DISSERTATION ON "TO EVALUATE THE CORRELATION OF ELECTROPHYSIOLOGICAL TESTS AND BEHAVIORAL RESPONSES IN COCHLEAR IMPLANT PATIENTS"

Submitted in partial fulfilment of the requirements for the award of degree of

M.S. DEGREE BRANCH-IV OTORHINOLARYNGOLOGY

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



UPGRADED INTITUTUE OF OTORHINOLARYNGOLOGY MADRAS MEDICAL COLLEGE, CHENNAI-600 003.

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CERTIFICATE

This is to certify that this dissertation entailed "TO EVALUATE THE CORRELATION OF ELECTROPHYSIOLOGICAL TESTS AND BEHAVIORAL RESPONSES IN COCHLEAR IMPLANT PATIENTS" submitted by Dr.P.NITHYA, for M.S. ENT., Branch IV Degree examination in May 2020 is a bonafide record of work done by her under my direct guidance and supervision in partial fulfilment of regulations of the Tamil Nadu Dr. M.G.R. Medical University, Chennai, Tamil Nadu, India.

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DECLARATION

THE CORRELATION OF ELECTROPHYSIOLOGICAL TESTS AND BEHAVIORAL RESPONSES IN COCHLEAR IMPLANT PATIENTS" is done by me at the Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai during 2017- 2020 under the guidance and supervision of Prof.Dr.R.MUTHUKUMAR, MS., DLO., DNB. This dissertation is submitted to The Tamilnadu Dr. M.G.R. Medical University, towards partial fulfilment of regulation for the award of M.S. DEGREE IN OTORHINOLARYNGOLOGY (BRANCH-IV)

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ABBREVIATIONS

BOA : Behavioral observation audiometry

PTA : Pure Tone Audiometery

OAE : Oto acoustic emissions

BERA : Brain stem evoked response audiometry

ART/ECAP : Auditory response telemetry / Evoked

compound action potential (ECAP)

ESRT : Electrically evoked stapedial reflex telemetry

EABR : Electrically evoked auditory brain stem

response

CAP : Categories of auditory performance score

RW : Round Window

TORCH : Toxoplasma, Rubella, Cytomegalo virus and

herpes

IQ : Intelligent quotient

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INTRODUCTION

Cochlear implants are the true bionic sense organs. They convert the sound signal to electrical signals which stimulates the nerve fibres of the vestibulocochlear nerve directly. Thus it replaces the transducer function of the hair cells of the cochlea, which was not functioning. Cochlear implantation (CI) is considered in patients with severe to profound hearing loss who are not benefited from appropriate and well-fitting hearing aids. Congenital profound hearing loss limits the ability to develop communication skills by affecting auditory and lingual development in children. Cochlear Implantation restores the important special sense. It provides greater access to sound, speech understanding, auditory abilities and linguistic development. Hearing impairment should be detected early so that implantation can be done earlier to improve the quality of life in deaf children. Hence the introduction of high-risk newborn hearing screening as a routine procedure in detection of hearing loss in early life helps in early rehabilitation with implantation in children with severe – profound hearing loss. It helps in improving the quality of life in children with hearing impairment and also earlier the implantation better the outcome in terms of speech and language development.

The success of the cochlear implant mainly depends upon the transmission of the signal to the auditory cortex through the auditory pathways from the ear, followed by appropriate central processing to ensure understanding of the signal. Hence proper functioning of device and electrodes is necessary to send signals to

the auditory nerve fibers. The multimodal electrophysiological tests are used to assess the proper functioning of the device and electrodes. These Electrophsiological tests should be done intra-operatively and post –operatively at regular intervals to assess the integrity of the device and the electrodes. The Electrophysiological tests are Electrically evoked stapedial reflex threshold (ESRT) and Auditory response telemetry (ART) intraoperatively and post – opeative ESRT ,ART and Electrically evoked auditory brain stem response (EABR). The outcome of the cochlear implant surgery is measured by several means of subjective and objective tests. The outcomes can be measured using behavioural responses using aided audiogram and auditory performance using CAP score. The purpose of this study is to evaluate to the correlation of multimodal electrophysiological tests and behavioural response in post cochlear implant patients. In our study behavioural response is assessed using aided audiogram.

AIMS AND OBJECTIVES

- 1. To evaluate the correlation of multimodal electrophysiological tests and behavioral responses in post cochlear implant children.
- 2. To evaluate the correlation of intraoperative ESRT and postoperative ESRT.

REVIEW OF LITERATURE

Articles related to our study:

Bas van den borne et al in 1996⁴ –he compared the intraoperative electrically evoked stapedius reflex thresholds with postoperative ESRT in 19 children undergoing cochlear implant. He noted that intraoperative thresholds were higher compared to the postoperative thresholds. The increase in intraoperative ESRT thresholds is due to the influence of anaesthetics used during surgery.

S.Mason et al in 2004 ⁹– conducted a study in 427 cases of cochlear implantation patients. He reviewed the electrophysiology and objective measures as monitoring tool for cochlear implant in operating room and their value in management of children. He did impedance telemetry, ESRT and ART. The results showed that intraoperative measures provided valuable assistance in the initial fitting of the device. The normal intraoperative findings provided immediate reassurance to the parents and the implant team that the implant was functioning fully and the stimulation activates the auditory pathways.

Kosaner et al in 2008 ¹⁸ – studied about use of ESRT in fitting of cochlear implant speech processor in young children. ESRT can be measured easily and quickly and it correlates with behaviourally measured maximum comfort levels.

Cosettei et al in 2010³¹-analysed the intraoperative neural response telemetry as a predictor of outcome in cochlear implant patients. He followed 24 children and 73 adults who has undergone cochlear implant over a period of 1year. He stated

that no significant correlation between NRT and performance at 1 year. Also absence of NRT does not indicates a lack of stimulation.

Kim et al in 2010²¹ –included 17 cochlear implant patients in their study. They evaluated the relationship between the electrically evoked compound action potential and speech perception. He concluded that ECAP has significant correlation to the performance with cochlear implant. ECAP measures useful to predict the outcomes with cochlear implant.

Kartas et al in 2011¹⁹ – he studied about intraoperative electrically evoked stapedius reflex thresholds (ESRT) in children undergone cochlear implantation. He compared the electrical stimulation thresholds using ESRT among the Round window and cochleostomy approaches. He concluded that the duration of electrically stimulation thresholds were shorter in round window approach compared to cochleostomy approach. ESRT measurements were recorded at lower threshold in the round window approach compared with the cochleostomy group. Hence he resulted that round window approach insertion offers best electrically stimulation relative to electrode insertion than cochleostomy.

Baysal et al in 2011¹- conducted a study with 65 prelingual hearing loss children undergoing cochlear implant. He studied about the correlation between intra- and postoperative electrically evoked stapedius reflex thresholds (ESRTs) in children with cochlear implants. He concluded that intraoperative ESRT measurements were unable to predict early postoperative ESRT. A correlation analysis did not

reveal any statistically significant correlation between intra- and postoperative ESRTs.

Oana manolache et al in 2012 ⁶-investigated 72 cochlear implant patients over a period of 3 months. He measured the electrode impedance variations in patients with cochlear implant. He noticed that increase in impedance in the post operative period. The increase in impedance in all electrodes is due to the absence of electrical stimulation, during the time between surgery and the device activation.

Goering et al (2013) ⁷- studied about the intraoperative ad postoperative cochlear implant electrodes impedance among 165 paediatric and adult patients. He concluded that intraoperative high impedance have a probability of resolving by initial activation. Surgical techniques or complications results in increased incidence of air bubble in cochlea play a role in abnormal intraoperative impedance results.

Kelly cristina lira de Andrade et al 2014 ⁵ – studied the importance of Electricaly evoked stapius reflex threshold (ESRT) in cochlear implants. He concluded that ESRT is useful in programming the cochlear implant, especially in patients with inconsistent responses.

Mohammed said abdelsalam et al in 2015 ²³-studied about electrically evoked auditory brain stem response in cochlear implant children. They conducted in 30 children undergone cochlear implantation. He stated that EABR proves to be effective method to evaluate the auditory function in children. There was a

positive correlation between EABR wave latencies and the age at implantation. Children who undergo implantation at younger ages tend to achieve higher levels of speech perception.

Kosaner et al in 2017 ¹⁸ – conducted a study in 52 paediatric cochlear implant patients, they compared the ESRT and ECAP measurements in those children. They concluded that ESRT were significantly higher than the ECAP thresholds.

Makhdoum et al in 2018 ³– conducted a study about the effect of volatile and intravenous anesthesia over stapedial reflex threshold. He concluded that use of inhalational anaesthetics affects the stapedius muscle contraction in the intraoperative period.

Bayrak et al in 2019 ²²conducted a study among 16 children who underwent cochlear implant, regarding the relationship between the electrically evoked compound action potential and electrically evoked auditory brain stem responses. He concluded that consistency was found between ECAP and EABR recordings. But one cannot be preferred over the other because the data quality of the two tests was different.

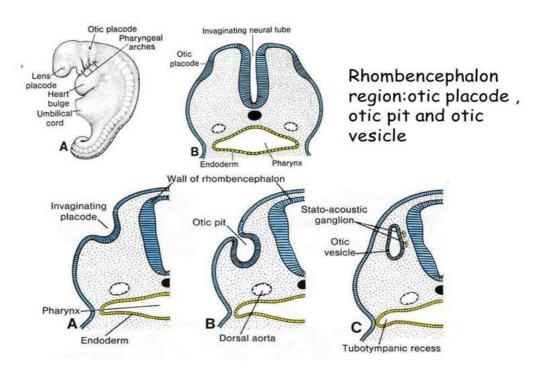
Mariappan et al in 2019 ³² –conducted a study among 21 cochlear implant children. EABR were recorded in all cochear implanted children.the morphology ,amplitude and latencies of the waveforms were analysed.he concluded that the apical electrode tends to show steeper (amplitude) and earlier (latency) EABR waveforms than the middle and basal electrode. These differences between the apical and other electrodes reflect the relative difference in the density of

surviving spiral ganglions and the possible difference in the neurophysiology of the nerves at different regions of the cochlea.

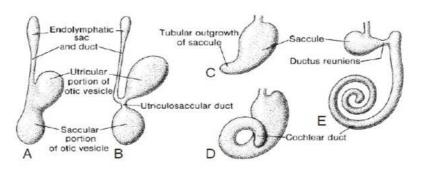
INNER EAR:

EMBRYOLOGY OF THE INNER EAR AND AUDITORY NERVE:12

Inner ear developed from ectoderm in the region of hindbrain. Thickening of the ectoderm (Auditory placode or otic placode) becomes invaginated to form Auditory (otic) vesicle. The otic vesicle detached from the surface and a layer of mesoderm surrounds it. It sinks in to the mass of mesoderm which is rudiment of petrous bone. The otic vesicle draws a tail behind it which is rudiment of ductus endolymphaticus. Saccus endolymphaticus develops as an expansion of distal end of ductus endolymphaticus.



The membranous labyrinth is formed from otic vesicle and it is the first part of ear mechanism to make its appearance.



A and B. Development of the otocyst showing a dorsal utricular portion with the endolymphatic duct and a ventral saccular portion.

C to E. Cochlear duct at 6, 7, and 8 weeks, respectively. Note formation of the ductus reuniens and the utriculosaccular duct.

By the 6th week of embryonic life – three semicircular canals are well formed. The ampullated ends becomes clearly defined. The dependent portion of vesicle not only elongated as cochlear pouch began to assume its snail shell coil.

By the end of 1st month –only the endolymphatic space developed. The perilymphatic space not developed.

The first perilymphatic space to form is just within the oval window in vestibule, the cisterna perilymphatica, occurs in 3rd fetal month. The second perilymphatic space is within round window is the scala tymani. The aqueductus of cochlea develops relatively late out pouching from subarachnoid space.

The Neuroepithelial structures of membranous labyrinth are basically similar in type, but modified in form in accordance with final respective function.

By 7th week – Macula develops from utricular and sacular epithelia at points where nerve enters their walls.

By 8 weeks- Epithelium of cochlear duct begins to differentiate in to basal turn then followed by middle and apical turns.

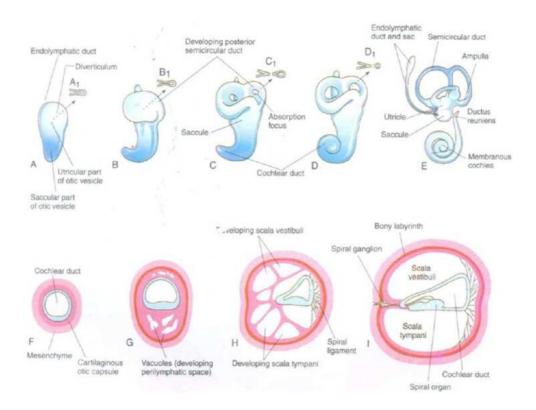
By 12th week – Differentiation of hair cells and supporting cells occurs. The organ of corti and tectorial membrane are recognisable in basal turn.

By 14th and 16th week –Otoconia appeared in gelatinous layer.

Crista ampullareis also forms at point where nerve fibres enter the ampulla of semicircular canals. They begin to develop at sametime as macula but instead of remaining flat, becomes elevated into the ridge covered by gelatinous cupula.

By 4 th month - Cochlea is almost adult form.

Inner ear is the only organ reaches adult size and complete differentiation by midterm, even before tiny fetus become a viable premature infant. Last to differentiate in labyrinth is recently acquired cochlearend organ (more subject to anamolies) than older vestibular organ.



DEVELOPMENT OF OTIC CAPSULE:

Otic capsule develops from mesoderm which surrounds the membranous labyrinth.

Mesoderm Precartilage True cartilage membranous labyrinth at 2nd month of fetal life ossification of otic capsule (5t month of fetal life)

Dedifferentiation and ossification of otic capsule occurs by process of incrustation ,from this onwards otic capsule is known as pars petrosa. The pars petrosa becomes encased with dermal or membranous bone ,so at birth 3 distinct layer of bones are discernible.

Outer layer- periosteal bone (which is lamellar in type)

Inner layer – Endosteal bone (also of lamellar type). It lines the contour of labyrinth and it is relatively thin.

Between the two layers of endosteal and periosteal bone a enchondral bone is present, which is charecterised by presence of cartilage cell rests/ Globuli interossei.

DEVELOPMENT OF AUDITORY NERVE: 12

The rudimentary eighn nerve appears in 4th week as Auditory ganglion, which lies between auditory vesicle and wall of hindbrain. At first it fused with ganglion of 7th cranial nerve (Acousticofacial ganglion) later it two separates. The cells of ganglion derived from neural crest—cells and also from neuroectoderm of auditory vesicles. The Auditory ganglion divides into vestibular and cochlear part, each associated with corresponding division of 8th cranial nerve. By 7^{tarah} week, cochlear nerve is laid down. Spiral ganglia recognised at 8th week. The spiral ganglion and cochlear nerve has been linked up with their sensory end organ by 12th week.

ANATOMY OF THE INNER EAR:

THE VESTIBULAR SYSTEM:

The vestibular system can be generally divided into two parts: the saccule and utricle. The saccule is anatomically a separate chamber from the utricle. The three semicircular canals which arise from and terminate in the utricle. They arehorizontal (lateral) scc, posterior scc and superior (anterior) scc. The utricle and

semicircular canals are evolutionarily and developmentally separate from the saccule.

THE COCHLEA:

GROSS ANATOMY: 30

The cochlea is formed of three parallel canals coiled in a spiral around a central 'stalk called the modiolus. The axons of the central projections of the auditory nerves that innervate the sensory epithelia, and the vessels of the cochlear blood supply, the cochlear artery and cochlear vein, run through the length of the modiolus. There are 2.5 turns in the cochlea.

