Dr. Jason Barnes:

Hey there. Welcome to another episode of ENT in a Nutshell. My name's Jason Barnes, and today we're joined by Dr. Matt Carlson and we will be discussing adult cochlear implantation. Dr. Carlson, thanks again for being here.

Dr. Matthew Carlson:

Thanks so much for having me.

Dr. Jason Barnes:

Now again we usually go through a given pathology when we have an episode, but for this one we wanted to focus specifically on cochlear implantation, and for this episode more specifically on adult cochlear implantation, understanding that there will be some overlap on some of the pathologies that we discuss in later episodes. But Dr. Carlson, to start when someone presents to your clinic who is likely a candidate for cochlear implantation, what do they usually present with?

Dr. Matthew Carlson:

Yeah so when we're specifically talking about adult cochlear implantation, most patients are presenting with gradual progressive hearing loss over time. Often times there's one ear that is better than the other ear. Most of the patients have been using hearing aids for a longer period of time, and they're starting to say their hearing aids just aren't cutting it. They've had their hearing aids reprogrammed, or they've had several different pairs over the years and even decades, and they're getting to the point that particularly when there's background noise, they're having more difficulty. It's often a very gradual or indolent thing. It's frequently that they'll say things like "I just don't go out as much as I used to. I don't like going out. I avoid social situations or social gatherings because when there's any background noise or there's multiple speakers, forget it. I just can't participate." Often there's this element of social withdrawal.

So there's different ways people can present, but that's in the adult population probably the most common presentation.

Dr. Jason Barnes:

When you see these folks, what are some specific questions you should ask, understanding that hearing loss is a large part of this, but what else would you ask them?

Dr. Matthew Carlson:

I think one thing that's really critical to think about when you're looking at this topic is the understanding that cochlear implantation in adults are highly underutilized. If you look at the number of people who are potential candidates for cochlear implantation compared to the number of people who are treated, the current statistic is that less than 10% of people that might qualify for an implant in adults don't undergo implementation. There's a lot of reason of that including limited awareness, poor understanding of the current guidelines, lack of best referral patterns between audiology and ENT surgery. Even within audiology training and ENT training, a lot of people just don't get a lot of exposure to cochlear implantation. So in my opinion, if you're starting to think that they might benefit from a cochlear implant and you're on the fence, in those situations for sure you should consider referring somebody.

I think we should err towards early referral rather than waiting and wondering if the person might benefit. There are some good screening questions, but before I go into a couple good screening questions, I want to emphasize the point that when you encounter a patient in the clinical setting for any sort of visit whether it's hearing loss or something else, I think we as a whole consistently underestimate the degree of hearing loss. Patients who can look directly at your face in a quiet setting and understand you does not mean that they have very good hearing. It's remarkable that always kind of astounds a lot of the residents when they're in the room and I'm talking face to face with a cochlear implant patient just speaking and they're understanding everything I'm saying, and I'm telling I'm going to implant them, and that the issue is the clinic setting does not replicate the real world situation where there's multiple talkers, a lot of background noise in social gatherings where most of these patients are most of their day.

So having said all of that, I think there are some generally good screening questions that you can help to get at whether or not a patient's a good candidate. So if you one question is can you carry out a conversation with another person if you can't directly see their face? Related to that, can you carry out a conversation on a telephone as long as the volume's turned up enough? That's kind of the same thing. You don't have visual cues to help guide you. Do you struggle to communicate with others in large gatherings such as church or social gatherings? Then related to that, do you no longer really participate in those sorts of events, because you have such a difficult time going to them that you feel frustrated or you get to the point where you think it's not worth it for you?

Those are very common good screening questions. Related to that, is the patient saying something like "I've tried multiple hearing aids, and they're just not really doing it for me" despite spending a lot of money on it. To me that's another red flag that they might be considered a very reasonable candidate for a cochlear implant.

Dr. Jason Barnes:

Can you speak a little bit to the epidemiology of these patients? Who's the typical patient who walks in ready for a cochlear implant?

Dr. Matthew Carlson:

So in this population, so we're specifically talking about adults, currently, it's uncommon at least in my practice that I'll see a prelingual adult coming in for an assessment, so somebody that was born with congenital hearing loss, or even perilingual, meaning they lost their hearing during the time of speech and language development. Most of the people I'm seeing are post-lingual, means they have adult onset or later in life onset hearing loss. Those situations it's the age range varies significantly. It can be anywhere from the 20s to 90s for patients. The factors that lead to sensorial hearing loss for these patients are variable. The most common reasons a person has hearing loss in adulthood include presbycusis, noise exposure throughout their lifetime, repetitive loud noise exposure either occupational or recreational, exposure to otoxicities, and then a lot of people have a hereditary component where they have a family history of hearing loss before the age of 50 or 60, and that's a major contributing factor.

