International Journal of Early Childhood Special Education (INT-JECSE)
DOI:10.9756/INTJECSE/V14I5.1000 ISSN: 1308-5581 Vol 14, Issue 05 2022

A Case Report On Bilateral Congenital Profound Sensorineural Hearing Loss

1] Ms. Shraddha Lokhande* 2] Ms. DhartiMeshram 3] Mrs. InduAlwadkar 4] Ms. Priyanka Meshram 5] Ashish Bhagat6]Anjali Alone

1] Ms. Shraddha Utamaro Lokhande* GNM 3rd year,
Nightingale Training College of Nursing, Sawangi (M) Wardha.
Datta Meghe Institute of Medical Sciences (Deemed to be University) Sawangi (M) Wardha.
Email I'd: shraddhauttamlokhande@gmail.com No.9552796249
2] Ms. Dharti Meshram** Nursing Tutor, Department of Mental Health Nursing.
Florence Nightingale Training College of Nursing,
Datta Meghe Institute of Medical Sciences (Deemed to be University) Sawangi (M) Wardha.
Email Id- dhartimeshram9@gmail.com Mobile No. 7507517801, 8605556166
3] Mrs. Indu Alwadkar*** Principal of FNTCN, Department of Community Health Nursing.
Florence Nightingale Training College of Nursing,
Datta Meghe Institute of Medical Sciences (Deemed to be University) Sawangi (M) Wardha
Email Id- indualwadkae@gmail.com Mobile No 9960278995
4] Ms. Priyanka Meshram**** Nursing Tutor, Department of Mental Health Nursing.
Florence Nightingale Training College of Nursing,
Datta Meghe Institute of Medical Sciences (Deemed to be University) Sawangi (M) Wardha.
Email Id- priyankameshramganvir@gmail.com Mobile No. 8600289201
5]Research Scientist, Jawaharlal Nehru Medical College, Datta Meghe Institute of Medical Sciences, Sawangi, Wardha, Maharashtra.
6] Department of Medical-Surgical Nursing, Smt. Radhikabai Meghe Memorial College of Nursing, Datta Meghe Institute of Medical Sciences, Sawangi, Wardha, Maharashtra.

ABSTRACT
One to three neonates out of every 1000 have congenital hearing loss. Many children who are born deaf have non-syndromic hearing loss, which is an isolated finding. If a patient suffers hearing loss in addition to other medical issues, it is referred to as syndromic hearing loss. Around 20% of kids with genetics also have other related symptoms. This exercise outlines the diagnostics and therapeutics of syndromic sensorineural loss of hearing and emphasizes the importanceof the interprofessional team in enhancing patient care. Different nations have varying rates of sensorineural loss of hearing Between 5 and 27 per100,000 people in the US experience abrupt SNHLeach year, with about 66,000 new occurrences. Congenital hearing loss can haveseveral etiologies and is almost always sensorineural.The author presented a case of 4 yrs. Male develop bilateral congenital profound sensorineural hearing loss due to infectious diseases passed from the mother to child in the womb. The current condition the patient received at birth but the patient's mother observes this deficiency after 2 years. Inability to speak, inability to hear in both ears, and he responds to only loud sounds. All initial Investigations were carried out such as history taken, physical examination, and laboratory testing. RT PCR Negative, HIV, HBsAg, HCV- non-reactive, Chest X-ray - within normal limits, the patient received all immunization according to schedule. BERA test was carried out using alternative click stimuli from 105 DB NHL to 90 DB NHL. No clear and repetitive v peak was obtained at 105 dB NHL for both ears, left ear- severe to profound hearing loss in the right ear. ASSR test-. severe to the left ear profound severe to profound hearing loss in the right ear. MDCT scan of temporal bone precision daintily scan. Cochlear implantation is done. The patient's response was good.

Keywords: Syndromic sensorineural hearing loss, SNHL RTPCR, HBsAg, HCV- non-reactive,

INTRODUCTION

Inborn hearing loss When the hearing capacity to transform the automated vibration into the electrical energy of nerve impulses from sound is compromised, a hearing loss exists at birth[1.] loss of hearing is divided into two categories based on where the lesion is located: While sensorineural hearing loss affects the inner ear, auditory

nerve, and/or middle ear, conductive hearing loss affects the middle or outer ear or central auditory pathway.[2] Congenital infectious causes, such as CMV, are uncommon in patients who receive comprehensive prenatal care, whereas genetic factors are more likely.[3]

