THE DEAF CHILD

MedMentor EDU

ENT Study Notes

THE DEAF CHILD

Childhood Hearing Loss | Diagnosis | Management | Cochlear Implant

SECTION 1 | Introduction & Definitions

1.1 Definition of Deaf Child

A deaf child is a child under 18 years of age with a significant bilateral hearing loss that interferes with normal speech, language, and educational development.

- Clinically significant: hearing loss >40 dB HL in the better ear
- Includes prelingual (before speech) and postlingual (after speech development) onset

1.2 Definition of Hearing Impairment

Hearing impairment is any degree of reduction in the ability to perceive sound, ranging from mild to profound.

- Measured by Pure Tone Audiometry (PTA)
- Average of hearing thresholds at 500 Hz, 1000 Hz, 2000 Hz, 4000 Hz

1.3 WHO Classification of Hearing Loss

Grade	Hearing Threshold (dB HL)	Description
0 — Normal	25 dB or better	No impairment
1 — Slight	26–40 dB	Difficulty with faint speech
2 — Moderate	41–60 dB	Difficulty with normal speech
3 — Severe	61–80 dB	Difficulty with loud speech
4 — Profound	81 dB or more	Unable to understand even amplified spee

COMMON MCQ: WHO Grade 4 (Profound, >80 dB) is the cochlear implant candidate range — most commonly asked.

1.4 Epidemiology & Burden

- 1–3 per 1000 live births affected by significant hearing loss
- Most common sensory disability in children globally
- ~50% of childhood deafness is preventable
- Estimated 34 million children worldwide affected (WHO data)
- India: ~6.3% of population has significant hearing loss; children are a major subset

1.5 Preventable Causes of Childhood Deafness

- Immunization against rubella, measles, mumps
- Antenatal care and TORCH screening
- Neonatal hyperbilirubinemia management
- Avoidance of ototoxic drugs in pregnancy and neonates
- Prompt treatment of meningitis and otitis media
- Noise exposure prevention

1.6 Importance of Early Diagnosis

- Brain plasticity is maximal in first 3 years of life
- Delayed diagnosis ? missed critical period ? irreversible language delay
- 1-3-6 Rule (JCIH):
    - Screening by 1 month of age
    - Diagnosis by 3 months
    - Intervention by 6 months

HIGH-YIELD: 1-3-6 Rule (JCIH): Screen by 1 month, Diagnose by 3 months, Intervene by 6 months — extremely high-yield MCQ.

1.7 Critical Period of Speech & Language Development

- Birth to 3 years: most critical period for auditory and speech development
- Language centres in brain require early auditory input for maturation
- Hearing loss during this period causes: absent speech development, poor language, educational failure
- Late implantation (>5 years) results in poor cochlear implant outcomes

1.8 Effect of Deafness on Communication & Education

- Absent or distorted speech development
- Inability to follow verbal instructions in school
- Social isolation and psychological impact
- Learning disability and academic underperformance
- Behavioral problems due to communication frustration

SECTION 2 | Normal Auditory & Speech Development

2.1 Auditory Development in Infancy

- Hearing begins in utero by 20–24 weeks of gestation
- Newborn responds to sudden loud sounds (Moro/startle reflex)
- Auditory brainstem pathways mature by 2 years of age

2.2 Auditory Milestones

Age	Expected Auditory Response
0–3 months	Startle to loud sound; quietens to mother's voice
3–6 months	Turns eyes toward sound source
6–9 months	Localizes sound to side; responds to name

9–12 months	Localizes sound accurately above and below
12–18 months	Identifies familiar objects by name
18–24 months	Points to named body parts; follows simple commands

2.3 Speech Milestones

Age	Speech Expected
0–2 months	Cooing; vegetative sounds
2–6 months	Babbling begins; vowel sounds
6–9 months	Reduplicated babble (ma-ma, ba-ba)
9–12 months	Jargon; mama/dada with meaning
12–18 months	3–10 single words with meaning
18–24 months	2-word phrases; 50-word vocabulary

2–3 years	Short sentences; strangers understand ~75%
3–4 years	Full sentences; 90% intelligible to strangers

2.4 Red Flag Signs of Deafness

HIGH-YIELD: These are favorite viva/MCQ topics — memorize age-specific red flags.

Age	Red Flag Sign
0–3 months	No startle to loud sound; does not quieten to voice
3–6 months	No babbling; does not turn to sound
6–12 months	No localization of sound; no response to name
12–18 months	No single meaningful words
18–24 months	No 2-word phrases; vocabulary <10 words
>2 years	Speech regression; unclear articulation

2.5 Delayed Speech Indicators

- Absence of babbling by 9 months
- No single words by 12–14 months
- No 2-word combinations by 24 months
- Sudden loss of language (acquired hearing loss)
- Speaks too loudly; watches lips closely (lip reading compensation)
- Inattention in class; apparent behavioral problems

SECTION 3 | Classification of Deafness

3.1 By Time of Onset

Type	Definition	Key Point
Congenital	Present at or before birth	Usually genetic or intrauterine
Acquired	Develops after birth	Meningitis, ototoxicity, noise
Prelingual	Before speech development (<2 yrs)	Severe language delay
Postlingual	After speech development (>2 yrs)	Better prognosis for CI

COMMON MCQ: Prelingual deafness has WORSE cochlear implant prognosis compared to postlingual deafness.

3.2 By Type of Hearing Loss

Conductive Hearing Loss (CHL)

- Defect in sound transmission through external or middle ear
- Air-bone gap present on audiometry
- Causes in children: otitis media with effusion, wax, aural atresia, ossicular abnormalities
- Maximum loss: 60 dB HL
- Usually treatable — surgical or medical

Sensorineural Hearing Loss (SNHL)

- Defect in cochlea (sensory) or auditory nerve (neural)
- No air-bone gap; both air and bone conduction reduced
- Causes: genetic, meningitis, ototoxicity, congenital CMV
- Usually permanent; managed with hearing aids or cochlear implant

Mixed Hearing Loss

- Combined CHL and SNHL components
- Air-bone gap present with elevated bone conduction threshold
- Example: chronic otitis media with co-existing SNHL from ototoxic drops

Central Hearing Loss

- Normal cochlea and auditory nerve but abnormal processing in CNS
- Child hears but cannot interpret speech
- Also called Auditory Processing Disorder (APD)

3.3 Syndromic vs Non-Syndromic Deafness

Feature	Syndromic	Non-Syndromic
Definition	Deafness + other organ involvement	Isolated hearing loss only
Proportion	~30% of genetic deafness	~70% of genetic deafness
Examples	Waardenburg, Usher, Pendred	DFNB1 (Connexin 26)
Inheritance	Various (AR, AD, X-linked)	Mostly AR

SECTION 4 | Etiology of Childhood Deafness

4.1 Genetic Causes

Overview

- ~50% of congenital deafness is genetic
- Majority autosomal recessive (~80%)
- Autosomal dominant: ~15–20%
- X-linked and mitochondrial: ~1–2% each

Autosomal Recessive Deafness

- Most common genetic cause
- Parents are carriers but have normal hearing
- DFNB1 locus — most common: Connexin 26 (GJB2 gene) mutation
    - GJB2 mutation: most common cause of non-syndromic SNHL worldwide
    - Disrupts gap junctions in cochlea ? impaired K+ recycling ? hair cell death

Autosomal Dominant Deafness

- DFNA loci — multiple mutations
- Often progressive hearing loss
- Parent usually affected

X-Linked Deafness

- Rare; affects males predominantly
- Mutations in PRPS1, POU3F4 genes
- POU3F4: associated with congenital X-linked perilymphatic gusher during stapes surgery

Mitochondrial Deafness

- Maternally inherited mutations in mtDNA
- 1555A>G mutation: aminoglycoside hypersensitivity
    - Even small doses of gentamicin cause profound SNHL in these patients

HIGH-YIELD: Connexin 26 / GJB2 mutation = most common cause of non-syndromic hereditary SNHL. 1555A>G mutation = aminoglycoside hypersensitivity.

4.2 Syndromic Deafness

Syndrome	Key Features	Type of HL	Inheritance
Waardenburg Syndrome	White forelock, heterochromia irides, dystopia canthorum, SNHL	SNHL	AD
Usher Syndrome	SNHL + Retinitis pigmentosa (progressive vision loss)	SNHL (profound)	AR
Pendred Syndrome	SNHL + Goitre (thyroid enlargement) + Mondini dysplasia	SNHL	AR
Alport Syndrome	SNHL + Nephritis + Ocular abnormalities	SNHL	X-linked
Jervell & Lange-Nielsen	Profound SNHL + Long QT syndrome (risk of sudden death)	SNHL	AR
Treacher Collins	Malar hypoplasia, micrognathia, EAC/middle ear atresia	CHL	AD
Down Syndrome	Trisomy 21; recurrent OM + occasional SNHL	CHL / Mixed	Chromosomal

CHARGE Syndrome	Coloboma, Heart defect, choanal Atresia, Retarded growth, GU and Ear anomalies	Mixed	AD

HIGH-YIELD: Jervell & Lange-Nielsen: SNHL + Long QT ? sudden cardiac death — extremely important. Usher = SNHL + retinitis pigmentosa. Pendred = SNHL + goitre.

COMMON MCQ: Treacher Collins causes CONDUCTIVE hearing loss (middle ear atresia) — not SNHL.

4.3 Prenatal Causes — TORCH Infections

Agent	Type of HL	Key Points
Rubella	SNHL	Most common infectious cause; 1st trimester most dangerous; also causes heart defects, cataracts
CMV (Cytomegalovirus)	SNHL	Most common non-genetic congenital SNHL; may be asymptomatic at birth; progressive HL
Toxoplasmosis	SNHL	Intracranial calcifications; chorioretinitis
Syphilis	SNHL	Hutchinson teeth, interstitial keratitis; bilateral SNHL

Herpes simplex	SNHL	Rare; neonatal HSV meningitis

HIGH-YIELD: CMV = most common non-genetic cause of congenital SNHL. Rubella = most common infectious cause. Syphilis triad = deafness + interstitial keratitis + Hutchinson teeth.

Other Prenatal Causes

- Maternal drug exposure: thalidomide, quinine, aminoglycosides
- Radiation exposure in first trimester
- Maternal diabetes ? SNHL in child
- Hypothyroidism in mother ? congenital hearing loss (Pendred-like)

4.4 Perinatal Causes

- Prematurity (<32 weeks gestation) ? immature cochlea, NICU exposure
- Low birth weight (<1500 g) — independent risk factor
- Birth asphyxia — hypoxic-ischemic cochlear damage
- Kernicterus — bilirubin deposits in cochlear nuclei and auditory brainstem
    - Causes auditory neuropathy/dys-synchrony — BERA absent but OAE present
- NICU stay >5 days — high-risk criterion for hearing screening
- Ototoxic medications in neonates: furosemide, gentamicin, vancomycin
- Mechanical ventilation — associated with aminoglycoside use and hypoxia

HIGH-YIELD: Kernicterus ? Auditory Neuropathy Spectrum Disorder (ANSD): OAE present but ABR absent or abnormal. Very important MCQ.

4.5 Postnatal Causes

- Bacterial meningitis: most common acquired cause of bilateral profound SNHL in children
    - Streptococcus pneumoniae — most damaging organism
    - Risk of cochlear ossification ? urgent cochlear implant consideration

- Viral infections: measles, mumps (unilateral SNHL), varicella
- Chronic otitis media — predominantly conductive HL; SNHL if cholesteatoma or ototoxics
- Noise-induced hearing loss — rock concerts, headphone use in adolescents
- Head trauma — temporal bone fracture
- Autoimmune inner ear disease — rare in children; bilateral progressive SNHL
- Ototoxic drugs (see table below)

Drug Class	Examples	Type of HL
Aminoglycosides	Gentamicin, Streptomycin, Tobramycin, Amikacin	SNHL (permanent)
Loop diuretics	Furosemide, Ethacrynic acid	SNHL (usually reversible)
Antineoplastic	Cisplatin, Carboplatin	SNHL (permanent, dose-dependent)
Antimalarials	Quinine, Chloroquine	SNHL (reversible)
Salicylates	Aspirin	SNHL (reversible, tinnitus)
Vancomycin	Vancomycin (esp. with aminoglycosides)	SNHL

COMMON MCQ: Cisplatin causes dose-dependent, cumulative, irreversible SNHL beginning at high frequencies — very common exam question.

4.6 Congenital Inner Ear Anomalies

Anomaly	Description	Key Points
Michel Aplasia	Complete absence of inner ear	Severest form; cochlear implant co
Mondini Deformity	Cochlea has only 1.5 turns (normal 2.5)	Risk of CSF gusher during surgery
Scheibe Dysplasia	Membranous labyrinth aplasia; bony labyrinth normal	Most common inner ear malforma
Common Cavity	Cochlea and vestibule fused into single cavity	No modiolus; CI possible with cau
Large Vestibular Aqueduct (EVA)	Vestibular aqueduct >1.5 mm at midpoint	Progressive SNHL; avoid head tra
Cochlear Nerve Deficiency	Absent/hypoplastic cochlear nerve	Cochlear implant contraindicated

HIGH-YIELD: Scheibe dysplasia = most common inner ear malformation. Michel aplasia = most severe (CI contraindicated). Mondini = 1.5 cochlear turns + CSF gusher risk.

COMMON MCQ: EVA (Enlarged Vestibular Aqueduct) = progressive SNHL triggered by head injury or straining.

