AUDITORY NEUROPATHY

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WHAT DO WE KNOW?

- Neuropathy vs. Dys-synchrony
  - If AN – site of lesion would indicate that CI is not beneficial
  - If AD – then CI should be beneficial
  - Currently there is no definitive test to differentiate these two categories
  - Current assessment procedures characterize auditory skill development –
- Course of the condition is unpredictable
  - are they progressing with HAs or without,
  - is there fluctuation – good days and bad days, good times-badtimes
2002-2009 STATE OF COLORADO

- Incidence of HL in 610,829 infants screened from a birth population of 626,701
- The screening rate was 97.5% and the follow-through rate ranged from 83 to 89%
- 873 infants were diagnosed with SNHL in this time period
- 67 infants were diagnosed with AN
- Of children with SNHL the incidence of AN was .87%
- Very close to 9% of children with SNHL

- Prevalence of AN was .01%
  - 1 in 10,000
- Prevalence of SNHL was .16%
CHILDREN WITH AN
 ▶ 21% were unilateral AN (14/67)
 ▶ 79% or 53 were bilateral AN(53/67)
 ▶ 60 were born in the NICU (90%)
 ▶ Most infants from NICU have bilateral AN – 82%
   ▶ 11 were unilateral (18%)
   ▶ 49 were bilateral (82%)
 ▶ 7 were born in the well-baby nursery (10%)
   ▶ 3 were unilateral (43%)
   ▶ 4 were bilateral (57%)

DEVELOPMENTAL DATA ABOUT ANSD
 ▶ 39 of the children in this birth cohort have developmental data in the birth through three age range
 ▶ 2 of the 11 (18%) are children with unilateral UANSd
 ▶ 37 of 53 children have bilateral ANSD (70%)
PROPORTION OF CHILDREN WITH SIGNIFICANT COGNITIVE DISABILITY (32%)

- 32% (12/37) have significant cognitive disability in addition to hearing loss
  - Almost 1 in every 3 children
  - 9 of these 12 children or 75% have significant cognitive and neurological involvement with developmental quotients ranging from 10 to 55 (9/37 or 1 in 4)
  - 3/4 of the children with cognitive disability have severe/profound involvement and multiple other issues

ADDITIONAL DISABILITIES BESIDES COGNITIVE - 57%

- 9 additional children had normal cognitive development but other disabilities including vision, motor/orthopedic, severe health issues
- 57% (21/37) of the population of children with bilateral AN have additional disabilities and the vast proportion of these issues are severe/profound cognitive delays, in many cases also neurological issues.
NORMAL COGNITIVE FUNCTION – 68%

- 68% (25/37) of the children (B-AN) had cognitive developmental quotients within the normal range.
  - 2 of every 3 children had cognitive developmental quotients within the normal range
  - 9/37 had normal cognitive quotients and additional disabilities (24%) (9/25 36% of children with normal cognition)

HISPANIC/LATINO

- 32% (12/37) Colorado children with AN identify as Hispanic/Latino
  - One of every Three children
  - 10 of these 12 children are Spanish-speaking in the home indicating that one of every four children with bilateral AN in the state of Colorado is born into a family that does not speak English as a native language.
GENDER

- 35% are female (13/37)
- 65% are male (24/37)
- The incidence of bilateral AN is two times greater for males than for females.

COCHLEAR IMPLANTS

- 10.8% (4/37) of the children received a cochlear implant/s
- 14% (5/37) were not amplified
- 76% (28/37) used hearing aids
EARLY INTERVENTION SERVICES

- All 37 families received weekly early intervention services from an early intervention provider with either a deaf education, speech/language pathology or an audiology degree.

SIGN LANGUAGE

- 82% (31/37) families chose to receive weekly sign language instruction from a deaf or hard of hearing native/fluent sign language instructor
- 1 of the children used Cued Speech
LANGUAGE QUOTIENTS

- 68% of the children had normal cognitive quotients.
- 57% (21/37) of the children had language quotients within the normal range.
  - Approximately 11% of the children with normal cognitive scores did not have language quotients in the normal range.
  - Recall that 57% of the children had hearing loss plus additional disabilities.
- 32.4% had cognitive disabilities.
- 24.3% had normal cognitive abilities with additional disabilities.

