CHET RIHAL: Hi. This is Chet Rihal. I have a very special guest with me here today. Dr. Joseph Dearani is chair of the Division of Cardiovascular Surgery at Mayo Clinic Rochester, and he's a specialist in pediatric and adult congenital heart surgery. Joe, welcome.

JOSEPH DEARANI: Thank you, Chet. It's nice to be here.

CHET RIHAL: Great to have you. Listen, we're here to talk about Ebstein's anomaly. This is an anomaly that we in the cardiology world have been learning about since the early days, but can you tell us? Refresh our memory. What is Ebstein's anomaly? Why is it important?

JOSEPH DEARANI: So Ebstein's anomaly is an anomaly, actually, of the right ventricle. And most of the attention focuses on the tricuspid valve, but the tricuspid valve develops from the right ventricle. And the right ventricle is the underlying abnormality. And as a result, we end up with a myopathy of the right ventricle. We end up with a dysplastic, nonfunctional tricuspid valve. And then there are other associated defects that can be related.

There are arrhythmias that frequently can be associated. For example, Wolff-Parkinson-White and, with time, atrial arrhythmias. There's often a hole in the heart, usually atrial septal defect or patent foraminal valley. And the anatomy from patient to patient is different. So in contrast to other congenital defects, like a standard atrial septal defect or VSD where it's very, very reproducible, the anatomic finding from patient to patient-- with this, the anatomy is different from every single patient. And so this has made it very challenging for cardiologists and surgeons to get into a comfort zone, because every one is a little bit different.

CHET RIHAL: So Joe, typically, we think of this presenting in infancy or childhood. But over the course of my career, we've stumbled-- I've stumbled across a couple of patients in their adult years with previously undiagnosed--

JOSEPH DEARANI: Yeah.

CHET RIHAL: --Ebstein's anamoly. So what are the common presentations?

JOSEPH DEARANI: So this is the most fascinating congenital anomaly, because you can have a presentation from a highly symptomatic newborn that is tied to a ventilator that needs urgent surgery, And you can have the other end of the spectrum, the 70-year-old that has snuck through all of these adult years, who then has a chest X-ray, has some cardiac enlargement, which prompts a cardiac work up and it's a mild to moderate form of Ebstein's anomaly. And then, of course, everything in between. In children, the most common, probably, clinical presentation is cyanosis, because an atrial septal defect is very commonly present. And of course, they have right-to-left shunting and they're desaturated, and this usually prompts medical attention.

But when the atrial septal defect is small, like patent foraminal valley, or it's absent, those are the patients that actually have a delayed presentation, because we know with many other disease entities that tricuspid regurgitation can be tolerated for extended periods of time. And so it may not get picked up until arrhythmias become apparent, or the rare patient where you actually have Frank right-sided heart failure. Fortunately, that's the exception, not the rule.

CHET RIHAL: Joe, what proportion of patients with this anomaly eventually undergo surgery?

JOSEPH DEARANI: Oh, I think, eventually, the vast majority of them will need surgery, either for symptomatic tricuspid regurgitation, right-sided heart failure, either sign cyanosis or paradoxical embolism, which we may see in the adult patient. I think once it's identified, at some point in time, the majority of them will need operation.

CHET RIHAL: Is it ever too late to operate on them, and what is the ideal timing of surgery?

JOSEPH DEARANI: Ha. You've really struck a sensitive question there. This has been the topic of conversation and still is frequently the buzz topic at many of our meetings. Timing of surgery with Ebstein's can be very, very difficult. Clearly, when they're symptomatic, surgery's indicated, but many patients are not symptomatic. And if you take the cyanotic element out of it, probably the most common symptom is just fatigue. And it can be very subtle.

And for a parent, it may be a few more naps. It may be going to bed a little bit earlier with their child. Fortunately, Frank right-sided heart failure is very, very rare, and that would be, probably, the patient where conventional surgery may be high risk, or, in some situations, it may be too late. But under most circumstances, it's really-- most everybody could benefit from surgery at some point in time.

CHET RIHAL: And Joe, in the range of operative repairs that you perform, is this on the relatively straightforward end of the spectrum, or the more complex end of the spectrum?

JOSEPH DEARANI: No, it's the complex end of the spectrum. I mean, I've committed my career to trying to understand and learn this diagnosis, and, of course, the hope is that you're going to be able to repair the valve. And this, interestingly enough, there are more valve repairs described for this anomaly than anything else in cardiac surgery.

