

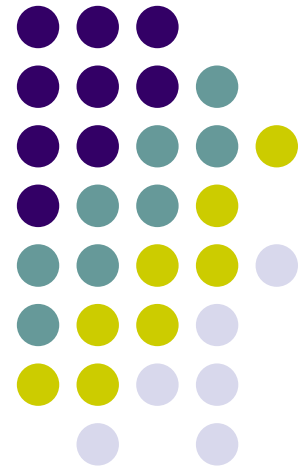
Management of Auditory Neuropathy: Mission Impossible?

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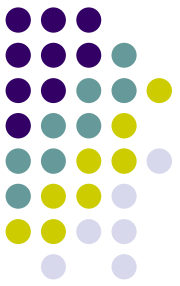
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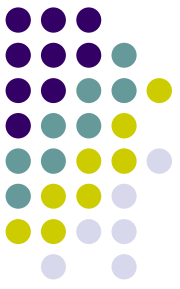
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Overview

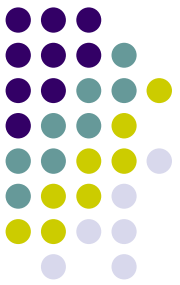


- What is auditory neuropathy/dys-synchrony (AN/AD)?
 - How does it differ from other types of hearing loss?
 - How can I tell which children have it?
 - Does my local universal newborn hearing screening program test for this?
- What can we do to help children with AN/AD?
 - Hearing aids ?
 - Cochlear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

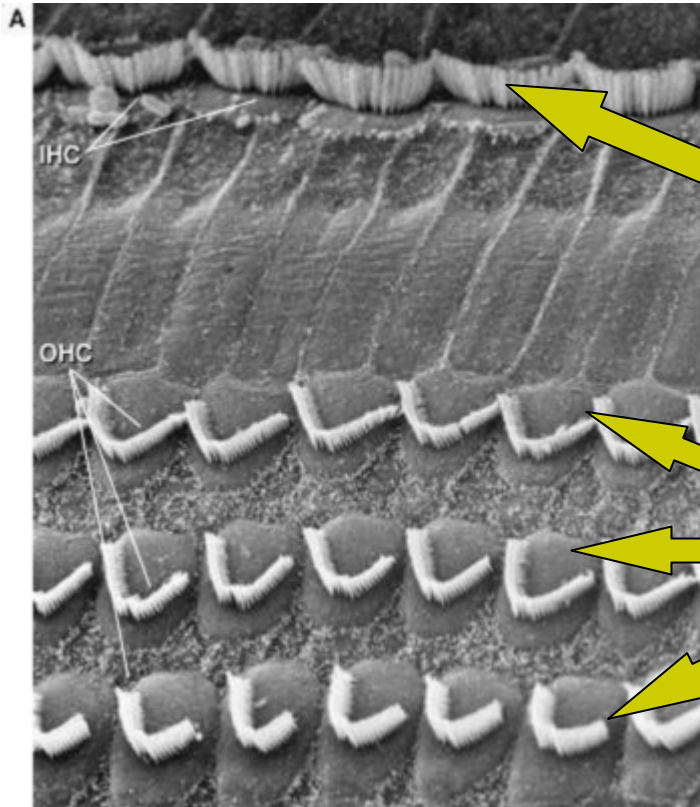


Auditory neuropathy

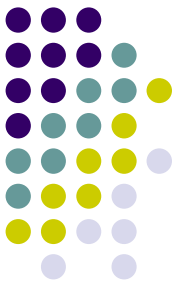
- Not a “new” type of hearing loss
 - Clinical advent of widespread use of OAEs
 - Mid 1990s – first cases emerged in literature
- Paradoxical ABR cases prior to that
 - People with no ABR and normal hearing
 - People with much less hearing loss than the ABR would indicate



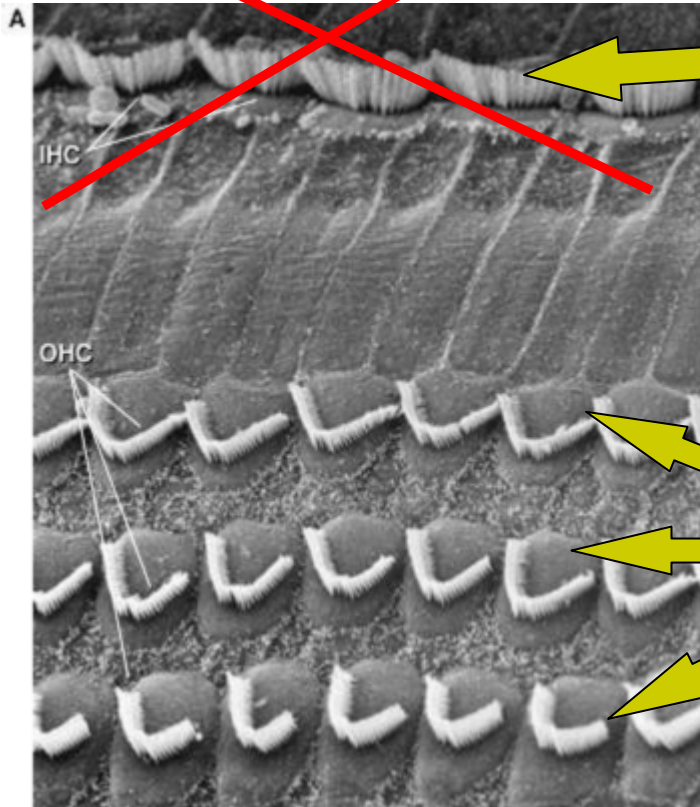
Inside your cochlea...



