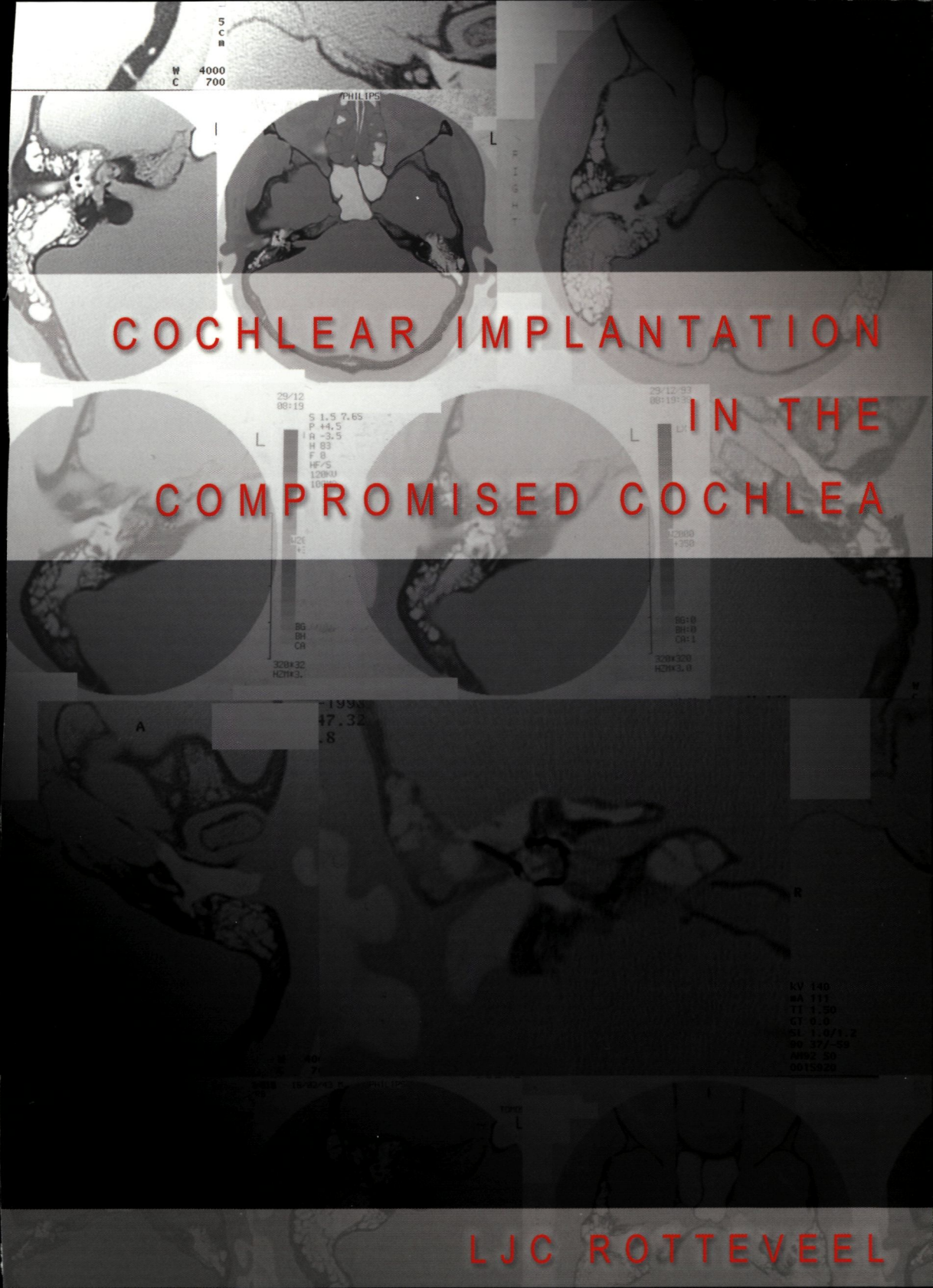


COCHLEAR IMPLANTATION IN THE COMPROMISED COCHLEA

LJC ROTTEVEEL

Cochlear Implantation in the Compromised Cochlea

Liselotte Rotteveel

Print: PrintPartners Ipskamp
Lay-out: Diny Helsper
Cover-design: Vincent Seekles

Rotteveel, Liselotte
Cochlear implantation in the compromised cochlea

Thesis Radboud University Nijmegen Medical Centre
Nijmegen, the Netherlands.

ISBN: 978-90-9023081-8

Cochlear Implantation

in the

Compromised Cochlea

Een wetenschappelijke proeve op het gebied van de
Medische Wetenschappen

Proefschrift

ter verkrijging van de graad van doctor
aan de Radboud Universiteit Nijmegen
op gezag van de rector magnificus prof. mr. S.C.J.J. Kortmann,
volgens besluit van het College van Decanen
in het openbaar te verdedigen op
maandag 9 juni 2008
om 13.30 uur precies

door

Liselotte Rotteveel
geboren op 16 Februari 1976
te Nijmegen

Promotores: Prof. dr. ir. A.F.M. Snik
Prof. dr. C.W.R.J. Cremers

Copromotor: Dr. E.A.M. Mylanus

Manuscriptcommissie:
Prof. dr. C. van Weel (voorzitter)
Prof. dr. Th. Lenarz (Hannover)
Prof. dr. ir. J.H.M. Frijns (Leiden)
Prof. dr. L.A.A. Kollée
Prof. dr. H.P.H. Kremer

The study in this thesis was financially supported by Cochlear Corporate Australia and Stichting Atze Spoor Fonds.
Additional sponsoring was offered by Atos Medical, Beter Horen, Carl Zeiss, Electro Medical Instruments, Schering-Plough, Veenhuis Medical Audio.

Aan mijn ouders

Contents

Chapter 1

Introduction

1.1 Hearing, deafness and cochlear implants

Hearing is the result of sound vibrations being transmitted along the ear canal, through the middle ear, to the inner ear. The inner ear, or cochlea, is a snail-like structure of 2½ to 2¾ turns embedded in bone (Figure 1). It houses the sense organ of hearing, i.e. the organ of Corti (Figure 2). Three spiral compartments: the scala media, scala vestibuli and scala tympani make up the cochlear turns and they are composed of wound around the modiolus tat contains the spiral ganglion cells of the cochlear nerves. The scala media contains the organ of Corti, which rests on the basilar membrane and lies between the scala vestibuli and scala tympani. High frequency sound causes maximum vibration of the basilar membrane at the beginning of the cochlear turn, while low frequency sound causes vibrations at the end of the cochlear turn. As a result of these basilar membrane vibrations, the hair cells in the organ of Corti move back and forth. Hair cell movement evokes action potentials in the cochlear nerve that form patterns of excitation. These patterns are transmitted to the higher brain centres of the auditory pathway where they are interpreted as sound and processed as pitch and loudness, as well as speech (Figure 3). Some 20,000 hair cells are required for normal hearing.

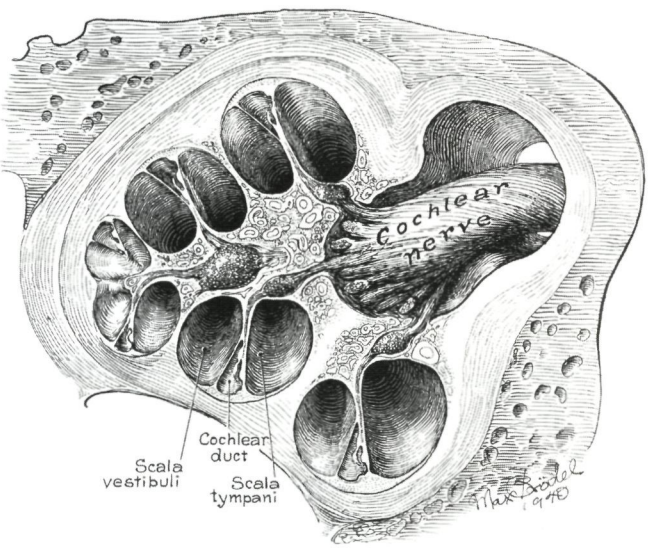


Figure 1. Schematic drawing of vertical section through right cochlea to show anatomy of cochlear duct, spiral ligament and cochlear nerve.
(By courtesy of G.T.Nager, from: Pathology of the Ear and Temporal bone, Chapter 6, p.6, C.W. Mitchel ed., Williams & Wilkins publ., USA)

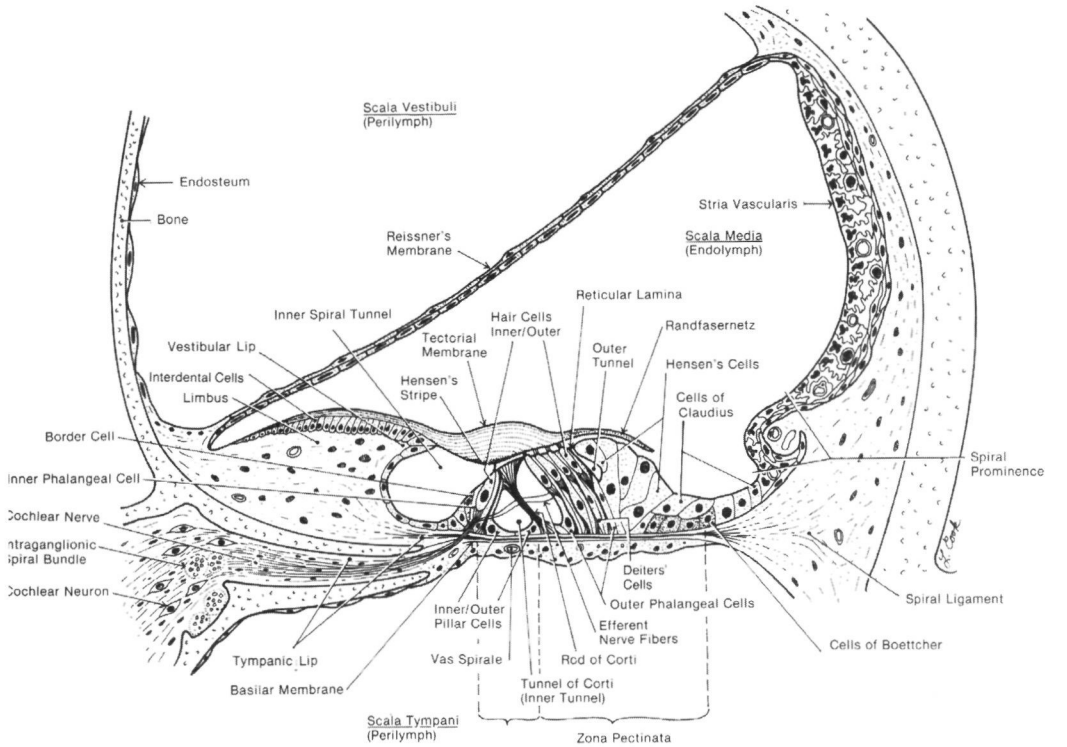


Figure 2. Cytologic structures of the cochlear duct.
(By courtesy of H.F.Schuknecht, from: Pathology of the Ear, Chapter 2, p.48, R.K.Bussy ed., Lea & Febiger publ, USA)

Loss of hair cells results in so-called sensorineural hearing loss (SNHL), which is one of the most prevalent disabilities in the world.[1] When the loss is severe, amplification with conventional hearing aids will not help the person to hear speech, as the auditory nerves that lead to the brain centres cannot be stimulated. To bypass the hair cells and provide information to the hearing centres of the central nervous system, the cochlear implant (CI) was developed. It is an electronic device that transforms acoustic vibrations electronically into an electrical current that directly stimulates the auditory nerve. Thus, a CI, also referred to as "the bionic ear", enables severely to profoundly deaf people who do not benefit from conventional amplification, to perceive sound, i.e. to hear.

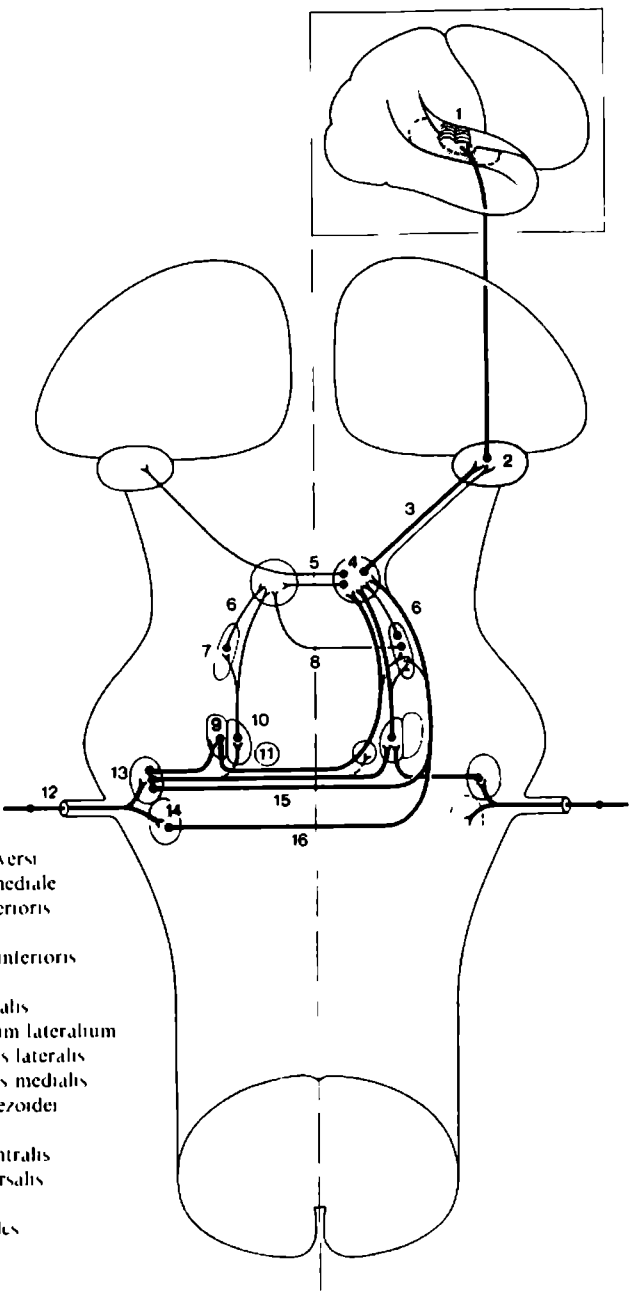


Figure 3 The central auditory pathway
The temporal and spatial patterns of excitation in the auditory nerve (12) are received in the cochlear nucleus (13,14) The information from the ventral and dorsal cochlear nucleus passes ipsilateral or contralateral through the above centres Further crossing-over takes place after which the auditory cortex is reached (1)
(By courtesy of Prof dr Nieuwenhuys from Das Zentral-nervensystem des Menschen Teil IV p 139 Springer-Verlag publ)

A CI consists of external and internal parts. The external parts include a microphone, speech processor and transmitter. The microphone is placed above the ear and worn like a hearing aid. It transduces the acoustic information of sound into electrical signals that are then sent to the speech processor. Depending on the processing strategy used, the speech processor transforms the input. The information is then transmitted across the skin electromagnetically to an implanted receiver-stimulator that decodes the signal and sends patterns of stimuli to the electrodes placed inside the cochlea (Figure 4).

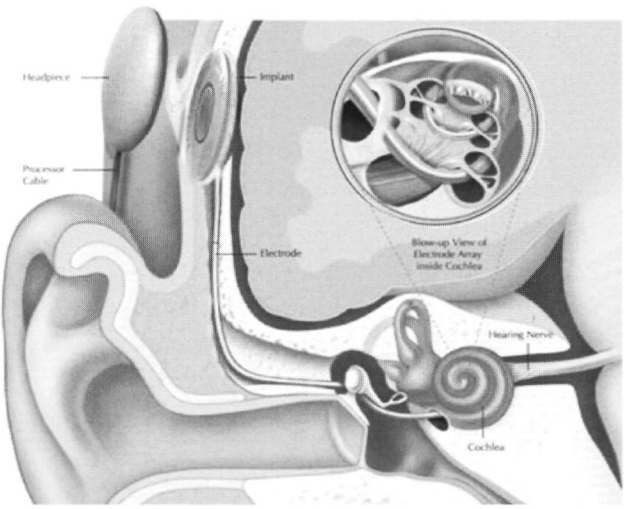


Figure 4. External and internal parts of a cochlear implant.
Blown-up view of an intracochlear electrode array.

1.2 History of cochlear implantation

In 1790, the idea of using electrical energy to produce hearing sensations was first put into practice by Alessandro Volta: he inserted metal rods into his ears and connected them to an electrical source. This caused him to lose consciousness, but he also remembered hearing bubbling noises in his ears. Many years later in France, around 1957, Djourno et al.[2] applied a single copper wire to the auditory nerve of a deaf man with a history mastoid surgery for cholesteatoma. An induction coil and an indifferent electrode were placed in the temporal muscle, while an active electrode was placed in the vestibule on a segment of the auditory nerve. When the coil was stimulated by induction currents, the subject reported hearing sounds like 'crickets'.

Despite the scepticism of many scientists about the practical feasibility of a CI in the 1960s, the field of cochlear implantation has grown from a small number of isolated experimental studies, to a diverse discipline investigated by many as a result of extensive

research Few medical advances have required the integration of so many disciplines as the CI it is the result of research into surgical anatomy, pathology, biology, biophysics, neurophysiology, psychophysics, speech science, engineering, surgery, audiology, rehabilitation, education and quality of life studies It could never have been foreseen that cochlear implantation would become such an important otological intervention

Since the 1960s, several types of CI devices have been developed They can be classified into extracochlear and intracochlear systems and further divided into single-channel and multi-channel systems Initially, it was feared that the placement of intracochlear devices would cause even more damage to the refined hearing organ in the cochlea Besides the possible effects of insertion trauma (e g loss of residual hearing[3]), other objections to intracochlear systems were the possible lack of biocompatibility of the device and, especially in children, the risk of middle ear infection spreading to the cochlea and meninges via the intracochlear electrode array Therefore, extracochlear systems were developed, in which the electrode was placed outside the cochlea, in the round window niche or on the promontory However, animal studies in Australia[4,5] and the United States[3] showed that the scala tympani, at the centre of the inner ear spiral, was the best place to stimulate the auditory nerve fibres connected to the different frequency regions of the brain Moreover, when electrode arrays with the correct mechanical properties were placed without excessive force, they did not cause any injury to the nerve fibres and neither did the current itself[6] The first commercially available intracochlear single-channel CI was developed by House and his group in Los Angeles [7] In this device, the entire speech signal was delivered to a single electrode located in the scala tympani Just one electrode in the single-channel systems was unable to transfer the spectral information needed to enable open speech recognition Later, multiple-electrode stimulation took advantage of the tonotopic organization of the cochlea In 1978, the first postlingually deaf volunteer was implanted with the multiple-channel electrode array developed at the University of Melbourne It consisted of 20 electrodes

Research aimed not only to achieve a larger number of electrodes in the cochlea, but also to optimize intracochlear placement of the electrode array Physiological data from animals[8] and modelling efforts[9] suggested that placement of the electrode array closer to the modiolus and spiral ganglion cells would result in more localized current flow and more effective stimulation of the neurons This would enhance discrimination ability and speech understanding and reduce power consumption Consequently, the Bionic Ear Institute at the University of Melbourne developed the Contour array, or perimodiolar, 'modiolus-hugging' electrode array [8,10] However, it has proven elusive to demonstrate the clinical efficacy, because some reports showed better speech recognition scores in Contour™ users, whereas others did not detect any difference in performance between Contour™ and straight electrode users [11]

As patients with labyrinthitis ossificans were no longer excluded from cochlear implantation, surgeons and researchers had to find ways to insert the electrode array as far as possible into an obliterated cochlea In labyrinthitis ossificans, the cochlear lumen becomes partly or completely filled with bone as a chronic stage of the healing process (for example after meningitis) In these patients, it is impossible to insert the full length of the electrode array, despite attempts to create a new lumen in the ossified cochlea by drilling Fairly recently, in collaboration with Cochlear Limited, a special implant has been developed, called the Nucleus double array implant It features two separate electrode arrays with 10 and 11 active electrodes, as well as a reference electrode on the receiver-stimulator package One electrode array can be placed into the hole drilled in the scala tympani of the basal turn, while the other can be placed in the scala vestibuli of the second turn Auditory test results in patients with a totally obliterated cochlea were significantly better as a result of an increased number of intracochlear electrodes [12]

To achieve even better speech understanding, several speech processing strategies have been developed for the different CI systems during more than 20 years of research These strategies present the complex speech patterns to the nervous system by electrical stimulation The first wearable speech processor was developed in 1979 [13] In the 1980s, various collaborations developed between research centres (University of Melbourne, House Ear Institute, Technical University of Vienna, University of California in San Francisco, University of Antwerp) and the industry (Cochlear Limited, 3M, Storz, Advanced Bionics, Philips, respectively) Thus, the 1980s was the decade in which CI research flourished and great progress was made Over the years, the development of new speech processing strategies has shown an almost linear rise in the speech perception scores of CI patients When cochlear implantation had become sufficiently efficacious in adults, research was extended to children Early results showed that the younger the age at implantation, the greater the improvement in performance [14] When implanting children of younger than two years, specific safety issues needed to be considered the effects of drilling on head growth, possible electrode extrusion in the long-term due to head growth, middle ear infection as a risk of causing bacterial meningitis and the effect of electrical stimulation on a maturing nervous system After several studies in the 1990s, these issues did not prove to be main causes of concern, provided that the electrode array entry point was properly sealed and implantation took place in the absence of middle ear infection [15 16]

1.3 Patient selection

To define criteria for implantation, it is necessary to establish when the advantages of implantation outweigh the disadvantages. In adults, preoperative evaluation for cochlear implantation aims to select patients with the highest probability of achieving better hearing than with their exiting (appropriate and optimally fitted) hearing aids. This requires careful assessment of preoperative speech perception and communication abilities. Further, the candidate must have realistic expectations about the benefits and risks and have adequate help available from family or social services to pursue the rehabilitation programme. When cochlear implantation was first introduced, it was only offered to the extreme cases, i.e. profoundly hearing impaired adults who did not derive any measurable benefit from conventional hearing aids. As a result of the highly positive outcomes, the criteria of candidacy have become more liberal.

To be able to predict the benefit of cochlear implantation, the results of cochlear implant recipients have been analysed extensively.[17-22] Several factors were found to influence performance in adults and children: age at onset of deafness, age at implantation, duration of deafness, duration of implant use, etiology of deafness, presence of progressive hearing loss, degree of residual hearing, speech reading ability and medical condition. Some of these factors influenced performance indirectly because of alterations to the anatomical, biophysical and biochemical properties of the central nervous system (CNS), nerves and cochlea. Hereby, these indirect factors can also influence the position of the electrode array in the cochlea and the number of active electrodes (Table 1).

Age at implantation correlated negatively with performance in adults only if the person was older than 60 years[23] and in children who were born deaf or became deaf early in life. In children, age at onset of deafness can be prelingual or postlingual. The former means that the child has had little or no language experience, which has proven to be a negative factor. This can be overcome by early implantation, which shortens the duration of deafness.[24] Another negative correlation was found with longer durations of deafness.[17,18] Several factors correlated positively with performance: the presence of some residual hearing[20], good speech-reading ability[10] and longer duration of implant use.[23]

General predictive factors that specifically apply to children are language level and communication mode, educational setting after surgery, parental support and cognitive and motor milestones. Language development was found to influence speech perception and vice versa. Children in auditory-oral communication programmes made better progress, whereas children with delayed cognitive and motor milestones took longer to learn and they did not achieve the same plateau of open-set speech recognition. Factors such as the extent of family support are hard to quantify, which makes the preoperative evaluation process even more complex.

Table 1. Factors that influence the performance of cochlear implant recipients

Direct factors		Indirect factors
Device	speech processor characteristics; coding strategy	
Electrodes	position in the cochlea; number in use	etiology of deafness (infection, ossification, demineralization, malformation, trauma, toxicity, genetics)
Cochlea	electrical properties; size; physical condition	etiology of deafness (ossification, demineralization, malformation, trauma, toxicity, genetics) age at implantation (natural degeneration)
Nerves	spiral ganglion cell survival; spiral ganglion cell function	etiology of deafness (ossification, demineralization, malformation, trauma, toxicity, infection) age at implantation (natural degeneration) duration of deafness (accelerated degeneration, auditory deprivation) duration of implant use (plasticity, learning)
CNS	central neural function; memory for spoken language; cognition	etiology of deafness (trauma, toxicity, infection, additional neurological disorders) age when deafened (maturation, plasticity, learning) age at implantation (natural degeneration) duration of deafness (accelerated degeneration, auditory deprivation) duration of implant use (plasticity, learning) communication mode (plasticity, learning)

The preoperative evaluation is undertaken jointly by the otologist and audiologist and also requires consultation from other specialists, such as a psychologist, speech pathologist, neurologist, social worker or general physician (CI team). The preoperative audiological assessment consists of pure-tone audiometry, middle ear impedance testing, speech perception and production tests and hearing aid evaluation. The otological evaluation involves medical history, standard ENT examination, high-resolution computed tomography (HRCT) and/or magnetic resonance imaging (MRI).

High-resolution CT and MRI of the temporal bone enable the surgeon to study the morphology of the temporal bone and identify any abnormalities that might affect the surgical procedure. The positions of surgical landmarks can be explored, such as the short incudal process, the vertical portion of the facial nerve, the oval and round windows, the carotid canal and the sigmoid sinus (Figure 5). Variations in morphology may be encountered, including abnormalities due to previous surgery, or pathological alterations caused by ossification (in meningitis and otosclerosis), spongiosis (in otosclerosis, Osteogenesis Imperfecta and Paget's disease) or congenital malformations. It is important to take the degree of ossification into account when considering which ear to implant. If there are any doubts, the surgeon can arrange to have a double array implant available at the operating theatre.

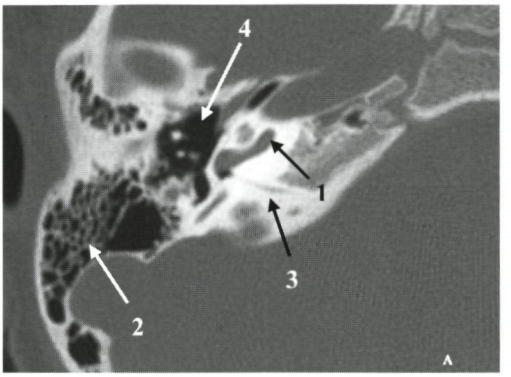


Figure 5A. Axial view of the normal petrosal bone
1. Basal turn of the cochlea; 2. Mastoid air cells;
3. cochlear aqueduct; 4. Tympanum

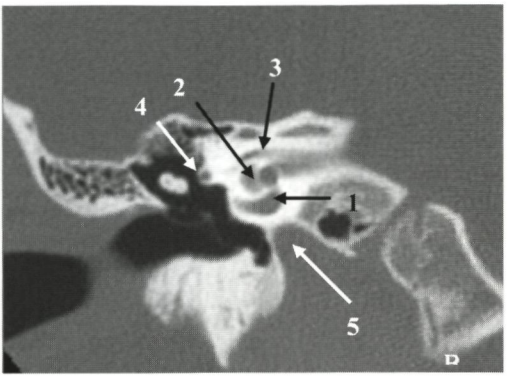


Figure 5B. Coronal view of the normal petrosal bone
1. Basal turn of the cochlea; 2. Middle turn of the cochlea; 3. Labyrinthine segment of the facial nerve; 4. Tympanic segment of the facial nerve; 5. Jugular bulb

Preoperative awareness of the morphology is crucial when there are congenital malformations in the inner ear: a modiolus-hugging device is best suited to the presence of a well-defined modiolus, whereas a straight array is preferred if the nerves are located peripherally in the cavity. The course of the facial nerve must be noted, because it is very likely to be aberrant in congenitally malformed ears (Table 2). Further, cochlear patency[25] and the presence of the cochlear nerve[26] must be evaluated. The presence of a cochlear nerve can be ascertained on MRI with gradient-echo techniques,[27] but also on HRCT based on the assumption that a normal cochlear nerve must be present when the cochlear nerve canal is normal.[28] MRI is the best technique to show *early* cochlear obliteration and central causes of hearing loss.[29] Bettman et al.[25] were more in favour of a CT protocol to scan the temporal bone of CI candidates preoperatively, whereas others[30,31] preferred MRI in the decision-making process of cochlear implantation.

Table 2a. Key points in preoperative imaging studies that lead to modification of the surgical strategy or device type.
Table 2b. Key points in preoperative imaging studies that imply an increased surgical risk.

a. Modification of surgical strategy or device type	b. Increased surgical risk
Cochlear ossification	Hypoplastic mastoid process
Hyperostosis of round window niche	Inflammation middle ear
Persistent membranous labyrinth inflammation	Dehiscent or aberrant facial nerve
Inner ear at risk of CSF gusher: • dilation of endolymphatic sac, semi-circular canal or vestibule • cochlear dysplasia	Mastoid emissary vein
Otosclerotic foci	Deep sigmoid sinus
M. Paget	Exposed jugular bulb
	Aberrant carotid artery
	Persistent stapedial artery

Recent research by Trimble et al.[32] has shown that although there is overlap in the type of abnormalities detected by HRCT and MR, preoperative dual-modality imaging of the petrosal bone and MRI of the brain in paediatric CI candidates can detect abnormalities related to the deafness that would not have been found using one of the modalities alone. They presented an algorithm to help select the best imaging modality, using the patient risk factors identified in their study.

1.4 Surgical implantation procedure

The surgical cochlear implantation procedure is similar in children and adults. Several modifications have been made to the skin incisions and approaches to the middle ear and cochlea.[33-35] In the Nijmegen/Viataal CI programme, the middle ear is reached via mastoidectomy and a facial recess approach, while the cochleostomy is made anterior and inferior to the round window. First, the position of the implant is marked on the skin with a fine needle and syringe that contains methylene blue. A single flap is created that comprises skin as well as superficial and deep fascia. The deeper periosteal flap is incised at a different location. After the landmarks of the mastoid bone have been exposed, a well is drilled to hold the receiver-stimulator. Then mastoidectomy and the facial recess approach are used to gain access to the middle ear and cochlea (Figure 6). A round eminence called the promontory is visible in the medial wall of the middle ear. This bulge contains the basal turn of the cochlea and lies in front of the oval and round windows. The upper portion of the basal turn lies under the tympanic/horizontal segment of the facial nerve, whereas the middle turn is more accessible. The round window is sealed by a membrane that overlies the scala tympani of the basal turn. To enable insertion of the electrode array into the scala tympani of the basal turn, the cochleostomy is made just anteroinferior to the round window (Figure 7). If

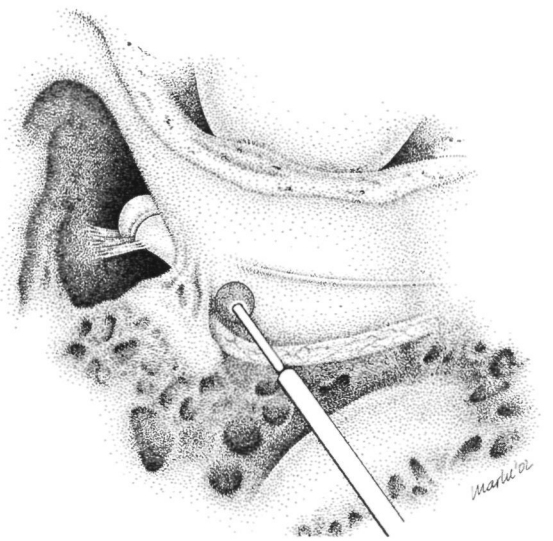


Figure 6. Site of the posterior tympanotomy after mastoidectomy.
(By courtesy of Prof. dr. C.W.R.J. Cremers)

resistance is felt during insertion, or the array is seen to buckle, it is important to stop immediatly: forceful insertion may damage the inner ear or electrodes.

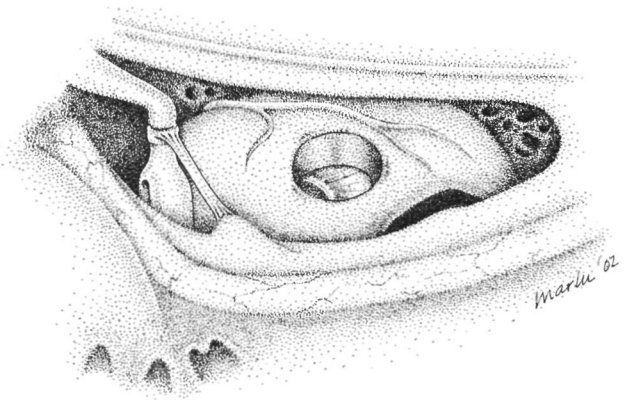


Figure 7. Cochleostomy
(By courtesy of Prof. dr. C.W.R.J. Cremers)

In case of ossification of the cochlear lumen, the location of the cochlear turns in relation to the medial wall of the middle ear are of particular importance, because drilling is necessary to create a new lumen and make (partial) insertion possible. When ossification is severe, the double array implant might be indicated. The first cochleostomy is routinely made anterior to the round window, which provides access to the basal turn (that contains the scala tympani and the scala vestibuli). Any connective tissue and bone can be removed until the anterior wall of the basal turn becomes visible. A second cochleostomy is performed in the second turn, caudal of the cochleariform process and 2 mm anterior of the oval window, after removal of the incus. Any excess tissue should also be removed. One of the electrode carriers is then placed into the scala tympani of the basal turn and the other into the scala vestibuli of the second turn. The remaining surgical procedure is identical to that used in patients with a patent cochlea. In patients with congenital malformation of the inner ear, the position of the facial nerve and cochlear windows may help to locate the scalae so that safe insertion can be performed.

The cochleostomy must be completely sealed circumferentially using fascial autograft or pericranium, to ensure that middle ear infection cannot spread to the inner ear, which would otherwise lead to labyrinthitis, or even meningitis. To prevent the electrode array from being extruded from the cochlea in growing children, it is fixed to the fossa incudis, because the distance from there to the round window does not change after birth.

Before sealing the cochleostomy, some CI centres perform intraoperative 3D rotational X-ray imaging to be certain that the electrode array is in the correct position.[36] In contrast,

most CI centres consider that intraoperative neural response measurements (NRTs) are sufficient in patients with normal petrosal bones on the preoperative CT scan and in whom the surgical procedure was uneventful. Even routine postoperative X-rays (Stenvers) to check the position of the electrode array[37] is a practice that some CI centres have abandoned: imaging might only be indicated in patients with abnormal postoperative clinical or electrophysical findings.

1.5 Rehabilitation

After recovery from CI surgery, the speech processor is fitted. To optimize the speech signal, threshold levels (T levels) and maximum comfortable levels (C levels) for electrical stimulation on each electrode are established and then programmed into the patient's speech processor, called the MAP. The electrical stimulation levels between T and C levels cover the dynamic range. The frequency boundaries of each electrode are also set to determine the pitch range per electrode. If the patient is not content with the sound perceived, the MAP can be fine-tuned. After programming, the patient attends training sessions in which he/she learns how to interpret the sensations created by the electrical stimulation. In adults, the rehabilitation mainly focuses on speech recognition, while in children the focus is broader: speech perception, speech production, receptive language and expressive language The speech material used in auditory training is age-appropriate and contains specific speech tokens, such as vowels and consonants, or sentences and words. Training exercises can also be used to asses the performance of the patient.
During rehabilitation, close attention must be paid to factors that predict the outcome, as described above. For example, patients with a long duration of deafness are more likely to require long rehabilitation to achieve adequate speech perception. Central nervous system pathology can influence learning abilities. Children with developmental delays and learning disorders may have poorer speech perception and progress at a slower rate.[38] Age at onset of deafness is also important, because there is a 'most sensitive period' in the development of language within the first few years of life. Early results of cochlear implantation in young children suggested that children with congenital deafness and children with early acquired deafness experienced similar benefit to adults with postlingual deafness and most of these children ultimately achieved open-set speech recognition.[39,40]

1.6 The Nijmegen/Viataal CI programme

1.6.1 History

In 1983, when there was much debate on the concept of cochlear implantation, an adult patient from the department of Otorhinolaryngology at the Radboud University Nijmegen Medical Centre, underwent cochlear implantation in Paris and was rehabilitated in Nijmegen.[41] In the Netherlands, the first cochlear implantation procedure in adults was performed in Utrecht in 1985, followed by Nijmegen in 1987. Initially, application of the technology was restricted to a government-sponsored study on 20 adult patients. The positive findings[42,43] led to the approval of cochlear implantation by the Dutch Minister of Health in 1997. Since then, the annual number of adult patients has risen (Figure 8). In 1989, the first child was implanted at the Radboud University Nijmegen, Medical Centre. A clinical study on cochlear implantation in children was conducted from 1993-1996.[44]

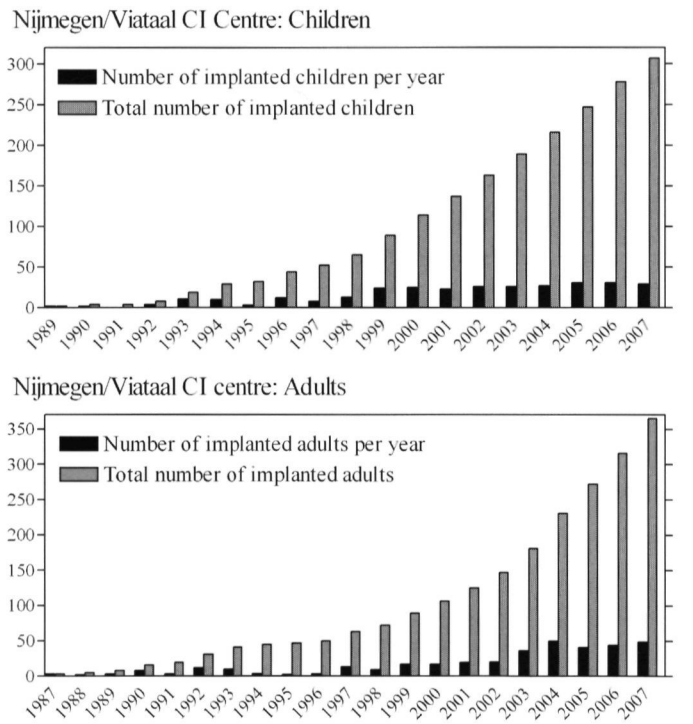


Figure 8. Number of patients implanted at the Nijmegen/Viataal CI centre.

The first child with congenital deafness was implanted in 1990 at 13 years of age. Implantation in the case of a congenitally malformed cochlea (Mondini's dysplasia) first took place in 1994 in a 7-year-old girl with congenital deafness. In 2001, the Nucleus Double array CI was used for the first time in a 6-year-old girl with postmeningitic

deafness and ossified cochleae. At the end of 1999, the Dutch Minister of Health approved cochlear implantation in prelingually deaf children.

