ROUGHLY EDITED FILE

2018 EARLY HEARING DETECTION &

INTERVENTION MEETING

DENVER, COLORADO

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AGATE A‑C

PEEL THE LABEL

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>>: All right. Minor technical difficulties, but I think we are ready to go.

Welcome. This is call peel the label. I'm Ashley Mahlstedt and I'm an early intervention teacher of the deaf in Pennsylvania.

>>: This is my colleague.

>> CART PROVIDER: Microphone needed.

>>: These are our objectives. You can see here I hope what you will get from this presentation. So before I start, just wondering if anyone in the room is familiar with Hunter Syndrome. By show of hands, anyone heard of it before? Okay so a couple of people.

What I have up here is probably the nicest way to describe Hunter Syndrome. As you can see there, the biggest take away here is progressive system deterioration. It eventually affects the muscles, bones, nervous system, and eventually brain function.

Hunter Syndrome is caused by a missing or malfunction enzyme. So this enzyme is responsible for the breakdown of complex molecules in the body.

A good way to look at it, and this is sort of an analogy the family uses to describe it, so imagine that your body is your house, and all of the molecules in your body are like bags of trash. So one week the trash collection doesn't come. No big deal. One week. You can get on with your life. You can wait for the next week. Everything will probably be fine.

But now the second week comes and trash collection doesn't come again. Now, you have two weeks’ worth of trash built up. Now, imagine a month's worth of trash. Six months’ worth of trash built up. A year. Eventually you're not going to be able to go about your daily activities because there's too much trash at your house. That's sort of the affect Hunter Syndrome has on the body. Your molecules start building up, building up, and building up and you eventually lose function of your joints user organs and your brains.

When the family which you will meet later on in the presentation, when they were given this diagnosis was quickly fouled with the phrase ‑‑ followed with the phrase don't [indiscernible]. What you'll find is not pretty. It's extremely rare. There's about 20 cases ‑‑ 2,000 ‑‑ I'm terrible with numbers. I'm sorry.

2,000 cases worldwide and approximately 500 in the United States and it primarily affects the male population.

So that's Hunter Syndrome in a nutshell. There's so much to learn about it and probably so much that we don't know about it.

So now I'd like you to meet Ethan, and I think I kind of went about this backwards because I introduced you to Hunter Syndrome, but I met Ethan before I met Hunter Syndrome. So what I met was a 20 month old just diagnosed with a bilateral moderate sensorineural hearing loss. Parents were so excited to be receiving services. Ethan had his hearing needs. They got the ball rolling. Things were going well.

Then I will never forget the day I went for my regularly scheduled session with Ethan and his family and I walked in and mom said to me, you know, my parents are coming to pick up our oldest son so that we can talk. And I kind of thought that was strange. We always included the older brother in our session, so it was a little offputting and my suspicions were almost immediately confirmed when big brother left and a floodgate of tears emerged from mom as she tried to explain the diagnosis they had just received from the genetic office.

When she finished, she looked at me and said you've heard of Hunter Syndrome, right? And I kind of panicked. I'm sure it was mentioned in grad school. I'm sure I learned about it. I don't know. I don't know.

And I left there that day and I decided I was going to do whatever I could to learn as much as I could about Hunter Syndrome so that we could form the partnership that we have now between myself, his early interventionist or team at the Clark school, Ethan's medical team, audiologist, speech‑language pathologist, everyone, and that in and of itself was probably the most critical component to Ethan's success.

So some of the services he was receiving, I went to his home and provided early intervention therapy in the home. He also received occupational and physical therapy mostly as a monitoring precaution right now just because we know there is a potential for progressive loss of his motor functions. He's just kind of monitored right now. Ethan began a toddler group program at our school, at Clarke, September so September after his second birthday, and our toddler program is family involved program where the parents accompany the child to school twice a week for three hours a day, and while the child participates in a language enriched curriculum class, the parents are participating in parent education session. They're able to communicate and share their stories and create relationships that are so valuable within this community of children with hearing loss, and so he did that for two days a week for a full year.

I often think of all these families that are coming every week, and the majority of the children, there were six in Ethan's class, their parents are sitting in the observation room talking about hearing loss and talking about audiology appointments, talking about the wonderful success they have seen in their child, and Ethan's dad had to sit there and think about that and worry about that but also think about 100 other labels that Ethan had received once he got this diagnosis of Hunter Syndrome. And I think that's kind of where we wanted to take this presentation, labels. How do we let labels define what we expect our children to do.

So that was Ethan when we met him, and what we saw ‑‑ I can tell you what we saw with Ethan, but no two children with Hunter Syndrome look alike. There's such a broad range of severity of the symptoms so it can range from mild to severe. Hearing loss can range from a moderate to severe hearing loss in degree.

So while we can't tell what you Hunter Syndrome looks like in all children, we can tell what you we saw with Ethan.

So I will let these load.

So what you can see here are just some of the typical behaviors we saw when Ethan started out our center. Hunter Syndrome is often misdiagnosed because of the behavioral component seen in children with Hunter Syndrome.

