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The Cross-Check Principle in Pediatric Audiometry Forty Years Later

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Abstract

"…we discuss a method of pediatric audiologic assessment that employs the 'cross-check principle.' That is, the results of a single test are cross-checked by an independent measure. Particularly useful in pediatric evaluations as cross-checks of behavioral test results are impedance audiometry and brainstem-evoked response audiometry…"

from *The Cross-Check Principle in Pediatric Audiometry*
James F. Jerger, Ph.D., and Deborah Hayes, M.A.
Archives of Otolaryngology, 1976, 102:59-65

Purpose of this Presentation

- Snapshot of the past ~ how much has changed in 40 years!
- Rationale for the Cross-check principle in 1976
  - Exciting new clinical developments
- Role of the Cross-check principle in 2016
  - Things we should do to improve clinical evaluation of infants and young children
    - Objective response detection of auditory evoked potentials
    - Click ABR
    - Ipsilateral and contralateral acoustic reflex measures
- Interesting patients yesterday and today ~ "…every patient is interesting…" James Jerger (N.D.)

Learning Outcomes

After this course, you will be able to:

- Define the cross-check principle and explain the history of its application to pediatric audiology.
- Explain how the application of the cross-check principle evolved over time with the development of new measures for audiology diagnoses, and its relevance today.
- Discuss how current techniques of objective response detection, click-evoked auditory brainstem response, and ipsilateral and contralateral acoustic reflexes can provide cross-checks and improve clinical pediatric audiology today.

What a change in 40 years! Things we no longer use

What a change in 40 years! Things we no longer do

- Electrodermal audiometry
- Crib-o-gram
- Auditory response cradle
- Heart rate audiometry
- Respiration audiometry
- Visual reinforcement audiometry with a flashing light bulb inside a plastic halloween pumpkin

What we did then: A patient from 40 years ago

- Baby A.B., a 12-month old female born after an uncomplicated pregnancy and delivery
  - Parents are concerned about baby's lack of response to sound and delayed verbal development
  - Audiological assessment consistent with profound hearing loss
    - No behavioral response in the sound field
    - Normal tympanograms with no acoustic reflexes (cross-check)
    - Click ABR under chloral hydrate sedation showed no response to clicks at 90 dB nHL (air conduction) or 40 dB nHL (bone conduction)

What we did then: A patient from 40 years ago

- What went right for baby A.B.
  - Early identification ~ she was only 12-months old when identified!
  - Probable recessive non-syndromic genetic deafness; no additional developmental delays or disabilities
  - High-achieving parents who were dedicated to her success
- What audiology contributed to her care
  - Body-worn hearing aid with a Y-cord
  - Recommendations for correspondence course for early intervention until preschool enrollment age (3)

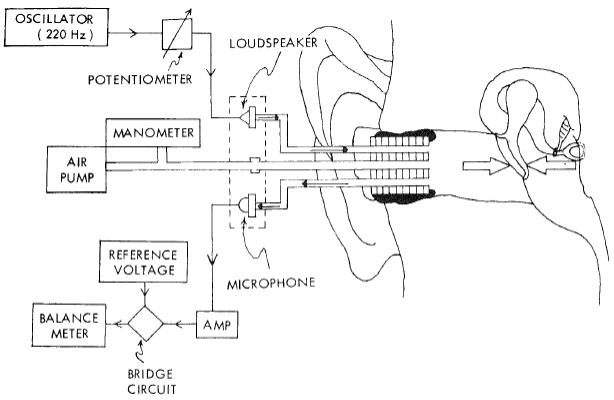
Why we needed the cross-check principle 40 years ago

- Rationale for the original article:
  - Children were receiving behavioral observation evaluations with no confirmatory physiological cross-check
    - Led to mis-diagnosis/missed diagnosis of hearing loss, especially in two groups of children
      - Young infants
      - Children with developmental delays
  - New techniques, introduced into the clinic in the '60s and '70s, were uniquely suited to evaluation of young children
    - Impedance audiometry ~ sensitive to middle ear disorder; predictive of sensory hearing loss; predictive of site of disorder from middle ear to brainstem
    - Brainstem-evoked response (BSER; now known as auditory brainstem response, ABR) ~ predictive of hearing loss above 500 Hz