The central canal, the scala media, is lined by epithelia (part of the membranous labyrinth) and is filled with endolymph. In cross sections of the scala media, appears triangular in shape the scala media is bounded by three walls. The basilar membrane, Reissners membrane and stria vascularis. The sensory epithelium, the **organ of Corti,** running along the **basilar membrane** which forms the floor of the triangle. The primary ion-transporting epithelium, the **stria** forms **vascularis**, forms the lateral side of the triangle and **Reissner's membrane** the roof of the triangle. The **scala vestibule is present above the Reissner's membrane** and under the basilar membrane the **scala tympani was present.**. These two scalae vestibule and tympani are filled with perilymph. Reissner's membrane acts as the barrier between endolymph and perilymph in the scala vestibuli. The Perilymph is freely permeable into the intercellular spaces of the spiral ligament that underlies the stria vascularis but there is a barrier for the direct diffusion of ions

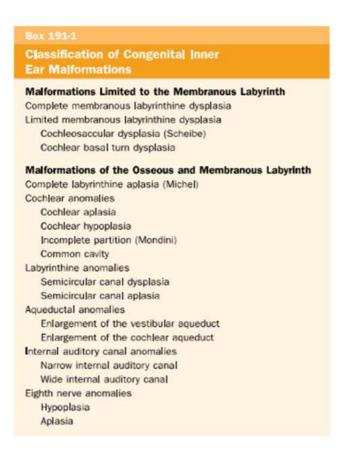
from the spiral ligament into the ion-transporting epithelium. The height and width of all the three scala vestibule, scala media and scala tympani decrease systematically from base to apex of the spiral. At the basal end, the scala tympani terminates at the **round window**. It is covered by secondary tympanic membrane formed of two epithelial sheets sandwiching connective tissue, containing collagen and blood vessels. The apical surface of the outer epithelium is exposed to air in the middle ear, whereas the inner epithelium is bathed in perilymph. The scala vestibuli at its basal end is continuous with the vestibule and the perilymphatic compartment of the vestibular system. The oval window, opening over the vestibule, is covered by footplate of the stapes and annular ligament. At the apical end of the cochlea, the scala media is closed by epithelial tissue, arising partly by extension of Reissner's membrane, leaving a small opening, the helicotrema, through which the scala vestibuli and scala tympani are connected. Sound-induced movements of the tympanic membrane drive pistonlike 'in-out' movements of the stapes footplate displacing incompressible perilymph along the scala vestibuli, through the helicotrema and down the scala tympani leading to 'out-in' movements of the round window. As fluid is displaced, the pressure difference across the scala media between the scala vestibule and scala tympani, produces vibrational movement of the basilar membrane, described by Von Bekesy. This 'travelling wave' stimulates the sensory cells housed in the organ of Corti that sits on the vibrating basilar membrane.

CONGENITAL MALFORMATION OF THE INNER EAR:

Most of the inner ear malformations arise due to the interruption in the formation of membranous labyrinth during first trimester of pregnancy. The syndromes associated with radiologically detectable inner ear malformation includes Waardenburg, Wilderwanck, Apert, Pendred, brancio-oto-renal syndrome. In utero viral infecions like rubella and cytomegalovirus can cause inner ear malformations.

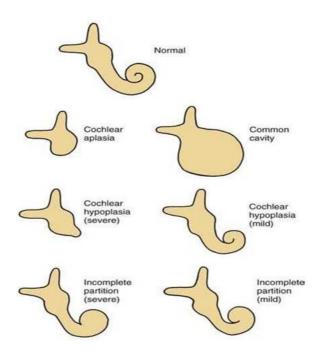
Congenital malformations broadly divided in to two categories – malformations limited to membranous labyrinth and malformations of both osseous and membranous labyrinth.

Sennaragolu classification of congenital inner ear malformations:³³



Type of malformation	Gestational week of origin	Malformation	
Complete labyrinthine aplasia	3 rd week	Complete absence of inner ear structures	
Cochlear aplasia	Late 3 rd week	Absent cochlea with normal or deformed vestibule and semicircular canals	
Common cavity	4 th week	Cochlea and vestibule in single cavity no internal architecture	
Type 1 incomplete partition	5week	Cystic cochlea vestibular malformation with absent moidiolus.	
Cochlear hypoplasia	6week	Small cochlear bud with less than one turn	
Type 11 incomplete 7week partition		Cochlea with normal basal turn with cystic apex	

CONGENITAL MALFORMATION OF THE INNER EAR:



HEARING LOSS: 30

Hearing loss is a common problem affecting all age groups in the world. It leads to marked disability. The World Health Organization (WHO) lists hearing loss in the 20 leading causes of burden of disease.

In 2012, the WHO estimated that there are 360 million children worldwide with a disabling hearing loss. It accounts for 5.3% of the world's population. Of these, 91% are adults and 9% children. The prevalence of hearing loss is high in South Asia, Asia Pacific and Sub-Saharan Africa both for children and for adults over the age of. The prevalence increases with age: in children the hearing loss is 1.7%, 7% in individuals over the age of 15. The hearing loss in adults over the age of 65 it is almost 1 in 3. However that half of all cases of hearing loss can be avoided through primary prevention.

LEVELS OF PREVENTION OF HEARNG LOSS: 30

1)Primary prevention:

- Genetic
 - Genetic counselling
- Infective (congenital or acquired)
 - Immunization
 - Early treatment

 Avoidance and education
• Traumatic (noise, physical trauma and barotrauma)
– Avoidance
- Early treatment
• Ototoxic medications
- Avoidance
- Monitoring
- Treatment
2) Secondary prevention:
• Screening
• Treatment
3) Tertiary prevention:
• Early rehabilitation of hearing loss.
CAUSES FOR HEARING LOSS: 30
The hearing loss can be genetic / Hereditary and Environental / Acquired
is further divided into syndromic and non-syndromic hearing loss. Non-
syndromic hearing loss manifestations accounts for about 70% of genetic hearing

loss. The common cause of genetic deafness is mutations in the gap junction beta

2 gene (*GJB2*) located in the chromosome 13q.It encodes for the protein connexin 26. Hearing disability is the second most common cause of disability in India. The incidence is 7/100000 population. The prevalence of hearing disability is 291 persons /100000 population. The prevalence is higher in rural areas than urban areas. In India, 1 out of 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 considered.

It has been noted that 80% of deafness is avoidable ,50% is preventable and about 30% treatable or can be managed with assistive devices. The current 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 fit for hearing aid.

CAUSES FOR HEARING LOSS:

50% Environmental		cytomegalovirus (CMV) meningitis rubella prematurity neonatal icterus ototoxicity (some are genetic susceptibilities) other infections	
50% Genetic	30% Syndromic	Alport Norrie Usher Pendred Waardenburg branchio-oto-renal Jervell and Lange-Nielsen	
	70% Nonsyndromic	Autosomal Dominant (DFNA1 - DFNA3) Autosomal Recessive (DFNB1 - DFNB30) X-Linked (DFN1 - DFN8 Mitochondrial	

CAUSES FOR NON- SYNDROMIC HEARING LOSS:

Autosomal Dominant Syndromes	Autosomal Recessive Syndromes	X Linked Disorders	Mitochondrial Syndromes:
1. Waardenburg	1.Pendred	1.Alport	1.MELAS
Syndrome	Syndrome	Syndrome	Syndrome
2. Branchio-oto-renal	2.Usher's	2.Mohr-	2.MERRF
Syndrome	Syndrome	Tranebjaerg	Syndrome
3. Stickler's Syndrome	3.Jervell and	Syndrome	
4. Neurofibromatosis	Lange-Nielson	3. Otopalatal –	
Type 2	Syndrome	digital	
5. Treacher Collins	4.Biotidinase	syndrome.	
Syndrome	deficiency	4.Norrie	
6. Apert syndrome	5.Refsum's	syndrome	
, ,	disease		

MATERNAL FACTORS FOR HEARING LOSS:

(a) Infections during pregnancy:

Infection that affects the developing foetus are toxoplasmosis, rubella, cytomegaloviruses,herpes type 1 and 2 and syphilis(TORCHES).

(b) Drugs during pregnancy:

Streptomycin, gentamicin, tobramycin, amikacin, quinine or chloroquine injestion during the antenatal period cross the placental barrier and causes damage to the cochlea. Thalidomide not only affects ear but also causes abnormalities of limbs (phacomelia) ,heart, face, lip and palate.

(c) Radiation to mother in the first trimester.

(d) Other factors: Nutritional deficiency, diabetes, and thyroid deficiency.

Maternal alcoholism is also teratogenic to the developing auditory system

PERINATAL CAUSES FOR HEARING LOSS:

a)ANOXIA:

It damages the cochlear nuclei and causes haemorrhage into the ear. Placenta praevia, prolongedlabour, cord round the neck and prolapsed cord all these can cause foetal anoxia.

b)PREMATURITY AND LOW BIRTH WEIGHT:

Infant born before term or with birth weight less than 1500 g (3.3 lb).

c) BIRTH INJURIES:

Birth injury due to forceps delivery may cause intracranial haemorrhage with extravasation of blood into the inner ear.

D) HYPERBILIRUBINEMIA(NEONATAL JAUNDICE):

Bilirubin level more than 20 mg% damages the cochlear nuclei.
E) NEONATAL MENINGITIS
F) SEPSIS
G) OTOTOXIC DRUGS.
AUDIOLOGICAL ASSESSMENT:
SUBJECTIVE TESTS:
1)Neonatal screening procedures
• ABR/OAEs

- Arousal test
- Auditory response cradle
- 2) Behaviour observation audiometry
- Moro's reflex
- Cochleopalpebral reflex
- Cessation reflex

3) Distraction techniques (6–18 months)

4) Conditioning techniques (7 months – 2 years)

- Visual reinforcement audiometry
- Play audiometry (2–5 years)

5) Pure tone audiometry.

OBJECTIVE TESTS:

- Impedance audiometry
- Otoacoustic emissions
- ABR

1) Screening Procedures:

Arousal test:

A high-frequency narrow band noise is given for 2 s to the infant when he/she is in sleep. A normal hearing infant can be aroused twice when 3 such stimuli are presented to him.

Auditory response cradle: It is a screening device for newborns, where baby is placed in a cradle and his trunk and limb movement, head jerk in response to auditory stimulation are monitored by transducers. It is useful in screening the babies with moderate, severe or profound hearing loss.

2)BEHAVIOUR OBSERVATION AUDIOMETRY(BOA):

Auditory signal is presented to an infant produces a change in behaviour is noted. For example cessation of an activity, widening of eyes or facial grimacing.

Moro's reflex: sudden movement of limb and extension of head in response to sound of 90dB

Aurapalpebral reflex or cochleopalpebral reflex: blink to loud sound

Cessation/Initiation reflex:

Baby starts crying in response to sound of 90 dB or stops his activity.

3) DISTRACTION TECHNIQUES:

Distraction techniques are used in children 6–7 months old. The child at this age turns his head to localise the source of sound. In this test, the child is seated in the mother's lap, an assistant distracts the child's attention then the examiner produces a sound from behind or from one side to see if the child tries to locate it. Sounds used for distraction testing are high frequency rate (8 kHz), low-frequency hum, warbled tones or narrow band noise (500–4000 Hz).

4) CONDITIONING TECHNIQUES:

Visual reinforcement audiometry:

The Child is trained to look for an auditory stimulus by turning his head & the child is conditioned for sound with visual stimuli. It is done in children 6-24

months. This test helps to determine the hearing threshold using standard audiometric techniques. The sound stimulus is delivered by headphones or by insert earphones which are accepted better in children and are also light weight.

Play conditioning audiometry:

Play conditioning audiometry used in children between 2-5 years. The child is conditioned to perform an act each time after the child hears the sound signal. The act can be placing a marble in a box, putting a ring in the stand, plastic block in a bucket each time he/she hears a sound signal. For the correct performance of the act is reinforced with praise, encouragement or reward. Ear specific thresholds can be determined by standard audiometric techniques.

Speech audiometry:

In Speech audiometry the child is asked to repeat the names of certain objects or to point them out on the pictures. The voice can be gradually lowered. In such a way the hearing level and speech discrimination can be assessed.

5) PURE TONE AUDIOMETRY:

It is a subjective test used to identify the type, degree and configuration of hearing loss. An audiometer is an electronic device which produces pure tones, the intensity of the pure tone can be lowered to 10 to 15db steps until it becomes audible or increased in 5 dB steps till the patient responds. This procedure is repeated till hearing threshold is obtained and it is known as "up 5 down 10 technique". Usually air conduction thresholds are measured in 125, 250, 500,

1000, 2000, 4000 and 8000 Hz frequencies, whereas the bone conduction thresholds are measured in 250, 500, 1000, 2000 and 4000 Hz. The amount of intensity that has to be raised above the normal level is a measure of the degree of hearing impairment at that frequency. The results are charted in the form of a graph called audiogram. Pure tone average is simply a mean of air conduction threshold at 500, 1000, 2000Hz.

Uses of Pure Tone Audiogram

- (a) It is used to measure the threshold of hearing loss by air and bone conduction and the degree and type of hearing loss.
- (b) A record can be kept for future reference.
- (c)PTA is useful in the prescription of hearing aid.
- (d) Helps to find degree of handicap for medicolegal purposes.

OBJECTIVE TESTS:

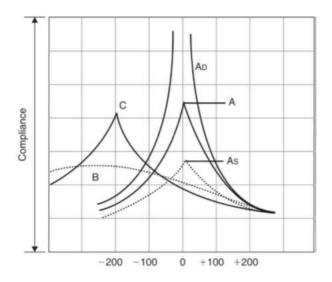
• Impedance audiometry:

Impedance audiometry consists of Tympanometry, Eustachian tube function tests, Acoustic reflex tests. It is based on a simple principle, when a sound strikes tympanic membrane, some of the sound energy is absorbed and some is reflected. The reflection of sound energy is more if the tympanic membrane is stiff than a compliant one. By changing the pressures in a sealed external auditory canal and then measuring the reflected sound energy, it is possible to

find the compliance or stiffness of the tympano-ossicular system and thus helps in finding the status of the middle ear. The equipment consists of a probe which snugly fits into the EAC and has three channels: (i) one to deliver a tone of 220 Hz, (ii) another to pick up the

reflected sound through a microphone and (iii) to bring about changes in air pressure in the ear canal from positive to normal and then negative. The results are interpreted in a chart.

TYMPANOGRAM:



Type A - Normal tympanogram.

Type As - Compliance is lower at or near ambient airpressure. Seen in fixation of ossicles, e.g. otosclerosis, malleus fixation.

Type Ad - High compliance at or near ambient pressure. Seen in ossicular discontinuity or thin and lax tympanic membrane.

Type B - A flat or dome-shaped graph. No change in compliance with pressure changes. Seen in middle ear fluid or thick tympanic membrane.

Type C - Maximum compliance occurs with negative pressure in excess of 100 mm H2O. Seen in retracted tympanic membrane.

ACOUSTIC REFLEX:

Stapedius muscle contracts in response to sound of 70 -100 dB above the threshold of hearing of a particular ear and this reflex can be recorded. Tone can be presented to one ear and the reflex picked from the ipsilateral or the contralateral ear.

The reflex arc involved is:

Ipsilateral: CN VIII \rightarrow ventral cochlear nucleus \rightarrow CN VII nucleus ipsilateral stapedius muscle.

Contralateral: CN VIII → ventral cochlear nucleus →contralateral medial superior olivary nucleus → contralateral CN VII nucleus → contralateral stapedius muscle.

CLINICAL USES:

- To test the hearing in infants and young children. It is an objective method.
- To find malingerers. A person who feigns total deafness and does not give any response on PTA but shows a positive stapedial reflex is a malingerer.