Since the start of the programme, the Radboud University Nijmegen Medical Centre works in close collaboration with Viataal, the former Institute for the Deaf in St. Michielsgestel for the rehabilitation of implanted adults and children. Together they form the Nijmegen/Viataal CI Centre. At the end of 2007, 743 implantation procedures had been carried out in 392 adults and 351 children, including bilateral implantations and reimplantation procedures.
In order to stay experienced in the various CI systems produced by the industry, the Nijmegen/Viataal CI centre uses implants from more than one manufacturer. This also enables the CI team to make the best choice in individual patients based on audiological criteria as well as ease of handling. Table 3 shows a list of CI devices used in Nijmegen through the years and their characteristics.

1.6.2 Patient selection, preoperative evaluation procedure

As mentioned above, it is a complex procedure to determine whether a deaf patient is a suitable candidate for cochlear implantation. Over the years, the Nijmegen/Viataal CI team have broadened the criteria for cochlear implantation (for example with respect to duration of deafness, amount of residual hearing and presence of additional handicaps) based on increasing experience and proven benefit of cochlear implantation. This trend has also been observed in CI teams worldwide. Patients who had been advised against implantation in the past, were considered suitable candidates later. For example, adults with prelingual deafness are no longer excluded, because cochlear implantation can not only make valueable contributions to speech perception, but also to sound perception and quality of life. Therefor, these individuals undergo thorough evaluation during counseling. This can even lead to the successful implantation of patients with psychomotor retardation, which in Nijmegen was first performed in 2002.
The standard evaluation procedure includes tests on various factors that are known to influence postoperative performance (as described in paragraph 1.3). Relevant data are obtained by means of medical history, ENT physical examination, MRI, CT scanning, ENG, pure tone audiometry, speech perception tests, BAER and psychological assessments, as well as speech-language tests in children. In some cases, it may be necessary to perform supplementary measurements, such as Auditory Steady State Responses (ASSR), electro cochleography (ECoG) and electrically round window stimulation.

Table 3. CI devices used in the Nijmegen/Viataal CI programme between 1987 and 2005

Company name (device name / electrode array name)	Speech Processor	No. of electrodes	Basilar - apical electrode	Coding strategy	Electrode placement	Implanted subjects	Period of application
Vienna (3M)	3M	1	-	Analogue	Extra	Adult	1987-1992
Antwerp Bionics Systems (Laura)	Laura	8	8 - 1	CIS	Intra	Adult	1994-1996
Cochlear (Nucleus 22)	WSP/ MSP/ Spectra/ Esprit 22	22	1 – 22	F0-F1-F2/ MPEAK/ SPEAK	Intra	Adult/child	1989-1997
Cochlear (Nucleus 24)	Sprint/ Esprit 24	22+2	1 – 22	SPEAK/ CIS/ ACE	Intra	Adult/child	1997-2001
Cochlear (Nucleus 24 Double Array)	Sprint/ Esprit 24	2x 11	1 – 22	SPEAK/ CIS/ ACE	Intra	Adult/child	2001-present
Cochlear (Nucleus 24 Contour)	Sprint/ Esprit 24/ 3G	22+2	1 – 22	SPEAK/ CIS/ ACE	Intra	Adult/child	2001-2004
Cochlear (Nucleus 24 Contour Advanced electrode)	Sprint/ Esprit 24/ 3G	22+2	1 – 22	SPEAK/ CIS/ ACE	Intra	Adult/child	2003-2005
Cochlear (Nucleus Freedom Contour Advanced electrode)	Freedom	22+2	1 – 22	SPEAK/ CIS(RE)/ ACE(RE)	Intra	Adult/child	2005-present
Med-El (M1)	COM	1	-	Analogue	Extra	Adult/child	1989-1992
Med-El (Combi 40+)	Tempo+	12	12 - 1	CIS	Intra	Adult/child	1996-1997
Advanced Bionics (C I / Enhanced Bipolar)	S-series/ PSP / P-BTE	8	8 – 1	CIS/ CA	Intra	Adult	1997- 1999
Advanced Bionics (C I / HiFocus 1)	S-series/ PSP / P-BTE	8	8 – 1	CIS/ CA	Intra	Adult	1999-2001
Advanced Bionics (C II / HiFocus 1)	PSP / BTE-II/ Auria / Harmony	16	16 – 1	CIS/ CA / HiRes / F120	Intra	Adult	2003
Advanced Bionics (C II / HiFocus 2)	PSP/ BTE-II/ Auria / Harmony	16	16 – 1	CIS/ CA / HiRes / F120	Intra	Adult/child	2001-2003
Advanced Bionics (Clarion 90K / HiFocus 1)	PSP/ BTE-II/ Auria / Harmony	16	16 – 1	CIS/ CA / HiRes / F120	Intra	Adult	2003-present
Advanced Bionics (Clarion 90K / Helix)	PSP/ BTE-II/ Auria / Harmony	16	16 1	CIS/ CA / HiRes / F120	Intra	Adult	2004-present

WSP = Wearable speech processor, MSP = Mini speech processor, MPEAK = Multiple peak, SPEAK = Spectral peak, CIS = Continuous interleaved sampling; CA = Compressed analogue, ACE = Advanced Combined Encoder

1 6 3 Postoperative evaluation protocol

In adults, the speech processor is fitted 3 weeks after the implantation procedure ('week 0'). The necessary equipment is provided and checked. T and C levels are established and programmed into the speech processor (the MAP). Several fitting sessions take place in the first year postimplantation. Free-field audiometric data are collected at 5 weeks ('week 5') with the CI and in case of preoperative residual hearing, without the CI. These tests are repeated one year postoperatively ('week 52') and the patient's speech perception is also also evaluated: word discrimination (AN spondee test) and speech perception in quiet and in noise (word and phoneme scores on the open set NVA monosyllable word lists). At Viataal, the rehabilitation therapist assesses quality of life using the Nijmegen CI Questionnaires (NCIQ). After the first year of follow-up, the equipment is checked annually and pure tone audiometry, word discrimination and speech perception are measured alternatively by the audiologist in Nijmegen and at Viataal.

In children, rehabilitation takes place at Viataal. The first fitting session is performed about 3 to 6 weeks after the implantation surgery. The CI is then activated and assessments are made of the hearing sensations produced by the implant. One week after the first fitting session an extensive week of fitting and stimulation led by the audiologist and rehabilitation therapist takes place. During this week the child and his/her parents can stay at the Guest House of Viataal. Auditory development is monitored at 1, 3, 6, 9 and 12-months follow-up by means of audiometry, speech perception and speech production tests. The rehabilitation therapist also makes regular home school visits. After the first year of follow-up, annual measurements are performed with special attention to the development of spoken language.

1.6.4 Nijmegen/Viataal CI centre: Results

The speech perception scores at 1-year follow-up of 450 postlingually deaf adult CI patients, enrolled at the Nijmegen/Viataal CI programme and implanted with different CI systems, are shown in Figure 9. The average speech perception scores 1 year after implantation gradually improves through the years, although a wide variation in performance persists.

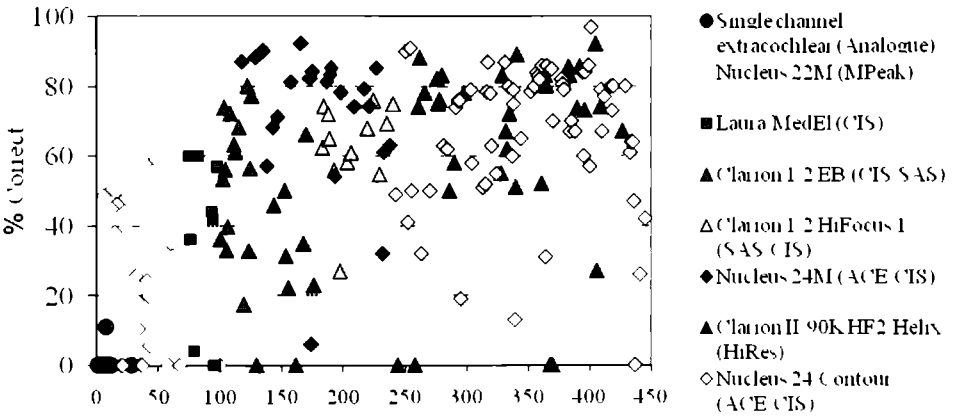


Figure 9 Monosyllable word recognition score (% correct) at 1-year follow-up of 450 consecutive implanted postlingually deaf adults, who used different types of CI devices and speech coding strategies

1.7 Aim of the thesis

Since the introduction of cochlear implantation, efforts have been made to explain the variability in outcomes in order to improve the results and select patients who are most likely to benefit from the intervention Several factors have been found to influence the performance of a patient with a CI When it became apperent that factors such as age at implantation and duration of deafness were important, research extended to the implantation of children In **Chapter 2**, the speech perception performance of 67 children implanted at the Nijmegen CI centre between 1986 and 1999, was evaluated by means of a broad battery of speech perception tests At that time, the criteria for implantation were still comparatively strict, which resulted in a fairly homogenous group of children To deal with the bottom and ceiling scores that inevitably occur when a broad battery of speech perception tests is used for the follow-up of children at different ages and developmental stages, the different speech perception results were reduced into one measure the "equivalent hearing loss" (EHL) [45] This outcome measure refers to the performance of a reference group of severely and profoundly hearing impaired children who were using conventional hearing aids An attempt was made to explain the variability in long-term post-implant speech perception performance on the basis of several factors, such as age at implantation, duration of deafness and communication mode

Relaxation of the criteria for cochlear implantation means that CI teams are being confronted by more and more etiologies of deafness that were once considered to be contraindications for cochlear implantation Etiology of deafness is important because it is another predictive factor which has been recognized to more or less influence the

performance of patients with a CI.[21,23] Changed histopathology of the cochlea can cause SNHL and altered electrical properties of the temporal bone, ganglion cell number and central neural survival or function, which are known to influence auditory performance with a CI. Diseases that alter the morphology of the cochlea and otic capsule are especially likely to compromise cochlear implantation and affect the number of active electrodes and their positions.

Meningitis is an important disease to consider in cochlear implantation, as it may cause deafness requiring cochlear implantation, as well be a rare complication of cochlear implantation itself (device-related meningitis). Infection may spread from the meninges to the cochlea through the cochlear aqueduct and foramina in the osseus labyrinth and cause labyrinthitis, but it can also spread in the reverse direction when it originates from the middle ear or inner ear. Meningogenic labyrinthitis ossificans is part of the healing phase after inflammation; this new bone formation often destroys stimulable spiral ganglion cells,[46] which is known to have a negative influence on the outcome of cochlear implantation. In addition, obliteration of the cochlea may require special surgical implantation techniques, such as various degrees of drilling,[47,48] alternative placement of the electrode array in the scala vestibuli or extracochlearly,[49,50] or the use of double array implants.[12] However, despite these surgical techniques to deal with ossification of the cochlea, in some patients only partial insertion of the electrode array can be achieved. To evaluate the effect of such partial insertion on postoperative performance with a CI, **Chapter 3** explored the outcome of children with postmeningitic deafness and partial insertion of the electrode array due to ossification of the cochlea and compared the results to those of children with postmeningitic deafness and full insertion of the electrode array.

In the early days of paediatric cochlear implantation, the majority of children had acquired, postmeningitic bilateral sensorineural deafness. Nowadays, increasing numbers of children scheduled for cochlear implantation have congenital deafness. Between 20-30% of the congenital cases have abnormalities of the bony labyrinth.[51,52] Malformations of the cochlea were classified by Jackler et al.[51] based on embryonic life. They vary from total aplasia, severe cochlear hypoplasia, mild cochlear hypoplasia, common cavity, severe incomplete partition, mild incomplete partition, to a subnormal cochlea that doesn't reach the full 2.5 turns. Besides variable functioning and possibly tonotopically disorganized cochlear neuroepithelium, some inner ear malformations are associated with aplasia or hypoplasia of the cochlear nerve. In cases with insufficient nerve fibres, or none at all, a cochlear implant will be of no benefit.[53] Other challenges during the surgical implantation procedure are an anomalous course of the facial nerve and the occurrence of cerebrospinal fluid (CSF) gusher.[54,55] Specific problems can also occur postoperatively, during activation

and programming [54 56 57] **Chapter 4** addressed the surgical aspects and performance outcome of cochlear implantation in children with malformed inner ears

Over the past few years, an increasing number of adults with an etiology of otosclerosis (7 to 9 5%) received a CI [58] Otosclerosis is a hereditary disease that only affects the bony structure of the temporal bone In the active phase (otospongiosis), the normal lamellar bone in the middle layer of the otic capsule is resorbed and replaced by thick, irregular bone (otosclerosis) [59] The subsequent hearing loss can be conductive, commonly due to stapes fixation, or sensorineural if the cochlea is involved SNHL in otosclerosis is thought to be the result of narrowing of the cochlear lumen with distortion of the basilar membrane,[60] or it may be induced by toxic enzymes that are released into the perilymph from otosclerotic foci [61 62] Histological studies have shown that otosclerosis has a relatively small effect on spiral ganglion cell survival compared to other causes of deafness [63] If treatment such as stapedectomy or stapedotomy fails, the patient may become a candidate for CI surgery, but there is a risk that the surgical implantation procedure will be hindered by obliteration of the round window or basal turn Alternatively, the otic capsule might be softened by otospongiosis so that it is easily penetrated when the electrode array is pushed forward To evaluate the feasibility of cochlear implantation in deaf patients with an etiology of otosclerosis, **Chapter 5** mapped the special features that occurred during surgery and rehabilitation in a group of 53 otosclerosis patients who received a CI

Another hereditary bone disease that can involve severe SNHL is Osteogenesis Imperfecta Osteogenesis Imperfecta is a heterogeneous disease of the connective tissue and bone matrix, caused by defective collagen type I production Hearing loss affects 35-60% of the patients with Osteogenesis Imperfecta and is mostly conductive or mixed [64] The sensorineural component is believed to be the result of abnormal bone encroaching on the cochlea, which leads to mechanical distortion of the basilar membrane, tiny fractures of the otic capsule, haemorrhage into the labyrinth, otosclerotic foci that steal blood from the cochlear microcirculation and interference with the mechano-electric function of hair cells by toxic enzymes [65 66] As the hearing loss progresses to deafness in 2-11% of patients with Osteogenesis Imperfecta,[66 67] cochlear implantation may be the only remaining treatment option in some cases At the Nijmegen/Viataal CI centre, 3 patients with Osteogenesis Imperfecta have been implanted and enrolled in the CI rehabilitation programme **Chapter 6** focused on the specific problems encountered during cochlear implantation surgery and rehabilitation in these patients To evaluate whether the affected temporal bones had lower electrical resistance and to gain an impression of the possible consequences, psychoacoustic, electrical and electrophysiological measurements were performed

References

1 Niparko JK. The epidemiology of hearing loss: how prevalent is hearing loss? In: Niparko J, editor. Cochlear Implants: principles and practices.Philadelphia· Lippincott Williams and Wilkins; 2000. p. 88-92.

2. Djourno A, Eyries C, Vallancien B. Electric excitation of the cochlear nerve in man by induction at a distance with the aid of micro-coil included in the fixture. *C R Seances Soc Biol Fil* 1957,151:423-5.

3. Simmons FB. Permanent intracochlear electrodes in cats, tissue tolerance and cochlear microphonics. *Laryngoscope* 1967,77:171-86.

4. Clark GM Hearing due to electrical stimulation of the auditory system *Med J Aust* 1969;1.1346-8.

5 Black RC, Clark GM. Differential electrical excitation of the auditory nerve *J Acoust Soc Am* 1980;67.868-74.

6 Shepherd RK, Clark GM, Black RC. Chronic electrical stimulation of the auditory nerve in cats. Physiological and histopathological results *Acta Otolaryngol Suppl* 1983;399·19-31.

7. House WF, Berliner KI. Safety and efficacy of the House/3M cochlear implant in profoundly deaf adults *Otolaryngol Clin North Am* 1986;19:275-86.

8. Shepherd RK, Hatsushika S, Clark GM. Electrical stimulation of the auditory nerve: the effect of electrode position on neural excitation *Hear Res* 1993;66·108-20.

9. Frijns JH, de Snoo SL, ten Kate JH. Spatial selectivity in a rotationally symmetric model of the electrically stimulated cochlea *Hear Res* 1996;95:33-48.

10. Staller S, Parkinson A, Arcaroli J, Arndt P. Pediatric outcomes with the nucleus 24 contour North American clinical trial. *Ann Otol Rhinol Laryngol Suppl* 2002;189:56-61.

11. Fitzgerald MB, Shapiro WH, McDonald PD, Neuburger HS, Ashburn-Reed S, Immerman S, Jethanamest D, Roland JT, Svirsky MA. The effect of perimodiolar placement on speech perception and frequency discrimination by cochlear implant users *Acta Otolaryngol* 2007;127:378-83

12 Lenarz T, Lesinski-Schiedat A, Weber BP, Issing PR, Frohne C, Buchner A, Battmer RD, Parker J, von Wallenberg E. The nucleus double array cochlear implant: a new concept for the obliterated cochlea. *Otol Neurotol* 2001,22:24-32

13. Tong YC, Clark GM, Seligman PM, Patrick JF. Speech processing for a multiple-electrode cochlear implant hearing prosthesis. *J Acoust Soc Am* 1980;68:1897-8.

14. Clark GM, Blamey PJ, Busby PA, Dowell RC, Franz BK, Musgrave GN, Nienhuys TG, Pyman BC, Roberts SA, Tong YC. A multiple-electrode intracochlear implant for children. *Arch Otolaryngol Head Neck Surg* 1987;113·825-8.

15 Luxford WM, House WF. Cochlear implants in children: medical and surgical considerations. *Ear Hear* 1985,6 20S-3S.

16. Dahm MC, Clark GM, Franz BK, Shepherd RK, Burton MJ, Robins-Browne R. Cochlear implantation in children: labyrinthitis following pneumococcal otitis media in unimplanted and implanted cat cochleas *Acta Otolaryngol* 1994;114:620-5.

17. Gantz BJ, Tyler RS, Knutson JF, Woodworth G, Abbas P, McCabe BF, Hinrichs J, Tye-Murray N, Lansing C, Kuk F. Evaluation of five different cochlear implant designs: audiologic assessment and predictors of performance. *Laryngoscope* 1988;98.1100-6.

18. Blamey PJ, Pyman BC, Gordon M, Clark GM, Brown AM, Dowell RC, Hollow RD. Factors predicting postoperative sentence scores in postlinguistically deaf adult cochlear implant patients *Ann Otol Rhinol Laryngol* 1992;101 342-8.

19. Cohen NL, Waltzman SB, Fisher SG. A prospective, randomized study of cochlear implants The Department of Veterans Affairs Cochlear Implant Study Group. *N Engl J Med* 1993,328:233-7.

20 Gantz BJ, Woodworth GG, Knutson JF, Abbas PJ, Tyler RS Multivariate predictors of audiological success with multichannel cochlear implants. *Ann Otol Rhinol Laryngol* 1993,102:909-16

21. Battmer RD, Gupta SP, Allum-Mecklenburg DJ, Lenarz T. Factors influencing cochlear implant perceptual performance in 132 adults. *Ann Otol Rhinol Laryngol Suppl* 1995;166:185-7.
22 Shipp DB, Nedzelski JM. Prognostic indicators of speech recognition performance in adult cochlear implant users: a prospective analysis. *Ann Otol Rhinol Laryngol Suppl* 1995;166:194-6.
23. Blamey P, Arndt P, Bergeron F, Bredberg G, Brimacombe J, Facer G, Larky J, Lindstrom B, Nedzelski J, Peterson A, Shipp D, Staller S, Whitford L Factors affecting auditory performance of postlinguistically deaf adults using cochlear implants. *Audiol Neurootol* 1996;1:293-306
24. Snik AF, Makhdoum MJ, Vermeulen AM, Brokx JP, van den Broek P. The relation between age at the time of cochlear implantation and long-term speech perception abilities in congenitally deaf subjects. *Int J Pediatr Otorhinolaryngol* 1997;41:121-31
25 Bettman R, Beek E, van Olphen A, Zonneveld F, Huizing E MRI versus CT in assessment of cochlear patency in cochlear implant candidates *Acta Otolaryngol* 2004,124:577-81.
26. Maxwell AP, Mason SM, O'Donoghue GM. Cochlear nerve aplasia: its importance in cochlear implantation. *Am J Otol* 1999;20:335-7.
27 Casselman JW, Offeciers FE, Govaerts PJ, Kuhweide R, Geldof H, Somers T, D'Hont G. Aplasia and hypoplasia of the vestibulocochlear nerve: diagnosis with MR imaging. *Radiology* 1997;202:773-81.
28. Stjernholm C, Muren C Dimensions of the cochlear nerve canal: a radioanatomic investigation. *Acta Otolaryngol* 2002;122:43-8
29 Nikolopoulos TP, O'Donoghue GM, Robinson KL, Holland IM, Ludman C, Gibbin KP. Preoperative radiologic evaluation in cochlear implantation. *Am J Otol* 1997;18:S73-4.
30 Parry DA, Booth T, Roland PS. Advantages of magnetic resonance imaging over computed tomography in preoperative evaluation of pediatric cochlear implant candidates. *Otol Neurotol* 2005;26.976-82.
31 Ellul S, Shelton C, Davidson HC, Harnsberger HR Preoperative cochlear implant imaging: is magnetic resonance imaging enough? *Am J Otol* 2000;21:528-33.
32 Trimble K, Blaser S, James AL, Papsin BC. Computed tomography and/or magnetic resonance imaging before pediatric cochlear implantation? Developing an investigative strategy. *Otol Neurotol* 2007;28.317-24
33 Aschendorff A, Jackel K, Schipper J, Maier W, Laszig R, Klenzner T. The freiburg incision for cochlear implantation -- initial results. *Laryngorhinootologie* 2005,84 408-11
34 Pau HW, Sievert U, Graumuller S, Wild E. Incisions for cochlear implant flaps and superficial skin temperature. Skin temperature/blood circulation in CI flaps. *Otolaryngol Pol* 2004;58.713-9.
35. Telian SA, El-Kashlan HK, Arts HA. Minimizing wound complications in cochlear implant surgery *Am J Otol* 1999;20:331-4
36. Carelsen B, Grolman W, Tange R, Streekstra GJ, van Kemenade P, Jansen RJ, Freling NJ, White M, Maat B, Fokkens WJ. Cochlear implant electrode array insertion monitoring with intra-operative 3D rotational X-ray *Clin Otolaryngol* 2007,32:46-50.
37. Xu J, Xu SA, Cohen LT, Clark GM. Cochlear view: postoperative radiography for cochlear implantation. *Am J Otol* 2000,21:49-56.
38. Pyman B, Blamey P, Lacy P, Clark G, Dowell R. The development of speech perception in children using cochlear implants. effects of etiologic factors and delayed milestones. *Am J Otol* 2000;21:57-61.
39. Dawson PW, Blamey PJ, Rowland LC, Dettman SJ, Clark GM, Busby PA, Brown AM, Dowell RC, Rickards FW. Cochlear implants in children, adolescents, and prelinguistically deafened adults. speech perception. *J Speech Hear Res* 1992,35:401-17.
40. Staller SJ, Dowell RC, Beiter AL, Brimacombe JA. Perceptual abilities of children with the Nucleus 22-channel cochlear implant. *Ear Hear* 1991,12:34S-47S.
41. Cremers CW, Brokx JP. Cochleaire implantaten. *Ned Tijdschr Geneeskd* 1987,131.735-7

42. van den Broek P, Brokx JP, van Olpen AF. Selectie van volwassen doven voor een elektrische binnenoorprothese (cochlear implant) en evaluatie van de met deze prothese verkregen resultaten. University Hospital Nijmegen; 1995
43. Hinderink JB, Mens LH, Brokx JP, van den Brock P. Performance of prelingually and postlingually deaf patients using single-channel or multichannel cochlear implants. *Laryngoscope* 1995;105.618-22.
44. van den Broek P, Brokx JPL, Baker AE. Cochleaire implantatie bij kinderen. University Hospital Nijmegen; 1996
45. Vermeulen AM, Snik AF, Brokx JPL, van den Broek P, Geelen CP, Beijk CM. Comparison of speech perception performance in children using a cochlear implant with children using conventional hearing aids, based on the concept of "equivalent hearing loss". *Scand Audiol Suppl* 1997;47:55-7.
46. Hinojosa R, Redleaf MI, Green JD, Jr , Blough RR. Spiral ganglion cell survival in labyrinthitis ossificans: computerized image analysis. *Ann Otol Rhinol Laryngol Suppl* 1995;166:51-4.
47. Cohen NL, Waltzman SB. Partial insertion of the nucleus multichannel cochlear implant: technique and results. *Am J Otol* 1993,14:357-61.
48. Rauch SD, Herrmann BS, Davis LA, Nadol JB, Jr. Nucleus 22 cochlear implantation results in postmeningitic deafness. *Laryngoscope* 1997,107:1606-9.
49. Gantz BJ, McCabe BF, Tyler RS. Use of multichannel cochlear implants in obstructed and obliterated cochleas *Otolaryngol Head Neck Surg* 1988;98:72-81.
50. Steenerson RL, Gary LB, Wynens MS. Scala vestibuli cochlear implantation for labyrinthine ossification. *Am J Otol* 1990;11:360-3.
51. Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear: a classification based on embryogenesis *Laryngoscope* 1987,97:2-14.
52. McClay JE, Tandy R, Grundfast K, Choi S, Vezina G, Zalzal G, Willner A. Major and minor temporal bone abnormalities in children with and without congenital sensorineural hearing loss. *Arch Otolaryngol Head Neck Surg* 2002;128:664-71.
53 Shelton C, Luxford WM, Tonokawa LL, Lo WW, House WF. The narrow internal auditory canal in children. a contraindication to cochlear implants *Otolaryngol Head Neck Surg* 1989;100:227-31.
54. Hoffman RA, Downey LL, Waltzman SB, Cohen NL. Cochlear implantation in children with cochlear malformations. *Am J Otol* 1997;18:184-7.
55. Graham JM, Phelps PD, Michaels L. Congenital malformations of the ear and cochlear implantation in children. review and temporal bone report of common cavity *J Laryngol Otol Suppl* 2000;25:1-14.
56. Luntz M, Balkany T, Hodges AV, Telischi FF. Cochlear implants in children with congenital inner ear malformations. *Arch Otolaryngol Head Neck Surg* 1997;123:974-7.
57. Tucci DL, Telian SA, Zimmerman-Phillips S, Zwolan TA, Kileny PR. Cochlear implantation in patients with cochlear malformations. *Arch Otolaryngol Head Neck Surg* 1995;121:833-8.
58 Woolford TJ, Roberts GR, Hartley C, Ramsden RT. Etiology of hearing loss and cochlear computed tomography: findings in preimplant assessment. *Ann Otol Rhinol Laryngol Suppl* 1995,166.201-6
59. Guneri EA, Ada E, Ceryan K, Guneri A. High-resolution computed tomographic evaluation of the cochlear capsule in otosclerosis: relationship between densitometry and sensorineural hearing loss. *Ann Otol Rhinol Laryngol* 1996,105:659-64.
60 Linthicum FH, Jr., Filipo R, Brody S Sensorineural hearing loss due to cochlear otospongiosis· theoretical considerations of etiology. *Ann Otol Rhinol Laryngol* 1975;84: 544-51.
61. Linthicum FH, Jr Histopathology of otosclerosis. *Otolaryngol Clin North Am* 1993;26:335-52.
62. Youssef O, Rosen A, Chandrasekhar S, Lee HJ Cochlear otosclerosis the current understanding. *Ann Otol Rhinol Laryngol* 1998,107.1076-9.

63. Nadol JB, Jr., Young YS, Glynn RJ. Survival of spiral ganglion cells in profound sensorineural hearing loss: implications for cochlear implantation. *Ann Otol Rhinol Laryngol* 1989;98:411-6
64. Garretsen AJ, Cremers CW, Huygen PL. Hearing loss (in nonoperated ears) in relation to age in osteogenesis imperfecta type I. *Ann Otol Rhinol Laryngol* 1997;106:575-82
65. Pedersen U. Osteogenesis imperfecta clinical features, hearing loss and stapedectomy. Biochemical, osteodensitometric, corneometric and histological aspects in comparison with otosclerosis *Acta Otolaryngol Suppl* 1985;415:1-36.
66. Tabor EK, Curtin HD, Hirsch BE, May M. Osteogenesis imperfecta tarda: appearance of the temporal bones at CT. *Radiology* 1990;175:181-3.
67. Pedersen U. Hearing loss in patients with osteogenesis imperfecta A clinical and audiological study of 201 patients. *Scand Audiol* 1984;13:67-74

Chapter 2

Cochlear implantation in children

Speech perception in congenitally, prelingually and postlingually deaf children expressed in an Equivalent Hearing Loss value

L.J.C. Rotteveel
A.F.M. Snik
A.M. Vermeulen
C.W.R.J. Cremers
E.A.M. Mylanus

Submitted 2008

Abstract

Objectives To investigate the speech perception performance of children with a cochlear implant (CI) after 3 and 4 years of follow-up and to study the influence of age at implantation, duration of deafness and communication mode on the variability in speech perception performance

Study design A broad battery of speech perception tests was administered to 67 children with a CI The results were reduced into one measure the "equivalent hearing loss" This outcome measure refers to the performance of a reference group of severely and profoundly hearing impaired children with conventional hearing aids

Patients The population comprised 35 congenitally, 17 prelingually and 15 postlingually deaf children implanted between 1986 and 1999 The population was homogeneous with respect to cognition, residual hearing and support at home as a result of conservative inclusion criteria

Results During the first 2 years after implantation, postlingually deaf children showed the fastest rate of improvement After 3 years of implant use, the early implanted prelingually deaf children and congenitally deaf children implanted under the age of 6 years caught up with the postlingually deaf children Prelingually deaf children implanted after a relatively long duration of deafness tended to show poorer performance than those with a shorter duration Performance of congenitally deaf children implanted after the age of 6 years was poorer and progress was slower In the congenitally deaf children, 36% of the variability in performance was explained by the duration of deafness, whereas in the children with acquired deafness, communication mode and age at onset of deafness explained 71% of the variance

Conclusions All children derived benefit from their CI for speech perception tasks, but performance varied greatly Several children reached EHL levels around 70 dB, their speech perception was equal to that of a child with conventional hearing aids who has 70 dB HL After early implantation, the levels of performance that were eventually achieved differed no more than 10 dB, irrespective of whether the onset of deafness was prelingual or postlingual In congenitally deaf children, the duration of deafness played a major role in speech perception performance, whereas in children with acquired deafness, communication was a major factor

Introduction

Research and clinical experience have established that profoundly deaf children derive substantial benefit from multichannel cochlear implants.[1-3] The main goal of implantation in children is to improve hearing and consequently spoken language development. Improvement in speech recognition with a cochlear implant (CI) depends on several factors that are directly or indirectly associated with the functionality of the auditory system. In the past years these factors are being identified and are still reason for debate. They include duration of deafness[4], age at onset of deafness, age at implantation[5,6], duration of implant use, length of daily device use[7], cause of hearing loss[5,8] and preoperative level of residual hearing.[9] Other relevant factors are related with the CI device, such as the number of active electrodes[4], device type[10], speech processing strategy[10], mode of stimulation[11] and individual factors, such as cognition[8], motivation, support at home[12], communication mode[5,13] and educational setting.[5,12,14]

In the early nineties, inclusion criteria for cochlear implantation in the Netherlands were conservative, which resulted in a fairly homogenous group of children who had no residual hearing, normal cognition, no known learning disabilities, good motivation and support at home and no suspicion of any retrocochlear involvement. Factors related with the CI device and strategy were also fairly homogeneous. However, these children form a diverse group concerning factors such as the duration of deafness, cause and age at onset of deafness and communication mode. During the late nineties, inclusion criteria with respect to e.g. residual hearing, cognitive ability and age at implantation changed. In addition, several new and different types of implant and coding strategy became available, which contributed to increasing diversity. Thus, the group of children implanted during the early nineties is unique. Their data enable us to study the relation between a limited number of variables (e.g. age at onset, duration of deafness, communication mode) and long-term speech recognition results, while ignoring other factors, because they can be considered as homogeneous owing to conservative inclusion criteria (e.g. residual hearing, cognition, device type, support at home). The present study investigated these relations in 67 consecutive children who underwent implantation in Nijmegen between 1986 and 1999. The results at 3 years follow-up were used in a multivariate analysis to establish the effect of age at onset of deafness, duration of deafness, communication mode and educational setting on postimplant performance. The main reasons for using the data at 3 years follow-up were that almost all (n = 60) the children were still at the same school 3 years after implantation and were using the same type of communication mode (either primarily aural-oral communication or primarily sign language) as they had been prior to the implantation. After 3 years, many of the children were mainstreamed or placed at special schools for children with a severe hearing impairment (not profoundly deaf). Because in the majority of children the communication mode remained the same throughout the 3-year study

period, communication mode can be considered as a variable in the population of implanted children Furthermore, at the 3-year evaluation point, most of the children (n = 56) were still using the MPEAK (multiple peak) or SPEAK (spectral peak) speech processing strategy A limited group of 11 children had converted to ACE (Advanced Combined Encoder) strategy Research has shown that the differences in overall performance after a change from SPEAK to ACE are relatively small [15]

The cause of deafness was not included in the statistical multivariate analysis due to the small numbers of patients with specific pathologies

In this report, we present longitudinal data on speech perception in a group of children who differed only on a limited number of aspects Although speech perception only partially reflects improvements in speech and language development, or the psychosocial and intellectual development of a child, it is probably the most direct measure of the benefit a child derives from a CI

Methods

Subjects

The study group comprised 67 deaf children whose evaluation data were available over a period of at least 3 years after receiving a Nucleus multichannel cochlear implant at the Radboud University Nijmegen Medical Centre between 1986 and 1999 Eight additional children were not included because of partial insertion of the electrode array, as this may lead to poor results [16] Thirteen other children were not included because no measurements at the 3 year follow-up interval were performed due to moving house or poor physical condition

All the subjects were profoundly deaf, with hearing thresholds at 1, 2 and 4 kHz that exceeded 110 dB HL Psychological tests performed as part of the selection procedure were within the range of normal nonverbal intelligence

Group demographics are shown in Table 1 Thirty-five children were born deaf with etiologies of deafness that ranged from pre- or perinatal infection, anatomical malformations of the inner ear, to hereditary forms of deafness (6 children have the Usher syndrome) and unknown reasons These congenitally deaf children were grouped by age at implantation (i e duration of deafness), according to arbitrarily chosen limits (before the age of 4 years, between 4 and 6 years of age and older than 6 years at implantation) as this factor is known to influence speech perception abilities [6]

Thirty-two children with an acquired form of deafness were grouped according to the age at which deafness had occurred Children whose onset of an acquired form of deafness

occurred before the age of 2 years were classified as prelingually deaf, whereas if they had been older than 2 years at the time of onset of deafness, they were classified as postlingually deaf.

Sixteen out of the 17 prelingually deaf children had suffered from meningitis, while in one case the cause of deafness was unknown. The mean age at onset of deafness was 0.8 years. The duration of deafness was defined as the period between onset of deafness and cochlear implantation. In 10 children, the duration of deafness was longer than 3 years, while the remaining 7 children had received a CI within a period of deafness less than 3 years.

All 7 postlingually deaf children whose duration of deafness was longer than 3 years had suffered from meningitis (mean age at onset of deafness 4.2 years). The subgroup of postlingually deaf children whose duration of deafness was shorter than 3 years comprised 6 children with a history of meningitis and 2 children with an enlarged vestibular aqueduct (mean age at onset 5.0 years).

Table 1. Group demographics

Group, duration of deafness (years)	n	Mean age at implantation in years (range)	Mean duration of deafness in years (range)	Causes of deafness
Congenital < 4	13	3.1 (1.4 – 3.8)	3.1 (1.4 – 3.8)	5 hereditary, 1 CMV infection, 1 infection/dysmaturity, 1 dysplasia of inner ear, 5 unknown
Congenital 4-6	8	5.0 (4.3 – 5.7)	5.0 (4.3 – 5.7)	4 hereditary, 1 prematurity, 1 Mondini's malformation, 2 unknown
Congenital > 6	14	8.4 (6.9 – 13.5)	8.4 (6.9 – 13.5)	9 hereditary (6 Usher's syndrome), 1 rubella infection, 4 unknown
Prelingual < 3	7	3.0 (2.2 – 3.6)	2.0 (1.4 – 2.4)	7 meningitis
Prelingual > 3	10	6.5 (3.3 – 11.4)	5.7 (3.1 – 9.8)	9 meningitis, 1 unknown
Postlingual < 3	8	7.0 (4.3 – 13.9)	2.0 (0.7 – 2.5)	6 meningitis, 2 EVA
Postlingual > 3	7	9.0 (5.6 – 12.3)	4.8 (3.2 – 7.8)	7 meningitis

CMV = cytomegalovirus; EVA = enlarged vestibular aqueduct

Communication mode

The main communication mode of the children varied from aural-oral communication to primarily sign language. They had received different types and quantities of auditory stimulation and training. Distinction was made between children predominantly using oral communication and those solely using sign language (Table 2). We defined oral communication as communication through audition and/or speech reading, whether or not in combination with speech supporting signs. In the Netherlands, there are separate schools

for hearing impaired children and deaf children. Schools for hearing impaired children are mostly oral-aural oriented, with speech supporting signs. Most Dutch schools for the deaf use sign language, while some have a bilingual approach in which sign language and spoken language are taught separately. Children at the latter school who did not use oral communication outside the lessons were classified in the "solely sign language" group.

Table 2. Communication mode before and after implantation, number of subjects per subgroup

Group, duration of deafness (years)	Before implantation		3 years after implantation	
	Oral communication	Sign language	Oral communication	Sign language
Congenital < 4	5	8	6	7
Congenital 4-6	4	4	5	3
Congenital > 6	7	7	6	8
Prelingual < 3	5	2	6	1
Prelingual > 3	5	5	5	5
Postlingual < 3	7	1	8	0
Postlingual > 3	3	4	5	2
Total	36	31	41	26

Audiometry

The speech perception test battery that was used comprised seven different tests that quantify the increasing complexity of speech perception; basal speech perception tests (i.e. tests on speech discrimination and supra-segmental speech identification tests), word identification tests (Dutch version of the closed-set Early Speech Perception tests) and open-set speech recognition tests (an open-set word recognition test using monosyllables).[17] Scores on this test battery were reduced to one single measure, called the 'equivalent hearing loss' (EHL).[17,18] In order to obtain the 'equivalent hearing loss' reference data, the speech perception tests were administered to a group of 46 severely and profoundly hearing impaired children. Their PTA (average hearing loss at 0.5, 1 and 2 kHz) ranged from 50 to 130 dB HL. They were all using binaural powerful conventional hearing aids and they had been participating in aural-oral training programmes for at least 3 years. A principal component analysis of the subtests showed that there was one main factor that was significant for all subtests and which explained 73% of the variance. This suggested that the subtest scores could be clustered, which enables a better overview. The best-fit curves for the individual speech perception scores as a function of hearing loss were used in reverse to relate the scores of an experimental case (e.g. a child with a CI) to

those of the reference group. This results in 'equivalent hearing loss' scores, however, it can only be applied on % correct scores between 10% and 90%. Thus bottom and ceiling scores that occur inevitably when using a broad battery of speech perception tests for the follow-up of children at different ages and developmental stages[19], were excluded in the calculations. The EHL values can vary between 50 and 130 dB HL.

