"ELECTROPHYSIOLOGY AND AUDITORY PERFORMANCE OF CHILDREN WITH PROFOUND SENSORYNEURAL HEARING LOSS AFTER COCHLEAR IMPLANT SURGERY"

Dissertation Submitted to

THE TAMILNADU Dr. M.G.R. MEDICAL UNIVERSITY

Submitted in partial fulfillment of the rules and regulations for the

M.S. DEGREE BRANCH- IV OTORHINOLARYNGOLOGY

UPGRADED INTITUTUE OF OTORHINOLARYNGOLOGY

MADRAS MEDICAL COLLEGE

CHENNAI-600 003

MAY- 2018

DECLARATION

I solemnly declare that the dissertation **"ELECTROPHYSIOLOGY AND AUDITORY PERFORMANCE OF CHILDREN WITH PROFOUND SENSORYNEURAL HEARING LOSS AFTER COCHLEAR IMPLANT SURGERY"** is done by me at the Upgraded Institute of Otorhinolaryngology, Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai during March 2016 – August 2017 under the guidance and supervision of **Prof. Dr. R. MUTHUKUMAR., MS, DLO., DNB.**

This dissertation is submitted to The Tamil Nadu Dr. M.G.R. Medical University, towards partial fulfillment of rules and regulations for the award of M.S. DEGREE IN OTORHINOLARYNGOLOGY (BRANCH - IV)

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BONAFIDE CERTIFICATE

This is to certify that the dissertation entitled "**ELECTROPHYSIOLOGY AND AUDITORY PERFORMANCE OF CHILDREN WITH PROFOUND SENSORYNEURAL HEARING LOSS AFTER COCHLEAR IMPLANT SURGERY",** is a bonafide record of work done by **Dr. R. MAHESH KUMARI,** under our direct guidance and supervision. The dissertation is submitted in partial fulfillment of rules and regulations of The Tamil Nadu Dr. M.G.R. Medical University, Chennai, for the award of **M.S. Degree in OTORHINOLARYNGOLOGY, (BRANCH IV)** in the examination of May 2018.

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ACKNOWLEDGEMENT

I would like to acknowledge with sincere gratitude to the great people who have made it possible for my successful completion of the thesis.

I would like to thank ***Prof. Dr. NARAYANA BABU, M.D.DCH.,*** *the DEAN, Madras Medical College, for having permitted me to conduct the study in the hospital and for using the resource materials.*

I would like to express my sincere gratitude to ***Prof. Dr. R. MUTHUKUMAR, M.S. D.L.O. D.N.B.,*** *the Director and Professor, Upgraded Institute of Otorhinolaryngology, and also my guide for the study. I am deeply indebted to my guide for giving me support in choosing the topic and for scholarly guidance in the conduct of the study. I am thankful to him for the supervision, constant encouragement, mental support and constructive suggestions at every stage of the study, which helped me to complete the study. I express my heartfelt gratitude for his painstaking efforts in completion of the study in a better way.*

I am grateful to ***Prof. Dr. G. SANKARANARAYANAN, MS, DLO., DNB, MNAMS.,*** *Professor, Upgraded Institute of Otorhinolaryngology, for the encouragement and support during the conduction of the study process.*

I am greatly indebted to ***Prof. Dr. SURESH KUMAR M.S. D.L.O.,*** *Professor, Upgraded Institute of Otorhinolaryngology, for his valuable support and guidance during the study.*

I am greatful to my co-guide, **Mrs. V. JAYA., M.Sc., (SP & HG)** for her valuable support, guidance and resources provided to the study and technical support related to the study.

*I express my sincere thanks to all the Assistant Professors, **Dr. S. SEMMANASELVAN, Dr. M. VENUGOPAL, Dr. S. SHENBAGAVALLI, Dr. VIVEK,** for their thoughtful guidance throughout the work.*

I thank the Secretary and Chairman of Institutional Ethical Committee, Government General Hospital and Madras Medical College, Chennai.

I also thank Dr. Sindhu, Dr. Kalaimani my colleagues and other post graduates for their valuable suggestions and help during the study.

I am thankful to all the staff members in the department for the help provided in the completion of the study.

I am very grateful to my family, my father and mother, my son, brother and inlaws, for standing with me all the time and for the mental support and encouragement in the completion of the study.

*I express my sincere thanks to my husband, **Dr. K. Kumaresan**, for helping in data analysis and for completing the study in a better way and also in standing with me during the difficult times.*

Finally and most importantly without the support and consent of the study participants, it would have been a futile exercise. I am thankful to all the study participants and their parents for giving consent, study related information and co-operating with the study procedures.

Above all I thank God Almighty for his immense blessings.

Dr. R. Mahesh Kumari.

ABSTRACT

Title: "Electrophysiology and Auditory Performance of Children with Profound Sensoryneural Hearing loss after Cochlear Implant Surgery"

Introduction: Cochlear implantation is a powerful tool to gain hearing ability and to achieve age appropriate communication skills in children with severe to profound sensorineural hearing loss.

Objective: To compare the intraoperative and postoperative telemetry of the children with Cochlear implants and to assess the auditory performance of children with sensorineural hearing loss after surgery.

Methodology: A prospective study was done involving 63 children operated for cochlear implant at Upgraded Institute of Otorhinolaryngology, Madras Medical College, Chennai. Intraoperative and postoperative electrode impedance as well as telemetry measurements were done. CAP score was used to assess the auditory performance preoperatively and at follow up.

Results: Majority (41.3%) of the children was in 13 – 24 months age group and 57% were males. Around 45% of children reached CAP score of 3 by 6 months, 38% achieved 4 and 14.3% reached score of 5 by 12 months with a significant increase in follow up. Surgery before 3 years of age had a significant relationship with performance. The electrode impedance and telemetry measurements were found to be predictors of device function.

Conclusion: Our results show that, early implantation leads to better auditory performance compared to implantation at later ages. Electrode impedance and telemetry measures provide valuable information regarding the output and response of the auditory system.

Key words: Cochlear implant, children, early implantation, CAP score, telemetry, impedance.

INDEX

1. INTRODUCTION

Hearing impairment is the most frequent sensory deficit in human populations,[1] affecting all ages from infancy to older age groups and the global burden of hearing impairment is increasing. According to World Health Organization (WHO) [2] estimates, 360 million people are living with disabling hearing loss, which constitutes about 5.3% of the world population. Among them, 328 million (91%) are adults and 32 million (9%) are children.

The Disability Adjusted Life Year's (DALYs) for hearing loss is estimated to be 1.8% of total DALYs and projected to increase to 2.9% by 2030.[3] Countries in South Asia, Asia Pacific and Sub-Saharan Africa have more number of people with disabling hearing loss. [2] The prevalence of disabling hearing loss ranges from 4.6% - 8.8% in South East Asian countries. [4]

The burden of hearing impairment is substantially high in India. In our country approximately 63 million (6.3%) suffer from moderate to severe impairment. The prevalence of adult onset and childhood onset deafness is estimated to be 7.6% and 2.00% respectively.[4] Hearing disability is the second frequent cause of disability in India, the incidence being 7/100000 population. The prevalence of hearing disability is 291 persons per 100000 population, higher in rural areas than urban areas. [5] In India, one in 1000 babies are born profoundly deaf (≥90 dB in better ear) and the burden would be higher, if nearly 40,000 births per day are taken into account. [6]

Hearing loss in children can be congenital or acquired. The consequences of hearing problems are well known. Many of the children with congenital permanent hearing impairment have difficulties in speech and language development. Mild to moderate hearing loss in children can result in developmental delays. Profound hearing loss can lead to significant speech and language delays, resulting in lack of communication skills.

Young children face challenges in developing spoken language, psychological functioning and academic achievement with severe to profound sensorineural hearing loss.[7] Permanent hearing loss have negative impact on the development of auditory skills, speech and language, and educational attainment of children.[8]

It has been noted that 80% of all deafness is avoidable - 50% preventable and about 30% treatable or can be managed with assistive devices.[4] The available treatment for children and other profoundly hearing impaired individuals are hearing aid and cochlear implant surgery. Surgery is indicated for individuals in whom the hearing aids fail or individuals not candidates for hearing aid. [9]

Cochlear implants are implantable biomedical devices, that will not cure deafness but will provide some degree of auditory perception to patients with sensorineural hearing loss.[10] Cochlear implantation is a powerful tool to gain hearing ability and to achieve age appropriate communication skills in children with severe to profound sensorineural hearing loss.[11] Based on evidence from other studies, indications for positive outcomes are shifting towards degree of hearing loss and age of implantation.[12]

The auditory performance and speech perception outcomes are significantly better in children who received implants earlier, compared to children who are implanted later. [12, 13]

The success of cochlear implants is based on the device's ability to send electrical signals to the auditory nerve fibers. The proper functioning of the device and the electrodes are assessed intra-operatively and post-operatively at regular intervals as part of monitoring the patient. [14] The outcome of cochlear implant surgery is measured by means of electrophysiology and others measures like auditory performance, speech and language outcomes, and quality of life can be assessed by various available subjective and objective methods.

2. AIMS AND OBJECTIVES

1. To compare the intraoperative and postoperative telemetry of the children with Cochlear implants.

2. To assess the auditory performance of children with sensorineural hearing loss after Cochlear implant surgery.

3. To study the electrophysiology of cochlear implant children with abnormal cochlear morphology.

3. REVIEW OF LITERATURE

Hearing loss is one of the most common sensory deficit in children, and broadly split into two categories - those having lost hearing before speech development (Pre-lingual) and those after speech development (Post-lingual). [15] Hearing loss can be classified into conductive or sensorineural hearing loss. It can occur both in children and adults. Sensorineural hearing loss (SNHL) occurs as a result of damage or development failure of hair cells of organ of corti in the cochlea. [16]

Before the advent of cochlear implants, the resources for hearing disability were limited to development of communication skills and use of hearing aids. The cochlear implant development has created an opportunity to gain an understanding of the auditory system and better prospects for individuals with profound sensorineural hearing loss.

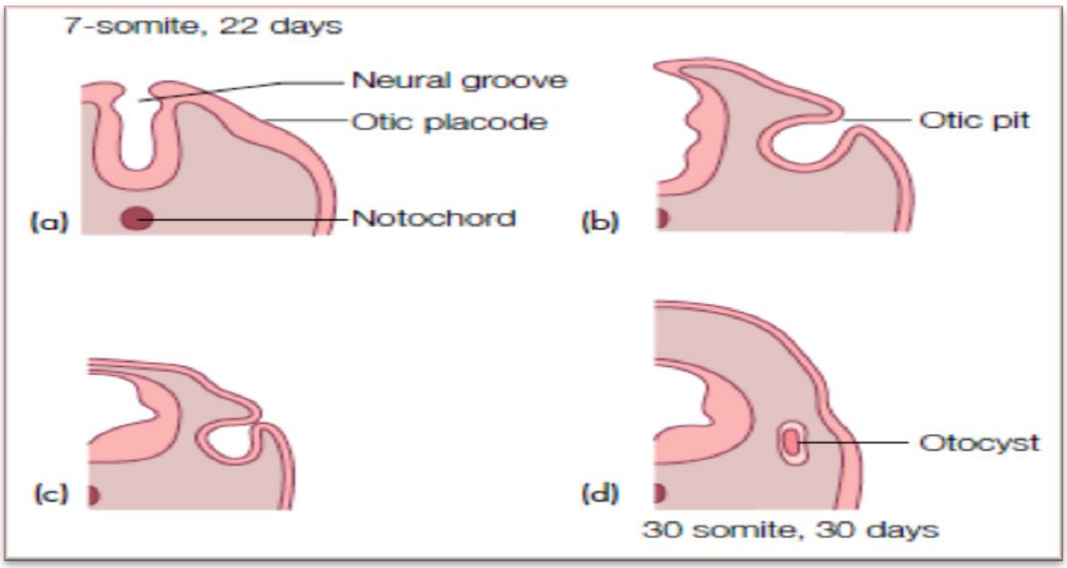
3.1 EMBRYOLOGY OF THE INNER EAR: [17]

The inner ear development is independent of middle and outer ear, interconnected by stapes superstructure thereby giving connectivity to auditory pathway. The development of labyrinth starts with formation of membranous labyrinth, followed by period of encasement by bony labyrinth and production of further series of spaces within the bony shell which in turn becomes the perilymphatic space.

Within first few days of embryonic life (day 22-23) ectodermal thickening forms the otic placode, which deepens and sinks from the surface to form otic pit, and eventually losses from the surface to form otocyst. The otocyst undergoes series of spectacular changes to result in full adult sized membranous labyrinth by 25 weeks of gestation [17]. The development of inner ear is given in Fig 3. 1:

Fig 3.1

Development of otocyst [18]

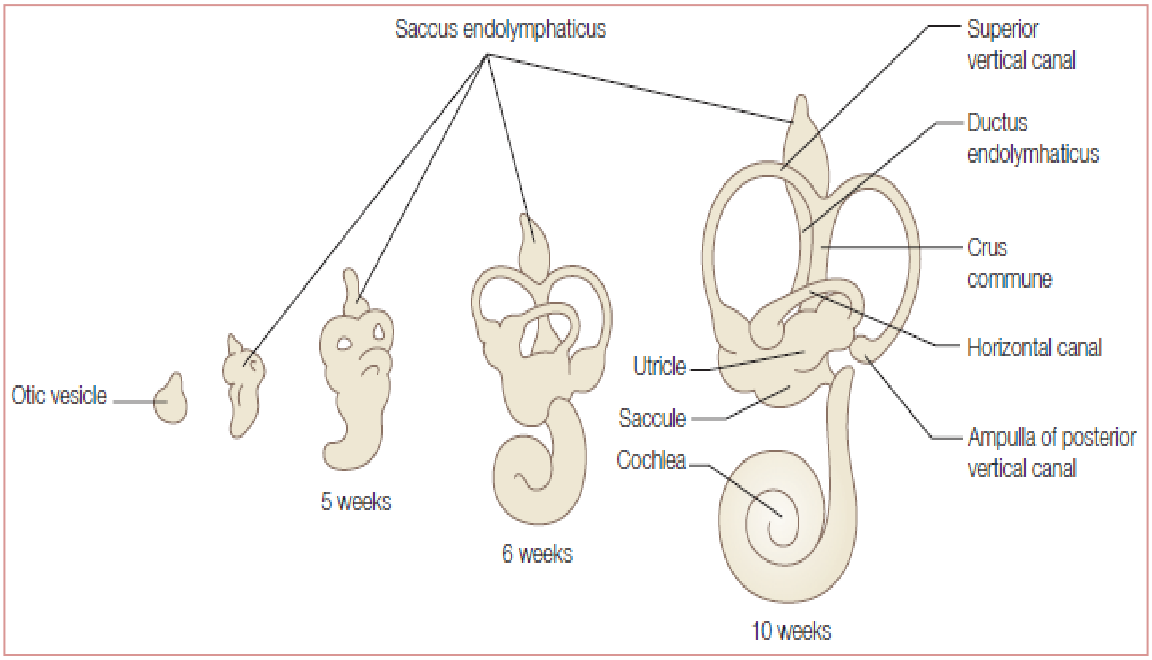


Semicircular canals develop around 35 days as three flattened pouches that grow out at right angles to each other from the utricle. The saccule puts out a single pouch like process that grows & begins to coil from base to apex to reach its full two and half coils by 25 weeks.

Organ of Corti develops as a single block from areas of ectodermal specialization at around 11 weeks. Within the mass develop the outer and inner hair cells along with supporting cells. Differentiation progresses from the base to apex, so at any point of time various stages of development can be seen.[17] The development of membranous labyrinth is depicted in Fig 3.2:

Fig 3.2

Development of membranous labyrinth [18]



BONY LABYRINTH:

The mesenchyme enclosing the otocyst becomes chondrified to form the otic capsule. Otic capsule remodels and undergoes differentiation to form fluid filled spaces that eventually become perilymphatic spaces. The spaces become continuous with CSF by development of cochlear aqueduct, which runs to the posterior cranial fossa from the base of cochlea. The development of communication channels passing through labyrinth is given in table 3.1:

Table 3.1

Development of communication channels passing through labyrinth [18]

	Channel
Internal auditory meatus	Persisting channel in cartilage model around VII and VIII nerves
Subarcuate fossa	Persisting vascular channel
Vestibular aqueduct	Fifth and sixth ossification centres fuse around the endolymphatic duct
Cochlear aqueduct	Resorption of precartilage
Fossula ante fenestram	Resorption of precartilage
Fossula post fenestram (inconstant)	Resorption of cartilage
Oval window	Otic capsule becomes footplate of stapes and annular ligament
Round window	Persisting cartilage becomes round window niche and membrane

3.2 ANATOMY OF THE INNER EAR: [19]

Bony Labyrinth:

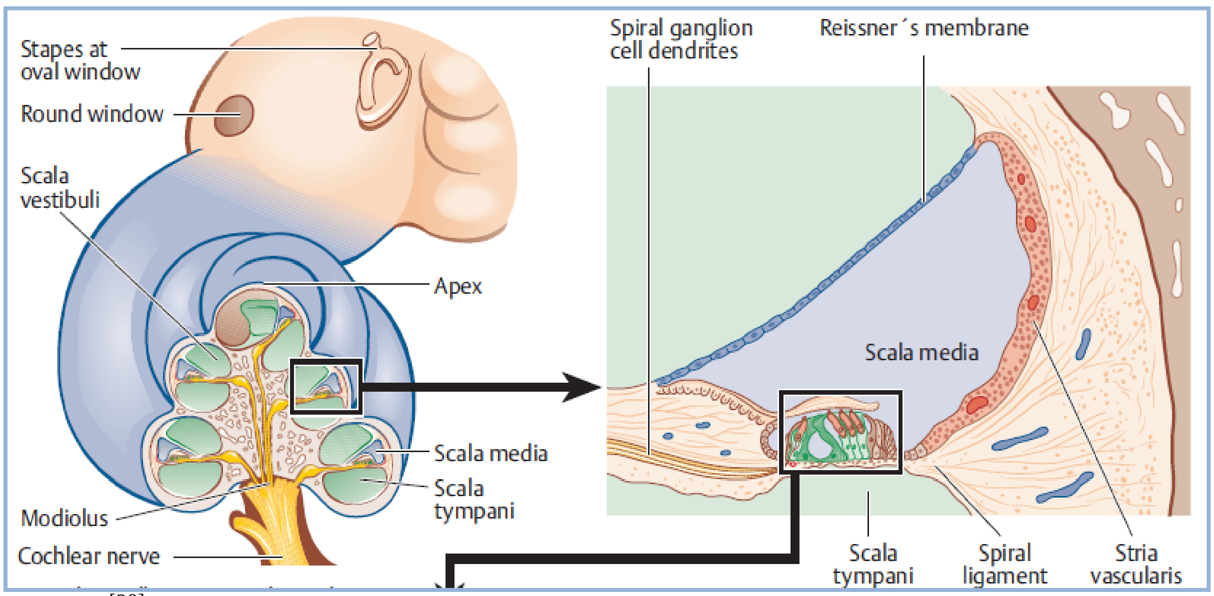
The bony labyrinth shelter's the sensorineural and membranous structures of the inner ear that comprises the vestibule, the semicircular canals, and the cochlea. The bone is trilaminar, with an inner endosteal layer, an outer periosteal layer, and a mixed layer of intrachondrial and endochondral bone, characterized by globuli interossei in between.

The vestibule, measuring 4 mm is the central chamber of the bony labyrinth. It is dominated by the depressions, housing the utricule (the elliptical recess), the saccule (the spherical recess), and the basal end of the cochlear duct (the cochlear recess).

The cochlea is a 32 mm bony spiral that winds 2 1⁄2 turns about its central axis called modiolus, to a total height of 5 mm. The base of the cochlea abuts the fundus of the internal auditory canal, and is perforated for the transmission of cochlear nerve fibers. The osseous spiral lamina also winds about the modiolus, partially subdividing the cochlear canal into the scala tympani and scala vestibuli. The interscalar septum separates the cochlear turns. The microscopic features of cochlea is shown in Fig 3.3

Fig 3.3

Microscopic anatomy of cochlea



Source [20]: Adunka OF, Buchman CA. Otology, Neurotology and Lateral Skull Base Surgery. An Illustrated Handbook. 2011; Thieme Medical Publishers.Pg.17

There are three semicircular canals: the lateral (horizontal), posterior (posterior vertical), and superior (anterior vertical). Each of the three ampullae, and the nonampullated ends of the lateral canal, posterior and superior canals fuse to form the crus commune, which open into the vestibule.

