# Cochlear Implantation

and

# Quality of Life Assessment

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# Cochlear Implantation

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# Quality of Life Assessment

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Whosoever wishes to know about the world must learn about it in its particular details.
Knowledge is not intelligence.
In searching for the truth be ready for the unexpected.
Change alone is unchanging.
The same road goes both up and down.
The beginning of a circle is also its end.
Not I, but the world says it: all is one.
And yet everything comes in season.
*Heraklietos of Ephesos*

# Contents

# General Introduction

# General introduction

Patients with moderate to severe hearing loss may benefit from a conventional hearing aid fitting. Nevertheless, for people with bilateral profound sensorineural hearing impairment, conventional hearing aids are of little or even no use. Sensorineural hearing loss occurs when there is damage to the inner ear (cochlea) or to the nerve pathways from the inner ear (retro cochlear) to the brain. The sensorineural hearing loss may be genetic or secondary to diseases, birth injury and drugs that are toxic to the auditory system. Sensorineural hearing loss may also occur as a result of noise exposure, viruses, head trauma, ageing and tumours[1]. Until several decades ago, it was not possible to provide patients with profound hearing loss with useful auditory input. Nowadays, the non-functional haircells in the cochlea can be bypassed by direct stimulation of the auditory nerve by means of a cochlear implant (CI).

Ideally, for deaf children, a CI creates the opportunity to develop spoken language and to improve mastery of language, in spoken and even in written form, and hereby it decreases the effects of deafness in a hearing society. Thus, a CI may alleviate to some extent the limitations brought along by being deaf in a 'hearing world'. However, it is important to recognize that deafness involves more than 'being unable to hear'. Furthermore, it is important to recognize that communication involves more than 'speaking to somebody'. It also presupposes a degree of interaction and understanding. The benefit of cochlear implantation differs from one young patient to another, depending on the child's age at onset of deafness (post-, perilingual or congenital onset), age at implantation, the child's character (motivation), cognitive abilities, the occurrence of additional handicaps and the duration and quality of support the patient receives[2]. Ultimately, the merit of a CI is determined by the ability of the recipient to maintain him- or herself within the hearing society, while retaining a positive identity[3]. Of course, the merit of a CI also depends on its technological factors. Most studies on cochlear implantation in the past decennia focused on speech perception and language development[4], but the impact on the patients' Quality of Life (QoL) has become an often used and acknowledged outcome measure as well[5]. This PhD-thesis focuses on the impact of a CI on diverse aspects of quality of life in different populations using various instruments to assess that quality.

After introducing the ear and the Deaf world, a description of the CI, its history, general functioning and selection criteria will be given in the following part of this introduction. Next, information regarding the Nijmegen/St Michielsgestel Cochlear Implant Team and the performance of CI patients in general will be presented. In the last part of the introduction, the term "Quality of Life" will be introduced, together with an introduction to the diverse instruments that are used in this thesis to assess quality of life. Finally, the aims of this thesis will be stated.

*I. Anatomy and Physiology of the ear*

Essential to the treatment of deafness with a CI, is understanding the inner ear's anatomy.

Centuries ago, the first investigations regarding the ear's anatomy were published. Alessandro Achillini (1463-1512) discovered the malleus and incus[6], though others claimed that it was Vesalius who first described these two ossicles. Approximately half a century later, Philippus Ingrassia (1510-1580) described the stapes and acknowledged the sound-conductivity of teeth[7]. Another anatomist, Julius Casserius (1561-1616), provided a wealth of comparative anatomy of the ear and larynx. He published *De Vocis Auditusque Organis Historia Anatomicae* in 1600. The quantity and quality of illustrations in Casserius' book greatly facilitated the understanding of the inner ear[8].

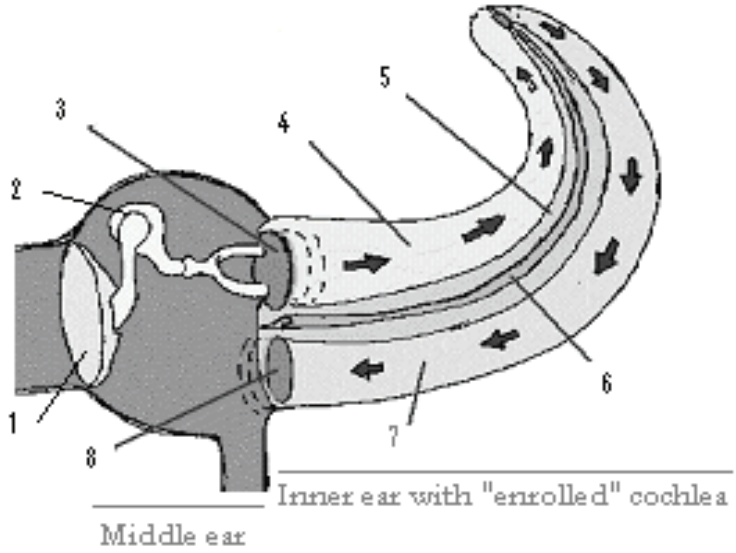
Portrait of Julius Casserius (1561-1616)

As described by the above mentioned researchers for the first time and elaborated upon by others, the human ear consists of three parts: the external, the middle and the inner ear. The external ear plays the role of an acoustic antenna: the auricle diffracts and focuses sound waves, the concha and the ear canal act as a resonator.

Acoustically, the eardrum is the final part of the external ear, which thus functions as a tube open only at one end. The eardrum or tympanic membrane separates the external auditory canal from the middle ear, which communicates with the nasopharynx via the Eustachian tube. The oval window (hidden by the stapes footplate) and the round window separate the middle and inner ear. The ossicular chain, malleus, incus and stapes, links the eardrum to the oval window [Figure 1]. The middle ear can be considered as an impedance adapter - without it most sound energy would be reflected by the fluid in the cochlea at the oval window. Two sensory organs are located in the inner ear: the organ of equilibrium and the organ of hearing, named the cochlea. The cochlea comprises a tube in the form of a snail's shell rolled up about 2 ½ times (35 mm in length)[9].

The cochlear implant works by using the tonotopic organization of the tectorial membrane of the inner ear. 'Tonotopic organization' stands for the way the cochlea sorts out different frequencies so that our brain can process that information. High-frequency sounds (i.e. high pitched sounds) do not pass very far along the tectorial membrane as the membrane close to the windows is specifically sensitive for high

frequency waves, whereas low frequency sounds do. In fact the tectorial membrane forms a short cut for high frequency waves travelling from the oval to the round window. The deflection of the stereocillia of the inner haircells, located all along the tectorial membrane, creates an electrical disturbance that is transported by the dendrites of the surrounding cochlear nerve cells. These nerve cells are uniquely connected to the haircells so that the frequency analysis performed by the tectorial membrane is maintained and available for subcortical neural processing. In cochlear implantation, the placement of the various electrode contacts along the cochlea enables the use of this tonotopic arrangement for speech coding and electric stimulation of selected regions of the cochlear nerve.




1. Tympanic membrane
2. Ossicular chain
3. Oval window
4. Scala vestibuli
5. Scala media
6. Tectorial membrane
7. Scala tympani
8. Round window

Figure 1.

*II. The Deaf and the dilemma of the Deaf Society*

*"An African American is not a non-White person, a woman is not a non-man and a Deaf person is not a non-hearing person"* [10]

The first written self-reflection of Deaf people in the Netherlands was found in letters of former pupils of the Dutch Guyot Institute for deaf education. Those 73 letters were written between 1809 and 1828. They showed a negative picture that the teachers had of the Deaf culture before entering the educational programme in the institute. The dissertation of Guyot's son (and director of the Guyot's institute from 1854 to 1861) contained a rather shocking quote: "The deaf and dumb are not entirely accountable because of mental deficits. They look like human beings and imitate human beings rather well. However, they essentially stand somewhere in the middle between animals and human beings, though closer to animals. They are not really part of society and even can be dangerous for society."[11] After having finished the

educational programme, the pupils were judged more positively. The deaf people themselves also reported this. In their letters they wrote about busy social lives, no experience of discrimination and the feeling of being useful to society. Nevertheless, the communication with the hearing was not always easy[12]. This first report already showed the different visions and feelings of the Deaf Society. In a more philosophical ethical way, the aspect of human dignity should be taken into account when speaking of the Deaf. Human dignity is more than a principle or a rule of behavior; it is the proper foundation of healthcare ethics[13]. Human dignity and human vulnerability are closely related concepts. The public debate on vulnerability is often related to the (re)habilitation process of disabled people. As far as Deaf people are concerned there is a strong public perception that they are vulnerable and that social solidarity is in accordance with the long-standing tradition of social and distributive justice. The Deaf Society does not agree with the perception of vulnerability of its culture. It is generally felt that although a 'normal' human being is a non-existent reality, there always are specific circumstances that might affect a social understanding of quality of life.
A study mentioned that the prevalence of subjective ill health, mental ill health and low appreciation of social contacts was higher in subjects with hearing disability. No association was found between hearing disability and the frequency of social contacts or with the functional content of the social contacts[14]. So, the Deaf feel that deafness is not a handicap and that in a broad perspective Deaf people living in a Deaf community, communicating only by sign language, are not disabled but just different. From a physiological perspective, however, deafness is always the result of some kind of pathology of the auditory pathway with characteristic histological and cytological changes. Hence, from the medical point of view adult and childhood deafness is always an impairment and should be treated accordingly. Notwithstanding the existence of a Deaf Society, accepted as such in a secular pluralistic society, deafness is associated with the lowest educational level and the lowest family income[15]. Cochlear implantation tries to overcome deafness, as an impairment, a disability and a limiting condition in the hearing world. When addressing the dilemma of cochlear implantation and the Deaf world, the otologic surgeon must be willing to think philosophically, to step out of the purely medical mode. For their part, the Deaf World needs to commit seriously to maintain dialogue with the otologic medical society. It means acquiring an accurate vision of hearing advances and disseminating that information to its members[16].

*IIIa. History of Cochlear Implantation*

Curiosity in stimulating hearing by way of electrical signals has existed since the discovery of the electrolytic cell by Volta in the first half of the nineteenth century. While some of Volta's contemporaries refined his rather crude and dangerous experiment, he himself placed a 50 Volt electrode in his ear. Nevertheless, these early

endeavours did not lead to any tangible results. A genuine breakthrough did not occur until the mid-twentieth century when Frenchmen Dijourno and Eyries drew scientific attention to the possibilities of electric stimulation of the inner ear, which they performed during otologic procedure for cholesteatoma eradication[17]. Dijourno and Eyries reported that it was indeed possible to recognize sounds through direct electric stimulation of the inner ear. Some years after the publication of their case-report several established otologic research groups were drawn to the field: the pioneering effect of scientists such as House, Simmons and Clark would prove to be momentous in the development of CI technology[18].

William House remembers the first steps he took towards developing a CI: "During the early sixties I implanted several devices in totally deaf volunteer patients. Unfortunately these were rejected due to the lack of biocompatibility of the insulating material. However, during the short time that these devices worked, it was obvious to me that this was an opening salvo to the conquest of sensorineural deafness." [19]

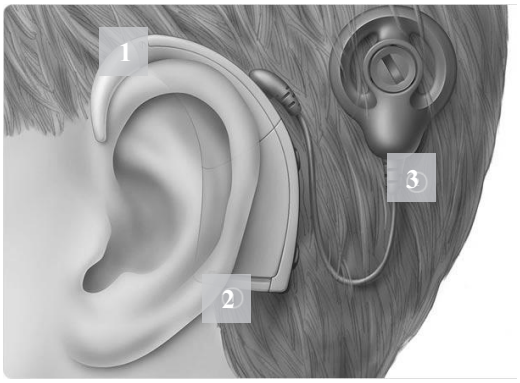
The first CI for clinical purposes was developed in the late sixties by the House Ear Institute in Los Angeles, CA, almost simultaneously with Clark and Simmons in Australia. In 1972 a wearable CI became available. In 1984 the FDA approved of the CI for application in post-lingual deafness. By 1990, several other countries had approved the CI for late deafened adults and- under some conditions- prelingually deafened children. Today more than 100.000 deaf individuals have received a CI (according to the FDA's website[20]), nearly half of which are prelingually deaf childen. Since the beginning the implant has evolved from single channel/single electrode systems to multiple channel/multiple electrode systems with sophisticated stimulation paradigms.

*IIIb. Functioning of the Cochlear Implant*

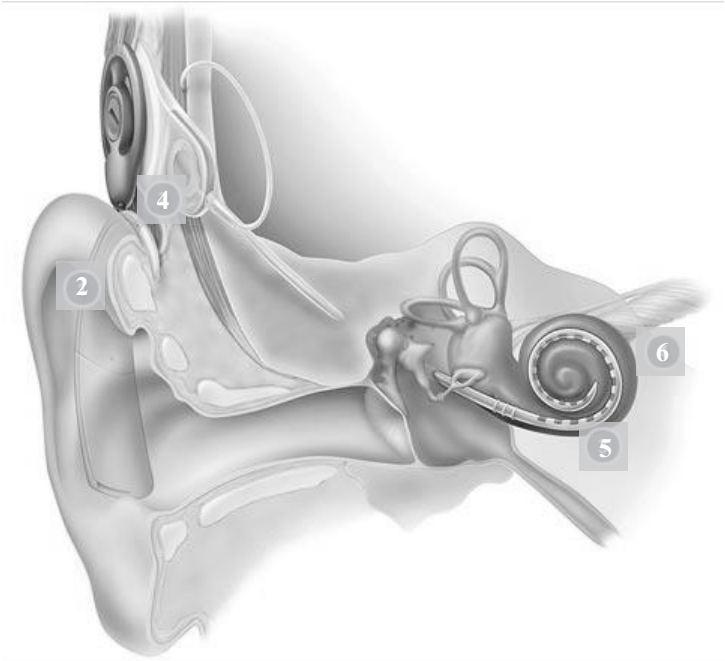
A CI is a semi-implantable technical device that provides hearing sensations to patients with sensorineural hearing loss. Sounds from the environment are registered by a microphone, converted by a so called 'speech processor' using a specific speech coding strategy. Subsequently, the encoded stimuli are transferred directly to the auditory nerve through electrodes, placed in the scala tympani [Figure 1]. In general, the CI can be divided into two parts, the part worn outside the body, and the part which is implanted. The external part contains the microphone, the processor and the transmitter, whereas the internal part of the device contains the receiver and the electrode array (wound through the cochlea in the scala tympani).

The microphone is usually fitted behind the ear [Figure 2 (1)]. The speech processor is a digital device, worn on the body or behind the ear like a conventional hearing aid [Figure 2 (2)]. The processed speech is transmitted by a coil through the skin by means of electromagnetic induction [Figure 2 (3)]. This signal is picked up by an antenna placed under the skin [Figure 2 (4)] and passed through then via an internal cable fed

to electrodes inside the cochlea [Figure 2 (5)]. The speech processor mimics the cochlea by spectral analysis of the microphone sounds. The resulting action potentials of the cochlear nerve dendrites are sent through the auditory pathways to the brain [Figure 2 (6)].




External part




Internal part

www.cochlear.com

Figure 2.

*IIIc. Selection and indication criteria*

The main indication for cochlear implantation is severe-to-profound sensorineural hearing loss that cannot be treated adequately with conventional hearing aids. Whether a CI works for a patient depends on patient factors, as well as on the (surplus) value of alternative ways to achieve appropriate profits. With respect to the former, the following aspects should be distinguished: anatomical and physiological prerequisites for proper processing of auditory signals, social, emotional and cognitive abilities and the social environment of the patient at home and/or at school. Alternative ways to achieve appropriate profits are, for example, using conventional hearing aids and communicating in Sign language.

Generally, the candidacy for implantation is considered separately for adults and children. As outlined in the 1995 National Institutes of Health (NIH) consensus statement on cochlear implantation, adult candidacy is noted as being probably successful in postlingually deaf adults with severe-to-profound hearing loss with no speech perception benefit from hearing aids[21]. In addition, the statement notes "most marginally successful hearing aid users implanted with a cochlear implant will have improved speech perception performance." Prelingually deafened adults, though

potentially suitable for cochlear implantation, must be counselled regarding realistic expectations. A strong desire for oral communication is, amongst others, paramount for this group of patients.

Currently, children with congenital and acquired severe to profound hearing loss are considered candidates for implantation from an age of six months on. In the case of a progressive obliteration of the cochlea after meningitis, even below the age of six months, an implantation may be considered. As for the adults, audiologic criteria include profound sensorineural hearing loss bilaterally and poor speech perception under best-aided conditions. As age at implantation is a crucial variable in the outcome of cochlear implantation, neonatal hearing loss needs to be detected at an early age by means of objective testing[22]. National screening programmes for hearing loss have contributed enormously. Objective tests in this age group include Auditory Brainstem Responses. This should be supplemented by Auditory Steady State Responses[23]. A hearing aid trial and various auditory training programmes are considered as essential before implantation is considered. Nowadays, also children with an additional handicap are considered for implantation (Chapter 1.3) as it is to be expected that those children, if sensitive for sensory input, can benefit from a CI as well[24].

*IIId. Nijmegen - Sint Michielsgestel Cochlear Implant Team*

The Radboud University Nijmegen Medical Centre and Viataal, formerly called the Institute for the Deaf, at Sint Michielsgestel have cooperated since 1986 in the counselling and rehabilitation of adult CI patients and since 1989 also for children with a CI. In 2000, this cooperation led to the formation of the official CI Centre Nijmegen/Sint Michielsgestel. During the past decades, this Implant Centre gained considerable expertise. In total, until 2006, 650 people received a CI in this Centre, 337 adults and 313 children. Annually about 100 CI patients are added to this number. In the Netherlands, about 100 to 150 deaf children are born yearly with a minimum hearing loss of 80 dBHL. Estimates of the annual number of candidates for CI vary widely from 30 to 150 or even 200 children. These estimates depend on the indication criteria used. Currently, in the Netherlands, the number of congenital deaf children, as well as the number of children becoming deaf after an episode of meningitis or rubella, is decreasing. The number of foreign deaf children and the number of children with an additional handicap are increasing.

*Adults*

In the past five years, between 55 to 80 deaf adults yearly applied for cochlear implantation. About half of them received a positive advice concerning the implantation. They have already received their implant, will receive it within the next months or are temporarily on a "parking-list" (progressive hearing loss, will receive a

CI when necessary). Others received a negative advice concerning implantation, mostly based upon residual hearing or prelingual deafness.
In the past 20 years 337 adults received a CI, 53 of which in 2006. Mean age at implantation varies as can be seen in the graph below [Figure 3]. Most people had been deaf for a relatively short period of time (0-10 years). The aetiology of deafness varies, with unknown deafness as most common cause. Other causes were for example meningitis, hereditary, trauma and otosclerosis[25, 26].

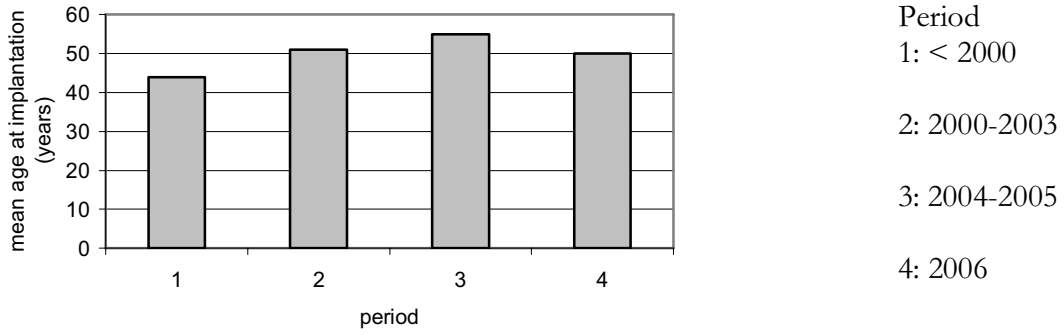



Figure 3.

*Children*
In the past five years, 251 children applied for a CI. In the past three years (2004 - 2006) most of those children received a positive advice concerning the implantation. In the past 17 years (1989 – 2006) 313 children received a CI, 58 in 2004 and 2005 and 31 in 2006. Mean age at implantation is decreasing, with in the past three years over one third being under two years of age. Most children are congenitally or prelingually deaf. The aetiology of deafness varies as mentioned in the adult section, with unknown congenital deafness as most common cause[25, 26].

*IIIe. Performance with a Cochlear Implant*
In several areas the importance and benefit of cochlear implantation seems undeniable. It was shown that deaf children with a CI perform almost equal to hearing impaired children with hearing loss of 70 dBHL or worse with conventional hearing aids on tests of speech perception, speech production and language skills, under the condition of early implantation[27]. Furthermore, the Reynell test demonstrated in other studies that the language skills improve faster for CI children when compared to children with severe hearing loss using conventional hearing aids. Yet it should be noted that there was a vast interindividual variability and the results of the CI children are generally poorer than the results of their hearing peers[28]. Geers mentioned that speech perception is the best documented effect of cochlear implantation. She

suggested that an increase in speech perception leads to better capability of spoken language development[4]. She showed that after receiving a CI, the development of grammatical structures increases rapidly, nevertheless, still stays behind in comparison with normal hearing children. Most progression in grammatical skills can be received when implanting a child at a young age[29]. The narrative abilities of CI users, including structure and use of language, are significantly related to speech perception, comprehension of syntaxes of spoken language and reading capacity. The narrative skills can even be a predictor for reading skills in a deaf child. Cochlear implantation can enhance the narrative skills to a level comparable with hearing peers, though the previously mentioned interindividual variability is also seen here[30].

It may be more demanding to investigate the cognitive skills of CI patients. Deaf patients obviously have normal learning capacities, but cognitive development may be different for deaf people. Initially, perception is mainly visual and the experiences over time are influenced by communication and language development[31]. An example of a specific cognitive task is reading. Deprivation of auditory stimuli and spoken language seems to negatively influence the ability to encode, rehearse and recall sequential information, the importance of early cochlear implantation in deaf children when concerning the cognitive abilities[32].

As the first CIs in children were placed about 20 years ago, results can now be presented of long-term follow up. However, it should be noted that the implants are improving and the age of implantation is decreasing (shorter period of deafness), which makes comparison with new CI users difficult: this is often referred to as the "moving target phenomenon" [33].

In an extensive research project, Stacey et al. described the long-term follow-up results of CI children by means of questionnaires on auditory skills, spoken language, school performance and quality of life. There was no clear correlation between school performance and quality of life. Children implanted before the age of 5 showed overall the best performance[34]. The long-term follow-up results in adults of the Nijmegen-Viataal Cochlear Implant centre are described in chapter 1.4.

All of the results mentioned above concern deaf people without developmental delay and a normal IQ. As implant criteria are expanding, children with additional handicaps are included in the implant procedure as well. In a retrospective study, the speech and language development of CI children with and without minor developmental delay was compared. All children showed significant improvement after implantation in both speech and language development. Only in the domain of auditory speech perception, the children without developmental delay outperformed the study group significantly. The differences between the two groups were very apparent in spoken language skills. Nevertheless, it was concluded by the researchers that the children with developmental delay do benefit from a CI[35]. Those findings were more or less confirmed by our current study (chapter 1.3).

An extensive overview of diverse aspects of the development of CI children and its possible consequences for the pedagogic management was recently written by the Nijmegen-Viataal Cochlear Implant centre [36].

*IVa. Quality of Life*

In general, the term quality of life is used by societies to indicate how happy people are compared to the people of another society. A discussion arises with the definition of 'happiness' because what 'happiness' means to one person is not necessarily what it means to another. Quality of life involves more than just the material things that one possesses, e.g. the feeling of wellbeing. It includes many different aspects. Although all the different aspects of quality of life are important, some societies place more emphasis on certain aspects than on others. Operational definitions of quality of life are diverse, with variability fuelled not only by use of societal or individualistic perspectives but also by the range of applicable theoretical models or academic orientations. Several authors have commented on this diversity. In 1976, Liu stated that there were as many quality of life definitions as people, emphasizing the axiom that individuals differ in what they find important[37]. Some years later, Baker and Intagliata (1982) point to there being as many definitions as the number of researchers studying the phenomenon, a comment that throws the spotlight on the lack of agreement between those attempting to operationalise the concept. In their view, writers had done little up to that point to achieve definitional consistency[38].

In a review study on quality of life in 1995, Felce et al. defined quality of life as an overall general well-being that comprises firstly objective descriptors, secondly subjective evaluations of physical, material, social, and emotional wellbeing and finally the extent of personal development and purposeful activity, all weighted by a personal set of values[39] [Figure 4]. The three elements were shown in dynamic interaction with each other. Changes in some objective facet of life may change satisfaction or one's personal values or both. Similarly, changes in values may change satisfaction and precipitate change in some objective circumstance. In the same way, they stated, a change in a sense of satisfaction may lead to reappraisal of values and lifestyle. As well as affecting each other, the three elements are capable of changing independently as a result of external influences. Such external influences might include genetic, social, and material inheritance, age and maturation, developmental history, employment, peer influences and reference points, and other social, economic, and political variables. As the three elements that define quality of life are all open to external influence, assessment of all three is necessary to any measurement system purporting to capture quality of life. Knowledge of one set cannot predict another because the relationships between them may not remain constant. Quality of life issues in general and specific in the CI area are more extensively discussed in the General Discussion chapter.

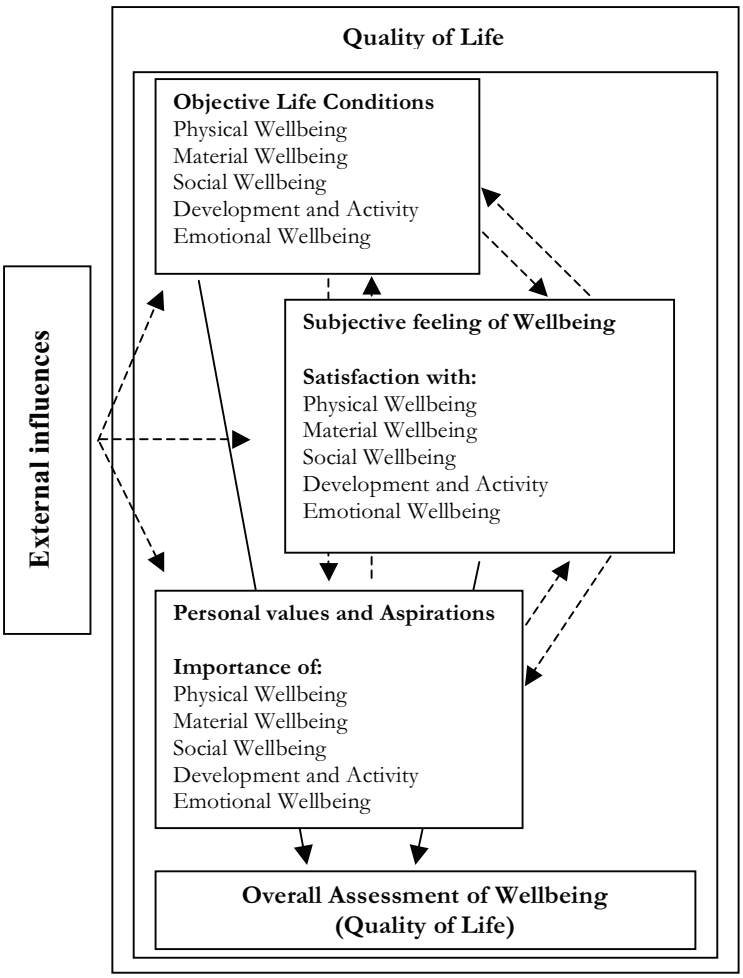



Figure 4. A model of quality of life.
*(Felce D., Perry J., Quality of Life: Its definition and measurement. Research in Developmental Disabilities1995)* [39]

*IVb. Quality of Life and Cochlear Implantation*

The social and emotional development of CI patients is an important focus of care[40]. This simultaneously applies to the short and the long run: the well-being of the patient and the ability of this patient, especially when being a child, to maintain itself in society when it grows older. Social and emotional well-being is related to the development of a positive identity, that is, a contended awareness of who one is and whom one belongs to. Such development requires that a CI user learns to be assertive, interacts with his or her social environment and is able to communicate[41]. Communication, especially language development, is a crucial prerequisite for developing a positive identity within the hearing world, because interaction with others has a linguistic nature. First studies on cochlear implantation and quality of life were performed in the late 80s, by means of interviews. Later on, self-reported measures of the benefits of a CI filled in by adults correlated well with the objective test results, but also revealed important information that was not available from objective tests[42]. In that period,

experiences and feelings of several adult CI patients of our implant centre were described. Very strikingly, a CI patient says: "What you experience is, by matter of speech, the shadow of the sound"[43]. Quality of life in CI children was described for the first time, in France, in 1992, where cochlear implantation showed a significant impact on the quality of life of all implanted children[44].

The goal of CI, naturally, is to improve or restore the hearing of deaf individuals. Hearing, however, is not a goal in itself. Many regard hearing as an aid in speaking and reading and consequently in communication, as well as in intellectual, social and emotional development. With respect to pediatric cochlear implantation, Boothroyd et al. explain:

"The long term goal of implantation is the reduction of developmental delays by virtue of the partial restoration of auditory capacity. The four areas to be covered are: language (knowledge of vocabulary, grammar, use of language, independent of modality, such as speech or words), speech (the ability to produce sounds, words and sentences recognizable, via hearing, to others), intelligence (particularly verbal IQ, reading ability and academic attainment) and social and emotional status (perception of and behaviour towards others and self)"[45]. This emphasises the importance of quality of life (social and emotional status) in evaluating cochlear implantation.

*IVc. Quality of Life Instruments*

In this thesis, ten different QoL instruments were used. A short overview of the instruments is provided in the table on the right [Table I]. It should be noted that two instruments were especially designed for cochlear implantation. These two are the NCIQ and the PP. More detailed information about these instruments can be found in different chapters of this thesis.

*V. Aims of this thesis*

Primarily, a CI aims to improve or restore the hearing of deaf subjects. Hearing is a vital prerequisite for normal development of speech and reading and consequently of importance in communication and appropriate intellectual, social and emotional development in a hearing world. The impact of a CI on aspects of daily life was investigated in specific CI populations (1.Assessment of specific populations). Different instruments used for the assessment of QoL were investigated (2.Assessment of specific instruments). Thus, an insight into the diversity in quality of life between differing populations receiving a CI might be obtained. For proper analysis, the validity and accuracy of diverse instruments was addressed, in order to obtain knowledge of which questionnaires should be used. A more detailed introduction to the topics of the chapters will be discussed next.

Table 1.

| Abbreviation | Full Instrument name | Type | Suitable for Adult (A)/Child(C) | Number of items | Number of domains | Domains | Score (range) | Used in this thesis (chapter) |
|---|---|---|---|---|---|---|---|---|
| AMP[46] | Assessment of Mainstream Performance | Specific | C | 22 | 2 | - Overall score Classroom performance<br>- Class ranking | Categorical | 1.1.<br>2.1. |
| GBI[47] | Glasgow Benefit Inventory | Specific | A | 18 | 3 | - Physical<br>- Emotional<br>- Overall benefit score | -100 – 100 | 1.2 |
| GCBI[48] | Glasgow Children's Benefit Inventory | Specific | C | 24 | 1 | - Overall benefit score | -100 – 100 | 1.2<br>1.3 |
| HUI 3[49] | Health Utility Index mark 3 | Generic Utility score | A, C | 15 | 8 | - Vision<br>- Hearing<br>- Speech<br>- Ambulation<br>- Dexterity<br>- Emotions<br>- Cognition<br>- Pain | 0 – 1 | 1.2<br>1.4 |
| NCIQ[50] | Nijmegen Cochlear Implant Questionnaire | Specific | A,C | 60 | 6 | - Sound perception basic<br>- Sound perception advanced<br>- Speech production<br>- Activities<br>- Social functioning<br>- Self esteem | 0 – 100 | 1.2<br>1.4 |
| PedsQL[51] | Pediatrics Quality of Life | Generic | C | 23 | 4 | - Physical functioning<br>- Emotional functioning<br>- Social functioning<br>- Functioning at school | 0 - 100 | 1.3 |
| PP[52] | Parent's Perspectives | Specific | C | 74 | 10 | - Communication<br>- General functioning<br>- Self-reliance<br>- Wellbeing, happiness<br>- Social relationships<br>- Process of implantation<br>- Education<br>- Effects of implantation<br>- Decision to implant<br>- Supporting the child | -100 – 100 | 2.3 |
| SF 36[53] | medical outcome study Short Form 36 | Generic | A,C | 36 | 8 | - Physical functioning<br>- Role functioning due to physical health problems<br>- Role functioning due to emotional problems<br>- Bodily pain<br>- Vitality<br>- Social functioning<br>- Mental health<br>- General health | 0 – 100 | 1.2<br>1.4 |
| SIFTER [54] | Screening Identification For Targeting Educational Risk | Specific | C | 15 | 5 | - Academics<br>- Attention<br>- Communication<br>- Class participation<br>- School behavior | Categorical | 1.1<br>2.1 |
| Usher Lifestyle[55] | Usher Lifestyle Survey | Specific | A,C | - | - | - | Descriptive | 2.2 |

The development of spoken language in deaf children with a CI has made mainstreaming a more reachable academic level[56]. To be able to get an insight into

how CI children perform in mainstream education in comparison with their normal hearing peers, several school-related skills were examined by means of the AMP and SIFTER questionnaires. The main research question addressed *(1.1)* is whether differences can be seen in the classroom performance and communicative skills between CI children and their normal hearing peers. As language development had not been investigated so far in relation to the AMP and SIFTER, it was decided that further research *(2.1)* was necessary to determine whether the mentioned variability in AMP and SIFTER results could be related to differences in language development. Before performing these analyses, the instruments' structures were examined.

Usher syndrome is a genetic disorder that causes sensorineural hearing loss, retinitis pigmentosa (RP), and sometimes balance problems. Usher type I is characterised by congenital profound deafness, absence of vestibular function and progressive vision loss due to RP. Owing to the double handicap, cochlear implantation might play a major role in the rehabilitation of these individuals and may be among those most worthwhile to consider for implantation. In a study *(1.2.a)* a survey of the benefits of cochlear implantation in Usher type I patients with regard to hearing, vision and quality of life is presented. In another study *(1.2.b)*, the audiological performance after cochlear implantation in Usher type I patients is analysed in more detail. Finally, a survey *(2.2)* especially designed for a project concerning Usher patients, was described.

Since 1989, deaf children have received cochlear implants at the Nijmegen/St. Michielsgestel CI team. Initially, only children who were expected to make optimal use of the CI were implanted. Only recently, children with developmental delay have received a CI. The advantages of cochlear implantation in children with an additional cognitive handicap besides deafness (either developmental delay: non-verbal IQ<80, or severe learning disabilities) have been mapped *(1.3)*. Results of language comprehension tests and quality of life questionnaires were compared to those obtained from a reference group of CI children with deafness alone (normal cognition).

Cochlear implantation has proved to be a successful and effective treatment for severely and profoundly postlingually deaf individuals. At present, little is known about the long-term quality of life of postlingually deaf CI users. A study *(1.4)* aimed to gain more insight into changes in quality of life in adult CI recipients during long-term follow-up. The following aspects were addressed: long-term effects of a CI on quality of life, changes in quality of life during follow-up and long-term changes in quality of life. In addition, the results of the quality of life instruments were evaluated in relation with speech perception scores.

It is generally believed that the parental view has the potential to add significant value to structured evaluations of CI children gained by professionals[57]. Archbold et al. developed a parent outcome instrument, commonly referred to as Parental Perspectives (PP) for cochlear implantation[52]. The importance of this instrument seems generally accepted. In a current study *(2.3)*, the main question relates to evaluation and validation of the PP by means of statistical analysis. Additionally, as the present PP is often considered as rather lengthy for workable practice, the possibility of developing a short version was explored.

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# 1 Assessment of specific populations

1.1

# Deaf Children with a Cochlear Implant in Mainstream Education

Classroom performance of children with cochlear implants in mainstream education.
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# Abstract

*Aim:* To compare classroom performance of children with a CI with their normal hearing peers in mainstream education.

*Methods:* Thirty-two CI children in mainstream education, congenitally or prelingually deaf, participated in this study and 37 hearing classmates. The teachers filled in two questionnaires: the Assessment of Mainstream Performance (AMP) and the Screening Instrument for Targeting Educational Risk (SIFTER). High Fletcher Index and open speech perception scores were obtained.

*Results:* CI Children scored above average in the AMP and sufficient in all but one area (communication) of the SIFTER questionnaire. Class ranking did not differ significantly between the CI students and their normal hearing peers. Overall, the normal hearing group outperformed the CI group. Classroom performance of CI children correlated negatively with duration of deafness and age at implantation. All longitudinal audiologic data of the CI children showed improvement in open speech recognition.

*Conclusion:* Although results are encouraging, the CI group scored significantly less than their normal hearing peers in most questionnaire domains of both the AMP and the SIFTER. Most important variables for the outcome in this study were age at implantation and the duration of deafness.

# Introduction

The availability of cochlear implant technology has made mainstreaming a more reachable academic level for deaf children. Latest technological advances in cochlear implants and gathered experience on paediatric cochlear implantation in the last 15 years have evolved candidacy criteria[1]. The age at implantation is decreasing. Because more children are implanted at an earlier age and obtain good results with their CI, it is likely they will be ready to enter mainstream education at the same age as their hearing peers. Though this may be a successful participation for some, others will need additional skills and/or facilities to be able to succeed in mainstream education. A provisional report on the needs of CI students in mainstream education showed that there are some skills with regard to communication (for example comprehension of classroom instruction) in which children with implants constantly show poorer performance than their hearing classmates[2]. Paediatric CI users have attained diverse levels of achievement in school[3].

Many articles described several factors influencing the outcome in speech, language and reading competence of the implanted child and hereby may be influencing (mainstream) school performance[4-7]. Factors discussed are age at onset of deafness, age at implantation, amount of residual hearing before implantation, period of implant use and technical and educational setting. It has been reported that cochlear implantation attended by aural rehabilitation tends to lead to higher rates of mainstream placement, with consequential cost savings. Data indicate that CIs offer valuable input for increasing literacy, next to increasing speech perception, production and language[8].

Since 1989, 235 children have received a cochlear implant at the department of ORL of the Radboud UMC. Thirty-eight CI children are at this moment enrolled in mainstream primary or secondary school. To be able to get an insight in how they perform in mainstream education in comparison with their normal hearing peers, we examined several skills in the classroom. All mainstream teachers of CI students were asked to complete two questionnaires for the implanted child as well as for a normal hearing classmate, selected by protocol. Besides the Assessment of Mainstream Performance (AMP), teachers also completed the Screening Instrument for Targeting Educational Risk (SIFTER). The Children's Hearing Institute, New York, USA, developed the Assessment of Mainstream Performance (AMP) to determine the skills that children require to be successful in mainstream school settings[2]. The SIFTER is a short teacher-rating test that explores several areas of school performance[9].

The main question addressed in this study is whether differences can be seen in the classroom performance and communicative skills of CI students and their normal hearing peers. Results of the questionnaires were analysed for correlation with audiologic test results. It is expected that the results may be of assistance in dealing

with the probable challenges CI students meet while mainstreaming as well as an effort to identify benchmarks of performance that children will require to be successful.

# Materials and methods

**Participants**

To date, a total of 235 children have been implanted at the Radboud University Medical Centre Nijmegen. Thirty-eight of these children were placed in mainstream school settings, a decision which was made individually per child in close collaboration with the rehabilitation specialists (among who psychologists and school representatives) of the Nijmegen/ St Michielsgestel Cochlear Implant team. Six different ambulatory coaches from all over the country counselled the CI children. Cooperation of the coaches was required to obtain information regarding the specific schools and teachers involved in these children. Thirty-six of the CI students received weekly extra support, like remedial teaching and speech therapy.

The control group consisted of normal hearing classmates of the CI children, chosen by randomised selection, described in the AMP protocol: "in order to ensure that there is no bias in the selection of the non-implanted child, a method of selection has been developed for this purpose. Teachers will list all children's names in alphabetical order, omitting the child with the implant from the list. When using the kindergarten/preschool AMP, the fourth child will be selected as the control. When using the elementary/high school AMP, the eighth child will be selected, If this cannot be accomplished due to small class size, then the teacher should complete the AMP for the last child on the list". The parents of the non-CI children were given a short questionnaire by the teacher which comprised 6 questions about the hearing of their child, in order to be able to objectify their normal hearing, which was confirmed in all cases. Informed consent for this study was obtained from the parents. Parents and teachers were offered help by email or by telephone when necessary.

Twenty of the children were congenitally deaf, 12 became deaf prelingually (≤ 3 years of age) and 5 postlingually (>3 years). It was decided to leave the postlingually deaf children out of further analyses as their results are generally expected to differ from congenitally or prelingually deaf children, making the final total number CI students 32. All received the implant between 1995 and 2003 (mean age at implantation 3;7 years, range 1;0 to 9;7 years). Twenty-nine children received the Nucleus 24-channel and 3 children the Nucleus 22-channel cochlear implant (Cochlear Corporate, Englewood, Australia). In all patients surgery was uneventful and a full function of the electrode array was achieved. Mean duration of deafness is 3;4 years (0;4 year to 9;7 years). At the time of this study the mean period of cochlear implant use was 5;0 years

(from 1;0 year to 9;1 years) and the children's ages ranged from 4;5 to 13;0 years (mean 9;0 years). Psychological reports were checked. None of the patients showed a delay in their psychological development and all obtained intelligence quotient scores (IQ) of > 80. The scores were determined before implantation as part of the (psychological component of the) selection procedure and obtained by means of standardized intelligence tests. At time of the investigation no child was diagnosed with an additional handicap, such as learning difficulties or pervasive developmental disorders. Patient characteristics are shown in Table 1. There was a vast interindividual variability in age at onset of deafness (period of hearing), age at implantation (period of deafness) and age at entering mainstream education. Hence, all age related characteristics of the cochlear implanted patients are shown in Figure 1.