We always think about whenever we say "What's the cause of hearing loss" we always want to put it on one thing and say "Oh that person has a genetic condition that's leading to hearing loss" but in actuality, like most things in medicine, the condition is multifactorial. Most people have more than one hit that's leading them to have the hearing loss. So for example, somebody over the age of 70 who has hearing loss has probably presbycusis, but a lot of times, they've had a history of loud noise exposure. In my area, a lot of people have noise exposure to farm work or occupational noise exposure, recreational noise exposure from hunting, things like that. So we'd like to make it nice and clean, but most of the time, we're losing hearing gradually from many different causes. We all start out with when we're born, we start out with about 15,000 inner hair cells, and every day we live, every time we're exposed to louder noise exposure or other insults, we lose those hair cells, and that's what contributes for most people contributes to sensorineural hearing loss.

Dr. Jason Barnes:

Is there a gender predilection?

Dr. Matthew Carlson:

Most of the epidemiological data particularly for presbycusis and occupational noise exposure would suggest that males are at higher risk for acquiring hearing loss than women.

Dr. Jason Barnes:

In the clinical setting, is there anything you're looking for on physical exam when you evaluate these patients?

Dr. Matthew Carlson:

So again in the adult population in contrast to the pediatric population, most of the adult patients at least in my practice don't have concombinant inner ear malformations, or at least certainly not severe ones. I will say occasionally maybe once a year twice a year I'll see an adult with bilateral EVA that's finally entering the age where they can consider cochlear implantation or enlarged vestibular aqueduct or Mondini malformation IP2. But most of the time they don't have malformations. So on examination findings, of course you'd look for craniofacial abnormalities, multiple pits in front of their ear, accessory tragus, you look for other conditions that might help you determine if they have underlying condition, but again, adult population that's not very common.

I do particularly for the elderly patients, I do really try to hone in on what their overall cognitive status is when I'm speaking with them. A lot of times and that's easy to disguise, a lot of times people with some cognitive impairment, their family will come with them, and they'll kind of rely on their family and the family members will answer the questions. Sometimes it goes unnoticed. So I try to directly ask the patient a lot of these questions. If I'm starting to get the feeling that they're not remembering things that they should remember closely, I'll be very frank and ask the family and the patient "Are you having problems with memory impairment?" I'll try to listen to some of that information. It doesn't mean they're not going to be a candidate for a cochlear implant, but [inaudible 00:08:41] can potentially because they may have an issue with central processing despite giving them good access to sound through the cochlear implant.

Dr. Jason Barnes:

Moving on briefly to pathophysiology you already discussed the main causes of hearing loss. Can you dive a little bit deeper into the pathology around hearing loss?

Dr. Matthew Carlson:

Yeah I think if you understand the pathophysiology of sensorineural hearing loss, you'll understand why cochlear implants work in most people. So as I alluded to earlier, we're all born with a certain amount of inner ear hair cells in spiral ganglion cells and a cochlear nerve. So sound, the way we perceive sound is that sound travels through airwaves that hit our tympanic membrane and they make our tympanic membrane malleus incus stapes vibrate, and that moves fluid within the inner ear. That deflects the hair cells, and that transduces mechanical into electrical potentials, neural action potentials that go along the so goes to the hair cells, to the spiral ganglion cells, to the cochlear nerve and then to the brain for processing where it's understood as sound. So anything at the cochlea or more distal or towards the brain you can cause some sort of sensorineural hearing loss.

As it is, most types of sensorineural hearing loss are caused by deficiency an end process and a deficiency of hair cells. So those are the cells that transduce mechanical into electrical signal, so noise induced hearing loss, otoxicities et cetera, the hair cell loss might be secondary, it might be stria vascularis injury for example, but ultimately, most of these have an end pathway where they lead, they have a hair cell insult. The reason a cochlear implant specifically works for most people is a cochlear implant bypasses absent or missing damaged hair cells. They stimulate the spiral ganglion cells, cochlear nerve directly. So that's why it works with most people. There are some other conditions that can cause sensorineural hearing loss that aren't necessarily related to hair cells or the inner ear. You can have auditory neuropathy where you have dyssyncronous signal sent from the cochlea and along the cochlear nerve to the brain.

You can have retrocochlear pathology such as an acoustic neuroma, a vestibular schwannoma for example that's pushing on the vestibular nerve, or you can have a central processing disorder. All those things can contribute to it. Interestingly, many times and contrary to what you'd intuitively think, many of these conditions are still rehabilitated with a cochlear implant. We found more and more that patients with vestibular schwannomas particularly if they've only had radiation or observation respond pretty well to a cochlear implant. Even sometimes after surgery, patients with auditory neuropathy can often have because the cochlear implant provides a super physiological electrical stimulation, it often resynchronizes or resets that asynchronous nerve [inaudible 00:11:25] signal along the cochlear nerve.

For some situations with central processing, a patient can still benefit significantly from a cochlear implant.

Dr. Jason Barnes:

You mentioned spiral ganglion cells. Where are those located?