SECTION 5 | Clinical Evaluation

5.1 History Taking

Antenatal History

- Maternal infections (TORCH screen), drug intake, radiation, diabetes, hypothyroidism
- Family history of deafness (first and second-degree relatives)

Birth History

- Prematurity, birth weight, APGAR scores
- Birth asphyxia, prolonged jaundice, NICU admission
- Ototoxic drug use in neonatal period

Developmental History

- Age at onset of hearing concern
- Speech milestones — age of babbling, first words, sentences
- School performance, attention, behavioral issues

Family History

- Consanguinity (autosomal recessive deafness risk)
- Affected siblings or parents
- Syndromic features in family

5.2 Examination

General Examination — Syndromic Features

- Eyes: heterochromia (Waardenburg), retinal pigmentation (Usher)
- Skin: white forelock (Waardenburg), hypopigmentation
- Neck: goitre (Pendred), branchial sinuses (Branchio-oto-renal syndrome)
- Face: Treacher Collins facies, CHARGE anomalies
- Hands/feet: syndactyly, polydactyly
- Renal: signs of nephropathy (Alport syndrome)

Otoscopic Examination

- External canal: atresia, stenosis, wax, foreign body
- Tympanic membrane: perforation, retraction, fluid (OME — amber TM, fluid level)
- Middle ear: cholesteatoma, granulations

Neurological Examination

- Cranial nerve assessment — facial nerve, balance
- Developmental assessment: gross motor, fine motor, social
- Vestibular assessment: balance, gait

SECTION 6 | Screening of Childhood Deafness

6.1 Universal Newborn Hearing Screening (UNHS)

- Recommended for ALL newborns before hospital discharge
- Gold standard screening tool: Otoacoustic Emissions (OAE) ± Automated ABR
- Follows the 1-3-6 Rule (JCIH 2007 and 2019 guidelines)

6.2 JCIH High-Risk Factors for Hearing Loss

Joint Committee on Infant Hearing (JCIH) High-Risk Criteria:

- Family history of permanent childhood SNHL
- NICU stay >5 days OR any of the following: ECMO, assisted ventilation, ototoxic drugs, hyperbilirubinemia
- In utero infections (TORCH)
- Craniofacial anomalies including EAC and pinna anomalies
- Physical findings associated with syndromes (Waardenburg, Usher, etc.)
- Neurodegenerative disorders (Hunter syndrome, Charcot-Marie-Tooth)
- Postnatal infections: bacterial meningitis, encephalitis
- Head trauma

- Parental concern regarding hearing/speech/language development

HIGH-YIELD: JCIH high-risk factors: NICU >5 days is THE most important perinatal risk factor for hearing screening.

6.3 Neonatal Screening Protocol

Stepwise Neonatal Hearing Screening Flowchart

All newborns: OAE screening before discharge

?

Pass ? Normal; follow-up if risk factors develop

?

Fail ? Repeat OAE in 2–4 weeks (avoid false positives from vernix/fluid)

?

Fail again ? Automated ABR (AABR)

?

Fail AABR ? Comprehensive audiological evaluation by 3 months

?

Confirmed loss ? Intervention (hearing aid/CI) by 6 months

6.4 OAE vs Automated ABR

Feature	OAE	Automated ABR (AABR)

Detects	Outer hair cell function	Brainstem auditory pathway
Time	~1–2 minutes	~15–20 minutes
Sensitivity	~80%	~99.7%
False positive	Higher (vernix, fluid, noise)	Lower
Auditory neuropathy	Normal OAE; ABR absent	Detects ANSD
Use	1st line screening	High-risk infants; 2nd line after fail

HIGH-YIELD: Auditory Neuropathy Spectrum Disorder (ANSD): OAE PRESENT but ABR ABSENT/abnormal — hallmark finding. Caused by kernicterus, prematurity, cochlear nerve deficiency.

SECTION 7 | Hearing Assessment in Children

7.1 Behavioral Audiological Tests

Test	Age Group	Principle	Notes
Behavioral Observation Audiometry (BOA)	0–6 months	Observer notes reflexive behavioral response to calibrated sounds	Subjective; not reliable for threshold

Visual Reinforcement Audiometry (VRA)	6 months–2.5 years	Child conditioned to turn head; rewarded with visual stimulus	Gold standard 6 months–2 years
Conditioned Orientation Reflex (COR)	6 months–2 years	Similar to VRA; loudspeaker-based	Free-field; bilateral average
Play Audiometry	2–5 years	Child performs play task in response to sound	Standard test 2.5–5 years
Pure Tone Audiometry (PTA)	>5 years	Child responds (button/hand raise) to pure tone frequencies	Standard adult-type test
Speech Audiometry	>5 years (cooperative)	Speech reception threshold + word recognition score	Functional hearing assessment

COMMON MCQ: VRA is the gold standard behavioral test for 6 months to 2 years. Play audiometry for 2.5–5 years. PTA >5 years.

7.2 Objective Audiological Tests

Otoacoustic Emissions (OAE)

- Generated by outer hair cells of cochlea
- Types:
  - TEOAE (Transient Evoked OAE): clicks ? response from 1–4 kHz range; used in neonatal screening
  - DPOAE (Distortion Product OAE): two simultaneous tones ? used for ototoxicity monitoring and more frequency-specific data
- OAE absent when hearing loss >25–30 dB HL
- OAE normal in auditory neuropathy (ANSD)

BERA / ABR (Brainstem Evoked Response Audiometry)

- Objective, no patient cooperation needed
- Measures neural responses along auditory pathway to click/tone-burst stimuli
- 5 waves (I–V): Wave V is most reliable threshold estimator
- Wave I: cochlear nerve; Wave III: superior olivary complex; Wave V: inferior colliculus
- Normal wave V threshold: ~10–15 dB nHL
- Used for: threshold estimation, retrocochlear lesions, neonatal screening, ANSD diagnosis, cochlear implant candidacy

HIGH-YIELD: ABR Wave V latency prolonged in retrocochlear lesions (acoustic neuroma). Absent Wave V in ANSD with present OAE = classic ANSD pattern.

ASSR — Auditory Steady State Response

- Frequency-specific objective threshold measurement
- More accurate than ABR for estimating hearing thresholds at specific frequencies
- Used for hearing aid fitting prescription in infants
- Excellent for severe-profound HL where ABR cannot estimate threshold

Tympanometry

- Measures middle ear pressure and compliance

Type	Compliance	Pressure	Interpretation
Type A	Normal	Normal	Normal middle ear
Type As	Reduced	Normal	Otosclerosis, tympanosclerosis
Type Ad	Increased	Normal	Ossicular discontinuity, flaccid T

Type B	Flat	Cannot determine	Otitis media with effusion (OME)
Type C	Normal/reduced	Negative	Eustachian tube dysfunction

COMMON MCQ: Type B (flat) tympanogram = most common in children with OME (glue ear).

7.3 Radiological Evaluation

- HRCT Temporal Bone: best for bony anatomy — canal atresia, middle ear, ossicles, Mondini, large vestibular aqueduct, cochlear ossification after meningitis
- MRI Internal Auditory Canal: best for cochlear nerve, cochlear fluid, central auditory pathway, cochlear nerve deficiency
- MRI Brainstem: auditory neuropathy, central causes

HIGH-YIELD: Pre-CI workup: HRCT temporal bone (bony anatomy) + MRI IAC (nerve presence). Both mandatory before cochlear implant surgery.

COMMON MCQ: Cochlear ossification after meningitis — detected on HRCT (white out of cochlea). Emergency indication for early cochlear implant before complete ossification.

SECTION 8 | Management

8.1 Medical Management

- Treat reversible causes: antibiotics for meningitis, antimalarials for malaria-related HL
- Manage otitis media with effusion: nasal decongestants, autoinflation, adenoidectomy + grommets
- Steroids for sudden SNHL (>2 years): systemic or intratympanic
- Prevent ototoxicity: monitor drug levels, use lowest effective dose, prefer alternatives
- Vaccination: see Section 10

8.2 Rehabilitation

Early Intervention

- Begin as soon as hearing loss confirmed; by 6 months of age
- Parent-centered program: parent is primary facilitator
- Home-based and clinic-based approaches

Auditory Verbal Therapy (AVT)

- Uses residual hearing maximally with hearing aids/CI
- Emphasizes listening and spoken language; no sign language
- Aims for mainstream schooling
- Best outcomes when started early (<2 years)

Lip Reading (Speech Reading)

- Uses visual cues from speaker's lip and facial movements
- Supplement to hearing aids; never a standalone substitute

Speech Therapy

- Trained SLP works on articulation, voice, language skills
- Essential component of CI rehabilitation

Family Counseling

- Explain diagnosis, prognosis, and realistic expectations
- Genetic counseling if hereditary etiology
- Support groups

Educational Rehabilitation

Option	Description	Suitable For
Special School (Oral)	Spoken language focused; uses AVT	Mild-moderate HL; good hearing aid
Special School (Sign Language)	Sign language as primary communication	Profound HL; poor CI candidates
Resource Room	Partial inclusion with resource teacher support	Moderate HL with hearing aids
Mainstream with FM System	Regular school with FM hearing loop	Post-CI, mild-moderate HL

8.3 Hearing Amplification

Hearing Aids

- First-line for all types/degrees of hearing loss
- Digital BTE (behind-the-ear) preferred for children
- Custom earmolds required; change as child grows
- Effective for mild-severe HL; limited benefit in profound HL
- Hearing aid trial minimum 3–6 months before CI candidacy

Bone Conduction Devices

- Indicated: CHL/mixed HL unsuitable for conventional hearing aids (atresia, chronic discharge)
- BAHA (Bone-Anchored Hearing Aid):
    - Implanted titanium screw in skull bone
    - Bypasses external and middle ear; transmits sound through bone to cochlea
    - Osseointegration needed (>5 years for implantation); use softband before 5 years

SECTION 9 | Cochlear Implant

9.1 Principles & Components

How It Works

Cochlear Implant Signal Processing Pathway

Microphone (external): picks up sound

?

Speech processor (external): converts sound to digital code

?

Transmitter coil (external): sends signal across skin

?

Receiver-stimulator (internal, implanted): receives signal

?

Electrode array (in cochlea): stimulates spiral ganglion neurons

?

Auditory nerve ? brain ? sound perception

Components

- External: microphone, speech processor, transmitter/headpiece
- Internal (implanted): receiver-stimulator, electrode array
- Electrode array: ~22 electrodes; placed in scala tympani of cochlea
- Different electrodes stimulate different frequency regions (tonotopic)

9.2 Indications

- Bilateral profound SNHL (>90 dB HL) — not benefiting from hearing aids after 3–6 months trial
- Bilateral severe SNHL (70–90 dB HL) with poor hearing aid benefit in children
- Single-sided deafness (SSD) — selected adult cases
- Post-meningitis cochlear ossification — urgent/early CI before complete ossification
- Age: optimal window 12 months to 3 years (prelingual); can be done from 6–12 months in selected cases

HIGH-YIELD: Optimal CI age: 12 months to 3 years. Earlier = better outcomes. Pre-lingual implantation before 2 years gives near-normal speech outcomes.

9.3 Contraindications

- Cochlear nerve aplasia (absent cochlear nerve on MRI) — absolute contraindication
- Michel aplasia (complete absence of inner ear)
- Severe developmental delay with inability to benefit from auditory rehabilitation
- Active middle ear infection — relative; treat first
- Unrealistic parental expectations without commitment to rehabilitation

COMMON MCQ: Cochlear nerve aplasia on MRI = absolute contraindication to cochlear implant. Michel aplasia = bony contraindication.

9.4 Candidate Selection

Audiological Criteria

- PTA average >70–90 dB HL bilaterally
- Minimal speech perception scores with optimally fit hearing aids (<30–50% on open-set sentence tests in adults)
- Failed hearing aid benefit after adequate trial

Radiological Criteria

- HRCT: patent cochlea, no complete ossification, patent cochlear turns
- MRI IAC: cochlear nerve present (mandatory)

Psychological & Rehabilitation Assessment

- Parental motivation and commitment to rehabilitation program
- Realistic expectations
- Absence of severe learning disability

9.5 Surgical Aspects

Approach

- Postauricular incision ? cortical mastoidectomy ? posterior tympanotomy (facial recess approach)
- Cochleostomy or extended round window approach to enter scala tympani
- Electrode array inserted via soft surgery technique to preserve residual hearing

Soft Surgery Technique

- Preserves cochlear microstructure
- Allows hearing preservation for electric-acoustic stimulation (EAS) in residual low-frequency hearing
- Slow, atraumatic electrode insertion

Neural Response Telemetry (NRT)

- Intraoperative testing of electrode-nerve interface
- Confirms cochlear nerve responses
- Guides programming/mapping

9.6 Postoperative Rehabilitation

- Device activation (switch-on): 4–6 weeks post-surgery
- Mapping/Programming: programming of each electrode's threshold (T-level) and comfortable level (C-level)
- Repeated mapping sessions over months to optimize performance
- Auditory rehabilitation: structured listening program
- Speech therapy: intensive speech and language work
- Progress monitoring: speech perception tests, language assessments

CLINICAL PEARL: CI outcomes are maximized when: early implantation (<2 years), strong parental involvement, intensive AVT, and mainstream educational integration are combined.

9.7 Bilateral Cochlear Implantation

Feature	Sequential	Simultaneous
Timing	Two separate surgeries, interval >6 months	Both ears in single surgery
Advantage	2nd ear if 1st fails; hearing assessment after 1st	Single anesthetic; shorter interval to bina lower total cost
Disadvantage	Longer period of unilateral hearing; 2 anesthetics	Higher surgical risk; 2nd ear cannot bene experience
Preferred in	Financial/system constraints; older children	Young infants; when simultaneous benef

CLINICAL PEARL: Bilateral CI provides binaural hearing advantages: sound localization, improved speech in noise, auditory streaming.

9.8 Complications of Cochlear Implant

Complication	Details
Device failure (hard failure)	Internal device malfunction; requires re-implantation
Facial nerve injury	Rare (<1%); most at second genu or mastoid segment
CSF leak / Perilymph gusher	Risk with Mondini deformity; manage with packing
Wound infection	Superficial; rare serious deep infection
Meningitis	Post-CI meningitis risk (pneumococcal); vaccination essential
Flap necrosis	Skin flap breakdown over implant
Electrode migration/extrusion	Device displacement
Cochlear ossification	Post-meningitis; may prevent full insertion
Tinnitus/vertigo	Usually temporary

HIGH-YIELD: Post-CI meningitis risk: caused by Streptococcus pneumoniae traveling via electrode. Mandatory vaccinations before CI: Pneumococcal + Meningococcal + Hib.

