

Nonobstructive septal hypertrophy in a young adult provoking recurrent polymorphic ventricular tachycardia successfully treated with transaortic and transventricular septal myectomy: A case report



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Introduction

In this report, we describe a case of an 18-year-old woman with massive septal hypertrophic cardiomyopathy (HCM) without left ventricular outflow tract (LVOT) obstruction that was discovered after resuscitation from a witnessed cardiac arrest. HCM is the most common genetic cardiomyopathy, with an incidence of 0.2%.¹ It is associated with a 1% annual rate of sudden cardiac death (SCD).² Septal myectomy, typically reserved for treatment of LVOT, has not previously been described in the literature as a means to debulk the substrate for recurrent ventricular arrhythmias.

Case report

The patient is an 18-year-old woman with no significant prior medical history. While dancing, she experienced a witnessed collapse and was found pulseless. Immediate cardiopulmonary resuscitation was started, and spontaneous circulation returned after 2 minutes. A defibrillator was not readily available, and therefore the presenting cardiac rhythm was unknown. The patient was subsequently transferred to a tertiary care hospital, where she denied experiencing any prodrome of presyncope, palpitations, or chest pain prior to the cardiac arrest. She did reveal a family

history of sudden death in a maternal aunt and a heart transplant in a maternal uncle at age 19, but no further details were available.

On admission, she was alert and oriented and in no acute distress. Physical exam findings were unremarkable. Basic complete blood count and electrolyte studies were within normal limits. An initial electrocardiogram demonstrated left ventricular hypertrophy with ST depression and T-wave inversion in the anterolateral leads (Figure 1). A transthoracic echocardiogram demonstrated marked asymmetrical ventriculoseptal hypertrophy of 29 mm, consistent with the reverse-curve subtype of HCM (Figure 2A). There was no evidence of systolic motion of the anterior mitral valve or evidence of an outflow tract obstruction by continuous wave Doppler echocardiogram (Figure 2B). The patient's familial history of sudden cardiac death (SCD) predisposed her to an increased risk of recurrent malignant ventricular arrhythmias,³ and therefore a single-chamber implantable cardioverter-defibrillator (ICD) was implanted prior to discharge and metoprolol tartrate 25 mg twice daily was started. Of note, the metoprolol could not be further titrated because of the development of symptomatic bradycardia.

Unfortunately, the patient returned 5 months later following 4 events of ventricular fibrillation with successful ICD defibrillation occurring during moderate exertion. A treadmill echocardiographic (ECG) stress test precipitated syncope due to polymorphic ventricular tachycardia (Figure 4). She ultimately received cardiopulmonary resuscitation with restoration of sinus rhythm after 1 ICD defibrillation therapy. The patient denied experiencing any angina or presyncope prior to this event. A subsequent electrophysiological study did not demonstrate an inducible monomorphic ventricular tachycardia or appreciable trigger for polymorphic ventricular tachycardia or ventricular fibrillation. She was started on sotalol 120 mg twice daily, and because of bradycardia, the metoprolol was discontinued. She subsequently underwent a left-sided video-assisted thoracoscopic surgery (VATS) sympathectomy. On her recovery from surgery, a repeat

KEYWORDS Hypertrophic cardiomyopathy; Ventricular arrhythmia; Septal myectomy; Sudden cardiac death; Myocardial bridging

ABBREVIATIONS HCM = hypertrophic cardiomyopathy; ICD = implantable cardioverter-defibrillator; LVOT = left ventricular outflow tract; SCD = sudden cardiac death; TTE = transthoracic echocardiogram; VATS = video-assisted thoracoscopic surgery; VT = ventricular tachycardia (Heart Rhythm Case Reports 2015;1:300–304)

Conflicts of interest: The authors declare no potential conflicts of interest with respect to the research, authorship, or publication of this article. Funding: The authors received no financial support for the research, authorship, or publication of this article. **Address reprint requests and correspondence:** Dr Samuel J. Asirvatham, Division of Cardiovascular Diseases, Department of Medicine, Mayo Clinic College of Medicine Department of Pediatrics and Adolescent Medicine, Mayo Clinic, 200 First Street SW, Rochester, MN 55905. E-mail address: asirvatham.samuel@mayo.edu.

