

Hearing Loss

Hearing loss is unfortunately one of the most common adverse side effects of standard treatment for high-risk neuroblastoma. According to recent research, up to 62% of children treated for high-risk NB will end up with hearing loss.¹ The hearing loss is caused by certain medications, most importantly the chemotherapy cisplatin. Your child may also have received other medications that damage the hearing, such as carboplatin, certain antibiotics, and Lasix (which is often administered if your child has extra fluid called ascites). The likelihood and severity of hearing loss seems to increase with higher doses of these medications and with younger age of the child. Older children seem less likely to be affected.²

Many families are disheartened upon learning the news that their child, having completed a long and arduous treatment course, must now cope with the challenges of hearing loss. However, since the majority of children with post-NB treatment hearing loss are at an age when speech is developing, it is important not to delay intervention. Even a mild hearing loss can profoundly affect a child's ability to process language. Children who are cancer survivors are already at risk for learning problems due to radiation, chemotherapy, emotional trauma, and other reasons. Delays in effective interventions for hearing loss can only compound these problems.

This section is intended to be a resource guide to hearing loss, targeted especially at the young NB survivor. Topics that will be discussed include specialized terminology, audiograms, hearing loss professionals, and educational considerations, starting with a discussion of the first step you should take -- getting the most precise evaluation possible of your child's hearing status by a hearing loss professional.

Getting Your Child Evaluated

When you read through the standard material on hearing evaluations for young children, you will see checklists of signs that a child may have a hearing loss. In some ways, your task is easier, since any child who has completed the standard protocol for high-risk NB may have hearing loss. **Any child who has received one of the platinum-based chemotherapy agents (cisplatin, carboplatin) should be evaluated for hearing loss.** If your child is very young, it is very important to find an audiologist who has experience working with young children, because it is notoriously difficult to get accurate audiograms with this age group. It is very common to hear parents of children with hearing loss tell stories of going through multiple audiologists before a hearing loss can be accurately measured. For example, it took over a year and three audiologists, before one two-year-old NB survivor was able to be evaluated precisely enough to equip him with hearing aids! The A.G. Bell website has good advice for finding a pediatric audiologist:

http://www.agbell.org/DesktopDefault.aspx?p=Find_an_Audiologist

Key points made include asking audiologists about their experience with young children, and asking other parents of hearing-impaired children for recommendations.

There are several methods of testing hearing, but for children over 6 months or so of age, behavioral assessments are most typical. In this type of test, the child's response to pure tones is assessed while sitting in a sound booth. For a very young child, this response may be as simple as a head turn. Preschoolers will be encouraged to make this into a game, tossing balls into a bin, for example, in response to the tones. Some young children may be frightened by the sound booth, which is often small and dimly lit. This is why it is so important to find an audiologist who has a lot of experience

with small children and who has a good rapport with your child. Sometimes, the test is best run with two evaluators – one to run the equipment and the other to observe the child.

Here is a list of resources on hearing tests and how to interpret them.

<http://kidshealth.org/parent/general/eyes/hear.html>

Hearing evaluation in children. This provides an overview of the types of tests used, types of hearing loss, and assistive technology such as hearing aids and FM systems.

http://www.agbell.org/DesktopDefault.aspx?p=Testing_for_Hearing_Loss

Another overview of the types of hearing tests that are used.

<http://www.audiology.org/aboutaudiology/consumered/guides/audiogram.htm>

How to read an audiogram. You'll want to be able to explain your child's audiogram when you are dealing with school personnel, who often have never worked with a hearing-impaired child before.

<http://www.earinfo.com/how-to-read>

Here is another explanation of how to read an audiogram.

http://www.handsandvoices.org/resources/coGuide/05_Lossvseffct.htm

<http://www.helpkidshear.org/resources/starter/degrees.htm>

Good explanations of the degrees of hearing loss. This is also useful when dealing with school personnel.

<http://www.utdallas.edu/~thib/rehabinfo/tohl.htm>

This site has simulations of what different types of hearing loss actually sound like.

Types of Hearing Loss

This section presents a quick overview of the types of hearing loss, and then describes the type of hearing loss suffered by most NB survivors. You will want to understand the differences between types of hearing loss so that you can more effectively communicate with your child's audiologist, speech therapist, and educational personnel. It is often useful to memorize a quick phrase that summarizes your child's hearing loss: "bilateral moderate-to-severe sensorineural loss," for example.