In the present study, a within-subject repeated-measure design was used to compare the children's preoperative performance with conventional hearing aids (t = 0) to their postoperative performance with the Nucleus cochlear implant after 3, 6, 12, 18, 24, 36 and 48 months of use. Missing values occurred in some children, because of unavoidable circumstances, such as intercurrent illness. Comparison of the results from the various subgroups was made using the Mann-Whitney-Wilcoxon two sample test, as the distributions were not normal. A probability value of $P < 0.05$ was considered to be significant. Multiple regression analysis was performed on the 3-year data (n = 67) to examine the influence of the different variables on the EHL: age at onset of deafness, duration of deafness and communication mode

Results

Postimplant development of speech perception

All the children showed improvement in speech perception over time, but at various rates and to various extents. Preoperative and postoperative performance expressed in EHL scores at each evaluation point are summarised in Figure 1. In Figure 2 the mean scores of the subgroups are shown. The mean preoperative EHL of the postlingually deaf children was 124 dB HL. This improved to 70 dB HL 3 years after implantation for the children who had been deaf for less than 3 year and to 77 dB HL in the children who had been deaf for a longer period (a non-significant difference: $P = 0.2$). Most of the improvement took place during the first year, in which the children with a short duration of deafness showed the fastest progress.

The prelingually acquired deaf children developed at a slower rate than the postlingually deaf children. However, 3 years after implantation, the results of the prelingually deaf children who had been deaf for less than 3 years were no different from those of the postlingually deaf children ($P = 0.11$) The results of the prelingually deaf children with a longer duration of deafness varied fairly widely (Figure 1) and their performance seemed to be poorer than that of the children with a short duration of deafness. This difference in outcome was not significant ($P = 0.28$).

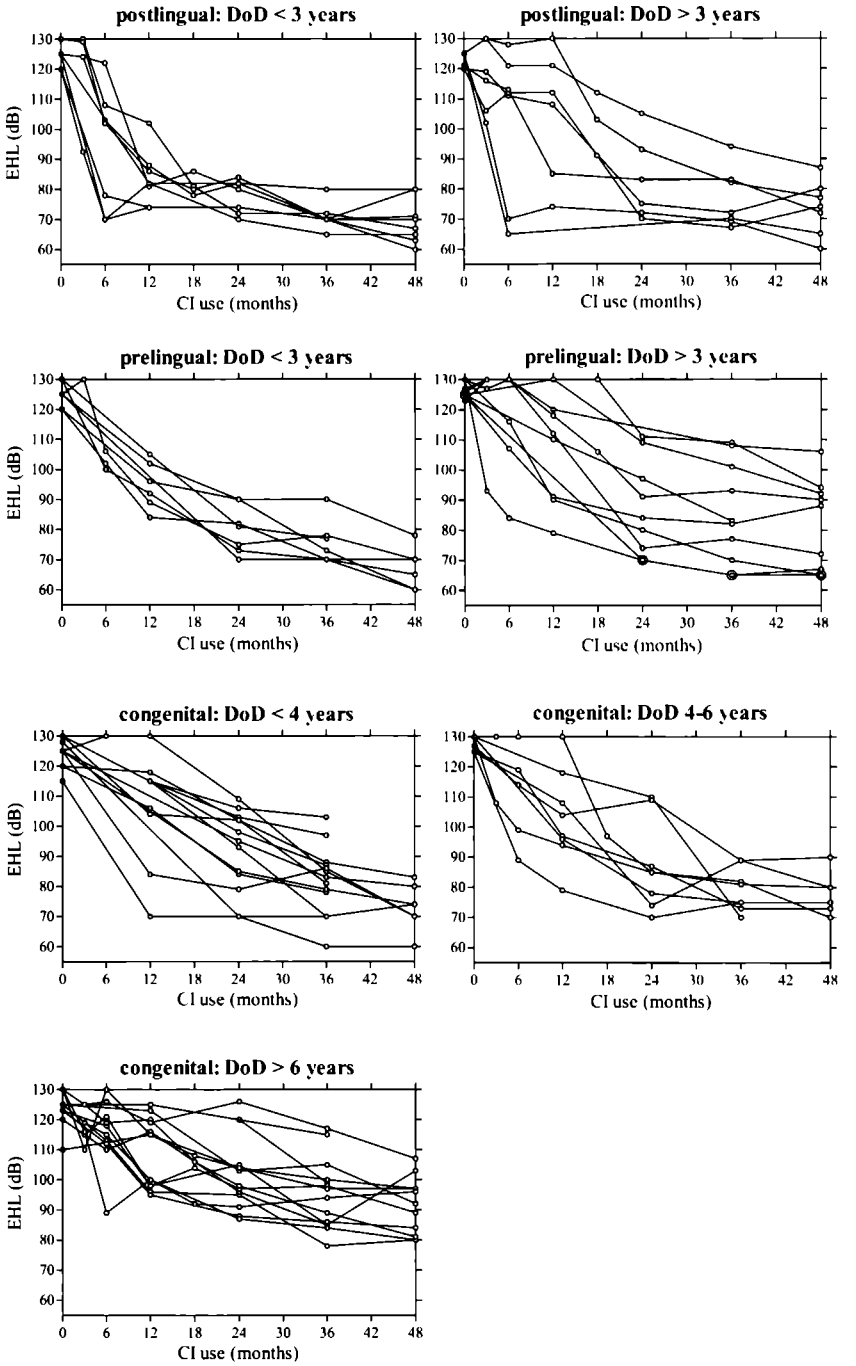


Figure 1 Individual longitudinal data of EHL in all subgroups DoD = duration of deafness

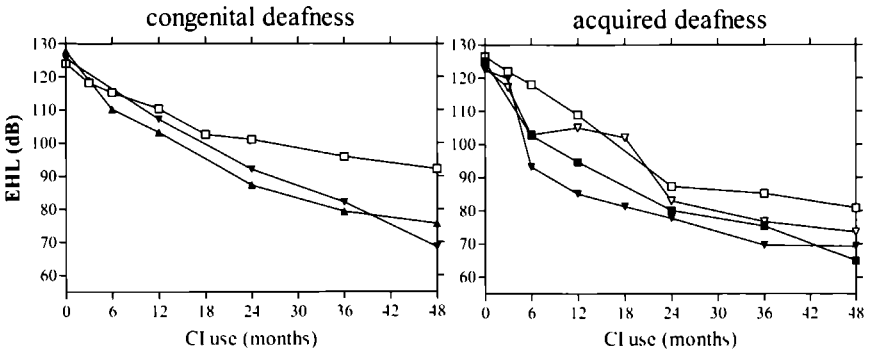


Figure 2. Longitudinal analysis of the mean EHL in prelingually and postlingually deaf children (right) and congenitally deaf children (left)

The symbols in the group with *congenital* deafness refer to. □= duration of deafness longer than 6 years, ▲=duration of deafness 4 to 6 years, ▼= duration of deafness less than 4 years. In the group with *acquired* deafness, the symbols refer to· □= prelingually deaf, duration of deafness longer than 3 years; ■= prelingually deaf, duration of deafness less than 3 years, ▽= postlingually deaf, duration of deafness longer than 3 years; ▼= postlingually deaf, duration of deafness less than 3 years.

Figure 2 shows that the congenitally deaf children also improved over time, but after the first year of CI use, the rate of progress was slower in the children with a longer duration of deafness. At 3 years follow-up, the children of older than 6 years at implantation had significantly poorer scores than the children implanted at a younger age (P = 0.001). They seemed to reach a plateau at a poorer EHL level.

Correlation between variables

Figure 3 shows the median EHL and range of the 7 subgroups at the specific evaluation points 3 and 4 years postimplantation in the form of boxplots. Mean scores were comparable between the postlingually deaf children, the prelingually deaf children implanted relatively early and the congenitally deaf children implanted before the age of 6 years (Mann-Whitney Test, $p < 0.05$). After 4 years of follow-up, most children in these 5 subgroups had reached an EHL of 75 dB HL or less (Figure 3b). Thus their speech perception abilities at that time were comparable with the reference group of hearing impaired children using well-fitted conventional hearing aids whose hearing loss was 75 dB HL. After 3 years of implant use (Figure 3a), the performance of the congenitally deaf children implanted before the age of 6 years was equal to that of the prelingually deaf children, but significantly poorer than that of the postlingually deaf children (P=0.009). Especially the congenitally deaf children implanted before the age of 4 years and the

prelingually deaf children with a short duration of deafness were still showing noticeable signs of progress during the fourth year of follow-up.

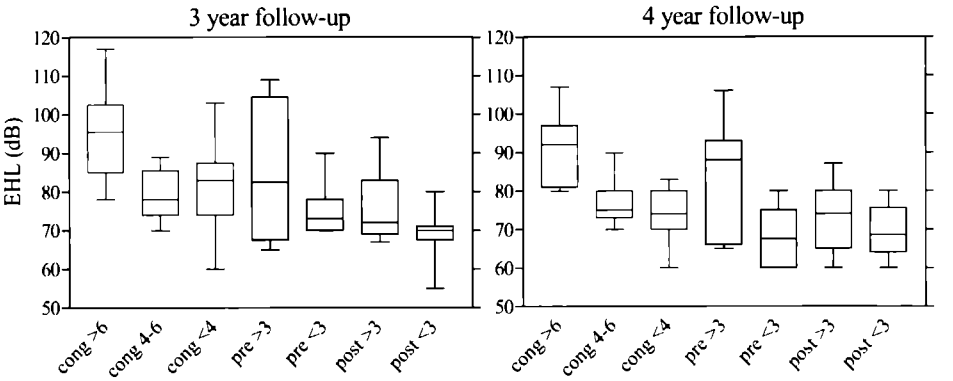


Figure 3a

Figure 3b

Figure 3. EHL obtained at 3 and 4 years postimplantation as a function of subgroup
The boxes extend from the 25th percentile to the 75th percentile, with a line at the median (the 50th percentile). The whiskers extend above and below the boxes to show the highest and lowest values.
Abbreviations:
Cong > 6 = congenitally deaf children, older than 6 years at implantation;
Cong 4 - 6 = congenitally deaf children, 4 to 6 years old at implantation;
Cong < 4 = congenitally deaf children, younger than 4 years at implantation;
Pre > 3 = prelingually deaf children, duration of deafness longer than 3 years;
Pre < 3 = prelingually deaf children, duration of deafness less than 3 years;
Post > 3 = postlingually deaf children, duration of deafness longer than 3 years,
Post < 3 = postlingually deaf children, duration of deafness less than 3 years

Table 3. Correlations between EHL at 3 years follow-up (EHL3), at 4 years follow-up (EHL4) and different variables

	Duration of deafness	Age at onset of deafness	Communication mode (1 = oral, 0 = signs only)
EHL3 of congenitally deaf children (n=35)	0.59 (S)	-	-0.15 (NS)
EHL4 of congenitally deaf children (n=25)	0.74 (S)	-	-0.27 (NS)
EHL3 of children with acquired deafness (n=32)	0.13 (NS)	-0.35 (S)	-0.83 (S)
EHL4 of children with acquired deafness (n=30)	0.28 (NS)	-0.28 (NS)	-0.75 (S)

S = significant correlation ($P < 0.05$), NS = non-significant correlation ($P > 0.05$)

Table 3 shows the correlations between the EHL at 3-year follow-up (EHL3) and at 4-year follow-up (EHL4) and the variables age at onset, duration of deafness and communication mode for the whole group of children. Multiple regression analysis was conducted with

EHL3 and EHL4 as dependent variable and the variables mentioned above as independent variables. Separate analyses were performed on the data sets from the children with congenital deafness and those with acquired deafness.

In the *congenital* cases, a statistically significant association was observed between EHL3 and duration of deafness ($P < 0.001$). Communication mode was not found to be associated with EHL3 ($P > 0.2$). Duration of deafness accounted for 36% of the variance in EHL3 and even 55% of the variance in EHL4. To illustrate this, in Figure 4, the EHL values of all the congenitally deaf children after 3 years of CI use are plotted against their age at implantation (i.e. duration of deafness). This relation between EHL and duration of deafness in congenitally deaf children persists after a longer follow-up period of 4 years. At 4 years follow-up less data were available but at 3 years follow-up there had occurred less change in educational setting so that most children used a form of communication comparable to that of the preoperative situation.

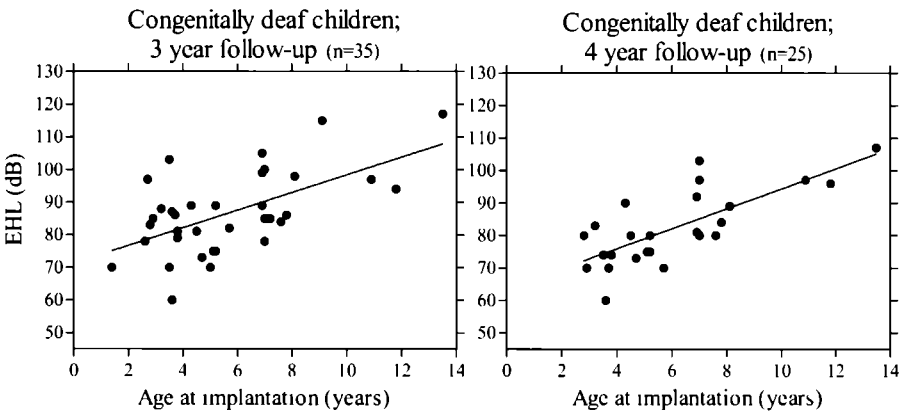


Figure 4. Equivalent Hearing Loss obtained at 3 and 4 years of follow-up as a function of the age at implantation for the group of congenitally deaf children

In the children with *acquired* deafness, a significant association was found between EHL3 and age at onset ($P < 0.05$) and communication mode ($P < 0.001$). At 4-year follow-up, there was no longer a significant correlation between age at onset of deafness and EHL4. Duration of deafness was neither associated with EHL3 ($P = 0.2$) nor with EHL4 ($P > 0.05$). The two significant variables age at onset of deafness and communication mode accounted for 71% of the variance in EHL3; communication mode alone accounted for 69% of the variance in EHL3. The only significant variable at 4-year follow-up, communication mode, accounted for 56% of the variance in EHL4.

Discussion

In children with a CI, the acquisition of auditory and spoken language skills develops steadily over time.[20,21] Speech perception is a basic outcome measure of the resolution provided by the implant. It does not fully reflect the attainment in language nor the ability to communicate. It is known that several child related factors, environmental factors and device-related factors influence the outcomes with a CI. The present data are unique because they concern a well-defined group owing to the conservative inclusion and exclusion criteria as generally used in the eighties and nineties. Several children were included that nowadays are no longer considered good candidates, for example the congenitally deaf children with long duration of deafness.

Age at onset of deafness

Early clinical trials have reported that prelingually deaf children make slower progress in the development of speech perception skills than postlingually deaf children, which is in accordance to our findings.[22,23] However, 4 years after implantation, the correlation between age at onset of deafness and the EHL of the children with acquired deafness was non-significant in the present study. This suggests that on the long run, the differences between postlingually and prelingually deaf children might disappear.

It has been reported that congenitally deaf children made slower progress than prelingually deaf children.[24,25] These differences might be due to the effects of prior auditory input, which is absent in congenitally deaf children and present, although limited in prelingually deaf children. The present study showed that compared to the early implanted children with acquired deafness, the congenitally deaf children implanted before the age of 6 years developed at a slower rate during the first 2 years. However, their progress remained steady over time, which resulted in comparable outcomes between postlingually, prelingually and early implanted congenitally deaf children after 4 years of CI use (Figure 3b). Others reported similar findings.[26,27] This suggests that the present congenitally deaf children implanted before the age of 6 years are relatively 'slow starters'.

Outcome comparison with conventional hearing aid users

Some researchers compared the performance of children using a CI to hearing impaired children using conventional hearing aids. This is also the central theme of the present study, incorporated into the EHL procedure. Somers et al.[28] showed that speech perception scores in the control group of profoundly hearing impaired children with unaided thresholds of between 100 and 110 dB HL were equal to those in the CI group after 1 year of follow-up. Svirsky and Meyer[29] reported that 12 to 18 months after implantation, the speech perception scores of prelingually deaf children with a CI were similar to those of

prelingually and congenitally deaf hearing aid users with residual hearing in the 90 to 100 dB HL range. Miyamoto et al.[30] found that 2.5 years after implantation, the mean speech perception scores of prelingually deaf children exceeded the average score of children with conventional hearing aids with a PTA of between 90 and 100 dB HL. In line with the results of these studies, the mean EHL values after 12 and 24 months of CI use in all the prelingually deaf children in the present study were 100 dB HL (n = 14) and 84 dB HL (n = 16), respectively. The longer follow-up period in the present study revealed further important improvement over time. Remarkably, after 3 years, the majority of the children were performing equally as well as hearing aid users with a PTA of 70-80 dB HL and in some individuals even 60-70 dB HL (Figure 1).

Duration of deafness in acquired deafness

Children with acquired deafness of long duration (i.e. a long period between age at onset of deafness and age at implantation) showed slower progress and more variability in scores than those with a short duration of deafness. There was a tendency towards better performance in children who had been deaf for less than 3 years (Figure 2), which underlines the negative effect of several years of auditory deprivation prior to implantation. However, duration of deafness and EHL3-4 were not correlated in the children with acquired deafness. This absence of a significant correlation might be explained by the fact that although the duration of deafness in the children with acquired deafness ranged from 0.7 to 9.8 years, the mean duration was only 3.6 years. Some studies in postlingually deaf adults also failed to find a strong negative effect of duration of deafness on postoperative performance.[4] Osberger et al.[1] showed that after 18 months of CI use, prelingually deaf children with a 3 year duration of deafness performed as well as those with a 7.5 year duration of deafness, although the former children seemed to improve more rapidly. It can be argued that to fully examine the effect of duration of deafness, a long follow-up is needed to ensure that all the different subgroups of implanted children reach a plateau in speech perception scores. This recommendation is based on the vast improvement made by the children with a short duration of deafness even in their fourth year (Figure 3b). Despite the similar scores in children with long and short durations of deafness after 3 to 4 years of implant use, the fact that implantation after a longer period of auditory deprivation causes delay in the development of speech perception may have a negative effect on spoken language development and the child's performance at school.

Duration of deafness in congenital deafness

In the congenitally deaf children, there was a relatively high correlation (0.59; n = 35) between duration of deafness (i.e. age at implantation) and the EHL (Table 3 and Figure 4). The earlier a congenitally deaf child is implanted, the better his or her speech perception

performance after 3 years of CI use No significant differences in speech perception were found between the congenitally deaf children implanted under 4 years of age and those implanted between 4 to 6 years Similarly, in a study by Papsin et al [31], congenitally deaf children implanted before 6 years of age made significantly better progress in open-set speech perception than children implanted after 6 years of age The influence of duration of deafness is generally accepted to be the result of the age-dependent plasticity of neurosensory development When Manrique et al [32] found poor results in prelingually deaf children implanted after 6 years of age, they argued that the period of auditory plasticity may span the first 6 years of life and auditory stimulation with a CI after this period might not be able to fully restore the loss of auditory plasticity

Communication mode

Evaluating the child's communication mode, and especially changes from signs-only to oral communication, can contribute to a more extensive view on the benefit a child can derive of it's CI In the children with acquired deafness, a signs-only communication mode was significantly correlated (-0 83, n = 35) with a poor EHL Multiple regression analysis showed that after three years of CI use, the group receiving non-oral education had an overall EHL that was 23 dB poorer than those using oral communication It is possible that the choice of education is influenced by the level of speech perception achieved by the child children with limited auditory capacities are more likely to be placed at a school that uses sign language However, in the nineties, there was little choice as 4 out of the 5 schools for the deaf in the Netherlands used only sign language for teaching their pupils In the congenitally deaf children, communication mode was not correlated with the EHL In this group, 15 children used mainly oral communication, while 20 used only sign language The 'poor performers' in this group were equally distributed over the two communication modes It should be noted however, that the number of patients is limited

The literature has also shown that children who followed oral communication programmes performed better and acquired auditory perception skills at a faster rate than children in total communication school settings [33] When children who previously communicated through sign language develop auditory perception and speech production skills with their CI, they are likely to become more auditory-oral communicators In the present study, the postlingually deaf children reached an EHL level of 71 dB HL after 4 years, while the prelingually and early implanted congenitally deaf children reached an EHL of 72 dB HL As can be expected with these outcomes, several children changed their communication mode during follow-up, this occurred primarily after more than 3 years of CI use

Conclusions

Most children derived substantial benefit from their cochlear implant in terms of speech perception, but performance varied greatly. The best performers were the postlingually deaf, the prelingually deaf implanted at a relatively early age and the congenitally deaf implanted at an age of younger than 6 years; postlingually deaf children showed the fastest rate of improvement and the best long-term scores. Children with a longer duration of deafness needed more time to catch up with the other groups.

These results once again emphasise the advantage of implanting congenitally deaf children at a young age. Age at onset of deafness, prelingual or postlingual, had little influence on speech perception scores after 3 years of CI use. Furthermore, in the children with acquired deafness, better performance was highly associated with an oral communication mode.

References

1. Osberger MJ, Kalberer A, Zimmerman-Phillips S, Barker MJ, Geier L Speech perception results in children using the Clarion Multi-Strategy Cochlear Implant. *Ann Otol Rhinol Laryngol Suppl* 2000;185:75-7.
2. Miyamoto RT, Osberger MJ, Robbins AM, Myres WA, Kessler K Prelingually deafened children's performance with the nucleus multichannel cochlear implant. *Am J Otol* 1993;14:437-45.
3. Gantz BJ, Tyler RS, Woodworth GG, Tye-Murray N, Fryauf-Bertschy H. Results of multichannel cochlear implants in congenital and acquired prelingual deafness in children: five-year follow-up. *Am J Otol* 1994;15 Suppl 2:1-7.
4. Hiraumi H, Tsuji J, Kanemaru S, Fujino K, Ito J. Cochlear implants in post-lingually deafened patients *Acta Otolaryngol Suppl* 2007;17-21.
5. O'Neill C, O'Donoghue GM, Archbold SM, Nikolopoulos TP, Sach T Variations in gains in auditory performance from pediatric cochlear implantation. *Otol Neurotol* 2002,23:44-8
6. Snik AF, Makhdoum MJ, Vermeulen AM, Brokx JP, van den Broek P. The relation between age at the time of cochlear implantation and long- term speech perception abilities in congenitally deaf subjects. *Int J Pediatr Otorhinolaryngol* 1997,41.121-31.
7. Fryauf-Bertschy H, Tyler RS, Kelsay DM, Gantz BJ, Woodworth GG. Cochlear implant use by prelingually deafened children: the influences of age at implant and length of device use. *J Speech Lang Hear Res* 1997;40:183-99
8. Pyman B, Blamey P, Lacy P, Clark G, Dowell R. The development of speech perception in children using cochlear implants. effects of etiologic factors and delayed milestones. *Am J Otol* 2000;21.57-61
9. Osberger MJ, Fisher L. Preoperative predictors of postoperative implant performance in children. *Ann Otol Rhinol Laryngol Suppl* 2000,185:44-6.
10. David EE, Ostroff JM, Shipp D, Nedzelski JM, Chen JM, Parnes LS, Zimmerman K, Schramm D, Seguin C. Speech coding strategies and revised cochlear implant candidacy. an analysis of post-implant performance. *Otol Neurotol* 2003;24:228-33.
11. Dowell RC, Blamey PJ, Clark GM. Potential and limitations of cochlear implants in children. *Ann Otol Rhinol Laryngol Suppl* 1995;166:324-7.
12. Hodges AV, Dolan AM, Balkany TJ, Schloffman JJ, Butts SL Speech perception results in children with cochlear implants contributing factors. *Otolaryngol Head Neck Surg* 1999,121:31-4.

13. Young NM, Grohne KM, Carrasco VN, Brown CJ. Speech perception in young children using nucleus or Clarion Cochlear Implants. effect of communication mode. *Ann Otol Rhinol Laryngol Suppl* 2000;185:77-9.
14. Knutson JF, Ehlers SL, Wald RL, Tyler RS Psychological predictors of pediatric cochlear implant use and benefit. *Ann Otol Rhinol Laryngol Suppl* 2000;185:100-3.
15. Psarros CE, Plant KL, Lee K, Decker JA, Whitford LA, Cowan RS. Conversion from the SPEAK to the ACE strategy in children using the nucleus 24 cochlear implant system: speech perception and speech production outcomes. *Ear Hear* 2002;23:18S-27S.
16. Rotteveel LJ, Snik AF, Vermeulen AM, Mylanus EA. Three-year follow-up of children with postmeningitic deafness and partial cochlear implant insertion. *Clin Otolaryngol* 2005,30:242-8
17. Snik AF, Vermeulen AM, Brokx JP, Beijk C, van den Broek P Speech perception performance of children with a cochlear implant compared to that of children with conventional hearing aids. I. The "equivalent hearing loss" concept. *Acta Otolaryngol* 1997,117.750-4.
18 Boothroyd A, Eran O. Auditory speech perception capacity of child implant users expressed as equivalent hearing loss. *The Volta Review* 1994,96.151-68
19 Snik AF, Vermeulen AM, Geelen CP, Brokx JP, van den Broek P. Speech perception performance of children with a cochlear implant compared to that of children with conventional hearing aids II. Results of prelingually deaf children. *Acta Otolaryngol* 1997,117:755-9.
20. Beadle EA, McKinley DJ, Nikolopoulos TP, Brough J, O'Donoghue GM, Archbold SM Long-Term Functional Outcomes and Academic-Occupational Status in Implanted Children After 10 to 14 Years of Cochlear Implant Use. *Otol Neurotol* 2005;26:1152-60.
21. Nikolopoulos TP, Archbold SM, O'Donoghue GM. The development of auditory perception in children following cochlear implantation *Int J Pediatr Otorhinolaryngol* 1999;49 Suppl 1:S189-91.
22. Waltzman SB, Cohen NL, Gomolin RH, Shapiro WH, Ozdamar SR, Hoffman RA Long-term results of early cochlear implantation in congenitally and prelingually deafened children *Am J Otol* 1994;15 Suppl 2:9-13.
23. Fryauf-Bertschy H, Tyler RS, Kelsay DM, Gantz BJ. Performance over time of congenitally deaf and postlingually deafened children using a multichannel cochlear implant. *J Speech Hear Res* 1992;35:913-20.
24 Fukushima K, Sugata K, Kasai N, Fukuda S, Nagayasu R, Toida N, Kimura N, Takishita T, Gunduz M, Nishizaki K. Better speech performance in cochlear implant patients with GJB2-related deafness. *Int J Pediatr Otorhinolaryngol* 2002;62:151-7.
25 Staller SJ, Beiter AL, Brimacombe JA, Mecklenburg DJ, Arndt P. Pediatric performance with the Nucleus 22-channel cochlear implant system. *Am J Otol* 1991;12 Suppl:126-36.
26 O'Donoghue GM, Nikolopoulos TP, Archbold SM, Tait M Speech perception in children after cochlear implantation. *Am J Otol* 1998;19:762-7.
27. Mitchell TE, Psarros C, Pegg P, Rennie M, Gibson WP. Performance after cochlear implantation: a comparison of children deafened by meningitis and congenitally deaf children *J Laryngol Otol* 2000;114:33-7.
28. Somers MN Speech perception abilities in children with cochlear implants or hearing aids. *Am J Otol* 1991,12 Suppl:174-8.
29. Svirsky MA, Meyer TA. Comparison of speech perception in pediatric CLARION cochlear implant and hearing aid users *Ann Otol Rhinol Laryngol Suppl* 1999;177:104-9.
30. Miyamoto RT, Kirk KI, Todd SL, Robbins AM, Osberger MJ Speech perception skills of children with multichannel cochlear implants or hearing aids. *Ann Otol Rhinol Laryngol Suppl* 1995,166:334-7.
31. Papsin BC, Gysin C, Picton N, Nedzelski J, Harrison RV. Speech perception outcome measures in prelingually deaf children up to four years after cochlear implantation. *Ann Otol Rhinol Laryngol Suppl* 2000;185:38-42.

32. Manrique M, Cervera-Paz FJ, Huarte A, Perez N, Molina M, Garcia-Tapia R Cerebral auditory plasticity and cochlear implants. *Int J Pediatr Otorhinolaryngol* 1999;49 Suppl 1:S193-7.
33. Geers A, Brenner C, Davidson L. Factors associated with development of speech perception skills in children implanted by age five. *Ear Hear* 2003;24:24S-35S.

Chapter 3

Cochlear implantation in the postmeningitic ossified cochlea

Three-year follow-up of children with postmeningitic deafness and partial cochlear implant insertion

L.J.C. Rotteveel
A.F.M. Snik
A.M. Vermeulen
E.A.M. Mylanus

Clin Otorhinolaryngol 2005, 30(3) 242-248

Abstract

Objectives To evaluate the long-term outcome of children with postmeningitic deafness and partial insertion of the Nucleus electrode array due to ossification of the cochlea, and to compare their speech perception performance to that of children with postmeningitic deafness and full insertion of the electrode array

Methods A battery of seven speech perception tests was administered to 25 children with a cochlear implant Results were reduced into one measure equivalent hearing loss (EHL) The partial insertion group comprised 7 children with postmeningitic deafness, mean age at implantation 5 5 years, mean duration of deafness 3 6 years The full-insertion control group comprised 18 children with postmeningitic deafness, mean age at implantation 4 4 years, mean duration of deafness 2 9 years All the children became deaf between 0 and 3 years of age

Results Over a 3-year follow-up period, the children with partial insertion showed continuing progress, although there was wide variation in performance and the rate of progression Some open-set comprehension could even be achieved with the insertion of only 8 electrodes of a nucleus device

Three years after implantation, speech perception in the partial insertion children was poorer than that in the control groups with long ($P < 0.01$, 95% confidence interval 7-43 dB EHL) and short duration of deafness ($P < 0.0001$, 95% confidence interval 28-53 dB EHL) They showed slower progress and reached a poorer EHL plateau Four of the seven children acquired open-set word recognition

Conclusions Patients with partial insertion of the electrode array benefit from a cochlear implant, although less than patients with complete insertion

Introduction

Hearing loss is a frequent complication of meningitis The incidence of hearing impairment following meningitis is reported to be 10 5% In developed countries, approximately 5% of survivors are left with permanent sensorineural hearing loss, depending on the causative organism Otherwise, meningitis is one of the most common aetiologies of acquired hearing loss in childhood [1] Hearing loss occurs when the infection spreads from the meninges to the inner ear and bacteria invade the cochlea This causes an acute inflammatory response the initial acute stage of suppurative labyrinthitis During labyrinthitis, the organ of Corti and hair cells may be damaged by the inflammation and subsequent fibrosis (fibrotic stage) and potential ossification (ossification stage) of the cochlea Some degree of cochlear ossification, i e the end point of severe inflammatory disease, is found in approximately 70% of cases with bacterial meningitis and profound hearing loss [2] The frequency and severity of ossification varies according to the causative organism [2] Neo-ossification is most marked in the scala tympani of the basal turn of the cochlea, whereas the more apical turns are less affected In regions of ossification, there is severe damage to the organ of Corti [3] Evidence also exists that lesions of the acoustic nerve, brainstem or higher auditory pathways may be responsible for postmeningitic hearing loss [4]

Although cochlear ossification used to be considered a contraindication for cochlear implantation, nowadays many centres implant these patients routinely

Meningogenic labyrinthitis ossificans is a concern in cochlear implantation because of loss of stimulable spiral ganglion cells and the technical difficulties of dealing with surgery of an obliterated cochlea Histopathological temporal bone studies are inconclusive on the relation between the severity of ossification and ganglion cell survival Hinojosa et al [3] observed variability in survival of ganglion cells in temporal bones of deaf subjects with and without ossification while others reported substantial spiral cell loss in middle and apical turns affected by ossification [5] Surgical techniques to overcome this cochlear obstruction require various degrees of drilling[6,7], alternative placement of the electrode array in the scala vestibuli or extracochlearly[8,9] and the use of double array implants [10]

Speech perception and production have been studied extensively in postmeningitic deaf children who received a cochlear implant (CI) [11,12] Significant improvement after cochlear implantation has been reported both in speech perception and production compared to their pre-operative performance with conventional hearing aids Electrical stimulation of the auditory nerve appeared effective despite the presence of new bone formation Comparable results were achieved in implanted patients with ossified cochleae and patent cochleae [2,7,13] The degree of ossification did not appear to affect speech perception performance [13] However, if the electrode array can only partially be inserted because of extensive cochlear

ossification, the results can be less favourable [7] Surgical procedure, electrode array placement and the number of electrodes in use affect the patient's performance [6 7] Although many patients with postmeningitic deafness have some degree of ossification, this can usually be overcome peroperatively Ossification inhibited the complete insertion of a multichannel electrode array in only a small number of these patients [6] Other factors that influence a child's performance with a CI are the duration of deafness, duration of CI experience, age at onset of deafness, age at implantation, level of residual hearing preoperatively, intelligence, motivation, psychological support at home, communication mode and educational environment

The purpose of this study on children with postmeningitic deafness was to compare the speech perception performance of those with partial insertion of the electrode array to that of a control group with full insertion

Methods

Subjects

Deafness was caused by meningitis in 52% of the children implanted between 1990 and 1998 in the CI programme in our centre Twenty-five consecutive children were selected to take part in this study according to the inclusion criteria listed in Table 1

The causative infections were *Streptococcus pneumoniae* in 17 children, *Neisseria meningitidis* in one child, *Haemophilus influenzae* in four children and unknown in two children (Table 2)

Seven children had partial insertion of the Nucleus multichannel CI electrode array Partial insertion was defined as the condition where electrodes remained visible outside the cochlea after inserting the array through the cochleostomy Mean age at onset of deafness in this partial-insertion group was 1 8 years, mean age at implantation was 5 5 years and mean duration of deafness was 3 7 years Duration of deafness was defined as the time between onset of deafness and implantation

Table 1 Inclusion criteria

Residual hearing	Hearing thresholds at 1, 2 and 4 kHz exceeding 95 dB HL, no open-set speech perception
Age at onset of deafness	0 to 3 years of age
Aetiology	Meningitis of any kind
Medical condition	No/minor additional disabilities
Cognition	Normal non-verbal intelligence
Motivation	Good motivation and support at home

The control group comprised 18 children with postmeningetic deafness and full insertion of the electrode array. Mean age at onset of deafness was 1.6 years, mean age at implantation was 4.5 years and mean duration of deafness was 2.9 years. As the duration of deafness influences speech perception,[14] these children were divided into a group with a duration of deafness longer than 3 years, and a group shorter than 3 years. Duration of deafness was longer than 3 years in eight of the full-insertion and in four of the partial-insertion children.

Surgical technique and device characteristics

In the selection period, cochlear imaging was performed using computed tomography (CT) scanning. These scans were reviewed retrospectively and compared to the degree of cochlear ossification observed at surgery.

If during surgery no lumen in the scala tympani or the scala vestibuli could be localized for insertion of all electrodes, limited drilling of 6 to 8 mm was performed in an anteromedial direction along the scala tympani in the obliterated basal turn, until a lumen became visible, or until a new channel had been created.

All the children received the Nucleus 22 or 24 CI. The speech processors were programmed 4 to 6 weeks after surgery. During rehabilitation, testing took place and readjustments were made to the programming parameters to improve each child's performance. Almost all the children were using the multiple peak (MPEAK) or spectral peak (SPEAK) coding strategy. Four children were using the advanced combination encoder (ACE) strategy (C16, C17, C18 and S1).

Performance

Aided sound-field thresholds were measured using narrow band noise with central frequencies from 0.25 to 4 kHz. Speech perception data were analysed and computed into one single measure, called the 'equivalent hearing loss' (EHL).[15] Speech perception test results obtained from a group of severely and profoundly hearing impaired children with well-fitted conventional hearing aids were used as a reference. Relations between the test scores and the degree of hearing loss were established. These relations were used to transform the scores of subjects with a CI into an EHL value. EHL values vary between 50 and 130 dB hearing level (HL). This measure can be used to summarize progress monitored with a battery of different speech perception tests and effectively handle bottom and ceiling test scores that occur when a broad battery of speech perception tests of varying difficulty is used for the follow-up of children at different ages and developmental stages.[16] The EHL value is based on three scores that quantify the increasing complexity of speech perception: basal speech perception score (i.e. the scores on the speech discrimination and suprasegmental speech identification tests), the word identification

score (the Dutch version of the closed-set Early Speech Perception tests) and the open-set speech recognition score (a phoneme and word recognition test using monosyllables).[15]

Statistics

A within-subject repeated-measure design was used to compare the children's preoperative performance with conventional hearing aids (t=0) to their postoperative performance with the CI after 3, 6, 12, 18, 24, 36 and 48 months of use. Most children were tested at each evaluation point, although missing data in some children occurred because of missed appointments or other unavoidable circumstances. Comparison of the results of the three groups was made using unpaired *t*-tests, or Mann-Whitney U tests when distributions were non-normal. A probability value of $P < 0.05$ was considered to be significant.

Results

Subjects

Patient characteristics are shown in Table 2. There were no significant differences between the partial-insertion and full-insertion groups concerning age at onset of deafness, age at implantation and duration of deafness (Table 3).

CT scanning

Ossification preoperatively identified by CT scanning, was confirmed during surgery in 10 children (sensitivity 53%). In 9 children, no ossification was visible on the CT scan, but was indeed encountered during surgery (false negative rate 47%; negative predictive value 40%). There were no false positive CT scans (specificity 100%) (Table 4). The presence and location of ossification (basal and/or apical) was diagnosed correctly in six of seven children with partial insertion. In the remaining child, ossification seemed to be absent in the one ear selected for implantation, but severe in the other ear.

