



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

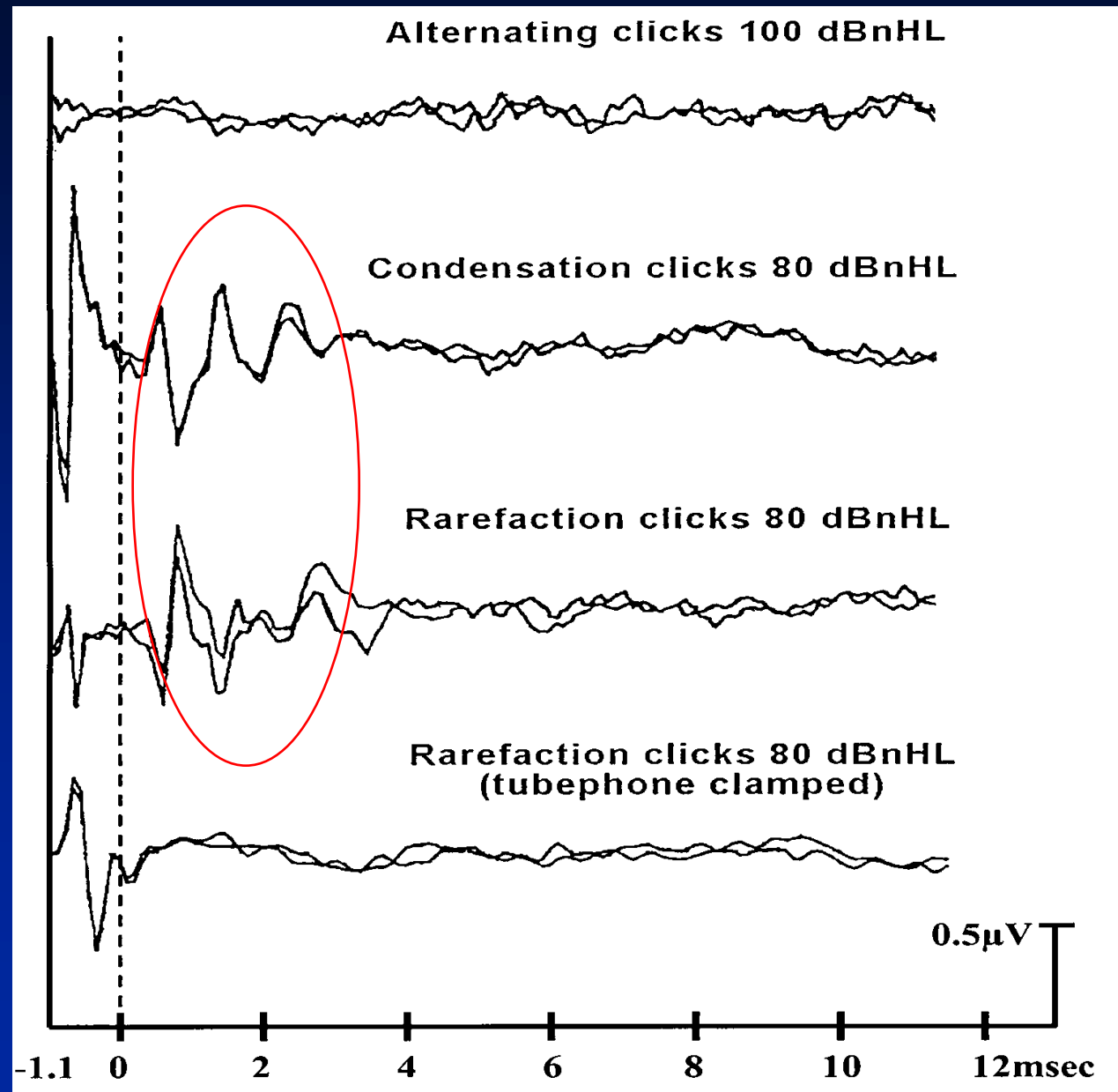
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The University of Melbourne

International Paediatric Conference, Shanghai, April 2014

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: \approx 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)

