

Phonak Compendium

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A Review of Unilateral Hearing Loss in Children

Introduction

The first mention of hearing loss in recorded history came from the ancient Egyptians, who in 1550 B.C. treated a “bewitched” ear by pouring a mixture of tree oils and medicinal clay into the affected ear (Mudry, 2006). Fortunately, our understanding of hearing loss has come a long way since then. Today, people with hearing loss have access to advanced amplification technologies, strong social support networks, and an array of protective legislation. As the detrimental impact of bilateral hearing loss on children is so well-established, federally-mandated protocols exist to identify, treat, and support those children diagnosed with hearing loss. Unfortunately, it has taken far longer to understand the impact of permanent unilateral hearing loss in the pediatric population.

Defined by the presence of normal hearing in one ear, and impaired hearing in the other ear, unilateral hearing loss (UHL) can be caused by a hereditary condition or acquired as the result of an infection, injury, or disease. The resulting hearing loss can be sensorineural, conductive, or mixed in nature, and range in severity from mild to profound. In many ways, the presence of normal hearing in one ear has complicated our ability to detect and treat children with UHL. Decades ago, it was thought one normal hearing ear was enough to get the child through school, but in the 1980s

researchers began to show the struggles these children experienced academically. Studies showed significantly higher rates of academic failure and grade repetition amongst these children in comparison to their normal hearing peers. These studies showed these children faced far greater challenges than previously thought, and resulted in further research to understand which treatments worked best for this unique population. Recently, research has increasingly focused on the neurological impact of UHL in children, offering fascinating insight into how structural changes in the brain correlate to academic and social difficulties often seen in these children.

Never before have clinicians had such a vast array of information available regarding UHL in the pediatric population. This paper is intended to summarize recent research into pediatric UHL and the unique challenges children face with this condition. The hope is that clinicians can use this information to make the appropriate clinical decisions for their patients, educate teachers and families regarding the needs of the child with UHL. Since each child with UHL has unique communication needs, it is paramount that audiologists work with these children and their families to find the most appropriate combination of amplification, intervention services, and follow-up monitoring.

Diagnosis, Prevalence, & Etiology

UHL can present with a number of different audiometric configurations based on the type and severity of hearing loss, as well as the side of impairment. Because children with UHL are less likely to demonstrate verbal delays associated with hearing loss, they were often not diagnosed until school-age, when academic and behavioral difficulties suggested a problem. Such a late diagnosis has made determining etiology difficult, as it was unknown whether the child was born with the hearing loss or it was acquired later as a result of trauma or illness. Determining the etiology of UHL is important, as the child may be at risk of progression or involvement of the contralateral ear resulting in later bilateral hearing loss.

With the widespread implementation of universal newborn hearing screenings [UNHS], children with unilateral hearing loss are able to be diagnosed earlier than ever before. Not only does this allow for intervention at an earlier age, but also improves our ability to identify etiology. This information can then be used to determine the appropriate follow-up protocol for these children. Questions still remain however, as to whether current UNHS protocols are sensitive enough to children with milder degrees of unilateral hearing loss. Data collected from individual US states show rates of diagnosis have yet to match prevalence data from multiple research studies. Further research is needed to determine the best way to screen children with unilateral hearing loss.

Ross, D.S., Holstrum, J., Gaffney, M., Green, D., Oyler, R.F., & Gravel, J.S. (2008). Hearing screening and diagnostic evaluation of children with unilateral and mild bilateral hearing loss. *Trends in Amplification, 12*(1), 27 – 34.

In June 2005, the Centers for Disease Control and Prevention and the Marion Downs Hearing Center convened a workshop to discuss current challenges faced in the diagnosis of children with "minimal hearing loss," including UHL and mild bilateral hearing loss. Following the introduction of UNHS in hospitals across the United States, data obtained from programs appeared to show a lower rate of diagnosis for UHL than was expected based on existing literature. A number of contributing factors are established, including an inability of existing protocols to detect losses less than 30 dB HL, a lack of reporting on results of hearing tests post-referral, and voluntary reporting from audiologists and physicians means some children with UHL may go unreported. While the authors state more children with minimal hearing loss could be diagnosed if another mandatory screening event were to be implemented at around 30 months of age, there is no widely recognized life event shared by children at that age (such as beginning of school or a mandatory physical examination).

Lundeen, C. (1991). Prevalence of hearing impairment among school children. *Language, Speech, and Hearing Services in Schools, 22*(1), 269 – 271.

In 1968-1969, the United States conducted the National Speech and Hearing Survey, obtaining reliable audiometric thresholds for 38,568 students in grades 1 through 12. Measurements were taken in school districts across 48 states, and this undertaking provided comprehensive hearing data, the scope of which had never before been attempted. Results showed the percentage of children with UHL to be 1.9%, which outpaced the number of students with bilateral hearing loss (0.73%). A trend noted in the data was that hearing loss of any kind was most common in first graders and prevalence of hearing loss seemed to decline with age.

Oyler, R.F., Oyler, A.L., & Matkin, N.D. (1988). Unilateral hearing loss: Demographics and educational impact. *Language, Speech, and Hearing Services in Schools, 19*(1), 201 – 210.

After reviewing seven years of audiologic records from a school district of 54,000 students in kindergarten through 12th grade, 106 students with UHL were identified (prevalence = 2/1000). Of this group, there were twice as many cases of right-ear UHL than there were left-ear UHL and there were slightly more boys with UHL than girls. Three-fourths of the hearing losses were sensorineural and any losses related to otitis media were not included in this population. Of the remaining children with conductive or mixed UHL, nine children had atresia.

In addition to a review of audiograms, the teachers of these children were surveyed to determine their academic performance. Results showed children with UHL were significantly more likely to have failed a grade than their normal hearing peers. Children with right-ear UHL were significantly more likely to have failed a grade than children with left-ear UHL and children with a severe-to-profound UHL were more likely to fail a grade than children with a lesser degree of UHL. Only 40% of children with UHL were receiving services. When teachers rated the overall academic performance of the children with UHL, there was no difference from normal hearing peers, with 50.9% being rated as having "average" academic performance. This is fascinating because these children with UHL have much higher rates of remission and access to special services, yet the teachers seemed to think they were performing adequately academically.

Ruscetta, M.N. & Arjmand, E.M. (2003). Unilateral hearing impairment in children: age of diagnosis.

Retrieved from

<http://www.audiologyonline.com/articles/unilateral-hearing-impairment-in-children-1123>

In a chart review of 77 children with sensorineural UHL (SNUHL) not caused by trauma or infection, the average age at diagnosis was 4 years, 11 months. The average age at diagnosis for a child with a mild degree of SNUHL (6 years, 9 months) was higher than those children with a moderate to profound degree of SNUHL (4 years, 3 months). Most children diagnosed with SNUHL were identified through school or pediatrician-led hearing screenings (54.5%), parental concern (22%), or (pre-universal) newborn hearing screening protocols (10%). Most of the children with SNUHL (55.5%) had an unknown etiology while 26% were a result of anatomical anomalies of the inner ear. The authors suggest children with a milder degree of SNUHL are at greater risk for late diagnosis as they may be less impacted communicatively than children with a greater degree of impairment.

Ghogomu, N., Umanski, A.M., & Lieu, J.E. (2014). Epidemiology of unilateral sensorineural hearing loss with universal newborn hearing screening. *The Laryngoscope*, 124(1), 295 – 300.

After implementation of a Universal Newborn Hearing Screening (UNHS) protocol in the state of Missouri, the average age of diagnosis for UHL decreased to 2.6 years of age (from 4.4 years of age). The percentage of children with UHL diagnosed by 6 months of age went from 3% to 42%. As a result of earlier detection and diagnosis, more instances of UHL could be diagnosed as having a congenital etiology than before the implementation of UNHS.

Fitzpatrick, E.M., Al-Essa, R.S., Whittingham, J., & Fitzpatrick, J. (2017), Characteristics of children with unilateral hearing loss. *International Journal of Audiology*, 1 – 10. doi: 10.1080/14992027.2017.1337938

A comprehensive, prospective chart study of children with UHL at a pediatric hospital in Ontario, Canada spanning 13 years found 537 children diagnosed with permanent hearing loss and 108 (20.1%) of those were diagnosed with permanent UHL. The median age at first assessment was 4 months, and the median age at diagnosis was 13.9 months. While those with congenital UHL had a median age of detection of 2.8 months, children with early onset UHL (before 6 months of age) had a median age of diagnosis of 4.3 months. Most cases of UHL were sensorineural (64.8%), while 25% were conductive losses and the remaining 10% had a mixed origin. The etiology of the hearing loss was known in 55.6% of all UHL cases, with a malformation of the ear accounting for a majority (51.7%) of these cases.

