Expanding Indications for Cochlear Implants to Children with Unilateral Hearing Loss
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Presenter: Lisa Park, AuD, CCC-A
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- [Moderator] It's my pleasure to introduce our presenter, Doctor Lisa Park. Doctor Park is an assistant professor in the Department of Otolaryngology/Head and Neck Surgery at the University of North Carolina at Chapel Hill. Her clinical research focuses on expanding cochlear implant indications for children who are deaf and hard of hearing. She investigates optimal programming for children who are considered nontraditional pediatric cochlear implant recipients and associated outcomes on measures of speech perception, spatial hearing, and quality of life. Doctor Park, it's great having you with us today, and I'll turn the microphone.

- [Doctor Park] Thank you so much, Carolyn. Hello, everyone. Good afternoon or good morning, depending on where you're coming to us from, very happy to be here today, excited you guys could all join us. This is a very timely topic, so let's get into it. Here are my disclosures. I do receive research grant support from MED-EL Corporation and this study is being completed under an investigational device exemption. The cochlear implant, the MED-EL cochlear implant system is not FDA-approved for single-sided deafness in children under five years of age. They just received that indication, for five years of age and up, so we're very excited about that. I'm glad this ended up being today because we have so many questions about the study and what our outcomes have been thus far, and ideas for protocols and ways to test and follow these kids. So we're happy to be able to share with you what we have so far. Here are our learning outcomes. So we'll be talking about characteristics in general of children with severe to profound unilateral hearing loss, talk about some challenges in testing and programming these children, and identify some potential benefits and outcomes that we're seeing so far in our cohort. So let's get started. We need two intact auditory pathways in order to know where a sound is coming from and to be able to listen to speech target in a noisy environment. When children have hearing loss in one ear that is not able to be amplified with a traditional hearing aid, they aren't able to access these cues. This can lead to distraction and fatigue which can have a snowball effect on learning and communication. Currently the available treatment options involve
routing the signal from the poorer ear to the better hearing ear. So let’s take a look at what the research shows in terms of how unilateral hearing loss impacts children and dive into the current state of treatment. Estimates from newborn hearing screening programs suggest that there’s about one congenital unilateral hearing loss per 1,000 births, but acquired etiologies abound with this population and incidents increases to as high as 6.3% among six to 19-year-olds and it’s estimated that 70% of unilateral hearing losses fall within the severe to profound range. Children with unilateral hearing loss are often regarded as being just fine. We expect that they will be able to learn to listen, talk and function with one normal hearing ear, but in fact the literature tells us rather clearly that this isn’t the whole story. We’ve known since the ‘80s that children with unilateral hearing loss face educational challenges. Studies have cited that between 22 to 35% of these children repeat a grade, and 12 to 41% need an additional support under an individualized education plan.

There’s also evidence suggesting that they struggle cognitively. Whether this is due to the hearing loss or etiology remains to be seen, but it is an additional hurdle that children with unilateral hearing loss face. Parents report more behavioral concerns as well. Studies consistently show that children with profound unilateral hearing loss in particular perform more poorly on measures of expressive and receptive language than their peers with normal hearing in both ears. The 2013 study by Lieu is particularly interesting as she studied pairs of siblings with and without unilateral hearing loss longitudinally. What she found was that the siblings with unilateral hearing loss struggled more with language and had higher educational needs than their brothers or sisters with normal hearing in both ears, and of course we know that children who aren’t able to hear from both ears struggle with hearing in noise and with localization. Children with unilateral hearing loss use more effort to understand speech and to communicate, and this can lead to challenges with quality of life and fatigue. As audition is integrated into neural function, unilateral input from an early age may negatively impact higher-level brain functions, and there are multiple studies listed here that support this theory. You may find that these children struggle with auditory
attention, executive function, and even sensorimotor control, and this is certainly something that we’ve seen in our clinic. These neural changes can be long-lasting. In fact studies have shown that this imbalance of input can result in cortical reorganization. Etiology-wise, cochlear nerve deficiency is very common in unilateral profound hearing loss and we’ve even seen it in children who do have some low-frequency residual hearing. They often present with unilateral auditory neuropathy. So the hair cells are there but there’s no nerve or not enough nerve to fire synchronously, so that’s why an MRI looking specifically at the auditory nerve is very important in this population. CMV is very common with unilateral hearing loss. I’ve seen citations of about 40%. It’s often progressive. Children may lose hearing in the contralateral ear over time, and unfortunately, unless they were caught at birth, and we don’t often know that CMV is the etiology. I know here in North Carolina we have difficulty getting a hold of those Guthrie cards to be able to even have an idea whether or not CMV may be something that we’re dealing with. These are often the kids who may be caught at their kindergarten screenings. Unilateral malformations aren’t uncommon either. Sometimes there will be a strictly unilateral Mondini or other malformation with a completely normal cochlea on the contralateral side.