- To identify cochlear pathology. Presence of stapedial reflex at lower intensities, e.g. 40–60 dB than the usual 70 dB indicates recruitment and thus a cochlear type of hearing loss.
- Stapedial reflex decay: To identify VIIIth nerve lesion. If a sustained tone of 500 or 1000 Hz, delivered 10 dB above acoustic reflex threshold, for a period of 10 s, brings the reflex amplitude 50%, it shows abnormal adaptation and is indicative of VIIIth nerve lesion.
- Level of lesions in facial nerve disorders: Absence of stapedial reflex when hearing is normal indicates lesion of the facial nerve, proximal to the nerve to stapedius. The reflex can also be used to find prognosis of facial paralysis, the appearance of reflex, after it was absent, indicates favourable prognosis.
- Identify the lesion of brainstem.- If ipsilateral reflex is present but the contralateral reflex is absent, lesion is in the area of crossed pathways in the brainstem.
- To identify whether the lesion is cochlear or retro-cochlear in sensorineural deafness
- Also used in objective differentiation between conductive and sensorineeural hearing loss.
 - Absent of acoustic reflex + normal tympanometry = SNHL
 - Absent of acoustic reflex + abnormal tympanometry = CHL

OTOACOUSTIC EMISSIONS (OAE):

Primary purpose of OAE is to determine cochlear status specifically hair cell function. OAE's are low intensity sounds produced by outer hair cells of normal cochlea. The sound emitted by the normal cochlea can be picked up and measured by the receiver placed in the deep external auditory meatus.

TYPES OF OAE:

- i) Spontaneous OAE
- ii) Evoked OAE Stimulus frequency OAE, Transient evoked OAE and Distortion product OAE.

SPONTANEOUS OAE'S:

Spontaneous OAE's are narrow band sounds emitted from the ear in the absence of stimulation. They are present in about 50% of normal subjects where hearing loss does not exceed 30Db. The limitations are they are found in different frequencies in different ear, amplitude varies over time, found in relatively restricted range of frequencies. they ay be absent in 50% individuals.

EVOKED OAE'S:

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

a) STIMULUS FREQUENCY OTOACOUSTIC EMISSIONS:

Stimulus frequency OAE are produced 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 EMISSION:

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 outer hair cell travels via the middle ear and external auditory canal which is picked by the microphone and is recorded graphically in a moving strip of paper. A series of click stimuli are presented at 80-85 dB SPL and response recorded. TEOAE'S are obtained in all normal individuals including newborn. It is reduced in factors causing hearing losses such as ototoxic drugs, hypoxia &noise exposure. It is absent in cochlear SNHL 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:

DPOAE's are produced by presenting two tones of different frequencies presented 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.

Clinical Uses of OAE:

- It is used as a screening test of hearing in neonates and to test hearing in uncooperative or mentally challenged individuals after sedation. Sedation does not affects the OAEs.
- 2) They helps to differentiate cochlear from retrocochlear hearing loss. OAEs are absent in cochlear lesions. For example sensorineural hearing loss due to ototoxic drugs. They detect ototoxic effects earlier than pure tone audiometry.
- 3) OAEs are also useful to diagnose retrocochlear pathology, especially auditory neuropathy. Auditory neuropathy is a neurologic disorder of CN VIII. The patient with Auditory neuropathy presents with absent or abnormal auditory brainstem response, show a retrocochlear type of lesion but OAEs are normal

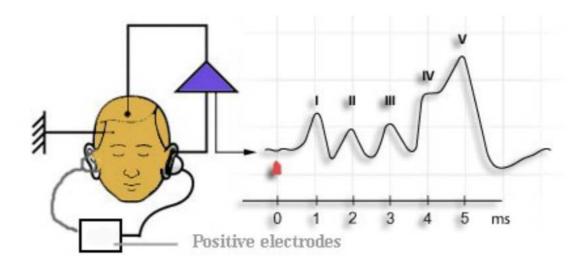
BRAIN STEM EVOKED RESPONSE AUDIOMETRY:

Brain stem evoked response audiometry (BERA) is a useful objective hearing assessment in infants. It is a measurement of synchronous neural activity of auditory nerve and brain stem in response to acoustic stimulus. The advantage of this procedure is its ability to test even infants in whom conventional audiometry may not be useful. This investigation can be used as a screening test in high risk infants. First described by Jewett and Williston .It is a neurological test of auditory brainstem function in response to click (auditory stimuli). The result is recorded in forms waveform 1-V11 ,generated at the level of brain stem in

response to click / tone impulse by placement of electrodes in the scalp. Stimulus is given by a transducer placed in the insert ear phone / head phone.

ELECTRODE PLACEMENT IN BERA:

The electrodes used to record BERA should be placed over the scalp so the scalp hair should be free of oil. Patient should be instructed to give shampoo bath to the hair on the day of the Investigation. The non inverting(Active electrode) electrode is placed over the vertex of the head, and the inverting electrode(Reference electrode) is placed over the ear lobe of ipsilateral ear or mastoid prominence. The Ground electrode or earthing electrode is placed over the forehead or the contralateral mastoid. This earthing electrode is important for the proper functioning of preamplifier. Electrodes that are placed over the mastoid process or ear lobe should be symmetrical.



RECORDING OF THE RESPONSE IN BERA:

BERA can be measured from 28 wks of gestation. Sound stimulus in the form of

clicks or tone burst stimuli are presented at an intensity level >90DB. The level is

then lowered and the responses is tracked until an intensity is reached at which

the response is no longer observable. The response observed is identification of

peak V wave. Multiple recordings were carried out to check the replicability and

morphology of the peak V wave obtained. The hearing threshold is assessed

using BERA. The minimum intensity at which wave V is traced is considered as

the hearing threshold.

Wave I Distal part of CN VIII

Wave II Proximal part of CN VIII near the brainstem

Wave III Cochlear nucleus

Wave IV Superior olivary complex

Wave V Lateral lemniscus

Waves VI and VII Inferior colliculus

USES OF BERA:

• Auditory brain stem response is a valuable objective measurement of hearing in

newborn infants, mentally challenged patients and malingering individuals.it is

used in estimation of hearing threshold in individuals who cannot be tested by

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behavioral methods.BERA is unaffected by sleep or sedation hence infants can be sedated before performing this test.

- It is used to differentiate central or peripheral disorders,
- Also used to identify the site of lesion in retrocochlear pathologies.
- It can be used in premature infants more than 30wks of gestation.
- Used to assess the maturity of Central nervous system in newborns.
- objective identification of brain death
- Used to assessing prognosis in comatose patients.
- It is detect demyelinating lesions involving auditory pathways.
- Also used to detect lesions and tumors involving auditory pathway.
- It helps the neurosurgeon in intraoperative period for the monitoring of the audiotory vestibular system during extensive neurosurgical procedures involving this area

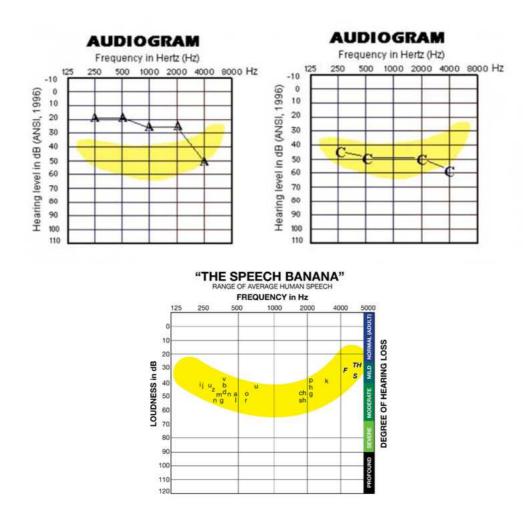
AIDED AUDIOGRAM:

The audiogram is performed with hearing aid or cochlear implants called an aided audiogram. The purpose of this testing is to determine if sounds can be detected by the child with the device on. A speech banana is a banana shaped range on a audiogram that covers the frequencies and decibel that are needed to understand speech. Audiologists are concerned with hearing loss that occurs

within speech banana because it can slow the development of a child's speech abilities and language development.

After cochlear implantation the child's behavioural response is assessed using aided audiogram, tones are presented to the implanted ear and the responses are recorded and plotted in a graph. The symbols of aided audiogram, represents the hearing level with amplification with hearing aids [A] or cochlear implants [C or CI].

The main goal of cochlear implant is to give hearing to all the sounds in the area of speech banana.



HISTORY OF THE EVOLUTION OF COCHLEAR IMPLANT

Cochlear implants are the 1st true bionic sense organs. Cochlear implants are not hearing aids. The fundamental concept of cochlear implants- to bypass the damaged hair cells. The device bypasses damage parts of auditory system and directly stimulates auditory nerve fibres (spiral ganglion cells) allowing individuals who are profoundly deaf to receive sounds. Many developments and newer technologies were made in the development of cochlear implant.

The history of cochlear implant begins with the use of electricity to stimulate the ear in an attempt to produce a sensation of sound.

- Volta was the first to experiment who tried stimulating ear with 30-40 connected cells connected to 2 metal rods.
- Ritter repeated it but with 100-200 cells in 1801.
- Stevens at Harvard, in 1937 through his experiments found 3 mechanisms for production of sound.
- 1.Direct stimulation of auditory nerve.
- 2.Stimulation of cochlear receptors at different frequencies.
- 3.Mechanical vibrations that stimulate the auditory organs.
- In 1957 usher ,during the surgery for facial nerve grafting for a case of cholesteatoma with facial nerve palsy , an electrode was implanted into the

stump of the remaining cochlear nerve. The patient was able to hear sound and differentiate frequency and intensity.

- Djourno is the first person to describe using promontory stimulation with a transtympanic needle to verify a functioning cochlear nerve.
- Chouard developed one of the early multichannel implants.
- House first implanted 2 patients with a single electrode system into the scala tympani in 1961
- Simmons, in 1964 and 1966, placed multiple electrodes into human subjects with no adverse effects.
- Clark developed an early multichannel implant using biphasic current stimulation in 1967
- Cochlear implant development at the Technical University of Vienna was started by Ingeborg and Erwin Hochmair in 1975
- FDA began regulation of cochlear is implant in 1980
- FDA approved the use of cochlear implant in adults in 1984
- FDA approved the use of cochlear implant in children above 2 years old in 1990
- FDA approved the use of cochlear implant in children above 18 months and above in 1998

- FDA approved the use of cochlear implant in children above 12 months old in 2002
- In 2005 first three recipients were implanted with cochlear's TIKI device, a
 totally implantable cochlear implant in Melbourne, Australia as a part of a
 research project.

COCHLEAR IMPLANT:

Cochlear implants are surgically placed electronic device that restore hearing sensation in people with severe to profound hearing loss, who has minimal or no benefit with the hearing aids.

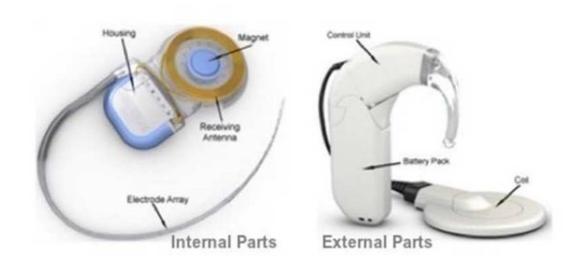
Parts of cochlear implant:

It consists of external and internal components. The external components consists of Microphone, Speech processor, Transmitting coil. The internal components consists of Receiver/Stimulator and Electrode array

Mechanism of cochlear implant:

Sound signals received by microphone are sent to speech processor. Speech processor analyzes and digitizes the sound impulses into coded signals. Coded signals sent into transmitting coil. Transmitter sends the code across the skin to the receiver/stimulator of the internal part. Signals sent to the electrodes to stimulate the nerve fibers. Signals are recognized as sounds by the brain producing a hearing sensation.





CODING STRATEGY:

Defined by which pitch, loudness and timing of sound are translated in to a series of electrical impulses..All three FDA approved devices are capable of using more than one type of strategy.

Two types of strategies are Simultaneous strategy and Non-simultaneous strategy

SIMULTANEOUS STRATEGY:

This strategy activates more than one electrode at the same time.it Provides a more natural quality of sound. Only advanced bionics is capable of SS. Disadvantage - when two electrodes are activated simultaneously there is chance of signal interference (channel interaction). So modiolus hugging electrode are developed- lies close to spiral ganglion, less intensity sound is required for activation, hence less chances of channel interaction.

NON-SIMULTANEOUS STRATEGY:

The non –simultaneous strategies are CIS(continued interval sampling),ACE (advanced combination encoder),SPEAK (spectral peak).CIS stimulates each active electrode serially in turn one after the other.No electrode is stimulated or bypassed out of order. Each electrode stimulates different frequency within the cochlea, the cochlea receives complete information about the frequency composition of incoming signal. Upto a certain point, the rapidity with which stimulation occurs leads to improved speech recognition. All three FDA approved devices use CIS strategy, but rates at which stimulation occurs are different.

COMPANIES MANUFACTURING COCHLEAR IMPLANT:

- Nucleus
- Advanced bionics
- MED EL

ELECTRODE OPTIONS IN COCHLEAR IMPLANT:

- Straight electrodes
- Peri modiolar electrodes /Contour advance electrodes
- Electrodes for malformed cochlea.

STRAIGHT ELECTRODES

- The longevity and efficacy is more in straight electrodes.
- It is used in patients with variety of anatomical variation, when structure of cochlea is not suitable for perimodiolar electrodes.
- Combined electric and acoustic stimulation have also been established for which electrode design is critical in preservation of residual hearing.
- The hybrid electrodes are straight with limited length to avoid intracochlear trauma.

PERIMODIOLAR ELECTRODES

- These electrodes are designed to coil during or after insertion to occupy a position closer to the modiolar wall of the cochlea where the spiral ganglion cells reside.
- Advantages :
- More selective stimulation of spiral ganglion sub-populations.

- Less current required for each stimulus thereby reducing power consumption.
- Less damage to the cochlear elements.
- These lead to better speech understanding, longer battery life, and preservation of residual hearing

ELECTRODES FOR MALFORMED AND OBSTRUCTED COCHLEA

- All the available electrodes can be used for obstructed cochlea; however helix electrode may not be desirable due to the larger diameter tip.
- The med-el split electrode or the cochlear corporation double array is used with the dual cochleostomy technique.
- These devices have 2separate electrode arrays with the number of contacts split between the arrays.

PRE – OPERATIVE EVALUATION

- 1) AUDIOLOGICAL EVALUATION –BOA(Behavioral observation audiomertry), BERA, OAE, IMPEDENCE, PTA
- 2) RADIOLOGICAL EVALUATION To look for anomalies, plan for the approach, to decide on the implant to be used, to assess duration of surgery, to assess implant position post op, to predict outcome.

HRCT Temporal bone with cochlear cuts – Assesses bony labyrinth,

Mastoid-middle ear complex-pneumatisation, cortical thickness and

IACs, Aqueducts, Bony intra - temporal facial nerve canal, jugular bulb, Bone based pathologies

MRI Brain with Internal acoustic meatus screening cuts- MRI essentially compliments CT because of its excellent soft tissue contrast. MRI is directed towards imaging of fluid containing spaces in temporal bone, vascular structures and their pathologies, Adjacent brain parenchyma and Evaluation of 7th and 8th nerve complex

- 3) GENERAL AND SYSTEMIC EVALUATION AND ANAESTHETIC FITNESS Paediatrician opinion, Psychologist opinion and psychological assessment, Ophthalmologist, Cardiologist opinion, TORCH screening, Genetic screening, Routine blood investigations (complete blood count, bleeding, clotting time, blood grouping, typing, viral markers).
- 4) Pneumoccocal polysaccharide vaccine and Meningococcal vaccine should be given prior to surgery.
- 5) Proper pre and post-operative counselling regarding realistic expectation of the outcomes of the cochlear implant surgery and challenges of surgery has to be explained to the parents of child and advice regarding post-operative audioverbal therapy and its importance.
- 6) Candidates with possible less favourable outcome like post meningitis, inner ear malformation, TORCH infection. These poor outcomes have to be explained to the patient prior to surgery.