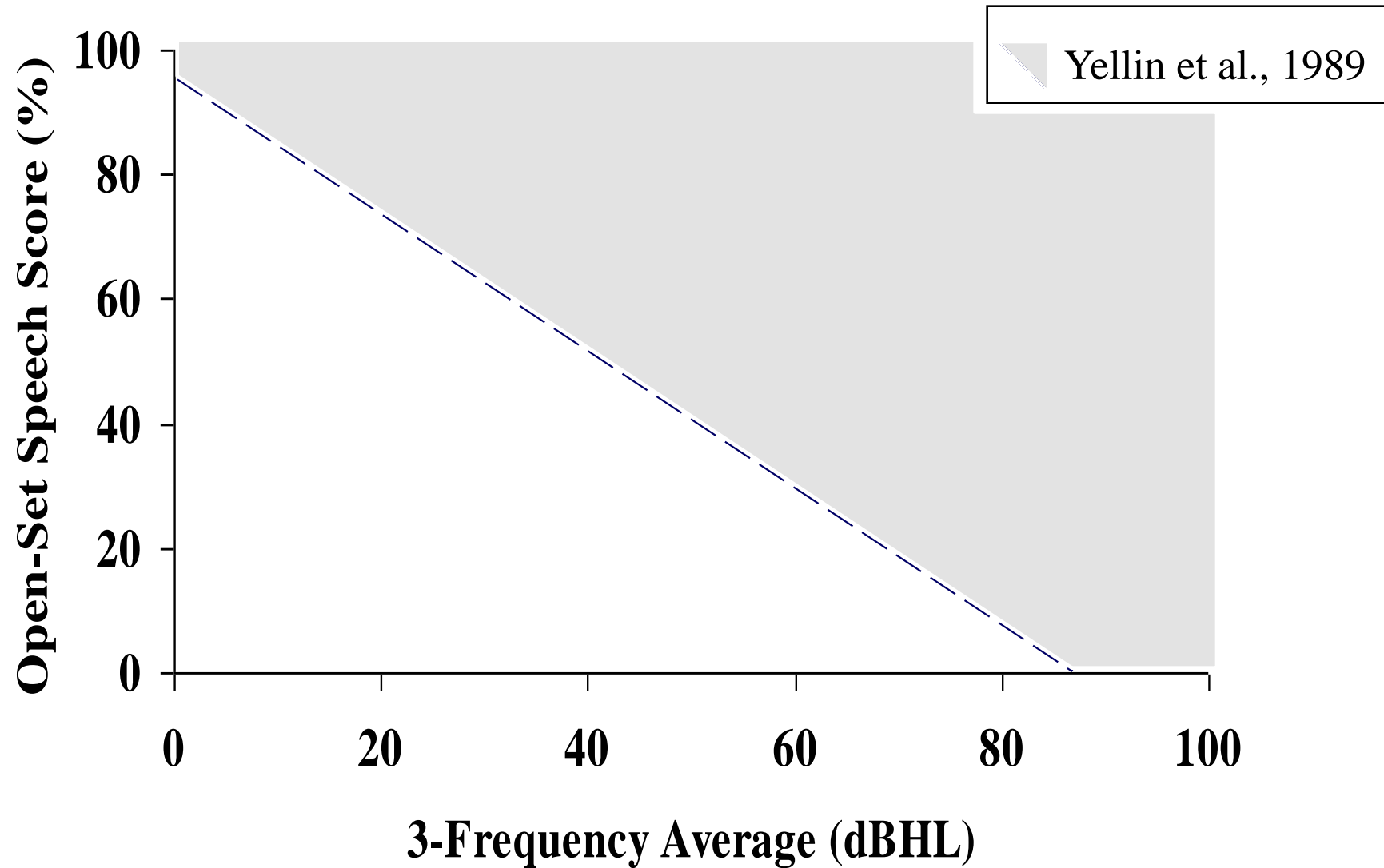
◆ Functional hearing

- Impaired speech perception

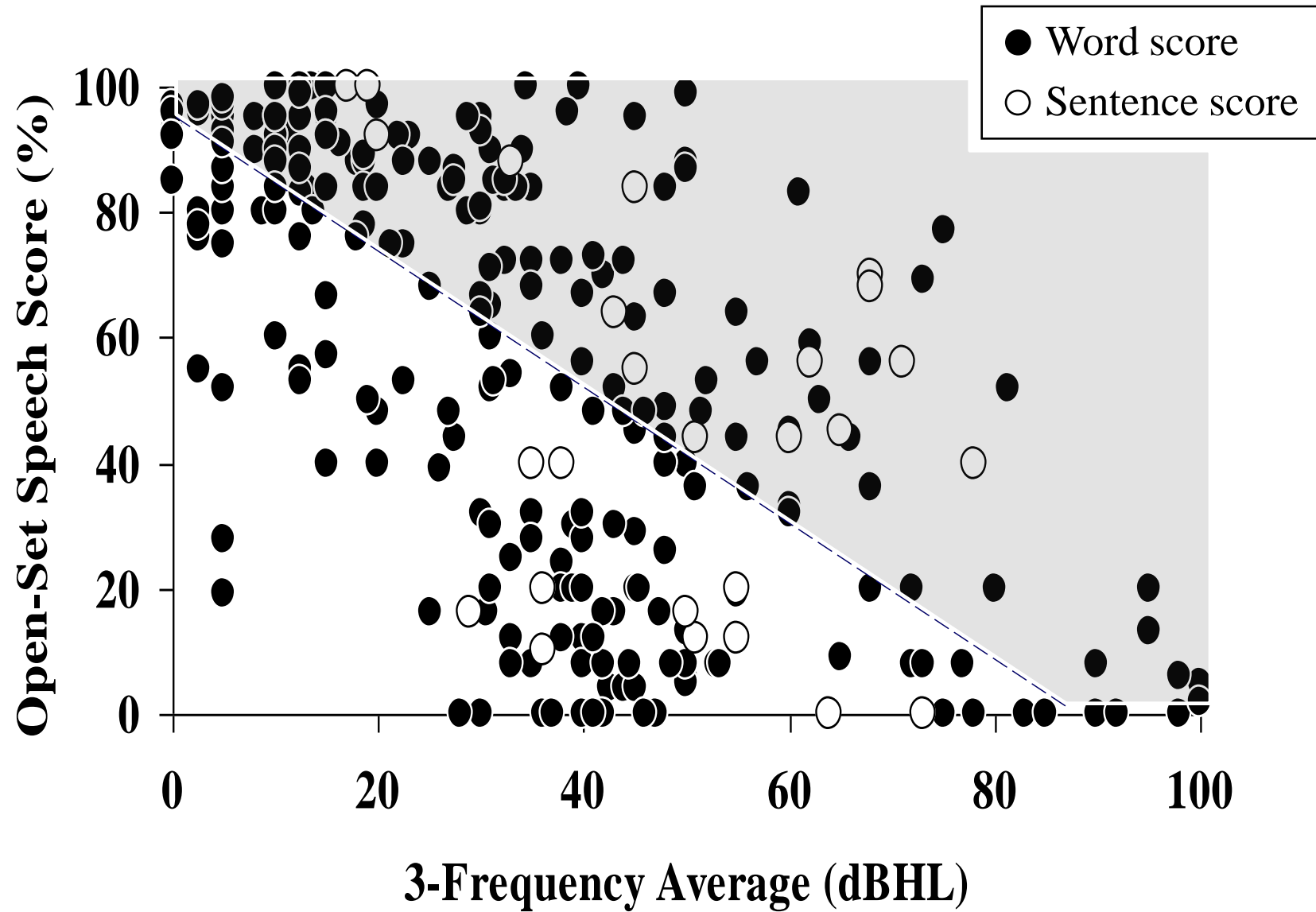
Speech Perception

- ◆ 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



Open-set Speech Perception v Hg Level for Children & Adults with ANSD



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

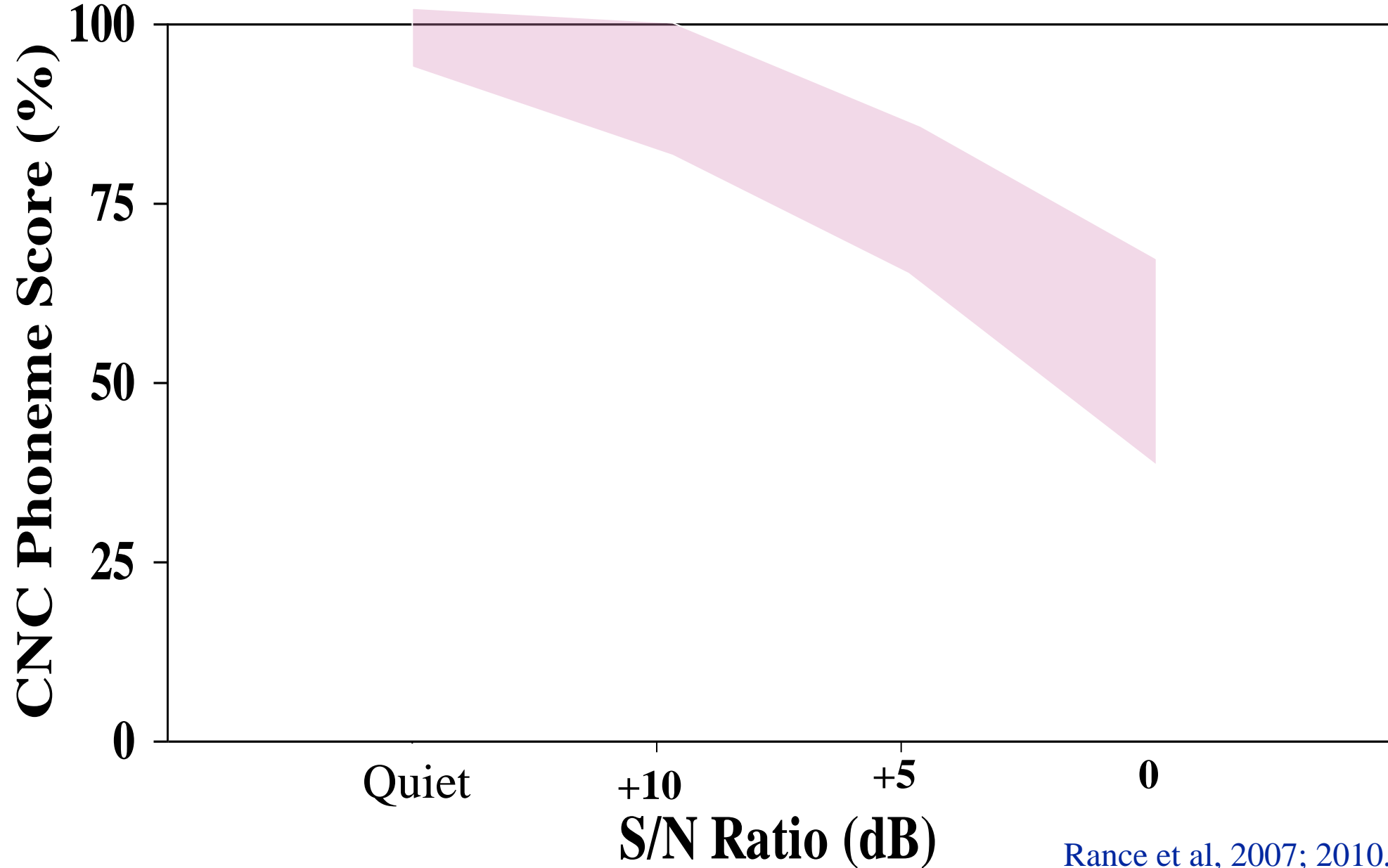
