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

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In their case series in this issue of the Journal, Nguyen and Schaff 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. 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. 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. 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. 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. The ventricular myocardium is also subject to electrical remodeling; however, the mechanisms of this electrical remodeling are less understood. The clinical 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, 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. 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. 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.

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.
In its severe form, it may lead to an apical aneurysm and the development of 2 distinct left ventricular chambers with flow abnormalities. Patients with HCM, midventricular obstruction, and left ventricular apical aneu-
yrsms represent a high-risk subgroup for severe complica-
tions, 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 reso-
nance 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, radiofre-
nquency 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 chal-
lenging subgroup of patients represented in this case series. These patients did not respond to the common clinical ap-
proaches and went to surgery with sustained ventricular arr-
hythmias and other complications. The surgical commu-
ity is not well familiar with the surgical approaches appro-
riate 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 commu-
nities 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.

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