"NEONATAL HEARING EVALUATION-A HOSPITAL BASED STUDY"

By

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Date: **Dr. Ciju K. George**

LIST OF ABBREVIATIONS

%	-	Percentage
AABR	-	Automated Auditory Brainstem Response
ABR	-	Auditory Brainstem Response
BERA	-	Brainstem Evoked Response Audiometry
CP	-	Cerebral Palsy
dB	-	Decibel
DPOAE	-	Distortion Product Otoacoustic Emissions
EAC	-	External Auditory Canal
ET	-	Exchange Transfusion
F1	-	Frequency 1
F2	-	Frequency 2
GDM	-	Gestational Diabetes Mellitus
HDN	-	Hemorrhagic Disease of Newborn
HIE	-	Hypoxic Ischemic Encephalopathy
Hz	-	Hertz
IHS	-	Infant Hearing Screening
IUGR	-	Intra Uterine Growth Restriction
JCIH	-	Joint Committee on Infant Hearing
LBW	-	Low Birth Weight
MAS	-	Meconium Aspiration Syndrome
MSL	-	Meconium Stained Liquor
MV	-	Mechanical Ventilation
NA	-	Not Applicable
NHB	-	Neonatal Hyperbilirubinemia
NHS	-	Newborn Hearing Screening
NICU	-	Neonatal Intensive Care Unit
OAE	-	Otoacoustic Emission

PPROM	-	Preterm Premature Rupture of Membrane
RDS	-	Respiratory Distress Syndrome
Sig	-	Significant
SNHL	-	Sensorineural Hearing Loss
SOAE	-	Spontaneous Otoacoustic Emissions
SPL	-	Sound Pressure Level
TEOAE	-	Transient Evoked Otoacoustic Emissions
TNHS	-	Targeted Newborn Hearing Screening
TTN	-	Transient tachypnea Newborn
UNHS	-	Universal Newborn Hearing Screening
VLBW	-	Very Low Birth Weight

ABSTRACT

Aim: To find the prevalence of sensorineural hearing loss in neonates admitted in BLDEU's Shri B. M. Patil Medical College, Hospital and Research Centre, Vijayapur.

Materials & Methods: A study group consisting 320 neonates from the department of Pediatrics were evaluated in the department of Otorhinolaryngology and Head & Neck Surgery, Shri B. M. Patil Medical College, Hospital and Research Centre, Vijayapur during the period October 2014 to August 2016. Neonates were subjected to DPOAE at 48-96 hours of life. For pass cases no further testing was done. For refer cases repeat DPOAE testing was done at 45-60 days of life. Those infants who failed the rescreening were subjected to Brainstem Evoked Response Audiometry (BERA) within 3 months.

STUDY DESIGN: Descriptive study

Results: Three hundred twenty neonates were screened by DPOAE. 39 infants had refer result for 1st DPOAE hearing screen. The second DPOAE screen was done at 45-60 days of life.19 infants who had failed the initial screen were rescreened. 20 infants failed to follow up. 3 infants failed the rescreening and were subjected to BERA within 3 months. On testing with BERA, 2 were found to have severe sensorineural hearing loss. Thus, the prevalence of sensorineural hearing loss in neonates admitted in our hospital was observed to be 6.67 per thousand neonates.

Conclusions: The prevalence of hearing loss was 6.67 per thousand neonates. Hence, Distortion Product Otoacoustic Emissions is an easy, cost effective and reliable method of testing of large number of infants for hearing loss. BERA introduced a new era in hearing screening, but its invasive nature, need for infant cooperation, cost and need for trained audiologist to conduct the test proves as limitations for the test to be used on large number of infants as a screening tool.

Key Words: Distortion Product Otoacoustic Emissions, Distortion product, Brainstem Evoked Response Audiometry, Newborn screening, Hearing screening, Evoked Otoacoustic Emissions.

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INTRODUCTION

"Blindness separates people from things; deafness separates people from people**."** –Helen Keller**.** Congenital and early childhood onset deafness or severe-to-profound hearing impairment may affect the auditory neuropathway of children at a later developmental stage if appropriate and optimal interventions are not provided within the critical period of central auditory pathway development. "One of the most crucial factors in any child's development is the acquisition of spoken language. Spoken language is the doorway to successful communication and the social interaction which is so important to normal human development".

The incidence of hearing impairment in a standardized population of neonates at risk and not at risk to develop hearing impairment ranges from 6-60 per 1000 neonates with an average of 4 per 1000 neonates.[1] Screening is one of the most important methods for early diagnosis of hearing loss.

Otoacoustic Emissions (OAE) is an audiological tool which is sensitive, non invasive, cost and time effective, making it an ideal method for neonatal hearing evaluation. Otoacoustic Emissions (OAE) reflect the status of the cochlea (outer hair cells). A probe microphone similar to that used in acoustic immitance measures the inaudible sounds reflected by vibratory motion in cochlea. One form of evoked OAE is termed the distortion product otoacoustic emission (DPOAE). DPOAEs are produced when two tone stimulus is presented to the ear. Due to non linearities in the mechanism of the inner ear, acoustic distortion products are created at algebraic combinations of the stimulus tones, f_1 and f_2. Though DPOAEs occur at a number of these algebraic combinations, the distortion product at $2f_1$-f_2 is by far the most robust and is the only one that is employed in clinical practice.[2]

BERA (Brainstem Evoked Response Audiometry) is an objective and non invasive method of hearing assessment which detects electrical activity from inner ear to inferior colliculus. It appears to be the most reliable and accurate newborn screening available.[3]

This study is undertaken in order to detect the prevalance of congenital deafness among neonates admitted in our hospital using Distortion Product Otoacoustic Emission and BERA as the testing tools.

OBJECTIVE

To find the prevalence of sensorineural hearing loss in neonates admitted in BLDE Hospital.

REVIEW OF LITERATURE

Neonatal hearing loss has a prevalence that is more than twice that of other newborn disorders amenable to screening such as congenital hypothyroidism and phenylketonuria.[4,5] Early identification and intervention for hearing loss by 6 months of age provides better prognosis in language development, academic success, social integration and successful participation in the society.[6] Late detection causes irreversible stunting of the language and development potential of the child. Detection and rehabilitation of hearing in infants by 6 months of age has proven advantage over those detected after 6 months to acquire normal language regardless of degree of hearing impairment.[7] To maximize the outcome of infants hard of hearing, a universal screening program has to be developed to identify infants by 3 months and provide appropriate intervention by 6 months.

CAUSES OF NEONATAL HEARING LOSS

CAUSES FOR CONDUCTIVE DEAFNESS: [8]

1) Congenital Disorders:

A) Genetic abnormality of external or middle ear:

i**) Deafness present at birth**

Down's syndrome

This is the most common chromosome abnormality syndrome typified by a wide range of abnormalities. Otolaryngologic findings are numerous in these patients and can affect every region of the head and neck. This includes small ears with over-folding of the superior helix, stenotic external auditory canal and eustachian tube dysfunction. There is also an increased incidence of chronic ear disease in affected children due to increased incidence of upper respiratory infections, reduction of B and

T cell function (immune system immaturity), and eustachian tube dysfunction. The hearing loss in Down's syndrome is usually conductive, secondary to the chronic middle ear disease but can also be due to ossicular chain abnormalities, especially the stapes.

Crouzon's Disease

This is an autosomal dominant trait with conductive deafness. The affected children have hypoplasia of mandible and maxilla with a parrot beak nose. There is usually skull deformity (craniostenosis) and exopthalmos. There may be stenosis or atresia of the external auditory canal, absence of tympanic membrane; malleus may be fused to the bony wall of epitympanum. Other features include a deformed stapes and a narrow round window niche. Conductive deafness is present in one –third of children with crouzon syndrome.

Marfan's syndrome

This is inherited as an autosomal dominant trait. Affected children are tall, often with scoliosis and have long fingers and toes. Other feature includes hypotonic muscles, a tendency for lens dislocation and cardiac problems, especially aortic aneurysm. Deafness is a rare finding.

Treacher Collins Syndrome.

Also known as Franceschetti-Zwahlen-Klein Syndrome or Mandibulo-Facial Dysostosis, this autosomal dominant entity is due to mutations on chromosome 5q11. Diagnostic criteria include microtia and malformed ears, midface hypoplasia, down slanting palpebral fissures, coloboma of outer 1/3 of lower eyelids, and micrognathia. The upper airway narrowing can be a major issue in infancy. The size of the nasopharynx is 50% smaller than normal and affected infants are more prone to

obstructive sleep apnea and sudden infant death syndrome. Hearing loss in this syndrome is usually conductive with a wide array of middle ear anomalies present such as monopodal stapes, ankylosed foot plate, malformed incus, cochlea and vestibule abnormalities. The EAC may be absent or stenosed. If sensorineural hearing loss is present, it usually occurs at high frequencies.

Pierre Robin Syndrome

This is an autosomal dominant trait although; in some cases it may be due to intra-uterine disease during the first trimester. The features of this syndrome include cleft palate, hypoplasia of the mandible, glossoptosis, congenital dislocation of the hip and club foot. There may be mental retardation associated with either microcephaly or hydrocephalus.

The external ear may be cup shaped and appear to be low set because of hypoplastic mandible. The middle ear cleft may be absent or there may be thickening of the stapes footplate and crura. The inner ear deformities include: abnormal communications between middle and apical turns of cochlea, a poorly developed modiolus and a narrow internal auditory canal. The audiogram shows a conductive deafness but in cases with inner ear anomalies, the hearing loss is mixed.

Achondroplasia

It is an autosomal dominant trait which mainly affects the skeletal system. In the middle ear the ossicles may be fused to bony margins. The cochlea may be deformed . The hearing loss if present is usually conductive, as a result of the middle ear abnormality and also is a predisposition to otitis media with effusion.

Duane Syndrome

This is an autosomal dominant syndrome with very short neck, congenital paralysis of the 6 th cranial nerve and enophthalmos with conductive deafness.

Apert's Syndrome

This is inherited as an autosomal dominant trait. These children have a high tower skull, a flat forehead, maxillary hypoplasia with high arched palate, cleft palate and saddle nose. The fingers and toes are fused. The audiogram shows conductive deafness of varying degrees. Surgical exploration has demonstrated congenital fixation of stapes footplate.

Otopalatodigital Syndrome

This is a X- linked trait characterized by bossing of frontal and occipital bones, hypertelorism, hypoplasia of the mandible, cleft palate, mild mental retardation, low set pinnae and conductive hearing deafness due to abnormalities of ossicular chain.

ii) Deafness appearing in childhood:

Osteogenesis Imperfecta- This is an autosomal dominant disorder displaying a triad of bone fragility, blue sclera and hearing impairment. The conductive component of the hearing loss is attributed to the thickened and fixed stapes footplate, similar to what is seen in otosclerosis.

B) **Congenital disorders predisposing to otitis media with effusion or infection:**

i) Cystic fibrosis

ii) Immotile cilia syndrome

iii) Cleft palate

iv) Immune deficiency disease

C) **Miscellaneous disorders:**

i) Isolated malformations

ii) Congenital Cholesteatoma

iii) Rhabdomyosarcoma

iv) Fibrous dysplasia

v) Goldenhar's syndrome

D) **Acquired Causes**:

Inflammation

I. Otitis Externa
II. Acute (suppurative) Otitis media.
III. Chronic (suppurative) otitis media.
IV. Acute otitis media with effusion.
V. Chronic otitis media with effusion.

CAUSES OF SENSORINEURAL DEAFNESS:

1) **Congenital disorders**:

A) Genetic:

i) Deafness present at birth-

Deafness alone

a) Michel dysplasia

Total absence of labyrinth as a result of failure of the otic vesicle to separate from the neural ridge.

b) Mondini dysplasia

It affects the cochlea and semicircular canals. The cochlear duct is reduced to the basal coil only. The organ of corti may be absent or reduced to a mound of undifferentiated cells.

c) Bing Sibemann dysplasia

The bony labyrinth is normal with underdevelopment of membranous part.

d) Scheibe (cochleosaccular) dysplasia

It is the least severe and is present in about 70% of cases of congenital deafness. The stria vascularis has alternating areas of aplasia and hyperplasia. The organ of corti is rudimentary and hair cells are sparse or absent. The saccule is collapsed.

Syndrome associated with deafness

a) Klippel-Feil syndrome

The etiology is unclear. Majority are inherited as autosomal dominant trait. The external ear may have microtia with preauricular appendages and atresia of the external auditory canal. Middle ear manifestations include deformity of the incudostapedial joint or stapes. The cochlea is short and there may be distortion of internal auditory meatus. Most have a sensorineural hearing loss.

b) Turner's syndrome

These patients have an abnormal genetic XO pattern. The external ears are low set with large lobes. The mastoid air cell system is poorly developed and there may be abnormalities of stapes. Anderson et al, stated that in their series 64% patients had sensorineural hearing loss.

c) Fanconi's syndrome

The autosomal recessive condition presents with congenital anaemia, skin pigmentation, skeletal deformities and mental retardation. The hearing loss appears to affect high frequencies first and is slowly progressive.

d) Pili torti

In this autosomal recessive disease, dry and brittle hair is associated with sensorineural deafness.

e) Usher's syndrome

It is inherited as an autosomal recessive trait. It is an association of retinitis pigmentosa with progressive sensorineural deafness.

f) Pendred syndrome

This is an autosomal recessive trait. A congenital defect in thyroxine synthesis eventually cause goiter. The sensorineural deafness is severe to profound.

g) Congenital hypothyroidism

It is associated with senorineural or mixed hearing loss, mental or physical abnormalities.

h) Waardenburg's syndrome

This is inherited as an autosomal dominant trait. These are of 2 types. In type 1, medial canthus of eye is displaced laterally and in type 2, medial canthus is not displaced laterally. Twenty percent have a white forelock. 4.5 % have heterochromia iridis. Twenty percent of type 1 Waardenburg's syndrome had a sensorineural hearing loss whereas, 55% of the type 2 were affected.

i) Jervell and Lange-Nielsen syndrome

It is an autosomal recessive disorder. The deafness is severe to profound and bilateral. It is associated with prolongation of QT interval.

ii) **Deafness appearing in childhood**:

Syndromes associated with deafness

a) Alport's syndrome

Children present with hematuria and albuminuria. In 50 % of patients, a high frequency sensorineural hearing loss begins around the age of 10 years. This loss usually progress to become severe.

b) Renal tubular acidosis

It is an autosomal recessive disorder which may be associated with sensorineural hearing loss worse at high frequencies.

c) Refsum's disease

Retinitis pigmentosa with peripheral neuropathy and cerebellar ataxia are features of this autosomal recessive disorder. Sensorineural deafness starts between 10 and 20 years of age

d) Cogan's syndrome

There is interstitial keratitis with sensorineural deafness and vertigo. It usually manifests first in the adolescence with sudden onset of vertigo, tinnitus and rapidly progressive deafness.

e) Norrie's syndrome

It is an X linked recessive disorder. There is progressive blindness, and in some cases, mental retardation. Progressive sensorineural deafness is present in about one third of cases.

B**) Nongenetic- due to intrauterine disease**

a) Infections

These are maternal infections which may be transmitted to fetus across the placenta or at the time of birth. The most important of these infections include rubella, cytomegalovirus, toxoplasmosis, congenital syphilis, herpes simplex.

b) Ototoxic Drugs

The otoxic drugs affect the cochlea of the fetus. The effect of these drugs on the fetal cochlea is similar to acquired causes of deafness. The two most important groups causing potential ototoxicity are loop diuretics and the aminoglycoside antibiotics

c) Other causes - irradiation, ultrasound, maternal diabetes, fetal alcohol syndrome.

2)Perinatal Disorders:

a) Hypoxia

Hall in 1964 described the otopathological findings in neonatal asphyxia, including a decrease in cell number in cochlear nuclei. The cochlea appeared histologically normal. Many authors have suggested that the periods of neonatal apnea and hypoxia strongly predispose to subsequent hearing loss.

b) Hyperbilirubinaemia

The auditory system is particularly sensitive to the toxic effects of bilirubin.[9] Bilirubin, at high levels, can damage retrocochlear structures such as the brainstem auditory nuclei, inferior colliculi, spiral ganglion neurons, and auditory nerve fibers. The effect of hyperbilirubinaemia on auditory dysfunction is generally dose dependent with greater dysfunction noted at higher total serum bilirubin levels.[10]

Bilirubin levels well above exchange transfusion thresholds were associated with sensorineural hearing loss.[11]

c) Preterm delivery and low birth weight

These infants have higher incidence of hearing loss than normal. They are more likely to have suffered episodes of hypoxia or acidosis. They also have immature metabolic functions and kernicterus which can result from smaller increases in serum bilirubin level than in mature neonates. There is also a possibility that deafness and low birth weight are concomitantly caused by the same factor, e.g. Rubella. In the immediate postnatal period, these children spend a variable amount of time in intensive care units in noisy incubators. They are very prone to life threatening infections and are given antibiotics which are potentially ototoxic.

3) **Acquired conditions:**

a) Infections-

i) Complications of otitis media

Toxins produced by cholesteatoma sac cross the round window membrane and cause irreversible cochlear hair cell loss mostly affecting the basal turn of the cochlea.

ii) Viral labyrinthitis

Viral labrynthitis due to mumps, measles, herpes simplex, varicella zoster, influenza virus. [12] Viruses have been implicated in the development of labyrinthine infections for decades. Although strong clinical evidence supports a causative relationship between many viral infections and the development of cochlea-vestibular symptoms, cytomegalovirus is the only perinatal viral infection that affects the labyrinth that has actually been isolated from perilymph or detected within inner ear tissue. Congenital cytomegalovirus infections occur primarily via transmission in

utero, although they may occur at the time of delivery or postnatally. The majority of children born with congenital cytomegalovirus are asymptomatic, and 80% never develop any significant symptoms or disabilities. Occurring rarely, the most severe form of congenital cytomegalovirus infections, termed cytomegalic inclusion disease, affects multiple organ systems and is associated with significant, permanent disabilities. In patients with this severe phenotype, SNHL, microcephaly, and learning difficulties are seen in nearly 50% of cases. Hepatosplenomegaly, jaundice, blueberry muffin rash, and computed tomography evidence of intracerebral calcifications are also typical of cytomegalic inclusion disease.[12]

b) Immunization

Tetanus immunization and vaccination are known to cause peripheral neuropathy. The risk of deafness after MMR vaccination is small and must be weighed against the risk of deafness due to natural disease.

c) Autoimmune deafness

Immunological destruction of the auditory and vestibular systems is a recognized feature of many autoimmune diseases. Immune complexes lodge in the microcirculation of the ear causing obstruction and hypoxia in the distal tissues.

d) Meningitis

The most important cause of acquired sensorineural deafness in children is meningitis. The incidence of post meningitic hearing impairment is reported as varying from 3.5 % to as high as 37.2%.[13] The ear may be affected in different ways by meningitis. Bacterial labyrinthitis due to direct spread of the infection from subarachnoid space through cochlear aqueduct, internal acoustic meatus or endolymphatic duct is associated with profound sensorineural hearing loss. In

children, who have partial or reversible hearing loss there maybe toxic or serous labyrinthitis. The deafness is usually bilateral and profound although maybe less severe and even unilateral. The common organisms associated with sensorineural hearing loss after meningitis are streptococcus pneumonia, haemophilus influenza, neisseria meningitidis

e) Ototoxic drugs

The term ototoxicity refers to the tendency of a drug or chemical agent to cause inner ear dysfunction that produces symptoms of hearing loss and/or dizziness. Many agents can cause ototoxicity, and inner ear tissues may be damaged either temporarily or permanently. The common ototoxic drugs are aminoglycoside antibiotics, antineoplastic agents, loop diuretics, analgesics, quinine and related drugs, erythromycin and related macrolide antibiotics, desferoxamine, and vancomycin.

f) Trauma

A blow to the head, sufficient to render a child unconscious can cause cochlear concussion with a fracture of temporal bone. Transverse fractures of the petrous temporal bone are associated with damage to cochlea or auditory nerve.

g) Metabolic disease

Disorders of microcirculation are common in conditions like diabetic mellitus and are quoted as a cause of deafness.

HISTORICAL PERSPECTIVES OF NEWBORN HEARING SCREENING AND METHODS OF SCREENING

1. SUBJECTIVE METHODS- Free field examination:

A) Behavioral Observation. (4months-2.5 years)

These were first used to screen hearing in United States of America in late 1960s using the auro palpebral response, startle response, limb and head movements to judge a response to high frequency narrow band noise at about 90-100 dB SPL.

It does assess the infants reflexive response to auditory stimuli including warbled pure tones, narrow band noise and speech signals presented through speaker in a sound field. Reflexive responses include full body startle, head/limb reflex and eye blink. Attentive responses include motion cessation, eye widening, and in older infants smiling, laughing, pointing, and cessation /initiation of crying or babbling. In general, responses should be seen within a few seconds of the stimulus presentation.[14]

This method was time consuming, subjective, and identified only infants with bilateral severe to profound high frequency hearing loss. It did not provide ear and frequency specific information and had a high false negative rate. [15]

Table 1: Auditory maturation of normal hearing infants

Auditory maturation of normal hearing infants	
Age	**Auditory maturation**
0-4 months	Newborn behavioral responses to auditory stimulus are limited to reflexive actions
4-7 months	Response to sound is a horizontal head turn toward the side of sound source 4 month-head turn is slow 6 month –head turn is definite and brisk
7 month	Localize sound source in lower plane(looking downward)
9 month	Locate sound source when presented over height
12 month	Locate sound source in any plane on either side of the body easily and briskly

B) Crib-O-Gram:

In order to decrease the observer error associated with behavioral testing, the Crib-O-Gram was developed in 1974 by Simmons and Russ.[16] It is a more objective method for screening which uses motion sensitive transducer placed under the crib mattress or between the crib and the frame. The equipment was designed to present a 3000Hz sound at 92dB SPL (sound pressure level) to the infant. A motion sensitive

transducer was placed under the mattress to detect a startle response. A strip chart recorder printed out the infant's activity prior to and following the stimulus presentation.

It was in 1985 that, Durieux-Smith and coworkers found that one-third of the infants with normal ABR responses failed the Crib-O-Gram. They concluded that only severe to profound losses were identified and that the Crib-O-Gram also failed to detect unilateral hearing losses.[17]

C) Auditory Response Cradle (ARC):

The Auditory Response Cradle is a fully automatic microprocessor that was designed in Great Britain. It consists of a pressure sensitive mattress and headrest that monitors head turn, head startle and body activity. The baby's respiration activity is monitored using a polyethylene band over the abdomen. A high pass band noise is presented bilaterally via earphones at 85dB SPL and was used to detect the more common congenital hearing losses in the high frequency regions. The infants motor and respiration responses are detected automatically and stored in the microprocessor. The Auditory Response Cradle also has the capability to present an equal number of silent trials to determine if the baby's responses are to the stimuli rather than spontaneous movement. The baby is considered a 'pass' when 97% of the responses, within 10 trials, are not by chance. The baby 'refers' when this criteria is not met. The screening procedure usually ranged from 2-10 minutes.

In 1992, Tucker and Bhattacharya[18] described the use of Auditory Response Cradle on 6000 infants. Infants who failed 2 screens were referred for an audiometric evaluation consisting of ABR, OAE, and acoustic reflex testing. The results of this research showed an initial 8.1% fail rate that was reduced 1.7% (N=102) after the

second screen. Seventy-nine (1.3%) were determined to have normal hearing following the audiologic evaluations indicating the high false positive rate of the ARC screening procedure. Twenty infants were found to have hearing loss, which included 5 with conductive hearing loss. The cohort was followed for three years and an additional 7 children were found to have permanent hearing loss. This technique showed great promise but the objective measures of Otoacoustic emissions and auditory brainstem response techniques that were emerging simultaneously based on physiologic responses were considered more reliable.

D) Visual reinforcement audiometry (children between 6 months and 2.5 years)

The child sits on his mother lap in sound treated room facing the examiner. The child is conditioned and trained to look to the light (toy) when sound is presented. The stimuli are warble tones from 250-8000Hz delivered from loudspeaker with variable intensities. The goal is to obtain ear and frequency specific information before the child loses interest. The method requires that a child turn his or her head toward the sound source that is coupled with conditioned reinforcement, such as a lighted toy. In an ideal situation, two audiologists are required : one to initiate the stimulus and one to observe child's response. The response should be time-locked to within a few seconds of the stimulus presentation for it be considered a true response. The paradigm is stimulus-response-reinforcement: a stimulus (either pure tone or speech) is presented and the child learns that a response (turning head towards the stimulus source) will result in reinforcement ,usually an animated toy enclosed in a smoked plexiglass.[14] To obtain ear specific information, insert ear phones or standard head phones should be used. It is possible that, some children will not accept any form of earphone and the sound field testing will have to be substituted. In this case

the results are a reflection of better hearing ear only. Moreover, it needs exceptional child and examiner cooperation.

E) Conditioned play audiometry (children 2.5 to 5 years)

Conditioned play audiometry is the next level of behavioral testing and can often be used in the children 2 years of age to 4-5 years of age. The primary goal of conditioned play audiometry is to obtain ear and frequency specific thresholds via air and bone conduction allowing for the diagnosis of conductive, sensorineural or mixed hearing loss. For air conduction testing, insert earphones should be used whereas a bone oscillator is placed for bone conduction testing. The test used a form of operant conditioning where the child is taught to wait, listen for a tone or speech signal, and then perform an activity as a response. Most popular task includes putting a block in a box, pegs in a board or doing a simple puzzle. In some situations with consent of parents or guardian, the child can be offered a tangible item (food or candy) as reward. It is important not only to choose a task that the child can perform with ease but also to switch task to avoid boredom.

2. OBJECTIVE METHODS

A) Conventional audiometry (Adult type)

It can be used for children 5years and older. The child is seated in a sound treated room wearing headphones. The child asked to raise his hand in response to sounds that are presented at variable intensities at frequency range of 250 – 8000Hz. In this way, both air-conduction and bone conduction thresholds are measured together with speech audiometry.

B)Tympanometry[19]

It measures the middle ear status and is not a measure of hearing thresholds. It can quickly differentiate between middle ear effusion, eustachian tube dysfunction and normal middle ear. It needs passive cooperation i.e. the child sits quietly or sleeps. A probe is placed in the ear canal of the child and acoustic admittance of the ear with various amount of air pressure in the ear canal is measured.

Types of Tympanograms:

- **Type A**: Normal

  Type As- middle ear fibrosis, Tympanosclerosis, Congenital fixation of footplate, osteoarthritis of malleo-incudal joint.

