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SIG 9, Hearing and Hearing Disorders in Children

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Collaborative Management of Auditory Neuropathy Spectrum Disorder

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Outline

• Overview and Definitions
• Variations in Presentation
• Protocol for Diagnosis and Management
• Counseling Families
• Intervention Strategies
• Case Examples
Auditory Neuropathy: A Definition

Clinical syndrome characterized by electrophysiological evidence of normal or near normal cochlear function and absent or abnormal auditory pathway transduction.
Audiologic Findings

- Normal outer hair cell function as measured by present otoacoustic emissions (OAEs) or the presence of a cochlear microphonic (CM).
  - OAEs may be present initially but disappear over time, or be absent at time of diagnosis (Starr et al, 1996)
- Abnormal auditory nerve response as observed by absent or markedly abnormal ABR
- Acoustic reflexes are absent in most cases (Berlin et al 2005, 2010)
Clinical Characteristics Reported

- Pure tone thresholds ranging from normal to profound
- Disproportionately poor speech recognition abilities for the degree of hearing loss
- Difficulty hearing in noise
- Impaired temporal processing
- Hearing fluctuation
- Some individuals with AN have little or no communication difficulties while others are functionally deaf
- Not all individuals diagnosed with ANSD experience the same problems or to the same degree
Prevalence

- Disorder initially thought to be rare
- Many published reports since late 90’s describing patients with similar audiologic test findings (absent ABR with present CM and/or OAEs)
- Estimates range from 7-10% of children diagnosed with permanent hearing loss

(Rance 2005)
Possible Etiologies and Associations

- Genetic Etiologies:
  - Syndromic:
    - Charcot-Marie-Tooth disease; Friedrich’s Ataxia; Hereditary motor and sensory neuropathy (HSMN)
  - Non-syndromic:
    - Recessive genetic mutations: Otoferlin (OTOF), Pejvakin (PJVK)
    - Autosomal dominant mutations: AUNA1 (onset of auditory symptoms in late teens)

- Perinatal Conditions:
  - Hyperbilirubinemia
  - Hypoxia
  - Low birth weight
  - More common in premature infants

Rance (2005); Rapin & Gravel (2003); Starr et al. (2003); Hayes 2011
Possible Etiologies and Associations

- **Congenital Conditions:**
  - Cochlear Nerve Deficiency

- **Infectious Processes**
  - Viral Infections (e.g. mumps, meningitis)

- **Head injury**
  - e.g. Shaken baby syndrome

Rance (2005); Rapin & Gravel (2003); Starr et al. (2003); Hayes 2011
Guidelines Development Conference: Identification of Infants and Children with Auditory Neuropathy

Lake Como, Italy, June 19-21, 2008

Guidelines available at:
(Denver Children’s Hospital Website)
Guidelines: Identification and Management of Infants and Young Children with Auditory Neuropathy Spectrum Disorder

- Terminology
- Diagnostic Criteria
- Comprehensive Assessments
- Audiological Test Battery
- Amplification Strategies
- Considerations for Cochlear Implantation
- Habilitation for Communication Development
- Screening
- Monitoring Infants with “Transient” ANSD
- Counseling Families of Infants with ANSD

Guidelines Development Conference:
Identification of Infants and Children with Auditory Neuropathy
Lake Como, Italy, June 19-21, 2008
Terminology Considerations

• Same constellation of findings with different sites of lesion:
  » Auditory nerve
  » Synaptic dysfunction at junction of inner hair cell/auditory nerve
  » Myelin disorder
  » Cochlear nerve deficiency (small or absent 8th nerve)

• Panel sought to identify simplified terminology to reflect an auditory disorder with a range of presentations secondary to variety of etiologies

• **AUDITORY NEUROPATHY SPECTRUM DISORDER (ANSD)**
DIAGNOSIS OF AUDITORY NEUROPATHY
Recommended Audiologic Test Battery

• Auditory Brainstem Response (ABR)
• Acoustic Immittance Measures
  » Tympanometry
  » Acoustic Reflex Testing
• Otoacoustic Emissions Testing
• Behavioral Audiometry
  » VRA, BOA, play audiometry
• Speech Recognition Testing
Auditory Steady State Response (ASSR)

- ASSR responses can be obtained to high signal levels (>80dBHL) with ANSD but responses are elevated even in children who later show normal behavioral audiograms.
- Therefore, ASSR cannot be used to predict behavioral thresholds in ANSD

