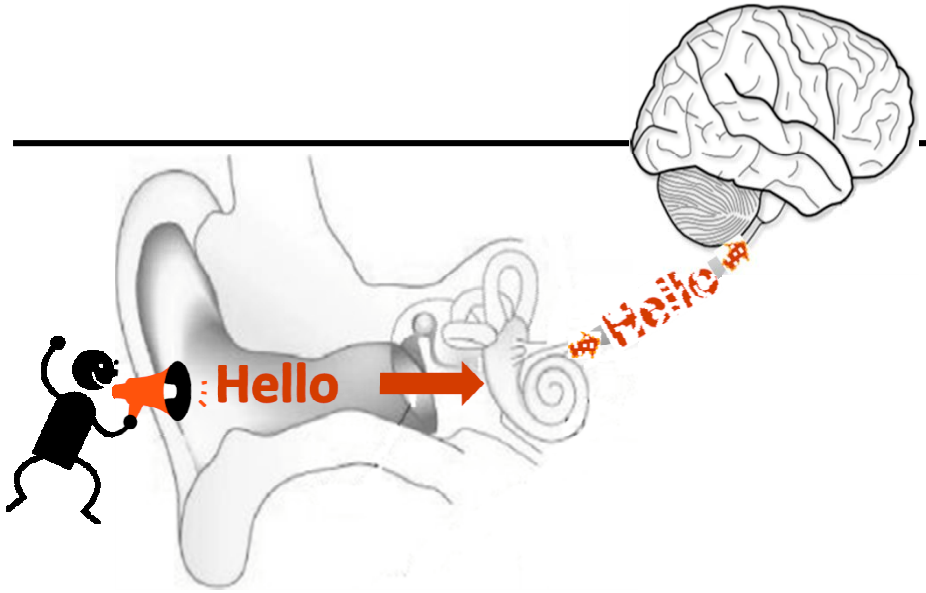


Impact of the presence of auditory neuropathy spectrum disorder on outcomes at 3 years of age

