1. INTRODUCTION

Hearing loss is the most common sensory impairment in humans, affecting people from infancy to old age. In 2012, World Health Organization (W.H.O) estimated that 360 million persons in the world are suffering from hearing loss (5.3 % of the world’s population). Of these 328 million are adults (183 million males, 145 million females). 32 million are children. Approximately one third of the persons over 65 years are affected by hearing loss.

Hearing loss is classified as conductive and sensorineural hearing loss. Sensorineural hearing loss (SNHL) is due to damage of hair cells of organ of corti in the cochlea. Sensorineural hearing loss may develop in children or adults. It may be congenital or acquired. Congenital causes of SNHL are genetic anomalies and syndromes, maternal infections such as TORCH (Taxoplasmosis, Rubella, Cytomegalovirus and Herpes). Acquired causes of SNHL include age related hearing loss (presbycusis), diseases of blood vessels, immune disease, infections such as meningitis, mumps, scarlet fever, injury, exposure to loud noise, meniere’s disease, acoustic neuroma, ototoxic drugs.

Cochlear implantation is performed to restore hearing in subjects with profound (>90 dB) and severe (70-90 dB) sensorineural hearing loss and thus enhance their ability to participate in aural-oral communication.
1.1 History of Cochlear Implants

Cochlear implant is an electronic device which converts sound signals into electrical impulses. It bypasses the inner ear and provides information to hearing centers in the brain through direct stimulation of the auditory nerve.

In 18th century (1790), Alessandro Volta was the first person to stimulate the auditory system electrically by connecting a battery of 30-40 ‘couples’ to two metal rods that were inserted into his ears.

William House stimulated the auditory nerve via the scala tympani by implantable receiver stimulators. Advancements in microelectronics, biocompatible materials and microscopic otologic surgery made William House to develop the first single-channel implant. In 1972, a speech processor was developed to interface with the House 3M single-electrode implant.

In 1978, Prof. G.M. Clark in Melbourne (Australia) performed the first successful cochlear implantation surgery on an adult patient who after many months of training could hear and speak even though he was deaf for many years since then cochlear implantation was performed in many adult patients. In 1984, FDA formally approved the use of cochlear implant in children.

In 1990’s multi-channel cochlear implant was developed. The multi-channel Cochlear implant enhanced the spectral perception and speech
recognition capabilities compared to single channel device. The late 1990’s and early 2000’s saw technologic improvements such as the peri-modiolar contour electrode, split electrodes, behind-the-ear processors, and implantation for children as young as 12 months \(^5,^6\).

1.2 Parts of Cochlear Implant

Cochlear implants have external (outside) parts and internal (surgically implanted) parts that work together to allow the user to perceive the sound (Figure 1.1).

![Cochlear Implant](https://www.chicagoear.com/cochlear_implants/how_implant_works)

Fig. 1.1: Parts of cochlear implant (Photo Courtesy of http://www.chicagoear.com/cochlear_implants/how_implant_works)
**External parts:** include a microphone, a speech processor and a transmitter.

The microphone picks up sounds- just like a hearing aid and sends them to the speech processor. The speech processor may be housed with the microphone behind the ear, or it may be a small box like unit typically worn in chest pocket.

The speech processor is a computer that analyses and digitizes the sound signals and sends them to a transmitter worn on the head just behind the ear. The transmitter sends the coded signals and sends the coded signals to an implanted receiver just under the skin.

**Internal parts:** The internal parts include a receiver and electrodes. The receiver is just under the skin behind the ear. The receiver takes the coded electrical signals from the transmitter and delivers them to array of electrodes that have been surgically inserted into the cochlea. The electrodes stimulate the fibers of the auditory nerve, and sound sensations are perceived.
1.3 Embryology of Ear

External ear

During fourth and sixth weeks of gestation auricle develops from the six swellings called branchial hillocks of His.

The external auditory canal is formed by the invagination of the first branchial cleft an ectoderm derivative that lies between first and second branchial arches.

The tympanic membrane originates from the endoderm of first pharyngeal pouch and ectoderm of first pharyngeal cleft \(^8,9,10\).

Middle ear

The middle ear develops from endoderm of the first pouch. The expansion of endodermal pouch opens the way of development of Eustachian tube, middle ear cavity and antrum.

During 20-38 weeks few mastoid air cells begin to develop, but most of the air cells develop post natally. Ossicles begin to form as mesoderm condensations from derivatives of first and second branchial arches.