Of the 108 children with UHL comprising this study, 92 had enough audiometric data points to analyze progression of their hearing loss over several years. Progressive losses were identified in 39 of the children (42.4%), with most experiencing a progression of hearing loss in the impaired ear. The authors demonstrate the importance of diagnostic imaging and monitoring to determine the etiology of loss as well as monitor for progression of loss in either ear. Once again, UNHS was proven to significantly lower the average age of diagnosis for children with UHL.

Friedman, A.B., Guillory, R., Ramakrishnaiah, R.H., Frank, R., Gluth, M.B., Richter, G.T., & Dornhoffer, J.L. (2013). Risk analysis of unilateral severe-to-profound sensorineural hearing loss in children. *International Journal of Pediatric Otorhinolaryngology*, 77(7), 1128 – 1131.

A retrospective chart review of 84 pediatric patients with severe-to-profound sensorineural UHL found an identifiable etiology in 41.5% of patients. The most common risk factors were perinatal events (16.2%) and a family history of hearing loss (10%). Of the 49 patients who had undergone MRI or CT scans, 20 (40.8%) were found to have unilateral or bilateral temporal bone anomalies, with the most common being enlarged vestibular aqueduct. With a mean-follow-up time of 31.9 months, no patient in this study demonstrated a clinically significant change of audiometric thresholds in the contralateral ear (change in contralateral PTA >10 dB). The authors state the risk of contralateral involvement with severe-to-profound UHL is low; however, this may be a result of selection bias. Since the patients included in this study were undergoing evaluation for a bone-anchored hearing aid (BAHA), the authors state these children were likely to have more stable thresholds given their age and hearing status and may not accurately represent the risk of progression in younger children with milder degrees of hearing loss.

Dodson, K.M., Georgolios, A., Barr, N., Nguyen, B., Sismanis, A., Arnos, K.S., ... Pandya, A. (2012). Etiology of unilateral hearing loss in a national hereditary deafness repository. *American Journal of Otolaryngology*, 33(1), 590 – 594.

Samples of DNA from 34 adults and children with permanent UHL were analyzed to determine genetic characteristics of those with UHL. The average age at first diagnosis for the subjects in this study was 7 years. Of the 34 subjects, 12 (35%) showed variations in gene sequences typically associated with hearing loss, indicating possible genetic involvement. However, the presence of these variations alone would not necessarily present with hearing loss. The authors suggest combinations of genetic sequences or the interplay of genes and environmental factors may play a role in certain UHL etiologies.

Uwiera, T.C., deAlarcon, A, Meinzen-Derr, J., Cohen, A.P., Rasmussen, B., Shott, G., & Greinwald, J. (2009). Hearing loss progression and contralateral involvement in children with unilateral sensorineural hearing loss. *Annals of Otolaryngology, Rhinology, & Laryngology*, 118(11), 781 – 785.

In a retrospective review of 198 children with permanent congenital SNUHL, temporal bone CT scans revealed 13% of these children exhibited temporal bone anomalies. The children with temporal bone anomalies were more likely to have a profound degree of hearing loss than the children without anomalies. Out of 142 children with sufficient follow-up data, 30 children exhibited a progressive loss in the impaired ear, and over half of these children had hearing losses which progressed at all measured frequencies. The median progression rate was 9 dB per year. An additional 15 children developed new-onset hearing loss in the normal hearing ear. The majority of these new losses developed in the high-frequencies only.

New-onset hearing loss in the contralateral ear developed more frequently in children with a high-frequency UHL (31%), opposed to those with a hearing loss across a wider range of frequencies (10.6%). The authors strongly recommend children with UHL be closely monitored for changes in audiometric thresholds in either ear. This line of research suggests some cases of sensorineural UHL are actually a subclinical presentation of binaural hearing loss.

Lieu, J.E.C. & Dewan, K. (2010). Assessment of self-selection bias in a pediatric unilateral hearing loss study. *Journal of Otolaryngology, Head & Neck Surgery*, 142(3), 427 – 433.

The goal of research involving a sample of subjects is that these subjects represent a random cross-section of the entire population, allowing the results of the research to be generalized to the larger population. This requires recognizing the impact of "self-selection bias," or the problem of people who choose to participate in research not accurately reflecting a randomized sample. In a study of children with UHL, key differences were found between the children participating in the study sample and the children who elected not to participate. Children included in the study were more likely to have an unknown etiology, have fewer siblings, have a later age of diagnosis, and live closer to the research facility. Participating children were also more likely to have a sensorineural or mixed component to their hearing loss and less likely to have a structural hearing loss component (e.g. microtia, atresia, etc.). To combat the problem of self-selection bias, the authors suggest facilitating the logistical needs of the parents and their children including transportation vouchers and flexible scheduling policies. Once recruited, it is important to work with families

as much as possible to ensure they do not drop out and skew the sample population demographics. Recognizing characteristics of self-selection bias can help determine how generalizable the results are to the population at large.

A Summary of Diagnosis, Prevalence, & Etiology

Children with unilateral hearing loss comprise a significant portion of the population of children with hearing loss. While UHL can have a variety of presentations and etiologies, most cases seem to be sensorineural in nature. This can be the result of genetic and/or environmental factors or the result of illness or trauma. The introduction of UNHS has allowed audiologists to diagnose UHL at a much earlier age than before while allowing for greater ability to determine etiology. Some etiologies may require specific follow-up testing to determine the presence of co-morbidities. As there is great variability in research outcomes, all children with UHL should be regularly monitored for changes in audiometric thresholds and involvement of the contralateral ear. More research is necessary to determine how UNHS protocols might be altered to better find children with UHL without overburdening the system with false-positive referrals.

UHL & the Inner Ear

In the fourth week of embryonic development, a primitive sensory cell called the "otic placode" forms on each side of the embryo. These placodes gradually ingress into the embryo and form the otic cysts, the precursor to the cochlear and vestibular structures. It takes a further sixteen weeks before the inner ear is fully developed. During that time a disruption in the development process can result in a congenital hearing loss.

For children with UHL, looks can be deceiving. The presence of normal hearing in one ear may mask the presence of an underlying bilateral condition. Imaging studies can help explain the presence of such a condition, and recent studies have given insight into the prevalence of inner ear anomalies in these children. Since the inner ear encompasses not only the hearing mechanism, but also the vestibular system, inner ear anomalies can sometimes be expressed as difficulties with balance or coordination.

Masuda, S., Usui, S., & Matsunaga, T. (2013). High prevalence of inner-ear and/or internal auditory canal malformations in children with unilateral sensorineural hearing loss. *International Journal of Pediatric Otorhinolaryngology*, 77(2), 228 – 232.

A retrospective review of the medical charts and temporal bone CT scans of 69 pediatric patients with permanent sensorineural UHL revealed 66.7% of patients exhibited some degree of inner ear malformation. When parsed out by age, infants under the age of one year had significantly higher prevalence of inner ear malformations at 84.6%. The most common inner ear malformation was stenosis of the cochlear nerve canal [CNC]. The authors postulate that the cause of CNC stenosis could be cochlear nerve hypoplasia. This is based on the hypothesis that a prerequisite for a normatively-sized cochlear nerve canal is a normatively-sized cochlear nerve. Temporal bone CT imaging is recommended for all children with permanent sensorineural UHL. It is further suggested that children with specific inner ear malformations be referred for genetic testing to determine etiology.

Haffey, T., Fowler, N., & Anne, S. (2013). Evaluation of unilateral sensorineural hearing loss in the pediatric patient. *International Journal of Pediatric Otorhinolaryngology*, 77(6), 955 – 958.

In a retrospective chart review of 89 pediatric patients with SNUHL over a 6-year period, it was discovered that one third of those patients who were given a CT scan of the temporal bone (CTTB) showed structural anomalies (e.g. EVA, Mondini Dysplasia, etc.). A post-CTTB MRI revealed no additional anomalies in these patients. When CTTB showed no anomalies, a post-CTTB MRI revealed structural anomalies in approximately 10% of patients. Of the 14 patients who underwent genetic testing, a genetic etiology was discovered in 43%. Referrals for ophthalmologic services showed 29% of patients required intervention. CTTB is a worthwhile diagnostic tool for pediatric patients with UHL. For those patients with an unremarkable CTTB, MRI can be utilized to further rule out physical anomalies. Patients with increased risk factors for genetic or acquired hearing loss should be referred for genetic testing or ophthalmologic evaluation as appropriate. Children with sensorineural UHL should be monitored for changes in audiometric thresholds, especially in the normal-hearing ear, as 11% of patients eventually progressed to a bilateral hearing loss.

