Hearing Loss, Deafness

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**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.

Classification, Diagnosis

|  |  |
| --- | --- |
| **Mild loss** – 20-40 dB loss  **Moderate loss** – 40-60 dB loss  **Severe loss** – 60-90 dB loss  **Profound loss** – 90-110 dB loss in speech frequency – unable to process language through audition (even with hearing aid).  **Deafness** > 110 dB loss. |  |

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



[Source of picture: Rudolf Probst, Gerhard Grevers, Heinrich Iro “Basic Otorhinolaryngology” (2006); Publisher: Georg Thieme Verlag; ISBN-10: 1588903370; ISBN-13: 978-1588903372 >>](http://www.amazon.com/gp/product/1588903370)

**I. Conduction deafness** – defect in **external ÷ middle ear**.

1. plugging of external auditory canals with wax or foreign bodies
2. destruction of auditory ossicles
3. eardrum thickening (following 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.
     + loss of ***spiral ganglion cells*** can occur, with no or minimal loss of hair cells.
   1. **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 conductions are impaired equally* – so they maintain normal relationship to each other.
* patients tend to ***speak with loud voice***.
* differentiation from conduction deafness is by simple tests with tuning fork (Rinne, Weber, Schwabach → see [p. D1ear >>](http://www.neurosurgeryresident.net/D.%20Diagnostics\D1-5.%20Neurologic%20Examination\D1ear.%20Otologic%20Examination.pdf#Tuning_fork_tests)).

Differentiation of sensory vs. neural hearing losses:

|  |  |  |
| --- | --- | --- |
| **Test** | **Sensory hearing loss** | **Neural hearing loss** |
| **Speech discrimination** | ***moderate*** decrement | ***severe*** decrement |
| **Performance-intensity function** for phonetically balanced words (i.e. discrimination with increasing intensity) | **improves** | **deteriorates** (“rollover”)!!! - characteristic of CN8 nerve lesions |
| **Recruitment** (abnormal increase in perception of loudness or ability to hear loud sounds normally despite hearing loss) | **present** (i.e. sensation of loudness in affected ear increases *more* with each increment in intensity than it does in normal ear) | **absent** or even **decruitment** (i.e. sensation of loudness in affected ear increases *less* with each increment in intensity than it does in normal ear) |
| **Acoustic reflex decay** | **absent** (or mild) | **present** |
| **Pathologic adaptation (tone decay)** | **absent** (or mild) | **marked** (i.e. patient cannot continue to perceive constant tone above hearing threshold) |
| **Otoacoustic emissions** | **absent** | **present** |
| Waveforms in **auditory brain stem responses** | well formed, with normal latencies | absent or with abnormally long latencies |

**III. Central deafness** – defect in **cochlear nuclei ÷ auditory CNS pathways** - extremely rare!

N.B. due to 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:

1. **Discrimination of distorted speech** (with low-frequency or high-frequency filters, periodic interruptions, or time compression) – lost in *cortical* lesions.
2. **Discrimination in presence of competing message** in other ear – 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: [also see p. S2 >>](http://www.neurosurgeryresident.net/S.%20Symptoms,%20Signs,%20Syndromes\S01-09.%20Language,%20Memory,%20Praxis\S02.%20Language%20disorders.pdf)

1. **pure word deafness** - disturbance of *spoken language* *comprehension* and repetition; no problems with reading or writing; nonverbal sounds are correctly identified.
2. **auditory agnosia** - relatively normal pure tone hearing on audiometry, but inability to *interpret (recognize)* *nonverbal sounds* (such as ringing of telephone) with preserved ability to interpret speech.
3. preserved *awareness of sound occurrence* (for instance, by startle reaction to clap).

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 “Ameslan”).

Management

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

Hearing Aids (s. Amplification)

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.

1. **Body aid** (for profound hearing loss) - most powerful - worn in shirt pocket or body harness.
2. **Postauricular aid** (for moderate to severe hearing loss) - fits behind pinna.
3. **In-the-ear aid** (for mild to moderate hearing loss) - contained entirely within ear mold.
4. **Canal aid** - contained entirely within ear canal; difficult for some persons (especially elderly) to manipulate.
5. **CROS aid** (Contralateral Routing Of Signals) - for severe unilateral hearing loss; microphone is placed in nonfunctioning ear, and sound is routed to functioning ear - enables to hear sounds from nonfunctioning side and to develop limited ability to localize sound; if better ear also has some hearing loss, sound from both sides can be amplified with **BICROS aid.**

**Bone conduction aids**

- provide sound conduction through temporal bone to otic capsule.

