Alyssa Smith:

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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 neurotologist Dr. Brad Kesser, and we'll be discussing congenital aural atresia. Dr. Kesser, thanks so much for being here.

Dr. Brad Kesser:

Thanks for having me, Jason. I appreciate the opportunity.

Dr. Jason Barnes:

Congenital aural atresia, it might sound a little bit straightforward, but can you tell us how children typically present to your clinic with congenital aural atresia?

Dr. Brad Kesser:

Children typically present in one of a couple ways. The deformity is often seen in association with microtia so it's seen from the day of birth, and it's not unusual for me or one of my residents to be called to the newborn nursery just to evaluate a child with "an ear deformity." The other way that they present to me is once they've kind of gotten out of the hospital in the ambulatory setting, child presents to the clinic with an ear deformity and then we go from there and I see them as an outpatient.

Dr. Jason Barnes:

Can you describe some of the epidemiology related to congenital aural atresia? How common is this and what is it associated with?

Dr. Brad Kesser:

Sure. The quoted rates of incidents of aural atresia is about one in 10,000 to about one in 20,000. So it's not terribly common, but in the course of someone's practice, you'll see that several times. I think it's important to be prepared and armed with how we work up children with aural atresia and what are the best and optimal options for hearing habilitation in these children. For reasons that we don't know, boys are typically more affected than girls. The right ear, again for reasons that we don't know, is more commonly affected than the left ear. It can also be associated with syndromes. Associated syndromes include Goldenhar or hemifacial microsomia, Treacher Collins where we see microtia atresia in both ears, sometimes in Crouzon and Pierre Robin. So I think as we initially evaluate children with aural atresia, it's important to get kind of a 30,000-foot view and look at the facial structure of the child to try to detect any other associated abnormalities.

Dr. Jason Barnes:

And that kind of goes to my next question. When you first evaluate these patients in clinic, what are you looking for on physical exam?

Dr. Brad Kesser:

Right. Prior to the physical exam, I will ask the family if there's a family history of microtia or aural atresia or a family history of hearing loss, were there any maternal drug exposures or maternal toxicity exposures, and just ask really about a brief prenatal, perinatal, and postnatal history for the child much as I would do any child with hearing loss. I think the other important consideration is whether the microtia atresia is unilateral or bilateral. Obviously, we can detect that on our physical exam, just looking at the auricles. If both auricles show a degree of microtia, again, it's important to look and determine if there's also associated aural atresia where the ear canal didn't develop. Sometimes there's a grade two microtia, and we can talk about the classification in just a second, with a stenotic ear canal and that's important to document. Other physical exam findings that are important to document include any ear pits, ear tags. Look along the sternocleidomastoid muscle for cysts or sinuses. Branchiootorenal syndrome is another syndrome that's associated with aural atresia. A quick survey in the medical record for any kidney or renal problems or vertebral problems can also be associated with microtia and aural atresia.

Dr. Jason Barnes:

And how often do you see a decrease in facial nerve function in these children?

Dr. Brad Kesser:

Not terribly often. Certainly in the children with Goldenhar, hemifacial microsomia, we can see some reduced facial motion on the affected side. As I've looked at people's smiles over the years and being an otologist, you'd be surprised how common a congenital weakness of the marginal mandibular branch of the facial nerve is. So definitely in terms of physical exam, we look for the crying response or any kind of startle response that might give us an indication as to facial nerve function.

Dr. Jason Barnes:

When you first visit with these patients, we'll come to find out that there aren't always actionable steps at the first visit. How do you counsel families in terms of other things regarding the child's care for the next several months or years apart from the workup and maybe surgical intervention?

Dr. Brad Kesser:

Right. So that involves kind of the next step in the evaluation. So once we've taken the history and done the physical exam and determined is this a bilateral problem or a unilateral problem, the next most important step in the evaluation is a hearing test. Now, I always ask about the results of the newborn hearing screening and if the child has bilateral aural atresia, it's very likely they failed on both sides. If there is a unilateral aural atresia, it's possible or actually likely that they would have passed in the normal structured ear and failed in the microtia atretic ear. That obviously is not good enough and so I recommend ABR testing within the first three months of life to document air conduction thresholds for the normally structured ear, the ear without microtia atresia, and I also think it's important to document bone conduction thresholds by ABR for the atretic ear. I want to know whether the inner ear, the cochlea is functioning in that atretic ear. It may not make a difference as to my treatment recommendations, but I think it's very important to document that the normally structured ear is normally hearing and the bone conduction thresholds for the atretic ear. That we can do in the first three months with an ABR really in the office. The mom feeds the baby, the baby goes to sleep, and then we can usually get reliable air and bone conduction ABR thresholds.