  Type Ad- Ossicular chain disarticulation, hyper mobility of tympanic membrane due to thinning
- **Type B**: middle ear effusion.
- **Type C**: Eustachian tube dysfunction.
- **Type D**-Flaccid or scarred tympanic membrane
- **Type E**-Thick graft following myringoplasty or tympanoplasty

3) OAE

As early as 1948, Gold reported that the outer hair cells of cochlea could produce energy by an active mechanical process.[20] It was not until 1978 that David Kemp demonstrated that the cochlea is capable of producing low intensity recordable sounds called Otoacoustic Emissions (OAE).[21] In 1986, the first commercially available instrument using a personal computer to record OAE, the ILO88 was designed by Kemp and colleages.[22]

Otoacoustic emissions (OAE) are low intensity sound caused by motion of the eardrum in response to vibrations from the outer hair cells of the cochlea only when the organ of Corti is in normal condition and when middle ear system is operating normally. They are of four types-

1) Spontaneous OAE (SOAE)

2) Transient OAE (TEOAE)

3) Distortion product OAE (DPOAE)

4) Sustained frequency OAE.

Kemp, the English biophysicist who discovered otoacoustic emissions and published the first scientific description of TEOAEs introduced DPOAEs shortly thereafter.[23] Distortion product Otoacoustic Emissions (DPOAE) is generated by frequency specific region of the cochlea & has the potential to test micromechanical properties of outer hair cells in frequency specific regions.[24]

C) Auditory brainstem response

BERA is an objective tool for assessment of auditory function in neonates.[25]. It is a far field recording of the synchronized response of a large number of neurons in the lower auditory portions of the auditory pathway. It was first described by Sohmer and Feinmesser in 1967.[26] Shortly thereafter, Jewett and his colleagues published a series of studies that contained detailed description of BERA.[27]

BERA is composed of seven scalp positive waves which occur within the first 10 milliseconds following acoustic stimulation by transient signals.[28] The ABR derived from the scalp is recorded electroencephalogram by averaging. The 3 major

waves are I, III and V. Wave I is generated by auditory nerve while wave V originates in the region of inferior colliculus. Hall et al found that BERA is a method with high sensitivity and specificity.[29]

PREREQUISITE

The recording is carried out in a sound treated room. Since the electrodes should be placed over the head, it should be oil-free. The subject is best tested in a supine comfortable, position thus cervical myogenic activity which generates electrical interference will be minimized. Active children can be sedated.

PROCEDURE

The stimulus either in the form of click or tone pips is transmitted to the ear via a transducer placed in the insert ear phone or head phone. The wave forms of impulse generated at the level of brainstem are recorded by placement of electrodes over the scalp.

Electrode placement – the standard electrode configuration for BERA involves placing a non-inverting electrode over the vertex of the head and inverting electrodes placed over the mastoid prominence. One more earthing electrode is placed over the forehead.

The waveforms recorded are very weak and they need to be amplified. This amplification is achieved by improving signal to noise ratio by means of 3 parallel approaches namely filtering, repeated stimulation, polarity alteration.

The impulses when recorded contain a series of peaks and troughs. The positive peaks are referred to by Roman numerals I to VII. These peaks are considered to originate from the following anatomical sites.

Wave I: distal portion of eighth cranial nerve

Wave II: proximal portion of eighth cranial nerve

Wave III: cochlear nucleus

Wave IV: superior olivary complex

Wave V: nuclei of lateral lemniscus

Wave VI & VII: inferior colliculus

The parameters compared at 70dB level stimulus are:

1. Absolute latency of waves I, III, V in milliseconds of each ear separately.
2. Amplitude in micro volts of wave I and V.
3. Wave V / I amplitude ratio.
4. Interpeak latencies of wave I, III, V in milliseconds.
5. Hearing threshold in dBHL of each ear separately.

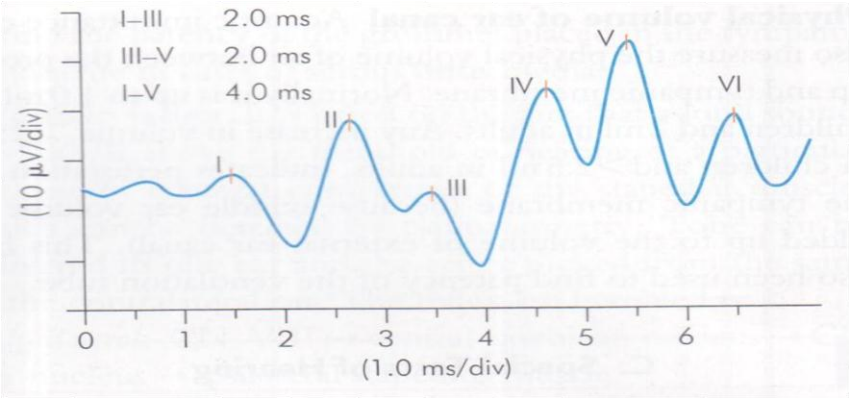

Figure 1: Brainstem auditory evoked potentials

SCREENING

Permanent childhood hearing loss has been described as a "neurologic emergency" as extended periods of auditory deprivation have a significant impact on the overall brain development and sensory integration of the child. According to Northern & Downs screening involves the application of tests, examinations and procedures to a large number of individuals, in order to make a differentiation between individuals who have a high probability of having a certain disorder, and those who have a low probability of having a certain disorder. Screening is not a diagnostic procedure, and individuals who fail the screening process are therefore referred for diagnostic testing.[30]

Research on neuroplasticity has suggested that early auditory stimulation is necessary for developing a child's auditory potential.[31] Hence early identification of hearing loss is absolutely necessary to ensure maximal communicative and literacy development for children with hearing loss.

NEWBORN HEARING SCREENING

Newborn hearing screening (NHS) involves the screening of auditory abilities of newborns. A newborn is any infant in its first four days of life. The goal of NHS is to identify newborns with a substantial hearing loss, so that treatment and early intervention can be implemented by the age of six months. Two commonly used techniques for NHS are the measurement of Otoacoustic emissions (OAEs) and automated auditory brainstem response (AABR) .

INFANT HEARING SCREENING (IHS):

Infant Hearing Screening refers to hearing screening of all newborns as well as young children up to the age of 12 months. Infant hearing screening (IHS), like NHS refers to application of techniques, such as AABRs and OAEs, in order to differentiate between children requiring further diagnostic hearing testing, and those with normal hearing.

UNIVERSAL NEWBORN HEARING SCREENING (UNHS):

UNHS denotes hearing screening provided for all newborns and infants in a health care facility. The principles of UNHS are all infants have access to hearing screening, using a physiologic measure, such as OAE's or ABR's. This can be achieved in the following way. Newborns who receive standard routine care should have access to hearing screening during their hospital stay at birth admission. Newborns born outside of a large hospital should be referred for hearing screening before the age of one month. And lastly, all newborns in the NICU must receive hearing screening before being discharged from the hospital.

TARGETED NEWBORN HEARING SCREENING (TNHS)

TNHS refers to risk-based NHS. Only infants who display risk factors for hearing loss receive hearing screening.[31] Risk factors for hearing loss are defined by the Joint Committee on Infant Hearing.[32] TNHS is a more cost-effective way of NHS, as only 10% of infants display risk factors for hearing loss. However, it has the disadvantage of not identifying a large number of infants with hearing loss, as 50% of infants with hearing loss display no risk factors [33]

TNHS aims to detect permanent bilateral hearing loss of at least 40 dB averaged over the frequencies 0.5, 1, 2, and 4 kHz.[34]

EARLY HEARING DETECTION AND INTERVENTION (EHDI)

Early hearing detection and intervention (EHDI) services for infants with hearing loss is endorsed by the JCIH Year 2000 Position Statement [32]. The goal of EHDI is to ensure maximal communicative and literacy development for children with hearing loss. EHDI comprises the following: All infants should be screened for hearing loss prior to hospital discharge, using objective or physiological tests, such as OAEs and AABRs. Confirmation of an infant's hearing loss should take place by the age of three months, and appropriate family-centered intervention should commence by the age of six months. Furthermore, infants who display risk factors for late onset or progressive hearing loss should receive ongoing audiologic monitoring for three years, at appropriate intervals.[32]

Components of a good screening program:

For infant screening program to be successful,

- The testing method must be fast, inexpensive, and simple to perform upon all infants prior to discharge from the hospital in order for the screen to cover a large population.
- Not require infant cooperation apart from lying still during the period of testing.
- It should have high sensitivity in order to prevent undiagnosed cases from passing through the screen.

- Should have high specificity in order to prevent false positives from burdening audiological staff with rescreening of failures and increasing parental anxiety.
- The instrument used must function well under standard clinical conditions and be operable by testers with a wide range of experience.
- The condition being screened for is not otherwise detectable by clinical means.
- There should be minimal loss to follow-up. Follow up of at least 70%.
- Alternative platforms for Infant Hearing Screening programs, besides hospital based screening including immunization clinics, should take the unique socioeconomic, demographic and healthcare infrastructures of each country and region within the country into account.

High Risk Registries:

A national committee on neonatal hearing screening, chaired by Marion Downs was formed in 1968, and lead to the development of the Joint Committee on Infant Hearing (JCIH).

In 1973, the Joint Committee on Infant Hearing Screening recommended five criteria for identifying infants at risk for hearing loss. In 1975, Mencher recommended that all infants should be universally screened using the JCIH five criteria and also recommended that the World Health Organizations, national and local governments legalize a program of infant screening. In 1982, the JCIH expanded the criteria to six . The Committee recommended the use of auditory evoked potentials, as part of the audiometric evaluation, for those infants who were identified as high risk. This led to the evolution of screening infants with electrophysiological measures rather than behavioral testing.

It was in 1987 that, Mahoney and Eichwald estimated that 15% of infants were subjected to the high risk register but less than half were actually tested for hearing.

High risk registries were also plagued with high false-positive information on family history. Research demonstrated that the high risk register criteria recommended by the JCIH identifies only 50% of infants with significant hearing loss.

The JCIH 1990 Position Statement once again expanded the high-risk criteria to include stigmata or findings associated with a syndrome known to include sensorineural hearing loss and prolonged mechanical ventilation for duration equal to or greater than 10 days.[35]

This has further been modified in 2007 to include:

1. Caregiver concern regarding hearing, speech, language, or developmental delay.
2. Family history of permanent childhood hearing loss.
3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: Extracorporeal membrane oxygenation, assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide /Lasix), and hyperbilirubinemia that requires exchange transfusion.
4. In utero infections, such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis.
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.

6. Physical findings, such as white forelock, that is associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher's syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
8. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
9. Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
10. Head trauma, especially basal skull/temporal bone fracture that requires hospitalization.
11. Chemotherapy.[36]

RELAVENT ANATOMY

THE COCHLEA

The human cochlea is situated in the inner ear and consists of a spiral tubular duct embedded in petrous bone, lined by membranes. Cochlea comprises 2¾ turns of a spiral. It has a central pillar, the modiolus, and a bony cochlear canal. The base of the spiral protrudes into the middle ear as the promontory of the medial wall. The bony wall of the cochlea has two defects; each covered by a thin membrane- the round window and the oval window. The latter contains the footplate of the stapes, which is held in place by the annular ligament. A cross-section of one turn of the cochlea shows that the cochlea is divided into three segments. From above down, these are the scala vestibuli, the scala media and the scala tympani and each scala is fluid-filled. The scala media contains endolymph and the other two contain perilymph. There is communication between the perilymph of the scala vestibuli and the scala tympani at the apex of the cochlea at a point known as the helicotrema. The scala media is a closed, triangular cavity bounded above by Reissner's membrane and below by the basilar membrane. The stria vascularis forms the base of the triangle lying against the bony wall of the cochlea. The organ of Corti sits on the basilar membrane and it is here that the transduction of sound happens.

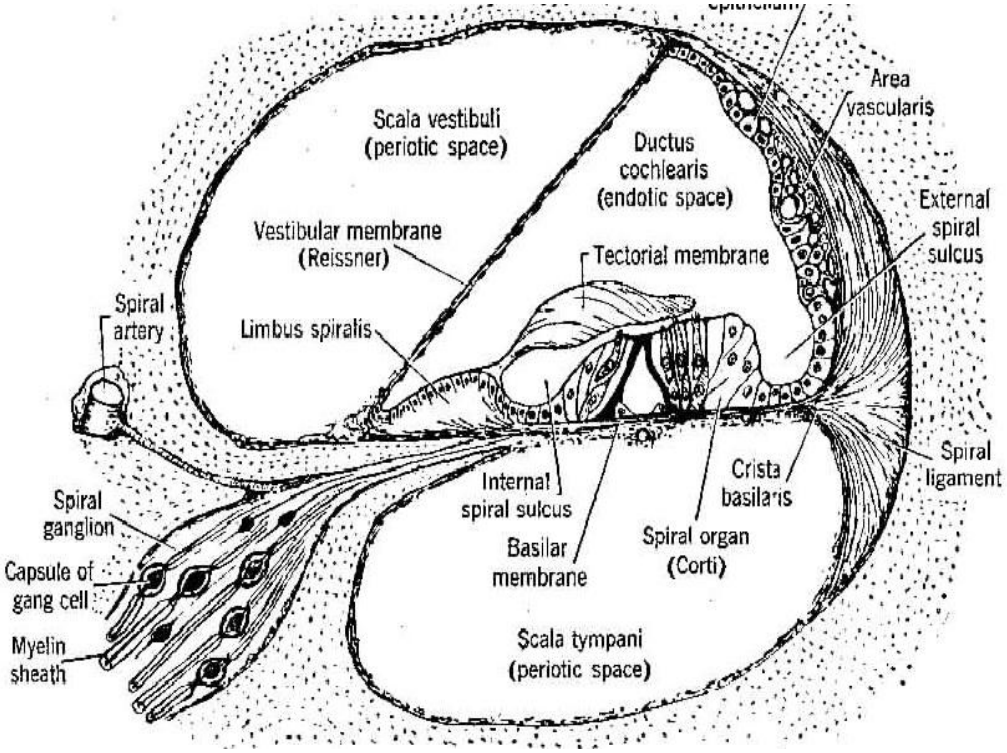

Figure 2: crossection of cochlea with organ of corti

THE ORGAN OF CORTI

The organ of Corti extends across the upper surface of the basilar membrane from the spiral limbus situated over the osseous spiral lamina to the Claudius cells that lie between the edge of the sensory region and the outer anchorage of the basilar membrane. The basilar membrane runs between the inner and outer bony spiral laminae. It is narrower and more taut at the base of the cochlea, wider and floppier at its apical end. At its inner end, sits the organ of Corti. This comprises the limbus, the tectorial membrane, the inner and outer rods of Corti with the tunnel of Corti between them, one row of inner hair cells, three rows of outer hair cells and supporting cells of Claudius, Deiter and Hensen. The organ of Corti contains hair cells that give rise to nerve signals in response to sound vibrations. There are approximately 12,000 outer hair cells and 3,500 inner hair cells in humans. The auditory branch of the eighth cranial nerve (the vestibulocochlear nerve) contains fibers that run from the cochlea to

the brain stem afferent fibers and efferent fibers that run in the opposite direction. Around 90% of the afferent fibers come from the inner hair cells. Each fiber comes from only one cell but each cell may have up to 10 fibers. The remaining 10% of the afferent fibers and all of the efferent ones are associated with the outer hair cells. Each of the nerves associated with the outer hair cells is connected with many cells. Each hair cell has many hairs (cilia). The outer hair cells have 80-100 stereocilia. The cilia of each outer hair cell are arranged in a 'W' shape (with a very shallow central notch) and those of the inner hair cells, the stereocilia are arranged in a U shaped manner. The hairs of the outer hair cells are embedded into the tectorial membrane whereas at rest the hairs of the inner hair cells are not. The cilia of each hair cell are connected by tip links. The outer hair cells contain contractile actin and myosin fibers which allow for the cells to alter their length. The spiral organ in basal turn of cochlea is concerned with high pitched sound, that in the apical turn with low pitched sound.

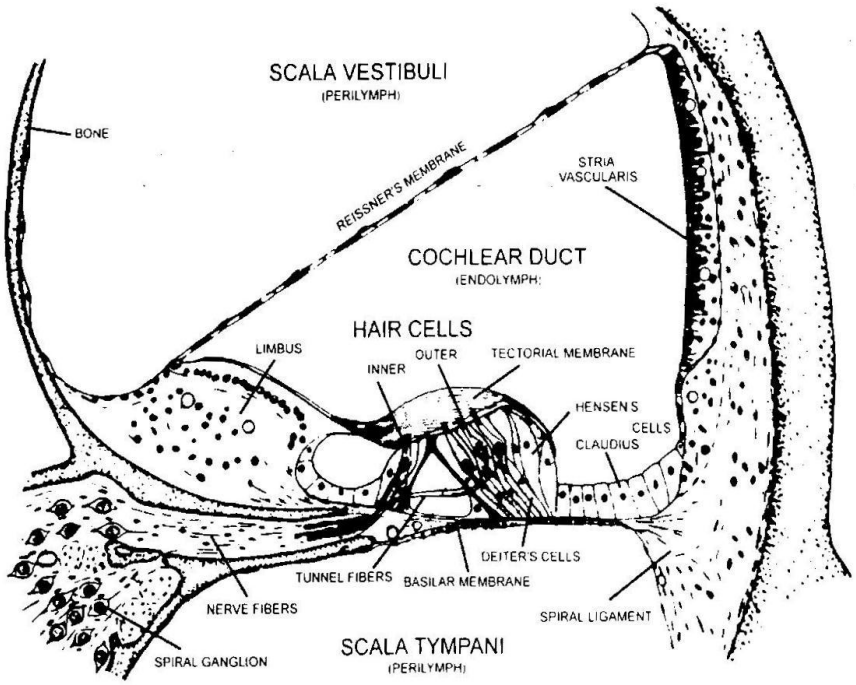

Figure 3: Organ of Corti

Physiology of conduction of sound[37]

Hearing is the vital basis for acquisition in speech and language and these skills in turn are most important tools of constructive thought. The sound conduction mechanism comprising the ossicles forms a link, extending from the pinna to the organ of Corti.

Its functions include:

1. Collection and transmission of sound energy involving impedance matching at every stage and particularly matching between the external air and cochlear fluids.
2. Protection of the inner ear from the excess loud sounds, a function carried out by the tympanic muscle sacrificing the sensitivity of low intensity sound levels.

Middle ear sound conduction[38]

The sound waves in air cannot be transmitted efficiently to the fluid medium which fills the cochlea without the help of some device to overcome the impedance mismatch. The role of middle ear is to match these impedances by acting as an acoustic sound pressure transformer. It also has to provide for acoustic separation of the round window from oval window if movement of stapedial foot plate is to be transmitted to the perilymph in the cochlea.

The round window membrane provides the elasticity needed for the transmission of the sound wave into the fluid medium. If an incident sound wave falls on the oval and round windows simultaneously, the perilymph column will be unmoved, because the pressure exerted by the footplate at the oval window would be exactly resisted by pressure, acting in the opposite direction on the round window

membrane. This difficulty is overcome by protection of the air cushion within the tympanum and by the preferential channeling of the sound waves from the tympanic membrane through the ossicular chain to the oval window. By these means the round window is acoustically isolated from the oval window.

Ossicular movement[37, 38]

The vibrations of the tympanic membrane are conveyed to the malleus through its handle. The malleus and incus move as one functional unit, except at very high intensity, rotating in and out through a tiny arc, about an axis which passes from the anterior process of the malleus backwards to the end of the short process of the incus. The oscillating movement of the stapes in the oval window, received from the long process of the incus, is in and out, like that of a piston, when amplitude is low. At higher amplitude the footplate executes a rocking motion about a vertical axis through its posterior edge. When the stapedius muscle contracts, in response to sound pressure levels 80 dB or more above the threshold, the mode of stapes movement may change to one of longitudinal rotation about its long axis. This form of vibration attenuates sound levels reaching the cochlea, especially in the low frequencies.

Sound pressure transformation

The problem of impedance matching, so that relatively light and inelastic air can impart its energy to relatively dense and highly elastic fluid, is solved by two mechanisms – ossicular lever action and hydraulic action. The pressure of sound waves on the stapes footplate is almost twenty times greater than on the tympanic membrane as a result of the combined effect of these mechanisms.

Ossicular leverage

It is provided by the movement of the malleus – incus complex acting as a lever about its axis of rotation. The handle of the malleus is about 1.3 times longer than the long process of the incus, when each is measured from its tip to the fulcrum at the axis. This factor of 1.3 is the size of the mechanical gain provided by the lever action.

Hydraulic action

It depends on the relative surface areas of the tympanic membrane and the stapes footplate. Anatomically, the surface area of the tympanic membrane is about 21 times that of the footplate. It is known that the central 2/3rd of area of tympanic membrane moves as a unit and it is this central part which provides the area to relate to that of the footplate. The functional ratio of tympanic membrane surface area to footplate area is then 2/3rd of 21:1 = 14:1, which is the mechanical advantage derived from the hydraulic action. The combined benefits of lever action and the hydraulic action provide an increase of pressure at the oval window of 14 x 1.3 or just over 18 times.

Auditory tube function

Effective sound transmission through the middle ear and into the cochlea requires that the air in the middle ear is maintained at a pressure level identical to that of the ambient air in the external acoustic meatus. Deviations from this ambient level of pressure, the impedance of middle ear increases. The pressure of air in the middle ear must at all times be kept at the ambient external level as a prerequisite for efficient middle ear function and inner ear sound conduction. Auditory tube maintains this pressure. Auditory tube obstruction raises the threshold of hearing by 30-40dB. In

normal individuals, the limits of variation for compliance and resistance are much narrower for female subject, but the average values are similar to both sexes.[39]

Sensorineural function

Air conducted sound waves are admitted to the cochlear perilymph through the oval window, and the information they convey emerges at the other end of the cochlea as nerve impulses in the afferent fibres of the cochlear nerve. The cochlea is a tube filled with perilymph, coiled on itself 2¾ times. Along the length this tube is divided into two channels by a cochlear partition. The upper channel is the scala vestibuli, into which the oval window opens. The lower channel is the scala tympani, which is sealed at its end by the round window membrane. These two perilymphatic channels communicate with each other only at the cochlear apex, through the helicotrema, scala media containing the endolymph is separated from scala vestibule by Reissner's membrane and from scala tympani by basilar membrane supporting the organ of corti and associated structure. The basilar membrane is 35 mm long gradually increases in width from 0.08 mm at the base near the oval window to 0.5 mm at the apex. There is progressive increase in mass and decrease in stiffness along the length of the membrane. An account of sensorineural function demands a description of movement of the cochlear partition by sound waves, of the conversion of the mechanical energy of movement to electrical energy (transduction) and of the electrical events induced in the fibres of cochlear nerve.

Movement of the cochlear partition[37]

1. Helmholtz's Place Theory[40]: He suggested that basilar membrane consist of a series of tuned resonators. In this theory, any segment of the basilar membrane is activated by a sound wave of the resonant frequency of that segment, with high

frequency waves exciting segments in the basal turn and low frequencies exciting the more apical regions.

2. Rutherford's Telephone Theory: According to this the frequency of activating sound wave is signaled by the rate of discharge in the cochlear nerve fibres. The latent period of nervous action limits this theory to the perception only of frequencies below 1000 Hz, if the relationship between sound wave frequency and nerve impulse has a simple ratio of 1:1.

3. Wever's Volley Theory: Combines both place and telephone principles postulating that:

a. High frequencies are perceived as per place theory (in the basal turn).
b. Low frequencies (below 1000 Hz) stimulate nerve action potentials at a rate equal to the stimulus frequency.
c. Intermediate frequencies are represented in the auditory nerve by asynchronous discharges in groups of neurons, whose combined activity represents the frequency of the stimulus.

4. Von Bekesy's Travelling Wave Theory[41]: Each wave increases in amplitude until it reaches a maximum at a place, which is specific for its frequency and then rapidly dies away. Successive trains of waves produced by a sustained tonal stimulus have an envelope with a maximal displacement at a site determined by the stimulus frequency. High frequency waves activate only the basal turn, which appears to move as one. Lower frequency waves travel farther along the whole length of the partition to the apex before reaching their maximum. Sharpening of this frequency sensitivity takes place partly in the cochlea and farther by neural mechanisms in the brain. The traveling wave uniquely represents the frequency of excitation and many of its physical character may subsequently be used by brain for finer pitch assessment.

Cochlear transduction

Endolymph has a composition different from that of perilymph, which is an ultrafiltrate of plasma. Particularly the high potassium level of endolymph (150 mEq/litre compared with 6 mEq/litre in perilymph) and the low endolymphatic sodium level (1.5 mEq/litre compared with 150 mEq/litre in the perilymph). Electrically endolymph in the scala media has a positive potential of +80 mV relative to that of perilymph. This is endocochlear potential. The interior of hair cell has a potential negative relative to that of perilymph, of the order of -70 mV, so there is a potential difference of 150 mV between the endolymph and the interior of the hair cell. Deformation of the cochlear partition by the traveling wave bends both the basilar membrane and the tectorial membrane, but since these pivot about different axes, the displacement 'wipes' the tectorial membrane with a shearing action, across the tops of hair cells. This changes the resistance of the surface of the hair cell in contact with the endolymph and so alters the amount of current flowing through the cell. In this way, movements of the cochlear partition modulate the current flowing through the hair cell body. This causes the electrical excitation of the afferent nerve endings. The stored electrical energy represented by the large potential differences, endows the cell with amplifier properties, so that tiny amounts of mechanical energy modulate the output of a greater electrical energy source. The endocochlear potential is changed by displacement of basilar membrane and resulting hair bending and the change is maintained as long as mechanical deformation persists. Movement upwards, which is the direction causing neural excitation, is associated with a reduction of the potential.

Cochlear nerve activity[37]

This nerves act on ‘an all or none’ basis, which implies that the nerve discharges only when its threshold of excitation is exceeded. A second action potential can follow only after a refractory period, during which the nerve regains its resting state. At rest, all the cochlear nerve fibres are discharging. Each nerve fibre responds most readily to a stimulus of a particular frequency its characteristic frequency and less readily to stimuli of frequencies differing from that. The threshold for excitation increases the more the stimulus frequency differs from the characteristic frequency. Tuning curve of acoustic nerve fibres shows that frequency sensitivity is much finer or the tuning is much sharper, than the mechanical response of the basilar membrane. The tuning curves overlap and broaden at high intensities, high intensity sounds excite fibres whose characteristic frequencies are more and more distant from the stimulus frequency. The tuning curves of low frequency fibres are symmetrical but those of fibres with high characteristic frequencies are asymmetrical with a sharp high frequency cut off. This means, at high intensity, all fibres with characteristic frequency below the test tone will be activated.

Thus, the intensity of a sound is indicated by the rate of spike discharge and the number of active fibres. Frequency information is available from the site of maximum excitation and from the spatial pattern and responses of excited fibres. The frequency and intensity determinants of nerve fibre impulse rates are finally separated centrally by the activity of neurons in the brain with which the auditory fibres eventually connect.

Cochlear microphonics

These are alternating potentials, originating in the hair cells, which accurately follow the pattern of the sound stimulus, and the movements of the cochlear partition. They persist after nerve conduction ceases and appear as responses in opposite senses with upward and downward movement of the partition. Summating potentials show as steady baseline shifts in the recording. They reflect steady changes in endocochlear potential.