The facial nerve arises from the facial nucleus. The chorda tympani arises from the mastoid segment of facial nerve passes between incus and malleus and exit the anterior petrous bone \(^8,9,10\).
**Inner ear**

The development of inner ear derived from the otic capsule is greatly independent of development of middle and outer ear which take their form from the first and second branchial arches.\(^{11}\)

During fourth week of gestation the inner ear develops from thickening of ectoderm adjacent to the hind brain called otic placode (Figure 1.2 A). The otic placode invaginates into the connective tissue (mesenchyme) adjacent to the rhombencephalon and becomes otic vesicle.

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**Fig. 1.2** (A) The development of otic vesicle. (Photo courtesy of James B snow and P. Ashley wackym).  (B) The development of semicircular canals, utricle, saccule and cochlea from otic vesicle. (Photocourtesy of Ulrich Drews)
The superior portion of the otic vesicle gives rise to vestibular apparatus. The inferior portion of the otic vesicle gives rise to cochlea (Figure 1.2 A).

In vestibular apparatus the semicircular canals appears as out pouching’s during the sixth week of development.

The superior semicircular canal develops first, followed by posterior semicircular canal. The lateral semicircular canal is formed last\textsuperscript{10,12} (Figure 1.2 B).

**The membranous and bony labyrinth**

The membranous labyrinth is usually surrounded by neural crest cells that form a connective tissue (mesenchyme) covering which becomes cartilaginous and then ossifies to become bony labyrinth.

Connective tissue which is closer to membranous labyrinth degenerates to form perilymphatic space which contains perilymph\textsuperscript{13}. The cochlea reaches adult size by 22\textsuperscript{nd} week of gestation\textsuperscript{12}. 
1.4 Applied Anatomy of Ear

The ear is divided into external, middle and internal

**External ear**

External ear consists of auricle or pinna, external auditory canal and tympanic membrane (Figure 1.3).

![Fig. 1.3: Parts of ear (Photo courtesy of Anne Waugh and Allison grant)]

**Auricle**

The auricle is expanded portion projecting from the side of the head. It is made of fibro-cartilage covered with skin.

**External auditory meatus**

This is ‘S’ shaped tube, 2.5-3.0 cm in length extending from the auricle to the tympanic membrane. It has two parts (a) cartilaginous and (b) bony parts. Sebaceous and ceruminous glands are present in the cartilaginous part of EAM.

**Tympanic membrane (Ear drum):** It separates the external auditory meatus from the middle ear. It is oval in shape with the slightly boarder edge upwards
and is formed by three types of tissue: the outer covering of hairless skin, the middle layer of fibrous tissue and the inner lining of mucous membrane continuous with that of middle ear\textsuperscript{14,15}.

**Middle ear**

This is an irregular shaped air-filled cavity within the petrous portion of the temporal bone. The middle ear extends beyond the tympanic membrane and is divided into (i) mesotympanum (ii) epitympanum or attic (iii) hypotympanum.

Middle ear has roof, floor, medial, lateral, anterior and posterior walls (Figure 1.4).

![Figure 1.4: Anatomy of middle ear (Photo courtesy of P.L Dhingra)](image)

**Roof:** It is formed by thin plate of bone called tegmen tympani. It separates middle ear cavity from middle cranial fossa.
**Floor:** It is also thin plate of bone which separates middle ear cavity from jugular bulb.

**Anterior wall** has a thin plate of bone which separates the cavity from internal carotid artery.

**Posterior wall** consists of bony projection called pyramid. Pyramid is present beneath the oval window. It contains stapedius muscle which passes forward to insert into the neck of stapes. Aditus is a large irregular opening connecting with mastoid antrum to the middle ear cavity. Below the aditus is a small depression known as the fossa incudis. Pyramid is present below the fossa incudis and closer to facial nerve.

Between the promontory and the tympanic annulus is the facial recess. The facial recess is bounded medially by the facial nerve and laterally by the chorda tympani and superiorly by fossa incudis.

**Medial wall**

It consists of prominent bulge caused due to basal turn of the cochlea called promontory. Oval window is present behind and above the promontory which is closed by foot plate of stapes.

Round window is present behind and below the oval window which is covered by secondary tympanic membrane. Above the oval window there is
canal for facial nerve. Just anterior to the oval window, the medial wall presents a hook like projection called processus cochleiformis.

Medial to the pyramid is a deep recess called sinus tympani which is bounded by subiculum below and ponticulus above (Figure 1.5).

![Fig. 1.5: Structures present on medial wall of middle ear (Photo courtesy of P.L Dhingra)](image)

**Anatomy of round window**

Round window was first described by Antonio Scarpa in 1772. Scarpa called the round window membrane as secondary tympanic membrane\textsuperscript{16}.

The round window membrane is located in the round window niche, inferior to oval window. The RWM lies in the roof of the round window niche and lies mostly in the horizontal plane. The round window niche is formed by two over hangs postero-superior and antero-inferior overhang (Figure 1.6 A).
The round window membrane is visible after removal of over hangs of the round window niche (Figure 1.6 B).

