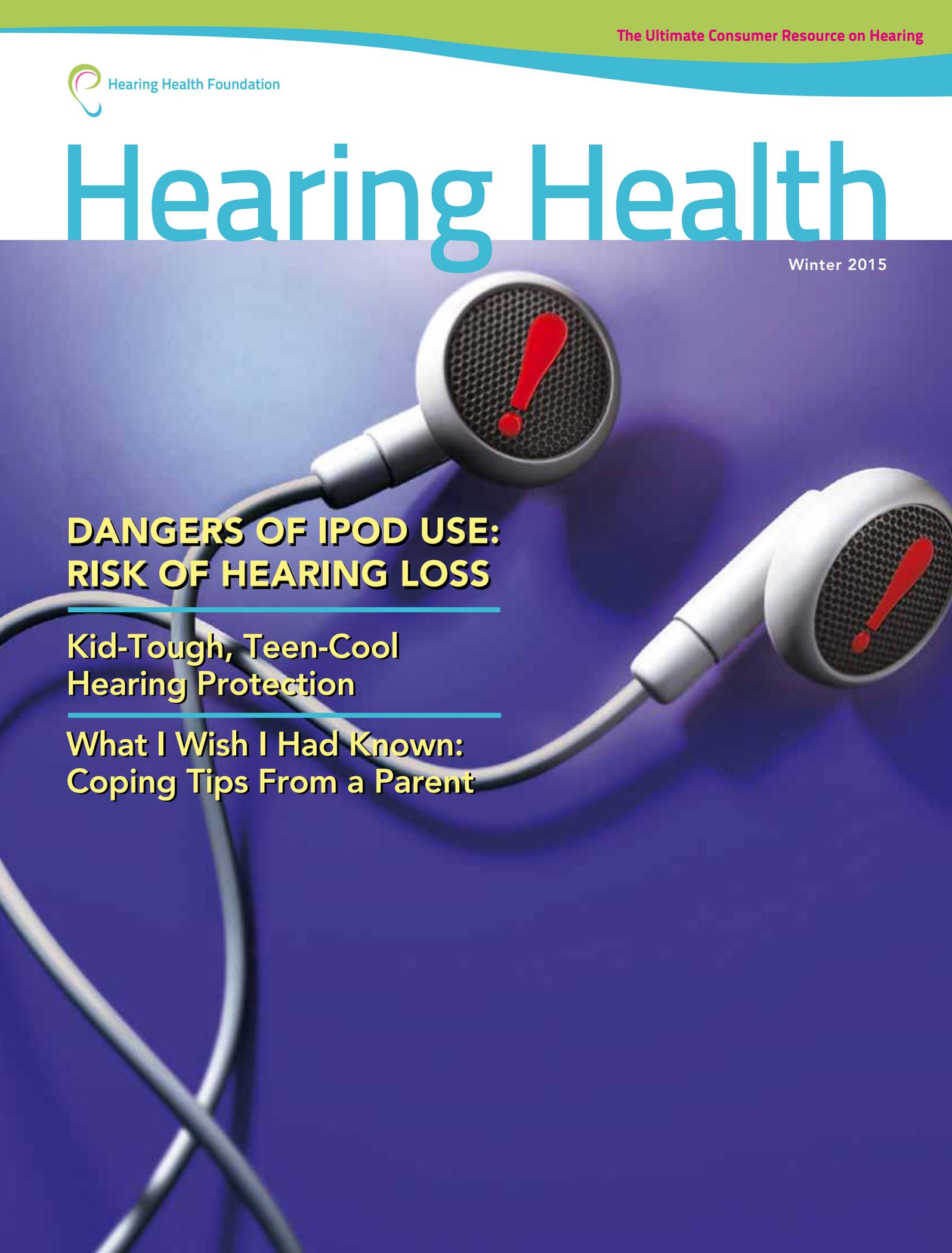


Hearing Health

Winter 2015



DANGERS OF IPOD USE: RISK OF HEARING LOSS

**Kid-Tough, Teen-Cool
Hearing Protection**

**What I Wish I Had Known:
Coping Tips From a Parent**



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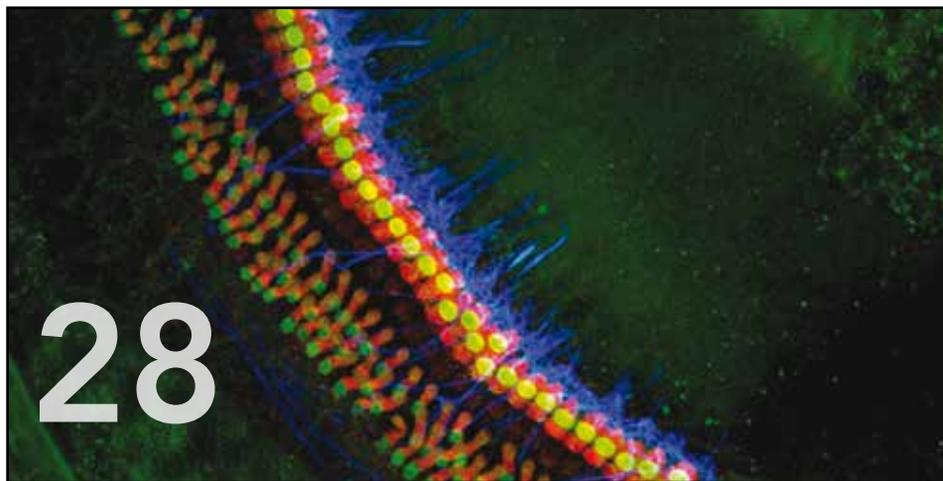
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Hearing Health

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I won't let my school-aged children wear earbuds to listen to music. This may sound extreme, but I have an adult-onset genetic hearing loss and I worry that I may have passed it onto them. I will take every precaution to make sure my children preserve the hearing they have now should problems strike later, and wearing earbuds is just too big a risk to take.

But can wearing earbuds really cause hearing loss? Not directly, but using them to listen to music at unsafe listening levels definitely can. In fact, listening unsafely with earbuds is likely the primary cause of the more than 30 percent rise in the incidence of teenage hearing loss that took place over the dozen years leading up to 2006. I can only imagine it has increased further in the past decade.

The good news is that this type of hearing loss is 100 percent preventable. At HHF, we are committed to helping educate parents, teens, schools, and other influencers on the importance of listening at safe levels. The story "The Dangers of iPod Use" (page 8) details some of the risks and provides important tips for parents and teens so that they are sure to listen as safely as possible.

Children are also exposed to noise in other venues, like sporting events, movie theaters, and even birthday parties. The article "The 95-Decibel Birthday Party" (page 24) provides reviews of several products, including earplugs and earmuffs, that can help children and teens (and even adults) protect their hearing.

Some children are born with hearing loss. In this issue we discuss new advances in testing for hearing loss in newborns and young children. The sooner hearing loss is diagnosed, the faster interventions can be made to ensure proper language development. Please see "5 Things I Wish I'd Known" (page 15), written by an HHF Junior Board member and parent of a child with hearing loss. She shares tips on getting the help you need when your young child has hearing loss.

As always we encourage you to learn more about HHF and help support our research for a cure for hearing loss and tinnitus. Visit hhf.org, where you can sign up for our informative monthly e-newsletter, create a fundraising event or giving page, make a tribute gift to honor a loved one with hearing loss, or make a tax-deductible donation. Be sure to "like" us on Facebook and follow us on Twitter @HearingHealthFn to stay up to date on the latest hearing research and news.

Warm regards,

Shari S. Eberts

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1. Buechner A, Dyballa K-H, Hehrmann P, Fredelake S, Lenarz T (2014) Advanced Beamformers for Cochlear Implant Users: Acute Measurement of Speech Perception in Challenging Listening Conditions. PLoS ONE 9(4): e95542. doi:10.1371/journal.pone.0095542

Dear Readers,

I am very pleased to announce that we are in the final stages of creating Hearing Health Foundation's strategic plan for 2015 to 2019. This is a big step forward for HHF. The strategic plan will define our organizational imperatives and set priorities that will help guide our decisions and actions over the next few years. This "road map" will lead us further down the path to a cure for hearing loss and tinnitus. I'll be sharing specifics of our new plan in the Spring issue of Hearing Health magazine that comes out in April. Here are some highlights:

The Hearing Restoration Project (HRP), our international hearing research consortium of leading scientists in the field, will continue to advance in its collaboration toward a cure for hearing loss and tinnitus by remaining focused on hair cell regeneration biology. In the current phase, the priority is on bioinformatics to compare and contrast the mountains of data we've collected by using finely tuned computer analyses. This will help identify the paths most important in regulating hair cell regeneration. (Learn about the unique mouse model developed by the HRP on page 28.)

Through our Emerging Research Grants (ERG) program, we will continue to identify and fund hearing loss, tinnitus, and balance research with the goal of increasing treatment options as well as protecting those people who are at risk. In 2015, we are funding research on central auditory processing disorders (CAPD), hyperacusis, Ménière's disease, and tinnitus. (Meet one of our ERG researchers on page 38.)

While HHF's primary goal remains research, we will begin a concerted effort in 2015 to empower adults, parents, caregivers, colleagues, and friends with information they need to prevent hearing loss and tinnitus and to improve the quality of life. This will include identifying and pursuing new ways to disseminate information about the importance of preventing hearing loss and tinnitus. To this end, the Safe and Sound Seal program for corporate partners will better connect us with like-minded people who can help us spread the message. We will also be encouraging grassroots support for the importance of hearing protection, hosting local events, and serving as a connection to the community for people living with hearing loss and tinnitus.

Building on successful communications efforts from the past, we will focus on engaging with the community and general public through the enhanced use of digital technology, leveraging the information in this magazine online as well as cultivating relationships with high-profile personalities who believe in HHF's mission. (Read about Ralph Johnson, a founding member of the legendary band Earth, Wind & Fire, and his commitment to hearing health on page 35.)