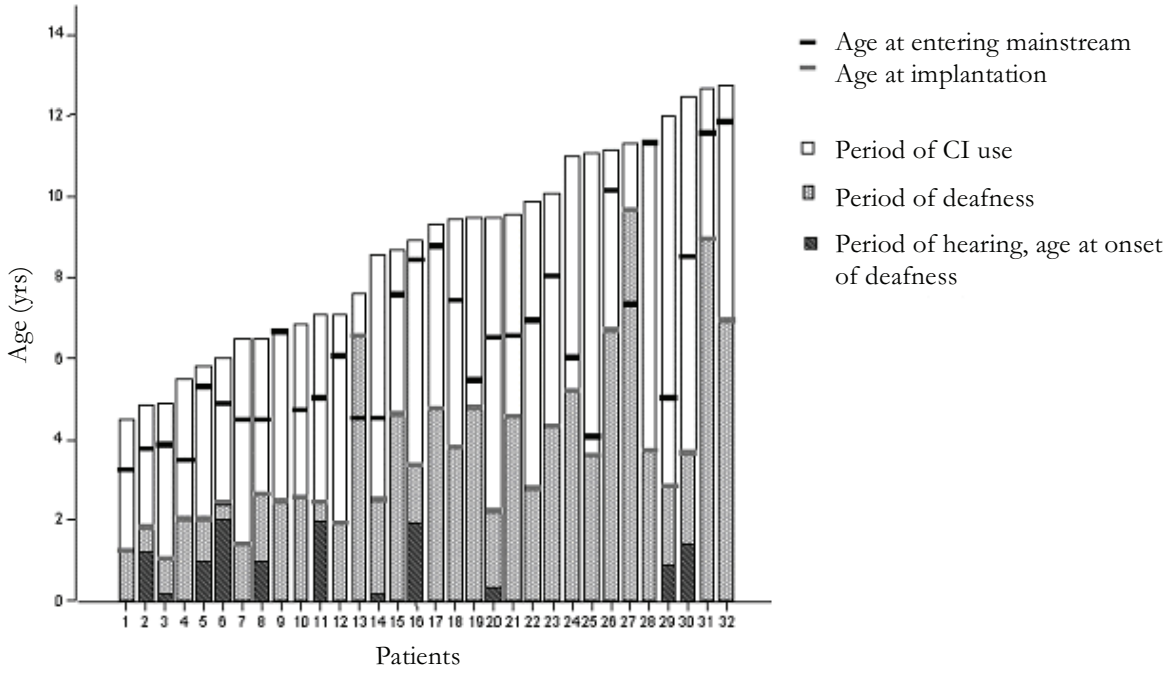


Figure 1. Patient numbers correspond with the numbers mentioned in Table 1. Total height of bars represents the age of the children in years. The different CI related periods and events are marked in the bar chart.

**Educational setting**

The teachers were contacted by telephone and given information about the research project. Two teachers in elementary school did not get permission from the parents of the normal hearing child to fill out the questionnaires, making the total number of participating children 67; 32 with CI (18 boys, 14 girls) and 35 without CI (14 boys, 21 girls). Twelve completed questionnaires were received from kindergarten and 55 from elementary schools.

Table 1. Patient characteristics

| Child | Current age | Onset deafness | Congenital Prelingual | Cause of Deafness | Duration of deafness | Age at implant | Period CI use | Residual hearing | Cochlear Type CI |
|---|---|---|---|---|---|---|---|---|---|
| 1 | 4,5 | 0,0 | con | unknown | 1,3 | 1,3 | 3,2 | - | N24 |
| 2 | 4,8 | 1,2 | pre | meningitis | 0,6 | 1,8 | 3,0 | - | N24 |
| 3 | 4,9 | 0,2 | pre | meningitis | 0,9 | 1,0 | 3,9 | - | N24 |
| 4 | 5,5 | 0,0 | con | dysplasia | 2,0 | 2,0 | 3,5 | - | N24 |
| 5 | 5,8 | 1,0 | pre | meningitis | 1,1 | 2,1 | 3,8 | - | N24 |
| 6 | 6,0 | 2,0 | pre | meningitis | 0,4 | 2,4 | 3,6 | - | N24 |
| 7 | 6,5 | 0,0 | con | Waardenburg | 1,4 | 1,4 | 5,1 | - | N24 |
| 8 | 6,5 | 1,0 | pre | meningitis | 1,6 | 2,6 | 3,9 | - | N24 |
| 9 | 6,8 | 0,0 | con | dysplasia | 2,5 | 2,5 | 4,3 | - | N24 |
| 10 | 6,8 | 1,0 | pre | meningitis | 2,6 | 2,6 | 4,2 | - | N24 |
| 11 | 7,1 | 0,0 | con | hereditary | 2,0 | 2,0 | 5,1 | - | N24 |
| 12 | 7,1 | 2,0 | pre | meningitis | 0,5 | 2,5 | 4,6 | - | N24 |
| 13 | 7,6 | 0,0 | con | unknown | 6,6 | 6,6 | 1,0 | - | N24 |
| 14 | 8,6 | 0,2 | pre | meningitis | 2,4 | 2,5 | 6,0 | - | N24 |
| 15 | 8,7 | 0,0 | con | hereditary | 4,7 | 4,7 | 4,0 | - | N24 |
| 16 | 8,9 | 1,9 | pre | meningitis | 1,4 | 3,3 | 5,6 | - | N24 |
| 17 | 9,3 | 0,0 | con | CMV | 4,7 | 4,7 | 4,6 | - | N24 |
| 18 | 9,4 | 0,0 | con | dysplasia | 3,8 | 3,8 | 5,6 | - | N24 |
| 19 | 9,5 | 0,0 | con | unknown | 4,8 | 4,8 | 4,7 | - | N24 |
| 20 | 9,5 | 0,4 | pre | meningitis | 1,9 | 2,2 | 7,3 | - | N24 |
| 21 | 9,6 | 0,0 | con | hereditary | 4,5 | 4,5 | 5,1 | - | N24 |
| 22 | 9,9 | 0,0 | con | unknown | 2,8 | 2,8 | 7,2 | - | N24 |
| 23 | 10,1 | 0,0 | con | unknown | 4,3 | 4,3 | 5,8 | - | N24 |
| 24 | 11,0 | 0,0 | con | Waardenburg | 5,2 | 5,2 | 5,8 | - | N24 |
| 25 | 11,0 | 0,0 | con | unknown | 3,6 | 3,6 | 7,5 | - | N20+2 |
| 26 | 11,1 | 0,0 | con | unknown | 6,7 | 6,7 | 4,5 | - | N24 |
| 27 | 11,3 | 0,0 | con | Pendred | 9,7 | 9,7 | 1,7 | + | N24 |
| 28 | 11,3 | 0,0 | con | Usher | 3,7 | 3,7 | 7,6 | - | N24 |
| 29 | 12,0 | 0,9 | pre | meningitis | 2,0 | 2,9 | 9,1 | - | N20+2 |
| 30 | 12,5 | 1,4 | pre | meningitis | 2,2 | 3,6 | 8,9 | - | N20+2 |
| 31 | 12,7 | 0,0 | con | Usher | 9,0 | 9,0 | 3,7 | - | N24 |
| 32 | 12,8 | 0,0 | con | unknown | 7,0 | 7,0 | 5,8 | - | N24 |
| Mean | 8,7 | 0,4 | | | 3,4 | 3,7 | 5,0 | | |
| Range | 4,5 - 12,8 | 0,0 - 2,0 | | | 0,4 - 9,7 | 1,0 – 9,7 | 1,0 – 9,1 | | |

All ages and periods are mentioned in years.
Con=congenital and pre=prelingual onset of deafness

All participating schools were registered at the Inspectorate of Education and had a certificate of quality, which gives an overview of the quality of a school. In all participating schools the educational test results were sufficient, indicating that results were as to be expected by standardised protocol measurements.

All children, but one, get extra support like remedial teaching and speech therapy. Some teachers use facilities like a microphone during instructions on subject matters. The amount of information about the extra facilities in classroom varied among divers schools and teachers, though most report to have sound-absorbing treatments in classroom (e.g., carpeting, window treatments, acoustic tiles).

**Questionnaires**

The Children's Hearing Institute, New York, USA, developed the Assessment of Mainstream Performance (AMP) to determine the skills that children require to be successful in mainstream school settings[2]. The AMP has two versions: one is utilised with preschool and kindergarten aged children (age 3-5) and comprises 16 questions. The other version, which consists of 22 questions, is for children educated in elementary or high school settings. The questions of both versions investigate the child's ability to participate in a range of typical classroom activities and behaviours that are age and content appropriate. In the AMP-K and AMP-E, 2 and 4 questions respectively are posed in opposite form, so these answers were recoded before computation. Answers to the questions are categorised in percentage of time that a child shows certain age and content appropriate behaviour (almost never; 0-10%, to almost always; 91-100%). Class ranking, a separate question within the AMP, indicates the child's level in regard to their peers, estimated by the teacher, ranging from failure (0-25%) to excellent (91-100%). Both versions were validly translated in Dutch (translated twice by different translators, compared and translated backwards) and sent to the teachers by mail. As an example, the AMP elementary school instrument is provided as Appendix 1.

The second questionnaire used is the Screening Identification For Targeting Educational Risk (SIFTER). We used this questionnaire complementary to the AMPs to rule out the possibility that children have an abnormal behaviour due to an unknown problem in education. The SIFTER is a test designed to provide a valid method by which children with hearing problems can be screened educationally. The SIFTER is a short, 15-item teacher-rating test that explores several areas of school performance including academics, attention, communication, class participation, and school behavior. The five content areas comprise three related questions answered through a ranking scale, from one to five. Scoring the SIFTER involves summing the responses of the three questions in each content area. Responses are then placed on a chart to develop a profile of the subject, composing three categorical outcome measures (failure, marginal or sufficient)[9;10]. There are two versions of the SIFTER; a

preschool version for young children (aged 3 – kindergarten) and one for older children. The SIFTER has been field-tested and has been shown to have good content and score reliability[11].

**Audiologic data**

We reviewed the medical and rehabilitation files of all CI patients for additional and audiologic data. High Fletcher Index (hFI), computed from the audiograms at 6 months and yearly after implantation, and open speech recognition scores were used as outcome measures. Two different Dutch Consonant-Vowel-Consonant (CVC) tests were used for open speech scores: the Bosman test[12] and the easier Gestel-Nijmegen (GN) test[13-15]. Preferably, tests were done in standardized conditions using a compact disc, if that was not possible, monitored live voice testing was done at 70 dB, at 6 months and yearly after implantation. The most recent hFI and open speech recognition score were used for regression and correlation analyses. The residual hearing before implantation was determined, using the principle of 'Count the Dots' audiogram[16]. As only 1 child had marginal functional residual hearing according to this principle, no further analyses have been performed on this subgroup.

**Data analysis**

Data were recoded and analyzed in SPSS 12.0.1. As numbers were small and data were not assumed to be normally distributed, a non-parametric equivalent to the T-test was used. Mann-Whitney exact tests determined potential statistical significance between the CI children and their normal hearing peers for the AMP means and the 5 areas of the SIFTER. Class ranking, a question within the AMP-list, was used as a separate variable.

General linear model analysis was used to investigate possible correlations between AMP-scores /SIFTER-scores/ class ranking and different variables. The audiologic variables were high Fletcher index and open speech scores and the analysed implant variables were duration of deafness, period of implant use and age at implantation. The number of years following mainstream education was used as separate variable.

# Results

**AMP**

In kindergarten both the CI children (n=6) as well as the children in the control group (n=6) were spending 75-90% of their time (mean AMP-K score 4.6 resp. 5.3) performing to their ability in class [Table 2], which means they often participate in classroom activities and show age appropriate behaviour. In elementary school the CI

children (n=26) had a mean AMP score of 4.1, which corresponds with regular participation and appropriate communicative behaviour in 51-75% of time. Their normal hearing peers (n=29) obtained a mean AMP-E score of 5.0, often (75-90% of time) performing to their ability. This resulted in a significant difference between the hearing children and the children with CI in the AMP-E scores (p=0.00). The mean AMP-E score of congenitally deaf children (3.9) was significantly lower than this mean score of the prelingually deaf children (4.7, p=0.01). The peak scoring differences between the CI students and the normal hearing children in elementary school were seen in questions on communication breakdown, engagement in group discussion and displaying turn taking abilities or a leadership role. Within the CI group, there were significantly better scores on these questions for the prelingually deaf children compared to the congenitally deaf children.

The question on class ranking investigated the teacher's estimation of the child's level with regard to its peers. All the CI children scored 'above average' and the total non-CI group scored 'good'. Mean scores of class ranking and its implications are shown in *Table 2*. No significant differences were seen between the CI students and their hearing classmates, though in elementary school the outcomes did definitely differ (p=0.08).

Furthermore, class ranking scores in elementary school showed positive correlation with the AMP-E score (Pearson: 0.58, p=0.00). A negative correlation was found between the AMP-E score and duration of deafness (Pearson: -0.66, p=0.00) as well as with the age at implantation (Pearson: -0.60, p=0.00), indicating the longer the duration of deafness or the older the child has received its implant, the lower the AMP-E scores. When groups were subdivided by duration of deafness (less than 2 years, 2 to 4 years and more than 4 years) significantly higher overall AMP results were encountered in the groups with a shorter period of deafness (Chi square test between the 3 groups < 0.01). Children with duration of deafness longer than 4 years obtained worse scores.

Audiologic test results demonstrated no coherence with the AMP scores. It should be noted that in the CI group of the kindergarten age, there are only two children who have performed the audiological tests; hence no analyses can be done with those two children.

Table 2. Results AMP kindergarten and AMP elementary school

| **AMP** | | CI+ | | | CI- | | | CI+ vs. CI- Significance |
|---|---|---|---|---|---|---|---|---|
| | | n | Mean (SD) | Meaning | n | Mean (SD) | Meaning | |
| AMP-K | Overall score | 6 | 4.58 (0.94) | Often (75-90% of total time) | 6 | 5.34 (0.25) | Often (75-90% of total time) | 0.22 |
| | Class ranking | | 3.33 (0.82) | Above average | | 3.58 (0.67) | Good | 0.70 |
| AMP-E | Overall score | 26 | 4.14 (0.68) | Regular (51-75% of total time) | 29 | 5.03 (0.59) | Often (75-90% of total time) | *0.00* |
| | Class ranking | | 3.07 (1.00) | Above average | | 3.55 (0.83) | Good | 0.08 |

Comparison of children with and without CI in both kindergarten and elementary school. Means (Standard Deviation) and significance (by use of exact Mann-Whitney U test, $p < 0.05$ is significantly different) were computed for the overall score and for class ranking . Meaning of the mentioned numbers is shown as well; the AMP overall scores represent the percentage of time that the child shows appropriate communicative behaviour and the class ranking is divided into 5 categories, 5; being the best score (outstanding achievement) to 1; being the worst score (failure). Class ranking, a separate question within the AMP, indicates the child's level in regard to their peers, estimated by the teacher, ranging from 5: excellent (top 91-100%) to 1: failure (lowest 0-25% of the class).

## SIFTER

Mean scores of the content areas were computed and recoded into 3 categorical outcomes; failure or marginal or sufficient. The results, divided into 5 areas, showed overall sufficient outcomes, though in elementary school the hearing students perform significantly better in four of the five content areas *[Table 3]*. The CI children in kindergarten scored marginal on communication (8.83), significantly worse than their hearing peers (12.17, p=0.01). In elementary school the CI students failed on communication with a score of 7.2, again significantly worse than their hearing classmates (11.4, p=0.00). These findings suggest a deprivation in the area of communication for the CI students, regardless their age.

In kindergarten a negative correlation was observed between duration of deafness and the area of communication (Pearson: -0,88, p=0.02). Period of implant use (in years) was positively correlated with two areas; attention (Pearson: 0.81, p=0.05) and social behaviour (Pearson: 0.84, p=0.04).

In elementary school duration of deafness correlated negatively with four areas; academics (Pearson: -0.53, p=0.01), attention (Pearson: -0.46, p=0.02), communication (Pearson: -0.52, p=0.00) and class participation (Pearson: -0.48, p=0.02). Age at implantation also correlated negatively with four content areas; academics (Pearson: -0.60, p=0.00), attention (Pearson: -0.40, p=0.05), class participation (Pearson: -0.41,

p=0.05) and communication (Pearson: -0.56, p=0.00). Concluding, the shorter the period of deafness and the longer the period of CI use, the better the performance in various SIFTER areas. Subdividing into groups based upon duration of deafness (less than 2 years, 2 to 4 years and more than 4 years) showed significantly higher SIFTER results, in the domains of communication (all children, Chi square test 0.02), attention and class participation (elementary school, Chi square test respectively 0.05 and 0.02), in the groups with a shorter period of deafness, best scores obtained in the group with shortest period of deafness. Children, who were deaf over 4 years, obtained worse scores in all three domains.

The only audiologic test that showed a weak relation with the SIFTER was the GN phoneme score, it correlated positively with the area academics (Pearson: 0.47, p=0.03).

Thus, it was shown that better speech recognition scores do not always implicate better classroom performance (in AMP or SIFTER).

Table 3. Results SIFTER

| **SIFTER kindergarten** | CI + (n=6) | | CI – (n=6) | | Significance CI+ vs. CI- |
|---|---|---|---|---|---|
| | Mean (SD) | Outcome | Mean (SD) | Outcome | |
| Pre academics | 10.33 (1.86) | Sufficient | 12.00 (0.63) | Sufficient | 0.12 |
| Attention | 10.67 (2.25) | Sufficient | 12.83 (0.98) | Sufficient | 0.08 |
| Communication | 8.83 (1.47) | Marginal | 12.17 (1.33) | Sufficient | *0.01* |
| Class participation | 11.33 (3.20) | Sufficient | 14.17 (1.17) | Sufficient | 0.10 |
| Social behavior | 11.00 (2.45) | Sufficient | 11.17 (1.47) | Sufficient | 1.00 |
| **SIFTER elementary** | CI + (n=*) | | CI – (n=**) | | Significance CI+ vs. CI- |
| | Mean (SD) | Outcome | Mean (SD) | Outcome | |
| Academics | 10.38 (2.52) | Sufficient | 12.15 (1.79) | Sufficient | *0.01* |
| Attention | 8.52 (2.79) | Sufficient | 10.96 (2.32) | Sufficient | *0.00* |
| Communication | 7.32 (2.53) | Failure | 11.43 (2.01) | Sufficient | *0.00* |
| Class participation | 9.17 (2.63) | Sufficient | 12.33 (2.25) | Sufficient | *0.00* |
| School behavior | 12.58 (2.23) | Sufficient | 13.48 (1.67) | Sufficient | 0.13 |

*: academics, class participation and school behavior n=24. Attention and communication n=25.
**: academics, class participation and school behavior n=27. Attention and communication n=28.
Comparison of children with and without CI; Means (Standard Deviation) and significance (by use of exact Mann-Whitney U test, $p < 0.05$ is significantly different) were computed for the 5 SIFTER content areas . Meaning of the mentioned numbers, the outcome, is shown as well

## Questionnaire comparison

Comparison of AMP and SIFTER results showed an obvious (significant) correlation between both questionnaires. In kindergarten, where the emphasis generally lies on social interaction and learning through play, the AMP-K score correlated positively with 4 areas of the SIFTER: pre academics (Pearson: 0.65, p=0.02), attention

(Pearson: 0.79, p=0.00), class participation (Pearson: 0.92, p=0.00) and social behaviour (Pearson: 0.58, p=0.05). The AMP-E score correlated positively with all SIFTER content areas; academics (Pearson: 0.63, p=0.00), attention (Pearson: 0.62, p=0.00), communication (Pearson: 0.74, p=0.00), class participation (Pearson: 0.78, p=0.00) and social behaviour (Pearson: 0.47, p=0.00). Class ranking correlated positively with the questionnaire results, AMP and SIFTER, of elementary school children. Thus the outcomes of both questionnaires seemed to reflect the description of the teacher of their students. Larger standard deviations within the CI group in results of AMP-K, AMP-E, and SIFTER [Table 2,3] suggested that the individual variance in the group of CI children was more pronounced than in the group of the normal hearing peers.

On an individual level, analysing the matched pairs, formed by classmates, showed no significant differences in class ranking or mean AMP scores. In the SIFTER only one content area varied significantly between the CI students and their normal hearing classmates; in elementary school the normal hearing children outperform the CIs in the area of communication (p=0.04).

**Audiologic results**

Longitudinal audiologic data of the CI children (high Fletcher Index, Bosman score and GN word and phoneme scores) are shown in Figure 2. All audiologic test results show an improving trend. The mean high Fletcher Index decreases from 50 dB shortly after first fitting to reach a plateau of approximately 35 dB in 3 to 4 years post-implantation. The mean Bosman and GN scores showed improvement till 6 years after implantation. Inter-test comparison demonstrated significant correlation between hFI and GN phoneme (Pearson –0.42, p=0.02), hFI and Bosman (Pearson –0.45, p=0.04) and strongly between both the GN tests (Pearson 0.95, p=0.00). The GN phoneme score correlated negatively with duration of deafness (Pearson -0.45, p=0.01) and age at implantation (Pearson –0.48. p=0.01). The period of CI use did not correlate significantly with any of the audiologic test results, but it did show a strong relationship with the number of years children were in mainstream education (Pearson 0.68, p=0.00). When comparing the audiologic data of the congenitally and prelingually deaf, no significant differences were seen.

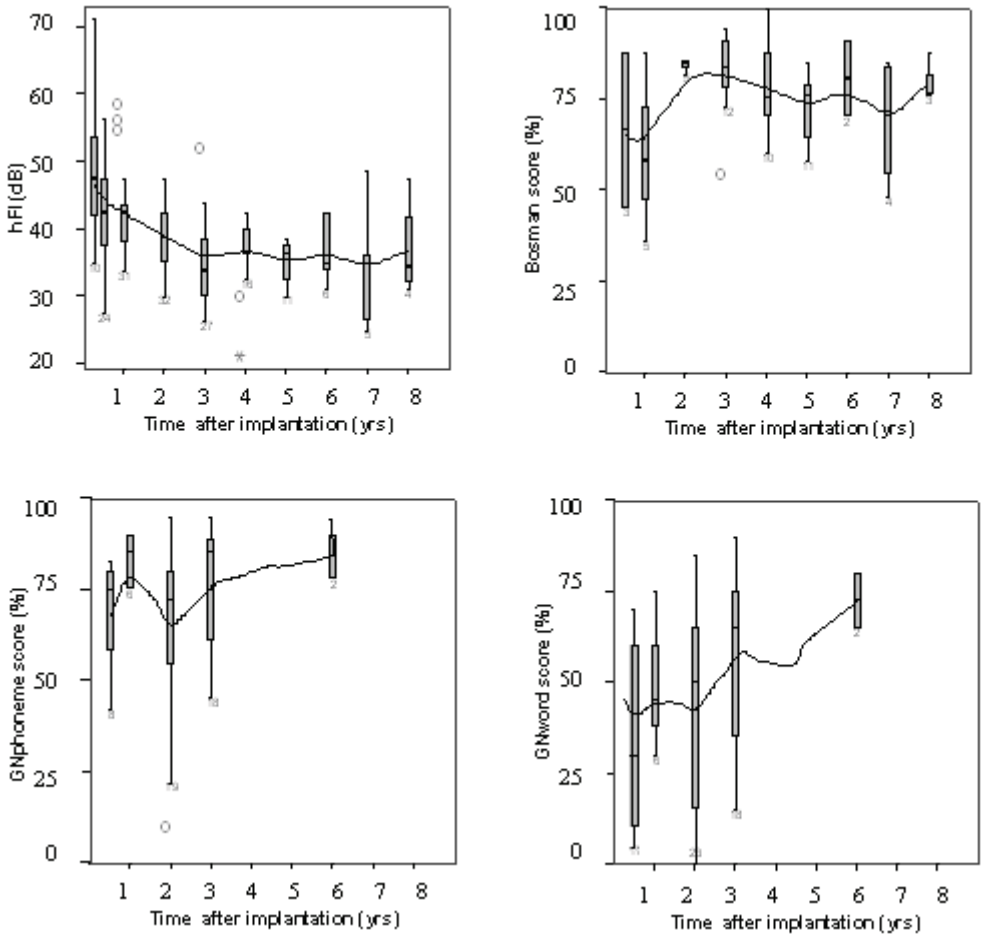


Figure 2. High Fletcher Index and speech perception scores. Boxplots show the longitudinal data of the implanted children of diverse audiologic tests performed at different times after implantation. Interquartile range (IQR), median, maximum and minimum within 1,5 IQR and outliners are shown. The thin line is a smooth fit line, connecting the means and showing the time trend in each graph.

# Discussion

All the CI students in the present study scored "above average" in the AMP, which is a remarkable positive finding. Nevertheless, a few specific questions did show a deprivation in communicative skills of CI students compared to their normal hearing peers. The AMP questionnaire has so far only been used in one other study[17]. Preliminary results of this previous study showed communicative difficulties for the CI students. The questionnaire still has to be objectified by retesting this questionnaire with large groups of CI, hard of hearing and normal hearing students. An outcome of this further analysis could be that the current AMP does not differentiate enough. AMP ranking could be objectified by comparing the scores with, for example, randomised intelligence tests. In the present study, one must take into account that the

children who entered mainstream schools are generally good performers. In the beginning of the mainstreaming of CI students in the Netherlands, only the best children got the chance to enter a very select number of schools. Nowadays, more children enter different schools in all parts of the country. It should be noticed though, that this could be an exceptional situation for the Netherlands. In our country there is a large variability in schools for deaf and hard of hearing students, so at the moment, generally only the children who are regarded capable of mainstream schooling by the CI-team enter mainstream, while other implanted children enter schools with systems more suitable for their specific needs. This may not be the case in other countries where schools for hard of hearing pupils may not be present or difficult to enter. Results from CI students in mainstream from all over the world may therefore not be compared automatically without further analyses, as entering levels could differ. Nevertheless, it would be very interesting to compare communicative performance of CI children in different countries to determine whether the expected differences will be seen.

The significantly lower mean AMP-E score of the congenitally deaf in regard to the prelingually deaf children could theoretically be explained by the fact the prelingually deaf have had normal hearing before turning deaf. Niparko et al. concluded that greater speech and language proficiencies may be expected from children who exhibit normal hearing for even a brief period after birth and receive a cochlear implant shortly after losing their hearing[18]. Data of Sharma et al. suggested that in the absence of normal stimulation, as in congenitally deaf children, there is a short sensitive period of about 3.5 years during which the human central auditory system remains maximally plastic, after that, plasticity is greatly reduced[19]. The twenty congenitally deaf children in our study were implanted between 1.0 and 7.6 years, only 6 before the age of 3.5 years and 2 children a couple of months later. Questionnaire and audiologic results from those congenitally deaf children did not, in contradiction with Sharma et al.[19], differ significantly from the congenitally deaf children implanted at older age.

The SIFTER questionnaire has an internal reference as the teacher is asked about the pupils level in comparison to other students. In relation to the main question in this study how CI children communicatively perform in mainstream education with regard to their normal hearing peers, the SIFTER outcome showed the CI children were delayed in communication in kindergarten as well as in elementary school. The teachers answered the three questions within the area of communication, on expressive and receptive language skills, less reassuringly for the CI students. In the SIFTER manual has been described that the cut off scores for the three categories (failure, marginal and sufficient) are to be interpreted as guidelines as opposed to highly accurate decision points. The difficulty remains, how to interpret numerical

statistical differences between the CI group and the normal hearing group within the same category. We feel that those differences could indicate a probable risk of a gliding scale towards serious difficulty in mainstream school. The preschool SIFTER manual described the means of large groups of hearing (n=114) and hard of hearing (n=110) children in all 5 areas. The outcomes of the CI children in this study are most comparable to children hard of hearing. The score difference between the CI and normal hearing group in this study was comparable to the score difference between the hard of hearing and normal hearing group from the manual. On communication in particular, the manual and the outcome of this study showed a greater difference between the normal hearing group and the CI group, hypothetically indicating the specific difficulty formerly deaf children with CI encounter in the classroom[10].

In the current study, the Bosman score and GN word and phoneme scores showed an improving trend over time [Figure 2]. Speech perception is known to improve in the years after implantation[7;20-22]. However, there are individual differences influenced by not only the quality of the sensory stimulus of the implant, but also by linguistic, world, and social knowledge[21].
In our study, two implant variables seemed to have a large effect on the questionnaire results: the duration of deafness and age at implantation. Duration of deafness correlated negatively with the AMP-E score and with four content areas of the SIFTER elementary school, as well as with the area of communication in the preschool SIFTER. Among others, Gordon et al. already discussed the fact that children who have spent many years without sufficient auditory stimulation, tended to achieve poorer speech perception skills than their peers with shorter durations of deafness[5]. Age at implantation correlated negatively with the AMP-E score. This finding was confirmed by the outcome of the SIFTER, which revealed a negative correlation between four content areas and age at implantation and by the obvious negative correlation between the age at implantation and GN phoneme scores. Many studies described the influence of age at implantation on speech perception and oral communication abilities[1;3;7;20;24-31]. It has been demonstrated that prelingually deaf children who were implanted before the age of 5 achieved significantly better outcomes in communication than children implanted at older age[1;32]. Govaerts et al. concluded their data added evidence to the importance of early implantation (before the age of 2 years) of congenitally deaf children[30]. Children with profound sensorieural hearing loss[20] or prelingually deaf children[32] who received their implants during preschool demonstrated on average stronger performance in speech production and perception over time than children who received the implant during their elementary school years. The children in Geers' study were all prelingually deaf and implanted before the age of 5, the mean age at implantation being 3.3 years[32]. As the current study contained congenitally deaf children partly implanted above the age of 5 as well,

not all results were comparable. However, in our study it was shown that better speech recognition scores do not always implicate better classroom performance. Besides speech perception, it is acknowledged that language development could have played a substantial role in the performance of CI children in mainstream educational settings. Therefore, further research will be done with analyses of receptive and expressive language development in combination with classroom performance.

# Conclusion

Cochlear implant students seem to perform well in mainstream education according to the AMP and SIFTER scores. Mean scores in the AMP questionnaire of overall class ranking were even above average for all students. Nevertheless, in the area of communication in the SIFTER, the CI pupils failed or scored marginal. The CI group scored significantly less than their normal hearing peers in most questionnaire domains of both the AMP and the SIFTER. The beneficial audiologic effect of the CI increased over time, as was seen in the improving results of speech perception tests. Age at implantation and the duration of deafness were important variables for the communicative outcome in this study. In children with a long period of deafness and/or late age at implantation, extra attention should be considered when entering mainstream education.

The AMP questionnaire can be a very useful tool in future, though it has to be retested in large number studies to evaluate its power and to prove its value for rehabilitation follow-up. Ambulatory coaches can use the questionnaire to determine whether a child is communicatively successful in mainstream education and it may help counselling of the parents of the child and teacher throughout the child's education. The ultimate goal is to use the AMP as a means of determining placement in the mainstream. Since the coaches made some of this determination the data for this set may be skewed but still useful as it provides information for children who should be successful.

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## **APPENDIX 1.** AMP elementary school

Assessing Mainstream Performance (AMP)
for Deaf and Hard of Hearing Students:
Elementary/High School

Pat Chute, Ed.D., Mary Ellen Nevins, Ed.D. Nicole Czarnecki, M.S.
The Children's Hearing Institute (CHI), Inc.

Student ID Number:_____ Date:_____
Grade Level: _____ Age:_____
Type of Device _____

THE CLASSROOM

Position of professional completing form: (circle appropriate)
Reg. Ed. Teacher Teacher of the Deaf Speech-Lang. Pathologist
Other (please specify):_____

Total years teaching experience:_____ Experience with this child_____

Previous experience with *Deaf/Hard of Hearing* students: Yes No
If yes, please explain:_____
_____
Previous experience with *Cochlear Implant* students: Yes No
If yes, please explain:_____
_____

Total number of students in class:_____
Number of years child has been in mainstream (not including this year):_____

Other personnel in room (circle all that apply): aide interpreter cued speech transliterator
other teacher FT other teacher PT

List type and amount of special services the child receives _____
_____

Assistive listening device(s) used by student (e.g., hearing aid(s), cochlear implant, personal or soundfield FM system). List all: _____

Please list all sound-absorbing treatments in classroom (e.g., carpeting, window treatments, acoustic tiles):
_____
_____

******PLEASE ANSWER ALL ITEMS. DO NOT LEAVE ANY ITEMS BLANK.******

STUDENT PERFORMANCE

1. **Percentage of time student demonstrates a general response to speech through audition only (e.g., cessation of activity, head turn):**
   □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

2. **Percentage of time student spontaneously attempts to communicate with others using speech-only:**
   □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

3. **Percentage of time student spontaneously attempts to communicate with others using any modality:**
   □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

4. **Percentage of time student demonstrates communication breakdown:**
   □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

5. **Percentage of time student spontaneously attempts to imitate models (speech/language provided):**
   □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

6. **Percentage of time student successfully follows verbal or signed directions:**
   □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

7. **Percentage of time student attends during teacher-directed activities:**
   □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

8. **Percentage of time student is an active participant in teacher-directed activities:**
   □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

9. **Percentage of time student makes a comment that is on-topic:**
   □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

10. **Percentage of time student makes a comment that is off-topic:**
    □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

11. **Percentage of time student makes a comment that is enriching to the discussion:**
    □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

12. **Percentage of time student comprehends classroom instruction:**
    □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

13. **Percentage of time student gives some indication that content was not understood:**
    □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

14. **Percentage of time student participates in typical recitation activity:**
    □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

15. **Percentage of time student responds or comments in lectures/teacher-directed activities:**
    □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

16. **Percentage of time student is engaged in group discussions:**
    □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

17. **Percentage of time student displays general turn-taking abilities:**
    □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

18. **Percentage of time student initiates general peer interactions through verbal means and/or sign:**
    □ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

**19. Percentage of time student exhibits distractibility when extraneous visual and/or auditory stimuli are present:**
□ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

**20. Percentage of time student demonstrates familiarity with classroom routines:**
□ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

**21. Percentage of time student willingly takes learning risks or participates in challenging tasks:**
□ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

**22. Percentage of time student assumes a leadership role in class:**
□ 0-10% □ 11-30% □ 31-50% □ 51-70% □ 71-90% □ 91-100%

Please describe any areas of concern or special needs for student:_____
_____
_____
_____

**Additional comments about student:**

_____
_____
_____

Based on the entire class, where does this student fall in the overall class rank:
□ top 91-100% □ 75-90% □ 51-74% □ 26-50% □ 0-25%

1.2a

# Patient with Usher syndrome Type I; is cochlear implantation beneficial?

Quality of life and cochlear implantation in Usher syndrome type I
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# Abstract

*Aim:* To evaluate quality of life, hearing and vision in Usher syndrome type I patients with and without cochlear implant
*Design:* descriptive retrospective study
*Methods:* Quality of life (QoL) of fourteen Usher type 1 (USH1) patients with a cochlear implant (7 adults, 7 children) was compared to those of fourteen USH1 patients without a CI (12 adults, 2 children) by means of three questionnaires; NCIQ, SF12 and the Usher Lifestyle Survey. Additional information on hearing level was obtained by the Equivalent Hearing Loss (EHL) principle and on the visual deterioration by the Functional Vision Score (FVS).
*Results:* A significant benefit of CI was seen in the hearing specific questionnaire NCIQ. This difference could not be detected in the generic SF12 survey. The Usher Lifestyle survey indicated that USH1 patients with a CI tend to be able to live an independent life more easily than the profoundly deaf unimplanted USH1 patients. EHL and FVS scores varied in both groups.
*Conclusions:* Overall quality of life can be enhanced by CI in USH1 patients, though effects are mostly seen in hearing related QoL-items.

# Introduction

Usher Syndrome is an autosomal recessive disorder that causes bilateral sensorineural hearing impairment, retinitis pigmentosa (RP), and occasionally vestibular dysfunction (Usher 1935). The prevalence of Usher syndrome is estimated at 3.5 to 6.2 per 100,000 inhabitants[1-3]. Three clinically different syndromes, designated Usher syndrome type I, II and III (USH1, 2 and 3) can be distinguished by differences in audiovestibular features[4]. USH1 is characterised by congenital profound deafness, vestibular areflexia and progressive vision loss due to RP. The disease progresses over the years with deterioration of visual field size and visual acuity, finally causing blindness or severe visual impairment. The mean age of RP diagnosis is estimated to be 13-14 years[4].

Cochlear implantation (CI) has a major role to play in the rehabilitation of deafblind individuals, and these patients may be among those most worthwhile to consider for implantation. Cochlear implantation is relevant in USH1 patients because of the combination of congenital profound deafness and visual deterioration. Several studies have shown the benefit of (early) implantation in USH1 children[5,6] to be indisputable. It has been stated that, in association with early implantation, speech therapy should start early to maximize the auditory-oral communication before the onset of loss of vision[6]. Cochlear implantation in deafblind adults was proven to be as successful as in other prelingually deaf adults; scores on closed-set speech recognition tests were comparable[5-7].

In this study a survey of the results of cochlear implantation in USH1 patients with regard to hearing (Equivalent Hearing Loss EHL), vision (Functional Vision Score FVS) and quality of life is presented. In a separate paper, the audiologic results of cochlear implantation and its benefit for these patients are described and correlated to the underlying genetic diagnosis[8]. As quality of life measurements three different questionnaires were used: a specific Usher questionnaire (the Usher lifestyle survey), a specific hearing impairment/CI questionnaire (NCIQ) and a generic health state questionnaire (SF 12). Results from fourteen USH1 patients with CI (7 adults, 7 children) were compared to the results of USH1 patients who have not been implanted (12 adults, 2 children).

# Materials and Methods

## Patients

*USH1 with CI:*

Sixteen USH1 patients, with a clinical diagnosis based upon audiovestibular and ophthalmologic examinations, had received a CI at the cochlear implant centre Nijmegen-St Michielsgestel in the past years and were asked to participate in this study. Fourteen patients (seven children and seven adults [1-14]) agreed to participate. One patient was staying abroad and hence could not participate and the other non-participant was a teenager, whose parents thought it was better for him not to participate. Written (parental) informed consent was obtained for this study. The average age of the children was 12.4 years (SD 2.9 yrs) and of the adults 30.7 years (SD 6.8 yrs). Four of the seven adults mentioned to have and live with a partner (three adults lived alone) and also four, though not exactly the same, had paid employment. All children were living with their families. Further demographic characteristics and implant data are shown in Table 1.

*Control group:*

For the control group, the patient database of Viataal, St Michielsgestel, the main Dutch institute for deaf and blind patients, was searched for USH1 patients. Thirty-one USH1 patients were selected, the diagnosis based on the same clinical findings as described above. The group of control patients was matched to the patient group as accurately as possible with the demographic characteristics. Fourteen patients [15-28] finally participated in the study. Ten patients replied that they were unwilling to cooperate, two further patients had received a CI and three others had a foreign nationality and were excluded (difficulty in understanding the questionnaires). The average age of the adults in the control group was 36.9 years (SD 16.0 yrs). Only 2 un-implanted children were included, aged 16.5 and 14.3 years. Four of the twelve adults had a partner and seven of them had paid employment. Both children lived at home with their families, two adults lived alone, nine lived together with others and one lived in an institution. Further demographic characteristics of the control group are also shown in Table 1. Informed consent was obtained similarly to the CI-group.

## Hearing

The principle of 'equivalent hearing loss' (EHL) was used to describe the audiologic performance of the CI-users. Scores on a battery of speech perception tests were reduced to one single measure, called the EHL, as described elsewhere[9].

Table 1. Patient characteristics

| Patient | Gender | Age (yrs) | Age at implantation | Follow-up period | Cochlear Nucleus | EHL (dBHL) | Test age (yrs) | FAS (%) | FFS (%) | FVS (%) |
|---|---|---|---|---|---|---|---|---|---|---|
| 1 | M | 14,3 | 9,1 | 5,1 | N24 | 115 | 8,9 | 98 | 58 | 57 |
| 2 | F | 12,2 | 6,9 | 5,2 | N24 | 103 | 6,7 | 90 | 79 | 71 |
| 3 | M | 22,8 | 11,8 | 11,0 | N22 | 94 | 17,6 | 90 | 59 | 53 |
| 4 | M | 10,7 | 3,7 | 7,0 | N24 | 70 | 8,5 | 93 | 66 | 61 |
| 5 | F | 21,9 | 10,9 | 11,1 | N22 | 90 | 21,8 | 100 | 53 | 53 |
| 6 | F | 17,4 | 6,9 | 10,5 | N24 | 72 | 16,9 | 89 | 48 | 43 |
| 7 | M | 27,7 | 13,5 | 14,3 | N22 | 107 | 23,0 | 76 | 46 | 35 |
| 8 | M | 33,4 | 22,8 | 10,6 | N22 | 120 | 17,4 | 90 | 28 | 25 |
| 9 | F | 33,6 | 20,6 | 13,0 | N22 | 110 | 32,6 | 95 | 51 | 48 |
| 10 | F | 35,8 | 20,1 | 15,7 | N22 | 114 | 32,1 | 90 | 39 | 36 |
| 11 | M | 39,9 | 30,4 | 9,4 | N22 | 115 | 34,3 | 94 | 58 | 55 |
| 12 | F | 8,4 | 3,5 | 4,8 | N24 | 83 | * | * | * | * |
| 13 | M | 12,0 | 9,0 | 3,0 | N24 | 78 | * | * | * | * |
| 14 | M | 11,4 | 4,7 | 6,6 | N24 | 70 | * | * | * | * |
| 15 | F | 39,9 | | | | | 35,5 | 53 | 73 | 39 |
| 16 | M | 46,7 | | | | | 44,6 | 21 | 89 | 19 |
| 17 | M | 20,4 | | | | | 19,8 | 18 | 93 | 17 |
| 18 | F | 19,2 | | | | | 19,0 | 55 | 99 | 54 |
| 19 | M | 41,2 | | | | | 36,7 | 41 | 72 | 30 |
| 20 | M | 35,8 | | | | | 25,0 | 44 | 59 | 26 |
| 21 | M | 32,7 | | | | | 32,7 | 53 | 90 | 48 |
| 22 | M | 36,4 | | | | | 29,2 | 40 | 89 | 35 |
| 23 | F | 23,4 | | | | | 23,1 | 53 | 84 | 36 |
| 24 | M | 49,3 | | | | | 46,2 | 45 | 80 | 36 |
| 25 | F | 21,8 | | | | | 21,9 | 50 | 99 | 50 |
| 26 | F | 76,0 | | | | | 74,4 | 50 | 80 | 40 |
| 27 | M | 16,5 | | | | | 16,3 | 99 | 99 | 98 |
| 28 | F | 14,3 | | | | | 13,4 | 56 | 90 | 51 |

Gender: Male (M) or Female (F), * missing data. EHL= Equivalent Hearing Loss in decibel hearing level (dBHL). Test age: age at time of investigation, FAS: Functional Acuity Score, FFS: Functional Field Score, FVS: Functional Vision Score (American Medical Association Chapter 12 The visual system. In: Cocchiarella L & Anderson GBJ (eds) Guides to the Evaluation of Permanent Impairment, 2001).

The EHL may vary between 50 and 130 dB hearing level (HL). For example, an EHL of 90 dB reflects speech recognition abilities comparable to a person with a 90 dB hearing loss fitted with hearing aids. EHL was measured at different moments after implantation, but only the most recent test results, which were obtained at least 3 years after surgery, were used in further analysis.

**Vision**

Data of visual function were obtained from the Vision Centre of Viataal in St Michielsgestel. The last-visit measurement was selected for cross-sectional analysis. Visual acuity (VA) was calculated according to regular standards by using Snellen charts. Best-corrected measurements of both eyes were used for further evaluation. The VA measurement was converted into a Functional Acuity Score (FAS)[10]. The visual field size was evaluated by Goldmann perimetry of both eyes and in most cases the isopters for the V-4, III-4 and I-4 test targets were measured. The Goldmann perimetric fields were quantified into a Functional Field Score (FFS) by plotting the III-4 isopter according to the description of the AMA in the Guides for the Evaluation of Permanent Impairment[10]. The Functional Vision Score (FVS) was determined by the FAS and the FFS based on the equation FVS = (FFS × FAS) / 100[10]. The FVS enabled us to interpret the percentage of vision score, a FVS < 50% indicating serious visual impairment in daily life. Visual characteristics of the patients are shown in Table I.