The round window membrane has anterior and vertical portions. The anterior portion lies in the vertical plane where as posterior portion lies in horizontal plane. The conical shape of the round window membrane results from fusion with the base of the primary osseous spiral lamina.
**Mastoid antrum:** It is large, air containing space in the upper part of mastoid and communicates with epitympanum through aditus. It is marked externally on the surface by supra meatal triangle (Macewan’s triangle) (Figure 1.7).

![Mastoid antrum and MacEwen’s triangle](image)

Fig. 1.7: MacEwen’s triangle. It is bounded by temporal line (a), Posterosuperior segment of bony external auditory canal (b) and the line drawn as tangent to the external canal (c). It is an important landmark to locate the mastoid antrum in mastoid surgery (Photo courtesy of P.L. Dhingra)

**Ossicles of the middle ear**

The malleus, incus and stapes are the ossicles of middle ear.

The malleus has head, neck, handle (manubrium) a lateral and anterior process. Head and neck lies in attic. Manubrium is attached to tympanic membrane. The lateral process forms a knob like projection on the outer surface of the tympanic membrane and gives attachment to the anterior and posterior folds. The incus has a body and a short process both of which lie in attic and long process which hangs vertically and attaches to the head of stapes.
The stapes has a head, neck, anterior and posterior crura and a foot plate. The foot plate is held in the oval window by annular ligament.  

**Inner ear**

The inner plays vital role in hearing and balance. It consists of bony labyrinth and membranous labyrinth. Bony labyrinth consists of vestibule, semicircular canal and cochlea. Vestibule is central part of labyrinth. Oval window is present at its lateral wall. Saccule and utricule are present inside the vestibule.

**Semicircular canals:** Lateral, posterior and superior semicircular canals lie at right angles to one another. Each semicircular canal has an ampullated end which opens independently into the vestibule and a non ampullated end.

The non ampullated ends of posterior and superior semicircular canals unite to form common crus. Thus the three canals open into vestibule by five openings.

**Cochlea:**

It is a coiled structure making around 2.5 to 2.75 turns around modiolus. From modiolus a bony ridge called osseous spiral lamina projects into the canal, winding around modiolus like the thread of a screw.
**Compartments of cochlea**

Two membranous partitions extend between the osseous spiral lamina and outer wall of the spiral canal.

1. Basilar membrane
2. Vestibular membrane

Basilar membrane stretches from tip of the osseous spiral lamina to tough dense fibrous called spiral ligament which lines the outer wall of the canal.

Vestibular membrane also called Reissner’s membrane. It is placed obliquely between the upper surface of osseous spiral lamina and upper part of spiral ligament. Both these membranes divide the spinal canal of cochlea into three compartments scala vestibuli, scala tympani, scala media (Figure 1.8).

![Fig. 1.8: Cross section of bony canal of cochlea (Photo courtesy of K. Sembulingam and Prema Sembulingam)]
Scalars vestibuli: Scala vestibuli is closed by the foot plate of stapes which separates it from the air filled middle ear.

Scalars tympani: Scala tympani is closed by secondary tympanic membrane; it is connected with the subarachnoid space through the cochlear aqueduct.

Scalar media: is also called cochlear duct or membranous cochlea. It is a triangular compartment enclosed by basilar and vestibular membrane.

Membranous labyrinth consists of cochlear duct, utricle, saccule and three semicircular canal ducts and endolymphatic sac.

Organs of corti: it is the sense of hearing situated on the basilar membrane. It consists of tunnel of corti, Hair cells, supporting cell.

Hair cells: They are important receptor cells of hearing and functions as transducer which converts sound energy into electrical energy. Inner hair cells form a single row while outer hair cells are arranged in three or four rows. Inner hair cells are richly supplied by afferent cochlear fibres and are probably meant in transmission of auditory impulses. Outer hair cells mainly receive efferent innervations from the olivary complex and are concerned with modulating the function of inner hair cells.
1.5 Patho-Physiology of Hearing

The sound received from the environment is collected by the pinna, which passes through the external auditory meatus and hits the tympanic membrane. The vibrations of the tympanic membrane are transmitted from the tympanic membrane through chain of ossicles of middle ear to the foot plate of stapes.

The movement of foot plate of stapes causes pressure changes in the labyrinthine fluids which move the basilar membrane. This stimulates the hair cells of the organ of corti. These hair cells act as transducer which converts mechanical energy into electrical impulse and travel along the auditory nerve (Figure 1.9).

