American Cochlear Implant Alliance Task Force Guidelines for Determining Cochlear Implant Candidacy in Adults with Single-Sided Deafness or Asymmetric Hearing Loss

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- At this time, it's my pleasure to introduce Donna Sorkin. She's the Executive Director of ACI Alliance. And as I mentioned a couple weeks back, somebody who has done more for cochlear implants than many people alive today. It is a pleasure working with her in this organization. And I'll turn it over to Donna.

- Thank you so much, Carolyn. It's really a pleasure to be back with AudiologyOnline today for this important session. And the session today, as Carolyn said, is on candidacy in adults with single-sided deafness or asymmetric hearing loss. Here's our presenter disclosures and our learning outcomes for the course. And this is on taskforce guidelines for Cochlear Implant Candidacy, which is part of a four-part series that we're doing on four populations in adults and children. So, you may wonder who we are and why there's been another organization created in hearing health. And we are a special organization that is membership-based and with members from across the care continuum, such as our speakers today, an audiologist and a physician, and we also have speech language pathologists and educators, and others on CI teams, as well as adults and parents.

We have a website that's really designed for people both in and outside of CI. And a lot of people from the general public and primary care physicians. Those in hearing healthcare, not in CI, come in and use that. And it's really designed for you. We are highly collaborative with other organizations like AudiologyOnline, and that's in our name Alliance. And we welcome your involvement. So, please do get involved. And we've given you our website there as well as Facebook and Twitter. So, we're very active on social. Please follow us. Our mission is to advance access to the gift of hearing provided by cochlear implantation through research advocacy and awareness, and to specifically address factors that contribute to under utilization of cochlear implants.

Very important to us is to improve awareness regarding candidacy and outcomes. And that is, in fact, our objective today, to share information on how CI candidacy is determined for a specific population. It's interesting that there's a set of criteria that are part of the FDA indications, but depending on the clinic, there are different ways of determining candidacy. And so, these four task forces recreated to look specifically at how we think best practices are carried out and see what those four topics are on adults and children. And we hope that by improving knowledge about candidacy, we will improve the referrals for CI from hearing healthcare professionals and others. Because right now, we're still hovering around five to 8% of US adult candidates.

Probably even lower if you include those who are candidates by virtue of the fact that they have single-sided deafness. So, this particular taskforce today will be looking at SSD in adults, but there are other talks that have been given on AudiologyOnline that you can view and see for those other populations. So, with that, I'm going to move into introduction of our speakers today. We're very honored that these two important clinicians and scientists could be with us to speak today. Margaret Dillon is an audiologist and Associate Professor and Director of the Cochlear Implant Clinical Research program in the Department of Otolaryngology at the University of North Carolina at Chapel Hill. And she is especially interested in new indications for cochlear implantations, such as SSD.

Dr. Matthew Carlson is a Professor of Otolaryngology at the Mayo Clinic in Rochester, Minnesota. He is the Cochlear Implant Program Medical Director. He's also involved in a lot of public policy issues related to CI access, and is the co-chair of the Hearing Healthcare Collaborative with works with clinicians and others from across the care continuum and in and out of cochlear implants. So, he has a wide range of interests, and we're really delighted that he would be involved in this issue of SSD. So, without, I'm gonna turn it over to our speakers today. Thank you so much for being with us.

- All right, thank you, Donna. So, we first would like to acknowledge our collaborators on this project. So, as Donna mentioned, this was some work conducted by a task force to look at the current literature on, and understanding of what's happening in the clinic with patients that have single-side deafness and asymmetric hearing loss. And so, we have collaborators from the Mayo Clinic, New York University, University of Miami, and WashU. And so, we were thrilled to be able to work with this team, and the work that we did together is what we are sharing today. First, we'll talk about the influence of single-side deafness and asymmetric hearing loss on speech recognition and spatial hearing, as well as quality of life.

And then introduce this idea of cochlear implantation as a treatment option, as well as go over the candidacy considerations for adults with single-side deafness and asymmetric hearing loss. And then talk about the post-activation management of cochlear implant recipients that have been implanted by this indication. And finally end with the ACI Alliance guidelines for adults with single-side deafness and asymmetric hearing loss. So, we know that patients that have single-side deafness, and so, in this case, we are defining that as a moderate to profound hearing loss in one ear, and normal or near normal hearing thresholds in the contralateral ear. For asymmetric hearing loss, the definition for that is a mild to moderate hearing loss in the contralateral ear.

So, globally, we can think about these cases as all cases of asymmetric hearing loss or unilateral hearing loss. And we know that these patients have difficulty with spatial hearing, meaning poor sound source localization, identifying where sounds are coming from. And then limited speech understanding in complicated background noise or complex maskers. And that's because if you're listening with one ear alone, we can't take advantage of these binaural benefits. This also results in poor quality of life and was observed in patients that have normal hearing bilaterally. And what we see is that these patients report the poor quality of life on these subjective questionnaires, as well

as an increased report of hearing handicap. Historically, the treatment options for patients that had single-side deafness or asymmetric hearing loss have included no treatment.

I think we can all remember a time where we would tell patients that one ear is good enough. And we now know that that is not true. Depending on the hearing loss and the impaired ear, they could be fit with a conventional hearing aid, but more popular are the contralateral routing of the signal hearing aids and the bone conduction devices, which pick up the signal from the poor hearing ear and bring that over to the normal hearing ear or the better hearing ear. So, it's all going up one auditory pathway. And even though this is providing patients with the sound from the impaired side, they have an increased sound awareness. The performance benefit is very limited.

And so, we can see poor performance on these facial hearing tasks, such as localization and speech recognition and noise. And often, the reason that these patients are pursuing technology is because of difficulty with spatial hearing. And so, if they're not getting that benefit with these routing technologies or re-routing technologies, then there is a poor associated quality of life with that as well. So, when cochlear implantation was first thought of for these cases, it was little bit controversial because the idea really was, can you take this crude sound quality from a cochlear implant, and can you fuse it with what you're getting from your contralateral normal hearing ear? So, even amongst our team, there were team members that said, you know, "This might be too far for cochlear implants.

