## Miguel Leal, MD (00:17):

Hello everyone, and welcome to the Hypertrophic Cardiomyopathy Podcast, titled invasive strategies, percutaneous and surgical; selecting the appropriate patients. Today is another one of a series of podcasts that has been presented by the American Heart Association, Hypertrophic Cardiomyopathy Initiative, sponsored by Bristol-Myers Squibb. I am Miguel Leal, Director of Electrophysiology at the University of Wisconsin, Madison. And I have the pleasure to be joined today by two colleagues and a patient representative.

#### Miguel Leal, MD (<u>00:48</u>):

With me here today, we have Dr. Carey Kimmelstiel, who is Director of the Cardiac Catheterization Laboratory and Interventional Cardiology at Tufts Medical Center, and a professor of medicine at Tufts University School of Medicine in Boston.

## Miguel Leal, MD (01:01):

I also have Dr. Hartzell Schaff, who is a cardiovascular surgeon at the Mayo Clinic in Rochester, Minnesota with specialty and expertise in hypertrophic cardiomyopathy management from a surgical standpoint. In addition, we have Adam Elliot join us today. We're always excited to have a patient representative who'll participate actively in our discussion.

#### Miguel Leal, MD (01:22):

Today we'll be exploring invasive strategies for patients with hypertrophic cardiomyopathy, including both percutaneous and surgical approaches in a management of this complex disease condition. Over the next 20 to 30 minutes, our goal is to clarify the state of the art available therapies, introducing those techniques, and also trying to explain which patients are more likely to benefit from them throughout their journey.

#### Miguel Leal, MD (01:48):

The first question that I'd like to ask our two colleagues here today is when is medical therapy not sufficient? Or in other words, when should we start considering invasive therapies such as percutaneous or surgical treatment of hypertrophic cardiomyopathy? And I'd like to start with Dr. Kemmelstiel, please.

## Carey Kimmelstiel, MD (02:07):

Thank you for the invitation. So in general, patients need to have obstruction. They need to have a hypertrophic cardiomyopathy with obstruction, which would roughly be about two-thirds of the patient population. And they need to be refractory to medical therapy. That is to say that they should have [inaudible 00:02:27] association probably greater than class two symptoms in order for you to consider invasive therapy.

## Miguel Leal, MD (02:35):

Thank you, Dr. Kimmelstiel. And Dr. Schaff, in your experience, what kind of patient typically arrives at your clinic to discuss the possibility of invasive surgical therapy?

#### Hartzell Schaff, MD (02:44):

Well, as Carey mentioned, most of the patients have had medical treatment that's been unsuccessful or partially successful, but I think there are nuances to consider in patients who are under medical

treatment. There are some patients whose symptoms of shortness of breath or chest pain are improved with medicines, but they have intolerable side effects say to beta blockers and are not satisfied. Other patients may have some improvement in their symptoms, but the cardiologist who's managing them has limited their activities. So the point is that what's intolerable symptoms to some patients maybe tolerable to others and vice versa. So it's a very individual issue.

#### Hartzell Schaff, MD (03:27):

In my view, the best approach is to mention to the patient at the outset that there are these possibilities of septal reduction therapy if medicines don't work. It seems to me that too often, we see patients who have been managed for several years, and by the time they're referred for septal reduction therapy, they've had this unnecessary period of limitation where they could have had that earlier and they say, well, I just didn't know. So part of it is the patient making the decision on whether the medical treatment is tolerable or intolerable.

#### Miguel Leal, MD (04:04):

Thank you, Dr. Schaff. And Dr. Kimmelstiel back to you, can you walk us through the percutaneous procedure? What exactly does it entail? A brief description of what a patient could expect, and again, if this is a procedure that works for everyone or for most people and so on.

#### Carey Kimmelstiel, MD (04:20):

The procedure is a percutaneous procedure. It takes approximately an hour and a half to perform. Virtually, all patients get a temporary pacemaker during and for a day or so following the procedure. Standard, we will do a right-heart catheterization. Obviously, we monitor our procedure doing a left-heart catheterization with a catheter in the ventricle. And then we do coronary angiography and find an appropriate septal artery or septal branch with the aid of myocardial contrast echocardiography. So within the Center of Excellence, one of the imaging experts is in the room with you and you try to find the area. You're obviously quite dependent on the patient's inherent coronary anatomy. And you then inject a branch of the LAD, the septal branch that has the area of what we would call SAM septal contact, that is to say, where the anterior leaflet of the mitral valve makes contact with the basal septum of the left ventricle.