KEY TEACHING POINTS

- Septal myectomy may be an effective treatment for ischemia-induced ventricular arrhythmia in hypertrophic cardiomyopathy (HCM) patients.
- Coronary ischemia due to myocardial bridging is a potentially reversible and possibly underrecognized cause of ventricular arrhythmias in HCM patients.
- Septal myectomy is a mainstay of treatment for symptomatic left ventricular outflow tract obstruction refractory to medical therapy.

treadmill ECG stress test was performed. At 1 minute 50 seconds into stage 1 of a Bruce protocol, ST depression in the anterior, anterolateral and inferior leads with ST elevation in AVR (Figure 3) was noted. A few seconds later, polymorphic ventricular tachycardia developed (Figure 4) that degenerated into ventricular fibrillation that was terminated by ICD shock delivery.

Because of the recurrent polymorphic ventricular tachycardia preceded by the ST changes that were concerning for ischemia, a coronary angiogram was performed. Results showed no coronary atherosclerosis but revealed marked myocardial bridging of large first and second septal perforators (Figures 5A and 5B). It was suspected that the patient's polymorphic ventricular tachycardia (VT) was mediated by an exertional increase in myocardial contractility exacerbating myocardial bridging, with resultant ischemia and polymorphic VT. Accordingly, a septal myectomy was scheduled with the intention to debulk the interventricular septum in hopes to decrease ischemia and thereby remove the arrhythmogenic substrate.

The septal myectomy was successfully performed with an approach through the aorta and the left ventricular apex to

optimize exposure. Cardiac bypass was commenced and the aorta was cross-clamped. Then, an oblique aortotomy was performed, and the aortic cusps were retracted, allowing exposure to the subaortic septum. An extended septal myectomy was performed to the level of the papillary muscles with sparing of the septal perforators. After closure of the aortotomy, an apical ventriculotomy was performed, followed by a wide excision of the septum until it joined the area of the subaortic excision. The removed tissue demonstrated moderate to marked myocyte hypertrophy with mild to focally moderate interstitial fibrosis with no evidence of endocardial fibrosis. Lastly, the ventriculotomy was closed with 2 layers of running sutures. The cross-clamp was released, and temporary extracorporeal circulation was successfully discontinued. The total aortic cross-clamp duration was 27 minutes, and estimated blood loss was <250 cm³. The patient successfully recovered and was discharged from the hospital 6 days postoperatively with the recommendation to continue sotalol therapy. Electroanatomic mapping done 4 months after myectomy showed evidence of septal scar on myocardium. A year and a half post myectomy, the patient had no recurrent ventricular arrhythmias. She had a follow-up ECG cardiac stress test that did not provoke ventricular arrhythmias.

Discussion

HCM is the most common genetic cardiomyopathy, with an incidence of 0.2%.¹ Among the population of HCM patients, the annual incidence of SCD is approximately 1%.² Of several precipitating factors for SCD previously identified,³ the patient demonstrated 3: prior history of SCD, spontaneous ventricular tachycardia, and a presumed family history of SCD due to HCM.

ECG stress testing is reasonable to consider in HCM to determine response to therapy aimed at relieving outflow

