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Research Article

AUDIOLOGICAL TEST OUTCOMES IN GOLDENHAR SYNDROME- A CASE STUDY

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ABSTRACT

Goldenhar syndrome (GS) is a rare congenital disease present since birth. Craniofacial anomalies affecting head and face have been seen in this syndrome which also indicates the probability of presence of hearing loss. Goldenhar syndrome was diagnosed based on the sign and symptoms of the child. Audiological tests include both subjectively and objectively. Two consecutive audiological evaluations were carried out over time. Results indicate the child is having auditory neuropathy/auditory dys-synchrony in both ears. Involvement of audiologist and proper follow up programs with other professionals like pediatricians, ophthalmologist, physiotherapist etc. is must to provide better service to the client.

INTRODUCTION

Goldenhar syndrome caused due to defects occurs in the development of 1st and 2ndbranchial arches during blastogenesis [1, 2, and 3].It was first introduced by Maurice Goldenhar, the Swiss ophthalmologist in 1952 [4, 5, 6,]. Incidence and prevalence lies between 1:3500 to 1:5600 live births with male/female ratio 3:2. Mostly sporadic in nature [7, 8, 10]. Due to still unclear basis of this syndrome and also unavailability of specific diagnostic test, diagnosis is based on the clinical aspects and its unique features such as mandibular hypoplasia, vertebral anomalies, ear abnormalities, preauriculartag, and hemifacial microsomia[9].

CASE REPORT

A three years old male client presented with complain of unable to speak & hear appropriately. Prenatal history reveals that his mother had suffered from diarrhea for three months during 6th month of pregnancy. Perinatal history was normal. According to the post natal history child was suffered from weakness & malnutrition after birth. He is a full term baby with normal delivery. His birth weight was about 2.7 kg. Motor milestone development is also delayed. Some of the physical anomalies present are mandibular hypoplasia, vertebral abnormality, and facial asymmetry on the right side with malocclusion of teeth. Also having preauricular tag in front of the right ear.

Auditory stimuli for behavioral observation audiometry was presented to the child with different warble tone (500, 1000, 2000, 4000Hz) with loudspeaker placed at 45 degree. Live speech and noise like BBN and narrow band noise (500Hz to 4000Hz) was also presented along with warble tone at different intensity level. Observable response and any behavioral changes due to the given stimuli were noted.

Immitance evaluation was done using a calibrated GSI- Tymp star diagnostic Immitance middle ear analyzer to obtain acoustic reflex thresholds and tympanogram. Tympanometry was performed bilaterally using 226Hz probe tones and measurements of ipsilateral acoustic reflex thresholds up to 110 dB were attempted at 500; 1000; 2000; and 4000 Hz.

Distortion product evoked Otoacoustic emissions elicited using 80 dB SPL peak equivalent level click, present in 5 successive trials, where P1- 65 dB and P2- 55 dB was set and a signal to noise ratio of at least 6 dB with a reproducibility score of at least 75% using Oto read screening INTERACOUSTICS OAE instrument was measured, in frequency band of 2KHz to 5KHz. A single channel recording was used for ABRE measurement with vertex as non- inverting electrode. The ear lobe or mastoid