**Quality of Life**

Three questionnaires were used (summer 2004): the Nijmegen Cochlear Implant Questionnaire, the Usher Lifestyle survey and the Standard Medical Outcome Study Short-Form 12. As mode of administration for the CI users the in-person mode was chosen. The patients or their parents filled in the questionnaires at home supervised by the first author in all cases. Questionnaires were sent by mail to the control group and patients were offered help by email with filling in the questionnaires. As only signed language assistance would have been helpful while filling in the questionnaires (which is different from the orally assisted CI group), the in-person mode was not preferred in the control group. The parents of the USH1 children did not fill in the SF12 questionnaire, as it is not suitable for children. All three questionnaires are discussed below.

The Nijmegen Cochlear Implant Questionnaire (NCIQ) measures health-related quality-of-life (HRQoL). In contrast with two other questionnaires that were used, the NCIQ is a hearing handicap specific QoL questionnaire[11]. Questions comprise three general domains: physical, psychological and social functioning. Each domain can be divided further in subdomains, consisting of 10 items. These items are formulated as a statement and have five possible answers. If a statement did not apply for a patient, a

sixth answer could be given: not applicable. Final scores for the subdomains ranged from 0 to 100 (optimal).

The Usher Lifestyle survey is a descriptive questionnaire consisting of nine main questions, divided into several subquestions on the topic of maintaining independence. This questionnaire was developed in the United Kingdom by Sense[12,13]. In 1980 the Nordic countries agreed to a common definition of domains of independence that are adversely affected in deafblindness. The domains are: ability to give and receive information (communication), access to information and mobility. There are open-ended questions as well as questions with multiple choice answer possibilities. Answers to the open-ended questions were recoded.

The standard Medical Outcome Study Short-Form 12 (SF12) is a short questionnaire based on the SF36, which was based on a large battery of health status and HRQoL instruments employed in the Medical Outcomes Study[14]. This questionnaire is a non-disease specific or generic HRQoL instrument. The SF12 contains 12 items that measure eight dimensions: Physical functioning, Role functioning due to Physical health problems, Role functioning due to Emotional problems, Bodily Pain, Vitality, Social Functioning, Mental Health and General Health perceptions. The number of response choices per item ranges from two to six. Item scores for each dimension are coded, summed and transformed to a scale from 0 to 100, with higher scores indicating better self-perceived health. Mental and Physical health summary scores were computed.

**Data analyses**

Data were analysed using SPSS version 12.0. Descriptives were computed for main characteristics: age, sex, age at implantation, partnership, paid employment and living situation [Table 1]. Final results of the questionnaire domains were computed following the guidelines of each questionnaire. Results were compared for groups differing in age (adults versus children) and cochlear implant use. These differences were analysed by nonparametric independent sample Mann-Whitney U test for multiple responses for continuous data and by the Chi square tests for categorical data. Correlation analysis was done using Spearman's correlation.

# Results

The patients were implanted between 1988 and 2001, following standard procedures. The mean follow up time at the moment is 9.2 years with a minimum of 3.0 years and a maximum of 15.7 years. No complications were mentioned in the post operative period, though two patients needed revision surgery due to a failure of the CI. Finally,

after revision surgery, two different types of implants were used: 8 Nucleus 22 devices and 6 Nucleus 24 devices (Cochlear corporate, Englewood, Australia). In all implantation procedures full insertion was obtained. The mean age at implantation varied from 6.3 years (sd 2.4 yrs) in the children to 18.6 years (sd 7.1) in the adult group.

EHL speech perception tests were performed at six months and yearly after implantation. The EHL scores of the USH1 patients with CI vary from 70 dBHL in a young child to 120 dBHL. The mean EHL score of the implanted group was 95.8 dBHL, when dividing the group into adults and children the means were 107.1 dBHL and 84.4 dBHL respectively. The non-CI users were all profoundly deaf with PTA (pure tone average)-scores above 110 dB at 0.5, 1 and 2 kHz and speech scores at chance level. This resulted in EHL scores of 130 dBHL. In a separate study, the audiological results and benefits of cochlear implantation will be reported[8].

The FVS varied from 17% to 98%. In two children Goldmann perimetry was not performed and in one child different test targets were used (I-4 and V-4 instead of III-4, which is needed to compute the standard FVS). Hence three children did not have the required data to be included in further FVS analyses. Mean FVS of the total USH1 group (n=25) was 45% (sd 17.4); when dividing the group into adults and children the means were respectively 39% (range 17 to 55%) and 63% (range 43 to 98%). As can be seen in Figure 1 the FVS correlates significantly with age. Vision deteriorates as patients get older.

**Questionnaire results**

**NCIQ**

First USH1 patients with and without CI were compared. Both adults and children with CI seem to perform better in this specific hearing related QoL questionnaire in multiple domains [Table 2]. Significant differences with favourable results in the patients who underwent cochlear implantation were seen in the domains of 'sound perception basic' and 'advanced' for both adults and children. In the domains of 'activity limitations' and 'social interactions' children with CI also scored significantly better than unimplanted children. When comparing adults and children with CI, the children seemed to benefit more; in both domains of sound perception they scored significantly better than the adults. A comparison in the children and adults without CI did not show any difference.

Table 2. NCIQ results

| | Adult CI + | | | Child CI + | | | Adult CI - | | | Child CI - | | Adult CI + vs. Child CI +* | Adult CI + vs. Adult CI -* |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | | | | | | Nº27 | Nº28 | | |
| | N | Mean | Range | N | Mean | Range | N | Mean | Range | score | | | |
| Sound perception basic | 7 | 47.4 | 5.0-85.0 | 7 | 75.4 | 52.5-85.0 | 10 | 6.5 | 0.0-55.0 | 0.0 | 42.5 | **0.04** | **0.00** |
| Sound perception advanced | 7 | 48.3 | 27.8-80.0 | 7 | 67.9 | 32.5-95.0 | 9 | 24.0 | 2.8-65.0 | 10.0 | 32.5 | 0.12 | **0.03** |
| Speech production | 7 | 25.4 | 12.5-42.5 | 7 | 42.5 | 22.5-57.5 | 9 | 32.1 | 15.0-60.0 | 20.0 | 75.0 | **0.04** | 0.36 |
| Self-esteem | 7 | 70.0 | 37.5-90.0 | 7 | 65.4 | 40.0-90.0 | 9 | 51.3 | 25.0-89.3 | 47.5 | 52.5 | 0.63 | 0.07 |
| Activity limitations | 7 | 76.0 | 32.5-90.0 | 7 | 74.2 | 63.9-88.9 | 10 | 58.6 | 22.5-93.8 | 60.0 | 47.2 | 0.84 | 0.11 |
| Social interactions | 7 | 65.0 | 37.5-80.6 | 7 | 70.9 | 60.0-77.5 | 10 | 55.8 | 27.5-75.0 | 60.0 | 47.5 | 0.35 | 0.24 |

Results of the six domains of the NCIQ for the three different USH1 groups and the two USH1 children without CI are shown. Separate data (N= number, Mean= mean score and Std. Dev.= standard deviation) as well as the comparative results of the nonparametric Mann Whitney U tests are presented. Significant differences, where p is <0.05, are bold type numbers. * p-values

Table 3. Usher Lifestyle survey, Maintaining independent

| Maintaining independent in deafblindness | | CI+ Adult | | | CI+ Child | | | CI- Adult | | | CI- Child | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | n | median | range | n | median | range | n | median | range | Nº27 | Nº28 |
| Access to information | Wake up | 7 | 2 | - | 7 | 2 | 1-3 | 12 | 2 | - | 2 | 2 |
| | Front door | 7 | 2 | 2-3 | 7 | 1 | 1-2 | 12 | 2 | - | 2 | 2 |
| | Access form | 4 | 2 | 1-3 | 4 | 3 | 2-3 | 9 | 2 | 1-3 | 3 | 2 |
| | Emergency | 7 | 3 | 2-4 | 7 | 3 | 2-4 | 11 | 3 | 1-4 | 3 | 4 |
| Communication | No. telephone | 7 | 2 | 0-4 | 7 | 1 | 0-3 | 12 | 2 | 0-3 | 2 | 1 |
| | No. written information | 7 | 5 | 2-6 | 7 | 5 | 4-5 | 12 | 3 | 1-6 | 2 | 2 |
| | Buy food | 7 | 0 | 0-1 | 7 | 0 | 0-1 | 11 | 0 | 0-1 | 0 | - |
| | Communicate doctor | 7 | 1 | 0-1 | 7 | 1 | - | 12 | 1 | - | 0 | 1 |
| Mobility | Visit shop | 7 | 0 | 0-1 | 7 | 0 | 0-1 | 11 | 0 | 0-1 | 0 | - |
| | Visit doctor | 7 | 1 | 0-1 | 7 | 1 | - | 12 | 1 | - | 1 | 1 |

Access to information: 1=independently, 2= with special equipment, 3= with others, 4= not aware. Communication: number of ways in which a telephone or written information is used, buy food/communicate doctor 0= no help, 1= with others. Mobility: 0= no help, 1= with others. Results of the three domains of the Usher Lifestyle survey for the three different USH1 groups and the two USH1 children without CI are shown (n= number).

## Usher Lifestyle Questionnaire

The Nordic definition of maintaining independence in deafblindness is based on tree domains: access to information, communication and mobility [Table 3]. In the domain of 'access to information', two significant differences were seen. Children with CI need less help of others or equipment to know that someone is at the front door than adults. Subjects without a CI answered the question about knowing whether an emergency situation occurs at home more reassuringly than subjects with CI: CI users, especially children, tend to need more equipment to detect such a situation or are afraid they will not notice at night. The domains of 'communication' and 'mobility'

both show that CI users need less help of others and less equipment. Overall, a trend is seen that USH1 patients with CI maintain independent more easily than unimplanted USH1 patients.

**SF12**

This generic questionnaire was filled in by adult patients only. When comparing USH1 adults with and without CI, no significant differences or trends could be detected in any of the eight domains or in the mental or physical health summary score [Table 4].

Table 4. SF12 results

| | CI + Adult | | | CI - Adult | | | CI+ Adult Vs. CI- Adult* |
|---|---|---|---|---|---|---|---|
| | N | Mean | Range | N | Mean | Range | |
| General Health | 7 | 71.4 | 25-100 | 12 | 81.3 | 25-100 | 0.42 |
| Physical Health | 7 | 100.0 | - | 12 | 83.3 | 25-100 | 0.12 |
| Role Functioning Physical | 7 | 67.1 | 0-100 | 12 | 79.2 | 10-100 | 0.47 |
| Role Functioning Emotional | 7 | 80.0 | 0-100 | 11 | 67.3 | 20-100 | 0.43 |
| Pain | 7 | 75.0 | 25-100 | 11 | 88.6 | 50-100 | 0.22 |
| Emotional wellbeing | 7 | 68.6 | 30-90 | 11 | 74.6 | 40-100 | 0.55 |
| Energy | 7 | 60.0 | 20-100 | 11 | 60.0 | 20-100 | 1.00 |
| Social Functioning | 7 | 78.6 | 0-100 | 11 | 75.0 | 25-100 | 0.82 |
| Physical summary score | 7 | 52.1 | 42.9-61.5 | 11 | 52.6 | 31.9-68.2 | 0.72 |
| Mental summary score | 7 | 46.4 | 15.7-57.2 | 11 | 46.7 | 23.0-59.4 | 0.72 |

Results of the eight domains of the SF12 for two adult USH1 groups are shown. Separate data (N= number, Mean= mean score) as well as the comparative results of the nonparametric tests are presented. * p-values

**Correlations**

Correlation analysis showed a significant correlation between the EHL and two domains of the NCIQ. When hearing impairment worsens (EHL increases), the 'sound perception basic' and the 'sound perception advanced' decrease significantly (correlations $p<0.01$, $n=28$). This correlation is not seen when analysing the EHL and the results of the generic health state questionnaire SF12. Investigating correlations between the FVS and the specific (NCIQ) or generic (SF12) questionnaires did not show any significant results.

# Discussion

Over the past years, cochlear implantation has evolved to become an established means of providing auditory perception to profoundly deaf individuals. As experience

with the assessment, surgery and rehabilitation of deaf patients has increased, the selection criteria also have changed. The global experience of implanting deaf-blind subjects is growing, but there are still only a few reports in literature discussing the results of cochlear implantation in deaf-blind patients[6,7,15].

The present data show that most people with Usher syndrome receive significant benefit from cochlear implantation in specific areas. EHL results illustrate an increase in speech perception abilities after implantation[8]. This increase in speech perception is also subjectively reported in the NCIQ results. The beneficial effect of CI in Usher patients confirms previous findings[7]. Loundon et al. reported this important benefit in speech perception after implantation as well and reported a relation with the age of implantation[6]. In addition, it should be noticed that in the present study EHL scores were not measured at the moment when the questionnaires were filled in. It was assumed that all patients had reached a plateau score as they all used their CI for at least three years.

To determine the impact of visual impairment the FVS was used. The FVS has proven to be a reliable and straightforward method for evaluating vision. A previous study showed that the FVS decreases with age in USH1 patients[16]. No studies have been conducted on quality of life and visual impairment in USH patients. Investigating the impact of retinitis pigmentosa on quality of life showed perceived difficulties in daily activities, which correlated with the actual clinical measures of visual function[17].

Few studies were conducted on cochlear implantation in visually impaired or multi handicapped patients. In 1994 four Usher patients were described by Hinderink et al[7]. El Kashlan et al. have studied a group of 8 visually impaired implanted patients, among who two USH patients (one child, one adult). A beneficial effect in speech perception tests, both open and closed set, was seen[5]. Even when there was a larger variability in etiology of multi handicapped children, speech perception results in open and closed sets improved after implantation[18]. Loundon et al. mentioned that there was no relation seen between the visual acuity and the, good, logopedic results in 13 implanted USH patients (11 of which USH1)[6].

Questionnaires were selected to assure a wide range of quality of life issues: generic, specific on USH syndrome and specific hearing/CI related. The implanted USH1 group filled in the questionnaires with an interviewer in person, whereas the unimplanted USH1 group received the questionnaires per mail and was offered help where needed (patients preferred way of administration). This could have caused a bias and difference in answers. Nevertheless, some domains in the SF12 (which do not reflect immediately the effect of a CI, for example 'pain') did not differ significantly between in personal and postal questionnaires results. The social domains of the NCIQ do not differ between implanted USH1 patients and non-implanted USH1 controls. Previously the NCIQ was used in postlingually deaf adults, so this might compromise a comparison. As expected, the results of implanted USH adults were

worse in four of the six domains of the NCIQ compared to previous results in postlingually deaf adults. In the unimplanted patients the three directly hearing related domains had worse scores compared results of to the earlier study[19].
The generic questionnaire, SF12, does not show a difference in overall quality of life in adults as well, suggesting only a benefit in the specific hearing domains of the quality of life questionnaires. In the Usher Lifestyle survey it was found that USH people with CI seem to be able to live a more independent life. One outcome was unexpected: people without a CI seemed to need less equipment or help of others to know when an emergency situation occurred at home. Possibly, people with a CI relied more on their implant and hence were afraid of what would happen at night when their implant was not operational. Whereas USH1 people without an implant are used to this feeling, some mentioned to have expanded their trust in for example the alarm function of their olfactory system. Furthermore, the children depended on their parents in such situations and the CI children in this study outnumbered the unimplanted CI children.
A significant positive impact on the quality of life of deafblind individuals who receive a CI was expected by others[5]. This present study supplies proof that this is not always the case; quality of life should be split into specific (hearing related) and generic (health and social circumstances) quality of life.

# Conclusion

Cochlear implants are important for most patients with USH1 in helping to remain independent during daily life. Overall quality of life can be enhanced by a CI, though especially hearing and hearing related quality of life items improve after cochlear implantation. The EHL seems to be a good measurement for hearing in people with CIs. EHL is related to disease specific QoL. The FVS is an instrument to determine the impact of vision loss as it combines visual acuity and visual field into one overall score. Measures such as the EHL and the FVS can have potential both in research settings and in the evaluation of clinical services for the purposes of audit and clinical governance.

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1.2b

# Patient with Usher syndrome Type I; is cochlear implantation beneficial?

Audiological performance and benefit of cochlear implantation in Usher syndrome type I
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# Abstract

*Aim:* to evaluate the benefit and performance of cochlear implantation in Usher syndrome type 1 (USH1) patients.
*Methods:* 14 patients with a clinical diagnosis of USH1 were included. Mutation analysis of USH1 genes was performed in all of them. All patients filled in the G(C)BI questionnaire, which measures the benefit of implantation. In addition, equivalent hearing loss scores (EHL) were calculated to measure performance. Correlations between the mentioned parameters were studied.
*Results:* One or two pathogenic mutations were identified in 7 of the 14 examined patients. Similar to previous studies, it was demonstrated that implantation at an earlier age results in better performance than implantation at higher age. Cochlear implantation performed within the first two decades of life was beneficial to 13 of 14 (93%) of the USH1 patients. Finally, the EHL score and the G(C)BI score showed a significant correlation; the benefit of implantation increases with a decreasing EHL score.
*Conclusion:* Cochlear implantation in USH1 patients improves the audiological performance when patients are implanted at and earlier age and is beneficial according to the G(C)BI, when performed within the first two decades of life.

# Introduction

Usher syndrome is named after Charles Usher, a British ophthalmologist who described families with inherited types of retinitis pigmentosa in his Bowman lecture published in 1935.[1] The Usher syndromes are characterised by sensorineural hearing impairment, retinitis pigmentosa and in some cases vestibular dysfunction. Current prevalence estimates of Usher syndrome range from 3.5 to 6.2 per 100,000.[2] Usher syndrome accounts for about 3-6% of all congenital types of deafness, about 18% of patients with retinitis pigmentosa and over 50% of all patients with deaf-blindness.[3] Three clinical types can be distinguished on the basis of audiovestibular features.[4] Usher syndrome type I (USH1) shows congenital profound deafness, retinitis pigmentosa and vestibular areflexia. Usher syndrome type II is characterised by high-frequency sensorineural hearing impairment, retinitis pigmentosa and intact vestibular responses on testing. Usher syndrome type III shows progressive hearing impairment, which in a few years may lead to profound sensorineural hearing impairment, retinitis pigmentosa and variable vestibular function.

Usher syndrome is inherited in autosomal recessive fashion. Genetic subtypes have been identified for the three clinical types of Usher syndrome. At first it was believed that each clinical type of Usher syndrome was caused by mutations in one single gene. At present, however, already eleven loci and eight genes have been identified [Table 1].[5] For USH1, five of the currently estimated seven genes have been identified.

Table 1. Clinical and genetic subtypes of Usher syndrome, subsequent loci and gene

| Usher type | Genetic subtype | Localisation | Gene |
|---|---|---|---|
| Usher type I | USH1a | 14q32 | |
| | USH1b | 11q13.5 | MYO7A |
| | USH1c | 11p15.1 | USH1C |
| | USH1d | 10q22.1 | CDH23 |
| | USH1e | 21q21 | |
| | USH1f | 10q21-22 | PCDH15 |
| | USH1g | 17q24-25 | SANS |
| Usher type II | USH2a | 1q41 | USH2A |
| | USH2b | 3p23-24.2 | |
| | USH2c | 5q14.3-21.3 | VLGR1 |
| Usher type III | USH3 | 3q21-25 | USH3 |

Cochlear implantation (CI) is useful in USH1 patients because they are profoundly deaf. In addition, these patients will develop progressive visual impairment in the course of their life. This first manifests itself by night blindness, mainly presenting in

the first decade of life. In the following decades, constriction of the visual field causing tunnel vision as well as a decrease in visual acuity will develop. Eventually, retinitis pigmentosa leads to severe visual impairment or blindness. Being able to hear with a CI instead of being profoundly deaf seems even more important to these patients than to deaf patients with normal vision. This study evaluates the audiological performance after cochlear implantation in USH1 patients by measuring the equivalent hearing loss. The present study is unique because of the spread in patient characteristics, especially regarding the age of implantation. The benefit of cochlear implantation is evaluated by using the Glasgow Benefit Inventory (GBI) and the Glasgow Children's Benefit Inventory (GCBI).[6,7] In addition, an attempt is made to evaluate the genotype to see whether variations in genotype have any bearing on the performance and benefit of CI in these patients. A separate paper in this issue describes the results of questionnaires concerning quality of life in the same patients.[8]

# Materials and Methods

**Patients**

This study includes 14 patients with a clinical diagnosis of USH1. Only patients #1 and #2 are sibs, all other patients represent isolated cases. All patients underwent cochlear implantation because of profound deafness. The age of implantation varied from 3.5 to 30.4 years of age (median age: 10.0 years). Audiovestibular examinations confirmed profound deafness and vestibular areflexia in all patients prior to cochlear implantation. Retinitis pigmentosa was confirmed by ophthalmologic examinations, as previously described.[9] All patients agreed to participate in this study by written informed consent.

**Mutation analysis**

Blood samples were obtained from all patients, their sibs and parents to perform mutation analysis of USH1 genes. Genomic DNA was extracted according to Miller et al.[10] Mutation analysis of USH1 genes was performed by heteroduplex analysis, WAVE/DHPLC, Amplification-Refractory Mutation System (ARMS) assay, sequence analysis or by micro-array analysis (Usher chip, Asper Biotech, Tartu, Estonia). Not all exons of all USH1 genes were sequenced, however, an attempt was made to screen as many as possible (performed tests available on request). Some of the identified mutations (#3, #5, #8, #9, #10) have been reported previously.[11,12]

**Cochlear Implantation**

All but one patient (#10) received multichannel cochlear implants according to current standard procedures and they all had a follow-up for at least 24 months. Patient #10 primarily received a single-channel implant, however, in a second stage she was upgraded with a multi-channel implant. The 'equivalent hearing loss' (EHL) concept was used to evaluate the audiological performance of the CI users. Scores obtained in a battery of speech perception tests were reduced to one single measure, designated the EHL.[13] Speech perception test results from a cluster of children with severe and profound hearing impairment and well-fitted conventional hearing aids were used as a reference. Functional relationships between test scores and degree of hearing impairment were established with statistical procedures and used in reverse to convert the speech perception scores of subjects into an EHL value.[13] The EHL can vary between 50 and 130 dB hearing level (HL).[14] An EHL score of 90 dB HL reflects the speech recognition abilities of a person with 90 dB hearing loss, who is provided with well fitted hearing aids.

**Measurement of benefit by G(C)BI**

The Glasgow Benefit Inventory (GBI) and Glasgow Children's Benefit Inventory (GCBI) were used to assess patient benefit. These are validated means of comparing and quantifying changes in quality of life resulting from an otolaryngological procedure for both adults and children, including cochlear implantation.[6,7] The GBI questionnaires consist of, respectively, 18 and 24 questions sensitive to changes in general, social and physical health benefits. Patients were assisted by one of the authors (G.D.) while filling in the questionnaires. Possible scores for each question were based on a five-point Likert scale. The numerical data from the questionnaire were converted into a G(C)BI score. This is an index score of +100 to -100, representing best to worst outcome.

**Statistical analysis**

Linear regression analysis (EHL score plotted against months after implantation) was performed to evaluate individual improvement in speech perception over time. Patients were grouped by age of cochlear implantation in 3 different groups (1-9 years, 10-19 years and >20 years of age). Linear regression analyses of G(C)BI and the last-visit EHL scores plotted against age of implantation were performed to evaluate the benefit of cochlear implantation and the reduction in EHL score with increasing age of implantation. For the sake of comparison, EHL scores of congenital profoundly deaf CI users (n=26) with normal vision were used to evaluate differences in performance with USH1 users. Linear regression analysis was also performed on the G(C)BI score related to the EHL score data. Improvement in score was designated significant when a significant negative slope was determined ($P < 0.025$). Prism 3.03

software (GraphPad, San Diego, CA, USA) was used in all statistical analyses. Nonlinear regression analysis was only performed on an arbitrary basis to obtain satisfactory curve fits. The nonlinear functions used related to either one-exponential decay with offset, i.e. of the form Y=Span(exp(-kX))+Offset, with Y for score and X for age where Span, the rate constant k and Offset were fitted, or to one-exponential association with offset, i.e. of the form Y=Span(1-exp(-kX))+Offset, with Span=Offset=65 dB where only k was fitted.

# Results

**Mutation analysis**

Mutation analysis of the USH1 genes revealed mutations in two different USH1 genes in 7 of the 14 affected participants in this study. Six patients had pathogenic mutations in *MYO7A* causing USH1b and in one patient a *CDH23* mutation was responsible for USH1d. All patients with USH1b had two pathogenic mutations in *MYO7A* identified, however, only one *CDH23* mutation was identified in patient #10 [Table 2]. The second mutation in this patient has so far not been identified. Unfortunately, it was not yet possible to identify all responsible pathogenic mutations in the remaining 7 patients.

Table 2. Genetic characteristics of patients involved in present study

| Patient | Age of cochlear implantation (y) | Subtype | Gene involved | Mutation 1 | Mutation 2 |
|---|---|---|---|---|---|
| 1 | 9.14 | - | - | - | - |
| 2 | 6.94 | - | - | - | - |
| 3 | 11.76 | USH1b | MYO7A | R1240Q | R1240Q |
| 4 | 3.72 | USH1b | MYO7A | R1602Q&1170K | R1240Q |
| 5 | 10.89 | USH1b | MYO7A | R1602Q&1170K | R1240Q |
| 6 | 6.91 | USH1b | MYO7A | R1602Q&1170K | R1240Q |
| 7 | 13.46 | - | - | - | - |
| 8 | 22.78 | USH1b | MYO7A | R666X | R302H |
| 9 | 20.60 | USH1b | MYO7A | R1602Q&1170K | R212H |
| 10 | 20.05 | USH1d | CDH23 | IVS20+1G>A | - |
| 11 | 30.44 | - | - | - | - |
| 12 | 3.53 | - | - | - | - |
| 13 | 8.96 | - | - | - | - |
| 14 | 4.71 | - | - | - | - |

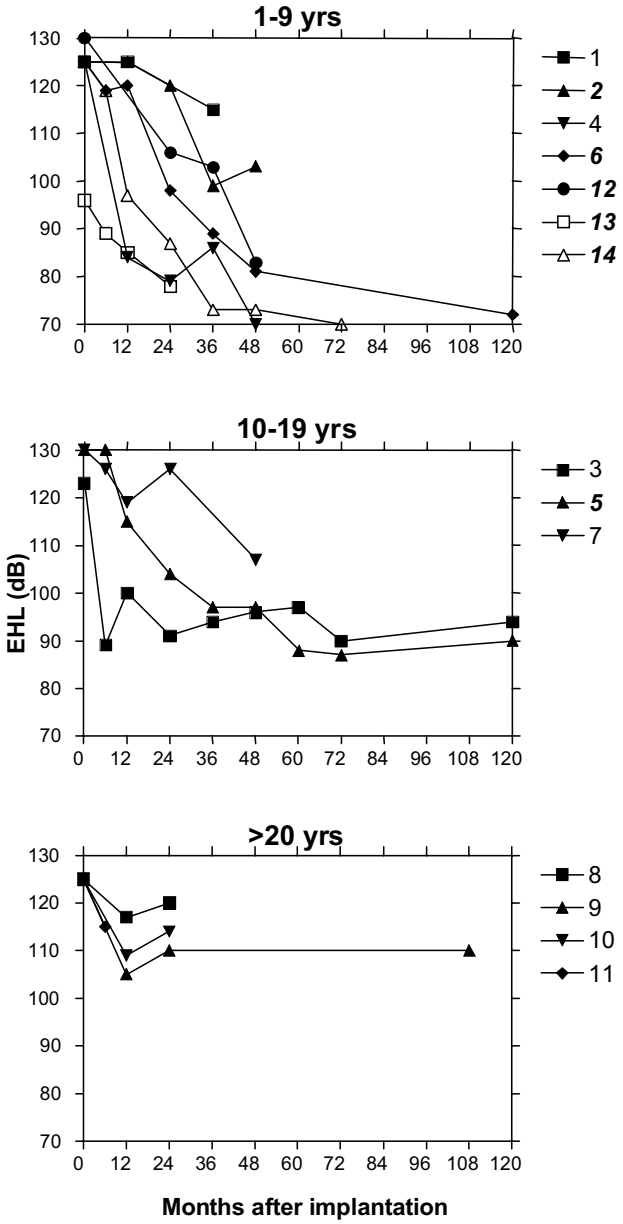


Figure 1. Development of the EHL score after cochlear implantation.
Bold and italic numbers relate to a significant reduction of EHL scores on linear regression analysis in the corresponding patients.

## Audiological performance after CI

Individual equivalent hearing loss values after cochlear implantation are shown in Figure 1. The improvement of performance is shown in relation to the number of months after implantation. In 5 out of 7 patients implanted below 10 years of age a significant reduction of EHL is seen. In this age group the mean last-visit EHL score was 84 dB HL. The youngest patient in the 10-19 years of age group also shows a significant reduction in EHL score, whereas the two remaining patients do not. This age group has a mean last-visit EHL score of 97 dB HL. Four patients older than 20

years of age at cochlear implantation also showed no significant improvement of hearing abilities on follow-up. The last age group has a mean last-visit EHL score of 115 dB HL after cochlear implantation.

## G(C)BI and CI

Figure 2 shows the G(C)BI scores plotted against age of implantation. Linear regression analysis (data not shown) demonstrated that cochlear implantation was significantly more beneficial at younger than at more advanced ages. The majority of the cochlear implant patients benefited from implantation performed within the first two decades of life. Only one patient (#10) reported a negative G(C)BI score. This patient was implanted twice and experienced facial nerve stimulation with her first single channel cochlear implantation. After re-implantation of a multi-channel cochlear implant she experienced uncomfortable auditory sensations and pain when turning on her cochlear implant device.

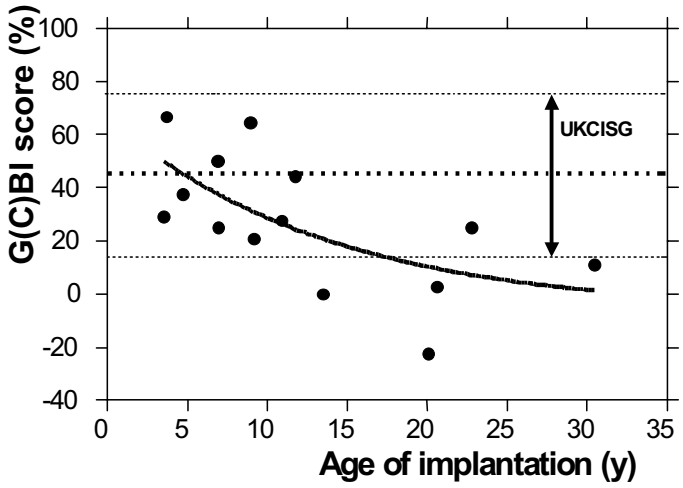


Figure 2. G(C)BI scores (in percentage) plotted against the age of implantation with an arbitrarily fitted curve (Methods). Dotted lines represent mean value and 2 sd lines for 227 adult postlingual cochlear implant patients.[26]

## EHL and CI

In Figure 3 the last-visit EHL score is plotted against the age of implantation. The black dots represent EHL scores of implanted USH1 patients and white dots the scores of congenital deaf patients with normal vision provided with a cochlear implant. Linear regression analysis (data not shown) indicated a significant reduction in EHL score when patients were implanted at an earlier age. From this plot it can be concluded that early implantation leads to better audiological performance than implantation at higher age.

**G(C)BI and EHL**

Finally, in figure 4, the results of the G(C)BI score are plotted against the EHL score results. With increasing EHL score the benefit of cochlear implantation deteriorated significantly. As the EHL score approaches 120 dB HL, the benefit score becomes 0% indicating that at this level of performance on speech recognition tests cochlear implantation can no longer be regarded beneficial.

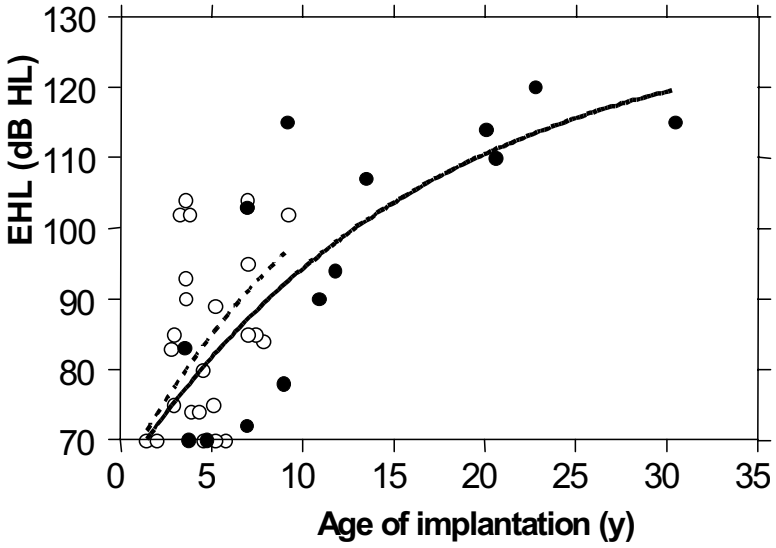


Figure 3. Last-visit EHL scores plotted against the age of implantation with arbitrarily fitted curves (Methods). Black dots and continuous line represent EHL score values and the fitted curve in USH1 patients (n=14). Circles and dotted line represent EHL scores and the fitted curve in congenitally profoundly deaf patients (n=26) with normal vision.

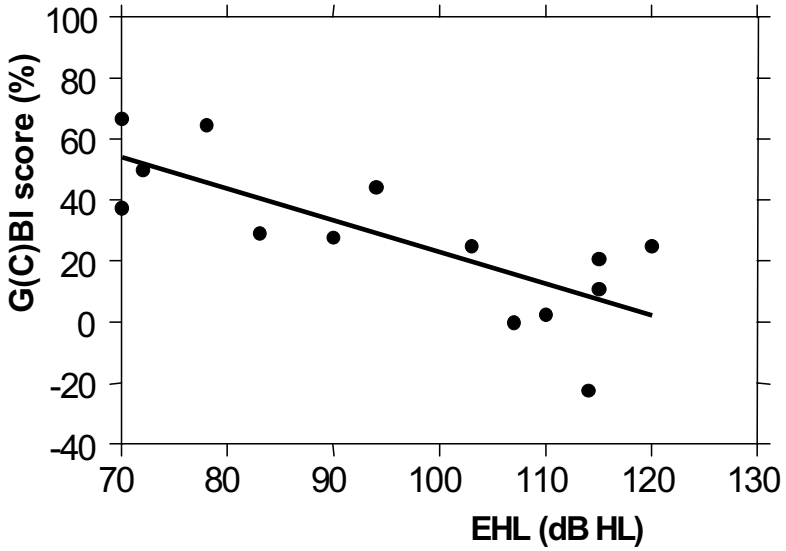


Figure 4. G(C)BI scores plotted against EHL scores. Black line depicts linear regression line.

# Discussion

This study analysed the audiological performance and the benefit of cochlear implantation in 14 USH1 patients. In a separate study, the quality of life in these patients was analysed.[8] The benefit of implantation was evaluated by measuring the G(C)BI score through a questionnaire. In addition, individual longitudinal analyses of EHL scores, measuring audiological performance, were performed. Mutation analysis of USH1 genes was performed to correlate the performance and benefit of CI in USH1 patients to the genetic subtype or to a specific type of mutation.

In 7 of the 14 patients in the present study, one or two pathogenic USH1 mutations were identified. In six of them, a mutation in the *MYO7A* gene was identified, causing USH1b and in one patient a mutation in *CDH23* causing USH1d was found. Astuto et al.[11] analysed the *MYO7A* gene in 151 USH1 families by linkage and mutation analysis. Pathogenic mutations were identified in 64 families (42,4%), which is fairly similar to the findings in the present study (6/14 = 42,9%). Although in some of these patients the Usher micro-array was used for mutation analysis, in only half of the patients one or two pathogenic mutations could be identified. This probably is related to the fact that many mutations are still unknown and not covered for by the Usher syndrome micro-array.

No clear relationship was found between the type of USH1 mutations and audiological performance after implantation. However, in only seven patients a genetic diagnosis could be made and the large variability in age of cochlear implantation does not permit any conclusions to be drawn. In patient #10, however, the benefit score was negative and this was also the only patient who had a *CDH23* mutation. Her problems are possibly related to insertion or to the used technique. It may, however, be interesting to find out whether other USH1d patients experience similar problems or not. Larger studies are certainly needed to establish genotype-phenotype correlations regarding audiological performance after cochlear implantation.

Several studies have evaluated cochlear implantation in Usher syndrome or deaf-blind patients.[15-18] This is, however, the first study that presents the audiological performance and the reported benefit of cochlear implantation in USH1. Loundon et al.[16] concluded that logopedic results are linked to the precocity of implantation and that an early diagnosis of Usher syndrome contributes to the optimisation of speech therapy. Two reports have speculated about the possibility that the performance of deaf-blind individuals after cochlear implantation may be superior to the performance of recipients with normal vision.[17,18] The report by El-Kashlan et al.[17] concluded this on the basis of the performance in three postlingually deafened cochlear implant

patients who became legally blind during life. The study by Saeed et al.[18] presented several patients who were congenitally blind and became deaf in the course of their life. These two studies differ from the present study that presents the results of cochlear implantation in congenitally deaf patients who develop progressive visual impairment in the course of their life. In this study, it has been shown that the speech recognition scores of USH1 patients behaved fairly similarly to those of profoundly deaf patients with normal vision.

The results of the present study confirm previous findings that early implantation leads to better performance than implantation at higher age.[19] This study also shows that cochlear implantation is beneficial to the majority of USH1 patients when performed within the first and second decades of life. Although there is some report on the benefit of CI in congenitally deaf adolescents and adults, the benefit seen in the patients in this study may possibly be related to the fact that these patients besides deafness also have retinitis pigmentosa. Although speculation, the visual impairment caused by retinitis pigmentosa possibly leads to less colonisation of the auditory cortex and therefore cochlear implantation may be longer beneficial, when compared to normal seeing deaf individuals. It needs, however, to be emphasised that Usher syndrome patients at young age hardly experience any visual problems and that they therefore probably use their visual cortex in a similar way like normal seeing subjects do.

The questions in the Glasgow Benefit Inventory were generated to measure a change in health status, where health status is defined as the general perception of well-being, as well as psychological, social and physical well-being.[6] It was mainly developed to assess patient benefit in otorhinolaryngological interventions, including for example tonsillectomy, insertion of ventilation tubes and bone-anchored hearing aids.[6,7] Recently, a similar health-related benefit measure for children was developed.[7] This study has shown that the G(C)BI questionnaires correlate clearly with the audiological performance in implanted Usher syndrome patients. When comparing the present GBI results to the reported benefit in cochlear implanted adults who became profound deaf postlingually [Figure 2],[20] it can be concluded that in 10 of the 14 patients the benefit of cochlear implantation is within two sd of the mean benefit (45) in the 227 patients studied by the UK Cochlear Implant Study Group. Obviously, reduction in EHL score is associated with an increasing benefit [Figure 4]. It therefore can be concluded that the G(C)BI questionnaires are reliable to asses patient benefit in cochlear implantation as well. In addition, it can be concluded that although cochlear implantation was performed in some patients more than 10 years ago, the benefit could still well be recorded and seems to justify the use of the questionnaires over a

longer period of time. Whether this is the case in other otorhinolaryngological procedures with less impact remains to be analysed.

# Conclusions

The present study described the results of cochlear implantation in a unique set of USH1 patients, who were implanted at various ages. It has shown that cochlear implantation in the majority of Usher syndrome type I patients is beneficial when performed within the first and even second decade of life. Similar to previous studies, it showed that early implantation leads to better audiological performance. No clear relationship could yet be established between the USH1 genotype and the phenotype regarding audiological performance or benefit of cochlear implantation.

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1.3

# Deaf children with a Cochlear Implant with developmental retardation or learning disabilities

Cochlear implantation in deaf children with developmental retardation or learning disabilities: quality of life and language comprehension.
(submitted)
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# Abstract

*Aim:* Comparison of quality of life and language comprehension after cochlear implantation in deaf children with multiple handicaps and children with deafness alone.

*Methods:* 20 implanted children with additional handicaps besides deafness (developmental retardation or learning disabilities) and a matched control group of 16 implanted children with deafness alone. The Glasgow Children's Benefit Inventory and the Paediatrics Quality of Life questionnaires and separate questions on the parents' worries regarding the cochlear implant (CI) were used. Furthermore, comparisons were made of language perception scores.

*Results:* The children with developmental retardation or learning disabilities tended to use their CI less than the control group. Only one multiple handicapped child was attending a regular primary school compared to nine control children. Questionnaire outcomes indicated that parents of children with developmental retardation or learning disabilities regarded the CI almost equally beneficial as parents of children with deafness alone. After three years of CI use, the language comprehension performance of children with additional handicaps was equal to that of the control children.

*Conclusion:* The results of the questionnaires did not reveal significant differences in quality of life and benefit of the CI between the two groups. Language comprehension of children with developmental retardation or learning disabilities was equal to that of control children with deafness alone. These results emphasize the importance of cochlear implantation in deaf children with developmental retardation or learning disabilities as additional handicaps.

# Introduction

Deaf children have been receiving cochlear implants from the Nijmegen/St. Michielsgestel CI team since 1989. Initially, only children who were expected to make optimal use of the CI were implanted[1]. These were totally deaf children without any other problems or disorders than their deafness. Children with an additional handicap (besides their deafness) were not usually considered as eligible for implantation; having low or no expectations regarding the possibilities of the child to develop spoken language. More recently, a number of children with developmental retardation have received a CI. In addition, some of the children who received a CI were later diagnosed with an additional disability.

Few reports have been published on the effect of cochlear implantation in children with an additional disability. Results suggest that children with multiple disabilities are able to benefit from a CI. After cochlear implantation, progress was observed in speech[2,3] and language development[4]. Nevertheless, in some studies, the children with learning disabilities as additional handicap made slower progress, achieved lower scores and showed wider variation in scores than the CI recipients without learning disabilities[5, 6]. In other studies, the performance of the children with an additional handicap was reported to be variable [7]. Studies investigating the effects of a CI on the development of children with an additional handicap, are rare and have shown that in most cases, cochlear implantation had a positive effect on communication and the parents expressed that they saw improvements in the quality of life of their children[8-12].

The present study mapped the advantages of cochlear implantation in children with an additional handicap besides deafness (either developmental retardation: non-verbal IQ<80, or severe learning disabilities). Results of language comprehension tests and quality of life questionnaires were compared to those obtained from a group of CI children with deafness alone. This study provides a unique insight in differences and challenges of CI children with and without an additional handicap.

Providing the parents with detailed information during the implantation process and rehabilitation phase is of great importance [3, 7, 13]. On the basis of the results of this study, we hope to compile further information for the parents of CI candidates and to help give them more realistic expectations about their child's future before they enter the implantation process.

# Materials and methods

## Patients

*Study group*

The study group comprised twenty deaf children who had an additional handicap besides deafness. A proportion of them (n=12) had developmental retardation (non-verbal IQ<80). Non-verbal IQs were between 52 and 79 prior to implantation according to standardised intelligence tests. The remaining eight children had been diagnosed with learning disabilities before implantation when they were enrolled in the implant selection procedure. Learning disabilities means that for example their reading, spelling or mathematical level was significantly lower than their intellectual ability. It had not been possible to catch up the difference in levels with the aid of remedial teaching [14].

