

# Clinical Applications of Electrocochleography (ECoChG) in Audiology Today

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**Presented by:**

**James W. Hall III, Ph.D.**

**Moderated by:**

**Carolyn Smaka, Au.D., Editor-in-Chief, AudiologyOnline**

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## **Clinical Applications of Electrocochleography (ECochG) in Audiology Today**

**James W. Hall III, Ph.D.**

*Adjunct Professor  
Nova Southeastern University*

*Adjunct Lecturer  
University of Florida*

*Adjunct Professor  
Salus University*

*Extraordinary Professor  
University of Pretoria, South Africa*

[jwhall3phd@gmail.com](mailto:jwhall3phd@gmail.com)  
[www.audiologyworld.net](http://www.audiologyworld.net)

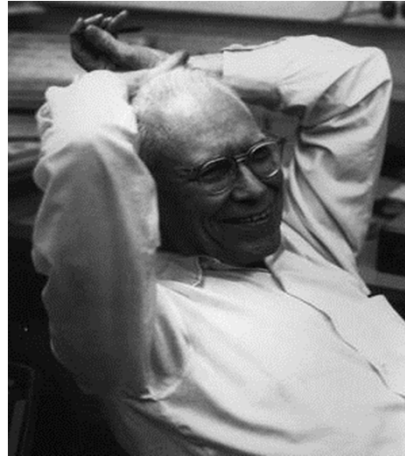
## **Clinical Applications of Electrocochleography (ECochG) in Audiology Today**

- ☐ Introduction: Historical overview of ECochG
- ☐ Review of test protocol for ECochG recording
- ☐ Review of test electrode options for ECochG recording
- ☐ Clinical applications of ECochG in adults
- ☐ Clinical applications of ECochG in the diagnosis of auditory neuropathy spectrum disorder (ANSO)
- ☐ Summary, Questions & Answers

## Original Description of Electrocochleography (ECochG)

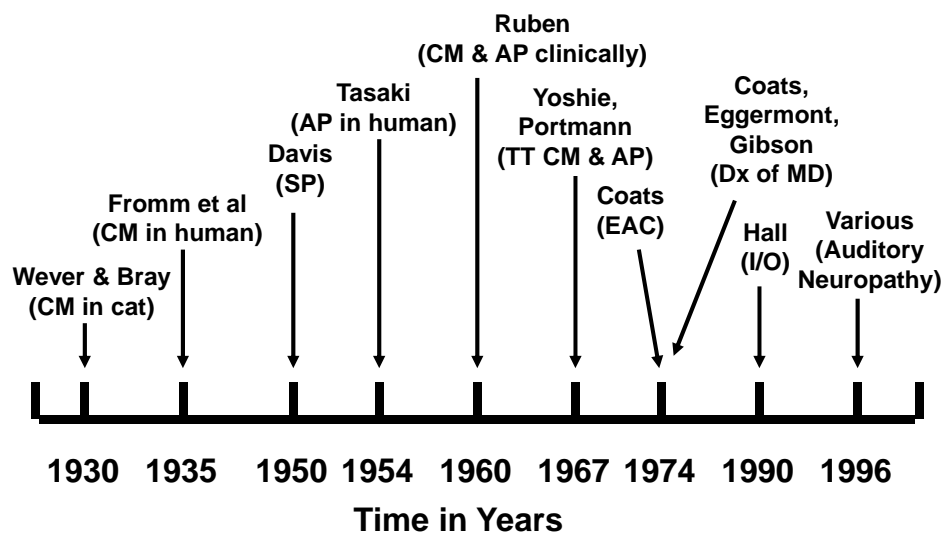
Wever EG and Bray CW. 1930.  
Action currents in the  
auditory nerve in response  
to acoustic stimulation.  
Proceedings of the National  
Acad of Science (USA) 16:  
344-350.

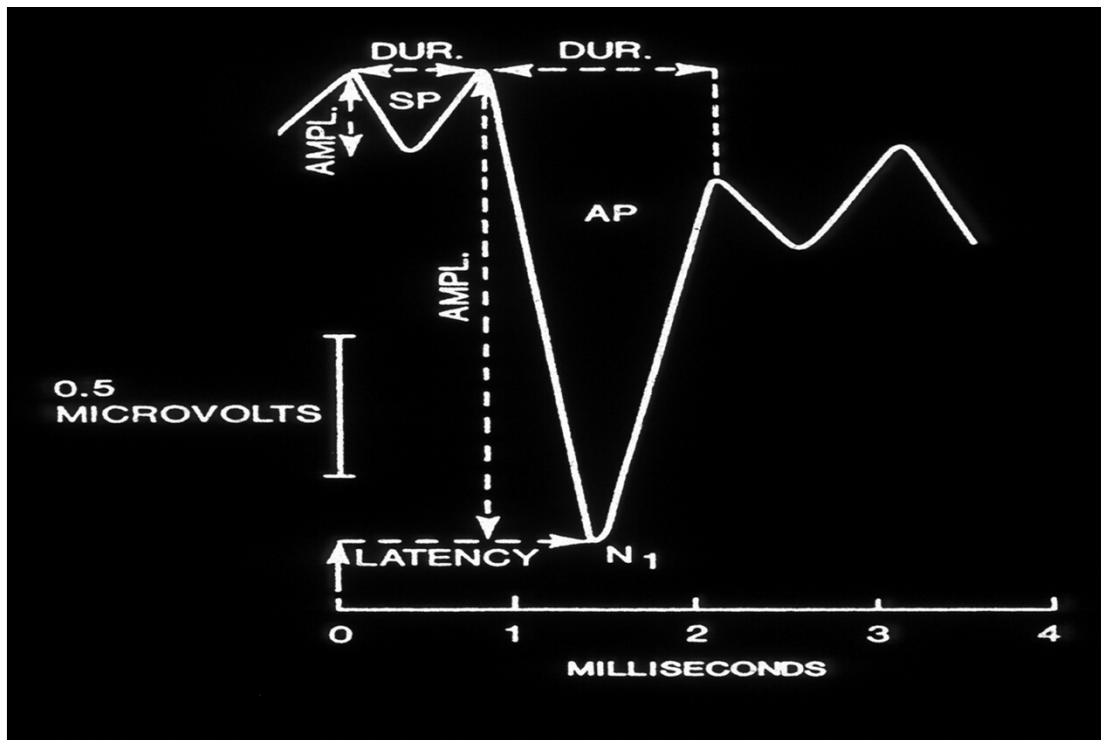
Wever EG and Bray CW. 1930.  
Auditory nerve impulses.  
Science 71: 215.