Conductive and sensorineural hearing loss are both considered types of mixed hearing damage. When someone has conductive hearing loss, sound waves pass by way of the ear due to improper middle ear, external ear, or both development, or as a result of temporary center ear obstruction brought on by using effusion (as in the case of otitis media) There are three types of sensorineural loss of hearing sensory (when the hair follicles disturbed), the middle (when the reason is found in the auditory pathway in the middle), and autistic disease on the neuropathy spectrum[4] Speech discrimination is hampered by Disorder of the Auditory Neuropathy Spectrum which encompasses a wide variety of medical diseases marked otoacoustic emissions and a cochlear microphonic are present, but auditory brainstem responses are lacking or have defects.[5]

The spectrum of Auditory Neuropathy's initial damage to the inner hair cells, and intervening synapses either in the auditory nerve or damage to certain populations of neurons in the auditory pathway can all contribute to a disorder.[6] Programs for newborn hearing screening are available for this. an issue in the majority of developed nations.[7] Within a month of birth, these coders hope to screen all newborns. Early diagnosis, intervention, and treatment help children develop more successfully later in life.[8] Neonatal hearing screening programs may overlook infants with progressive loss of hearing since, Over time, hearing loss can get worse. For newborns who are at risk, frequent screening at regular intervals is indicated. According to the cause and kind of loss of hearing, congenital loss of hearing is treated medically and with supportive measures.[9]

Congenital infections, genetic components, and cranial-facial anomalies are the most typical contributors to hearing loss, such as non-syndromic types in which the only clinical condition is hearing loss symptoms in both syndromes including Lange-Nielsen, Jervell, and Usher syndromes. In this introduction, we specifically address persistent hearing loss that is congenitally present in both ears, which hearing loss is referred to as 40 dB averaged over the frequency range of the ear with superior hearing crucial for speech recognition, which is commonly evaluated by hearing screening (500, 1,000, 2,000, and 4,000 Hz) programmers.There is a discussion of epidemiology, causes, diagnostics, and management.

Hearing evaluation loss genetically. Genetic testing is used for some patients who might identify the cause that they heard loss and support a medical diagnosis.[10] However, due to the rapidly growing number of hearing loss that runs in the family (HHI) genes revealed both the comparably poor clinical distinction of the HHIs,[11] a gap is emerging among fundamental genetic studies on hereditary deafness. likewise clinical otology. Trying to follow along with the overview includes progressive low bilateral cochleovestibular impairment, frequency hearing loss late-onset progressive high loss of hearing frequency, and bilateral congenital hearing loss. Also discussed are several syndrome variants of HHI and the availability of genetic diagnostics. Finally, a quick description of the prerequisites for successful linkage analysis, a crucial tool in genetic research for identifying probable genes for a trait on a chromosome

PRESENTATION OF CASE

This case was selected from Rural Hospital Sawangi Meghe Wardha. The author presented a known case of 4 yrs. a male child who came to the hospital with their mother with chief complaints of developmental delay (inability to speak and bilateral hearing loss) and developed congenital profound sensorineural hearing loss due to an infectious disease passed from the mother to child in the womb. Past Pediatrics History Before approaching these hospitals they don't take treatment in a private clinic and had a history of hearing loss. After the child's mother observe the child's condition inability to speak, and bilateral hearing loss they came to the present hospital. All initial Investigations were carried out such as history taken, physical examination, and laboratory testing. RT PCR HIV, HBsAg, test negative. He received all immunization according to the national immunization schedule. BERA test was carried out using an alternative click stimulus from 105 DB NHL to 90 DB NHL. No clear and repetitive v peak was obtained at 105 DB NHL for both ears, left ear- extreme to profound severe to profound hearing loss in the right ear ASSR test- left ear- severe to profound hearing loss, right ear - severe to profound hearing loss, MDCT scan of temporal bone precision daintily scan. Impression - no significant abnormality is seen in the present study portion of inner ear structures. No abnormal soft tissue or bone erosion is seen in the expected course of descending facial nerve. Bilateral mastoid air cells show a normal pneumatization pattern. MRI brain for cochlear nerve evaluation done. Bilateral cp angle cisterns appear normal. The vii - viii cranial nerve complexes are symmetrical, arising from the anterolateral surface of pons, and show normal course through the cp angle with usual signal intensity. Bilateral cochlear nerves appear normal in course and caliber (approximately 1 to 1.2mm) Limited MRI study of the brain

does not reveal any significant structural or parenchymal abnormalities. DPOAE test -done and bilateral 'refer', s/o OHC dysfunction. He received treatment of injectable KCL 4ml, injectable cefuroxime 700 mg injectable pan 15mg, injectable Emset 1.5 mg, injectable Neomol 20ml, SypIbugesic plus 7ml for 2 days Syp Taxol dx 3.5 ml for 3days, Sypsinarest 1 tsp for 3 days, l/a venusia max lotion for 2 days. Cochlear implantation is done. Patient response was good advice given to patient relatives review in ENT OPD in case of following signs and symptoms watch for headache, earache, ringing sensation, dizziness and review in Ent. Follow-up.

