
2

Audiometric Tests and Diagnostic Workup

Paul J. Govaerts

Children with suspected congenital hearing loss are referred for audiological and diagnostic workup. The suspicion is often based on parental anxiety, but more and more children are referred after failing neonatal hearing screening.

The audiological workup aims at establishing the type and degree of hearing loss. Pure tone audiometry is the standard test in daily clinical practice. The technique of pure tone audiometry is explained and some issues that are useful for a good interpretation of audiometric reports are highlighted. Standards are given for age-related deterioration of hearing. In addition, attention is paid to objective tests, such as auditory brainstem responses (ABR), otoacoustic emissions, and tympanometry. These tests play an important role in assessing the hearing of infants or otherwise uncooperative subjects.

The diagnostic workup of hearing loss also aims at refining the diagnosis, excluding associated pathology in case of syndromic hearing loss, and identifying the molecular defect. To exclude associated pathology means looking for syndromes. Typical features such as facial anomalies may be indicative of a syndrome. A thorough clinical examination focusing on any signs of an underlying syndrome is therefore mandatory. In addition, several organs are known to be at risk in case of a congenital hearing problem. These are mainly the kidneys (e.g., BOR syndrome), the heart (e.g., long Q-T syndrome), the thyroid (e.g., Pendred syndrome), and the eyes (e.g., Usher syndrome). Special attention should therefore be given to these organs.

I. AUDIOLOGICAL WORKUP

A. Pure Tone Audiometry

Pure tone audiometry is the standard test to assess hearing and hearing loss. It is a way to measure hearing thresholds at different frequencies and at both ears. It is a subjective test as it involves the cooperation of the test person and this is in contrast to objective tests like brainstem evoked audiometry (ABR) or otoacoustic emissions.

1. Technique

The technique of measuring hearing thresholds is standardized (American National Standards Institute, ANSI). The thresholds are expressed in “decibels hearing level” (dBHL), whereby zero dBHL at a given frequency is defined as the lowest sound level that a normal person of 18 years is just able to hear. A hearing loss of, for example, 30 dB means that the intensity of sound has to be 30 dB above the zero level before the subject starts hearing it (the average intensity of conversational speech is 50–60 dB).

In general, hearing is tested in a soundproof room with a calibrated audiometer and earphones that present sounds of varying intensity and frequency to each ear separately. Different test procedures exist. Conventional procedures use attenuation steps of 5 dB or more, thus introducing a minimum error of ± 5 dB. The test frequencies used are 125, 250, 500, 1000, 2000, 4000, and 8000 Hz. Once the threshold at a given frequency is assessed, it is plotted with a specific symbol on the audiogram. Both the graphic representation and the symbols are defined in international recommendations (1) (Table 1). For unmasked air conduction thresholds the symbols are O for the right ear and X for the left ear (Fig. 1).

Air conduction (AC) audiometry uses earphones to present the different sounds. Bone conduction (BC) audiometry uses a bone-conducting vibrator

Table 1 Common Symbols Used for Audiometric Representation (ASHA 1990)

	Right ear	Left ear
Air conduction (earphones)		
Unmasked	O	X
Masked	Δ	\square
Bone conduction (mastoid)		
Unmasked	<	>
Masked	[]

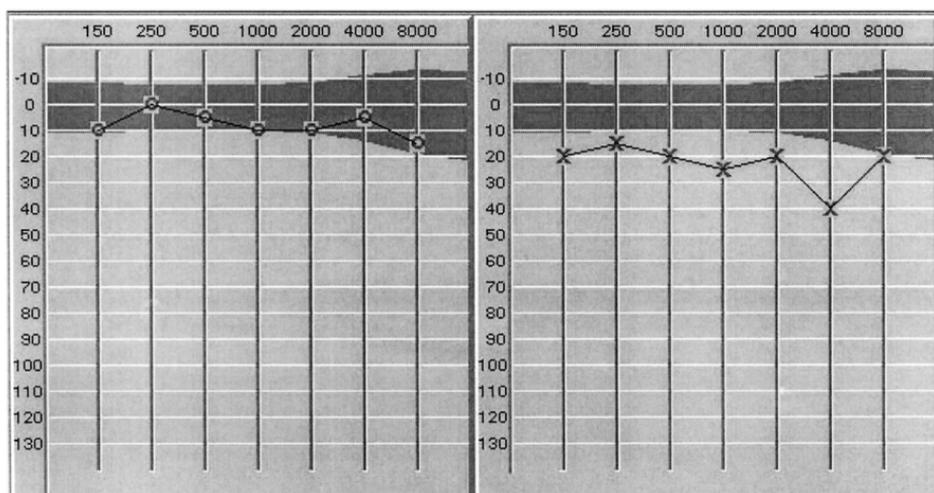


Figure 1 Typical audiogram showing the air-conduction thresholds of the right ear (depicted with the symbol 0 at the left panel) and the left ear (depicted with the symbol X at the right panel). The dark-gray area is the 95% confidence region for an 18-year-old person.

that is positioned on the mastoid of each ear. Air-conducted sound reaches the auditory nerve through the external, the middle, and the inner ear. Bone-conducted sound bypasses the external and the middle ear and reaches the auditory nerve directly through the inner ear. The symbols for unmasked BC thresholds are < for the right and > for the left ear.

