N.B. even moderate unilateral hearing loss may have implications for learning!

### Table 8.2. Classification of hearing loss by severity

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Normal hearing</th>
<th>Hearing loss in dB</th>
<th>Hearing loss in %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>&gt; 30 dB</td>
<td>0–30</td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>30–40 dB</td>
<td>30–40</td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td>40–60 dB</td>
<td>40–60</td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>60–90 dB</td>
<td>60–90</td>
<td></td>
</tr>
<tr>
<td>Profound</td>
<td>90–110 dB</td>
<td>90–110</td>
<td></td>
</tr>
<tr>
<td>Deafness</td>
<td>&gt; 110 dB</td>
<td>100</td>
<td></td>
</tr>
</tbody>
</table>

### I. CONDUCTION deafness

- defect in external or middle ear
  1. plugging of external auditory canals with wax or foreign bodies
  2. destruction of auditory ossicles
  3. eardrum thickening (followed repeated middle ear infections)
  4. abnormal rigidity of attachments of stapes to oval window.
  - air conduction is impaired while bone conduction remains normal (Rinne test “negative”; “air-bone gap” on audiometry). diagnosis can be confirmed by tympanometry.
  - hearing aids work well.
  - normal functional tympanic membrane contributes > 20 dB to hearing level.
  - patients speak with soft voice because, to them, their own voices sound louder than background sounds in environment.

### II. SENSORINEURAL deafness

- defect in inner ear ± CN VIII.

#### 1. SENSORY deafness - cochlear lesion (usually not life threatening but also incurable).
- 1. acoustic trauma (prolonged exposure to noise damages outer hair cells)
- 2. viral labyrinthitis
- 3. ototoxic drugs: aminoglycoside antibiotics (obstruct mechanosensitive channels in stereocilia = cell degeneration).
- 4. Ménière’s disease
- 5. presbycusis (gradual cumulative loss of hair cells and neurons).
  - most common is loss of hair cells of cochlea; spiral ganglion cells are often preserved for period of time but eventually degenerate because of lack of trophic factors (such as brain-derived neurotrophic factor) from hair cells.

#### 2. NEURAL deafness - CN VIII lesion (potentially fatal but curable):
- cerebellopontine angle tumors*, other neurologic disorders.
  - *in progressing unilateral sensorineural hearing loss perform MRI
  - to exclude cerebellopontine angle tumor (e.g. acoustic neuroma)!!
  - air and bone conduction are impaired equally – so they maintain normal relationship to each other.
  - patients tend to speak with loud voice.
  - differentiation from conduction deafness is by sample tests with tuning fork (Rinne, Weber, Schwindach – see p. Diagn 2)

**Differentiation of sensory vs. neural hearing losses:**

<table>
<thead>
<tr>
<th>Test</th>
<th>Sensory hearing loss</th>
<th>Neural hearing loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>Speech discrimination</td>
<td>moderate decrement</td>
<td>severe decrement</td>
</tr>
</tbody>
</table>

**Hearing Loss, Deafness -** general term for hearing loss, without designation of degree or cause (i.e. any degree of hearing loss may be described as deafness).

By age 75 years, 360 of 1000 adults have disabling hearing loss.
Amplification of sound

1. Cortical deafness is essentially combination of:
   a. Pure word deafness - disturbance of spoken language comprehension and repetition; no problems with reading or writing; nonverbal sounds are correctly identified.
   b. Auditory AGNOSIA - ability to fuse incomplete or partial messages.
   c. Discrimination of distorted speech.
   d. PURE WORD DEAFNESS - inability to interpret (recognize) auditory stimuli, but inability to interpret (recognize) nonverbal sounds.

Pathologic adaptation (tone decay)

1. Acoustic reflex decay: absent or mild
2. Pathologic tuning (tone decay): absent or mild
3. Waveforms in auditory brainstem responses: absent or with abnormally long latencies

Hearing Aids (s. Amplification)

Methods of Communication for Deaf

1. Auditory-oral - enhancement of residual hearing (amplification) + lipreading skills.
2. Cued speech - hand cues to supplement information received from lipreading.
3. Manualism - manual alphabet (fingerspelling) and sign language (e.g. American Sign Language "ASL").

Management

- Intratympanic (IT) methylprednisolone and oral prednisone are equally effective for treatment of idiopathic sudden sensorineural hearing loss.

