Hearing loss in infants and children
An update on the evaluation and management paradigms

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The advent of universal newborn hearing screening and the subsequent monitoring efforts in many developed countries have led to a significant increase in the diagnosis of hearing loss in infants and children. Otolaryngologists are now routinely asked to evaluate and treat hearing deficits in children at a very young age. Alongside this new development is the substantial improvement in our knowledge of the genetics of hearing loss, which has led to the increasing use of genetic testing as part of the diagnostic paradigm. On the treatment front, advances in cochlear implant technology, along with enhanced awareness of the safety of implant surgery, have resulted in expansion of the cochlear implant candidacy to include children as young as 4-6 months. The goal of this article is to review how these new developments have changed the ways otolaryngologists evaluate and treat infants and children with hearing loss. The authors seek to outline the types of hearing loss commonly encountered in this group of patients, the relative merits and pitfalls of the clinical, radiological, laboratory, and genetic testing, and the indications and effectiveness of current medical and surgical treatment options.

KEY WORDS: Hearing loss - , Infant, newborn - Screening - , Genetic testing.

The evaluation and management of hearing loss has traditionally been an integral part of the practice of most otolaryngologists. However, several recent developments in the United States have significantly altered the ways otolaryngologists evaluate and manage pediatric patients with hearing loss. First, as state-

mandated Universal Newborn Hearing Screening (UNHS) programs are implemented across the country, otolaryngologists are expected to manage hearing loss in infants and very young children. Second, our understanding of the molecular basis of hereditary hearing loss has been significantly enhanced over the last decade. With increasing familiarity with the mutation screening process of specific deafness genes, many otolaryngologists are beginning to employ molecular genetic testing as part of the initial evaluation of children with sensorineural hearing loss (SNHL). Third, with increasing recognition of the impact of congenital hearing loss on language development, early intervention with surgical or non-surgical means are becoming the assumed standard of care.

In 2000, the Joint Committee on Infant Hearing (JCIH) and the American Academy of Pediatrics (AAP) endorsed revised UNHS guidelines, with a goal to achieve a screening capture rate of 95% prior to hospital discharge, hearing loss confirmation by 3 months of age, and intervention by 6 months of age. The impetus for the recommendations was rooted in the findings that children whose hearing losses were identified and treated before 6 months of age had receptive and expressive language quotients significantly high-

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er than children whose hearing losses were identified after 6 months of age.\textsuperscript{9,10} Other proven consequences of failure to detect prelingual hearing loss include personal-social maladjustments, emotional difficulties, and poor academic performance.\textsuperscript{11} Recognizing the urgency of this issue, legislatures around the country were receptive to the report and quick in enacting the recommendations. By 2004, legislation for UNHS was in place in 35 states. With these new mandates, otolaryngologists are increasingly called upon to provide accurate and timely diagnosis of hearing loss, and to deliver the essential treatments that would minimize the deleterious effects of untreated deafness.

In this article, the authors seek to review the types and causes of hearing loss that are commonly encountered in this patient population and the steps required for proper diagnosis. The relative merits and pitfalls of the clinical, radiological, laboratory, and genetic testing are reviewed in an effort to formulate an evidence-based paradigm. Finally, the indications and efficacies of both the surgical and non-surgical treatment options are discussed.

**Hearing loss in infants and children**

Moderate to profound hearing loss occurs in approximately 1-3 per 1,000 births in the United States.\textsuperscript{12,13} The European rates are similar, with ranges from 1.4-2.1 per 1,000.\textsuperscript{14,15} But the actual incidence is known to vary with time and place. For example, the last rubella pandemic in 1964 produced a significant spike in the incidence of congenital deafness.\textsuperscript{14} Conversely, the incidence is lower in areas with improved neonatal care and immunization programs, where many major causes of acquired prelingual deafness, such as rubella and Haemophilus influenzae type B, have been virtually eliminated.

