

Darren and Emily Viti

Parents of Parker (15) Christopher (4) and Jacob (2). Christopher has Hurler Syndrome (MPS Type 1) and has had a stem cell transplant.

1	<p>TITLE: <i>Docs didn't look at the Whole Child: It took a year to get the Hurler diagnosis.</i></p> <p>https://courageousparentsnetwork.org/videos/docs-didnt-look-at-the-whole-child-it-took-a-year-to-get-the-hurler-diagnosis/</p> <p>Description: Parents of a 4-year old son with Hurler Syndrome talk about the road to diagnosis and all the signs the doctors missed. "When you have children coming in with multiple problems, look at the child as a whole. The hand doctor was looking at his hands. the hip doctor was looking at his hips." Mom talks about how she has made peace with that.</p> <p>Transcript:</p> <p>Emily: When we tell the story, I think a really important piece of the story is that Darren and I were trying to have kids for three years and couldn't. And then finally we went through IVF. That was a very difficult decision for us, deciding whether or not that's the route that we wanted to go through. A lot of emotional roller-coaster through that process and we made that decision, it was great. And it was kind of a lot easier than I thought, anyone thought it could be. And then Christopher was born and the first thing that we noticed, I always say this when I speak at places, but noticed that he had his grandfather's ears, his uncle's nose and this huge lump on his back. And so that, we kind of spiralled into asking questions all the time. Christopher had shown some other health issues that just weren't normal, but weren't severe.</p> <p>We always go into these doctors' appointments and we are like something is not right and everyone is like, no, his hands are here, his back is here, his hips don't look right. But there was never a whole kind of package approach until we walked into Tufts. And Dr. Jody Hoffman was there, and yeah she is amazing. And she -- I remember the look on her face and I was like, this isn't good. I always say like it was the second we walked in, we both I think knew immediately they knew what was wrong, but they had to get the tests to confirm it. I think our appointment was like at 4 o'clock or something in the afternoon, we didn't leave there until 6:30 or 7 and we -- at first we were like the doctors are so nice, they are all staying late like they are just going to run some tests, they don't want us to have to come back and that's so sweet and I remember Darren and I being like this probably isn't them being sweet, this must be an emergency. And that was the first day that we had heard lysosomal storage disease. And then I was like Darren don't go</p>
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home and google this, like this is not very good and I was actually sitting right here when we got home. Because I think we both still were like but he is basically fine like he had this great personality --

Darren: -- Because he was an IVF child we thought "hey what else could be going on" --

Emily: All this genetic testing that you have to do beforehand, I was actually really irritated on the way to the geneticist. Like this is going to be a huge waste of time, he is IVF, they know basically every strand of his DNA at this point. And that obviously was not true. Because they don't test for obviously everything that would be incredibly expensive, so we came home and I was like yeah but he is fine, they think it's a lysosomal -- I still couldn't get it in my head. And I -- after pleading with Darren not to google it, sat myself right here in this corner of the coach and started googling lump on your back, cloudy eyes, that was the other thing Christopher has brown eyes, Darren has blue eyes, I have brown eyes and Christopher had these like really cool like Michel Pfeiffer-looking eyes. And I was like, man, he is lucky; he got these really cool eyes. And at the ophthalmologist, that was like she had said something like, have his eyes always been that colour? And I remember being like, yeah, they're so cool. And she was like yeah, and then it started to kind of hit like, there is a lot of other stuff that we don't get that's going on here. And so she labelled it as cloudy corneas and so I was googling cloudy cornea, lump on your back, and sinus infection and everything that popped up was hurler. I was like -- okay --

Darren: And then the photos came.

Emily: Well and it's funny because there are like a 100 symptoms for hurlers and he had like three of them. So I think he had three of them that I was -- in a place where I could accept. And so I was like well this isn't true, he is not really developmentally delayed, though he wasn't walking or really talking so I don't know why I was telling myself he wasn't. But it all seemed kind of like normal and he was a really happy kid. How could he be in that much pain and be so happy, I didn't really -- that didn't really make sense to me. And I googled, I did an image search and then I just like lost it on the couch like just crying, sobbing and I remember Darren walking through that doorway and he was like what's the matter and I was like just showed him a picture I mean they were identical images, even the faces.