The clinical application of new techniques

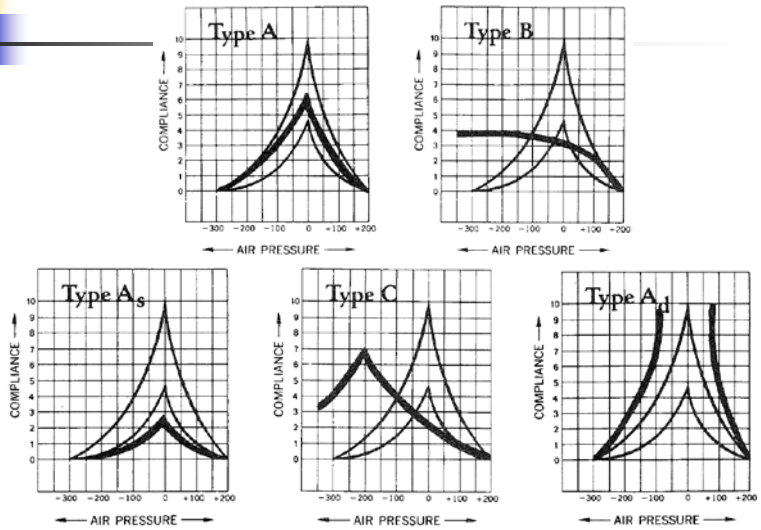
- Impedance audiometry; immittance measures
  - Tympanometry provided an indication of middle ear dysfunction with a unique signature for
    - Tympanic membrane perforations and/or presence of PE tubes
    - Middle ear effusion
    - Eustachion tube dysfunction
    - Ossicular fixation and ossicular disarticulation
  - Acoustic reflexes provided an estimate of presence of sensorineural hearing loss
  - Acoustic reflexes provided a window into the crossed and uncrossed reflex pathway
    - Middle ear – cochlea – eighth nerve – brainstem – seventh nerve

The 1970s Impedance Audiometer



Arch. Otolaryngol. 92: 311-324, 1970.)

The 1970s Tympanogram Types



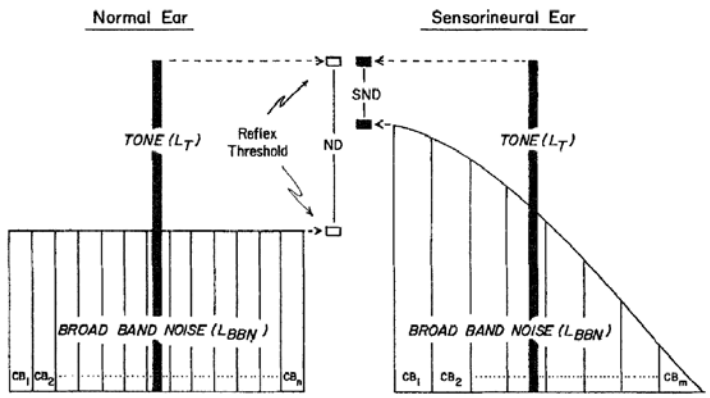
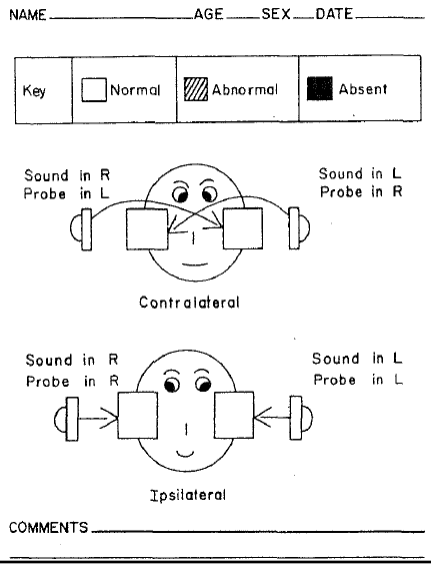


Figure 1. Hypothetical explanation for the phenomenon that the normal difference between tone and noise reflex thresholds is reduced in ears with sensorineural hearing loss.

