Auditory Neuropathy/
Dys-synchrony:
Shades of Gray

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Auditory Neuropathy

...term to characterize a hearing disorder in which patients presented with:

- Evidence of poor auditory neural function
- Evidence of poor auditory function
- Evidence of present hair cell function

- Incidence widely variable ranging from 1-20% of HI population

- HI = involvement of auditory nerve either as part of generalized neuropathy or isolated auditory nerve disorder

Starr et al, 1996
Auditory Neuropathy

- Term “morphs”: auditory neuropathy/dys-synchrony or AN/AD
- Important because the auditory nerve is not always affected always affected
Six Categories of AN/AD

- **Berlin, 2001**
  1. Children with absent ABR but otherwise normal hearing ability who develop speech and language
  2. Children with OAEs and CM initially, but disappear over time; behavior consistent with severe-profound profound HL—hearing function may appear improved at times
  3. Similar to #2, but pt. functions as severely impaired majority of time; OAEs eventually disappear/CM activity remains
Six Categories of AN/AD

- Berlin, 2001
  - 4. Pts with no ABR and behavior consistent with profound loss
  - 5. Children “normal” at birth that develop problems w/hearing, speech, language and are later dx with AN/AD as part of a general peripheral neuropathy (Charcot-Marie Tooth)
  - 6. Adults with no ABR but otherwise normal auditory and language function
Possible Causes of AN/AD

- May be multiple underlying causes
  - Abnormality in the synapse between primary neurons and IHC leads to temporal “jitter”; nerves are no longer phase locked to the stimulating waveform
  - Loss of function of IHCs and/or auditory neurons so that fewer or no spikes are evoked in the auditory nerve; consistent with the fact that some pts have near-normal thresholds since only a few functioning IHCs are required
  - Might be associated with “patchy” dead regions over a large part of the cochlea
Etiology of AN/AD

- **Yes:** Hyperbilirubinemia; perinatal asphyxia; prematurity; ototoxicity; family history; consanguinity; other neuropathies

- **Maybe:** IVF-6 of 26 (Raveh et al, 2007)

- **No:** 25-35% w/no known risk factors for AN/AD
Communication Characteristics: Shades of Gray

- Inconsistent response to sound (*but some exhibit consistent response to sound*)
- Speech understanding poorer than predicted by audiogram (*but not always*)
- Speech understanding poor in presence of background noise (*OK…almost always*)
- Often difficult to learn spoken language through listening alone (*but can happen*)
- Range of vocal quality (*can vary day to day*)

Sininger & Starr, 2001
AN/AD

Rapin and Gravel, 2003 & 2006

- “urge that the term auditory neuropathy be reserved for demonstrable involvement of 8th nerve as a whole or selective involvement of the spiral ganglion cells or their processes”

- “should not be used for pathologies of uncertain or mixed locations”
<table>
<thead>
<tr>
<th>Anatomic Site of Pathology</th>
<th>Proposed Nomenclature</th>
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</thead>
<tbody>
<tr>
<td>Hair Cells</td>
<td>Sensory Hearing</td>
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<tr>
<td>Spiral Ganglion Cells/VIII nerve</td>
<td>Auditory Neuropathy</td>
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<tr>
<td>Spiral ganglion cells/VIII nerve and/or central auditory pathway (when locus of pathology is undetermined)</td>
<td>Neural Hearing Loss</td>
</tr>
<tr>
<td>Hair cells and/or spiral ganglion cells/VIII nerve and/or central auditory pathway (when locus of pathology is undetermined)</td>
<td>Sensorineural Hearing Loss</td>
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<td>Rapin and Gravel, 2006</td>
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</table>
Protocol for Assessment and Management

- ABR; tymps, ART, OAEs, Case Hx
- Cochlear nerve MRI (absent/deficiency)
- Developmentally appropriate behavioral/speech perception/language assessment at frequent intervals (every 3 mos)
- Once behavioral sensitivity is established, amplification trial (DSL, etc.) w/counseling and monitoring
- Genetics, Opthalmology, Otology, EI
Protocol for Assessment and Management

Hearing Aids: Benefit or Not?

- Timeframe varies due to:
  - Developmental level
  - Consistency of amplification use
  - Clinical “wavering” of professionals
  - Progress in Speech and Language Development
  - Results of subjective evaluations (ITMAIS, ELF, etc.)
Protocol for Assessment and Management

Cochlear Implants: No, Maybe, Yes

- No: Early cases of AN thought to be due to poor function of the VIII cranial nerve
- Maybe: Due to results of pts implanted prior to OAEs
- Yes: Outcomes vary, but are similar to those w/SNHL
Cochlear Implants and AN/AD

Why does it work?

May bypass the site of lesion (IHC, synaptic junctions)

Electrical stimulation may restore synchronous firing of cochlear nerve

Post Implant: EABR and electrically evoked stapedial reflexes indicates that neural synchrony has been enhanced/achieved
Case 1

- 8 months old w/ hx of NICU stay
- Congenital anemia, hyperbilirubinemia (double volume exchange transfusion, peak direct bili level of 24.4)
- Passed NBHS Phase I (OAEs); referred Phase II (AABR)
B.W. - 1st AER

- Diagnostic eval at 3 months
  - Present OAEs, Absent Acoustic Reflexes, ABR
  - Re-eval in 3 months
B.W. - 2nd AER
B.W. - behavioral