Hearing Loss, Deafness

### Performance-Intensity Function (fication with increasing intensity)

<table>
<thead>
<tr>
<th>Performance-Intensity Function</th>
<th>improves</th>
<th>deteriorates</th>
</tr>
</thead>
<tbody>
<tr>
<td>Function</td>
<td>tone losses</td>
<td>hair loss</td>
</tr>
</tbody>
</table>

### Recruitment (abnormal increase in perception of loudness or inability to hear loud sounds normally despite hearing loss)

<table>
<thead>
<tr>
<th>Recruitment</th>
<th>normal or</th>
<th>absent or</th>
<th>even recruitment</th>
</tr>
</thead>
<tbody>
<tr>
<td>function</td>
<td>sensation of</td>
<td>loudness in</td>
<td>intensity than</td>
</tr>
<tr>
<td>affected ear</td>
<td>it does in</td>
<td>normal ear</td>
<td></td>
</tr>
</tbody>
</table>

### Acoustic reflex decay

<table>
<thead>
<tr>
<th>Acoustic reflex decay</th>
<th>absent or mild</th>
</tr>
</thead>
</table>

### Pathologic adaptation (tone decay)

<table>
<thead>
<tr>
<th>Pathologic adaptation (tone decay)</th>
<th>absent or mild</th>
</tr>
</thead>
</table>

### Huschke's response

<table>
<thead>
<tr>
<th>Huschke's response</th>
<th>absent or present</th>
</tr>
</thead>
</table>

### Waveforms in auditory brainstem responses

<table>
<thead>
<tr>
<th>Waveforms in auditory brainstem responses</th>
<th>absent or with abnormal latency</th>
</tr>
</thead>
</table>

### III. CENTRAL deafness - defect in cochlear nuclei + auditory CNS pathways - extremely rare

1. Bilateral representation, unilateral CNS lesions do not produce deafness (i.e. do not result in elevation of pure-tone thresholds or in decreased discrimination of single words). Diagnosis is not made by pure tone audiogram (which often yields normal result) - special tests are required.

2. Discrimination of distorted speech (with low-frequency or high-frequency filters, periodic interruptions, or time compression) - lost in cortical lesions.

3. Ability to fuse incomplete or partial messages delivered to each ear into meaningful message - lost in brainstem lesions.

4. Ability to localize sound in space (median plane localization) when acoustic stimuli are delivered simultaneously to both ears - lost in brainstem lesions.

Cortical deafness is essentially combination of:

- Auditory-oral - enhancement of residual hearing (amplification) + lipreading skills.
- Cued speech - hand cues to supplement information received from lipreading.
- Manualism - manual alphabet (fingerspelling) and sign language (e.g. American Sign Language "ASL").

### METHODS OF COMMUNICATION FOR DEAF

- Amplification of sound - helps almost all persons with conductive or sensorineural hearing losses. With many models, microphone can be switched off and magnetic coil used to enhance clarity when talking on telephone.
- Best models are adjusted to particular pattern of hearing loss.
  - Gain refers to difference between input and output of hearing aid (more severe hearing loss, more gain is required).
  - Frequency response - gain as function of frequency; as general rule, frequency response is selected to provide gain consistent with patient's audiometric configuration.
  - Saturation level - maximum output of hearing aid regardless of input - important consideration for patients with reduced tolerance to sound (as in recruitment).

### Major side effect - feedback (esp. with high gain) H: devices implantable into middle ear cavity.

### Air Conduction Aids

- coupled to ear canal with airtight seal or open tube.

- A. Body aid (for profound hearing loss) - most powerful - worn in shirt pocket or body harness.
- B. Postauricular aid (for moderate to severe hearing loss) - fits behind pinna.
- C. In-the-ear aid (for mild to moderate hearing loss) - contained entirely within ear mold.
- D. Canal aid - contained entirely within ear canal; difficult for some persons (especially elderly) to manipulate.

### Bone Conduction Aids

- Provide sound conduction through temporal bone to stapes capsule.
  - Indicated in conductive hearing loss when external (e.g. euraxis) or middle (e.g. chronic otitis media) ear factors make use of conventional hearing aids impossible.
  - Bone conduction must be 45 dB or better and speech discrimination score of 60% or better.
  - Oscillator is placed in contact with head (usually over mastoid), with spring band over head.
  - May not reduce noise, but better comfort to wear.

### Cochlear Implants

- Depend on stimulation of surviving spiral ganglion neurons (not hair cells that are degenerated).
- Indicated for profound bilateral sensorineural hearing loss that cannot be helped by hearing aids.
  - In case of congenital deafness, implantation must be performed before puberty (later, symmetrical in both of Held degeneration - implantation is not effective).
  - Contraindications - cochlear aplasia, absence of auditory nerve, active middle ear infection, lesions of brain stem.
  - Contains battery-powered processor (converts sound into modulations of electric current), internal and external induction coil system (transmits electrical impulses through skin), and array of electrodes connected to internal induction coil (stimulates remaining fibers of auditory division of CNS).
  - Always perform preimplantation high-resolution imaging (CT or T2-weighted fast spin echo MRI*) to evaluate cochleovestibular apparatus and internal auditory canals.

