

Language Acquisition in Very Young Children with a Cochlear Implant: Introduction

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Abstract

This symposium aims at sharing the preliminary data that are available on language acquisition in very young children with a cochlear implant. Congenital sensorineural hearing loss occurs in approximately 2 per 1000 newborns and results in significant and often irreversible retardation in the development of speech and language. Early detection is possible thanks to universal hearing screening programs. Early intervention consists of hearing aids or cochlear implants in case of severe losses followed by intensive (re)habilitation. Hearing aids are provided at ages as young as 3 months, but cochlear implants are not yet provided routinely before the age of 2 years. A trend however exists towards younger implantation, even before the age of 1 year, and it is anticipated that this will significantly influence the speech and language development of these children. Preliminary data are available on the audiological outcome of very young implantation and they will be presented. In addition, it is also important to assess the speech and language development of these children and to compare this with both normally hearing children and hearing impaired children with a hearing aid during the first years of their life. An attempt will be made to define relevant outcome measures in terms of speech and language development and some first results will be presented.

Congenital hearing loss

Congenital bilateral sensorineural hearing loss (>30 dB HL) occurs in approximately 1.2 to 3.2 per 1000 live births (Watkins 1991, White 1993, Mauk 1993, Parving 1993, Davis 1994, Northern 1994, Fortnum 1997, Stein 1999). This hearing loss is permanent and results in significant delay in speech and language development and consequently in important integration problems in the mainstream educational system (Brannon 1966, Davis 1974, Davis 1986, Andrews 1991, Geers 1989). Deaf-mutism is the most extreme consequence and this has been part of all cultures in human history. Until recently, no other therapy than hearing aids existed. Because of factors that will be discussed later, even hearing aids were unable to restore hearing sufficiently to prevent these severe consequences of congenital deafness.

This situation has dramatically changed in the last decade. The reason for this is the development of cochlear implants in the late seventies. These are implantable electronic devices that aim at replacing the cochlear function. Initially these implants were used to restore hearing in elderly patients with acquired deafness. With time, and encouraged by improving results and technology, the field of indications broadened towards younger patients and lower degrees of hearing loss. Initially congenital (or “prelingual”) deafness was considered a relative contraindication for cochlear implantation, because it was observed that these persons with severe speech and language retardation hardly improved after the intervention. However it was felt by many professionals in the field that cochlear implants could have significant impact on the speech and language development if they could be implanted at sufficiently young an age, meaning before the onset or at a very early stage of the linguistic development. For this to become possible, it would be crucial to detect congenital hearing losses at a very early stage and to develop proper diagnostic tools to gain certainty about the type and degree of hearing loss.

Fortunately and in parallel with the development of cochlear implants, new techniques became available to easily detect hearing losses in newborns. These techniques were based on the otoacoustic emissions that were discovered as a physiological entity in the late seventies (Kemp 1978). Commercial equipment became available in the late eighties and this was the incentive to start thinking of universal neonatal screening programs in order to detect all congenital hearing losses immediately after birth (White 1993). To date, universal neonatal hearing screening is a fact in several regions in the world. Infants with congenital hearing loss are receiving elaborate diagnostic work-up and they typically receive their first hearing aids by the age of 3 months. Audiological tools exist that allow early selection for cochlear implantation, which can be safely done before the age of 1 year.

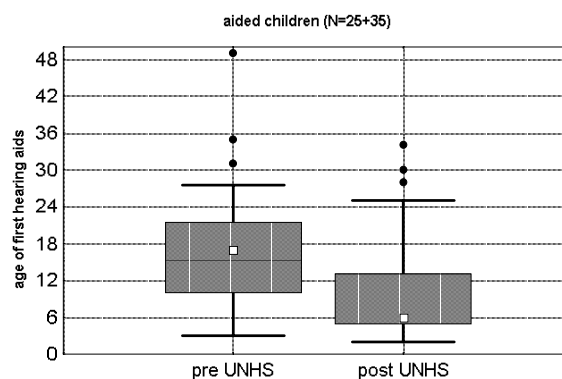


Figure 1. Results of a study of one birth cohort just before (“pre UNHS”) and another just after (“post UNHS”) the introduction in 1998 of universal neonatal hearing screening in Flanders, showing the decrease of the age at which the hearing impaired children received their first hearing aids. Both graphs are box and whisker plots in which the whiskers represent the lower and upper extremes (P0 and P100), the box the lower and upper quartile (P25 and P75) and the central dot the median (P50). (PJ Govaerts, unpublished results)

