Electrophysiology Assessment of Infant Hearing: An Evidence-Based Protocol

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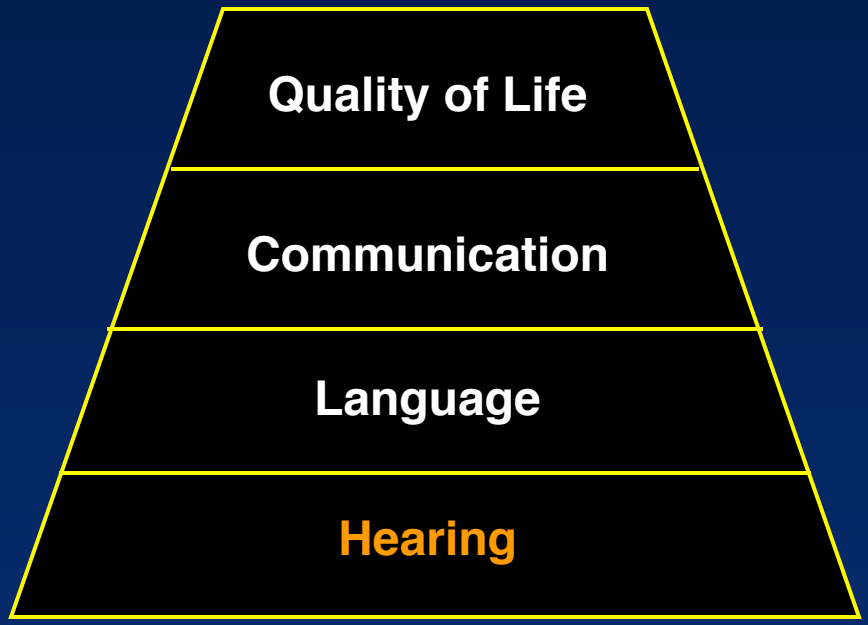
Electrophysiology Assessment of Infant Hearing: An Evidence-Based Protocol

- Early diagnosis and intervention improves communication
- Accurate assessment of infant hearing is standard of care
- Efficient identification of (screening for) hearing loss
- The cross-check principle still rules
- An objective auditory test battery
  - Acoustic immittance measures
  - OAEs
  - ABR
  - ASSR
  - ECochG
- Pulling it all together

Hearing: An Important Building Block in the Foundation for Communication and Quality of Life



Evidence-Based Efficient and Effective Identification and Diagnosis of Infant Hearing Loss

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A Common Evidence Grading System: *Four Categories*

- Grade 1
  - 1a: Well-designed meta-analysis of randomized controlled trials
  - 1b: Well-designed randomized controlled trials
- Grade 2
  - 2a: Well-designed controlled studies without randomization
  - 2b: Well-designed quasi-experimental studies
- Grade 3: Well-designed non-experimental studies, i.e.,
  - Correlational studies
  - Case studies
- Grade 4:
  - Expert committee reports, consensus conferences and clinical experience

Evidence-Based Efficient and Effective Identification and Diagnosis of Infant Hearing Loss: *Clinical Guidelines*

- Joint Committee on Infant Hearing Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Pediatrics, 120*, 2007-2333, 2007
- 2008 Guidelines on Identification, Diagnosis, and Management of Auditory Neuropathy Spectrum Disorder in Infants and Young Children
- 2012 American Academy of Audiology: Audiologic Guidelines for the Assessment of Hearing in Infants and Young Children
- 2013 American Academy of Audiology Clinical Practice Guidelines: Pediatric Amplification
- Australian Professional Practice Standards. Version 1. (July 2013). Standard Assessment … Paediatric

Example of A Practice Guideline in Audiology: Year 2007 JCIH Position Statement Protocol for Evaluation for Hearing Loss In Infants and Toddlers from Birth to 6 months

- Child and family history
- Evaluation of risk factors for congenital hearing loss
- Parental report of infant's responses to sound
- Audiological assessment
  - Auditory brainstem response (ABR)
    - ✓ Click-evoked ABR with rarefaction and condensation single-polarity stimulation if there are risk factors for auditory neuropathy
    - ✓ Frequency-specific ABR with air-conduction tone bursts
    - ✓ Bone-conduction stimulation (as indicated)
    - ✓ Auditory steady state response (ASSR) is optional
  - Otoacoustic emissions (distortion product or transient OAEs)
  - Tympanometry with 1000 Hz probe tone
  - "Clinical observation of infant's auditory behavior. *Behavioral observation alone is not adequate for determining whether hearing loss is present in this age group, and is not adequate for the fitting of amplification devices."*

Australian Professional Practice Standards (July 2013)
Advanced Audiological Assessment *Clinical Processes*

- Purpose and Aim
- Expected Outcomes
- Clinical Indicators
- Clinical Processes
- Documentation
- Correspondence
- Settings
- Safety
- Equipment Specifications
- Related References

Australian Professional Practice Standards (July 2013)
Advanced Audiological Assessment *Clinical Processes*

- Detailed case history
- Otoscopy
- Audiometry (behavioral AC and BC)
- Speech perception assessment (formal or informal)
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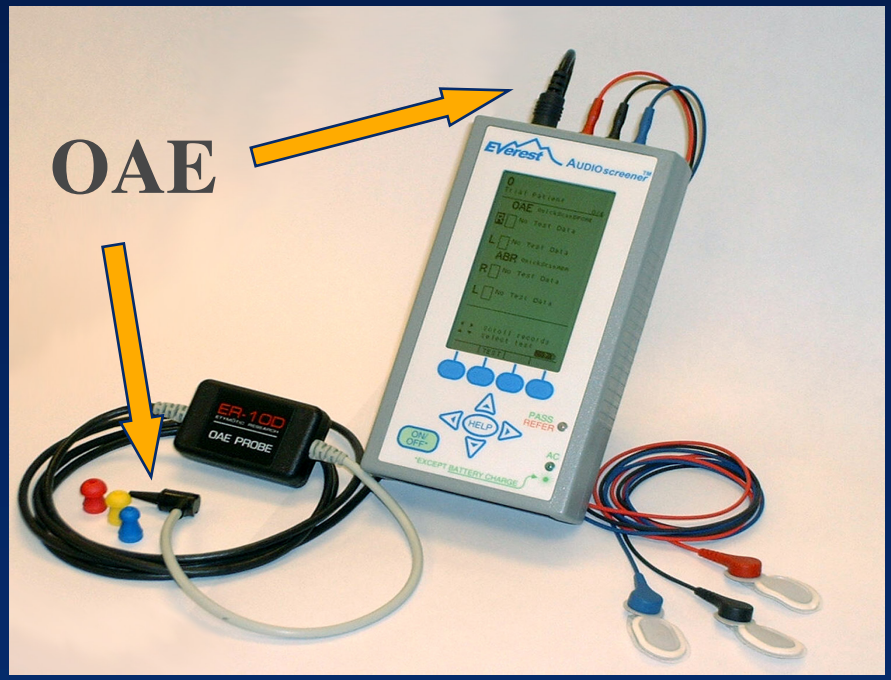
Combined Automated OAE and ABR Technique

AUDIOscreener (GSI)



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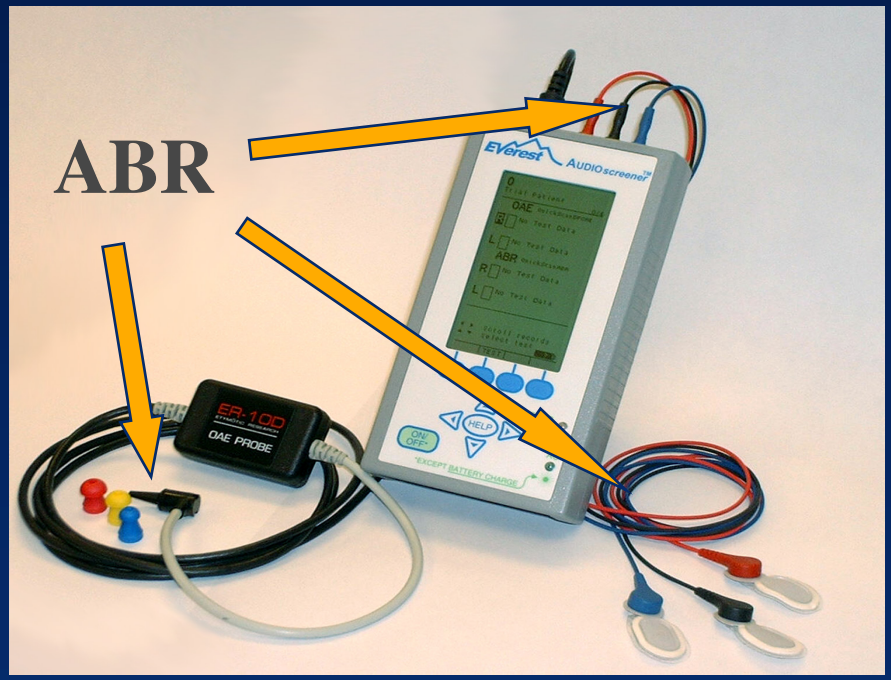

Combination OAE/ABR Screening: Differentiation of Peripheral Auditory Dysfunction

(Hall, Smith & Popelka. Journal of the American Academy of Audiology, August 2004)

Type of dysfunction	OAE	ABR
None	Normal	Normal
External/middle ear *	Abnormal	Normal
Sensory (OHC)	Abnormal	Abnormal
Neural	Normal	Abnormal

** Minor dysfunction in most cases*

Combined OAE and AABR Study: Results (N = 600)

Diagnostic Outcome	Screening Outcome: Pass	Refer	N
Normal	590	2	592
Hearing Impaired	0	8	8
N	590	10	

Sensitivity = 100.0%
Specificity = 99.7%
Refer Rate = 1.7%
Positive Predictive Value = 80.0%

OAE and AABR Screening Techniques: 2007 Joint Committee on Infant Hearing Recommendations

- Well baby nursery (WBN)
  - Screening with OAEs or AABR
  - Refer outcome for AABR
    - ✓ Schedule for diagnostic follow up assessment < 3 months
  - Refer outcome for OAEs?
    - ✓ Immediate follow up screening with AABR
  - Refer outcome for OAEs and AABR?
    - ✓ Schedule for diagnostic follow up assessment < 3 months
- Neonatal intensive care unit (NICU) or intensive care nursery (ICN)
  - Screening AABR (to detect auditory neuropathy)
  - Pass outcome for AABR?
    - ✓ Follow as indicated by risk factors for progressive/delayed onset hearing loss
  - Refer outcome for AABR?
    - ✓ Perform OAEs to identify possible auditory neuropathy
    - ✓ Schedule for diagnostic follow up assessment < 3 months

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- The cross-check principle still rules almost 40 years later
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The Cross-Check Principle in Pediatric Audiology

(Jerger J & Hayes D. Arch Otolaryngol 102: 1976)

The Cross-Check Principle in Pediatric Audiology

(Jerger J & Hayes D. Arch Otolaryngol 102: 1976)

"We have found that simply observing the auditory behavior of children does not always yield an accurate description of hearing loss"…

"The basic operation of this principle is that no result be accepted until it is confirmed by an independent measure."

Test Battery:

- Behavioral audiometry
- Immittance (impedance) measurements
  - ✓ Tympanometry
  - ✓ Acoustic reflexes (contralateral only with SPAR)
- Auditory brainstem response (brainstem-evoked response audiometry or BSER)
  - ✓ Click stimulus air conduction
  - ✓ Click stimulus bone conduction

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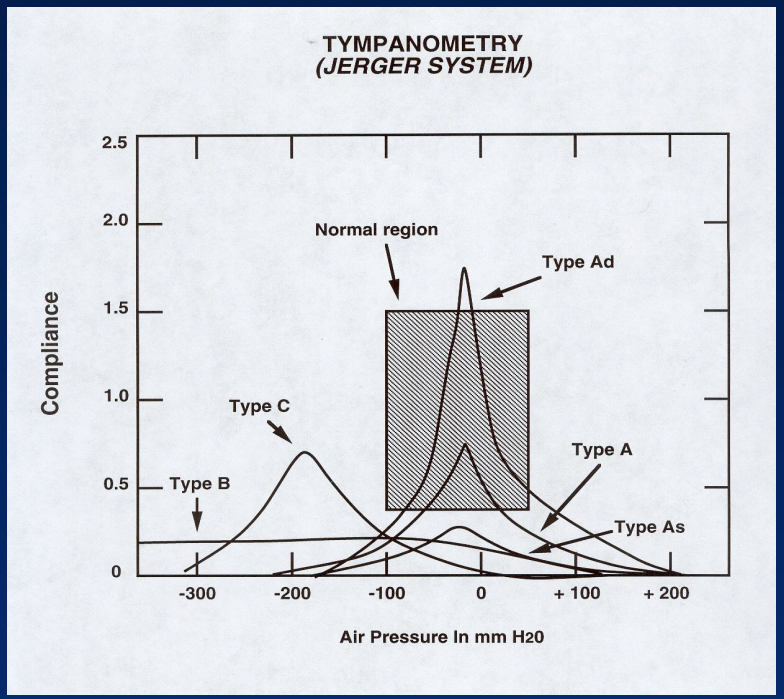
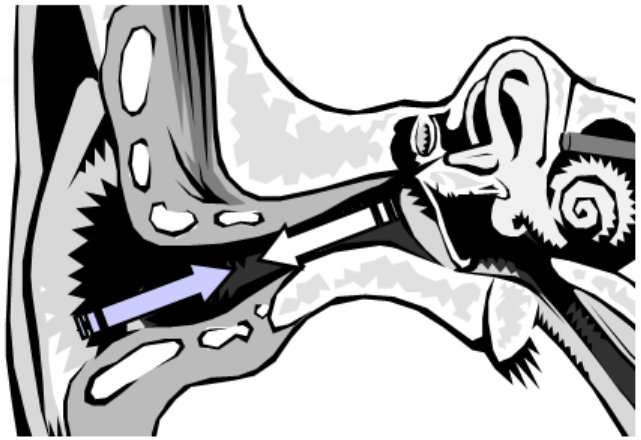
Acoustic Immittance Measures:
Tympanometry with a 1000 Hz Probe Tone



