



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** relative to expected degree of hearing threshold
- **A trial of personal amplification which is not of benefit for improved speech comprehension**

Terminology

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

Anatomical Terminology

- Inner hair cell damage/loss
- Synaptic block
- **Neuronopathy (ganglionopathy)**
- **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).

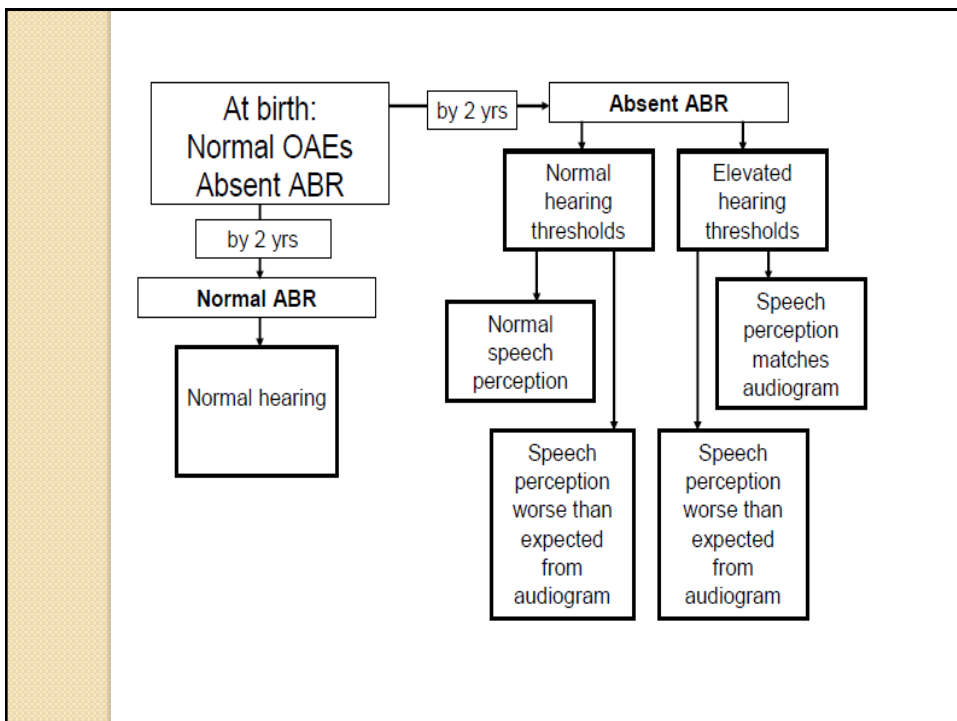
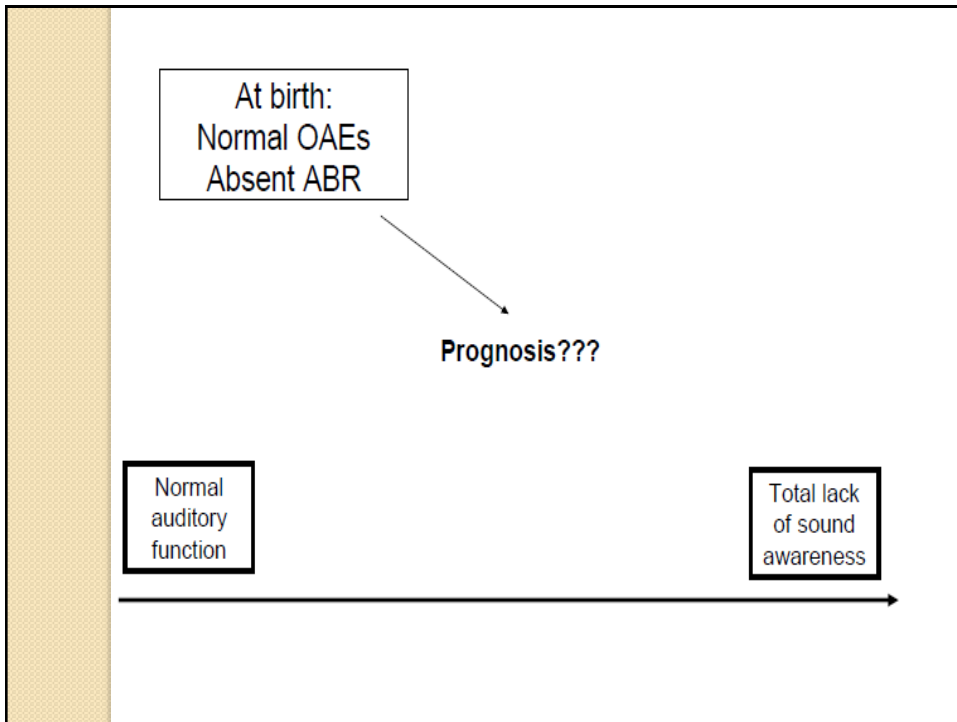
Aetiology in **Well-baby** population

