Poster Highlights
ANSD Subtypes
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ANSD Guidelines

Guidelines exist for medical work-up and management

– Developed by experts in the field in 2008
– “Guidelines for Identification and Management of Infants and Young Children with Auditory Neuropathy Spectrum Disorder”
– Provide needed direction for clinicians and medical teams
– However, management remains elusive.
  – Proposal is intended to supplement current medical and behavioral guidelines.
Goal for Proposed ANSD Subtypes

Clarification for projected outcomes and expectations

Accessible and clear format

Provide clinicians with additional confidence

Improve counseling and management

Reduce parental anxiety

Reduce delays
ANSD Management Based on Etiology

• An increasing amount of research that supports the notion that management should be made with consideration for etiology.

• Dr. Santarelli, et al, concluded, “...the choice and effectiveness of assistive devices proposed for patients with AN depend critically on the mechanisms underlying the alteration of auditory nerve firing and thus, on the etiology of AN disorders.”

• In 2015, Dr. Rance and Dr. Starr reported, “The efficacy of cochlear implantation in auditory neuropathy is closely related to the site(s) of lesion.”

• Using anatomical or etiologic subtypes or descriptions, can help clarify the potential projected pathway families may take and guide audiologists.
Projected Pathways for Some Etiologies Become Clear:

– i.e. OTOF (Otoferlin) – Pre-synaptic disorder
Projected outcomes with CI are good

- i.e. CND (Cochlear Nerve Deficiency) – Often poor CI outcomes due to poor neural survival
Other Projected Pathways Remain Variable

- **Post-Synaptic:**
  - Variable outcomes
  - Conditions where the auditory nerve and its brainstem connections are affected. (Rance and Barker, 2009)
  - Demyelinating and axonal neuropathy are the two most commonly encountered neural abnormalities in patients with ANSD. (He, S, et al, 2015)

- **Neonatal Complications** (Preemies, Anoxia, Hyperbilirubinemia):
  - Variable outcomes with HA and CI (half seem to benefit form HA and/or CI)
  - There is promise for benefit with HA or CI
  - Findings suggest dysfunction of IHCs – may be underlying mechanism
  - Increase in acoustic input could possibly compensate for the loss of hair cells – depending upon number of residual receptor elements and proportion of spared OHCs and IHCs.
ANSD Projected Outcomes

Flowchart for proposed etiological approach to ANSD management
CONCLUSION

By categorizing ANSD with consideration for etiologic factors and site of lesion, a child’s team - audiologist, neurologists, otologists, speech-language pathologists, and deaf/hard-of-hearing specialists will proceed with increased confidence to carryout treatment and education plans.

Efforts to determine the site of lesion along the auditory nerve and brainstem are vital to project outcomes with cochlear implants for ANSD.

Widely available and improved techniques to identify site of lesions are needed.

With etiological guidelines, increased understanding can improve counseling and provide clearer direction on a path that is not always straightforward. As a result, this can translate into reduced language delays for children with ANSD.