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Candidacy Considerations for Bone Conduction Hearing Devices in Pediatrics Recorded June 2, 2020

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- [Ravi] Everyone, this is Ravi Sockalingam, from Oticon Medical, U.S. I would like to take a few moments to first and foremost, welcome all of you to the Oticon Medical webinar series, a seven part series that runs for seven consecutive weeks, except for the week of the 4th of July. Today, we are very fortunate to have Dr. Laurie Mauro, a senior pediatric audiologist from the Children's Hospital of Philadelphia. Laurie has done incredible work in the field of bone conduction hearing, particularly from a clinical perspective. She is an integral member of a working group on Bone Conduction Hearing Devices, totally dedicated to developing evidence based and best practice guidelines. So today, Laurie will speak to us on the candidacy considerations for bone conduction devices in children. So I'll just pass the mic over to Laurie and thank you Laurie.

- [Laurie] Great, thank you. Good afternoon, everyone. Again, my name is Laurie Mauro from Children's Hospital of Philadelphia. I'll be discussing candidacy considerations for Bone Conduction Hearing Devices in pediatrics. Throughout today's presentation, I'll be referring to Bone Conduction Hearing Devices as BCHD. As Ravi said, this lecture is the first presentation of a bone conduction seminar series that's hosted by Oticon Medical. At the end of this course, you should be able to explain the non surgical and surgical audiologic candidacy criteria for BCHD in infants and children, identify specific concerns or challenges that impact children with unilateral hearing loss, list features desired in a pediatric BCHD, and identify the benefits of fitting BCHD's to children. Today I'll be discussing both non surgical and surgical audiologic candidacy criteria for BCHD's in infants and children, as well as some of the other factors to consider during the selection and the fitting process. I'll be showing some case study examples and then we'll have some time for question and answer at the end. Please hold all questions till the end of the course. So we are gonna just start off with the basics. As we all know, Bone Conduction Hearing Devices are known by many names, BAHA, Bone Anchored Hearing Aid, Bone Conduction Hearing Implant, Hearing Apparatus. I'm sure I can go on. The term Bone Conduction Hearing Device describes non

conventional amplification used for treatment of hearing loss through direct bone conduction. This type of device can be non surgical or surgically implanted. A BCHD should be recommended to individuals who are unable to use conventional air conduction amplification. So, to date, fitting protocols for BCHD's are not standardized, and often lack the detail that we desire as clinicians, leaving gaps in our clinical practice. At Children's Hospital of Philadelphia or CHOP, like many other institutions, we relied on creating our own fitting practices from our own clinical experience and trials using evidence based literature as a guide. The information discussed today will include manufacturer recommendations, clinical experience and practice guidelines from Children's Hospital of Philadelphia audiologists, as well as data from the Pediatric Bone Conduction Working Group. This group consists of pediatric audiologist throughout the U.S. and Canada of a special interest in bone conduction. This group was developed to collectively design and execute informative projects related to BCHD in children. In addition, we're developing fitting protocols that includes use of DSL prescription targets and verification with the skull simulator. We've already completed several projects.

In 2015, a survey was conducted of audiologist, BCHD selection and fitting practices and found that 16% indicated they did not feel confident in their fitting procedures. As a follow up to that survey in 2016, a multicenter retrospective chart review across six sites was completed. This found inconsistencies and lack of evidence based protocols for selection, verification and validation of BCHD's for children. At the end of 2018, 2019, two surveys were administered, one went to pediatric audiologists, and then another one went to parents to assess their perspectives about bone conduction technology, candidacy, counseling, recommendations, as well as their decision making for children with unilateral conductive or mixed hearing loss. And presently the group is developing guidelines for fitting non surgical BCHD's in children with conductive or mixed hearing loss. So we are first gonna start with candidacy. Just for review, the three main candidacy requirements for BCHD's are conductive hearing loss and mixed

hearing loss which can be in one and both ears, and single sided deafness or SSD, which is unilateral severe to profound sensorineural hearing loss. Manufacturers as well as most of the literature reports, candidacy for conductive and mixed hearing loss as using a pure tone average for bone conduction thresholds of 500, 1,000, 2,000 and 3,000 hertz up to about 65 dB HL. A PTA of less than 45 dB HL can typically be considered for most devices. For more of those mixed hearing losses of 55 to 65 dB HL, a power or suit power device is needed as the degree of the sensor or component affects the device choice. Having said that, even though the manufacturers have this candidacy criteria, additional considerations are that clinically, there's limited evidence that supports fitting those mixed hearing losses that are between 45 and 65 dB. You need to consider that benefit and the process or options may be limited as you would need to find sufficient gain for that sensorineural component. And incorporating 4,000 hertz into the PTA over 3,000 hertz is really a more clinical, feasible approach in pediatrics, as it can be obtained on ABR testing and it usually adds overall more clinical value than 3,000 hertz. And finally, the airborne gap should be greater than 30 dB as the greater the airborne gap, the greater the benefit.

