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Case Report

Auditory Brainstem Response Characteristics in Spastic Diplegic Cerebral Palsy Secondary to Periventricular Leukomalacia - A Single Case Illustration

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ABSTRACT

Auditory behaviours and objective audiological findings have limited clarity to explain the underlying pathological conditions in children with periventricular leukomalacia (PVL). The purpose of this study was to examine auditory brainstem response (ABR) characteristics in a child with spastic cerebral palsy (CP) secondary to PVL and to discuss correlation between behavioural observation audiometry (BOA) and ABR findings. Audiological evaluations consisted of BOA, immittance audiometry, ABR and cochlear microphonics followed by a detailed case history. Informal observation of auditory behaviours was also carried out with verbal and non-verbal stimuli. Behavioural responses in terms of informal and BOA results found to be correlated well nevertheless, ABR findings indicated poor correlation with behavioural responses. In the test results, it had been evident that ABR findings were over estimating the actual hearing thresholds of the child. The neurological conditions underlying CP will affect myelination process of the central and peripheral nervous system. The changes in myelination process and altered neural firing may have disturbed the ABR finding. Disorders affecting neural maturation and synchronized firing are among the common neurological impairment seen in children with CP and there for the pathological conditions alter their responses in multiple ways.

Key words: ABR, Periventricular leukomalacia, Cerebral Palsy, Behavioural Observation Audiometry, Cochlear Microphonics

INTRODUCTION

Periventricular leukomalacia (PVL) is a type of brain injury which involves death of brain tissues surrounds the ventricles. PVL is commonly seen in premature babies as ischemic brain injury. [1] The ischemia occurs in white matter (WM) adjacent to the lateral ventricles lead to deep white matter injury which most commonly results from insults occurring between 24 to 34 weeks of gestation. During this period WM is more susceptible to injury as it attempts to complete the complicated and dynamic process of maturation. WM plays a significant role in cortical organization and neural networking which directs long term neurocognitive outcome.

Parrot explained that periventricular lesions as infarcts which seen in the lateral corner of periventricular WM. [2] The term 'periventricular leukomalacia' was first used

by Banker and Larroche in 1962 and described the neuro pathological changes that corresponded to coagulation necrosis and defined clinic pathological correlations. [3] The neuropathology of PVL consists of two principal components: focal component is characterized by localized necrosis of all cellular elements with subsequent cyst formation. It may disrupt remote pathways or interfere with differentiation and maturation. But diffuse component is less severe and cell-specific, which categorized by diffuse injury to oligodendroglia (OL) precursors. It designed to develop as mature OLs to form myelin of the cerebral WM. PVL not only affect the myelination process of CNS but it also affect the myelination process of PNS. It is a prototypical cause for cerebral palsy (CP) and babies with PVL typically do not show any obvious symptoms initially. But they are at risk for delayed motor development, delayed mental development, coordination problems and visual and hearing impairments. It is the main cause and approximately 60-100% of infants with PVL are diagnosed with CP. [4] Spastic diplegia is the most common form of CP following mild periventricular leukomalacia. Severe periventricular leukomalacia is frequently associated with quadriplegia. [5]

In children with CP, hearing assessment is having a significant role and it is important to plan further rehabilitation process. Auditory brainstem responses (ABR) are an important tool to assess auditory nerve function in terms of threshold estimation in children with multiple impairment, to assess functional integrity of auditory system and for neurophysiological examination. Hence it is used for children with CP to check their auditory functions. ABR characteristic is not respectable in terms of morphology, repeatability and latency in children with CP, even if they have normal hearing sensitivity. This pin point towards underlying neurological problem in children with CP. [6] Currently available audiological tests methods both behavioural and non-behavioural tests and its correlation are important for audiological interpretation. Accurate responses of Behavioural Observation Audiometry (BOA) provides the estimate of entire auditory system [7] and children with multiple impairment who cannot do conditioned play audiometry also relay on BOA to find the correlation of subjective and objective audiological tests. Hence both of these audiological tests BOA and ABR are commonly used to elicit auditory responses of children with CP.

