Differential Assessment and Management of Auditory Neuropathy in a Premature Infant

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- 33,000 outpatient visits to ENT clinic/year
- Approximately 1000 infants and children with hearing loss followed at UNC
  - ~500 using amplification
  - ~500 with cochlear implants
Characteristics of Auditory Neuropathy/Auditory Dysynchrony

● Electrophysiologic Findings
  – Absent or grossly abnormal ABR with presence of cochlear microphonic
  – Otoacoustic emissions may be present but may disappear over time
  – Absent acoustic reflexes

● Clinical Characteristics
  – Speech recognition abilities poorer than expected for degree of hearing loss
  – Pure tone thresholds vary from normal to profound
Auditory Neuropathy/Dysynchrony: Possible Etiologies

- Selective inner hair cell loss
- Synaptic disorder
- Myelin disorder
- Auditory nerve disorder (e.g. with other peripheral neuropathies)
Case #1

- First child born at 25 weeks gestation
- Ventilated for 6 weeks
- Oxygen 3 ½ months
- Hyperbilirubinemia
  - Treated with lights, exchange transfusion
- Treated with antibiotics and diuretics
- Hospitalized 4 ½ months
- No family history of hearing loss
- Did not pass newborn hearing screen at hospital discharge
Age 6 Weeks:
Hospital #1 First Diagnostic Auditory Brainstem Response (ABR) Evaluation

- “ABR consistent with severe to profound sensorineural hearing loss bilaterally”
- Repeat testing recommended one week later to confirm results
Age 7 Weeks:
Hospital #1 Second Diagnostic ABR

- ABR showed no response in either ear at maximum intensity levels to click stimuli or to tone bursts at 500Hz, 1000Hz, 2000Hz or 4000Hz

- Recommendations:
  - Referred to otologist for medical clearance for hearing aids
  - Fitted with high gain hearing aids following otologic exam
First Contact with UNC Hospital
Age 5 months (1.5 Months Adjusted Age)

- Mother makes first phone contact with UNC explaining family is moving to our area, requests appointment for ear impressions.
- Mother counseled that repeat ABR needed even though child has already had two ABRs.
- Mother concerned that earmolds too small so appointment scheduled prior to ABR to take ear impressions.
Mother’s Comment During Appointment for Ear Impressions:

– “It’s strange, I know he’s been diagnosed as being profoundly deaf but the other day I dropped the remote control on the TV and he startled.”
Age 6 Months (2 1/2 Months Adjusted Age): UNC Evaluation

- ABR at high intensity level with both rarefaction and condensation clicks shows only a cochlear microphonic with no evidence of neural response
- OAEs absent
ABR Obtained at UNC, Age 6 Months (2 1/2 Months Adjusted Age)
Family Meeting to Discuss Test Results:

- ABR results consistent with AN/AD
- Explained that in cases of AN/AD ABR is not useful in predicting hearing threshold levels
- Recommended discontinuation of high gain hearing aids until behavioral audiometry can be obtained
- Discussed controversy surrounding treatment options, (amplification, communication strategies etc.) as well as the approach to AN/AD our clinic uses
- Scheduled otologic exam
- Referral for early intervention services
Otologic Exam

- Normal otoscopic examination
- Referrals made:
  - MRI → No CNS problems, inner ears normal
  - pediatric neurology → normal
  - ophthalmology → normal
  - genetics → no abnormalities detected
Age 9 Months (5 1/2 Months Adjusted Age)  
First Attempt at Visual Reinforcement Audiometry (VRA):

- Child beginning to perform VRA task but responses probably “suprathreshold”
- Acoustic Immittance:
  - Right: flat tympanogram, Left: normal
- OAEs:
  - Right: not tested
  - Left: absent
Age 10 Months (6 1/2 Months Adjusted Age): Behavioral Audiometry with VRA

- Sound Field Audiogram:
  - moderate hearing loss for “better ear”
- Bone conduction thresholds confirm sensorineural HL
- Acoustic Immittance:
  - Right: normal
  - Left: normal
- Discussion with family
  - Decision made to proceed with amplification
Child fitted with new binaural digital hearing instruments with ear level FM receivers
- Appropriate for moderate degree of hearing loss
- Set to match DSL targets

Parents counseled to carefully observe child’s behavior and notify us if any signs of loudness discomfort

Return appointment scheduled to continue assessment and obtain individual ear measures
Age 10 ½ Months (7 Months Adjusted Age)
Hearing Aid Fitting
Verification Measures Using DSL Targets
Age 12 Months (8 1/2 Months Adjusted Age): VRA with Insert Earphones Attached to Child’s Earmolds