9.9 Vaccination Protocol for CI Candidates

- Pneumococcal vaccine (PCV13 + PPSV23) — most critical
- Haemophilus influenzae type b (Hib) vaccine
- Meningococcal vaccine
- All vaccines should be given at least 2 weeks before surgery if possible

SECTION 10 | Prognosis & Prevention

10.1 Consequences of Delayed Diagnosis

- Permanent language deprivation — cannot be fully reversed after critical period
- Auditory deprivation: central auditory pathway fails to mature
- Learning disability: academic failure
- Psychological sequelae: social isolation, low self-esteem, depression
- Poor CI outcomes if implantation delayed beyond 5–7 years

10.2 Prevention

Primary Prevention

- Universal immunization: MMR (measles, mumps, rubella), Hib, pneumococcal, meningococcal
- Antenatal TORCH screening and treatment
- Avoidance of ototoxic drugs during pregnancy
- Genetic counseling for high-risk couples (consanguinity, affected sibling)
- Safe noise exposure: education, hearing protection

Secondary Prevention

- Universal newborn hearing screening — 1-3-6 Rule
- Early hearing aid fitting
- Early speech therapy and educational placement

Tertiary Prevention

- Cochlear implantation for profound SNHL
- Auditory verbal therapy to maximize use of implant
- Mainstream educational integration

SECTION 11 | Master Comparison Tables

11.1 Conductive vs Sensorineural vs Mixed HL

Feature	CHL	SNHL	Mixed
Site of lesion	External/middle ear	Cochlea/auditory nerve	Both
Air conduction	Reduced	Reduced	Reduced
Bone conduction	Normal	Reduced	Reduced (less
Air-bone gap	Present (>10 dB)	Absent	Present
Rinne test	Negative (BC>AC)	Positive (AC>BC, both reduced)	May be negat

Weber lateralization	Lateralizes to worse ear	Lateralizes to better ear	Variable
Max severity	60 dB HL	120 dB HL (profound)	120 dB HL
Management	Often surgical/medical	Hearing aid / CI	Combined

11.2 OAE vs BERA (ABR)

Feature	OAE	BERA/ABR
Origin	Outer hair cells	Auditory pathway (CN VIII ? inferi
Reflects	Cochlear function	Neural synchrony along auditory pa
Cooperation	Not needed	Not needed (sedation in children)
Frequency specific	Partial (DPOAE better)	Poor (click ABR); good (tone burst
Threshold estimate	Indirect (present/absent)	Direct threshold estimation
ANSD pattern	Present (normal)	Absent / severely abnormal

Use	Screening	Diagnosis + threshold + retrocochle

11.3 Hearing Aid vs Cochlear Implant

Feature	Hearing Aid	Cochlear Implant
Mechanism	Amplifies residual hearing	Bypasses cochlea; directly stimulat
Surgery	Not required	Required
Degree of HL	Mild–severe (best); some profound	Severe–profound SNHL
Cochlea required	Must have residual hair cells	Spiral ganglion neurons sufficient
Reversible	Yes	No (device is permanent)
Best age to start	As early as possible	<2 years for best outcomes
Outcome	Good for mild-moderate HL	Near-normal speech for early bilate

11.4 Prelingual vs Postlingual Deafness

Feature	Prelingual	Postlingual
Onset	Before speech development (<2 years)	After speech development (>2 year
Language impact	Severe; no spontaneous speech	Speech retained initially; deteriorat
CI timing	Best <2 years	Good at any age; faster re-adaptatio
Prognosis	Depends on age of CI	Generally better
Example	Congenital SNHL	Meningitis at age 5; sudden SNHL

11.5 Key Genetic Syndromes — Quick Comparison

Syndrome	HL Type	Other Feature	Inheritance
Waardenburg	SNHL	White forelock, heterochromia	AD
Usher	SNHL (profound)	Retinitis pigmentosa	AR

Pendred	SNHL	Goitre + Mondini	AR
Alport	SNHL	Nephritis	X-linked
Jervell-Lange-Nielsen	SNHL (profound)	Long QT — sudden death	AR
Treacher Collins	CHL	Malar hypoplasia, micrognathia	AD
Branchio-oto-renal	Mixed	Branchial cysts, renal anomalies	AD

SECTION 12 | High-Yield Exam Pearls

12.1 Epidemiology & General

- 1-3-6 Rule: Screen by 1 month ? Diagnose by 3 months ? Intervene by 6 months
- Most common non-genetic congenital SNHL = CMV
- Most common genetic cause of non-syndromic SNHL = Connexin 26 (GJB2) mutation
- Most common cause of acquired SNHL in children = Bacterial meningitis
- Most severe inner ear malformation = Michel aplasia (CI contraindicated)
- Most common inner ear malformation = Scheibe dysplasia

12.2 Audiology

- VRA = gold standard behavioral test for 6 months–2 years
- ANSD: OAE present, ABR absent — caused by kernicterus, prematurity
- Type B flat tympanogram = OME (glue ear) in children
- ASSR: best objective frequency-specific test for infants; aids in hearing aid fitting

- Wave V of ABR = most reliable wave for threshold estimation
- Cisplatin: irreversible, dose-dependent SNHL at high frequencies first

12.3 Cochlear Implant

- Optimal CI age: <2 years for best speech outcomes
- CI contraindications: absent cochlear nerve (MRI), Michel aplasia
- Pre-CI mandatory vaccines: Pneumococcal + Hib + Meningococcal (at least 2 weeks before)
- Post-meningitis: urgent CI before cochlear ossification occurs
- Mondini deformity ? CI possible but risk of CSF gusher during surgery
- EVA (Enlarged Vestibular Aqueduct) ? progressive SNHL; avoid head trauma

12.4 Syndromes

- Jervell-Lange-Nielsen = SNHL + Long QT ? cardiac arrest; dangerous ? ECG mandatory
- Usher syndrome = SNHL + progressive blindness (retinitis pigmentosa)
- Pendred = SNHL + goitre + Mondini dysplasia + perchlorate discharge test positive
- Alport = SNHL + hematuria + nephritis + ocular anomalies; X-linked
- Waardenburg Type I: dystopia canthorum (wider spacing of medial canthi) — distinguishes from Type II

12.5 Embryology / Congenital

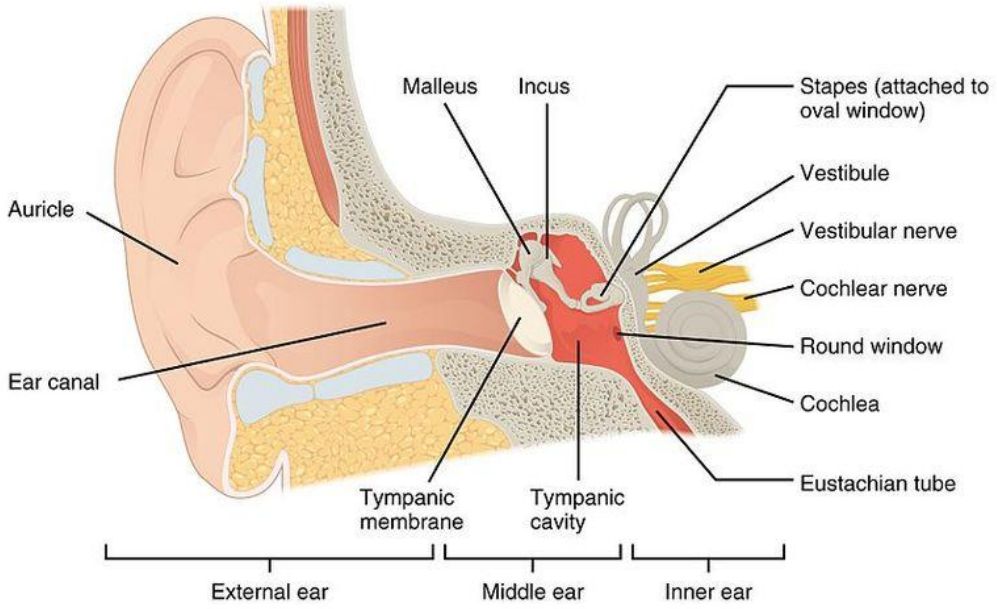
- 1555A>G mitochondrial mutation: aminoglycoside hypersensitivity ? profound SNHL even with single dose
- Rubella: 1st trimester = most dangerous; causes SNHL + congenital heart disease + cataracts
- Congenital X-linked deafness with POU3F4 mutation ? perilymphatic gusher during stapes surgery
- Cochlear nerve deficiency ? CI contraindicated; consider auditory brainstem implant (ABI)

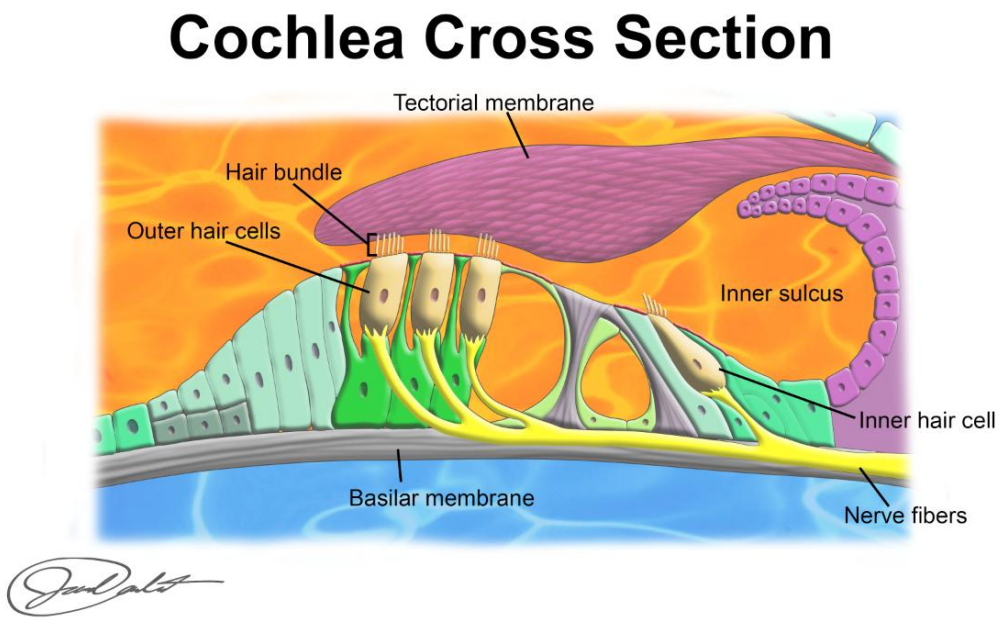
SECTION 13 | Important Diagrams & Figures

13.1 Anatomy Diagrams

Image 1

- Normal ear anatomy — external, middle, inner ear labeled cross-section
- Cochlear cross-section showing scala vestibuli, scala media, scala tympani, Organ of Corti, basilar membrane
- Organ of Corti: inner hair cells, outer hair cells, tectorial membrane, pillar cells