Table 1. Study group data

| | Gender | Age at onset of deafness (yrs) | Onset of deafness | Aetiology | Handicap | Duration of deafness (yrs) | Age at implantation (yrs) | CI Nucleus type | Follow-up period (yrs) | IQ score |
|---|---|---|---|---|---|---|---|---|---|---|
| 1 | F | 0.4 | pre | meningitis | D | 2.2 | 2.7 | 24 | 3.3 | 70 |
| 2 | M | 0.0 | cong | hereditary | L | 2.5 | 2.5 | 24 | 4.8 | 80 |
| 3 | F | 0.0 | cong | unknown | D | 2.9 | 2.9 | 24 | 5.0 | 70 |
| 4 | F | 0.0 | cong | Waardenburg's syndrome | D | 2.7 | 2.7 | 24 | 3.8 | 57 |
| 5 | M | 0.0 | cong | Waardenburg's syndrome | D | 1.6 | 1.6 | 24 | 3.6 | 78 |
| 6 | M | 0.0 | cong | premature | L | 5.0 | 5.0 | 24 | 7.4 | 104 |
| 7 | F | 0.0 | cong | CHARGE association | D | 6.6 | 6.6 | 24 | 2.9 | <70 |
| 8 | F | 0.0 | cong | unknown | L | 6.7 | 6.7 | 24 | 5.2 | >80 |
| 9 | F | 0.0 | cong | cochlear immaturity | D | 2.5 | 2.5 | 24 | 3.8 | 55 |
| 10 | M | 0.0 | cong | rubella infection | L | 5.8 | 5.8 | 24 | 7.0 | 80 |
| 11 | M | 0.0 | cong | unknown | D | 6.9 | 6.9 | 24 | 7.1 | 70 |
| 12 | M | 0.0 | cong | unknown | D | 3.8 | 3.8 | 24 | 2.5 | 52 |
| 13 | F | 0.4 | pre | meningitis | L | 1.6 | 1.9 | 24 | 3.0 | >80 |
| 14 | F | 0.0 | cong | Usher's syndrome | D | 3.6 | 3.6 | 24 | 6.2 | 78 |
| 15 | F | 5.8 | post | meningitis | L | 0.8 | 6.6 | 24 | 4.5 | >80 |
| 16 | M | 0.4 | pre | meningitis | D | 0.3 | 0.7 | 24 | 3.3 | 70 |
| 17 | F | 0.0 | cong | rubella infection | D | 3.1 | 3.1 | 24 | 5.5 | 70 |
| 18 | F | 0.0 | cong | Johanson-Blizzard's syndrome | D | 5.8 | 5.8 | 24 | 2.5 | 78 |
| 19 | F | 3.8 | post | cytomegalovirus | L | 1.5 | 5.4 | 24 | 6.4 | 108 |
| 20 | M | 0.0 | cong | hereditary | L | 11.0 | 11.0 | 24 | 4.4 | >80 |
| Mean<br>Range | | 0.5<br>0 - 5.8 | | | | 4.0<br>0.3 - 11.0 | 4.5<br>0.7 - 11.0 | | 4.6<br>2.5 – 7.4 | <br>52-108 |

Gender: M = Male, F = Female; Onset of deafness: cong = congenital, pre = prelingual (age>3 years), post = postlingual; Handicap: D = developmental (non-verbal IQ<80), L = learning disabilities

All the children in the study group had been implanted with a Nucleus 24 (Cochlear Corporation, Englewood, Colorado, USA) between 1994 and 2002. Demographic characteristics are shown in Table 1. On average, the children were 4.5 years of age at time of implantation.

*Control group*

The database of the Nijmegen/St. Michielsgestel CI team was searched for control children who matched the study group as closely as possible regarding age at onset of deafness, age at implantation and duration of deafness. A total of 16 control children were included in the study. None of these children had, before implantation, been diagnosed with developmental retardation or learning disabilities by means of standardised psychological and IQ tests. 12 of the children had been implanted with a Nucleus 24, while the other four had a Nucleus 22 (Cochlear Corporation, Englewood, Colorado, USA). Implantation had taken place between 1993 and 2003. Demographic characteristics are shown in Table 2.

Table 2. Control group data

| | Gender | Age at onset of deafness (yrs) | Onset of deafness | Aetiology | Duration of deafness (yrs) | Age at implantation (yrs) | CI Nucleus type | Follow-up period (yrs) |
|---|---|---|---|---|---|---|---|---|
| 1 | M | 0.6 | pre | meningitis | 2.3 | 2.9 | 24 | 6.9 |
| 2 | F | 0.0 | cong | hereditary | 6.6 | 6.6 | 24 | 1.8 |
| 3 | F | 0.0 | cong | hystiocytosis | 5.5 | 5.5 | 24 | 5.3 |
| 4 | M | 0.4 | pre | meningitis | 1.8 | 2.2 | 24 | 6.8 |
| 5 | M | 0.0 | cong | hereditairy | 4.8 | 4.8 | 24 | 5.4 |
| 6 | F | 0.0 | cong | unknown | 10.9 | 10.9 | 24 | 3.5 |
| 7 | F | 0.0 | cong | unknown | 3.5 | 3.5 | 22 | 9.0 |
| 8 | M | 0.0 | cong | unknown | 2.7 | 2.7 | 24 | 7.9 |
| 9 | F | 5.4 | post | unknown | 1.4 | 6.8 | 24 | 3.0 |
| 10 | F | 0.0 | cong | unknown | 5.8 | 5.8 | 22 | 10.8 |
| 11 | F | 0.0 | cong | dysplasia | 2.5 | 2.5 | 24 | 5.0 |
| 13 | M | 0.0 | cong | hereditary | 1.5 | 1.5 | 24 | 2.2 |
| 14 | F | 0.0 | cong | infection | 2.8 | 2.8 | 22 | 9.0 |
| 15 | F | 0.7 | pre | meningitis | 0.4 | 1.0 | 24 | 4.6 |
| 17 | F | 0.0 | cong | Usher's syndrome | 6.9 | 6.9 | 22 | 11.9 |
| 18 | M | 0.0 | cong | cochlear immaturity | 3.8 | 3.8 | 24 | 6.3 |
| Mean<br>Range | | 0.4<br>0 – 5.4 | | | 4.0<br>0.4 – 10.9 | 4.4<br>1.0 – 10.9 | | 6.2<br>1.8 – 11.9 |

Gender: M = Male, F = Female; Onset of deafness: cong = congenital, pre = prelingual (age>3 years), post = postlingual. All children had standardised IQ scores >80 (categorical outcome)

It can be noted that the control group contains one child with the Usher syndrome. Although this is an additional disability, the child had at the time of investigation no visual deterioration or other complaints and had a normal IQ. This is why we decided to include the child into the control group, as it had no developmental retardation, learning disabilities or manifest other disabilities at time of this study.

**Questionnaires**

A number of data collection instruments were sent to the parents of all the children in this study: two questionnaires *(Glasgow Children's Benefit Inventory and Paediatrics Quality of Life)*, a list of general questions about the child and his/her CI and a short questionnaire developed by Nikolopoulos et al. about the expected future perspective and worries of the parents about their child[15]. The answers to some of the questions of this short questionnaire - on the theme of: Are you concerned about the development of your child's communication / listening / speech and language skills? - were used to produce descriptive statistics. Answer categories were "definitely", "to some extent", "slightly" and "not at all". The pre-implant questions and the questions on positive changes after cochlear implantation were left out, as this was not currently regarded relevant.

*Glasgow Children's Benefit Inventory (GCBI)*

To evaluate the effect of the CI, the parents of the study group children and control group children filled in the Glasgow Children's Benefit Inventory (GCBI). This instrument is designed to measure changes in quality of life after an otological intervention (e.g. cochlear implantation). The inventory comprises 24 questions on changes in various fields of social development and physical health. Each question can be answered on a 5-point Likert scale and the responses can be transposed into a GCBI score of between +100 (the most positive outcome) and -100 (the most negative outcome)[16]. To our knowledge, the GCBI has not been used in children with CI.

*Paediatrics Quality of Life Measurement (PedsQL)*

The PedsQL is a modular approach to measuring the general health-related quality of life of children and adolescents. It is a short and practical questionnaire, suitable for administration to healthy children and to children with acute or chronic health problems. There are four versions of the PedsQL that parents can complete depending on the age of their child: 2-4 years, 5 to 7 years, 8-12 years and 13 to 18 years.

The PedsQL comprises 23 questions on four domains (Physical functioning, Emotional functioning, Social functioning and Functioning at school) that can be answered on a 5-point Likert scale. Answers are recoded on a linear scale from 0-100; higher scores indicate better quality of life. The PedsQL has good reliability and validity and is commonly used world-wide[17, 18], though not yet in children with CI.

**Reynell Developmental Language Scales (RDLS)**

To evaluate the receptive language development of the participants in this study, the Reynell's test for language comprehension, 2nd revised edition, was used[19]. The communication method for all the children was oral, when needed supported by sign language. Results were obtained by administering the test during the evaluation sessions after implantation. Scores at 3 years after implantation were used in the present study. Reynell's test for language comprehension is a translation of the Reynell Developmental Language Scales[19,20]. Scores can be expressed as a standard score, percentile or age-equivalent. In this study, it was decided to express the speech comprehension score in the number of months a child was lagging behind his or her calendar age, i.e. speech comprehension test age in months was subtracted from the calendar age in months. We chose to express the results in this manner, as several children in our study were older than the oldest age at which the Reynell test quotient outcome has been standardised (6;3 years).

**Education**

The children's schools were classified into three groups according to the Dutch educational system (see Table 3). Group 1 comprised the regular primary schools and regular secondary schools; Group 2 comprised special primary and secondary schools for the deaf; Group 3 comprised special primary schools for deaf children with additional disabilities.

**Data analysis**

Data were collected in the spring of 2005 and most recent language test results were used in (correlation) analyses. SPSS version 12.0 and Prism version 3.03 were used to perform the analyses. Descriptive statistics were used to express demographics [Tables 1 and 2]. The answers filled in on the questionnaires were compared on group level. A non-parametric equivalent of the T test was used for a proportion of the analyses (Mann-Whitney U test). Analysis of the categorical data was conducted using Fisher's exact test in which an exact P value was calculated.

# Results

Table 3 shows the results of the comparison between the study group and the controls on the number of hours of CI use per day, their preferred method of communication and the types of school they were attending.

The analyses showed that the majority of children in the two groups were using their CI for more than 12 hours per day (study group 56% vs control group 83%). The whole of

the control group were using their CI for a minimum of 6 hours per day, while one child in the study group was hardly using his CI (6% <6 hours per day). Fisher's exact test showed that these differences were not statistically significant.

Table 3 also shows the preferred communication method of the children at home and at school. The most striking finding was that the children in the control group were all using verbal communication at school. At home, the vast majority of them were also using verbal communication (79%), while 21% were using (partly) sign language for support. In the study group, differences between school and home were less evident. A large proportion was using sign language to support verbal communication (school 47%, home 44%).

The distribution over the various types of school is again shown in Table 3. In the control group, more than half of the CI children (56%) were attending normal primary or secondary schools, while only one child in the study group (5%) was receiving this type of education. A large proportion of the children with an additional handicap was attending schools for the Deaf and one quarter was receiving education for children with an additional handicap. In addition, we evaluated the distribution of children with their specific additional handicap over the schools (developmental retardation or learning disabilities). In both groups, developmental retardation and learning disabilities, the majority follows education for the Deaf.

Table 3. Supplementary data on study and control group

| | | Study group | | Control group | |
|---|---|---|---|---|---|
| | | percentage | n | percentage | n |
| Duration of CI use per day (hours) | < 6 hours | 6% | 1 | - | - |
| | 6 – 12 hours | 38% | 6 | 17% | 2 |
| | > 12 hours | 56% | 9 | 83% | 10 |
| Communication method at home | NGT | 25% | 4 | 7% | 1 |
| | NMG | 44% | 7 | 14% | 2 |
| | Oral | 31% | 5 | 79% | 11 |
| Communication method at school | NGT | 26% | 4 | - | - |
| | NMG | 47% | 7 | - | - |
| | Oral | 27% | 4 | 100% | 3 |
| Type of school | BO/VO | 5% | 1 | 56% | 9 |
| | SBO/SVO | 70% | 14 | 44% | 7 |
| | SSBO | 25% | 5 | - | - |

Communication; NGT: Dutch sign language, NMG: Dutch supported by sign language
Type of school; BO/VO: regular primary or secondary school, SBO/SVO: special primary or secondary school, SSBO: special primary school for the deaf with additional handicap

**Questionnaires**

Data from questionnaires are shown in Table 4. Generally speaking, there was a trend towards the parents of the study group having more worries about the future of their child than the parents of the control group, based on the Nikolopoulos questionnaire. On all three subjects, more than half of the parents of the study group children mentioned that they were 'definitely' or 'to some extent' worried about the future, whereas the same percentages of parents of the control group children mentioned 'hardly' or 'not at all'. The study group scores showed more worries on all three subjects, but the differences were not statically significant. Even after combining the 'definitely' or 'to some extent' answers and the 'hardly' or 'not at all' answers and comparing those between both groups, results did not differ significantly.

Table 4. Questionnaire scores

| Questionnaire | | | Study group | | Control group | | |
|---|---|---|---|---|---|---|---|
| "Are you concerned about the development of your child's…" | | | Percentage | n | Percentage | n | Fisher exact* |
| | | Definitely | 31% | 6 | 15% | 2 | 0.42 |
| | | To some extent | 32% | 6 | 15% | 2 | 0.42 |
| | Communication | Slightly | 16% | 3 | 39% | 5 | 0.22 |
| | | Not at all | 21% | 4 | 31% | 4 | 0.68 |
| | | | | | | | |
| | | Definitely | 21% | 4 | 7% | 1 | 0.63 |
| | | To some extent | 48% | 9 | 31% | 4 | 0.47 |
| | Listening | Slightly | 5% | 1 | 31% | 4 | 0.13 |
| | | Not at all | 26% | 5 | 31% | 4 | 1.00 |
| | | | | | | | |
| | | Definitely | 26% | 5 | 15% | 2 | 0.67 |
| | | To some extent | 48% | 9 | 23% | 3 | 0.27 |
| | Speech & language skills | Slightly | - | - | 23% | 3 | 0.06 |
| | | Not at all | 26% | 5 | 39% | 5 | 0.70 |
| | | | | | | | |
| | | | Mean | Sd | Mean | Sd | Mann-Whitney U* |
| GCB1 | | | 36.2 | 18.66 | 29.9 | 21.74 | 0.66 |
| | | | | | | | |
| PedsQL | Physical | | 82.03 | 19.71 | 88.70 | 20.19 | 0.21 |
| | Emotional | | 72.47 | 12.40 | 71.92 | 18.77 | 0.75 |
| | Social | | 68.17 | 23.91 | 77.40 | 20.49 | 0.28 |
| | School | | 73.96 | 11.48 | 82.44 | 17.58 | 0.14 |

*Fisher exact test, $p < 0.05$ is considered statistically significant
*Mann-Whitney U exact test, $p < 0.05$ is considered statistically significant
GCBI: Glasgow Children's Benefit Inventory, PedsQL: Pediatrics Quality of Life
Mean; GCBI optimal score = 100, minimum score = -100, Sd: standard deviation

In the study group, the mean GCBI score (benefit of CI in comparison with situation before implantation) was 36.2 compared to 29.9 in the control group. The study group had a higher score than the control group, but further analyses showed that the difference was not statistically significant. Scores varied (seen by the standard deviation), particularly in the control group.
With the aid of the PedsQL parents can judge the quality of life of their child. Scores were calculated for the two groups on the various domains of the PedsQL to obtain a broader view of the quality of life of CI children. The scores of the study group were lower than those of the control group in three out of the four domains (physical, social and school), but the differences were not statistically significant.
Besides these group means and scores, we also analysed the differences in questionnaire results of the children with developmental retardation or learning disabilities. No significant differences were found in the answers to the different questionnaires. The scores on the questionnaires did not differ significantly between the children with the Nucleus 22 and those with the Nucleus 24.

**Reynell's test for language comprehension**
Language development, in number of months a child was lagging behind his or her calendar age, was measured at three years after implantation with Reynell's test. Figure 1 shows the data from the study group and the control group. After three years of implantation, the results of the study group were equal to those of the control group. There were no significant differences between the two groups.

# Discussion

The results of the children with developmental retardation or learning disabilities were compared to those of a control group of children with deafness alone ('normal' paediatric CI recipients). The results showed that there were minimal differences in the number of hours of CI use per day, the types of school, preferred communication method, quality of life measurements and language comprehension scores between the children with developmental retardation or learning disabilities and the control children. In interpreting all results it should be noted that the follow-up time of the two groups differed, nevertheless patients were matched for age at onset of deafness, age at implantation and duration of deafness. Besides those variables, more variables could be of influence, for example the open speech perception test results. Further research has to be carried out to investigate the impact of other variables.

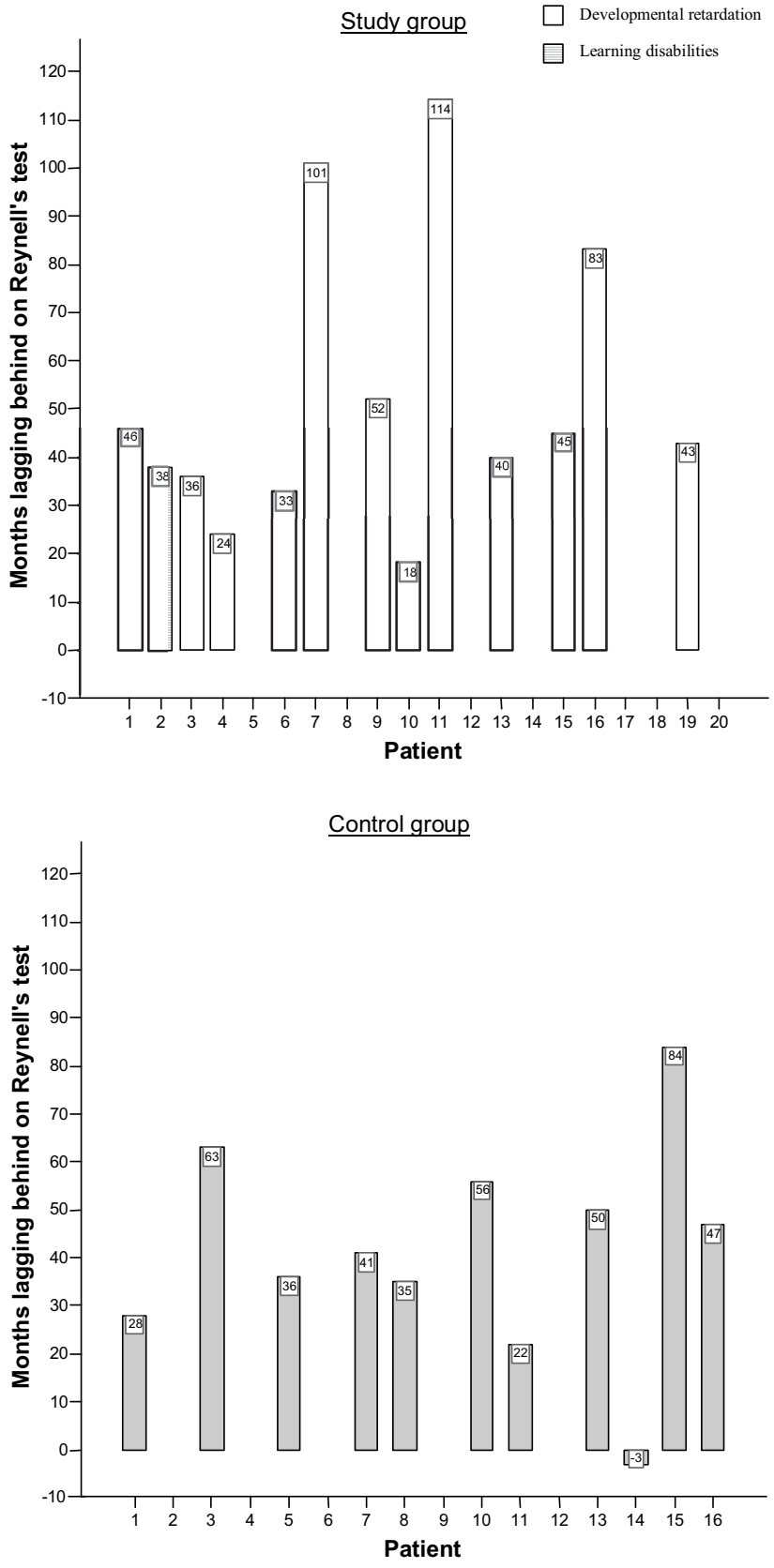


Figure 1. Number of months the children were lagging behind Reynell's norm.
The graphs show the scores on Reynell's test for language comprehension in the study group and the control group for all individual children, patient numbers correspond with the tables. Data obtained 3 years after implantation.

In terms of mean values, the children in the control group were using their CI for more hours per day than the children in the study group, but the difference was not statistically significant. This result was also mentioned in the study by Knutson et al.[21]

Studies on the preferred communication method in children with multiple disabilities showed that they were making less use of verbal communication than the children with deafness alone[5,11, 22]. Our results supported this finding. It was striking that although the majority of children in the study group were using sign language (in some cases to support spoken language), their language comprehension scores on tests that did not involve sign language were not significantly lower than those obtained from the control group.

Although the children in the study group did not have statistically significantly lower scores on the language comprehension tests or the quality of life questionnaires than the control group, a large proportion of them were attending 'special' schools. It is possible that some of the children with extra disabilities may have been placed in an inappropriate educational setting. In the study by Archbold et al., a larger proportion of the children with deafness alone were attending normal schools[23], though hereby it should be noted that the UK has a different educational system for people with a handicap than in the Netherlands.

Parents of the study group children were slightly more worried about communication, listening and speech development than the parents of the control group. Nevertheless, the study group's language comprehension development was almost equal to that of the CI children with deafness alone. It may be possible that the parents of the study group were more worried about language and speech development, because of the smaller amount of information existing on the development of multiple handicapped CI children. In theory, this could have left them with no frame of reference for the language comprehension development of their child. For CI children with deafness alone, more and univocal information is available. Providing full and correct information is an important factor to help parents form realistic ideas and expectations about the effect of cochlear implantation[13,24]. On the other hand, it should be noted that in the current study, no expressive spoken language skills were investigated. Parents may also be concerned about this aspect of their children's communication abilities, though this was not studied in the present report.

According to the answers that the parents of the study group and control group gave to the questions in the PedsQL, children living with a CI have a positive feeling about their QoL. In the study group, the score was even slightly higher than in the control group. The parents of the two groups of children in this study judged the quality of life of their children to be about the same as that reported by the parents of healthy children in the study by Varni et al.[25].

The present study had a retrospective design, so no data were available on quality of life prior to cochlear implantation. However, the results of the GCBI showed positive impact

of the CI on the QoL of their child. This is in agreement with the findings in other studies. Chmiel et al. and Vlahovic et al. asked parents to judge quality of life factors on a closed questionnaire[8,9]. Wiley et al. also invited parents to give their views on quality of life using open questions[10]. All three studies showed that the parents of children with extra disabilities regarded their child's quality of life to be better after cochlear implantation than before.

Language comprehension tests revealed that our group of children with developmental retardation or learning disabilities performed almost equally to the control group children. This was a remarkable finding, particularly because the children with extra disabilities had a lower mean non-verbal IQ than the children with deafness alone. Hypothetically, non-verbal IQ has little effect on the language comprehension process. It should be noted that all children are far behind compared to the normal hearing children though. However, studies on language development after cochlear implantation observed that children with several disabilities made slower progress and ultimately achieved poorer results than the children with deafness alone[5,6,22]. A poorer result generally meant that the children with multiple disabilities had more difficulty with the complex tasks (e.g. open set speech perception) than the children with deafness alone. In this study, we only assessed language comprehension and a slower development was not visible.

# Conclusion

The results of the questionnaires on cochlear implantation and quality of life, showed that the mean scores of the group of children with developmental retardation or learning disabilities were only slightly lower than those of the group of children with deafness alone. Language comprehension tests revealed that within three years after cochlear implantation, the language development of the children with developmental retardation or learning disabilities, seemed equal to that of the control group of children with deafness alone. Based on these data, cochlear implantation in children with developmental retardation or learning disabilities seems to show the same additional benefit in speech comprehension, quality of life as that in children with deafness alone.

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1.4

# Postlingually Deaf Adults with a Cochlear Implant

Cochlear implantation and quality of life in postlingually deaf adults: long-term follow-up.
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# Abstract

*Aim:* Investigate long-term quality of life (QoL) in postlingually deaf adults after entering the cochlear implantation programme.
*Methods:* Follow-up study from 1998 onwards in tertiary university medical centre. Long-term CI users, patients who have not received a CI and relatively short-term CI users were re-evaluated six years after initial data collection in 1998 by using three questionnaires (NCIQ, HUI3 and SF36) and speech perception tests.
*Results:* In general, the beneficial effect of CI remained stable during long-term follow-up, though scores on the questionnaires decreased slightly. Outcomes before and after cochlear implantation were significantly different. The group without a CI demonstrated slightly decreasing trends in outcomes. Long-term speech perception performance improved in time.
*Conclusions:* This is the first study to investigate long-term follow up of CI patients, in all aspects of QoL combined with speech perception performance, in comparison with postlingually deaf adults without CI.

# Introduction

Cochlear implantation has proved to be a successful and effective treatment for severely and profoundly deaf individuals[1]. A cochlear implant (CI) enhances speech perception and speech production in adults and improves hearing in all respects. Over the past 10 years, the general health status of patients, often referred to as health-related quality of life (HRQoL), has received increasing attention. Several studies have shown that a CI leads to substantial improvement in HRQoL, for example in the domains self-esteem and social functioning[2,3]. Few studies have been conducted on the long-term effects of cochlear implantation on HRQoL. Previous work has shown that initial improvements in psychological status after cochlear implantation tended to subside one and a half years after surgery[4]. At present, little is known about the long-term HRQoL of postlingually deaf adults after cochlear implantation. This study aimed to gain more insight into changes in HRQoL in adult CI recipients during long-term follow-up.
Background: In 1998, Hinderink et al. developed a disease-specific health-related QoL questionnaire for adults with a CI: the Nijmegen Cochlear Implant Questionnaire (NCIQ)[5]. To compare different aspects of HRQoL in a group of CI recipients, the authors administered the new NCIQ and two generic HRQoL instruments: the Health Utilities Index (HUI-3) and the Medical Outcome Study Short Form (SF36). Results showed that cochlear implantation led to improvements in HRQoL[6].
In the present study, six years after the initial assessment, the same three questionnaires were re-administered to the subjects. Data from the first study were compared to the present data (2004) and statistical analyses were performed. The following HRQoL aspects were addressed: long-term effects of a CI on HRQoL, changes in HRQoL during follow-up, differences in HRQoL between 1998 and 2004. In addition, the results of the HRQoL instruments were evaluated in relation with speech perception scores.

# Materials and methods

**Patients**

In 2000, Hinderink et al.[6] used three questionnaires to evaluate HRQoL in 47 adult patients who had received a CI at the Radboud University Medical Centre Nijmegen between 1989 and 1997. All the patients were postlingually deaf adults (without any functional residual hearing) who had received a multichannel implant at least one year before entering the study. Their HRQoL scores were compared to those obtained from a control group of 46 postlingually deaf patients who were on the waiting list for a CI at our institute.

In April 2004, we contacted all the patients and control subjects (n=93) and invited them to participate in the present study by filling in the same three questionnaires.
A total of 37 out of the initial 47 CI recipients agreed (group I). The remaining 10 patients were excluded, because three had died, three were lost to follow-up and for were unwilling to cooperate for various reasons.
In the initial control group of 46 patients on the waiting list for a CI, 29 patients had received an implant in the interval prior to the current investigation. Seven of them had to be excluded, because three had died, two were lost to follow-up and two were unwilling to take part in the study. Questionnaires were therefore sent to 22 patients who had received a CI between 1999 and 2004 (group III).
The remaining 17 patients had not received a CI. Ten of them agreed to fill in the questionnaires and seven patients had to be excluded, because two had died, two were lost to follow-up and three were unwilling to participate. These 10 patients (group II) had remained unimplanted for various reasons: fear, lack of motivation, unrealistic expectations, arbitrary residual hearing and a long period of deafness.
Our study population therefore comprised three groups of patients: group I (CI users in 1998 and in 2004), group II (non-implanted in 1998 and 2004) and group III (non-implanted in 1998, implanted between 1999 and 2004) [Figure 1]. Demographic characteristics are shown in Table 1.

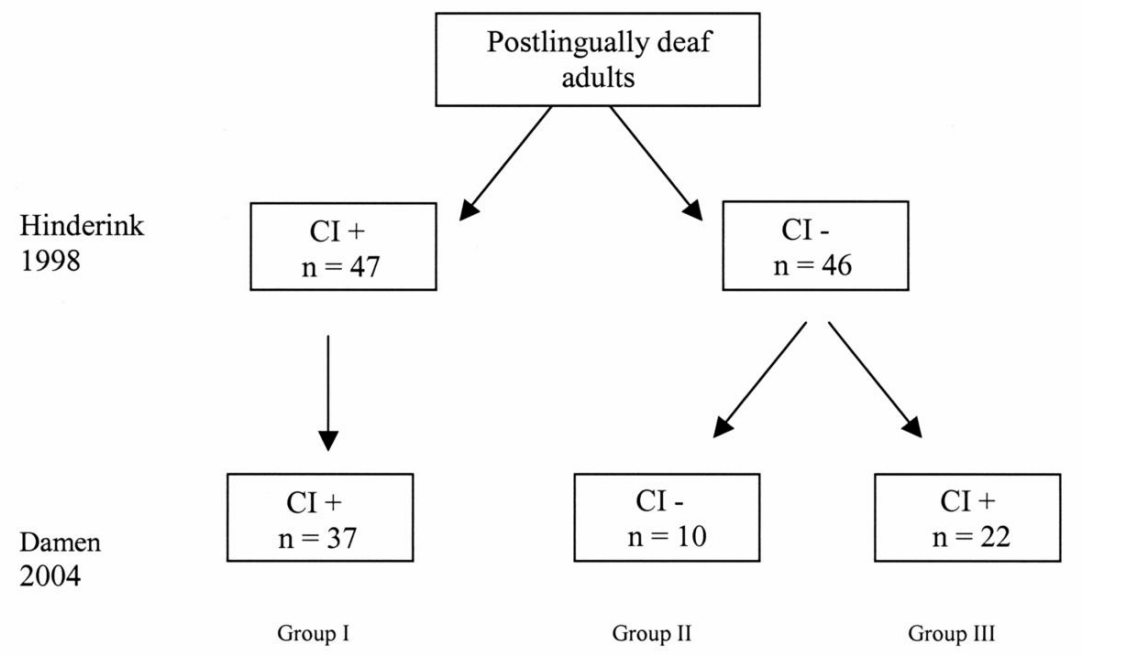


Figure 1. Demographic characteristics of the three groups who entered the CI programme

In group I, four different types of CI had been implanted: eight Clarion C1 devices (Advanced Bionics Corp., USA), two Laura devices (Antwerp Bionic Systems, Belgium), 21 Nucleus 22M and six Nucleus 24M (Cochlear Corp., Australia). In group III, two

different devices had been implanted: 11 Clarion C1 and 11 Nucleus 24M devices. In all the cases, surgery had been uneventful and no complications occurred in the direct post-operative period.

Data obtained in the previous study were compared to data obtained in the present study. The subjects were not provided with their previous answers. The study was approved of by the Institutional Review Board.

Table 1. Demographic characteristics of the three groups who entered the CI programme

| Characteristic | Group I (n=37) | Group II (n=10) | Group III (n=22*) |
|---|---|---|---|
| | '98: CI +<br>'04: CI + | '98: CI -<br>'04: CI - | '98: CI -<br>'04: CI + |
| Gender | | | |
| Male | 46% | 50% | 68% |
| Female | 54% | 50% | 32% |
| Paid Employment | | | |
| Yes | 41% | 60% | 27% |
| No | 60% | 40% | 73% |
| Education level | | | |
| Lower | 30% | 20% | 29% |
| Secondary | 46% | 60% | 52% |
| Higher | 24% | 20% | 19% |
| Living situation | | | |
| Alone | 19% | 30% | 23% |
| With others (parents, partner) | 81% | 60% | 77% |
| Care centre | - | 10 % | - |
| Age (mean yrs, sd) | 55.1 (16.0) | 50.5 (21.9) | 61.5 (13.1) |
| Age onset deafness (mean yrs, sd) | 30.8 (16.6) | 24.4 (30.6) | 47.6 (15.4) |
| Age CI (mean yrs, sd) | 45.2 (5.4) | - | 57.0 (13.4) |
| CI use (mean yrs, sd) | 9.9 (2.5) | - | 4.4 (1.2) |

* Education level, age CI and CI use n=21
CI +: patients with CI
CI -: patients without CI, on the waiting list (see text)

**Health Related Quality of Life Instruments**

Three instruments were used to obtain data on HRQoL: the Nijmegen Cochlear Implant Questionnaire (NCIQ), the Health Utility Index (HUI) and the Medical Outcome Study Short Form 36 (SF36).

The Nijmegen Cochlear Implant Questionnaire (NCIQ) is a disease-specific HRQoL instrument [5]. It addresses three general domains: physical (communication-related), social and psychological functioning. Each domain has one or more subdomains. The physical domain has subdomains: basic sound perception, advanced sound perception

and speech production. The social domain consists of 2 subdomains: activities and social functioning. The psychological functioning domain has one subdomain: self-esteem. Each subdomain contains 10 items. These items are formulated as statements with five answer categories that vary from 'never' to 'always' (55 statements) or from 'no' to 'good' (5 statements). If a statement does not apply to a patient, a sixth answer can be given: 'not applicable'. After computation, the scores on the subdomains range from 0 (very poor) to 100 (optimal).

The second questionnaire was the Health Utility Index (HUI) mark 3, a 15-item self-report health-status classification[7]. It provides a description of the health status of subjects and consists of eight subdomains: vision, hearing, speech, ambulation, dexterity, emotions, cognition and pain, with 5 or 6 response levels per item that vary from disabled to normal[8]. The HUI3 has proved to be a reliable, responsive and valid instrument in a wide variety of clinical studies[7]. Classifications on the 8 domains provide all the information necessary to calculate a single metric HRQoL summary score. These summary scores (also called utility scores) range from 0 to 1 on a generic scale, in which a value of 0 means as bad as being dead and a value of 1 means perfect health [9]. Health utility scores also proved useful in cost-utility analyses and related studies [7].

The third questionnaire was the Medical Outcome Study Short-Form 36 (SF36) that is based on a battery of health status instruments employed in the Medical Outcomes Study[10]. This questionnaire is a non-disease specific, generic HRQoL instrument. It contains 36 items that measure eight domains: physical functioning, role functioning due to physical health problems, role functioning due to emotional problems, bodily pain, vitality, social functioning, mental health and general health perceptions. The number of response choices per item range from two to six. Item scores on each dimension are coded, summed and transformed onto a scale from 0 to 100, in which higher scores indicate better self-perceived health. A physical component summary score (PCS) and a mental component summary score (MCS) can be computed using the standardised scoring system.

**Auditory test material**

Two Dutch standardised speech perception tests on compact disc were used: the NVA test (an open speech recognition test that consists of monosyllabic wordlists)[11] and two subtests from the Antwerp-Nijmegen test battery, AN-test, to assess suprasegmental identification (a closed set spondee identification test and a closed set number of syllables test)[12]. All auditory tests were presented at 70 dBSPL (conversation level) in a double-walled soundproof room with low reverberation. At least two lists consisting of 11 monosyllables each were presented for the NVA-tests; phoneme scores were obtained.

**Statistical Analysis**

Long-term effects of CI use were determined by comparing the results obtained from group I in 1998 and 2004. For baseline purposes, i.e. to study differences over time regardless of cochlear implantation, we reviewed the two sets of results obtained from the subjects who had not received a CI (group II). By comparing pre- and post CI results of 1998 (group I) and 2004 (group III), the potential change in benefit of CI could be established in two sequential groups of patients.

Besides the diverse domain scores, mean scores for the NCIQ and SF36 were determined as well as the utility score of the HUI3.

Scores on the three instruments were declared as missing values if nothing was filled in or if ambiguous information was provided. The maximum number of incomplete answers for a specific subdomain of the NCIQ was arbitrary set at three items per domain; above this number the domain was not scored. As the distribution of scores on the majority of separate domains appeared to be skewed, nonparametric tests (Wilcoxon's Signed Rank exact tests) were used to analyse whether the scores obtained from the three groups were statistically significantly different. The same statistics were used to analyze audiological data. Pearson and Spearman correlation analyses were computed to identify statistical relation between diverse domains and implant variables. Analyses were done by using SPSS software package 12.0.

# Results

**Health Related Quality of Life measurements**

Table 2 presents the mean NCIQ outcomes. In group I, there was very little change over time. Statistically significant deterioration was only seen in the domain 'social interactions'; 32 out of the 37 patients had poorer scores in the 2004 evaluation.

In group II, scores on all but one of the NCIQ domains decreased over time (i.e. showed deterioration in HRQoL). Only the domain 'speech production' decreased significantly.

The potential change in benefit of cochlear implantation was established by comparing pre- and post-CI results obtained from two sequential groups of patients in 1998 (group I) and 2004 (group III). In 1998, the NCIQ showed significant improvement in all the domains after implantation. The same effect, with comparable size, was seen in 2004. Changes in the mean scores of group I over time on the NCIQ showed that long-term HRQoL had decreased slightly during the further six years of CI use, although not statistically significantly [Figure 2a].

Nevertheless the beneficial effect of the CI was still clearly apparent compared to the preimplantation scores. Mean scores in group I and group II obtained in 1998 and 2004 had the same slope, which demonstrated minor deterioration over time. The beneficial

effect of cochlear implantation was visible in the parallel increasing slopes in group I and group III after implantation, which indicated similar significant benefit of cochlear implantation in the present study and the former study by Hinderink et al.

Table 2. NCIQ results

| NCIQ | Group I (n=37) | | | Group II (n=10) | | Group III (n=22) | |
|---|---|---|---|---|---|---|---|
| | Pre CI | '98 CI+ | '04 CI+ | '98 CI- | '04 CI- | '98 CI- | '04 CI+ |
| Sound perception basic | 3.2 (6.0) | 65.5 (24.2) | 60.7 (25.1) | 17.3 (15.2) | 15.0 (14.7) | 10.0 (13.8) | 63.5 ** (23.2) |
| Sound perception advanced | 14.6 (11.7) | 55.2 (19.3) | 54.4 (20.0) | 27.1 (9.1) | 22.5 (10.9) | 14.6 (10.8) | 51.7 ** (21.1) |
| Speech production | 60.5 (20.7) | 83.3 (17.6) | 83.3 (17.7) | 56.4 (16.0) | 41.6 ** (17.0) | 68.8 (18.3) | 80.3 * (17.7) |
| Self-esteem | 43.0 (20.1) | 67.7 (17.2) | 66.8 (19.2) | 52.5 (19.7) | 44.9 (21.4) | 43.6 (20.7) | 69.4 ** (13.0) |
| Activity | 50.0 (21.9) | 75.1 (16.0) | 73.6 (19.6) | 44.2 (16.3) | 48.7 (18.4) | 45.0 (23.9) | 71.7 ** (18.2) |
| Social interactions | 53.7 (18.0) | 74.5 (14.1) | 63.7 ** (14.8) | 51.9 (10.3) | 44.9 (12.8) | 42.0 (21.4) | 60.6 ** (14.2) |

Means and standard deviations (between brackets) on the domains of the NCIQ. Significant changes after 6 years are indicated with an asterisk (Wilcoxon's Signed Rank) * $p < 0.05$; ** $p < 0.01$

Most utility scores from the HUI 3 [Table 3] did not alter significantly in group I in the long-term, although the 'pain' domain seemed to show a slight significant decrease. There was a general trend towards slight deterioration in HRQoL, but the amount of change rarely reached significance. The HUI 3 did not detect any significant changes in group II over time. Comparison of pre- and post-implantation data showed similar effects in 1998 and 2004: the same 3 domains ('hearing', 'emotions' and the total utility score) improved significantly. Although the HUI utility score [Figure 2b] decreased over time, the final results were still significantly better than those obtained before cochlear implantation (pre-CI vs long-term post-CI 2004: $p > 0.01$). In Figure 2b, similar slopes are visible in group I and group II between 1998 and 2004. The increase in HUI utility score after cochlear implantation in group III (2004) was also significant, but to a smaller extent than in 1998 [Table 3].

Table 4 shows that the SF36 detected greater decreases in HRQoL in the long-term than the NCIQ and the HUI3. In group I, all eight domains deteriorated over the years, five of which statistically significantly [Table 4].

