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West Syndrome: A Case Study Through Audiological Perspective

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ABSTRACT

West Syndrome is a disorder with combination of symptoms such as infantile spasm along with its typical age of onset and specific kind of electroencephalogram pattern termed as hypsarrhythmia Here in this article I report a case of West syndrome with left hemi paresis, diagnosed in a 2.5 year old baby boy

Key words: West Syndrome, Hearing Evaluation, Auditory brainstem response

INTRODUCTION

The disorder was first described by William James West in the year 1841 and thereby named as "West Syndrome" (WS). Thereafter evolution of nomenclature was witnessed by the literature, leading to periodical up-gradation of the terminology to infantile spasm in conjunction with the most relevant clinical presentation of the disorder. Again the terminology had an update and was termed as epileptic spasm considering the onset of the disorder even after infancy [1]. Now the umbrella term Infantile Spasm syndrome is used to collectively mention these groups of disorders commonly exhibiting EEG (Electroencephalography) abnormalities, cognitive deficits and other neurodevelopment delays [3].

West syndrome is a rare neurological syndrome that can affect males and females. The X-linked form of West syndrome affects males more often than females. It has been estimated to affect 0.31 per 1000 live births in the United States. West syndrome accounts for approximately 30 percent of all cases of epilepsy affecting infants [2].

According to an article on WS from Northern India, 10.1 % (15 out of 148) of children were identified with hearing impairment, 62.2 % with spastic cerebral palsy, 64.9 % with microcephaly and a male preponderance accounting to 81.1 %. It is a difficult-to-treat disorder characterized by poor response to antiepileptic drugs and consequent intellectual disability [4]

Adrenocorticotropic hormone (ACTH), Prednisolone, vigabatrin, sodium valproate, clonazepam and. levetiracetam and modified Atkins diet are common drugs used to pharmacologically treat spasm in patients with West syndrome. These drugs were found to have variable results ranging from cessation of spasm to partial improvement and even no improvement [5]. Hypsarrhythmia [4, 5] and, modified hypsarrhythmia [5] of was also reported in the electroencephalogram (EEG) finding in in children diagnosed with West syndrome.

EEG showing high voltage chaotic slow waves intermixed with spike and sharp wave discharges are termed as hypsarrhythmia whereas, increased interhemispheric synchronization-consistent voltage asymmetries; consistent focus of abnormal discharge; episodes of generalized/regional or lateralized voltage

attenuation; or primarily high-voltage bilaterally asynchronous slow wave activity are the characteristics reported for modified hypsarrhythmia [6].

High rates of comorbidities such as cerebral palsy, microcephaly, vision and hearing impairment, and feeding difficulties as sequelae of perinatal insults were also found as additional problems in children with WS [4].

Perinatal asphyxia which is a known predictor for hearing loss in neonate babies is often a know etiology for WS as well [7]. Other such known causes of West Syndrome which are also included as risk factor for hearing loss are meningitis, hypoglycaemia and prematurity [4]. But a good number of children are found to have no known etiology.

These etiological data also put forward the inferences that the children with West Syndrome have high probabilities to manifest motor milestone lags and intellectual disability of varying degrees. Thus the speech, language and auditory development can be compromised in cases with WS leading to spoken language disorder which can be secondary to multiple factors such as impairment, intellectual disability, seizure disorders and/or cerebral palsy.

CASE PRESENTATION

Background Information

A 2 year 5 month old male child was bought to the department with suspicion of speech, language and motor milestone developmental delays. The child is a known case of West Syndrome with left hemi paresis as reported by Geneticist from a private hospital. After otoscopic examination the child was found to have intact ear drums in both ears, and normal structural oral peripheral mechanism.

Birth History

No significant pre-natal, peri-natal or antenatal history reported.

Radio-logical evaluation:

The MRI reports revealed No abnormality of brain and inner ear, bilateral vestibular cochlear nerve bundle normal.