DISCUSSION

Since most so-called industrialized nations successfully implemented prenatal children with congenital hearing loss are increasingly subjected to hearing testing. discovered early and auditory therapy, including cochlear implants, is well established.[12-21]Aetiological focus, however, is frequently absent. An evidence-based strategy for an efficient aetiological work-up is not widely agreed upon. There is not a lot of agreement on both the content and methodology of screening. 13 To enhance evidence-based management and pinpoint areas for future action, this qualitative analysis sought to clarify the current state of aetiologies and investigations for kids with bilateral congenital SNHL.

A paper from Sierra Leone published a few years ago showed an opposite correlation between the prevalence of measles- loss of sensorineural hearing and the country's higher immunization rate (1995; Wright & Leigh).However, the authors note that there may be circumstantial evidence connecting the dramatic reduction in hearing loss brought on by the measles incidence with the reduction in the number of measles cases that have been recorded. They assert that an examination of sensorineural hearing loss in a pertinent group could provide more concrete proof of the association (Wright & Leigh, 1995).

Malaria. is the most frequent reason for febrile sickness in a child in Nigeria and West Africa 1(Mc Pherson&Holborrow, 1985). Numerous investigations in the West Africa region have linked deafness to the etiology of malaria and the medications used to treat it Holborow et al., 1982; Wright & Leigh, 1995; Bondi, 1992; Chukuezi, 1991. Further research is required to determine whether malaria can make children deaf. Malaria infection is thought to have the potential to lead to deafness either locally through microvascular alterations in the cochlea's end arteries or generally by reducing disease resistance and amplifying the negative effects of other illnesses.

In current society, the reasons for sensorineural hearing loss inherited and congenital are poorly understood and rarely detected at an early stage.[21-30] Heredity has been linked to childhood hearing loss in South Africa (8 percent) and Gambia (11 percent) (Sellars & Beighton, 1983; Mc Pherson&Holborrow, 1985). The associated stigma makes family tracing challenging. In this investigation, we were unable to pinpoint a particular hearing loss in families. Here is most likely because of the aforementioned factor. In the majority of healthcare delivery facilities, genetic research is practically nonexistent. Nigeria is not unique in this regard (Brobby, 1988). It is still unknown how parents will react to a deafness diagnosis made during pregnancy. More thorough research is required in this area because unidentified causes make up a third of the etiological components.[31]

CONCLUSIONS

9.0 percent of cases of Children with bilateral and unilateral sensorineural hearing loss (SNHL) Children with bilateral and unilateral sensorineural hearing loss (SNHL) are brought on by congenital cytomegalovirus infection with (CMV). In cases of bilateral profound SNHL, the diagnosis of the use of A combination CMV DNA detection test and genetic deafness test was reported to be 46.4 percent. In the current investigation, congenital CMV infection prevalence was examined in till the age of 12, children with unilateral or bilateral SNHL.CMV infection in utero was retrospectively determined by CMV DNA detection in dried umbilical cord tissues. Methods: 134 dried umbilical cords were preserved kids with either unilateral (88 kids) or bilateral (46 kids) SNHL were gathered. The dried cords of the womb DNA were taken, and statistical PCR was used to find CMV DNA. Bilateral SNHL in children underwent genetic hearing loss studies. according to the intruder assay.

Conflict of interest: - No conflict.

REFERENCES:

1. Cole EB, Flexer C. Children with hearing loss: Developing listening and talking, birth to six. Plural Publishing; 2019 Jul 22.

