

Hypertrophic Cardiomyopathy (HCM)

Announcer: Welcome to the Mayo Clinic Cardiovascular Continuing Medical Education podcast. Join us each week to discuss the most pressing topics in cardiology and gain valuable insights that can be directly applied to your practice.

Dr. Klarich: Hi, my name is Kyle Klarich and I'm here in Rochester, Minnesota where I'm a member of the Department of Cardiology. And we are doing a session on "Interview with the Experts." And today our topic is septal reduction therapy in hypertrophic cardiomyopathy. And we have one of our outstanding surgeons, Dr. Bagameri. Gabor Bagameri, who's a surgeon at the St. Mary's Hospital, Rochester, Minnesota, along with me. And I refer a lot of my patients to him and he has agreed to talk to us about this very important topic for patients with hypertrophic cardiomyopathy. So, Gabor, welcome.

Dr. Bagameri: Thank you. Thank you for having me.

Dr. Klarich: Absolutely. It's a pleasure. So I'll start right in. What are the indications for septal myectomy in patients with hypertrophic cardiomyopathy?

Dr. Bagameri: Thank you. It's an excellent question. So hypertrophic cardiomyopathy is one of the most common inherited cardiomyopathy. It's primary heart muscle disease. The people are born with a genetic predisposition. The common symptoms usually when they have, so hypertrophic cardiomyopathy, people can have obstruction or without obstruction. When people, probably two-thirds of the patients, they have symptoms from obstruction, which can be addressing or can be provocative on exertion. The symptoms usually can be chest pain, and in general like chest pain, shortness of breath, or sometimes syncope. The first treatment for obstructive hypertrophic cardiomyopathy is medical treatment. And sometimes people, despite aggressive medical treatment, they still are symptomatic. That could be an indication for surgery, or they have developing side effect from the medication which is intolerable and that could be also an indication for surgery.

Dr. Klarich: Great. So basically we're taking patients who have failed medical therapy and then referring them for consideration of septal reduction therapy.

Dr. Bagameri: Yes.

Dr. Klarich: And they've failed because they're still having symptoms of either chest pain or syncope or pre-syncope, meaning almost passing out. Or, they're very short of breath of activity. And so then we would ask one of our surgical colleagues to see them. And what kind of surgical approaches then, are you going to describe to your patients with obstructive hypertrophic cardiomyopathy?

Dr. Bagameri: So the surgical approach requires open heart surgery, through the median sternotomy. And there is two way to approach. There is a lot of morphologic variation between the septal hypertrophy. It can be very just immediately below the valve which is the most common, the sigmoid. It could be in the middle of the ventricle or closer to the apex. And depending on the location of this obstruction or the hypertrophy, depends on what surgical

approach we are using. The most commonly used, so-called transaortic extended myectomy. When after going on the heart-lung machine we stop the heart, open up the aorta and retracting gently, the aortic valve, we're we looking down into the left ventricle and try to excise the enlarged hypertrophic muscles. Sometimes the hypertrophic obstruction is deeper in the ventricle and it necessitates a different approach. Sometimes you have to approach it from the apex.

Dr. Klarich: Really, you have to know the anatomy based on either imaging with cardiac MR or echocardiography to really plan the surgery up front. And maybe sometimes in cases, you might need both to understand not only where the obstruction is occurring, whether it's more at the base, which is obviously the most common in our patient population or can be at the mid or even at the apex or some combination. And it sounds like the most common surgical approach is through the sternum. So like opening up the chest and looking down on the heart and then making an incision on the aorta so you can actually see where the bulging muscle is.

Dr. Bagameri: Yes.

Dr. Klarich: If it's a basal septum. And then if it's the mid or at the apex, you may have to also or preferentially go to make an incision in the apex of the heart, and then look up at the bulge or the obstructive area. Is that correct?

Dr. Bagameri: Yes. So depending, again, if it's a middle of the ventricle, or sometimes it can be really hard to reach it all the way from the aorta, and then we use both approach. But sometimes it's just localized to the apex and it could be just the apical myectomy.