Fig. 1.9: Mechanism of hearing (Photo courtesy of K Sembulingam and Prema sembulingam)
Theories of mechanism of hearing

1. Travelling wave theory

Movement of foot plate of stapes against oval window causes movement of perilymph in scala vestibuli. This fluid does not move all the way from oval window to round window through helicotrema. It immediately hits the vestibular membrane near oval window. This causes movement of fluid in scala media, since vestibular membrane is flexible. Movement of fluid in scala media causes bulging of basilar membrane towards scala tympani (Figure 1.10).

Bulging of basilar membrane increases the elastic tension in basilar fibers in that portion of basilar membrane. Elastic tension in basilar fibers initiates wave, which travels along basilar membrane towards the helicotrema. It is called travelling wave.²²

Fig. 1.10: Displacement of fluid (arrows) (Photo courtesy of K Sembulingam and Prema Sembulingam)
Tonotopic organization of cochlea

Physical characteristics of the basilar membrane cause different frequencies to reach maximum amplitudes at different positions. Higher frequencies are transduced at the base of the cochlea whereas low frequencies are transduced at the apex (Figure 1.11).

Volley theory

In 1949, wever postulated this theory. According to this theory, the impulses of sound waves with frequency above 1,000 cycles per second are transmitted by different groups of nerve fibers.

Place theory

According to this theory, nerve fibers from different portions (Places) of organ of corti on basilar membrane give response to sounds of different frequency. Accordingly, corresponding nerve fiber from organ of corti gives
information to the brain regarding the portion of organ of corti that is stimulated.

**Diseases of external ear**

**External otitis**

It is inflammation of the external ear canal. Staphylococcus aureus is the usual cause of localized inflammation (boils) in the external auditory canal. When more generalized, the inflammation may be caused by bacteria or fungi or by an allergic reaction to dandruff, soaps, hair dyes.

**Cerumen or wax**

One of the more common conditions found is obstruction of ear canal caused by mass of wax. Wax is mixture of dead skin, oily secretions and sweat.

**Exostoses**

Sometimes there are bony narrowing’s of the ear canal known as exostoses, which are often found in people who have swum a great deal in cold water. They sometimes look like white pearls and are frequently mistaken for cysts. Their only importance is that they may obstruct the view of tympanic membrane and may be mistaken for pathology.

**Atresia of EAC:** It is absence or closure of the External auditory canal. The malformations of the middle ear bones may be affected including the narrowing or ear canal known as canal stenosis.
Microtia

It is congenital deformity affecting the outer ear (pinna) where the ear does not fully develop during the first trimester of pregnancy. A microtia is often smaller in size, can have a peanut shaped appearance or completely absent at birth.\(^\text{24}\).

Diseases of middle ear

Acute otitis media

This is the inflammation of the middle ear cavity usually caused by upward spread of microbes from an upper respiratory tract infection via the auditory tube. It is very common in children. Microbial infection leads to accumulation of pus and outward bulging of the tympanic membrane.

Chronic otitis media

In this condition there is permanent perforation of the tympanic membrane following otitis media (especially when recurrent, persistent or untreated). During the healing process stratified epithelium from the outer ear canal sometimes grows into middle ear forming a cholesteatoma. This is a collection of desquamated epithelial cells and purulent material. Continued development of cholesteatoma may lead to destruction of ossicles and conduction deafness,
erosion of the roof of middle ear and middle ear, spread of infection to inner ear that may cause labyrinthitis.

**Serous otitis media** is collection of fluid in the middle ear cavity. It is also known as glue ear.

**Otosclerosis:** Abnormal bone develops around the foot plate of stapes suing it to oval window, reducing the ability to transmit sound waves across the tympanic cavity. It is common cause of progressive conductive hearing loss in young adults 14.

**Diseases of inner ear**

**Congenital malformations of inner ear:**

**Labyrinthine aplasia:** It is also known as Michel’s aplasia. It occurs due to developmental arrest of otic placode in the third week of gestation. On imaging, complete absence of entire cochlea and vestibular structures are seen. Internal auditory canal is typically absent or small in size.

**Cochlear aplasia:** It occurs due to developmental arrest in the late third week of gestation. It is characterized by complete absence of the cochlea. The vestibule and semicircular canals are developed but typically deformed.
**Common cavity deformity:** It results from developmental arrest during fourth week of gestation. There is no differentiation between the cochlea and the vestibule and they appear as common cavity. Semicircular canals are usually absent but may be normal or dysplastic. Internal auditory canal is usually present and may be large or small in size.

**Incomplete partition type – I:** It results from otic placode developmental arrest in the 5th week of gestation. Cochlea appears cystic and entire modiolus is absent. Vestibule is always dilated. IAC is usually large. Vestibular aqueduct is normal.

