Initial diagnostic testing of infants who fail newborn screening

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Best textbook chapter ever written on this subject !

Thanks to:

Dr David Stapells
University of British Columbia
Vancouver, Canada

Frequency-Specific Threshold Assessment in Young Infants Using the Transient ABR and the Brainstem ASSR

Chapter 21, in: RC Seewald & AM Tharpe (Eds)
Comprehensive Handbook of Pediatric Audiology
San Diego, Plural Publishing

Challenge: after AABR screen fail or direct high-risk referral:

- Significant hearing loss? YES / NO
- If YES, what type?
  Conductive (T or P) Conventional cochlear
  ANSD Brainstem neural
  Any mixture of these !
- If NOT Conductive (T), ANSD or neural:
  Thresholds sufficient to specify amplification,
  Implant candidacy, baseline for progression

Additional challenges

- Validity & accuracy, given limited data in young infants with proven permanent hearing loss, especially for ASSR & for bone-conduction
- Efficiency & completeness, given three pressures: access, baby EEG state & early intervention

Present diagnostic tools

- Tonepip ABR (air & bone conduction)
- Brainstem (80 Hz) ASSR: single/multiple-frequency
- Click ABR & cochlear microphonic (CM)
- (Long-latency cortical potentials)

- OAE (Distortion Product or Transient): 1-4 kHz
- Tympanometry:1 kHz probe < 6 months
- Middle-ear muscle reflexes: ipsi, 1k, wide-band

AABR screen will NOT fail:

- ANY abnormal audiogram with ANY threshold better than 30 dBHL in the range 1-8 kHz

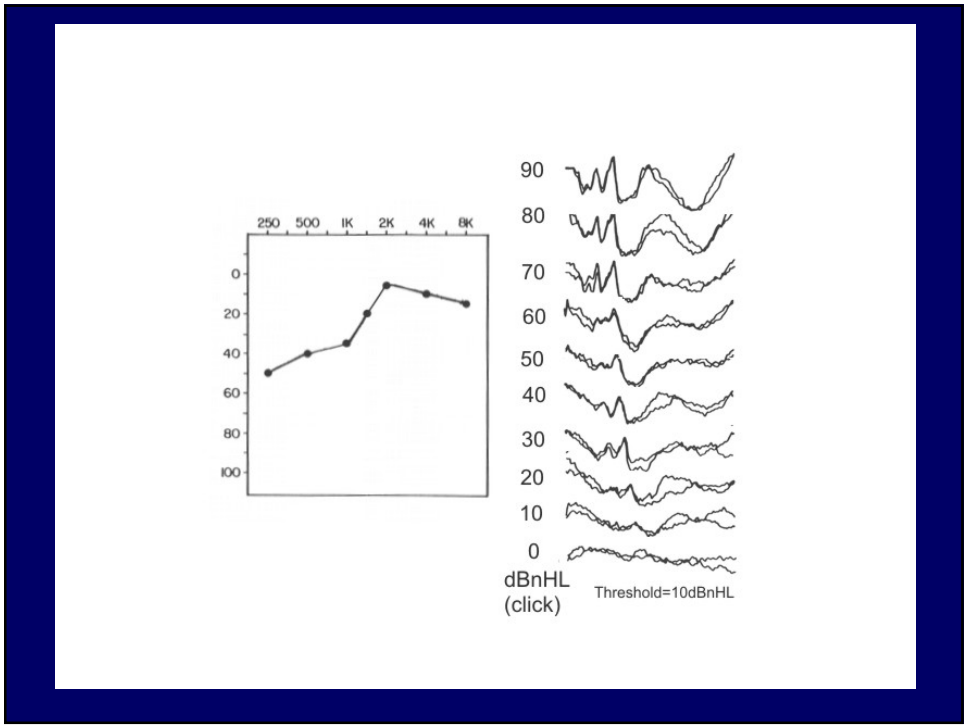
- These include:

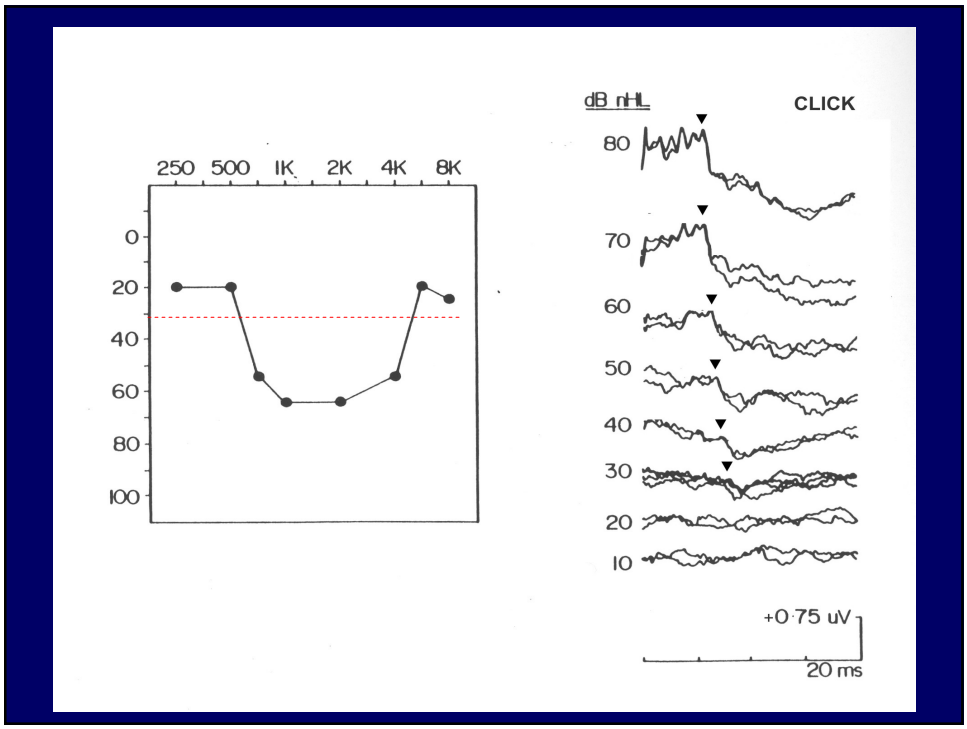
  Sloping losses normal at 1kHz or above

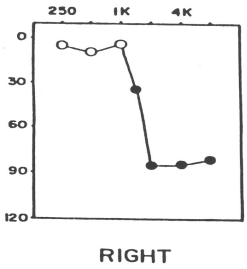
  Steep high-frequency losses above 1kHz

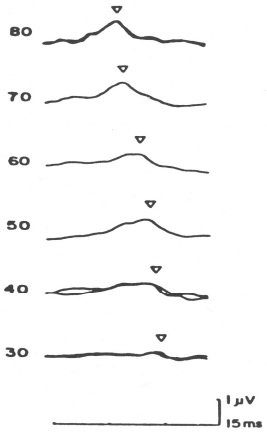
  Reverse-slope losses normal at 8kHz or below

  U-shaped losses normal at 1kHz or 8kHz


250 500 1K 2K 4K 8K
0
20
40
60
80
100
90
80
70
60
50
40
30
20
10
0
dBnHL
(click)
Threshold=10dBnHL

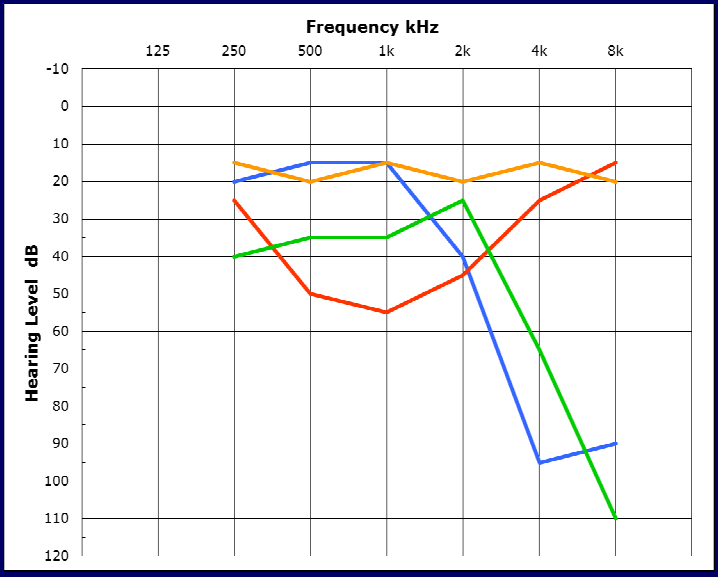

250 500 1K 2K 4K 8K
0
20
40
60
80
100
dB nHL
CLICK
80
70
60
50
40
30
20
10
+0·75 uV
20 ms



