

Heart Catheterization in Adults with Congenital Heart Disease: When, Why, and How

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 back to "Interview with the Experts", a podcast series from Mayo Clinic Cardiovascular Education. I'm your host, Dr. Luke Burchill, and I'm leading the heart failure care pathway here at Mayo Clinic. And joining me today is Dr. Will Miranda, my colleague in the adult congenital heart disease clinic and the person that I look to when I need catheterization and need to understand the results of those cath in our adult congenital heart disease patients. So thank you for joining us today, Dr. Miranda.

Dr. Miranda - Oh, thank you Luke. Thanks for having me here. So hopefully we'll be able to clarify and answer some questions.

Dr. Burchill - Yeah, so I thought we'd really start simple and let's talk about exactly what is heart catheterization? When we talk about that with our patients, how do you explain it to them?

Dr. Miranda - Sure. And I think that's a great question. So I think cardiac catheterization basically involves two components. So one is measuring pressures and flow, and the second one is looking at structures. And that can be the pump, for example. It can be the vessels, it can be the vessels feeding the heart. So it is doing a functional assessment, that's pressure and flow, and doing a structural assessment, looking at vessels or chambers for example. So taking pictures similar to what we would do with a CT scan, for example.

Dr. Burchill - And so why do we need to do them? I mean, we've got echoes, we've got CTs, we've got MRIs. Why do we actually need to do heart or cardiac catheterizations?

Dr. Miranda - Yeah, so historically, for example, everything started with cardiac catheterization and then echo came along and then CT and MRI came along and then cardiac catheterization became less and less used, mainly because it's an invasive procedure. But we have to remember that every test that we do is not perfect. So you can still have an echocardiogram or a CT that is not fully diagnostic, it doesn't give you the full answer, or you have two pieces of data that don't match. They're discordant, we call, that's the medical term we use. And therefore your cardiac catheterization can be the tiebreaker. So although we don't use it as often as we used before, today the cardiac catheterization tends to be the final test, the one that we use to make a final decision when things don't add up.

Dr. Burchill - That's so well explained. And if we focus now on our adult congenital heart disease patients, these are patients with a high prevalence of residual valve problems, other

congenital heart lesions. Maybe if you can comment on how we're using cardiac catheterization for adult congenital heart disease patients.

Dr. Miranda - So I think there are two main issues or questions. So the first one is establishing that the symptoms are cardiac. I think there's perhaps a preconception or a tendency to say that just because one has had congenital heart disease the symptoms are cardiac, versus, for example if you saw somebody in clinic that had hypertension and was short of breath, the first question to answer is this cardiac related or not? So I think before making the diagnosis of cardiac shortness of breath or cardiac symptoms in someone with congenital heart disease, we should go through the same exercise that we would do in any cardiac patient. So that's the why. And I think the second one is try to understand what's causing the symptoms, if we do think that the symptoms are cardiac. So this is a little bit different, perhaps more challenging than in patients without congenital heart disease because there are several imperfections, if you will. You mentioned that patients have had surgery before, they might have some residual leakage of one of the valves. So the idea is not so much to see if things are perfect, but try to understand how much these imperfections are contributing to symptoms, 'cause this is how we're gonna treat, right? If the valve is the main issue and the valve leaks a lot, therefore it would make sense by tackling the valve would help. If we don't think that the valve, despite the valve leaking, the valve is the big problem, then it's a complete different story. So the second thing is try to explain what is the main driver, if you will, in causing the patient's symptoms.

Dr. Burchill - And so I'm thinking about some of our listeners out there. It will include cardiologists who are seeing adult congenital heart patients. And those patients will definitely have symptoms. And as you explained, their echo may not fully explain their symptoms or they may be discordant. It's not uncommon for us to meet the patient who has really significant symptoms and they don't seem to be fully explained by that echo or non-invasive imaging data. So this is the kind of patient where we would generally in our practice, say, think about cardiac catheterization sooner rather than later. And don't just attribute those symptoms to them having lived with congenital heart disease all their life. Do you agree?

Dr. Miranda - I agree. And I think you're kind of trying to get into, I think, talk about how we do this actually. If you were to do this especially if you're doing this in a proactive approach, you know, how thorough should we be? And I think this is important. I think, you know, it's been shown over and over in patients without congenital heart disease that when things are obvious or overt or if they're completely obvious at rest, then perhaps you missed the boat a little bit and you really wanna recognize things and diagnose things when they're early to of course treat and tackle in an earlier state instead of just waiting until more events disease has developed.

