

An Auditory Verbal Approach for Children with Auditory Neuropathy / Dys-synchrony

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INTRODUCTION

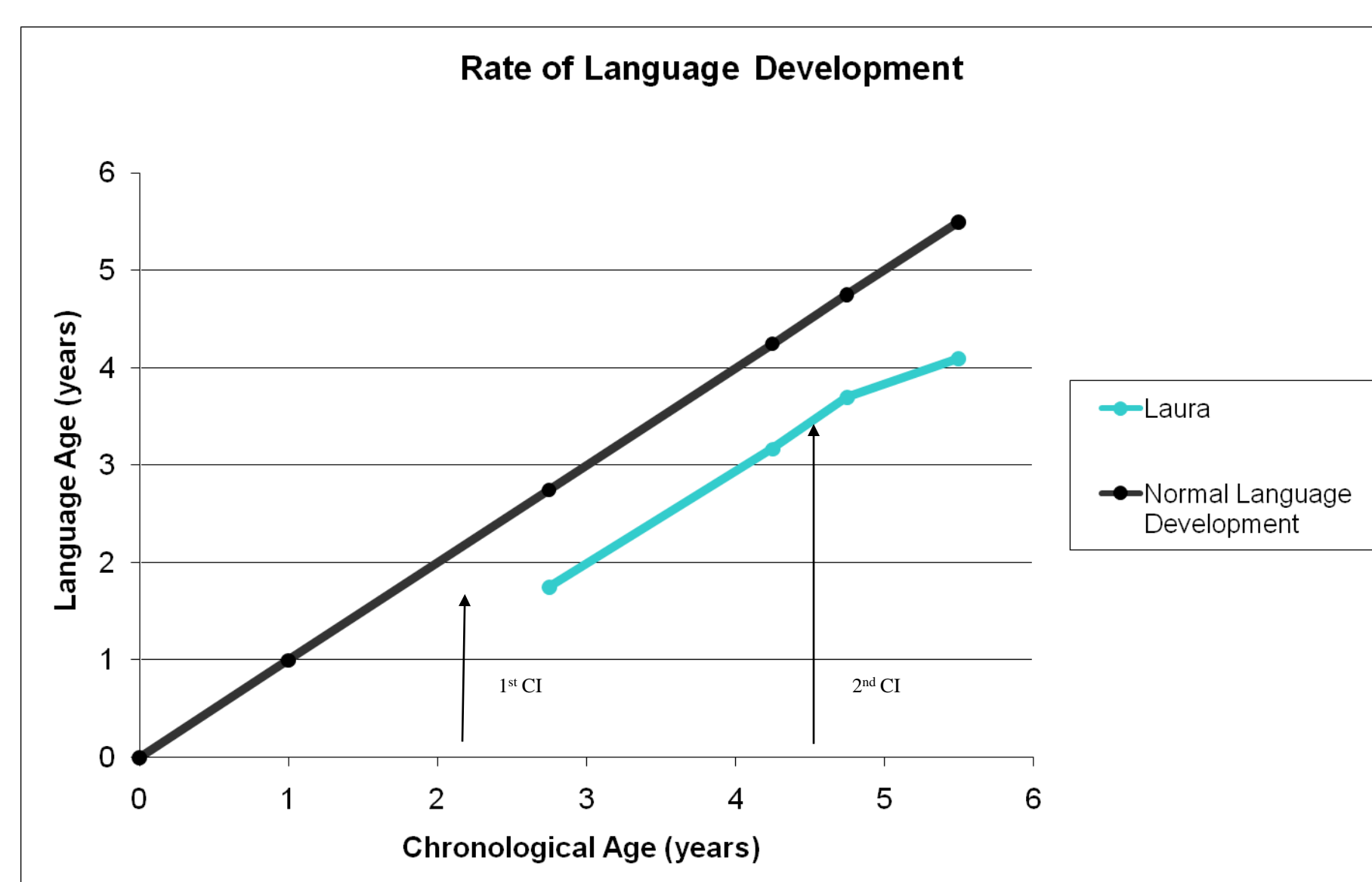
The habilitation of children diagnosed with Auditory neuropathy/ auditory dys-synchrony (AN/AD) needs to be considered on an individual basis because of the wide range of auditory capabilities demonstrated within this sub-population of children with hearing impairment: Some children are not fitted with amplification, others with low-gain hearing aids, yet others with cochlear implants; some children follow an oral-aural approach while others use Total Communication or Sign Language.