It's just someone thinking we are crazy and doctors just saying like no, no and that was when Christopher was seven months old. He could have been diagnosed with Hurlers at seven months, he could have been in transplant at eight months but he -- at a year he was incredible during transplant. So like I said I was so angry that moment and I go back to maybe he wouldn't

	<p>have needed hip surgery if he had got diagnosed earlier but also maybe he wouldn't have survived transplant if he had gotten diagnosed earlier and so I think those are like, when I try to reframe my thinking, I just kind of always try to stay on that side of it, like you don't have the luxury of being angry when you are dealing with all those other stuff. And I feel like that's really unhealthy for everyone if that's the kind of stuff you hang on to. But I mean it's part of the journey for sure, but it's -- those are the parts that you just kind of have to let go sometimes though I bring it up every time I talk to someone, so I haven't really let go of it, but it's tough to think about that.</p> <p>I think taking any opportunity that we can to talk to other doctors who maybe -- haven't even had an opportunity to misdiagnose or not diagnose. But going to be -- when we went to be with Dr. Hoffman, going to Genzyme and speaking there and just anything that we can do to encourage doctors to not just look at MPS, but when you have got parents coming in with multiple problems to look at all of the problems. And I think that's what was missing for Christopher; they were looking at his hands. The hand doctor was looking at his hands; the hip doctor was looking at his hips -- when he got his adenoids out finally at nine months old, they were just looking at his adenoids. And so, that kind of stuff -- trying to help everyone see the importance and as a teacher right we are always told, you got to look at the whole child and I think doctors have to be reminded of that too.</p>
2	<p>TITLE: <i>Enzyme replacement therapy, Stem Cell Transplant, and Living into the Unknown</i></p> <p>https://courageousparentsnetwork.org/videos/enzyme-replacement-therapy-stem-cell-transplant-and-living-into-the-unknown/</p> <p>Description: Parents of a 4-year old son with Hurler Syndrome talk about their decision to pursue a stem cell transplant. "For me it was, well Hurlers is a 100 percent fatal, stem cell transplants, whatever the mortality rate is, is lower which is better than a 100 percent mortality rate. We took a shot. The quality of life question is unanswerable so we didn't ask it." He's currently doing really well, but what's next?</p> <p>Transcript:</p> <p>Darren: There's a lot of things you have to juggle with, within like a six week time period -- actually probably quicker than that</p> <p>Emily: It was quick, we were in enzyme replacement within three weeks.</p> <p>Darren: Three weeks, and then we met our stem cell team or we interviewed. I guess the best thing is to call it an interview. That's when we met doctor Dunkin and had a conference with her, I think within about 40 minutes after that we felt extremely comfortable and it was almost like a</p>

brother sister relationship. Just her attitude and the way she was confident that hey you are in good hands. I think we had no question --

Emily: Yeah, the second I met her I was like this is where we need to be. And she was just -- there is something about her, I always say it to her but she is like a magician. And there was something about her that was like oh no, she is the one for Christopher and maybe she is not the one for every MPS kid but she was the one for Christopher and I think it was so clear right from the second we met her.

I think also for me it was well, Hurlers is a 100 percent fatal, stem cell transplant whatever the mortality rate of that is, in my mind I was like well if you average those together it's a 60 percent survival rate, which is better than a 100 percent mortality rate like in my mind it wasn't even a question so --

Darren: We never debated it; we were on the same page --

Emily: Which is interesting because it was actually -- there have been families who have declined that as a treatment, and when we heard that at Boston Children's, one of the doctors that we work with was like well not all families do this, and I was just like oh, I don't like I don't understand that and they probably also don't understand the other end of it from where we are. And then when I see it on the MPS society on like Facebook and in the group that we are in, people will ask often like does anybody regret getting a stem cell transplant, does anybody wish they had done just ERT, or and it's I always think like gosh did we not ask enough questions and it just like worked out really well for us, but I mean to me that's how I balanced it out, I am not really good at math but in my mind I was like well if Hurlers is going to kill him then I feel like even if stem cell transplant in the end does, we took a shot and if we just didn't, in my head it didn't make any sense to do it any other way because it raised his chances of survival. It was the only way I could think of it because if you got a 100 percent and no matter what the mortality rate is, whatever that number in between is, is his now his survival rate which he doesn't have at all without that. And ERT is wonderful but it wasn't going to save his life and I think that for us was --

Interviewer: They didn't have enough information about the success rate --

Emily: -- other families? I -- you mean the other families?