Membranous Labyrinth:

The membranous labyrinth consists of the cochlear duct, the three semicircular ducts with their ampullae, the otolithic organs (the utricle and the saccule), and the endolymphatic duct, housed within the bony labyrinth. The membranous labyrinth is filled with endolymph, with ductus reuniens connecting the major structures, the utricle and sacule.

The cochlear duct or scala media, is an epithelial duct that spirals from the vestibular cecum in the vestibule to the cupular cecum at the apex of the bony cochlea. The epithelium of the floor of the cochlear duct is dominated by the organ of Corti, which rests on the basilar membrane. The inner and outer hair cells are the primary auditory receptors, partially enveloped by the synaptic terminations of cochlear nerve fibers. The spiral ligament is a specialized layer of periosteum, upon which rests the stria vascularis. Reissner's membrane forms the anterior wall, or roof of the cochlear duct, and the tectorial membrane is a gelatinous leaf blanketing the organ of Corti.

The utricle is an elliptical tube and its macula, oriented in the horizontal plane is the sense organ of the utricle. The saccule is a flattened sac, and its macula lies in the spherical recess, predominantly in the vertical plane. The saccule is characterized by a reinforced area, and its endolymphatic space communicates with that of the cochlea by means of the ductus reuniens.

The endolymphatic sac lies about 10 mm posterolateral to the porus of the internal auditory canal in a slight depression called the endolymphatic fossette. The endolymphatic duct runs from the posterior wall of the saccule and joins the utriculosaccular duct, passes along with the vestibular aqueduct and ends in the endolymphatic sac. The cochlear aqueduct is a bony canal that connects the basal turn of the cochlea to subarachnoid space, and the duct parallels the inferior margin of the internal auditory canal. The vestibular aqueduct runs from the vestibule in a transverse direction to the long axis of the petrous temporal bone to the posterior cranial fossa and lodges the endolymphatic duct and sac.

Internal Auditory Canal:

The internal auditory canal is an osseous channel within the petrous part of temporal bone that lies between posterior cranial fossa and the inner ear. It is traversed by the superior and inferior vestibular nerve, along with cochlear, facial, and nervus intermedius, as well as the labyrinthine artery and vein.

Inner ear malformations:

Congenital malformations of the inner ear may be divided into two broad categories: [21]

(a) Malformations with pathologic changes that involve only the membranous labyrinth.

(b) Malformations that involve both the osseous and the membranous labyrinth (malformed otic capsules).

Membranous labyrinth malformations:

It constitutes about 80% of the inner ear malformations. Several types of membranous labyrinth malformation includes, complete membranous labyrinth dysplasia (Bing- Siebenmann malformation), cochleosaccular dysplasia (Scheibe malformation), and cochlear basal turn dysplasia (Alexander dysplasia).

Bony and Membranous Labyrinth Malformations:

It constitutes about 20% of inner ear malformations. The Sennaroglu and Saatci classification of cochlear malformations [22] is widely used and is given in table: 3.2

Table: 3.2

Classification of cochlear malformations

Type of Malformation	Gestational Week of Origin*	Manifestations	Percentage of Patients Affected†
Complete labyrinthine aplasia	3rd	Complete absence of inner ear structures	1
Cochlear aplasia	Late 3rd	Absent cochlea with normal or deformed vestibule and semicircular canals	3
Common cavity	4th	Confluent cochlea and vestibule forming a cystic cavity with no internal architecture; normal or deformed semicircular canals	25
Type I incomplete partition	5th	Cystic cochleovestibular malformation with absence of modiolus; cystic vestibule present but separated from cochlea; figure-eight or snowman-like appearance on axial CT and MR images	6
Cochlear hypoplasia	6th	Small cochlear bud with less than one turn; normal or deformed vestibule and semicircular canals	15
Type II incomplete partition	7th	Cochlea with normal basal turn and cystic apex; strong association with enlarged vestibular aqueduct	50

Source: [22, 23]

Incomplete partition Type III or X linked deafness is a rare type of bony and membranous malformation and constitutes about 2%. In this type, modiolus is deficient and the interscalar septum is partially present [23].

3.3 AUDITORY PATHWAY:

Auditory pathway connects peripheral auditory system to the central nervous system. Afferent auditory neurons are bipolar with central bodies within the spiral ganglion connecting the hair cells to the central auditory system. There are about 12000 outer hair cells and 3500 inner hair cells. Auditory pathway travels through VIIIth nerve via cochlear nucleus, superior olivary complex, lateral lemniscus, inferior colliculus and medial geniculate body to the auditory cortex.

Cochlear nucleus is divided into dorsal and ventral nuclei. Ventral nucleus is further divided into anterior and posterior ventral nuclei. Low frequency vibrations travel ventrally and high frequency towards dorsal nuclei, thus maintaining the tonotopic organization. The dorsal pathway goes directly into the inferior colliculus, while the ventral pathway relays in the ipsilateral and contralateral superior olivary complex from spherical or bushy cells and making the superior olivary complex the first site in auditory pathway where binaural comparisons are made.

Inferior Colliculus receives direct input from the cochlear nucleus via the lateral lemniscus. Main function of the inferior colliculus is understanding about the nature of sound and also involved in auditory motor responses (i.e) stapedial reflex that helps in attenuating loud sounds and hence protecting ear.

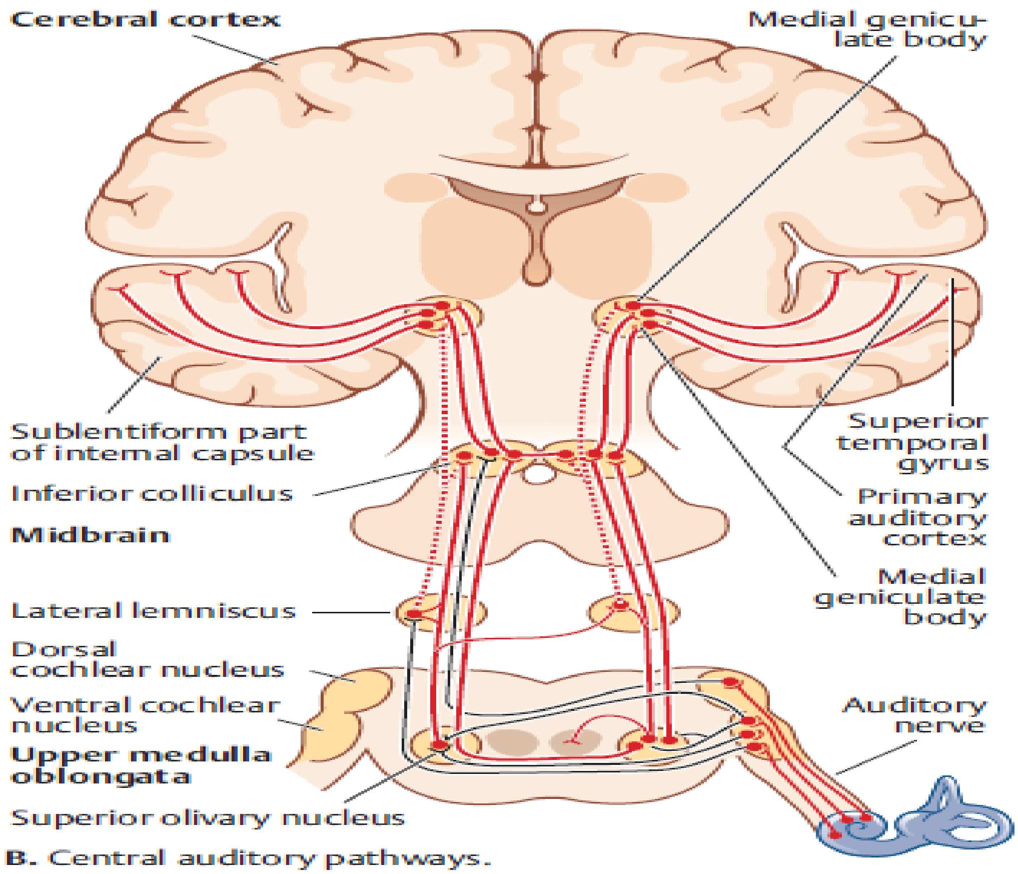
Medial Geniculate body **-** Each geniculate nuclei has three divisions receiving separate pathway from inferior colliculus. The Ventral part receives input from central nucleus of inferior colliculus and is organized tonotopically and the dorsal part receives from dorsal cortex of inferior colliculus and respond to complex sounds, whereas the medial part receives multimodal inputs from lateral cortex of inferior colliculus involving other sensory inputs.

The main auditory portion of the cerebral cortex resides in the temporal lobe, close to the sylvian fissure namely the primary auditory cortex (Area 41) and the association auditory cortex (Area 22 & 42). The primary auditory cortex is tono topically tuned, with high frequencies being represented more medially and low frequencies more laterally and is involved with integrating and processing complex auditory signals, which includes language comprehension while auditory association cortex plays an important role in speech perception.

In addition to the primary and association auditory cortex, auditory information from the subcortical structures also project to other parts of the brain, such as the amygdala, which is a part of the limbic system which explains why sounds such as music can evoke strong emotional responses. The auditory pathway is depicted in Fig 3.4:

Fig 3.4

Auditory Pathway



Source [20]: Adunka OF, Buchman CA. Otology, Neurotology and Lateral Skull Base Surgery. An Illustrated Handbook. 2011; Thieme Medical Publishers.

3.4 CAUSES OF CONGENITAL HEARING LOSS:

Congenital hearing loss can be of genetic (50%) and acquired origin (50%). The genetic hearing loss is divided into syndromic and non-syndromic hearing loss. Non-syndromic hearing loss in the absence of other phenotypic manifestations accounts for about 70% of genetic hearing loss. The causes of congenital hearing loss are given in table 3.3:

Table 3.3

Causes of congenital hearing loss

<table>
<tr><th colspan="2">Hereditary / Genetic [50%]</th><th>Acquired [50%]</th></tr>
<tr><td>Syndromic [30%]</td><td>Non-syndromic [70%]</td><td rowspan="2">Idiopathic
Intrauterine infection
(TORCH)
Low birth weight
Hypoxia
Hyperbilirubinemia
Non-genetic syndromes
Goldenhar’s Syndrome
Fetal Alcohol Syndrome</td></tr>
<tr><td>Autosomal Dominant:
• Waardenburg Syndrome
• Branchio-oto-renal Syndrome
• Stickler’s Syndrome
• Neurofibromatosis Type 2
• Treacher Collins Syndrome
Autosomal Recessive:
• Pendred Syndrome
• Usher’s Syndrome
• Jervell and Lange-Nielson Syndrome
• Biotidinase deficiency
• Refsum’s disease
X-linked:
• Alport Syndrome
• Mohr-Tranebjaerg Syndrome
Mitochondrial Syndromes:
• MELAS Syndrome
• MERRF Syndrome</td><td>Autosomal Dominant:
• DFNA 1 - 64
Autosomal Recessive
• DFNB 1 - 30
X-linked
• DFNX 1 - 4
Mitochondrial:
• MTR NR 1</td></tr>
</table>

3.5 HEARING ASSESSMENT: [24]

The hearing assessment in infants and children is done using both subjective and objective methods and is given in table 3.4:

Table 3.4

Methods of Hearing Assessment

SUBJECTIVE AUDIOMETRY	OBJECTIVE AUDIOMETRY
1. Pure Tone Audiometry (PTA) 2. Behavioural Observation Audiometry (BOA) 3.Visual Reinforcement Audiometry (VRA) 4. Play Audiometry (PA). 5. Impedance Audiometry (IA)	1. Brainstem Evoked Response Audiometry (BERA) 2. Oto-Acoustic Emissions (OAE)

PURE TONE AUDIOMETRY (PTA):

It is a subjective test used to identify the type, degree and configuration of hearing loss. The test is reliable only in adults and older children, as it depends on patient's response to pure tone stimuli. In children aged 3 to 6 years, conditioned audiometry is used.

Test signals are presented by both air and bone conduction or in a sound field with and without masking. The thresholds obtained are used to find the degree and type of hearing loss. The tone is adjusted to a level below the patient's threshold and is lowered in 10 to 15 dB steps until it becomes audible and increased in 5 dB steps till the patient responds. This procedure is repeated till hearing threshold is obtained and known as "up 5 down 10 technique". Pure tone average is the mean of air conduction threshold at 500, 1000, 2000 HZ (Speech frequency).

BEHAVIOURAL OBSERVATION AUDIOMETRY (BOA):

This test involves watching baby's responses to sudden and intense stimulus presented in a sound field such as speech signal, warble tones, narrow band noises, hand-held noisemakers. The responses are in the form of

1) Moro's reflex: Sudden movement of limbs and extension of head in response to loud sounds.
2) Auro-palpebral reflex: Child responds by blink , opening or widening of eyes to a loud sound
3) Auro-oculogyric reflex: Eye shifts in response to the direction of sound
4) Cessation reflex: Infant stops activity or starts crying in response to sound.

VISUAL REINFORCEMENT AUDIOMETRY (VRA):

The hearing measurement is done by using a conditioned localization response and a visual reinforcer. This was described by Suzuki & Obiga in 1961, who called it as conditioned orientation reflex (COR). This test is usually performed after 4 months, as the child turns the head towards sound source and starts listening to music. The test is performed in a room with loudspeakers on both sides with visual reinforcer on top of it. The characteristics of the test are

1) Beginning the test without conditioning (training) trials in which the test tone and reinforcer are paired.
2) An initial test level of 30 dB (which is raised in 20 dB steps if there is no response at 30 dB)
3) The use of an up 10 dB / down 20 dB technique.

Tangible Reinforcement Operant Conditioning Audiometry:

It is done in difficult to test patients due to physical, developmental, perceptual, cognitive emotional and other problems. Upon hearing a tone, child is asked to push a button or to make another simple but specific motor response within his range of neuromotor capabilities. Correct responses are reinforced with tangible reward like cereal, candy.

Visual Reinforcement Operant Conditioning Audiometry:

The child is asked to press a response button instead of turning towards loud speaker, after which a visual reinforcer is presented. The visual reinforcer is the same kind used in VRA. When VROCA is used in sound field testing, the loudspeaker is kept in front of the child so he is not distracted from the response box.

PLAY AUDIOMETRY (PA):

Play audiometry is used to test the hearing of very young toddlers and preschoolers to determine the type and degree of hearing loss. It involves training the child to listen for stimulus and then make a specific motor response within the framework of the game, usually in combination with social reinforcement such as smiles, praises, etc.

The basic game used here is making the child hold a peg up to his ear while listening for a tone, and then placing it into a hole in the pegboard when a tone is heard. Tell and demonstrate what to do and praise them for correct action. Once the child has learned the task, we need to obtain threshold efficiently before habituation occurs. Northern and Downs (1991) recommended starting in 40 dB and descending in 15dB steps, and then to use ascending presentations to find the threshold, using two responses as the criteria. It is desirable to obtain thresholds at 500 and 2000 HZ first for both ears. The other frequencies can be filled as long as the child is in task with 1000 HZ, 250 HZ, 4000 HZ.

IMPEDANCE AUDIOMETRY (IA):

Impedance audiometry comprises of tympanometry, Eustachian tube function tests and Acoustic reflex tests. It is used in objective differentiation between conductive and sensorineeural hearing loss, to measure middle ear pressure and evaluate Eustachian tube function. It is also used to identify the site of lesion in facial paralysis, to identify whether the lesion is cochlear or retro-cochlear in sensorineural deafness and objective estimation of average hearing threshold level.

BRAINSTEM EVOKED RESPONSE AUDIOMETRY (BERA):

Auditory brainstem response audiometry is a neurological test of auditory brainstem function in response to auditory (click) stimuli and first described by Jewett & Williston in 1971. The principle behind BERA is, when sound reaches the cochlea, it is converted into electrical impulses and passes from the cochlea to auditory cortex which is recorded in waveforms of 1-V11, within 10 minnutes of onset of stimuli. These responses are called short latency responses. BERA threshold is defined as the lowest intensity level of sound at which a detectable, repeatable and replicable response is observed in BERA tracing.

BERA TRACING:

Active electrode - Vertex

Reference electrode - mastoid/ear lobe of i/l ear

Ground electrode - forehead just above the nasion or over the c/l mastoid

Sound stimulus - given at fixed suprathreshold level ie at above 60dB.

Sound - broad band clicks of 0.1ms duration.

Stimulus rate - 11.1 clicks/sec.

RECORDING OF ABR:

The recording of ABR were started from 60db intensity and then successively increased or decreased as per identification of wave v peak. Hearing threshold of the patient is the minimum intensity at which the wave v is traced. Multiple recordings are carried out for each intensity and were superimposed, to check the reproducibility of the waves thus obtained. The minimum time taken for ABR is 30min including 10min of patient preparation.

WAVEFORM COMPONENTS:

WAVE I: This response is the representation of compound auditory nerve action potential in the distal portion of V111th nerve as they leave the cochlea and enter the internal auditory canal. It's is a sharp peak beyond 1ms mark on the BERA graph. It should be recognized properly as it gives an idea of stimulus crossed over from the cochlea and distal end of the Eighth nerve.

WAVE II: This nerve is generated in the proximal eighth nerve as it enters the brainstem. It peaks immediately preceding wave III. It should be looked for at just before 3ms mark on BERA tracing.

Wave III: It arises in the cochlear nucleus in the caudal portion of pons. This wave precedes wave 1v.its consistent as wave 1and v. It is identified as upward peak between waves II and IV, just beyond 3ms mark on the graph.

WAVE IV: It arises from the superior olivary complex, but additional contributions may come from cochlear nucleus and nucleus of lateral lemniscus. Identified as a peak just preceding wave V. Distinct and separately identifiable wave present in only 50% of subjects.

WAVE V: It is believed to originate from the vicinity of inferior colliculus or lateral lemniscus. Mostly it is reliable and easily identifiable. Hallmark of wave V is sharp negative deflection immediately following peak, always bigger than wave I (twice the size).

CLINICAL USES OF BERA:

It is used in screening newborn infants, mentally retarded or malingering patients. Used in identifying the nature of deafness, to differentiate central or peripheral disorders, identify the site of lesion in retrocochlear pathologies, can be used in premature infants more than 30 weeks of gestation, used to assess the maturity of CNS in newborns, objective identification of brain death, assessing prognosis in comatose patients.

OTOACOUSTIC EMISSIONS (OAE):

These are low intensity sounds (biological sounds) produced by outer hair cells of normal cochlea. The sound emitted by the normal cochlea can be picked up and measured by the microphone receiver placed in the deep external auditory meatus. There are three types of OAE'S.

Spontaneous OAE’s: These are narrow band sounds emitted from the ear in the absence of stimulation. These emissions occur only in about 50% of normal subjects. The limitations are they are found in different frequencies in different ear, amplitude varies over time, found in relatively restricted range of frequencies.

EVOKED OTOACOUSTIC EMISSIONS:

These are sounds emitted from the ear as a result of stimulation and there are three types of evoked emissions.

a) STIMULUS FREQUENCY OTOACOUSTIC EMISSIONS:

These are elicited by presenting sweep-frequency tone to the ear. Though it provides useful information it cannot be used as a viable clinical tool due to complications in terms of technology and interpretation.

b) TRANSIENT EVOKED OTOACOUSTIC EMISSIONS:

The sound generated by the loud speaker travels via the middle ear in to the cochlea, where the sound energy is processed and the biological sound generated by the OHC reaches EAC through middle ear, which is picked by the microphone and is recorded graphically in a moving strip of paper. TEOAE’S are obtained in all normal individuals including newborns. It is reduced or obliterated in factors causing hearing losses such as ototoxic drugs, hypoxia &noise exposure. It is absent in cochlear sensorineural losses greater than 30 to 50 dB. The interpretations are OAE’s absent if there is defect in middle ear or cochlea.

c) DISORTION PRODUCT OTOACOUSTIC EMISSIONS:

These are produced by presenting two stimulus of different frequencies simultaneously. The lower stimulus tone is f1 and the higher stimulus tone is f2. In response to this stimulus the cochlea will generate a tone of different frequency called as distortion product. This distortion product is transmitted back to the ear canal as otoacoustic emissions. The frequency of DPOAE is 2f1-f2.

SPEECH AUDIOMETRY (SA):

It is used to assess the degree and type of hearing loss, to examine the word recognition abilities, to find the discomfort or tolerance level to speech stimuli and to know the proper gain and the maximum output of amplifying devices.

Speech reception threshold (SRT) is the threshold of a person for speech at the lowest level at which the presence of speech signal can be heard or identified 50%of the time. Speech detection threshold (SDT) is the lowest level at which the presence of speech signal can be detected 50% of the time. SDT is always better than SRT as SDT is just to identify whereas, SRT is to comprehend the speech signal and have to repeat the speech stimulus. The average difference between them is 10 dB. Speech discrimination score is the percentage of number of correct responses to the total number of stimulus presented. Normal score is 100%.