- Two types of sensory cells
 - Inner hair cells
 - Sensory receptors
- Outer hair cells
 - Have motility and can expand and contract
 - Produce OAEs

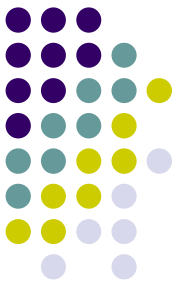


In patients with AN/AD . . .

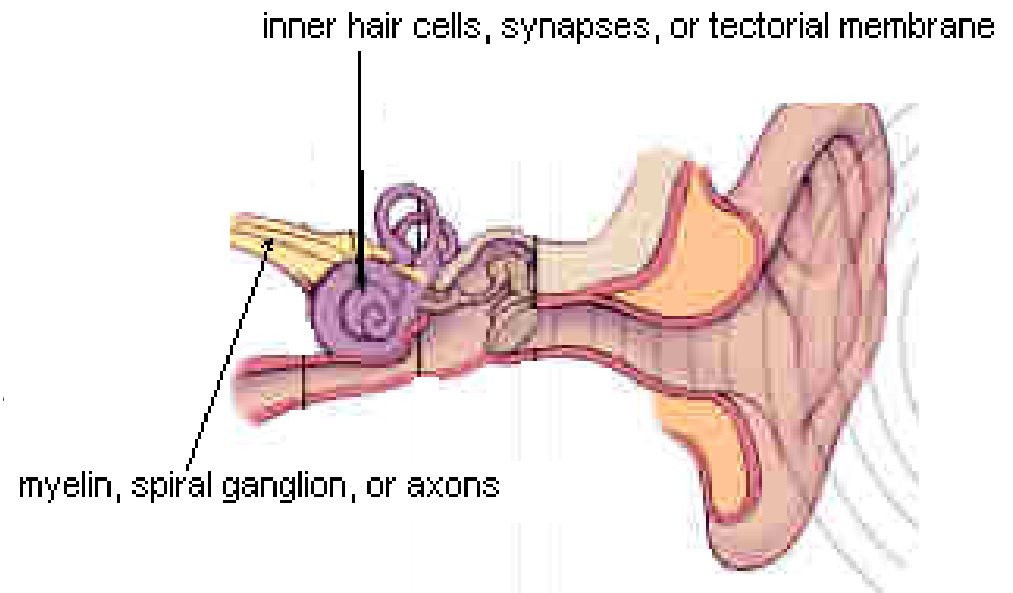


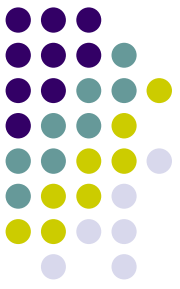
- Inner hair cells (IHC)
 - Neural transmission is impaired
 - Abnormal/absent ABR
 - Could be IHC problem
- Outer hair cells (OHC)
 - Normal
 - Produce OAEs and/or cochlear microphonic

Site of Lesion



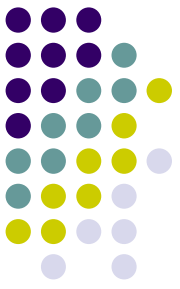
- Neuropathy?
 - Myelin or conduction problem
 - Associated with other neuropathies
- Inner Hair Cells or synapse?
- Spiral Ganglion or Axons
- Tectorial Membrane?
- Neurotransmitters?
- Clinical presentation does not differentiate





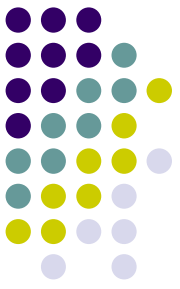
Why call it neuropathy ?

- 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-synchrony ?
 - Auditory neuropathy ?
 - Neural hearing loss ?



Auditory neuropathy ?

