Hearing Loss in Children: The Otologist’s Perspective

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Background and Protocol

Universal newborn hearing screening was mandated in North Carolina in 1999. The North Carolina program appears to be highly effective with 98.2% of the 133,823 infants born in 2007 receiving a hearing screening (personnel communication, North Carolina Early Hearing Detection and Intervention [EHDI] Program). Since the inception of screening in the state, there has been a dramatic increase in the number of children presenting for comprehensive hearing evaluation and management. The age at identification has decreased from 24–30 months down to 2 to 3 months (Harrison, Roush and Wallace 2003). Currently, our center is serving more than 1300 children with hearing loss (roughly 650 using hearing aids and 650 using cochlear implants). To serve such a large population, we have developed a multidisciplinary approach and standardized protocols for the evaluation and treatment of these children. The multidisciplinary endeavor requires input from a diverse group of professionals with expertise in a variety of hearing-related disciplines. These areas are broadly depicted in figure 1. It is common that certain disciplines might predominate during one phase of the process while others frequently become more active later.

Cooperation through mutual respect for one another’s skills and opinions forms the backbone for successful collaboration for the child and family’s benefit.

In an effort to create a timely diagnosis and early intervention, we have created a timeline for the events of the first year of life (figure 2). While not set in stone, we believe this serves as a rough guide for the events of the first year of life as it relates to hearing loss and its management. The cornerstone goals of the first year are:

1. **Identification** of hearing loss and establishing precise auditory thresholds;
2. **Diagnosis** of the etiology for the hearing impairment;
3. **Intervention** through provision of appropriate treatment and/or technologies; and
4. **Education** by providing information for families to help make decisions.

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Figure 1. Schematic representation of the Hearing Loss Team at the University of North Carolina at Chapel Hill.
A comprehensive diagnostic hearing evaluation is the first requirement following a confirmed “fail” or “refer” indication by the newborn infant hearing screening examination. At our center, this evaluation is carried out by a group of highly experienced audiologists with expertise in pediatric hearing testing. The initial follow-up testing for each child occurs under one of three environments: natural sleep, conscious sedation, or general anesthesia. Infants younger than about 3 months of age are tested in natural sleep, if possible. Babies older than this, without other medical contraindications, are sedated prior to testing. After medical clearance by a physician, a nurse from the hospital’s pediatric sedation team administers a sedative and remains at bedside to monitor the entire session. Sedation is typically accomplished with chloral hydrate delivered orally or midazolam delivered intravenously. In cases where the infant is scheduled for a procedure under general anesthesia (e.g., surgery or imaging), the evoked potential testing is incorporated into the procedure sequence, if appropriate. Some children are not deemed candidates for safe monitored conscious sedation and so testing is carried out under the supervision of an anesthesiologist (i.e. general anesthesia). The duration of the test is usually dictated by the test environment, ranging from about 30 minutes in the operating room to over an hour under conscious sedation.

No single test is sufficient for precisely identifying the degree of hearing impairment in a baby (American Academy of Pediatrics and Joint Committee on Infant Hearing 2007). Rather, a test battery is needed that might include a variety of measures such as: auditory brainstem response audiometry (ABR), auditory steady-state responses (ASSR), otoacoustic emissions (OAE), and tympanometry (immittance). While reasonably accurate, each of these testing procedures has a variety of potential shortcomings. It is paramount that the individual administering the protocol be able to recognize and interpret the various tests results within the context of these shortcomings (American Academy of Pediatrics and Joint Committee on Infant Hearing 2007).

ABR, OAEs, and tympanometry form the backbone of the diagnostic testing protocol at our institution for infants who fail newborn screening. Our protocols have been detailed previously (Buchman et al. 2006). ASSR has also been used at our institution, although we have found such testing adds very little diagnostic information beyond conventional ABR. In a manner similar to ABR, ASSR can over- or underestimate the degree of hearing loss with overestimation being more common (Stapells, Herdman, Small, Dimitrijevic and Hatton, 2005; Gorga et al. 2006; Ahn, Lee, Kim, Yoon and Chung 2007). ASSR is notably less accurate in patients with lesser degrees of hearing loss and is unable to identify those ears with the auditory neuropathy/dys-synchrony (AN/AD) phenotype (Stapells et al. 2005; Tlumak, Rubinstein and Durrant 2007). In subjects with profound hearing loss, ASSR may produce artifactual responses that are recorded as auditory thresholds when, in fact, they are produced by non-auditory centers (Gorga et al. 2004; Small and Stapells 2004). Thus, it is critical that clinicians understand that although these electrophysiological modalities are useful for estimating thresholds for the purposes of amplification fitting in children, they are not precise enough for making decisions re-

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**Figure 2.** Timeline for diagnostic and therapeutic interventions for infants that fail a newborn infant screen.
garding surgical intervention when destruction of the underlying residual hearing may occur as a sequela of that intervention.

The otologist’s role in caring for the hearing impaired child is to:

1. **Diagnose** hearing loss by identifying:
   - (a) Etiology and severity;
   - (b) Specific anatomical relationships to functional findings;
   - (c) Possible associated problems; and
   - (d) Referrals to related professionals.