## Carey Kimmelstiel, MD (05:20):

We then inject a small volume of alcohol monitoring the patient for a variety of different side effects, including heart block and arrhythmias primarily. We allow this alcohol to stay beyond a balloon that is placed in the [septal artery 00:05:37] inflated, an angioplasty balloon, and we keep that balloon inflated and inject the alcohol through the distal port of the balloon. We leave that balloon up for at least 10 minutes, and then we flush it afterwards and then remeasure our hemodynamics.

# Carey Kimmelstiel, MD (<u>05:53</u>):

In general, we judge a successful procedure by a greater than 50% reduction in the gradient during the procedure in which it's different than surgery, where they have an elimination of the gradient instantly. In our case, the gradient frequently comes back the next day because the stunning of the myocardium resolves. And then following that over a period of approximately two to three months, there'll be fibrosis of the basal septum. And so there will be a stepwise reduction in the gradient that is measured noninvasively following the procedure.

# Miguel Leal, MD (<u>06:26</u>):

That was excellent. Actually, without being an interventional cardiologist myself, I was able to sort of get a mental picture of the procedure or sequence that you so well described. So thank you for that. And Dr. Schaff, how about surgery? Can you just walk us through the surgical myectomy in any terms of how this procedure also has evolved over time in the treatment of these patients?

# Hartzell Schaff, MD (06:46):

Sure. Well, we review what the patient has to do with the preoperative preparation and intraoperative procedure and recovery. Intraoperatively, we explain to the patients that the operation takes about four hours, always seems a little longer for the families. The time on the heart-lung machine is about 30 minutes while we work down through the aorta to do the myectomy.

# Hartzell Schaff, MD (07:10):

Now, how we do the myectomy differs a little from the standard Morrow's procedure, but if the patient has isolated basal septal hypertrophy, we would just go through the aorta. If the patient has a more complex length of septal obstruction, we may have to do the procedure through the aorta and through the apex of the heart. And if the patient had pure mid-ventricular obstruction, we would probably approach that through the apex.

# Hartzell Schaff, MD (07:36):

So what the surgeon does during the operation depends very much on the phenotype, whether it's isolated, basal septal hypertrophy, mid-ventricular obstruction, or in some cases, apical hypertrophic cardiomyopathy. The stay in the hospital ordinarily is about five days if there's no complications. We tell patients that they can expect to be able to do normal activities of daily living when they get out of the hospital, we'd ask them not to drive for two weeks and not to do any heavy lifting for six weeks to allow the sternum to heal.

## Hartzell Schaff, MD (08:10):

But as Carey mentioned, the effect of the surgery is immediate in terms of relief for the gradient. And so many of them, even though they may be somewhat limited by their sternal incision will notice improvement in the symptoms of dismia or angina or syncope very early after surgery.

## Miguel Leal, MD (08:29):

Thank you, Dr. Schaff. So clearly that is lightened once there, in terms of how quickly are the symptoms expected to resolve, depending on what approach is undertaken. At this point, I'm going to pause it, my colleagues, and bring Adam Elliot to the discussion. And Adam, as a patient yourself, who has lived with diagnosis of hypertrophic cardiomyopathy, when you listen to two experts like Dr. Kemmelstiel and Dr. Schaff talk about percutaneous and surgical approaches, what are your thoughts as a patient, namely, how is an invasive procedure of percutaneous or surgical nature as seen by a patient such as yourself?

#### Adam Elliott (09:04):

Thank you, Dr. Leal. Whenever I found out that I was a candidate for the surgery, it was just like whenever I found out that I was going to have to have the ICD implanted. Even though I knew that was something that was probably coming down the road, still finding out that day it was challenging. And with that said, I was at a Center of Excellence and everything was explained in a way, just like most of

the other doctors where I was able to track and follow the reasons why. So once we started focusing on the treatment itself and the outcomes, then I was able to process it and get everything planned out.

#### Miguel Leal, MD (09:42):

And Adam, tell us a little bit about that informed consent moments. So when you're meeting with the proceduralist or the surgeon, and you're having that eye-to-eye conversation, what do you think is important for the patient to fully understand about the procedure that he's about to go through?

