

Speech Audiometry– A Reliable Pointer

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Citation

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Abstract

Auditory Neuropathy coexisting with bilateral conductive hearing loss has not been reported so far .This case report highlights how the underlying Auditory Neuropathy can be unmasked by Speech Audiometry and that subsequently changes the entire management. Unlike Pure Tone Audiometry (PTA), Speech Audiometry is not necessarily done always for all patients, especially in developing countries and we highlight the need for it to be used routinely.

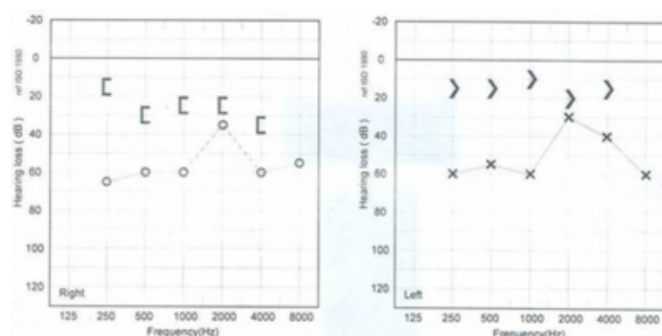
CASE REPORT

A 13 year old girl presented with complaints of bilateral hearing loss for one year. Her symptoms were slowly progressive, particularly the difficulty in understanding speech. She had associated bilateral tinnitus but there was no history of vertigo or ataxia, headaches, vomiting, loss of consciousness or seizures. Her birth and early development were normal and there was no past history of jaundice or familial hearing loss. The patient's two siblings were normal and her parents were distantly related.

Her tympanic membranes were normal and the tuning fork tests revealed bilateral conductive hearing loss. Standard blood tests were normal. Pure Tone Audiometry (PTA) showed bilateral moderate severe conductive hearing loss with mixed component at 2 kHz on the left side (a carharts notch)

Figure 1

Figure 1A &B: Pure tone audiogram of the patient's right and left ears



She had bilateral 'A' curve with reflexes absent on impedance audiometry and in speech audiometry, there was

an inability to establish speech reception threshold scores bilaterally due to very poor speech perception.

Otoacoustic Emissions (OAE) were present bilaterally and Brain Stem Evoked Response Audiometry (BERA) revealed bilateral poor wave form morphology.

Figure 2

Figure 2 A & B Distortion products (OAE) of the patient's right and left ears

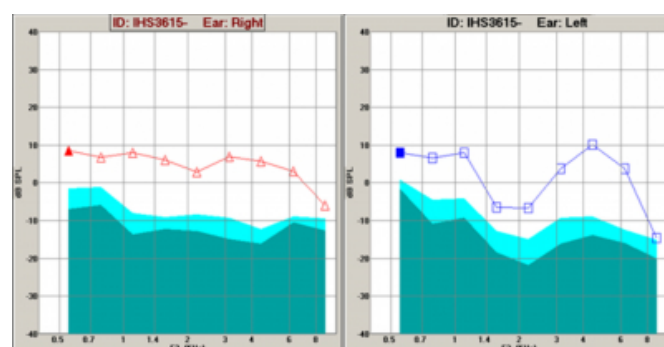


Figure 3

Figure 3: A & B Brainstem response audiometry of the patient's left and right ears



She did not benefit from hearing aids and was counselled regarding lip reading and further evaluation for implantable hearing devices. Her neurological evaluation was noted to be normal, except for the hearing loss. Magnetic resonance imaging (MRI) of the brain and nerve conduction studies

were also normal. However the visual evoked potential assessment showed bilateral optic nerve dysfunction. She was planned for cerebrospinal fluid analysis and ophthalmology evaluation. However the patient did not complete these evaluations and was thereafter lost to follow-up.

DISCUSSION

Auditory Neuropathy was first reported in the late 1970's as paradoxical findings because of a discrepancy between absent waves on BERA and the hearing thresholds on PTA. This disorder was initially referred to as central auditory dysfunction or auditory neural synchrony disorder, and it was Starr et al in 1996 who described this as a clinical entity of auditory neuropathy^[1]. These patients characteristically demonstrate hearing loss for pure tones (mild to severe sensory neural hearing loss), impaired speech discrimination out of proportion to pure tone loss^[2], absent or abnormal auditory brainstem responses, normal outer hair cell function as measured by otoacoustic emissions and cochlear microphonics^[3] and the absence of bilateral acoustic reflexes. This combination of findings suggests that cochlear function and in particular outer hair cell function is normal in these patients and also suggests that the inner hair cell / VIII nerve functional unit is abnormal^[4]. Auditory Neuropathy includes dysfunction of peripheral synaptic coding of sound by inner hair cells (synaptopathy) and/or of the generation and propagation of action potentials in the auditory nerve (neuropathy)^[5].