Why would anyone want to listen to a cochlear implant when you have a normal hearing ear to compare this to?" And so, the big question was, can we integrate these two signals, this cochlear implant electric signal with what we're getting from our normal hearing ear to have improvement on spatial hearing, and ultimately, subjective benefit and quality of life. The history of cochlear implantation for cases of single-side

deafness is really interesting. So, it started with patients that had incapacitating tinnitus in the impaired ear. So, they really were looking for a treatment for the tinnitus and not necessarily an improvement in binaural hearing. And what they found was that patients that received a cochlear implant had the significant reduction in tinnitus severity, but that they also were reporting and then demonstrating improvements in spatial hearing abilities.

So, speech recognition and spatially separated noise, and these complex listening situations, as well as sound source localization. So, that got the field very excited about cochlear implantation as a treatment for patients with single-side deafness who were pursuing it for improvements in bilateral or binaural hearing. And then, comparisons of cochlear implant use with these alternative technology. So, either rerouting technologies, or in an unaided condition showed that there was superior performance when listening with a cochlear implant as compared to these alternative treatment options on measures of spatial hearing. So, sound source localization, speech recognition, noise, and as well as reported improvements of subjective benefit and quality of life. And so, these findings were so compelling that in 2013, the EU approved single-side deafness and asymmetric hearing loss as indications for cochlear implantation for children and adults.

And then in 2019, the FDA approved these indications for children and adults five years of age and up. So, what we know from the current evidence is that cochlear implantation is an effective treatment option for adults with single-side deafness. And if you're interested in the literature on children, that will be presented next week by our colleagues, Doctors Park and Young. And then there are special considerations for candidacy for this patient population, as well as test methods and mapping that we need to think about so that we're providing the best care for these patients. But we can expect benefits of cochlear implant use in these patients to be improvements in speech recognition in the impaired ear. So, when listening with the cochlear implant, as well as

improvements in spatial hearing abilities when listening in the combined condition, tinnitus suppression, and improvements in subjective benefit as compared to their preoperative abilities. And I'll turn that over to Dr. Carlson.

- All right, thanks a lot, Dr. Dillon. I just wanna take the brief opportunity to say thanks to AudiologyOnline for allowing me to share the stage with these two incredible thought leaders in cochlear implantation. I think Dr. Dillon is a foremost expert in this topic and has really already changed largely from her work came the labeling expansion, which is really incredible. And Donna, of course, is just a huge advocate and leader for policy change and everything. And so, thank you to both of you. With the background of being a surgeon, I'm gonna present some aspects that I think are, hopefully, relevant to most of the audience, primarily being on the audiology side. If I go through something a little bit more quickly, it's because I think it might be less interesting to you.

But if it is something that you have questions on afterwards, I'm certainly very glad to circle back and expand on different topics. But, at least at this part, I'm gonna primarily talk about candidacy considerations. And when you think about that, I think broadly, there's kind of three questions and the answers often align, but they don't always. And the first one is, can you do it? So, is it safe to do? Is the patient reasonable of medical surgical candidate? And when you say, can you do it, you're also wondering. You need three elements for a cochlear implant to work. You need something that resembles a cochlear. You need a cochlear nerve of some sort, and you need to have brain that is able to process the signal.

And so, that's the, can you do it? Should you do it? I think, balances whether you think that the benefit outweighs the risk, that the patient's gonna be better off on the other side for having done it. And then, will it be covered? So, we did see that the FDA labeling has expanded for two of the companies now, or two of the cochlear implant manufacturers. But, insurance often is delayed. And so, it doesn't necessarily mean it'll

be covered. Fortunately for most people and most situations, it is covered, but not completely. And so, we often look at prior authorizations. Still in Medicare, you cannot implant based on my interpretation of Medicare criteria. You cannot implant SSD at the current time.

So, those are the three questions, and we'll kind of work through those etiology, potential contraindications. And I always like the word potential before contraindications, because you'll see that that's often very nuanced and patient-specific. And what would be a contraindication in one patient might not actually be in another patient, depending on the whole clinical picture. Imaging the impact of duration of deafness for SSD, which I think is fundamentally different than patients with binaural hearing loss or more significant, binaural hearing loss. Age at implantation, tinnitus severity, which is really fascinating to me. We'll talk about that in a little bit. And then how it compares and contrast to other hearing technologies where you might get out of the different ones, and then patient benefit and quality of life.

So, you can probably advance slides. So, broadly, the etiology of SSD is different than the adult population, the pediatric population. In the adult population, by far, the most common cause of single-sided deafness is sudden sensory hearing loss. So, adult acquired sudden sensory hearing loss, and most of those cases are idiopathic, meaning you do the whole workup and you don't find any other cause. Certainly, there is a cause for it. There's a cause for everything, but we can't clearly identify, and it's something probably on the molecular level. It's not structural where you can see on a scan, or do a test and easily identify it. The other groups that are common in the adult are reasonably common that make up the other maybe 50% or lesser, Meniere's disease, trauma.

So, temporal bone trauma to capsule fractures, iatrogenic, or retrocochlear pathology. So, oftentimes, a vestibular schwannoma cause a single-side deafness or the

treatment of a vestibular schwannoma will accelerate that hearing loss in that affected ear. And then you can have an adult with single-side deafness because they were congenitally unilaterally deaf. So, they could live their whole life and develop into an adult and have it then. But that's a smaller number, at least that we see. Most of those patients have grown accustomed to their unilateral hearing loss. They don't present. Most of the people we see are the people that have an abrupt change. So, they were living their life binaural hearing and they had a major disruption in their life.

They lost hearing in one ear and they might have concomitant symptoms of tinnitus, maybe vertigo from labyrinthitis, that sort of thing. And they're coming in because it's a big change and they wanna find out what caused it, what they can do to improve it. In the pediatric population, the most common reasons you have SSD are you were born with it, either idiopathic, or from a genetic cause, you could have a labyrinthine malformation, or actually what's important to recognize is a significant proportion of patients in the pediatric population have cochlear nerve hypoplasia or aplasia, which reduces outcomes, or might even make it kind of a contraindication in some cases. And then an important one that's developing in the pediatric group is CMV.