* indicated in **conductive hearing loss** when external (e.g. aural atresia) or middle (e.g. chronic otorrhea) 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.
* require more power, introduce more distortion, and are less comfortable to wear.
* can be implanted in mastoid process.

Cochlear Implants

* depends 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, synapses in ***bulb of Held*** degenerate – implantation is not effective).
* contraindications - cochlear aplasia, absence of auditory nerve, active middle ear infection, lesions of brain stem.
* consists of *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 CN8).
* always perform *preimplantation high-resolution imaging* (CT or T2-weighted fast spin echo MRI\*) to evaluate cochleovestibular apparatus and internal auditory canals.

\* MRI better reveals fluid spaces of cochlea.

* better use general anesthesia.
* 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.

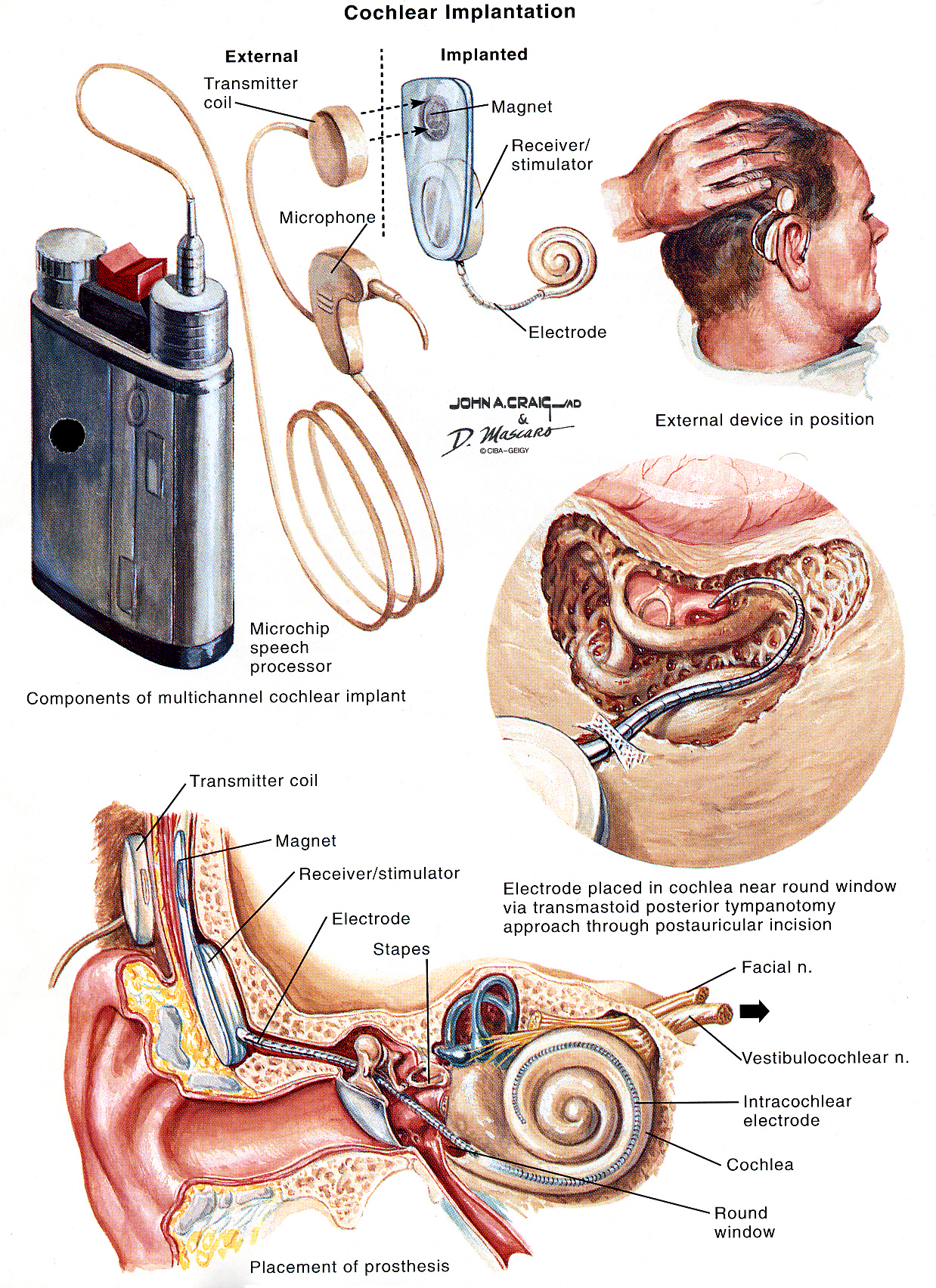
* **multichannel** implants are more effective than **single-channel** ones.
* enable deaf persons to hear and distinguish environmental sounds and warning signals; some wearers can discriminate words without visual clues and can talk on telephone; also help deaf persons modulate their voices to make 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).