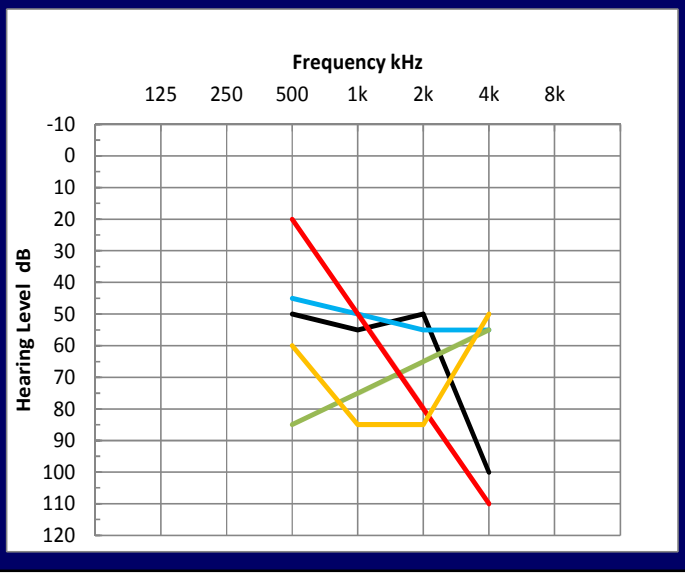


Adapted from: Picton, 1978

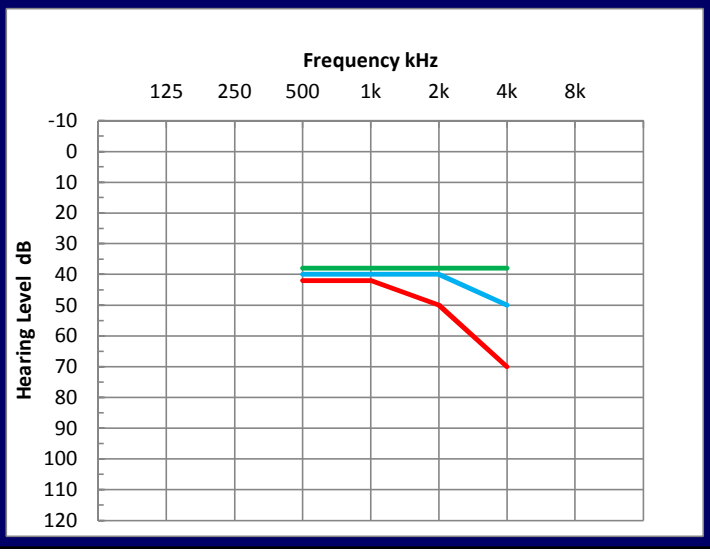
Click ABR present at 30 dBnHL



Click ABR threshold 60 dBnHL
Same amplification…???



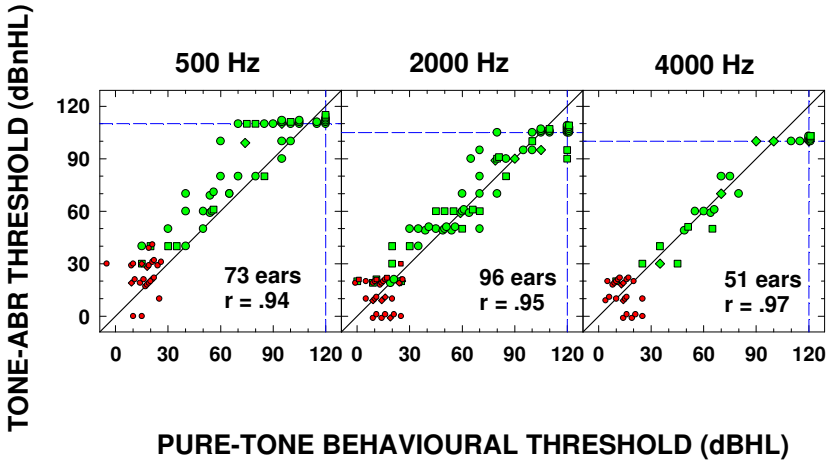
Click ABR threshold 50 dBnHL
Monitoring: no progression..???