And the idea is you could develop a unilateral hearing loss in that population, but then later on in life, you could acquire contralateral hearing loss. And so, the consideration for SSD implantation, that group is even greater, I think. When you think about your etiology, I think it's really important to understand what the likelihood of developing hearing loss in the other ear is from that driving etiology. So, if it's idiopathic, sudden sensory hearing loss, a very small number actually will acquire hearing loss in the other ear and the estimates are less than 1%. So, if somebody, you know, woke up one day and had sudden hearing loss in one ear and they're devastated by it and they ask what the risk is in the other ear, it's about 1%.

Meniere's disease is 30% or higher, depending on how long you find them. You might have an adult with bilateral EVA or something like that, but usually they're gonna be asymmetrical. It's usually rare that they'd have normal hearing into adulthood in one ear. And then the NF2 population, they're destined to have bilateral hearing loss if they follow the storyline for 99% of people with NF2. And so, if you have a person that has unilateral, so, SSD, but the probability of developing hearing loss in the other year is much greater. I think that's a major motivation factor for you to consider at least suggesting the idea of implanting that SSD ear first. And the reason is, you could wait until the other ear goes out, but it's nice for them to gain experience in the one ear before the other ear goes out, and there is some impact.

We don't know exactly what the impact is, but there is some impact of SSD, or sorry, of duration of deafness. So, if you wait too long for the other ear to go out, the first ear will be deprived from auditory stimulation for a decade, two decades, three decades maybe, and then you've maybe changed your outcome in that SSD ear. So, etiology is a critical feature. The other thing about etiology is if it's a sudden sensory hearing loss, you have to understand the natural history of that disease and not intervene too early for a lot of reasons. But most of the people with adult SSD have idiopathic sudden sensory hearing loss. It's important to understand that 1/3 of those patients will return to normal in that ear, or very close to it.

About 1/3 will have partial recovery, and 1/3 will not improve at all. And there are some prognosticators for improvement, but they're imperfect and there's no way to know if they're gonna return or not. So, if you put a cochlear implant in too early, you have taken away their chance to recuperate or recover their natural hearing. And the general rule of thumb is it's very rare a sudden hearing loss will return after three months. So, at three months, it's pretty safe to assume that it's not gonna come back, but to be safe, most people say, "Well, you should probably really wait at least six months to give that

margin to really feel good about it." But the other part of that, what's important is not just if it's gonna return or not, but you have to give them a little time to acclimate.

If you have a person who has sudden hearing loss and they come in and they have horrible tinnitus, they're dizzy, and they have hearing loss, and you talk to them about all the options, they're gonna do everything at that time to return to their pre-morbid state. They're gonna want... It's gonna be difficult for them because they haven't acclimated. And they're in that very acute period. And by giving them time to acclimate, you'll find out some of the patients will find out that they're actually doing okay and they might not wanna push rehabilitation. If you look at the major, so, you know, the prevalence of SSD varies based by study, but it's probably something like 0.1%. If you take the entire world with SSD and the number of people that actually have a cochlear implant right now, it's a very, very small number.

And that's because it's a growing indication in many different factors. But just to say, it's not crazy to not rehabilitate SSD if the patient's acclimating really well and they don't feel like it's a major quality of life modifier. I think you need to look to the patient and figure out what's best for them. But giving them at least at six months to a year to acclimate to the hearing loss, to consider other technologies, and we'll talk about that. They're not great, I think, compared to a cochlear implant, but we can consider those other technologies. And also tinnitus changes in the first six months. And in my opinion, that's a major driver of whether or not a birth consider getting a cochlear implant for SSD.

There's this idea that for tinnitus, the idea is generally the six-month benchmark. Meaning if you just have a sudden onset tinnitus, it might fluctuate a bit in the first six months. And if it stays persistent beyond six months, it's usually gonna be there for a very long time. And so, that's kind of helpful to have that benchmark in your head. But understanding the etiology and the workup is important. You also wanna understand

the etiology because it might have implications for their prognosis with their implant and the timing of such. So, if they had labyrinthitis as if they had meningitis and they developed an asymmetric hearing loss or something, it might be more time sensitive. They might develop progressive ossification.

And so, you might push to do it earlier. If they have a vestibular schwannoma, and you are gonna treat that, sometimes you don't know if you're gonna be able to preserve a nerve or not, depending on what treatment of modality you go down. And we'll talk about that in a little bit. And if you have a vestibular schwannoma, you also have to consider tumor surveillance long term in whether or not you retain the internal magnet and the impact that will have on your ability to see the tumor for a mater of fact. Fortunately, you're able to do that pretty well, but there are some centers that are reluctant to scan people with retained magnets, and that causes a big issue in disruption in clinical workflow in some situation.

So, it's complex on many different levels, but most of the time, most etiologies do not preclude the ability to gain benefit from a cochlear implant for SSD. So, we kind of talked about those risks of future. I think we covered those thing items pretty well. Potential contraindications. So, basically, we said earlier, you need three things for cochlear implant to work. And the most important one, one of the most important things is the whole clinical picture and patient motivation and what consequences they're having and their goals of what they are hoping to achieve. But backing up from that, just what are some potential things from a physical standpoint that might reduce your ability to give them a cochlear implant or give a good outcome.

So, severe, so, you need a cochlear, you need a cochlear nerve that's healthy and able to propagate a stimulus, and then you need a brain that's receptive to it so it's enabled to process it. So, severe cochlear ossification, white it out cochlear. You can do the drill out procedure, the double tunnel, the double labyrinthotomy, the split array, all these

sorts of things. But the reality is the outcome's gonna be usually pretty poor for a severe cochlear ossification, and in a person with normal hearing in the other ear and only sound awareness in the one ear, they're probably not gonna use it very much. So, probably not a great use of CI for SSD. Cochlear nerve aplasia and cochlear nerve hypoplasia, of course, that's gonna be a pediatric onset hearing loss and they've grown into adulthood and now they still have it as an adult.

If the nerve is completely to be... So, fortunately, MRI is really good at detecting cochlear nerve aplasia. And it's specifically these parasagittal views on a type of MRI called T2 cisternography, also called CISS, or FIESTA, or heavily T2 weighted imaging. They're all the same. It just means that the CSF becomes very, very white and anything else becomes very, very dark. And you can see this contrast on a very nice level. And the parasagittals mean you take the IAC as a tube and you're cutting it this way. So, in cross section, you can actually see four different nerves, the tube, vestibular nerves, cochlear nerve, and facial nerve. And you can actually see, is the cochlear nerve present?