Out of 51 families currently engaged with a programme of Auditory Verbal therapy at Auditory Verbal^{UK}, 4 (8%) have a confirmed diagnosis of AN/AD. This is in line with the suggested prevalence of 7% AN/AD within the population of children with permanent hearing loss (Rance, 2005). The listening and spoken language skills of a further 2 (4%) of children have led therapists to query whether these children have AN/AD.

CASE STUDY 1

Laura was born prematurely at 35 weeks. Her behavioural thresholds for PTA were indicative of a profound, bilateral hearing loss for which Laura received sequential bilateral cochlear implants (CI) at ages 2;3 years and 4;6 years. During investigation for suitability for CI, Laura was found to have cochlear nerve deficiency. She was diagnosed with CHARGE syndrome at 3;9 years. She is now 5;9 years old.

Age at diagnosis of hearing loss	3 months
Age at hearing aid fitting	3 months
Age at diagnosis of AN/AD	2;0 years
First contact at AVUK	2;6 years
Age at last assessment	5;6 years
Equivalent language age	4;1 years



CASE STUDY 2

Neil, an identical twin, was born very prematurely at 29 weeks gestation. His twin brother has typical hearing. The aetiology of his hearing loss is thought to be premature birth caused by twin-to-twin transfusion syndrome. Neil's parents initially used BSL to communicate with him, but describe him as wanting to use his hearing and talking to learn. He had a vocabulary of 20-30 single words on arrival at AV^{UK}.

Behavioural thresholds obtained via VRA through earmoulds:

	0.5	1	2	4	KHz
Right	45	35	45	60	dBHL
Left	40	40	35	50	dBHL

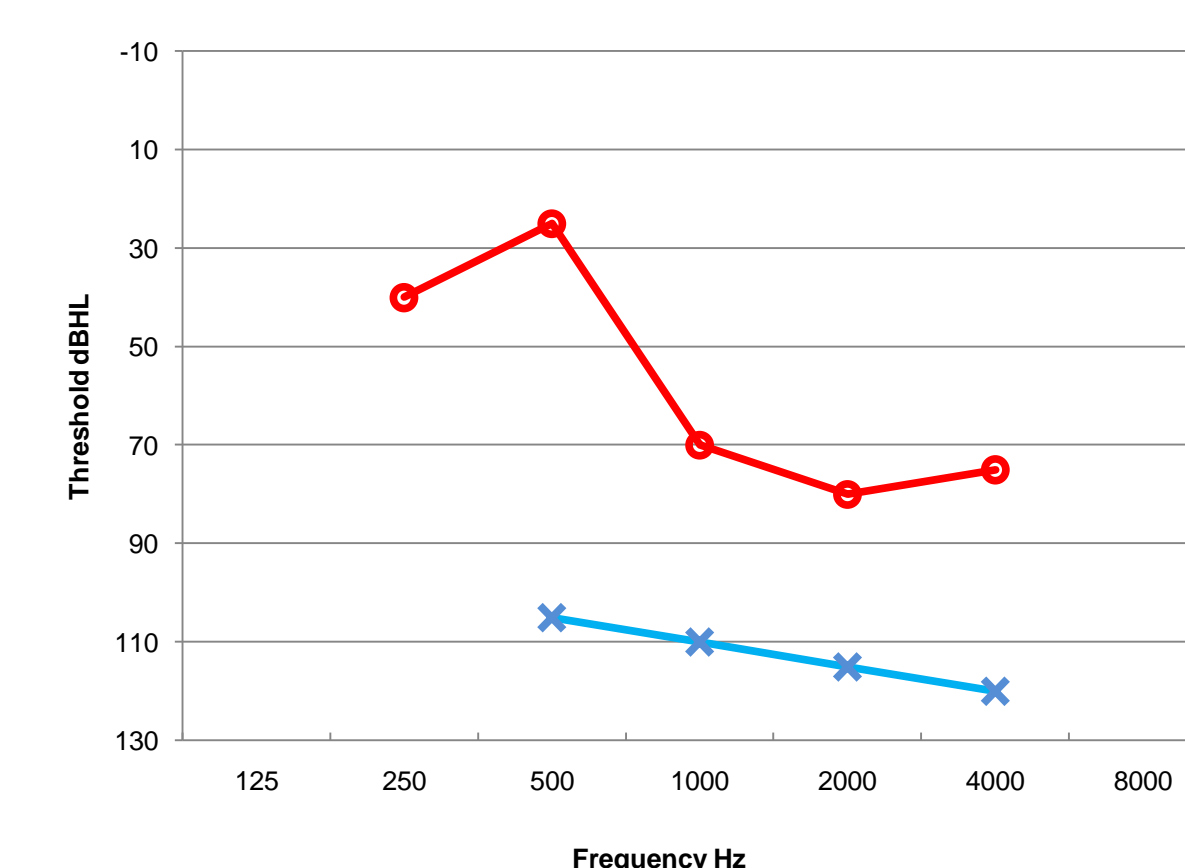
There was no evidence of middle ear effusion at the time of test.