Table 3. HUI3 results

| HUI3 | | Group I (n=37) | | Group II (n=10) | | Group III (n=22) | |
|---|---|---|---|---|---|---|---|
| | Pre CI | '98 CI+ | '98 CI- | '98 CI- | '98 CI- | '98 CI- | '04 CI+ |
| Vision | | 93.9 (16.0) | 93.7 (16.0) | 97.0 (2.6) | 95.7 (1.8) | 95.7 (1.8) | 93.7 (1.8) |
| Hearing | | 56.6 (22.1) | 55.1 (24.9) | 19.3 (31.1) | 13.8 (20.5) | 13.8 (20.5) | 59.2 ** (23.5) |
| Speech | | 95.3 (10.4) | 94.2 (12.9) | 75.5 (17.7) | 90.1 (17.4) | 90.1 (17.4) | 94.4 (9.8) |
| Ambulation | | 98.7 (6.0) | 96.8 (8.6) | 98.3 (5.4) | 96.3 (14.0) | 96.3 (14.0) | 92.6 (16.4) |
| Dexterity | | 98.2 (9.2) | 97.9 (9.4) | 98.8 (4.8) | 96.4 (17.1) | 96.4 (17.1) | 98.9 (3.5) |
| Emotion | | 94.9 (5.8) | 91.5 (15.7) | 96.4 (4.7) | 90.0 (14.9) | 90.0 (14.9) | 97.6 * (4.1) |
| Cognition | | 96.9 (12.1) | 95.8 (8.9) | 84.6 (20.9) | 96.3 (9.1) | 96.3 (9.1) | 84.2 (26.7) |
| Pain | | 93.1 (11.4) | 87.8 * (20.2) | 91.6 (15.8) | 91.9 (9.9) | 91.9 (9.9) | 85.1 (24.8) |
| HUI 3 utility | 0.32 (0.15) | 0.64 (0.20) | 0.58 (0.24) | 0.37 (0.22) | 0.31 (0.18) | 0.38 (0.21) | 0.53 * (0.24) |

Means and standard deviations (between brackets) on the domains of the HUI3. Significant changes after 6 years are indicated with an asterisk (Wilcoxon's Signed Rank) * $p < 0.05$; ** $p < 0.01$

Table 4. SF36 results

| SF36 | Group I (n=37) | | | Group II (n=10) | | Group III (n=22) | |
|---|---|---|---|---|---|---|---|
| | Pre CI | '98 CI+ | '04 CI+ | '98 CI- | '04 CI- | '98 CI- | '04 CI+ |
| Physical functioning | 88.4 (18.5) | 86.6 (20.4) | 80.5 * (22.5) | 86.0 (17.5) | 74.6 * (25.0) | 76.0 (30.0) | 68.6 (27.2) |
| Social functioning | 58.1 (28.0) | 85.1 (20.0) | 77.1 * (22.3) | 77.5 (22.7) | 73.8 (23.2) | 76.7 (26.5) | 79.0 (26.8) |
| Role functioning (physical) | 57.4 (40.8) | 83.1 (33.4) | 70.6 (38.7) | 80.0 (30.7) | 52.5 (44.8) | 61.4 (40.6) | 58.3 (43.5) |
| Role functioning (emotional) | 62.2 (39.4) | 86.5 (30.9) | 71.6 (39.5) | 86.7 (28.1) | 50.0 (42.3) | 68.3 (37.2) | 81.8 (36.5) |
| Pain | 88.4 (17.6) | 84.5 (17.1) | 76.9 (28.1) | 83.5 (21.1) | 85.0 (20.7) | 80.8 (21.5) | 80.3 (25.7) |
| Mental health | 63.1 (18.8) | 78.2 (16.7) | 70.9 * (21.4) | 74.8 (17.7) | 66.0 (19.4) | 74.4 (16.1) | 82.7 * (16.1) |
| Vitality | 68.7 (18.0) | 72.4 (17.8) | 62.9 ** (22.8) | 66.0 (19.8) | 71.7 (30.0) | 68.9 (19.8) | 67.5 (25.6) |
| General health perception | - | 74.9 (18.9) | 61.4 ** (25.2) | 70.0 (22.1) | 60.9 (26.5) | 66.8 (21.4) | 61.6 (21.5) |
| Physical Summary Score | - | 51.6 (8.7) | 48.3 ** (10.4) | 50.7 (7.6) | 48.1 (12.4) | 47.2 (10.1) | 43.0 (11.9) |
| Mental Summary Score | - | 52.4 (9.8) | 47.9 ** (12.9) | 50.1 (8.8) | 45.3 (11.6) | 49.5 (8.7) | 54.5 * (10.1) |

Means and standard deviations (between brackets) on the domains of the SF36. Significant changes after 6 years are indicated with an asterisk (Wilcoxon's Signed Rank) * $p < 0.05$; ** $p < 0.01$

The physical and mental summary scores also decreased statistically significantly. Time effects in group II were ambiguous on the SF36; only the scores on the 'physical functioning' domain decreased significantly.

The SF36 benefit scores for cochlear implantation measured in 2004 (group III) were compared to the results of the previous study (1998). Whereas in 1998 all but two domains ('pain' and 'vitality') improved significantly, in 2004 only 'mental health' and the mental summary score increased significantly. The 'general health' domain had not been measured before implantation in 1998. Results on the eight domains were combined into one mean SF36 score [Figure 2c]. Deterioration in group I ran parallel with that in group II. This decrease was significant in group I ($p < 0.01$) in which 25 out of the 37 patients had a poorer score in 2004, but not significant in group II ($p = 0.08$). Group III showed a small, non-significant increase after cochlear implantation that was less apparent than the increase in mean SF36 score in group I after cochlear implantation in 1998. Generally, the long-term benefit of a CI on HRQoL remained stable, but the effects were less clear on the generic SF36 than on the NCIQ and the HUI.

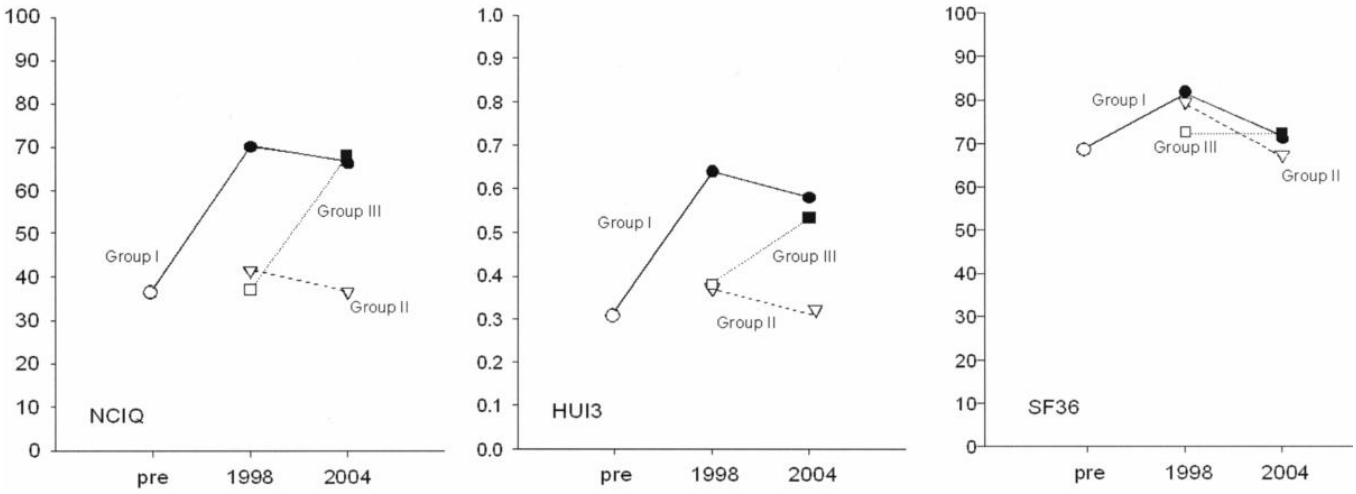


Figure 2. Mean questionnaire scores obtained at each measurement session
2a. NCIQ scores ; 2b. HUI3 scores ; 2c. SF36 scores; group I (CI users in 1998 and 2004), group II (no CI in 1998 or 2004) and group III (no CI in 1998; CI in 2004)
open symbol: no CI, filled symbol: with CI

**Follow-up of speech perception after implantation (Group I)**

Follow-up speech perception scores were not obtained from nine patients for different reasons (emigration, unwilling to cooperate, illness).

The two subtests of the Antwerp-Nijmegen test battery showed relatively high scores. During long-term follow-up, the mean AN spondee percentage in group I hardly changed: 82% in 1998 compared to 88% in 2004. Similar scores were obtained on the syllable identification test (80% in 1998; 82% in 2004). Figure 3 shows that the patients had lower scores on the speech recognition NVA test than on the AN test.

Significant improvements in the NVA word scores and phoneme scores were seen over time in group I (tested by Wilcoxon's Signed Rank exact tests for dependent variables): from 24% in 1998 to 36% in 2004 and from 45% in 1998 to 60% in 2004, respectively [Figure 3].

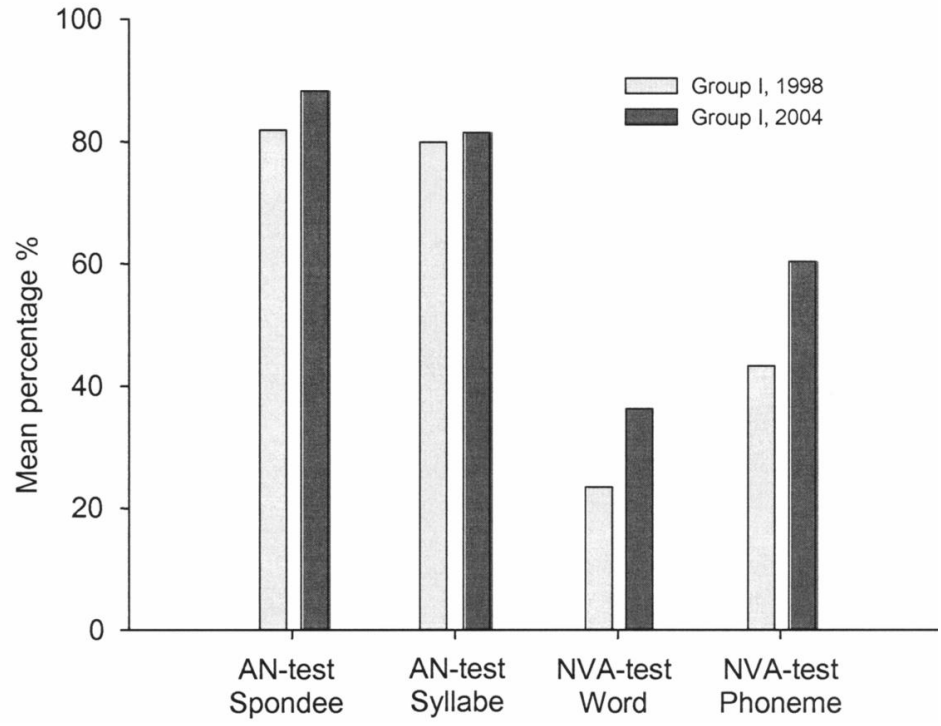


Figure 3. Mean speech perception test results in group I (CI users in 1998 and 2004) on the AN spondee and syllable tests and on the NVA word and phoneme tests are shown for .

# Discussion

In the current long-term follow-up study, cochlear implantation and multiple aspects of specific and generic HRQoL were combined with utility scores. To our knowledge, this is the first publication on this issue. Our results showed that the beneficial effects of a CI on HRQoL were stable in the long-term. Especially the hearing-related HRQoL scores (NCIQ) increased after cochlear implantation and this beneficial effect remained clearly visible. Nevertheless a small but non significant trend towards deterioration was observed over time. A similar trend was detected in group II, which suggests that the decline was not necessarily connected with cochlear implantation. The deterioration could for example be a natural effect of aging. Group III showed beneficial effects of cochlear implantation similar to those found in 1998. Others have described the positive effect of a CI on QoL, although not many researchers used the NCIQ[1,2]. Recently, Cohen et al[13] used the NCIQ (but no generic instruments) to compare HRQoL between 26 CI users and 30 hearing aid (HA) users, all with postlingual deafness. The results of their analyses supported the value of this

instrument in the HA group and CI group. They found beneficial effects of a CI in all the subdomains[13], which were equivalent to our results. As these authors had not used generic HRQoL instruments, it was not possible to compare the effect of a CI on hearing-specific and generic HRQoL.

In group I, the Health Utility Index mark 3 did not show any significant changes in the long-term. All the domains and the utility score reflected a small but non-significant decrease in HRQoL. Utility scores in group I and group II showed comparable deterioration, which was also apparent in the mean NCIQ scores. These findings may confirm our hypothesis on the natural effects of aging. Similar deterioration in HUI scores over time was described by the UK group in cochlear implant patients[14]. In group III, scores on four of the domains increased after implantation (2 of which statistically significantly), whereas an equal number of domain scores decreased. The utility score of group III improved significantly after they had received a CI. Only one other study reported the use of the HUI mark 3 to determine changes in HRQoL after a CI, but this was in 22 prelingually deaf children[15]. The smaller level of improvement in our study than in the report by Cheng et al. could be due to the fact that all the patients in our study group were postlingually deaf adults.

In our long-term evaluation, a significant decrease was detected in five out of the eight SF36 domains. Group II results were ambiguous and did not show any type of trend. In general, these unclear findings seem to confirm the variable SF36 results after cochlear implantation mentioned by Krabbe et al.[6]. Group III results showed only one significant increase (in the domain of mental health), whereas in 1998, five domains had shown a strong increase after implantation. A lack of sensitivity of the SF36 to detect changes in HRQoL after cochlear implantation was also seen in an earlier study on 27 postlingually deaf adults[16] and in a number of Usher type I subjects who had received a CI[17]. As far as we know, these are the only studies that used the SF36 in CI patients, although the instrument has been used before in hearing impaired adults. According to the literature, the SF36 lacks the necessary sensitivity to detect clinically meaningful improvements in patients with hearing impairment[18]. Therefore, on the basis of the previous and present observations of low sensitivity, we believe that the SF36 should not form the first choice of generic QoL questionnaire to evaluate hearing impaired patients.

It has been reported that HRQoL and utility scores are based on two main factors: the dimensions used to describe a person's health state and the technique used to assign a value to the health state descriptions elicited by each of the HRQoL questionnaires[19]. The Health Utilities Index focuses on a person's capability to undertake certain tasks, such as hearing and speech (production), but it does not consider the implications of any impairments. In contrast, the SF-36 focuses on performance rather than the underlying level of impairment. This could be an additional explanation for the difference in outcomes and trends between the HUI and the SF36.

Long-term effects of a CI on speech recognition tests showed progressive increases in suprasegmental scores and segmental speech perception tests. In group I patients, the initial improvement in speech recognition after cochlear implantation increased significantly over the subsequent six years. This is an impressive finding, because other authors have demonstrated no increase in speech perception outcomes[15,20] or only improvement over a shorter follow-up period[21].

# Conclusions

The initial benefit of cochlear implantation in a group of postlingually deaf adults was found to be stable in the long-term, although the HRQoL scores decreased slightly over time. This decreasing trend was observed with the hearing-specific NCIQ and the utility-based HUI3. On the whole, the benefit of a CI was maintained, which is an important finding with respect to cost-benefit analyses and very encouraging for policy makers and health care providers. The SF36 was also used, but it showed ambiguous results. We feel that this instrument should not be the first choice of generic QoL questionnaire in further CI research projects. The control group (who had not received a CI) showed the same slight decrease in QoL as the long-term CI users. The currently reported beneficial effects of cochlear implantation on quality of life were equal to the former results from 1998. Speech perception scores still continued to increase over time, even after long-term CI use.

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# 2 Assessment of specific instruments

# 2.1

# The assessment of mainstream performance

Classroom performance and language development of CI students placed in mainstream elementary school.
Otology & Neurotology 2007;28(4):463-72.

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E.A.M. Mylanus

# Abstract

*Aim:* Investigation of the relation between classroom performance and language development of Cochlear Implant (CI) students in mainstream education. Structural analyses of Assessment of Mainstream Performance (AMP) and Screening Instrument For Targeting Educational Risk (SIFTER) instruments.

*Methods:* 26 CI children in elementary school with congenital or prelingual deafness were included. At the time of this study, mean period of multichannel CI use was 5.3 years and children's ages ranged from 6.5 to 12.8 years. Main outcome measures were AMP and SIFTER instruments that measured classroom performance. Language development was measured by means of Reynell and Schlichting tests.

*Results:* AMP and SIFTER domains showed good reliability (Cronbach's alpha > 0.6), but factor analyses only showed the expected instrument structure in the AMP. In both questionnaires and within all domains, individual variability is detected. Spearman's correlation analyses showed the probable explanation of individual questionnaire variability by language test results (p-value mostly <0.01). The AMP and SIFTER instruments showed a predictive capacity for language development, based upon general linear model univariate and linear regression analyses.

*Conclusions:* Individual classroom performance, measured by AMP and SIFTER questionnaires, of CI children in mainstream education varies. Correlation analyses showed strong significant relation between questionnaire results (classroom performance) and both expressive and receptive language test results (Schlichting and Reynell tests). Structural questionnaire analyses of the AMP and SIFTER demonstrated good reliability. The predictive value of the AMP can monitor the actual linguistic functioning of the child.

# Introduction

The present cochlear implant technology has made mainstreaming a more reachable academic level for a considerable number of profoundly deaf children. As more children are implanted at an earlier age and obtain good results on speech recognition and language acquisition tests with their CI sooner, it is likely that more children may be ready to enter mainstream education at the same age as their hearing peers. Spencer et al. have shown that cochlear implant users attain comparable levels of academic achievement measures[1]. This is in concordance with previous findings from a provisional report by Chute et al. and from an earlier study conducted at the Radboud University Medical Centre Nijmegen[2;3]. Nevertheless, paediatric CI users seem to attain diverse levels of academic achievement in school. These differences observed between CI-users needed further evaluation. Several articles described factors influencing the outcome in speech, language and reading competence of an implanted child and with probable impact on (mainstream) school performance. These factors diverge from nonverbal intelligence, family size and socio-economic status[4], family concerns and programming difficulties[5] or oral education programmes[6] to motor and cognitive delay[7].

In the earlier study[3], it was examined whether differences could be seen in the classroom performance and communicative skills of CI students and their normal hearing peers. Teachers of CI students in mainstream educational settings were asked to complete the Assessment of Mainstream Performance (AMP) and the Screening Instrument for Targeting Educational Risk (SIFTER) for the implanted child as well as for a normal hearing classmate. Results of the questionnaires were analysed for correlation with audiologic test results. It was concluded that even though CI children in mainstream education face special challenges, they seemed to perform quite well, except for the instrument domain regarding communication. This was an important finding as communication is regarded to be imperative for the social and educational development of children in mainstream education. There was a vast inter-individual variability in outcomes between the CI students. No definite explanatory factors for this variance were found at that time, even though age at implantation and duration of deafness did show correlation with the questionnaire results.

As language development was not investigated so far in relation to the AMP and SIFTER, it was decided that further research was necessary to determine whether the mentioned variability in AMP and SIFTER results could be related to differences in language development. The language development was tested by means of standardised language production tests, performed yearly after CI (longitudinal follow up till 5 years post CI). Before performing the mentioned analyses, the instruments' structures were examined (reliability and factor analyses) as both the two earlier reports emphasized the need of further investigation of the AMP questionnaire[2;3].

# Materials and Methods

**Subjects**

Over 250 children have been implanted at the Radboud University Medical Centre Nijmegen since 1989, in close collaboration with Viataal, Sint Michielsgestel. In a former study, data were gathered of thirty-two CI children currently in mainstream education. In the current study twenty-six successive CI children in elementary school with congenital or prelingual deafness (≤ 3 years of age) were included. Those twenty-six children were, besides children in kindergarten, also included in the former study. The decision for school placement was made individually per child in close collaboration with the rehabilitation specialists (psychologists and school representatives) of the UMC Nijmegen/ Viataal Cochlear Implant team. All 26 children received the implant between 1995 and 2003 (mean age at implantation 4.2 years, range 1.4 to 9.7 years). Twenty-three children received a Nucleus 24-channel and three children a Nucleus 22-channel cochlear implant (Cochlear Corporate, Englewood, Australia). In all patients surgery was uneventful and a full function of the electrode array was ascertained. Mean duration of deafness is 3.9 years (0.5 year to 9.7 years), 18 children were congenitally deaf and 8 children became deaf prelingually. At the time of this study the mean period of cochlear implant use was 5.3 years (varying from 1.0 year to 9.1 years) and the children's ages ranged from 6.5 to 12.8 years (mean 9.5 years). Demographic characteristics for each child individually are shown in Table 1.

Cooperation of the ambulatory coaches of the CI children was required to obtain information regarding the schools and teachers involved in these children. Informed consent for this study was obtained from the parents. The teachers were contacted by telephone and given information about the research project. All participating schools were registered at the Inspectorate of Education and had a certificate of quality, which gives an overview of the quality of a school. In those schools the educational test results were sufficient, indicating that student results were as to be expected by standardised protocol measurements.

Medical and rehabilitation files of all CI patients were reviewed for additional and audiologic data (only one child had marginal functional residual hearing). Psychological reports were checked. None of the patients showed a delay in their psychological development and all obtained intelligence quotient scores (IQ) of > 80.

A control group consisted of 26 normal hearing classmates of the CI children in elementary school, chosen by randomised selection, described in the AMP protocol: "in order to ensure that there is no bias in the selection of the non-implanted child, a method of selection has been developed for this purpose.

Table 1. Demographic characteristics

| Child | Current age | Onset deafness | Congenital Prelingual | Cause of Deafness | Duration of deafness | Age at implant | Period CI use | Residual hearing | Cochlear Type CI |
|---|---|---|---|---|---|---|---|---|---|
| 1 | 6.5 | 0.0 | con | Waardenburg | 1.4 | 1.4 | 5.1 | - | N24 |
| 2 | 6.5 | 1.0 | pre | meningitis | 1.6 | 2.6 | 3.9 | - | N24 |
| 3 | 6.8 | 0.0 | con | dysplasia | 2.5 | 2.5 | 4.3 | - | N24 |
| 4 | 6.8 | 1.0 | pre | meningitis | 2.6 | 2.6 | 4.2 | - | N24 |
| 5 | 7.1 | 0.0 | con | hereditary | 2.0 | 2.0 | 5.1 | - | N24 |
| 6 | 7.1 | 2.0 | pre | meningitis | 0.5 | 2.5 | 4.6 | - | N24 |
| 7 | 7.6 | 0.0 | con | unknown | 6.6 | 6.6 | 1.0 | - | N24 |
| 8 | 8.6 | 0.2 | pre | meningitis | 2.4 | 2.5 | 6.0 | - | N24 |
| 9 | 8.7 | 0.0 | con | hereditary | 4.7 | 4.7 | 4.0 | - | N24 |
| 10 | 8.9 | 1.9 | pre | meningitis | 1.4 | 3.3 | 5.6 | - | N24 |
| 11 | 9.3 | 0.0 | con | CMV | 4.7 | 4.7 | 4.6 | - | N24 |
| 12 | 9.4 | 0.0 | con | dysplasia | 3.8 | 3.8 | 5.6 | - | N24 |
| 13 | 9.5 | 0.0 | con | unknown | 4.8 | 4.8 | 4.7 | - | N24 |
| 14 | 9.5 | 0.4 | pre | meningitis | 1.9 | 2.2 | 7.3 | - | N24 |
| 15 | 9.6 | 0.0 | con | hereditary | 4.5 | 4.5 | 5.1 | - | N24 |
| 16 | 9.9 | 0.0 | con | unknown | 2.8 | 2.8 | 7.2 | - | N24 |
| 17 | 10.1 | 0.0 | con | unknown | 4.3 | 4.3 | 5.8 | - | N24 |
| 18 | 11.0 | 0.0 | con | Waardenburg | 5.2 | 5.2 | 5.8 | - | N24 |
| 19 | 11.0 | 0.0 | con | unknown | 3.6 | 3.6 | 7.5 | - | N22 |
| 20 | 11.1 | 0.0 | con | unknown | 6.7 | 6.7 | 4.5 | - | N24 |
| 21 | 11.3 | 0.0 | con | Pendred | 9.7 | 9.7 | 1.7 | + | N24 |
| 22 | 11.3 | 0.0 | con | Usher | 3.7 | 3.7 | 7.6 | - | N24 |
| 23 | 12.0 | 0.9 | pre | meningitis | 2.0 | 2.9 | 9.1 | - | N22 |
| 24 | 12.5 | 1.4 | pre | meningitis | 2.2 | 3.6 | 8.9 | - | N22 |
| 25 | 12.7 | 0.0 | con | Usher | 9.0 | 9.0 | 3.7 | - | N24 |
| 26 | 12.8 | 0.0 | con | unknown | 7.0 | 7.0 | 5.8 | - | N24 |
| Mean | 9.5 | 0.3 | | | 3.9 | 4.2 | 5.3 | | |
| Range | 6.5 - 12.8 | 0.0 - 2.0 | | | 0.5 - 9.7 | 1.4 - 9.7 | 1.0 - 9.1 | | |

All ages and periods are mentioned in years. con=congenital and pre=prelingual onset of deafness

Teachers will list all children's names in alphabetical order, omitting the child with the implant from the list. When using the kindergarten/preschool AMP, the fourth child will be selected as the control. When using the elementary/high school AMP, the eighth child will be selected, if this cannot be accomplished due to small class size, then the teacher should complete the AMP for the last child on the list". The parents of the non-CI children were given a short questionnaire by the teacher, which comprised six questions about the hearing of their child, in order to be able to objectify their normal hearing, which was confirmed in all cases. Results of this control group were used for questionnaire analysis.

**Questionnaires**

The Children's Hearing Institute, New York, USA, developed the Assessment of Mainstream Performance (AMP) to determine the skills that children require to be successful in mainstream school settings[2]. The AMP has two versions: one is utilised with preschool and kindergarten aged children (age 3-5) and comprises 16 questions. The other version, which consists of 22 questions, is for children educated in elementary or high school settings. The items of both versions investigate the child's ability to participate in a range of typical classroom activities and behaviours that are age and content appropriate. In the AMP-Elementary, four questions are stated negatively so these answers were recoded before computation. Answers to the questions are categorised in percentage of time that a child shows certain age and content appropriate behaviour (almost never; 0-10%, to almost always; 91-100%). Class ranking, a separate question within the AMP, indicates the child's level in regard to their peers, estimated by the teacher, ranging from failure (0-25%) to excellent (91-100%). Both versions were validly translated in Dutch (translated twice by different translators, compared and translated backwards) and sent to the teachers by mail.

The second instrument used, is the Screening Identification For Targeting Educational Risk (SIFTER). This instrument was used complementary to the AMPs to rule out the possibility that children have an abnormal behaviour due to an unknown problem in education. The SIFTER is a test designed to provide a valid method by which children with hearing problems can be screened educationally. The SIFTER is a short, 15-item teacher-rating test that explores several areas of school performance, being: academics, attention, communication, class participation, and school behavior. The five content areas comprise three related questions answered through a ranking scale, from one to five. Scoring the SIFTER involves summing the responses of the three questions in each content area. Responses are then placed on a chart to develop a profile of the subject, composing three categorical outcome measures (failure, marginal or sufficient)[8;9]. There are two versions of the SIFTER; a preschool version for young children (aged 3 – kindergarten) and one for older children. The SIFTER has been field-tested and has been shown to have good content and score reliability[10].

**Audiologic data and Language tests**

The Reynell test[11] was used to measure comprehensive language development, while the Schlichting test[12] was used for the expressive language development.

The Dutch Reynell language test is a translation of the original Reynell developmental language scales (RDLS). The RDLS consists of a set of scales, 87 items in 12 sections in the Dutch/Flemish translation, for testing receptive language development for children between 3 and 6;3 years of age. The Schlichting test was first described by Schlichting et al.[12] and later validated and augmented by Eldik et al.[13]. The Schlichting test consists of tests for sentence and word production development. The sentence

development test contains 40 items (with emphasis on imitation) and the word development test 62 items, divided over 10 sections. In addition, the results of the last visit Gestel-Nijmegen (GN) test[14-16], a Dutch Consonant-Vowel-Consonant (CVC) test for open speech scores, are mentioned in Table 4 as well. Analyses of the GN-scores and the questionnaire outcomes were previously described[3].
Tests were administered by a speech therapist. The test results can be expressed in standard scores, Q-scores or age-equivalent scores. Scores were obtained as age-standard and equivalent age. The Q-scores (percentile range) were not used for further analyses, as it was not possible to compute the Q-score for all CI children in the current study. Q-scores can only be computed for children within the application range, nevertheless the number of months behind in language development could be obtained for older children as well, as they did not exceed the maximum language age of 6 years and 3 months. Language development delay was expressed as the difference between equivalent age in months and calendar age in months.

**Statistical analyses**
Data were analysed using SPSS version 12.0. Final results of the AMP and SIFTER domains were computed following the guidelines of each questionnaire. Correlation analysis was based on the non-parametric Spearman's correlation.
Predictive function of AMP and SIFTER with regard to language tests was established by univariate analyses (full factorial, type III sum of squares) of the individual relation of the diverse questions of both questionnaires. A linear regression analysis, forward approach, was done afterwards on questions that correlated significantly. This analysis was performed to determine strongest relating and predicting questions.

# Results

**Factor analyses and Internal consistency of AMP and SIFTER**
Factor analysis was performed to identify the subscales of the AMP and the SIFTER. The number of factors to be retained was determined based on screeplot and eigenvalues > 1; varimax rotations were performed afterwards. These analyses were exploratory in order to reveal whether items supposed to belong to the same scale, grouped coherently into a factor. Factor analyses of both the AMP and SIFTER revealed the presence of several factors.
In the AMP there were 5 factors with an eigenvalue > 1, nevertheless there was only one factor that contributed significantly to the amount of variance. This one factor counted for over 46% of the variance, whereas the other factors with eigenvalue > 1,

showed only not even 8% of the variance. Hence it can be concluded that one strong factor within the AMP was detected.
This is in concordance with the supposed structure of the AMP (one general scoring domain) [Table 2].

Table 2. AMP factor analyses

| Factor | Initial Eigenvalues | | | Extraction Sums of Squared Loadings | | | Rotation Sums of Squared Loadings | | |
|---|---|---|---|---|---|---|---|---|---|
| | Total* | % of Variance | Cumulative % | Total | % of Variance | Cumulative % | Total | % of Variance | Cumulative % |
| 1 | 10,19 | 46,30 | 46,30 | 10,19 | 46,30 | 46,30 | 5,63 | 25,58 | 25,58 |
| 2 | 1,70 | 7,71 | 54,01 | 1,70 | 7,71 | 54,01 | 4,19 | 19,04 | 44,62 |
| 3 | 1,49 | 6,77 | 60,78 | 1,49 | 6,77 | 60,78 | 2,72 | 12,36 | 56,98 |
| 4 | 1,33 | 6,03 | 66,81 | 1,33 | 6,03 | 66,81 | 1,77 | 8,04 | 65,01 |
| 5 | 1,20 | 5,43 | 72,24 | 1,20 | 5,43 | 72,24 | 1,59 | 7,23 | 72,24 |
| 6 | 0,91 | 4,12 | 76,36 | | | | | | |
| 7 | 0,83 | 3,79 | 80,15 | | | | | | |
| 8 | 0,71 | 3,24 | 83,39 | | | | | | |
| 9 | 0,63 | 2,85 | 86,24 | | | | | | |
| 10 | 0,49 | 2,23 | 88,47 | | | | | | |
| 11 | 0,44 | 3,00 | 90,46 | | | | | | |
| 12 | 0,40 | 1,81 | 92,27 | | | | | | |
| 13 | 0,31 | 1,40 | 93,67 | | | | | | |
| 14 | 0,29 | 1,30 | 94,97 | | | | | | |
| 15 | 0,25 | 1,12 | 96,09 | | | | | | |
| 16 | 0,19 | 0,86 | 96,95 | | | | | | |
| 17 | 0,17 | 0,77 | 97,71 | | | | | | |
| 18 | 0,13 | 0,58 | 98,29 | | | | | | |
| 19 | 0,12 | 0,53 | 98,82 | | | | | | |
| 20 | 0,10 | 0,45 | 99,26 | | | | | | |
| 21 | 0,09 | 0,41 | 99,67 | | | | | | |
| 22 | 0,07 | 0,33 | 100,00 | | | | | | |

The SIFTER showed a total of four factors with eigenvalue > 1, indicating a four-dimensional structure. Here again, one main factor can be detected, though the difference in explained variance is less than within the AMP (first factor 44% and the next 12%). Unfortunately, this outcome, however, is not consistent with the expected structure, as the SIFTER is supposed to have 5 domains [Table 3].

Table 3. SIFTER factor analyses

| Factor | Initial Eigenvalues | | | Extraction Sums of Squared Loadings | | | Rotation Sums of Squared Loadings | | |
|---|---|---|---|---|---|---|---|---|---|
| | Total* | % of Variance | Cumulative % | Total | % of Variance | Cumulative % | Total | % of Variance | Cumulative % |
| 1 | 6,62 | 44,14 | 44,14 | 6,62 | 44,14 | 44,14 | 4,45 | 29,66 | 29,66 |
| 2 | 1,86 | 12,37 | 56,51 | 1,86 | 12,37 | 56,51 | 3,31 | 22,07 | 51,73 |
| 3 | 1,36 | 9,09 | 65,60 | 1,36 | 9,09 | 65,60 | 2,08 | 13,87 | 65,60 |
| 4 | 1,07 | 7,15 | 72,75 | 1,07 | 7,15 | 72,75 | 1,76 | 11,73 | 72,75 |
| 5 | 0,89 | 5,96 | 78,72 | | | | | | |
| 6 | 0,69 | 4,62 | 83,34 | | | | | | |
| 7 | 0,61 | 4,06 | 87,39 | | | | | | |
| 8 | 0,44 | 2,91 | 90,30 | | | | | | |
| 9 | 0,37 | 2,46 | 92,77 | | | | | | |
| 10 | 0,32 | 2,12 | 94,88 | | | | | | |
| 11 | 0,27 | 1,80 | 96,68 | | | | | | |
| 12 | 0,16 | 1,08 | 97,76 | | | | | | |
| 13 | 0,14 | 0,92 | 98,68 | | | | | | |
| 14 | 0,12 | 0,78 | 99,47 | | | | | | |
| 15 | 0,08 | 0,53 | 100,00 | | | | | | |

Extraction Method: Principal Component Analysis. * Total is the amount of variance in the observed variables accounted for by each factor

Table 4. Questionnaire analysis

| Questionnaire | | Scale statistics | | |
|---|---|---|---|---|
| | | Cronbach α | Mean | SD |
| AMP-Elementary | | 0.93 | 101.88 | 16.76 |
| SIFTER | Academics | 0.68 | 11.31 | 2.32 |
| | Attention | 0.79 | 9.81 | 2.81 |
| | Communication | 0.87 | 9.49 | 3.06 |
| | Participation | 0.74 | 10.84 | 2.89 |
| | Behavior | 0.63 | 13.06 | 1.98 |

Reliability analyses results. Scale statistics: Cronbach α, scale means and standard deviation (SD).

Reliability analyses (internal consistency) were performed to study the properties of the measurement scales and their items. The reliability analysis procedure calculates a number of commonly used measures of scale reliability and also provides information about the relationships between individual items in the scale. The Cronbach alpha was estimated, based on the average inter-item correlation. Both the AMP and SIFTER domains showed good reliability, as all Cronbach α scores were > 0.60 [Table 4.].

**Questionnaires**

The teachers of CI children in mainstream school settings filled in the AMP and SIFTER questionnaire. Questionnaire results of the children included in the current study are provided in Table 5. As can be seen, in both questionnaires and within all domains, inter-individual variability is detected. The AMP overall scores (maximum 6, minimum 1) ranged from 2.9 to 5.5, indicating that appropriate behavior was shown nearly sometimes (score 3) to often/always (score 5/6). Class ranking scores (best score 1, worst score 5) ranged from 1.0, being outstanding, to 4.0, being below average. The divers SIFTER domain-scores showed even greater ranges from failure to sufficient (academics 6-15, attention 3-13, communication 4-13, class participation 4-13 and school behaviour 7-15). In general, the study population showed 'often' appropriate classroom behavior (AMP) and scored overall 'above average' in their class ranking (AMP). Three out of five SIFTER domains were regarded 'sufficient' (academics, class participation and school behavior), one was in between 'marginal' and 'sufficient' (attention) and the domain of communication was rated as 'failure'.

**Language tests**

The last visit (mostly at 3 to 5 years after implantation) Reynell and Schlichting test results are presented in Table 6. Again the inter-individual variability is clearly present. Age equivalent scores were obtained and the number of months behind in normal language development was used for further analyses. A negative data indicate a better language development than expected at that specific age. In total, it can be said that children lag 30-35 months on the average in expressive and receptive language development and that only two children scored equal or better than their normal hearing peers. The results of the last visit Gestel-Nijmegen (GN) test, open speech scores, are shown as well. The GN word scores vary widely from 5 to 90% and the GN phoneme scores are higher and vary a little less from 42 to 95%.

Table 5. Questionnaire results

| Child | AMP elementary | Class ranking | SIFTER academics | SIFTER attention | SIFTER communication | SIFTER class participation | SIFTER school behaviour |
|---|---|---|---|---|---|---|---|
| 1 | 4.9 | 4.0 | 15 | 12 | 11 | 13 | 14 |
| 2 | 4.1 | 2.0 | 10 | 8 | 7 | 7 | 13 |
| 3 | 4.7 | 4.0 | 13 | 7 | 6 | 11 | 12 |
| 4 | 4.1 | 3.0 | 11 | 8 | 10 | 7 | 13 |
| 5 | 5.5 | 4.0 | 13 | 11 | 9 | 12 | 13 |
| 6 | 4.7 | 4.0 | 12 | 12 | 10 | 12 | 14 |
| 7 | 3.4 | 2.0 | 7 | 7 | 4 | . | . |
| 8 | 4.5 | 4.0 | . | . | . | . | . |
| 9 | 2.9 | 3.5 | 11 | 11 | 7 | 5 | 8 |
| 10 | 4.8 | 4.0 | 9 | 13 | 6 | 13 | 11 |
| 11 | 2.9 | 1.0 | 6 | 3 | 4 | 4 | 7 |
| 12 | 4.5 | 4.0 | 12 | 10 | 8 | 11 | 15 |
| 13 | 3.6 | 2.0 | 12 | 3 | 6 | 7 | 15 |
| 14 | 5.5 | 4.0 | 14 | 9 | 12 | 11 | 14 |
| 15 | 4.1 | 4.0 | 11 | 7 | 10 | 7 | 12 |
| 16 | 3.8 | 2.0 | 10 | 6 | 6 | 9 | 11 |
| 17 | 3.5 | 1.0 | 7 | 7 | 4 | 7 | 14 |
| 18 | 4.4 | 3.0 | 13 | 10 | 12 | 12 | 15 |
| 19 | 4.1 | 3.0 | 10 | 12 | 7 | 9 | 13 |
| 20 | 4.1 | 3.0 | 12 | 8 | 8 | 9 | 11 |
| 21 | 3.5 | 3.5 | 6 | 4 | 3 | 6 | 12 |
| 22 | 4.5 | 3.5 | 9 | 9 | 8 | 8 | 9 |
| 23 | 4.2 | 2.0 | 10 | 7 | 7 | 9 | 15 |
| 24 | 4.6 | 4.0 | 9 | 12 | 8 | 13 | 12 |
| 25 | 3.6 | 3.5 | . | 8 | 5 | 8 | 14 |
| 26 | 3.6 | 2.0 | 7 | 9 | 5 | 10 | 15 |
| Mean | 4.1 | 3.1 | 10.4 | 8.5 | 7.3 | 9.2 | 12.6 |
| Range | 2.9 - 5.5 | 1.0 - 4.0 | 6 - 15 | 3 - 13 | 3 - 12 | 4 - 13 | 7 - 15 |

| AMP | AMP Elementary (appropriate behaviour) | Class Ranking | | SIFTER | Academics | Attention | Commu-nication | Class participation | School behavior |
|---|---|---|---|---|---|---|---|---|---|
| 1 | Almost never | Outstanding | (91-100%) | Sufficient | ≥ 10 | ≥ 9 | ≥ 11 | ≥ 9 | ≥ 10 |
| 2 | Seldom | Good | (75-90%) | Marginal | 8-9 | 7-8 | 8-10 | 7-8 | 8-9 |
| 3 | Sometimes | Above average | (51-74%) | Failure | ≤ 7 | ≤ 6 | ≤ 7 | ≤ 6 | ≤ 7 |
| 4 | Regularly | Below average | (26-50%) | | | | | | |
| 5 | Often | Weak | (0-25%) | | | | | | |
| 6 | Almost always | | | | | | | | |

Patient numbers correspond with the patient numbers in Table 1

Table 6. Language development. Outcomes of Reynell and Schlichting test are expressed in number of months lagging in language development compared to normal development at last visit. Negative sign indicates a language development better than expected at a certain age in normal hearing children. Gestel-Nijmegen (GN) test results are expressed in percentages.

| Child | Reynell | Schlichting Sentence | Schlichting Word | GN Word | GN Phoneme |
|---|---|---|---|---|---|
| 1 | 7 | 8 | . | . | . |
| 2 | 9 | 20 | 13 | 75 | 90 |
| 3 | 19 | 24 | 16 | 75 | 88 |
| 4 | 22 | 25 | 17 | 85 | 95 |
| 5 | 15 | 17 | 13 | 65 | 85 |
| 6 | -11 | 3 | 3 | 50 | 90 |
| 7 | 60 | 59 | 55 | 60 | 83 |
| 8 | -3 | . | . | . | . |
| 9 | 54 | 56 | 51 | 30 | 67 |
| 10 | 37 | 42 | . | 20 | 55 |
| 11 | 48 | 55 | 45 | 21 | 54 |
| 12 | 44 | 55 | 46 | 35 | 67 |
| 13 | 34 | 35 | 18 | 80 | 93 |
| 14 | 21 | 20 | 7 | 80 | 90 |
| 15 | 30 | 30 | 24 | 5 | 42 |
| 16 | 30 | 24 | 15 | 15 | 45 |
| 17 | 58 | 68 | 57 | 65 | 85 |
| 18 | 25 | 17 | 17 | . | . |
| 19 | 38 | 47 | 25 | 65 | 78 |
| 20 | 44 | 41 | 41 | 70 | 83 |
| 21 | . | . | . | . | 45 |
| 22 | 39 | 43 | 36 | 30 | 55 |
| 23 | 13 | 32 | 27 | 50 | 72 |
| 24 | 32 | 34 | 33 | 90 | 93 |
| 25 | . | . | . | 40 | 68 |
| 26 | 69 | 67 | 66 | 35 | 50 |
| Mean | 30.6 | 35.7 | 29.8 | 51.9 | 72.7 |
| Range | -11 - 69 | 3 - 68 | 3 – 66 | 5 - 90 | 42 - 95 |

Patient numbers correspond with the patient numbers in other tables

## Correlation analyses

To be able to establish whether the detected variability in AMP and SIFTER results could be explained by the variability in language test results, divers correlations were examined by Spearman's rho correlation. Outcomes are presented in Table 7 and the most significant outcomes also graphically in Figure 1. The number of months behind in language development correlated negatively with the instrument results, indicating that a better score on the instruments was related to a shorter period behind in language development.