And then you can also see caliber. So, if broadly, the cochlear nerve should be larger than the facial nerve. And we know that from scans and also seeing them when we do posterior fossa surgery. So, if the cochlear nerve is smaller than the facial nerve, it's a good rule of thumb to say that they have cochlear nerve hypoplasia. So, that means it's smaller. And if it's absent, you can have aplasia. If you don't have a nerve, you're not gonna be able to propagate a signal, but I will say, sometimes it's not totally straightforward in the group it's in particular, And I know this is an adult talk, but the charge group in particular, all bets are off for nerve anatomy.

Charge children typically have a smaller cochlear or less turns and they often lack portions of the vestibular system, and their nerves are different. Their nerves, you can have a nerve bundle that does multiple things. Your cochlear and vestibular nerves can

come together. Even your facial nerve can bundle with one of your cochlear nerves. So, they're really tricky to figure out, and you often combine it with CT scan to see if there's a cochlear aperture also. It's more in the weeds than we need to be right now, but I just wanna bring that up. And then retrocochlear tumors, it used to be considered a contraindication. The idea was that if you had a tumor, so, we believe that cochlear implants work because they bypass missing or nonfunctional hair cells and stimulate the cochlear nerve directly.

So, if you believe that that's how they work. That if you had a vestibular schwannoma that was pushing on the nerve, even though you're stimulating the cochlear, maybe the signal can't get through the power cord, but it turns out that the reason people lose hearing in the case of a vestibular schwannoma or retrocochlear tumors, isn't always because of just a problem with the nerve, or critical damage to the nerve. It's actually a combination of things. So, long story short, in many cases, a cochlear implant can work in retrocochlear pathology, and that's, so, auditory neuropathy resynchronizes the nerve retrocochlear tumors. You often develop a super-physiological stimulation that can overcome some of that blockage, that conduction block from having a tumor there, and even conditions like superficial siderosis and some of these more obscure rare things.

It turns out cochlear implants often will work on these things remarkably, which kind of challenges how we understand those disease processes and cochlear implants in general. So, imaging, if you have an adult that developed, so, if they were born with normal hearing and they have normal hearing through childhood, adolescence, and into adulthood, the inner ear anatomy is gonna be normal 99% of the time. It's very rare that you would have a labyrinthine malformation or something abnormal and you wouldn't have hearing loss earlier. And I bring that up to say, because this topic is primarily adults. Most of the time, an MRI is sufficient, and you don't need to get both.

In MRI, we'll see if the cochlear, what the status of the cochlear nerve is, what the cochlear looks like, cochlear anatomy, whether or not the cochlear is patent.

So, there's a lot of conditions that can cause the cochlear to scar shut. The one we most commonly think of is meningitis with labyrinthine ossification. Otosclerosis can cause it. Temporal bone trauma can cause it. It can be iatrogenic. It can be also from chronic otitis media from a labyrinthitis, or if you have a labyrinthine fistula, so, a cholesteatoma erosion causing a labyrinthine, or cochlear fistula or something. Then what's interesting is also for vestibular schwannoma, is there are cases where we attempt to preserve hearing on an acoustic neuroma and we did not drill into the inner ear, but there's a vascular event. There's this artery called the labyrinthine artery, which comes off of a loop of ICA, which is a branch of the posterior circulation of the brain.

There's a single artery that feeds mainly most of the inner ear. And if that goes into spasm or it's injured, it loses its blood supply. So, you have ischemia or loss of oxygenation to the cochlea, and it will actually scar many times. So, even though you didn't enter the cochlea, and you know you didn't 'cause you're operating, you can see two or three. If you lose hearing two or three years later, sometimes the cochlear scar is shut still. So, even in those cases, it's still prudent to get an MRI to make sure the signal inside the cochlear, the fluid is still there so to make sure you can get an electrode in there. CT scan might be a roadmap for surgeons that don't do this procedure as much, and just helps them think about the numerisation of the temporal bone and things, but it's definitely not a requirement in most cases.

But I will say this is nuanced. And I think we probably shouldn't be too prescriptive on saying what people can and can't do. But usually, an MRI is enough, I guess is the punchline. Now, this is where I think it gets, I mean, all of this is super interesting, but this is where it gets really interesting to me. I think, having single-sided deafness, both congenital or later acquired is very different than having bilateral hearing loss with

regard to duration of deafness. And the idea is that the brain, the ears are not independent. They interconnect through the central nervous system. And so, there's crossover and duplication or redundancy in the system. And in many ways, even though one ear is deaf, the brain can still be primed globally.

And so, maybe it doesn't have as much of an impact. It's also like you can't really have a pre-lingual SSD kid, as long as they're otherwise developmentally normal. Like, it does just doesn't apply 'cause you have the other ear and you're gonna develop speech and language. And so, it's just a completely fundamentally different thing. I also think rehabilitation is different. Dr. Dillon knows a lot more about this than I do, but the idea that you have one normal ear and one deaf ear, you can probably use that good ear to coach your bad ear to hear what you can reconcile, what your cochlear implant ear is hearing and what your normal ear is hearing. And you can reconcile those sounds faster.

You have an internal coch, I call it. And so, maybe that affects maybe the timing or rate of getting better, or your performance or whatever else. There's a lot of things that go into it. But I just think it's fascinating. But I just wanna say that right now, I don't think you should withhold a cochlear implant from a patient has prolonged deafness because you're extrapolating data from the bilateral hearing loss population. They are not the same. And until we know more information, look at the whole clinical picture and don't use that as a reason to not implant somebody. There's all these different reasons why we consider implanting for SSD. It's not just sound localization, speech understanding and noise, it's also tinnitus suppression.

So, imagine the person who's born congenital unilaterally deaf and has horrible tinnitus in that ear or whatever. And you say, well, they were born congenitally deaf and now they're adults, they're not gonna good outcome. They might not get a great outcome for speech perception, but you might do neuromodulation with tinnitus suppression on

that side. So, there's so much things we don't know yet. And I think until we, in a situation where the risk of the procedure is extremely low and the benefit might be there, I don't think we should be restrictive. I think we should be more exploratory safely to understand, and what's actually happening and to push the envelope and to understand where that point of diminishing return is to the benefit of our patients, as long as we're doing it safely.

