It is the time for a heart team approach for patients with hypertrophic cardiomyopathy

Niv Ad, MD, and Lawrence M. Wei, MD

From the Department of Cardiovascular and Thoracic Surgery, West Virginia University, Morgantown, WV.
Disclosures: Ad is a consultant for Medtronic, a member of the speaker's bureau for AtriCure, a proctor and a member of the speaker's bureau for LivaNova, on the advisory board for Nido Surgical, and co-owner of Left Atrial Appendage Occlusion, LLC. Wei has nothing to disclose with regard to commercial support.
Received for publication Aug 8, 2017; accepted for publication Aug 26, 2017; available ahead of print Sept 27, 2017.

Address for reprints: Niv Ad, MD, Department of Cardiovascular and Thoracic Surgery, West Virginia University,

1 Medical Center Dr, Morgantown, WV 26506 (E-mail: nivadmd14@gmail.com).

J Thorac Cardiovasc Surg 2017;154:e105-6

0022-5223/\$36.00

Copyright © 2017 by The American Association for Thoracic Surgery

<http://dx.doi.org/10.1016/j.jtcvs.2017.08.083>

In their case series in this issue of the *Journal*, Nguyen and Schaff¹ from the department of cardiovascular surgery at the Mayo Clinic present the results of a surgical approach to treat patients with midventricular obstruction and hypertrophic cardiomyopathy (HCM). All 3 patients were seen with significant history of electrical storm and sustained ventricular tachycardia. One of the patients also had significant thromboembolic events occur.

HCM is a heterogeneous disease that is classically transmitted in an autosomal dominant fashion. In HCM, the association between a genetic sarcomeric disorder and myocyte disarray and scarring is well linked to sudden cardiac death and malignant ventricular arrhythmias. The myocardial changes found in patients with HCM are myocardial fibrosis, microvascular ischemia, and cellular disarray. Interestingly, the histologic changes of myocardial fibrosis (scarring and disruption of the normal cellular architecture) can be found throughout the myocardium, but they are most commonly seen in the thickest area of hypertrophied cardiac muscle.²⁻⁴ It is documented that silent, low-grade myocardial ischemia and altered myocardial blood flow lead to the conditions needed for scar formation and electrical remodeling arrhythmogenesis.⁵ These changes are directly linked to reentrant ventricular arrhythmias in patients with HCM.

Electrical remodeling of the myocardium is defined as a persistent change in the electrophysiologic properties of myocardium in response to a change in rate or activation sequence.⁶ When it comes to the atrial myocardium and atrial fibrillation, electrical remodeling has been shown to have significant implications for the maintenance and stability of atrial fibrillation.⁷ Multiple studies have demonstrated that the fibrillatory activity is associated with changes in ion channels, shortened atrial refractoriness, and more established and persistent atrial fibrillation.^{7,8} The ventricular myocardium is also subject to electrical remodeling⁶; however, the mechanisms of this electrical remodeling are less understood. The clinical

Niv Ad, MD, and Lawrence M. Wei, MD

Central Message

Both the surgical and the cardiology communities should be aware of a surgical approach to hypertrophic cardiomyopathy and the importance of its consideration by the heart team when indicated.

See Article page e101.

implications of the ventricular electrical remodeling are significant and well associated with sustained ventricular tachycardia, heart failure, and mortality.

Although relatively uncommon, sudden death and syncope are well-documented complications of hypertrophic cardiomyopathy,⁹ and although not fully understood, they may be related to lethal ventricular arrhythmias, ischemia, and the degree and severity of the left ventricular outflow obstruction. Polymorphic ventricular tachycardia and ventricular fibrillation are the most common documented arrhythmias during electrophysiologic studies.¹⁰ The availability of implantable cardioverter-defibrillator (ICD) therapy significantly improved the treatment approach to such patients, leading to a clear reduction in sudden cardiac death, with reported rates of 7% per year in termination of ventricular tachycardia or ventricular fibrillation with a very high success rate.¹¹ It is well established that ICD therapy is efficacious in preventing sudden cardiac death; however, the challenge remains in patient selection for such therapy. Catheter ablation in conjunction with an ICD is usually offered and performed in patients with significant numbers of ventricular arrhythmias.⁹