Table 2 Group characteristics

ubject	Onset of deafness (years)	Causative organism	Duration of deafness (years)	Age at implantation (years)	Drill-out procedure	No. of inserted electrodes*	No. of active electrodes at 3-yr CI
1	2.7	S. pneumoniae	3.5	6.2	Total	8	8
2	2.6	Unknown	3.8	6.4	Total	10	Non user
3	0.6	S. pneumoniae	8.6	9.3	Total	10	13
4	0.4	H. influenzae	3.3	3.7	Total	13	11
5	2.6	S. pneumoniae	3.0	5.6	None	13	13
6	3.3	H. influenzae	1.8	5.2	Total	12	13
7	0.4	S. pneumoniae	1.8	2.3	Partial	18	17
:1	2.7	S. pneumoniae	2.4	5.1	None	27	22
:2	2.7	S. pneumoniae	2.5	5.2	Partial	32	18
:3	2.5	S. pneumoniae	3.5	6.0	Partial	32	20
:4	2.6	S. pneumoniae	3.8	6.4	Partial	27	19
:5	2.9	H. influenzae	2.3	5.2	Partial	27	22
:6	2.3	H. influenzae	3.2	5.6	Partial	32	21
:7	3.0	H. influenzae	1.3	4.3	Partial	32	22
:8	0.2	S. pneumoniae	5.9	6.2	Partial	32	22
:9	1.8	S. pneumoniae	4.0	5.7	None	32	21
:10	1.4	S. pneumoniae	1.9	3.3	Partial	26	20
:11	1.7	S. pneumoniae	4.1	5.8	None	32	20
:12	0.9	N.meningitidis	2.0	2.9	None	27	20
:13	1.4	S. pneumoniae	2.2	3.6	None	29	20
:14	0.8	Unknown	3.1	3.9	None	22	20
:15	0.0	S. pneumoniae	3.2	3.3	None	26	20
:16	0.3	S. pneumoniae	1.9	2.2	Partial	30	20
:17	0.5	S. pneumoniae	2.4	2.9	None	32	20
:18	0.2	S. pneumoniae	2.4	2.5	Partial	32	20

S1-7 = subjects 1 to 7 with partial insertion of electrode array, C1-18 = control group subjects 1 to 18 with full insertion of electrode array; No. = number, *the number of electrodes inserted in the control group, including the number of retaining rings; 3-yr CI = 3 years of CI use.

Table 3. Comparison of group characteristics and outcomes

	Partial insertion	Full insertion	Difference
Number of subjects	7	18	-
Age at onset of deafness (years)	1.8	1.6	NS
Age at implantation (years)	5.5	4.5	NS
Duration of deafness (years)	3.6	2.9	NS
EHL at 3-year follow-up (dB)	112 (range 82-130)	DoD > 3 subgroup: 87 (range 70-109)	S
		DoD < 3 subgroup: 72 (range 70-78)	S

EHL = equivalent hearing loss, DoD > 3 subgroup = full-insertion subgroup with duration of deafness of longer than 3 years; DoD < 3 = full-insertion subgroup with duration of deafness of shorter than 3 years; S = significant, NS = non-significant.

Table 4. Ossification encountered at surgery and diagnosed on CT scan in 25 children with postmeningitic deafness

Ossification at surgery	Ossification on CT scan		
	present	absent	total
present	10	9	19
absent	0	6	6
total	10	15	25

Sensitivity = 53%; specificity = 100%; negative predictive value = 40%; positive predictive value = 100%.

Surgery

During surgery, ossification was present in 19 of the 25 children (Table 4).

In the partial-insertion group, the basal turn was totally obliterated in five of seven children. After drilling for up to 8 mm still no lumen was found. Further drilling was limited by the coiling of the cochlea and the proximity to the carotid canal. Therefore, the electrode arrays were partially placed in the drilled tunnel. In two children, partial insertion was caused by ossification of the apical turns. In one of these cases, some basal ossification was also present, required limited drilling.

In the control group, cochleostomy revealed a fully patent basal turn in six children. In two children, some bony ridges were seen, but they were not causing any obliteration of the basal turn. In 10 children, it was necessary to drill 1 to 6 mm before the natural lumen in the scala tympani was reached (Table 5). There were no cases of scala vestibuli or total drill out insertions.

In 18 children with full insertion, the number of retaining rings that could also be inserted has been added to the number of electrodes. In nine children, all 10 retaining rings could be inserted (Table 2).

Table 5. Degree of ossification and causative organism in the partial-insertion and full-insertion control group

Causative organism	Partial insertion (n=7)			Full insertion (n=18)			
	n	Ossification (7/7)		n	Ossification (12/18)		
		basal	apical		basal	narrowed	none
S. pneumoniae	4	2	2	13	7	2	4
H. influenzae	2	2	0	3	3	0	0
N. meningitides	0	-	-	1	0	0	1
unknown	1	1	0	1	0	0	1

In the *partial-insertion group,* the degree of ossification was as follows:
Basal = complete ossification of the basal turn, no lumen encountered after up to 8 mm of drilling; Apical = ossification of the apical turn.
In the *full-insertion group*, the degree of ossification was as follows:
Basal = obliteration that required drilling to reach the natural lumen in the scala tympani; Narrowed = some bony ridges not causing any obliteration; None = fully patent basal turn.

Performance

Figure 1 presents the group minimum, mean and maximum preoperative unaided thresholds in the ear that was later implanted and the sound-field thresholds with a CI at 3-year follow-up as a function of frequency. Hearing thresholds that exceeded 130 dB HL were plotted at 130 dB HL. One child in the partial-insertion group was excluded, because he became a non-user one year postimplantation and therefore, no 3-year follow-up thresholds were available. Postimplantation hearing thresholds between the two groups were comparable, except at 0.25 kHz and 4 kHz. Unpaired t-tests showed significantly poorer aided thresholds in the partial-insertion group ($n = 6$) at 0.25 kHz ($P < 0.05$; 95% confidence interval 0.5 to 15 dB) and 4 kHz ($P < 0.01$; 95% confidence interval 2.4 to 15 dB) than in the control group ($n = 18$). The thresholds at 0.5, 1 and 2 kHz did not differ.

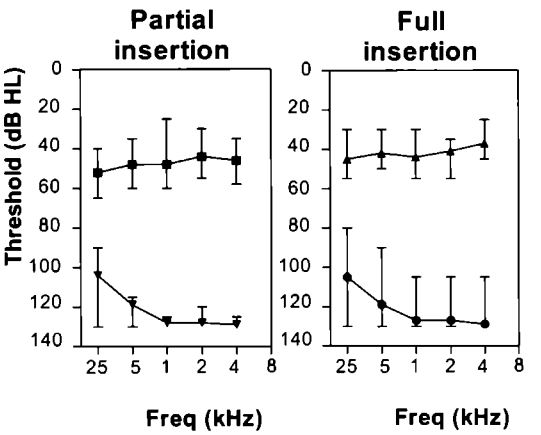


Figure 1. Unaided and aided thresholds
Minimum, mean and maximum preoperative unaided thresholds in the ear that was later implanted (▼ partial-insertion group, ●: full-insertion group) and the sound-field thresholds with a CI at 3-year follow-up (■ partial-insertion group; ▲ full-insertion group).

Speech perception scores expressed in EHL of all individuals are shown in Figure 2. Figure 3 shows the mean scores and standard deviations at each follow-up measurement, obtained from the partial-insertion group, the full-insertion group whose duration of deafness was longer than 3 years and the full-insertion group whose duration of deafness was shorter than 3 years. Speech perception improved over time in nearly all the children, but this occurred at various rates.

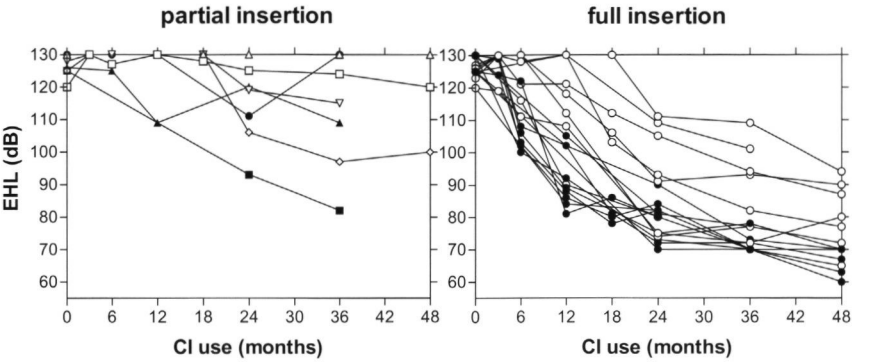


Figure 2. Speech perception expressed in EHL scores as a function of follow-up.
Left: children with partial insertion of the electrode array. The symbols refer to the different subjects: □ S1, Δ S2, □ S3, ◊ S4, ● S5, ▲ S6, ■ S7.
Right: control group with full insertion. Data from children with duration of deafness longer than 3 years (○) or shorter than 3 years with (●).

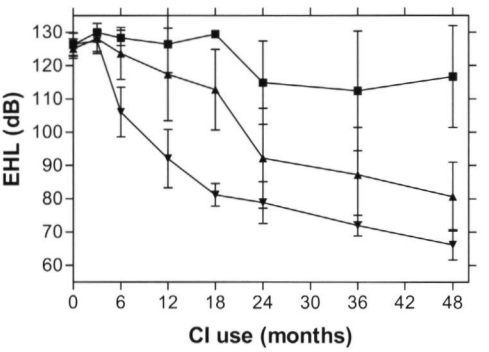


Figure 3. EHL group means and standard deviations as a function of follow-up.
The symbols refer to: ■ = partial-insertion group (n = 7); ▲ = full-insertion control group whose duration of deafness was longer than 3 years (n = 8); ▼ = full-insertion control group whose duration of deafness was shorter than 3 years (n = 10).

During the first 18 months postimplantation, most subjects with partial insertion made little or no progress. After one year of CI use, patient S3 became a non-user and in the long-term analysis his performance was plotted as 130 dB EHL. After 3 years of CI use, patient S7, the youngest implanted child with the shortest duration of deafness, had an EHL of 82 dB. This means that on a battery of speech perception tests, his performance was as good as that of severely hearing impaired children with well-fitted conventional hearing aids whose hearing loss was 82 dB HL. He was performing well within the range (70-109 EHL dB) of the prelingually deaf children with full insertion who had been deaf for longer than 3 years. The performance of the partial-insertion group was significantly poorer than that of the control group with a long ($P < 0.01$) or a short duration of deafness ($P < 0.0001$; Table 3).

When S3, i.e. the non-user, was excluded from the analysis, the difference between the partial-insertion group and the control group with a long duration of deafness was no longer significant. The children in the control group whose duration of deafness was longer than 3 years had significantly poorer scores on speech perception tasks than the children with a shorter duration of deafness.

Individual open-set phoneme scores of the children with partial insertion are shown in Figure 4, together with the mean scores of the control children with a long or short duration of deafness. Three years postimplantation, only patients S4 and S7 had achieved open-set word recognition scores that fell within the standard deviation of the control group with a longer duration of deafness. Patients S1 and S3 were able to recognize some phonemes. Patients S2, S5 and S6 were unable to perform the open-set speech recognition tests.

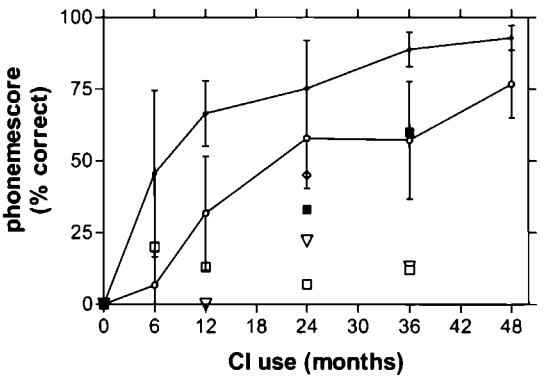


Figure 4. Individual phoneme scores of the children with partial insertion of the electrode array as a function of follow-up and the mean scores of the 2 control groups with full insertion.
The symbols refer to: ♦ = mean (with standard deviation) of the full-insertion control group whose duration of deafness was shorter than 3 years; ○ = mean (with standard deviation) of the full-insertion control group whose duration of deafness was longer than 3 years; partial insertion subjects: □ S1, Δ S2, ▽ S3, ◊ S4, ● S5, ▲ S6, ■ S7.

The small number of patients with partial insertion did not allow multivariate analysis to determine the influence of the number of active electrodes on speech perception performance. Patient S7 has the highest number of active electrodes and the best EHL score. However, patient S1 has only 8 active electrodes, her speech perception was better than that of patients S2 (the non-user) and S5 with 10 and 13 electrodes respectively (Figure 5). Thus, a higher number of active electrodes not necessarily mean higher speech perception scores.

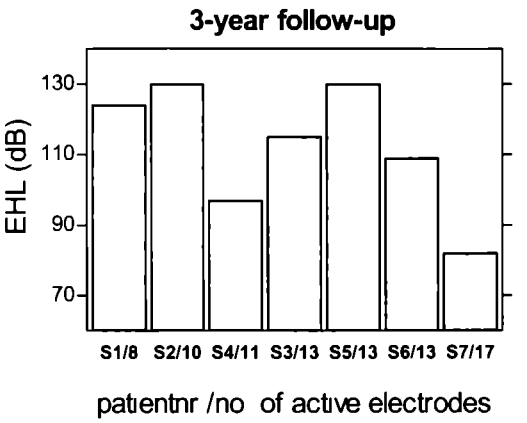


Figure 5 Number of active electrodes in the partial-insertion group (Patients S1-7) versus the EHL at 3-year follow-up

Discussion

Meningitis and deafness

In the present study, 76% of the children with postmeningitic deafness had some degree of ossification, which is compatible with previous reports [2] Obliteration was complete in five cases, in two of them, meningitis had been caused by *S pneumoniae* In 10 cases, the natural lumen was reached by drilling through the initial total obliteration, in seven of them, meningitis had been caused by *S pneumoniae* Eisenberg et al [17] and Becker et al [2] showed a definite relation between extensive ossification and *S pneumoniae* In 20 out of the 25 children (80%) with postmeningitic deafness Eisenberg found some degree of ossification Six had total obliteration and in five of them, meningitis had been caused by *S pneumoniae* To establish a statistical relation between the degree of ossification and the causative organism, larger numbers of subjects are required In smaller groups, as in this study, the incidence of meningitis and its etiological pathogens have to be borne in mind In the Netherlands, the incidence of *H influenzae* meningitis decreased rapidly after the introduction of Hib vaccination in 1993, whereas the incidence of *S pneumoniae* meningitis increased slightly Between '92 and '96, the number of cases with *N meningitidis* meningitis stabilised, at that time it was the most frequent cause of bacterial meningitis in the Netherlands The risk of developing postmeningitic hearing loss depends on the causative organism A review of the literature by Fortnum[1] showed that the incidence of permanent sensorineural hearing loss ranged from 21% to 50% for *S pneumoniae*, 5% to 10% for *N meningitidis* and 6% to 18% for *H influenzae*

Cochlear imaging

It is important to obtain accurate information on cochlear patency in the preoperative assessment process. The low sensitivity of CT scanning, as also reported by Young et al.[18] might be the result of the less dense structure of postinflammatory bone, which may be more fibrous, contain less calcium and consequently be less visible on CT scans. However, the CT technique, as applied, is relevant: higher sensitivity in predicting cochlear patency has been reported for recent CT techniques using axial and semi-longitudinal planes.[19] Otherwise, MRI is considered the imaging of choice to evaluate cochlear patency because it can detect the presence of fluid in the cochlear coil and visualize fibrosis.[20]

Auditory performance

The purpose of this study was to compare the performance of children with partial electrode insertion with that of children with full insertion. In children with full insertion aided thresholds were between 25 and 55 dB HL at frequencies of 0.25 to 4 kHz. These values are in agreement with those reported in the literature.[21] In the partial-insertion group, aided thresholds were somewhat poorer. In a battery of speech perception tests, however, the children with partial insertion had significantly poorer scores. Although children with a reduced number of electrodes achieve awareness of sound, apparently the auditory stimuli received are not always sufficient for them to recognize speech.

During 3 years of implant use, the speech perception of the children with partial insertion showed consistent but slow improvement. As a result of this slow rate of improvement, the difference between the partial-insertion group and the control group became more distinct as follow-up progressed (Figure 3).

Speech perception improved in six out of the seven partial-insertion subjects, whereas one child became a non-user. At 3-year follow-up, patient S4 had better speech perception than two of the control children with full insertion and a long duration of deafness. Remarkably, S7 had better scores than the mean score of the control group with a long duration of deafness. With 17 active electrodes his number of electrodes comes close to full insertion.

In S2, electrical stimulation of the implanted ear was no longer effective 1 year postimplantation. During surgery, we did not find any identifiable electrically evoked auditory brainstem responses (EABR) or stapedius reflex thresholds. Six weeks after surgery, electrical stimulation elicited a response from 10 electrodes. Responses to auditory stimuli during the rehabilitation period were inconsistent and further speech processor programming sessions were troublesome. At 9-months follow-up, no behavioural response could be obtained when the 10 electrodes were stimulated. Integrity measurements of the implant showed no abnormalities.[22] EABR measurements were performed under general anaesthesia, but again no identifiable responses were found up to the highest stimulation levels. CT scanning ruled-out extrusion of the electrode array from

the cochlea. In retrospect it might be questioned whether this subject had adequate hearing sensations at any time. He had a long duration of deafness (8 years and 7 months), which suggests that deprivation of the auditory system might have contributed to failure of the implantation.

The performance of patients with cochlear ossification and partial electrode insertion has been investigated by Kemink et al.[23] and Kirk et al.[24] They did not find differences in scores on selected speech perception tests after 6 to 18-month of follow-up between children with partial insertion and children with full insertion of the electrode array. Long-term results (4 to 5 years) were only reported in 2 of these children with partial insertion and their performance was similar to that of the control group with full insertion.[24] In these reports, the shorter follow-up period and longer duration of deafness in the control groups might explain the discrepancy with the present study. After 1 year of implant use, the difference between the partial and full-insertion groups was only significant for the subgroup with a duration of deafness of longer than 3 years. The subgroup with a shorter duration of deafness and the control group as a whole had better speech perception, even as early as at 1 year of implant use. Mean duration of deafness in the control group in Kirk et al.'s study[24] was 4.1 years, and 5 years in the study by Kemink et al.[23]

In studies performed in children and adults with partial insertion, Cohen and Waltzman[6] found poor speech perception results in most of their cases, while Beiter et al.[25] concluded from their experiments that the patients with partial insertion benefited from a CI, although not to the same degree as the patients with complete insertion. The present study is compatible with these findings. Rauch et al.[7] observed poor performance in patients with complete ossification that required total drill-out procedures (radical cochleotomy according to Gantz[8]). The range of performance in patients who required partial drill-out to achieve full electrode insertion most closely resembled that in patients who did not require any drilling.

Presumably, patients who experience more specific auditory stimuli, as delivered by an implant with a larger number of different channels, can detect the features of speech more accurately and thus achieve better scores on speech perception tasks than patients who experience a less differentiated auditory environment, as delivered by fewer active channels. Kileny et al.[26] investigated how speech recognition was affected by a reduced number of active electrodes inserted into the basilar end of the cochlea. They observed a trend towards increased scores in open-set speech recognition tasks when all 20 electrodes of the array were activated, compared to activation of the 10 basal channels only. In the present study also, the full-insertion group, with an average of 20 active electrodes, had significantly better speech perception than partial-insertion subjects in whom 8 to 13 electrodes had been implanted. In the partial-insertion group with 8 to 13 active electrodes, there was no relation between speech perception scores and the actual number of active

(/

electrodes (Figure 5) Besides the relatively small number of subjects with partial insertion, other factors such as duration of deafness and age at onset may also play a role in performance variability

Besides the reduced number of electrodes, there are other explanations for poor speech perception when severe ossification leads to partial insertion In these cases, it is the goal to drill-out the ossified cochlea and place the electrode array as close to the modiolus as possible, without disrupting it by drilling The typical structure of the neo-ossification serves as a guide to the direction of the axis of the pars inferior of the scala tympani In some cases, it might not be possible to achieve optimal modiolus-array proximity Another explanation is that the electrical current in a drilled tunnel may be broadly spread, which is less favourable Furthermore, the integrity of surviving spiral ganglion cells and auditory nerves are of concern in determining the benefit of cochlear implantation, especially in children with postmeningitic deafness Patients with auditory nerve lesions have been known to benefit from cochlear implantation In fact, most patients with severe primary end organ (hair cell) disease have retrograde neural degeneration to some degree, and also these patients became successful CI users [27] It is not easy to make a preoperative evaluation of the functional capacity of the peripheral or central auditory system in children with prelingually deafness Some believe that intra-operative brainstem and cortical AEPs measurements and neural response telemetry (NRT) may be helpful to predict neural integrity, the survival rate of spiral ganglion cells, the integrity of the central auditory pathways and the functional prognosis of cochlear implantation [28] However, these measurements are still under evaluation and have not yet become fully implemented in CI assessment

Conclusions

Ossification of the cochlea is not necessarily a contraindication for cochlear implantation Despite normal cochlear appearance on CT scans, the presence of ossification must be expected in a child with postmeningitic deafness, thus additional MRI is mandatory

Some open-set comprehension could even be achieved with the insertion of only 8 electrodes of a Nucleus device Over a 3-year follow-up period, the children with partial insertion showed continuing progress, although there was wide variation in performance and the rate of progression On average, their rate of progress was lower than that of the control group The long-term results at 3-year follow-up were significantly poorer than those in children with postmeningitic deafness and full insertion of the electrode array

References

1. Fortnum HM. Hearing impairment after bacterial meningitis: a review. *Arch Dis Child* 1992;67:1128-1133
2. Becker TS, Eisenberg LS, Luxford WM, House WF. Labyrinthine ossification secondary to childhood bacterial meningitis. implications for cochlear implant surgery. *AJNR Am J Neuroradiol* 1984;5:739-741.
3. Hinojosa R, Redleaf MI, Green JD, Jr, Blough RR. Spiral ganglion cell survival in labyrinthitis ossificans: computerized image analysis. *Ann Otol Rhinol Laryngol* 1995; Suppl 166:51-54.
4. Jiang ZD., Liu XY, Wu YY, Zheng MS, Liu HC Long-term impairments of brain and auditory functions of children recovered from purulent meningitis. *Dev Med Child Neurol* 1990,32.473-480.
5. Nadol JB, Jr., Hsu WC. Histopathologic correlation of spiral ganglion cell count and new bone formation in the cochlea following meningogenic labyrinthitis and deafness *Ann Otol Rhinol Laryngol* 1991;100:712-716
6. Cohen NL, Waltzman SB. Partial insertion of the nucleus multichannel cochlear implant: technique and results. *Am J Otol.* 1993;14:357-361.
7. Rauch SD, Herrmann BS, Davis LA, Nadol JB, Jr Nucleus 22 cochlear implantation results in postmeningitic deafness. *Laryngoscope* 1997,107:1606-1609.
8. Gantz BJ, McCabe BF, Tyler RS. Use of multichannel cochlear implants in obstructed and obliterated cochleas. *Otolaryngol Head Neck Surg* 1988;98:72-81.
9. Steenerson RL, Gary LB, Wynens MS (1990) Scala vestibuli cochlear implantation for labyrinthine ossification. *Am J Otol* 1990;11:360-363.
10. Lenarz T, Buchner A, Tasche C, Cristofoli T, Lesinski-Schiedat A, Wallenberg EV, Battmer RD, Busby PA, Frohne C. The results in patients implanted with the nucleus double array cochlear implant: pitch discrimination and auditory performance. *Ear Hear* 2002;23:90S-101S.
11. Mitchell TE, Psarros C, Pegg P, Rennie M, Gibson WP. Performance after cochlear implantation: a comparison of children deafened by meningitis and congenitally deaf children. *J Laryngol Otol* 2000;114:33-37
12. O'Neill C, O'Donoghue GM, Archbold SM, Nikolopoulos TP, Sach T. Variations in gains in auditory performance from pediatric cochlear implantation. *Otol Neurotol* 2002;23:44-48.
13. Hodges AV, Balkany TJ, Gomez-Marin O, Butts S, Ash SD, Bird P, Lee D. Speech recognition after implantation of the ossified cochlea. *Am J Otol* 1999;20:453-456
14. Osberger MJ, Fisher L. Preoperative predictors of postoperative implant performance in children. *Ann Otol Rhinol Laryngol Suppl* 2000;185:44-6
15. Snik AF, Vermeulen AM, Brokx JP, Beijk C, van den Broek P. Speech perception performance of children with a cochlear implant compared to that of children with conventional hearing aids. I. The "equivalent hearing loss" concept *Acta Otolaryngol* 1997;117:750-754
16. Snik AF, Vermeulen AM, Geelen CP, Brokx JP, van den Broek P. Speech perception performance of children with a cochlear implant compared to that of children with conventional hearing aids. II. Results of prelingually deaf children *Acta Otolaryngol* 1997;117:755-759.
17. Eisenberg LS, Luxford WM, Becker TS, House WF Electrical stimulation of the auditory system in children deafened by meningitis. *Otolaryngol Head Neck Surg* 1984,92:700-705.
18. Young NM, Hughes CA, Byrd SE, Darling C Postmeningitic ossification in pediatric cochlear implantation *Otolaryngol Head Neck Surg* 2000,122, 183-188
19. Langman AW, Quigley SM Accuracy of high-resolution computed tomography in cochlear implantation. *Otolaryngol Head Neck Surg* 1996,114.38-43.
20. Iwasaki S, Atsumi K, Ocho S, Mizuta K Facial nerve stimulation by a cochlear implant in a hemodialysis patient with bone of low mineral density. *Eur Arch Otorhinolaryngol* 1998;255:352-354.

21. Staller SJ, Dowell RC, Beiter AL, Brimacombe JA. Perceptual abilities of children with the Nucleus 22-channel cochlear implant. *Ear Hear* 1991;12.34S-47S.
22. Mens LH, Oostendorp T, van den Broek P. Identifying electrode failures with cochlear implant generated surface potentials. *Ear Hear* 1994,15:330-338.
23. Kemink JL, Zimmerman-Phillips S, Kileny PR, Firszt JB, Novak MA. Auditory performance of children with cochlear ossification and partial implant insertion. *Laryngoscope* 1992;102·1001-1005.
24. Kirk KI, Sehgal M, Miyamoto RT. Speech perception performance of nucleus multichannel cochlear implant users with partial electrode insertions. *Ear Hear* 1997;18:456-471.
25. Beiter AL, Brimacombe JA, Fowler-Brehm N, Sinopoli TA, Segel PA. Results with a multichannel cochlear implant in individuals with ossified cochleae. In Hochmair-Desoyer IJ, Hochmair ES (eds) Advances in cochlear implants, pp. 462-466. 1994 Manz, Wien
26. Kileny PR, Zimmerman-Phillips S, Zwolan TA, Kemink JL. Effects of channel number and place of stimulation on performance with the Cochlear Corporation multichannel implant. *Am J Otol* 1992;13:117-123.
27. Spoendlin H, Schrott A. Analysis of the human auditory nerve. *Hear Res* 1989;43:25-38
28. Gordon KA, Ebinger KA, Gilden JE, Shapiro WH. Neural response telemetry in 12- to 24-month-old children. *Ann Otol Rhinol Laryngol* 2002;Suppl 189·42-48.

Chapter 4

Cochlear implantation in the malformed cochlea

Congenital malformation of the inner ear and pediatric cochlear implantation

E.A.M. Mylanus
L.J.C. Rotteveel
R.L. Leeuw

Otology & Neurotology 2004, 25 308-317

Abstract

Objectives To study the surgical aspects and performance outcome of cochlear implantation in children with malformed inner ears

Study design Clinical and audiometric evaluation in 13 patients

Methods Patient data concerning surgery, postoperative follow-up, and pre- and postimplantation audiometry were obtained from the cochlear implant center's database and evaluated A review of the literature has been included

Patients The patients had a variety of inner ear malformations and profound hearing loss One patient with recurrent meningitis had a severe cochlear malformation (common cavity)

Results Major complications did not occur In one patient with an abnormal position of the cochlea and concurring middle ear pathology, it was difficult to find the scala tympani during surgery Cerebrospinal fluid gusher was encountered in two patients and an aberrant facial nerve in another, which did not lead to any complications Patients with mild cochlear malformation like an incomplete partition demonstrated a good performance in speech perception tests Even the child with the common cavity deformity had some open-set speech perception 1 year after implantation

Conclusions Viewing the patients from this study and patients from a review of the literature concerning cochlear implantation in children with malformed inner ears including severe cochlear malformations, the occurrence of an aberrant facial nerve was 17%, which increases to 27% if one reviews the surgical findings in children with severe malformed cochleae such as common cavity or a severe cochlear hypoplasia In the latter patients, results in speech perception vary Although the result of cochlear implantation may be promising, as in our patient with a common cavity, during preoperative counseling the child's parents must be informed that the result is uncertain

Introduction

In the early days of pediatric cochlear implantation, the majority of the patients consisted of children with postmeningitic profound bilateral sensorineural hearing loss. Nowadays, an increasing proportion of the children scheduled for cochlear implantation have congenital profound hearing loss. According to Jackler et al.[1], 20% of all cases of congenital profound hearing loss have bony abnormalities of the labyrinth. More recent studies report this incidence to be even more (i.e., 30%) because of improvements in high-resolution computed tomographic (CT) scan techniques and a heightened awareness of cochlear malformations.[2] It is therefore not surprising that there has been an increase in the number of reports on the results of cochlear implantation in malformed cochleae in the past decade.[3-12]

To classify the various malformations and correlate surgical issues and rehabilitation outcome to certain types of malformation, most reports make use of the classification based on embryonic life suggested by Jackler et al.[1] The stage at which the embryonic development of the cochlea is arrested produces a malformation with a certain degree of severity. Thus, a malformation of the cochlea may vary from total aplasia, severe cochlear hypoplasia, mild cochlear hypoplasia (basal turn only), common cavity, severe incomplete partition, mild incomplete partition to a subnormal cochlea that does not reach a full 2.5 turn. The cochlear malformation may be presenting with a variety of bony abnormalities of the vestibule or semicircular canals or an enlarged vestibular aquaduct. Cochlear malformation presents technical problems for cochlear implant (CI) surgery, most notably the anomalous facial nerve and cerebrospinal fluid (CSF) gusher.[9,15] Also, postoperatively, during activation and programming, specific problems can occur and frequent reprogramming may be needed.[4,5,9]

In this study, the surgical aspects of 13 children are described. The audiometric results of cochlear implantation in 10 children with inner ear malformations are discussed and compared with those of a control group consisting of 10 matched implanted children with a normal cochlea. A review of the literature is presented, focussing on the results of cochlear implantation in children with severe cochlear malformations; the common cavity and severe cochlear hypoplasia.

Methods

Between 1994 and 2002, 13 children with inner ear malformations and severe hearing loss or deafness underwent multichannel cochlear implantation at the Nijmegen/Viataal CI centre. Ten of the 13 children with a follow-up of at least one year were matched with children with a normal cochlea who had received CIs. They were matched for variables

that are known to influence performance with CI: age at implantation, duration of deafness and electrode insertion depth. In Tables 1 and 2, the most important patient characteristics and surgical aspects are shown. Eleven children with malformed cochlea were considered deaf from birth, and underwent implantation at an average age of 4.0 years. Patients 2 and 3 suffered progressive fluctuating hearing loss as a result of the enlarged vestibular aqueduct (EVA) syndrome and had confirmed profound deafness for 2 years. They underwent implantation at an age of 6.5 and 7.3 years, respectively. Their matched pairs, one subject deafened by an unknown progressive cause and the other by meningitis, had been deaf for 1.3 and 2.5 years and underwent implantation at an age of 4.5 and 7.7 years, respectively. The other congenitally deaf control subjects were implanted at an average age of 3.3 years. Patients 12 and 13 had CHARGE (coloboma, heart defects, atresia choanae, retardation of growth and/or development, genital hypoplasia, and ear anomalies and/or deafness) association. During preimplant assessment, all children were tested with tonal and behavioural audiometry in an unaided and aided situation to confirm severe hearing loss or deafness.

Table 1 Patient characteristics and surgical aspects

Patient	Hearing Loss	Vestibular tests	Age at Impl (years)	CT: Cochlear malformation	CT: labyrinthine, IAC or vestibular aqueduct malformation	Intraoperative complications
1a	Congenital	areflexia	5 7	Severe IP	Dysplastic vestibule and canals	CSF gusher
2a	Progressive	normal	6.5	Normal	EVA	
3a	Progressive	normal	7 3	Mild IP	EVA	
4a	Congenital	areflexia	3 8	Mild IP	Dysplastic LSC, wide IAC	
5a	Congenital	normal	2 9	Mild IP	Normal	
6a	Congenital	normal	2 5	Mild IP	Normal	
7a	Congenital	normal	1 1	Normal*	Dysplastic vestibule and canals	Exposed carotid artery
8a	Congenital	normal	2 0	Mild IP	Normal	
9a	Congenital	hypofunction	2.5	Severe IP	EVA, dysplastic vestibule and canals	
10a	Congenital	NT	6 2	CC	Aplastic canals	Aberrant facial nerve
11	Congenital	normal	7 2	Severe IP	EVA, dysplastic vestibule	CSF gusher
12	Congenital	NT	6 7	Mild CH	Aplastic canals, obliterative oval window	
13	Congenital	NT	3 1	Mild CH	Aplastic canals, obliterative oval window	Stapes and incus removed for access

NT = not tested, Age at Impl = age at implantation, CT = computed tomographic scan; IP = incomplete partition; CC = common cavity; CH = cochlear hypoplasia, EVA = enlarged vestibular aqueduct, IAC = internal auditory canal, LSC = lateral semicircular canal; CSF = cerebro spinal fluid, * flat promontory and medially rotated cochlea

Table 2. Matched pairs control group, patient characteristics and surgical aspects

Patient	Onset of HL [age in years]	Cause of Hearing Loss	Vestibular tests	Age at Impl (years)	CT scan findings	MRI findings	Intra-, and postoperative complications
1b	Congenital	Unknown	Areflexia	5 1	No abnormalities		
2b	Postlingual [3 2]	Unknown, progressive	Normal	4 5		No abnormalities	
3b	Postlingual [5 2]	Meningitis	Areflexia	7 7	Normal cochlea, ossification SSC		
4b	Congenital	Unknown	Areflexia	3 7	No abnormalities		Postoperative otorrhea
5b	Congenital	Unknown	Areflexia	2.9	No abnormalities		
6b	Congenital	Unknown	Areflexia	2 4	No abnormalities		
7b	Congenital	Meningitis	Areflexia	1 0	No abnormalities	Basal ossification	
8b	Congenital	Hereditary	Normal	2 0	No abnormalities		
9b	Congenital	Unknown	Normal	2 5	No abnormalities		
10b	Congenital	Unknown	Areflexia	6 7	No abnormalities		

Age at Impl = age at implantation; CT = computed tomographic; MRI = magnetic resonance imaging, SSC = superior semicircular canal

The inner ear malformations were diagnosed with high-resolution CT (HRCT) scanning. For patients 9 to 13, this was supplemented with magnetic resonance imaging (MRI). The severity of the cochlear malformation was graded on the basis of the embryologic concepts underlying cochlear malformations outlined by Jackler et al.[1] (total aplasia, severe cochlear hypoplasia, mild cochlear hypoplasia (basal turn only), common cavity, severe incomplete partition and mild incomplete partition). All images were reassessed for this study by a radiologist specialised in the imaging of the petrosal bone. Any abnormalities of the internal auditory canal, vestibule, semicircular canals, facial nerve and vestibular aqueduct were noted.

