Recovery of Functional Hearing Abilities with Auditory Neuropathy

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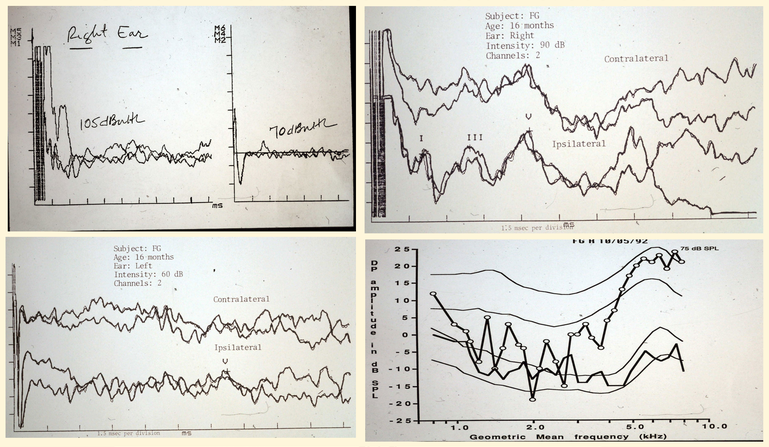
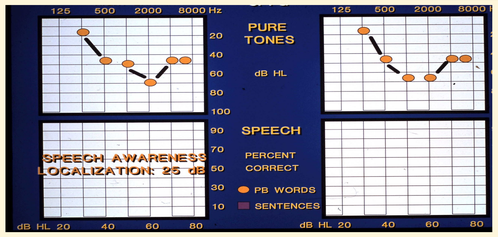
Introduction. Auditory neuropathy is classically defined as an absent or abnormal auditory brainstem response (ABR) with normal otoacoustic emissions and a present cochlear microphonic.[1] Although onset can be at any age, it is diagnosed most in infants on the basis of the objective tests. Not uncommonly, behavioral responses may reflect little or no response to sound stimuli. But in a relatively small number of children, functional recovery of hearing abilities seems to occur spontaneously over the first 18-24 month of life.

An added complication is the addition of autism spectrum disorder that eventually becomes the primary diagnosis, although the ABR continues to be abnormal reflecting ongoing dys-synchrony of the auditory nerve function. When this happens, it becomes difficult to determine whether the child's auditory response patterns are due to auditory neuropathy or autism spectrum disorder. Over the course of approximately 30 years, we have seen three children with recovery of auditory function but left with a residual diagnosis of autism spectrum disorder. We will discuss the first child, seen in 1993, and the child initially seen 11 years ago whom we are still following. The third child, born almost three years ago, appears to be following the developmental trajectory of the earlier two children. All three children serve as illustrative cases highlighting the contribution of parent report, clinical observations, objective test results combined with behavioral audiometry to determine an accurate diagnosis and the need for close monitoring of auditory status. But perhaps the most intriguing element of these case reports is the residual diagnosis of autism spectrum disorder revealed through the recovery of peripheral auditory abilities.

FG was born at 32 weeks GA and spent approximately one month in the NICU due to hypoxic episodes after birth. The early ABR showed no response to click stimuli leading us to believe that he had profound SNHL bilaterally. Three months after discharge, his father asked me if hearing loss ever gets better. Behaviorally, he was responding to softer sounds. The ABR at 16 months of age showed improved responses as did OAEs for mid- and high-frequencies. Final audiogram showed mid- and high frequency hearing loss bilaterally. Based on developmental delays, he was diagnosed with autism spectrum disorder. His last audiogram was in 2014 with little change from the one shown here. After that, he was lost to follow-up.

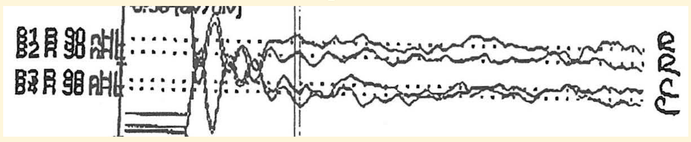
FG birth ABR and 16 months later. Final audio below.



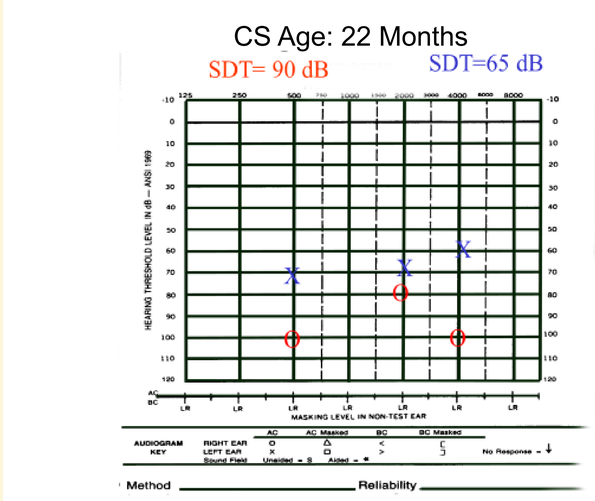


CS was born in 2010 at 33 weeks GA. He had a NICU stay of five weeks and was discharged with typical well-child follow-up. CS failed the newborn hearing screening but was not seen for follow-up until 22 months. Inattention to sounds and speech delay were presenting symptoms. At 30 months his vocabulary was growing, and he was putting several words together for sentences. He wore hearing aids until age 3 years and then could not tolerate the amplification. Repeated audiograms showed normal hearing sensitivity until September 2022 when he failed a school screen. The most recent audio was Sept. 2022 which began to show high frequency SNHL. He has started wearing mild gain hearing aids. He retains the residual diagnosis of autism spectrum disorder and needed support services for the first five grades. Now, he no longer needs support services and is on the school honor roll.