Dr. Matthew Carlson:

So you have the vestibulocochlear nerve leaving the pons going into the IC. When it gets to the cochlea, it will kind of fan out along the medialis. That's where that one single cable of the cochlear nerve starts to fan out into these small appendages or ends of the cochlear nerve. Spiral ganglion cells are located distally within the medialis. There's something called the Canal of Rosenthal where a lot of these terminal appendages of the cochlear nerve fan out before they go and innervate along their dendritic appendages, and to the organ of corti. So that's what you're probably stimulating directly as your spiral ganglion cells. Different disease processes can affect hair cells and primarily or secondarily affect your total spiral ganglion cell counts.

Dr. Jason Barnes:

Moving on to work up, when you see a patient in clinic who seems like they might be a good candidate for cochlear implantation, what's the first step at work up?

Dr. Matthew Carlson:

So sometimes a patient's referred directly in for a cochlear implant assessment. The outside provider thought that they would be a good candidate, and they go straight to cochlear implant testing, but also pretty commonly, a patient will come in just with an audiogram with a chief complaint of sensorineural hearing loss. You have to make that first step yourself. Are they a potential candidate or not? As a general rule of thumb, so just to back up a little bit, the first assessment for somebody with hearing loss in the ENT clinic, in the audiology clinic is an audiogram. An audiogram includes behavioral tone testing, or pure tone audiometry where the patient is presented with different tones at different frequencies at different levels and they point out which frequencies at what level they can hear. That's graphically presented in an audiogram. The second is speech audiometry.

One of the main components of speech audiometry is word recognition scores. So monosyllabic words are presented to the patient, and the patient should respond to those. The number they have correct is your word recognition score. As a general rule of thumb, people who scored less than 50% on word recognition score testing on routine speech audiometry are in the ball park of where you should consider the possibility of a benefit of a cochlear implant. It's a good general rule of thumb. You have to remember that the audiogram is separate from more formal cochlear implant candidacy testing. More formal cochlear implant candidacy testing is much more involved, it's a much longer visit, but the audiogram at the beginning is the first step to kind of think "Is this person a candidate or not?"

Dr. Jason Barnes:

What's involved with the actual CI evaluation, their specific tests, sentence scores, that kind of thing? So what's involved in that?

Dr. Matthew Carlson:

So when a person goes into audiological testing to determine if they're a candidate for cochlear implantation, the first thing they'll do is they'll either have their own hearing aids tested, or they'll get a pair of loner hearing aids to make sure they have the best hearing aids or the best hearing that they could obtain with conventional hearing aids. It's in that setting they'll go into this testing. All candidacy criteria require that the patient be tested in the "best-aided condition." After you've made sure their hearing aids are working well, you'll get a series of different tests. The tests usually encompass word testing and sentence testing. All of this testing, basically all of this testing is performed using open set speech recognition testing. That means that the patient doesn't get visual cues, and they also don't have a list or something else in front of them to pick from.

So if you say car, they don't have a list of 10 words that include ball, boat, house, car that they can choose from, because of course your score would be higher. It's open set meaning they have to come up with the word without any type of cue. So historically, we would use testing such as HINT or what's called the hearing in noise test, or the CUNY testing. Right now, the most common tests that are used, at least within the United States are CNC, which stand for consonant-nucleus-consonant. They're monosyllabic words, and sentence testing. The most common sentence testing used in the United States are AzBio tests. AzBio tests are a little bit more difficult that HINT and CUNY testing for example, and the reason is they're presented at a more typical speech rate. They'll have different speakers at different times, which make it a little bit more difficult for the average person to hear, but it also is more fair because it's replicating what a person might actually encounter in the real world.

So both word and sentence score testing are presented as a percent correct, and what the person can present back without any visual cues, and just hearing the words alone. You usually present it in ear specific and also best-aided condition. So we'll talk about the candidacy criteria a little bit, but they're usually said in the phrase of in the ear to be implanted, or the best-aided condition. So you'll say left best-aided condition, right best-aided condition, and together what's the best hearing the person has. The last thing I want to point out is increasingly we're testing with background noise. So earlier in this podcast, I alluded to the fact that hearing, an assessment or estimate of someone's degree of hearing loss is not really that accurate if you're just seeing them in a quiet clinical consultation, you're looking directly at them, and they're getting visual cues and they can watch your lips moving, et cetera.

Real world listening more typically involves multiple talkers without being able to see the one talker's face very well, with competing background noise, which can really be difficult for most of these patients. So more and more we're also testing in background noise to get a better assessment of how they're actually functioning. When we talk about testing in background noise, you'll it commonly presented as +10 or +5 dB SNR. That's the signal to noise ratio. The lower the number, the more difficult. So +5 dB SNR is more difficult than +10 dB SNR. This allows us to again as I said test them in a more accurate way to determine if they're really going to benefit from a cochlear implant and to understand how they're functioning in their daily life.

Dr. Jason Barnes:

Is there a role for imaging in cochlear implant candidacy?

