

# Multiple alcohol septal ablations in a young patient with hypertrophic cardiomyopathy

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## Abstract

*A 16 year old female with hypertrophic cardiomyopathy was treated with alcohol ablation for NYHA class III symptoms on medical therapy. Three months later, patient underwent a second alcohol ablation procedure for continued symptoms. Follow-up, for 4 years now, continues to show resolution of symptoms. (Cardiol J 2007; 14: 301–304)*

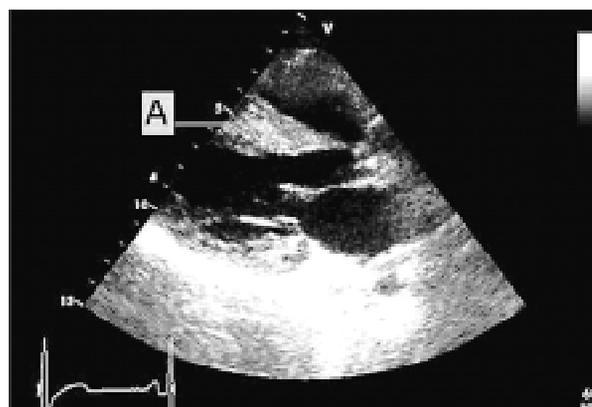
**Key words:** hypertrophic cardiomyopathy, alcohol ablation, surgical myectomy

## Introduction

Hypertrophic cardiomyopathy (HCM) is a genetic disease characterized by hypertrophy of the left ventricle and in a subset of patients, obstruction of the left ventricular outflow tract (LVOT) [1, 2]. Significant outflow tract obstruction is an independent predictor of poor prognosis in patients with HCM [3]. The incidence of LVOT obstruction has been demonstrated to be present in higher than previously reported percentage of patients when exercise stress testing was used to provoke a gradient [4]. In the treatment of obstructive HCM non-pharmacologic options include surgery and transcatheter ethanol septal reduction. Dual chamber pacing has only a limited role. Patients with obstructive HCM are candidates for septal reduction therapy if they are severely symptomatic (New York Heart Association functional class III or IV) or have recurrent syncope despite pharmacologic therapy.

## Case report

A 16 year old female with a family history of hypertrophic cardiomyopathy presented with symptoms of increasing fatigue and NYHA functional class III. She had a loud systolic ejection murmur at rest, which increased with Valsalva maneuver. Echocardiography showed septal hypertrophy with septal thickness of 27 mm (Fig. 1), systolic anterior motion (SAM) of mitral valve leaflet, resting LVOT gradient of 70 mm Hg which increased to 100 mm Hg with Valsalva maneuver. She had an episode consistent with presyncope during physical



**Figure 1.** Pre-ablation parasternal long axis echocardiograph showing thickened septum (A) and narrow left ventricular outflow tract.

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activity and a family history of ventricular arrhythmias. Patient underwent ICD implantation and was on medical therapy with beta blockers and calcium channel blockers. Medications were very poorly tolerated because of hypotension and fatigue. Patient was no longer able to attend school. Options of surgical *vs.* transcatheter ethanol septal reduction procedures were discussed with her family and her. A decision was made to proceed with ethanol septal ablation. First septal perforator branch of the LAD was engaged with angioplasty catheter and slow infusion of ethanol was performed. Immediate resolution of LVOT gradient was noted. Systolic ejection murmur was no longer present. Patient was monitored in the intensive care unit, cardiac troponin peaked at 30 and ECG showed septal Q waves. The next post-procedure day, physical examination demonstrated recurrence of systolic murmur at rest. Echocardiogram confirmed recurrence of LVOT gradient and presence of SAM of mitral valve leaflet. The hope was that recurrence of gradient was in part due to increased edema of infarcted proximal septal tissue and that with development of scar and left ventricular remodeling there will be improvement in LVOT gradient. Clinically, patient reported symptomatic improvement in her fatigue and she was able to return to school. Serial echocardiograms showed development of small scar in the proximal septum but persistence of LVOT gradient and SAM. Within three months patient also had recurrence of symptoms. A decision was made to repeat the transcatheter ethanol septal reduction procedure with injection of ethanol into the second and third septal perforator branches of LAD. Immediate results of this procedure were equally rewarding as the first one. There was immediate resolution of LVOT gradient in the catheterization laboratory and disappearance of systolic murmur. This time troponin peaked at 50. We were disappointed when on the second post-procedure day the murmur returned with the same intensity and again LVOT obstruction was demonstrated by echocardiography. But this time patient felt significantly better and had objective improvement in physical activity tolerance. She returned to school full time and was able to finish high school. Serial echocardiograms demonstrated gradual thinning of the interventricular septum, disappearance of LVOT obstruction and SAM by the third month post second procedure. Septal remodeling with progressive septal thinning was observed to occur over period up to 1 year (Fig. 2). Patient was able to attend college and is now working full time as a paramedic. She has no limitations of physical activity and is able to exercise regularly.