LEVELS OF OUTCOME

- 1) Signal function (awareness of sounds)
- 2) Support of lip-reading skills
- 3) Open set speech understanding

INDICATION FOR COCHLEAR IMPLANT IN CHILDREN

- Severe to profound sensorineural hearing loss in both ears > 90db in better ear
 [FDA guidelines relaxed to >70db in better ear].
- Lack of benefit from hearing aids and therapy
- No medical contraindications.
- No anatomical contraindications.
- High motivation and expectations for child and family.
- Auditory neuropathy (CI restores synchrony by bypassing cochlear hair cells & stimulating the auditory nerve directly and synchronously).
- Post lingual deaf children (initial USFDA approval limitation).
- Pre lingual deafness with age >12 months
 - o Maturation of long latency cortical responses (decreased latency) occurs reliably when implanted <3 -1/2 yrs, never occurs >7yrs.

o Post implant speech recognition scores directly proportional to implant age.

CONTRAINDICATIONS FOR COCHLEAR IMPLANTATION:

ABSOLUTE:

- Mild to moderate SNHL
- Neurofibromatosis II, mental retardation, psychosis, organic brain dysfunction,
- CT finding of cochlear agenesis (Michel deformity) small IAC (8th CN atresia)

RELATIVE:

- Active middle ear disease
- Labyrinthitis ossificans
- Advanced otosclerosis
- H/o CWD mastoidectomy
- Marked difference in vestibular function between ears (previous trauma)

SURGICAL TECHNIQUE:

The majority of cochlear implant surgeries undertaken was transmastoid approach, accessing the cochlea through the posteriortympanotomy/facial recess.

There are other surgical techniques have been used. They are transcanal approaches (with or without use of an endoscope) and suprameatal approach. The steps of transmastoid approach alone is discussed.

POST AURICULAR SKIN INCISION;

4 types of skin incisions:

Lazy S Incision

• Wide C shaped retro auricular incision

• Normal retro auricular incision

Minimal access incision

Lazy S incision:

Advantage: it gives proper implant coverage and good access for drilling the

mastoid.

Dis advantage: incision has to be avoided over the mastoid tip, it will lead to

injury to the facial nerve.

Wide C shaped retro auricular incision:

Indication: Well pneumatised mastoids, where more access is needed in case

of subtotal petrosectomy

Dis advantage: Wound & scar over receiver stimulator.

MINIMAL ACCESS INCISION:

Dis advantage: severe stretching of skin creates problematic scarring of the skin,

sometimes two incisions may be needed for access mastoid and middle ear and

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another incision for receiver stimulator placement, this creates infections & flap related problems.

Most surgeons use a lazy S incision, it should be sufficient in length to allow the implant to be introduced. The surgeon should avoid placing the incision line directly over the receiver–stimulator package to prevent post-operative wound complications such as dehiscence of the wound and exposure of implant.

MUSCULO PERIOSTEAL FLAP:

The soft tissue is dissected down to the periosteum, which is incised to create an anteriorly based palva flap or posteriorly based flap.

CORTICAL MASTOIDECTOMY:

A cortical mastoidectomy is then performed.

RECEIVER WELL CREATION AND IMPLANT FIXATION

A subperiosteal pocket is created to house the receiver–stimulator. Some surgeons drill a well in the bone to secure the implant and tie is applied over the implant to secure it in position using 3-0 ethibond. However, with the evolution of thinner implants now-a —days—surgeons are choosing simply to create a subperiosteal pocket. Often, a gutter is drilled for the electrode as it passes into the mastoid cavity

POSTERIOR TYMPANOTOMY / FACIAL RECESS APPROACH:

A posterior tympanotomy is created to give access to the round window niche, taking care to preserve the chorda tympani. Then the round window niche has been drilled away), Depending on the surgeon's preference and the access achieved, the entry into the cochlea may be via a cochleostomy or through the round window membrane by opening the round window in an anterior and inferior direction.

The advantages of round window approach are trauma during insertion is less, preserves the residual hearing, less postoperative vestibular complaints, time taken for opening the window is less.

Cochleostomy has more disadvtages than round window approach.it is used in case of difficulty in identification of the round window. Time needed for drilling is more and more postoperative vestibular complaints. Residual hearing is affected.

ELECTRODE INSERTION

After opening the round window the electrode is inserted in the scala

Tympani in an a traumatic fashion. After the insertion of the electrode soft-tissue seal is placed around the niche, to prevent the leakage of perilymph.

WOUND CLOSURE:

The residual electrode wire is coiled within the mastoid cavity, and the wound is closed in layers.

COMPLICATIONS OF COCHLEAR IMPLANT SURGERY:

EARLY COMPLICATIONS:

- Facial paralysis
- Wound infection
- Wound dehiscence
- Flap necrosis
- Electrode migration
- Device failure
- CSF leak
- Meningitis
- Postoperative dizziness/Vertigo

LATE COMPLICATIONS:

- Exposure of device and extrusion
- Pain at the site of implant
- Migration/displacement of device

- Late device failure
- Otitis media.

INTRAOPERATIVE MONITORING:

After cochlear implantation, Intraoperative monitoring of various electrophysiological testing should be done as well as the radiological imaging which helps to assess the function of the implanted electrode and correct placement of the electrode array and the device. The correct placement of electrodes results in successful transfer of stimulating signals from the electrode towards the auditory nerve fibers.

ELECTROPHYSIOLOGICAL TESTING DURING THE

INTRAOPERATIVE PERIOD:

The telemetry system used to test the basic functions of a cochlear implant and to detect the problems in each electrode [short circuit (SC) between electrodes, and open circuits (OC) because of the cable being cut off] [11].

The various multimodal electrophysiological testing was done during the intraoperative period to check the device functioning and neural responsiveness to electrical stimulation. They are

- Intra operative Electrode impedance
- Auditory response telemetry -ART(Evoked compound action potential)
- Electrically evoked stapedial reflex telemetry (ESRT)

ELECTRODE IMPEDANCE FIELD TELEMETRY:

The electrode impedance field telemetry is a method of measuring the resistance encountered by electricity passing through wires, electrodes, and biological tissue 10 . Presently, all CI devices include a telemetry system for checking the operation of the impedance of each electrode in the system and the electrical interaction between them 13 . Measurement of electrode impedance field telemetry provides an information regarding electrode integrity. It is calculated as the ratio of the effective voltage applied to a particular circuit and the actual amount of electrical power intensity absorbed by the circuit. The unit of impedance is the Ω . Short circuits indicate low impedance values whereas Open circuits indicate high impedance

Intra cochlear lesions and new tissue formation (new bone and fibrous tissue) induced by electrode insertion should be minimized by surgical technique and electrode design. Because it increases the electrical impedance. Also air bubble entry during the electrode insertion, faulty insertion techniques and traumatic insertion increases the electrode impedance. So surgeons should try to minimise traumatic insertion using flex electrodes, precise surgical technique and round window approach which significantly reduces the electrode impedance.

Impedance value may be low in cochlear malformation, excess solution in mastoid cavity.

ELECTRICALLY EVOKED STAPEDIAL REFLEX TELEMETRY (ESRT):

The Stapedial reflex is a response to loud sound that results in reflexive contraction of stapedius muscle whereas in the electrically evoked stapedius reflex telemetry electrical stimulus is used to stimulate the reflex contraction of stapedius muscle. Electrical stimulus is delivered to the receiver –stimulator using telemetry coil intraoperatively. ESRT is a measurement for monitoring the stimulation of cochlea. During the intraoperative period, the contraction of the stapedius muscle/tendon can be observed either by microscope or visually by the surgeon before the closure of post auricular incision.¹²

Intra operative ESRT measurement is affected by

- Muscle relaxant duration of action of muscle relaxant.
- Type of anesthetic agent.
- Intraoperative impedence.
- Air bubble entry during Electrode insertion
- Blood and bone dust entry affects intraoperative ESRT.
- Status of middle ear

ADVANTAGES OF ESRT:

• Less time consuming

• Intra operatively – direct visualization of stapedial muscle contraction confirms the integrity of electrode.

LIMITATIONS OF ESRT:

• Affected by many factors- middle ear pathology

AUDITORY RESPONSE TELEMETRY/ NEURAL RESPONSE TELEMETRY -ART(EVOKED COMPOUND ACTION POTENTIAL)

The Neural Response Telemetry (NRT)/Neural response imaging (NRI)/Auditory response telemetry(ART) is a synchronous response from auditory nerve fibres which is stimulated electrically and it is mainly electrical form of wave I of the brainstem evoked response. ART is described as an easy tool in measuring the electrically evoked compound action potential (ECAP) generated by the auditory nerve following electrical stimulation of the cochlea via an electrode of the cochlear implant. ART has negative peak and positive peak. The negative peak as a latency of 0.2 -0.4 ms, which is followed by a positive peak(2)

CLINICAL USES OF ECAP:

- Intra-operatively, ART can be used to verify auditory nerve integrity and complete electrode insertion during surgery.
- Objective verification of auditory nerve function in response to electrical stimulation

 Art thresholds can be used as a clinical tool for programming the speech processor and processing strategies.

• It helps in programming the speech processor who cannot provide reliable information regarding the behavioral response.

• Also useful in verification of accuracy of questionable behavioral response.(2)

Intraoperative ECAP measurement is easier rather than post operative ECAP measurement. Because during the intraoperative period, child is in general anesthesia so high level of electrical stimulation can be given, which gives good resonse, whereas in the post operative period high stimulation causes discomfort to the children. So the given stimulation cannot exceed the loudness acceptance level.

POSTOPERATIVE MAPPING (PROGRAMMING):

Activation of the implant is usually done in 3–4 weeks after implantation. Following activation the implant is "programmed" or "mapped." Mapping is done at regular intervals during postoperative rehabilitation to fine-tune the processor so that the best performance of hearing with the implant can be achieved.

POST OPERATIVE ELECTOPHYSIOLOGICAL TESTS:

Post operative impedence measurement

Post operative Auditory response telemetry (ART)

Post operative ESRT

Post operative EABR

POST OPERATIVE IMPEDENCE MEASUREMENT:

Post operative impedence measurement is measured during the intial activation of the device i.e during switch on in the 3-6weeks after implantation, before doing ECAP measurement.

POST OPERATIVE AUDITORY RESPONSE TELEMETRY (ART):

It is done after switch on ,ECAP measurements were done. Post –operative ART can be used to monitor progress of the cochlear implanted children. It is used as an objective tool for fitting the sound processing system. Fitting the speech processor after cochlear implantation (CI) mainly relies on the determination of the 'threshold' (T) and 'comfort' (C) levels.

ADVANTAGES OF ART:

- Quick procedure and less time consuming.
- Sedation not required
- Cost effective
- Also used in assessing the implantee response subjectively also.

DISADVANTAGES OF ART:

- Cannot be used as a stand alone procedure in assessing the function of electrodes. Has to be correlated with other electrophysiological tests.
- Only a screening tool

POST OPERATIVE ESRT:

Postoperative ESRT was measured in the contralateral ear. The electrical stimulus is given through the telemetry coil to the receiver-stimulator device in the implanted side and reflex contraction of the stapedius muscle is recorded in the contralateral ear. Post operative ESRT is affected by the middle ear pathology.

POST-OPERATIVE EABR:

Electrically evoked auditory brainstem response is measured in the postoperative during the follow up period in the 4 and 6 months after cochlear
implantation. It is same as BERA whereas in EABR instead of sound stimulus
electrical stimulus is used and the appearance of peak V wave form is observed.
The latencies of EABR waveforms were 1 to 1.5 ms earlier than acoustic ABR
waves. This earlier latency can be due to the direct stimulation of spiral ganglion
cells which reduces conduction time that takes place in acoustic ABR.. Even
though EABR is more accurate and reliable in assessing the functional integriy of
electrodes but it has its own limitations. In such cases ART and ESRT will be
useful.

ADVANTAGES OF EABR:

- Superior than other electrophysiological tests.
- Used as confirmatory and diagnostic tool in assessing the function of electrodes.

LIMITATIONS OF EABR:

- Expert skills needed in interpreting the wave forms and morphology.
- Time consuming
- Sedation needed in younger children
- Artifacts will be more
- Costly
- Requires sound proof room and with proper good electrical connection and good earthing is needed.
- Because of time constraint study done only in random selection of electrodes in apical, mid and basal regions.

REHABILITATION:

Rehabilitation is an essential part for those who have undergone cochlear implantation. All patients need AVT (Audio-verbal therapy).

STAGES IN AVT:

- Auditory awareness 2months
- Identification and discrimination 3 to 4months
- Listening in noisy environment 6 to 7 months
- Comprehension -7 to 8 months
- Memory and Sequencing- 9 to 10 months
- Telephone conversation- 1 year

CATEGORIES OF AUDITORY PERFORMANCE (CAP) SCORE:

• The Categories of Auditory Performance (CAP) score is a categorical, nonlinear scale that ranges from 0 to 7. All levels are determined by the ability to perform every-day auditory tasks, with 0 representing no awareness of environmental noise, and 7 representing the ability to talk on a telephone. The CAP scores were recorded in the postoperative period.

CAP score	
0	No awareness of environment sound
1	Awareness of environmental sound
2	Response to speech sounds
3	Identifies environmental sound
4	Discrimination of some speech sounds without lipreading
5	Understands common phrases without lipreading
6	Understands conversation without lipreading
7	Use of telephone with known speaker

FACTORS AFFECTING THE OUTCOMES OF COCHLEAR IMPLANT:

- Age at implantation -- the earlier the better, definitely by age 3, preferably by age 2
- Duration of profound loss -- the shorter, the better
- Duration of cochlear implant use -- maximum benefit not seen until at least 3 5 years post-implant
- Training with amplification/early linguistic experience -- if some residual hearing present and used, results are better with CI
- Communication environment -- patients in oral only environment have better open-set
- Presence of other disabilities -- reduced performance in word recognition compared to patients without disabilities.
- Family support

MATERIALS AND METHODS

STUDY DESIGN:

Prospective study

STUDY CENTRE:

The study was conducted at Upgraded Institute of otorhinolaryngology and Institute of speech and hearing, Madras Medical College, Rajiv Gandhi Government General Hospital, Chennai

DURATION OF STUDY:

The study was conducted from September 2017 to October 2019 – 2 years period.

INCLUSION CRITERIA:

- 1. Patients with severe to profound hearing loss.
- 2. Age :1- 6yrs
- 3. Patients with normal inner ear anatomy.

EXCLUSION CRITERIA:

1. Patients with inner ear abnormality.

ETHICS CLEARANCE

 Ethical clearance was obtained from Institutional Ethics Committee before starting the study. Also the study protocol was approved by the Institutional Ethics Committee.

- Informed written consent was obtained from the parents of the cochlear implant participants before the study.
- The information collected was only used for the study purpose and strict confidentiality is maintained throughout the study.

