Sawbones 185: The Tetralogy of Fallot

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[theme music plays]

Justin: Hello everybody, and welcome to Sawbones, a marital tour of misguided medicine. I'm your cohost, Justin McElroy.

Sydnee: And I'm Sydnee McElroy.

Justin: No middle name this time, Syd.

Sydnee: Oh no.

Justin: I like to get straight into business.

Sydnee: Let's get straight to it.

Justin: Okay.

Sydnee: Are you gonna get straight?

Justin: You do. You're the business.

Sydnee: Well, I am the—I know.

Justin: For the party. If we were a mullet—

Sydnee: [laughs]

Justin: As a couple…

Sydnee: Uh-huh. I'll be the front.

Justin: You'd be the front, and I'd be the back.

Sydnee: Right.

Justin: For sure.

Sydnee: Uh, so, I would be what gets us hired.

Justin: Right.

Sydnee: You'd be what gets us fired.

Justin: Fired. Exactly.

Sydnee: Right?

Justin: Exact-a-mundo.

Sydnee: [laughs] That's fair. So, Justin, occasionally, it's nice to cover a topic that a lot of people are talking about.

Justin: Mm-hmm.

Sydnee: Are hearing about. Um, right now, a lot of people probably watched Jimmy Kimmel's, uh… talk, speech, monologue…

Justin: Monologue.

Sydnee: Story.

Justin: Monologue. Monologue.

Sydnee: Story.

Justin: Story, yeah.

Sydnee: Uh, that he told on—either because you watched his show, or because you saw it on Facebook, like me. Because I, it was shared—

Justin: It went viral, as they say.

Sydnee: Yes. Yes.

Justin: And that's kind of internet slang.

Sydnee: Is that what that—

Justin: Web professionals. People like me in web media. We say 'go viral.'

Sydnee: It also has to do with an infectious disease that is a virus, like that term.

Justin: Ah, well that's—yeah, that's… not my best choice of words then. So sorry about that everyone.

Sydnee: [laughs] Uh, but if you haven't seen it, Jimmy Kimmel is telling story. It's a very touching, moving story of the birth of his child who had a congenital heart disease. Heart defect.

Justin: Mm-hmm.

Sydnee: Excuse me, a congenital heart defect called tetralogy of Fallot and uh a lot of people probably aren't familiar with it. What that is, what that means.

Justin: I know I wasn't, certainly.

Sydnee: So, I thought, and so did Shantell, one of our listeners who wrote in and suggested this topic. Shantell thought that this would be an interesting thing maybe to talk about.

Justin: For something that is so, um, uh, serious, it's certainly a fanciful name. Isn't it?

Sydnee: Tetralogy of Fallot?

Justin: It seems very fanciful to me.

Sydnee: If, if you break it down, what it means it doesn't, it's not—I mean, do you know tetralogy and what it's referencing?

Justin: Look at my eyes Sydnee. Look, look into our 11 plus years together.

Sydnee: You don't know.

Justin: Do you think I know what it's referencing Sydnee?

Sydnee: [laughs] Okay.

Justin: Tetra is five, right?

Sydnee: Four. [laughs]

Justin: Sooo, this is off the a great start. Excellent work everyone.

Sydnee: Uh, the tetralogy reference is for the four components of it, and Fallot is the person who called it a tetralogy, so there you go.

Justin: Mm-hmm.

Sydnee: That's the last name, Dr. Fallot.

Justin: Got it. Are you spoiling the story for me right now?

Sydnee: No, no.

Justin: Okay.

Sydnee: So, because the first thing I wanna tell you is what it is. Uh, as if you've watched the video, you may be aware that it is a—it's a congenital heart defect, meaning it is something that you are born with that is a malformation of the heart. It doesn't come together the way that traditionally does while you are in utero. It is a complex congenital heart defect, and it occurs in about one in two thousand newborns.

Justin: Do you mean complex to mean something clinical, or like…

Sydnee: Yeah, it's as opposed to—there are some simple heart defects. So, some people will say—uh, you may have heard this before, "I have hole in my heart," and there's a little teeny hole in between two of the chambers.

Justin: Mm-hmm.

Sydnee: That may not necessarily ever be clinically significant. It may just have been found and is there and may not do anything.

Justin: Mm-hmm.

Sydnee: Uh, this one's more complex.

Justin: Okay.

Sydnee: And it is the most common of the complex heart defects. About ten percent of congenital heart defects are tetralogy of Fallot. Fairly

common. Um, I have seen it. It's common enough that it—me and my—I'm still—I'm still pretty young.

Justin: Yeah.

Sydnee: I've had a few years of practice.

Justin: Yeah.