Speech Perception in Noise



- ◆ 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

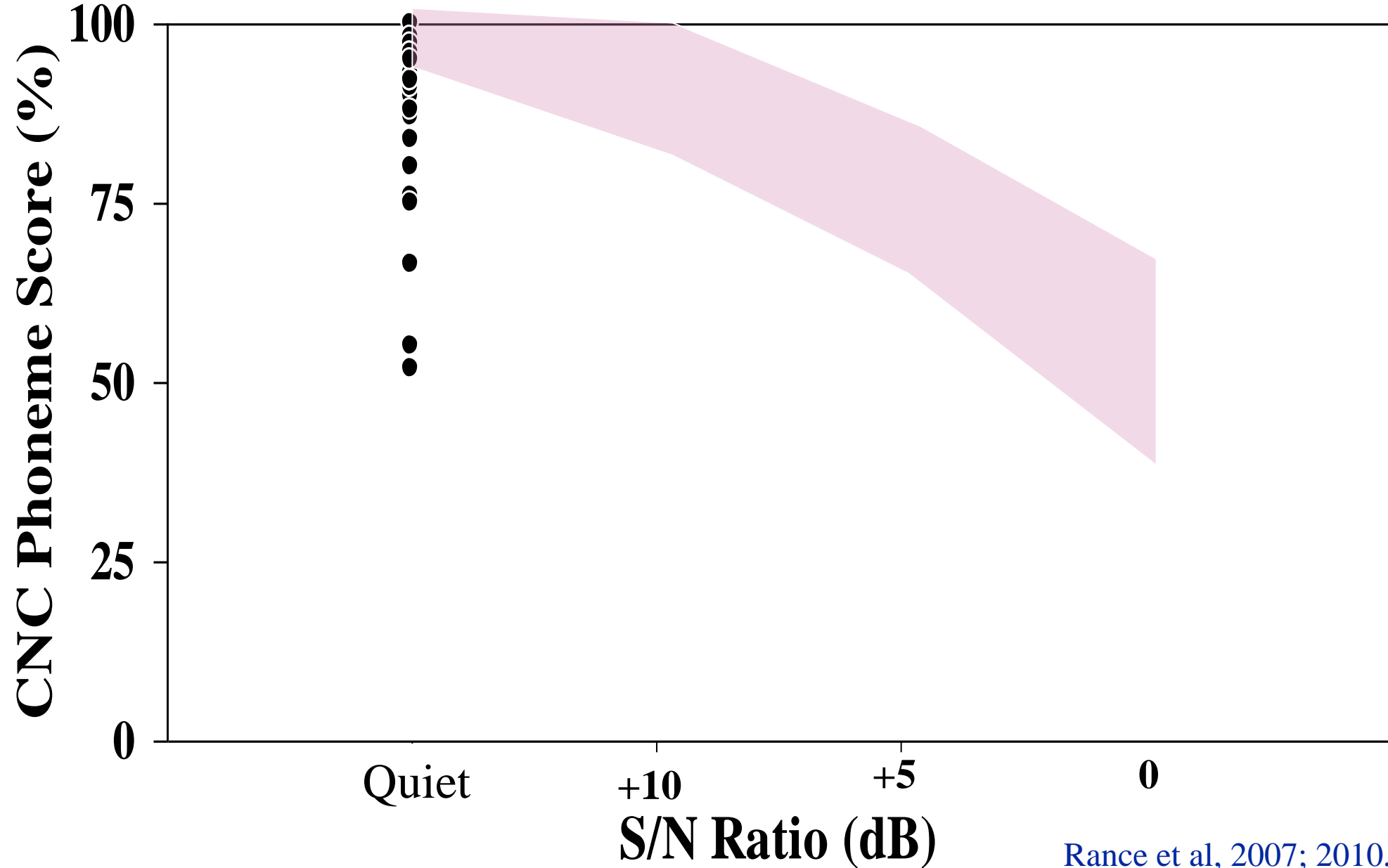
Speech Perception in Noise for Children with ANSD

(normal sound detection)



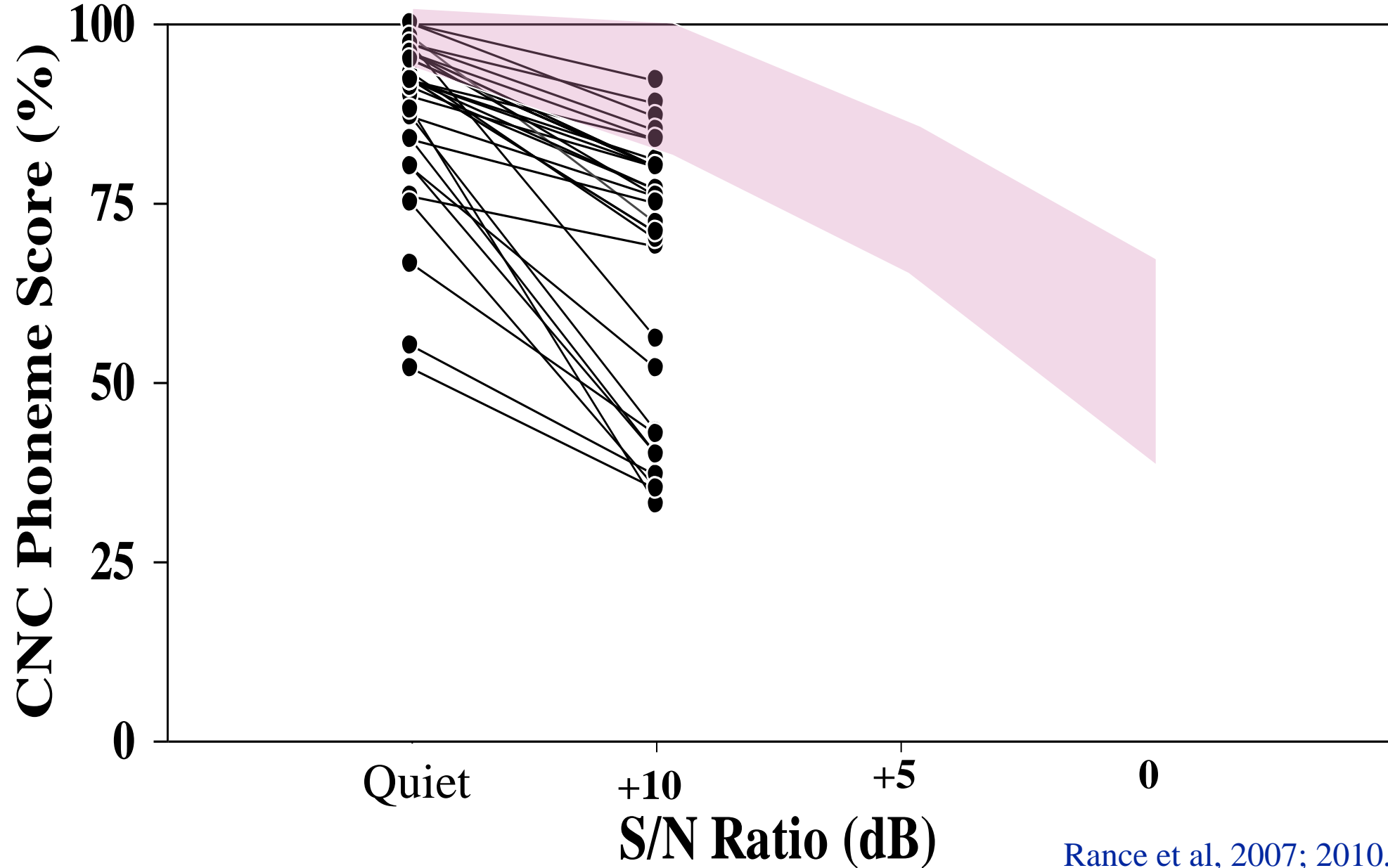
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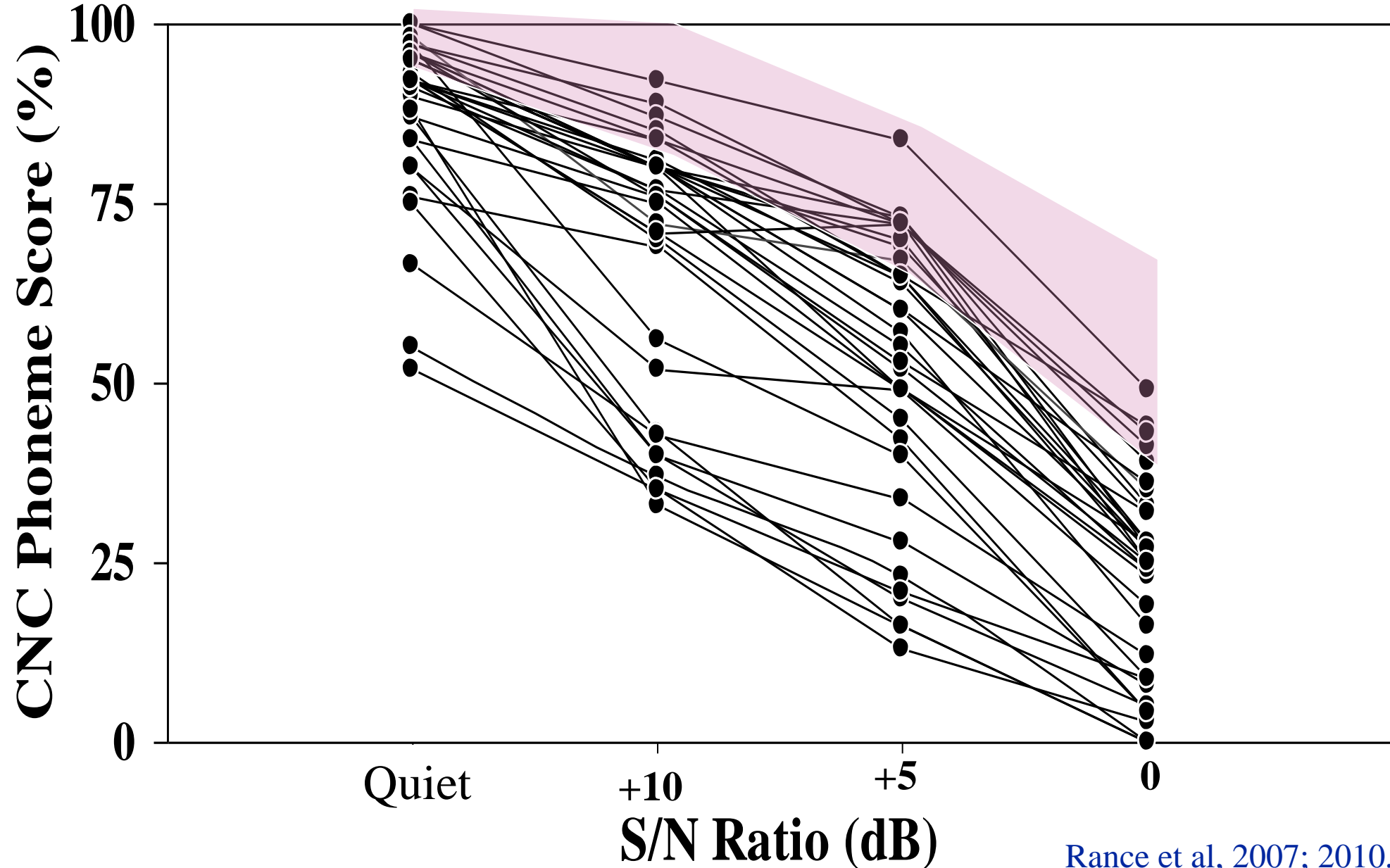
Speech Perception in Noise for Children with ANSD

