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Auditory neuropathy: pathophysiology

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Auditory neuropathy (AN) describes a recently hearing disorder characterized by dysfunction of the auditory nerve (ABRs are absent or severely impaired) in the presence of preserved cochlear outer hair cell functions (OAEs and CMs are preserved). A low frequency or flat hearing loss are the most common audiometric configurations. The major symptoms of the disorder are impaired speech comprehension that is disproportionately affected by environmental noise. AN is not rare and surveys suggest an incidence of up to 10% of hearing impaired subjects. The etiologies are diverse and include toxic-metabolic disorders (hyperbilirubinemia and anoxia) neonates, infectious disorders (e.g., mumps) in adolescence, and hereditary disorders affecting peripheral and auditory nerves (e.g., Charcot-Marie-Tooth disorders, Friedreich's ataxia), and hereditary disorders without peripheral neuropathy (e.g, otoferlin). One-half of AN subjects do not have an etiology that can be clearly identified. The sites of auditory nerve dysfunction include the ganglion cells and axons (Type I) or the synaptic junction between inner hair cells and nerve terminals (Type II). The pathophysiology and psychoacoustics underlying the hearing loss as well as current treatments of the disorder will be discussed.

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