3.6 COCHLEAR IMPLANTS OVERVIEW:

3.6.1 History of Cochlear Implants:

A cochlear implant is an electronic device that converts external sound signals to electrical impulses in place of the defective hair cells and provides information through direct stimulation of the auditory nerve to hearing centers in the brain. [25, 26] Cochlear implants (CI's) are true bionic sense organs that can provide meaningful sound and speech perception to individuals with sensorineural hearing loss.[10] The "Tonotopic organization", also called as "frequency-to-place" mapping of the basilar membrane with in the inner ear is used by the implant, to process the information with different frequencies within our brain. [27]

The impact of the cochlear implants is greater considering its development over brief time. To elicit hearing, the implants had progressed from direct electrical stimulation of the auditory nerve to a commercially available device that has restored varying degrees of hearing to tens of thousands of deaf patients in less than four decades.[28] Many discoveries and new technologies were made from late 18th century to twentieth century that had influenced the rapid and remarkable development of cochlear implants. The time line of the events in the history of cochlear implants is given in the table 3.5:

Table 3.5

Timeline regarding history of cochlear implants [28, 29, 30]

Year	Events
18th Century (1790)	**Alessandro Volta** – First person to stimulate the auditory system electrically with two metal rods inserted into his ears by connecting to a electric battery.
1855	**Duchenne de Boulogne** – Stimulating cochlea with an alternating current rather than direct current
1930	**Wever and Bray** – Electrical potentials in cochlea that closely followed the waveform of the sound stimulus [Wever and Bray effect] **S.S. Stevens and his colleagues** – Described the mechanism by which the cochlear elements respond to electrical stimulation to produce hearing.
1939	**Homer Dudley** – demonstrated a real time voice synthesizer that produced intelligible speech using circuitry designed to extract the fundamental frequency of speech, intensity of spectral components and named as “vocoder”
1950	**Lundberg** – Performed the first direct stimulation of auditory nerve
1956	**Jack Urban and Dr. William House** – designed a workable / wearable implant
1957	**Andre Djourna and Charles Eyries** – First direct electrical

	stimulation of human auditory system. Device fails few weeks after implantation
1961	**Dr. William House** – American Otologist developed the first single channel cochlear implant. Scala tympani insertion of electrodes, advancements in microelectronics, biocompatible materials and microscopic otologic surgery was done
1966	**F. Blair Simmons** – Performed the first temporary human implantation of a multichannel system in the trunk of the auditory nerve itself, in a deaf volunteer.
1967	**Prof. G. M Clark -** created a team to conduct basic research on the pathophysiology of profound deafness in animals and on the tolerability of implanted materials.
1972	**Dr. William House** – builds the first wearable signal processor
1973	**R. Michelson** (UCSF - San Francisco) – Chronically implanted a deaf man with an experimental multichannel implant, using pair of antennas for each channel.
1975	**Ingeborg and Erwin Hochmair -** Started cochlear implant development at the Technical University of Vienna.
1976	**CH Chouard & Bernard Meyer -** Performed first implant at Saint-Antoine hospital, Paris with bulky transmitter.
1977	**Prof. G. M Clark & team -** filed a patent for a system with three

	functional electrodes but using only a limited part of the speech information. **Prof. Kurt Burian -** World's first microelectronic multichannel cochlear implant (8 channels) developed by Ingeborg and Erwin Hochmair was implanted in Vienna.
1978	**Prof. G. M Clark and Dr. Brian Pyman** – Performed first successful cochlear implantation surgery.
1980	US Food and Drug Administration (FDA) began regulation of cochlear implants
1982	**CH Chouard -** Employing the Born reconstruction technique which he had previously used was first to demonstrate in animals, the need for early implantation to avoid the atrophy of the central auditory structures that occurs very rapidly in case of persistent neonatal deafness.
1984	FDA approved the use of cochlear implant in adults
1985	Cochlear Corporation Nucleus 1 system approved by USFDA
1989 - 90	Medical Electronics Corporation (MED-EL) founded by Ingeborg and Erwin Hochmair.
1990	U.S. FDA approved the use of cochlear implant in children above 2 years old

1991 - 92	Multi-channel cochlear implant was developed. It enhanced the spectral perception and speech recognition capabilities compared to single channel devices.
1994	Med-el presented the world's first electrode array capable of stimulating the entire length of the cochlea to allow a more natural hearing.
1996	Advanced Bionics Corporation (AB) implant approved by USFDA
2000	FDA approved the use of cochlear implant for infants above 12 months of age.

3.6.2 Parts of Cochlear Implant:

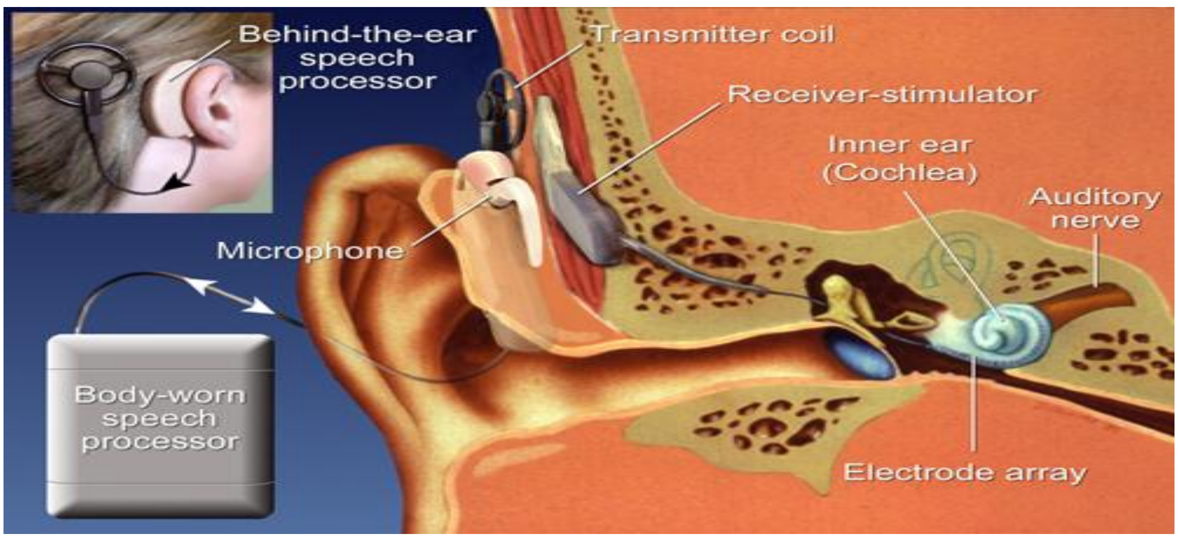
The implant consists of external parts (worn outside by the patient) and internal parts, (surgically implanted) that communicate transcutaneously via radio frequency (RF) signals. [10]

The external parts include a microphone, a speech processor, and a radio transmitter. The microphone collects the sound and converts it into electric signals and then processed by speech processor, through a series of band-pass filters according to the device's speech encoding strategy. After processing, the transmitter sends the coded signals to the receiver transcutaneously. The transmission coil is held by the magnets at the center of each internal and external coil in place over the receiver. [31]

The internal part consists of a receiver – stimulator system, secured in skull bone behind the ear and an array of multiple (12 – 22) electrodes. The receiver – stimulator system converts the signals into a series of bipolar square-wave signals and delivers to the electrode array in cochlea. The electrodes stimulate the fibers of the auditory nerve to send information to brain, where it is interpreted as a meaningful sound. [31] The parts of cochlear implant are shown in Fig 3.5:

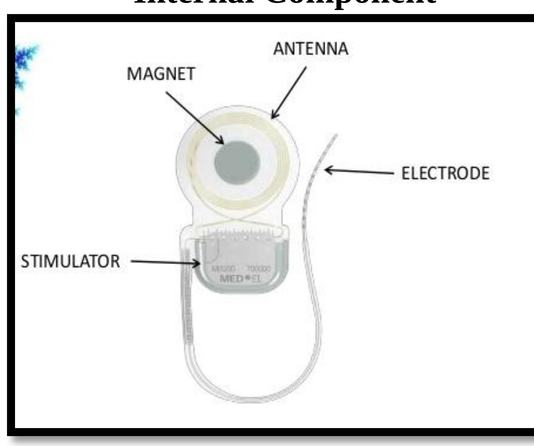
Fig 3.5

Parts of Cochlear implant



External Component

Internal Component



Source: https://www.boystownhospital.org/knowledgeCenter/articles/hearing/Pages/howaCochlearImplantWorks.aspx

3.6.3 Coding Strategies:

Coding strategies are defined by which the pitch of sound, loudness and timing of sound are translated into a series of electrical impulses. Two type of strategies are used - Simultaneous and Non simultaneous.

a) Simultaneous strategies:

This strategy permits the activation of more than one electrode at a time, thus providing a better speech outcome and natural quality of sound. The only disadvantage is the channel interaction.

b) Non simultaneous strategies:

The strategies are Continued Interval Sampling (CIS), Advanced Combination Encoder (ACE), Spectral PEAK (SPEAK). CIS stimulates each electrode serially, one after the other. Each electrode is stimulated by means of different frequency and the cochlea receives complete information about the frequency composition of incoming signals. The rapidity with which stimulation occurs leads to improved speech recognition.

3.6.4 Evaluation for Cochlear Implant Surgery:

The evaluation of a child with sensorineural hearing loss for cochlear implant surgery is done by a team of experts, comprising of otolaryngologist, audiologist, radiologist, pediatric neurologist, clinical psychologist and speech pathologist. Initially, the otolaryngologist examines the middle and inner ear to ensure that no active infection or presence of any other abnormality. The audiologist performs the hearing assessment tests to find out the severity of hearing loss and also performs pre-implant assessment and counseling for prospective clients. Radiology imaging like High Resolution Computerized Tomography (HRCT) of temporal bone and Magnetic Resonance Imaging (MRI) brain and inner ear are done to assess the morphologic status of the ear and the brain. Basic laboratory investigations are also done with screening of TORCH infections. Anaesthetists will do a complete physical examination and analysis of investigations to identify any potential problems with the general anesthesia. Opinion from pediatric neurologist and clinical psychologist regarding fitness for cochlear implant surgery is also taken.

3.6.5 Cochlear Implant Surgery:

The basic principles and major goals of cochlear implant surgery are: [31]

1. To insert the electrode array without trauma, as far as possible into the scala tympani
2. To place the device on the side of the head in a manner that, it protests from trauma
3. To ensure that the device and electrode array are secure enough to prevent movement.

The surgery is performed under general anaesthesia and lasts for about 2 – 4 hours. Under aseptic precautions, a post auricular incision of 4 – 5 cm made extending superiorly or posteriorly according to surgeon’s preference. The vertically oriented incision is least likely to impair blood flow into the skin flaps, thus preventing wound gaping. After skin incision, T shaped periosteal incision made with superior border paralleling the temporalis muscle and descending limb extending to the tip of mastoid. Mastoid cortex is exposed, well is created for receiver - stimulator in the flat portion of the skull far posterior to the post auricular incision, so that it does not under lie the ear level processor.

A template is used as guidance and temporal squama is drilled to a depth of 2 – 3 mm to accommodate the receiver / stimulator. Drill holes are made superior and inferior to the stimulator and sutures are passed through the holes to secure the stimulator in position. Cortical Mastoidectomy is done exposing middle fossa dural plate, sigmoid sinus and sinodural angle. Cavity should not be saucerized and edges are left as acute as possible to retain electrodes within the confines of mastoid cavity. Facial recess is then widely opened to visualize the stapes tendon and round window.

The inferior portion of facial recess is necessary in identifying round window niche and superior portion is used to achieve an appropriate angle of insertion. After visualization of round window niche, posterior bony overhang of niche is removed using small 1mm burrs until round window membrane is seen. Round window membrane is always darker, much shiner and appears to be tightly stretched. Round window membrane is an extension of the endosteum of scala tympani and identification of round window membrane identifies scala tympani unequivocally.

The size of cochleostomy varies between 0.6 – 1.2 mm. Round window cochleostomy is commonly used and is important to ensure that tip of electrode array is directed anteriorly and inferiorly to avoid contact with modiolus. The electrode insertion is advanced slowly and smoothly, and stopped at first resistance. By gentle twisting movements the electrode is allowed to pass the resistance and go deeper into scala tympani. Sealing the cochleostomy site with soft tissue prevents electrode extrusion and ascending infection leading to meningitis. Closure is done in all three layers, deeper layer closure should completely cover the receiver stimulator and electrode leads. Post-operative care is given as per protocols.

3.6.6 Complications of Surgery:

Cochlear implant surgery is a safe and effective procedure with low complication rates. World literature had reported major complication rates of about 2 – 6 %. [32-35] The early complications associated with the surgery are poor electrode placement, infections, Chorda tympani and Facial nerve injury, CSF leak, damage to electrodes and haemorrhage. Late complications reported are receiver-stimulator extrusion, device failure, electrode migration and facial nerve stimulation.

3.7 INTRAOPERATIVE MONITORING:

Intra-operative monitoring is used to confirm the integrity of the implant and the electrical output of the device. It also gives immediate feedback to the operating surgeon, audiologist and the family members about the success of the procedure.[36] Intraoperative monitoring during surgery is used as a guiding factor to assess the functioning of device and correct placement of the electrode array [37]. The success or failure of cochlear implant patients depends on the transfer of stimulating signals from the electrode to the auditory nerve fibers. Intraoperative monitoring is done by electrophysiological testing of the device and radiology imaging.

ELECTROPHYSIOLOGICAL TESTING (TELEMETRY):

"Telemetry is used as an important measure to check the normal functioning of the external and internal components. It provides communication between external component and the receiver-stimulator to detect any electrode failure during and after implantation." [38, 39] Various testing measures used in telemetry are:

1. Electrode Impedance (EI)
2. Electrically Evoked Stapedial Reflex Telemetry (ESRT)
3. Electrically Evoked Compound Action Potential (ECAP)
4. Spread of Excitation (SOE)

ELECTRODE IMPEDANCE (EI):

Electrode impedance is a major aspect of the electrode design, which in turn depends on the electrode surface area, its morphology, and the signals initiated by electrical stimulation. [40] Impedance is concerned with power consumption and reducing the amount of power will lead to further minimization of the processors. [41] Electrode impedance is derived by measuring the voltage drop across an electrode for a given electrical signal. [38] It implies the function of electrode integrity, like short or open circuits and resistive characters of fluid and tissues surrounding the electrode. [42]

Short circuits indicate low impedance values due to cochlear or common cavity malformations, excess solution on the mastoid cavity. A reduction in electrode impedance might be due to formation of a hydride layer on the surface of the electrodes, by increasing the surface area. [43] Open circuits indicate high impedance due to air bubble around the electrode, insertion or extrusion of the electrode (partial insertion) and rarely damaged or faulty electrode. Presence of tissue and/or bone growth near the electrode array may lead to high impedance, displaying positive correlation. The open or short circuits should be solved by repositioning the electrodes or replacing them before closing the cavity. [38]

In most of the cases, abnormal impedance usually settles in 5-10 minutes .The low incidence of abnormal impedance was due to associated factors like post meningitis, partial insertion, explant/re-implant etc. [38]

Normal impedance telemetry has also been recorded when the electrode array is placed in the carotid, superior semicircular canal or internal auditory canal, thereby concluding that electrode impedance does not confirm the electrode placement and it does not replace imaging studies after the implant. [44]

ELECTRICALLY ELICITED STAPEDIAL REFLEX TELEMETRY (ESRT):

The Stapedial reflex is an autonomic response to loud sound that results in reflexive contraction of the Stapedius muscle. ESRT is used as a guide for postoperative comfort level settings during initial cochlear implant programming in children and difficult to test patients. ESRT requires intact acoustic reflex pathway and functional device but it not used in testing device function or electrode position.

NEURAL RESPONSE TELEMETRY (NRT):

The Neural Response Telemetry (NRT) / Neural Response Imaging (NRI) or Auditory Response Telemetry (ART) as described by different manufacturers is used as a tool for recording auditory nerve responses to electrical stimulation. The neural responses can be measured electrically by, evoked compound action potentials (ECAP), evoked stapedial reflex (ESRT), evoked auditory brainstem response (EABR) and electrically evoked middle latency responses.

EVOKED COMPOUND ACTION POTENTIAL (ECAP):

Electrically evoked compound action potentials (ECAP) is one of the most commonly used measure by the recent cochlear implant systems with their inbuilt hardware and softwares. "Compound action potential is a synchronous response due to electrical stimulation of cochlear nerve fibres, and it is the electrical version of the Wave 1 of the acoustically stimulated auditory potentials of the brainstem. [45] The response of the auditory nerve resulting from a stimulus presented at one location is recorded from a neighboring location within the cochlea. [46]

ECAP recordings can be done intraoperatively and postoperatively and will have a negative peak (N1) followed by a positive peak (P1) with approximate latency of 0.2 – 0.4 ms. [47] Intra-operatively, it is used to assess the auditory nerve integrity and post-operatively used to fit the sound processing system and monitor the recipient progress.

Fitting the speech processor relies on the determination of the ‘threshold’ (T) and ‘comfort’ (C) levels. Postoperative ECAP measurements will be helpful in device programming by providing threshold values for initial MAP creation, especially in very young children and other difficult-to-program patient populations. [48]

The ECAP values have no relation to age at presentation, etiology of deafness, duration of hearing loss, hearing aid use duration and preoperative PTA values. Postoperative absence of NRT responses do not have any correlation with open-set word discrimination, suggesting that lack of measurable ECAP does not necessarily indicate a lack of auditory response to electrical stimulation or a dysfunctional device. [48]

ECAP responses done immediately after implantation, where the patient is still under anesthesia, high stimulation levels can be applied resulting in high success rate. But whereas in post-operative setting, high stimulation levels might cause some discomfort by exceeding the patient’s subjectively determined acceptable sound level, leading to lower recording of ECAP responses.

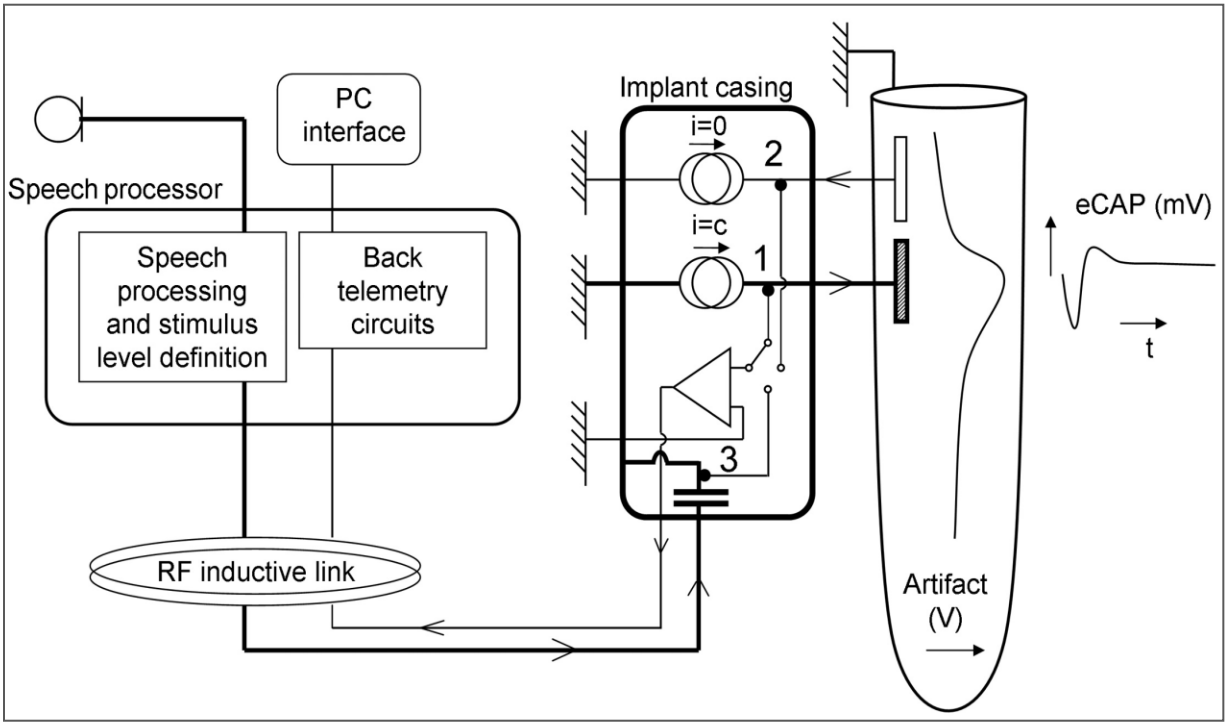
SPREAD OF EXCITATION (SOE):

Spread of Excitation (SOE) is used to detect the tip rollover of the electrode array and provide information regarding the selectivity of neural excitation fields around each electrode. [37] When tip roll over occurs, two portions of cochlea are affected. The area distal to fold over is deprived of stimulation, but the proximal region receives competing stimulation from multiple electrodes. It is this channel interaction in the area of electrode fold over that is detected by SOE.

