American Hospital Dubai

ent NEWSLETTER

Fall 2008

Newborn hearing screening

Dr. Muaz Tarabichi

"As otolaryngologists, we have a moral responsibilities towards our patients, especially children. One issue that needs to be communicated to society at large is the need for early identification and intervention for hearing loss. The advent of cochlear implants have increased the importance of finding out as early as possible the status of the hearing in every child. We need to do more to get the message out on newborn hearing screening."

Newborn Hearing Screening why Universal?

Lubaina Sharafally, M.A. , CCC-A Clinical Audiologist

Babies begin developing speech and language from the moment they are born. They learn by listening to and interacting with the sounds and voices around them. Listening in the first few months of life prepares the baby to learn language. Before their first birthday, they babble many of the sounds they hear spoken around them. By the first birthday, infants are already figuring out what words mean. These early steps are building blocks for communication.

When these sound and voices can't be heard, language learning is often delayed. This can lead to slow development of speech and language. It can create difficulties in family communication. Delays in speech and language can also lead to school problems, both academic and social. Detecting hearing loss early helps to prevent these delays. Unfortunately, hearing loss due to its silent nature is easily missed in the early years of life; the most important period for language development, the foundation for all learning and human interaction. Hearing loss is one of the most common congenital anomalies, occurring in approximately 2-4 infants per 1000. Hospitals routinely screen babies for some specific problems, like phenylketonuria (PKU). Interestingly, hearing loss occurs more often in babies than any of the other problems that are screened for at birth.

Traditionally, hearing screening had been targeted to high risk populations, such as those infants who have been in the neonatal intensive care unit, family history of hearing loss or those with craniofacial anomalies. Screening only those infants who meet the high-risk register (HRR) criteria is not enough because as many as 50% of infants born with hearing loss have no known risk factors. This led to the 1993 joint commission for infant hearing – (USA) position statement which recommended all newborns (universal newborn hearing screening) be screened for hearing loss before leaving the hospital. This was further amended in 1994 to read "all infants with hearing loss should be identified before 3 months of age and receive intervention by 6 months of age".

Universal newborn hearing screening aids in reducing the mean age of identification and intervention. Studies in the US, where hearing screening since 1993 has been their mean age of identification, reduce from 12 months to 3-6 months and age of intervention reduces from 13-16 months to 5-7 months. Research has also shown that a deaf child can develop language at the same rate as their hearing peers if deafness is identified by the age of six months and an effective early intervention program is provided. Working as a clinical audiologist in the UAE since 1998, I have felt a strong need for universal newborn hearing screening services, as congenital or early acquired hearing loss in the Middle East seems to be an epidemic problem due to the social and cultural structure. Also, the age of diagnosis of children's hearing loss among Arab populations is quite late with most children being identified past the critical language learning period. The average age at which children are found to have hearing loss is 2-3 years. Children with mild-to-moderate hearing loss are often not identified until 4 years of age. My first task upon joining the American Hospital Dubai in 2000 was to initiate a newborn hearing screening program. This concept was welcomed and well supported at the hospital. Newborn hearing screening services are offered to all our babies delivered at the hospital and our program is modeled as per NIH guidelines 1993. The screening uses a quick and non invasive test – Otoacoustic emissions (OAE's). Babies are tested in the newborn nursery prior to discharge. In the event of a retest a repeat OAE/ Auditory Brainstem Response (ABR) testing is done. Both tests are painless for the baby. OAE's and ABR testing provide the audiologist with valuable information about an infant's hearing loss. If a hearing loss is detected, the next step is intervention. Early intervention involves a range of professionals - our team comprises of Dr, Muaaz Tarabichi ENT, Ms. Lubaina Sharafally, Clinical audiologist; Dr. Rana Batterjee, Bilingual audiologist; and speech and language pathologists Ms. Nadine Ewanchyshyn and Ms. Ghada Ajamieh.

With the ability to detect and diagnose an infant with hearing loss soon after birth, there is now no reason why any infant born with a hearing loss should experience anything but normal speech and language development as a result of early intervention.

GENE THERAPY FOR HEARING LOSS

Dr. Rana Batterjee, Au.D., CCC-A, ASHA-F, F-AAA,
Clinical Audiologist

I recently read an article on gene therapy for hearing loss. It reminded me of a parent who decided not to implant her child with profound hearing loss until geneticists could figure out how to "grow cochlear hair cells" in replacement of Cochlear Implant surgery. Surely you'll find the following edited article by Mark Parker and Robert Martin interesting.