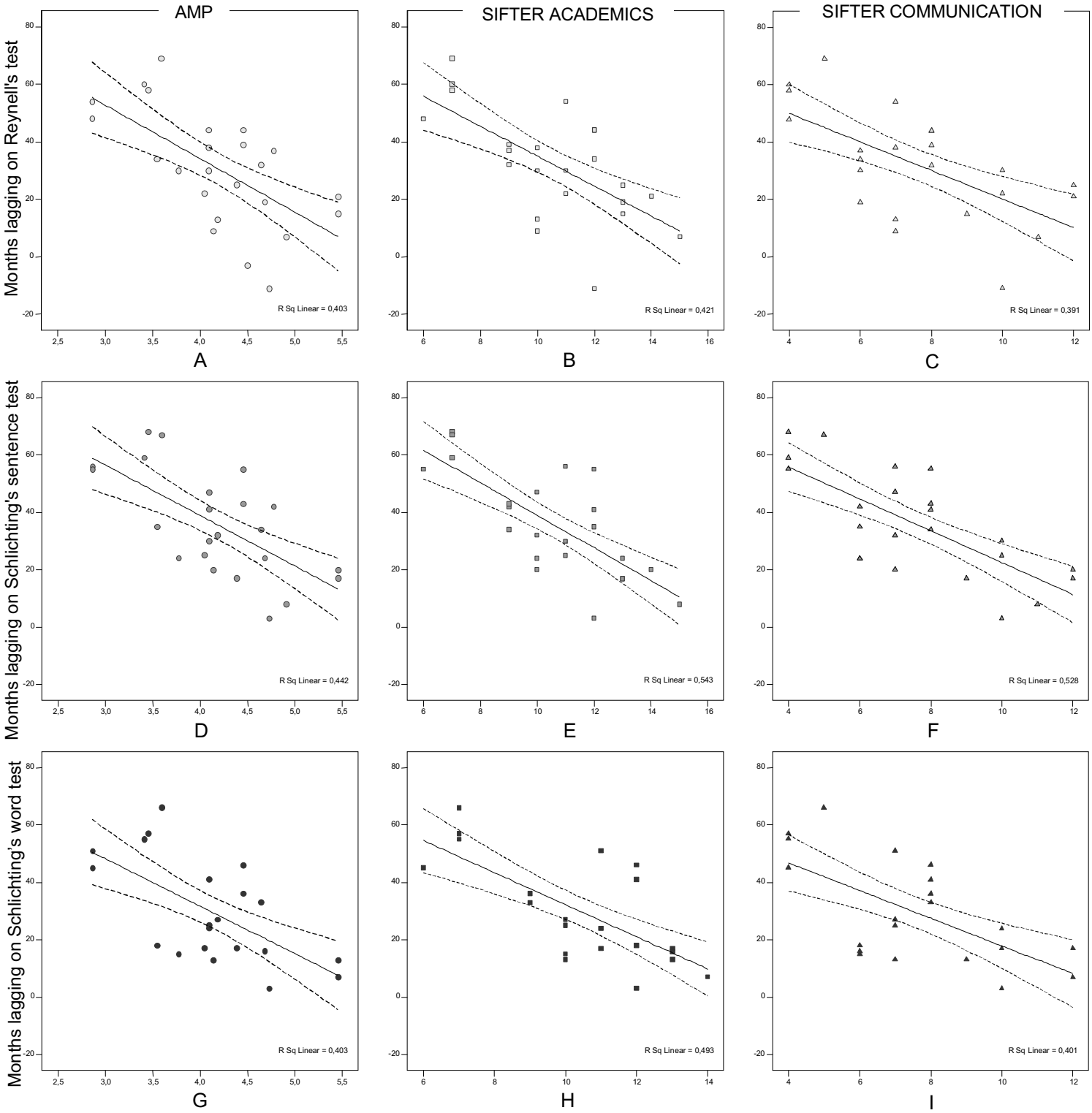


Figure 1. Scatterplot of the diverse language tests (results in number of months that a child stays behind in language development) and the questionnaire results of the AMP and SIFTER Academics and Communication, based on the estimates of Spearman's correlation. Linear fit line and 95% confidence interval lines are shown.
1A: Correlation Reynell test result and mean AMP score; 1B: Correlation Reynell test result and mean SIFTER Academic score; 1C: Correlation Reynell test result and mean SIFTER Communication score; 1D: Correlation Schlichting sentence test result and mean AMP score; 1E: Correlation Schlichting sentence test result and mean SIFTER Academic score; 1F: Correlation Schlichting sentence test result and mean SIFTER Communication score; 1G: Correlation Schlichting word test result and mean AMP score; 1H: Correlation Schlichting word test result and mean SIFTER Academic score; 1I: Correlation Schlichting word test result and mean SIFTER Communication score

Table 7. Relation Language development and questionnaire results
Analysis of the diverse language tests (results in number of months that a child stays behind in language development) and the questionnaire results of the AMP and SIFTER, based on the estimates of Spearman's correlation coefficient.

| | | Reynell | | Schlichting sentence | | Schlichting word | |
|---|---|---|---|---|---|---|---|
| | | Coëff. | p-value | Coëff. | p-value | Coëff. | p-value |
| Class ranking (AMP questionnaire) | | -0.46 | 0.02 | -0.48 | 0.02 | -0.40 | 0.07 |
| AMP elementary | | -0.64 | <0.01 | -0.67 | <0.01 | -0.63 | <0.01 |
| SIFTER | Academics | -0.65 | <0.01 | -0.72 | <0.01 | -0.64 | <0.01 |
| | Attention | -0.18 | ns | -0.20 | ns | -0.12 | ns |
| | Communication | -0.60 | <0.01 | -0.70 | <0.01 | -0.56 | 0.01 |
| | Participation | -0.40 | ns | -0.47 | 0.03 | -0.36 | ns |
| | Behavior | -0.25 | ns | -0.19 | ns | -0.10 | ns |

Coëff.: Correlation coefficient, Sign. : Significance (2-tailed), ns: non significant

Significant correlations were detected when comparing the language tests and both questionnaires. These findings illustrate the probable explanation of individual questionnaire variability by language test results (AMP, Class ranking, SIFTER academics and SIFTER communication).
Possible relations between language development and child/implant variables were investigated [Table 8]. Current age correlated significantly with Reynell and Schlichting results, surprisingly as both tests are age-corrected. The age at onset of deafness only showed a significant relation with the Reynell results: the younger children became deaf, the worse their comprehensive Reynell language test results (the longer the period behind in language development). Duration of deafness and age at implantation were both implant variables with a very strong significant correlation with language developmental results. Period of CI use did not show any relation with language test results.

Table 8. Relation Language development and implant variables
Analyses of the diverse language tests (results in number of months that a child stays behind in language development) and implant variables, based on the estimates of Spearman's correlation coefficient.

| | Reynell | | Schlichting sentence | | Schlichting word | |
|---|---|---|---|---|---|---|
| | Coëff. | p-value | Coëff. | p-value | Coëff. | p-value |
| Child/Implant variables | | | | | | |
| Age | 0.47 | 0.02 | 0.45 | 0.05 | 0.45 | 0.04 |
| Age at onset of deafness | -0.43 | 0.04 | -0.30 | ns | -0.41 | ns |
| Duration of deafness | 0.73 | <0.01 | 0.60 | <0.01 | 0.70 | <0.01 |
| Age at implantation | 0.82 | <0.01 | 0.70 | <0.01 | 0.77 | <0.01 |
| Period of CI use | -0.11 | ns | -0.10 | ns | -0.03 | ns |

Coëff.: Correlation coefficient, Sign. : Significance (2-tailed), ns : non significant

Finally, we have looked at the probable relationship between language tests and speech perception tests [Table 6]. It was found that there is a strong significant relation between GN test (speech recognition, word and phoneme) and expressive as well as receptive language development ($p = 0.01$ to $0.05$). Additional analyses were performed to determine the relation between the language tests. Results of correlation analyses illustrate a strong significant relation between Reynell and both Schlichting language tests (correlation coefficient $> 0.93$ and p-value $< 0.01$).

**Predictive function AMP/SIFTER on language development**

The General Linear Model Univariate procedure provided regression analysis and analysis of variance for one dependent variable (language test results, months behind in development) by more variables (questionnaire items). Using this General Linear Model procedure, the null hypotheses about the effects of questionnaire items on the single dependent variable (language test results) were tested. These analyses showed that ten out of twenty-two items of the AMP and three out of fifteen items of the SIFTER correlated significantly with the language test results ($p < 0.03$). Those items, that showed the significant relation, were further analysed by linear regression. Linear regression estimated the coefficients of the linear equation, involving one or more independent variables (the questionnaire items), that best predict the value of the dependent variable (language test result). This way it is possible to predict the language development, the dependent variable, of a CI child in mainstream elementary school from questionnaire items, the independent variables.

Closing results showed that four AMP questions have a strong predictive value for the language developmental outcomes, namely question 4, 6, 9 and 22. These items comprise of questions regarding communicative breakdown, following directions, giving appropriate comment and taking leadership role. Within the SIFTER questionnaire only one item showed this predictive capacity: question 1, where the level of achievement in class was asked (academic achievement).

# Discussion

In the current study two instruments were used for the evaluation of CI children in mainstream education and results were compared with language test results. The variability in questionnaire results within this group of 26 CI users appeared to be related to on language performance.

In an earlier study the AMP and SIFTER questionnaires were used as well to measure classroom performance[3]. In that study all the CI students scored "above average" in the AMP, which was a notable positive finding. However, a few specific items did

show a disturbing deprivation in communicative skills of CI students compared to their normal hearing peers. Furthermore, considerable differences were observed between the children. Besides that study, the AMP questionnaire has only been used in one other pilot study[2]. Preliminary results of this pilot study also showed communicative difficulties for the CI students. Both these two earlier reports emphasized the need of further investigation of the questionnaire. In the current study we have examined the structure of the AMP questionnaire by means of factor and reliability analyses, which appeared to be good. Nevertheless, it should be noted that we were not able to include significant amounts of patients and/or controls.

As mentioned above, the SIFTER questionnaire was used in the former study by Damen et al. as well[3]. It showed that CI children were delayed in communication in kindergarten as well as in elementary school. The teachers answered the three questions within the area of communication, on expressive and receptive language skills, less reassuringly for the CI students, in accordance with our current results. During the development of the SIFTER questionnaire in 1989, several areas of risk for children with hearing problems in educational settings were determined, based upon literature research and other instruments. After content validity tests and item analysis, five areas with three questions each remained. Regrettably, the proposed structure was not entirely confirmed with the current analyses. Based upon the small number of patients in our current report, no conclusions can be drawn about the comparison with previous questionnaire structure analyses. The reliability of the SIFTER was previously tested in a large population ($n > 500$) and appeared to be moderate. It was concluded that the questionnaire was appropriate for screening and use in educational settings[10]. This reliability finding was confirmed by our reliability analysis results.

Spoken language development is an important outcome of cochlear implantation for children in mainstream education. By enhancing speech perception, cochlear implantation has allowed children to develop their use of spoken language. Spoken language grammar acquisition, measured by the multiple choice Test for Reception of Grammar, designed to assess the understanding of grammatical contrasts in the English language, was found to be considerably delayed according to Nikolopoulos et al.[17]. However, there was a tendency towards the development of grammar skills following implantation. Highest improvement was seen in children who were implanted before the age of 4, supporting the idea of implantation at a younger age if grammatical competence in spoken language is to be achieved. These findings were corroborated by the current study, as (long-term) language test results (and classroom performance) correlated significantly with the age of implantation.

Few studies addressing the language development of children with implants have used the Reynell tests. Robbins et al. found that language development could be above and beyond that anticipated from maturation alone, showing an increase in rate of language development[18]. Miyamoto et al. showed that rate of learning expressive

language skills of prelingually deaf children with a CI was equivalent to that of normal hearing peers and that no plateau in language learning was observed. They also did see a large variability in language development and suggested that this may have been due to intersubject differences in speech perception. We have looked at this possible relationship and found that there is a strong significant relation between GN test (speech recognition, word and phoneme) and expressive as well as receptive language development[19].

Others have to some extent investigated spoken language development and mainstream educational settings. Several studies were conducted on cost-benefit analyses according to mainstream placement, though few on language development of CI pupils in mainstream educational settings. Higher speech intelligibility scores in 8- to 9-year-old congenitally deafened CI students were associated with educational settings that emphasize oral communication development, such as mainstream education[20]. Children with cochlear implants seemed to have increased educational opportunities, with those children in mainstream and those who have moved toward mainstream demonstrating improved progress in speech perception ability[21]. Those studies did not investigate the child's classroom performance; hence no conclusions about their capacity of mainstreaming could be made. The current study is as far as known, the first one to investigate both classroom capacity of the child and their language development. It was found that within the AMP and SIFTER questionnaires respectively 4 questions and 1 question have a strong predictive value for language development. This implies that the questionnaires may be used for screening in future, as they seem to be able to detect language developmental problems. This way, a CI team can follow (ambulatory) a CI child's performance in mainstream elementary school, without any strain for the child itself, as the teachers fill in the questionnaire. By means of the prognostic value of (certain questions within) the AMP, the actual linguistic functioning of the child can be observed.

Even though substantial statistical analyses were performed on the AMP and SIFTER questionnaire, the relatively small amount of CI pupils should be taken into consideration. The reproducibility of the outcomes of analyses should be retested in large group settings. Nevertheless, the vast statistically significant correlations between questionnaires and language test results show an obvious predictive value of certain questions within the AMP and SIFTER for language development.

# Conclusion

Individual classroom performance, measured by the AMP and SIFTER questionnaires, of CI children in mainstream education varies. Correlation analyses

showed a strong significant relation between questionnaire results (classroom performance) and both expressive and receptive language test results (Schlichting and Reynell tests). Structural questionnaire analyses of the AMP and SIFTER demonstrated good reliability. It was found that within the AMP and SIFTER questionnaires respectively 4 questions and 1 question have a strong predictive value for language development. The AMP is an additional instrument for a CI team to follow (ambulatory) a CI child's performance in mainstream elementary school, without any strain for the child itself, as the teachers fill in the questionnaire. By means of the predictive value of (certain questions within) the AMP, the actual linguistic functioning of the child can be monitored. Longitudinal follow up studies have to establish the final importance and value of the AMP.

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# The Usher lifestyle survey

2.2

The Usher lifestyle Survey: maintaining independence; a multi-centre study.
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# Abstract

*Aim:* Patients with Usher syndrome face a special set of challenges in order to maintain their independence when their sight and hearing worsen. An insight in these challenges is described. Three different types of Usher (I, II and III) were investigated.

*Methods:* In this study 93 Usher patients from 7 European countries filled out a questionnaire on maintaining independence (60 patients type I, 25 patients type II, 4 patients type III and 4 patients type unknown). Results of Usher type I and II patients are presented.

*Results:* Following the Nordic definition of maintaining independence in deaf-blindness, three domains are investigated: access to information, communication and mobility. Research variables in this study are: age and type of Usher, considered hearing loss and the number of retinitis pigmentosa related sight problems.

*Conclusion:* Usher type I patients tend to need more help than Usher type II patients and the amount of help that they need grows when patients get older or when considered hearing loss worsens. No patterns in results were seen for the number of retinitis pigmentosa related sight problems.

# Introduction

Usher syndrome is a genetic disorder that causes sensorineural hearing loss, retinitis pigmentosa (RP), and sometimes balance problems. The prevalence is estimated to range from 3.5 to 6.2 in 100.000, making it the most common cause of deafblindness[1]. Distinction is made between three clinically different syndromes, Usher type I, II and III. The different types are distinguished by differences in onset, progression and severity of hearing loss and by presence or absence of balance problems[2].
Usher type I is characterised by congenital profound deafness, absence of vestibular function and progressive vision loss due to RP. The mean age of the identification of RP is around 13-14 years of age. In Usher type II, prelingual moderate to severe hearing loss, RP and an intact vestibular function are seen. The audiogram has a characteristic down sloping pattern that is mildly progressive[3]. RP is usually identified in the second decade. In Usher type III a considerable variety is seen in type and degree of progressive hearing loss, RP and vestibular function[4]. With young children and teenagers, type III cases appear like type II but, in adulthood, their audiological phenotype resembles type I.
Patients with Usher syndrome face a special set of challenges in order to maintain their independence. In 2001, Sense has launched the project CAUSE (Charge Association Usher Syndrome Europe). One of the aims of this project was to explore the day-to-day challenges that people with Usher syndrome have to deal with. Sense is the largest specialist organisation in the UK working with people with deafblindness and associated disabilities. A survey, especially designed for this project, comprises items focussed on maintaining independence. Research issues addressed in this study are:

- Is it more difficult to remain independent while getting older, with regard to the type of Usher?
- Is it more difficult to remain independent when hearing worsens, regardless of the type of Usher?
- Is it more difficult to remain independent when the number of RP problems increases?

# Materials and methods

**Background of the CAUSE project**

The CAUSE project was an 18-month European Union Project, designed at promoting awareness of Usher Syndrome and CHARGE association. The project, which commenced in November 2001, was supported by the European Usher

Syndrome Network and the CHARGE Network, together with project partner organisations: CRESAM and Retina France (France), Fighting Blindness (Ireland), ONCE (Spain), Lega del Filo d'Oro (Italy), Sense (UK), and Blindeninstitutsstiftung (Germany). The partner organisations are large corporate members of "deafblind international" (DBI), a world association for the benefit of international networking and sharing info for deafblind people. Usher patients from Viataal/St Radboud UMCN (the Netherlands), not part of CAUSE, completed the CAUSE questionnaire at a later date. The CAUSE project was co-ordinated by Sense. Some results were presented at the CAUSE conference in March 2003[5].

**Patients**

The project partners distributed the questionnaires to members with Usher syndrome, irrespective the type of Usher. Project partners were responsible for providing support necessary to enable the person with Usher to complete the questionnaire. All data were coded, in order to assure anonymity for the cooperating patients. In April 2004 the Radboud University Nijmegen Medical Centre, in cooperation with Viataal in St. Michielsgestel the Netherlands, started their participation in this project. Patients with Usher type I were asked to participate in the survey as part of the cochlear implant (CI) quality of life research project. Patients with CI were addressed in person. The Usher type I patients without CI answered the questionnaire by mail. The total number of participating patients in this study was 93 (Table 1), consisting of 50 male (53.8 %) and 43 female (46.2 %) patients. Three age groups of patients with arbitrary limits were defined: 18 people (19.4%) were younger than 25, 41 people (44.0%) were aged 25 to 45 and 34 people (36.6%) were aged older than 46.

Table 1. Data participating partner organisations

| Country | Usher Types | | | | |
|---|---|---|---|---|---|
| | Type I | Type II | Type III | Type unknown | TOTAL |
| France | 6 | 0 | 0 | 0 | 6 |
| Germany | 4 | 7 | 2 | 0 | 13 |
| Ireland | 2 | 7 | 0 | 0 | 9 |
| Italy | 8 | 4 | 0 | 3 | 15 |
| Spain | 9 | 2 | 1 | 0 | 12 |
| United Kingdom | 5 | 5 | 1 | 1 | 12 |
| The Netherlands | 26 | 0 | 0 | 0 | 26 |
| TOTAL | 60 | 25 | 4 | 4 | 93 |

**Survey**

A cross-sectional survey study was undertaken. The original survey in the Sense study (Appendix) contained three components: first general information about the patient (component 1), then clinical items (component 2, 3, 4, 5) and, thirdly, items about remaining independent (component 6, 7, 8). Items were open-ended questions and questions with multiple choice answer possibilities. Some items were left out or were unanswered by some patients.

In 1980 the Nordic countries agreed to a common definition of domains of independence that are adversely affected in deafblindness. These domains are: ability to give and receive information (communication), access to information and mobility. With regard to the survey the domains are represented in specific subdomains or items. Each of these specific items is addressed to get an overview of the issues involved. (Table 2)

Table 2. Nordic definition of independence in deafblindness

| Nordic Defenition | Domains | Questionnaire items | Recoded answers |
|---|---|---|---|
| Independence in Deafblindness | Information | Wake up (Q. 6.3.1) | 1= independently<br>2= with equipment<br>3= with others<br>4= not aware |
| | | Front door (Q.6.3.2) | |
| | | Access form (Q.6.3.5) | |
| | | Emergency (Q.6.3.6) | |
| | Communication | Telephone use (Q.6.1.3) | 0 = no use to 7 = 7 different ways |
| | | Written communication (Q.6.2) | 0 = no use to 8 = 8 different ways |
| | | Buy food (Q.6.3.3b) | 0= independently<br>1= with others |
| | | Communicate doctor (Q.6.3.4b) | |
| | Mobility | Visit shop (Q.6.3.3a) | |
| | | Visit doctor (Q.6.3.4a) | |

Four items were thought to relate to the domain "access to information". These items are: 'the way of waking up in the morning' (6.3.1), 'the way of realising someone is at the front door' (6.3.2), 'the way in which access to a form is gained' (6.3.5) and 'the way in which an emergency situation at home is discovered' (6.3.6). Answers to these open-ended questions were recoded into 4 answering categories; 1= independently, 2= independently with extra equipment (e.g. vibrating devices or flashlight indicators), 3= with help of others (e.g. family or social workers) and 4= not aware or no access. Only results of first choice answers are shown.

The domain of "communication" contains four items, namely (1) 'the use of a telephone', (2) 'the use of written information', (3) 'the ability to buy food in a shop' and (4) 'the ability to communicate with a doctor independently'. In item 6.1.3 is asked whether a patient uses a telephone and if so, what kind of telephone(s). A second item

covers the capacity and the ease of use of mainly written communication (6.2). In both these items we looked at the number of ways in which the patient is able to use this type of communication. The items of 'buying food in a shop' and 'communication with the doctor' are addressed in questions 6.3.3b and 6.3.4b. These were open-ended questions, but answers were easily recoded into "no help needed" (=0) and "help of other people needed" (=1).
The last domain is "mobility" and is divided into two items; 'visiting a shop' (6.3.3.a) and 'visiting a doctor' (6.3.4.a). These questions were open-ended and answers were recoded (0= alone, 1= with others).

**Data analyses**
Before computation of the domains, items were recoded (Table 2). Data were analysed using SPSS version 12.0. Means (standard deviations) and percentages were computed. Descriptives were computed for main characteristics: age, sex, type of Usher, considered hearing impairment and number of RP related sight problems. Item means were compared for groups differing in type of Usher and age, considered hearing loss and number of RP related sight problems. These differences were analysed by non-parametric Kruskal-Wallis tests for multiple responses for continuous data and by the Pearson Chi square tests for categorical data. Where significant differences were found, further analyses were done with Kruskal-Wallis exact testing or Pearson Chi square exact testing between subgroups.

# Results

As seen in Table 1, 93 patients filled out the questionnaire. As only 4 patients with Usher type III answered the questionnaire, we decided to leave Usher type III out of the results (as well as we left out the people who did not know their type of Usher for sure, n=4). Hearing impairment is considered worse in type I than in type II patients.

Type I patients with Cochlear Implants did not consider themselves as profoundly deaf (Figure 1). Visual impairment worsens with age in type I patients. Type I patients report significantly fewer RP-related sight problems compared to type II patients (Figure 2). Support (Q. 7.1) is given by various groups of people. Support by family and friends is reported most in both Usher type I and II (Table 3). Considered identity differed in Type I and II. Type I patients mostly see themselves as Usher people and Type II Usher patients consider themselves mostly hard of hearing and visually impaired (Figure 3).

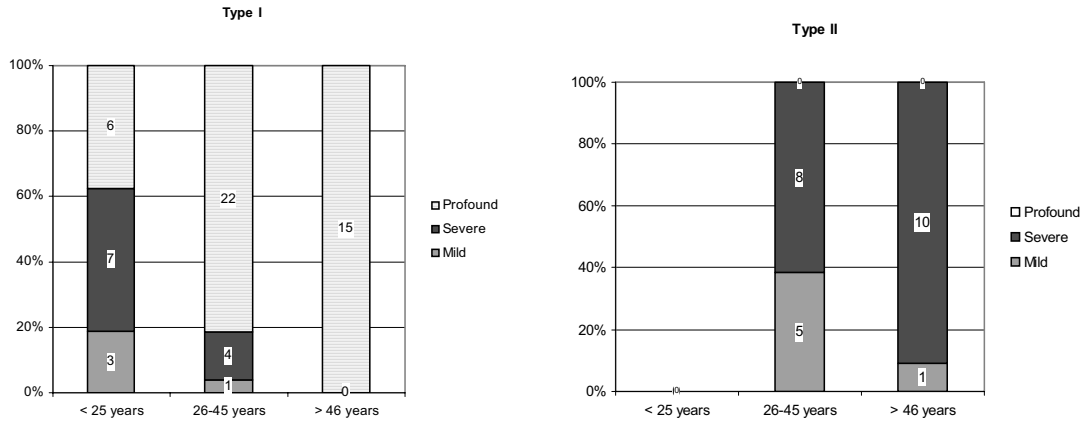


Figure 1. Considered hearing impairment and age per type of Usher (Q.3). Usher type I has Cochlear Implant users in both the younger age groups, this explains why not all type I patients consider themselves profoundly deaf.

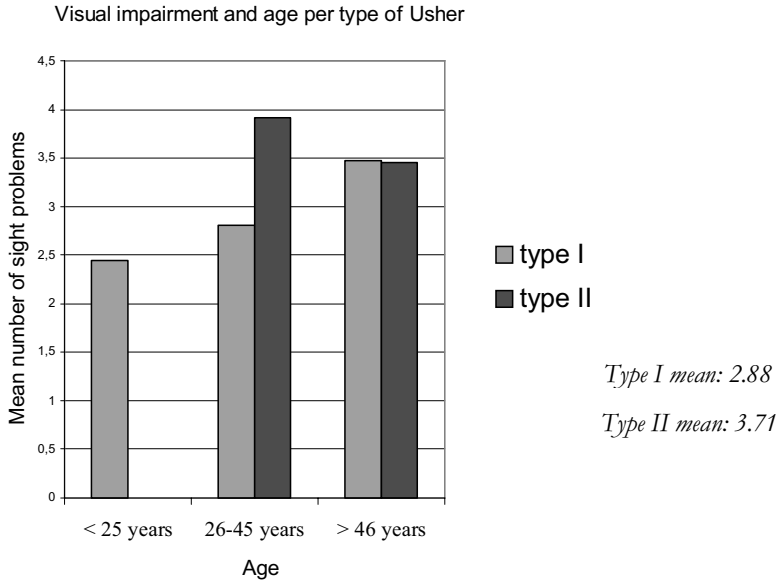


Figure 2. Visual impairment (mean number RP related sight problems) and age, with regard to the type of Usher (Q.4). Patients answered a question about the different RP related sight problems. More than one answer could be ticked, options: night blindness, tunnel vision, scotomas, visual disturbances, and cataract. The mean number of ticked problems is shown per age group of Usher patients. The mean number of RP-related sight problems is shown for type I and II respectively. Type I patients report significantly fewer RP-related sight problems compared to type II patients (p=0.001).

Table 3. Social and emotional support in Usher type I and II patients

| Support % (n) | Family | Friends | Volunteers | Professionals |
|---|---|---|---|---|
| Type I | 81.0 (47) | 86.7 (52) | 27.6 (16) | 62.1 (36) |
| Type II | 96.0 (24) | 92.0 (23) | 40.0 (10) | 28.0 (7) |

In Q. 7.1 people were asked from what individuals they receive support. More answers could be ticked.

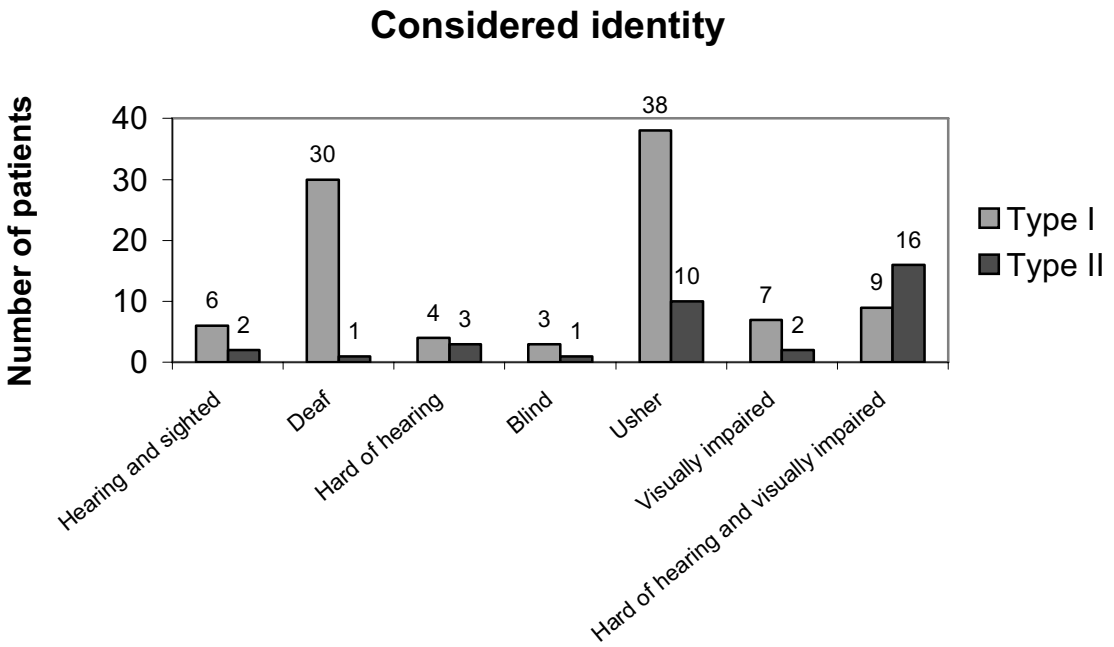


Figure 3. Considered identity per type of Usher (Q.7.3). More answers could be given

Table 4. Maintaining independence, based on the Nordic definition of deafblindness, with regard to age and type of Usher

| Nordic definition of deafblindness | | Usher Type I | | | Usher Type II | | |
|---|---|---|---|---|---|---|---|
| | | ≤25 | 26-45 | ≥46 | ≤25 | 26-45 | ≥46 |
| Access to information | Wake-Up [m (sd)] | 2.13 (0.74) | 2.04 (0.34) | 2.00 (0.76) | - | 2.23 (0.44) | 1.70 (0.68) |
| | Front door [m (sd)] | 1.63 (0.50) | 2.13 (0.45) | 2.07 (0.48) | - | 2.00 (0.00) | 2.30 (0.68) |
| | Access form [m (sd)] | 2.00 (0.82) | 2.13 (0.81) | 2.18 (0.40) | - | 1.83 (0.83) | 2.20 (0.79) |
| | Emergency [m (sd)] | 2.53 (1.06) | 3.09 (0.95) | 2.46 (1.20) | - | 2.58 (1.16) | 2.55 (1.04) |
| Communication | Telephone [m (sd)] | 1.75 (1.13) | 1.41 (1.28) | 1.33 (1.23) | - | 2.31 (1.11) | 1.67 (1.16) |
| | Written [m (sd)] | 4.63 (1.59) | 3.04 (1.48) | 1.87 (1.19) | - | 3.38 (1.85) | 1.92 (0.90) |
| | Buy food [% help from others] | 0.00 | 55.0 | 57.0 | - | 30.8 | 54.5 |
| | Communicate doctor [% help from others] | 93.7 | 95.8 | 100.0 | - | 15.4 | 41.7 |
| Mobility | Visit shop [% help from others ] | 14.3 | 47.8 | 35.7 | - | 25.0 | 36.4 |
| | Visit doctor [% help from others] | 93.8 | 80.8 | 83.3 | - | 46.2 | 66.7 |

Means (sd) and percentages are shown. "*Access to information*": higher means indicate more help needed or more difficulty to act independently. Only first choice answers are presented. "*Communication*": Mean number of ways in which a telephone or written communication can be used. Higher means indicate more different ways of communication possible or more possibilities to remain independent. In "Buy food" and "Communicate with a doctor" as well as in both items of "*Mobility"* higher percentages indicate more help needed or more difficulty to act independently.

**Effect of age and type of Usher**

Table 4 shows means and percentages calculated for different variables with regard to the type of Usher and age. First we compared Usher type I and type II. Significant differences were found in the subdomains of 'communicate with a doctor' and 'visit a doctor' (Table 5).

Table 5. Maintaining independence, based on the Nordic definition of deafblindness, results of comparing means (Kruskal Wallis) and percentages (Chi square) between groups

| | | Type<br>I – II | Hearing<br>Mild-Severe-Profound | Vision<br>Number of RP-related sight problems (1 to 5) |
|---|---|---|---|---|
| | | Kruskal-Wallis | | |
| Access to information | Wake up | 0.83 | 0.77 | 0.74 |
| | Front door | 0.17 | 0.04 | 0.72 |
| | Access form | 0.70 | 0.02 | 0.43 |
| | Emergency | 0.45 | 0.78 | 0.40 |
| Communication | Telephone use | 0.09 | 0.24 | 0.24 |
| | Written communication | 0.25 | 0.07 | 0.55 |
| | | Chi square | | |
| | Buy food | 0.61 | 0.62 | 0.82 |
| | Communicate doctor | 0.00 | 0.00 | 0.23 |
| Mobility | Visit shop | 0.79 | 0.86 | 0.18 |
| | Visit doctor | 0.01 | 0.15 | 0.17 |

When further investigating where the differences in 'communicating with a doctor' were located between age groups, the following results were found: comparing any age group of Usher type I with any age group of Usher type II showed significant difference ($p=0.00$).

Mobility of Usher patients varies according to the situation. When visiting a shop a large number of both Usher type I and II patients is able to go independently, but visiting a doctor in a hospital requires more help of others for type I patients ($p=0.01$).

Overall high means in the item of 'knowing that an emergency situation occurs at home' (Table 5) indicate that a lot of Usher type I and II patients are not aware of an emergency situation at home.

When we took age into account we saw some trends with ageing. In the domain of access to information the difficulty of 'access to a form' increased with age in both type I and II patients and the item of 'knowing someone is at the front door' only became more difficult for Usher type II patients. In the domain of communication, results of all four items indicate more help needed with ageing in both patients with Usher type I and II. In the mobility domain, type II patients report the need of more help of others when getting older.

In general, Usher type I patients tend to need more help than Usher type II patients and the amount of help needed seems to grow when patients get older.

**Effect of hearing**

Means and percentages were calculated as described before for the three groups of considered hearing impairment: mild, severe and profound.

In the domain of access to information 'waking up' and 'knowing some one is at the front door' independently becomes more difficult, when hearing impairment is considered worse (means 'waking up' 1.92(mild)-1.97(severe)-2.07(profound) and 'front door' 1.75(mild)-2.07(severe)-2.14(profound), where higher means indicate that more help is needed). Significant differences were found for the item 'knowing some one is at the front door' between the three groups ($p=0.05$, Table 5). Significantly different results were also seen in the item of 'access to a form' when comparing mild and severe hearing loss ($p=0.02$) and mild and profound ($p=0.03$). No coherence in answers can be seen in the item 'knowing about an emergency situation at home', though overall high means (>2.59) showed that patients have difficulty finding out about an alarming situation at home. Communication in three subdomains (telephone use, written information and communication with a doctor) becomes more difficult when hearing impairment worsens. The item of 'communication with a doctor' showed a significant inter group difference in the percentage of people who need help of others ($p=0.00$). In the item of 'buying food' no such trend can be seen. In the domain of mobility no significant differences between the different groups of considered hearing loss could be found. Visiting a doctor independently is more difficult than visiting a shop and both mobility items show that more help of others is needed when hearing gets worse.

Overall results indicate that when hearing impairment worsens, patients tend to need more help from others to be able to remain independent.

**Effect of number of RP related sight problems**

As described previously, means and percentages were calculated for the five groups of the number of RP-related sight problems. In all three domains of remaining independent, there is a large variability in the answers to the questions. In the domains of access to information and communication no answering trends can be seen. A lot of difficulties in 'knowing whether an emergency situation occurs at home' were reported in all groups of RP-related sight problems (means >2.20). In the domain of mobility the percentage of Usher patients who need help of others to visit a doctor decreases with the number of RP problems (number of sight problems 1 to 5; help needed 100.0%-90.0%-75.0%- 68.8%-55.6%).

# Discussion

No specific Usher questionnaire has been developed so far. This survey is conducted by applying a descriptive questionnaire with both closed and open-ended questions. We have calculated means and percentages of items to present an overview of aspects and their impact on remaining independent.

Before computation we checked whether our population answered basic questions about their Usher in accordance with what was to be expected according to literature. Hearing is considered worse in type I, followed by type II (and III) (Figure 2). The difference in measured hearing loss has been extensively described [6-9]. When we looked at the change of hearing loss with ageing, we observed an increase in considered hearing impairment in type I and in type II. In type II this has been found in psycho-acoustic measurements as well[10]. The explanation for the observation in type I patients might be that, of all type I patients born with profound deafness, patients who did not consider themselves as profoundly deaf were mainly CI users. Further clarification may be found in the following. In the hearing impaired population the effect of age of onset of deafness and the disadvantages throughout life have been studied. People with long-standing deafness and hearing impairment, especially Usher type I patients, have better learned to live with their situation than patients who acquired hearing loss as a result of the presbyacusis[11].

In our population different types of Usher differed to the extent in which R.P. problems are present. Type I patients reported significantly fewer RP problems than type II patients (Figure 3). Others found no significant differences in various visual signs and symptoms between type I and II[12,13,14] or more specific between genetically based Usher type IB and type IIA[13]. Some reported even more visual impairment in Usher type I than type II[14,15]. If we took age into account, we saw a rough increase in type I and a decrease in type II in number of RP problems when ageing. In the highest age group of type II the mean number of RP problems decreased. Though we did not notice this trend in type I, hypothetically this could be related to the fact that patients get used to living with their sight problems, so that they do not realize that they still have those problems[16]. We have assumed that all RP problems contribute equally to the degree of perceived vision impairment. An earlier study of patients with RP reported that visual acuity correlated significantly with perceived difficulty in everyday tasks. This result is consistent with the results from studies of ageing, which showed that visual acuity is a risk factor predictive of perceived difficulty among older populations. The patients' reports were significantly highly correlated with the reports of the verifiers, and with the assessments by the orientation and mobility specialist[17]. Other studies mention the importance of contrast sensitivity for RP patients in every day life[18, 19].

We addressed three questions with regard to what variables contribute to remaining independent. An overall trend was seen that type I Usher patients encounter more difficulty in preserving their independence than type II patients. The same increase in encountered difficulties was seen with regard to age. Results indicated that worse hearing means more help is needed. Dalton et al. showed that the severity of hearing loss was significantly associated with hearing impairment and with self-reported communication difficulties[20] . Studies have been conducted on communication of deaf people with their specialists. In our study it was obvious that patients with profound hearing loss were hardly ever able to communicate with the doctor by themselves. Other studies showed that deaf and hard-of-hearing persons report substantial communication difficulties with doctors. Results of our study emphasise the fact that deaf and hard-of-hearing persons constitute a minority population that experiences substantial difficulties in the patient-doctor relationship[21,22]. The use of written communication decreased with the increase of hearing loss. A possible explanation why people with a greater hearing loss use written communication less, is that their first language may well be sign language rather than the written and spoken forms of their country's language. People who sign often lack confidence in using written communication.

There seemed to be a reverse trend as when the number of RP problems increased, the amount of help needed to visit a doctor decreased. Szlyk et al. also reported that individuals with severe levels of vision loss could have minimal impairment in carrying out tasks of everyday life. Age was not significantly correlated with any of the functional task categories and is therefore unlikely to be the source of this variability[16]. This raises the hypothesis that vision may not be a major dependent variable for remaining independent or that, in our study, the number of RP problems is not consistent with the severity of vision loss.

Besides in Usher people, age-related vision and hearing impairments are becoming prevalent conditions in the general older population. Dual sensory loss, decreased communication performance and psychosocial functioning impacts on one's quality of life. Hence also rehabilitation services for older adults with age-related sensory loss need to accommodate these difficulties[23].

# Conclusion

People with Usher syndrome meet with particular challenges in order to maintain their independence as their sight and hearing worsen. Usher people need equipment and help from others to remain independent, especially when they get older. Type I people tend to need more help than Usher type II people. According to this study, perceived

hearing loss is a more important variable than impaired vision (based on the number of RP problems) for Usher patients in attempting to remain independent. When considered hearing loss worsens, it is more difficult to remain independent. An important outcome of the survey was the high percentage of Usher people that report their fear about not knowing when an emergency situation occurs at home.
We feel that this study, whilst providing some interesting insights into the lifestyle of people with Usher with the help of the survey, serves to highlight the need for detailed research into the support that Usher people need in order to expand their possibilities of remaining safely independent.

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## Appendix
Usher Lifestyle Survey, Maintaining Independence

### CAUSE Usher Lifestyle Survey Maintaining Independence

1. Your contact details

Family name (in capitals) _____
First name _____
Address _____
_____
_____
Country _____
Phone (Home) _____ (voice / text / mobile)
Phone (Other) _____ (voice / text / mobile)
Fax _____
Email _____

*2.* Your personal and Usher details

2.1 Are you: O Male O Female

2.2 What is your age group? Please tick the correct box
O 16-25 O 26-35 O 36-45 O 46-55 O 56-65 O 66-75 O 75+

2.3 What type of Usher do you have?
• Type 1 (profound deafness and poor balance from birth, RP usually evident by the teenage years) O
• Type 2 (partial hearing from birth, RP may not be diagnosed until adulthood) O
• Type 3 (normal or partial hearing at birth, sometimes poor balance from birth, RP and increased hearing loss often start together in teens or twenties) O
• Unknown O
Any other comments about your Usher?
_____

2.4 Some people with Usher have poor balance from birth. Have you had poor balance from birth?
O Yes O No O Not sure

2.5 Do you have any other long-term disabilities or conditions? O Yes O No
Please explain if you wish:
_____

3. Your hearing

3.1 What do you consider your hearing loss to be? Please tick one box only.
• Mild (You can hear some speech without a hearing aid, with or without lip reading) O
• Severe (You cannot hear speech without a hearing aid and you rely on lip reading as well) O
• Profound (You are too deaf to hear speech, even with a hearing aid. Many profoundly deaf people use sign language) O
• Unknown O

3.2 Do you consider that your hearing is getting worse?
O Yes O No O Not sure

4.Your sight

4.1 Please tick all the sight problems associated with RP below which apply to you
- O Night blindness
- O Tunnel vision
- O Scotomas (patchy vision)
- O Visual disturbances (flashing lights, floaters)
- O Cataracts (not removed)
- O Cataracts (one removed)
- O Cataracts (both removed)
- O Other RP symptoms _____

4.2 Do you have any other eye condition apart from RP?
O Yes O No
If "Yes,", please explain: _____
_____

4.3 Do you wear spectacles or contact lenses?
O Yes O No
If "Yes", do you wear them:
O All the time O Just for reading O Just for distance

4.4 Do you have an official registration scheme for visual impairment in your country?
O Yes O No
If your answer to question 4.4 is Yes, please answer these two questions:
4.4.1 Are you registered?
O Yes O No
4.4.2 What as? _____

5. Your preferred communication methods

5.1 Do you use hearing aid(s)? O Yes O No

If your answer to question 5.1 is "No", please move on to question 5.5.

5.2 How many hearing aids do you use? O 1 O 2

5.3 Is it possible to get hearing aids free in your country? O Yes O No

If you answered "Yes" to this question, do you use free, state provided, hearing aids? O Yes O No
Or did you choose to pay for a hearing aid? O Yes O No

5.4 Do you use a cochlear implant? O Yes O No

5.5 Do you use sign language or tactile communication? O Yes O No

If "Yes", is it: (you may tick more than one)
- The standard sign language for your country O
- Visual frame sign (The standard sign language is done using a small space within the narrow Usher visual field) O
- Hands on sign (The sign language is felt by the hands of the Usher person who cannot now see sign) O
- Manual alphabet (words are spelt onto the hand, often using a variation of the alphabet used with deaf people) O
- Block (the shapes of ordinary written letters are "written" on the hand of the Usher person) O

6. Your current access to communication, information and mobility

6.1 Communication and mobility

6.1.1 Do you have access to the following human aids to communication whenever you need it:
• Sign language or visual frame interpreter
O Yes O No O Not needed
If you need this service, even if you don't receive it:
How many hours support per week do you get? ____ hours
How many hours support per week do you feel you need? ____ hours
• Hand on hand sign language interpreter
O Yes O No O Not needed
If you need this service, even if you don't receive it:
How many hours support per week do you get? ____ hours
How many hours support per week do you feel you need? ____ hours
• Speech to text reporter (A person types out what is said and you read the typing, either from a computer or from a screen on the wall)
O Yes O No O Not needed
If you need this service, even if you don't receive it:
How many hours support per week do you get? ____ hours
How many hours support per week do you feel you need? ____ hours
• Deafblind manual or other tactile communicator
O Yes O No O Not needed
If you need this service, even if you don't receive it:
How many hours support per week do you get? ____ hours
How many hours support per week do you feel you need? ____ hours
• Communicator guide (A person who provides guiding and communication support to enable you to take part in a range of activities)
O Yes O No O Not needed
If you need this service, even if you don't receive it:
How many hours support per week do you get? ____ hours
How many hours support per week do you feel you need? ____ hours

6.1.2 Do you use a guide dog? O Yes O No
Do you use a mobility cane for visually impaired people? O Yes O No
Do you use a mobility cane for deafblind people? O Yes O No

6.1.3 Do you regularly use a telephone? O Yes O No
If yes, which is your preferred type of phone? (You may tick more than one)
O Standard phone O Phone with volume control O Textphone O SMS (text messaging)
O Mobile O Videophone O Other

6.2 Access to written / other information
Do you use (tick all which apply):
O Small print O Large print O Audio tape O Braille (tactile methods)
O Standard print O Video tape O Email O Computer disk

6.3 Equipment and helpful people
Technical equipment, as well as other people, can be of great help to Usher people in their quest to live an independent life. Please describe how you manage the everyday situations listed below. You may use other people, technical equipment or both.