From Jerger, Crump, and Mauldin, JSHD, 1974

A simplified method for comparing ipsi vs contra acoustic reflexes



From: Handbook of Clinical Impedance Audiometry, J. Jerger (ed), 1975

The clinical application of new techniques

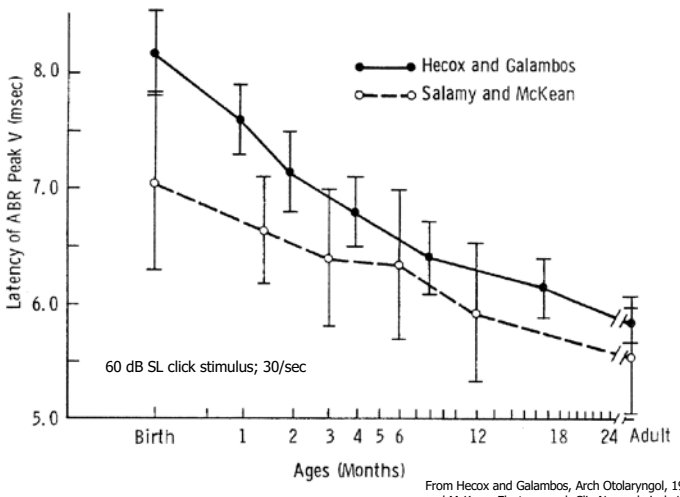
- Auditory brainstem response (ABR): The early years
  - Reliably present in individuals of all ages ~ newborns to adults
  - Independent of subject state ~ present in awake, sleeping, and sedated/anesthetized individuals
  - Well-defined response morphology
  - Predictable relationship between stimulus intensity and response latency
  - Response generators identified as eighth nerve and structures in the brainstem

Marion Downs holding *Hearing in Children,* 2nd Ed. (1978) with a 1970s era auditory brainstem evoked response system, the Nicolet CA1000.

The clinical application of new techniques

- ABR for hearing assessment in infants and children
  - Responses are present at adult threshold levels in very young children and even newborn infants
  - Response latency and interwave intervals are prolonged relative to adult values
  - Degree of hearing loss and site of dysfunction (outer/middle ear, inner ear, auditory nerve or brainstem) could be predicted with substantial accuracy from the pattern of ABR results
    - Conductive ~ elevated response threshold with delayed latency
    - Flat sensory ~ elevated response threshold with normal latency
    - Steeply sloping sensory ~ elevated response threshold with delayed latency at near-to threshold levels

Maturation of the ABR



From Hecox and Galambos, Arch Otolaryngol, 1974; Salamy and McKean, Electroenceph Clin Neurophyisol, 1976. Reprinted from Auditory Evoked Potentials Course Syllabus, The Neurosensory Center of Houston, May 10 – 12, 1980. Coats, A.C., and Jerger, J.

Summary: Cross-checks of the 1970s

- Impedance (immittance) measures confirmed:
  - Presence and nature of middle ear dysfunction
  - Site of auditory disorder (middle ear to brainstem)
  - Degree of sensory hearing loss based on SPAR
- Click BSER (auditory brainstem response) confirmed:
  - Nature of hearing loss (conductive, sensory, neural)
  - Site of auditory disorder (middle ear, cochlea, eighth nerve, brainstem)
  - Degree and configuration of hearing loss

What we do now: A patient from today

- Baby E.G., a five-week old male born after an uncomplicated pregnancy and delivery.
  - Referred on newborn hearing screening; re-screening
  - Audiological assessment consistent with profound hearing loss
    - No behavioral response was observed to any acoustic signal presented while infant was in light sleep
    - Unsedated click ABR showed no response at 90 dB nHL (air conduction) or 45 dB nHL (bone conduction)
    - ASSR showed no response at 80 dB HL at octave frequencies 250 through 8k Hz (cross-check)
    - DPOAEs were absent (cross-check)
    - 1000 Hz tympanograms were normal

What we do now: A patient from today

- Things that will go right for baby E.G.
  - Early identification ~ he is only 5-weeks old
  - Probable recessive non-syndromic genetic deafness; no additional developmental delays or disabilities
  - High-achieving parents who were dedicated to his success
- What audiology will contribute to his care
  - High-power ear level hearing aids
  - Referral to community resources and public, home-based early intervention program
  - Candidacy evaluation for cochlear implants before age 1 year

Is there a role for the cross-check principle today?

- Expectation of screening, diagnosis, and intervention before conditioned behavioral measures can be employed (Joint Committee on Infant Hearing "1-3-6 standard")
  - Newborn hearing screening is well established; in 2013, 97.2% of newborns in the US received NHS*
  - Infants who refer on NHS receive diagnostic audiological evaluation *by physiologic measures* by age three months
  - Infants identified with hearing loss are fitted with amplification based on results *of physiological measures* by six months of age

*http://www.cdc.gov/ncbddd/hearingloss/2013-data/2013_ehdi_hsfs_summary_a.pdf

Pediatric Audiological Test Battery: Birth to Six Months

- Behavioral observation ~ infant's interaction with the auditory environment
- Acoustic immittance measures ~ middle ear and acoustic reflex pathway
- Otoacoustic emissions ~ cochlear sensory cells
- Click-evoked auditory brainstem response ~ presence, degree and nature of hearing loss (normal, conductive, sensory, neural)
- Frequency-specific auditory evoked potentials ~ audiogram