SYDNEY COCHLEAR IMPLANT PROGRAM (GARDNER-BERRY, 3/15/2012)

- 43% of children with AN had additional disabilities (21% SNHL).
- 16% of children with AN had abnormalities on CT scan (x-ray computed tomography) (N=142) (6% of SNHL).
  - Mondini deformity
  - Wide IAMs (internal auditory meatus)
  - Dysplastic apical turn
  - Abnormal vestibule & lateral SSC (semi-circular canals)
AN & COMPROMISED AUDITORY NERVE - MRI

- N=142 (<10 years of age)
- 26% had compromised Auditory Nerve
  - 20% bilateral
  - 6% unilateral

Developing An Action Plan

TREATMENT OPTIONS
THE IMPORTANCE OF EFFECTIVE COUNSELING

- Need to help parent overcome feelings of helplessness and confusion due to uncertainties:
  - With unknowns, outcomes, treatment plans and variabilities
  - Finding comfort in making choices that may change
- Work with parents to develop an action plan
- Gather data/audiologic & developmental
- Need to establish an effective team of professionals

AMPLIFICATION CONSIDERATIONS

- No amplification
- Hearing Aids
- Cochlear Implantation
  - Consider cognitive competency of the child to use the auditory perceptual information and auditory access provided by the cochlear implant.
  - Consider the visual communication development of the child
  - Consider the auditory skill development of the child
COMMUNICATION APPROACHES/OPPORTUNITIES
VISUAL

- Sign language – visual conceptual communication
  - Requires visual ability (e.g. cortical blindness or sensory blindness)
  - Requires motor ability (e.g. severe cerebral palsy)
  - Requires visual linguistic ability (e.g. autism and difficulty with visual attention, visual interpretation, visual symbolic gesture or play) – signs are conceptual

COMMUNICATION OPPORTUNITIES/APPROACHES: VISUAL

- Cued Speech – visual code for speech reading
  - Requires visual ability (e.g. blindness, other visual impairment)
  - Requires motor ability (e.g. cerebral palsy or other significant gross motor disability)
  - Requires visual linguistic ability (e.g. visual attention, visual integration of cue and speech reading)
  - Cues require integration of speech reading cues and speech/motor
COMMUNICATION APPROACHES/OPPORTUNITIES
AUDITORY/ORAL

► Auditory/Oral- spoken language communication
  ▶ Requires auditory access (consistent and stable access-fluctuation of thresholds, good days/bad days)
  ▶ Requires auditory attention (ability to attend auditorally)
  ▶ Requires auditory integration (integration of sounds to verbal words- meaning)
  ▶ Can be successful with hearing aids, without HAs, with resolved AN or with cochlear implants

DEVELOPING A PLAN

► Amplification
► Functional Auditory Skill Set
► Communication
► Language
► Speech
► Cognition
KEY TO A SUCCESSFUL PLAN IS ONGOING ASSESSMENT AND FLEXIBILITY

- Consistent Team Communication
- Parent and Child Centered Clear Objectives
- Connections to Resources

ASSESSMENT AND ADAPTATIONS

- Obtain baseline data
- Retest to measure rate of progress
- Goal: Attempt to maintain development commensurate with cognitive age – additional disabilities complicate developmental progress
FUNCTIONAL AUDITORY SKILL DEVELOPMENT

- Closely observe and monitor listening skills in a variety of conditions. Changes may be noted:
  - quiet vs. noise
  - music and singing
  - time of day
  - weekly and even monthly
  - aided and unaided
- Look for consistency and quality of responses
- Auditory behaviors may not be hierarchical pre-implantation for AD children

AUDITORY SKILL DEVELOPMENT

- Monitor with trial amplification
  - Awareness vs. speech discrimination
  - Cortical auditory evoked potentials show promising use for fitting of amplification with AN
- With a cochlear implant, expect hierarchical auditory skill development
  - Allow time for spontaneous recovery
  - Monitor development of speech & language
  - Identify auditory discrimination skills vs. pure tone hearing levels
TOOLS TO MEASURE FUNCTIONAL AUDITORY SKILL DEVELOPMENT -

- DASL, Developmental Approach to Successful Listening
- ASC – Cincinnati Auditory Skills Checklist
- IMP - Infant Monitor of Vocal Production
- CASLLS Cottage Acquisition Scales for Listening, Language and Speech- Sounds and Speech
- Little Ears Auditory Questionnaire
- Checklist of Auditory Communication Skills

