Implant candidacy assessment in patients with cochlear nerve deficiency

Introduction

- Cochlear Nerve Deficiency (CND)
  - MRI “gold standard” (Parry et al, Buchman et al)
    - Absent
    - Hypoplastic
      Smaller than facial nerve
Assessment

• Despite absence of CN on MRI, the auditory system may be at least partially functional.

• Severe-to profound HL degraded ABR response.

• AN/AD, which often co-exist with cochlear nerve deficiencies, also result in abnormal ABR findings (Buchman)
P1 Central Auditory Evoked Potential

- The P1 is generated within the auditory cortex.
- The latency of the P1 CAEP decreases systematically as age increases.
- Therefore it may be used as a biomarker to assess central auditory development and function in hearing impaired children (Sharma et al., 2005; Sharma and Dorman 2006).

10 dB SL
- Less synchrony than ABR

Normal Hearing Children (N=190)

Sharma et al., Ear and Hearing, 2002
Methods

- Retrospective case review
- /ba/ stimulus
- Via HA or insert earphones

(Sharma, Dorman and Spahr, 2002)

Case 1

- 21 month w AN/AD by ABR
- *no visible cochlear nerve bilaterally.*
- IACs normal in size
- No Behavioral audiometric testing
Case 1

Left Insert Phone

Right Insert Phone

2 μV

Latency (msec)

-100 0 100 200 300 400 500 600

Frequency (Hz)

Hearing Level (dB)

250 500 1000 2000 3000 4000 8000
Case 2

- ABR @ 22 months suggested severe to profound bilateral SNHL
- *mild cochlear dysplasia*
- Cochlear nerve could not be visualized on either side.
Case 2

- ABR testing at age 2 months showed severe-to-profound loss bilaterally.
- Binaural hearing aids at age 5 months based on the estimated ABR result.
- MRI hypoplastic cochlear nerves bilaterally.

Case 3
Case 3

- Aided P1 results revealed a robust P1 response with normally developing morphology and latency.
- A second ABR was completed at age 8 months which yielded the same result as her initial testing; estimating a severe-to-profound hearing loss for the right ear, and a severe hearing loss for the left ear.
- P1 latencies, tested longitudinally up to 23 months post hearing aid fitting, continued to remain normal.
Case 3

Left Insert Phone

Right Insert Phone

2 μV

Latency (msec)

Case 3

Case 4

- Patient 4, failed her newborn hearing screening
- A profound bilateral hearing loss was estimated using ABR measures at age 12 months.
- Her MRI revealed severe cochlear and vestibular dysplasia with absent cochlear-vestibular nerves bilaterally.

Southwestern Medical Center
Case 4
Case 4

- Absent P1 responses at very high intensity levels, indicated that no acoustic information was being relayed to the central auditory pathways.
- Based on the collective evidence of MRI, behavioral, and P1 CAEP test results, cochlear implantation was not recommended for this patient.

Conclusion

- Absence of cochlear nerves on MRI does guarantee absence of function.
- Both behavioral and electrophysiologic testing helps clarify the extent of functional compromise and can help select the appropriate intervention.
Conclusion

- In these cases, P1 CAEP testing correlated well with behavioral testing suggesting that in the absence of behavioral testing (very young) the P1 is adequate for decision making.