GRAND ROUNDS IN COCHLEAR IMPLANTS

October 16, 2012

Today's Agenda

- Classroom format and details
  - Asking a question or posting a comment
  - Downloading the handouts
- Introduction of our guest clinician
  - Holly FB Teagle, AuD
- Discussion and Questions

**The information presented during today's webinar may not necessarily reflect the views or opinions of Advanced Bionics.*

Announcements

- Upcoming Events and Webinars
- SoundWaves Professional Newsletter
- Next Cochlear Implant Grand Rounds will be in February, 2013
- Certificate of Attendance: Carissa.moeggenberg@advancedbionics.com

Your Questions

Please type your questions or discussion points into the Q&A box and click submit.

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Meet Holly FB Teagle, AuD

"Cochlear Implants for Children with Auditory Neuropathy Spectrum Disorder: What Are We Learing?"

Department of Otolaryngology
University of North Carolina School of Medicine

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Cochlear Implants for Children with Auditory Neuropathy Spectrum Disorder: What are we learning?

October 16, 2012

Holly FB Teagle, AuD
Department of Otolaryngology
University of North Carolina School of Medicine

University Of North Carolina
Chapel Hill, North Carolina

Large tertiary care center with referrals within NC and surrounding states

Approximately 1900 infants and children followed
- 1100 using amplification
- 825 with cochlear implants
- 200+ with ANSD diagnosis

Universal newborn hearing screening legislation 1999

University of North Carolina
Department of Otolaryngology
UNC Hospitals
Pediatric Audiology
Carolina Children's Communicative Disorders Program
Center for the Acquisition of Spoken Language through Listening Enrichment

Audiologists:
Nissele Franco, AuD
Corinne Macpherson, AuD
Sarah Martinho, AuD
Laura Fleenor McCall, AuD
Jill Ritch, AuD
Patricia Roush, AuD
Debora Hatch, AuD
Lisa Park, AuD
Jennifer Woodard, AuD
Holly Teagle, AuD

Speech-Language Pathologists:
Hannah Eskridge, AVT
Lillian Henderson, AVT
Sandra Hancock, AVT
Erin Thompson, AVT
Maegan Evans, PhD

Otolaryngologists:
Craig Buchman, MD
Oliver Adunka, MD
Carlton Zdanski, MD
Harold Pillsbury, III, MD

Auditory Physiologists:
John Gross, Ph.D
Shuman He, Ph.D

Outline

- Overview and Definitions
- Variations in Presentation
- Electrophysiological Findings
- Clinical Outcomes
- Implications for Clinical Management

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
- Abnormal auditory nerve response as observed by absent or markedly abnormal ABR
- Acoustic reflexes are absent in most cases

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 ANSD 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

(Starr et al 1996, Zeng et al 1999, Kraus et al 2000, Rance et al; 2002; 2004; 2005, Zeng and Liu, 2006)

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)

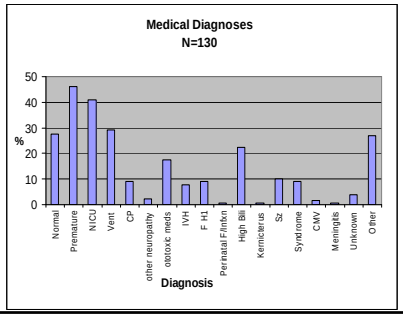
Diagnostic Criteria

- Minimum Test Battery Required for Diagnosis:
  - » Tests of cochlear hair cell (sensory) function:
    - Otoacoustic emissions and/or
    - Cochlear microphonics
- Tests of auditory nerve function:
  - » Click-evoked auditory brainstem response (ABR) to high-level click stimuli

Comprehensive Evaluations Following Diagnosis with ANSD

- Otologic
- Radiologic imaging (MRI/CT)
- Neurologic
- Medical Genetics
- Ophthalmologic
- Pediatric and Developmental Evaluations
- Communication Assessment

Why Comprehensive Medical Evaluation is Important
72% have positive history of other medical diagnoses



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); Buchman et al. (2006)