**FREQUENCY IN HERTZ**

**EXAMER:**

Jan S. 1-02-08

**REFERRAL:**

AUD: EXT - Report AER

**CMT:**

COULD NOT TEST

**MCL:**

UNCOMFORTABLE LEVEL

**NR:**

COULD NOT TEST

**MOST COMFORTABLE LEVEL**

**SRT:**

NO RESPONSE

**SPR:**

SPEECH RECEPTION THRESHOLD

**A:**

BOTH EARS

**SAT:**

SPEECH AWARENESS THRESHOLD

**A:**

RIGHT EARS

**NB:**

NARROW BAND NOISE

**AE:**

OTOACUSTIC EMISSIONS

**RIGHT**

**LEFT**

**AIR**

X

**BONE**

<

> 

**BONE MASKED**

<

> 

**SOUNDFIELD**

S

**UNAIDED RESPONSES**

U

**COCHLEAR IMPLANT**

C

**BAHA**

B

**EVOKE POTENTIALS**

EPa

EPb

**TEST CONDITIONS**

PROCEDURE:

• COR / VRA

• PLAY

• CONVENTIONAL

RELIABILITY:

• GOOD

• FAIR

• POOR

TRANSDUCER:

• PHONES

• INSERTS

• SPEAKER

**NEWBORN**

PASS

FAIL

**SCREEN**

PASS

FAIL

**SPEECH AUDIOMETRY**

<table>
<thead>
<tr>
<th>EAR</th>
<th>SRT / SAT</th>
<th>Aided SRT / SAT</th>
<th>Word Recognition</th>
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<tbody>
<tr>
<td>RIGHT</td>
<td></td>
<td></td>
<td>% CORRECT / LEV.</td>
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<tr>
<td>LEFT</td>
<td></td>
<td></td>
<td>% CORRECT / LEV.</td>
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<tr>
<td>SF</td>
<td>65?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>BC</td>
<td>40</td>
<td></td>
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**TYMPANOMETRY**

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<th>Right</th>
<th>Left</th>
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</table>
| 226 Hz / 1K Hz | C  | B
| Volume       | 0.4 | 0.7 |
| Peak         | -240 | -9.5 |
| Static Compliance | 0.3 | 0.1 |
Behavioral testing limited
Parents note some responses to louder stimuli – but not much.
Began trial with mild gain amplification and speech/language therapy
Monitor and adjust!
Case 2

- (Foster Care) possible preterm birth, no prenatal care, exposure to cocaine/methamphetamines in utero, abnormal external ear shape
- Failed NBHS in right ear
- Previously diagnosed with suspected AN/AD in another state; mild bilateral amplification fit, inconsistent use
E.C.

- Initial visit at ACH – 15 months:
  - CT scan, repeat AER, behavioral, speech/lang. eval
  - Present OAEs, absent ABR - left ear
  - Absent OAEs/ABR – right ear
### Speech Audiometry

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<tr>
<td>BC</td>
<td>30?</td>
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### Tympanometry

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<tr>
<td>Volume</td>
<td>6.8</td>
</tr>
<tr>
<td>Peak</td>
<td>-20</td>
</tr>
<tr>
<td>Static Compliance</td>
<td>0.5</td>
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E.C.

- Now has full-time HA use (mild gain on left; fit to loss on right)
- Currently getting speech tx 3xs month
- Recent MRI – normal eighth nerve on left; absent/deficient eighth nerve on right
- CI has been discussed, but due to MRI findings and recent acquisition of full-time HA use/tx, no decision has been made yet.
E.C.

Plans:

- Continue to monitor rate of speech/language development
- Possibly increase amount of speech/language therapy
- Remove right HA (no VIIIth nerve)
- Consider CI
K.M.

Case 3

- 6 year old female
- Normal hearing, speech/language, development
- Seen as in-pt initially with hx of viral cerebellar ataxia
- Unsteady gait
- Unable to understand parents following viral attack
K.M.
K.M.

- Mild/moderate behavioral hearing loss
- Cannot obtain SAT/SRT
- Present OAEs bilaterally
- Absent/elevated Acoustic Reflexes
K.M.

Absent ABR - AN/AD
K.M.

- Due to sudden onset – conservative approach
- Trial with personal FM
- Repeat behavioral testing
- Speech/language tx (short-term)

- Almost one year out – no noted improvement
  - Just began trial with bilateral amplification
  - CI has been discussed, but parents very uncertain at this time
  - Discussed further neurological/genetic testing
Case 5

- Ten year old male who referred following hearing screening with teacher concerns
  - OAE present and WNL, AU
  - Type A tympanograms, AU
  - Audiometric results
    - Hearing within normal limits, AD
    - No reliably, obtained thresholds, AS
- “normal” “malingering”???
**ABR:**

- Right ear: good wave morphology and absolute latency of components in the expected range
- Left ear: poor wave morphology, inverting CM when stimulus polarity reversed

**Reflexes:**
- MEMR absent ipsi, elevated contra, AS
- MEMR present ipsi, absent contra, AD
B.A.

- **Recommendations**
  - **Classroom**
    - Optimal seating
    - Confirmation of understanding what has been said
    - Additional visual aids/media to supplement the spoken information of the lesson
  - **Medical**
    - Referral to see otologist
    - Follow-up re-evaluation
    - Trial use of an earplug
    - Lost to f/u
S.C.

Case 6

Complaints:
- Trouble listening
- Trouble in school
- TV up
- Some days better/worse

- Currently in a facility for teenagers with emotional disorders/hx of sexual abuse
- Initial school problems considered to be related to ESL (moved to US at 6) although bilingual
- Educated, concerned family
S.C.

- SRTS at 40-50 dB
- PT essentially WNL
- Malingering?
- Absent/elevated reflexes
- Present OAEs
- Reports speech as “bzup bzup, bzup bzup”
S.C.

Outcomes:

- Family/individual/staff counseling
- Trial comparing HA and personal FM
- Preferred FM
- Unanticipated environmental interference
Auditory Neuropathy/
Dys-Synchrony

Summary

- The cross check principle continues to be the gold standard “only the rules are different”

- The one constant about AN/AD/?? is variability!