**Cochleo-vestibular hypoplasia:** It results from developmental arrest during sixth week of gestation. Cochlea is small in size and often with a single turn. Vestibule is hypoplastic or may be absent.

**Incomplete partition type-II:** It is the Classic Mondini malformation. It occurs due to developmental arrest of otic placode during seventh week of gestation. The defect is limited to the apical and middle turns of the cochlea with deficient intercalar septum which results in cystic appearance of cochlear apex only. Over all the cochlea and the vestibule are normal in size but the cochlea has only 1.5 turns. The basal turn of cochlea is normal. Vestibular aqueduct is enlarged.
**Semicircular canal anomalies**

**Lateral Semicircular canal -vestibular dysplasia:** In this lateral semicircular canal is short and fuses with dysplastic vestibule.

**Large vestibular aqueduct syndrome:** Most common imaging finding in children with congenital SNHL is large vestibular aqueduct. It is usually bilateral. Width of vestibular aqueduct should be more than 1.5 mm measured at the midpoint between common crus and external aperture. It occurs due to insult at seventh week of gestation\(^\text{25}\).

**Meniere’s disease**

In this condition there is accumulation of endolymph causing distension and increased pressure within the membranous labyrinth with destruction of sensory cells in the ampulla and cochlea, it is usually unilateral first but both ears may be affected later. It is associated with episodes of dizziness (vertigo), nausea and vomiting\(^\text{14}\).

**Labyrinthitis ossificans**

It most commonly occurs as sequel of inflammation of the inner ear that results from bacterial meningitis. On HRCT the lumen of labyrinth shows ossification and on MRI the lumen of the labyrinth shows reduced fluid signal intensities on T2 weighted images\(^\text{26}\).
Hearing loss

According to World Health Organization (W.H.O), disabling hearing loss refers to hearing loss greater than 40 decibels (dB) in better hearing ear in adults and a hearing loss greater than 30 dB in better hearing children.

A person who is not able to hear as well as someone with normal hearing-hearing thresholds of 25 dB or better in both ears is said to have hearing loss. Hearing loss may be mild, moderate, severe or profound. It can affect one ear or both ears, and leads to difficulty in hearing conversational speech or loud sounds.


‘Hard of Hearing’ refers to people with hearing loss ranging from mild to severe. They usually benefit communicate through spoken language and can benefit from hearing aids. People with more significant hearing loss may benefit from cochlear implants.

There are two principal kinds of hearing loss, Conductive and Sensorineural hearing loss. Hearing loss can also be mixed where there is combination of conductive and sensorineural deafness in one ear.
Conductive hearing loss is due to impaired transmission of sound waves from outside to the oval window, i.e. an abnormality of the outer or middle ear.

Common causes of conductive deafness are wax or foreign body, acute otitis media, serous otitis media, chronic otitis media, Barotrauma, otosclerosis, External otitis, injury of tympanic membrane.

Sensorineural hearing loss (SNHL)

Results from damage to the neural receptors of the inner ear (the hair cells, organ of corti), the nerve pathways to the brain (notably the auditory nerve), or the area of the brain that receives sound information.

Deafness of this type is usually permanent. It can be congenital or accompany other birth related problems such as erythroblastosis fetalis (Rh incompatibility) or Anoxia (lack of oxygen during delivery). Before vaccines were available, German measles (rubella) and common measles (rubeola) were leading causes; maternal cytomegalovirus and genital herpes simplex virus.

Tumors, injury, stroke, toxic substances and certain over the counter and prescription drugs (eg: Streptomycin) are additional factors that can affect auditory pathways and the brain and lead to sensori-neural hearing loss.

Continued exposure to loud noise, as in certain industries or from loud music can result in damage to the inner ear, causing irreversible hearing loss 28.
1.6 Assessment of Hearing loss

Audiometric tests are done to determine subjects hearing levels, type and severity of hearing loss.29.

There are two types of audiometric tests

1. **Subjective Audiometry**: Pure Tone Audiometry (PTA), Behavioural Observation Audiometry (BOA), Visual Reinforcement Orientation Audiometry (VROA), Play audiometry.

2. **Objective Audiometry**: Otoacoustic emission Audiometry (OAE), Automated Auditory Brainstem Response Audiometry (AABR).

**Subjective audiometry**

**Pure Tone Audiometry**: Pure tone audiometry is the standard behavioral assessment of individual’s hearing by using pure tones. In this the subject is instructed to listen carefully for a beeping sound and when he or she hears, a sound, even it is very soft, to raise his or her hand. The audiologist finds the softest sound the subject can hear across a range of frequencies and determines the hearing thresholds. The hearing thresholds are plotted on a graph called audiogram. Frequency ("pitch") is measured in Hertz plotted on X-axis with low frequencies on the left to high frequencies on the right. Loudness ("Volume") measured in decibels plotted on the Y-axis, with very soft sounds
at the top of the graph increasing to very loud sounds at the bottom \(^{30}\) (Figure 1.12).

