INTRODUCTION
Reports since 1979 have noted that patients without reproducible brain stem auditory evoked response have thresholds within the normal range in pure tone audiometry. However, it is not until seventeen years later in 1996 that Starr et al described this to be a clinical entity called auditory neuropathy (AN). This is a type of hearing impairment due to a disorder of auditory nerve function and may have, as one of its cause, a neuropathy of the auditory nerve, occurring either in isolation or as part of a generalized neuropathic process. This is not a new clinical entity but a disorder that new diagnostic techniques make easier to detect. This disorder has been diagnosed in all ages from newborn to adults and it has been related to different etiologies. Auditory neuropathy has also been identified in peripheral, metabolic, toxic and inflammatory neuropathies or as an isolated neuropathy of the VIII cranial nerve. Starr et al in their report of 10 patients showed clinical evidence of peripheral neuropathy in seven and chorea in one patient. Hearing disorder preceded the latter in these patients by several years. In a retrospective study of 22 children, hyperbilirubinemia was found in 50% patients and the remaining having predisposing factors of prematurity / ototoxic drug exposure, cerebral palsy and family history of hearing loss.

The diagnostic criteria on AN is (1) sensorineural hearing loss, usually bilateral, of any degree; (2) normal outer hair cell function as evidenced by the presence of otoacoustic emissions (OAEs) and/or cochlear microphonics (CM); (3) abnormal evoked potentials beginning with wave I of the ABR; (4) poor speech perception; and (5) absent acoustic reflexes to the ipsilateral and contralateral tones at a 110 dB hearing level.

The typical patients with AN present as children or adults with dissociation between pure-tone and speech audiometry. There is mild or moderate hearing loss accompanied by disproportionate difficulties in speech discrimination and absent or severely distorted ABRs in the presence of preserved OAEs and/or EchocGst. The prevalence of AN is not unknown. There has been varying reports of 0.5%, 15% and 11%7,8. There has been no report of this entity from India. We present a series of four patients with this clinical entity.

REPORT OF PATIENTS
This is a report of 4 patients who attended the ENT outpatient department in the past 6 months with the history of severe inability to understand speech along with hearing loss. All patients were males and their ages ranged from 18 to 27 years. Details of the patients are tabulated in Table 1.

These patients were administered standard pure tone and bone conduction audiometry (250 – 8000Hz), speech recognition and speech discrimination tests. The pure tone audiograms of all the patients showed a low frequency loss of rising slope pattern (Figures 1-4). We were unable to establish speech recognition and discrimination scores in four patients in spite of their mild to moderate hearing loss. SRT was found in 1 patient, but not the discrimination scores.

1Professor, 2Professor & Head, 3Senior Audiologist, 4Lecturer, 5Lecturer, 6Audiology Technician, Dept of ENT, Christian Medical College, Vellore, INDIA
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Fig V Non recordable discrimination scores in all the four patients in spite of their mild to moderate hearing loss

Fig VI Absent ABRs in all four patients

Fig VII Normal DPOAEs in all four patients

Brainstem evoked response audiometry were recorded on the 'intelligent hearing systems' using electrodes on vertex, ipsi and contra lateral on either mastoids and ground on the forehead. Clicks were presented via insert earphones and the responses were recorded. All four patients had absent ABRs (Fig VI).

Distortion product emissions (DPOAE) were measured with Intelligent hearing systems. The starting frequency was 500 Hz and the end frequency was 8000 Hz. The loudness level ranged from 55-65 dB SPL. These were presented at the rate of 32 sweeps, averaged and stored in two buffers. All four patients had normal OAEs (Fig VII) at least in one ear. In patient 2, emissions were normal at low and high frequencies, low amplitude between 1.5 - 3.5 kHz in right ear.

Tympanometry and acoustic reflex testing for pure tone stimuli from 500 Hz - 4000 Hz were also performed. Acoustic reflexes were considered absent when both ipsilateral and contra lateral stimuli fail to elicit responses even at 110 dB HL. Tympanometric results showed normal pattern in all patients and reflexes absent in 3 of the four patients. Ipsa and contra lateral reflexes were present in one of the patients.

The pure tone audiograms of the 4 patients showing a low frequency loss of rising slope pattern (Fig V)

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