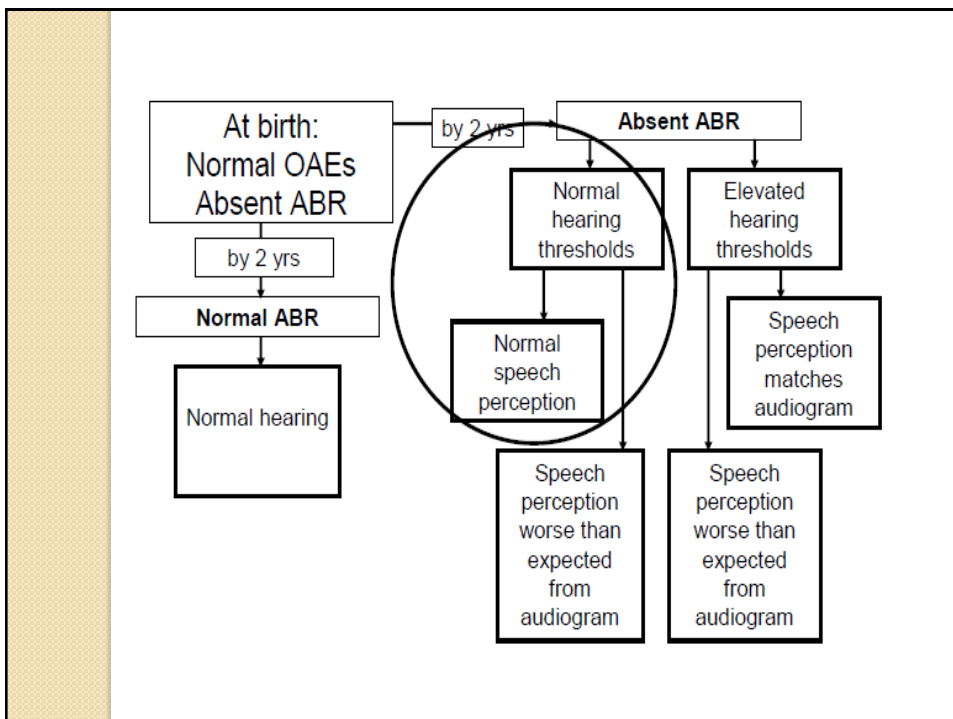
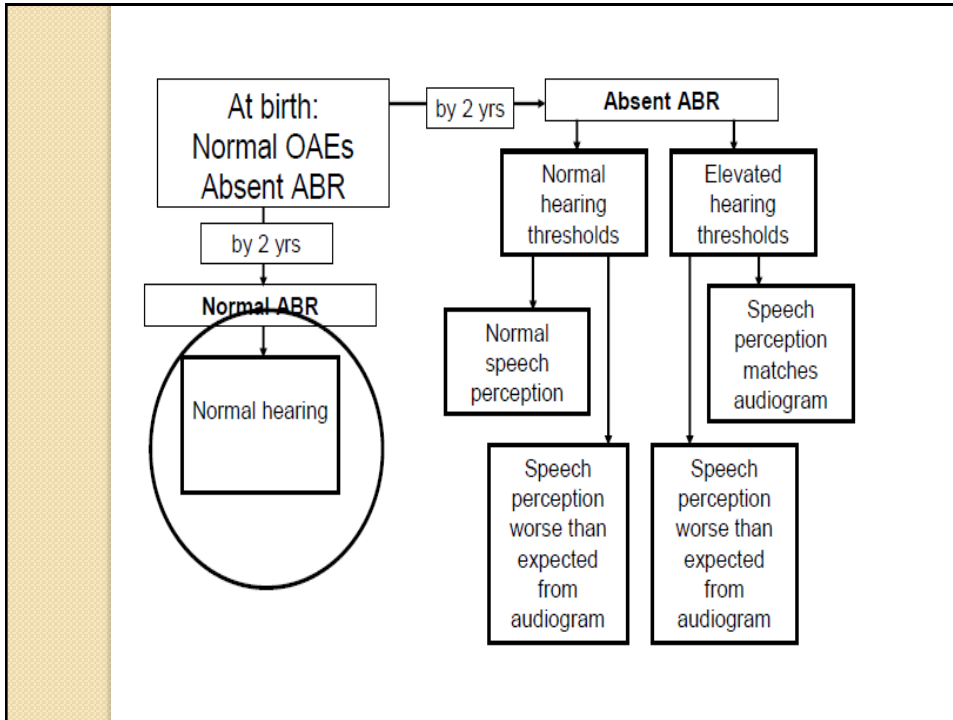
- Heredity:
 - autosomal recessive isolated
 - syndromes e.g. Waardenburg
- Cochlear nerve deficiency
 - developmental aplasia
- tumor or cyst