As you can see a couple of things I just wanted to highlight. Transition time was a huge trigger. I know it's a huge trigger for a lot of children, but we found an extreme response in Ethan when was time to transition. He had very repetitive play often coming into school two days a week and choosing the same exact toys, playing with the toy the same exact way. He kind of had these encrypts he ‑‑ these scripts he created. He had some rote phrases he used and probably had a vocab of 25 to 30 words he would use to label a request.

At home, behavior was similar. Oftentimes I would just show up in the window and that was enough to put him into a tantrum. [simultaneous speaking]

And if he wanted to do something and you tried to direct him the other way, it often ended poorly. But that's what we started with and I want to talk a little bit about what we ended with. By the end of the year, Ethan came into school. He was smiling. He was saying hi, to strangers in the hallway. He would hang up his backpack in his cubby and take out his communication book, a tool we use at our program. He would come in and same hi, to his friends. He could name all of his friends. He sat in his hair. He was excited to participate in group opportunities. I feel like 180 degrees is enough to describe the difference we saw Ethan in September to Ethan in July. So we come to July and now the family has to transition out of the early intervention. If any of you are EI providers, you know how scary that is for families. They dread that transition, and Ethan's family dreaded that transition like any family would, but they had this added fear, because of this label he had, was he going to be able to continue at our program? Was him continuing at our program going to affect the other children in our program? What were they going to do and together as a team and staff we ensured the family we were not going to let this label stop us from helping Ethan reach his full potential and meet the expectations we had for him. So at this point I turned Ethan's case over so I will turn it over to Sherri.

>>: It sounds like a really sad story, but I'm going to tell you, I get a little choked up. I said to Ashley, I hope I don't cry! I don't know that I'll ever have a story that I'll feel so proud of regardless of what happens to Ethan. I think we gave his family a lot of Joy, and I think that the only reason we were able to do that is because we decided as a team and I'm kind of on the periphery, I'm and administrator at the school so I try to make sure things are in place. I'm not the people doing the amazing day‑to‑day work but I'm so proud of our team and I'm proud of the fact that together we decided there's no such thing as a bad day. There's days where we get to learn about Ethan and what works and what doesn't work, and we just decided we were going to stay the course.

So as I sniffle a little bit, we're going to talk about Ethan transitioning to preschool and ‑‑ here's what it looked like. We had a strong partnership between EI and preschool. Because I spent a lot of time in my career in EI I knew what that looked like on both sides to pass the baton. So we made sure the family still felt comfortable going to Ashley about that and she spent a lot of time in the observation room with dad in the beginning of the year to ease the transition. We had Dad come in and give an in‑service to the staff and talk about his expectation and his fear. You know what his fear was? He was worried about us. He was worried about the strain on the of that because he was really concerned about Ethan's behaviors which as Ashley described to you were pretty significant.

We had a personal ‑‑ we had a behavioral specialist on the team that was critical, and we had Ethan assigned a personal care assistant so a one‑on‑one who was with him who could monitor his behavior and also keep him safe because he was head banging.

We had a framework. We closely monitored all of his levels of development. Because of the disease we knew there could be a pretty rapid decline but we needed to know right away because the medical intervention needed to be involved.

Our motto just became, let's give it a try. Let's see what's gonna work. Let's see what's gonna happen.

So I guess what we started to say to ourselves and this whole idea of labels kids, what if we didn't partner with the parents? What if we focused on what Ethan couldn't do instead of what Ethan could do? What if we let Hunter Syndrome scare us because it's scary. It's a scary diagnosis and it's an emotional diagnosis for everybody that's involved in it. The life expectancy for a child with hunter is not very long. We have had some students that are ‑‑ I think the oldest person with hunters is maybe in their early 20s and that's pretty extreme and you don't know when that's gonna happen.

We were worried could we meet his needs. Not only could we meet his needs could we keep him safe but what about the other children in the class when Ethan is off in the corner screaming at the top of his lungs within children are trying to listen and talk. So we were concerned about that.

And what if we just let his diagnosis drive the plan and we didn't decide we're going to see what's going to happen so it was a collaboration with everybody.

This is a slide. I'm not going to go into it. But I would encourage anybody who wants to learn about trust either in relationships in your personal life or with your teams, and building trusts within your team and between parents, Renee Brown is ‑‑ I love Renee brown. She has a small little webinar called the anatomy of trust. Because we want to see Ethan. This is a short little clip of Ethan. He's working with his speech‑language pathologist here. And we'll give a listen to Ethan.

>>: At the beginning of the year, we have 30 minutes of auditory speech and language for every child that's in our program. At the beginning we couldn't get Ethan out of the classroom without a major tantrum. And then we got him halfway down the hall and he would throw himself down in the hallway and have a meltdown. And then we got him three quarters of the way to the speech room. Then we got him into the speech room, and this was done ‑‑ this video is probably about a month ago. I think it's a success and the same thing happened with going out to the playground, out to recess. All transitions were really difficult for him but he really worked hard and worked his way past it.

