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INVERVENTION CONSIDERATIONS FOR CHILDREN WITH COCHLEOVESTIBULAR MALFORMATION USING COCHLEAR IMPLANTS AND AUDITORY BRAINSTEM IMPLANTS

Topical Session 7

Capitol 2

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>> DEBRA SCHRADER: Good afternoon. It's so quiet in here, which is really nice. This has been a really nice facility. My name is Debbie Schrader, and I am a teacher of the deaf and hard of hearing. I am the Educational Specialist at the USC Caruso Family Center for Childhood Communication, and I'm here this afternoon representing the L.A. pediatric auditory brainstem implant study team, and I'll be talking with you about auditory brainstem implants and cochlear implants in children who have significant cochlear malformation, hypoplastic auditory nerves, and then sharing some intervention considerations with you this afternoon.

Our funding has been provided by the FDA. We have an FDA clinical safety trial at the University of Southern California that we are currently conducting We also receive funding from the National Institutes of Health, the National Institute for the Center on Deafness and other Communication Disorders. The principal investigators on our L.A. pediatric auditory brainstem implant study are Laurie Eisenberg, a research audiologist, and Dr. Eric Wilkinson, an otologist with the House Ear Clinic. I have no disclosures to report this afternoon.

I'm part of our clinical team. Our program is a program for children with hearing loss in Southern California. We follow approximately 5,000 children that have hearing loss annually. We see about 1,000 children in our clinic that are using hearing aids, bahas, auditory brainstem implants. We've been part of the Department of Otolaryngology at the University of Southern California since October 2013, when the late Dr. John Niparko established the pediatric program, and our clinical faculty includes otologists. We have 7 full‑time audiologists in our program, speech‑language pathologists, and the Educational Specialist.

Our clinical goal is to provide comprehensive care and support for the children that we serve in our community through providing multidisciplinary services in collaboration with our community partners. We have on going research studies being conducted here at the Caruso Family Center. We are part of the childhood development after cochlear implantation study. I believe we're in the 4th year of data collection and USC is one of the 7 centers that are contributing data to the study team. We hold the L.A. pediatric auditory brainstem implant study and we're one of two clinical trials currently being conducted here in the United States.

We just received funding to conduct a study that really is going to focus on novel coding strategies. Historically we've relied on coding strategies that were used with adults to program pediatric cochlear implants, and to program our auditory brainstem implants, and through collaboration with New York University, Laurie Eisenberg and David Landsberger, we've gotten a grant, and we'll begin to study novel coding strategies, which I'm excited about, in our pediatric population. And we also hold a grant that has supported our Come Read With Me at USC Program, and today I'm going to focus on outcomes associated with the children that were enrolled in the L.A. pediatric auditory brainstem implant study. Our ABI study was designed to provide intervention to determine the clinical safety and the efficacy of the auditory brainstem implant in children that we enroll in the study that were between 2 years of age and 5 years of age. As I shared earlier it's an FDA clinical trial obtained from a grant.

And there's collaboration among three facilities, the USC Caruso Family Center and then surgery and related services that are provided why Children's Hospital of Los Angeles, and the Huntington Research Medical Institute in Los Angeles. At this point in time, we have evaluated records for approximately 30 to 40 children. We have enrolled 10 children in the L.A. pediatric auditory brainstem implant study. 6 children have moved from enrollment to be included and have met the criteria, and have received surgery for auditory brainstem implants, and we've been following the children longitudinally now for 4 years.

Our multidisciplinary team on the ABI is comprehensive. We've been talking throughout this conference about the importance of a multidisciplinary team. And I wanted to make sure that we included the child and the parent on that team. We have a neurosurgeon or neurotologist, we have intraoperative monitoring, audiologists that are part of the ABI surgical team. We have our programming audiologists, the speech‑language pathologists. We've had Occupational Therapists supporting the children. The educational Psychologist and myself. As the Educational Specialist I review all the children's records before they enroll in the study. I've had the opportunity to read a number of IFSPs, a number of IEPs, looking at the services that the child receives prior to enrollment in this study, and then I follow the children longitudinally and collected information from their service providers in the community to follow their overall growth and development.