2. **Treat** hearing loss by providing medical and/or surgical interventions;

3. **Referral** for amplification and/or speech therapy;

4. **Prevent** further hearing loss and other related complications through education of parents, children, and other health care providers; and

5. **Communicate with professionals** on the hearing loss team.

The otologist’s role usually commences once a hearing disorder has been identified. One exception might be the child who requires diagnostic audiology, but is either unable to be tested under natural sleep or sedation because of associated medical conditions or when it is apparent that middle ear effusions need to be addressed in order to garner accurate test results. These children are taken to the operating room for general anesthesia. In this setting, the ears are examined using the operating microscope and a determination regarding middle ear status is made. When fluid is present, we prefer to place tympanostomy tubes, and the audiological testing protocol is subsequently carried out. Ear canal bleeding must be avoided as this might negatively impact testing results. Placement of otic drops is deferred until after the auditory testing has been concluded. If a hearing impairment has been identified, ear canal impressions for future hearing aid molds are usually taken during the same setting for convenience. Findings are subsequently entered into the newborn infant hearing screening database.

Following the initial audiological assessment, we have found that a search for a hearing loss etiology and associated medical conditions is critical and frequently impacts the treatment paradigm. In North Carolina, referrals are also made to BEGINNINGS (www.beginningssvcs.com) and the Early Intervention Services Program (www.ncei.org) from the State at this juncture. BEGINNINGS was established as a non-profit organization, incorporated under the laws of North Carolina since 1987, to provide emotional support and access to impartial information as a central resource for families with deaf or hard of hearing children, age birth through 21 years. Early Intervention Services refers to a variety of public state agencies working together to provide services for children with special needs age birth to 3 years. Early Intervention Services is usually the mechanism whereby assistive technologies (AT) are funded and a speech and language pathologist (SLP) becomes involved in the child’s care plan. In North Carolina, there are a growing number of SLPs that are trained and certified in the methods of Auditory Verbal Therapy (AVT). Emerging evidence suggests that in the setting of an adequate auditory signal (either through amplification or a cochlear implant), auditory-based intervention provides for better acquisition of spoken language in hearing-impaired children than communication modes that include sign language (Moog and Geers 2003).

**Medical Diagnostic Evaluation**

The medical evaluation focuses on trying to identify an etiology for the hearing loss and associated problems that may negatively impact communication or other health issues. Implicit is the fact that a detailed understanding of the causes of hearing loss in children is needed to identify the salient issues in a particular patient. An excellent review of the potential etiologies of hearing loss has been previously published (Morton and Nance 2006). In addition to searching for the etiology of hearing loss, careful evaluation must identify disorders in vision, craniofacial malformations, and primary speech and auditory processing disorders to allow a comprehensive approach to the communication needs of a child and his/her family. Referrals among a variety of medical professionals are often needed.

Currently, available data estimates the prevalence of hearing loss at birth to be 186 per 100,000 and approximately 270 per 100,000 at 4 years of age (Morton and Nance 2006). Genetic factors appear to play a greater role than acquired causes at birth, while acquired cases become more prevalent later in life. There are more than 300 distinct hearing loss syndromes that have been identified by their association with other clinical features. Syndromic forms of hearing loss are less common, accounting for only 10–20% of new cases. However, a thorough understanding of these syndromes allows for relatively simple and rapid identification of the genetics and associated comorbidities that might affect a particular child. Non-syndromic (55%) and acquired (35%) forms of hearing loss are responsible for most of the newly identified cases.
cases emerging from the newborn hearing screening process and the etiologies can be somewhat more difficult or even impossible to identify (Morton and Nance 2006). For all types of hearing impairment in children, evaluation by a geneticist with expertise in pediatric hearing loss can provide additional information for families.

A careful history, physical examination and selective use of imaging studies and laboratory testing can identify the etiology of a child’s hearing loss in many cases. In addition to knowing the details of the newborn infant screening and diagnostic auditory testing, the **medical history** should be thorough in the areas of pregnancy and complications, past medical/surgical history, and family history. Some of the details that should be extracted include:

1. Did the pregnancy progress to full term? Were there associated complications such as eclampsia, fetal distress, oligo- or polyhydramnios, bleeding, Rh incompatibility, premature rupture of membranes, preterm labor?
2. Was there perinatal infection such as toxoplasmosis, herpes simplex, rubella, syphilis, or cytomegalovirus infection, Group B streptococcus, or other infection (TORCHes)?
3. Was antibiotic treatment required? If yes, were aminoglycosides used?
4. Was perinatal hypoxia or meconium aspiration evident at the time of delivery?
5. Did the child spend time in the neonatal intensive care unit (NICU)? If so
   a. What was his/her birth weight? (Below 1500 grams?)
   b. Was the child on a ventilator? What duration?
   c. Where high oxygen concentrations needed?
   d. Were there blood transfusions?
   e. Intracranial hemorrhage?
   f. Necrotizing enterocolitis? This might be associated with the use of aminoglycoside antibiotics.
   g. Retinopathy of prematurity
   h. Heart defects? If so what type?
6. Was the child jaundiced? If so, how high was the bilirubin concentration, for what duration and how was it treated?
7. Did the child have meningitis? Bacterial? If so, what organisms were present?
8. Did the infant require any surgeries?
9. What other medical disorders does the child have?
10. Are there difficulties with vision, feeding, or problems with other bodily functions?

