Hearing Loss: A Sequela of Prematurity: A Case Study

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Abstract*: Hearing impairment becomes a very common sensory deficit seen in premature infants and its association has long been recognized. Understanding the principal lead for hearing impairment in pre-term infants becomes difficult as they show spectrum of high-risk registers which may contribute to the hearing loss. An 8-month- old preterm baby was reported with the complaint of hard of hearing and failure of OAE tests. A detailed Audiological evaluation was done and the baby was diagnosed to have severe to profound hearing loss in both the ears. Further counseling was given regarding the better management options available. Objective of this case study is to show the prime pathophysiology behind the possible etiological factors for hearing loss and to emphasize the need for a detailed Audiological evaluation in preterm infants.*

Keywords*:* pre-term, high-risk registers, hearing loss, audiological evaluation.

1.Introduction

Prematurity is defined as the birth of the child before 37 weeks of gestation (World Health Organization, 2015). It is categorized as Late preterm, born between 34 and 36 completed weeks of pregnancy, Moderately preterm, born between 32 and 34 weeks of pregnancy, Very preterm, born at less than 32 weeks of pregnancy and Extremely preterm, born at or before 25 weeks of pregnancy.

The major causes of prematurity are conceiving through in vitro fertilization, problems with uterus, cervix or placenta, previous history of miscarriage or abortions, physical injury or trauma, intrauterine infections, vaginal bleeding, high blood pressure for the mother [1]. Hearing loss result as a consequence of prematurity [2]. The prevalence of hearing impairment is estimated to be 1 – 3% among the premature population and it may vary depending on associated risk factors that accompany [4].

Some high risk factors for hearing impairment in premature infants includes a family history of hearing loss, craniofacial anomalies, congenital anomalies in relation with congenital hearing loss , congenital infections(TORCH complex), very low birth weight (<1500g), low APGAR score, hyperbilirubinemia, ototoxic medications, bacterial meningitis which require mechanical ventilation and intensive care[2]. They have a higher probability of acquiring conditions such as sepsis, iron deficiency anemia, hypoxia, respiratory distress syndrome followed by which the prolonged intensive care with incubation and ventilation, antibiotic courses that are ototoxic may be given. These may in turn affects the otolith organs [2].

Since they have a better chance of acquiring hearing loss, it is therefore important to carry out a detailed Audiological Evaluation in order to categorize the diagnosis accurately and provide appropriate recommendations to prevent the difficulties faced by a hearing handicapped.

2.Case Study

An 8-Month-old/male baby was brought to the department of Audiology for a detailed Audiological evaluation in MERF Institute of Speech and Hearing (P)LTD on 10/09/2019, with the complaint of hard of hearing and delayed speech and language development. The onset of the problem was reported to be congenital and the severity has remained unchanged. Hearing screening was done by 8 months which revealed “Refer” in OAE test.

Pre-natal history revealed that the mother was under medication for calcium. Peri-natal history revealed pre-term delivery e with a low birth weight of 1.35kgs, APGAR score obtained was 7/10. Post-natal history revealed that the child had respiratory distress and neonatal sepsis, for which the child was shifted to SNCU (Sick Newborn Care Unit) and was under CPAP (Continuous Positive Airway Pressure). Pulmonary surfactant and antibiotics (AMIKACIN 20mg)

were stepped up. On day 5, there was a setback with intermittent gasping respiration, mottling of skin, tachycardia, and fresh bleed from oronasal cavity with apnea. The infant was then incubated and put on ventilator support. FFP (Fresh Frozen Plasma) was transfused. Ophthalmic opinion reveals Retinopathy of Prematurity and a single hemorrhagic spot with exudates in inferonasal region. This is Suggestive of (?) CMV Retinopathy and (?) Anemic Retinopathy. On day 8, the baby was diagnosed to have anemia and advised to have blood transfusion. But, it was denied by the parents and hence, not carried out. The child was further recommended for hearing and vision assessment considering various risk factors. This shows the necessity of carrying out a detailed audiological evaluation in this child.