CREATING A FUNCTIONAL DEVELOPMENTAL PROFILE

- Assess at regular intervals – every 3-6 months
- Expect developmental gains at a rate that is commensurate with that child’s cognitive skills – with the exception of multiply disabled children who may have extreme difficulty learning language at a normal rate
- Review data on ANAD/ANSD - % cognitive delay, % other disabilities
TYPES OF ASSESSMENT

- Parent/caregiver report – parent questionnaires
- Direct observation of the child
- Observation of child’s interaction with a parent
- Videotaped interaction
- Clinician-administered assessments
- Multi-disciplinary – all developmental domains

CASE 1: EDDIE

- Age of ID: 18 mo.
- Right UE congenital amputation
- Maternal Level Ed: some college
- NR ABR – Unknown degree – initial audio
- CI: 20 months of age
- Second CI: 23 months of age
MACARTHUR: EXPRESSIVE VOCABULARY AGE (EVA) 22 MO. AT CA 26
EVA 29 MO. AT CA 33

SALT (MORPHOLOGY SYNTAX)

- 26 mo. (4 mo post CI)
- 115 utterances
  - 53% Sign Language
  - 71% Spoken
- MLU 1.08
- 144 words 66 diff
  - 81 Spoken (37/81 diff)
  - 63 Signed (29/63 diff)

- 33 mo (13 mo post CI)
- 248 utterances
  - 27% Signed (N=66)
  - 96% Spoken (238/248)
- MLU 1.11
- 204 words 89 diff
  - 69 Sign (40/69 diff)
  - 55/69 intell Oral
### 26 MO.
- Unintelligible speech
- 14 vowels 51% correct
- 9 consonants 35% correct

### 33 MO.
- EOWPVT 29 mo. 34%ile
- Speech Intelligibility: 6 Unintelligible
- 12 vowels 16% correct
- 13 consonants 32% correct

### EDDIE – JULY 2012 – AT AGE LEVEL STANDARDIZED TESTS
- CELF P2, Clinical Evaluation of Language Fundamentals (45%ile) – in spoken English
  - Sentence Structure Scaled Score: 10
  - Word Structure Scaled Score: 9
  - Expressive Vocabulary Scaled Score: 10
  - Core Language Standard Score: 98
Language scores improved with age
Speech intelligibility improves with age
Mainstream success – at age level without additional services
Auditory dyssynchrony – CI worked well for auditory access
Intelligible speaker

CASE II: CARLOS
Well – baby nursery
Parents – both had a college degree
Severe hearing loss bilaterally – pre-implant
Spanish-speaking home
First cochlear implant – about 20 months
Second cochlear implant – 3 years, 11 months
CASE 6 = CARLOS

- ANSD
- NRT absent with first CI
- Slow progress in spoken and visual language systems
- P1s present prior to implanting second side
- One year later due to poor outcomes, P1s were absent when repeated
- Second CI activated 12/2010
  - NRT present
  - Making significant gains in visual communication
  - Responding to high frequency sounds

CARLOS – LENA DATA – QUALITY OF HIS SPOKEN LANGUAGE DAILY DIET

- Adult word count – 23,990 95th%ile
- Conversational Turns – 674 72nd%ile
- Child Vocalizations -  2312 57th%ile
- ***AVA Standard Score – 73.9 4th%ile

- High quantity of vocalizations but they were not speech-like vocalizations – no meaningful spoken language
- He used his vocalizations for conversational turn-taking.
IS SOUND REACHING THE CORTEX – CORTICAL AUDITORY EVOKED POTENTIALS – P1s

- Carlos had P1 testing – cortical index of audibility
- Time 1 – P1 was within normal limits with CI
  - However, behaviorally, Carlos was responding inconsistently to sound
- Time 2 – P1 was absent – with CI
- Time 3 – P1 was present, then, absent, then, present – fluctuated within session

VOWEL & CONSONANT DEVELOPMENT INSUFFICIENT FOR INTELLIGIBLE SPOKEN ENGLISH - BY 26 MONTHS SHOULD HAVE 18-20 CONSONANTS
MINNESOTA EXPRESSIVE LANGUAGE (EL) SIGN LANGUAGE- INCONSISTENT – LANGUAGE QUOTIENT (ELQ) DROPPING BY AGE

- EV = Expressive Vocabulary Age (MacArthur Communicative Development Inventories)
- EL = Expressive Language Age – Minnesota Child Development Inventory
- LC = Language Comprehension Age – Minnesota Child Development Inventory
- ELQ = Expressive Language Quotient (Expressive Language Age/chronological age) x 100
- LCQ = Language Comprehension Quotient (Language Comprehension Age/Chronological Age) x 100
- EVQ = Expressive Vocabulary Quotient (Expressive Vocabulary Age/Chronological Age) x 100
DO ANAD (ANSD) CHILDREN RECOVER?

