WHY DO COCHLEAR IMPLANTS HELP PATIENTS WITH AUDITORY NEUROPATHY/DSYCHRONNY

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We have implanted 35 children & 2 adults with auditory neuropathy/dys-synchrony (AN/DYS)

In this presentation, I will review:
1. the characteristics of auditory neuropathy and
2. how we have used objective measures to verify neural synchrony

Pre-operative characteristics of our patients with Auditory Neuropathy

- variable ranges of sensorineural hearing loss
- reduced speech perception
- abnormal temporal processing
- abnormal evoked potentials (ABR)
- middle ear reflexes absent
- otoacoustic emissions usually present

BACKGROUND INFORMATION

- Starr et al. (1991) identified auditory neuropathy (AN) as a timing disorder
- Zeng et al. (1999) demonstrated temporal processing problems in adults with auditory neuropathy.

BACKGROUND INFORMATION

- Berlin et al. (2001), and Hood (2002) proposed auditory dys-synchrony as a more descriptive term Typical findings include:
  - abnormal evoked potentials
  - otoacoustic emissions (usually present)
  - absent middle ear reflexes
  - lack of benefit from amplification, but not in all cases.

IDENTIFYING AUDITORY NEUROPATHY

- Berlin et al. (2001): Recommend 4 standard tests in addition to pure tone audiometry
  - Tympanometry
  - Middle ear reflexes
  - Otoacoustic emissions
  - Click / toneburst ABR (phase specific)
This child was seen for an evaluation at 15 months of age due to delayed speech and language. Her parents reported:
- normal birth history
- no complications during pregnancy
- normal labor & delivery
- mild jaundice at birth
- treatment for hyperbilirubin was not required

Her medical evaluations yielded negative findings for ENT, neurology, and ophthalmology. Initial medical genetics findings were negative for any syndromes. Magnetic resonance imaging studies were negative for any abnormalities.

However, subsequent genetic studies have shown that this patient and her brother have a mutation of the Otoferlin Gene.


Pre-operative Tests
- Audiogram & speech recognition
- Hearing Aid assessment & trial
- Tympanometry & acoustic reflexes
- Otoacoustic emissions
- Auditory Brainstem Response
- Condensation vs. rarefaction click stimuli

ANC01: Her pre-operative hearing tests were typical of all of the auditory neuropathy children that we have implanted.

Note that the CM is phase reversing.

Outer hair cells intact. Inner hair cells ??
Post-operative Testing

IOM pNRT values are above her initial behavioral thresholds and comfort levels

pNRT @ 60 mo approximates her behavioral thresholds

“Growing Programs”
OBJECTIVE: To evaluate the viability of cochlear implantation in children diagnosed with Auditory Neuropathy/Dyssynchrony (AN).

DESIGN: Six pediatric subjects were included in this study. Preoperative testing included otoacoustic emissions, auditory brainstem response testing, tympanometry and behavioral audiologic testing. Although audiometric thresholds ranged from mild to profound, performance with hearing aids was poor for all subjects. Five subjects were implanted with the Nucleus CI24RCA prosthesis and one with the Advanced Bionics 90K device. Length of implant use ranged from 3–18 months. Post-operative testing included behavioral audiologic thresholds and speech perception testing including the ESP, GASP, MLNT and Common Phrases.

RESULTS: At the last evaluation, 5 of the 6 subjects demonstrated significant improvement in auditory and linguistic skills, and one subject showed marginal improvement.

CONCLUSION: Children with AN can benefit from cochlear implantation.

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Green et al. 2005

Children diagnosed with Auditory Neuropathy/Dys-synchrony have the potential to benefit from cochlear implants beyond what is obtained from traditional amplification. As with other cochlear implant recipients the presence of developmental disabilities influences outcome. A coordinated team effort including audiologists, speech language pathologists and teachers of the deaf is necessary to maximize outcome.

Summary and Conclusions

- Educational setting is a significant factor
- Electrical stimulation provides neural synchrony which enables children to hear clear speech
- AN children and adults can benefit from cochlear implants

Auditory Neuropathy and Cochlear Implantation in Children

Alyce Breneman, M.A.