In general, hearing loss can be characterized by three distinct descriptors: congenital versus non-congenital, hereditary versus non-hereditary, and syndromic versus non-syndromic. By definition, congenital hearing loss refers to a hearing deficit that is present at birth. It may or may not have a genetic/hereditary etiology. Likewise, hereditary hearing loss implies a genetic inheritance, and it may or may not be present at birth. Finally, "syndromic" means the presence of a group of abnormal features that share a common etiology. Most syndromic hearing losses are congenital and have a hereditary basis; however, exceptions do exist, which include fetal alcohol syndrome and the syndrome associated with prenatatal thalidomide exposure.\textsuperscript{16} With the overlapping and sometimes confusing descriptors, it is prudent to divide all types of hearing loss into subcategories in a systematic fashion. Figure 1 segregates the various known causes of pediatric hearing loss according to the 3 descriptors described above. Six separate categories/regions can be identified using the 3 descriptors.
Hereditary noncongenital nonsyndromic hearing loss

Region 1 represents the hereditary delayed-onset nonsyndromic hearing loss. The vast majority of the patients in this category are afflicted with autosomal dominant nonsyndromic deafness (ADNSD).\textsuperscript{15,17} Approximately 70% of all hereditary hearing loss are nonsyndromic.\textsuperscript{18} Of those, 10-20% are autosomal dominant (which usually has a delayed onset), and about 75% are autosomal recessive (which has an early onset, and will be discussed in the next section). Even though more than 40 genetic loci are currently known to be associated with ADNSD, no single locus has emerged as the dominant gene. The phenotype for ADNSD is typically a moderate bilateral asymmetrical SNHL below 4,000 Hz and normal high-frequency hearing. However, certain subsets of ADNSD patients may experience rapid progression and ultimately a profound loss across all frequencies.\textsuperscript{15} The inconsistency makes it difficult to correlate audiologic phenotype with a particular genotype. Consequently, no mutation-screening test is commercially available for the diagnosis of ADNSD. However, for patients with presumed ADNSD due to family history, it is still useful to construct a pedigree, which may aid in family counseling and in the future research of ADNSD.

Hereditary congenital nonsyndromic hearing loss

Region 2 represents the hereditary congenital nonsyndromic hearing loss. This group consists mainly of patients with prelingual autosomal recessive nonsyndromic deafness (ARNSD). This group represents about 75% of all nonsyndromic hearing loss. The first genetic locus for ARNSD, DFNB1, was mapped to chromosome 13q12.\textsuperscript{19} This gene was later identified as GJB2 (Gap Junction-Beta 2), which is responsible for encoding a protein called connexin 26 (Cx26) — an essential component of gap junctions.\textsuperscript{20} These gap junctions are found in the stria vascularis and the supporting cells of the cochlea, and are believed to play a significant role in the ionic homeostasis of the endolymph by allowing potassium ions transport after auditory stimulation.\textsuperscript{21} Since the defect is on a cellular basis, patients with ARNSD have no radiological abnormality of the temporal bone computed tomography (CT) scan.

Aside from the GJB2 gene, 33 additional loci have been localized, and allele variants of more than 15 genes have been related causally to ARNSD. However, GJB2 mutations were found to be responsible for up to 50% of the ARNSD in most of the United States and the European countries.\textsuperscript{15,20} The patients with GJB2-related deafness usually have bilateral severe (30%) to profound (50%) SNHL. Mild to moderate SNHL have also been reported (20%).\textsuperscript{15,22} Kenna et al.\textsuperscript{23} recently reported that 30% of their patients with sensorineural or mixed hearing loss of unclear etiology were found to have GJB2 mutation. They concluded that Cx26 mutations, especially the homozygous and compound heterozygous mutations, were likely the cause of the hearing loss. However, the pathogenicity was less certain where only a single Cx26 mutation was present. Similar results were reported by Preciado et al.\textsuperscript{3} who also pointed out that the positive rate for GJB2 screening was significantly higher in patients with severe to profound SNHL than in those with less severe hearing loss.