Dr. Jason Barnes:

I next wanted to move onto pathophysiology. Sometimes for residents, it's hard to talk too much in detail about embryology, but this certainly requires some discussion of embryology. Could you describe to us maybe some of the embryology around congenital aural atresia and what happens in children with congenital aural atresia?

Dr. Brad Kesser:

Sure. I mean this is a classic embryological problem and I think there are really two important points to understand about the embryology of the ear that has absolute implications for our clinical management of these children. The first point is that the inner ear, the cochlea and the auditory nerve derive from a completely separate analogue as compared to the middle ear and the ear canal. In other words, the inner ear, the cochlea and the auditory nerve develop from the otocyst, the neuroepithelium. However, the ear canal and the middle ear develop from the branchial arch apparatus. The implication for this embryologic division between the inner ear and the outer and middle ears is that most children with aural atresia have normal inner ear function. In other words, the otocyst has developed normally along its embryologic path and it's usually fairly well developed by the 12th week of gestation. However, there's been an arrest in the development along the branchial apparatus usually much later in development where the middle ear and the ear canal are hypoplastic. The ear canal didn't develop and the middle ear, usually there's an ossicular abnormality along with reduced middle ear volume.

The second point that I'd like to make about the embryology is that congenital aural atresia is really a first arch abnormality. The first arch as everyone remembers is the ear canal and really kind of what you cannot see through the eardrum. In other words, the head of the malleus and the body of the incus. So the implication for aural atresia being a first arch abnormality is that the second arch structures, namely the stapes and the facial nerve, are generally normally developed or fairly well developed. So the typical middle ear atresia or middle ear hypoplasia that I see is a smaller head of the malleus with an absent manubrium of the malleus, the body of the incus is usually fairly well formed although it may be a little bit hypoplastic with a lenticular process and incudostapedial joint that is fairly normal and a normal stapes bone, and then the facial nerve can take a lateral and more anterior course.

But I think those two embryologic points are really important to know because they have implications clinically. Number one, the inner ear develops separately. So children with aural atresia generally have normal bone conduction thresholds, but you got to do the tests, the ABR test to make sure. Number two, aural atresia is typically a first arch abnormality so that there is maybe some hypoplasia of the mandibular, Meckel's cartilage, as well as the fused malleus-incus complex so that there aren't separately developed malleus bones from the incus bone, and that the second arch derivative, the stapes bone, is usually fairly well developed.

Dr. Jason Barnes:

Do we know if there are any genetic risk factors for congenital aural atresia?

Dr. Brad Kesser:

There is one named genetic abnormality and that's distal 18q deletion. This is a very unusual condition in which children have normal auricles, so they do not have microtia, but they have complete atresia or very, very stenotic ear canals with associated conductive hearing loss. Most unilateral isolated patients with aural atresia do not have a known or named genetic abnormality.

Dr. Jason Barnes:

So next, I wanted to go onto workup. We've talked a little bit about the ABR and the importance of obtaining that early. What else are you considering in terms of workup for these patients?

Dr. Brad Kesser:

Right. So the workup in the first year of life really is the ABR and then as the child gets older, behavioral audiometry, either visual reinforced audiometry or conditioned play audiometry. I want to stress the CT scan can wait and in my opinion should wait. There's no reason to get a CT scan on a one-year-old with aural atresia because we're not going to be doing anything about the aural atresia surgically anyway and you're delivering a small dose of radiation to a small child. So the total body dose of radiation is higher than if you wait til the child is five or six to get the CT which is the time we're starting to consider canal surgery. So I think it's very important to get hearing data, but I also think it's very important to wait before getting the CT scan. As far as other workup, if the child has Treacher Collins or looks syndromic, I think you could have a genetics workup, but as I mentioned earlier, isolated unilateral aural atresia in the absence of any other syndromic findings does not have a known or named genetic etiology.

Dr. Jason Barnes:

And you suggested that the CT scan can wait, meaning that there won't be an intervention for some time. In that time period, what are some things you counsel patients on or parents on in terms of things to be aware of, the unaffected ear, and maybe how they're going to develop and learn?