Dr. Matthew Carlson:

Absolutely. I'll say that this is a controversial point, and there's no standard guideline, and there's no standard recommendation across all insurance carriers, or Medicare. It's more surgeon preference. I would say that that's kind of broadly under the umbrella of a medical surgery candidacy assessment. So we had just talked about audiological work up. The medical surgical work up includes imaging of some sort. For adults, I almost always obtain just an MRI and not a CT scan, but I know other providers who will just get a CT scan and not get an MRI scan. Most I guess I don't know for sure, but I would say that most providers don't usually get both for adults in contrast to kids. The reasoning for me just getting an MRI and not getting a CT scan is most of these adults have asymmetrical hearing loss to some level, and there's already an established guideline that if they have asymmetrical hearing loss, they should be getting an MRI anyway to look for retrocochlear pathology.

So I think that most of them weren't getting one, but then on the MRI, I can look for inner ear malformations which are pretty rare for adult onset hearing loss can occur but pretty rare. I just don't see the need for getting a CT scan. Other people like I said will just get a CT scan alone. A CT scan might be particularly beneficial if you are suspecting a temporal bone inner ear malformation, or if they've had multiple surgeries before. The other aspect that's really important to think about MRI and or CT scan is the conditions where you're losing, where you're having ossification within the inner ear. There's a couple conditions that can lead to that. The three most common ones we talk about are labyrinthitis ossificans post meningitic. You can also get labyrinthitis ossificans after acute otitis media where you have bad labyrinthitis.

There's different phases to labyrinthitis ossificans. The first after a person develops some hearing loss, they'll have early fibrosis, granulation tissue formation, and then over time they'll have bony reaction and bony changes. A CT scan can underestimate cochlear lumen patency because it's only really seeing bone formation well, whereas an MRI and particularly on heavily T2 weighted MRI with sub-millimeter slice acquisitions, you can see the patency of the cochlea perilymph just like the CSF on heavily T2 weighted imaging should be bright white, and when you start to lose that signal if it becomes muddy or dirty, we call it, or if you lose it all together, you're worrying about your cochlear patency.

So again, most commonly, that's in the setting of labyrinthitis ossificans, but you can also get it with otosclerosis with far advanced otosclerosis or cochlear otosclerosis. You can also get it after trauma, so otic capsule fracture would be another situation where you might worry about cochlear patency. The whole issue of cochlear patency is if the cochlea scar shut, it's hard to get a electrode fully in there. It also just the ossification itself produces results in loss of spiral ganglion cells, which may make your cochlear implant outcome suboptimal from that standpoint also, so a long answer to your question.

Dr. Jason Barnes:

We've talked about work up, we've talked about pathophysiology. So what are the official criteria for someone to be an implant candidate?

Dr. Matthew Carlson:

So that's evolving. It's a moving target. I think the best way to answer that is briefly talk about the evolution of candidacy criteria. So single channel cochlear implants were first approved by the FDA in 1984. The multichannel cochlear implant was first approved by the FDA in 1985. That was approved for adults with bilateral profound sensorineural hearing loss. In the 1990s, children were approved for cochlear implantation. Then the candidacy have continued to become more and more encompassing or inclusive. As cochlear implant outcomes are improving, as the devices are better, as their selection is better, we're seeing better outcomes. Whenever you're thinking about implanting somebody, really the bottom line is is a cochlear implant going to reliably provide them a better outcome than a hearing aid is? As their outcomes are getting better or more and more likely to implant patients more and more degrees of residual hearing.

I think it's worth pointing out several recent expansions in cochlear implant FDA labeling criteria. In 2014, the hybrid cochlear implant the hybrid med el cochlear implant was first approved by the FDA for implantation. We'll talk about hybrid implantation a little bit more in a second. More recently, just this past year, cochlear implantation for asymmetrical hearing loss in single sided deafness was approved, which is my opinion is wonderful. It'll take a little bit of time before insurance carriers are covering it, but this is the first big step towards allowing us to implant these patients. In that study, single sided deafness was described as normal hearing in one ear, and contralateral ear having essentially profound hearing loss, with less than five percent word recognition or sentence recognition.

An asymmetrical hearing loss was anything from mild to moderate sensorineural hearing loss in the affected ear, and profound hearing loss in the other ear. Again, that was just approved this past year, but it will take a little bit of time before insurance approval allows it. But the most common criteria that insurance carriers are using right now for adults is no better than 50% on sentence scores in the ear to be implanted, and no better than 60% in the best-aided condition, and no better than moderate to profound sensorineural hearing loss. Those are the criteria for adults for what we call a conventional length cochlear implant electrode. Medicare is more restrictive. The current criteria for Medicare cochlear implantation is no better than 40% in the best-aided condition on sentence scores, and the person must have worse than moderate to profound sensorineural hearing loss.

Medicare criteria are somewhat vague in what they describe. The two areas that they're particularly vague on are they don't give ear specific information, and they don't give anything about what type of sentences are being used, and if there's background noise. So that allows for some latitude on the provider's part to ensure that you're serving the needs of the patient the best way.

Dr. Jason Barnes:

So we spent the first part of this episode talking about the candidate, the patient, the pathophysiology work up, and for the second part, I wanted to talk more about the technology and what a cochlear implant is, the procedure and that kind of thing. Can we first start with you explaining what is a cochlear implant?

