Chapter 6
The Etiologies of Childhood Hearing Impairment

N. Wendell Todd, MD, MPH

The purpose of this chapter is to explore the causes of a child's hearing impairment. We suggest a practical, organized approach to attending a baby with presumed hearing impairment.

Often one consultation with a specialist is not sufficient to determine the cause of a hearing impairment. Etiology is often enigmatic.

Why Be Concerned about the Cause of Hearing Impairment?

- Knowing if anything can be done to resolve the hearing impairment. Early focused treatment of some congenital infections is quite effective.
- Knowing if anything can be done to keep the hearing impairment from progressing.
- Understanding which modes of communication may be practical for the child.
- The level of concern for the hearing health of other family members.
- Scientific study to understand mechanisms of hearing.

Often one consultation with a specialist is not sufficient to determine the cause of a hearing impairment. Etiology is often enigmatic. Determining why a child has a hearing impairment is most feasibly done as part of the child's ongoing care, since it is rarely a one-time process. The often-elusive explanation for a child's hearing impairment is all-important for that particular child. Additionally, it is vital to know which portions of the auditory system are malfunctioning and to take into consideration that the underlying explanation may not lie in the common group of causes of hearing impairment.

Categorizing permanent childhood hearing impairments is helpful in guiding the diagnostic process (see *Table 1*).

Succinct Background

About 3 in 1,000 children are born with some degree of permanent hearing loss, with about 1 in 1,000 coming from the well-baby nursery and about 1 in 100

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Table 1
Permanent Childhood Hearing Impairment Categories

Conductive, sensory, neural, or mixed	Congenital or acquired	Syndromic or non-syndromic
Only a battery of audiologic assessments will enable accurate determination of the type of hearing impairment.	Delayed diagnosis—considered by some to be the result of false negative auditory physiologic newborn screening or inattentive parents or doctors—may actually be the result of genetic problems that manifest after the first few months of life. Some patients with anatomic inner ear differences anomalies (recognizable with CT or MRI scanning) do not manifest hearing impairment in the first few years of life.	Although the combination of deafness plus syncope and long Q-T interval default to the label of Jervell Lange-Nielsen syndrome, and deafness plus white forelock and heterochromia lead one to the label of Waardenburg syndrome, most patients with syndromic hearing impairment are recognized only upon delving further into "secondary" symptoms. The heralding symptoms of Usher syndrome, beyond deafness, are easier noted in retrospect than in real time. More than 400 genetic syndromes that include hearing impairment have been described (Smith et al., 2014). Yet, most children with hearing impairment do not have an underlying syndrome (or at least one that is not known).

Unilateral or bilateral or asymmetric vs. symmetric	Fluctuating or non-fluctuating	Degree or hearing impairment
Bilateral auditory impairment is more communicatively hampering than unilateral impairment. When compared to normal hearing children, those with unilateral impairment suffer worse oral language scores (Lieu, Tye-Murray, Karzon, & Piccirillo, 2010) in addition to increased rates of academic failure and problems with sound localization.	Many hearing impairments are stable over months and years. Three impairments, however, characteristically fluctuate over relatively short periods of time: perilymphatic fistula, large vestibular aqueduct syndrome (Mori, Westerberg, & Atashband, 2008), and auditory neuropathy spectrum disorder (Rance et al., 1999).	Mild, moderate, severe, or profound.

Photo courtesy of NCHAM

Types of Hearing Impairment

Compartmentalizing hearing impairment determines the region(s) of the auditory system that are affected.

Approximately 15% of genetic hearing loss is part of a syndrome; hence, the recognition of hearing impairment may be the initial clue to a more involved diagnosis.

1
Conductive hearing impairment

The problem between the external ear and the cochlea. A common, often spontaneously resolving etiology in about one-fourth of newborns less than age 48 hours is vernix in the external ear canal (Doyle, Rodgers, Fujikawa, & Newman, 2000). Middle ear effusion bilaterally was present in about 12% of babies. Otitis media in the first 2 months of life heralds an increased risk of otitis at least during infancy. The most common cause of congenital maximum conductive hearing impairment (i.e., thresholds approximating 60dBHL) is aural atresia. The child develops without an external ear canal. Overall, ear atresia occurs in about 1 in 10,000 births; approximately 15% of cases are bilateral. Though about 20% of patients with congenital aural atresia have inner ear anomalies identifiable with computed tomography, a minority has sensorineural hearing impairment—usually mild (Vrabec & Lin, 2010).