- Infant Monitor of vocal Production (IMP)
  - 3 measures in the first year of life
  - 4-5 month, 7-8 month, 10-11 month
- Issue: Children diagnosed with AN who recover because of prematurity issues
- Candidacy for early CI – pre 12 months
- Mandatory that you investigate auditory-vocal development

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THE INFANT MONITOR OF VOCAL PRODUCTION (IMP)
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RIDBC Renwick Centre / University of Newcastle
Sydney, Australia.

IMP PRÉCIS

- Criterion referenced instrument
- Clinical evaluation & parent education tool
- Successive measure of infant, pre-linguistic vocal competence
- Diagnostic aid to habilitation programming
TYPICAL INFANT VOCAL DEVELOPMENT

(Kuhl, 2004; Lewkowicz & Hansen-Tift, 2012)

STAGES OF INFANT VOCAL PRODUCTION

Adapted from: Oller (2000); Nathani, Ertmer, & Stark (2006)
IMP PROBE-QUESTIONS

37. 
When playing alone Are sounds described as general, stereotyping, mixing?

40. parent: Do you hear "I'll" to himself when he is playing aloud? What does he do?

INTERPRETING THE IMP

SCORE: Assessment Date ..........................................

Child’s Age:........... yrs ........... mths.
**INTERPRETING SEQUENTIAL IMP RESULTS**

**IMP: INFANT (ANSD) CASE STUDY 102**

**SCORE:**
Assessment Date: "Lily"
Child's Age: 11.5 months

[Diagram showing assessment results with measures of Chronological Age (Corrected), Question calling, Hanging age (CT), Innate, Transition to A-P Loop, Integrity of A-P Loop.]
IMP: INFANT (ANSD) CASE STUDY

SCOR:
Assessment Date .......... "Lily" ..............
Child's Age........... yrs. 8.5 mths.

IMP: INFANT (ANSD) CASE STUDY

SCOR:
Assessment Date .......... "Mary" ..............
Child's Age........... yrs. 9 mths.
**IMP : INFANT (ANSD)**

**CASE STUDY**

**SCORE:**
Assessment Date: "Mary"
Child’s Age: 9 weeks

**IMP : INFANT (ANSD)**

**CASE STUDY**

**SCORE:**
Assessment Date: "Ian"
Child’s Age: 1 yr 7 mths.

**Graphs:**
- Innate
- Transition to A-P Loop
- Integrity of A-P Loop
**IMP** : INFANT (ANSD) CASE STUDY 35

**IMP** : INFANT (ANSD) CASE STUDY 135

**SCORE:**

Assessment Date ..........“Cameron” ............

Child’s Age: 1 yrs. 1 mths.

<table>
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<th>Chronological Age (Corrected)</th>
<th>Physiological</th>
<th>12</th>
<th>15</th>
<th>18</th>
<th>24 months</th>
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Innate Transition to A-P Loop Integrity of A-P Loop
**IMP**: INFANT (ANSD)  
CASE STUDY

135

"Cameron"

DIFFERENCE IN PROGRESS (ANSD)  
(Kuhl, 2004).
Registration

This training is free of charge, however you are required to register before you can access the training modules. We also ask that you share data collected using the IMP via the online form, available in the training website.

Go to http://www.ridbcrenwickcentre.com/imp to register. Once registered, you will receive an email containing your login credentials.

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WHAT DOES AN AUDITORY SPOKEN LANGUAGE MATRIX CONSIST OF? ?? DO WE WANT TO USE THIS WITH CARLOS?

- Questionnaires
  - Minnesota
  - MacArthur CDI
  - Auditory Skill Checklist
  - Little Ears
- Visual Reinforcement Infant Speech Discrimination (VRISD)
- Language Environment Analysis (LENA)
- Cortical Evoked Auditory Potentials (P1s)
- Vestibular Evoked Myogenic Potential (VEMP)