Dr. Matthew Carlson:

Absolutely. So there's currently three FDA approved cochlear implant manufacturers that provide devices within the United States, and there are some small variations between them, but they all really share common features that drive the functionality of a cochlear implant. The external components include a microphone, a sound processor, a transmitting coil, and the internal components include a radiofrequency receiver, a micro processor based stimulator, and a multi channel cochlear implant electrode. So just real briefly, the external sounds in the environment will come to the microphone, that'll get picked up and processed by the sound processor, and that'll go to the transmitting coil. The transmitting coil is that little circle that you'll see on everyone's temporal parietal scalp that is connected to the ear level processor.

That gets transferred through a transcutaneous radio frequency signal. So there's no wire between them or anything. It's through the RF signal that it goes to the internal device. When you think about the internal device, that little microcomputer, we usually refer to that as a receiver stimulator, receiver in that it's receiving that input from the external coil, and then stimulator in that that little computer is taking that sound and sending them down preferentially down different channels. So if you think of what a cochlear implant electrode looks like, I think it'll help you understand how it works. So all the intracochlear portion of a cochlear implant electrode contains a single cable or single electrode. Within that are multiple contacts. Depending on the manufacturer, there's anywhere from 12 to 22 different contacts. Each one of those individual contacts is spaced out relatively evenly along the electrode, and each one of those contacts are independent of the one right next to them.

So you can send electrical current down just one of those and not another one. That allows you to preferentially stimulate different parts of the cochlea at different times, which is primarily responsible to your frequency that you're giving to the cochlea when you stimulate. So each one of those contacts are individually hermetically sealed from one another, or isolated from one another. When you think about how a cochlear implant works, you have to once again go back to the tonotopic distribution of the cochlea, that anatomy. If you can recall, the basal part of the cochlea is responsible for high frequency hearing loss, and the apical regions are responsible for low frequency. So just in the same way as the organ of corti is stimulated by transducing mechanical and electrical signal through hair cells, when you stimulate the spiral ganglion cells, the match up isn't exactly the same as the tonotopic map for acoustic stimulation, but it's relatively close for spiral ganglion cell stimulation.

So that's how I'd say in a nutshell how a cochlear implant works, and the different components of it.

Dr. Jason Barnes:

Can you describe the different types of cochlear implants?

Dr. Matthew Carlson:

Yeah so there's a lot of different ways you can break up or divide or talk about cochlear implants. Probably broadly the best way to divide them up are conventional length cochlear implants and hybrid designs. Conventional cochlear implants are just what the name implies. They're the ones that have been developed for a long time. They're usually for patients with more degrees of hearing loss, although the line is blurring between it, between the two. Generally the electrode goes in the cochlea at least 360 degrees, but more commonly 400 degrees or 500 degrees. When I say degrees, that's number of times it goes around the cochlea itself. In contrast, hybrid cochlear implants use shorter often thinner electrodes. The idea behind it all is that many patients with sensorineural hearing loss have preserved low frequency hearing. If you have really good low frequency hearing, or even what we would call functional low frequency hearing, but you have really bad high frequency hearing, it'd be great if you could just stimulate the areas that were bad, and save the parts that were good.

That's what the hybrid idea is. So the idea behind hybrid cochlear implantation is that by putting in a shorter electrode that only goes around say 270 degrees or less than 360 degrees is that you can preferentially rehabilitate the high frequency hearing loss that is caused by the basal regions of the cochlea needing stimulation, but protect the apical regions that are responsible for low frequency hearing. So you give the patient the best of both worlds. Then can use the cochlear implant for the high frequencies, and then they can use a hearing aid for the low frequencies. Hybrid stimulation is the situation where you're using a hearing aid and a cochlear implant in the same ear. That's different than bimodal stimulation. A lot of people confuse that. Bimodal stimulation is where you use a cochlear implant in one ear, and a hearing aid in the contralateral ear which also has benefit.

Now when I presented that, it might make the listener think that anybody with a lot of low frequency hearing would only be implanted with a hybrid device and anybody who had profound hearing loss would be implanted with a conventional hearing device. But I have to say that the line is blurring a lot. A lot of centers are only using conventional length cochlear implants even for patients with significant degrees of residual hearing, because even with an experienced cochlear implant surgeon when performed very carefully, you can still preserve hearing in a lot of patients. So the general rule of thumb is that you can preserve low frequency acoustic hearing at the one year time point with a hybrid device in about 66 to 70% of people. Most of the literature would show that with a conventional length cochlear implant, you can still save hearing in about 50% of patients.

So the difference is about 15 or 20%, but the potential advantage of using a conventional length cochlear implant in that population is if they continue to have progressive hearing loss, and they lose their low frequency hearing, they can still be remedied by continuing to use the same device and they wouldn't necessarily need to change to a different electrode, or perhaps you do lose the hearing during your surgery, and there's now left with a shorter cochlear electrode. They'll cover more of the cochlea by having a longer electrode and provide more thorough electrical stimulation for usually which provides better performance overall.