11. In addition to the usual medical history regarding medicines, allergies, and past surgeries, it is important to assess other family members with hearing disorders and/or disorders related to hearing loss. Attempts should be made to uncover both first and second degree relatives that have or have had hearing loss.

The **physical examination** is focused on trying to identify syndromic features, associated ear-specific disorders, and anatomic situations that would adversely affect communication. While a detailed listing of these issues is beyond the scope of this manuscript, some specifics that are sought in the physical examination might include:

1. General appearance traits such as wide set eyes, pigmen
tary changes such as heterochromic irides and a white forelock of hair that are consistent with Waardenburg syndrome.
2. Cervical fistulas and pits with ear deformities suggesting branchio-oto-renal (BOR) syndrome.
3. Cleft lip/palate, down slanting eyes, coloboma, low-set small external ears, and mandible and maxillary hypoplasia in association with a conductive type of hearing loss would possibly indicate Treacher-Collins Syndrome.
4. Palatal and lip clefts in association with choanal atresia, external ear deformity, and facial paralysis might raise the suspicion for CHARGE association or similar syndromes.
5. Microcephaly might be seen in association with perinatal CMV or rubella infection or other events such as birth asphyxia or brain underdevelopment.
6. The general neurologic status of the child should be assessed. While this may not provide direct evidence for the etiology of the hearing loss, global neurologic control might influence the effectiveness of a variety of interventions.
7. Otitis externa and otitis media should be assessed in all cases as these can adversely affect precise hearing loss assessment and the institution of amplification.

**Radiographic Imaging**

Radiological imaging is a critical aspect of the assessment of every child with newly identified hearing loss. In our program, it is recommended immediately after the diagnosis of hearing loss has been established by electrophysiological measures. Early anatomical assessment of the temporal bones, auditory, vestibular and facial nerves as well as brain may:
(1) further characterize the hearing loss etiology,
(2) identify anatomical markers for hearing loss progression,
(3) predict poor prognosis from interventions such as amplification and/or cochlear implantation, and
(4) identify lesions of the central nervous system that require medical/surgical intervention for the overall health of the patient.

Classical studies of temporal bone and ear morphol-ogy have been carried out using histological/pathological techniques in a variety of conditions (Schuknecht 1993). Thus, the structural characteristics of many of the hearing loss syndromes have been described and can broadly be classified into those with or without radiographically detectable abnormalities. Patients with isolated inner ear cellular or membranous labyrinthine disorders are currently not identifiable based on current imaging resolution. Conversely, malformations of the external, middle, and inner ears and internal auditory canal (IAC) are clearly detectable using currently available imaging. Structural anomalies of the nerves of the IAC and brain are also resolvable in many cases. In general, high resolution computed tomography (HRCT) is well suited for assessing the osseous structures (external auditory canal and middle ear) while magnetic resonance imaging (MRI) provides excellent soft tissue detail for looking at the cranial nerves and brain. The inner ear is well visualized using either MRI or HRCT. HRCT shows the osseous labyrinthine shell well while MRI shows the fluids within the inner ear that conform to the otic capsule outline. The protocols that we use for these studies have been described previously (Adunka et al. 2006; Buchman et al. 2006; Adunka, Jewells and Buchman 2007).

There currently remains some debate regarding which of the various imaging modalities is most appropriate for assessing children with hearing loss (Parry, Booth and Roland 2005; Adunka et al. 2006; Buchman et al. 2006; Adunka et al. 2007; Trimble, Blaser, James and Papsin 2007). This controversy stems mostly from the familiarity of otologists and radiologists with interpreting HRCT for inner ear morphological changes. For cases of aural atresia and other conductive hearing losses, HRCT remains superior to MRI for assessing bony detail. Conversely, we prefer MRI rather than HRCT in all children with newly identified sensorineural hearing loss since it allows direct imaging of the cochlear nerves and brain. The consequences of missing either isolated cochlear nerve deficiency or unsuspected retrocochlear/brain pathology could be profound and might ultimately result in inappropriate treatment of the child. For example, cochlear implantation in an ear without a cochlear nerve or in an ear affected by a tumor could be devastating for the child and family. In cases of sensorineural hearing loss, we use supplementary HRCT only in cases where (1) semicircular canal defects are identified, so that the anatomy of the facial nerve is determined, (2) inner ear obstruction is evident on MRI, to further determine if the lesion is osseous or fibrous (post-meningitis), (3) the IAC is narrow, to determine patency of the bony cochlear nerve canal, and (4) temporal bone pathology has been identified such as in cases of tumors (Adunka et al. 2006; Buchman et al. 2006; Adunka et al. 2007).

Inner ear malformations that are detectable on imaging studies are common in children with SNHL. Some studies have estimated that 20–30% of children with SNHL have some morphological abnormality of their inner ear (Coticchia, Gokhale, Waltonen and Sumer 2006). They can be conveniently divided into abnormalities of the cochlear and/or vestibular apparatus or abnormalities of the neural structures. Cochlear abnormalities can take the form of aplasia (absence or Michel aplasia; figure 3), hypoplasia (small cochlea; figure 4), or dysplasia. Cochlear dysplasias are usually character-
ized as an incomplete partitioning as in the classical Mondini malformation (figure 5) or modiolar deficiency as in X-linked stapes gusher syndrome (figure 6). Vestibular morphological variants can also have aplasia, hypoplasia or dysplasias and can affect the semicircular canals, otolithic organs, and the vestibular aqueduct. Vestibular aplasia is shown in figure 7.

