



Auditory Neuropathy/Auditory Dys-synchrony

Audiological diagnosis and management

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Terms

- Auditory Neuropathy (AN)
- Auditory Dys-synchrony (AD)
- Auditory Neuropathy Spectrum Disorder (ANSD)



What is AN/AD?

- Patient displays auditory characteristics consistent with normal outer hair cell function and abnormal neural function at level of the VIIIth nerve
- Speech perception abilities out of proportion with behavioral audiograms
- Starr et al. 1996 – first use of term AN



Why are we seeing it so much more?

- Universal Newborn Hearing Screening
- Audiologists becoming more familiar with characteristics and management
- “It is not a new disease, but an entity, which is recognized because of improvement in hearing assessment.” – Ngo et al.



Causes of AN/AD

- Inner hair cell (IHC) loss or damage
- myelinization disorder
- auditory nerve disorder with other peripheral neuropathies
- damage to IHC/VIIIth nerve synapse

Risk Factors AN/AD

- Lack of oxygen in neonatal period (i.e. asphyxia, anoxia, hypoxia)
- Prematurity
- Infectious processes
- Hyperbilirubinemia
- Low birth weight
- Other genetic disorders with accompanying neuropathies

- Some have no risk factors!



Incidence

- Unknown
- ~ 7-10% of those diagnosed with permanent hearing loss
 - (studies range from 5-15%)
- Up to 40% of the hearing loss in NICU babies



Diagnostic Tests (Audiology)

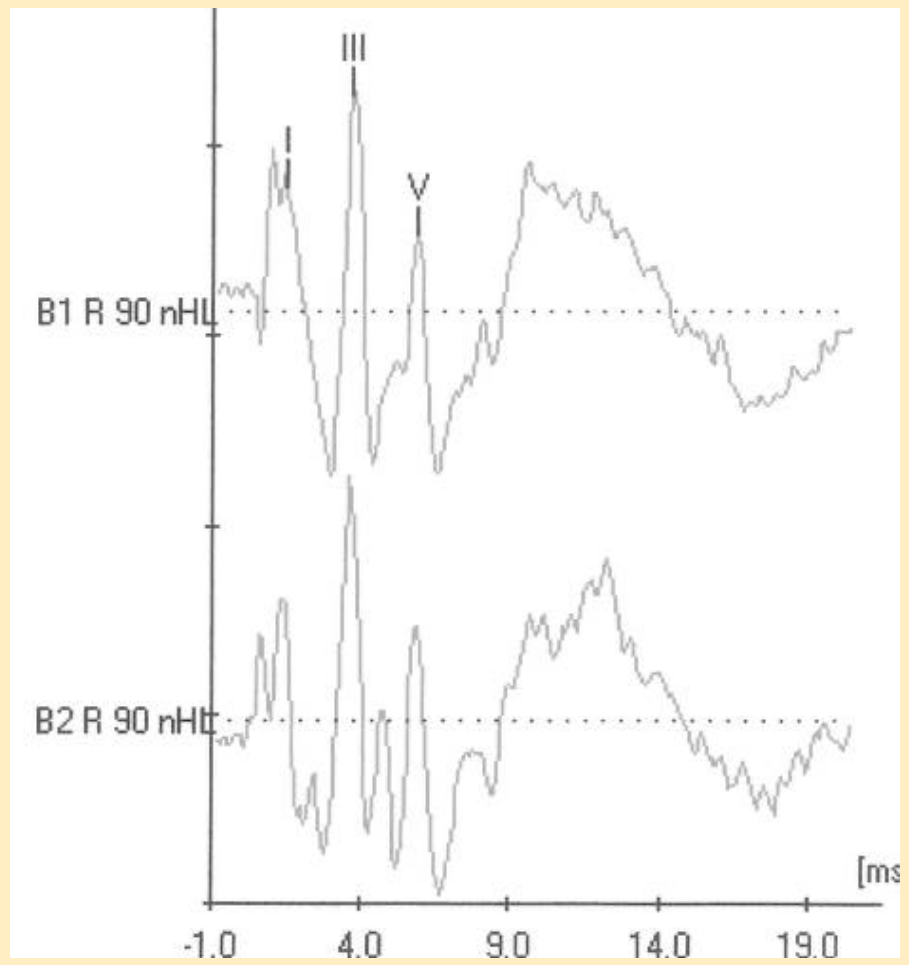
- Auditory brainstem response (ABR) evaluation
 - Click stimulus (rarefaction AND condensation)
- Otoacoustic emissions (OAE)
- Middle ear muscle reflexes
- Pure tone thresholds
- Speech recognition in noise



Diagnostic Findings in AN/AD

- **ABR: abnormal**, with only the presence of cochlear microphonic (CM)
- **OAE's: present**
- **Middle ear reflexes: absent**, in presence of otherwise normal middle ear function
- **Pure tone thresholds: variable**, range from normal to profound range
 - Speech recognition disproportionately poorer in comparison to what expected for thresholds
 - Speech in noise performance: Poor





Clicks

Right

0.20 [uV/div]

Refractation

B1 R 95 nHL

condensation

B2 R 95 nHL

[ms]

[ms]

0

-1.0

4.0

9.0

14.0

19.0



Audiological intervention

- Recommendations have evolved over the last decades
- Initially recommended using American Sign Language (ASL) or Cueing
 - at first thought to be pathology of auditory nerve



Intervention continued

- Audiologists then began experimenting with low gain hearing aids for one ear
- Also some recommendations for no amplification at all
- FM system



Audiological intervention today

- Amplification after obtaining reliable behavioral thresholds, if residual hearing
 - Hearing aids
 - FM system
 - Cochlear implant (CI)
- Communication modalities
 - Spoken language
 - ASL
 - Cued speech



How do we decide which intervention is best for these children?

- Trial traditional amplification
- Monitor speech-language milestones closely, using parent report, SLP report, standardized speech perception measures
- FM system for noisier situations
- Cochlear implantation only if lack of progress with traditional amplification



How do we decide which intervention is best for these children?

- Length of time required to evaluate benefit or lack of benefit from amplification will vary based on developmental level of child, consistency of amplification use, and clinical judgment



Why is cochlear implant an option?

- If site of lesion IHC/synaptic transmission: CI bypasses these to directly stimulate auditory nerve fibers with electric current
- If site of lesion auditory nerve – electrical stimulation much more synchronized than acoustic stimulation
- *Expectations should be set more realistically for AN/AD implantees compared to non-AN/AD implantees

Outcomes

- Variable
- Small chance auditory function can spontaneously improve during first 1-2 years of life
- Some studies suggest that less than 10% can develop normal speech-language
- Other studies show AN/AD children performing similar to peers with sensorineural hearing loss



Outcomes

- Receptive language – delayed compared to normal hearing peers, but similar to matched groups with sensorineural hearing loss (when aided)
- Variable speech production, though most intelligible



Take Home Messages

- Importance of newborn hearing screening
 - Use of multiple tests in NICU
- Importance of team evaluation [Audiology, Speech Pathology, Medical Team (ENT, Geneticist, Pediatrician)]
- Do not be afraid to try traditional amplification
 - Nor to begin trying to help child develop spoken language
 - Get behavioral information, in addition to other audiological test battery information, in order to fit hearing aids
- Monitor speech-language progression CLOSELY
- If lack of benefit, CI's are a possibility



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