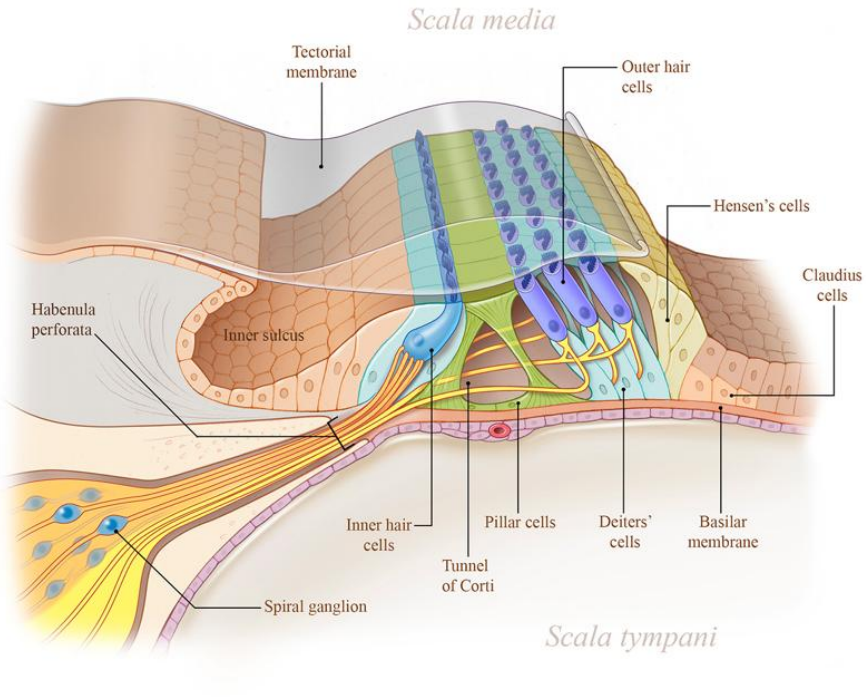
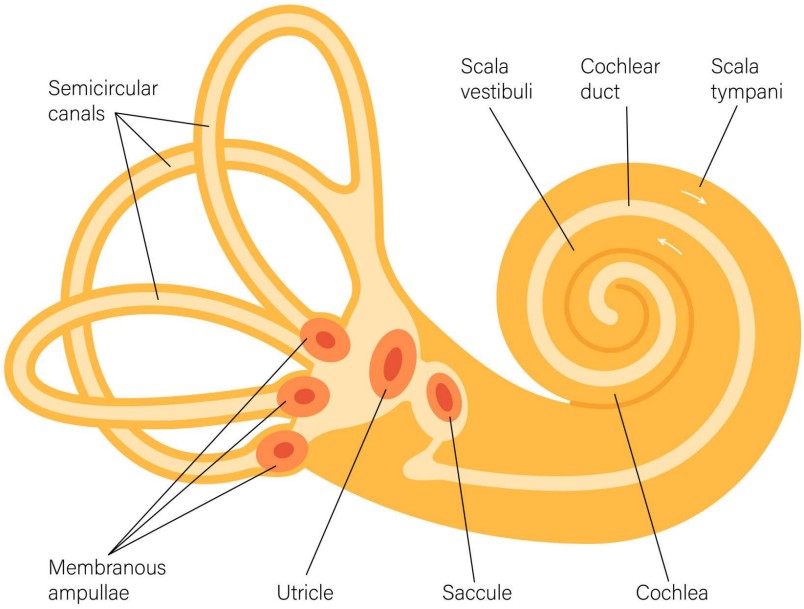
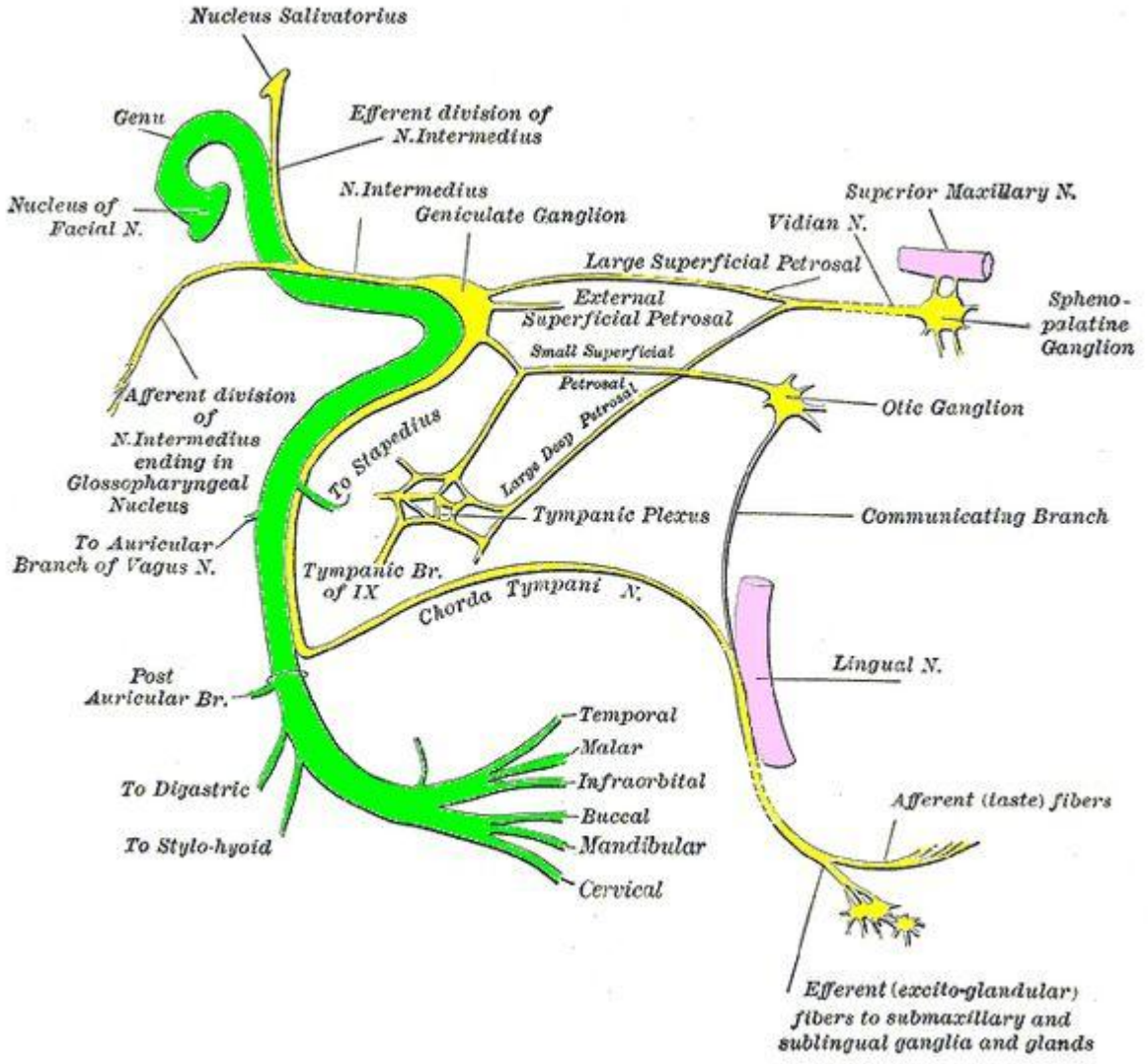


Image 2

- Vestibular apparatus: utricle, saccule, semicircular canals, crista ampullaris
- Facial nerve course in temporal bone — all segments

VESTIBULAR APPARATUS

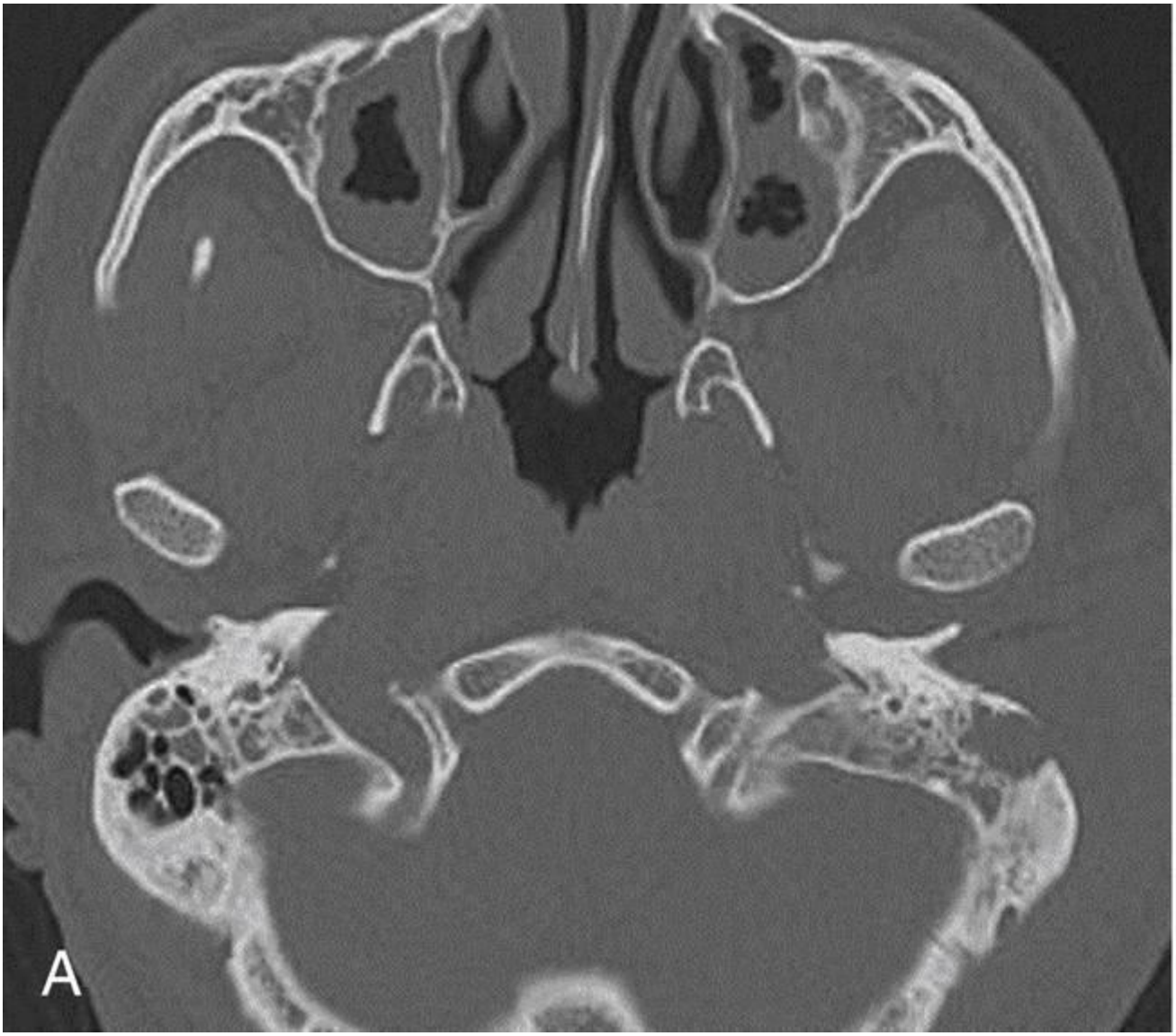


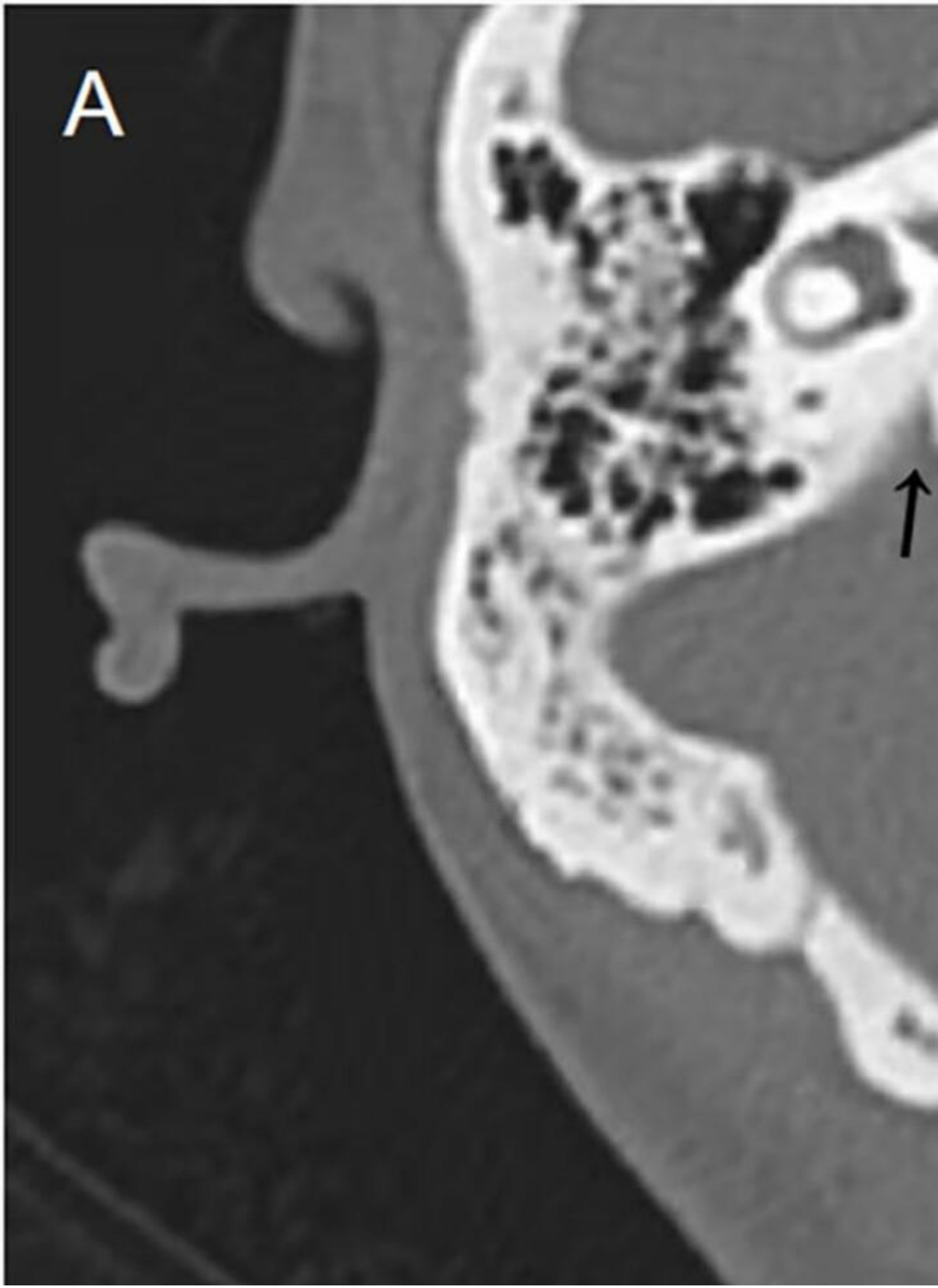


13.2 Pathology Diagrams

Image 3

- HRCT temporal bone: normal cochlea vs Mondini deformity vs common cavity
- HRCT temporal bone: enlarged vestibular aqueduct
- HRCT temporal bone: post-meningitis cochlear ossification