E. Glen Weaver, Ph.D.  
(October 16, 1902 — September 4, 1991)

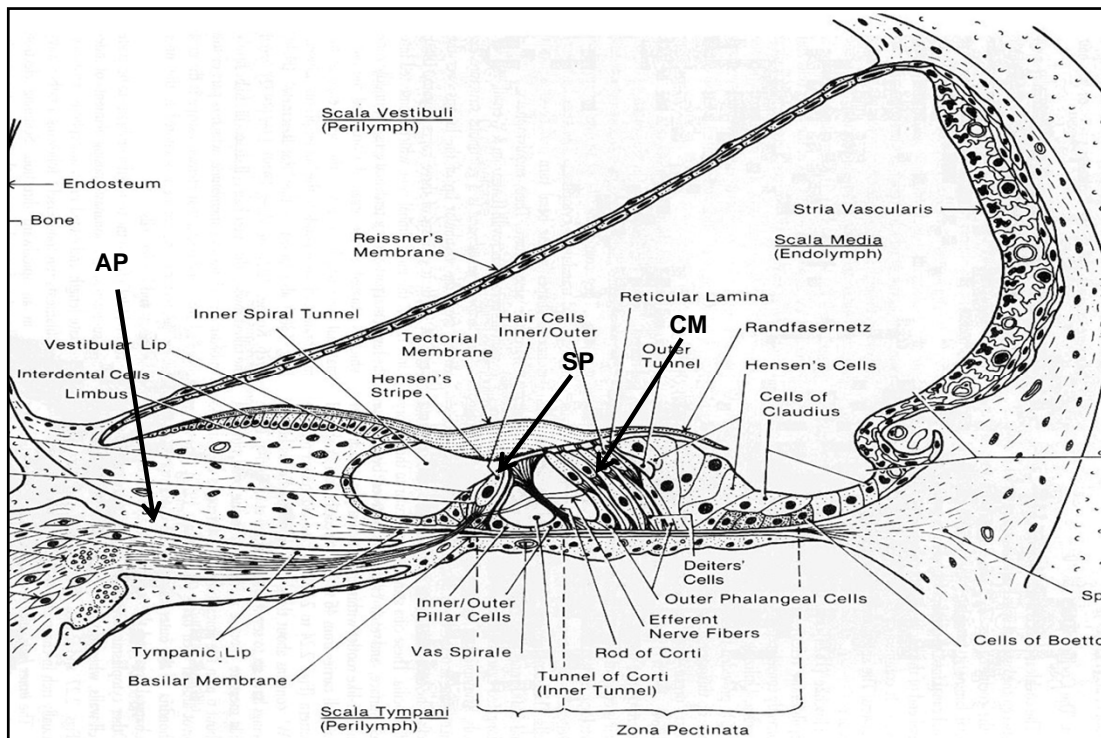
## ELECTROCOCHLEOGRAPHY: *83 Years Old and Still Clinically Important!*





## ELECTROCOCHLEOGRAPHY: Generators

- ❑ Cochlear microphonic (CM)
  - Outer hair cells
  - Receptor potentials
- ❑ Summing potential (SP)
  - Inner hair cells (> 50%)
  - Outer hair cells
  - Organ of Corti
- ❑ Action potential (AP)
  - Afferent fibers in distal 8th cranial nerve
  - Spiral ganglion



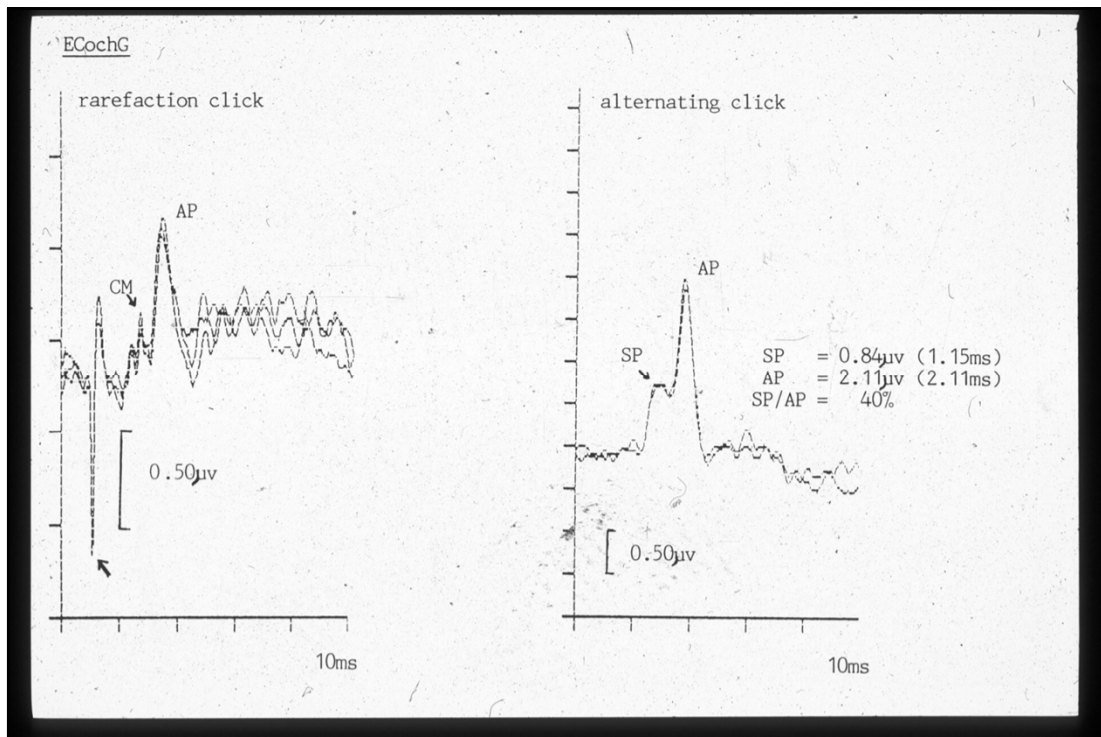
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## ECochG TEST PROTOCOL (1)