Neil has bilateral hearing aids, which his mother uses on days when she suspects he is not hearing well. His parents have decided to proceed down the Auditory Verbal route for a diagnostic period of six months to assess Neil's auditory potential. He is now 2;9 years old.

Age at diagnosis of hearing loss	4 months
Age at hearing aid fitting	18 months
Age at diagnosis of AN/AD	17 months
First contact at AVUK	29 months
Age at initial assessment	29 months
Equivalent language age	19 months

CASE STUDY 3

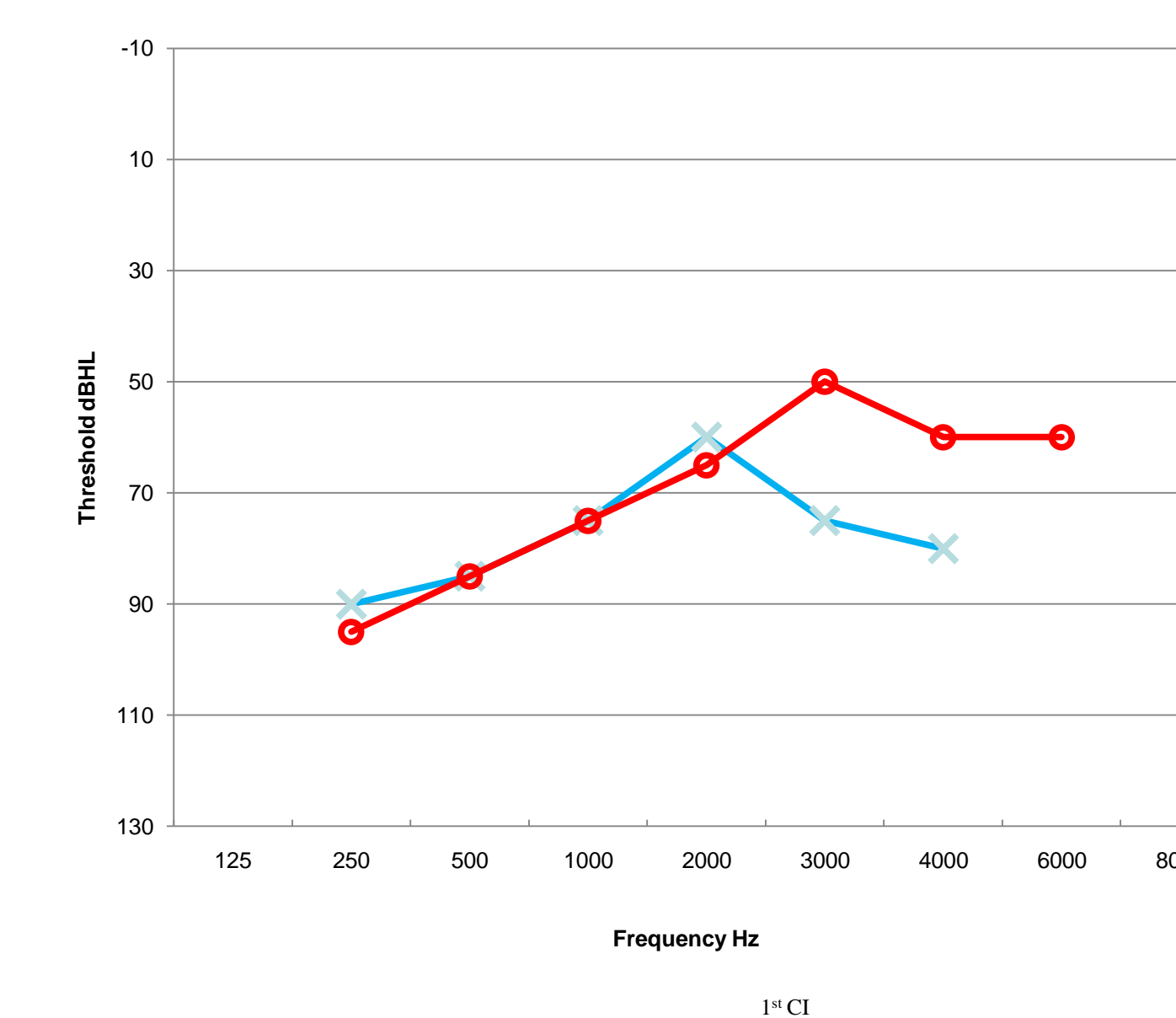
Eddie was diagnosed with a bilateral hearing loss of unknown aetiology following his newborn hearing screen. His behavioural thresholds are suggestive of a moderate loss of sensitivity at the right ear and a profound loss at the left ear. Eddie has also responded to minimal presentation of the Ling sounds in quiet conditions. He was assessed for a cochlear implant, but it was not recommended, largely due to his age appropriate language and communication skills. It was decided that his speech and language development would be closely monitored because the diagnosis of AN/AD puts him at risk of developing speech and language disorder. His speech production is behind his other areas of language development. He is now 2;1 years.