Hereditary syndromic hearing loss

Region 3 of Figure 1 represents hereditary syndromic hearing loss, which may be congenital or acquired. Currently more than 400 syndromes that involve hearing loss are known. This is a widely heterogeneous group, and the cause of the hearing loss may be conductive, sensorineural, or mixed. Since the hearing loss in these patients occurs in conjunction with a group of other features, early diagnosis may be possible. Craniofacial dysmorphism is often associated with hereditary syndromic hearing loss, which may often be the initial basis of referral to an otorlaryngologist. Familiarity with syndromic features associated with hearing loss may prompt hearing evaluation that could uncover subtle hearing loss that would otherwise be missed. Genetic counseling and treatment for the concomitant problems may then be provided if needed.

While most hereditary syndromic hearing losses are congenital in nature, the hearing loss or the syndromic features may be mild or are not readily recognizable after birth. An example is the Pendred’s syndrome, the most common form of hereditary syndromic hearing loss. Pendred’s syndrome is characterized by SNHL that is congenital in most cases and commonly severe to profound, although mild to moderate progressive hearing loss have also been reported.\textsuperscript{24} Bilateral enlargement of the vestibular aqueduct with or without cochlear malformed, along with either thyroid goiter or abnormal perichondral discharge test
complete the syndrome. Since thyroid abnormality is not present at birth, and temporal-bone abnormality is not a routine screening test, Pendred’s syndrome is seldom recognized in the neonatal period.

Other more common syndromes that are associated with SNHL include Waardenburg, Branchio-Oto-Renal, Alport, Jervell and Lange-Nielson, Usher and Biotinidase Deficiency. Common syndromes with prominent conductive hearing loss component are Apert, Treacher-Collins, Crouzon, and Goldenhar.16

Details and comprehensive reviews with up-to-date information on these syndromes may be found on several internet sites (Online Mendelian Inheritance in Man, http://www.ncbi.nlm.nih.gov/omim; and Hereditary Hearing Loss Homepage, http://web-host.ua.ac.be/hhh).

Interestingly, delayed-onset conductive hearing loss secondary to otitis media has been linked to several genetic syndromes, such as Down, velocardiofacial, and Turner syndromes.16

Nonhereditary syndromic hearing loss

Region 4 of Figure 1 represents the subgroup of syndromic hearing losses that are nonhereditary. Fetal alcohol syndrome,82 prenatal exposure to streptonycin, quinine, and thalidomide16,26,27 are all potential causes of syndromic hearing loss. Fetal alcohol syndrome is particularly common, and it refers to a pattern of anomalies that includes craniofacial, central nervous system, growth, and various sensory anomalies. The hearing disorders may include developmentally delayed auditory function, SNHL, intermittent conductive hearing loss owing to recurrent serous otitis media, or central hearing loss.25

A wide variety of prenatal infections can affect the fetus and result in syndromic hearing loss. The most common causes of hearing loss in this category are the maternal TORCH infections (toxoplasmosis, syphilis, rubella, cytomegalovirus, herpes simplex virus). In developing countries without a rubella vaccination program, congenital rubella syndrome, which consists of hearing loss, cataracts, and cardiac anomalies, remains the most prominent cause of nonhereditary congenital hearing loss.28 In most developed countries, the introduction of the rubella vaccine has greatly decreased the incidence of congenital rubella infection.

In the United States and Europe, cytomegalovirus (CMV) is generally recognized as the most frequent cause of nonhereditary hearing loss in newborns. In the United States, 40,000 CMV-infected infants are born each year. More than 4,000 of these infants have hearing loss detectable during infancy or childhood.29,30 Approximately 10% of all CMV patients also have systemic disease that include jaundice, hepatospleno-megaly, rash, intrauterine growth retardation, or respiratory distress. Half of the patients with systemic symptoms have SNHL, and most would expect progressive deterioration in their hearing.24 Also as important, approximately 8-10% of CMV-infected infants who were asymptomatic at birth could later develop some degree of SNHL. Therefore, long-term audiological follow-up is essential for all CMV-infected newborns.