![Audiogram with pitch on x-axis and hearing levels on y-axis.](http://www.hearingaidknow.com)

**Fig. 1.12: Audiogram with pitch on x-axis and hearing levels on y-axis.**

(Photo courtesy of http://www.hearingaidknow.com)

The pure tones used may be presented via: Head phones, Insert ear phones, Bone conductor.

Head phones and insert ear phones tests the entire hearing system. Thresholds obtained with these are called air conduction thresholds. The bone
conductor directly stimulates the cochlea. Thresholds obtained with these are called bone conduction thresholds.

In conductive hearing loss: Air conduction thresholds indicate hearing loss but bone conduction thresholds are normal.

In sensori neural hearing loss: Bone conduction thresholds match air conduction thresholds.

In mixed hearing loss: both bone and air conduction thresholds are elevated, but air conduction thresholds are poorer than bone conduction thresholds.

Pure tone audiometry is used for testing in older children and adults.

Play Audiometry

Play audiometry is suited for children in the age of 3-7 years. In this the child is taught to respond, using a pre-determined task, whenever they hear pure tone. Examples of tasks given are putting a colorful peg into a peg board, dropping a marble in to marble ladder game. 

Visual Reinforcement Orientation Audiometry (VROA)

It is suited to infants aged 7 to 8 months to 3 years developmentally. The child is taught to turn their head when a sound is heard. Initial conditioning is
achieved by introduction of stimuli at moderately high levels and audiologist waits until the child looks for the source of sound. They are then shown a colourful, moving puppet or toy under illumination as a reward. This puppet or toy reinforces child’s turning behavior or orientation. Once this conditioned response is reliably observed, the stimuli can be presented at ever decreasing levels until auditory levels have been reached.

**Behavioral Observation Audiometry (BOA)**

It is suited to infants aged less than seven months. It might also be used for older children who cannot taught to respond for VROA or Play audiometry because of disabilities that affect motor skills or cognition (learning). Young infants generally show little in tonal stimuli when they hear good hearing. BOA testing therefore is performed using stimuli or “noisemakers” such as baby rattles, drums, squeaking toys.

There are some significant difficulties with BOA testing. First, the infants response to these sounds is strongly influenced by their age and state on the day of the test. If they are sleepy, hungry, sick or crying then outcomes will be unreliable. Second, many infants may not show behavioral changes to sound until they are quite loud. Third, some infants may be more interested in some types of sounds than others.\(^{30}\)
Objective Audiometry

Otoacoustic emission Audiometry (OAE)

The primary purpose of Otoacoustic emission audiometry is to determine cochlear status especially hair cell function. The outer hair cells produce otoacoustic emissions (Low intensity sounds) as they expand and contract. It is used for testing new born babies. In this a small ear piece containing a microphone and a mini loud speaker is placed in the ear. The loud speaker makes clicking sound in the ear. If the cochlea is working normally it responds sending a sound back to ear canal. This is detected by microphone.  

Automated Auditory Brainstem Response (AABR)

In this test a small ear phone plays click into the baby’s ear. If the baby can hear the click, a signal in the hearing nerve can be measured from the sensors that are placed on baby’s skin.
1.7 Preoperative Radiological Evaluation for Cochlear Implantation

The radiological evaluation during preoperative Cochlear implant work up includes High Resolution Computed Tomography (HRCT) of temporal bone and Magnetic Resonance Imaging (MRI) of inner ear.

HRCT temporal bone has become an important part of the preoperative assessment for cochlear implantation. It also has a significant role in making decision for performing implantation and choosing the side of implantation\textsuperscript{32,33}.

HRCT Temporal bone is useful in evaluation of the degree of pneumatisation of mastoid air cells, mastoiditis, anatomical variations in position of sigmoid sinus, carotid artery, and facial nerve, dehiscent jugular bulb, presence of Korner’s septum, persistent stapedial artery, integrity of middle ear ossicles, anomalies of the bony labyrinth, semicircular canals dehiscence, width of Internal auditory canal, cochlear duct, vestibular duct and ossification of cochlea\textsuperscript{5,34,35,36}.

HRCT temporal bone has limitations in demonstration of fluid in the inner ear, the early detection of labyrinthitis ossificans in post meningitic cases and in identification of presence/absence of Cochleo vestibular nerve in cases of narrow internal auditory canal\textsuperscript{5}. Preoperative MRI of inner ear is useful in evaluation of Cochleo-vestibular nerve, fluid status of the membranous labyrinth, Cochleo-vestibular anomalies\textsuperscript{5,37}.  