Auditory Neuropathy Spectrum Disorder: Challenges/Questions

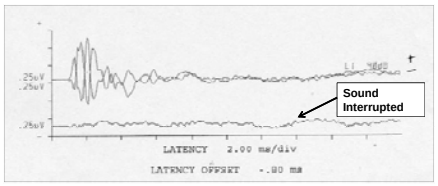
- Making the diagnosis and counseling families
- Determining if hearing aids will be helpful for the short term or the long term?
- What constitutes an adequate trial period with amplification?
- How do we determine who will benefit from hearing aids or cochlear implants?
- Will alternative hearing aid or cochlear implant speech processing strategies result in better performance?
- What communication approach is best?

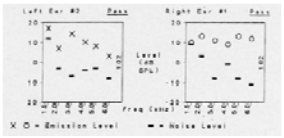
DIAGNOSIS OF AUDITORY NEUROPATHY

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

Typical Pattern of ANSD Abnormal ABR with Present CM





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

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
- Amplification should be fitted as soon as ear specific 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

Factors affecting outcomes after hearing aid fitting

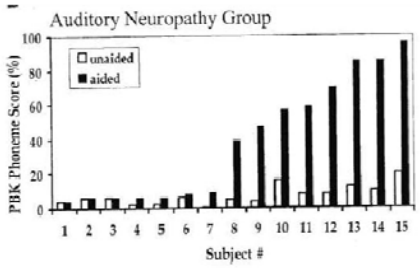
For all children benefit from a particular technology will depend on several factors including

- » 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

Evidence regarding amplification in children with ANSD

- Rance et al 2002
  - » Comparison of unaided and aided speech perception abilities in group of 15 children with AN/AD compared to group of children with typical SNHL
  - » Results show ~50% of group showed significant open-set speech improvements; ~50% of group showed no open-set speech perception ability.

Hearing Aids in Children with AN/AD: 50% Benefit from Hearing Aids



Rance et al Ear and Hearing 2002

What have we learned about ANSD and cochlear implants at UNC?

How does this impact cochlear implant patient candidacy considerations and counseling?

How do we manage cochlear implants in children with ANSD?

Cochlear Implant Candidacy 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

What have we learned?

- **Etiology matters**
- **Tremendous variability exists**
- **Other co-morbid conditions affect performance and make patient management challenging**

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%)

Buchman, C, Roush P, Teagle H, Brown C, Zdanski C, Grose J. Auditory neuropathy characteristics in children with cochlear nerve deficiency. *Ear Hear*. 2006 Aug;27(4):399-408

Cochlear Nerve Deficiency

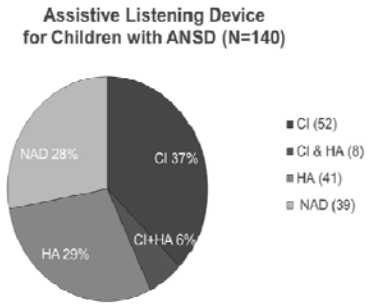
- Absent 8th Nerve
  - not uncommon
  - can result in auditory neuropathy phenotype
  - commonly has normal internal auditory canal (IAC) morphology
  - commonly has normal labyrinth
- Need MRI instead of CT in all kids
  - with profound hearing loss
  - with auditory neuropathy phenotype
  - Audiological, educational, and medical recommendations will be influenced by these findings e.g. CI or HA candidacy

UNC-Chapel Hill : ANSD Study 2010

- 140 infants and children with ANSD at UNC
- All patients evaluated by pediatric audiologist & otologist
- 58 (37%) received cochlear implants
  - » 50 (88%) had bilateral ANSD
  - » 8 (12%) had unilateral ANSD
  - » 52 received CI in ANSD ear
  - » Mean age @ report 88 months (range 14-241 mos)
  - » Mean age @ implant 47 months (range 12-203 mos)
  - » Mean duration of use 41 months (range 6 to 118 mos

Teagle, HFB, Roush, PA, Woodard, JS, Hatch, D.R., Buss, E, Zdanski, CJ, Buchman, CA, Cochlear Implantation in Children with Auditory Neuropathy Spectrum Disorder *Ear and Hearing,* 31(3), 2010