2
Sensory hearing impairment

The problem is in the cochlea. Causes include genetics, infections, ototoxins, hypothyroidism, and leukemia. Discovery of genetic causes of hearing loss has been rapid in recent years. Approximately 70-80% of genetic hearing loss is autosomal recessive, 15-20% is autosomal dominant, and 2% is X-linked or mitochondrial. Understanding a family's history of hearing loss is essential in determining if a child's hearing loss is genetic in origin. Approximately 15% of genetic hearing loss is part of a syndrome; hence, the recognition of hearing impairment may be the initial clue to a more involved diagnosis. Down syndrome, Usher syndrome, and Waardenburg syndrome are just a few examples. Currently over 400 syndromes involve deafness as one of their defining traits. Infectious causes of sensory hearing impairment are also numerous, though less common in the United States than in Second or Third World countries. Cytomegalovirus (CMV), rubella, syphilis, and toxoplasmosis can all cause moderate to profound hearing loss, both unilaterally as well as bilaterally. CMV characteristically manifests in the months after birth.

3
Neural hearing impairment

Is attributable to a failure of the neural portion of the auditory pathway. The auditory nerve can be implicated in hearing loss by failure of the nerve to be present (aplasia) or smaller than normal (hypoplasia). The nerve itself may be normal in appearance at imaging (e.g., MRI) but not functioning due to genetic or environmental factors, such as metabolic stress, especially in the child born prematurely. Rarely, a tumor, such as a schwannoma, can be present along the auditory nerve and render it nonfunctioning. Congenital malformations, toxins (e.g., hyperbilirubinemia), bleeding, and infections (e.g., syphilis, CMV, toxoplasmosis, and rubella) may affect the auditory pathways in the brain.

4
Combinations of the types of hearing impairments

Combinations do occur and are termed "mixed" type of hearing impairment.

from the neonatal intensive care unit. The prevalence of hearing loss increases to 6 in 1,000 by the time children begin kindergarten. In the United States, overall, about half of the children with hearing impairment have a genetically determined explanation—some manifesting after the newborn period. Regardless of the cause of the hearing impairment, early detection and intervention enable the child to have an increased likelihood of better communication, better performance in school, and a better quality of life.

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Congenital vs. Postnatal Hearing Impairment

Numerous factors can contribute to auditory problems that occur after birth. While congenital hearing loss affects 3 in 1,000 births, at least the same number of children acquires hearing impairment over the next several months of life. As with congenital hearing losses, late onset hearing impairments can be categorized based on the location of the problem. Although there are few solutions to "cure" or completely reverse congenital hearing impairment, the prognosis for some acquired postnatal impairments is often more encouraging. Return of function is sometimes an option. The last millennium's line dividing congenital and acquired hearing impairments was easy and logical. Delayed recognition of genetic congenital etiologies was often blamed on nonobservant parents or hurried practitioners. The delayed presentation of hearing impairment at 11-60 months of age in cases of biallelic *GJB2* is now well documented (Tranebjærg, 2008). Postnatally manifesting hearing losses are sometimes classified as delayed onset, progressive, and acquired (Weichbold, Nekahm-Heis, & Welzl-Mueller, 2006).

Postnatally manifesting conductive hearing loss has a number of causes. These can best be understood when divided into those external to the tympanic membrane (TM) and those medial to TM but in front of the cochlea. Causes in the external ear canal include impacted cerumen, foreign body, and otitis externa. A rare cause is a tumor in the external ear canal. Impacted cerumen is often the result of an over-anxious parent or caregiver attempting to "clean" the external canals with Q-tips or a similar object. This action, rather than removing the cerumen, actually impacts it further into the canal in an area where the ear cannot remove it through its own self-cleansing mechanisms. Foreign bodies are often a result of an overzealous child attempting to understand their own anatomy. Otitis externa is a painful inflammation of the external ear often caused by bacterial overgrowth.