Sometimes there will be bilateral malformations, but one ear just loses hearing before the other. In cases like this, or in CMV cases where there’s a real risk of progression in the contralateral ear as well, we push strongly for early implantation of the deafened ear because there’s a real chance that bilateral hearing loss will develop over time. Meningitis can also result in unilateral hearing loss. In these cases you need to be in clinic quickly before ossification sets in and cochlear implantation becomes difficult. Trauma is also a possibility. Temporal bone fracture can result in unilateral hearing loss, but oftentimes we just don’t know the cause. So we always support binaural input for children with bilateral hearing loss, but the treatment of options available to children with significant unilateral hearing loss don’t provide binaural input. They involve rerouting the signal to the better hearing ear via a CROS or a bone conduction device. The problem with this is that neither of these options provide auditory stimulation to
the neural pathways of the affected ear and for many children this does not provide enough benefit for localization and hearing in noise particularly when it comes to using squelch and binaural summation. In the classroom, assistive listening systems are often recommended and while they increase the signal-to-noise ratio and improve hearing in noise, they do not provide binaural cues. We aren’t able to fit any of this technology on infants and young children and need to wait until they are physically big enough for the technology. So the only way that we are able to bring stimulation to the auditory pathways and neural structures of both ears of a child with hearing loss too significant for traditional amplification on a deafened ear is through a cochlear implant. Enter the Pediatric Unilateral Hearing Loss clinical trial. This is an investigator-initiated study. Doctor Kevin Brown is the principal investigator and the study is sponsored by MED-EL Corporation. We have an investigational device exemption from the FDA to perform the study, so it’s not currently approved for children under five. We started enrollment in 2017 and currently we’re fully enrolled with 20 participants. The aim of this study is to investigate the effectiveness of cochlear implantation in children with moderate to profound unilateral hearing loss using aid within subject’s design.

We'll talk a little bit about our protocol development, including our inclusion criteria, the characteristics of our enrolled patients, and our test battery. The inclusion criteria are listed here. Generally our study includes typically developing children who are preschool and kindergarten-aged at the time of implantation. All were cochlear implant candidates in the ear to be implanted. English is their primary language and none of them have cochlear nerve deficiency, major inner ear malformations or ossification. They all receive a MED-EL SYCHRONY device and two SONNET speech processors. All subjects are programmed in an omnidirectional mode and we are not enabling any wind noise reduction for the purposes of this study. We’re also only using ear level processors in an attempt to avoid any possible mic placement affects particularly when we’re looking at spatial hearing. Here are the details on our participants: ages at implantation, etiologies, and suspected length of profound hearing loss at the time of device activation, and here’s why I say suspected. Only seven of these children were
identified via newborn hearing screening. One was not tested, he was born out of the country, and four of them, which is 20% of the full cohort mind you, initially referred and then passed on rescreen with their newborn hearing screenings. So we suspect that there may have been at least some hearing loss at birth in these children that was missed. Of the remaining eight who passed their newborn hearing screening, two reported a moment where the child reported that, "That's my bad ear," and then the parents realized that the child couldn't hear from that side. Three had sudden loss related to a specific event. So aside from those sudden cases and the children identified at birth or identified prior to progressing to a profound hearing loss, it's very difficult to know the length of profound loss for at least eight of these cases so we've based these numbers on parental estimates. The two older children noted with asterisks were a bit older and were implanted under compassionate use as part of the study. Etiology-wise, we don't have any cases of C&D as it was an exclusion in this study. CMV is known in three of the children and we suspect that many of these unknown cases may be CMV related as well. Two of our participants have Mondini malformations unilaterally. We also have a child with Waardenburg syndrome. Subjects one through 11 have completed the 12-month postop assessment and will receive the bulk of the attention in our talk today.

Our test battery includes measures of detection, single word recognition, hearing in spatially separated noise, localization and several questionnaires meant to look at device acceptance and quality of life, and we'll go into each of these in more detail. So first we get an unaided audiogram and we repeat that every three months or so. It's so important to monitor that contralateral ear religiously as studies have indicated that as many as 40% of children with unilateral hearing loss eventually go on to have bilateral hearing loss. We can't tell when this is going to happen or even if it's going to happen, so we wanna make sure we keep a close eye on that other ear. We also get a plug-and-muff audiogram at each visit. We plug the normal hearing ear with a foam plug that has a 33 dB noise reduction rating, and then we add bands earmuffs. So they have a noise reduction rating of 28 dB, so adding muffs gives us a little boost in
attenuation. But when you’re talking about kids with zero dB thresholds or even better in the contralateral ear, every little bit counts. We obtain a sound field audiogram, plugged and muffed, CI processor off first, and then we add the processor to the mix without adjusting the headphones or the plug or the muff, and then we go ahead and we test them again. Let me show you a little bit more about why we’ve chosen to do it this way. So here we have unaided left ear thresholds and we’ve marked SL as the thresholds obtained in the sound field left ear plugged and muffed without the speech processor. When we add the speech processor, marked as CR, our thresholds are within test-retest differences of that plug and muff and we probably aren’t testing detection with the cochlear implant. For this child, we can feel more confident that we are testing detection with the cochlear implant. And yes most plug-and-muff thresholds are falling between 50 and 60 dB HL, but we’re dealing with a pretty razor-thin range here in trying to make sure we’re measuring what we think we’re measuring. Not every child has the same attenuation, even between visits. The placement of that plug and those muffs is really important. Let me show you an example here.