3.Clinical Audiological Procedure

An Audiological Test Battery Approach begun with a detailed case history, followed by Behavioral and Electrophysiological tests.

Behavioral Observation Audiometry was done and it revealed (?) severe to profound hearing loss.

Immitance Audiometry was done with flute inventis and tympanometry finding shows bilateral 'A' type tympanogram, indicating no middle ear pathology in both the ears.

Oto-acoustic Emission was done with Intelligent hearing system (IHS), finding revealed absent DPOAE's which suggests Outer hair cell dysfunction in both the ears. As it is depicted in the figure A.

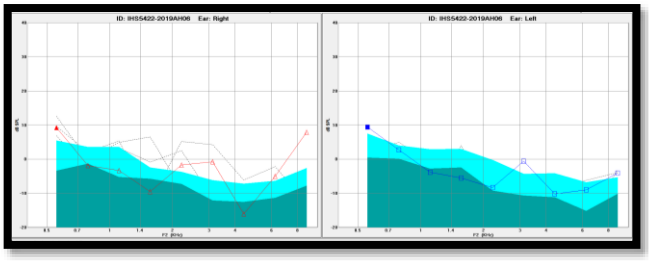

Figure A: Dp Gram showing absences of Dp OAE in both the ears

Auditory Brainstem Response was done using Intelligent hearing system (IHS) and findings shows that ABR V peak could not be obtained at 88dBnHL at the rate of 19.3/s using click stimulus which is presented in both the ears using ER3A-insert hear phones in both rarefaction and condensation polarity. It gives an impression that both ears have severe to profound hearing loss. As it is clearly depicted in the figure B.

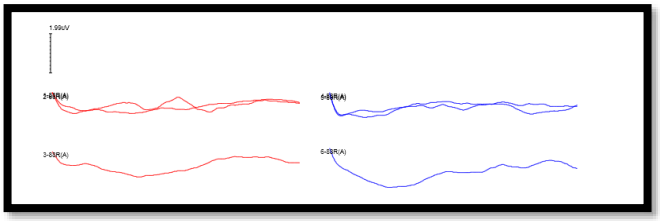

Figure B: ABR Waveform showing absence of Response in both the ears

Hearing Aid Trial was done using Beltone Boost BTE and shows that the aided responses are out of the speech spectrum. This shows hearing aid can only be used for awareness purpose not for speech perception.

Counseling regarding cochlear implant was done and baby was referred to undergo radiological evaluation in order to know the status of cochlear structures and the auditory nerve to continue with further management.

4.Discussion

Hearing loss remains a distinct cause for handicap in surviving pre-term infants and is notably associated with multiple factors than in general newborn population. The pathophysiology of hearing impairment in such infants is quite complicated and prematurity is not the only factor contributing, means that it is also linked with other high risk factors that influence the auditory system and impairing the hearing mechanism. Etiology of hearing impairment in pre-mature neonates is crucial to prevent such impairments, to promote early intervention and to promote better management options and recommendations.

One such factor says that the rapidity of fetal Audiological development occurs, during the course of 33-35 weeks of gestational age, with specific responses to different sound frequencies such as 250,500,1000 and 3000Hz [3]. Therefore, premature infants who are born before a complete development of the auditory system or during these weeks of gestation may pose as a threat for a hearing impairment.

Hearing loss among Low-birth weight infants is a common finding. Pre-term infants, with birth weight of $\leq$ 1500g are at a greater risk for hearing impairment as the incidence of high-risk registers is comparatively more confounding than those with birth weight >1500g. Even though hearing screening for such LBW fails due to transient middle ear fluid accumulation and other conductive hearing loss, it does resolve within weeks of discharge from hospital. The linkage of sensorineural hearing loss in infants with LBW still remains unknown but episodes of other risk-factors such as ototoxic drugs, hyperbilirubinemia, hypoxia, etc. may lead to progressive hearing loss. Moreover, it is also known that hearing impairment in early years and progression in later stages has an impact on central auditory processing and intellectual functioning. Thus, long-term

plans for monitoring the hearing status in infants with LBW and in those exposed to the risk factors are crucial [4]. Male individuals are more prone to the development of hearing loss as they are more vulnerable to severe neo-natal illness, such as sepsis and HMD [8].