To achieve most benefits of binaural hearing, bone conduction threshold should be symmetrical. Within 10 dB difference for the PTA, between the ears, or up to a 15 dB difference at those individual frequencies. There are several benefits for fitting BCHD's in both ears for those with bilateral conductive hearing loss with symmetrical bone conduction thresholds. Bilateral fitting gives audibility from both sides. It allows for localization and binaural summation, or that perceived feeling of loudness. It can improve your spatial awareness allowing overall better hearing and binaural processing, which leads to improved hearing in quiet as well as in noise. All of these things leads to higher patient satisfaction. Binaural processing is expected when one has symmetrical bone conduction thresholds. In cases of those greatly asymmetric threshold, it's less likely to obtain the same benefits of binaural hearing, such as localization and improved speech perception and noise. However, these patients still

may benefit from the reduction of the head shadow effect. Candidacy for unilateral conductive or mixed hearing loss is basically the same as what we went over for bilateral hearing loss. Additional criteria includes, that the air conduction PTA in the indicated ear should be greater than 40 dB HL and less than 20 dB HL in that normal hearing ear. And as previously discussed, the PTA airborne gap should be greater than 30 dB, with the most benefit actually perceived when the airborne gap is greater or equal to 50 dB. It's well documented that children with unilateral hearing loss may have reduced audibility, especially when sounds are from a distance. Their localization abilities as well as hearing in noisy environments are often decreased. As a result, children with unilateral hearing loss are at increased risk for language delays, educational challenges, auditory fatigue and social emotional difficulties. The survey that was conducted to pediatric audiologist administered by the Pediatric Bone Conduction Working Group found that a majority of respondents recommend a BCHD at diagnosis for children with the unilateral conductive or mixed hearing loss. However, their responses were highly variable when asked about their understanding of what the benefits were.

There are many reported benefits to fitting BCHD's in children with unilateral conductive or mixed hearing loss. It can provide binaural hearing and potentially avoid auditory deprivation. In addition, several studies have shown improved directionality localization, improved speech perception and noise and reports of improvement in both objective and subjective hearing. Single sided deafness or SSD is categorized as severe to profound sensorineural hearing loss in one ear with normal hearing in the other. Now, there continues to be a lack of consensus amongst pediatric audiologists, as well as limited evidence and outcomes data available that supports fitting of a BCHD in young children with SSD. CHOP's decision to add an age criteria was a direct result of our clinical experience, the existing evidence and the out of office patient trials that we conducted in house. In terms of age, we know that older children are able to monitor their listening environment and determine when the device can be beneficial or

when it can be detrimental. Previous publications on cross studies show that speech understanding in noisy listening environments was more difficult because the noise on the impaired side is routed along with speech to the normal hearing ear. This was reported by our patients who participated in our in house patient trials. They noted that hearing their friends in the cafeteria was more difficult than when they were in the library. Now, we do not solely rely on age, but more their maturity level. My six-year-old daughter would be able to complete all testing and give me tons of specific examples of how she determined if she heard better or worse. While my nine-year-old son would shrug his shoulders and say, I don't know if I heard any difference and it would be like pulling teeth getting something out of him. First and foremost, a parent needs to understand that BCHD will not provide binaural hearing for a child with SSD. All appropriate expectations should be reviewed as well the fact that there's still limited evidence and outcomes that supports fitting BCHD in young children with SSD. Choosing not to fit is not the same as doing nothing. At CHOP, we monitor their hearing and speech and language development.

