Auditory Neuropathy Spectrum Disorder ANSD in children

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Aims

- To promote understanding of the condition known as **Auditory Neuropathy Spectrum Disorder**

Definition in young infants

- **Condition is characterised by:**

– ABR absent or severely abnormal at high levels

– OAEs present and/or CM present

- **Normal cochlear function at the level of OHCs but dys-synchronous auditory brainstem responses**

Definition in older children and adults (old definition)

- **Speech perception impaired beyond what would be expected for the degree of hearing threshold**
- **A trial of personal amplification which is not of benefit for improved speech comprehension**

relative to expected degree of hearing threshold

Terminology

- Auditory neuropathy
- Auditory dys-synchrony
- Auditory de-synchrony
- Auditory mismatch
- Primary auditory neuropathy

Anatomical Terminology

- Inner hair cell damage/loss
- Synaptic block
- **Neuronopathy (ganglionapathy)**
- **Neuropathy:**

– **demyelinating**
– **axonal**
– **mixed**

- Brainstem disorder

Prevalence in **Adult** population

- Likely to be prevalent in certain patient groups e.g. conditions affecting nervous system:

- Hereditary

- Charcot-Marie-Tooth disease
- Friedreich's ataxia

- Acquired

- systemic diseases (e.g. diabetes mellitus) infections and autoimmune disorders (e.g. HIV)

Prevalence in **At-risk** population

Population	%
special care nursery	4.00
intensive care unit	1.96
"at-risk" infants	0.23
babies in NICU for ≥48 h	**0.2**

Aetiology in **At-risk** population

- Prematurity
- Low birth weight
- Hyperbilirubinaemia
- Anoxia/hypoxia

Prematurity

- **Kernicterus** often occurs at lower bilirubin concentrations in premature newborns as compared with term newborns.
- Higher sensitivity to **hypoxic-ischemic damage** has been observed in premature infants

Low birth weight

- Neuromaturational delay

  less than 3 pounds or 2,5 kg

Hyperbilirubinaemia

- The most common aetiological factor in neonates
- Bilirubin is by-product of red blood cell breakdown
- Normally broken down by liver and excreted by kidneys
- Up to 60 percent of term newborns have clinical jaundice in the first week of life
- Results in yellow pallor to skin and eyes (jaundice)
- Treated with phototherapy & exchange transfusions

Hyperbilirubinaemia

- **Where does bilirubin damage the auditory system?**

– inner ear: **NO**
– spiral ganglion and auditory nerve: **YES**
– brainstem auditory nuclei: **YES**
– thalamus and auditory cortex: **NO**

Anoxia/Hypoxia

- Chronic mild hypoxia selective **inner hair cell** loss:

Prevalence in **Well-baby** population

- Low???
- 1:500,000 (Mehl 2002)
- 1:200,000 (Australian unpublished data 2005)
- But:
- 1:5,700 (Owen et al 2008)
- .09:1000 (Boudewyns A et al 2016).

