Hypertrophic Obstructive Cardiomyopathy
Surgical Myectomy and Septal Ablation

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Abstract: Hypertrophic cardiomyopathy is a genetic disorder characterized by marked hypertrophy of the myocardium. It is frequently accompanied by dynamic left ventricular outflow tract obstruction and symptoms of dyspnea, angina, and syncope. The initial therapy for symptomatic patients with obstruction is medical therapy with β-blockers and calcium antagonists. However, there remain a subset of patients who have continued severe symptoms, which are unresponsive to medical therapy. These patients can be treated with septal reduction therapy, either surgical septal myectomy or alcohol septal ablation. When performed by experienced operators working in high-volume centers, septal myectomy is highly effective with a >90% relief of obstruction and improvement in symptoms. The perioperative mortality rate for isolated septal myectomy in most centers is <1%. Alcohol septal ablation is a less invasive treatment. In many patients, the hemodynamic and clinical results are comparable to that of septal myectomy. However, the results of alcohol septal ablation are dependent on the septal perforator artery supplying the area of the contact between the hypertrophied septum and the anterior leaflet of the mitral valve. There are some patients, particularly younger patients with severe hypertrophy, who do not uniformly experience complete relief of obstruction and symptoms. Both techniques of septal reduction therapy are highly operator dependent. The final decision as to which approach should be selected in any given patient is dependent upon patient preference and the availability and experience of the operator and institution at which the patient is being treated. (Circ Res. 2017;121:771-783. DOI: 10.1161/CIRCRESAHA.116.309348.)

Key Words: athletes ■ blood pressure ■ cardiomyopathy, hypertrophic ■ dyspnea ■ hypertrophy

Obstruction in Hypertrophic Cardiomyopathy

Introduction
Hypertrophic cardiomyopathy (HCM) is a genetic disorder of the heart muscle, 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. HCM is highly heterogeneous with a diverse anatomy, pathophysiology, and clinical course. Some patients present with severe dyspnea, angina, and syncope, but many patients remain asymptomatic throughout life. There is an increased risk of sudden cardiac death, more commonly in younger patients; indeed, HCM is the most common cause of sudden cardiac death in trained athletes. Yet the overall survival of most patients with HCM is comparable to an age- and sex-matched population without heart disease.

It is obstruction to left ventricular outflow that has become the major hallmark of the disease. The unique pathophysiology underlying the obstruction is its functional dynamic nature, which is greatly influenced by alterations in the load imposed on the left ventricle and its contractility (Figure 1). Although the clinical significance of the outflow pressure gradient had been questioned in the past, it is now well accepted that true obstruction to outflow does exist. This obstruction causes an increase in left ventricular systolic pressure, which then leads to a complex interplay of pathophysiologic abnormalities including a high wall stress, prolongation of ventricular relaxation, impairment of left ventricular filling, elevation of left ventricular diastolic pressures, secondary mitral regurgitation, myocardial ischemia, and a reduction in cardiac output.

Among all patients presenting with HCM, resting left ventricular outflow tract obstruction (Figure 2; defined as a peak pressure gradient at rest >30 mmHg) is present in approximately one third and latent obstruction (no obstruction at rest but obstruction upon provocation) occurs in another third. The remaining third have no obstruction either at rest or on provocation during their initial evaluation, but it is unclear how many of these patients will later develop outflow tract gradients. It is the dynamic left ventricular outflow tract obstruction and its secondary pathophysiologic consequences, which play the major role in producing exertional dyspnea, angina, and near syncope. It is important to establish the presence or absence of obstruction because symptomatic relief after treatment of obstruction with septal reduction therapy is excellent. In patients with symptoms who have a latent obstruction that is evident only with provocation, symptomatic relief after septal reduction therapy may also occur.

Pathophysiology of Obstruction

Obstruction to left ventricular outflow was initially thought to be caused solely by encroachment of a hypertrophic basal septum into the left ventricular outflow tract during systole. Echocardiographic studies then described systolic anterior motion (SAM) of the mitral valve (Figure 2), which contacts the septum during mid to late systole, and this is now recognized as a primary component of the obstruction in the majority of patients. Initially, SAM was thought to be because of a Venturi effect from the septal hypertrophy sucking the mitral valve leaflets into the left ventricular outflow tract. It is now recognized that ejection occurs against an abnormally positioned and elongated mitral valve apparatus, which results in a drag force on a portion of the mitral valve leaflets and pushes the leaflets into the outflow tract, thereby causing obstruction. Distortion of the mitral valve leaflets frequently results in secondary mitral regurgitation, which may be a major cause of severe symptoms. The mitral regurgitation is a late systolic event, usually directed posterolaterally, and its severity is dependent on the degree of outflow obstruction. The overall sequence of the pathophysiologic events in patients with HCM and obstruction with secondary mitral regurgitation has been described as eject, obstruct, and leak. It is important to understand this pathophysiology because the treatment of the obstruction often also treats the mitral regurgitation (see below).

