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Review

What can we expect of normally-developing children implanted at a young age with respect to their auditory, linguistic and cognitive skills?

Astrid van Wieringen*, Jan Wouters

KU Leuven – University of Leuven, Dept Neurosciences, Experimental Oto-rhino-laryngology, Herestraat 49, Bus 721, Leuven, Belgium

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ABSTRACT

As a result of neonatal hearing screening and subsequent early cochlear implantation (CI) profoundly deaf children have access to important information to process auditory signals and master spoken language skills at a young age. Nevertheless, auditory, linguistic and cognitive outcome measures still reveal great variability in individual achievements: some children with CI(s) perform within normal limits, while others lag behind. Understanding the causes of this variation would allow clinicians to offer better prognoses to CI candidates and efficient follow-up and rehabilitation. This paper summarizes what we can expect of normally developing children with CI(s) with regard to spoken language, bilateral and binaural auditory perception, speech perception and cognitive skills. Predictive factors of performance and factors influencing variability are presented, as well as some novel data on cognitive functioning and speech perception in quiet and in noise. Subsequently, we discuss technical and non-technical issues which should be considered in the future in order to optimally guide the child with profound hearing difficulties.

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1. Introduction

Newborn hearing screening and subsequent early intervention with a cochlear implant have provided children with profound hearing impairment access to sound at a very early age. Newborn hearing screening has been implemented in Flanders (Belgium) since 1998 and assesses, on average, 96% of newborns (Van Kerschaver et al., 2012; Desloovere et al., 2013). This high coverage is important since 1.2 to 2.05 per 1000 infants are born with bilateral hearing loss (>40 dB HL). Of these children about 35% suffer from severe to profound sensorineural hearing loss (71 - > 95 dB HL) and cannot benefit from conventional hearing aids (Van Kerschaver and Stappaerts, 2011). These infants can be treated with a cochlear implant (CI), a device which partially restores hearing by electrical stimulation of the auditory nerve. For more than 350.000 profoundly deaf persons, of whom 80.000 children, the CI is a life changing opportunity (e.g. O'Donoghue, 2013; Kral and O'Donoghue, 2010). In Flanders, 93% of children with severe to profound hearing loss receive CIs at a young age (De Raeve, 2010). Since 2010 two CIs are fully reimbursed by The Belgian National Health Insurance Institute for children under 12 yrs of age (one CI for adults). The ultimate aim of pediatric cochlear implantation is to enable deaf children to have good access to sound, achieve good spoken language levels, and to provide optimal opportunities for social and academic development in an oral society.

Yet, despite improved accessibility to sound, the large variability in outcomes remains a significant concern, even for those children who have normal intellectual abilities (non-verbal IQ > 80). Although some children achieve adequate language levels, others lag behind (e.g. Svirsky et al., 2000; Niparko et al., 2010; Peterson et al., 2010; Ching et al., 2013; Hess et al., 2014). Many of the 'good' performers do not achieve normal speech perception levels and most CI users experience severe difficulties when performing

Abbreviations: BMLD, binaural masking level differencel; CI, cochlear implant; CVC, consonant – vowel-consonant; dB, decibel; IQ, intelligence quotient; LQ, language quotient; MAE, mean absolute error; RDLS, Reynell Developmental Language Scales; SELT, Schlichting Expressive Language Test (_W = word, _S = sentence); SNR, signal-to-noise ratio; SRM, spatial release from masking; SRT, speech reception threshold; WISC III, Wechsler Intelligence Scale for Children

* Corresponding author. Tel.: +32 16 330478; fax: +32 16 330486.
E-mail address: astrid.vanwieringen@med.kuleuven.be (A. van Wieringen).

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more challenging listening tasks. The aim of this paper is to summarize what we can expect of normally developing children with CI(s) with regard to spoken language, bilateral and binaural auditory perception, speech perception, and cognitive skills.

The topics addressed in this paper are based on data from two multicenter studies with Dutch speaking children and studies reported in the literature. Very often clear interpretation of the literature is difficult because of the limited sample size, and other methodological limitations, such as lack of matching of subject groups, or lack of sensitive outcome measures which, for example, do not tap into language sub skills or do not reflect speech in noise difficulties. The research group in Leuven has performed two multicenter studies with Dutch speaking young implanted children. The first study dealt with the auditory, spatial and binaural abilities of young children with two CIs, at a time when 2 CIs were not common practice yet (2004–2009, Van Deun et al., 2009, 2010a,b). The 2nd study investigated spoken language development in a contemporary group of young implanted children with one or two CIs (2009–2013, Boons et al., 2012a,b; 2013a,b,c). In the 2nd multicenter study all 288 Dutch speaking children received their first CI before the age of 5. Nowadays, many of them attend mainstream school and are eventually expected to achieve educational and employment levels similar to those of their normal hearing peers (e.g. Venail et al., 2010). However, although they are generally doing very well, most children lag behind with regard to spoken language, auditory and cognitive abilities, and experience large difficulties understanding speech, especially in more challenging listening tasks. We will first address some issues which are known to influence performance, and then suggest other technical and non-technical factors which should be better considered more in the future to optimally guide the child with profound hearing difficulties. This is by no means an exhaustive review; we offer our thoughts to professionals worldwide who deal with children with hearing impairment.