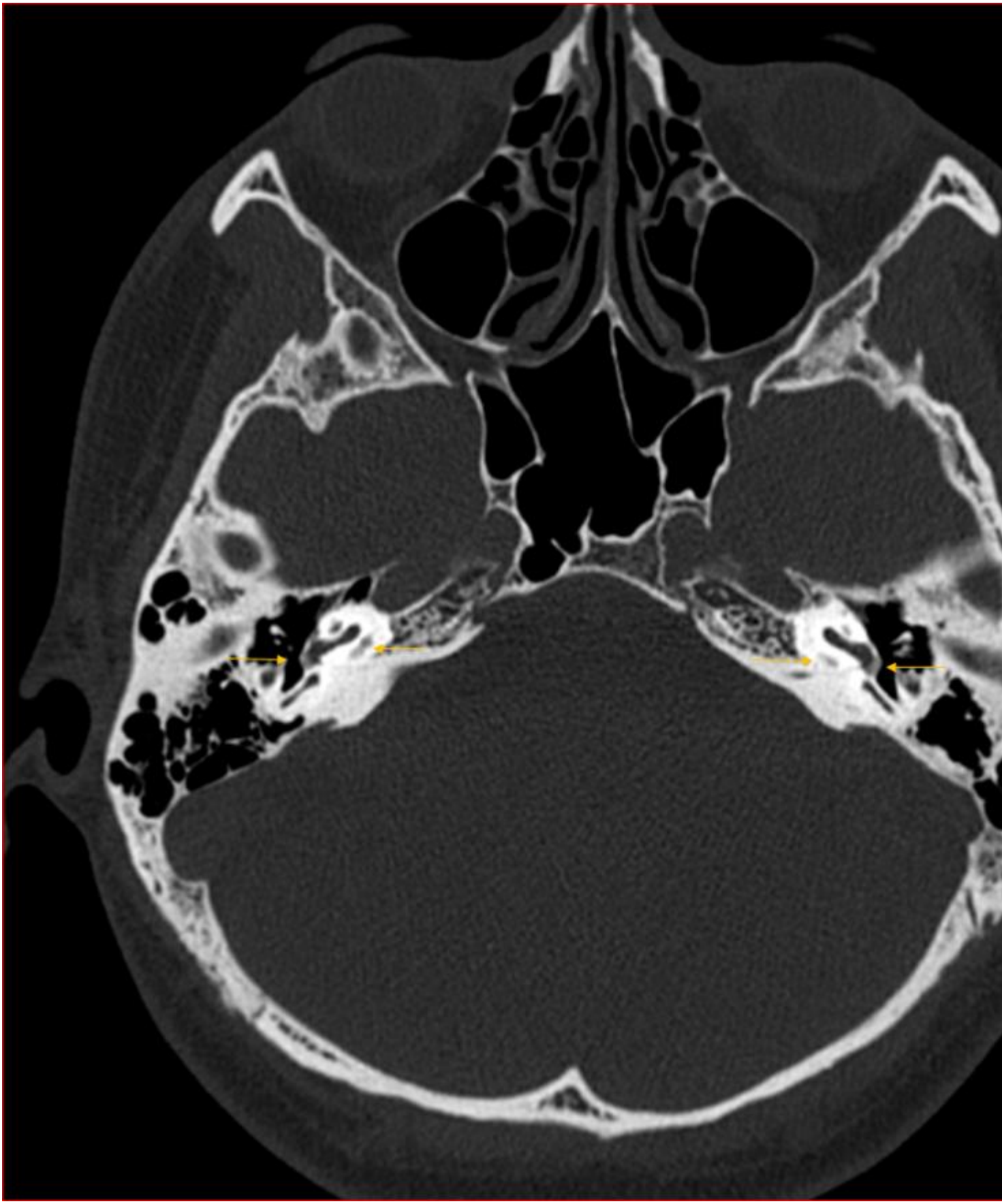
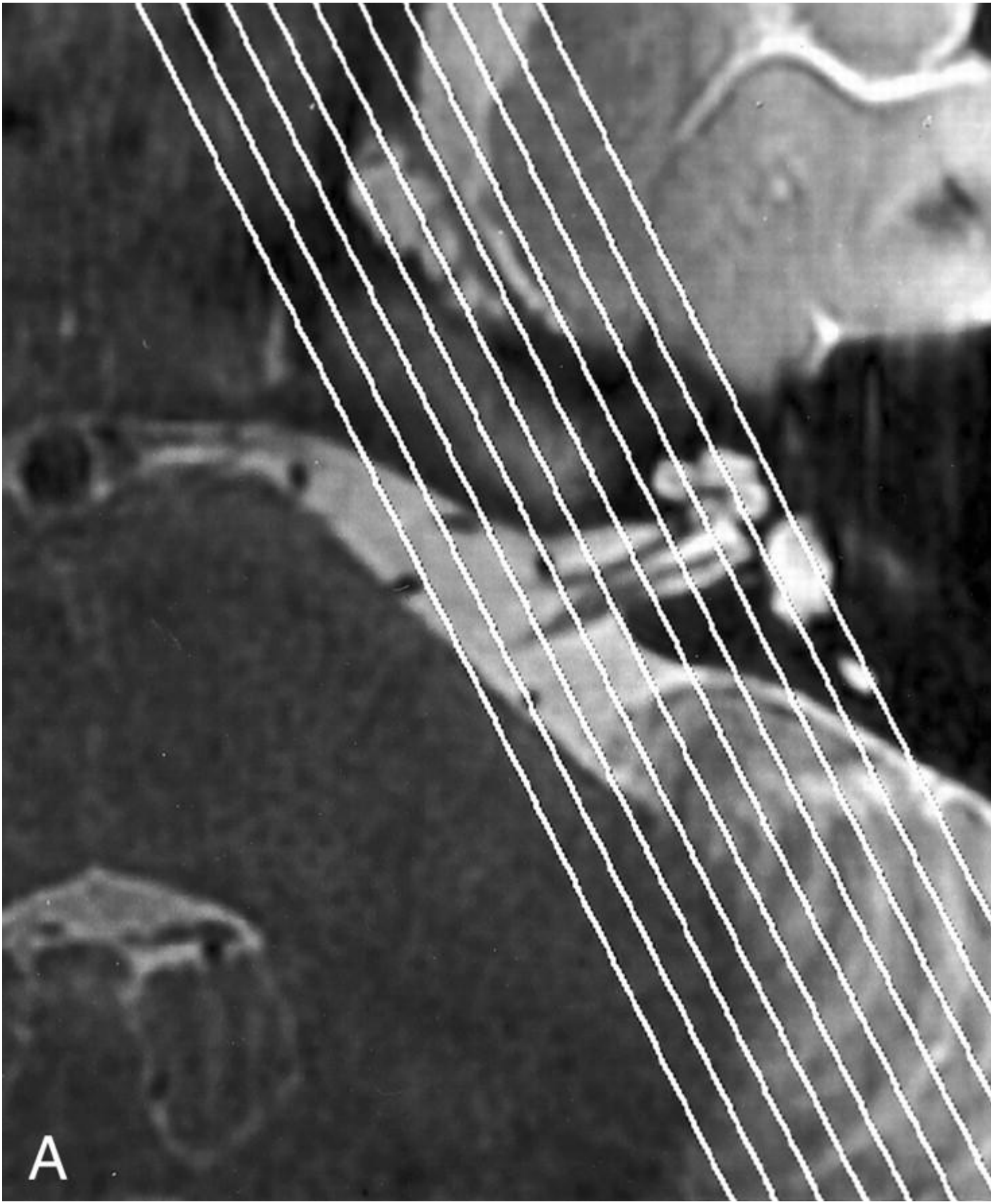


Image 4

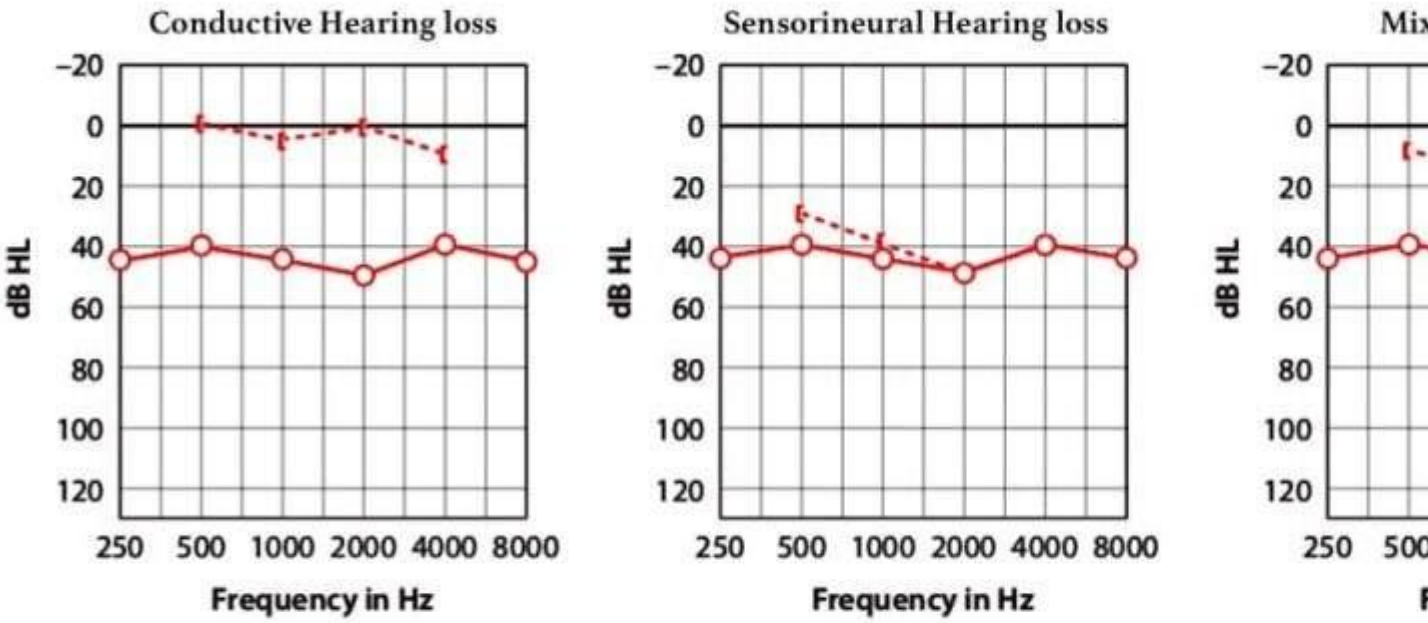
- MRI IAC: normal cochlear nerve vs cochlear nerve aplasia



13.3 Audiological Figures

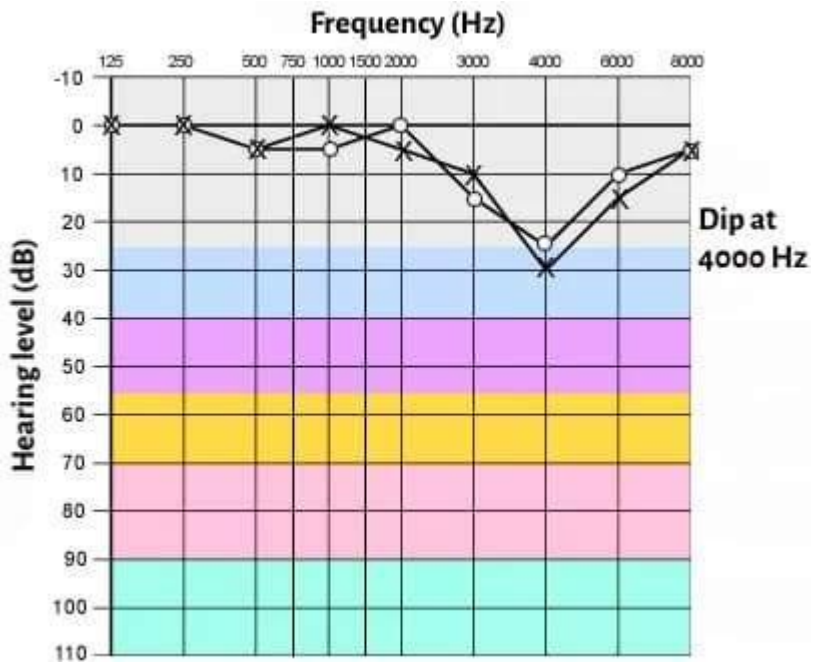
Image 5

- Pure tone audiogram: CHL, SNHL, mixed hearing loss
- Audiogram: high-frequency notch vs flat genetic SNHL
- Speech banana



Types of Hearing loss on PTA.

CHL - BC thresholds are normal, increased AC thresholds with significant AB
SNHL - Both AC, BC thresholds increased with no significant AB gap.
Mixed HL - Both AC, BC thresholds increased with significant AB gap.


Frequency (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
-10
0
10
20
30
40
50
60
70
80
90
100
110
Hearing level (dB)
Dip at
4000 Hz

