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# Auditory Neuropathy in Childhood

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**Objectives:** Auditory neuropathy is a recently described disorder in which patients demonstrate hearing loss for pure tones, impaired word discrimination out of proportion to pure tone loss, absent or abnormal auditory brainstem responses, and normal outer hair cell function as measured by otoacoustic emissions and cochlear microphonics. We have identified eight pediatric patients having hearing deficits that are most likely due to a neuropathy of the eighth nerve. In this study, the results of audiologic testing performed with these eight children are described. **Study Design:** Retrospective review of audiologic findings in eight children with auditory neuropathy. **Methods.** Each subject was tested with pure tone and speech audiologic testing, auditory brainstem response, and click-evoked otoacoustic emissions. Results of these tests were tabulated and summarized. **Results:** Pure tone audiologic testing revealed five children with upsloping sensorineural hearing loss, two with high frequency loss, and one with a mild, flat configuration. Six children demonstrated poor word discrimination scores, and the other two had fair to good word discrimination. All eight subjects had normal distortion product and transient otoacoustic emissions. All eight children demonstrated absent or marked abnormalities of brainstem auditory evoked potentials. These findings suggest that while cochlear outer hair cell function is normal, the lesion is located at the eighth nerve. **Conclusions:** Recent advances in otoacoustic emissions testing permit differentiation of neural deafness from sensory deafness. This paper describes the clinical presentation and audiologic findings in pediatric auditory neuropathy, as well as the recommended management of these patients. Otolaryngologists should be aware of this disorder and implications for its management, which differs from treatment of sensorineural hearing loss. **Key Words:** Auditory neuropathy, childhood, hearing loss, auditory brainstem response, evoked otoacoustic emissions.

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## INTRODUCTION

A group of pediatric patients has been identified who have hearing loss and absent or severely abnormal auditory brainstem responses (ABRs), yet have normal cochlear function as measured by otoacoustic emissions (OAEs). Starr et al.<sup>1</sup> named this disorder *auditory neuropathy* and studied the auditory abilities of these patients. Starr et al.<sup>2</sup> described an 11-year-old girl who developed progressive hearing loss from the age of 7 years, and who had such great difficulty understanding speech that she was dependent on lip-reading. Her pure-tone audiogram progressed from mild hearing loss to bilateral, moderately severe neurosensory hearing loss with poor word discrimination scores (16% right and 8% left). Evoked auditory potentials, repeatedly measured throughout her clinical course, consisted of high-amplitude cochlear microphonics to 87-dB nHL clicks, and completely absent ABR to clicks or tone bursts up to 95-dB nHL. Starr et al. believed that a disorder occurring anywhere along the afferent auditory system from the inner hair cells to the eighth nerve cell body could account for the audiometric loss and the loss of auditory evoked potentials. They thought that cochlear microphonic preservation was due to the presence of normal outer hair cell function in this child.

Starr et al.<sup>1</sup> reported on 10 patients, including five adults and five children, all with pure-tone hearing loss ranging from mild to severe, poor speech discrimination, absent or severely abnormal ABRs, and normal click-evoked OAEs. Magnetic resonance imaging of the brain revealed no brainstem or eighth nerve abnormalities to account for the electrophysiologic findings. Since that report, we have identified eight additional children with auditory neuropathy. In this paper, we present the audiologic findings in these eight pediatric patients, and suggest possible rehabilitative strategies for this unusual disorder.

## METHODS

### *Subjects*

Subjects include eight children aged 4 years to 15 years, all of whom were identified and referred over a 2-year period by four audiologists from four audiology clinics serving children in Los Angeles and Orange counties in California. There are five boys and

three girls. All children were initially diagnosed as having sensorineural hearing loss. Further workup by the referring audiologists confirmed that they had absent or abnormal ABRs and normal OAEs. Table I summarizes demographic data for the eight subjects. Subject 2 developed normal speech and language until age 18 months, when he contracted Stevens-Johnson syndrome in response to sulfamethoxazole treatment. His parents reported that after that illness his speech and language development ceased and he could no longer hear their voices, although he still responded to music. Subject 8 has Friedreich's ataxia and reportedly had pneumonia before diagnosis of hearing loss at the age of 18 months. Subjects 3 and 8 have absent deep tendon reflexes on neurologic examination. There are no other known neurological or medical abnormalities in the other children.