Wideband Reflectance/Absorbance

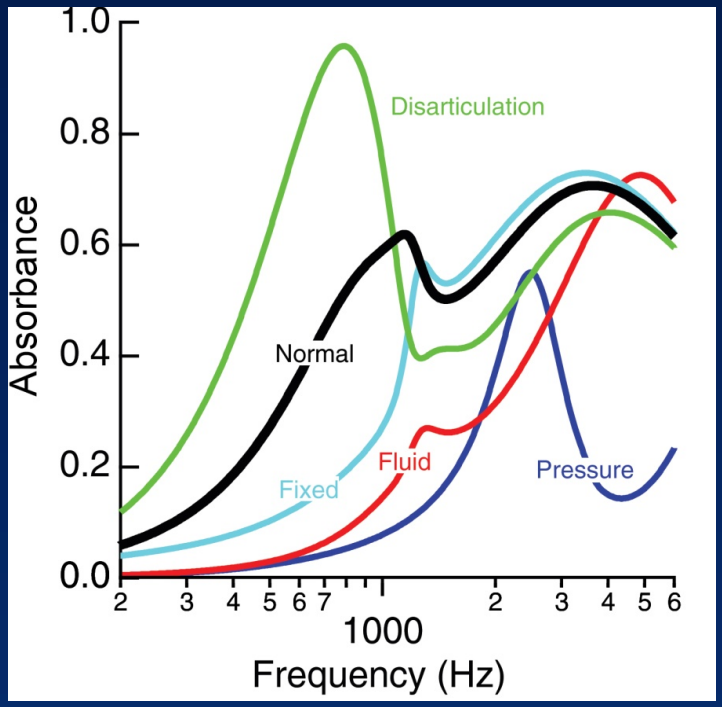
(Figure courtesy of Bue Kristensen, Interacoustics, 2013)



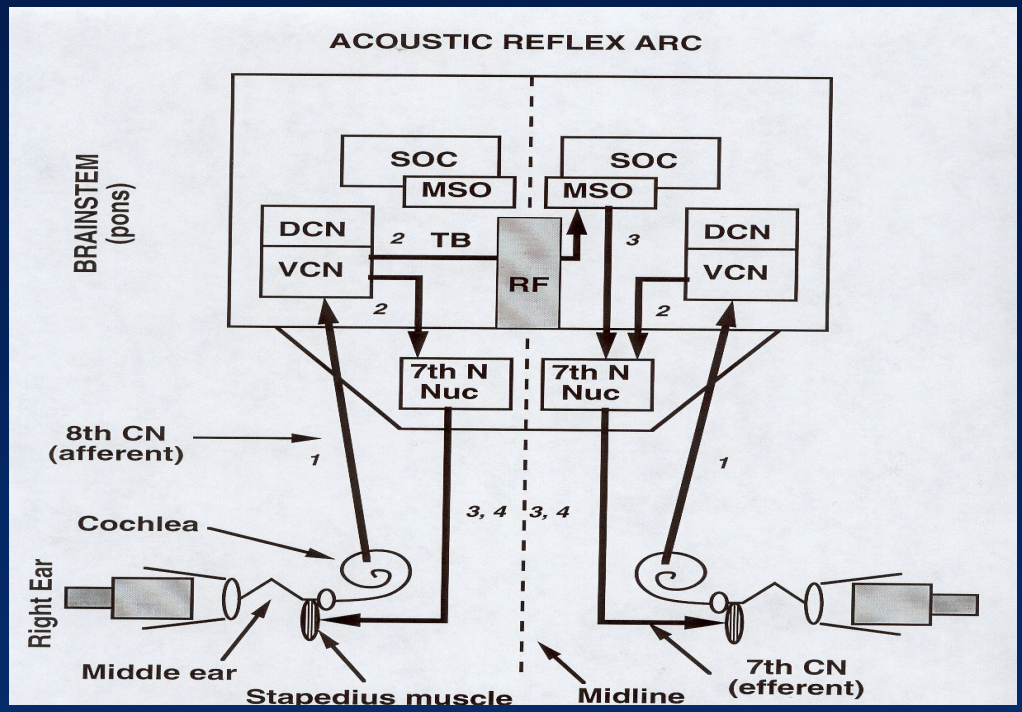
$$\text{Energy Absorbance} = \frac{\text{Absorbed Power}}{\text{Incident Power}} = 0$$

$$= 1 - \text{Energy Reflectance}$$

Wideband Reflectance/Absorbance (Voss et al. Ear & Hearing, 2008)



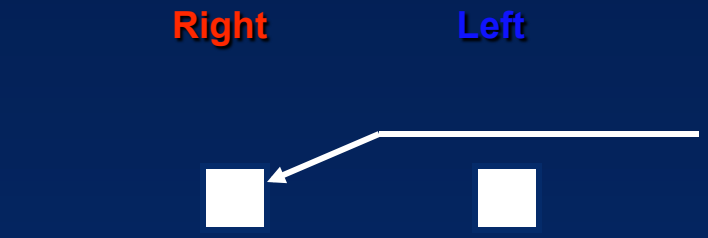
Measurement of Acoustic Stapedial Reflex with 1000 Hz Probe: When Feasible for Broad Band Noise Stimulus



Plotting the Results of Acoustic Reflex Measurements: Objective Differentiation Among Types of Auditory Dysfunction

Abnormal Acoustic Reflex

Diagnosis Based on Pattern of Abnormalities
Middle ear, Cochlea, 8th CN, Brainstem, 7th CN



Contralateral (Crossed) Sound Right Probe Left

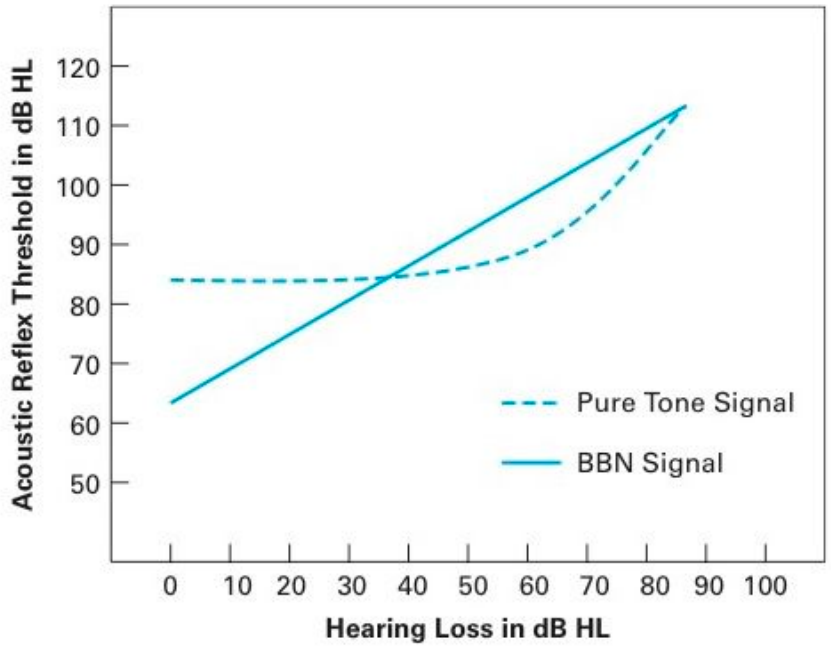
Contralateral (Crossed) Sound Left Probe Right

Ipsilateral (Uncrossed) Sound Right Probe Right

Ipsilateral (Uncrossed) Sound Left Probe Left

Differentiation of Normal Hearing Sensitivity versus Hearing Loss Using BBN Stimulus for Acoustic Reflex Threshold

(Figure from Hall JW III. Introduction to Audiology Today. Boston: Pearson, 2014)



Contribution of Aural Immittance Measurement to Diagnosis and Intervention of Infant Hearing Loss

- Diagnostic information
  - Differentiation of middle ear versus sensory auditory dysfunction
  - Objective confirmation of sensory hearing loss (acoustic reflexes)
  - Objective evidence of retrocochlear auditory dysfunction and of auditory neuropathy spectrum disorder (acoustic reflexes)
  - Objective evidence of central auditory nervous system dysfunction (acoustic reflexes)
- Impact on Intervention Outcome
  - Prompt medical management of middle ear disorder
  - Cost effective and lower risk decisions regarding further diagnostic test procedures (e.g., ABR under anesthesia)
  - Timely referral for multi-disciplinary referrals for prompt and accurate diagnosis of ANSD

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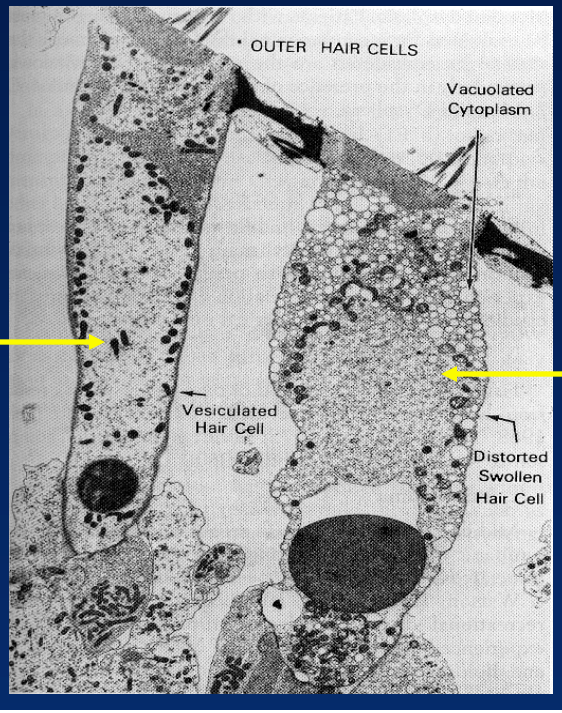
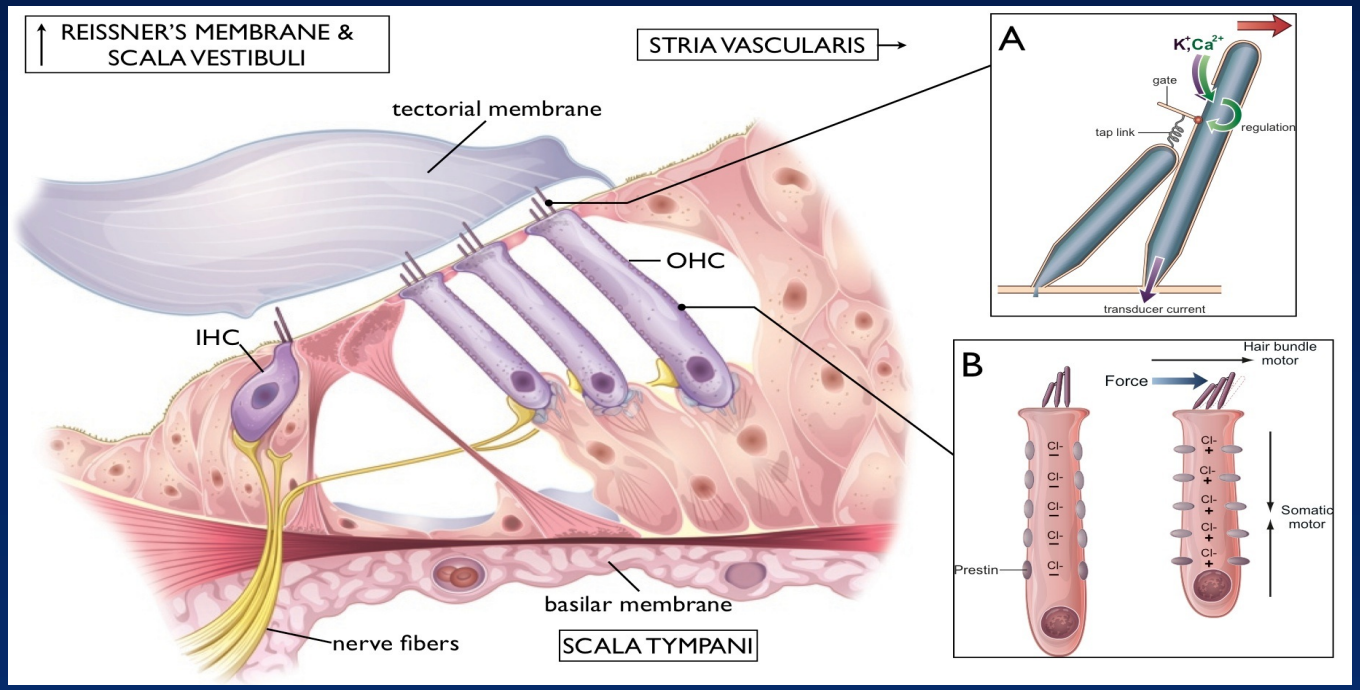
OAEs in Early Detection of Outer Hair Cell Dysfunction

