

Item 49 : Clinical and functional assessment of a sensory disability: hearing loss

Collège Français d'ORL et de Chirurgie Cervico-faciale

2013

Table des matières

Introduction.....	3
1. Hearing loss.....	3
1.1. Categories of hearing loss severity.....	3
1.2. Functional assessment of hearing loss.....	4
1.2.1. In children.....	4
1.2.2. In adults.....	4
2. Sensory disabilities in children.....	4
2.1. Normal development of spoken language in children.....	4
2.2. Warning signs – approaches to management.....	5
3. Sensory disabilities in adults.....	5
3.1. General.....	5
3.2. Repercussions.....	5
3.3. Approaches to management.....	6
4. The specific case of profound or total unilateral hearing loss.....	6

Objectifs ENC

- Assessing an impairment or disability
- Analyzing a disability's implications in terms of the patient's career and social life
- Knowing the direct and indirect signs of hearing loss in children
- Knowing how hearing disorders are screened for in children of different ages

Introduction

Auditory disabilities are the result of hearing loss. The seriousness of a patient's disability depends on when hearing loss sets in, how severe and permanent it is, and whether it affects one or both ears. Hearing loss may originate in the outer, middle, or inner ear or in the central auditory pathway. Depending on where the dysfunction occurs, hearing loss may be classified as either conductive or sensorineural, or mixed (a combination of the two).

1. Hearing loss

Conductive hearing loss results from dysfunction in the :

- outer ear (pinna and external auditory canal);
- middle ear (tympanic membrane, ossicles, mastoid, Eustachian tube).

Such problems may be due to malformations, and thus congenital in origin. More often they are acquired following an inflammation or infection (chronic otitis). Hearing loss can also be caused by degenerative conditions leading to otosclerotic stapes fixation, such as in otospongiosis in adults. Auditory impairments of this kind occur when the middle ear fails to perform its impedance-matching function properly. Loss of hearing is thus at most 60 dB (50-60 dB in major malformation syndromes).

Sensorineural hearing loss is most often caused by damage to the inner ear, the acoustic nerve, or very rarely the central auditory pathways.

Inner ear involvement is predominant in congenital bilateral hearing loss, often of genetic or idiopathic origin. The severity of such hearing loss varies on a case-by-case basis from 20 to over 120 dB. *Bilateral profound congenital sensorineural hearing loss is a major sensory disability and can lead to severe communication disorders.*

1.1. Categories of hearing loss severity

Determined as follows (based on the better ear) :

- mild hearing loss: 20-39 dB lost;
- moderate hearing loss: 40-69 dB lost;
- severe hearing loss: 70-89 dB lost;
- profound hearing loss: more than 90 dB lost.

The 40-dB level is the first major disability level, because speech can only be heard if the speaker raises his or her voice. At 90 dB, speech can no longer be heard.

1.2. Functional assessment of hearing loss

Functional assessment is based, in children and adults, on behavioral audiometry.

1.2.1. In children

Behavioral audiometry techniques can generally be used starting at the age of 3 months (once psychomotor development is far enough along for a baby to hold its head up). Tests are performed using headphones and skull vibrators to determine response thresholds elicited by air and bone conduction across all frequencies. Conditioning techniques and audio equipment are selected in accordance with the child's age. This data is crucial for establishing indications and recommending hearing prosthetic aids, which need to be performed as effective as possible beginning as early as 6 months of age, currently the optimal time for diagnosing congenital hearing loss.

Other techniques, such as oto-acoustic emissions or automated testing of short-latency auditory evoked potentials, can be used in maternity wards to screen for neonatal hearing loss. Brainstem auditory evoked potentials are also useful for obtaining initial data on high-frequency hearing thresholds. Impedance measurement is used to facilitate diagnosis of middle-ear disorders.

Screening for hearing loss is recommended not only in newborns but also systematically at the ages of 9 months, 2 years, and 4 years. Such screening involves risk factor analysis, parent interviews, vocal stimulation tests, and hearing tests with sound-emitting toys.