### Coaches for Students

- Voice projection
- Public speaking
- Auditory processing
- Time management
- Test anxiety

### Conclusion

- DEAFNESS can be a serious problem that affects many people around the world. It is important to understand the different types of hearing loss and the methods available to treat them. By using proper communication techniques, individuals with hearing loss can lead fulfilling lives and participate fully in society.

- In conclusion, hearing loss can have a significant impact on an individual's quality of life. It is important to seek medical attention and explore available options for treatment and communication.

- Again, the key to managing hearing loss is early detection and intervention. If you or someone you know is experiencing difficulties with hearing, it is important to consult a medical professional for a proper diagnosis and treatment plan.
• **MRI** better reveals fluid spaces of cochlea.

• **determine side of implant**:
  - implanting better-hearing ear, allows greater population of surviving spiral ganglion cells to receive electrical stimulation and, hence, potentially results in better outcome.
  - some patients are reluctant to implant their best-hearing (although poor-hearing) ear out of fear of implant failure.

• **electrode array** is inserted into scala tympani of basal turn in cochlea (via mastoidectomy and posterior tympanotomy);
  - **internal induction coil** is implanted into bone of skull posterior and superior to ear;
  - **external conduction coil** is held in place on skin over induction coil by magnets in two coils.

• **multichannel implants** are more effective than single-channel ones.

• enable deaf persons to hear and distinguish environmental sounds and warning signals; some wearers can discriminate words without visual clues and can talk on telephone; also help deaf persons modulate their voices to make their speech more intelligible (overall prognosis for hearing improvement and improved quality of life in properly selected patient is excellent!!!).

N.B. patients should be aware that any residual hearing in operated ear is lost after implantation.

N.B. patients deafened after **meningitis** must be followed closely with **serial imaging** - may develop **labyrinthitis ossificans** - implant as soon as diagnosis of early ossification or fibrosis is made; otherwise at least 6 months observation is indicated (fairly high hearing recovery rates in at least one ear).

---

**Fig. Cochlear implant (C)**

![Diagram of cochlear implant](source)

The sound is received by a directional microphone worn on the ear and fed to the speech processor as an analog signal. The speech processor, which is worn externally, processes the microphone signal, extracts the speech components that are necessary for comprehension, and converts them into a series of electrical impulses. A transmitting coil worn behind the ear transmits the impulses as radio frequencies (through the skin to the implanted portion of the C) to the implant. The necessary power supply is also transmitted; the implant itself does not require a separate power source.


**Current FDA cochlear implant guidelines**:

- SRT (speech reception threshold), PTA (pure-tone average)
Candidates aged 18 months to 18 years

- profound sensorineural hearing loss in both ears (SRT/PTA rated at “not useful” [i.e. ≥ 90 dB hearing loss])
- children < 5 years also must score ≤ 20% on sentence recognition tests under best-aided conditions (i.e. with best-fit hearing aids in place).
- all cochlear implant recipients must be vaccinated against pneumo coccal (1 of the 3 pneumoniae meningitis).

Adult candidates (no upper age limit)

- severe-to-profound sensorineural hearing loss in both ears (SRT/PTA rated ≤ 70 dB hearing loss) or score ≤ 30-40% on sentence recognition tests under best-aided conditions (i.e. with best-fit hearing aids in place).

**AUDITORY BRAINSTEM IMPLANTS**

- typically used in neurofibromatosis type 2, where tumors involving both cranial nerve VII & VIII complexes have rendered patient anacusic.
- device is implanted into 4th ventricle adjacent to cochlear nucleus (usually after tumor has been resected, during same operation).

**PREVENTION**

- single most successful way – limit damaging noise (< 85 dB / 8 h per day).
- also avoid ototoxic medications.

**OTITIS MEDIA**

- 1800 to 1/1000 newborns have severe to profound hearing loss at birth.
- hearing loss is No.1 birth defect (ahead of congenital heart defects and cleft lip/palate).
- N.B. most common cause (33-50% of childhood deafness is genetic).
- during childhood, another 2.5/1000 children acquire moderate to severe hearing loss.
- can result in lifelong impairments in receptive - expressive language skills.

**ETIOLOGY**

N.B. most common are acquired conductive losses associated with otitis media.