Dr. Burchill - Yeah, I think another key difference between congenital and acquired heart disease is with an acquired patient, particularly if I'm sort of thinking with my heart failure hat on, we're more likely to start some treatment based upon our heart failure guidelines and based upon something like say reduced ejection fraction and see how the patient responds. I think we don't have that evidence base in our adult congenital heart patients. And I really appreciate the emphasis that you are putting on. We really first need to ask the question, what is the diagnosis

here? And does that explain this patient's symptoms? So we are going to use cardiac catheterization sooner in our evaluation compared to the standard acquired heart disease or heart failure patient. Do you agree?

Dr. Miranda - No, I totally agree. And I think you can also take the other end of the spectrum. For example, somebody that has been worked up for transplant. This is something that is a fear or perhaps even an expectation for some patients with some forms of congenital heart disease. And sometimes we see patients that are quite symptomatic. And again, everything would point towards that this is cardiac, right? And we're talking about cardiac transplant, it doesn't get bigger than that. And yet, even in some of those patients we've seen that there's a big component of non-cardiac disease. So I think it's important that we do it early, but even in the later stages when everything would point towards cardiac, I still think that we should be as meticulous. So you can argue that there's a role in the early detection but even in the sicker patient, the quote unquote if you will, there's still a role for being meticulous in ensuring that again, we're addressing the primary issue.

Dr. Burchill - Yeah, I think that that's so valuable. Equally important for understanding when someone's symptoms are non-cardiac. I think that's a perfect point for people to take away from this conversation. Cardiac catheterization plays a role there. And of course it's not a standalone test. We are looking at how it integrates with the other findings, for instance, on exercise tests and things such as the non-invasive imaging that we might be doing with echo and MRI. So I wanted to talk to you about some of the newer research that you've been doing, particularly using exercise cath. The first question I get is, how can you safely exercise people at the time of a cath? So I might start there and let you explain what is actually involved in exercising people on the table in the lab and why would we do that?

Dr. Miranda - Yeah, and that's a question that comes up a lot. And as you can imagine, both patients and providers might see this idea a little bit different than what we typically do. And, but perhaps the first thing to remember is that people have been doing this for 60 years. So people started doing this in the early days of cardiac catheterization with way less fancy hardware compared to now. So again, this idea has been around and in fact the first studies in exercise cardiac authorization came from congenital heart disease. So I can argue that we're just coming back full circle. Now the way we do it, and again, that's when things become a little bit different. I think it's critical that we explain the patients and inform the patients properly because you know this is typically done from the neck and from the radio approach. So it's different than the typical femoral approach, than most congenital procedures are done. Of course, this cannot be done under general anesthesia. That's a big change from pediatric cath, for example. And even if your prior cath wasn't done under GA, this one, since you're gonna pedal, so bike for most of the cases, you have to be awake. So there's some inherent differences here that even from the access standpoint, from the amount of personnel that is involved, to what actually entails. And I think it's critical that would tell the patients. But I'll tell you, as crazy as this might sound, I think it is really, really critical and it provides important incremental data. And I think perhaps the simplest way to explain why you would do this is that if you think about the standard patient that you see in clinic, the patient typically has exertional symptoms. And then the next step would be to do an outpatient cardiopulmonary exercise test, since your symptoms are cardiac and exertional. Now

then when we come to the cardiac catheterization laboratory, everything is done under a fasting state and at rest. So by definition, and this has been the standard approach for several different reasons and I think we can talk about that, but we're by definition comparing the exercise test that is done in the outpatient setting to the resting invasive data. And the idea is that maybe we should be comparing apples to apples. So if you go on the treadmills as an outpatient, we should go ahead and exercise you in the cath lab as well and we can do that safely. And I'll tell you, you know, the number of diagnosis that have been unmasked by using the exercise, even if you use the non-congenital data, about 50% of patients would go undiagnosed if you didn't exercise. And I think in our congenital practice, in our experience, the numbers actually have been a bit higher. So there is value and I think it does change management.

Dr. Burchill - And so that term unmasking, what are some examples of the kinds of diagnoses that you are unmasking by performing these exercise caths?

Dr. Miranda - Several things. And I think this is be, the idea is to try to do exactly what we do on the treadmill in the cath lab. So we're gonna look for, first is this cardiac or not? Is there pulmonary limitation or not? If this is cardiac, is it because the feeling pressure, so the pressure side of the heart are high? Is it because the pressures in the lungs are high? Is it because there's not enough blood being pumped out the heart? And then if so, is it because the heart rate is low? So all these different components and then, and at the end we say, okay, this is not cardiac. So what's the issue? Is this related to, for example your muscles being deconditioned, your legs. So it's kind of going to the drawing board and then actually going through all these steps and all these components that can make one winded and ensuring that these are there or these are not there because again, their therapy will be completely different.

Dr. Burchill - And would you say that the HFpEF, the heart failure with preserved ejection fraction, they're group that have really benefited from the use of these studies for actually validating that they do have an issue that may not have been detected on resting imaging?