Sydnee: I've seen it. So, it involves—the four thing it involves, first of all, is a ventricular septal defect. Now, to, to describe these things just picture this.

Justin: Okay.

Sydnee: The heart has four chambers. Right? We're aware of that.

Justin: Yep.

Sydnee: There's two bottom ones, the ventricles. Two top ones.

Justin: The aorta!

Sydnee: Nope. Atria.

Justin: [laughs]

Sydnee: [laughs]

Justin: Okay. Alright. I'm doing my best here.

Sydnee: The uh—

Justin: I was so excited.

Sydnee: And the basic way the heart works. Do you know how the heart works? Do you know how it—what it does?

Justin: Um, yeah. It's like a big pump.

Sydnee: Uh-huh.

Justin: And the bad blood goes in, and the good blood comes out.

Sydnee: Okay, where does—okay, what's the bad blood?

Justin: Uh, blood that is not oxygenated.

Sydnee: Right, and it goes into…

Justin: The heart.

Sydnee: Which side?

Justin: The… right?

Sydnee: Yeah.

Justin: Crushed it!

Sydnee: And it—from the right side of the heart it goes to the…

Justin: Left.

Sydnee: Lungs.

Justin: [laughs]

Sydnee: And from the lungs it goes back to…

Justin: [sighs]

Sydnee: The heart.

Justin: The heart. The—

Sydnee: The left. The left side.

Justin: The left side of the heart.

Sydnee: Right.

Justin: Okay.

Sydnee: And from the left side of the heart, it gets pumped out through the whole body. Through the aorta, by the way.

Justin: Perfect.

Sydnee: Yeah.

Justin: I knew that was up in the mix. [laughs]

Sydnee: So, a ventricular septal defect means that there is a hole in between the two ventricles. So, the two ventricles are supposed to be separate. They have a septum between them.

Justin: Okay.

Sydnee: Separation. A wall between them and in this, the first defect is there's a hole there.

Justin: So, does that mean that the old blood can mix with the new blood?

Sydnee: Yes.

Justin: Okay.

Sydnee: Uh, then, the second part is hypertrophy of the right ventricle. That means is that the right ventricle gets big and thick. And it's not really supposed to be. The left ventricle is thicker normally, because it's gotta pump blood to the whole body, right? Well, the right side just has to pump blood into the lungs.

Justin: Mm-hm.

Sydnee: So, it's not as hard. So, in this case, the right ventricle is very big and thick.

Justin: Okay.

Sydnee: Because been working really hard to try to pump blood through the next defect, which is a—which is pulmonary stenosis. So, the pulmonary artery that comes out of the right side of the heart, just like the aorta does the left side. The pulmonary artery is tight. It's too tight in this condition.

Justin: Mm.

Sydnee: So, here's this ventricle trying to pump blood through it, and this is really tight, so the ventricle gets all big and beefy trying to pump blood through it.

Justin: Okay.

Sydnee: It makes sense?

Justin: Yep.

Sydnee: And then the last part is what we call an overriding aorta, meaning that the aorta now is actually getting blood from both ventricles. It's just supposed get it from the left, but instead, it's like crossing over and getting blood from the right side too.

And all this is really hard—I know this is hard when you're trying to visualize it. If you look at a picture of the heart – and this is really easy, you can Google a picture of tetralogy of fallout and that can kind of help to describe it. But if you imagine that what happened to the embryo, what happened developmentally that caused this, is that the division between the pulmonary artery and the… and the aorta just didn't happen in the right place.

Justin: Okay.

Sydnee: So it moved too far over, making this tiny little pulmonary branch, making this big large aorta, this hole between the septums. And the result is that, just like you said, you're not getting enough oxygenated blood and you're getting blood that isn't properly oxygenated pumped out through the body, and there's blood being shunted from the right side of the heart to the left side of the heart. So instead of going through the lungs, it's just being—

Justin: It's cutting the line, basically.

Sydnee: —shunted over back, and pumped back out into the body. We remember, by the way we learned a mnemonic for this in med school to remember the four different parts of tetralogy of Fallot, and it was IHOP. I just thought I'd share that. [laughs]

Justin: Oh.

Sydnee: I remember it that way. IHOP. Uh, so, like I said this causes a right to left shunt. All this unoxygenated blood.

Justin: What's the IHOP stand for?

Sydnee: IHOP stands for the intraventricular septal defect hypertrophy of the right ventricle overriding aorta, and then, pulmonary stenosis.

Justin: And it's just that easy?

Sydnee: And it's just that easy.

Justin: And now you at home can remember that entire sequence thanks to the acronym, IHOP.

Sydnee: [laughs] I remember it.

Justin: Perfect.

