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EHDI – Florence

Making the Case: Following 1:3:6 for Children with Microtia/Atresia

Casey Judd

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>> Hello, my name is Meredith Berger. I'm here with my colleague Michelle Kraskin to talk about Following 1:3:6 with children with microtia/atresia. We didn't have time to get to number three in the learning outcomes previously in our time slot, but we'll do our best. Michelle needs to leave, literally, when we're out of time here today, to catch her plane home... but her e‑mail address is in the PowerPoint and I could talk about this for days, so... I will stay as long as the last person wants to be here to talk about this.

So... Michelle and I started working together and discussing children with microtia, atresia, seeing more and more children in our respective locations showing up who have microtia, atresia to out of proportion to what we thought the statistics should represent in our populations, so... we started asking a lot of questions, because we were observing that the way parents were supported or not supported through screening, testing, diagnosis, referral, things like that, were very, very different from what we were seeing happen with children with sensorineural hearing loss. It made us start to ask a lot of questions about what best practice really is and how the regulations and rules apply to this population of children.

So... we're kind of curious, for the people who are here... what brought you here and... if you have had similar experiences. So... one of the things I want to ask is... if you could raise your hand, if you already know what microtia/atresia is. So... I think most of the people ‑‑ Michelle's going to do a brief overview, but that let's us know how much detail to get into and if someone isn't following what we're saying, let us know and if it's a deeper conversation, I'm more than willing to stay and I think I might have been an audiologist in a past life, so hopefully I can answer your questions.

And... for the other ‑‑ for everyone who is here ‑‑ can you raise your hand if you're a parent of a child with microtia/atresia? Any parents? Teachers of the deaf? Audiologists? Great. EHDI coordinators? Who am I missing? Speech pathologists, sorry... and I'm sure I'm still missing someone ‑‑ feel free to call [breaking up]. Anything else? Anyone else we missed? I'm sorry ‑‑ beginnings, as far as the early intervention system? Terrific... great... yes... so... welcome, thank you, everyone.

>> Michelle: Pretty much everyone raised their hand for microtia, atresia, and know what it was. Sometimes we think "we have a lot of Hispanic children, and Asian children," you're not crazy. Numbers are higher for that population. Male versus female as well.

Grades, obviously it goes from no out of ear to a closed or absent ear canal ‑‑ a lot of times we'll counsel in terms of saying "it's just a plug in your ear, it's like hearing under water." In actuality ‑‑ it's not. We have developed speech and language ‑‑ we have a good foundation where to go through. If we stick our fingers in our ears ‑‑ yeah... you can think about the information and you know what's being said. If you're thinking about a baby who has no language, they're not making those connections.

So... JCIH, clearly indicates in the ‑‑ in their position statements that children with bilateral or even lateral conductive hearing loss are part of the targeted population. And... you know... when we talk about making the case ‑‑ there's part of me that thinks we shouldn't have to ‑‑ it's kind of already made for us, right? We do ‑‑ we know there's a big disconnect in many states between what JCIH recommendations may be and what we all feel or most of us feel strongly about, should happen for all children who are deaf or hard of hearing, as far as eligibility for early intervention and services, but... it varies tremendously by state and... we are very lucky in New York State that any child with any degree of hearing loss, at all, is automatically eligible and the evaluations are determined baseline.

But... they should be automatically eligible, we also benefit from home‑based and center‑based services being allowed, and... amplification is paid for at no cost to the parents through the early intervention program, that includes BAHA, PONTO, softband wearings. There isn't another option in New York State. We have a lot of advantages that we believe every child in the country should have and is supported already by the JCIH position.

We talked earlier about the fact that we feel these families are counselled differently, but... they also have different experiences. And... I'm just going to give you a second to read this. It is a ‑‑ a blog post that was written by one of the family ‑‑ the mother in one of the families we work with. Is everyone done?

Okay... so... this experience is quite common. If you can imagine that you're a parent who ‑‑ you're the mother giving birth or the father waiting to catch the baby, often the father notices that something's not quite right. Many times the parents will describe the father looking at the baby and then looking at the doctor and then looking at the baby and looking at the doctor, waiting to see if what they're seeing is normal and maybe their ears are just a little squished from coming through the birth canal or something like that.