Normal OHC (OAEs)



Abnormal OHC (OAEs)

OAEs Originate in the Cochlea …
But That's Not the Entire Story



Auditory Anatomy Involved in the Generation of OAEs

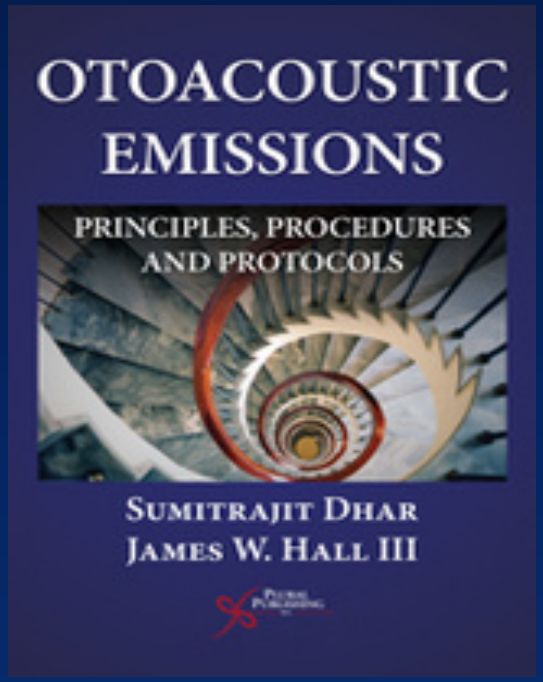
- Outer hair cell motility
  - Prestin motor protein
- Stereocilia
  - Motion
  - Stiffness
- Tectorial membrane
- Basilar membrane mechanics
  - Dynamic interaction with outer hair cells
- Stria vascularis
- Middle ear (inward and outward propogation)
- External ear canal
  - Stimulus presentation
  - OAE detection

Contribution of Otoacoustic Emissions to Diagnosis and Intervention of Infant Hearing Loss

- Diagnostic information
  - Confirmation of outer hair cell integrity versus dysfunction
  - Early detection of cochlear (outer hair cell) dysfunction, e.g., in
    - ✓ Ototoxicity
  - Differentiation of outer versus inner hair cell dysfunction
  - Objective confirmation of sensory hearing loss
  - Identification and diagnosis of auditory neuropathy spectrum disorder
- Impact on Intervention Outcome
  - Earlier and more accurate diagnosis leads to more effective intervention
  - Cost effective and lower risk decisions regarding further diagnostic test procedures (e.g., ABR under anesthesia)
  - Timely referral for multi-disciplinary referrals for prompt and accurate diagnosis of ANSD

Otoacoustic Emissions:
Principles, Procedures, and Protocols

Plural Publishing
(www.pluralpublishing.com)
150 pages, Softcover, 5 x 7.5"
ISBN10: 1-50756-342-0
ISBN13: 978-1-59756-342-0
$45.00



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    - ✓Click
    - ✓Tone burst
    - ✓Chirp
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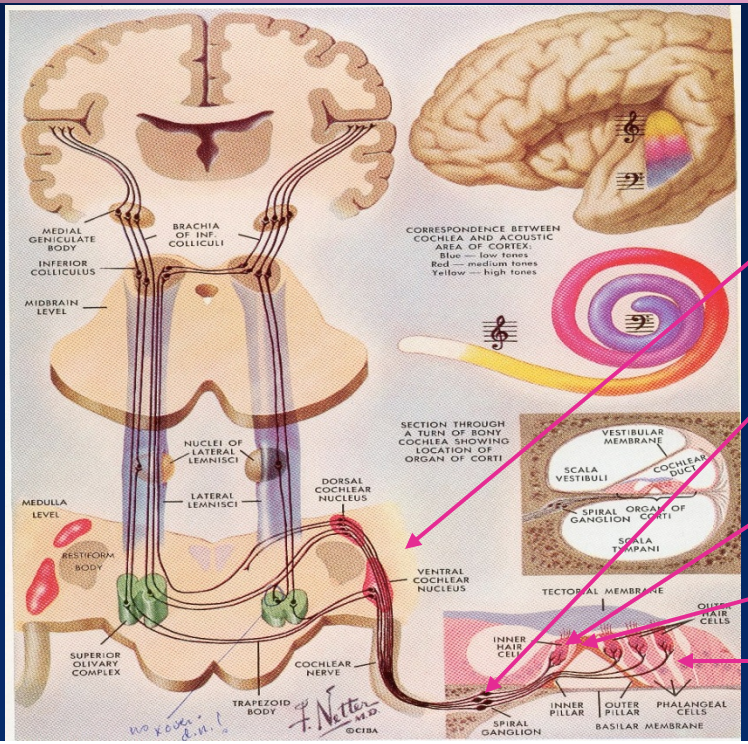
Sensory and Neural Asssessment



Brainstem
(ABR, ASSR, ARs)

Spiral ganglion cells
(ABR, ECochG)

IHC - 8th CN Synapse
(ABR)

Inner hair cells
(ECochG, ABR, ASSR, ARs)

Outer hair cells
(OAE, ECochG, ARs)

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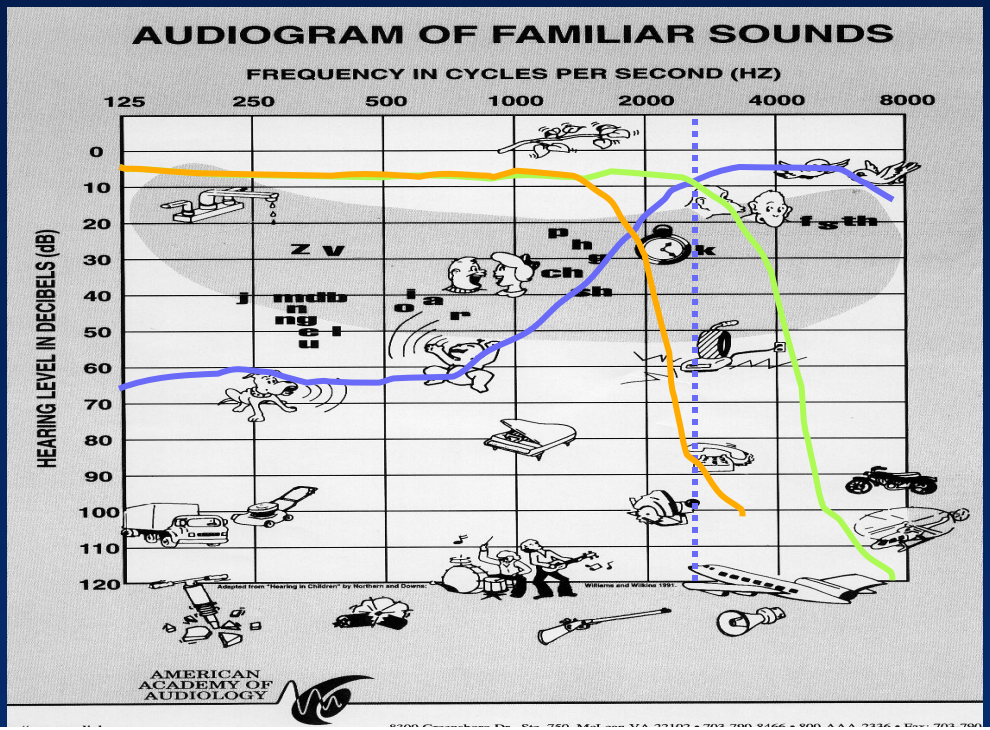
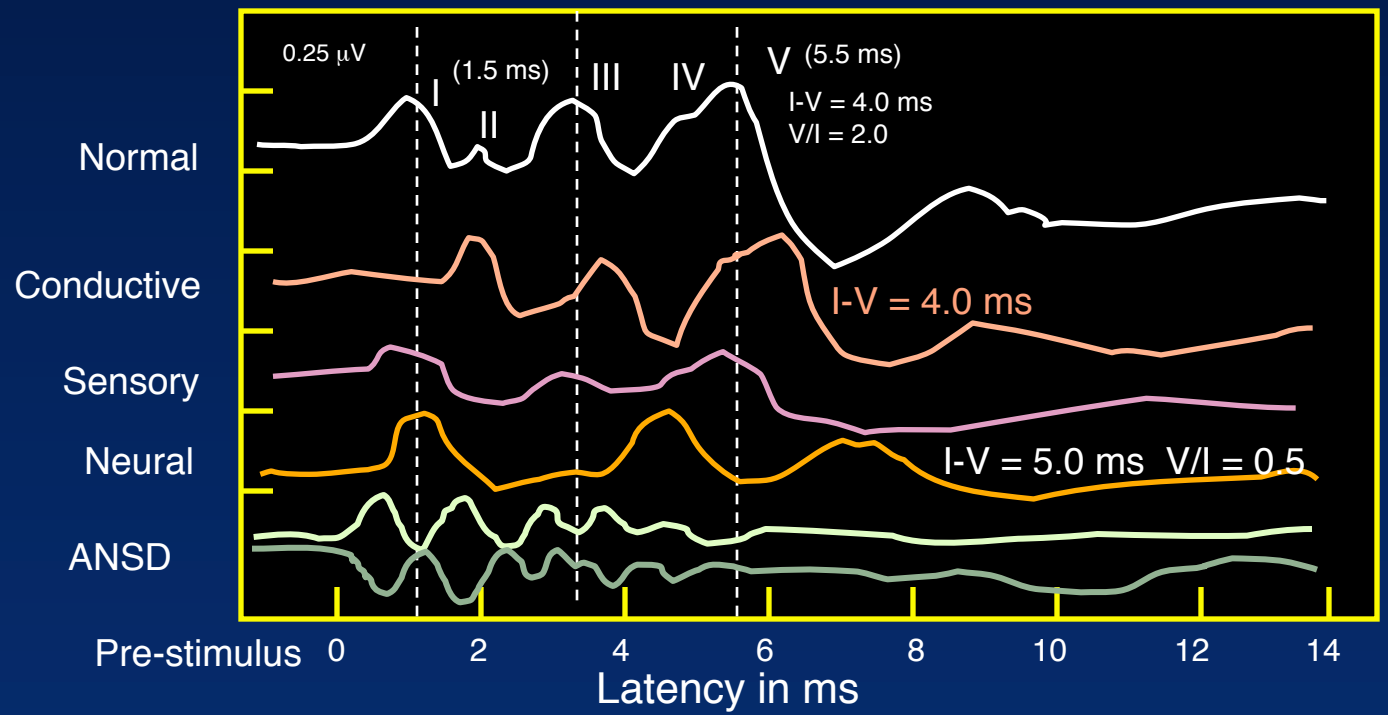
Frequency-Specific Estimation of Auditory Thresholds: An Essential Step in Diagnosis of Infant Hearing



Normal click ABR

Abnormal or no click ABR

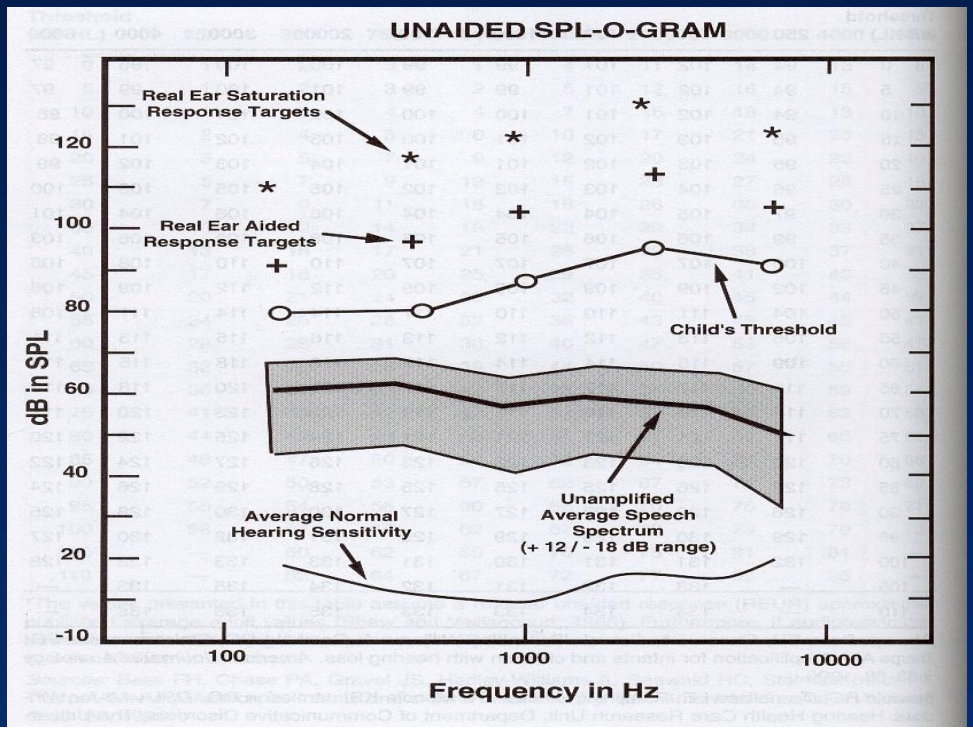
Diagnostic Value of the Click-Evoked ABR: Differentiation Among Types of Auditory Dysfunction *(Waveforms for 85 dB nHL Click Stimulus)*