ABR findings significantly differ in children with spastic CP as compared to typically developing children. Depressed auditory wave form responses indicate decreased or alter neural firing or synchronization in the auditory brainstem. [8] As mentioned in literature ABR abnormalities, depressed auditory wave responses and auditory neuropathy spectrum disorders (ANSD) were discussed in children with CP devoid of considering the causes of CP. CP can occur due to several reasons and PVL is one of the main causes which can lead to mild to severe issues in the neuronal level. However, significant discussion about ABR characteristics in particular to PVL is limited in literature. Even if abnormalities can expect in ABR wave forms, how it will affect the threshold estimation and at what extent variation can expect in its wave characteristics need to be investigated; as PVL interrupt myelination process and neural network of both central as well as peripheral nervous system. In the same time the auditory behaviours and audiological findings in PVL need to be specified. The purpose of this case study was to examine ABR characteristics in a child with spastic CP secondary to PVL and to discuss correlation between BOA and ABR results.

METHOD

A female child, 6 years 1 month with spastic diplegic cerebral palsy reported to our clinic for ruling out hearing loss. History of the child comprised of low birth weight of 910 gm, prematurity (26 weeks),

delayed developmental milestones and inadequate speech and language skills. Neonatal jaundice and respiratory distress soon after birth were remarked on the evaluation. Hence, child was kept in neonatal intensive care unit for three days for phototherapy and for ventilation. MRI was done and the result reveals bilateral periventricular white matter lesions suggestive of periventricular leukomalacia. Child's speech and language characteristics milestones found to be delayed. Child started to speak at 1year 4 months of age and two word phrases at 4 years of age. Currently child is having verbal communication in three to four word sentences. Child is undergoing for both physiotherapy and speech therapy since 5 years on regular basis.

Procedure:

Detailed case history of the child was taken and observation sessions carried out informally to check auditory response for different auditory stimulus were as name call, conversational speech (simple and familiar questions and statements) and nonverbal stimulus. Audiological tests were performed which comprised BOA, impedance audiometry, Cochlear Microphonics (CM) and ABR. Conditioned play audiometry was tried but the child was not performed well to avoid false positive responses. The child was reluctant to wear headphones to complete test frequencies of conditioned play audiometry. Hence, BOA with Narrow Band Noise (NBN), warble tone and Sound Awareness Threshold (SAT) were done using Grason-Stadler Inc (GSI), AudioStar Pro two channel audiometer. Stimulus were introduced using speakers which were placed at 45 degree angle (free field audiometry) and stimulus frequencies were at 250Hz, 500Hz, 1 KHz, 2 KHz & 4KHz. BOA was performed twice to avoid inconclusive responses. GSI, TympStar was used to perform immittance audiometry. Tympanometry was carried out using probe tone of 226 Hz and reflexometry was done using activator tones of frequency 500Hz, 1KHz, 2KHz and 4KHz. Admittance of minimum .03ml was considered as reflex for both ears. ABR was done using Intelligent Hearing systems (IHS), version 5.33. It was elicited with acoustic click stimuli at the rate of 33.1/second and at 11.1/second with 2000 sweeps for each repetition by using insert form tip. Before placing four surface electrode, skin was cleaned thoroughly to develop impedance <5 kΩ with no more than 3kΩ difference between the electrodes. Monaural recording with rarefaction polarity was used to carry out the test. Cochlear microphonics was also performed at 88dBnHL with the polarity of condensation and rarefaction for both ears at 33.1/second using the same instrument, IHS.

RESULT

Parents were not suspected hearing loss as the child had better responses to speech and to other auditory stimuli in day to day situations. BOA responses to warble tone and narrow band noise were elicited at around 45- 50 dBHL for the frequencies 250Hz, 500Hz, 1KHz, 2KHz and 4KHz and at 50-55 dBHL for speech awareness threshold. Observation session which was carried out informally with verbal and non-verbal stimuli at normal conversational level also correlated with behavioural responses. The child responded to name call and familiar conversational speech at moderate and moderately loud level by head turn, eye movement and by responding to speech. But auditory responses were not observed below 40 dBHL and at soft level verbal stimuli for both BOA and informal observation respectively. 'A' type tympanogram and reflexes below .03ml were obtained bilaterally for immittance audiometry. ABR with click stimuli showed no significant V peak at 88 dBnHL bilaterally at two different rates, 11.1/second and 33.1/second.