This coming year promises to be a turning point for HHF. I look forward to reporting back to you on our progress throughout 2015 so you can share in all of the activities and initiatives we aim to achieve. HHF is excited by what lies ahead, and we look to you for continued support of our mission to prevent and cure hearing loss and tinnitus.

Sincerely,



Claire Schultz

CEO, Hearing Health Foundation



Remembering Marion Downs

A force to be reckoned with, Marion P. Downs, Ph.D., was regarded as “the mother of pediatric audiology.”

By Amy Gross



I interviewed Marion Downs in 2007, shortly after her sassy “senior advice” book, “Shut Up and Live! (You Know How)” was published. It was only my second assignment for Hearing Health magazine, and I knew little about hearing loss, hearing research, or hearing technology. I had no idea how influential Marion Downs had been—and at the time, still was—regarding newborn infant screening, but it didn’t take much research to discover that this woman was a big, big deal. I don’t know why, but her passing on November 13, 2014, caught me by surprise. It didn’t matter that she had reached her 100th birthday; I, like many of her fans, found it difficult to accept that the force known as Marion Downs had moved on, peacefully, in her sleep.

Marion (she wouldn’t let me use any honorific) was 92 when we spoke. She was still skiing and swimming and playing tennis competitively, and one of the photos in “Shut Up and Live!” showed her gleefully skydiving with a handsome young instructor (she made sure to point out the “handsome” part several times). I had read every word of her book, in which she provided candid advice for anyone dealing with the aging process: the importance of weight training, why hearing aids are one of the secrets to a happy marriage, and how to maintain a healthy sex life into one’s senior years. I loved that she was able to make me blush more than a few more times; the woman minced no words.

But what had put Marion Downs on the map, audilogically speaking, were her pioneering efforts, beginning in the early 1960s, in the essentially

unheard-of area of infant hearing testing. An audiologist herself, Marion and a research partner started hearing testing for newborns before those infants had even left the hospital, fitting even the tiniest babies with hearing aids. Today, thanks to universal newborn hearing screening and early hearing detection and intervention programs, more than 90 percent of newborns have their hearing screened. Knowing what we know now about the importance of hearing with respect to language and even cognitive development in extremely young children, there’s no telling how many infants with hearing loss were identified as such in a timely manner, and their developmental skills saved, because of Marion Downs’s work. (To learn more about newborn screenings, see “This Is a Test,” page 10.)

The Marion Downs Center in Denver, Colorado, a nonprofit organization that espouses, as Marion did herself, a cradle-to-grave approach in dealing with hearing loss, will continue her efforts in advocating for those with hearing loss. Marion was a visionary in the world of hearing health. Her legacy lives on, quite visibly, in the children whose lives she touched. 

Senior Editor Amy Gross has been contributing to Hearing Health magazine since 2006.

Hearing Health Foundation was proud to have Marion P. Downs, Ph.D., as a member of its honorary board since 2009. Downs was a Distinguished Professor Emerita at the University of Colorado School of Medicine.



The Dangers of iPod Use

Listening to music at unsafe volumes—whether on a personal digital music player, at a concert, or even during band practice—for prolonged periods of time puts kids’ hearing at risk. Get the answers to common questions about hearing protection for children and teens.

By Nannette Nicholson, Ph.D., and Noelle Bonneannée



NOISE IS AN UNDERESTIMATED THREAT TO the health of children, adolescents, and young adults. The impact of noise is much like that of sun exposure to skin: The greater the cumulative exposure, the greater the impact. Excessive noise levels—and “noise” includes music—can cause a range of health problems, such as sleep disturbances, cardiovascular effects, poorer performance at school, hearing loss, and tinnitus (ringing in the ears).

The explosion in portable technology such as cell phones and music devices, like the Apple iPod, have brought about an increased use of earbuds and headphones, sometimes at unsafe listening volumes. As a result, noise-induced hearing loss (NIHL), especially among adolescents and young adults, has emerged as a public health concern.

A 2010 report in the *Journal of the American Medical Association* made waves when it found that documented hearing loss among adolescents ages 12 to 19 jumped by a third over about a dozen years. The study estimated that one in five teens has a hearing loss.

With the near-ubiquity of personal technology, the risk of NIHL in children and teens is real. Here are answers to common questions.

Are babies particularly sensitive to noise?

It is likely that your baby and young child’s hearing ability is better than yours. Human hearing begins to age

from the day you are born, just like your skin. In fact, by the time a person is 20 years old, the ability to hear the very high frequencies (8,000 to 20,000 Hertz, or Hz) has often been lost, due to the accumulated effects of lifetime noise exposure. Young children are often uncomfortable around noisy kitchen appliances, vacuum cleaners, and other loud sounds such as airplanes. Proactively remove your child from these environments, or invest in child-size hearing protection, such as infant earmuffs. (Learn more about hearing protection on the following page.)

Are some children more susceptible to NIHL than other children?

Current training material for healthcare providers prepared by the World Health Organization (WHO) says, “It is logical to consider certain subgroups of children (since conception) to be particularly at risk for harm from excess noise exposure.” These include fetuses small for their gestational age and very young infants born preterm and/or with low birth weight.

The training material adds that children who have learning disabilities or attention difficulties “may be more likely to develop early problems with mild hearing loss” from noise exposure compared to children without these challenges. Children on ototoxic medications (drugs that damage hearing), which include treatments for cancer or

severe bacterial infections, “may have a higher likelihood of developing problems from exposure to excess noise.”

The WHO says it remains unproven whether the developing cochlea in fetuses are more susceptible to environmental noise or vibrations in utero, and whether this may have been associated with premature birth or slowed development. But it does say infants and children generally are more vulnerable to noise because they may not recognize dangerous exposures, and they may not be able to identify and avoid the source of noxious noise.

Can NIHL be prevented?

Yes, it is the only type of hearing loss that can be prevented. Teach your children about the harmful effects of loud noise, including music, and encourage the use of earmuffs, earplugs, and volume-limiting headphones. Set a good example by protecting your own hearing in noisy situations, such as when using power tools.

How does loud noise damage hearing?

Excessive noise causes damage to sensitive structures in the inner ear. These sensory cells, known as hair cells, convert sound wave energy that reaches the eardrum into electrical nerve signals that travel to the brain. When hair cells are damaged or die, the result is permanent hearing loss. (Hearing Health Foundation’s Hearing Restoration Project has been taking the lead in investigating hair cell regeneration to cure hearing loss and tinnitus. See “Meet the DTR Mouse,” page 28.)

What do I need to know about gear for hearing protection for children?

Hearing protection, such as earmuffs, should be specially designed for children’s smaller heads and ears. It needs to fit well in order to work properly. It should also be able to withstand a child’s rough use. Built-in volume limiters in earbuds or headphones remove the guesswork about unsafe listening levels. The maximum volume should not exceed 85 dBA (A-weighted decibels, which measure how the human ear perceives sound). Limit use to two hours a day—even if the maximum is within the safe limit. If no built-in limiter is available, older children and teens can be taught to use a digital music player’s settings to set the volume to 75 percent of the maximum. (For a selection of kid-tough, teen-cool hearing protection, see “The 95-Decibel Birthday Party,” page 24.)

When should my children wear protection?

Hearing protection should always be worn in noisy environments where the average sound level measurement is 90 dBA or louder, such as at concerts, parades, and racetracks, and during activities such as target shooting,

hunting, and fireworks. Loud toys and video games are other potential sources for loud noises, as are sporting events, dance parties, and even band or orchestra practice.

How do I know when a sound is too loud?

You can download a sound level meter on your smartphone or tablet to measure the decibel level. After testing the accuracy of volume-measuring apps, researchers at the National Institute for Occupational Safety and Health cited NoiSee by Noise Lab (\$1), Noise Hunter by Inter•net2day (\$6), and SoundMeter by Faber Acoustical (\$20)—all available for Apple iOS—as providing the most accurate A-weighted sound level measurements. The report, in the *Journal of the Acoustical Society of America*, said the poor uniformity among Android devices and the lack of features and functionality of the Android-based apps precluded them from inclusion in the study.

What else can I do to prevent NIHL in children and teens?

A number of hearing awareness and education programs have been developed for children and their families. Some include curricula to use in elementary, middle, and high schools. Use these materials to advocate for safe hearing practices in school, sports, and social situations. Hearing Health Foundation, the publisher of this magazine, has partnered with the National Institute on Deafness and Other Communication Disorders on its “It’s a Noisy Planet” campaign (noisyplanet.nidcd.nih.gov), which includes fact sheets for parents and educators as well as online activities and quizzes aimed specifically at tweens (ages 10 to 12) and teens.

Learn more about NIHL at hhf.org/what-is-nihl. For references cited in this article, please see hhf.org/winter2015_references. 

Nannette Nicholson, Ph.D., is an associate professor and director of audiology in the Department of Audiology and Speech Pathology at the University of Arkansas for Medical Sciences in Little Rock. She has a joint faculty appointment at the University of Arkansas at Little Rock and a clinical staff appointment at Arkansas Children’s Hospital. She coauthored the chapter “Automated Hearing Screening” in the textbook “Auditory Electrophysiology: A Clinical Guide” (2012), edited by Samuel Atcherson, Ph.D., and Tina Stoodly, Ph.D.

Noelle Bonneannée is a graduate student in human development with an emphasis in social activism at Pacific Oaks University in Pasadena, California.



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This Is a Test

Actually, a number of tests. Get the lowdown on the simple and painless, though possibly ticklish, hearing screenings for infants and young children. *By Betty S. Tsai, M.D.*

One to three of every 1,000 infants are born with some degree of hearing loss, and even more children lose hearing later during childhood. The most critical period of speech and language development occurs during the first three years of life. Because hearing loss can limit a child's exposure to language, children with known hearing loss often have difficulty with the development of spoken as well as signed language. Children who receive intervention by age 6 months are often able to develop language skills on par with their peers.

As a result, many Early Hearing Detection and Intervention (EHDI) programs throughout the U.S. attempt to identify children with hearing loss by age 3 months. These programs work to identify children and infants with hearing loss and provide follow-up checkups and services for these children and their families. While every state has its own EHDI program, only 43 states at this time have mandatory newborn hearing screening.