Dr. Brad Kesser:

Right. So good question and I think we'll start with the easy clinical scenario first and that is the child with bilateral aural atresia. I think no one would argue that a child with bilateral aural atresia needs to be fitted with a bone conducting hearing device. Now, the FDA has not approved the osseointegrated bone-conducting hearing devices. That would be the BAHA Connect, the BAHA Attract, the Oticon Ponto for children under five years of age, but these children do need amplification and that comes in the form of either a soft band or a hard band where the bone oscillator is fit to the mastoid cavity or fits to the mastoid bone and the child is amplified through bone-conducted sound. That will certainly promote speech and language development for the child with bilateral aural atresia.

Now, many parents ask, "Do I need bone-conducting hearing aids for both ears in my child with bilateral aural atresia?" There's no data at this point to support using bilateral bone conductors. Some parents do want to go down that road. I would say in my clinical experience, again I haven't seen any data, but in my clinical experience one bone conductor is sufficient for normal speech and language development in these children. I don't know that you need two bone conductors. Two bone conductors will not give the child sound localization and we don't know about hearing and background noise, but my suspicion is two is not necessarily better than one when it comes to sound localization. So that's the case, that's the child with bilateral aural atresia. Absolutely needs a bone-conducting hearing device to support speech and language development.

Now, the child with unilateral aural atresia and a normal hearing ear is a little bit of a ... I wouldn't say controversy, but do we put a bone-conducting hearing device on every child with unilateral aural atresia and a normal hearing ear? First, let's talk about the normal hearing ear. I tend to be a little bit more aggressive should that ear develop otitis media with effusion in putting a tube in. I think a tube if carefully placed is a low-risk procedure and certainly will help with hearing and support speech and language develop since the normal hearing ear is really the only hearing ear that that child has for speech and language development. So I do tend to be a little bit more aggressive placing a PE tube in a child with OME, otitis media with effusion, in the normal ear and atresia on the other.

The question about a bone-conducting hearing device in a child with unilateral normal hearing and the other side aural atresia is a difficult one, and my approach is I never discourage families from putting a bone-conducting hearing device on their child with unilateral atresia, but I'm never so dogmatic as to say, "If you don't put a bone-conducting hearing device on your child, your child will never develop speech and language." We know that normal hearing in one ear is sufficient for normal speech and language development. So I give parents the option. We do know that the earlier that a bone conductor is placed on the child, the better that the child is able to integrate the signal.

So for example, I have parents who try them on their three and four-year-olds and man, they will not keep a bone conductor on because at the end of the day, that bone conductor with normal hearing in one ear does not give the child better sound localization, it does not help the child with hearing in background noise. So at the end of the day, it's a strap they got to wear around their head and it's uncomfortable and it can be hot and there are cosmetic issues and the three or four or five-year-old ends up just throwing the bone conductor off the head. So again, I never discourage families from trying bone conductors, but I also reassure them that we have a lot of battles to fight as parents and having your five-year-old with normal hearing in one ear wear a bone-conducting hearing device on the atretic ear is not necessarily a battle that we have to fight.

Dr. Jason Barnes:

Now, you mentioned the possible insult to speech and language development. How do you counsel parents during this time on pursuing speech therapy and those kinds of therapies?

Dr. Brad Kesser:

So I tell parents, "You need to do all the things that you know as good parents to support speech and language development. Read to your child. Talk to your child. Sing to your child. Do all those things." I do support speech therapy as an initial evaluation. Most of these children are normally developed with regard to speech and language and don't necessarily need speech therapy. Again, same with the bone conductor, I'm not so dogmatic as to say, "Your child needs to be enrolled in speech therapy tomorrow," but I certainly support a family's desire to go down the road of speech therapy. Most of the time the speech therapist says, "Your child with unilateral aural atresia is developing appropriately." With bilateral atresia, I tend to be a little bit more aggressive in at least having the child evaluated for speech as the child hits one, two years old just to make sure that the child is on task and on his or her curve for speech and language development.

Dr. Jason Barnes:

There's some additional considerations when taking care of children with aural atresia, and I was hoping we could talk about a few of these briefly. Could we first start with microtia? Could you tell us about how you evaluate microtia, what the grading system is, and how this plays a role in your potential timing of surgical options?