### Stimulus Parameters

Type	Clicks
Duration	0.1 ms
Rate	7.1/sec; slower if needed or faster if possible
Polarity	Alternating (for SP and AP) Rarefaction and condensation (for CM)
Intensity	Maximum or lower
Transducer	Insert
Masking	Never needed (Components are biological markers for ear-specific response)

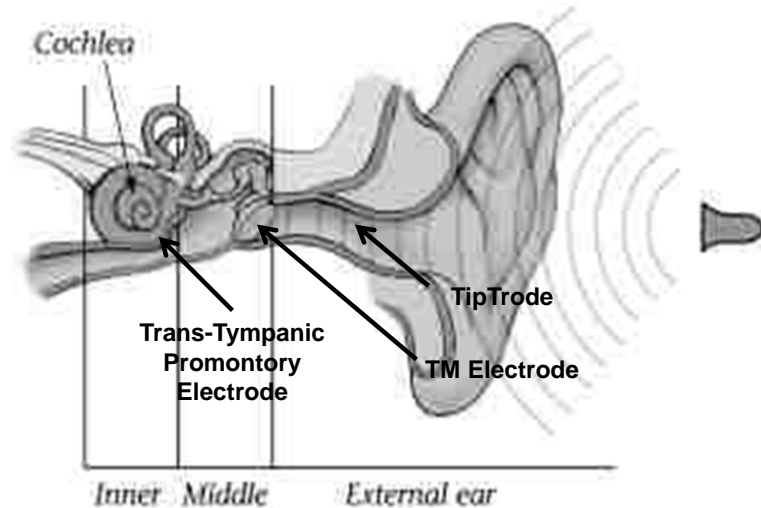


## ECoG TEST PROTOCOL (2)

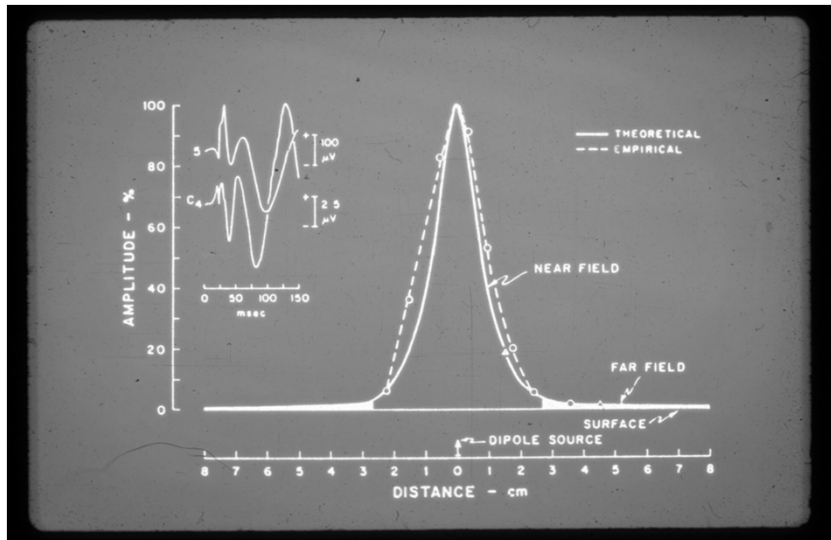
### Acquisition Parameters

Amplification	75,000 or less
Analysis time	5 or 10 ms
Sweeps	500 or less (depends on SNR)
Filters	10 to 1500 Hz
Notch filter	Never
Electrodes	
Option 1	Fz to trans-tympanic needle
Option 2	Fz to tympanic membrane
Option 3	Fz to Tiptrode

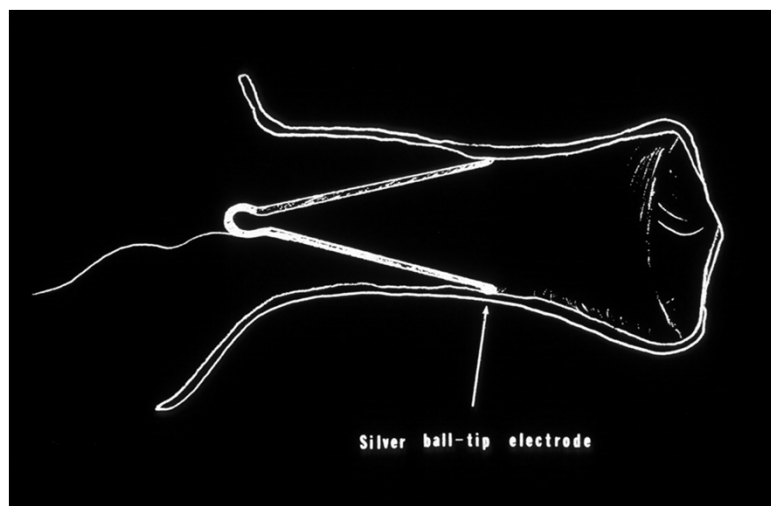
### ECoG Electrode Options: *The Closer to the Cochlea, the Better*