Recommended Audiologic Test Battery

- Auditory Brainstem Response (ABR)
- Acoustic Immittance Measures
  - Tympanometry
  - Acoustic Reflex Testing
- Otoacoustic Emissions Testing
- Behavioral Audiometry
  - VRA, BOA, play audiometry
- Speech Recognition Testing
Normal ABR
Estimating the Audiogram from Tone Burst ABR in Non-AN type hearing loss
Absent ABR with No Cochlear Microphonic: Child with profound hearing loss
Typical Pattern of ANSD
Abnormal ABR with Present CM

![Graph showing sound interrupted]

- Latency: 2.00 ms/div
- Latency Offset: -0.80 ms

![Graph showing emission and noise levels]

X O = Emission Level
- = Noise Level
What is a Cochlear Microphonic (CM)?

- Pre-neural response (occurs before Wave I in the ABR)
- Unlike the ABR, the CM shows a direct phase relationship to the acoustic wave form. When the polarity of the stimulus is changed there is a reversal of CM waveform
- Considered to have limited clinical use in past; renewed interest in diagnosis of ANSD
- CM can be recorded in normal ears, ears with “typical SNHL” and ears with ANSD
- Significance in ANSD is when CM is present when neural response is absent or markedly abnormal
ABR Protocol for Evaluating CM

- Must have adequate recording conditions
  - Infant ready to sleep (natural or sedated sleep)
  - Avoid electrodes positioned over transducer
- Single polarity clicks at 90dBnHL with rarefaction and condensation polarities
- Must use insert earphones
  - Excessive stimulus artifact with standard headphones obscures cochlear microphonic
- Sound interrupted run with stimulus on but sound tube disconnected or clamped to check for stimulus artifact
CM vs stimulus artifact

If response remains with sound tube disconnected from transducer, response obtained is stimulus artifact and not CM as in case below.
Example of child with normal hearing incorrectly diagnosed with ANSD
~Note poor quality of ABR on left compared to right
Example of child with ANSD incorrectly diagnosed with normal hearing

~Note incorrect identification of waveforms on left
Otologic Examination

• Medical History
• Ear Exam
• Etiology
• Evaluate for other associated problems
  » Seizures
  » Motor delays
  » Visual problems
  » Ear canal problems
  » Otitis media
• Radiologic Studies (MRI/CT)
  » Inner ear malformations
  » Cochlear nerve integrity
• Other studies as needed
Medical Diagnosis in 130 Children with ANSD at University of North Carolina
72% have positive history of other medical diagnoses
Cochlear Nerve Deficiency (CND) (May present with ANSD pattern)

- Small or absent VIII nerve
- Must perform MRI to determine if VIII nerve is small or absent
- CT may show normal internal auditory canal when cochlear nerve is absent
- In cases when there is question of CND both CT and MRI imaging may be needed
- Imaging is especially important when behavioral audiometry shows profound hearing loss
Child with bilateral deafness
No VIII\textsuperscript{th} nerve on right
UNC Children with Characteristics of ANSD and Available MRI (2009)
N=140

- 35/140 (25%) Cochlear Nerve Deficiency (CND) (absent or small cochlear nerve) in one or both ears
  - Unilateral (n=24; 69%)
  - Bilateral (n=11; 31%)

Recommended Audiologic Test Battery

- Auditory Brainstem Response (ABR)
- Acoustic Immittance Measures
  - Tympanometry
  - Acoustic Reflex Testing
- Otoacoustic Emissions Testing
- Behavioral Audiometry
  - VRA, BOA, play audiometry
- Speech Recognition Testing
ANSD Protocol for Infants: Behavioral Audiometry

- Behavioral assessment with VRA beginning at 6-7 months (developmental age) with goal of obtaining individual ear measures and bone conduction thresholds by 8-9 months of age
  - May be difficult (or impossible) with children who have additional developmental or medical challenges
  - Behavioral Observation may be needed
VRA Six Month Old
Hearing Aid Fitting in Infants with ANSD

- Behavioral thresholds cannot be predicted from ABR or ASSR
- Determination of hearing thresholds is delayed until infant developmentally able to perform task (6-9 months of age for most typically developing infants)
- Many children with ANSD are at risk for cognitive impairments resulting in a lengthier and more complicated process of threshold determination
- This results in delays in hearing aid fitting and greater amount of time without adequate audibility of speech signal
ANSD Guidelines (Como 2008): Recommended Amplification Strategies

