Case Study

Auditory Stimulation in Auditory Maturation Delay: A Case Study

Mathew Nishanth A[1], Steven Michael J[2], Anusha Rao S[3], Jaya Lakshmi P B[4]

[1,3,4]Assistant Professor, MERF Institute of Speech and Hearing (P) Ltd, Chennai, India, [2]Undergraduate student, MERF Institute of Speech and Hearing (P) Ltd, Chennai, India.

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Abstract

Introduction: Auditory deficiencies have profound implications for the development of communication abilities, social exchange, and cognitive skills. Although advances in technological instrumentation have decreased the age at which it is possible to detect auditory deficits, an absence of electrophysiological or behavioural reaction to sound does not always reflect auditory loss in younger children.

Aim of the study: To profile audiological findings of a child with auditory maturation delay associated with Chudley McCullough Syndrome post auditory stimulation.

Method: An extensive audiological evaluation, comprising Behavioral Observation Audiometry, Immittance Audiometry, Oto- Acoustic emission, Auditory Brainstem Response, and Cortical Auditory Evoked Potentials, was performed on a 6-month-old infant. After the audiological evaluation, the infant received auditory stimulation for two months. A comparable audiological assessment was conducted after auditory training.

Results: The infant was diagnosed to have severe to profound hearing loss in both ears with noticeable cochlear microphonics in ABR on pre stimulation testing. As a result, he was recommended and had undergone auditory stimulation for two months. Audiological testing post auditory stimulation revealed significant improvements in auditory responses and noticeable betterment in auditory thresholds.

Conclusion: Detailed audiological test battery is necessary in determining auditory maturation delay. Intensive auditory stimulation should be provided before concluding on the auditory management.

Key words: Auditory maturation, Auditory stimulation, Chudley McCullough Syndrome, Maturation delay.

Introduction

The central auditory system is immature at birth. The period of greatest neuronal maturation occurs until the first two years of life, leading the brainstem maturation[1]. The first year of life is critical for child development which includes the acquisition and development of language, since it is intrinsically related to the auditory nervous system maturation. The pediatric audiological assessment must be

Corresponding Author: Mathew Nishanth A, Assistant Professor, MERF-Institute of Speech & Hearing (P) Ltd, No.172/212, Royapettah high road, Royapettah Chennai, Tamil Nadu, India.

E-mail: mnishanth001@gmail.com

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carried out differently from adults. The audiologist has to give more attention to each phase of the child's maturation and development[2]. Bilateral Severe to profound sensorineural hearing loss(SNHL) is present in 1 to 3 out of 1000 newborns and 2 to 4 out of 100 infants who require neonatal intensive care [3]. To avoid significant delay of speech and language acquisition as well as auditory maturation followed by undetected congenital hearing loss (HL), an early identification and intervention is required. The absence of response may result from compromised auditory reception or processing. Thus, delayed maturation of the auditory pathway should be considered in the differential diagnosis of young children with sensorineural hearing loss[4]. Although advances in technological instrumentation have decreased the age at which it is possible to detect auditory deficits, an absence of electrophysiological or behavioral reaction to sound does not always reflect auditory loss. The ability to obtain reliable behavioral responses in infants is useful clinically. Even though reliable response of behavioral audiometry are recognized in infants from 5 to 6 months onwards, this method of exploring hearing is often considered to be unfeasible or unreliable below this age. Maturational changes of the auditory system from the periphery to the brainstem level are clearly seen in the Auditory Brainstem Response test[5]. The click-evoked ABR provides reliable estimates of the behavioral pure-tone thresholds in the frequency range 2–4 KHz. Hence this study aims to profile audiological findings of a child with auditory maturation delay associated with Chudley McCullough Syndrome post auditory stimulation.