Dr. Brad Kesser:

Sure. We still use the Altman classification for microtia where grade three is ... Well grade four is anotia. There is nothing there. Grade three is the classic peanut ear where there's a small remnant auricle with maybe a little bit of cartilage. Grade two is a relatively well-formed helix but missing some of the interior cartilaginous components, the triangular fossa or the antihelix. Also, the auricle is smaller. Grade one has all of the normal cartilaginous components, but is just smaller. So with microtia, it's obviously always important to look in the canal and see if there is a canal. We define stenotic canal as less than four

millimeters, and there was a study by my mentor Bob [Jahrsdoerfer 00:20:24] and [inaudible 00:20:26] that looked at children with less than four millimeter diameter canals and showed a higher risk of congenital, or not so much congenital, potentially acquired, but ear canal cholesteatoma. In other words, the canal is so stenotic that the natural epithelial migration is disrupted. The dead skin can't get out and the skin becomes trapped in the canal.

Now, that is an indication for surgery and if there is a stenotic canal, I always ask about drainage, ask the parents, "Have you ever seen drainage from the canal? Have you ever seen anything come out of the canal like dead skin, keratin? Does it have a foul odor? Does the child ever complain of pain?" Those are things that would push me to get a temporal bone CT scan sooner, looking for the possibility of an ear canal cholesteatoma in the setting of a stenotic ear canal. There are certain CT scan findings that are suggestive of the canal cholesteatoma. There's bony remodeling. The dead skin can kind of bulge from the canal into the middle ear. There's a widening of the canal in certain areas. So I don't see canal cholesteatoma in children under four. By four, four and a half, five, that's when I might consider getting a temporal bone CT scan in the setting of a stenotic canal, looking for the possibility of a canal cholesteatoma.

Dr. Jason Barnes:

And talking a bit more about the temporal bone CT scan, what would you say is the optimal timing for obtaining the first temporal bone CT scan? Maybe what are some indications for getting one earlier? Can you talk us through how you evaluate this and what kind of criteria or classification systems are used?

Dr. Brad Kesser:

The temporal bone CT scan, as I mentioned earlier, is not necessary to do until we're starting to think about surgical reconstruction of the ear canal and the middle ear system. In the setting of grade three microtia with complete atresia of the ear canal, if the family is not interested in atresia surgery or canal surgery, there's no reason to get a temporal bone CT scan. I don't necessarily think that you have to get a CT scan just to take a look or to see what's going on. All that you need to know is whether the inner ear is working or not and whether that child is a candidate for a bone-conducting hearing device. Obviously if the inner ear is not working which is pretty rare, there was a study by Jeff [Rayback 00:23:12] that showed about a 19% incidence of structural abnormalities of the inner ear that are associated with aural atresia. That's not really been my experience, although those structural abnormalities included the facial nerve and there are some facial nerve abnormalities associated, as I mentioned earlier, with microtia atresia.

But in any event, my clinical experience has been that the bone conduction threshold is the inner ear is working fine even if there is a higher 10 to 15 or 19% incidence of structural inner ear abnormalities seen on temporal bone CT scan in atresia patients. So if the family is not interested in canal surgery and there's complete atresia of the ear canal, the CT scan is not even necessary. If the family is interested in canal surgery, then it is the temporal bone CT scan by which I assess their anatomic candidacy for surgery. So they have to have normal bone conduction thresholds obviously if we're going to try to open the ear canal. We don't want to open the ear canal in a deaf ear. So normal bone conduction thresholds and then I still use Bob Jahrsdoerfer's classic 10-point grading scale, looking at each anatomic structure and assessing each structure one point and giving the stapes bone two points and adding up the points to determine candidacy for atresia surgery. That has been, in my hands, extremely valuable. It is not only good to assess candidacy, but I think in some ways it is somewhat predictive of hearing outcomes. Children with better anatomy will have better hearing outcomes.

We showed several years ago that children with seven, eight, and nine will have as high as an 80 to 90% chance of achieving normal or near normal hearing with atresia surgery whereas children with six or below had about a 40 to 45% chance of achieving normal or near normal hearing with atresia surgery. So I carefully pore over these temporal bone CT scans, looking at each anatomic structure, assigning the points, two points to the stapes bone, point for the oval window, one for the pneumatization or the middle ear volume. I think middle ear volume is an important predictor also. If you have a nice wide, large middle ear volume, you've got room to open the middle ear space and there's also room for the ossicles to vibrate. If it's a narrow, constricted middle ear space, I think that's a greater risk for loss of hearing gains longterm after surgery. In other words, a subset of children after canal surgery will do well, but over time, one year, two years, three years, will lose their initial hearing gains. So their bone conduction thresholds stay the same, but their air conduction thresholds decline. I think that's secondary to either lateralization of the eardrum or re-fixation of the ossicular chain. I think a larger middle ear volume protects or prevents against re-fixation of the ossicular chain.