(normal sound detection)



Speech Perception in Noise for Children with ANSD

(normal sound detection)



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

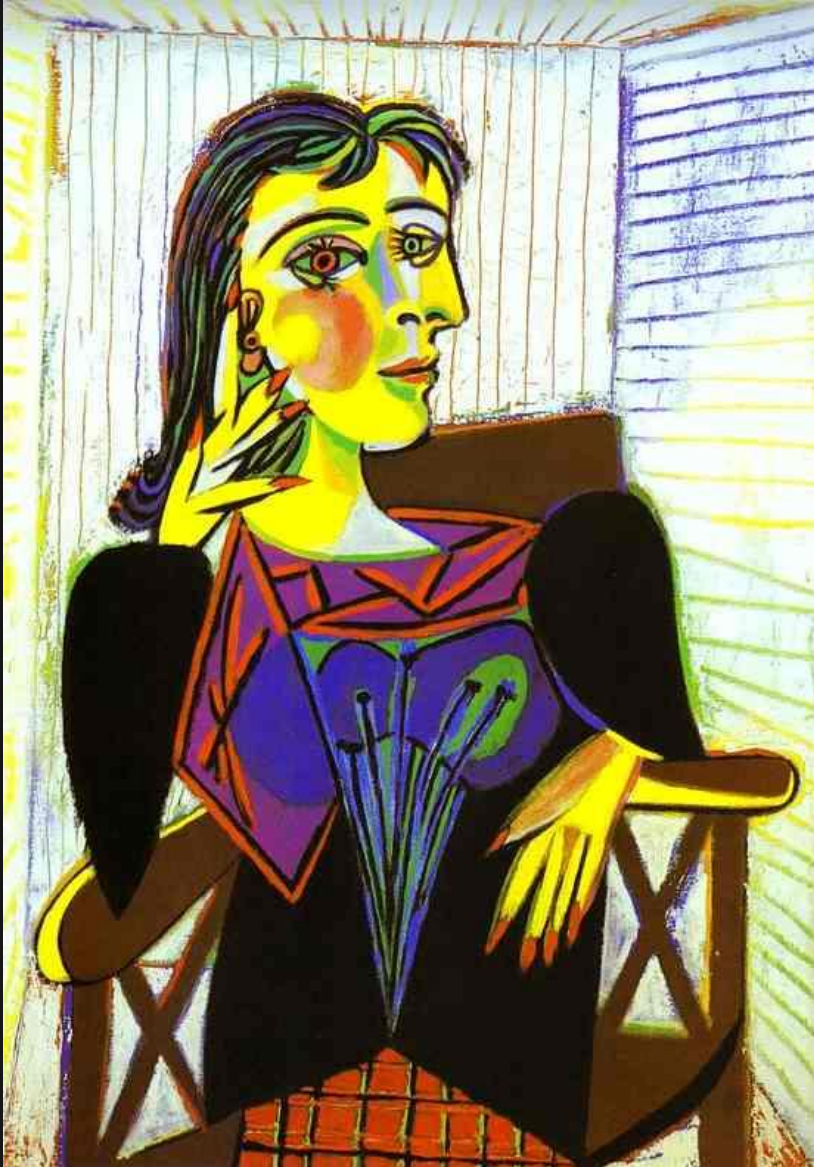
Management of Children with ANSD

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)