So if you were to obtain the CI audiogram with the left ear plugged and muffed, assuming that those plug-and-muff thresholds were at 50, you might think that she could use some reprogramming, but that she was detecting something with her processor. In reality attenuation with that plug and muff wasn’t what we would expect on this day for whatever reason and we really can’t be sure which ear those CI responses are coming from for the majority of the frequency range, so it’s important to check that plug-and-muff threshold without the CI whenever you are testing children with a CI and normal hearing in the other ear using this paradigm. So that’s how we’re making sure they are detecting. Let’s get into some outcomes, shall we? Let’s talk about outcomes for the 11 kids who have completed the first 12 months of the study. We’ll start with quality of life and questionnaire data. We’ll talk about fatigue, perceived benefit and listening effort. So the Peds-QL measures how tired a child feels generally: how sleepy they are, how cognitively fatigued they may be, and a higher score indicates less fatigue. While it does look like we are seeing a trend for improvement on
parental ratings of child fatigue over time, it hasn’t reached significance, and there are a few things to mention here. More than one parent has mentioned that they wish they could go back in time and report on the scale preop again because they didn’t know how tired their children really were, until they weren’t anymore. We’re talking about kids who were six years old and taking naps, but their parents didn’t think this was unusual because it was the status quo for their child. We’re also noticing that children who entered kindergarten during this year aren’t showing as much improvement. So we wonder how much new demands of school are coming into play, but we need to wait until we have a full 20 participants before we can delve into those factors, and as a mom of a child who just started kindergarten last week, I’m seeing that that’s, yeah, they’re really tired when they start kindergarten. So parents give their input, but for the children who are five or older, they are able to rate their fatigue as well. It’s interesting that the children are rating themselves as much more fatigued than their parents rate them, and this is a finding that has been reported in the literature before.

While we are questioning some of the validity of this measure for these kids, we do feel that it’s important to note that children may be feeling more tired than their parents think that they are so we wanna get the input from the kids as well. Parents are filling out a pediatric version of the Speech Spatial and Qualities Questionnaire as a proxy for their children. It’s very similar to the adult version of the SSQ that’s used very frequently in clinics and in research, but the language is more child friendly. Thus far we’re finding a statistically significant trend for linear improvements over time in all three domains, which is very exciting. Parents feel that their child’s understanding of speech, their spatial hearing, and even their perception of sound quality is improving with the implant, especially the quality has really surprised us. When we look at questions on the SSQ meant to target listening effort, we see the same linear trend for improvement. So parents are seeing that their children aren’t working as hard to listen since using the implant and we’re very excited about this because we feel it may have trickle-down effects to behavior and education. So let’s talk word recognition because that’s what audiologists like to target. We like to talk about that word recognition. So
we used direct connect testing to isolate the cochlear implant ear for our pediatric unilateral hearing loss patients, and here is my ear. I have plug-in bone conduction headphones for myself so that I can follow where we are in the word list. These kids are really good at picking up cues from leaky earbuds and headphones and I find that using bone conduction phones works nicely. The children are connected to their SONNETs with the FM sleeve and the 90-10 cable. To get us both connected to the audio jack on the computer, we use this particular splitter. What’s really nice about this splitter that I just found on Amazon is that each jack has a separate volume control so the child is able to turn their volume to a level that they deem most comfortable and the audiologist can turn theirs to where it’s just audible to avoid any possibility of leakage. So even though they’re bone conduction headphones, they still can have some leakage so I keep it turned way down to just enough so that I can tell where we are in the list. The splitter is plugged into the headphone jack of the computer and we turn the volume control on the laptop up to about 95% to avoid some clipping and the child adjust their volume on the splitter independently to where they feel the volume is the most comfortable. We make sure we do this each time we switch to a different list.

So you may be asking, why are you using this setup? Why not just mask the contralateral ear or plug and muff? Well we have a few concerns about both of these options when it comes to word recognition and the sound field. So we don’t like to use masking with this population of children for a couple of reasons. First children are more susceptible to masking than adults to begin with, and for speech shaped noise, they don’t develop the same skills adults have until about age 10. Add to that the evidence that children who have unilateral hearing loss may have developed cortical changes that influence their ability to make use of auditory selective attention, and you have a situation where adding masking to the normal hearing ear for a speech perception testing in the sound field isn’t really testing isolated speech perception in the CI ear. Central masking in an auditory system that’s been compromised for selective attention could really impact your results here. We also had concerns about plugging and muffing in the normal hearing ear. This is an unaided air conduction audiogram for
inserts for the left ear and what’s marked as as SL was measured with a plug and muff in the sound field with the cochlear implant off. Presenting speech stimuli in the sound field at 60 dB SPL might be okay for speech perception, but there is a possibility that the child could pick up some fundamental frequency cues from that normal hearing ear even with a plug and muff. In fact when we enter this plug-and-muff audiogram into the loss simulator on the two as a conductive loss measured in the sound field and then we run average speech as part of a simulator, you can see that some of the energy is still within the range of audibility for the plug-and-muff ear alone. For that reason we didn’t feel it was enough isolation for research purposes and felt direct connect testing was our best option. We can go ahead and run this video really quickly so everybody can see how some of that energy can really get above that audibility line so they may be picking up some cues. Even though it’s not fully audible, there may be a little bits and pieces here and there that help them figure out what that signal is, and go back to the presentation, that’d be great.