The septal hypertrophy may extend distally and obstruction can occur in the midcavitary region because of a hypertrophied papillary muscle abutting a hypertrophied ventricular septum (Figure 3). This may result in a high pressure gradient between the apex and base of the heart, with the high left ventricular apical pressures causing abnormally elevated wall tension, and myocardial ischemia. Patients with midventricular obstruction with or without basal outflow obstruction may develop apical aneurysms. These patients are frequently severely symptomatic and at risk for ventricular arrhythmias and embolic events.

Diagnosis and Initial Treatment of Obstruction

Patients with HCM should be educated on the disease, including its genetic nature and the need to screen all first-degree relatives. Genetic testing and counseling may be useful in patients with large families or in whom multiple members have been affected. Risk stratification for sudden death should be performed irrespective of symptoms or the presence or absence of obstruction. In patients with HCM presenting with exertional symptoms, it is essential to determine whether obstruction is present, either at rest or during provocation. If obstruction is present, its relief is likely to reduce or abolish these symptoms. Thus, a directed physical examination coupled with comprehensive 2-dimensional and Doppler echocardiography are needed to identify the presence, location, and severity of obstruction. The classic finding of obstruction is a loud systolic ejection murmur that increases in intensity with reductions in preload or afterload or an increase in left ventricular contractility, all of which tend to reduce ventricular volume and thereby increase obstruction. A murmur that increases in intensity from the squatting to the standing position or during the strain phase of the Valsalva maneuver is highly suggestive of a dynamic outflow obstruction. If this murmur is not present at rest or during these maneuvers, auscultation should be repeated during or immediately after exercise. In the absence of a murmur, under these several circumstances, the presence of clinically important obstruction should be questioned.
Two-dimensional and Doppler echocardiography can identify the degree and extent of hypertrophy and document the presence of SAM and midventricular obstruction\(^4,13\) (Figure 2). A late peaking systolic velocity jet across the outflow tract detected by continuous-wave Doppler echocardiography is a classic finding in obstructive HCM (sometimes referred to as HOCM), and the modified Bernoulli equation should be applied to the peak velocity to determine the severity of the obstruction. Midventricular obstruction is diagnosed by turbulent color flow Doppler in the midventricular region, accompanied by high velocity across the midventricle, which has a longer duration of flow extending into early diastole, from dyssynergy of contraction and relaxation of the apical portion of the myocardium (Figure 3).\(^2,22\) If SAM and a resting gradient are not present, then provocation with a Valsalva maneuver, amyl nitrite inhalation, or exercise with repeat imaging should be undertaken. In patients with severe symptoms in whom a high gradient cannot be elicited noninvasively, cardiac catheterization with isoproterenol challenge may be necessary to establish the presence of provokable obstruction\(^25\) (Figure 1).

The mainstay of therapy to relieve obstruction, and thereby alleviate symptoms, has been the combination of lifestyle changes and medical therapy, with the target of altering the contractility and load on the left ventricle to maintain its volume.\(^1-5\) Patients should always be well hydrated to maintain

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**Figure 1.** Cardiac catheterization pressures showing the dynamic nature of a left ventricular outflow tract obstruction in a patient with hypertrophic cardiomyopathy (HCM). **Top.** In the resting state, there is no left ventricular outflow tract obstruction between the left ventricle (LV) and aorta (AO). The left atrial (LA) pressure is normal. **Middle.** During infusion of isoproterenol, which increases contractility and reduces afterload, there is the onset of a dynamic outflow tract obstruction with a gradient of 35 mm Hg between the LV and AO. There is an elevation of LA pressure because of diastolic dysfunction. **Bottom:** At peak isoproterenol infusion, there is a marked reduction in central aortic pressure to 75 mm Hg with an LV outflow tract gradient of 45 mm Hg. There is a spike and dome pattern in the central aortic pressure. There is severe elevation of the LA pressure with a high V wave because of both diastolic dysfunction and concomitant secondary mitral regurgitation.
adequate left ventricular preload. They should avoid situations in which sudden changes in preload and afterload might occur, such as prolonged hot showers, hot tubs, or saunas. Rapid postural changes should be avoided, particularly after meals, when obstruction may be exacerbated. Medical therapy is successful in many patients, starting with $\beta$-blockade to reduce the ventricular contractility and heart rate, specifically to counter the increase in contractility that occurs during exertion. $\beta$-Blockade is usually titrated with increasing dosages to either eliminate the symptoms or attain a resting heart rate of $\approx 60 \text{ bpm}$. If a patient is intolerant of $\beta$-blockers, calcium channel blockers such as verapamil can be tried. Some patients may respond to the negative inotropic properties of disopyramide, which should be given with a $\beta$-blocker in patients with atrial fibrillation to prevent rapid conduction down the atrioventricular node.