# Adam Elliott (09:57):

That's a great question. I very vividly remember that it's, I think it's important to have a family member or two there with you. That was something that was important for me, because while I'm pretty much able to track what and everything we're talking about and what the doctor's explaining, I think just having your family here all at the same time, that way, I think if there's any questions that they'd have that you might not think of or vice versa, it's important during that informed consent to have at least one, if not two other family members with you.

# Miguel Leal, MD (<u>10:29</u>):

Thank you, Adam. Dr. Kimmelstiel, in your opinion, is there such thing as an ideal sequence of procedures here? In other words, should the patient always try a percutaneous approach first or not necessarily? So what has been your experience in terms of the sequence of procedures, if both were to ever be needed and even a route that topic, can a patient go through both at some point?

## Carey Kimmelstiel, MD (10:53):

Yeah. I've evolved in my thinking over the years concerning this procedure, I think trying a percutaneous approach, and if that doesn't work going on to surgery is not a good idea. I think it's best to plan the best procedure for the particular patient population. I think the guidelines are very clear as it relates to this. And I certainly am supportive of the guidelines, and so far still understanding that surgical myectomy is the gold standard. And in general, we would perform alcohol septal ablation on poor surgical candidates.

## Carey Kimmelstiel, MD (11:26):

In general, in patients that don't have associated abnormalities of the mitral valve that are adding to the patient's obstruction, left ventricular outflow tract obstruction, I would say that one thing that gets glossed over that is important to understand and consider, that the guidelines kind of infer, but don't come out and say, is that what the patient wants is very important. So if the patient is otherwise a reasonable surgical candidate, and it's just not motivated to undergo surgery, in those cases, I really do not have a problem offering them alcohol septal ablation.

## Carey Kimmelstiel, MD (12:08):

The corollary of that is, we have taken patients that have had abnormalities of their mitral valve, for instance, papillary muscles that are adding to obstruction and we've ablated them because they were really poor surgical candidate and done fairly well. That would be my approach. I'm not a believer in everybody go percutaneous first. And if it fails, go to the surgeons, I don't think that's fair to the patient. I don't think that's fair to the surgeon, frankly, because when there's alcohol instilled in a coronary

artery, it's not a pristine case for the surgeon. And I would think, and Dr. Schaff can certainly answer this better, but I would think that would add to surgical risk and complications.

## Miguel Leal, MD (<u>12:54</u>):

That's a perfect segue there. Dr. Schaff, when you treat a patient that has undergone a previous alcohol septal ablation, do you notice a difference as far as the surgical complexity of the case?

# Hartzell Schaff, MD (13:00):

Well, if we could just back up for a minute in terms of the decision-making, I think that it was well said, but what you also should consider the fact that with alcohol septal ablation you'll often get a right bundle branch block, and with surgery, about a third of our patients get a left bundle branch block. And so if a patient has had a previous alcohol septal ablation in a right bundle, and we do a myectomy in the usual way and give the patient a pacemaker, that's not a good outcome for a young patient. It may be acceptable for someone who's older, but we know from long-term followup of our own patients who had myectomy, that having a pacemaker in place and having heart block is a risk factor for late death.

# Hartzell Schaff, MD (13:46):

So I think that especially for a young patient, they should be informed that their risk, if they choose the path of having alcohol septal ablation and then surgery if it doesn't work, does increase the risk of heart block and need for transvenous pacing.

# Miguel Leal, MD (<u>14:02</u>):

Thank you very much for that, that really adds perspective to the fact that this is not going to be a one size fits all, or maybe even a one size fits most approach. And it highlights the importance of the individualized care that we all recommend for this complex disease condition.

## Miguel Leal, MD (<u>14:17</u>):

Adam, just bringing you back to our conversation here, after this procedures take place, that are obvious follow-up needs, and both Dr Kimmelstien and Dr. Schaff have referred and alluded to some of them, from a patient's perspective, especially when you have to have surgery or a procedure done at a site that is remote from your traditional primary clinic sites, what do you think is important to be insured so that the patient's long-term follow-up needs are met?