Dr. Jason Barnes:

Can you briefly describe the approach to cochlear implant? What does the procedure consist of?

Dr. Matthew Carlson:

So the surgical procedure's performed under general anesthesia. It usually requires about an hour to two hours. It involves a small incision behind the ear. Most of the time, you don't have to shave any hair, or if you do, a limited amount of hair. You expose the mastoid cortex, you perform a cortical mastoidectomy with antrotomy. You see the lateral semicircular canal in the incus. These serve as your landmarks to open the facial recess. The facial recess is an artificial anatomical corridor. It's not natural. You make the opening yourself, and it's between your facial nerve which is located posterior medially in your chorda tympani nerve, which is located anterolaterally. This gives you a window. Usually you're using a size two diamond drill to go through there, so it gives you an estimate about the size of it. It's about two millimeters, a little bit smaller. Then you can see the round window.

The wound window is located below the oval window by several millimeters. It's located inferior to your pyramidal eminence. Most of the time you'll drill off the bony overhang of the round window niche to see your full round window membrane. There's often a pseudomembrane or mucosa sitting over the top that can fool people sometimes. You can also have a real prominent intracochlear air cell track that might fool people also, but it should be immediately below your pyramidal eminence and very close to your oval window. Then there's two ways you can put the cochlear implant electrode in, or actually I should back up and say there's technically three ways. Historically, cochlear implantation was always performed through a cochleostomy. Originally cochleostomies were performed basically anywhere on the promontory. You just drilled until you saw an opening, and you put the electrode in. But as you can imagine, that can result in significant cochlear trauma.

So more and more surgeons have found that if you put the cochleostomy inferior to the round window or anteroinferior, you're more likely to enter the correct part of the cochlea. I'll talk about that in a second. The other way of putting it in the cochlea is just going through the round window membrane. This has become more popular particularly as electrodes have become smaller and smaller. You can put most of today's conventional cochlear implants through the round window membrane without any difficulty. Sometimes you have to perform what's called an extended round window approach, or some people will call it a marginal cochleostomy. I think those are two synonymous terms, and that's basically you're opening the round window and you're drilling off the anterior inferior lip of the round window just to make a bigger opening.

Your goal for electrode insertion is to put the entire electrode within the scala tympani. If you remember cochlear anatomy, you'd remember that there's three scalae. There's the scala tympani, scala media and scala vestibuli. The scala tympani is located if you look at the cross section of the cochlea, the scala tympani is located inferiorally, then there's the interscala partition that's encompassed by the osseous spiral lamina and the basilar membrane. That kind of separates your inferior compartment which is the scala tympani from your superior compartment. Superior compartment is comprised of the cochlear duct, which is also called scala media and also your scala vestibuli. The scala media is located between your basilar membrane and Reissner's membrane. Superiorly you have your scala vestibuli.A lot of data has shown at least recent data has shown that if you can put the electrode fully in the scala tympani you're more likely to preserve natural acoustic hearing, and you're probably you provide better electrical stimulation as well.

Dr. Jason Barnes:

I've heard of different types of electrodes being perimodiolar or lateral wall. Can you talk about that briefly?

Dr. Matthew Carlson:

Yeah you'll hear a lot particularly in the last 10 years about the different types of electrode designs. Besides being shorter or longer, what we call conventional hybrid are the natural shape of the electrode. So there's perimodiolar electrodes, and those are ones are also commonly called modiolar hugging electrodes. The idea is that there's memory built into the electrode where they naturally hold close to the modiolus. There's a couple characteristics that define those versus lateral wall. The other type is lateral wall electrodes. They are naturally very flexible, but straight electrodes, meaning if you pull them out of the sheath, they'd want to stand straight, and the perimodiolus will actually want to curve. You'd wonder how you can get a perimodiolar electrode with into the cochlea when it's already curving. Well most of the time they have an internal system, an internal stylet.

More recently, they can have an external sheath. That allows you to advance it through what's a more straight part of the basal turn of the cochlea and start to curl around the modiolus. There's also a third type of design, and it's called the Mid-Scala design. That has a pre curve to it, and it does hug the modiolus a little bit more. I really do classify that as a perimodiolar design, even though it's called a Mid-Scala design. There are some potential advantages between the two. I would say most commonly now, and the pendulum swings back and forth all the time, but most commonly now, people are performing round window electro insertions using lateral wall designs, and there's probably less trauma related to that.

We'll talk about both of those in a second, but there are again, advantages and disadvantages to both. So the theoretical advantages of a perimodiolar electrode are they put the electrode closer to the spiral ganglion cells, which probably gives more specific stimulation and theoretically, it might give better speech perception outcomes, although practically that hasn't been really consistently demonstrated in any study because it's putting the electrode right on the modiolus. You also theoretically have more efficient energy use, meaning you don't have to have a such high levels of stimulation, very limited potential benefit I think from that standpoint. There has been data that show that electrode impedances are less erratic than using lateral electrodes, which may have some benefit.