The working of cochlear implant system is given in Fig 3.6:

Fig 3.6

Cochlear implant system



Block diagram of a cochlear implant system. The microphone signal is sent to the external speech processor. Through an inductive radio frequency link, energy and instructions are coupled to the implant. The bold lines indicate the power and signal path. The implanted electronics are capable of sensing voltages at different points of the circuitry and signal the status of the system to the speech processor and, if connected, the fitting station. Voltages sensed within the active output circuits (1) show open circuits. Voltages on nonstimulated electrodes (2) show short circuits, how the electrical field generated by the stimulus itself spreads across scala tympani, and the electrically elicited compound action potential (eCAP). The adjustment of the energy on the RF link is carried out by monitoring the supply voltage available to the current sources (3). In actual implant systems, the number of current sources varies between 1 and 16 and the number of intracochlear electrodes between 12 and 22.
Source: [39]

RADIOGRAPHY IMAGING:

Intra operative imaging is used to provide confirmation of electrode array placement inside the cochlea. The modalities used are x-ray imaging, fluoroscopy and

CT scan. Plain radiography by modified Stenver's method is safe and a reliable tool for assessing the electrode location, position and presence of tip roll over. [37, 48, 49]

Intra operative CT scan is very useful to confirm electrode position in cochlear malformations. If any malposition of electrodes is present, it can be corrected under the same sitting, as the patient is under anaesthesia.

Fluroscopy is unique in assessing two dimensional planar visualization ofelectrode position during insertion. In cases of abnormal cochlear anatomy, intraoperative fluoroscopy is advocated as a dynamic ''real-time'' assessment of electrode placement.[37]

Immediate intraoperative determination of device functionality and optimal electrode placement using radiology imaging is advantageous. Among the intraoperative testing modalities like electrode impedance, tNRT, and plain radiograph, the radiographic results had an impact on the intraoperative surgical decision making and led to the use of the backup device. [37]

POST OPERATIVE PROGRAMMING:

The initial stimulation [Switch ON] of the cochlear implant starts, 3 – 6 weeks after surgery by the rehabilitation team using computer aided software. The electrode impedance is checked in all the electrodes. The Auditory Response Telemetry (ART) measurement is also done. Mapping is done initially and in subsequent sessions periodically to determine the threshold levels, comfortable levels for each electrode's and for balancing the parameters. The data is stored in the speech processor. Audiology assessment, speech and language perception is also tested at regular intervals.

The participation of parents and family members is of paramount importance in the training of the children after implantation. Training is given to parents in creating listening environment, language modeling and enabling daily routines to train the child at home. Post-operative programming is a team work and needs technical support of all the persons, involved in the rehabilitation of the children.

3.8 ASSESSMENT OF AUDITORY PERFORMANCE:

To assess the auditory performance, Pure Tone Audiometry (PTA) is reliable and a direct measure, but cannot be used in very young children. Therefore indirect measures like Categories of Auditory Performance (CAP) score, speech and language development scores are used.

Categories of Auditory Performance (CAP) Score:

The Categories of Auditory Performance (CAP) score [50] is a categorical, nonlinear scale that ranges from 0 to 7. The levels are assessed by the ability to perform every-day auditory tasks, with 0 being no awareness of environmental noise and 7 representing the ability to use telephone. [10] It is the only supraliminal auditory receptive outcome measure applicable to all children irrespective of the age. [12] The Categories of Auditory Performance (CAP) score used in the study is given in table 3.6:

Table 3.6

Categories of Auditory Performance (CAP) score [50, 51]

Category	Criteria	Working definition
7	Uses telephone	The child can sustain a simple unscripted conversation on the telephone with a familiar talker.
6	Understands conversation	The child can carry out a simple unscripted conversation with a familiar talker (e.g. a parent or teacher) without lip reading in a quiet setting.
5	Understands phrases	Child is able to identify common phrases in a familiar constraining context without lip reading
4	Discrimination of sounds	Child discriminate consistently any combination of two of Ling's five sounds presented at conversational level without lip reading
3	Identifies environmental sounds	Child identifies environmental sounds via audition
2	Response to speech sounds	Child responds to simple commands delivered in a normal conversational sound level at 1 – 2 feet distance
1	Awareness of environmental sounds	The child detects environmental sounds
0	No awareness to environmental sounds	Child does not respond spontaneously to any environmental sounds

3.9 FACTORS AFFECTING THE OUTCOME:

The outcome of cochlear implantation depends on several factors. Many factors like age at onset of deafness, duration of deafness, presence of multiple disabilities, age at implantation, implant technology, implant use duration, preoperative level of residual hearing, social and rehabilitative factors may influence the better outcomes in implantation. The factors affecting the outcome in cochlear implantation are given in table 3.7:

Table 3.7

Variables affecting cochlear implantation performance [52]

1. Cochlear implant technology a. Processing strategy b. Electrode design c. Device reliability	6. Preoperative function: a. Hearing level and speech performance
2. Neuronal cell physiology and function a. Age at implantation b. Duration of deafness/auditory deprivation c. Auditory neuroplasticity d. Auditory pathway development	7. Education/rehabilitative environment a. Mode of communication b. Education and postimplantation rehabilitation services 8. Auditory training
3. Binaural hearing 4. Multiple disabilities a. Autism b. Auditory neuropathy/ auditory dysynchrony	9. Social factors a. Socioeconomic status b. Parent/family expectations and motivation
5. Medical/surgical issues a. Anatomic abnormalities b. Meningitis c. CHARGE syndrome (Coloboma, central nervous system anomalies, heart defects, atresia of the choanae, Growth retardation and/or development, ear anomalies and/or deafness)	

3.10 REVIEW OF LITERATURE

Spivak and Chute et al (1994) [53] have shown that post-operative objective electrophysiological tests like electrically elicited acoustic reflex threshold correlate well with behavioural comfort levels and these measurements may provide valuable information regarding programming the cochlear implant.

Ponton et al. (1996) [54] studied auditory evoked potentials (AEPs) in children with cochlear implants. The authors concluded that (1) the auditory cortex does not develop without stimulation; (2) the plasticity of the cortical auditory system is maintained during sensory deprivation; and (3) maturation of the auditory system begins upon initiation of stimulation.

Archbold et al (1998) [51] had undertaken an inter-user reliability of Categories of Auditory Performance (CAP) score among 23 children followed up at various intervals after implantation.. It provides one means of rating and presenting information that is quick and easy to use, and illustrates progress in a group of children with a wide range of achievement over a long period of time. Also the study had demonstrated its repeatability, with an extremely high degree of agreement between users, attesting to the robustness of the measure.

Nikolopoulos et al. (1999) [55] had done a prospective trial consisting of 126 prelingually deaf children, implanted before the age of 7 years and followed up for four years. Categories of Auditory Performance (CAP) Score is used to assess the auditory performance. The study results showed that speech and language outcomes were negatively correlated with age at implantation, strongly in favour of implantation at young age. However, the author noted that large variations exists between individual children and so age of implantation alone should not be used as a predictive criteria for cochlear implant surgery.

Osberger et al (2000) [56] and **Kirk et al (2002)** [57] performed longitudinal studies among prelingually deaf children, using different scales like Glendonald Auditory Screening Procedure (GASP) and Reynell Developmental Language Scales (RDLS) for assessing auditory and language performance respectively. They concluded that early age of implantation had better outcomes compared to children implanted at later age group.

Govaerts et al (2002) [58] evaluated the outcome of cochlear implantation in young children less than 6 years in relation to age of implantation using CAP score. For all the children the CAP score increased after implantation and intervention before the age of 2 years resulted in early normalization of CAP scores, with 90% probability of integration into mainstream school system.

Eggermont and Ponton et al (2003) [59] studied auditory system maturation by means of auditory evoked potential recordings compared to maturation of axon neurofilaments and some critical stages in speech perception. The parallels strongly suggest that the emergence of the N1 component reflects the maturation of the axons in layer II and upper layer III of the auditory cortex. The absence of N1 in cochlear implant subjects who have been deaf for a period of at least 3 years below the age of 6 years suggests a critical period in the maturation of the upper cortical layers and potentially poor future performance in the perception of masked and degraded speech.

Teoh et al (2004) [60] studied that ultrastructural organization of auditory brainstem is affected by prolonged deafness and further urged the use of aural based therapies with hearing aids prior to implantation and aural-only rehabilitation after cochlear implantation to protect the auditory plasticity.

Mason et al (2004) [61] studied the electrophysiology and objective monitoring of cochlear implant during surgery and showed that normal intra-operative findings provided immediate reassurance to the implant team and parents of young children that the implant was fully functioning and that electrical stimulation was activating the auditory pathways

Miyamoto et al. (2005) [62] explored the benefits of early intervention with cochlear implants in children less than 12 months of age. They used two new behavioural procedures for evaluation of infant speech perception abilities, both based on the time an infant spends looking at an object presented in association with a sound and reported that early implanted infants were capable of forming word-object associations.

Sharma et al (2005) [63] examined the longitudinal development of the cortical auditory evoked potential (CAEP) in 21 children with cochlear implants fitted either before age 3.5 years or after age 7 years. Early-implanted children by age 3.5 years showed rapid development in CAEP waveform morphology and P1 latency. Late-implanted children showed aberrant waveform morphology and significantly slower decreases in P1 latency postimplantation.

Kameswaran et al (2006) [64] assessed the outcome of cochlear implant in 100 patients using Category of Auditory Performance (CAP) scores and Speech Intellegibility Rating (SIR) scores. The study concluded that early cochlear implantation tends to yield normalization of audiophonologic parameters, which enables us to consider the performance of children implanted very early as being similar to that of their normally hearing peers.

Dorman et al, (2007) [65] did a study in larger population of 245 children less than seven years with cochlear implant by dividing in to three groups. Children who were implanted by the age of 3.5 years obtained normal P1 latencies, usually within 3–6 months after initial stimulation. Approximately half of the children implanted between ages 3.5 and 7 years, had decreased P1 latencies compared with normal children at same age. However, children who had experienced more than 7 years of auditory deprivation had an elevated P1 latency, even in measurements taken years after onset of stimulation. They hypothesized that, the P1 response in late implanted children is generated from a different source as compared to normal hearing and early-implanted children, thereby emphasizing better outcome in early implantation.

Kral et al (2007) [66] addressed the issue of sensitive periods and cross-modal reorganization for auditory development and concluded that sensitive period exists between 2nd and 4th year of life. Cross modal reorganization acts as a barrier for processing signals, resulting in higher-order auditory areas occupied by visual functions. In the absence of aural stimulation, descending modulation from higher auditory areas is also decoupled from primary areas in the brain. The study implies that, the neuroplasticity of the auditory system decline sharply after 4 years and thereafter the benefits of cochlear implantation are reduced greatly.

Basta et al in 2007 [67] suggested that strong, intact and possibly synchronized functional correlation between the activity of lower (Cochlear Nerve) and higher (auditory pathway of the CNS) auditory structures (measured by evoked compound action potential) is required to facilitate an improved speech performance after cochlear implantation.

Holt et al (2008) [68] conducted a study with 96 prelingually deaf children who were implanted before the age of 4 yrs and were stratified into four groups based on age at implantation. He proved that age at implantation did not significantly influence the rate of the word recognition development, but did influence the rate of both receptive and expressive language acquisition. Children implanted earlier in life had faster rates of spoken language acquisition than children implanted later in life.

Cosetti et al (2010) [48] did a retrospective review to determine whether intraoperative neural response telemetry (tNRT) is predictive of postoperative speech perception in children and adults. This study suggested that there is no significant correlation between intraoperative tNRT and speech perception performance at 1 year. At the time of surgery, tNRT provides valuable information regarding the electrical output of the implant and the response of the auditory system to electrical stimulation and preliminary device programming data. Furthermore, the absence of tNRT does not necessarily indicate a lack of stimulation.

Forli et al (2011) [69] did a systematic review on the clinical effectiveness of cochlear implantation in children. The authors identified seven studies comparing post-CI outcomes in children implanted within the first year of life with those of children implanted after one year of age. The findings in these studies suggested improvements in hearing and communicative outcomes in children receiving implants prior to one year of age.

Cosetti et al (2012) [37] generated an evidence-based algorithm for the use of intraoperative testing during cochlear implantation to assess device functionality and electrode placement. Intraoperative testing included the individual electrode impedance measurements; neural response telemetry (tNRT) levels for electrodes E20, E15, E10, and E5; and plain film radiograph assessment of electrode position and found to be normal in majority of the patients.

Goehring et al in 2013 [70] had done a retrospective analysis of 303 cochlear implants electrode impedance data, from 2004 to 2011. His objective was to assess the intraoperative impedance abnormalities which resolve by initial activation and normal intraoperative impedances that present as abnormal at the initial activation. The study results showed that, 82% of abnormal intraoperative impedance resolved by initial activation and 0.17% of normal intraoperative electrode impedance were abnormal postoperatively. Intraoperative impedance testing has significant clinical value, showing whether the device is functioning appropriately and also provides notice of potential issues that can be managed postoperatively. Finally, intraoperative device testing provides a baseline for device function over time.

Zhou et al (2013) [71] assessed the auditory performance and speech perception of 34 children with profound sensorineural hearing loss. The CAP and SIR scores of both groups increased with increasing time of implant use during the follow-up period, and at each time point, the median scores of the two groups were comparable. The study results indicate that great communication benefits achieved by early implantation (< 18 months) and exemplify the importance of enhanced social environments provided by everyday life experience for human brain development and reassure the parents regarding cochlear implant surgery.

Shennawy et al (2015) [38] conducted a cross sectional study on cochlear implant recipients, to monitor changes in impedance telemetry and evoked compound action potentials (ECAP) measured during surgery versus the same measures at post implant follow-up visits. The study also correlated the recorded ECAP measures with the patient's postoperative performance and to evaluate any abnormal telemetry measurements and their changes at device activation. The authors' found that, in the absence of electrical stimulation, there was an increase in impedance on all electrodes at the initial stimulation, which decreased at the 9 – 12 months follow-up. No correlation existed between ECAP thresholds and post-operative patient performance at the one-year follow up, thereby concluding that telemetry provides valuable information regarding the electrode integrity and the neural responsiveness of the auditory nerve to electrical stimulation

4. MATERIAL AND METHODS

4.1 STUDY DESIGN:

- Prospective study

4.2 STUDY SETTING:

- The study was conducted at Upgraded Institute of Otorhinolaryngology, Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai – 600003 and at Institute of Speech and Hearing, Rajiv Gandhi Government General Hospital, Chennai – 600003.

4.3 STUDY PERIOD:

- The study was conducted during March 2016 to August 2017, over a period of 18 months.

4.4 STUDY SUBJECTS:

- Children admitted for cochlear implant surgery during the study period at Upgraded Institute of Otorhinolaryngology, Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai, who satisfy the inclusion criteria

4.4.1 INCLUSION CRITERIA:

- Children aged between one to six years
- Intact cochlear nerve and patent cochlea

- Bilateral profound sensory-neural hearing loss with PTA > 90dB in better ear
- No appreciable benefit with hearing aids
- No medical or anatomical contraindications
- Motivated parents for cochlear implant surgery

4.4.2 EXCLUSION CRITERIA:

- Children more than 6 years of age.
- Children with cochlear nerve aplasia and massive cochlear ossification
- Deafness due to lesions in central auditory pathway
- Children with mental and behavioural disorders

4.5 SAMPLING AND SAMPLE SIZE:

- Non probability (Convenient) sampling was used for the study. The children admitted for cochlear implant surgery at the institute was selected consequently, who satisfy the inclusion criteria
- A total of 70 children were included in the study initially and after lost to follow up, 63 children were included for analysis.

4.6 DATA COLLECTION:

- The data was collected from the study subjects using semi-structured questionnaire / case proforma.
- Clinical examination and other relevant investigations were done before and after the surgery.

- ❖ Complete audiological examination like BERA, OAE, BOA, Impedance was done before the surgery.
- ❖ **Impedance measurement:**

  Intra-operative:

  - Impedance was measured during surgery on all electrodes after electrode insertion and before the electrically evoked compound action potential was measured. Impedance was measured using the manufacturers default modes

  Post-operative:

  - Impedance was measured at the beginning of the initial activation, before any electrophysiological or behavioural measurements, and then before each speech programming appointment.

- ❖ **Auditory Response Telemetry (ART) Measurement:**

  Intra-operative:

  - Evoked Compound Action Potentials (ECAP) was recorded from all 12 electrodes intra-operatively.

  Post-operative:

  - Evoked Compound Action Potentials (ECAP) was recorded post-operatively at initial switch on, 3 – 6 weeks after surgery.

All the intra-operative ART measurements were recorded with the clinic's speech processor, whereas the post-operative measures were recorded with the patient's own speech processor.

- **Categories of Auditory Performance (CAP) score:**

  - Auditory performance of the children was assessed using Categories of Auditory Performance (CAP) Score.

  - CAP score was assessed before the surgery and follow assessment was done at 3, 6 and 12 months after the cochlear implant surgery.

4.7 DATA ANALYSIS:

- The data collected in the case proforma was entered into Microsoft excel sheet and the data analysis was done by using Statistical Package for the Social Sciences (SPSS) software version 18.
- Quantitative data was expressed in mean and Standard deviation, whereas the qualitative data is expressed in proportions.
- Association between the study variables and outcome measures was done using appropriate statistical methods. Mann Whitney U test and ANOVA for more than two groups was used to test the association. Wilcoxon Signed Rank Test was used for pre and post test comparisons.
- P value of less than 0.05 is taken as the level of significance.[$p < 0.05$]

4.8 ETHICAL CONCERNS:

- The study protocol was approved by the Institutional Ethics Committee (IEC) and after that only, the study was started.
- Informed Written Consent was obtained from the parents or guardians of the study children in local language, after explaining about the purpose of the study.
- The information collected was used, only for the study purpose and strict confidentiality was maintained throughout the study.

5. RESULTS

In this study, a total of 70 children were enrolled after satisfying the inclusion criteria and were followed up for 12 months. At the end of study, 63 children completed the follow up and seven children were lost due to follow up at different intervals. The response rate was 90% and loss to follow up was 10%. The children lost to follow up were excluded from final data analysis and the results of sixty three (63) children were given here.