2. Conductive, Sensorineural, and Mixed Hearing Loss

Anomalies of the inner ear and/or the central auditory pathways (from the auditory nerve via the brainstem to auditory cortex) result in a “perceptive” or “sensorineural” hearing loss and will affect both AC and BC thresholds (Fig. 2, left ear). Many forms of congenital hearing loss and most types of nonsyndromic hearing loss are sensorineural. Anomalies of the outer and middle ear will affect only AC thresholds. The difference between AC and BC threshold is called the “air-bone gap” and reflects a middle- or outer-ear problem. This is called a “conductive” hearing loss (Fig. 2, right ear). This type of hearing loss is found, for example, in congenital atresias of the outer ear canal, malformations of the middle ear ossicles, and stapedial ankylosis due to otosclerosis.

Problems at different levels (e.g., middle and inner ear) may result in a “mixed” hearing loss, with a sensorineural component given by a BC loss

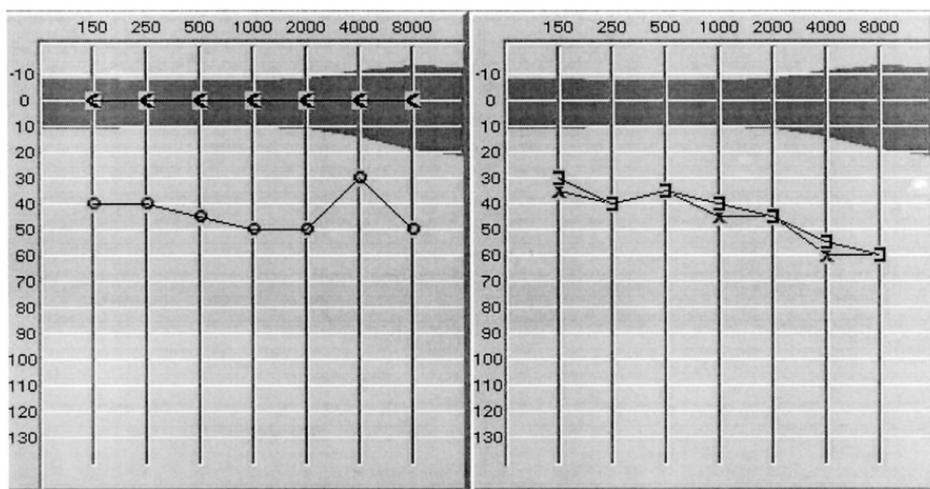


Figure 2 Typical audiograms of a conductive hearing loss at the right ear (left panel) and a sensorineural hearing loss at the left ear (right panel). The right ear shows normal bone-conduction thresholds (PTA 0 dBHL) and abnormal air-conduction thresholds (PTA 47 dBHL). The left ear shows abnormal bone- and air-conduction thresholds with a PTA of 40 dBHL and 43 dB, respectively.

and a conductive component given by a superimposed air-bone gap. This type can be found for instance in otosclerosis affecting both the cochlea and the stapes.

3. Masking

Sound that is presented to one ear may also reach the contralateral ear, which should be avoided. The interaural attenuation of AC and BC sound is known for the different test frequencies and in case a risk of crossover exists, the nontest ear should be “masked.” Masking is difficult and both the decision whether to mask and the execution of it should not be underestimated and, although strict criteria and guidelines exist, it should be left to audiologists. Masking leads to corrected AC and BC thresholds and special symbols exist to represent these.

4. PTA or Fletcher Index

To summarize the audiometric findings several indices have been introduced, among which the pure tone average (PTA) or “Fletcher index” is the most commonly used. The PTA is the average of the AC thresholds at the frequencies 500, 1000, and 2000 Hz. For instance, the audiogram in Figure 2

has a PTA of 47 dBHL for the right ear and 40 dBHL for the left ear. Hearing losses are often described in a qualitative way that is based on the PTA. Different classifications exist. The WHO recommends the classification of Table 2. In addition one may describe the curve of the audiogram, such as “down-sloping or U-shaped.

5. Feasibility

Since pure tone audiometry requires a responsive subject, it may be unfeasible in children or in persons who are not able to give reliable responses because of mental or other problems. For small children special observational or conditioning techniques exist that allow fairly reliable audiometric results from the age of approximately 2 years onward. At younger ages, more than one session may be required or alternative techniques, like ABR (see below), may be used.