Nonhereditary congenital nonsyndromic hearing loss

Region 5 of Figure 1 represents the nonhereditary nonsyndromic congenital hearing loss. Certain forms of TORCH infections, such as subclinical CMV, syphilis, and herpes simplex virus infections, may cause a hearing loss that are unaccompanied by other syndromic features. Isolated middle or inner ear anomalies can also occur without systemic defects. For example, isolated middle ear malformations, which include malformations or incus fixation, incudostapedial disconnection, and stapes fixation, have been reported in 1.2% of neonates with congenital conductive hearing loss.31,32 Temporal bone trauma during the delivery process can likewise lead to conductive hearing loss at birth.

Transient conductive hearing loss in neonates may be a result of persistent amniotic fluid in the middle ear. Moderate amounts of amniotic fluid have been found in up to 77% of neonates, and it may persist for up to 243 days postpartum.33 Such transient hearing loss may result in false positive screening after birth.

Nonhereditary noncongenital nonsyndromic hearing loss

The last category (Region 6) shown in Figure 1 is the nonhereditary acquired hearing loss. The most common causes of acquired hearing loss in infants and children are acute otitis media, otitis media with effusion, and chronic otitis media.34 Such hearing loss is typi-cally mild to moderate in severity and is conductive in nature. The hearing loss is reversible with treatment of the underlying condition.
The most common cause of acquired SNHL in infants and children is bacterial meningitis. Approximately 6% of all cases of SNHL in children can be attributed to bacterial meningitis, with *Pneumococcus* and *Haemophilus influenzae* as the most common pathogens. The use of vaccines against *Streptococcus pneumoniae* and *Haemophilus influenzae* type B and the administration of steroid therapy early in the disease course have decreased the incidence and the sequelae of these infections. Postmeningitic hearing loss can be either unilateral or bilateral, but commonly bilateral loss is observed.

Several perinatal conditions are considered high-risk predictors of hearing loss. Among the most significant are birth weight less than 1.5 kg, mechanical ventilation lasting 10 days or longer, and hyperbilirubinemia at levels requiring exchange transfusion. All 3 factors are commonly associated with neonatal ICU patients, thus underscoring the importance of early hearing screening in the NICU setting. Interestingly, hyperbilirubinemia produces a neural hearing loss, as evidenced by abnormal brain stem responses (ABRs), that can often be reversed with exchange transfusion and establishment of a normal bilirubin level.

Another common cause of acquired SNHL is the administration of ototoxic medications such as aminoglycoside antibiotics, diuretics, and platinum-based chemotherapeutic agents. The ototoxicity of these drugs is closely related to a prolonged administration and higher total doses, particularly aminoglycosides and furosemide. The resulting damage to the cochlear hair cells and the stria vascularis are permanent, thus the hearing loss is typically irreversible.

### Evaluation of hearing loss in infants and children

With the myriad causes of hearing loss in infants and children, a systematic approach to the evaluation process is critical. A careful and thorough history and physical examination could provide clues that assist otolaryngologists in differentiating between congenital *versus* acquired, hereditary *versus* nonhereditary, and syndromic *versus* nonsyndromic types of hearing loss. Based on the discussion presented in the previous section, the differential diagnosis could be narrowed, and a more focused selection of laboratory tests, radiographic imaging, and genetic testing could be ordered.

### History

A complete history that includes the prenatal, perinatal, and postnatal periods should be elicited from the mother or the primary caregiver. Prenatal information includes the gestational age at birth, maternal teratogenic exposure, medications taken during the pregnancy, and gestational illnesses. Any TORCH screening results should be verified. Specific conditions during the perinatal period that could predispose the newborns to hearing loss, such as low birth weight, prolonged mechanical ventilation, hyperbilirubinemia, hypoxia, and birth trauma should also be inquired.