The average follow-up was 3.5 years (range 1.0 to 9.0 years) in the group of children with inner ear malformations and 4.7 years (range 2.0 to 7.5 years) in the group with matched pairs. Postimplant performance was tested using free-field thresholds and two open-set word tests consisting of lists of CVC monosyllables. The difference between the Gestel/Nijmegen test and the Bosman test is the difficulty of the word material, the latter using more uncommon words.[14] In case the perception scores could not be obtained because of a limited follow-up or young age, the reaction of the child with a CI to sound was commented on

Table 3. Pre- and postoperative performance

atient	Follow-up (yrs;mo)	Preoperative unaided thresholds (dB HL) (0.5-1-2-4 kHz)	Preoperative aided thresholds (dB HL) (0.5-1-2-4 kHz)	Postoperative thresholds CI (0.5-1-2-4 kHz)	GN open-set phonemes (follow-up)	Bosman open-set phonemes (follow-up)	Comments
a	9,0	110-NM-NM-NM	70-75-NM-NM	45-40-40-35	83% (5 yrs)	73% (8 yrs)	
a	7;2	105-115-110-105	75-65-75-65	45-50-45-40	95% (5 yrs)	89% (6 yrs)	
a	4;6	80-80-80-75	40-40-30-35	35-40-40-35	90% (1 yr)	88% (4 yrs)	
a	4,7	115-NM-NM-NM	75-80-80-NM	40-40-35-40	82% (4 yrs)		
a	4,0	100-NM-NM-NM	80-90-NM-NM	40-40-35-30	80% (3 yrs)		
a	4,2	NM-NM-NM-NM	65-60-70-65	45-40-40-35	70% (3 yrs)		
a	2,6	NM-NM-NM-NM	80-75-85-90	55-50-55-50			54% Erber (2 yr
a	2,4	105-120-NM-NM	65-50-70-70	40-40-45-35	78% (2 yrs)		
a	2,0	110-110-120->120	55-60-75-NM	55-55-65-45			75% Erber (2 yr
0a	2,0	NM-NM-NM-NM	95-95-NM-NM	50-40-45-45	40% (1 yr)		
1	1,1	90-105-110-NM	40-45-65-100	45-40-40-30	68% (1 yr)		
2	1,0	NM-NM-NM-NM	90-NM-NM-NM	55-55-55-55			Discriminates sounds
3	1;0	105-120-120-120	80-85-90-95	45-45-45-50			Discriminates sounds

Preoperative unaided and aided thresholds were measured in a free-field set-up Only the lowest thresholds are given, irrespective of the ear In patients 1a, 2a and 3a the contralateral (worse) ear was implanted. Yrs = years, mo = months; HL = hearing loss; NM = not measurable, GN = Gestel/Nijmegen open-set phoneme test, Bosman = open-set phoneme test, less usual words; Erber = spondee recognition test for young hearing-impaired children, CI = cochlear implant

Table 4. Matched pairs control group. Pre- and postoperative performance

atient	Follow-up (yrs;mo)	Preoperative unaided thresholds (dB HL) (0.5-1-2-4 kHz)	Preoperative aided thresholds (dB HL) (0.5-1-2-4 kHz)	Postoperative thresholds CI (0 5-1-2-4 kHz)	GN open-set phonemes (follow-up)	Bosman open-set phonemes (follow-up)	Comments
b	6,9	NM-NM-NM-NM	100-NM-NM-NM	50-50-50-45	67% (6 yrs)		
b	4;6	125-135-NM-NM	85->110-NM-NM	30-30-25-30	98% (4 yrs)		
b	7,5	110-125-125->130	75-70-NM-NM	30-30-35-35	80% (4 yrs)	94% (5 yrs)	
b	6,7	NM-90-NM-NM	60-55-70-NM	45-40-40-35	55% (3 yrs)	91% (5 yrs)	
b	5,8	105-NM-NM-NM	85-90->90-NM	40-35-30-30	100% (4 yrs)	64% (5 yrs)	
b	2,0	120-130-130-NM	95-100-95->100	40-35-35-35			100% Erber (yrs)
b	2,10	NM-NM-NM-NM	100-110-110-NM	45-45-45-45	87% (2 yrs)		
b	3,11	NM-100-115-120 *	NM-NM-65-85 *	45-45-40-40	90% (3 yrs)	73% (3 yrs)	
b	4;1	NM-NM-NM-NM	75-75-80-NM	40-35-40-40	23% (3 yrs)		
0b	3,4	100-105-NM-NM	70-65-80-95	45-35-35-35	82% (3 yrs)	85% (3 yrs)	

Yrs = years, mo = months, HL = hearing loss, NM = not measurable, GN = Gestel/Nijmegen open-set phoneme test; Bosman = open-set phoneme test; ; Erber = spondee recognition test, CI = cochlear implant, * dB(A)

Results

Adequacy of matching

Descriptive data and performance data for the case patients and control subjects are shown in Tables 1 to 4. For the matched congenitally deaf children, the mean age at implantation

(also duration of deafness) in both groups was 3 3 years As all children had full insertion, the subjects were completely matched for insertion depth The control group comprised only mentally and physically healthy children Patient 9a however, with severe incomplete partition and EVA syndrome, has a slight cognitive developmental delay and attends a special school for the deaf She was born with an occipital meningocele and a cerebral Arnold-Chiari malformation Type 2 Patients 12 and 13 have CHARGE-association with typical findings including retardation of growth and cognitive development In all other children, the malformation was an isolated finding

Surgery

Intraoperative complications are shown in Tables 1 and 2 A standard surgical procedure was performed in all patients with malformed cochlea except for Patient 10a, who is presented in more detail below Cortical mastoidectomy and the posterior tympanotomy approach of the middle ear provided access to the round window niche without damage to the chorda tympani or to the facial nerve In Patient 1a, who had a severe incomplete partition, and in Patient 11, with mild cochlear hypoplasia, CSF gusher was encountered but managed with packing of the cochleostomy with periosteum In all patients a complete insertion of all active electrodes was accomplished In Patient 7a, great difficulties were encountered in performing the cochleostomy Aside from a flat promontory and a medial rotation of the cochlea, there was an abundant hypertrophy of the middle ear mucosa in this child who had a history of recurrent otitis media with effusion The first attempt to locate the scala tympani resulted in exposure of the adventitia of the carotid artery After removal of all middle ear mucosa from the promontory and maximum exposure of the sinus tympani, the position of the oval and round window could be assessed and the scala tympani was found A small tear of the dura occurred at the site of the implant package The resulting CSF leak was managed adequately with the use of tissue glue and bone dust The implant was fully recessed in the temporal bone (contrary to most children who have undergone implantation in recent years) As in all children, a headdressing was maintained for one week postoperatively In this child, a headtrauma occurred resulting in a swelling over the implant site Aspiration demonstrated sanguinous fluid and a new headdressing was placed for another week No complications occurred afterwards in this or in any other child

Patient 10a is described in more detail This congenitally deaf girl was presented to us at an age of 5 5 years She had a history of recurrent meningitis that had been treated with intravenous antibiotics High-resolution CT-scanning showed bilateral common cavities (Figure 1), aplasia of the semicircular canals, and at the left side a fluid-filled mastoid The internal auditory canals were normal MRI demonstrated the presence of the VIIIth cranial

nerve and a probable intact cribriform plate (Figure 2). An explorative tympanotomy was performed in the left ear, confirming a small leak of CSF through the anterior portion of the oval window. The oval window was malformed and in a more inferior position than normal. The leak was sealed with temporal fascia and tissue glue. As suspected on high-resolution CT-scanning an aberrant facial nerve was present, with it's course in a more inferior position than normal. Unfortunately, the meningitis recurred several months later. It was decided to perform a subtotal petrosectomy and a cochlear implantation, followed by a total obliteration with abdominal fat and closure of the external auditory canal.[15] The approach to the common cavity was through a labyrinthotomy, where one would expect the lateral semicircular canal in normal inner ears. No CSF gusher was found and complete insertion was accomplished with an uncoiled, straight electrode array. The final position of the electrode was checked radiographically before packing the cochleostomy. There were no postoperative complications.

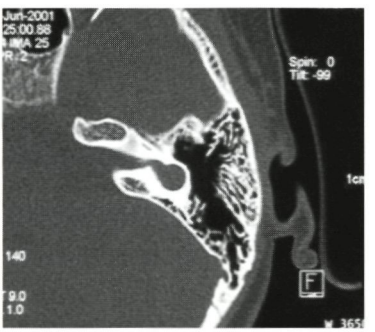


Figure 1. Patient 10a, CT scan, axial view of the left petrosal bone: common cavity, aplastic semicircular canals.

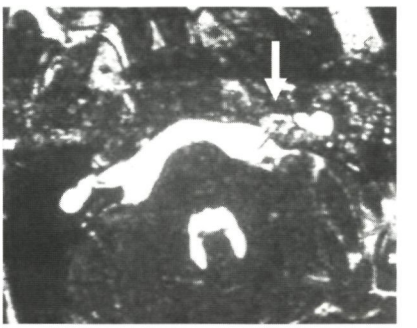


Figure 2. Patient 10a, MRI, axial view: bilateral common cavities, intact cribriform plate (*arrow*).

Audiology

In Tables 3 and 4, the most important results of audiometric testing and speech tests are shown. Preoperative aided and unaided free-field thresholds of the implanted ears are shown. Bilateral profound hearing loss was demonstrated in 11 children with congenital malformations. In Patient 2a, who had progressive hearing loss, a severe hearing loss was detected in the non-implanted and a profound loss in the ear planned for implantation. In Patient 3a, the preoperative thresholds (aided and unaided) would normally have precluded cochlear implantation. However, in this child the thresholds were fluctuating in a decapacitating manner. In Table 3 this child's lowest thresholds in the non-implanted ear are shown.

After implantation, in 12 children with congenital malformations threshold (T) and maximum comfortable (C) levels did not show abnormalities compared with children with

normal cochleae and could be measured in a normal way with respect to their age. In Patient 10a only a limited number of electrodes had a normal T level. On the remaining electrodes, T levels approached the limits of the equipment. Therefore, in the latter electrodes, the dynamic range between T and C levels was small. The thresholds with the CI in the free field for narrow-band noises were in accordance with the expected ones for that microphone sensitivity, although in some patients the thresholds were somewhat elevated. This might be explained by age and follow-up (Tables 3 and 4).

Speech perception scores obtained at the most recent audiometric session are shown in Tables 3 and 4. At 1-year follow-up, for most children open-set phoneme scores could be measured. Some patients, however, had limited language abilities and did not have an open speech perception yet (Patients 6b, 7a, 9a, 12, 13), possibly because of young age, long duration of deafness or short follow-up. The poor language skills of these children precluded the use of standard tests of speech perception. However, they demonstrated closed-set speech perception, or at least an increased awareness of environmental sounds by responding to sounds or their names. The length of device use is said to be one of the most important variables influencing performance in young congenitally deaf children with implants.[3] Therefore, for the first 6 patients with considerable follow-up and their matched pairs the open-set speech perception scores at 1-year intervals after implantation are shown in Figure 3. Although some data are missing, there is no great difference in performance between the two groups of patients. As a result of a recent change in coding strategy, Patient 5b showed some deterioration in speech perception at the 5-year postimplantation evaluation session.

Discussion

Of foremost importance in the technical feasibility of cochlear implantation in profoundly deaf children with malformed cochlea is to determine whether there is sufficient cochlear lumen for electrode placement and to rule out VIIIth cranial nerve aplasia or hypoplasia. This means that imaging is of great importance. The degree of patency of the cochlear duct can only be reliably assessed by magnetic resonance imaging (MRI). Aplasia of the VIIIth cranial nerve also needs to be ruled out with MRI, especially in patients with a common cavity abnormality, a narrow internal auditory meatus visualised on CT scan (i.e., < 2 mm in diameter), or in patients with CHARGE syndrome.[2,16,17] Isolated absence of the cochlear nerve is very rare.[18] If appropriate, electrophysiologic tests, such as promontory auditory brainstem responses(ABR), can give additional information on the neural pathway.[17,19] At our CI centre, MRI scanning is routinely performed in postmeningitic deaf children and in children with inner ear malformations.

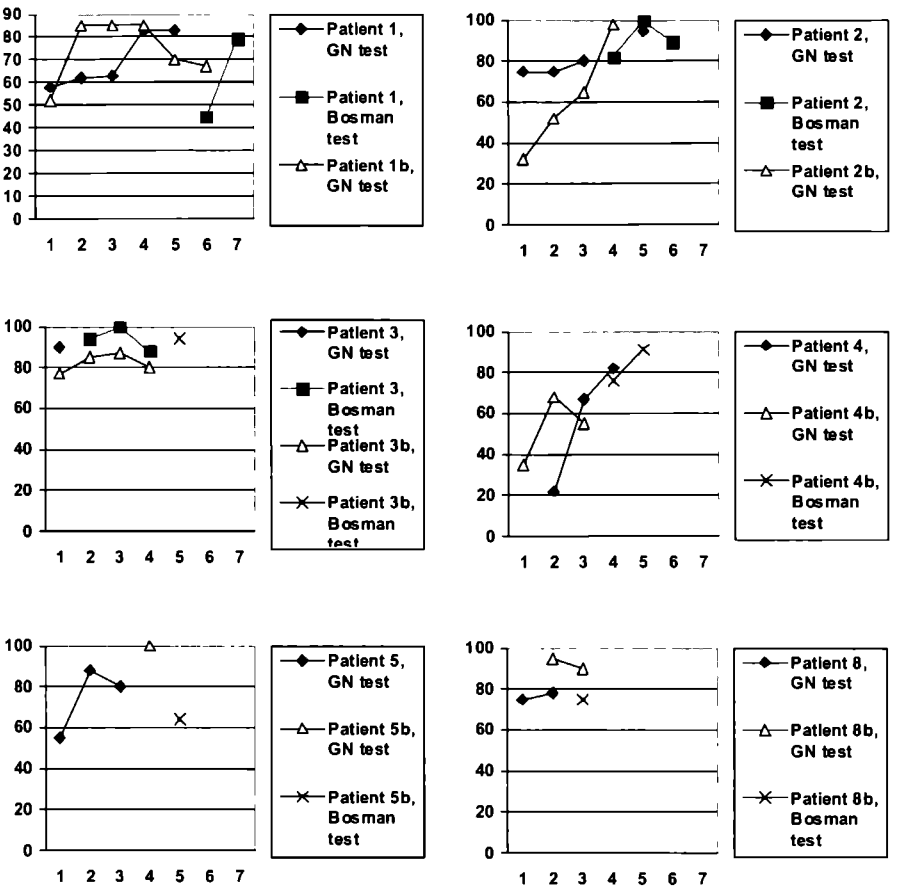


Figure 3. Open-set speech perception, phoneme scores (% correct) of 6 matched pairs.
GN test = Gestel/Nijmegen open-set phoneme test; Bosman = Bosman open-set phoneme test; X-axis, years of follow-up.

Although the surgical procedure is considered feasible, cochlear implantation may be more difficult in children with malformed inner ears as a result of the abnormal anatomy of the temporal bone, the possibility of an aberrant course of the facial nerve, and the occurrence of CSF gusher.

Aberrant facial nerves were reported in 16% of inner ear malformations in general[9], and noted more frequently in patients with a severe malformation such as a common cavity or a severe hypoplastic cochlea.[8,20] In some patients with an aberrant facial nerve a canal-wall-down procedure was performed instead of the standard transmastoid facial recess approach, to gain safe access to the cochlea.[5,12] McElveen et al.[8] described the transmastoid

labyrinthotomy approach to common cavity malformations to minimize the risk of injuring an aberrant facial nerve and to have better control of a possible CSF gusher The use of a facial nerve monitor in this particular group of patients is strongly advised by most surgeons In our patient with the common cavity deformity and the aberrant facial nerve, the facial nerve monitor was considered a valuable attribute during surgery

Gushers of CSF usually are the result of a bony defect of the cribriform plate, causing abnormal communication between the cochlea and the subarachnoid space The gusher in enlarged vestibular aqueduct syndrome tends to be significantly less than in other malformations [21] In our patient group only 2 patients with a severe incomplete partition had a CSF gusher, which was managed with simple packing of the cochleostomy with periosteum Because CT scanning and MRI of the girl with the common cavity deformity ruled out a patulous cribriform area, CSF gusher was not expected preoperatively and indeed did not occur In the study of Eisenman et al [17], preoperative CT scanning demonstrated a patulous communication between the lateral portion of the internal auditory canal and the cochlea in all 7 patients who had intraoperative flow of CSF However, sometimes CT scanning demonstrated patulous communication in patients in whom CSF outflow did not occur Sufficient packing of the cochleostomy with soft tissue is of importance Postoperative leakage of CSF poses a risk for meningitis which may even occur several months postoperatively [3]

Some children with inner ear malformation are at risk for meningitis as a result of an abnormal communication between the anomalous cochlea and internal auditory canal, whether a cochlear implantation has been performed or not [22] The common cavity malformation is an important precursor of otogenic meningitis and surgery is indicated in the case of a suspected leakage In our patient group, only Patient 10a with bilateral common cavities, suffered from recurrent meningitis preoperatively During an explorative tympanotomy, the leakage of CSF through the oval window, the alleged cause of infection, was sealed In order to further reduce the chance for meningitis, after cochlear implantation, the ear was obliterated and the external auditory canal was closed None of the patients have had postoperative meningitis

In Table 5, the surgical results are given for 12 previous studies concerning cochlear implantation in children with malformed inner ears that have included patients with severe cochlear malformations such as common cavities and severe cochlear hypoplasia Including our own data, a total of 81 children with inner ear malformation are listed, including 23 ears with a common cavity deformity and 10 ears with cochlear hypoplasia (one patient was implanted bilaterally) In 3 of the 7 studies that included patients with

cochlear hypoplasia, it is clear that the patients described had a severe cochlear hypoplasia according to Jackler's classification.[1,5,7,11] Including the current study, an aberrant facial nerve was found in 14 ears (17%), of which (at least) 9 were in the 33 ears with severe cochlear malformations, which is 27%. Postoperative facial nerve palsy has only been reported twice, of which one was transient.[4,7] CSF gushers were encountered in 32 ears (40%), of which (at least) 9 in severely malformed cochleae and at least 11 in cochleae with an incomplete partition. These data are still in agreement with the data presented by Hoffman[9] from a literature review (23 patients) and a questionnaire study (23 patients).

A CI depends on the presence of spiral ganglion cells and cochlear nerve fibres. As the exact location of neural tissue within a severely malformed cochlea such as a common cavity deformity is unknown, optimal insertion of the electrode array may be difficult to achieve. Histologic studies have shown that neural elements may be present on the outer wall of the cavity.[13] For this reason, an uncoiled electrode was used in our patient with a common cavity. Because of the risk of entering the internal auditory canal with this straight electrode array, we performed perioperative anteroposterior X-ray imaging to check the final position of the electrode before packing the cochleostomy, as has been suggested by others.[9] Considering the mobility of the electrodes in the cavity, initial fluctuation in thresholds may be expected, requiring frequent reprogramming of the electrodes.[5,13] In our patient with a common cavity no fluctuations were found. By performing peroperative neural respons threshold (NRT) measurements, a more optimal positioning of the electrode array can be achieved.

Several clinics have reported worthwhile benefit of cochlear implantation in children with inner ear malformation.[3,4,9,10,13] This is certainly true for children with labyrinthine abnormalities and normal cochleae, as in the isolated enlarged vestibular aqueduct syndrome.[23,24] Generally, in patients with mild cochlear deformities as mild or severe incomplete partition, full insertion of the electrode array is possible and results can be obtained comparable to those obtained in profoundly deaf patients with normal cochleae.[13] This was also observed in our Patients 1a to 6a. In children with CHARGE-association, mild cochlear dysplasia occurs, allowing full insertion of the electrode array, yet results may vary as motor and cognitive delays may impede the improvement of speech perception after cochlear implantation.[25]

Table 5 Summary of surgical data obtained from studies which include children with severely malformed cochleae (common cavity, severe cochlear hypoplasia)

Author(s)	N	Normal cochlea	Severe cochlear malformation	Aberrant facial nerve	Canal wall down, earcanal closure (indication)	Approach for CC	CSF gusher	Facial nerve stimulation	Insertion depth
Jackler et al (1987)	4	0	CC (2) and CH (2)	1 dehiscent	0	Facial recess	1 (CC)	2 (1 IP, 1 CC)	-
Slattery and Luxford (1987)	10(3 ad)	2 (ad)	CC (2) and CH (1)	2 (CH* IP)	1 (CH) (access)	Facial recess	4 (2 CC 1 IP, 1 EV)	0	CC single electrode CC/CH incomplete
Molter et al (1993)	1	0	CC (1)	1 (CC)	0	Labyrinthotomy	0	1	Complete
Tucci et al (1995)	6(1 ad)	0	CC (1) and CH (2)	1 (CH)	1 (CC) (access)	Canal wall down	3 (1 IP, 1 CH, 1 CC)	4 (1 CC 1 CH)	At least 10 electrodes 2 CH into IAC
Luntz et al (1997)	10	3°	CC (3)	2 (CC), *(1)	2 (rec meningitis obliteration)	Labyrinthotomy (2)	5 (2 NC)	3	2 CC/7 IP complete CC incomplete
McElveen et al (1997)	4	0	CC (4)	2 (CC)	0	Labyrinthotomy (4)	0	1 (at high current levels)	Complete
Weber et al (1998)	12	0	CC (2) and CH (2)	2	2 (rec meningitis, obliteration)	Facial recess	6	2	-
Woolley et al (1998)	4	1 (EVA)	CC (1)	0	0	Facial recess	3 (1 CC*, 1 EVA, 1 IP)	N IX stimulation	2 IP complete EVA/CC incomplete
Ito et al (1999)	1	0	CC (1)	1 (CC)	1 (access)	Labyrinthotomy	0	0	Complete
Beltrame et al (2000)	1	0	CC (1)	0	0	Labyrinthotomy	0	0	Complete
Eisenman et al (2001)	17	0	CC (4) and CH (2)	1	1 (access)	Facial recess	7 (5 IP, 2 CC)	0	-
Incesulu et al (2002)	2	0	CH (1)	0	0	-	2 (1 IP 1 CH)		Incomplete
Mylanus et al (2003)	13	2	CC (1)	1 (CC)	1 (rec meningitis, obliteration)	Labyrinthotomy	2 (IP)	1 (CC)	Complete

Abbreviations: ad adults, CC = common cavity, CH = cochlear hypoplasia, IP incomplete partition, EVA = enlarged vestibular aqueduct, IAC = internal auditory canal, ° 2 patients with normal cochlea and labyrinth on CT but peroperative CSF gushers, revision surgery after 7 months as a result of CSF leakage and meningitis, * facial nerve injury

Table 6. Summary of performance data obtained from previous published studies which include children with severely malformed cochleae

Author(s)	Cochlear Malformation	Age (yrs) at implantation	Follow-up (months)	Thresholds with CI	closed set speech perception	open set speech perception	Comments
Jackler et al. (1987)	CC CH CC CH	5(right ear)* 7(left ear)* 5 9	12 2 12 10	73 dB HL to NM 53 to 68 dB HL 62 to 69 dB HL 39 to 63 dB HL			All implants were single electrode. *patient became a non-user as a result of facial nerve stimulation. The other ear (left) was implanted 2 years later
Slattery and Luxford (1987)	CC CH CC	3.5 4.5 3	42° 42 2	55 to 60 dB HL 20 to 30 dB HL 55 to 80 dB HL	No Yes NT		°Single electrode implant.
Molter et al. (1993)	CC	4	10		39% MTS (W)		
Tucci et al. (1995)	CC CH CH	3.5 4 4	12 18 18	Range of all patients: 30 to 40 dB HL	<10% MTS(W) 70% <10%	35% GASP(W) 10%	Fluctuating thresholds: patients with abnormal cochleae may require frequent monitoring of psychophysical responses
McElveen et al. (1997)	CC	2, 4, 1.3 and 7	No follow up	No data	No data	No data	Audiometric data submitted for publication in subsequent paper
Luntz et al. (1997)	CC CC CC	3 3 4	87 9 8	20 dB HL SRT 25 dB HL SRT 15 dB HL SRT	Yes No No	56% PBK 0% 0%	Results in patients with inner ear malformations other than CC are comparable to those of other deaf children with CI
Weber et al. (1998)	CC CC CH CH	3.5 3.5 3.3 4.1	22 7 19 37		At 9 months - at 7 months at 8 months	At 15 months - - -	Slower rate of development compared to normal cochleae
Woolley et al. (1998)	CC	4.5	6	Detection within speech spectrum	40% ESP/L	0%	Revision surgery after 7 months as a result of CSF leakage and meningitis
Ito et al. (1999)	CC	4	3	No data	70% WI		
Beltrame et al. (2000)	CC	2	2	No data			Reaction to sounds and good detection and identification of sounds
Eisenman et al. (2001)	CC CC CC CC CH	7.7 3.8 2.1 4.8 8.5	24 24 24 24 6		21% ESP 8% 8% 100% 29%	0% GASP(W) 0% 0% 75% 17%	No statistical difference between malformations and controls. No statistical difference between mild and severe malformations (small numbers!). Slower rate of development.
Incesulu et al. (2002)	CH	5	10	35 to 40 dBA			Discrimination and proper reaction to sounds in this multihandicapped child.
Mylanus et al. (2003)	CC	6.2	24	40 to 50 dB HL		40% GN	Results in patients with inner ear malformations other than CC are comparable to those of other deaf children with CI

Abbreviations: CC = common cavity, CH = cochlear hypoplasia, SRT = speech reception threshold, IE = inner ear, NT = not tested, ESP = early speech perception test, ESP/L = low verbal version of ESP, GASP(W) = Glendonald auditory speech perception test for words, MTS(W) = monosyllable trochee spondee (word) identification test, WI = word identification 1-, 3- and 5-syllable words, PBK = Phonetically Balanced-Kindergarten test, GN = Gestel/Nijmegen open set phoneme test.

Patients with severe inner ear malformations are expected to perform less than patients with normal cochlea because of the likelihood of a decreased number of spiral ganglion cells associated with cochlear malformation and meningitis, and because the more complex surgical challenges in such malformed ears.[12] Research has shown that in patients with severe malformations, postoperative speech perception results are highly variable and less certain.[3,4] This is reflected in Tables 5 and 6, in which special attention was given to the outcome of implantation in severely malformed cochlea. Table 6 summarizes the follow-up period of the various children and their results in speech perception tests. In most children, the follow-up period was short. Thresholds with the CI vary enormously. Results after 24 months vary from no speech perception at all to 100% closed-set word identification and 75% open-set word identification. Most studies state that all children are users of their implant, benefit from it, and perform better than with their hearing aids. Perhaps one of the most interesting studies in this regard is the case-control study by Eisenman et al.[12] In their study, at 24 months after implantation, there were no significant differences in performance on standard measures of speech perception between children with radiographically malformed cochleae and those with normal cochleae, although the former group developed at a slower rate. Moreover, they could not find a significant difference in performance between children with mild malformations and severe malformations, although numbers seem too small for a definite conclusion. Weber et al.[10] noted that children with hypoplastic cochleae seemed to be progressing within the same range of ability as those with incomplete partition of the cochlea. In this study also, patient numbers and follow-up period were too small to allow comparison between the degree of malformation and performance with a CI. Knowing that implantation of a severe hypoplastic cochlea will often involve a partial insertion, even when a compressed electrode array is used, and implantation of a common cavity may involve a "functional" partial insertion, results may turn out to be comparable to those obtained in post-meningitic children with obliterated cochleae and partial insertions. It has been shown that these children develop speech perception skills at a slower rate and often do not reach the same outcome level as the children with complete insertions.[26] An unpublished report of more than 40 implanted children with cochlear malformations indeed showed poor results in children with severe cochlear hypoplasia and a great variability in the results in children with a common cavity deformity.[27] In only two published cases on results of common cavities some open set capabilities have been reported.[4,10]

In our patient group, as was to be expected, the children with a severe or mild incomplete partition and the child with the isolated EVA syndrome perform well. Three of the 6 children with a follow-up of more than 2 years are in mainstream schools and 1 child is in a school for the hard of hearing. Even the girl with the common cavity and recurrent

meningitis has adequate postoperative thresholds with her CI and has a 40% open-set phoneme perception 2 years after implantation. In her case, as a result of facial nerve stimulation, a limited number of electrodes had to be switched off. Facial nerve stimulation has been reported in other cases of cochlear implantation in a common cavity[20] and tends to have an overall higher incidence in patients with a malformed cochlea. This may be related to the facial nerve's aberrant course, dehiscence over the nerve, or its proximity to the electrode array. When electrodes have to be deactivated, the patient's performance may decrease. Despite the difficulties in measuring speech perception in children with limited language skills, all children showed gains in auditory awareness with the implant compared with preimplantation performance.

Conclusions

With congenital sensorineural hearing loss now a common cause of deafness in the pediatric cochlear implant patient pool, familiarity with unusual anatomic configurations will become increasingly more important.[9] Reports on results of cochlear implantation in this specific group of CI candidates are important. In this study, in which own results and reports from the literature were combined, the incidence of an aberrant course of the facial nerve in inner ear malformations was, after Hoffman[9], confirmed at 17%, which increases to 27% in severe cochlear malformations. The incidence of CSF gushers was 40%. Our own experience and the literature have shown that the surgical procedure is safe, provided the surgeon is aware of the fact that the facial nerve is more at risk than normal. Preoperative HRCT scanning and MRI and facial nerve monitoring are essential. Concomitant middle ear problems may lead to potentially hazardous situations, as was shown in one of our patients. Perioperative imaging should be considered when implanting a malformed cochlea to rule out insertion in the internal auditory meatus.

The performance of implanted children with severe cochlear malformations varies considerably. The majority of the children reviewed in the literature have results presented after a follow-up of less than 24 months. At this stage, there seems to be no indication that children with congenital anomalies of the inner ear will eventually have a lesser performance. Long-term follow-up studies of larger number of patients will offer the possibility to take other confounding factors into account, such as age at implantation and communication mode. Studies should provide detailed information concerning the anatomy of the inner ear. To obtain this knowledge is especially important so that it can be used when counseling parents before implantation.

References

1. Jackler RK, Luxford WM, House WF Congenital malformations of the inner ear a classification based on embryogenesis. *Laryngoscope* 1987;97·2-14
2. McClay JE, Tandy R, Grundfast K, Choi S, Vezina G, Zalzal G, Willner A. Major and minor temporal bone abnormalities in children with and without congenital sensorineural hearing loss. *Arch Otolaryngol Head Neck Surg* 2002;128:664-71.
3. Woolley AL, Jenison V, Stroer BS, Lusk RP, Bahadori RS, Wippold FJ. Cochlear implantation in children with inner ear malformations *Ann Otol Rhinol Laryngol* 1998;107:492-500.
4. Luntz M, Balkany T, Hodges AV, Telischi FF. Cochlear implants in children with congenital inner ear malformations. *Arch Otolaryngol Head Neck Surg* 1997,123·974-7
5. Tucci DL, Telian SA, Zimmerman-Phillips S, Zwolan TA, Kileny PR. Cochlear implantation in patients with cochlear malformations *Arch Otolaryngol Head Neck Surg* 1995;121.833-8.
6. Molter DW, Pate BR, Jr., McElveen JT, Jr. Cochlear implantation in the congenitally malformed ear. *Otolaryngol Head Neck Surg* 1993;108·174-7
7. Slattery WH, III, Luxford WM. Cochlear implantation in the congenital malformed cochlea. *Laryngoscope* 1995;105·1184-7.
8. McElveen JT, Jr., Carrasco VN, Miyamoto RT, Linthicum FH, Jr. Cochlear implantation in common cavity malformations using a transmastoid labyrinthotomy approach. *Laryngoscope* 1997;107·1032-6
9 Hoffman RA, Downey LL, Waltzman SB, Cohen NL. Cochlear implantation in children with cochlear malformations *Am J Otol* 1997;18:184-7
10. Weber BP, Dillo W, Dietrich B, Mancke I, Bertram B, Lenarz T. Pediatric cochlear implantation in cochlear malformations. *Am J Otol* 1998;19:747-53
11. Jackler RK, Luxford WM, House WF Sound detection with the cochlear implant in five ears of four children with congenital malformations of the cochlea. *Laryngoscope* 1987,97.15-7.
12. Eisenman DJ, Ashbaugh C, Zwolan TA, Arts HA, Telian SA. Implantation of the malformed cochlea. *Otol Neurotol* 2001;22:834-41
13 Graham JM, Phelps PD, Michaels L. Congenital malformations of the ear and cochlear implantation in children: review and temporal bone report of common cavity. *J Laryngol Otol* Suppl 2000;25·1-14.
14. Snik AF, Vermeulen AM, Geelen CP, Brokx JP, van den BP. Speech perception performance of children with a cochlear implant compared to that of children with conventional hearing aids. II. Results of prelingually deaf children. *Acta Otolaryngol* 1997;117:755-9.
15. Bendet E, Cerenko D, Linder TE, Fisch U. Cochlear implantation after subtotal petrosectomies. *Eur Arch Otorhinolaryngol* 1998,255 169-74.
16. Bamiou DE, Worth S, Phelps P, Sirimanna T, Rajput K. Eighth nerve aplasia and hypoplasia in cochlear implant candidates: the clinical perspective. *Otol Neurotol* 2001;22:492-6.
17. Casselman JW, Offeciers FE, Govaerts PJ, Kuhweide R, Geldof H, Somers T, D'Hont G. Aplasia and hypoplasia of the vestibulocochlear nerve diagnosis with MR imaging. *Radiology* 1997;202·773-81.
18. Sennaroglu L, Saatci I, Aralasmak A, Gursel B, Turan E Magnetic resonance imaging versus computed tomography in pre-operative evaluation of cochlear implant candidates with congenital hearing loss. *J Laryngol Otol* 2002;116.804-10.
19. Phelps PD. The common cavity deformity of the ear. A precursor of meningitis but now being implanted. *JBR -BTR* 1999;82:239-40
20. Beltrame MA, Bonfioli F, Frau GN. Cochlear implant in inner ear malformation: double posterior labyrinthotomy approach to common cavity. *Adv Otorhinolaryngol* 2000;57·113-9.

21 Aschendorff A, Marangos N, Laszig R. Large vestibular aqueduct syndrome and its implication for cochlear implant surgery. *Am J Otol* 1997,18:S57.
22. Incesulu A, Vural M, Erkam U, Kocaturk S. Cochlear implantation in children with inner ear malformations: report of two cases. *Int J Pediatr Otorhinolaryngol* 2002;65:171.
23. Au G, Gibson W. Cochlear implantation in children with large vestibular aqueduct syndrome *Am J Otol* 1999;20:183-6.
24. Miyamoto RT, Bichey BG, Wynne MK, Kirk KI. Cochlear implantation with large vestibular aqueduct syndrome *Laryngoscope* 2002,112:1178-82.
25. Bauer PW, Wippold II FJ, Goldin J, Lusk RP. Cochlear implantation in children with CHARGE association. *Arch Otolaryngol Head Neck Surg* 2002,128:1013-7
26 van den Borne S, Snik AFM, van den Broek P, Vermeulen AM. Performance of postmeningitic deaf children with a multichannel cochlear implant and partial insertion of the electrode array. Thesis 1999. University of Nijmegen.
27 Weber, BP, Grimaldi H, Götz F, Meyer V, Bertram B, Lenarz T. Cochlear implantation in cochleovestibular malformations: surgical and rehabilitation results in more than 40 children. Book of abstracts of the 6th European Symposium on Paediatric Cochlear Implantation, Las Palmas de Gran Canaria 2002:160.

5.1

Cochlear implantation in otosclerosis

Cochlear implantation in 53 patients with otosclerosis: demographics, CT scanning, surgery and complications

L.J.C. Rotteveel
D.W. Proops
R.T. Ramsden
S.R. Saeed
A.F. van Olphen
E.A.M. Mylanus

Otology & Neurotology 2004, 25(6) 943-952

Abstract

Objectives: to collect data of a large number of cochlear implant recipients with otosclerosis and to make an assessment of these patients' clinical characteristics, computed tomographic (CT) scans, surgical findings and complications, and to quantify the occurrence of postoperative facial nerve stimulation.

Patients: Fifty-three patients with otosclerosis from four cochlear implant centres in the United Kingdom and the Netherlands were reviewed. Sixty surgical procedures were performed in these patients: 57 devices were placed in 56 ears.

Results: The patients had varying rate of progression of hearing loss.The CT scans demonstrated retrofenestral (cochlear) otosclerotic lesions in the majority of the patients. Although not statistically significant, the extent of otosclerotic lesions on the CT scan as categorized in 3 types, tends to be greater in patients with rapidly progressive hearing loss, in patients in whom there is surgically problematic insertion of the electrode array and in patients with facial nerve stimulation. In four patients, revision surgery had to be performed. Twenty of 53 (38%) patients experienced facial nerve stimulation at various periods postoperatively.

Conclusions: Cochlear implant surgery in patients with otosclerosis can be challenging, with a relatively high number of partial insertions and misplacements of the electrode array demanding revision surgery. A very high proportion of patients experienced facial nerve stimulation mainly caused by the distal electrodes. This must be discussed with patients preoperatively.

Introduction

Cochlear implantation is a well established and cost-effective means of rehabilitating selected congenitally deaf individuals or those with acquired deafness [1] In adults with acquired deafness, the cause in some cases is otosclerosis Cochlear implantation in this particular group of patients may present the surgeon with specific challenges The rehabilitation team may have to deal with a difficult postoperative fitting as a result of partially inserted electrode arrays, a misplaced array or facial nerve stimulation (FNS) In order to acquire more insight into these matters, a multicentre study was undertaken Thus, a relatively large number of cochlear implant (CI) recipients with otosclerosis could be evaluated

The patient with otosclerosis typically presents with a history of slowly progressive hearing loss that is usually bilateral and asymmetrical Hearing loss in fenestral otosclerosis may be conductive (CHL) In addition there may be a progressive sensorineural hearing loss (SNHL) causing a mixed hearing loss pattern [2] In far-advanced otosclerosis or retrofenestral otosclerosis pure SNHL may exist [3] Otosclerosis occurs more frequently in Caucasians and usually presents between the ages of 15 to 45 years [4] There may be a family history of deafness There is rapid progression of the hearing loss in younger patients, during pregnancy and in women on oestrogen therapy The disease is equally common in both sexes However, in clinical practice the disease is seen more frequently in females which is possibly due to a combination of a higher incidence of bilateral and severe disease in females and exacerbation due to hormonal influences [4] Tinnitus and vestibular symptoms are also common features

There is more or less a consensus about the way otosclerosis is inherited autosomal dominant with incomplete penetrance and variable expression [5,6] Based on the assumption that otosclerosis is an inherited collagen disorder, otosclerosis has historically been associated with other connective tissue disorders like osteogenesis imperfecta [4,7] However, genetic and histopathological studies showed that otosclerosis is not a localised form of osteogenesis imperfecta [8,9] Otosclerosis only affects bone derived from the otic capsule In the active vascular phase (otospongiosis), the normal lamellar bone is resorbed and, as the disease progresses, replaced by thick, irregular bone (sclerotic phase), although this sequence has been questioned [10] Otosclerotic bone may invade the stapes footplate causing stapes ankylosis and CHL SNHL is possibly caused by lytic enzymes that are released from otosclerotic foci into the perilymph[11] or by narrowing of the cochlear lumen with distortion of the basilar membrane [12] Long-term follow-up studies suggest that about 10% of ears with otosclerosis and CHL also develop SNHL [13,14]

High-resolution computed tomography (HRCT) is at present the imaging modality of choice for the assessment of the osseous labyrinth, labyrinthine windows and cochlear capsule HRCT can detect abnormalities of the oval window area in 80 to 90% of patients

with surgically proven otosclerosis Sensitivity approaching 90% for fenestral otosclerosis has been demonstrated [15] One cannot conclude that otosclerosis is not present when demineralization is not present in HRCT, but one can be virtually certain that this disease is present when it is seen HRCT is therefor highly specific [16] The diagnosis of fenestral otosclerosis is usually made clinically, but HRCT evaluation can assess potential involvement of the cochlea Cochlear otosclerosis is the less common of the two forms and is rarely observed without fenestral involvement [17]

Resorbed bone on HRCT appears as areas of decreased density, lucent zones, which may give the impression that unusual canals and ducts exist, which may have surgical implications in CI surgery In fenestral otospongiotic lesions, the margin of the oval window may become decalcified, which makes the window look larger than normal, whereas mature otosclerotic foci narrow or even close the window In retrofenestral otosclerosis, a typical sign of otospongiosis is the 'double ring' or 'halo effect' The ring represents pericochlear confluent foci surrounding the cochlear lumen [18] Sclerotic foci cause abnormal irregularity and narrowing of the cochlear turns, best evaluated at the basal turn [19]

Management of patients with otosclerosis and severe or profound hearing loss may be stapedectomy or stapedotomy and subsequent hearing aid amplification Further, there have been some studies that demonstrated that sodium fluoride reduces the rate of SNHL [20 21] If treatment fails, the patient may become a candidate for CI surgery The changes in the temporal bone caused by otosclerosis may pose several challenges for the surgeon and for the rehabilitation team The surgeon may be confronted with an obliterated round window or basal turn Further, the cochlea may consist of soft, otospongiotic bone in which an electrode array that is pushed forward easily penetrates The speech processor programming might be hampered by the occurrence of facial nerve stimulation

The aim of this multicentre study was to collect data from a large number of CI recipients with otosclerosis and to make an assessment of these patients' clinical characteristics, CT scans, surgical findings and complications, and to quantify the occurrence of postoperative facial nerve stimulation (FNS)

Materials and Methods

Patient selection

The databases with prospectively collected data of 4 CI centres in the Netherlands and United Kingdom - University Hospital Birmingham, Manchester Royal Infirmary, Radboud University Nijmegen Medical Centre and University Medical Centre Utrecht - were searched for patients with otosclerosis Information regarding 61 patients were retrieved from the databases These patient's clinical notes, rehabilitation notes and CT

scans were fully reviewed at each implantation centre by the first and last authors. Included in the study were patients with either retrofenestral and/or fenestral otosclerotic lesions on CT scan, patients with normal CT scans but with a history of stapes surgery, and patients with otosclerosis diagnosed at the CI procedure. Eight patients did not meet the inclusion criteria and were excluded from this study. Thus, 53 patients were included: 19 patients (36%) had signs of otosclerosis on CT scan, 28 patients (53%) had a positive scan and had a history of stapes surgery and 5 patients (9%) had normal CT scans but a history of stapes surgery. One patient was diagnosed solely by the finding of a fixed stapes during the implantation procedure.

