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Diagnostic challenges for symptomatic obstructive HCM

Hello, I'm Pablo García-Pavia from Hospital Universitario Puerta de Hierro and Centro Nacional de Investigaciones Cardiovasculares, CNIC, both in Madrid and Spain. It's my pleasure to talk today about diagnostic challenge with symptomatic obstructive hypertrophic cardiomyopathy. Here are my disclosures.

Hypertrophic cardiomyopathy is a very heterogeneous disease. The underlying pathophysiology of this disease is common, no matter what is the type of presentation of patients. Patients show hypertrophy, increased fibrosis, hypercontractility, impaired relaxation, and altered myocardial energetics. As you probably know, in a majority of patients with hypertrophic cardiomyopathy, there is an underlying genetic mutation in one of the sarcomeric genes.

The clinical course of hypertrophic cardiomyopathy is pretty heterogeneous as well. There are patients who suffer from sudden cardiac death, from heart failure, or develop atrial fibrillation and embolic events, but there is a wide majority of the patients who do pretty well and remain stable over the disease course.

HCM is characterized by left ventricular hypertrophy in the absence of another cardiac systemic or metabolic disease capable of producing the magnitude for hypertrophy. In adults, this represents 15 millimeters, and in children, this means adjusted z-score more than two standard deviations above the mean. This is why it's very important to accurately measure left ventricular hypertrophy in these patients. For doing this, we should take into account that we should measure the degree of hypertrophy in the short axis, echocardiographic view. As you can see here, in this patient, it's very important to do the measurement this way, because otherwise, the degree of left ventricular hypertrophy will have not been correctly measured. Assessing what is the magnitude of left ventricular hypertrophy is certainly important for sudden cardiac death prevention as this parameter is included in both the American and European guidelines for assessing sudden cardiac death prevention.

When the echocardiographic views are not good enough, we have the possibility of using cardiac MRI which allows us to have a better definition of left ventricular walls. Also, MRI allows us to identify phenocopies, which play a fantastic, important role in the diagnostic part of hypertrophic apathy.

The pathophysiology or the pathology of left ventricular hypertrophy goes beyond left ventricular hypertrophy. We know nowadays that hypertrophic cardiomyopathy also affects mitral valve, coronary arteries and more structures in the heart. In this sense, we now know that the papillary muscles exhibit abnormalities, and they have anterior insertions in the mitral valve apparatus, and they are located in different positions across the left ventricle. Here, we have also a study highlighting how these patients exhibit mitral valve abnormalities. They have longer mitral valves than patients who do not have hypertrophic cardiomyopathy. This finding represents early alteration in hypertrophic cardiomyopathy. It has been shown that even patients who have subclinical hypertrophic cardiomyopathy and who are carriers of sarcomeric mutations exhibit these alterations in the mitral valves. The combination of hypertrophy plus long mitral valves and altered papillary muscles contribute all them to the physiology of left ventricular outflow tract obstruction, as you can see here in this beautiful example.

Assessing the presence of obstruction is certainly important because a majority of the patients with hypertrophic cardiomyopathy who develop progressive heart failure suffer from obstructive hypertrophic cardiomyopathy. Obstruction is associated with worse prognosis,

as you can see here in the slide. Both death and advanced heart failure are more common in patients with hypertrophic cardiomyopathy who exhibit obstruction.

An obstruction is seen in one-third of patients with hypertrophic cardiomyopathy at rest. It's very important to investigate this finding in patients who suffer from shortness of breath because when we do provocation maneuvers, we see that 70% of patients with hypertrophy cardiomyopathy at referral centers show obstructive physiology. For this, we should perform Valsalva maneuvers or exercise testing. As you can see here in this example from our review of our group published in 2018, where the patient did not have any degree of obstruction at rest, but with exercise, you can see how left ventricle outflow tract obstruction appear. To provoke left ventricle outflow tract obstruction, we have a variety of maneuvers. Obviously, we can do Valsalva at rest, as you can see here in the slide. There is also the possibility to perform exercise and assess the degree of obstruction at the peak exercise and also post-exercise. Moreover, these patients frequently complain about symptoms postprandial. It's possible also to assess the presence of obstruction after a heavy meal, as was done here in this beautiful study from Italy. As you can see here in these patients, only one-third of them showed obstruction in fasting exercise, but postprandial even at the rest, three-quarters of the patients had obstruction.

It's important to highlight that when assessing obstruction, we should differentiate between the flow through the arctic valve and the left ventricular outflow tract, which is this dark shape in red, and not the mitral insufficiency that frequently accompanies these patients when they have obstruction. Here, you can see it in blue.

In conclusion, HCM is a heterogeneous disease with a heterogeneous clinical course. LVH characterized hypertrophic cardiomyopathy, and it should be correctly assessed. HCM pathology expands beyond left ventricular hypertrophy and involves the papillary muscles and mitral valve apparatus. These structures play a pivotal role in left ventricular outflow tract obstruction. This finding is present in up to two-thirds of patients with hypertrophic cardiomyopathy. An appropriate evaluation of obstruction with provocation techniques is required in symptomatic patients.

Thank you very much for your attention.