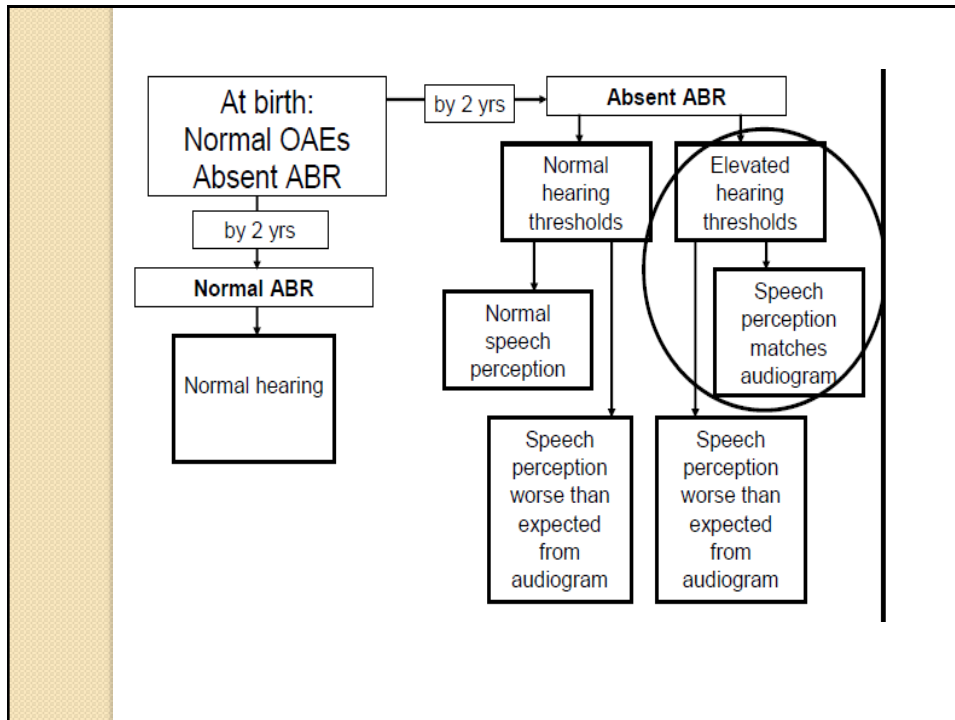
Autosomal recessive isolated ANSD

- 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







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

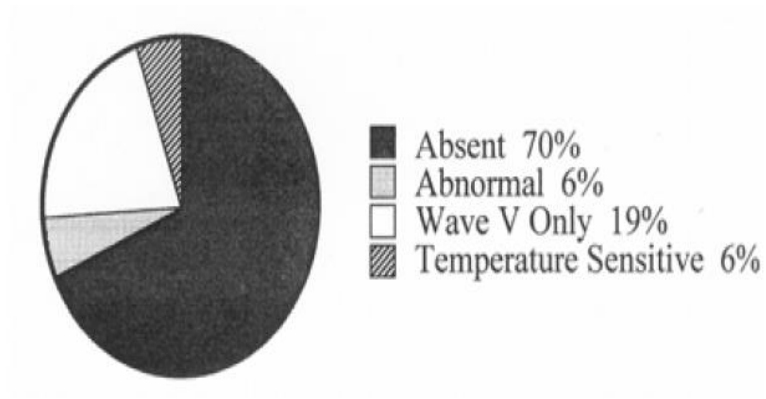
- The reported aetiological/risk factors:
 - low birth weight
 - hyperbilirubinaemia
 - anoxia
 - genetic factors