Okay, so to illustrate this issue, we tested a few of our participants using contralateral masking that’s shown here in the dark gray, the plug-and-muff technique shown in the red and the direct connect technique in the light gray. The error bars that you see here are the 95% critical difference ranges from Thornton and Raffin. Mask scores are significantly lower than the direct connect scores and plug-and-muff scores for most of the subjects, and some of these kids really really struggled as soon as we introduced that noise. Plug-and-muff scores are similar to direct connect scores, although you can see subjects two’s plug-and-muff scores are close to being significantly higher. So the next question is: how can we be so sure that the direct connect technique is equivalent to sound field testing? There is literature that is out there supporting this method in terms of using it for telehealth, but we tested a few of our long-term bilateral SONNET users just to double check. They were tested unilaterally with CNC in the sound field at 60 dB SPL as part of their annual visit. We then took that same processor and connected it to the same computer using the direct connect technique and tested them with a different list. No one had a significant difference and that helped us feel
more confident in our choice of direct connect testing for our protocol. Here we have the CNC word score results for just the 11 children who have completed the 12-month postop interval. On the left are their preoperative scores with a traditional hearing aid all the way through the 12-month interval. In the purple here are 12-month CNC word scores for matched peers from our clinic database who have similar preoperative hearing histories and were implanted at a similar age. So mostly these are children with progressive losses who waited a little before implantation or they're bilateral cochlear implant users who waited before they received their second side, so they had good language skills at the time this ear received an implant.

On the far right in gray are the scores for the trial participants' normal hearing ear at 12 months showing that they were able to complete the task. So there is a significant difference between scores and specifically postop testing shows that at three months we don't have significant improvement yet, but we do by six months, nine months, and 12-months postop. Interestingly there's no significant difference between the CNC word scores of the unilateral hearing loss ears and these matched typical cochlear implant recipients. There is a child here at 12 months whose score had dropped down to 0%. Her device use decreased significantly between nine and 12 months as well and we're hoping to see that rebound by 18 months, but we'll talk about her a little bit more in a bit here. So moving on to localization. This is how we get it done. The kids wear a supercool adjustable headlamp and sit happily in the center of our 11-speaker arc array, just like this smiling young man. They actually do get really excited to do this task. So here's an illustration of our arc on the left and on the right is a photo that looks a little something like this before we turn the lights out. So the speakers are 18 degrees apart, each with a picture of an animal on top. We turn out the lights and present a 200 millisecond speech burst at 70 dB SPL from a randomized speaker. The child turns their head towards the sound, lighting up their respective animal and we record what speaker they turned their head toward. There's an assistant in the center to provide encouragement and help direct them and center their head and light back between trials to look towards those. You see there's little magnets up here. Those are tar heels.
there 'cause North Carolina. We have them look at the tar heels and wait till they hear the next speech burst, and on the next slide we have a little video showing you what it looks like and you'll see Doctor Gagnon centering the child, but this child is not from North Carolina so he calls them shark teeth. He didn't know what they were. I know it hurt Doctor Gagnon's heart a little bit, but here's a brief snippet of what it looks like. There's a little shot with the lights on, and then moving towards turning the lights off.

- All right so it's kinda hard to tell, make sure, there you go. All right tell the shark teeth, good job! Good job. All right, back to the shark teeth. Good.

