Sawbones 130: Hemophilia

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Intro (Clint McElroy):

Sawbones is a show about medical history, and nothing the hosts say should be taken as medical advice or opinion. It's for fun. Can't you just have fun for an hour and not try to diagnose your mystery boil? We think you've earned it. Just sit back, relax, and enjoy a moment of distraction from that weird growth. You're worth it.

[theme music plays]

Justin:

Hello, everybody. Welcome to Sawbones, a marital tour of misguided medicine. I'm your co—host Justin McElroy.

Sydnee:

And I'm Sydnee McElroy.

Justin:

I still don't sound normal. It's been six weeks since I had a cold and I still can't snap back from it. What's up, Syd?

Sydnee:

You're fine.

Justin:

I mean, I'm fine. I'm fine.

Sydnee:

I mean, you just talk a lot.

Justin:

That is true. I can't give my instrument the time it needs to recuperate.

Sydnee:

You call it your instrument. That's your real problem is you call it your instrument.

Justin:

I was especially taxed during the Max Fun Drive, uh, which, uh, we... 'cause we recorded a lot of extra stuff and extra long episodes. And uh, I just want—

Sydnee:

[mockingly] "I had to do so many podcasts."

Justin:

Oh—

Sydnee:

"It was so hard."

Justin:

Eat my butt—

Sydnee:

"It's what I love doing. And everybody..."

Justin:

Eat my—

Sydnee:

"...loves me for it."

Justin:

Eat my butt, Smirl.

Sydnee:

"It was so difficult to do!"

Justin:

Kiss my grits. Um, no I want to thank everybody for being so generous during the drive we had, like, 9,400, uh, new and upgrading donors to the Maximum Fun network.

Sydnee:

Yes. Thank you all so much.

Justin:

So much.

Sydnee:

Your support means everything to us. We really appreciate it.

Justin:

And because you all were so good to us, I am positive, uh, Sydnee assures me that we are going to have, uh— Last week. It was kind of a lot of poopy talk and um...

Sydnee:

I didn't realize how much poop would bother everybody.

Justin:

Mm-hmm, yeah.

Sydnee:

Uh, I thought— I mean, I know it's something that, like, I understand that it's gross, inherently.

Justin:

Hmm.

Sydnee:

Like I get that.

Justin:

Yeah.

Sydnee:

But I kind of thought it, it's one of those things that, like, when you're a little kid you like to talk about poop and then as you're an adult, you pretend like you don't like to talk about poop, but everybody secretly does. That is not the case. You were all...

Justin:

Yeah.

Sydnee:

Not you were all, but a lot of you were a little unsettled.

Justin:

We were all put out by it now.

Sydnee:

By it. And I am sorry for that.

Justin:

So this week Sydnee promises that she's got something a lot more, uh, less squirm inducing.

Sydnee:

Yeah.

Justin:

Uh, a lot more relaxed, uh, sort, just a light topic. So what do we got Sydnee?

Sydnee:

Absolutely. Uh, there are a lot of people who have been asking for me to talk about this for a while. And so, um, and I thought, you know, following on the heels of poop, what could be easier for everyone to deal with than blood?

Justin:

Right.

Sydnee:

So, I thought we would talk about hemophilia this week.

Justin:

You don't actually think that's...

Sydnee:

Uh, because—

Justin:

You don't actually think that that's, like, okay.

Sydnee:

What do you mean?

Justin:

You don't actually think that that's okay. Like, I know we all have a lot of fun here, but you don't think blood's, like, another cool topic. Like, you know, we go to say okay, chilled out topic.

Sydnee:

Well, it's not— I mean, there's no, like, there's no fecal material in it. I mean, there shouldn't be, hopefully there aren't, like, there's not—

Justin:

I—

Sydnee:

Inherently there are bacteria in it. I mean, unless there's a problem. Like, it's a very— It's, it's clean, it's sterile.

Justin:

I under—

Sydnee:

It's just blood. It's, like, it carries oxygen and it's got all kinds of...

Justin:

Okay—

Sydnee:

Good cells in there that protect you from infection—

Justin:

Mm-hmm.

Sydnee:

And clot you and you're... well, hopefully when you have a cut, I mean, like, it's great. Blood's great—

Justin:

You, you—

Sydnee:

Is blood—

Justin:

Okay—

Sydnee:

Is... wait, is blood gross?

Justin:

I understand that you're a physician. And like... but you are also— You have been swapped out with, like, kind of a pod, some kind of pod person. Like you still have human emotions and stuff, right?

Sydnee:

I do— Does blood... Is that a bothersome thing?

Justin:

Sydnee, this is not... I— You're scaring me right now. Uh, you don't actually think this, right?

Sydnee:

I really don't understand why blood is upsetting.

Justin:

You know it, but you—

Sydnee:

I mean, other than, like, if it was, like, I mean—

Justin:

Like, you know it is for some people, right?

Sydnee:

It is in the emotional sense that, like, I don't want people to bleed because that's usually problematic. And so I am worried about their wellbeing or, or my own, certainly if I am bleeding. It's gross though. Right? Like that's I'm—

Justin:

Yeah, it's gross—

Sydnee:

What I'm gathering from you.

Justin:

Yeah. It's gross—

Sydnee:

It's gross too.