This is our team. And today's objective is to provide you with basic information about what an auditory brainstem implant is, to compare and contrast the auditory brainstem implant to a cochlear implant, and identify potential outcomes for children with cochleovestibular malformation, who are cochlear implant recipients. We've got a larger number of children that are being implanted across the United States that have cochleovestibular malformation, and we have seeing a larger number of pediatric patients using ABIs across the United States. We have a large number of children that have been implanted. We have ABIs in Italy, in Turkey that reside here in the United States that have gone outside of our clinical trials and obtained ABIs, and we do mapping on compassionate use for many of those children that receive their surgeries in another part of the world.

And I want to be able to identify some intervention considerations, short‑term, long‑term considerations for children's habilitation plans.

So I think what we have to begin with is really what those three critical questions: What exactly is cochleovestibular malformation? And historically we've looked at the research by Jackler et al., and looked at embryology. Our surgeons are asking themselves, one: Is the cochlea normal? Does it have a full two and three‑quarters turns? Is it abnormal? Do we have a rudimentary basal turn? Could that abnormal cochlea support insertion of an electrode array? Is there a nerve that they can see, moving from the cochlea to the auditory pathway?

Our surgeons are looking at their semicircular canals, what does the bony vestibule look like? Are they normal? Are they missing the posterior semicircular canal. The surgeons talk as they look at MRI imagings, can they see all four nerves, can they see three? Can they see two? Is one nerve perhaps behind another nerve and they may take a second or third look at that MRI to determine first if the child can be a candidate for a cochlear implant, if the answer is yes then they move to cochlear implant surgery, before they would be moving to an ABI.

And then we're also taking a look at the child's medical history, their global development, we're looking at speech and language considerations, what's the child's joint attention? What's the child's ability to use sign language, to communicate? Audiological considerations. We've had a number of children that have come whose parents and service providers have reported no benefit from a cochlear implant. And our audiologists have put the child in the booth, and they've been able to get responses with a cochlear implant. We've sent the child back home to the area they came from, suggested additional interventions, increased services, brought the child back 6 months later to see if the child possibly would be a candidate to enroll in the ABI study and the child's demonstrating as great a capacity with the cochlear implant as we would anticipate getting with an ABI, just as a lack of poor use of the cochlear implant and not the proper intervention to support the child's early auditory learning, and I think for some of the families, there was that self‑fulfilling prophecy that we don't know what the outcomes could be using the cochlear implant, because your child had abnormal imaging, so there wasn't the fidelity of intervention provided. And that's happened in a couple cases. So we're really looking at audiologically, is the child a candidate? Educational considerations, does the child have the intervention in his or her home community to support efficacy of use of the device so they can maximally achieve the outcomes that we'd like them to obtain?

What is an auditory brainstem implant? How many of you in here are audiologists? I'm just curious. We have a large number of audiologists. Speech‑language pathologists? Teachers of the deaf and hard of hearing?

Early Interventionists? Parents?

So we've got a good representation in the audience today.

The difference, there are similarities and differences between cochlear implants and an ABI. An ABI is somewhat like a cochlear implant in it has two parts. It has an external processor, and it has an internal processing device. It has an internal receiver stimulator that has an electrode, but the electrode goes to a paddle and on the paddle there are the electrode array goes to a paddle and on the paddle there are electrode plates. And the electrode paddle is placed on the superior cochlear nucleus, on the brainstem.

And an ABI is provided when a child that has cochlear malformation or an absent auditory nerve was unable to obtain benefit from a cochlear implant. Then we've looked at the ABI.

