IVS: interventricular septum; RV: right ventricle





muscles, repair of the MV, and closure of atrial-level shunts. Complications noted involved two cases of iatrogenic injury to the MV and seven cases involving injury to the aortic valve, all of which were successfully repaired. Additionally, they reported one case of an iatrogenic ventricular septal defect. Although there were four late fatalities, most patients experienced symptom improvement, with 96% classified within New York Heart Association class I or II. Six patients required repeat septal myectomy<sup>(21)</sup>.

### **Recurrent Obstruction Following Septal Myectomy**

In cases where complete and adequate septal myectomy is performed, recurrence rates tend to be low. As previously mentioned, it is essential to exclude anatomic factors contributing to recurrence-such as mitral subvalvular abnormalities involving anomalous papillary muscles and chordae-during the initial myectomy procedure to prevent reoperation or persistent symptoms.

Specific mechanisms leading to redo myectomy were identified in >50 patients: insufficient initial myectomy (especially critical in younger children), midventricular obstruction, and anomalous papillary muscles<sup>(22)</sup>. The option for repeat septal myectomy remains both safe and viable; and it should continue to be considered the primary treatment modality for patients experiencing recurrent or persistent LVOT gradients following initial limited resection.

## Conclusion

Extended left ventricular septal myectomy remains the gold standard for treating symptomatic children diagnosed with the obstructive variant of HCM. The trans-aortic and/or transapical approaches are most commonly used in this population, depending on the IVS morphology and the level of obstruction. Other adjunctive procedures that may be considered include concomitant unroofing of significant MB and placement of epicardial ICD in patients who meet the criteria. Concomitant right ventricular outflow tract obstruction, especially in patients with genetic syndromes. The outcomes remain excellent with low risk of recurrence if complete and satisfactory myectomy is initially performed.

## Ethics

**Financial Disclosure:** This study did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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