Alcohol Septal Ablation after Suboptimal Surgical Septal Myomectomy

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INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a common inherited cardiovascular disease characterized by hypertrophy of a nondilated left ventricle in the absence of any other cardiac or systemic disease (such as hypertension) that could account for the observed hypertrophy. HCM is caused largely by mutations in genes encoding thick and thin contractile myofilament proteins of the cardiac sarcomere. Phenotypically, HCM can be obstructive (in 70% of patients), with the presence of left ventricular outflow tract (LVOT) obstruction, or nonobstructive (in 30% of patients).1

In symptomatic patients with LVOT obstruction, symptom relief may be achieved by medical therapy,2 surgical myomectomy,3 or alcohol ablation.4

Alcohol septal ablation (ASA) was first described in 1994 as a less invasive modality for the treatment of symptomatic hypertrophic obstructive cardiomyopathy compared with surgical myomectomy.4 The procedure has been improved in recent years, especially with the use of myocardial contrast echocardiography for localization of the target area, which results in improved safety and a decreased amount of alcohol used.5

Since it was first described in 1975, surgical myomectomy remains the gold standard for obstruction relief in symptomatic patients.5

The choice between the two modalities depends mainly on expert opinion and individualized decision making, because of the lack of head-to-head comparisons in randomized trials.

In case of suboptimal ASA results, surgical myomectomy results in less favorable outcomes compared with ASA-naive patients. A case series of 31 patients with surgical myomectomy showed that those with prior failed ASA had an increased risk for cardiac death, advanced heart failure, and implantable cardioverter-defibrillator discharges compared with patients without prior ASA.6

However, sufficient data are lacking regarding outcomes of ASA in patients with prior failed surgical myomectomy. Here we describe a patient with obstructive HCM who underwent ASA after a prior failed surgical myomectomy.

CASE PRESENTATION

The patient was a 63-year-old man who presented to our clinic in May 2015 with exertional dyspnea of 2-year duration with insidious onset and a slowly progressive course. His medical history was noncontributory except for hypertension. His vital signs included blood pressure of 160/100 mm Hg, resting heart rate of 120 beats/min, respiratory rate of 18 breaths/min, and oxygen saturation of 98%. Cardiac auscultation revealed normal first and second heart sounds and a 2/6 systolic murmur over the apex. There were no congested neck veins. Neither lower limb edema nor signs of pulmonary congestion were observed.

Initial electrocardiography showed normal sinus rhythm and ST-segment elevation in precordial leads V1 to V3.

Transesophageal echocardiography showed hypertrophic left ventricular (LV) ejection fraction > 70% and grade 1 diastolic dysfunction. Asymmetric septal hypertrophy measuring 24 mm in diastole was observed (Figure 1). There was systolic anterior motion (SAM) of the mitral leaflet (Figures 2A and 2B, Video 1), with LVOT obstruction and a resting systolic peak pressure gradient of 46 mm Hg increasing to 118 mm Hg with sublingual nitrate (Figures 3A and 3B). A moderate posteriorly directed mitral regurgitation jet was also noted (Figure 4, Video 2).

The patient was started on atenolol and up-titrated to a maximum dose of 100 mg/d, with no significant improvement. The case was discussed by the heart team, and septal myomectomy was decided. The procedure was done in January 2016. Intraoperative transesophageal echocardiography was not performed, as it was considered nonessential in this particular case from the surgeon’s perspective.

After discharge, the patient experienced the same previous exertional symptoms, which continued to affect his daily activities.

Follow-up echocardiography performed in May 2016 showed normal LV systolic function (ejection fraction 56%), maximal septal thickness of 17 mm (Figure 5), residual SAM of the anterior mitral leaflet (Figure 6), and a resting gradient of 47 mm Hg that increased to 60 mm Hg with the Valsalva maneuver (Figures 7A and 7B). Mild mitral regurgitation was also present.

Medical therapy was maximized, including atenolol 100 mg orally in the morning and 50 mg at night.

The patient’s symptoms did not diminish. A treadmill stress test was done and showed an abnormal response to exercise and a decrease of systolic blood pressure, with reduced exercise tolerance, as the patient was not able to exceed 6 METs.

Repeat echocardiography in November 2016 showed a resting peak LVOT gradient of 85 mm Hg that increased to 136 mm Hg with the Valsalva maneuver (Figures 8A and 8B). There was SAM of the anterior mitral leaflet with a moderate posteriorly directed regurgitation jet (Figure 9).

Upon discussion with the patient, he rejected the option of redo surgery, so we decided to proceed to ASA.