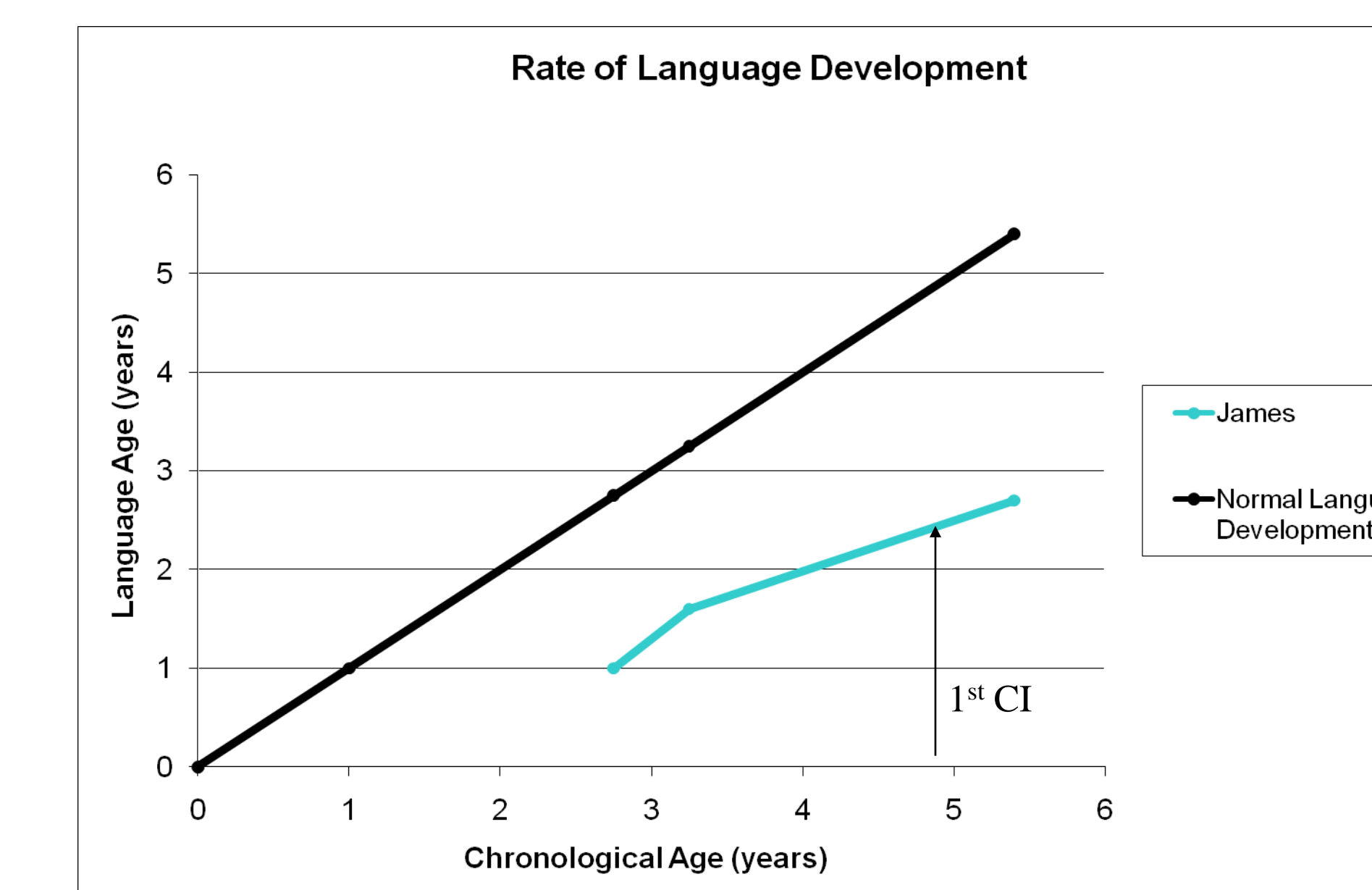
Age at diagnosis of hearing loss	8 weeks
Age at hearing aid fitting	10 weeks
Age at diagnosis of AN/AD	16 months
First contact at AVUK	16 months
Age at initial assessment	21 months
Equivalent language age	23 months