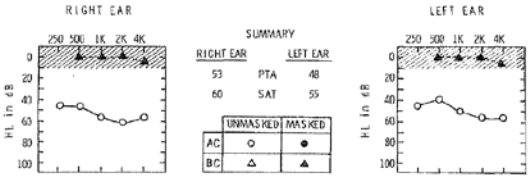
Why we need the cross-check principle today

- The pediatric audiological test battery should yield an internally consistent and complete picture of an infant's auditory function from outer/middle ear through brainstem. The audiologist must:
  - Understands the advantages and limitations of each component of the test battery ~ how is each test vulnerable to misapplication and misinterpretation?
  - Understands the relationship among tests comprising the test battery ~ are the results of test A consistent or inconsistent with the results of test B?
  - Resolves discrepancies in test results ~ what is the unique signature of different forms of auditory dysfunction?

Cases from the Cross-Check Principle

- Five cases discussed in the 1976 article
  - Case 1 ~ 15 year old boy mis-diagnosed with autism who was deaf
  - Case 2 ~ 2 year, 9 month old boy mis-diagnosed with moderate conductive hearing loss who had severe, high-frequency mixed hearing loss
  - Case 3 ~ 4 year, 6 month old boy with 4 different hearing test results from 4 different facilities
  - Case 4 ~ 5 year, 9 month old girl with auditory agnosia
  - Case 5 ~ 6 month old girl with first arch syndrome and bilateral atresia

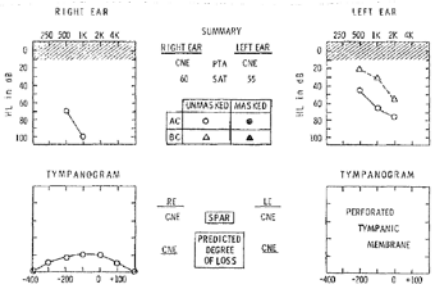
Everything old is new again: Case 2 from 40 years ago



Case 2 from "The Cross-Check Principle." Audiogram of a 2-3 year old boy from an otologist's office. Youngster had been in his care for 18-months for recurrent middle ear disease.

Results from The Methodist Hospital for Case 2 at age Audiogram and impedance audiometry results on case 2 at age 2-9.

Auditory brainstem response yielded **no response** to air-conduction clicks at 80 dB nHL and responses to bone conduction clicks at 55 dB nHL, confirming at least a severe sensorineural hearing loss bilaterally



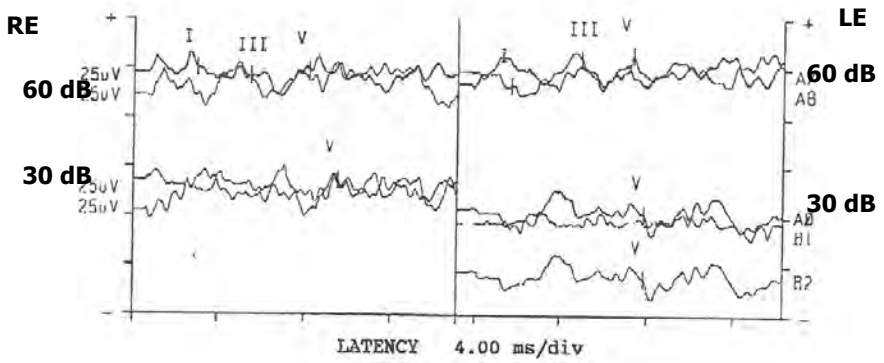
Everything old is new again: Case EM from today

- Full-term male infant; uneventful pregnancy and delivery
- Referred from NHS; did not pass screening or outpatient re-screening in either ear
- Family history of childhood hearing loss including mother and maternal grandfather
- Diagnostic audiological evaluation conducted at age 2 months at birth hospital and age 3 months at Children's Hospital Colorado

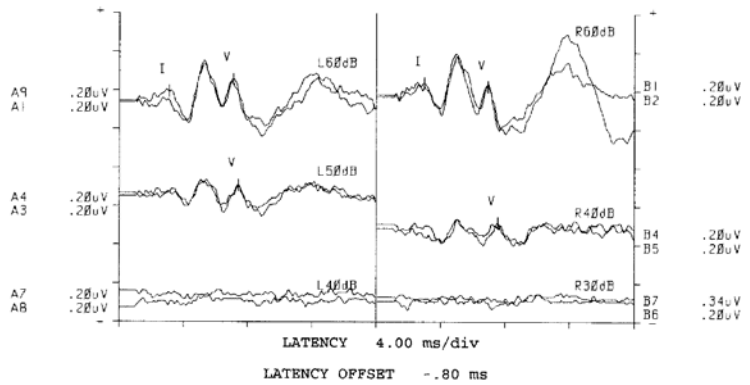
Child: EM Gender: M Age: 2 months Diagnostic Evaluation at Birth Hospital