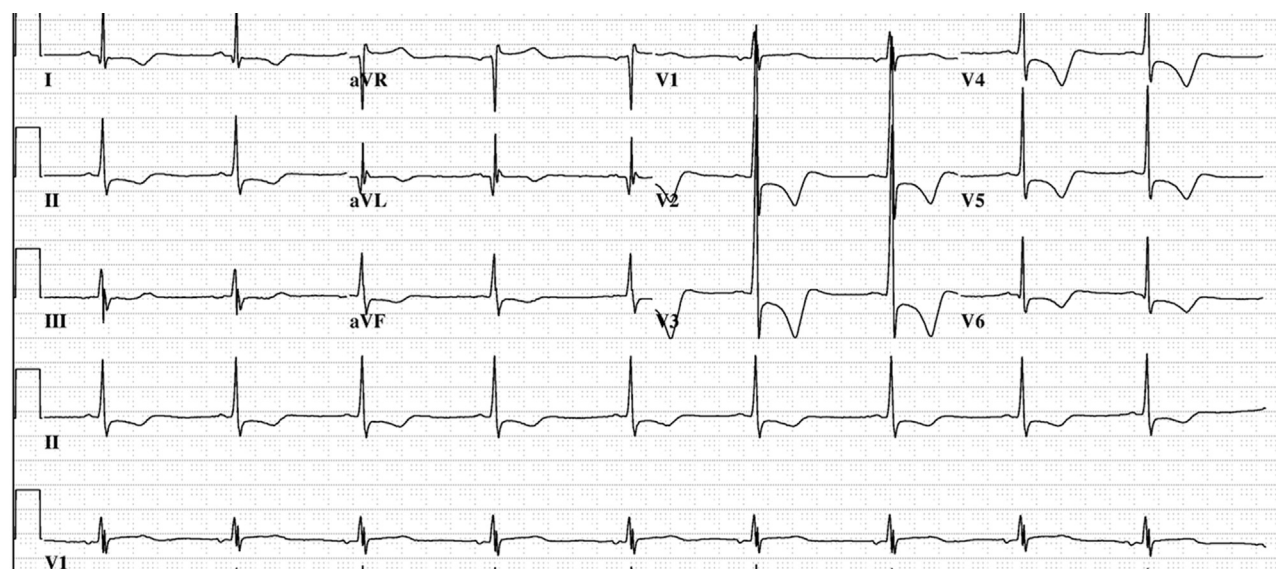


Figure 1 A 12-lead resting electrocardiogram demonstrating increased voltage consistent with left ventricular hypertrophy and anterolateral ST depression and T-wave inversion consistent with hypertrophic cardiomyopathy.