Again, just going down the list, I give a point to the facial nerve, the malleus-incus complex, mastoid pneumatization, the incus-stapes joint, IS joint, round window, and then of course almost everyone gets a point off for the external ear if they have any degree of microtia at all. So in summary, I find the Jahrsdoerfer grading scale, the 10-point grading scale that has been around for, gosh, 20 years now, 30 years actually, I think he published it in '91, to be reliable, a good predictor of hearing outcomes, and the means by which I assess a child's anatomic candidacy for canal surgery.

Dr. Jason Barnes:

And moving onto canal surgery, we've talked around it a little bit this episode. Can you tell us how you counsel parents on their options for the treatment of congenital aural atresia, both atresiaplasty but also other options moving forward?

Dr. Brad Kesser:

Sure. So essentially if the child has bilateral aural atresia, as I mentioned earlier, absolutely with a bone-conducting hearing device. When the child is five or over, we then talk about surgical options for hearing habilitation, surgical options being the osseointegrated bone-conducting hearing devices versus canal surgery. So I really go through the risks and complications of both and/or the family can elect to stay with the bone conductor on a soft band or a hard band. In children with unilateral atresia, again, the options would be observation. You don't have to do anything if the other ear is a normal hearing ear. There is the option of a bone-conducting hearing device, but I tell families that in a normal hearing ear on one side and an atretic ear on the other, a bone-conducting hearing device will not allow the child to improve sound localization and will not improve hearing in background noise. It's very hard to get children with a normal ear to wear a bone conductor. Then of course canal surgery.

Areas of my research interest include sound localization and hearing in background noise in children with unilateral aural atresia who have undergone either bone-conducting hearing device or canal surgery. What we found with sound localization is that they do improve their sound localization after surgery, but it is not as good as two normally hearing ears. In addition after surgery, children do hear better in background noise after canal surgery. So I think there are definite advantages and in the anatomically favorable candidate, canal surgery can be a great boon to their quality of life.

Dr. Jason Barnes:

And if atresiaplasty or ear canal surgery is pursued, what is the timeline or when are you offering this intervention? Why do you use that cutoff and how do you coordinate it with possible microtia repair if that's also desired?

Dr. Brad Kesser:

Right. Those are very important points. I'll take the second one up first. The timing of atresia surgery is greatly dependent upon the family's choice for microtia repair. So if there's a grade two microtia or even a grade one microtia and the family is not considering microtia repair, then it's certainly easy to just go ahead and do the canal surgery and put the meatus where it belongs, where the auricle is. If the family is going to choose rib graft microtia repair, then the rib graft microtia repair goes first and the atresia surgery, the canal surgery goes after the rib graft microtia repair. The reason is if I make an incision in that postauricular area, that tissue becomes scarred and the rib graft microtia surgeon can't develop the pocket necessary for the sculpted rib graft. So canal surgery after rib graft.

If the family chooses the MEDPOR or the Su-Por, a porous polyethylene implant for the microtia repair, then the canal surgery goes first. The reason is if I were to do the canal surgery after the polyethylene implant, the MEDPOR goes in and I inadvertently expose that MEDPOR polyethylene, it will not heal. So they have to go back for additional surgery to cut back the MEDPOR and develop a flap to cover the exposed area. So it's very important to work with your plastic surgeon such that the canal surgery goes before the MEDPOR and goes after the rib graft.

Now, as far as timing, how old or what is the ideal age to do canal surgery, I truly feel that the youngest I would recommend is probably around five and a half. Now, I will give you the caveat that there are four-year-olds that I have done in the setting of canal cholesteatoma and they've done fine, but my feeling is that at five and a half to six or even seven, there are several features that make doing it at this age much preferred. Number is the maturation of the eustachian tube. If children have eustachian tube dysfunction, I don't want my beautiful middle ear and ear canal and then have to fill them with fluid or they get otitis media and have to put a tube in a reconstructed ear. So let all the eustachian tube issues pass and let the child's eustachian tube and skeletal framework mature before we do the canal surgery.