Prior to the implementation of EHDI programs, the average age of a child identified with hearing loss was 2.5 years old. Since their implementation, the average age of diagnosis was just under age 4 months, with intervention by around 6 months of age, according to a 2005 study.

Newborn hearing screening takes place soon after birth, often prior to the discharge of the infant from the hospital. Screening can occur for babies with no birth complications as early as 6 hours after birth, although many centers will wait about 24 hours. Babies in the neonatal intensive care unit are screened just prior to discharge or when the newborn is in a stable condition in an open crib. Some neonatal babies may be screened multiple times throughout their hospitalization stay. For births that occur at home or birthing facilities, most states recommend that the screening occur within the first two weeks of life.

Babies who do not pass—or “refer,” another term for not passing—the initial screen are rescreened in the upcoming weeks. Those who continue to fail will be offered a full diagnostic test and may be referred to an audiologist for the full testing.

If a baby has been diagnosed with a hearing loss, a referral will be placed to an otolaryngologist (ear, nose, and throat physician) who has knowledge of pediatric hearing loss for a formal evaluation. The child may also be referred to a geneticist for a genetics consultation, since half of patients with congenital hearing loss have a genetic component, and to an ophthalmologist (eye doctor) for a formal vision screen. The infant may also be referred to an audiologist for hearing aid fittings.

Additionally, a formal speech-language evaluation may be performed followed by counseling about all communication options. Many states also have programs to assist families of infants with hearing loss as part of their early intervention services.

Depending on each state's and hospital's guidelines, the newborn hearing screening may be an otoacoustic emission (OAE) test or an auditory brainstem response (ABR) test, or a combination. The tests are safe and comfortable even for babies. (For details about these tests, in addition to other hearing screening tests for infants and children, see “Simple, Painless: Objective Hearing Tests,” page 12.)

While the OAE and ABR tests are complementary, some centers may prefer the use of one over the other or prefer to use both. OAE testing is more cost effective and takes less time but has a false positive detection rate between 5 and 21 percent within the first three days of life. On the other hand, ABR has a false positive rate of 4 percent. The increased false positive rate in OAEs has been attributed to the presence of amniotic fluid within

TEEN HEARING: DANGER AHEAD

Most teens are engaged in dangerous listening habits

NEARLY HALF

of teens showing potential signs of hearing loss

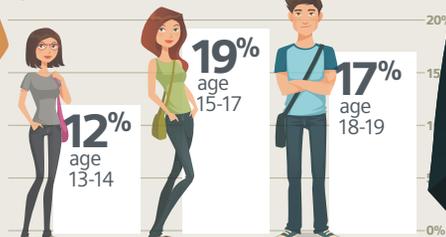
46%

(ringing, roaring, buzzing or pain)

1 in 6 teens have symptoms often or all the time

(about 5 students in the average classroom)

Breakdown of teens with at least one symptom, often or all the time



Risky Habits

Teens know there are risks and are still leaving their hearing unprotected

88%

Nearly 9 in 10 teens engage in at least one risky hearing behavior



81%

Listen to loud music with earphones



21%

use mowers & other loud tools



16%

use noisy powered toys

Signs of hearing loss in teens is a growing problem. By joining our Siemens Online Hearing Health Community you'll gain access to information, special offers, and eNewsletters about hearing health and products that can help. Join now to start receiving important news and offers at usa.siemens.com/didyouhear.

the ear canal in newborns, which disappears within the first couple of days of life. If there is concern about hearing loss in the child, both tests are used together to determine if a hearing loss exists.

Early detection with intervention of hearing loss has been shown to improve child developmental outcomes, especially in terms of speech and language development. Should there be any patient or provider concern, each

state has resources to help direct these children to the right places. Links to each state's information can be found at InfantHearing.org, the online resource center for EHDI.

Betty S. Tsai, M.D., is an assistant professor specializing in otology and neurotology in the Department of Otorhinolaryngology at the University of Oklahoma Health Sciences Center.

Simple, Painless: Objective Hearing Tests

By Samuel R. Atcherson, Ph.D.,
Sarah E. W. Kennett, Au.D.,
and Sarah Holland

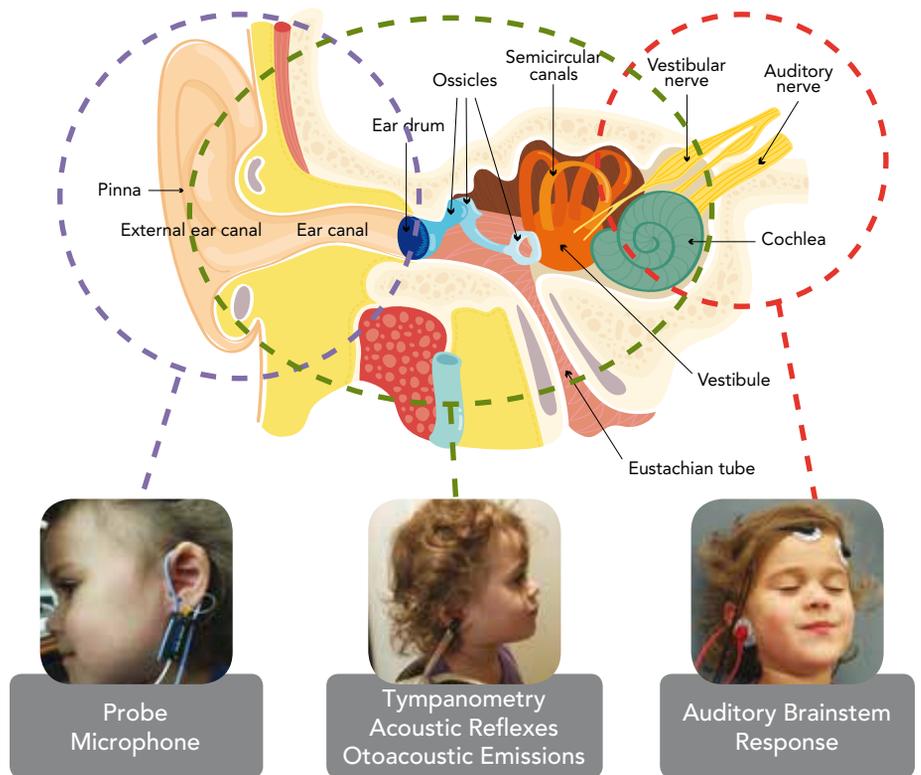
Objective hearing tests are those that do not require infants and young children to perform a task. Infants do not yet have the communication and motor skills to press a button in response to hearing a tone. Even in young children who do have the communication and motor skills, hearing tests can be so uninteresting or intimidating that it may take more than one audiologist and more than one appointment to obtain adequate information about their hearing status.

Some babies and young children may not like, or are tickled by, things placed in their ears. But since these simple, painless tests do not rely on the patient responding to instruction, they are invaluable for the information they provide about whether there is a hearing loss and whether hearing aids or an implant device may be helpful.

The tests are ordered as such below because they go from outer ear to the brainstem, which is also a logical testing order. The order also indicates the tests' importance.

Tympanometry

This test provides information about movement of the eardrum, the health of the middle ear, and the



The Auditory System

The ears that are on the outside of the head are only a small part of a larger, more complex system with many moving parts. The outer ear (above, left) collects sound waves and delivers them through the ear canal to the eardrum. Beyond the eardrum is the middle ear (above, center), which is an air-filled space and has three tiny bones that translate the movements of the eardrum into the waves in the cochlea. Located in the inner ear (above, right), the

cochlea is a snail-shaped structure with a ribbon lined with thousands of hair cells that respond to the sound-related movements of the middle ear. When the hair cells are excited, they cause the hearing nerve to send signals to the brain. The first entry into the brain is the brainstem. Within the brainstem are many nerve stations that continue to process sounds, which eventually make their way to the outer surface of our brain, called the cortex. It is within the cortex that we really "hear" sound.

Every child deserves the
best chance to learn



Oticon | Sensei

Study proves effectiveness of Oticon's advanced adaptive FM strategy, VoicePriority *i*TM



A recent study* by Erin Schafer, PhD, a leading expert in educational audiology and FM research, has validated the ability of VoicePriority *i*TM to support hearing in background noise. Her team showed that Oticon Sensei with VoicePriority *i*TM provides a significant increase in speech recognition in complex listening environments compared to traditional FM systems or a hearing instrument alone - particularly in localized noise.

Integrated into Oticon Sensei, VoicePriority *i*TM automatically adjusts the gain of the incoming FM signal to optimize the signal to noise level at the child's ear. This advanced adaptive FM strategy instantly responds to changing noise levels, ensuring consistent speech recognition wherever the child is located. And the more a child understands, the better her learning opportunities. It's as simple as that.

*) Peer-reviewed & accepted for publication in Journal of Educational Audiology, Schafer EC, Sanders K, Bryant D, Keeney K, & Baldus N (2013) Effects of Voice Priority in FM Systems for Children with Hearing Aids.

To learn more about Sensei, VoicePriority *i*TM and Dr. Schafer's study, contact your hearing care professional or Oticon Pediatrics at pediatrics@oticonusa.com

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PEDIATRICS

size of the ear canal. Tympanometry uses gentle air pressure in the ear canal to move the eardrum slowly from one position to another. As the eardrum is moved, a tone is played, and the energy of the tone within the ear canal is measured. The test result is plotted on a graph called a tympanogram.

Acoustic Reflexes

The acoustic reflex is a response from a small muscle in the middle ear, called the stapedius muscle. Controlled by the facial nerve, this muscle is connected to one of the three tiny middle ear bones and reacts to loud sounds from our own voice as well as to sounds presented through the ear canal. Sound must make its way from the ear canal to the cochlea, up the hearing nerve to the lower brainstem, and down the facial nerve to the stapedius muscle.

The goal of acoustic reflex testing is to find the softest sound at different pitches that causes this muscle to contract. Acoustic reflex testing uses the same equipment as tympanometry, but with some differences. During muscle contraction, the eardrum stiffness changes, and we are able to see the movement through pressure changes in the ear canal. These results can be helpful in giving a rough estimate of a child's hearing

status, since conductive (stemming from a problem in the outer and/or middle ear) hearing loss and severe hearing loss will most likely not have acoustic reflexes.