PHYSIOLOGY OF OTOACOUSTIC EMISSIONS

The answer lies in an understanding of current theories of cochlear function. The travelling wave theory was developed by Von Bekesy in the 1940's.Von Bekesy studied the effect of sound upon the excised human cochlea. He discovered that sound caused the motion of stapes which set up a wave in the basilar membrane that progressed from base to apex. When the basilar membrane moves, the hair cells are set into motion and an electromechanical response is elicited. While an afferent signal is transmitted and an efferent signal is emitted.[42]

When the frequency of the stimulus was altered, the travelling wave would peak at different sites on the membrane. High frequencies caused maximal vibration at the base of the cochlea whereas lower frequencies had a greater effect near the apex. In fact, each site on the basilar membrane has its own characteristic frequency that always produces maximum resonance at that point. Von Bekesy proposed that the peak of the travelling wave was detected by the local hair cells, which responded by depolarizing and initiating a sensory nerve signal. In this way, the ear was able to distinguish between sounds of different frequency.

The travelling wave theory suggests that the cochlea is entirely passive. Over the last two decades, however, experiments on live cochleas have produced findings which imply an additional, active process. Also, the displacement of the basilar membrane is far greater at its characteristic frequency than the theory would predict. It was in 1978 that David Kemp found, that sounds can be recorded in the external ear that are almost certainly of cochlear origin. On some occasion these emissions contain more energy than the eliciting stimulus. These findings led to the theory of a 'cochlear amplifier'.[43]

The exact nature of the cochlear amplifier is unknown but it almost certainly involves the mechanical action of the outer hair cells upon the basilar membrane. Outer 28 hair cells are located in the organ of Corti on the basilar membrane arranged in 3 rows and are motile. The outer hair cells have stereo cilia which are linked to each other and, therefore, move as a unit. These outer hair cells are believed to underlie OAE generation. It is generally agreed that some active mechanism causes the travelling wave to reach a very abrupt peak at, or near, its site of characteristic frequency. It is the sharpness of the peak which accounts for the exquisite sensitivity of the mammalian ear. As a by-product, the amplifier produces a secondary disturbance of the basilar membrane which generates a lower amplitude wave which passes back along the membrane and through the middle ear. It is this wave which emerges in the external ear as an OAE. Thus, an efferent signal is transmitted back through the auditory pathway, and the signal is measured in the outer ear canal.

The cochlea behaves more like a room with its natural acoustics enhanced by an imperfect acoustic amplification, hence the potential for feedback howl (SOAE) and inter-modulation distortion(DPOAE).Thus, OAE's reflect the status of outer hair cells of the cochlea. OAE's are a byproduct of sensory outer hair cell transduction and are reflected as echoes into the external auditory canal. OAE's are pre-neural in origin and dependant on the integrity of outer hair cells. Evoked emissions are essentially an all-or nothing response. Normal ears produce OAEs, whereas ears with hearing losses over 30 dB fail to produce a response.

Otoacoustic Emissions are generated only when the organ of Corti is in near normal condition, and they emerge only when middle ear system is operating normally. The sounds generated by the cochlea are small but potentially audible, sometimes amounting to as much as 30 dB SPL. They can emerge spontaneously

because sound already in the cochlea perpetually re-circulates, but more commonly OAE's follow acoustic stimulation. No electrodes are needed to observe OAE's. They are not electrical in nature but vibratory responses. In fact, microphones are used to detect them and they are converted to an electrical signal to process them more easily. OAE's are created by motion of the ear drum driven by the cochlea through the middle ear chain. To record OAEs a healthy middle ear with good sound conduction is needed.

Closing the ear canal is an essential part of the OAE technique and enables any oscillatory movement of the ear drum to compress and rarefy the air that otherwise would flow silently in and out of the ear canal, without creating sound.

Furthermore, the loudness of the stimulus required to elicit an emission bears little resemblance to the subject's auditory threshold. For these reasons, evoked emissions are of little use in determining the severity of any hearing loss.

Healthy cochlea does contain a mechanism capable of returning sound to the middle ear, and significantly impaired cochleas don't do the same. This makes OAE a uniquely valuable clinical tool.

Following Kemp's discovery, several other types of OAEs have been identified.

These include four types of Otoacoustic emissions have been described:

- Spontaneous Otoacoustic Emissions (SOAE's)
- Transient Otoacoustic Emissions (TOAE's) or Transient Evoked Otoacoustic Emissions (TEOAE's)
- Distortion Product Otoacoustic Emissions (DPOAE's)
- Sustained-Frequency Otoacoustic Emissions (SFOAE's)

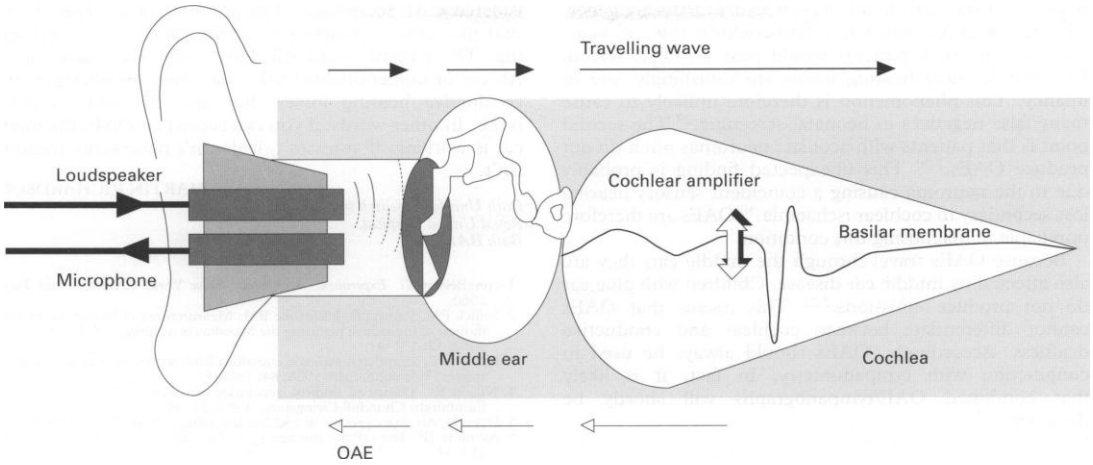

Figure 4: Physiology of OAE

TYPES OF OTOACOUSTIC EMISSIONS:

The sound emitted by the biological activity of the normal cochlea which can be picked up, recorded and measured by placing a microphone in the deep external meatus is called otoacoustic emissions.[44]

Spontaneous Otoacoustic Emissions

The spontaneous otoacoustic emissions are generated automatically, i.e., spontaneously and they don't require any external stimulation for being generated. The SOAE are found in 50% of normal hearing subjects. The spontaneous Otoacoustic emissions are low intensity, continuous, very narrow band or pure tone sounds that can be picked up by placing a very sensitive miniature microphone in the external auditory meatus just lateral to the ear drum. If the spontaneous Otoacoustic emissions can be detected in an individual, it indicates that the individual has normal hearing at the frequency in which SOAE is being generated. The frequency of SOAE is usually between 3000-4000 Hz in infants and 1000-2000 Hz in adults. Over 60,

years the intensity and the prevalence of SOAE decreases even if the hearing is normal.

Sustained-frequency Otoacoustic Emissions

SFOAEs are responses recorded to a continuous tone. Because the stimulus and the emission overlap in the ear canal, the recording microphone detects both. Therefore, interpretation depends on reading a complicated series of ripples in the recording. At present, SFOAEs are not used clinically.[45]

Transient Evoked Otoacoustic Emission(TEOAE)[44]

TEOAE are measurable in essentially all normal hearing persons, irrespective of the age. The character i.e. the frequency, latency, duration etc of the evoked Otoacoustic Emission's depends upon the character of the evoking stimulus. The OAE in response to a sound stimulus of a particular frequency occurs from that part of the cochlea which is tuned for that particular sound. If a low frequency sound is presented to the cochlea, the evoked otoacoustic emissions will originate from the apical region of the cochlea. Similarly if a high frequency is presented to the cochlea, the evoked OAE will originate from the basal turn of cochlea. TEOAES can be elicited by click stimuli as well as tone bursts (pure tone sounds). The TEOAE elicited in response to a click or a tone burst will be a broad band sound with more energy in the mid frequency region and the TEOAE generated by presenting a tone burst will have the same frequency as that of the evoking sound. TEOAE's are usually elicited by sounds of 80-85dB SPL. The average loudness recorded with a sound stimulus of 80-85 Db is about 20 db in neonates and less than 10 db in adults. The evoked OAEs reduce very rapidly as the deafness increases and are undetectable when the deafness is above 30 to 35 dB SPL

DISTORTION PRODUCT OTOACOUSTIC EMISSIONS

When two tones are presented simultaneously to healthy cochlea, the response measured in the ear canal will contain several tones that are not present in the eliciting stimuli. These additional tones are called distortion products. These are attributable to nonlinear processes of normally functioning cochlea. They are vibratory in response, not electrical in nature.

Stimuli consist of two pure tones at two frequencies (i.e., f1, f2 [f2>f1]) and 2 intensity levels (i.e., L1, L2). By convention, the lower frequency pure tone is referred to as the f1 primary and its level as L1. The higher frequency pure tone is referred to as f2 primary, and it's level as L2. The relationship between L1-L2 and f1-f2 dictates the frequency response. The two tones are selected such that the frequency ratio between the tones f2/f1 is 1.22 which is known to produce the largest (2f1-f2) distortion product at most frequencies in humans, robust DPOAE are produced. The most frequently measured acoustic inter-modulation – distortion product is at frequency 2f1-f2, although the cochlea also produces concurrently DPOAEs at other frequencies (f2-f1, 2f2-f1, 3f1-2f2) in response to such bi-tonal stimulation. Indeed, the only emitted distortion component utilized for clinical purposes has been the 2f1-f2. Therefore clinical DPOAE measurements are generally made with both stimulus intensity and frequency ratios optimized for maximum DPOAE 2f1–f2 intensity.[46] Although 2f1-f2 DPOAEs can be detected in essentially all normal human ears, they are typically extremely small i.e.5-15 dB SPL. However, they generally are charted according to f2 because that region approximates the cochlear frequency region generating the response. To yield an optimal response, intensities are set so that L1 equals or exceeds L2. The most appropriate stimulus intensity for evoking DPOAE in most clinical applications is in the range of 50-70db SPL. With higher stimulus

intensities there is a chance that passive responses will be recorded, responses that do not reflect outer hair cell activity. However lower intensities often fail to generate detectable DPOAE, even from persons with reasonably normal hearing sensitivity. An appropriate relative intensity difference is 10-15db. A setting of 65/55 dB SPL L1/L2 is frequently used clinically. DPOAE's are measured by using signal averaging techniques. The averaged waveform detected in ear canal, then undergoes spectral analysis to determine SPL at 2(f1-f2) for each tonal pair. In general, the inter modulation distortion that produces DPOAEs is thought to arise from fundamental processes within the cochlea, particularly those associated with the non linearity of outer-hair cell motion. Like other types of OAEs, DPOAEs are thought to be generated by the active cochlear process responsible for enhancing basilar membrane vibration.

The most common format of plotting a DPOAE is DP gram. The DP gram is measurement of DPOAE amplitude as a function of stimulus frequency. The stimulus frequency as the f2 primary in Hertz is plotted along horizontal axis. The DP amplitude is plotted in db SPL on vertical axis. For each stimulus frequency, the noise floor in the region of DP frequency (2f1-2f2) is also plotted. To be considered a valid DPOAE, the DP amplitude must exceed the noise floor by at least 3db. The noise floor is determined by calculating the SPL in a narrow band of frequency above and below either side of 2f1- f2. With exception of frequency below 1500 Hz, noise estimated lowered around 6db, distortion product response to noise ratio of 20db or greater throughout most of frequency range tested. Below approximately 1500Hz, noise amplitude increases secondary to biologic noise.

Their primary advantage is that they offer objectivity in evaluating frequency specific regions in the cochlea. The frequency range of the DP gram is approximately 500-8000Hz. However, OAE data do not translate into threshold data.

In infants and newborn, OAE amplitude is 5db greater than those measured for older children and adults. The precise mathematical relationship of DPOAE to the eliciting tones is a unique characteristic of mammalian ear. DPOAEs allow greater frequency specificity and can be used to record at higher frequencies than TEOAEs. Therefore, DPOAEs may be particularly useful for early detection of cochlear damage as they are for ototoxicity and noise-induced damage. Reliability of DPOAEs is greatest above 1000 Hz. DPOAEs can be elicited by asymmetrical protocols (75-65 dB SPL) testing the frequencies 1.5, 2.0, 3.0, 4.0 and 6.0 kHz. The DPOAEs are found to be more immune to noise than TEOAEs and are very useful in PASS borderline cases.

OAE's and ABR reflect functional integrity of the portions of the auditory pathway that are most susceptible to diseases or lesions associated with hearing loss. Hence, these instruments are used as a proxy for hearing tests that can only be performed when the infant is older. [24]

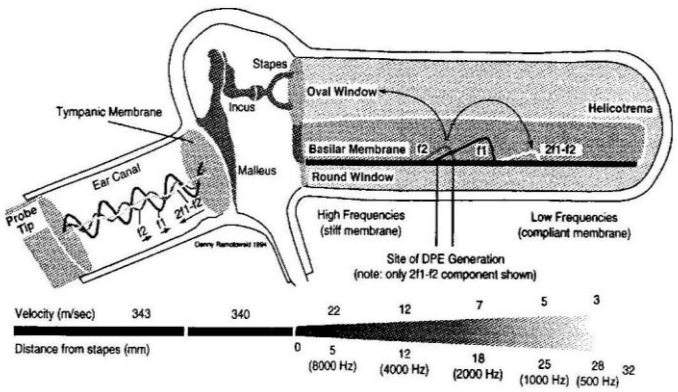

Figure 5:Production of DPOAE

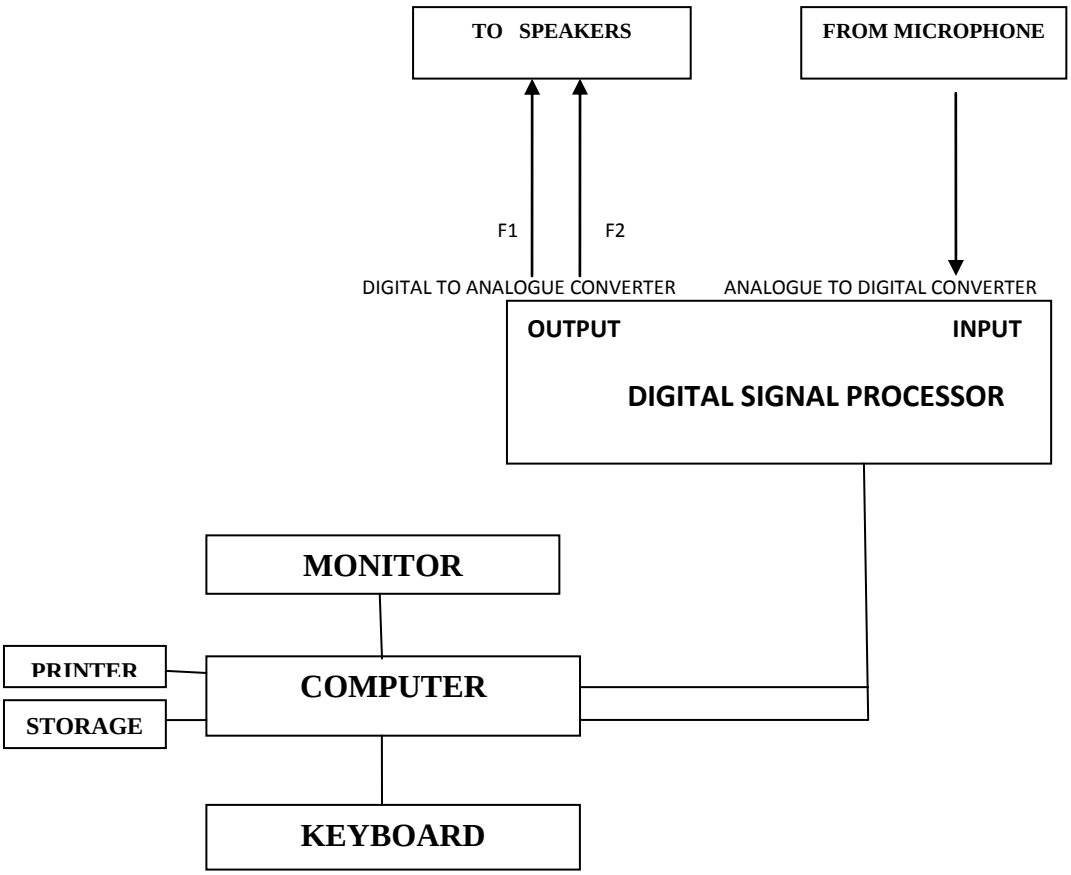

Figure 6: schematic diagram of components of a typical DPOAE measurement device

RECORDING PARAMETERS.[47]

Prerequisite required for conducting DPOAE screening include:

- Unobstructed outer ear canal-The transmission of DPOAE's in the reverse direction of the primaries from the cochlea back to external canal depends upon the integrity of ossicles and tympanic membrane.
- Seal of the ear canal with the probe- Proper probe fit is critical in the usage of OAE instruments, without which, background noise levels of 45dB SPL can prevent obtaining a response via OAE device.
- Optimal positioning of the probe- Manipulation of the pinna can allow for the opening of collapsed ear canals found in newborns.
- Absence of middle ear pathology.
- Functioning cochlear outer hair cells
- A quiescent patient: Excessive movement or vocalization may preclude recording.
- Relatively quiet recording environment: A sound booth is not required, but a noisy environment may preclude accurate recording. As instruments may be unable to differentiate background noise from the expected output, and thus be unable to calculate a result. This would necessitate immediate retesting until a determination could be made, increasing time and costs of the screening program.
- Screener training allowing for the proper handling of instrumentation as well as minimizing other problems.

INTERPRETATION

PASS -This means that the infants' outer hair cell functioning in each ear is normal at the time of testing.

REFER: Though a refer result is suggestive of outer hair cell dysfunction, other causes for refer result includes debris in external ear, fluid in middle ear, noisy environment or if the infant is very restless. Such infants require reassessment a retest with OAE. If retest also indicates refer, confirmation by BERA is required.[48]

FACTORS AFFECTING DPOAE SCREENING:

1. Impact of Ear canal obstruction and Middle ear disorder on OAE screening:-

It has been observed that OAE testing has a high false positive rate (up to 15.6%) in the first 24 hours of life, falling to about 4% by 72 hours. Bonfils and colleagues in 1992, reported results on the screening of 27 ears of 27 normal neonates, aged 1 to 5 days (mean 2.4 days) via DPOAE. Six presented without low frequency emission, who on otoscopic examination revealed middle ear effusion. The authors concluded these effusions as well as nasal respiration stated to be low frequency noise led to difficulties in detecting emissions in the lower frequencies.[49]

Tsui and coworkers [50] in 2008 reported no increase in percentage of normal results for infants who received 2nd DPOAE in neonatal period compared to those screened beyond age of 30 days, as vernix and middle ear fluid were less likely to mask the true negative result. Thus, it can be concluded that the high referral rates on initial screening may be due to the presence of vernix caseosa in the external auditory canal or the presence of fluid in the middle ear that is expected to resolve within a few days following birth. Since OAE technology uses reverse transmission of cochlear emissions through the middle and outer ear it, is more likely to be influenced by the

presence of debris in the auditory canal than an ABR device. Thus, unless multiple rescreens of failures is anticipated and accounted for, or some form of cleaning the external auditory canal is attempted, using this technology in the initial hours postpartum might result in unacceptable levels of referral rates.

2. Effects of Environment and Impact of noise on DPOAE screening

It was in 1994, that Brass and Kemp[51] suggested that the screening test be conducted in a quiet or soundproof room. However, a soundproof room was not essential if a quiet room was available.

In 1992, Lasky et al, [52] used DPOAE technology to examine frequencies below 3 kHz infants and in adults, showed noise levels 5-15 dB higher in infants than in adults. While screening in a NICU environment there is difficulty in obtaining clear DP emissions from neonates aged 3 days to 5 months at f2 frequency of 977 Hz due to elevated noise floor. He concluded that noise levels in ten full-term newborns were significantly higher than in ten adults for 19 out of 20 f2 frequencies measured from 537 to 10,000 Hz. Their opinion was that this was due to the subject movements, muscle-tone, or snoring that are characteristic of neonates and also background noise in a well baby nursery prevented reliable recordings below 1.7 kHz by a DPOAE device in the testing of 54 full-term infants.

Sheppard et al,[53] noted that during the screening of 77 full-term infants under maternity ward conditions, respiratory noise could be observed in the spectra of a DPOAE device below 3 kHz. The greatest numbers of emissions were detected in the 2 to 4 kHz range, with an f2 of 3 kHz eliciting the greatest number of emissions among the infants tested. As environmental noise would have been excluded by the

noise rejection system, hence the main source of noise in the study was the infants themselves.

In general terms thus, the influence of noise appears to be particularly problematic in the screening of newborns. Noise can be attributable to environmental noise in the ward, subject noise of the infant, and equipment noise within the instrument itself. As the activity level of the infant influences the degree of subject noise, screening the infant while he or she sleeps aids in reducing the noise, yet loud breathing of the infant may still interfere.

3. Age at screening

In 1993, Vohr and coworkers also reported that waiting as long as possible prior to discharge before screening the baby provides more opportunity for debris in the external auditory canal to clear naturally. This was confirmed by data from Rhode Island Hearing Assessment Project (RIHAP) in neonates tested before 24 hours of age. When the examiners waited until the infants were at least 24 hours of age, the "pass" rate for a sample of over 4,000 infants at RIHAP increased from 70% to 82%.[54]

In a study conducted by Gabbard et al, newborn screening procedures, BERA and DPOAE's on 110 neonates with a mean age of 15 hours was performed. 107 (97%) passed the BERA whereas 69 neonates (63%) passed the OAE. A significant difference (at the level 0.05) was found between neonates younger than ten hours of age, neonates 10 to 24 hours of age and those more than 24 hours of age. Hence they showed that a significant difference in the age-related effect was identified during the OAE screen test.[55]

In 1998, Bantock and Croxson [56] reported that testing at three to four weeks of life could lower the initial failure rate of OAE. They reported a nine percent failure rate for both ears when the OAE screening test was carried out in the first few weeks of life. However, when they were retested one month later, only 0.8% failed.

OAE has been reported to have a high false positive rate (about 15% at the first screen on day one and then reduces by about 50% with each retest). Screening using auditory brain responses (ABR) technique is associated with a much lower false-positive rate. About 5% on day one and reduces to about one percent by the second retest.[57] Hence we can conclude that young neonates were less likely to pass the OAE screen test than older neonates.

4. Inappropriate probe size:

Especially for small premature babies, it will cause false-positives. In addition, with the DPOAE, probe fit can influence the attenuation of background noise entering into the probe. According to Rhode Island Hearing Assessment Project (RIHAP), probe must be stabilized before the test can be conducted.[56]

5. Loss in follow-up

Infants not being brought for follow up screening are another reason for reduction in sensitivity. In a study done at Malaysian hospital, there were many defaulters during both the second stage (18.4%) and third stage (35%) of screening. Some of the reasons identified were, parent's misconceptions about their infants hearing, parents being too busy to bring their infants for follow-up, or were not fully briefed on the importance of this test and therefore did not see the need for follow-up. [56]

6. Auditory Neuropathy

This includes central auditory nervous system dysfunction and Cranial Nerve VIII auditory dysfunction. The advent of OAE recordings has opened a new area of auditory investigation in auditory neuropathy. Classic auditory neuropathy is characterized by the presence of OAEs or enlarged cochlear microphonics, abnormal ABR findings, and, often, absent or abnormal behavioral responses to sound. ABR abnormalities consistent with auditory neuropathy include absence of all ABR waveforms or prolonged interpeak latencies. The disorder can be idiopathic. The cause of auditory neuropathy sometimes is unknown; however, the following conditions may be associated with pediatric auditory neuropathy: hyperbilirubinemia, neurodegenerative diseases, neurometabolic diseases, demyelinating diseases, hereditary motor sensory neuropathologies (e.g., Charcot-Marie-Tooth diseases with deafness), inflammatory neuropathy, hydrocephalus, severe and/or pervasive developmental delay, ischemic-hypoxic neuropathy, encephalopathy, meningitis, cerebral palsy.[47]

7. DPOAE and ABR

In 1991, Kennedy and colleagues[58] studied the auditory functioning 370 infants, of both high and low risk using standard Auditory Brainstem Response (ABR), automated analysis of ABR and automated analysis of evoked Otoacoustic emissions (OAE). They reported that automated OAE testing was quickest (median 12.5 min) and least invasive (no scalp electrodes). Bilateral failure rates (and upper 95% confidence limits) with a stimulus 35-36 db above normal hearing threshold level 9nHL) were 3.0% with automated OAE, 3.2% with ABR and 2.7% automated

ABR. Hence, automated OAE was the most sensitive for subsequently confirming hearing impairment.

Independent studies in Sheffield and Southampton compared OAEs with ABR in a total of over one thousand infants. Both groups found that OAEs are generally very well tolerated and can be recorded in half the time of ABRs. Compared with hearing status at 8 to 18 months, both OAEs and ABR had a sensitivity of 100% for sensorineural hearing loss. OAEs had a specificity of around 80% whereas the figure for ABR was about 90%.[58] OAE screening therefore failed more normal ears. Nevertheless, the Sheffield group concluded that OAE screening is still worthwhile because of its simplicity. They also suggested that OAEs could be used as an initial screen with the more complicated ABR being reserved for those who fail. The Southampton investigators also reached similar conclusions. They calculated that the two tier screen would have a specificity of over 99%.

ABNORMAL RESULTS IN OTOACOUSTIC EMISSIONS TESTING:

Non pathologic problems that can cause absence of OAEs

- Poor probe tip placement or poor seal: Most current equipment alerts clinicians to these problems.
- Standing waves: Most current equipment alerts clinicians to standing waves.
- Amniotic fluid occluding the canal or blocking a probe port
- Debris and foreign objects in the outer ear canal
- Vernix caseosa in neonates: This is common immediately after birth.
- Uncooperative patient: Usually, recordings simply are not obtained.