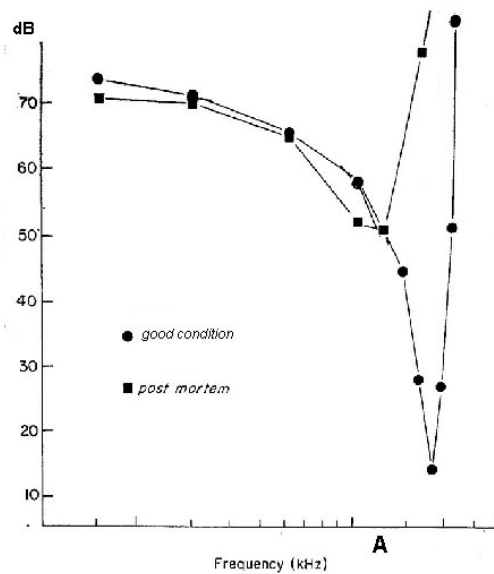


Figure 2. Typical tuning curve of a cochlea in good physiological condition. When compared to the tuning curve post mortem, it is clear that the sensitivity has increased (lower thresholds, especially near the characteristic frequency) and that the tuning has become sharper. This is the result of the active outer hair cell mechanism (figure modified after Sellick 1982).

A congenital sensorineural hearing loss is almost always characterized by a malfunctioning cochlea. The two major functions of the normal cochlea are (1) amplification and (2) frequency-resolution. This is expressed by the tuning curve (figure 2). In case of sensorineural hearing loss, the outer hair cells are virtually always affected. Only in rare cases of isolated retrocochlear types of hearing loss, this may not be the case. If the outer hair cells are affected, the tuning curve shows a higher threshold and a broader tip. The higher threshold results in an elevated threshold on pure tone audiometry. The broadened tip results in a lower frequency resolving power of the cochlea, which is more difficult to assess in the clinical setting. But a good frequency resolving power is essential for normal speech and language development. So this is the key problem in hearing impairment and it is the link between the hearing loss and the speech and language retardation.

Conventional hearing aids unfortunately don't interfere with the tuning, they only amplify the sound. Figure 3 shows how this affects the tuning curve. The result for the patient is that with a hearing aid the detection level of sound decreases but that the frequency resolution of his hearing does not really improve. The patient will therefore report to hear sound better with a hearing aid, without necessarily better understanding the words.

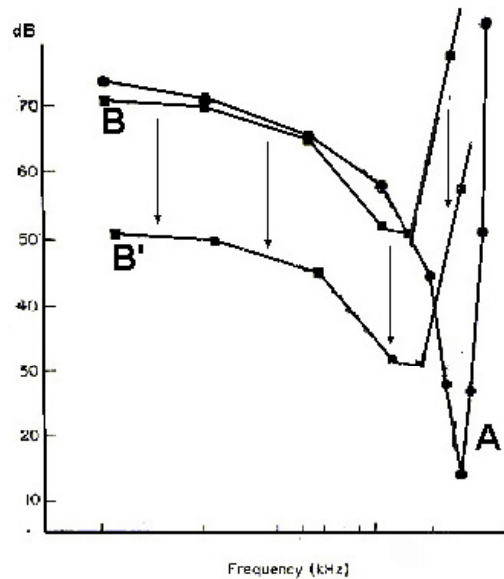


Figure 3. Effect of a hearing aid on the tuning curve of a cochlea. This Figure is based on Figure 2. The sharply tuned curve (A) is the typical curve of a normal cochlea. Curve B is of a hearing impaired cochlea with elevated thresholds and broad tuning. With a hearing aid, this curve shifts downward but the shape does not change (B'). The effect is that the cochlea will detect sound at lower levels, but that the frequency resolving capacity of the cochlea does not improve.