Otoacoustic Emissions

Otoacoustic emissions (OAE) are tiny sounds produced in the cochlea that make their way back to the ear canal. In order to record these sounds, a small speaker and microphone are placed gently into the ear canal. As sounds are delivered to the ear, the OAEs that come back are recorded and processed.

OAE tests provide some information about how healthy the ear's hair cells are, both the inner and outer hair cells. When there is wave movement in the cochlea, the outer hair cells will respond by trying to make the movements stronger, which cause the sound to be amplified in the cochlea. Because of all the moving parts, this amplified movement can make its way back into the ear canal. When outer hair cells are healthy, OAEs are recorded. OAEs are also recorded at several different pitches.

Auditory Brainstem Response

The auditory brainstem response (ABR) is a measurement of how the cochlea, hearing nerve, and brainstem respond to sound. It is tested while

the child is resting, sleeping, or sedated. Small surface electrodes are placed on the patient's head—two on the forehead and one behind each ear being tested. Those electrodes will be connected to a computer controlled by an audiologist. A small earphone is placed in the ear canal. Sounds are delivered through the earphone, or a headband, in the forms of clicks or short bursts of pitch-specific tones.

The electrodes pick up the nerves' responses to sound and display the readings as waves on the computer screen. The audiologist can then interpret the waves to conclude how the child hears, such as an estimation of how well the child can hear soft sounds at certain frequencies. An ABR test typically lasts 30 minutes to one hour or longer, depending on the type of information to be collected.

A related common newborn hearing test called an automatic auditory brainstem response, or AABR, takes only a few seconds. This screener will only tell the operator if the nerve responses are normal or if the child needs additional testing. If a child does not pass the AABR, an ABR may be scheduled to obtain more information about what the child is hearing. 



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Measuring How the Ear Canal Shapes Sound

Ears come in all shapes and sizes, which is why it is important that hearing aid probe microphone measures are used when programming a child's hearing aid. The shape of a child's ear canal can affect how things sound.

Using a tiny rubber microphone probe tube placed in the child's ear canal, an audiologist can measure how the ear canal shapes sound (i.e., the ear canal

response) so that the hearing aid is neither too loud nor too soft. After the probe tube is placed the child will look directly toward a speaker and hear a recorded passage or tones. (See page 12, photo on far left.)

The audiologist has several ways to fit hearing aids for infants and children. The hearing aid can be placed alongside the probe tube and the aid then adjusted to match the child's hearing loss. If the child cannot

5

Things I Wish I'd Known

The majority—more than 90 percent—of children born with hearing loss have parents who have typical hearing. It can be overwhelming receiving the diagnosis for your newborn, suddenly and without warning. Here, the parent of a newborn diagnosed with congenital hearing loss shares advice about the next steps, based on her experience with her daughter, who is now almost 4 years old.

By Nicole Kolodny

1 If your child has failed the newborn screening, contact an audiologist. Do not see an ENT (ear, nose, and throat doctor, also known as an otolaryngologist). Our experience is that ENTs perform the same test as the hospital and will not get you any definitive results.

2 Find an audiologist you feel comfortable with and whom you really trust. The first audiologist we saw, who ultimately gave us the same results as the audiologist we currently use, had no bedside manner. She didn't understand that we were first-time parents with a child who was just diagnosed with hearing loss.

3 Ask your audiologist for help with what you need to do next. He or she should be able to provide the names of doctors and schools that can help you. We would not have been able to get through step one of any process without all of the helpful advice from Blake's audiologist.

4 Be around people who are there to help. Don't let people tell you that the tests are wrong and that everything is going to be fine. Yes, everything will be fine, but not right away. Keep those people at arm's length. The people around you should be there to help, not pretend nothing is wrong.

5 You will not be okay with the diagnosis immediately, and there is no specific time frame for when you will be okay. Everyone deals with this differently, and some people need to experience an "aha" moment before acceptance. Mine was the day, a few months after Blake was diagnosed at age 1 month, when I looked at her and thought, "If Blake did not have a hearing loss, that would change a part of who she is, and I am in love with the person sitting in front of me, exactly the way she is."

Residents of New York City, Nicole Kolodny and her husband Michael Kolodny are founding members of HHF's National Junior Board.

sit long enough for programming, the audiologist may be able to obtain a quick ear canal response and use that response to program the hearing aid without the child being present. Or, if the child will not tolerate the probe tube, the audiologist can use estimated ear canal responses based on the age of the child. As the child grows and matures, further adjustments will be made based on one of these techniques.

Samuel R. Atcherson, Ph.D., is an associate professor in the Department of Audiology and Speech Pathology at the University of Arkansas at Little Rock/University of Arkansas for Medical Sciences. Sarah E.W. Kennett, Au.D., is a pediatric audiologist at Arkansas Children's Hospital. Sarah Holland is a second-year Doctor of Audiology student at the University of Arkansas for Medical Sciences.

On a Quest to Learn

Siblings, each fitted with bilateral hearing aids at age 3, have thrived in their local schools with the help of teachers, specialists, technology, and their desire to learn. *By Lisa N. Brownlie*

It's hard to believe that it has been three years since I wrote about my son and daughter—both born with bilateral moderate sensorineural hearing loss—for *Hearing Health* magazine.

My son Sean, who was fitted with his first hearing aids at age 3, is now 14 years old and a high school freshman at the progressive Quest to Learn (Q2L) Upper School in Manhattan. He graduated from Q2L Middle School with a 94 percent grade point average. Sean continues to work hard to meet the individualized education program (IEP) goals created around his congenital hearing loss and diagnosis of central auditory processing disorder (CAPD).

Sean is diligent about wearing his hearing aids in class now that he realizes just how crucial it is for him to hear and be connected in his school setting. He also continues to practice self-advocacy in class. He asks the teacher to repeat or further explain something that he does not fully understand. He asks plenty of questions and his teachers are happy to help in any way they can. Thankfully, they appreciate Sean's efforts.

His sister Samantha is 10 years old and in the fifth grade at P.S. 3 The Charrette School in Manhattan. In preschool, Sam's teacher noticed that Sam did not respond to the teacher's calls when her back was turned. She was also fitted for her first hearing aid around age 3. Today, Samantha also diligently wears her hearing aids and FM (frequency modulation) unit in class.

My husband Des and I decided that because both kids only had moderate hearing loss, it would be most appropriate if they attended local public schools. Our public schools provide Hearing Education Services (HES), as well as speech therapy, FM units, and other modifications

as needed. They have been very accommodating.

Sean's CAPD adds another set of challenges. He was diagnosed when he was 8 and in second grade. But Sean is hardworking and disciplined and thrives on structure in a classroom setting. Coincidentally or not, Sean has an amazing memory and is excelling in both Spanish and French.

As a high school student, he is able to ace tests and projects with less and less oversight by his parents or by his special ed teachers—"learning strategists" as they are called at Q2L. He is a kindhearted person, and people see that. Speech services and hearing education services are still mandated on Sean's IEP for reinforcement and extra help as needed.

As for Samantha, she is a bright, friendly, and participatory "alpha female." She also is holding her own quite well academically. Her book, "Samantha's Fun FM and Hearing Aid Book," continues to sell—find it on Amazon!—and provides people of all ages with clear and simple information about hearing aids and FM units.

Samantha wrote the book at age 6 and it explains how she and her teacher use the FM unit and how they work so that kids (and adults) can easily understand. Our public relations machine consists of my husband Des using social media to publicize the book.

Samantha's book received enough attention online that in 2012, CNN picked up her story and interviewed her on camera. She explained how and why she wrote the book and how it has helped people of all ages. She had previously won second place in Oticon's Focus on People Award in the children's category as a result of writing her book.

Both of our kids are staunch self-



"Both of our kids are staunch self-advocates," Brownlie says.

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advocates. They ask questions, ask people to repeat spoken requests, and ask for improved seating in class as necessary. Their self-advocacy enables them to reach their academic goals.

We connect with their teachers and special educators on a regular basis to work out any scholastic bumps in the road. It is very reassuring to be able to communicate with educators who care, respect, and

are willing to work with our children to help them achieve their personal bests. 

Lisa N. Brownlie lives with her family in New York City. Find her previous story about Sean and Samantha, "Passing It On," in the Fall 2011 issue at hearinghealthmag.com.



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Notes
from



Sean &
Samantha

"Don't be afraid to wear your hearing aids," advises Sean, with Samantha.

HI, MY NAME IS SAMANTHA.

HELLO, MY NAME IS SEAN.

I am 14 years old, I live in Manhattan, and I have enjoyed my school, Quest to Learn, for the three years I've been there. I was born with hearing loss, like my sister Samantha, and I wear hearing aids in both ears. I've worn them since I was little. When I wear my hearing aids, it makes the sound louder, and that way I can hear other people's voices loud and clear. Without them, I wouldn't really be able to hear much, but I might hear a few things. All of my friends and classmates in my school know that I wear hearing aids. I have been doing well in my classes with my hearing aids. I also have CAPD.

Some advice I'd like to give to those who are uncomfortable with wearing their hearing aids is: Don't be afraid to wear your hearing aids when you need them. I have experienced this myself. I was afraid to wear my hearing aids at one time because I felt that people would make fun of me. But now I feel confident in wearing them so I don't feel left out and will always be connected to others, and you should feel confident as well.

I am 10 years old. I wrote my book about hearing aids when I was 6 years old, and I had the help of my hearing teacher Karen, who is now retired. At P.S. 3 I have a funny and awesome teacher named Alan and he is always singing or making funny jokes in class. I am now writing a book about the life of a 13-year-old girl. I am not really done with it yet, but it's coming along.

My FM system is way more different than last year's FM system. Instead of wearing it as a necklace, it clips onto my teacher's belt buckle. In school I am on level X in reading which means I only have two more levels till I get to level Z.

