Treating Heart Failure in Adults with Congenital Heart Disease: Heart Transplant

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. Burchill: Welcome to Interviews with the Experts, a podcast series from Mayo Clinic, Cardiovascular Education. My name is Dr. Luke Burchill, and I lead heart failure for adults with congenital heart disease, here at Mayo Clinic. And today, I am joined by my colleague and friend, Dr. Mauricio Villavicencio, the Director of heart-lung transplant, here at Mayo Clinic.

Dr. Villavicencio Theoduloz: Yeah, thanks very much for the wonderful invitation. I'm thrilled, you know, to talk about, you know, the congenital issues that are pertained to a heart transplant. Because I'm not by anywhere near to be a congenital heart surgery expert. But I come from the transplant world, and then there's a connection in there that's very exciting to talk about.

Dr. Burchill: And I think that together, we're just seeing more and more of these patients, aren't we?

Dr. Villavicencio Theoduloz: Yeah, I think we are, especially here at Mayo, there's a fair amount of referrals, you know, for heart transplantation. But all over the country, you know, the congenital teams are doing such an amazing job just to get this patient through and get through the adulthood. And then, you know, we get, have this wonderful tool, and we are learning more and more how to get good outcomes, you know, in heart transplant. So, as you transplant the patients, you know, quicker and with better outcomes, that you get more people coming around for this, you know, amazing tool that we have.

Dr. Burchill: And Mauricio, so many of my patients that I meet, they come and tell me the same story, and that is that they've been told that at some point, they're going to need a transplant. This is particularly some of the young Fontan patients that I'm meeting. So they've been told that they might need a transplant, but actually they haven't been told a lot about what actually is a transplant. So, I'm interested from a surgeon's perspective, what do you say to patients and families the first time you're meeting them to discuss the possibility of transplant?

Dr. Villavicencio Theoduloz: Well, I think I tell them that it is, you know, something that until, I didn't see, you know, how amazing it is. I didn't have a great sense of it, because essentially, you know, from one moment to another, you get from a patient that has end-stage heart failure, you know, to have normal function, and then we can, you know, reconfigure his anatomy, and then have, you know, a normal anatomy. So all of a sudden, after so many years, you know, with, you know, congenital heart disease, and have the trouble, you know, all of a sudden you have a normal heart. And that's simply amazing, when you're able to, you know, to see that. One patient of mine told me a little bit ago, once in a heart transplant, I didn't know how it was feel just to walk around the hospital, like I wanted all the time, but I couldn't. And right a few days after transplant. So it's, so I would say, you know, we provide a substantial benefit, you know, because of the gift of life that a donor give us, and we can make a great transformation. And then the downside will be, you know, we'll need immune suppression, and you'll need a lot of special care. So that's what we're here for.

Dr. Burchill: That's right. I think that that's something we all emphasize, is that it's often exchanging one chronic condition for another. It does require lifelong care, just like congenital heart disease does. And that relationship with the transplant team is going to be the new relationship that our patients forge after their transplant. One of the questions that I'm asked by the referring cardiologist is, so, when is the timing right? And who is the ideal candidate for transplant? Who's most likely to benefit from transplant? And again, from a surgeon's perspective, what are your thoughts on that?