Cochlear implants in contrast not only amplify the sound, but they also aim at a (partial) restoration of the frequency resolution of the cochlea. This is achieved by the spatial selectivity of the stimulation at different points in the cochlea. A cochlear implant has an electrode array with multiple electrode contacts. The Nucleus® 24 device (from Cochlear Ltd, Australia) will be described to illustrate this. This implant has 22 intracochlear and 2 extracochlear electrodes. Different stimulation modes are possible, of which the monopolar mode is commonly used. This means that the current flows between the intracochlear and the extracochlear electrodes. In consequence the spatial current spread at the site of the electrode is small and results in a local stimulation of the cochlear nerve. The smaller the spatial spread, the more selective the stimulation will be. This should be reflected in the tuning curves.

With a cochlear implant, tuning curves not only show better thresholds, they also show remarkably fine tuning (Figure 4). This is the major advantage of a cochlear implant over a hearing aid. Hearing aids are doing fine as long as the hearing loss is not too severe and cochlear tuning is still acceptable. In such a case, amplification alone is sufficient. If dynamic compression strategies (e.g. wide dynamic range compression), noise suppression paradigms and other quality-improving features are added, modern -often digital- hearing aids may serve the moderately to severely hearing impaired patient well. But as soon as the cochlear tuning becomes deficient, amplification alone doesn't suffice any longer and cochlear implants may yield better results.

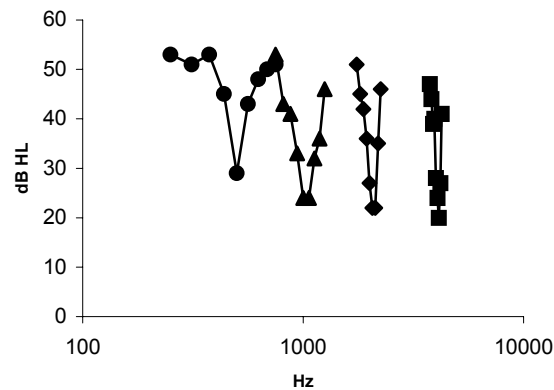


Figure 4. Psychoacoustic tuning curves of a patient with a Nucleus® 24 multichannel cochlear implant in monopolar stimulation mode. Tuning curves with probe frequencies at 500, 1000, 2000 and 4000 Hz and a simultaneous masking paradigm are shown. It can be inferred from the figure that the thresholds are approximately 25 dB HL and that the tuning is sharp (Govaerts 2002a).

How to assess the frequency resolving power of the cochlea?

One of the challenges in handling the pediatric hearing-impaired population is the assessment of hearing. Pure tone audiometry, otoacoustic emissions, automated brainstem audiometry etc. only assess hearing at its detection level. This may be sufficient to know whether a hearing problem exists or not, but it hardly reflects the capacity of the hearing impaired child to discriminate or identify language. So far, too little attention has been given to the fact that hearing impairment means both an increase in detection threshold and a loss of frequency discrimination. In consequence, improving the detection threshold to “within the speech zone” (e.g. 40 dB) does not imply that the aided subject also discriminates the phonemes presented at or above this sound level. Although this limitation has always existed, cochlear implants have forced us to look for supraliminal evaluation techniques. These are needed both in the selection of cochlear implant candidates and the evaluation of cochlear implantees. Supraliminal features of hearing are discrimination and identification of sounds. Tests for discrimination or identification of words and sentences exist, but especially in the preverbal child the results are strongly biased by their individually variable language impairment or cognitive skills. A “preverbal” child is a child with no or very limited functional speech, both comprehensive and productive. Normal hearing children use to become verbal by the age of 1 year (Barrett 1994, Gillis 2000). In hearing impaired children this age is very variable. It depends on the level of hearing loss and the type and intensity of stimulation. Their preverbal stage may typically last till the age of 4-5 years. Tests for this “preverbal” population are difficult and should be conceived in such a way that the dependence on the child’s linguistic and cognitive skills is minimal and that no reading and speech skills are required. Furthermore, the distinctive features

