

# CASE REPORT

## AUDITORY NEUROPATHY WITH BILATERAL BAT EARS – A RARE CASE REPORT

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### HOW TO CITE THIS ARTICLE:

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**ABSTRACT:** Auditory neuropathy is a hearing disorder in which sound enters into the inner ear normally but the transmission of signals from the inner ear to auditory cortex is impaired due to the poor brain development. We report a case of 7 year male child presented with hard of hearing, deformed both external ears and inability to speak since birth. On examination bilateral bat ears , accessory auricles, hypertelorism, hyperactive attention deficit and from auditory brainstem response and otoacoustic emission reports it is diagnosed as auditory neuropathy and associated with various forms which is not included in ENT syndromes till date.

**KEYWORDS:** Auditory neuropathy, Bilateral bat ears, hyperactive attention deficit.

**INTRODUCTION:** The finding of normal cochlear function accompanied with abnormal brainstem responses was defined as auditory neuropathy (AN) by Starr, Picton, Sininger, Hood & Berlin, in 1996. Hearing disorder in which the transmission of signals from inner ear to the brain is impaired. It may have normal hearing, or hearing loss ranging from mild to severe. The main cause is due to damage to the inner hair cells. The hallmark of auditory neuropathy is a negligible or very abnormal ABR reading together with a normal OAE reading. The treatment is Occupational Therapy, Hearing Aids, Frequency Modulation Systems, Lip Reading, and Auditory - verbal Therapy. We report a case of auditory neuropathy with associated features like bilateral bat ears, hyperactive attention deficit.

### MATERIALS AND METHODS:

**CASE REPORT:** Patient Dharun 7 year male came to the ENT outpatient department with the history of hard of hearing, deformed both ears, inability to speak since birth. Perinatal history was prolonged labour, Neonatal hypoxic episode after delivery with prolonged assisted ventilation.



Fig. 1



Fig. 2



Fig. 3

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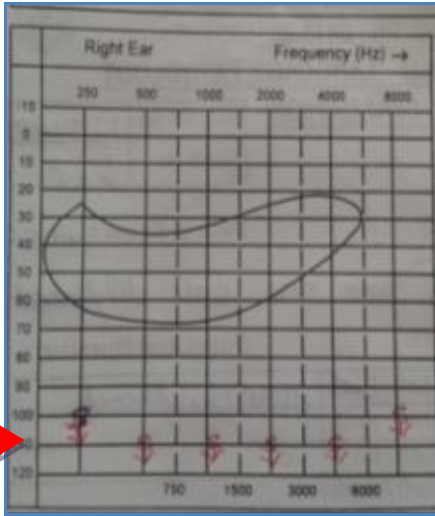


Fig. 4

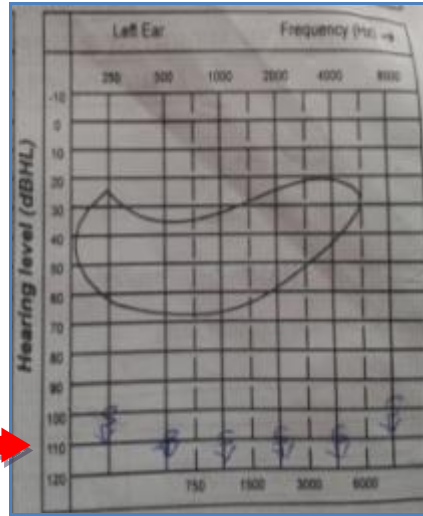


Fig. 5

On examination bilateral bat ears, accessory auricles (Fig 2, 3), hypertelorism (Fig 1), hyperactive attention deficit. Play audiometry shows bilateral sensorineural hearing loss (Fig 4, 5), Brain stem evoked response shows both ears -no repeatable peaks could be obtained for click stimuli at 90 dbnHL with rate of 21.1. Cochlear microphonics was observed which reversed with change in polarity of stimuli suggestive of auditory neuropathy (Fig 6, 7). Otoacoustic emission we recorded which was present in both ears (Fig 8, 9).