2. Sininger YS, Doyle KJ, Moore JK. The case for early identification of hearing loss in children: auditory system development, experimental auditory deprivation, and development of speech perception and hearing. Pediatric Clinics of North America. 1999 Feb 1;46(1):1-4.
3. Korver AM, Smith RJ, Van Camp G, Schleiss MR, Bitner-Glindzicz MA, Lustig LR, Usami SI, Boudewyns AN. Congenital hearing loss. Nature reviews Disease primers. 2017 Jan 12;3(1):1-7.
4. Moser T, Predoehl F, Starr A. Review of hair cell synapse defects in sensorineural hearing impairment. Otology & Neurotology. 2013 Aug 1;34(6):995-1004.
5. Eggermont JJ. Hearing loss: Causes, prevention, and treatment. Academic Press; 2017 Feb 22
6. Shearer AE, Hansen MR. Auditory synaptopathy, auditory neuropathy, and cochlear implantation. Laryngoscope investigative otolaryngology. 2019 Aug;4(4):429-40.
7. Swanepoel DW, Hugo R, Louw B. Infant hearing screening at immunization clinics in South Africa. International journal of pediatric otorhinolaryngology. 2006 Jul 1;70(7):1241-9.
8. Yoshinaga-Itano C. From screening to early identification and intervention: Discovering predictors to successful outcomes for children with significant hearing loss. Journal of deaf studies and deaf education. 2003 Jan 1;8(1):11-30.
9. Fligor BJ, Neault MW, Mullen CH, Feldman HA, Jones DT. Factors associated with sensorineural hearing loss among survivors of extracorporeal membrane oxygenation therapy. Pediatrics. 2005 Jun;115(6):1519-28.
10. Alford RL, Arnos KS, Fox M, Lin JW, Palmer CG, Pandya A, Rehm HL, Robin NH, Scott DA, Yoshinaga-Itano C, Professional Practice and Guidelines Committee. American College of Medical Genetics and Genomics guideline for the clinical evaluation and etiologic diagnosis of hearing loss. Genetics in medicine. 2014 Apr 1;16(4):347-55.
11. Topsakal V, Van Camp G, Van de Heyning P. Genetic testing for hearing impairment. B ENT. 2005 Jan 1;3:125.
12. Morton CC, Nance WE. Newborn hearing screening—a silent revolution. New England Journal of Medicine. 2006 May 18;354(20):2151-64.
13. Collins SA, Currie LM, Bakken S, Vawdrey DK, Stone PW. Health literacy screening instruments for eHealth applications: a systematic review. Journal of biomedical informatics. 2012 Jun 1;45(3):598-607.
14. Estivill X, Govea N, Barceló A, Perelló E, Badenas C, Romero E, Moral L, Scozzari R, D'Urbano L, Zeviani M, Torroni A. Familial progressive sensorineural deafness is mainly due to the mtDNA A1555G mutation and is enhanced by treatment with aminoglycosides. The American Journal of Human Genetics. 1998 Jan 1;62(1):27-35.
15. Estivill X, Govea N, Barceló A, Perelló E, Badenas C, Romero E, Moral L, Scozzari R, D'Urbano L, Zeviani M, Torroni A. Familial progressive sensorineural deafness is mainly due to the mtDNA A1555G mutation and is enhanced by treatment with aminoglycosides. The American Journal of Human Genetics. 1998 Jan 1;62(1):27-35.
16. Akhuj, Aditi, SnehalSamal, Rakesh Krishna Kovela, RaginiDagal, and Rebecca Thimoty. “Impact of Balance Training and Co-Ordination Exercises in Post-Operative Left Cerebellopontine Angle Tumor: A Case Report.” Journal of Pharmaceutical Research International, December 14, 2021, 413–16. https://doi.org/10.9734/jpri/2021/v33i57A34014.
17. Akther, Jawed, Y. R. Lamture, Varsha P. Gajbhiye, Ranjit Ambad, and Aditya V. Ghunage. “Assessment of Proximal Radio-Median Cubital/Radio-Cephalic Arterio-Venous Fistula.” Journal of Pharmaceutical Research International, December 15, 2021, 612–19. https://doi.org/10.9734/jpri/2021/v33i58A34158.
18. Ali, Sabir, Renu Rathi, and Bharat Rathi. “A Comparative Study on the Efficacy of Kantkari and Vasa Lozenges in Children with Kasa (Cough)-Study Protocol.” Journal of Pharmaceutical Research International, June 11, 2021, 25–33. https://doi.org/10.9734/jpri/2021/v33i31B31685.
19. Alnewar, Pragati, Seema Singh, and Vaishali Tembhare. “Effectiveness of Planned Teaching on Knowledge Regarding Noonan Syndrome among Nursing Students.” Journal of Pharmaceutical Research International, October 29, 2021, 709–13. https://doi.org/10.9734/jpri/2021/v33i47A33065.