Hearing loss can be categorized by general cause. A **conductive hearing loss** occurs when sound is not conducted through the outer and middle ears. This can be caused by otitis media or problems in the Eustachian tube. A **sensorineural hearing loss** is caused by damage to the inner ear, the cochlea, or to the nerves in the inner ear. This can be caused by disease, certain medications, genetic syndromes, and exposure to noise. This is a permanent hearing loss that cannot be corrected by medication or surgery. Finally, **central auditory processing disorders** originate in the auditory centers of the brain, and may be caused by injury or disease.

A hearing loss is also characterized in terms of its "shape". This refers to the shape of the line on the audiogram. A **sloping** hearing loss means that hearing is better in the lower frequencies and gets worse in the high frequencies. A **flat** hearing loss means that hearing is the same across all frequencies. A **rising** hearing loss means that hearing is worse in the low frequencies and better in the high frequencies. Finally, a **cookie-bite** hearing loss means that hearing is worst in the middle frequencies.

The type of hearing loss that is most common among children who have received ototoxic chemotherapy medicines is a bilateral, sloping, sensorineural hearing loss. The hearing loss may range from mild to profound, although moderate or moderate-to-severe seems to be most common. What this means is that the hearing loss is about the same in both ears, is worse in the higher (and

often middle) frequencies, is caused by damage in the inner ear, and is serious enough to require amplification. The fact that the hearing loss is often in the middle and high frequencies is important since those are the frequencies at which many speech sounds occur. It is very common for young children with this type of hearing loss to need speech therapy as a result. NB survivors often finish treatment at an age when language is being acquired, so it is important to keep this in mind.

Here are resources on types of hearing loss:

<http://www.helpkidshear.org/resources/starter/types.htm>

Mild Loss

<http://www.sciencedaily.com/releases/2004/11/041123204642.htm>

http://www.audiologycentre.com/child_faq_management.htm

Terminology

<http://www.helpkidshear.org/resources/starter/glossary.htm#S>

A glossary of the types of terminology you are likely to hear.

<http://www.listen-up.org/htm/acronyms.htm>

A glossary of acronyms that you are likely to encounter. Be forewarned – the area of hearing impairment abounds with acronyms.

Now What Do We Do?

The current thinking among audiologists is to aggressively aid hearing-impaired children as early as possible. This helps minimize speech and language delays. Most children with moderate or worse hearing impairments will need hearing aids, and many children with mild hearing impairments will need hearing aids as well. You will find that you will be spending a lot of time on hearing aid issues, so it is worth understanding the technology.

Besides aggressive amplification, the other important strategy is to obtain educational services for your child as quickly as possible. Such services may be through an early intervention program or through your school district, depending on your child's age.

Hearing Aids

Hearing aids work by amplifying sounds coming into the ear. Hearing aids can be programmed to boost sounds selectively at different frequencies, depending on the individual needs of the wearer. Hearing aids may be either **analog** or **digital**. Digital hearing aids provide a cleaner signal and seem to be most typically recommended for children.

Hearing aids for children are most typically **behind-the-ear** hearing aids (BTE). These consist of the hearing aid itself, which fits behind the ear, and an **ear mold** which fits into the ear. This both anchors the hearing aid to the ear and sends sound into the ear. This type of hearing aid works well for children because it is easy to insert and because only the ear mold needs to be replaced as the child grows. Ear molds are available in many child-friendly colors – we have spotted children with swirling tie-dye style ear molds, and pink sparkly ear molds! Besides the BTE style hearing aids, **in-the-ear (ITE)** and **in-the-canal (ITC)** hearing aids are available and may be appropriate for teenagers whose ears have stopped growing.

Resources on hearing aids follow.

http://www.childrenshearing.org/custom/hearing_aids.html

A discussion of hearing aid types, as well as of other assistive technology.

http://www.agbell.org/DesktopDefault.aspx?p=Hearing_Aids

A great overview of hearing aids.

<http://www.boystownhospital.org/Hearing/hearingaids/index.asp>

Another overview, with more technical detail on digital hearing aids.

http://www.asha.org/public/hearing/treatment/child_aids.htm

Another overview of children's hearing aids, along with information on funding.

<http://www.utdallas.edu/~thib/rehabinfo/ao.htm>

This page has pictures of the various kinds of hearing aids, and an extensive section on hearing aid maintenance and troubleshooting.