2. Early implantation, and bilateral input

Beyond doubt newborn hearing screening has a great positive impact on the development of communication abilities in the profoundly deaf child. Early diagnosis and early cochlear implantation are very important for the development of auditory processing, spoken language, reading, and other academic skills (e.g. Archbold et al., 2008; Niparko et al., 2010; Tajudeen et al., 2010; Yoshinaga et al., 2010; Boons et al., 2012b; Grieco-Calub and Litovsky, 2012; Tobey et al., 2013; Ching et al., 2013; Dettman et al., 2013; Leigh et al., 2013). Early access to sound enables 'early neural wiring' in the brain, which is advantageous for auditory and cognitive development, and which, in turn, will facilitate onset of babbling, a precursor for the development of spoken language (e.g. Schauwers et al., 2008; Moreno-Torres, 2014). Although the ability of the brain to adapt as a result of experience persists throughout life, certain changes are more predominant during the first few years of life (e.g. Kral and Sharma, 2012). In the following sections we will discuss which factors translate to better language, auditory and speech perception skills, and effects of bilateral input.

2.1. Spoken language

Communication through language is vital to humans. While some children with hearing impairment learn to sign, the vast majority wants to communicate through oral language. In order to be able to fully participate in society, these children need to acquire full competence in spoken language, both receptively and expressively. This will also lead to better reading and writing skills.

The Reynell Developmental Language Scales (RDLS), a verbal comprehension test, and two tests of the Schlichting Expressive Language Test (SELT) were administered to 288 children in 5 Dutch speaking CI centers in Flanders and the Netherlands. The RDLS and SELT provide norm-referenced scores, as well as age-equivalent scores, which are based on typical language levels for normally developing children with normal hearing. The age-equivalent score can be divided by the child's chronological age to calculate a language quotient (LQ), which indicates the ratio between the expected level of performance based on the chronological age and the actual performance of the child. An LQ close to 1.00 indicates an age-appropriate language level, and an LQ of 0.50 corresponds to a delay of half the chronological age. All children received their 1st CI before the age of five. In order to examine whether 2 CIs translate to better spoken language, 25 children with 1 CI were matched with 25 children with 2 CIs on 10 factors (Boons et al., 2012a,b). These children were selected from the sample of 288 children.

Mean LQs for the receptive RDLS and expressive SELT word development (SELT_W), and SELT sentence development (SELT_S) at 3 years post cochlear implantation are presented in Fig. 1. Each test illustrates four conditions: 1) the language quotients of the entire sample ('all'), 2) of the children who received their 1st CI under 2 years of age (CI < 2 yrs), 3) of 25 children with 2 CIs (match_bi), and 4) of 25 carefully matched children with 1 CI (match_un). All boxes demonstrate large and similar variability regarding performance on the three tests. However, children implanted before the age of two perform significantly better on all three tests than children who were implanted at an older age ($p < 0.001$). Also, the bilaterally implanted children outperform the unilaterally implanted ones on receptive and expressive skills (Boons et al., 2012a). Still, even these children do not approach the age equivalent score.

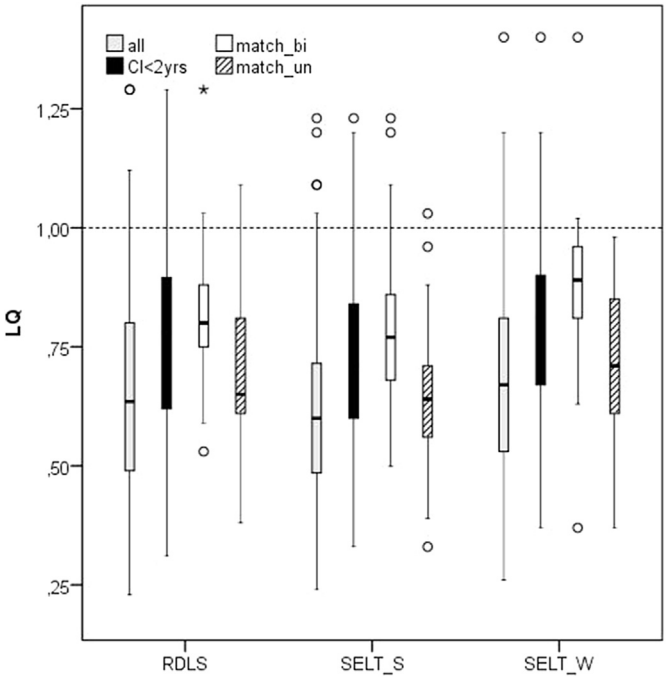

Fig. 1. Language quotients of the RDLS, SELT_S and SELT_W 3 yrs after implantation. RDLS, Reynell Developmental Language Scales; SELT_W, Schlichting Expressive Language Test—word development; SELT_S, Schlichting Expressive Language Test—Sentence development. For each language test the Language quotients of the entire set is given ('all', $n = 288$), of the children who received their CI before the age of 2 (CI < 2 yrs), of the bilaterally implanted children (match_bi) and their matched peers with unilateral CI (match_un).

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