But... it's very common for the family member ‑‑ whether the father or somebody else, to Google in the delivery room or when they get back to whatever the recovery room is for the mother. And... they will look up information because they can see that something isn't quite right. Grade three microtia, which almost looks like a little packing peanut is the most‑common. It's unusual that you'd have microtia without atresia for grade three. They can see right away ‑‑ their baby doesn't have an ear canal. They'll Google something like... my baby has no ear. What they come up with, often, it's gotten a little better ‑‑ what they usually come across is the websites for otologists or plastic surgeons who do reconstructive surgery, or... canal opening surgery, or BAHA surgery or something like that. We'll talk about why that's kind of a problem. Many of these doctors websites will say if your child has a unilateral ‑‑ who is unilateral microtia atresia, you don't need to do anything unless a delay develops ‑‑ now... we know that that undermines the entire premise of the EHDI system ‑‑ the whole point of screening and identification is to do something. And... so... we have a population of parents that are get ‑‑ the first information they're getting is "everything will be fine, wait for a delay."

And... this chart might be a little confusing, so... bear with me. Down the side... it has age ranges, so... birth to three, older than three, older than five and... around seven or older and across the top, it has things we need to consider for children who have microtia atresia. Audiological things can be done. Ear canal‑related reconstruction ‑‑ reconstruction of the pinna and therapies and supports. And... if you look... these are the only two things that can be done birth to three. But... based on what I said, as far as families Googling and getting information, sometimes in the delivery room, all the things they're talking about or that they're reading first... have to do with things that they can't do until their child is three at the youngest.

So... again, it goes against the entire reason we're all here.

So... when we're talking about EHDI and we're talking about the referrals into early intervention ‑‑ we really need to bring ourselves, our colleagues, possibly our systems, wherever you may be and whatever your role is, and parents back to what can we do now and how do we best‑support your child so that a delay doesn't develop? It's so much easier to keep up with typical development, than try to play catch‑up later as we know from all the work we do.

And when we have longer presentations, the recurring theme that Michelle and I end upcoming back to is counseling and we don't necessarily mean counseling as you know... we're providing therapy to the family or let's talk about this, but thinking more about the topics of discussion and the recognition of the learning curve and the journey parents are on and often, many of the challenges they face... and... helping to support them in gathering the information so that they can make informed decisions. That's... one of our parent/child groups. The biggest thing we say at the beginning, the most‑important thing, I believe, for any family of any deaf or hard of hearing child is connecting with other parents because there are things that they get from people who are walking or have walked the same path they're on ‑‑ that they cannot possibly get from us. Some of the topics we cover from an early intervention point of view has to do with that critical time for auditory input and... language development and brain development.

The challenge that we have and we'll talk about this more, this is a really under‑researched population. And... so... we have to kind of look to neighboring research to really, you know... draw comparisons and make our case. The idea for keep up versus catch‑up ‑‑ we say to parents all the time ‑‑ you're going to meet adults who say "I was fine." Look at me, I was okay. I had a single‑sided hearing loss, unilateral microtia, atresia. They might, you know, they're successful ‑‑ the fact they had to adapt to their environment at an earlier age than other children ‑‑ didn't stop them in many ways.

But... we don't know, when we're looking at that two‑week‑old or four‑week‑old, six‑week‑old. If we hear from a family at two months whose child has microtia atresia, it feels late. The baby in this picture was 19 days old on the first day she came for therapy wearing a loner BAHA.

So... we talk all the time about really getting in there. We can't tell when we look at this baby, is she going to be the one who has a hard time or the one who learns to adapt really easily?

Being near a center or program that has experience, it's how do we find really good therapists who are really curious and interested in learning?

And... we have to acknowledge contradictory advice. There are going to be parents that come to us and say "I don't want to do that, my ENT or my otologist, my pediatrician said my child will be fine"... and we talk about all of the reasons why we believe doing something now is better than waiting to intervene later. Parents have to make a decision based on all the information they get.