Interviewer: Yeah.

Emily: I think the questions come when you are going -- seeing the other

kids that are not surviving it and I think it just makes this it's --

Interviewer: -- surviving transplant?

Emily: Yeah because then I think you are thinking you did that to your kid, that is the only way I can kind of make sense of it --

Darren: Or it could be religious beliefs.

Emily: Yeah, some of it, I think, is religious. And there was one family who I think, I don't know if this should go on there, but there was one family that I think they already had a child that didn't survive stem cell transplant, but did for a long time after and it was like the -- some of the side effects of it were such torture that I think in their head they thought this is their path and let this be their path as least painful. And you know, I -- on a lot of levels, I do understand Christopher has graft versus host disease on his skin. It's basically eczema, and it tortures him on his skin. Kids have graft versus host in their eyes, in their bellies and their -- I mean, it tortures him on his skin, and that is awful for us as his parents to see. If he couldn't eat food and needed a bag to digest or to go to the bathroom and that was his I don't know maybe that would have been different and again like sometimes I think like God did we ask enough questions. But to me it was survival versus not survival and I think for other people that big question of quality of life comes in. Which to me is just unanswerable? So it wasn't worth asking.

Darren: Yeah I mean I think we just -- we made our decision we wanted his life to -- roll the dice, and with a lot of families that are dealing with MPS if they don't treat they know the consequences. Again like Emily was saying some kids don't make it thought transplant, others make it through, but have a very whole host of problems thereafter their stem cell transplant fails a year, two years into it and they -- now they are three years old and the option of doing it again or not doing it again and just go to ERT I mean you are really rolling the dice and in our story it's successful. The terms of having the disease were, I always use the term, he is like a one percenter, he is at one percent of that population that is doing extremely well, what's next though. That's always in the back of our mind, what is next. We always ask how far along in a stem cell transplant can fail, will it fail. And her saying well if you get past three years, four years --

Emily: Right the big years, the big -- the 100 days is the big day then it's like a year, then it's five years but --

Darren: -- but there is not enough research to say if you get passed to your next transplant -- you are good to go.

<https://courageousparentsnetwork.org/videos/as-a-parent-you-do-what-you-have-to-do/>

Description: Parents of a 4-year old son with Hurler Syndrome who has received a stem cell transplant talk about staying present and getting through his transplant.

Transcript:

Darren: When we were going through all this, we did make light of a lot of things in a joking way because I think that was our only way of saying, hey, we're in this together. I mean we basically did this together. We didn't have -- I don't want to say support from our family. Our family don't -- they don't live around the area and stuff. So Emily spent the two and a half almost -- two months in the hospital around the clock with the exception of time that she got sick. I worked continuously throughout the whole thing. Would literally run down there after work at 5 o'clock and spend the time till 10 o'clock at night and come back here because the house was under renovation and the hospital was very determined saying that the child cannot go back in the house with all that construction because of bacteria, his immune system and everything. So I will work on the house from 10 o'clock to 4 o'clock in the morning, wake up at 7 and go back to work and do that routine together. So --

Emily: It was crazy.

Darren: We thought that was the norm for us because okay hey this is what we have to do for family and then we look back at it today and I am like I can't believe we got through this. And we look at those pictures and we look at us together as a family and saying wow, who in their right mind would be have a positive mind or a positive thought process during this whole thing which was crazy and that's why I will say this -- it's -- we were a great team, I couldn't have done this with anybody else with Emily, Emily is a very strong minded person. She keeps me in check but that bond of saying hey this is what we are going to do, and our plan. I mean it was -- if we were to do it again for some reason and stuff it would just come second nature to us.

Emily: Yeah. People say -- people will say all the time like you guys are so strong and like there was a moment where I was like wasn't that a privilege thing for you to be able to say because actually you would be that strong too because you don't -- there is no option.

Darren: -- you don't understand, it's not the norm.