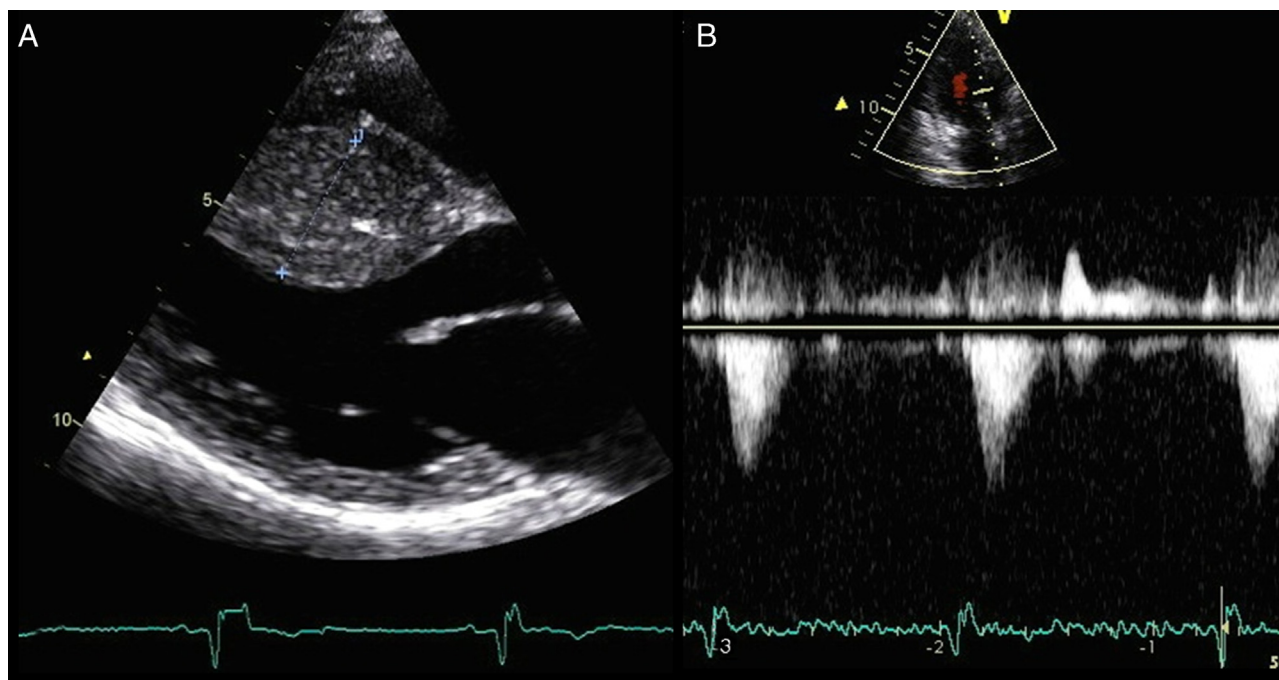


Figure 2 A: Parasternal long-axis transthoracic echocardiogram demonstrating marked septal hypertrophy (29 mm) and reverse-curve morphology. B: Continuous wave Doppler echocardiogram through the left ventricular outflow tract in the apical long-axis view without evidence of obstruction (highest recorded gradient was 0.8 m/s).

tract obstruction, systolic anterior motion of the mitral valve, and mitral regurgitation (Class IIa).³ In the current case, repeat ECG treadmill testing was performed to determine if either pharmacotherapy or VATS would be successful in precluding recurrent ventricular arrhythmias. Typically, the development of ventricular tachycardia in HCM stress testing is rare: in a prior series of

>3000 patients, only 1 (0.03%) had developed ventricular tachycardia.⁴ However, in the current case, exertional polymorphic ventricular tachycardia did occur, underscoring the importance to rule out structural cause for an ischemic trigger such as coronary atherosclerosis, myocardial bridging, or an anomalous coronary artery.



Figure 3 Echocardiogram stress test results revealing pronounced ST depressions in the anterior, anterolateral, and inferior leads with ST elevation in aVR. These findings developed 1 minute 50 seconds into a Bruce-protocol stress test and were concerning for ischemia.