Birdane, L., İncesulu, A., Özüdoğru, E., Cingi, C., Caklı, H., Gürbüz, M.K., & Adapınar, B. (2016). Evaluation of the vestibular system and etiology in children with unilateral sensorineural hearing loss. *The Journal of International Advanced Otolaryngology*, 12(2), 161 – 165.

In a study of thirty-three children with permanent sensorineural UHL, 52% of patients presented with additional inner ear abnormalities, with the majority consisting of lateral semicircular canal hypoplasia. Of the 33 subjects with UHL, 24 reported some degree of vestibular symptoms including vertigo, vestibular migraines, nausea, and/or imbalance. Electronystagmography was completed on 31 patients revealing 67.7% had canal paresis on the side of the impaired ear. VEMP testing revealed the children with UHL had prolonged N23 latencies in comparison to a normal hearing control group. The authors underscore the importance of diagnostic imaging (MRI or CT scan) for children with UHL, both for identifying the etiology of the hearing loss as well as monitoring for other inner ear abnormalities present in this population that may impact hearing or balance.

Wolter, N.E., Cushing, S.L., Vilchez Madrigal, L.D., James, A.L., Campos, J., Papsin, B.C., Gordon, K.A. (2016). Unilateral hearing loss is associated with impaired balance in children: a pilot study. *Otology & Neurology*, 37(1), 1589 – 1595.

Children with UHL demonstrate significantly poorer balance ability than children with normal hearing. Results of time to fall testing showed that these children performed poorer than normal hearing controls only on the most challenging of tasks (for example, standing on one foot on a stable surface with eyes closed) and as a result clinical testing may not be challenging enough to reveal differences. Balance testing performed with and without visual stimuli showed the children with UHL relied more heavily on visual inputs to maintain balance than the children with normal hearing. The authors suggest this could be a result of monaural auditory input or concurrent vestibular disorders present in children with UHL.

A Summary of UHL & the Inner Ear

Children with sensorineural UHL are at increased risk for physiologic anomalies of the inner ear, including the unimpaired ear. A diagnostic CT scan of the temporal bone can show whether the child with UHL exhibits these inner ear abnormalities, and if they do not, an MRI can be used to further explore potential abnormalities. In conjunction with diagnostic imaging, balance testing can be performed to determine if the child has concomitant vestibular dysfunction.

The test battery may need to be more rigorous for these children and include tasks with eyes closed, as visual compensation means problems may only surface during extremely difficult balance tasks.

UHL & the Brain

The brain is the true organ of hearing, receiving auditory input collected by the ears and translating the incoming signal into meaningful information that we use to communicate. This process of audition involves electrical impulses travelling a complex pathway of ipsilateral and contralateral fiber tracts connecting the auditory nerve to the primary auditory cortex of the brain. In normal hearing children, fMRI studies have shown significant asymmetry in cortical activation in response to sound. This is thought to be a result of the left brain hemisphere being the primary site for language and auditory processing. Because the contralateral pathways are more robust than the ipsilateral pathways, input from the right ear crosses the ipsilateral pathway directly to the auditory cortex and is dominant over the signal from the left ear.

While the cochlea is fully formed at 30 weeks gestation, the auditory pathways are not fully developed until the child is between the ages of four and eight years old. Changes in non-auditory neural structures will have some impact on the development of the auditory system during this time. In children with bilateral hearing loss, it is well-established that reduced stimulation of auditory centers of the brain during the early years of life can result in maladaptive reorganization of auditory neural pathways. However, it was unknown what impact monaural input would have on these same neural structures.

Propst, E.J., Greinwald, J.H., & Schmithorst, V. (2010). Neuroanatomic differences in children with unilateral sensorineural hearing loss detected using functional magnetic resonance imaging. *Archives of Otolaryngology, Head & Neck Surgery, 136*(1), 22 – 26.

Using fMRI, changes in neural bloodflow can be monitored to determine what areas of the brain are activated in response to specific stimuli. For this study, 12 children with severe-to-profound UHL and 23 normal-hearing children were stimulated with narrowband noise chirps and speech presented in noise while undergoing an fMRI scan. Activation patterns in the children with UHL differed significantly from those of the normal hearing control group. In response to the narrowband chirps, both normal-hearing and UHL groups showed bilateral activation to the monaural stimuli. However, the children with UHL had reduced activation of the auditory

centers of the brain, failed to activate secondary auditory centers associated with more advanced sound integration, and showed no activation of auditory attention networks. For the speech-in-noise stimuli, children with UHL showed activation of the secondary auditory processing areas on the left side only, while the normal hearing controls showed bilateral activation.

The speech-in-noise task showed interesting differences in activation related to the side of impairment. Children with right-side UHL showed no activation of the auditory attention centers, while the normal hearing children and the children with left-side UHL showed typical activation. Interestingly these auditory attention centers are in the right hemisphere of the brain. This suggests contralateral stimulation of the left hemisphere is dominant over contralateral stimulation of the right hemisphere. The children with left-side UHL also demonstrated activation of the visual centers of the occipital lobe in response to the speech-in-noise task, suggesting cross-modal plasticity. The normal hearing controls and the children with right-side UHL showed no such activation, although previous studies have shown visual activation to auditory stimuli in children with normal hearing when they close their eyes while listening. This visual center activation only in children with left-side UHL may also be explained by the dominant nature of the left-hemisphere in processing auditory stimuli. The results of this study demonstrate that higher thresholds of activation in the attention centers of the brain in response to auditory stimuli may be responsible for increased rates of attentional and behavioral difficulties experienced by children with UHL.

Tibbetts, K., Ead, B., Umansky, A., Coalson, R. Schlaggar, B.L., Firszt, J., & Lieu, J.E.C. (2011). Inter-regional brain interactions in children with unilateral hearing loss. *Otolaryngology Head & Neck Surgery, 144*(4), 602 – 611.

Unlike fMRI, which shows patterns of neural activation in response to a specific stimuli, a resting state functional connectivity MRI (rs-fcMRI) shows the spontaneous neuronal activity of the brain as well as the functional connections that exist between neural structures. In this study, 16 children with severe-to-profound UHL and 10 normal-hearing sibling controls were placed in an MRI machine to obtain an accurate picture of resting-state neural activity. Imaging showed significant differences in the functional arrangement of the region of the brain responsible for sustained task maintenance in the children with UHL compared to the normal hearing controls. Additionally, the children with UHL had this region more closely correlated to another region thought to be responsible for echoic memory and generalized working memory. The authors speculate that children with UHL utilize

"subvocal rehearsal" to maintain attention to a task, resulting in a stronger connection between these two brain regions.

Children with UHL demonstrated atypical neural connectivity in four key brain regions: the left medial globus pallidus (associated with impulsivity and attentional disorders), the left middle temporal gyrus (associated with sentence comprehension), the mid-cingulate (associated with attention, decision-making, and error detection), and the right parahippocampal gyrus (associated with place processing, episodic memory, and contextual associative processing). All regions showed atypical neural connections to sensorimotor neurons, including those responsible for movement of the mouth for speech, possibly explaining difficulties in speech production seen in children with UHL. The authors state further research is necessary to determine which educational and outcome variables are associated with these atypical neural interactions, as they may guide targeted interventions in the future.

Ead, B., Hale, S., DeAlwis, D., & Lieu, J.E.C. (2013). Pilot study of cognition in children with unilateral hearing loss. *International Journal of Pediatric Otorhinolaryngology*, 77(1), 1856 – 1860.

Seven children with severe-to-profound UHL and a control group of seven normal hearing siblings underwent tests of cognition and phonological processing. Results showed the children with UHL demonstrated reduced phonological accuracy and efficiency when attending to unfamiliar verbal information, and demonstrated impaired executive functioning when attempting to maintain verbal information in working memory and listen to novel auditory stimuli simultaneously. These results suggest phonological processing is a significant component of normal working memory, and children with UHL have reduced capacity for multitasking. This corroborates previous MRI studies showing altered development of regions of the brain associated with dual-task control.

Rachakonda, T., Shimony, J.S., Coalson, R.S., & Lieu, J.E.C. (2014). Diffusion tensor imaging in children with unilateral hearing loss: a pilot study. *Frontiers in Systems Neuroscience*, 8, 87.

In this study of 49 children with severe-to-profound UHL, each subject underwent a specialized MRI referred to as "Diffusion Tensor Imaging" or DTI. This scan allows for visualization of the white matter tracts in the brain. A selection of auditory and non-auditory white matter tracts were visualized and compared to scans from a control group of normal hearing siblings. Results showed children with UHL had reduced microstructural integrity in two auditory regions and two non-auditory regions when compared to normal hearing controls. Correlations were found linking reduced structural integrity of certain auditory regions to specific

outcomes such as Individualized Education Plans, speech therapy, language ability, verbal IQ, performance IQ, and full IQ. Given that many children diagnosed with UHL undergo an MRI to determine etiology, the authors suggest DTI could someday be used to determine the educational needs of the child before academic difficulties actually manifest.