- Conductive hearing loss
  - A. Otitis media (esp. before age 6 months)!!1 (most commonly temporary hearing loss)
  - B. Malignations of external auditory canal / middle ear.
  - C. Cholesteatoma.

- Sensorineural hearing loss
  - If occurs prenatally - CONGENITAL sensorineural hearing loss.
  - If occurs during first two or year of life - EARLY-ONSET PROGRESSIVE sensorineural hearing loss.
  - If occurs later – PROGRESSIVE sensorineural hearing loss of childhood.

Causes of CONGENITAL sensorineural hearing loss:

- a) endogenous causes – genetic (> 50% congenital cases), teratologic, prematurity.
- b) exogenous causes – anoxia, infections (rubella, syphilis, CMV, toxoplasmosis, herpes), Rb incompatibility, ototoxic drugs given to mother.

Causes of ACQUIRED sensorineural hearing loss:

1. autoimmune disorders
2. ototoxic substances
3. bacterial meningitis (causes = 9% of childhood deafness), bacterial endotoxins and exotoxins
4. congenital / acquired viral infections (e.g. mumps!!, rubella, CMV)
5. sound trauma
6. head trauma resulting in temporal bone concussion / fracture.
7. malformed bones of bony labyrinth.
8. perilymphatic fistulas 
9. primary diseases of CNS in childhood are rare (most common are schwannomas in neurofibromatosis II).
10. severe kernicterus.

**DEAFNESS DUE TO GENETIC MUTATIONS**

- occurs in 0.1% newborns:
  - a) nonsyndromic deafness (70%)
  - b) syndromic deafness (30%) - associated with abnormalities in other systems.

**Non syndromic deafness**

- can first appear in adults rather than children (so incidence is > 0.1%).
- products of >100 gene complexes have rendered patient anacusic.
- one form of long QT syndrome - mutation of one of K+ channel proteins (KVLQT1) in stria vascularis (essential for maintaining high K+ concentration in endolymph), and in heart (helps maintain normal QT interval).
- Usher syndrome - sensorineural hearing loss & retinitis pigmentosa.
- Waardenburg syndrome
- Alport syndrome
- Cockayne syndrome
- Alstrom syndrome
- Reesem disease
- Branchiosotoral syndrome
- Mucopolysaccharidoses, sphingolipidoses

**PEDIATRIC HEARING DEFICITS**

- 1/800 to 1/1000 newborns have severe to profound hearing loss at birth.
  - hearing loss is No.1 birth defect (ahead of congenital heart defects and cleft lip/palate).
- N.B. most common cause (33-50% of childhood deafness is genetic).
- during childhood, another 2.5/1000 children acquire moderate to severe hearing loss.
- can result in lifelong impairments in receptive - expressive language skills.
Hearing Loss, Deafness

2. Eye:
   - Auricular deformity - Treacher-Collins syndrome, Goldenhar syndrome
   - External canal atresia or stenosis - Treacher-Collins syndrome, Goldenhar syndrome
   - Protrusor pits - Branchiootorenal syndrome
   - Prominent skin tags - Goldenhar syndrome

3. Integument:
   - Eccomedical dysplasia - Scharlach
   - Hypopigmentation - Albinism
   - Lentigines - Lentigines, electrographic abnormalities, ocular hypertelorism, pulmonary stenosis, abnormalities of genitalia, retardation of growth, and deafness (LEOPARD) syndrome
   - Whitefook - Waardenburg syndrome

4. Cardia:
   - Conductive defects - Jervell and Lange-Nielsen syndrome
   - Mitral insufficiency - Forney syndrome

5. Renal:
   - Dysfunction - Alport syndrome
   - Malformation - Goldenhar syndrome

6. Dental:
   - Abnormal dentos - Orofaccioaner imperfecta
   - Pegged (Hutchinson) incisors - Congenital syphilis

7. Endocrine / metabolic:
   - Goiter - Pendred syndrome
   - Hypogonadism - Alström syndrome
   - Obesity - Laurence-Bloom-Blind syndrome
   - Mucopolysaccharidosis - Hunt-Heller syndrome

8. Chromosomal abnormalities:
   - Trisomy 13 - Patau syndrome
   - Trisomy 18 - Edwards syndrome
   - Trisomy 21 - Down syndrome

9. Neurologic:
   - Ataxia - Spinocerebellar degeneration
   - Epilepsy - Hermann syndrome
   - Peripheral neuropathy - Flynn-Aird syndrome
   - Polynuropathy - Rilfan disease