ABR interpreted as "normal";

baby discharged from follow-up

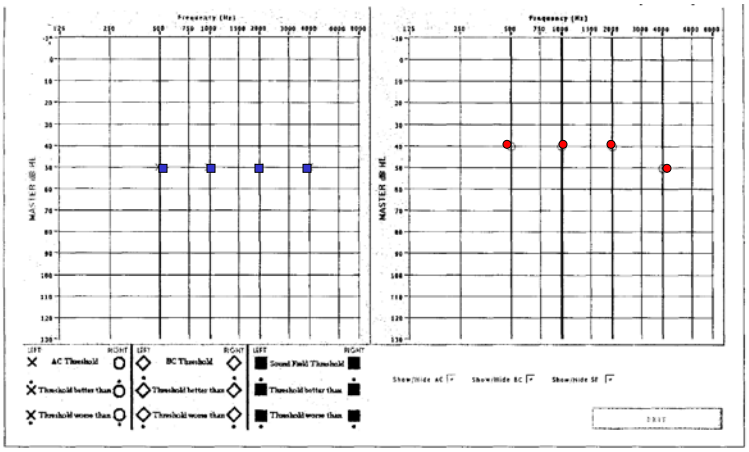


Child: EM Gender: M Age: 3 months
Diagnostic Evaluation at Children's



DPOAEs absent bilaterally

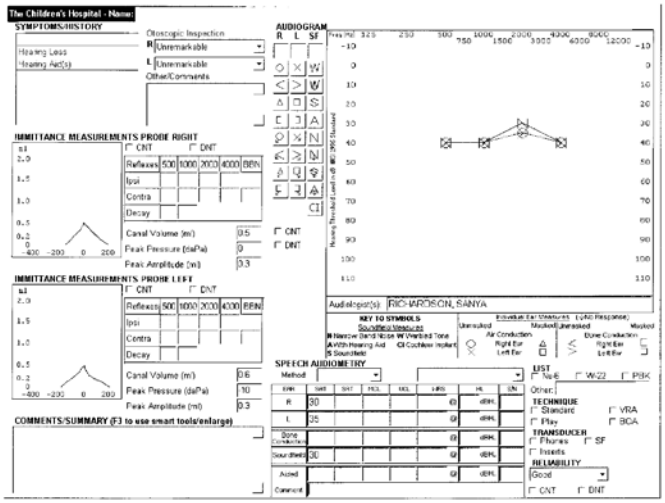
Child: EM Gender: M Age: 3 months
Diagnostic Evaluation at Children's



Child: EM Gender: M Age: 3 months
Children's Hearing Clinic Evaluation

- Otologic and Genetic Assessment
  - Connexin 26 - Negative
  - CMV screening – Negative
  - Laboratory studies – Negative
  - CT Imaging – Not significant
  - Diagnosis: probably autosomal dominant nonsyndromic hearing loss (mother and maternal grandfather with mild, bilateral, apparently congenital hearing loss)

Child: EM Gender: M Age: 9 months
Behavioral Evaluation at Children's



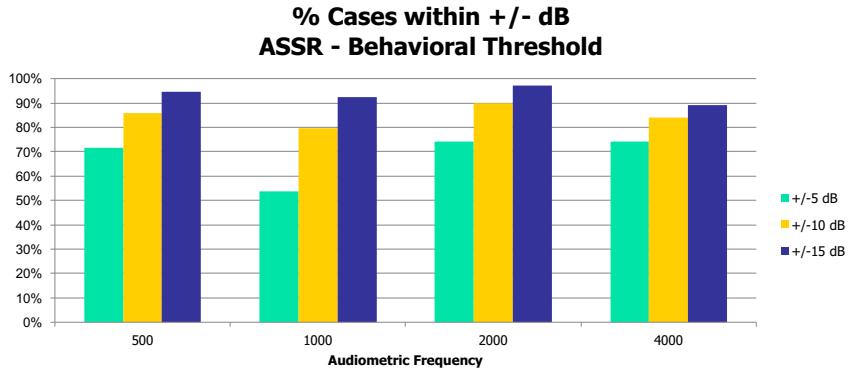
What can we learn from EM? Diagnostic audiological evaluations for infants