All subjects with UHL, regardless of side of impairment, exhibited asymmetries in white matter organization between the left and right brain hemispheres similar to what is seen in children with normal hearing. The authors suggest that fMRI studies showing reduced activation of these structures supports the notion that some auditory areas have been recruited for use by other regions of the brain.

Schmithorst, V.J., Plante, E., & Holland, S. (2014). Unilateral deafness in children affects development of multi-modal modulation and default mode networks. *Frontiers in Human Neuroscience*, 8, 164. <http://doi.org/10.3389/fnhum.2014.00164>

In this fMRI study, 21 children with moderate to profound UHL and 23 normal hearing controls underwent an MRI while completing a receptive language task (Token Sentence test). The child heard a sentence, then saw an animation, and the child pressed a button if what was shown in the animation matched what was heard in the sentence. Results showed the children with severe-to-profound UHL or right-side UHL had significantly less activation of right hemisphere regions responsible for secondary visual processing, indicating changes in neural modulation of cross-modal stimuli. The children with left-side UHL or a moderate degree of UHL did not show this reduction in visual activation. Children with UHL for less than two years demonstrated increased activation of the left superior temporal gyrus, possibly as a result of recent neural restructuring in response to a change to primarily monaural input.

The subjects with UHL also demonstrated reduced deactivation in multiple "default-mode networks" or DMN. Default-mode networks are considered responsible for resting state tasks, or tasks not requiring significant cognitive demand. When DMNs are not adequately suppressed during a cognitively-demanding task, attention and focus can be impaired. The authors state this is evidence that receiving only a monaural auditory signal in the brain leads to changes in higher-order cognitive resources.

A Summary of UHL & the Brain

Imaging studies of the brains of children with unilateral hearing loss have given us remarkable information as to why

these children demonstrate deficits in specific domains. Abnormal neural connectivity between areas of the brain associated with impulsivity, attention, episodic memory, and sentence comprehension correlate to specific behaviors commonly noted in this population. These children also require more auditory stimulation to achieve levels of neural activation similar to that seen in normal hearing peers. A reduced ability to suppress their "default-mode networks" may contribute to children with unilateral hearing loss having an impaired ability to maintain focus, and evidence of cross-modal plasticity suggests maladaptive changes to the organization of sensory neural structures. In the future, neural imaging may allow us to determine areas of future deficit in children with unilateral hearing loss, and allow for the implementation of targeted management strategies before these deficits are expressed.

Outcomes in Children with UHL

Although classified as "minimal hearing loss," it could be argued that UHL is anything but minimal. Research in the 1980s demonstrated that children affected with UHL had significantly poorer performance both socially and academically when compared to their normal hearing peers. In the decades since, the importance of this outcome data has not waned. By tracking performance data for these children, we cannot only determine the effectiveness of common gold-standard management strategies, but also gauge how effectively these children integrate into mainstream classrooms.

In the United States, intervention services are mandated by the federal government, but each state decides who qualifies for these services. As it stands now, children with UHL are eligible to receive services in just over half of states. Outcome data for this group of children is important in the fight for expanded access to services for children with UHL and other "minimal" hearing losses.

Johnson, C.E., Stein, R.L., Broadway, A., & Markwalter, T.S. (1997). Minimal high-frequency hearing loss and school-age children: Speech recognition in a classroom. *Language, Speech, and Hearing Services in Schools, 28*(1), 77 – 85.

Children with minimal high-frequency hearing loss (including UHL) were less able to correctly identify consonants in quiet than normal hearing children. When noise was added to the task, or when the task was changed to vowel identification, there was no difference in performance between the two groups. A qualitative analysis of the results showed the children with minimal high-frequency hearing loss received

43% less sibilance information from the auditory signal in quiet than the normal hearing children. The authors strongly suggest children with minimal high-frequency hearing loss utilize amplification and/or an FM system in a quiet classroom.

English, K., & Church, G. (1999). Unilateral hearing loss in children: An update for the 1990s. *Language, Speech, and Hearing Services in Schools, 30*(1), 26 – 31.

Prior to this study, research had shown that children with UHL were more likely to repeat a grade than their normal hearing peers. Beginning in the early 1990s it was discovered that forcing a child to repeat a grade had a temporary positive effect, but was followed by a significant and long-lasting detrimental impact on their overall academic success. Following that finding, it was recommended that students be given individualized educational services rather than repeating a grade. The authors sought to uncover what services children with UHL were typically receiving. For this study, surveys were mailed to the families of 423 children with UHL. Responses indicated 46% of students were not actively receiving services as it was believed their performance was adequate for their age and instead were being monitored for detrimental changes in academic performance. Of the 54% of children with UHL receiving services, the most common service was fitting with amplification and subsequent monitoring by an educational audiologist. Asked to rate the academic performance of the child, 63% were rated to have average academic performance, and 24% were rated as having below average academic performance. Results suggest the percentage of children with UHL exhibiting below average academic performance is unchanged since the 1980s.

Davis, A., Reeve, K., Hind, S., & Bamford, J. (2002). Children with mild and unilateral hearing impairment. *Proceedings of the Second International Conference "A Sound Foundation Through Early Amplification" (14).*

Retrieved from <http://www.audiologyonline.com/files/content/01000/01051/phonak-2001proceedings-20-chapter14.pdf>

A quality of life questionnaire was mailed to 150 families of children with a mild bilateral hearing loss or a UHL. Responses from parents indicated most had few concerns about their child's ability to hear in quiet or noise, although those children fit with hearing aids showed improved ease of listening in noise. 80% of the children with UHL or mild bilateral hearing loss had been fit with amplification. The average age at initial fitting of amplification was 5 years. Of the children with UHL who had been fit with amplification, 50% reportedly never wore the device, while 26% wore it "all of the time." Two-thirds of respondents stated their child's

hearing loss impacted the child's life "not too much", "very little", or "not at all." Interestingly, parents of children with UHL rated their children as significantly more clumsy than did parents of children with bilateral mild hearing loss. When results were aggregated and compared to earlier studies of children with greater severities of hearing loss, it was found the children with UHL and mild bilateral hearing loss had a similar impact on the family as a child with a moderate bilateral hearing loss.

Kiese-Himmel, C. (2002). Unilateral sensorineural hearing impairment in children: analysis of 31 consecutive cases. *International Journal of Audiology, 41*(1), 57 – 63.

Children with sensorineural UHL had no delay in utterance of first-words compared to normal hearing children, but there was a significant delay (an average of 5 months) in their utterance of two-word phrases. When administered standardized language tests, the children with sensorineural UHL scored within the normal range for their age and gender. The author suggests sensorineural UHL has minimal impact on language performance, at least on the language tasks tested in this study.

Niedzielski, A., Humeniuk, E., Blaziak, P. & Gwizda, G. (2006). Intellectual efficiency of children with unilateral hearing loss. *International Journal of Pediatric Otorhinolaryngology, 70*(9), 1529 – 1532.

This study sought to determine the impact of UHL on a child's intellectual abilities. The authors hypothesized that there would be differences in intellectual development based on the side of the affected ear. 64 children with UHL were administered subtests from the Wechsler Intelligence Scale for Children-Revised (WISC-R), including tests of verbal ability and performance ability. Results showed that although children with UHL had overall intelligence quotient levels on par with their normal hearing peers, there were specific areas of deficiency related to side of impairment. Those children with right-side UHL had significantly poorer performance on measures of verbal intelligence when compared to children with left-side UHL. Deficiencies were noted in the areas of logical thinking, abstract thinking, classifying, verbal learning ability, and conceptual understanding. Children with left-side UHL performed significantly poorer on measures of non-verbal intelligence compared to children with right-side UHL, and these children demonstrated deficiencies in areas of visual-motor coordination, spatial imagination, and visual memory. These results are important, as they suggest side of impairment can be used to determine in which domains a child with UHL may experience difficulty. Further research is needed to determine if addressing these areas of concern before the child demonstrates academic difficulty might be beneficial to overall outcomes in children with UHL.

McFadden, B. & Pittman, A. (2008). Effects of minimal hearing loss on children's ability to multitask in quiet and in noise. *Language, Speech, and Hearing Services in Schools, 39*(7), 342 – 351.

