Briefing: Auditory Neuropathy
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What is auditory neuropathy (AN)?

- Hearing disorder affecting the connections between the ear and the brain.
- Classic diagnostic definition: Absent ABRs despite present OAEs[1].
- Better definition: A hearing phenotype characterized by a lack of subcortical neural synchrony, manifesting as absent ABRs that cannot be explained by sensorineural or conductive hearing loss.
- Causes: unknown, genetic, or acquired (typically acute illness in infancy, such as hyperbilirubinemia).
- CDC estimates about 5-7% of pediatric hearing loss cases are AN[2].

AN is a relatively new hearing disorder

- First identified in the early 1980's[3], "auditory neuropathy" coined in the 1990's[4].
- Not mainstream consideration for pediatric hearing loss until late 1990's/early 2000's.
- As a result, the first major cohort of AN patients are now in their 20's.
- There may also be adults with hearing problems attributable to undiagnosed AN.

Evaluating hearing in AN is challenging

Hearing Problems
- Inconsistent response to sound
- Poor sound awareness
- Extraordinary difficulty listening in noise

- Auditory function can fluctuate .
- AN is not mutually exclusive from sensorineural hearing loss
- Tremendous variability *within* the AN population itself.
- But, two clinical hallmarks: absent or grossly abnormal ABRs and absent acoustic reflexes.

Expected results from comprehensive hearing evaluation

Test	Expected Results	Comments
Tymps	Normal	
Acoustic Reflexes	**Always absent**	**Red flag, particularly in children where an ABR study is difficult**
OAEs	Typically present, but variable	Typically OAE amplitudes are larger than normal, but can be absent or disappear over time
Audiograms	Variable	Runs the gamut from completely normal to moderate-severe sensorineural hearing loss
ABRs	**Always absent**	**Easy to mistake cochlear microphonic for ABR**
MLRs	Typically present	
CAEPs	Typically present	
Speech in Quiet	Typically normal	Usually excellent, assuming accounting for audibility in cases of SNHL
Speech in Noise	**Severely impaired**	**Substantial difficulty hearing speech in noise**

[1] Thus the importance of using OAEs *and* ABRs in newborn hearing screening!
[2] https://www.cdc.gov/ncbddd/hearingloss/2018-data/14-type-and-severity.html
[3] Kraus et al. (1984) *Laryngoscope*
[4] Starr et al. (1996) *Brain*

AN listeners have extreme difficulties understanding speech in noise, but normal recognition in quiet

- Word recognition: similar to controls in quiet, identify 50-75% fewer words in noise[5].
- Sentence recognition: similar to controls in quiet, but <1st percentile in noise[6].
- Listening with 2 ears boosts sentence recognition considerably.

AN listeners struggle to process fast temporal cues in sound

- Poor at detecting a brief tone embedded in noise[7].
- Perceptual and neurophysiological (MMN) evidence of poor processing of temporal contrasts in speech sounds, such as /b/ vs. /g/[8], but, normal processing of slower temporal contrasts, such as /b/ vs. /w/.

AN listeners do not exhibit a frequency-following response (FFR), confirming the FFR relies on subcortical neural synchrony

- Double dissociation: subcortical neural synchrony necessary and sufficient for FFR; auditory cortices necessary and sufficient for CAEPs. [9]
- Even for stimuli within the range of auditory cortex phaselocking, subcortical neural synchrony is necessary to generate the FFR[10]; cortical activity may contribute.

AN provides fundamental insights into the importance of subcortical neural synchrony

- Neural synchrony is necessary to hear in noise and generate an FFR.
- FFR requires subcortical synchrony. FFR reflects neural mechanisms underlying speech-in-noise recognition.
- Normal OAEs and hearing thresholds do not guarantee normal auditory function.
- By extension, mild forms of dyssynchrony may account for everyday listening challenges[11].

Our speculations about AN and what it tells us about normal hearing

- Lack of acoustic reflexes means AN listeners live without the ear's protective mechanisms in noise. This may elevate risk of developing sensorineural hearing loss in adulthood.
- AN illustrates important role of binaural hearing to recognize speech in noise.
- Subcortical synchrony is important for sound awareness – ability to detect a new sound amid a complex soundscape.

For more information: www.brainvolts.northwestern.edu

[5] Kraus et al. (2000) *JARO*
[6] White-Schwoch et al. (2020) *JAMA-Oto*
[7] Kraus et al. (2000) *JARO*
[8] Kraus et al. (1993) *Ear Hear*; Kraus et al. (2000) *JARO*
[9] White-Schwoch et al. (2019) *J Neurophysiol*
[10] White-Schwoch et al. (2021) *J Neurophysiol*
[11] Anderson et al. (2012) *J Neurosci*