Justin:

Um, why don't you, uh, why don't you. I'm sorry—

Sydnee:

I know some people pass out with blood. I just, I guess I didn't think of it. I don't know. Okay.

Justin:

I'm sorry. Well, let's—

Sydnee:

I see where I've gone wrong—

Justin:

Let's, let's keep trucking [laughs].

Sydnee:

All right. Well, I— A lot of people have requested this topic. Thank you, Andrew and Lauren and Syd, not me. Another Syd.

Justin:

That would be weird.

Sydnee:

Yeah. Not, not—

Justin:

You're Sydnee—

Sydnee:

Thanking myself.

Justin:

It's you.

Sydnee:

No, Syd. And Sarah and Jolene and Diane and Mike, thank you all for recommending this topic. Um, because it's very interesting and there's a lot of history and I'm sorry, I guess it is somewhat upsetting.

Justin:

Yes.

Sydnee:

For some people. Well, let's talk about it anyway.

Justin:

Okay.

Sydnee:

Do you know what hemophilia is?

Justin:

Uh, it's when you bleed a lot and can't clot properly.

Sydnee:

That's pretty good.

Justin:

Thanks.

Sydnee:

I'm impressed.

Justin:

Thank you.

Sydnee:

I think most people have, like, a passing understanding of hemophilia.

Justin:

Oh. Cool. Kind of like a... kind of, like, a compliment and then diminish me, like, in the same breath. That's pretty cool.

Sydnee:

[laughs] I didn't mean to.

Justin:

No, keep going. That's good.

Sydnee:

Okay. Sure. Uh, most people are aware that it is a bleeding disorder. Um, but it... it's actually kind of complex as to what causes it. I mean, why?

Justin:

Okay—

Sydnee:

I mean, I'm guessing you probably don't understand, like, what's... what has gone wrong that someone has...

Justin:

Probably some—

Sydnee:

...bleeding issues.

Justin:

Probably some blood cells not doing something properly. Right?

Sydnee:

Sorta. Yeah. Sorta like that. So our blood clots and it's important that our blood clots. Right?

Justin:

Right.

Sydnee:

Because... not just because we accidentally bleed from time to time, you know, when we get, um, cut or when we fall and scrape something or if something, you know, is going wrong in our colon or in our bladder or something, we bleed for lots of different reasons. Uh, but we also sometimes need surgery.

Justin:

Right.

Sydnee:

Right?

Justin:

Right.

Sydnee:

Sometimes we wanna get something pierced—

Justin:

And we need—

Sydnee:

I don't know—

Justin:

We need to get better—

Sydnee:

Or get a tattoo.

Justin:

We need to get—

Sydnee:

There are lots of reasons why we would need our blood to clot. And there are lots of ways that our blood clots, there are lots of different factors involved.

Justin:

Okay.

Sydnee:

We call them clotting factors. So that's convenient.

Justin:

Mm-hmm. yeah.

Sydnee:

There—

Justin:

It's kind of, like, works on two levels.

Sydnee:

There's a whole cascade of them that takes place. If you were to get cut, a whole, like, domino effect of factors that get activated, we call that the clotting cascade or coagulation cascade. And then there are other things involved. Things like platelets, you've probably of, things called, like, fibrin and plasmin.

And, uh, there are lots of different parts of your blood that are involved in clotting. The problem in hemophilia can actually be different. When we say the word hemophilia, we can mean different things. Um, for instance, let's say that, uh, you have hemophilia A—

Justin:

Okay—

Sydnee:

Because there are different types.

Justin:

All right.

Sydnee:

What that means is that you're missing factor VIII.

Justin:

Okay.

Sydnee:

So that's one of the factors in the clotting cascade. This is why the disease was tricky to treat for a long time. Because if you have hemophilia A, you've got the other parts of the clotting pathway, you don't have factor VIII.

Justin:

And is it, like a, I mean, is domino an applicable metaphor here, in that, if you take one out, like the whole thing breaks down?

Sydnee:

Others won't be activated. Yes.

Justin:

Okay.

Sydnee:

And there are... I mean it is, if you look at the clotting cascade, it's not a direct domino, it's almost like one of those really fancy domino setups with like a web where there are different dominoes that could hit one domino.

Justin:

Okay.

Sydnee:

So it is not as simple as to say, if you're missing one factor, it just stops in the chain. There are other things involved and kind of other side pathways that can happen. But each factor is absolutely very important.

Justin:

Okay.

Sydnee:

And some are more important than others and factor VIII is particularly important for its place in the cascade.

Justin:

Okay.

Sydnee:

Uh, similarly, if you have hemophilia B, you're missing factor IX, and that is also very important in the clotting cascade. Um, so different types of hemophilia equals missing different clotting factors. Does that make sense?

Justin:

Yeah, for sure.

Sydnee:

Uh, factor VIII deficiency or hemophilia B by the way is also called Christmas disease.

Justin:

Oh, festive.

Sydnee:

So if you ever hear that, that's what they're talking about. It's a type of hemophilia—

Justin:

Did we talk about Christmas disease once?

Sydnee:

We didn't, we talked about discussing Christmas disease for our Christmas episode.

Justin:

Yes. Right.

Sydnee:

And then we didn't, but um, it's actually not named for the season [laughs] or the holiday.