#### Adam Elliott (14:45):

As far as follow-up for patient needs, I was fortunate whenever I had the surgery I had in May of 2019, that facility was fairly close to my house. One of the things we actually talked about was the possibility of going to Mayo Clinic. And I think the importance of that is kind of finding out, the determining factors for me would have been, okay, if I am going there for the surgery, how often do I need to go for a follow-up? Is it typically two, three times the first year, that it maybe two times next year? So that way I can kind of plan out. You have that expectation upfront because of the travel involved and everything involved in that. Those were things that we did talk about as far as I was referred to another hospital. Fortunately, we're able to do everything here in town, but those are things that I would've liked to have considered.

Miguel Leal, MD (<u>15:36</u>):

Thank you, Adam. And Dr. Kimmelstiel back to you, you're a busy practitioner and academic medical center. And my question would be, is alcohol septal ablation a procedure that would be offered throughout the country? Is this something that most academic and non-teaching hospitals are capable of performing or delivering, or is this something that you would still see should be restricted to a relatively finite number of centers, which have expertise on the topic?

#### Carey Kimmelstiel, MD (16:02):

This is of course, a hot button issue. And I have to say, I feel very strongly that this is not a procedure that should be done at a standalone facility that doesn't have a center of excellence. You really need to have clinicians that can define the disease optimally, that can advise the family, which is very important in terms of genetic testing. You need expert imagers that really understand the important aspects of disease that affects the mitral valve. You need to have well-trained competent surgeons to be colleagues in a team approach to discussing these patients. You need to have expert electrophysiologists as well.

# Carey Kimmelstiel, MD (16:52):

The other point that I would make is that having interventionalists, and I'm a card carrying interventionist, coronary interventionists, but thinking that you can translate your skills in atherosclerotic coronary artery disease to performing septal ablation in a very different disease is a very, very dangerous sort of assumption and it's incorrect. So I think these procedures, surgical myectomy and alcohol septal ablation, need to be done at centers of excellence that have all the clinical subspecialties and can contribute their expertise to the patient with hypertrophic cardiomyopathy. And in fact, this is something relatively new in the guidelines this year.

#### Miguel Leal, MD (<u>17:43</u>):

Thank you, Dr. Kimmelstiel. Dr. Schaff, as Dr. Kimmelstiel referred to, this is a hot button issue that has led some degree of discussion and controversy. What is your take on this question?

#### Hartzell Schaff, MD (17:54):

We agree that the patient is best served by going to a center where there are multiple specialists that can participate in the decision to go ahead with a procedure. And as regards the operation, what we see is that in centers with less experience, there is a tendency to do more to the mitral valve, and in some cases, to replace the mitral valve, because that does relieve obstruction, but it leaves the patient with a prosthesis and all the hazards of having an artificial valve in place for the rest of your life. So in the surgical world, the increased experience at centers of excellence will result in more patients getting an operation that is just a myectomy with fewer mitral valve interventions. And I think that's important.

#### Hartzell Schaff, MD (18:45):

Now, it is a difficult operation technically to teach because the exposure is poor, but we have a number of ways that we teach this to our residents and to visiting surgeons that have to do with the endoscopic visualization of the myectomy. We have a library of procedures that we've done, where people can see the myectomy done. And then it's also very important we think for the surgeons to study the images and to understand the difference between isolated basal septal hypertrophy, long segment septal hypertrophy, mid-ventricular obstruction, and [inaudible 00:19:26] hypertrophic cardiomyopathy.

Miguel Leal, MD (<u>19:28</u>):

I would like to thank my colleagues, Dr. Kimmelstiel and Dr. Schaff, and our patient representative, Adam Elliott for participating in today's podcast, during which we were able to discuss a lot of important points about the invasive strategies that are available in the management of hypertrophic cardiomyopathy, especially with an obstructive component to the disease that appears to be refractory to medical therapy. This podcast is part of the American Heart Association, HCM initiative, hypertrophic cardiomyopathy initiative, which is sponsored by Bristol Myers Squibb.

# Miguel Leal, MD (20:00):

In closing, I would like to remind everyone who is listening to us today to encourage your patients to play an active role in their medical care, always advocating for themselves and their family members. As we just heard from Mr. Elliott today, who kindly shared part of his personal journey with us, it is very important that you have a full understanding of your disease condition and healthcare providers such as ourselves have that responsibility to our patients. Anyone is also invited to go to the American Heart Association, hypertrophic cardiomyopathy patient website for additional educational materials. Thanks everyone.