I use my hearing aids to hear better. My brother Sean wears hearing aids as well, and we both have them in both ears. Last year my hearing teacher helped me learn about the inner and middle and outer part of the ears, and I know about how I got hearing loss. My inner ear has a kind of snail-like blue part that has hair-shaped cells and liquid inside, and in my snail-like thing I somehow lost a few hair-shaped cells, and that's basically how I got hearing loss.

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FM systems are the go-to devices to benefit children with hearing loss in the classroom. Simple to use, unobtrusive, and affordable, they make use of existing technology wired into the child's hearing aid. (See "The 'FYI' on FM Systems," page 32.)

FrontRow's Juno system leverages the FM system to provide playback of classroom lessons. At the push of a button, the audio system records the teacher speaking, integrates multimedia and any student responses or comments, and packages it all together in a single audio-video stream that can be accessed online. This allows a child with hearing loss to review classroom lessons.

Regardless of whether a child



Redcat by Lightspeed subtly amplifies the teacher's voice.

natural voice throughout the classroom, without the need to raise their voice or even face the students. The wireless, installation-free system makes use of the existing sound-reflecting properties of the walls, floor, and ceiling.

has a hearing loss, classroom technology can help students hear better and, as a result, focus better. Lightspeed's Redcat audio tool is comprised of a small (16-by-9-inch) flat screen, which sits unobtrusively on top of a bookshelf or a table in a corner of the classroom, and a small microphone that the teacher wears around the neck.

The technology subtly amplifies and evenly distributes the teacher's

"To me it's a win-win to give every child an advantage," says Margaret Leining, Ed.D., the principal of Travell Elementary School in Ridgewood, New Jersey, who experienced the system in use in a neighboring school district and began installing the systems in classrooms in 2011. "Anything that gives students a way to improve their access to learning is worthwhile."

—Yishane Lee



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Proof That Everything Will Be Okay

After every setback, a father whose daughter has Usher syndrome comes to this realization, over and over.

By Mark Dunning

WHEN MY DAUGHTER BELLA was 8 months old, she held her thumb and her forefinger up to her bald head and sent a toothless smile in my direction. She was either signing “daddy” or “loser,” but either way she meant me. I can’t overstate the relief I felt. My deaf daughter and I were going to be able to communicate. I was going to be able to tell her I loved her, and she was going to understand. Her hearing loss had been a shock and had filled me with such dread and fear. Here, finally, was proof that everything was going to be okay.

When Bella was 3 years old we were in the yard, and she began to look around with a mixture of surprise and consternation. She pointed at the sky and signed “bird.” A year earlier she had gotten a cochlear implant. This was the first time she clearly heard a sound

and associated it with something. I had feared the surgery and therapy were an empty ordeal. Here, finally, was proof that everything was going to be okay.

When Bella was 4, my wife Julia and I founded the Decibels Foundation to provide specialized early intervention, educational services, family support, and access to essential technologies for children with hearing loss, from infancy through high school. As we had learned through many hours of work, kids with hearing loss can live normal, happy, productive lives with the right support. Through Decibels we could help other families to know that everything was going to be okay.

When Bella was 8, she was diagnosed with Usher syndrome. Suddenly those countless hours adjusting and adapting, studying and practicing, and accepting and incorporating hearing loss into our lives meant nothing. My daughter—my beautiful, wonderful daughter—had Usher syndrome. She was going to go blind.

Usher syndrome is the most common cause of combined deafness and blindness. It is classified as a rare disease but it’s not as rare as many think, at least not among those with hearing loss. Upward of 10 percent of people with congenital, bilateral, sensorineural hearing loss have Usher syndrome. This translates to

about 45,000 people in the United States and 400,000 worldwide, more than ALS (Lou Gehrig’s disease), more than Huntington’s disease, and more than Duchenne muscular dystrophy.

When Bella was 10, Julia and I founded the Usher Syndrome Coalition to raise awareness about Usher syndrome as the most common cause of combined deafness and blindness, to accelerate research, and to provide support to individuals and families affected by the condition. In July 2014 we held the International Symposium for Usher Syndrome at Harvard Medical School in Boston. It was the largest gathering of the Usher syndrome community in history.

Standing before more than 300 researchers and families at the symposium, Bella, then 15, spoke clearly, confidently, and passionately about how much Usher syndrome had improved her life. She spoke about the dear friends she had made through the Usher Syndrome Family Network and acknowledged the hope in the many treatments in development. “Thank you all for coming,” she concluded. “My life is great because of you.”

The auditorium rose in a standing ovation. All the hope generated by the symposium and the pride in my daughter hit me like the tide. I can’t overstate the relief. Here, finally, was proof that everything was going to be okay. 🦻

Mark Dunning and his family live in Massachusetts. To learn more, see DecibelsFoundation.org and Usher-Syndrome.org.

CHILDREN & HEARING LOSS ISSUE



The author with his daughter Bella.

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The 95-Decibel Birthday Party

All of us, including children, increasingly live in a noisy world. Here's what you need to know about kid-tough, teen-cool gear to protect their hearing.

By Yishane Lee

Can having fun harm your hearing? The answer to that question appears to be yes. The volume level at popular recreational activities such as going to the movies, playing video games, or even attending a birthday party can be loud enough to cause permanent hearing damage and tinnitus (ringing in the ears), especially if the sound is repeated for a long time or at a very close range.

The upper safe limit for hearing is 85 decibels (dB)—about the sound of traffic. But the way decibels are measured is on orders of magnitude. For example, volume that is 95 dB (like at a DJ dance party) is twice as loud as 85 dB, but 105 dB (the sound emitted by some toys) is four times as loud as 85 dB. Explosions and other loud scenes in movies can measure 115 to 120 dB.

According to a 2010 study published in the *Journal of the American Medical Association*, one in five adolescents between the ages of 12 and 19 shows a hearing loss. Population-based, cross-sectional data collected from 1988 to 1994 and from 2005 to 2006 showed the rate of hearing loss among adolescents had jumped 33 percent.

While most of the hearing loss was slight, the study was a wakeup call, not only because of the increased prevalence, but also because the hearing loss was unilateral and in the high frequencies. This can indicate noise-induced

hearing loss (NIHL) stemming from the use of earbuds or headphones at unsafe volumes.

NIHL is the only type of hearing loss that can be prevented. Limiting exposure is your first line of defense, as is using protective gear. But there are challenges. Can the hearing protection for younger children take a beating? Will your teens think it's cool enough to use every time they need to? No matter which product you choose, remember that hearing protection must fit properly and be comfortable in order to be effective—and worn consistently in noisy situations. Here are our picks.

Volume-Limiting Headphones and Earbuds

Ear gear with a built-in, volume-limiting feature can help you make sure children and teens are listening safely. Some limiters are “always on,” and some use a switch. With exceptions noted below, all of the ear gear listed in this section tops out at 85 dB.



Califone SoundAlert Headphones (\$86, califone.com) use an LED light to indicate when the volume goes above the safe limit—and then it's up to you or your child to turn it down. These institutional-



grade headphones sport rugged plastic and rubber parts, an adjustable headband, replaceable padded ear cups that fit completely over the ear, and an extra-long, six-foot cable and reinforced jack.

Griffin Technology Crayola MyPhones (\$25, griffintechnology.com) have an always-on volume limiter, an adjustable headband, and padded ear cups that can be customized with stickers and Crayola markers (included). **Griffin KaZoo MyPhones** (\$20) have the same fit and technology but come in two animal designs: a frog with a tadpole jack and a penguin with a fish jack.

Etymotic Ety-Kids5 Earphones (\$40, etymotic.com) are high-fidelity earbuds that come with three eartip sizes so you can be sure that there's always a good fit—and seal—when using. The volume limiter is always on, and the cable is reinforced with Kevlar.

Fuhu Nabi Headphones (\$70, nabi.com) have ear cups that light up when the 80 dB volume-limiting switch—hidden in one of the cups—is flipped on. The cushioned ear cups can be customized with cartoon characters, the cord is replaceable, and the adjustable headband fits adult-size noggins so you can use these headphones, too. (We suggest keeping the volume limiter turned on.)

JLab JBuddies Folding Headphones (\$50, jlabaudio.com) fold down using hinges that won't pinch little fingers. The volume limiter is always on, and the padded ear cups can be decorated with 3-D stickers.

The **Kidz Gear KidzControl** line (gearforkidz.com) comes in a wired (\$20) and wireless version (\$30) and has a separate volume control cord that sets volume at a maximum of 80 percent of its 108 dB max—or about 86 dB. If you happen to lose the detachable cable that turns it on, Kidz Gear will replace it for free.

MarBlue Headfoams (\$30, thegrommet.com), for toddlers to teens, have an always-on volume limiter and

are designed to be indestructible. Covered with tough EVA foam, they have a cord and padded ear cups that are nonremovable, to prevent breakage and loss. (See photo, opposite page, bottom.)

The **SMS Audio KidzSafe** line (smsaccessgranted.com), featuring always-on volume limiting, includes **MyDesign D.I.Y. Headphones** (\$30) and **MyDesign D.I.Y. Earbuds** (\$20), both of which can be personalized with removable stickers. Two sets of snap-on padded ear cups are included with the headphones, along with two removable five-foot cords. With the earbuds, you get three pairs of silicone eartips to help ensure a perfect fit.

Protective Earmuffs



In 2010, pro football player Drew Brees put children's hearing protection on the map when his 1-year-old son Baylen was shown wearing Peltor earmuffs on the football field immediately after Brees and his team, the New Orleans Saints, won the Super Bowl.

Professional sports are notoriously loud events. The dubious achievement of a Guinness World Record for the loudest crowd roar at a sports stadium goes to Kansas City Chiefs fans, at over 142 dB. This is a scary number, because at 150 dB—equivalent to hearing a plane take off from 80 feet—rupturing an eardrum is likely.

Noise protection products often list an NRR, or noise reduction rating (NRR). “Confusingly, the NRR is actually unrelated to the actual attenuation levels,” says Melissa Heche, Au.D., of New York Speech and Hearing in New York City. So a product listed with “NRR 12” can still promise a 20 dB reduction, when used properly. The NRR—achieved using a standard formula to measure

potentially achievable protection—is helpful as a rough guideline for how many decibels are being reduced.