In an academic setting, children are often expected to multi-task. For instance, a child could have to write out an assignment while the teacher or classmates speak. Because minimal hearing loss (defined as unilateral, high frequency, or mild) can impact performance on certain auditory tasks, the authors sought to understand the nature of this impact on the ability to multitask in quiet and in noise. For the purpose of this study, 10 normal hearing children and 11 children with minimal hearing loss were placed in a room and given two simultaneous tasks. One was a simple series of connect-the-dot games and another was categorizing a list of nouns played through a speaker. Three rounds were completed, one in quiet and two with varying signal-to-noise ratios. Results showed children with minimal hearing loss had poorer performance on the more difficult auditory task as the signal-to-noise ratio decreased, while performance on the connect-the-dots task remained the same. While the normal hearing children diverted cognitive resources from the visual task to complete the auditory task, the children with minimal hearing loss did not demonstrate this same ability. The authors recommend children with minimal hearing loss be given accommodations to reduce or eliminate the necessity of multitasking.

Borton, S., Mauze, E., & Lieu, J. (2010). Quality of life in children with unilateral hearing loss: A pilot study. *American Journal of Audiology, 19*(1), 61–72.

UHL has been shown to have a varying degree of negative impact on a child's classroom performance and behavioral aptitude, but no study had investigated the impact of UHL on the child's overall quality of life. For the first phase of this study, children with UHL and their parents were divided into separate focus groups and asked to discuss their experiences. The children's group stressed the importance of adapting to situations they found challenging. Parents discussed a variety of experiences regarding their child's UHL, with many expressing concern for their children's social and academic performance as well as a need to constantly educate teachers and other professionals about their child's hearing needs. In the second phase, a generic pediatric quality of life survey was distributed to children with normal hearing, unilateral hearing loss, or bilateral hearing loss and their parents. Results from both parents and children indicated there were no statistically significant differences in the quality of life between these three populations. However, the children with UHL and their parents did express significantly more variability than the two control groups in the domain of

Social and School Functioning, indicating UHL may impact quality of life in specific environments.

Lieu, J.E.C., Tye-Murray, N., Karzon, R.K., & Piccirillo, J. (2010). Unilateral hearing loss is associated with worse speech-language scores in children. *Pediatrics*, 125(6), 1348 – 1355.

Many studies of UHL in children consist of small retrospective case studies. While these small sample sizes allow for greater efficiency and more specific or rigorous testing, they can be more vulnerable to influence from outside variables unaccounted for in the study. To combat this problem, the authors of this study specifically sought a large sample size with an age-matched control group to further account for variables impacting the success of children with UHL. In this case-control study, 148 children with UHL (ages 6 to 12) were compared to their siblings with normal hearing. Of the children in this study, less than half had trialed any form of amplification (hearing aid, CROS, BAHA, or FM system). UHL was associated with a significant reduction in performance on the Oral and Written Language Scales (OWLS) when compared to their normal hearing siblings. Maternal education level and socioeconomic status were also associated with poorer oral expression and oral composite scores.

The children with UHL were more likely to have an IEP and be receiving speech therapy than their normal hearing siblings. The authors point out that because each state determines which students are eligible for 504c accommodations, children with UHL are often not eligible. The authors strongly suggest children with UHL receive accommodations and academic support similar to those given to children with bilateral hearing loss.

Lieu, J.E.C., Tye-Murray, N., & Fu, Q. (2012). Longitudinal study of children with unilateral hearing loss. *The Laryngoscope*, 122(1), doi: 10.1002/lary.23454

This was a three-year longitudinal study of children 6 to 8 years of age with permanent UHL. While verbal and full-scale IQ, oral expression, and oral composite scores significantly improved over the three-year period, there was no change in rate of Individualized Education Plans, enrollment in speech therapy, or academic difficulty. This would indicate delays experienced by children with UHL are persistent over time.

Lieu, J.E.C., Karzon, R.K., Ead, B., & Tye-Murray, N. (2013). Do audiologic characteristics predict outcomes in children with unilateral hearing loss? *Otology & Neurotology*, 34(9), 1703 – 1710.

Children with UHL had lower mean vocabulary and language scores, as well as lower full-scale IQ scores than normal hearing sibling controls. The children with UHL also had poorer word recognition scores in quiet and in noise. Severity of UHL was correlated with poorer performance on oral and expressive language ability. No meaningful differences were found in children with left-ear UHL versus those with right-ear UHL. Two prominent variables correlated with better performance across tasks were maternal education and socioeconomic status of the family, a correlation that is well-established in pediatric research. In discussing the results of word recognition testing, the authors question whether or not improving the word recognition ability of children with UHL could lead to improvements in academic performance.

Kishon-Rabin, L., Kuint, J., Hildesheimer, M., & Roth, D.A.E. (2015). Delay in auditory behaviour and preverbal vocalization in infants with unilateral hearing loss. *Developmental Medicine & Child Neurology*, 57(1), 1129 – 1136.

Until the advent of Universal Newborn Hearing Screenings, children with UHL were simply not found in time to study their first-year vocalizations. Researchers took normal hearing infants and those with permanent UHL and sorted them into four groups based on their hearing status and degree of risk for developmental delays based on the risk factors published by the Joint Committee on Infant Hearing in 2000. The goal was to understand how UHL impacted the vocalizations and expressive language abilities of children with UHL compared to their normal hearing peers while controlling for factors that might put either group at high-risk for speech-language delay or other developmental problems.

Data was obtained via parent questionnaires, consisting of a series of interviews regarding the auditory behaviors of their child. Of the infants with UHL, 21% demonstrated delays in auditory behavior and 41% demonstrated delays in preverbal vocalization ability. Infants with UHL were four times more likely to experience delayed auditory behavior and nine times as likely to experience delays in preverbal vocalizations than their normal hearing peers. There were no differences in abilities between the high-risk and low-risk groups in each hearing category.

Anne, S., Lieu, J.E.C., & Cohen, M.S. (2017). **Speech and language consequences of unilateral hearing loss: a systemic review.** *Otolaryngology-Head and Neck Surgery, 154*(4), 572–579.

This was a systemic review of thirteen studies to quantify the objective measures of speech and language delay among children with UHL. For children with severe to profound UHL, there is an overall detrimental effect on speech and language testing results. For children with mild to moderate UHL, evidence suggests there may be a small effect on speech and language measures. When studied longitudinally, speech and language delays appear to improve or resolve. The authors conclude there is no uniformity in the testing methods and the severity of the hearing loss tested in these studies. It was unclear if hearing aids or school support mechanisms were included or evaluated in the children. The data regarding effects of UHL on speech and language outcomes for children are inconsistent.

Lewis, D.E., Smith, N.A., Spalding, J.L., & Valente, D.L. (2017). **Looking behavior and audiovisual speech understanding in children with normal hearing and children with mild bilateral or unilateral hearing loss.** *Ear and Hearing, 1*–12. doi: 10.1097/AUD.0000000000000534

Eighteen children with normal hearing (NH), eight children with mild bilateral hearing loss (MBHL), and ten children with unilateral hearing loss (UHL), from the ages of eight to twelve years, performed audiovisual tasks. The children performed in a complex multi-talker environment where they were evaluated for where they looked, for looking patterns as a function of hearing status, and whether looking behavior was related to performance on the task. Results showed children's looking to be quite variable, however, the children were able to complete the audiovisual tasks. The children with NH performed higher on the behavioral task than either group of children with hearing loss. No differences in performance were shown between children with UHL and children with MBHL. The authors of the study suggest that the performance of some children, whether they have NH or MBH/UHL, is not dependent on visual fixation to relevant talkers.

A Summary of Outcomes in Children with UHL

From an early age, UHL begins to negatively impact the child. Incidence of preverbal delay is significantly higher than normal hearing infants, and delays experienced in early childhood can persist for several years. Children with UHL demonstrate poorer expressive language skills, reduced verbal intelligence, and a decreased ability to multi-task.

Presentation of the hearing loss including severity and side of impairment will have an impact on specific receptive and expressive abilities due to the nature of the brain. While some research suggests these deficits have a minor impact on the child's overall quality of life, these skills seriously impact the child's ability to perform academically. As a result, these children require special services like speech-language therapy and Individualized Education Plans at a much greater rate than normal hearing children.

Management of UHL in Children

Only decades ago, management of UHL rarely consisted of more than preferential classroom seating. The burden of management relied significantly on the normal hearing ear, expected to perform well enough for sufficient academic performance. Once research began showing the challenges faced by these children, various amplification options were explored with varying success. Today, audiologists have more evidence-based treatments for children with UHL than ever before. Having recognized the need to tailor treatment to the individual communication needs and preferences of the child and their family, these options are diverse in nature and can accommodate a range of needs.