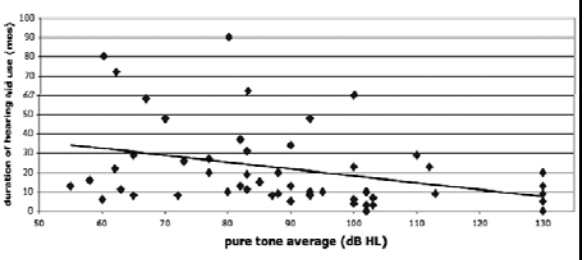
UNC-Chapel Hill: ANSD Study



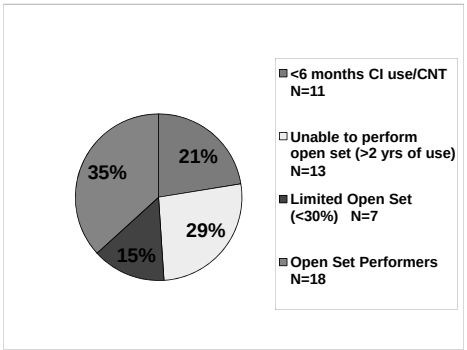
Patient Characteristics

- 42% history of prematurity (<36 weeks gestation)
- Significant co-morbidies
  - » Bronchopulmonary dysplasia (54%)
  - » Necrotizing enterocolitis (18%)
  - » Retinopathy of prematurity (14%)
  - » Intraventricular hemorrhage or hydocephalus (27%)
  - » Hyperbilirubineamia (17%)
  - » Seizure disorder (12%)
  - » Progressive sensorimotor neuropathy (8%)
- 22% no other medical diagnoses
- 27% positive history for family hearing loss

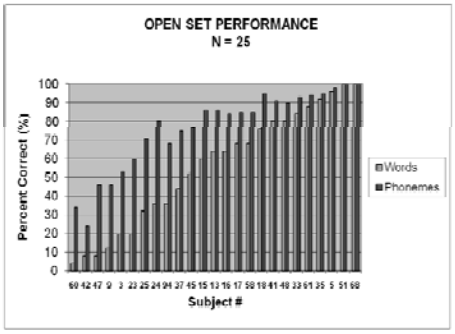
Pre-op PTA for ANSD children receiving CI



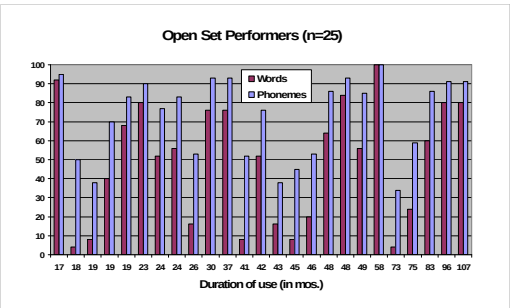
UNC ANSD Children with CI N=52



UNC ANSD Children with CI



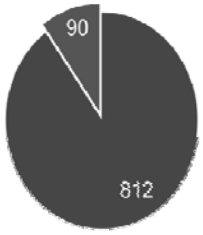
UNC ANSD Children with CI



Second look in 2012…

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?

UNC Children with Cochlear Implants & ANSD (September 2012)

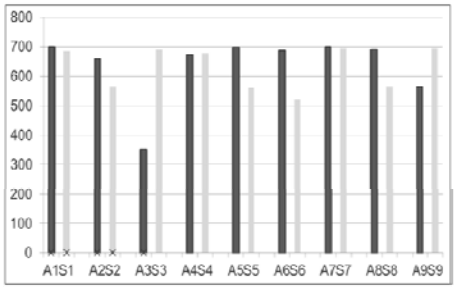


Approximately 10% identified with ANSD (Definitive diagnosis of ANSD not obtained routinely in clinical care until 2000)

Speech Perception Test Battery

- IT-MAIS or MAIS (Parent Questionnaire)
  (Zimmerman-Phillips, et al., 2000; Robbins, et al.,1991)
- Early Speech Perception Test battery (ESP)
  (Moog and Geers, 1990)
- MLNT/LNT words and phonemes
- (Kirk, et al, 1995)
- PB-K words and phonemes
  (Haskins, 1949)
- HINT sentences in quiet and noise conditions

SRI-Q - cumulative quotient of test hierarchy. Child must met 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)



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

ANSD
SNHL

What are we learning?