Sydnee: So, uh, so all this blood that is not oxygen rich is being sent through the aorta, uh, because it can't get through that pulmonic valve. And as a result, you're not getting enough oxygen to your body, and you become cyanotic, so you don't have enough oxygen. You're hypoxic, you know, you don't have enough oxygen.

Justin: Okay.

Sydnee: As a result, what would this look like? Well, babies can have a bluish tint, especially around their mouths. They can actually look kind of blue, because they're not—they're deprived of oxygen.

Justin: Right.

Sydnee: Um, which is one of the things Jimmy Kimmel mentioned. Uh, you may hear a murmur. A heart murmur is just the sound of blood moving through a valve, by the way.

Justin: Okay.

Sydnee: You're not—

Justin: The murmur is the sound?

Sydnee: Yeah. We're not supposed to hear the blood as it moves through the valve. We just kind of hear the valves opening and closing. If you can hear the blood move through the valve, it's called a murmur.

Justin: I always thought it was a hole in the heart.

Sydnee: No.

Justin: Okay.

Sydnee: No.

Justin: Bad job. Sorry.

Sydnee: You could have a hole in your heat, but that's not what it is. Uh, these babies might have difficulty doing things like breastfeeding for instance, because they tire out really easily while they're doing it.

Justin: Mm-hmm.

Sydnee: So, they might turn blue or even pass out. They can have these things that are called tet spells, where they do exactly that. They might cry or, even when they're having a bowel movement, they might turn blue and pass out.

Justin: Tet?

Sydnee: Tet. Tetralogy. Tet spells.

Justin: Oh, tet. I got it.

Sydnee: Yeah. It can also cause something called clubbing. It's not just seen in this particular disorder. Anything that results in hypoxia, so a lot of lung diseases, a lack of oxygen, can result in clubbing, which is this bulbous enlargement of the fingertips that's pretty… I mean, you'd noticed right away. That's a really old sign, by the way. It's maybe the oldest sign in medicine.

Justin: Wow, really?

Sydnee: They used to be called Hippocratic fingers, because Hippocrates talked about them, too.

Justin: Ah.

Sydnee: They're not just in tetralogy of Fallot, but that is one thing you can see them in. Um, so like I said, the whole thing has to do with early on in the development, the separation between the aorta and the pulmonary vessel do not form in the right place. All of this leads to, like I said, the enlarged right ventricle that isn't pumping or is trying to pump through this tight vessel.

Justin: Mm-hmm.

Sydnee: And it came to be related to certain genetic mutations. Um, there are certain syndromes. DiGeorge Syndrome is one, Down Syndrome can be related to this. Um, if mom gets rubella while she's pregnant, this can cause tetralogy of Fallot.

Justin: Would tetralogy of Fallot cause those things, or would they be an offshoot of those things?

Sydnee: They would be a—yeah. Yeah. Exactly. So, if you have somebody who has Down Syndrome or DiGeorge Syndrome, or mom had congenital rubella, um, advanced maternal age, or, um, drug or alcohol abuse. Any of these things, you have a higher likelihood of tetralogy of Fallot.

Justin: Mm.

Sydnee: And then sometimes, you just see it.

Justin: It just happens.

Sydnee: Yeah. Uh, as… now, as complex as all of this sounds, because it does, we have known about tetralogy of Fallot for over three hundred years. We've figured this out a long time ago.

Justin: Wow.

Sydnee: In 1673 the Danish anatomist, Steno, wrote the first description of tetralogy of Fallot. It was obviously based on an autopsy, and he noted all of the abnormal findings, and he basically said, "I wouldn't even attempt to tell you why this happened."

Justin: [snorts]

Sydnee: Which—

Justin: "Hey, listen."

Sydnee: [laughs]

Justin: "Listen. You're gonna hear a lot of people around my time period try to feed you a load of bull crap that they know what's happening, and I'm here to tell you… listen historians of the future – you are not going to look back and laugh at me. Not at Steno."

Sydnee: "I'm just telling you…"

Justin: "I'm telling you I don't know."

Sydnee: "Flat out. I got—"

Justin: "Here it is."

Sydnee: "I got no clue what's going on here." But he did—he did say this, "The baby was noted to have what is called a harelip." What was colloquially called a harelip.

Justin: What's the technical term for that?

Sydnee: Like a cleft.

Justin: Oh, cleft palate?

Sydnee: A cleft lip.

Justin: Cleft lip, okay.

Sydnee: The baby was noted to have what they were calling a harelip, and the mom had said, "I think this is probably because I was really craving rabbit stew while I was pregnant and I ate a lot it."

Justin: Mm.

Sydnee: And Steno was like, "Now, that sounds dead on."

Justin: Yeah.

Sydnee: Yeah. Definitely, that's why that happens. [laughs]

Justin: And this is why I'm not guessing about what is actually causing it.