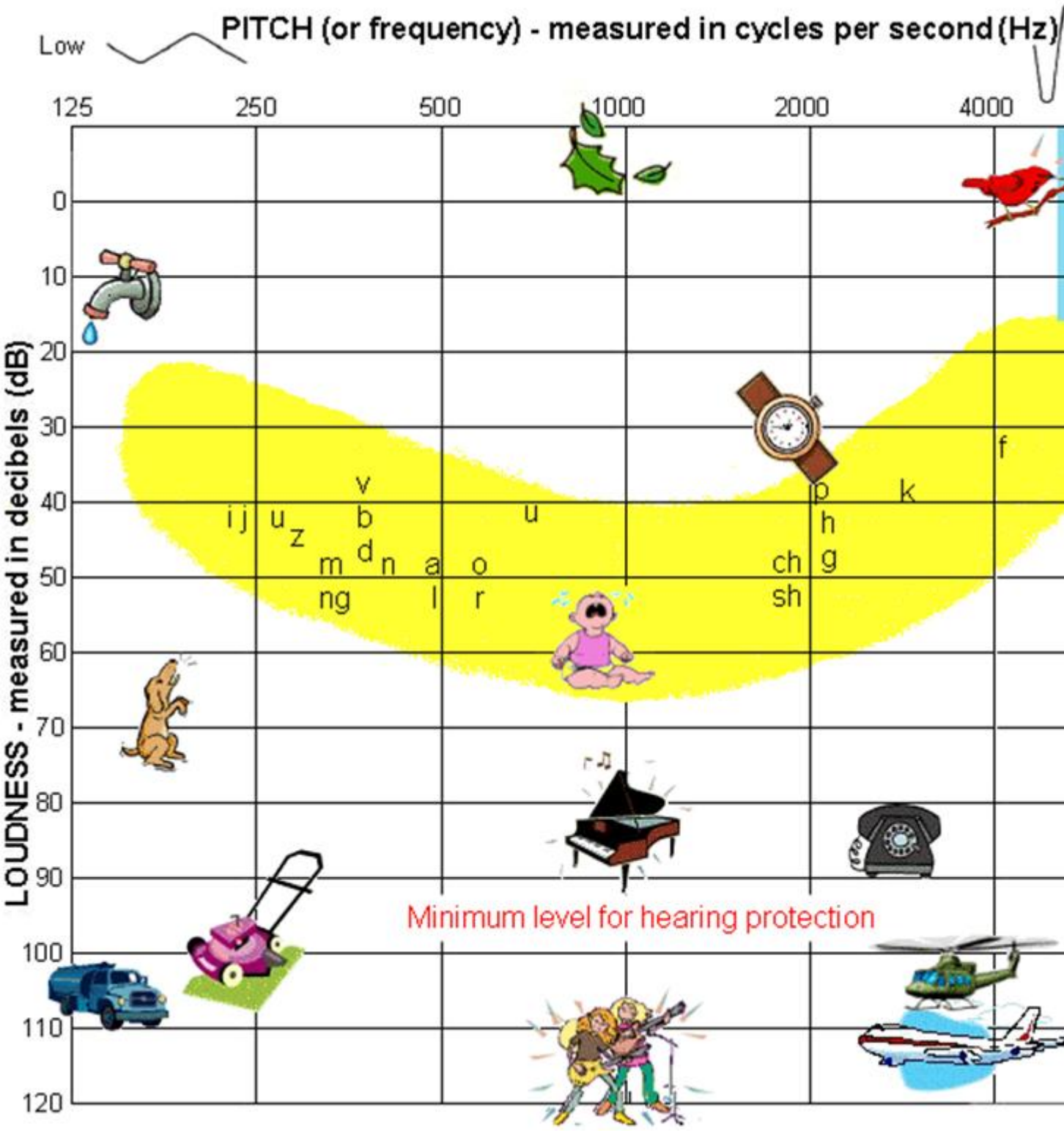
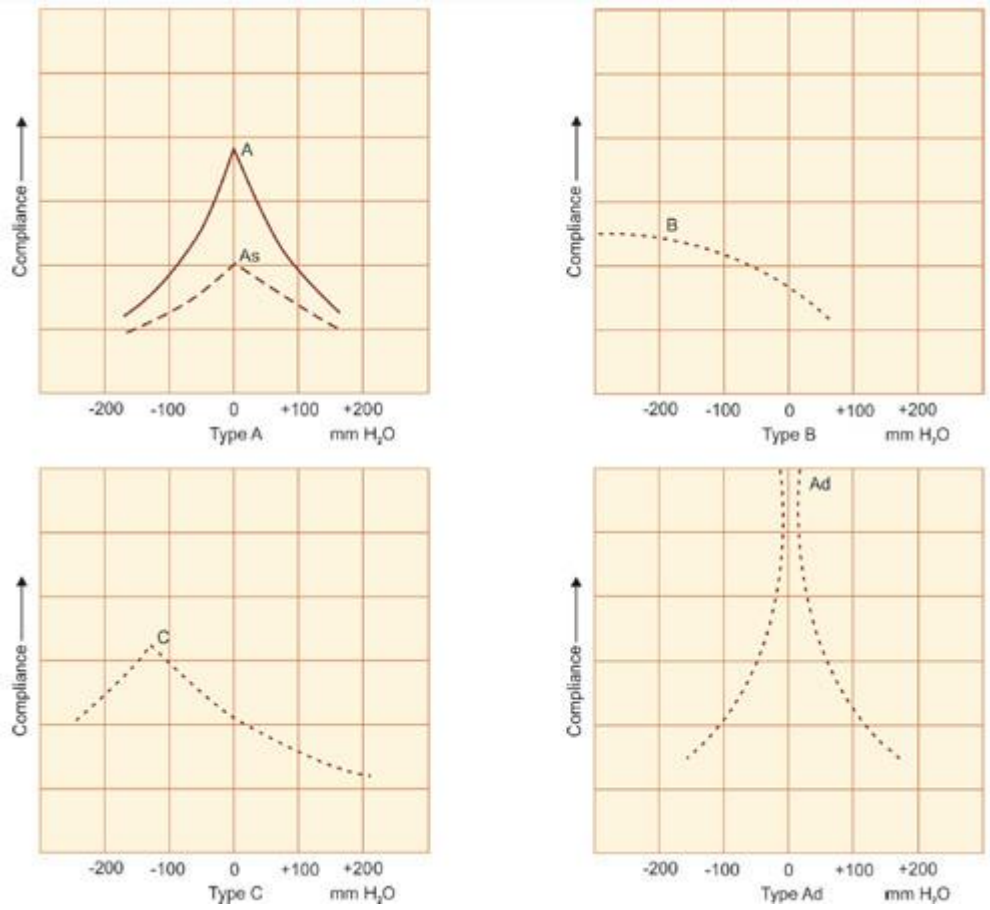


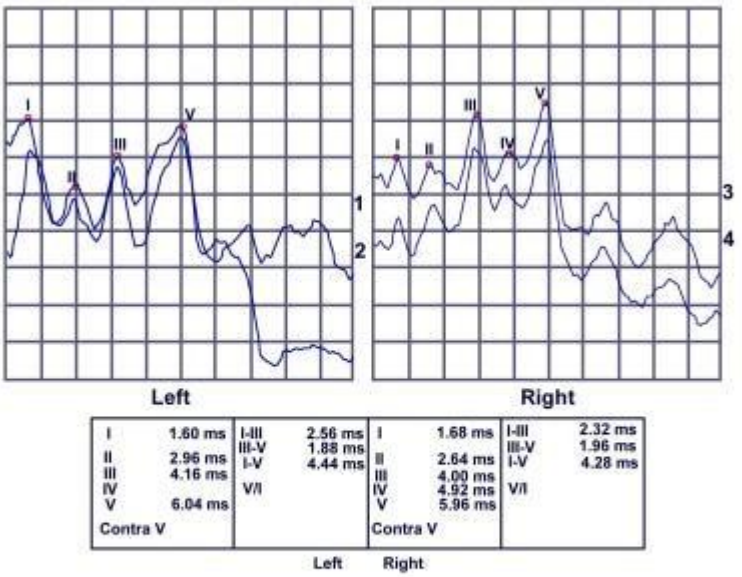
Image 6

- Tympanogram types: A, As, Ad, B, C

- BERA/ABR waveform: Waves I–V labeled

- OAE result: present vs absent waveform





Normal adult ABR waveform response. I-V absolute latencies and interpeak intervals (I-III, III-V, I-V) are within normal limits bilaterally. Interaural differences for the I-V interpeak intervals (1.16ms) and wave V absolute latencies (.08 ms) are within normal limits.

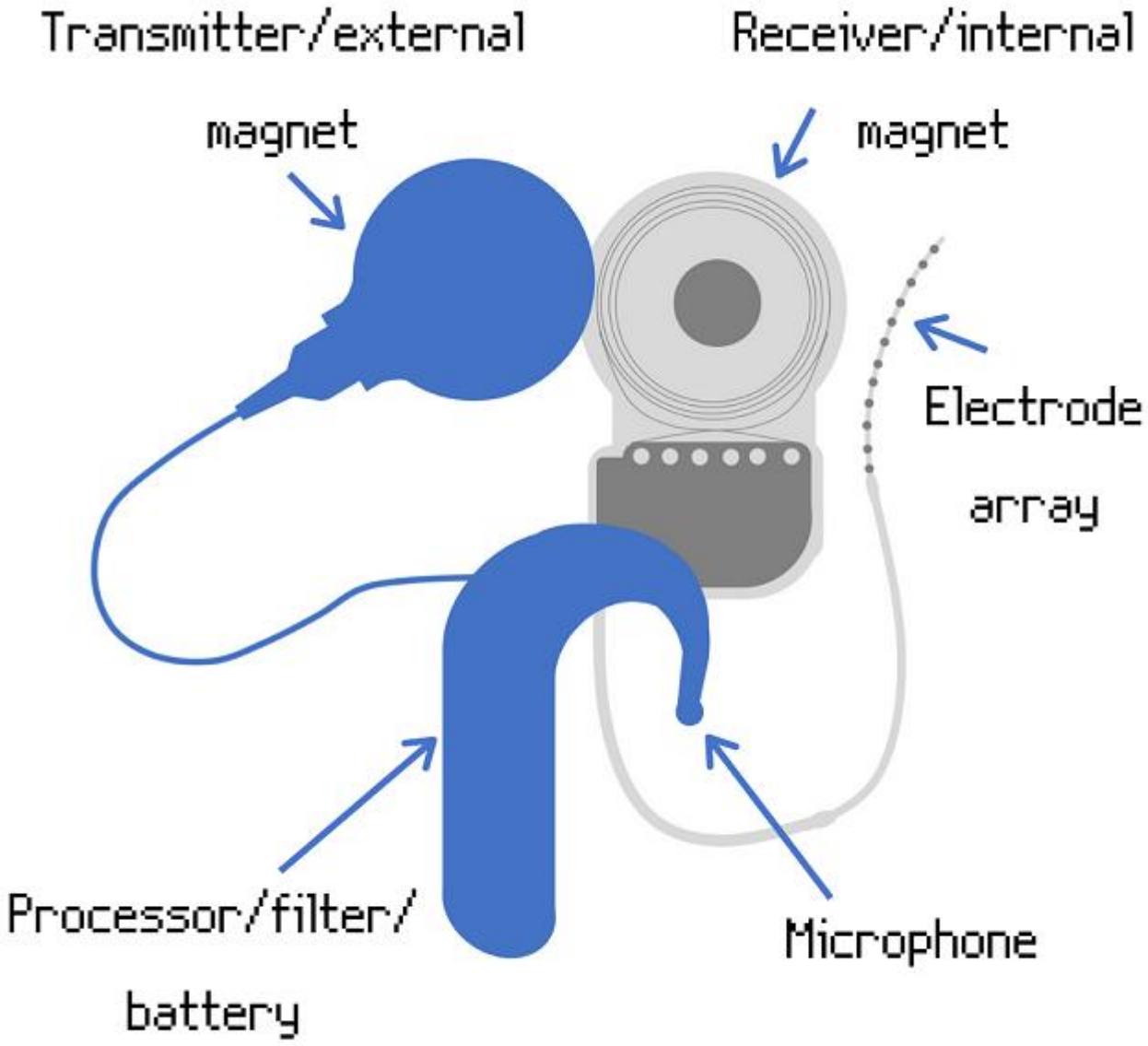
- Visual inspection, and preferably tympanometric measurement prior to OAEs recordings, will help determine if middle ear and external ear abnormalities might reduce or block acoustic transmission of OAEs from the cochlea to the microphone, and in the case of TEOAEs and DPOAEs, of the evoking sound to the cochlea.
- Reduction of ambient noise picked up by the microphone is achieved by a tight fit of the probe into the ear canal.
- Patient -generated sounds can be minimized by instructing the patient to be still and not to talk during testing.

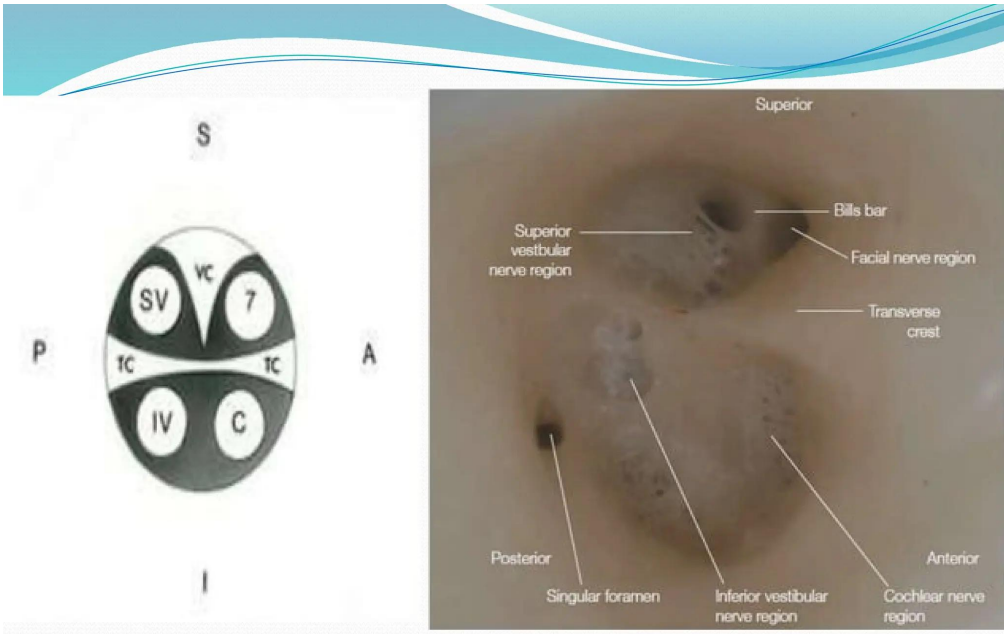
13.4 Surgical & Cochlear Implant Diagrams

Image 7

- Cochlear implant external and internal components
- Facial recess approach and posterior tympanotomy

