HYPOSPADIAS CONVERSATIONS

TRANSCRIPT: DOCTOR'S ROUNDTABLE PART 3

Episode 17: Discussion with Doctors: Vinaya Bhatia, MD, Lauren Corona, MD, Pankaj Dangle, MD, Christopher Long, MD, Hadley Wood, MD

***JOHN**: Welcome to **Hypospadias Conversations** with cohosts Bonnie Steinberg and John Filippelli. We are members of the community that have experienced living with hypospadias, epispadias, the surgeries that are often used to correct for them, and we want to talk to people who are members of that community and their family and friends about many of the feelings and issues that we all have faced. We are not doctors we do not represent the medical community and we want to be clear that we are discussing our personal experiences—experiences that too often are not shared, leaving many boys, men and families feeling that they are alone. You are not alone. Our goal is to offer frank conversations about our thoughts, give many people company, give parents who are wondering what to do with their new babies that have been born with hypospadias or epispadias, some resources. to think about treatment. To think about parenting boys with this difference. The conversations are personal, frank, and we hope that you are aware of how vulnerable we feel, how risky it feels to open ourselves up in public. We hope to cultivate your compassion and understanding and create more safety to have these conversations.*

JOHN: Dr. Bhatia made a really interesting point that I'd love to hear your thoughts on, on the outcomes. She said to ask the patient because they're the one using the surgical site, how they feel about the results of the surgery. So, I was curious your thoughts on that and also specifically what criteria you would gauge to determine the success of a surgery.

DR. LONG: I think from our standpoint, we've, you know, one of the other paradigm shifts for us at CHOP is that we really focus on long-term outcomes. I think you know, it also begins, in that initial sort of conversation with families. This is not something where, say, we do a surgery when the patient's 12 months old, we see them back a couple months later, maybe six months after that, and say, you're good, we'll see you later. Call us if there's a problem.

What we know now is that we really need to follow them at least past potty training. Ideally, we really like to follow them into adulthood. So, in terms of getting the patient inputs on the outcomes and how it's working for them, it's invaluable. So we've built ways to monitor patients that have maybe missed appointments or they're over 15 years old and we wanna see them back.

There's actually a system within CHOP that we can sort of, you can look at a graph and you can look at a hundred patients within minutes and sort of see who's overdue so we can try

and contact them. I think one of the challenging things is that we run into is that so many patients are doing well, we'll give them a call, and they don't want to come in to be evaluated just because they say everything's great, you know, and we don't want to be seen.

So, we end up seeing a lot of the patients that—maybe more patients that have an issue as opposed to the ones that don't have an issue. But we're trying to work around that. And I think setting the expectations when children are young with the parents to say, hey, this is, you know, we're gonna follow patients into adulthood, number one, they seem very engaged by that because they're making the decision for their child, but ultimately, you know, from my standpoint, I really wanna be able to talk to that child when they're older and approaching puberty and adulthood to make sure that they're doing okay. So, I think it's invaluable.

Another caveat to that is the patient-reported outcome measures. There's a couple of sort of metrics that are coming out soon so that we can sort of give these—there are basically some surveys that are coming out that we can give to patients, particularly when they're older, so that we can get basically a validated questionnaire that can really sort of ask them very pointed questions, what has been validated in other patients that they find very important, so that we can get those questions answered and really find out if they're doing okay. So looking forward to that.

JOHN: Absolutely.

BONNIE: In our discussions with one surgeon in a prior podcast, he mentioned the importance of hypospadiologists being forthcoming about the published complication rates of up to 70% for proximal hypospadias from the Children's Hospital of Philadelphia of hypospadias research and also their personal complication rate. What are your thoughts? Is this related, Dr. Corona, to what you're saying with decisional regret and informing people about what the outcomes might be?

DR. CORONA: Yeah, I think, like I mentioned, if there is any type of outcome that occurs that was not discussed previously or not discussed in counseling, then it's a surprise or unexpected for the family. And that's when I think decisional regret is more likely.

DR. BHATIA: I will echo what Dr. Corona said. And I will mention that I think most of the people who mentored me in the area of hypospadiology recommended that we do our own internal audits so that we know our outcomes. And we can also make sure that our patients are acquiring the type of follow-up that will allow us to give ourselves an honest representation of our outcomes, right? And so, I try to be that person for our center. So, I review our hypospadias outcomes, but I know the centers represented in this panel, we all do this. We all look at our long-term outcomes so that we can tell patients what our outcomes are. So, this has become part of my standard practice and training. And I have my mentors to thank for that, right. For telling me to do this early and often.