We refer to early intervention, complete validation questionnaires during visits that can identify hearing concerns. When we do decide to fit, it's a collaborative decision between audiology, the family, and ENT. We do require an out of office amplification trial of at least four weeks with a device that can be utilized at home, school and social environments before making the final recommendation to obtain their own device. Let's take a look at another question from the survey. We asked the surveyed audiologist about device trials. Over 90% of the respondents reported that they offer an in office trial, while only 67% offered out of office trials. There are studies that show that a trial with a BCHD on a soft headband, may provide an indication of expected benefit. Out of office trial may be critical for families who deny hearing difficulties by highlighting the benefits of using a BCHD in different listening environments. Let's now talk about some at risk populations you may consider fitting a BCHD. Typically, when you think of bone conduction devices, it's most commonly recommended for those genetic

syndromes with ear anomalies such as atresia microtia. Those listed in that first column are common syndromes that have a high incidence of conductive or mixed hearing loss. Most children with abnormalities of the outer or middle ear are unable to use traditional air conduction amplification. Now, children with otitis media, chronic drainage and middle ear surgeries, for example, can all have fluctuating conductive hearing loss and can be at risk for hearing difficulties that can be just temporary or maybe more chronic. Often fitting of a BCHD may not come to mind for children with typical ear anatomy, or for those with just the common diagnosis of otitis media. As we all know that otitis media is extremely common in young children, and it's considered to be easily treated just by placing PE tubes. However, there may be a delay in medical and surgical intervention, and any hearing difficulties can affect a child's language development or educational learning. Examples of children that may fall into this category are babies with trisomia 21 who are risk for chronic otitis media, and are often unable to receive timely surgical intervention, as their ears are often too synoptic for PE tubes. Or children with tracheostomy and ventilator dependence, or those with unrepaired cleft palate. All have high incidence of chronic otitis media, but by placing tubes would only result in chronic ear drainage. BCHD should be considered for children who have fluctuating conductive hearing loss due to middle ear pathology.

Those who are waiting longer for medical intervention, or those who may have interment middle ear issues. An advantage of fitting a non surgical BCHD is that it does not need to be readjusted if the air conduction thresholds fluctuate. Programming an air conduction hearing aid for a patient with chronic otitis media and fluctuating hearing loss is like trying to hit a moving target. The biggest concern with pediatrics is not providing consistent and appropriate amplification. And with air conduction aids, you risk either over or under amplifying. You do not need to worry about any of this when you're dealing with bone conduction. Right, we're now going to move into evaluation and follow up. Determining candidacy, and evaluating a child for a Bone Conduction Hearing Device, starts with an audiologic assessment by obtaining frequency specific

thresholds. This could be done via ABR or with audiologic behavioral testing. Oftentimes, children start with a failed newborn hearing screen, followed by a diagnostic ABR, which is recommended for all children that are six months of age and younger, or for those children who cannot complete behavioral testing. Both bone conduction threshold as well as air conduction threshold should be obtained when applicable. In order to fit a device, it's recommended that minimally you obtain at least one low and one high frequency for bone conduction in that indicated ear. Common referrals that are made for all children with hearing loss are to an otolaryngologist to investigate the etiology of the hearing loss and to provide medical clearance to fit amplification. Too early intervention to determine support services. Speech language pathologists to evaluate and monitor their communication skills. This could be provided through early intervention, the school district or privately.

A genetics evaluation is important as oftentimes hearing loss can be related to additional medical issues. And an ophthalmologist as well for an evaluation. In addition to those five recommended referrals, at CHOP, we include family wellness services. All children diagnosed with hearing loss are referred to one of our child and family wellness therapists following that hearing loss diagnosis. The first appointment is typically within a few months after their diagnosis, to check in on how the family and the child are adjusting to their hearing loss. Future appointment goals are to help monitor and promote their child's healthy, social, emotional and behavioral development. During a BCHD evaluation, really like any other hearing aid evaluation, you're really discussing all types of amplification as well as remote microphones. This may include some information on non surgical as well as surgical options. Minimally, you should program the device by completing custom in situ measurements, well, I'll discuss further on the next slide. You may perform aided testing such as the Ling 6 and recorded speech perception testing using a test band or a soft headband, depending on their age. We're fortunate to have a bank of devices at CHOP that can be lent to patients for out of office trial. So the device can be trialed at home and at

school. An out of office trial is required for any patient that is considering surgery or for those with SSD. In situ audiometry, for the purpose of fitting a Bone Conduction Hearing Device is when pure tone audiometry is completed using the sound processor as the transducer rather than using the bone oscillator. The frequency specific stimuli is presented from the manufacturer programming software using the child's own BCHD and connection. This can be with a non surgical soft headband or with a surgical abutment. These measurements are incorporated into the software's prescription making the fitting more accurate because it eliminates the variability of the bone oscillator. In situ audiometry can be completed in a programming room or in any audiometric soundbooth by using a Noahlink Wireless or High Pro with cables that are patched through the wall panel. The top picture shows how I obtained in situ measurements in a programming room. I'm setting up the computer presenting the sounds and I have the child who's about two and a half facing away from the computer sitting at a small table, with another audiologist who is acting as the test assist.