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

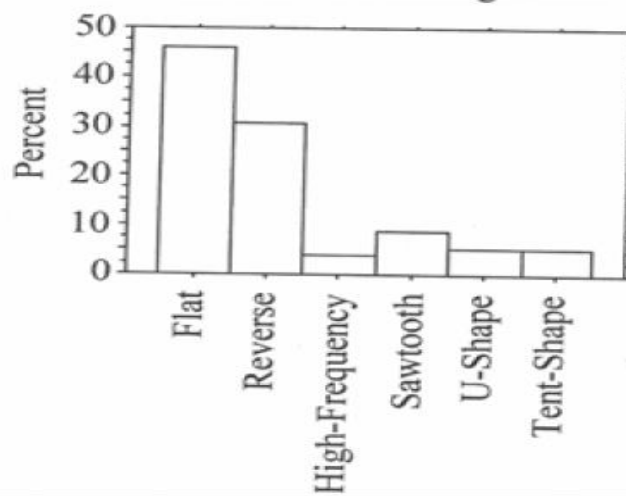
OAEs and/or CM

- **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

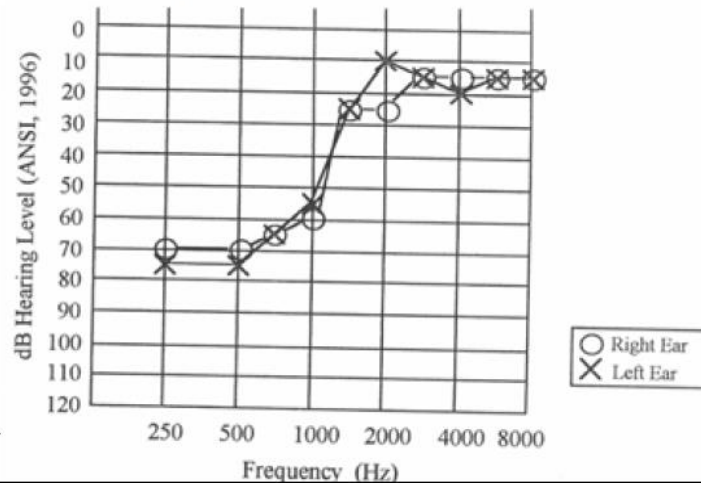
CM

- **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

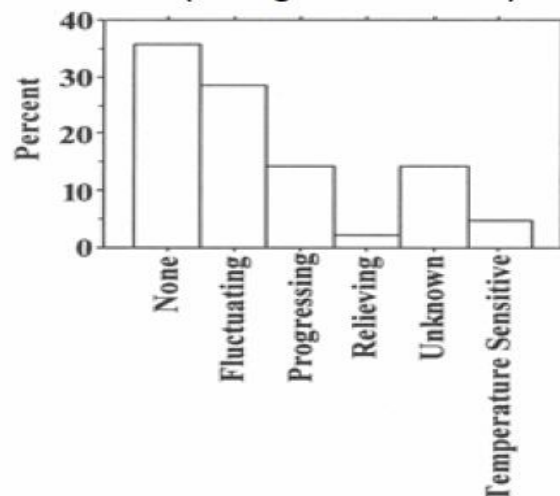
Audiometric Configuration



Example of low frequency hearing impairment

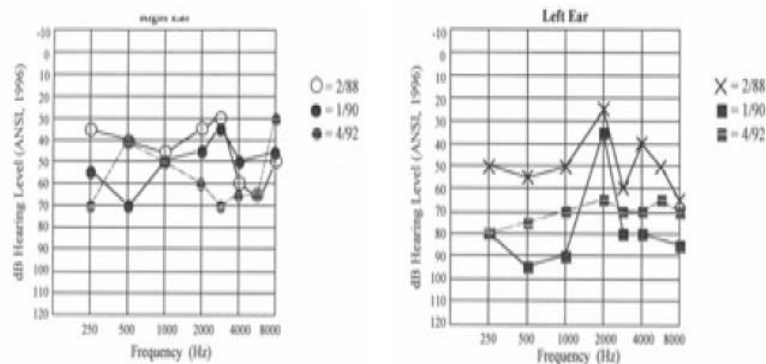


Changes in thresholds over time



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