So we talked about our starting point. That was what was typical then and here's what we're dealing with now. We have transitions without difficulty. He participates in our music class. We have four classrooms at Clarke. So not only does he move within his little group,but he has large music which is ‑‑ but he has large music which is 32 children, and probably about ten staff when we do group music once a month, and he just is ‑‑ loves every minute of it. Is involved with it. You can see all the great things. We saw a huge change in him. We've been talking about fading out his personal care assistance to maybe moving her outside of the room. She was really critical in our ability to have Ethan engaged in preschool setting.

Oh, the other great by‑product and Ashley might talk to this a little bit better than I would, as you can imagine with all these behaviors audiology appointments were a bit of a nightmare. You should tell the story.

>>: As Sherri said I do still get to see the family a lot and in those early stages when I was worked with the family, audiology was a big piece. And time and time again we would go to audiology and he wouldn't let them put the headphones on or insert earphones in. He wouldn't do the condition play game so we would leave with nothing. I would say probably in the last three months, with great collaboration with his audiologist who he sees much more frequently than a typical child with hearing loss might, he goes and checks in with her at least once a month to keep their relationship fresh. He will walk in. He sits down. They put the headphones on he gets his blocks and he dozen the whole audiological example. Dad called me. I'm calling you before I call my wife. Ethan did it! The whole thing!

>>: So I think what's really fun to look at is we had a child to came to us late diagnosis at 20 months. We had a child that came with a complicated medical history and I'm happy to tell you right now that his cognitive skills are age appropriate and his listening skills are near age appropriate. We're not kind of quite there yet, but he's three years, five months, three years six months? He'll be four in April says his early interventionist and his language is coming along too. On top of that the quality of life that the parents report at home, his interactions with his brother, which were really pretty nonexistent early on have become great too.

We've enlisted Ethan's parents to send us messages. We thought they were a critical part from you to hear from, so let's hear from Emily and Steve.

(Video clip played ).

>>: Sorry. I don't think that got cut properly.

Here's the second part of interview with Emily and Steve.

(Video clip played ).

>>: So Ethan is in this video. The first time I watched it I had to watch it like five times because I was so intent on watching Ethan that I wasn't listening at all to what Emily and Steve were saying but you can choose who you want to pay attention too. But Ethan is here playing with his ‑‑ playing with a puzzle, alphabet puzzle and if you listen to Ethan on and off, you'll hear him naming his letters too (video clip playing ).

>>: Sorry it cut off at the end.

I would like to say that our contact information is on the last slide there. I talked to Ashley pretty quickly after she spoke to the parents because then she said to me, you know about hunter's right and I went, no. But we went out to other professionals in the field. It's the one really great thing about our field. Three degrees of separation. So we're happy to share our story with anyone. Somewhere along the line you may come across a family who has the hunter diagnosis as well. So we're more than happy to share what we've done. While in some indications labels can be ‑‑ cases labels can be a good things because his label of hearing loss is actually what led to the hunter diagnosis which then led to much earlier medical interventions which we hope will have some lasting effects on Ethan's life.

We have a few minutes for questions. I will let everybody know that there's a snowstorm in Philadelphia, 20 inches, so Ashley and I literally out the door at 330 so I promise you will be too. Does anyone have any questions for us about how we handled this situation or hunter or anything else? Yes. Let me bring you the microphone.

>>: Were you able in Pennsylvania to get public school support for the ‑‑ what he had.

>>: Yeah, that's a great question. So the personal care system was a very critical part of the program. So, yes, we got full support through his individual education plan. So Clarke is a nonprofit school. We're approved private school so we partner with the public school system so we were able to get complete funding mainly because of his medical diagnosis. So, yeah, we were. Happy to say. Any other questions?

>>: As an audiologist that gets to try to get headphones on kids that sometimes are not cooperative, do you have any suggestions or want to share any of the things they did that help moved him towards that.

>>: So like I said, because Ethan is ‑‑ I might have forgotten to mention it, Ethan goes to the hospital once a week for eight hours to receive an enzyme transfusion which helps to replace the enzyme that his body is missing and because he's at the hospital so frequently, not only for that appointment but just for his vast variety of medical appointments he does check in with his audiologist very, very frequently. So a lot of the time they would just go for five, ten minutes, go in the booth, play around. Let Ethan hold the headphones and play with the inserts, Dad or mom would put them on, they would try them on big brother so I feel that was the biggest key to his success at getting to that point where he can do a full audiogram is the fact that he was able to play and explore a and be so familiar with that environment. We did some prep and practice as well at home. So that's sort of what worked for us.

>>: And the other thing that sometimes works which we encourage the families to do, we can often accompany them to the audiology appointments if our schedules line up, but the other thing we do is we use the same toy over and over and over again and then we give the family that toy to take to the audiology appointment so the child knows exactly what they're supposed to do with it. We do listening checks every morning at Clarke and then they do another listening check before they beginning their auditory speech and language individual session. So there was just a lot of practice with that task which I think helps all kids but especially some resistant friends.

We probably have time for one more question if anybody has one more question?

I just want to say I wish I had brought chocolate for all the people that stayed till the very end. So thank you so much for coming and again of you our contact information so please feel free to reach out if anybody has any questions that come up later. So thanks everybody. Safe travels. Thanks.

(End of session.)