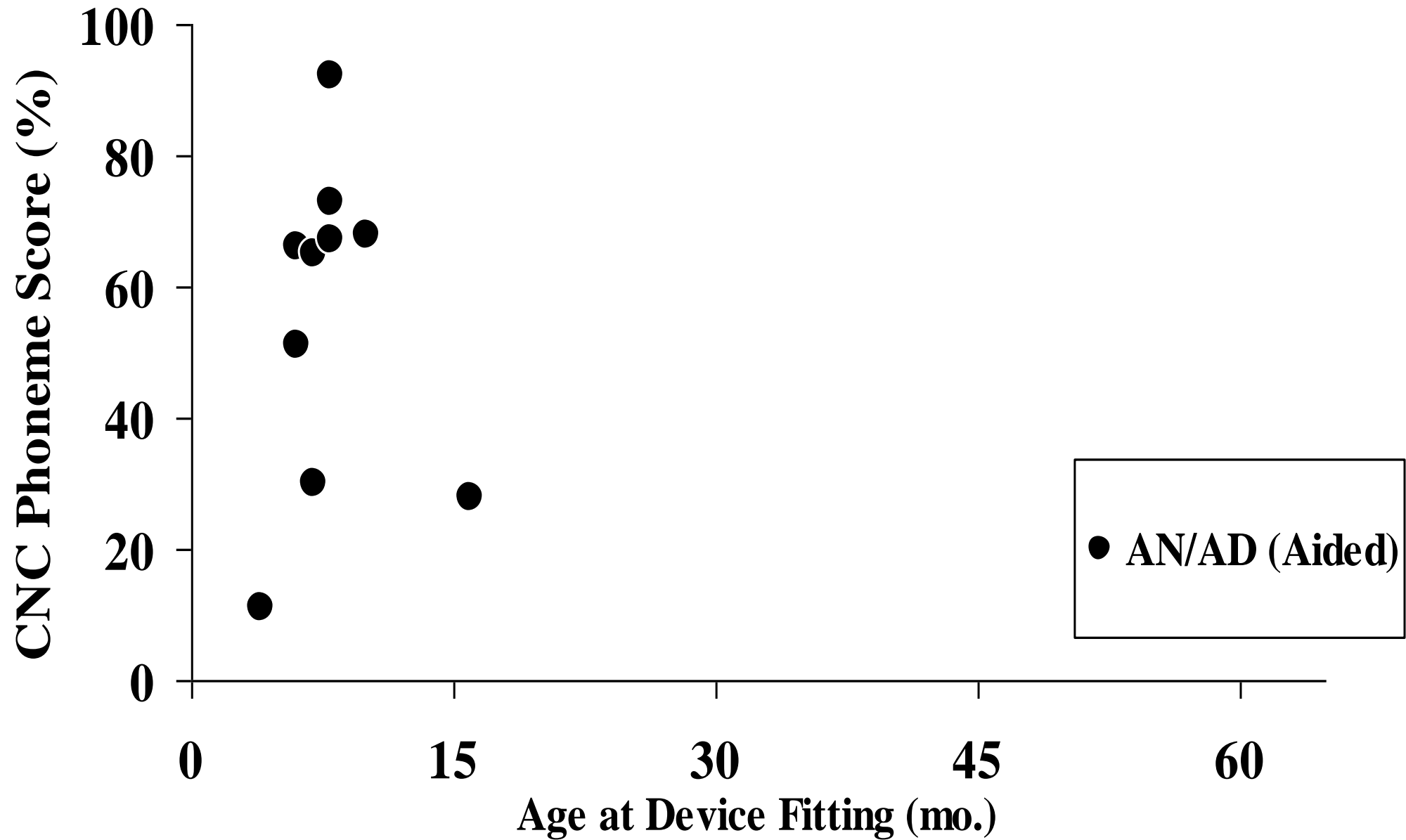
◆ Subjects

- Aided ANSD children (N=10)
- Implanted ANSD children (N=10)
- Implanted SNHL children (N=10)

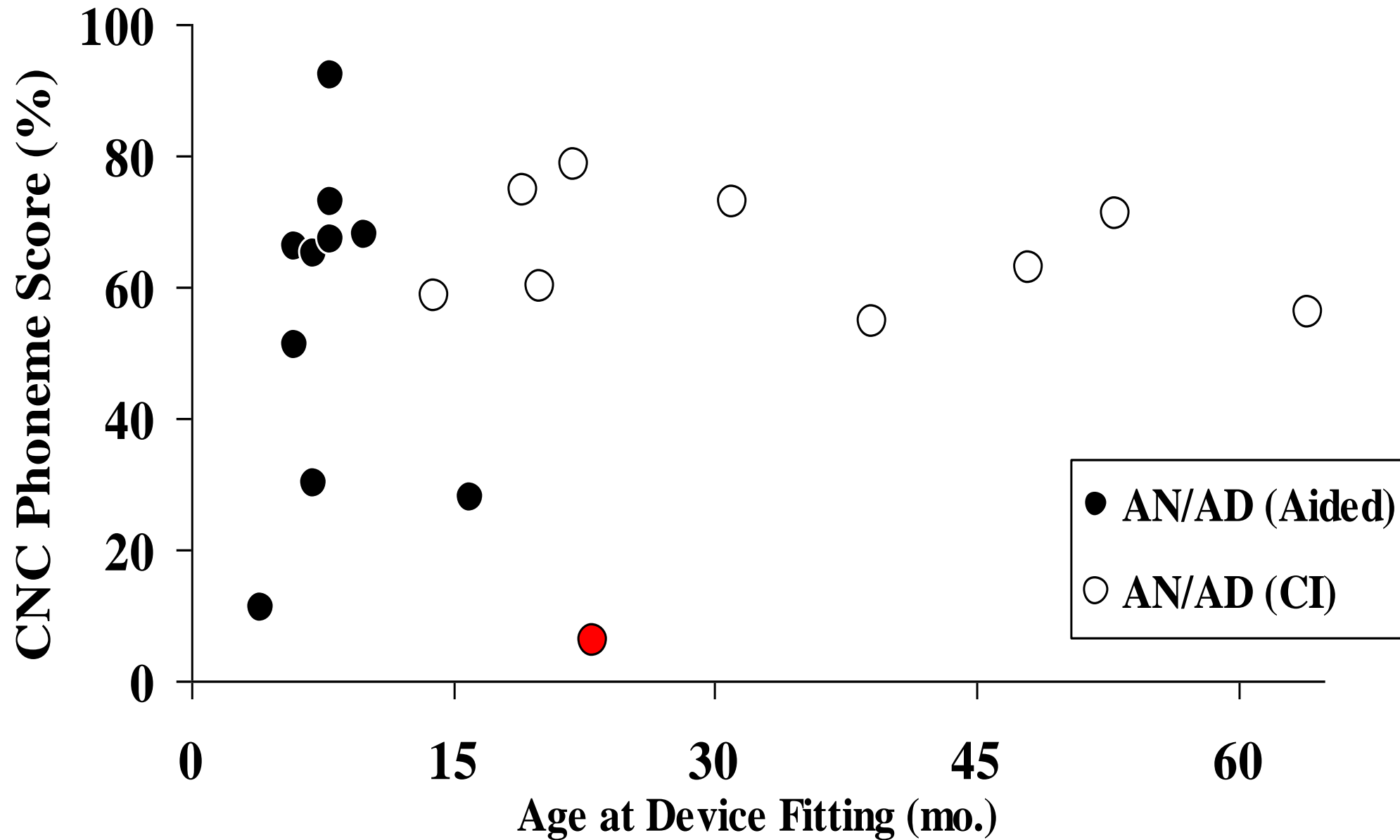
◆ Assessment

- Open-set speech perception (CNC-Words)
- Free-field presentation
- Normal listening condition

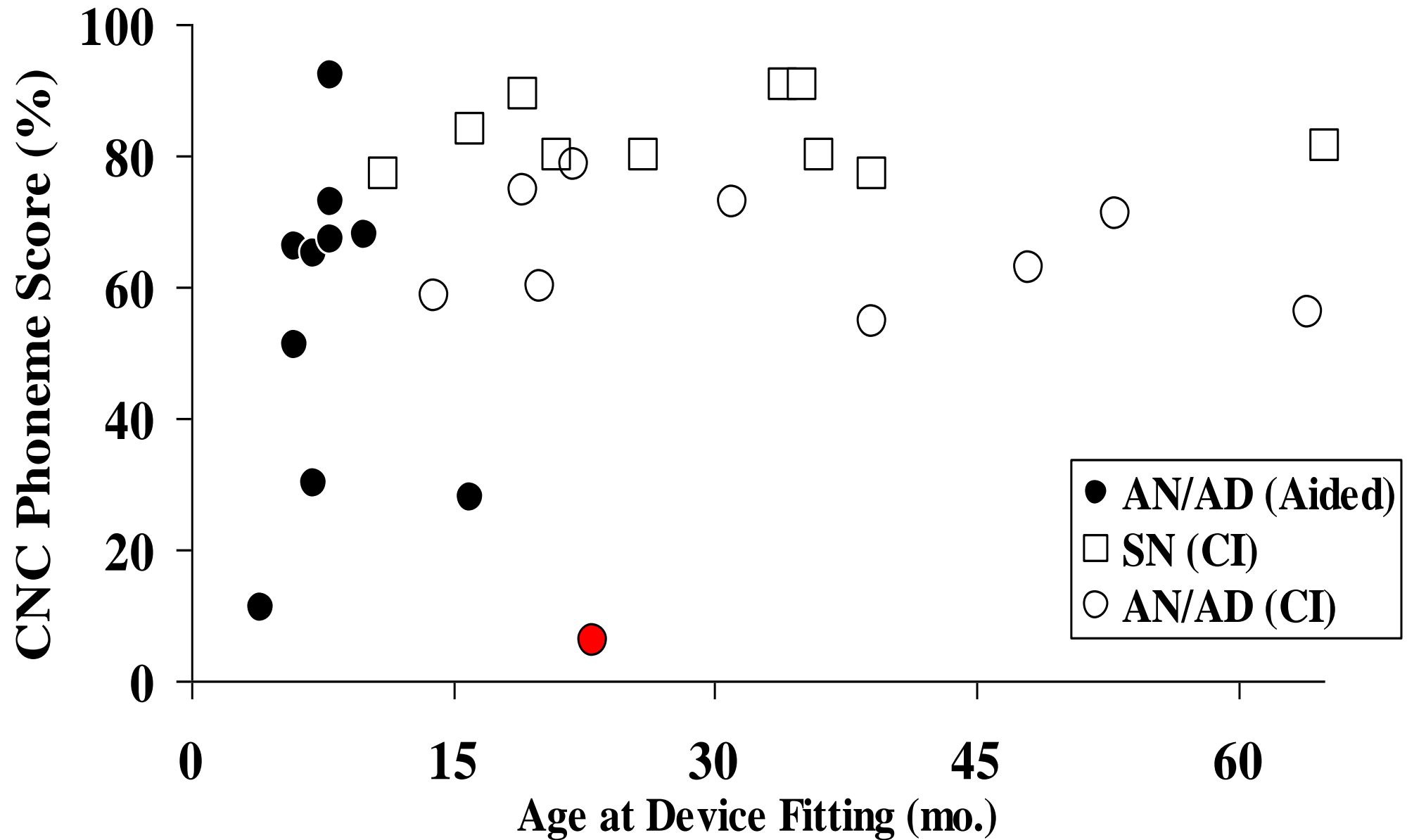
Open Set Speech Perception



Open Set Speech Perception



Open Set Speech Perception



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)