## ECochG is a Near Field Response



## Coats Silver-Ball Extra-Tympanic Electrode (Coats, 1974)





## TIPtrode: Part Transducer and Part Electrode

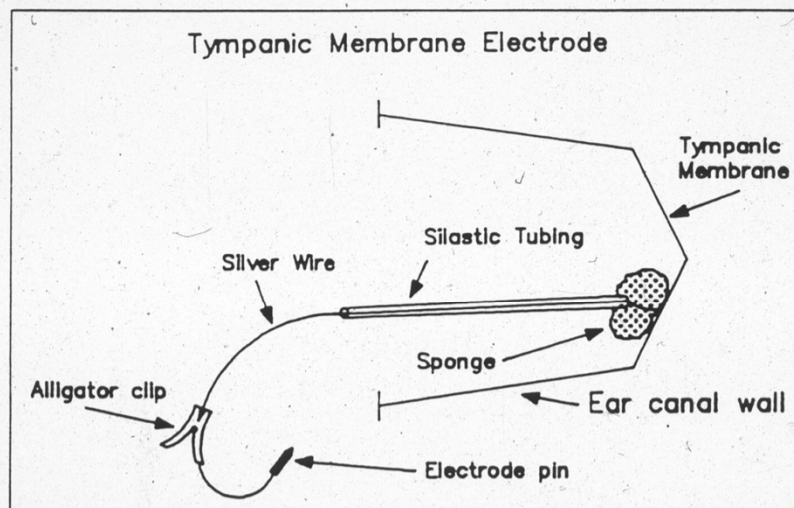
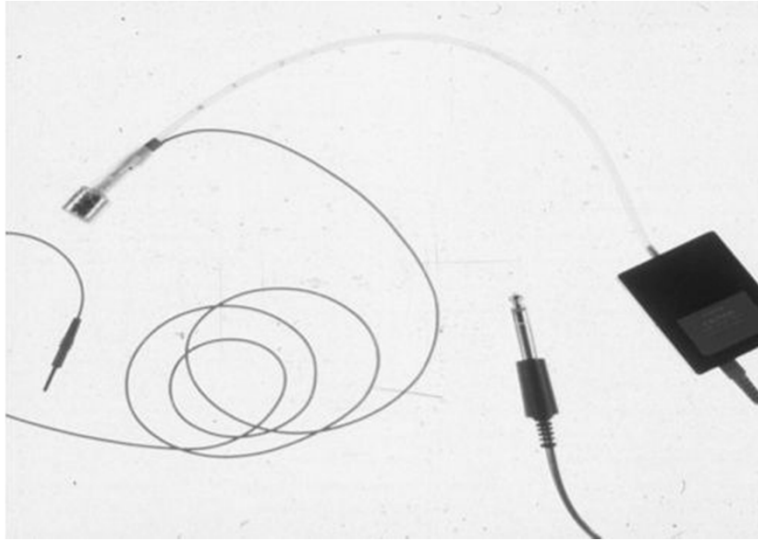
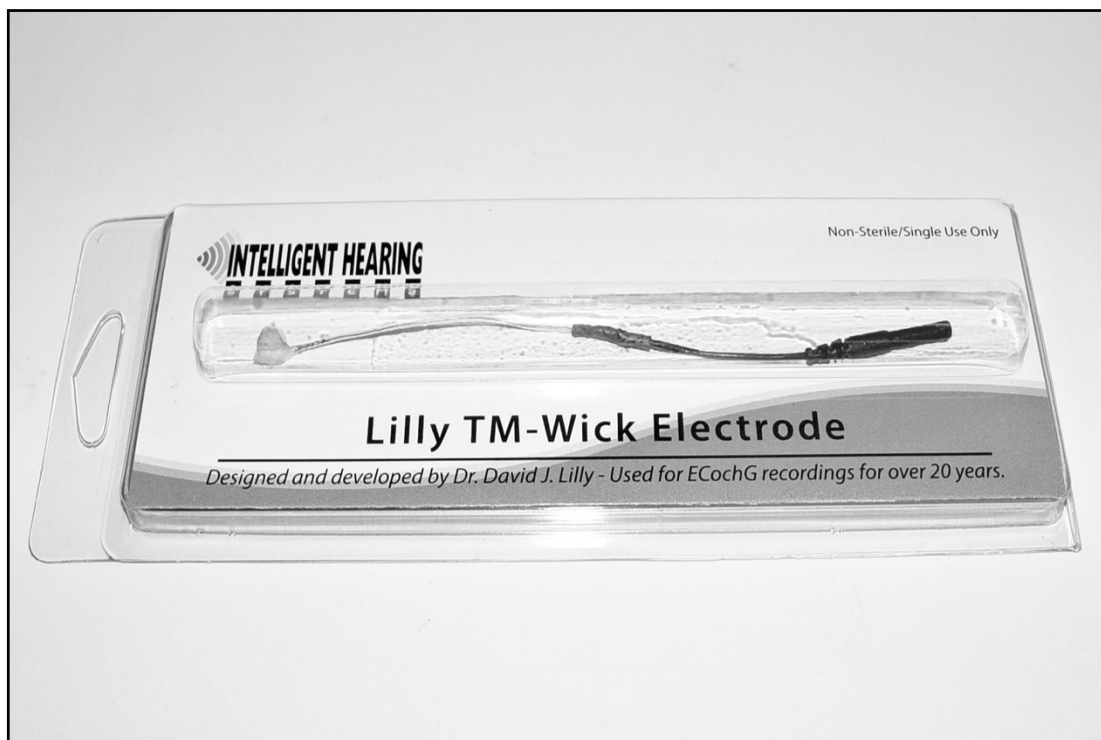
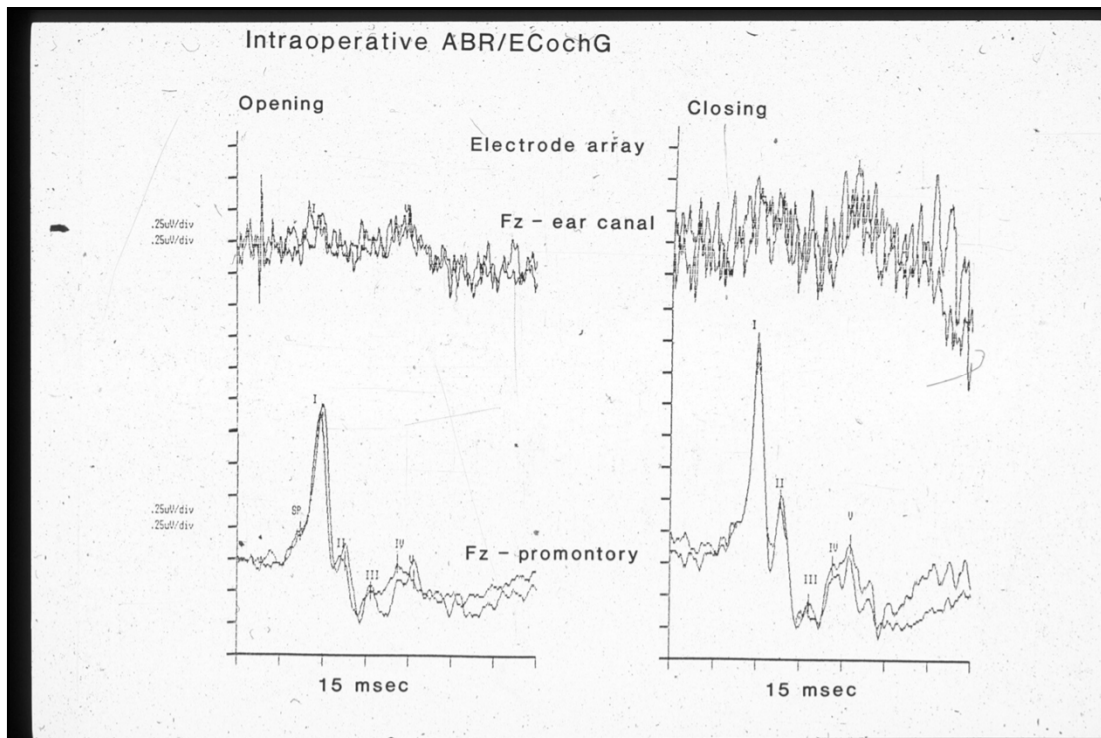


Figure 5-3. TM electrode design (sometimes called a "tymptrode"). Silver wire runs through a flexible plastic tube and connects with sponge or cotton at the tip. When the tymptrode is inserted into the ear canal, the tip makes contact with the lateral surface of the TM.