The year of implantation ranged from 1990 to 2002. Type and progression of hearing loss were assessed, as were notes on family history and complaints of tinnitus. All data concerning history including previous stapes surgery, implantation procedure(s) and postoperative follow-up were collected in a database. During the preoperative workup for CI, most patients had filled out a questionnaire in which they were asked to note the time of onset of hearing loss and the time at which their hearing loss became profound (when conventional hearing amplification was no longer effective). Although a subjective measure, these questionnaire data were used to calculate the duration of progressive hearing loss and duration of profound deafness (Figure 1).

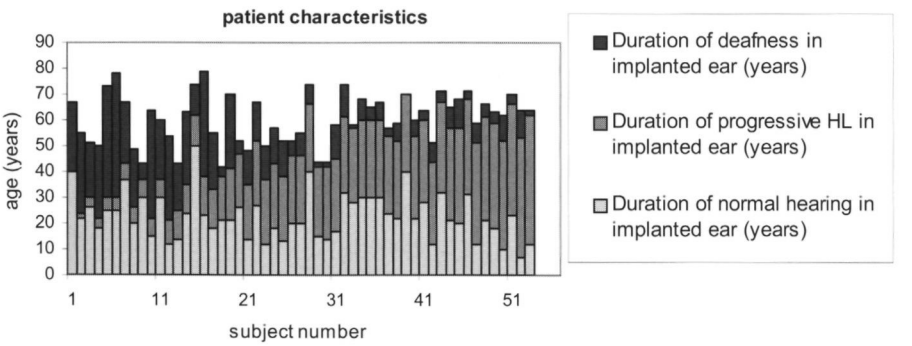


Figure 1. Patient characteristics

Imaging

As part of the standard evaluation for cochlear implantation of all CI centres, patients had undergone CT scanning of the temporal bone. When available, these CT scans were reviewed by the senior author. The scans of 17 patients had been destroyed. Their results were based on the official reports of the (neuro)radiologist at the CI centre at which the examination was performed. Fenestral involvement of the otosclerotic process (narrowed

or enlarged window, thickened footplate) and/ or retrofenestral involvement (double ring effect, narrowed basal turn) was noted After Lindsay's[22] histologically based subdivision of otosclerosis in fenestral and retrofenestral types, we propose a new categorization of findings on CT scan Type 1, solely fenestral lesions (spongiotic or sclerotic lesions), Type 2, retrofenestral lesions (double ring effect, narrowed basal turn, or both) with or without fenestral lesions, and Type 3, severe retrofenestral lesions with loss of the normal architecture of the cochlea To investigate the reproducibility of our categorization, an experienced neuroradiologist reviewed a subset of 18 CT scans independently and categorized the 36 ears according to the categories described There was good agreement between the two observers (κ = 0 77) Most disagreement (4 of 5 ears) concerned Types 2 and 3 The CT scan findings of the implanted ear at the time of primary implantation were used to investigate possible correlations with the progression of hearing loss, duration between onset of hearing loss and implantation, age at onset of hearing loss, sex, surgical problems and the occurrence of FNS

In all but one patient (implanted with an UCH mid Mk2 single-channel device), multichannel implant systems were used These comprised 6 Clarion devices (S, CI, CII, Enhanced Bipolar, HiFocus, Advanced Bionics, Sylmar, CA, U S A), 46 Nucleus devices (20+2, 22, 24, Contour, double array, Cochlear Corp , Lane Cove, Australia), and 4 Combi 40+ devices (Med-El, Innsbruck, Austria) Surgical problems and revision surgery are discussed in more detail Programming notes were searched for the occurrence of FNS and the causative electrodes

Results

In the total group of 53 patients with otosclerosis that eventually led to profound hearing loss, the onset of the hearing loss (of the eventually implanted ear) ranges from 7 to 50 years of age (median, 22 9 yrs, standard deviation (SD), 8 8 yrs)(Figure 1) Greater variation was found in the duration of progressive hearing loss to profound hearing loss or deafness, when the conventional hearing aid was no longer beneficial (range, 0-50 yrs, median, 26 yrs, SD, 13 yrs) On the whole, patients with a short duration of progressive hearing loss had a relatively long period of deafness before cochlear implantation and vice versa, which accounts for the fact that the actual age at implantation only shows a moderate variation (range, 42-79 yrs, median, 62 yrs, SD, 9 5 yrs) This finding probably results from the relatively late availability of cochlear implantation for this patient group

Hearing loss in the contralateral ear was also noted Seven patients had acquired bilateral profound hearing loss within 10 years, in 16 patients, one of both ears developed deafness at a similar rate, and in 30 patients the duration of progressive hearing loss was greater

than 10 years for both ears. No correlation was found between the rate of progression of hearing loss and the prevalence of stapes surgery (Table 1). Stapes surgery had been performed in 33 patients. The mean age at the time the first stapes surgery took place was 32 years (n = 30; range, 15-60 yrs; SD, 11.6 yrs). None of the ears deafened after stapes surgery (n = 5) has later undergone implantation. Patients with a more rapid progression of hearing loss did not have stapes surgery at an earlier age (Table 1). Tinnitus was absent in 15 patients, occasional but not bothersome in 27 patients, and definite in 10 patients, but again no correlation was found with the rate of progression of hearing loss (Table 1).

None of the CI centres systematically gathered information on family history. Retrospectively, records on family history of 15 patients (28%) could not be retrieved. Nineteen patients (19 out of 38; 50%) had a positive family history (i.e., family members with early-onset progressive hearing loss and/or a history of stapes surgery). A positive family history was not correlated with the duration of progressive hearing loss (Table 1).

Table 1 Correlation between the duration of progressive hearing loss of implanted ears and various factors

	A@OHL	A@OD	DoD	A@Imp	sex	stapes	A@1st stapes	tinnitus	Pos fam
ırman r	-0.19	0.79	-0.72	0 24	-0.12	0.04	0.15	0.03	0.09
lue	0.18	< 0.0001	< 0.0001	0.09	0.41	0.76	0.44	0.83	0.61
ı-tailed)									
ificant	no	yes	yes	no	no	no	no	no	no

Abbreviations A@OHL = age at onset of hearing loss; A@OD = age at onset of deafness; DoD = duration of deafness; A@Imp = age at implantation; stapes = history of previous stapes surgery in the later implanted ear; A@1st stapes = age at the time of first stapes surgery in any ear; Pos fam = positive family history

Table 2. Extent of otosclerosis in 106 ears on 53 preoperative CT scans of 53 patients. In 17 (16%) ears no signs of otosclerosis were detected.

Otosclerotic lesions of the otic capsule		No. of ears (%)
Type 1	Solely fenestral involvement (thickened footplate and/or narrowed or enlarged windows)	7 (7%)
Type 2	Retrofenestral with or without fenestral involvement	55 (52%)
	Type 2a: double ring effect	26 (25%)
	Type 2b: narrowed basal turn	4 (4%)
	Type 2c: double ring and narrowed basal turn	25 (23%)
Type 3	Severe retrofenestral (unrecognizable otic capsule), with or without fenestral involvement	27 (25%)

Imaging

The results of the CT scans of the 53 patients are shown in Table 2. Of all 106 scanned ears 17 (16%) were unaffected; in 7 (7%), only fenestral otosclerosis was present (Type 1)(Figure 2a); and 55 (52%) had retrofenestral lesions with or without fenestral involvement (Type 2)(Figures 2b and 2c). Fenestral involvement was present in 34 ears (32%); in 21 (20%) it was not. In 27 (25%) ears, the normal structure of the otic capsule was almost unrecognizable because of extensive otosclerosis (Type 3)(Figure 2d). In such severe cases, assessing fenestral involvement was found very difficult.

In 11 patients (20%), the severity of otosclerosis was asymmetrical: 3 patients had one ear without signs of otosclerosis, whereas the remaining patients had one side with solely retrofenestral otosclerosis and the other side with retrofenestral and fenestral otosclerosis. Seven of these patients were implanted in the less affected ear. Still, the severity of otosclerotic lesions on CT scan of the implanted ears as categorized in 3 types was not significantly different from the nonimplanted ears (Figure 3a).

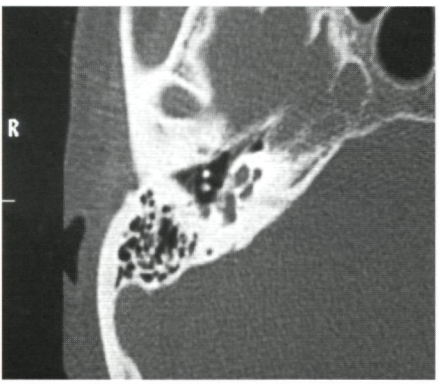


Figure 2a Anterofenestral focus and thickened footplate: Type 1 (*Case 35*)

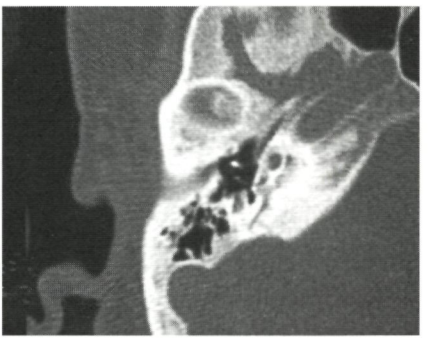


Figure 2c Double ring effect and a narrowed basal turn: Type 2c (*Case 29*)

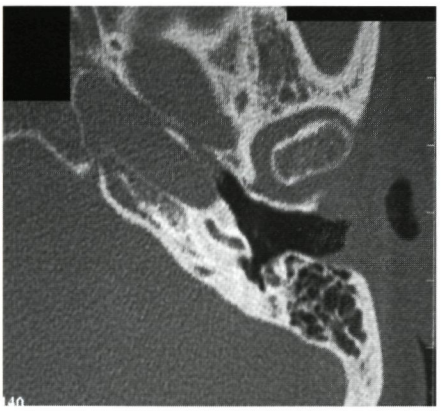


Figure 2b. Double ring effect or halo effect (hypodensity around the basal turn), no narrowing of the basal turn: Type 2a (*Case 41*)

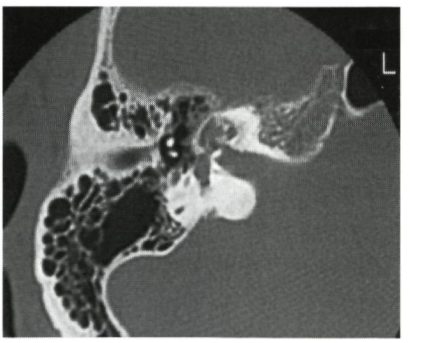


Figure 2d. Severe retrofenestral involvement of otosclerosis, no otic capsule recognizable: Type 3 (*Case 2*)

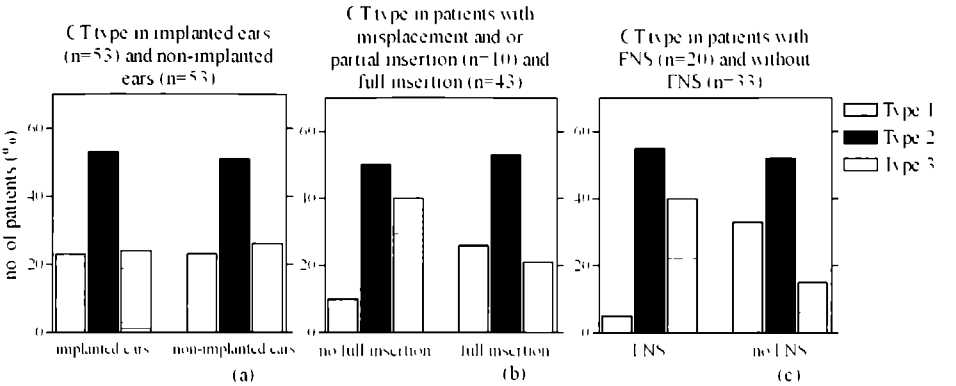


Figure 3. Comparisons of CT types in different groups

In 20 out of the 53 patients, the type of progressive hearing loss had been mixed; fenestral otosclerosis was detected on CT scan in 15 of these patients. In 14 patients with pure sensorineural progressive hearing loss, 8 patients had fenestral otosclerosis. In the remaining 19 patients the type of hearing loss in the progressive phase was unknown.
The extent of otosclerosis on CT scan (Types 1, 2 and 3) was significantly correlated with the age at onset of hearing loss and the age at onset of deafness, but not with the duration of progressive hearing loss, the duration of deafness, the total duration of hearing loss (time between onset of hearing loss and time of implantation), the age at implantation or sex (Table 3).

Table 3 Correlation between the extent of otosclerosis on CT scan of implanted ears and various factors

	A@OHL	A@OD	DoHL	DoD	TotalDoHL	A@Imp	sex	stapes
Spearman *r*	-0.30	-0 34	-0 12	0.20	0 08	-0 26	-0 08	-0.07
P value (two-tailed)	0.03	0 01	0 39	0.16	0 57	0.06	0 58	0.60
Significant	yes	yes	no	no	no	no	no	no

Abbreviations. A@OHL = age at onset of HL; A@OD = age at onset of deafness, DoHL = duration of progressive HL, DoD = duration of deafness; TotalDoHL = total duration of hearing loss, time between onset of hearing loss and time of implantation, A@Imp = age at implantation, stapes = history of previous stapes surgery in the later implanted ear

Surgery

Fifty-three patients underwent cochlear implantation. In 5 patients, a subsequent surgical procedure was undertaken of which one patient was included in a bilateral implantation programme. In 4 patients, revision surgery was necessary, involving the contralateral ear in 2 patients (Table 4). Thus, in 53 patients, 57 devices were implanted in 56 ears and 1

patient was eventually explanted. One of the revision cases involved a patient (*Case 33*) implanted with a single channel device who was later implanted in the contralateral ear with a multichannel device resulting in a partial insertion of the electrode array. In a second revision case (*Case 11*), primary implantation involved a partial insertion of the electrode array. Because of limited benefit of the implant, a double array cochlear implant was implanted at the contralateral ear 4 years later. The other 2 revision cases will be discussed in more detail below.

Table 4. Partial insertions, misplacements of electrode arrays and revision surgeries in 10 patients.

Patient no.	CT type	Primary implantation	First revision	Second revision	Third revision
10	3	partial, 16 e			
11	2	partial, 13 e	double array (CL ear)		
13	2	misplacement; lateral semicircular canal	partial, 4 e	new device, partial, 4 e	explantation
14	2	partial, 14 e, scala vestibuli			
19	2	partial, 18 e			
21	3	partial, 19 e			
33	3	single channel device	partial, 10 e (CL ear)		
37	1	misplacement; superior semicircular canal	withdrawal, complete insertion		
46	2	partial, 10 e			
47	2	misplacement, otosclerotic cavity			

partial = partial insertion of electrode array; e = electrodes; CL = contralateral.

Review of all the surgical notes of the implantation procedures demonstrated no abnormalities at inspection of the middle ear in 28 operations. Round window ossification was noted in 4 cases, stapes fixation in 5, the presence of a stapes prosthesis in 13, an eroded incus in 2, surgically removed ossicles in 3, middle ear adhesions in 2, mobile stapes in 2, tympanosclerosis in 1, and an oval window fistula was described in one surgical note. Although after the cochleostomy a full insertion of the electrode array could be achieved in the scala tympani in 42 patients, in one patient the scala tympani turned out to be obliterated and a full scala vestibuli insertion was performed (*Case 25*).

The insertion of a multichannel electrode array was problematic in 10 (19%) patients and resulted in a partial insertion of the electrode array in 7 cochleae and a misplacement of the electrode array in 3 cochleae (Table 4) The number of active electrodes in the cochleae ranged from 4 to 19 The misplacement of the electrode array in one patient (*Case 47*) has been described earlier in a case report by Ramsden et al [23] In all but two of the patients (*Cases 13 and 37*) with a partial insertion or misplacement, the presence of basal turn obstruction or narrowing could be identified in the preoperative CT scan A total of 27 patients (51%) had a narrowed or obstructed basal turn on CT scan, of whom 8 (30%) patients had a partial insertion or misplacement (Figure 4) After cochleostomy, the surgeon observed an obstructed scala in 17 patients and a patent scala in 36 patients Insertion of the electrode array in the latter group nevertheless led to one partial insertion and two misplacements In Figure 3b, the CT scans of patients with partial insertion and/or misplacement are compared with the CT scans of patients with full insertion Cochlear abnormality (Types 2 and 3) seems more extensive in the group with partial insertion and/or misplacement However, Chi square tests do not show any significant differences in the prevalence of Type 1, 2 or 3 between both groups Few other complications occurred during surgery One Clarion positioner was partially inserted, the postoperative CT scan showed a bent-over tip of the electrode array In one patient (*Case 7*) an oval window fistula was noted and closed with muscle Postoperative complications were not seen

Revision Case descriptions

Case 11

In this male patient progressive hearing loss first became apparent at the age of 30 years By the time he was 37 years old he was deaf ADS He underwent stapedectomy AD at the age of 42 years Twenty-three years after deafening he was referred for cochlear implantation He had a 0% speech perception score CT scanning showed a thickened footplate, the double ring effect and a narrowed basal turn on both sides (Type 2c) A Nucleus 22 device was implanted in his left ear The cochleostomy revealed a basal turn filled with easily bleeding fibrous tissue and bone, which required drilling Thirteen electrodes were placed with great difficulty A postoperative X-ray showed a straight course of the electrode array Four years later, a new CT scan showed, besides the presence of a CI in the left ear, progression of the otosclerosis (Type 3) A double array device was implanted in his right ear, 6 electrodes of the upper array and 11 electrodes of the lower array could be inserted Unfortunately, postoperative stimulation of the electrodes of the upper array did not lead to any auditory sensation and consequently they were switched off

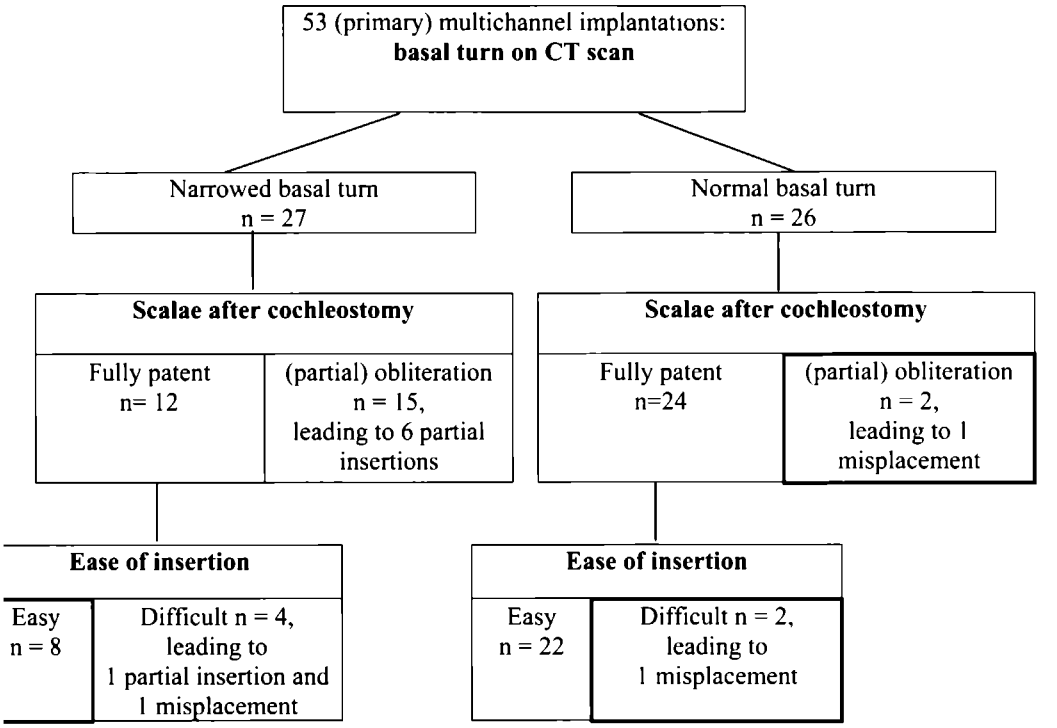


Figure 4. The presence or absence of a narrowed basal turn on CT scan, confirmed at surgery by observation after cochleostomy and ease of electrode array insertion.
The *boxes with bold lines* represent the patients with false-negative scan results (n = 4), the *box with double lines* represents the patients with false positive scan results (n = 8).

Case 13

This woman had progressive hearing loss since the relative young age of 14 years. Her father had undergone stapedectomy in the past. During pregnancy, her hearing had rapidly deteriorated. She was profoundly deaf at the age of 25 years. At the age of 19, a Teflon prosthesis had been placed in her right ear, and during the implantation assessment, a stapedectomy of her left ear took place. She had occasional tinnitus in her right ear. Electronystagmography showed areflexia in the right ear and hyporeflexia in the left ear. Because her hearing did not improve after the stapedectomy, 2 years later, the left ear was scheduled for an implantation with a Med-El Combi 40. Preoperative CT scan showed fenestral otosclerosis of the left ear and otic capsules heavily affected by otospongiosis showing abundant pericochlear lucencies. However, the basal turns did not appear narrowed or obstructed (CT scan Type 2a). During implantation, it was impossible to identify the round window because of round window obliteration. The promontory was

highly vasculated and the stapes piston was encountered Cochleostomy was performed using only the oval window as orientation a fully patent space emerged and the electrode array could totally be inserted without any difficulty Postoperative X-ray showed that the electrode array was not placed in the cochlea but in the horizontal semicircular canal Thirteen days later, the patient underwent reoperation the array was taken out and the cochleostomy was widened by drilling A heavily obstructed basal turn emerged No natural lumen could be reached by drilling, and four electrodes were placed in the drilled canal Perioperatively performed X-ray showed a well-positioned, but partially inserted electrode array However, during follow-up, lack of auditory sensation and short circuits made programming impossible In less than a year after implantation, the device was explanted and replaced by a new one of the same model Again, some drilling was required and only four electrodes could be inserted This implant also provided her for just a short period of time with minimal auditory sensation awareness of sound but no speech perception She developed various physical and mental complaints After 2 5 years the device was explanted

Case 37

Progressive mixed hearing loss in both ears first became apparent in this woman by the age of 24 years A sudden drop in hearing left her profoundly deaf in her left ear by the age of 52, and in her right ear by the age of 54 years She had severe tinnitus, no vertigo and a negative family history for otosclerosis She had never had a stapedectomy When referred for cochlear implantation at the age of 57 years, she had no residual hearing, no speech perception, and a positive result on promontory stimulation test The report of the preoperative CT scan (which could not be reviewed) did not mention the presence of any abnormalities During the implantation procedure, no abnormalities were encountered in the middle ear After cochleostomy, a fully patent basal turn emerged However, the insertion of the electrode array of a Nucleus 22M device into the cochlea took place with great difficulty Postoperatively, she had severe dizziness, nystagmus and pain in her mouth and throat A postoperative X-ray revealed that the array had gone straight up into the superior semicircular canal, presumably through otosclerotic bone Programming was troublesome because of discomfort, all but three electrodes had to be switched off Five months later, she was operated again and, after the device was pulled back, it was fully placed in the cochlea On the postoperative X-ray, the electrode array made a wide circle of 180 degrees, which appeared like a partial insertion During surgery however, there had been no electrodes visible outside the cochleostomy in the middle ear Fewer electrodes caused discomfort gradually more electrodes could be activated, up to 16 active electrodes The tinnitus had diminished compared to prior to surgery, and became less once the implant was activated

Facial nerve stimulation

During rehabilitation (mean follow-up, 5.6 yrs; range, 0.5-13 yrs), 20 patients (38%) developed FNS when the implant was activated at various time intervals. Two of these patients underwent implantation with a Clarion device, of which one was placed with a positioned; the remaining patients all had a Nucleus device (of which none had received a Contour device). Of all 53 patients, 10 were implanted with perimodiolar or 'modiolus hugging' devices (Clarion implants with positioner and Nucleus Contour implants). Only one of these patients (10%) developed FNS, whereas 19 of the remaining 43 patients implanted with nonmodiolus hugging devices (44%) suffered from FNS. This difference did not reach significance ($\chi^2 = 4.04$; P = 0.05; df = 1), because of the small numbers.

Five patients that suffered from FNS had partial insertions of the electrode array. The electrodes causing FNS in the patients with full insertion of the active electrodes of a Nucleus electrode array are shown in Figure 5. When comparing the CT scans of patients with FNS to those of patients without FNS (Figure 3c), Type 1 otosclerosis is significantly less frequent in patients with FNS (Fisher's exact test, P = 0.02). Although Type 3 seems more frequent in patients with FNS, this difference is not significant (Fisher's exact test, P = 0.05). The management of FNS usually consisted of a reduction in stimulus levels or eventually a switch off of the causative electrode, if only temporarily. In one of the CI centres an attempt was made to treat 2 of 5 patients with FNS with fluoride. In one of these patients (*Case 3*), FNS occurred already during surgery. During rehabilitation, more and more electrodes had to be switched off because of FNS until only 8 electrodes remained active. In the 2 years of fluoride treatment, the FNS remained stable. The other patient stopped using fluoride because the side effects of the treatment.

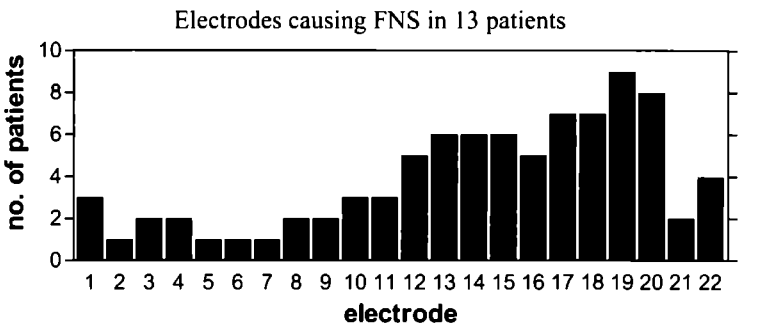


Figure 5. Electrodes causing facial nerve stimulation (FNS) in 13 patients with a multichannel Nucleus device with a complete insertion of the electrode array

Discussion

By conducting a multicentre study on cochlear implantation in patients with otosclerosis, it has been possible to collect data on a relatively large group of patients Off all adults who received a CI at the 4 implant centres up to the end of 2002 (n = 788), 6 7% has otosclerosis as the cause of deafness Thus, otosclerosis is not a rare indication for cochlear implantation, although far-advanced otosclerosis was once considered a contra indication for cochlear implantation [3]

In these patients deafened by otosclerosis, hearing loss became apparent in their early twenties The hearing loss was either mixed or pure sensorineural, and the rate of progression varied greatly In general, clinical otosclerosis is more frequent in woman and is seen most often between the ages of 30 and 49 years [16] In this group of otosclerosis patients who eventually received a CI, 38% were women, and the mean age at onset of hearing loss was 23 6 years in the implanted ears and 26 3 years in the nonimplanted ears Thus, a female dominance in this selected group of otosclerosis patients is no longer present In accordance to the eight otosclerosis patients receiving CIs in the study by Ruckenstein et al [17] the majority of patients were older adult men with a long history of progressive hearing loss Three of these eight patients (38%) had a family history of otosclerosis It has been shown that about 70% of patients inherit the disease (autosomal dominant) [16] In the current study, 50% of the patients had a positive family history Possibly, when data on family history are systematically collected, such as is performed for genetic research, higher incidences will be found

CT imaging

Although the earliest implantations date back to 1990, it was possible to review two-thirds of the CT scans and collect the original radiology reports of the remaining patients In this study, in 20% of patients the severity of otosclerosis was asymmetric In 77% of the patients, retrofenestral involvement was present on CT scan Isolated cochlear otosclerosis was present in at least 20% of the patients In severe cochlear otosclerosis, distinguishing whether or not fenestral involvement was also present is very difficult

In 7% of the patients, only fenestral involvement could be identified It is possible that the retrofenestral otosclerotic lesions were too small to be detectable on CT (i e , < 1 mm)[15] or that the chemical process in otosclerotic lesions damaged neural elements in the cochlea [11] Patients may have SNHL before otosclerosis becomes apparent on CT images and bilateral hearing loss may be present with unilateral or asymmetric CT findings [24] Such cases are also speculated to be the result of ototoxic enzymes reaching the endosteal cochlear capsule through small bony channels, leading to spiral ligament hyalinization and stria vascularis atrophy [25]

A comprehensive staging of CT images of the otic capsule in otosclerosis was suggested

We defined 3 categories of increasing local involvement of the otic capsule. Valvassori[18] characterized the otosclerotic lesions by hypodensity of the otic capsule or footplate thickening into 4 categories: anterior (fenestral) focus (1), pericochlear focus without (2) or with (3) endosteal extension, and footplate thickening (4). Rather than local extension, Valvasorri's classification is based on maturation of the lesions. However, a narrowed basal turn and severe pericochlear involvement seems to have more predictive value for the insertion procedure in cochlear implantation.

Although not significant, the severity of the cochlear lesions and postoperative FNS seem related. In this study, a higher extension of otosclerosis on CT scan was associated with a younger age at onset of hearing loss and onset of deafness. This finding is in agreement with earlier histopathologic findings in otosclerosis in which the type of otosclerosis involving the otic capsule, as opposed to the type of otosclerosis limited to the fenestra, is more active, with multiple foci that form early in life.[22] The duration of progressive hearing loss was not correlated. Patients deafened very quickly after the first signs of hearing loss, did not have more extensive lesions on CT scan than patients that had a long period of slowly progressing hearing loss. Also, patients with long duration of profound deafness did not have more extensive lesions on CT scan.

Surgery

Fayad et al.[26] stated that new bone formation in otosclerotic bones is limited to the scala tympani and is not a contraindication for implantation. In their study, ossification was present in 6 of 20 otosclerosis patients, and drilling up to 5 mm was required. They found insertion of the electrode array difficult in only one patient. Overall, ossification did not preclude CI surgery and did not influence the clinical performance. In the study by Ruckenstein et al.[17] in 1 of 8 otosclerosis patients a drill-out of the basal turn had to be performed. No partial insertions occurred. Despite ossified scalae tympani in two patients, full insertions were achieved by insertion of the scala vestibuli. In the current study, in 17 (32%) patients, a partial or complete obliteration of the basal turn was observed during surgery that required drilling forward the scala tympani. The insertion of the electrode array led to either partial insertion or misplacement in 10 (19%) patients, which is at least comparable to the occurrence of partial insertions in children deafened by meningitis.[27] Obstruction of the scalae may not be evident on CT scan, especially at the more apical turns. Regarding narrowing of the basal turn, the CT scan does have predictive value, as 37% of the cases with this particular finding proved to be surgically difficult, resulting in partial insertion or misplacement of the electrode array. MRI has been useful in the assessment of the membranous labyrinth with it's neural elements and of the cochlear lumen before to cochlear implantation, but has had limited application in the diagnosis of otosclerosis with involvement of the cochlear capsules because it does not image bone.[18]

Revision surgery has been necessary in 4 patients of which 1 patient unfortunately had 3 operations after the primary implantation. Two of the 3 patients with a misplaced electrode array underwent revision. Some feel that an attempt should be made at mapping the electrodes inside the cochlea and that any electrodes outside the cochlea, whether in the middle ear or in a cavity in the temporal bone, should be turned of, rather than to remove and attempt to replace the electrode in a second surgical procedure.[23] In such a case, proximity to the internal carotid artery or the meninges has to be assessed for potential danger. In general, it can be stated that implantation in patients with otosclerosis is surgically feasible but may be more demanding and revision surgery may be required more frequently than in the general CI population.

Facial nerve stimulation

Facial nerve stimulation (FNS) after cochlear implantation has been reported with a variable incidence. FNS may be a serious problem even leading to explantation.[28] The incidence of FNS in the general CI population as reported in the literature varies from 0.9%[22], 3% [29] to 14.6%[30] and is more frequent in patients with otosclerosis and otosyphilis.[17,29 31-33] A large percentage of patients (38%) in this study experienced FNS at various periods postoperatively.

When a CI is activated, electrical fields are generated that produce regional current flow. The distribution of these currents may be influenced in such a way that the facial nerve becomes stimulated. Because stimulus intensities needed for thresholds are not higher in otosclerotic bones, FNS must result either from lowering of the electrical impedance of the bone by the disease or by a reduced distance from the electrode to the facial nerve by loss of bone and cavity formation.[23] Both mechanisms are probable in otosclerosis.[33-35] Sometimes, electrodes positioned at the round window are responsible for FNS at the tympanic segment or even vertical segment of the facial nerve.[30] This may be the result of the proximity of the electrodes to the facial nerve, as the array crosses the facial ridge at the posterior tympanotomy. A low-impedance shunt at the basal cochlea has also been suggested. Most frequent FNS, however, has been reported to be caused by electrodes deeper in the cochlea, especially those electrodes positioned at the most superior part of the basal turn, which is closest to the geniculate and labyrinthine segment of the facial nerve [29] Bigelow et al.[33] demonstrated in a temporal bone study using the Nucleus 22 electrode array that the electrodes 8 to13 were closest to the labyrinthine portion. Indeed, in their 7 patients with FNS, the electrodes causing stimulation most frequently (in more than 2 out of 7 (> 29%) patients) ranged from electrodes 9 to 14. In our study however, the electrodes most frequently involved in FNS (in more than 4 out of 13 (> 31%) patients) ranged from electrodes 12 to 20 (i.e., the more distal electrodes on the array). The difference is explained by the fact that in the current study, calculations were based on the number of

inserted electrodes plus a variable number of 0 to 10 supporting rings measured from the cochleostomy, whereas in the study by Bigelow et al.[33] in all cases, all 10 supporting rings were included in the number of inserted electrodes, resulting in an overall deeper insertion. Small variations in cochlear length or bending of the array may give further rise to variations in the exact position of an electrode, but given the data above, it may be concluded that the facial nerve in most patients with FNS in this study was stimulated by electrodes positioned closest to the labyrinthine and geniculate segments.

Electrode position within the cochlea is considered to be an important variable in cochlear implant outcomes measures.[36,37] To improve stimulation of specific neuronal populations and to decrease power consumption, perimodiolar or modiolus hugging electrode arrays were developed. Indeed, electrical threshold levels tended toward lower values in the patients with tighter coils, although the number was not significant.[36] It can be hypothesised that when lower thresholds are needed in an otosclerotic bone, less leakage of current possibly resulting in FNS will occur. Battmer et al.[38] reimplanted 3 patients who had previously been implanted with a Nucleus Mini22 device and who were suffering from severe FNS with a Nucleus 24 Contour device. In all 3 patients, after reimplantation, none of the postoperatively stimulated electrodes caused FNS. In the present patient group, a trend was observed towards fewer occurrences of FNS in patients implanted with modiolus hugging devices.

As this study has demonstrated, FNS in implanted patients with otosclerosis is common and occurred with Nucleus as well as Clarion devices. Most often, FNS is successfully managed by reprogramming the responsible electrodes, but this may limit the efficacy of the implant. The preoperative CT scans in patients with FNS more frequently showed more extensive abnormality than in patients without FNS, although the difference was not statistically significant. In the preoperative counseling of a patient the CT scan is possibly helpful in predicting FNS and may be decisive in determining the side of implantation.

Conclusions

To our knowledge, this multicentre study describes the largest number of otosclerosis patients provided with CIs to date. Within the patient group, the rate of progression of hearing loss had varied greatly. The CT imaging demonstrated retrofenestral otosclerotic lesions in the majority of the patients. The severity of the pathologic changes of the otic capsule related to early onset of hearing loss and deafnes. Although not statistically significant, the extent of otosclerotic lesions on the CT scan tends to be greater in patients with rapidly progressive hearing loss, surgically problematic insertion of the electrode array and facial nerve stimulation. CI surgery in patients with otosclerosis can be challenging, with a relatively high number of partial insertions and misplacements of the

electrode array demanding revision surgery. A very high percentage of patients was confronted with FNS mainly caused by the distal electrodes, which must be discussed with patients preoperatively.

References

1. Summerfield AQ, Marshall DH, Davis AC. Cochlear implantation: demand, costs, and utility *Ann Otol Rhinol Laryngol Suppl* 1995;166:245-8.
2. Browning GG, Gatehouse S Sensorineural hearing loss in stapedial otosclerosis. *Ann Otol Rhinol Laryngol* 1984;93:13-6.
3. Wiet RJ, Morgenstein SA, Zwolan TA, Pircon SM. Far-advanced otosclerosis. Cochlear implantation vs stapedectomy. *Arch Otolaryngol Head Neck Surg* 1987,113:299-302.
4. Smyth GDL. Otosclerosis In AG Kerr, editor. Scott-Brown's Otolaryngology Oxford: Butterworth Heinemann; 1997. p. 3-14/1-3/14/35.
5. Menger DJ, Tange RA. The aetiology of otosclerosis: a review of the literature. *Clin Otolaryngol* 2003;28·112-20.
6. Chen W, Campbell CA, Green GE, van den Bogaert K, Komodikis C, Manolidis LS, Aconomou E, Kyamides Y, Christodoulou K, Faghel C, Giguere CM, Alford RL, Manolidis S, Van Camp G, Smith RJ Linkage of otosclerosis to a third locus (OTSC3) on human chromosome 6p21 3-22 3. *J Med Genet* 2002,39.473-7.
7. Nager GT. Osteogenesis imperfecta of the temporal bone and its relation to otosclerosis. 1988;97·585-93.
8. Pedersen U. Osteogenesis imperfecta clinical features, hearing loss and stapedectomy. Biochemical, osteodensitometric, corneometric and histological aspects in comparison with otosclerosis *Acta Otolaryngol Suppl* 1985,415.1-36.
9. Morrison AW, Bundey SE. The inheritance of otosclerosis. *J Laryngol Otol* 1970;84:921-32.
10. Guneri EA, Ada E, Ceryan K, Guneri A. High-resolution computed tomographic evaluation of the cochlear capsule in otosclerosis: relationship between densitometry and sensorineural hearing loss. *Ann Otol Rhinol Laryngol* 1996;105:659-64
11. Linthicum FH, Jr. Histopathology of otosclerosis. *Otolaryngol Clin North Am* 1993;26.335-52.
12. Linthicum FH, Jr., Filipo R, Brody S. Sensorineural hearing loss due to cochlear otospongiosis: theoretical considerations of etiology. *Ann Otol Rhinol Laryngol* 1975;84:544-51.
13 Ramsay HA, Linthicum FH Jr. Mixed hearing loss in otosclerosis: indication for long-term follow-up *Am J Otol* 1994,15 536-9.
14 Browning GG, Gatehouse S. Sensorineural hearing loss in stapedial otosclerosis. *Ann Otol Rhinol Laryngol* 1984,93.13-6.
15. Mafee MF, Henrikson GC, Deitch RL, Norouzi P, Kumar A, Kriz R, Valvassori GE. Use of CT in stapedial otosclerosis. *Radiology* 1985,156.709-14.
16. Donaldson J, Snyder JM. Otosclerosis. In. Cummings CW, Fredrickson JM, Harker LA, Krause CJ, Schuller DE, editors. Otolaryngology-head and neck surgery. 2nd ed. St Louis: Mosby; 1993. p. 2997-3016
17. Ruckenstein MJ, Rafter KO, Montes M, Bigelow DC. Management of far-advanced otosclerosis in the era of cochlear implantation. *Otol Neurotol* 2001;22.471-4.
18. Valvassori GE. Imaging of otosclerosis. *Otolaryngol Clin North Am* 1993;26:359-71.
19. Swartz JD, Mandell DW, Berman SE, Wolfson RJ, Marlowe FI, Popky GL. Cochlear otosclerosis (otospongiosis): CT analysis with audiometric correlation *Radiology* 1985;155 147-50.

