International Journal of Early Childhood Special Education (INT-JECSE)
DOI:10.9756/INTJECSE/V14I5.997 ISSN: 1308-5581 Vol 14, Issue 05 2022

A Case Report On 2.5-year-Old Child with Bilateral Congenital Profound Sensorineural Hearing Loss with Developmental Delay with Cochlear Implant Surgery

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Abstract:
Most people have congenital hearing loss. This type of hearing loss happens during pregnancy. The causes are Prematurity, maternal hyperglycemia, anoxia before birth, genetics, and infectious disorders like rubella transmitted from mother to baby in utero.Early diagnosis of hearing loss in some neonates allows for timely treatment with cochlear implants or hearing aid treatment to support language development. Cochlear implants may enable better speech comprehension in both quiet and noisy situations with the aid of therapy. We discuss Bilateral Congenital Profound Sensorineural Hearing Loss with Developmental Delay and Cochlear Implant Surgery.This2.5-year-old female child registered in the ENT department, with the chief complaint of inability to hear in both ears and inability to speak; Static noticed when the child was at the age of 2.5 years responded only to loud sounds. An examination and evaluation of a doctor follow the diagnosis of bilateral severe to profound hearing loss with developmental delays. Medication was given to a child as per the doctor's order, Ivf DNS 400ml +Inj. KCL 4ml 8 Hourly, Inj. Cefuroxime 700 Mg Bd, Inj Pan 15mg Od, Inj. Emeset 1.5 Mg Tds, Inj. Neomol 20ml Tds, Inj. Dexa 2 Mg Bd, SypIbugesic Plus 7ml Tds ,Syp. Taxol Dx 3.5 mlTdsSyp.Sinarest 1 Tsp, N/D Otrivin (P) 2 Drops Tds.After treatment child's condition improved.

Keywords: Bilateral severe profound hearing loss, Sensorineural hearing loss, congenital, developmental delays, Cochlear Implant Surgery.

Introduction:
Sensorineural hearing loss develops when the inner ear or hearing nerve functions incorrectly.[1] An audiometric pure-tone average of more than 70 dB hearing level indicates severe to profound hearing loss, which makes normal conversation nearly impossible without amplification.[2]congenital hearing loss that is present at the birth
Hereditary factors have a significant role in congenital deafness in children, and the majority of risk factors for deafness are preventable. Therefore, many occurrences of hearing loss in childrencan be avoided with genetic counselling before consanguineous marriage, early diagnosis, and timely management.[3]
Recurrent otitis media and neurologic abnormalities such as cerebral palsy, developmental or motor delays, and seizure disorders were the most frequent medical problem seen.[4] Patients with bilateral severe to profound sensorineural hearing loss may benefit from auditory rehabilitation with a cochlear implant (SNHI).[5]
More than 500 million individuals worldwide have deafness, making it the most prevalent neurological disease.[6] One to three out of every 1000 new babies in developed nations is thought to have sensorineural hearing loss (SHL). It is anticipated that this rate will be higher in developing countries.[7] Severe to profound sensorineural hearing loss (SNHL), which affects 1 in 2 000 newborns, affects 6 in 1000 people by 18 years.[8]
The most frequent medical issues were recurrent otitis media, followed by neurologic abnormalities such as cerebral palsy, developmental or motor delays, and seizure disorders. In unilateral SNHL, the average age at diagnosis was

3.9 years; in cases of bilateral moderate or worse SNHL, the average age was 3years.[9]Computed tomography scanning was the most beneficial diagnostic investigation.[10]