Electrophysiological Estimation of the Audiogram in Infants and Young Children with Clicks, Tone Bursts, and Chirps

- Why it's a good strategy to begin the ABR assessment with click stimulation
  - Waveform analysis permits differentiation among types of hearing loss
  - Waveform analysis indicates test ear (presence of wave I)
  - Auditory neuropathy spectrum disorder (ANSD) can be ruled out or identified
  - Findings help to determine next steps in the assessment, e.g.,
    - ✓ Bone conduction ABR or tympanometry?
    - ✓ ASSR?
  - Only requires a few minutes of test time
  - Recommended by the 2007 Joint Committee on Infant Hearing

Estimation of Frequency-Specific Auditory Thresholds with Tone Burst ABRs: Initial Data Points for Hearing Aid Fitting



Frequency-Specific ABR Test Protocol: Stimulus Parameters and Research Needs

Parameter	Selection	Rationale/Research Needs
Transducer	ER-3A inserts	Numerous infant advantages Accurate in-ear SPL verification
Type	Tone bursts	Available on all systems Clinical trials of chirp stimuli
Ramping (window)	Blackman	Less spectral splatter
Frequencies	1, .5, 4, 2 K Hz	Sequence varies clinically High frequency option (> 4000Hz) Normative data for infants
Duration	2-0-2 cycles	Abrupt onset frequencies Equivalent intensity for each frequency 0 plateau < spectral splatter

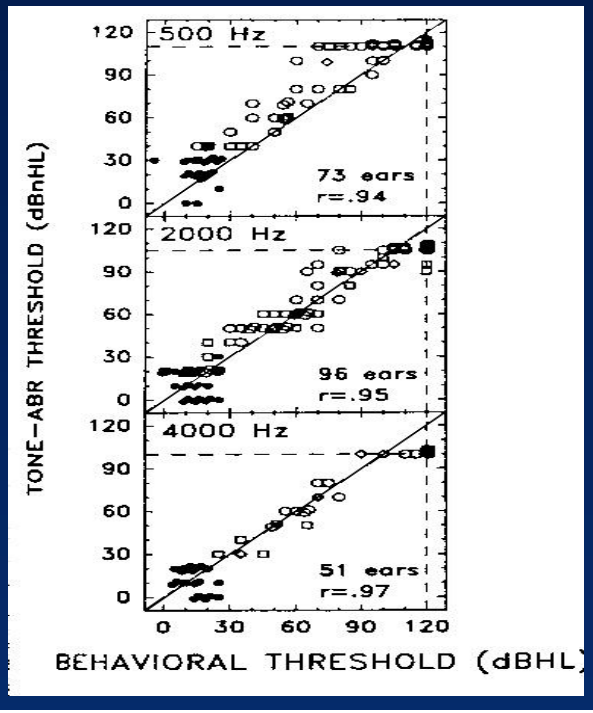
Frequency-Specific ABR Test Protocol: Acquisition Parameters and Research Needs

Parameter	Selection	Rationale/Research Needs
Artifact reject	On	Minimize muscle artifact Weighted averaging of all data
Analysis time	20 ms	Encompass delayed wave Vs and SN10 after wave V
Sweeps	1000 or 2000	Produce adequate SNR Automated detection of adequate response based on SNR
Reliability	2 or 3 runs	"If it doesn't replicate, you must investigate!"

Frequency-Specific ABR Test Protocol: Acquisition Parameters and Research Needs

Parameter	Selection	Rationale/Research Needs
Electrode type	Disc & ear clip or disposable	Amplification of response at or near the electrode
Electrode location	Fz - Ai Fpz ground	Optimal infant response Good for BC stimulus Permits ipsi/contra meas't
Filter settings	30 - 3000 Hz No notch filter	Encompass infant spectrum
Artifact reject	On	Minimize muscle artifact Weighted averaging of all data

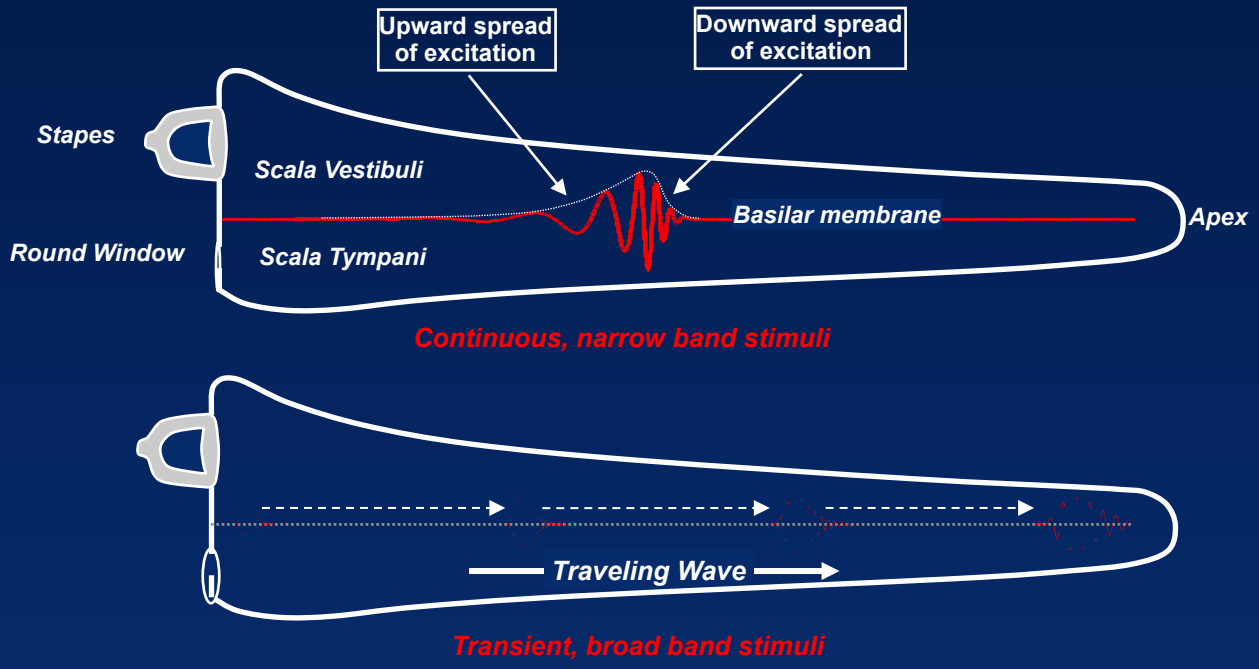
FREQUENCY-SPECIFIC AUDITORY BRAINSTEM RESPONSE (ABR): Relation to Audiogram (Oates & Stapells, 1998)



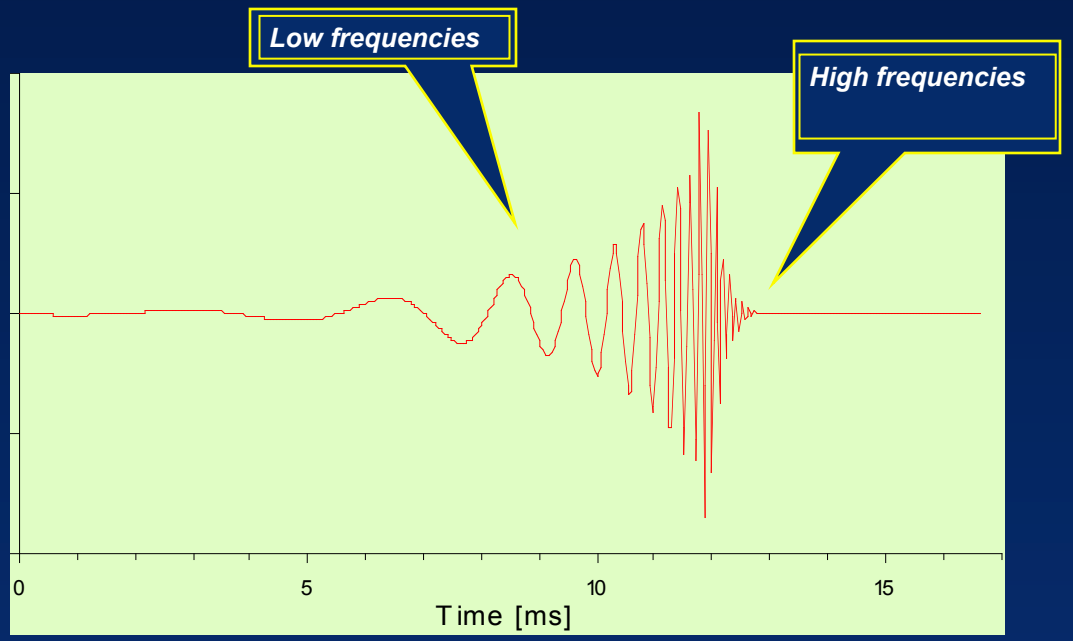
Advantages of CE-Chirp Stimulation of the Auditory Brainstem Response (ABR)

- What is a chirp?
- What are the advantages of chirp stimuli?
- Are intensity and frequency characteristics comparable for conventional versus chirp stimuli?
- Do chirp stimuli really result in larger ABR amplitude?
- Examples of chirp-evoked ABR waveforms
- Summary of the advantages of chirp stimuli and a remaining question about clinical application of chirp-evoked ABR

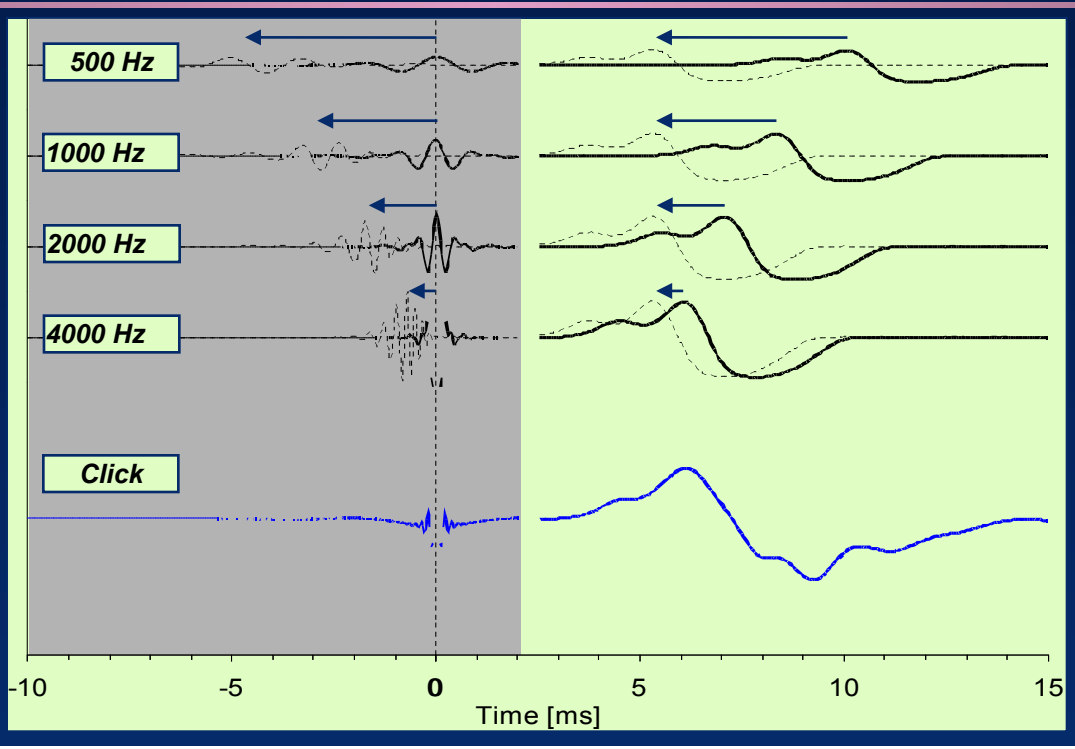
Cochlear Excitation Patterns for Click versus Narrow Band Stimulation



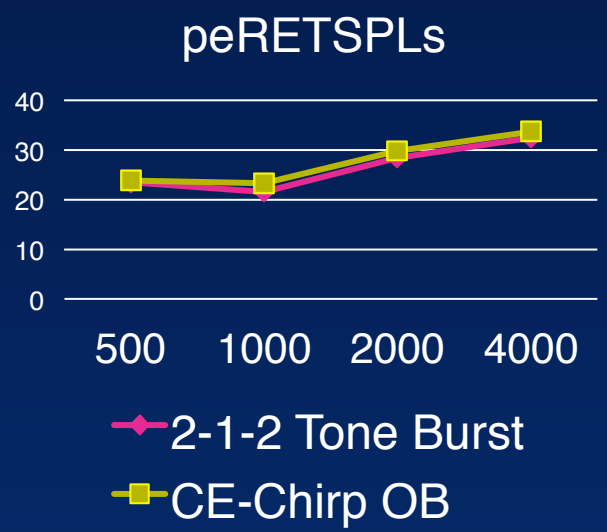
Chirp Temporal Waveform



Temporal Compensation via Input Compensation (Courtesy of Claus Elberling)