I think that's probably covering that well. Age at implantation is not a contraindication. I think that this should be applied to both the bilateral hearing loss and the SSD. We always, in medicine, it's much better to look at a biological age rather than a chronological age, like some 100-year-old people look like they're 60 and some 60-year-olds look like they're 120 years old and they're biologically that aged. So, we generally look at all overall health status and longevity, anticipated longevity. The procedure, it's very, very rare that we have a person in a bigger medical center with experienced anesthesiologists. It's very rare that we have a person that's so sick that we would say we can't do an implant on.

You medically optimize them. In fact, I can't think of somebody where we said, "We can't give you an implant. I'm sorry, you're too sick." I mean, there's people that have a short lifespan for whatever reason, but actually physically saying you couldn't do it for that reason or because you're older is not a viable contraindication, in my opinion. The oldest person we've implanted at the Mayo Clinic is 96. And I'm sure other groups have implanted people older. I had a 100-year-old woman who came in and she wanted a cochlear implant. Her hearing wasn't good enough, so I told her to come back in 10 years. So, age should not be a contraindication to cochlear implantation. And there are some data that says the outcomes are just slightly poor.

And there are some studies that show that the outcomes are the same, but I'm not aware of anything that really shows it's significantly worse, and that's speech

perception. And so, we're not talking about quality of life, which is a whole nother facet that's independent of speech perception outcomes. About tinnitus severity is also really interesting. A lot of people thought, so, it turns out that cochlear implants suppressed tinnitus in most people. Rarely, they can make it worse, or rarely it's a net even. Most of the time it suppresses it. So, a lot of people for single-sided deafness, they'll come in and they'll say, so, I think hearing is important. I think it's very important, but I'm just gonna say, sometimes I hear patients say this.

They'll say, "I don't care about the hearing in my one ear, 'cause I have my other ear," they'll say this. But they'll say, "My tinnitus is so bad, I can't function." And obviously, you wanna work through comorbidities of depression, anxiety, but for a lot of people, they're just completely reasonable people and they have really bad unilateral tinnitus. And a cochlear implant might, they'll say, "If you could just get rid of this tinnitus, I don't even care about the hearing. Can you make it better?" And so, in my practice, one of the main drivers is that population 'cause it does really significantly reduce tinnitus for most people. There's several systematic reviews, meta-analysis that say tinnitus reduction or complete suppression occurs in 90% of people who have incapacitating unilateral tinnitus in the SSD population.

Maybe a 5% risk of making it worse, a 5% risk of making it about the same. What's super fascinating to me is it's not purely masking. So, there's this idea that people who are deaf in one ear and you tell them, "You can't use masking. You can't mask a deaf ear." So, what do you do? Well, cochlear implants might be able to do it. And people thought, "Well, you're giving them sound back in that ear." So, you're suppressing their tinnitus through masking. What's interesting is a lot of the benefit people get from cochlear implantation is actually after the device is off even. It's this idea of residual inhibition. So, even when they're not hearing sound and getting stimulus into their implant, or even after they take it off, they'll have hours or days or months sometimes.

The longer they use the device in time, so, for longer months or years of experience, the more residual inhibition they get. There's all also not a huge level of habituation or tolerance development, meaning you might say, well, maybe it helped them at beginning, but after they use it for a while, it's not gonna help them as much, and the tinnitus, it actually doesn't seem to happen. They actually don't build up a tolerance or an issue with it. So, the benefits are probably, you know, tinnitus is probably more like a neural damage issue, and giving stimulation is kind of more like a neuro-stimulator, a neuromodulation rather than masking. I think we should fundamentally think differently about how cochlear implant actually mask or changes tinnitus.

Candidacy considerations. I gotta stop talking 'cause most of this stuff is Dr. Dillon's. So, I'll get through this quick. Before you undergo a cochlear implant consideration, you should at least talk to them about the different options. But broadly, as Dr. Dillon said, you can do nothing. You can give a technology, surgical, or nonsurgical technology that routes to the other side, or you can try to restore binaural hearing to some level with a cochlear implant. CROS systems, BiCROS systems, bone conduction device for routing to the other side, Do not give the binaural benefit back. You do not get sound localization back to any degree, I believe, despite limited data says you get some sound localization back. And then it helps speech understanding in background noise in some configurations, but actually hurts in other configurations.

You think about a person driving in a car and whose right ear is good, and they're talking to the passenger on the side, and they have a CROS system, and the window's down. So, their window's down and they're driving, and the person's talking to their good ear, but all the wind noise is getting shuffled back their other ear. And so, they can't, they would hear better if they didn't even use the device. So, it helps in some situations, and it makes it worse in some situations. And so, at least in our experience, we actually do more CIs by a decent margin for SSD than we use CROS systems, implantable or not implantable. You can go to next.

Candidacy considerations. It's nice to document benefit pre and post. And if you're able to do that, it helps understand how much you've helped the patient. Also helps you learn from your experience. It also helps the world literature to be able to do this. So, these are different measures that help you document benefit. And the idea is that you're giving them better speech understanding and noise. You're doing tinnitus reduction, you're giving better spatial resolution or ability to localize sound. And then what's really kind of grown more and more recently and particularly over the last 10 years is the benefits to quality of life. So, there are some people that actually don't have magnificent outcomes for speech perception, but they'll report a substantial improvement in quality of life.

And there's great work by several people, including Dr. McCracken, that basically says that your speech perception performance and your reported subjective quality of life are not, they're somewhat independent from another. So, you can get significant benefits to your quality of life beyond what you're actually seeing in your speech perception scores. And so, it's just another completely different way to think about the benefits of cochlear implantation beyond just the conventional ones of speech perception. All right, maybe next slide. So, I think we talked about most of this. Cochlear implantation for SSD is the same if they have unilateral or bilateral hearing loss. As far as the actual procedure, it doesn't change at all. I don't think we have time to get into osseo Bonebridge, BAHA tract, all these sorts of things. So, we'll skip a bit. If there's questions, we can talk about it later.

