Obstruction in Hypertrophic Cardiomyopathy
How Often Does It Occur? Should It be Treated? If So, How?

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Hypertrophic cardiomyopathy (HCM) is the most common monogenic cardiac disorder and has been estimated to occur in 1 of every 500 people in the general population, amounting to a total of ~600,000 persons in the United States. Its pathophysiology and optimal management have been the subject of conjecture and debate for more than a century. The issues surrounding left ventricular outflow tract (LVOT) obstruction in HCM have evoked the most discussion.

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In 1907, a German pathologist, A. Schminke, described 2 hearts from women in their mid-50s. Decades before the development of left heart catheterization, and before any pressure gradients had ever been measured in humans, he wrote the following: “Diffuse muscular hypertrophy of the left ventricular outflow tract causes an obstruction. The left ventricle has to work harder to overcome the obstruction. So, the primary hypertrophy will be accompanied by a secondary hypertrophy, causing an incremental (further) narrowing of the outflow tract.” Thus, Schminke presciently understood that the obstruction can only be explained by muscular hypertrophy of the left ventricular outflow tract.1-3

As open-heart surgery exploded in the early 1960s, patients with this condition (the name HCM had not yet been agreed on) were encountered with increasing frequency by cardiologists and cardiac surgeons around the world; the British surgeon, Sir Russell Brock provided especially useful insights.4 Indeed, HCM soon became the poster child for the hemodynamic era. Although, by definition, all patients had forms of left ventricular hypertrophy similar to those in the first 2 groups, but did they not exhibit LVOT obstruction even with provocation.5

Because LVOT obstruction could be provoked or intensified by β adrenergic stimulation,6 it was logical to try to prevent it with β adrenergic blockers, which had just been developed and which proved helpful to many patients with HCM, perhaps less so to patients with severe LVOT obstruction in the basal state.7 Morrow at the National Institutes of Health8 and Kirklin and Ellis at the Mayo Clinic9 turned their attention to the development of a corrective operation, surgical septal myectomy (SSM). However, in those early years, the operation was found to be technically challenging, the risk was substantial, and it was carried out in only a small number of centers. Some observers even doubted that obstruction ever occurred in HCM and that the pressure differences between the left ventricle and the aorta, on which we based our recommendations about surgery, were artifacts.10,11

Now, to fast-forward to 2006, when the Marons and their colleagues12 showed by echocardiography that LVOT gradients at rest or during exercise occur in 70% of patients with HCM, are frequently associated with symptoms and adverse clinical outcomes,13 and concluded that when these obstructions are not responsive to pharmacological therapy, they require mechanical relief. Two approaches to accomplish this are available today; the first is SSM, sometimes referred to as the Morrow procedure, which has gradually become both more extensive and safer over the years. In 1995, alcohol septal ablation (ASA) was introduced,14 and because it relieved obstruction without requiring open-heart surgery, it quickly became the more frequently used procedure. However, SSM has continued to be carried out, mostly at specialized centers, and it is still considered to be the gold standard.15 The question most frequently asked today is which of these procedures is preferable and for whom?

Three major comparisons are available. Argawal et al16 performed a comparison of the results in 8 institutions in which both procedures were used; 326 patients received SSM and 380 ASA. Leonardi et al17 compared the results on 1887 patients who received SSM with those on 2153 patients who received ASA, usually in different hospitals. Several studies found their way into both comparisons. Both analyses concluded that the mortality rates were similarly low with both procedures. However, Agarwal et al16 pointed out that ASA increased the need for implantation of a permanent pacemaker and left patients with low, but significantly higher, LVOT pressure gradients than did SSM.

In this issue of Circulation, the excellent study by Sorajja et al18 from the Mayo Clinic, Rochester provides the largest
References


Disclosures

None.
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