7th Australasian Newborn Hearing Screening Conference

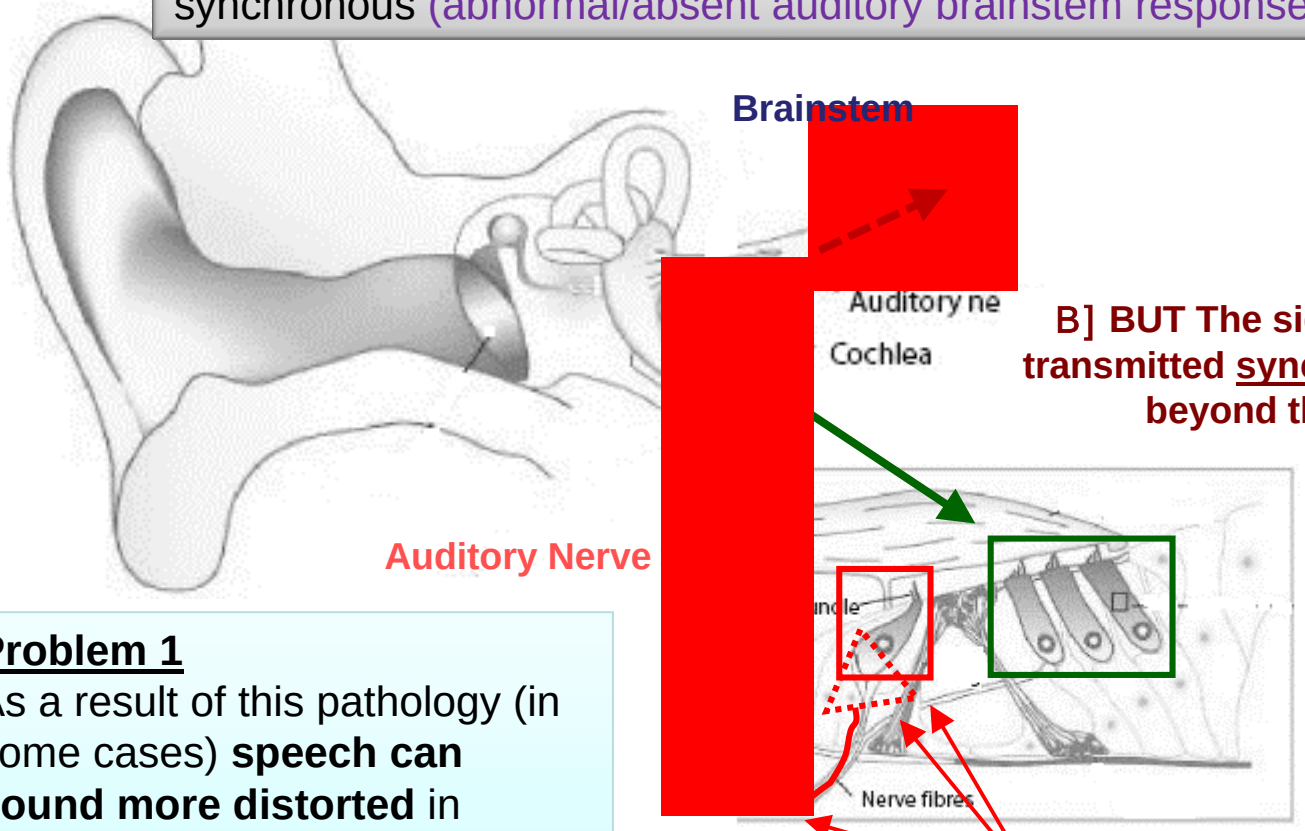
17th-18th May2013

Auckland, New Zealand

Ching TYC, Day J, Dillon H, Gardner-Berry K,
Hou S., Seeto M, Wong A, Zhang V

What is ANSD? in a nutshell……

ANSD is a type of hearing loss where there are signs that parts of the cochlear are still functioning (presence of OAEs and/or cochlear microphonic) but the neural signals along the auditory nerve & brainstem are not synchronous (abnormal/absent auditory brainstem response (ABR)



B] BUT The signal is not being transmitted <u>synchronously/clearly</u> beyond the cochlear

A] Parts of the cochlea are still working (<u>outer</u> hair cells)