Justin:

It's been named for Herb Christmas,

Sydnee:

Stephen Christmas.

Justin:

Oh,man.

Sydnee:

Yeah [laughs]. Good—

Justin:

All right.

Sydnee:

Good guess. But it was named for Stephen Christmas, who was the first person diagnosed officially with hemophilia B back in the fifties—

Justin:

The first person— Not the person to discover it, the first person to, like, discover it, the first person to get it?

Sydnee:

Yeah. Well, no, he wasn't the first person to have it. Lots of people had had it—

Justin:

If— Be diagnosed with it—

Sydnee:

But the first person to actually, officially be diagnosed with hemophilia B was Stephen Christmas. And so—

Justin:

And he had to go and ruin—

Sydnee:

Christmas disease.

Justin:

He just ruined Christmas, just like that. You ruin the whole world. I mean, at least—

Sydnee:

The Stephen who stole Christmas.

Justin:

At least his name wasn't Steven Pizza. That would've been bad.

Sydnee:

[laughs] Pizza disease?

Justin:

Pizza disease that would've been so tragic—

Sydnee:

I call that heartburn—

Justin:

[imitates a trumpet] Fart noise. Okay, go ahead.

Sydnee:

[laughs] Um, it is... hemophilia is an X-linked disease. Do you know what that means?

Justin:

Absolutely not.

Sydnee:

Justin. You have, yeah, you had some genetics—

Justin:

Yeah.

Sydnee:

At some point in science class.

Justin:

Oh yeah. Does that mean it affects mainly men?

Sydnee:

You read ahead.

Justin:

I did [laughs].

Sydnee:

Okay. Uh, uh, sure it does, Justin—

Justin:

You put me on the spot—

Sydnee:

Could you—

Justin:

You could have just let my ignorance lie, but you had to put me on blast and, like, really do that thing that I love where you're, like, "Are you sure you're a dummy?" And I had to be, like, "No, no, no. I am for sure a dummy." So I got defensive. I got... you know what? I don't have your knowledge base, but you know what I do have?

Sydnee:

What's that?

Justin:

Savvy.

Sydnee:

Okay, Justin. So, you know, it largely affects men—

Justin:

[crosstalk] ...over here reprogramming the whole simulation.

Sydnee:

Sure. Why would an X-linked disease largely affect men?

Justin:

Because men have XX.

Sydnee:

Nope.

Justin:

Men have XY—

Sydnee:

Back, backwards, backwards. Okay.

Justin:

Men have XY.

Sydnee:

Anyway, when we're talking, uh, purely, and again, this is purely based on, uh, genotype. We're referencing kind of some gender terms—

Justin:

Sure, men are XX. Men are XY, men are—

Sydnee:

Okay, if we are talking genotypically male—

Justin:

Yeah. Sorry. Yes. I should have been more— Genotypically male—

Sydnee:

Yes, genotypically male, XY.

Justin:

Okay.

Sydnee:

Genotypically female we say is XX.

Justin:

Got it.

Sydnee:

Okay. Got that.

Justin:

Got it.

Sydnee:

So we're at— We're talking purely about sex chromosomes at this point. Um, and not the larger issue of gender, but, uh, an X-linked disease means that the gene, the problem gene, that that's causing hemophilia is on the X chromosome. So a woman, genotypically, has two X chromosomes.

Justin:

Mm-hmm.

Sydnee:

Whereas a male would have one.

Justin:

Okay.

Sydnee:

So if you're getting your X chromosome from your mother, which you do, right? If you're a— If you are born a male—

Justin:

Mm-hmm.

Sydnee:

You get an X chromosome from your mom and a Y chromosome from your daddy.

Justin:

Right.

Sydnee:

Okay? You can't get a Y chromosome from your mom 'cause—

Justin:

She don't have one—

Sydnee:

She doesn't have one. Got it. Okay. So if you, you only have one X, so if you get the affected chromosome from your mom, that's it, you've got hemophilia.

Justin:

Okay.

Sydnee:

Because you only got one X, whereas a female gets two Xs. So she's gonna get an X from her dad who either has hemophilia or doesn't and then an X from her mom where she's rolling the dice. One of those Xs has hemophilia, the other one doesn't.

Justin:

Mm-hmm. okay.

Sydnee:

So a woman is going to probably not be affected because she's got a good gene on the other X chromosome.

Justin:

Okay. All right.

Sydnee:

So the only, the only way a woman could be affected is if both mommy, daddy give her an affected X.

Justin:

Okay. Got it—

Sydnee:

And the statistics on that are just low.

Justin:

Okay. Interesting.

Sydnee:

Does that make sense?

Justin:

Yes.

Sydnee:

Okay. You don't get it, do you?

Justin:

I do. I do get it.

Sydnee:

I'm gonna—

Justin:

I'm like—

Sydnee:

Draw you some Punnett squares later.

Justin:

No, I'm like Gregor Mendel over here. I got it.

Sydnee:

This dates back to ancient times, most likely even though we don't—

Justin:

Hemophilia you mean?

Sydnee:

Hemophilia does, even though we don't see, like, the word hemophilia dating back to ancient times. But there's some hints of it. For instance, you know, circumcision is something that has been practiced, um, in the Jewish faith for a very long time.