Sydnee: [laughs] And this idea, just on a side note, I think we may have mentioned this briefly before, but this particular idea that things that mom did, saw, encountered, thought about, emotions that—that the pregnant person had during the pregnancy would have these kinds of effects on the baby. This was widespread, and even some of the, you know, way ahead of their time luminaries of, y'know, congenital heart defects and other congenital issues kind of had this concept that…

Justin: It's truly a pervasive idea even in modern age. Right? Like, you hear about women being told that they shouldn't go see scary movies.

Sydnee: Mm-hm.

Justin: Like, while they're pregnant, because they, you know, could have an effect on the baby.

Sydnee: Exactly.

Justin: Or frightening things, I should say.

Sydnee: Yeah, well and this is—that's exactly what they used to tell women is that, you know, if you're distressed in anyway, if you're putting any undue emotional or physical stress, so like hearing about something upsetting, reading a very thrilling book… You shouldn't read anything too exciting or thrilling. Uh, don't see a scary movie. Don't even think sad thoughts. Uh pregnant people were advised to basically like stay positive. Stay away from anything intense. Just be really cheerful all the time.

Justin: [snorts]

Sydnee: Rest.

Justin: No problem. No problem.

Sydnee: Rest a lot, and if you—except for housework, which is always great exercise for pregnancy. So pregnant, pregnant people were were advised to do lots of housework. [laughs]

Justin: Yeah.

Sydnee: That's very convenient.

Justin: Yeah.

Sydnee: Yeah. That's the work that's best for you, we found.

Justin: Yeah, just uh, keep on cleaning.

Sydnee: Is uh, doing the dishes and sweeping the floor.

Justin: Mm-hm.

Sydnee: Uh, in 1777, a Dr. Sandifort wrote a case for what he called a blue boy who had what he described as 'sinking spells,' and upon autopsy, was found to have these similar findings that we've already talked about. The tetralogy findings.

Justin: Mm-hmm.

Sydnee: And shortly after that, there was a Dr. Hunter, who wrote of a case of a boy who lived to the age of 13 with the same defect. He had the same sorts of spells, these what they would call sinking spells, basically just passing out. Which was becoming hypoxic and passing out, especially with exertion, so doing something stressful or demanding.

But he also seemed to have some growth issues as well. And he even—Dr. Hunter even theorized that maybe a lack of pulmonary blood flow, this tight pulmonic valve, maybe this has something to do with it. Um, and why you don't have proper growth and stuff. It was way ahead of his time, the idea that this has anything to do with anything. He—proposing this… right now, this seems really obvious, but proposing this at the time was kind of brilliant.

Justin: Yeah.

Sydnee: Uh, he also advised that while, again, he didn't know what caused it or what to do about it at all, definitely don't do any of the popular cures of the day like bleeding or purging or blistering or giving caustic agents or anything like that. So that was good.

Justin: That was good. He's on point. Good job, Dr. Hunter.

Sydnee: In general, in many of these cases, the patients didn't live very long lives. Um, if they made it out of infanthood, we have a lot of these, you know, like children and teenagers, and that was about as long as anybody was expected to survive with this defect. Uh, and they were weak and sickly throughout their lives. They were not able to, you know, go to school, or have a job, or get married, or have kids, or any of those kinds of things.

Uh, so what kind of advances did we start to make? So, this story gets a little happier.

Justin: Thank you.

Sydnee: In the 1850s, the stethoscope was still new and exciting, and Dr. Peacock used it to correlate various murmurs with different defects that

cause them, including being able to recognize a murmur that was associated with tetralogy of Fallot.

Justin: And I bet people who had them then all had like, cool moves that they did with them, like really getting showy with their stethoscope.

Sydnee: Like swing it around.

Justin: Swing it around. "Oh no problem, I can listen to your heart with my super hearing."

Sydnee: [laughs] Like, wrap it around your neck and swing it around like a hula-hoop.

Justin: Yeah. Exactly.

Sydnee: [laughs]

Justin: Very cool. Very hip look.

Sydnee: I do that now. Is that good?

Justin: Yeah, that's excellent, Syd. That's a good way to inspire confidence in your patients.

Sydnee: And it'll get me lots of friends. In the 1880s, we finally see the Dr. Fallot of renown. Dr. Fallot, who publishes this article where talks about—he has actually encountered numerous cases now of the tetralogy, and he writes up all these different studies, and what happened to them, and their clinical findings and upon autopsy and all this stuff. And he's the first one who dubs it the tetralogy, who actually says like, "If you find these four things, they go together. This is indicative of this disorder. We know what this is. It's called the tetralogy." And everybody was like, "Of Fallot, I guess."