Management: Wireless Microphone Systems

Kenworthy, O.T., Klee, T., Tharpe, A.M. (1990). **Speech recognition ability of children with unilateral sensorineural hearing loss as a function of amplification, speech stimuli, and listening condition.** *Ear and Hearing, 11*(4), 264 – 270.

Children with UHL underwent speech recognition testing with an FM system, a CROS device, and unaided. Each round of testing simulated a different seating position in the classroom (signal coming from midline, signal reaching impaired ear first, and signal reaching normal hearing ear first). Listening with the FM system was the only condition that resulted in consistently high speech recognition scores regardless of the position of the child or the type of speech stimulus used.

Maxon, A.B., Brackett, D., & van den Berg, S. (1991). Classroom amplification use: a national long-term study. *Language, Speech, and Hearing Services in Schools, 22*(10), 242 – 253.

In an effort to understand the ways in which schools adopt and use FM systems and to examine possible changes in attitudes and adoption of FM systems over time, two surveys of speech-language pathologists, audiologists, and teachers of the hearing impaired were completed. The first round of questionnaires was distributed in 1981 – 1982 and a subsequent questionnaire distributed in 1988 – 1989. For both distributions, the questionnaire was the same and consisted of three components, the first being questions related to involvement of academic staff with FM use, the second was attitudes regarding FM use, and finally each participant was asked questions about the population of children they serve.

Responses showed that audiologists were frequently not consulted before schools purchased FM systems, and hearing-impaired children and their parents had little involvement in the selection or purchasing process. There was a decline in full-time FM use by the second questionnaire, with more children using FM systems incidentally or using other means of amplification instead. A trend toward greater professional acceptance of FM systems was noted in the second questionnaire; however, teachers still only preferred to use the FM microphone and transmitter about 50% of the time. While the authors noted some positive trends, there were still significant concerns regarding the adoption and implementation of FM systems in schools.

Kopun, K., Stelmachowicz, P.G., Carney, E., & Schulte, L. (1992). Coupling of FM Systems to Individuals with Unilateral Hearing Loss. *Journal of Speech and Hearing Research, 35*(1), 201 –207.

The purpose of this study was to examine the occlusion characteristics of five coupling options commonly utilized for FM systems at time of publication. Because children in a classroom would need to hear classmates throughout the day as part of learning, it is assumed occlusion from headphones or an earmold could negatively impact the audibility of non-FM speech. Results indicated the only fitting option with no occlusion was a tube-fitting with no earmold attached. Of the four remaining options, the lightweight headphones had the least amount of occlusion, with attenuation of less than 5 dB through 4 kHz. The remaining options all involved an earmold, which resulted in 15-30 dB of attenuation in the high-frequencies. The authors recommend performing real-ear measures to verify changes in ear canal resonance when fitting a child with an FM-system, as this can negatively impact audibility of speech from other sources.

Urdike, C.D. (1994). Comparison of FM auditory trainers, CROS aids, and personal amplification in unilaterally hearing impaired children. *Journal of the American Academy of Audiology, 5*(3), 204 – 209.

Six children with UHL (ages 5 to 12 years) were fit with either an FM system, a CROS system, or a hearing aid and underwent speech recognition testing in quiet and in noise (+6 dB SNR). The CROS system and hearing aid resulted in no improvement in performance in noise and only the child with a mild degree of UHL showed improvement in quiet with either of these devices. While using the FM system, every child in the study demonstrated improvement in performance in noise, and a majority showed improvements in performance in quiet as well. The author urges the use of FM systems for children with UHL.

Tharpe, A.M., Ricketts, T., & Sladen, D.P. (2003). FM systems for children with minimal to mild hearing loss. In *ACCESS: Achieving Clear Communication Employing Sound Solutions* (chapter 20). Retrieved from https://www.phonakpro.com/content/dam/phonak/b2b/FM_eLibrary/ACCESS_Chapter_20_Anne_Marie_Tharpe.pdf

In a study of fourteen children with minimal hearing loss (three of which had UHL), each child was fit with an FM system in three conditions (closed fit monaural, open fit monaural, and open fit binaural). The children utilized the FM system for two weeks in each condition, and each two-week period was followed by speech-in-noise testing with the device, a teacher questionnaire to understand the child's academic performance while using the device with each tested configuration, and a self-report questionnaire to measure the child's opinions and preferences regarding each FM configuration.

Results showed that for speech-in-noise testing, performance in the bilateral FM condition was significantly better than in the monaural open condition for speech coming from 0 or 270 degrees azimuth, but there was no significant difference in performance when speech came from 90 or 180 degrees azimuth. While on average teachers gave the children slightly higher scores after use of the FM, this increase was not statistically significant. An analysis of those students with scores in the marginal or failure range on questionnaire subtests pre-FM fitting showed that after consistent FM use the number of students in the marginal or failure score range dropped from 54% to 26%. Answers to the self-report questionnaire showed 74% of the children liked wearing the FM system as it made hearing the teacher easier, all of the children preferred the monaural condition, all but one child wanted to keep the FM system, and 90% of the children had no preference between earmold types.

McKay, S. (2008). Frequency modulated (FM) considerations for children with minimal/mild or unilateral hearing loss. *Perspectives on Hearing and Hearing Disorders in Childhood*, 10 – 18.

Children with UHL often struggle with hearing in noisy environments and at a distance, two situations in which FM systems offer considerable benefit. When selecting an FM system, it is important to consider what difficulties the child experiences in the classroom, what amplification they are currently using or have used in the past, and any special considerations given the nature of their hearing loss. Caution is recommended when considering soundfield FM systems. While proven beneficial for even normal hearing children, they are insufficient for those with higher degrees of UHL and may hinder the child from receiving additional academic services. Existing evidence neither confirms or denies the benefit of FM systems for infants and toddlers with UHL, but it may be useful for hearing-in-noise.

Should the audiologist choose to validate performance with FM systems, several options exist. Functional measures should be conducted with a stimulus and masker that closely resemble the classroom environment. Functional measures can also be completed in the classroom, with the teacher completing a performance evaluation with and without the FM system. Questionnaires can also be used to find out how the child functions in the classroom and at home with an FM system. This also allows for greater involvement of the parents in the treatment process. Audiologists should work with parents to ensure they are aware of ways in which they can optimize the at-home listening environment for the child with UHL.

Stewart, E., Houston, B., & Holstad, B. (2016). Optimization of device fittings for two children with unilateral hearing loss: The benefit of bilateral Roger use. Poster session presented at the 7th International Pediatric Audiology Conference, Atlanta, GA.

The authors sought to determine the optimal amplification configuration for two children with UHL fit with Bone-Anchored Hearing Aids [BAHA], one with single-sided deafness and the other with unilateral microtia and atresia. In both cases, speech recognition in noise was maximized with bilateral use of a Roger wireless microphone system via a receiver attached to the BAHA and a Roger Focus receiver in the better ear. The authors encourage audiologists to think beyond traditional amplification for children with UHL.

Rance, G. (2018). Remote Microphone Listening Devices for Individuals with Unilateral Hearing Loss. *Field Study News*.

Eight school-aged children and six adults with unilateral sensory hearing loss ranging from mild to profound, with no previous hearing aid experience, were fitted with a device configuration that provided them the highest speech perception score in background noise. In most cases, the device setup was either a Roger Focus worn on the better ear and a hearing aid plus Roger on the poorer ear. A subjective assessment of performance was completed by the children (and their teachers) and the adult subjects post-fitting. The results showed significant improvement in their real-world listening and comprehension abilities when wearing the auditory devices. The authors suggest that remote microphone systems, specifically a Roger Focus device fitted to the better ear, either alone or in combination with a hearing aid/Roger Integrated Receiver on the poorer side, can achieve significant perceptual benefits for children and adults with unilateral hearing loss.

Management: Hearing Aids

Kiese-Himmel, C. (2002). Unilateral sensorineural hearing impairment in children: analysis of 31 consecutive cases. *International Journal of Audiology*, 41(1), 57 – 63.

Twenty-six children (1 – 10 years of age) diagnosed with a sensorineural UHL were fit with unilateral amplification. Approximately 81% of the children accepted their hearing aid and wore it for most of the day (excluding weekends).

McKay, S. (2002). To Aid or Not to Aid: Children with Unilateral Hearing Loss. Retrieved from <http://www.audiologyonline.com/articles/to-aid-or-not-children-1167>

In a study of 28 children (ages 2 to 17) with UHL fit with a hearing aid, 20 families completed retrospective surveys. Responses to the survey indicated most children showed improvements in academic and social areas where auditory abilities had been questioned before receiving amplification. A majority of parents stated they wished they had pursued amplification sooner. Although the responses from the children indicated some had cosmetic concerns regarding the device, they still chose to wear them due to their beneficial impact on hearing. Regardless of performance in the classroom, the author believes children with UHL should be amplified due to the positive impact on overall quality of life.