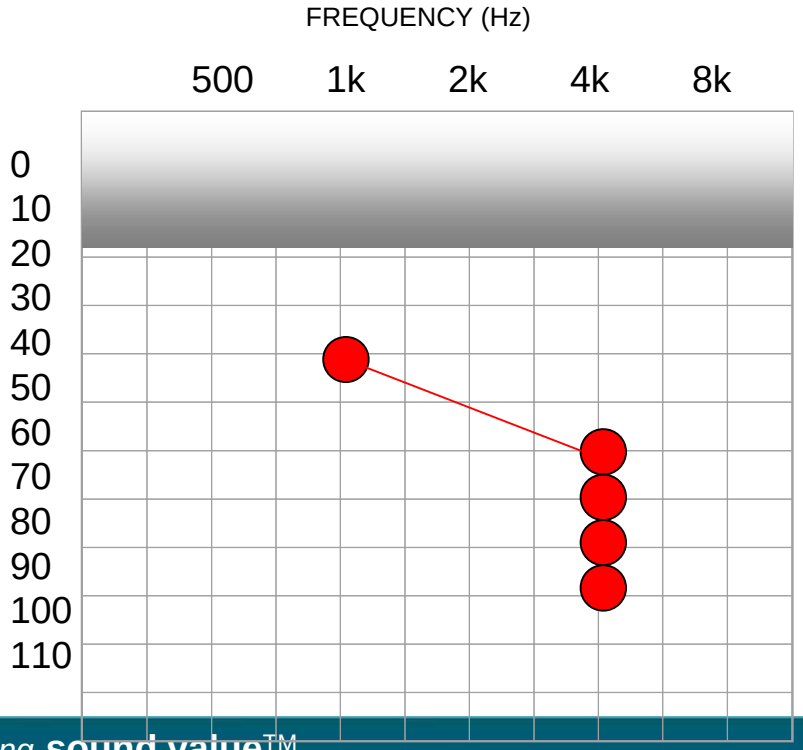
<u>Problem 1</u>

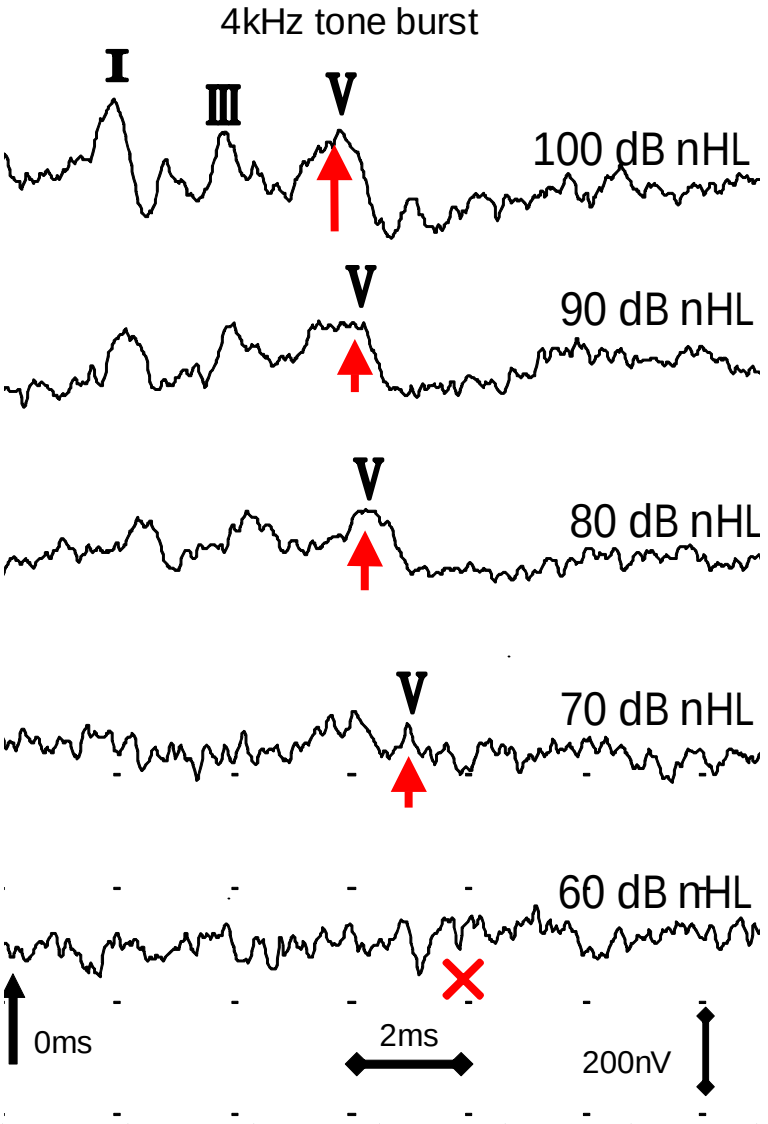
As a result of this pathology (in some cases) **speech can sound more distorted** in patients with ANSD than it would be with a typical sensorineural hearing loss (SNHL)

Could be a problem with:
- <u>Inner</u> hair cells
- <u>Synapse</u>
- <u>Auditory nerve</u>

Background:
- Estimating the audiogram with ABR - SNHL

- When we diagnose a young baby with a hearing loss we use Auditory Brainstem Response (ABR) testing and track wave V down to threshold.
- The wave V thresholds across different frequencies can then be used to predict an audiogram.