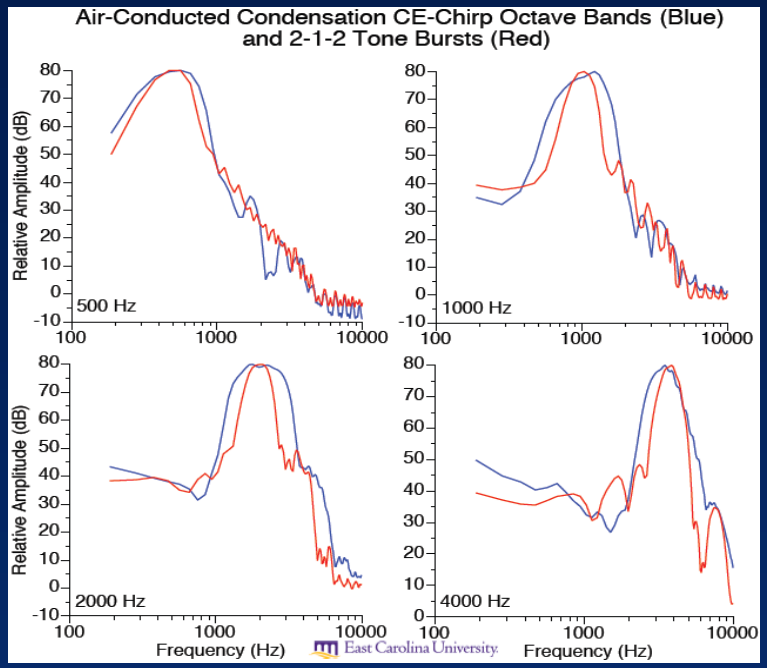
peRETSPLs:
CE-Chirp Octave Bands vs. Tone Bursts



- ISO 389-6: 2-1-2 Tone Burst peRETSPLs (blue = tone bursts)
- 3A Insert Earphones using 711 ear simulator
- Range of 0.4 to 1.8 dB difference

Reference: Gotsche-Rasmussen, Poulsen, Elberling, Reference Hearing Threshold Levels for Chirp Signals Delivered by an ER-3A Earphone, International Journal of Audiology, 2012, Early Online: 1-6

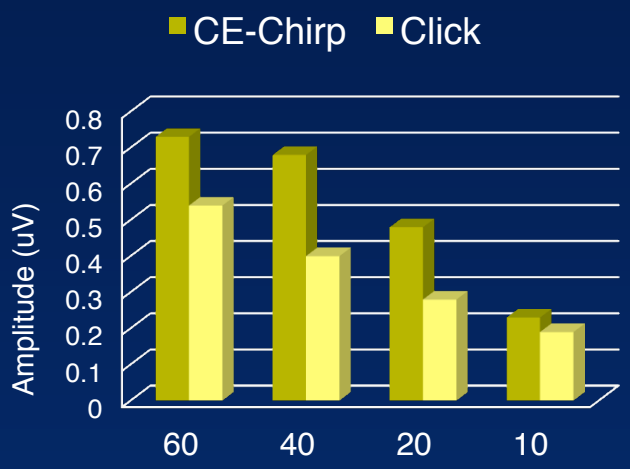
Acoustic Spectrum:
CE-Chirp Octave Bands vs. Tone Bursts



Courtesy of East Carolina University

Adults:
CE-Chirp Amplitudes

Amplitude Comparison

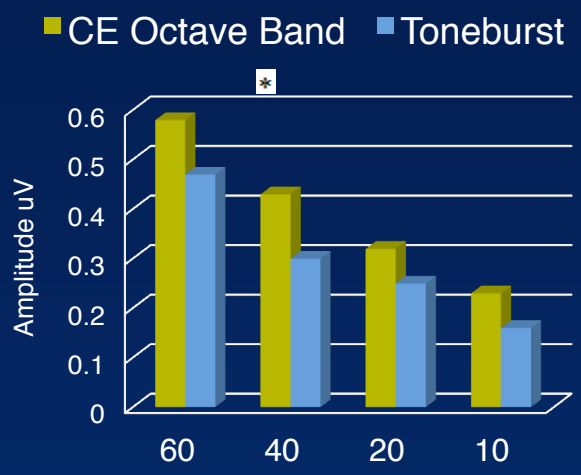


- **Wave V amplitudes were significantly greater at 60, 40, 20 dB nHL**
- **Greater amplitudes are consistent with previously published research**

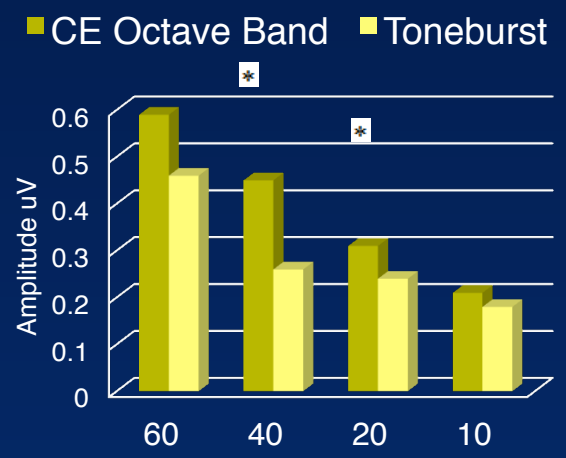
Stangl S, Rentmeester L, Hood LJ. (2013). Auditory brainstem responses to clicks, chirps, tonebursts, and octave-band chirps. Poster presented at the 2013 Meeting of the American Auditory Society, Scottsdale, Arizona.

Adults:
CE-Chirp Octave Bands

4000 Hz



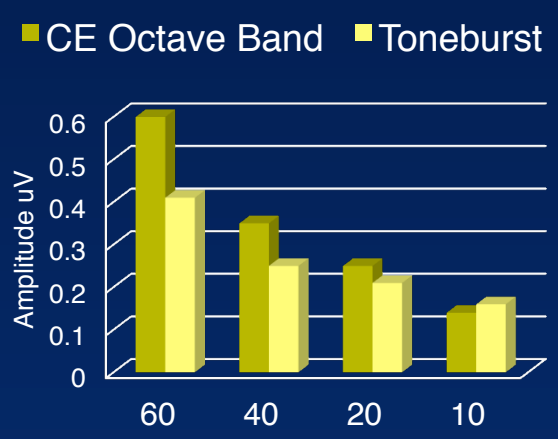
2000 Hz



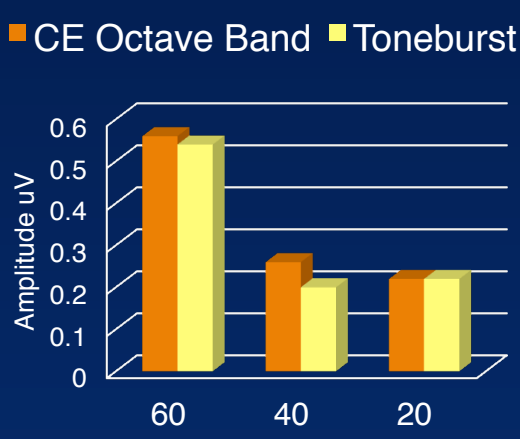
Stangl S, Rentmeester L, Hood LJ. (2013). Auditory brainstem responses to clicks, chirps, tonebursts, and octave-band chirps. Poster presented at the 2013 Meeting of the American Auditory Society, Scottsdale, Arizona.

Adults: CE-Chirp Octave Bands

1000 Hz



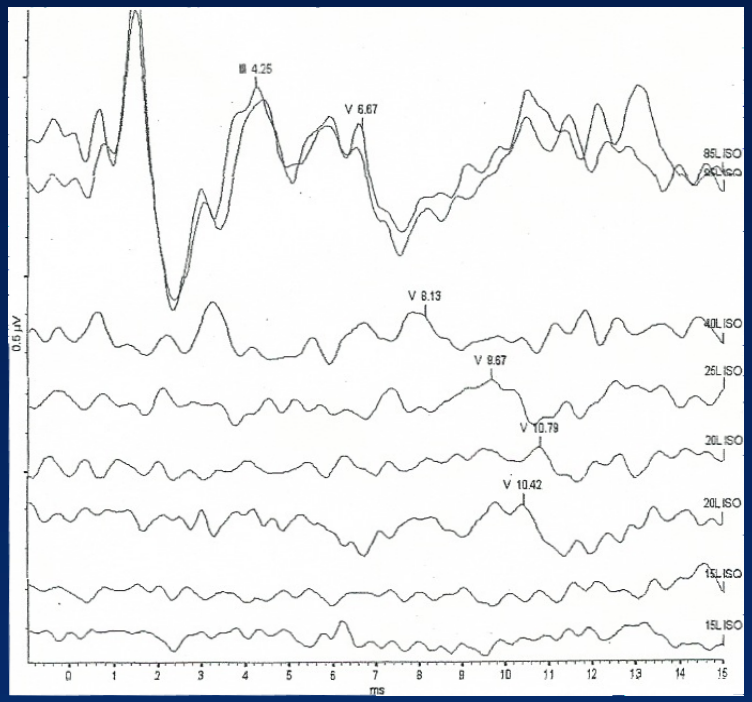
500 Hz



Stangl S, Rentmeester L, Hood LJ. (2013). Auditory brainstem responses to clicks, chirps, tonebursts, and octave-band chirps. Poster presented at the 2013 Meeting of the American Auditory Society, Scottsdale, Arizona.

Conventional Click versus CE Chirp Evoked ABR

(1 year 4 month old boy with speech & language delay who failed hearing screening in nursery. Parents do not speak English)