We also recognize that, for deaf and hard of hearing children, when they have a sensorineural hearing loss, that at the beginning, denial is a very healthy and appropriate coping strategy. Parents can tune out to all of the appointments they need to go to because they can take their child's hearing aids off ‑‑ or implants off ‑‑ wherever they may be, and they can just be with their baby. When your child has microtia, atresia, you never don't see it. It's always there. For a parent who is raw and vulnerable ‑‑ remember ‑‑ this could be a parent who has a seven‑day old and they think they'll never be able to shower again. We need to remember that people stare at their baby if we say put a headband on a boy who has lost an ear. We need to be up front with that and just acknowledge... this is hard. Car seats, high chairs, strollers are the bane of many family's existence. Guess what happens when a baby puts their head back on whatever they're sitting on and they turn their head, it pops off. It's really annoying. Recognizing that mom or dad may be driving on the highway, they're on the highway, someone else is driving and they're hanging over the back seat trying to find the BAHA that's feeding back under the baby without waking the baby or something like that. Hats and helmets are really challenging too. Most ‑‑ even with the advanced programming that minimized or reduces the feedback, the reality is, putting a hat on a baby is much more complicated with this type of device than some of the others we may be used to dealing with. So... helping them find one ‑‑ you know... find that ‑‑ you know... we usually say a beret‑style hat that's several sizes too big, but has a tight brim and buy it in every color you can. When you lose it ‑‑ you won't be able to go back to the store to get it.

There's also ‑‑ there are a number of parents who have created hats or decorative soft bands or what have you ‑‑ that they sell to other parents. It makes me cringe a bit because the softband ‑‑ whatever the device is, whatever manufacturer ‑‑ the softband is part of the FDA approval. So... whatever off market thing the parent is using doesn't necessarily provide enough tension for the bone conduction to be effective, so... we always advise families, if they're doing that, before they use it as part of their every day activity, to go to their audiologist and have their audiologist do booth testing to see how much, if any, sound they're losing from using that off‑label device.

>> And just to sort of ‑‑ it's been as much ‑‑ when I tested patients ‑‑ it's been as much as 15 to 25 decibels, so... when you think about that... you might as well not even wear it ‑‑  
 >> Sometimes that's a gamble. I didn't mention ‑‑ I have a daughter who has microtia atresia and she's bilateral. It's hard to work around two BAHAs and stuff. There's a point in the winter, it's so cold ‑‑ this type of weather, you can't have your baby outside without covering, so... we kind of felt like, at the point that she was at, when it was a cold spell, that... you know, losing 15 or 20 dB was better than her either not wearing them or being cold.

So... we knew, and we limited how that happened and that's part of that supporting parents and understanding things. I will say... I, I tend to ‑‑ and I'm not sure ‑‑ I can't recall who we have photo approval for ‑‑ I often use my daughter as pictures and things ‑‑ and I'm sure she's getting to the age where she's going to tell me to stop... but... the ‑‑ every time we go to the pediatrician, the picture of the little girl wearing the facemask... was during a flu outbreak, where everybody who had a cough and a fever had to wear the mask. If you don't have ears, it's really hard to wear that. I'm not ‑‑ I'm not ‑‑ I'm not sensitive ‑‑ this isn't a trigger for me as a parent. I know a lot of parents, especially when their children are still young... that going to put it on, that could be the thing that causes them to climb into bed for the rest of the day.

So... we just, you know... we're able to put the little rungs around the post for her BAHAs and all is right with the world. But... every time we go to the pediatrician ‑‑ there's a moment where the pediatrician takes out the ear thermometer, then looks at my daughter ‑‑ then looks down at the thermometer and puts it back in the drawer and tries to find something else. So... I know if I'm going through this... that other families are as well and just recognizing that it is ‑‑ it's hard and it's painful ‑‑ sometimes... and it's going to be okay... and we need them to ‑‑ we need those parents to be okay... because them being okay or not okay... will have a direct link to the child's self‑identity and their social emotional health going forward. And things ‑‑ even for those really healthy kids ‑‑ it's hard to be different ‑‑ we really need everybody to be kind of at their best, emotionally to deal with those challenges and conversations.

So... as an audiologist ‑‑ a lot of conversation for me, personally, happens at the time of diagnosis and continues, whether or not they're having surgery. And... I think it's really important as clinicians to, to discuss the impact of unilateral hearing loss. I will always tell families ‑‑ when I first started practicing 17 years ago... my philosophy was very different than it is now. We know more. The research shows us more and we do better. I always counsel families, what would you feel like hearing on only one side? We have safety issues ‑‑ we lose all effects of bi‑normality. We discuss the ear bone gap and throw these phrases out as if the family knows what that means. When you stop and think about what does that ear gone gap really mean? A lot of times you're thinking about the surgical timeline. You're having babies in your office who are a few weeks old ‑‑ surgery's not happening until at least three, four, five, seven ‑‑ from that point until the age of surgery ‑‑ we're basically saying walk around with no hearing. I look at it as very black and white in that sense. You have to think about the surgery and the success of surgery.