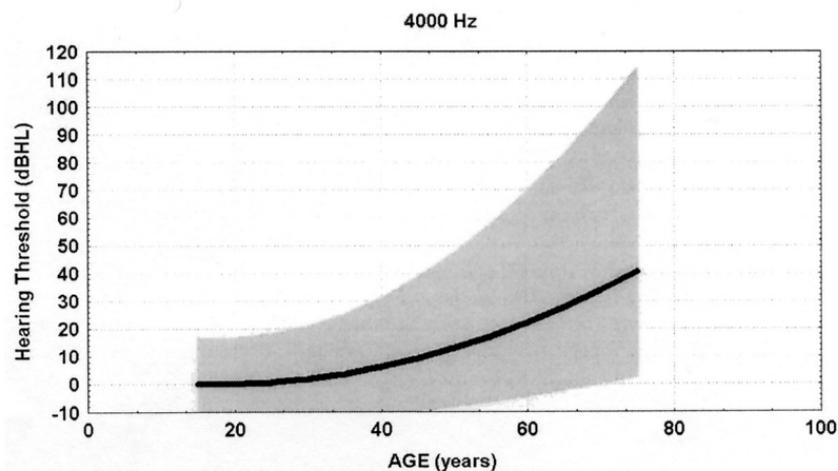
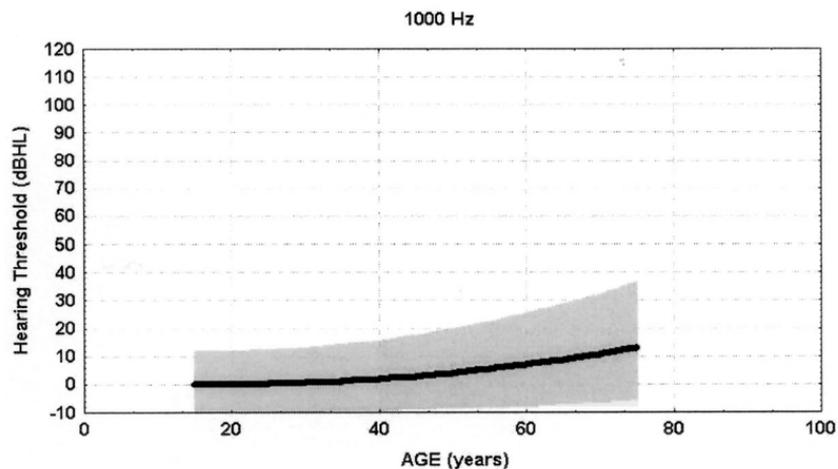
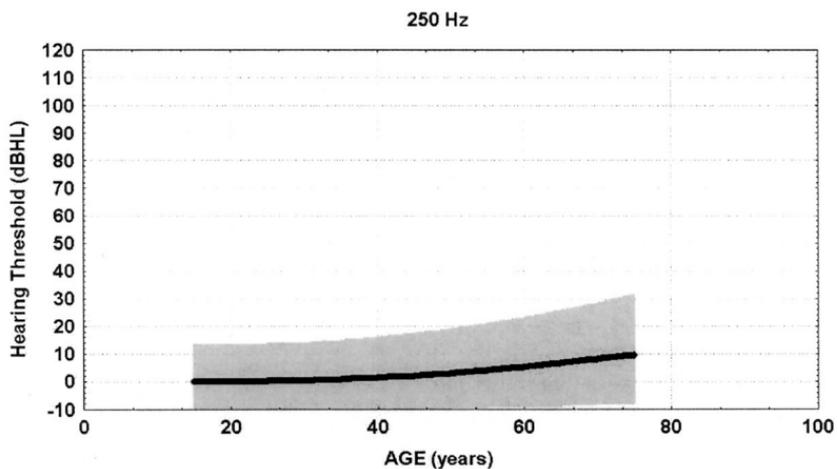
6. ISO 7029

A special issue of interest is the definition of “normality” in hearing. Zero dBHL is defined as the average threshold in 18-year-old persons with a history free of otological disease. It is, however, known that hearing deteriorates with age and a hearing loss of 25 dBHL at 500 Hz and even 80 dBHL

Table 2 Grades of Hearing Impairment

Grade of impairment	Corresponding audiometric ISO value (average of 500, 1000, 2000, 4000 Hz) of the better ear	Performance
0 No	25 DB or better	No or very slight hearing problems Able to hear whispers.
1 Slight	26–40 DB	Able to hear and repeat words spoken in normal voice at 1 meter
2 Moderate	41–60 dB	Able to hear and repeat words using raised voice at 1 meter
3 Severe	61–80 dB	Able to hear some words when shouted into better ear
4 Profound	81 dB or greater	Unable to hear and understand even shouted voice

Source: World Health Organization (<http://www.who.int/pbd/pdh/Docs/GRADESTable-DEFs.pdf>). Adapted from the report of the informal working group on prevention of deafness and hearing impairment programme planning, WHO, Geneva, with adaptations from the report of the first international consultation on future program developments for the prevention of deafness and hearing impairment, WHO, Geneva.



at 8000 Hz is not abnormal for a 70-year-old man. The age- and gender-related distribution of hearing thresholds is defined by the International Organization for Standardization (ISO) 7029 standard [ISO 7029 (1984) “Acoustics—threshold of hearing by air conduction as a function of age and sex for otologically normal persons” (International Organization for Standardization, Geneva)]. The hearing threshold of any given patient can therefore be expressed as the number of standard deviations below or above the median value for the given age and gender. The corresponding percentile can be derived from these data in any table of a normal distribution. For example, the median hearing loss at 500 Hz for a normal 70-year-old man is 8 dBHL according to the ISO 7029 standard with a positive standard deviation of 10 dBHL. A hearing loss of 25 dBHL can be expressed as 1.7 standard deviations ($25 \text{ dBHL}/10 \text{ dBHL}$) above the median and this corresponds to the ninety-sixth percentile (or P96). If normality is defined as the group between P2.5 and P97.5, this is to be considered normal (2) (Fig. 3) Since genetic hearing losses affect different generations in one family, it is important to bear in mind this concept of normality and abnormality.