The postnatal childhood history could reveal problems related to either congenital or acquired hearing loss. Childhood infections such as meningitis, measles, mumps, rubella, and encephalitis are all common causes of hearing loss. Conductive and SNHL after acoustic and temporal bone trauma have been reported. A history of syncope should prompt an evaluation to rule out prolonged QT interval that implies Jervell and Lange-Nielsen syndrome. Other histories of childhood illnesses should be sought to rule out syndromic causes of hearing loss. Information regarding the use of specific ototoxic medications, such as aminoglycoside, loop diuretics, cisplatin, and retinoic acid can help establish the etiology of the hearing loss.

A thorough family history could provide insights into the possible hereditary nature of the hearing loss. If there are multiple family members with either early-onset hearing loss or severe to profound hearing loss, analysis of the pedigree may define the mode of inheritance. Finally, for infants and young children, the developmental history that includes hearing-specific developmental milestones could be compared to age-associated norms to determine the onset of hearing loss.

### Physical examination

Most patients with hearing loss do not have gross physical abnormality. Nonetheless, a complete physical examination is crucial to avoid missing features that may suggest syndromic hearing loss or congenital malformation. The physical examination should begin with a thorough head and neck examination. Any otorhinolaryngologic abnormalities, such as microtia or atresia, congenital cholesteatoma, middle ear effusions, or tympanic membrane anomalies should be noted. Craniofacial dysmorphic features should be docu-
mented. The otolaryngologist should also examine the eyes for coloboma, hypertelorism, or other abnormalities. It has been suggested that an ophthalmology consult is very helpful in the evaluation process, as nearly half of severely profoundly deaf children have ocular abnormalities. The majority of the findings are consistent with simple refractory errors, but its correction is often needed to aid in auditory rehabilitation. The neck should be examined for branchial abnormality or thyroid enlargement. Oral cavity examination may reveal cleft palate or submucous clefting, which predispose the patient to chronic Eustachian tube dysfunction and conductive hearing loss secondary to otitis media.

A general examination includes inspection of the skin for areas of pigmentation changes or café-au-lait spots, which is related to neurofibromatosis. Abnormal number, size, and shape of the digits are suggestive of Alpert’s syndrome. Unusually small patients may have a history of prematurity or intrauterine growth retardation, while very tall patients may have an underlying Marfan or Stickler syndrome.

**Laboratory testing**

Multiple studies have found that if the etiology of hearing loss is not apparent after a complete history and physical examination, routine laboratory testing is unlikely to yield the diagnosis. Blood tests used by Mafong et al. to assess autoimmune, blood dyscrasias, endocrine abnormalities, renal function, and infection did not yield a diagnosis in 114 consecutive children with sensorineural or mixed hearing loss. Only one case of Jervell and Lange-Nielsen syndrome was diagnosed by prolonged QT interval on electrocardiogram. Similar results and conclusions were reached by Preciado et al., who studied 650 children with SNHL of unknown etiology.

The evidence thus suggests that a laboratory “shotgun” approach to diagnosing hearing loss should be discouraged. Instead, choosing the specific test based on clinical impression may be the more prudent course. For instance, electrocardiogram should be ordered for a patient with history of unexplained syncope or family history of sudden death. Likewise, laboratory tests for other syndromic hearing loss should be ordered only on the basis of history and physical examination findings.

If intrauterine infection is suspected, IgM antibody assays can be obtained. An elevated IgM level is highly suggestive of intrauterine infection because maternal IgM does not cross the placenta. If positive, the serial titers should be followed by a pediatric infectious disease specialist for further evaluation and treatment.

**Radiographic imaging**

Unlike the laboratory testing, radiologic abnormalities of the inner ear are common in patients with hearing loss of unknown etiology. Among that patient population, 29-39% of the CT and magnetic resonance (MR) imaging of the temporal bone have been abnormal. Large vestibular aqueduct was the most common finding. Other noted abnormalities include cochlear dysplasia, common cavity deformity, lateral semicircular canal dysplasia, and small internal auditory canal. Of particular interest, the diagnostic yield was significantly higher in children with unilateral SNHL than bilateral hearing loss (37% vs 25%).