Surgeons will always say it's a success and we have normal hearing. Meredith and I cringe when you go on the websites and they show you audiograms and deem normal hearing as mild or moderate hearing loss. To me, that's not normal.

Also, amplification, post‑surgery. I think back to a patient who had microtia atresia repair, she went back to school ‑‑ she had surgery. Her doctor said everything is normal. No amplification ‑‑ went to school, first day of kindergarten, came home and asked her mom for her BAHA back. Children know ‑‑ it's very important to think about that.

Thinking about the ear bone gap and what that means. Basically for me, a normal bone line, that's what you do with amplification ‑‑ you'd hear normally and we look at what's lost in the gap. The bone line, again, is how you'd be performing. You'd hear normally. Sorry?

And then... without amplification ‑‑ you're at the bottom of the gap. What's in between all speech? Your child is missing all that without an amplification.

>> So... one of the things ‑‑ when we first started working together and have continued to do since... unfortunately for this population research doesn't tell us very much. If you're looking for information about surgical options, there are too many articles, research studies to count, but... again, if we're talking about birth to three and birth to five, we're not talking about surgical options yet... so... that doesn't help us. There are a handful of articles and research studies regarding children with bilateral microtia atresia, but it's really uncommon to have bilateral microtia atresia. That doesn't help us very much. Everyone would agree ‑‑ it's clear if you can't hear out of either ear ‑‑ you have to do something. We have disagreements with some of the things out there about what should be done. But that's another topic.

And then, for unilateral, at the time, when we started working together there, were no studies at all. And the handful of studies that include ‑‑ that were on unilateral hearing loss that included children with microtia, atresia, in them, some of the larger studies, didn't pull out the data. You didn't know about performance on various tests, where these children fell and so... that was ‑‑ that was you know ‑‑ that's a problem for us.

One of the more‑recent studies that we thought was very interesting had to do with the readability and the content on microtia atresia websites that parents may be looking at. And so... for the 30 sites that were analyzed, the content was considered good for microtia and if it was information just about oral atresia, the content was fair and for readability, the overwhelming number of sites that they looked at were at a 10th grade reading level. That's a problem when I think of the families that we are working with. And honestly ‑‑ this article didn't ‑‑ I assumed that all these sites are in English and we have a lot of families that even if they have English competency, for spoken conversations, I wouldn't assume, unless I had a conversation with them, that their reading of English matched that ability in the problem area. That's a problem for us.

There was a study, I'm so excited, that came out in the middle of 2018 that talked about parent preferences for that initial information. There were 87 parents, a quarter of them reported getting no information, when their child was born. And I had that experience where I said to a family on the phone during our first phone call, something about ‑‑ something to the effect of ‑‑ so... have you started considering ‑‑ or do you have questions about your child's hearing loss and the parent will say... oh... so... you're saying he does have a hearing loss? Because nobody told me that? And that was for a five week old who had other medical needs and was identified at birth because of the other medical needs and had been to probably more specialists than I've been to in my lifetime in a short period of time, but nobody confirmed for this parent that her child had a hearing loss.

Most parents describe their experience in that initial consultation as terrible or bad... and... I realize I didn't put down the exact number ‑‑ but a very high number of families didn't have an audiologist as part of that initial team‑sharing information.

Sometimes I put in things that I shouldn't ‑‑ let me go through this ‑‑ oh no ‑‑ everything's missing. Do you want to talk about this? We have two minutes... so... we have the effect of amplification on speech and language ‑‑ there are 16 children and it was retrospective and this is what you're going to see as the... this is a retrospective at a conference parents paid to go to and stay at a hotel. Doesn't represent the range of families I work with in New York City. And it didn't look back at their history. It made some ‑‑ it drew some conclusions, but it didn't look back at what had been done early on and... yet... if you read an article ‑‑ a research study, on microtia atresia, this article is cited in every single one ‑‑ even though they, themselves, point out their weaknesses within the article.

