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Hypertrophic cardiomyopathy: What are the concerns?

Hello. In the next few minutes, I will discuss with you when one should be concerned about a condition called hypertrophic cardiomyopathy.

Hypertrophic cardiomyopathy is a condition characterized by a thick heart muscle, hypertrophied heart muscle. It's not just the thickness. It also changes within the heart muscle, such as development of scar tissue, fibrosis, lack of energy due to overconsumption of the energy by the muscle, and reduced oxygen supply to the cardiac cells because of microcirculation abnormalities. The thick heart muscle occupies space from the left ventricle usually and therefore, the capacity of the left ventricular pump is reduced. It can come in many different patterns. Sometimes, it is symmetrical. Most frequently, it is asymmetrical, but it's always concentric, meaning that it goes towards the center of the left ventricle. This reduced space within the left ventricle causes a number of functional problems. One of them is that the ventricle cannot handle the volume of blood in diastole, and together with the stiffness of the muscle, makes the filling time of the ventricle a struggle. Also, the systolic function of the heart is severely changed. There are areas where the heart is hypercontractile, meaning that there is very strong systolic function. There are areas which, due to fibrosis and the loss of the myocyte architecture, which we call disarray, are hypocontractile, so they have reduced performance during systole. There are also areas which overcompensate for the hypocontractile areas of the heart and therefore, pump much faster and stronger. On top of all of this, there is also obstruction in the outflow tract of the left ventricle, which means that when the heart function is expected to give the most to the body and the circulation, the flow of the blood is obstructed inside the heart.

All these are significant problems associated with hypertrophic cardiomyopathy, but from the patient's end, the manifestation of these problems is different. Obviously, the patient will not be concerned about the intraventricular space and volume, will not be concerned about the fibrosis. The main concern of the patient is the risk of sudden death because this has historically been linked to hypertrophic cardiomyopathy, and although not very frequent, in actual fact, quite rare, most of the patients who are diagnosed with hypertrophic cardiomyopathy are really concerned about the risk of sudden death.

Here is one big challenge, because the tools we have to evaluate the risk of sudden death are statistical, so we can only learn about the risk of sudden death by looking at patients with hypertrophic cardiomyopathy who have experienced sudden death and look at what risk factors they have had. The statistics are helpful but then they need to be brought down to the level of the patient again. They need to be individualized because the patient is not so worried about the general statistics but is worried about what will happen to them. Therefore, it's a big challenge to individualize the statistics and make them relevant to the particular individual patient.

Patients will also need to understand what it means when we tell them that they have a risk, but this is not high enough to warrant prevention, and we will continue monitoring. For them, it's a risk which is unprotected sometimes but, for us, it's a risk which is small enough to be lower than the level where any prevention would offer benefit. This is a concept that is different from patient to patient, and they have different approaches. At the end, the patient who nowadays want to become our partner in this discussion needs to be fully informed and understand all this, otherwise, a non-informed partner in the discussion will not be helpful and will not set up the ideal situation for the doctor and the patient.

The other big concern for the patient is the symptoms. The symptoms can be variable from day to day, from month to month, and from

year to year, and they need to be evaluated continuously. We, medical professionals, know how to evaluate the symptoms in the hospital, but what is more important for the patient most of the times is the real-life symptoms, so what they experience when they go up the stairs at home? They're not so much interested how they perform on the exercise bicycle in the hospital, but they're really concerned if they cannot pick up their small child from the floor or they cannot carry shopping bags and this is where the patient's main concern is.

The final issue which is sometimes very worrisome for the patient is the fact that they may inherit the condition, they may pass on the condition to their children, or they may share the condition with other members of the family. Nowadays, we have genetic testing available which can give us the genetic diagnosis in patients with hypertrophic cardiomyopathy in over 50% of the cases, which is good, but the next thing to do there is to interpret this for the patient, make them understand what this means for those who have the genetic changes and for those who don't have the genetic changes, how this information can be beneficial for them, and what we can predict, what we cannot predict. This information will help them and their families live with the condition and learn how to prevent the condition in some members of the family.

Coming to the healthcare professionals, obviously, the first challenge is how to accurately diagnose the condition, how to differentiate between obstructive and nonobstructive hypertrophic cardiomyopathy, how to describe the overlaps, how to identify other causes of symptoms in patients with this condition such as severe diastolic dysfunction and sometimes restrictive physiology. It is very important to have an open mind when we are investigating these patients because one size does not fit all and patients sometimes have multiple causes of symptoms, multiple reasons to be limited by symptoms, we need to look at all of them.

It is important for the healthcare professional to evaluate the risk of sudden death, to do that at regular intervals and to also evaluate the risk of thromboembolism and prevent them where appropriate. This is a very challenging procedure and it often requires the contribution of other colleagues of other disciplines. It requires to take into account multiple parameters, so nowadays, we're talking about a multiparametric and multidisciplinary approach in the evaluation and prevention of the risk of sudden death and thromboembolism.

Eventually, what is important to do is to manage the patient's condition and to manage it in the best possible way. We may set our own targets as healthcare professionals, but the main target for the patient is to have a near-normal life and near-normal quality of life and to be able to do things that they couldn't do before the diagnosis and the initiation of management. This may include nowadays new medical options such as the myosin inhibitors, the mavacamten and the aficamten, will include a lot of discussion with the patient, management of expectations, counseling, will include lifestyle modifications, advice about exercise, advice about family screening, and will also need to be individualized because no individual is the same with another one.

If I summarize what I have discussed with you in the last 10 minutes, the main concern is not to miss the forest for the trees. In other words, not to focus too much on small details and miss the opportunity to offer the patient a holistic approach for the diagnosis and management, and this is indeed the right solution for this type of disease. Thank you for your attention.