- Okay, so, now that the patient has a cochlear implant, or qualifies for a cochlear implant, we also have some different things that we need to think about as far as the management of these patients. And so, in this section, I'll talk about audiologic assessment, mapping, which is my favorite, and then aural rehabilitation. So, with audiologic assessment, the test battery is going to include unaided hearing detection

thresholds, as well as this need with this particular patient population to isolate the input to the affected ear so that we can assess performance with that ear alone, independent of what's happening with the normal hearing ear. And then look at their spatial hearing abilities. So, for the audiologic assessment, one aspect of this as we do with all of our patients is the unaided hearing detection thresholds.

And so, preoperatively, this is to diagnose the hearing loss, and then postoperatively, this is to look at the residual hearing in the implanted ear. Earlier, I said that we think about unilateral hearing loss as moderate to profound hearing in one ear, and then normal to near normal hearing in the contralateral ear. And we know that with these new surgical techniques and new designs for electrode arrays, the patients can have hearing preservation. And so, if you have patients with particularly good low frequency hearing preoperatively, you're gonna wanna monitor that long term to see if they could potentially be fit with EAS. And then as well as monitoring the hearing in the contralateral ear. And so, as Dr.

Carlson was mentioning earlier, that is something that we need to think about with particular etiologies is the status of that contralateral ear, and is it going to change? When we're looking at performance with the affected ear, again, we're considering the need to isolate the input from that normal hearing ear, and preoperatively, at least at our clinic, we are assessing performance in that affected ear with a conventional hearing aid, much like what we do with our traditional cochlear implant assessment test battery. And then post-activation, we're looking at the performance with the cochlear implant alone. So, we're wanting to see that the effectiveness of the treatment of a cochlear implant for that affected ear. And so, when we're thinking about that, we're thinking about the need to isolate the cochlear implant from the normal hearing ear.

Probably, many of you when testing in the sound field with your traditional patients, you're taking off the contralateral technology, or masking that contralateral ears so that you can assess performance with a cochlear implant alone. Now that we have normal hearing thresholds, that becomes a little bit more complicated. And so, test methods that have been used in various studies, and that are also being implemented clinically include the use of direct audio input technology so that you know that you're only providing that stimulation to the cochlear implant. The plug and muff method. So, we're sitting in the sound field, we put an insert into the contralateral, the normal hearing ear, and then put either the TDH phone or a headphone overtop.

And then to make sure that we're really isolating the input if you're testing in the sound field. Applying masking. So, in our case, we'll put an insert phone in to the ear, overlay the TDH phone, balance the contralateral part on top so that we're not interfering with the cochlear implant, and then present masking to the normal hearing ear. And of course, you have to make sure that the tube isn't crimped so that the masking is actually being delivered and that you are applying effective masking in that case. Something that Doctors Park and Young will likely talk about next week is for children, that that method is not preferred because of central masking that can occur. And the distraction that can occur when you're presenting masking to one ear and you're wanting them to focus on the cochlear implant.

And so, there is this push to use these direct audio input techniques with adults and children. For test materials, what has been used in the literature traditionally are more syllabic words, so CNC words, like what we use for the minimum speech test battery, as well as AzBio sentences in quiet. There are some groups that also assess sentence recognition in noise. I do not see the utility for that. I think the point of the cochlear implant in these cases is to provide this improvement in spatial hearing, at least in many cases. Not so much to improve the speech recognition in the implanted ear. At

least listening alone, many times these patients their goal is not, "I wanna listen to my cochlear implant alone."

The point of doing these tests of testing and isolating just of what's happening with the cochlear implant is to make sure that the cochlear implant is working and that our mapping procedures are effective. So, in that case, I think what's needed is your unaided, or your aided sound filled thresholds that we're looking to see, are we providing an equal balance of the speech information across the frequency range? And then CNC word recognition so that you can track performance growth over time and identify potential red flags later that would indicate either a change for mapping techniques, or a need to investigate the integrity of the internal device. But as far as sentence recognition in quiet and noise, I think efforts could be spent more towards looking at their spatial hearing performance now that you've ruled out that performance is growing with a cochlear implant.

So, again, for spatial hearing, this is often the reason in addition to tinnitus suppression for many patients, that they are pursuing cochlear implantation over alternative treatment technologies. And an important thing to think about is their performance with the cochlear implant alone is not reflective in majority of cases of how they do in the combined condition. So, there have been a number of studies that have looked at CNC word recognition with the cochlear implant alone, as compared to the benefits that happen on spatial hearing task. And we're not seeing significant correlations with that. So, you can't just say, "Well, I've measured performance with the CI alone. So, I know they're doing better in this binaural condition." So, there's a real need for our field now to think about assessing binaural hearing and not just speech front, noise front all the time.

Now, that becomes a little bit more complicated with current clinical setups. So, not many current setups can allow for a multi-speaker array in order to assess sound

source localization, but we can use our current clinical setups to test spatial hearings. So, for a speech recognition and spatially separated noise task. And we have a depiction of that here. This is a traditional clinical setup where we have the listener facing one speaker, and then we have another speaker offset 90 degrees. And in this case, we're presenting the target sentence and the background noise from the same speaker. And so, if we've tested with the cochlear implant on versus off, this would tell us about the improvement because of binaural summation of having two ears, as opposed to listening in a monaural condition.

And this is our traditional setup, right? We put them in the sound booth, we present CNC words in quiet, AzBio sentences in quiet, and then start to introduce noise. And you will see growth on these conditions. However, when we're thinking about patients that have normal hearing, they may already be showing pretty good performance in this condition. And so, you're needing to think about your signal to noise ratio that you might be using with these patients. With our single-side deafness patients, we are presenting it at 0 dB SNR so that we get them around 50% in this speech front noise, front condition. And then we test them at that same signal to noise ratio for the other conditions, so that we have it consistent and can observe a benefit across the conditions.

So, next we have the target from the front, we've moved the masker over towards the normal hearing ear. So, this will show us if the cochlear implant's on versus off, or preoperative versus postoperative. The effect of the head shadow benefit that you would get. And then you simply move the patient's chair and now they're facing that other speaker, and you present the target from that speaker and deliver the noise towards the cochlear implant ear. And that could show you the benefit of binaural squelch in those conditions. One thing to point out is that across studies, we see significant improvements on the head shadow effect. And so, that's shown very

reliably. It is variable as to whether patients also show significant improvements on the summation effect and the squelch effect.