5.1 AGE GROUP DISTRIBUTION

The age group distribution of the study participants is given in Table 5.1:

Table 5.1

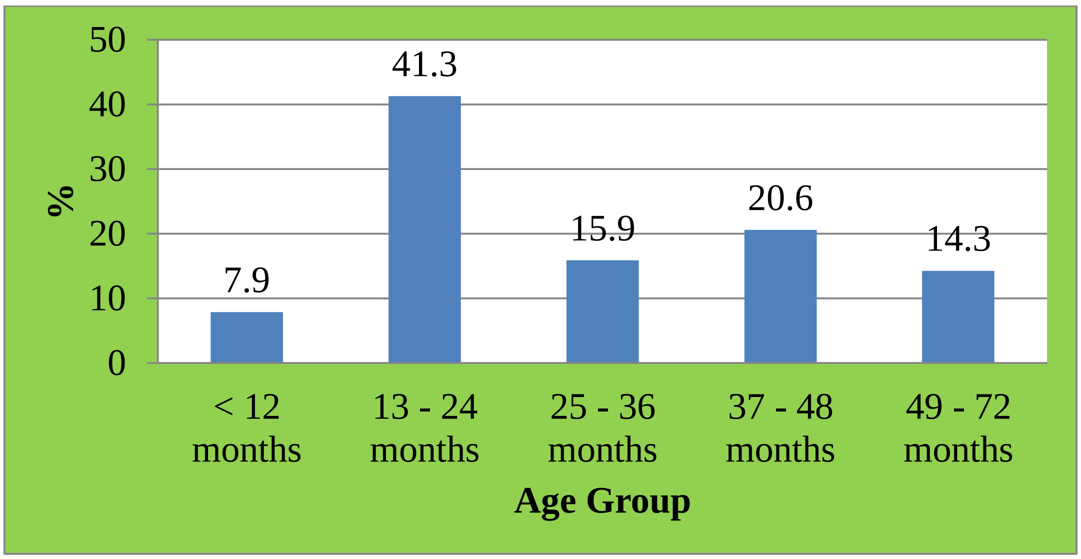
Percentage distribution of study participants by age group

S. No	Age Group	Number	Percentage
1.	≤ 12 months	5	7.9
2.	13 - 24 months	26	41.3
3.	25 – 36 months	10	15.9
4.	37 – 48 months	13	20.6
5.	49 – 72 months	9	14.3
Total		63	100
Mean Age [Mean ± SD]		34.00 ± 15.54 months	

Among the study children, Majority 26 (41.3%) was in 13 – 24 months age group, followed by 37 – 48 months age group (20.6%). About, 10 (15.9%) were 25 – 36 months old and 14.3% belong to 5 – 6 years category. Around 8% were infants. The mean age of the study children was 34.00 ± 15.54 months and the median age was 30 months. [Range = 12 – 66 months]

Fig 5.1

Percentage distribution of study participants by age group



5.2 GENDER DISTRIBUTION

Regarding the gender distribution of participants, male children were 57.1% and female children were 42.9 %. The gender wise details, is given in table 5.2:

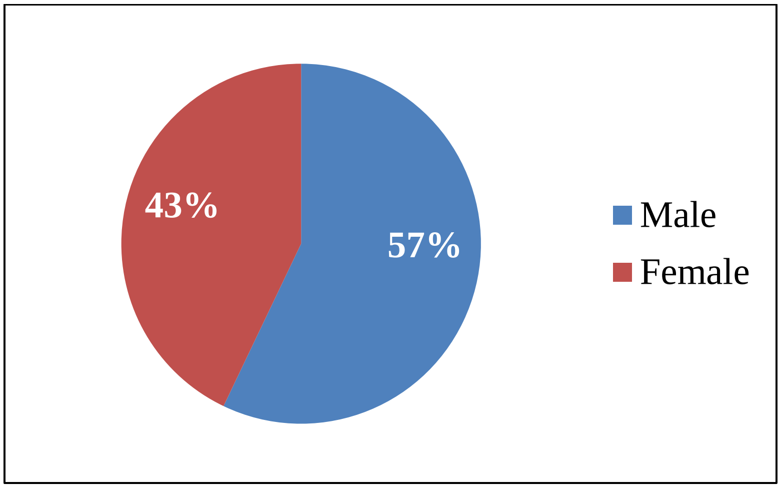
Table 5.2

Percentage distribution of study children by Gender

Gender	Number	Percentage
Male	36	57.1
Female	27	42.9
Total	63	100

Fig 5.2

Percentage distribution of study children by gender



5.3 RELIGION WISE DISTRIBUTION

Majority (92.1%) of the study children were Hindus, followed by Muslims (4.8%) and Christians (3.2%). The distribution of participants according to religion wise is depicted in table 5.3:

Fig 5.3

Study subjects by religion wise distribution

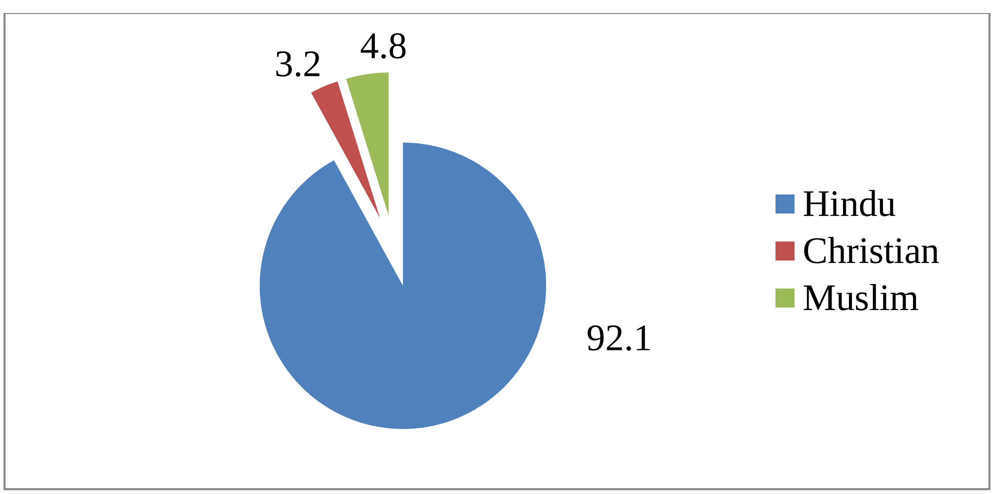


Table 5.3

Distribution of study participants by Religion

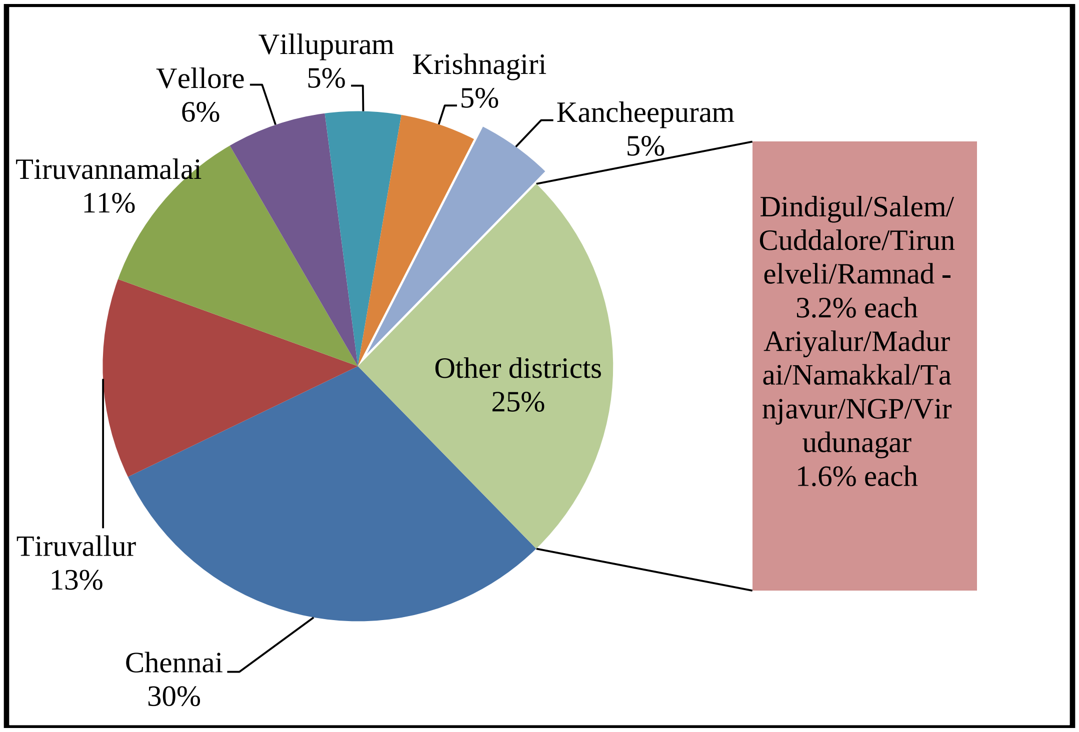
Religion	Number	Percentage
Hindu	58	92.1
Christian	02	3.2
Muslim	03	4.8
Total	63	100

5.4 DISTRICT OF ORIGIN

Among the study participants, nearly 70% of the children were from the northern districts of Tamil Nadu. A total of 19 children (30.2%) were from Chennai, 8 (12.7%) from Tiruvallur, 11.1% from Tiruvannamalai and 6.3% from Vellore. About 15% children were from Villupuram, Krishnagiri, and Kancheepuram districts. The rest of the children (25.4%) were from other districts like Dindigul, Ramnad, Cuddalore, Salem, Tirunelveli, Madurai, Ariyalur, Pudukottai, Tanjavur, Namakkal and Nagapattinam districts. The district of origin of study participants is depicted in the Fig 5.4

Fig 5.4

Percentage distribution of study participants by district of origin



5.5 EDUCATIONAL STATUS OF PARENTS:

Regarding the educational status of the parents, majority of the father's 28 (44.4%) were Diploma / Degree holders, followed by higher secondary schooling (30.2 %). About 15.9% have studied up to high school.

Regarding educational status of mother, 19 (30.2%) had studied up to higher secondary and 11(17.5%) had diploma / degree. About 14.3% of the mother's went to primary school and high school respectively. Among mothers, 14.3% had no formal schooling. The educational status of the subjects is showed in table 5.4:

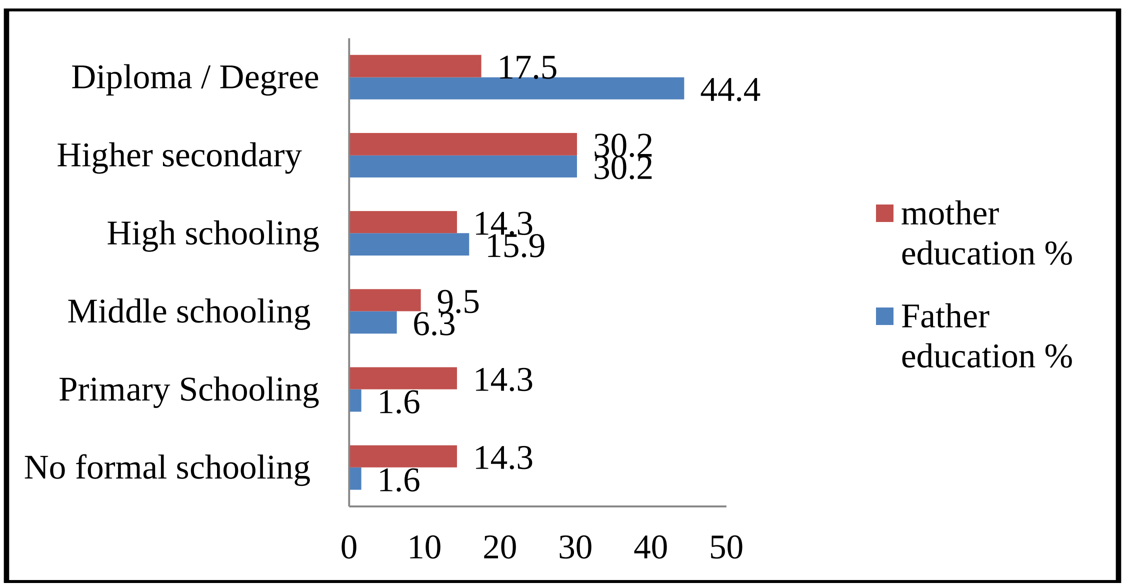
Table No: 5.4

Distribution of respondents by Education level

S. No	Educational status	Father		Mother	
		Number	% (n = 63)	Number	% (n = 63)
1.	No formal Schooling	1	1.6	9	14.3
2.	Primary Schooling	1	1.6	9	14.3
3.	Middle schooling	4	6.3	6	9.5
4.	High schooling	10	15.9	9	14.3
5.	Higher secondary	19	30.2	19	30.2
6.	Diploma / Degree	28	44.4	11	17.5

Fig No: 5.5

Distribution of respondents by Education level



5.6 OCCUPATIONAL STATUS OF THE PARENTS:

The parent's occupational status of the study population is given in table 5.5:

Table No: 5.5

Distribution of subjects by main occupation - Father

Occupation	Number	%
Unskilled worker	10	15.9
Semi skilled worker	18	28.6
Skilled worker	27	42.9
Business / Others	08	12.7
Total	63	100

Majority of the children's father are skilled workers (42.9%), followed by semi-skilled workers (28.6%) and unskilled workers (15.9%). Around 12.7 % are involved in business.

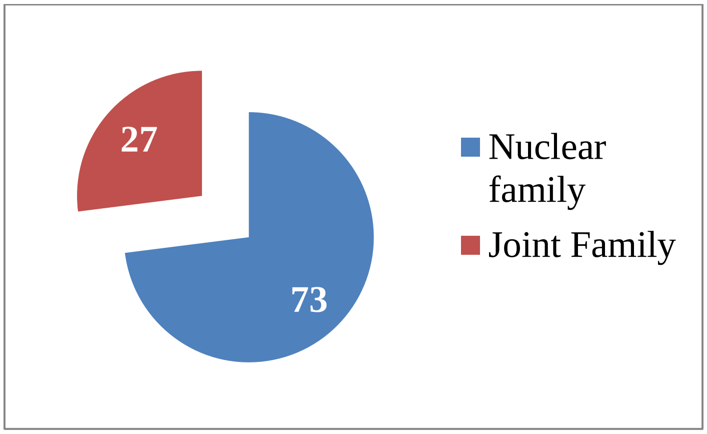
Regarding the occupational status of mother's, nearly 90 % are homemaker's and 10% are employed in unskilled or semi-skilled work.

5.7 FAMILY TYPE:

In the study group, about 46 (73 %) belongs to nuclear family and 17 (27 %) belongs to joint family 17 (27 %). The type of family is given in Fig 5.6:

Fig No: 5.6

Distribution according to Family Type



5.8 FAMILY HISTORY OF HEARING LOSS

Among the study participants, 20 (31.7%) had family history of hearing loss and 43 (68.3%) did not have family history of hearing loss. The family history of hearing loss is shown in table 5.6:

Table No: 5.6

Distribution by Family history of hearing loss

Family H/O	No	%
YES	20	31.7
NO	43	68.3
Total	63	100.0

5.9 CONSANGUINEOUS MARRIAGE OF PARENTS:

With regard to consanguinity, about 25(39.7%) of the study subjects parents had history of consanguineous marriage and 38 (60.3 %) had no history of consanguineous marriage. The consanguinity is given in table 5.7:

Table No: 5.7

Percentage distribution of study subjects according to consanguinity

Consanguineous marriage	**No**	**%**
YES	25	39.7
NO	38	60.3
Total	63	100

5.10 PRENATAL AND NATAL HISTORY

i) TORCH Screening:

Intra uterine infections with Toxoplasmosis, Rubella, Cytomegalovirus and Herpes Simplex (TORCH) may lead to sensorineural hearing loss. Presence of any one of the infection is taken as positive for analysis. TORCH screening was positive in 20 (31.7%) of the children and the remaining 43 (68.3%) tested negative. Among the positives, infection with Toxoplasmosis was higher 11 (55%), followed by Cytomegalovirus 4 (20%), Rubella 3 (15%) and Herpes Simplex virus, 2 (10%).

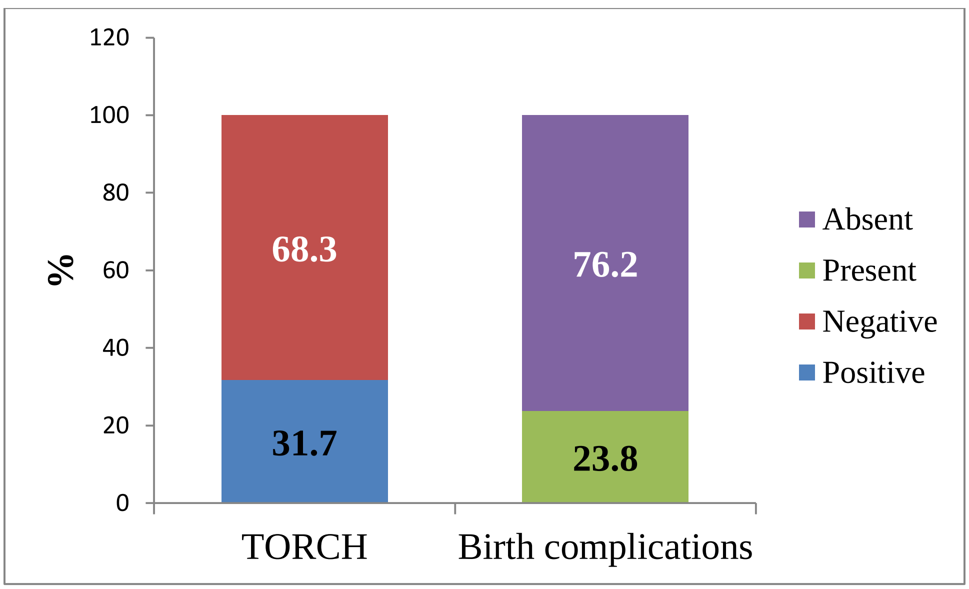
ii) **Birth Complications:**

History of presence of low birth weight, preterm baby, hypoxia and hyperbilirubinemia were taken as birth complications and as single variable for analysis. Birth complications was present in 15 (23.8%) of the children and absent in 48 (76.2%) of the subjects.

The results of TORCH screening and birth complications is summarized in Fig 5.7:

Fig No: 5.7

Distribution according to TORCH and Birth complications



5.11 DEVELOPMENT HISTORY

i) Development Milestones:

The development milestones of the children included in the study were normal in 37 (58.7%) of the children and 26 (41.3%) reported delayed milestones.

ii) Development Quotient (DQ):

Development quotient of ≤ 80 is taken as low score and > 80 as normal. The Development quotient is normal in 43 (68.3%) of the children and low in 20 (31.7%) of the children. The development history is depicted in Table no: 5.8:

Table No: 5.8

Distribution of study subjects according to Development history

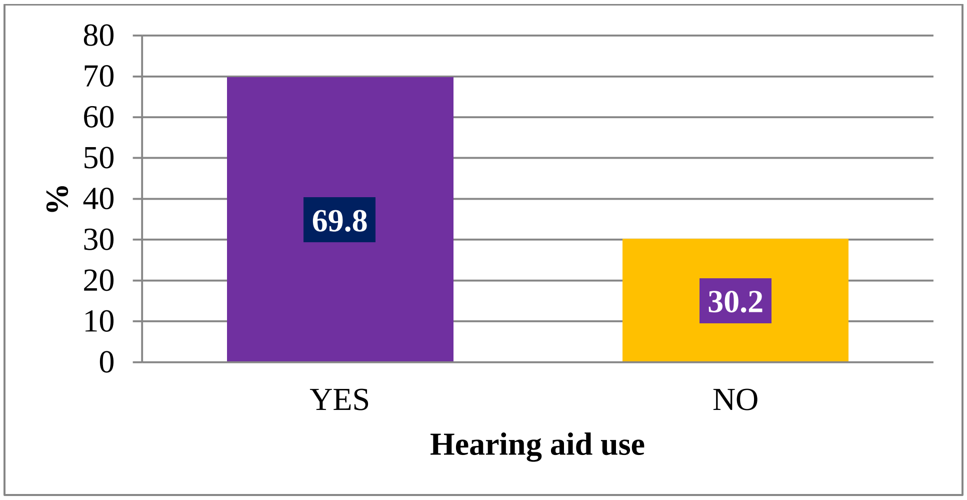
Development history	**No (n = 63)**	**% (n = 63)**
i) Development Milestones		
Normal	37	58.7
Delayed	26	41.3
ii) Development Quotient		
≤ 80	20	31.7
> 80	43	68.3

5.12 HEARING AID USE

Nearly 44 (70%) of the study children had used hearing aids before surgery and 19 (30%) did not have history of hearing aid use. The hearing aid use is given in Fig 5.8:

Fig 5.8

Hearing Aid use distribution



Duration of hearing aid use:

Table No: 5.9

Hearing aid duration of study subjects

Hearing aid use duration	No (n = 44)	% (n = 44)
< 6 months	17	38.6
$\geq$ 6 months	27	61.4
Mean duration (Months) [Mean $\pm$ SD]	7.45 $\pm$ 4.58	

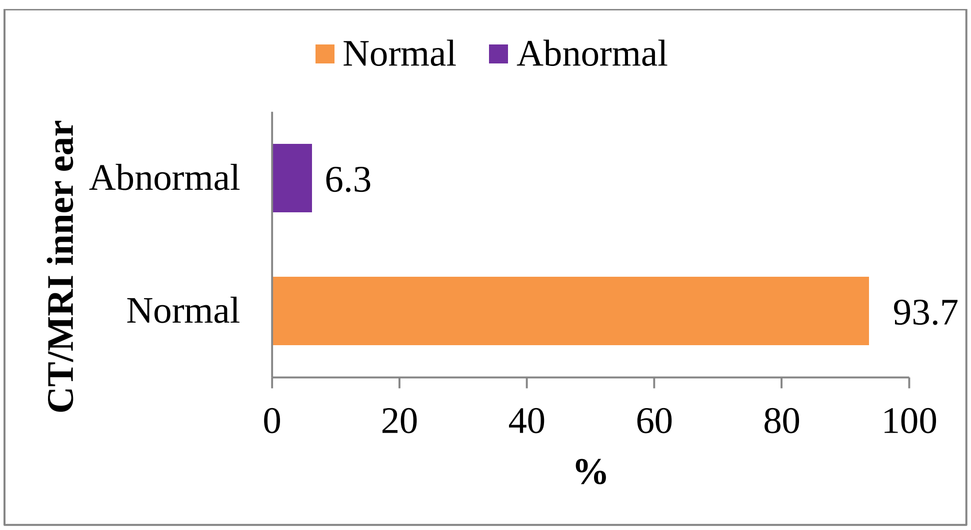
Majority, 27 (61.4%) of the study children had used hearing aids for more than 6 months and 17 (38.6%) had used for less than 6 months duration. The mean duration of hearing aid use was 7.45 $\pm$ 4.58 months. [Median duration = 6 months]

5.13 Radiology

CT and MRI scan was used to assess the inner ear and cochlear malformations. As far as radiology imaging is concerned, majority 59 (93.7 %) had normal CT / MRI findings and about 4 (6.3%) showed abnormal findings in imaging. Abnormal inner ear malformations noted are incomplete partition type I, Bilateral Superior Semicircular Canal Dehiscence and Mondini deformity. The radiology findings are given in Fig no 5.9:

Fig No: 5.9

Percentage distribution of children by imaging of inner ear



5.14 AGE OF COCHLEAR IMPLANT SURGERY

The age at which cochlear implant was done is given in the table 5.10:

Table No: 5.10

Distribution by age at surgery

Age of Surgery	**No**	**%**
≤ 3 years	41	65.1
> 3 years	22	34.9
Total	63	100
Mean age of surgery [Mean ± SD]	2.91 ± 1.3 yrs	

Majority, 41 (65.1%) of the children were operated for cochlear implant by less than 3 years of age and rest 22 (34.9%) after 3 years of age.