And um, there are provisions specifically that you can find throughout, um, kind of ancient history and in the Talmud where they say, if you have two male infants, like two brothers who are, who both died during circumcision—

Justin:

Mm-hmm.

Sydnee:

Then the third— And then you have another male in that family, they actually do not have to go through circumcision. There's actually provisions that...

Justin:

Oh—

Sydnee:

...say you can forgo it. The thought process being that if two boys in the family had bleeding problems to the extent that they died following circumcision, the third may as well. And this is kind of an understanding of a genetic disease, even though we didn't understand what it was yet, you know?

Justin:

Yeah. I think it's weird that it takes two. Like, for me as a parent, if my first kid beefed it after he got circumcised, I would be like, "You know what? Let's leave it on. We're good."

Sydnee:

Well, I mean, it may have just been, you know, may have just been a chance at that one.

Justin:

Yeah. You know what? I'm not a risk taker. After my first kid did that whole thing. I'm just gonna go ahead and stick with... Stick that foreskin right on there.

Sydnee:

I think that's fair. I think my scientific inquiry probably ends where my child begins.

Justin:

Right, right, right.

Sydnee:

For me personally.

Justin:

Yeah.

Sydnee:

Um, there is mention in the Bible of a woman who had hemorrhages for, like, 12 years and then Jesus healed her of those hemorrhages, just these bleeding episodes. So that could have been a reference to hemophilia. 'Cause let me mention hemophilia can affect women. It's just much more common in men—

Justin:

I got it, no, I got it.

Sydnee:

Um, and there was, uh, Albucasis who was an Arabian physician from the 10th century who wrote kind of these, like, pedigrees, these histories of families, where there were men in them who bled to death and nobody knew why. So people sort of understood that there was this genetic bleeding disease, but nobody knew why it was happening or exactly how it was passed from person to person. And it would seem to skip generations and nobody knew why.

Justin:

Okay.

Sydnee:

Now before I get into how we figured all this out and some of the crazy things we tried to do for it before we knew what to do for it, um, I think this is a good moment to talk about why hemophilia is known sometimes as the royal disease or the disease of kings.

Justin:

Okay.

Sydnee:

Um, you may know this Justin, but hemophilia played a major part in the royalty of Europe.

Justin:

Yes, I did—

Sydnee:

Have you—

Justin:

Yes.

Sydnee:

Have you heard that reference?

Justin:

I don't know how, but yeah, I know basically.

Sydnee:

So I'm gonna try to walk you through some of this 'cause there's some interesting facts, but let me recommend, um, that if you are really interested in understanding it, right now you go Google, like, European, Royal family hemophilia pedigree. Something to the, some mixture of those words.

Justin:

Okay.

Sydnee:

You know, hemophilia, pedigree, Royal family, something like that. Pedigree is like one of those charts of inheritance.

Justin:

Does this happen on Stuff You Missed in History Class? They say like, "Listen guys, we're gonna try our best here. But uh, just, just Google it."

Sydnee:

I just think this is something that... no, I think if you could see it, if you can see the family tree, it's easier to follow.

Justin:

Yeah.

Sydnee:

'Cause I'm gonna give you a lot of names and who had hemophilia and who didn't and I think if you can see the little boxes and they're like, "This one is shaded for a carrier and this one is colored this color if it's a person with hemophilia." I think it's easier to see.

Justin:

Okay. Got it. I'm with you.

Sydnee:

So Queen Victoria of England who reigned from 1837 to 1901—

Justin:

So I remember from the Doctor Who episode with werewolves.

Sydnee:

Perfect. Okay. She was a carrier for the hemophilia gene. Okay?

Justin:

Okay.

Sydnee:

So she did, she was not a hemophiliac. She did not have hemophilia, but she had it on one of her X chromosomes. Right?

Justin:

Mm-hmm. right.

Sydnee:

So she carried it and could pass it along to her children.

Justin:

With you.

Sydnee:

Which she did. Uh, so let's start talking about some of her children, we're gonna talk first about Leopold. Leopold was her eighth child, her fourth son. And he had hemophilia. He had hemorrhages throughout his life. You can get spontaneous bleeding, not just like when you get cut, you bleed a lot or you can get a nose bleed that won't stop. You can get spontaneous bleeding in your joints. Um, worst case scenario in your brain.

Justin:

Oh.

Sydnee:

Uh, so yes, it can be a very crippling—

Justin:

Woof.

Sydnee:

Uh, dangerous disease, or at least at that point in time, it could have been.

Justin:

Mm-hmm.

Sydnee:

And it was for Leopold. He died from a brain hemorrhage as a result of it. Uh, but before he did, he had some kids, which is important. Um, on a side note he had a couple different doctors. One of them was Jenner. You've probably heard of Jenner—

Justin:

No.

Sydnee:

Like, Dr. Jenner. Okay.

Justin:

No.

Sydnee:

He studied him. And, uh, and he came up with a theory based on studying Leopold that hemophiliacs just had more blood than the average person.

Justin:

They had to let it off, like, let off steam. Yeah.

Sydnee:

[laugh] And they had really small blood vessels. So it just kind of spilled out all the time. Uh, that's not true. Um, his second doctor Dr. Legg thought that, uh, staying in warm climates would help, which is why Leopold spent a lot of time where it was warm. Um, he actually, uh, died in Cannes, in France.