Clicks are an otoneurologic tool, not a sufficient threshold tool

- Would you do an adult hearing test with white noise only?
- Infants deserve the best possible audiometry
- Clicks have many technical deficiencies & may soon be replaced by chirps

Best published data to date + long experience

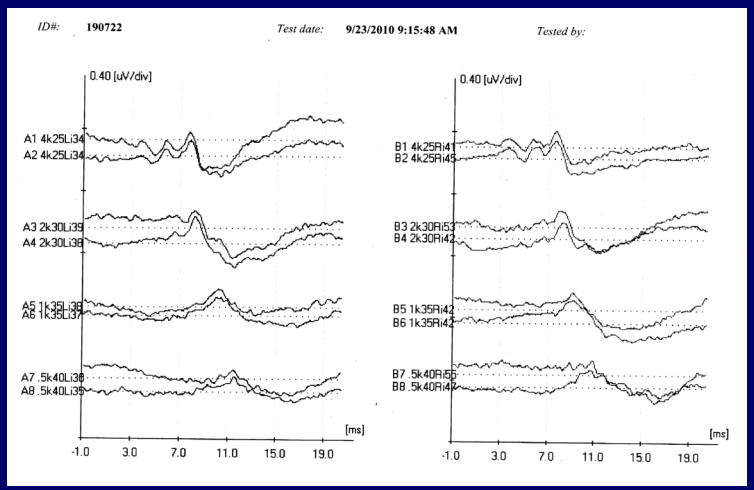


Stapells, Gravel & Martin, 1995

Estimated Hearing Level (eHL)

- Determine tone-ABR threshold:
  ABR present at x dBnHL, absent at x-10
- Correction factor C is normative median difference (ABR threshold – HL threshold)
- eHL threshold = ABR threshold – C
- AC 500 15 dB 1k 10 dB 2k 5 dB 4k 0 dB

Minimum intensity levels (25 dBeHL)
Recording time < 10 mins



Successful diagnostic ABR testing

- Sufficient caseload to develop/maintain skills
- 4-8 weeks corrected age, 4 weeks after screen
- Natural sleep, arrive tired & hungry, feed after electrode attachment
- TWO insert transducers, whenever possible
- Hand-held BC if trained, otherwise use Velcro band

- High-efficiency (top-down, progressive) strategy:

Every test condition is chosen so its result will make the greatest difference to baby MANAGEMENT if the test stopped at that point….

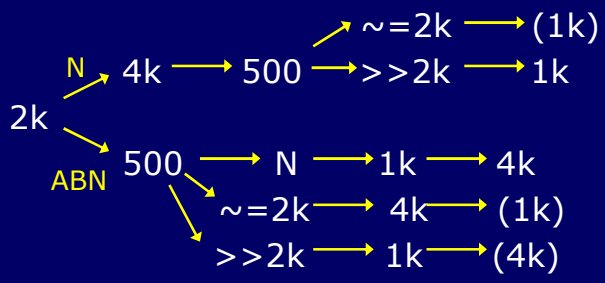
Minimum test for AABR failures?

Loss probability: No risk 0.1, High risk 0.4

- Statistical false positive AABR – N hearing
- ABR detection failure – high EEG noise levels
- Transient hearing loss/debris/probe blockage
- Permanent hearing loss (1-8 kHz)/ANSD
- Test options: 2k, 4k, 1k, 500
  2k, 500 or 4k, 1k
  2k only !?
- ABR at minimum required level? Yes: STOP
  No: ENTER EFFICIENT AUDIOGRAM STRATEGY

Minimum test for direct Dx referral (eg very high risk, no AABR screen)

- ANY hearing loss profile is possible, so:
- ENTER EFFICIENT AUDIOGRAM STRATEGY



Efficient loss type strategy

- Try OAE first if baby not asleep (esp. high risk)
- If OAE 2k present & 2k ABR absent at 90 dBnHL, ANSD is present
- If first ear 2k 'normal', change ears immediately
- If first ear 2k threshold >=40 dBnHL go to BC
- If BC 2k ABR present at min level, stop