What we see is that there's not a decline in performance though, with the introduction of the cochlear implant. So, in this particular condition, now we're delivering noise towards the cochlear implant. If you thought that the cochlear implant is going to be too distracting, you would see that performance would be worse in this condition relative to having the cochlear implant off and them listening with just their normal hearing alone, with the noise being delivered towards the CI. So, the fact that we don't see performance drop significantly in this condition shows that patients are benefiting from having two ears together, and that this is not a detriment in performance that they could experience. So, important to assess the binaural hearing abilities on these different conditions.

So, what we would recommend based off of what we know so far as for a reasonable clinical test battery is to look at word recognition with cochlear implant alone, and then have them listen with both ears on a speech recognition and spatially separated noise task, so that you can look at the benefit of the cochlear implant, listening with two ears in comparison to what they were doing preoperatively, or if you have time to test it on versus off, and then to use some of these questionnaires that are specific to tinnitus perception. At our clinic, we use the tinnitus handicap inventory. It's a 25-item questionnaire. It's very easy for the patients to complete quickly prior to being brought back to see the clinical audiologists, as well as the speech spatial and qualities of hearing questionnaire, which is really tailored towards the patient's perceived abilities of listening with two ears.

And so, it's beneficial to think about the sound source localization in those cases, there is that subset of looking at spatial hearing abilities particular to localization, that's one way to capture that benefit for the patient as well. With mapping, this is one of my

favorite topics. So, English King and I, who was the clinical audiologist at the time, spent a lot of time before we started our clinical trial, talking about how we would map these patients. Because something that you'll find is that when you first activate them, the sound quality is so different from the normal hearing ear that their initial response is to turn it down. They think it's too loud, it's overwhelming their normal hearing ear, and they want you to turn it down.

And so, you'll turn that stimulation level below where we would think to have it to provide better speech recognition. Well, now, are they not improving with the cochlear implant because the cochlear implant is not effective for single-side deafness, or is it because it's not at a loud enough sound quality to provide them with speech information and spatial hearing information? So, the way that we're doing this at our clinic, and the way that we did it as part of the clinical trial was we were pretty aggressive with trying to obtain sufficient simulation levels at activation so that they were leaving the clinic with a sound quality that we thought would be, or a sound level that we thought would be sufficient to improve speech recognition with the cochlear implant.

So, we would plug the normal hearing ear, measure the threshold levels, electric threshold levels, and the reason to plug the normal hearing ear in this condition is to limit the environmental noise that's happening in the clinic, on their ability to do that task, and then have them measure comfort levels. And you can do this either behaviorally, or with Electrical Stapedial Reflex Threshold test. And then we would also balance loudness across channels to make sure that we are providing equal levels of loudness across these different speech frequency ranges. Then we would remove the plug from the contralateral ear and talk with them a little bit, do some environmental sounds to have them loudness balance with the normal hearing ear.

And if they were wanting to turn it down, we would maybe decrease slightly, but would not move it more beyond that. And counsel a lot about the importance of trying to acclimate at an appropriate loudness level and not sort of stair-step them into it because we want them to have improvement early. And hopefully, that would increase the rate of use. One thing that is starting to come out is the rates of non-use in patients. And I do wonder a little bit about if we start them off too low and they're not experiencing those benefits on spatial hearing task within these early months because the level is too low, is that causing this higher rate of non-use.

And non-use rates are varying across different reports, but that's just something to think about is trying to provide them with the best sound quality, the best levels early, so that they are achieving these benefits that they are looking for on these spatial hearing task. Something that is emerging for our area of the field is thinking about the way that we are providing the speech information. So, how important is it to align our frequency filters with the cochlear place frequency? So, we know that frequency to place mismatch can interfere with spatial hearing abilities. And what frequency to place mismatch is is the discrepancy between the frequency information you're providing for a given contact, and the cochlear place frequency that's adjacent to that contact.

And if we have shifts in what we are providing, so, if I'm presenting 500 Hertz to a 1000 Hertz cochlear place frequency, the patient has to acclimate to those shifts in order to show an improvement. However, we know from patients that have bilateral cochlear implants, that these frequency to place mismatches, which create an inner aural mismatch, disrupt their ability to benefit from some of these binaural cues. And so, patients that have mismatched inner aural information, have poor spatial hearing abilities than when we hit the line with this information. And what's really interesting about patients that have bilateral cochlear implants is you can put the same electrode in each ear, but you still could be stimulating different cochlear regions with the same

filters because of differences in our cochlear morphology and differences in surgical approach.

And that that can also disrupt their spatial hearing abilities. So, if we take that into our single-side deafness patients, now we have patients that have normal hearing in the contralateral ear. And so, getting it right with where we're presenting that frequency information could be wildly important. In two ways, you can try to match the frequency information to the cochlear place frequency. One is to implant a long electrode array so that your default filters are closer to that cochlear place frequency. Even then, we know that there can be a lot of variability in frequency to place mismatch across patients. An alternative that we are currently investigating is, or in addition to long arrays, is to map the filter frequencies to match the cochlear place frequency.

So, if you have some information about where that electrode array is, you can individualize the mapping of your frequency information to match that cochlear place frequency, and that could support better spatial hearing ability. So, that's something that is emerging in our area of the field and something to keep an eye on. And then finally, aural rehabilitation. We know that this can be beneficial for all patients. Unfortunately, it's reimbursed more for children than it is with adults. A lot of studies for cochlear implantation in adult patients who are using aural rehabilitation within those initial weeks to months of CI use. Some were recommending 20 to 30 minutes a day. I think doing the direct audio input early on helps the patient recognize the appropriate loudness they need for better speech recognition with the cochlear implant alone.

And that helps them in the combined condition as well. There is also some methods looking at bilateral training. And Dr. Firszt at WashU is doing some work looking at localization training for this patient population. So, right now we're still in a bit of a guessing game of what's the best form of aural rehabilitation for these patients. But we know that if those that have it early and are doing these tasks, they tend to do better,

faster than those that are not doing some sort of formal training, or books on tape at home, or things like that. And that leads us into our guidelines that have been developed by the ACIA task force.