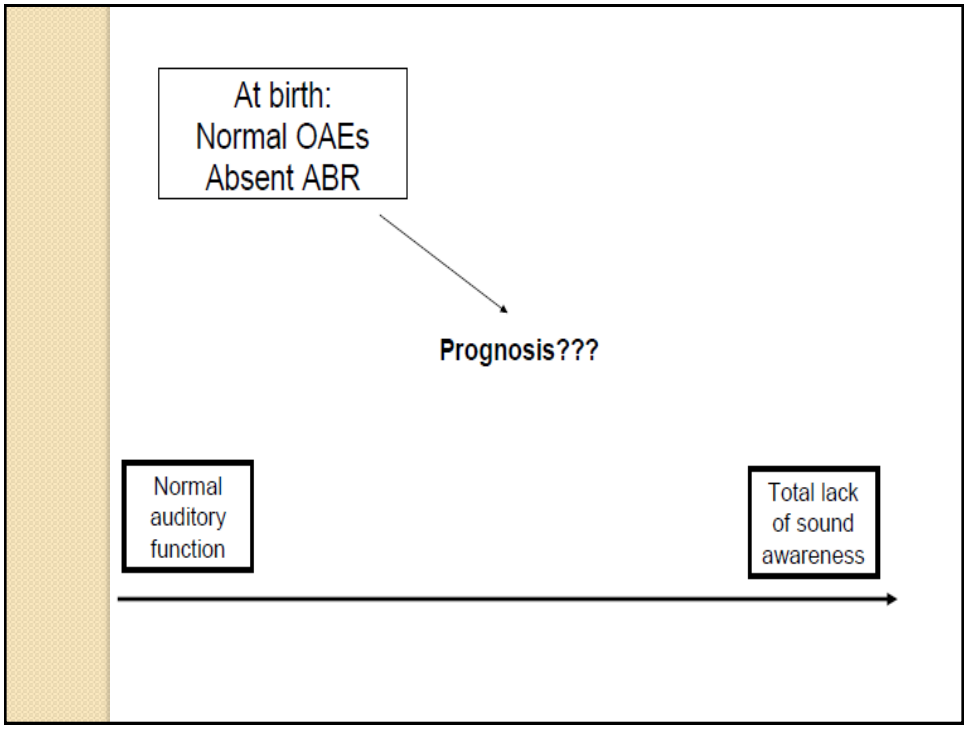
Aetiology in **Well-baby population**

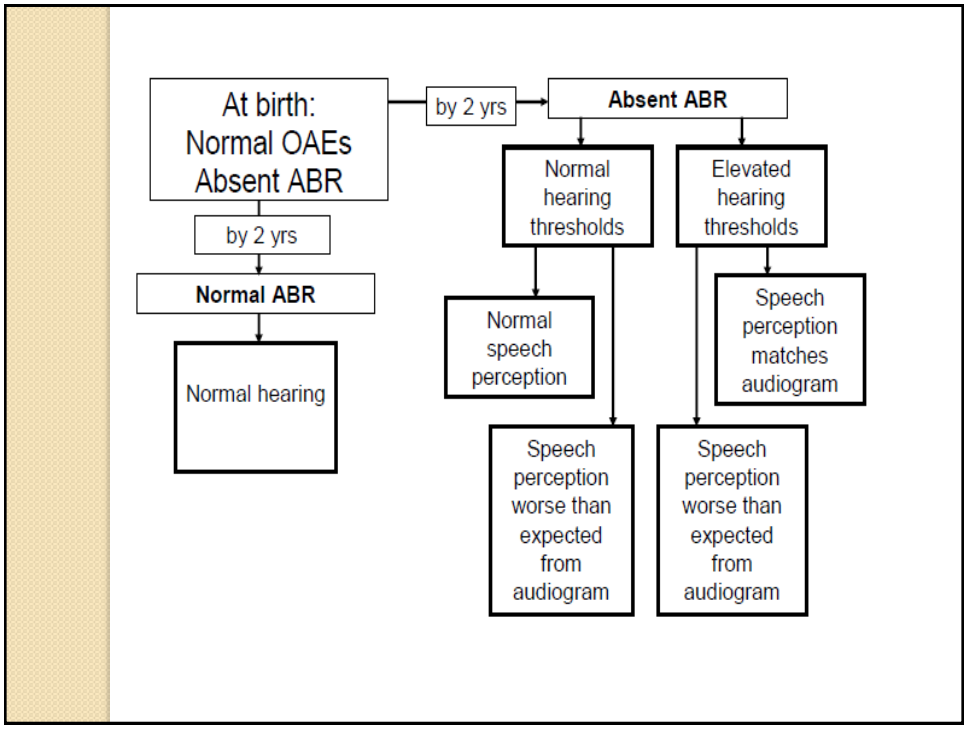
- Heredity:
  - autosomal recessive isolated
  - syndromes e.g. Waardenburg
- Cochlear nerve deficiency
  - developmental aplasia
- tumor or cyst