[Source of picture: Rudolf Probst, Gerhard Grevers, Heinrich Iro “Basic Otorhinolaryngology” (2006); Publisher: Georg Thieme Verlag; ISBN-10: 1588903370; ISBN-13: 978-1588903372 >>](http://www.amazon.com/gp/product/1588903370)



[Source of picture: Frank H. Netter “Clinical Symposia”; Ciba Pharmaceutical Company; Saunders >>](http://www.amazon.com/gp/product/1933247401)

**Current FDA cochlear implant guidelines**:

SRT (speech reception threshold), PTA (pure-tone average)

**Candidates aged 18 months ÷ 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 pneumococci! (↑risk of Str. 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**.

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*.

Etiology

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

**Conductive hearing loss**

1. Otitis media (esp. before age 6 months)!!! (most commonly temporary hearing loss)
2. Malformations of external auditory canal / middle ear.
3. Cholesteatoma.

**Sensorineural hearing loss**

If occurs **prenatally** - congenital sensorineural hearing loss.

If occurs during **first year or two of life** - early-onset progressive sensorineural hearing loss.

If occurs **later** - progressive sensorineural hearing loss of childhood.

Causes of **congenital** sensorineural hearing loss:

1. **endogenous causes** – genetic (≈ 50% congenital cases), teratologic, prematurity.
2. **exogenous causes** – anoxia, infections (rubella, syphilis, CMV, toxoplasmosis, herpes), Rh 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. **malformations** of bony labyrinth.
8. **perilymphatic fistulas** (associated with minor head trauma, labyrinthine malformations).
9. **primary diseases of CN8** in childhood are rare (most common are schwannomas in neurofibromatosis II).
10. severe **kernicterus**.

Deafness due to genetic mutations

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

Nonsyndromic deafness

* can first appear in adults rather than children (so incidence is > 0.1%).
* products of > 100 genes are essential for normal hearing (deafness loci have been described in all but five of 24 chromosomes).
* examples of proteins which when mutated cause deafness:

1. **connexin 26** (normal recycling of K+ through sustentacular cells) ≈ 40% genetic deafness cases!!!
2. three **nonmuscle myosins**:

* **myosin-VIIa**, associated with actin in hair cell processes;
* **myosin-Ib**, part of "adaptation motor" that adjusts tension on tip links;
* **myosin-VI**, essential in some way for formation of normal cilia.

1. **α-tectin** (one of major proteins in tectorial membrane).

Syndromic deafness – more than 70 phenotypically distinct syndromes are described:

1. **Pendred syndrome** - mutant *sulfate transport protein* causes deafness and goiter.
2. 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).
3. **Usher** **syndrome** - sensorineural hearing loss & retinitis pigmentosa.
4. **Waardenburg** **syndrome**
5. **Alport** **syndrome**
6. **Cockayne syndrome**
7. **Alström syndrome**
8. **Refsum disease**
9. **Branchiootorenal syndrome**
10. Mucopolysaccharidoses, sphingolipidoses

Selected physical findings important in evaluation of syndromic hearing loss:

1. **Ear**:

* Auricular deformity - *Treacher-Collins syndrome, Goldenhar syndrome*
* External canal atresia or stenosis - *Treacher-Collins syndrome, Goldenhar syndrome*
* Preauricular pits - *Branchiootorenal syndrome*
* Preauricular skin tags - *Goldenhar syndrome*

1. **Eye**:

* Cataracts - *Congenital rubella*
* Coloboma - Colomba of iris, heart deformities, choanal atresia, retarded growth, genital and ear deformities *(CHARGE) association*
* Dystopia canthorum - *Waardenburg syndrome*
* Heterochromia iridis - *Waardenburg syndrome*
* Keratitis - *Cogan syndrome*
* Ocular palsy - *Duane syndrome*
* Retinal atrophy - *Cockayne syndrome*
* Retinitis pigmentosum - *Usher syndrome*

1. **Integumentum**:

* Ectodermal dysplasia - *Ichthyosis*
* Hypopigmentation - *Albinism*
* Lentigines - Lentigines, electrocardiographic abnormalities, ocular hypertelorism, pulmonary stenosis, abnormalities of genitalia, retardation of growth, and deafness *(LEOPARD) syndrome*
* White forelock - *Waardenburg syndrome*

1. **Cardiac**:

* Conduction defects - *Jervell and Lange-Nielsen syndrome*
* Mitral Insufficiency - *Forney syndrome*

1. **Renal**:

* Dysfunction - *Alport syndrome*
* Malformation - *Goldenhar syndrome*

1. **Dental**:

* Abnormal dentin - *Osteogenesis imperfecta*
* Pegged (Hutchinson) incisors - *Congenital syphilis*

1. **Endocrine / metabolic**:

* Goiter - *Pendred syndrome*
* Hypogonadism - *Alström syndrome*
* Obesity - *Laurence-Moon-Biedl syndrome*
* Mucopolysaccharidosis - *Hunter/Hurler syndrome*

1. **Chromosomal abnormalities**:

* Trisomy 13 - *Patau syndrome*
* Trisomy 18 - *Edwards syndrome*
* Trisomy 21 - *Down syndrome*

1. **Neurologic**:

* Ataxia - *Spinocerebellar degeneration*
* Epilepsy - *Herman syndrome*
* Peripheral neuropathy - *Flynn-Aird syndrome*
* Polyneuropathy - *Refsum disease*

1. **Skeletal**:

* Dwarfism - *Achondroplasia*
* Fusion of cervical vertebrae - *Klippel-Feil syndrome*
* Limb deformities - *Osteogenesis imperfecta, Hurler syndrome*
* Scoliosis, elongated limbs - *Marfan syndrome*
* Syndactyly - *Apert syndrome*

1. **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*
* Midface hypoplasia - *Crouzon syndrome*
* Ocular / auricular anomalies - *Goldenhar syndrome*



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Diagnosis

[Also see p. D5 >>](http://www.neurosurgeryresident.net/D.%20Diagnostics\D1-5.%20Neurologic%20Examination\D5.%20Pediatric%20Neurologic%20Examination.pdf#Cranial_nerves)

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

* diagnosis is usually significantly delayed:
* severe losses are usually diagnosed by age 2 yr;
* mild to moderate and unilateral losses are typically not recognized until school age.
* diagnosis must be made as early as possible → adequate linguistic input for optimal language development.
* **special audiometric techniques** can assess hearing *starting at birth* - tests measure reflexive, behavioral, and physiologic responses to auditory stimuli of controlled intensity; examples:
  1. **startle responses** or **blinking** – children < 6 months.
  2. **turning head to sound source** – 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!

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

N.B. effect of unilateral hearing losses is often underestimated - difficulty in identifying speech in moderately noisy backgrounds → *significant language deficits*. H: system at school that allows teacher to speak into microphone that sends signals to hearing aid in child's good ear.

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 good ear toward speaking voice.
* patients with true long-standing hearing loss may speak loudly.

Tests to distinguish true unilateral deafness from psychogenic deafness:

1. patient ***wears headphones and listens to story*** being told, part in one ear and some in another → 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 >>](http://www.neurosurgeryresident.net/Ear.%20Otology\Ear.%20Bibliography.pdf)

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