Pathologic problems that can cause absence of OAEs

Outer ear

- Stenosis
- Atresia of EAC
- Otitis externa
- Cyst
- Tympanic membrane - Perforation of the eardrum

Middle ear

- Otosclerosis
- Middle ear disarticulation
- Cholesteatoma
- Cyst
- Abnormal middle ear pressure
- Bilateral Otitis Media

Cochlea

- Exposure to ototoxic medication or noise exposure: OAE changes may precede threshold changes in the conventional frequency range.
- Any other cochlear pathology

Conditions that do not affect OAEs

- CN VIII pathology: only if the cochlea is also affected (e.g., vestibular schwannoma that decreases cochlear vascular supply), OAEs are affected.
- Central auditory disorder

Conditions that elicit abnormal OAEs and normal behavioral thresholds

- Ototoxicity
- Vestibular pathology
- Tinnitus: OAEs may be abnormal in the frequency region of the tinnitus.
- Excessive noise exposure

Conditions that elicit normal OAEs and abnormal behavioral thresholds

- Auditory neuropathy
- Functional hearing loss, Attention deficits, Autism

Thus the information obtained by OAE testing can be used to:

- Screen hearing.
- Estimate hearing sensitivity within a limited range.
- To differentiate between sensory and neural components of sensorineural hearing loss.
- Test for functional hearing loss.[47]

SENSITIVITY AND SPECIFICITY OF DPOAE:

The result of the OAE screening performed by White et al in 1993, on 1,850 neonates showed a sensitivity of around 100% and a specificity of 73%. According to that study, OAE was moderately specific but very sensitive. Ng and Yun reported the sensitivity and specificity of OAE with respect to BAER was 95% and 93%, respectively.[56]

Bantock and Croxson and coworkers[56] in 1998 conducted OAE screening on 700 neonates with risk factors for hearing loss and on 1,492 infants without any risk

factors. They found that the sensitivity was l00% in both groups. The specificity in both groups was 94% and 91%, respectively. On the second stage-screening test done about six months after, the sensitivity remained 100% and specificity improved to 99.3% in a group with no risk factors for hearing loss.

In 1998, Salata and colleagues[59], calculated sensitivity and specificity of DPOAE on 104 infants screened in a quiet room by comparing it to results of BERA. He concluded that Otoacoustic emission screening protocols have been associated with higher false-positive rates than BERA screening protocols.

In a study done by P Torroco and colleagues[60], from June 1999 to June 2001, a total of 2,567 infants were screened with DPOAE's. Following first test, 77% infants passed the test, 6.9% had bilateral refers another 16.1% had unilateral refer. Of the 591(21%) infants who had failed the first screen, underwent subsequent DPOAE screen. During the second DPOAE screen, it was observed that 568 (96%) showed bilateral pass, 11 infants (1.8%) showed unilateral pass and 13 infants (2.2%) showed bilateral refer results. On further investigating, these 24 infants with DPOAE, 3 infants passed the screen and 18 were classified as deaf. He thus concluded that sensitivity of 1st OAE was 100% & the specificity of 1st test 77.49% ,positive predictive value of 3.05 and quotient of probability 4.44.The specificity of second test is 99.88, positive predictive value 85.7, quotient of probability of 84.8%.This suggests that, the first test if not normal the probability of having hearing loss is 3.05 % and the second test shows this the clinical suspicion rises to 85.7 %.It thus can be observed that, if the first OAE screen is a refer, the probability of having hearing loss is 3.05%, while a refer on second OAE testing clinical suspicion rises to 85.7.

Other studies on neonatal hearing evaluation

It was in 1998, that Ochi and co-workers[61], tested the ears of 36 neonates (aged from 3 days to 5 months) in a NICU environment with both DPOAE and ABR devices. Comparing the ABR results to the DPOAE device, both devices showed pass rates of 68.7% (46 out of 67 ears passed according to test criteria), yet there were 4 out of 36 infants with differing results. Two infants appear to have been passed by the DPOAE device and not the ABR, and 2 failed the DPOAE device and passed ABR. Yet based on the ABR thresholds chosen, the authors concluded the DPOAE did not fail to detect severe hearing loss.

A study by M Owen et al in 2001, showed that the first test pass rate in their study was 92.7% (bilateral) and 95.7% (unilateral), rising to 96.3% and 97.7% respectively after the second test. The unilateral or bilateral screening pass rate of 97.7% in this study is comparable with that of the Wessex study of 98.4%. Of the 13 babies referred to the audiology department for bilateral OAE test failures on two occasions, all but one subsequently went on to have normal BERA.[62] This suggests that, in most cases, there was sustained inhibition of the OAE response by factors such as fluid, wax, or debris in the external or middle ear.

Similar study was done at Hospital University Kebangsaan, Malaysia during April 2003 to December 2003. 4,219 infants were born in the hospital, 3,762 (89.2 percent) underwent OAE screening. In the second stage screening at three months of age, only 39 (65 percent) patients turned up. Of these, ten infants passed the OAE test and 29 failed. However, when these infants underwent BERA, 13 had normal BERA and 16 have abnormal BERA. The prevalence of hearing loss in this study was 0.42 percent (16/3,762).[56]

At the University Hospital of Ferrara, a total of 4269 full-term newborns were tested during period January 2000-December 2004, by DPOAE-DPOAE-ABR. Screening of well-babies (newborns with no risk factors) was carried out within the first 48 hours of life, using a three-phase (DPOAE-DPOAE-ABR) procedure. The presence of an OAE response in both ears was considered as a "PASS". The criteria for a PASS were based on previously established signal and signal to noise ratio (S/N) values. In the event, if an acceptable OAE response was not present even in one ear, a second test OAE test was performed within 15-30 days. If the result was again a "REFER", an ABR test within the third month of age was done. The prevalence of bilateral hearing loss, in this group, was estimated as 0.07%.Of the 654 NICU babies tested, the prevalence of bilateral hearing loss was, therefore, 1.07%. Despite the combined use of OAEs and ABR, in the NICU population, since 2003, no cases of Auditory Neuropathy have been identified.[63]

A study was done, at Christian Medical College Hospital, Vellore from February 2005 to July 2005. Five hundred neonates were screened with distortion product Otoacoustic emission for hearing loss, 9.2% of whom had one or more risk factors. Although 6.4 % had hearing loss at initial assessment only 1.6% had hearing loss on retesting with DPOAE .Thus retesting with OAE before an ABR helped to exclude patients without hearing loss. The frequency of moderate to moderately severe hearing loss in this study was 0.6%.[64]

From 1st September 2007 to 31st March 2008, a retrospective study was conducted at Union Hospital (Hospital A) and Queen Elizabeth Hospital (Hospital B) to assess the feasibility of a staged combined distortion product otoacoustic emission test (DPOAE) and ABR screening protocol for implementation in local hospitals. A

group of 3,006 infants at Hospital A underwent hearing screening using a combined DPOAE and ABR screening protocol. Results were compared with the results of an ABR-only screening protocol administered to a group of 3,330 infants at Hospital B. The combined DPOAE and ABR protocol had a final referral rate similar to that of the ABR-only protocol, but was about 2.5 times cheaper and almost 3 times faster.[65]

The results of screening by the otoacoustic emissions application in 904 newborns, at the Delivery Ward in the Clinics for Gynecology and Obstetrics at the Clinical Center "Zvezdara" in Belgrade, revealed passing on the first test in 86.3%, and in the second in 99.3% of newborns. In the study, two newborns with unilateral hearing impairment was detected.[66]

Studies conducted in the Clinical Center Kragujevac, in 2009, 1994 newborns out of who 1778 full term and 216 premature born children were examined. Analysis showed that a higher test passing was reached in the full term delivered children(92.5%), compared to 55.1% in preterm newborns, which represents a statistically significant difference.[67]

MATERIALS AND METHODS

The present study "**Neonatal Hearing Evaluation- A Hospital Based Study**" was done in BLDEU's Shri B. M. Patil Medical College, Hospital and Research Centre, Vijayapur during the period of October 2014 to August 2016.

Study group and method of collection

320 neonates were selected at random were evaluated by means of:

1. Proper history.
2. Clinical examination including anthropometry, general examination and otoscopy.
3. Distortion Product Otoacoustic Emissions (DPOAE) testing of neonates was done at 48-96 hours of birth. For pass cases, no further testing was done. For refer cases repeat DPOAE testing was done at 45-60 days of life, failing which such infants were subjected to Brainstem Evoked Response Audiometry testing (BERA) within 3 months to confirm hearing loss.
4. For DPOAE testing, Echolab Otoacoustic Emssions system, Labat, Italy was used. BERA was tested using Clarity Octopus 4CH BERA machine.

Procedure:

A proper history was taken from the parents about the antenatal period, mode of delivery and postnatal period. Also, the following information of the infant was noted: gestational age, sex, prenatal risks, birth weight, APGAR score at 1^{st} and 5^{th} minute and postnatal complications if any. After otoscopic examination of the ears, DPOAE screening was done. With the infant lying comfortably on the bed or the mother's lap, testing was carried out in a sound treated room. Probe with a soft flexible tip was gently inserted into the outer part of the ear canal so as to obtain adequate seal. Once the probe tip was in place, the test was started. The machine delivers two pure tone stimuli with frequencies f1, f2 (f2/f1 is 1.2) and intensity L1, L2 (L1: 65, L2:55). A robust DPOAE was obtained with FDp: 2f1-f2 .The DPOAE amplitude and noise floor were recorded by machine.

Multiple responses were averaged. All DPOAEs were analyzed relative to the noise floor. A Sound to Noise ratio 5 or more was taken as cut off as pass for a f2 frequency. The test for an ear is considered pass by the machine if pass result were obtained for two out of 5 f2 frequencies. For a quiet and cooperative infant, recording usually required less than a few minutes per ear. For an uncooperative or noisy infant, recordings took significantly longer or had to be postponed till infant slept.

BERA testing done in a sound treated room with infant lying supine after adequate sedation. Clarity Octopus 4CH BERA machine was used. After securing the electrodes, a click stimulus was presented to each ear individually and characteristic wave forms produced were noted.

Inclusion Criteria

A study group consisting 320 neonates chosen at random from the department of Pediatrics was evaluated in the department of Otorhinolaryngology and Head and Neck Surgery, BLDEU's Shri B. M. Patil Medical College, Hospital and Research Centre, Vijayapur during the period October 2014 to August 2016. Neonates were subjected to DPOAE at 48-96 hours of life. For pass cases no further testing was done. For refer cases, repeat DPOAE testing was done at 45-60 days of life, failing which such infants were subjected to Brainstem Evoked Response Audiometry (BERA),within 3 months to confirm hearing loss.

Exclusion Criteria

1. Infants with obvious congenital aural and head & neck deformities.
2. Infants, whose parents did not give consent for the procedure.
3. Infants with acute illness.

Sample Size

Incidence of hearing loss in neonates is 4 per 1000.[1] At 99% confidence interval and ± 1 margin of error, the sample size is 263, using the statistical formula

$$N = (Z\alpha)^2 \times P \times Q \; / \; d^2$$

$$= 2.57 \times 2.57 \times 0.4 \times 99.6 \; / \; 1 \times 1$$

$$= 263$$

Where, N = sample size

p = incidence rate,

q = 100 – p

d = margin of error.

Dropout rate = 20%

So, N = 263 + 53 = 316. Approximately 320

Statistical analysis

All characteristics were summarized descriptively. For continuous variables, the summary statistics of N, mean, standard deviation (SD) were used. For categorical data, the number and percentage were used in the data summaries. Chi-square (χ^2)/Fisher exact test was employed to determine the significance of differences between groups for categorical data. If the p-value was < 0.05, then the results will be considered to be significant. Data were analyzed using SPSS software v.23.0. Results were tabulated using bar diagrams.

RESULTS

It was seen that of 320 neonates who underwent first screening, 281 infants passed and 39 failed the first screen.

Table 2: Distribution of the cases according to first DPOAE screening

First DPOAE screening	N	Percentage
B/L refer	19	5.9
B/L pass	281	87.8
L refer, R pass	10	3.1
R refer, L pass	10	3.1
Total	320	100.0

Graph 1: Distribution of the cases according to first DPOAE screening

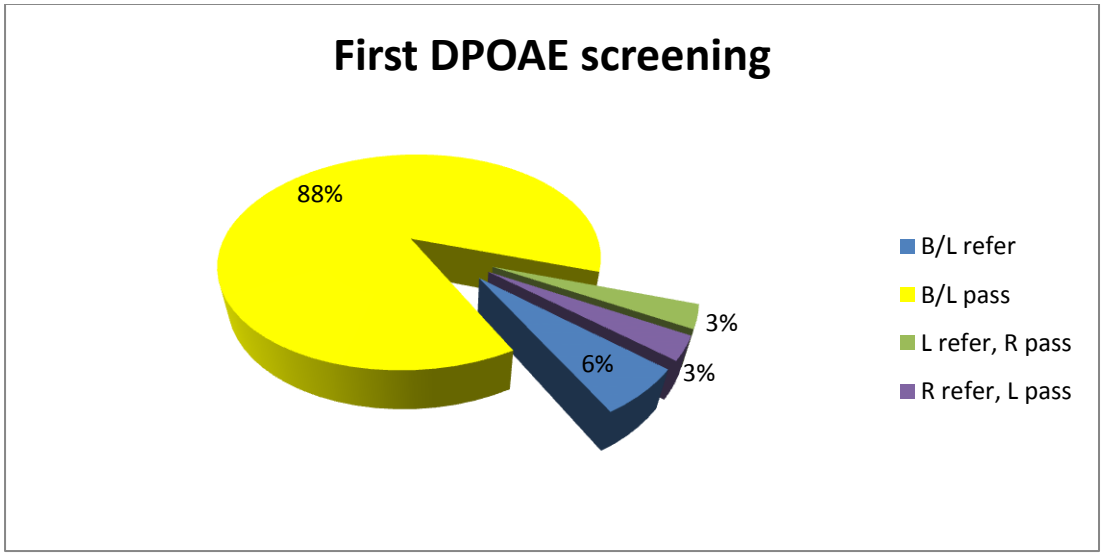

Table3: Distribution of the cases according to second DPOAE screening

39 infants failed in the first DPOAE screening where called for second DPOAE screen at 45-60 days of life. 20 infants who failed the first screening failed to follow up for the subsequent screening. 19 infants were screened of which only 3 failed the screening test.

Second DPOAE screening	N	Percentage
B/L Refer	3	7.7
B/L Pass	16	41.0
Failed to follow up	20	51.3
Total	39	100.0

Graph 2: Distribution of the cases according to second DPOAE screening

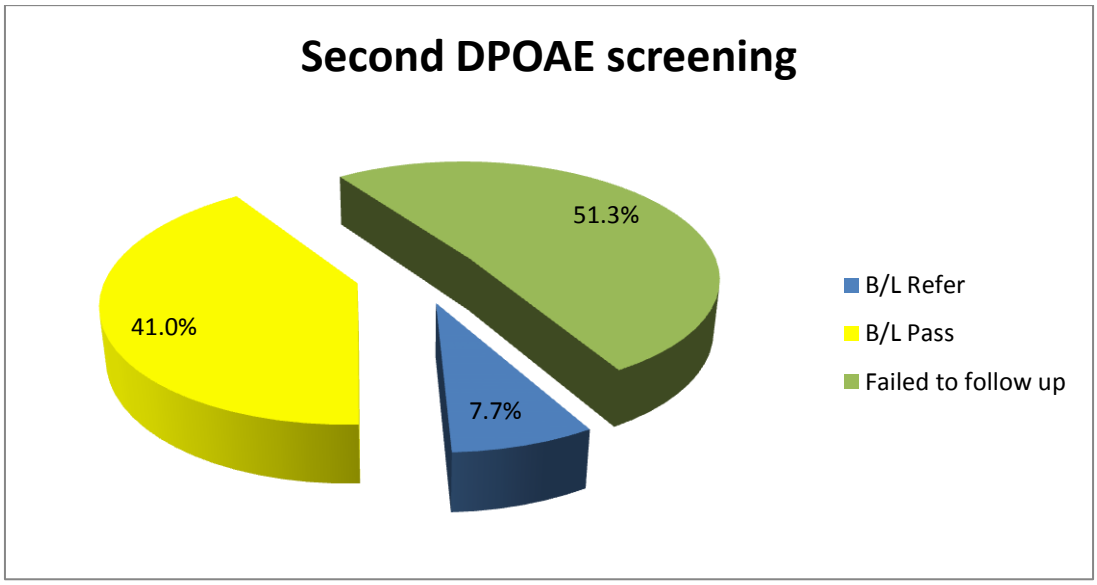

Table 4: Distribution of the cases according to BERA

BERA was conducted on 3 infants who had failed the second DPOAE screen. It was observed 2 infants had bilateral sensorineural hearing loss.

Therefore, the 2 out of 300 infants had hearing loss (excluding 20 failed to follow up cases). Thus prevalence of hearing loss is 6.67 per thousand newborns admitted in our hospital.

BERA	N	Percentage
Hearing loss	2	66.7
Normal	1	33.3
Total	3	100

Graph3: Distribution of the cases according to BERA

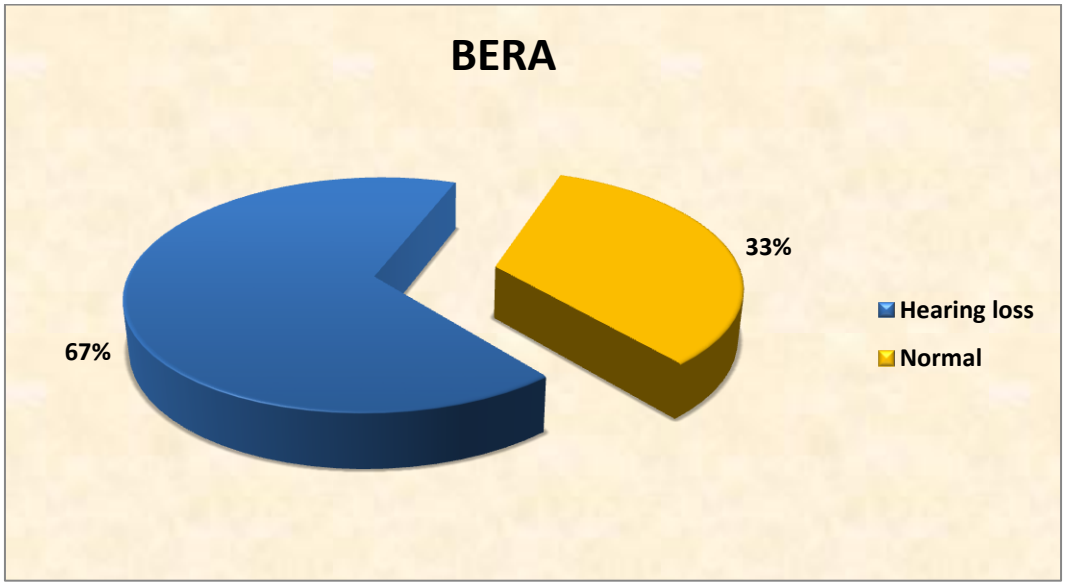

Table 5: Distribution of the cases according to first DPOAE screening and gender

It was seen that of 320 neonates screened, 178 were males and 142 were females.

Of the 281 neonates that passed the first DPOAE screen, 158 males and 123 females.

A total of 39 had failed the first test, 20 were males and 19 were females

On applying χ^2 (chi square) test, p value of 0.56 was obtained. Based on gender distribution, refer result among males and females are not significantly different. No significant association with gender and infant failing first DPOAE screening test was inferred.

Gender	Refer		B/L Pass		Total		χ^2 p value
	N	%	N	%	N	%	
Male	20	51.3%	158	56.2%	178	55.6%	
Female	19	48.7%	123	43.8%	142	44.4%	0.56
Total	39	100.0%	281	100.0%	320	100.0%	

Graph 4: Distribution of the cases according to first DPOAE screening and gender

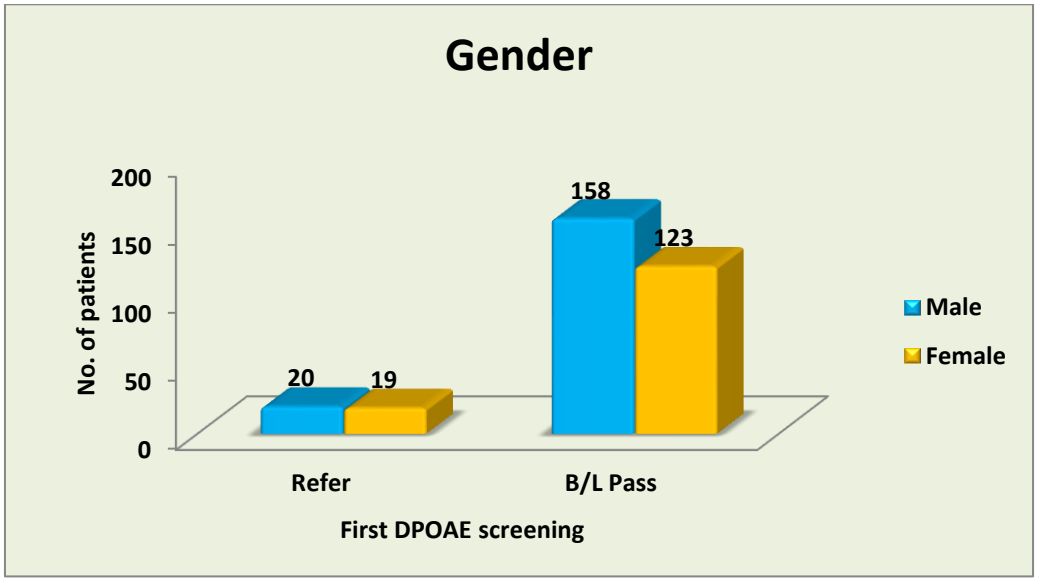

Table 6: Distribution of the cases according to second DPOAE screening and gender

The second DPOAE screen was conducted on 19 infants at 45-60 days, 12 males and 7 females were screened .A total of 1 male infant and 2 female infants had failed the second DPOAE screen.

Hence, on applying Fisher exact test, p value of 0.523 was obtained. Hence no significant difference in hearing loss in males and females was observed for the second OAE screen.

Gender	Refer		B/L Pass		Total		Fisher exact test p value
	N	%	N	%	N	%	
Male	1	33.3%	11	68.8%	12	63.2%	0.523
Female	2	66.7%	5	31.2%	7	36.8%	
Total	3	100.0%	16	100.0%	19	100.0%	

Graph 5: Distribution of the cases according to second DPOAE screening and gender

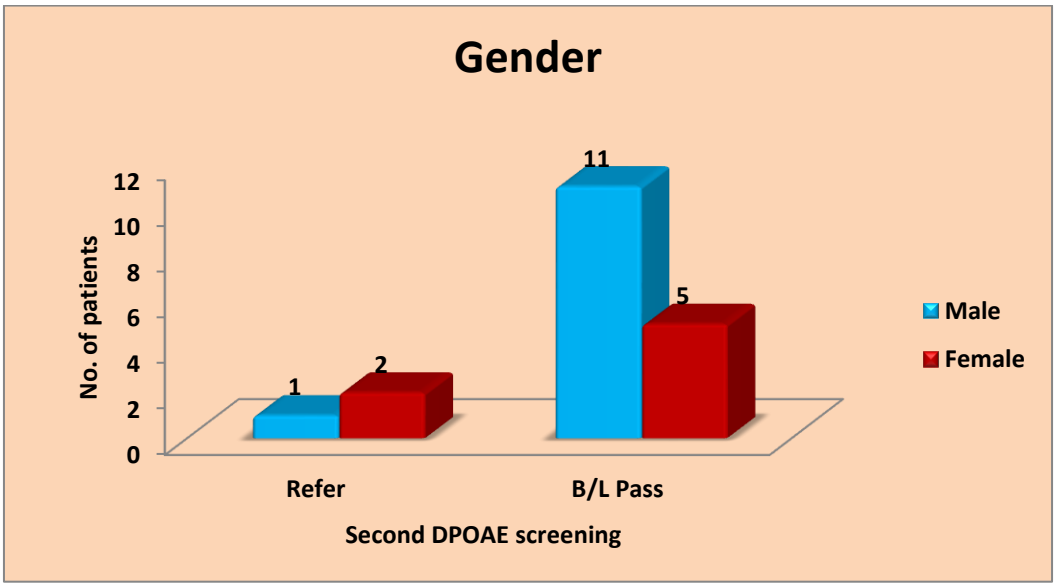

Table 7: Distribution of the cases according to BERA and gender

BERA was conducted on 3 infants who had failed the second DPOAE screen, at about 3 months of age of the infant .It was observed that 1 male and 1 female infant were diagnosed as having hearing loss.

On applying Fisher exact test, p value of 0.999 was obtained which was not significant. Based on gender distribution, almost same incidence of hearing loss in males and females was seen.

Gender	Hearing loss		Normal		Total		Fisher exact test p value
	N	%	N	%	N	%	
Male	1	50.0%	0	0.0%	1	33.3%	0.999
Female	1	50.0%	1	100.0%	2	66.7%	
Total	2	100.0%	1	100.0%	3	100.0%	

Graph 6: Distribution of the cases according to BERA and gender

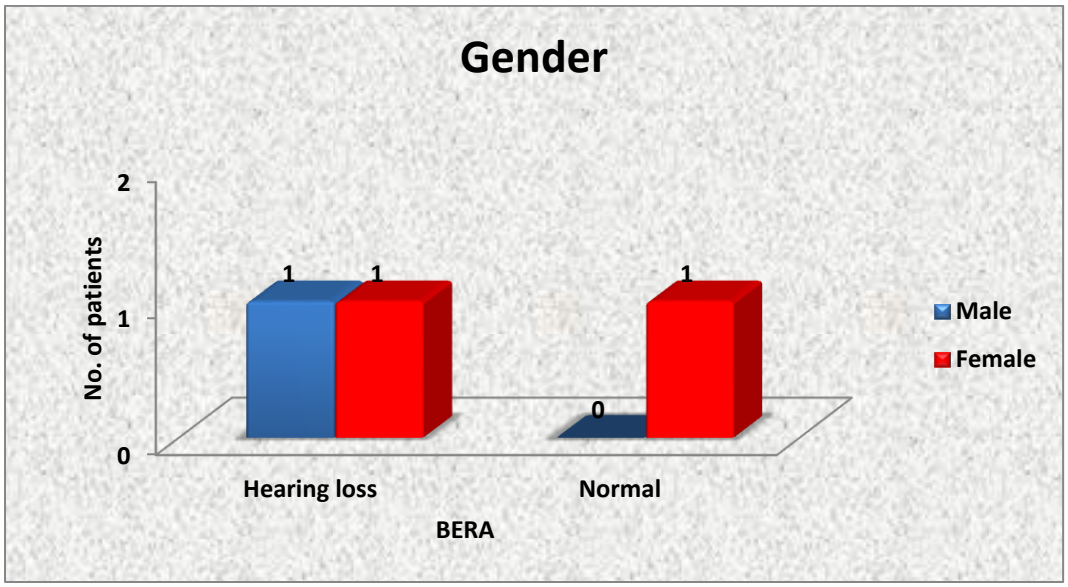

Table 8: Distribution of the cases according to First DPOAE screening and parity

Of the 320 neonates screened, a total of 201 mothers were multiparas, of which 176 neonates passed and 25 infants referred. Of the 119 infants of primipara, 105 passed and 14 referred. On applying χ^2 test, p value of 0.859 was obtained. No significant association with parity of the mother and infant failing screening test was seen.