Acquired conductive hearing loss (CHL) can also be attributed to a hole in the TM created by a previous ear tube, an episode of otitis media, or by manipulation of a foreign body. Otitis media, cholesteatoma, and head trauma are the most common causes of acquired conductive loss medial to the TM. Otitis media is inflammation in the middle ear. Cholesteatoma is a tumorous accumulation of skin-like debris in the middle ear space. Finally, head trauma can cause the ossicles to disarticulate.

Sensory hearing loss can manifest in the months and years after birth. Causes include a genetic origin, severe metabolic disturbance (e.g., with diaphragmatic hernia or ExtraCorporeal Membrane Oxygenation [ECMO]), ototoxic agents, infectious etiology, trauma, or noise (Dedhia, Kitsko, Sabo, & Chi, 2013). Genetic hearing loss can manifest after birth in approximately 50% of hearing-impaired children. It is not unusual for a child to pass newborn hearing screening and subsequently fail another exam at a later date. Children with genetic hearing loss can progressively lose their hearing throughout their first several months of life, either prelingually (prior to acquisition of speech) or postlingually (after acquisition). These hearing impairments are discovered by incidental screening, from an observer raising concern about the child's hearing, or the loss of previously utilized speech. Ototoxic agents, such as aminoglycosides and cisplatin, can also result in sensory hearing loss. The severe end-result of cochlear inflammation is labyrinthitis ossificans in which the cochlear channels turn into concrete-like structures. The resulting sensorineural hearing loss

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cannot routinely be well addressed by a cochlear implant. These infectious sources can also be categorized as a neural cause of acquired hearing loss, since they can also directly affect the cochlear nerve. Trauma resulting in temporal bone fractures can result in a sensory hearing loss, especially if the fracture passes through the otic capsule (bone surrounding the cochlea). Finally, noise trauma can cause mild to profound hearing loss depending on the insulting agent. Up to 15% of children ages 6-18 years have some acquired hearing loss from noise trauma, such as loud music.

Postnatal manifestations of neural hearing loss have three main causes: trauma, infection, and tumor.

VATICINATE

With childhood hearing impairment having half-dozen categories and hundreds of causes, an organized system is best used in addressing the needs of the patient and family. The mnemonic VATICINATE has been employed as a standardized method of working-up and caring for a patient with hearing impairment—whether it be for a 2-day, 2-month, or 2-year-old child or an adult. Vaticinate—a real word—means to sing or foretell about the future. Ironically, if diligently and repeatedly applied, it can help foretell the future of a child with hearing impairment.

V Verify That a Hearing Impairment Exists

- Was an age-appropriate battery of tests as outlined in the Joint Committee on Infant Hearing (JCIH) 2007 Position Statement used?
- Does the compilation of information make sense? Are the results of the various auditory physiologic and behavioral tests consistent with one another?
- Do family members agree that a hearing problem exists? Has a family member's observations brought into question the validity of the audiologist's findings? Did the family interpret the audiologist's work as sloppy?
- Do fluctuations in the patient's hearing explain the discrepancy? Auditory abilities sometimes fluctuate, day to day, in patients with otitis media, auditory neuropathy spectrum disorder, perilymphatic fistula, and some inner ear dysplasias, such as:
  - Incomplete partition of the cochlea.
  - Large vestibular aqueduct syndrome.
  - X-linked fixed stapes gusher syndrome.
- Is a family member considering the responses to low-frequency vibrations or visual stimuli (e.g., elicited by clapping hands or slamming doors) hearing?
- Does the infant babble? Does the child talk? Does the speech include sibilants?

Until there is verification that a hearing problem exists, any family is unlikely to comply with recommendations.

Do not accept a dB without a reference. The dB is a logarithmic expression of an output divided by a reference value, so a dB without a reference is not a dB—remembered by contemplating a line in the song "Casablanca," written and sung by Bertie Higgins: "A kiss is not a kiss without your sigh." In hearing measurements, the dB is typically referenced to hearing level (HL) or sound pressure level (SPL). Be aware that dB involving pressure (acoustical energy is expressed in pressure) has the logarithmic expression squared, but that dB involving other entities (e.g., electricity) does not have the logarithmic expression squared.

Any discussion of dB levels of hearing or hearing impairment is very likely to be confusing for parents. If the professional is going to quantify the hearing in dB rather than more functional terms the family can understand, he/she must be prepared to offer a relatively detailed description of the audiogram. For example, a hearing threshold of 30 dB SPL is very different than a hearing threshold of 30dB HL.