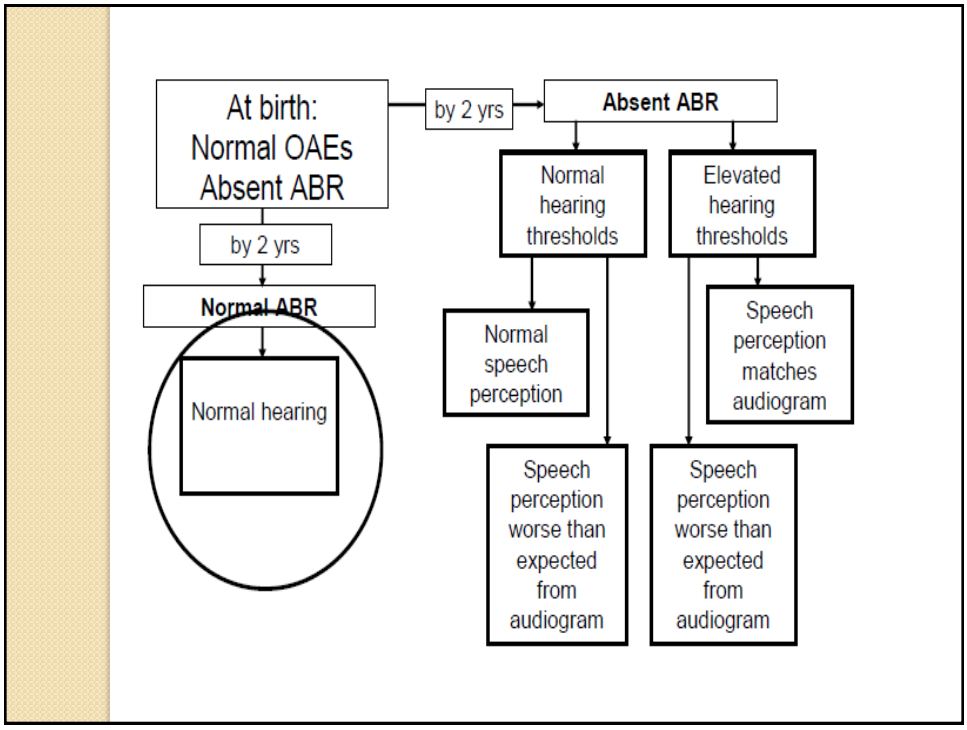
Autosomal recessive isolated ANSD

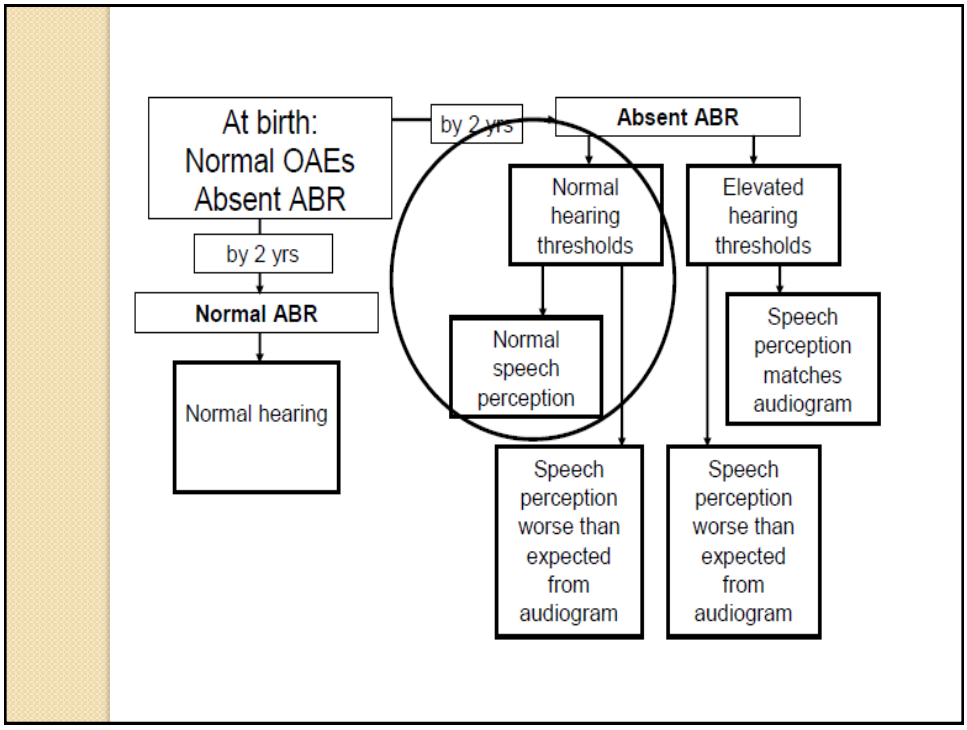
- Primary lesion **at the level of the inner hair cells (IHC), the IHC synapse to the afferent nerve fibers**
- **Or**
- Primary lesion **neurons in the spiral ganglion and the brainstem auditory nuclei**