Justin:

Mm-hmm.

Sydnee:

Um, but uh, strangely Legg also advocated in papers that he wrote on the subject that men with hemophilia not be allowed to marry or have kids, which is weird because Leopold did both of those things.

Justin:

Yeah.

Sydnee:

And if the Royal family kind of had wind of this, you would've think they may have thought like, "Well this is not gonna be good for our descendancy."

Justin:

Yeah, "Let's get this... Let's weed out. This particular branch from the family tree."

Sydnee:

Exactly. But they, they didn't, they didn't seem to mind. They, they actually wanted to keep it all kind of under wraps and secret, um, which they were pretty good at doing. People—

Justin:

Some awkward dinner parties, I'm assuming when people just started bleeding out of their joints.

Sydnee:

Well, not— Like, into your joints, like, your knee will get all swollen.

Justin:

Yeah.

Sydnee:

With blood in there.

Justin:

Yes. "Percival, my Lord Percival, is that blood coming from your tear duct? Percival, excuse—"

Sydnee:

That's not how this—

Justin:

"No mother. It's some tomato sauce. Mother no." "Percival, are you bleeding from your ear holes?" "No mother—"

Sydnee:

So—

Justin:

"You're embarrassing me, mother."

Sydnee:

When Leopold died... Okay—

Justin:

[makes gurgling noises] I'm sorry about that.

Sydnee:

This is on you. I'm not making this hard [laughs].

Justin:

All right.

Sydnee:

When Leopold died—

Justin:

We got a lot to get through, Syd. You gotta pick it up.

Sydnee:

I know. I'm sorry. This is the most interesting part though.

Justin:

Okay.

Sydnee:

Okay.

Justin:

This next sentence or the whole thing? Sorry.

Sydnee:

When Leopold died suddenly, uh, there was no mention of what he died from, even though it was very clear that he died from complications related to hemophilia. Um, but if you looked at journals from the time, the Lancet and the British Medical Journal, which were both, you know, very popular at the time and still are, uh, there were obituaries for him published in both journals and also huge articles about hemophilia—

Justin:

Like in, [crosstalk] like in alternately facing pages [laughs]?

Sydnee:

Exactly. Exactly. Like, "We're not gonna..." it was like out of respect for the Royal family. Like, "We're not gonna say it, but we kind of all know." Like—

Justin:

Right.

Sydnee:

"He's had two doctors who are experts in hemophilia, who are his personal doctors."

Justin:

Yeah.

Sydnee:

"We're not stupid." Um, but a good note, uh, Leopold's death actually hugely accelerated research in the area of hemophilia.

Justin:

Mm-hmm. I bet Leopold wished we had maybe gotten on that a little bit sooner, but okay—

Sydnee:

Sure.

Justin:

Yeah.

Sydnee:

Um, his daughter, Alice was a carrier of it and she also had a son who had it. More importantly though, Queen Victoria had two daughters who were carriers. So Leopold's uh, um, sisters. Okay?

Justin:

Okay.

Sydnee:

Um, Alice was one daughter. She would give birth to a son with hemophilia and to carrier daughters. Okay? Carriers are people who aren't affected, but continue to carry the gene along. Right?

Justin:

Right.

Sydnee:

So Alice's two daughters. So Queen Victoria's granddaughters, um, this is important. One, uh, was Irene. Irene married Prince Henry of Prussia, bringing the hemophilia gene into what would later become like members of the German Royal family.

Justin:

Okay.

Sydnee:

So this is where, like, hemophilia starts to become part of the German aristocracy.

Justin:

Mm-hmm.

Sydnee:

Uh, Alexandria ma— almost married Prince Albert, her cousin, Prince Albert. He proposed to her. She refused 'cause she didn't love him. And if she had married him, she could have introduced hemophilia into what is now our current British family—

Justin:

Oh, wow—

Sydnee:

Like the current line of British royalty. But she refused. Instead, she married Nicholas, the czar of Russia.

Justin:

Oh, gosh.

Sydnee:

Gave birth to Alexi who did, of course, have hemophilia. And who was tended mainly... Do you know who was the person... medical person to take care of Alexi?

Justin:

Rasputin.

Sydnee:

Rasputin. Exactly. Who was thought to be the only person who was able—

Justin:

From downtown, Smirl? You can't gimme anything for that? Sheesh.

Sydnee:

Okay. I'm impressed your knowledge of Russian aristocracy—

Justin:

I don't have a fancy sheet of paper in front of me. Okay? I'm flying, uh, by seat of my brain here.

Sydnee:

That was good though.

Justin:

Yeah. No kidding it was good—

Sydnee:

That was good though. Okay. I'm impressed. I'm impressed.

Justin:

Woof.

Sydnee:

I'm impressed. So—

Justin:

Now, I know how Rasputin felt when he got stabbed all those times. 'Cause you have pierced—

Sydnee:

I've wounded you?

Justin:

You've wounded me deeply, my wife.

Sydnee:

Rasputin was thought to be able to alleviate Alexi's um, suffering, uh, largely through hypnosis.

Justin:

Sure.

Sydnee:

And, and of course, I don't know, of course the strain of the illness and the effects of Rasputin on the Royal family were supposedly, you know, big factors, uh, thought to be big factors in the Russian revolution that would follow.