Hearing aids are unfortunately quite expensive. It is common for digital hearing aids to cost well over \$2000. And sadly, many insurance plans will not cover hearing aids. Many families who have been through NB treatment are already reeling financially, so this is a very unwelcome blow. In some states, Early Intervention and other state programs will pay for hearing aids, so check these options in your state. Medicaid will also cover hearing aids. Some states are introducing mandates that insurance pay for hearing aids. In some cases these mandates are so new that even the insurance company representatives may not be aware of the requirement, so always check the laws in your state if you have been turned down for insurance coverage.

Here is an entire page of links on funding for hearing aids.

<http://www.listen-up.org/haidfund.htm>

Other Assistive Technology

FM systems are commonly used in classroom settings, although they can also be useful in any noisy situation. These systems enhance the "speech to noise ratio" when the speaker is in a noisy setting (such as a classroom) and the listener is not physically close. The speech to noise ratio is the relationship between the loudness of the speaker and the loudness of the background noise level. Hearing-impaired children have difficulty picking out a speaker's voice when there is a lot of background noise. It isn't enough for a teacher to simply raise his or her voice since the hearing-impaired child will still have trouble picking out the differences between speech sounds, which is necessary to understand the speech. This is where an FM system can be a big help.

A **personal FM system** consists of a small microphone that the teacher wears and a receiver worn by the child. A common type of system involves 2 tiny receivers (called "boots") that plug into the child's hearing aids. The receivers pick up the signal transmitted from the microphone and transmit it directly into the hearing aids, thus boosting the teacher's voice significantly.

Soundfield systems consist of a microphone worn by the teacher, and small speakers that transmit the signal. Such systems are useful for children with mild hearing loss, but personal FM systems are more effective for children with higher degrees of hearing loss. Interestingly, some school districts have been experimenting with soundfield systems for all students, finding that amplification boosts attention and helps all children learn.

Cochlear implants are used in situations where a child has severe-to-profound hearing loss, with little or no residual hearing. This type of device bypasses the inner ear and directly stimulates the auditory nerve to send information. Most children who have lost hearing due to chemotherapy do not have this degree of hearing loss, so it is not common to see cochlear implants in this group.

Closed captioning is used in the classroom for films or video. It is common to write this accommodation into a child's IEP or 504 educational plan (more on those topics below in Educational Considerations). It is also possible to place the FM system mike in front of the speakers or to plug it directly into the audio output of the VCR.

When assistive technology is used in a school setting, it is important for the teacher(s) and other staff who interact with the child to be trained in the proper usage of the devices. These devices are fragile and unfortunately, break fairly often. The teacher and at least one other person in the school should know how to insert and remove the hearing aids and FM system receivers, and how to change batteries. As the child grows older, he or she can also take more responsibility for these tasks. If there is an IEP or 504 plan, these responsibilities should be spelled out. It is also useful if the school has the services of an educational audiologist, or can communicate with the child's private audiologist directly for assistance when there are problems. For example, one NB survivor's audiologist runs an in-service training session for his teachers each year, which is specified in the child's IEP.

Educational Considerations

Classrooms, as we all may remember, tend to be noisy, bustling places. A significant amount of the material learned in a classroom is learned through the ears, by listening. And part of that is learned not directly from the teacher, but through listening to other conversations. Children with hearing loss, even if they have hearing aids, tend to have trouble distinguishing speech sounds in a noisy situation. Hearing aids amplify background noises as well as speech. Thus, children with hearing loss have a lot of difficulty simply accessing the material to be learned. In addition, children with hearing loss often are behind other children in terms of vocabulary, which has a great impact when learning to read. Many children with hearing loss end up performing below grade level in reading, and the gap often progresses over time. (<http://www.readingrockets.org/article/5135>).

In addition, children with hearing loss often have social delays since they have trouble communicating with other children, and picking up information and social cues from other children's conversations. Children with hearing loss are often described by teachers as being "out of it" or having problems paying attention. Anecdotally, some parents have noticed that many children are misdiagnosed as having attention difficulties before finally being diagnosed with hearing loss.

Assistive technology such as personal FM systems are very important in overcoming the barriers experienced by children with hearing loss. If your child uses hearing aids, it is important that he/she wear them every day, and that they be maintained in working order. If your child uses an FM system, it is important that every teacher use the microphone when speaking to the class. For older children, it may also be useful to pass the microphone around when classroom discussions are taking place.