Midventricular obstruction is an uncommon presentation of HCM; however, there is an increased recognition of the importance of relatively unusual thin-walled, scarred left ventricular apical aneurysms associated with this variant

of HCM.⁹ In its severe form, it may lead to an apical aneurysm and the development of 2 distinct left ventricular chambers with flow abnormalities. Patients with HCM, midventricular obstruction, and left ventricular apical aneurysms represent a high-risk subgroup for severe complications, including ventricular arrhythmias and sudden death, thromboembolism, and end-stage heart failure.⁹ The sudden death events among such patients can be as high as 5% per year, and these patients therefore represent a unique high-risk group among those with HCM. Modern imaging, including contrast echocardiography and magnetic resonance imaging, may assist in identifying the higher risk group for life-threatening complications; however, the small number of patients with such a phenomenon makes it difficult to establish and validate a risk stratification model. The clinical approach to complications associated with HCM and apical aneurysm is not different from that for other forms of HCM and has shown to be very effective. This includes primary prevention, ICD therapy, radiofrequency ablation for recurrent ventricular tachycardia, and prophylactic anticoagulation for stroke prevention. Catheter ablation can be very effective in such patients, because in many cases, the mechanism for the ventricular arrhythmia in this specific subgroup of patients is often associated with a single macro reentry circuit around the neck of the aneurysm.

It is clear that there needs to be more discussion and refinement of the clinical approach for the unique and challenging subgroup of patients represented in this case series. These patients did not respond to the common clinical approaches and went to surgery with sustained ventricular arrhythmias and other complications. The surgical community is not well familiar with the surgical approaches appropriate for this subgroup of patients, mainly because of

the condition's rarity. It is important to recognize, however, that a well-skilled surgical team can achieve outstanding outcomes. Both the surgical and the cardiology communities should be aware of such a surgical approach and the importance of its consideration when indicated as part of the heart team approach for HCM. This case series surely demonstrates that appropriate indications and surgical execution result in excellent long-term outcomes.

References

1. Nguyen A, Schaff HV. Electrical storms in patients with apical aneurysms and hypertrophic cardiomyopathy with midventricular obstruction: a case series. *J Thorac Cardiovasc Surg.* 2017;154:e101-3.
2. Ismail TF, Jabbour A, Gulati A, Mallorie A, Raza S, Cowling TE, et al. Role of late gadolinium enhancement cardiovascular magnetic resonance in the risk stratification of hypertrophic cardiomyopathy. *Heart.* 2014;100:1851-8.
3. Varnava AM, Elliott PM, Mahon N, Davies MJ, McKenna WJ. Relation between myocyte disarray and outcome in hypertrophic cardiomyopathy. *Am J Cardiol.* 2001;88:275-9.
4. Chan RH, Maron BJ, Olivetto I, Pencina MJ, Assenza GE, Haas T, et al. Prognostic value of quantitative contrast-enhanced cardiovascular magnetic resonance for the evaluation of sudden death risk in patients with hypertrophic cardiomyopathy. *Circulation.* 2014;130:484-95.
5. Basso C, Thiene G, Corrado D, Buja G, Melacini P, Nava A. Hypertrophic cardiomyopathy and sudden death in the young: pathologic evidence of myocardial ischemia. *Hum Pathol.* 2000;31:988-98.
6. Libbus I, Rosenbaum DS. Remodeling of cardiac repolarization: mechanisms and implications of memory. *Card Electrophysiol Rev.* 2002;6:302-10.
7. Wijffels MC, Kirchhof CJ, Dorland R, Allessie MA. Atrial fibrillation begets atrial fibrillation: a study in awake chronically instrumented goats. *Circulation.* 1995;92:1954-68.
8. Yue L, Feng J, Gaspo R, Li GR, Wang Z, Nattel S. Ionic remodeling underlying action potential changes in a canine model of atrial fibrillation. *Circ Res.* 1997;81:512-25.
9. Rowin EJ, Maron BJ, Haas TS, Garberich RF, Wang W, Link MS, et al. Hypertrophic cardiomyopathy with left ventricular apical aneurysm implications for risk stratification and management. *J Am Coll Cardiol.* 2017;69:761-73.
10. Alfonso F, Frenneaux MP, McKenna WJ. Clinical sustained uniform ventricular tachycardia in hypertrophic cardiomyopathy: association with left ventricular apical aneurysm. *Br Heart J.* 1989;61:178-81.
11. Trivedi A, Knight BP. ICD therapy for primary prevention in hypertrophic cardiomyopathy. *Arrhythm Electrophysiol Rev.* 2016;5:188-96.