- Electrode array position in scala tympani





Inner aspect of lateral end of IAC

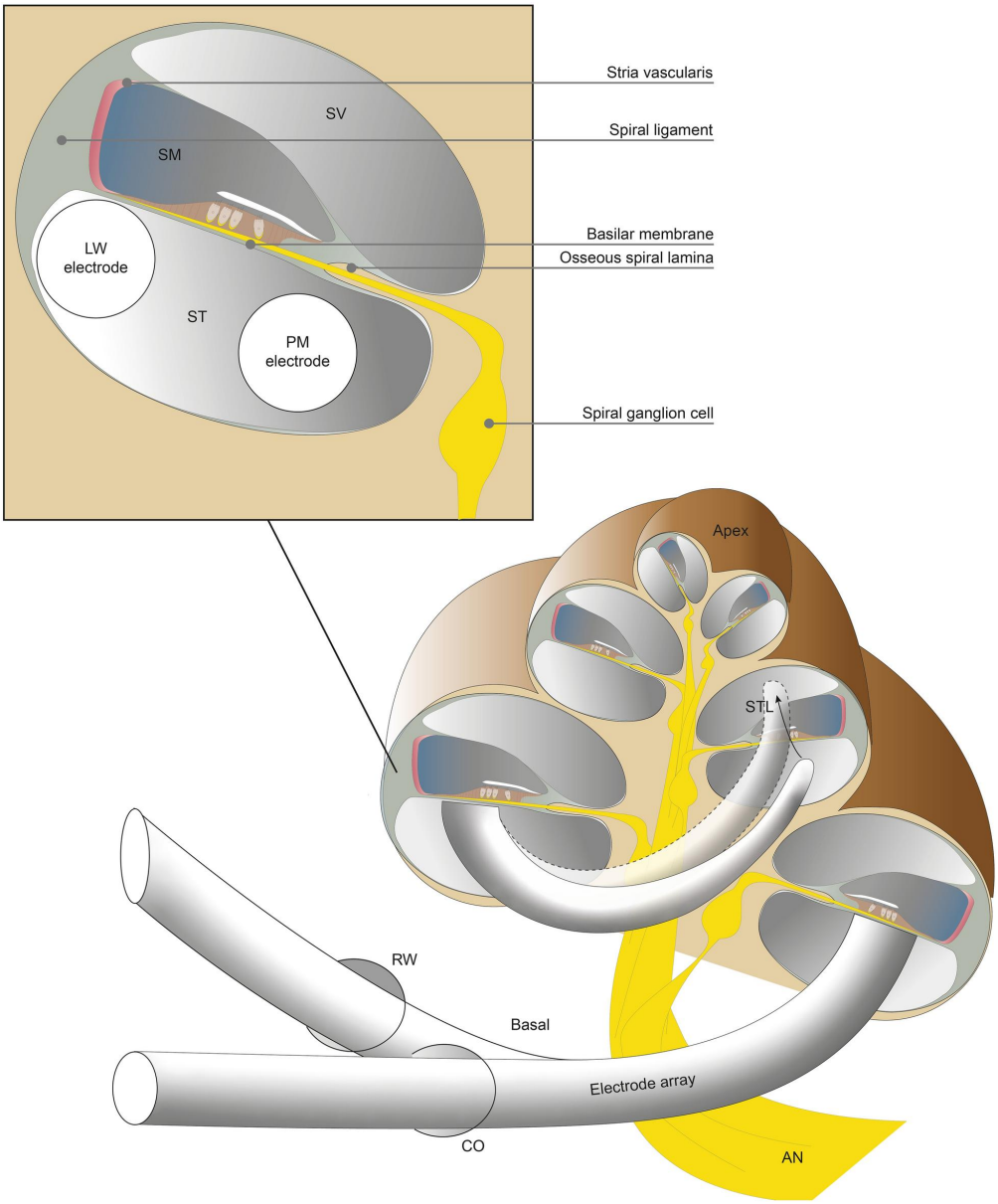
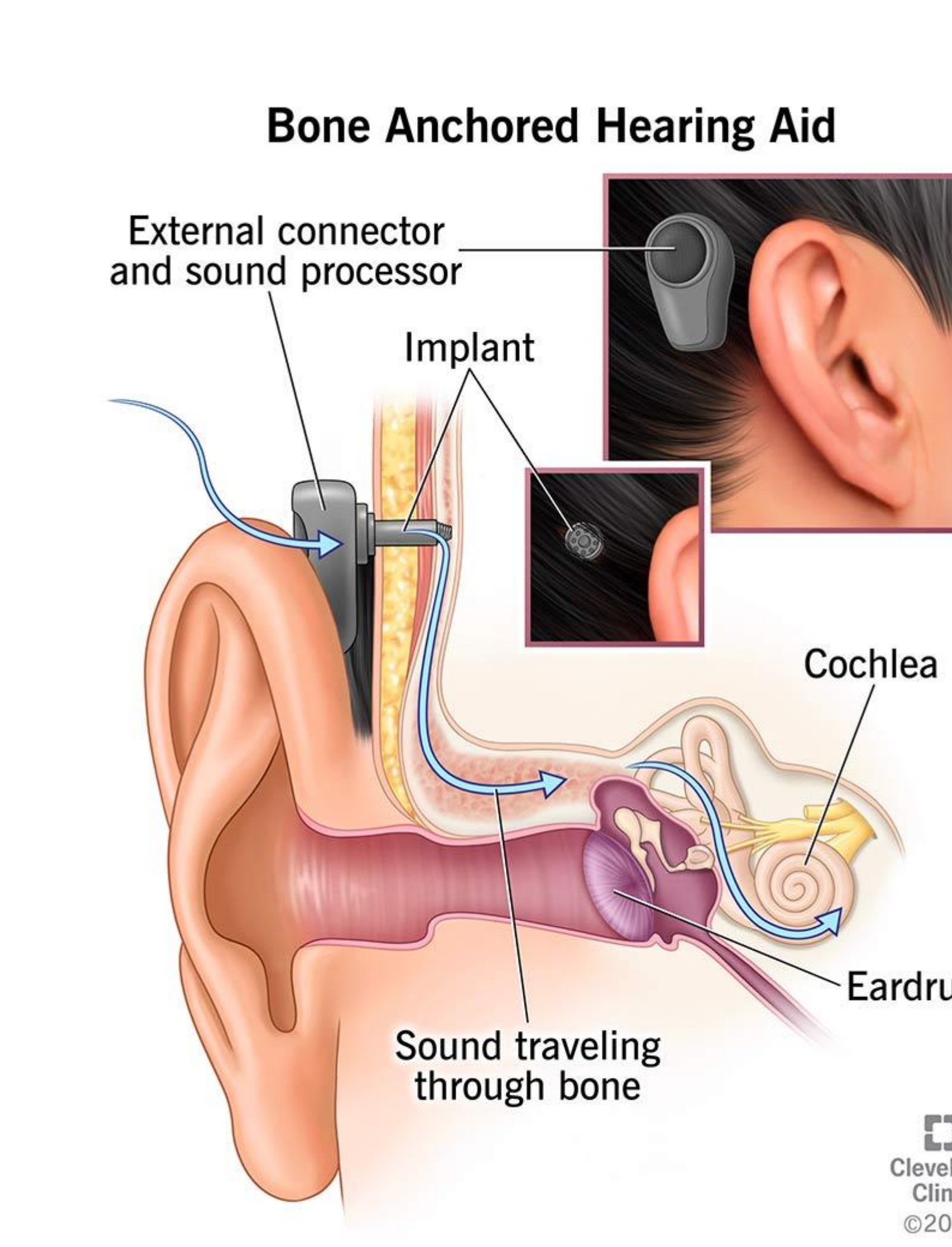
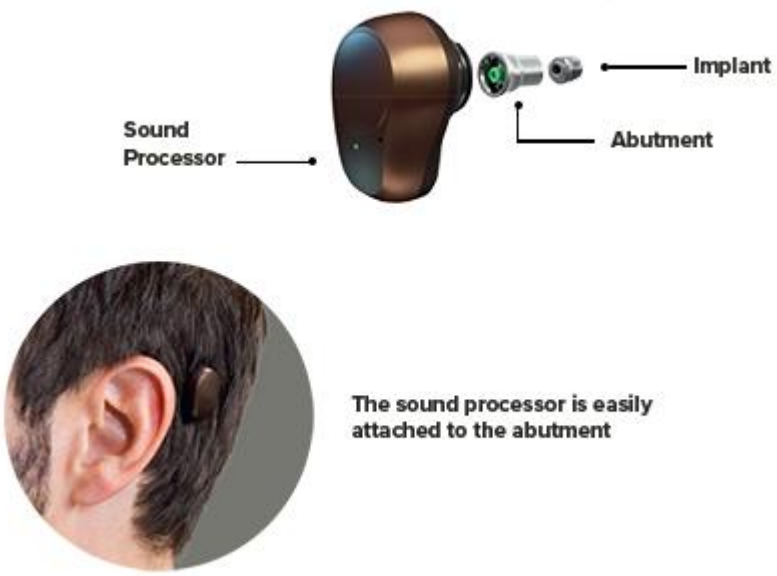


Image 8

- BAHA: bone-anchored hearing aid with titanium screw integration

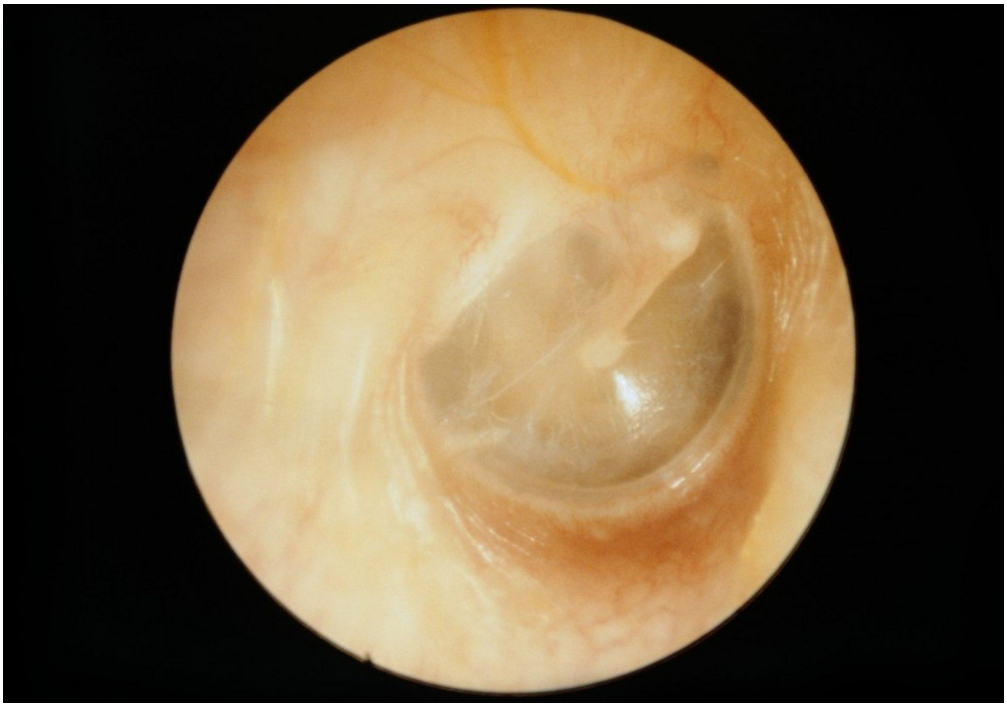

Bone Anchored Hearing Aid
External connector and sound processor
Implant
Cochlea
Eardru
Sound traveling through bone
Clevel
Clin
©20

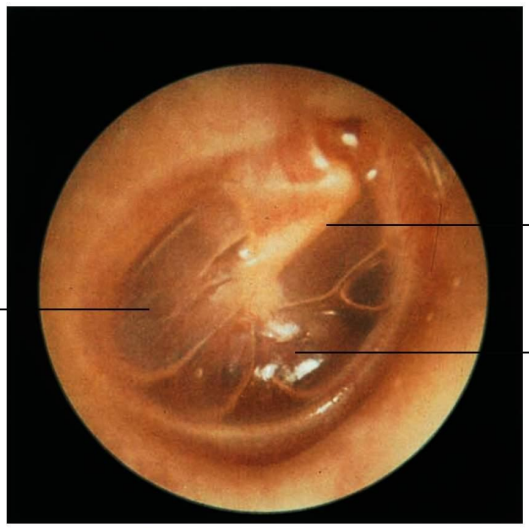


13.5 Clinical Photographs

Image 9

- Normal tympanic membrane
- OME glue ear
- Child undergoing play audiometry





Otitis Media with Effusion

Image 10

- Child wearing BTE hearing aid with earmold
- Cochlear implant processor and headpiece in child
- Waardenburg syndrome clinical features


Transmitting Coil
Ear Level
Speech Processor

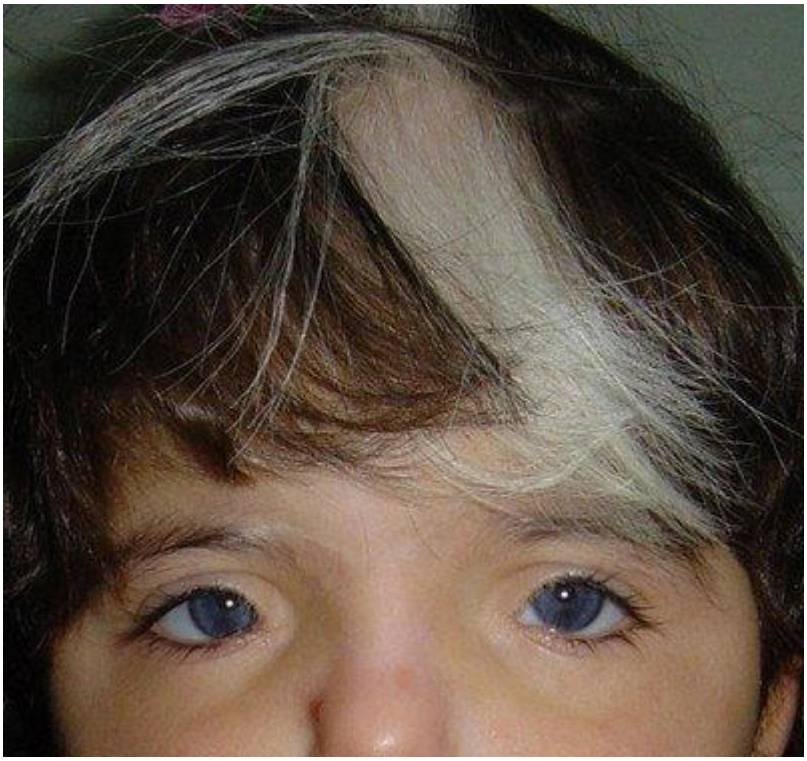


Image 11

- Treacher Collins syndrome: malar hypoplasia, micrognathia, ear anomalies

Characteristic clinical features of Treacher–Collins syndrome

Eyelids
- Antimongoloid obliquity of palpebral fissures
- Coloboma of lower eyelids
- Dystopia of lateral canthi
- Shortening of palpebral fissure
- Absence of eyelashes
- Notching of eyebrows and upper eyelids

Orbits
- Inferior portion of lateral wall is often absent
- Inferior migration of superolateral portion of frontal bone

