Management of Auditory Neuropathy: Mission Impossible?

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Overview

- What is auditory neuropathy/dys-synchrony (AN/AD)?
  - How does it differ from other types of hearing loss?
  - How can I tell which children have it?
  - Does my local universal newborn hearing screening program test for this?
- What can we do to help children with AN/AD?
  - Hearing aids?
  - Cochlear implants?
  - Classroom support
Auditory neuropathy

- Evidence of normal outer hair cell function in the cochlea
  - Otoacoustic emissions (OAEs) – present
  - Cochlear microphonic response – present in auditory brainstem response (ABR)
- Evidence of neural impairment
  - ABR is abnormal
  - Acoustic stapedial reflexes are absent/abnormal
- Audiogram
  - Ranges from normal to profound (any audiogram)
Auditory neuropathy

- Not a “new” type of hearing loss
  - Clinical advent of widespread use of OAEs
  - Mid 1990s – first cases emerged in literature
- Paradoxical ABR cases prior to that
  - People with no ABR and normal hearing
  - People with much less hearing loss than the ABR would indicate
Inside your cochlea...

- Two types of sensory cells
  - Inner hair cells
    - Sensory receptors
  - Outer hair cells
    - Have motility and can expand and contract
    - Produce OAEs
In patients with AN/AD . . .

- Inner hair cells (IHC)
  - Neural transmission is impaired
  - Abnormal/absent ABR
  - Could be IHC problem

- Outer hair cells (OHC)
  - Normal
  - Produce OAEs and/or cochlear microphonic
Site of Lesion

- Neuropathy?
  - Myelin or conduction problem
  - Associated with other neuropathies
- Inner Hair Cells or synapse?
- Spiral Ganglion or Axons
- Tectorial Membrane?
- Neurotransmitters?
- Clinical presentation does not differentiate
Why call it neuropathy?

- Original group of patients (Starr, 1996)
  - Adults with additional peripheral neuropathy
- Current group, including children –
  - Only 30-40 % have other sensory neuropathies
- Change the name of the disorder?
  - Auditory dys-sychrony?
  - Auditory neuropathy?
  - Neural hearing loss?
Auditory neuropathy?

- Currently available tests do not differentiate site of lesion
- Patients with AN are heterogenous in their auditory performance
  - Likely related to varying degrees of impairment across various sites of lesion
- More specific tests and imaging may help us to be more specific
How common is AN/AD?

- 2 to 3 children in 1000 live births has permanent hearing loss
- Estimates of AN/AD range
  - Foerst et al. 2006
    - 0.94% (Children at-risk for hearing loss)
    - 8.4% (Children with profound hearing loss)
- ~ 1.4 in 10,000 live births has AN/AD
Implications for universal newborn hearing screening

- Most hospital use OAEs only (saves $)
- Babies with AN/AD have normal OAEs
- OAEs alone will not identify AN/AD
- ABR screening will identify AN/AD
- Cost difference justified by small numbers?
Pediatricians can -

- Know the type of hearing screening test (OAE or ABR) used at your local hospitals
- Recognize that OAE only screening programs will miss every child with AN/AD
- Refer children with hearing or speech and language concerns to audiologist
  - Even with a normal newborn hearing screening
Most Common Causes of AN/AD in Children

- Anoxia
- Hyperbilirubinemia
- Infectious Disease (Mumps)
- Immune Disorders
- Non-syndromic Recessive AN/AD

15 – 25% of AN/AD cases – no risk factors
Hyperbilirubinemia / Kernicterus

- Commonly reported in cases of AN/AD
- Extent of neurological effects may depend on:
  - TSB Levels
  - Duration of exposure
  - Gestational age
  - Infection
  - Pre-maturity
  - Genetic susceptibility (?)
- Shapiro (2005) suggests ≥ 20 mg/dl TSB as possible criteria for further studies.
- Cases of AN exist in patients with < 20 mg/dl.
Reported Non Genetic Causes of AN/AD

- **Systemic/Metabolic**
  - Bilirubinemia/ Kernicterus
  - Diabetes mellitus
  - Uremia
  - Alcoholism/nutritional
  - Paraproteinemias
  - Anoxia

- **Infection/Inflammatory**
  - Leprosy
  - AIDS
  - Lyme Boreliosis

- **Toxins**
  - Drugs (cisplatin)
  - Heavy metals
  - Vascular diseases
  - Neoplasia
  - Trauma