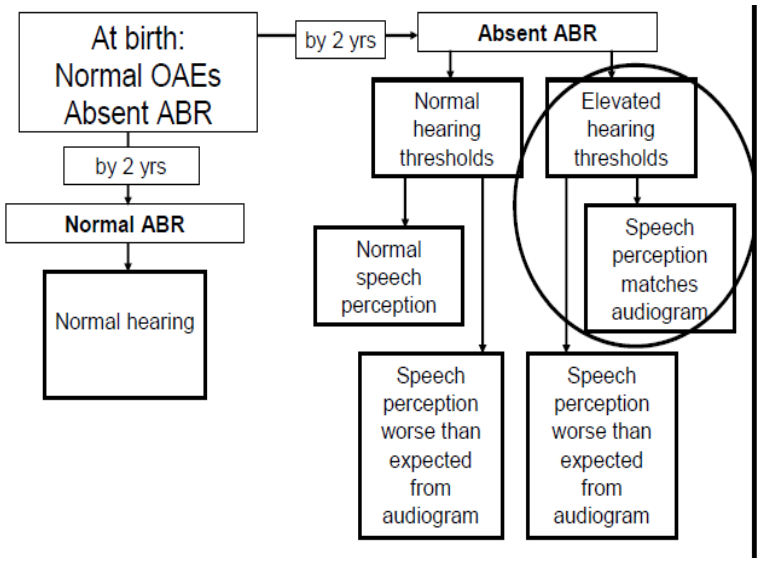
PROGNOSIS


At birth:
Normal OAEs
Absent ABR
Prognosis???
Normal auditory function
Total lack of sound awareness


At birth:
Normal OAEs
Absent ABR
by 2 yrs
Normal ABR
Normal hearing
by 2 yrs
Absent ABR
Normal hearing thresholds
Elevated hearing thresholds
Normal speech perception
Speech perception matches audiogram
Speech perception worse than expected from audiogram
Speech perception worse than expected from audiogram







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

- Sometimes transient:
  - 65% Psarommatis et al 2006

- Traditionally called maturational, however…

Transient ANSD

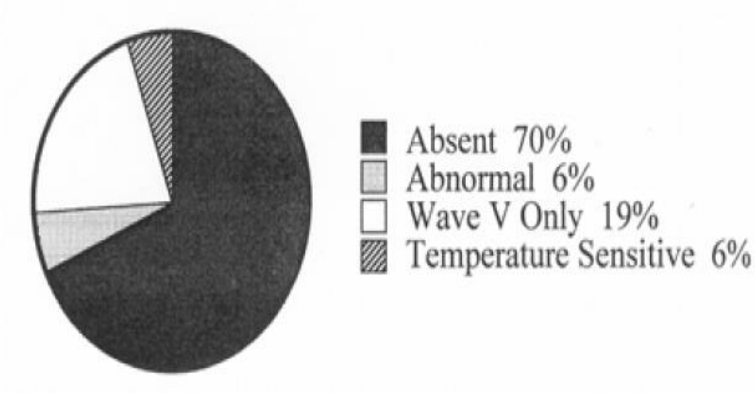
- The reported aetiological/risk factors:
  - low birth weight
  - hyperbilirubinaemia
  - anoxia
  - genetic factors