The second is co-operability. There are some very good three-year-olds and there are some not as cooperative three-year-olds, but by the age of five or six, children understand what's going on. They know they're going to have their ear operated on and they're going to hear after their ear and they jump up in the chair and they allow me to do the necessary postoperative care to ensure a good result. That postoperative care involves removing the packing from the ear canal that I put in at the time of surgery, that's one week after surgery, and then cleaning the ear one month after surgery where I elevate the dead skin layer and peel away that desquamated epithelium. So the older child just gets it and understands and is much more cooperative than the younger child.

I also think, as I mentioned earlier, we don't need to get the CT scan at the age of one or two. Let's get the CT scan at the age of five or six when the total body dose of radiation is much smaller and let's get it one time. So we don't have to get it at the age of one and then it doesn't look that great and so we decide, "Okay. Let's get another one at the age of five to see if there's better pneumatization of the mastoid and the middle ear space." So I think earliest is five and a half to six unless there's evidence of canal cholesteatoma in which case I would go a little bit earlier at four, four and a half.

I would also mention that I do not do the combined microtia atresia surgery. I do not think that is in the best interest of the child, and so you may hear about combined microtia atresia and so that is not something that I do or that I recommend. One other issue about doing it at the age of maybe five and a half or six is that I think they may be less likely to stenos at that age compared to a three or four-

year-old. That's just purely anecdotal. There's no data to support that statement, but I've just found that the older children, they can certainly tolerate if we need to do a steroid injection. They can tolerate a steroid injection and I think they're potentially less likely to stenos.

Dr. Jason Barnes:

And could you briefly walk us through the procedure of atresiaplasty and what your awareness is of the facial nerve and its course during this procedure?

Dr. Brad Kesser:

So very important to track the facial nerve on the CT scan to make sure that it is not so anterior and so lateral that it is in the drill path of your drill, but essentially you make a postauricular incision or a small incision behind the auricular remnant if I'm doing the surgery prior to MEDPOR, and then get down to the temporalis fascia. We'll harvest some fascia and then make periosteal incisions along the linea temporalis just like we do for chronic ear and then anteriorly along the glenoid fossa so it's actually a posteriorly based mastoid periosteal flap that then is reflected posteriorly. Under the microscope, then I elevate the mastoid periosteum anteriorly to define the three really important bony landmarks. I don't drill until I've absolutely defined the root of the zygoma, the posterior bony ledge of the glenoid fossa, and the mastoid tip. Now, I don't completely expose the mastoid tip, but I just want to know kind of where the mastoid tip is starting.

So using those three bony landmarks, I start with a five-millimeter cutting drill burr and I stay anterior and superior, so kind of in the angle between the linea temporalis, root of the zygoma, and the posterior bony margin of the glenoid fossa. I stay right up there anterior superior, and the first thing I do is find the tegmen. Once I find the tegmen and that is where the air cells end, I follow the tegmen medially and as the tegmen courses medially, the air cells start to get bigger until I open into the epitympanum. Once I've opened into the epitympanum, I identify the fused malleus-incus complex and use progressively smaller drill burrs, three, two, 1.5 millimeter diamond drill burrs to drill away the atretic plate, isolate the ossicles, and then the final portion is to take a small 59 Beaver blade and incise the ligamentous attachments between the neck of the malleus and the atretic plate. That frees the ossicular chain so that the chain is mobile.

Next thing is just the harvest the skin graft. I've been taking the skin graft lately from the upper lateral thigh. Actually, I take the skin graft with the two-inch guard on a Zimmer Dermatome with 0.004 to six inches, extremely, extremely thin skin graft. It's a tough part and I've had to take them a couple times in a few patients, but it's got to be very, very thin. Then once I've taken the skin graft, I'll place the temporalis fascia graft in an overlay fashion with care taken to drape one or two millimeters of the fascia up onto the canal wall. The entire middle ear space has to be protected and covered by the fascia graft including the aditus. Then I put the skin graft in. I notch the end of the skin graft, the medial end of the skin graft, and align the notches over the temporalis fascia such that the entire fascia graft is covered by the split thickness skin graft. The skin graft I drape up onto the bony canal wall. So I work my way around clockwise and the edges of the skin graft meet anteriorly.