Emily: You just have to be that strong and only like you are saying, only

	<p>when you have the opportunity to look back and say like oh wow I can't believe we did that, and we did it like that, and we did really well like we did that. And -- but that was I remember that just kind of every time someone said that to me I was just like, oh it's like, I know that is meant to be a huge compliment but like it's actually not. It's not an insult, but anybody -- any parent who wants their kid to survive would do whatever you are being told is the right thing to do, or whatever your gut is telling you the right thing. We were told stem cell transplant and we were told these two options and my gut and I think Darren's gut too was like doctor Dunkin is the right person for him. And so I think we learned a lot about ourselves as human beings too. Like there is a lot to be said for how you question your gut and like I just -- we are not in that -- we don't have the privilege or questioning our gut --</p> <p>Darren: We are good at decision making and I became more of an emotional person.</p> <p>Emily: That is true.</p>
4	<p>TITLE: <i>He is brave, doesn't complain. He makes our life a pleasure.</i></p> <p>https://courageousparentsnetwork.org/videos/he-is-brave-doesnt-complain-he-makes-our-life-a-pleasure/</p> <p>Description: Parents of a 4-year old son with Hurler Syndrome who has received a stem cell transplant talk about his courage, his light and how they draw strength from him.</p> <p>Transcript:</p> <p>Darren: Every single procedure that he's had; he knows how to retract, lay low, get through it and then when he is ready, he just shines with a light switch. And that's the way it was his life, he is just a happy-go-lucky individual and makes our life a pleasure.</p> <p>Emily: And he knows like when it's time -- and I think that's exactly it. He just -- he may be feeling a little bit better. Because all the doctors were like yesterday he was like -- we were thinking you guys would be here another week; now today, we are talking about you guys getting out at four. But I think he just -- at three or two just listens to his body so well. And I was like though I feel better than I did yesterday. I think he knows he doesn't feel 100 percent, so he is just going to wait it out. And then when he feels a 100 percent, then he is right back to a 100 percent. It's really interesting; I think we could learn something from that because you know, you push yourself so hard. And he pushes himself when he feels really good, he pushes himself to these limits but when he is not feeling good he just stops. And you are like, how do you know how to do that. And there is always these you are</p>

	<p>always asking yourself questions like how can a kid go through everything that he has been through and have this kind of personality and not be scared of adults and not be scared of doctors and he is not like he is so interesting.</p> <p>Darren: Yeah he manages it well. When we have over 27, 30 doctors appointments a year, a lot of it is just maintenance, where is he at, and everything, and every time you have to explain it and he accepts it. Am I going to get shots, no, what are they going to do, are they doing to take off my shirt and so he knows the routines now. Even when he realised that he had to go into the next surgery and it wasn't like okay, I mean he sees the yellow Johnny go on and stuff and you know to him that is -- surgery time and but he just fights through it and he doesn't complain about it. So, which is again having him as my son he makes me a stronger person, don't sweat the small things in life because there is a lot other big things in life that are problems and in his situation he looks at it and he manages it and then I am like okay if you are happy I am happy.</p> <p>Emily: Yeah.</p> <p>Darren: If you are not, we are going to help you get, you know, you are a happier person.</p> <p>Emily: And when Jacob has to get shots with the doctors he wants to go so he can hold Jacobs hand, like that's like so interesting to me. Because I think he in his head is like oh that would be good for me that's what I -- so I always say that when I describe him like he is incredibly empathetic and part of his journey has brought him to that piece of his personality like more so than a four year old, it's sometimes very odd.</p> <p>Darren: Yeah he is like an adult, he is like a small adult and he could have an adult conversation with you, we always say that that's the reason why he was in isolation for almost a little bit over a year and people around were adult doctors so I think he bonded with that the adult individuals and became an adult himself but he obviously has a child-like side of him as well.</p>
5	<p>TITLE: <i>We don't know what the future holds and how we handle that.</i></p> <p>https://courageousparentsnetwork.org/videos/we-dont-know-what-the-future-holds-and-how-we-handle-that-post-stem-cell-transplant/</p> <p>Description: Parents of a 4-year old son with Hurler Syndrome who has received a stem cell transplant talk about how they handle living into an uncertain future for their son. "We know he's going to need a lot of surgeries in the future. What's next? We don't have that crystal ball and that</p>

drives me nuts." "We don't ask these questions about our healthy son so I try not to think about it with our son who has a diagnosis."