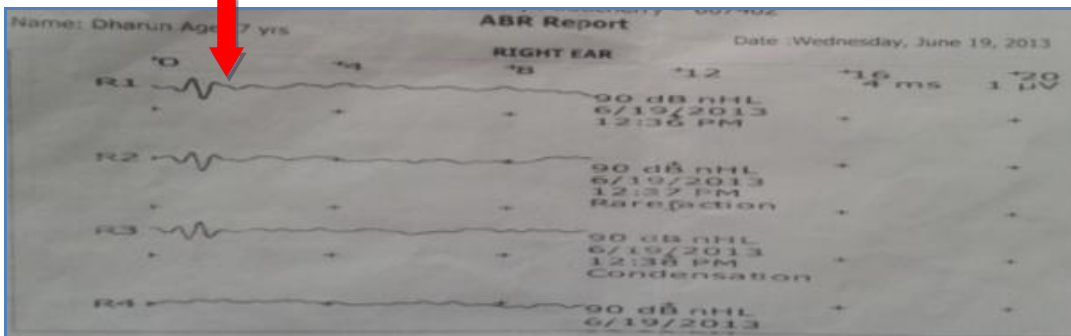


Fig. 6

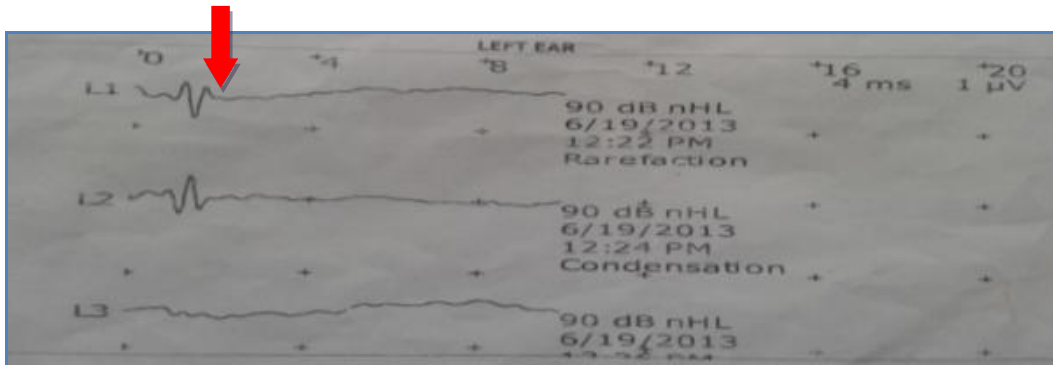


Fig. 7

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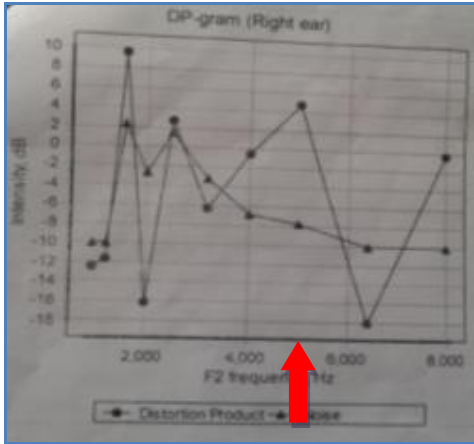


Fig. 8

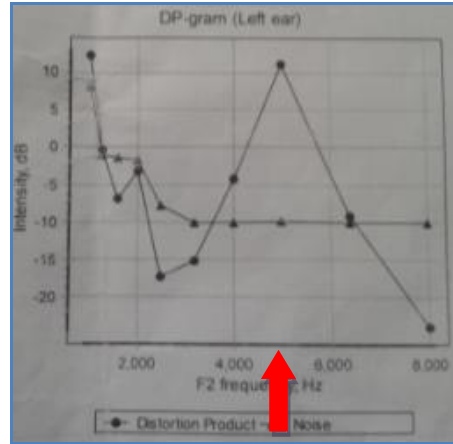


Fig. 9

Computerised tomography shows normal study (Fig 10 to 15). Treatment given was occupational therapy for increasing attention, complete inside canal (CIC) hearing aid trial for communication, speech stimulation and speech education for language development, sign language, and auditory verbal therapy lip reading.