During this particular session, we were first using VRA with a portable puppet that's positioned right to the left of the child. It's a little hard to see in that picture. We then introduced CPA when we realized he's two and a half and really not into puppets anymore, and we thought he might be ready for play. I typically use this type of setup in a programming room where the child faces away from the computer to ensure they're not seeing any presenting cues. Now this child has bilateral hearing loss, and I knew he would likely not hear the ambient noise around him. By completing in situ in a sound booth allows for VRA, CPA, and voluntary audiometry to be obtained in a quiet environment. The software can be installed through Noah or using the manufacturer's standalone software as shown in that bottom picture. We use standalone software in our booths. So I write down my thresholds on a piece of paper and manually add them into the Noah file when we go back into the programming room. The Noahlink Wireless is plugged into the computer on the tester side, and actually maintain good connection to the device that was located on the patient on their side of the booth. Moving on to

non surgical and surgical devices. Every pediatric audiologist has a wish list of what features are important for them to consider for children. These can include a tamper resistant battery door that meets industry standards. The device must be durable as we know how rough kids are in anything they own. Having a safety liner clip that attaches to the processor can help prevent the device from being lost. Disabling a volume control and programming button ensures that the sound is not changed accidentally. The indicator light provides some information about how the processor is working without having to ask the child. A parent or a teacher can tell from across the room if the device is on and working, or if the battery is getting low, or if the remote microphone is connected. Which brings me to remote microphones or other wireless accessories. All device being considered for children should have an option for remote microphone system for use in the classroom. And having wireless connectivity to phones and tablets are becoming more standard as well. Having a soft headband that's adjustable, secure and comfortable is very important. And finally, it's just an extra bonus if there are multiple color options to choose from.

For non surgical candidacy, we're primarily talking about a soft headband connection. This can be fitted any age. A soft headband is the best option for a child who is under five who is not yet a surgical candidate or for those at risk populations that we talked about earlier, who may only need a device for a short term for an acute hearing loss. A BCHD and on a soft headband can be worn in one or both ears. The manufacturer soft headband is recommended over a hard headband due to the adjustability and comfort that we'll discuss further on the next slide. So as I typically advise my families to be cautious when using some of the self made headbands that are not adjustable as these can often be difficult to ensure that they have a good fit. Studies have shown that the best position for a BCHD is when it's placed on the mastoid or right above on the temporal bone rather than placing it on the forehead. Soft headband should fit snug so that it's secure in position, but not so tight that you would not be able to fit one or two fingers in between the band and the head. Manufacturer soft headbands when properly

fit can provide sufficient and consistent tightness, so the output is not changed. When considering a BCHD for a baby with unilateral hearing loss, or for a bilateral fitting, adequate head control may be needed to ensure that you have appropriate placement of the processor to obtain the most benefit to the indicated cochlea. Now, I'm not suggesting that a BCHD should not be recommended a diagnosis. I'm rather conveying the importance of appropriate placement and acknowledging that this is often very difficult on a baby that cannot lift their head. There'll be another entire webinar that discusses surgical Bone Conduction Hearing Devices on June 16th. So I'm not going into a lot of detail about surgical options. But we're primarily talking about percutaneous, meaning there's an abutment that goes through the skin, or transcutaneous connection, meaning the processor is worn across the skin, typically held in place by a magnet. For candidacy, a child who is five or older without any medical contraindications can be considered a surgical BCHD candidate. They must have sufficient skull thickness, usually greater than 2.5 millimeters and sufficient temporal bone quality.

These are typically addressed by the anti surgeon. We make sure the family understands and able to maintain and clean the abutment site when applicable. And children with significant developmental delays or behavior problems that may jeopardize or harm that surgical site should be considered on a case by case basis. There have been many studies that show improved hearing with a surgical abutment over a non surgical connection. These include improvement in aided thresholds and speech perception testing, better sound quality and performance, an increase in learning speed and enhancing working memory. All of these benefits are a result of high frequency emphasis achieved with direct surgical transmission over non surgical connection since high frequencies are weakened by the passing through the skin. I've worked with many families over the years who may not be interested in pursuing any surgical options. The child may be doing well using their soft headband and they may not think that there will be any additional benefits of surgery. We looked at this as this

question from the audiologist survey inquired about potential concerns that led the patient or family not to proceed with the devices recommended. Remember, this is for children with unilateral conductive mixed hearing loss. Almost 60% of the responding audiologist reported that the reason the family did not proceed with the recommended non surgical device was due to cosmetic issues, cosmetic concerns. And then cosmetics as well as other concerns regarding the surgical procedure and maintenance of the surgical site were reported as additional reasons why they would chose not to proceed with surgery when recommended. Once a surgical solution is chosen, the process or fitting cannot take place until the soft tissue is healed and they're medically cleared by the ENT. In some cases, it may be beneficial to wait until the bone conduction implant is fully osseointegrated. Typically for a single stage surgery, both the implant and the abutment are placed in one surgery. The processor typically can be fit on the abutment about two to six months after surgery.