A Amplify: Hearing Aids

- If the family concurs, refer to an audiologist for fitting of hearing aids. Obtaining insurance approval and custom earmolds takes time. The parents may need gentle guidance to move from acceptance of the hearing impairment to amplification considerations.
- If the baby has profound hearing impairment, amplification through the use of hearing aids can provide vibro-tactile stimulation (awareness, rhythm) and accustom the child to a foreign object being on/in their ears. Rarely is a hearing aid trial inappropriate before cochlear implantation is considered. If the child cannot feasibly use earmolds (e.g., aural atresia, small external canals, icthyosis), then a bone conduction aid should be considered.

T Typify

- This step involves efforts to determine the type of hearing impairment: conductive, sensory, neural, central, or mixed.
- In general, conductive hearing impairment is addressed either surgically or with hearing aids, and sensory loss with hearing aids or cochlear implantation.
- If the neural impairment is determined to be auditory neuropathy spectrum disorder (ANSD), cochlear implantation may be considered. An appropriate trial with amplification prior to implantation is prudent in cases of ANSD.
- Of course, with any type of hearing impairment, family preference and/or particular patient features (e.g., bilateral absence of cochlear nerves) may dictate the patient best communicate with sign language or some alternative mode of communication.

I Investigate

Attempt to determine the cause of the hearing loss through:

- A complete history that includes query about hearing impairments in the family, pregnancy, infections, and baby's history, including type of physiologic auditory screening, gestational age, hyperbilirubinemia, severe metabolic disturbance (e.g., with diaphragmatic hernia or ExtraCorporeal Membrane Oxygenation [ECMO]), and ototoxic drugs.
- A thorough physical exam, including height, weight, head circumference, birthmarks, facial asymmetry, and otomicroscopic exam of the tympanic membranes.
- Laboratory data, including results of maternal screenings for toxoplasmosis, rubella, and syphilis, as well as the baby's newborn metabolic screenings. In the United States, screening for congenital hypothyroidism is standard; however, complete follow-up of abnormal results is a concern (American Academy of Pediatrics, 2008). Centers for Disease Control and Prevention (CDC) does not recommend routine maternal screening for CMV infection during pregnancy. There is no drug licensed to treat congenital CMV infection.Nevertheless, at least in Utah, the law mandates CMV testing for every baby who does not pass universal newborn hearing screening. "Congenital CMV infection can be diagnosed if an infant has the virus detected in his or her urine, saliva, blood, or other tissues within 2-3 weeks after birth" (CDC, 2011). Other laboratory tests, such as urinalysis, cholesterol, and complete blood count, are unlikely to contribute to a cause for the hearing impairment—unless additional physical signs

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or symptoms queue the physician (Preciado et al., 2005).

- Obtaining an electrocardiogram (ECG) has an extremely low diagnostic yield but is important for ruling out the long QT of Jervell and Lange-Nielsen syndrome (JLNS). Reportedly, half of individuals had cardiac events before age 3 years, and half of untreated children die prior to age 15 years. Consider obtaining an ECG (or cardiology consultation or 24-hour monitoring of cardiac electrophysiology) if the hearing impairment is bilateral, severe-profound, and the patient or a family member has history of unexplained syncope (seizure or "falling-out" spell). The definitive diagnosis of JLNS requires all (1) congenital sensorineural deafness, (2) long QT interval—often manifest as syncope—most often elicited by emotion or exercise, and (3) presence of biallelic pathogenic variants in either KCNQ1 or KCNE1 (Tranebjærg et al., 2014).

Consult: Ophthalmologist, Radiologist, and Geneticist

Ophthalmologist

Consultation ensures that the patient's vision is optimized and assists in assessing the stigmata of syndromes (e.g., Usher) that may explain the hearing impairment. Ophthalmologists can facilitate early diagnosis for the timely management of potentially treatable congenital infections affecting hearing, such as syphilis, toxoplasmosis, and CMV. Sharma, Ruscetta, and Chi (2009) report that about 22% of children with sensorineural hearing loss have ophthalmic problems; the rate in deaf children is about 50%. Children with non-syndromic sensorineural hearing impairment have a 2- to 3-fold increased occurrence of ocular abnormalities (Johnston, Curry, & Newborough, 2010).