THE GENOME PROJECT

Hearing scientists have focused on identifying genes that might be used to restore hearing loss. Using gene therapy to treat lost hearing has produced encouraging results in animals and holds great promise for future treatments in humans. It's no wonder that hard-of-hearing people are excited when they see news reports on this topic. The collection of all of the genes that make us human is called the genome. In 2003, researchers deciphered the human genome. The result of their work, known as the Genome Project, is a database containing a complete set of codes for a full set of human chromosomes.

The Genome Project has published an online database of the genetic codes for the approximately 20,000 genes that make us human (www.ncbi.nlm.nih.gov/sites/entrez). This has led to the identification of genetic mutations that result in hearing loss. To date, more than 70 genetic mutations are known to lead to hearing loss (http://webh01.ua.ac.be/hhh/). Now, researchers are using this genetic information to try to solve human problems like restoring hearing. For example, scientists have discovered that a gene called Math1 (also known as Atoh1) is both required and sufficient for hair cell genesis. This means that a cell in the inner ear that expresses the Math1 gene will develop into a hair cell. In several recent studies researchers have inserted the Math1 gene into the inner ears of deafened rodents, and measured regenerated hair cells. Furthermore, these regenerated hair cells improved ABR thresholds in deafened animals.This technology will need to be refined before genes can be used on humans. Nonetheless, these studies indicate that gene therapy may be used to treat human hearing loss in the future.

EMERGING TECHNOLOGIES

New technologies have facilitated the use of gene therapy. For instance, ordering a gene today is about as easy as ordering a hearing aid. If you want to insert a gene into the cochlea, you first need a gene of interest, and then a delivery vehicle for the gene. The delivery vehicle is typically a virus that has had its own DNA mutated to make it benign. If one wanted to use the pro-hair cell Math1 gene, all that is required is to go to the Human Genome Project, copy the text sequence for Math1, and e-mail this text file to a company that creates synthetic DNA molecules (e.g.,www.blueheron.com). The company would generate a synthetic strand of DNA containing an exact sequence of the gene you specified. The Math1 gene could then be inserted into a commercially available virus that has been engineered for optimal efficiency in infecting human cells (e.g., www.invitrogen.com). Finally, the virus could be injected into the cochlea. In theory, cochlear cells infected by this virus would express Math1, and develop into hair cells. While viral-mediated gene delivery has demonstrated promise, this technology is not ready for the clinic. One problem with current viruses is that they may infect cells at random. That could cause Math1 to be expressed in cells that shouldn't become hair cells in a normal cochlea. Expression of Math1 in specific cochlear cells, such as supporting cells, is a focus of research.

IN CONCLUSION

This is an exciting time in biological research. Most of the technical code used to create all the structures in the human body is now known. Scientists are working to use this information to re-grow defective structures, such as hair cells in the cochlea. Success has been achieved with animal studies. However, much work needs to be done before this approach can be tried on humans.

Reference: Parker,M., & Martin,R. (2008). Gene therapy for hearing loss: An update. The Hearing Journal, 61 (6), 46.

Early hearing detection and intervention:
Audiologists and SLPs Collaborate in Successful Program

Nadine Ewanchyshyn, MS, S-LP (C), CCC-SLP
Speech Language Pathologist

At the American Hospital Dubai, in the ENT Clinic and the Cochlear Implant Center, the audiologists and the speech-language pathologists work together, enabling us to serve children with hearing loss and their families. We have found, as is discussed in the article, Early hearing detection and intervention: Audiologists and SLPs Collaborate in Successful Program, that the "communication and cooperation among audiologists and speech-language pathologists (SLPs) is pivotal to the success …for early hearing detection and intervention." Since the 1999 implementation of universal newborn hearing screening in North Carolina, USA, audiologists and SLPs have forged successful collaboration by working in partnership in programs serving children with hearing loss and their families.

Professional Roles and Responsibilities

Essential roles and responsibilities of audiologists and speech-language pathologists were reaffirmed recently by the Joint Commission on Infant Hearing (JCIH), Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs (EHDI) (JCIH, 2007).