10. Skeletal:
    - Deformations - Achondroplasia
    - Fusion of cervical vertebrae - Klippel-Feil syndrome
    - Lamb deformities - Orofaccioaner imperfecta, Blalock syndrome
    - Scoliosis, elongated limbs - Marfan syndrome
    - Synosty - Apert syndrome

11. Craniofacial:
    - Acrocephaly (tower skull) - Apert syndrome
    - Branchial fistulas - Branchiootorenal syndrome
    - Cleft palate, small mandible - Pierre Robin sequence
    - Cranial synostosis - Crouzon syndrome
    - Malar / facial bone anomalies - Treacher-Collins syndrome
    - Microcephaly - Crouzon syndrome
    - Occul / auricular anomalies - Goldenhar syndrome

Table 9.1 Congenital syndromes that are associated with hearing loss

<table>
<thead>
<tr>
<th>Type of hearing loss</th>
<th>Year of hearing loss</th>
<th>Typical features</th>
<th>Inheritance</th>
<th>Anomalies of the external ear</th>
<th>Mandibulofacial dysplasia(Treacher-Collins syndrome)</th>
<th>Autosomal-dominant</th>
<th>Anomalies of the external and middle ear</th>
<th>Autosomal-recessive</th>
<th>Autosomal-dominant</th>
<th>Autosomal-recessive</th>
<th>Autosomal-recessive</th>
</tr>
</thead>
</table>

Hearing Loss, Deafness

Table 9.2: Causes of acquired hearing loss in newborns and infants

<table>
<thead>
<tr>
<th>Type of Hearing Loss</th>
<th>Example</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prelingual</td>
<td>Infections, trauma, ototoxicity, congenital malformations, genetic syndromes, trauma, neoplasms</td>
</tr>
<tr>
<td>Perinatal</td>
<td>Congenital infections, ototoxicity, congenital malformations, genetic syndromes, trauma, neoplasms</td>
</tr>
</tbody>
</table>

Diagnosis

All newborns, infants, and children should be screened for hearing loss!!!

- Diagnosis is usually significantly delayed:
  1. Severe losses are usually diagnosed by age 2 yr.
  2. Moderate and unilateral losses are typically not recognized until school age.

- Diagnosis must be made as early as possible to ensure adequate linguistic input for optimal language development.

- Special audiometric techniques can assess hearing starting at birth:
  1. Reflexive responses (startle responses or blinking in children < 6 months).
  2. Behavioral responses (turning head to sound source in children > 6 months).
  3. Conditioned orientation response audiometry (lighted toy mounted on loudspeaker is flashed after presentation of test tone; after undergoing brief conditioning period, child localizes toward tone, if audible, in anticipation of flashing toy; recorded threshold is called minimal response level, since true thresholds may be slightly lower than levels required to elicit these behavioral responses).
  4. Auditory brain stem response audiometry
  5. Otoacoustic emissions testing

- Many states are mandating universal newborn screening with auditory brain stem response audiometry & otoacoustic emissions testing.

- From 3-5 years formal (pure tone) audiologic screening may be used; failure to respond to 1000 or 2000 Hz at 20 dB or 4000 Hz at 25 dB in either ear → formal audiologic testing.

- If child does not develop speech normally → consider deafness / mental retardation / aphasia / autism.

- Many children with sensorineural hearing losses have vestibular deficits → delayed / regressive motor development.

Treatment

- Objective is to support optimal language development.
  1. First year of life is critical period for language development.
  2. Deaf children will develop language only with special training, ideally beginning as soon as hearing loss is identified.
  3. Special educational intervention! (seating in front of classroom, placement in residential school).
  4. Amplification (hearing aid) should be started as early as possible (even by 6 weeks of age).
  5. Children age ≥ 2 yr with profound bilateral hearing loss may be candidates for cochlear implant (more effective in those who already have developed language).
  6. Children whose acoustic nerves have been destroyed by tumor may be helped by implantation of brain stem auditory stimulating electrodes.
  7. Final indication of success of habilitative program is child's language capability and not level of hearing.

Psychogenic Deafness

- More common in malingering than in conversion disorder (hysteria).
- Truly deaf patients seek sensory input from their environment, watch examiner intently, and turn their good ear toward speaking voice.
- Patients with true long-standing hearing loss may speak loudly.

PSYCHOGENIC DEAFNESS

- True unilateral deafness from psychogenic deafness:
  1. Patient wears headphones and listens to story being told, part in one ear and some in the other → patient is tested on information.
  2. Auditory evoked responses and measurement of stapedius reflex.
  3. If psychogenic hearing loss is bilateral, loud noise (e.g. clapping hands) may produce oculopalpebral reflex.

BIBLIOGRAPHY for ch. “Otology” → follow this LINK >>