Electrophysiological measures provide insight on clinical management

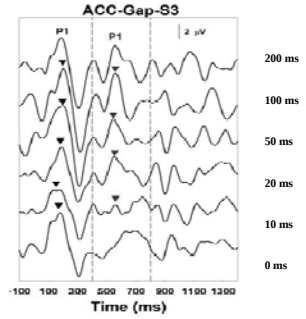
Speech processor programming should be carefully optimized

Cortical Evoked Potentials (CAEPs)

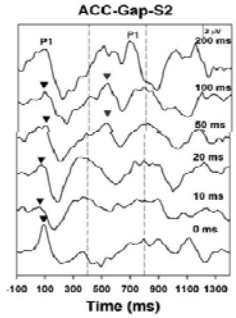
- ABR evaluates outer ear to lower brainstem
- CAEP evaluates outer ear to auditory cortex
- CAEPs not as reliant on timing as earlier evoked potentials and may be present when ABR is not
  - » Hood, 1998, Rapin and Gravel, 2003
- Unlike ABR must be completed in awake (but quiet) infants
  - » Cone Wesson and Wunderlich, 2003)
- CAEP research is ongoing with normal infants and infants with SNHL and ANSD
- One measure is Acoustic Change Complex (ACC)

Cortical Evoked Potentials

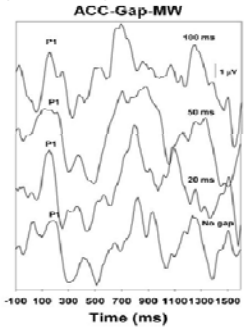
Acoustic Change Complex
ANSD, 60 dB HL, excellent hearing aid user



Acoustic Change Complex
ANSD, 60 dB HL, moderately successful HA user



Acoustic Change Complex
ANSD, 60 dB HL, candidate for cochlear implant



Electrically Evoked Auditory Change Complex in Children with Auditory Neuropathy Spectrum Disorder (ANSD)

Shuman He, PhD; John H. Grose, PhD, Holly FB Teagle, AuD; Craig A. Buchman, MD

University of North Carolina at Chapel Hill

Research Questions

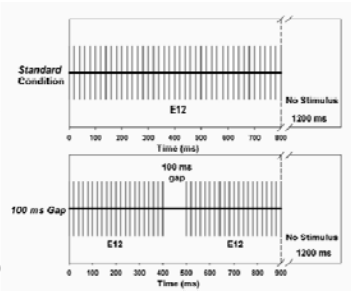
- Do poor performers have temporal processing deficits?
- Do poor performers have spectral resolution deficits?

Methods

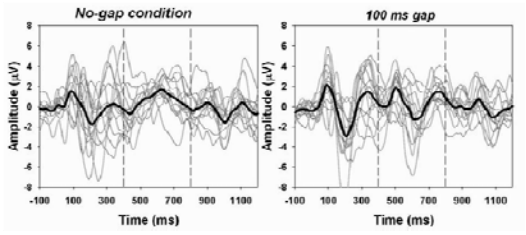
- Subjects:
  - 15 ANSD children with Cochlear Nucleus devices (5.3 to 17.2 yrs; mean: 9.0 yrs).
  - All subjects received their first implants before 4.5 years of age.
  - Good performers: >70% correct on PBK words
- Two stimulating conditions:
  - *Standard condition*
  - *Gap condition*

Methods

- *Standard condition*: 800-ms biphasic pulse train sent to e12
- *Gap condition*:
  - Two 400-ms stimulus bursts separated by a silent interval (i.e. gap)
  - Gap duration tested: 5, 10, 20 50, and 100 ms