In our study, several variables, such as a severe respiratory difficulty resulting from respiratory distress syndrome(hyaline membrane disease) which lacks a molecule surfactant in the immature lungs, are treated and have consequences such as an increased exposure to other risk factors such as acidosis, antibiotic courses, prolonged incubator and ventilator noise, which may result in hearing impairment and other neurological impairments[8].

Aminoglycosides and Non-Aminoglycosides have deleterious impacts in the cochlea and vestibular organs by causing hair cell death. Aminoglycosides that are cochlear specific contains neomycin, amikacin, kanamycin and vestibular specific drugs contains gentamycin, tobramycin and streptomycin. The pathophysiology behind these drugs show that the ionic currents are blocked via mechanoelectrical transduction channels in stereocilia which are then taken by the hair cells via apical endocytosis. Free radicals are formed that lead to cell damage via the reactive species and the damage predominantly affects the high-frequency and then progressing to the low-frequency [4]. These may have predicted a hearing deficit not because of specific ototoxicity, affecting the cochlear functions, but because of poor immune infants who are pre-term.

Hypoxia has a greater link with hearing loss due to inadequate oxygenation and perfusion which affects the cells of spiral ganglion. As the severity increases, it causes irreversible cellular damage to the cochlea especially the outer hair cells and stria vascularis, thus impairing the normal cochlear function. At this point, it is important to note that hyperventilation due to respiratory failure may further decrease the oxygenation of the auditory system [4].

An experimental study on otopathy due to sepsis showed that sepsis leads to significant hearing impairment. A prospective study was done on laboratory mice where regularly hearing animals were infected using cecal ligation puncture technique and a remarkable hearing loss was observed at all frequencies. The pathophysiology contributing to such loss is due to apoptosis in the supporting cells of organ of corti and glutamate excitotoxicity at the basal pole of inner hair cells. Thus, it was proven that hearing loss could occur in the absence of ototoxicity and it also highlights that hearing evaluation is a must for those suffering from sepsis syndrome [5].

Iron Deficiency Anemia is one of the major health problems and it posing as a causative variable for hearing impairment in neonates has been supported. Even though the exact cause for IDA leading to hearing impairment nor its treatment options promotes hearing restoration have not been supported, it's related to the sensitivity of the cochlea to vascular and neurologic effects of IDA. It is known that cochlea is supplied by labyrinthine artery and lack of its circulation might make it more prone to ischemic effect of IDA. Since, IDA compromises tissue oxygen delivery, cochlea becomes more vulnerable to reduced oxygen in blood and thus impairing its normal functions, resulting in a sensorineural hearing loss [6].

The long-term impact of neonatal and infant hearing loss are given major importance as it has an impact on all walks of life such as communication, behavior, cognition, social and emotional development, academic outcomes and other phases, too. Fortnum et al. reported that the prevalence of permanent hearing handicap continues to rise until the age of 9 years and 205 per 100,000 for the general population[4]. Similarly, the prevalence of hearing handicap for the high-risk infants is comparatively higher and hence, a need for a detailed Audiological evaluation per annum is necessary.

5.Conclusion

It is thus of highest priority, for those infants who are born pre-term and those with sensitive results from a neonatal hearing screening or with high-risk registers, to undergo a detailed hearing evaluation so that a correct diagnosis can be arrived at, in order to be recommended the immediate possible management options suitable for that particular individual. In fact, the early identification and intervention follows a better understanding of the pathophysiology of the associated conditions. The sooner and better the habilitation, the easier it is to overcome the difficulties of a handicapped.

6.Reference

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