should be very clear and unambiguous so as to leave no doubt which features are perceived by the child and which are not (Boothroyd 1997). At least some of the tests should provide the fitter with phoneme-based analytical information to guide the fine-tuning of the cochlear implant.

A common way to investigate auditory performance is the identification test. Identification tasks presuppose a degree of linguistic knowledge and higher functions that are not always present in the hearing impaired child. Thus most of the existing identification tests are only fit for verbal children. In normal hearing children they are feasible from the age of 2-3 years onwards but in deaf children or children with additional problems in language development they cannot be done at this young age.

Another and possibly more correct way to test preverbal children with minimal bias related to the level of linguistic development is testing discrimination instead of identification. No knowledge of the stimulus is required. The child has to discriminate between two or more successive stimuli and has to show a behavioural response (Dillon 1995, Bochner 1992). An additional advantage of discrimination tests as part of a test battery is that they allow for the assessment of the cause of systematic confusions as they may occur in identification tests. The Auditory Phoneme Evaluation (APE®, Melakos nv, Antwerp, Belgium, www.melakos.net) is an audiological evaluation tool that uses strictly defined phonemes as stimulus material for detection, discrimination and identification tests. The APE® was designed as a language-independent test to yield supraliminal information on the auditory function with as little cognitive bias as possible. The main purpose of the test is to evaluate the discriminatory power of the cochlea of very young, preverbal hearing-impaired children with hearing aids.

The phoneme discrimination test of the APE® is an oddity test in which two phonemes are presented and the infant is conditioned to react to the odd phoneme. Table 1 shows the basic set of the phoneme pairs as routinely used by the authors and coworkers in the assessment of the cochlear function.

The discrimination test of the APE® is routinely used by the authors to evaluate the cochlear function in hearing impaired children and adults. Infants as young as 7-8 months can be tested. As a measure of the frequency resolving capacity of the aided cochlea (with hearing aids), it has become an essential tool in the selection of cochlear implant candidates. If the patient fails to discriminate on several phoneme pairs, it is anticipated that his/her discrimination will be better with an implant. If all 22 phoneme pairs of the basic set are assessed, discrimination of less than 19 pairs is an indication to consider cochlear implantation. If only the minimal set of 7 phoneme pairs is assessed, discrimination of less than 6 is an indication to consider cochlear implantation. The phoneme pairs that are often the first fall-outs in hearing aid wearers, are /z/-/s/, /m/-/z/, /u/-/l/ and /v/-/z/. Obviously the phoneme discrimination is not the only selection criterion for cochlear implant and the results should be combined with other audiological and other results before a final decision is made.

a-r	l-E
u-ʃ	œ -E
u-l	œ -l
l-a	y-l
u-a	u-y
o-a	z-s
u-o	m-f
œ -a	m-z
œ -u	m-r
œ -o	s-ʃ
E-a	v-z

Table 1. “Basic set” of phoneme pairs of the APE®. The first phoneme of a pair is presented as the background phoneme and the second as the odd phoneme. The black fields represent the phoneme pairs of the “minimal set”.

Cochlear implantation before the age of 2 years

Thanks to new selection tools and to the ever-improving performance of the implants, the number of implants performed worldwide has increased exponentially over the years (Fig 5).

The first children below 6 years of age at our department were implanted in 1994, below 2 years in 1996 and below 1 year in 2000. This steady shift in the age of implantation (Figure 6) has resulted in a significant improvement in outcome.