### Audiologic Testing

Standard pure-tone audiometric testing for the frequencies 250, 500, 1000, 2000, 4000, and 8000 Hz was carried out for all eight children. Masked bone conduction testing could not be accomplished for the three youngest children only. Speech audiometry was not possible in the four youngest subjects, who have little spoken language.

Click-evoked OAEs were measured with the ILO-88 OAE system (Otodynamics, LTD., United Kingdom). Click level ranged from 80- to 87-dB peak sound pressure. Responses to 260 stimuli were averaged and stored in two separate buffers. The presence of normal click-evoked OAEs was determined by response signal-to-noise ratio of at least 4 dB and waveform reproducibility in at least three octave bands of more than 75%.

Auditory brainstem evoked potentials were recorded in two electrode configurations: in a vertical channel, vertex to seventh cervical vertebra to optimize detection of wave V and vertex to the ipsilateral ear using band-pass from 30 or 100 Hz to 3000 Hz. Click stimuli were rarefaction clicks presented monaurally at rates from 5 to 25/s and at intensities of 65-, 75-, 85-, and in most cases, 95-dB nHL. Two averages were made at each test level and reproducible components of the ABR were evaluated.

### RESULTS

Table II presents the results of pure tone audiometric testing, word discrimination scores, click-evoked OAEs, and ABRs for the eight children. Seven ears had an upsloping hearing loss, four had high-frequency hearing loss, three had flat configuration, and two had profound hearing loss. Severity ranged from mild to profound. In the four youngest children, who had insufficient spoken language, word discrimination could not be assessed and is assumed to be poor on this basis. Of the remaining four children, three had poor-to-fair word discrimination. Subject 5 had very good word discrimination (88% to 92%).

Because auditory neuropathy is defined here as the presence of normal evoked OAEs and absent or abnormal ABRs, all of the children in this study have normal OAEs and abnormal or absent ABRs, as shown in (Table II). The exception is subject 2, a 6-year-old boy who exhibits the auditory neuropathy pattern in the left ear, but who has profound sensorineural hearing loss in the right ear without OAEs.

### DISCUSSION

Traditionally, the site of lesion of hearing loss is classified as sensorineural or conductive, with sensorineural hearing loss being subdivided into cochlear versus retrocochlear. The eight children presented in this paper have a hearing loss that would be more accurately classified as neural, but the available data only permit identification of the site of lesion as being "post-outer hair cell." The presence of OAEs indicates that the outer hair cells are functional, but the abnormality in auditory neuropathy could be at the inner hair cells, the synapse between the inner hair cells and their dendrites, the spiral ganglion, the eighth nerve fibers, or combinations of the above. It is likely that as more is learned about the disorder, different forms of auditory neuropathy will be demonstrated at each of these anatomic areas. Because wave I of the ABR is absent in auditory neuropathy patients, the lesion cannot be restricted to the cochlear nucleus, although it is possible that there are brainstem auditory pathway abnormalities in addition to more peripheral lesions. Post-mortem histological techniques currently can differentiate these lesions,<sup>3</sup> but there are at this time no diagnostic tests that differentiate, for instance, an inner hair cell lesion from a spiral ganglion lesion. At least two studies have been published outlining the pathological changes in the temporal bones of individuals with hearing loss and peripheral neuropathies. Spoendlin<sup>3</sup> described the temporal bones of two individuals with Friedreich's ataxia. He noted that the organ of Corti was normal, but that there was damage to the spiral ganglion cells in these patients. Hallpike et al.<sup>4</sup> also found normal hair cells, but degeneration of spiral ganglion cells and auditory nerve fibers in a patient with hereditary hearing loss, poor speech comprehension, and peripheral neuropathy.

Other researchers have identified adults and children with auditory neuropathy. Kraus et al.<sup>5</sup> described four patients with audiometric findings ranging from normal hearing to moderate hearing loss, all of whom had absent ABRs.

TABLE I.  
Demographic Data for Eight Children With Auditory Neuropathy.

Subject No.	Sex	Age (y)	Onset Age	Other Disease
1	M	4	Congenital	None
2	M	6	1 y	Stevens-Johnson syndrome (1 y)
3	M	6	Congenital	None
4	M	7	Congenital	None
5	F	11	Unknown	None
6	F	13	Unknown	None
7	F	13	Unknown	None
8	M	14	1 y	Friedreich's ataxia

TABLE II.  
Audiologic Data for Eight Children With Auditory Neuropathy.