Children with inner ear or neural malformations may have an identifiable clinical syndrome. For instance, absent semicircular canals (figure 7) with or without cochlear hypoplasia and cochlear nerve deficiency are very common in children with CHARGE association, VATER syndrome, as well as BOR syndrome. An enlarged vestibular aqueduct (and endolymphatic duct) when seen in isolation or in association with an incompletely partitioned cochlea (Mondini’s deformity; figure 5) might indicate that Pendred’s syndrome is present. Children with Waardenburg’s syndrome might also have inner ear malformations along the Mondini spectrum of findings as well. A bulbous or dilated IAC that widely communicates with a deficient cochlear modiolus is suggestive of the X-linked stapes gusher syndrome (figure 6; Morton and Nance 2006).

Children with hearing loss can also have a variety of congenital and acquired changes to the central nervous system that are evident on imaging. Dandy-Walker syndrome is a congenital, developmental abnormality involving the cerebellum and posterior cranial fossa. The key features of this syndrome are enlargement of the fourth ventricle, a partial or complete absence of the cerebellar vermis, and cyst formation near the base of the skull. An increase in the size of the fluid spaces surrounding the brain as well as an increase in pressure may also be present (Paris and Dobyns 2003). Congenital CMV may result in microcephaly and/or cerebral calcifications and gliosis, which can be prominent in the temporal lobes. However, imaging findings can be difficult to distinguish from other viral infections of the cen-

Figure 5. Mondini malformation (cochlear dysplasia, large vestibule, small horizontal canal and enlarged vestibular aqueduct) as seen on HRCT (a), and MRI (c). A normal HRCT is shown in (b) for comparison.

Figure 6. Cochlear dysplasia (modiolus deficiency) on the left from X-linked stapes gusher syndrome compared to normal right.

Figure 7. Vestibular aplasia left and normal inner ear right.
Hearing Loss in Children: The Otologist’s Perspective

(8) eye examination/electro-retinography (Usher syndrome)

In general, an EKG, CMV assessment, and connexin testing are offered to all families while an eye examination with an ophthalmologist should be considered. While the Jervell and Lange-Nielsen syndrome is exceedingly rare, a properly performed EKG can identify

Laboratory Assessment

Laboratory assessment is dictated, in part, by the patient’s presenting situation. For children that have failed a newborn screening test and have undergone confirmatory testing that documents a sensorineural hearing loss (SNHL), testing might include:

- (1) electrocardiogram (EKG) [Jervell and Lange-Nielsen syndrome]
- (2) Guthrie card PCR for CMV infection
- (3) Connexin 26 and 30 mutation testing
- (4) VDRL (syphilis)
- (5) erythrocyte sedimentation rate (ESR), complete blood count (CBC)
- (6) rheumatoid factor (RF), antinuclear cytoplasmic antibody (ANCA), antinuclear antibody (ANA), anticardiolipin antibody,
- (7) renal ultrasound (BOR) and urinalysis (Alport’s syndrome)

Figure 8. Cochlear nerve deficiency (right ear) in a child that experienced sudden hearing loss. MRI (A) shows axial and parasagittal reconstructed images in a plane perpendicular to the IAC. In the left ear, four nerves (superior and inferior vestibular, facial and cochlear) are well-visualized. In the right ear, the cochlear nerve is absent. The arrow points to the normal cochlear nerve on the left and the region of the absent cochlear nerve on the right. HRCT (B) shows normal inner ear morphology and a satisfactorily placed cochlear implant in the ear without a cochlear nerve. The ABR (C) shows a distinct cochlear microphonic in the right ear and no response in the ear with a normal nerve. The cochlear implant in the right ear provided limited perceptual abilities and left cochlear implantation resulted in normal, open set speech perception.
AN/AD phenotype can result from a variety of lesions. Rather than a single lesion, it is plausible that the AN/AD phenotype can result from a variety of lesions throughout the auditory pathway. Mutations in a number of genes (MPZ, NDRG1, PMP22, OTOF, AUNA1) have now been characterized in hereditary cases of AN/AD (Varga et al. 2006). Associations have also been made between infectious (measles, mumps), metabolic (diabetes, hyperbilirubinemia, hypoxia), and neoplastic processes (acoustic neuroma) as well as prematurity (Rance et al. 1999; Starr et al. 2001). In most children with AN/AD, the cochlear nerve is known to be anatomically present because residual hearing abilities exist. Many of these affected individuals have varying levels of pure tone thresholds with disproportionately poor speech perception abilities. Since some children with AN/AD who have received cochlear implants have had robust electrically-evoked intracochlear compound action potentials (ECAPs) and good performance (Buss et al. 2002; Madden et al. 2002; Mason, De Michele, Stevens, Ruth and Hashisaki 2003), the abnormal hearing that some of these children have is thought to be due to disordered signal transduction at the inner hair cells, hair cell-dendrite synapse, or the cochlear neurons (i.e. auditory dys-synchrony) (Starr et al. 1996; Berlin, Morlet and Hood 2003; Berlin et al. 2003; Fuchs et al. 2003).

