Diagnosis and Management of ANSD: Outcomes of Cochlear Implants versus Hearing Aids

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Auditory Neuropathy Spectrum Disorder (ANSD)

- Also referred to as: auditory neuropathy, auditory dys-synchrony, auditory synaptopathy…

- Hearing impairment in which cochlear outer hair cell function is “normal” but afferent neural transmission is disordered

- Indicated by the presence of pre-neural responses (OAE / CM) with absent or severely disrupted auditory neural responses (ABR)
CM/ABR tracings for a 3mo old with ANSD (1989)
Possible mechanisms producing the ANSD result pattern

- Cochlear damage restricted to the inner hair cells (IHC)
- IHC/auditory nerve synapse
- Auditory nerve abnormality
  - reduced neuronal population
  - disruption of neural synchrony
  - cochlear nerve deficiency
  - tumour
Paediatric ANSD

◆ Congenital/Perinatal
  » anoxia
  » hyperbilirubinaemia

◆ Progressive
  – Neurodegenerative disease
    » Onset physical symptoms usually in adolescence
    » Identified earlier (routinely see 1-4 yr olds in clinic)
    » Hearing difficulties often the first presenting symptom
ANSD Clinical Profile

◆ Prevalence

– Congenital/Perinatal ANSD
  » 1 in 800-1000 children show permanent hearing loss
  » 5-15% of those present with the ANSD result pattern

– Neurodegenerative disease
  » List of diseases associated with ANSD growing
    ◆ FRDA/CMT/LHON/ADOA...
  » relatively rare
  » Friedreich ataxia most common: ≈ 1 in 20,000
ANSD Clinical Profile

- **Behavioural audiogram**
  - Level: normal hearing to profound loss
  - All configurations: \( \approx 30\% \) low frequency
  - Fluctuating hearing

- **Acoustic reflexes**
  - Typically absent (regardless of hearing level)

- **Functional hearing**
  - Impaired speech perception
Speech Perception

- Consistently reported problem in both adults and children with ANSD

- Difficulties out of proportion with the behavioural audiogram
  - Abnormal speech perception in subjects with “normal hearing”
  - Subjects with elevated hearing thresholds show speech perception poorer than for SNHL of equivalent degree
Open-set Speech Perception v Hg Level for Children & Adults with ANSD

Yellin et al., 1989
Open-set Speech Perception v Hg Level for Children & Adults with ANSD

- Word score
- Sentence score

3-Frequency Average (dBHL) vs. Open-Set Speech Score (%)
Why is speech perception often poorer than expected?

◆ Signal distortion
◆ Timing of neural conduction disrupted
◆ Impaired perception of temporal cues in speech
  – Inability to judge vowel duration
    » eg. hid vs heed
  – Inability to discriminate consonants based on timing cues
    » eg. pin vs bin
      tin vs din
◆ Different to sensorineural hearing loss
  – Temporal processing typically normal
Speech Perception in Noise

- Extreme difficulty reported in adults and children with ANSD (Kraus et al., 2000; Rance et al., 2007; 2010; 2012; Starr et al., 1998)

- Some cases show normal understanding in quiet and negligible perception in “everyday” listening conditions
Speech Perception in Noise for Children with ANSD
(normal sound detection)

CNC Phoneme Score (%) vs. S/N Ratio (dB)

Rance et al., 2007; 2010, 2012
Speech Perception in Noise for Children with ANSD
(normal sound detection)

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Speech Perception in Noise for Children with ANSD
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CNC Phoneme Score (%)

S/N Ratio (dB)

Rance et al, 2007; 2010, 2012
Clinical Management of ANSD

- Children with ANSD form a heterogeneous group
  - Range of different aetiologies
  - Different clinical presentations

- Neural distortion (to varying degrees)
  - Perceptually quite different to children with SNHL

- Standard management approaches may not apply
Management of Children with ANSD

Hearing Aids vs Cochlear Implants
Conventional Amplification

- **Arguments against amplification**
  - Inherent pathway limitations
  - Potential for cochlear damage

- **Argument for amplification**
  - Increased access to the speech signal (if sufficient gain is provided)

- **Speech perception outcomes**
  - 40-50% show significant benefit
Cochlear Implantation

– Currently the option of choice for most individuals with ANSD

– Speech Perception Outcomes

» Most reported cases have performed at levels similar to peers with SNHL

» Some poor results

» Teagle et al. (2010)

» 52 children with open-set scores

» 27% of cases showed speech perception scores <30%
ANSD Management: Hearing Aids / Cochlear Implants

◆ Speech Perception (Rance & Barker, 2008)

◆ Subjects
  – Aided ANSD children (N=10)
  – Implanted ANSD children (N=10)
  – Implanted SNHL children (N=10)

◆ Assessment
  – Open-set speech perception (CNC-Words)
  – Free-field presentation
  – Normal listening condition
Open Set Speech Perception

CNC Phoneme Score (%)

Age at Device Fitting (mo.)

- AN/AD (Aided)
Open Set Speech Perception

CNC Phoneme Score (%)

Age at Device Fitting (mo.)

- AN/AD (Aided)
- SN (CI)
- AN/AD (CI)
ANSD Management: Hearing Aids / Cochlear Implants

◆ Melbourne Long-Term Outcome Study
  – Infant ANSD first identified in Melb (1989)
  – Tracking these individuals from infancy to adulthood

◆ Longitudinal data
  – Audiometry
  – Basic auditory perception (temporal/frequency processing)
  – Speech perception (quiet/noise)
  – Hearing disability ratings
  – Expressive/receptive language development
Long-term Language Development in ANSD

◆ Receptive Language
  – Peabody Picture Vocabulary Test (PPVT)
  – determines an “equivalent language age” based on norms for normally hearing/developing children

◆ Longitudinal data: (4 yrs – 20 yrs)

◆ Subjects (April 2014)
  – Aided ANSD children (n=8)
  – Implanted ANSD children (n=6)
  – Implanted SNHL children (n=12)
Receptive Language (PPVT)

Equiv. Language Age (yrs)

Age at Assessment (yrs)
Receptive Language (PPVT)
Receptive Language (PPVT)

![Graph showing the relationship between Age at Assessment and Equiv. Language Age. The graph includes data points for ANSD (CI).]
Receptive Language (PPVT)
Receptive Language (PPVT)
Conclusions

Most implanted children with ANSD show speech perception and language outcomes equivalent to those of young implantees with SNHL.

Some children with ANSD managed with conventional hearing aids can perform as well as the average implantee.
Clinical Challenge

◆ How to predict whether a newly diagnosed baby will perform better with conventional hearing aids or CI?

◆ Considerations
  – Anatomy: if a child has no nerve then a CI will not be beneficial
  – Sound detection thresholds: if hg levels are in the severe/profound range the child is unlikely to benefit from amplification (same audiologic selection criteria as for SNHL)
  – Auditory capacity: perceptual ability in cases with mild/severe loss range determined by the degree of temporal distortion

◆ Current Research Objective
  – Measure auditory processing (in infancy) predict long-term outcomes
Summary

- 20+ years of experience with paediatric ANSD has led to significant advances
  - Understanding of mechanisms
  - General pattern of functional outcomes

- Results in individual children are highly variable and so the management of affected youngsters remains a challenge...
Thankyou