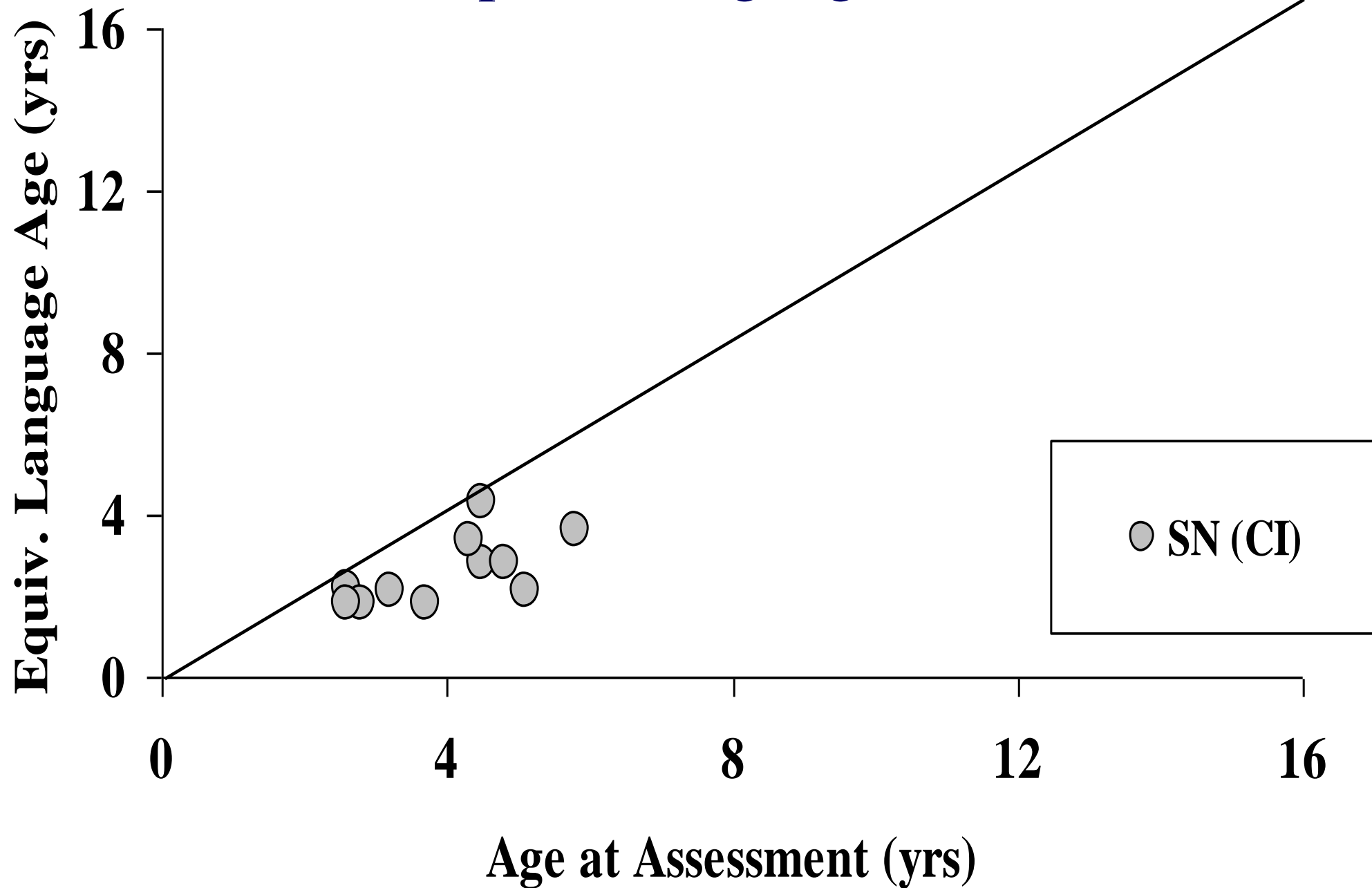
◆ Subjects (April 2014)

- Aided ANSD children (n=8)
- Implanted ANSD children (n=6)
- Implanted SNHL children (n=12)

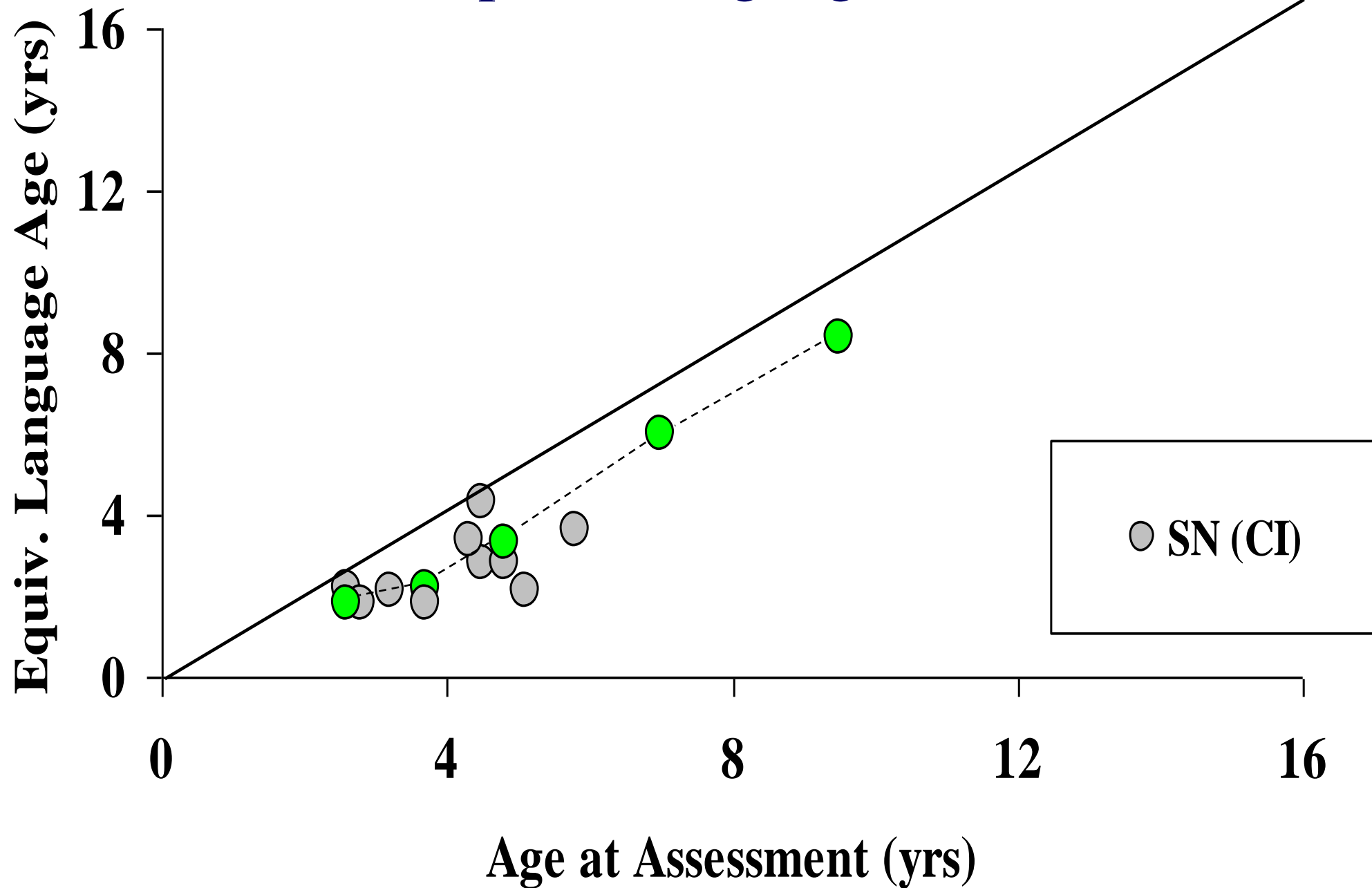
Receptive Language (PPVT)