The role of the audiologist, according to the position statement, includes management of newborn hearing-screening programs; quality assessment; service coordination; and audiological diagnosis and treatment, including the timely fitting and monitoring of amplification and other sensory devices. Average age of diagnosis is approximately 3 months of age. Initial hearing aid fitting is usually completed within two weeks. Once infants reach a developmental level of approximately 6 months, physiologic tests are supplemented by behavioral measures.

SLPs, according to the position statement, are among the specialists who provide early intervention services to young children with hearing loss and who can provide appropriate assessment of language, speech, and cognitive-communication development, as well as offer their expertise in auditory skill development. The position statement recommends a global screening of developmental milestones for all infants and young children, provided in/by the family's "medical home" and further recommends immediate referral to an SLP for further evaluation if there is evidence of delays in speech, language, or communication development.

Diagnosis and Management

Acoustic amplification alone is not sufficient. Cochlear implantation evaluation and mapping by audiologist is required. In addition, management includes an auditory-oral preschool where comprehensive assessment and intervention services, including auditory-verbal therapy, are provided by SLPs and other early intervention specialists. Children's progress is monitored through weekly team meetings.

An important strength consistent with the recommendations of the 2007 JCIH position statements, is that "all professionals are specialists rather than generalists. Furthermore, every effort is made to enroll children and begin intervention immediately following referral. At no time in the referral process is a child placed on a waiting list."

A Vital Role

Audiologists and SLPs play a vital role in early identification, diagnosis, and intervention for young children with hearing loss and their families. Challenges persist, such as those related to health conditions or disabilities that exist in addition to hearing loss, limited financial coverage of hearing instruments, transition from early intervention to the local school system, and a shortage of early intervention providers with expertise needed to serve these special populations. But despite these challenges, progress continues through advances in technology combined with the dedicated efforts of audiologists, SLPs, and other service providers committed to parent-professional collaboration and comprehensive EHDI services.

Roush, J., Wilson, K., Alberg, J. (2008, Feb. 12). Early hearing detection and intervention: Audiologists and SLPs collaborate in successful program. The ASHA Leader, 13(2), 14-17, 45.

Early identification of children with hearing loss

Ghada Ahmad, MA, Speech-language Therapist

The screening allows us to identify children and to provide them with access to communication through early intervention which begins with amplification and then the therapy.

Early intervention

Early intervention actually begins at the point at which we diagnose hearing loss and we provide counselling and support to parents, because obviously the vast proportion of parents with new born babies getting screened don't even imagine that their baby is going to have a significant hearing loss and so as opposed to past generations where they were noticing developmental delays in their children and they were looking for the answer as to why their children were not developing appropriate speech and language skills, these parents are anticipating that they are going to have a perfectly normal child in every way...and so when we're telling a parent that we have the capability of diagnosing this hearing loss with this new born baby, they are frequently very shocked. It's also at a point in time especially for mothers when they're biochemically at a point where they can develop serious depression, so the counselling strategies need to be immediate and they need to be appropriate for both the grief process for the hearing loss and also for the cultural competence. One of the things of which we are very acutely aware is that we've become a global society and there really is no country in the world anymore that is completely homogenous in culture or race so the different ways that people react [to hearing loss] because of their culture, race or religion can completely determine whether or not they are going to accept therapeutic intervention. Early intervention begins with the counselling, it typically almost always begins with the amplification and provision of appropriate amplification; and as soon as we have the ability to provide access to communication, whether it's auditory or whether it's visual, then we are training parents to adapt within their normal environment so that they optimise the access to language and communication.

What are the wider implications for parents?

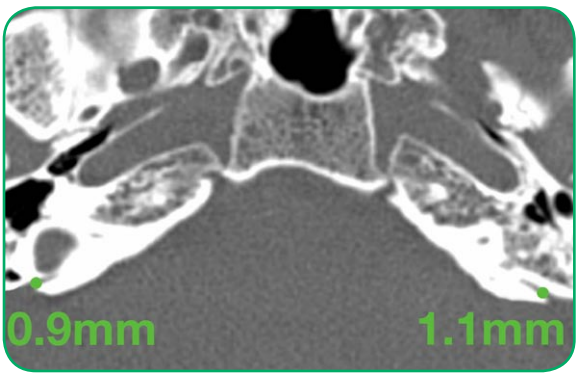
The wider implications are that, a generation ago and prior to universal newborn hearing screening, the message that parents got when their children were diagnosed with significant hearing loss was pretty dismal. You could even tell it on the face of the audiologist who were giving the diagnosis...it was a horrible diagnosis basically because the outcomes and in terms of education were not very positive and the outcomes in terms of speech intelligibility were not very positive. A vast proportion of these children couldn't function in a hearing society without incredible accommodations.(in orange so it is up to you if you want to keep it or omit it) Today, audiologists are diagnosing hearing loss and they're saying to the parents that we have the technology and educational expertise so that their children can be pretty normal.