For a two stage surgery, the implant is placed first, followed by a second surgery to place the abutment, which could be about three to six months later after the osseointegration period. The processor can typically be fit on the abutment 2-3 weeks after that second surgery. These fitting times may vary slightly depending on the surgeon's preference. At CHOP, we recommend that the first follow up be scheduled about one month post fitting. Then every three months for the first year, every three to six months for the second year, and every six to 12 months thereafter. Additional visits may be needed if concerns arise or if limited information is obtained during a single visit. At least one validation tool should be selected for use during each appointment as deemed appropriate based on the patient's age, ability and their developmental level. Validation tools are divided into three categories, access to information such as device usage, aided speech perception testing, and outcome measures. Whenever possible, both aided speech perception and outcome measurements should be assessed for cross comparison and to provide the most comprehensive information possible. Use of recorded materials versus live voice are preferred in order to ensure a

standardized presentation of test items to increase reliability. At CHOP, depending on what the goals are of testing, the following conditions may be used. Speech at conversational levels in quiet presented at 60 dBA, speech at soft levels in quiet presented at 50 dBA, and conversational speech in noise at 65 dBA. A higher intensity level for completing testing and noise was selected to approximate the higher speech levels that are observed when one is in background noise. A plus five dBA signal to noise ratio was chosen as it is a typical classroom setting. Additional speech in noise testing paradigm is recommended for unilateral hearing loss. This test can first be completed unaided during the evaluation and then aided during follow up visit. Here's some examples of additional testing you might do with unilateral hearing loss, especially SSD. At CHOP, across our eight locations, we have different booth configurations. Some booths have speakers positioned at 45 degrees and some at 90 degrees.

The top two pictures show two speakers at 45 degrees on each side. In the first top left picture, we turn the patient so that they're facing one speaker at zero degrees azimuth and present speech at a conversational level of 65 dBA, while presenting a plus five multi talker babble from that second speaker at 45 degrees azimuth towards that left ear. In the top right picture, we then turn the patient to face the other direction. So speech again is from the front and then noise is being presented to the right ear. In the bottom left picture, speech is presented from the front at zero degrees azimuth and that plus five multi talker babble is presented from behind. The right bottom picture has speech and noise presented from that same speaker at zero degrees azimuth simultaneously, which can be used really for any speaker booth configuration. The speech and noise testing paradigm looks at the effects of noise as it crosses the normal cochlea. Now, this is not research by any means. It's not scientific, and it's not without flaws. As clinicians and not researchers, we created a clinical practical approach using speech information to address our clinical questions and needs. Comparing the unaided to the aided scores can assist with counseling of appropriate

expectations and further recommendations. Here are some examples of common outcome measures. LittleEars questionnaire is recommended for use for children birthed to two years of age. A peach for children, three years of age through school aged. And a sifter can give you useful insight on how the child is performing in school. This unilateral amplification questionnaire is another validation tool that we use at CHOP that was adapted and modified from a variety of other tools. It's a post amplification questionnaire that compares the current amplification to no amplification at all. The questions consists of listening situations which will be the most difficult for a child with unilateral hearing loss, including localization of a sound source. It looks at their confidence, frustration level and attention and it asks certain questions to elicit subjective feelings about the device. There are two versions, one for the parent and one for the child to complete. You would complete a new questionnaire following each device trial, which again just compares the current amplification that they're using to when they were no amplification at all.

It does not compare each device to one another. We find that this information on the questionnaire is helpful when counseling and for obtaining feedback. In addition to the questionnaire, we also developed a unilateral amplification journal. It's at least a four week daily journal that asks how many hours the device is worn in school and at home. It asks for specific situations that using the device made listening easier, and when listening was more difficult. Week four or whatever final week, ask some additional questions about the comfort of the device, the sound quality, ease of use and blatantly ask, would you use this device? Back in 2008 and 2009, CHOP audiologists were not routinely recommending BCHD's for any children with SSD. At that time, most of the current peer reviewed literature was based on adults that had acquired hearing loss, not children with congenital hearing loss. We did not want to conduct research. Rather, we wanted to answer our own clinical questions about whether or not to introduce new devices into our clinical practice without the evidence to support their benefit. So first, we decided to start with children who were 10 years of age or older, keeping in mind