Justin: Of Fallot, and he's like, "Well, I'm not crazy about it."

Sydnee: [laughs]

Justin: "But, so this is my… just wanna check, my legacy? Okay. Great."

Sydnee: Are you kidding me? No way. He was like, "Yes."

Justin: Yes.

Sydnee: "Yeah, of course. Well…"

Justin: You took the words right out of my mouth.

Sydnee: "Well, I mean, I don't mean to brag, but…"

Justin: If you insist.

Sydnee: "But I guess. "

Justin: I guess I do sort—

Sydnee: "I guess it kind of is my tetralogy." So—

Justin: Why not Fallot's Tetralogy?

Sydnee: I don't really know.

Justin: It's weird. It's usually—that's usually the structure that you see right?

Sydnee: There—but it sounds good. Tetralogy of Fallot.

Justin: And that has a good flow to it for sure.

Sydnee: Yeah. It's abbreviated well. I have TOF all through my documents. It's abbreviated well, TOF. Tetralogy of Fallot. In 1936, Mon Abbot, who was a famous Canadian physician, and which is really cool to talk about. Year 1936, Mon Abbot, who was female, was a famous Canadian physician expert on congenital heart disease, um, who published the Atlas of Congenital Cardiac disease, which was the bible of congenital cardiac disease, um, recognized in its time as one of the finest works anybody put together.

She actually, um, ended up pursuing her course of study in pathology and studying congenital heart disease and going down this road because she was denied the other internships, the other paths in medicine she wanted to take.

Justin: Hm.

Sydnee: Because of her gender. Uh, but…

Justin: Well, I guess that time, though…

Sydnee: [laughs]

Justin: Sexism worked out great for everybody. I guess this one goes to sexism. Right?

Sydnee: Yeah, I—

Justin: I guess it worked out pretty good that time!

Sydnee: Maybe some of us are just—were just gonna succeed no matter how hard…

Justin: Well…

Sydnee: … you, the man, try to hold us down.

Justin: Fair enough.

Sydnee: Uh, she p—so in it were drawings, clinical descriptions, EKG findings, chest x-ray findings. The heart looks boot-like on a chest x-ray if you have tetralogy of Fallot.

Justin: Oh, odd.

Sydnee: Uh, and pathology and all this together for tetralogy of Fallot, and this is really important because if you think about it, we're not—we're still not at a time where we can do what we would do now, which is an echo. The echocardiogram, which is like an ultrasound of the heart, which can show us all this stuff. We weren't doing all that yet, so any way you could diagnose it—

Justin: Right.

Sydnee: —was a pretty, pretty good idea.

Justin: Well we diagnose it, but I wanna fix it, Syd.

Sydnee: Well, I'm gonna—we're gonna—I'm not gonna do anything. Other smart people who could do surgery are gonna something about it. But first, why don't we head to the billing department?

Justin: Let's go.

[theme music plays]

[ad break]

[theme music plays]

Justin: You were gonna, uh, be a conduit for history as it affects the tetralogy of Fallot.

Sydnee: Right. I'm gonna tell you about the—this is the good part. So, the first—the beginning of this story actually starts with a successful surgery to fix something else. A different congenital heart defect, called a patent ductus arteriosus, that happened in 1938. Now, I really—I wish that—you know, I love podcasting, but this is an episode where, if this were a visual medium, it would be helpful.

So, there is a, something called the ductus arteriosus that you have when you are a fetus. It's a connection. It's a vessel that connects the aorta and the pulmonic vessel, and it is supposed to exist in the developing fetus.

Justin: Okay.

Sydnee: And then it goes away, because we don't need it anymore in our adult life. The reason is because it's… in the fetus, it's shunting blood back to the aorta and away from the pulmonary vessel, because it doesn't need to pump blood through those fluid filled, you know, amniotic fluid filled lungs.

Justin: Right.

Sydnee: But then after we're born, we need blood to be pumped through our no longer fluid filled lungs.

Justin: Okay.

Sydnee: So, this, this closes off itself. That's part of the process. It closes off. Just becomes a ligament. A remnant that's left there. Now, this doesn't always happen. Sometimes things go wrong.

Justin: Sure.

Sydnee: And so sometimes, this little connection between these two vessels is left open. It causes a machine-like murmur.

Justin: Mm. What's that mean?

Sydnee: It sounds like machinery.

Justin: Oh.

Sydnee: Like a grinding machine.

Justin: Wow. Okay.

Sydnee: If you hear it. Anyway, it—so, surgeons were trying to figure out, "How can we close this thing? We need to know how to close this thing, because it shouldn't still be open. About a week after birth, you know, at the latest, it closes. It shouldn't be open after that. We need to figure out how to close it."