- [Doctor Park] Great, thank you. So it's pretty simple and the kids really do look forward to doing it. They have a good time. We found that turning the lights off really helps get away from some of the, "Oh, I really wanted to look at the cats, "so I'm going to pick cat this time." All right so here our localization outcomes and I'm including all subjects because we're really excited about it. This is RMS error, so lower scores are better here. Open symbols are device off and closed are device on. We do record this measure each visit, device on and device off, because we wanna be able to account for any possible developmental effects that may be happening here. So scores are significantly improved, device on versus off, as early as three months postop, and this continues at nine months. We're even better at 18 months. What's really exciting is that most of the kids are starting to approximate the skills of age-matched normally hearing peers after 18 months of use. You'll notice that we aren't seeing improvement over time device off, so it doesn't look like this is a developmental thing. This is actually watching localization skills develop over time with the cochlear implant, and it's fun to watch how this changes over time and I'll do my best to illustrate it here. On the top you see the picture of our arc and on the graph you see a bubble chart. The red bubbles are responses with the device on and blue are with device off. So on the x-axis here, we see the source speaker degrees, and on the y-axis, we have their response. The bigger the bubble, the more responses for that particular combination of source and response. So the line of agreement is on the diagonal. Responses there
would be spot on. So for this child, device off, the blue bubbles, he thought all of the responses were at minus 90 degrees no matter where the source was, and his left ear is his normal hearing ear. So he was constantly turning towards the left when his device was off. With device on, he can at least lateralize a little bit or tell us which side the sound is coming from. So when the source was at 90 degrees all the way on the right, he could tell it was at least on that side of the arc. When it was closer to the middle, he defaulted back to that left side, but he starts to improve with more red dots getting closer to the line of agreement at nine months. Device off, he gets a little bit better also, so he's learning to pick up on some cues there and use them even with the device off. He's really only lateralizing when he's not wearing his implant though. He's even better device on at 18 months and he's still just lateralizing for the most part with the device off.

Here's where we are after 24 months of use, pretty close to spot on when he's using his implant. So moving on to look at speech in spatially separated noise. We are testing the children using the BKB-SIN with speech and babble located in front, speech front and babble to the CI side, and speech front noise to the normally hearing ear. We do have to move the child depending on where we need the noise to be directed and since we wanna be able to account for any developmental effects, again we test both device on and device off at each test interval. So we're looking for spatial release from masking and here are some average results from age-matched peers with normal hearing to illustrate. Threes in the BKB-SIN and we're getting SNR 50 results, so a lower score is better here, indicating that the child can tolerate a lower signal-to-noise ratio in that condition and still understand 50% of the sentences. So the gray in the middle is the SNR score when speech and noise, which in this case is a four-talker babble, are co-located in front. This is the more challenging condition. When noise moves to either side, we would expect to see that spatial release from masking. Score should improve as they do here. The bar in blue is when noise is moved to the right side and the black when noise is moved to the left. We wanna see a difference on the order of about two dB at least and we expect to see similar improvements when noise...
is moved to either ear. This is what we hope to see develop in children with unilateral hearing loss who are using a cochlear implant. Here’s what we’re seeing thus far with this early cohort of children. The chart on the left are the scores at six months and on the right are scores at 12 months. The striped bars are with the cochlear implant off and the solid are with the cochlear implant on. Here in the center are the speech front noise front scores, in blue are the speech front noise to the normal hearing ear scores, and in black are the speech front noise to the cochlear implant side scores. Overall they’re performing better CI on versus CI off, so device use helps with hearing in noise. At six months we’re not seeing much spatial release from masking in either condition. What we are seeing is mostly due to the head shadow, so noises on the cochlear implant side, and they’re making use of that normal hearing ear that’s on the side that ends up with the more favorable signal-to-noise ratio because the head is blocking that noise. This happens with or without the implant. We aren’t really seeing spatial release from masking when noise is to the normal hearing ear, the sixth point, but you’ll see that at 12 months, we’re starting to see that spatial release from masking when noise is directed to either side with the cochlear implant on.

When the processor is off in the striped bars, we aren’t seeing release from masking when noise is directed to the normal hearing ear, but there’s a larger improvement in scores when the device is on. It’s very promising and exciting stuff to see that the cochlear implant is really helping these kids hear in noise, but if you take home one message from this talk, let it be this: this population is different. We need to create specialized test protocols for these children in the clinic. They don’t come to us for an implant because they wanna understand single words in their deaf ear in isolation. That can’t be our primary metric for these kids because that metric so far correlates with absolutely nothing that these families are hoping to see a cochlear implant do for their child. Here are some scatter plots where we try to see if there’s any connection between that isolated CNC score and a number of our outcomes of interest. Thus far CNC word scores in isolation are not correlated to localization, they’re not correlated to listening effort or any of our SSQ results. We’re not seeing any correlations with fatigue.

continued
or hearing in noise benefits. So remember the child with the 0% word recognition skills at 12 months? Well, here she is right here. She has monstrous benefits in noise and she's seeing a ton of spatial release from masking. She's showing benefits from head shadow, summation, and even some squelch. She has very little report of fatigue as well. So if we went on CNC word scores alone, we would say that this child receives no benefit from her cochlear implant when clearly that is not the case. It's really important to be able to measure and demonstrate these spatial hearing outcomes. In our clinic we're implementing spatial hearing-in-noise protocols by calibrating speech perception stimuli in two corners of the booth as illustrated earlier. We're trying to carry those sorts of measurements over into clinic use for this population of children. Programming considerations. Unilateral malformation cases may benefit from some advanced mapping. One of our Mondini kids, we ended up needing to do some pitch ranking and some comparisons. He just wasn’t making the progress that we expected and we did end up turning off a couple of electrodes and it got him on track to do some better word recognition, and he started enjoying his implant more as well. When you’re measuring Ts, go ahead and plug that normal hearing ear. These kids have really learned to use all sorts of cues to figure things out. I'm a fan of measuring Ts even with the MED-EL device and these kids will hear you click a mouse and they'll figure out that you did something, and then they will respond.