7. Supraliminal Audiometry

So far only “pure tone” audiometry has been discussed. This is called “liminal” audiometry since it assesses the threshold of hearing. Hearing, however, is far more complex than simply detecting low-level sounds. To understand the sounds one needs to discriminate different sounds. These supraliminal aspects of hearing can be assessed by other tests, such as speech audiometry. Supraliminal tests in general are less standardized than liminal audiometry and they are most often language-dependent. In addition, they assess not only hearing but also higher cognitive and other functions. This renders them less applicable for genetic research.

8. Cochlear Conductive Loss

A rare finding is the so-called “cochlear conductive hearing loss.” This is typically found in an enlarged vestibular aqueduct (2) but it may also be found in other types of cochlear anomaly. It shows a conductive hearing loss on audiometry (an air-bone gap). Tympanometry (see below) shows normal

Figure 3 Evolution of normal hearing thresholds for 250 Hz (top), 1000 Hz (middle) and 4000 Hz (bottom) in function of age according to the ISO 7029 formula (see text for details). The solid lines are the median hearing thresholds and the gray areas represent the 9% confidence interval.

middle-ear pressure but stapedial reflexes are absent. These audiometric findings are commonly interpreted as an ossicular problem (ossicular malformation or fixation) but exploratory tympanotomy or high-resolution CT scan does not reveal any such condition. Although the underlying mechanism is not quite understood, it is thought that the minor cochlear malformation prevents the traveling wave within the cochlea from proceeding smoothly. Although the sound is well transferred through the middle ear and thus reaches the cochlea without problems, it encounters a mechanical resistance within the fluids of the cochlea itself before arriving at the inner hair cells. This should explain the relatively normal BC thresholds and abnormal AC thresholds.

B. Auditory Brainstem Responses

The evaluation of ABR, also called evoked response audiometry (ERA) or “brainstem evoked response audiometry (BERA), refers to an objective technique of assessing hearing thresholds. It is objective since it does not require the cooperation of the test person. It can even be done under anesthesia. It is based on the recording of electrical responses from the cochlea and the auditory nerve after acoustical stimulation either via AC or via BC. An averaging paradigm is used with a short stimulus (typically a click of 100 μ sec) that is presented some 1000–2000 times to allow averaging of the response. Although the test is objective, physical principles allow it merely to assess the thresholds at the higher frequencies (2000 and 4000 Hz). The thresholds at other frequencies are far less reliable. In addition, the confidence interval of the thresholds is higher than the ± 5 dB of the pure tone audiometry. This renders ABR a test of second choice that should only be used when regular audiometry is not feasible or reliable, e.g., in infants or mentally disabled persons.

Special devices have been developed to record ABR and to automatically interpret the responses in terms of hearing thresholds. The output is either “pass” or “fail” meaning normal hearing or, respectively, a hearing loss of at least 20–30 dBHL. These are called automated ABRs (AABRs) and are used for screening purposes and especially for neonatal hearing screening.

C. Otoacoustic Emissions

Otoacoustic emissions are acoustic signals that are emitted from the ear (3). Different types exist, but in clinical practice, the so-called transient evoked otoacoustic emissions (TEOAEs) are most commonly used. They are produced by the outer hair cells if they are in good physiological condition and stimulated by an external sound (4,5). The technique and interpretation

are well established. If no TEOAEs can be elicited, this is interpreted as a hearing loss of 30 dBHL or more. Presence of TEOAEs means normal or near-normal hearing. The test is fast and no cooperation of the test person is needed. On the other hand, it is rather aspecific, since both sensori-neural and conductive hearing loss will result in the absence of TEOAEs and hardly any frequency information can be derived. However, it is a high-quality screening tool and as such it is widely used for neonatal and other hearing screening. Thanks to the many nationwide (“universal”) neonatal hearing screening programs, early detection of congenital hearing loss is becoming more and more common practice (6,7).

D. Tympanometry

Tympanometry is a test to register the acoustic impedance of essentially the middle ear. It reflects the pressure within the middle ear cavity and as such it is often used to detect middle ear ventilation problems. A special feature is the possibility to record stapedial reflexes, which are small movements of the stapes, due to a contraction of the stapedial muscle as a response to high-intensity sounds. The movement of the stapes makes the incus, the malleus, and the tympanic membrane move, which can be measured with an external probe. The test is simple and gives information on the mobility of the ossicular chain. An immobile chain fails to give recordable stapedial reflexes. This typically occurs in otosclerosis and also in different types of congenital ossicular malformation with fixation of one of the ossicles.