Justin:

Right.

Sydnee:

Right. Um, one interesting side note, a couple. One, Prince Albert who Alexandria, almost married, who would've maybe revisited, you know, hemophilia on the current British Royal family, Prince Albert for a while was thought to be Jack the Ripper [laughs].

Justin:

Oh, really?

Sydnee:

Yeah. There were some theories floating around about that. He wasn't though. Don't worry. Um, and uh, although Irene and Alexandria, both were carriers as we've demonstrated and carried on, you know, the gene to different Royal families, uh, she also gave birth to a daughter who wasn't a carrier, Alice.

This is important because, again, another close, another close call for the current Royal family. Alice gave birth to Prince Philip, who of course is currently married to Queen Elizabeth.

Justin:

Mm-hmm.

Sydnee:

Another opportunity for it to kind of affect the current family. But it didn't. Um, the other part of this that's interesting is Beatrice who we really haven't talked about yet. She, uh, gave birth to two affected sons as well as a daughter, who was a carrier, Victoria who married king Alfonzo the 13th of Spain, bringing hemophilia to the Spanish Royal family. None of the current members of any Royal families are affected by hemophilia though.

Justin:

Okay.

Sydnee:

Yeah. So don't worry about that. Everybody's gonna be just fine.

Justin:

[laughs] I was very, very worried about the European aristocracy. Thank you. Or I guess is aristocracy even accurate if we're talking about Royal families, there's probably another word for it—

Sydnee:

No, I don't know.

Justin:

But anyway. I don't know.

Sydnee:

Anyway, so there you go [crosstalk] there is hemophilia and all the European Royal families and why everybody... it was all, it is all Queen Victoria.

Justin:

So in closing, go Google it. Um, so how did we start to understand, uh, hemophilia a bit better, Syd?

Sydnee:

Justin, I'm gonna tell you about that, but first, why don't you follow me to the billing department?

Justin:

Let's go.

[theme song plays]

[ad break]

Jesse:

I'm Jesse Thorn, the host of Bullseye. Bullseye is your guide to what's good in pop culture. Every week, I'll sit down with people like Elvis Costello, Elizabeth Banks, and Spike Lee, to talk about their creative work and their lives. Find your new favorite TV show, book, movie, or album, and gain new insights into the things you already love. That's Bullseye from MaximumFun.org and NPR.

[ad break ends]

Justin:

So Sydnee, I'm ready to take a stand against hemophilia and I hope you are too, uh, how do we start to fight back?

Sydnee:

So in the 1800s, we finally began to understand, uh, hemophilia a little bit. Um, first there was John Conrad Otto a doctor from Philadelphia who traced back, um, familial, what he called bleeders in different families started— And then, uh, the term hemophilia actually didn't come about until 1828. A researcher in Zurich, Friedrich Hopff, who finally named it hemophilia. So we weren't just—

Justin:

Now doesn't philia mean like, you love it. Like you're crazy for it?

Sydnee:

Uh—

Justin:
Like, so it's like, you're like, you're crazy for—

Sydnee:
Yes. I mean, like—

Justin:
Like you love to bleed—

Sydnee:
It could be the like to—

Justin:
Like you're crazy about it?

Sydnee:
Well, I mean, it's like you're prone to bleeding is what this is really.

Justin:
Right.

Sydnee:
Like apt to bleeding.

Justin:
Mm-hmm.

Sydnee:
It's not like you're—

Justin:
And hemo means blood.

Sydnee:
Yeah. Yeah. Not like you just really love bleeding.

Justin:

Just love to bleed.

Sydnee:

No, that's not. I don't... no, huh.

Justin:

No, that's not right.

Sydnee:

But I can understand why you would think that.

Justin:

Thanks.

Sydnee:

Um, throughout the mid 1900s, we started to discover all the different types of hemophilia, by different factors. As we were able to isolate different clotting factors in the blood, then we began to understand what people—

Justin:

[crosstalk] Yeah. Stuff like blood typing and more. Yeah.

Sydnee:

Exactly. So then we began to figure out different things, but, um, even as we began to understand that people had problems clotting and that there were different factors involved in clotting and that was a problem. We still didn't have a great way to treat it because we didn't have a great way to store blood yet.

Justin:

Oh, right—

Sydnee:

We didn't know exactly how to... yeah. It took us a while to figure out how to break it all down. And then it took us a while to figure out how to keep it fresh. And so often you... if you were to get a blood transfusion, it would be a straight, fresh whole blood transfusion from a family member.

Justin:

Mm-hmm.

Sydnee:

Was kind of like our best bet. Like, "I don't know. Let's get somebody related to you 'cause then it's probably fine. And we're just gonna take blood out of them and put it directly in you."

Justin:

That'd be exhausting. You'd be like following around your cousin, like, "Please don't trip, please, please. No, don't fight that bully. I'll fight him for you. It's fine. Fine. Ugh."

Sydnee:

[laughs] Uh, this was obviously less than ideal. It might not have matched. And if you look at, um, the average life expectancy, when we go back to the early 1900s, it was, like, 13.

Justin:

Wow.