Once the skin graft is in, I place a little 0.04-inch Silastic disc over the skin graft and fascia graft and then place 3/4 length Merocel wicks. I hydrate the wicks with Floxin Otic solution, Ofloxacin otic solution, and then I've either done the meatoplasty if it's a MEDPOR. I just suture the skin graft to the native skin around just behind the auricle in a circumferential fashion, in a circular fashion, or if it's a MEDPOR, I will carve out the meatus from the rib graft and then sew the rib graft back down so the postauricular incision and then deliver the skin graft out through the newly created meatus and put additional packing in. So that's essentially the operation of atresiaplasty.

Dr. Jason Barnes:

Could you speak to how you counsel parents on outcomes in terms of prognosis, hearing outcomes, and when do you find you need to use a prosthesis and how does that affect outcomes?

Dr. Brad Kesser:

So it's fairly rare that I use an ossicular prosthesis, although I have used them for sure. I would estimate maybe 5%, maybe even as high as 10% I'll have to disarticulate the incudostapedial joint, remove the fused malleus-incus complex, and place a partial ossicular replacement prosthesis on the mobile stapes bone. Times when I do that are times when I just can't seem to mobilize the ossicular chain without drilling on the ossicular chain. Now, I don't want to drill on the ossicular chain because that high energy of the drill would be transmitted through the stapes and into the cochlea and give the patient a sensory neural hearing loss. So those are the situations where I would disarticulate the IS joint and put a porp on.

As far as hearing outcomes, I tell parents a couple things about the expected outcomes. Number one, I can never make the reconstructed ear as good as the normal hearing ear. Number two, I cannot bring every frequency of sound into the normal range. My goal is to bring the human voice range, 500 to 3000 or 4000, into the normal range. So with good anatomy ... And I will tell you I cherry pick these. If the anatomy isn't favorable, I will not offer surgery. So with favorable anatomy, I can generally get those mid-range tones in the normal range in probably 80 to 90% of children. It's extremely gratifying, extremely rewarding. We don't, as we mentioned, know the learning effects. We don't the sound localization and hearing in background noise effects longterm. I mentioned them, I alluded to them earlier in the show where after surgery, children can hear better in background noise and do locate sounds somewhat better than they did before surgery, but definitely not as well as two normal hearing ears.

What we don't know if whether there's a learning effect, whether they are able to over time integrate the new signal from the new ear into tests of binaural hearing. So what I'm trying to work on is some longterm results looking at how do they locate sound in speech and how well do they hear in background noise a year or a couple of years after surgery. But I think those couple of points early in my answer, I can't make your hearing as good as the other ear and I can't get every frequency in to the normal range, are really important to tell parents before surgery.

Dr. Jason Barnes:

And you mentioned earlier kind of your postop follow up with debridement, but how do you follow up with these patients longterm?

Dr. Brad Kesser:

So the postop care, right, one week to remove packing, one month to clean the ear, and I tell parents and the patient that you're going to need to have the ear cleaned once or twice a year for the rest of your life. The reason is a normal canal has this epithelial migration phenomenon where skin migrates or skin moves in the ear canal, and that's how the ear canal cleans itself. There's lots of research that puts ink spots on the eardrum and watches the ink spots over time, and they over time will migrate out to the lateral canal. When you lay a skin graft in there, the skin doesn't move. There is no epithelial migration. So the cleaning involves carefully lifting off the dead skin layer and peeling that dead skin away.

It's kind of like peeling the dead skin off a sunburn. It doesn't hurt and actually I will tell you these kids are insensate in the canal so you don't have to be absolutely meticulous in your technique. I

mean if you bang a little bit, they're not going to feel it. So you have to just elevate that desquamated epithelial layer and peel that dead skin away. A lot of patients know when it's time for a cleaning because their hearing gets a little bit worse. I think that dead skin layer somewhat tethers the eardrum and so when you clean that dead skin layer out, it's like you open the ear up all over again and they notice improvement in their hearing.

Dr. Jason Barnes:

Well Dr. Kesser, this has been such a great discussion about an interesting topic. I really appreciate you being willing to be on this episode. Before I move into our summary, is there anything that we haven't talked about that's worth mentioning or adding?