Patient Information:
A 2.5-year female child was brought to a tertiary care rural hospital in the ENT department with the chief complaint of Inability to Speak and Bilateral Hearing Loss since Birth.
Primary concern and symptoms: Narrated by mother a 2.5-year female child was admitted to the ENT ward, herchild was two years five months old with chief complaint of inability to hear in both ears and inability to speak, static, noticed when a child responded to only loud sounds since birth. After the general examination and investigation, case of Bilateral Congenital Profound Sensorineural Hearing Loss was diagnosed with developmental delays. After all, the investigation resulted the doctor did her right ear cochlear implant surgery.
Medical History and Psychosocial History: History of Developmental Delay was Delayed Walking by 3-4 Months. The child belongs to a Joint Family background. She had a positive relationship with her parents. History of Consanguinity Present, No Family History of Deafness or Other Impairment, her Sleep, Appetite, Bowel, and Bladder were Normal.
Birth History: C-section delivery at the hospital, birth weight of 3.75 kg, and no ICU admission history. She immunized Up To AgeandThey were also given Pneumococcal Covalent, Homophiles Influenza, Type B Meningococcal vaccine.
Physical examination and clinical findings: on admission, physical examination child were well oriented and conscious. She looks anxious. Vital parameters area febrile, Blood pressure was 102/68mmhg, Pulse was 102 bpm, respiration was 22bpm, BMI was 1.08, BSA was 1.18. General Examination was fair. Decubitus and Nutritional Status was normal. **Systemic examination:** The abdominal review was normal, respiratory and cardiovascular systems also normal. **Local examination:** EAR was pre auricular –Bilateral regular, auricular was Bilateral normal, post auricular was Bilateral normal, Tragal tenderness was absent, EAC was normal, TM was Bilateral intact. **Nose:** external deformity- absent, DNS to the right, bilateral inferior turbinate hypertrophy presentsPara nasal sinus examination: tenderness absent. Oral cavity: mouth opening: adequate, gums - normal, the floor of mouth- normal, hard palate- normal, throat: PPW: clear
Timeline: Narrated by the mother A 2.5-year female child was admitted to the ENT ward, the child was two years five months old with developmental delay and complaint of inability to hear in both ears and inability to speak, static, noticed when a child was at the age of 2.5 years responds to only loud sounds and also the history of Developmental Delay was Delayed Walking By 3-4 Months After the general examination and investigation found out diagnosis of Bilateral Congenital Profound Sensorineural Hearing Loss with developmental delays. Her right ear cochlear implant surgery was done.
Diagnostic assessment:A routing investigation was done.HIV, HBsAg, and HCV were non-reactive,and **Chest X-Ray** and **ECG** were within normal limits.**BERA**Test Was Carried Out Using an Alternative Click Stimulus From 105db NHL To 90db NHL. No Clear and Repetitive V Peak Was Obtained at 105 Db NHL for Both Ears. Left Ear and Right Ear - Severe To Profound Hearing Loss.**ASSR TEST**-LEFT EAR- Severe To Profound Hearing Loss, RIGHT EAR - Severe To Profound Hearing Loss. **MDCT Scan Of Temporal Bone:** 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** was done, and the Impression was bilateral CP angle cisterns appears 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 the course and calibre (approximately 1 to 1.2mm). Limited MRI Study Of Brain Does Not Reveal Any Significant Structural or Parenchymal Abnormalities, Bilateral CP Angle Cisterns, Internal Auditory Canals, And Inner Ear Structures Appears Normal; Bilateral Cochlear Nerves Appear Normal.**Tympanometry** done s/o- acoustic reflex was absent**, DPOAE TEST** wasdone Impression - bilateral s/o OCH dysfunction, **USG** abdomen: Impression was USG of the abdomen is within normal limits, **Children Expectations Questionnaire** was done on, Paediatric Neurology Callby a doctor - can consider cochlear implant procedure as planned, Paediatrician Call:I/V/O fitness for surgery advice: a patient can be taken for surgery under due risk of anaesthesia, Anaesthetist Call was donefitness for surgery.
Diagnostic challenges: No challenges during diagnostic evaluation.
Diagnosis: After the physical examination and investigation, a doctor diagnosed a Bilateral Congenital Profound Sensorineural Hearing Loss with developmental delays.

Prognosis:-prognosis of this case was satisfied.

Therapeutic Interventions:
Medication was given to a child as per the doctor's order, DNS 400ml intravenous fluid with 4ml Inj KCL given 8 Hourly, Inj Cefuroxime 700 Mg Bd, Inj Pan 15mg Od, Inj. Emset 1.5 Mg Tds, Inj. Neomol 20ml Tds, Inj. Dexa 2 Mg Bd, SyrupIbugesic Plus 7ml Tds ,Syrup. Taxol DX 3.5 MlTds, Syrup Sinarest 1 Tsp, Nasal dropOtrivin (P) 2 Drops Tds.
Follow-up and outcome: patient incision site was healed. Doctor Advice the mother to avoid sudden head movement of child. Take care of incision site clean daily and pat dry. Give a healthy balanced diet, do regular health check-ups, maintain personal hygiene, and give proper medication as per the doctor's order.

Discussion:
A 2.5-year female child was admitted to the ENT ward with a complaint of inability to hear in both ears and inability to speak, static, noticed when a child responds to only loud sounds with developmental delay. Doctor carried out all necessary and routing investigation, and the case of Bilateral Congenital Profound Sensorineural Hearing Loss with Developmental Delay was diagnosed with Cochlear Implant Surgery. After treatment child's condition improved. Doctor allowed for discharge. [13-24]
The study is one of the first to look into the developmental outcomes of young a child who are hard of hearing (as opposed to deaf), who were detected early, and for whom intervention services (including amplification) were started soon after hearing loss was officially verified.[11]
In the Netherlands, 49 per cent of people with severe and profound hearing impairment have inherited causes for their permanent hearing loss. With different populations, inherited factors may affect permanent hearing loss differently.[12]Children with parental consanguinity have a higher hearing loss, according to Bener et al.[25-30]
Compared to adults, kids are more flexible and more capable of learning. As a result, a cochlear implant can help them more. When compared to children who simply have acoustic hearing aids, cochlear implant users' vocal quality and speech intelligibility appear to be better.[31-34]
Some researchers found no changes in language outcomes for very young infants with hearing loss detected early (6 months of age). These findings suggest that toddlers with lesser hearing loss who are discovered early and get intervention programmes can acquire developmentally good language skills between 12 and 18 months.[35]

Conclusion:
A type of hearing loss that is present from birth is called congenital hearing loss. Early discovery, timely diagnosis, and appropriate treatment can prevent delays in a child's capacity to communicate and adapt to their surroundings. It can be caused by genetic or environmental risk factors. We discuss a 2.5-year-old female child with bilateral profound sensorineural hearing loss and developmental delays in this case report. The child's condition became better after getting a cochlear implant.
Informed consent: His parents were informed before taking on this case.
Conflict of Interest: No conflict of Interest.
Funding: Datta Meghe Institute of Medical Sciences,Wardha.

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