Opinions regarding management of children with AN/AD vary widely (see also Chapter 17 in this volume). When hearing loss has been documented, conservative amplification has been proposed (Rance et al. 2002). On the contrary, other investigators believe that the distorted speech perception abilities resulting from AN/AD precludes effective use of amplification (Berlin et al. 2003). Regarding the utility of cochlear implantation in children with AN/AD, limited data has demonstrated efficacy in many cases, implying that electrical stimulation may restore neural synchrony in some of these patients (Buss et al. 2002; Madden et al. 2002; Sininger and Trautwein 2002; Mason et al. 2003; Peterson et al. 2003).

At UNC-Chapel Hill, we established a prospective, institutional review board (IRB)-approved protocol five years ago to study the clinical characteristics and outcomes of children with newly identified AN/AD. The protocol was based on the premise that children with AN/AD can have a diversity of clinical characteristics and auditory abilities, suggesting that some might benefit from certain interventions while others might not. We believed that some children with AN/AD might have severely distorted perceptual abilities as described above while others might not. Thus, we chose to approach children with AN/AD individually, assessing their perceptual abilities with and without amplification.

Protocol Variation: Auditory Neuropathy/Dys-synchrony (AN/AD)

AN/AD is a clinical syndrome characterized by the presence of otoacoustic emissions (OAEs) and/or cochlear microphonics (CM) suggesting normal hair cell function in conjunction with absent or grossly abnormal auditory brainstem responses (ABRs) (Starr, Picton, Sininger, Hood and Berlin 1996). AN/AD is thought to account for up to 10% of newly diagnosed cases of hearing loss in children (Madden, Rutter, Hilbert, Greinwald and Choo 2002). Less than 10% of AN/AD cases are thought to involve only one ear. With bilateral presentation, patients exhibit a wide range of auditory capabilities. Hearing thresholds for pure tone detection can range from normal to profound levels (Madden et al. 2002; Rance, Cone-Wesson, Wunderlich and Dowell 2002). Recent studies in older children and adults suggest that these patients’ perceptual abilities can be severely impaired for both pitch discrimination in the low frequencies as well as temporal processing tasks (Rance, McKay and Grayden 2004; Zeng, Kong, Michalewski and Starr 2005). It has been hypothesized that lesions in the inner hair cells, the synapse between the inner hair cell and the auditory nerve, and the auditory nerve itself may account for the clinical findings (Starr et al. 1996; Berlin, Morlet and Hood 2003; Berlin, Hood, Morlet, Rose and Brashears 2003; Fuchs, Glowatzki and Moser 2003).

The etiology of AN/AD appears to be multifactorial. Rather than a single lesion, it is plausible that the AN/AD phenotype can result from a variety of lesions...
prior to considering cochlear implantation. While this assessment and management paradigm sounds remarkably similar to that previously described for children without AN/AD, some modifications were made in an attempt to better understand each child’s perceptual abilities prior to instituting amplification or cochlear implantation. This management paradigm has been discussed in previous publications (Zdanski, Buchman, Roush, Teagle and Brown 2006).

In our program, when a child is identified with the electrophysiological characteristics of AN/AD, the parents are told that their child has clear evidence of an auditory disorder. We also tell them that we cannot predict their child’s auditory thresholds or speech perceptual abilities and thus a period of careful observation ensues until an age when auditory thresholds can be determined. With highly experienced audiologists, thresholds are usually attainable using visual reinforcement audiometry (VRA) around 7 to 9 months corrected age. Should VRA demonstrate a significant hearing loss, a trial of amplification is instituted, based on real ear measures and DSL targets. Should amplification prove fruitless in the setting of ongoing diagnostic and therapeutic speech and language therapy, cochlear implantation is then considered. While this protocol does result in minor delays, we believe the approach is justified so that children who can benefit from amplification are identified and inappropriate cochlear implantation is avoided.

In multiply-handicapped children, it can sometimes be impossible to complete VRA successfully. For these children and their families, as well as the clinicians who care for them, rehabilitative decision-making can be very difficult.

We are currently following approximately 130 children with AN/AD, all of whom had diagnostic electrophysiological testing carried out at our institution similar to that described previously. What is clear from looking at this group of children is that AN/AD is a very heterogeneous group of disorders that presents with a common electrophysiological profile. Some children are profoundly affected by global and progressive neurological disease while others are otherwise completely normal. Some children have a genetic form that is similar among affected siblings while others have an association with prematurity, a stay in the NICU, hyperbilirubinemia, etc. When behavioral audiometry becomes possible, some children have near normal pure tone thresholds while others have a profound hearing loss. Following identification of pure tone thresholds, some children benefit from appropriately fit amplification similar to those children with “typical sensorineural hearing loss” while others go on to cochlear implantation. Most of the children that have gone on to cochlear implantation have had a severe-to-profound hearing loss and have met criteria for implantation based on conventional parameters. Very few children with AN/AD and less than a severe-to-profound hearing loss have received implants at our institution. Of this very select group of children with substantial residual hearing and AN/AD, decision-making on their behalf was very challenging for both the families and clinicians. In general, cochlear implantation has been very successful for most children with AN/AD although exceptions do exist. One group of children that provided significant insight into this disorder was those with anatomical cochlear nerve deficiency as detailed above (Buchman et al. 2006).