There are a number of classroom accommodations that can be made to further assist your child. These often need to be explicitly explained to the teacher, or better yet, specified in your child's IEP if he/she has one. Many teachers are not familiar with hearing loss issues and do not know the types of accommodations that can help. Here is a list of helpful accommodations:

- Preferential seating. A child should be seated near the front of the classroom where he/she can easily see the teacher. If the teacher regularly changes position, the child may need to be able to change seating as well. Many children with hearing loss use some degree of lip reading to augment understanding, so being able to see the teacher can be very important. The child should also be seated away from noisy equipment such as air conditioners or fans.
- The teacher should face the children when speaking, rather than facing a board.
- The teacher should write down important information, such as new vocabulary words, assignments, and announcements.
- The teacher should ask the child if important instructions have been understood.

- The teacher should help the child understand when transitions are occurring, by using phrases that make the transition explicit. For younger children, it is especially important to make transitions to new types of activities explicit. One child's kindergarten teacher used a set of cards with pictures of children playing (for free time), children sitting quietly (for circle time), and children writing (for writing activities).
- The teacher should also assist with new vocabulary, since this is an area of particular difficulty for hearing-impaired children. The meaning of new words should be explicitly explained before using the new words to teach material. If the child has a Teacher of the Deaf as a support service, this specialist will usually preteach and reteach new vocabulary.

Here is a link which describes strategies for teaching hearing impaired children:
<http://www.as.wvu.edu/~scidis/hearing.html>

In addition, there are steps that can be taken to reduce classroom noise. Noise often comes from sources such as air conditioning systems, pencil sharpeners, hallway noise, chairs and desks sliding on the floor, and of course, other children talking. Reverberation in the classroom can make such noise worse. Steps that can be taken include:

- Installing acoustic panels on the walls.
- Installing carpeting.
- Putting tennis balls on the chair legs – this is a common solution since most districts will balk at installing carpeting as an accommodation.
- Requesting that tests be taken in quiet settings. This can be written into the child's IEP.
- Seating the child away from noise sources such as windows or corridors.
- Requesting that landscaping services (especially mowing) be done during non-instructional time.

Remind your school district that accommodations that reduce background noise will help all the students in the class, not just your child.

Finally, many children with hearing loss can benefit from the services of specialized support staff. Many parents are not even aware that some of these support services exist. One of the most important types of specialists that may be available to your child is the **Teacher of the Deaf**, or sometimes called an itinerant teacher of the hearing-impaired. This type of teacher, not to be confused with sign language interpreters, usually has completed a degree in Deaf Education. Typically, a school district will employ this type of teacher to travel to all the schools in the district and work with hearing impaired children. Smaller districts may use teachers contracted through regional consortia or a regional School for the Deaf. For example, in New York, such teachers are usually contracted for through a regional educational consortium called BOCES. A teacher of the deaf can provide many services, but the most important roles are to help other teachers develop strategies for working with your child, serve as a liaison between the various staff members and the parents, and provide one on one tutoring of your child in the area of vocabulary and strategies for understanding language. A Teacher of the Deaf is different from a speech therapist, whose role is to help a child produce comprehensible language. Often, a child with a serious hearing impairment will work with both types of specialists. School districts will usually have speech therapists on staff, and may try to argue that a hearing-impaired child only needs the speech therapist, so it is important to understand the distinction between the two types of specialists. Older children with hearing loss may also benefit from the services of transcribers to take notes, or from peer notetakers. Children who use sign language (which is less typical among cancer survivors) may also use a sign language interpreter.

IEPs and 504 plans

This topic often causes the most stress for parents with special needs children, because these are the laws and regulations that govern your child's access to specialized services and accommodations.

The **Individuals With Disabilities Education Act (IDEA)** is the main law that governs access to specialized educational services. Schools are required to provide children with disabilities with a **free appropriate public education (FAPE)**. A child who is determined to require such services will have an **Individualized Educational Plan (IEP)** which lists the educational goals to be met and the types and frequencies of services. Unfortunately, there are many areas of controversy, and possibilities for disagreements, within this law, especially as it relates to which children qualify for an IEP, and which types of services are appropriate. A child with hearing loss, and especially a child who is a cancer survivor, has a high likelihood of qualifying, so you should initiate the process of having your child evaluated as early as possible.

This article lists steps to take in order to request that your child be evaluated.

<http://www.fetaweb.com/01/faqs.evals.htm>

One tip – your child will almost certainly have a speech evaluation as part of this process. Request a speech evaluator that is trained in working with hearing-impaired children, since speech delays often manifest themselves differently in this group. Under the regulations, if you are not satisfied with an evaluation provided by the school, you can request an independent evaluation. Here is an article which discusses that process.