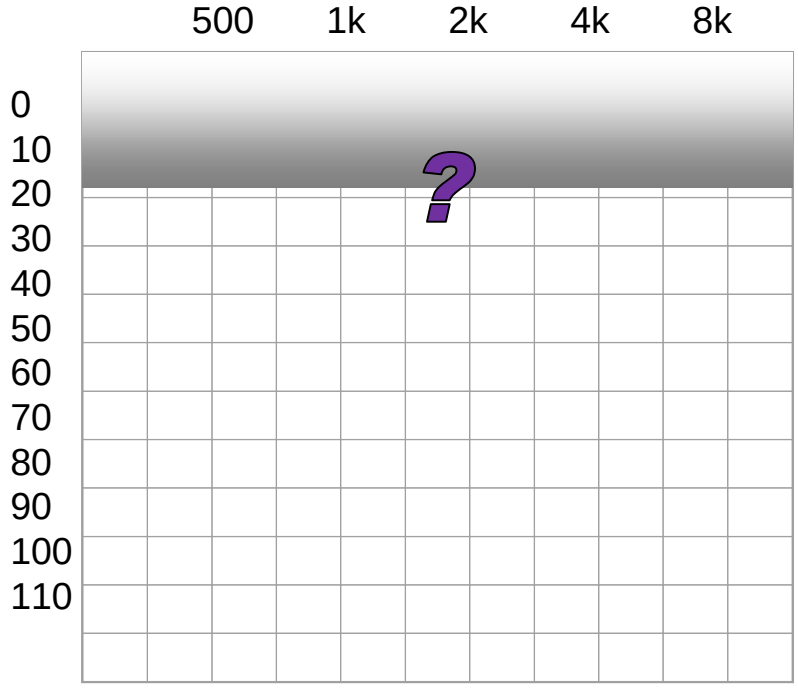


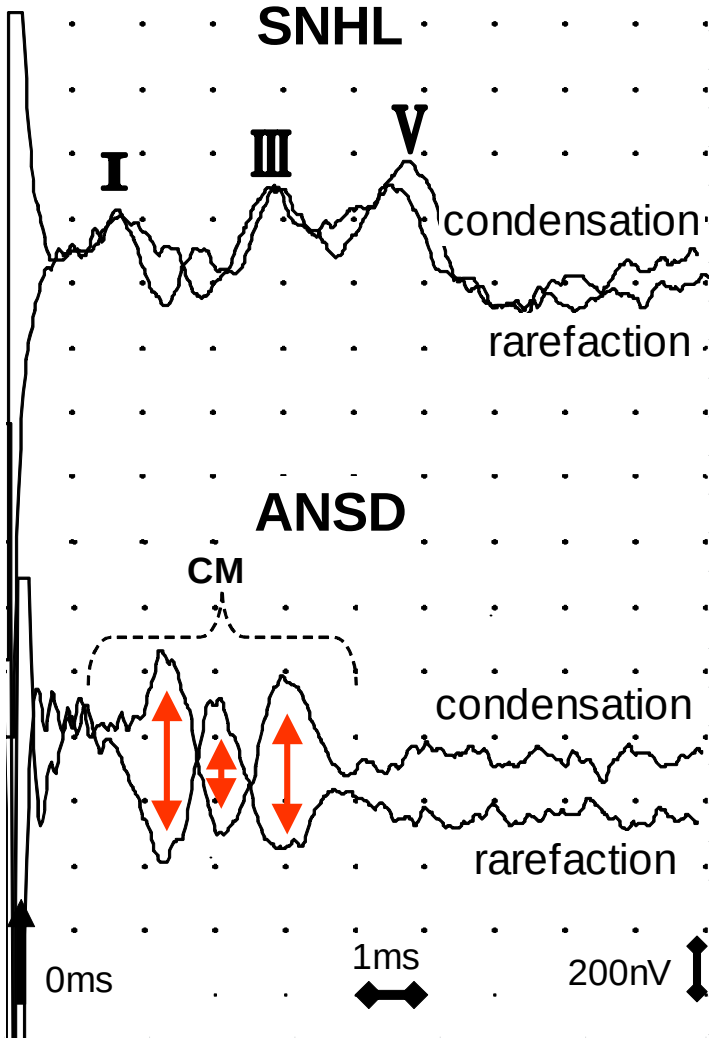
Background:
- Estimating the audiogram with ABR - ANSD

- In ANSD **there is no ABR or is poorly formed** so thresholds can not be reliably determined

Problem 2

As a result of this it is not clear to the clinician whether they should fit hearing aids, and if so how the **hearing aids should be set**.





Background:
– - Early intervention

They need a visual form of communication

Don't use AVT

Problem 3
There is conflicting information in the literature (& on the Internet in general) regarding how to best manage this population & what the likely outcomes will be.

How can we best advise parents about the most appropriate way forward for their baby?

Hearing Aids don't work

Cochlear Implants work in some

Question:
What do we really know about the outcomes for infants diagnosed with ANSD?

Auditory Neuropathy Spectrum Disorder (ANSD)

Systematic review of the literature

Roush et al. (2011)
American Jnl Audiology Vol 20, p159-70

When amplification is used..

- What is the effect on auditory outcomes?
- What is the effect on speech & language outcomes? ✗
- What is the effect on academic outcomes? ✗
- What is the effect on social emotional/parent outcomes? ✗

When cochlear implants are used..