Among the imaging modalities, high resolution CT scan is the test of choice for the evaluation of patients with craniofacial anomalies, conductive hearing loss, and unilateral SNHL of unknown etiology. As will be discussed in the next section, patients with bilateral SNHL may benefit from genetic testing for CX26 mutations as the initial diagnostic evaluation. Since patients with homozygous CX26 mutations have a microscopic abnormality that is not visible on imaging studies, CT scanning should only be used for patients with negative genetic test results. CT scan is also needed for pre-surgical planning and surgical decision making in temporal bone surgery, such as for cochlear implantation or congenital aneryxia repair. It helps to delineate the course of the facial nerve, and to check for cochlear patency and middle ear anatomy.

MR imaging complements CT scanning by providing superior images of the cochlear, facial, and vestibular nerves traveling in the internal auditory canal, as well as showing greater resolution of the endolymphatic sac and duct. It is indicated in cases in which neurofibromatosis or retrocochlear pathology is suspected, or if the hearing loss is progressive but the CT scan is normal. Some authors also advocated the use of MRI as part of the pre-operative evaluation for cochlear implantation, as it provides superior information regarding cochlear patency in post-meningitic patients when compared to the CT scan.
Genetic testing

Prior to the availability of genetic testing, many patients with nonsyndromic SNHL remained undiagnosed even after undergoing a careful review of the medical history, a thorough physical examination, and preliminary laboratory and imaging studies. Molecular genetic testing promises to improve the diagnosis rate by unlocking the genetic information associated with previously unrecognized hereditary hearing loss.

The discovery that mutations in the GJB2 gene are responsible for nearly half of all cases of autosomal recessive nonsyndromic hearing loss has allowed for the creation of an excellent screening test. One specific mutation, the 35delG, was found to account for as high as 70% of all CX26 mutations in the United States and several European countries. Several other mutations of CX26 have been noted to be highly prevalent in other populations, including the 167delT mutation in the Ashkenazi Jews and the 235delC mutation in the Japanese.

The most common phenotype for a child with CX26 abnormality is bilateral severe to profound SNHL. This phenotype is commonly linked to 35delG homozygotes. Milder levels of bilateral hearing loss have been attributed to 35delG heterozygotes, 35delG compound heterozygotes, or more recently a digenic inheritance of GJB2 and GJB6 (a Cx30 gene) mutations.

Of particular importance, the incidence of children identified with a GJB2 mutation having other concurrent diseases is exceedingly rare. It is therefore recommended that all patients with bilateral nonsyndromic SNHL of uncertain etiology should initially undergo GJB2 screening. If mutations are identified, additional testing can be avoided. Additional evaluations, such as high resolution CT scan or clinically-directed laboratory studies, are ordered only if the screening is negative.

Genetic screening for other forms of hearing loss is also becoming increasingly available, although mainly in research settings. For example, Pendred syndrome, the most common hereditary syndromic hearing loss, can be diagnosed by screening for mutations in the SLC26A4 gene. It is safe to predict that over the coming years, the number of genetic tests available will increase substantially, thus having a major impact on the diagnosis and management of hearing loss in infants and children.

Management of hearing loss in infants and children

The universal newborn screening programs were founded on the premise that early detection and intervention (screening by 1 month, confirmation by 3 months, intervention by 6 months) could maximize the development of auditory skills and facilitate spoken language development. Once a hearing loss is suspected or confirmed, the patient is often referred to an otolaryngologist for further evaluation and treatment. The initial evaluation process should proceed as discussed above, but the eventual treatment options depend largely on whether the hearing loss is conductive or sensorineural in nature.

Conductive hearing loss

For the most common cases of conductive hearing loss secondary to otitis media, placement of pressure equalization tubes are indicated if the middle ear effusion persists for more than 3 months. The procedure is usually curative with near complete restoration of hearing. Other common middle ear pathologies that could result in conductive hearing loss, such as tympanic membrane perforation, cholesteatoma, and chronic otomastoiditis, are treated with tympanoplasty with or without mastoidectomy. If a patient is not a surgical candidate, does not desire surgery, or has significant post-operative residual conductive hearing loss, a hearing aid should be recommended. Most patients with conductive hearing loss derive significant benefit from well-fitted hearing aids.