So in 1938, they figured out how to just ligate it and close it. And this was a big success, because we hadn't been doing surgeries on hearts, especially baby hearts, so this was, you know…

Justin: Yeah.

Sydnee: A huge deal. So, because they fixed that, this inspired somebody. So, this is Dr. Helen Taussig, who was working in the cardiology clinic at Johns Hopkins, who was a brilliant, maybe the most known pediatric cardiologist in history. Definitely one of the most known.

So, she began to wonder about a lot of the children she was seeing in her clinic with congenital heart defects, and whether there would be some surgery that might help them as well, because that seemed like the only approach to this. It's gotta be a surgical solution.

Justin: Yeah. Right.

Sydnee: There has to be a surgery here, right?

Justin: Yeah.

Sydnee: Um, she also, by the way, had worked there for some time, because she was denied a spot in the medical internship she wanted, because they would only allow one woman on the house staff at any given time.

Justin: Wow.

Sydnee: And she was beaten out for this spot by two points by another woman. So anyway, she devoted her life to this cardiology clinic, and to these children, and it was a wonderful thing that she did. Because she started to ask the question, "If we can close a ductus, that little connecting vessel in the heart…"

Justin: Sure.

Sydnee: "If we can close it… could we not build one?"

Justin: Like a, like—okay.

Sydnee: "Can we not construct a duct? We can close a duct. Could we not construct a duct?"

Justin: Construct a duct is more fun to say.

Sydnee: [laughs]

Justin: Certainly, we should be able to.

Sydnee: She actually went to Dr. Gross, who was the doctor who had done this procedure in 1938. The surgeon who had done it. She went to him and said—

Justin: That's just an unfortunate name for a doctor.

Sydnee: Dr. Gross.

Justin: Dr. Gross is rough. That's a rough one.

Sydnee: [laughs] She went to Dr. Gross and—

Justin: He was a surgeon?

Sydnee: Yeah.

Justin: Oh man. Ah Dr. Gr—my heart goes out to you my friend. That's a tough putt.

Sydnee: [laughs] She went to Dr. Gross who had just done this, this successful procedure and said, "Hey!"

Justin: You gotta work really clean.

Sydnee: "Ca-" [laughs]

Justin: That's the thing. If people come in to sit on operating table and they're like, "Oh man, there's guts everywhere.' They're gonna be like, 'And not just in name," And I'd be like, "I get it. I've heard it my entire life. Okay, I'm a messy guy. It has nothing to do with my name." I'm just saying, you gotta work really clean.

Sydnee: But the—that's true.

Justin: I'm done talking about Dr. Gross.

Sydnee: [laughs] You done with his name?

Justin: Yeah.

Sydnee: Well-

Justin: I'll be looking in your nose today. My name is Dr. Boogers. [laughs]

Sydnee: [laughs]

Justin: I'll be your ENT.

Sydnee: You're—you're not gonna be done, because she went to Dr. Gross and said, "Hey, you figured out how to do this and it's amazing. Do you think you can do the opposite? Do you think you can build a duct?" And he said, "Yeah, definitely. Like, for sure. No problem. I could. I'm not going to. Not really feeling that right now. Everybody's super excited about how I closed that other duct…"

Justin: Mm-hmm.

Sydnee: "So, that's kind of where the heat is. I'm not feeling—"

Justin: Sure.

Sydnee: "I'm not picking up what you're putting down."

Justin: Right. There's still heat behind making ducts.

Sydnee: [laughs] So—so he said no. So she went back to Johns Hopkins and she bided her time because she knew that another surgeon, Dr. Alfred

Blalock, along with his assistant Vivien Thomas, were coming to work there, and she thought, this is a guy that I'm gonna talk into the surgery, because I know this surgery—I know that this is gonna mean something.

So, she told him all of her observations on congenital heart defects and what she's seen and what she thinks… that this might work. So, this was her theory on constructing a duct.

Justin: Okay.

Sydnee: She noticed that the worst symptoms started about a week after birth.

Justin: Hm.

Sydnee: When that thing I told you about, that ductus arteriosus, typically had closed. So, what she was thinking was that that was allowing some of the blood to go back. Here all this blood is being shunted to the body and not making it through the lungs.

Justin: Mm.

Sydnee: This open little—this open tunnel was like a back channel. It was allowing blood to go back through the lungs.

Justin: Yeah. I, I—

Sydnee: Right?

Justin: I understand. Yeah.

Sydnee: So, if it stayed open, maybe we can keep allowing blood to go back through the lungs.

Justin: Unless it—

Sydnee: You get more oxygen in the blood. And you get more oxygenated people.

Justin: Right. Good, I mean you could stop at more oxygen in the blood.

Sydnee: Right.