Case Report

A 6 month old infant was brought to the Department of Audiology with the complaint of not responding to sounds. Detailed interview of the parents on the birth and developmental history revealed that the child had birth asphyxia and water accumulation in lungs and was kept in neonatal intensive care unit (NICU) for 20 days. Motor milestones development was reported to be delayed and partial ocular albinism in vision was reported. The child was subjected to a detailed audiological test battery in which Behavioral Observation Audiometry (BOA) revealed Severe hearing loss in both ears. 'B' type tympanogram was obtained in both ears due to middle ear effusion. Distortion product Otoacoustic emission (DPOAE) testing revealed outer hair cells dysfunction. Auditory Brainstem Responses revealed absence of wave V in low rate (11.1/s) using rarefaction, condensation and alternating polarities (Figure 1). The child was recommended to the otolaryngologist for further management related to middle ear effusion and was recommended for audiological follow up after one month.

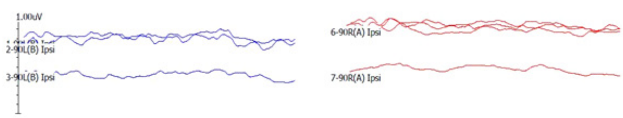

Fig 1 shows the Auditory Brainstem Response of the child

After 1 month, the child was followed up with detailed audiological evaluation. As reported by the parents, the child was under medical management for middle ear effusion and they had also undergone radiological assessment and genetic testing as referred by the otolaryngologist. Radiological evaluation reports revealed "diffuse hypomyelination & diffuse corpus callosal hypogenesis and simplified gyral pattern". Genetic testing revealed abnormal mutation of GPSM 2 genes which indicates the presence of Chudley-McCullough syndrome. Audiological testing revealed Severe hearing loss in both ears using BOA and A type tympanogram in both ears using tympanometry. DPOAEs were performed which revealed partial outer hair cells functioning. Electrophysiological test findings revealed presence of clear and replicable ringing cochlear microphonics with no replicable peak in low rate (11.1/s) using rarefaction, condensation and alternating polarities (Figure 2). Hence the child was given a diagnosis of Severe to Profound hearing loss with (?)Auditory Dyssynchrony. Although ABR testing indicated severe to profound hearing loss, unaided Cortical Auditory Evoked Potential testing revealed presence of P1-N1 complex in both ears (Figure 3). Based on the current findings, the child was recommended for an intensive auditory stimulation therapy with monthly monitoring of auditory thresholds.