II. GENERAL WORKUP

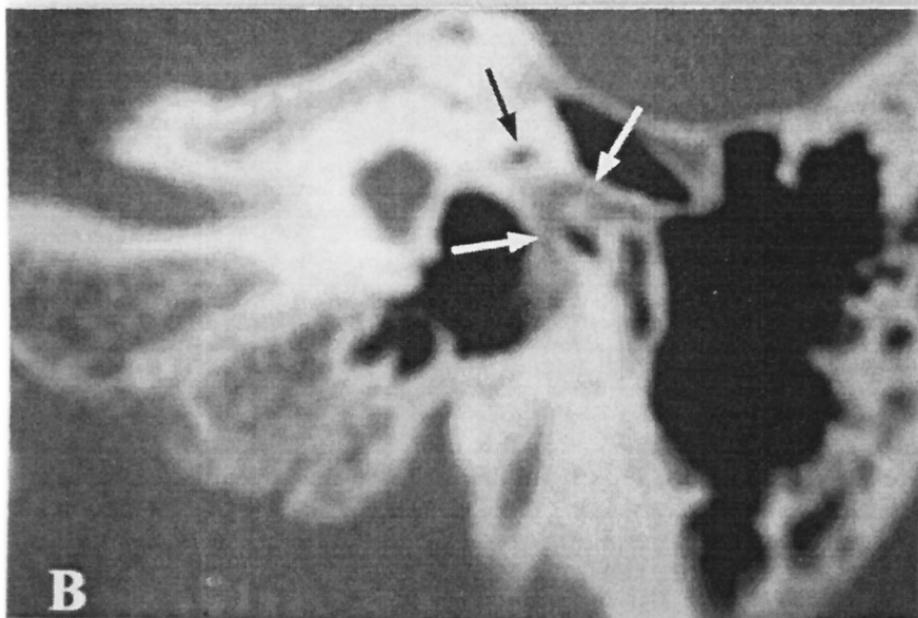
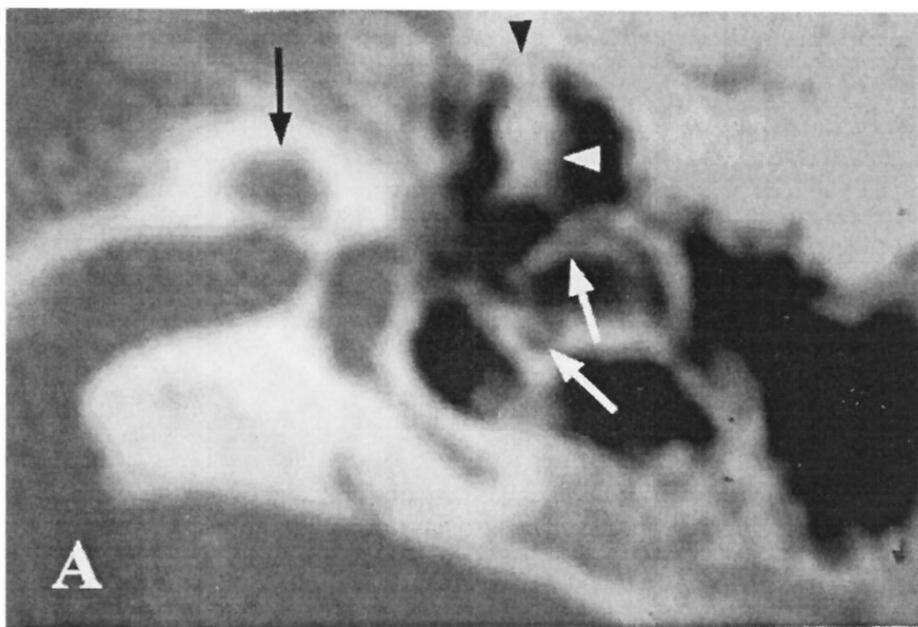
A. Blood Examination, Including Connexin 26

Thanks to the universal neonatal hearing screening programs, an increasing number of neonates are referred for diagnostic workup. The amount of blood that is available for laboratory tests is limited. Therefore, not all tests that may contribute to the diagnosis may be done. In the absence of indications for specific underlying pathologies, genetic (connexin 26) and serological examinations may suffice.

Thyroid hormone tests are not useful. Patients with Pendred syndrome are euthyroid at birth. Only half of them become hypothyroid with the development of a goiter after the age of 10 years (8).

B. Genetic Examination

In case of syndromic hearing loss, either karyotyping or specific biochemical and molecular investigations may be useful. Even in the absence of



phenotypic indications suggestive of an underlying syndrome, it is recommended to actively exclude the three most frequent syndromes, namely, the autosomal recessive Usher and Pendred syndromes and the X-linked Alport syndrome. The former two syndromes typically have negative familial histories. Ophthalmological examinations with electroretinography (at the age of 5 years), medical imaging, and urine examination are required.

An increasing number of genes causing nonsyndromic hearing loss have been described [see web-page V Camp (9)]. At present it is not possible to routinely search for many of these mutations for practical reasons. Mutations in the connexin 26 gene (GJB2) account for probably 20% of the nonsyndromic congenital hearing losses (10–12). In some populations this figure may even be higher [e.g., 80% in Jewish Ashkenazi children (13,14)]. The most common mutations are the 35delG and the 162delT mutations and these can be routinely found by simple and inexpensive restriction enzyme analysis. Additional mutations should be ruled out by sequencing the gene.