85 dB nHL Click, rarefaction, 21.1/sec
I = 1.46 ms
V = 6.67 ms
I-V = 5.21 ms

45 dB nHL Click

25 dB nHL Click

20 dB nHL Click

20 dB nHL CE Chirp

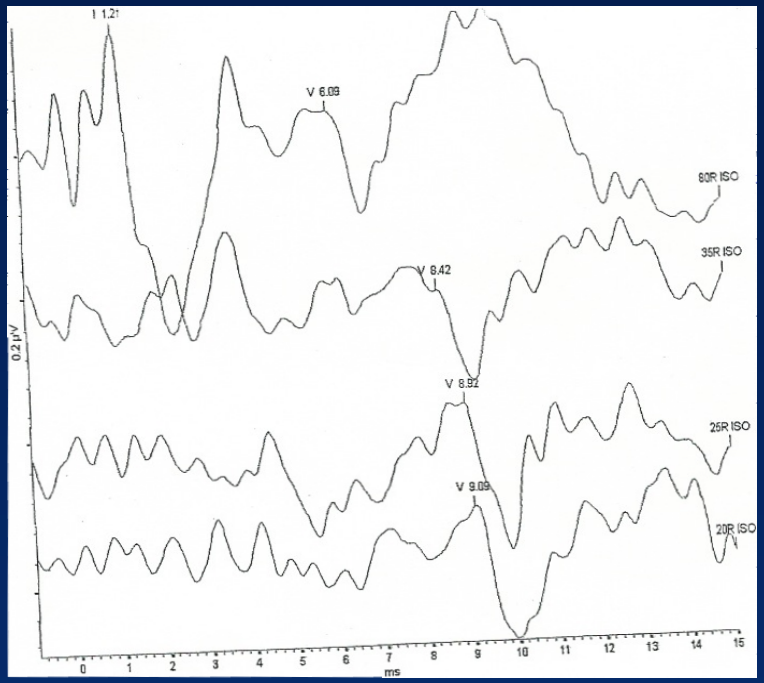
15 dB nHL Click

15 dB nHL CE Chirp

2000 Hz Chirp Evoked ABR

Stimulus rate = 37.7/sec

Total sweeps = 2318 ; Total test time = 61 seconds



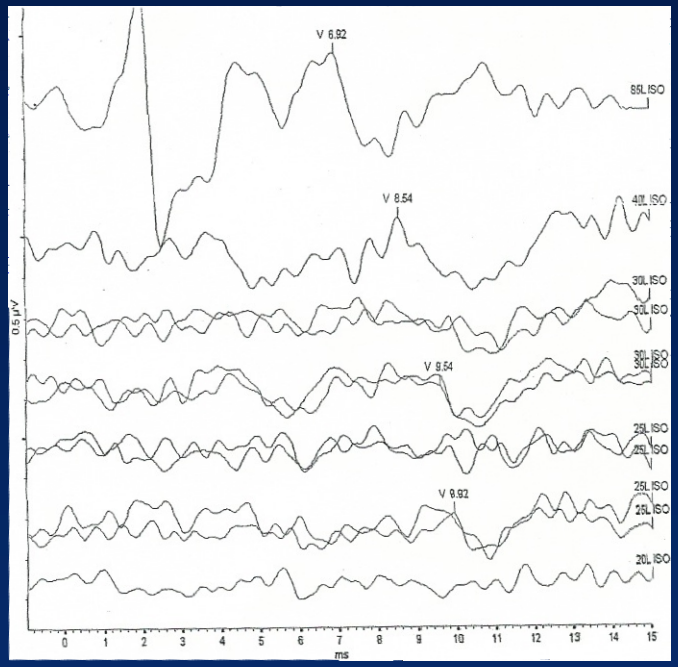
80 dB nHL
722 sweeps

35 dB nHL
570 sweeps

25 dB nHL
456 sweeps

20 dB nHL
570 sweeps

4000 Hz Conventional versus Chirp Evoked ABR



Left Ear
85 dB nHL
Tone Burst

40 dB nHL
Tone Burst

30 dB nHL
Tone Burst

30 dB nHL, Chirp Tone Burst

25 dB nHL, Tone Burst

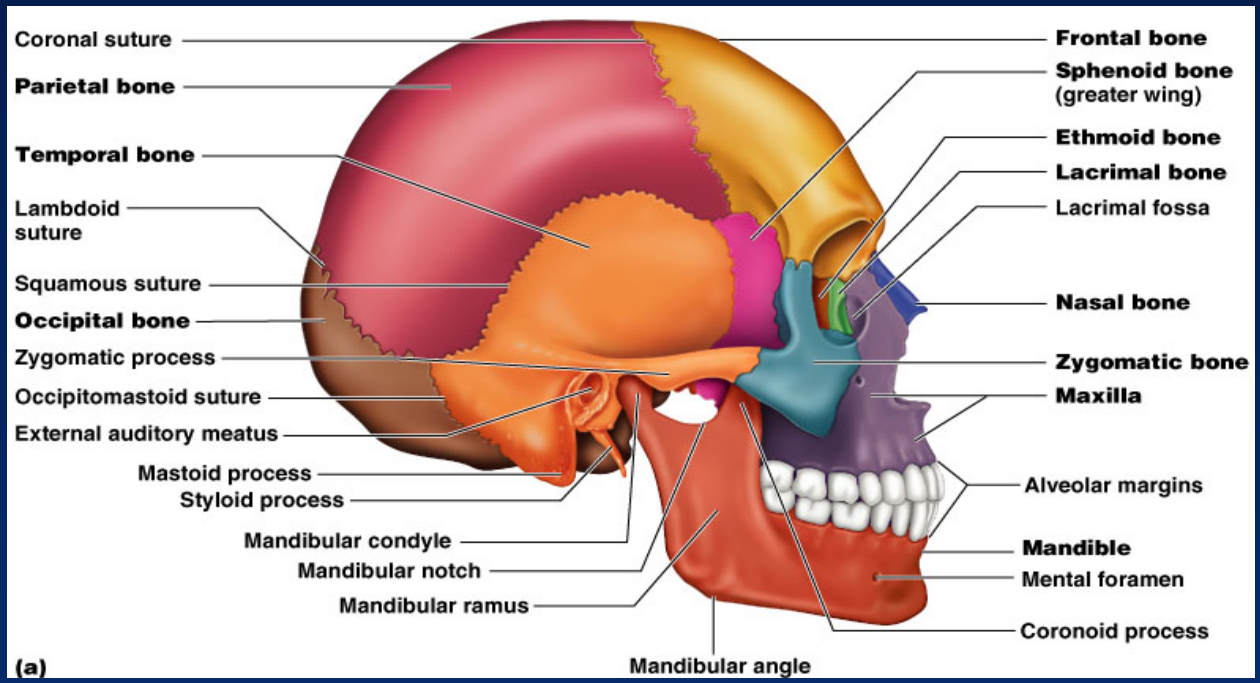
25 dB nHL, Chirp Tone Burst

15 dB nHL, Chirp Tone Burst

Advantages of CE-Chirp Stimulation of the Auditory Brainstem Response (ABR): Advantages of Chirp Stimulation

- ABR amplitude is larger for chirp stimulation
- Larger amplitude contributes to:
  - More confident identification of wave V
  - Shorter test time is needed to identify wave V
  - Reduced test time for each stimulus frequency permits more complete estimation of auditory threshold in speech frequency region
  - More accurate thresholds are sometimes possible with chirp stimulation
- More data are needed on the clinical measurement of ABR with high intensity (> 60 dB) chirp stimuli

Ear Specific Bone Conduction ABR Assessment: Clinically Feasible and Essential

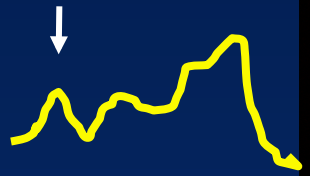


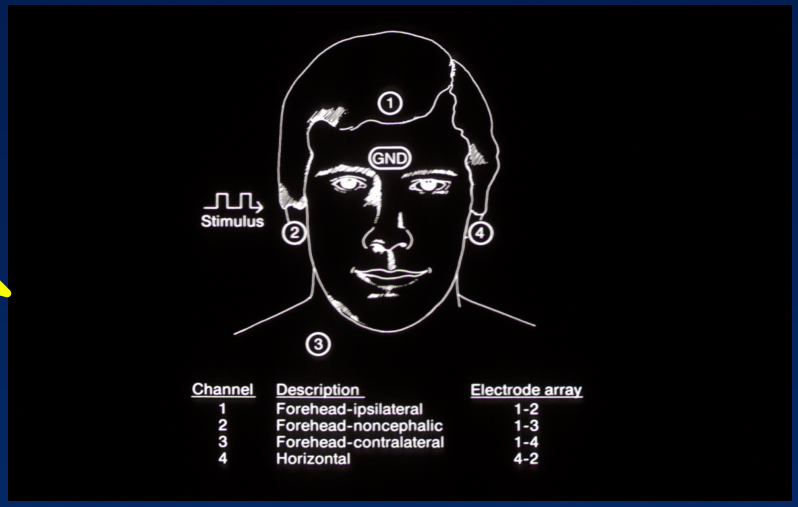
Maturation of Bone Conduction ABR: Increased Inter-Aural Attention in Infants

- Yang EY, Rupert AL & Moushegian G (1987). A developmental study of bone conduction auditory brainstem responses in infants. Ear & Hearing, 8, 244-251
- Small SA & Stapells DR (2008). Normal ipsilateral/contralateral asymmetries in infant multiple auditory steady-state responses to air- and bone conduction stimulation. Ear & Hearing, 29, 185-198
- Conclusion: Bone conduction stimulation up to 30 dB nHL in infants will activate only the ipsilateral cochlea

Two-Channel Bone Conduction ABR Recording: Applying ECochG Principles to Verify the Test Ear

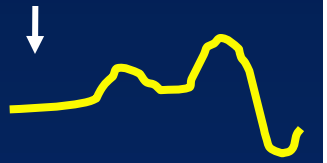
Ipsi Channel
Wave I





Channel	Description	Electrode array
1	Forehead-ipsilateral	1-2
2	Forehead-noncephalic	1-3
3	Forehead-contralateral	1-4
4	Horizontal	4-2

Contra Channel
No Wave I



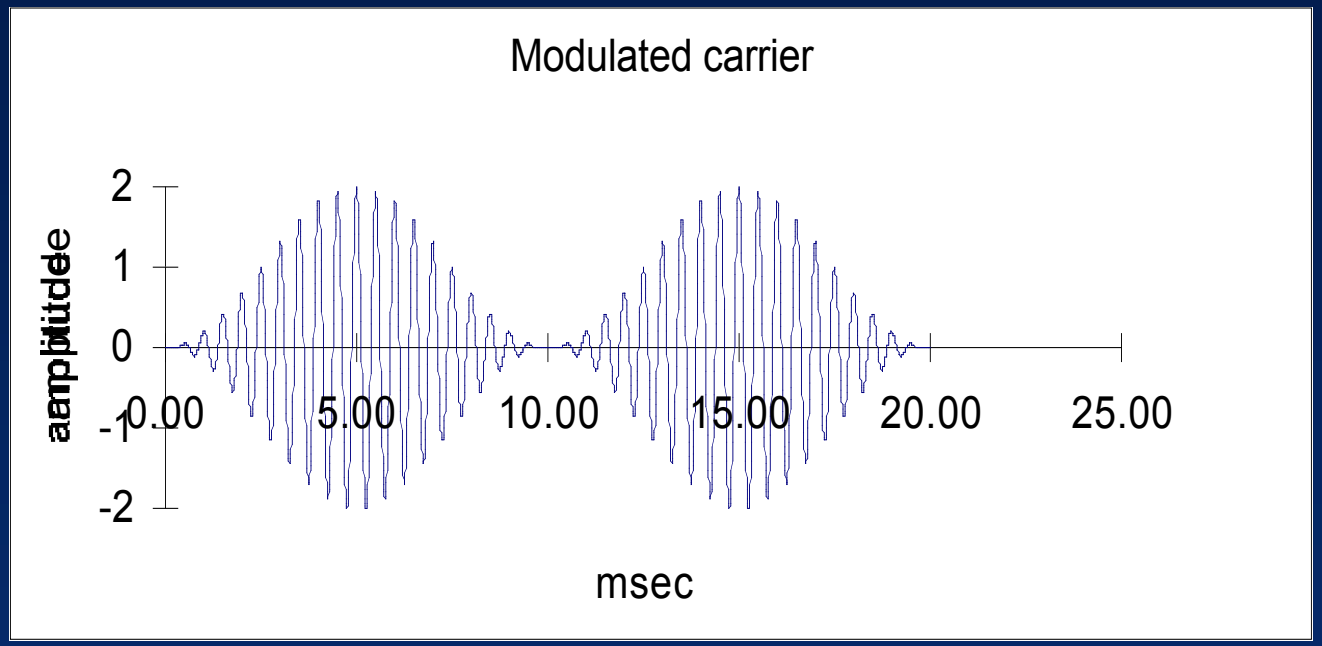
Clinical Measurement and Applications of Bone Conduction ABR: Click or Tone Burst Bone Conduction Stimulation

- Rationale for click only
  - Air conduction tone burst information is most useful
  - Test time is unacceptably lengthy with addition of tone burst bone conduction recordings
  - Confident identification of ABR is more likely with click versus tone burst stimulation
  - Provides information needed for management decisions
- Rationale for tone burst stimulation
  - Consistent with protocol for behavioral audiometry
  - Click stimulation may underestimate conductive component
    - ✓ Only estimates air-bone gap in the high frequency region
    - ✓ Conductive hearing loss is usually greatest in low frequency region

Australian Professional Practice Standards (July 2013)
Advanced Audiological Assessment *Clinical Processes*

- Detailed case history
- Otoscopy
- Audiometry (behavioral AC and BC)
- Speech perception assessment (formal or informal)
- Tympanometry (226 Hz and high frequency)
- Acoustic reflexes (BBN, multi-frequency, ipsi, contra)
- Otoacoustic emissions
- **Auditory evoked potentials** (ABR, **ASSR**, ECochG, other)
- Interpretation of tests performed and test battery
- Feedback, counseling and health promotion
- Recommendations for future management

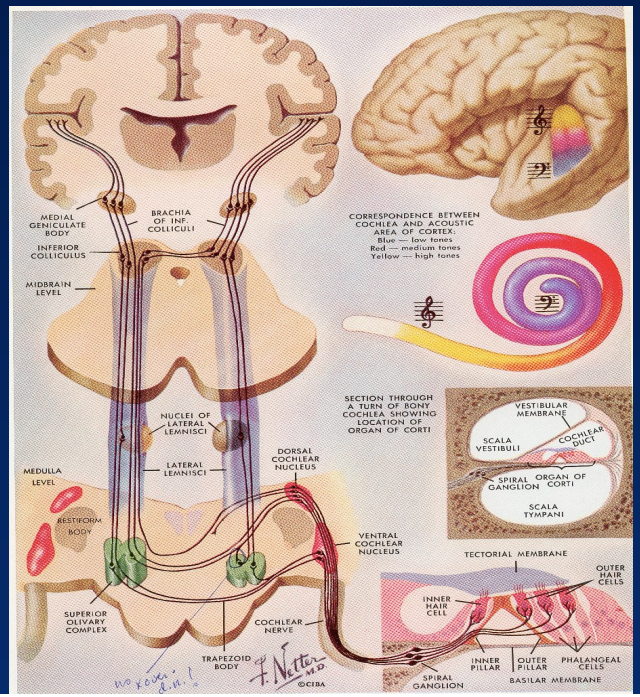
Auditory Steady State Response (ASSR): 2000 Hz tone modulated at rate of 100 Hz