Neuromaturational Delay

- Changes in myelination
- Changes in synaptic efficiency

AUDIOLOGICAL PROFILE

ABR



ABR criteria

1) flat ABR with no evidence of peaks
2) presence of early peaks (waves up to III)
3) some poorly synchronised but evident later peaks (wave V)

OAEs

- In 30% OAEs may disappear, but CM persists
- Disappearance of OAEs does not appear to be linked to amplification

OAEs and/or CM

- **OAEs** may deteriorate, but **CM** persists:

– subtle middle ear pathology

– significant OHC loss; CM produced by IHC

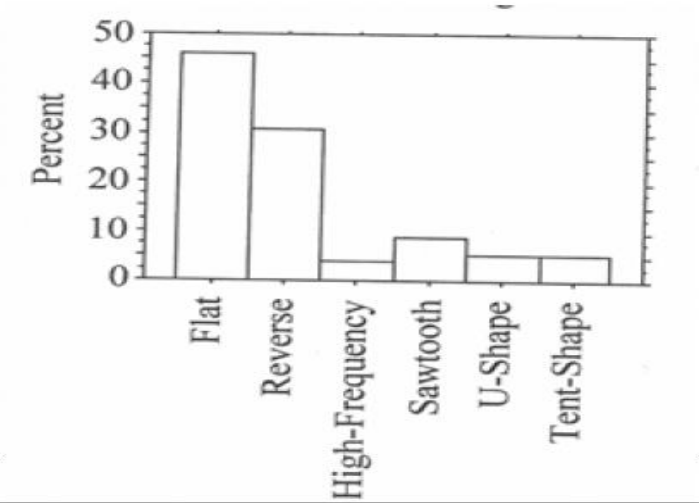
– OHC present, but impaired function: able to polarise and depolarise producing CM, but unable to generate the mechanical cochlear processes producing OAEs

CM

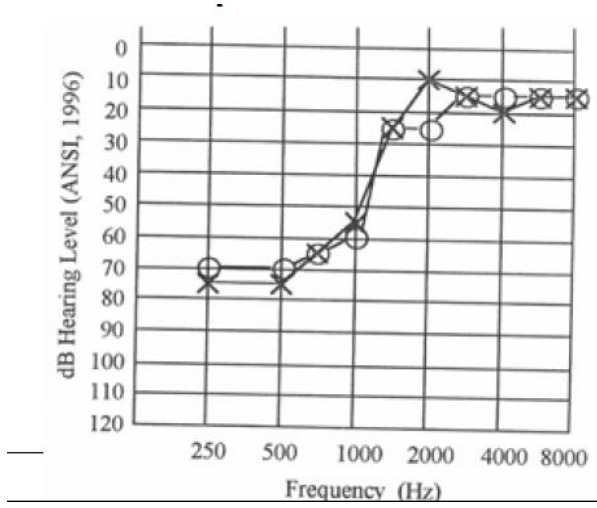
- **Displacement of basilar membrane in response to stimulus**
- **Results in a stimulus-related potential called the cochlear microphonic**
- **Features:**

– pre-neural response from cochlea
– shape follows stimulus polarity
– does not change in latency (1-2ms)
– cannot be used to estimate threshold