6.3.1 How do you wake up at a set time each morning?
_____
What other help would you like to have?
_____
6.3.2 How do you know when someone is at the front door of your home?
_____
What other help would you like to have?
_____
6.3.3 You need to buy some food at the local shop. What help do you receive in order to a) get to the shop and b) buy the food?

_____
_____
_____

What other help would you like to have?

_____

6.3.4 You have to see the eye specialist. Who will help you to a) get to the hospital and b) communicate with the specialist?

_____
_____

What other help would you like to have?

_____

6.3.5 You have just received a form to fill in which you cannot read because the print is too small. How will you a) access the form and b) fill it in?

_____
_____

What other help would you like to have?

_____

6.3.6 How do you know when there is an emergency in your home, for example, a fire or someone has tried to enter your home?

_____

What other help would you like to have?

_____

6.3.7 What activities would you like to do but can't because you do not have access to the appropriate equipment you need as an Usher person?

_____
_____

6.3.8 Additional equipment

Do you use any of the following (you may tick more than one):

O Magnifier
O Magnifier with light
O CCTV (closed circuit television)
O Extra lighting

6.4 Environment

People with Usher often benefit from good lighting and good colour contrast in their environment. Please answer the questions below about your environment at home and out of doors.

6.4.1 Is the lighting throughout your home suitable for you?

O Yes O No

6.4.2 Do you often bump into furniture, cupboard doors, and articles left on the floor etc in your home?

O Yes O No

6.4.3 Do you often trip over obstacles on the pavements in your street?

O Yes O No

6.4.4 Do you often fall or miss stairs or kerbs?

O Yes O No

6.4.5 When travelling how do you access information about delays or problems?

_____

6.5 Disability Benefits

6.5.1 Do you receive any disability benefits because you have Usher?

O Yes O No

7. Social and emotional support

7.1 Usher people receive support from many different people. Please tick all the boxes below, which apply to you.

I receive support from the following individuals:

O Family

O Friends: O Hearing and sighted
O Deaf
O Hard of hearing
O Blind
O Usher
O Visually impaired
O Hard of hearing and visually impaired

O Volunteers (People who are not necessarily friends and who are not paid to help you)

O Professionals

Please tell us of any difficulties you have in accessing support

_____

7.2.1 Who supports you in your household? (You may tick more than one box):

O Partner
O Parent
O Child(ren)
O Sibling(s)
O Other family member Give relationship _____
O None

7.2.2 Do you feel that any support which your partner or family provides places any stress on the relationships in your family? O Yes O No

Write any comments here: _____

_____

7.3 We all have an identity and that we fit best into certain groups and cultures. Do you consider yourself as:

O Hearing and sighted
O Deaf
O Hard of hearing
O Blind
O Usher
O Visually impaired
O Hard of hearing and visually impaired

8. What do you need to live an independent Usher life?

8.1 How do you know what support and equipment is available for people with Usher?

_____
_____

8.2 What support (equipment, environment and people) works well for you now?

_____
_____

8.3 What extra support do you need?

_____
_____

8.4 Which of the subjects listed below are the most important for you and why? You may tick more than one box.

O Research into Usher
Why? _____
O Research into RP
Why? _____
O Access to communication support
Why? _____
O Disability access / human rights
Why? _____
O Equality: Access to work
Why? _____

O Equality: Education
Why? _____
O Equality: Leisure activities
Why? _____
O Any other important subjects _____
Why? _____
8.5 As a person with Usher, what do you see as the biggest challenges in your life in adapting to change?_____

9. Last questions!
If funding permits, we would like to video some of the Usher people's replies to this questionnaire. The video recording will take place during the CAUSE conference on 27-30 March 2003 at the Hanover Hotel, Hinckley, Leicestershire, UK.
• Are you planning to come to this conference? O Yes O No
• Would you be prepared to take part in the video taping of this questionnaire? O Yes O No

Thank you for taking the time to reply to this survey. You will be sent a copy of the results in Spring 2003.
*We will do our best to produce this document in any print size, in braille, on audiotape and on email.*
Confidentiality
The results of this questionnaire will be used in the preparation of a report. The report will not identify anyone by name, so, unless you have agreed to also take part in the video questionnaire, your name will remain confidential within the CAUSE Project.

2.3

# The parents'perspectives instrument

Evaluation of the Parental Perspective instrument for pediatric cochlear implantation to arrive at a short version.
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# Abstract

*Aim:* Evaluation of the well-known and widely used Parental Perspectives questionnaire (PP) by means of statistical analysis and exploring the possibility to develope a short version, as the instrument is often regarded as being rather lengthy with 74 questions.

*Methods:* 130 parents of children participated in this study. To assess internal consistency among the PP items of the domains, Cronbach's alpha coefficients were calculated. Corrected item-total correlations were carried out to investigate the strength of individual items' associations with the construct. Factor analysis was performed to identify the statistical factors of the original PP and in exploring a revised short form PP.

*Results:* Instead of the expected eight-factor structure (eight suggested domains), factor analyses found a 15-factor structure. Nevertheless, when the proposed eight-domain structure is followed, some items can be disposed of, based upon the Cronbach's alpha analyses and consistent reasoning. After reducing the number of factors based on standard criteria, a three-domain structure was shown as main concept. The cumulative variance explained by this new model was 39.4% and the final number of items in this probable revised version is 23. Reliability analyses of the new domains of the proposed short version PP (sPP) showed good internal consistency (Cronbach's alpha 0.79). The corrected item-total correlations represent strong individual items' associations with the construct as R items-total varies between 0.34 and 0.64.

*Conclusions:* The Parental Perspectives instrument (PP) is an important tool to assess the impact of cochlear implantation of a child for the quality of life for the family and the child itself. This statistical investigation showed a possible option for development of a short form usable in prospective follow-up studies.

# Introduction

A range of studies has confirmed the effectiveness of cochlear implantation in children from a professional point of view, with emphasis on speech perception and speech production[1-4]. Nevertheless, it was found that solely clinical tests of speech and hearing might not adequately reflect the child's performance at home or school after cochlear implantation[5]. It is generally believed that the parental view has the potential to add significant value to the evaluations gained by professionals, as formal objective measures may not reflect the child's functioning in everyday situations. Parents are often the ones who make the decision for a cochlear implant (CI) for the child and it could help them to know more about the views of other parents who already have experience with cochlear implants. Furthermore, parents whose children received implants could offer comments of great value to implant teams and policy makers. They can provide valuable information about the child's functioning, the process of implantation, the additional interventions needed and the benefits and limitations experienced. An instrument to assess parents' views can be used across different children's age levels and can rely on the perspective of people profoundly interested in the process and outcomes of implants. It must be recognized though, that any perspective of outcomes is limited and should not be used as single source of information[6].

For those reasons, Archbold et al. developed a parent outcome instrument, originally titled "Parents' views and experiences with pediatric cochlear implant"[7]. The items (formal term for the questions) of the instrument were based upon responses to open ended questions in interviews with parents of CI children. The initial 107 items were reduced to 74 multiple-choice questions. In previous work, the authors collected empirical data that showed that the instrument is robust and repeatable[8]. Reliability and validity of the instrument were addressed by Nunes et al[6,9]. The instrument is now commonly referred to as Parental Perspectives (PP).

The importance of this instrument seems generally accepted, with numerous implant centres in for example the United Kingdom, Belgium and the Netherlands, using the instrument. An internet version of the PP including a software package for the PP is worldwide accessible on the website from the UK Ear Foundation[10]. As both previous investigating studies did not have large numbers of participants, validation from other centres is wanted. The complex nature of computing the measurement properties of an instrument implies caution of interpretation of results in literature. This complexity made us realise the need of a general statistical structure analyses with a large numbers of participating parents.

In the current study, the main question relates to evaluation and validation of the PP by means of statistical analysis. Additionally, as the present PP is often considered as

rather lengthy for workable practice[6], the possibility of developing a short version was explored.

# Materials and methods

**Patients**

Data for this study were derived from a project on quality of life in children who received a CI at the Radboud University Medical Centre Nijmegen. All children were implanted and rehabilitated consecutively by the Nijmegen-Viataal Cochlear Implant Team and received their multichannel implant at least one year before participation in the study. Audiologic criteria for implantation in the Netherlands are profound hearing loss (pure tone average > 110dB, speech perception results < 25%) and no functional residual hearing. Besides those criteria, also psychological factors (of child and caretakers) are taken into account. Children with a substantial residual hearing are placed on a waiting list so they can be implanted when needed. The PP (amongst other questionnaires) was sent to the parents of 175 children. In total, 130 parents of children between 1.9 and 18.2 years of age (mean 8.9) participated in this study by returning the filled in instruments and signing an informed consent form. Age at implantation of deafness varied from 0.7 till 14.3 years of age (mean 4.6) and the duration of deafness was 0.2 till 14.3 years (mean 3.8). Aetiology of deafness was variable with meningitis, unknown aetiology and hereditary disease as main aetiologic factors (respectively 33%, 19% and 8%). The non-responders had a mean age of 12.2 years (range 4.1-18.5), age of implantation varied from 1.3 till 15.6 years of age (mean 5.6) and the duration of deafness ranged from 0.3 till 15.5 years of age (mean 4.3), being fairly similar to the participant demographics. Demographic characteristics are shown in Table 1. The present heterogeneous group made it possible to analyse the instrument without measurement of inclined answer patterns.

**Instrument**

The PP instrument was developed by Archbold et al. and is based on themes that were regarded relevant by parents in open interviews[7]. The PP instrument covers ten themes or domains, identified as significant on the basis of the interviews with parents: namely communication, general functioning, self-reliance, well-being and happiness, social relationships, process of implantation, education, effects of implantation, decision to implant, and supporting the child. In order to avoid a halo effect in the responses to items referring to the same domain, the items were placed in random order for presentation to the parents. The PP instrument is constructed as a closed-format instrument consisting of a series of statements with which the respondent

could 'strongly agree', 'agree', 'neither agree nor disagree', 'disagree', or 'strongly disagree'. Responses on these five categories were coded in ascending order 1 - 5. A failure to respond was classified as a missing value. Some questions were phrased in opposite form (negatively stated). As two domains of the PP, 'process of implantation' and 'decision to implant' contain qualitative items, which are impossible to be numerically sensibly coded, these domains were left out of further analyses. Those domains were also excluded in the software package by the Ear Foundation[10]. A validated Dutch translation of the instrument was obtained from ONICI, Zonhoven, Belgium.

Table 1. Demographic characteristics

| | | Percentage | |
|---|---|---|---|
| | | Participants | Non-responders |
| Onset of deafness | Congenital | 59% | 52% |
| | Prelingual | 33% | 32% |
| | Postlingual | 8% | 16% |
| Gender | Male | 47% | 32% |
| | Female | 53% | 68% |
| Use of CI per day (hours) | < 6 | 3% | |
| | 6-12 | 39% | |
| | >12 | 58% | |
| Living situation | Alone | 2% | |
| | With family | 96% | |
| | Institute | 2% | |
| Education | Special | 55% | |
| | Regular | 45% | |
| Communication School | Oral | 59% | |
| | Sign | 15% | |
| | Oral supported by sign | 26% | |
| Communication at home | Oral | 57% | |
| | Sign | 9% | |
| | Oral supported by sign | 34% | |

**Data analyses**

To assess internal consistency among items of the domains Cronbach's alpha coefficients were calculated. The higher the alpha, the stronger the internal consistency reliability, indicating that all items are measuring aspects of the same underlying construct (domain). A Cronbach's alpha of more than 0.70 indicate good reliability[11]. Corrected item-total correlations were carried out to investigate the strength of individual items' associations with the construct. Factor analysis (unweighted least squares) was performed to identify the underlying factors (i.e. similar to domains, but

here derived based on statistical analysis) of the original PP and in the exploration of a revised short form PP. The number of factors to be retained was determined based on screeplot and eigenvalues > 1. Subsequently, a factor analysis with varimax rotations was performed. These analyses were exploratory in order to reveal whether items supposed to belong to the same domain, grouped coherently. Only items with a factorial weight of > 0.4 were regarded meaningful. It should be noted that in using the PP after implantation, some items regarding the situation before implantation become relatively less relevant. Based upon consistent reasoning (identifying ambiguous items or items that refer to the period before implantation, as they can be currently regarded less relevant in an instrument filled in after implantation and in a short form to be used in prospective follow-up studies) some items could be excluded in a short version. The questionnaire contained 74 items in total, 48 quantitative items and 26 qualitative items (could not be included in final analyses). The statistical data evaluation was performed using the SPSS Version 12.0 program package.

# Results

The descriptive statistics of each item of the eight quantitative domains of the original PP instrument and their reliability parameters are given in Table 2. Most domains of the PP instrument did not reach the critical Cronbach's alpha of > 0.7, indicating that the subdivision of the items in the proposed domains is not optimal.

The results of the factor analyses showed that instead of an eight-factor structure (eight suggested domains based on theoretical considerations), a 15-factor structure was found. The cumulative variance explained by this 15-factor model is 66.5%, with main loadings on the first two factors (total variance 31.1%). Even though two principal factors within the total of fifteen factors with eigenvalue > 1 were established, no clear structure of the PP instrument could be detected, nor after rotation. In addition, a factor plot (after varimax rotation) of all items of the parental perspective instrument showed no apparent division of the factors.

These results suggest that the PP instrument does not have the anticipated structure in eight domains. When investigating the possibility of a short form based upon the previously mentioned statistical analyses, some items could be left out of the instrument. At this point, when the proposed division of items into the eight domains is followed, some items would have to be disposed of, solely based upon the Cronbach's alpha criterion. This concerns the following eleven items: 5, 12, 15, 19, 20, 30, 59, 61, 64, 65 and 67. Furthermore, consistent reasoning (see materials and methods) results in exclusion of the following 12 items: 3, 4, 8, 26, 31, 33, 45, 48, 53, 62, 73, 74, making the total number of excluded items 23.

Table 2. Descriptive statistics and reliability analyses on the eight quantitative domains of the original parental perspective instrument.

| Domain | Item no. | Mean | SD | Cronbach α total | Cronbach α if deleted | R item-total |
|---|---|---|---|---|---|---|
| Communication | 1 | 3.81 | 1.18 | 0.86 | 0.86 | 0.56 |
| | 18 | 3.87 | 1.20 | | 0.85 | 0.61 |
| | 27 | 3.67 | 1.24 | | 0.85 | 0.64 |
| | 66 | 3.90 | 1.20 | | 0.85 | 0.61 |
| | 71 | 4.27 | 1.06 | | 0.82 | 0.81 |
| | 72 | 4.10 | 0.99 | | 0.83 | 0.75 |
| General Functioning | 4 | 1.98 | 1.32 | 0.46 | 0.55 | 0.04 |
| | 6 | 3.42 | 1.26 | | 0.42 | 0.23 |
| | 7 | 4.46 | 0.75 | | 0.40 | 0.29 |
| | 35 | 3.94 | 0.93 | | 0.33 | 0.40 |
| | 51 | 3.70 | 0.96 | | 0.33 | 0.40 |
| | 53 | 3.84 | 0.95 | | 0.45 | 0.15 |
| Self Reliance | 12 | 4.13 | 0.95 | 0.24 | -0.09 | 0.32 |
| | 32 | 3.65 | 0.79 | | 0.44 | -0.14 |
| | 33 | 2.48 | 0.97 | | 0.35 | -0.00 |
| | 47 | 3.51 | 1.16 | | -0.23 | 0.35 |
| Wellbeing Happiness | 16 | 3.37 | 1.02 | -0.08 | -0.03 | -0.05 |
| | 59 | 3.33 | 0.89 | | -0.28 | 0.11 |
| | 61 | 3.41 | 0.91 | | 0.23 | -0.24 |
| | 64 | 3.17 | 0.90 | | -0.13 | 0.02 |
| | 70 | 4.60 | 0.57 | | -0.18 | 0.10 |
| Social Relations | 5 | 4.24 | 1.07 | 0.27 | 0.22 | 0.13 |
| | 30 | 3.33 | 1.24 | | 0.31 | 0.02 |
| | 39 | 3.82 | 1.07 | | 0.02 | 0.38 |
| | 41 | 4.38 | 0.73 | | 0.17 | 0.25 |
| | 45 | 3.24 | 1.08 | | 0.42 | -0.16 |
| | 65 | 4.07 | 0.91 | | 0.13 | 0.28 |
| | 74 | 3.48 | 0.84 | | 0.30 | -0.00 |
| Education | 9 | 3.51 | 1.37 | 0.58 | 0.44 | 0.51 |
| | 23 | 4.10 | 0.73 | | 0.54 | 0.35 |
| | 31 | 4.03 | 0.71 | | 0.58 | 0.18 |
| | 37 | 3.08 | 1.19 | | 0.53 | 0.34 |
| | 48 | 3.66 | 1.10 | | 0.63 | 0.07 |
| | 50 | 4.16 | 0.85 | | 0.48 | 0.52 |
| | 69 | 3.56 | 1.17 | | 0.57 | 0.23 |
| Effects of Implantation | 2 | 4.06 | 0.98 | 0.59 | 0.55 | 0.29 |
| | 8 | 2.81 | 1.07 | | 0.60 | 0.17 |
| | 11 | 3.02 | 1.32 | | 0.45 | 0.51 |
| | 14 | 4.47 | 0.73 | | 0.51 | 0.49 |
| | 17 | 4.00 | 0.93 | | 0.52 | 0.39 |
| | 26 | 3.94 | 1.18 | | 0.61 | 0.14 |
| | 73 | 3.55 | 1.09 | | 0.57 | 0.25 |
| Support the child | 3 | 3.53 | 1.07 | 0.36 | 0.43 | 0.01 |
| | 15 | 4.13 | 0.85 | | 0.20 | 0.35 |
| | 19 | 2.88 | 1.06 | | 0.28 | 0.21 |
| | 20 | 3.09 | 0.95 | | 0.24 | 0.26 |
| | 62 | 1.58 | 0.65 | | 0.46 | -0.16 |
| | 67 | 3.58 | 1.03 | | 0.21 | 0.30 |

- Number of answered items ranges from minimum 90 (social relations) to maximum 116 (self reliance)
- Items numbers phrased negatively and hence recoded: 1, 18, 4, 53, 12, 33, 16, 61, 5, 30, 39, 9, 37, 2, 8, 11, 14, 17, 73, 15, 62, 67, 10, 22, 29, 55, 63, 28, 57
- Possibly afterwards deleted items based on Cronbach α and logics: 4, 53, 12, 33, 59, 61, 64, 5, 30, 45, 65, 74, 31, 48, 8, 26, 73, 3, 15, 19, 20, 62, 67

The factor analyses on the revised PP showed seven principal domains with an eigenvalue > 1. However, for several of these factors, less than three variables were loaded with a factor loading > 0.4. After further reducing the number of factors, the three-domain structure (eigenvalue > 1) showed equally spread of item loadings per domain. Hence, the three-domain structure was chosen as main concept. The cumulative variance explained by this new model was 39.4%. The rotated factor matrix showed equally divided item numbers over the three factors (Table 3). As can be seen in Table 3, the item numbers 32 and 51 have factor loadings < 0.4 and were therefore excluded. This makes the total number of excluded items 25. As there were 48 quantitative items in the original instrument, the final number of items in this revised version is 23.

Table 3. Rotated Factor Matrix (Varimax with Kaiser Normalization) from unweighted least squares factor analyses of items of the revised parental perspective instrument.

| Item no. | Factor | | |
|---|---|---|---|
| | 1 | 2 | 3 |
| 1 | | | 0.54 |
| 2 | | | 0.52 |
| 6 | 0.53 | | |
| 7 | 0.44 | | |
| 9 | | 0.53 | |
| 11 | | | 0.47 |
| 14 | | | 0.56 |
| 16 | | 0.54 | |
| 17 | | | 0.53 |
| 18 | | | 0.52 |
| 23 | | 0.53 | |
| 27 | 0.69 | | |
| 32* | | | |
| 35 | 0.59 | | |
| 37 | | 0.45 | |
| 39 | | 0.55 | |
| 41 | | 0.56 | |
| 47 | | 0.58 | |
| 50 | | 0.54 | |
| 51* | | | |
| 66 | 0.59 | | |
| 69 | 0.64 | | |
| 70 | | 0.41 | |
| 71 | 0.62 | | |
| 72 | | | 0.53 |

*Item 32 and 51 were excluded

Reliability analyses of the new domains of the proposed short version PP (sPP) are shown in Table 4. All three Cronbach's alpha were 0.79, indicating good reliability of the three factors. The corrected item-total correlations represent rather strong individual items' associations with the construct as R items-total varies between 0.34 and 0.64. Analyses of the items of the three factors suggest three specific domains: General Communicative functioning (seven items), Social and Educational functioning (nine items) and Effects of Implantation (seven items).

Table 4. Reliability analyses on three new domains of the revised Parental Perspective instrument.

| Domain short<br>Parental Perspectives | Item no. | Mean | SD | Cronbach α total | Cronbach α if deleted | R item-total |
|---|---|---|---|---|---|---|
| General Communicative functioning | 6 | 3.35 | 1.28 | 0.79 | 0.80 | 0.34 |
| | 7 | 4.47 | 0.74 | | 0.77 | 0.50 |
| | 27 | 3.61 | 1.24 | | 0.75 | 0.60 |
| | 35 | 3.97 | 0.90 | | 0.77 | 0.50 |
| | 66 | 3.88 | 1.15 | | 0.74 | 0.64 |
| | 69 | 3.56 | 1.17 | | 0.76 | 0.53 |
| | 71 | 4.26 | 1.08 | | 0.75 | 0.59 |
| Social and Educational functioning | 9 | 3.52 | 1.37 | 0.79 | 0.77 | 0.48 |
| | 16 | 3.31 | 1.15 | | 0.77 | 0.45 |
| | 23 | 4.11 | 0.73 | | 0.76 | 0.52 |
| | 37 | 3.09 | 1.20 | | 0.77 | 0.46 |
| | 39 | 3.78 | 1.10 | | 0.76 | 0.47 |
| | 41 | 4.30 | 0.79 | | 0.76 | 0.50 |
| | 47 | 3.58 | 1.13 | | 0.75 | 0.57 |
| | 50 | 4.16 | 0.85 | | 0.76 | 0.52 |
| | 70 | 4.60 | 0.58 | | 0.78 | 0.42 |
| Effects of Implantation | 1 | 3.83 | 1.17 | 0.79 | 0.73 | 0.64 |
| | 2 | 4.05 | 0.99 | | 0.78 | 0.40 |
| | 11 | 3.00 | 1.29 | | 0.76 | 0.54 |
| | 14 | 4.45 | 0.71 | | 0.77 | 0.48 |
| | 17 | 4.03 | 0.92 | | 0.77 | 0.42 |
| | 18 | 3.87 | 1.17 | | 0.75 | 0.57 |
| | 72 | 4.11 | 0.97 | | 0.75 | 0.57 |

- Number of answered items ranges from minimum 110 (effects of implantation) to maximum 117 (general communicative functioning)
- Items numbers phrased negatively and hence recoded: 1,2, 9, 11, 14, 16, 17, 18, 37, 39

# Discussion

The aim of our study was to evaluate the Parental Perspectives instrument for CI children. The results from statistical analyses to derive the underlying structure of the PP instrument indicated the absence of the anticipated structure in eight domains. The investigation of the possibility of a reliable and valid short version of the PP, based upon the original domain structure of the instrument showed a possible option for a short form for example usable in prospective follow-up studies.

The original instrument, developed and validated by Archbold et al.[7], has been extensively studied by various research institutes. The first to examine the PP, at that time called the "Children with cochlear implants: parental perspectives", were Dujardin et al.[12]. A total of 68 respondents, response rate of 63%, were included in their study. The length of the instrument was regarded the most important reason for the low response rate. This was one of the reasons why the authors suggested an exploration of a short form PP. It was concluded, based upon factor analyses, that the original structure of ten domains could not be detected. Reliability analyses did only show good reliability for the domain of communication, which is in concordance with our current results. Dujardin et al. stated that the low reliability scores in the other domains could be due to the relatively small number of respondents. Their final advice was to further investigate this instrument[12] Unfortunately, Dujardin et al. did not report their exact factor loadings and Cronbach's alpha's, so independent comparison with our current results was not possible. Their study emphasizes, on the other hand, the importance of a large number of participants. To date, we have, with 130 respondents, the largest number of PP instrument responses used in analyses.

O'Neill et al. have investigated the reliability of the PP[8]. Parents of 20 congenitally or prelingually deaf with at least two years of CI experience participated in their study. Parents filled in the instrument twice with a one month interval. Responses over time were compared. Test-retest reliability appeared to be > 95% for most instrument items. This high test–retest reliability indicated the capacity of elicit parental views in a meaningful manner. They suggested that an extensive use of the instrument could result in valid comparisons of outcomes among different cochlear implant centres and they highlighted the need for an acceptable, valid and reliable instrument for parental perspectives in cochlear implantation[8]. However, a limited number of participants was used.

Nunes et al. investigated the reliability and validity of the instrument, at that time called "Parents views and experiences with pediatric cochlear implant questionnaire" (PVECIQ)[6,9]. Parents of 61 children with at least three years of CI experience responded and used the instrument and were engaged in an interview. Based upon the interview with parents an extra domain, "functioning before the implantation" was added and the domains were redefined (different items within each domain) and 23

items were excluded. The Cronbach's alpha reliability of the new 11 domains varied between 0.41 (Process of implantation) and 0.74 (Education). Relatively good content validity was assessed by comparison with parents' responses to an interview. A factor analysis of the total instrument scales identified four components. To some extent, this analysis is uncommon as the individual items are not analysed and therefore the loading of the divers items on a specific domain was not determined. It was concluded that the PVECIQ could be used to describe how pediatric cochlear implants affect the children's lives according to their parents' perceptions and that it could fulfil the aim of making it possible to obtain data relatively quickly and in quantifiable ways. They did realize that some scales produced more robust and valid results than others.

As different methodological approaches were used in the studies mentioned above, it is hard to directly link the former studies to the current investigation. Nevertheless, an overview of the diverse studies is important in understanding the involvedness of the PP questionnaire.

The need for a parental instrument to determine the impact of CI on the family and QoL of the child was previously investigated by only few others as most reports focussed on speech perception. The Parenting Stress index has been used to evaluate the stress of parenting children with CI compared with children with hearing aids (and with normal hearing children)[13]. Two studies used an open structured assessment as method to address parental stress in cochlear implantation. An interview-based study was performed by Quittner et al. to determine the effect of cochlear implantation on parental stress [14] and Kelsay et al. used an open format instrument to ask parents about benefits and problems after cochlear implantation[15]. These studies described interesting outcomes of cochlear implantation regarded by parents, but results could not be used in further analyses or could not be compared with other results, as the methods used, were neither universal nor quantitative. This emphasizes the need for a short, generally accepted, reliable and validated instrument.

The importance of the PP questionnaire seems, based upon the large numbers of users and extensive research conducted previously, inevitably clear. In an attempt to analyse an existing instrument, it is impossible to find the one sole correct way to do so. In the current report, an analytical methodology was applied, contemplating the use of this instrument for prospective follow-up. The methodology applied at present is a proposition and is not claimed to be the answer to every measurement property about this instrument. In this study the PP instrument was analysed from a psychometric point of view, nonetheless a more clinimetric (qualitative, clinical) approach would also have been possible. A psychometric point of view emphasis the statistical characteristics and quantitative outcomes of an instrument, whereas a clinimetric point of view tries to find as much information as possible by means of a descriptive narrative approach. These principles of clinimetrics and psychometrics were first described by Feinstein in 1987[16].

The possibility that differing populations may change the analyses, may be only partly true as minor differences between best and worst performing CI children may be reflected within the PP results. It should be noted that an instrument such as the PP is developed for a specific population (CI children) with a specific application (parent directed retrospective investigation) and the performance range should have been taken into account at the time of its development. Therefore, it is not realistic to assume great variance in instrument analyses based solely upon different populations CI children.
At last, it is realised that reducing the PP questionnaire length is attractive but will inevitably result in loss of information. In the present paper, a short version, based upon the original PP, was analyzed. This resulted in exclusion of a number of, according to some parents and caretakers possibly useful items, that might have still been included when another instrument structure would have been used. The revision of the proposed eight-domain structure could be investigated in future in close collaboration with the developers of the instrument. Moreover, in order to be able to test the validity of the instrument (and to determine whether another structure is more valid) in comparison with other instruments, supplementary research needs to be done.

# Conclusion

The Parental Perspectives instrument (PP) is an important tool to assess the impact of CI of a child for the quality of life for the family and the child itself. The originally named "Parents' views and experiences with pediatric cochlear implant" has been analysed before, but no conclusive results based on purely statistical analyses, were reported so far. By means of statistical analyses, the original PP instrument was currently evaluated. Most domains of the PP instrument did not reach the critical Cronbach's alpha of $> 0.7$ and no clear structure of the PP instrument could be detected. These results indicate that the PP instrument does not have the anticipated structure in eight domains. Moreover, as the PP, containing 74 items, is regarded rather lengthy, the possibility of a reliable and valid short version of the PP, was explored. This investigation showed a possible option for development of a short form usable in prospective follow-up studies. It should be realised that reducing the questionnaire length is attractive but will inevitably result in loss of possible valuable information.

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# General discussion

# General discussion

When investigating cochlear implantation and quality of life assessment, some questions arose. This discussion is built upon those questions.

I. The first dilemma to be addressed, before even mentioning quality of life in CI patients, is whether or not to implant in the first place. This still is a very realistic and actual dilemma for deaf adults and parents of deaf children. How is the perceived quality of life in deaf people? How do they feel about their quality of life? And how do they feel about cochlear implantation? In order to answer these questions, we need to get some insight into the Deaf culture and the self-perception of Deaf people.

II. Furthermore, after having chosen for cochlear implantation, it is important to realise that there are numerous ways to assess quality of life in general and in CI patients. What are the different ways to study quality of life? What are the advantages and disadvantages for one method over another? By what factors is quality of life influenced? And how are we supposed to investigate the diverse questionnaires used in measuring quality of life in an objective way? After all, nothing seems more subjective than quality of life!

III. Finally, the quality of life of CI patients has been studied by quite a lot of research groups and implant teams. What different results were overall mentioned in literature? And how do those studies relate to each other and most importantly to this thesis?

*I. The Deaf, the Hearing World and Cochlear Implantation*

It should be noted that the existence and fulfilment of minorities – ethnic, racial, religious, linguistic or other – is a fundamental milestone of peaceful democratic societies. The Deaf Society is one such minority. Studying the quality of life in deaf people incorporates certain difficulties: the questions have to be translated properly into sign language or have to be in written format questionnaires, which can be a problem, as not all deaf people are able to read accurately. Those dilemmas were overcome in a study of Fellinger et al. in 2005. They created an interactive computer based assessment package to assess mental distress and quality of life in deaf people[1]. It turned out that the mental distress correlated very well with the reported quality of life in a negative way. This means that the more distress is mentioned, the worse the reported quality of life is. The Deaf population showed a significant higher rate of distress than the general population, which is in accordance with previous studies. They assumed that the increase in mental distress could probably be explained by a lack of communication in childhood (which can lead to emotional problems), higher levels of common mental disorders in adult life and the fact that sometimes etiology of deafness can be associated with cerebral pathology (e.g. rubella, meningitis)[2].

Deaf adults are autonomous people that are able to give informed consent for cochlear implantation. However, some controversy surrounds its use in deaf children. In his book "The Mask of Benevolence – Disabling the Deaf Community", Harlan Lane speaks of genocide and ethnocide believing that auditory (re)habilitation, for example by means of a CI, would, in the end, overwhelm and destroy the Deaf culture[3]. Also the National Association of the Deaf (NAD) first disapproved of cochlear implantation in children. Their position paper on cochlear implants in children (1993), now withdrawn, stated that "The NAD deplores the decision of the Food and Drug Administration (to approve the commercial distribution of cochlear implants for surgical implantation of children aged two through seventeen) which was unsound scientifically, procedurally and ethically" and that "far more serious is the ethical issue raised through decisions to undertake invasive surgery upon defenseless children, when the long-term physical, emotional and social impacts on children from this irreversible procedure – which will alter the lives of these children – have not been scientifically established"[4]. The former clearly emphasizes the importance of quality of life research in this area.

It follows that deaf children's civil rights might be at stake and that legal authorities could overcome parental custody and enforce a medical treatment – cochlear implantation and auditory (re)habilitation – if it is regarded as being in deaf children's best interests. When a person has never been competent – as a deaf child – Engelhardt's contention is that "guardians may be in authority to choose particular understandings of an individual's best interests in terms of the values embraced by the community within which the ward lives and to which, it can often be presumed, the ward will or would subscribe"[5]. Most deaf children are not born into the Deaf-World and therefore the standard of best interest should be to choose as a reasonable and prudent person would choose and it might be added as a reasonable and prudent hearing person would choose. Parents are the legitimate surrogate decision-makers on behalf of their deaf children. Particular moral communities should respect parental rights to make decisions on behalf of their wards. It is generally accepted, as Dena Davis argues that "the autonomy of the individual is ethically prior to the autonomy of the group"[6].

The debate about the personal decision for cochlear implantation was characterized by Enerstvedt by the following quote: "Imagine a culture in which deaf people were considered holy, selected by the gods; the cochlear implant would surely not have been invented there".[7]

What is it, then, about Western cultures and their views of deafness that make medical solutions so appealing? Public attention to medical technologies is still rooted in a common and persistent concept of deafness as a disability. The availability of such technologies as cochlear implantation, can have unintended negative effects on families with deaf children. That is why it is important that parents should receive

comprehensive, detailed, and objective (i.e. neutral) information in order to make a decision[8].

Several books, articles and reports have been written on the ethical topic of cochlear implantation. It is far beyond the scope of this discussion to give an overview of those investigations. A different and inspiring way to approach the dilemma of cochlear implantation, was described by Gonsoulin: 'Deaf World representatives have defined the profoundly deaf as a separate culture from the mainstream hearing society. A fundamental premise is that deafness is not a disability. The cochlear implant community has challenged this perspective, maintaining that deafness is a disability. Efforts to reduce deafness are regarded as ethical, consistent with the beneficence principle. The Deaf World challenged the performance of cochlear implants in children as a matter of ethical debate. The first challenge is that of diminution of a minority culture. In heated rhetoric, this diminution is labelled genocide or ethnocide. On the other hand, the cochlear implant community contends that deafness is a disability. In order to look at how the opposing groups differ in their perspective, an analysis of bottom elephants is helpful. As described by Bernard Loomer, a theologian, the term bottom elephant referred to the fable that asked the question: what holds up the world? Answer: an elephant. What holds up the elephant? Another elephant. The question keeps getting asked until we come to a bottom elephant, the one that holds up all of the rest of the elephants. In an unpublished manuscript from 1970, his thesis said "that every system of thought (theological, philosophical, economic, political, or scientific), to the extent to which it is a system, rests on an elephant." The elephant consists of a core of perhaps two or three foundational ideas, concepts, assumptions. These are ultimate with respect to that system of thought. One cannot go beyond them for more basic meaning. Their validity is unquestioned. In the Deaf World, deafness constitutes the major part of the bottom elephant. It is more than a physiologic phenomenon. It constitutes fundamental identity. 'If the essence of my being, deafness, is a disability, what does that say about me?' With regard to the mainstream hearing world, particularly the cochlear implant community, a major part of the bottom elephant is the ethical principle of beneficence. Knowing and understanding the two bottoms, could help in discussing the cochlear implant dilemma, as was previously described in the introduction"[9].

*II. Quality of Life Measurement*

The development of quality of life measures in health has been encouraged both by the need to assess the relative merits of rival health programmes in a context of increasing pressure on health resources, and by a desire to be able to comprehensively assess the impact of clinical therapies. With respect to the latter, Revicki noted that advances in medical research and therapy have shifted health care resources from the diagnosis and treatment of infectious disease to the prevention and control of chronic

disease: with this has come an increased emphasis on changes in functional status and quality of life outcomes[10]. Deyo and Patrick have also pointed out that medical interventions may result in improved functional health status without evidence of physiologic improvement [11].

Health-related quality of life represents those domains of overall quality of life that are affected by one's health[12]. Instruments that measure (health-related) quality of life can be divided into different categories. One possible division is health surveys (for example the SF36) and health utility instruments (for example the HUI3). Health index surveys comprise batteries of items organized into scales, each of which measures a dimension of health-related quality of life. With health indices, scores are compiled across questions querying each different domain, and a single summary score or domain-specific score can be reported. These metrics provide a rich source of information for approaching clinical questions or understanding the health status of patients. Health utility metrics focus on a subject's broader conceptualization of health-related quality of life through approaches that differ from health index surveys. In trying to capture a patient's overall sense of well-being, patients are asked to assign a value to their current health state, ranging from 0 to 1 Although health utility metrics do not provide detailed clinical information, they are useful for assessing the cost-utility of medical treatments [13, 14].

With respect to paediatric cochlear implantation, two major issues arise. Firstly, what the salient domains of health-related quality of life would be for a child and secondly it should be noted that (health-related) quality of life measures in children are often completed by parental-proxy rather than by the child. Previous studies have demonstrated poor correlation between parent and child scores for mental- and social-related domains of health-related quality of life and better correlation for physical-related domains[15]. The interpretation of health-related quality of life results must, therefore, take into consideration the choice of survey-respondent, and when possible, both parent and child-reported scores should be used. An example of such an instrument is the PedsQL, used in this thesis.

A large number of measures of health status and associated notions are available[16]. Another specific way to characterise the instrument is, instead of health survey and health utility instrument, is in terms of disease specific versus generic measures[17, 18]. In this thesis both disease specific and generic instruments were used. Measures vary in the degree to which they are developed to measure a specific disease or to be capable of application to many or all illness states. Disease specific measures have greater salience for physicians, better focus on functional areas of particular concern, and may possess greater responsiveness to disease-specific interventions[11]. On the other hand generic measures permit comparisons across interventions and diagnostic conditions, which can be important for policy makers. They also allow dysfunction to be

quantified for an individual experiencing several disease conditions [19, 20]. The use of generic measures has several advantages, including the reduced need for developing and testing different instruments for all patient groups separately, and uniformity of measurement in rehabilitation facilities. An important advantage is that, when using generic measures, the burden of different diseases and disabilities can be compared among patient groups and, in some cases, with the healthy population[21]. In other words, generic measures intend to measure the same construct (activity limitations or participation restrictions) across different patient groups, while specific measures are developed for application in one diagnostic group only.

As both specific and generic measures have vast advantages, it is plausible to use both types of instruments. Measurement instruments, whether generic or disease specific, usually consist of one or more subscales (domains), where items are summed up to form a total subscale score. It is important that the subscales measure one clearly defined underlying construct, preferably based on the domains of the International Classification of Functioning, Disability and Health (WHO International classification of functioning, disability and health 2001). All items of the same (sub)scale are supposed to measure one construct, and should therefore be related to the construct that is intended to be measured[21] (also see measurement statistics).

*Measurement Statistics*

Measurement in rehabilitation research typically concerns a group of patients. The measurement of functioning and disability (quality of life) of such a group generally has one of three aims: diagnosis, prognosis or evaluation. In evaluation (most important in this thesis), the aim of measurement is to evaluate changes in functioning and disability over time. Traditional methods for evaluating reproducibility, validity and feasibility have been supplemented with methods to evaluate responsiveness and interpretability, thereby extending the evaluation of the quality of measurement instruments[21]. Descriptive information on the instruments includes the goal of measurement, the nature of the measurement instrument (e.g. questionnaire, rating of performance, measurement of physical properties such as force or pressure), the specific populations for which the instrument was developed, the format of the measurement instrument (e.g. number of items, response options, minimum and maximum score) and issues related to feasibility (e.g. time needed to perform the measurement, required equipment and training). Recently, a checklist has been developed which facilitates the systematic evaluation of clinimetric properties of measurement instruments[22]. This checklist focuses on questionnaires and contains items on validity, reproducibility, responsiveness, interpretability and feasibility (practical burden). The concept of validity refers to the degree to which an instrument measures what it is supposed to measure. Reproducibility is the extent to which an

instrument is free of measurement error. The checklist focuses on test-retest reliability and agreement. Responsiveness refers to an instrument's ability to detect important change over time in the concept being measured. Responsiveness can be conceptualized as longitudinal validity: does the instrument measure changes in the concept that it is supposed to measure? Concerning feasibility, the checklist focuses on time required for administration and ease of scoring. Interpretability can be defined as the degree to which one can assign meaning to quantitative scores: information is required on the clinical meaning of scores and on which difference between scores can be regarded as clinically meaningful.

In order to be able to calculate sum scores from the items, the dimensions of the measurement instrument have to be consistent across groups. It should therefore be tested whether the items correlate with the same subscale scores (i.e. the dimensions they belong to) in all patient groups. If items behave differently (i.e. do not measure the supposed construct) it should be reconsidered whether this item can be used in this patient group. This is especially important in rehabilitation research because pooled analyses are frequently performed, evaluating outcome in a diagnostically mixed group of patients. Obviously, the above also applies when using disease specific instruments in other patient groups than those they have been developed for. It may be possible that the same instrument can be used in other patient populations, but this should be tested in advance. Traditional methods to investigate the dimensional structure of measurement instruments are factor analysis (or principal component analysis) and determining internal consistency of the dimensions (subscales) by calculating Cronbach's alpha. These methods have been used and described in diverse parts of this thesis.

*Factors of influence on Quality of Life*

While there is general agreement on the potential value of quality of life measures as key evaluation variables, there is an absence of clear agreement on a definition of quality of life: definitions of quality of life in the health context are mostly vague or absent. Conceptions relevant to health and quality of life are diverse, scattered through many disciplines, and use many different labels (e.g. health status, functional status, disability scale, quality of life)[11]. Bergner notes that the notion of quality of life has been a category in Index Medicus since 1966, yet quality of life is usually not defined in the reports of clinical trials[17]. None the less there seems to be acceptance that health-related quality of life is a multi-dimensional concept that encompasses the physical, emotional, and social components associated with an illness or treatment [10]. The term quality of life can have several meanings. It may be used to refer to outward material circumstances, so that good quality of life is represented by good physical health, material security, supportive family and friends, etc. Alternatively it can refer to subjective well-being, meaning an individual's sense of happiness or satisfaction,

typically reflecting a global assessment of all aspects of his or her life. Both emotional and cognitive factors may be referred to as part of subjective well-being, while objective conditions such as health, wealth, and comfort are seen to be potential influences but not inherently or necessarily part of the notion.