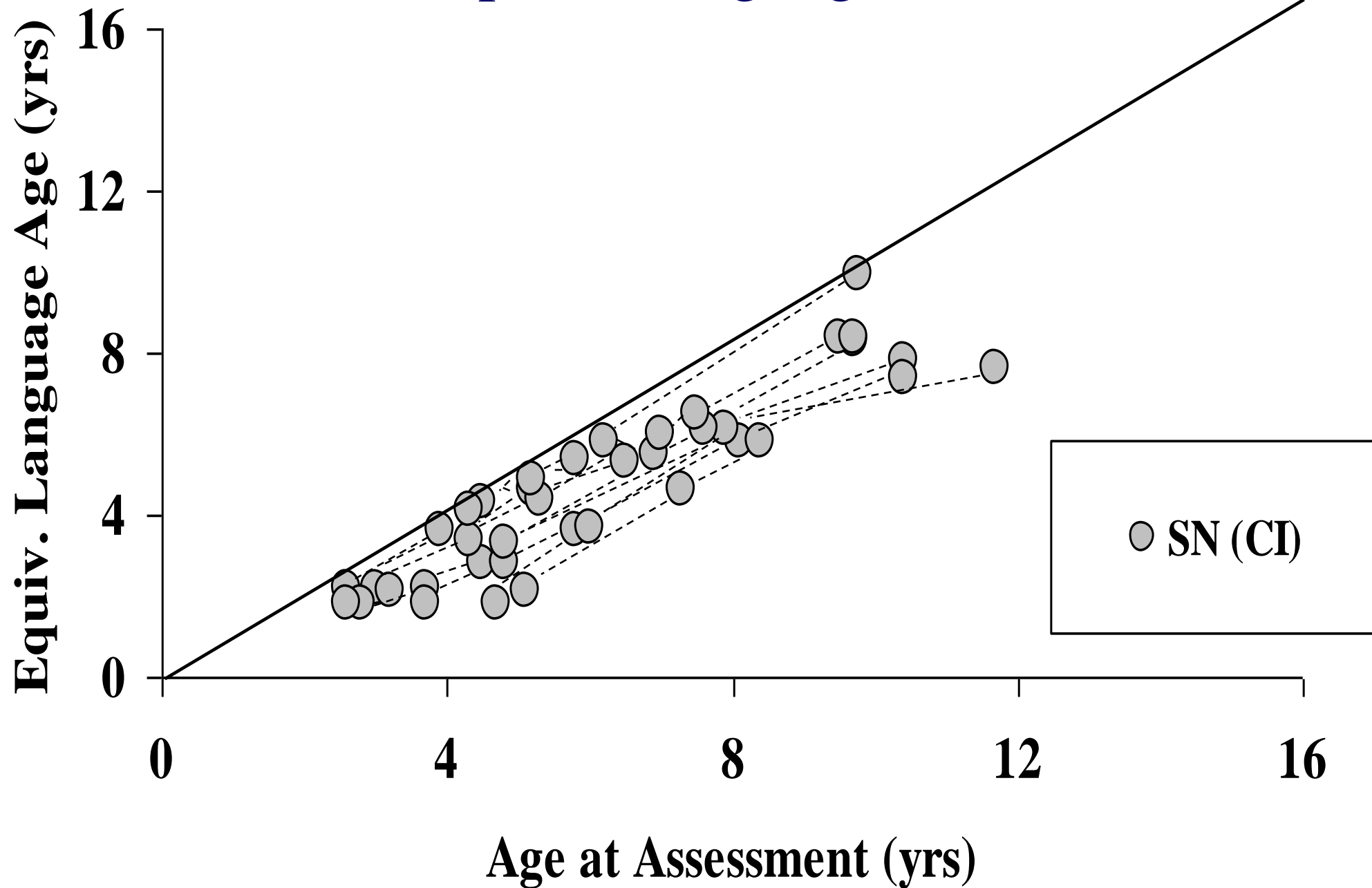
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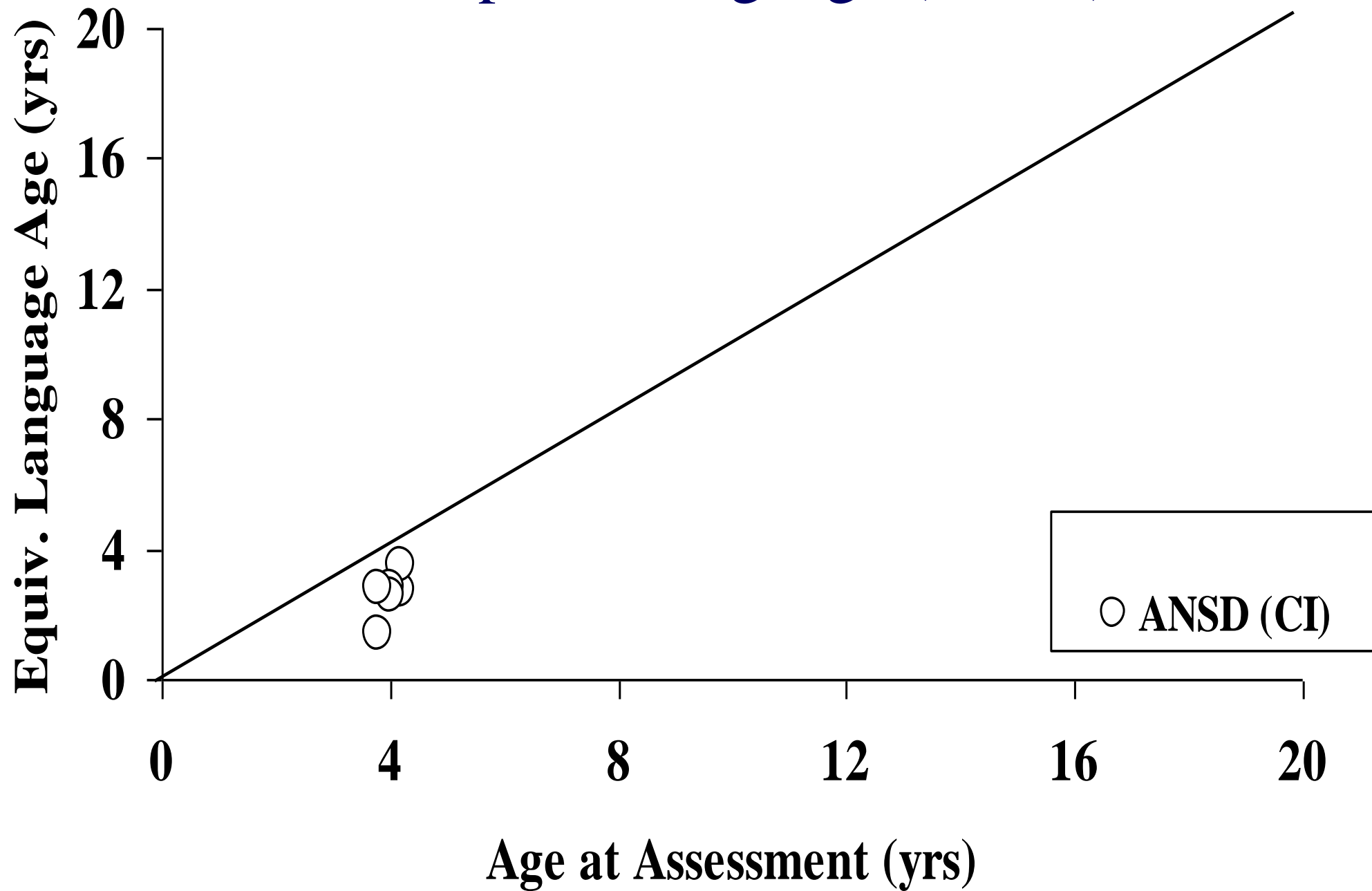
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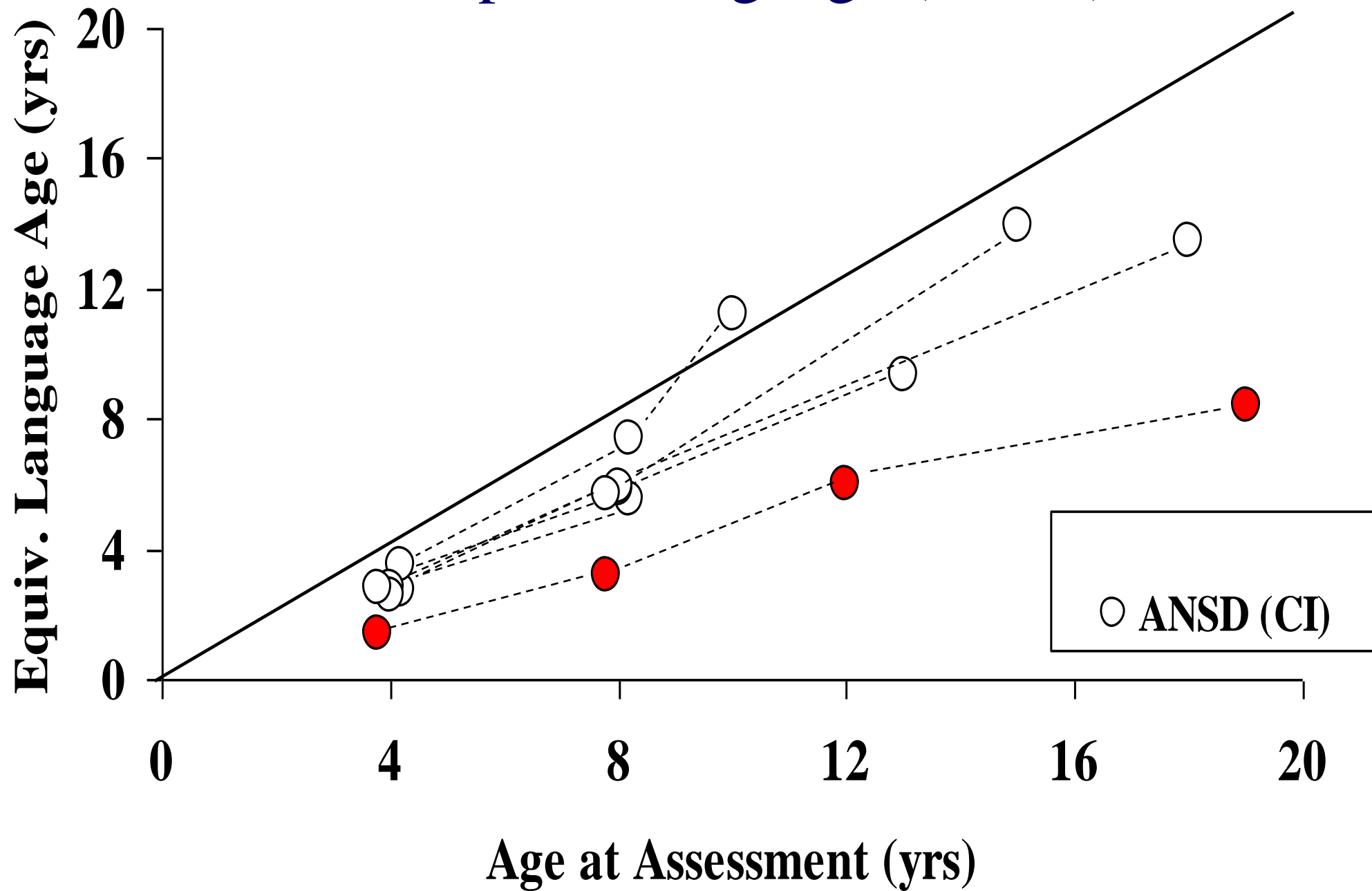