SAMPLE SIZE

Total of 17 children who fit under inclusion criteria was taken for study

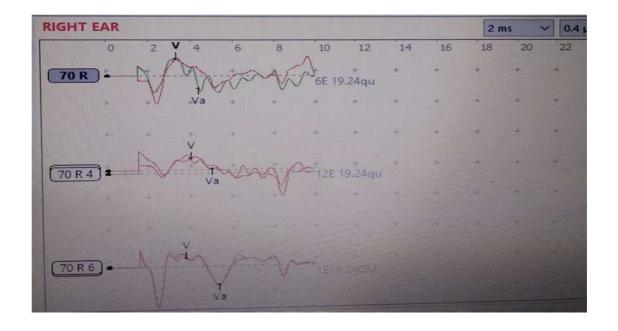
DATA COLLECTED:

- The data was collected from the parents using semi-structured questionnaire / Case pro forma.
- Clinical examination and other relevant investigations were done before and after the surgery.
- Complete audiological examination was done before and after the surgery.
- Pre operative audiological examinations such as BOA,PTA, OAE,BERA,
 Impedence audiometry.
- Intra-operative impedance measurement During surgery Impedance
 were measured on all electrodes after electrode insertion. It was measured
 using the manufacturers default modes

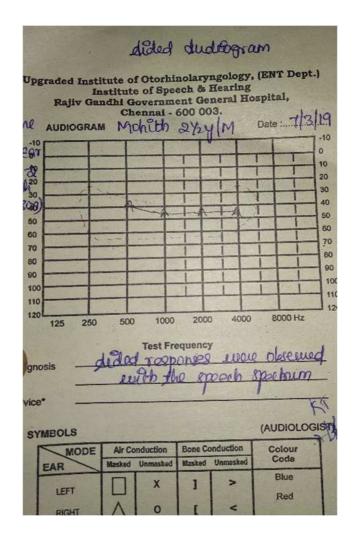
- Post-operative impedance measurement It was measured during the initial device activation appointment and before each programming appointment.
- Intra-operative Electrically evoked stapedial reflex measurement
 Electrical stimulus is given through the telemetry coil and reflex contraction of stapedius muscle was assessed visually in the intraoperative period before closure of the wound.
- Post -operative Electrically evoked stapedial reflex measurement in the post-operative period electrical stimulus is given through the telemetry coil and reflex contraction of the stapedius muscle is recorded in the opposite ear. It was measured in the apical, mid and the basal electrodes randomly.



- Evoked compound action potential (ECAP) Intra-operative Auditory response telemetry (ART) measurements were recorded in all the electrodes using the telemetry coil after impedance measurement.
- Post-operative ART It was measured at the initial device stimulation visit. The post-operative ART measures were made with the patient's own speech processor.
- Post-operative Electrically evoked auditory brain stem response
 (EABR) EABR was done in the postoperative follow up visit .Electrical stimulus was given to apical ,mid and the basal electrodes and appearance of peak V wave was observed.



• **Behavioral responses assessment** – It was assessed using aided audiogram and recorded 2 times in the post-operative follow up period.



Post operative Categories of auditory performance (cap) score : CAP score was recorded in the postoperative period .

DATA ANALYSIS:

 The Data collected in the case proforma was entered into Microsoft excel sheet and data analysis was done by using SPSS statistical software.
 Comparison was done using appropriate statistical methods and appropriate tests of significance were used.

RESULTS

A) AGE WISE DISTRIBUTION:

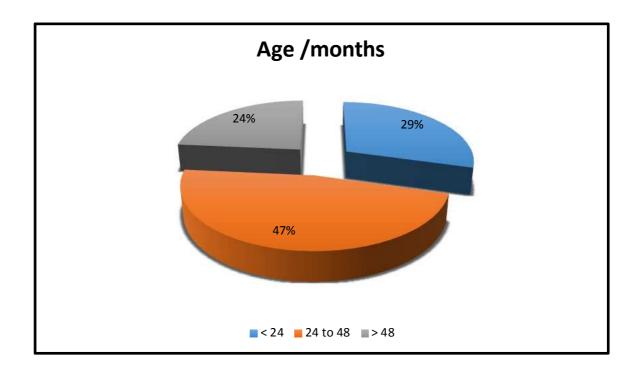
The age wise distribution of the study group is given in the following table.

Table-5.1 Percentage distribution of study group by age

Age in months	Frequency	Percentage
< 24	5	29.4
24 to 48	8	47.1
> 48	4	23.5
Total	17	100.0

In the total of 17 children, 47.1%(8) were between 24-48 months of age, 29.4%(5) were below 24 months and 23.5% (4) were above 48 months of age.

Figure 5.1- Percentage distribution of study group by age



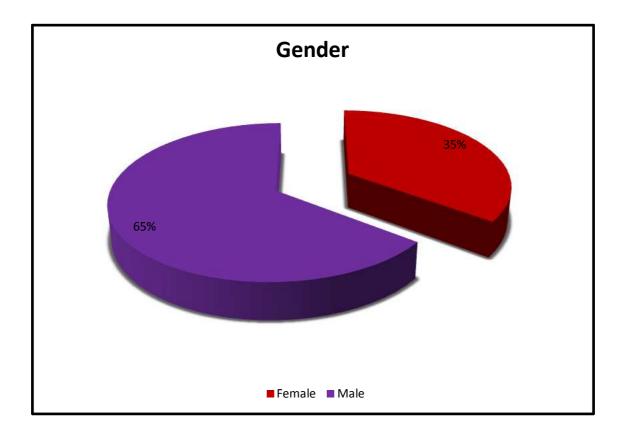
B) GENDER WISE DISTRIBUTION:

The gender wise distribution is given in following table 5.2. Among the study population,35.3% were female and 64.7% were male.

Table 5.2- Percentage distribution of study group by gender.

Sex	Frequency	Percent
Female	6	35.3
Male	11	64.7
Total	17	100.0

Figure 5.2 - Percentage distribution of study group by gender



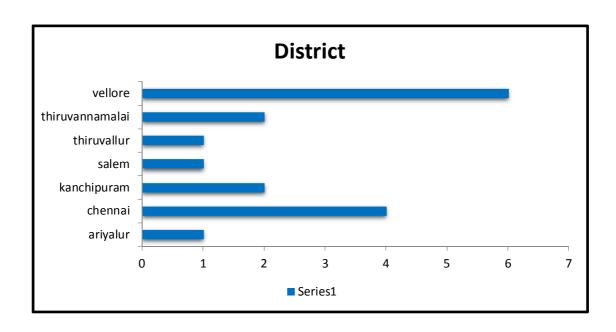
c) DISTRICT WISE DISTRIBUTION:

In the study children among 35.3% children from vellore,23.5% from Chennai,11.8% from thiruvannamalai, 11.8% from kancheepuram, 5.9% from salem,5.9% from thiruvallur, 5.9% from ariyallur district.

Table- 5.3Percentage distribution of study group by district

	Frequency	Percent
ariyalur	1	5.9
Chennai	4	23.5
kanchipuram	2	11.8
Salem	1	5.9
thiruvallur	1	5.9
thiruvannamalai	2	11.8
Vellore	6	35.3
Total	17	100.0

Figure 5.3-Percentage distribution of study group by district



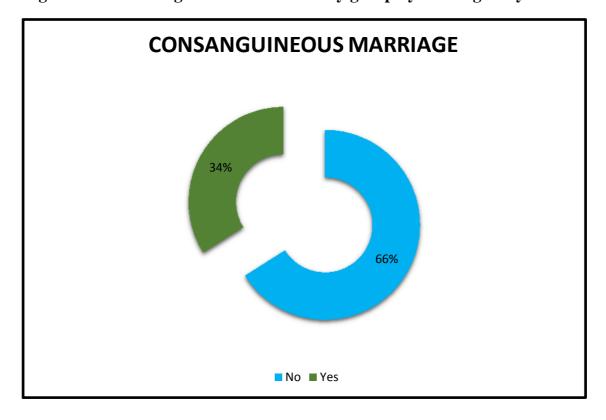
D) CONSANGUINITY OF THE PARENTS:

Among the study group, 33.3% study group parents had consanguineous marriage, whereas 64.7% had no history of consanguineous marriage. The Percentage distribution of study group by consanguinity is given in table 5.4

Table 5.4 :Percentage distribution of study group by consanguinity

Consanguineous Marriage	Frequency	Percent
No	11	64.7
Yes	6	33.3
Total	17	100.0

Figure 5.4 -Percentage distribution of study group by consanguinity



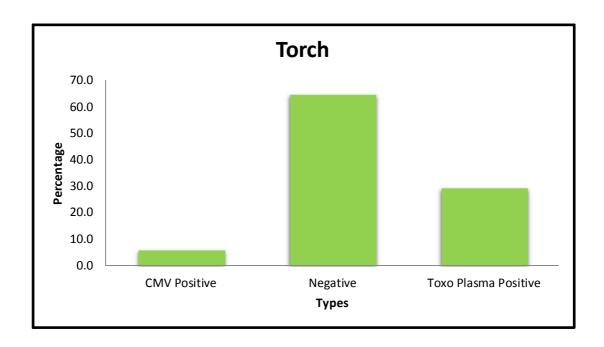
E)TORCH INFECTIONS:

TORCH (Toxoplasma, Rubella, Cytomegalo, Herpes simplex) infection is one of the cause for sensorineural hearing loss in our study group 1(5.9%) were positive for cytomegalovirus infection,5(29.4%) were positive for Toxoplasma infection and 11 (64.7%) were negative for TORCH infection.

Table 5.5 Percentage distribution of study group based on TORCH infection

TORCH	Frequency	Percent
CMV Positive	1	5.9
Negative	11	64.7
Toxo Plasma Positive	5	29.4
Total	17	100.0

Figure 5.5 Percentage distribution of study group based on TORCH infection



F) INTELLIGENT QUOTIENT:

In the study group, about 12% had intelligent quotient between 60-70, 59% between 70-80,24% between 80-90 and 5% had more than 90.

Percentage distribution of study group based on IQ

IQ	No.of children	Percentage
60-70	2	12%
70-80	10	59%
80-90	4	24%
90-110	1	5%

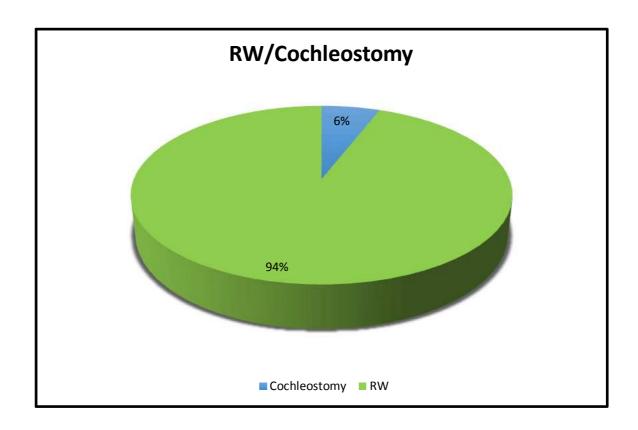
G) COCHLEAR IMPLANT SURGERY DETAILS:

Among the study population, 1 (5.9%) patient had cochleostomy approach and 16 (94.1%) had round window approach. The electrode insertion was full in all the 17 patients i.e 100%. The Percentage distribution of study group based on cochlear implant surgery is given in table 5.7

Table 5.7 Percentage distribution of study group based on cochlear implant surgery

RW/COCHLEOSTOMY	Frequency	Percent
Cochleostomy	1	5.9
RW	16	94.1
Total	17	100.0

Figure -The Percentage distribution of study group based on cochlear implant surgery

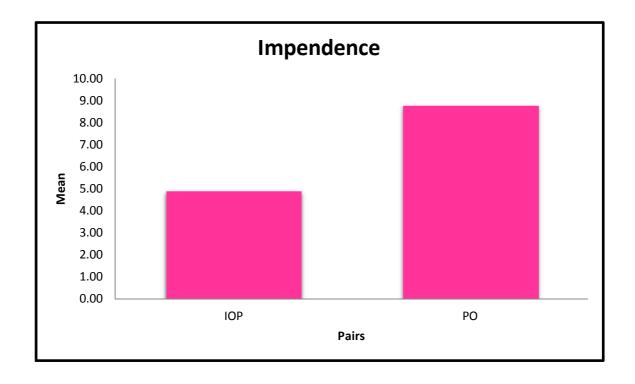


H)IMPEDANCE:

The electrode impedance were measured in the study children during the intraoperative period and the postoperative period during the initial switch on visit at around 3-6 weeks following surgery. The mean intraoperative impedance was 4.89 kohm and the mean postoperative impedance was 8.78kohm. The difference between the intraoperative and the postoperative values were highly statistically significant. (p value -0.0005).

Comparison of Impendence with Paired t-test						
Mean N Std. Deviation t-value P-value						
Doin 1	Impedance Intra Op	4.89	17	1.016	12 102	0.0005 **
Pair 1	Impedance Post Op	8.78	17	1.225	12.193	0.0005 **
** Highly Significant at P < 0.01 level						

Comparison of Intraoperative and postoperative impedance values.



I) AUDITORY RESPONSE TELEMETRY / NEURAL RESPONSE TELEMETRY:

The Auditory response telemetry / neural response telemetry were measured during the intraoperative period and the postoperative period during the initial switch on. The ART were measured in the apical, mid and basal electrodes. The ART measurements were given in the following table.

Table 5.9(1) - Comparison of ART measurement in apical electrodes in the intraop and postop period.

Art Intra Op Apical	Frequency	Percent
Absent	6	35.3
Present	11	64.7
Total	17	100.0

Art Post Op Apical	Frequency	Percent
Absent	4	23.5
Present	13	76.5
Total	17	100.0

ART INTRAOP APICAL WITH ART POSTOP APICAL							
			ART PO	ART PO APICAL		w 2 - waltus	D volvo
			Absent	Present	Total	χ2 - value	P-value
ical	Absent	Count	3	3	6		
Art In Op Apical	Ausent	%	17.6%	17.6%	35.3%		
In O	Duagant	Count	1	10	11	2 611	0.000 #
Art	Present	%	5.9%	58.8%	64.7%	3.611	0.099 #
Total		Count	4	13	17		
Total		%	23.5%	76.5%	100.0%		
	# No Statistical Significance at P>0.05 level						

The ART measurements were compared between intra operatively and post operatively in the apical electrodes. The ART measurement were present in 64.7% and absent in 35.3% in the intraoperative period, whereas ART were present in 76.5% and absent in 23.55 in the postop period.

Figure 5.9(1) - Comparison of ART measurement in apical electrodes in the intraop and postop period

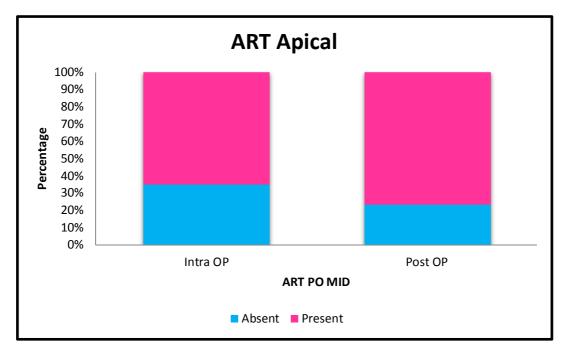


Table 5.9(2) - Comparison of ART measurement in mid electrodes in the intraop and postop period.

Art Intra Op Mid	Frequency	Percent
Absent	7	41.2
Present	10	58.8
Total	17	100.0

ART POSTOP MID	Frequency	Percent
Absent	4	23.5
Present	13	76.5
Total	17	100.0

ART INTRAOP MID WITH ART POSTOP MID							
			ART PO MID		Total	w 2 - walnus	P-value
			Absent	Present	10tai	χ2 - value	r-value
75	Abcont	Count	3	4	7		
p Mic	Absent O uI the pin do Present Present	%	17.6%	23.5%	41.2%	2.471	0.250 #
In O		Count	1	9	10		
Art	Present	%	5.9%	52.9%	58.8%		
Total		Count	4	13	17		
		%	23.5%	76.5%	100.0%		
	# No Statistical Significance at P>0.05 level						

The ART measurements were compared between intra operatively and post operatively in the mid electrodes. The ART measurement were present in 58.8% and absent in 41.2% in the intraoperative period, whereas ART were present in 76.5% and absent in 23.5% in the postop period.