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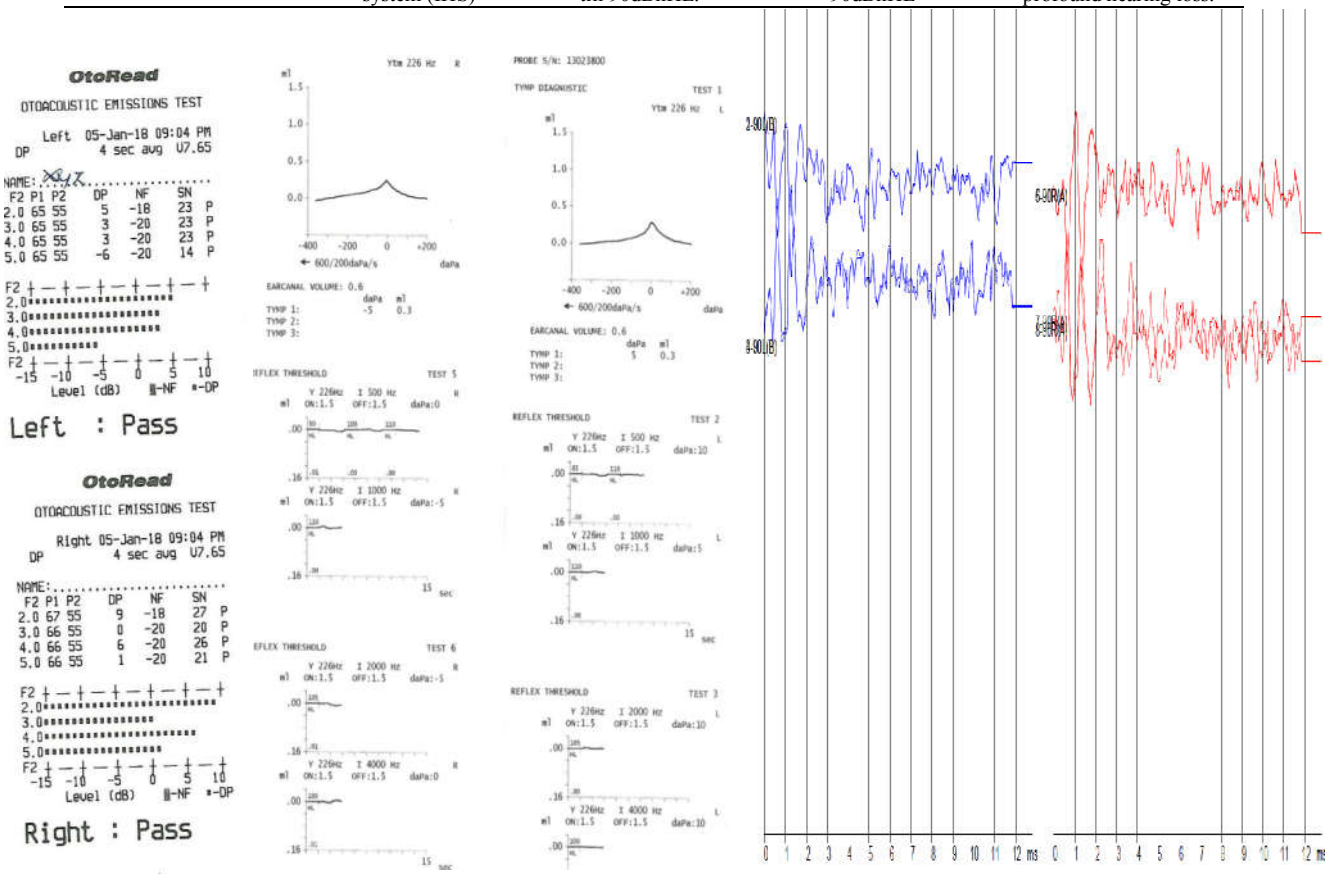
ipsilateral to the stimulus was used as inverting electrode site while the ear lobe or mastoid contralateral to the stimulus was used as ground electrode. All impedance was less than 5 KOhms. A high level ABR was obtained by presenting 90 dBnHL, 100 microsec. rarefaction clicks through insert earphones using a rate of 7.1 clicks per second. Two recordings were obtained to ensure repeatability. The recording parameters consisted of a 12ms time window. The bioelectric activity was amplified 100,000 times and filtered 300-3000 Hz. Approximately 1500 averages were obtained for each recording.

tympanogram which objectively suggest no middle ear pathology.

The present reports outline the possibility of auditory neuropathy. Though conductive and sensorineural loss has been reported in literatures but auditory neuropathy have not been reported till yet. The entire tests performed provide valuable information regarding the hearing sensitivity of the client.

Table 1 Details of test administered, instrumentation and results

Test administered	Instrument used	Right ear	Left ear	Impression
OAE	DPOAE Oto read (Inter acoustics)	PASS	PASS	Bilateral normal outer hair cells functioning
Immitance audiometry tympanometry and reflexometry	GSI- Tymp star	A type tympanogram with acoustic reflex absent.	A type tympanogram with acoustic reflex absent.	No indication of middle ear pathology.
Behavioral Observation audiometry	AC-40 interacoustics diagnostic type 2 audiometer.	Head turning response till 40 dB (no ear specific information)	Head turning response till 40 dB (no ear specific information)	? normal hearing sensitivity
BERA	Intelligent Hearing system (IHS)	No peaks observe till 90dBnHL.	No peaks observe till 90dBnHL	Bilateral severe to profound hearing loss.



RESULTS AND DISCUSSION

Screening Otoacoustic emission test revealed bilateral pass suggestive of normal outer hair cells functioning while the auditory brainstem response audiometry revealed bilateral severe to profound hearing loss, as no peak observed even after repeated trials at high intensity 90dBnHL. Next evaluation carried out was BOA evaluation, revealed observable response like head turning, stilling, sound searching were noted till 40dBHL, which is suggestive of normal hearing sensitivity. Results of impedance audiometry reveal bilateral 'A type'

We can conclude that a case with Goldenhar syndrome can also have possibilities of having not only conductive and sensorineural loss but they can also be suspected as a case of auditory neuropathy. Hence it is important to perform audiological screening tests right after birth, followed by diagnostic test batteries. So that, syndromes having hearing disorders can be detected as early as possible. Because hearing loss can also negatively affect a child's speech, Therefore Involvement of audiologists among other professionals like pediatricians, ophthalmologists, otorhinolaryngologists,

physiotherapist and other medical specialist for the diagnosis and management of this syndromes is important in terms of hearing and speech Language development and also Proper follow-up programs with this professionals are also very necessary for the betterment of the child.

This report also ensures that objective and subjective audiological tests are equally important in the clinical and prognostic evaluation.

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