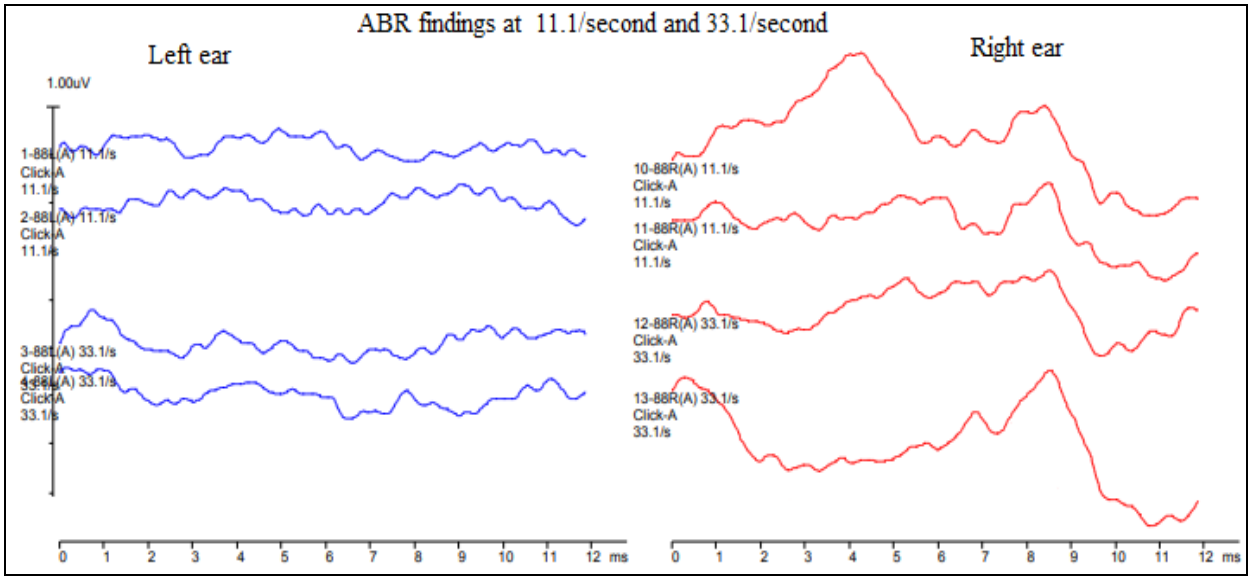

Figure: 1 shows both right and left ear ABR findings. First two wave form of right and left ears done at 11.1/second and last two wave form done at 33.1/second. No definable V peaks identified at 88 dBnHL within the latency of 5 to 8 milliseconds.

As prolonged latencies were expected in children with CP, latency of 5 -8 milliseconds considered to mark the V peak at 88 dBnHL. Polarity differences with condensation and rarefaction could not made noticeable CM at 88 dBnHL bilaterally.

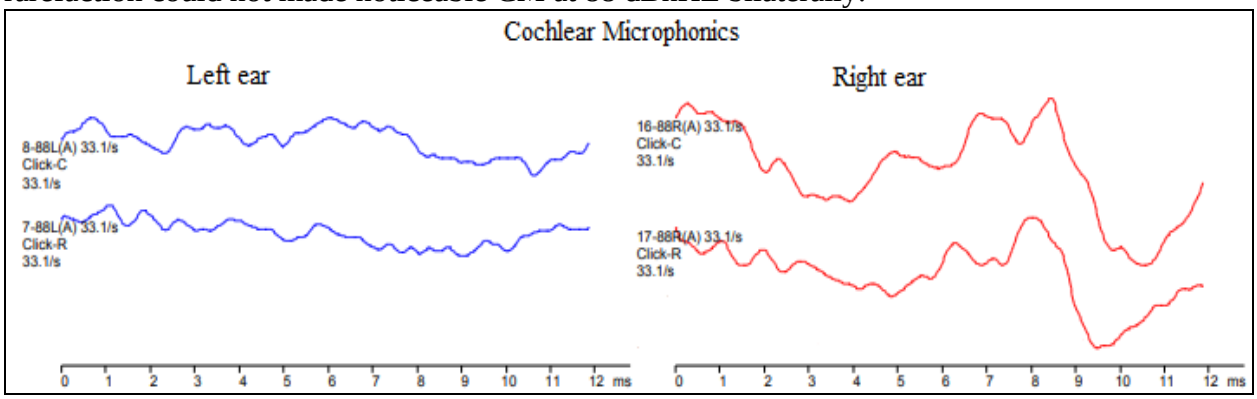

Figure: 2 shows cochlear microphonics of right and left ear at a rate of 33.1/second. There was no significant CM identified within 1millisecond in both ears.

