Temperature-induced auditory neuropathy is a rare disease. It is a clinical diagnosis based upon hearing loss associated with:

1. Fever
2. Normal cochlear microphonics (i.e., otoacoustic emissions)
3. Absence or abnormal auditory brainstem response

Pathophysiology is thought to be secondary to demyelinating neuropathy of the auditory nerve, specifically at the node of Ranvier. Demyelination may account for the abnormal auditory brainstem response. Normal cochlear microphonics confirm normal cochlear function.

Patients often return to baseline hearing once they have defervesced; however, the auditory brainstem response remains abnormal.

HPI: 24 y/o female presented with bilateral hearing loss and isolated fever to 100°F. She was unable to hear anything secondary to the loud bilateral tinnitus. Denied upper respiratory symptoms.

PMH: Similar symptoms had occurred in the past since one year of age. There were increases in temperature followed by complete resolution.

Audiometric evaluation (Figure 1): Pure tone audiometry demonstrated bilateral severe pan-frequency SNHL. Cochlear microphonics, both DPOAE and ABR were within normal limits. Auditory brainstem response (ABR) demonstrated wave one only. These findings were consistent with evaluations performed during prior febrile episodes.

Follow-up 2 weeks later (Figure 2): Audiogram returned to baseline of mild high frequency sensorineural hearing loss. ABR remained abnormal.