CASE STUDY 4

James passed the hearing screen at birth. He exhibited feeding difficulties and delay in development of gross motor movements. His parents first expressed concern about his responsiveness to sound aged 2;1 years. He only had a few recognisable single words. Insertion of grommets did not improve his hearing levels and hearing tests continued to show variable responses. James was fitted with hearing aids and then received a unilateral cochlear implant aged 4;9 years. He is now 5;7 years.



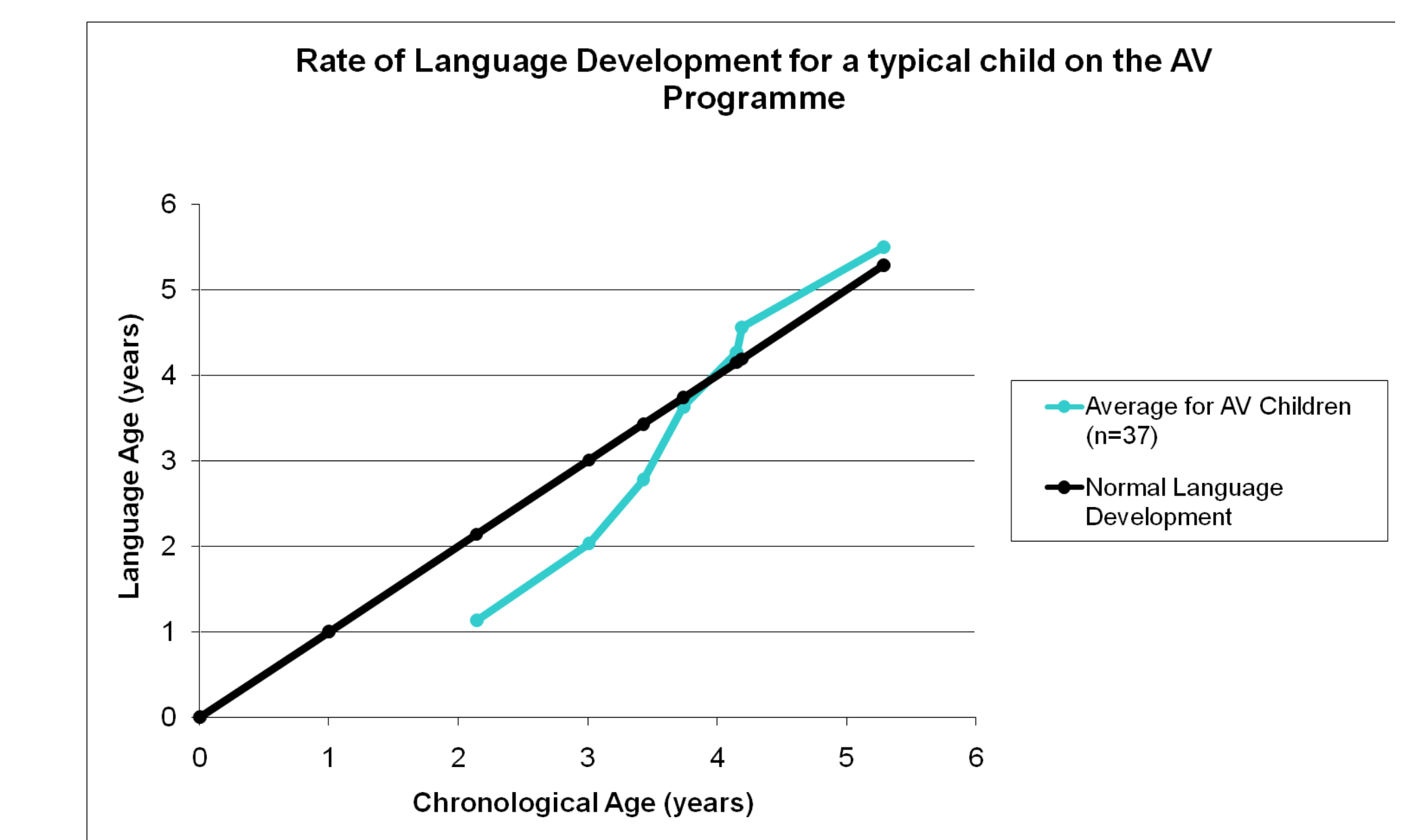
Age at diagnosis of hearing loss	2;7 years
Age at hearing aid fitting	2;8 years
Age at diagnosis of AN/AD	4;11 years
First contact at AVUK	2;9 years
Age at last assessment	5;5 years
Equivalent language age	2;8 years



CONCLUSIONS

The case studies of these four children serve to demonstrate the disparate nature of a condition in which the specific locus of the auditory processing abnormality may be unknown but for which a specific collection of audiological test results and behavioural observations lead to the label of AN/AD.

The expectation for children on an AV programme is that their rate of language progress will meet or exceed that of their hearing peers, all other things being equal.



The two children who have received CIs are both progressing in spoken language communication but the gap between themselves and their typically hearing peers is not reducing. We will continue collecting data for all of these children to monitor their rate of progress on the AV programme.

Could the age of diagnosis of AN/AD have made a difference?

For Laura, the limiting factor may be her cochlear nerve deficiency whereas for James, his outcomes may have been better at this stage had his AN/AD been diagnosed earlier. The NDCS Quality Standards in Paediatric Audiology Vol IV (2000) states that for children with permanent childhood hearing loss (PCHL) the following should be done at the same time or shortly after ABR testing:

- ME impedance and stapedius reflex testing
- Recording startle and auro-palpebral reflex (APR) activity

To the best of our knowledge, APR activity is not routinely investigated in the UK. If APR activity could act as an indicator for possible AN/AD then we should be conducting this test for children with PCHL.

Rance G (2005) Trends Amplif 9: 1-43 Auditory Neuropathy/dys-synchrony and its perceptual consequences.