- So, just going through these. And I think the big thing to remember is that there's not a ton of data, and this is based on our understanding of the current data. And we don't wanna be overly prescriptive, meaning really you need to highly individualize your recommendations for the patient based on your understanding and what their priorities are. So, having said those things, we'll go through the guidelines. It's recommended that individuals with sudden and rapid, progressive single-side deafness undergo appropriate medical workup and have a period of at least three to six months to see if they spontaneously improve and whether or not they adapt to a point where they don't need to, where they don't want to pursue something.

There are some time sensitive conditions, such particularly ones that involve progressive ossification where you might intervene earlier. Labyrinthitis ossificans from meningitis is one of those. Another one is if you're, particularly, if you're doing a concurrent tumor resection with consideration of cochlear implant. If you do it through a trans-lab approach, so, going through the labyrinth, most people believe you should place an electrode, or at least the dummy electrode space holder at the same time, because ossification happens pretty quick than otic capsule fractures, but otherwise, it's better to give the patient some time, except maybe in these situations, do it, intervene earlier. Consider the probability of them developing bilateral hearing loss over time. So, if the underlying etiology would predict that they have a high risk of acquiring hearing loss on the other side, or if they even have started to demonstrate some level of what we call threatened hearing in the other ear, that might be a driver to consider earlier intervention.

Number three, preoperative imaging, by definition, people with asymmetrical or SSD warrant an MRI. I would consider it standard of care in the United States. Some people would argue that a little bit, but basically, you need an MRI. If you wanna get a CT scan, you can. Most people don't necessarily need it, but if it's helpful for surgical planning, it's totally reasonable to do an MRI, will basically take care of 99.9% of the things you need to understand how to manage the patient. Cases of advanced cochlear ossification, severe labyrinthine dysplasia, and cochlear nerve aplasia are potential contraindications to cochlear implantation, particularly in a setting of SSD where patients already have one good ear and they might be at an elevated risk of not using the device again.

This is nuanced, but those are pretty, if you really believe they don't have a nerve based on all the information, or if it's a horrible malformation, there's barely little of a cochlear left, or they are probably better not, at least at this point, not to do it. Yeah, one idea or one thing to think about is the duration of deafness. You should not take the data from the bilateral hearing loss group and extrapolate and apply to this. The groups are fundamentally different, and we might find out that there is overlap, but I think that we will probably find there's just as many differences as there are similarities. And even in the bilateral severe to profound hearing loss group, single, or sorry, duration of deafness only accounts for a very small amount to the variance seen between people.

So, even in that population, I think we wait that way too much in general. And certainly don't wait it too much in the SSD population until we have better data. Advanced age for both groups shouldn't be a contraindication to implantation. And think about the overall patient's biological health rather than their age. Tinnitus reduction is a major driver, and particularly if the person's bothered by it, they're not rehabilitated through other means such as cognitive behavioral therapy masking, or whatever else you're gonna try. And it's still bothering them a lot. You can consider implantation 'cause it

does have a good chance of improving tinnitus perception. You should at very least discuss nonsurgical options or even surgical routing options like bone conduction devices.

But again, broadly, you either do nothing, you route on the other side through surgical nonsurgical means, or you try to restore some binaural cue through implantation on that side. And they all have their individual pros and cons. Preoperative counseling is important to going through the device, giving them appropriate counseling and expectations. The biggest thing, I think, is the patient has to be very motivated. I always tell patients, if you only use your device, for SSD patients, if you only use your device when you need it, it's never gonna be good enough for you, because you haven't practiced when you didn't need it. So, they have to wear it most of the time. Dr. Dillon has great data that basically says, the longer you use it, the better your performance is.

And so, probably over six or eight hours a day, if you can do that, you're gonna have better performance with your device. And that's just you're growing with your device use. You're getting better performance from it, things like that, but they can't only use it when there's a lot of background noise and say it didn't work for 'em. And they shouldn't even undergo implantation if they don't, you know, or if they have not been tested.

- For the post-activation test battery, we should include some subjective questionnaires, as well as looking at that include quality of life, but also tinnitus severity since a lot of these patients are reporting different levels of tinnitus severity and their reasons for pursuing cochlear implantation have included tinnitus suppression. For CI recipients that have preoperative hearing, which we put here as moderate or better acoustic low-frequency thresholds, we are wanting to monitor that hearing postoperatively. So, we're assessing unaided thresholds in both ears. One

consideration when assessing the impaired ears that need to isolate the contralateral normal hearing ear, and that can be done either with direct audio input technology, or plugging the contralateral ear and putting a circumaural phone over the top, or presenting masking and then having the circumaural phone over top as well.

But remembering to make sure that insert foam is not crimped. 13, we're looking at the inclusion of spatial hearing into the post-activation and preoperative test battery. So, something that can be implemented clinically is a task of speech recognition in spatially separated noise, which we have those images to show how that can be used in most clinical setups. And then the behavioral measurement of electric thresholds, it's recommended to plug the contralateral ear so that you're limiting environmental noise. And then, for the behavioral measurement of MCLs, first, doing that with the ear plugged, and then removing the plug and comparing it with the contralateral ear. And as was just mentioned, if you're not going to wear it, then you're not going to show a benefit.

And so, we would recommend a minimum of eight hours of device use per day. The patients that think that, "Oh, I'm just going to go to the grocery store today. And so, now I'll put on my cochlear implant and I'll show a benefit." That's not what's going to happen. You have to wear it every day for at least eight hours to have that benefit. And one of the patients that we had early on was not improving the way that she thought she was. And it came out that she was only wearing it about three hours a day. And she got a little competitive about that. And after she started wearing it eight to 10 hours a day, she had these significant improvements on these spatial hearing tasks.

So, you need to wear it to benefit from it. And then finally, auditory training is recommended particularly within these initial months of device use to help facilitate the benefits of the cochlear implant. And with that, thank you so much for attending today. Thank you, Dr. Carlson, for joining me on this. And thank you, Donna. And we

appreciate AudiologyOnline for giving us this opportunity to share these data with you today.

- Thank you both so much for such an informative presentation. I do encourage, I saw a couple questions come in about looking for citations, and you do have a separate reference list. So, please, download that. It's one of your handouts. Thank you so much to both of our presenters today and to Donna, and we hope everyone will tune in to continue this topic next week with children. So, thank you so much. With that, we'll wrap our class and wish everyone a great afternoon.