[MUSIC PLAYING]

STEVE R. OMMEN: The hypertrophic cardiomyopathy is a condition that has been under-diagnosed and overly feared throughout the world. In the United States alone, there are over a half a million people that have hypertrophic cardiomyopathy, many of whom are completely asymptomatic and unaware of their diagnosis. Some people can die suddenly. Sudden cardiac death occurs randomly without warning.

HARTZELL V. SCHAFF: Over 2/3 of the patients will have obstruction. And the obstruction to the left ventricular outflow tract is an indication for operation in patients that have symptoms. So we know now that 2/3 of the patients with hypertrophic cardiomyopathy and obstruction are candidates for surgery.

STEVE R. OMMEN: Hypertrophic cardiomyopathy is the most common inherited cardiomyopathy or heart muscle disease. People are born with the genetics for it, but the hypertrophy doesn't appear to start developing until adolescence, growth spurts, or beyond. It is possible for infants to born with thick heart muscles, but that's really quite rare and usually more severe expressions of the disease. And it's also been described as not coming on until people were in their fifth or sixth decade of life. So really, the onset can be at any time of life. And certainly the symptoms can occur throughout life.

HARTZELL V. SCHAFF: The common symptoms that patients have when they have obstructive hypertrophic cardiomyopathy are shortness of breath, angina-like chest pain and syncope. And unfortunately, some of these symptoms develop so slowly and over such a long time that the patients don't really understand how limited they are.

STEVE R. OMMEN: For patients who have symptoms due to hypertrophic cardiomyopathy, the first line of therapy is always to use medical management, medications. Usually, that's adding specific medications, but sometimes patients are on medications that can make their situation worse. And so some of the most effective therapy is removing the wrong agents, and then perhaps having to add in the right agents to help them with their symptoms down stream.

For patients who don't respond to those medical changes, or for whom those medications caused side effects that are intolerable, then that's when we move to things like surgical myectomy, which can more definitively relieve their symptoms.

HARTZELL V. SCHAFF: The patients who are referred for surgery almost always have either failed medical treatment or have side effects from the medicines that limit them just as much as the symptoms from hypertrophic cardiomyopathy. So operation to relieve outflow tract obstruction is to relieve the symptoms. And in some patients, to allow them to get off of the medications that are having unwanted side effects.

STEVE R. OMMEN: Surgical myectomy has been a very successful operation for many of our patients. However, it is not utilized as much as it maybe could be in part owing to prior perceptions about increased risk with the operation, lack of universal availability of surgeons who can do it. But in the hands of expert centers, the complication rates are very low and our success rates are very high.

HARTZELL V. SCHAFF: We now do a more extensive septal myectomy that extends towards the apex of the heart. And we've learned over the years that it's this distal portion of myectomy that's the most important in terms of relieving symptoms. The few patients who have had a second operation, who have been referred to us after having an operation that was unsuccessful, we found that the myectomy was not carried far enough into the ventricle. It's really not a regrowth of muscle. It's just an inadequate initial operation.

STEVE R. OMMEN: With surgical myectomy, the surgeon removes a portion of the hypertrophied septum, which is narrowing the path of blood, out of the heart. By doing this, it changes the direction the blood flows through the ventricle. It allows the mitral valve to function normally. And allows blood to leave the heart without increasing pressures or increasing forces. This muscle doesn't regrow over time. It is a permanent fix.

HARTZELL V. SCHAFF: We found that it's rarely necessary to do something to the mitral valve. And the hazard with doing something to the mitral valve, where it turns out to be unnecessary, is there is a chance of injury. So we would prefer to do a septal myectomy, come off bypass, assess the mitral valve with an echocardiogram during surgery, before then addressing the mitral valve if there is residual regurgitation.

We can tell if the mitral regurgitation is relieved immediately after the myectomy once the aorta is closed and the heart is restarted. The echocardiogram is done in the operating room and we know right away whether the mitral regurgitation has been relieved. An operation is available for some patients that have non-obstructive hypertrophic cardiomyopathy. And these are patients with the apical distribution of hypertrophy.

Some of those patients have diastolic heart failure related to very small ventricular cavities.

And in these patients, doing a transapical myectomy to enlarge the ventricle can improve their symptoms of heart failure.

STEVE R. OMMEN: While we see great results from the surgical myectomy as performed now, it is still something that should only be performed at true centers of excellence. Recent data that have come out have suggested that at low, medium, and even quote unquote "high volume" centers, there is a gradient of mortality, meaning it's highest at the low volume centers and lowest at the high volume centers.

But even those so-called high volume centers have mortality rates that are dramatically higher than what is reported from the true expert centers. And this is a procedure that should be done by those who are very familiar with this procedure, and doing a lot of them.

HARTZELL V. SCHAFF: At Mayo Clinic, we've done over 3,000 operations for hypertrophic cardiomyopathy. We do 200 to 250 operations each year. The mortality for the procedure is less than 1%, especially for patients who are otherwise healthy.

STEVE R. OMMEN: One of the biggest parts of every interaction that I have with patients is helping them understand what their individual risk for sudden cardiac death might be, and whether they might consider having an implantable defibrillator. Our patients who have had surgery have a lower rate of sudden cardiac death and a lower rate for their defibrillators to discharge among those who have had them.

HARTZELL V. SCHAFF: One of the things we've learned after doing septal myectomy is that actually the incidence of ventricular arrhythmia seems to be reduced. And this is shown in studies that look at defibrillator discharges and rates of sudden death.

STEVE R. OMMEN: The inheritance pattern of hypertrophic cardiomyopathy is autosomal dominant, which means each of the children of a patient with HCM has a 50/50 chance of inheriting this disease. We do recommend screening for all first-degree relatives, which is either genetic testing or echocardiographic-based surveillance. When a family has chosen to use echocardiography as their screening tool, we recommend that adult first-degree relatives get screened every five years. First-degree relatives who are adolescents or athletes, we usually screen every 12 to 18 months.

HARTZELL V. SCHAFF: Septal myectomy cures the symptoms of hypertrophic cardiomyopathy when it relieves the obstruction. But of course, patients still have hypertrophic cardiomyopathy, still need to be

followed by their physician for the other problems related to hypertrophic cardiomyopathy. But hopefully, are relieved of the shortness of breath, chest pain, or light-headedness that leads to the operation.

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