Figure 5.9(2) - Comparison of ART measurement in mid electrodes in the intraop and postop period

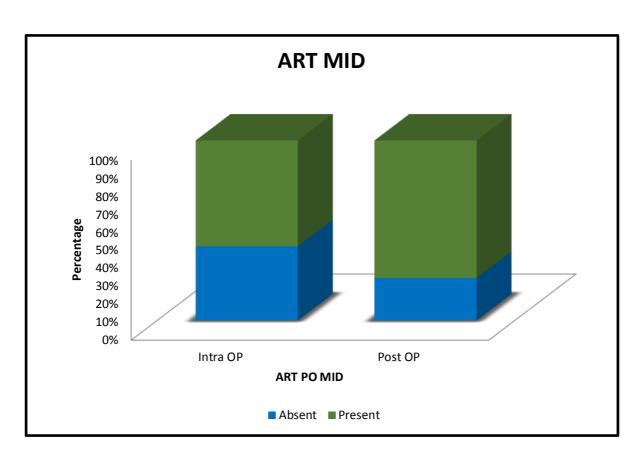


Table 5.9(3) - Comparison of ART measurement in basal electrodes in the intraop and postop period.

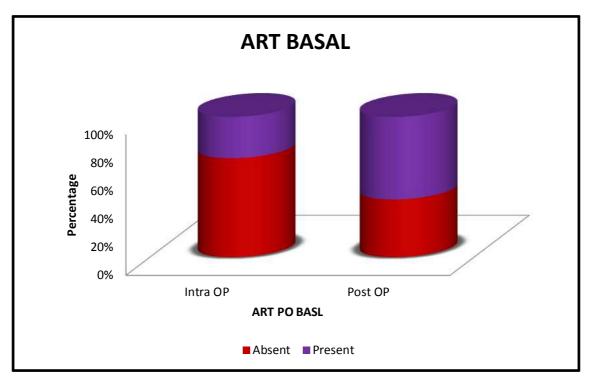
Art Intra Op Basal	Frequency	Percent
Absent	12	70.6
Present	5	29.4
Total	17	100.0

Art Post Op Basal	Frequency	Percent
Absent	7	41.2
Present	10	58.8
Total	17	100.0

ART INTRAOP BASAL WITH ART POSTOP BASAL							
			ART PO BASAL Absent Present		Total	χ2-	D al a
					10tai	value	P-value
d	Abcont	Count	7	5	12		
Absent Absent	%	41.2%	29.4%	70.6%			
Art Intra Op Basal	Basal	Count	0	5	5	4.958	0.044 *
₹ Present	%	0.0%	29.4%	29.4%	4.938	0.044	
Total Cou		Count	7	10	17		
		%	41.2%	58.8%	100.0%		
	* Statistical Significance at P < 0.05 level						

The ART measurements were compared between intra operatively and post operatively in the basal electrodes. The ART measurement were present in 29.4% and absent in 70.6% in the intraoperative period, whereas ART were present in 58.8% and absent in 41.2% in the postop period. There was a statistical significance was present between these two values. (pssss

Figure 5.9(3) - Comparison of ART measurement in basal electrodes in the intraop and postop period



J) ELECTRICALLY EVOKED STAPEDIAL REFLEX TELEMETRY (ESRT):

The electrically evoked stapedial reflex telemetry were measured during the intraoperative period and the postoperative period during the initial switch on.

The ESRT were measured in the apical, mid and basal electrodes. The ESRT measurements were given in the following table.

Table 5.10(1) - Comparison of ESRT measurement in apical electrodes in the intraop and postop period

ESRT Intra Op Apical	Frequency	Percent
Absent	14	82.4
Present	3	17.6
Total	17	100.0

ESRT Post Op Apical	Frequency	Percent
Absent	5	29.4
Present	12	70.6
Total	17	100.0

ESRT Intra Op Apical With Esrt Postop Apical							
			ESRT PO APICAL		Total		D svalsta
			Absent			χ2 - value	P-value
cal	Absont	Count	5	9	14		
Api	Absent	%	29.4%	52.9%	82.4%	1.518	0.515 #
ESRT IO Apical	Duagant	Count	0	3	3		
ESR	Present	%	0.0%	17.6%	17.6%		
Total		Count	5	12	17		
		%	29.4%	70.6%	100.0%		
	# No Statistical Significance at P>0.05 level						

The ESRT measurements were compared between intra operatively and post operatively in the apical electrodes. The ESRT measurement were present in 17.6% and absent in 82.4% in the intraoperative period, whereas ESRT were present in 70.6% and absent in 29.4% in the postop period.

Figure 5.10(1)- Comparison of ESRT measurement in apical electrodes in the intraop and postop period

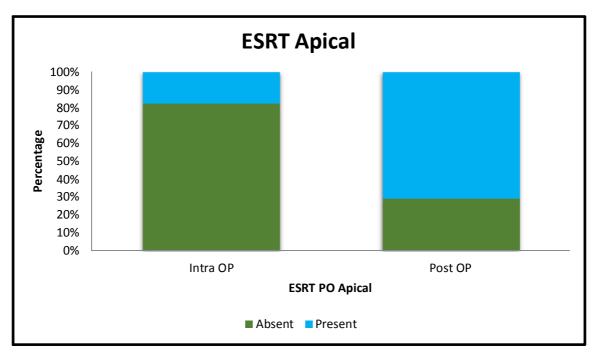


Table 5.10(2) - Comparison of ESRT measurement in mid electrodes in the intraop and postop period.

Esrt Intra Op Mid	Frequency	Percent
Absent	14	82.4
Present	3	17.6
Total	17	100.0

Esrt Post Op Mid	Frequency	Percent
Absent	4	23.5
Present	13	76.5
Total	17	100.0

	ESRT Intra Op Mid With Esrt Postop Mid								
			ESRT I	PO MID	Total		Devolves		
			Absent	Present	Total	χ2 - value	P-value		
	Absont	Count	4	10	14				
MII (Absent	%	23.5%	58.8%	82.4%	1.121	0.541 #		
ESRT IO MID	Present	Count	0	3	3				
ESF	Fresent	%	0.0%	17.6%	17.6%				
Total		Count	4	13	17				
		%	23.5%	76.5%	100.0%				
	# No Statistical Significance at P>0.05 level								

The ESRT measurements were compared between intra operatively and post operatively in the mid electrodes. The ESRT measurement were present in 17.6% and absent in 82.4% in the intraoperative period, whereas ESRT were present in 76.5% and absent in 23.5% in the postop period.

Figure 5.10(2)- Comparison of ESRT measurement in mid electrodes in the intraop and postop period

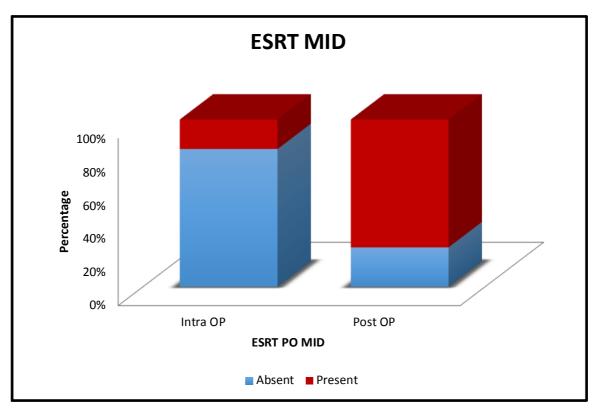


Table 5.10(3) - Comparison of ESRT measurement in basal electrodes in the intraop and postop period.

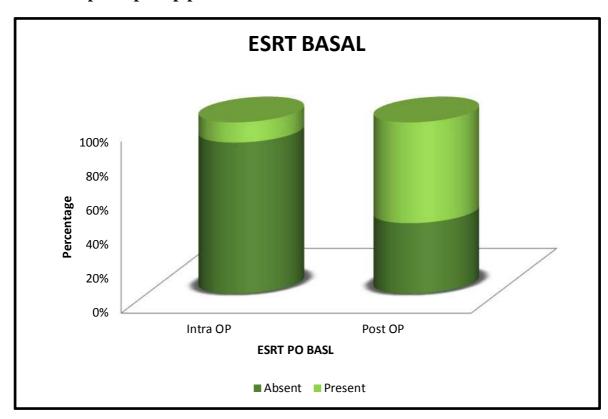
ESRT Intra Op Basal	Frequency	Percent
Absent	15	88.2
Present	2	11.8
Total	17	100.0

ESRT Post Op Basal	Frequency	Percent
Absent	7	41.2
Present	10	58.8
Total	17	100.0

ESRT INTRAOP BASAL WITH ESRT POSTOP BASAL							
			ESRT PO BASAL		T 4 1		D .1 .
			Absent	Present	Total	χ2 - value	P-value
	Absent	Count	6	9	15		1.000 #
ESRT IO BASAL		%	35.3%	52.9%	88.2%	0.073	
	Present	Count	1	1	2		
		%	5.9%	5.9%	11.8%	0.073	
Total		Count	7	10	17		
		%	41.2%	58.8%	100.0%		
# No Statistical Significance at P>0.05 level							

The ESRT measurements were compared between intra operatively and post operatively in the mid electrodes. The ESRT measurement were present in 11.8 % and absent in 58.8% in the intraoperative period, whereas ESRT were present in 88.2% and absent in 41.2% in the postop period.

Figure 5.10(3)- Comparison of ESRT measurement in basal electrodes v``in the intraop and postop period



K) ELECTRICALLY EVOKED AUDITORY BRAINSTEM RESPONSES (EABR):

The electrically evoked auditory brain stem responses are recorded in the postoperative period in the apical,mid and basal electrodes randomly. The presence of peak V was considered as a positive response and absence of Peak V considered as negative response.

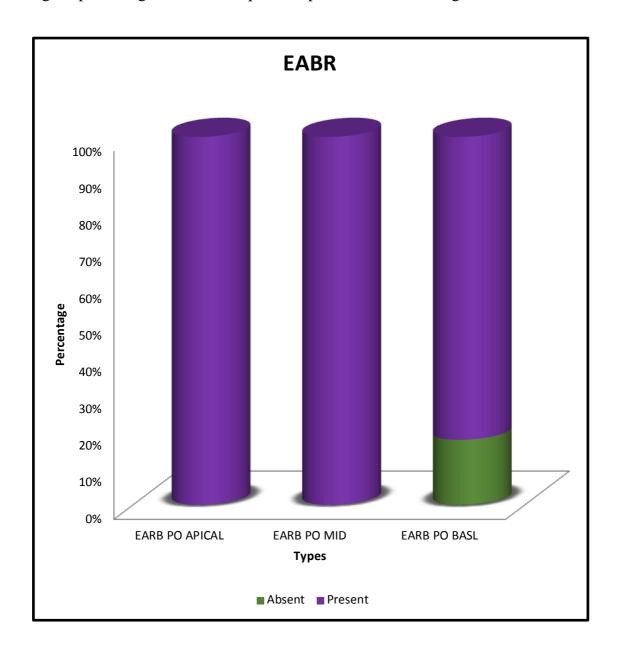
EABR Post Op Apical	Frequency	Percent
Present	17	100.0

EABR Post Op Mid	Frequency	Percent
Present	17	100.0

EABR Postop Basal	Frequency	Percent
Absent	3	17.6
Present	14	82.4
Total	17	100.0

The EABR responses were present in 100% in apical and the mid electrodes whereas in the basal electrodes the response was around 82.4% and absent in 17.6%

Figure-percentage of EABR response is plotted in the bar diagram



L) AIDED AUDIOGRAM

The behavioural responses in the postoperative patients were assessed using aided audiogram. The aided audiogram responses were present in all patients (100%).

M) CATEGORIES OF AUDITORY PERFORMANCE (CAP) SCORE:

The preoperative CAP score was 0 in all children (100%). CAP score at 6 month postoperative period was measured. CAP score of 1 was attained by 6%, score of 2 by 12%, 3 by 29%,4 by 41%,5 by 6% and maximum score of 6 by 6% children in our study.

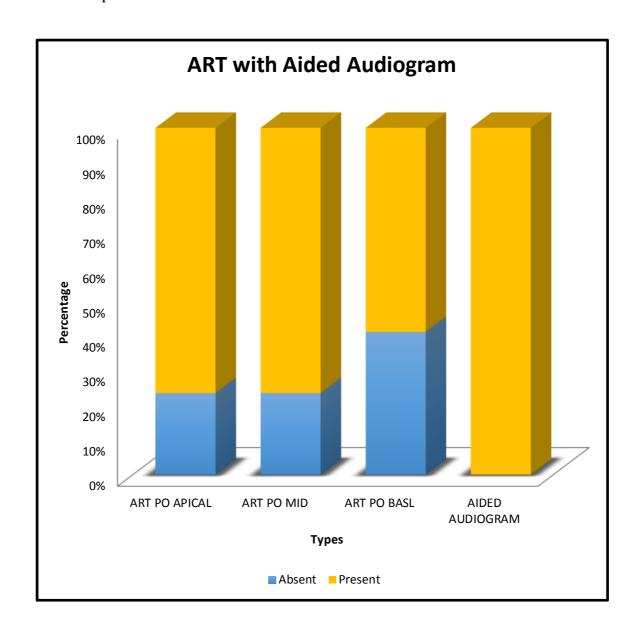
CAP score measurements preoperative and postoperative period given in the table.

CAP SCORE	Preoperative No of children	Preoperative % of children	Postoperative No of children	postoperative % of children
0	17	100%	0	0%
1	0	0	1	6%
2	0	0	2	12%
3	0	0	5	29%
4	0	0	7	41%
5	0	0	1	6%
6	0	0	1	6%
7	0	0	0	0%

About 7 children 41% attained a CAP score of about 4 and those were implanted before the age of 31/2 years.

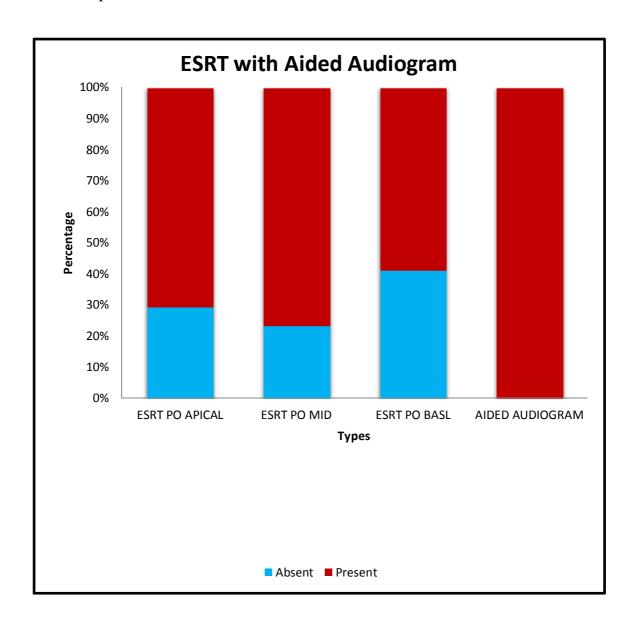
N) ASSOCIATION BETWEEN THE ART AND AIDED AUDIOGRAM:

The ART responses in the apical ,mid and basal electrodes are 76.5%,76.5% and 58.8% respectively in the post operative period, but the aided audiogram shows 100% response.