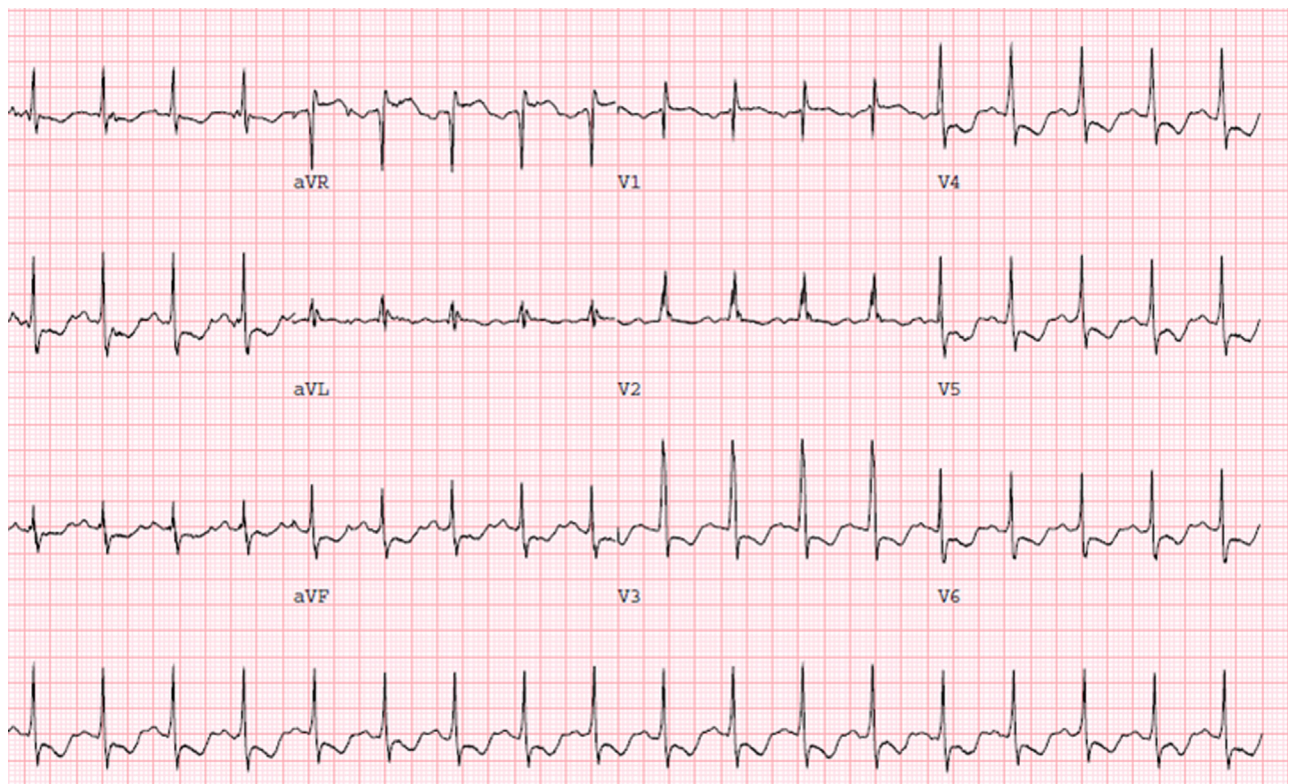


Figure 4 Echocardiogram stress test results revealing polymorphic ventricular tachycardia that developed 4 minutes 6 seconds into a Bruce-protocol stress test (2 minutes 16 seconds after the ST changes described in [Figure 3](#) occurred).

Pharmacotherapies including beta blockers and antiarrhythmic agents should not be used in lieu of ICD when ICD is indicated. In the current case, sotalol therapy was started in an attempt to preclude recurrent ventricular arrhythmias that were inciting ICD tachyarrhythmia therapies. A small, double-blind, crossover study has demonstrated the effectiveness of sotalol in suppression of ventricular arrhythmias

in patients with HCM.⁵ In the study, 54 percent of patients experiencing ventricular arrhythmias on placebo had resolution of these arrhythmias when sotalol therapy was started ($P < .05$). Moreover, this same study population improved the ECG stress test duration by 1 minute ($P < .01$). In the current case, another option to be considered would be beta-blockade titration as a means to reduce myocardial bridging.