5.15 COCHLEAR IMPLANT SURGERY

i) Approach to Cochlea:

The Round Window or cochleostomy technique is used to approach the cochlea. In our study, Round window technique was used in 47 (74.6%) of the children and Cochleostomy approach was used in 16 (25.4%) of the subjects.

ii) Electrode insertion:

Most of the patients, 58 (92.1%) had full insertion of the electrode and in minority of the subjects 5 (7.9%) partial insertion of electrode was done.

The approach to cochlea and electrode insertion findings can be noticed in table 5.11:

Table No: 5.11

Distribution of children according to cochlear implant surgery details

Cochlear implant Surgery	**No (n = 63)**	**% (n = 63)**
i) Approach to Cochlea		
Round Window	47	74.6
Cochleostomy	16	25.4
ii) Electrode insertion		
Full	58	92.1
Partial	05	7.9

5.16 ELECTRODE IMPEDANCE

The Electrode impedance was measured intra-operatively and post-operatively at initial switch on around 3 – 6 weeks. The mean impedance during surgery was 4.86 $\pm$ 0.83 kohm and increased to 7.92 $\pm$ 1.01 kohm at the initial switch on. The difference between the two values was highly statistically significant [Paired t test, $p < 0.0001$]. The electrode impedance values are shown in table 5.12:

Table No: 5.12

Intra-operative and Post –operative impedance measurements

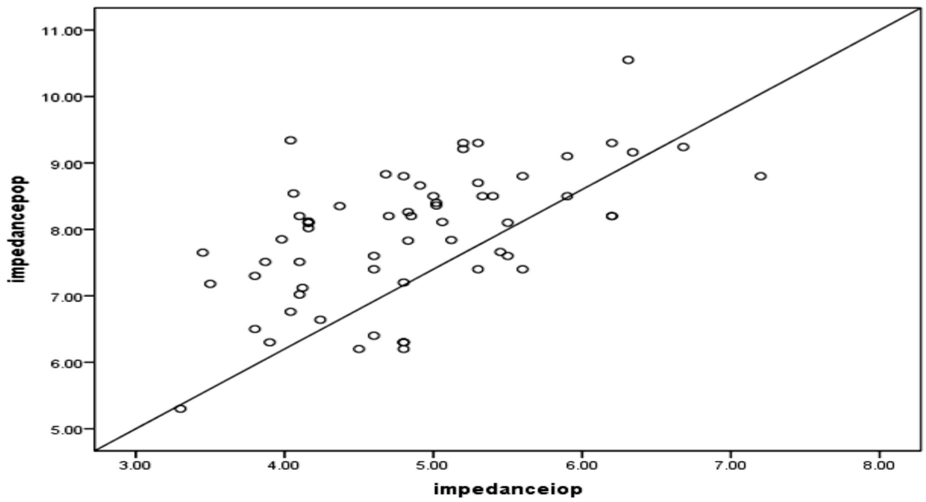
Impedance	Mean	SD	SE	95% CI	Mean Diff.	t value	P value
Intra-operative (kohm)	4.86	0.83	0.10	4.66 – 5.06	3.058	27.485	0.0001*
Post-operative (kohm)	7.92	1.01	0.13	7.66 – 8.18			

(*p < 0.05 Significant)

The Scatter diagram showing the correlation between intra-operative and post –operative impedance measurements is given in Fig 5.10:

Fig No: 5.10

Correlation between Intra-operative and Post –operative impedance



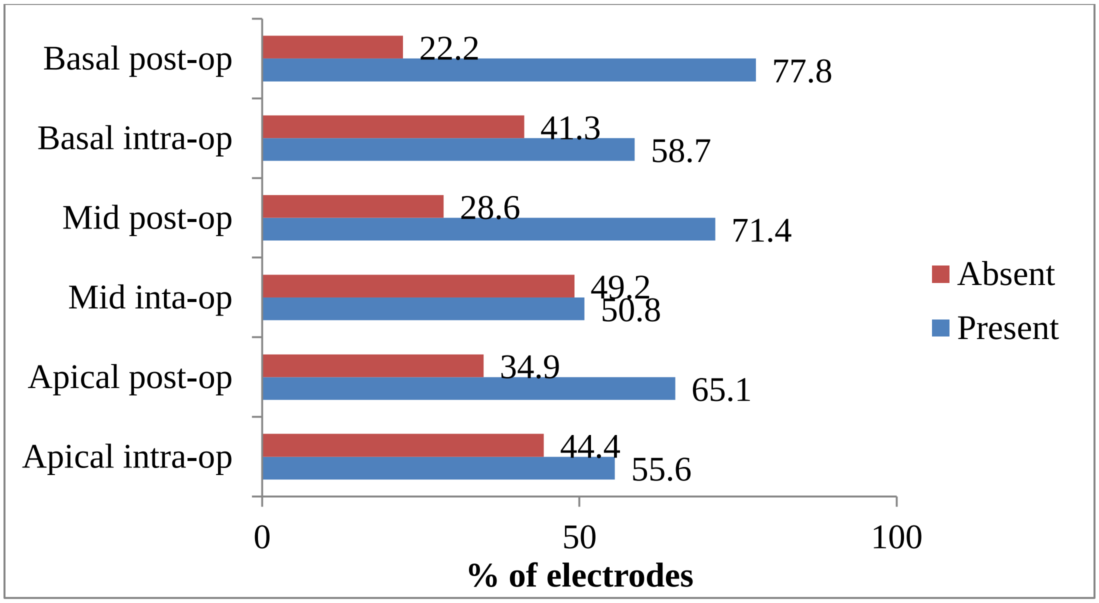
[Pearson correlation - 0.555, p value <0.0001 Highly Significant]

5.17 AUDITORY RESPONSE TELEMETRY [ART]

The Evoked Compound Action Potentials was measured at apical, mid and basal turns of cochlea, during surgery and at switch on 3 – 6 weeks after surgery. The ART measurements are depicted in Fig 5.11:

Fig No: 5.11

Bar diagram showing the ART measurements [Intra-op and Switch on]



When ART measurements are compared intra-operatively and at initial switch on, there was a highly statistically significant association between these two values. [Chi Square – 54.986, p <0.0001] The evoked compound action potentials was absent in 14 (22.2%) of children intra-operatively and present at all electrodes in 20 (31.7%) of subjects. Whereas the evoked compound action potentials were absent in only 8 (12.7%) of children and present in 32 (50.8%) participants post –operatively. The results are given in table 5.13:

Table No: 5.13

ART – Intra operative and Post operative switch on comparison

ART Intra op	ART Post-op [Switch On]							
	Absent		Partial		Present		Total	
	N	%	N	%	N	%	N	%
Absent	8	100.0	2	8.7	4	12.5	14	22.2
Partial	0	0.0	20	87.0	9	28.1	29	46.0
Present	0	0.0	1	4.3	19	59.4	20	31.7
Total	8	100.0	23	100.0	32	100.0	63	100.0

[Chi Square Value = 54.986, p value = < 0.0001* *p <0.05 Significant]

5.18 CATEGORIES OF AUDITORY PERFORMANCE (CAP) SCORE

The Pre-operative CAP score was 0 in 98.4% of the children. At 3 months, 65.1% achieved CAP score of 1 and 25.4% reached the score of 2. By 6 months, 44.4% reached CAP score of 3 and 30.2% had score of 2. About 11.1% reached the highest score of 4 by 6 months. After the follow up of one year, about 14.3% achieved maximum CAP score of 5. About 38.1% and 34.9% of the children reached the CAP score of 4 and 3 respectively. The attainment of CAP scores at different intervals is summarized in table 5.14:

Table No: 5.14

CAP Scores at Pre-operative and Follow up 3, 6 & 12 months

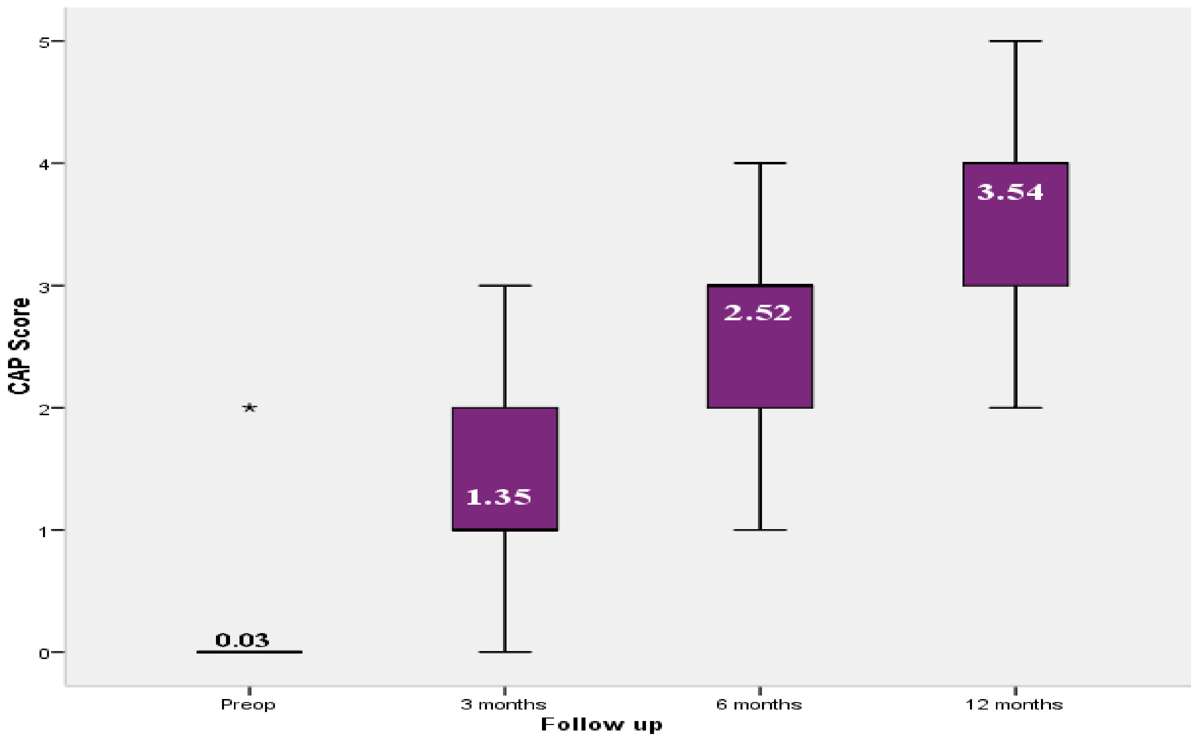
CAP Score	Pre - op		Post - op					
	n	%	3 months		6 months		12 months	
			n	%	n	%	n	%
0	62	98.4	2	3.2	0	0	0	0
1	0	0	41	65.1	9	14.3	0	0
2	1	1.6	16	25.4	19	30.2	8	12.7
3	0	0	4	6.3	28	44.4	22	34.9
4	0	0	0	0	7	11.1	24	38.1
5	0	0	0	0	0	0	9	14.3
6	0	0	0	0	0	0	0	0
7	0	0	0	0	0	0	0	0

The mean CAP score before the implant was 0.03 $\pm$ 0.25 [Median score = 0]. After the cochlear implant surgery, the mean CAP score increased to 1.35 $\pm$ 0.62 [Median score = 1] at 3 months follow up and increased to 2.52 $\pm$ 0.86 [Median score = 3] at 6 months. At the end of 12 months, the mean CAP score was 3.54 $\pm$ 0.88 [Median score = 4]. The box plot showing the CAP scores are given in Fig 5.12:

The increase in scores from baseline to follow up at 3, 6, and 12 months showed a statistically significant difference at each points of follow up. [Wilcoxon Signed Ranks Test, p value < 0.0001]

Fig No: 5.12

Box and Whisker Plot showing the comparison of CAP scores at Pre-operative period and follow up at 3, 6 & 12 months after implant.



5.19 ASSOCIATION BETWEEN VARIABLES AND CAP SCORE

5.19.1 Age at surgery:

The mean CAP score of **3.86 ± 0.89** at 12 months was higher for children who are implanted before 3 years of age, when compared to mean score of **3.37 ± 0.86** for children implanted after 3 years of age. A statistically significant association was observed for children implanted before 3 years of age. [Mann Whitney U test, p value - 0.046] The association between age at surgery and CAP score is shown in table 5.15:

Table No: 5.15

Association between age at surgery and CAP Score

Age at surgery	No	CAP SCORE 12 Months				Mean Rank	'U' value	'p' value
		Mean	SD	SE	95% CI			
$\leq$ 3 years	41	3.86	0.89	0.18	3.58 – 4.24	37.95	320.00	0.046*
> 3 years	22	3.37	0.86	0.13	3.11 – 3.64	28.80		

[Mann Whitney U test , *p <0.05 Significant]

5.19.2 Gender:

The mean CAP score of female children (3.67 $\pm$ 0.92) was slightly higher than the score of male children (3.44 $\pm$ 0.88), but was not statistically significant.

5.19.3 Family Type, Family History and Consanguinity:

The children coming from joint family had a higher mean CAP score (3.65 $\pm$ 0.93) than children from nuclear family. Those children having a positive family history of hearing loss reported higher CAP scores (3.65 $\pm$ 0.87) compared to children without family history of hearing loss. Consanguineous marriage also does not have any significant association with mean CAP scores. The relationship between these variables and CAP score is given in table 5.16:

Table No: 5.16

Association between Gender, Family type, Family history and CAP Score

Variables		No	CAP SCORE 12 Months				'U' value	'p' value
			Mean	SD	SE	95% CI		
Gender	Male	36	3.44	0.88	0.14	3.17 – 3.73	418.00	0.319
	Female	27	3.67	0.92	0.17	3.32 – 4.03		
Family H/O	Yes	20	3.65	0.87	0.19	3.27 – 4.00	384.50	0.478
	No	43	3.49	0.91	0.14	3.23 – 3.76		
Family type	Nuclear	46	3.50	0.89	0.13	3.23 – 3.74	354.00	0.545
	Joint	17	3.65	0.93	0.22	3.20 – 4.09		
Consanguineous marriage	Present	25	3.56	0.96	0.19	3.21 – 3.95	459.50	0.818
	Absent	38	3.53	0.85	0.14	3.24 – 3.80		

5.19.4 Development History:

The children with normal development milestones achieved a greater mean CAP score of 3.97 $\pm$ 0.76, than children with delayed milestones (2.92 $\pm$ 0.69). Likewise, the children with Development Quotient (DQ) of $\leq$80% had lower CAP scores (3.20 $\pm$ 0.95) compared to children with DQ of >80% (3.70 $\pm$ 0.83). The development milestones and DQ was found to have a strong significant association with increase in CAP scores. [Mann Whitney U test, p value < 0.05] The association between development history and CAP score at 12 months is given in table 5.17:

Table No: 5.17

Association between Development history and CAP Score

Development H/O		No	CAP SCORE 12 Months				'U' value	'p' value
			Mean	SD	SE	95% CI		
Development milestones	Normal	37	3.97	0.76	0.12	3.73 – 4.20	418.00	0.001*
	Delayed	26	2.92	0.69	0.13	2.65 – 3.18		
Development Quotient	≤ 80%	20	3.20	0.95	0.21	2.78 – 3.62	299.00	0.040*
	> 80%	43	3.70	0.83	0.13	3.44 – 3.96		

[Mann Whitney U test, *p <0.05 Significant]

5.19.5 Birth Complications:

The birth complications like low birth weight, hypoxia, prematurity are associated with lower CAP scores, compared to children with no birth complications. (3.08 ± 0.87 Vs 3.66 ± 0.89) This association was found to be statistically significant and the details are given in table 5.18:

Table No: 5.18

Association between Birth complications and CAP Score

Birth complications	No	CAP SCORE 12 Months				Mean Rank	'U' value	'p' value
		Mean	SD	SE	95% CI			
YES	15	3.08	0.87	0.24	2.62 – 3.55	23.73	217.50	0.048*
NO	48	3.66	0.88	0.12	3.41 – 3.90	34.15		

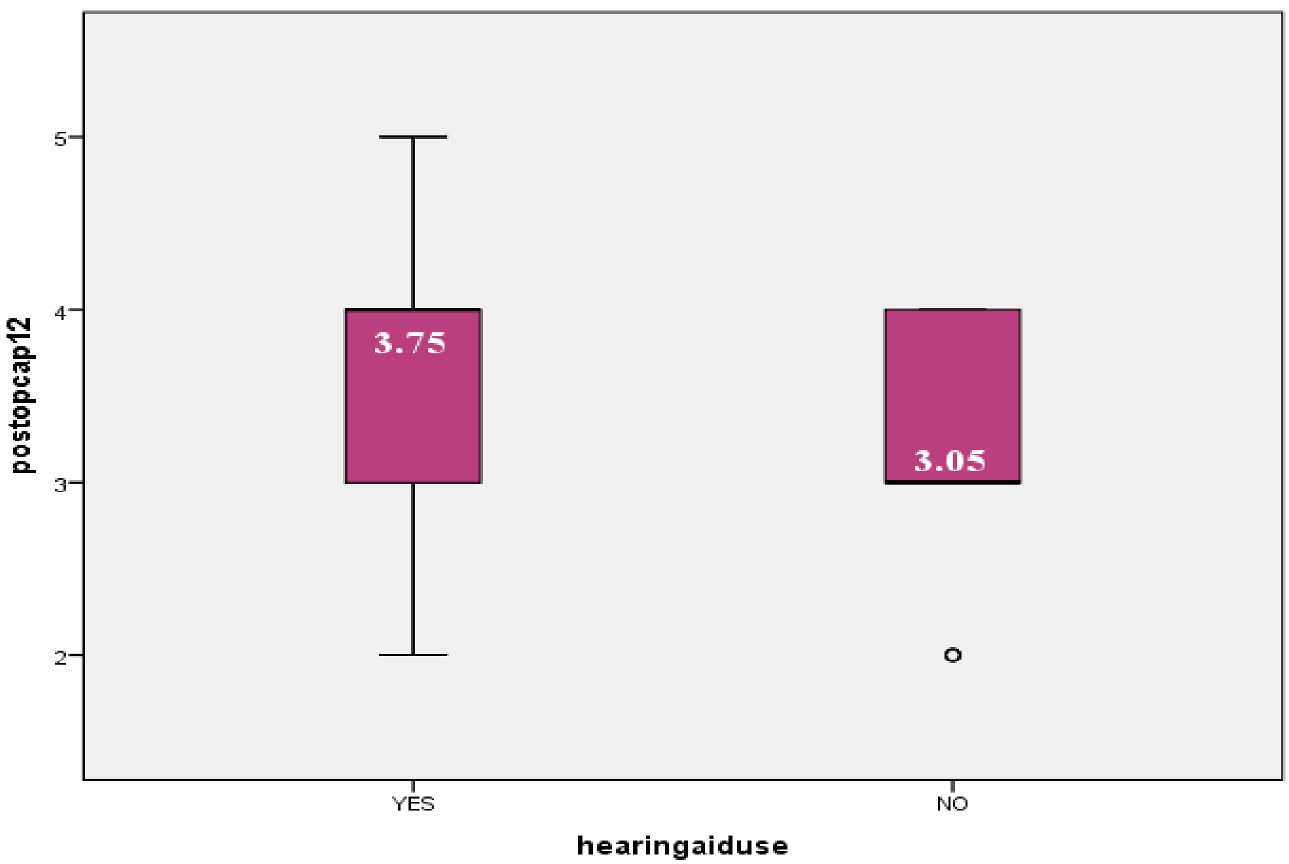
[Mann Whitney U test *p <0.05 Significant]

