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Predicting Cochlear Implant Outcomes in Children With Auditory Neuropathy

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Objective: To examine the outcome of cochlear implantation in children with auditory neuropathy (AN) and cochlear nerve deficiency (Group A). Results are compared with a cohort of children with AN and normal cochlear nerves (Group B).
Study Design: Retrospective cohort study.
Setting: The Sydney Cochlear Implant Centre and the Children's Hospital at Westmead.
Patients: Children younger than 15 years with bilateral profound sensorineural hearing loss and the diagnosis of AN confirmed on electrophysiologic testing. All children underwent cochlear implantation with Nucleus 24 cochlear implants from 1997 to 2006.
Interventions: Magnetic resonance imaging was examined for deficiency of the vestibulocochlear nerve. Brain and inner ear abnormalities were recorded. Cochlear implant outcomes and demographic variables were compared.
Main Outcome Measures: Melbourne speech perception score (MSPS) at 1 year and implant evoked electric auditory brainstem response (EABR).
Results: Group A performed significantly worse on both parameters than Group B. In Group A, median MSPS was 1, compared with a median score of 4 in Group B ($z = -3.010$; $p = 0.003$). EABR was abnormal in 13 of 15 (87%) children in Group A, compared with 9 of 39 (23%) in Group B. Children in both groups with abnormal EABR had significantly worse MSPS ($z = -2.780$; $p = 0.005$). Fourteen of 15 children with cochlear nerve deficiency had associated inner ear abnormalities.
Conclusion: Children with AN can have associated cochlear nerve deficiency. These patients have worse speech perception scores at 1 year post cochlear implantation, higher rates of abnormal EABR, and more associated inner ear abnormalities than children with AN and normal cochlear nerves. **Key Words:** Auditory neuropathy— Cochlear nerve deficiency—Cochlear implant—Cochlear nerve— Electric auditory brainstem response—Magnetic resonance imaging—Pediatric.
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Auditory neuropathy is an otologic syndrome characterized by moderate-to-profound sensorineural hearing loss (SNHL), preserved otoacoustic emissions (OAEs) and/or cochlear microphonic potentials, and abnormal or absent auditory brainstem response (ABR). Outer hair cells remain intact, but there is no synchronous neural activity of the cochlear nerve when measured with ABR. Typically, speech discrimination scores are worse than predicted by pure tone audiology, and response to amplification with hearing aids is poor (1). The incidence of AN has been estimated as 10% to 14% of children diagnosed with severe to profound SNHL (2–4). The natural history may be progressive or transient. Even in cases of mild hearing loss, those with prelingual onset often do not develop speech (5). Hearing aids, rarely beneficial, are usually trialled for lack of other options.

Theories proposed about the pathophysiology include lesions at any of the following sites: inner hair cell, inner hair cell synapse with the cochlear nerve, spiral ganglion cells, axon, myelin sheath, and nerve dendrite (6,7). Clinical testing does not differentiate between these entities. Starr et al. (8) recognized that the condition could reflect pathologic abnormality at any of these sites. The term *auditory neuropathy* was coined because many of these patients had an associated peripheral neuropathy, and it seemed likely that the auditory nerve was the source of the pathologic abnormality. Further studies revealed that only one third of patients with AN had an associated peripheral neuropathy (9).

Many suspected that this was due to different clinical entities being described under the blanket term of *AN*. This was confirmed in several histologic studies that identified different pathologic sites in the auditory

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