Efficient threshold strategy

- Start with 2kHz at minimum level & go up in LARGE steps (20-30 dB)

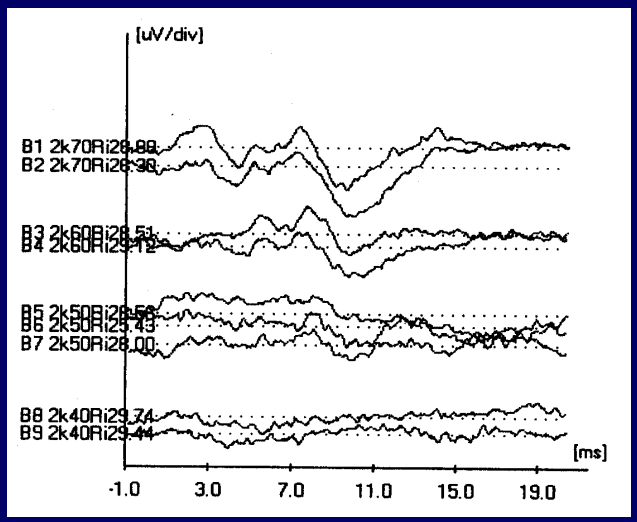
  Never do input-output function with 10 dB steps !

- Use 1-2000 sweeps with repetition if needed

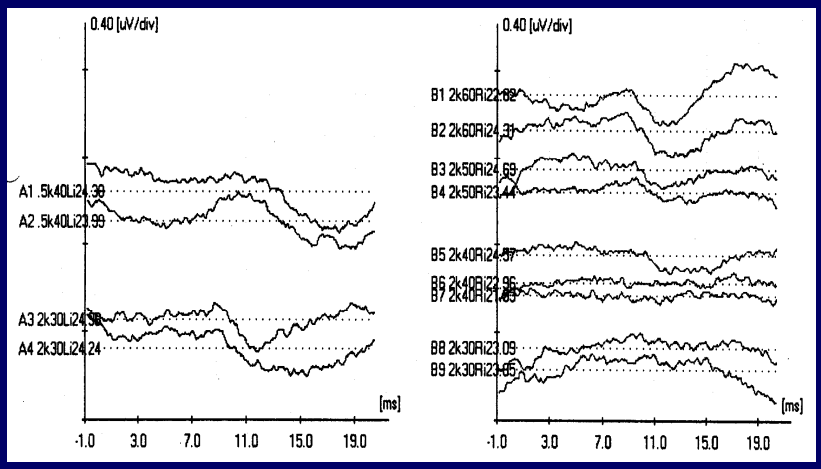
- Bracket threshold with 10 dB step, repeated averages & use residual noise level (RNL)