Dr. Brad Kesser:

I'll just say a quick word about complications. Facial nerve injury is very, very rare as long as you've plotted out and tracked the facial nerve on the CT scan and understand it's course. Facial nerve lives medial to the ossicle so you're going to hit the ossicles before you hit the facial nerve typically. The biggest risks are stenosis and loss of hearing over time. Patients can also have some drainage. I'll talk about drainage first. Sometimes the skin graft or part of the skin graft dies. It's up against an air cell or it's up against some mucosa and it what I call [mucusalizes 00:45:47]. It turns into mucus membrane and that mucus membrane can be moist. If it's small areas, I can treat that in the office with just either some antibiotic powder or some silver nitrate. Larger areas that mucosalize and just weep all the time probably require revision surgery.

With regard to stenosis, the best treatment for stenosis is prevention. I will often put Kenalog, inject Kenalog in the perimeatal skin before the child wakes up to get a little Kenalog going to prevent stenosis. I can also inject Kenalog in that perimeatal skin in the office. I mix it with a little lidocaine. So the first injection is a little uncomfortable, but kids seem to tolerate it pretty well. Then I'll have parents use a foam earplug coated in a water-soluble lubricant like Surgilube or KY Jelly at night so that they scrunch it up in their finger, they lubricate it, they slide it in, it expands, they sleep with it overnight, and then they take it out in the morning. Sometimes they have to do that for three to six months to prevent stenosis. So that can be a challenge.

Then finally, some children will lose some hearing over time. I think that could be secondary either to lateralization of the eardrum or re-fixation of the ossicular chain. If it's not too bad, they could certainly wear a conventional hearing aid in their new ear canals. So it's not unheard of for families to go down the road of a conventional hearing aid, and that is quite adequate in amplifying sound. The other option would be to go back in and either release the ossicular chain that has become re-fixed or to do a new tympanic membrane graft if the tympanic membrane has lateralized.

Dr. Jason Barnes:

Well Dr. Kesser, thanks again so much for this discussion. I'll now move into our summary. Congenital aural atresia is a middle ear deformity that's often seen in conjunction with microtia and can be associated with various pediatric syndromes. Workup includes ABR and can include a CT scan later on down the road if intervention is being considered. The Jahrsdoerfer criteria is a grading scale used to assess the degree of aural atresia and can be used in choosing correct surgical candidates. Treatment options for atresia include bone conduction with possible prosthesis versus canal surgery or atresiaplasty. Atresiaplasty is a complex surgery. So you need to be considering the facial nerve and this requires thoughtful preoperative evaluation. In terms of outcomes when the correct patient is selected,

they can be very good in terms of bringing hearing levels back to the normal level and speech thresholds.

Dr. Kesser, anything else you'd like to add?

Dr. Brad Kesser:

Jason, only to thank you for the opportunity to speak with you about my second passion, secondary to my family, but it's been a great session talking with you. Again, thank you for the opportunity.

Dr. Jason Barnes:

Yeah, thank you so much. This has been great.

I'll now move onto the question-asking portion of our time. As a reminder, I'll ask a question, wait a few seconds, and then give the answer. So the first question is what are some pediatric syndromes that can be related to congenital aural atresia?

Some of the more common pediatric syndromes seen with congenital aural atresia are Goldenhar syndrome or hemifacial microsomia, Treacher Collins, Crouzon, and Pierre Robin.

For the next question, what are the four grades of microtia?

So reviewing these Altman classification system, grade one is a small ear with most of the key features. Grade two is with some features missing, the two-thirds of the ear is present. Grade three is what's classically referred to as the peanut ear. Grade four is total anotia.

For our next question, what are the aspects of the Jahrsdoerfer criteria? Recall that there are 10 total points.

So to review these criteria as Dr. Kesser said, the presence of a normal stapes is given two points, and then the following are given one point: the oval window being open, the middle ear space being present, the facial nerve having a favorable path, the malleus-incus complex not having severe deformity, mastoid pneumatization, the incus-stapes connection or the IS joint, the round window being open, and the appearance of the external ear.

For our next question in terms of the Jahrsdoerfer criteria, what score portends a good outcome for atresiaplasty?

Although not specifically mentioned in this episode, a Jahrsdoerfer criteria of seven portends a much better outcome and atresiaplasty is often not offered to patients with a Jahrsdoerfer score of less than seven.

For our last question, when should atresiaplasty be offered to children?

Again, as Dr. Kesser described, atresiaplasty isn't often offered before the age of five and a half and the main reasons for this are that the eustachian tube is more fully developed which decreases the risk of fluid collection in the newly created ear space and also kids are able to tolerate postsurgical debridement as well.

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