The primary benefits of a lateral wall electrode are that they probably limit trauma more. So they are just very slowly inserted over time. They'll follow the natural curvature of the cochlea. In contrast to the perimodiolar electrodes, they use markers, so when you're inserting it, you look at a marker and you start to deploy the stylet or the sheath, and that presupposes that all cochlea have the same size basal turn, they have the same dimensions cross sectional area and everything else. It's a one size fits all with the perimodiolar design versus the lateral wall electrode which is passive. So in my opinion and most of the studies would also demonstrate the lateral wall electrodes are safer are better for hearing preservation overall. But again, I'll say it's a very controversial aspect.

Dr. Jason Barnes:

What are some of the risks involved with surgery?

Dr. Matthew Carlson:

So fortunately, this is quite a routine surgery that's performed very often at many high volume centers, and fortunately, the anatomy is very straightforward in most adult cochlear implants. You're operating through a well pneumatized mastoid. Usually there's no cochlear malformations, and most patients don't have chronic ear disease, but there are some complications associated with it. The overall risk of a wound infection is one or two percent in adults. The risk of hematoma's probably about one percent. Some people uncommonly can have chronic surgical site pain, that's under one or two percent, and some people have persistent vestibular symptoms. Having transient dizziness is not overly uncommon, but to have somebody say that they have permanent long term dizziness after cochlear implantation in uncommon but it can occur. The risk of permanent facial nerve paralysis is less than 0.1% by most large series, and the risk of temporary or transient facial nerve paralysis is probably one percent overall.

It's usually not related directly to a direct facial nerve injury, but a secondary inflammation which is sometimes I hypothesize to be a secondary reactivation of herpes virus, much like kind of like a Bell's Palsy can happen. You can also have post-operative meningitis. There are three risk factors for developing meningitis after cochlear implantation, that's age under five or six, that's when recurrent otitis media is most common, having a cochlear malformation and particularly any that result in incomplete partition or an abnormal communication between the subarachnoid space in the inner ear. Probably the best example of this is an IP3 or gusher type malformation. But many of the malformations can result in an increased risk of meningitis. Then lastly, historically the use of a electrode positioner. In the early 2000s, there was an implant manufacturer that used a positioner that was meant to push the electrode closer to the modiolus to get some of those theoretical advantages of a perimodiolar electrode that we already talked about.

That resulted in an area for otitis media or infection to leak into the inner ear, and it would cause ascending meningitis in a group of patients. So in children, there's a elevated risk of meningitis, although it's still very, very, very rare today. In adults, the risk is even less. In adults, the risk of acquiring post implantation meningitis in a normal shaped cochlea is less than one in 1000. That risk can be even further reduced or mitigated by following the Center for Disease Control recommendations for pneumococcal vaccination prophylaxis. So currently, the CDC recommends that adult or and children regarding cochlear implantation receive a pneuma vaccination as well as the PCV13 vaccination. They cover different strains, but it's the same bug, pneumococcal pneumonia that causes pulmonary infections. It can also cause otitis media and meningitis.

So that's a good way that the risk for meningitis can be lowered. The risk of having a device failure is separate, so in children, the risk of having a device failure is a little bit higher because children are a little bit more likely to fall and hit their head on something or break their device. In adults, the risk is something that increases with time. It's an electronic device, and just like anything else, the longer its been around, the more likely it is to fail. Overall, the risk at 10 years of having a device, a hard device failure that requires replacement is two percent or less. It's highly dependent on how you define device failure and how long a person's been followed up.

Dr. Jason Barnes:

When we talk about outcomes, how do patients generally fare following cochlear implantation, and are there any factors related to the patient that might [inaudible 00:41:10] better or worse outcomes?

Dr. Matthew Carlson:

Yeah those are very important questions. I think when you look at outcomes, it all goes back to appropriate selections. So you're identifying patients who are more likely to benefit. If you're implanting a lot of people who have pretty good hearing, you're risking having them not get a lot better with a cochlear implant, versus people with more hearing loss, you're more likely to see there's more significant benefits. But if you're implanting conventional cochlear implant candidates, I would say over 90% of patients are doing better than their preoperative scores. A large percentage of patients are able to talk over the telephone with their device. I think maybe one of the best ways to answer your question is so what's the average CNC and AZBio score before cochlear implantation, and what are those average scores after cochlear implantation, and what can you get for sound perception?

So as we talked about earlier, most patients who are cochlear implant candidates have moderate to severe, or moderate to profound sensory hearing loss. A cochlear implant will usually reinstate normal pure tone levels. So you can take that really bad hearing and bring it up to being able to detect very even quiet sounds, even in the high frequencies, but more importantly is word and sentence recognition. So there's several studies. One study that's coming from our center and also a previous study from Vanderbilt has shown that the average person who's implanted with a cochlear implant has less than 10% on word and sentence scores in the ear to be implanted, which is really remarkable when you think about it because our guidelines stipulate they have to have less than 50%. So that means that it further emphasizes the idea that we're implanting people way too late with much worse scores from when they might benefit from actually getting implanted. So again, if you think somebody might be a candidate, send them for testing.