## **Sub-Dermal Needle Electrode for Trans-Tympanic Promontory ECoG Recording**





## Clinical Applications of Electrocochleography (ECochG) in Audiology Today

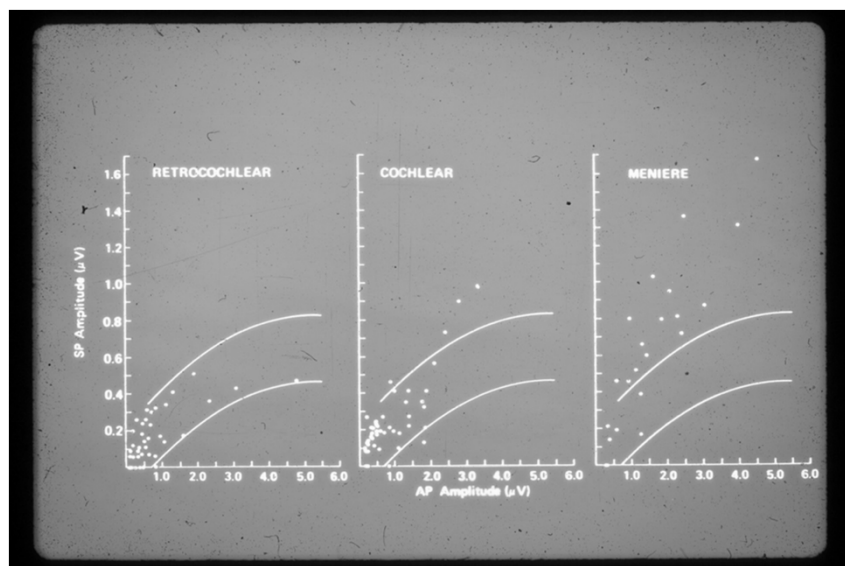
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## **ELECTROCOCHLEOGRAPHY (ECochG): Clinical Applications in Adults**

- ❑ Enhancement of wave I in ABR measurement with air- and bone conduction click stimulation to:
  - Enhance inter-wave latency analysis
  - Confirm ear-specific test findings (biological marker)
  - Minimize the need for masking non-test ear
- ❑ Intra-Operative Neurophysiological Monitoring
  - Prompt documentation of cochlear status
  - Enhance inter-wave latency analysis
  - Minimize interference of electrical artifact
- ❑ Diagnosis of Meniere's disease

### **ECochG in Diagnosis of Meniere's Disease: Abnormally Large SP/AP Ratio**

*Coats AC (1981). The summing potential in Meniere's and non-Meniere's ears. Archives of Otolaryngology, 107, 199-208*



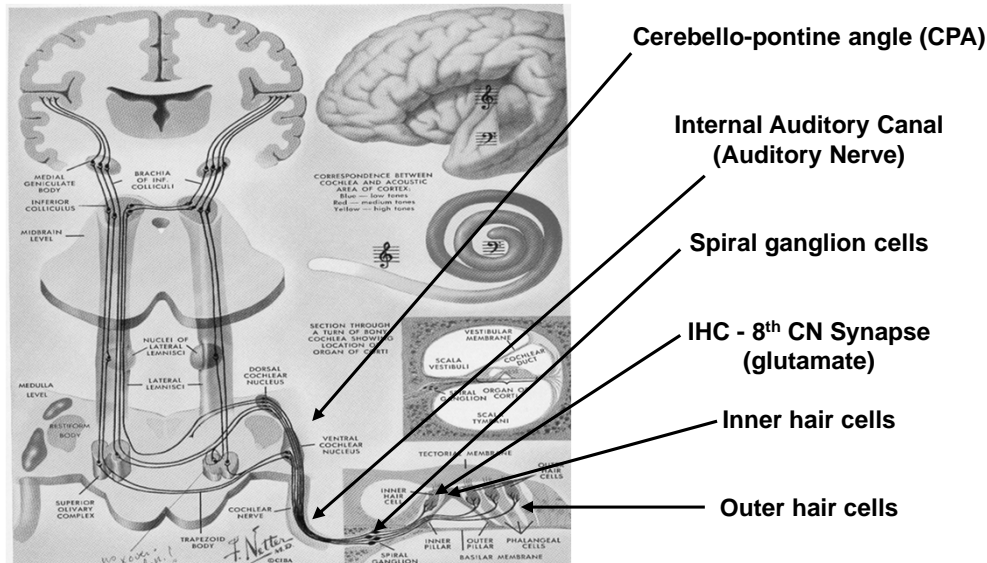
**ELECTROCOCHLEOGRAPHY: Selected Papers**  
(PubMed Search at [www.nlm.nih.gov](http://www.nlm.nih.gov) with key words “Meniere’s Disease” and “electrocochleography” showed 1400 publications)

- ❑ Pou, Hirsch, Durrant et al. (1996). Efficacy of tympanic ECoChG in the diagnosis of endolymphatic hydrops. *AJO*, 17
- ❑ Durrant et al. (1998). Are inner or outer hair cells the source of summing potentials recorded from the round window? *JASA*, 104
- ❑ Levine, Margolis, Daly. (1998). Use of ECoChG in the diagnosis of Meniere’s Disease. *Laryngoscope*, 108
- ❑ Orchik, Shea, Ge. (1998). Summing potential and action potential ratio in Meniere’s Disease before and after treatment. *AJO*, 19
- ❑ Ferraro JA. (2010) Electrocochleography: A review of recording approaches, clinical applications, and new findings in children and adults, *JAAA*, 21, 145-152
- ❑ Hall JW III (2007). *New Handbook of Auditory Evoked Responses*. Boston: Pearson
- ❑ Atcherson SR & Stoodly TM (2012). *Auditory Electrophysiology: A Clinical Guide*. New York: Thieme

