>> Welcome to what's new? Recent research on microtia/Atresia to inform practice. Our speaker is Meredith Berger.

>> Welcome everyone. I'd love to get a sense of who is in the room. So if you are a parent of a child with microtia/atresia?

Me too. A parent of the deaf?

Interventionist? Audiologist?

We don't have a lot of time so I'll do my best to get through the material and do my best to share any of the articles. It's a lot to upload.

 So for those of who work with families, some of the questions you might see are the same as the questions you see with parents working with children with sensorineural hearing loss and some are different. Obviously the what causes, how does this happen comes off quite often. Does my child need a hearing aid or a bone duction type of device, those types of questions. What about other medical issues, how will they do in school, and I think bullying is probably the biggest thing after did I do something wrong. And so prior to like 2019, there wasn't a lot of research on this group of children and that's starting to change which is nice. And so the question about risk factors comes up with professionals a lot. I often have professionals ask me about you know it seems like I have a lot of kids I work with who have microtia/atresia who are of Hispanic descent, is it just my imagination? It's not your imagination, there are higher rates in certain populations. So that tells us that families from Indigenous communities in the U.S., Central and South America, Asian families have higher rates than European descent white families.

The lowest population are those of African or African American descent which is interesting to see. These are some of the new articles of new research that's come up in the last couple of years. History of miscarriage and if you have had three or more miscarriages, the likelihood is five and a half times higher than the general population. This has been across a number of research studies, not just the ones cited in here which is really interesting.

There are chemical risk factors.

I was talking to someone on Sunday maybe who mentioned that they had worked in Texas near oil fields and that almost their entire caseload was children with microtia/atresia which is something I haven't seen yet, but it makes sense that those chemicals might have an effect on a developing fetus. Twins or more, children who are a multiple are more likely to have microtia/atresia. It's interesting, the one or both parents Hispanic or the mother being Hispanic but born outside of the U.S. So we know that there's probably a combination of environmental and genetic causes that are interacting.

Also a lot of research we're seeing the type one or type two diabetes as a risk factor.

 Parents in online communities often exchange information about whether you should or shouldn't follow up on whatever recommendations you may have received from medical professionals. Sometimes the parents are right because there's a lot of ignorance regarding needs and follow up and what else should be done, and sometimes the parents are not right. For this, there is a look at what else is happening with children, this whole section, what else might be happening that we should be aware of as professionals when parents are asking where else should I go, should I be concerned about anything else?

And so when you see that these percentages, if someone told me that my child had a 5% chance of having a cardiac issue, I would check it out. You'd probably play the lottery with those odds, so wouldn't you want to know if you were a parent. A big thing that comes up, it's inconsistent if doctors refer parents for renal checks.

Statistics indicate yes, they should be screened if there will be a lot of children that have no issues at all. But there are enough children that do that we should be checking that out to make sure. And it's not just children who are syndromic, which I think a lot of times we think if it's just microtia, if it's one ear, they have no health issues, do they really need that? But what the research is showing is it's regardless of whether it's syndromic or not syndromic.

 The research on heart defects and scans was shocking to me because we talk a lot about kidneys. Sometimes you hear of midline issues that heart could be part of it. But to see this bluntly that this many children could have heart issues and if they're not checked early on, at some point they're going to have surgery for something either reconstruction, it could be to have a tooth pulled and they need to be put out that not knowing that there is a possible cardiac defect could be really serious in terms of anesthesia.

So the recommendation is that children if they don't get an echoechocardiogram at birth, there is a good chance that that child will have some type of surgery in childhood where they might not think to run any kind of cardiac eval as part of their pretesting or presurgery protocol. I'll give you a second to look at this.

 So there's a growing, it's relative for a small population, there's a growing research in children with microtia/atresia that we often think of them as either syndromic or maybe like hemifacial microsomia, so they have some facial asemytry. And the growing research is saying that maybe there isn't someone that just has microtia/atresia, that it's really on the milder end of OAVS which includes hemifacial microsomia. That's one extreme. And what we think of as microtia/atresia as the mildest. So often children who just have microtia/atresia don't have deeper evaluations in terms of the oral structures. So nobody looks to see is there anything else. They look symmetrical. They seem okay, but I've had this experience and some of you may have as well that often you'll find some of these children have really great language, but they are completely unintelligible. And they have speech errors that are not typically based on typical developmental errors, and they're not based on what you would expect for a child with the same degree of sensorineural hearing loss. They're just a little wonky, a little off.