- **Sarcoidosis**
- **Polyarteritis nodosa**
- **Rheumatoid Arthritis**
- **Ramsey Hunt syndrome**
- **Gullain-Barre Syndrome**
- **Vascular diseases**
- **Neoplasia**
- **Trauma**
Genetic Causes of Auditory Neuropathy/Dys-synchrony

- Adrenoleukodystrophy
- Amyloid polyneuropathy
- Charcot-Marie-Tooth 1A, 1B, 2, 2a, 2B, 4A, 4B, 2D, X
- Complex I, subunit ND4
- Dejerine-Sottas syndrome
- Friedreich Ataxia
- H Sensory Neuropathy type 1
- Hereditary myelinopathy
- HMSN (Lom)
- HMSN and deafness
- HNPP
- Infantile-onset spino-cerebellar ataxia
- Leber optic atrophy
- Moebius syndrome
- Mohr-Tranebjaerg syndrome
- Myopathy, Distal 2C
- Neuraminidase deficiency
- Neurofibromatosis type 2
- NMSN, type II, with deafness
- Phosphoribosyl PPi synthase I
- Refsum disease (HSMN IV)
- Refsum disease, infantile form
- Spastic paraplegia 8
- tRNA, mitochondrial, lysine
- Wolfram syndrome
Etiology of AN

- Multiple possible sites of lesion
- Multiple etiological factors:
  - Infectious
  - Teratogenic
  - Genetic
  - Traumatic
  - Interactions may exist
- Multiple etiologies may explain variability of auditory skills
Pediatricians can -

- Recognize risk factors
- Refer children with significant risk factors for an ABR, if OAE was only hearing screening
  - Particularly children with extended periods of high TSB (> 20 mg/dl)
When to refer older children

- Hearing concerns
- Concerns for speech and language development
- History of other neuropathies or neurological diagnoses
- Family history of childhood hearing loss
Intervention

- Based on individual auditory skills
- Trial with hearing aids is essential
- Other medical needs may take precedence
- Goal: provide access to language and child’s environment in most efficacious manner
- Evidence base is growing – much work left to do
Amplification – Hearing Aids

- First step in intervention process
- ABR generally provides threshold data
  - In AN/AD patients, ABR is always absent or abnormal at high levels
  - Does not give threshold data?

- How do we know how to set the hearing aids?
Amplification – Hearing Aids

- Unfortunately, must wait to have behavioral threshold data prior to fitting hearing aids
  - May be 6 – 12 months (or later) before this data is reliably available
- Contradicts all philosophies related to early identification and intervention with hearing loss
- Audiogram may range from normal to profound hearing loss
- 50% of children with AN/AD receive benefit from amplification (Rance, 2005)
Cochlear Implants

- If hearing aids are not effective, cochlear implantation may be an option.
- If you have a nerve problem, how does a cochlear implant help?

**Figure 11–18.** Electrical auditory brain stem response recording for Child B.
CI and AN: Peterson et. al., 2003

- 10 children with AN/AD and CIs
- Matched to 10 control children with CIs
- No significant outcome differences
  - Aided audiograms
  - Speech perception measures
  - Educational placement/communication mode
  - EABRs
CI: The Universal Cure?

- Some reports of lack of benefit from CI
  - Miyamoto et al, 1999; Rance et al, 1999; Lesinski-Schiedat et al, 2001
- Additional neuropathies/medical complications
  - Charcot-Marie-Tooth disease
  - Friedreich ataxia
  - Demyelination or axonal damage
- Mild to moderate hearing loss per audiogram?
- Recovery of function? (Berlin et.al., 1999)
Cochlear Nerve Deficiency

- Buchman et al. 2006 Ear and Hearing
- Nerve is small or absent on MRI
  - May have narrow IACs
  - May have normal IAC diameter
- Present in 9/51 AN patients (18%)
- Consideration for CI
  - Poor CI outcomes
  - MRI (CT may be normal)
Support

- FM systems in classroom environments
- Sign language
- Cued speech
- Captioned telephone
Pediatrician’s Toolkit for Auditory Neuropathy

- Recognize tests used for newborn hearing screening
  - OAE vs. ABR
- Refer infants with risk factors, if OAE is only screening available
- Children with unexplained speech and language concerns should have a hearing evaluation
Pediatrician’s Toolkit for Auditory Neuropathy

- Treatment approach is based on individual auditory skills and needs
- Trial with hearing aids should be initiated once behavioral audiometric results are obtained
- Cochlear implantation can be an effective solution
  - MRI should be performed to rule out VIIIth nerve deficiency
Questions / Discussion ?

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