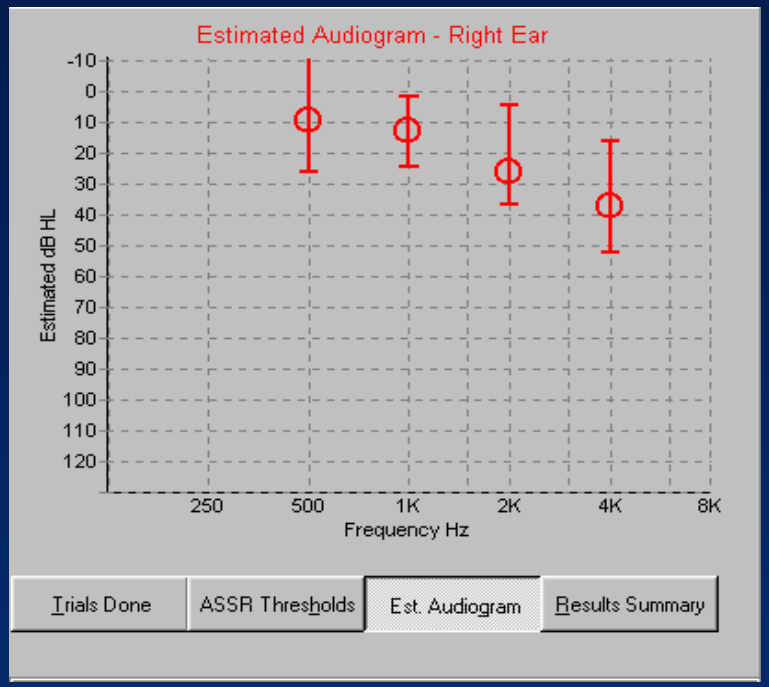
Anatomy & Physiology of ASSR: Generators (Kuwada et al, 2002

Slower modulation rates (< 80 Hz) = Cortical regions

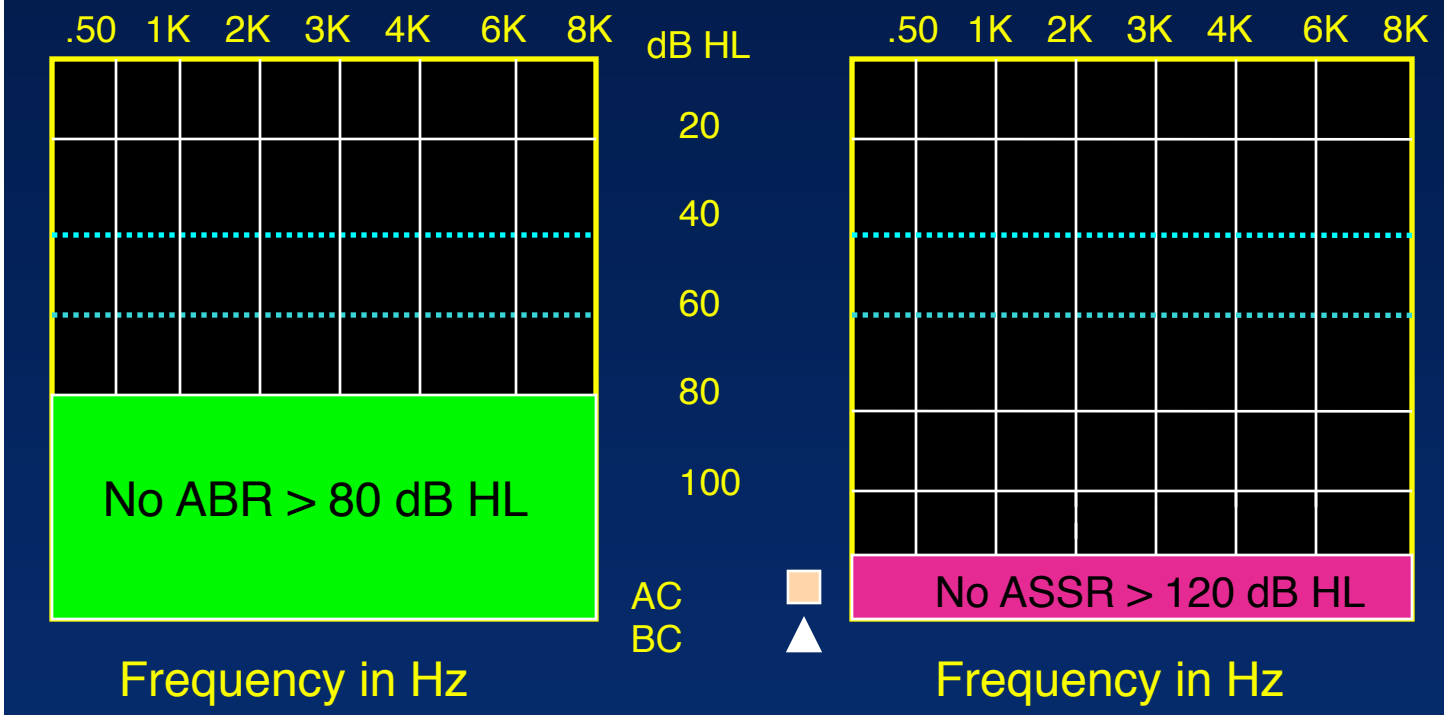
Faster modulation rates (> 80 Hz) = Brainstem



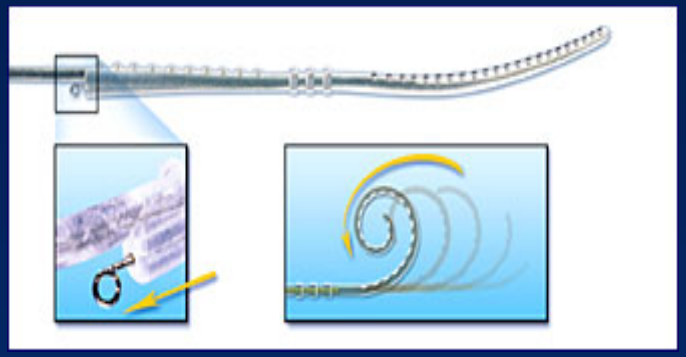
Estimating the Audiogram with ASSR



Limitation of Tone Burst ABR and Advantage of ASSR in Severe-to-Profound Hearing Loss



ASSR Contributes to Timely Management of Infant Hearing Loss: Determining the Need for Cochlear Implants and Confirmation of 8th Nerve Integrity in ANSD



Evidence-Based Efficient and Effective Identification and Diagnosis of Infant Hearing Loss

- Early diagnosis and intervention improves communication
- Accurate assessment of infant hearing is standard of care
- Efficient identification of (screening for) hearing loss
- The cross-check principle still rules
- An objective auditory test battery
  - Acoustic immittance measures
  - OAEs
  - ABR
  - ASSR
  - ECochG
- Pulling it all together

Australian Professional Practice Standards (July 2013)
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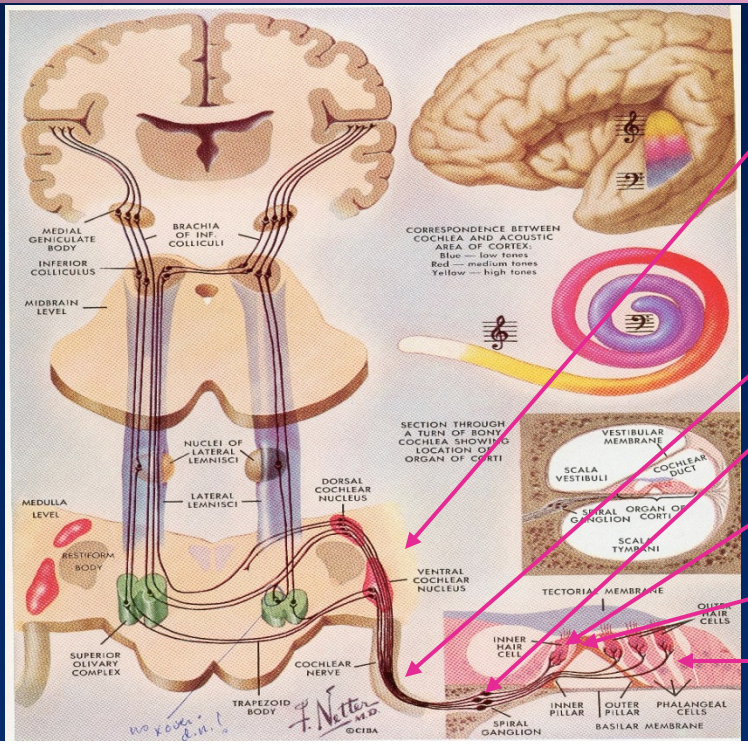
ECochG in the Diagnosis and Management of Auditory Neuropathy Spectrum Disorder (ANSD)



Cerebello-pontine angle (CPA)

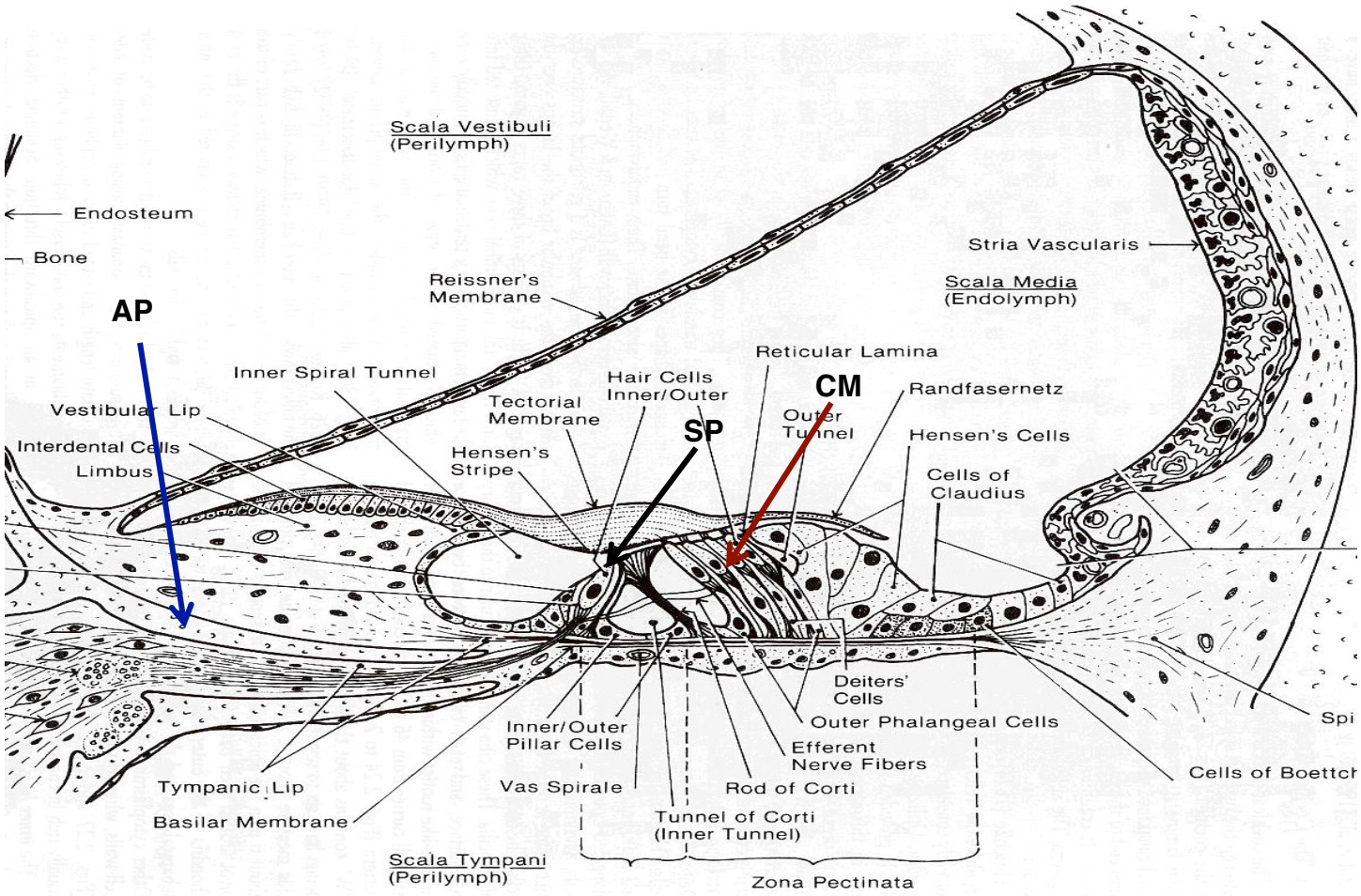
Internal Auditory Canal (Auditory Nerve)

Spiral ganglion cells

IHC - 8th CN Synapse (glutamate)

Inner hair cells

Outer hair cells


Scala Vestibuli
(Perilymph)
Endosteum
Bone
AP
Reissner's
Membrane
Stria Vascularis
Scala Media
(Endolymph)
Reticular Lamina
Inner Spiral Tunnel
Hair Cells
Inner/Outer
CM
Randfasernetz
Vestibular Lip
Tectorial
Membrane
SP
Outer
Tunnel
Hensen's Cells
Interdental Cells
Limbus
Hensen's
Stripe
Cells of
Claudius
Deiters'
Cells
Outer Phalangeal Cells
Inner/Outer
Pillar Cells
Efferent
Nerve Fibers
Cells of Boettch
Tympanic Lip
Vas Spirale
Rod of Corti
Basilar Membrane
Tunnel of Corti
(Inner Tunnel)
Scala Tympani
(Perilymph)
Zona Pectinata

Electrocochleography (ECochG) Test Protocol (1)