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1.8 Principles of Multiple-row Detector Computed Tomography (MDCT)

Multislice CT has opened new avenues and opportunities to improve the quality of care offered to the patients. Multislice CT provides faster scanning (Figure 1.13 B) and higher resolution with the introduction of several new concepts relating to detector technology, the geometry of data acquisition, multislice image reconstruction algorithms. Multislice CT has lead to the development of Functional CT imaging. Multislice CT uses the helical principle and allows for acquisition of more than one slice of data simultaneously with the help of multiple row of detector elements \(^{38,39}\) (Figure 1.13 A).

Fig. 1.13: (A) Acquisition of more than one slice of data simultaneously with Multidetector CT (MDCT) compared to Single Slice CT (SSCT) in equal scanning times. (B) Scan time is less and Volume coverage is more with MDCT compared to SSCT in equal scanning times
Physical principles:

Data acquisition

Unique features of multi slice data acquisition are collimation, beam geometry, and pitch.

Collimation

The beam is collimated by a pre collimator to fall on the entire multi row detector array. The multiple slices are a result of division of the division of the total X-ray beam in to multiple beams depending on the number of detector rows. To minimize the scatter radiation, post collimation is used.

In multislice CT, the detector row collimation is 1/N of the X-ray beam collimation. N= no. of detector rows. This makes it possible to simultaneously achieve high volume coverage speed and high z axis resolution.

Beam geometry

Cone beam geometry is used to cover the wide area of the detector array.

Pitch

Pitch is an important parameter that combines the table distance travelled per 360-degree gantry rotation with slice thickness.

Pitch = Table translation (mm) per 360° gantry rotation / Total nominal slice width (mm).
Pitch relates to the volume covered and also to the patient dose.

**Instrumentation:**

**X- ray tube**

The x ray tube is a rotating- anode tube capable of high heat storage capacity with high anode and tube housing cooling rates.

**X- ray generator**

The X- ray generator is a compact, light weight, high frequency generator that provides a stable high voltage to the X- ray tube to ensure efficient production of X- rays.

**Detectors**

Gather information by measuring the X- ray transmission through the patient & convert the x ray transmission data into electrical signals. In MDCT an assembly of multiple solid state detector array modules is used. There are basically two types of detectors used in multislice CT scanning $^{38,39}$ (Figure 1.14).

- **Fixed width detector array**
  - Matrix detector array

- **Variable-width detector array**
  - Hybrid detector array
  - Adaptive detector array

![Fig. 1.14: Types of Detector configuration](image-url)
Detector configuration:

The term detector configuration describes the number of data collection channels and the effective section thickness determined by the data acquisition system settings.

Data acquisition system:

The detector electronics responsible for digitizing signals from the detectors before they are sent to the computer for post processing.

Computer system:

The computer system for multislice CT receives data from the data acquisition system and the operator, who inputs patient data and various examination protocols. These systems must be capable of handling vast amount of data collected by the multi detector elements.

Advantages of MDCT:

- Increase in speed and volume coverage.
- Improved z-axis spatial resolution.
- Increased temporal resolution.
- Acquire true isotropic voxel by means of thinner slice thickness.
- Dedicated CT scanners for imaging the heart.
1.9 HRCT and MRI Protocol for Inner ear:

**HRCT**

The axial images of the temporal bone are obtained with thickness of 0.6-0.7 mm using High resolution spatial frequency (bone plus) algorithm, large matrix size (512x512) with smaller Field of view (FOV).

**MRI**

A routine T2 weighted axial sequence through brain is obtained in all patients. For evaluation of inner ear 3D FIESTA (Fast Imaging Employing Steady State Acquisition) is done in axial. In addition, especially for evaluation of nerves a 3D FIESTA is also acquired in a direct oblique sagittal plane perpendicular to the VII –VIII nerve complexes.

1.10 Steps in Cochlear Implantation Surgery

Cochlear implant surgery is performed under general anesthesia. Adequate hair is shaved on the implantation side. The template is placed on the head and its position is marked with ink (Figure 1.15 A).

The internal receiver has to be placed slightly behind the edge of the auricle, such that it does not interfere with the external process behind the ear. The tip of the implant is positioned above the canthomeatal line.
After the position of the device and the centre of the well are marked, an incision is drawn on the skin (Figure 1.15 B).