Audiological evaluation

Test battery approach was used for the audiological evaluation of the child. The routine protocol of test battery was used for the detailed audiological evaluation and the tests included Conditioned Play Audiometry, Immittance audiometry, Otoacoustic emission testing and, auditory brainstem response evaluation

Instruments used

Periodically calibrated audiological instruments were used. The conditioned play audiometry was done using Piano Inventis dual channel audiometer in free field mode using loudspeakers. Immittance audiometry was done using the GSI-Tympstar Middle ear Analyzer. Otoacoustic emission testing was carried out using Interacoustic Titan and auditory brainstem response evaluation was done using Biologic Navigator Pro with ER-3 insert ear phones.

RESULTS OF THE AUDIOLOGICAL EVALUATION

Conditioned Play Audiometry

Could not be completed as the child was not cooperative while during the conditioning phase of the testing.

Immittance audiometry

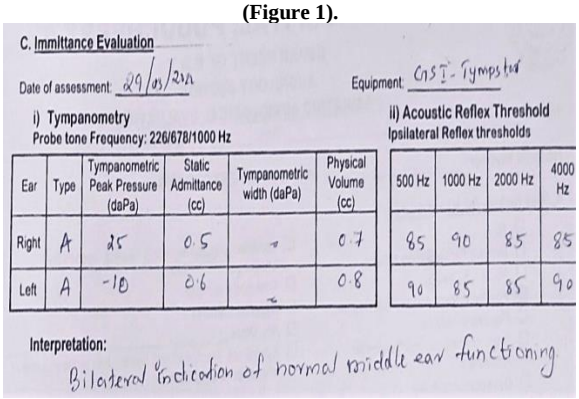
Child was sedated during the testing

Findings: Both ears: “A” TYPE tympanogram with reflexes present.

Impression

Both ears: Indication of normal middle ear Functioning.

(Figure 1).



C. Immittance Evaluation

Date of assessment: 29/05/22 Equipment: GSI - Tympstar

i) Tympanometry
Probe tone Frequency: 226/678/1000 Hz

Ear	Type	Tympanometric Peak Pressure (daPa)	Static Admittance (cc)	Tympanometric width (daPa)	Physical Volume (cc)
Right	A	25	0.5	-	0.7
Left	A	-10	0.6	-	0.8

ii) Acoustic Reflex Threshold
Ipsilateral Reflex thresholds

500 Hz	1000 Hz	2000 Hz	4000 Hz
85	90	85	85
90	85	85	90

Interpretation:
Bilateral indication of normal middle ear functioning

Figure 1: Immittance Audiometry results

Oto-acoustic emission

Findings: Bilateral Transient Evoked OAE's are present.

Impression: Bilateral Indication of normal Outer hair cells function.

Auditory Brainstem Responses

Findings: Both ears: clear and identifiable ABR Ⅴth peak absent at 30dbHL for click stimuli at a rate of 21.1/s using rarefaction polarities (Figure 2).

Impression: Bilateral hearing sensitivity within normal limits.

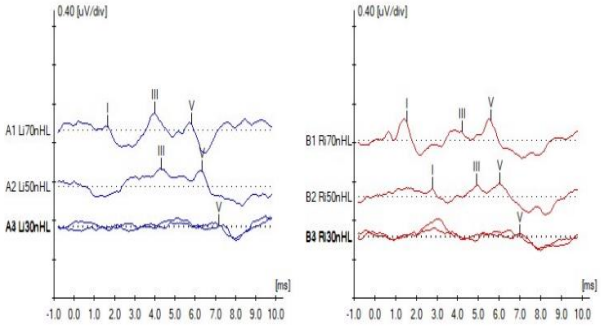


Figure 2: Auditory Brainstem Response results

Recommendation given was

1. Conditioned Play Audiometry
2. Speech and Language Intervention
3. Psychological Evaluation
4. Physiotherapy Intervention
5. Genetic Counseling
6. Follow up.

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Conflict of Interest: None

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