Intern Seminar

Ri 陳盈曦 林珮璇 薛雅元
Supervised by HP 許權振
2009/7/29
• Auditory neuropathy: Clinical characteristics and therapeutic approach

• Cochlear implantation in children with auditory neuropathy: Outcomes and rationale
  Acta Oto-Laryngologica, 2007; Suppl 558: 36–43

• Predicting Cochlear Implant Outcomes in Children with Auditory Neuropathy

• Results of Cochlear Implantation in two children with mutations in the OTOF gene
Auditory Neuropathy

- Often misdiagnosed
- Also known as auditory dys-synchrony
Definition of AN/AD

• A neural hearing disorder:
  – Abnormal neural function characterized by absent or abnormal ABR.
  – Normal outer hair cell function: OAEs are present
  – Middle ear muscle reflexes are absent
Etiology/Location of Pathology

• Inner hair cells /IHC afferent synapse
• Kernicteric deposits anywhere from the spiral ganglion fibers to the brainstem
• Demyelinating diseases of VIIIth nerve and other peripheral nerve
Risk Factors

- Genetic
- Prematurity
- Histories of hyperbilirinemia
- .......
Audiological presentation

- Absent or abnormal ABR.
- Normal OAEs and/or CM.
- Absent middle ear muscle reflexes
- PTA can be very variable
- Fluctuating functional hearing loss: normal, mild, severe or profound.
Screening Pitfalls

• OAE:
  – Universal Newborn Hearing Screening
  – Normal $\rightarrow$ 10% Serious auditory synchrony problems
Diagnosis

• Preaudiometric triage
  – Tympanometry, Reflexes, OAEs
<table>
<thead>
<tr>
<th></th>
<th>Normal hearing</th>
<th>Auditory neuropathy</th>
<th>Conductive loss</th>
<th>Sensory loss</th>
<th>Nonorganic loss or central auditory disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Tympanometry Reflexes</strong></td>
<td>Normal</td>
<td>Normal</td>
<td>Abnormal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td>Present at 75–90 dB ipsilateral and contralateral</td>
<td>Absent</td>
<td>Elevated or absent</td>
<td>Usually present if loss does not exceed 75–80 dB</td>
<td>Present</td>
</tr>
<tr>
<td></td>
<td>Present</td>
<td>Present and often robust</td>
<td>Absent or diminished</td>
<td>Absent or diminished</td>
<td>Normal</td>
</tr>
<tr>
<td><strong>Emissions</strong></td>
<td>Present</td>
<td>Most often only a large ringing cochlear</td>
<td>Normal, by bone conduction; delayed, by air conduction</td>
<td>Robust responses with very steep latency-intensity function until threshold is reached. Then response quickly disappears</td>
<td>Normal</td>
</tr>
<tr>
<td><strong>Auditory brain stem response to ± clicks if used</strong></td>
<td>Follows latency-intensity curve, cochlear microphonic at the beginning is seen to invert</td>
<td>with no increase in latency when intensity decreases</td>
<td></td>
<td>Normal latency-intensity function, just as in normal-hearing patients</td>
<td></td>
</tr>
</tbody>
</table>
• GA: 31 weeks
  - Tympanometry: normal
  - Middle ear Muscle reflexes(-)
  - TEOAE (Lt): (+)

• Long ringing CM
• Reversed polarity
Management

• Spontaneous recovery or no obvious symptoms of an absent ABR
• Amplification
• Language learning and acquisition via oral, auditory, or visual means
• Cochlear implantation
Summary

• Auditory neuropathy:
  – Normal OAEs and CM
  – Absent or grossly abnormal ABR
  – Absent middle ear muscle reflexes

• OAEs or ABR alone → 10% error

• Tympanometry, Reflexes, OAEs
Joint Committee on Infant Hearing 2007

- AN included in the target group identified in NICU

- Hearing-screening and -rescreening protocols
  - NICU infants admitted > 5 days → include ABR
  - All infants readmitted within 1 month old, when there are conditions associated with potential hearing loss (e.g., hyperbilirubinemia that requires exchange transfusion or culture-positive sepsis), repeat hearing screening is recommended before discharge
Acoustic Simulations of Auditory Neuropathy

Original waveform
Mild
moderate
Severe
Profound neuropathy
Thank You!