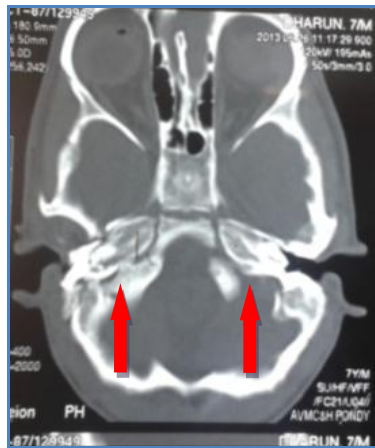


Fig. 10

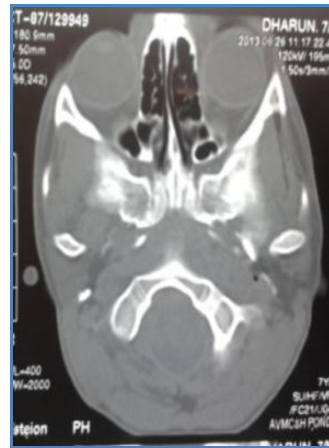


Fig. 11



Fig. 12

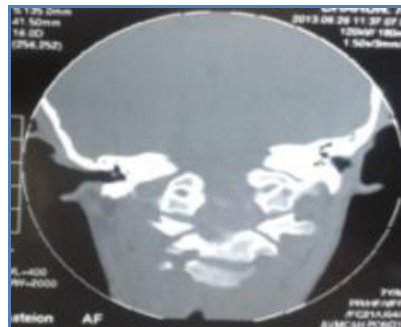


Fig. 13

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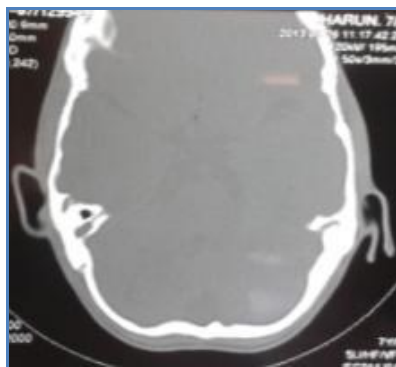


Fig. 14

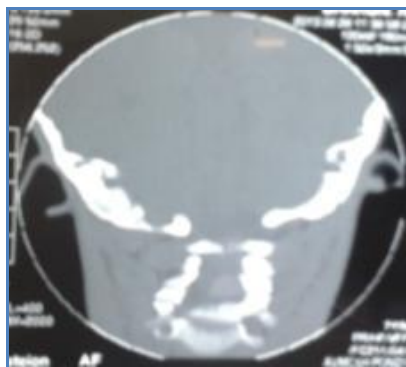


Fig. 15

**RESULTS:** A child with hard of hearing and inability to speak with negligible or very abnormal brainstem evoked response reading together with a normal otoacoustic emission reading suggestive of auditory neuropathy . A normal otoacoustic emission reading is a sign that the outer hair cells are working no Fig. 15

**DISCUSSION:** Auditory neuropathy is a hearing disorder in which sound enters the inner ear normally but the transmission of signals from the inner ear to the brain is impaired. It can affect people of all ages, from infancy through adulthood. People with auditory neuropathy may have normal hearing, or hearing loss ranging from mild to severe; they always have poor speech-perception abilities, meaning they have trouble understanding speech clearly. Often, speech perception is worse than would be predicted by the degree of hearing loss. The cause is mostly damage to the inner hair cells specialized sensory cells in the inner ear that transmit information about sounds through the nervous system to the brain. Outer hair cells help amplify sound vibrations entering the inner ear from the middle ear. When hearing is working normally, the inner hair cells convert these vibrations into electrical signals that travel as nerve impulses to the brain, where the impulses are interpreted as sound. Some children who have been diagnosed with auditory neuropathy experienced certain health problems as newborns, or during or shortly before birth. These problems include jaundice, premature birth, low birth weight, and an inadequate supply of oxygen to the unborn baby. The hallmark of auditory neuropathy is a negligible or very abnormal ABR reading together with a normal OAE reading. A normal OAE reading is a sign that the outer hair cells are working normally.

For Neonatal screening an ABR test monitors brain wave activity in response to sound using electrodes that are placed on the person's head and ears. An OAE test uses a small, very sensitive microphone inserted into the ear canal to monitor the faint sounds produced by the outer hair cells in response to stimulation by a series of clicks. ABR and OAE testing are painless and can be used for newborn babies and infants as well as older children and adults. Treatment includes was occupational therapy for increasing attention, complete inside canal (CIC) hearing aid trail for communication , speech stimulation and speech education for language development ,sign language ,auditory verbal therapy lip reading , frequency modulation (FM) systems are helpful. Cochlear implants and auditory brain implants are not useful effectively.

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**CONCLUSION:** The child presenting with deaf mute and perinatal insult along with abnormal brainstem evoked response and normal otoacoustic emission suspect auditory neuropathy. The features of auditory neuropathy along with bilateral bat ears, hyperactive attention deficit, and hypertelorism are rarest clinical findings which are not reported in any ENT syndromes till date.

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