And the effects of oral atresia on speech development and learning. Again... we're dealing with some interesting information, but we don't have context ‑‑ we don't know what's cause and what's effect. So... for next steps ‑‑ almost all the research is either retrospective or parent survey. Clearly, we have a lot of work to do to be able to support these families and these children and... give them information to help them make well‑informed decisions. None of the ‑‑ almost none of the research starts at birth. There's almost nothing that I've seen that's perspective or if it is ‑‑ the data isn't highlighted. We're really excited because we know that for people who are aware of the Locki Study from Australia, the same group is starting a longitudinal hearing loss study and they're prolific publishers which makes me optimistic, when they collect data on children with microtia atresia, they'll publish it separately. We need more information to do better.

And we thank you for being here ‑‑ we thank the families tomorrow letting us learn on them and support and grow with them. Michelle is going to leave in a couple minutes ‑‑ we have a minute to answer a couple questions. I'm getting a nod in the back ‑‑ I'm willing to stay for anyone that would like to discuss this more. Any questions? No? Yes... there's a hand in the back.

>> [Speaker off mic]. What was the name of that study?   
 >> The study that has already been done is called the Locki Study, but that's not on children with unilateral hearing loss. If you look it up, you'll get to them ‑‑ the National Acoustic Laboratory in Australia. And Dr. Ching, I believe, is the head of that lab. So... that might help you find the information as well.

>> Awesome, thank you.

>> My pleasure. Any other questions? No? Oh... yes? Is this on?   
 >> Did you ever find out why the prevalence was higher than you expected? Because... we ‑‑ I've seen that in our state as well. But... we can't really figure out why. It seems to be increasing each year ‑‑  
 >> I mean, I have a lot of theories. One of ‑‑ one of my theories is that similar to after newborn hearing screening started becoming the law of the land that there were places where doctors didn't report children who had, like mild or unilateral sensorineural hearing loss. My suspicion is the same thing is happening with children with unilateral microtia atresia. That people think of it as a birth defect. That's how the CDC tracks it, as opposed to a hearing loss. There's actually... I cringe every time I say it. It's not my wording, but there is a birth defect registry that provides lots of information. I think they have it by state and you can look at the incidence of microtia atresia. They refer to it as anotia, which means there's no ear at all ‑‑ but I think they use that blanket term. You can look to see what the prevalence is by state. There hasn't been enough concerted research to document these children, as well as other areas as we would like. Yeah? Yep?

>> Thank you, in the state where I come from, we find a lot of challenges, surprisingly that kids with unilateral atresia don't seem to go on for diagnostic, ABR testing early on. They get their nonatretic ear screened and leave it there. It seems to be very hard sell.

>> I think a lot of it starts with the birthing centers. Our ‑‑ in our hospital ‑‑ we're fortunate enough that all of our newborn screeners ‑‑ it's literally like red flag high alert. Any time a baby has microtia atresia, couldn't screen‑print baby, mom has your information to follow‑up for diagnostic ‑‑ any baby, before they even see an ENT, they go right to audiology and we're the ones referring for medical work‑up. It starts with birthing centers and educating them on our pediatrician who runs the newborn nursery, very aware of it ‑‑ she was the one who implemented our CMD program. She's very proactive, so... it helps.

>> Okay... I think one of the other things that Michelle mentioned, earlier, in the presentation, on one of the slides, is that children with microtia atresia are more‑likely to need tubes on the non‑atresia side. There are some other research indicating possible internal malformations on the non‑atresia side. Even the idea that we would not be monitoring these kids more actively seems to go against what research we have, but... you know... I think it kind of comes back to the old bias that a unilateral hearing loss isn't a big deal because... you have ‑‑ the other ear is fine. So...

>> Do you know what portion of those cases are just ENTs being cautious and placing a tube?   
 >> I can get ‑‑ when we end ‑‑ I can give you the article. I don't know ‑‑ we uploaded this after the upload period closed, so... someone might call it today that we actually did this. So... I'm not sure if it is on ‑‑ I'm not sure if it's available on the, on the conference website, but I'm more than happy to send it to you or send you the link to the article.

Betsy says she thinks it will show up after the conference. I'm glad I'm in good company with all the people who uploaded late and they'll be there ‑‑ but... if not, I'm more than happy to share the resources. I love that so many people came and are interested in this topic, because we do believe it's an under‑addressed population. Other questions?

Thank you very much for coming.   
[applause]

[Presentation concluded at 5:10:00 p.m. ET].

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