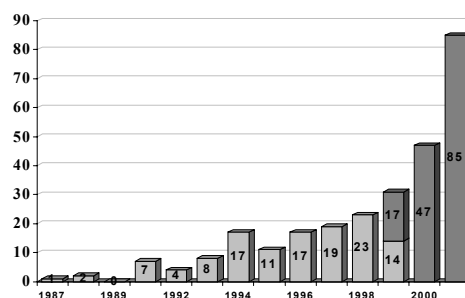


Figure 5. Annual number of cochlear implants at the University ENT Department of the St.-Augustinus Hospital. Light grey: LAURA™ implants; dark grey: Nucleus® 24 implants

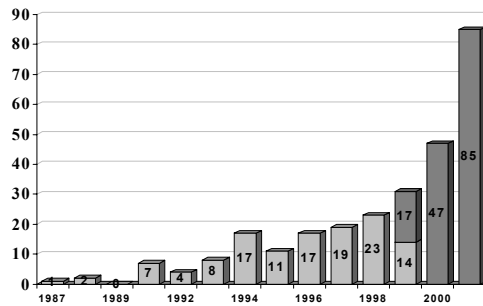


Figure 6. Age distribution of the cochlear implantees at our department for the period 1995-2000. The small figure zooms in on the youngest group (less than 5 years) and it shows that the age distribution in this group has clearly shifted to the younger than 2-year-old children in 2000. (Govaerts 2002a).

Briefly, in children with congenital severe to profound hearing loss, implantation above the age of 4 years gives a moderate auditory performance (even in the long run) with only 33 % of the children being able to integrate in the mainstream educational system. Implantation between 2 and 4 years of age gives good auditory performance, be it with a significant delay of 2-3 years, and a mainstream integration in two out of three. Implantation at 12-18 months gives immediate high auditory performance with an integration rate of 90 % already in the first year of the kindergarten (Figure 7 and Table 2).

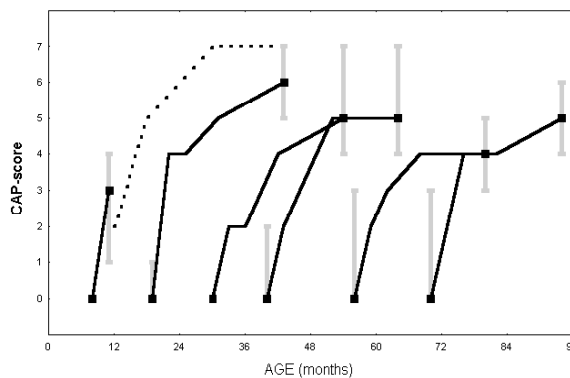


Figure 7. Results of a longitudinal study with the consecutive median CAP-scores (categories of auditory performance, Archbold 1995 and 1998) for six age cohorts defined by the age of implantation. Five cohorts have a follow-up of two years. For each cohort, the range of the CAP-score is

given preoperatively and 2 years postoperatively. The dotted line is the median CAP-score of the control group. (Govaerts 2002b)

Age group	Age (with range of first hearing aids (months))	Mainstream integration (%)	Age of integration (months)
0	2 (1-4)		
1	7 (3-12)	67 (89)	37
2	13 (9-21)	57 (63)	67
3	13 (3-32)	23 (54)	96
4	15 (10-37)	17 (33)	79
5	20 (10-44)	14 (14)	84

Table 2. Integration of cochlear implantees. The figures in the third column refer to the percentage of children that have been integrated in the mainstream school system so far. The figures between brackets are the same ones plus those that are anticipated to be able to integrate in the near future. (Govaerts 2002b)

Conclusions

In conclusion, the age of implantation in congenital hearing loss is decreasing. The selection criteria are shifting thanks to new evaluation tools. Evidence is being built up of the audiological outcome of young implantation and it seems that implantation before the age of 18 months has advantages. Time has come to assess the outcome in terms of speech and language development.

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