- Evoked potential recording conditions must be optimum to obtain valid, artifact-free, unambiguous test results
  - Infants must be soundly sleeping to avoid movement artifact or "noisy" recordings.
  - Utilizing objective response detection methods can limit errors related to observer bias
- Diagnostic evaluations must include more than a single measure of auditory function
  - OAEs and immittance measures can serve as a cross-check of auditory evoked potentials; if results are inconsistent, follow-up evaluation is warranted

Objective response detection in auditory evoked potentials

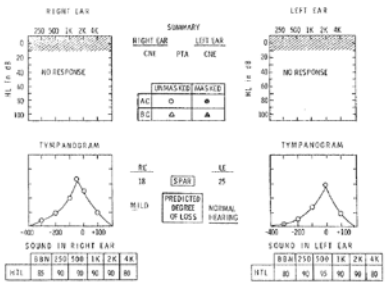
- Visual waveform analysis ("peak picking") of evoked potentials is dependent on audiologist experience and informal criteria resulting in observer bias. Confounding variables include recording quality, subject quiescence, and inter-subject variability.
- Statistically-validated objective response detection criteria for auditory evoked potentials include:
  - Fsp/Fmp ~ based on the estimated signal to noise ratio in the averaged ABR response; compares variance in the response (signal) plus noise to the variance in the noise to calculate the probability of presence of a response.
  - F ratio ~ probability that amplitude at stimulus modulation frequency is within the distribution of amplitudes at neighboring frequencies
  - Phase coherence ~ analysis of response amplitude and phase to determine probability that response is phase-locked to stimulus
- These techniques are available on commercial AEP systems.

Predictive accuracy of ASSR by F-ratio objective response detection



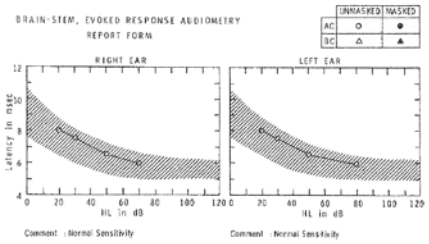
Results from 205 children tested by both ASSR and behavioral audiometry at Children's Hospital Colorado.

Everything old is new again: Case 4 from 40 years ago



Case 4 from "The Cross-Check Principle." Audiogram and impedance results of a 5 - 9 year old girl from The Methodist Hospital. Youngster had documented rapidly progressive hearing loss following a high-fever illness and grand mal seizure at age 2 – 6 years.

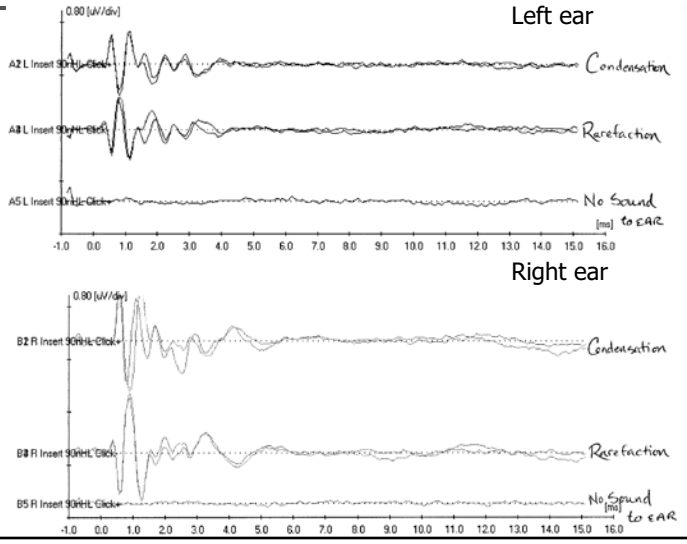
ABR from case 4 obtained at The Methodist Hospital. Results of acoustic reflex measures and ABR are consistent with normal peripheral hearing sensitivity in both ears. Child's ultimate diagnosis was *auditory agnosia*, an exceedingly rare neurological condition.