Justin: We know that's the goal here.

Sydnee: Which is good. Uh this wouldn't fix the problem, but kind of palliate the problem somewhat. So, make things better.

Justin: Thank you.

Sydnee: Yeah. So, she, you know, pulled all this together, presented him with all this information and uh, he said, "You know what? This sounds reasonable. I'm gonna have to test out your theory for a while." So, he attempted it a lot on some animal models. We won't go into detail there.

Justin: Thank you.

Sydnee: Yeah. And after two years, he decided, "You know what? I think I can do this surgery. If you think this surgery can work, let's go for it."

Justin: All right.

Sydnee: So in 1944 they started doing the procedure to connect this subclavian artery, which arises from the aorta, to the pulmonary artery. So not exactly replicating the natural ductus arteriosus, but inspired by a similar concept.

So, they started doing these procedures, and they worked. So, imagine with me, if you will, the moment. It's 1947, and these two doctors, Dr. Blalock and Dr. Taussig, are giving a lecture in the big giant hall at the British Medical Association in London.

And they're talking about the procedure, and they're showing diagrams, and they're going through the intricacies of the surgery, and how like, with each procedure, they alter it just a little to make it a little better, and why the results got better with each one, and all this stuff. They're telling this story, and then at the climax of the story, a spotlight appears on this gorgeous little two and a half year old, curly hair, pink cheeks, looks healthy as can be, and they said, "And here is a surgery we did last week. And here stands this healthy baby girl that, you know, would never have been this healthy to just be here with us today. And a week ago, we did this surgery on her."

Justin: Since the drama is so important when you're presenting this stuff, you're gonna get people hyped up. That's very good.

Sydnee: I don't usually—

Justin: They should let—they fly her in from like the… put her on wires and just like fly her in. Somebody shoots some sparklers off. Stuff like that.

Sydnee: [laughs] She actually, in one of the articles I read, was described as cherubic.

Justin: Cherubic.

Sydnee: Cherubic.

Justin: That's adorable.

Sydnee: Uh, so the kids did oxygenate better. They had less, you know, problems with turning blue and passing out and all that. And they could play, and this sounds like a dumb thing to say, but these families had kids who could go play again. Who could run around outside, who could play games, who weren't stuck on the couch all day. And the medical community just was on fire, so—

Justin: And that's before TV, so it really was sad back then.

Sydnee: [laughs]

Justin: If you think about it.

Sydnee: So, by the 1950s—

Justin: I can understand while you're getting teary eyed, because like, and what did they watch? Because that's what I'm—

Sydnee: Their kids could play!

Justin: And that—and not sit on the couch and not watch any TV.

Sydnee: Some of these kids were like 11 years old, and for the first time in their life, they were able to run around and play.

Justin: 11 years of not watching TV. I mean, can you think about it?

Sydnee: [laughs] So by the 1950s, uh, different procedures were tried, and there actually were some open-heart procedures done. You know, it took us a while to get to the point… obviously, anesthesia was a big part of this too, to get to the point where we could, um, open people up and fix problems.

Justin: Sure.

Sydnee: You know? For longer periods of time, more advanced problems, and they eventually figured out how to repair the defects, closing the hole, and um, opening up the pulmonary vessel and reducing the size of that right ventricle, and all that kind of stuff. And throughout the ‘70s, procedures became better, and of course, as I already alluded to, once we had the echocardiogram where we could ultra sound and look at the heart ahead of time, we didn't have to guess about what we were getting into. We could, you know, noninvasively know exactly what the structure of the heart was. Which was obviously a big advantage, and we can do that in utero. Right?

Justin: Yeah.

Sydnee: We can see that in utero.

Justin: Mm-hmm.

Sydnee: While you are, while you—a patient is still pregnant, we can look in at the developing fetus and look for heart defects.

Justin: Sure, yeah.

Sydnee: Uh, so, now we do surgery. Uh, we don't typically do that shunt that I described. The Blalock-Taussig shunt. Taussig-Blalock shunt. I forget which direction it's in. Anyway, they shunt that’s named from them. We don't typically do that. You can still do procedures like that. Shunts sometimes are still done if, in certain patients where the defects in so severe that you kind of have to do it stepwise, tou need to do something immediately to help get oxygen into the blood and stabilize the patient, and then, down the road, you'll do the complete fixing of it, the complete corrective surgery.

Justin: Mm-hm.

Sydnee: But most people now end up, sooner or later, getting the complete corrective surgery, usually within the first year of life. Um, there are patients that, as recently as the ‘90s, who got it later than that.

Justin: Hm.

Sydnee: But usually within the first year of life. And like I said, it could be one surgery, it could be a series of surgeries, depending on exactly what type of… you know, the defect isn't exactly uniform in every single person.