Results: Feasibility



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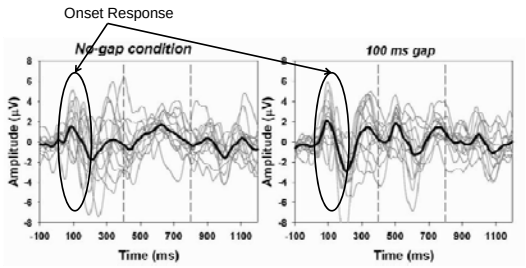

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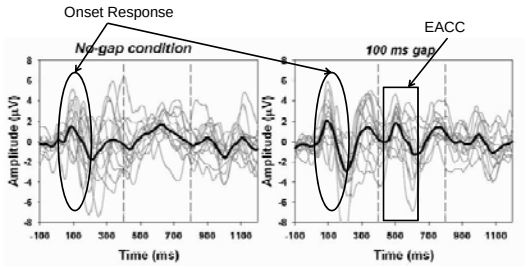
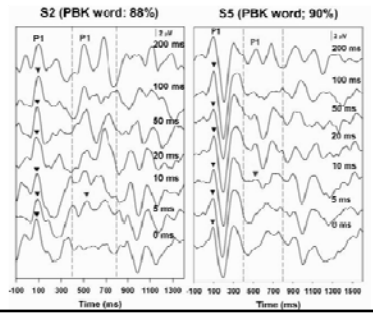


EACC responses were recorded from all ANSD patients using gap stimuli.

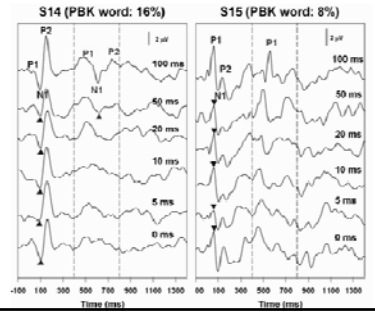
Results: Good Performers

Gap detection threshold (GDT): 5 ms (n=6) or 10 ms (n=4)

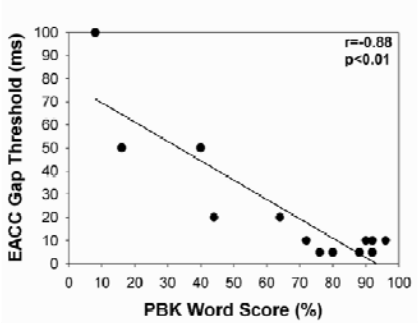


Results: Fair and Poor Performers

GDT: 20 ms (n=2), 50 ms (n=2) or 100 ms (n=1)



Results: Correlation Between the EACC Gap Threshold and PBK Word Score



Research Questions

- Do poor performers have temporal processing deficits?
  - YES.
- Do poor performers have spectral resolution deficits?

Research Questions

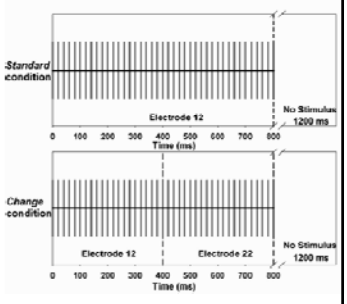
- Do poor performers have temporal processing deficits?
  - YES.
- Do poor performers have spectral resolution deficits?

Methods

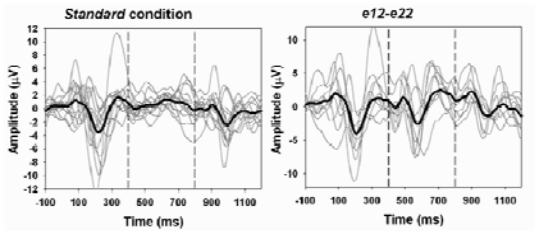
- Subjects
  - 14 ANSD children with Cochlear Nucleus devices (5.4 to 17.1 yrs; mean: 9.1 yrs).
  - All subjects received their first implants before 4.5 yrs of age.
  - Good performers: >70% correct on PBK words
- Two stimulating conditions:
  - *Standard condition*
  - *Change condition*