An auditory brainstem implant works in a very similar way to a cochlear implant. Your external processor has a microphone. Microphone picks up speech sound. It's coded by your speech processor. The speech processor is sending that electrical signal through the transmitting cable through the external headpiece head coil that's going to send information to the internal receiver stimulator and then you can see the electrode array and then you can see how it ends with the implant array that's positioned on the superior cochlear nucleus along the brainstem.

Here's just another view of the ABI placement. The ABI surgery is a much longer surgery than a cochlear implant surgery. It's a much more invasive surgery. It's placed surgically along the brainstem. It's about a 7 to an 8 hour surgery in length. The children are in the hospital for approximately 4 to 5 days, after they've had their surgery. The paddle there is representative of the cochlear paddle, the children enrolled in our study were implanted with a cochlear electrode ABI electrode, and it actually has 21 electrodes on the paddle, but what we know is that those 21 electrodes on that ABI paddle may not provide the same type of frequency specific information that the electrode array on a cochlear implant provides.

If you take a minute and compare and contrast a cochlear implant to an ABI, the cochlear implant takes advantage of the tonotopic organization within the cochlea. And a cochlear implant would be indicated for individuals that have malformed cochleas but they have an auditory nerve. They have missing inner ear hair cells resulting in sensory loss and that would be the indication for a cochlear implant.

An ABI probably provides variability in frequency information. What our ABI programming audiologists typically say is that the cochlear implant will provide many children with place of production cues. The ABI because we don't know exactly what the frequency coding is at the level of the brainstem, all the way up the auditory pathway to the auditory cortex, the child may get suprasegmental information. The child will probably get pattern information. The child will get manner of production information but specific place cues may be very difficult for a child to obtain who's using an ABI. The ABI is indicated in individuals who have absent cochlea, or absent auditory nerves.

Any questions before I move on? Yes?

[ Off Microphone ]

It is actually the entire ‑‑ I'm going to go back a slide.

The surgeon takes this trans‑labyrinth approach where they come across the inner ear space, and use ‑‑ a retrosigmoid approach where they go in through the cerebellar pontine angle and as the surgery ‑‑ it's that approach that takes such a long time for the surgeon to follow that pathway, and the actual placement on the superior cochlear nucleus also takes time, and then they ‑‑ after they place it, they do the intraoperative monitoring to make sure that they have the paddle placed at the proper place on the cochlear nucleus, so that if any ‑‑ if they need to make any type of slight modification, they can do that during the surgery, but I wouldn't want to answer anymore of the questions because I'm the teacher on the team.

[ Off Microphone ]

It's not robotic surgery, no. And typically, there are three surgeons. There's the neurotologist, the otologist and the neurosurgeon are present during the ABI surgery.

And what we see are children with cochlear implants with abnormal anatomy ‑‑ yes. Yeah?

[ Inaudible Comment ]

Yes.

[ Off Microphone ]

Yes. She's asked about, I made a comment that we see a large number of children with auditory brainstem implants at USC for mapping post ‑‑ after they've had their auditory brainstem implant placed by a surgeon in a different part of the world. We have FDA approval to what we call provide compassionate use, because the child got the device in another country, our audiologists through the approval of the FDA can provide subsequent mapping of the auditory brainstem implant at USC but for every child that would be mapped with an ABI at USC, we've got to go through the process of gaining compassionate use through the FDA, and the FDA limits the number of compassionate use mapping that can be provided at the present time.

So we see children making greater progress with abnormal anatomy than we expected. We see children with no additional disabilities making progress, and we're seeing children with additional disabilities with cochlear malformation making progress. And as I mentioned earlier, we had several children that were ‑‑ their outcomes were sometimes more favorably than what we would have anticipated with an ABI, using their cochlear implants.

So ABIs may be an option for some children, but ABIs are not better, just because they're newer technology, than cochlear implants, because again, there's the lack of that tonotopic organization. We don't know how pitch information is really going to be coded by the young child who's using an auditory brainstem implant so our expectations are not equal for children using auditory brainstem implants who have had no benefit from audition, from any type of technology than children in cochlear implants.