McKay, S., Gravel, J.S., & Tharpe, A.M. (2008). Amplification considerations for children with minimal or mild bilateral hearing loss or unilateral hearing loss. *Trends in Amplification, 12*(1), 43 – 54.

The authors explore considerations in fitting children with minimal hearing loss (including UHL) with amplification. Children with UHL are sometimes not identified until later in development, and as a result they can be significantly impacted by auditory deprivation in the impacted ear. These children also lack important binaural advantages like binaural summation, interaural timing and level differences, and binaural release from masking. Without these binaural advantages, children with UHL often demonstrate difficulty not only hearing, but also localizing to sounds and hearing in noisy environments.

Underscoring the importance of fitting children with UHL with amplification strictly on a case-by-case basis, the authors state that some will benefit just as much from preferential seating as they would from a hearing aid in the impacted ear. In a review of existing literature on acceptance and utility of a variety of amplification options, it is shown that children with a mild-to-moderate degree of UHL seem to have higher rates of acceptance of hearing aids than children with a severe-to-profound degree of UHL. Regardless of the management strategy that is decided upon, it is important to use appropriate validation measures to monitor performance. Finally, parents and caregivers of children with UHL should be given appropriate information and follow-up resources to make informed decisions regarding their child's health and to optimize their at-home communications.

Johnstone, P.M., Nábělek, A.K., & Robertson, V.S. (2010). Sound localization acuity in children with unilateral hearing loss who wear a hearing aid in the impaired ear. *Journal of the American Academy of Audiology, 21*(8), 522 – 534.

Localization ability of children with sensorineural UHL who had worn hearing aids for at least three months was measured with and without their hearing aid. Results showed the benefit of a hearing aid for localization depended on the age the child was first fit, the current age of the child, and the degree of hearing loss. Localization ability was greatest in those children who had been fit with a hearing aid before the age of 5. The older children in the study who were fit with a hearing aid after the age of 7 showed a lack of binaural benefit, resulting in greater errors in localization. Without hearing aids, the older children in the study demonstrated significantly better localization ability than the younger children without hearing aids. Results of this study

seem to indicate that hearing aids are unable to restore localization ability to normal levels in children with UHL.

Briggs, L., Davidson, L., & Cho Lieu, J. (2011). Outcomes of conventional amplification for pediatric unilateral hearing loss. *Annals of Otology, Rhinology, and Laryngology, 120*(7), 448 – 454.

Eight children (ages 7 to 12) with mild to moderately-severe UHL were fit with conventional amplification in the impaired ear. Speech perception in quiet and in noise was measured pre- and post-fitting, following an acclimatization period of three months. Results showed no difference in performance between the aided and unaided conditions. Subjective assessments were given to the children, their parents, and their teachers at regular intervals pre-fitting and post-fitting. Responses from these subjective surveys showed children with UHL received significant benefit from the conventional hearing aid while in the classroom, at home, and in other challenging listening environments. The authors encourage children with UHL to receive a trial with amplification along with monitoring to determine their level of benefit with the device.

Purcell, P.L., Jones-Goodrich, R., Wiseneski, M., Edwards, T.C., & Sie, K.C.Y. (2016). Hearing devices for children with unilateral hearing loss: patient- and parent-reported perspectives. *International Journal of Pediatric Otorhinolaryngology, 90*(1), 43 – 48.

In interviews of parents of 50 children with UHL, it was shown that 34 of the 50 children (68%) had at some point tried amplification (hearing aid, CROS, or BAHA). Of these, 20 children (59%) had chosen to wear the devices long-term indicating they received positive benefit from the devices. Children with UHL who no longer wore amplification cited discomfort and lack of benefit as the primary reasons.

In interviews of 16 children with UHL, 73% reported being given preferential classroom seating. However, half of these children reported experiencing problems related to social stigma when finding or obtaining appropriate seating. FM usage was reported by 10 (63%) of the participants, but 4 children complained of poor sound quality and 3 children reported stigma associated with using an FM system in the classroom. Children with UHL who had never tried amplification cited social stigma as the primary reason. The authors cite the high uptake rate found in this study as proof that children with UHL should be considered candidates for amplification, and audiologists should work with patients and their families to determine the most suitable type of amplification for that patient.

Management: Cochlear Implants

Hassepass, F., Aschendorff, A., Wesarg, T., Kroger, S., Laszig, R., Beck, R.L., ... Arndt, S. (2012). Unilateral deafness in children: audiologic and subjective assessment of hearing ability after cochlear implantation. *Otology & Neurotology*, 34(1), 53 – 60.

Two children with post-lingual single-sided deafness (SSD) and one child with peri-lingual SSD received a cochlear implant (CI) in the impaired ear. After twelve-months of use, both of the post-lingually deafened children demonstrated significant improvement in hearing-in-noise and localization abilities as compared to their pre-implant performance. Responses to subjective assessments by both children and their parents showed positive benefit and improved listening outcomes. This suggests cochlear implants may restore binaural hearing abilities in children with profound UHL. As the child with peri-lingual hearing loss was too young for most of this test battery, only subjective assessment was obtained. One year post-implantation, the peri-lingual child demonstrated significant improvement in everyday listening. Further research is necessary to determine the impact of factors like duration of deafness and etiology on CI outcomes for these patient populations.

Plontke, S.K., Heider, C., Koesling, S., Hess, S., Bieseke, L., Goetze, G., & Rahne, T. (2013). Cochlear implantation in a child with posttraumatic single-sided deafness. *European Archive of Otorhinolaryngology*, 270(1), 1757 – 1761.

In this case study, an 8-year old boy with profound UHL following a traumatic temporal bone fracture showed evidence of cochlear fibrosis. Given that treating post-lingual SSD with a cochlear implant had proven successful in adults, the decision was made to implant the child before the cochlear fibrosis limited the ability to implant in adulthood. Three months post-implantation, the child demonstrated significant improvement in speech recognition in the implanted ear as well as localization ability in noise. Six months post-implantation the child continued to demonstrate significant improvement in speech recognition and localization. These results suggest cochlear implants can at least partially restore binaural hearing ability in children with post-lingual onset UHL.

Távora-Vieira, D. & Rajan, G.P. (2015). Cochlear implantation in children with congenital and noncongenital unilateral deafness: a case series. *Otology & Neurology*, 36(1), 235 – 239.

Four children with UHL (three congenital and one acquired) were implanted in the impaired ear. The child who acquired UHL post-lingually demonstrated improvement in speech

recognition in noise and localization, suggesting restoration of binaural hearing ability following implantation. For the children with congenital UHL, it is posited that binaural integration of the electrical and acoustic signals can only occur if implantation happens in the "critical period" of 24 months of age. The youngest child in the study was 17 months old at time of implantation, and demonstrated no issues accepting the implant, but was too young for formal measures of binaural hearing. After twelve months, the other two children with congenital UHL, ages 4.5 and 6.8 years, demonstrated no binaural benefit from the cochlear implant. The authors suggest because these children were implanted later in childhood their brain was unable to adapt to bilateral input. It is recommended that children with congenital UHL undergo intervention as soon as possible following diagnosis, especially if a cochlear implant is sought.

Arndt, S., Prosse, S., Laszig, R., Wesarg, T., Aschendorff, A., & Hassepass, F. (2015). Cochlear implantation in children with single-sided deafness: Does aetiology and duration of deafness matter? *Audiology & Neurotology*, 20(1), 21 – 30.

Thirteen children with congenital, peri-lingual or post-lingual SSD, implanted with a cochlear implant were tested pre-implantation and post-implantation to determine the impact of duration and etiology of deafness on CI treatment outcomes. It was noted that of the original pool of children with SSD over 50% had Cochlear Nerve Deficiency (CND.) This made them ineligible to receive a cochlear implant. This makes MRI imaging of these children vital when determining candidacy and alternative amplification strategies should be explored.

Of the implanted children, there was a significant correlation between duration of deafness and treatment outcome as well as etiology and outcome for those children with congenital SSD. Subjects implanted after four years of age demonstrated much poorer outcomes than the subjects implanted before age four. The child implanted at approximately two years of age demonstrated evidence of binaural integration post-implantation. More research is needed to determine if the critical period for CI intervention in children with SSD is wider or narrower than for bilaterally deaf children. In an examination of etiology of congenital loss and treatment outcome, those children with congenital SSD as a result of cytomegalovirus infection in utero had overall poorer outcomes than children with other congenital etiologies. This same trend of poor CMV-related outcomes has been noted in studies of adult CI recipients as well.