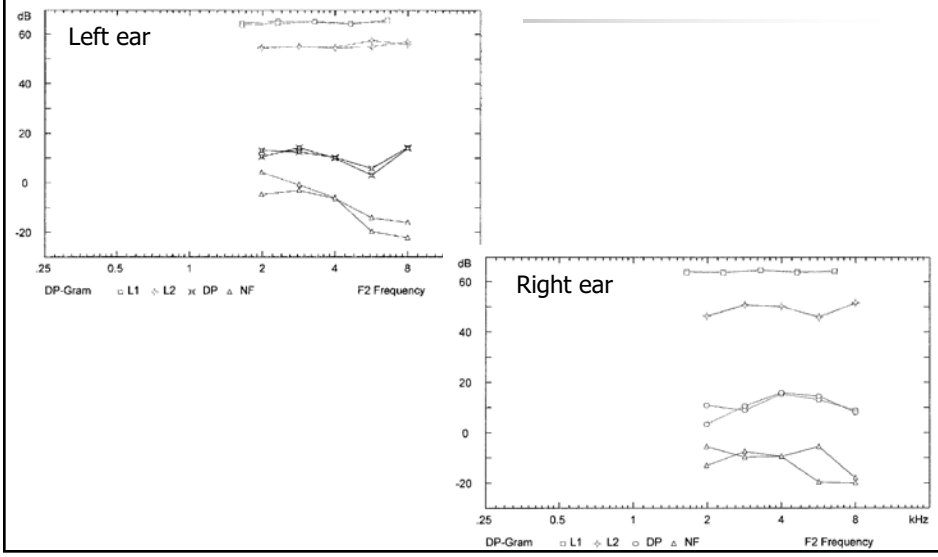
Everything old is new again: Case DD from today

- Born at 33 weeks gestation; no prenatal care; unattended birth; birthweight 1660 grams; appropriate for gestational age
- Delayed resuscitation (? duration); spontaneous respirations when EMS arrived
- Stabilized and transported to Children's Hospital Neonatal Intensive Care Unit (NICU)
- Hospital course included (6 weeks)
  - Mild hyperbilirubinemia
  - Respiratory distress; surfactant and supplemental oxygen
  - Rule out sepsis; ampicillin and gentamicin

Child: DD Gender: M Age: <1 month Diagnostic Evaluation at Children's



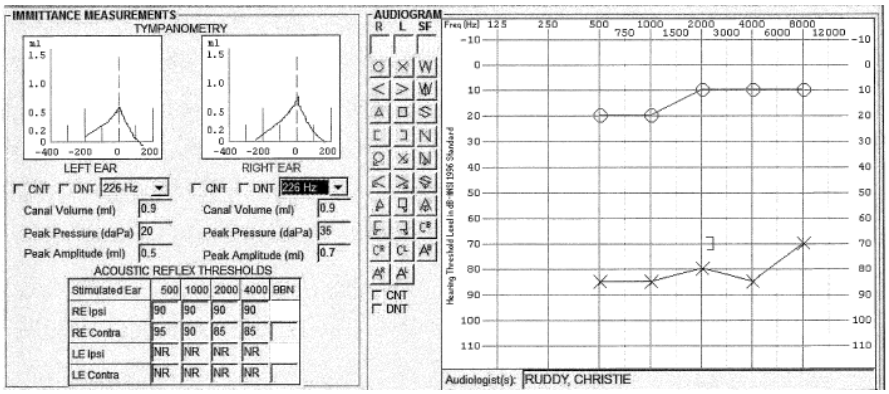
Child: DD Gender: M Age: <1 month Diagnostic Evaluation at Children's



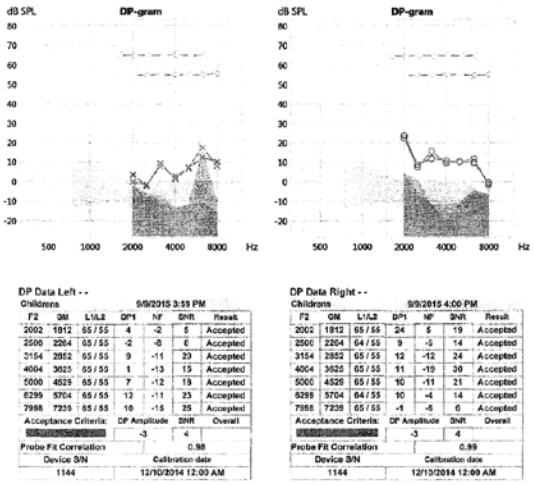
Another interesting case: LM Gender: M Age: 4 - 3

- Typically developing child; normal pregnancy and delivery; no significant family or medical history
- Passed NHS; parents report no suspected hearing problems and normal speech and language development
- At age 3 – 10, received audiological evaluation through local Child Find program; unilateral left hearing loss identified
- Age 4 – 3, received audiological evaluation at Children's Hospital Colorado