5.19.6 Hearing aid use:

The mean CAP scores of the children who had used hearing aid (3.75 ± 0.89) are higher as compared to children not used hearing aids (3.05 ± 0.70). This relation was found to be highly significant and the results are depicted in Fig 5.13:

Fig No: 5.13

Association between hearing aid use and CAP Score



[Mann Whitney U value – 235.50, p value – 0.004*, *p <0.05 Significant]

Duration of hearing aid use with age at surgery:

Table No: 5.19

Hearing aid duration and age at surgery

Hearing aid use duration		**Age at Surgery**		**Total (n = 44)**
		≤ 3 Years	**> 3 Years**	
< 6 months	N (%)	11(64.7%)	6 (35.3%)	17 (100%)
	CAP score (Mean ± SD)	3.81±1.07	3.5 ± 1.04	
≥ 6 months	N (%)	13 (48.1%)	14 (51.9%)	27 (100%)
	CAP score (Mean ± SD)	3.94±0.83	3.46 ± 0.66	

[F value – 1.259, p value – 0.302]

The mean CAP score of children less than 3 years who had used hearing aid for more than 6 months is 3.94±0.83, which was higher compared to children > 3 years. But no statistically significant association was found between CAP score with hearing aid use and age at surgery. The results are given in table 5.19:

5.19.7 Radiological findings:

The children with normal inner ear anatomy had achieved a greater mean CAP score, than children with inner ear malformations (3.61 ± 0.87 Vs 2.50 ± 0.58). This association was found to be highly statistically significant and the details are given in table 5.20:

Table No: 5.20

Association between Radiological findings and CAP Score

CT/MRI - Inner ear	No	CAP SCORE 12 Months				Mean Rank	'U' value	'p' value
		Mean	SD	SE	95% CI			
Normal	59	3.61	0.87	0.11	3.39 – 3.82	33.36	38.00	0.017*
Abnormal	04	2.50	0.58	0.29	1.92 – 3.08	12.00		

[Mann Whitney U test, *p <0.05 Significant]

5.19.8 Cochlear implant Surgery:

There was no significant difference between round window and cochleostomy approach with CAP score in this study. Full insertion of electrode is associated with increased levels of CAP scores compared to partial insertion of electrodes, but not statistically significant. The association between approach to cochlea and electrode insertion is shown in table 5.21:

Table No: 5.21

Association between Surgery techniques and CAP Score

Surgery techniques	No	CAP SCORE 12 Months				Mean Rank	'U' value	'p' value
		Mean	SD	SE	95% CI			
i) **Approach to Cochlea**								
Round Window	47	3.55	0.88	0.13	3.28 – 3.81	32.41	356.50	0.745
Cochleostomy	16	3.50	0.97	0.24	3.06 – 4.00	30.78		
ii) **Electrode insertion**								
Full	58	3.55	0.92	0.12	3.31 – 3.79	32.28	128.50	0.658
Partial	05	3.40	0.54	0.24	3.00 – 3.88	28.70		

5.19.9 Auditory Response Telemetry:

The mean CAP score at 12 months is compared with evoked compound action potentials at initial switch on. The mean CAP score increased from 3.38 ± 0.26 in absent electrodes to 3.61 ± 0.99 in present electrodes. But the association between ART measurements and CAP score is not statistically significant. The results are summarized in the table 5.22:

Table No: 5.22

Association between ART measurement and CAP Score

ART Switch on	No	CAP SCORE 12 Months				Mean Square	'F' value	'p' value
		Mean	SD	SE	95% CI			
Absent	08	3.38	0.74	0.26	2.86 – 3.90	0.164	0.20	0.819
Partial	23	3.53	0.88	0.16	3.21 – 3.85			
Present	32	3.61	0.99	0.21	3.19 – 4.03			

6. DISCUSSION

A Prospective study was done to assess the auditory performance following cochlear implant surgery among children aged less than six years at Upgraded Institute of Otorhinolaryngology, Rajiv Gandhi Government General hospital, Chennai. A total of 70 children were enrolled for the study satisfying the inclusion criteria and followed up for 12 months. At the end of study, 63 children successfully completed the follow up and seven children were lost due to follow up. Electrode impedance and Evoked compound action potential telemetry was recorded intra operatively and at the initial switch on. The auditory performance was assessed using Categories of Auditory Performance (CAP) scores.

Among the study participants, Majority 41.3% was in 13 – 24 months age group, followed by 37 – 48 months age group (20.6%). About 8% were one year old. The median age of the children was 30 months. [Mean age – 34.00 ± 15.54 months].

In our study, 65.1% of the children achieved CAP score of 1 by 3 months, 44.4% reached score of 3 by 6 months and 38.1% had score of 4 by 12 months. About 14.3% reached maximum score of 5 by 12 months. After cochlear implant surgery, there was a statistically significant increase in median CAP scores during the follow up period. At 3 months, the median CAP score was 1, which increased to 3 at 6 months and 4 at 12 months.

Zhou et al, [71] studied thirty-four congenital deaf children who underwent cochlear implant before the age of 18 months. The median CAP score before surgery was 0, and it increased to 4 at 6 months, 5 at 12 months, and 7 at 24 months after cochlear implantation in China.

Kameswaran et al, [64] in Chennai had evaluated the quality of life after cochlear implantation with Category of Auditory Performance scores. Out of the 100 implantees, 39% were between1-5 yrs of age and adults were 14%. The results of CAP score showed that 10% implantees achieved category 7 in 12 months in 1-5 yrs age group and 13% achieved category 6 in 12 months, whereas in 6-10 yrs, 4% got category 7 in 12 months and 9% achieved category 6. Children responded better with very good outcomes with early cochlear implantation.

The age of implantation has significant impact on the auditory performance of the children. Regarding the age at surgery in our study, 65.1% of the children were operated by less than 3 years of age and rest 34.9% after 3 years of age. The mean age of surgery was 2.91 $\pm$ 1.3 years. The mean CAP scores of the children operated before 3 years was higher, when compared to children undergone surgery after 3 years and it was found to be statistically significant ($P<0.05$).

Early age of implantation had better performance than children implanted at later age due to loss of neural plasticity. Although cochlear implants done before the age of 6 years can provide significant benefit, better outcomes are seen with implantation at early age. Various studies done in different parts of the world have also emphasized the importance of early implantation for better outcomes.

Goverts et al, [12] compared the CAP scores with age of implantation in 48 children under six years of age and concluded that CAP scores increased in all children after implantation. Implantation of children less than 2 years resulted in immediate normalization of CAP scores with 90% probability of integration into normal school when compared to children of higher ages. Children implanted between 2- 4 years resulted in normal CAP scores by 3 years with 66% probability of integration into primary school, whereas surgery after 4 years hardly resulted in normal CAP scores with 20% probability of integration into primary school.

Colletti et al, [72] in 2005 reported that children implanted between 1- 3 years had delayed auditory performance than children implanted before 1year of age, thereby stressing the need for implantation before first birthday.

Tomblin et al, [73] showed that children implanted before the age of 42 months demonstrated auditory evoked P1 response latencies that mirrored those of children who were born with normal hearing, compared to children implanted after the age of 84 months.

Nikolopoulos et al, [55] reported better CAP scores in children implanted before four years and poor results among children who underwent surgery after four years.

The early age of cochlear implantation leads to better and faster performance in children has been reported by many authors around the world, like **Holt et al,** [68] **Osberger et al** [56] **Toeh et al** [60] in 2004 and **Kirk et al.** [57]

The presence of evoked compound action potentials was recorded intra-operatively and post-operatively in this study. The evoked compound action potentials was absent in 22.2% of children intra-operatively and present at all electrodes in 31.7% of subjects. Whereas the evoked compound action potentials were absent in only 12.7% of children and present in 50.8% of children post-operatively, showing a significant difference between the two measurements.

The mean CAP score increased from 3.38 $\pm$ 0.26 in absent electrodes to 3.61 $\pm$ 0.99 in present electrodes, but there was no significant association between ART and post-operative performance.

Shennawy et al, [38] studied 44 subjects (12 adults, 32 children) implanted at the Cochlear Implant Unit, Cairo University. ECAP thresholds and electrode impedance measures were collected intra-operatively, at initial stimulation and at 9 – 12 months post initial stimulation. There was significant increase in impedance from intra-operative to initial stimulation, and a significant decrease from initial stimulation to the one-year post implant visit. Intra-operative ECAP measurements compared with post-operative performance showed no significant correlation. He concluded that telemetry gives valuable information regarding the electrical output of the implant and the response of the auditory system. However, it is not a valuable predictor of the patient's post-operative performance.

Cossetti et al, [37, 48] had evaluated the importance of intra operative telemetry monitoring. Intra operative abnormal impedances are transient; due to air bubbles generated by electrode insertion and may resolve quickly, thus explaining the frequent normalization of these measures. Intra operative ECAP measurements can provide a valid basis for initial programming, especially in difficult-to-program populations such as very young children or those with multiple disabilities. Absent tNRT responses on 1 or more electrodes occurred in 14% of patients, although complete lack of response was rare (1.4%) and did not correlate with a dysfunctional device or the post operative performance.

In our study, the Electrode impedance was measured intra-operatively and post-operatively on initial switch at around 3 – 6 weeks. The mean impedance during the surgery was 4.86 $\pm$ 0.83 kohm and increased to 7.92 $\pm$ 1.01 kohm at the initial switch on. We noticed a highly statistically significant correlation between these two measurements [$p < 0.001$]. The presence of abnormal electrodes was also less than 1%, signifying the performance of electrodes. But we could not find any significant association between electrode impedance and auditory performance of the children.

Goehring et al, [29] conducted a retrospective study to review the intra operative and postoperative cochlear implant electrode impedances among 165 children and adults. He found that, 97.5% had normal impedance both intra operatively and postoperatively. About 0.17% had normal impedance intra operatively and abnormal measures postoperatively, in contrast to 1.9% of electrodes having abnormal intra operative impedance and normal measures postoperatively. Finally, the incidence of an electrode having an abnormal value for both the intra-operative and post-operative periods was 0.41%.

Despite the low incidence of abnormal intra operative impedance and chance of high resolution post operatively, intra operative device testing is a useful tool in assessing the device integrity and gives a peace of mind for surgeon and family that the device is functioning normally. Lastly, the author concludes that impedance monitoring should be done intra-operatively and at regular post operative intervals.

Among the 63 children implanted, Round Window approach was used in 74.6% and 25.4% through cochleostomy approach. The mean CAP scores of both the groups at 12 months were more or less comparable, and the association was also not significant. The results of this study are comparable with other studies also.

BJ Kang et al, [74] compared the cochlear implant performance with round window or cochleostomy approach in a group of 84 children in New York. He found that there was no significant difference in post operative performance at 12 months between the two groups. He concluded that patients with favourable round window anatomy, who underwent round window electrode insertion showed comparable auditory outcome scores with the traditional cochleostomy group.

Hamerschmidt et al, [75] studied the comparison between the neural response telemetry via round window or cochleostomy approach in cochlear implantation. This prospective study comprised of 23 patients, six approached by cochleostomy and seventeen through round window insertion. They concluded that, both techniques equally stimulated the cochlear nerve and the choice of approach depends on the surgeon's preference and experience.

Regarding the electrode insertion, majority (92%) had full insertion of electrodes and rest 8% had partial insertion in our study. Full insertion of electrode is associated with increased levels of CAP scores (3.55 $\pm$ 0.92) compared to partial insertion of electrodes (3.40 $\pm$ 0.54) at 12 months. But we could not find any significant difference in the insertion of electrodes as seen in other studies.

Nayak et al, [76] assessed the outcome of cochlear implantation with the depth of electrode insertion in 30 children with congenital SNHL undergoing cochlear implant surgery. The results indicate that auditory performance was better in children with complete insertion than with partial insertion.

Yukawa et al [77] also studied the depth of insertion of electrode array with post operative speech perception in post lingually deaf adults, and concluded that full insertion resulted in better speech perception.

In our study, analysis was done to find any association between perinatal complications and auditory performance. About 23.8% of the children had perinatal problems like low birth weight, hypoxia, prematurity and hyperbilirubinemia and 31.7% had any one of the TORCH infections.

A statistically significant association was noted among children with perinatal complications, compared with normal children [$p<0.05$]. The mean CAP score at 12 months was higher among normal children than children with birth complications.

Kang DH et al, [78] evaluated the outcomes of cochlear implant in children with perinatal problems and inner ear anomalies. In their study, perinatal disorders most commonly encountered are infections, prematurity, low birth weight, hyper-bilirubinemia and the post operative outcome in these patients is highly variable but favourable.

In this study, 6.3% of the children had inner ear malformations like incomplete partition type I, Bilateral Superior Semicircular canal dehiscence and Mondini deformity. The children with abnormal findings of inner ear were found to have lower mean CAP scores than normal children at 12 months (2.50 ± 0.58 Vs 3.61 ± 0.87). This association was found to be highly statistically significant [$p<0.05$].

Wermeskerken et al, [79] analyzed audiological performance after cochlear implantation in a sample of children with radiographically detectable inner ear malformations. The responses are delayed in these children compared to their normal peers, but finally they reach a favourable outcome over a period of two years.

Kang DH et al, [78] also studied the outcome in patients with inner ear anomalies and concludes that, they have poorer outcomes compared to their normal peers.

7. CONCLUSION

Cochlear implantation is a safe and effective procedure for hearing rehabilitation in children with profound sensorineural hearing loss and had created a new opportunity for development of communication skills.

1. Our study results show that, early implantation leads to better auditory performance compared to implantation at later ages. All children less than 6 years got benefit from cochlear implants, however surgery done before 3 years played a significant role in attaining age appropriate auditory skills.
2. Other factors like Development milestones, Development quotient, perinatal complications, hearing aid use and normal cochlear morphology have significant impact on the outcome and associated with higher CAP scores.
3. Cochlear implant done in children with abnormal cochlear morphology had lower CAP scores compared to children with normal cochlea.
4. The Electrode impedance and telemetry measurements during intraoperative and postoperative settings can provide valuable information regarding the integrity and response of the auditory system, which helps in programming the device.
5. Further measures like abnormal impedance or absence of tNRT does not indicate a lack of stimulation and do not correlate with post operative performance.

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9. APPENDIX

1. ABBREVIATIONS
2. CASE PROFORMA
3. CONSENT FORM
4. MASTER CHART
5. INSTITUTIONAL ETHICS COMMITTEE APPROVAL
6. PLAGIARISM CERTIFICATE

ABBREVIATIONS

ART	Auditory Response Telemetry
BERA	Brainstem Evoked Response Audiometry
BOA	Behavioral Observation Audiometry
CAP	Categories of Auditory Performance
CI	Cochlear Implant
CI	Confidence Interval
CT	Computed Tomography
dB	Decibels
EABR	Evoked Auditory Brainstem Response
ECAP	Evoked Compound Action Potentials
EI	Electrode Impedance
IA	Impedance Audiometry
MRI	Magnetic Resonance Imaging
NRT	Neural Response Telemetry
OAE	Oto-Acoustic Emissions
PA	Play Audiometry

PTA	Pure Tone Audiometry
SA	Speech Audiometry
SD	Standard Deviation
SE	Standard Error
SNHL	Sensorineural Hearing Loss
SOE	Spread of Excitation
VRA	Visual Reinforcement Audiometry

ELECTROPHYSIOLOGY AND AUDITORY PERFORMANCE OF CHILDREN WITH PROFOUND SENSORYNEURAL HEARING LOSS AFTER COCHLEAR IMPLANT SURGERY

QUESTIONNAIRE

1. Name: ……………………… 2. Age: ___ yrs 3. Sex: Male / Female

4. District of origin:………………

5. Religion: Hindu / Christian / Muslim / Others

6. Education of parents:

Father: Illiterate / Primary schooling / Secondary / High school / Higher secondary / Degree / Others

Mother: Illiterate / Primary schooling / Secondary / High school / Higher secondary / Degree / Others

7. Occupation of parents:

Father: Unemployed/ Unskilled Worker / Semiskilled Worker / Skilled Worker / business / Professional / Others …………..

Mother : Homemaker/ Unskilled Worker / Semiskilled Worker / Skilled Worker / business / Professional / Others …………..

8. Income : ________/ Month

9. Family details:

Type of family: Nuclear / joint

Family history: Yes / No

Consanguinous marriage: Yes / No

10. Degree and type of hearing loss:

11. Hearing aid use: Duration:

12. Development milestones: Normal / delayed

13. TORCH infections:

14. Perinatal complications:

15. Birth weight:

16. Development Quotient:

17. Age of surgery:

18. Type of implant:

19. Audiological assessment:

S.no	Tests	Preop
1.	BERA	
2.	Impedence	
3.	OAE	
4.	BOA	

20. CT / MRI Findings:

21. Cochlea approach: Round window / Cochleostomy

22. Electrode insertion: Full / Partial

23. Electrode impedance:

    Intraop:

    Postop:

24. ART:

	Intra-op	Post-op
Apical:		
Mid :		
Basal :		

25. Categories of Auditory Performance (CAP):

Category	Criteria	Pre-implant	Post implantation			
			0	3 months	6 months	12 months
7	Uses telephone					
6	Understands conversation					
5	Understands phrases					
4	Discrimination of sounds					
3	Identifies environmental sounds					
2	Response to speech sounds					
1	Awareness of environmental sounds					
0	No awareness to environmental sounds					
Total numbers						

Signature of Investigator:

Date:

PATIENT CONSENT FORM

Title of the Study:

"ELECTROPHYSIOLOGY AND AUDITORY PERFORMANCE OF CHILDREN WITH PROFOUND SENSORYNEURAL HEARING LOSS AFTER COCHLEAR IMPLANT SURGERY"

Institution : Upgraded institute of Otorhinolaryngology, Rajiv Gandhi Government General Hospital, Chennai – 600003.
Institute of Speech and Hearing, Rajiv Gandhi Government General Hospital, Chennai – 600003

Name : Date:

Age : IP No:

Sex : Study Patient No:

The details of the study have been provided to me in writing and explained to me in my own language.

I confirm that I have understood the above study and had the opportunity to ask questions.

I understood that my participation in the study is voluntary and that I am free to withdraw at any time, without giving any reason, without the medical care that will normally be provided by the hospital being affected.

I agree not to restrict the use of any data or results that arise from this study provided such a use is only for scientific purpose(s).

I have been given an information sheet giving details of the study.

I fully consent to participate in the above study.

__________________ ________________ ____________

Name of the subject Signature Date

__________________ ________________ ____________

Name of the Investigator Signature Date

INFORMATION SHEET

- We are conducting a prospective study on **"ELECTROPHYSIOLOGY AND AUDITORY PERFORMANCE OF CHILDREN WITH PROFOUND SENSORYNEURAL HEARING LOSS AFTER COCHLEAR IMPLANT SURGERY"** at the Upgraded Institute of Otorhinolaryngology, Madras Medical College & Rajiv Gandhi Government General Hospital, Chennai – 600003.

- Cochlear implant surgery is a minimally invasive surgical procedure performed to treat hearing loss and eventually leads to better quality of life.

- In this surgery, the cochlear implant (Receiver with electrode) is placed under the skin just behind the ear. The electrode array which transmits the sound impulses to the cochlear nerve is inserted into the inner ear.

- In this surgery, nothing is inserted into the brain and no part of brain tissue is exposed.

- At the time of announcing the results and suggestions, name and identity of the patients will be confidential.

- Taking part in this study is voluntary. You are free to decide whether to participate in this study or to withdraw at any time; your decision will not result in any loss of benefits to which you are otherwise entitled.

Signature of Investigator: **Signature of Parent/guardian:**

Date:

ஆராய்ச்சி தகவல் தாள்

சென்னை மருத்துவக் கல்லூரியில், நரம்பு சம்பந்தமான காது கேளாமை உள்ள குழந்தைகளுக்கு உட்செவிச்சுருள் பதியம் அறுவை சிகிச்சை செய்தபின், அதனுடைய மின்யியங்கியல் மற்றும் செவிப்புல செயல்திறன் பற்றி ஆய்வு நடத்துகிறேன்.

உட்செவிச்சுருள் பதியம் அறுவை சிகிச்சை என்பது நரம்பு சம்பந்தமான காது கேளாமை உள்ள குழந்தைகளுக்கு குறைபாடு சரி செய்யப்படுவதுடன், அவர்களுடைய வாழ்க்கைத் தரமும் உயர வழிவகுக்கிறது.