3M Peltor Kid Earmuffs (\$19, solutions.3m.com) have soft, padded ear cups and a slim, low-profile headband that helps make sure the muffs stay on, comfortably. The ear cups protect but allow air circulation over the ear, to help reduce the buildup of moisture and heat. Adjustable to fit babies up through adolescents, the muffs have an NRR of 22 dB.

Em’s 4 Bubs Baby Earmuffs (\$31, earmuffsforkids.com) are specially sized for infants ages newborn to 18 months and have an NRR of 26 dB. These patented earmuffs use an adjustable fabric headband and Velcro to keep ear cups in place, which is gentler on a baby’s head. (See previous page for a photo.) The company also makes adjustable earmuffs for ages 6 months and up, **Em’s 4 Kids** (\$26).

My-T-Muffs (\$16, earplugstore.com), with an NRR of 19 dB, have padded adjustable headbands and soft vinyl ear cups. Foldable for easy transport, they fit babies through teens.

Pro Ears ReVO Earmuffs (\$80, proears.com) are made by a company that specializes in hearing protection for recreational shooters. These rugged, child-size earmuffs

have an NRR of 25 dB and come with an adjustable headband and padded leather ear cushions.

Silenta Kid Earmuffs (\$29, silenta.com), from a Finnish hearing protection company, are foldable with a reflective headband and fluorescent, padded ear cups. With an NRR of 21 dB, they are sized for infants and very young children.

Tasco Kidsafe Hearing Protector Earmuffs (\$17, tascocorp.com) have an NRR of 25 dB. The ear cups are padded and deep for extra comfort, and the padded headband has a low clamping force and evenly distributed pressure points.

Earplugs

When kids get older and can be counted on to keep track of their own things (and when small parts won’t pose a choking hazard), earplugs for sports or concerts are a must. The models with flanges limit the volume while still allowing for clear hearing of music or speech.

Generally speaking, they have a lower NRR than earplugs



Smartphone Smarts



If you have to shout to be heard, that’s an indication you are in a loud environment. But if you want to know exactly how loud it is, download a decibel-measuring app for your smartphone or tablet. While they are not substitutes for a professional sound meter, they will give you a general idea as to when you need hearing protection.

Always keep in mind that distance from the noise and duration of the noise are just as important as decibel levels. Avoid sounds that are too loud, too close, or that last too long.

Andrew Smith’s AudioTools (\$20) is the closest app to a professional-grade audio and acoustic analysis tool. For one thing, it measures sound as perceived by the human ear, the A-weighted decibel (dBA), and not just the decibel level. According to Hearing Health staff

writer Kathi Mestayer, this app, made for the Apple iPhone or iPad, gets high marks from acousticians she has interviewed.

Skypaw’s Decibel 10th (free) is a basic app that will give you a sense of how loud your surroundings are. Available for Apple or Android devices, the app compares the current decibel reading to a common sound environment—e.g., a reading of 50 dB is an “average quiet street”—and includes the peak level for the current session. You can tap to send the data via email. Because of these features, among the many free decibel apps sampled, this is the one we liked the best.

(The National Institute for Occupational Safety and Health tested nearly 200 decibel-measuring apps and rated three the most accurate. See page 9.)

As smartphone use has expanded, so have smart accessories. Here are two new products to check out.

Fuse Chicken’s Bobine (\$35, fusechicken.com) is a cable, dock, tripod, and stand all in one. Made of a sturdy but lightweight and bendable cable cord, it charges your Apple iPhone or Android phone via a USB port. Bend the cord to place your phone at hands-free eye level for video calls or to read recipes while cooking, or set it up as a tripod.

Nomad’s NomadPlus (\$39, hellonomad.com) is a unique battery charger that literally piggybacks on Apple’s square wall charger. Place NomadPlus over the Apple charger and charge both your device and the battery at the same time. When it is fully charged, NomadPlus provides an on-the-go, 100 percent charge to your Apple iPhone. —Y.L.

that are made from foam or silicone, whose more solid construction reduces additional decibels.

Another thing to look for is “flatter attenuation.” This means sound is reduced equally across frequencies. “Most earplugs reduce high frequencies more than low, resulting in a muffled sound quality,” says Joscelyn Martin, Au.D., an instructor in audiology at the Mayo Clinic in Minnesota.

Model healthy hearing behavior for your children by using earplugs when mowing the lawn, attending a parade, watching a sports event, and in other noisy situations. Prices listed here are for one pair, unless noted.

Etymotic Ety Plugs ER20 (\$13, etymotic.com) come in two sizes, with the standard size working for average and smaller ears. Etymotic says these reusable earplugs, used correctly, reduce sound by 20 dB across all frequencies (the NRR is 12 dB), making music and speech quieter and safer but still clear. (See opposite page for a photo.)

Hearos Pearls Silicone Earplugs (\$30 for five pairs, hearos.com) are “pre-rolled” silicone balls that allow you to easily, quickly, and comfortably position them over the ear canal to protect from noise (and water, if needed). With an NRR of 21 dB, they come in a reduced size for

children and smaller adults, as well as a standard size.

Killnoise Sound Plugs (\$9, us.killnoise.com) are a stylish answer to the question of how to use earplugs at concerts and still look cool. The minimalist, four-level design in flexible, translucent plastic delivers an NRR of 18 dB in the small size while maintaining fidelity and clarity. The earplugs are reusable, and the slim carrying case available in more than a dozen colors easily fits into a pocket or purse. (See photo, pages 24 and 25.)

Mack’s Hear Plugs (\$12, macksearplugs.com) are washable and reusable and have an NRR of 12 dB. An inconspicuous, low-profile design and two comfort-tip sizes encourages consistent use during band practice, concerts, nightclubs, sporting events, and other noisy recreational events.

Westone Audio Universal TRU Earplugs (\$30, westoneaudio.com), with an NRR of 13 dB, use advanced acoustic filter technology and are designed for recreational use at concerts or clubs. The universal fit should work for average-size ears. 



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Meet the DTR Mouse



The goal of the Hearing Restoration Project (HRP) is to define drugs or other therapies that can restore hearing in humans. Most hearing loss is due to the loss of the tiny cells in the inner ear that change mechanical stimulation of the fluids in the cochlea into electrical signals that can be transmitted to the brain; the mechanoreceptive cells are called hair cells and are absolutely necessary for hearing and balance.

Unlike other vertebrates such as fish, birds, and reptiles, mammals (including humans) do not spontaneously regenerate these hair cells after they die or have been damaged. For the past two years, the HRP's efforts have been discovery-oriented and aimed at identifying genes, or pathways, that control hair cell regeneration.

As the HRP progresses, we hope to have pathways and then candidate drugs to test for their efficacy in restoring hair cell numbers and auditory function. At that time it will instantly become essential to have well-documented and characterized animal models for creating controlled hair cell damage and hair cell loss in mature subjects. These animal models will allow us to effectively, efficiently, and systematically evaluate candidate pathways, find or create new drugs, and take the next steps toward creating a cure for hearing loss and tinnitus.

The most advantageous experimental system would enable complete, selective, and rapid loss of hair cells at any age in a reproducible fashion. With this approach, evidence for new hair cell production will be inherent in the re-emergence of hair cell profiles over time.

Existing Methods

Other methods for inducing hair cell loss in mice exist, but there are limitations.

Noise: Until recently, very little work on the relationship between hair cell loss and noise exposure has used mice. However, recent systematic studies convincingly demonstrate that reliable loss of hair cells only occurs in conditions when the noise levels are loud enough to induce cellular damage elsewhere in the cochlea. In such cases, we cannot know if the hair cell damage is the primary

result of the noise or a secondary result of the surrounding cellular damage. Furthermore, hair cell loss tends to be highly variable, and the collateral damage from noise may disrupt the ability of supporting cells to produce new hair cells.

Finally, noise damage to hair cells is extremely nonlinear. Even small variations of a mouse's position in its enclosure can make large differences on the pattern of hearing loss. This makes standardization across labs very, very difficult.

Aminoglycosides (AGs): Many scientists in the hearing field have studied the death of inner hair cells due to antibiotic therapy. This is because an unfortunate side effect of the first, most powerful class of antibiotics, gram-negative AGs, is that they kill hair cells, causing hearing and balance disorders. But at high doses, they also cause rapid damage to the stria vascularis and ganglion cells as well as to hair cells. Therefore damage to these other cochlear components will render attempts to attribute recovery to new hair cells difficult. In other words, AGs are relatively nonspecific in the cells they affect. These issues will make quantitative studies of hair cell regeneration using mice whose hearing loss was caused by AGs a serious challenge.

A more immediate problem is the variability of results using AGs to kill hair cells. HRP investigators Elizabeth Oesterle, Ph.D., and Yehoash Raphael, Ph.D., together with Andrew Forge, Ph.D., have produced large lesions in mature mice in vivo using AGs coupled with diuretics. The variability seen with these treatments and with AGs alone is very large. Inner hair cells seemed to survive—or die more slowly and more variably. Oesterle has stressed the inconsistency between animals and within the cochlea of single animals.

A New Mouse Model

During the past 30-plus years, my laboratory has worked on trying to understand how hearing loss that is first evident at different ages influences development of the structure and function of brain regions that are

A designer laboratory mouse—whose hearing loss can be induced in a stable, reproducible way—will become critical when regeneration therapies are eventually tested.

By Edwin W Rubel, Ph.D.

critical for hearing. In 1986–87 my laboratory and the group led by Douglas Cotanche, Ph.D., discovered hair cell regeneration in birds—initiating a new field of research with immense potential clinical importance. Clearly, we needed a new mammalian model to study both topics.

With this in mind, we teamed up with Richard Palmiter, Ph.D., a pioneer in the production of biologically engineered mice. We developed a mouse line that has had the human diphtheria toxin receptor inserted into the gene, *Pou4f3*, that is exclusively expressed in the hair cells in the inner ear. While some other cells in the body express this gene, no other cells in the inner ear do so.

