Normal Hearing and Language Development in a Deaf-Born Child


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Background: Congenital deafness leads to major problems in speech, language, education, and social integration. Neonatal hearing screening and cochlear implantation now allow early hearing restoration. This article reports on a prospective longitudinal study of the first infant ever who received two cochlear implants in the prelexical period of her life.

Methods: The first deaf-born girl ever who received two implants at the ages of 5 and 15 months, respectively, was followed-up with repeated and detailed quantitative assessments from birth to 4 years of age. This consisted of 1) audiologic evaluation (audiometry, speech audiometry, and Categories of Auditory Performance score), 2) linguistic evaluation (monthly video analyses and tests of vocabulary, language skills, grammar, and intelligibility of the child’s speech), and 3) descriptive assessment of the educational setting.

Results: All results lie within the 95% confidence interval of hearing peers. The audiologic performance lies at or above average from age 2 years onward. The child started babbling at the normal age of 8 months. Her linguistic skills increased from low percentiles before age 2 to above average from age 2 for comprehension and from age 3 for production. The grammar and intelligibility of the child’s speech increased from low percentiles to average at age 4. The girl entered preschool at the normal age of 2.5 years, and this with only very limited special assistance.

Conclusion: This case illustrates the fact that congenital deafness no longer has to lead to abnormal hearing and abnormal speech development. It opens the debate of the ethics of not implanting a deaf child in the first few months of life. Key Words: Bilateral cochlear implantation—Congenital deafness—Hearing impairment—Speech development.

pends on early auditory input. A child’s prelexical babbling is in part determined by the auditory input during the first months of life (13). The vocal utterances of deaf infants are clearly different from those of their hearing peers (13,14), and some of the linguistic delays and differences found in deaf children also seem to be irreversible (15). Therefore, if CIs are to have a true impact on a congenitally deaf person’s hearing, speech, and language capabilities, it can be anticipated that they have to be implanted at a very young age (16).

Thanks to universal neonatal hearing screening programs that have been implemented in many states and countries during the last decade (17,18), children with congenital deafness can now be identified immediately after birth. For the first time ever, this allows intervention at the beginning of the infant’s life and thus at the very beginning of speech and language development. Many cochlear implant centers around the world have lowered the age limit for implantation to 2 years of age or even below (19,20). These policies are based on the growing body of evidence, though often indirect and circumstantial, of the benefit of early implantation. Evidence is accumulating that implantation below the age of 2 may prevent the irreversible problems encountered in terms of audiology, speech and language development, and educational functioning of children implanted at a later age (21).

This article reports on a girl with congenital deafness who received a CI in the right ear at the age of 5 months (i.e., before the normal onset of prelexical babbling in hearing children). She was also implanted at the other side at 15 months of age. To the best of our knowledge, this is the youngest implantee ever. The girl has now been followed-up for 48 months at monthly intervals, and detailed results are available on her hearing (i.e., sound perception), speech and language development (i.e., language understanding and production in communicative settings), and educational setting (i.e., integration into mainstream education).

PATIENT AND METHODS

The patient was born in May 2000 after an uncomplicated pregnancy. She has an older sister who was born with congenital deafness of unknown origin in 1997 and an older hearing brother who was born in 1998. The sister received a cochlear implant at the age of 13 months and a second one at the age of 25 months. The patient did not pass the neonatal hearing screening consisting of two consecutive tests for otoacoustic emissions (22). She was then referred for diagnostic workup. Auditory brainstem responses showed no responses to clicks of 120 dB hearing loss (HL). Bilateral hearing aids were fitted at 3 months of age, but aided thresholds were hardly better (110 dB HL) than the unaided ones. On the basis of the good experiences with the older sister, the parents decided to have the patient implanted as soon as possible. She received an implant in her right ear at the age of 5 months and in her left ear at the age of 15 months. Both implants were programmed using an automated method developed by the senior author (P. J. G.) (method for automated fitting of cochlear implants, obtained cochlear implant and computer programs therefore: European patent application No. 02447028.8 [February 21, 2002], U.S. provisional patent application No. 60/361,385 [February 28, 2002]). The patient was vaccinated against meningitis according to the Food and Drug Administration recommendations (summarized by Cohen et al. (23)). After informed consent of the parents, the patient was enrolled in a longitudinal prospective study to monitor her audiologic, linguistic, and educational development.