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(ECoChG) in Audiology Today**

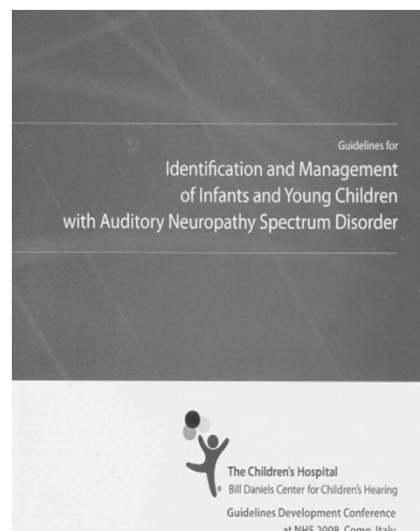
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## Essential Role of Electrocochleography (ECoG) in the Diagnosis and Management of Auditory Neuropathy Spectrum Disorder (ANSD)



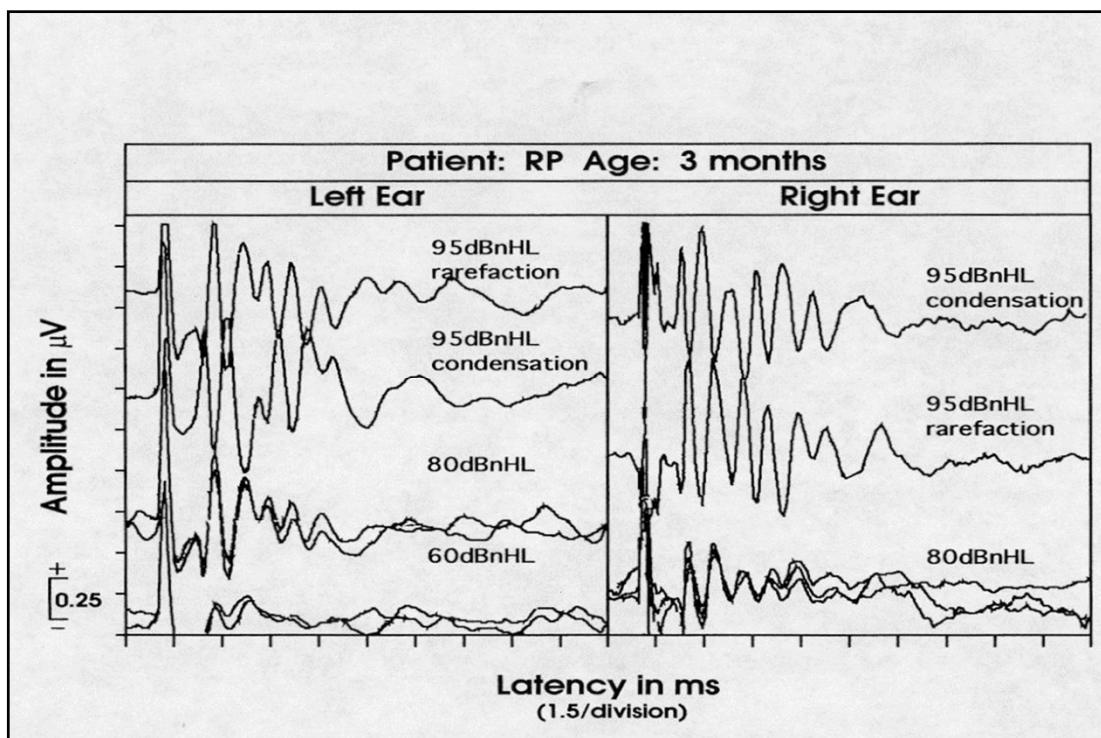
## Electrococheography (ECoG): Diagnosis of Auditory Neuropathy Spectrum Disorder

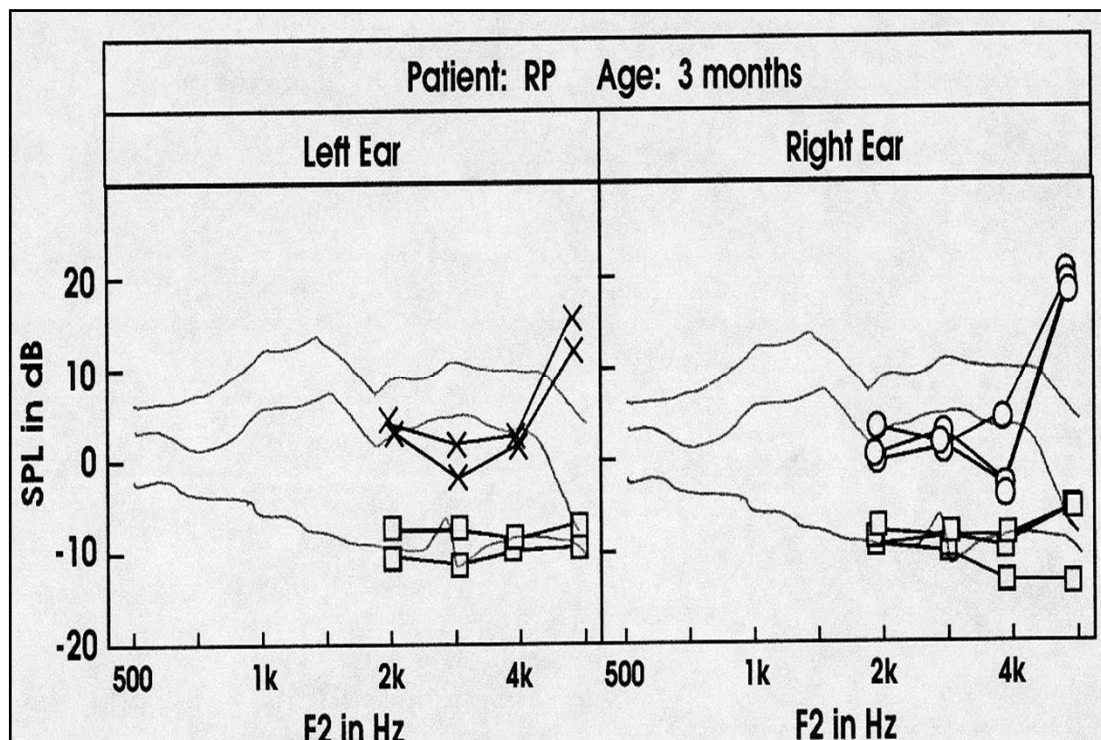
- ❑ In June 2008, at the invitation of Deborah Hayes, a panel of experts met in Como, Italy, at the NHS 2008 Conference to develop Guidelines for the Identification and Management of Infants and Young Children with Auditory Neuropathy.
- ❑ The panel consisted of:
  - Yvonne Sininger, Ph.D.
  - Arnold Starr, M.D.
  - Christine Petit, M.D., Ph.D.
  - Gary Rance, Ph.D.
  - Barbara Cone, Ph.D.
  - Kai Uus, M.D., Ph.D.
  - Patricia Roush, Au.D.
  - Jon Shallop, Ph.D.
  - Charles Berlin, Ph.D.