Dr. Klarich: Okay, so the mid sounds like it might be more complicated because you have to come from the aortic side of the heart, the septum and the apical side for the mid.

Dr. Bagameri: Yes, yes. Over the year, sorry.

Dr. Klarich: Unfortunately those are the most rare of our patients.

Dr. Bagameri: Yes. So over the years, what we learned, the most important aspect of the myectomy, that makes sure that we extend it far enough towards the apex. So sometimes when people come back with residue obstruction, it's not because the hypertrophies is going to come back, or because of scar tissue. It's usually because of the inadequate first surgery when they don't run deep enough down towards the apex.

Dr. Klarich: Yeah, that's a great point. And maybe we should emphasize that. So I always get asked by patients, when we take the muscle out, does it grow back?

Dr. Bagameri: The short answer is no. The muscle is not going to go back. When again, as I mentioned earlier, if they have persistent obstruction, it's usually implies that the resection wasn't carried far enough towards the apex. And that's when the surgical judgment and the review that cared for the imaging, the echocardiogram or the MRI to see whether apart from the transaortic action, we need additional trans-apical approach to get the complete resection of the bulging muscle.

Dr. Klarich: Great, thank you very much for that clarification. So a lot of our patients will also have arrhythmias, atrial fibrillation being the most common at the time of myectomy or septal reduction therapy. Are there arrhythmic approaches that surgeons can use at the time of the myectomy?

Dr. Bagameri: Yes, we have some adjunctive surgical procedures we can do to try to decrease the likelihood of atrial fibrillation. As you mentioned, it's quite common and patients with hypertrophic obstructive cardiomyopathy, they don't tolerate the atrial fibrillation because it takes out the atrial kick, less feeling of the ventricle. Usually the procedure, it implies pulmonary vein isolation when we are encircling pulmonary vein and try to isolate, which is the most common focus for the paroxysmal fibrillation. And at the same time, we are going to exclude either with surgical or we clip the left atrial appendage. This procedure adds minimal time and minimal risk to the surgery.

Dr. Klarich: Oh, that's great to know. So, some of our listeners might not know exactly why we are worried about the left atrial appendage. And maybe you want to say a few words about why we exclude the left atrial appendage either by clipping it or resecting it.

Dr. Bagameri: When patient go into the paroxysmal fibrillation and there is a irregular activity of the upper chamber of the left atrium, there is no regular contraction. So the blood can be stagnant. The left atrial appendage is a heart-pouching remnant from embryo even when the heart was developing. And the blood can get stasis, stagnant, and can form clot. And there is this clot, there is a possibility this clot can travel into the systemic circulation, go to the brain, and with devastating consequences of causing a stroke.

Dr. Klarich: Yeah. So being sure that the left atrial appendage, which is the most high-risked for having that stagnant blood. And when blood slows down, it clots. And when clots are formed, if the heart starts being, again as you mentioned, because the atrial fibrillation is intermittent or paroxysmal, then that clot can be passed into the circulation. It could go anywhere, but if it goes to the brain of course it causes a stroke, which is bad. That's great. So that's an important thing to consider if someone's going to surgery to also reduce risk of other complications of this disease, hypertrophic cardiomyopathy. How do you manage, so a lot of our patients will have mitral regurgitation, or leaky mitral valve, when they have the obstruction to their flow of blood out of the heart. For mitral regurgitation, how do you manage that at time of surgery?