So go ahead and plug that ear when you're measuring Ts and also they are easily distracted by noises in the hallway. It’s hard to remember that they can hear you when you're so used to seeing kids who have hearing loss in two ears, and don’t be afraid to use some unique methods for scaling. ESRTs are your friend. Some of our kids in the study have really had a hard time setting their MCLs. Some have overset them. I think that they are equating loudness with more understanding, and so they've been really turning up, turning up, turning up their sound. Don't be afraid to move more quickly than you would with your typical cochlear implant population. I know sometimes we like to keep things turned down in the pediatric population and move slowly through programs, gradually introducing loudness, but these kids can really tolerate things a lot
more quickly. They know what sound is. We also have created some of our own scaling charts to measure our MCLs. These are kids who know what sound is. They know what it means for a sound to be soft. They know what it means for a sound to be loud and they know what that means in the context of typical hearing, so not through an electrical dynamic range. So we’ve created some of our own charts to introduce these concepts to some of our preschoolers. I’ll show you what one of them looks like. You don’t have to be a good artist to make this happen, as you can see. So we’ll have the kids talk about what is a soft sound to them. So for this kid, it was a whisper, so we drew a picture of whispering. What is a comfortable sound? Well, they watch Pepper Pig on TV, so we drew a little picture of Pepper there. What’s a loud sound, something that’s allowed, but it doesn’t hurt you? To them, this was a hairdryer, and what’s something that is loud and hurts your ear? Well, that was his sister yelling, and I have heard the sister yelling and I would have to agree with him on that one. So for him, we work off of this scaling chart because this makes a lot more sense to a kid who knows what hearing is in acoustic dynamic range than the monkey charts or things that we typically use with children who have cochlear implants.

So in summary just to go back over some of the things that we’ve talked about: Clinical considerations. When you’re looking at threshold testing, be sure you’re being aware of what the normal hearing ear is contributing to the sound field audiograms when you really want to know what they’re detecting just with their cochlear implant alone. For word recognition testing, make sure that you’re isolating that cochlear implant ear, preferably with a direct connect system. That’s what we like to use here. Another clinical consideration when you’re thinking about programming, block the normal hearing ear when measuring those thresholds. Don’t be afraid to think outside the box when you’re doing scaling and don’t forget about ESRTs. They have been a really great tool for this population. When you’re dealing with a contralateral ear that doesn’t have a cochlear implant and is typically pretty healthy, you can get some really robust, really nice ESRTs to help aid you in your programming. Spatial hearing. I encourage everyone to incorporate some form of testing that takes spatial hearing into account.
Each of these children have unique goals. You may wanna tailor your test battery to take into account the goals for that particular child. Sometimes even just adding some questionnaires could be a good helpful tool for you as you’re trying to work towards getting some sort of a spatial hearing protocol going. Outcomes for word recognition. Isolated speech understanding is improved with this cochlear implant, but it is not the be-all and end-all of everything. We’re not seeing a whole lot of correlation to other outcomes thus far. Localization improves over time with cochlear implant use in children with unilateral profound hearing loss and many children are approximating the skills of kids with normal hearing by about 18 months of age, 18 months of use, sorry. Hearing in noise. Hearing in noise generally better cochlear implant on versus cochlear implant off. Spatial release from masking is becoming evident when noise is directed to either side by 12 months postoperatively. Parents report that their children are exerting less effort to listen over time with the cochlear implant, which is very exciting to see. All subscales of our quality of life data on the Speech Spatial and Qualities data, parents are noting improvement over time. This is my contact information if you needed to get in touch with me or if you had any questions, more than happy to answer those. It looks like we have plenty of time for any questions that you may have.

- [Carolyn] Thank you so much, Doctor Park. We have quite a few questions already queued up and I would encourage our participants to enter any others that they have. We'll start at the top. Jacob asked, "At the beginning you mentioned "that MED-EL is not FDA approved "for single-sided deafness for under five. "Could you talk a little bit about that?"

- [Doctor Park] Yes so the FDA approved the MED-EL SYCHRONY system for single-sided deafness for children who were five years of age and older. It’s mainly for adults, but they went down to children as young as five years of age with 10 years or less of deafness, of profound deafness in that ear. So they didn’t go under five years of age for their FDA approval.
- [Carolyn] Great, thank you, and then Kristi asked about, when you were talking about your equipment setup, "What is the name of the splitter that you use?" I don’t know if you wanna go back to that slide or you just know that off the top of your head.

- [Doctor Park] No, that’s a good question, and I’m not sure off the top of my head, I just, so let’s see, unless there’s a picture of it somewhere. I honestly just went to Amazon and I searched for splitter with two separate volume controls. Let me see if we can see it here, oh, there it is. It’s iLuv. Now that I see that little bit, I can remember seeing it. So it’s I-L-U-V.