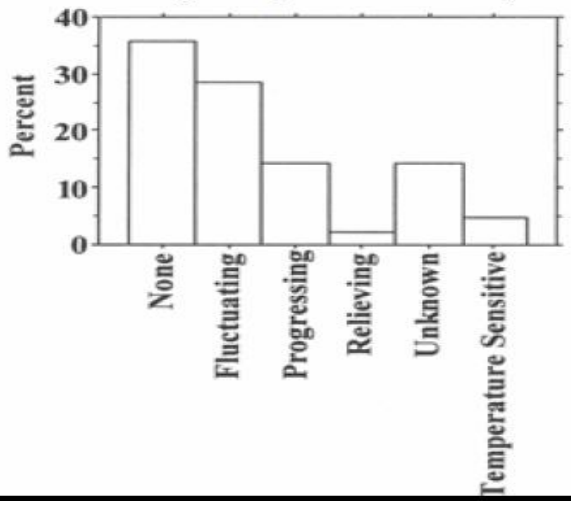
Audiometric Configuration



Example of low frequency hearing impairment

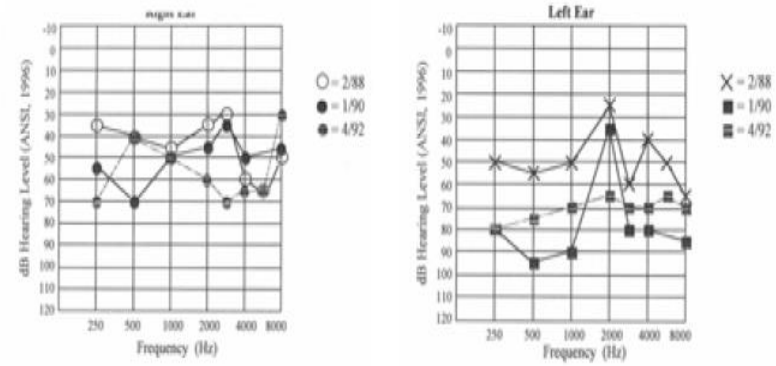


Changes in thresholds over time



ANSD Audiometry: flat \ reverse \ fluctuating..... low frequency

Example of fluctuating hearing impairment



Diagnostic Issues

- Differentiate from HF SNHL
- ABR results **do not** predict behavioural thresholds
- Behavioural thresholds **do not** predict speech perception
- Monitoring and follow-up:

– repeat ABR, CM and OAEs at 8-10 weeks and 9-15 months

– tympanometry and stapedial reflexes when possible

– vestibular testing if possible

- Multidisciplinary approach

MANAGEMENT

management

- The only absolute rule:
  – *First, not to harm!*
- Varied and very controversial
- *'the earlier the better'* rule does not necessarily apply
- Attention to child's development (especially communication development)

multidisciplinary management

- Family
- Teacher of the deaf
- Audiologist
- Speech and language therapist
- ENT
- Paediatrician
- Neurologist
- …

Monitoring

- Monitoring child's development:

– global development with a special emphasis on communication development

– repeat the audiological tests

– other needs and additional medical conditions need to be identified and considered

Medical treatment

- Surgical (e.g. tumour, cyst, hydrocephalus)
- Medication (e.g. corticosteroid therapy in demyelinating conditions)

Amplification

– NOT BEFORE BEHAVIOURAL THRESHOLDS

– have not been successful in adult population

– potential risk of noise-induced hearing loss

– under-amplification also detrimental

Amplification

- – 50% children show auditory perceptual
- skills consistent with their SN peers (Rance et al 2002, 2004)

FM systems

- Either alone or with hearing aids

– because of the severe breakdown of speech perception particularly in noisy situations

Cochlear Implantation

– very beneficial in some children with ANSD

– do not allow normal-mild-moderate audiogram be a contraindication

– **however, be aware of the possibility of spontaneous recovery up to 2 years**

– success may be dependent on *site of lesion*

– family education and expectation management

Brainstem Implantation

– suitable where site of lesion more central

– suitable in auditory nerve insufficiency (Buchman et al 2006)

Visual communication

– sign language

Additional needs

– as compared with children with SNHL much larger proportion of children with ANSD have additional needs which are crucial in choosing intervention strategies

family-friendliness!!!

- Do not forget that behind this very puzzling condition is a child and a family
- Be honest with the family: tell them what we know and also what we don't know
- Written information to the family and as well as to other key professionals involved with the family

Thank you