If any hearing loss is suspected, age-appropriate behavioral audiometry (as described above) should be carried out by a specialized team.

1.2.2. In adults

Audiometric assessments should always include pure-tone and speech audiometry, as well as impedance measurements. Auditory brainstem evoked potential testing should be reserved for the topographic diagnosis of sensorineural hearing loss.

2. Sensory disabilities in children

2.1. Normal development of spoken language in children

Involves the following stages :

- at birth, child reacts to sounds;
- begins burbling and babbling around 3 months;
- reacts to his/her name around 4 months;
- imitates sounds and intonations around 6 months;
- gives an object when asked around 8 months;
- communicates using first words at 12 months;
- uses a vocabulary of 50 words and can string 2-3 words together at 18 months;
- at around 3 years, the child can :
 - understand language about his/her daily activities;
 - use "I";
 - communicate and make sentences with subject/verb/object;
 - ask questions;
- at 5 years, the child can:

- speak without mangling words;
- use a wide vocabulary;
- understand and build complex sentences;
- talk about something that happened or tell a story.

The more serious the (bilateral) hearing loss, and the earlier its onset during spoken and written language acquisition, the more severe the disability will be.

2.2. Warning signs – approaches to management

In infants, any departure from the normal pattern of development should be met with special testing as quickly as possible.

In preschool-aged children, any small delay in language or speech acquisition should quickly lead to testing for hearing loss.

Chronic bilateral otitis with effusion leading to conductive hearing loss, which may in turn disrupt language acquisition, requires treatment.

Bilateral sensorineural hearing loss may occur in the course of meningitis or following ototoxic treatments. Its onset may also be progressive, secondary, and genetically driven.

The same goes for school-aged children.

Any bilateral sensorineural hearing loss should be addressed very quickly by a specialized team who will provide assessment and follow-up in the ENT, pediatric, SLP, and auditory prosthetics fields, as well as parental coaching and educational support.

Full coverage is provided for bilateral hearing aids until a patient's 20th birthday.

Cochlear implants are reserved for patients with profound bilateral hearing loss and no benefit from conventional hearing prostheses. They are advisable under such circumstances from the age of 12-18 months for congenital hearing loss and as early as possible under the same circumstances for secondary hearing loss.

3. Sensory disabilities in adults

3.1. General

Conductive hearing loss should be specially managed, potentially using with surgery.

Bilateral sensorineural hearing loss is most often progressive, primarily affecting the high frequencies, at least initially. Profound bilateral hearing loss may occur under specific circumstances:

Bilateral hearing loss occurring in adulthood may have various repercussions depending on its severity and the patient's age.

3.2. Repercussions

Any bilateral hearing loss of 35-40 dB in medium and high frequencies may be a serious social impediment:

3.3. Approaches to management

It should ideally be possible to rapidly equip any adult in need of bilateral hearing prostheses, given recent technological progress.

Speech therapy should be reserved for patients with profound/severe hearing loss who need rehabilitation with lip reading. Patients with profound hearing loss, if unable to benefit from conventional hearing aids, are also candidates for cochlear implantation.

Administratively speaking, workplace hearing loss is listed under workplace illnesses, Table 42, and conditionally entitles the claimant to compensation.

The various rubrics for assessing auditory disabilities under ordinary or labor law, or in the case of civil-service workers, define the degree of PPD (partial permanent disability) as ranging from 5% for mild bilateral hearing loss to 80% for bilateral hearing loss of 80 dB and above.

4. The specific case of profound or total unilateral hearing loss

In children, there are usually no repercussions for speech acquisition, but vigilance is recommended because the prevalence of this condition appears higher in children with dyslexia or dysorthographie than in the general population.

In adults, the onset of this condition (which systematically necessitates testing for retrocochlear involvement) leads to various complaints related mainly to a difficulty discerning speech in noisy environments: meetings, group meals, or vehicles (driver with right-side hearing loss).

These patients should be able to benefit from special bone-conduction hearing aids.