There's a strange kind of breathiness and nasality in places you wouldn't expect it. I suspect that we'll see more research like this that shows that it's actually related to the soft pallet movement and that it should be part of a protocol, both the medical and the intervention to really make sure that the soft pallet is symmetrical and identify if it's not, what the steps might be to follow up on that. I like to think of these as evidence-informed considerations based on the research that we have. Right off the bat, children with microtia/atresia should be followed by a craniofacial team. That is not always easy for families in more rural areas. So that is a challenge. But it's often the case that families are referred not because of the severity or because of a syndrome, it's very haphazard. So we need to think about who the people are who have information, who have experience with this, because parents sometimes run into ENTs at birth who will say, I think it's just folded, it will come down. You can't assume that every ENT or every audiologist or every teacher of the deaf or speech therapist has had any experience at all with these children. So all children with microtia/atresia should be assessed for cardiac issues definitely before any surgeries, but preferably earlier than that as part of just the follow-up steps after birth. A detailed family and pregnancy history obviously will help as well because parents just want information, they want to know why something happened or if they could do something differently if they're having other children. It's a very natural fear. And children need an in depth evaluation of soft pallet movement to identify whether they are having speech reduction issues related to the asymmetry of the pallet. Any questions? Sorry, I feel like it's a little dry.

 The genetics testing, I think, I would almost want to ask a geneticist, there's a lot happening on the research of genetics that I don't really go into, it's a lot out of my field of practice, but it's also hard for me to read it and fully explain it. There's a lot of really interesting research. To my knowledge, none of it has really translated into useable tests. So there's some syndromes they might do genetic testing for. But a lot of the things to answer parents questions of how did this happen would be harder to find.

>> So regarding the soft pallet and insufficiency like with VPI, I work in early intervention of developmental disabilities so we have a team. But I'm constantly asking about BPI for not just this but for a variety of reasons. And it seems that there are different, it's a niche, right. So there's different professionals who are really we'll take this up. And where I'm at, we do have a couple of SLPs who will pick this up who are part of the cranial facial team. But especially for people who don't have access to this really beautiful cranial facial team, and even though we do have a cranial facial team, it doesn't always get in the right hands. What is your recommendation for basically everyone in this room with a vested interest on where should we be sending these people?

Generic ENT I know isn't the answer.

>> I think you're right.

Sometimes it's not the title, it's the knowledge. And it is hard to give like just a concise answer of you should go to this person. I think it is a matter of crowd sourcing with families about what experiences they've had or other professionals and sometimes finding the person who has like related knowledge and is curious who is going to work with you. In this particular study, I believe they have a protocol for assessing for this as well. And so I think the first thing is figuring out what assessments we can do to identify it and then what do we do to address it and sometimes some of that information I think would come also from SRPs who are really embedded within cleft pallet support because I think there's some of the therapeutic strategies are going to be the same. There's someone who has a question.

>> I am a pediatric ENT.

>> No offense to your profession.

>> I think what you said is absolutely true. So if people are looking where to refer, the craniofacial team is going to be number one because those people have a protocol set up to do this stuff. As you said people in very rural areas, usually you only have to go once every couple of years, every year or two to be seen there and they can get local care with some support. So I'm in Arizona, and so we provide a lot of support to really rural providers all over the state because people are coming from eastern California and western New Mexico and all over the place to come in and can't come every time they have an ear problem or something else going on. So we try to support the local people.

Your second best bet is if you have a pediatric trained ENT in your area, even if they're not part of a craniofacial team, this is part of our practice and training. Someone who is fellow trained in pediatric would be a good choice. An SRP or audiologist who kind of runs the show, so knowing who the person is in your area is the best answer. That may be better than any physician you have because they're going to know these kids and how to get them to all of the right places. That's my two cents.

>> Thank you for sharing that.

>> In New York City there's an organization called My Face, and they work closely with hospitals, pediatric craniofacial team. So they do a lot of supports for families and adults with craniofacial. There are times when family is in a rural area for example and they're coming into New York for like craniofacial team support, there's funding to help the family travel. And so that's a good resource as well. But I do think that often connecting with other parents whether it's through one of the Facebook groups to find out either who to avoid, like you know don't go to these people. And go to this.

And you're right, I've heard parents talk about like a geneticist they met who said these are the people, it's like three towns over, it might be an hour and a half drive or something. But sometimes at least to get started, that's a good place.

 If you haven't yet had a look at the surgical, I highly recommend it. It came out in 2021, and again a lot of these articles are hard to get ahold of if you don't have some kind of account with them. But I have them all saved so I'm more than happy to share them. This is specifically about nonsurgical options for pediatrics. And if we're thinking about the goal of EHDI is to fit within one month of diagnosis. I've worked with babies fit at three weeks of age. There's no medical reason why they can't be. In this document they say two months just thinking that's how much time you would need to work through insurance. And if you have bilateral, you should have two devices. So this was a newer study that came out that was looking at craniofacial microsomia which isn't necessarily, doesn't necessarily always mean hearing loss. But they did a systematic review, so they looked at all of these studies that were already out there during a given timeframe.