Audiologic development

The patient’s hearing was regularly assessed by means of several tests: pure-tone audiometry (behavioral audiometry, yielding hearing threshold shifts expressed in decibels of HL), speech audiometry (yielding word scores, i.e., the percentage of correctly identified words from an open-set word list), and Categories of Auditory Performance (24). The latter test is a global outcome measure of auditory functioning consisting of a nonlinear hierarchical scale along which the parents and a professional therapist rate the infant’s developing auditory abilities according to eight categories of increasing difficulty, with category 7 “use of telephone with a familiar talker” as the highest one. For all audiologic tests, values in hearing children at different ages are available and have served as controls.

Linguistic development

In order to monitor the patient’s linguistic development, we relied on monthly video recordings. Each of these lasted approximately 80 minutes and were made starting from the first month after activation of her first implant (i.e., at 7 mo of age). Selected video segments were transcribed in a very detailed manner. The patient’s prelexical utterances were coded according to the sensorimotor description model for early infant vocalizations developed by Koopmans-van Beinum and van der Stelt (25). Because babbling is a major landmark in prelexical speech development, both its onset and its quality were defined and analyzed (26). In particular, the quality of the babbles was analyzed in terms of manner of articulation (stops, nasals, glides, fricatives, and liquids). For further technical details on the video recordings and their analyses, see Schauwers et al. (26). As soon as the patient started producing conventional words and short sentences, we were able to administer the following formal tests:

1. The Dutch adaptation of the MacArthur Communicative Development Inventories (27,28): The Communicative Development Inventories assesses the vocabulary skills between 8 and 30 months of age, based on parental reports, and yields age-specific percentile values that can be compared with those of a normalized hearing sample; for this study, only the productive skills were assessed.
2. The Dutch adaptation of the Reynell Developmental Language Scales (29,30): The Reynell Developmental Language Scales evaluates receptive and expressive language (mainly vocabulary and syntax) from 2 to 5 years of age, based on item scoring by speech and language therapists, and yields percentile scores for comprehension and production separately.
3. The Language Assessment, Remediation and Screening Procedure (31) adapted for Dutch (taal analyze remediering en screening procedure) (32): The Language Assessment, Remediation and Screening Procedure analyzes morphosyntax from 1 to 4 years of age. It is scored by speech and language therapists and yields categorical levels corresponding roughly with age-specific percentiles, 10, 50, and 90%.
4. The Speech Intelligibility Rating (33): The Speech Intelligibility Rating assesses the intelligibility of the child’s speech as rated by speech and language therapists and yields a hierarchy ranging from unintelligible speech (rating 1) to speech that is intelligible to all listeners (rating 5) that can be translated to age-specific quartiles (0–25%, 25–50%, 50–75%, and 75–100%).

For all tests, normality was defined as the 95% confidence interval (between 2.5% and 97.5%) as found in normally hearing age-matched controls.

**Educational development**

Hearing children in Belgium usually attend a daycare center from the age of approximately 3 months onward and enter preschool at the age of 2.5 years. For hearing-impaired children, special facilities exist at all educational levels. In the preschool years, hearing-impaired infants are typically enrolled in a multidisciplinary program consisting of family guidance at home, auditory stimulation and training, speech and language therapy, and physiotherapy or ergotherapy. When deaf children attend school, whether it is a mainstream school or a special school, speech and language therapy continues for approximately 2 hours per week (depending on the degree of hearing loss). Below, we describe the patient’s educational setting.

**RESULTS**

**Audiologic performance**

Table 1 shows the audiologic data at different moments and in different conditions. The pure-tone averages (PTAs) (or the mean of the thresholds at 500, 1,000, and 2,000 Hz) were compared with the average and standard deviation in hearing children as given by Hodgson (34) and percentile values were calculated accordingly.

