Auditory Neuropathy in Children
- an Otologist’s Perspective

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Based on a Presentation by Craig A. Buchman, MD

Objectives

• What is Auditory neuropathy?
  » Etiology & Pathogenesis
  » Diagnosis of AN
  » Evaluation of AN & SNHL
  » Classification

• Management of ANSD
  » Conventional amplification
  » Cochlear implants

• Cochlear Nerve Deficiency
  » Why important for AN?
  » Diagnosis, Evaluation, Management

Auditory Neuropathy

• Term ‘Auditory Neuropathy’ first used in 1996
• Described as:
  » ‘Abnormal auditory nerve function in the presence of normal cochlear receptor hair cell activity reflected by preserved otoacoustic emissions (OAEs) and/or cochlear microphonics (CMs)’ in 2003
• Not entirely new
  » Children w/ flat ABRs who respond to sound
  » Children w/ flat ABRs who do not respond
  » Matured technology allows identification

Controversy

Multiple aspects of ANSD
• Terminology
  » Auditory neuropathy vs. Auditory dys-synchrony
  » Neural hearing loss?
  » Neural conduction disorder?
• Etiology
• Pathomechanisms
• Management

Basic Assumption

Early approach @ UNC started in 2003:
AN/AD is a heterogeneous group of disorders with a similar electrophysiological profile
• Several causes
• Variety of clinical appearance
• Different auditory perceptual abilities
• Necessitates individualized management
• Expect variable outcomes

Individualize the clinical protocol

What is Auditory Neuropathy?

Auditory Neuropathy

Interaction of Hearing Professionals

• Diagnose or confirm diagnosis
  » Single polarity click ABR, OAEs
• Imaging (MRI)
  » CNS & individual nerves
  » Observation & amplification until VRA possible
• Behavioral testing
  » Normal: close f/u, repeat ABR?
  » Abnormal: conventional amplification
  » Intensive predominately auditory based therapy
• Absent or inadequate speech & language production/speech perception
  » Cochlear implantation?

Timeline

Early Diagnostic Evaluation & Management of SNHL & ANSD

Medial Assessment

• Otologic examination
• Imaging (MRI)
• Referrals
  » Neurologic consultation
  » Genetics consultation
  » Ophthalmology consultation

What we are looking for?

• Etiology
  » Hereditary or acquired
• Associated problems
  » Seizures
  » Motor delays
  » Visual impairment
  » Ear canal problems
  » Otitis media
• Inner ear morphology
• Cochlear nerve integrity

Possible Etiologies of ANSD

…Associations

• Prematurity
• Neonatal distress
  » Hyperbilirubinemia
  » Perinatal asphyxia
  » Artificial ventilation
• Genetic abnormalities
  » OTOF, PMP2, MP2, NDRG1
  » Charcot-Marie-Tooth syndrome, Guillain-Barre syndrome
• Infectious processes
  » Viral infections (mumps, meningitis)
  » Ototoxicity
  » Head injury

UNC ANSD Cohort

- Prospective study, 10 years
  - Clinical characteristics of ANSD
  - Document benefits from assistive devices
- Infants & children, n>150
  - Large number of very small children, just beginning to see outcomes
- Evaluation
  - All ABRs performed at our institution
    - Pediatric audiologist
    - Otologist

Biographical Facts & Associations

Management of ANSD

Bilateral (n=104) vs. Unilateral (n=26)

Device/HA Utilization

No Assistive Device, n=18
Bilateral AN CI (n=45)
Bilateral AN HA (n=41)

CI ECAP Recordings

(n=30)
Why Cochlear Implants Work?

...in many cases of ANSD
- Maybe not always a neural HL?
- Synchronization of neural response?

Cochlear Nerve Deficiency

Facts about CND
- Small or absent CN
- What do we know?
  - Previously described in small IACs
  - Failed stimulation w/ CI
  - Contraindication to CI

Cochlear Nerve Deficiency
- Studies: IAC morphology (CT): diagnostic of cochlear nerve deficiency
- TB Histology: absent CN...with normal IAC & inner ear
- Recent studies: MRI can identify IAC nerves image absent CN directly

First Case
- Bilateral profound HL
- CI right w/ no NRTs®, no benefit
- CI left, normal NRTs®, 100% PBK @ 3 years

Change in Protocol
- MRI for children w/ SNHL started around 2002 @ UNC
- High resolution protocol
  - Glastonbury et al. protocol, CISS imaging
- ABR using single polarity stimuli
- Behavioral data (VRA)

Patient Characteristics I

- **Audiological Profile**
  - CM present in ~70% of ears
  - Absent neural responses
  - Profound or severe-to-profound SNHL
  - Represents ~10-15% of ANSD population!!

- **MRI Characteristics**
  - ~60% of ears have normal IAC
  - ~40% of ears have normal labyrinth
  - many of them have hypoplasic malformations
  - absent semicircular canals, small cochlea

Patient Characteristics II

- **CT Findings**
  - 19 children CT & MRI
  - ~50% have normal IAC
  - All w/ normal IAC have patent BCNC
  - CT completely normal!
  - ...in a subgroup of children
  - and thus miss CND!

Normal IACs

- **Axial HRCT, MRI, parasagittal MRI**
  - Normal BCNC
    - no stimulation after CI, absent CN, normal CV anatomy
  - Small BCNC
    - partial bony obliteration, normal CV anatomy, absent CN

Small IACs

- **Axial HRCT, MRI, parasagittal MRI**
  - Small BCNC
    - single nerve, NI ABR, normal cochlear anatomy, profound HL
  - Absent BCNC
    - single nerve, CM potential, profound HL

CND Conclusions

- ~10-15% of AN ears
  - ~70% have CM, absent distal waves
- MRI primary imaging modality
- Normal IAC: MRI identifies all cases
- Small IAC: MRI identifies the small IAC
  - HRCT further defines status of CN ~70% of ears
    - Absent BCNC - suggestive of absent CN
    - ~30% remain inconclusive
- HRCT for small IACs

For indeterminate ears: electrical ABR

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Really deficient Nerve? 
…or can we just not see it on MRI?
• CI in 5 ears w/ CND
  » All had sound awareness
  » None had ECAPs
  » None gained open-set speech perception
  » Very poor users
• Why sound awareness?
  » Non-auditory somato-sensory stimulation?

Single Nerve & Patent BCNC
• Small IAC on MRI
  » Normal facial function
  » Patent BCNC (via CT scan) indicating possibility of some auditory nerve fibers
• Implantation or not?

Conclusions

Conclusions I
• ANSD heterogeneous group
  » Multiple causes
  » Study population still very young
  » Many w/ multiple challenges
• Rarely normal hearing
• Some benefit from:
  » Amplification
  » Cochlear implantation
• Some do not benefit from HA or CI

Conclusions II
• Early involvement of otologist
  » …as part of the evaluation team!
• MRI instead of CT for screening SNHL including AN!
• CND ~10-15% of AN ears!
  » ~70% electrophysiological AN pattern!
• CT selectively
  » IAC < 3 mm, Single nerve in IAC, Cochlear obstruction
  » Semicircular canal malformation
  » To identify position of the facial nerve
  » Temporal bone pathology

Conclusions III
• Select cases remain inconclusive
  » Small IAC, single nerve, patent BCNC
  » Use everything you have when IAC is small
  » Electrical stimulation ABR for unclear ears!
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