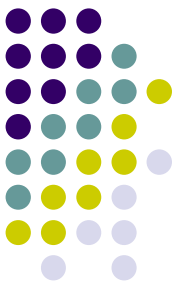
- Currently available tests do not differentiate site of lesion
- Patients with AN are heterogenous in their auditory performance
 - Likely related to varying degrees of impairment across various sites of lesion
- More specific tests and imaging may help us to be more specific



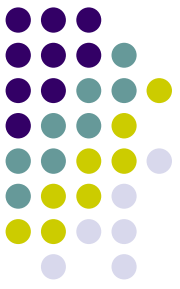
How common is AN/AD ?

- 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



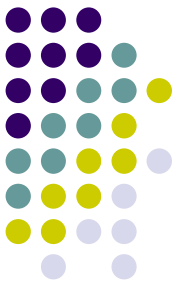
- Most hospital use OAEs only (saves \$)
- Babies with AN/AD have normal OAEs
- OAEs alone will not identify AN/AD
- ABR screening will identify AN/AD
- Cost difference justified by small numbers ?



Pediatricians can -

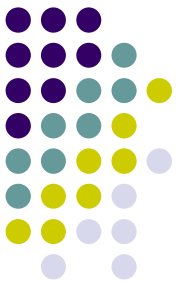
- Know the type of hearing screening test (OAE or ABR) used at your local hospitals
- Recognize that OAE only screening programs will miss every child with AN/AD
- Refer children with hearing or speech and language concerns to audiologist
 - Even with a normal newborn hearing screening

Most Common Causes of AN/AD in Children



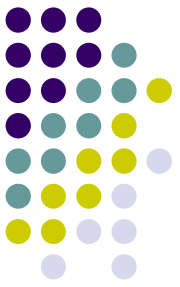
- Anoxia
- Hyperbilirubinemia
- Infectious Disease (Mumps)
- Immune Disorders
- Non-syndromic Recessive AN/AD
- 15 – 25% of AN/AD cases – no risk factors

Hyperbilirubinemia / Kernicterus



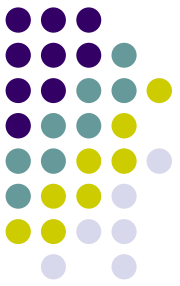
- Commonly reported in cases of AN/AD
- Extent of neurological effects may depend on:
 - TSB Levels
 - Duration of exposure
 - Gestational age
 - Infection
 - Pre-maturity
 - Genetic susceptibility (?)
- Shapiro (2005) suggests ≥ 20 mg/dl TSB as possible criteria for further studies.
- Cases of AN exist 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
- Sarcoidosis
- Polyarteritis nodosa
- Rheumatoid Arthritis
- Ramsey Hunt syndrome
- Gullain-Barre Syndrome
- ***Toxins***
- Drugs (cisplatin)
- Heavy metals
- Vascular diseases
- Neoplasia
- Trauma

Genetic Causes of Auditory Neuropathy/Dys-synchrony



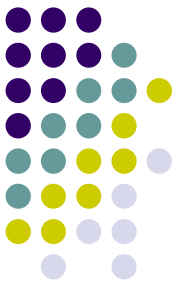
- Adrenoleukodystrophy
- Amyloid polyneuropathy
- Charcot-Marie-Tooth
1A,1B,2,2a,2B,4A,4B,2D,X
- Complex I, subunit ND4
- Dejerine-Sottas syndrome
- Friedreich Ataxia
- H Sensory Neuropathy type 1
- Hereditary myelinopathy
- HMSN (Lom)
- HMSN and deafness
- HNPP
- Infantile-onset spinocerebel. ataxia
- 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

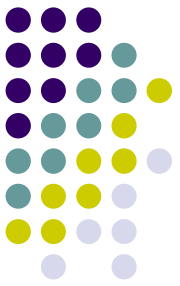
- Multiple possible sites of lesion
- Multiple etiological factors:
 - Infectious
 - Teratogenic
 - Genetic
 - Traumatic
 - Interactions may exist
- Multiple etiologies may explain variability of auditory skills

Pediatricians can -

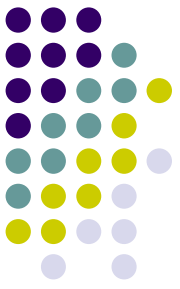


- Recognize risk factors
- Refer children with significant risk factors for an ABR, if OAE was only hearing screening
 - Particularly children with extended periods of high TSB (> 20 mg/dl)

When to refer older children

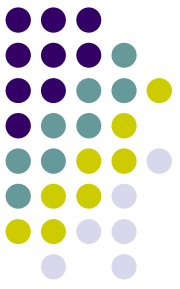


- Hearing concerns
- Concerns for speech and language development
- History of other neuropathies or neurological diagnoses
- Family history of childhood hearing loss



Intervention

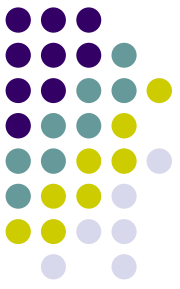
- Based on individual auditory skills
- Trial with hearing aids is essential
- Other medical needs may take precedence
- Goal: provide access to language and child's environment in most efficacious manner
- Evidence base is growing – much work left to do



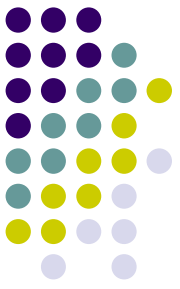
Amplification – Hearing Aids

- 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 ?