the potential detrimental effects that were reported from cross studies. In 2009 and 2010, we fit about 12 children who completed a four week out of office trial using different devices including BCHD's on a soft headband. We utilized the unilateral speech perception testing paradigm, as well as the unilateral amplification questionnaires and journals throughout the trial. Some children liked the device and decided to get their own and proceed with surgical solutions. The majority reported there were some situations that they did not hear well, such as the cafeteria, restaurants, the mall and gym class. This information both guided and validated our candidacy criteria, demonstrating that children needed to be old enough to have the maturity to determine when the device would be beneficial or when it would be detrimental. Throughout the last 10 years, we've lowered the suggested age from 10 to eight and presently have it at school age with a focus more on maturity level and the ability to be a reliable reporter.

All of these tools helped look at both objective and subjective benefit to help make a decision on how to proceed with amplification. We're gonna be moving on to case studies. These case studies highlight bone conduction devices over the years and you'll actually see how each case shaped my growth as a clinician. Let's first meet Bianca. She's a patient I've been following for 15 years. So think back to the year 2005. Bianca has Treacher Collins Syndrome, bilateral microtia and permanent bilateral conductive hearing loss with additional hearing fluctuations complicated by middle ear pathology. Treacher Collins is a genetic syndrome categorized by distinctive abnormalities of the head and face. This condition mostly affects the development of bones and other tissues of the face. Typically they have downward slanting eyes, a notch lower eyelid, underdeveloped or malformed ears, including the external and middle ear structures and hearing loss, which is primarily conductive. I first met Bianca at three weeks of age when she was scheduled for a natural sleep ABR as a follow up to her failed newborn hearing screen. At that visit, I noticed she had atypical facial features with bilaterally malformed ears. She was not yet given a genetic diagnosis.

ABR testing revealed a moderately severe conductive hearing loss in both ears with abnormal temp anagrams. Bianca was referred to ENT and scheduled for follow up testing in a few weeks. Between two and six months of age, amplification was recommended, but medical management delayed the fitting as Bianca was diagnosed with otitis media. At three months of age, a PE tube was placed in her left ear only, as her right ear canal was twistomatic for any surgical intervention. ABR results that followed revealed a moderately severe hearing loss in the right ear and a moderate to mild hearing loss in the left ear. When discussing amplification, the benefits of bone conduction devices versus air conduction were reviewed. During this time, Bianca was given an official Treacher Collins diagnosis. And even though her facial features suggested a genetic syndrome, her parents were definitely having difficulty coping. A BCHD and a soft headband was recommended, however, her parents were reluctant to proceed as they expressed cosmetic concerns at the soft headband.

I hesitantly agreed to trial an air conduction hearing aid in the left ear knowing that it would be challenging giving her extremely synoptic ear canals and cupped penas. Now for the next six months just as predicted, we were pretty much losing the retention battle. We tried everything. Huggy aids, double sided tape, hats, anything and everything that was available to keep that in her ear and keep that hearing aid behind her ear. A BCHD was again recommended to ensure that Bianca had consistent access to sound 'cause I knew this was not being achieved. With an air conduction aid, that would not stay in her ear. But her parents saw a benefit and did not wanna pursue any other options at that time. Between one and two years of age, some speech and language concerns were reported. And since Bianca was already receiving early intervention support services, I recommended that we try binaural amplification, so a BCHD was fit in her right ear at two years of age. For the next several years, Bianca utilized a BCHD and soft headband for the right ear and an air conduction hearing aid in the left ear. Here's an example of some additional testing completed during a visit when Bianca was three. She was successfully using binaural amplification

as seen by all of her verification and validation measures. The next few years were generally uneventful. At around four or five years of age, we started to discuss surgical options. The family did not want to consider percutaneous bone conduction surgery at that time as this was the only option available. When other people transcutaneous surgical options emerged, being that benefit would be comparable to the results with her soft headband, the family decided to just continue using her BCHD without surgery. Bianca did continue to use both devices during all waking hours. Next, I want to introduce Teddy. Teddy is only a few years younger than Bianca. Teddy has Goldenhar Syndrome, unilateral atresia, conductive hearing loss, chronic otitis media, and respiratory failure with tracheostomy due to an obstructive airway issue as an infant. Goldenhar Syndrome is a cranial facial syndrome that includes hemifacial microsomia. The eyes, ear and or spine may be affected. The most common ear abnormalities include atresia and or microtia as well as hearing loss.