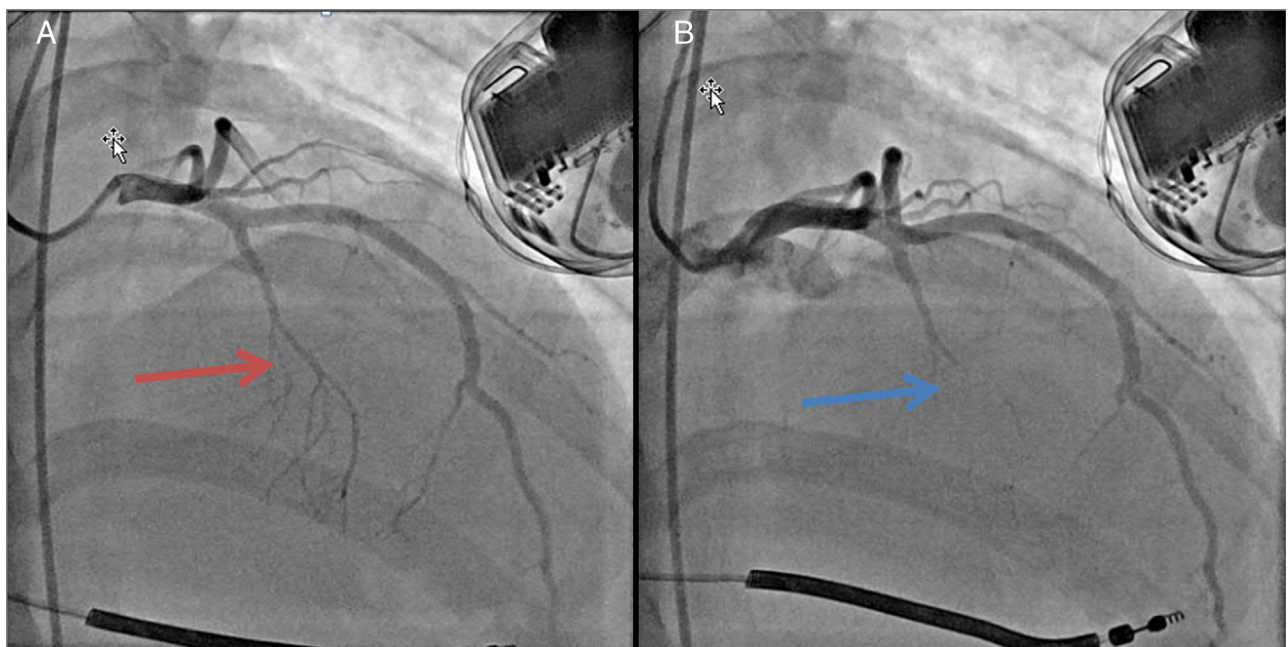


Figure 5 A: Coronary angiogram demonstrating a large septal perforator (red arrow). B: During systole, marked myocardial bridging is noted (blue arrow).

However, the patient developed bradycardia with low doses of metoprolol therapy, inhibiting further titration.

A left VATS sympathectomy decreases noradrenergic stimulation of the left ventricle by removing portions of the left stellate and thoracic ganglia. Prior experiences in the HCM population have demonstrated variable response to sympathectomy, with some patients having complete remission and others having persistent arrhythmias.⁶ Unfortunately, in the current case, left-sided sympathectomy did not prove successful. Consideration of a bilateral sympathectomy would have subjected the patient to the risks of intervention, including the development of Horner syndrome as well as a pneumothorax, without any evidence of benefit.

Electrophysiological ablation has been demonstrated to successfully treat recurrent monomorphic ventricular arrhythmias in the HCM population⁷ and can be considered to eliminate ventricular ectopy known to trigger polymorphic ventricular tachycardia or ventricular fibrillation.⁸ However, in the current case, the patient had no inducible monomorphic ventricular arrhythmias or appreciable triggers for polymorphic ventricular tachycardia or ventricular fibrillation during electrophysiological testing, suggesting that the unroofing of the myocardial bridges and debulking ischemic substrate by myectomy were the main mechanisms for eliminating the arrhythmia. Cardiac transplant for incessant ventricular arrhythmias in the HCM population may also be considered.⁹

However, given the patient's young age at presentation, an innovative approach was chosen in an attempt to avoid the long-term morbidity associated with cardiac transplantation. Coronary stenting of the septal perforators to inhibit bridging was not attempted, given evidence of prior trials suggesting increased rates of postintervention in-stent stenosis.¹⁰ Compared with septal myectomy, alcohol septal ablation would be limited in its capability to debulk the myocardium. Moreover, septal ablation would induce ischemia and predispose the patient to further ventricular arrhythmias.¹¹ Therefore, septal myectomy was the chosen intervention. Although septal myectomy has proven to be a mainstay of treatment for symptomatic left ventricular outflow tract obstruction that is refractory to medical therapy,¹² its use to successfully debulk an arrhythmogenic substrate in non-obstructive HCM is novel.

Conclusion

HCM is a common cardiomyopathy and may be associated with life-threatening ventricular arrhythmias necessitating ICD implantation. Invasive procedures, such as septal ablation or septal myectomy, are typically reserved for treatment of symptomatic LVOT. Treatments for ventricular arrhythmias include ICD implantation for primary or secondary prevention,³ beta-blockade, antiarrhythmics, and

VATS. In refractory cases, cardiac transplantation may be considered.

The current case describes a rare phenomenon of exercise-induced ventricular arrhythmia in a patient with HCM. Moreover, prior to the development of polymorphic ventricular tachycardia, the echocardiogram demonstrated changes consistent with ischemia. Therefore, the presence of polymorphic ventricular tachycardia in patients with HCM may be a clue to a potentially reversible and possibly underrecognized cause for ventricular arrhythmias, such as coronary ischemia due to myocardial bridging. In the current case, a coronary angiogram was negative for atherosclerosis but did show marked bridging of large first and second septal perforators. Extended septal myectomy successfully precluded any recurrent arrhythmias. This case describes the novel use of ventricular myectomy to debulk ischemic substrate in HCM to prevent recurrent arrhythmias.

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