Summary and Conclusions

- Educational setting is a significant factor
- Electrical stimulation provides neural synchrony which enables children to hear clear speech
- AN children and adults can benefit from cochlear implants

Bringing clinical experience & research information to you

- Are there certain risk factors or case history information that we should look for regarding early identification of AN/AD?
- What are some of the management issues that we should be aware of?

Clinical profile

- In most cases AN/AD presents with specific medical risk factors
- However, it can occur in the absence of any obvious medical problems or hearing-related risk factors


Risk factors in infants

- Hyperbilirubinemia
- Premature birth
- Perinatal asphyxia
- Exchange transfusions

Risk factors in infants

- Neonatal insults
  - Anoxia
  - Hyperbilirubinemia
- More than 50% of early onset AN/AD cases have shown one or both of these conditions in their neonatal histories

Rance, 2005

Profile: permanent or fluctuating hearing loss

- Fluctuation in both hearing level and perceptual ability
- Threshold variances are significant enough to produce clear differences in functional hearing and speech understanding (Rance, 1999)
- Parents report: good/bad hearing days

Profile: speech perception

- Speech perception is usually poorer than expected
- Good speech perception is possible
- Problems in noise

Profile: Hearing aid use

- Won’t wear hearing aids in spite of good efforts at keeping them on
- Not making progress as expected with hearing aids even when the hearing aids are providing access to speech

Summary: red flags from the case history

- Neonatal risk factors:
  - Anoxia, hyperbilirubinemia, exchange transfusion
- Good/bad hearing days
- Won’t wear hearing aids
- Progress with hearing aids poorer than expected despite good audibility of speech.

Management issues

- AN/AD may require different management approaches
- Cochlear implantation
- Hearing aid issues
- Mode of communication
Hearing aid issues

- Use of hearing aids for children with AN/AD is controversial
- Potential damage to the outer hair cells
- Because of the dys-synchrony, hearing aids will produce a louder but still distorted signal
- No consensus on the HA fitting approach

Hearing aids may damage the outer hair cells

- Disappearance of the OAE is cited as proof of this damage
- No correlation has been established between hearing aid use and loss of emissions (Rance, 2005)
- Emissions may not be related to speech perception ability (Starr et al, 2000)
- Delays in HA fitting may contribute to limited outcome

Hearing aids only produce a louder but distorted signal

- The potential for improvement in signal clarity with hearing aids in ears with AN/AD is likely limited
- Some suggest that hearing aids should not be fit at all (Berlin, 1999)
- If good speech perception is possible, we won’t know if hearing aids work or not unless we try them.

Hearing aid fitting approach

- Some have recommended that HA should only be fit monaurally and should be low-gain aids even for those with severe or profound HL (Hood, 1998)
- More recent research has indicted that the best responses are obtained when the HA is set to the prescribed target for the degree of hearing loss (Rance, 2005)
- They did not benefit significantly from the aids set at lower gain or MPO levels

Our approach

- Hearing aid trial is required as part of the evaluation for a cochlear implant
- Parents are informed of the potential benefits and disadvantages of hearing aid fitting
- Recommend binaural fitting, unless parents have objection
- Hearing aids should be set at target levels

Management: communication approach

- Pre-implant: visual focus
  - Cued speech
  - Total Communication
- Post-implant
  - Auditory/oral is possible
  - Approach used must take individual differences into account
  - Age at implantation, associated medical/learning issues, parent goals
Candidacy level

- A = Auditory oral/verbal communicator
- B = Both: Auditory oral/verbal communicator with visual assist
- C = Complimentary: Primary visual communicator with auditory assist
- D = Quality of life benefits

McClatchie & Therres, 2003

Mode of communication: our patients post implantation

Review of management issues

- Fit binaural hearing aids ASAP
- Hearing aids should be set to target
- Pre-implant, child will likely benefit from visual communication approach
- If child is not making sufficient progress with hearing aids, consider CI
- Post-implant, synchrony is restored so AO is possible.

Summary

- “Cochlear implantation is currently the most successful remediation strategy for patients with poor sensitivity and speech understanding caused by auditory neuropathy/dys-synchrony” (Rance, 2005)
- Management of these children needs to take individual differences into account

Thank you

Questions?