CS ABR showing CM and abnormal ABR



CS Age: 22 Months



CS
38 months old

		Threshold (dB HL)				
		500Hz	1000Hz	2000Hz	4000Hz	SDT
AIR	Right	0	0	5	10	0
	Left	0	0	0	5	0

CS
Current audiogram:
September 2022

		250 Hz	500 Hz	1000 Hz	2000 Hz	3000 Hz	4000 Hz	6000 Hz	8000 Hz
AIR	*Right*	25 dB HL	20 dB HL	30 dB HL	40 dB HL	30 dB HL	40 dB HL	30 dB HL	75 dB HL
AIR	*Left*	25 dB HL	30 dB HL	30 dB HL	40 dB HL	35 dB HL	45 dB HL	40 dB HL	75 dB HL
BONE	*Left*	DNT	25 dB HL	20 dB HL	25 dB HL	DNT	40 dB HL	DNT	DNT

NS was born in 2020 at 37 weeks GA. His newborn hospital stay was two days. He failed his newborn hearing screen but was lost to follow-up until 14 months of age. An ABR showed significant hearing loss in the left ear. The right ear was tested two days later with similar results. He was fitted with bilateral hearing aids and wore them essentially all waking hours. His mother reported improved but inconsistent responses to sound stimulation but no advance in vocalizations or language. We had to decrease amplification three times with his behavioral complaints of sound being too loud. Simultaneously, audiometric results showed improvement in sensitivity to music, speech, narrow bands of noise, frequency analyzed noise-makers, and eventually warble tones. As time continued, his behavior and responses to sounds appeared to be more like a child with autism instead of a child with simple auditory neuropathy. An interdisciplinary team evaluation in October 2022 confirmed a diagnosis of autism spectrum disorder. The latest audio was obtained via VRA with insert earphones using pulsed warble tones, pulsed narrow band noise, music, and speech and showed normal hearing sensitivity at each test frequency. His latest ABRs have shown no response at 105 dB nHL.

Ear	Stimulus	Intensities Tested (dB nHL)	Wave V Threshold (dB nHL)	Wave V Threshold (dB eHL)	**Interpretation**
Left Ear	Click	70, 60	70	70	**Moderate Hearing Loss**
	500 Hz	50, 60, 70	60	60	**Moderate Hearing Loss**
	2000 Hz	70, 80, 90	90	90	**Severe Hearing Loss**

Audio date: 1/9/23	500 Hz	1000 Hz	4000 Hz
LEFT EAR	25 dB HL	25 dB HL	20 dB HL
RIGHT EAR	25 dB HL	25 dB HL	25 dB HL

Discussion. There exist scattered reports in the literature of puzzling hearing function improvement with a diagnosis of auditory neuropathy with recovery taking as long as 2 years.[2,3] And it is even more unusual to have the additional diagnosis of autism spectrum disorder as an additional diagnosis. Even with recovery of hearing function, the ABR may remain abnormal . But one of the most important "take-aways" from this presentation is the need for and the role of behavioral testing as a complement to the objective test procedures.[2] Observations from the parents as well as audiologists in the clinic are also important as a cross-check on objective results. Clinically, we rely heavily on tests of auditory physiology to lead us toward appropriate diagnoses of hearing status. But there must be agreement between the test outcomes, parental observational reports, and clinical observations. If there is disagreement among these factors, something is missing from this diagnostic picture, and continued diagnostic exploration is essential. For these three children, final status blended diagnostic efforts of audiologists, speech-language pathologists, and psychologists to explain the totality of each child's development. These children also demonstrate the need for observation of overall development, apart from hearing alone, in order to arrive at a correct, all-encompassing diagnosis. Each child's behavioral responses to hearing aid amplification and developmental patterns emphasized the need to look beyond the hearing status and bring in colleagues of other disciplines to supplement what we did.

We do not yet know the link between ANSD, recovery of hearing function, and ASD, but these three case studies suggest that there may be a link. These children illustrated the necessity not to be overly aggressive toward cochlear implants until the child's status is confirmed and stable. Measurable improvements in hearing sensitivity justify watchful waiting for a period of time as long as documentable improvements continue. But this is also not to suggest waiting an extraordinary amount of time waiting for improvements to appear. In each case presented here, progressive improvements in hearing sensitivity began appearing before 12 months of age.

References.

1. Berlin CI, Hood L,Morlet T, Rose K, Brashears S. *Auditory neuropathy/dyssynchrony:diagnosis andmanagement.* Ment Retar and Devel Dis Res Rev. *2003;9(4):225-231.*
2. Madden C, RutterM, Hilbert L, Greinwald J, Choo D. Clinical and audiological features in auditory neuropathy. *Arch Otolaryngol Head Neck Surg.* 2002b;128:1026-1030.
3. Psarommatis I, RigaM, Douros K, et al. Transient infantile auditory neuropathy and its clinical implications. *Int J Pediatr Otorhinolaryngol*. 2006;70(9):1629-1637.