However, there remains a subset of patients with a dynamic left ventricular outflow tract obstruction who either cannot tolerate medical therapy or remain significantly symptomatic despite such an optimal therapy. In these patients, septal reduction therapies, either surgical septal myectomy, or catheter-based percutaneous alcohol septal ablation (ASA) are effective in relieving the severe limiting symptoms. Implantation of a dual-chamber pacemaker with a short atrioventricular delay was attempted several decades ago to reduce obstruction by causing dysynchronous contraction of the septum. However, this approach has not been found to be efficacious.26

**Surgical Septal Myectomy**

**Historical Perspective**

Surgical septal myectomy for HCM has now been performed for >5 decades. The first surgical resection was described by Morrow and Brockenbrough, Kirklin and Ellis, and Brock. The original operation was a myectomy of the region of the septum projecting into the left ventricular outflow tract. The extent of septal excision, both width and length, initially described by Morrow and Brockenbrough and Morrow is considerably less than the extent of myectomy performed currently. Excision of protruding septal muscle results in enlargement of the left ventricular outflow tract with decrease in severity or complete elimination of the left ventricular outflow tract obstruction. Some surgeons have proposed implantation of a low-profile mechanical mitral prosthesis to treat both the obstruction and mitral regurgitation. However, in patients with concomitant mitral regurgitation secondary to SAM, the mitral regurgitation resolves from myectomy alone in >95% of patients. In addition, mitral valve replacement has its own adverse long-term consequences of a mechanical prosthesis,
as opposed to a septal myectomy, which is usually a definitive and long-lasting effective treatment.

There have been continued developments in this surgical procedure. Septal resection is now often extended distally to the level of the papillary muscles, to avoid residual midventricular obstruction (Figure 4). In some patients with abnormalities of the papillary muscles, dissection and reduction of the anomalous papillary muscle apparatus is performed. Intraoperative transesophageal echocardiography and direct measurements of left ventricular pressures are helpful because they can inform the surgeon if the myectomy has been adequate. There are some surgeons who perform a mitral valvuloplasty or plication of the deformed and elongated mitral valve leaflets with removal of secondary chordae and mobilization and reorientation of the papillary muscles. However, in the hands of experienced surgeons the extended septal myectomy alone has been shown to be effective in nearly all cases.

Immediate Outcomes and Complications

The early operative risk of septal myectomy included a 5% to 10% mortality rate with a high incidence of major complications including heart block, aortic regurgitation, and ventricular septal defect. When performed by experienced surgeons, isolated septal myectomy can now be performed with a <1% mortality. The other major complications are also rare now, with heart block requiring a permanent pacemaker occurring in <3% of patients and other major complications occurring in <1%. Hemodynamic results are excellent with most patients having a residual gradient <10 mm Hg, even during provocation. These results, however, are dependent on the skill and experience of the surgeon and cardiac center (Figure 5).

Long-Term Outcomes

The clinical results of surgical myectomy are outstanding, with >90% of patients being free of significant symptoms and most being able to return to a normal lifestyle. Importantly, postoperative functional improvement of patients with latent provokable outflow tract obstruction preoperatively is similar to that in patients with severe resting outflow tract obstruction. There are now data on several decades of follow-up of patients undergoing septal myectomy. Patients have been shown to maintain long-lasting improvement in symptoms and objective measurements of exercise capacity. In long-term follow-up, there has not been recurrence of obstruction because of regrowth of septal muscle. Recurrent symptoms and outflow tract obstruction is almost always related to inadequate septal excision at initial operation or the development of midventricular obstruction.