Dr. Villavicencio Theoduloz: Well, it's quite a broad question, because I think, you know, it's essentially for me, in general terms, is somebody that, you know, the conventional therapies have been exhausted. So, everything that we can offer, there's no transplant, you know, have been done. And then the patient is still in heart failure, and brings a really bad quality of life for the patient. And we, one thing is, you know, really quality of life, but this could progress you know, to, you know, pulmonary edema, cardiogenic shock, the most extreme circumstances of heart failure. So we try not, you know, not to get there as much as we can. But, so first, you know, end-stage heart failure not susceptible to other therapies. And I would say that who's not appropriate for transplant? I would say that is very important that the patients could be compliant and then follow medical advice. Because what happens, I have had this before, that the patient has gotten a successful transplant. They have gone home, and then they have simple not taking the, you know, the medication. So after, you know, there's so many people wanted a heart transplant, and waiting on the list, and then get a heart, and then just lost the graft and lost the life. And because you don't take the medication, it is something that is so shocking, that, you know, for me as a surgeon, you know, if I, we do as a team, all this effort and then it is just, you know, not get to be successful in the long term, just because you don't take the immunosuppression, is quite, you know, shocking. So we like somebody compliant, would like somebody has support, because there are many issues, especially, you know, they're quite acute, you know, in the first three months after transplant, I would say. So we need some support. I would like somebody that would have, hopefully, you know, single organ failure but, you know, we have been progressing on this realm, that, you know we could do heart-kidney, we could do heart-liver, and we could do a heart-lung transplant with this. All of these, especially applicants, to the congenital heart disease patients. And so, but obviously, the less sick the patient it is in other organs, the better. And I would say something important, because we can, you know, do combined organ transplants that then get away with it. But I think something very important is the patient, is not that much the condition, you know, kind of poor nutrition, not moving that much, in a quiet terminal ill state sometimes, you know. We can do a surgically perfect heart, liver, or heart-kidney transplant, and then the patient doesn't bounce back. And that's all also very, you know, heartbreaking to see. So we like somebody that has that potential, you know, to bounce back from the.

Dr. Burchill: Yeah.

Dr. Villavicencio Theoduloz: You know, long-term consequences for heart failure and then, you know, get a, you know, a good life.

Dr. Burchill: Yeah, so much, so many important things that you just said. So I'm just gonna emphasize them. I want to talk to you about the combined organ and Fontan patients, particularly

heart-after-liver transplants. But just before I get to that, I wanted to summarize some of the things that I completely agree with. These patients do need a comprehensive evaluation upfront, because sometimes there are further things that we can do on the adult congenital side. It might be another surgery, even a high-risk surgery. It could be transcatheter interventions. It might be getting their heart rhythm under control. But that initial adult congenital evaluation, I think is really critical. I think that the cardiologist, particularly the local cardiologist, is a great partner in this process, because they often have that in-depth understanding and relationship with the patient. And people might be surprised to hear you, the surgeon, talking so much about psychosocial and adherence issues. But you're absolutely right, this is a scarce resource in the form of donor hearts, and we really wanna make sure that the people who receive these transplants have great outcomes. We don't see great outcomes with people who can't take their medications reliably. And the other groups of patients would be people who continue to smoke nicotine, which is an absolute contraindication. Continuing issues with substance use. Again, we don't see great outcomes. And people who don't have great family supports. So highlighting that information for us can be very helpful, and that's why our social workers play such a critical role in the evaluation of our patients. Your last point was on deconditioning, and we see that unfortunately, don't we? We see patients coming in very late stage in their disease and it starts to become a big question as to whether they're gonna actually be able to recover and benefit from their transplant and live their best life. So, thank you for all those great points. But let's talk about heart-after-liver transplant. I was discussing this with a colleague recently, a cardiologist, and they said, oh, so are you bringing the patient back, like in two different surgeries? And how far apart is their heart and their liver transplant? So maybe we need to really break it down, keep it simple. Can you explain heart-after-liver transplant? Why would we go in that direction, and why are we now doing that here at Mayo Clinic?

