Clinical Applications of Electrocochleography (ECochG) in Audiology Today

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- 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 (ANSD)
- Summary, Questions & Answers
Original Description of Electrocochleography (ECoChG)


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

ELECTROCOCHLEOGRAPHY: 83 Years Old and Still Clinically Important!


Time in Years
ELECTROCOCHLEOGRAPHY: Generators

- Cochlear microphonic (CM)
  - Outer hair cells
  - Receptor potentials

- Summating 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**

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

![Graphs showing rarefaction and alternating clicks](image)

- **Rarefaction Click**
  - CM
  - AP
  - SP = 0.8 μV (1.15 ms)
- **Alternating Click**
  - AP
  - SP = 2.1 μV (2.11 ms)
  - SP/AP = 40%
ECochG 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

ECochG 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 ECochG Recording
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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 summating 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 with key words “Meniere’s Disease” and “electrocochleography” showed 1400 publications)

- Durrant et al. (1998). Are inner or outer hair cells the source of summating potentials recorded from the round window? *JASA, 104*
- Orchik, Shea, Ge. (1998). Summating potential and action potential ratio in Meniere’s Disease before and after treatment. *AJO, 19*

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

- Cerebello-pontine angle (CPA)
- Internal Auditory Canal (Auditory Nerve)
- Spiral ganglion cells
- IHC - 8th CN Synapse (glutamate)
- Inner hair cells
- Outer hair cells

Electrocochleography (ECochG): 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 (ECoG) 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%).

  - 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)

  - 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)

  - 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)

ECochG waveforms:
- CM (Condensation)
- SP (Sponaneous Potentials)
- N1, N2
- DP (Dendritic Potential)

Analysis Time: 10 ms

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)
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Thank You!

Questions?
Update on Auditory Electrophysiology: Evidence-Based Clinical Applications

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

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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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