CHET RIHAL: Really?

JOSEPH DEARANI: And that's because the anatomy's variable. So, you know, you apply different techniques that you've learned from one approach versus another, and you adapt it to whatever anatomy you find. Now, there are certain underlying principles that you try to stick to, but at the end of the day, every repair is a little bit different.

CHET RIHAL: So what are those principles? Can you tell us about the repair that you performed here and perhaps Dr. Danielson's role in developing this?

JOSEPH DEARANI: Well, Dr. Danielson was one of my mentors, and it is a signature diagnosis at Mayo. Our experience in surgery now exceeds 1,000 cases, and probably 2/3 in children and about 1/3 in adults. So we have the whole age spectrum covered. And I think the cardiologists and the surgical teams are in a really good comfort zone right now. The goals of surgery are the following-- repair the valve. And I think now the wide variety of techniques that you can apply to repair and preserve native tricuspid valves has gotten really good. So fortunately, we can repair the vast majority.

Second, the right ventricle. We talked about this myopathy. There's a specific area of the ventricle that is very, very thin in the atrialized right ventricle. We generally treat that with a plication. Any intra-atrial shunts we typically close, with rare exception. There might be some situations in an infant where we would do a subtotal closure. Arrhythmia surgery is going to be dependent. I mean, arrhythmia surgery is a standard part of the operation in a 50-year-old. It's going to be rarely--

CHET RIHAL: Is it for WPW, or--

JOSEPH DEARANI: No, or atrial arrhythmias.

CHET RIHAL: Yes.

JOSEPH DEARANI: No, the WPW is something different. That, in general, is treated by your colleagues in the cath lab with a mapping and an ablation strategy. But atrial tachyarrhythmias develop with time with this diagnosis with or without surgery. So in the middle-aged adult patient, it is very common for there to be a history of palpitations, and maybe a halter shows some PACs or transient runs of atrial tachychardia. And so [INAUDIBLE] surgery is pretty much part of the whole operation in a 50-year-old, where it would be very unlikely in a three-year-old. And then--

CHET RIHAL: Joe, what's the outlook for these patients--

JOSEPH DEARANI: It's good.

CHET RIHAL: --after surgery?

JOSEPH DEARANI: Yeah, so the outlook-- first, the survival is excellent, and that's the most important thing, I think, for the patient community. The second thing is is that while we have gotten very good with surgery, it still is one of those lesions where the potential need for reoperation down the road is a very real thing. And the evolution of tricuspid valve repair has gotten better and better and better. In our own experience, by 20 years, almost 50% of the patients have needed another operation.

And we think with the more contemporary repair techniques that will be much better, but this is clearly a lesion where it's, I think, important to counsel parents and/or patients themselves, depending upon their age, that there is a possibility that they might need something, another reoperation or an intervention-- arrhythmia, for example-- at some point in time down the road, which really emphasizes the lifelong surveillance thing with this diagnosis. It's critically important. This is a serious disease, and they always need to have oversight by you and your colleagues.

CHET RIHAL: Joe, is this one of those diagnoses that can be made be antenatally--

JOSEPH DEARANI: Yes.

CHET RIHAL: --and perhaps even treated antenatally as fetal surgery?

JOSEPH DEARANI: Yeah, that one is a tough one. I think it definitely can be identified prenatally. And in fact, that is essentially a risk factor. And when it's can be diagnosed prenatally, that usually is because the size of the heart is very, very large. And we know that those newborns, we struggle with. They're a challenge for the intensivists in the NICU. And in the current era, it still is the highest risk operation we do in the newborn period when it's required. Even in experienced hands, we're talking about a mortality rate of 20%, 25%, which, for a surgeon, is quite a discouraging number when you can do an arterial switch procedure, which is very complex, with a risk of 1% to 2%.

CHET RIHAL: My guest today has been Dr. Joe Dearani, Chair of the Division of Cardiovascular Surgery here at Mayo Clinic Rochester. We've been talking about Ebstein's anomaly, and Joe has gone through it in great detail. It's been very informative. I hope that you have found it as educational as I have. Thank you for listening today. Joe, thank you very much for being here.

JOSEPH DEARANI: Thank you for the invitation.