In sharp contrast, our patient presented with moderately severe conductive hearing loss with a dip at 2 kHz in the left ear – a typical audiogram which made the first author to suspect stapes fixation. The impedance audiometry showed bilateral 'A' curves with bilateral absent stapes reflexes. This led us to a provisional clinical diagnosis of bilateral Otosclerosis/Congenital stapes fixation and she underwent speech audiometry in order to measure her cochlear reserve. The poor speech scores were the first indicator that the child had something more than what her PTA indicated. The absent BERA and normal OAE confirmed the diagnosis of Auditory Neuropathy.^[6] In view of the coexisting bilateral optic nerve dysfunction, it is likely that this patient has a non isolated auditory neuropathy.^[7]

The normal OAE in the presence of conductive hearing loss could not be explained adequately as typically in conductive hearing loss OAE is absent. In retrospective, the absence of acoustic reflexes was attributed to either Auditory

Neuropathy and /or stapes fixation.

The role of speech audiometry in the management of conductive hearing loss is usually considered to be limited as it primarily used to evaluate sensory neural hearing loss. The decision to proceed to stapedotomy in otosclerosis is primarily based on the pure tone audiometry and impedance audiometry. If speech audiometry is not done in such a case, then the patient will undergo stapedotomy and post operatively no hearing improvement will be noticed. This would again initially be attributed to surgical causes of failure of stapedotomy – slipped prosthesis, short prosthesis or even sensory neural loss that can occur as a post operative complication.

The common scenario will be that the clinician will wait for 6 to 8 weeks for the surgical site to stabilize before which no clear diagnosis can be made on a definitive basis. The repeat audiogram at this stage would show a near normal hearing on the operated side since the air bone gap would have closed due to stapedotomy. Inability to hear inspite of such an audiogram would be first factor that would lead the clinician to think that there could be some other coexisting disorder and finally the diagnosis of auditory neuropathy will be made on further evaluation in a retrospective manner. This entire scenario can be preemptively avoided if speech audiometry is included in the investigative work up for conductive hearing loss as it was done in this case.

Auditory Neuropathy is a disorder which presents with different clinical and audiological findings and hence management must be individualized^[8]. Although patients with Auditory Neuropathy may demonstrate improvement in thresholds with amplification, the temporal encoding dysfunction and the consequent speech perception degradation is not alleviated by amplification^[9]. Cochlear implantation is often a good solution for failures of conventional rehabilitation^{[10][11]}. Our patient did not benefit from hearing aid amplification and therefore the other option was to consider cochlear implantation.

Although a High Resolution Computerised Tomogram and High Resolution MRI scan of the temporal bone could have provided more information regarding the ossicular status and the presence/absence of otosclerotic focus, the gold standard to establish the cause of conductive hearing loss would have been an exploratory tympanotomy under general anaesthesia. However in presence of proven auditory neuropathy, any corrective measure for the conductive hearing loss was unlikely to change the final prognosis. On

the contrary, this patient would certainly benefit from an implantable hearing device for her hearing problem.

CONCLUSION

1) Although the incidence of the association of Auditory Neuropathy with conductive hearing loss is unknown, it is worth subjecting all patients who present with findings typical of Stapes fixation for a speech audiometry. The pure tone audiogram may not by itself reveal the retrocochlear component and the absence of stapedial reflexes may get attributed to stapes fixation instead of auditory neuropathy. A poor speech discrimination score can be a screening test for Auditory Neuropathy and this can alter the entire management, as stapedotomy will not benefit such patients.

2) Auditory Neuropathy can coexist with conductive hearing loss – a fact not high-lighted in literature so far as all cases of Auditory Neuropathy classically presents with different grades of sensory neural hearing loss. Our current knowledge on Auditory Neuropathy as a retrocochlear pathology does not explain an element of conductive loss in it and hence we cannot consider this as a variant/ atypical presentation of Auditory Neuropathy. The clinician should be aware of such rare associations so that he is able to make the right diagnosis and thereafter carry out the correct management.

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