But I think we're lucky that Dr. Long is a part of this panel and that he really pioneered this approach to looking at hypospadias for a lot of other centers.

DR. DANGLE: If I may jump in, I agree with Dr. Bhatia and Dr. Corona on this. But I have just one thing to point out now with this data from Dr. Long is this is a very highly selected patient population which has very severe complex hypospadias. Now when you take hypospadias, about 80% of those are more common or we call it distal hypospadias. Now the outcomes between a distal hypospadias and a proximal hypospadias are completely different.

We are talking about a severe variety of hypospadias which may sometimes takes two operations or sometimes three operations and we are trying to create more towards normalcy or a functional penis for not only urinary function, sexual function, and psychological function. So as both of our colleagues mentioned, it's extremely important to kind of do the surgeries which are tailor-made for individual patient. And that's the key. And again, looking at our outcomes and complications, reporting honestly is the key to learn from our own mistakes and kind of just use that patient-centered approach and individualized approach for each of these patients. So, if you look at the distal hypospadias, success rate is about 90-95%. It's the proximal which are more challenging and that's a very unique population. And again, it's on the rise, and we all have to get kind of used to dealing with this more and more complex patient.

And one thing I would say, it is very important for us as surgeons to manage these patients as a team, not only of pre-op evaluation and drop and post-op, but even as a surgical team. So, we should have a senior colleague and a surgeon, primary surgeon, doing the surgeries together because the first operation is the best operation for this child. And I continue to do that even today after 10, 12 years of my practice and traveling almost three times a year to doing this same operation over and over again.

DR. BHATIA: And I do want to mention that what Dr. Dangle is saying is becoming standard practice at what are considered centers of excellence for hypospadias. So generally, for more complex cases, those are booked now with two surgeons to support a better outcome. And the data is promising, but obviously, we need another 15 or 20 years to ascertain whether it's durable, right?

DR. LONG: And we weren't the first or the only group to put forth our efforts there or outcomes there. I mean, I think there was a group from Texas, Boston, Vanderbilt. There were a few different places, and Toronto. So, there was a few different places that sort of published in a similar time frame. But that paper for us, there's a big paradigm shift in terms of management of, you know, the more severe form or proximal hypospadias in that, you know, there was a lot of thought process that everything could be fixed in one surgery. And there was a big push over the years to do that surgery, which is a very complex surgery and very complex anatomy, to do everything in one surgery.

And for that paper, we broke it down and the patients that had the 70% complication rate were those patients that had a single stage repair. A lot of them had what we call a plication in terms of straightening penile curvature. And part of that was that ultimately those patients had a lot of sort of issues afterwards. They would come back with persistent or

recurrent penile curvature, which for those patients is a big deal. You know, I think anything we can do to lengthen the penis, make it the penis nice and straight. I think that there's a lot of studies that show that patient satisfaction, comfort with their own body, confidence in sexual activity as adults, there's a lot of studies that show that that has a huge impact for those patients. And so, part of what we found was that those patients had a little bit of a, you know, by just doing the plication, the penis would be a little bit shorter. They would have recurrent curvature when they were 8 or 9 or 10 years old, and then they would have to go through multiple surgeries, sometimes 3 and 4 surgeries in order to achieve a better outcome.

And so, for us, that was very sobering data. And, you know, the alternative to doing a single stage is a multi-stage. And so part of doing the multi-stage is we weren't really doing placation as the primary repair there. We were actually doing what we consider like a ventral lengthening where you're making the front of the penis, which is the part of the penis that is tethered and pulls the penis forward—by cutting the urethra, by lengthening the front of the penis, what we found is that had better outcomes.

Now, again, complicated surgeries, but the complication rate in those patients was around 40 percent or so. And I think there are some caveats there, right? And so, you know, one of the things that Dr. Canning wanted to—the study that he wanted to do was something called No Man Left Behind. And so the idea behind that paper was, okay, some patients, they'll get a complication after surgery. And there's a big difference between the patient's penis looks great. They have a small fistula, where the original opening was. It's a half an hour surgery to fix the fistula. Otherwise, erections are great. You know, the urine stream is coming from the top of the penis. Penile length is good. You know, there's a big difference between that, which is no doubt about it, that it is a complication, but it's a complication that is what I would consider like a near miss. But those patients that had the single stage repair where they had to get two and three and four surgeries, that's a bigger deal and for those patients, that's a bigger miss.