- Amplification should be fitted as soon as ear specific elevated pure-tone and speech detection thresholds are demonstrated by conditioned test procedures.
- Hearing aid fitting strategies…should follow established guidelines for the fitting of amplification in infants and toddlers:
  - e.g. American Academy of Audiology Pediatric Amplification Protocol, 2003
- Since improvement in auditory function has been reported in some cases, careful monitoring needed to adjust and modify amplification as needed.
Verifying Audibility of Speech Spectrum

Speechmap/DSL 5.0a child

Jan 31, 2008 3:58pm

[Graph showing auditory data with markers indicating different conditions and levels.]
Evaluation of Speech Perception Following Hearing Aid Fitting or Cochlear Implantation

- Parent Questionnaires (e.g. PEACH, IT-MAIS or MAIS) (Zimmerman-Phillips, et al., 2000; Robbins, et al., 1991)
- Early Speech Perception Test battery (ESP) (Moog and Geers, 1990)
  - Standard
  - Low Verbal
- MLNT/LNT words and phonemes (Kirk, et al, 1995)
- PB-K words and phonemes (Haskins, 1949)
- CNC words
- HINT sentences in quiet and noise conditions
Early Speech Perception Test (ESP)
Factors that may affect outcomes

For all children benefit from a particular technology will depend on several factors including:

» Age at diagnosis and treatment
» Appropriateness of device fitting
» Consistency of use
» Quality of intervention
» Extent of family involvement
» Cognitive abilities of child
» Presence of other medical conditions
ANSD Guidelines (Como 2008):
Special Considerations for Cochlear Implantation

- Evidence of auditory nerve sufficiency should be obtained prior to surgery using appropriate imaging technology (Buchman et al., 2006)
- Children with ANSD who do not demonstrate good progress in speech recognition and language development should be considered candidates for cochlear implantation regardless of audiometric thresholds.
- Families should be informed that spontaneous improvement in has been reported. CI should not be considered until test results are stable and demonstrate unequivocal evidence of permanent ANSD…
CI Criteria-Children

- Advanced Bionics
  - Children-age 4 or less:
    - Failure to reach auditory milestones or <20% on MLNT at 70 dB SPL
  - Children > age 4: <12% on PBK words or < 30% on open set sentences at 70 dB SPL

- Cochlear Corporation
  - Children-12 months though 17 years
  - Bilateral profound SNHL in children 12 months to 2 years
  - Bilateral severe to profound SNHL in children 2 years and older
  - 30% or less on open set MLNT or LNT
  - 3-month trial with HA if not previously amplified

- Med El
  - Children- 12 months to 17:11 (17 years, 11 months)
  - Profound SNHL specified as 90 at 1K Hz
  - Lack of progress in auditory skills with habilitation and amplification provided for at least 3 months
  - Less than 20% on MLNT or LNT
  - 3-6 month HA trial without previous fitting; waived if ossification
UNC ANSD Children with CI N=52

CI in AN EAR

- 35% <6 months CI use/CNT N=11
- 29% Unable to perform open set (>2 yrs of use) N=13
- 15% Limited Open Set (<30%) N=7
- 21% Open Set Performers N=18
OPEN SET PERFORMANCE
N = 25
Variable Presentations of ANSD
Case Examples
Case #1:
Present CM and OAEs

- 24 wk preemie, 940 grams
- NICU 4 months, ventilated
- ABR at 4 and 5 months of age abnormal
- ABR repeated at 18 months-no change
Case #1
Normal thresholds, Present CM and OAEs

Audiogram at 14 months
Audiogram at 18 months
Case #1
Speech Perception Test Results

- Age 2 yrs-11 months:
  - ESP monosyllabic word test (closed set test of speech perception):
    - 12/12 correct for each ear at 50dBHL
- Age 5 years:
  - PBK words: 80% and 84% at 60dBHL for right and left ears
Case #2
Abnormal ABR with Present CM

Sound Interrupted

Sound Interrupted
Case #2
Present OAEs
Case #2
Child with Profound Bilateral HL
Present CM and OAEs

Ear exam: Normal
EKG: Normal
MRI: Normal
Connexin test: Negative
Otoferlin test: **POSITIVE**
Received CI at 24 months of age
Case # 3 Background

- 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
- Initially diagnosed with profound bilateral SNHL at an outside clinic and fitted with high gain hearing aids
Case #3
Child with “moderate loss”
CM present, absent OAEs
Case #4: Large CM; Present OAEs; Distal Waveforms Present

Caution needed when interpreting ABRs that show abnormal waveform morphology at high intensity levels.