Dr. Bagameri: So as you mentioned, people, with patients with obstructive hypertrophic cardiomyopathy, they have mitral regurgitation. It can be very dynamic. Sometimes they don't have it addressed, but an exertion gets worse. The main problem is when the heart squeezes and the blood coming out from the left ventricle, it comes through, we call the left ventricle outflow tract. Because bordered by the septum, and anterior leaflet of the mitral valve. In normal condition when the heart squeezes, this is a wide open channel and the blood can travel. In hypertrophic cardiomyopathy, especially with obstruction, because of the change of the hypertrophy and the blood flow vectors, the mitral valve gets moved into the left ventricular outflow tract. So, it creates an obstruction. At the same time and the mitral valve moves the entry in the posterior leaflet, they don't really close properly. And it allows the blood going back from the left ventricle into the top chamber, left atrium. And it cause increasing pressure in the

pulmonary vasculature, causing significant shortness of breath. Most commonly, even though there is some morphologic changes to the mitral valve, people would be elongated mitral leaflet. But increasingly the mitral valve is almost always normal, which means when you relieve the adequate obstruction, the mitral regurgitation is going to completely resolve. In rare instances, it's possible that we still have some underlying increasing mitral valve pathology. Like for example, a flare segment of the mitral leaflet. Our practices here is to perform an adequate myectomy, and become of the heart-lung machine. And then we look at that under transesophageal echocardiogram, and we should be able to see immediately the resolution of them, and the mitral regurgitation. If there is any evidence of the persistent atrial fibrillation, and we know, or mitral regurgitation, and we know for sure it's not because of the inadequate receptor myectomy, maybe a segment, then we usually go back on the bypass and fix the mitral valve. The biggest danger is try to intervene, because we can cause unnecessary injury, and then we know that we don't want to replace the valve because there is poorer survival.

Dr. Klarich: Yes. And it's interesting that you say that, I'm glad you made that point. Because, years ago we used to think that at least some institutions, not at Mayo Clinic but at other places, there were replacing mitral valve thinking that would relieve the obstruction. which it does do. But it's at the expense of adding another issue, which is a prosthetic valve which requires anticoagulation. So, to summarize your very elegant description, many patients with mitral, with hypertrophic cardiomyopathy will have mitral valve regurgitation and the vast majority of that regurgitation can be relieved with a successful reduction of septum, or the septum myectomy. But we have a safety net and we look with echo-cardiography, it's a generally a transesophageal echo-cardiography done intraoperatively, so that even before we leave the OR, the patients will be assessed to see that we've resolved the mitral regurgitation. If we have not, we can go back on bypass and make more adjustments to that valve. Would you say it's a correct summary?

Dr. Bagameri: Yes.

Dr. Klarich: Great. Is there a way to manage residual obstruction? So, that's something the surgeons never wanna hear. Okay. The mitral valve regurgitation is better, but we have still some turbulence of blood flow. We might have a gradient still, as we come off of the cardiopulmonary bypass machine. So what do you do about that, if you hear that from your echo or anesthesia colleagues?

Dr. Bagameri: We have to very carefully evaluate it. I have to make sure that we were far enough done to the myectomy. So we check with echocardiogram. We also check direct pressure measurement in the aorta, inserting a very small needle, and checking the pressure across the obstruction. If we have the residual obstruction, turbulent fluid that always implies that probably we are inadequate with receptor myectomy. So it's most commonly we go back on the heart-lung machine, we reopen the aorta, and we try to extend it. And what's very important here, it's not really deepening the previous myectomy incision, but it means that we are trying to travel further down more towards the base of the papillary muscle of the apex of the heart.

Dr. Klarich: We did touch briefly on it earlier. And you made the comment that the most common obstruction is at the base of the ventricle. The ventricle is actually shaped like a cone, if you will. I think, it's maybe patients don't realize that but certainly physicians are aware that the

heart's divided into a base portion, a mid portion, and apical portion. Apical hypertrophic cardiomyopathy is usually non-obstruction of the blood leaving the heart, but can still cause a lot of problems with heart function. And we have developed at St. Mary's Hospital with our talented surgical group. A procedure called an apical myectomy. It's not done that often. And I'm often interested that a lot of times when I talk about apical hypertrophic cardiomyopathy, not that many people are aware that this is available. Again, for patients that have failed medical management. So, but are there other indications, or what do you, what can you tell us about apical myectomy? When is it indicated, and what are you thinking about from the surgical of the evaluation?