Dr. Villavicencio Theoduloz: Well there's several things, because sometimes, you know, sometimes patients might need just a heart early, or a, you know, as you know, a heart-liver, because they have, you know, low standing, high venous pressure, which, you know, ends up in have a liver disease, and then a liver cirrhosis. And then, it is quite tricky to do a transplant if you have, suppose that you have sort of an early liver cirrhosis, and you would try to do just a heart transplant, and then what you have after the operation is the terrible vasoplegia and coagulopathy, 'cause the liver would not be working. So, we struggle a little bit to define the line where we need the combined heart-liver. But, when the cirrhosis is established, we definitely wanna do a heart-liver. And we have been trying to switch the paradigm, and to do, you know, simultaneous heart-liver transplant for these patients with cirrhosis. And then, but we have been changing it to do it first, the liver. And as I said this is, you know, has several reasons to do this. As I said, first, you know, if you do the liver first, at the time that you do the transplant, you go on the heart and lung bypass machine, then, you know, your vasoplegia is less. So you suffer less, you know, with vasoconstrictors, which can end up in organ failure. And then, if the liver, you know, you implant the liver, liver start working immediately. And then, you know, the coagulation and, you know, most of these patients have multiple sternotomies, you know, and a complex surgical disease. So, you know, quite a lot of raw surfaces. You would like to have the best coagulation possible. And when the liver is strong, it's that much easier to accomplish. And a third reason that we would like to do that, to do the liver first, is just to, because if patients that are highly sensitized, they have had many operations, and they have antibodies, like human tissue, they have antibodies, then would like the, when we do a liver first, that helps taking care

of those antibodies. So there are multiple reasons to do that. And then we, in the last few years, have emerged this great tool, which is the so-called heart-in-a-box, that you could preserve the heart for a longer period of time. So what happens is, while you are doing the liver, you don't want the, you know, the liver surgeons to be rushing, you know, in doing their liver transplant. So they, you could put the heart in the box, and do the liver transplant, which is most of the time. At least the operation that the heart transplant, because there hasn't been any surgeries on the abdomen, like in the Fontan transplanted patients. So, usually a primary abdominal operation. So you could do the liver quick, not much problem. Leave the heart in a box and then transplant the heart. And that brings all the benefits that we just talked about.

Dr. Burchill: And if people were to sort of watch a video of the heart-in-a-box, it literally looks like a heart beating inside a special medical box. Correct?

Dr. Villavicencio Theoduloz: Yes, it's kinda, if you would like to think about it, the way I conceptualize this, it's that it's kind of of ECMO thing essentially, but it's a kind of sophisticated, in terms that you put the heart in this device, and you perfuse the aortic root essentially, most of the time kind of 700 or 800 units per minute of blood, of, you know, a normothermia. So you perfuse it, and you perfuse it with, and then that gets drained into a pump. The pump, you know, against, pushes the blood flow, and then goes through an oxygenator, and then goes again into the heart. So, pretty much like a sophisticated ECMO circuit. So, and then you can monitor the hematocrit, the pressure on the aortic root, how's the degree of your saturation. And one thing that we have been using, and it's the lactate, because the heart has this ability to consume lactate that's different than other tissues. So, as soon as the lactate levels on the heart involved are coming down, then we think the heart, you know, is working better, better preservation. But there's a fair amount that's still, that we need to know, because we kind of take a look, like over the top of the device, how the heart is beating, you know, sort of, if it is beating strongly, you know, we'll proceed with transplant anyway. In case, you know, you have a good lactate, but then it looks like quite weak. Then we get scared, we don't proceed with the transplant. So it's a very nice tool, and allow us to get the heart, you know. Before we have this threshold, four hours to do the transplant, and now we're comfortable, you know, take eight hours, with total out of the body time with the heart in, for a liver-heart transplant.

Dr. Burchill: Amazing innovation. So, the heart-in-a-box, as you were just explaining. And also heart-after-liver transplant, putting that liver in first, because it's gonna help with our blood pressure, counteracting the vasoplegia. It helps with all of those coagulation products that deliver manufacturers, and it's also helping with sensitization. So, some really exciting things happening with transplant, here at Mayo Clinic, particularly benefiting our adult congenital heart disease patients. Thank you for all your leadership here for our program. And thanks for coming and spending time with us today. This has been very informative. Thank you.

Dr. Villavicencio Theoduloz: Appreciate the invitation. Thanks very much. Have a good one.

Announcer: Thank you for joining us today. Feel free to share your thoughts and suggestions about the podcast by emailing cvselfstudy@mayo.edu. Be sure to subscribe to the Mayo Clinic Cardiovascular CME podcast on your favorite platform and tune in each week to explore today's most pressing cardiology topics with your colleagues at Mayo Clinic.