Christine Yoshinago-Itano is Professor of Audiology at in the Department of Speech, Language and Hearing Sciences at the University of Colorado in the United States.

Progressive Sensorineural Hearing Loss in Childhood A case report

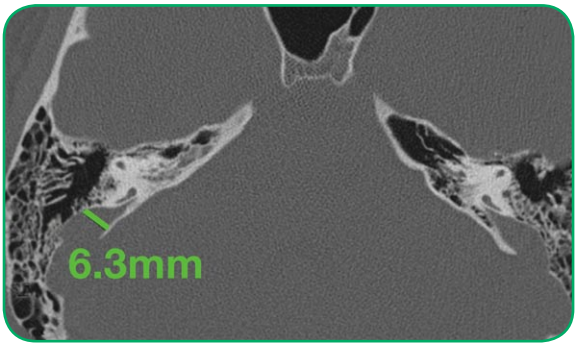
Dr. Omar Nazhat, MD

Introduction: Large Vestibular Aqueduct Syndrome (LVAS) may result in sudden, fluctuating, or progressive sensorineural hearing loss. It is defined as the combination of the clinical presence of Sensorineural hearing loss in a child and the identification of the large vestibular aqueduct on a CT scan, or the MRI identification of the enlarged endolymphatic duct and sac. A child with LVAS may retain a good hearing until a triggering factor launch the hearing loss, commonly head trauma or infection. The vestibular aqueduct is a narrow bony canal (aqueduct) that runs through the skull, connecting the inner ear (vestibule) to the cranial cavity-hence its name. At the half-way point, the diameter normally ranges somewhere between 0.5 mm and 1.4 mm and averages 0.8 mm (fig.1). LVAS is defined as a vestibular aqueduct with a diameter greater than 1.5 mm at the midpoint on CT scan of Temporal bones.



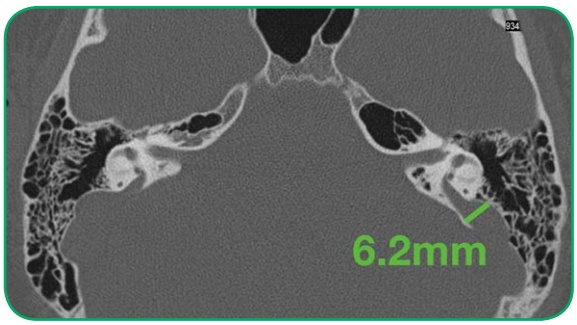
(fig.1)

A fifteen years old male, presented to our ENT clinic with recent acute sudden hearing impairment, from the history the patient's father stated that his son has had meningitis 12 years ago after which the hearing loss established and the patient has been using hearing aids since then on both sides and did well regarding speech development and communication skills, until recently when acute sudden deterioration in hearing occurred mainly on the left side with dizziness. No history of ear pain, discharge on either side. No other symptoms regarding the nose and throat. ENT examination was normal, patient sent for hearing evaluation tests which showed profound bilateral Sensorineural hearing loss, more on the left side with poor speech discrimination and normal middle ear pressure on both sides.



(fig.2a)

An urgent CT scan of the brain and temporal bones was scheduled and a short course of steroids (60 mg daily orally for one week) prescribed for the patient started at once in an attempt to improve his acute sudden hearing loss. The CT scan showed an enlarged vestibular aqueduct bilaterally, but more on the left side, the vestibular aqueducts measured more than 6mm in the midpoint indicating unusual enlargement of the aqueduct (fig. 2a, b). Although the short course of steroids did improve the hearing on low frequencies level after one week, yet the progressive nature was thoroughly explained to the family and that ultimately the hearing deterioration will leave the patient with cochlear implantation as the last resort.



(fig.2b)