Before implantation, the PTA in the best-aided condition with two hearing aids was not better than 110 dB HL, thus corresponding to an extremely low percentile. With bilateral implants, PTAs improved almost immediately. The patient obtained thresholds within the 95% confidence interval of hearing children (2.5–97.5%) immediately after the first implantation. When she reached 3 years of age, her thresholds were close to the average thresholds of hearing peers.

Speech audiometry cannot be performed before the age of approximately 3 years. Hearing children obtain a 90 to 100% word score at 40 dB sound pressure level.

Table 1 shows that the patient obtained near to normal scores at the same age as hearing peers. Table 1 also shows the Categories of Auditory Performance scores at different ages. The results of the patient always fell between the 25th and 50th percentiles of the hearing children. The patient reached the highest score at 3 years of age.

**Speech and language development**

**Prelexical babbling**

Onset of babbling The average age at onset of babbling in hearing children is 30.8 weeks (standard deviation, 6.3 wk; 95% confidence interval, 18–43 wk) (25). The patient started to babble at 8 months of age or 2 months after activation of her first implant. This corresponds to the 28th percentile of normal hearing children.

Quality of babbling Figure 1 shows the percentage of occurrence of six consonant types (stops, nasals, glides, fricatives, and liquids) up to the appearance of the first word(s) in our group of 10 normally hearing children and in our CI patient. It is commonly taken for granted that the quality of babbling between hearing and hearing-impaired children differs most in the stops and the fricatives (35), where hearing-impaired children produce less stops and more fricatives. In addition, Van Hapsburg also claims that hearing impaired children produce more nasals and less glides. For all these distinctive features, our patient tends to show more normal preferences, with more stops and glides and less nasals and fricatives. It is unclear how to explain the high occurrence of stops (83%). Similar analyses on more CI children will have to show whether this is a consistent finding or not.

**Lexical development**

Table 2 lists the patient’s scores on the different linguistic tests at consecutive ages. Her lexical development was within normal ranges from the earliest recordings onward. However, it remained below median until 2 to 3 years of age. From 3 years onward, both her receptive and productive vocabularies were beyond the median of hearing peers. The patient’s grammatical development and her speech intelligibility were normal from the first recordings onward but only reached the 50th percentile.
at age 4 years. These combined findings show that at age 4, the patient’s language was rich and expressive in terms of vocabulary, language content, and morphosyntax. The patient’s language use compares very favorably with that of her normally hearing peers.

Educational setting

In line with the standard procedures of good clinical practice, the patient was enrolled in an early (re)habilitation program as soon as her hearing loss was discovered. This program consisted of family guidance and four home sessions per week of 30 minutes for hearing (re-)habilitation and speech stimulation. The patient was raised orally with support of a limited number of Dutch signs. At the age of 2 years 8 months, she was successfully integrated in the first preschool class of a mainstream school. Besides 4 hours per week of additional assistance by an educational therapist at school, the patient continued to receive 2 hours of speech and language therapy, auditory rehabilitation, and physiotherapy or ergotherapy at home.

DISCUSSION

In this article, we have presented a case study of a deaf-born girl who at the moment of implantation was the youngest person to receive a CI. She was first implanted at the age of 5 months and received a second implant at the age of 15 months. We followed-up the child longitudinally until the age of 4 years. At these young ages, children are hard to test, and test results are known to vary significantly with age. This is why we opted for the combination of many tests and the expression of test results in terms of percentile values referring to the normal population.

The results show unequivocally that very early cochlear implantation in the prelexical period of the infant is able to restore hearing to normal levels. This resulted in normal speech and language development and a normal integration into the mainstream educational system. This is in contrast to the results obtained in profoundly hearing-impaired children wearing hearing aids, who hardly ever reach aided thresholds with PTA levels of 30 to 40 dB HL or word scores beyond 40%. Profoundly hearing-impaired children typically show a substantial delay in the onset of babbling. Most of them do not start babbling before the age of 18 months (13,14,36), and the quality of their babbles differs significantly from their hearing peers (Fig. 1). Also, the lexical development of profoundly hearing-impaired children is known to be significantly impaired, with typical rates of language growth of approximately 50%, meaning that at the age of 4 years, the lexical development averages around the stage of a 2-year-old hearing peer (37).