Children with congenital anatomic anomalies such as aural atresia and ossicular malformations could be reconstructed with a variety of surgical techniques. Various ingenious middle-ear prostheses made with biocompatibility materials such as titanium and hydroxyapatite have been used, and the reported success rates are generally good. An alternative to reconstructive surgery is the use of bone-conduction hearing aids. An implantable version of the hearing aid, the bone-anchored hearing aid (BAHA), has been gaining popularity, especially for slightly older children with permanent conductive hearing losses who could not tolerate or do not gain benefit from conventional hearing aids.

Sensorineural hearing loss

There are few surgical or medical treatment options
available for children with SNHL. For these patients, a trial of hearing aids or frequency-modulation device is recommended. Aggressive speech and language therapy should also be offered. If the assistive hearing devices are of limited help, such as for children with bilateral severe to profound SNHL, the parents are faced with the difficult decision of choosing a communication mode for the patient. Currently, there are 3 general categories: visual based sign languages, auditory-oral communication (e.g. auditory verbal training, lip reading, and cued speech), and total communication (a combination of sign and speech). Intensive speech and language therapy is needed for the patient to be proficient in any of the 3 communication modalities.

Thus far, the only surgical treatment available for the bilateral severe to profound SNHL patients is cochlear implantation. A cochlear implant is an electronic prosthetic device that is surgically implanted in the temporal bone. It functions by selectively stimulating a portion of the spiral ganglion cells of the cochlea in accordance with the frequency of the incoming acoustic signals, thereby generating useful sound perception. The early users of cochlear implants were mostly postlingually deafened adults, but with continuing improvement in cochlear implant technology and surgical techniques, the current candidacy criteria have been expanded to include children 12 months of age. At certain centers, many infants as young as 4-6 months have been safely implanted, and the children have generally enjoyed excellent long-term audiologic outcomes.

Several studies have shown that early cochlear implantation is related to improved oral language outcomes. It was found that implantation before the age of 2 resulted in speech perception and language advantages that were significant both statistically and from a practical point of view. With the expansion of UNHS, cochlear implantation will undoubtedly play an increasingly prominent role in helping restore hearing to many newly diagnosed infants and children with significant hearing loss.

Riassunto

Perdita di udito nei neonati e nei bambini: un aggiornamento sulla valutazione e gestione dei paragoni

L’avvento universale della valutazione preventiva dell’udito nei neonati e i conseguenti sforzi di monitoraggio ha portato in molti Paesi sviluppati un significativo aumento della diagnosi di perdita di udito nei neonati e nei bambini. Attualmente agli otorinolaringoiatri viene richiesto di valutare e di trattare i deficit di udito nei bambini. Accanto a questo nuovo sviluppo è il miglioramento sostanziale della nostra conoscenza della perdita di udito su base genetica che ha consentito di aumentare l’uso dei test genetici quali parte del paradigma diagnostico. Sul fronte del trattamento, i miglioramenti della tecnica di impianto coclea, assieme con l’aumentata consapevolezza della sicurezza dell’impianto chirurgico, hanno portato a un’espansione del numero di candidati a questo tipo di intervento, includendo i bambini con 4-6 mesi di età. L’obiettivo di questo articolo è rivedere come questi nuovi sviluppi abbiano causato le modalità con le quali gli otorinolaringoiatri valutano e trattano i neonati e i bambini con perdita di udito. Gli Autori cercano di sottolineare i tipi di perdita di udito comunemente presenti in questo gruppo di pazienti, i meriti e i difetti dei test clinici, radiologici, laboratoristici e genetici e le indicazioni e l’efficacia delle attuali opzioni mediche e chirurgiche.

Parole chiave: Perdita dell’udito - Età pediatrica - Screening - Test genetici.

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