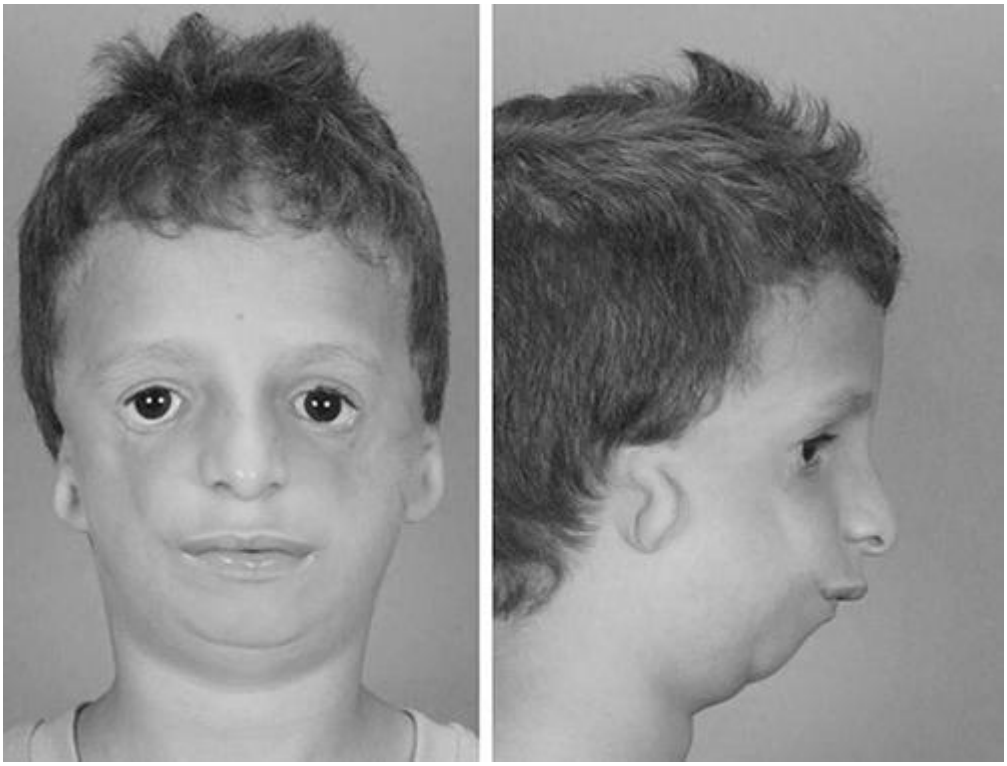
Malar bone
- Hypoplastic or absent
- Absence of zygomatic arch

Maxilla
- Narrow and underprojected
- High and narrow palate

Mandible
- Hypoplastic
- Vertical occlusal plane
- Class III malocclusion with anterior open bite
- Long and retruded chin

Nose
- Protruded with a broad base
- Flattened frontonasal angle
- Narrow pharynx

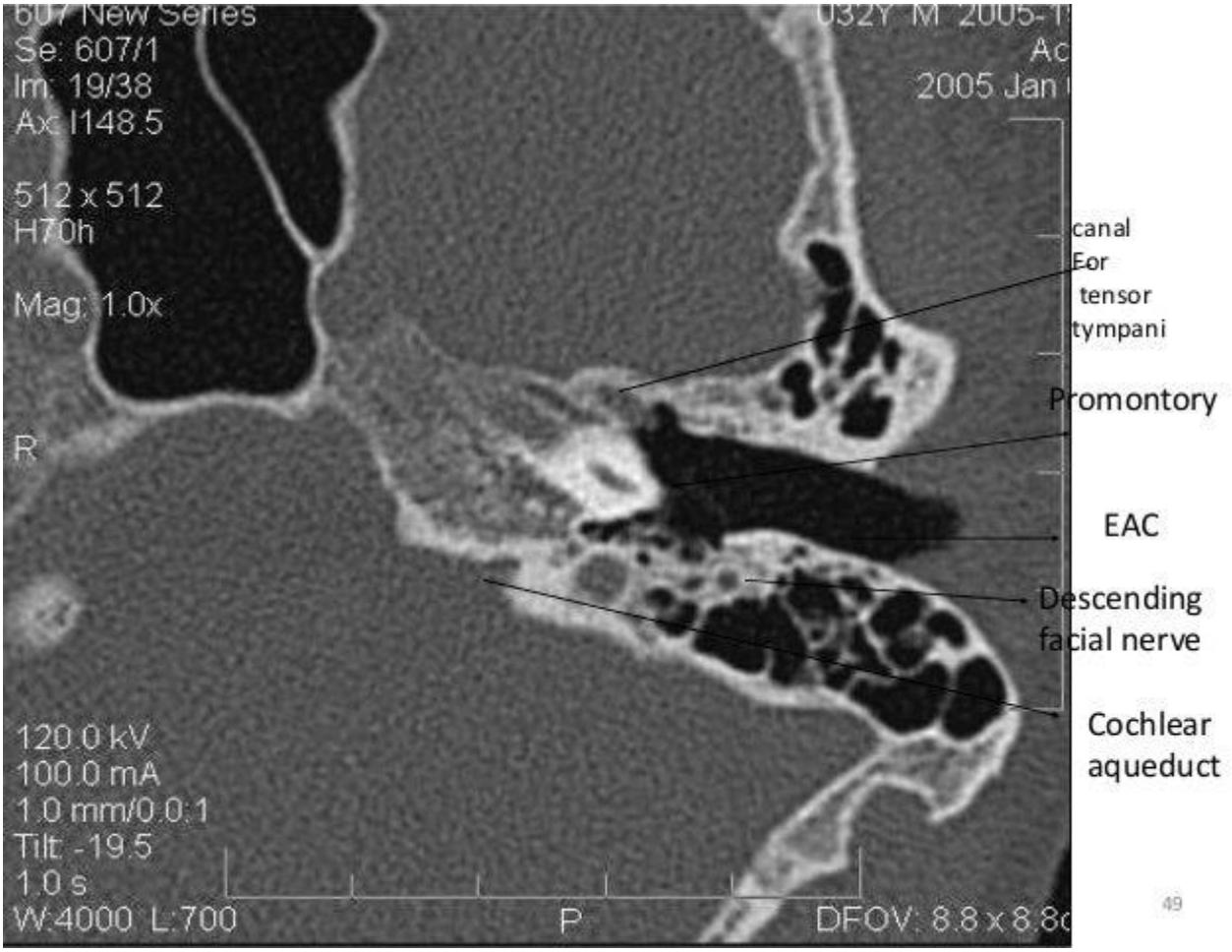
Others
- Microtia and ear deformities
- Absence of external auditory canal
- Abnormalities of middle ear
- Macrostomia
- Possible velopharyngeal insufficiency



13.6 Radiology Images

Image 12

- HRCT temporal bone: normal cochlea vs Mondini deformity
- HRCT temporal bone: large vestibular aqueduct coronal view
- HRCT temporal bone: labyrinthitis ossificans white cochlea



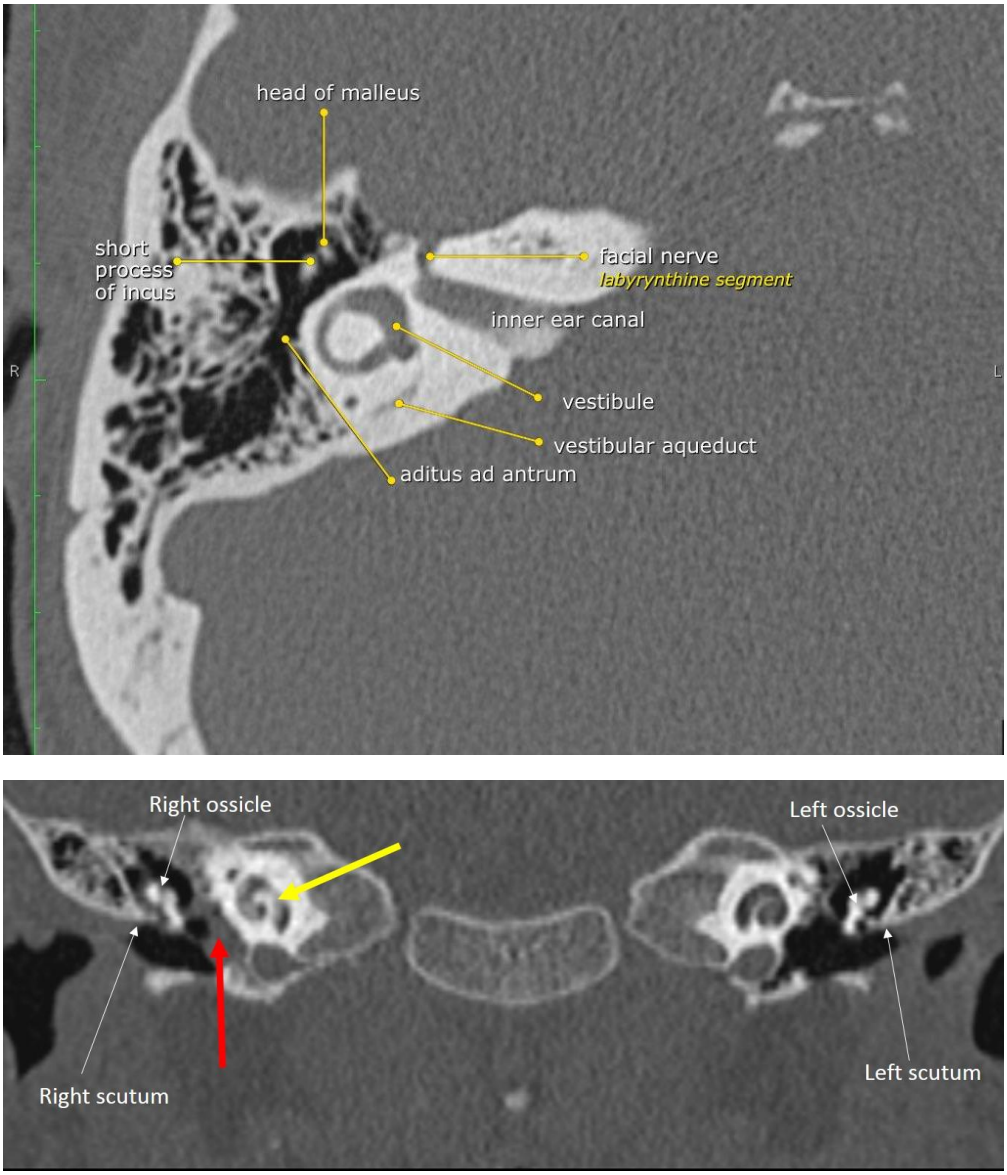
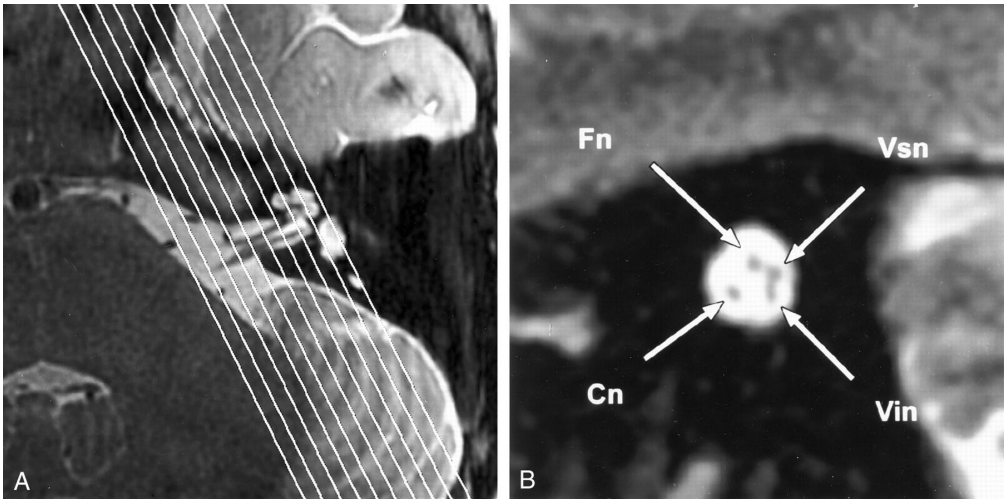
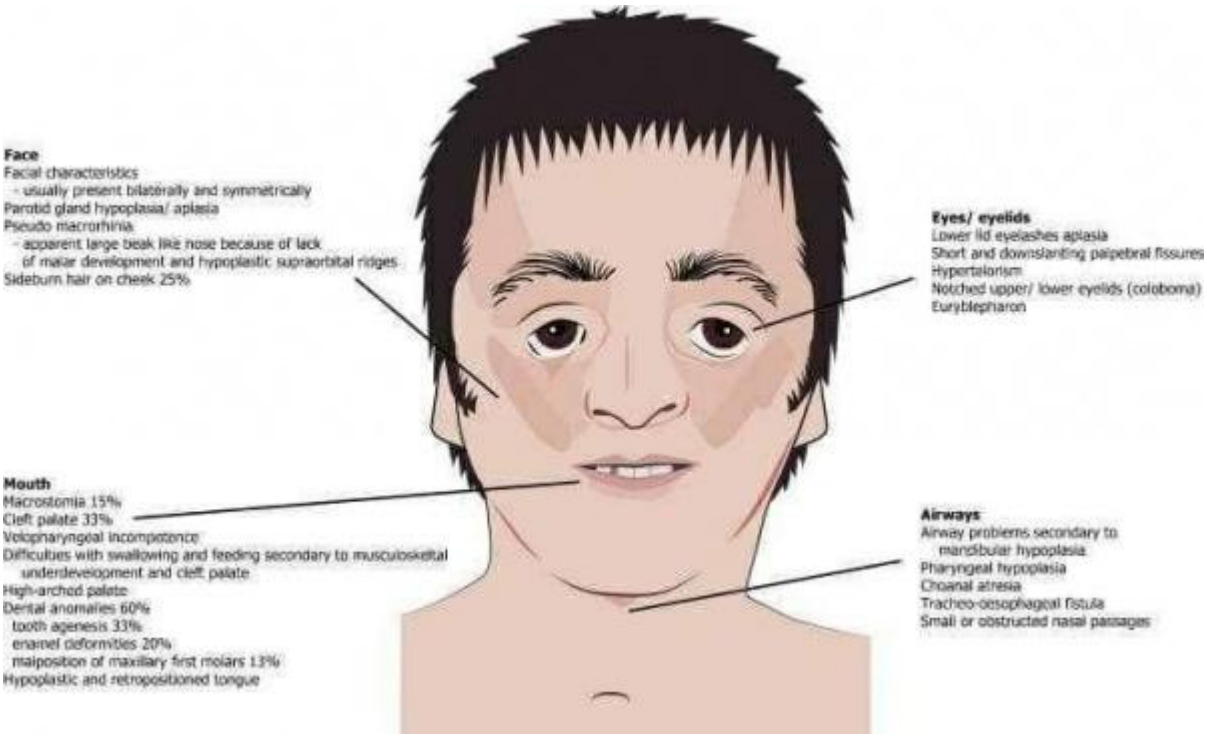


Image 13

- MRI IAC FIESTA: normal vs absent cochlear nerve
- HRCT: middle ear and ossicles in Treacher Collins





End of Notes — The Deaf Child

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