The procedure was done on February 27, 2017. Invasive peak-to-peak pressure gradient across the LVOT was 80 mm Hg at rest, with typical Brockenbrough-Braunwald phenomenon after inducible premature ventricular beats (Figures 10A and 10B).
Two large septal perforators were identified. The first septal perforator was crossed with Terumo RUNTHROUGH wire (Terumo, Tokyo, Japan) and a Sprinter OTW balloon (1.5 mm x 10 mm; Medtronic, Minneapolis, MN). The balloon was inflated at 10 mm Hg, and because of a lack of echocardiographic contrast agent, simple agitated saline was used for myocardial contrast echocardiography, which clearly opacified the basal part of the septum (Figures 11A and 11B, Video 3); no other nontarget segments were opacified. No collateral circulation was identified in continuation of the target septal. Alcohol was injected twice, with a total delivery of 1.5 mL 98% alcohol (Figure 12, Video 4). Immediately after alcohol injection, the highest peak-to-peak gradient was 19 mm Hg (Figures 13 and 14). No post–premature ventricular contraction aortic pulse pressure change was noted.

The patient developed complete heart block and required a temporary pacemaker, which was converted to a dual-chamber permanent pacemaker.
pacemaker when the patient’s rhythm did not recover beyond 48 hours.

The patient had a remarkable clinical response, with complete relief of exertional symptoms 6 months later and markedly improved exercise tolerance; currently, he is able to carry on his daily activities without any limiting symptoms.

In October 2017, the patient was seen in our cardiology clinic. He was completely asymptomatic (New York Heart Association functional class I), and electrocardiography showed sinus rhythm with recovered heart block (Figure 15).

Follow-up echocardiography in October 2017 showed normal LV size and systolic function, septal hypokinesia, trace mitral regurgitation, and an LVOT peak pressure gradient < 10 mm Hg at rest; the highest provoked peak pressure gradient with the Valsalva maneuver was 13 mm Hg (Figure 16, Video 5).

**DISCUSSION**

Given the risk of redo open heart surgery in patients with obstructive HCM after failed septal myomectomy, ASA when performed properly is a valid and safe therapeutic option for persistent symptoms of obstruction.

Myocardial contrast echocardiography is an important intraprocedure tool for the selection of the proper septal branch and success of the procedure. Suboptimal ASA results are due largely to suboptimal scar location. Intraoperative myocardial contrast echocardiography can lead to a change in the selection of the septal branch and even cancellation of the procedure.7 The use of intraprocedural contrast echocardiography results in improved procedural and patient outcomes by limiting the infarct to the targeted region of interest. Advantages include shorter intervention and fluoroscopy times, fewer occluded vessels, less ethanol use, a smaller infarct size, a lower likelihood of heart block, and a higher likelihood of success.11

The American Society of Echocardiography recommends the use of echocardiographic contrast agent for myocardial contrast.8 However, if contrast agent is not available, simple agitated saline may be used successfully as an alternative.

High-degree atrioventricular block is common after ASA; however, the conduction system may recover after some time. Precise assessment of symptoms and functional capacity aid the decision for intervention in patients with obstructive HCM. Intraoperative transesophageal echocardiography is an essential tool during surgical myectomy. It allows evaluation of the immediate results of the myectomy, ensuring that SAM and LVOT obstruction have been resolved. The development of a heart team with optimal communication among the surgeon, cardiac interventionalist, and echocardiographer is essential for managing challenging cases.

**CONCLUSION**

ASA after failed surgical septal myomectomy when performed with the aid of myocardial echocardiography contrast is safe and may be a very effective therapeutic option. Agitated saline may be used safely as an alternative when echocardiographic contrast agent is not available.
Figure 7 Postoperative Doppler imaging showed an LVOT of 40-45 mm Hg, which increased to 80 mm Hg. (A) The resting peak pressure gradient across the LVOT was 47 mm Hg. (B) The provoked peak pressure gradient with Valsalva maneuver increased to 80 mm Hg.

Figure 8 Follow-up echocardiogram after maximizing medical therapy. (A) The resting peak pressure gradient across the LVOT was 85 mm Hg. (B) The provoked peak pressure gradient with Valsalva maneuver increased to 136 mm Hg.
Figure 9 Echocardiographic findings for maximum medical therapy. (A) Persistent systolic anterior motion of the anterior mitral leaflet. (B) Color Doppler showing mild posteriorly directed mitral regurgitation.

Figure 10 Invasive hemodynamic assessment before ablation. (A) The peak-to-peak pressure gradient was 80 mm Hg. (B) Typical Brockenbrough-Braunwald sign after inducible premature ventricular beats.
**Figure 11** Two-dimensional echocardiography after injection of “agitated saline” covering the SAM-septum contact area. **(A)** Modified apical four-chamber view. **(B)** Modified apical three-chamber view.

**Figure 12** **(A)** The culprit septal is wired with Terumo RUNTHROUGH wire. **(B)** Alcohol injection through an over the wire balloon.

**Figure 13** Doppler assessment after ablation.
Figure 14 Pressure tracing immediately after ablation showed a peak-to-peak gradient of less than 20 mm Hg.

Figure 15 (A) Complete heart block with right bundle branch block. (B) On temporary pacemaker. (C) On atrial sensed ventricular paced rhythm. (D) Sinus rhythm with left bundle branch block.
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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.02.004.

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Figure 16 Two-dimensional echocardiography in October 2017 after ASA showed no significant gradient across the LVOT. (A) Baseline peak pressure gradient. (B) Provoked peak pressure gradient after Valsalva maneuver (13 mm Hg).