Methods

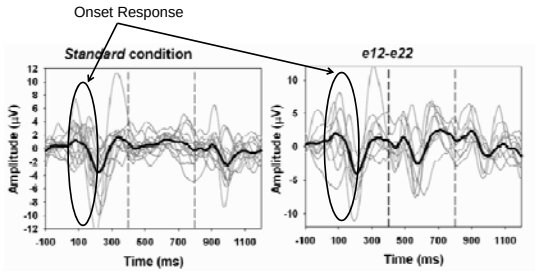
- *Standard condition:* 800-ms biphasic pulse train presented on e12
- *Change condition:*
  - A 800-ms pulse train presented initially on electrode 12, changing to more apical electrodes at 400 ms.
  - Second stimulating electrode: e13-e22.



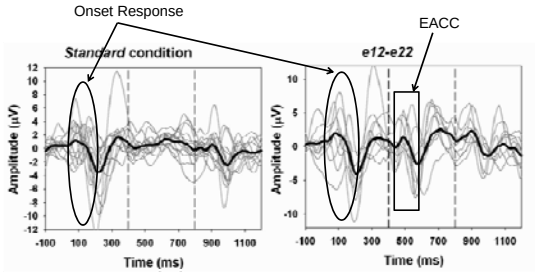
Results: Feasibility



Results: Feasibility

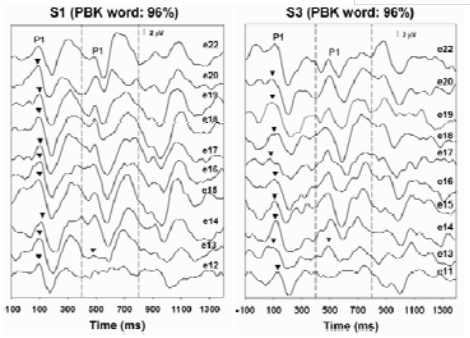


Results: Feasibility

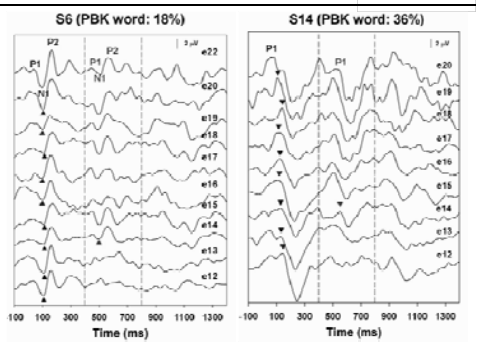


- The EACC in response to changing positions of stimulating electrodes was recorded from all ANSD patients.

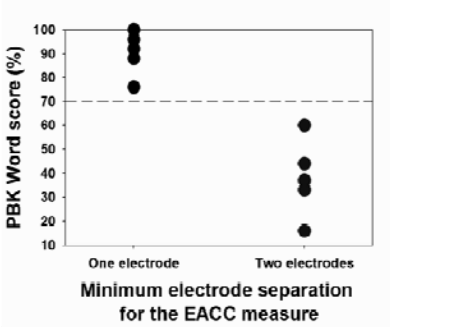
Results: Good Performers



Results: Fair and Poor Performers



Results: EACC thresholds vs PBK Word scores



Research Questions

- Do poor performers have temporal processing deficits?
  - YES.
- Do poor performers have spectral resolution deficits?
  - YES.

Take Home Messages

- Poor performers have temporal processing deficits and/or spectral resolution deficits.
- The EACC can potentially be used as an objective tool to evaluate auditory functions of ANSD patients with CIs.

How do we optimize CI programming for children with ANSD?

- Slow the rate of stimulation
  - » Allow for longer refractory period
- Widen the pulse width
  - » Excite more neural elements at a slower rate
- Measure loudness growth
  - » Optimize the real dynamic range of neural system
- Pitch rank and eliminate channels that create the same percept
  - » Replicate the tonotopic potential that exists for better spectral resolution
- Don't employ strategies that create more channels
  - » Decrease the possibility of spectral and temporal smearing

All these techniques can be implemented through the programming software but they are **not** the default settings and they take more time.