20. Bretlau P, Causse J, Causse JB, Hansen HJ, Johnsen NJ, Salomon G. Otospongiosis and sodium fluoride. A blind experimental and clinical evaluation of the effect of sodium fluoride treatment in patients with otospongiosis *Ann Otol Rhinol Laryngol* 1985,94:103-7
21. Causse JR, Causse JB, Uriel J, Berges J, Shambaugh GE, Jr, Bretlau P. Sodium fluoride therapy *Am J Otol* 1993,14:482-90.
22. Lindsay JR. Histopathology of otosclerosis. *Arch Otolaryngol* 1973;97:24-9.
23. Ramsden R, Bance M, Giles E, Mawman D. Cochlear implantation in otosclerosis: a unique positioning and programming problem. *J Laryngol Otol* 1997;111:262-5.
24. Weissman JL. Hearing loss. *Radiology* 1996;199 593-611
25. Parahy C, Linthicum FH, Jr. Otosclerosis: relationship of spiral ligament hyalinization to sensorineural hearing loss *Laryngoscope* 1983;93:717-20.
26. Fayad J, Moloy P, Linthicum FH, Jr Cochlear otosclerosis: does bone formation affect cochlear implant surgery? *Am J Otol* 1990,11.196-200.
27. Mylanus EA, van den Broek P. Clinical results in paediatric cochlear implantation. *Cochlear Implants International* 2003,4:137-47.
28. Shea JJ, III, Domico EH. Facial nerve stimulation after successful multichannel cochlear implantation. *Am J Otol* 1994,15:752-6.
29. Kelsall DC, Shallop JK, Brammeier TG, Prenger EC. Facial nerve stimulation after Nucleus 22-channel cochlear implantation. *Am J Otol* 1997;18:336-41
30. Niparko JK, Oviatt DL, Coker NJ, Sutton L, Waltzman SB, Cohen NL Facial nerve stimulation with cochlear implantation VA Cooperative Study Group on Cochlear Implantation. *Otolaryngol Head Neck Surg* 1991;104 826-30.
31. Muckle RP, Levine SC. Facial nerve stimulation produced by cochlear implants in patients with cochlear otosclerosis. *Am J Otol* 1994;15:394-8
32. Ross UH, Laszig R, Bornemann H, Ulrich C. Osteogenesis imperfecta clinical symptoms and update findings in computed tomography and tympano-cochlear scintigraphy *Acta Otolaryngol* 1993,113:620-4.
33. Bigelow DC, Kay DJ, Rafter KO, Montes M, Knox GW, Yousem DM. Facial nerve stimulation from cochlear implants. *Am J Otol* 1998;19:163-9
34. Mens LH, Oostendorp T, van den Broek P. Cochlear implant generated surface potentials: current spread and side effects *Ear Hear* 1994;15:339-45.
35. Weber BP, Lenarz T, Battmer RD, Hartrampf R, Dahm MC, Dietrich B. Otosclerosis and facial nerve stimulation. *Ann Otol Rhinol Laryngol Suppl* 1995;166 445-7
36. Marrinan MS, Roland JT Jr, Reitzen SD, Cohen LT, Cohen NL.. Degree of modiolar coiling, electrical thresholds, and speech perception after cochlear implantation. *Otol Neurotol* 2004;25:290-4.
37. Lenarz T, Battmer RD, Goldring JE, Neuburger J, Kuzma J, Reuter G. New electrode concepts (modiolus-hugging electrodes). *Adv Otorhinolaryngol* 2000,57:347-53.
38. Battmer RD, Pesch J, Goldring JE, et al Eliminating facial nerve stimulation by reimplantation of a Nucleus 24 Contour implant system. Conference Proceeding 2004;127

Chapter 5.2

Cochlear implantation in otosclerosis

Speech perception after cochlear implantation in 53 patients with otosclerosis: multicentre results

L.J.C. Rotteveel
A.F.M. Snik
H.R. Cooper
D.J. Mawman
A.F. van Olphen
E.A.M. Mylanus

(Submitted)

Summary

Objectives to analyse the speech perception performance of 53 cochlear implant recipients with otosclerosis and to evaluate which factors influenced patient performance in this group The factors included disease-related data, such as demographics, preoperative audiological characteristics, the results of CT scanning and device-related factors

Methods data were reviewed on 53 patients with otosclerosis from four cochlear implant centres in the United Kingdom and the Netherlands Comparison of demographics, pre-operative CT scans and audiological data revealed that the patients from the four different centres could be considered as one group Speech perception scores had been obtained with the English AB monosyllable tests and Dutch NVA monosyllable tests Based on the speech perception scores, the patients were classified as poor or good performers The characteristics of these subgroups were compared

Results There was wide variability in the speech perception results Similar patterns were seen in the phoneme scores and BKB sentence scores between the poor and good performers The two groups did not differ in age at onset of hearing loss, duration of hearing loss, progression, age at onset of deafness, or duration of deafness

Conclusions the clinical presentation of the otosclerosis (rapid or slow progression) did not influence speech perception Better performance was related to less severe signs of otosclerosis on CT scan, full insertion of the electrode array, little or no FNS and little or no need to switch off electrodes

Introduction

Nowadays, cochlear implantation is a well-accepted and effective intervention in patients with profound hearing loss. A large number of studies have shown that the majority of adults and children with a cochlear implant (CI) achieve word scores of more than 50% on speech perception tasks.[1-3] However, performance varies widely and there are still a number of users who do not reach this level of performance.

Several attempts have been made to explain this variance in order to predict the benefit of cochlear implantation.[4,5] Well-known factors related to open-set speech perception are age at onset of deafness, duration of deafness, residual hearing with extensive use of hearing aids before implantation and whether the deafness was progressive or sudden.[5-8] In addition, device-related factors, such as type of CI device, speech processing strategy and number of active electrodes, are of importance.[9] Analysis of speech perception across devices and patient cohorts suggested that about two thirds of the variance can be explained by the above mentioned variables. The remaining one third of the variance is due to other, less obvious factors, e.g. the etiology of deafness.[5,8-10] The relation between speech perception scores and the etiology of deafness was reported to be weak.[8] However, etiology might affect auditory performance indirectly via other factors, such as number and position of active electrodes (e.g. in congenital malformation of the cochlea, or basal turn ossification in meningitis), electrical properties of the temporal bone (e.g. decalcification in otosclerosis), ganglion cell survival or function and central neural survival or function (e.g. in meningitis). Unfortunately, it is not possible to study such effects in heterogeneous groups, but studies on subgroups of patients with the same aetiology might be of value to assess the importance of the disease-specific factors. This information will be useful for counseling purposes.

In order to draw firm conclusions, a sufficiently large number of patients must be available. In the present study, a retrospective multicentre design was employed to evaluate the effect of otosclerosis on cochlear implantation. Over the past years, an increasing number of patients who received a CI have been diagnosed with otosclerosis (7 to 9.5%).[11,12]

Otosclerosis is a heritable disease that affects the bony structure of the temporal bone. In the active phase, so-called osteospongiosis, the normal lamellar bone is resorbed and through a vascular stage is replaced by thick, irregular bone in the normal middle layer of the otic capsule.[13] The subsequent hearing loss can be conductive, which is most commonly caused by stapes fixation due to plaque formation around the oval window, or sensorineural in the case of cochlear involvement. Sensorineural hearing loss (SNHL) in otosclerosis is thought to be the result of narrowing of the cochlear lumen with distortion of the basilar membrane[14] or it is believed to be caused by lytic enzymes that are released

into the perilymph from otosclerotic foci [15 17] Long-term follow-up studies showed that about 10% of ears with otosclerosis and conductive hearing loss also developed SNHL [18 19]
In otosclerosis patients, there seems to be a trend towards fewer active electrodes and poorer scores on postoperative open-set sentences tests than in CI recipients with other causes of deafness [20] Histological studies have shown that otosclerosis has a relatively small effect on spiral ganglion cell survival compared to other causes of deafness [21] Thus the poorer scores in otosclerosis patients might be explained by the lower number of active electrodes, the altered bone properties in the otic capsule that may affect the current distributions produced by the electrodes and possibly the older average age at implantation, rather than be caused by diminished neural response

In this multicentre study, a group of 53 otosclerosis patients with a CI were reviewed at the CI centres in Manchester, Birmingham, Utrecht and Nijmegen Patient characteristics, CT scans, surgical findings and the incidence of facial nerve stimulation (FNS) have been described in a previous paper [11] First, a search was made for inter-clinic differences in factors that might affect auditory performance Second, longitudinal speech perception scores were analysed to establish relations between speech perception scores and several factors related directly or indirectly to otosclerosis

Materials and methods

Subjects
Patients diagnosed with otosclerosis were retrieved from the databases of 4 CI centres in the Netherlands and United Kingdom that hold prospective data University Hospital Birmingham, Manchester Royal Infirmary, Radboud University Nijmegen Medical Centre and University Medical Centre Utrecht The diagnosis of 'otosclerosis' was based on the presence of otosclerotic lesions on the preoperative CT scan, history of stapes surgery, or the finding of fixation of the stapes during the surgical implantation procedure A total of 53 patients were included 19 patients (36%) had signs of otosclerosis on the CT scan, 28 patients (53%) had a positive CT scan and a history of stapes surgery, 5 patients (9%) had a normal CT scan and a history of stapes surgery and one patient was diagnosed solely by the finding of a stapes fixation during the implantation procedure The year of implantation of the patients ranged from 1990 to 2002 There was no difference in the mean and median year of implantation between the 4 centres
Table 1 shows the device types that had been used at each of the centres No differences were found in the distribution of the previous generation (Nucleus 22 and Clarion I) and the more recent generation CI devices (Nucleus 24 and Clarion II) between the 4 centres (Kruskal-Wallis test, P = 0 52)

Table 1. Number and types of implanted devices, previous and recent generations, per CI centre

Device type	Nijmegen (n = 13)	Utrecht (n = 9)	Birmingham (n = 17)	Manchester (n = 14)
Previous devices	9	5	11	10
Nucleus 22	4	3	11	9
Clarion S	2	0	0	0
Clarion I	3	0	0	0
Med-el 40+	0	2	0	1
Recent devices	4	4	6	4
Nucleus 24	3	4	6	4
Clarion II	1	0	0	0

Over half of the patients had undergone stapes surgery prior to cochlear implantation. The proportion of patients with a history of stapes surgery in either ear was significantly higher in the patient group from Utrecht (100%) than in the patient groups from Birmingham (41%) and Manchester (57%); there were no differences in stapes surgery between the other groups (Fisher's exact test). The proportions of patients who had preoperative experience with a conventional hearing aid (CHA) at the time of implantation did not differ between the 4 centres (Kruskal-Wallis test, $P = 0.67$).

Preoperative evaluation data

As part of the selection procedure for cochlear implantation, the patients at all four CI centres had undergone CT scanning of the temporal bone. When available, these CT scans were reviewed by the same experienced otologist. It appeared that the CT scans of 17 patients had been destroyed. In these cases, the diagnoses were based on the original reports by the radiologists at the CI centres. The CT scans were reviewed for fenestral involvement (narrowed or enlarged window, thickened footplate) and retrofenestral involvement (double ring effect, narrowed basal turn) of the otosclerotic process and were categorized into three types (Table 2).

Postoperative evaluation data

First, at each CI centre, the patients' speech processor programming notes were evaluated to gather information on the need to lower stimulation levels or switch off electrodes to eliminate non-auditory effects, such as FNS and pain or stinging sensations in the middle ear or throat.

Table 2. Extent of otosclerosis on the preoperative CT scans. 3 types

Otosclerotic lesions of the otic capsule	
Type 1	Solely fenestral involvement (thickened footplate and/or narrowed or enlarged windows)
Type 2	Retrofenestral, with or without fenestral involvement Type 2a: double ring effect Type 2b: narrowed basal turn Type 2c: double ring and narrowed basal turn
Type 3	Severe retrofenestral (unrecognizable otic capsule) involvement, with or without fenestral involvement

Second, longitudinal speech perception scores were retrieved from the medical files. At all 4 clinics, speech perception measurements had been carried out in special sound-treated booths. The speech material was recorded on tape or CD and presented by a loudspeaker placed in front of the patient. Although speech perception measurements were part of the regular evaluation visits at all 4 centres, the time interval between measurements varied. The English CI centres had recorded data on the open-set Bamford-Kowal-Bench (BKB) sentences test[22] and/or phoneme scores on the open-set Arthur Boothroyd (AB) monosyllables test.[23] Phoneme scores had also been obtained by the two Dutch CI groups, using the open-set NVA monosyllables tests.[24] The AB and NVA are largely comparable; the two tests comprise a large number of lists that consist of 10 isophonemic balanced CVC (consonant-vowel-consonant) words. As the speech recognition-intensity curves obtained from subjects with normal hearing were fairly comparable[24,25] and the test scores had been obtained at a fixed level of 40 dB above the speech reception threshold (SRT) of controls with normal hearing, it was decided to pool these data for statistical analysis. This presentation level of 40 dB above SRT resembles about 65 dB SPL, the overall level of normal speech.

In this study, the data from the 4 CI centres were compared with respect to demographics and preoperative audiological characteristics, CT scan results and the types of implants (previous or recent generation) used. For further analysis, the patients were grouped into poor and good performers, based on the 25th percentile of the speech perception scores of a large reference group of postlingually deaf adult CI patients using the same device type. The reference group for the phoneme scores comprised 76 Dutch CI patients implanted between 1991 and 2001, the reference group for the BKB-sentences test scores comprised 100 English patients. The characteristics of the two subgroups of 'better' and 'poor' performers were compared.

Results

Inter-clinic differences

Demographic data

Some demographic data from the patients at the CI centres are shown in Table 3. The patients at the 4 centres did not differ in age at onset of progressive hearing loss, duration of progressive hearing loss, age at onset of deafness, duration of deafness and age at implantation (Kruskal-Wallis nonparametric test, $P > 0.05$). The proportions of female patients per centre ranged from 23 to 44%; the differences were not significant (Kruskal-Wallis test). On the basis of these demographics, the patients at the 4 centres were considered to be largely comparable.

Preoperative evaluation data

At all 4 centres, the patients had to be profoundly deaf to enter the cochlear implantation programme, so variations in residual hearing were limited. Mean preoperative unaided hearing thresholds at 0.5, 1, 2 and 4 kHz exceeded 110 dB at all 4 centres.

The extent of otosclerosis on the CT scans was categorized into 3 types (Table 2). Although the patient group from Utrecht seemed to have a somewhat higher proportion of patients with Type 3 (i.e. severe retrofenestral otosclerosis/ unrecognizable otic capsule) and fewer patients with Type 2 (i.e. retrofenestral involvement) (Figure 1), chi-square tests revealed that these differences in occurrence of Type 1, 2 and 3 at the 4 centres were not significant. In addition, there were no significant differences in the proportions of patients with partial insertion of the electrode array between the 4 centres (Kruskal-Wallis tests; $P = 0.87$).

Table 3. Patient characteristics of implanted ears per CI centre (years)

Centre	Nijmegen (n = 13)			Utrecht (n = 9)			Birmingham (n = 17)			Manchester (n = 14)			P value
	25%	M	75%	25%	M	75%	25%	M	75%	25%	M	75%	
A@OHL	17 5	26.0	30 0	13 0	20 0	21 5	16 5	23 0	30 0	15 0	21 0	28 0	0 36
DoHL	26 0	30 0	33 5	7.5	26 0	30 5	7 0	17 0	35 5	7 5	25 0	33 5	0 36
A@OD	49 5	54.0	60 0	24 5	44 0	51.5	36.0	40 0	60 5	32 5	43 0	56 5	0.19
DoD	4 0	6 0	10 5	7 5	13 0	23 0	4 0	11 0	25 0	6 0	14 0	28 5	0 09
A@Impl	53 0	64.0	67 5	50 0	52 0	60 0	56 0	64 0	71 5	55 5	61 0	70 5	0 24

Abbreviations: A@OHL = age at onset of progressive hearing loss (years), DoHL = duration of progressive hearing loss (years), A@OD = age at onset of deafness (years), DoD = duration of deafness (years), A@Impl = age at implantation (years), 25% = 25th percentile; M = median, 75% = 75th percentile, P-value on Kruskal-Wallis test (Sign < 0.05)

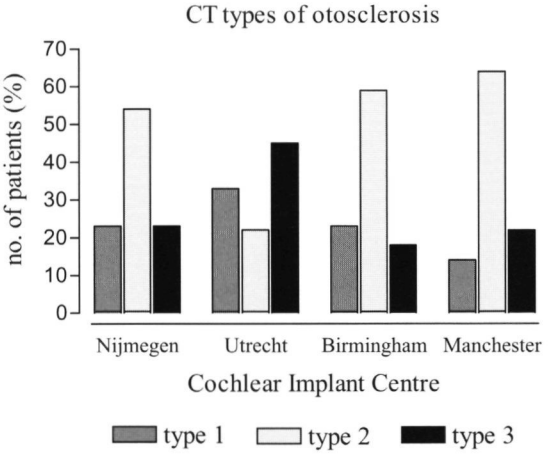


Figure 1. Severity of otosclerosis as categorized in 3 types per CI centre

Categorization according to patient performance

Phoneme scores

Phoneme scores were available of 19 out of the 31 English-speaking patients on the AB monosyllable test and of all 22 Dutch-speaking patients on the NVA monosyllables tests. The English and Dutch phoneme scores were pooled. In Figure 2, the phoneme scores at follow-up '0' were obtained directly after the sound processor had first been fitted. The figure shows that performance varied widely. Scores improved most sharply during the first 9 months, after which they seemed to stabilize. The patients were grouped according to their performance after 9 months of implant use. An evaluation of the phoneme scores of the reference group of 76 postlingually deaf adult CI patients showed a mean phoneme score of 55% and the 25th percentile at 40%. This 25th percentile was used as the criterion for inclusion in either the "better performance subgroup" (group 1) or the "poor performing subgroup" (group 2). Patients with a phoneme score of higher than 40% (n = 24) after more than 9 months follow-up, were categorized as better performers;whereas patients with a score of lower than 40% (n = 17), which is the 25th percentile in the adult postlingually deaf CI population, were categorized as poor performers (group 2).

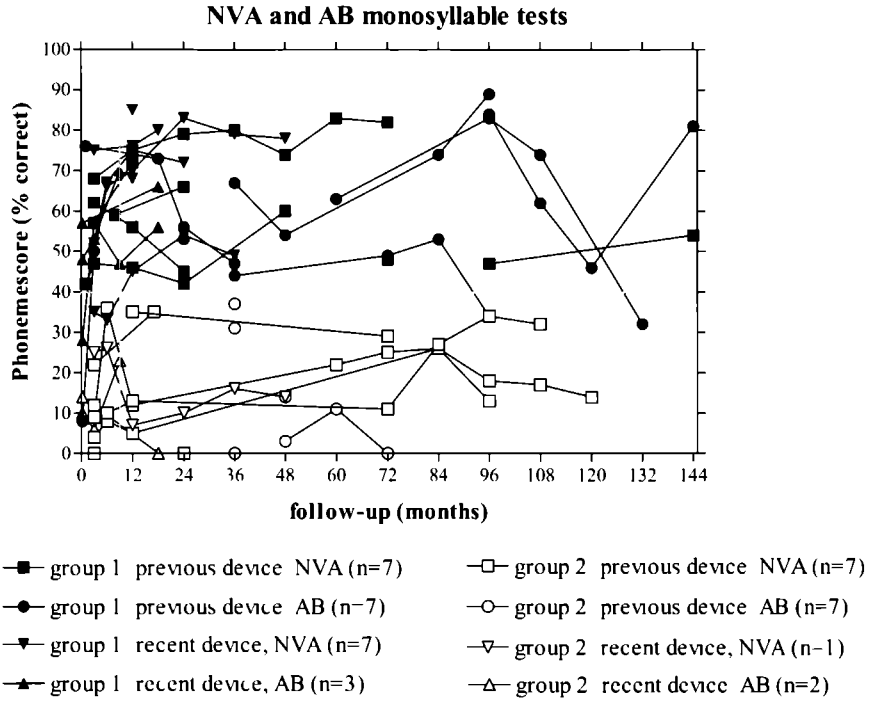


Figure 2 Phoneme scores of 19 English-speaking patients on the AB monosyllable test and of 22 Dutch-speaking patients on the NVA monosyllable test
Group 1 patients with a phoneme score of higher than 40% after more than 9 months follow-up (better performers), Group 2 – patients with a score of lower than 40% (poor performers)

Group 1 and 2, the better and poorer performers, did not differ in age at onset of hearing loss (Mann Whitney t-test, P = 0 32), duration of progressive hearing loss (P = 0 87), age at onset of deafness (P = 0 46) or duration of deafness (P = 0 65) The distributions of recent and previous generation devices and of NVA and AB monosyllable tests in the two groups were not significantly different (Fisher's exact test, P = 0 17) Analysis of the extent of otosclerosis on the CT scan between the better and poor performers revealed a tendency towards a lower proportion of patients with Type 1 otosclerosis and a higher proportion of patients with Type 3 otosclerosis in group 2, the poorer performers (Figure 3a), although significance was not reached (Fisher's exact test Type 1, P = 0 26, Type 2, P = 0 76, Type 3, P = 0 50) Figure 3a also shows that partial insertion of the electrode array, FNS and inactive electrodes (i e switched off during rehabilitation) were less common in group 1, the better performers, but again statistical significance was not reached in these groups (Fisher's exact test, P = 0 14, 0 10 and 0 06, respectively)

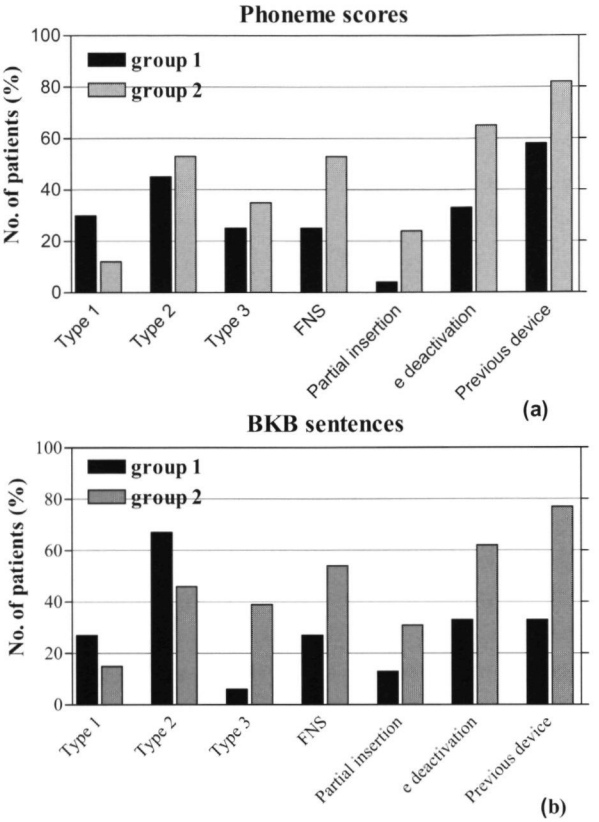


Figure 3. Comparisons between the better (group 1) and poor (group 2) performers on NVA and AB monosyllable test results (Figure 3a) and between the better (group 1) and poor (group 2) performers on BKB sentences test results (Figure 3b). Comparisons concerned CT Type (1-3), facial nerve stimulation (FNS), insertion depth (partial), electrode (e) deactivation during programming and generation of CI device used.

BKB sentences test scores

BKB sentences test scores had been obtained from 28 English-speaking patients. Data from the 2 English centres were combined, because there were no differences in patient characteristics, preoperative residual hearing, extent of otosclerosis on the CT scan, device type-related factors and test procedures. Figure 4 shows the scores on the BKB sentences test: performance varied widely. The patients were grouped according to their performance after more than 9 months of implant use, using the 25th percentile of the BKB data of the reference group as a criterion, which was 47% correct. The criterion for inclusion in group 1 was a score of higher than 47% (n = 15); individuals with a score of lower than 47% were placed in group 2 (n = 13). Group 1 and 2 did not differ in age at onset of hearing loss (Mann Whitney t-test, P = 0.78), age at onset of deafness (Mann Whitney t-test, P = 0.66),

duration of progressive hearing loss (Mann Whitney t-test, P = 0.55) nor duration of deafness (Mann Whitney t-test, P = 0.68).

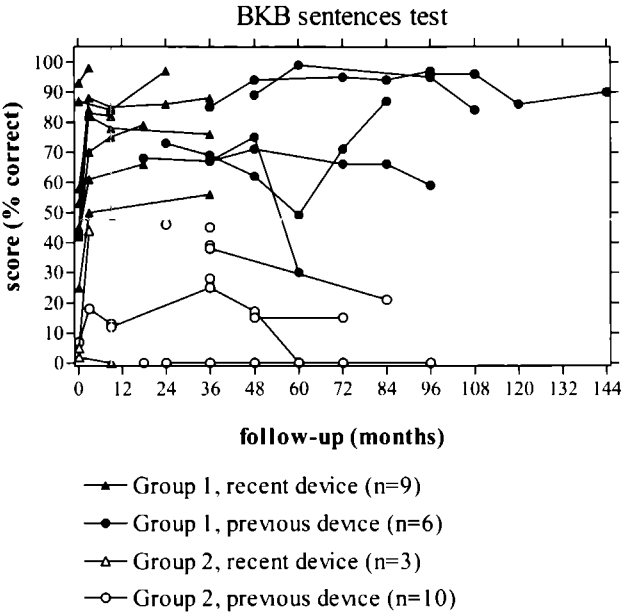


Figure 4. Scores on the BKB sentences test of 28 English-speaking patients Group 1 = patients with a score of higher than 47%, Group 2 = patients with a score of lower than 47%.

Figure 3b shows the extent of otosclerosis (Type 1, 2 or 3) on the preoperative CT scan per group. A larger proportion of patients in group 2 (poor performers) had Type 3 otosclerosis, i.e. severe retrofenestral otosclerosis with an unrecognizable otic capsule (Fisher's exact test, P = 0.07). Type 1 otosclerosis, i.e. solely fenestral involvement, was more frequent in group 1 (Fisher's exact test, P = 0.65). However, these differences were not significant.

Figure 3b further shows the percentages of patients with full and partial insertion per performance group. There were trends towards more patients with partial insertion among the poor performers (group 2) (Fisher's exact test, P = 0.37), a lower percentage of patients with FNS in group 1 (Fisher's exact test, P = 0.25) and a lower percentage of patients who had one or more inactive electrodes (i.e. that had been switched off at some point during rehabilitation to control FNS or other types of discomfort) in group 1 (Fisher's exact test, P = 0.25). Group 2 contained a larger proportion of patients with relatively older generation devices than group 1 (Fisher's exact test, P = 0.07).

Discussion

Results were available on 53 CI users with otosclerosis as the cause of deafness at 4 different CI centres. Similarities in demographic data, preoperative CT scans and audiological data meant that the patients from the 4 different centres could be considered as one group. The preoperative audiological data reflected that all 4 CI teams had employed conservative inclusion criteria. Nevertheless, significantly more patients at the Utrecht centre had undergone stapes surgery than the patients at the other centres. This was not considered to have had any important influence on later performance with a CI.

The phoneme scores obtained from the English and Dutch patients were pooled, although different tests had been used (AB and NVA monosyllable tests, respectively). Pooling was considered feasible, because the AB monosyllable test and the NVA monosyllable test have the same set-up, scoring procedure and level of presentation of the CVC words. Moreover, analysis of the distribution of NVA and AB monosyllable test results showed that these were equally distributed in the two groups (Fisher's exact test; $P = 0.54$). By pooling these data, the statistical power increased significantly.

A wide variation in the speech perception scores was observed between our subgroups of better and poor performers. No differences were found in demographic factors between the poor en better performers: the clinical presentation of the disease (rapid or slowly progressive) did not influence performance with a CI. Also, there were no differences in age at onset or duration of deafness between the two groups, although these factors were reported to be (more or less) influential in reports by other authors.[5,7,8] The differences between the poor and better performers comprised factors directly related to the disease (extent of otosclerosis on the CT scan, nonauditory sensations such as FNS) and factors indirectly related to the disease (fewer electrodes due to partial insertion or deactivation of electrodes). Obvious trends were seen: compared to the poor performers, the better performers had less severe otosclerosis on the CT scan, the majority had full electrode array insertion, very few had FNS and very few had deactivated electrodes. Similar patterns were seen in the phoneme scores and BKB sentences scores in the poor and good performers. Although many of the differences did not reach statistical significance, the similarities between the scores on these two speech recognition tests indicate that these differences are of importance.

Conclusions

A previous paper showed that CI surgery in patients with otosclerosis can be challenging with a relatively high number of partial insertions and misplacements of the electrode array demanding revision surgery. A very high percentage of patients was confronted with FNS mainly caused by the more distal electrodes on the array.

The present study showed wide variation in speech perception scores in patients with otosclerosis. Pooling of the data for statistical analysis was found feasible after analysis of the different test procedures. Several factors were identified to influence patients' performance. Good performance in patients with otosclerosis was related to less severe otosclerosis on the CT scan, full electrode array insertion, little or no FNS and little or no need to switch off electrodes. One indirect disease-related factor, the number of active electrodes, appeared to be the most important determinant of the outcome. Knowledge of these factors is of clinical importance during the patient selection period prior to implantation: in patients with this specific disease affecting the otic capsule, special emphasis can be put on the assessment of the cochlear structure. During counseling, the probability of a successful rehabilitation with the CI may be estimated by the CT scan obtained and by the acknowledgement of a potential partial electrode array insertion. Although exact predictions about the benefit remain uncertain and unwise, this knowledge may be of value for the patient with otosclerosis in order to develop realistic expectations.

References

1. Dowell RC, Mecklenburg DJ, Clark GM. Speech recognition for 40 patients receiving multichannel cochlear implants. *Arch Otolaryngol Head Neck Surg* 1986,112 1054-1059.
2. Oh SH, Kim CS, Kang EJ et al. Speech perception after cochlear implantation over a 4-year time period. *Acta Otolaryngol* 2003,123:148-153.
3 Valimaa TT, Sorri MJ, Lopponen HJ Speech perception and functional benefit after multichannel cochlear implantation *Scand Audiol Suppl* 2001;45-47.
4. Hamzavi J, Baumgartner WD, Pok SM et al. Variables affecting speech perception in postlingually deaf adults following cochlear implantation. *Acta Otolaryngol* 2003,123:493-498.
5. Battmer RD, Gupta SP, Lum-Mecklenburg DJ et al. Factors influencing cochlear implant perceptual performance in 132 adults. *Ann Otol Rhinol Laryngol Suppl* 1995;166:185-187.
6. Shipp DB, Nedzelski JM Prognostic indicators of speech recognition performance in adult cochlear implant users. a prospective analysis *Ann Otol Rhinol Laryngol Suppl* 1995;166·194-196.
7. Shea JJ, Domico EH, Orchik DJ. Speech recognition ability as a function of duration of deafness in multichannel cochlear implant patients *Laryngoscope* 1990,100:223-226
8. Blamey P, Arndt P, Bergeron F et al. Factors affecting auditory performance of postlinguistically deaf adults using cochlear implants. *Audiol Neurootol* 1996;1:293-306.
9. Rubinstein JT, Parkinson WS, Tyler RS et al. Residual speech recognition and cochlear implant performance: effects of implantation criteria. *Am J Otol* 1999;20:445-452

10. Skinner MW. Optimizing cochlear implant speech performance. *Ann Otol Rhinol Laryngol Suppl* 2003;191 4-13.
11. Rotteveel LJ, Proops DW, Ramsden RT et al. Cochlear implantation in 53 patients with otosclerosis: demographics, computed tomographic scanning, surgery, and complications *Otol Neurotol* 2004;25:943-952.
12. Woolford TJ, Roberts GR, Hartley C et al. Etiology of hearing loss and cochlear computed tomography: findings in preimplant assessment. *Ann Otol Rhinol Laryngol Suppl* 1995;166:201-206.
13. Guneri EA, Ada E, Ceryan K et al. High-resolution computed tomographic evaluation of the cochlear capsule in otosclerosis. relationship between densitometry and sensorineural hearing loss. *Ann Otol Rhinol Laryngol* 1996;105:659-664.
14. Linthicum FH, Jr., Filipo R, Brody S. Sensorineural hearing loss due to cochlear otospongiosis theoretical considerations of etiology. *Ann Otol Rhinol Laryngol* 1975,84:544-551.
15. Mafee MF, Valvassori GE, Deitch RL et al. Use of CT in the evaluation of cochlear otosclerosis. *Radiology* 1985,156:703-708.
16. Youssef O, Rosen A, Chandrasekhar S et al. Cochlear otosclerosis: the current understanding. *Ann Otol Rhinol Laryngol* 1998;107:1076-1079.
17. Linthicum FH, Jr. Histopathology of otosclerosis. *Otolaryngol Clin North Am* 1993;26: 335-352.
18. Ramsay HA, Linthicum FH, Jr. Mixed hearing loss in otosclerosis indication for long-term follow-up. *Am J Otol* 1994;15:536-539.
19. Browning GG, Gatehouse S. Sensorineural hearing loss in stapedial otosclerosis. *Ann Otol Rhinol Laryngol* 1984;93:13-16
20. Blamey PJ, Pyman BC, Gordon M et al. Factors predicting postoperative sentence scores in postlinguistically deaf adult cochlear implant patients. *Ann Otol Rhinol Laryngol* 1992,101:342-348.
21. Nadol JB, Jr., Young YS, Glynn RJ Survival of spiral ganglion cells in profound sensorineural hearing loss: implications for cochlear implantation. *Ann Otol Rhinol Laryngol* 1989;98:411-416.
22. Bench J, Kowal A, Bamford J. The BKB (Bamford-Kowal-Bench) sentence lists for partially-hearing children *Br J Audiol* 1979;13:108-112
23. Boothroyd A. Statistical theory of the speech discrimination score *J Acoust Soc Am* 1968; 43:362-367.
24. Bosman AJ, Smoorenburg GF. Intelligibility of Dutch CVC syllables and sentences for listeners with normal hearing and with three types of hearing impairment. *Audiology* 1995; 34:260-284
25. Michael M. Practical aspects of Audiology. Speech Audiometry. Whurr Publishers Ltd, 1987.

Chapter 6

Cochlear implantation in Osteogenesis Imperfecta

Cochlear implantation in 3 patients with Osteogenesis Imperfecta: imaging, surgery and programming issues

L.J.C. Rotteveel
A.J. Beynon
L.H.M. Mens
J.J. Mulder
A.F.M. Snik
E.A.M. Mylanus

Audiology & Neurotology 2008, 13 73-85

Summary

Objectives: to describe the surgery and rehabilitation after cochlear implantation of patients with severe sensorineural hearing loss due to Osteogenesis Imperfecta.

Methods: 3 patients with Osteogenesis Imperfecta were retrieved from the Nijmegen/Viataal Cochlear Implant Centre's database. The patient's perioperative imaging, medical charts and programming notes were evaluated. Objective electrophysiological measures (evoked compound action potentials (ECAPs), averaged electrode voltages (AEVs) and spatial spread of neural excitation) as well as subjective psychoacoustical measures, such as electrical threshold and comfortable level determination and pitch scaling estimation were performed.

Results: Most of the specific observations in ear surgery on patients with Osteogenesis Imperfecta, such as brittle scutum, sclerotic thickening of the cochlea, hyperplastic mucosa in the middle ear and persistent bleeding, were encountered. In Case 3, with severe deformities on the CT scan, misplacement of the electrode array into the horizontal semicircular canal occurred. In all 3 Cases, programming was hindered by nonauditory stimulation. Even after reimplantation, nonauditory sensations lead to Case 3 becoming a nonuser. AEVs in Case 3 were deviant in accordance with an abnormally conductive otic capsule. Spatial spread of neural excitation responses in Cases 1 and 2 suggested intracochlear channel interaction for several electrodes, often in combination with facial nerve stimulation (FNS). In Case 1, the estimated pitch of the electrodes that caused FNS varied consistently. However, after 1-year follow-up, open-set phoneme scores of 81% and 78% were reached in Cases 1 and 2, respectively.

Conclusions: When aware and prepared for the specific changes of the temporal bone in Osteogenesis Imperfecta, cochlear implantation can be a safe and feasible procedure. Preoperative imaging is recommended to be fully informed on the morphology of the petrosal bone. In case of severe deformities on the CT scan, during counseling the possibility of misplacement should be mentioned. Rehabilitation is often hindered by FNS requiring frequent refitting. Despite the electrophysiological changes, Cases 1 and 2 had high phoneme scores.