Radiologist

Consultation for imaging of the temporal bones is an important diagnostic endeavor. Abnormal findings are noted in about 30% of patients and somewhat more of patients with asymmetric hearing impairments (Licameli & Kenna, 2010). The explanations usually involve an abnormal architecture of the inner ear—most commonly large vestibular aqueduct. First described in 1978 (Valvasorri & Clemis) using radiographic polytomography, large vestibular aqueduct is probably best considered a proxy for labyrinthine dysplasia involving sensorineural hearing impairment—often progressive (Mori et al., 2008). There are several classifications of inner ear architectural differences. The classification of Sennaroglu and Saatci (2002) may be considered practical and embryologically based. Older terminologies (e.g., Mondini, Michele, Alexander) are of more historic origin than scientifically grounded. Since the older terminologies are no longer meaningfully communicative, they can best be placed in an "old word home."

Computed tomography and magnetic resonance imaging (MRI) are complementary tools, each having advantages and disadvantages (Licameli & Kenna, 2010). For a child with hearing impairment and any neurologic or cochlear nerve concern, MRI is our preference. If cochlear implantation is considered, the question is whether the ear is implantable or not? Is there a cochlea, a cochlear nerve, and otitis that is controlled or controllable? Our general preference is to request imaging at age 8 or 9 months, so as to obtain more data about otitis media. Imaging at an earlier age uncommonly affects what is done with the patient.

Geneticist

Consultation can be rewarding, especially in providing etiologic information, comorbidity risks, relieving guilt, and estimating risk of occurrence in another family member (Alford et al., 2014). Certainly the consultation is extremely helpful to the patient if the geneticist identifies an

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explanation for the hearing impairment that involves an important health concern, such as the sudden death risk associated with long QT syndrome or the impending blindness of Usher syndrome. Additional benefits of genetic consultation are:

- Family members becoming aware of their risks of having the hearing impairment.
- The affected individual knowing her/his risks for transmitting the auditory situation. An updated consultation when the patient anticipates her/his own family would be in order.
- The satisfaction of curiosity having been addressed.

While any physician may order a genetic laboratory test (e.g., for the common connexin gene associated with the majority of genetically determined hearing impairment), the geneticist is best prepared for interpretation and counseling about the results of the test. The most optimistic estimate in 2014 is that genetic testing can identify the explanation for congenital hearing impairment in about two-thirds of cases.

A nongeneticist is debatably best able to address "that genotype cannot necessarily predict phenotype due to the complexity of the genome, the proteome interacting with transcriptome, and the dynamically couples systems that are involved" (Yan & Liu, 2010).

Ethnicity is debatably an important determinant of optimally sequencing the diagnostic workup of the child with sensorineural hearing impairment (Chan, Schrijver, & Chang, 2011).

Genetic hearing impairments. Waardenburg is the most common type of autosomal dominant syndromic hearing impairment. Usher syndrome, which involves retinitis pigmentosa, is the most common type of autosomal recessive syndromic hearing impairment. Usher syndrome affects more than 50% of the deaf-blind persons in the United States (Smith et al., 2014). A child with congenital severe-profound hearing impairment, who seems otherwise healthy but exhibits late motor developmental milestones (e.g., does not walk by age 18 months), is considered high risk for having Usher Type I. A syndromic X-linked hearing impairment is Mohr-Tranebjaerg syndrome (deafness-dystonia-optic atrophy syndrome).

At least 70% of patients with genetically determined hearing impairments do not have a syndrome. These impairments—named for their gene loci—are designated DFN (for DeaFNess):

- DFNA: Autosomal dominant
- DFNB: Autosomal recessive
- DFNX: X-linked

Of patients with prelingual hearing impairment, about 75-80% are DFNA, about 20-25% are DFNB, and about 1-1.5% are DFNX. The number following the above designations reflects the order of gene mapping and/or discovery. For example, DFNB1 involves mutation in the gene GBJ2, which encodes the protein connexin 26, maps to chromosome 13q12 (Smith et al., 2014). Of antosomal recessive non-sydromic hearing loss, about 50% can be attributed to DFNB1. Many different connexin mutations have been reported.