Medical Management of Hearing Loss in Children

The medical intervention for children with hearing loss depends on the type of hearing loss (conductive, sensorineural, mixed) and the functional-anatomical (or pathological) correlations that exist. Moreover, the desires and wishes of the family and child (in older children) are paramount. Patients and families not committed or interested in auditory-oral communication are by no means coerced in to hearing restorative interventions unless medical necessity dictates such treatment. While the details of every medical intervention for every type of hearing loss are beyond the scope of this work, some generalizations can be made which may act as a rough guide for families and professionals.

Behavioral Audiometry

In our program, behavioral audiometric testing continues to form the backbone for decision-making when medical/surgical intervention is being contemplated for a child. The shortcomings of both ABR and ASSR have been detailed previously. While these tests are clearly useful for estimating initial thresholds for the purposes of fitting amplification, the published literature and our experiences suggest that they are not precise enough for making decisions regarding irreversible procedures. Specifically, since cochlear implantation usually results in a loss of the native or residual hearing, we believe that an ear-specific behavioral audiogram using VRA is essential prior to considering cochlear implantation or other major ear interventions, although rare exceptions exist.
Hearing restorative medical interventions can broadly be classified into those that are disease-specific and those that are not disease-specific. Some examples of interventions that are not disease-specific include referral for hearing aid evaluation and fitting or surgical placement of hearing devices such as cochlear implants, bone conduction devices, or auditory brainstem implants.

**Cochlear implants** are reserved for those children with a severe-to-profound sensorineural hearing loss (> 90 dB) in the presence of an anatomically intact cochlea and cochlear nerve. These children should also be enrolled in an educational program committed to an auditory-oral approach and demonstrate limited progress with speech and language development while using appropriately fit amplification (http://www.nidcd.nih.gov/health/hearing/coch.asp). Since the hearing loss is severe to profound, high gain amplification is required and these devices must be fitted using real ear measurements and according to prescribed DSL targets. In addition to the auditory and speech criteria listed above, participation in such a program requires significant commitment by the family members. In the best scenario, children are identified following birth by a newborn infant hearing screening program. Following verification in the first month of life, the diagnosis is confirmed by way of electrophysiological testing methods described above and the trial of amplification is instituted. At the same time, the child is followed by a trained speech therapist with experience in auditory-oral approaches. By 6–9 months of age, behavioral audiometry confirms the degree of hearing loss and the therapist can provide feedback regarding auditory awareness and the development of the earliest vocalizations such as canonical babbling. When progress is evident, continued observation and therapy occurs. Conversely, when the child is making limited progress, cochlear implantation is considered with the goal of getting the device implanted around the end of the first year of life (figure 2). Factors that might delay cochlear implantation beyond the first year of life include: (1) delayed or inaccurate diagnosis, (2) delayed, under-fitted, or non-use of prescribed amplification, (3) lack of/or inappropriate participation in a speech/language therapy trial, (4) medical co-morbidities that delay diagnosis or preclude surgical intervention, (5) severe motor or cognitive developmental delays that hinder accurate auditory assessment, (6) diagnosis of AN/AD, and (7) lack of commitment by the family. Later ages at implantation are also expected in children with progressive hearing loss since these children achieve the severe-to-profound benchmark later in life and usually have better speech and language development because of their residual hearing.

Cochlear implant surgery takes roughly 90 minutes to two hours to perform at our institution. Following placement, intraoperative electrical telemetry is used to interrogate the device for integrity and to estimate a starting point for programming in the postoperative period. While complications are possible, they remain very unusual (Francis et al. 2008). Most children are implanted on an outpatient basis with anesthesia provided by a pediatric anesthesiologist. The children wear a head bandage for 3 to 5 days and return for a check approximately 1 week postoperatively. The device is usually activated 3 to 4 weeks postoperatively.

Outcomes following cochlear implantation in children are often truly remarkable. Most children demonstrate significant closed-set (using visual cues) speech perception ability after one year of usage. Open-set (without visual cues) speech perception skills are usually evident after two to three years of device experience. Figure 9 below demonstrates the results of open-set speech perception testing using the phonetically-balanced kindergarten (PBK; Haskins 1949) word test for a group of 315 prelinguistic children implanted in our program as a function of years of implant usage and age at implantation. It is evident from the graph that earlier implantation is better for developing this skill.