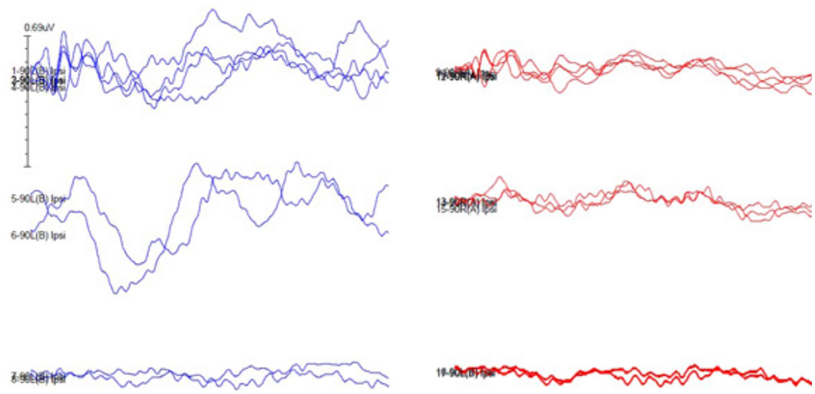

Fig 2 ABR in the 1st follow up evaluation

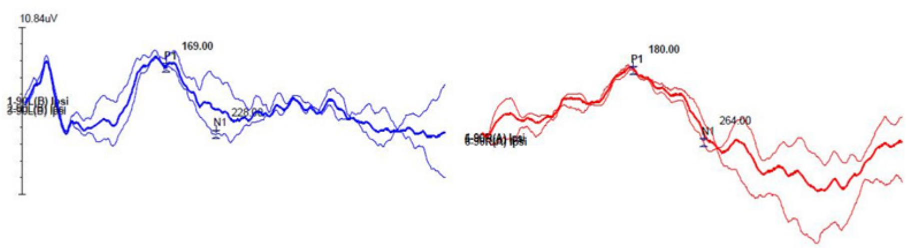

Fig 3 shows the CAEP responses of the child

The child was enrolled in auditory stimulation therapy. The child was provided with 2 sessions of therapy per week with 45 minutes each. The parents were also counseled and were trained in providing similar auditory stimulation at home intensively. The goals for the session were to improve detection of environmental sounds, ling sounds, LTL sounds and localisation of sounds with varying vocal intensities based on the child's response. Informal observations post 1 month of therapy showed significant progress from the child. He was able to localize sounds although not accurately but searching for sounds were present. He was able to detect ling sounds with 75% accuracy even with soft vocal loudness. The child was also subjected for electrophysiological testing to objectively profile on the progress. ABR testing revealed presence of peak V until 30 dBnHL in the right ear and 70 dBnHL in the left ear indicating normal hearing sensitivity in the right ear and moderate hearing loss in the left ear. Clear and replicable ringing cochlear microphonics was also observed in both ears (Figure 4). Cortical Auditory Evoked Potential revealed presence of both P1-N1 and P2-N2 complex in left ear and P1-N1 complex in right ear (Figure 5). The child was recommended to continue auditory stimulation therapy and the parents were counseled on intensive auditory stimulation at home.

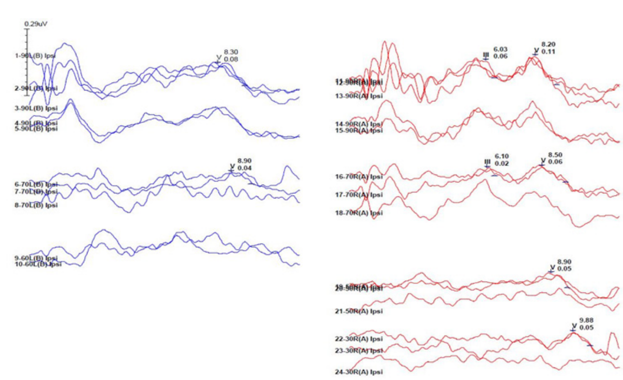

Fig 4 shows the ABR after providing auditory stimulation

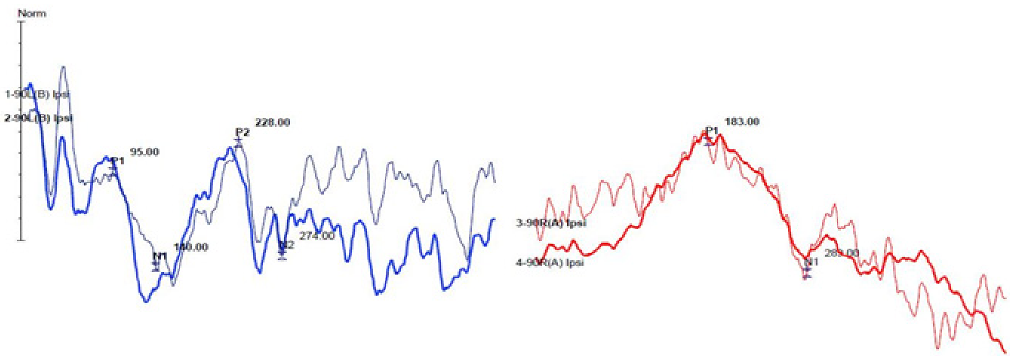

Fig 5 shows the CAEP after providing auditory stimulation

Discussion

Auditory maturation or development refers to changes in the peripheral and central nervous system along the auditory pathways [1]. This maturation occurs in response to acoustic stimulation. Auditory pathways mature until 2 years of age which can clearly be reflected in an electrophysiological testing like ABR through variations in latencies and amplitudes of ABR waveforms [4]. No auditory brainstem responses in ABR testing is considered as Severe to Profound hearing loss. Auditory deprivation can have long term detrimental effects on language and learning ability of an individual. Thus, early identification of the underlying problem is advised for appropriate early management. A comprehensive audiological test battery can help identify hearing loss. The auditory management for severe to profound hearing loss with no benefit from hearing aids is Cochlear implantation [6]. However, to rightfully identify that a severe to profound hearing loss in an objective testing like ABR can be contributed by other factors, a detailed audiological test battery with auditory follow-ups and appropriate referrals is a must.