Because the human diphtheria toxin receptor is several thousand times more sensitive than the normal mouse receptor, this created a mouse model in which hair cells can be rapidly, selectively, and completely eliminated at any age by injecting tiny amounts of diphtheria toxin into the mouse. Although the use of mice expressing parts of the diphtheria toxin (DT) gene or the human diphtheria toxin receptor (DTR) in specific cell types has been available for about 25 years, this appears to be a unique model for hearing research in its versatility and specificity.

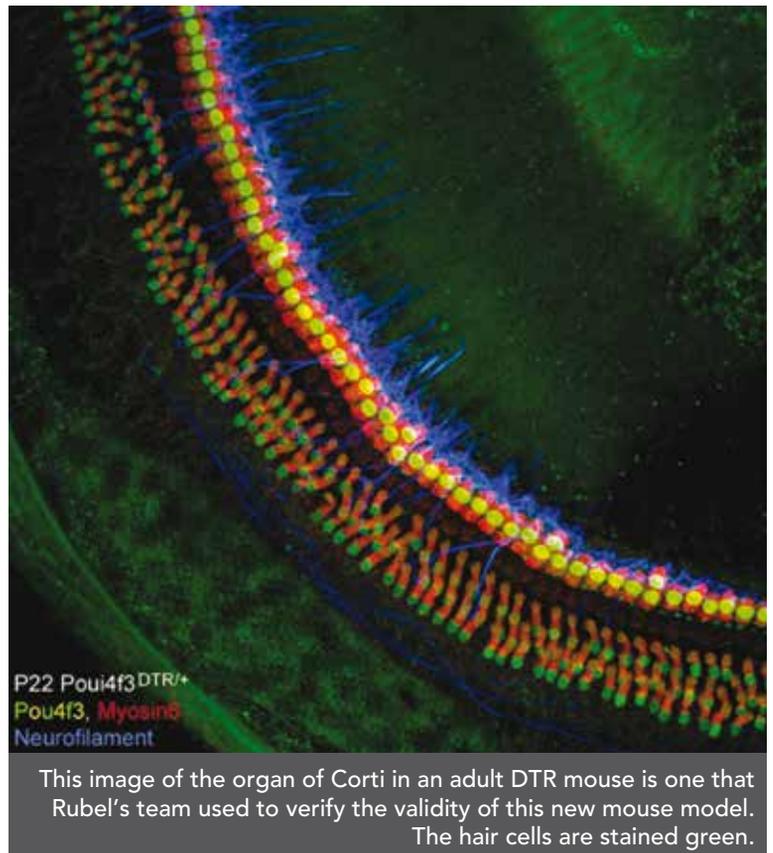
Mice with both copies of the transformed *Pou4f3* gene are deaf because all of their hair cells degenerate shortly after they are formed. On the other hand, mice with one copy—which we refer to as *Pou4f3*^{DTR/+} or DTR mice—have normal hair cell numbers and normal hearing, and seem otherwise perfectly healthy until they are given a single, very small injection with diphtheria toxin. Then, within three to five days the mice become totally deaf, and all of the hair cells in the cochlea are gone soon after.

We have been thoroughly characterizing this DTR mouse model to collect all of the information needed for studying hair cell regeneration. This includes gathering detailed DT dosage information such as the timing of hearing loss and hair cell loss

at different ages and different DT dosages; collecting data on exactly what happens to each kind of supporting cell and other components of the cochlea and vestibular organs, such as the stria vascularis and ganglion cells, at specific times ranging from a few days to several months; and identifying the parameters for using this model for in vitro (tissue culture and organ culture) studies.

So far we have determined that in adult mice, supporting cells in the organ of Corti of the cochlea appear to survive intact, as do most of the ganglion cells. This is very good news since the supporting cells are thought to be the potential source of regenerated hair cells, and the ganglion cells are necessary for connecting these cells to the brain.

We are continuing these studies with the help of funds from the HRP and the Hamilton H. Kellogg



and Mildred H. Kellogg Charitable Trust.

Several HRP labs are now using this model for in vivo and in vitro studies to screen for and analyze pathways for hair cell regeneration. Using the same well-characterized model across different laboratories and across different kinds of experiments optimizes the transition from an effective in vitro experiment to determining how that treatment behaves, and whether it is effective, in a live animal, as well as improving the transition from basic, discovery science to translational research.

While I may not be aware of the unpublished details on other models, it appears to me that the DTR mouse is the best live model we have at this time for studies attempting to initiate hair cell regeneration in the mammalian inner ear. 

Edwin W Rubel, Ph.D., is the Virginia Merrill Bloedel Professor of Hearing Science at the University of Washington in Seattle, where he is also a professor in the Departments of Otolaryngology, and Physiology and Biophysics.

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CHILDREN & HEARING LOSS ISSUE

The 'FYI' on FM Systems

By George Khal

Ear-level FM systems are dispensed only by audiologists because they must match the electronics of the user's hearing aid. They are especially useful in classrooms but also work at home for any listening situation. A microphone is plugged into the transmitter and worn by the speaker. The receiver in this case is a tiny "boot" plugged into the bottom of a behind-the-ear hearing aid. Sound is transmitted through the hearing aid, already optimized to the user's hearing loss. The range is about 50 feet and the cost, which is not normally covered by insurance, is about \$2,500.

Large-area FM systems use an electric transmitter and transmit the audio from the sound system of the establishment to an unlimited number of receivers in an auditorium, playhouse, theater, or place of worship. The transmission range is 500 to 2,000 feet and the cost ranges from \$900 to \$3,000.

Tour guide/courtroom FM systems operate much like a personal FM system but use 10 to 30 receivers to accommodate many listeners at the same time. The tour guide systems come with a boom microphone worn on the head. The transmission range is 100 to 150 feet and prices start at \$2,000.

These FM systems, including the ones for institutional use, can usually be purchased from assistive listening device vendors such as Harris Communications (harriscomm.com) and Teltex (teltex.com). 

George Khal founded Sound Clarity, an international retail company for assistive devices, serving as its president from 2000 to 2010. He has a severe bilateral hearing loss and was the Iowa chapter coordinator for the Hearing Loss Association of America.

Frequency modulation (FM) systems are electronic devices that broadcast sound on special FM frequencies. They work much the same way music is transmitted via an FM radio station, but the frequencies used in FM systems are outside the range of FM radio. People with and without hearing loss can use them to hear better.

With a transmitting range of between 60 and 2,000 feet, FM systems help a person with hearing loss hear better in a variety of situations, such as in the classroom, during meetings, in tour groups, when watching TV, or during conversations in noisy settings.

FM systems transmit the sound directly to the person's hearing aid, neck loop, or headphones, which eliminates the problem of background noise. Depending on the model and type, FM systems generally consist of two battery-powered parts: the transmitter, used by the speaker, and the receiver, used by the listener.

There are several types of FM systems. The first two detailed below are for personal use, and portable. The other two you may encounter in public settings. If you are wondering how FM systems differ from wireless Bluetooth technology, the main difference is the range. The range for Bluetooth is 20 to 30 feet, much shorter than FM systems, whose minimum range is 60 feet. However, with the use of streamers, Bluetooth

has become the standard for pairing hearing aids to your cell phone and other personal electronics.

Personal FM systems consist of a transmitter and receiver, both the size of a deck of cards. The transmitter is used with either a lapel microphone, a boom headset microphone for noisy situations, or an omnidirectional microphone placed on a table for a small group meeting. The receiver can be used with a neck loop, headphones, or earphones. Both transmitter and receiver are worn on the body using a supplied clip. The system is very versatile because it can be used for conversations in the car, around the dinner table, while watching TV, and in the classroom. And over the past five years, built-in microphones in the receiver allow the listener to hear sounds that are close by.

Many university student disability services loan out personal FM systems to students for classroom use. The latest models now transmit high-definition digital sound and transmit on the 2.4 GHz global frequency standard. Some also use secure digital channels for communication in places like courtrooms. Some models use the 72–76 MHz frequencies that are protected by the Federal Communications Commission for hearing assistance use. The transmission range is 90 to 150 feet and the cost is \$700 to \$950, depending on features and accessories.

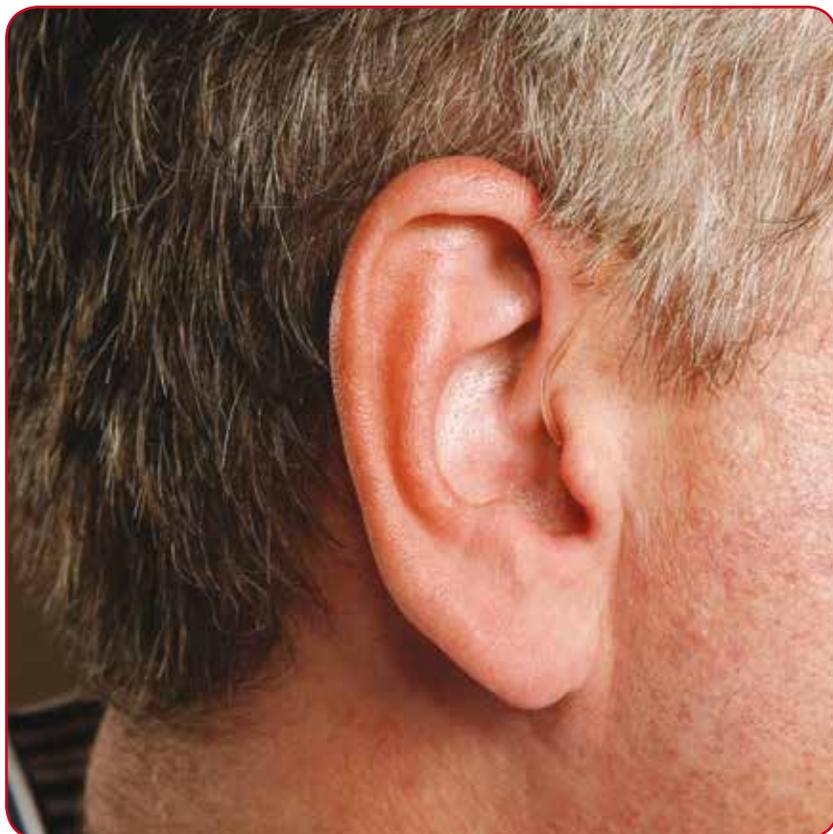


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What to Consider When Choosing Hearing Aids for Kids



By Courtney M. Campbell, Au.D.