Quality of life is seen to be influenced by quite idiosyncratic factors, with a major determinant of an individual's quality of life being the perceived discrepancy between what is and what could have been [17]. There is no doubt that objective external factors such as income, length of survival, change in tumour volume, etc., influence quality of life. Generally such factors are assessed to be influences on quality of life, not the quality of life itself, however there are those who appear to argue that quality of life should be identified with physical conditions only. In the context of health status measurement, Kaplan et al. stated that "most investigators believe that symptoms and mortality do represent quality of life" [23], contrasting this approach with those who regarded quality of life as "subjective appraisals of life satisfaction" or those who combine a patient's subjective evaluation of well being with physical symptoms, sexual function, work performance, emotional status, etc.[24].

How people judge their quality of life or subjective well-being depends on various factors. Satisfaction within a given domain could conceivably be a consequence of subjective well-being. For example, Heady et al. have argued that satisfaction with work, standard of living, and leisure satisfaction, are largely the result of overall life satisfaction, and that satisfaction with friendship and general fitness (as opposed to illness) appear to be explicable solely on the basis of personality[25]. It has been argued that personality can heavily mediate the impact of exogenous life events, with each person having a "normal" equilibrium level of life events and subjective well-being, predictable on the basis of age and personality; only when events deviate from equilibrium levels is subjective well-being seen to change[25]. Individuals have also been shown to vary in coping strategies, which in turn can affect physical factors such as health outcomes[26]. In addition to individual-specific variables, there are general psychological mechanisms that act to increase subjective well-being independently of direct physical effects. For example patients frequently experience release of anxiety and stress in the initial stages of recovery following surgery[27]. Other effects can develop more steadily, for example Cassillet (cited Breetvelt and Van Dam[28]) reported that patients with newly diagnosed illness had greater anxiety and depression than patients who had been living with the illness for longer periods. Adaptation can be so great as to apparently eliminate subjective well-being differences between people chronically ill and controls, or even those who have recently had very positive experiences. These findings may sometimes reflect inadequacies in the quality of life instruments. However they also suggest fundamental homeostatic processes, such as the re-setting of expectations, change in reference standards, etc. Diener concluded

that health does seem correlated with subjective well-being, but that adaptation markedly reduces its influence[29].

*III. Quality of life and Cochlear Implantation in selected groups*

In this thesis it was investigated whether differences could be seen in the classroom performance and communicative skills between CI children and their normal hearing peers, by means of the AMP and SIFTER questionnaires *(1.1)*.

All the CI students scored "above average" in the AMP questionnaire. In some questions on communicative skills the CI students scored less than their normal hearing peers. The AMP questionnaire has so far only been used in one other study[30]. This other American study reported similar results as our study, with CI children staying behind on communicative skills. Results from CI students in mainstream educational setting from all over the world may not be compared automatically, as entering levels could differ. Nevertheless, it would be very interesting to compare communicative performance of CI children in different countries, for example by means of the AMP. In a second study on CI children in mainstream education *(2.1)*, we have examined the structure of the AMP questionnaire by means of factor and reliability analyses, which appeared to be good. Hereby, it should be noted that we were not yet able to include significant amounts of patients and/or controls.

In relation to the question how CI children communicatively perform in mainstream education with regard to their normal hearing peers, the SIFTER outcome showed the CI children were delayed in communication in kindergarten as well as in elementary school *(1.1)*. A recent report on educational performance of CI students in mainstream education in Malaysia also used the SIFTER. They found, as we did, that the area of communication shows the worst pass rate of the five areas[31]. SIFTER results are organized in categories (failure, marginal and sufficient), not in actual numeric outcomes[32]. This shows a difficulty in interpreting numerical statistical differences between the CI group and the normal hearing group within the same category. During the development of the SIFTER questionnaire in 1989, several areas of risk for children with hearing problems in educational settings, were determined, based upon literature research and other instruments. After content validity tests and item analyses, five areas with three questions each remained. Current questionnaire analyses, reported in chapter 2.1, did not confirm the proposed structure. Based upon the small number of patients in our current report, no conclusions can be drawn about the comparison with previous questionnaire structure analyses. The reliability of the SIFTER was previously tested in a large population ($n > 500$) and appeared to be moderate[10]. This reliability finding was confirmed by our reliability analysis results.

In our studies, two implant variables seemed to have a large effect on the questionnaire results: the duration of deafness and age at implantation. Many studies described the influence of age at implantation on speech perception and oral

communication abilities[33-44]. Speech perception and language development could also play a substantial role in the performance of CI children in mainstream educational settings. The current study showed that (long-term) language test results measured by the Reynell tests (and classroom performance) correlated significantly with the age of implantation. Few studies addressing the language development of children with implants have used the Reynell tests[36, 45, 46]. Robbins et al. found that language development could be above and beyond that anticipated from maturation alone, showing an increase in rate of language development[18]. Miyamoto et al. showed that rate of learning expressive language skills of prelingually deaf children with a CI was equivalent to that of normal hearing peers and that no plateau in language learning was observed. They also saw a large variability in language development and suggested that this may have been due to intersubject differences in speech perception. We looked at this possible relationship and found that there is a strong significant relation between speech recognition and expressive as well as receptive language development. However, our studies showed that better speech recognition scores do not always imply better classroom performance. Therefore, further research was done with analyses of receptive and expressive language development in combination with classroom performance.

In Chapter 2.1 it was found that within the AMP and SIFTER respectively 4 questions and 1 question have a strong predictive value for language development. This implies that the questionnaires may be used for screening in future, as they seem to be able to detect language developmental problems.

A recent review article described, by means of category referenced scales, the association of CI with significant enhancements in academic abilities and participation or engagement in the process of education, as mentioned by teachers. This review article also emphasized the need of studies on CI children in educational achievements and quality of life[47].

Finally, it should be taken into account that although considerable statistical analyses were performed on the AMP and SIFTER questionnaire, a relatively small amount of CI pupils was included. We feel that reproducibility could be retested in large group settings, even though the strong statistically significant correlations between questionnaires and language test results showed an obvious predictive value of certain questions within the AMP and SIFTER for language development.

The global experience of implanting deaf-blind subjects is growing, but there are still only a few reports in literature discussing the results of cochlear implantation in deaf-blind patients [48-50]. The present data *(1.2)* show that most people with Usher syndrome receive significant benefit from cochlear implantation in specific areas. EHL results illustrate an increase in speech perception abilities after implantation[51]. This increase in speech perception is also subjectively reflected in the Nijmegen Cochlear Implant

Questionnaire results. The beneficial effect of CI in Usher patients confirms previous findings. An important benefit in speech perception after implantation was described and these results showed a relation with the age of implantation. There was no relation seen between the visual acuity and the logopedic results in 13 implanted Usher patients (11 of which Usher type I)[48]. In our own research centre the first four Usher patients who received a CI were described in 1994[49]. This study has found no clear relationship between Usher type I mutations and audiological performance after implantation. These results substantiate previous findings that early implantation leads to better performance than implantation at older age[52]. It was also shown that cochlear implantation is beneficial to the majority of Usher type I patients when performed within the first or second decade of life.

Quality of life, focusing on remaining independent, of Usher patients type I, II and III was measured by means of the Usher Lifestyle survey *(2.2)*. An overall trend illustrated that type I Usher patients encounter more difficulty in preserving their independence than Usher type II patients. An increase in encountered difficulties was seen with regard to age. Results indicated that worse hearing means more help is needed, which obviously decreases after cochlear implantation. Dalton et al. showed that the severity of hearing loss was significantly associated with hearing impairment and with self-reported communication difficulties[53] .

The questionnaires currently used to assess quality of life in the three Usher studies, showed a benefit in the specific hearing domains of these quality of life instruments. In the Usher Lifestyle survey it was found that Usher people with CI seemed to be able to live a more independent life. A significant positive impact on the quality of life of deafblind individuals who receive a CI was expected by others[5]. Nevertheless, quality of life should be split into specific (hearing related) and generic (health and social circumstances) quality of life. The G(C)BI questionnaires correlate undoubtedly with the audiological performance in implanted Usher type I patients. Perceptibly, decline in EHL score is associated with an increasing benefit. In conclusion, the G(C)BI questionnaires are reliable to asses patient benefit in cochlear implantation as well. Furthermore, even though cochlear implantation was performed in some patients more than 10 years ago, the benefit could still well be recorded. This seems to justify the use of the retrospective questionnaires over a longer period of time.

In Chapter 1.3 the results of the children with developmental retardation or learning disabilities were compared to those of a control group of children with deafness alone ('normal' paediatric CI recipients). The results showed that there were minimal differences in the number of hours of CI use per day, the types of school, preferred communication method, quality of life measurements and language comprehension scores between the children with developmental retardation or learning disabilities and the control children.

In terms of mean values, the children in the control group were using their CI for more hours per day than the children in the study group, but the difference was not statistically significant. This result was also mentioned in the study by Knutson et al. [54]. Studies on the preferred communication method in children with multiple disabilities showed that they were making less use of verbal communication than the children with deafness alone [34, 55, 56]. Our results supported this finding. Although the children in the study group did not have statistically significantly lower scores on the language comprehension tests or the quality of life questionnaires than the control group, a large proportion of them were attending schools for the deaf or schools for deaf children with an additional handicap. This is quite a remarkable finding. It is possible that some of the children with extra disabilities have been placed in an inappropriate educational setting. In the study by Archbold et al., a larger proportion of the children with deafness alone were attending normal schools [57] , though hereby it should be noted that the UK has a different educational system for people with a handicap than the Netherlands.

Parents of the study group children were slightly more worried about communication, listening and speech development than the parents of the control group. Nevertheless, the study group's language comprehension development was almost equal to that of the CI children with deafness alone. It may be possible that the parents of the study group were more worried about language and speech development, because of the smaller amount of information existing on the development of multiple handicapped CI children. For CI children with deafness alone, more and univocal information is available. Providing full and correct information is an important factor to help parents form realistic ideas and expectations about the effect of cochlear implantation [58, 59]. It should be noted that in the current study, no expressive spoken language skills were investigated. Parents may also be concerned about this aspect of their children's communication abilities, though this was not studied in the present report.

According to the answers that the parents of the study group and control group gave to the questions in the PedsQL, children living with a CI have a positive feeling about their QoL. In the study group, the score was even slightly higher than in the control group. The parents of the two groups of children in this study judged the quality of life of their children to be about the same as that reported by the parents of healthy children in the study by Varni et al. [60].

The present study had a retrospective design, so that no data were available on quality of life prior to cochlear implantation. However, the results of the GCBI showed positive impact of the CI on the QoL of both study groups. This is in agreement with the findings in other studies. Chmiel et al. and Vlahovic et al. asked parents to judge quality of life factors in a closed format questionnaire [61, 62]. Wiley et al. also invited parents to give their views on quality of life using open questions [63]. All three studies

showed that the parents of children with extra disabilities regarded their child's quality of life to be better after cochlear implantation than before.
Language comprehension tests after three years of CI use, revealed that our group of children with developmental retardation or learning disabilities performed almost equally to the control group children. This was a remarkable finding, particularly because the children with extra disabilities had a lower mean non-verbal IQ than the children with deafness alone. Hypothetically, (non-verbal) IQ has little effect on the language comprehension process. It should be noted that all the children with extra disabilities are far behind compared to the normal hearing children though. However, studies on language development after cochlear implantation observed that the children with several disabilities made slower progress and ultimately achieved poorer results than the children with deafness alone [34, 64, 65]. A poorer result generally meant that the children with multiple disabilities had more difficulty with the complex tasks (e.g. open set speech perception) than the children with deafness alone. In this study, we only assessed language comprehension and a slower development was not visible.

In the current long-term follow-up study, cochlear implantation and multiple aspects of specific and generic quality of life were combined with utility scores *(1.4)*. Our results showed that the beneficial effects of a CI on quality of life were constant in the long-term. The hearing-related quality of life scores (NCIQ) increased after cochlear implantation and this beneficial effect remained clearly visible. Nevertheless, a small but non significant trend towards deterioration was observed over time. A similar trend was detected in the unimplanted control group, which suggests that the decline was not necessarily connected with cochlear implantation. The deterioration could for example be a natural effect of aging. The positive effect of a CI on QoL has been described by others, although not many researchers used the NCIQ[66, 67]. Recently, Cohen et al. used the NCIQ to compare quality of life between CI users and hearing aid (HA) users, all with postlingual deafness. The results of their analyses supported the value of this instrument in the HA group and CI group[68]. They found beneficial effects of a CI in all the subdomains, which were equivalent to our results. As these authors had not used generic quality of life instruments, it was not possible to compare the effect of a CI on hearing-specific and generic quality of life.
The Health Utility Index mark 3 (HUI 3) did not show any significant changes in the long-term. All the domains and the utility score reflected a small and non-significant decrease in quality of life. Utility scores in the groups with and without CI showed comparable deterioration, which was also apparent in the mean NCIQ scores. These findings may confirm our hypothesis on the natural effects of aging. Similar deterioration in HUI scores over time was described by the UK group in cochlear implant patients[69]. The utility score of the more recently implanted group improved significantly after they had received a CI. Only one other study reported the use of the

HUI mark 3 to determine changes in HRQoL after a CI, but this was in prelingually deaf children[70]. The smaller level of improvement in our study than in the report by Cheng et al. could be due to the fact that all the patients in our study group were postlingually deaf adults.

In our long-term evaluation, a significant decrease was detected in five out of the eight SF36 domains. Results of the unimplanted group were ambiguous and did not show any type of trend. In general, these unclear findings seem to confirm the variable SF36 results after cochlear implantation mentioned by Krabbe et al.[71]. A lack of sensitivity of the SF36 to detect changes in quality of life after cochlear implantation was also seen in an earlier study on 27 postlingually deaf adults[72] and in a number of Usher type I subjects who had received a CI[73]. According to the literature, the SF36 lacks the necessary sensitivity to detect clinically meaningful improvements in patients with hearing impairment[74]. It has been reported that quality of life and utility scores are based on two main factors: the dimensions used to describe a person's health state and the technique used to assign a value to the health state descriptions elicited by each of the quality of life questionnaires[75]. The Health Utilities Index focuses on a person's capability to undertake certain tasks, such as hearing and speech (production), but it does not consider the implications of any impairments. In contrast, the SF-36 focuses on performance rather than the underlying level of impairment. Therefore, on the basis of the previous and present observations of low sensitivity, we believe that the SF36 should not be the first choice of a generic quality of life questionnaire to evaluate hearing impaired patients.

Long-term effects of a CI on speech recognition tests showed progressive increases in suprasegmental scores and segmental speech perception tests. In long-term CI users, the initial improvement in speech recognition after cochlear implantation increased significantly over the subsequent six years. This is an impressive finding, because other authors have demonstrated no increase in speech perception outcomes[70, 76] or only improvement over a shorter follow-up period[77].

The Parental Perspectives instrument for CI children *(2.3)* has been currently investigated. The results from statistical analyses to derive the underlying structure of the PP instrument indicated the absence of the anticipated structure in eight domains. The original instrument, developed and validated by Archbold et al.[78], was extensively studied by various research institutes. The first to examine the PP, at that time called the "Children with cochlear implants: parental perspectives", were Dujardin et al.[79]. A low response rate made the authors realize the importance of an exploration of a short form PP. It was concluded that the original structure of ten domains could not be detected. Reliability analyses only showed good reliability for the domain of communication, which is in concordance with our current results. Their study emphasized the importance of a large number of participants. Afterwards, O'Neill et

al. investigated the reliability of the PP[80]. Test-retest reliability appeared to be > 95% for most instrument items. They suggested that an extensive use of the instrument could result in valid comparisons of outcomes among different cochlear implant centres and they highlighted the need for an acceptable, valid and reliable instrument for parental perspectives in cochlear implantation. Then Nunes et al. investigated the reliability and validity of the instrument, at that time called "Parents views and experiences with pediatric cochlear implant questionnaire" (PVECIQ)[81, 82]. Based upon the interview with parents an extra domain, "functioning before the implantation", was added, domains were redefined and 23 items were excluded. Relatively good content validity was assessed by comparison with parents' responses to an interview. A factor analysis of the total instrument scales identified four components. Finally, just recently, Lin et al. studied this questionnaire and its literature as well and noted that a substantial factor analysis had so far not been performed (which is what we did in the current study)[83].

As different methodological approaches were used in the studies mentioned above, it is hard to directly link the former studies to the current investigation. Based upon the large numbers of users and extensive research conducted previously, the importance of the PP questionnaire seems inevitably clear. It was realised that reducing the PP questionnaire length will inevitably result in loss of information. We feel that statistical analyses of the instrument, including validity testing, should be done in large group sessions in the future.

In an attempt to analyse an existing instrument, it is impossible to find the one singular correct way to do so. We applied an analytical methodology, contemplating the use of this instrument for prospective follow-up. In this study the PP instrument was analysed from a psychometric point of view, nonetheless a more clinimetric (qualitative, clinical) approach would also have been possible. The principles of clinimetrics were first introduced by Feinstein in 1987[84].

Clinimetric and psychometric strategies are the two different scientific frameworks used for the development of multi-item health measurement scales[85]. The term "clinimetrics" indicates a domain concerned with indexes, rating scales and other expressions that are used to describe or measure symptoms, physical signs, and other distinctly clinical phenomena in medicine. Clinimetric strategies used in clinical medicine rely primarily on the judgements of patients and clinicians and aim to measure clinical phenomena that are generally believed to comprise several unrelated patient characteristics or attributes (scales measuring several attributes are called "heterogeneous" scales)[84].

Psychometry is discipline pertaining to psychological and mental testing, and to any quantitative analysis of a person's psychological traits or attitudes or mental processes. Psychometric strategies used in psychology and education rely more on mathematical techniques and generally (although not exclusively) aim at developing one scale (or

multiple scales) measuring single patient characteristics or attributes (scales measuring a single attribute are called "homogeneous" scales)[86].

Thus, the two methods differ in aims and strategy. The differential aspects of clinimetrics and psychometrics have become the focus of debate[87]. De Vet et al. and Fava et al. argued for the importance of clinimetrics as a methodological discipline which is concerned with measurement issues in clinical medicine and emphasized the substantial overlap between clinimetrics, psychometrics and biometrics together with the need for a better integration of the fields[88, 89]. Emmelkamp states that although clinimetrics may have a number of advantages over the more classical psychometrically based measures, such as being more sensitive to change, there are still a number of methodological issues that deserve to be studied before we may abandon the psychometrically based methods[90]. Clinimetric theory offers the conceptual ground for a substantial revision of assessment parameters and for linking co-occurring syndromes. From a clinical viewpoint, it may allow more flexibility and be more in tune with the clinician's reasoning, both in terms of diagnostic assessment and evaluation of co-morbidity, once variables are no longer considered as equal. From a research viewpoint, it may pave the way for inclusion criteria and assessment tools, which are more suitable for the purposes of clinical research. In summary, a scale developed with a clinimetric strategy can measure a complex (so-called heterogeneous) clinical phenomenon (thought to be composed of several patient attributes), but still fulfil psychometric criteria for "homogeneity." Thus, these strategies for the development of health measurement scales, which for a long time have been considered potentially opposite or conflicting, may be complementary[91]. In the rest of this thesis mostly the clinimetric point of view was taken and implemented.

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# Summary and Conclusions

# Summary and conclusions

Cochlear implantation aims to restore the hearing of deaf individuals. Main outcomes, directly related to hearing with a CI, e.g. language perception and speech production, have been extensively studied. It is important to realize that hearing can be an aid in speaking and reading and consequently can enable communication. However, the impact of a CI on quality of life has been investigated to a lesser extent; therefore, in this thesis, the quality of life in diverse CI populations (part 1) and by means of various instruments (part 2) was studied. An insight into the diversity in quality of life between differing populations receiving a CI was obtained. Diverse instruments were studied, in order to obtain knowledge of which questionnaires can be used for quality of life research in a CI population.

In the General Introduction short overviews are presented concerning both cochlear implantation and quality of life. The first part contained information regarding the functioning of a cochlear implant, our cochlear implant team and the performance of patients with their CI. In the second part, the definition of quality of life and the diverse instruments used in this thesis were discussed. The introduction was finished by declaring the aims of this thesis.

*PART 1*

Chapter 1.1 described the comparison of classroom performance of CI children in mainstream education with that of their normal hearing peers. The teachers filled in two questionnaires: the Assessment of Mainstream Performance (AMP) and the Screening Instrument for Targeting Educational Risk (SIFTER). The CI children scored above average in the AMP and sufficient in all but one area (communication) of the SIFTER questionnaire. Class ranking did not differ significantly between the CI students and their normal hearing peers. Overall, the normal hearing group outperformed the CI group. Classroom performance of CI children correlated negatively with duration of deafness and age at implantation. It was concluded that although results are encouraging, the CI group scored significantly less than their normal hearing peers in most questionnaire domains of both the AMP and the SIFTER. Most important variables for the outcome in this study were age at implantation and the duration of deafness. Unfortunately, no data on hearing impaired subjects were available.

Chapter 1.2a evaluated the quality of life, hearing and vision in Usher syndrome type I patients with and without CI. The quality of life of fourteen Usher type I patients with a CI was compared to that of Usher type 1 patients without a CI by means of three questionnaires: Nijmegen Cochlear Implant Questionnaire (NCIQ), the Standard Medical Outcome Study Short-Form 12 (SF12) and the Usher Lifestyle Survey. A

significant benefit of CI was seen in the hearing specific questionnaire NCIQ. This could not be detected in the generic SF12 survey. The Usher Lifestyle survey indicated that Usher type I patients with a CI tend to be able to live an independent life more easily than the unimplanted Usher type I patients. In general, overall quality of life can be enhanced by CI in Usher type I patients, though effects are mostly seen in hearing related quality of life-items.

Chapter 1.2b investigated the benefit and performance of cochlear implantation in Usher syndrome type I patients. 14 patients with a clinical diagnosis of Usher type I were included. Mutation analysis of Usher type I genes was performed. All patients filled in the Glasgow (Children's) Benefit Inventory (G(C)BI) questionnaire. In addition, equivalent hearing loss scores (EHL) were calculated to measure auditory performance. Correlations between the mentioned parameters were studied. Cochlear implantation performed within the first two decades of life was beneficial to 13 out of the 14 of the Usher type I patients. The EHL score and the G(C)BI score showed a significant correlation: the benefit of implantation increases with a decreasing EHL score. It can be said that cochlear implantation in Usher type I patients improves the audiological performance when patients are implanted at and earlier age and is beneficial according to the G(C)BI, when performed within the first two decades of life.

Chapter 1.3 compared the quality of life and language comprehension after cochlear implantation in deaf children with additional interfering handicaps (mild developmental retardation or learning disabilities) with that of a matched control group of implanted children with deafness alone. The Glasgow Children's Benefit Inventory (GCBI), the Paediatrics Quality of Life (PedsQoL) questionnaires and separate questions on the parents' worries regarding the CI were used. Furthermore, comparisons were made of language perception scores. The children with developmental retardation or learning disabilities tended to use their CI less than the control group. Only one multiple handicapped child was attending a regular primary school compared to nine control children. The results of the questionnaires did not reveal significant differences in quality of life and benefit of the CI between the two groups. Language comprehension of children with the extra disabilities was after three years of implant use, comparable to that of the control children. These results highlight the importance of cochlear implantation in deaf children with developmental retardation or learning disabilities as additional handicaps.

Chapter 1.4 investigated long-term quality of life in postlingually deaf adults after entering the cochlear implantation programme. This study was a follow-up study from 1998. Long-term CI users, patients who have not received a CI and relatively short-

term CI users were re-evaluated six years after initial data collection in 1998 by using three questionnaires: the NCIQ, the SF36 and the Health Utility Index mark 3 (HUI3), as well as speech perception tests. In general, the beneficial effect of a CI remained stable during long-term follow-up, although the scores on the questionnaires decreased slightly. The group without a CI demonstrated slightly decreasing trends in outcomes over time. Long-term speech perception performance with CI improved in time. This was the first study to investigate long-term follow up of CI patients, in all aspects of quality of life combined with speech perception performance, in comparison with postlingually deaf adults without a CI.

*PART 2*

Chapter 2.1 reported on an investigation on the relation between classroom performance and language development of CI students in mainstream education. Structural analyses of Assessment of Mainstream Performance (AMP) and Screening Instrument For Targeting Educational Risk (SIFTER) instruments were performed. AMP and SIFTER instruments measured classroom performance and language development was measured by means of Reynell and Schlichting tests. AMP and SIFTER domains showed good reliability (Cronbach's alpha > 0.6), but factor analyses only showed the expected instrument structure in the AMP. In both questionnaires and within all domains, individual variability was detected. Spearman's correlation analyses showed a probable explanation of individual questionnaire variability by language test results (p-value mostly <0.01). The AMP and SIFTER instruments showed a predictive capacity for language development, based upon general linear model univariate and linear regression analyses. Individual classroom performance, measured by AMP and SIFTER questionnaires, of CI children in mainstream education varies. However, correlation analyses showed strong significant relation between classroom performance and both expressive and receptive language test results (Schlichting and Reynell tests). The AMP is an important new instrument, as it seems to monitor the actual linguistic functioning of the child.

Chapter 2.2 describes a study on the descriptive Usher Lifestyle Survey, which is developed for patients with different types of Usher syndrome. Three different types of Usher (I, II and III) are distinguished by differences in onset, progression and severity of hearing loss and by presence or absence of balance problems. In this study, 93 Usher patients from 7 European countries participated. Following the Nordic definition of maintaining independence in deafblindness, three domains were investigated in the questionnaire: access to information, communication and mobility. Research variables in the study were: age, type of Usher, hearing loss and the number of retinitis pigmentosa related sight problems. Results of Usher type I and II patients were presented. Usher type I patients tend to need more help than Usher type II

patients and the amount of help that is needed increases as patients get older or as considered hearing loss worsens. No patterns in results were seen for the number of retinitis pigmentosa related sight problems.

Chapter 2.3 evaluated the consistency and validity of the widely used Parental Perspectives questionnaire (PP) by means of statistical analysis. It also explored the possibility to develop a short version of the PP as the instrument is often regarded as being rather lengthy. Parents of 130 CI children participated in this study. To assess internal consistency among the PP items of the domains, Cronbach's alpha coefficients were calculated. Corrected item-total correlations were carried out to investigate the strength of associations with individual items with the construct. Factor analysis was performed to identify the statistical factors of the original PP and to explore a revised short form of the PP. Some items could be disposed of, based upon the Cronbach's alpha analyses and upon consistent reasoning. After reducing the number of factors based on standard criteria, a three-domain structure with 23 items was shown as main concept. Reliability analyses of the new domains of the proposed short version PP showed good internal consistency and strong associations of the individual items with the construct. It was concluded that the Parental Perspectives instrument is an important tool to assess the impact of cochlear implantation of a child for the quality of life for the family and the child itself. This statistical investigation showed a possible option for the development of a short form usable in prospective follow-up studies.

The General Discussion presented an overview of the divers dilemmas and problems which are faced in investigating Quality of Life, especially in a clinical setting. The discussion is divided into three parts, being cochlear implantation and the Deaf society, quality of life measurement and finally cochlear implantation and quality of life.

# Samenvatting en conclusies

Cochleaire implantatie beoogd herstel of verbetering van het gehoor bij doven of ernstig slechthorenden. Er is inmiddels uitgebreid onderzoek verricht naar gehoor gerelateerde uitkomsten bij CI patiënten, zoals bijvoorbeeld taalperceptie of spraakproductie. Het is van belang te realiseren dat gehoor essentieel is voor communicatie. Desalniettemin zijn de effecten van een CI op de verschillende aspecten van het leven (oftewel de kwaliteit van leven) in veel mindere mate onderzocht. Vandaar dat in dit proefschrift de kwaliteit van leven in verschillende populaties (deel 1) en verschillende instrumenten om de kwaliteit van leven te meten (deel 2) werden bestudeerd. Op deze manier verkregen we een inzicht in de kwaliteit van leven van verschillende groepen CI gebruikers. De consistentie van de verscheidene instrumenten werd ook getest, daar op die wijze informatie kan worden verkregen betreffende de bruikbaarheid van deze instrumenten voor (prospectief) onderzoek bij CI patiënten.

De Algemene Introductie presenteerde een kort overzicht over zowel cochleaire implantatie als mede het begrip kwaliteit van leven. Het eerste gedeelte van de inleiding betreft informatie over de werking van een CI, over ons Cochlear Implant Team en over de prestaties van patiënten met hun CI. Tevens werd de definitie van kwaliteit van leven besproken, even als de verscheidene vragenlijsten die in dit proefschrift gebruikt werden. Tenslotte werden de doelstellingen van dit proefschrift gepresenteerd.

*Deel 1*

Hoofdstuk 1.1 beschreef de vergelijking van de prestatie van de CI kinderen en hun normaalhorende klasgenootjes in het reguliere onderwijs. De docenten vulden twee vragenlijsten in, zijnde de Assessment of Mainstream Performance (AMP) en de Screening Instrument For Targeting Educational Risk (SIFTER). De CI kinderen scoorden boven gemiddeld op de AMP vragenlijst en voldoende op haast alle onderdelen van de SIFTER (niet op het onderdeel betreffende 'communicatie'). Niveau van het kind volgens de docent, verschilde niet significant tussen de groep CI kinderen en hun horende klasgenoten. In het algemeen genomen, presteerden de horende kinderen echter beter dan hun CI dragende klasgenoten. De prestatie van de CI kinderen in de klas correleerde negatief met de duur van doofheid en met de leeftijd waarop zij hun CI ontvingen. Er kan worden geconcludeerd dat de resultaten van de CI kinderen in het reguliere onderwijs bemoedigend zijn, maar echter achter blijven bij de horende klasgenoten, volgens de meeste domeinen van de AMP en SIFTER vragenlijsten. Meest in het oogspringende variabelen van invloed op de uitkomsten van deze studie waren leeftijd van implantatie en duur van de doofheid.

Hoofdstuk 1.2a evalueerde de kwaliteit van leven, gehoor en visus van groepen Usher syndroom type I patiënten met en zonder CI. De kwaliteit van leven werd vergeleken met behulp van drie vragenlijsten; de Nijmegen Cochlear Implant Questionnaire (NCIQ), de Standard Medical Outcome Study Short-Form 12 (SF12) en de Usher Lifestyle Survey. Een significant voordeel van CI werd gezien in de gehoorspecifieke vragenlijst NCIQ. Een verschil tussen de groep met en zonder CI werd niet geconstateerd in de algemene SF12 vragenlijst. De Usher Lifestyle Survey gaf aan dat Usher type I patiënten met CI gemakkelijker een onafhankelijk leven leiden dan Usher type I patiënten zonder CI. In het algemeen gold dat kwaliteit van leven van Usher type I patiënten vergroot kan worden met een CI. Dit effect is met name terug te zien in gehoorspecifieke kwaliteit van leven.
Hoofdstuk 1.2b beschreef het onderzoek naar de variabelen die de resultaten van cochleaire implantatie bepalen bij Usher type I patiënten. 14 Patiënten met de klinische diagnose Usher type I werden geïncludeerd in deze studie. Mutatie analyse van Usher type I genen werd bij allen verricht. Alle patiënten vulden de Glasgow (Children's) Benefit Inventory (G(C)BI) vragenlijst in. Tevens werden Equivalent Hearing Loss scores (EHL) berekend om auditieve prestatie te kwantificeren. Correlaties tussen de genoemde parameters werden bestudeerd. Cochleaire implantatie binnen de eerste twee levensdecaden bleek voordelig voor 13 van de 14 Usher type I patiënten. Tenslotte lieten de EHL score en de G(C)BI score een significante correlatie zien; het voordeel van implantatie stijgt naarmate de EHL score afneemt. Er kan gesteld worden dat cochleaire implantatie bij USH1 patiënten de audiologische prestatie verbeterd wanneer de patiënten in een vroeg stadium geopereerd worden en dat implantatie voor de leeftijd van 20 jaar, het meest voordelig is, gebaseerd op de resultaten van de GCBI.

Hoofdstuk 1.3 vergeleek de kwaliteit van leven en taalbegripscores na cochleaire implantatie tussen dove kinderen mèt en zonder ontwikkelingsachterstand of ernstige leerproblemen. De Glasgow Children's Benefit Inventory (GCBI), de Paediatrics Quality of Life (PedsQoL) vragenlijsten en separate vragen betreffende de zorgen die ouders kunnen hebben met betrekking tot de implantatie werden gebruikt. Er werden tevens vergelijkingen gemaakt tussen de taalbegripsscores van de beide groepen CI kinderen. De kinderen met een meervoudige handicap bleken hun CI iets minder uren per dag te gebruiken dan de controle groep. Slechts een van de CI kinderen met extra handicap volgde op het moment van het onderzoek regulier onderwijs, in vergelijking met negen kinderen van de controle groep. Uitkomsten van de vragenlijsten toonden aan dat de ouders van de kinderen met ontwikkelingsachterstand of ernstige leerproblemen het CI bijna even profijtelijk voor hun kind ervoeren als ouders van de controle kinderen. Na drie jaar CI gebruik, bleken de taalbegripsscores van de kinderen met ontwikkelings-achterstand of ernstige leerproblemen vergelijkbaar met de scores van de controle groep.

Deze resultaten bevestigen het belang van cochleaire implantatie bij dove kinderen met een ontwikkelingsachterstand of ernstige leerproblemen als additionele handicap.

Hoofdstuk 1.4 beschreef een onderzoek betreffende de lange termijn kwaliteit van leven bij postlinguaal dove volwassenen na begin van de CI procedure. Deze follow-up studie liep vanaf 1998. Lange termijn CI gebruikers, patiënten die geen CI hadden en patiënten die relatief kort hun CI gebruiken, werden vergeleken middels drie vragenlijsten, de NCIQ, de SF36 en de Health Utility Index mark 3 (HUI3), en spraakverstaanscores, dit alles zes jaar na eerste dataverzameling in 1998. In het algemeen kan worden gesteld dat het positieve effect van het CI op de kwaliteit van leven, zoals in 1998 vastgesteld, zichtbaar bleef tijdens de lange termijn follow-up, al namen de scores van de vragenlijsten wel iets af. De groep zonder CI toonde een kleine afname in uitkomsten. Spraakverstaanscores bleken in loop van de jaren nog te verbeteren. Dit was de eerste studie die de lange termijn effecten, zijnde alle aspecten van kwaliteit van leven en spraakverstaanscores, van een CI heeft gemeten, en vergeleken met een groep postlinguaal doven zonder CI.

*Deel 2*

Hoofdstuk 2.1 beschreef het onderzoek naar prestatie in de klas en taalontwikkeling van kinderen met een CI in het reguliere onderwijs. Structurele analyse van de Assessment of Mainstream Performance (AMP) en de Screening Instrument For Targeting Educational Risk (SIFTER) vragenlijsten werd eveneens verricht. Prestatie in de klas werd gemeten met behulp van de AMP en SIFTER vragenlijsten, de taalontwikkeling door middel van de Reynell en Schlichting testen. De verschillende domeinen van de AMP and SIFTER toonden goede betrouwbaarheid (Cronbach's alpha > 0.6), factor analyse bevestigde enkel de vooraangenomen structuur in de AMP, niet in de SIFTER. In beide vragenlijsten, binnen alle domeinen, werd een individuele variabiliteit gezien. Spearman's correlatie analyses toonden een mogelijke verklaring voor deze individuele variatie; variatie in taaltest resultaten komt overeen met variatie in vragenlijst scores (p-waarde voornamelijk <0.01). De AMP en SIFTER vragenlijsten zijn significant geassocieerd met taalontwikkeling, dit gebaseerd op lineaire univariate analyse en lineaire regressie analyse. Kortom, individuele prestatie in de klas, gemeten door AMP en SIFTER lijsten, van CI kinderen varieert. Correlatie analyse toonde een sterk significante relatie tussen vragenlijst resultaten (prestatie in de klas) en resultaten van zowel expressieve als receptieve taaltesten (Schlichting en Reynell testen). Structurele vragenlijst analyse van de AMP en SIFTER toonde goede betrouwbaarheid. De AMP is een nieuw en belangrijk instrument daar het daadwerkelijke taalgerichte functioneren van een kind in kan beeld brengen.

Hoofdstuk 2.2 beschreef een studie over de Usher Lifestyle Survey, welke descriptief gebruikt kan worden voor patiënten met verschillende typen van het Usher Syndroom.

Drie verschillende types van Usher (type I, II en III) worden onderscheiden op grond van verschillen in begin, progressie en ernst van gehoorsverlies en aan- of afwezigheid van evenwichtsproblemen. In deze studie namen 93 Usher patiënten deel, afkomstig uit zeven Europese landen. Op basis van de Noord-Europese definitie van het behoud van onafhankelijkheid bij doofblindheid, werden drie domeinen onderzocht, namelijk: toegang tot informatie, communicatie en mobiliteit. Onderzoeksvariabelen waren leeftijd, type Usher, gehoorsverlies en aantal aan retinitis pigmentosa gerelateerde visus problemen. Resultaten van Usher type I en Usher type II patiënten werden besproken. Usher type I patiënten bleken meer hulp nodig te hebben dan Usher type II patiënten en dit neemt toe naarmate de patiënten ouder worden of subjectief meer gehoorsverlies hebben. Aantal retinitis pigmentosa gerelateerde visusproblemen leek hierop geen significante invloed te hebben.

Hoofdstuk 2.3 evalueerde de bekende Parental Perspectives (PP) vragenlijst door middel van statistische analyse en onderzocht de mogelijkheid van het verkrijgen van een verkorte versie van de lijst, aangezien de originele versie vaak als te lang wordt beschouwd. Ouders van 130 CI kinderen namen deel aan de studie. Cronbach's alpha coëfficiënten werden berekend ter evaluatie van de interne samenhang van de vragen van de verschillende domeinen. Om de mate van associatie van de individuele vragen met de bouw van de lijst te onderzoeken, werden gecorrigeerde 'vraag ten opzichte van lijst' correlaties verricht. Factor analyse werd gedaan om de statistische factoren van de originele lijst te detecteren en een eerste aanzet te geven voor het ontwikkelen van de verkorte versie. Sommige vragen konden geëxcludeerd worden, op basis van Cronbach's alpha analyse of logische redenatie. Na het verwijderen van deze vragen, werd een drie domeinen structuur gevonden, met nu een totaal aantal vragen van 23. Betrouwbaarheidsanalyse van de drie domeinen van de verkorte versie toonde goede interne samenhang  en een hoge mate van associatie van de individuele vragen met de bouw van de lijst Er kon worden geconcludeerd dat de Parental Perspectives vragenlijst een belangrijk instrument is om een indruk te verkrijgen van de impact van cochleaire implantatie op de kwaliteit van leven voor het gezin en het CI kind zelf. Deze statistische analyse toonde een mogelijke aanzienlijk verkorte versie van de vragenlijst.

De Algemene Discussie presenteerde een overzicht van de diverse dilemma's en problemen die men tegenkomt bij het onderzoek naar kwaliteit van leven, in het algemeen en bij cochleaire implantatie van doven en ernstig slechthorenden in het bijzonder. De discussie is onderverdeeld in drie delen, zijnde cochleaire implantatie en de Doven wereld, meting van kwaliteit van leven en cochleaire implantatie en kwaliteit van leven.

# Dankwoord

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# Curriculum Vitae

Godelieve Wilhelmina Jozefina Andrew Damen werd op 18 april 1978 geboren in Eindhoven. In 1996 behaalde zij het eindexamen VWO aan het Lorentz Lyceum te Eindhoven. Omdat ze werd uitgeloot voor de studie geneeskunde in Nederland, is ze in 1996 begonnen met geneeskunde aan de Katholieke Universiteit Leuven, België. Na de theoretische drie kandidatuurs- en twee doctoraatsjaren met succes te hebben afgerond, werd in 2001 een overstap naar de Universiteit Utrecht mogelijk, voor de afronding van de opleiding middels co-schappen. In 2003 werd het artsexamen met speciale felicitaties behaald. Voor het onderzoek dat ten grondslag ligt aan dit proefschrift, werd ze in 2004 aangenomen op de afdeling KNO van het UMC St Radboud te Nijmegen. Per 6 maart 2006 begon ze alhier aan haar opleiding tot KNO-arts onder leiding van Prof. Dr. K. Graamans en Prof. Dr. C.W.R.J. Cremers.

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# List of abbreviations

| | | |
|---|---|---|
| A | | |
| | AMA | American Medical Association |
| | AMP | Assessment of Mainstream Performance |
| | AMP-E | Assessment of Mainstream Performance Elementary school |
| | AMP-K | Assessment of Mainstream Performance Kindergarten |
| | AN-test | Antwerp Nijmegen speech perception test |
| | ARMS | Amplification-Refractory Mutation System |
| C | | |
| | CAUSE | Charge Association Usher Syndrome Europe |
| | CI | Cochlear Implant |
| | CVC-test | Consonant Vowel Consonant test |
| D | | |
| | dBHL | decibel Hearing Level |
| E | | |
| | EHL | Equivalent Hearing Loss |
| F | | |
| | FAS | Functional Acuity Score |
| | FDA | Food and Drug Administration |
| | FFS | Functional Field Score |
| | FVS | Functional Vision Score |
| G | | |
| | GBI | Glasgow Benefit Inventory |
| | GCBI | Glasgow Children's Benefit Inventory |
| | GN test | Gestel Nijmegen speech perception test |
| H | | |
| | HA | Hearing Aid |
| | hFI | high Fletcher Index |
| | HL | Hearing Level |
| | HRQoL | Health Related Quality of Life |
| | HUI3 | Health Utility Index mark 3 |
| I | | |
| | IQ | Intelligence Quotient |
| M | | |
| | MCS | Mental Component Summary score |

| | | |
|---|---|---|
| N | | |
| | NAD | National Association of the Deaf |
| | NCIQ | Nijmegen Cochlear Implant Questionnaire |
| O | | |
| | ONICI | ONafhankelijk Informatiecentrum over Cochleaire Implantatie |
| | ORL | Oto- Rhino- Laryngology |
| P | | |
| | PCS | Physical Component Summary score |
| | PedsQL | Pediatrics Quality of Life |
| | PP | Parental Perspectives questionnaire |
| | PVECIQ | Parents Views and Experiences with pediatric Cochlear Implant Questionnaire |
| Q | | |
| | QoL | Quality of Life |
| R | | |
| | RDLS | Reynell Developmental Language Scales |
| | RP | Retinitis Pigmentosa |
| S | | |
| | SD | Standard Deviation |
| | SF12 | medical outcome study Short Form 12 |
| | SF36 | medical outcome study Short Form 36 |
| | SIFTER | Screening Identification For Targeting Educational Risk |
| | SPL | Sound Pressure Level |
| | SPSS | Statistical Package for the Social Sciences |
| U | | |
| | USH1 | Usher Syndrome type I |
| W | | |
| | WHO | World Health Organisation |