So it becomes essential ‑‑

[ Off Microphone ]

Because I think it's probably important to keep the dialogue going. I thought about waiting till the end for your questions but I do think it's important to keep the dialogue going, so go ahead.

[ Off Microphone ]

I should have said that, too. The children that are enrolled in our ABI study, the inclusion criteria is on clinical trials.gov, and one of the inclusion criteria on our study includes that if the child had an implantable cochlea present auditory nerve they had to have a cochlear implant first before they moved to an ABI. That's just the enrollment criteria on our trial. But that was one of the criteria,.

Yes.

[ Off Microphone ]

Yes, and you're going to see a child that had absent cochlea bilaterally.

[ Off Microphone ]

So they were able to enroll in the trial, exactly, because of the way the guidelines are written. I could have spent the talk I put together, I thought I'd have more time and so I really had to condense when I found out exactly how much time I'd have. So parent counseling, as you can tell, becomes essential, helping parents understand and set realistic expectations for surgical outcomes, initial activation outcomes, device use, helping the parents understand the importance of having a communication plan in place where the parents are partners in developing language abilities, the importance of reviewing the IFSP and the IEP and reviewing objective data we obtain during the enrollment part of the study and then post‑enrollment as we followed the children longitudinally. We have an article that's just been accepted by the Journal of deaf studies, Deaf Education, early communication outcomes of children using auditory brainstem implants, and it should be May 2018, it should be in the May issue where we follow the children the first 4 years longitudinally and share the data we're collecting that you may be interested in. What we need to know about ABI maps. ABI maps are not the same as cochlear implant maps. Our audiologists have to actually program each electrode individually with an ABI. Detection doesn't equal discrimination or identification.

More electrodes on an ABI are not necessarily better.

Recipients demonstrate sound awareness pretty quickly and recipients experienced improved perception of rhythm pretty quickly and the intensity of speech. And children may experience improvement in recognition of environmental sounds.

In summary, we can safely implant an ABI. We can safely activate and program an ABI. And we can provide access to sound with an ABI. So the big question this afternoon is: How do we teach children how to use that information? How to make sense of the electrical stimulation that's been provided? So the listening journey begins and we have to have active parent participation.

And to maximize the potential of the device, their intervention needs to be intensive, it has to incorporate the child's communication approach, and we are recommending that the children use visual language, that oral habilitation is provided, and it may need to be offered more frequently, and that auditory activities have to be embedded throughout all learning contexts to support generalization of the skills. Overall, the children enrolled in the study are having difficulty showing evidence that they can use their hearing outside of the therapy or the clinical sessions.

Parents have to be active partners in the habilitation process and our service providers need to be keenly aware of the differences between a cochlear implant and an auditory brainstem implant and there has to be some time for sensory reorganization to occur in ABI users and children using cochlear implants, and careful scaffolding of tasks is essential.

The detection audiogram can be misleading. We can't assume just because we can get a detection response on a child with an ABI at 40 dB at 2,000 hertz that that child has the ability to use that information to identify the place of production for different consonants. And we can't base our expectations regarding child performance on an audiogram.

Full‑time device use is essential and it's been hard to get full time device use. I think one the parents have had a history of the child had an implant and we didn't get any benefit from the implant so now we introduce the ABI and we have to really encourage the parent to keep it on full‑time.

And that early listening requires intentionality in an environment that supports listening and extraneous perceptual information distracts the learner. Too much out there, too much information, creates cognitive overload.

And parents need opportunities to apply their learning, and one of those crucial questions I ask as a therapist is: What is the parent's prior knowledge? I talk with the families a lot about how the session is designed so that I can quickly release responsibility to practice listening to the parent. So the parent has some strategies for making listening observable to their child.

They've got to have opportunities to transfer language learning while embedding speech, auditory, cognitive and literacy skill development across domains, and that's difficult to achieve that.