Justin: Of course. Yeah.

Sydnee: It's a little different. Um, untreated, tetralogy of Fallot used to have a 35% mortality rate in the first year of life, and 50% mortality in the first three years of life.

Justin: Ugh.

Sydnee: And as I already mentioned, most people were not living out of their teens, in the majority of people. Now, early mortality is less than five percent. People can live long, healthy lives. Uh, they do need monitoring, and sometimes other complications down the road can occur, but a lot of patients have the surgery, have, you know, everything corrected, and never have problems from it. As an example, Shaun White, the snowboarder, had tetralogy of Fallot repaired.

Justin: America's favorite only snowboarder. [laughs]

Sydnee: It's really fascinating. I read the… [laughs] I read the paper by Helen Taussig where she was kind of describing like, one of many… I mean obviously, she—they wrote about this a lot, you know. If you developed this procedure, you're gonna write about it a lot.

But in one of her kind of uh, going back and thinking about the whole—her whole course of everything, how she started there and working at the cardiology clinic, and figuring out the surgery and everything and she was talking about, that they went back to see like how many of their patients down the road, how long did they live, did they have other problems, but also, did they get a job? Did they get to finish school? Did they get married, did they have kids? and talking about the like, three hundred and some children that resulted from all these lives that were able to be lived, you know, as a result, and it was…

I mean, it's amazing to read about, because they just kind of said, "Hey, I think this will work," and tried it out, and did. Um, she also mentions specifically, towards the end of the paper, she says, "To the best of my knowledge, no patient was ever refused because he could not pay. Further, the hospital established the policy that 'cyanotic patient'…" so somebody comes in blue, "… no cyanotic patient who arrives at the doors of the hospital seeking help should leave without seeing me." Um, so you were never turned away.

Justin: Mm.

Sydnee: And it didn't matter if you could pay. And she also said, "That this study indicates that a handicap in childhood does not preclude success in adult life. On the contrary, a handicap may act as a stimulus to do one's best." Which I thought was a really, really inspiring way to look at it.

Justin: Well, I'm glad that we got the—that was harrowing. I would describe that as harrowing. [laughs]

Sydnee: [laughs] That was harrowing?

Justin: I would say that was harrowing. It was very—it was very daunting and little upsetting at the beginning, and then things got better towards the end so thanks. And that's great story telling.

Sydnee: [laughs]

Justin: And history apparently so rarely works in that fashion, so…

Sydnee: It wasn't me. It wasn't me.

Justin: Imagine my relief.

Sydnee: Thank these brilliant physicians who came up with it. And especially, it's nice to hear a story like this set in a time period where uh, so many female physicians played a role.

Justin: Yeah. Absolutely.

Sydnee: Uh, big—a big role and you know the amazing thing is, this isn't something that has to devastate families anymore.

Justin: Mm-hmm.

Sydnee: It doesn't have to, because we can fix it, and I don't understand a world that would deny anybody this life-saving procedure, no matter what the cost, ever.

Justin: Folks, thank you so much for listening to our show. We hope you have enjoyed it. If you did, please leave us a rating or review on iTunes, or you know, just tweet about this show. Say, "Hey, um, listened to this show. You know that thing you heard about on Jimmy Fal, Jimmy—not Jimmy Fallon. Jimmy Kimmel."

Sydnee: Yeah.

Justin: Why do we have two Jimmies?

Sydnee: [laughs]

Justin: Uh, Jimmy Kimmel. Here's more information about it. It's got a fascinating history.

Sydnee: And say the name of our show, Sawbones.

Justin: Sawbones, and then my name, and then Sydnee's name after it.

Sydnee: [laughs] You can just leave my off, Justin's the real star.

Justin: Shut up.

Sydnee: [laughs]

Justin: Um, and thank you to Taxpayers for letting us use their song 'Medicines' as the intro and outro of our program. Uh, thank you to the Maximum Fun network for having us as part of their extended podcasting family. And folks, that is gonna do it for us.

Actually, no wait. You know what? We have a P.O. box where people send us stuff sometimes. P.O. box 54, and it's West Virginia 25706. And uh, I want to say thank you so much to Christina for the lovely book that is currently sitting on our coffee table.

Sydnee: Ah, it's a great book.

Justin: And Cecil sent us a scarf that they made, and Cecil knitted us some snakes.

Sydnee: Yes.

Justin: Like in our logo. They're awesome.

Sydnee: They're adorable and Charlie loves the scarf.

Justin: Um, and that is going to do it for us, folks. So, until next week, my name is Justin McElroy.

Sydnee: I'm Sydnee McElroy.

Justin: And as always, don't drill a hole in your head.

[theme music plays]

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