Sydnee:

So obviously we needed better treatments and there was a lot of other stuff tried at this time. Stuff that wasn't necessarily great ideas. Calcium was one of the earliest things recommended. There was some thought that maybe if you didn't—

Maybe you didn't have enough calcium in your blood. So calcium salts were prescribed. Um, people started taking extracts of, like, thyroid gland and bone marrow and injecting it into people thinking maybe that somehow would make you stop bleeding. Um, oxygen was a common treatment. I don't know.

Justin:

Oh. Yeah.

Sydnee:

I mean, I guess it doesn't hurt.

Justin:

No.

Sydnee:

Um, lime was thought to be a treatment for a while.

Justin:

Absolutely not. Yeah. [crosstalk] I can call fake, fake-a-roo on that one.

Sydnee:

Hydrogen peroxide was popular, which I guess I can kind of understand that in the sense that, like, I feel like every... I have lots of people I see who, um, will have any kind of wound or ailment and they'll just kind of dump some hydrogen peroxide on it. So I can see where that's just—

Justin:

That was my dad, man, when I was a kid. I have so many memories of my dad hurting himself and just dumping half a bottle of hydrogen oxide, I think, 'cause it foams up a little bit, you think like, "There it goes."

Sydnee:

"There, it's really helping."

Justin:

Now it's getting in there.

Sydnee:

Just don't do that.

Justin:

No?

Sydnee:

No.

Justin:

No?

Sydnee:

No.

Justin:

Is it not good for anything?

Sydnee:

It's not really... It's not really helping very much.

Justin:

Not with anything?

Sydnee:

No. It's not—

Justin:

Oh, no—

Sydnee:

Really helping. It's not, I mean—

Justin:

[crosstalk] What about gargle—

Sydnee:

I guess if you're—

Justin:

What about gargling it for like—

Sydnee:

[crosstalk] Gonna use it once, it's not really gonna— It's not— No, don't do that ever. Using it—

Justin:

My dad did that a lot.

Sydnee:

No, don't gargle... what? No, don't gargle hydrogen peroxide.

Justin:

It's like a solution mix with water.

Sydnee:

No, don't do that.

Justin:

Oh, man.

Sydnee:

No. Like, if you wanna dump it on a cut, I guess that's fine. But then please stop dumping it on the cut. Because you're just gonna inhibit healing.

Justin:

Okay.

Sydnee:

So like if it makes you feel good to do it the one time—

Justin:

What's it for?

Sydnee:

I don't know, people like the foaming, they feel like it's...

Justin:

[laughs]

Sydnee:

They feel like it's helpful.

Justin:

[laughs] You could have the same effect dumping a bottle of Dial on it.

Sydnee:

When was the last time you went to a doctor and they dumped hydrogen peroxide on you—

Justin:

I assume—

Sydnee:

Or told you to, or—

Justin:

I assume I'm going for, like, a folklore, like, home remedy type thing.

Sydnee:

Uh-huh.

Justin:

I would assume they got better stuff there.

Sydnee:

Yeah. And if you listen to our show a lot, you know that those folklore home remedies always work so well.

Justin:

Okay. Uh, this has been very eye opening, but thank— So, thank you. I appreciate it.

Sydnee:

Gelatin was recommended, which I can see where, like, people thought, like, I don't know that'll... That makes things sticky [laughs].

Justin:

Sticky. Yeah.

Sydnee:

Um, in the 1930s diluted snake venom was used.

Justin:

Okay.

Sydnee:

Because it did... There were some snake venoms that could cause you to clot. So that was probably dicey. That was risky business. In the 1950s, there was, uh, one doctor who kind of started recommending you let bees sting you.

Like, literally like he had hives and he would, like, hold a bee to your skin until it stung you. Can you imagine being that kid, who's just sitting there watching the bee knowing that like the goal here is that any second it's just gonna sting you.

Justin:

Yeah.

Sydnee:

Um, whoof, uh, birth control pills were recommended, which actually isn't a crazy thought because we know that birth control pills, um, estrogen treatments in general can put you at higher risk for clotting.

Justin:

Oh, okay.

Sydnee:

So obviously this was not the best course of treatment.

Justin:

Right.

Sydnee:

But this is not crazy thinking. Um, uh, as opposed to peanuts, which were recommended [laughs] by one doctor who had hemophilia, who noticed that he had a bleeding episode stop suddenly one day and he connected it to when he ate a handful of peanuts. And so for a while, peanut extract was [laughs] given to people. That was a...

Justin:

He was going for it. Huh?

Sydnee:

Yeah. I mean, I like peanuts, but um, even once we figured out how to give people plasma, there was, uh, there's so little of each clotting factor in, like, a whole big thing of fresh frozen plasma, blood plasma.

Justin:

Mm-hmm.

Sydnee:

That, uh, you would have to give kids tons of plasma in order to replace enough of the clotting factors they were missing,

Justin:

Which, for something that's, like, a finite resource that's kind of tricky.

Sydnee:

Exactly. And, like, you're just trying— And you wouldn't be doing this unless the kid was already bleeding. So you're trying to shove as much of this into a kid who's bleeding as fast as possible to try to stop bleeding. And it was not a great situation. Um, so by the 1960s, we really hadn't imp— This is 1960s.

Justin:

Yeah.

Sydnee:

We really hadn't improved stuff much to... the life expectancy was just under 20. So in those years that followed, we had a lot of great breakthroughs. In '64, um, the clotting cascade was published in Nature. And we really began to understand all the different things that were involved with the process.