For the children with post-lingual SSD, results showed evidence of binaural hearing abilities including improved sound localization and speech recognition in noise. These

results are similar to those seen in adults with SSD who have been implanted. While more research is needed to determine the optimal time window for implantation of children with pre-lingual and peri-lingual SSD, children with post-lingual SSD receive significant benefit from the restoration of binaural hearing abilities resulting from cochlear implantation in the impaired ear.

Management: Miscellaneous

Holstrum, W.J., Gaffney, M., Gravel, J.S., Oyler, R.F., & Ross, D.S. (2008). Early intervention for children with unilateral and mild bilateral degrees of hearing loss.

Trends in Amplification, 12(1), 35 – 41.

Children with UHL and their parents face many challenges when seeking treatment. These challenges include navigating differences in eligibility for Early Intervention services in each state, parents misinterpreting UHL as a "minimal" hearing loss and subsequently seeking treatment with less urgency, losing Early Intervention services after age three when the child transitions to Part B services, and a lack of understanding regarding the specific deficits faced by these children as well as the ways in which typical development is impacted. Audiologists should provide families with information to address these challenges, including resources to help parents navigate obtaining intervention services, as well as information regarding the optimization of communication in the home. Parents should be informed as to how they can monitor their child's developmental progress, as this will help determine efficacy of any services received as well as selected amplification strategies.

Noh, H. & Park, Y. (2012). How close should a student with unilateral hearing loss stay to a teacher in a noisy classroom? *International Journal of Audiology, 51*(6), 426 – 432.

The optimal distance from the speaker for an older child (10 – 19 years of age) with UHL is between 4.35 and 6.27 meters. When seated 4.35 meters from the speaker, the child with UHL will have speech discrimination scores similar to those of a normal hearing adult. When sitting 6.27 meters from the speaker, the child with UHL will have speech discrimination scores similar to those of normal hearing peers at 10 meters distance from the speaker. As the distance between the speaker and the child with UHL increases, there is a larger drop in speech discrimination performance than seen in normal-hearing controls. Younger children may need to be seated closer to the instructor than older children, and individual variations in performance should be taken into account when determining preferential seating for children with UHL.

Christensen, L. & Dornhoffer, J.L. (2008). Bone-anchored hearing aids for unilateral hearing loss in teenagers.

Otology & Neurotology, 29(8), 1120–1122.

Three teenagers (ages 16 to 18) with UHL were implanted and fit with a BAHA. All three teens demonstrated significant improvement in speech recognition in quiet and in noise with the BAHA. A subject assessment of performance was completed by the teenager and a parent pre-fitting and post-fitting and results showed significant improvement in hearing abilities following fitting of the BAHA. The authors suggest bone-anchored hearing aids are a viable alternative for teens who refuse to accept traditional amplification options.

Doshi, J., Banga, R., Child, A., Lawrence, R., Reid, A., Proops, D., & McDermott, A. (2013). Quality of life outcomes after bone-anchored hearing device surgery in children with single-sided sensorineural deafness. *Otology & Neurology, 34*(1), 100 – 103.

A cohort of 8 children with SSD were implanted with a BAHA and administered pre-fitting and post-fitting questionnaires to determine impact of the device on quality of life and listening ability. In total, 7 out of 8 children demonstrated significant improvement in overall quality of life with the BAHA. The child who did not show benefit was later shown to be suffering from self-confidence issues as a result of bullying. While this child scored poorly on measures of emotional impact, they still demonstrated neutral to positive benefit in terms of health, vitality, and learning measures. While the authors suggest a larger cohort is necessary to generalize results to the greater population, these results show BAHA as a promising treatment for children with SSD.

Appachi, S., Specht, J.L., Raol, N., Lieu, J.E.C., Cohen, M.S., Dedhia, K., & Anne, S. (2017). Auditory outcomes with hearing rehabilitation in children with unilateral hearing loss: a systematic review. *Otolaryngology-Head and Neck Surgery, 154*(4), 565–571.

A systemic review of twelve studies to evaluate the auditory outcomes of hearing rehabilitation in children with UHL. Bone-conduction hearing devices were commonly studied and shown to improve the functional and objective auditory outcomes for those with moderate to profound UHL. Inconclusive results were drawn on the benefit of CROS hearing aids. FM systems in the educational setting clearly benefit children with UHL. Conventional hearing aids appear to benefit children with mild to moderately severe UHL. The data are limited in the role of conventional hearing aids and FM systems in the classroom for children with UHL. The authors conclude the data supporting functional and objective auditory measures following amplification in children with UHL are limited, and further studies are needed.

A Summary of Management of UHL in Children

Wireless microphone systems are a powerful tool, and have been shown to significantly improve classroom listening abilities regardless of hearing status or primary means of amplification. As such, any child with unilateral hearing loss should have a wireless microphone solution integrated into their management protocol, if appropriate. For those children with mild to moderate UHL a trial with amplification should be given. This ensures any benefit from amplification may be measured by teachers and parents before committing to the technology. Although they may see no enhancement in performance, these children may still benefit emotionally and socially from the device. For children with severe-to-profound UHL, cochlear implants represent the only method proven to restore at least some of the benefits of binaural hearing in post-lingually deafened children or children still within the "critical period" at time of implantation. A BAHA may also offer children with SSD positive benefit, especially if contraindicated for a cochlear implant.

Gold-standard treatment of the child with UHL includes involvement of the parents and family. Unique cultural considerations as well as communication preferences must be taken into account when selecting the appropriate management strategies. Family involvement also facilitates optimization of the at-home listening environment. Teachers are also a valuable resource and can provide additional feedback regarding the efficacy of selected management strategies based on academic performance and classroom behavior. For the child with UHL, the ideal treatment involves a collaborative approach.

Conclusions & Final Thoughts

In just four decades, our understanding of UHL in children has grown tremendously, aided in part by the widespread implementation of universal newborn hearing screenings allowing for earlier identification of this population. At the same time, there is still so much to be learned. Continuations of advanced imaging studies will allow us not only to pinpoint neural mechanisms behind observed behaviors, but also design targeted interventions before academic and social deficiencies manifest. Until then, research conducted thus far gives insight into the best manner in which to identify and treat children with unilateral hearing loss: comprehensively and individually.

Each child with UHL faces unique communicative challenges brought about by a degradation or complete loss of binaural

hearing abilities. These can vary based on degree of impairment and side of impairment as well as the timing of onset and duration of hearing loss. For children identified with UHL, cranial imaging can be utilized to help determine etiology, identify further anomalies in the inner ear, and show any structural changes to the brain. Imaging studies can also be used to determine appropriateness of certain amplification strategies such as cochlear implantation. Additional diagnostic testing including genetic, vestibular, or ophthalmologic testing may be necessary to further diagnose hereditary etiologies or concomitant issues.

Selecting the appropriate amplification and accommodative strategies for children with UHL can be a complex process. Research shows acceptance of amplification for this population is mixed and navigating Early Intervention services can be challenging for parents unfamiliar with the process. Children with UHL should be given a trial with amplification to determine benefit and utilize a wireless microphone system in the classroom to ensure maximum audibility. Children with single-sided deafness, unable to benefit from traditional amplification, now show promising results with cochlear implants, which allow for at least partial restoration of binaural hearing if implanted post-lingually or within the "critical period." Involvement of the child's parents and teachers ensures that regardless of the selected amplification strategy, there are people who can monitor the progress of acceptance and subsequent benefit in multiple listening environments.

Over the past four decades, a growing body of research has explained the tremendous obstacles faced by children with UHL. As a result, comprehensive, individually-tailored treatment strategies can be formulated to help these children succeed socially and academically. Although we know more now than ever before, there is still much we do not know about the condition. How permanent are changes in neural architecture as a result of monaural auditory stimulation? How can newborn hearing screenings be modified to find more cases of UHL? When is the critical period for cochlear implantation for those children with congenital UHL? Hundreds of other questions still pervade the literature, and it may be several decades before we firmly understand how to best serve children with UHL. Until then, partnering with parents to determine the best combination of amplification, academic accommodations, and follow-up monitoring is the gold-standard of care for children with UHL.

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Authors of this compendium



Author

Chase joined Sonova in 2016 as Audiology Trainee, working at Advanced Bionics, Connect Hearing, Phonak, and Sonova HQ. He is now a Territory Manager for Phonak US. He received his Doctor of Audiology degree from Northwestern University.



Co-author

Jacqueline Drexler received her Doctorate of Audiology from the University at Buffalo. She joined Sonova in 2017 for a one-year formal development program. During that time she worked at Unitron, Connect Hearing Canada, Advanced Bionics, and Phonak.