Dr. Bagameri: You previously mentioned people with apical hypertrophic cardiomyopathy. Usually they don't have obstruction, but what happens is there is extreme thickened of the heart muscle and apex is not contracting. And the left ventricle volume is extremely small. So when they're not able to accommodate the fresh oxygenated blood coming from the lungs. And when the heart squeezes, there is very small amount of blood getting into the aorta. So they have a very low forward flow. So they develop heart failure symptom, extremely symptomatic, even though they don't have obstruction. And apart from heart transplantation the only real good option for them is the apical myectomy. But, basically have to forfeit certain requirements. Because certain numbers about the left ventricle and diastolic cavity, the number is usually it has to be 50 or less. Or certain amount of stroke volume, less than 30 milliliter, that could be an indication for apical myectomy. But it entails that we are making an incision approximately an inch, an inch and half incision, at the apex of the heart, which the apex in this disease process is not contracting. So it's not going to have any negative effect. And then we try to enlarge the volume of the left ventricle, excising muscle. And they were, and we were very successful with the instrumental doctor, developing this new technique for the apical myectomy.

Dr. Klarich: Great. Well that's a really nice explanation of how we evaluate. So we want someone that doesn't have a lot of blood leaving the heart. And you mentioned the value of a stroke volume of less than 30 millimeters, or 30 CCs, of blood per heartbeat. And the other number that you mentioned could you just repeat that was it the left ventricular and?

Dr. Bagameri: Diastolic. Yes.

Dr. Klarich: And what was that number?

Dr. Bagameri: Fifty, 50 millimeter.

Dr. Klarich: That's a really small.

Dr. Bagameri: Yeah.

Dr. Klarich: Left ventricular chamber size. And what's the most common way that we evaluate those two numbers?

Dr. Bagameri: Echocardiogram or MRI.

Dr. Klarich: Okay, great. Well, this has been a really interesting conversation about a very important technique for patients with hypertrophic obstructive cardiomyopathy that are failing medical therapy, which is always the first line. And we have a lot of different medicines that are available nowadays. But still there are a number of reasons either they don't tolerate the medications, or the medications don't completely get rid of their symptoms. That we can move onto very successful surgical interventions. Most commonly, that would be a basal septal myectomy, very rarely the mid-ventricle, which would require both incision in the aorta and at the apex. Or an apical myectomy, if they have a very small left ventricular chamber size, and a very slow, or a very low stroke volume. So, are there any other comments that you'd like to make before we leave our "Interviews with the Experts" on this topic, Dr. Bagameri?

Dr. Bagameri: Yeah, I think what's important to know that, this disease process is extremely underdiagnosed, and it's very feared mostly because of the perceived surgical risk. In expert centers, we know there is a direct volume outcome relationship. If you go search people go into surgery where it's frequently performed, the mortality and the morbidity is extremely low. It's around less than one percent. So this procedure can safely performed. It usually results in complete resolution of the symptoms. It doesn't necessarily cure the disease. The people, even if they became symptom free, they still have the hypertrophic cardiomyopathy. So they need lifelong surveillance and treatment by the cardiologists. But it's a better treatment option, because it's resolution symptoms. And it puts them back in the age-matched normal life expectancy.

Dr. Klarich: That's a really important point, and I'll just repeat it, because I think it's so amazing that if you have a successful myectomy, which is ninety-nine plus percent likelihood, that you'll go back to the age-matched lifelong survival. So this is a fantastic symptom-reliever, as well as allowing patients to have a normal longevity. So this has been a really fascinating conversation, and I appreciate you taking your time out of your busy day to inform our audience, in another "Interview with the Experts". Dr. Gabor Bagameri, Cardiovascular surgeon, Mayo Clinic, in Rochester, Minnesota. Thank you.

Dr. Bagameri: Thank you for the invitation. Thank you.

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