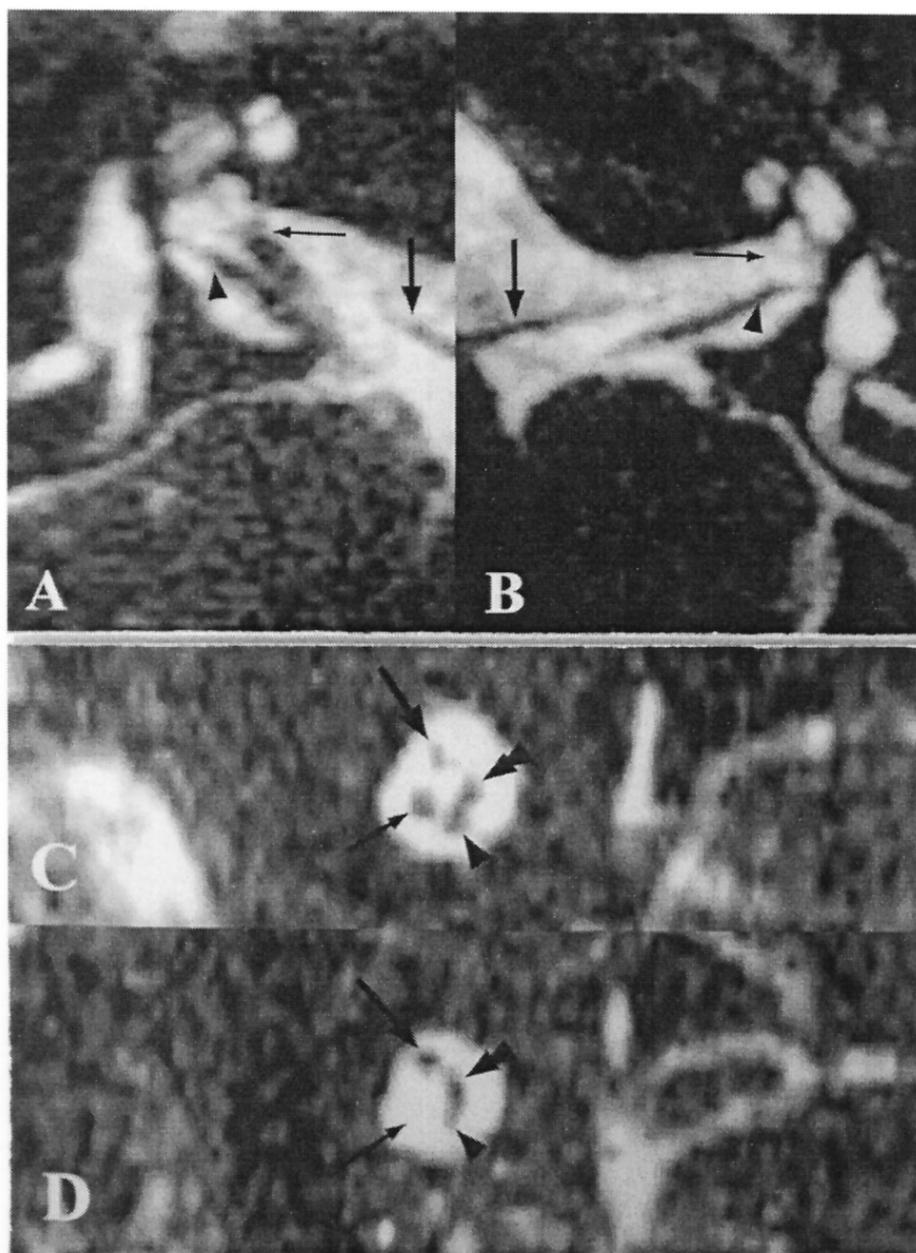
A frequent anomaly found in congenital nonsyndromic hearing loss is an enlarged vestibular aqueduct (2). This may account for over 20% of all congenital hearing losses (15). A number of them [15% (16)] have been shown to be related to mutations in the Pendred syndrome gene (PDS). If an enlarged vestibular aqueduct is found by medical imaging, it may be worthwhile investigating this gene.

C. Serological Examination

Maternal or congenital infections are becoming rare as a cause of congenital deafness in the Western world.

Thanks to widespread vaccination, the rate of confirmed congenital rubella syndrome has decreased to approx 0.05 per 10,000 life births (17). In

Figure 4 CT scan for congenital conductive hearing loss (courtesy J. Casselman). Axial and coronal CT image with bone window through the left middle and inner ear of a patient with congenital conductive hearing loss related to the branchio-oto-renal syndrome. (A) The malleus and incus are fused (white arrowhead) and are fixed against the anterior wall of the middle ear cavity (black arrowhead). The facial nerve descends in an abnormal position through the middle ear cavity and splits in two descending branches (white arrows). Note the hypoplastic cochlea with absence of a normal modiolus. (B) The facial nerve leaves its normal position under the lateral semicircular canal (black arrow) and gives rise to two branches, which are descending, through the middle-ear cavity (white arrows).



areas where rubella vaccination is not common practice, serological examination is probably justified.