Sound Interrupted
Case #4 (continued)
VRA with insert earphones
Age 14 months
Case #5

- Child born at full term
- No family history of hearing loss
- Presented to clinic with left profound unilateral hearing loss at 4 years of age.
- Passed newborn hearing screen using OAEs
Case #5 OAEs
Case #5
ABR Completed at Age 4 years
Case #5

- **Results of MRI:**
  - Right ear: Normal inner ear anatomy
  - Left ear: Consistent with small or absent nerve VIII

- At age 7 years child has above average speech and language development, no academic problems

- Managed as we do other cases with profound unilateral hearing loss.
Case #6: 8 year old with ANSD

- Child with progressive neurologic disease
- Speech recognition scores 5 years post CI in right ear:
  » 6% words: 38% phonemes
- Recently began wearing HA again in non-CI ear
- Mom reports increased benefit compared to CI alone
- Many additional medical issues:
  » Ataxic (in wheelchair now)
  » Optic neuropathy (only sees at close range)
- Probably “true” neuropathy
Six Cases with ANSD pattern on ABR…Six Different Outcomes

1. Normal hearing sensitivity no device needed, limited services required
2. Child with profound bilateral hearing loss; doing well with CI
3. Child with moderate HL benefitting from amplification
4. Child with ANSD pattern on ABR but distal waveforms present; normal hearing sensitivity
5. Child with unilateral profound HL and absent cochlear nerve
6. Child with progressive neurologic disease; limited benefit from either HA or CI alone; child feels best benefit from CI and HA combined
Evidence re Amplification

- Evidence regarding outcomes from amplification is limited
- Few peer-reviewed studies re outcomes with amplification or CI have been published
- Existing literature is based on small number of children
- Many of published reports are anecdotal
- Only a few published studies document use of a prescription-based fitting strategy that ensures audibility of speech signals
Audiological Management of Auditory Neuropathy Spectrum Disorder: A Systematic Review of the Literature

Roush, P., Frymark, T., Venedictov, R., and Wang, B.

American Journal of Audiology 2011, Sept. 22 (epub ahead of print)
Cortical Evoked Potentials (CAEPs)

- ABR evaluates outer ear to lower brainstem
- CAEP evaluates outer ear to auditory cortex
- CAEPs not as reliant on timing as earlier evoked potentials and may be present when ABR is not
- Unlike ABR must be completed in awake (but quiet) infants
  - Cone Wesson and Wunderlich, 2003
- Further CAEP research needed with normal infants and infants with SNHL and ANSD
Cortical Evoked Potentials (CAEP)
Acoustic Change Complex-Gap Detection
Preliminary Findings
Shuman He, PhD
University of North Carolina at Chapel Hill
Acoustic Change Thresholds (ACC)-Gap Detection
Acoustic Change Complex (ACC) Gap Detection Thresholds and PBK Scores
Counseling Families
Counseling in ANSD: What Do We Say to Families?

- Child has an auditory disorder; difficult to know prognosis at time of ABR evaluation
- Degree of deficit may be mild or severe
  - a small number have normal hearing sensitivity
- Results of behavioral testing are necessary before specific recommendations can be made
- Hearing aid use is helpful in some cases but not in others; benefit can only be determined with appropriate fitting and consistent use
- Cochlear implantation may be a better option if adequate benefit from amplification not received
UNC Team Approach for Children with Hearing Loss

- Parents & family
- Medical Doctor
- Auditory Verbal Therapist
- Other Professionals
- Education
- Audiologist
Lions, and Tigers and Bears, Oh My!