- [Carolyn] Oh, that’s awesome. Thank you for grabbing that. The next question is from Lisa. She said, "Are the 12 month spatial release "for masking scores with cochlear implant on "statistically significant with the smaller cohort?"

- [Doctor Park] So far, not yet. So the statistics that I ran I used the six-month and the 12-month data together and I used time as one of the factors and so I’m not seeing any sort of an interaction effect just yet. I’m not sure what it would look like if I looked at just the 12-month data. So right now it’s looking mostly like they do better overall device on versus device off and overall they’re doing better when they’re benefiting from a head shadow effect.

- [Carolyn] Jane asked, "Do you know what the status is "hearing loss in children?"

- [Doctor Park] I do not know. I do know that typically cochlear implant companies tend to grow off of each other in terms of new indications, but honestly I’m not sure what the other companies are doing in terms of even if they’re pursuing approval under that indication, but that’s a really good question.

- [Carolyn] Yeah, I guess you would expect it, just like the hearing aid industry that when one company comes out with an innovation, then you kind of see it go through,
but I would encourage Jane to contact those other companies directly maybe and ask if there's anything they can share. Jen asked, "How do you counsel," yeah, "how do you counsel parents who may hear "from other professionals that one ear is good enough?" Good question.

- [Doctor Park] Yeah, well, when they come to us typically, when they get so far that they’re coming for a cochlear implant visit they’ve kind of already noticed that their child is struggling or they’re concerned that they may start struggling. I think one of the things that we don’t know yet is if implanting children earlier will head off some of those educational concerns that they see down the road. So it’s tricky for a lot of families is that they see their three-year-old who doesn’t seem to be struggling and seems to be doing just fine, but we don’t know if once they enter school if they’re going to start struggling. So we talk about what the evidence shows in terms of what kids struggle with in school when they have unilateral hearing loss and really what we talk about a lot is that there is a risk that they would lose hearing in their other ear. So we don’t want to to find out that they lose hearing in their other ear when they’re 10-15 years old and they’ve gone 10-15 years without having hearing in that first ear. If we were to implant that ear at that point after 10 to 15 years of deafness, then they would really struggle and we wouldn’t be able to get them hearing with two ears. So that’s something we talk about a lot in terms of if you want binaural hearing, we should get binaural hearing going as early as possible in case they lose hearing in that other ear as well.

- [Carolyn] That was something I, you mentioned in the presentation as well and you gave a statistic and I remember thinking, wow, that’s higher than I thought. Do remember what that was?

- [Doctor Park] Yeah, it’s 40%. There’s some work that’s been done out of Canada and they’ve been able to look pretty systematically at some of these kids especially when it comes to the CMV data because they are better able to track that sort of thing and
Guthrie cards are kept for longer so they can go back and look and see if there's any evidence of congenital CMV, but yeah, it happens. It happens more frequently than you would think.

- [Carolyn] Reagan asked, "Were all the children aided pre-cochlear implant "if some had "long durations?

- [Doctor Park] Yeah, very few actually were aided. There was only one child who came to us with a traditional hearing aid and he had had a progressive hearing loss. There were a couple of kids who had been using a bone anchor device. None of them have been using CROS, but for the most part, none of them had really had any sort of amplification. We did test them with a clinic hearing aid for purposes of the study. That's where those aided word recognition scores came from.

- [Carolyn] Really great questions from everyone here. So this is from Jennifer. She said, "Can you comment more about FM "not being useful with unilateral loss "by not giving binaural cues?"

- [Doctor Park] So they are useful in that they are going to increase the signal-to-noise ratio. So they are going to help the child hear the teacher a little bit better. I think what is tricky is that you have to balance putting a receiver on the normal hearing ear and keeping it un-occluded so that they can hear their classmates, so that's concern number one. Concern number two is you're still not sending sound to that deafened ear. You're only sending sound to the normal hearing ear. So we're not using two auditory pathways, which is what's gonna help you be able to know where sounds coming from and to hear a noise. So it's gonna give you a little bit of a boost in terms of hearing the teacher's voice, but it's a balance in terms of making sure that they're hearing the teacher's voice and still able to participate in classroom communication.
- [Carolyn] Now Jacqueline has another question that I was wondering if it would come up regarding reimbursement. "Typically insurance companies deny "cochlear implants for single-sided deafness. "Do you have any ideas or expectations "on how long it will take them to catch up?"

- [Doctor Park] That's a good question. Sometimes it does take them a little while to catch up. That's where peer-to-peer can often come in to play, and then one of the physicians or the surgeon can talk to the insurance company and explain, "Look, this isn't experimental anymore," and that can help, at least as long as they're five years old. In North Carolina, Medicaid covers single-sided deafness cochlear implantation. So we're very lucky in that aspect and that actually helps us be able to go to other insurance carriers and say, Look, if you have Medicaid here, you can get this, and that helps. Also if you wanted to check the ACIA website, American Cochlear Implant Alliance. We wrote a white paper that talked about all the evidence that is out there that supports cochlear implantation in single-sided deafness in hopes of helping families who are trying to seek insurance coverage. We do have some trouble. It seems to kind of ebb and flow. We'll get a whole lot of approvals, and then all of a sudden it'll be just Blue Cross Blue Shield is all of a sudden not covering and or it seems to ebb and flow, but we have had a whole lot of luck getting coverage for kids, getting insurance coverage for kids.