There's a good chance they are. But again if preoperative scores in the ear to be implanted less than 10% CNC and AZBio, the average scores after implantation are approximately 60% on CNC word scores, and about 75% on AZBio sentences. So that's about a six times increase in CNC scores, and about a seven times increase on average for AZBio scores, so that's a pretty big improvement for most patients. There are some things that might help you predict the outcome, but equally important is the observation that we're still pretty poor at predicting who's going to do well and who's going to do bad. The traditional prognostic indicators for having a poor outcome was a long duration of deafness. So if you're an adult and you have prelingual hearing loss, you're not going to do very well with an implant.

If you have meningitis particularly with ossificans, you have a poor outcome, and there's been different theories as to why that is, but one of them is that it's hard to get the full cochlear implant electrode insertion. But separately, the spiral ganglion cells are probably poor in that population. Patients who have cognitive impairment and central processing disorders will have more difficulty understanding that sound. There's a clinical pearl that I think is worth mentioning in that for helping to identify these patients. It doesn't work perfectly, but it should heighten your suspicion for cognitive impairment. On preimplant testing and even post implant scores, if a person so generally a person should score better on sentence testing than they should on word testing. The reason is a word has no context around it. If I just say ball, it's harder to repeat ball than if I said "I was bouncing the ball" because if you just heard "I was bouncing" you would know that the next word was probably ball and not hall for example.

So that context helps most people who have good central processing make up for gaps. So the average person who has good central processing will score better on sentences. If you have a person who scores really similarly on sentence and word scores, you should worry about it. If you have somebody that scores even higher on word scores than sentence scores, that should be a red flag in your mind that this person might have central processing disorder. Those are probably the main predictors of poor outcome after cochlear implementation, but I also think it's worth equally emphasizing that even in the best multi variate models and the best studies, we can still only account for 20 to 40% of the variance in outcomes for cochlear implant patients, meaning we're not very good at really predicting who's going to do well and who's going to not do as well with an implant.

Dr. Jason Barnes:

I'd next like to move into our summary, but before I do, is there anything else you wanted to add?

Dr. Matthew Carlson:

I think that summarizes cochlear implants pretty well. No I don't have anything else to add.

Dr. Jason Barnes:

So in summary, patients presenting for a cochlear implant evaluation commonly present with progressive hearing loss over time, and hearing aids not working over several years. Work up includes audiometry with pure tone thresholds, and word recognition scores, and then CI evaluation with includes CNC word scores and AZBio sentence scores. The actual cochlear implant is made up of a microphone and processor with a transmitting coil, that transmits the sound signal to a receiver stimulator, and then the channel electrode that's implanted. The cochlear implant works by stimulating the spiral ganglion cells in the cochlear nerve. There are conventional types of cochlear implants which are standard length or the full length of the cochlea, and hybrid types which are shorter in an effort to preserve the function of low frequency hair cells. Then there are also perimodiolar and lateral wall electrodes that could theoretically be beneficial for different reasons.

The actual procedure involves a mastoidectomy, and then approach through the facial recess and placement of the electrode into the round window and scala tympani. Risks for this procedure include facial nerve weakness, meningitis and device infection, but all of these risks are very low. Patients tend to do worse if they've had a long duration of deafness, have a history of meningitis, or cognitive impairment, but overall post operative scores come up significantly for both word and sentence scores to the tune of six to seven times improved. Dr. Carlson, anything you'd like to add?

Dr. Matthew Carlson:

I think that summarizes everything well in a nutshell.

Dr. Jason Barnes:

Well thanks so much for being here.

Dr. Matthew Carlson:

Thanks for having me.

Dr. Jason Barnes:

I now want to move into the question asking portion of the episode. Recall that I'll ask a question, wait a few seconds for you to press pause or to contemplate the answer, and then I'll give the answer. So the first question is what is the audiologic workup for CI candidacy? Generally patients are worked up for hearing loss starting with a standard pure tone audiometry and word recognition scores provided that their word recognition is usually less than 50%. That would be a tip off that they should undergo CI candidacy which includes additional work up like CNC word scores and AZBio sentence scores. For the next question, what are the guidelines for cochlear implantation today? To succinctly state it, the guidelines for receiving a cochlear implant are that a patient has to have less than 50% sentence scores in the ear to be implanted, and less than 60% in the best-aided condition, and they cannot be better than moderate to profound hearing loss.

In terms of specific Medicare guidelines, they need to have moderate to profound hearing loss with less than 40% in the best-aided condition. Again, these guidelines are somewhat vague and allow different interpretations and signal to noise ratio is applied. Finally, for our last question in regard to scala location, what is the idea location for placement of a cochlear implant electrode? Again, when we discussed the procedure of cochlear implantation, this involved a mastoidectomy and facial recess approach. The idea location for the electrode is in the scala tympani, and this could be accessed through the round window through an extended round window approach, or a cochleostomy if clinically required.

Thanks so much, and we'll see you next time.