This study presents a case of a 6 month old infant who was initially diagnosed with severe to profound hearing loss due to an early absence of observable response to auditory stimuli in behavioral and objective testing had improvement in auditory responses with auditory stimulation. An audiological test battery comprising of Behavioral observation audiometry, tympanometry, oto-acoustic emissions and electrophysiological testing (ABR) was performed on a 6 month old infant with the complaint of not responding to sounds. The behavioral and objective testing of auditory thresholds indicated Severe to Profound hearing loss. However, tympanometry results suggested indication of middle ear pathology which could have contributed to absent DPOAEs in the physiological testing. Hence, Hearing aid trial was not indicated and the family was advised for a follow up audiological visit for a re-evaluation after medical management for middle ear effusion. The child was also diagnosed with Chudley McCullough Syndrome through genetic testing. This syndrome is characterized by early-onset sensorineural hearing loss with specific brain anomalies on radiological evaluation including corpus callosal hypogenesis[7] After 1 month of medical management, a repeat tympanometry suggested no indication of middle ear pathology. Test results of a follow up ABR revealed no replicable ABR peaks with noticeable cochlear microphonics which is a clear indicator of OHC integrity and susceptible Auditory dys-synchrony. Hence, CAEPs which is an objective measure to quantify auditory pathway maturation were performed to confirm the presence of auditory responses. The presence of the P1-N1 complex in CAEPs suggested the presence of auditory responses. However the latencies were prolonged which suggested a delay in central auditory pathway maturation. A delay in the auditory maturation can be contributed by several high risk factors such as preterm birth, low birth weight, perinatal anoxia, post natal seizure and infection [8]. In this case, high risk registers such as birth asphyxia, lung infection along with hypomyelination and corpus callosal hypogenesis which is indicated in radiological evaluation could be the contributing factors to auditory maturational delay. A delay in auditory maturation can often result in abnormal or absent

ABR waveforms which need not indicate a severe to profound hearing loss. Hence, a comprehensive test battery should include appropriate additional tests when indicated with follow up visits to confirm diagnosis. The child was advised to undergo auditory training. Upon intensive acoustic stimulation, observable subjective and objective auditory responses were noted with 1 month of auditory habilitation. Subjective responses include detection of acoustic stimuli presented at soft to conversational levels through behavioral responses. Objective responses include presence of ABR peaks with noticeable cochlear microphonics in ABR testing indicating normal hearing sensitivity in the right ear and moderate hearing loss in the left ear and presence of replicable peaks in CAEPs with an early latency shift in the left ear. These results suggest that acoustic stimulation can aid in auditory maturation which is reciprocated through electrophysiological testing. Thus, this case study highlights the importance of a detailed audiological test battery with inclusion of appropriate tests when indicated; correlation of test results; appropriate referrals and communication with interdisciplinary teams; advising auditory training and follow-up visits to confirm diagnosis before deciding on auditory management.

Conclusion

Even though congenital deafness is a characteristic of Chudley McCullough Syndrome, through appropriate & detailed audiological test battery an auditory maturation delay was identified resulting in early auditory stimulation and improvement in auditory responses.In summary, when suspecting auditory maturation delay it is advised to suggest routine auditory follow ups with auditory training to conclude on management.

This suggests a short period of postponement to confirm diagnosis is better than misdiagnosis which can result in inappropriate management.

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Ethicalclearance: Informed consent was obtained from the parents

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