Parity	Refer		B/L Pass		Total		χ^2 p value
	N	%	N	%	N	%	
Multi	25	64.1%	176	62.6%	201	62.8%	0.859
Primi	14	35.9%	105	37.4%	119	37.2%	
Total	39	100.0%	281	100.0%	320	100.0%	

Graph 7: Distribution of the cases according to first DPOAE screening and Parity

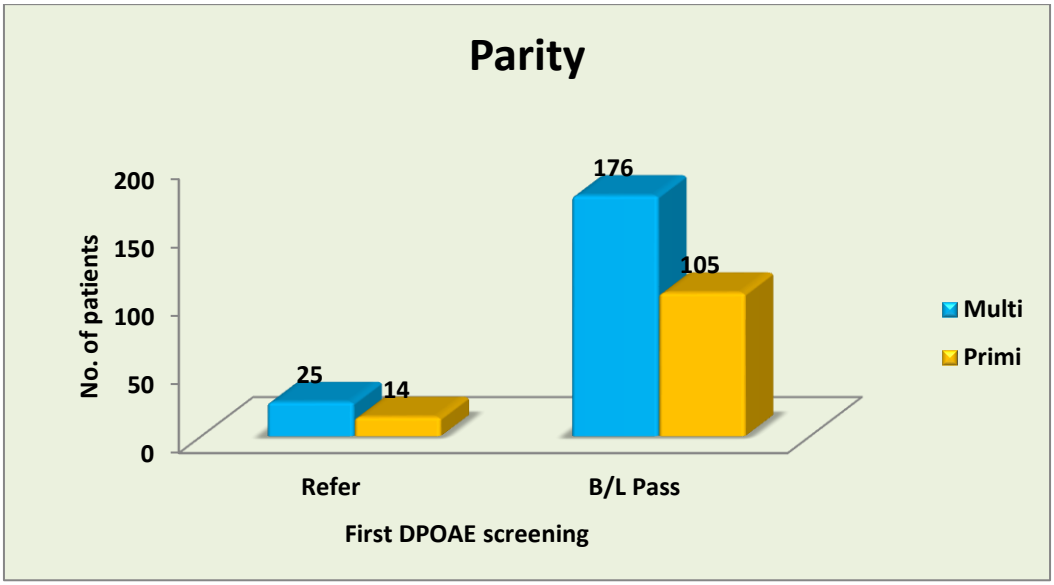

Table 9: Distribution of the cases according to second DPOAE screening and parity

Of the 19 infants screened with 2nd DPOAE, a total of 11 infants mothers were multipara and 8 primipara. On applying Fisher exact test, p value of 0.999 was obtained .No significant association with parity of the mother and infant failing second DPOAE screening test was inferred.

Parity	Refer		B/L Pass		Total		Fisher exact test p value
	N	%	N	%	N	%	
Multi	2	66.7%	9	56.2%	11	57.9%	
Primi	1	33.3%	7	43.8%	8	42.1%	0.999
Total	3	100.0%	16	100.0%	19	100.0%	

Graph 8: Distribution of the cases according to second DPOAE screening and parity

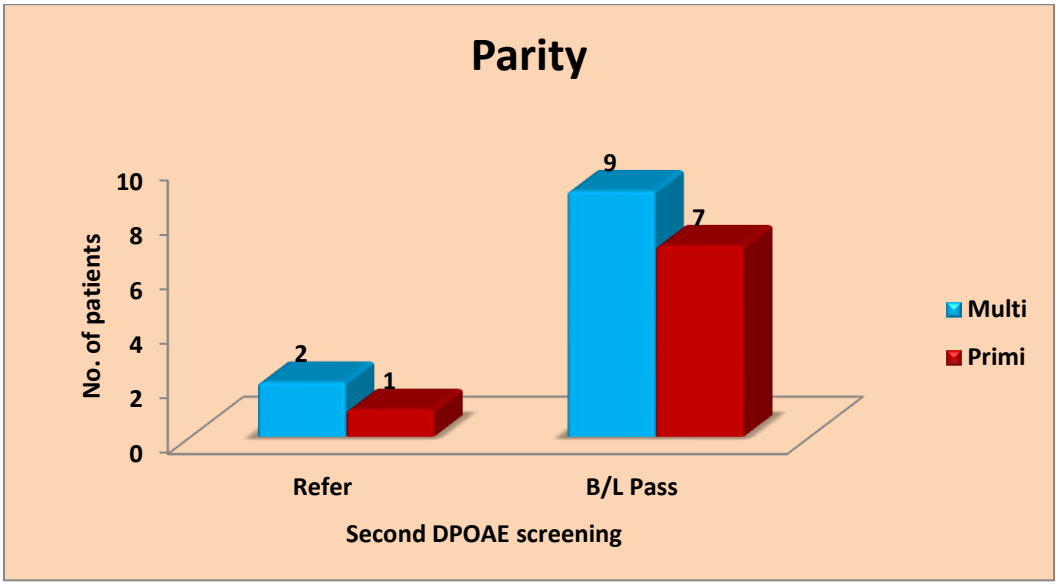

Table 10: Distribution of the cases according to BERA and parity

Of the 3 infants who underwent BERA, a total of 2 mothers were multipara and 1 primipara. On applying Fisher exact test, p value of 0.999 was obtained .No significant association with parity of the mother and hearing loss in infant was concluded.

Parity	Hearing loss		Normal		Total		Fisher exact test p value
	N	%	N	%	N	%	
Multi	1	50.0%	1	100.0%	2	66.7%	0.999
Primi	1	50.0%	0	0.0%	1	33.3%	
Total	2	100.0%	1	100.0%	3	100.0%	

Figure 9: Distribution of the cases according to BERA and parity

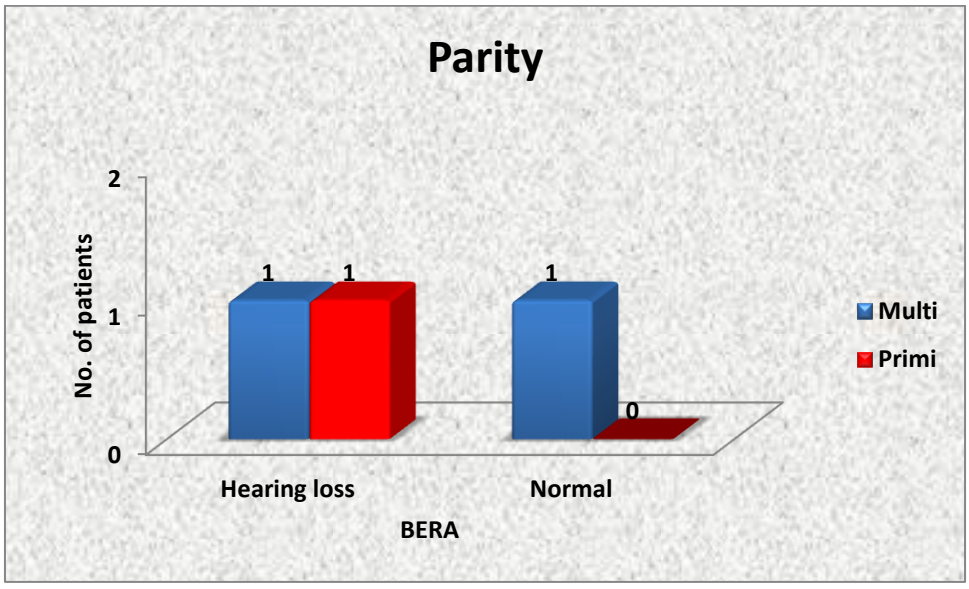

Table 11: Distribution of the cases according to first DPOAE screening and family history

Of the 320 neonates screened, a total of 289 had no relevant family history, of which 259 neonates passed and 30 neonates referred.30 of them had the history of consanguinity, of which 8 failed and 22 passed the first screening test.1 of them had the family history of deafness and the infant failed for the first test. On applying Fisher exact test, p value of 0.001 was obtained. So, the family history is significantly associated with referring at first DPOAE screening.

Family History	Refer		B/L Pass		Total		Fisher exact test p value
	N	%	N	%	N	%	
Absent	30	76.9%	259	92.2%	289	90.3%	0.001 (Sig)
Consanguinity	8	20.5%	22	7.8%	30	9.4%	
Deafness	1	2.6%	0	0.0%	1	0.3%	
Total	39	100.0%	281	100.0%	320	100.0%	

Graph 10: Distribution of the cases according to first DPOAE screening and family history

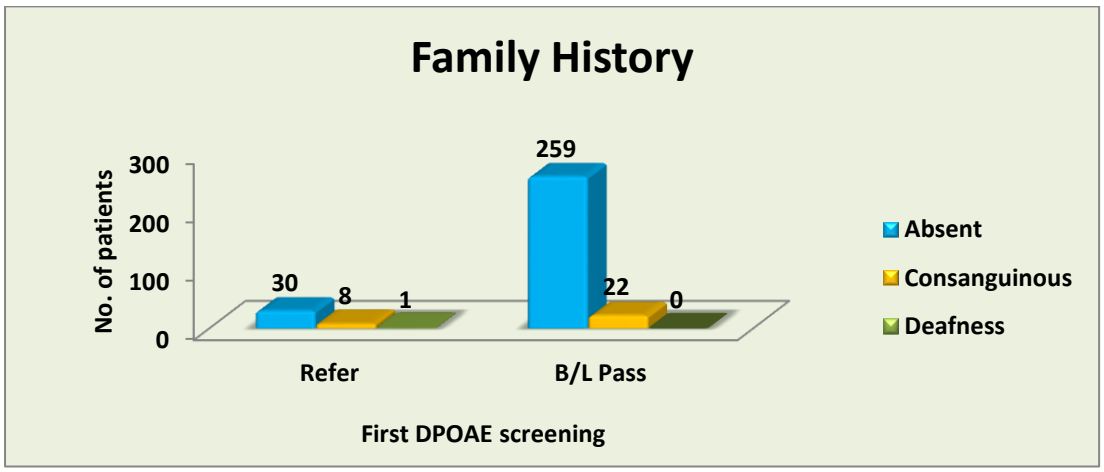

Table 12: Distribution of the cases according to second DPOAE screening and family history

Of the 19 infants screened with 2nd DPOAE, a total of 15 infants had no relevant family history of which 2 failed the test .4 of them had history of consanguinity of which 1 failed the test . On applying Fischer exact test, p value of 0.860 was obtained .No significant correlation with family history and infant failing the second DPOAE screening test was inferred

Family History	Refer		B/L Pass		Total		Fisher exact test p value
	N	%	N	%	N	%	
Absent	2	66.7%	13	81.2%	15	78.9%	0.860
Consanguinity	1	33.3%	3	18.8%	4	21.1%	
Deafness	0	0.0%	0	0.0%	0	0.0%	
Total	3	100.0%	16	100.0%	19	100.0%	

Graph 11: Distribution of the cases according to second DPOAE screening and family history

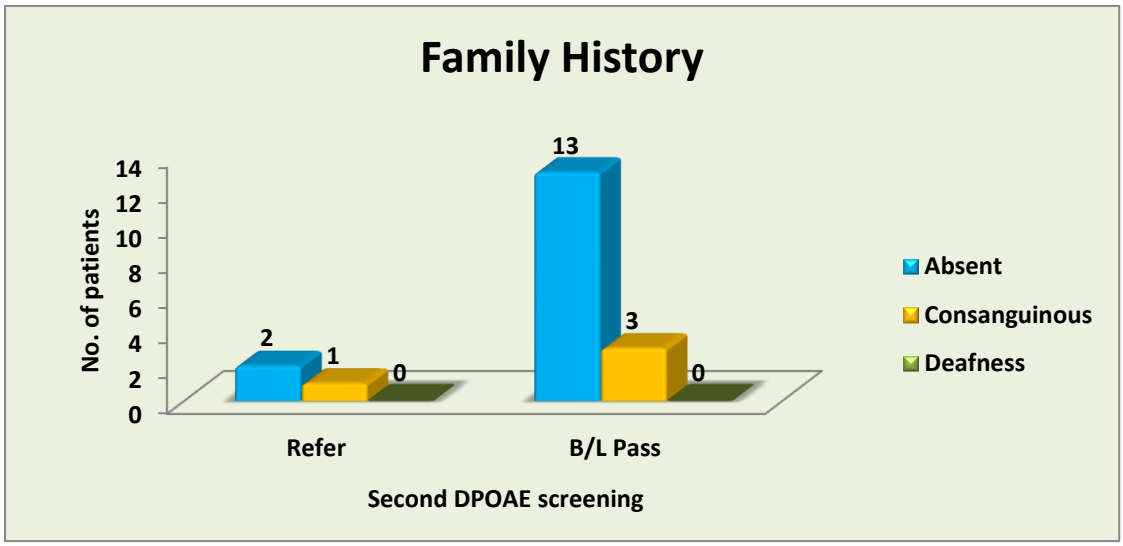

Table 13: Distribution of the cases according to BERA and family history

Of the 3 infants who underwent BERA, 2 failed the test and there was no relevant family history in both. On applying Fischer exact test, p value of 0.083. There was no significant correlation between family history and hearing loss.

Family History	Hearing loss		Normal		Total		p value
	N	%	N	%	N	%	
Absent	2	100.0%	0	0.0%	2	66.7%	0.083
Consanguinity	0	0.0%	1	100.0%	1	33.3%	
Total	2	100.0%	1	100.0%	3	100.0%	

Graph 12: Distribution of the cases according to BERA and family history

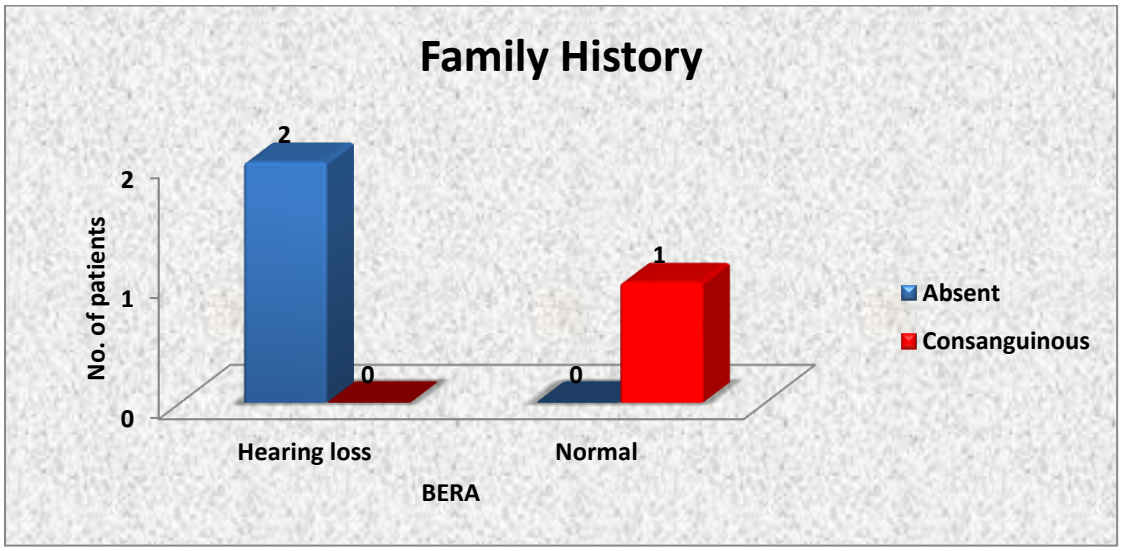

Table 14: Distribution of the cases according to first DPOAE screening and gestational age

It was seen that of 320 neonates screened, 39 neonates failed in the first DPOAE screening, of which 8 of them were late preterm, 4 of them were pre term and 27 were term and post term neonates. On applying χ^2 (chi square) test, p value of 0.026 was obtained. So the association of gestational age and first DPOAE screening is significant.

Gestational age	Refer		B/L Pass		Total		χ^2 p value
	N	%	N	%	N	%	
Late Preterm	8	20.5%	26	9.3%	34	10.6%	0.026 (sig)
Preterm	4	10.3%	13	4.6%	17	5.3%	
Term and post term	27	69.2%	242	86.1%	269	84.1%	
Total	39	100.0%	281	100.0%	320	100.0%	

Graph 13: Distribution of the cases according to first DPOAE screening and gestational age

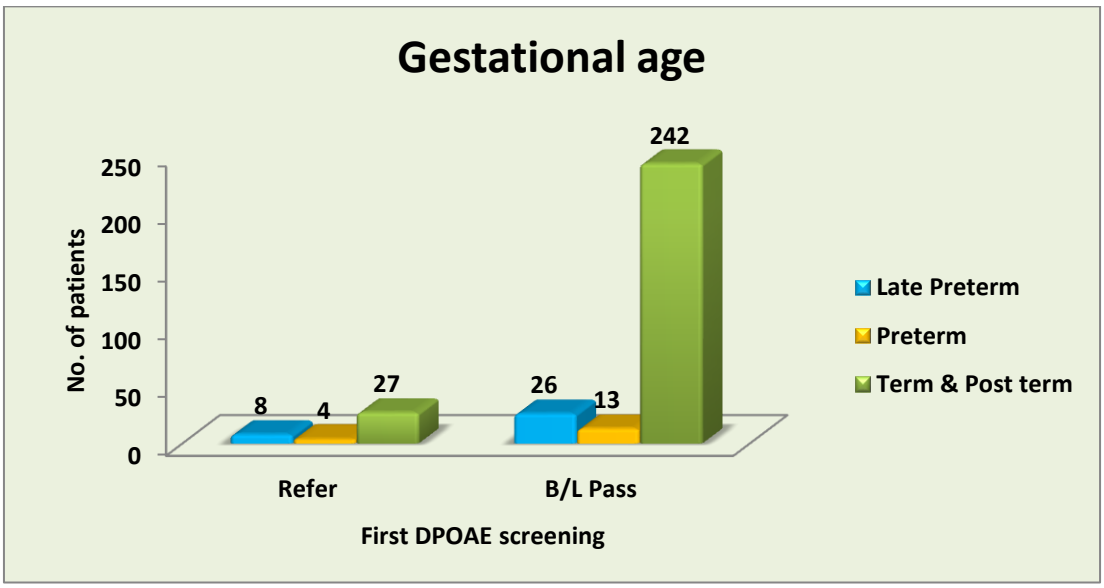

Table 15: Distribution of the cases according to second DPOAE screening and gestational age

Of the 19 infants who underwent the second DPOAE screening, 3 of them failed the test and all the 3 were term infants. On applying fisher exact test p, value of 0.659 was obtained which was not significant. There was no significant association between the gestational age and infant failing the second DPOAE screening.

Gestational age	Refer		B/L Pass		Total		Fisher exact test p value
	N	%	N	%	N	%	
Late Preterm	0	0.0%	6	37.5%	6	31.6%	0.659
Preterm	0	0.0%	2	12.5%	2	10.5%	
Term	3	100.0%	8	50.0%	11	57.9%	
Total	3	100.0%	16	100.0%	19	100.0%	

Graph 14: Distribution of the cases according to second DPOAE screening and gestational age

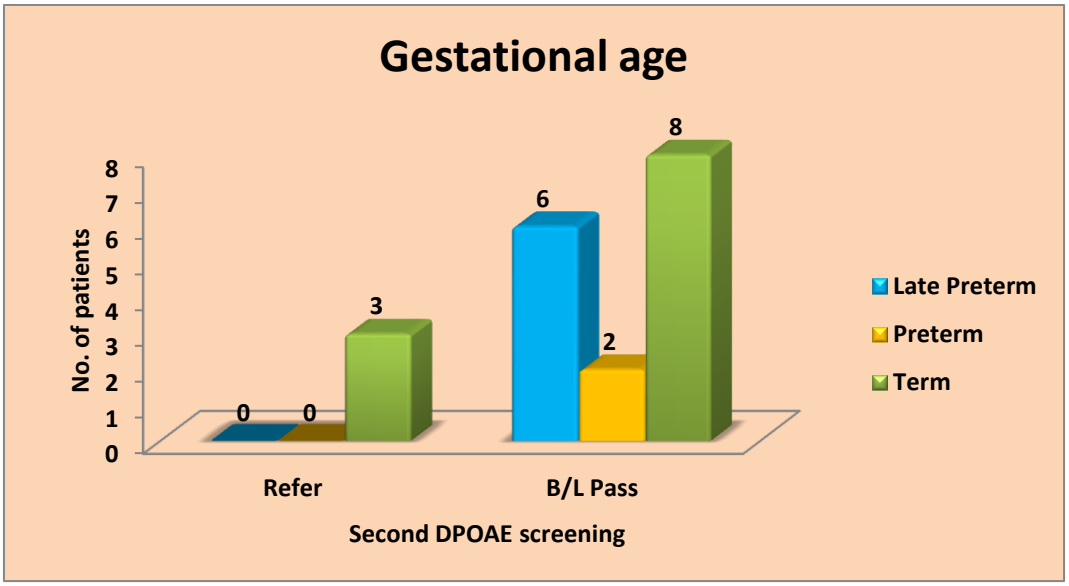

Table 16: Distribution of the cases according to BERA and gestational age

3 term infants who failed the second DPOAE screening underwent BERA. Two of them had hearing loss.

Gestational age	Hearing loss		Normal		Total		Fisher exact test p value
	N	%	N	%	N	%	
Late Preterm	0	0.0%	0	0.0%	0	0.0%	NA
Preterm	0	0.0%	0	0.0%	0	0.0%	
Term	2	100.0%	1	100.0%	3	100.0%	
Total	2	100.0%	1	100.0%	3	100.0%	

Graph 15: Distribution of the cases according to BERA and gestational age

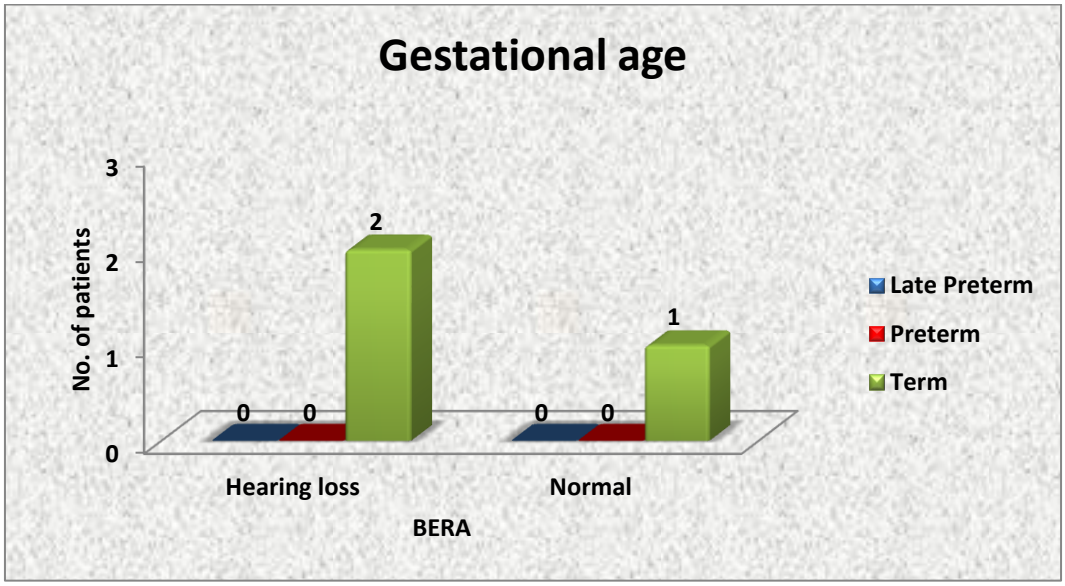

Table 17: Distribution of the cases according to first DPOAE screening and birth weight

It was seen that of 320 neonates screened, 36 neonates had birth weight of <2.5kg of which 31 had passed the first DPOAE screen, and 5 were refer. A total of 284 neonates had birth weight of ≥ 2.5kg, of which 250 had passed the first DPOAE screen, and 34 were refer for the 1st DPOAE screen.

On applying χ^2 (chi square) test, p value of 0.74 was obtained. Hence, no significant difference in hearing loss based on birth weight was seen on screening with 1st DPOAE test.

Birth weight	Refer		B/L Pass		Total		χ^2 p value
	N	%	N	%	N	%	
<2.5	5	12.8%	31	11.0%	36	11.2%	
≥2.5	34	87.2%	250	89.0%	284	88.8%	0.74
Total	39	100.0%	281	100.0%	320	100.0%	

Graph 16: Distribution of the cases according to first DPOAE screening and birth weight

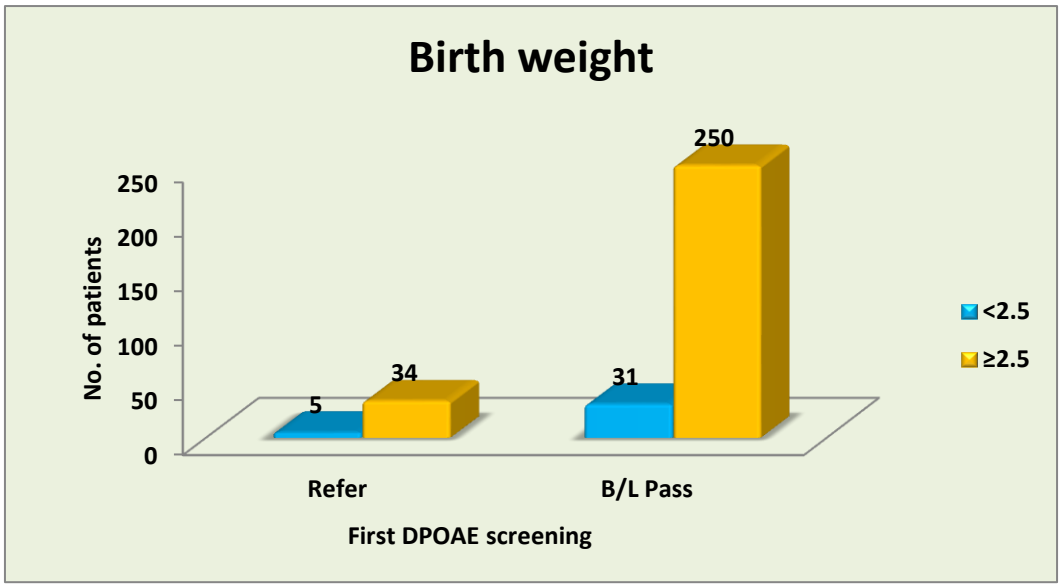

Table 18: Distribution of the cases according to second DPOAE screening and birth weight

It was seen that of 19 infants screened by 2nd DPOAE, 4 infants had birth weight of <2.5kg and all the 4 passed the 2nd DPOAE screening. A total of 15 infants had birth weight of ≥ 2.5kg of which 12 had passed the 2nd DPOAE screen, and 3 were refer for the 2nd screening test.

On applying fisher test, p value of 0.999 was obtained. Hence, no significant association of birth weight and a refer result in 2nd DPOAE screening test was inferred.