## Identification and Diagnosis of Auditory Neuropathy Spectrum Disorder (ANSD): Minimal Test Battery (2008 ANSD Guidelines)

- ❑ Tests of cochlear hair cell function
  - Otoacoustic emissions (OAEs)
  - Cochlear microphonic (ECochG and ABR)
    - ✓ CM may be present when OAEs are absent (e.g., with middle ear dysfunction)
- ❑ Tests of auditory nerve function
  - ABR for high intensity click stimulation (e.g., 80 to 90 dB nHL) with separate averages for:
    - ✓ Rarefaction stimulus polarity
    - ✓ Condensation stimulus polarity
- ❑ Additional tests
  - Acoustic reflex measurement (generally acoustic reflexes are absent in ANSD)
  - Suppression of otoacoustic emissions (abnormal, e.g, no suppression in ANSD)





### Essential Role of Electrocochleography (ECoChG) in the Diagnosis and Management of Auditory Neuropathy Spectrum Disorder (ANSD): Medical diagnoses (1)

#### □ Perinatal Diseases

- Hyperbilirubinemia
- Hypoxic insults
- Ischemic insults
- Prematurity

#### □ Neurological Disorders

- Demyelinating diseases
- Hydrocephalus
- Immune disorders, e.g., Guillain-Barre syndrome
- Inflammatory neuropathies
- Severe developmental delay



## **Essential Role of Electrocochleography (ECoChG) in the Diagnosis and Management of Auditory Neuropathy Spectrum Disorder (ANSD): Medical diagnoses (2)**

- ☐ **Neuro-metabolic diseases**
- ☐ **Genetic and Hereditary Etiologies**
  - Family history
  - Connexin mutations, e.g., GJB3 (D66del)
  - Otoferlin (OTOF) gene
  - Non-syndromic recessive auditory neuropathy
  - Hereditary motor sensory neuropathies (HMSN), e.g., Charcot-Marie-Tooth syndrome
  - Leber's hereditary optic neuropathy
  - Waardenburg's syndrome
  - Neurogenerative diseases, e.g., Friedreich's ataxia
- ☐ **Mitochondrial disorders, e.g., mitochondrial enzymatic defect**

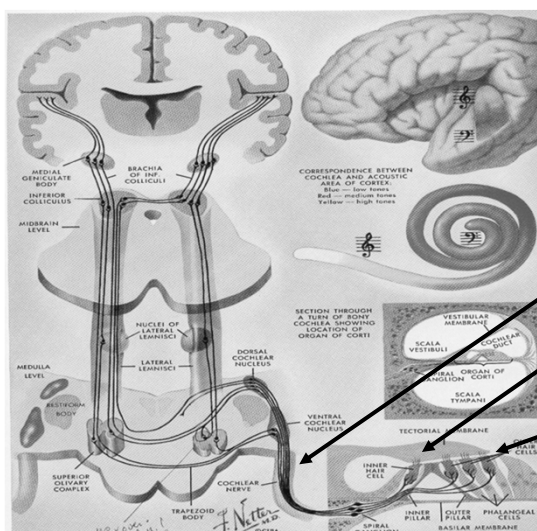
## **Other Procedures Important in the Diagnosis and Management of Auditory Neuropathy Spectrum Disorder (2008 Guidelines)**

- ☐ **Components of assessment**
  - Pediatric and developmental history
  - Otologic evaluation, plus
    - ✓ Imaging of cochlea with CT
    - ✓ Imaging auditory nerve with MRI
  - Medical genetics evaluation
  - Ophthalmologic evaluation
  - Neurological evaluation to assess:
    - ✓ Peripheral nerve function
    - ✓ Cranial nerve function
  - Communication assessment

## Comprehensive Assessment of Auditory Neuropathy Spectrum Disorder (ANSD): MRI of Auditory Nerve

- ❑ **Brainstem and inner ear abnormalities in children with auditory neuropathy spectrum disorder and cochlear nerve deficiency.** Huang et al. (UNC). *American J Radiol*, 31, 2010
  - CND was identified in 33.0% of children and 26.9% of ears with ANSD
  - Significantly more patients with bilateral CND had intracranial abnormalities than those with unilateral CND (60.0% versus 15.8%).
- ❑ **Unilateral auditory neuropathy caused by cochlear nerve deficiency.** Liu et al (China). *Int J Otolaryngol*, 2012.
  - Cochlear nerve deficiency can be seen by electrophysiological evidence and may be a significant cause of unilateral AN.
  - Inclined sagittal MRI of the internal auditory canal is recommended for the diagnosis of this disorder.