• I just got a child on my caseload with ANSD. I have no idea what to do.
• We can’t expect them to make typical progress, they have ANSD.
• But, they are diagnosed with ANSD.
• Additional disorder? I’m sure it is the ANSD.
ANSD and Therapy: IT’S NOT MAGIC
Evaluate and Monitor Progress

• **Expectations for Progress: 3 months of listening**
  *given child is attending appropriate Listening and Spoken Language therapy and audiology visits*

  » Child detects sound

  » Child is demonstrating some bonding to hearing aids/CI, measured by parent report, observation/clinical judgment and IT-Mais (shows increasing ability to wear device daily)

  » Parents report some sound awareness such as turning to environmental sounds
Evaluate and Monitor Progress

• Expectations for Progress: 6 months of listening *given child is attending appropriate Listening and Spoken Language therapy and audiology visits and age appropriate goal

  » Consistent and full time use and child indicates in some way if not hearing
  » Sound awareness repertoire is growing identifying more sounds, pointing to ear
  » Identification of 15 LLS in a set of 2
  » Identification of at least 2-3 songs
  » Identification of 10 stereotypic phrases
  » Increased vocalizations to communicate desires
  » Increased changes in vocalizations (duration, intensity and pitch) as well as different vowels
Evaluate and Monitor Progress

• Expectations for Progress: **1 year of listening**
  *given child is attending appropriate Listening and Spoken Language therapy and audiology visits and age appropriate goal

  » Completed Year 1 Auditory Learning Guide Goals
  » Depending on age and hearing history, some spoken language development including 7 to 10 words
  » Have most vowels and some early developing consonants
Factors Affecting Outcomes

- Age at diagnosis and treatment
- Appropriateness of device fitting
- Consistency of use
- Quality of intervention
- Extent of family involvement
- Cognitive abilities of child
- Presence of other medical conditions
When Targets Aren’t Met

• Ask these questions:

» Is the family consistently attending therapy with a professional trained in the development of spoken language through listening?

» Does the family carry over goals at home on a daily basis?

» Is the child wearing the hearing technology consistently?

» Have alternate hearing technology been considered (i.e. hearing aids to a cochlear implant)

» Is the child displaying signs of other developmental delays or disabilities?
When Targets Aren’t Met

• Consider additional evaluations
  » UNC Hearing and Development Team
    • Speech-Language Therapist, LSLS Cert. AVT
    • Psychologist
    • Special Educator
    • Occupational Therapist
    • Physical Therapist

• Consider alternative methods of communication
  • Cued Speech
  • Signed Exact English
72% have positive history of other medical diagnoses

Medical Diagnoses
N=130

Diagnosis

- Normal
- Premature
- NICU
- Vent
- CP
- Other neuropathy
- Ototoxic meds
- IVH
- F H1
- Perinatal F/Infxn
- High Bili
- Kernicterus
- Syndrome
- CMV
- Meningitis
- Unknown
- Other

%
Why Cued Speech?

• Development of spoken language through typical patterns
• Clarification of grammatical concepts (plurals, possessives)
• Increased reading skills
• Visual representation of perceived auditory signal
• Parents and therapists can learn system in 10-20 hours and use efficiently with continued practice
Counseling Families Regarding Spoken Language Prognosis

• The etiology and other medical conditions impact auditory performance thus impacting learning spoken language through listening
• It is crucial to enroll in therapy early with a qualified therapist
• Results of behavioral responses to HA or CI are necessary before communication decisions are clear
• We must monitor continuously, adapt with time and experience
• Working with a team of professionals in communication with each other also crucial to determine most effective communication methods
Current research in CI population with ANSD

Consider only ANSD CI children with no multiple medical diagnoses and match them to children who have SNHL. How do they compare after some duration of CI use?
SRI-Q scores for matched ANSD/SNHL CI Children

Matched for age at implant, first language, communication mode, uni- or bilateral implant, years of CI experience (3-9), lack of other medical diagnoses, social/educational status

SRI-Q - cumulative quotient of test hierarchy. Child must meet a level of performance (70%) before moving on to next level of test. Each level is a 100 point scale. (CDaCI, Wang et al 2008)
ANSD:

Assessing on a Case by Case Basis
Case #1 “J”
Meet “J”
Background

- 27 Week Preemie
- Diagnosis of Spastic CP combined with dystonia
- Visual Deficit
- Identified at 3 months with ANSD
- Later testing with VRA showed thresholds in moderate-severe range
- Parents initially reluctant to use hearing aids but proceeded after stable audiogram demonstrated
- Parent report indicating some speech and language progress with early hearing aid use
- Parents used Cued Speech
At 2 years of age parents began to pursue cochlear implantation