- What is the effect on auditory outcomes?
- What is the effect on speech & language outcomes? ✓
- What is the effect on academic outcomes? ✗
- What is the effect on social emotional/parent outcomes? ✗

202 articles were assessed for their methodological quality

Only **18** were deemed to be of sufficient quality to answer the review questions

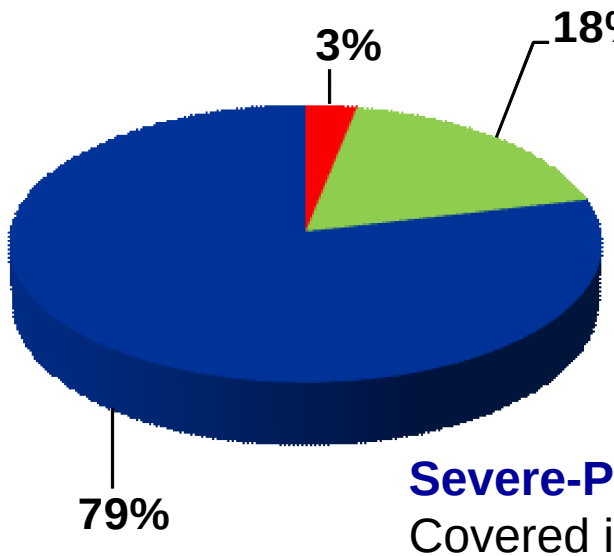
Overall: "The findings from this review do not resolve the controversies surrounding the audiologic treatment of ANSD in children.
The methodological limitations, heterogeneity of the participants, and the varied outcomes reported provide insufficient clinical evidence to guide the practicing clinician."

Hearing Aid vs. Cochlear Implant Outcomes

Imbalance in the literature

Roush et al. (2011) cont'd

"Especially lacking are HA performance data for children with ANSD whose pure-tone thresholds are in the mild to-moderate range "



Mild-Moderate hearing loss
Only covered in **4 articles**
N=**12 subjects**!

Severe-Profound hearing loss
Covered in **14 articles**
(N **>88 subjects**)

Aims

Given the limited literature on speech & language development in children with ANSD the aims of this study were to:

- Investigate the impact of the presence of ANSD on speech and language development of children at 3 years of age.
- To compare the outcomes of children with ANSD to those with SNHL.

Methods - Participants

N = 45 children with ANSD (28 male, 19 female)
[44 screened at birth]