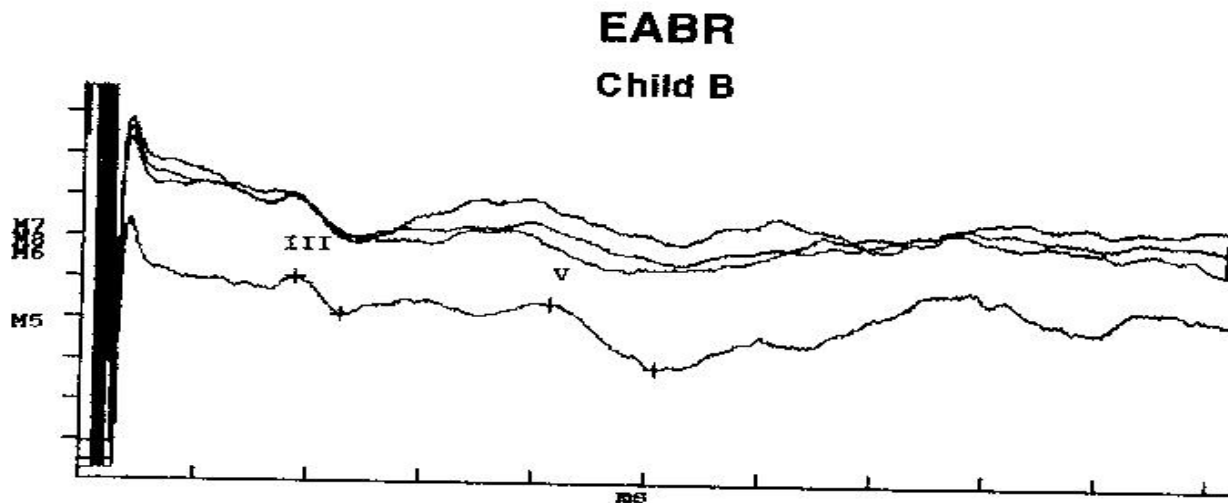
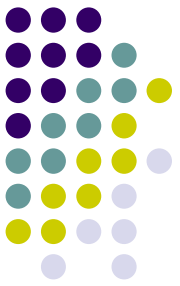


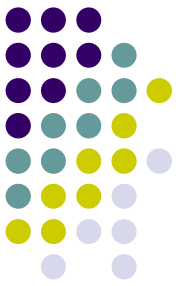
Figure 11-18. Electrical auditory brain stem response recording for Child B.

CI and AN: Peterson et. al., 2003

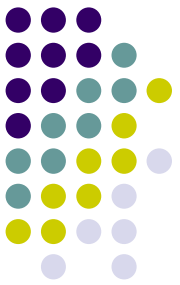


- 10 children with AN/AD and CIs
- Matched to 10 control children with CIs
- No significant outcome differences
 - Aided audiograms
 - Speech perception measures
 - Educational placement/communication mode
 - EABRs

CI: The Universal Cure?



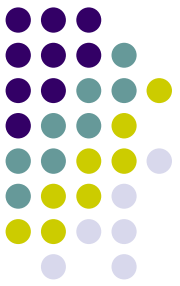
- Some reports of lack of benefit from CI
 - Miyamoto et al, 1999; Rance et al, 1999; Lesinski-Schiedat et al, 2001
- Additional neuropathies/medical complications
 - Charcot-Marie-Tooth disease
 - Friedreich ataxia
 - Demyelination or axonal damage
- Mild to moderate hearing loss per audiogram?
- Recovery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



Cochlear Nerve Deficiency

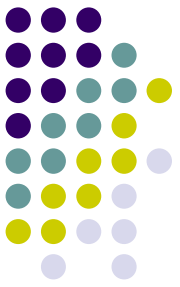
- Buchman et al. 2006 Ear and Hearing
- Nerve is small or absent on MRI
 - May have narrow IACs
 - May have normal IAC diameter
- Present in 9/51 AN patients (18%)
- Consideration for CI
 - Poor CI outcomes
 - MRI (CT may be normal)

Support



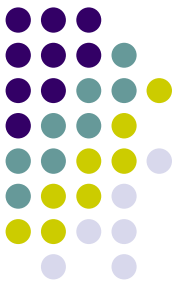
- FM systems in classroom environments
- Sign language
- Cued speech
- Captioned telephone

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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