Prevention of Sudden Death

There have been no randomized trials that have demonstrated that myectomy reduces the frequency of sudden death in patients with HCM. However, it seems that long-term survival may be improved, particularly in younger patients with severe outflow tract obstruction. Survival after myectomy has
been shown to be equivalent to the expected survival of an age- and sex-matched general population and superior to that observed in a contemporary cohort of patients with outflow tract obstruction not undergoing myectomy. When compared with patients who are managed with medical therapy alone, a composite end point of sudden death or implantable cardiac defibrillator (ICD) discharge is reduced in patients who underwent septal myectomy. Patients who underwent ICD implantation for primary prevention had a discharge rate, which was significantly lower after septal myectomy versus those patients who did not undergo myectomy.

Because severe obstruction to outflow reduces coronary flow, relief of obstruction may prevent transient decreases in myocardial perfusion and subsequent subendocardial

**Figure 4.** Top. Septal myectomy performed through a low oblique aortotomy extending into the noncoronary sinus. With a no. 10 blade on a long handle, an incision is made in the septum beginning just to the right of the nadir of the right aortic sinus. This incision is carried upward initially and then leftward toward the anterior leaflet of the mitral valve (broken line). In the inset, the initial excision is carried further toward the apex of the left ventricular to remove hypertrophied septum beyond the endocardial scar.

**Figure 5.** Intraoperative left ventricular and aortic pressures taken before and after septal myectomy. **Top.** There is a severe obstruction before myectomy with a pressure gradient of 120 mm Hg, increasing to 200 mm Hg after a premature contraction (arrow). **Bottom.** After myectomy, there is no residual obstruction either during normal sinus rhythm nor after a premature contraction (arrow).
ischemia, which could trigger malignant ventricular arrhythmias. It is also possible that long-standing pressure overload results in deleterious secondary myocardial hypertrophy and fibrosis acting on the genetically abnormal myocardium, which increases the development of heart failure and enhances the substrate for arrhythmias. It would not be expected that any type of surgical procedure would be able to totally prevent sudden death but perhaps relief of obstruction may be able to decrease significantly the incidence of sudden death, as well as of heart failure, particularly in young patients who are at increased risk.

Additional Surgical Interventions

Surgical intervention provides the ability to treat other concomitant cardiac abnormalities, which might not be treated with medical therapy or catheter-based therapy alone. In ≈6% of patients undergoing septal myectomy, there are intrinsic abnormalities of the mitral valve apparatus, such as ruptured chordae tendineae, contributing to severe mitral regurgitation. In these patients, relief of the obstruction alone would not provide relief of severe mitral regurgitation and additional mitral valve repair is required. Another subset of patients present with symptoms felt to be because of a dynamic left ventricular outflow tract obstruction but are found also to have a fixed obstruction at the time of operation. This can be due either to congenital discrete subaortic stenosis or the occurrence of a fibrotic area of scarring of the interventricular septum at the site of contact with the systolic anterior motion of the mitral valve in HCM. This requires resection of the discrete subaortic stenosis and the extended septal myectomy. Rarely, patients may have coexisting calcific or rheumatic mitral valve disease in association with HCM and dynamic outflow tract obstruction, which may require surgical correction.

Patients with the apical variant of HCM have severe cavity obliteration and severe diastolic dysfunction. Novel surgical techniques for a myectomy using an apical approach have been shown to improve the compliance of the left ventricle and, thus, improve severe dyspnea (Figure 6). In patients with midventricular obstruction and development of an apical aneurysm, resection of the midventricular obstruction through the apical approach and resection of the apical aneurysm is an innovative surgical technique that has been shown to improve significantly the intractable symptoms in this subgroup of patients.

Outcomes at HCM Centers of Excellence

The results of surgical myectomy are highly dependent on the experience of the surgeon and the entire surgical team. There are HCM centers of excellence in which patients are fully evaluated by teams of experts in the field of HCM, coupled with highly experienced surgeons who have developed great expertise in this operation. Recent data from the nationwide inpatient registry suggests that the real world mortality rate associated with myectomy ranges from 4% to 16% as compared with the low mortality rates of <1% found in the best high-volume centers. At the less experienced centers, the complications of ventricular septal defect, complete heart block requiring permanent pacemaker, and inadequate relief of the obstruction are higher than at the HCM centers of excellence. The ACC/AHA Guidelines for the Diagnosis and Treatment of Patients with HCM have recommended that septal reduction therapy should be performed only by experienced operators in the context of a comprehensive HCM clinical program, with the goal of a <1% operative risk for isolated septal myectomy and a major complication rate of <3%.

