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Diversity of Mitral Valve Abnormalities in Obstructive Hypertrophic Cardiomyopathy

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Abstract Hypertrophic cardiomyopathy (HCM) is a complex and common genetic disease. The left ventricular outflow tract obstruction is an important determinant of symptoms and outcomes. Its pathophysiology is determined by the complex interaction of the mitral valve, papillary muscles, chordae tendinea, and interventricular septum. The knowledge and importance of the mitral valve apparatus in the physiology of the HCM are expanding with the new imaging modalities, in particular, cardiac magnetic resonance. Several abnormalities of the mitral apparatus have been described in patients with HCM including abnormal papillary muscles, leaflets' lengths, and mitral regurgitation. Ignoring these variables can lead to unnecessary or incomplete surgical treatments and worse outcomes. This review discusses the role of the mitral apparatus in HCM with a focus on a multimodality imaging approach and the clinical importance of each abnormality. (Prog Cardiovasc Dis 2012;54:517-522)

Keywords: Hypertrophic cardiomyopathy; Mitral valve apparatus; Mitral regurgitation; Papillary muscles; Mitral leaflets

Hypertrophic cardiomyopathy (HCM) is a complex and common genetic disease with an autosomal dominant pattern of inherence, variable phenotypic expression, and diverse clinical outcomes. The prevalence of phenotypically expressed HCM is approximately 1:500 of adult individuals,[1] with identifiable mutation seen in 63% of patients with HCM.[2] The phenotypic expression of the disease is wide and characterized by predominantly asymmetric hypertrophy of the left ventricle that, although commonly affects the anterior septum, can also involve the apex, left ventricular free wall, and right ventricle and be symmetric as well.[3-5]

After 50 years since the first modern description of HCM, the complex pathophysiology of the disease is yet to be completely understood and continues to be a source of controversies in the medical literature.[6-8] This pathophysiology includes a complex interaction of the mitral valve (MV), the papillary muscles (PMs), chordae tendineae, and the ventricular septum that causes systolic anterior motion (SAM) leading to dynamic left ventricular outflow tract (LVOT) obstruction (LVOTO) and mitral regurgitation (MR). This dynamic LVOTO producing an intraventricular pressure gradient was recognized to be one of the most important features of the HCM since the first reports[7,9] and later recognized as one of the important determinants of outcomes.[10,11]

The increased flow velocity with asymmetric septal hypertrophy and narrowing of the outflow tract causing SAM (Venturi theory) was hypothesized as the mechanism that the obstruction occurs,[12,13] but LVOTO is also found in patients independent of asymmetric septum hyperthrophy.[14,15] The Venturi phenomenon also failed to explain the fact that SAM starts at low outflow velocities.[16] Because of this and other observations, investigators have presented evidence that flow drag, the pushing force of

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Abbreviations and Acronyms

AML = anterior mitral leaflet

CMR = cardiac magnetic resonance

HCM = hypertrophic cardiomyopathy

LVOT = left ventricular outflow tract

LVOTO = left ventricular outflow tract obstruction

MR = mitral regurgitation

MV = mitral valve

PM = papillary muscle

SAM = systolic anterior motion

TEE = transesophageal echocardiography

flow, is the dominant hydrodynamic force that causes SAM. The knowledge of the influence of the mitral apparatus in the pathophysiology of HCM has therapeutic implications because septal myectomy alone without intervention to the mitral apparatus could lead to worse outcomes.[17-19]

Within this context, it is important to understand the diversity of MV abnormalities and its components in patients with HCM and the potential clinical implications for diagnosis and management.

Papillary muscle

In the 1960s, Simon et al[20] performed angiographic studies in patients with HCM and hypothesized that hypertrophy and shift in the axis of PMs were contributors to the development of the LVOTO. Since then, several studies further characterized the anatomical abnormalities leading to an understanding of their clinical relevance. The currently described abnormalities are PM hypertrophy, PM insertion directly in the anterior mitral leaflet (AML), PM fusion to ventricular septum, PM fusion to left ventricular free wall, anterior apically displaced PM, double bifid PM, and accessory PM.[4,15,18,21,22]

Klues et al[23] studied the MVs in 10 patients with hypertrophied PMs inserting directly into the AML. The PM had an anterior displacement and, during systole midcavity obstruction, was produced by direct contact of the PM and midseptum, 6 of them without any SAM. Eight patients underwent valve replacement and had good outcomes in contrast to 2 patients who underwent myectomy alone with poor outcomes (1 postoperative death and 1 had persistent high gradient and symptoms after surgery). The initial echocardiogram failed to demonstrate the abnormality in 9 of 10 patients.

Advances in cardiac imaging technologies have allowed an understanding of novel insights into the complex features leading to dynamic LVOTO. Using cardiac magnetic resonance (CMR) imaging, Kwon et al[15] found that anteroapical displacement of anterolateral PM and a double bifid PM were associated with a higher peak resting LVOT gradient compared with those without these characteristics. Importantly, this association was not affected by the degree of septal thickness. Figs 1 and 2 illustrate a case in which anteroapical displacement of anterolateral PM and double bifid PM cause dynamic LVOTO. Clip 1 is a clinical example of a 44-year-old man with family history of HCM presenting with exertional dyspnea. Note that CMR can show several abnormalities of which the anterior tenting of the AML by the anomalous insertion of the PM favors significant dynamic LVOTO at rest.

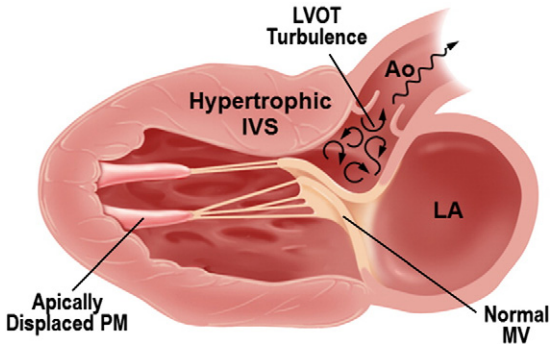

Fig 1. Schematic representation of the left ventricular tract obstruction in HCM with apically displace PMs. *Abbreviations:* Ao, aorta; LA, left atrium; IVS, intraventricular septum.

A real-time 3-dimensional echocardiographic study also found that compared with controls, patients with HCM have medial and anterior displacement of both PMs resulting in a shorter inter PM distance, an independent determinant of LVOT obstruction.[22] In vitro studies supported the theory that displacing the PM anteriorly as observed in patients with HCM creates SAM in absence of septal hypertrophy.[24,25] Anterior displacement of the PM was also surgically induced in dogs and produced SAM and outflow gradient.[26] Displacement of the MV apparatus places the MV into the flow stream, directly subjecting it to the pushing forces of flow.

Studies in patients undergoing septal myectomy showed that these abnormalities are frequently not identified by transthoracic echocardiography and often are only identified during direct inspection at the time of operation.[18,27]

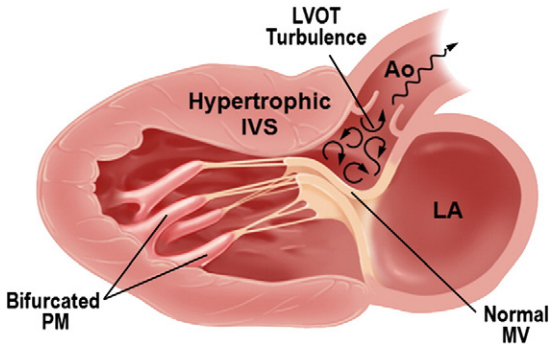

Fig 2. Schematic representation of the left ventricular obstruction in HCM with bifurcated PMs.