Teddy was first seen in audiology during his inpatient stay at CHOP as a follow up to his failed newborn hearing screen. Teddy was diagnosed with Goldenhar Syndrome and had a normal appearing left ear with complete atresia of the right ear. We completed several natural sleep ABR's during his hospital stay before he was diagnosed with a severe rising to mild hearing loss in the left ear. At five months of age, Teddy was fit with an air conduction hearing into the left ear as conclusive bone conduction responses could not be obtained for the right ear. For the next several months, we battled chronic otitis media in his left ear, which is often seen in children who have tracheostomies and are ventilator dependent. A PE tube was first placed at eight months of age in the left ear. Now, behavior results could not be obtained as Teddy was completely petrified of the VRA puppets. He would cry and scream every single time we stepped into the sound booth. After numerous attempts, he was referred for another ABR which was completed at 11 months. Results revealed a moderate rising to mild hearing loss of the left ear. Bone conduction results for the right ear were essentially normal. So since there were no reported hearing concerns, I

recommended that Teddy continue use of his air conduction hearing aid in the left ear. Now, you might be thinking, why didn't I recommend a BCHD for the right ear? At that time, CHOP was not routinely recommending BCHD's for children with unilateral hearing loss due to lack of evidence to support its use. And I was not concerned that his hearing in the left ear would fluctuate as long his PE tube was in place. Even though Teddy had bilateral hearing loss, at that time, my recommendations were similar to what I would give for someone with unilateral hearing loss as he was successfully using his air conduction hearing aid. I'm about to show you why I regretted that decision. From one to two years of age, chronic otitis media affected his ability to consistently wear his hearing aid. He had his PE tube replaced two more times during the year, and he still was unable to complete any behavioral testing. Still continue to scream and cry whenever we even got near the sound booth.

So since I wasn't able to monitor his hearing, I had concerns about the possibility of over or under amplifying, as well as the fact that he was getting inconsistent access to sound just like Bianca. So we decided to trial a BCHD and a soft band for the right ear while using his air conduction hearing in the left ear. This ensured that he had consistent access to the sound at all times. And finally around three, we were able to obtain behavior results using play audiometry, which was in a small booth that did not have any VRA puppets. These audiograms highlight some of the fluctuations in hearing for air conduction to chronic middle ear issues, even after four different PE tube surgeries. You're wondering about lack of aided test results obtained. Remember, Teddy still would not step foot into any booth that had VRA puppets, even if they were covered. And all of our booths with speakers were equipped with them. He was so happy when he finally grew out of that phase and he cannot believe when I tell him stories about how he was when he was younger. So Teddy's outcome, he was getting good benefit from his BCHD soft headband. We decided to discontinue use of his air conduction hearing aid in the left ear following improved hearing as shown on that most recent audiogram. BCHD for the left ear was considered and trialed. However, a

lot of benefit was not reported. Most likely it was due to the improvement in air conduction thresholds, leaving not much of an air grown gap as his higher frequencies were pretty much around the normal range. And as low as were only about 30 dB HL. When it was time for Teddy to consider surgical options, unlike Bianca who was bilaterally fed, when they decided not to proceed with her surgery, I knew she would still have access to those higher frequency sounds from our air conduction hearing aid. Now, Teddy needed the best hearing outcome being that he was fit unilaterally. So he proceeded with a percutaneous solution which was completed at five years of age. Now finally, let's talk about Ben. I first met Ben as a follow up to his newborn hearing screen just a few years ago. He has trisomy 21, fluctuating conductive hearing loss in both ears, and chronic otitis media. Children with trisomy 21 have a higher risk of hearing loss that could be conductive, mixed or sensorineural. In addition, they often have other factors, which put them at risk for progressive hearing loss. And majority of children with trisomy 21 will have otitis media.

I first saw Ben during a natural sleep ABR as a follow up to his failed newborn hearing screen just in one ear. The ABR revealed a mild conductive hearing loss for the right ear and a moderate conductive hearing loss in the left ear. He was referred to ENT for an audiologic consult. From three to six months, Ben had two additional ABR's which showed some degree of conductive hearing loss. ENT did recommend PE tubes, however, his ear canals were twistonotic. This time I did not hesitate to recommend amplification and from my previous experience with otitis media and fluctuating conductive hearing loss, a BCHD is the most appropriate option. Initially, only one BCHD was recommended as Ben had poor head control and trunk support and appropriate placement of two devices would have been difficult. Around one, he was fit with his second bone conduction device using a bilateral soft headband. His hearing and speech development was closely monitored. He was enrolled in early intervention and was receiving speech, physical, occupational therapies, as well as hearing support services. Due to some medical issues, Ben did not undergo PE tube placement until he