Fig. 1.15: (A) Marking of template on the side of implantation. (B) The incision line extending from the auricle to posteriorly. (C) Well created for the internal receiver (black arrow) and groove for placing the electrode from receiver into mastoid cavity (black dotted arrow). (Photo Courtesy of H. Hildmann, H. Sudhoff)

Planning the flap:

The incision is made in the post auricular sulcus near the mastoid tip, extending to the attachment of auricle in a curved fashion. The incision is made with cutting and coagulation current to reduce blood loss from the scalp.

The external skin muscle periosteum flap is raised, exposing the bone of the skull above the mastoid area. The flap and scalp are retracted using self retaining retractors (Figure 1.15 C).
**Preparation of the implant bed for the receiver:** The site for the internal receiver is prepared by drilling the bony evacuation according to the shape of the template. Holes are drilled above and below the anterior and posterior parts for tying the internal receiver. A groove is drilled for leading the electrode array from the receiver to the mastoid cavity (Figure 1.15 C).

**Mastoidectomy:** The mastoidectomy includes the removal of the mastoid cells, exposing the sinus dural angle, the bony shell of sigmoid sinus, the skeletonization of the labyrinth and the opening of the antrum to the epitympanum. The external landmarks such as spine of henle and temporal line are identified and limited mastoidectomy is performed to reach the facial recess. The landmarks such as sigmoid sinus lateral semicircular canal and short process of incus in fossa incudis are identified (Figure 1.16 A-B).

![Fig. 1.16](image.png)

Fig. 1.16 (A) The structures that are identified after performing mastoidectomy. (B) Intraoperative photograph of left ear showing the structures that are identified after performing mastoidectomy
Posterior tympanotomy:

The facial recess is a triangular space bounded posteriorly by the upper vertical portion of the facial nerve, superiorly by the short process of the incus, and anteriorly by the chorda tympani nerve. This space must be opened adequately to visualize the round window area and the electrode placement.

The facial nerve is identified under a thin layer of bone opening the facial nerve. The facial recess is widened inferiorly and posteriorly. The bony buttress between the facial recess and the fossa incudis is preserved. The buttress is used to fix the cochlear implant electrode. The distal long process of incus, the incudo stapedial joint, the stapedial tendon, the promontory and round window has to be identified (Figure 1.17 A-B).

Figure 1.17 (A) The structures that are identified after performing posterior tympanotomy (B) Intraoperative photograph of left ear showing the structures that are identified after performing posterior tympanotomy
The extent of drilling is limited initially by four landmarks: the facial nerve medially, the annulus laterally, the bar of bone to the left covering the short process of incus superiorly and chorda tympani inferiorly.

**Cochleostomy:**

The scala tympani of the cochlea is opened by either taking down the round window membrane or by direct cochleostomy.

**Cochleostomy approach:** Cochleostomy is performed anterior and inferior to round window membrane. Then electrode is inserted through cochleostomy (Figure 1.18 A-B).

![Figure 1.18](image)

**Fig. 1.18** (A) Showing the cochleostomy which is done anterior and inferior relative to round window membrane (B) Showing the insertion of electrodes through the cochleostomy site. (Photo Courtesy of H. Hildmann and H. Sudhoff)
**Round window membrane approach:** The tegmen of the niche is removed in order to visualize the membrane. Then electrode is inserted through round window membrane \(^{11,41,42}\) (Figure 1.19 B).

![Fig. 1.19 (A) Showing the cochleostomy which is done anterior and inferior relative to round window membrane (B) Showing the round window membrane approach where the over hangs are completely drilled to visualize round window membrane](image)

### 1.11 Postoperative Radiographic Evaluation for Cochlear Implantation:

Modified Stenver’s and Perorbital or Trans orbital view are taken to assess integrity, positioning and depth of insertion of electrodes.

**Modified Stenvers view:** It is taken with patient positioned prone or erect facing vertical bucky and the implanted side is kept in contact. The head is adjusted such that mid sagittal plane form an angle of 50° with the plane of the film (Figure 1.20 A). The central ray is directed 2 cm above and parallel to the infraorbitomeatal line \(^{43,44}\).
Method for counting electrodes:

Marsh et al.\textsuperscript{43} have proposed the method of counting the number of electrodes inside the cochlea on modified stenvers view (Figure 1.21).

Fig. 1.21: Method for counting the electrodes inside the cochlea on modified stenvers view.
Trans-orbital view or Per-orbital view:

It is taken with patient positioned prone or erect facing the vertical bucky. The neck is flexed so that the orbito-meatal baseline is perpendicular to the bucky. The central ray is angled 5-10° caudally, centering between the orbits (Figure 1.22 A).

Fig. 1.22 (A) Patient positioned prone, the central ray is directed caudally at an angle of 5-10° to orbitomeatal baseline. (B) Post operative perorbital view radiograph showing cochlear implant on left side; and electrodes inside the cochlea (white arrow)