## Auditory Neuropathy Spectrum Disorder (ANSD): Defining Site of Dysfunction is the Key to Accurate Diagnosis and Effective Management



Action potential (AP)

Summating potential (SP)

Cochlear microphonic (CM)

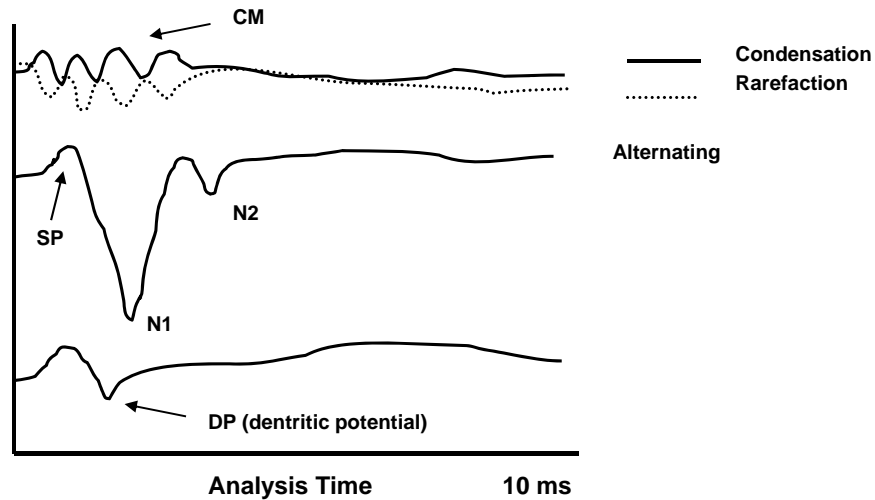
## **ECochG in ANSD: Refining diagnosis of “site of lesion” (1)**

- Santarelli, Starr, Michalewski & Arslan (2008). Neural and receptor cochlear potentials obtained by transtympanic electrocochleography in auditory neuropathy. *Clinical Neurophysiology*, 119, 1028-1041.
  - 8 subjects (with AN versus 16 normal subjects)
  - AN subjects between 5 and 48 years of age
  - Diagnosis based on presence of DPOAEs and absence of ABR (incl. wave I)
  - Enlarged CM in AN patients (Starr et al, 2001; Santarelli & Arslan, 2002)
  - Etiology
    - ✓ Hereditary (3)
    - ✓ Immunological (3)
    - ✓ Degenerative (1)
    - ✓ Congenital (1)
  - ECochG measures included
    - ✓ CM
    - ✓ SP
    - ✓ AP
    - ✓ Adaptation of AP determined by AP elicited by a first click versus AP elicited by a train of 10 rapid clicks (2.9 ms ISI)

## **ECochG in ANSD: Refining diagnosis of “site of lesion” (2)**

- McMahon, Patuzzi, Gibson & Sanli. (2008) Frequency-specific electrocochleography indicates that presynaptic and postsynaptic mechanisms of auditory neuropathy exist. *Ear & Hearing*, 29, 314-325.
  - 14 subjects (7 male and 7 female) with AN versus 2 normal subjects
  - AN diagnosed between 3 and 24 months of age
  - Diagnosis based on large CM potentials and absence of ABR (incl. wave I)
  - Genetic etiology for 6 subjects
  - Severe to profound audiometric thresholds for all subjects
  - All subjects received cochlear implants
  - Purpose of study was to better define physiology mechanisms of AN to guide management (including cochlear implantation)
  - ECochG recorded with
    - ✓ Non-inverting (“active”) electrode near round window “golf club” electrode (via myringotomy)
    - ✓ Inverting electrode on ipsilateral earlobe
  - ECochG in AN consistent with:
    - ✓ Pre-synaptic mechanism (abnormal SP) = good EABR and CI benefit
    - ✓ Post-synaptic mechanism (normal SP + dendritic potential) but no AP = poor or absent EABR and poor CI benefit

### ECochG in ANSD: Examples of ECochG Components (McMahon et al, 2008)



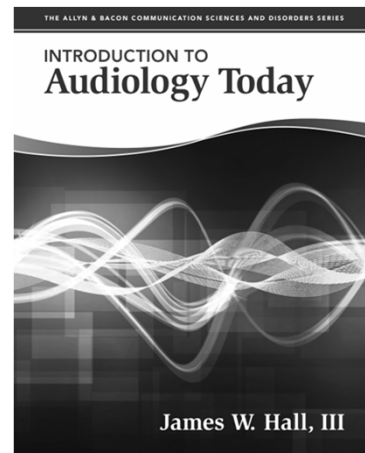
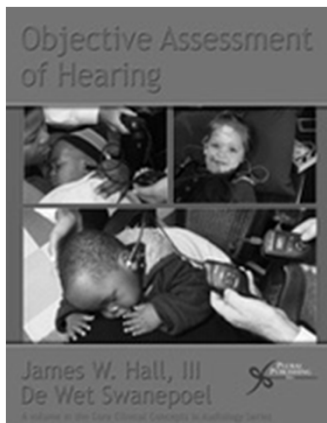
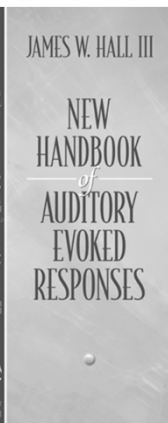
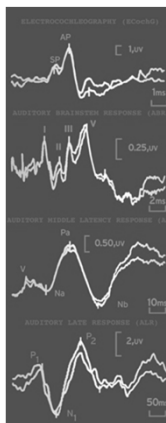
### Auditory Neuropathy Spectrum Disorder (ANSD): Audiological Management

- ❑ Close monitoring every three months until behavioral audiometry is complete
  - More accurate results are obtained over time
  - Significant improvement, including “recovery”, is possible
- ❑ Monitor OAEs
- ❑ Referral to other disciplines (pediatric neurology, genetics, ophthalmology, ENT)
- ❑ Hearing aids on trial basis with evidence of either:
  - Elevated pure tone or speech thresholds
  - Behavioral observation consistent with abnormal auditory sensitivity
- ❑ Cochlear implant for pre-synaptic forms of ANSD
- ❑ Assistive listening devices
  - Phonak EduLink
  - With hearing aids
  - With cochlear implants
- ❑ Alternative communication strategies
  - ♦ Cued speech
  - ♦ Visual emphasis aural approaches
  - ♦ Signing options (e.g., [www.BabySigns.com](http://www.BabySigns.com))

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**Thank You!**  
***Questions?***



**Pearson**  
**July 2013**

## **Update on Auditory Electrophysiology: Evidence-Based Clinical Applications**

Application of ABR in Objective Assessment of Infant Hearing  
James W. Hall III, PhD

Clinical Applications of Electrocochleography in Audiology Today  
James W. Hall III, PhD

Neurodiagnostic Auditory Evoked Responses Applications  
Samuel R. Atcherson, PhD

Cortical Response Applications for Audiometric and Audibility  
Assessment  
Samuel R. Atcherson, PhD

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