Septal Ablation and Other Catheter-Based Treatments

Historical Perspective

After the introduction and widespread adoption of percutaneous coronary angioplasty, interventional cardiologists began to develop percutaneous approaches to the treatment of valvular and structural heart diseases. Important ideas of catheter intervention for septal reduction therapy in patients with HCM were the observation of Sigwart, Sigwart et al, and Kuhn et al, who reported on reduction of systolic wall motion by temporary balloon occlusion of the coronary artery perfusing the septum. Furthermore, it had been shown that myocardial infarction may result in disappearance of left ventricular obstruction in patients with HCM. Septal artery occlusion by injection of alcohol was described as a treatment for patients

Figure 6. Schematic diagram of apical hypertrophic cardiomyopathy (HCM). Top Left. In some patients with apical HCM, severe hypertrophy reduces left ventricular end-diastolic volume. Bottom Right. Transapical incision and myectomy of the septum and midventricle, as illustrated in yellow shading, can improve effective compliance with resulting increase in ventricular stroke volume and decrease in end-diastolic pressure.
with malignant ventricular tachycardia by Brugada et al., Sigwart, and G. Berghoefer (personal communication) developed the hypothesis that occlusion of a septal coronary branch with subsequent infarction would treat obstruction in patients with HCM.

Sigwart was the first to treat patients with HCM with percutaneous ASA and published favorable results of the first 3 patients in 1995. His article raised questions about the long-term effect of an induced myocardial scar in patients who were already prone to malignant ventricular arrhythmias. However, despite this concern, especially after introduction of myocardial contrast echo guidance, ASA has become popular worldwide as a less invasive alternative to surgical septal myectomy for the treatment of symptomatic patients with HCM (Online Table I).

**Procedure**

**Description**

Most centers now use echocardiography-guided ablation, which was introduced by Faber et al. and Seggewiss et al. (Figure 7). Before the ablation, an initial diagnostic catheterization to measure the left ventricular outflow tract is performed. Coronary arteriography is required to exclude coronary artery disease and define a potential target septal artery; the first large septal branch or one of the first side branches is considered the target artery. Both catheterization and Doppler echocardiography can also be used to measure the outflow pressure gradient.

Because of the risk of development of complete heart block, a temporary pacemaker lead should be inserted in patients without a permanent pacemaker or ICD in place. A guidewire is advanced into the target septal artery; an over-the-wire balloon is then advanced into the target septal artery and inflated to avoid backflow of alcohol into the left anterior descending artery, thereby avoiding infarction of nontarget myocardial areas. After the guidewire is withdrawn, echocardiographic contrast agent is injected through the balloon catheter with simultaneous transthoracic echocardiography. Selective angiography of the target septal branch through the inflated balloon catheter should document the adequate sealing of the septal branch and exclude filling of any other coronary artery through septal collaterals.

Up to 3 mL of absolute alcohol is then injected slowly through the central lumen of the balloon catheter under continuous fluoroscopic, hemodynamic, and electrocardiographic observation. The quantity of injected alcohol should be determined by the septal thickness or septal artery diameter. After withdrawal of the balloon catheter, a final angiogram is performed to document complete occlusion of the septal branch and normal flow in the left anterior descending artery. Measurement of the outflow tract pressure gradient at rest and with provocation is repeated. At least 48 to 72 hours of hemodynamic and electrocardiographic surveillance are necessary. Cardiac enzyme measurements every 6 to 8 hours allow documentation of peak creatine kinase value. Depending on the rhythm, a decision is made on implantation of a permanent pacemaker.

**Figure 7.** Sequence of echo-guided alcohol septal ablation. A, Simultaneous pressure recording of left ventricular inflow tract pressure and aortic pressure with high gradient at rest and post extrasystole. B, Baseline angiogram of the left coronary artery with estimated target septal branch (black arrow) and documentation of temporary pacemaker lead (white arrow). C, Position of the small over-the-wire balloon in the target septal branch (black arrow). D, Baseline modified 4-chamber view echo with systolic anterior motion (SAM)–septal contact (arrow). E, Documentation of echo-contrast depot in the subaortic part of the septum at the site of SAM–septal contact point (arrow). F, Injection of angiographic contrast media through the lumen of the over-the-wire balloon (arrow). G, Occluded septal branch (arrow) after balloon retraction 10 min after last alcohol injection without damage of the left anterior descending artery. H, Final hemodynamic result without gradient at rest and post-extrasystolic beat.
pacemaker or of a defibrillator if there is an increased risk of 
sudden cardiac death according to the clinical risk stratification 
types.24

Immediate Hemodynamic Results and Complications
Since the first report of ASA in patients with HCM,53 many 
interventional cardiologists have documented acute hemody-
namic improvements with a gradient reduction of >50% 
in ≥90% of patients.65.68–81 Faber et al82 compared the results 
in 61 patients treated using echo-guided ASA with those in 
the initial 30 patients without echo guidance. The acute grad-
ient reduction (>50%) was 92% with echo guidance and 70% 
without. The need for permanent pacemaker implantation was 
reduced as well (7% versus 17%). This reduction of left ventric-
ular outflow tract obstruction resulted in symptomatic im-
provement and in increased exercise capacity.