- Aim to locate threshold in <= 6 averages

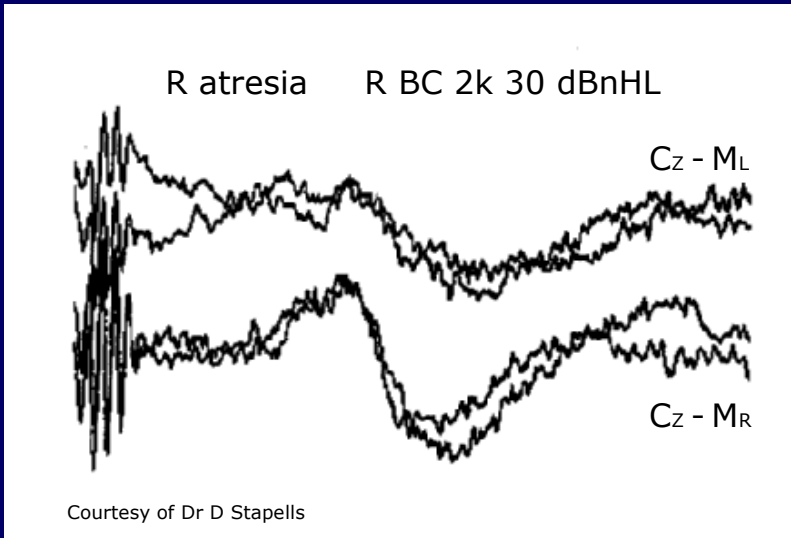
Example of inefficiency



False +ve & low-noise -ve

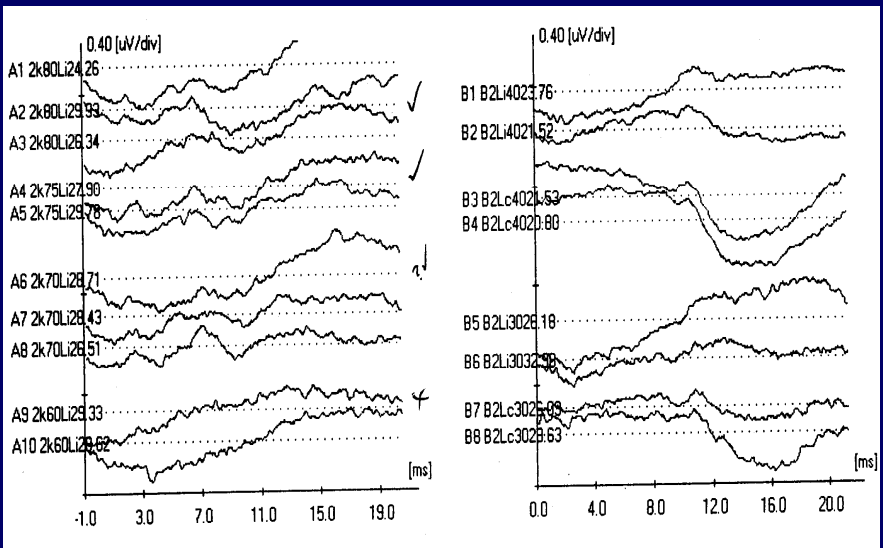


BC: Inference of responding cochlea



Courtesy of Dr D Stapells

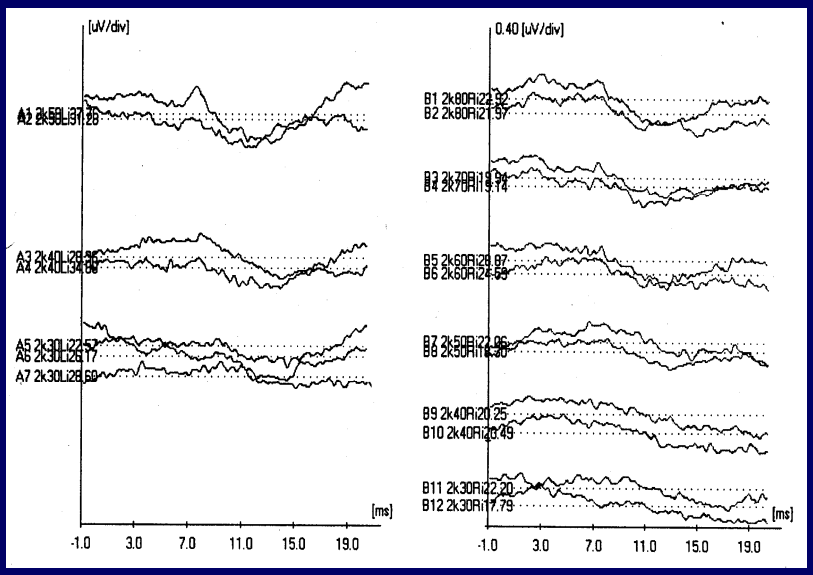
2 kHz BC ipsi-contra in a unilateral



Common error in ABR detection

- There are THREE decision outcomes, not TWO: Present, Absent, and INDETERMINATE
- If EEG noise level is not low enough, you cannot and must not say that response is absent or present (different criteria for each)
- Guessing is much worse that saying 'I don't know'
- Should use residual noise level if available

Not so easy

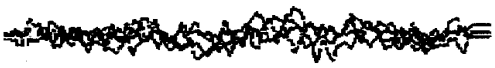


Using SNR & residual noise (IHS)