And so, the no man left behind sort of idea by Dr. Canning was, you know, we're going to identify those patients that maybe didn't get ultimately a good outcome, and maybe those patients that, you know, sort of had what we consider like a salvage procedure, where maybe they were urinating from the bottom of the penis there, maybe they had to sit down to urinate, maybe their penis was still curved. And so for us, I think that that paper was very important for us in terms of like seeing those outcomes. And yes, the complication rate is high, but you know, it *really* sort of shifted how we manage those patients. And we're going to update that paper because it's been eight years from then and hopefully with much better outcomes as we've changed our techniques to improve it.

JOHN: What would you like maybe spouses, partners to know if their partner has been diagnosed with hypospadias? What is the understanding that you would like to convey over to those people who are affected even indirectly?

DR. WOOD: I mean, I think there's some important medical things to point out. There's pretty strong evidence that men who have had hypospadias, even if they had normal testicular descent at childhood, have a little bit elevated risk of needing assistive reproductive technologies or IVF type of technologies for fertility. And so, one thing you need to hear is if you are attempting conception and unable to conceive within about six months of trying, that you probably should see a specialist to make sure that if you needed additional evaluation like semen analysis or something like that, that that can be done sooner rather than later.

So that's one thing that's—that's really important I think for people with hypospadias and their partners to know. The second thing is is that I think a lot of men, especially young men coming into adult life, a lot of what their perception is of normal genitalia is really based on maybe images they've seen online and that sort of thing, which are not sort of bell curve normal.

And so, I think a lot of men say like, oh, my penis is so much shorter than anything I've ever seen, you know, or that sort of thing. And so, I do do a lot of reassuring in these young men, like your penis is normal. It looks like normal within the 50th percentile of the guys that come in that are your age. And if there is something that's really bothering him, like curvature or a skin tag or some scarring, you know, go in and see someone and say, is there anything you can do about this? Because especially if that's limiting, you know, his confidence out in the world or his willingness to go and seek partners. I think those are, those are sort of three really important points that I, that I try to drive home to, to young men that come in with hypospadias repairs.

JOHN: And this also kind of reminds me of what you'd said in terms of really needing to understand the goals of the patient when you're treating them. And it sounds like that's a key here in terms of getting everyone involved to understand, you know, what's going on, what the goals are, and what the outc omes could be.

DR. WOOD: Yeah, I mean that's for sure. And I think that as a urologist, you know, we're always thinking first and foremost like, oh, we want to make sure the kidneys are protected and the patient's not getting recurrent infections. And so, in a lot of cases, we're always thinking about any blockages that could be in the urethra. But the fact of the matter is, a patient may come with a completely different thing on his mind. He may be concerned because of a cosmetic concern or because of curvature. And so, it’s really important to go through all of those and to really have a meaningful conversation. To have a meaningful conversation about what are your goals? How can we address them? What realistically, what could you expect if we did options A, B, or C?

These are the things in pediatric to adult care in urology. And that would be patients with spina bifida or bladder exstrophy or some of these other conditions that that that require ongoing care through childhood and surveillance.

Most—most young—young boys who have hypospadias repair sort of get it repaired and then they sort of go back to their lives and I'll say many young men don't even know that they had a hypospadias repair. It is recommended, I think, by most pediatric urologists to touch base with the pediatric urologist at the time of graduation from, you know, around 16 or 18 to make sure everything's okay. But we see lots of men who are—you know had a repair in childhood and then they're 40 and they have a problem. So…

JOHN: Can you tell us a little bit more about that Dr. Wood in terms of you know what you feel is the importance of parents discussing this with their child, their surgical history, so that they're informed for when they're older so they can understand if they do look a little bit different or if things function a little bit differently. Can you talk a little bit about that for us?

DR. WOOD: Yeah, I mean that's tricky because I think parents know their child best and what they're capable, what information they're capable of processing and learning, but it's so important that boys or young men are informed about if they had a surgery on their urethra as a child, because they are at higher risk of having a problem. And I'm referring specifically to a urethral problem with drainage of urine from the bladder.