An example of non-syndromic X-linked hearing impairment is DFNX3—characterized by a mixed conductive-sensorineural hearing loss. The conductive component is a result of stapes fixation (Smith et al., 2014). If the stapes is removed, there is gushing cerebrospinal fluid due to an abnormal communication between perilymph and the cerebrospinal fluid.

Mitochondrial hearing impairments involve mutations of mitochondrial DNA (mtDNA). The majority of mutations in mitochondrial genes cause a broad spectrum of maternally inherited multisystem syndromic disorders (Smith et al., 2014). A non-syndromic mtDNA mutation affects the ototoxicity of aminoglycosides. In some individuals with the 1555A>G mutation, hearing loss is induced by the administration of nominal doses of aminoglycosides (Pandya, 2011).

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I Initiate Discussion about Psychological Aspects

Initiate discussion of the psychological implications of raising a hearing-impaired child and how the family will cope with anger, stress, denial, grief, guilt, and over-compensation. Additionally, address the stress of communication strategies; hearing aids and cochlear implant accommodation; behavioral issues, including sleep; and mismatch of chronologic and communicative ages.

Photo courtesy of Oticon A/S

N No Harm

Meticulous care of remaining hearing is to be stressed. This includes advising about good ear hygiene and avoidance of ear-risk situations (e.g., ototoxicants, noise, and head trauma). Restriction of sporting activities is to be discussed if the patient has a labyrinthine dysplasia. Precautions related to aquatic activities may be indicated for the individual patient. Remember, ear wax (cerumen) is normal and protective. Do not use cotton-tip swabs or other objects inside the ear canal. To remove water from ear canal, evaporate with hair dryer (lowest heat and fan speed) held several inches from the ear. Caution about safety for localization of a sound's source is especially important if the patient has asymmetric hearing impairment. Keeping up to date with immunizations is important: childhood hearing impairment (notwithstanding inner ear dysplasia) is associated with an approximately five-fold increased relative risk of meningitis (Parner et al., 2007).

Our terminologies can be confusing, inadequate, and noncommunicative. For example, hearing "impairment" and hearing "loss" are often used interchangeably, as if they are synonymous terms. Strictly speaking, that is not the case—how can something that was never there be lost? Hearing impairment means deviation from the outer limits of an established standard. Hearing loss means deviation from one's own baseline threshold, which has been established and documented and which may or may not be impairment. It suggests progression. Epidemiologically, "hearing loss" would be a longitudinal term, whereas "hearing impairment" would be cross-sectional (Hannley, 2012).

It is important that the parents receive a realistic and consistent message regarding available communication options.

A Assure Communication

Provide the comfort that the patient will be able to communicate in some manner. If not the customary listening and speaking mode facilitated by hearing aids or cochlear implantation preferred by hearing parents, then "total communication" or sign (manual) communication. Any discussion of communication options should be done in conjunction with the child's team of medical personnel, audiologists, and early intervention providers. It is important that the parents receive a realistic and consistent message regarding available communication options. Deaf-blind patients are a special group, necessitating extraordinary anticipation and care.

T Test Repeatedly

Continue to evaluate as the child grows. This will enable the audiologic exam to become more accurate due to the ability of an older child to be more capable of responding to more sophisticated testing techniques. Regular testing will allow changes to be identified and provide a more accurate understanding of the etiology of the hearing loss. It is vital to ensure that amplification is appropriate for the type and degree of hearing loss. Testing should proceed at regular intervals as recommended by the audiologist and early intervention providers. At least once, test each of the first-degree relatives to determine if any has a hearing problem. It will also help them to understand the difficulties/rigors of having hearing tests.

E Educate

Families must learn about hearing impairment and what can be done. The earliest possible education for the child and family are vital to successful outcomes. Best communicative outcomes involve a multidisciplinary team of at least the primary care physician, audiologist, therapist(s), educator(s), otolaryngologist, ophthalmologist, and geneticist. Ideally, all care should be coordinated within the medical home. It is often very helpful to have a parent-maintained scrapbook of all the various tests and assessments of the child. This not only enhances parent involvement and empowerment, but also enables the professionals to have immediate access to all the relevant information necessary for optimum care of the child.

Families must learn about hearing impairment and what can be done. The earliest possible education for the child and family are vital to successful outcomes.

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