Although the child in our case study shows “normal” results on all reported items, her scores lie in the low percentiles on almost all aspects tested until she reaches 3 to 4 years of age. This initial temporary delay probably results from the initial auditory deprivation. Similar analyses with more subjects are needed to further explore this issue.

Other reports in the literature have suggested that earlier implantation would yield better outcome in congenital deafness (38,39), but many health care systems and providers are still hesitant to implant in the first or even the second year of life. This hesitation is mainly due to

TABLE 2. Prelexical and lexical development of the patient

<table>
<thead>
<tr>
<th>Age</th>
<th>Status</th>
<th>Onset of babbling</th>
<th>Vocabulary/language</th>
</tr>
</thead>
<tbody>
<tr>
<td>0: 8</td>
<td>Implant right ear</td>
<td></td>
<td>CDI</td>
</tr>
<tr>
<td>1: 0</td>
<td>Implant right ear</td>
<td></td>
<td>3</td>
</tr>
<tr>
<td>1: 6</td>
<td>Bilateral implants</td>
<td></td>
<td>5–10</td>
</tr>
<tr>
<td>2: 0</td>
<td>Bilateral implants</td>
<td></td>
<td>60–70</td>
</tr>
<tr>
<td>3: 0</td>
<td>Bilateral implants</td>
<td></td>
<td>60–70</td>
</tr>
<tr>
<td>4: 0</td>
<td>Bilateral implants</td>
<td></td>
<td>60–70</td>
</tr>
</tbody>
</table>

a All results are given in percentiles based on norms obtained from normal hearing children acquiring Dutch.

CDI, Communicative Development Inventories; LARSP, Language Assessment, Remediation and Screening Procedure; SIR, Speech Intelligibility Rating.
the absence of evidence from large numbers of children who have been implanted at young ages and to the ensuing cautiousness with regard to the new implantation techniques. However, it is clear that nature does not allow a "wait-and-see" approach without irreversible damage to the child. Not implanting at an early stage is bound to result in severe and irreversible handicap and disability. Although the debate about possible risks of early implantation, important as it is, lies beyond the scope of this article, it seems that these risks are limited if surgery is not performed before the age of 5 to 6 months and if the meningitis vaccination schemes as recommended by the Food and Drug Administration are strictly followed (24).

CONCLUSION

The present case does not stand alone. It has already been demonstrated that implantation before the age of 2 years is beneficial to the deaf-born child (21). The single case reported here, however, is unique because of the early age at implantation, because both ears received an implant, and because of the lengthy and detailed follow-up. It shows for the first time that a child with congenital deafness may have a completely normal development in terms of hearing, speech and language, and educational integration. It is highly probable that the early intervention is the most significant factor in these unprecedented findings. Further research will have to clarify the importance of the different contributing factors. In pharmaceutical research, it is common practice to stop experiments in case intermediate results strongly suggest an important and irreversible benefit of one therapeutic strategy compared with the other(s). In analogy to this, and in view of the accumulating scientific indications in favor of very early implantation that is supported by the hard evidence of the present single case, it is worth considering the ethics of not implanting a deaf-born child in his or her first year of life. The responsibility of caregivers deciding to postpone implantation until ages 2 to 4 years or even later is tremendous. Implantation at 4 years definitely leads to irreversible damage, with hardly one child in three ever being able to integrate into the mainstream school (21). Even implantation between 2 and 4 years of age cannot rule out significant and often irreversible delays in several aspects of the child’s development. Thanks to the universal neonatal hearing screening programs that are being implemented worldwide, deaf-born children can now be detected at birth. Once identified, these infants have the right to receive the best medical treatment possible. Very early cochlear implantation is an essential element of this best medical treatment.

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REFERENCES