Age at **Diagnosis** = **3.3** mths *(SD 2.2)*

Age at **Hearing Aid fitting** = **6.2** mths *(SD 3.5)*

Age at **Cochlear Implantation** = **18.2** mths *(SD 6.6)*

Additional Disabilities: ***(top 3)***

**NB many children had more than 1 disability*

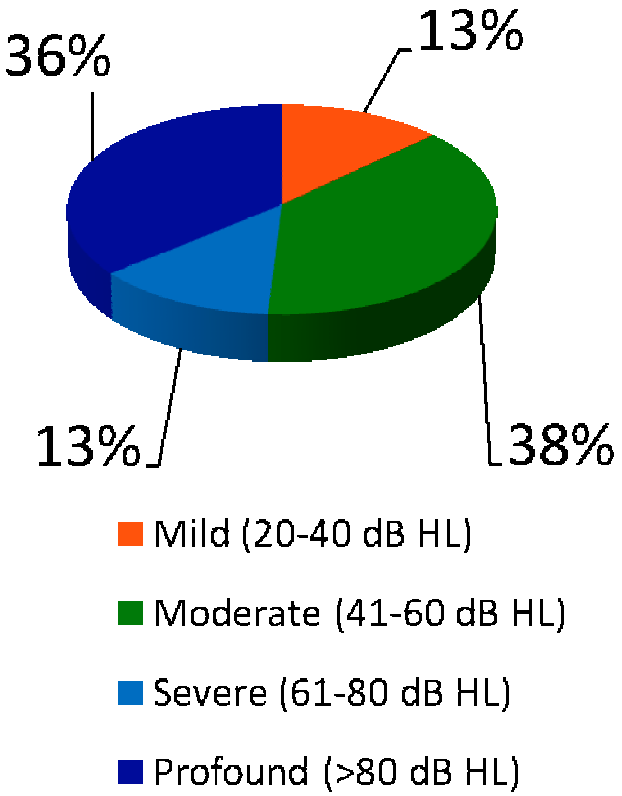
Prematurity (<37 weeks gestation) = 70%

Mechanical **ventilation** = 38%

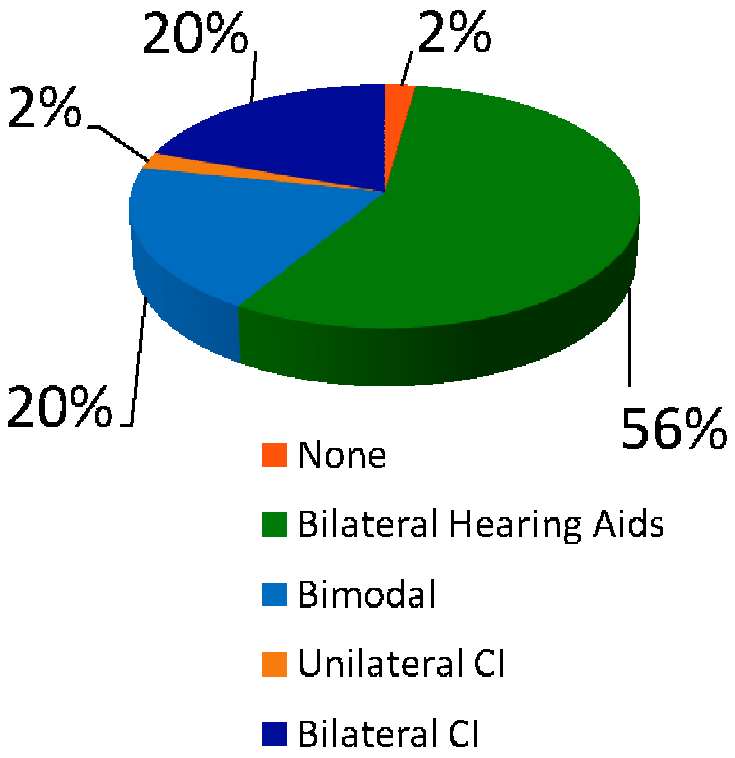
Jaundice = 30%

Methods: Participants

Degree of hearing loss



Devices used



Mode of communication

Aural/oral only	80%
Oral and sign	20%
Sign only	0%

Methods - Assessments

Pre-school Language Scale 4 **(PLS-4)** Child Development Inventory *Part I* **(CDI)**	**Expressive & Receptive language**
Peabody Picture Vocabulary Test **(PPVT)**	**Receptive vocabulary**
Diagnostic Evaluation of Articulation and Phonology test **(DEAP)** *Articulation and Phonology subtest*	**Speech production**
Child Development Inventory *Part II* **(CDI)**	**Social** **Self-help** **Gross motor** **Fine motor**
Parent Evaluation of Aural/Oral performance in Children **(PEACH)**	**Functional communicative performance in real-world situations**