Dr. Miranda - Oh, absolutely. And I think there's some subset of patients, for example, the Fontans. I mean, the typical example is a patient that is profoundly symptomatic or even in some we've used the term Fontan failure, and yet they come to the cardiac catheterization laboratory and the numbers are completely normal. So as you can imagine that creates a lot of frustration for the patient, a lot of frustration from the providers. And sometimes this becomes a barrier, for example during transplant evaluation. 'Cause you say, well, listen, you're profoundly symptomatic, your treadmill doesn't look good at all, but yet your cath is completely normal. So you have this big discrepancy here that sometimes I think gets in the way. Now, I think is also important that we remember one of Barry Borlaug's, you know, pet peeves, that the way the disease occurs is that we start with everything being normal. The next step is that things are only normal or ab, I'm sorry, only abnormal with exercise. And then in more advanced stages things are abnormal at rest. So what you want is actually to detect things early on. And by definition, when that happens everything is normal at baseline. So the question is, do we have somebody that has normal resting data and normal exercise data? Well, we have somebody that has normal

resting data but abnormal exercise data. So the next step in terms of disease progression and I think we can only unmask that to go back to the word we used, by exercising them.

Dr. Burchill - And I think the planets are aligning at this moment in time because we have more effective treatments, more effective treatments than we ever had particularly for that first step from being normal to having problems with exercise. I think that's been part of the difficulty in clinical practice. You know, a decade or so ago, maybe we just didn't have much to offer, or at least an evidence base to inform the treatments that we were offering these patients that had that subclinical disease. I think that that's really changed even in the last few years. What are your thoughts on that?

Dr. Miranda - Yeah, and I think the trend is exactly what you said. I mean, the earlier we diagnose, the earlier we treat, and the hope is that then by that we'll slow down or even improve some of the disorders we face. I mean, I think a great example is valve disease, right? We started in the sixties and seventies dogma was that you wait until the very last minute because your intervention is actually more dangerous than the disease. Now things have changed, you know? The amount of invasiveness of the intervention has really gone down and then the medical therapy has evolved tremendously. So I think we just have to change our goal here. And the goal is no longer wait until the disease is really advanced or bad, you know, to use simple terms, but really to pick up things at the very beginning so that we're actually avoiding all the consequences of untreated disease for decades to come.

Dr. Burchill - And I'm just, as we get close to finishing here, I wanna really emphasize a key point that you've made. These studies can be so helpful for patients that are highly symptomatic, but where we're ruling out significant cardiac disease. And I can think of a couple of patients certainly that we've collaborated on that had been part of the healthcare system, seeing different providers, different specialists, because they were highly symptomatic. They had discordant data, discrepant data on various tests. And doing this study was really the first time we were able to say we've excluded significant cardiac disease. We can reassure you. Your oxygen levels are not actually dropping, but you do have some issues say with your peripheral muscle and metabolism. You are safe. There's nothing terrible that's going to happen. We had one patient who didn't need to continue using continuous oxygen because we're able to demonstrate that that patient really didn't need it and they didn't have a shunt which had been diagnosed or misdiagnosed locally. So I do think this is just as important and for those listeners out there, that patient that keeps coming back in with symptoms where the tests just don't add up, I think that this is an important test that can provide reassurance for the patient, but also for the provider.

Dr. Miranda - And I think I would add one last thing and I think this is one of the lessons we learned from the HFpEF practice here. And to be honest, I think the only reason we can do all this fancy stuff in congenital heart disease is because of Barry Borlaug, you know? Who has really paved the way from, you know, from an infrastructure standpoint, from an expertise. So I think we're very fortunate, spoiled here, to have all this infrastructure already available to us. And then in HFpEF, even when we had no therapies, so this is before SGLT 2, all these newer drugs, sometimes just given the patient a diagnosis. They say, listen, unfortunately we don't have

a drug that is shown to work, but this is cardiac dyspnea, okay? So all these symptoms that you had for decades and they've remained elusive, this is cardiac. And some patients would feel so empowered or relieved to say, listen, finally, I have an explanation for my symptoms. And I think that's important. I mean, that's one of the reasons to do a test is to make a diagnosis. Sometimes you might not have the perfect treatment for that, but I think just giving the patient answers I think that's important too. So I think you're absolutely right. Ruling out disease is critical, especially when we're treating things that we don't know. But sometimes even giving the patient the answer, this is what you have, I think is actually quite beneficial from a quality of life standpoint.

Dr. Burchill - Yeah, I've certainly seen that too. I think that that's a great point and a really positive one to end on. So thank you so much for your time today, Will. Appreciate it. Any final comments?

Dr. Miranda - No. Anytime. My pleasure. And hopefully we'll keep understanding and pushing forward and helping our patients with their early diagnosis and therapy.