இந்த அறுவை சிகிச்சையில், உட்செவிச்சுருள் பதியம் (மின்முனையுடன் கூடிய ரிசீவர்) காதின் பின்புறம் உள்ள தோலின் கீழ் வைக்கப்படும். உள் காதினுல் வைக்கப்படும் மின்முனையின் வழியாக ஒலிவிசைகள் பரிமாற்றம் செய்யப்படுகிறது. இதன் மூலம் காதுகேட்கும் திறன் ஏற்படுகிறது.

இந்த அறுவை சிகிச்சையில், மூளையின் உள் எதுவும் வைக்கப்படுவதில்லை. அதனால் மூளை திசுக்கள் எந்தவகையிலும் பாதிக்கப்படுவதில்லை.

இந்த ஆய்வில் உங்களுடைய சொந்த விருப்பத்தின் அடிப்படையில் கலந்துகொள்ளலாம். எந்த தருணத்திலும் இந்த ஆய்வில் இருந்து விலகுவதற்கும் அனுமதி உண்டு.

ஆய்வின் முடிவுகள் வெளியிடும்போது, நபர்களுடைய பெயரோ மற்ற குறிப்புகளோ வெளியில் தெரியாதவாறு நம்பத் தகுந்த முறையில் பாதுகாக்கப்படும்.

ஆராய்ச்சியாளர் கையொப்பம்

பெற்றோர்/
பாதுகாவலர் கையொப்பம்

தேதி :

ஆராய்ச்சி ஒப்புதல் கடிதம்

ஆராய்ச்சி தலைப்பு

நரம்பு சம்பந்தமான காது கேளாமை உள்ள குழந்தைகளுக்கு உட்செவிச்சுருள் பதியம் அறுவை சிகிச்சை செய்தபின், அதனுடைய மின்யியங்கியல் மற்றும் செவிப்புல செயல்திறன் பற்றி ஆய்வு

நிறுவனம்: Upgraded Institute of Otorhinolaryngology,
சென்னை மருத்துவக் கல்லூரி, சென்னை-3.

பெயர் :

வயது :

பாலின் :

தேதி :

எண் :

1) ஆய்வு விவரங்களை என் சொந்த மொழியில் எனக்கு எழுதியும் விளக்கியும் வழங்கினார்.

2) நான் இந்த ஆய்வை புரிந்து மற்றும் கேள்விகள் கேட்க வாய்ப்பு கிடைத்தது என்று உறுதிப்படுத்துகிறேன்.

3) இந்த ஆய்வு முடிவு அல்லது தகவல் அறிவியல் நோக்கத்துக்கா எதிர் காலத்தில் பயன்படுத்த எனக்கு எந்த தடையும் இல்லை.

4) நான் இந்த ஆய்வு தொடர்பாக ஒரு தகவல்தாள் பெற்றுக்கொண்டேன்.

5) நான் முவுமையாக மேலே கூறிய ஆய்வில் பங்கேற்க சம்மதிக்கிறேன்.

பெயர் கையொப்பம் தேதி

புலன் விசாரணை பெயர் கையொப்பம் தேதி

name	agemonths	sex	district	religion	fatheredu	motheredu	fatheroccp	motheroccp	income	familytype	familyh/o	consgmarriage	hearingaiduse	duration	devpmilestones	agesurgery	ctmri	preopcap	postopcap3	postopcap6	postopcap12	intraopimpedence
srihari	24	1	tvm	1	5	2	3	1	6000	1	1	1	1	3	1	2	1	0	1	3	4	1
sharmila	36	2	dgl	1	5	3	3	1	7000	1	1	1	1	7	1	3	1	0	2	3	4	1
tharun	24	1	chen	1	6	5	4	3	8000	1	2	1	1	6	2	2	1	0	1	2	3	1
omritheswaran	18	1	chen	1	6	5	4	1	8000	1	1	1	1	3	2	1.6	1	0	1	1	2	1
rithikadevi	18	2	chen	1	6	6	4	1	8000	1	2	2	1	3	1	1.6	1	0	1	2	4	1
kavinash	48	1	tvm	1	6	5	4	1	8000	1	2	1	1	5	2	4	1	0	1	1	2	1
moorthy	24	1	cgl	1	4	4	3	1	7000	1	2	2	2		2	2	2	0	1	1	2	2
jeevitha	24	2	kgri	1	4	4	3	1	6000	1	1	1	2		1	2	1	0	1	1	2	1
brindashree	60	2	chen	1	6	6	4	3	8000	1	1	1	1	6	2	2	1	0	0	1	2	1
nishanthan	36	1	dgl	1	6	6	5	4	9000	1	2	2	1	4	2	2	1	0	1	1	2	1
manikandan	48	1	dgl	1	5	1	4	1	6000	1	1	1	1	12	1	5	1	0	2	3	5	1
yaswanth	42	1	slm	1	4	3	4	1	5000	1	1	1	1	12	2	3	2	0	1	2	3	1
nishanthi	36	2	vel	1	6	4	3	1	6000	1	1	2	1	9	2	4	1	0	1	1	3	1
sajithkumar	48	1	cdl	1	3	1	2	1	6000	1	2	2	2		2	3	1	0	1	2	3	1
manoj	24	1	tnvi	1	6	5	4	1	7000	1	1	1	1	6	2	3.6	1	0	1	2	3	1
karthiga	24	2	tvm	1	5	1	3	1	6000	1	2	2	2		2	3	1	0	1	2	3	1
kalyansundaram	48	1	chen	1	6	5	5	1	7000	2	2	2	1	8	2	3	1	0	1	2	3	1
kamalesh	48	1	vpm	1	3	1	2	1	5000	1	1	1	2		2	4	1	0	2	3	4	1
abdulrahim	24	1	chen	3	4	4	3	1	6000	2	1	1	1	3	2	4	1	0	2	3	4	1
sahayarohith	36	1	tnvi	2	6	6	4	1	7000	2	2	2	1	4	2	2	1	0	1	2	3	1
logavarshini	60	2	chen	1	6	6	4	1	8000	1	2	2	1	6	1	2	1	0	1	3	4	1
tamilarasu	30	1	tivl	1	4	2	2	2	7000	1	1	1	2		2	5	1	0	1	2	3	1
deepti	24	2	tvm	1	4	1	2	1	5000	2	2	2	1	6	1	4	1	0	1	4	5	1
kishorekumar	18	1	rmd	1	5	5	3	1	6000	2	1	1	1	3	2	4	1	0	2	3	4	1
thigaloviyan	60	1	vel	1	6	5	4	1	8000	1	2	2	2		1	1	1	0	1	4	4	1
monisha	60	2	chen	2	4	1	2	1	6000	2	1	1	1	3	2	2	1	0	1	3	4	1
sabarish	60	1	ariy	1	3	1	2	1	4000	1	1	1	2		1	2	1	0	1	2	4	1
hashini	66	2	chen	1	6	6	4	1	6000	2	2	2	1	8	1	3	1	0	1	3	4	1
chandran	48	1	slm	1	6	6	5	1	7000	1	1	1	1	12	2	5	1	0	1	2	3	1
rithickroshan	60	1	tjr	1	5	2	4	2	7000	1	2	1	1	3	1	2.6	1	0	1	3	5	1
sadhana	36	2	tivl	1	6	6	5	1	8000	2	2	2	2		2	1.3	2	0	1	1	2	1
abdulajeesh	42	1	chen	3	5	2	3	1	5000	2	2	1	1	4	1	2	1	0	1	3	5	1
lakshmankandan	30	1	chen	1	6	4	4	1	7000	1	2	2	2		1	1	1	0	2	3	4	1
ashwin	36	1	vel	1	5	3	3	1	7000	2	1	1	1	3	1	1.6	1	0	1	3	4	1
suseendran	42	1	kgri	1	5	3	3	1	6000	2	1	1	2		2	2	1	0	1	1	2	1
sridharsini	18	2	vel	1	6	2	4	1	6000	1	2	2	1	12	1	5	1	0	1	3	4	1
hemanthkumar	48	1	chen	1	5	3	3	1	5000	1	1	1	1	6	1	5	1	2	3	4	5	1
damodharan	66	1	kgri	1	4	4	3	1	6000	1	2	1	1	12	1	5	1	0	2	3	4	1
mahalakshmi	18	2	ngp	1	5	4	3	1	5000	1	2	1	1	3	2	6	1	0	1	2	3	1
sarathy	24	1	tvm	1	4	3	2	1	5000	1	2	2	2		2	2	2	0	1	2	3	1
ashwini	24	2	tvm	1	3	1	2	1	5000	1	1	1	2		2	4	1	0	0	3	4	1
sridaran	48	1	vpm	1	1	1	2	2	8000	1	2	1	2		1	1.6	1	0	1	3	3	2
kaviyadarshini	18	2	rmd	1	6	5	4	1	7000	2	2	1	1	12	1	4	1	0	1	2	3	1
dhansika	24	2	vpm	1	5	4	3	1	6000	1	2	1	1	3	1	5	1	0	1	2	3	1
malarvizhi	24	2	chen	1	6	5	4	1	6000	1	2	2	1	6	1	3	1	0	1	3	4	1
dharshan	24	1	chen	1	6	6	4	1	8000	2	2	2	2		1	1	1	0	1	2	3	1
indumathi	36	2	chen	1	6	5	4	1	8000	1	2	2	1	12	1	3.6	1	0	3	4	5	1
rishita	36	2	tivl	1	5	5	4	1	7000	1	2	2	1	6	1	2.6	1	0	2	3	4	1
keerthanasri	42	2	chen	1	6	5	4	1	8000	2	1	2	2		1	2	1	0	1	2	3	1
goopika	60	2	chen	1	5	5	5	4	9000	1	2	2	1	12	1	3	1	0	2	3	3	1
iniya	12	2	nam	1	5	5	4	1	7000	1	1	2	1	12	1	3.6	1	0	3	4	5	1
dinesh	24	1	chen	1	6	6	4	1	8000	1	2	2	1	6	1	1.6	1	0	2	3	3	1
pugalendhi	12	1	cgl	1	5	5	4	1	8000	1	2	2	2		1	2	1	0	1	3	3	1
rahul	12	1	cdl	1	4	2	3	1	6000	1	2	2	1	18	1	4	1	0	1	3	4	1
gopika	24	2	chen	1	6	5	5	1	9000	1	2	2	2		2	1	1	0	2	3	3	1
manikandan	24	1	virudh	1	5	2	4	1	8000	2	2	2	1	24	1	5.6	1	0	2	3	4	1
thavanesh	48	1	tivl	1	6	5	4	1	8000	1	2	2	1	3	1	1.6	1	0	2	3	4	1
mohamedraheem	18	1	nam	3	6	2	5	1	8000	1	2	2	1	12	1	4	1	0	2	3	4	1
nivedha	12	2	tivl	1	5	4	3	1	6000	2	1	1	1	6	1	2	1	0	1	2	4	1
radhika	24	2	pud	1	6	5	3	1	8000	1	2	2	1	12	1	4	1	0	3	4	5	1
akshaya	24	2	mad	1	6	5	4	1	8000	2	2	2	1	8	1	1.6	1	0	2	4	5	1
pratheksha	12	2	tivl	1	5	6	5	1	8000	1	2	2	1	6	1	2	1	0	2	3	4	1
ravichandran	24	1	tivl	1	2	2	2	1	5000	1	2	2	2	0	2	2.6	1	0	1	2	3	1

ARTiopapical	ARTiopmid	ARTiopbasal	ARTpopapical	ARTpopmid	ARTpopbasal	impedanceiop	impedancepop	cochleostomy	electinsert	TORCH	DQ	birthwt	birthcomp
0	0	0	0	0	0	3.87	7.51	1	2	2	79	3.4	2
0	0	0	0	0	0	4.37	8.35	1	1	2	94	2.6	2
0	0	0	1	0	1	7.2	8.8	1	1	2	84	3.2	2
1	0	1	1	1	1	4.68	8.83	2	1	2	70	2.5	2
1	0	0	1	1	0	5.02	8.36	1	1	2	86	3.2	2
0	0	1	0	0	1	4.16	8.1	1	1	1	78	2.4	1
0	0	0	1	1	1	5.9	9.1	1	1	2	76	2.2	1
0	1	1	1	1	1	6.31	10.55	1	1	2	86	3.6	2
0	0	1	0	1	1	5.02	8.4	1	1	2	73	2.6	1
0	0	1	0	0	1	4.8	6.3	1	1	1	84	3.1	2
1	1	1	1	1	1	4.6	7.6	1	1	2	77	3.2	2
1	1	0	1	1	1	4.24	6.64	1	1	1	83	3	2
0	0	0	1	1	1	4.1	7.51	1	1	1	88	2.8	2
0	1	1	0	1	1	5.2	9.21	1	1	2	82	3.4	2
1	1	1	1	1	1	5.06	8.11	1	1	1	80	2.4	2
1	1	1	0	0	1	4.04	9.34	2	1	1	88	3	2
1	1	1	1	1	1	5.12	7.84	1	1	2	94	2.8	2
1	1	1	1	1	1	5.4	8.5	1	1	1	90	2.6	2
1	0	1	0	1	1	6.2	8.2	2	1	2	84	2.8	2
1	0	1	1	0	1	4.1	8.2	1	1	1	79	3.5	2
1	1	0	1	1	0	4.12	7.12	1	2	2	87	3.1	2
0	1	1	0	1	1	5.3	7.4	2	1	2	73	3.2	2
1	0	0	1	1	0	4.6	7.4	1	1	2	83	3.4	2
0	0	0	0	0	0	4.8	6.3	1	2	2	72	3.2	2
0	0	1	0	0	1	5.5	7.6	2	1	1	94	2.9	2
0	0	0	0	0	0	3.8	7.3	2	1	1	85	2.1	1
0	1	1	1	1	1	4.7	8.2	1	1	2	84	3.2	2
1	1	1	1	1	1	3.8	6.5	1	1	2	77	1.75	1
0	1	1	0	1	1	4.8	6.2	1	1	2	75	3.6	2
1	1	0	1	1	1	3.3	5.3	2	1	2	84	3.1	2
0	0	0	0	0	0	4.5	6.2	2	1	1	74	2.1	1
0	0	1	1	1	1	5.6	7.4	2	1	2	86	2.9	2
1	1	0	1	1	1	4.8	7.2	1	1	1	75	3.1	2
1	1	1	1	1	1	5.5	8.1	1	1	1	80	2.7	1
1	0	0	1	1	1	3.9	6.3	1	1	2	84	3.5	2
1	1	0	1	1	0	6.2	9.3	1	2	2	90	3.6	2
0	0	0	1	1	1	5.9	8.5	1	1	2	82	2.9	2
0	0	1	0	0	1	4.6	6.4	1	1	1	85	3.1	2
1	1	1	1	1	1	5.3	8.7	1	1	2	76	2.1	1
0	0	0	0	0	0	4.16	8.1	2	1	1	79	2.9	1
1	1	1	1	1	1	4.8	8.8	1	1	2	76	2.4	1
0	0	0	1	1	1	4.8	6.3	2	1	2	75	2.7	2
1	1	1	1	1	1	4.04	6.76	2	1	2	82	3.3	2
1	0	0	1	0	0	3.5	7.18	1	1	2	84	3.6	2
1	1	1	1	1	1	6.34	9.16	1	1	1	79	3.2	2
1	1	1	1	1	1	5.2	9.3	1	1	2	90	3.5	2
0	0	1	0	1	1	4.16	8.02	1	1	2	90	3.2	2
1	1	1	1	1	1	5.6	8.8	1	1	2	84	3.4	2
1	1	1	1	1	1	4.91	8.66	1	1	2	82	1.8	1
0	0	0	0	0	0	4.1	7.02	1	1	2	83	2.8	2
0	0	1	0	0	1	3.45	7.65	2	1	2	84	2.9	2
1	1	0	1	1	1	4.16	8.12	1	1	1	78	3	2
1	1	1	1	1	1	6.68	9.24	1	1	2	82	2.3	2
1	1	1	1	1	1	5.45	7.66	1	1	2	80	2.4	2
0	0	0	0	0	0	5	8.5	2	1	2	82	3.6	2
1	1	1	1	1	1	4.85	8.2	1	1	2	83	3.2	2
1	1	1	1	1	1	4.83	7.83	1	1	1	86	3.1	2
1	0	1	1	0	1	6.2	8.2	2	1	1	81	2.5	1
1	1	1	1	1	1	4.83	8.26	1	1	1	88	2.6	2
1	0	0	1	1	0	4.06	8.54	1	2	2	80	2.5	2
0	0	0	0	1	1	3.98	7.85	1	1	2	79	2.8	2
1	1	1	1	1	1	5.3	9.3	1	1	2	95	3.5	2
0	1	0	0	1	1	5.33	8.5	2	1	2	80	1.8	1

INSTITUTIONAL ETHICS COMMITTEE
MADRAS MEDICAL COLLEGE, CHENNAI 600 003

EC Reg.No.ECR/270/Inst./TN/2013
Telephone No.044 25305301
Fax: 011 25363970

CERTIFICATE OF APPROVAL

To
Dr.R.Mahesh Kumari
Post Graduate in MS ENT
Madras Medical College
Chennai 600 003

Dear Dr.R.Mahesh Kumari,

The Institutional Ethics Committee has considered your request and approved your study titled " **ELECTROPHYSIOLOGY AND AUDITORY PERFORMANCE OF CHILDREN WITH PROFOUND SENSORYNEURAL HEARING LOSS AFTER COCHLEAR IMPLANT SURGERY " NO. 10062016.**

The following members of Ethics Committee were present in the meeting hold on **07.06.2016** conducted at Madras Medical College, Chennai 3

1.Dr.C.Rajendran, MD.,	:Chairperson
2.Dr.Isaac Christian Moses,MD.Ph.D.Dean(FAC)MMC,Ch-3	:Deputy Chairperson
3.Prof.Sudha Seshayyan,MD., Vice Principal,MMC,Ch-3	:MemberSecretary
4.Prof.B.Vasanthi,MD., Prof.of Pharmacology.,MMC,Ch-3	: Member
5.Prof.P.Raghumani,MS, Prof. of Surgery,RGGGH,Ch-3	: Member
6.Prof.Baby Vasumathi, Director, Inst. of O&G,Ch-8	: Member
7.Prof.K.Ramadevi,MD, Director,Inst.of Bio-Chem,MMC,Ch-3	: Member
8.Prof.M.Saraswathi,MD.,Director, Inst.of Path,MMC,Ch-3	: Member
9.Tmt.J.Rajalakshmi, JAO,MMC, Ch-3	: Lay Person
10.Thiru S.Govindasamy, BA.,BL,High Court,Chennai	: Lawyer
11.Tmt.Arnold Saulina, MA.,MSW.,	:Social Scientist

We approve the proposal to be conducted in its presented form.

The Institutional Ethics Committee expects to be informed about the progress of the study and SAE occurring in the course of the study, any changes in the protocol and patients information/informed consent and asks to be provided a copy of the final report.

28/6/16

Member Secretary – Ethics Committee

MEMBER SECRETARY
INSTITUTIONAL ETHICS COMMITTEE
MADRAS MEDICAL COLLEGE
CHENNAI-600 003

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DISSERTATION TITLE “ELECTROPHYSIOLOGY AND AUDITORY PERFORMANCE OF CHILDREN WITH PROFOUND SENSORYNEURAL HEARING LOSS AFTER COCHLEAR IMPLANT SURGERY”

1. INTRODUCTION

Hearing impairment is the most frequent sensory deficit in human populations,[1] affecting all ages from infancy to older age groups and the global burden of hearing impairment is increasing. According to World Health Organization (WHO) [2] estimates, 360 million people are living with disabling hearing loss, which constitutes about 5.3% of the world population. Among them, 328 million (91%) are adults and 32 million (9%) are children. The Disability Adjusted Life Year’s (DALYs) for hearing loss is estimated to be 1.8% of total DALYs and projected to increase to 2.9% by 2030.[3] Countries in South Asia, Asia Pacific and Sub-Saharan Africa have more number of people with disabling hearing loss. [2] The prevalence of disabling hearing loss ranges from 4.6% - 8.8% in South East Asian countries. [4] The burden of hearing impairment is substantially high in India. In our country approximately 63 million (6.3%) suffer from moderate to severe impairment. The prevalence of adult onset and childhood onset deafness is estimated to be 7.6% and 2.00% respectively.[4] Hearing disability is the second frequent cause of disability in India, the incidence being 7/100000 population. The prevalence of hearing disability is 291 persons per 100000 population, higher in rural areas than urban areas. [5] In India, one in 1000 babies are born profoundly deaf (≥90 dB in better ear) and the burden would be higher, if nearly 40,000 births per day are taken into account. [6] Hearing loss in children can be congenital or acquired. The consequences of hearing problems are well known. Many of the children with congenital permanent