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Hearing Loss in Leukodystrophy

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Abstract: ***Aim: This study aims to emphasize the role of audiologist in audiological diagnosing and providing audiological rehabilitative management for person identified with Leukodystrophy. Design: A single case study. Method: Audiological assessment is done using Pure Tone Audiometry, Immitance, Audiometry, Oto-Acoustic Emissions, Auditory Brainstem Response. Result: The child is identified to have almost all features of Leukodystrophy such as severe to profound hearing loss , delay in speech and language development, walking difficulties resulting from spasticity.***

Keywords: Leukodystrophy, Myelin sheath, Central Nervous System

1. Introduction

This study aims to emphasize the role of audiologist in audiological diagnosing and providingaudiological rehabilitative management for person identified withLeukodystrophy. It is also called as hereditary white matter disorders, inherited leukoencephalopaties. Congenital/ hereditary hearing loss are increasing day by day. Leukodystrophiesare a group of rare, progressive, metabolic, genetic diseases that affect the brain, spinal cord often the peripheral nerves [2,3]. There are several types ofLeukodystropy caused by specific gene abnormality that leads to abnormal development or destruction of white matter (myelin sheath) of the brain. It is an autosomal recessive disorder and may also be due to spontaneous mutation. Leukodystrophies is found in the ratio of 1:7663.

It is characterised by muscle rigidity, decrease or loss in hearing and vision, cognitive deterioration, epilepsy is common. In children spastic activity often proceeds progressive ataxia. They may have weakness in deglutition. Here we describe complete audiological assessment and rehabilitative management of a nine years with Leukodystrophy.

2. Method

A case of nine years old female child came with a complaint of not responding to soft and loud sounds and inadequate speech and language skills. She was identified as hearing impaired at four years of age . On audiological assessment the child has no significant issues in pre and perinatal history. Radiological findings (Magnetic Resonance Imaging) revealed Leukodystrophy with bilateral temporal lobe cyst. Barkovich classification of Leukodystrophy. The child has no vision problem. The child had difficulty in walking reported by neurology department.

On otological evaluation both tympanic membrane were found to be intact. On detailed audiological evaluation audiometry report revealed bilateral total hearing loss. Immittance audiometry showed “A” type tympanogram with absence ipsilateral and contralateral acoustic reflexes. Oto acoustic emissions were absent in all frequencies indicating inadequate functioning of outer hair cells. No peaks were observed on 90dBnHL for click stimulus and with repetition rate of 21.1 clicks/sec on Auditory Brainstem Response, indicating severe to profound hearing loss. In Scales for Early Communication Skill the child’s Combined Receptive and Expressive Language Age was below 2 to2.11 years. The child comprehends common lexical categories, commands through gestures. The child vocalize occasionally while requesting.

3. Result and Discussion

The child is identified to have almost all features of Leukodystrophy such as severe to profound hearing loss, delay in speech development, walking difficulties resulting from spasticity. Strong class Behind The Earhearing aid is prescribed to the child to get awareness from environment. Counselled about Cochlear implant and its realist expectations tothe parents for better speech and language development [1] and the importance of auditory verbal therapy after cochlear implant surgery was explained for better child's communication.

4. Conclusion

Leukodystrophies, while primarily affecting the CNS have a varied range of presentations with symptoms at any age. As an audiologist holistic assessment of the child’s hearing loss is important to rule out various unseen aetiologies such as demyelination of white matter for better intervention and welfare of the patient. Multidisciplinary approach brings forth better diagnosis and better intervention.

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