- **Speech Detection Thresholds:**
  - Unaided:
    - Right 40dBHL, Left 45dBHL
  - Aided
    - 20dBHL
- **Tympanometry**
  - Right: normal
  - Left: normal
- **Sound field audiogram (unaided and aided) completed for demonstration to parents**
- **Parental Report:**
  - Child began babbling with consonant sounds in past week: e.g. la, la, la, da, da, da
Age 14 Months (10 1/2 Months Adjusted Age): VRA with Insert Earphones

- SDTs:
  - Unaided
    - Right: 55dBHL
    - Left: 55dBHL
  - Aided
    - 25dBHL

- Tympanometry
  - Right: Type C
  - Left: Type C
Age 18 Months (14 1/2 Months Adjusted Age): VRA with Insert Earphones

- Tympanometry
  - Right: Type A
  - Left: Type A
- Parental Report:
  - Child now using a few words and beginning to understand simple commands
Age 24 Months (20 ½) Months Adjusted Age): VRA with Insert Earphones

- Child conditioned for play audiometry procedure but limited attention span
  - Results similar to previous audiograms

- Tympanometry
  - Right: normal
  - Left: normal
Age 24 Months (20 1/2 Months Adjusted Age): Communication Status

- **Parental Report:**
  - Child understands several words, using two word combinations
  - Comprehension of language seems very good

- **Early Speech Perception Test (ESP) administered**
  - Aided (auditory only condition) at 50dBHL:
    - Able to accurately identify from closed set of objects for spondee and monosyllabic words
Key Points

- Important to listen to family’s description of how child responds to sound
- May be value in repeating ABR especially if unfamiliar with testing facility where previously obtained
- If necessary to rely on ABR from another facility, ask to see waveforms and evaluate quality of test results and presence of cochlear microphonic
- Importance of testing with high level click stimuli with reversed polarity (condensation and rarefaction)
Key Points

- Electrophysiologic measures are critical, but including behavioral audiometry in test battery is essential for optimal management.
- Middle ear status must be closely monitored and managed.
- Adequate medical work-up essential (e.g. pediatrics, otolaryngology, genetics, ophthalmology, neurology).
- Important to evaluate aided speech perception abilities as soon as possible.
- Communication with early intervention specialists familiar with sensory hearing loss and knowledgeable about AN/AD essential.
Hearing Aids in Children with AN/AD

Rance et al Ear and Hearing 2002
AN and Cochlear Implants at UNC (n=18 children)

- Age at Implantation: Mean 3.25 yrs (10 mo-10yrs)
- Duration of Implant Usage: Mean 1.91 yrs (6mo-5 yrs)
Characteristics of UNC Patients with Auditory Neuropathy/Auditory Dysynchrony

- 60 infants and children
  - 23 hearing aids
  - 26 cochlear implants
  - 11 in evaluation process
- 29/60 premature (48%)
- Other problems:
  - Peripheral neuropathies
  - Seizure disorders
  - Family history of hearing loss
  - Head injury
  - Only a few without risk factors
- Longitudinal study underway to determine benefit from assistive devices
Case #2
Unilateral Auditory Neuropathy?
Or No 8th Nerve???

Right Ear

Left Ear
UNC Children with Absent 8\textsuperscript{th} Nerve

- 15 children with absent 8\textsuperscript{th} nerve identified at UNC by MRI since 2003
  - 5 bilateral, 10 unilateral

- 10 have ABR data
  - 10/70 (14\%) children at UNC who present with abnormal ABR and present cochlear microphonic have either unilateral or bilateral absence of 8\textsuperscript{th} nerve
Summary Points

● Absent 8th Nerve
  – Not uncommon 15 cases in 2 years
  – Can result in the appearance of auditory neuropathy
  – Over half have normal internal auditory canal morphology
  – Over one-third have normal labyrinth
  – Audiological, educational and medical recommendations will be influenced by this information

● Need MRI instead of CT in all children
  – With profound hearing loss
  – With audiological tests consistent with auditory neuropathy
Where Do We Go From Here? What Are the Issues?

- Heterogeneous group of children, manage on an individual basis
- Our team recommends early imaging with MRI
- Evaluate speech perception abilities as soon as possible
- Observe child carefully for any neurologic signs
- Consider supplemental communication methods
- Consider cochlear implantation if acoustic amplification not sufficient
Controversies/Unresolved Issues

- Acoustic Amplification?
- Match prescriptive targets?
- Monaural or binaural amplification?
- High frequency emphasis?
- Who will benefit from cochlear implantation?
- Which communication mode is best?
- How do we predict what will be needed?
- More research needed…