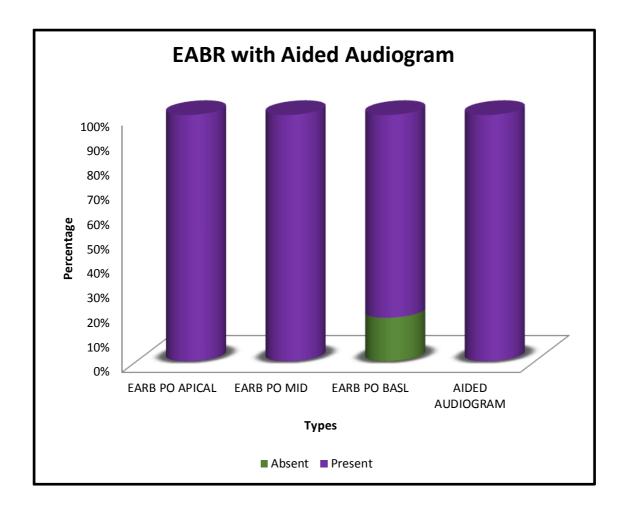
O) ASSOCIATION BETWEEN THE ESRT AND AIDED AUDIOGRAM

The ESRT responses in the apical ,mid and basal electrodes are 70.5%,76.5% and 88.2% respectively in the post operative whereas the aided audiogram shows 100% response.



P) ASSOCIATION BETWEEN THE EABR AND AIDED AUDIOGRAM;

The EABR responses in the apical ,mid and basal electrodes are 100%,100% and 82.4% respectively in the post operative period whereas the aided audiogram shows 100% response.



So among the 3 electrophysiological tests compared to ART and ESRT, EABR has high association between the aided audiogram.

DISCCUSSION

A Prospective study was done at Upgraded Institute of Otorhinolaryngology in the Department of ENT and Institute of speech and hearing, Rajiv Gandhi Government General hospital (RGGGH), Chennai to evaluate the correlation of multimodal Electrophysiological tests and behavioural response in post cochlear implant patients. A total of 17 children were involved in the study, who are satisfying the inclusion criteria and followed up for 12 months. Intra operatively the following Electrophysiological tests were done. They are Electrode impedance measurement, Evoked compound action potential/Auditory response telemetry (ART), Electrically evoked stapedial reflex telemetry (ESRT) and the responses were recorded. During the initial switch on period, Electrode impedance measurement, Evoked compound action potential were measured and in the subsequent follow up period Electrically evoked stapedial reflex telemetry (ESRT) and Electrically evoked auditory brain stem response (EABR) were recorded postoperatively. The behavioural response in the post cochlear implant children were measured using Aided audiogram. The auditory outcome was measured using CAP score.

AGE:

In the total of 17 children in our study, 47.1%(8) were between 24-48 months of age,29.4%(5) were below 24 months and 23.5% (4) were above 48 months of age. All the children in our study showed behavioural response in the post operative period.i.e100%. But subjectively the children implanted at the younger age showed better responses compared to the older ones.

CAP score at 6 month postoperative period was measured. CAP score of 1 was attained by 6%, score of 2 by 12%, 3 by 29%,4 by 41%,5 by 6% and maximum score of 6 by 6% children in our study. About 7 children 41% attained a CAP score of about 4 and those were implanted before the age of 31/2 years.

Harrision et al in 2000 ²⁸ – studied about the effect of age in the outcomes in terms of speech in cochlear implant children. He concluded that significant differences in rate of improvement of scores in the younger implanted children compared with those children implanted in later age.

Nikolopoulous et al in 1999 ²⁴ – studied about the correlation between the age and outcomes in 126 cochlear implant children who were implanted before the age of seven and they were followed a period of 4 years. He stated that a negative correlation between age and outcomes, because outcomes became apparent only after age of 3-4 years following implantation.

Gantz et al in 1994 ²⁷ – conducted a study in 59 children over a follow up period of 5years. He concluded that age of implantation between 2 and 13years has less impact on the outcomes of the cochlear implantation.

Osberger et al in 1999 ²⁵ - conducted a study in 58 children over a follow up period of 18 months in the post cochlear implant patients. He found significant improvement in performance over time.

Several studies have insisted that earlier the age of implantation better the behavioral and audiological outcomes in the children. As the age increases the

loss of neural plasticity has become the reason for the poor behavioural and audiological outcomes.

ELECTRODE IMPEDANCE:

The electrode impedance were measured in our study children during the intraoperative period and the postoperative period. In the postoperative period during the initial switch on visit at around 3-6 weeks following surgery. The mean intraoperative impedance was 4.89 kohm and the mean postoperative impedance was 8.78kohm. The difference between the intraoperative and the postoperative values were highly statistically significant. (p value – 0.0005). But in our study there was no correlation between the electrode impedance and the behavioural response of the children.

Goehring et al in 2013 ⁷ – conducted a retrospective study in 165 post cochlear implant patients. He studied about intraoperative and postoperative impedance in those children. In their study, he found increased intraoperative impedance was resolved in the postoperative period. He stated that abnormal intraoperative impedance is due to the traumatic insertion and variation in the surgical technique and air bubble entry results in the abnormal impedances.

Manolache et al ⁶ studied about electrical impedances variations in patients with cochlear implant. He investigated the changes noted in impedance values of the electrodes implanted in 72 patients over a period of 3 months. He concluded that increase in the post operative impedance is due to absence of electrical stimulation, there is an increase of impedance on all electrodes in the

postoperative period in all cochlear implant models. After activation, the impedances values varies which depends upon the type of the device used. The intraoperative impedance values and their fluctuations are part of the postoperative common trends for each specific device. Other than the device related parameters, the individual variation of the inner ear play an important role in the distribution of electrical impedance profile.

The abnormal intraoperative impedance and the increased postoperative impedances has high chance of resolution in the postoperative regular follow ups. So regular monitoring of the intraoperative and the postoperative impedance should be done for assessing the integrity and function of the electrodes.

AUDITORY RESPONSE TELEMETRY:

The Auditory response telemetry / neural response telemetry were measured during the intraoperative period and the postoperative period during the initial switch on. The ART measurement were present in 64.7% and absent in 35.3% in the intraoperative period, whereas ART were present in 76.5% and absent in 23.5% in the postop period in the apical electrodes, whereas in mid electrodes the ART measures were present in 58.8% and absent in 41.2% in the intraoperative period, whereas ART were present in 76.5% and absent in 23.5% in the postop period. In the basal electrodes the ART were present in 29.4% and absent in 70.6% in the intraoperative period, whereas ART were present in 58.8% and absent in 41.2% in the postop period. In our study compared to the intraoperative ART responses there is a increase in the postoperative ART measures. The

behavioural responses were present in all patients. So there is no one to one correlation of ART responses with behavioural response in our study.

Kim et al in 2010 ²¹ – he included 17 cochlear implant patients in their study. He studied about the relationship between the electrically evoked compound action potential and implant outcome. He concluded that ECAP has significant correlation to the performance with cochlear implant. ECAP measures useful to predict the outcomes with cochlear implant.

Cosetti et al in 2010 ^{31 -} he did a study about the importance of intraoperative telemetry monitoring. He stated that intraoperative telemetry monitoring helps in programming the implant especially in very young children and those with multiple disabilities. Also the abnormal intraoperative impedances is due to air bubble entry during the electrode insertion which may resolve quickly. The results of his study, absent NRT response in 1 or more electrodes were 14% and does not correlate with a dysfunctional device or the postoperative performance.

ELECTRICALLY EVOKED STAPEDIAL REFLEX TELEMETRY (ESRT)

The Electrically evoked Stapedial Reflex Telemetry (ESRT) were measured during the intraoperative period and the postoperative period. The ESRT measurement were present in 17.6% and absent in 82.4% in the intraoperative period, whereas ESRT were present in 70.6% and absent in 29.4% in the postop period in apical electrodes, in mid electrodes. ESRT measures were present in 17.6% and absent in 82.4% in the intraoperative period, in the postop period ESRT were present in 76.5% and absent in 23.5%. in basal electrodes the

responses were present in 11.8 % and absent in 58.8% in the intraoperative period, in the postop period ESRT were present in 88.2% and absent in 41.2%. In our study compared to the intraoperative ESRT responses there is a increase in the postoperative ESRT measures. Because intraoperative ESRT was affected by many factors. The behavioural responses were present in all study children. In this study intraoperative ESRT shows no correlation but postoperative ESRT responses increased and shows correlation with behavioural response.

Baysal et al in 2011¹- did a study with 65 children undergoing cochlear implant. He studied the correlation between intra- and postoperative electrically evoked stapedius reflex thresholds (ESRTs). He stated that intraoperative ESRT measurements were unable to predict early postoperative ESRT. A correlation analysis did not reveal any statistically significant correlation between intra- and postoperative ESRTs.

ELECTRICALLY EVOKED AUDITORY BRAINSTEM RESPONSES (EABR):

The electrically evoked auditory brain stem responses are recorded in the postoperative period in the apical, and basal electrodes. The EABR responses were present in 100% in apical and the mid electrodes whereas in the basal electrodes the response was around 82.4% and absent in 17.6%. the behavioural responses were 100% in study participants. In our study nearly 14 patients showed positive EABR responses in all electrodes, only 3 patients showed absent responses in basal electrodes, hence EABR shows significant correlation with behavioural responses.

Mariappan et al in 2019 ³² – did a study among 21 cochlear implant children. EABR were recorded in implanted children, amplitude and latencies of the waveforms were analysed. He stated that the apical electrode tends to show steeper (amplitude) and earlier (latency) EABR waveforms than the middle and basal electrode.

COMPARISON OF ART, ESRT AND EABR WITH BEHAVIOURAL RESPONSES:

In our study, the overall postoperative response in ART is 70.5%, ESRT response is 78.4% and EABR response is 96%, whereas the behavioural response is 100%. So ART shows least responses, then comes ESRT whereas EABR responses shows more correlation with behavioural responses.

S.Mason et al in 2004 ⁹ – conducted a study in 427 cases of cochlear implantation patients. He reviewed the electrophysiology and objective measures as monitoring tool for cochlear implant in operating room and their value in management of children. He did Impedance telemetry, ESRT and ART. He concluded that these electrophysiological tests helps in assessing the implant was functioning properly and their outcomes.

Bayrak et al ²²- did a study among 16 children who underwent cochlear implant. He studied about the relationship between the electrically evoked compound action potential and electrically evoked auditory brain stem responses. He concluded that consistency was found between ECAP and EABR recordings. But one cannot be preferred over the other because the data quality of the two tests was different.

CONCLUSION

In summary, cochlear implantation is a surgical procedure, which provides a sense of hearing in hearing challenged individuals. So the proper functioning of the device and electrodes should be checked periodically. There are several electrophysiological tests available to check the device function. These electrophysiological tests are useful in the intraoperative period to check the proper position and function of the electrodes immediately. Thus it gives an assurance about device function and position to the surgeon in the intraoperative table itself.

The electrophysiological tests in the postoperative period helps in the assessing the device function and function of the individual electrodes, thus it helps in assessing the behavioural response and audiological outcomes in post cochlear implant patients. The device dysfunction significantly affects the behavioural responses and audiological outcomes, hence the electrophysiological tests helps in early identification of the device failure.

Among the various electrophysiological tests, intraoperative ESRT is useful in assessing the electrode function accurately because visual or microscopic assessment of the contraction of the stapedius was made by the surgeon and gives assurance. But the intraoperative ESRT was affected by several factors, which limits its usage. But postoperative ESRT responses significantly increased compared to the intraoperative responses. Hence

there is no correlation between the intraoperative and postoperative ESRT measurements.

In the postoperative period, compared to ART and ESRT responses, the percentage of EABR responses are higher, hence it is useful in assessing the electrode function accurately and shows significant correlation with objective test like aided audiogram and subjective audiological assessment using CAP score than other tests. But expert skills are needed to predict the EABR wave morphology and interpretation limits its use.

Hence there is no single electrophysiological tests should be used in assessing the device function and behavioural response assessment. Each tests is compliment to each other and each tests assess the different parameters. So combination of electrophysiological tests should be used.

Hence for the successful cochlear implant outcomes all cochlear implant centre should have adequate equipment for audiological assessment and trained audiologist. The audiologist must be specially trained to do the electrophysiological tests and should be expert in picking up the behavioural responses from the children. Also they should be trained to do switch on, mapping, Auditory verbal therapy and management in trouble shooting.

REFERENCES

- 1. Baysal E,et al,intraoperative electrically evoked stapedius reflex thresholds (ESRT) in children undergone cochlear implantation,int J otorhinolaryngol.2012.may;76(5)
- 2. Michelle l.hughes ,fundamentals of clinical ECAP measuresin cochlear implants;part 1:use of ECAP in speech processor programming (2nd edition)nov 2010.
- 3. M.J. Makhdoum, A.F. Snik, M.H. Stollman, P.M. de Grood, P. van den Broek, The influence of the concentration of volatile anesthetics on the stapedius reflex determined intraoperatively during cochlear implantation in children, Am. J. Otol.
- B. Van den Borne, A.F.M. Snik, L.H.M. Mens, J.P.L. Brokx, P. van den Broek, Stapedius reflex measurements during surgery for cochlear implantation inchildren, Am. J. Otol. 17 (1996) 554–558. 19 (1998) 598– 603.
- 5. AndradeKC et al,the importanceof electrically evoked stapedial reflexin cochlear implant,braz J otorhinolaryngol.2014 jan-feb.
- 6. Manolache O, Olariu R, Radulescu L, Cozma S. Electrical impedances variations values in patients with cochlear implant. Romanian J Oral Rehabil 2012; 4:22–28.

- 7. Goehring JL, Hughes ML, Baudhuin JL, Lusk RP,intraoperative impedance measures predict postoperative electrode function Otol Neurotol 2013; 34:239–244.
- 8.J. Kosaner, I. Anderson, Z. Turan, M. Deibl, The use of ESRT in fitting children with cochlear implants, Int. Adv. Otol. 5 (2009) 62–71.
- Mason, Electrophysiologic and objective monitoring of the cochlear implant during surgery: implementation, audit and outcomes, Int. J. Audiol. 43 (2004) 33–38.
- Finley CC, Holden TA, Holden LK, Whiting BR, Chole RA, Neely GJ, et al. Role of electrode placement as a contributor to variability in cochlear implant outcomes. Otol Neurotol 2008; 29:920–928.
- 11. Schulman JH. Using impedance telemetry to diagnose cochlear electrode history, location and functionality. Ann Otol Rhinol Laryngol 1995; 166
- 12. Scott brown,the text book of otorhinololaeryngology head ansd neck surgery, 4th edition volume 3.
- 13. K. Stephan, K. Welzl-Muller, H. Stiglbrunner, Acoustic reflex in patients with cochlear implants (analoiz stimulation), Am. J. Otol. 12 (1991) 48–51. (Suppl):85–87.
- 14. Tykocinski M, Cohen LT, Cowan RS. Measurement and analysis of access resistance and polarization impedance in cochlear implant recipients. Otol Neurotol 2005; 26:948–956.

- 15. J. Kosaner, Generating speech processor programmes for children using ESRT measurements, Cochlear Implants Int. 11 (Suppl 2) (2010) 20–24.
- 16. Kasper AU: Electrically Evoked Activity in the Human Auditory System:Thesis No. 2472, U niversity of Geneva. 1991.
- 17. Abbas PJ. Brown CJ: Electrically evoked auditory brainstem responses:

 Refractory properties and strength-duration functions. Hear Res
 1991:51:139-148.
- 18. Abbas PJ. Brown CJ: Electrically evoked auditory brainstem responses:

 Refractory properties and strength-duration functions. Hear Res
 1991:51:139-148.
- 19. Kartas ,Denis aud , Electrically evoked stapedius reflex thresholds in children undergone cochlear implantation,int J otorhinolaryng,2012.
- 20. Kosaner ,comparing eSRT and eCAP measurements in pediatric cochlear implant users,int J cochlear implant .19(11)jan 2018.
- 21. Ryong kim, paul abbas,the relationship between Electrically evoked compound action potential and speech perception in cochlear implant users,otology-neurotology.31;1041-1048
- 22. Bayrak, Examination and Comparison of Electrically Evoked Compound Action Potentials and Electrically Evoked Auditory Brainstem Response Results of Children with Cochlear Implantation without Inner Ear Anomaly,turk arch otorhinolaryng-jan 19;10.5152.