- [Carolyn] That's really good to hear and I don't work in cochlear implants, but my understanding is that the cochlear implant companies, I'm assuming, MED-EL has a reimbursement department that works on that. Is that correct?

- [Doctor Park] They do, yes, they do. All the cochlear implant manufacturers have reimbursement contacts who can help families navigate those waters a bit.

- [Carolyn] Great, we have a question here from Dori. "Are you seeing any difference in outcomes "between children with congenital SSD "versus acquired progressive?"
- [Doctor Park] That's a good question and that's something we really wanted to look at, but like I said, some of these kids, it's really hard to know when they really lost their hearing. Some of these kids were acquired and we really expected them to pick up more quickly than they did. I think some of it was they tended to be a little bit older and were having a hard time with full-time use right off the bat just because they didn't want to look different, those sorts of things, but once they started wearing it more, they took off a little bit more quickly, but so far we're not seeing anything come out in terms of single word recognition for congenitally deafened kids or acquired and certainly not seeing a whole lot in terms of localization or hearing in spatially separated noise either. I will say that the malformation kids, those two malformation kids have taken a little bit longer to catch up, but they certainly are catching up to the rest of the cohort.

- [Carolyn] Great, great question. There's a couple of questions here on setup. Before I get to that, Reagan asked, "How is habilitation provided to children? "So is it direct connect? "And was specific habilitation provided for localization?"

- [Doctor Park] Oh, those are really, really great questions. Yes, so every kid in the study does get auditory therapy. It's mostly done through tele-therapy-like models, some of it is done in person, but really we take big advantage of teletherapy models. It is done via direct connect. They follow a hierarchy based on the Auditory Learning Guide. If you wanna go to our website at childrenscicenter.com, there is a link there. They have their own webinar talking about how they do the habilitation. So all the kids did get auditory therapy for twice a week for the first six months of the study, I mean, sorry, twice a month, every other week for the first six months of the study, and then once a month for the next six months of the study. We did not include any localization as part of the habilitation, but we have been encouraging families to do some of it at home, which has been really easy and fun for them to do. They do some games like find your sister, find the phone, hide and go seek with just sounds, those sorts of things, and I do think that that does help working on those skills at home. Some of the
kids have gotten to a level where they are working on some hearing in noise in therapy as well.

- [Carolyn] Great, and then we have two questions on setup. One is, "How do you set the ESRT or how do you use it?"

- [Doctor Park] So ESRT, so for our Electrical Stapedial Reflex Thresholds we use the protocol that MED-EL has. You can get that. They have a pamphlet you can get from them as well. So we set it up so that we are measuring their reflexes from their contralateral ear and we have them watch a movie while we're measuring it, which works really, really well. All they have to do is sit quietly. We measure that ESRT and we use it to set their MCLs. I will say that we use it as a starting point. Most of the kids, after we measure their MCLs, we have gone live and have them try to see if it balances out to their other ear. Does it sound just as loud? Does it need to be a little bit louder? Most of the kids have turned their MCLs up a little bit above that ESRT level, but we use it pretty early on so we have an idea of where we would expect their MCLs to fall.

- [Carolyn] Great and then Dori asked about direct connect speech discrimination testing. If you could go into a little more detail about how that is set up.

- [Doctor Park] We have a laptop that's connected to our audiometer and I just pull that laptop off of that exact same laptop off of the deck and I go into the booth with the child just 'cause it's a little bit quieter in there, and then I connect them directly to that very same laptop.

- [Carolyn] And then the last question we'll take here is from Claudia. I just wanna thank our participants again for all these great questions. Claudia's regarding bone conduction headphones. "Is there anything specific you look for "if somebody wanted to purchase those?"
- [Doctor Park] I wanted something that were wired versus the wireless kind. I do have some wireless kind that I have for personal use from AfterShokz, and so I like those. So I got the AfterShokz brand that plugs in. You do have to remember to charge it. It does have its own on and off and power button there. So you have to remember to charge it, but I knew I liked them and it worked for me, so I just use that.

- [Carolyn] Great. Thank you so much, Doctor Park, for this extremely informative presentation and I think we're gonna have to do a follow-up soon because this area's changing so quickly. So maybe next year foresee what is new in this area, but it’s so great that you're working with it and can provide so many tips and tricks, as well as all this great information. So thank you very much. It’s been great. For all of our participants, thanks for logging in, for all your great questions. Hope to see you in another webinar online soon and again if you're earnings CEUs, remember to login to your account and take the exam. It will expire. So if you're earning CEUs, you're gonna wanna do that soon. Thanks, everyone. Have a great afternoon.