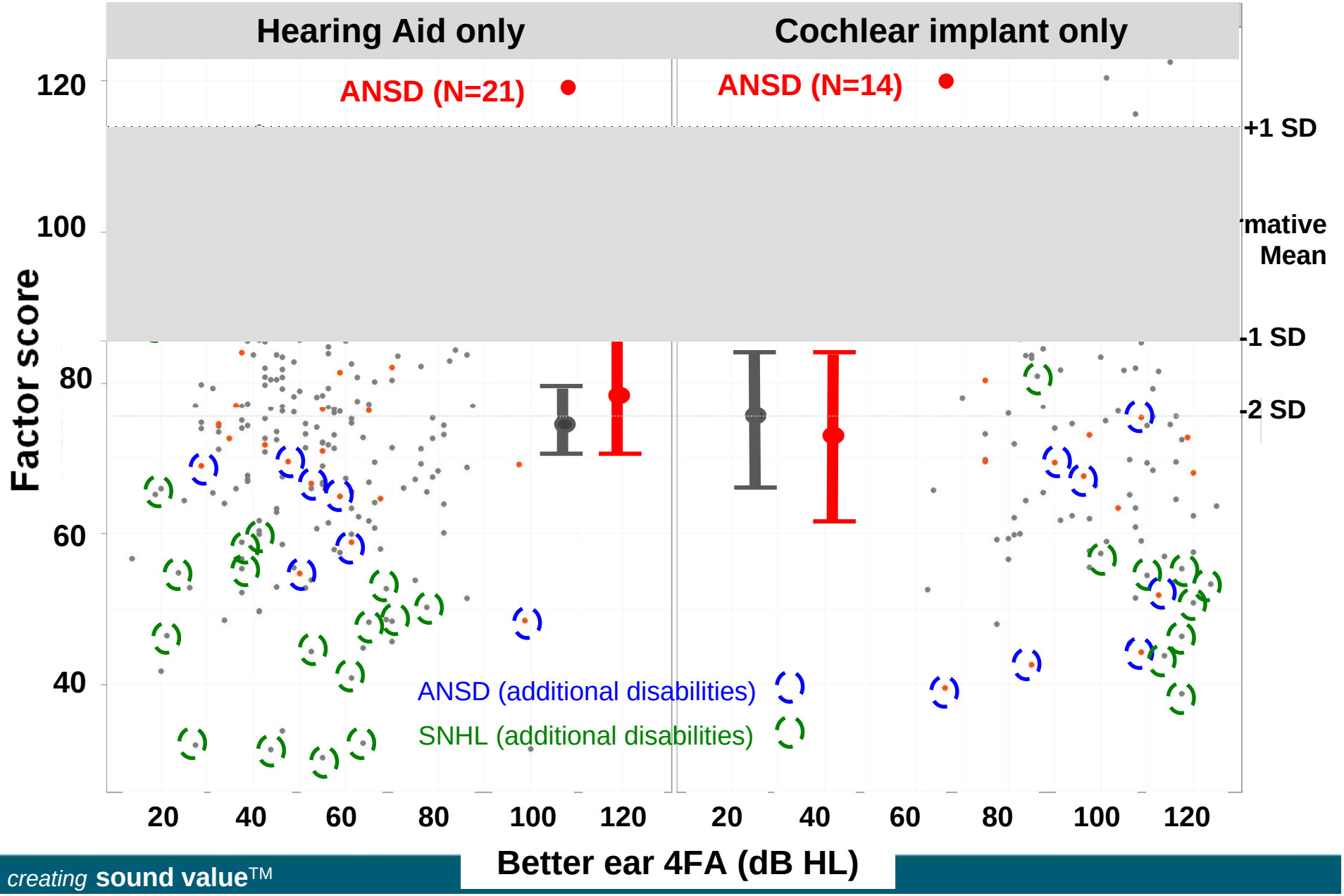
Forming a global factor score

Primary Outcomes Scores

Outcome	Factor loading
PLS language expression	0.92
CDI language comprehension	0.90
CDI expressive language	0.87
Peabody picture vocabulary	0.86
PLS auditory comprehension	0.85
DEAP vowel production	0.78
DEAP consonant production	0.73
PEACH	0.63
CDI social score	0.63
TEACH	0.53

For a full description see
Ching et al. (2013)
Ear & Hearing
(published ahead of print)

Results - Factor score, SNHL vs. ANSD



Conclusions

Finding 1: - No significant group differences

Speech, language, and psychosocial outcomes of children with ANSD did not differ significantly from those with SNHL – both for children with hearing aids, and children with cochlear implants.

Hearing aids ($p = 0.36$)
Cochlear implants ($p = 0.61$)

Finding 2: - No significant difference in variability of scores

The variability in outcomes of children with ANSD did not differ significantly from those with SNHL – both for children with hearing aids, and children with cochlear implants.

Hearing aids ($p = 0.45$)
Cochlear implants ($p=0.12$).

We know that <u>some</u> children with ANSD demonstrate unusual auditory behaviour & poorer than expected speech & language ability,
however when we look at this group as a whole the situation is not necessarily as dire as we previous thought!

Discussion

This cohort of children differs to many other studies in that:

- All identified early through newborn hearing screening
- All received **early** language engagement/intervention
- All were fitted early with hearing aids using a standard fitting procedure
- This was a prospective study

It is appropriate to start the audiological & early language engagement journey for children with ANSD in a similar way to what we do for children with SNHL
The important point is to continue monitoring language development over time to identify if a change in the approach to management is needed
(this is true for both ANSD and SNHL!)

Analysis of the 5 year outcomes data is currently underway to investigate whether any differences between the two groups develop over time.
This will include measures of speech discrimination ability in quiet & noise

Collaborating parties

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From left to right: Laura Street, Lauren Burns, Henrik Dahl, Vanessa Raymond, Gerrie Krynda, Angela Wong, Vicky Zhang, Kirsty Gardner-Berry, Angel Yeh, Julia Day, Melanie Reid, Patricia Van Buynder, Jessica Sjahalam-King, Kelly Stroud, Paola Incerti, Vivienne Martin, Jessica Thomson, Kathryn Crowe, Megan Gilliver, Sanna Hou, Christopher Flynn. www.outcomes.nal.gov.au

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The full article for this paper is currently “in press” with the **International Journal of Audiology** (2013)