- 23. Mohammed said abdelsalam, Electric Auditory Brainstem Response in Cochlear Implant children, eygpt J ent 2015,vol 16 145-150.
- 24, Nikolopoulous TP, O'Donoghue GM, Archbold S. Age at implantation: its importance in pediatric cochlear implantation. Laryngoscope 1999;109:595–9.
- 25. Osberger MJ, Zimmerman-Phillips S, Barker M, GeierL. Clinical trial of the Clarion cochlear implant in children. Ann Otol Rhinol Laryngol 1999;108:88–92.
- 26. Snik AF, Markhdoum MJ, Vermeulen AM, Brokx JP, van den Broek P. The relation between age at the time of cochlear implantation and long term speech perception abilities in congenitally deaf subjects. Int J Pediatr Otorhinolaryngol 1997;41:121–31.
- 27. Gantz BJ, Tyler RS, Woodworth GG, Tye-Murray N,Frauf-Bertschy H. Results of multichannel cochlear implants in congenital and acquired prelingual deafness in children: . ve year follow-up. Am J Otol 1994; 15(Suppl 2):1–7.
- 28. Harrison RV, Nedzelski J, Picton N, et al. The paediatric cochlear implant program at the Hospita Ifor Sick Children, Toronto. J Otolaryngol 1997;26:180–
- 29. Harrison, The effects of age of cochlearimplantation on speech perception outcomes in prelingually deaf children, scand audiology 2001;30 suppl 53:73-78.

- 30. Scott brown,the text book of otorhinololaeryngology head ansd neck surgery, 8th edition volume 3.
- 31. Cosetti MK, Shapiro WH, intraoperative neural response telemertry as a predictor of performance, otol neurotol .2010.sep 31(7):1095-9
- 32. Mariappan, narendra kumar ,Electrically evoked auditory brainstem responses in complete cochlear coverage,ty journ 2017,vol 1.
- 33. Cummings, the text book of otorhinololaeryngology head ansd neck surgery, 6^{th} edition volume 3.

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.P.Nithya
I Year Post Graduate in M.S. ENT
Upgraded Institute of Otorhinolaryngology
Madras Medical College
Chennai

Dear Dr.P.Nithya,

The Institutional Ethics Committee has considered your request and approved your study titled "TO EVALUATE THE CORRELATION OF ELECTROPHYSIOLOGICAL TESTS AND BEHAVIORAL RESPONSES IN COCHLEAR IMPLANT PATIENTS" - NO.16022018

The following members of Ethics Committee were present in the meeting hold on **06.02.2018** conducted at Madras Medical College, Chennai 3

1. Prof.P.V.Jayashankar	:Chairperson
2. Prof.R.Jayanthi, MD., FRCP (Glasg) Dean, MMC, Ch-3 : Dep	outy Chairperson
3. Prof.Sudha Seshayyan, MD., Vice Principal, MMC, Ch-3 : N	Member Secretary
4. Prof. N. Gopalakrishnan, MD, Director, Inst. of Nephrology, MMC, C	h : Member
5. Prof.S. Mayilvahanan, MD, Director, Inst. of Int. Med, MMC, Ch-3	: Member
6. Prof.A.Pandiya Raj, Director, Inst. of Gen.Surgery, MMC	: Member
7. Prof.Shanthy Gunasingh, Director, Inst. of Social Obstetrics, KC	H : Member
8. Prof.Rema Chandramohan, Prof. of Paediatrics, ICH, Chennai	: Member
9. Prof. Susila, Director, Inst. of Pharmacology, MMC, Ch-3	: Member
10. Prof. K. Ramadevi, MD., Director, Inst. of Bio-Chemistry, MMC, C	ch-3 : Member
11. Prof. Bharathi Vidya Jayanthi, Director, Inst. of Pathology, MMC	C,Ch-3: Member
12. Thiru S. Govindasamy, BA., BL, High Court, Chennai	: Lawyer
13.Tmt.Arnold Saulina, MA., MSW.,	:Social Scientist
14.Thiru K.Ranjith, Ch- 91	: Lay Person

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.

Member Secretary - Ethics Committee



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Submitted: 10/24/2019 5:07:00 AM Submitted By: nithihari89@gmail.com

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MRI TEMPORAL BONE:
INTRAOPERATIVE FINDING:
Intraoperative Impedance field telemetry
Intraoperative ESRT
Intraoperative ART
POST OPERATIVE ASSESSMENT:
Postoperative Impedance field telemetry
Postoperative ART
Postoperative ESRT

Postoperative EABR

BEHAVIORAL RESPONSE ASSESSMENT – using Aided audiogram

Post operative CAP score:

INFORMATION SHEET

TITLE: "TOEVALUATE THE CORRELATION OF ELECTROPHYSIOLOGICAL TESTS AND BEHAVIORAL RESPONSES IN COCHLEAR IMPLANT PATIENTS"

Name of Investigator: Dr. PNITHYA.

Name of Participant:

Purpose of Research: To determine the importance of electrophysiological tests and its correlation with behavioral responses in cochlear implant patients..

Study Design: Prospective and Retrospective Observational Study

Study Procedures: Patient will be subjected to electrophysiological tests like ESRT during intraoperative period and ESRT and EABR in postoperative period .

Possible Risks: No risks to the patient

Possible benefits

To patient : Correlation of Electrophysiological tests and the behavioral responses of patient is assessed over first year of implantation.

To doctor & to other people: This study help to know the importance of electrophysiological tests and its correlation with behavioral responses.

Confidentiality of the information obtained from you: The privacy of the patients in the research will be maintained throughout the study. In the event of any publication or presentation resulting from the research, no personally identifiable information will be shared

Can you decide to stop participating in the study: Taking part in this study is voluntary. You are free to decide whether to participate in this study or to withdraw at any time

How will your decision to not participate in the study affect you: Your decision will not result in any loss of benefits to which you are otherwise entitled.

`Participant

Date:

PATIENT CONSENT FORM

Study Detail	ELECTROPHYSIO	THE CORRELATION OF LOGICAL TESTS AND SPONSES IN COCHLEAR IMP	LANT
Study Centre	: Rajiv Gandhi Gover	nment General Hospital, Chennai.	
Patient's Name	:		
Patient's Age	:		
In Patient Number	:		
Patient may check () these boxes		
		for the above study. I have the opportunity been answered to my complete satisfaction.	٥
• •	ipation in the study is voluntary without my legal rights being a	and that I am free to withdraw at any time ffected.	-
records, both in resper relation to it, even if I was identity will not be	gulatory authorities will not not of current study and any family withdraw from the study I agree a revealed in any information re-	orking on the sponsor's behalf, the Ethics need my permission to look at my health further research that may be conducted in the to this access. However, I understand that beleased to third parties or published, unless see of any data or results that arise from this	
study.			
faithfully cooperate wit		the instructions given during the study and iately inform the study staff if I suffer from expected or unusual symptoms	0
I hereby consent to participa		expected of unusual symptoms.	_
	to undergo complete clinical e	examination and diagnostic tests including indergo treatment	-
Signature/thumb impres	sion	Signature of Investigator	
Patient's Name and Add	lress:	Study Investigator's Name:	

INFORMED CONSENT

"TO EVALUATE THE CORRELATION OF ELECTROPHYSIOLOGICAL TESTS AND BEHAVIORAL RESPONSES IN COCHLEAR IMPLANT PATIENTS"

At Madras Medical College Government General Hospital, Chennai

நான் இந்த ஆராய்ச்சியில் விவரங்களை முற்றிலும் புரிந்துகொண்டேன். ஆய்வில் பங்குஎடுத்துபோது, சாத்தியமான அபாயங்கள் மற்றும் பயன்களை பற்றி நான் அறிந்துள்ளேன்.

நான் எந்தவொருவேளையிலும் ஆய்வில் இருந்து திரும்பமுடியும், அதன் பின்னர், நான் வழக்கம்போல் மருத்துவசிகிச்சை பெற முடியும் என்று புரிந்துகொள்கிறேன்.

நான் ஆய்வில் பங்கு எடுத்து பணம் எதையும் பெறமுடியாது என்று அறிந்துள்ளேன்.

இந்த ஆய்வின் முடிவுகள் எந்த மெடிக்கல்ஜர்னலில் வெளியிடப்பட இருந்தால் நான் எதிர்க்கவில்லை, என் தனிப்பட்ட அடையாளத்தை வெளிப்படுத்தப்பட்டு இருக்ககூடாது.

நான் இந்த ஆய்வில் பங்கெடுப்பதன் மூலம் நான் என்னசெய்யபோகிறேன் என்று தெரியும்.

நான் இந்த ஆய்வில் என் முழுஒத்துழைப்பையும் கொடுப்பேன் என்று உறுதியளிக்கிறேன்.

தன்னார்வளர் பெயர்மற்றும்முகவரி கையொப்பம் / விரல்ரேகை: சாட்சி பெயர்மற்றும்முகவரி கையொப்பம் / விரல்ரேகை:

ஆராய்ச்சியாளராக

கையொப்பம்மற்றும்தேதி

MASTER CHART

S. No	NAME	Date of birth	AGE	AGE OF IMPLANT	SEX	IP NO	Date of surgery	Date of SWITCH ON	DISTRICT	Consanguineous marriage	HEARING AID USE	DURATION	IQ	CT/MRI	TORCH
1	sasikumar	16.9.14	5y1m	40m	M	97315	25.1.18	23.2.18	kanchipuram	y	N	N	82	normal	negative
2	santhosh	16.1.15	4y9m	38m	M	98432	20.2.18	28.3.18	vellore	Y	N	N	79	normal	toxoplasmapositive
3	tamilselvan	18.9.15	4y1m	30m	M	98837	10.3.18	4.4.18	chennai	N	N	N	89	normal	negative
4	harishraj	27.5.16	3y5m	24m	M	48311	5.5.18	1.6.18	ariyalur	Y	N	N	68	normal	negative
5	mohammed fowzan	3.8.16	3y1m	22m	M	59604	1.6.18	16.8.18	vellore	Y	N	N	74	normal	negative
6	karkuzhali	14.12.16	2y10m	20m	F	48958	12.7.18	17.8.18	thiruvannamalai	N	N	N	72	normal	negative
7	ayansh	21.7.14	5y2m	48m	M	81657	26.7.18	21.8.18	chennai	N	N	N	76	normal	toxoplasmapositive
8	pugazhlendhi	8.5.13	6y5m	63m	M	82409	27.8.18	10.9.18	vellore	N	N	N	64	normal	negative
9	mathiyabanu	18.9.16	3y5m	22m	F	82685	30.7.18	3.9.18	vellore	N	N	N	72	normal	negative
10	lokisha	23.6.14	5y4m	38m	F	82410	6.8.18	3.9.18	salem	Y	N	N	102	normal	negative
11	salvarani	19.7.14	4y2m	37m	F	82756	7.8.18	3.9.18	chennai	Y	N	N	78	normal	toxoplasmapositive
12	hareeshwaran	22.11.13	6у	51m	M	88455	21.8.18	19.9.18	thiruvannamalai	N	N	N	84	normal	toxoplasmapositive
13	arun	12.4.14	5y5m	52m	M	92771	238.18	19.9.18	vellore	N	N	N	76	normal	negative
14	mohit	26.10.16	Зу	22m	M	93034	29.8.18	27.9.18	vellore	N	N	N	80	normal	negative
15	aishwarya	6.3.16	3y6m	35m	F	5864	6.2.19	9.3.19	chennai	N	N	N	74	normal	negative
16	dharshika	27.9.16	Зу	30m	F	25054	14.3.19	16.4.19	kanchipuram	N	N	N	76	normal	CMVpositive
17	samson	25.8.13	6у	68m	M	25067	15.3.19	22.4.19	thiruvallur	N	N	N	85	normal	toxoplasmapositive

MASTER CHART

PREOP OAE	PRE OP IMPEDENCE	PREOP BERA	ART INTRAOP APICAL	ART INTRAOP MID	ART INTRAOP BASAL	ART PO OP APICAL	ART POST OP MID	ART POST OP BASAL	IMPEDENCE INTRAOP	IMPEDENCE POSTOP	ESRT INTRAOP APICAL	ESRT INTRAOP MID
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Present	Present	Absent	Present	Present	Present	3.8	6.3	Present	Present
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Present	Present	Present	Present	Present	Present	4.3	8.8	Absent	Present
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Present	Absent	Present	Present	Present	Present	7.3	10.7	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Present	Absent	Absent	Absent	Absent	Present	4.78	8.7	Absent	Absent
OHC dysfunction	b/l 'B' CURVE	prof SNHL	Absent	Absent	Absent	Absent	Absent	Absent	5.6	7.15	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Present	Absent	Absent	Present	Present	Absent	6.9	9.7	Absent	Absent
OHC dysfunction	b/l 'B' CURVE	prof SNHL	Absent	Present	Present	Present	Present	Present	4.1	8.56	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Absent	Absent	Absent	Present	Present	Absent	5.1	10.7	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Absent	Absent	Absent	Present	Absent	Absent	5.9	6.9	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Present	Present	Absent	Present	Absent	Absent	4.8	8.4	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Absent	Present	Absent	Absent	Present	Absent	3.9	8.5	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Present	Absent	Absent	Present	Present	Present	4.3	8.4	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Present	Present	Absent	Present	Present	Present	3.9	9.7	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Absent	Present	Absent	Absent	Present	Present	4.9	10.2	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Present	Present	Absent	Present	Present	Absent	4.8	9.1	Present	Absent
OHC dysfunction	b/l 'B' CURVE	prof SNHL	Present	Present	Present	Present	Present	Present	4.6	8.8	Absent	Absent
OHC dysfunction	b/l 'A' CURVE	prof SNHL	Present	Present	Present	Present	Present	Present	4.17	8.57	Present	Present

MASTER CHART

ESRT INTRAOP BASAL	ESRT POSTOP APICAL	ESRT POSTOP MID	ESRT POSTOP BASAL	EABR POST OP APICAL	EABR POST OPMID	EABR POSTOP BASAL	ELECTRODE INSERTION	ROUND WINDOW/ COCHLEOSTOMY	AIDED AUDIOGRAM 1	AIDED AUDIOGRAM 2	CAP SCORE
Absent	Present	Present	Present	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	4
Absent	Present	Present	Present	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	2
Absent	Present	Present	Present	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	4
Absent	Absent	Absent	Absent	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	3
Absent	Absent	Absent	Absent	Present	Present	Absent	Full	RW	within speech spectrum	within speech spectrum	4
Absent	Present	Present	Present	Present	Present	Present	Full	COCHLEOSTOMY	within speech spectrum	within speech spectrum	4
Present	Absent	Absent	Absent	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	3
Absent	Absent	Absent	Absent	Present	Present	Absent	Full	RW	within speech spectrum	within speech spectrum	3
Absent	Present	Present	Present	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	1
Absent	Present	Present	Absent	Present	Present	Absent	Full	RW	within speech spectrum	within speech spectrum	4
Absent	Absent	Present	Absent	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	3
Absent	Present	Present	Present	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	6
Absent	Present	Present	Present	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	5
Absent	Present	Present	Present	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	4
Absent	Present	Present	Absent	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	4
Absent	Present	Present	Present	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	3
Present	Present	Present	Present	Present	Present	Present	Full	RW	within speech spectrum	within speech spectrum	2