Birth weight	Refer		B/L Pass		Total		Fisher exact test p value
	N	%	N	%	N	%	
<2.5	0	0.0%	4	25.0%	4	21.1%	0.999
≥2.5	3	100.0%	12	75.0%	15	78.9%	
Total	3	100.0%	16	100.0%	19	100.0%	

Graph 17: Distribution of the cases according to second DPOAE screening and birth weight

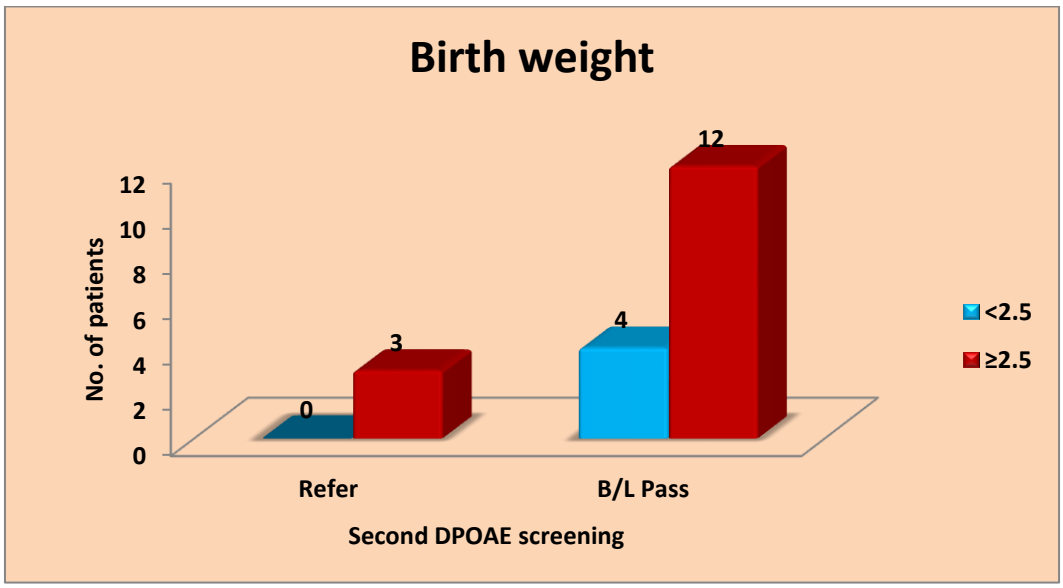

Table 19: Distribution of the cases according to BERA and birth weight

Of the 3 infants that underwent BERA, 3 infants who had Birth weight of ≥ 2.5kg, one had normal hearing and 2 had hearing loss.

Birth weight	Hearing loss		Normal		Total		Fisher exact test p value
	N	%	N	%	N	%	
<2.5	0	0.0%	0	0.0%	0	0.0%	NA
≥2.5	2	100.0%	1	100.0%	3	100.0%	
Total	2	100.0%	1	100.0%	3	100.0%	

Graph 18: Distribution of the cases according to BERA and birth weight

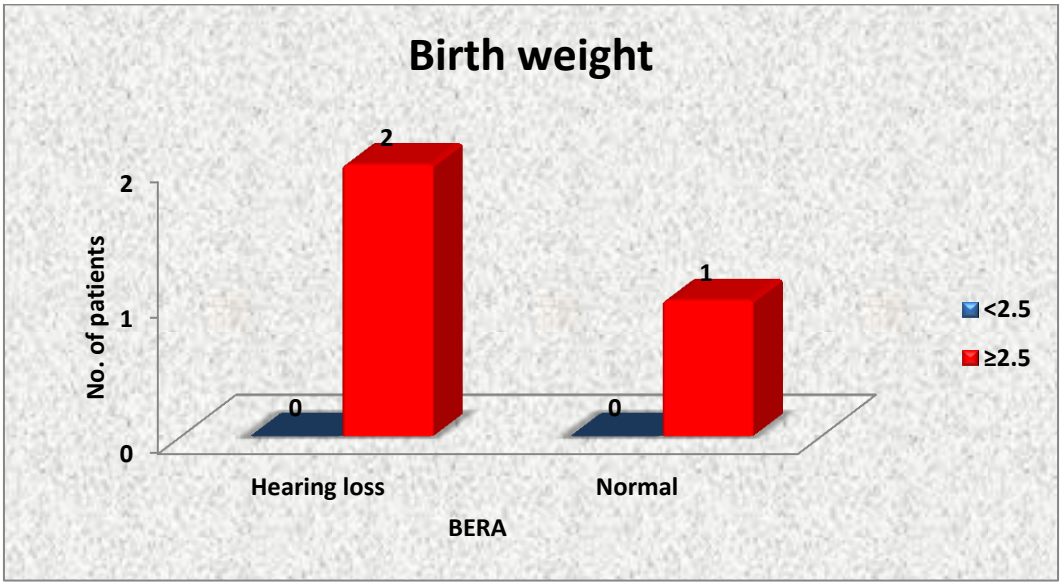

Table 20: Distribution of the cases according to first DPOAE screening and APGAR score.

It was seen that of 320 neonates screened, 9 infants with low APGAR score at 1 minute of birth, were refer. Neonates who had APGAR 7-10, of which 260 were pass and 30 refer. On applying chi square test, p value of 0.002 was obtained.

Hence, infants with an APGAR score more than 7 at 1 minute of birth are more likely to give pass results. There is significant association of APGAR score at 1 minute and first DPOAE screening result.

APGAR score at 1 minute	Refer		B/L Pass		Total		χ^2 p value
	N	%	N	%	N	%	
4-6	9	23.1%	21	7.5%	30	9.4%	0.002 (sig)
7-10	30	76.9%	260	92.5%	290	90.6%	
Total	39	100.0%	281	100.0%	320	100.0%	

Graph 19: Distribution of the cases according to first DPOAE screening and APGAR score.

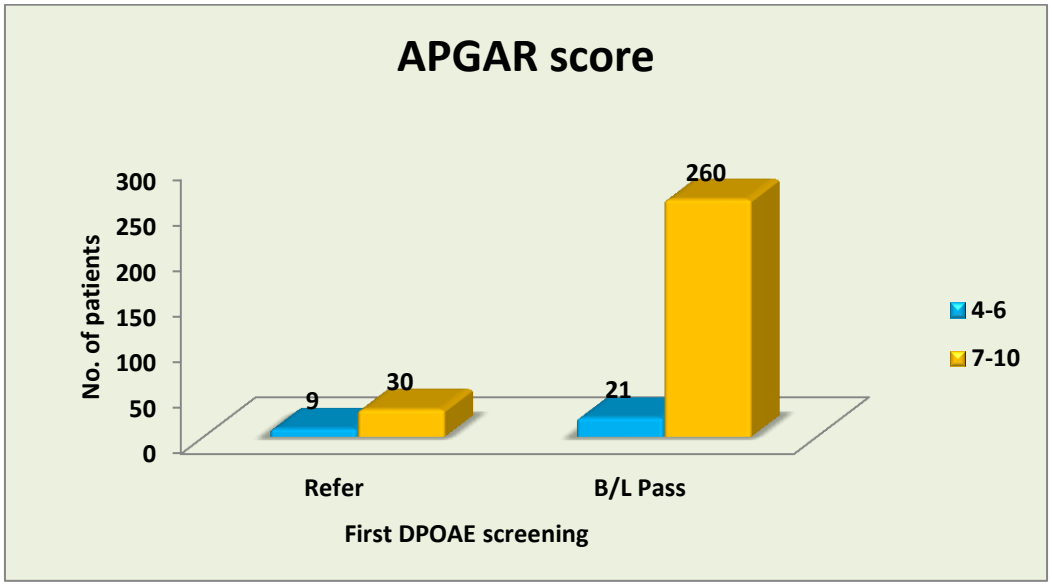

Table 21: Distribution of the cases according to second DPOAE screening and APGAR score.

It was seen that of 19 infants who underwent second DPOAE screening, 2 infants with low APGAR at 1 minute of birth were refer. Of infants with APGAR 7-10, 12 were pass and 1 refer.

On applying χ^2 tests, p value of 0.154 was obtained. Hence, no significant association of APGAR score at 1 minute was seen on screening with 2nd DPOAE screen.

APGAR score at 1 minute	Refer		B/L Pass		Total		χ^2 p value
	N	%	N	%	N	%	
4-6	2	66.7%	4	25.0%	6	31.6%	0.154
7-10	1	33.3%	12	75.0%	13	68.4%	
Total	3	100.0%	16	100.0%	19	100.0%	

Graph 20: Distribution of the cases according to second DPOAE screening and APGAR score

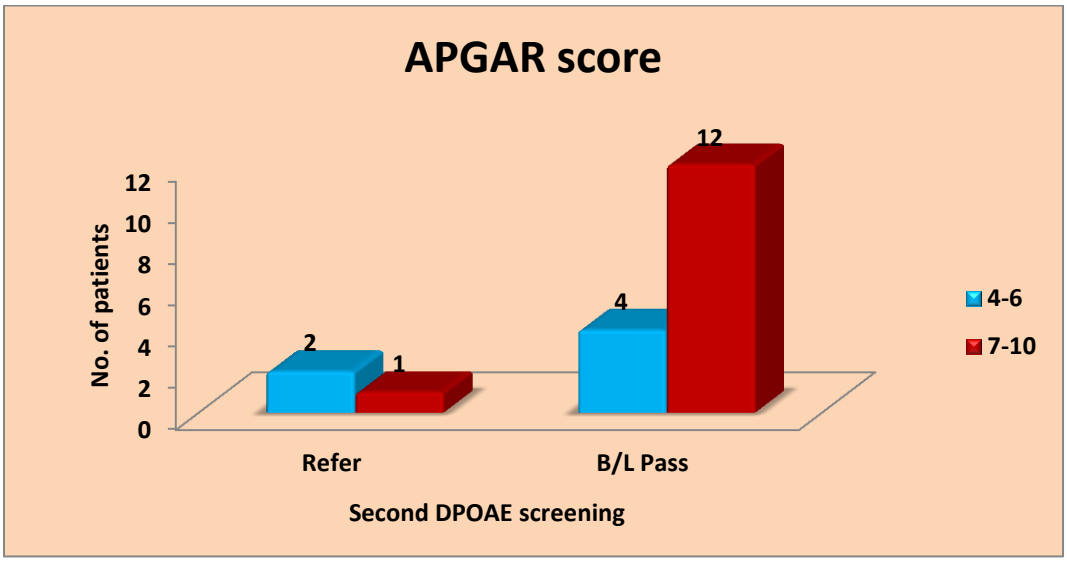

Table 22: Distribution of the cases according to BERA and APGAR score

It was seen that of 3 infants tested with BERA, 1 infant with low APGAR of score of 4-6 at 1 minute had hearing loss. Also 1 infant with APGAR 7-10, had hearing loss.

On applying Fischer exact test, p value of 0.999 was obtained. Hence, no significant association was observed between APGAR score and hearing loss.

APGAR score at 1 minute	Hearing loss		Normal		Total		Fisher exact test p value
	N	%	N	%	N	%	
4-6	1	50.0%	1	100.0%	2	66.7%	0.999
7-10	1	50.0%	0	0.0%	1	33.3%	
Total	2	100.0%	1	100.0%	3	100.0%	

Graph 21: Distribution of the cases according to BERA and APGAR score

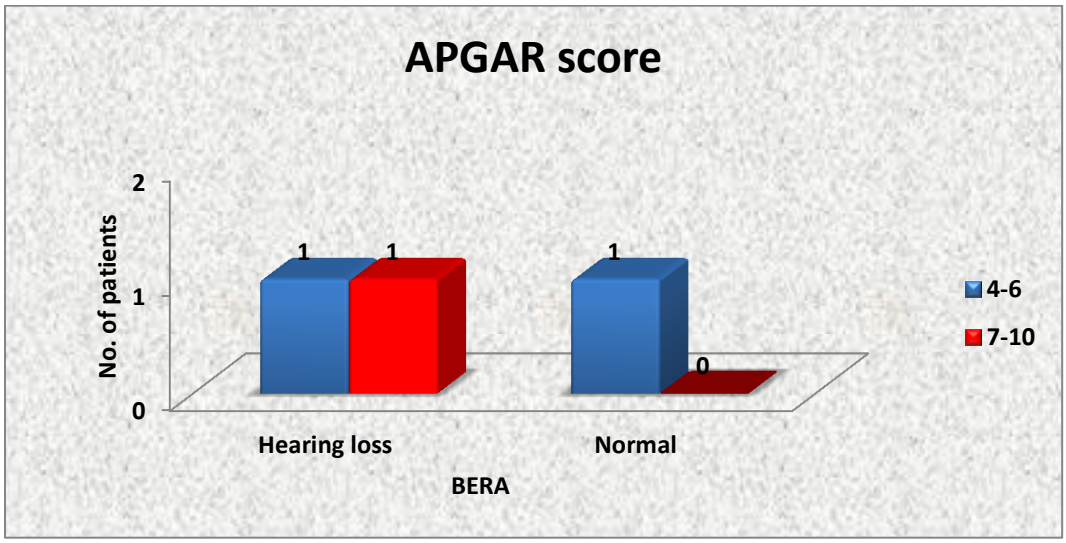

Table 23: Distribution of the cases according to first DPOAE screening and prenatal risk.

It was seen that of 320 neonates screened, 244 neonates had no prenatal risk factors. Of which, 212 passed and 32 failed 1st DPOAE screen. Of the 76 neonates that had prenatal risk factors, 69 passed and 7 failed first DPOAE screen.

On applying fisher exact test, p value of 0.364 was obtained. Hence, no significant association of prenatal risk factors and results of first DPOAE screening were observed in the study.

Prenatal risk	Refer		B/L Pass		Total		Fisher exact test p value
	N	%	N	%	N	%	
Absent	32	82.1%	212	75.4%	244	76.2%	0.364
Present	7	17.9%	69	24.6%	76	23.8%	
Total	39	100.0%	281	100.0%	320	100.0%	

Graph 22: Distribution of the cases according to first DPOAE screening and prenatal risk

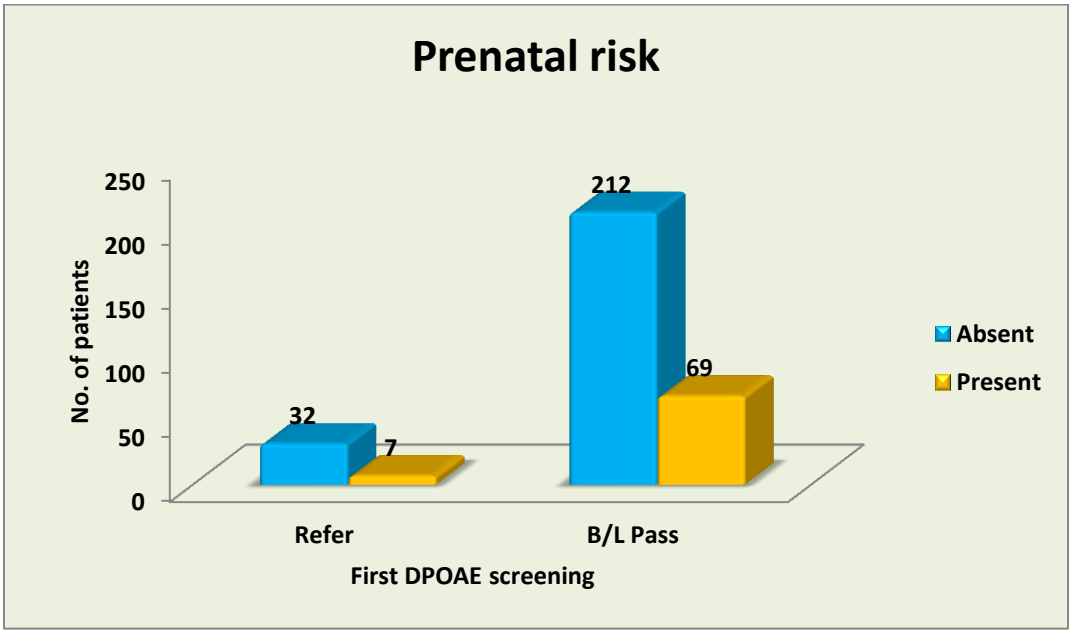

Table 24: Distribution of the cases according to second DPOAE screening and prenatal risk

Of the 19 infants that underwent 2nd DPOAE screen, 13 infants had no prenatal risk factors, of which 11 passed and 2 failed. A total of 6 infants had risk factors prenatally, of which 5 passed and 1 failed 2nd DPOAE screen.

On applying fisher exact test, p value of 0.096 was obtained. Hence, no significant association of prenatal risk factors was seen on screening with 2nd DPOAE screen.

Prenatal risk	Refer		B/L Pass		Total		Fisher exact test p value
	N	%	N	%	N	%	
Absent	2	66.7%	11	68.8%	13	68.4%	0.096
Present	1	33.3%	5	31.2%	6	31.6%	
Total	3	100.0%	16	100.0%	19	100.0%	

Graph 23: Distribution of the cases according to Second DPOAE screening and prenatal risk

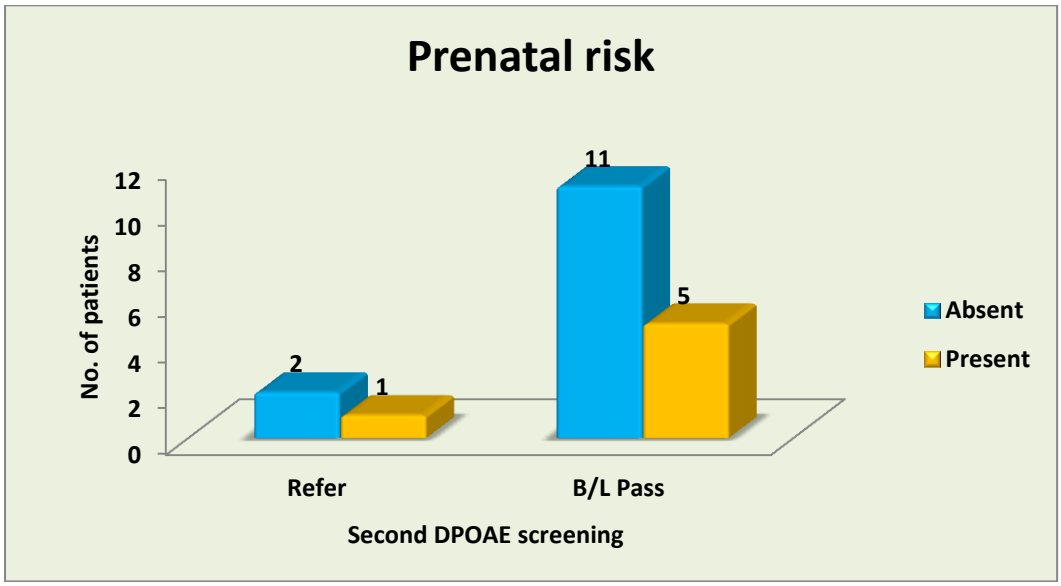

Table 25: Distribution of the cases according to BERA and Prenatal risk

During the BERA test, 1 of the 2 infants who had no prenatal risk factors passed and 1 failed. One infants who had risk factors failed BERA.

On applying Fischer exact test, p value of 0.386 was obtained. Hence no significant difference in hearing loss in groups with and without prenatal risk factors was observed.

Prenatal risk	Hearing loss		Normal		Total		Fisher exact test p value
	N	%	N	%	N	%	
Absent	1	50.0%	1	100.0%	2	66.7%	0.386
Present	1	50.0%	0	0.0%	1	33.3%	
Total	2	100.0%	1	100.0%	3	100.0%	

Graph 24: Distribution of the cases according to BERA and Prenatal risk

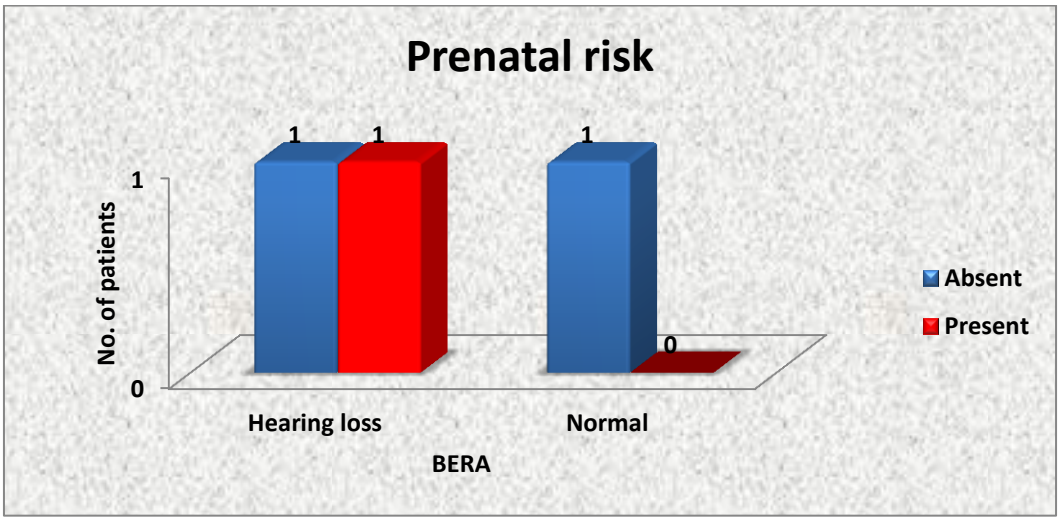

Table 26: Distribution of prenatal risks in first DPOAE screening

32 out of 244 neonates who failed the first DPOAE screening were having no prenatal risks. 7 out of 24 neonates with prenatal risk failed the first DPOAE screening.

Prenatal risks	**N**	**%**	**Result of screening**	**N**	**%**
Absent	244	76.2	Refer	32	82.1
Fetal distress	19	5.9		4	10.3
PPROM, Gestational hypertension	1	0.3		1	2.6
Twin	3	0.9		1	2.6
Twin, PPROM	1	0.3		1	2.6
Anaemia	2	0.6	B/L Pass		
Eclampsia	2	0.6			
Eclampsia, anaemia	2	0.6			
Fetal distress, MSL	1	0.3			
Forceps Delivery	1	0.3			
GDM	1	0.3			
Gestational hypertension	9	2.8			
IUGR	2	0.6			
MSL	6	1.9			
Oligohydraminos	11	3.4			
PPROM	3	0.9			
Pre eclampsia	3	0.9			
Prolonged labour	1	0.3			
PROM	5	1.6			
PROM with IUGR	1	0.3			
PROM, oligohydraminos	1	0.3			
vaccum delivery	1	0.3			
Total at first screening	320	100	Total referred	39	

Graph 25: Distribution of prenatal risks in first DPOAE screening

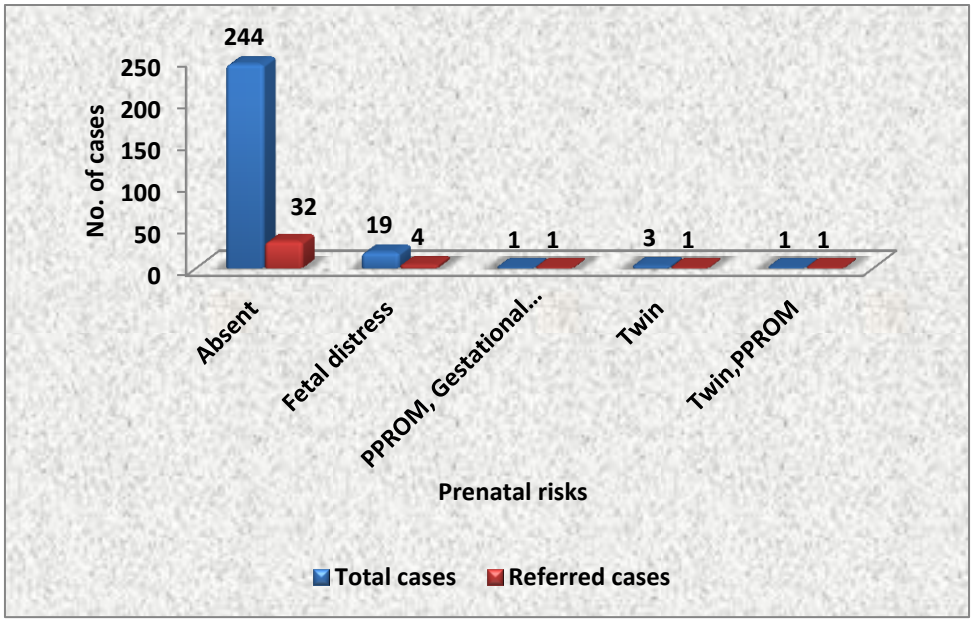

Table 27: Prenatal risks in referred cases of first DPOAE screening

The major pre natal complications which associated with a "refer" in first DPOAE screening were fetal distress, multiple pregnancy, PPROM, gestational hypertension.

Prenatal risk	**Referred cases (N=39)**	**%**
Fetal distress	4	10.3%
Twin	2	5.1%
PPROM	1	2.6%
Gestational hypertension	1	2.6%

Table 28: Distribution of the cases according to first DPOAE screening and postnatal risk

It was seen that of 320 neonates screened, 240 neonates had no postnatal risks. Of these, 212 of these infants passed the first DPOAE screen and 28 failed the first test. Eighty of the screened infants had postnatal risks, of which 69 of these infants passed the first DPOAE screen and a total of 11 failed the first test On applying χ^2 (chi square) test, p value of 0.622 was obtained. Hence, there is no significant association of postnatal risks and results of first DPOAE screening.

Postnatal risks	Refer		B/L Pass		Total		χ^2 p value
	N	%	N	%	N	%	
Absent	28	71.8%	212	75.4%	240	75.0%	0.622
Present	11	28.2%	69	24.6%	80	25.0%	
Total	39	100.0%	281	100.0%	320	100.0%	

Graph 26: Distribution of the cases according to first DPOAE screening and postnatal risk

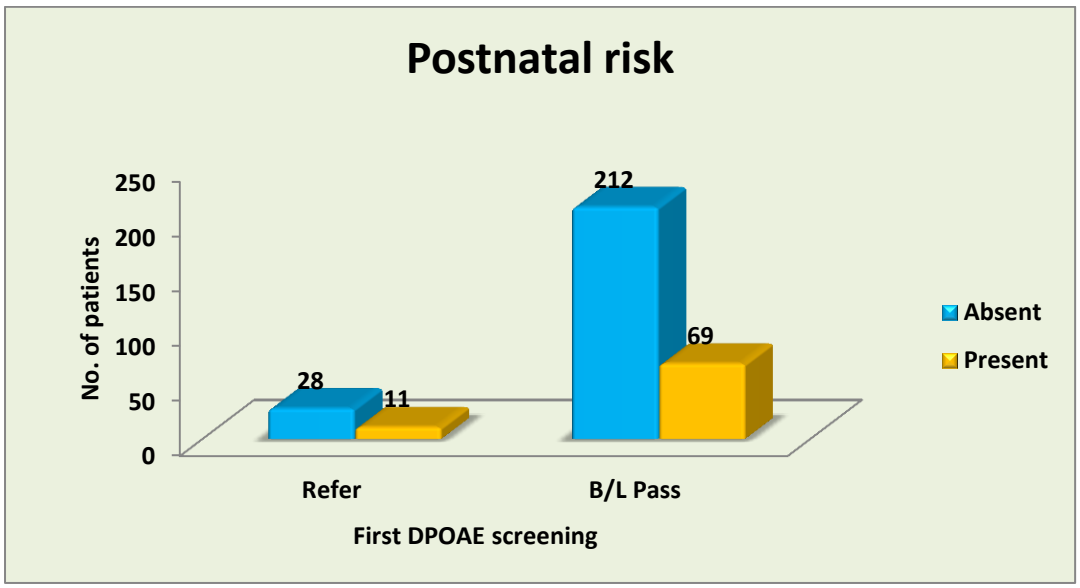

Table 29: Distribution of the cases according to second DPOAE screening and postnatal risk

When the 19 infants were screened with 2nd DPOAE, 12 infants had no postnatal risks, 11 of these infants passed the second DPOAE screen and 1 failed the test. Of the infants screened, 7 had postnatal risks, of which 5 of these infants passed the second DPOAE screen and a total of 2 failed the second test. On applying fisher exact test, p value of 0.523 was obtained. Hence, there is no significant association of postnatal complication and results of second DPOAE screening.