- Had enough residual hearing to afford detection of speech and environmental sounds with hearing aids
- Responded to name in quiet 75%
- IT MAIS score of 28/40
- ESP mono-syllable 80%
- Parents report limited growth in spoken communication, speech sounds remained limited

Received first CI at age 2.5 given a guarded prognosis

Supportive Family: Continued use of Cued Speech post CI

LSLS Therapist
Speech and Language Scores

• 1 year post CI:
  » PLS-4 Scores: AC 71   EC 75   TL 70
  » PB-K Words 20% Phonemes 64%

• 2 years post CI:
  » PLS-4 Scores: AC 85   EC 81   TL 81
  » PB-K Words 32% Phonemes 74%

• 3 years post CI:
  » CELF-4 Scores: AC 67   EC 75   TL 69
  » PB-K Words 76% Phonemes 93%
“J” Today

- Speech perception- 100% PBK words and phonemes
- Speech production- 95% intelligible (according to his mom an SLP)
- OWLS II- Oral Comprehension- 75, Oral Expression- 77 Overall- 74
- Fully mainstreamed in the 5th grade with FM and cued speech transliterator making A’s and B’s
- Receives ST 1xweek/30 min, HI 2Xwk/ 30 min; PT and OT on quarterly consultation
- Very organized, planner, gets his work done
Video
Case #2
Case #2
Background

• Born at 30 weeks gestation (2 lbs. 4 oz)
  » Twin to twin transfusion syndrome (twin deceased)
  » Hospitalized in NICU 3 months
  » Did not require ventilator
• No family history of hearing loss
• Failed AABR NB hearing screen, passed OAEs
• Diagnostic ABR recommended but not completed
Background

• Followed up for second opinion at UNC at age 21 months
  » Diagnosed with bilateral ANSD
  » Thresholds in mild to moderate range
• Received hearing aids at 22 months of age
• Began weekly AV sessions at 2 years of age until present
• Parents used a listening and spoken language approach.
Case #2

[Graphs and data plots demonstrating various measurements and sound responses]
Audiogram at 21 months
Audiogram at 5 ½ years
Speech and Language:

- **Hearing Age: 1 year (CA- 3 years)**
  - PLS-4: AC 81  EC 92  TL 92
- **Hearing Age: 2 years (CA- 4 years)**
  - PLS-4: AC 90  EC 89  TL 89
  - CELF-P: CL 79  RL 86  EL 81 LC: 81 LS 86
- **Hearing Age: 3 years: (CA- 5 years)**
  - OWLS: LC 89 81 89 OC 83
Case #2 Video
Case #3
Case # 3 Background

• Born at 25 weeks gestation
  » Hospitalized 4 ½ months
  » Ventilated for 6 weeks
  » Oxygen 3 ½ months
  » Hyperbilirubinemia
    • Treated with lights, exchange transfusion
  » Treated with antibiotics and diuretics

• No family history of hearing loss
• Did not pass NB hearing screen at hospital discharge
• Diagnosed with profound bilateral SNHL and fitted with high gain hearing aids
ABR Obtained at UNC, Age 6 Months
(2 1/2 months adjusted age)
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
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
VRA with Insert Earphones Age 24 Months (20 ½ months adjusted age):

Child conditioned for play audiometry procedure but limited attention span

Results similar to previous audiograms

Tympanometry

Right: normal
Left: normal
Communication Status
Age 24 Months (20 1/2 months adjusted age):

- **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 50dBJHL:
    - Able to accurately identify from closed set of objects for spondee and monosyllabic words
Age 5

- Mainstreamed in kindergarten
  - Using personal FM in classroom
- Receiving services from auditory verbal therapist and speech and language pathologist
- Functioning in average range in receptive and expressive language development
- Working on articulation errors
Age 8 years

- Now mainstreamed in 2nd grade
  - Using personal FM in classroom
- Receiving services from auditory verbal therapist and speech and language pathologist
- Aided monosyllabic word score 88% at 57dBHL.
- Functioning in average range in receptive and expressive language development
- Working on articulation errors
Speech and Language
Age 9 years

• Testing October 2012 (CA: 9 years)
  » OWLS: LC 99  OE 114  OC 107
  » GFII: 106
Case #4
Case # 4 Background

