

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



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aim of the talk

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?

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prevalence in **at-risk** population

Study	Population	%
Stein <i>et al</i> 1996	special care nursery	4.00
Psarommatis <i>et al</i> 1997	intensive care unit	1.96
Rance <i>et al</i> 1999	“at-risk” infants	0.23
NHSP Evaluation 2004	babies in NICU for ≥ 48 h	0.2

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1:200,000 (Australian unpublished data 2005)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.

Risk Factors	Family or Genetic History		Total
	Yes	No	
Hyperbillirubinemia	2	1	3
Prematurity	1	1	2
Multiple risk factors	0	7	7
No other risk factors	8	5	13
Total	11	14	25

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

Study	Population	%
Vohr et al 2001	Universal screening	1.80
Berlin et al 2000	1000 HI infants	8.70
Kraus et al 1984	48 HI infants	14.58
NHSP Evaluation 2004	169 HI infants	10.1

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Is transient ANSD worth talking about?

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transient ANSD

- 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:
 - low birth weight (Psarommatis *et al* 2006, Attias *et al* 2007)
 - hyperbilirubinaemia (Lafreniere *et al* 1993, Madden *et al* 2002, Attias *et al* 2007)
 - 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?

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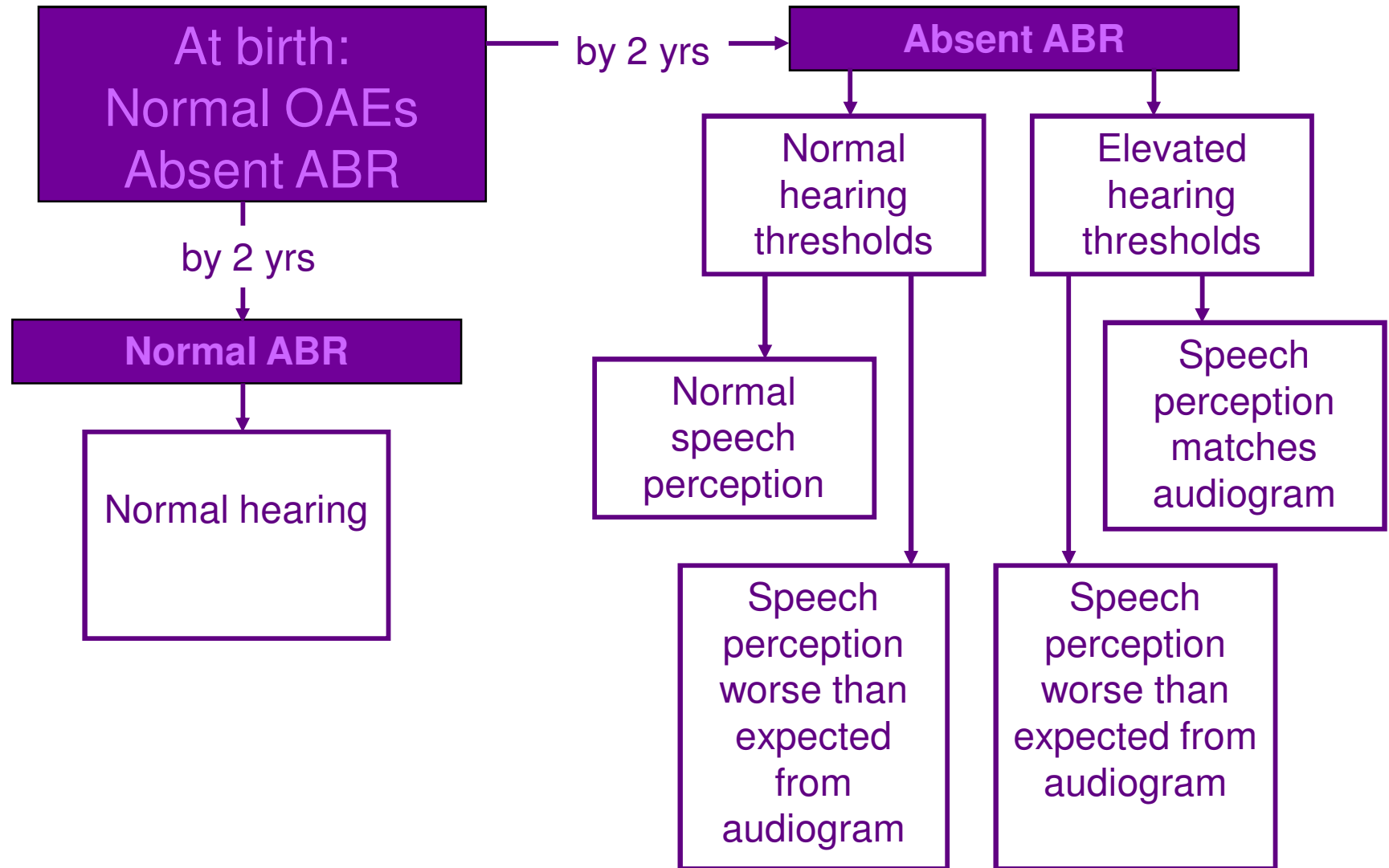
At birth:
Normal OAEs
Absent ABR

Prognosis???

Normal
auditory
function

Total lack
of sound
awareness

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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***

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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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