So I came up with three practice principles for this afternoon and we've just got a little bit of time left so I'm going to go through them as quickly as I can. First one is when children have had no experience listening, it takes effort to make listening an observable action. Consider the developmental age of the child. Consider the child's prior knowledge.

Go back to educational theory, Dale's Cone of Experience, when a child has no prior knowledge, they need as much sensory input, as much information so that they can construct their schemata for learning. Constructs tasks that replicate real life listening. The first video that I'm going to share is pattern perception. This is a child who had absent cochlea that was implanted with an auditory brainstem implant at 5 years of age.

He's now 3 years post‑device, or post‑activation. He's enrolled in an ASL program, and the activity that we came up with is he's got a key, and he's got a set of stamps and he is going to be the zookeeper at the zoo and his job is to listen as he moves around the zoo for different animals so if he hears the ‑‑ he's to pick up the hoof stamp and mark it here and then he goes to the next station so he's pretending to be the zookeeper so he dramatizes leaning out the elephant area and realizes he's supposed to listen and attend to the next sound. He identifies what it is. He's a second grader, he's going to make a map. This is pattern perception ‑‑ I'm sorry ‑‑ with ‑‑ if I can make it go.

He comes right back with a pattern and he picks up the stamp.

And so then we've got to define our roles. We've got to construct learning tasks that provide salient information, multisensory learning, and the task that we pick has to support the learner outcomes. We select the outcomes first, and then pick the activity or the task.

This is increasing a child's awareness of vocal turn‑taking, and he definitely has imitation skills. This is 3 months post‑ABI.

[ Video ]

I think he hears it there.

[ Video ]

[ Captioner has unstable audio ]

So again, just highlighting the importance of scaffolding. The reason why I didn't use the point prompt for mom was because I knew that he was going to immediately attend to the point prompt. And so I was going to try to help scaffold for the parent that one, mom, you've got to be aware, wait for his eyes, wait for that joint attention, wait for the mutual gazing and mom was a little bit anxious, and so I decided to prompt after dad explains in mom's first language, prompt him by just using a physical prompt that's easier to fade than the look or the point cue.

So there was some intentionality in scaffolding there.

And children with additional disabilities will require greater attention during the planning process. So I've got to constantly think about: How am I going to support independent application of intended learning outcomes?

The next video you'll see is a child who is deaf and hard of hearing, and he also has the additional diagnosis of autism. He had a rudimentary basal first turn and a hypoplastic nerve, and parent did work to get the child a cochlear implant.

This is about the 28th session that we have a therapy that you're going to see on the video. It took quite a bit of scaffolding to teach the child that, number one, I had to teach the child that when I placed an object in his hand, we were working on the conditioned response, he wasn't to throw it and that when I placed an object in his hand, we introduced the board maker stop sign that you were supposed to stop and then you're supposed to wait for the sign to flip over, put in.

And so he finally learned that okay, I get something in my hand that she gives me. I'm supposed to stop. I have to wait. And then I have to put it in when I see the change of the sign. We were able to pretty quickly fade the use of the board maker sign, wait, put in, so then I taught the child that now what I put in his hand, he was to put to his chin, and we were using that same sequence of learning to put in.

Before I even introduced speech, I taught all the physical motor actions of the conditioned response. And you'll see, this is the session when I think dad and I finally decide he's got it, that he understands and he's able to do this independently.

So thinking about the materials, what I realized too was that the heavier the object was in his hand, the more apt he was to hold it and not let it go. That was the proprioceptive input that helped him attend to this. And we'll finish up with this last video.

I'm trying to find it on the touchscreen here. There.

And being aware of all the extraneous information that distracted ‑‑

[ Video ]

I couldn't get him to put it in any other bucket, that's where the strongest visual contrast was.

[ Unstable audio ]

And just basically to accelerate learning considerations.

I'll be happy to answer any questions maybe out in the hall afterwards. I appreciate your attention and your participation this afternoon.

[ Applause ]

[ End of session ]

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