<http://www.wrightslaw.com/info/test.iee.steedman.htm>

Hearing-impaired children who do not qualify for an IEP will usually qualify for a 504 plan. **Section 504** is a law that prohibits discrimination against individuals with disabilities. Part of it (subpart D) mandates that children with disabilities have equal access to an education. Children who fall under this regulation (as all children with hearing loss do) will have a 504 plan drawn up by the school, which will typically mandate usage of hearing aids, an FM system, captioning of films/videos, notetakers, and sign language interpreters if needed. Very confusingly, some school districts will also provide services such as speech therapy in this plan, even though the child does not have an IEP. Keep in mind that a parent has far fewer procedural safeguards under Section 504 than under IDEA, so if your child needs support services to succeed in school, it is usually better to pursue an IEP.

The area of special education law is vast and difficult. It is impossible to delve into all of its complexities here. The WrightsLaw website and book series is an excellent resource which covers all of this material in depth. Links are provided below. School districts will also often have a parent advocate who participates in IEP meetings with families. This person may be helpful so you should try to locate your district's parent advocate. It is also possible to hire an independent educational advocate to assist you in obtaining needed services for your child from your school district. And finally, there are lawyers who specialize in special education law who can represent you at hearings. Hopefully you will never need such services!

Additional Resources

Organizations

www.handsandvoices.com

Hands & Voices is a nationwide non-profit organization dedicated to supporting families and their children who are deaf or hard of hearing, as well as the professionals who serve them.

<http://www.agbell.org/>

The Alexander Graham Bell Association for the Deaf and Hard of Hearing helps families, health care providers and education professionals understand childhood hearing loss and the importance of early diagnosis and intervention. Through advocacy, education, research and financial aid, AG Bell helps to ensure that every child and adult with hearing loss has the opportunity to listen, talk and thrive in mainstream society. They have chapters located in the United States and a network of international affiliates.

<http://www.clarkeschool.org/>

The Clarke School for the Deaf has a number of branches in Massachusetts, Pennsylvania, and New York. In addition to educational programs, they also provide independent educational evaluations. Many other schools for the deaf will also provide this service; however, Clarke is particularly well known.

Online Resources

www.listenup.org

A wonderful collection of articles, and most importantly, the Listen-Up mailing list, for families of children with hearing loss. The mailing list is fairly high traffic, but you can learn almost everything you need to know there about topics from cheap sources of batteries and diagnosing problems in hearing aids, to negotiating with school districts for services.

www.wrightslaw.com

The main “go-to” site for learning how to navigate the special education world, particularly focusing on IDEA regulations and parents’ rights under that law. The people who run this site also run training sessions for parents all over the country.

Books and Publications

Laverdière C, Cheung NK, Kushner BH, Kramer K, Modak S, LaQuaglia MP, Wolden S, Ness KK, Gurney JG, Sklar CA. “Long-term complications in survivors of advanced stage neuroblastoma,” *Pediatric Blood Cancer*, September 2005, pp. 324-332.

Y . Li, R. Womer, J. Silber, “Predicting cisplatin ototoxicity in children: the influence of age and the cumulative dose.” *European Journal of Cancer*, vol. 40, pp. 2445 -2451.

James G. Gurney, Jean M. Tersak, Kirsten K. Ness, Wendy Landier, Katherine K. Matthay, and Mary Lou Schmidt, “Hearing Loss, Quality of Life, and Academic Problems in Long-term Neuroblastoma Survivors: A Report From the Children's Oncology Group,” *Pediatrics*, vol. 120 No. 5, November 2007, pp. e1229-e1236.

From Emotions to Advocacy, The Special Education Survival Guide, 2nd Edition, Pam Wright and Peter Wright, Harbor House Law Press, 2006.

Wrightslaw: Special Education Law, 2nd Edition, Pam Wright and Peter Wright, Harbor House Law Press

Your Child’s Hearing Loss: What Parents Need to Know, Debbie Waldman, Jackson Roush, Berkeley Publishing Group, 2005.

Please contact info@cncfhope.org with any comments

¹ Lavadiere et al, *Pediatric Blood Cancer*, Sept. 2005, 324-32

² Li, Womer, and Silber: “Predicting ototoxicity in children: Influence of age and the cumulative dose,” *Eur. J. Cancer* 40:2445-2451, 2004.