Postnatal risks	Refer		B/L Pass		Total		Fisher exact test p value
	N	%	N	%	N	%	
Absent	1	33.3%	11	68.8%	12	63.2%	0.523
Present	2	66.7%	5	31.2%	7	36.8%	
Total	3	100.0%	16	100.0%	19	100.0%	

Graph 27: Distribution of the cases according to second DPOAE screening and postnatal risk

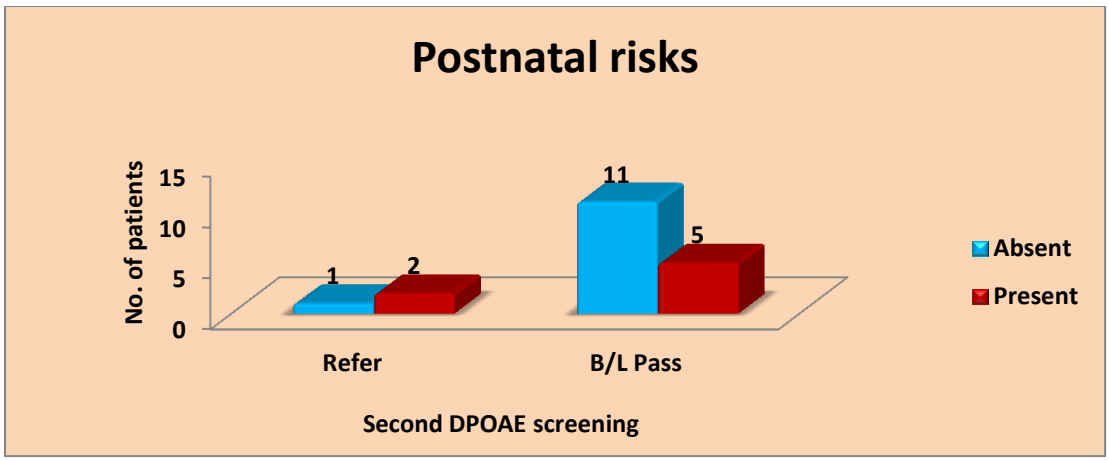

Table 30: Distribution of the cases according to BERA and Postnatal risk

Of the 3 infants who underwent BERA, 2 infants had postnatal risks. Of these 1 infant had normal hearing and 1 had hearing loss. Of the screened infants, 1 had no postnatal risk, and had normal hearing on testing with BERA. On applying Fischer exact test, p value of 0.999 was obtained. Hence, no significant difference in hearing loss between the two groups was observed in the study.

Postnatal risks	Hearing loss		Normal		Total		Fisher exact test p value
	N	%	N	%	N	%	
Absent	1	50.0%	0	0.0%	1	33.3%	0.999
Present	1	50.0%	1	100.0%	2	66.7%	
Total	2	100.0%	1	100.0%	3	100.0%	

Graph 28: Distribution of the cases according to BERA screening and postnatal risk

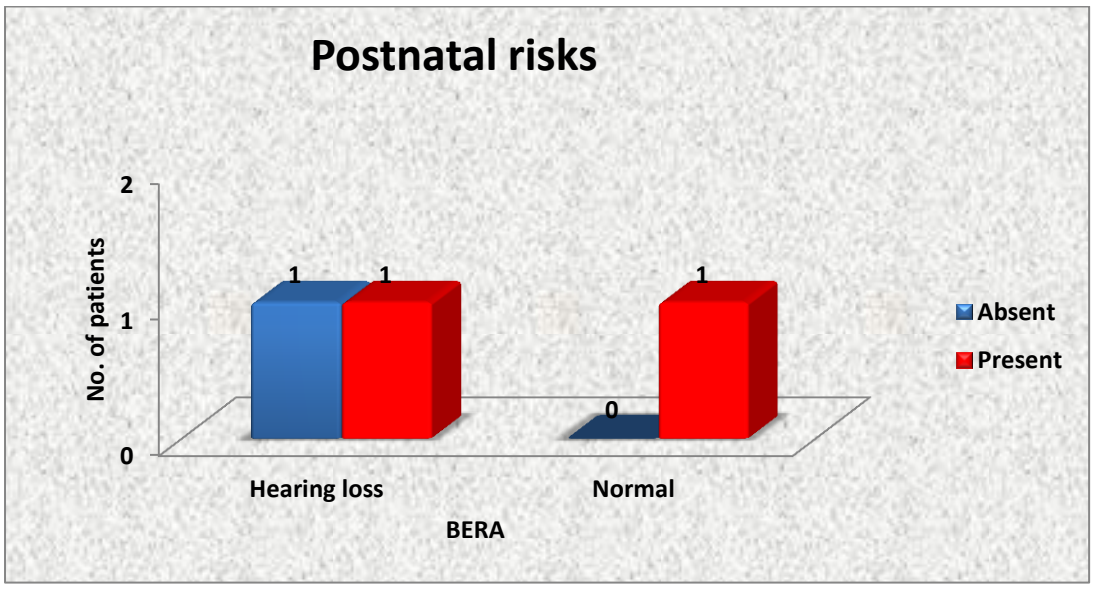

Table 31: Distribution of postnatal risks in first DPOAE screening.

Of the 320 cases, 240 cases had no postnatal complications. 80 cases had postnatal complications. 28 cases of 240 with no postnatal risks failed the first screening. 11 out of 80 cases who had post natal risk factors failed the first DPAOE screening.

Postnatal risks	**N**	**%**	**Result of screening**	**N**	**%**
Absent	240	75.0%	Refer	28	71.8%
LBW	41	12.8%		2	5.1%
CP, Seizures	1	0.3%		1	2.6%
NICU, Amikacin	1	0.3%		1	2.6%
NICU, HIE, Amikacin	4	1.3%		1	2.6%
NICU, HIE, Amikacin, CP	1	0.3%		1	2.6%
NICU, HIE, MV, Amikacin	2	0.6%		1	2.6%
NICU, HIE, Sepsis, Amikacin	1	0.3%		1	2.6%
NICU, LBW, Amikacin	2	0.6%		1	2.6%
NICU, RDS, MV, VLBW, Amikacin	1	0.3%		1	2.6%
NICU, VLBW, Amikacin	2	0.6%		1	2.6%
Cleft palate	1	0.3%	B/L Pass		
NICU, HDN, LBW, Amikacin	1	0.3%			
NICU, HIE, LBW	1	0.3%			
NICU, HIE, MAS, Amikacin	1	0.3%			
NICU, HIE, NHB, LBW, Amikacin	1	0.3%			
NICU, HIE, RDS, LBW, Amikacin	1	0.3%			
NICU, LBW, RDS, Amikacin	2	0.6%			
NICU, LBW, Sepsis, Amikacin	1	0.3%			
NICU, MAS, Amikacin	2	0.6%			
NICU, MAS, RDS, Amikacin	1	0.3%			
NICU, NHB	3	0.9%			
NICU, NHB, Amikacin	1	0.3%			
NICU, NHB, ET, Amikacin	1	0.3%			
NICU, RDS	1	0.3%			
NICU, RDS, Amikacin	1	0.3%			
NICU, Sepsis, LBW, Amikacin	1	0.3%			
NICU, TTN	1	0.3%			
NICU, TTN, Amikacin	1	0.3%			
NICU,RDS, Sepsis,Seizures,VLBW, MV, Amikacin	1	0.3%			
Neonatal anaemia,Blood transfusion	1	0.3%			
Total	320	100.0%	Total referred		39

Graph 29: Distribution of postnatal risks in first DPOAE screening

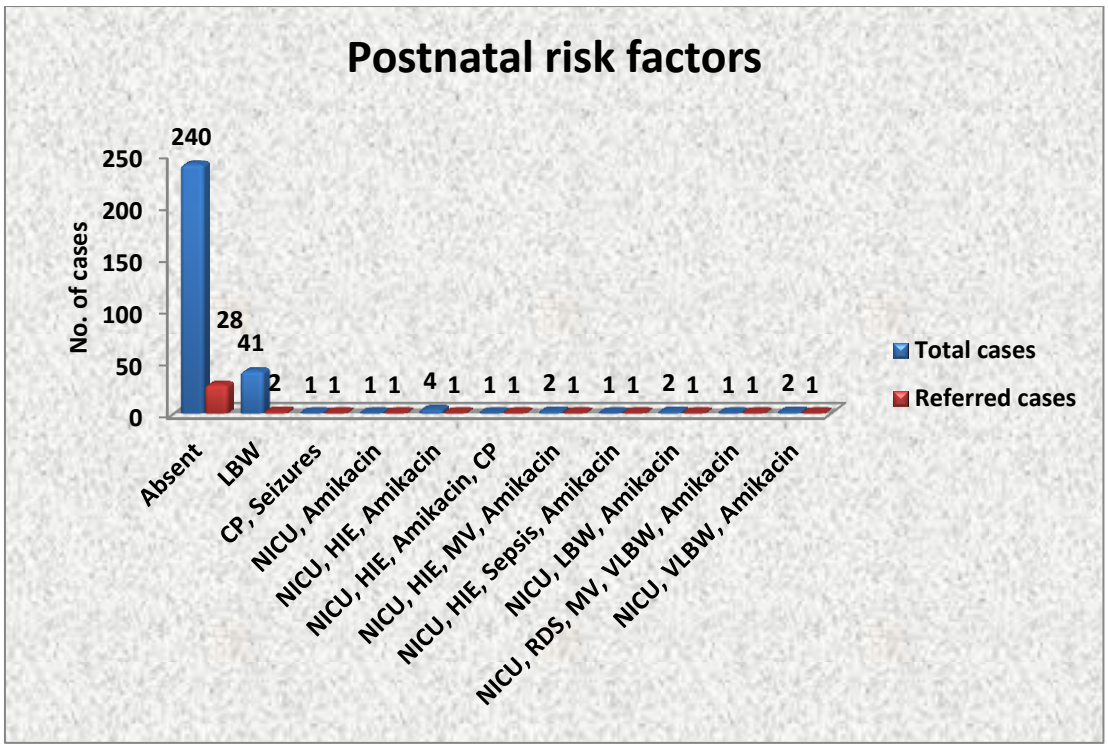

Table 32: Post natal risks in referred cases of first DPOAE Screening

The major post natal risks which associated with a "refer" in first DPOAE screening were, NICU admission, aminoglycoside like amikacin administration, Hypoxic Ischemic Encephalopathy and low birth weight .

Postnatal risks	Referred cases (N=39)	%
NICU	8	20.5%
Amikacin	8	20.5%
HIE	4	10.3%
LBW	3	7.7%
VLBW	2	5.1%
CP	2	5.1%
MV	2	5.1%
Sepsis	1	2.6%
RDS	1	2.6%

Table 33: Efficacy of first DPOAE screening

At first screening (excluding 20 lost to follow-up) 19 babies had a refer result in first DPOAE.281 babies had a pass result. Thus, sensitivity of the first DPOAE screening was 100 % and specificity was 94.3 %.Positive predictive value was 10.5% and negative predictive value was 100%. The accuracy of the test was 94.3%.

Table 33(a)

At first screening (excluding 20 lost to follow up)			
	Hearing Loss	**Normal**	**Total**
Referred	2	17	19
Passed	0	281	281
Total	2	298	300

Table 33 (b)

Sensitivity	100.0%
Specificity	94.3%
PPV	10.5%
NPV	100.0%
Accuracy	94.3%

Table 34: Efficacy of second DPOAE screening

Of the 19 infants who underwent second DPOAE screening, 3 failed the screening test and 16 passed the test. Thus, sensitivity of the first DPOAE screening was 100 % and specificity was 94.1 %.Positive predictive value was 66.7% and negative predictive value was 100%. The accuracy of the test was 94.7%.

Table 34(a)

At Second screening			
	Hearing Loss	**Normal**	**Total**
Referred	2	1	3
Passed	0	16	16
Total	2	17	19

Table 34(b)

Sensitivity	100.0%
Specificity	94.1%
PPV	66.7%
NPV	100.0%
Accuracy	94.7%

DISCUSSION

Congenital hearing loss is one of the most common congenital anomalies which can be identified early in life. The early recognition and intervention helps in the overall development of the child. There is a clear consensus that hearing screening and intervention at an early age improves later speech and language development outcomes.[68]

The developed countries are aware of the burden of congenital hearing loss and have taken significant steps by way of government policies for identification and rehabilitation. Children with disability in developing countries are more likely to face discrimination, restricted access to social services, be malnourished, and face physical abuse.[69]

WHO estimates that globally the number of people with hearing loss, has more than doubled from 120 million in 1995 to at least 278 million in 2005, thus making this condition the most prevalent sensory deficit in the population. In India, it is estimated that 18.49 million persons have disability that equivalents to 1.8 percent of the total population of the country where 10 percent of this figure are likely to have hearing disability of moderate to profound degree. Moreover, this number is likely to go up if we add lower degree of hearing disability.[70]

The adverse affects of hearing loss on language and cognitive development, as well as on psychosocial behavior are widely reported against the established benefits of early intervention. Children with a disabling hearing loss are at risk of delayed speech and language development with consequent poor academic performance.[71] The income of individuals with hearing loss is reported to be 40 to 45% less than the hearing population in developed countries and will be even more pronounced in developing countries.

The definition of early identification and intervention has evolved over the years. In the past, early identification was defined as intervention before the age of 18 months. However, now early identification is defined as the diagnosis as early as 3 months with intervention by 6 months.

Screening for hearing loss in infants should be done with a screening test that is simple, cost effective, quick, sensitive, efficient, reliable and effective. 50% of children with moderate to profound congenital hearing loss exhibit no risk factors for hearing loss.[71] In the absence of such objective screening test, hearing loss may not be detected until the child is 2–6 years of age, when intervention outcomes may be suboptimal.

In the present study conducted, a total of 320 neonates selected at random were screened with DPOAE at 48-96 hours of birth. Infants who had failed the first DPOAE screen were subjected to a second DPOAE screening at 45-60 days of life. For the infants that had failed second DPOAE screen were subjected to BERA at about three months age, to confirm for presence of hearing loss. The study was conducted at BLDE U's Shri B. M. Patil Medical College Hospital and Research Centre, Vijayapur, from October 2014 to August 2016. A total of 320 neonates were screened by DPOAE of which 39 failed the initial screen (12.18%). 19 of 39 infants who failed the DPOAE were rescreened (20 infants failed to follow up) .On rescreening, 3 infants failed in the second DPOAE screening. BERA was done for the 3 infants, of which 2 were found to have severe sensorineural hearing loss.

1) PREVALANCE:

In this study, the prevalence of sensorineural hearing loss was found to be 6.67 per thousand infants. According to the WHO estimates, globally up to 6 per 1000 live-

born infants annually, or 7,98,000 babies worldwide, suffer permanent hearing loss at birth or within the neonatal period. At least 90% of them are in developing countries.[56]

As per WHO estimates in India, there are approximately 63 million people, who are suffering from significant auditory impairment; this places the estimated prevalence at 6.3% in Indian population[64]

The Rhode Island hearing assessment program revealed 111 infants with permanent hearing loss, resulting in an impairment rate of 2 per 1000.[54] The Colorado Study showed a prevalence of 2/1000 population.[72] In 2013, a study done at Christian Medical College Vellore, showed that the prevalence of hearing loss among neonates was 4.1 per 1000 babies screened.[73] In the study conducted in St. Johns Medical College Hospital, Bangalore the weighted incidence of standardized population of neonates seeking care in a tertiary centre in India is 5.65 per 1000.[74]

2) GENDER:

We observed that of 320 neonates screened, 178 were males and 142 were females. Of the neonates that had passed the first DPOAE screen, 158 males and 123 females were present. A total of 39 had failed the first test, of which 20 were males and 19 were females. On screening with second DPOAE screen at 45-60 days, 11 male and 5 female infants passed the test respectively. It was observed that 1 male infant and 2 female infants had failed the second DPOAE screen. BERA was conducted on the 3 infants who had failed the second DPOAE at about 3 months of age of the infant. It was observed that 1 male and 1 female infant was diagnosed as having hearing loss. Hence, based on gender distribution, almost same incidence of hearing loss in males and females was seen. This was similar to the conclusion from the research by Vanessa Sinelli and researchers at Clinical Audiology Section of the

Communication Disorders Education and Rehabilitation Division. A total of 138 infants were evaluated from May to December 2004, 70 males and 68 females were evaluated, with ages between 6 to 65 days. They obtained $p>0.05$ in all frequencies, concluding, that there is no gender effect in the response level.[75]

However, in a study by Saitoh et al, 352 neonates were screened and it was found that there was a significant effect of gender on the signal-noise ratio, response level, whole wave and band reproducibility values. The females had a higher value in these aspects.[76]

3) LATERALITY:

With relation to the side of ear tested, according to the study of Raineri [77] and colleagues in 2001, there was no statistical difference between right and left ears for the DPOAE response level. In the study by Saitoh et al, it was seen that right ear had higher values of signal-noise ratio, response level, whole wave and band reproducibility values.[76]

In our study, on testing with the first DPOAE screen, number of infants that failed only left or only right ear was at an equal rate. Similarly, during the second DPOAE screen conducted on 19 infants, 3 infants had bilateral refer. Hence, there is no significant difference in incidence of hearing loss between right and left ear.

4) PARITY :

In our study, during the first DPOAE screen, 201 mothers were multipara and 119 were primipara. On applying statistical evaluation, no significant difference was seen in incidence of hearing loss in infants of mulipara and primipara. On repeat DPOAE screening and on conducting BERA, similar conclusion was arrived at. This is in concordance with Chu and colleagues in 2003, who concluded that there were no differences between groups when compared for maternal age, parity, race, and

exposure to prescribed or illicit drugs, clinical or histologic chorioamnionitis, and various perinatal outcome variables.[78]

5) FAMILY HISTORY :

It was observed a statistically significant relation between absence of Otoacoustic Emissions and family history of deafness and consanguinity in the first DPOAE screening. In the second DPOAE screening and BERA such a correlation could not be established. However, JCIH position statement consider family history of permanent hearing loss to be associated with neonatal hearing loss.

6) PRETERM AND TERM INFANTS:

It was observed a statistically significant relation between absence of Otoacoustic Emissions and preterm neonates in the first DPOAE screening. In the second DPOAE screening and BERA all the infants who gave a refer result were term infants. In 1997, Doyle et al, affirmed that there are two conditions that can be attributed to temporary hearing loss in newborns: vernix or debris in the external acoustic meatus and fluid in the middle ear. Chang et al and Del Buono et al concluded the same. Hence, on repeat testing it was seen that more infants gave pass result.[79] This could be due to the uneven distribution of two groups. Also, inappropriate probe fit due to small volume of external ear canal could be responsible for the inadequate attenuation of the noise. Hence, a louder noise floor recording could be responsible for larger refers in the initial screen, with improvement on subsequent screening. In the study done by Smolkin et al, to compare late preterm and term infants on first OAE test, showed that late preterm infants had two fold higher rates of failure on first OAE and needed repeated tests.[80]

7) BIRTH WEIGHT:

In our study of 320 infants, 36 were LBW, of which 5 were refer on 1st DPOAE screen. No significant association of birth weight and hearing loss could be established on DPOAE screening and BERA. However, the study conducted at Bobby R Alford, Department of Otolaryngology-Head and Neck Surgery, Baylor College of Medicine, Houston, concluded that although VLBW alone may not have a severe impact on hearing, it is commonly associated with multiple other risk factors that can alter hearing in a synergistic fashion.[81]

A total of 2,284 neonates were screened for the presence of OAE in Haydarpaşa Numune Educational and Research Hospital, Istanbul, Turkey found that birth weight <1,500 g is a risk factor to failure of screening with OAE in the study.[82]

8) APGAR SCORE:

There was significant association of APGAR score at one minute and first DPOAE screening in my study, with p value of 0.002. The newborns with low APGAR scores (4-6) are more likely to have a higher risk of hearing loss than infants with normal APGAR scores was observed in the first screen.

On second DPOAE screening and BERA, no such association could be established. This confirmed with the study of Christensen M et al, 2008.[81] In a study done by Augustine et al , DPOAE screening in 9448 babies, 58 babies were referred, of which 3.4 percent of the babies had low APGAR score.[73] It was seen that as infants matured a pass result was more likely.[81]

9) PRENATAL RISK FACTORS:

There was no significant association of prenatal risk factors and results of first DPOAE screening. Prenatal and maternal risk factors observed in failed cases of first DPOAE screening in my study were fetal distress, PPROM, gestational hypertension, multiple pregnancies. In a study done at Sao Paulo Hospital the most frequent risk factors for auditory deficiency were congenital infection, familial antecedent, ototoxic drugs, small for gestational age (SGA) and mechanical ventilation. In our study, on testing with second DPOAE screening and BERA no association were established. This could be attributed due to uneven distribution of infants in two groups.[79]

10) POSTNATAL RISKS FACTORS:

Postnatal risk factors observed in the referred cases of first DPOAE screening were NICU admissions, administration of ototoxic drugs like amikacin, hypoxic ischemic encephalopathy, low birth weight, very low birth weight, respiratory distress syndrome, sepsis and mechanical ventilation. Of the 320 infants, 80 had postnatal complications. Of these, 11 gave refer on 1st DPOAE screen. In 28 cases which gave a refer result in first DPOAE screening, had no post natal complications. On subsequent screening with DPOAE, 2 infants with postnatal complication gave refer results. On testing with BERA, two patients had hearing loss of which only one had post natal complication. The postnatal risk factors observed in these infants includes NICU admission, Hypoxic Ischemic Encephalopathy and administration of amikacin. No significant association could be demonstrated between postnatal complications and hearing loss.

A study done by Mietzsch et al, confirms that auditory function is transiently disrupted in almost all newborns with moderate to severe HIE. Outer hair cells as assessed by DPOAEs were a site of insult, although middle ear involvement could not

be ruled out.[83] Rebillard et al have demonstrated that DPOAEs are temporarily suppressed by inducing hypoxia.[84] The ABR waveform was delayed, although the inter-wave intervals were normal. These results suggest a cochlear insult that spared the auditory brain stem pathway and that newborns with severe HIE are at a significantly increased risk for sensorineural hearing loss.[83] However, normal brain stem transmissions in ABR studies were shown by Jiang et al. The effects Jiang et al recorded was increased by day 3 after birth and then recovered (faster for wave I than wave V) with subtle deficits remaining at 1 month.[85]

In the study of Azevedo et al, in 2004, the following risks for hearing loss were observed-ototoxic drug use, newborn with very low weight or SGA, mechanical ventilation and congenital infection, familial antecedent of hearing loss, birth asphyxia. [79]

Many factors might play a role in placing these NICU babies at an increased risk of hearing loss, including underlying disease processes as well as the treatment they receive. In addition to periods of profound hypoxia-ischemia, infants with respiratory failure may be treated with hyperventilation or alkalizing therapy, which might compromise the oxygenation and perfusion of the cochlear organ and auditory pathway, resulting in hearing loss. The use of ototoxic drugs, including loop diuretics and aminoglycosides, has been associated with increased vulnerability of the cochlea to damage from preexisting hypoxia.[86] In our study of the two infants with sensorineural hearing loss, one had prololonged NICU stay of 15 days.

11) EFFICACY OF DPOAE SCREENING :

In our study, the sensitivity of the first and second DPOAE screening was 100%. Specificity of the first and second DPOAE screening was 94%. In both stages of screening, negative predictive value was100 %. The positive predictive value of the first screening test was 10.5% and of the second screening test was 66.7%. Thus, the subsequent OAE screening helps in identifying true positive cases. Hence, the importance of repeating OAE screen cannot be overlooked. As this will decrease the burden of testing all infants with a screening test like BERA which is more invasive, costly, time consuming and requires cooperation of infant. Also the economic burden associated with the need for audiologist required for screening for hearing loss will be decreased. As the infants were chosen randomly, there were more chances of infants being normal. Also, this prevented bias when analyzing test values. As with other infant screening studies, our study also identified that screening with DPOAE is a cheap, cost effective, quick noninvasive method to be developed to screen all infants.

LIMITATIONS OF STUDY:

1) Random method of sampling was used, hence association between risk factors could not be demonstrated.
2) About 50% of the infants who failed the first screening did not appear for the rescreening (2^{nd} DPOAE Screening).
3) Due to small sample size, there were fallacies in comparison to larger studies .
4) It was a hospital based study, hence the prevalence of hearing loss in the community could be different.

STRENGTHS OF THE STUDY:

1) The prevalence of hearing loss was 6.67 per 1000 newborns, which was in concordance with world average of 6 per 1000.
2) Results of the study will be used to initiate universal newborn hearing screening in our hospital.
3) The concept “a centralized screening program, with a common data base in all hospitals in the state need to be established for universal hearing screening” was put forward. This might reduce the fail to follow up cases.

CONCLUSIONS

- Neonatal hearing evaluation was done for 320 neonates using DPOAE and BERA. It was observed that 2 infants had sensorineural hearing loss. The prevalence of hearing loss was 6.67 per thousand neonates admitted in our hospital.
- No significant association of hearing loss could be established with gender, laterality, parity, family history, gestational age, birth weight, APGAR score at 1 minute, prenatal and postnatal risk factors.
- Distortion Product Otoacoustic Emissions is an easy, cost effective and reliable method of testing of large number of infants for hearing loss. OAE is a screening test which holds good promise in hearing screening.
- BERA introduced a new era in hearing screening, but its invasive nature, need for infant cooperation, cost and need for trained audiologist to conduct the test proves as limitations for the test to be used on large number of infants as a screening tool.
- Screening programs requires the cooperation of different people, including pediatricians, nursing staff, material management personnel, medical record staff and audiologists.
- Need of the hour is to develop a screening test protocol that is community based and culturally competent. More community based studies are required, to find definite association of risk factors of hearing loss.
- OAE machine which can work effectively even in a noisy environment has to be designed.

SUMMARY

- The present study was conducted in 320 neonates selected at random admitted in our hospital. First DPOAE screening was done on neonates aged between 48-96 hours of life. 281 infants passed the first DPOAE screen and 39 had refer result for the first test.
- The second DPOAE screen was done at 45-60 days of life.19 infants who had failed the initial screen were rescreened. 20 infants failed to follow up.3 infants failed the rescreening.
- The 3 infants were subjected to BERA and 2 were found to have severe sensorineural hearing loss.
- The prevalence of sensorineural hearing loss in neonates admitted in our hospital was observed to be 6.67 per thousand neonates.
- In the present study, no significant difference in incidence of hearing loss in right and left ear was observed.
- No significant difference in hearing loss based on gender was found.
- It was seen that, parity of mother, birth weight of neonate, prenatal risks and postnatal complications had no association on the hearing loss of the infant.
- Although on initial DPOAE screen significant association between family history and hearing loss(p value-0.001), gestational age and hearing loss (p value-0.026), APGAR score and hearing loss(0.02) was inferred, no such association was seen on repeat screening with DPOAE and BERA.