And they look at how many participants they had and they looked at the numbers they report for different metrics and they try to give you a comparison to show like there's agreement on question X for children. So they found there were 5,122 records and that it ranged from 52% to 100% of patients depending on the study that had ear malformations. And it ranged from 29% to 100% of the children in that study having hearing loss which was mostly conductive. And it didn't give us any additional information on prevalence and it suggested that more research is needed which is what we know. So I thought that, you have to have these studies sometimes to go to the next study which is to actually look at prevalence. But it's a little frustrating when there isn't that much research in this field to have that confirmed.

 So this one also, course speech recognition, reorganization of brain activity in children with unilateral microtia/atresia so that there actually is differences in brain development based on access to sound or not. I think that's a big, we don't necessarily know what that brain difference does, but there is a difference and I think particularly when parents are told they don't really need a device if it's only one ear, it's important to know because I have yet to meet a parent who wants to do the wrong thing for their child and they may decide not to use amplification which is their right as a parent. But for them not to have access to the information to make the decision is unfair. It's tragic.

This is also a study regarding the lack of barriers for getting devices. In this study what they found was that a great many families never really did a follow up as part of 1-3-6. So they missed that opportunity to counsel and explain what the hearing loss meant and what amplification could do. And so right off the bat at that hospital point before discharge, something needs to change. Of the 67 of 94 who completed the ABR and were counseled on the benefits, 50 were fit with amplification. And if you look at the ages they were fit, it's very different than 1-3-6. So some of the barriers were related to communication and insurance coverage. Parents had concerns about their child using a device and if they would be dependent on it or what other people would say. But the other risk factor was not showing for the ABR which makes sense.

Children with, I need to go fast because we don't have a lot of time left. Children with unilateral conductive hearing loss report that they are not more tired than children with loss -- but their parents report that they typically are. Just to confirm we're all on the same page, that there is research to support bilateral fitting for bilateral kids. I'll give you one second to look at this. As we know families from vulnerable backgrounds need more support, information in their language, more outreach and counseling at the point of birth. And families from vulnerable backgrounds are often some of the same families who are at higher risk for microtia/atresia, so it's the perfect storm. I love that there was a behavioral study on the adjustment of preschoolers. It's nice to see that. And one of the big takeaways for me is the stress and anxiety parents felt and that some of the children had internalizing behaviors of anxiety and thinking about what that relationship might be like and what the parent support might need to be to help the parent and child both feel less stressed about these types of transitions and separations from each other. Behavioral performance and self-report measures. The takeaway is that preferential seating is not enough for classrooms. I think most of us would agree that we kind of know that, but classrooms are noisy and you have to know what your needs are to advocate for them. And you have to hear enough about your needs to advocate for them and if you're an elementary age child or middle school, high school, you're more concerned about standing out and looking different. So it was interesting to me in this one because of the teasing concern that parents have. This was a study that took place in both the U.S. and South America. And teasing started around age five in the U.S. and age seven in southh America. And that's the difference in aging when schooling starts. 40 % of parents reported their children being teased. 49% were some of the time and 39% almost never.

And 85% of teasing occurs at school. That says a lot for the work we need to do with schools in general regarding what might be happening when kids get there. I would hope things were getting better, but unfortunately it seems like from this for some children it's not.

Exploring experiences with adults which is another question parents often have, what's going to happen later. For many individuals there was anxiety around if you share information.

If you've met adults who have microtia/atresia, if it's a woman who has longer hair, it almost feels like they're hiding something to them. So they run into these barriers with like how do I now share this information. They didn't overall look at microtia as having a negative impact on their life but there were things that their anxiety or stress didn't necessarily diminish related to surgical reconstruction. But the things that helped were age and maturity and family and friends who supported them. We have two minutes left. I'm actually in this room for my next presentation, so I'm willing to stay if people have questions.

And I can send the powerpoint because we can't get through everything. But there's some school related outcomes that are probably of interest to a lot of people here. 34% of children repeated a grade in this study and two were in special education classes of the 29 participants. And so we need more research like this to help us start at those earlier points because it's hard to know if children needed services, repeated grades, or were in special ed because they didn't get the early intervention and it's hard to tease that out later. Services isn't necessarily a bad thing. My daughter still has services and she will until she graduates high school. But we don't know why they had services and I think often it's a reaction to something not going well later where we could have intervened earlier. This just confirms that as of 2018, there were only two studies and they weren't great in terms of the quality of the studies on outcomes for children with microtia/atresia. I think I'll stop here so I don't give our lovely moderator anymore stress. I will upload this for people who want to read through, I tend to put a lot of info in my slides so that you don't have to take a ton of notes. And if you email me, I'm more than happy to share any of these articles because I know how hard it is to get them.

>> Any questions? .

>> Thanks everyone.