Case LM
Audiogram and Acoustic Immittance



Case LM
Otoacoustic Emissions



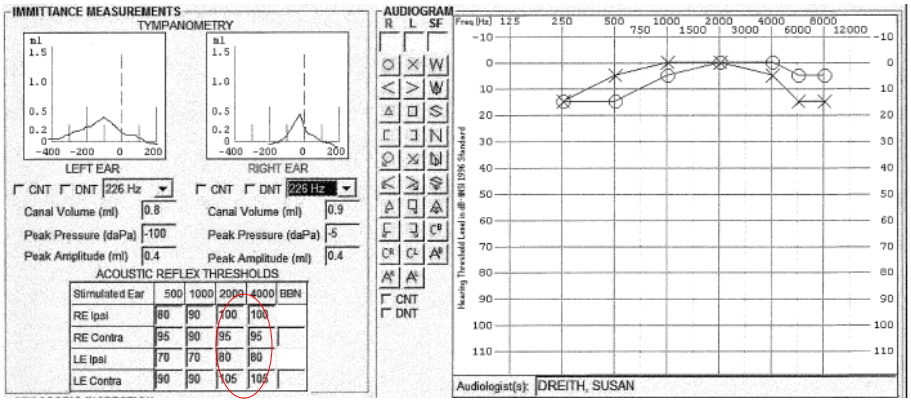
What can we learn from DD and LM?

- Auditory neuropathy presents in many forms ~ bilateral, unilateral, NICU, and well-babies.
  - Because AN is more common in NICU infants, ABR must be the hearing screening technology.
  - Regardless of screening technology, parents must be informed that hearing screening does not detect all forms of hearing loss.
- In a well-baby is often related to absent or abnormally small auditory nerve; MRI has been ordered for LM.
- For children who can be tested behaviorally, otoacoustic emissions are a valuable cross-check.
- When results are apparently inconsistent, further evaluation with an independent cross-check is warranted.

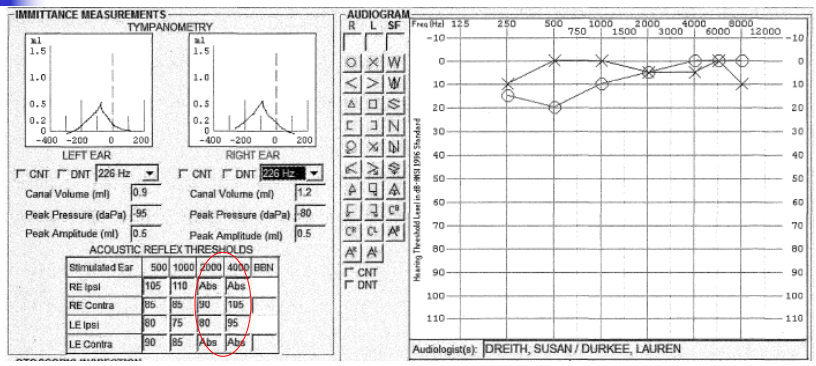
One more interesting case
MSM Gender: F Age: 12

- Youngster with complex medical history including prematurity, failure-to-thrive, multicystic single kidney, persistent asthma
- Newly diagnosed with neurofibromatosis type II; mother deceased from disease at age 30
- Bilateral vestibular schwannomas and right trigeminal schwannoma
- No complaints of ear pain, hearing loss, or vertigo

Case: MSM Gender: F Age: 12
Diagnostic audiology; 12/17/14



Case: MSM Gender: F Age: 12
Diagnostic audiology; 6/10/15



What can we learn from MSM?

- The acoustic reflex can provide information about the neural integrity of the pathway including the eighth nerve, rostral brainstem, and seventh nerve.
- Comparison of ipsilateral vs. contralateral acoustic reflexes yields specific patterns of acoustic reflex results that are consistent with afferent, efferent, and mixed afferent and efferent pathway abnormalities.
- Abnormal right ipsi and left contra on MSM's second test suggest right efferent (seventh nerve) pathway effect.

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  - Exciting new clinical developments
- Role of the Cross-check principle in 2016
  - Things we should do to improve clinical evaluation of infants and young children
    - Objective response detection of auditory evoked potentials
    - Click ABR
    - Ipsilateral and contralateral acoustic reflex measures
- Interesting patients yesterday and today ~ "…every patient is interesting…" James Jerger (N.D.)

The Cross-Check Principle
The Next 40 Years

- The Cross-check principle will continue to age well
- The basic premise is robust:
  - to incorporate new, validated test techniques to improve accuracy in audiological diagnosis and management of infants who are deaf or hard-of-hearing.
- I am confident that future generations of audiologists will continue the progress we have made and improve audiological care of this precious popuation.