Identification of ANSD in Neonates: From the Newborn Hearing Screening Perspective

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Speculate on (not necessarily answer) the following questions:

• What is the prevalence of ANSD in newborns?
• Is *transient* ANSD worth talking about?
• How to provide a prognosis for a newborn identified with ANSD?
• To screen or not to screen? Whom to screen?
What is the prevalence of ANSD in newborns?
## Prevalence in at-risk population

<table>
<thead>
<tr>
<th>Study</th>
<th>Population</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stein et al 1996</td>
<td>special care nursery</td>
<td>4.00</td>
</tr>
<tr>
<td>Psarommmatis et al 1997</td>
<td>intensive care unit</td>
<td>1.96</td>
</tr>
<tr>
<td>Rance et al 1999</td>
<td>“at-risk” infants</td>
<td>0.23</td>
</tr>
<tr>
<td>NHSP Evaluation 2004</td>
<td>babies in NICU for ≥48 h</td>
<td>0.2</td>
</tr>
</tbody>
</table>
aetiology in at-risk population

• prematurity and/or low birth weight
• hyperbilirubinaemia
• anoxia/hypoxia
• …
prevalence in well-baby population

Low???
1:200,000 (Australian unpublished data 2005)

…

But:
1:5,700 (Owen et al 2008)

And:
prevalence in well-baby population  
*(Sininger & Oba 2001)*

Table 2–1. Patients with onset of auditory neuropathy before age 2 years, grouped by family history and other neonatal risk factors.

<table>
<thead>
<tr>
<th>Risk Factors</th>
<th>Family or Genetic History</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Yes</td>
</tr>
<tr>
<td>Hyperbilirubinemia</td>
<td>2</td>
</tr>
<tr>
<td>Prematurity</td>
<td>1</td>
</tr>
<tr>
<td>Multiple risk factors</td>
<td>0</td>
</tr>
<tr>
<td>No other risk factors</td>
<td>8</td>
</tr>
<tr>
<td>Total</td>
<td>11</td>
</tr>
</tbody>
</table>
aetiology in well-baby population

- heredity:
  - autosomal recessive isolated:
    - otoferlin (Varga et al 2003)
    - pejvakin (Delmaghani et al 2006)
  - syndromes e.g. Waardenburg (Pau et al 2006)

- cochlear nerve aplasia/agenesis (Buchman et al 2006)

- tumor or cyst (Boudewyns et al 2008)
prevalence in PCHL population

<table>
<thead>
<tr>
<th>Study</th>
<th>Population</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vohr et al 2001</td>
<td>Universal screening</td>
<td>1.80</td>
</tr>
<tr>
<td>Berlin et al 2000</td>
<td>1000 HI infants</td>
<td>8.70</td>
</tr>
<tr>
<td>Kraus et al 1984</td>
<td>48 HI infants</td>
<td>14.58</td>
</tr>
<tr>
<td>NHSP Evaluation 2004</td>
<td>169 HI infants</td>
<td>10.1</td>
</tr>
</tbody>
</table>
Is transient ANSD worth talking about?
• ABRs have been reported to recover (or improve)

• ABR recovery (or improvement) may happen by up to as late as **two years of age** (Madden *et al* 2002)

• perceptual ability may improve even when ABR remains abnormal
prevalence of transient ANSD

- 24% in our pilot data
- 65% Psarommatis et al 2006
transient ANSD

- the reported aetiological/risk factors:
  - hydrocephalus (Russell et al 2001)
  - anoxia (Attias et al 1990, 2007)
  - metabolic toxic and/or inflammatory factors (Alexander et al 1995)
  - genetic factors
    - familial isolated delay of auditory maturation (Neault & Kenna 2004)
    - syndrome such as maple syrup urine disease (Spankovich et al 2007)
    - coexisting alongside delayed visual maturation in the absence of any known risk indicators has been described (Aldosari et al 2003)
transient ANSD

- Changes in myelination
- Changes in synaptic efficiency
- Other???
How to provide a prognosis for a newborn identified with ANSD?
At birth: Normal OAEs, Absent ABR

Prognosis???

Normal auditory function

Total lack of sound awareness

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At birth:
Normal OAEs
Absent ABR

by 2 yrs

Normal ABR

by 2 yrs

Absent ABR

by 2 yrs

Normal hearing

Normal hearing
tresholds

Elevated hearing thresholds

Normal speech perception

Speech perception worse than expected from audiogram

Speech perception worse than expected from audiogram

Speech perception matches audiogram
predicting prognosis

- attention to global development of the child
- better understanding of aetiopathology and risk factors associated with ANSD may help
- multidisciplinary approach
To screen or not to screen? That is NOT the question
screening principles (Wilson & Jung, 1968)

1. the condition should be an important health problem in the society concerned;
2. there should be an accepted and effective treatment for the cases identified;
3. facilities for assessment and treatment should be available;
4. there should be a recognisable latent or early symptomatic stage;
5. there should exist a simple predictive test suitable for screening;
screening principles (Wilson & Jung, 1968)

6. the test should be acceptable to the population;
7. the natural history of the condition should be understood;
8. there should be an agreed policy on whom to treat as patients;
9. the cost of case-finding (incl. further assessment and treatment) should be non-wastefully balanced in relation to possible expenditure on medical care as a whole;
10. case-finding should be a continuing process and not a ‘once and for all’ project.
role of professionals

• change our frame of mind
• learn to communicate uncertainty
role of families

• qualitative study (funded by National Deaf Children’s Society) explores parents’ experiences with an objective to use parents as experts
Thank you!

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