A Different Approach to Programming Advanced Bionics 90k Recipients: Return to Traditional Methods and Concepts

Teagle, Finley, Hatch, Park, Woodard, Strader & Buchman

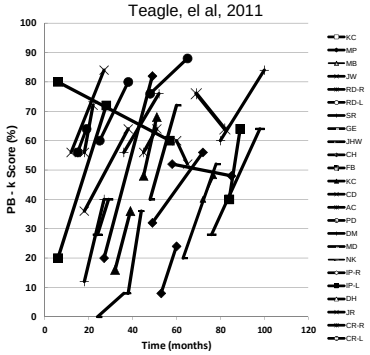
Presented at CI2011 Chicago, IL

- Describe the programming strategy used to improve patient outcomes for children with Advanced Bionics 90K cochlear implants
- Report the outcomes of two subsets of children with 90k devices who have been programmed using this alternative method
- 117 90k Implants
- 69 Attempted re-programming, 3 rejected, 66 preferred
- 48 Using manufacturer advocated approach and are demonstrating progress as expected OR they are no longer followed by our program

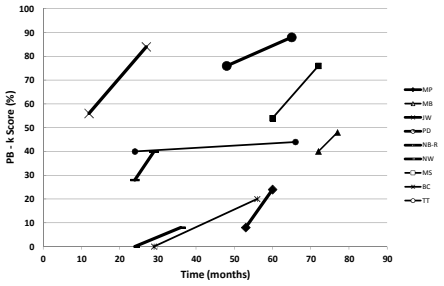
1. Turn off Fidelity 120
2. Use HiRes-S
3. Pitch rank two channels at a time if possible.
4. Turn off out of order or same percept channels.
5. Change the pulse width manually to achieve channel rate of about 1500 pps.
6. Measure Ts and scale Ms using tone bursts on every channel once channels, rate, and PW have been established.

Ears originally programmed in default parameters

Teagle, el al, 2011



Subgroup of 90K children with ANSD



Counseling in ANSD: What Do We Say to Families?

- Child has an auditory disorder
- Degree of deficit may be mild or severe
- Etiology and other medical conditions impact auditory performance
- Child should be enrolled in early intervention as soon as family is ready
- Frequent follow up visits will be necessary
- Results of behavioral response to HA or CI are necessary before communication mode decisions are clear
- Electrophysiological measures may be predictive but we are still learning
- Cochlear implant may be helpful but we will only know if child is fit appropriately and has consistent use
- Monitor continuously, adapt & adjust with time & experience
- Most effective communication strategy will need to be determined with input from family, teachers, therapists, and audiologist

Conclusions

- ANSD is more complicated than originally thought and population more heterogeneous. Therefore it is unlikely that a single approach to management will meet the needs of all children.
- 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. Hearing aids should be fit to behavioral audiograms.
- Some children will benefit from hearing aids either in the short term or the long term, others will require cochlear implantation.
- Regardless of residual hearing, if a child does not make sufficient progress based on measures of communication development, cochlear implantation should be considered.

Conclusions

- A complete medical work up with imaging by MRI is critical.
- Once implanted, speech processor programming should be customized and individualized.
- Many children with CIs will do well using default parameters for speech processor programming; others will benefit from slowing overall rate, widening pulse width and by optimizing tonotopic perception by pitch ranking and eliminating redundant channels
- Electrically evoked auditory event related potentials recorded from ANSD children with CIs may be predictive of speech perception performance.
- Important to use team approach to carefully monitor child's progress in meeting communication goals. Visual support for communication should be discussed and considered on a case by case basis.

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Closing Remarks

- Thank you to our guest presenter, Holly FB Teagle
- Upcoming Events
  - *CEO Web Chat: Thursday, Nov 1st 8 PM Eastern/5 PM Pacific*
  - *Next Cochlear Implant Grand Rounds will be in February, 2013*
  - *Next issue of SoundWaves Newsletter will be released November, 2012*
- Certificate of Attendance: Carissa.moeggenberg@advancedbionics.com

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Thank you for your participation!

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