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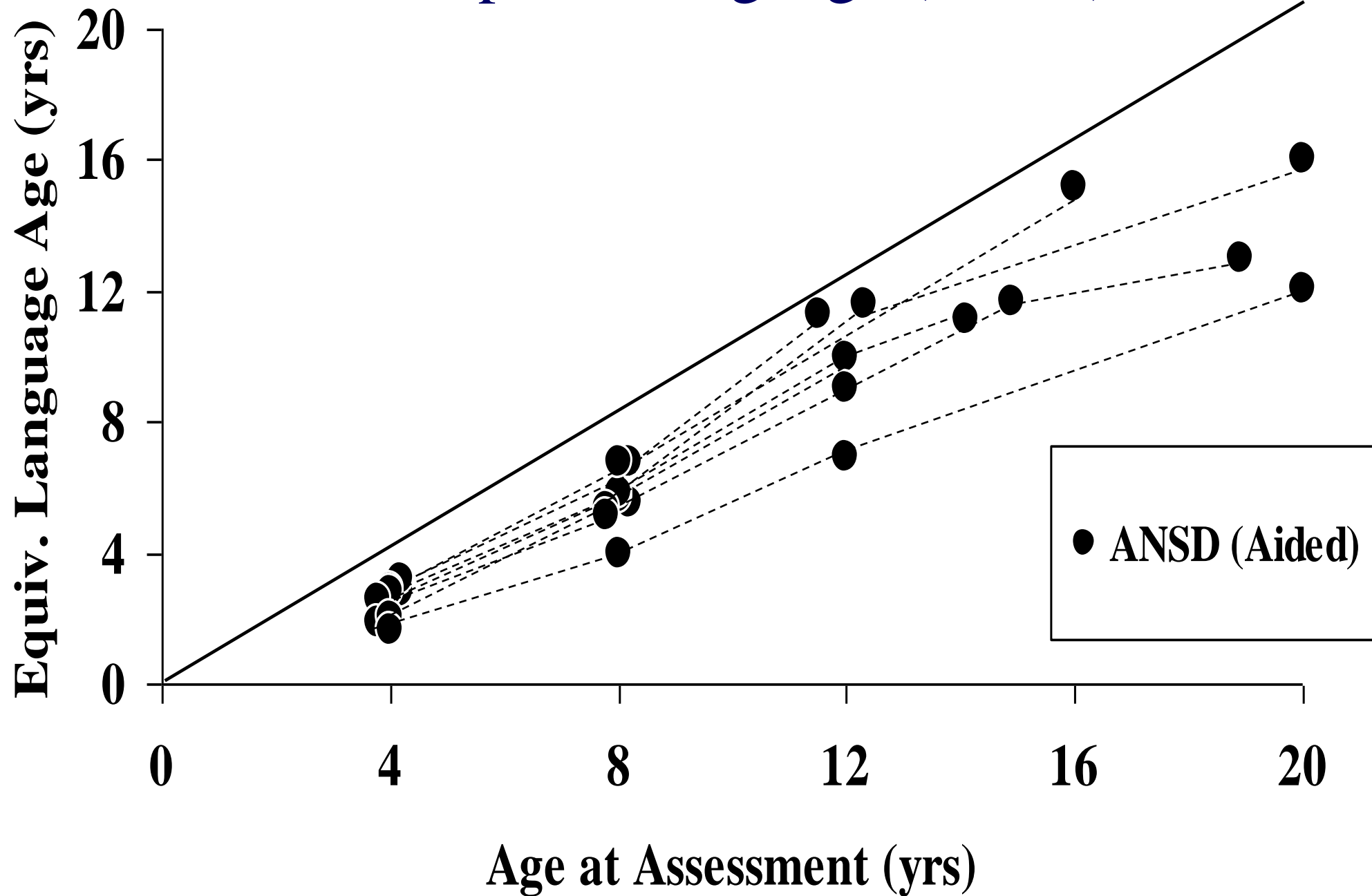
Receptive Language (PPVT)



Receptive Language (PPVT)



Receptive Language (PPVT)



ANSD Management: Hearing Aids / Cochlear Implants

◆ 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