20. Alwadkar, Sagar, and Deeplata Mendhe. "Effectiveness on Simulation Teaching Regarding Cardio Pulmonary Resuscitation among Workers of State Transport Depot." Journal of Pharmaceutical Research International, October 12, 2021, 111–15. https://doi.org/10.9734/jpri/2021/v33i46A32846.
21. Alwadkar, Sagar, Mayur B. Wanjari, Pratibha Wankhede, and Deeplata Mendhe. "Cerebral Palsy with Gross Developmental Delay with Lower Respiratory Infection with Hypoxia- A Case Report." Journal of Pharmaceutical Research International, September 22, 2021, 199–202. https://doi.org/10.9734/jpri/2021/v33i44B32666.
22. Alwadkar, Sagar, Pratibha Wankhede, and Mayur Wanjari. "Promotion of Children Health with Giving Supplement of Rainbow Nutrition: A Review." Journal of Pharmaceutical Research International, October 12, 2021, 88–92. https://doi.org/10.9734/jpri/2021/v33i46A32843.
23. Ambad, Ranjit, Roshan Kumar Jha, NandkishorBankar, and Sachin Patil. "Prevalence of Undernutrition and Its Associated Factors in Tribal Population of Gadchiroli (Vidarbha Region)." Journal of Pharmaceutical Research International, December 15, 2021, 167–73. https://doi.org/10.9734/jpri/2021/v33i58B34187.
24. Ambad, Ranjit S., Suryakant Nagtilak, GangaramBhadarge, and Meghali Kaple. "Circulatory Glutathion S Transferases Estimation in Chronic Alcoholics Vising Urban and Rural Health Center." Journal of Pharmaceutical Research International, December 15, 2021, 132–36. https://doi.org/10.9734/jpri/2021/v33i58B34181.
25. Ambad, Ranjit S., Suryakant Nagtilak, GangaramBhadarge, and Meghali Kaple. "Glutathione-S-Transferase Pi and Malondialdehyde in Alcoholic Patients Attending Smhrc and Avbrh Hospital." Journal of Pharmaceutical Research International, July 13, 2021, 26–30. https://doi.org/10.9734/jpri/2021/v33i37A31975.
26. Ambad, Ranjit S., Suryakant Nagtilak, DattuHawale, and Ashish Anjankar. "Comparitive Study of Glutathione-S-Transferase Isoenzyme and Vitamin D Levels in Smokers and Non-Smokers." Journal of Pharmaceutical Research International, December 15, 2021, 118–23. https://doi.org/10.9734/jpri/2021/v33i58B34179.
27. Ambad, Ranjit S., Suryakant Nagtilak, Rakesh Kumar Jha, and Meghali Kaple. "Study of Glutathione-s-Transferase and Reduced Glutathione Receiving Chemotherapy." Journal of Pharmaceutical Research International, July 29, 2021, 134–40. https://doi.org/10.9734/jpri/2021/v33i39A32151.
28. Ambad, Ranjit S., Suryakant Nagtilak, Ankita Kondalkar, and Ashish Anjankar. "Study of Glutathione S Transferases and Malondialdehyde Levels in Male Smokers from Vidharbha Region, India." Journal of Pharmaceutical Research International, July 12, 2021, 167–72. https://doi.org/10.9734/jpri/2021/v33i36B31964.
29. Ambade, Ratnakar, and Ankit Jaiswal. "Anthropometric Analysis of Human Knee Joint in Indian Population." Journal of Pharmaceutical Research International, December 15, 2021, 478–84. https://doi.org/10.9734/jpri/2021/v33i58A34141.
30. Ambekar, Shrikant Madhukar, S. Z. Quazi, Abhay Gaidhane, and Manoj Patil. "Steps towards Universal Health Coverage through Health and Wellness Center under Ayushman Bharat Programme Delivering Comprehensive Primary Health Care in Bhandara District." Journal of Pharmaceutical Research International, June 30, 2021, 34–38. https://doi.org/10.9734/jpri/2021/v33i34A31820.
31. Ambekar, Shrikant Madhukar, S. Z. Quazi, Abhay Gaidhane, Manoj Patil, and Roshan Umate. "Bibliometric Analysis of Publications from Pubmed on Non Communicable Disease and Ayushman Bharat Health and Wellness Center." Journal of Pharmaceutical Research International, August 3, 2021, 220–28. https://doi.org/10.9734/jpri/2021/v33i39B32198.