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Echocardiographic workup for HCM – making the diagnosis and evaluation of cardiac function

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Dr. Edvardsen:
Hello, everyone. My name is Thor Edvardsen and I'm happy to present the echocardiographic workup for hypertrophic cardiomyopathy. Here are my disclosures.

Echocardiography gives you a quick very good overview of the typical echocardiographic findings in severe hypertrophic cardiomyopathy. Here you can see the most typical features in hypertrophic cardiomyopathy. Like upper left you have the systolic anterior motion, SAM of the mitral valve. It's caused by the elongation of the mitral leaflets. Also, the very thick myocardium you can see there. Due to SAM, you have often mitral regurgitation, but you should also be aware that you have mitral regurgitation without SAM. Also in HCM in 10% to 20% of all the cases. LVOT obstruction is found in 60% of all patients with hypertrophic cardiomyopathy. The Doppler, you can see down left, the typical dagger-shaped envelope pattern in the Doppler pattern. Also, this is to discover with the colors, the LVOT obstruction, as you can see down right.

The diagnostic criteria, briefly. In adults, the thickest part of the wall should be equal or bigger than 15 millimeter in any myocardial segment, and it should not be explained by loading conditions. In relatives more or equal to 13 millimeters of maximum wall thickness. In children, the pediatricians, they use a predicted mean or a z-score. More than two standard deviation from the normal in their age groups.

Remember, you should measure the thickest part of the wall anywhere where you can find it. If it's basal or the mid or the apical level as shown in this example here to the right. That was the biggest, thickest part in this patient.

Here's the typical example sent to our hospital. Man, 37 years old, he had dyspnea, and he was in NYHA class II. Here is his echo. You can see the very thick walls and very good systolic performance. Here's from the apical four-chamber view, and it's very easy to discover the obstruction by a color Doppler as you can see to the right. His obstruction was severe. He had higher velocity than five meter per second, which gives gradient bigger than 100 millimeter of mercury. He was genotype positive, and we calculated his risk score, five-years risk score. It was 8.3% Then he received an ICD, of course. We gave him Verapamil but also beta block here, but it had only minor effect on his symptoms. He was referred for possible septal reduction therapy in May '24. In our hospital, we do both myectomies, the surgeons, and also alcohol septal ablation. We do more than 50 alcohol septal ablations every year, so he was referred for that type of therapy. Here is on the operating table before the ablation. You can see the high obstruction there. Here is when he received contrast in his septal branch. At the basal part of the septum, you can see contrast lightning up, so we know it's a safe place to ablate, and it was ablated. To the left, you can see the Doppler signal before ablation, to the right, after. There is not a huge difference here, but that was taken immediately after the ablation. Usually, we wait three months to conclude the results. He will come back in

December. Well, this month, actually, for a control and status after ASA. He had a maximum CK-MB of 118, so he had an infarct. We hoped for him that it was a successful treatment.

In all patients, we also do CMR at the first consultation. Here's another man, 35 years old, three cases of sudden cardiac death in his family. You can see the contrast uptake. Relatively large area at the right there, the white areas in the left ventricle, so he has a huge area of contrast. We also do strain imaging on all patients. Here is a GLS of the same patient. As you can see, the white areas, they correspond to the uptake of the contrast from the CMR. If you report the systolic function in these patients, his ejection fraction is 65. You can imagine and think that the systolic performance is normal, but it's not. As you can see, the global longitudinal strain is only -9% so he has severe systolic dysfunction. That goes for many of these patients.

Here's another one, a woman, 72 years old. She had palpitations and dyspnea, and NYHA class III. She had midventricular obstruction, as you can see on the image at the right, so we did Valsalva to see if the obstruction had a high gradient. The Doppler pattern, be careful. This is the mitral regurgitation. Inside the mitral regurgitation signal, you can also see the obstruction. It was not very high, two meter per second. Then we did squats. The maximum velocity we got was 3.7 meter per second, corresponds to 55 millimeter of mercury, so she had, also, a significant obstruction. She was also scheduled for exercise echocardiography, and she could do maximum 80 Watts. The obstruction we measured there was only two meter per second. Actually, the highest velocity was measured during 60 Watts. If you already have a significant obstruction from squats, for Valsalva, you don't necessarily have to do exercise echocardiography.

Also, remember that 40% of all our patients with HCM, they do not carry a non-pathogenic mutation. At the left, you see a typical genotype-negative HCM patient with the asymptomatic septal hypertrophy. At the right, you see a genotype positive phenotype, negative HCM patients. Remember, genetic tests, they have their limitations.

Strain imaging in HCM is very important. We do it in every patient. Here is one with a mild phenotype. Maximum wall thickness is 13 millimeter and signs of systolic dysfunction by strain. GLS was -16% while ejection fraction is 60% They also relatively often have a heterogenic or dispersed contraction pattern. It means that this maximum systolic contraction is occurring at different time spots during systole and diastole. We can calculate that, and we call it mechanical dispersion. There is a link between abnormal strain and ventricular arrhythmias and fibrosis in all these patients. We have shown that the more heterogenic pattern you got from the strain curves the more risk of having malignant arrhythmias. If you add strain GLS to the risk stratification by the ESC, you will have more accurate prediction of malignant arrhythmias. If you add mechanical dispersion on top, it will even better. You will have even better accuracy to predict arrhythmias.

Mechanical dispersion is also useful and most patients with hypertrophic cardiomyopathy who have a dispersed systolic contraction pattern by strain. While another one at the left here, athlete, also with hypertrophy, has a much more homogenic contraction pattern than the patient at the right with the hypertrophic cardiomyopathy.

Echocardiography is essential for the management of HCM patients for diagnosis, for family screening, for therapeutic strategies, and also for risk stratification.

How to follow these patients. Well, we do echocardiography every year or every second year if they are stable, and we also do CMR at least once at the initial evaluation. It could be repeated according to changes in their clinical status.

To summarize my presentation, cardiac imaging plays a central role in cardiac phenotyping and disease monitoring in HCM patients. Echocardiography is the main imaging tool from the initial diagnostic to the follow-up. An exercise echocardiography can be challenging but should be done if you don't have a significant obstruction by Valsalva or squats. Ejection fraction is very often preserved in HCM patients although systolic function is severely depressed. I think you should add advanced techniques as GLS and mechanical dispersion. CMR, of course, allows you for tissue characterization and see how widespread the fibrosis is. Combining Echo and CMR facilitates risk stratification and particularly sudden cardiac death.

Thank you for your attention.

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