was about three years of age. Since Ben seemed to say about his terrible two's for when he turned three, ear specific behavioral information could not be obtained. Therefore sedated ABR testing was recommended which revealed essentially normal hearing in both ears. It was recommended that he discontinue use of amplification at that time, but have close audiologic monitoring. As Ben got older, he still had middle ear issues. His ears would sometimes drain or the tubes would get blocked. Over the next several years, Ben continued to have fluctuating conductive hearing loss depending on the status of his PE tubes. His parents were instructed to use his BCHD as needed. So as time went on, as Ben got older, he would actually ask to use his BCHD if he was not hearing well. Since there was no risk of over amplifying just like there would be with air conduction hearing aids, Ben did not need to come in for a visit every single time he battles an episode of otitis media. Now let's look again at Bianca. Remember, I started seeing her in 2005.

Looking back, my biggest regret is not addressing the ill feelings about bone conduction devices and waiting to fit amplification in one ear. Maybe I didn't discourage trialing the air conduction hearing aid because I thought the sound quality and the frequency response would be better and maybe I convinced her parents of that too. I also didn't recommend bilateral BCHD's. Why, you might ask? Well, again, this was 15 years ago and during that time period, there was not a lot of evidence that supported fitting of BCHD's in both ears. When speech concerns were reported around two, I did not hesitate to revisit the BCHD recommendation for other ear. Bianca became my first bimodal patient to use both an air conduction hearing aid and a bone conduction hearing device. I've since used the experience and lessons learned from Bianca and applied it for future use. Now, let's look back on Teddy. Remember those at risk populations we discussed earlier, Teddy is quite a few of those. He has Goldenhar Syndrome, had an inpatient stay of several months and had a tracheostomy with ventilator dependence, which caused him to be more at risk for chronic otitis media. Looking back, I focused on amplifying his normal appearing ear with an air

conduction aid just like I did with Bianca. Often fitting a BCHD may not be come to mind for children with typical ear anatomy or for those with a common diagnosis of otitis media. For Bianca and Teddy, even though they had a diagnosed cranial facial syndromes, they had chronic otitis media, which we all know just complicates everything. For Ben, I did not hesitate to recommend a BCHD in both ears early on. I knew his issues with otitis media would be more like a war dragged out over many years and a quick battle. Providing consistent access to sound early was a priority. Now, those of you who've been practicing for as long as I have, or even longer, know how much bone conduction has changed over the years, from the old Starkey BC1 air conduction converted to bone aids using metal headbands, to the scientific compacts that required a screwdriver for programming.

BCHD's have better frequency responses than ever before, which can make you less fearful of recommending one over an air conduction aid. As for fitting one or both ears, there's now more than enough research that supports fitting BCHD's in both ears for bilateral hearing loss and for fitting BCHD for unilateral conductive mixed hearing loss. It goes without saying that additional research is needed on young children with SSD. I've learned that the fitting of transcranial amplification appears to be a good option for some children with SSD in some listening conditions, and one child's outcome does not predict the outcome of another. We need to remember that our clinical decisions, judgment and guidelines evolve as new technology or evidence arises. So thank you for joining me today and thank you to Oticon Medical for inviting me. I hope this information today was exactly what you were looking for. And now for the remainder of these few minutes, I'm available to answer any questions. So I answered everything perfectly.

- [Ravi] Thank you, Laurie. I think that was a great class, that was a great session. It was very, very enlightening, at least for me because you took us through the whole process of selection, treatment outcome measures that we'll gimmick. So that was

wonderful. So if any of you have further questions, please feel free to email Laurie. She has the email address there. And she would be most happy to answer all of your questions. Thank you again everyone for attending our very first session in the webinar series. Again, it was a great class, and I look forward to seeing you in the coming weeks for all the other sessions. Thank you.

- [Laurie] So we have a couple of questions coming in. Some people mentioned that they're happy that I brought up issues with children with trisomy 21 'cause everyone knows that it ends up being a long battle. There's a question about, do most kids wear their headband all day long? I find that they do. I think that most of them put it on as soon as they wake up in the morning and then take it off when they're getting ready to go to bed. I haven't had a ton of patients complain about comfort with those soft headbands, the manufacturer soft headbands.

- [Ravi] I think those are 30 questions.

- [Laurie] Yeah.

- [Ravi] Wonderful.

- [Laurie] Great, thank you.

- [Ravi] Thank you.

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