**Figure 2.** Post-ablation (2<sup>nd</sup> ablation) parasternal long axis echocardiography showing septal scar (A) with resolution of left ventricular outflow tract obstruction.

## Discussion

With surgical myectomy, the proximal septum is approached through the aortic valve via an aortotomy and 3 to 15 g of septal muscle is removed [5–8]. Perioperative mortality is reported to be less than 1% and centers with extensive experience report even lower mortality in uncomplicated cases. Complications of myectomy include a ventricular septal defect due to excessive removal of septal muscle, ischemia of the hypertrophied muscle due to inadequate intraoperative protection and resulting left ventricular dysfunction, aortic valve regurgitation due to traction on the aortic valve to improve visualization of and access to the interventricular septum, and left bundle branch block (LBBB) or complete heart block (CHB) requiring a permanent pacemaker in approximately 5–10% of patients. Reduction of LVOT gradient, improvement in functional class and long-term survival is excellent after surgical myectomy at experienced centers [9].

Transcatheter ethanol septal ablation consists of infarction and thinning of the proximal interventricular septum via infusion of ethanol into the first septal perforating branch of the left anterior descending coronary artery through an angioplasty catheter. Ethanol septal ablation reduces LV outflow obstruction, improves symptoms, and increases exercise capacity. Long-term benefit results from the creation of localized septal infarction and scarring, which increase LV outflow diameter as a result of septal thinning and “therapeutic remodeling” [10–13]. This procedure significantly reduces the resting LVOT gradient, the inducible gradient and improves functional class [14]. A subset of patients who do not show initial benefit can have later improvement [15]. These late responders had

a similarly improved LVOT gradient at one year compared to those who improved immediately after the procedure. Complete heart block (CHB) requiring a pacemaker occurs in 14–25% of patients after ethanol septal ablation [16, 17].

Both surgical myectomy and ethanol septal ablation reduce LV outflow tract obstruction and improve NYHA class in HCM. When efficacy of the two techniques was compared the resting LVOT gradient fell to a similar degree with both ethanol ablation and surgical myectomy, there was similar reduction in septal thickness and improvement in exercise duration in both groups. The incidence of CHB was higher with ethanol ablation, but surgery was associated with a significant increase in mild aortic regurgitation [18]. A recent study showed consistent anterior basal septal reduction with myectomy but not with alcohol ablation, as assessed by cardiac magnetic resonance imaging [19].

The advantages of ethanol ablation include avoidance of cardiopulmonary bypass, shorter hospital stay and recovery time and reduced expense. Advantages of surgical myectomy include more immediate and complete relief of resting and provoked obstruction and concomitant mitral regurgitation, lower incidence of CHB requiring pacemaker insertion and proven long-term (> 20 years) efficacy; similar data with ethanol ablation are not yet available.

In our patient, four year follow up shows satisfactory results with improved functional capacity, lack of LVOT gradient and preservation of left ventricular function without development of complete heart block. However the degree and extent of the left ventricular septal thinning is concerning to us. In retrospect, we probably should have shown more restraint and allowed more time for the remodeling before the second ablation procedure.

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