There has been a continuous reduction of complications 
because of increased experience of the interventional cardiol-
gists. The most common complication after ASA is complete 
heart block, which may require permanent pacing. Patients 
with pre-existing left bundle branch block are at higher risk 
of developing complete heart block as nearly one half of the 
patients develop complete right bundle branch block during 
the procedure.83 Preprocedural ICD implantation is reason-
able for patients who have an increased risk of sudden cardiac 
decath under commonly used risk stratification mod-
els.1,2,24 Contemporary practice involving myocardial contrast 
echocardiography and the use of lower alcohol volumes has 
resulted in a reduced need for permanent pacing after the in-
tervention, now approaching <10%,64,84,85 Other complications 
include coronary artery dissection, coronary spasm, ventricu-
lar fibrillation, cardiac tamponade, pulmonary embolism, car-
diogenic shock, stroke, and problems with the puncture site.69

There is a steep learning curve for the optimal perfor-
mances of ASA.78,86 Complications are rare with experienced 
operators working in high-volume centers where the 30-day 
mortality for septal ablation is now <1%, with major adverse 
cardiac events occurring in <2% of patients.87–89

Intermediate-Term Outcomes

Hemodynamic and Symptomatic Outcomes
Follow-up studies have shown ongoing hemodynamic im-
provement with further reduction of the left ventricular out-
flow tract obstruction during the first year after ASA (Online 
Figure 1).72 Because of the anatomic variation of the septal 
artery system,70 14% of patients in one series required a sec-
tond ablation to get an adequate result.90 Successful ablation 
results in reduction of SAM-related mitral regurgitation72 and 
pulmonary artery pressure.64 Hemodynamic improvement 
after ASA is associated with symptomatic improvement and 
increased exercise capacity.92 The subjective improvement has 
been verified by objective measurements of exercise perform-
ance on cardiopulmonary exercise testing.64,93,94 In recent 
registries and multicenter studies, the mean New York Heart 
Association Functional Classification (NYHA) class of dis-
ability decreased from an average of 2.9 to 1.6 after 5 years. 
Whereas ≥95% of patients were NYHA class III–IV before 
the procedure, ≥20% of patients remained in these classes, 
with the remaining patients having minimal or no symptoms. 

The mean residual left ventricular outflow tract gradient de-
creased from 60 to 70 mm Hg to 15 to 20 mm Hg. Patients 
with higher residual gradients have an increased all-cause 
mortality at follow-up.55,88,89,95

With introduction of ASA for symptomatic patients with 
HCM, concern was raised on the potential negative effect of 
creating a myocardial scar in patients who were already at risk 
for ventricular arrhythmias.54 This was borne out by initial 
case reports of ventricular arrhythmias occurring after ASA.96 
Magnetic resonance imaging scanning with gadolinium en-
hancement demonstrated a large scar in the myocardium at the 
site of the ablation, similar to that of a localized myocardial 
infarction.97 Several studies showed a high rate of defibrillator 
failings after ASA in patients who had defibrillators in place 
before the procedure.98,99

The first published intermediate-term follow-up (≤8 years) 
included 100 consecutive symptomatic patients (NYHA class 
2.8±0.6) treated with echocardiography-guided technique.100 
Only one sudden cardiac death was observed, and event-free 
survival was 74%. A more recent study on 178 consecutive pa-
tients treated with ASA provided similar results. Survivals after 
1, 5, and 10 years were 97%, 92%, and 82%, respectively, and 
did not differ from survival in an age- and sex-matched general 
population. The only independent predictor of all-cause mortal-
ity was age at the time of ASA.10 A study on 470 consecutive 
patients treated with echo-guided ASA between 1996 and 2010 
in Germany and Denmark addressed the question of sudden car-
diac death during follow-up. Before ablation, 25% of the pa-
tients had ≥2 risk factors for sudden death compared with 8% after ab-
lation (P<0.001). The observed 10-year survival was 88%, and 
survival free of sudden death was 95%, which was similar to that 
of a matched general population.95 Veselka et al reported on the 
results of the Euro-ASA registry in which 1275 patients under-
went the procedure in 10 European tertiary referral centers in 7 
countries. The 10-year overall survival was 77%, whereas sudden 
decath-free survival was 90%. Follow-up in this study exceeded 
7000 patient-years with a sudden death rate of 0.6% per year. The 
combined rate of sudden and cardiovascular mortality was rela-
tively low at 1.2% per patient-year.101,102 Thus, in the most recent 
registries and multicenter studies of patients undergoing ASA at 
highly experienced centers for an average follow-up of 5 years, 
there is no evidence that there is an increased risk of sudden death 
or increased mortality after septal ablation.54,87,88,95,100,103,104