Cytomegalovirus (CMV) is a frequent cause of congenital infections [0.4–2.3% of live births (18)] and may cause congenital hearing loss. Prenatal diagnosis is possible and provides the optimal means for both diagnosing fetal infection and identifying fetuses at risk of severe sequelae (19). Serological evaluation is possible on blood samples and PCR diagnosis is possible on urine samples.

D. Imaging

Refining the diagnosis means searching for the etiology.

Middle ear problems often consist of atresia and/or ossicular malformations. In most cases these can be readily seen on high-resolution CT scan (Fig. 4). Therefore, a congenital conductive hearing loss makes a CT scan mandatory. This can be done under sedation or anesthesia if necessary.

Sensorineural hearing losses are mainly caused by cochlear problems (large vestibular aqueduct, Mondini dysplasia, semicircular canal malformations) and to a lesser extent by central auditory lesions, situated in the auditory nerve (aplasia or hypoplasia), the auditory pathways, or the auditory cortex. Magnetic resonance (MR) imaging (Fig. 5) may elucidate the details and is often complementary to CT, especially in the study of inner-ear malformations (20). Routine T2-weighted spin-echo images of the brain are

Figure 5 MRI for congenital sensorineural hearing loss (courtesy J. Casselman). Axial 0.7-mm-thick T2-weighted gradient-echo images and sagittal reconstructions perpendicular to the course of the nerves made at the level of the fundus of the internal auditory canal (IAC) through both the right (A, C) and the left (B, D) inner ear in a patient with congenital deafness on the left side. (A) Normal facial nerve (large black arrow), inferior vestibular (black arrowhead), and cochlear (small black arrowhead) branch of the VIIIth nerve can be seen near the floor of the IAC. (B) The facial nerve (large black arrow) and inferior vestibular branch of the VIIIth nerve (black arrowhead) can again be distinguished; however, the cochlear branch of the VIIIth nerve is absent (small black arrow). (C) The facial nerve (large black arrow), superior vestibular (double arrowhead), inferior vestibular (arrowhead), and cochlear (small black arrow) branch of the VIIIth nerve can all be seen on the reconstruction made at the fundus of the right IAC. (D) The facial nerve (large black arrow), superior vestibular (double arrowhead), and inferior vestibular branch (arrowhead) of the VIIIth nerve can again be seen. This image confirms the congenital absence of the cochlear branch of the VIIIth nerve (small black arrow).

ideally suited to exclude white-matter disease or other lesions in the cochlear nuclei and along the auditory pathways. Only MR can demonstrate the abnormal course, hypoplasia, or aplasia of the vestibulocochlear nerve and facial nerve (21). MR techniques have to be adapted for inner-ear malformations. Only heavily T2-weighted gradient-echo sequences, such as three-dimensional Fourier transform constructive interference in steady state (3DFT-CISS), true-fast imaging with steady precession (true-FISP), or 3D-fast spin-echo (3D-FSE) sequences, are suitable for this purpose (22). Moreover, some changes inside the membranous labyrinth can only be seen on submillimetric (0.7 mm thickness) heavily T2-weighted MR images. A CT can be complementary to evaluate the abnormal internal auditory canal or facial nerve canal. Thin CT images (reconstructed 1- or 1.25-mm-thick images every 0.5 or even 0.1 mm) yield detailed information on the bony labyrinth, including modiolus and even the osseous spiral lamina, but they do not give any information on the fluid in the labyrinthine compartments, which can only be seen on MR images.

E. Ophthalmological Examination with Fundoscopy

One of the most frequent ocular problems linked with congenital hearing loss is retinitis pigmentosa (Usher syndrome). This is seen on fundoscopy. The first signs of retinitis, however, appear no sooner than at the age of a couple of years and often even much later. Therefore, a negative ophthalmological examination is no proof of the absence of Usher syndrome. Electroretinography may provide early diagnosis from the age of 2 years onward and some authors claim that all children with severe to profound, prelingual sensorineural hearing loss should be screened by ophthalmological examination including electroretinogram (23).

Other syndromes may consist of early onset eye problems, such as retinopathy, optic atrophy, or iridal or corneal anomalies.

F. Electrocardiogram

An electrocardiogram is routinely taken to exclude the long Q-T syndrome (Jervell and Lange-Nielsen syndrome).

G. Ultrasound Examination of the Renal System

Ultrasound examination of the kidneys is important to rule out any renal structural malformation, e.g., in the branchio-oto-renal syndrome (BOR).

H. Urine

Urine examination should exclude microscopic hematuria and proteinuria, although the latter is uncommon in children. It is also useful for detecting cytomegalovirus by PCR techniques.