But we know that men who've had hypospadias repair in pediatric life often also have sexual problems either with curvature of their erections or with cosmetic concerns and so that may impact their willingness or ability to seek partners and that sort of thing and that may not be something that a young man really wants to talk to his parents about and so you know it sort of puts the ball in the parents court to sort of say like hey this happened and if—if that young man or boy is not interested in talking to his parents about it, at least set up an appointment with the urologist to talk about, well these are the things that may be going on, how are things looking, is there anything we can help you with, and if something pops up down the road, this is how you can get in contact with me. So, it's, you know, it can be uncomfortable and every parent has a different relationship with their child and every young person has a different ability or capability of sort of having that information sink in. So, I usually leave it a little bit to the parents to decide, but we do recommend that parents disclose that information to their child.

BONNIE: Dr. Wood, you referred to hypospadias as a disease and sometimes I think that's such a harsh description. It's really a congenital difference. One is not truly diseased from that condition. So, I just wanted to throw that in there.

DR. WOOD: Your perspective is very true. I think of the condition of hypospadias when it trickles into ongoing problems in life. I think of that as a disease because it's something that requires intervention from medical specialists. So, I guess I was sort of using it within that context. Of course, I think a lot of patients with hypospadias probably would have a completely normal life if they never had a hypospadias repair.

And in fact, what made me think about that was many years ago, I was in Germany at a meeting and I had dinner with a bunch of—in Germany, pediatric urologists actually

become pediatric surgeons first, *so* they don't come up through urology, they come up through surgery. And so, I was having dinner with a bunch of these more senior pediatric urologists and one of them had lived, spent most of his career in East Germany. And he said to me in East Germany, before the wall came down, we didn't have the resources to fix hypospadias. We never fixed a hypospadias. So, there's this whole population of East German men who were born with hypospadias, who never had a repair in child life. And he's like, and we don't see problems in these patients.

Now we do know that patients who don't get repairs can have problems, and we do see that in patients who have just natural hypospadias that was never attempted to be fixed or corrected in pediatric life, but it does sort of beg the question, like, if it's a difference, then why is it operated on? You know, if it's part of a continuum that is within normal, then it's considered a difference. But once it gets fixed or there becomes a problem associated with like a blockage or a fistula or ongoing problems with curvature or erectile dysfunction, in my mind, then it's a disease because then we have to do something about it. So maybe we're creating some of these problems by fixing them in pediatric, quote unquote, fixing them in pediatric life that maybe… and it's also within the context of how sort of generally society has changed about what sort of the norm is for genital expectations. Like certainly when I was growing up, there was sort of male and female genitalia. And with time, I think because there's been a lot more patients out in the world who have maybe genitoplasty for trans surgery and that sort of thing, there's something called a metatoidioplasty. It's the kind of surgery that a trans man would have. Actually, those patients look a lot like pretty severe hypospadias patients, genital-wise. And so, you know, as we've sort of moved away from this concept of sort of a dichotomous definition of male and female, like I wonder if maybe going into the future there will be better acceptance of genitalia that don't look fully male, fully female, and exactly like, you know, the pictures in the atlases, in medical atlases.

BONNIE: The East German experience is so interesting and the discussion that you just had about the hope that our society could be accepting of a range of variations?

DR. WOOD: Well, young people are just engineered differently. I mean, I have teenage and college children and accepting different morphologies of all types of bodies is, you know, much—is much different than it was when I was their age. And so, I do think that just sort of culturally in America and probably across the world there's greater acceptance of people not looking like this this stereotypic quote-unquote norm. And I hope that will translate to genital acceptance and maybe mean that many of these young people who are born with maybe mild forms of hypospadias, distal hypospadias, like the parents don't feel compelled to have that operated on as a child.

And, you know, I don't know what the future will hold, but certainly there's, you know, ongoing changes at the level of policy, particularly in Europe, that's preventing operations on children, genital operations on children, even when cisgendered surgery is wanted. So that extends to hypospadias repair. And in some countries, like the country of Malta, does not permit genital operations in children. What that will look like for people coming into adult life, particularly if they have more severe forms of hyposppadias and what the chances

are of getting a good repair in a post-pubertal patient versus a pre-pubertal patient, we don't even know. It's all sort of a question at this point because we haven't gotten to the point where there's whole countries of people that aren't permitted to have operations in pediatric life coming into post-pubertal care.

JOHN: And it's also very interesting, I believe it was Alice, Alice Drager's research, but that in different countries, the criteria of what constitutes hypospadias is different, and that could also affect the treatment options for people is, I think you're pointing out, Dr. Wood, right?