Transcript:

Darren: In terms of not knowing what his future holds, we think about it a lot when we go to the doctors we always say okay what's next. We are relieved when they say we will evaluate him a year from now. Those years are just going to keep on passing, and eventually we know that he is definitely going to have carpal tunnel syndrome or carpal tunnel surgery, back surgery. Everything else, organ wise is status quo, but is that heart valve going to get worse. He does have a small issue, but nothing more than it needs a repair. Hydrocephalus, they monitor that. He just has a slight swelling, but it's basically baseline right now.

So with all that said, what's next? Is it going to happen at 10, 15 or would be dealing with this when he is 20, we don't know. I mean we don't have that crystal ball and that is always in my head and I know it's in Emily's head but it does drive me nuts because it's like you see this child that's got this unbelievable attitude, he is just a happy child I mean there is a lot of kids his age that are not, they are grumpy or they had an attitude and stuff. This kid is just straight across the board, very even keeled, vey neutral, happy and stuff. My biggest fear is, okay, when he is 10, will that change when he has to have that next major surgery and say why is this happening to me. And I think we'll fight that a lot when he understands what he has. That's our next phase of this whole thing. How are we going to manage, how are we going to talk to him about that.

Emily: I look at Christopher and he doesn't care what tomorrow is going to bring because he is good today and so I kind of try to -- Darren will say things like I hope he, and I am like yeah I don't know. Like I just kind of try to not think about what is going to happen next because I think for me it's a balance of just trying to deal with what's happening right now. Because even though there is not a lot of crisis happening right now it's still -- you still got a kid with MPS and that's tough, and so not knowing how tall he is going to be or what's going to happen with his heart. The reality is I don't know those things about Jacob. So like why should Christopher's future be that much more important? And they are both important, incredibly important. So that's kind of -- I am kind of trying to wrap my mind around that like, I don't actually know the answers. All of those questions that you give yourself because you have a diagnosis; you don't actually ask those questions of your healthy children.

So I don't have worries about Jacob in school until he goes to school and the teachers are like you should be concerned about it and then I am like oh

	<p>okay. So until someone tells me to be concerned I kind of try not to think about it.</p>
6	<p>TITLE: <i>Discovering I was pregnant and being petrified to find out if the fetus is also affected with MPS.</i></p> <p>https://courageousparentsnetwork.org/videos/discovering-i-was-pregnant-and-being-petrified-to-find-out-if-the-fetus-is-also-affected-with-mps/</p> <p>Description: Parents of a 4-year old son with Hurler Syndrome talk about learning they were pregnant and the fear that this second child would also have MPS.</p> <p>Transcript:</p> <p>Emily: And then I got a positive pregnancy test and I was equally if not more devastated. I mean I was petrified. And so then I immediately called, I mean this was like in the greatest place in the world if I was going to have this, so I called our metabolism doctor and I was like hey I just got this result like what do I do. I mean I was completely freaking out. And she was like, we'll take you down to Brigham and Women's, which was down the hallway from where we were. And I had a CVS test done which was awful. But it was literally like what am I going to do. And again, this moment of like I am basically going to tell myself I am not pregnant until I have an idea. The problem was I had found out so late, that by the time we found out whether or not Jacob tested positive for MPS or was a carrier, I was going to look pregnant. And so people were going to be asking me questions and I didn't know what we were going to do, we didn't actually, we hadn't talked that far but we had talked to doctor Dunkin and she was like well the great thing is if you know before he is born, she was like, the day he is born we can take him into transplant and it will be nothing for a newborn. So we were like oh okay, so we had this hope, again like hope built up slowly but Jacob was really lucky. And then when we got the phone call, what the test results were, and he was not even a carrier. It's like -- I mean that's incredible and so again I am like well Christopher forced his way here for a purpose, Jacob forced his way here for a purpose. I always say that like there is no good reason why, I mean there are a bunch of great reasons but like there is no reason why Christopher should still be here other than he is just really freaking lucky.</p> <p>Darren: He definitely won the lottery --</p> <p>Emily: It's just crazy to me to think that these two children, one has everything stacked against him. And he's, for all intents and purposes, got a perfectly normal life. And Jacob, who we had decided we were not having any more kids and is like, I think the best thing for Christopher is to have a little brother to take care of. and I think for Jacob he is showing a lot of signs</p>

	<p>of being really empathetic and a lot of the same personality traits as Christopher and I am like man, like I don't know where this journey will take the two of them but they were meant to do it together.</p>
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Darren: They are a good team.