REFERENCES

1. ASHA American Speech-Language-Hearing Association. Guidelines for audiometric symbols. ASHA 1990; 20(suppl 2):25–30.
2. Govaerts PJ, Casselman J, Daemers K, De Ceulaer G, Peeters S, Somers TH, Offeciers FE. Audiological findings in the large vestibular aqueduct syndrome. *Int J Pediatr ORL* 1999; 51:157–164.
3. Kemp DT. Stimulated acoustic emissions from within the human auditory system. *J Acoust Soc Am* 1978; 64:1386–1391.
4. Khanna SM, Leonard DG. Measurement of basilar membrane vibrations and evaluation of cochlear condition. *Hearing Res* 1986; 23:37–53.
5. Sellick PM, Patuzzi R, Johnstone BM. Measurements of basilar membrane motion in the guinea pig using the Mössbauer technique. *J Acoust Soc Am* 1982; 72:131–141.
6. White KR, Vohr BR, Maxon AB, Behrens TR, McPherson MG, Mauk GW. Screening all newborns for hearing loss using transient evoked otoacoustic emissions. *Int J Pediatr Otorhinolaryngol* 1994; 29(3):203–217.
7. Govaerts PJ, De Ceulaer G, Yperman M, Van Driessche K, Somers TH, Offeciers FE. A two-stage, bipodal screening model for universal neonatal hearing screening. *Otol Neurotol* 2001; 22(6):850–854.
8. Reardon W, Coffey R, Chowdhury T, Grossman A, Jan H, Britton K, Kendall-Taylor P, Trembath R. Prevalence, age of onset, and natural history of thyroid disease in Pendred syndrome. *J Med Genet* 1999; 36(8):595–598.
9. Van Camp G, Smith RJH. Hereditary Hearing loss Homepage. World Wide Web URL: <http://www.uia.ac.be/dnalab/hhh/>. (October, 1996).
10. Dahl HH, Saunders K, Kelly TM, Osborn AH, Wilcox S, Cone-Wesson B, Wunderlich JL, Du Sart D, Kamarinos M, Gardner RJ, Dennehy S, Williamson R, Vallance N, Mutton P. Prevalence and nature of connexin 26 mutations in children with non-syndromic deafness. *Med J Aust* 2001; 175(4): 182–183.
11. Kenna MA, Wu BL, Cotanche DA, Korf BR, Rehm HL. Connexin 26 studies in patients with sensorineural hearing loss. *Arch Otolaryngol Head Neck Surg* 2001; 127(9):1037–1042.
12. Milunsky JM, Maher TA, Yosunkaya E, Vohr BR. Connexin-26 gene analysis in hearing-impaired newborns. *Genet Test* 2000; 4(4):345–349.
13. Morell RJ, Kim HJ, Hood LJ, et al. Mutations in the connexin 26 gene (GJB2) among Ashkenazi Jews with nonsyndromic recessive deafness. *N Engl J Med*. 1998; 339:1500–1505.

14. Lerer I, Sagi M, Malamud E, Levi H, Raas-Rothschild A, Abeliovich D. Contribution of connexin 26 mutations to nonsyndromic deafness in Ashkenazi patients and the variable phenotypic effect of the mutation 167delT. *Am J Med Genet* 2000; 6:53–56.
15. Zhang S, Zhao C, Yu L. Analysis of sensorineural hearing loss in 77 children. *Lin Chuang Er Bi Yan Hou Ke Za Zhi* 1997; 11(6):252–254.
16. Scott DA, Wang R, Kreman TM, Andrews M, McDonald JM, Bishop JR, Smith RJ, Karniski LP, Sheffield VC. Functional differences of the PDS gene product are associated with phenotypic variation in patients with Pendred syndrome and nonsyndromic hearing loss (DFNB4). *Hum Mol Genet* 2000; 1709–1715.
17. Zimmerman L, Reef SE. Incidence of congenital rubella syndrome at a hospital serving a predominantly Hispanic population. *Pediatrics* 2001; 107(3):E40.
18. Witters I, Van Ranst M, Fryns JP. Cytomegalovirus reactivation in pregnancy and subsequent isolated bilateral loss in the infant. *Genet Couns* 2000; 11(4): 375–378.
19. Azam AZ, Vial Y, Fawer CL, Zufferey J, Hohfeld P. Prenatal diagnosis of congenital cytomegalovirus infection. *Obstet Gynecol* 2001; 97(3):443–448.
20. Casselman JW, Offeciers FE, De Foer B, Govaerts P, Kuhweide R, Somers TH. CT and MR imaging of congenital abnormalities of the inner ear and internal auditory canal. *Eur J Radiol* 2001; 40(2):94–104.
21. Casselman JW, Offeciers FE, Govaerts PJ, Kuhweide R, Geldof H, Somers TH, D’Hont G. Aplasia and hypoplasia of the vestibulocochlear nerve: Diagnosis with MR Imaging. *Radiology* 1997; 202:773–781.
22. Casselman JW, Kuhweide R, Deemling M, Ampe W, Dehaene I, Meeus L. Constructive interference in steady state-3DFT MR imaging of the inner ear and cerebellopontine angle. *Am J Neuroradiol* 1993; 14:47–57.
23. Mets MB, Young NM, Pass A, Lasky JB. Early diagnosis of Usher syndrome in children. *Trans Am Ophthalmol Soc* 2000; 98:237–242.