Are bilateral cochlear implants better than unilateral implants in children? Recent evidence suggests that bilateral implantation in postlingually deafened adults provides significant improvements for hearing in noise and sound localization abilities (Buss et al. 2008; Grantham, Ashmead, Ricketts, Labadie and Haynes 2007). In children, the data are only recently starting to

![Figure 9. Phonetically-balanced kindergarten (PBK) word scores as a function of duration of implant usage by age at implantation for 315 children with prelinguistic hearing loss. The dashed line indicates the scores for a group of children with normal hearing. In general, children implanted earlier ultimately achieve better scores at an earlier age than those children implanted later in life.](http://www.nidcd.nih.gov/health/hearing/coch.asp)
emerge but similar conclusions seem evident (Litovsky et al. 2006; Peters, Litovsky, Parkinson and Lake 2007). Only further research will determine whether there is a critical time window for developing these binaural skills in prelinguistic infants (see also Chapter 4 in this volume). Whether binaural implants will allow children to develop speech and language faster and to a higher level remains the critical unanswerable question. What is clear regarding binaural implantation is that having a second device provides a backup should there be trouble with the equipment in one ear. Should the device problems require surgery, having the backup or second side device will prevent unwanted time “off the air”.

Recent evidence suggests that children with sensorineural hearing loss with or without cochlear implants are at a higher risk for developing bacterial meningitis than the population in general (Biernath et al. 2006; Parner et al. 2007). It does appear from these data that cochlear implants impart an additional risk for contracting meningitis beyond that of having hearing loss alone. While the factors responsible for this increased risk have not been completely elucidated, one particular model of cochlear implant device that used a separate electrode positioner was implicated as predisposing to meningitis at a much greater rate than other models. This model has been removed from the market. The findings of these studies also prompted the United States Food and Drug Administration (FDA) and the Centers for Disease Control (CDC) to recommend the routine vaccination against the common bacteria implicated in cases of meningitis for all cochlear implant patients. Thus, *streptococcus pneumoniae* (i.e., pneumococcus) and *hemophilus influenzae* type B vaccination are indicated in all children with cochlear implants. The pneumococcal vaccines include the heptavalent conjugate vaccine (Prevnar®, Wyeth, Madison NJ) for children less than age 2 years and the 23-valent polysaccharide vaccine (Pneumovax®, Merck & Co, Whitehouse Station, NJ) after age 2 and again five years later (before age 10). The precise recommendations of the CDC and FDA vaccine programs are available on the world wide web (http://www.fda.gov/cdrh/safety/101007-cochlear.html). Currently, this is required prior to cochlear implantation in all children and adults in our program.

**Implantable bone conduction devices** are used in children when significant conductive or mixed hearing loss exists and direct reconstruction of the hearing apparatus is either not feasible or is not desired. This is relatively common in children with unilateral or poorly developed aural atresia. The currently FDA-approved system for this application is the BAHA (Cochlear Corp. Englewood CO, USA). It is based on the concept of implanting an osseointegrated fixture in to the skull to provide direct access to bone conduction of sound without the detrimental effects of soft tissue attenuation by the scalp. Low-gain amplification via osteo-oscillation is highly effective and very well tolerated. Implicit in the placement of such an implant is the need for a percutaneous connection, thus skin tolerance issues can be significant. Moreover, the skull of young children can be very thin precluding effective osseointegration. The BAHA is approved for use in children after age 5 years and is possible when skull thickness is sufficient (~3 to 4 mm). A preoperative CT scan can be useful in assessing skull thickness. The operation takes approximately one hour to perform and is usually outpatient surgery, although in children the operation is frequently performed in two stages. The device must heal for roughly 3 months prior to activation. It can be expected to provide auditory detection in the range of the bone conduction thresholds present on preoperative audiometric testing. The binaural benefits are only now becoming evident in congenitally affected patients (Kunst et al. 2008).

**Auditory brainstem implants (ABIs)** are currently only approved for usage in the United States for children with cochlear nerve deficiency resulting from neurofibromatosis type II. While the initial results of ABIs in children without NF2 are interesting, further data regarding the risks and potential benefits are needed (Colletti 2007).

Certain ear diseases that result in hearing loss can or must be addressed by way of disease-specific treatment of the underlying disorder. Examples of these disorders include: *otitis externa* and *otitis media*, congenital ossicular malformations and atresia, sensorineural hearing losses resulting from inflammatory conditions, and tumors of the temporal bone and brain.

**Otitis externa** is defined as inflammation of the external auditory canal and commonly results from bacterial or fungal infection. The hallmark signs and symptoms of this disorder are pain with auricular motion, minimal discharge, and conductive hearing loss when canal swelling is substantial. These findings should prompt a visit to a physician with experience treating ear disorders. It may follow ear canal trauma or contamination such as that which occurs from swimming pools or bathtubs. In some instances, a poorly fitted hearing aid mold might cause pressure on the ear canal skin, thereby predisposing to infection. Treatment requires removal of the offending agent (foreign body or hearing aid ear mold) when present and application of an otic
drop preparation containing either an acidifying or antimicrobial agent. When skin infection occurs outside of the ear canal, involving the pinna or surrounding soft tissues, an oral antimicrobial agent is also indicated. Patients with an immunocompromised state need to be treated particularly aggressively to prevent systemic infection and complications. Prevention in recurrent cases can usually be accomplished through careful cleaning and drying of hearing aid molds, removal of ear canal wax buildup, and prophylactic usage of drying agents (e.g., 50 : 50 mix of white vinegar and isopropyl alcohol) when the tympanic membrane is known to be intact (Buchman and Wambach 2002).