DR. WOOD: Right. Yeah.

JOHN: Yeah. And along with that too, that there are some cultures, even like the Middle East and such, where the men are encouraged to sit down to urinate and it's accepting. It's considered like from a cleanliness standpoint. So, in certain cultures, they are accepting in some fashion of things that in other countries, maybe in the United States, for instance, that men aren't really supposed to be sitting to urinate. So, it's actually pretty fascinating to think about, you know.

DR. WOOD: No, and it's not just with sitting, right? It's with circumcision habits. So, I would say I think the United States is the most circumcised country on earth, but we know circumcision rates have really reduced as our population has changed and also with time. And so, things like that, like a hypospadias repair in a patient who is circumcised versus uncircumcised is very different. Whether the patients, you know, these affected children and men live in a culture where they even have the expertise or the medical wherewithal to have surgery on these.

You know, in many countries, you don't get surgery unless it's a life-or-death thing. And s,o things that may be considered sort of quote unquote elective, like a cosmetic repair in child life, would not be something that would even be considered. And so, there's lots of populations out there we can learn from.

But understanding what their lived experience is, you know, young man with natural hypospadias or quote-unquote uncorrected hypospadias is going to have a very different lived experience socially in a culture where all the other men may have not had any surgery and also where the focus on a circumcised phallus and so much focus on cosmesis as our country has, is very different. So, and even with sitting and standing that sort of thing for sure.

JOHN: Thank you for sharing that.

BONNIE: Yeah those examples are so important. Again when a parent is making the decision, how do they understand the complications for the results of surgery versus unaddressed if they let their kid go natural? What might be the complications for the results of surgery and how are they addressed and what might the complications be for unaddressed?

DR. WOOD: Right. And we don't know the full answer to that question. I think that's actually one of the most important questions that we need to get to, but of course we haven't had like randomized controlled trials of children born with hypospadias saying you get surgery, you don't get surgery.

We do know for patients, for young boys who have distal hypospadias, about 12% of them will go on to have a complication requiring a secondary surgery. And we know that there is this… and then that rate goes way up the more proximal or the hypospadias become. So, the more the hole goes towards the anus is what that's what I mean by proximal. So those rates are somewhere in about the 50th percentile for patients with very proximal hypospadias. So, we know that many patients are going to require second surgeries after the first surgery in childhood.

And the patients that I think have the most problem are ones where they have surgery, surgery, surgery, surgery, and then they have complication after complication. And then the skin, there's lack of skin and other tissues to cover it to get a good appearance. And that's sort of the term, you know, it's an outdated term. Nobody really uses it anymore. But this concept of a hypospadias cripple, some young man who's had multiple surgeries on his penis such that his penis is sort of scarred and unable to sort of be fixed because there's no substrate left to fix it. So that's really what you want to avoid of course is the patient who has multiple surgeries particularly extending into the period of childhood where they have memory and they may be able to develop fears around being in the hospital, being treated, being touched in their genitalia.

And those are the patients I worry most about, carry a lot of this into adult life and may have significant trauma associated with that. On the flip side, we also know, we did a study many, many years ago looking at all the patients that showed up in our clinic with hypospadias problems, which for the most part is blockages in their urethra because we were a urethral reconstructive clinic. So, they [had] blockages or they had fistulas, which is a connection to the skin that's in, you know, not at the end.

And a minority of those men had never had surgery for their hypospadias, about 17% of them had never had surgery, but they did have urethral blockages. So, we know that a man with hypospadias has an abnormally developed urethra, particularly like the last about half inch or so of the urethra is not enveloped with the smooth muscle that a normally developed urethra has. And I think that those men are particularly susceptible to getting urethral stricturing because that part of the urethra is very delicate. So there probably is a subset of men who don't have their hypospadias corrected, who may get into trouble later in life related to stricturing just from that abnormal distal end of the urethra. But we can't characterize that because we just see the numerator. We don't know what the denominator is for any of these. We don't have centralized congenital anomaly data collection systems in America.

Our discussion will continue with part 4…

BONNIE*: The hosts of this podcast are not medical professionals, and the information presented during the podcast is not intended as a substitute for medical advice. If you or someone you love has a medical question concerning hypospadias, please consult your If you or someone you love has a medical question concerning hypospadias, please consult your physician.*