• Born at full term
• Developmental milestones normal until two years of age
• At age two, child developed peripheral neuropathies including optic neuropathy
• Hospitalized at age 3 and underwent several diagnostic studies including:
  » Electromyography
  » Muscle Biopsies
• Numerous medical consultations including:
  » Otolaryngology, Genetics, Neurology, Ophthalmology and Infectious Disease
• Etiology for her medical problems never determined
  » Guillain-Barre, Charcot-Marie-Tooth and mitochondrial disease were all ruled out
Audiogram at Age 6 years

- Bilateral rising pattern
- Unaided speech recognition using monosyllabic words:
  - 100% right
  - 84% left
Audiogram Age 10 years

- Bilateral rising pattern: mild on right, moderate left
- Unaided speech recognition using monosyllabic words:
  - 40% right
  - 24% left
Audiogram at Age 11 years

- Child continued to have fluctuating speech recognition scores
- ABR showed AN pattern
- Hearing aids tried but child and family reported minimal benefit
- Despite significant residual hearing, child was no longer able to repeat any words on monosyllabic word test
- Successful communication could only be accomplished at close range with speech reading
Otoacoustic Emissions Age 11 years
Follow up

• Family counseled extensively regarding potential benefits and limitations of cochlear implantation, particularly in view of multiple peripheral neuropathies.

• After careful consideration, family decided to proceed with left CI.

• After one year of use with CI:
  » Monosyllabic words: 32% words, 66% phonemes
  » Parents reported that while she continued to have significant communication difficulty, they felt need for repetitions was reduced with device on.
Follow up

• After 4 years of device use:
  » Monosyllabic words: 20%
  » Continued deterioration in motor abilities
  » Parents report significant difficulty understanding anyone other than family members
  » Since child still had significant residual hearing in her right ear; decision made to attempt hearing aid use age.

• At age 17 and after six years of CI use and with a hearing aid in contralateral ear:
  » Monosyllabic word score only 20%
Case Study Conclusions

- ANSD treatment plan requires significant collaboration between parent(s), audiologist and therapist.
- Each child should be evaluated and monitored on an individual difference as no ‘one size fits all’ approach works.
- Many children with ANSD also have other medical and developmental delays. These delays need to be considered when monitoring progress.
- There is no magical therapy process. It is critical for families to work with a therapist with experience in listening and spoken language if their desired communication mode is spoken language.
Summary of UNC Protocol for Management of Infants with ANSD

• Diagnose ANSD using ABR with single polarity clicks
• Counsel family about recommended steps in first year of life
• Enroll in early intervention
• Complete otologic exam including imaging with MRI (and CT if needed)
• Attempt behavioral audiometry with VRA beginning at 6-7 months developmental age
• Fit child with hearing aids as soon as behavioral thresholds have been established
Summary of UNC Protocol for Management of Infants with ANSD (continued)

- Set hearing aids to match targets for gain and output using prescriptive formula
- Perform hierarchical battery of speech perception tests
- Regularly communicate with early intervention teacher and parent regarding communication progress
- Consider CI if benefit from amplification insufficient for continued progress in communication skill development
- Use cortical evoked potentials to aid with management when needed
- Refer for comprehensive developmental evaluation when child has complex needs
Conclusions

- ANSD is more complicated than originally thought and population more heterogeneous.
- It’s unlikely that a single approach to management will meet the needs of all children.
- Some children will benefit from hearing aids either in the short term or the long term, others will require cochlear implantation.
- Visual methods to support communication may be required for some children even those who have received cochlear implants.
Conclusions

- The available clinical evidence does not support withholding audibility from infants with ANSD. Although audibility does not ensure good speech recognition, lack of audibility is certain to result in poor speech recognition.

- Important to consider the needs of the whole child, not only the auditory neuropathy diagnosis.

- Important to use team approach to carefully monitor child’s progress in meeting communication goals.
Research Needs

- Evidence regarding clinical management and use of amplification is still limited. More research needed especially with infants and young children.

- Studies aimed at evaluating hearing aid outcomes should include evidence-based prescriptive hearing aid fitting methods and real-ear verification methods appropriate for use with infants and children.

- Better ways to predict who will benefit from amplification vs cochlear implantation.

- Continued research needed into the role of CAEP and other electrophysiologic tests in evaluation and management.
Thank you for your attention!
Selected References and Resources


References and Resources


References and Resources


Thank You!

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