Otitis media is a primary inflammation of the middle ear space and commonly results from a bacterial and/or viral infection. Otitis media represents the most common ailment following the common cold for which a child will visit the doctor’s office, with a peak occurrence between the ages of 6 months and 2 years. Otitis media is commonly associated with episodes of upper respiratory tract infection and thus day care is a significant risk factor (Buchman and Das 2003). The signs and symptoms of otitis media include some degree of conductive hearing loss with or without associated ear pain, fever, nausea, irritability, cold symptoms, and ear drainage if the tympanic membrane has ruptured. During episodes, fluid is evident behind the intact tympanic membrane and signs of inflammation might be present. While the natural resolution rate is high, treatment is instituted to resolve pain more quickly, clear middle ear fluid, and prevent complications that at times can be severe and life threatening. Treatment of acute otitis media requires an oral antimicrobial agent and analgesics with follow-up to insure the clearance of middle ear fluid. In otherwise uncomplicated cases, persistent middle ear fluid can take more than 3 months to resolve. Thus, associated conductive hearing loss should be expected. Tymanostomy tubes are considered for middle ear ventilation and drainage in an effort to decrease or eliminate the duration and severity of otitis media episodes as well as to treat fluid-related hearing loss. Tymanostomy tubes are not routinely indicated for otherwise healthy children unless middle ear fluid persists for a prolonged period (> 3–4 months) and is associated with significant hearing loss (>30 dB) or when the severity of otitis media episodes are severe and disabling. However, when children have other associated communication disorders such as permanent hearing loss, developmental delays in speech and language, craniofacial disorders, immune system disorders, or anatomical changes in the tympanic membrane that can predispose to complications, tube placement is considered earlier (Paradise and Bluestone 2005).

Congenital conductive hearing loss can occur as a result of an isolated ossicular malformation or when external auditory canal atresia is present. For children with an isolated conductive loss with otherwise normal ear findings, stapes fixation represents the most common form of the disorder. Management of these children as infants is similar to those children with sensorineural hearing loss. That is, thresholds are determined and amplification is fitted as early as possible. When an ear canal lumen is not present bilaterally, a bone conduction hearing aid is applied to take advantage of the excellent speech discrimination abilities that these children usually have. When the child is older, reconstructive surgical options versus BAHA can be considered.

For a child with a congenital conductive hearing loss, a non-specific approach to the restoration of audition in the affected ear includes implantation of the BAHA device as described above (Kunst et al. 2008). For some families and children, the necessity for a percutaneous connector is unacceptable. Some payors will also not support this intervention because the price is considered prohibitive.

Another option for children with congenital conductive hearing loss is direct, surgical reconstruction. For the ear with an intact external auditory canal, this can be accomplished either through the canal or from a post-auricular approach. When stapes fixation is identified, a stapedectomy or stapedotomy is possible with excellent hearing results anticipated. When the malleus or incus is involved, hearing outcomes may be slightly worse. Prior to considering middle ear exploration for congenital conductive hearing loss, preoperative imaging is needed to rule out an inner ear malformation that might predispose to intraoperative cerebrospinal fluid leakage and further hearing loss. This surgery is probably best carried out when the child is very cooperative and can understand the risks associated with such a surgery. This usually occurs around the age of 8 years or older (Welling, Merrell, Merz and Dodson 2003).

Reconstruction can also be considered for children with external auditory canal atresia. Indications for this procedure might include (1) bilateral involvement, (2) the presence of a developmental temporal bone cholesteatoma in the affected ear; or (3) when observation of the BAHA are unacceptable to the family for the previously mentioned reasons. This procedure is major otological surgery and requires a very cooperative patient in the
postoperative period. The operation can take more than four hours and requires, in addition to an ear incision, a split thickness skin graft harvested from a site other than the head or neck (such as the leg or arm). Atresiaplasty is usually carried out after the time when the associated microtia surgery has been performed. Thus, most children undergo this surgery after the age of 7 or 8 years.

The results of atresiaplasty are quite variable and are dependent on the operating surgeon’s experience, the healing of the patient, and the patient’s anatomy. In general, roughly 75% of patients achieve a postoperative air bone gap of less than 30 dB. In those with a patent ear canal that is dry, a conventional hearing aid is also possible, thereby obviating the need for a bone conductor (McKinnon and Jahrsdoerfer 2002).

**Inner ear malformations** are present in 20–30% of children with congenital sensorineural hearing loss. These children can present in a variety of ways. While most will fail a newborn screen, some may not. Children with these anomalies clearly have a higher incidence of progressive hearing loss. It has been posited that sudden drops in hearing may be related to minor degrees of head trauma in these children. Many of these children will ultimately go on to cochlear implantation, which has a number of special considerations in this population (Buchman et al. 2004).

While there is currently no available method to repair developmental anomalies of the inner ear, a few facts are important for managing these children. First, it important for families to recognize that progressive hearing loss is common in these children. Moreover, children with inner ear malformations are at a higher risk for developing meningitis than children without malformations. Families need to be educated regarding the signs and symptoms of meningitis and about aggressive treatment of otitis media. In this regard, these children should also be considered for preventive vaccination similar to those children with cochlear implants. Finally, because of the association of sudden hearing changes and head trauma in these children, families are cautioned regarding participation in contact sports and activities.

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**References**