In '65, uh, Jude— Dr. Judith Graham figured out that if you thawed out plasma, those bags of frozen plasma, if you thaw it out, the stuff that the precipitate that's left, um, it's called cryo precipitate. It's just this really concentrated stuff that contains a ton of factor VIII.

Justin:

Huh.

Sydnee:

And when she figured that out, you could give that to patients instead of the whole bag of plasma. And it was a really concentrated solution of what they needed. So you could give them a lot less, a lot faster.

Justin:

It's like pan scraping—

Sydnee:

With a ton more—

Justin:

After you have—

Sydnee:

Yeah.

Justin:

Have cooked the, the steak, that was just the...

Sydnee:

Exactly—

Justin:

...really, really good stuff.

Sydnee:

Exactly. This was the really, really good stuff that hemophiliacs needed. Um, after that, we figured out how to give just single factors. And we started making, like, "Here's a bag of factor VIII, here's a bag of factor IX." And we started giving people prophylactic treatments so that instead of just treating them when they bled, you could come in and get regular injections of things to stop you from bleeding.

Justin:

Okay.

Sydnee:

Right? To give you what you needed. So you wouldn't bleed so much. Um, and then we started making synthetically available factors and we had to start finding ways to... 'cause some people's bodies started fighting back and creating antibodies against things. And we found ways to circumvent that as well.

And without getting into it, it started to become much more manageable. Um, the only major setback and I won't belabor this point, but I think you can't talk about hemophilia without mentioning it. Was in the eighties, which we were figuring out better and better ways to give people blood products. But we had not yet figured out screening for blood products.

Justin:

Mm-hmm..

Sydnee:

So we had a lot of major setbacks for people who have hemophilia in the eighties because of HIV. And then after that hepatitis C, um, uh, at a great number of people with hemophilia were affected by HIV and hepatitis C as a result of these transfusions.

Justin:

How do you mean? There, there weren't enough blood donations?

Sydnee:

Um, weren't screening blood products for these things. So they got HIV.

Justin:

Oh, okay.

Sydnee:

As a result of that. Um, so obviously we are much, much better at that process now.

Justin:

Mm-hmm.

Sydnee:

And it is exceedingly rare that you would ever get something like that from a transfusion.

Justin:

Okay.

Sydnee:

Thank goodness. Um, in addition, we have new drugs, we have recombinant stuff. Uh, like I said, synthetics, um, we need less frequent injections. There are new drugs coming out now that, so because it used to be something you would have to get an injection of almost every single day.

Justin:

Wow.

Sydnee:

Yeah. To prevent bleeding episodes, we're getting much better. Um, it's still really expensive. It's still pretty time consuming, but we're trying to make that a better process. And uh, now there are gene therapy trials underway to find ways if we could inject patients with hemophilia, with the genes that they need to make those clotting factors and they would become like part of their own DNA and then they could make some of that factor...

Justin:

Uh, interesting—

Sydnee:

...or maybe enough of that actor on their own.

Justin:

Yeah.

Sydnee:

Yeah. So that is, those trials are now underway to see if maybe we could find an even better way of treating it so...

Justin:

Well, I'll do—

Sydnee:

We've come a long way.

Justin:

That, yeah, absolutely. Especially from a life expectancy of 13, that's, that's awesome.

Sydnee:

It's amazing. When you look at, I mean, this is all just really been in the 1900s and largely since the sixties, I mean like that's really— That's that recent, that stuff has gotten so much better.

Justin:

Um, wow. That's wild. Uh, if you are somebody who's living with hemophilia, we'd love to hear from you, uh, sawbones@maximumfun.org is our email address. Uh, we always like to hear from people who have a little more firsthand experience with these topics than we do.

Sydnee:

Yeah.

Justin:

Um, uh, we are @sawbones on Twitter. We are, uh, sawboneshow.com, if you ever wanna link somebody to our program, or you can just send them our, our iTunes link. I believe it's itunes.com/sawbones, will work just fine.

Sydnee:

And if you do check us out on iTunes and you feel so inclined, why don't you, uh, give us a review and let us know what you think?

Justin:

Yeah. That's really the only way we have of, uh, of helping the show to grow. 'Cause we don't do advertising or anything like that. We just rely on your

word of mouth. So, uh, please like our show on Facebook and our Facebook group, if you wanna join that, it's Sawbones, just Sawbones. Come on over.

Sydnee:

Yeah. Also, please like our show.

Justin:

Uh, please just like it—

Sydnee:

Like just like it—

Justin:

Just like it. Um—

Sydnee:

Because we like you [laughs].

Justin:

Hey, we liked you first. Uh, that is, uh, I think that's gonna do it for us. Oh, thanks to The Taxpayers for letting us use their song Medicines as the intro and outro of our program, uh, go search for them on the internet and you can, uh, download all their music and give them a lot of money for it. An exorbitant fee, I should think.

Sydnee:

And thank you again, everybody who helped us out in the Max Fun Drive or just listened and told their friends about it. We appreciate it so much.

Justin:

We do. Uh, but until next Wednesday, my name is Justin McElroy.

Sydnee:

I'm Sydnee McElroy.

Justin:

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

[theme song plays]

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