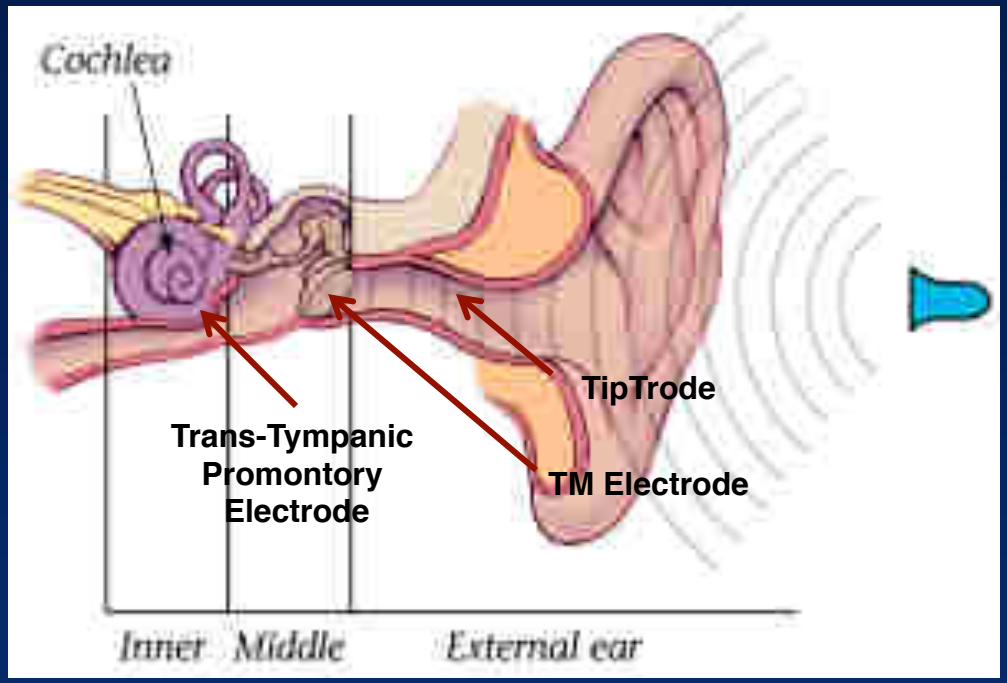
Stimulus Parameters

Type	Clicks
Duration	0.1 ms
Rate	7.1/sec or slower as necessary
Polarity	Alternating (for SP and AP)
	Rarefaction and condensation (for CM)
Intensity	Maximum or lower
Transducer	Insert
Masking	Never needed (response is ipsilateral)

Electrocochleography (ECochG) Test Protocol (2)

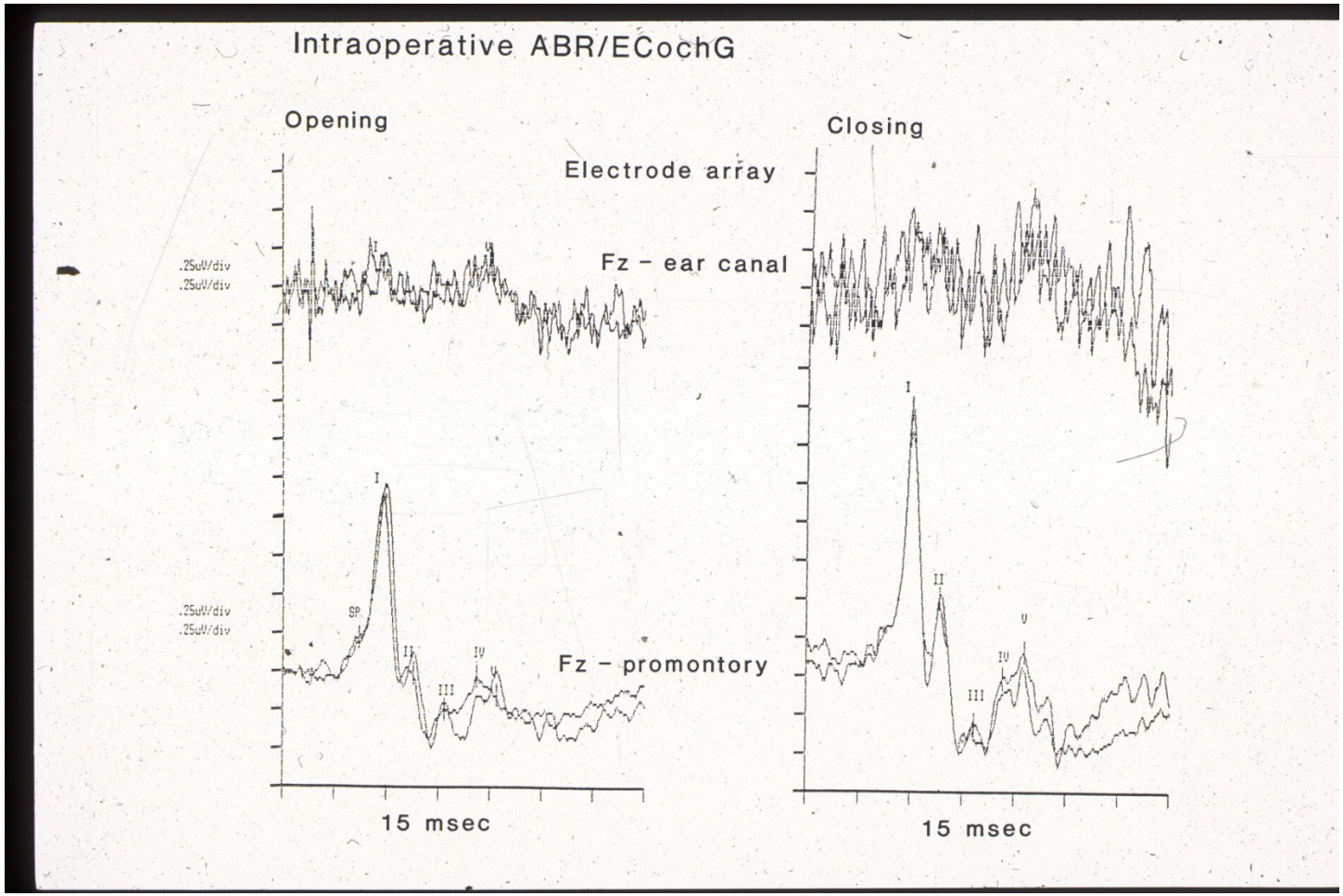
Acquisition Parameters	
Amplification	75,000 or less
Analysis time	5 or 10 ms
Sweeps	500 or less
Filters	10 to 1500 Hz
Notch filter	Never
Electrodes	
Option 1	Fz to trans-tympanic needle
Option 2	Fz to tympanic membrane
Option 3	Fz to Tiptrode

ECochG Electrode Options:
The Closer to the Cochlea, the Better



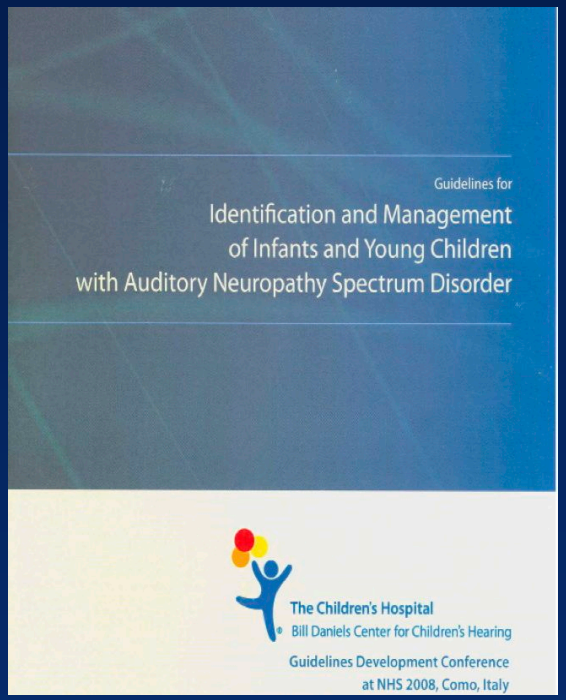

INTELLIGENT HEARING
Non-Sterile/Single Use Only
Lilly TM-Wick Electrode
Designed and developed by Dr. David J. Lilly - Used for ECochG recordings for over 20 years.

Sub-Dermal Needle Electrode for Trans-Tympanic Promontory ECochG Recording


Intraoperative ABR/ECochG
Opening
Closing
Electrode array
Fz – ear canal
.25uV/div
.25uV/div
.25uV/div
.25uV/div
Fz – promontory
SP
I
II
III
IV
V
15 msec
15 msec

Identification, Diagnosis, and Management of Auditory Neuropathy Spectrum Disorder (ANSD)

- 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 of Auditory Neuropathy Spectrum Disorder: Minimal Test Battery (2008 ANSD Guidelines)

- Tests of cochlear hair cell function
  - Otoacoustic emissions (OAEs)
  - Cochlear microphonic (ECochG and ABR)
    - ✓CM may be present with absent OAEs (e.g. middle ear disorder)
- Tests of auditory nerve function
  - ECochG
  - ABR for highand moderate intensity click stimulation with separate averages for:
    - ✓Rarefaction stimulus polarity
    - ✓Condensation stimulus polarity
- Additional tests are supplemental
  - Acoustic reflex measurement (generally absent in ANSD)
  - Suppression of otoacoustic emissions (no suppression in ANSD)

Diagnosis of Auditory Neuropathy Spectrum Disorder (2008 Guidelines)

- Diagnostic audiologic 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

Electrophysiology Procedures in the Diagnosis of ANSD: Refining Site of Dysfunction

- 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
  - ECochG recorded with
    - ✓ Non-inverting ("active") window "golf club" electrode near round electrode
    - ✓ 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 + dentritic potential) but no AP = poor or absent EABR and poor CI benefit

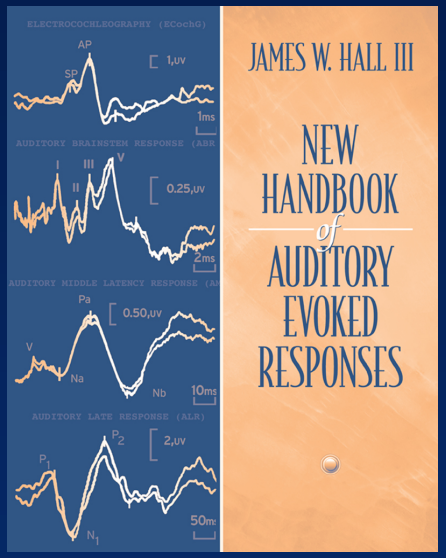
Electrophysiology Assessment of Infant Hearing: An Evidence-Based Protocol

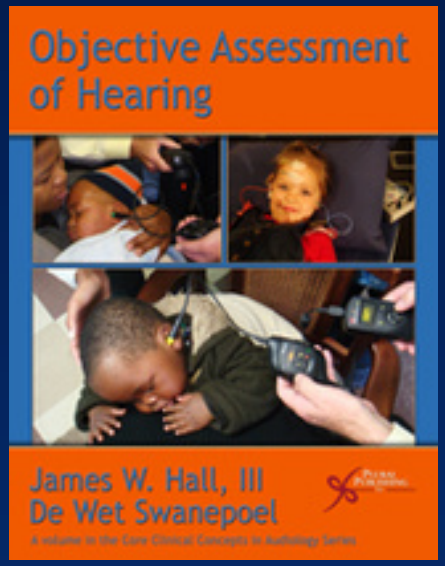
- Early diagnosis and intervention improves communication
- Accurate assessment of infant hearing is standard of care
- Efficient identification of (screening for) hearing loss
- The cross-check principle still rules
- An objective auditory test battery
  - Acoustic immittance measures
  - OAEs
  - ABR
  - ASSR
  - ECochG
- Summary

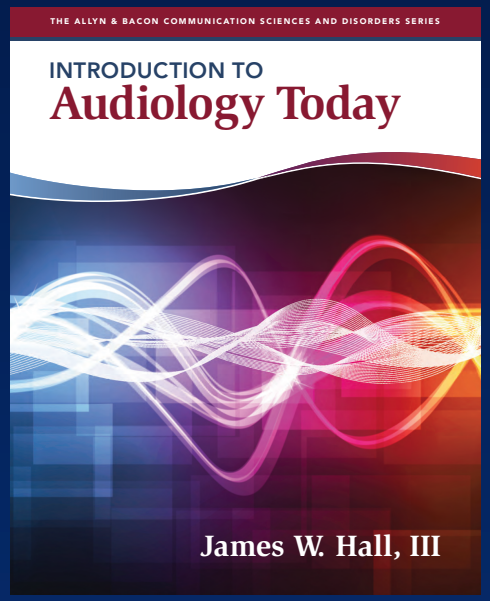
Electrophysiology Assessment of Infant Hearing: An Evidence-Based Protocol Summary

- Objective auditory tests permit early and accurate diagnosis of infant hearing loss
- The cross-check principle still is essential in pediatric audiology
- Each objective measure contributes importantly and rather uniquely to the diagnosis of infant auditory dysfunction
  - Immittance measures
  - OAEs
  - ABR and ASSR
  - ECochG
- Standard of care in infant hearing assessment is evidence-based and defined by peer-reviewed clinical guidelines

Thank You!
Questions?







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