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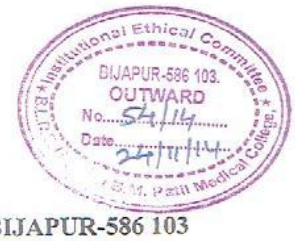
ANNEXURES

ETHICAL CLEARANCE CERTIFICATE



B.L.D.E. UNIVERSITY'S
SHRI.B.M.PATIL MEDICAL COLLEGE, BIJAPUR-586 103
INSTITUTIONAL ETHICAL COMMITTEE

INSTITUTIONAL ETHICAL CLEARANCE CERTIFICATE

The Ethical Committee of this college met on 22-11-2014 *at* 3-30pm *to scrutinize the Synopsis of Postgraduate Students of this college from Ethical Clearance point of view. After scrutiny the following original/corrected & revised version synopsis of the Thesis has been accorded Ethical Clearance.*

Title "Neonatal hearing evaluation – A Hospital - based study"

—x—x—x—

Name of P.G. student Dr. Ciju. K. George
Dept of Otorhinolaryngology.

Name of Guide/Co-investigator Dr S.R. Malipatil
Associate professor of Otorhinolaryngology

for

DR.TEJASWINI. VALLABHA
CHAIRMAN
INSTITUTIONAL ETHICAL COMMITTEE
BLDEU'S, SHRI.B.M.PATIL
MEDICAL COLLEGE, BIJAPUR.

Following documents were placed before E.C. for Scrutinization
1) Copy of Synopsis/Research project.
2) Copy of informed consent form
3) Any other relevant documents.

PROFORMA

PATIENT DETAILS

Name : Age: Sex : Hospital No :

Address :

Phone No.:

Date of Birth : Time of Birth : am/pm

Place of Birth :

Date of admission :

ANTENATAL HISTORY: G P L A POG-

Maternal Blood group:	TORCH	Diabetes Mellitus
HIV	Anaemia	VDRL
Thyroid Dysfunction	PIH	Hydramnios
Chorioamnionitis	1st Trimester Radiation exposure	Features suggestive of IUGR in Antenatal scans
Detailed drug intake during pregnancy:		

NATAL HISTORY:

Mode of delivery: Vaginal/Forceps/Vaccum/ Caesarian section (indication):

APGAR Score:

POSTNATAL HISTORY:

1) NICU Admission : A) Indication B): Duration

C) Intervention if any

Mechanical ventilation

ECMO

2) Post Natal infections:

A) Sepsis B) Pneumonia C) Meningitis

3) CNS Diseases:

A)Hypoxic Ischemic Encephalopathy

B) Interventricular Hemorrhage

C) Seizures

4) Hyperbilirubinemia

A) Cause

B) Highest Bilirubin levels : Total: Direct : Indirect:

C) Exchange transfusion: D)Phototherapy

5) Birth Trauma:

6) Ototoxic Medications to the child:

7) Others:

FAMILY HISTORY:

Consanguinous Marriage:

Family history of deafness in either parents/siblings/blood relatives:

HEAD TO TOE EXAMINATION:

ANTHROPOMETRY:

Birth weight :

Head circumference: (Microcephaly, Macrocephaly , Normal)

.

CRANIOFACIAL:

Facial symmetry and Stigmata

Cleft lip/cleft palate

Eye:

Ear:	Right	Left
1. Preauricular area A) Skin tags B) Pre auricular sinus		
2. Pinna A) Shape B) Size C) Position D) Obvious malformations		
3. Post auricular area. A) Sinuses B) Cysts/Swellings		
4. External auditory canal A) Patency B) Contents		
5. Tympanic membrane:		

Otoacoustic Emissions

RESULTS:

LEFT EAR

L1	L2	F1	F2	DP	NF	DP-NF

RIGHT EAR

L1	L2	F1	F2	DP	NF	DP-NF

COMMENTS

BERA

NEONATAL HEARING EVALUATION IN SHRI. B. M PATIL MEDICAL COLLEGE, VIJAYAPUR.

RESULT OF OAE SCREENING PROGRAM IN YOUR BABY

Name:

Date of birth:

Hospital number:

Date of test :

- **Passed the initial OAE screening which suggests a normal hearing at the time of screening.**

- **Failed the initial OAE screening in the right ear/left ear/both ears.**

[Failure of initial OAE screening can be due to blocked external auditory canal, middle ear fluid, or sensorineural hearing loss. Hence further assessment is recommended on your next visit to our hospital after 1 month at department of otolaryngology, head and neck surgery (room number: 15**)]**

Date of re-screening:

- **Passed the OAE screening which suggests a normal hearing at the time of screening.**
- **Failed the OAE screening in the right ear/left ear/both ears.**

  (*The test results are to be confirmed by BERA. This can be arranged in our hospital in department of otolaryngology, head and neck surgery)*

The child should be periodically assessed by the parents with the help of key language milestones provided below. In case of any delay in child's language milestones, you may contact your doctor further evaluation and management.

The key language milestones

Age	**milestones**
1 month	alerts to sound
3 months	coos
4 months	laughs loud
6 months	monosyllables (ba,da,pa),ah –goo sounds
9 months	bi-syllables (mama , baba ,dada)
12 months	1-2 words with meaning
18 months	8-10 word vocabulary
2 years	2-3 word sentences , uses pronouns I, me.
3 years	asks questions,know full name and gender
4 years	say song or poem, tells stories.
5 years	asks meaning of words

CONSENT STATEMENT

I confirm that Dr. Ciju K. George has explained to me the purpose of research, the study procedure that my baby will undergo, and the possible risk and discomforts as well as benefits that the baby may experience in my own language. I am willing to allow my baby to undergo the screening test. Therefore, I agree to give consent for my baby to participate as a subject in this research project.

_____

Parent/Guardian Date:

_____

_____

Signature of witness Date:

_____

I have explained to _____ the purpose of research, the procedures required and possible risks and benefits to the best of my ability in patient's own language.

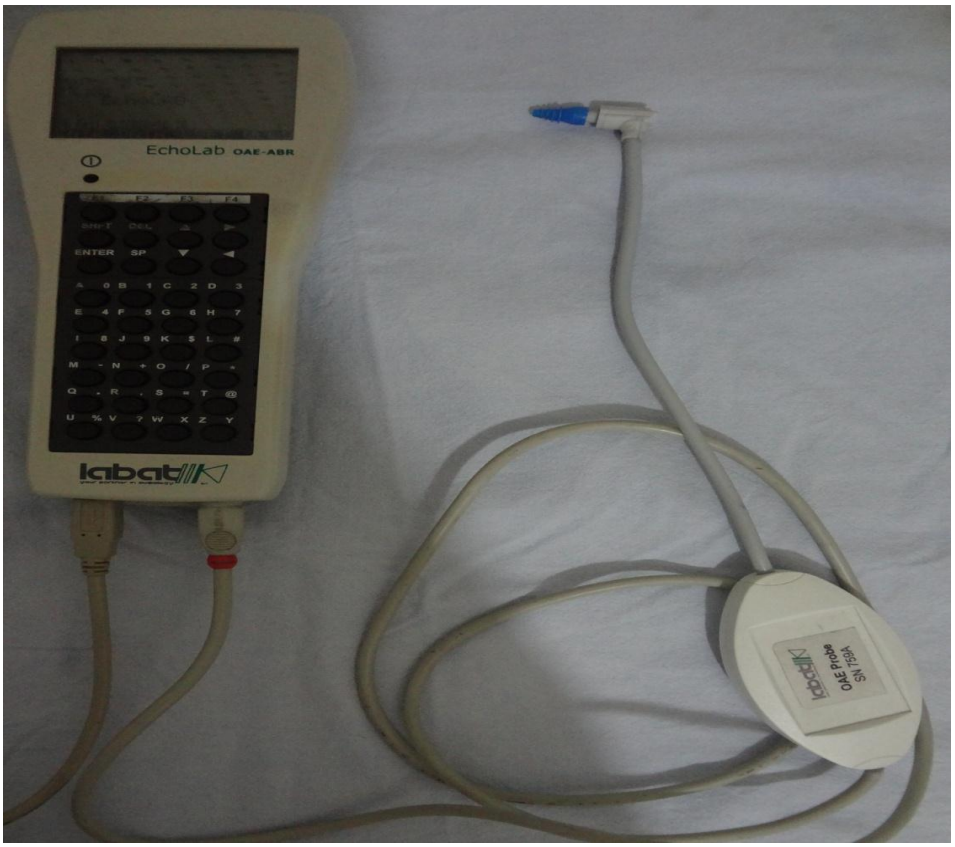
_____ _____ _____

Dr.Ciju K. George Dr. S. R. Malipatil Date

(Investigator) (Guide)

PHOTOGRAPHS

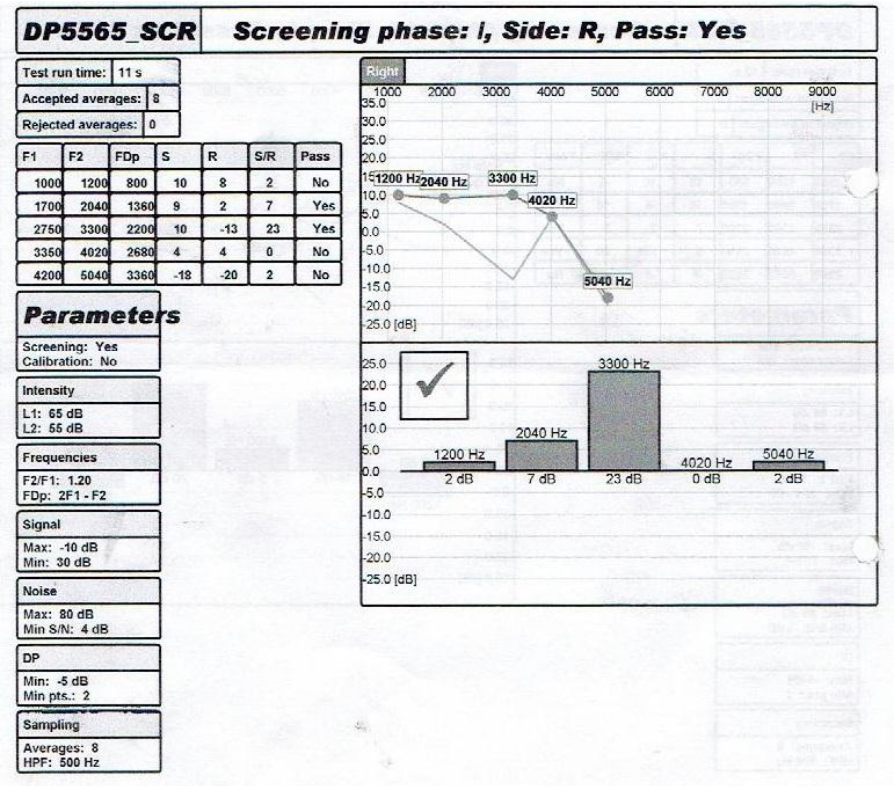
1) OAE machine



2) OAE machine connected to laptop

3) DPOAE screening of neonate

4) DPOAE test report of right ear showing pass result



DP5565_SCR — **Screening phase: I, Side: R, Pass: Yes**

Test run time: 11 s
Accepted averages: 8
Rejected averages: 0

F1	F2	FDp	S	R	S/R	Pass
1000	1200	800	10	8	2	No
1700	2040	1360	9	2	7	Yes
2750	3300	2200	10	-13	23	Yes
3350	4020	2680	4	4	0	No
4200	5040	3360	-18	-20	2	No

Parameters

Screening: Yes
Calibration: No

Intensity
L1: 65 dB
L2: 55 dB

Frequencies
F2/F1: 1.20
FDp: 2F1 - F2

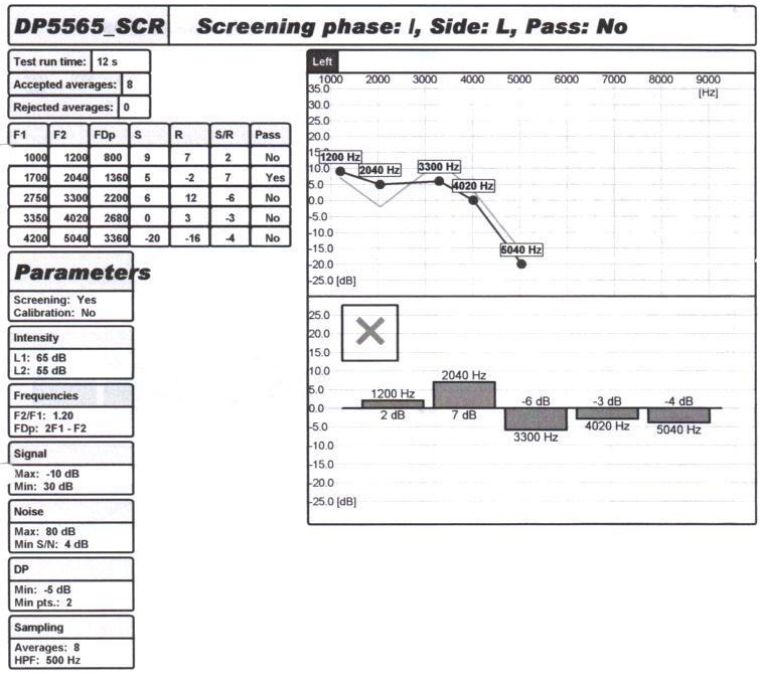
Signal
Max: -10 dB
Min: 30 dB

Noise
Max: 80 dB
Min S/N: 4 dB

DP
Min: -5 dB
Min pts.: 2

Sampling
Averages: 8
HPF: 500 Hz

5) DPOAE test report of left ear showing refer result



DP5565_SCR — **Screening phase: I, Side: L, Pass: No**

Test run time: 12 s
Accepted averages: 8
Rejected averages: 0

F1	F2	FDp	S	R	S/R	Pass
1000	1200	800	9	7	2	No
1700	2040	1360	5	-2	7	Yes
2750	3300	2200	6	12	-6	No
3350	4020	2680	0	3	-3	No
4200	5040	3360	-20	-16	-4	No

Parameters

Screening: Yes
Calibration: No

Intensity
L1: 65 dB
L2: 55 dB

Frequencies
F2/F1: 1.20
FDp: 2F1 - F2

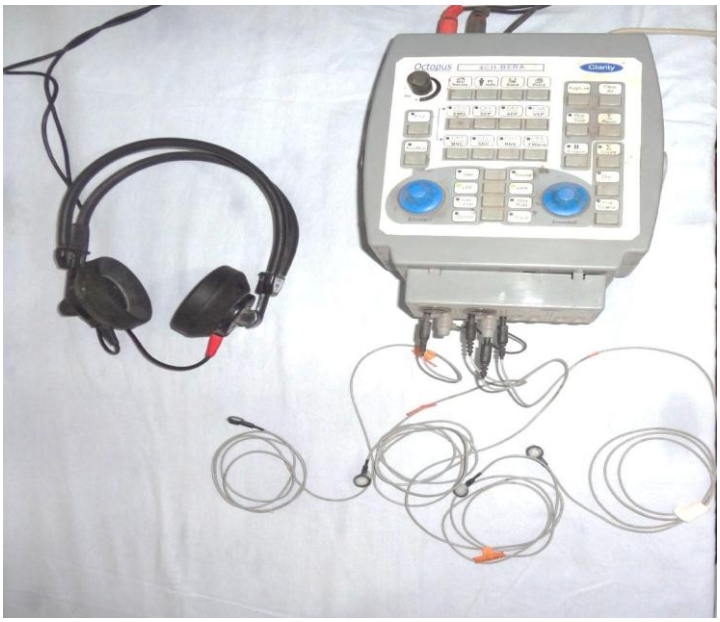
Signal
Max: -10 dB
Min: 30 dB

Noise
Max: 80 dB
Min S/N: 4 dB

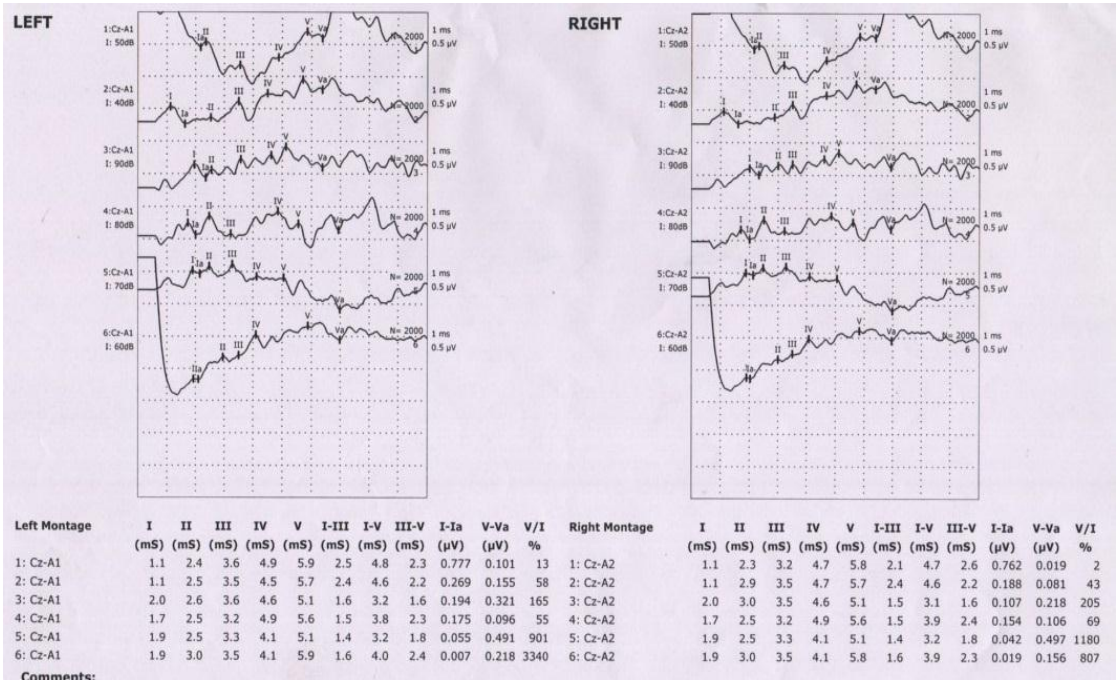
DP
Min: -5 dB
Min pts.: 2

Sampling
Averages: 8
HPF: 500 Hz

6) BERA apparatus



7) BERA report showing normal hearing

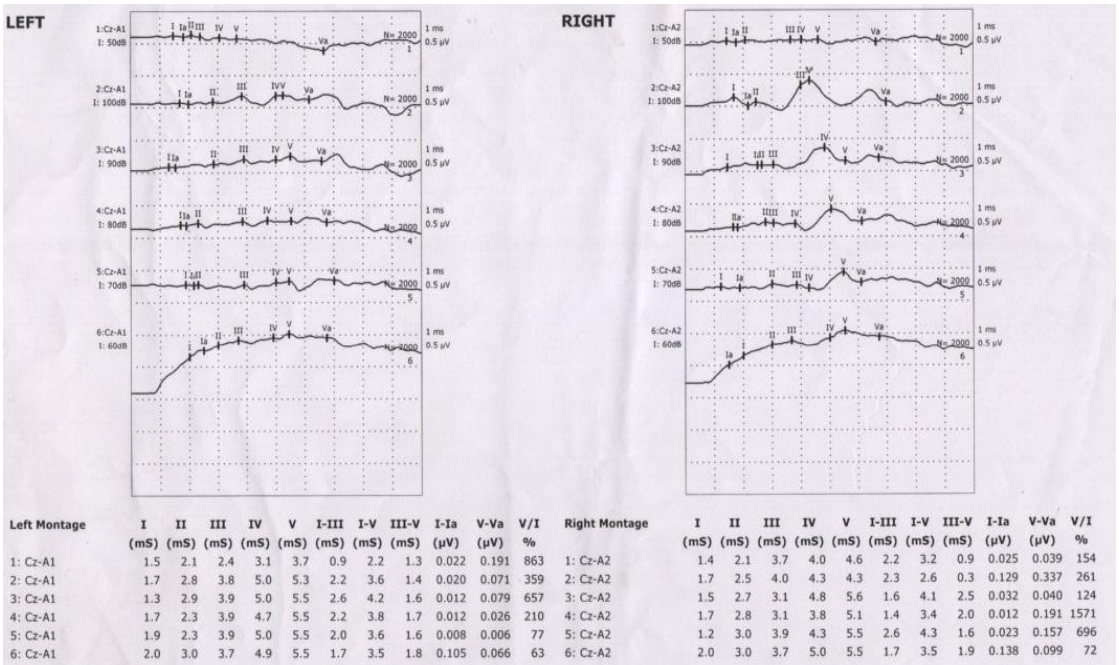


Left Montage	I (mS)	II (mS)	III (mS)	IV (mS)	V (mS)	I-III (mS)	I-V (mS)	III-V (mS)	I-Ia (µV)	V-Va (µV)	V/I %
1: Cz-A1	1.1	2.4	3.6	4.9	5.9	2.5	4.8	2.3	0.777	0.101	13
2: Cz-A1	1.1	2.5	3.5	4.5	5.7	2.4	4.6	2.2	0.269	0.155	58
3: Cz-A1	2.0	2.6	3.6	4.6	5.1	1.6	3.2	1.6	0.194	0.321	165
4: Cz-A1	1.7	2.5	3.2	4.9	5.6	1.5	3.8	2.3	0.175	0.096	55
5: Cz-A1	1.9	2.5	3.3	4.1	5.1	1.4	3.2	1.8	0.055	0.491	901
6: Cz-A1	1.9	3.0	3.5	4.1	5.9	1.6	4.0	2.4	0.007	0.218	3340

Right Montage	I (mS)	II (mS)	III (mS)	IV (mS)	V (mS)	I-III (mS)	I-V (mS)	III-V (mS)	I-Ia (µV)	V-Va (µV)	V/I %
1: Cz-A2	1.1	2.3	3.2	4.7	5.8	2.1	4.7	2.6	0.762	0.019	2
2: Cz-A2	1.1	2.9	3.5	4.7	5.7	2.4	4.6	2.2	0.188	0.081	43
3: Cz-A2	2.0	3.0	3.5	4.6	5.1	1.5	3.1	1.6	0.107	0.218	205
4: Cz-A2	1.7	2.5	3.2	4.9	5.6	1.5	3.9	2.4	0.154	0.106	69
5: Cz-A2	1.9	2.5	3.3	4.1	5.1	1.4	3.2	1.8	0.042	0.497	1180
6: Cz-A2	1.9	3.0	3.5	4.1	5.8	1.6	3.9	2.3	0.019	0.156	807

Comments:

8) BERA showing hearing loss



Left Montage	I (mS)	II (mS)	III (mS)	IV (mS)	V (mS)	I-III (mS)	I-V (mS)	III-V (mS)	I-Ia (µV)	V-Va (µV)	V/I %
1: Cz-A1	1.5	2.1	2.4	3.1	3.7	0.9	2.2	1.3	0.022	0.191	863
2: Cz-A1	1.7	2.8	3.8	5.0	5.3	2.2	3.6	1.4	0.020	0.071	359
3: Cz-A1	1.3	2.9	3.9	5.0	5.5	2.6	4.2	1.6	0.012	0.079	657
4: Cz-A1	1.7	2.3	3.9	4.7	5.5	2.2	3.8	1.7	0.012	0.026	210
5: Cz-A1	1.9	2.3	3.9	5.0	5.5	2.0	3.6	1.6	0.008	0.006	77
6: Cz-A1	2.0	3.0	3.7	4.9	5.5	1.7	3.5	1.8	0.105	0.066	63

Right Montage	I (mS)	II (mS)	III (mS)	IV (mS)	V (mS)	I-III (mS)	I-V (mS)	III-V (mS)	I-Ia (µV)	V-Va (µV)	V/I %
1: Cz-A2	1.4	2.1	3.7	4.0	4.6	2.2	3.2	0.9	0.025	0.039	154
2: Cz-A2	1.7	2.5	4.0	4.3	4.3	2.3	2.6	0.3	0.129	0.337	261
3: Cz-A2	1.5	2.7	3.1	4.8	5.6	1.6	4.1	2.5	0.032	0.040	124
4: Cz-A2	1.7	2.8	3.1	3.8	5.1	1.4	3.4	2.0	0.012	0.191	1571
5: Cz-A2	1.2	3.0	3.9	4.3	5.5	2.6	4.3	1.6	0.023	0.157	696
6: Cz-A2	2.0	3.0	3.7	5.0	5.5	1.7	3.5	1.9	0.138	0.099	72

KEY TO MASTER CHART

A	- Abortion
AM	- Amikacin
APGAR	- Apgar Score (1, 5 min)
B/L	- Bilateral
B/O	- Baby of
BERA	- Brainstem Evoked Response Audiometry
BRP	- Breech presentation
BW	- Birth weight
CM	- Consanguineous marriage
CP	- Cerebral Palsy
CPD	- Cephalopelvic disproportion
D	- Days
DOB	- Date of Birth
E	- Eclampsia
ET	- Exchange Transfusion
Family h/o	- Family history of
FD	- Fetal distress
FP	- Failure to Progress
FTFP	- Fail to follow up
G	- Gravida
GA	- Gestational age
GDM	- Gestational diabetes mellitus
GH	- Gestational hypertension
HDN	- Hemorrhagic Disease of Newborn

HIE	- Hypoxic Ischeaemic Encephalopathy..
Hosp no	- Hospital number
IUD	- Intrauterine Death
IUGR	- Intrauterine Growth Restriction
L	- Living
LBW	- Low Birth Weight
L-p	- Left ear pass
LPT	- Late Preterm
L-r	- Left ear refer
LSCS	- Lower Segment Caesarean Section
MA	- Maternal Anemia
MAS	- Meconium Aspiration Syndrome
MSL	- Meconium Stained Liquor
MV	- Mechanical ventilation
NHB	- Neonatal Hyperbilirubinemia
NICU	- Neonatal Intensive Care Unit
OBH	- Obstetric History
OH	- Oligohydraminos
P	- Parity
PE	- Pre- eclampsia
PL	- Prolonged Labour
PPROM	- Preterm Premature Rupture of Membrane
Prev	- Previous
PROM	- Premature Rupture of Membrane
PST	- Post term

PT - Preterm

RDS - Respiratory Distress Syndrome

ROD - Route of Delivery

R-p - Right ear pass

R-r - Right ear refer

SL No. - Serial Number

SNHL - Sensorineural Hearing Loss

SS - Short Stature

S - Stage

ST - Scar tenderness

SZ - Seizure

TTN - Transient tachypnoae Newborn

VD - Vaginal Delivery

VLBW - Very Low Birth Weight