Comparisons Between Septal Myomectomy 
and ASA
As summarized above, multiple studies have demonstrated a high 
success rate and low complication rate with both septal myomect-
tomy and ASA, leading to excellent reduction in outflow tract 
obstruction and sustained improvement in symptoms. The choice 
of procedure is dependent on many factors including the exper-

cise and availability of the operators, the presence of comor-
btant cardiac problems, accompanying medical comorbidities, and 
ultimately patient choice (Table). Patients who are candidates for 
this therapy should undergo a full evaluation from a team with ex-
pertise in the diagnosis and management of patients with HCM. 
Both procedures should be performed by experienced operators. 
In choosing which of those procedures should be selected, it is
Table. Selection of Septal Ablation vs Septal Myectomy

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<th>Septal Ablation</th>
<th>Septal Myectomy</th>
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<tr>
<td>Patient preference: less invasive,</td>
<td>Patient preference: most effective,</td>
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<tr>
<td>shorter recovery</td>
<td>longest follow-up</td>
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<td>Expertise widely available</td>
<td>Expertise limited to few HCM centers</td>
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<td>Increased risk PPM with normal</td>
<td>Increased risk PPM with RBBB</td>
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<td>Multiple comorbidities at high</td>
<td>Address other problems: fixed</td>
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<td>atrial arrhythmias, multivessel CAD,</td>
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<td></td>
<td>and mitral and apical hypertrophy</td>
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<td>Mild hypertrophy, elderly patients</td>
<td>Massive hypertrophy, younger patients</td>
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CAD indicates coronary artery disease; HCM, hypertrophic cardiomyopathy; LBBB, left bundle branch block; PPM, permanent pacemaker; and RBBB, right bundle branch block.

table: Selection of Septal Ablation vs Septal Myectomy

important to understand that no randomized clinical trial comparing the 2 approaches has been conducted.

Septal ablation is more widely available worldwide than septal myectomy. In experienced centers, clinical outcomes are similar to those of septal myectomy in most patients, Nonetheless, there is a subset of patients in whom ASA will not be effective if the area of the SAM–septal contact cannot be targeted through a septal perforator artery. Overall, the residual outflow pressure gradient is somewhat higher after ASA (average 20 mm Hg) versus myectomy (average <10 mm Hg). Myectomy is more effective than ASA in the presence of massive septal hypertrophy, which may be accompanied by midventricular obstruction for which an extended myectomy can completely relieve all levels of obstruction. In high-volume centers that offer both myectomy and ASA, it has been observed that in younger patients, particularly those with massive septal hypertrophy, there is a higher percentage of patients who experienced complete relief of symptoms after myectomy than after ASA.

In older patients who may have a lesser degree of hypertrophy, the symptomatic outcomes of the 2 procedures were similar.

All patients undergoing either septal myectomy or ASA should be evaluated for risk of sudden death and appropriate implantation of an ICD for primary prevention according to conventional guidelines and risk scores. It is still unclear as to whether overall survival and incidence of sudden death are similar after the 2 procedures. Vriesendorp et al reported that the overall mortality was similar between patients undergoing myectomy, ASA, and medical therapy; however, the risk of sudden cardiac death was lowest after myectomy. In other studies, there did not seem to be a difference in overall mortality comparing septal myectomy versus ASA. In 1 study of 601 patients referred for invasive therapy, 138 underwent ASA. Although patients <65 years of age showed greater symptomatic improvement with myectomy, the overall 4-year survival after the 2 procedures was similar. This documented efficacy of ASA was confirmed in another nonrandomized comparison of 177 patients who underwent ASA and followed up for 5.7 years and who were compared with an age- and sex-matched cohort of patients who underwent myectomy. Survival after both procedures was equal and did not differ from survival in an age- and sex-matched general population. Age and residual outflow pressure gradient were independent predictors of long-term survival free of all-cause death. Steggerda et al reported a single-center study in the Netherlands, which compared 161 patients after ASA with 102 patients after myectomy during a follow-up period of ≤11 years. Annual cardiac mortalities after ASA and myectomy were comparable (0.7% versus 1.4%; P = 0.15).