Subject No.	Age (y)	Hearing Loss Configuration (AD/AS)	Severity (AD/AS)	WDS (AD/AS)	OAE (AD/AS)	ABR (AD/AS)
1	4	Upslope/flat	Severe/profound	CNT/CNT	+/+	abs/abs
2	6	NR/upslope	Profound/severe	CNT/CNT	-/+	abs/abs
3	6	Upslope/flat	Severe/severe	CNT/CNT	+/+	abs/abs
4	7	Flat/flat	Mild/mild	CNT/CNT	+/+	abs/abs
5	11	High freq./high freq.	High freq./high freq.	88/92	+/+	abn/abn
6	13	Upslope/upslope	Moderate/moderate	76/72	+/+	abn/abn
7	13	High freq./high freq.	High freq./high freq.	56-64	+/+	abs/abs
8	14	Upslope/upslope	Moderate/moderate	40/68	+/+	abs/abs

WDS = word discrimination score; OAE = click-evoked otoacoustic emissions; ABR = auditory brainstem response; CNT = could not test; NR = no response; + = present; - = absent; abs = absent; abn = abnormal.

What was remarkable to these researchers was that the ABR abnormalities were far out of proportion to the pure-tone loss. The localization of the disorder to the eighth nerve could not be made at that time, as methods for defining outer hair cell function were not yet widely available (OAEs) or not routinely used (cochlear microphonics). Kraus et al. felt that these cases probably had neuropathology of the auditory brain stem. Cases of absent ABR in the presence of normal hearing had been identified nearly 20 years ago.<sup>6,7</sup> More recently, additional cases of adult and pediatric auditory neuropathy have been found<sup>8-12</sup> (Sininger et al., Miniseminar presented at the American Speech Language Hearing Association Convention, Anaheim, CA, November, 1993). Berlin et al.<sup>12</sup> described three patients having auditory neuropathy as part of Charcot-Marie-Tooth disease, a hereditary sensorimotor neuropathy. Subject 8 in this paper has auditory neuropathy probably as a consequence of Friedreich's ataxia, a disorder that has previously been described as producing abnormal ABRs in more than half of those affected.<sup>14</sup> The incidence of auditory neuropathy is not yet known, but is seen infrequently relative to sensory hearing loss. However, it is possible that some individuals who were diagnosed with sensorineural hearing loss before OAE testing came into common use actually have undetected neuropathy. We recommend routine OAE testing in children newly diagnosed with hearing loss.

Auditory neuropathy presents therapeutic challenges distinct from usual "sensorineural" hearing loss. In particular, speech understanding is so greatly impaired that patients with prelingual onset fail to develop spoken language, as in subjects 1 through 4. In particular, subject 4 has only a mild hearing loss for pure tones, yet has failed to develop spoken language. As with patients with acoustic neuroma and "retrocochlear hearing loss," hearing aids are seldom of benefit in adult or pediatric patients with auditory neuropathy. Indeed, some would point out that hearing aids are contraindicated in the face of intact outer hair cell function, because of the risk of noise-induced damage to the outer hair cells. However, anecdotally, some of these individuals benefit from hearing aids, which provide amplification of sounds, if not improved discrimination. We therefore usually recommend a trial of hearing aids when counseling parents of these children, despite our pessimism that hearing aids will be

of any benefit. We have not seen any adults with auditory neuropathy who benefitted from the use of hearing aids, but Widen et al.<sup>13</sup> report a case of a woman with auditory neuropathy who initially wore hearing aids. Speech-reading classes are strongly recommended for adults with auditory neuropathy; sign language, and intensive speech and language therapy provide the mainstay of habilitation for children with the disorder. Two of the patients presented in this paper use FM auditory trainers in their classrooms for the hearing-impaired. Some children in other centers have received benefit from vibrotactile devices. Cochlear implants are being considered for neuropathy patients, but it is unknown whether electrical stimulation will "resynchronize" neural activity.

## CONCLUSION

In this paper we have described what is currently known about the clinical presentation and audiologic findings in pediatric auditory neuropathy, as well as the recommended management of patients with this disorder. Otolaryngologists should be aware of the disorder and implications for its management, which differs from treatment of sensorineural hearing loss.

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