The audiological results of this child indicated poor correlation among BOA and ABR. Since ABR responses are depicted to auditory nerves, structural and functional variation of auditory nerves can affect its results.

DISCUSSION

Usually ABR measures showed statistically significant difference in children with CP as compared with typically developing children due to neurological abnormalities. Increased threshold, reduced wave V amplitude, decreased V/I amplitude ratio and prolonged I-V interval were seen in children with neuro developmental deficits [9] as compared with typically developing children. But in this child ABR wave patterns indicate absence of definable V peak bilaterally at 88 dBnHL even though the child had better behaviour responses. Myelination process in the auditory pathway of human brainstem occurs between 26th and 29th weeks of gestation. Subsequent to the 29th gestational week, density of myelination increases in all pathways until at least 1 year postnatal age. The 26th to 28th gestational weeks are critical period in the onset of human central auditory function. [10] Prematurity is responsible for complex changes in brain development because it affects the process of

myelinogenesis, cortical development and neural migration. These changes stem from the destruction or inadequate development of the oligodendrocytes which leads to an impaired development of cortical region. [11] As a result of damage to precursors of OL, there are chances for myelination problems in CNS.

The changes also affect the myelination of peripheral nervous system (PNS). As auditory nerve is the part of PNS, Schwann cells are responsible for the myelination process. [12] Peripheral myelination process involves recognition of the target axon, adhesion of glial process to the axon, synthesis and transportation of myelin component, enwrapping of the plasma membrane around the axon, continuous growth of glial cytoplasmic process and formation of compact myelin layer and nodes of Ranvier. It is a step by step process; disruption in any of these steps which can lead to neurological problem like CP. [13-14] The disruption of myelination process can affect the myelination of auditory nerve. Synchronised firing of auditory system will be get affected or delayed due to myelination problem. This can be a reason for absence of definable ABR peaks in this child.

Child showed better responses to behavioural stimuli while doing BOA rather than ABR findings. A poor correlation between BOA and ABR findings is evident in the audiological profile of the child. ANSD conditions generally show better behavioural responses, presence of Otoacoustic emissions/CM and absence or severely affected ABR peaks. In this case, CM was found to be absent bilaterally and absence of ABR peaks at higher intensity level; but consistent BOA responses were present at moderate level. Hence the audiological profile of the current case was met the ANSD criteria. The behaviour responses were correlating with speech and language skills which also not limited to explain severe to profound degree of hearing loss. The ABR wave patterns may be exaggerating the threshold level due to underlying neurological conditions and myelination problem. As it is a neurological impairment, higher cortical potentials can show a limited response due to periventricular lesions affects thalamus and midbrain areas. Disorders affecting neural maturation and synchronised firing are among the more common neurological impairment seen in children with CP and there for the pathological conditions alter their responses in multiple ways. Marked difference of ABR characteristics in spastic cerebral palsy positively associated with adverse neurological conditions. [6] Hence in this case ABR response may be undetectable due to limited synchronization of the auditory nerve.

CONCLUSION

PVL lead to spastic diplegic and quadriplegic CP and severe lesions may develop greater motor difficulties as well as associated issues. Adverse neurological conditions and underlying pathologies can exaggerate the response characteristics of CP, in particular to auditory responses. As ABR is a good tool to assess the functional integrity of the auditory system, abnormal neurological conditions undesirably affect the ABR wave characteristics. From the clinical observations it has to be understood that PVL can have an influence on audiological findings of the present case. Poor correlation between BOA and ABR characteristics in this child may be due to difference in the pathways which sub servers the test procedures. [15] Hence, the case report illustrates importance of both behavioural and electrophysiological test while performing audiological evaluations in children with PVL for better rehabilitation and prognosis; rather than confining tests to electrophysiological test due to restricted motor movements, developmental delay and difficulty to confirm behavioural responses in children with CP.

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