From the patients’ standpoint, septal myectomy involves the discomfort and longer recovery time associated with open-heart surgery, as opposed to the less invasive catheter-based therapy in ASA. In patients who are at higher risk for open-heart surgery because of other comorbidities, multiple previous cardiac operations, or frailty, ASA poses less overall risk. However, septal myectomy can address other concomitant cardiovascular problems at the time of the procedure, such as primary mitral valve and aortic valve disease, fixed subaortic obstruction, midventricular hypertrophic obstruction, coronary artery disease, and atrial arrhythmias. Despite substantial improvement in the technique, the risk of requiring a permanent pacemaker remains somewhat higher with ASA than septal myectomy. Also, although there is a lower (but still high) chance of complete symptomatic response after a first ASA, infrequently a repeat procedure may be necessary to achieve complete resolution of obstruction and relief of symptoms.

Ultimately, the final decision as to which procedure for septal reduction should be performed is dependent on 2 major factors. The first is patient preference, and a shared decision-making approach should be pursued, discussing the risks and benefits of each approach, then understanding the needs and preferences of the individual patient. The second important factor is the availability and experience of the operator and institution at which the patient is being treated. Both procedures require skill and expertise by highly experienced operators, as well as a multidisciplinary approach for patient evaluation. HCM centers with high-volume surgical programs performing septal myectomy are not universally available to all patients who are candidates for and require septal reduction therapy, and the results of operation in less experienced centers are associated with higher mortality and complication rates. There is less difference in the outcome of ASA between low- and high-volume centers, but certainly the optimal results of ablation are from the highly experienced operators.

There are continuing improvements in both techniques of septal reduction. Longer-term follow-up of larger number of patients will be important to understand the optimal roles of each procedure in the management of these patients. Transparency and reporting of an institution’s outcome are essential so that all patients can make an educated decision on the choice of procedure.

Disclosures

None.

References


Nishimura et al Treatment of HCM 783


### Table I: Acronyms of different techniques of alcohol septal ablation techniques with different acronyms

<table>
<thead>
<tr>
<th>Acronym</th>
<th>Name</th>
<th>Identification of target septal branch</th>
<th>Criterion of completion</th>
</tr>
</thead>
<tbody>
<tr>
<td>PTSMA</td>
<td>Percutaneous Transluminal Septal Myocardial Ablation</td>
<td>Transthoracic myocardial contrast echocardiography</td>
<td>Alcohol depot in echocardiography</td>
</tr>
<tr>
<td>TASH</td>
<td>Transcoronary Ablation of Septal Hypertrophy</td>
<td>Haemodynamic effect of temporary branch occlusion</td>
<td></td>
</tr>
<tr>
<td>NSRT</td>
<td>NonSurgical Septal Reduction</td>
<td>Echocardiographic study with the use of angiographic contrast dye</td>
<td></td>
</tr>
<tr>
<td>NSMR</td>
<td>NonSurgical Myocardial Reduction</td>
<td>Haemodynamic effect of temporary branch occlusion</td>
<td>Occlusion of several septal branches</td>
</tr>
<tr>
<td>PTSA</td>
<td>Percutaneous Transluminal Septal Ablation</td>
<td>Transthoracic myocardial contrast echocardiography</td>
<td>Repeated alcohol injection in the same branch</td>
</tr>
<tr>
<td>ASA</td>
<td>Alcohol Septal Ablation</td>
<td>Lately prevalent general term mainly (but not always) referring to the echocardiography guided technique</td>
<td></td>
</tr>
</tbody>
</table>

Acronyms of different techniques of alcohol septal ablation techniques with different acronyms
Mean ± SD of left ventricular outflow tract (LVOT) obstruction gradients at rest and during the stress of the Valsalva maneuver in 100 consecutive patients with echo-guided alcohol septal ablation up to 58±14 months following the procedure. Reproduced with permission from Seggewiss et al. Clin Res Cardiol 2007;96:A856 [104]
Correction to: Hypertrophic Obstructive Cardiomyopathy
Surgical Myectomy and Septal Ablation

In the article by Nishimura et al, “Hypertrophic Obstructive Cardiomyopathy: Surgical Myectomy and Septal Ablation,” which published in the September 15, 2017 issue of the journal (Circ Res. 2017;121:771-783. DOI: 10.1161/CIRCRESAHA.116.309348), a correction was needed.

The corresponding author’s email address has been corrected.

This correction has been made to the current online version of the article, which is available at http://circres.ahajournals.org/content/121/7/771.