Learning that your child has been diagnosed with a hearing loss, no matter the severity, can be overwhelming. But early detection and treatment of the hearing loss is extremely important. Hearing loss in children can affect a child's ability to develop appropriate communication, language, and social skills. The earlier the intervention, the more likely your child will thrive.

For most congenital hearing losses, the treatment is hearing aids. It is amazing that nowadays children as young as 4 weeks old can be fit with hearing aids.

There are some factors to consider when a child needs to be fit with a hearing aid. One major consideration is the style of hearing aid. All children, no matter the severity of their hearing loss, should wear a behind-the-ear (BTE) style. The primary reason for this is because as a child grows, their ears will change shape rapidly. Having BTEs with custom earmolds that can be remade as the ears grow and change will ensure a consistent, comfortable fit for the child, without having to get an entirely new hearing aid.

These earmolds tend to be made of soft, hypoallergenic materials that are safe for tiny ears. Keep in mind that these earmolds will need to be created multiple times throughout the child's life at regularly scheduled intervals (to be determined by the fitting audiologist).

The BTE style, being larger than

other styles, is easy for the parent or caregiver as well as the child to handle and insert into the ear. BTEs are also usually very durable, and they can be programmed to accommodate a wide range of hearing losses, from very mild to profound.

In addition, BTEs are easily connected to assistive listening devices. ALDs are used when (as commonly occurs) a child who wears hearing aids still has difficulty hearing and understanding a speaker's words in background noise or when there is a great distance between the child and the speaker. The FM (frequency modulation) system is an ALD that works well in such situations, like in a noisy classroom.

An FM system is comprised of a microphone, receiver, and transmitter. The speaker wears the microphone; the receiver is attached to the child's hearing aid; and the transmitter sends the speech signal directly from the microphone into the child's hearing aid for a consistent, audible, and understandable input regardless of classroom position or background noise. (For more on FM systems, see page 32.)

Finally, it is crucial that the child wears the hearing aid consistently. This means every day, and during all waking hours. This is especially true when it comes to young children who are acquiring language skills.

Certain accessories can help keep

CHILDREN & HEARING LOSS ISSUE

the hearing aids in place for young, active children, such as double-sided tape and lightweight hearing aid clips that connect the hearing aids to the child's clothing. Many audiologists who work with children recommend Huggie Aids, which are lightweight hearing aid retainers made of flexible, clear plastic that help keep hearing aids from falling out during sports or other activities.

To promote consistent hearing aid usage, encourage your child to make their hearing aids their own. Pediatric hearing aids come in an array of colors, and earmolds can be customized in various colors and patterns as well. My hearing aids are my favorite color, pink—bright colors also make them easier to find!

There are also some companies that make "jewels" and stickers just for hearing aids, which is another great way to help your child personalize their hearing aids. When you treat your child's hearing aids like cool, high-tech devices that are instrumental for their hearing, and when you celebrate wearing—instead of hiding—them, your child will use and benefit from them. 

An audiologist at A&A Hearing Group in Chevy Chase, Md., Courtney M. Campbell, Au.D., received her undergraduate and doctorate degrees from the University at Buffalo in New York. She has a hearing loss and has been wearing hearing aids for over a decade.

Music for Our Cause

Earth, Wind & Fire's Ralph Johnson and singer-songwriter Siedah Garrett, who recently collaborated on a holiday single, share what helping Hearing Health Foundation means to them.

Percussionist Ralph Johnson is a founding member of the legendary band Earth, Wind & Fire. He says:

“My entire career, including 43 years with Earth, Wind & Fire, is centered around my hearing, so supporting Hearing Health Foundation is a natural fit. There is some hearing loss in my left ear, but it in no way impairs my ability to perform, either personally or professionally.

Here's a news flash: Louder is not better; louder is more distorted. You need high power for quality listening, not high volume.

My collaboration with the incredible Siedah Garrett was rather serendipitous. We met in 2013 while Earth, Wind & Fire was appearing at the Hollywood Bowl in Los Angeles. Later, I needed someone to help complete a track and reached out to her. Siedah loved what she heard, so together we recorded the single 'Have a Very Merry Christmas.' I'm very proud of the work we've done together and look forward to more creative moments.

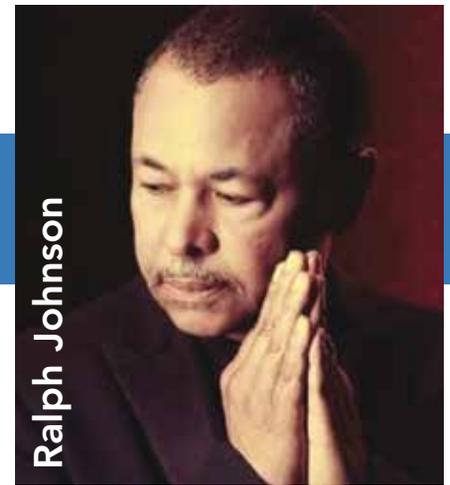
To those who are dealing with hearing loss, my advice is to become educated about what you're dealing with, be proactive, and know that you are not alone. ”

Siedah Garrett has written songs for and performed with Michael Jackson, Madonna, Quincy Jones, and many others. She says:

“I was fortunate to have cowritten Earth, Wind & Fire's single 'My Promise' from their 2013 album 'Then, Now & Forever.' Ralph contacted me about a music track and chorus that needed a melody and lyrics for the verses. Ralph felt so good about the song's potential that he decided to release it as a holiday single under his name, and I could not have been more delighted.

I have several family members who suffer from hearing loss. They sometimes display embarrassment or self-consciousness about their inability to hear or understand everyone's conversation. I try to be accommodating by speaking louder or sitting closer. However, even for those who are able to afford a hearing aid, there is an unfortunate stigma attached to wearing one. That is a perception that we as a culture can change, and should.

In my business, my hearing is as important as my voice. When performing onstage I normally wear in-ear monitors, which offer a certain level of protection. At live



Ralph Johnson

concerts or in the recording studio, I always wear custom-fit ear plugs with attenuators. I recognized the importance of protecting my hearing early on in my career.

The purpose of playing music loudly is to 'feel' the vibration of the music, or to make certain that every musical nuance is heard. But when the volume acts to destroy one's hearing, it is defeating the purpose. In the studio, the best mix engineers will balance all of the musical elements to be heard at low volume.

My advice to musicians starting out is to not fall for the short-term gratification of hearing the music loudly at the long-term expense of permanent hearing loss. ”



Siedah Garrett



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Ten percent of the proceeds raised from the single "Have a Very Merry Christmas" are being donated to HHF. Please consider extending the spirit of the season with the 99-cent purchase of this new classic on iTunes and Amazon. To learn more about the artists, please see earthwindandfire.com and siedah.com.



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The Hearing Restoration Project (HRP) is Hearing Health Foundation's groundbreaking consortium with a goal of offering a cure for hearing loss and tinnitus. Visit hhf.org to learn more about the cure.

hhf.org
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866.454.3924

NAME:

Cynthia Grimsley-Myers, Ph.D.
Emory University

BIO:

Grimsley-Myers received her Ph.D. in pharmacology at the University of Virginia, where she also conducted postdoctoral research. Now a research instructor at Emory University in Atlanta, Grimsley-Myers is funded by Hearing Health Foundation’s National Junior Board.

IN HER WORDS:

Inner ear hair cells and their formation is the focus of my research. Hair cells are specialized cells in the inner ear located within the cochlea and are responsible for our ability to hear. These cells have a unique apparatus on their surface, called a hair bundle, which is composed of tiny fingerlike protrusions resembling hairs. These “hairs” are highly organized into a specific structure and decrease in length across the hair cell surface. The hair bundle is deflected in a directional fashion toward the longer hairs by sound waves within the ear, leading the hair cell to send a signal to the brain.

Usher syndrome, a hereditary disorder, often results in hearing loss due to abnormalities in the hair bundle structure or its orientation. My research focuses on the role of a previously uncharacterized protein, URB, in the formation of the auditory hair bundle and cochlea duct. Our early data using mouse models indicates that URB is required for early hair bundle formation and cellular patterning during cochlear formation.

Our work aims to identify and characterize the roles of specific genes and signaling pathways in hair cell development. In this way, we hope to have a better understanding of how defects in these genes lead to hearing loss, eventually allowing for the design of efficient therapies for the treatment of hereditary hearing loss.



My dad, a psychology professor who teaches neuroanatomy, definitely had an influence on my early love of biology. I also worked at a nursing home for a while in college. Seeing patients struggling with various medical conditions motivated me to go into biomedical research. My husband also works in the biomedical research field. We hope to impart some of our love for science to our daughters, a 2-year-old and a newborn.

It always amazed me to learn how living things have adapted to survive in their environments. As a child, I once tested how centrifugal force affects the rate and angle of plant growth by putting bean plants on a record player and spinning them around for weeks while they grew. I spun them until the motor burned out!

The intricate design of the organ of Corti, in the cochlea, always fascinated me. The first images I saw of the hair bundles were beautiful, with their precisely coordinated shapes, polarity, and length. I was fascinated to learn how these hair bundles functioned to transmit a sound signal to the brain, and how important their precise design is to our ability to hear. Even slight deviations in the structure of the hair bundle can lead to hearing impairment. The organ of Corti is a complex organ, and we still have much to learn about it.

—Tine Aakerlund Pollard

To read more about this researcher, see the full interview at hhf.org/winter15_meet_the_researcher.

Hearing Health Foundation needs your help to continue funding excellent hearing researchers like Cynthia Grimsley-Myers, Ph.D. To donate today to Hearing Health Foundation and support groundbreaking research, visit hhf.org/donate.

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