Present

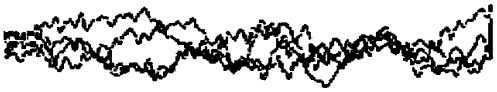
SNR 1.26 RN 0.07µV

Absent



SNR 0.64 RN 0.04µV

Indeterminate



SNR 0.16 RN 0.15µV

ASSR: some comments

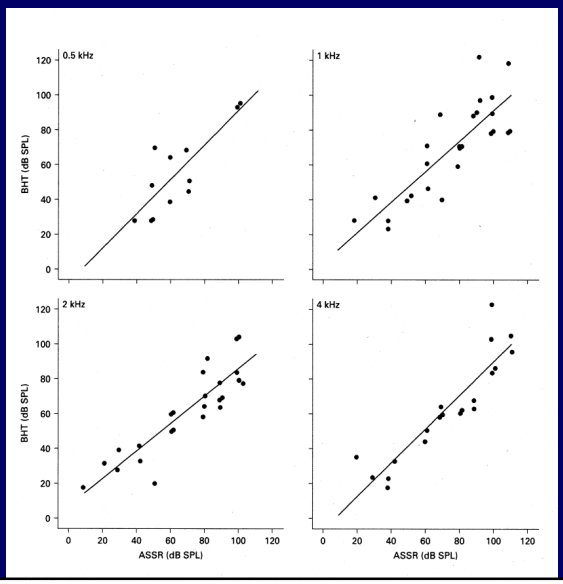
- 80Hz ASSR is probably ABR V-V' – difficult to prove
- High stimulus rate is efficient, objective detection is convenient, immunity to 60 Hz interference is useful
- 'Better frequency-specificity' NOT proven in infants with definite SNHL
- 'Can measure greater hearing losses' is probably an artifact of dBnHL vs HL (or vestibular response)

Tone-ABR vs s-ASSR vs m-ASSR

- m-ASSR less efficient for frequency-intensity strategy
- In young infants with proven SNHL:
  Very few *good* studies of s- or m-ASSR accuracy
  No normative studies of BC
  No good studies of relative efficiency
  Insufficient data on stimulus interactions
  Optimal test parameters are not well-established

Behavioural (VRA) vs m-ASSR, 21 infants

Luts et al Audiol Neurotol 2006;11:24-37



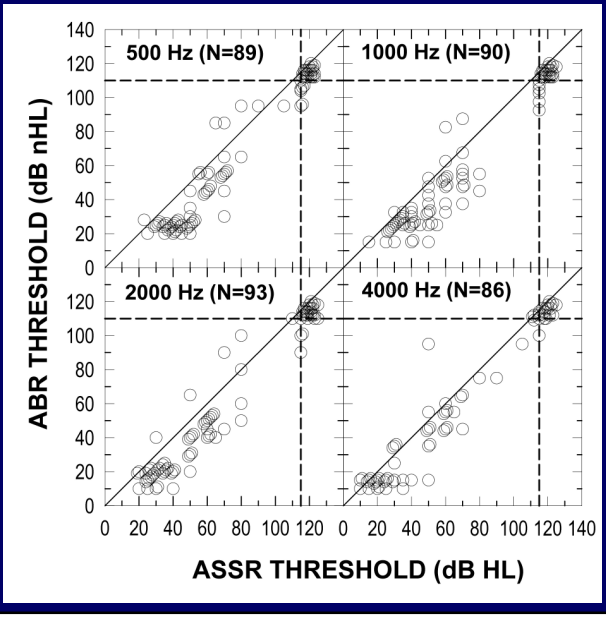
BUT…we *can* say that in infants:

- Multiple ASSR (one or two ears, AC) is a valid & efficient tool to prove absence of significant hearing loss, given response at:

	500 Hz	1 kHz	2kHz	4kHz
dBHL	50	45	40	40

- Recent data show a strong relationship between multiple-ASSR & tone-ABR thresholds (Van Maanen & Stapells, JAAA 2010)

Multiple ASSR (AC) vs tone-ABR in infants with hearing loss *(Van Maanen & Stapells 2010)*



Mensajes para llevar…

- Clicks are not good enough
- Tone ABR AC & BC is valid & practicable
  High-efficiency standard protocol is essential
- DO NOT need local norms. Google: 'BC Early Hearing Program Diagnostic Audiology Protocol'
- ASSR is efficient for normal/abnormal decision
- Insufficient ASSR data for complete audiometry