Introduction

Cochlear implantation is nowadays the treatment of choice for rehabilitation of motivated patients affected by severe to profound sensorineural hearing loss (SNHL) who do not benefit from traditional amplification Great advances have been made in restoring auditory perception to both children and adults alike However, the benefit provided by the cochlear implant (CI) varies widely [1] Numerous factors have been related to postoperative performance as reflected in speech perception tests [2] These factors include residual hearing, previous hearing aid use, age at onset of deafness, duration of deafness, age at implantation, integrity of the auditory nerve and central auditory pathways, intelligence, postoperative communication mode and educational setting, device type, insertion depth and number of activated electrodes Etiology of deafness has also been recognized as a factor of influence on performance [3,4] Constraints on performance such as a limited use of intracochlear electrodes (e g in congenital malformation of the cochlea or basal turn ossification in meningitis), histological alterations of the temporal bone (e g decalcification in otosclerosis), or neuronal lesions (e g ganglion cell and/or central neural survival in meningitis) may be related to etiology

At the Nijmegen/Viataal CI centre, 3 patients with a rare bone disease, Osteogenesis Imperfecta (OI), have been enrolled in the CI rehabilitation programme OI is a heterogeneous disease of the connective tissue caused by a defective gene (COL1A1 and COL1A2 located on chromosome 17 and 7, respectively) that is responsible for the production of collagen type I, leading to defective bone matrix and connective tissue Bones become brittle and are easily fractured The inheritance may be autosomal dominant or recessive The incidence of about 1 in 20000 subjects is maintained by a high rate of new mutations in most families [5] Various tissues are involved in the disease bone, dentine, tendon, blood vessels, heart valves and skin The severity of the disease is roughly correlated with the reduction in collagen I synthesis

According to Sillence et al [6], the disease can be classified in four different types (I, II, III and IV) with further division into subgroups depending on the presence of blue sclerae, abnormal dentition, the severity of bone fragility and hearing impairment (Table 1)

Although histological, biochemical and clinical features of OI and otosclerosis frequently coexist, otosclerosis and OI are different diseases Unlike otosclerosis, OI is not limited to the otic capsule In OI, the bone of the otic capsule shows more resorption spaces filled with connective tissue and a greater degree of structural disorganization In otosclerosis, spongiotic lucencies and sclerotic dense areas of bone with narrowing of the cochlear lumen are present predominantly at the basal turn [7]

Table 1. Classification of Osteogenesis Imperfecta

Bone fragility	Stature	Blue sclerae	Dental defects	Hearing impairment	Inheritance
Mild to severe bone fragility Late fractures	Normal or slightly short stature	Yes	Some	Some (highest incidence of all types)	AD
Extreme bone fragility Perinatal lethal fractures	NA	Yes	Some	NA	Sporadic new mutations
Neonatal severe bone fragility, Progressive skeletal deformity	Short stature	Blue at birth Not as adults	Some	Some	AR or sporadic new mutations
Mild to severe bone fragility Moderate skeletal deformity	Often short stature	No	Some	Some (lowest incidence of all types)	AD

NA = not applicable because of intrauterine or early infantile death, AD – autosomal dominant, AR = autosomal recessive

The CT findings of the petrosal bone in OI may be as follows: (1) extensive demineralised bone involving all or part of the otic capsule, which has a much lower attenuation on the CT scan, resulting in a so called 'halo' around the cochlea or 'double ring effect'; (2) fenestral manifestations caused by proliferation, such as a narrow middle ear cavity, enveloped stapes footplate and obliterated windows with irregular and indistinct margins; (3) extension of the dysplastic, demineralised bone as high as the upper margin of the superior semicircular canal; (4) involvement of the facial nerve canal in the dysplastic process, resulting in facial nerve paresis or paralysis.[8] The two entities most closely resembling OI of the temporal bone on a CT scan are Paget's disease (osteitis deformans) and otosclerosis. In Paget's disease, which is characterized by an abnormally rapid rate of bone turnover, the temporal bone involvement is usually accompanied by changes of the skull. Severe otosclerosis may be indistinguishable on a CT scan from OI, except for differences in degree and extent. In OI the thickness of the prolific bone appears to be much greater. The bony labyrinth is more frequently involved and to a more extensive degree, extending even above the superior semicircular channel.[5,8] In magnetic resonance imaging (MRI), pericochlear, ring-like enhancing soft tissue lesions can be found in both OI and otospongiosis, the active stage of otosclerosis.[9] In a previous study on patients with otosclerosis, the severity of the petrosal bone pathology was categorized according to the affected region on the CT scan, i.e. fenestral and/or retrofenestral.[10] The same can be done for patients with OI.

Hearing loss affects 35-60% of the patients, most often in the form of the conductive or mixed type.[11] Conductive hearing loss may be caused by fixation of the stapes footplate, by fracture or aplasia of one or both stapedial crura, or by distal atrophy or absence of the long process of the incus. The sensorineural component has been thought to be the result of abnormal bone encroaching on the cochlea causing mechanical distortion of the basilar

membrane, tiny fractures of the otic capsule, haemorrhage into the labyrinth, otosclerotic foci stealing blood from the cochlear microcirculation and interference with the mechano-electric function of hair cells by toxic enzymes [8 12] Pure SNHL in OI is rare (10%) [12] The hearing loss usually begins in the late teens, with the sensorineural component appearing in the third decade [13] It gradually leads to profound deafness, tinnitus, and vertigo by the end of the forth to fifth decade [12] As the hearing loss will progress to deafness in a varying reported amount (2%[11] - 11%[14]) of OI patients, cochlear implantation may become the only remaining treatment option in some patients

The aim of this study was to describe a series of 3 patients with OI and the specific problems encountered during surgery and rehabilitation after cochlear implantation It was hypothesized that in patients with OI and affected temporal bones the electrical resistance in the bone may be lower, causing extracochlear current spread leading to nonacoustic nerve stimulation, such as facial nerve stimulation (FNS), and a lesser frequency specificity from multichannel stimulation To evaluate this, objective electrical and electrophysiological measures (evoked compound action potentials, ECAPs, averaged electrode voltages, AEVs, and spatial spread of neural excitation) were performed Further, subjective psychoacoustical measures (electrical threshold, comfortable (C) level determination and pitch scaling estimation) were performed

Patients and Methods

Preoperative findings

The clinical diagnosis of OI was based on the presence of blue sclerae, a history of multiple fractures and a strong family history of OI The patients' preoperative CT or MRI scans and postoperative imaging were examined The CT scans in a high-resolution osseous window-level setting (HRCT) were performed in the axial and coronal planes The section thickness was 1 0 mm using contiguous sections The MRI examination was performed in the axial and coronal plane using T_1- and T_2-weighted spin echo sequences The CT scans were reviewed for fenestral involvement (i e narrowed or enlarged window, thickened footplate) and retrofenestral involvement (i e double ring effect, narrowed basal turn) of the petrosal bone They were categorized in three types (Table 2) [10] To evaluate for specific differences between the preoperative CT findings of patients with OI and patients with otosclerosis, the CT scans of 13 CI subjects with otosclerosis Type 2 (n = 8) and Type 3 (n = 5) were reviewed to evaluate the degree of demineralization with special emphasis on the superior semicircular channel

Table 2. Manifestations of otosclerotic or otospongiotic lesions on the CT scans: 3 types

Hypertrophic or demineralized lesions of the otic capsule	
Type 1	Solely fenestral involvement (thickened footplate and/or narrowed or enlarged windows)
Type 2	Retrofenestral, with or without fenestral involvement Type 2a: double ring effect Type 2b: narrowed basal turn Type 2c: double ring and narrowed basal turn
Type 3	Severe retrofenestral (unrecognizable otic capsule) involvement, with or without fenestral involvement

Surgery and programming

The medical charts and programming notes were evaluated with special attention to the surgery reports, the occurrence of FNS and deactivation of electrodes.

Electrophysiological measures

Electrophysiological measures comprised measurements of electrically evoked compound action potential (ECAP) threshold, average electrode voltages (AEVs) and spatial spread of neural excitation.

Neural response telemetry (NRT) has been widely used intra- and postoperatively to measure ECAPs in CI subjects[15,16] using a 'masker electrode /probe electrode -stimulus paradigm' stimulating the *same* electrode. The ECAP threshold measured by NRT is referred to as T-NRT.[16]

AEVs are implant-generated, far-field surface potentials recorded from scalp electrodes during stimulation through a CI. AEVs are used clinically to provide an objective assessment of internal device function and to identify malfunctioning electrodes, especially in devices lacking a back-telemetry facility. In the present two cases with a Nucleus device (Cases 1 and 2), AEVs were recorded with surface electrodes typically placed at the ipsilateral mastoid (positive), high forehead (reference), and the wrist (ground). AEVs of Case 3, implanted with a Clarion device, were obtained similarly, except that the reference electrode was placed on the contralateral mastoid.

In contrast to threshold determination to obtain T-NRTs, the 'masker electrode /probe electrode -stimulus paradigm' can also be used to determine electrode or channel interaction.[17,18] Here, ECAPs are recorded from one specific probe electrode while the masker stimulus is stimulating *another* electrode. With increasing distance between the stimulated masker electrode and the stimulated probe electrode, the masker becomes less sufficient and the amplitude of the ECAP measured at the probe electrode will decrease. Generally, electrode interaction function shows a peak at the electrode where probe and masker stimuli are stimulating the same electrode, i.e. when the distance between probe

and masker electrode is nil. Spatial spread of neural excitation is expressed as a function of ECAP amplitude over varying 'probe electrode /masker electrode'-distances. These measurements were performed using standard clinical NRT v3.1 software (Cochlear Ltd.).

Psychoacoustical measures

Subjective psychoacoustical measures comprised determination of the behavioural electrical threshold (T level) and comfortable level (C level), as well as measurements of pitch scaling estimation and speech perception. T and C levels were obtained using standard clinical fitting software.

To establish behavioural pitch estimation in Cases 1 and 2, separate electrodes were randomly stimulated at C level with 1000 ms biphasic pulse trains according to Busby and Clark.[19] The subjects were asked to judge the pitch in a scale ranging from 0 to 100; '0' is defined as a sound representing a low pitch and '100' a high pitch. All electrodes were randomly stimulated 4 times per electrode. Mean subjective ratings on the 100-point scale of these 4 sessions were calculated.

Speech perception was tested by obtaining mean phoneme scores on standardized open-set monosyllabic wordlists.[20]

Results

Preoperative findings

Case 1 represents a female with blue sclera and a history of multiple fractures had been known with progressive mixed hearing loss since the age of 13 for which conventional hearing aids were fitted. At otoscopy a positive Schwartz's sign was noticed. The hearing loss progressed to profound hearing loss at the age of 43 years. Her preoperative aided open set speech recognition score was 30% phonemes. She complained of tinnitus and vertigo present in a variable degree. Calorisation tests demonstrated no abnormalities. Electrocochleography ruled out the presence of an airbone gap suitable for stapes surgery. The MRI scan performed during the selection period for cochlear implantation is shown in Figure 1.

Case 2 represents a female with blue sclera and a history of multiple spontaneous fractures as a child and a progressive hearing loss of the left ear since her youth which was reported to have been a conductive hearing loss initially. The right ear had been deaf since the age of 6, presumable after stapes surgery. This was confirmed by an audiogram at the age of 19. Otoscopy at the left ear showed a positive Schwartze's sign and at the right ear a partially retracted ear drum.

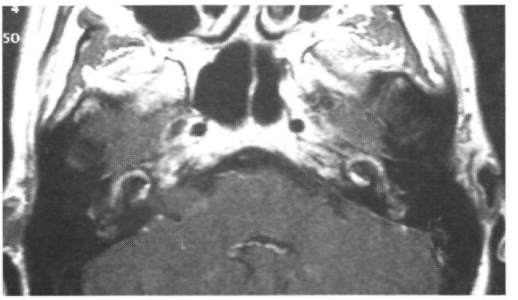


Figure 1a. Preoperative MRI (T_1 weighed with contrast) of the petrosal bone (transversal view): pericochlear enhancement

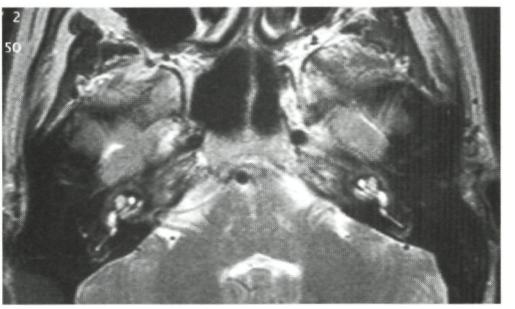


Figure 1b. Preoperative MRI (T_2 weighed) of the petrosal bone (transversal view): cochlea and labyrinth are both patent, on the right more eminent than the left side. There is a pericochlear high signal.

Figure 1. Imaging in Case 1

The bone conduction threshold of the left ear started to deteriorate 7 years later. She suffered from tinnitus. At the age of 17 a conventional hearing aid had been fitted at the left side, but was no longer useful at age 49. There was a positive family history for OI with progressive hearing loss in her mother, two sisters, son and a nephew. In the work-up for cochlear implantation, the calorisation test showed areflexia. She had no (aided) open-set speech recognition. Preoperative imaging is shown in Figure 2 and described in Table 3.

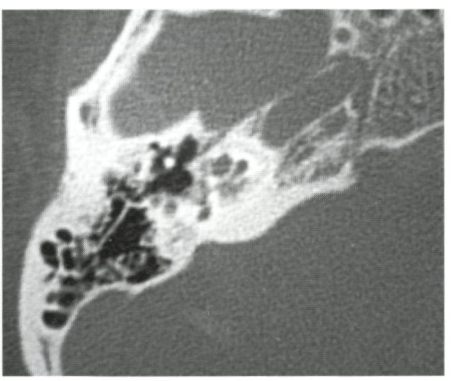


Figure 2. Imaging in Case 2; preoperative CT scan right petrosal bone (transversal view): pericochlear lucency - double ring effect - and narrowed basal turn (Type 2c).

Table 3. Case reports in the literature

Author	OI type / diagnosis	Type of HL, DoD	Temporal bone CT findings	A@I (yrs)	Surgery	Programming	Speech perception
Szyfkowski et al. (1998)	NM / "a history of established OI"	Progressive SNHL, 19 years	Loss of cochlear architecture with demineralization, annular osteolysis in otic capsule	50	Ossification of ossicular chain; easy full insertion of Nucleus 22	FNS, e9-13 switched off	NM
Huang et al. (1998)	NM / clinical diagnosis	Progressive mixed HL, 3 years	Otospongiotic change of both cochleas with pericochlear lucency	42	Hypervascular ME mucosa; brittle cochlear bone; easy, full insertion of Nucleus 22*	NM	Vowel perception 94% ; consonant perception 62% ; sentence perception 59%
Migirov et al. (2003)	NM / clinical diagnosis	Profound HL, NM	Normal	6	Normal ossicles and ME; easy, full insertion of Nucleus Contour	No FNS; normal electrical stimulation levels and electrode impedance values	Monosyllabic word identification 25% at 6-months follow-up
Streubel et al. (2005) Case 1	Type Ia / clinical diagnosis	Progressive SNHL, NM	"A pattern similar to significant cochlear otosclerosis"	35	Hypervascular ME mucosa; extensive fenestral bony growth; vascular bone; easy, full insertion of MedEl Combi 40	FNS, "several electrodes", management NM	Phoneme score 75%, word score 54% at 1-year follow-up (CNC)
Streubel et al. (2005) Case 2	Type Ia / clinical diagnosis	Progressive SNHL, NM	Some demineralisation fenestral and lateral to the basal turn	NM	Hypervascular ME mucosa; sclerotic promontory, easy, full insertion of Nucleus Contour	No FNS	Phoneme score 83%; word score 70% at 1-year follow-up (CNC)
Present study Case 1	Type I / genetic diagnosis	Progressive mixed HL, 2 years	Demineralisation of otic capsule, patent cochlea (MRI)	45	Hypervascular ME mucosa; brittle cochlear bone; easy, full insertion of Nucleus 24	FNS, e13-18 switched off and current levels of e14 and e19 lowered	Phoneme score 84%; word score 60% at 1-year follow-up (NVA)
Present study Case 2	Type I / clinical diagnosis	Progressive mixed HL, 2 years	Fenestral abnormalities and pericochlear lucencies AIDS; patent basal turns	51	Hypervascular ME mucosa; incus and stapes not identifiable; easy, full insertion of Nucleus 24	FNS, e20-22, e1 and e2 switched off	Phoneme score 78%; word score 56% at 1-year follow-up (NVA)
Present study Case 3	Type I / clinical diagnosis	Progressive mixed HL, 5 years	Loss of architecture of the cochlea, demineralization but patent scalae	54	Hypervascular ME mucosa; gusher; extensive fenestral bony growth; misplacement of Clarion CI array	FNS and other severe discomfort for which all e's switched off	No speech perception (non-user)

Abbreviations: Clinical diagnosis signifies the diagnosis "OI" based on clinical symptoms; Genetic diagnosis signifies the diagnosis "OI" based on genetic research; NM = not mentioned; Progr = progressive; FNS = facial nerve stimulation; ME = Middle ear; HL = hearing loss; DoD = duration of deafness; SNHL = sensorineural; A@I = age at implantation; yrs = years; e = electrode; * postoperative CT demonstrated curl of the electrode array; CNC = monosyllabic word recognition test (Consonant-Noun-Consonant); NVA = monosyllabic word recognition test (Nederlandse Vereniging van Audiologen, Dutch Society of Audiologists)

Case 3, representing a man with blue sclerae, dental defects and a history of multiple fractures, started to suffer from bilateral progressive hearing loss at the age of 13, which progressed to total deafness of both ears at the age of 25. At the age of 26, an attempt was made to improve thresholds by stapes surgery on both ears. This led to a temporary improvement and conventional hearing aids were fitted on both ears. After 2 years, the hearing aid was no longer beneficial at the left ear, followed by the right ear 23 years later; he no longer had profitable residual hearing. He experienced tinnitus occasionally. There was a positive family history for OI with hearing loss in father, mother and one uncle. Vestibular tests showed severe hyporeflexia. Both CT and MRI scans demonstrated a loss of architecture of the cochlea, demineralization but patent scalae of the cochlea, especially at the right ear (Figure 3a, Table 3).

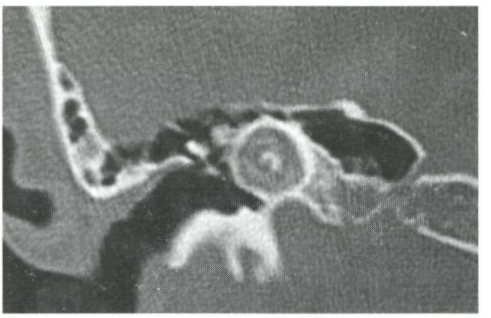


Figure 3a. Preoperative CT scan right petrosal bone (coronal view): unrecognizable otic capsule (Type 3).

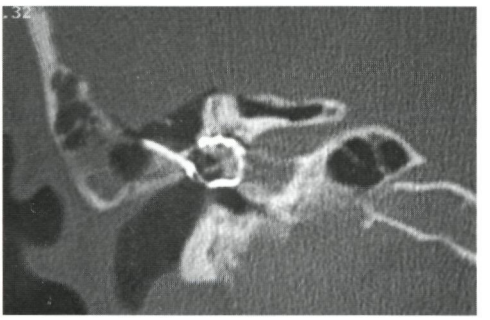


Figure 3b. Postoperative CT scan right petrosal bone (coronal view): the electrode array is in the lateral semicircular canal.

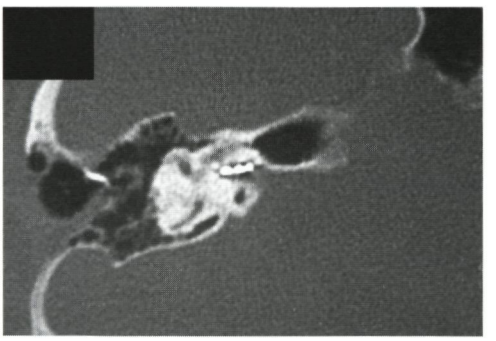


Figure 3c. Postoperative CT scan right petrosal bone (coronal view): the electrode array penetrates the internal auditory canal.

The CT scans of all 3 Cases with OI showed demineralization extending as high as the superior semicircular channel The CT scans of 13 CI subjects with otosclerosis Type 2 (n = 8) and Type 3 (n = 5) showed demineralization extending as high as the superior semicircular channel in only 3 subjects

Surgery and postoperative findings

In Case 1, at the age of 45 a Nucleus 24M was implanted at the right ear The promontory was covered by a highly vasculated and hyperplasic mucosa Cochleostomy was hindered by brittle and easily bleeding bone A full insertion of the electrode array in a patent scala tympani was achieved A postoperative CT scan showed a good position of the electrode array Reviewing this CT scan for classification (Table 2) showed a Type 2c for the left, non-implanted ear

In Case 2, the cochlear implantation performed at the right ear at the age of 51 was uneventful The partially retracted ear drum showed a perforation posterior of the malleus for which a tympanoplasty Type I was performed The long proces of the incus had been eroded The stapes suprastructure and chorda tympani could not be identified, probably due to the previous stapedotomy that presumably had caused acute deafness Cochleostomy revealed a patent lumen in which full insertion took place The third day postoperatively the head bandage was removed and a haematoma had to be aspirated This further resolved spontaneously Postoperative modified Stenvers X-ray showed a normal insertion

In Case 3, at the age of 54 cochlear implantation was performed at the right ear, leaving the stapes prosthesis in situ Middle ear mucosa was hyperemic After cochleostomy, perilymph leakage occurred The insertion of the electrode array of a Clarion 1 2 device (without positioner) was easy and complete in a patent scala tympani The electrode array on the postoperative X-ray appeared to make a turn of approximately 180 degrees but was slightly wrinkled at the tip Unfortunately, the postoperative switch-on could not elicit hearing sensations The patient experienced severe vertigo A subsequent CT scan 5 months postoperatively, demonstrated that the array had entered the lateral semicircular canal (Figure 3b) During revision surgery, a new cochleostomy was made, this time slightly more towards the round window niche, which was ossified extensively The same device was pulled back and reimplanted, after a check of the technical integrity using back-telemetry On the second postoperative X-ray the array seemed to be in the basal turn A control CT scan could confirm this, but also showed that the tip of the electrode array had entered the internal auditory canal (Figure 3c)

In all 3 Cases, intraoperative electrode impedances measured with the clinical back-telemetry system were within normal range

Programming

In Case 1, postoperative electrode impedances showed normal values for all electrodes. The patient was fitted optimally with a standard monopolar Advanced Combination Encoder (ACE) speech coding strategy. Mean threshold (T) levels were 180 (SD = 10) current units (cu) and mean C levels 207 (SD = 13) cu. Electrodes 15 to 18 showed FNS when activated above 177 cu and were therefore switched off. Stimulation levels of electrodes 14 and 19 were lowered below C levels, but above T levels, in order to control for FNS.

All electrodes in Case 2 had normal impedances. The patient was fitted optimally with a standard monopolar ACE speech coding strategy. Mean T levels were 158 (SD = 7) cu and mean C levels were 217 (SD = 6) cu. Because of FNS, 3 apical (electrodes 20 to 22) and 2 basal (electrodes 1 and 2) electrodes were switched off. Electrodes 20 to 22 showed FNS above behavioural C level, electrodes 1 and 2 showed FNS below C level.

Unfortunately even after revision surgery, Case 3 had no hearing sensations at all. Rehabilitation proved quite difficult. Nonauditory sensations such as discomfort, pain and FNS were present when the device was switched on, so that several electrodes had to be switched off. Even brief use of the CI caused extensive tinnitus and headache. Within 3 months time the number of usable electrodes was reduced to two, despite frequent refitting. Eventually, this patient became a nonuser. His vertigo worsened, possibly due to progression of the OI.

Electrophysiological results

ECAP thresholds obtained using standard neural response telemetry in Case 1 revealed stable T-NRTs for all electrodes with hearing sensations. Electrodes 14 to 20 caused FNS, making it impossible to obtain ECAP thresholds.

ECAP thresholds in Case 2 revealed T-NRTs for all electrodes with hearing sensations, except for electrodes 1, 2, 20, 21 and 22: because of FNS the ECAPs could not be obtained on these electrodes. Mean T-NRTs were measured at 202 (SD = 9) cu.

In Case 3 there was no auditory sensation at all; therefore ECAPs, speech perception scores and behavioral pitch estimation could not be obtained.

Figure 4a shows AEVs of Cases 1 and 2 obtained with monopolar stimulation (MP) at 100 cu for all 22 electrodes. Figure 4b shows AEVs of Cases 1 and 2 obtained with bipolar stimulation between different configurations (BP+3). Responses were evoked by standard biphasic pulses (25 us/phase, with 8 us interphase gap) with stimulation rate of 900 pps. Both measurements show consistent AEVs similar to that of other CI subjects.

In Figure 4c, the mean peak-to-peak value of the AEVs of Case 3 after medial monopolar, lateral monopolar and enhanced bipolar stimulation are shown. Note that the 8 electrodes

of the Clarion electrode array are numbered from apical to basal, in contrast to the reversed numbering used in the Nucleus device. Responses were evoked by biphasic pulses of 300 us/phase with an amplitude of 16 cu in monopolar mode and 50 cu in bipolar mode. The AEV recordings of Case 3 appear to be decreased compared to 4 control patients (postlingually deaf adults with normal petrosal bone anatomy, implanted with the same devices in the same year as Case 3). The bipolar recordings of Case 3 show phase-reversed AEVs (below '0') for all except the two most apical electrodes.

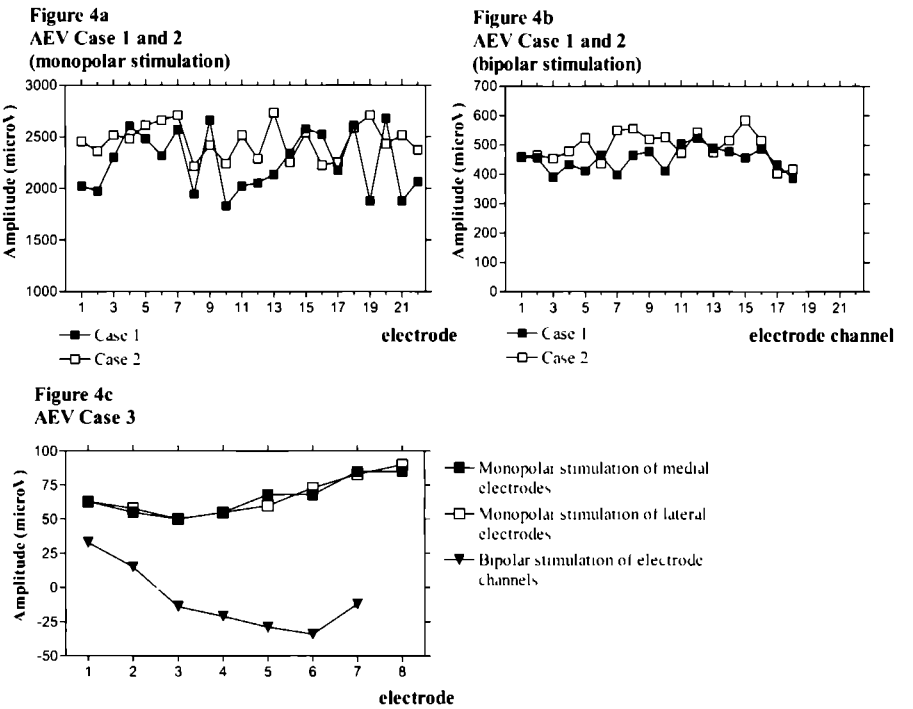


Figure 4. AEV measurements obtained with monopolar (MP) and bipolar stimulation

To analyse intracochlear channel interaction, spatial spread of neural excitation expressed as a function of ECAP amplitude was measured for several electrodes. Steeper slopes around an electrode imply less channel interaction and might imply better frequency specificity. Figure 5a shows the spread of excitation (SOE) responses of Case 1 for a basal, medial and apical electrode (electrodes 4, 11 and 22, respectively). Stimulating electrodes 14 to 18 did not produce consistent ECAPs, but caused FNS instead. Nevertheless, in two electrodes, 14 and 16, a spread of excitation pattern, although rather flat, could be elicited. Figure 5b shows spatial spread of neural excitation responses of Case 2 for electrodes 2, 4, 12, 15 and 16 Amplitudes were much higher compared to Case 1. Apical and medial

electrodes 16, 15 and 12 showed steeper slopes compared to the basal electrodes 2 and 4. With respect to electrodes that did not cause FNS, both Cases 1 and 2 seem to show similar SOE patterns as CI subjects with normal petrosal bones, i.e. highest ECAP amplitude around the stimulating probe electrode.

Figure 5a:
Spatial Spread of Neural Excitation in Case 1

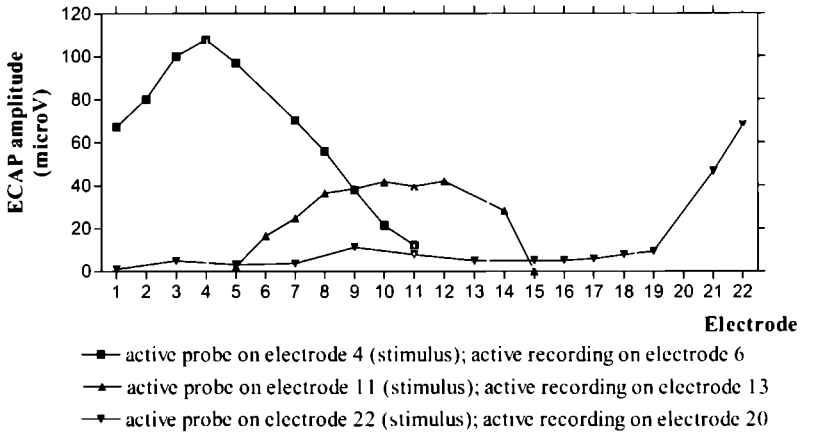


Figure 5b:
Spatial Spread of Neural Excitation in Case 2

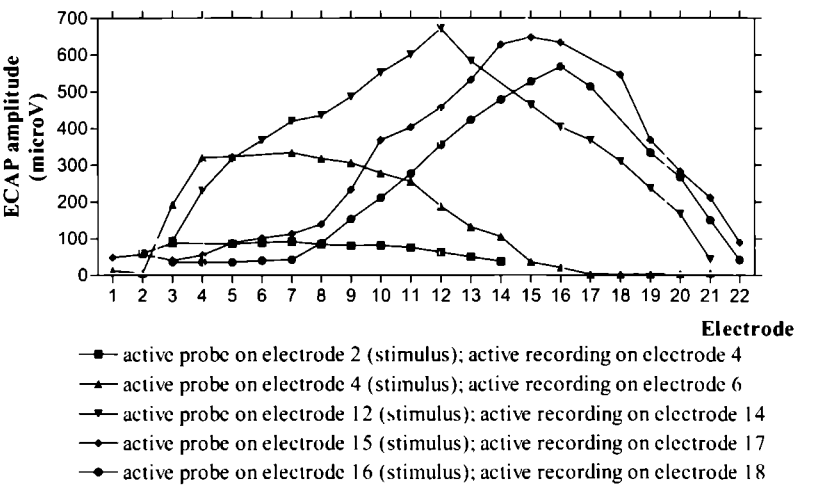


Figure 5. Spatial spread of neural excitation.
Stimulus electrodes are indicated with vertical dashed lines; electrodes lacking an ECAP amplitude measurement are the electrodes on which the active recording took place; the masker active electrode varied; areas of electrodes that had no reproducible ECAPs are indicated with the grey squares, in Case 1 FNS occurred at electrodes 14 to 20, in Case 2 at electrodes 1, 2, and 20 to 22.

Psychoacoustical measures

The results of the subjective pitch estimation per electrode by Case 1 are shown in Figure 6a. Electrodes that caused FNS are indicated within the grey square: the estimated pitch of these electrodes varied consistently. Figure 6b shows the subjective pitch estimation by Case 2. The estimated pitch varies the most for the medial electrodes. However, the mean scores (a high pitch on the basal and a low pitch on the apical electrodes) reveal that generally, the tonotopy of the cochlea is well perceived.

Figure 6a:
Mean and SD of estimated pitch in Case 1

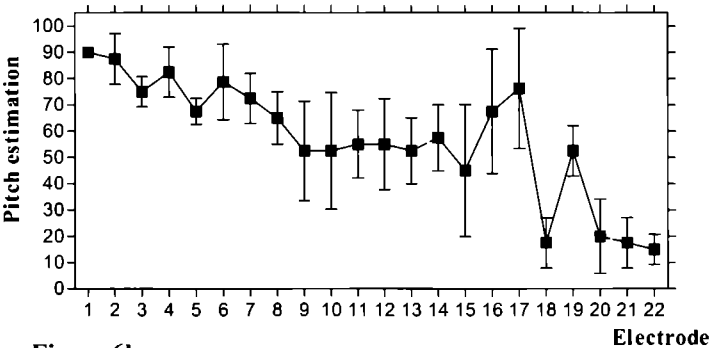


Figure 6b:
Mean and SD of estimated pitch in Case 2

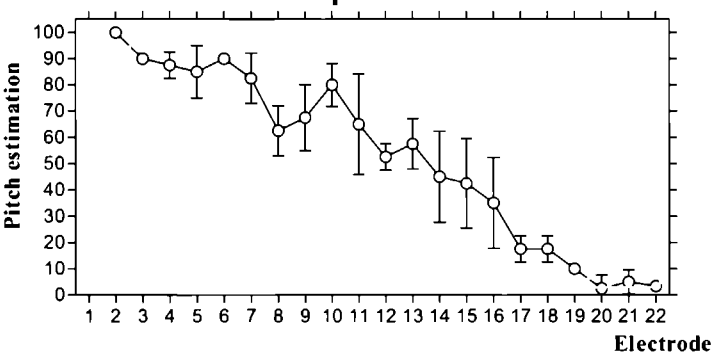


Figure 6 Pitch estimation The behavioural responses were obtained using a pitch estimation experiment on a 100-point scale. Each electrode was measured 4 times Mean scores and standard deviation of these 4 trials are shown. Electrodes that caused FNS are indicated in the grey squares.

After 1 year of implant use, Case 1 reached a phoneme score of 81% (Figure 7) and a word score of 60%. Case 2 had a 78% phoneme score (Figure 7) and a 56% word score at 1-year follow-up. At 6-years follow-up, these scores remained stable. Figure 7 also shows the phoneme scores of 8 subjects with varying CT types of otosclerosis, who also had a full insertion of identical Nucleus 24 devices. Cases 1 and 2 have comparable phoneme scores to the subjects who had CT scans showing the less severe otosclerosis Type 1 and 2.

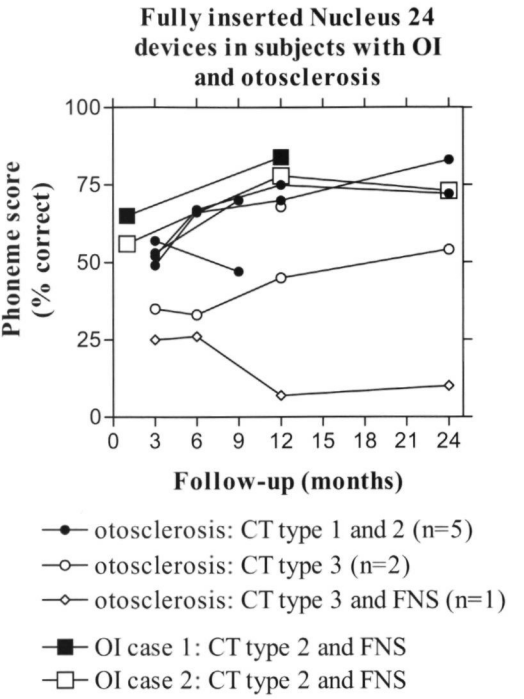


Figure 7. Phoneme scores in 2 patients with OI and 8 patients with otosclerosis implanted with a Nucleus 24 device.

Discussion

Preoperative findings

We report 3 patients with OI who received a CI. The diagnosis OI is based on clinical (i.e. increased fragility of bone associated with involvement of other connective tissue, such as blue sclerae, abnormal dentition, hearing loss, or a combination) and genetic criteria. Without knowledge of the clinical symptoms, imaging modalities such as CT or MRI can hardly differentiate between otosclerosis and OI. D'Archambeau et al.[25] described the differential diagnosis of otodystrophic lesions of the temporal bone and state the importance of HRCT as the primary imaging modality in evaluating osseous lesions of the temporal bone and labyrinth. In agreement with the literature[5,8], demineralization extending as high as the superior semicircular channel was present on the CT scans in only 3 out of 13 subjects with otosclerosis, whereas it was present in all CT scans of the 3 subjects with OI. It can, however, not be considered diagnostic for OI. While at present the direct molecular characterization is not feasible in the majority of cases, demonstration of

reduced synthesis of procollagen I by dermal fibroblasts is indictive for the disorder The diagnosis of OI in the present cases could be confirmed by the clinical features described in the patients medical charts The patient described in Case 1 had also participated in research by Garretsen et al [26] describing otological and clinical genetic aspects in OI type I Imaging in all 3 cases corresponded with the diagnosis

In the literature, 5 cochlear implantations in OI patients, of which one child, have been described [21 24] The CT scans of these patients and the present 3 cases showed pericochlear demineralization of varying extend in all patients, except for one normal CT scan in the only child in the series [23] Previously, the findings on the CT scan in OI patients had proven not to be correlated to the severity of the hearing loss [5] The diagnosis OI in this child with an established bilateral profound SNHL since the age of 6 months, was based on the presence of blue sclera, a history of fractures and the occurrence of these features in other family members CI surgery and rehabilitation were uneventful The CT scan of Case 3 showed the most severe lesions of all 3 cases described in the present study Cochlear implantation in this patient was the most complicated, even requiring revision surgery

Surgery

In all the adult OI patients presented here and in the literature, the implantation was technically more challenging compared to a routine procedure, mainly due to the vascularity of the spongiotic bone In the present first 2 cases, no major surgical complications were encountered, except in Case 3 Identifying the location of the round window niche by approximation from the stapes super structure was found especially difficult in Case 3 because of bone proliferation, a problem that had been encountered in OI patients before [24] In case of bone proliferation, obliteration of the basal turn should also be expected This is a common feature seen after bacterial meningitis with labyrinthitis ossificans and might require special drilling procedures [27] Preoperative MRI can be helpful in predicting cochlear patency and determining which ear to implant In none of the present OI patients or the patients reported in the literature was obliteration of the basal turn encountered and in all patients a full insertion of the electrode array could be achieved In Case 3, in which the CT scan showed severe deformities, misplacement of the electrode array in the otospongious bone occurred The possibility of misplacement of the electrode array in an otospongiotic otic capsule has also been described in patients with otosclerosis the array might penetrate an anatomical lumen such as a semicircular canal[10], mastoid cavity or internal meatus, or it might penetrate a newly formed osteolytic cavity in the otic capsule [28]

Vertigo

Vertigo was most disabling in Case 3, progressing long after he had stopped using his CI. Vertigo has been found to be common in patients with OI, and in most cases this is secondary to inner ear pathology.[29] After the first implantation in Case 3, when the electrode array had been misplaced in the lateral semicircular canal, the vertigo worsened. After reimplantation with removal of the array out of the semicircular channel, the vertigo lessened to a degree comparable to the preoperative status. The prevalence of postoperative vertigo after cochlear implantation varies considerably, ranging from 4% to 75%, the most common type being delayed in onset.[30] Four years after the reimplantation, calorisation tests in Case 3 showed hypofunction of the left vestibular system and no responses on the right implanted side. Because of this areflexia on the implanted side, the vertigo was believed to be caused by progression of the OI.

Speech perception

The success of the implantation in Cases 1 and 2 is reflected in their relatively high phoneme scores. Good speech perception in OI patients has also been reported by Streubel et al.[24] The elimination of some basal, medial or apical electrodes in Cases 1 and 2 does not seem to influence the speech perception by a lack of spectral or temporal information. This is in agreement with earlier studies by Wilson[31] reporting that interleaved stimulation in quiet is sufficient with only 7 active channels.

Facial Nerve Stimulation

FNS in Cases 1 and 2 was relatively easily treated by deactivation of some electrodes or stimulating below C level. Nonauditory sensations in Case 3, however, could not be controlled by programming adjustments. FNS is a common complication of cochlear implantation in patients with otosclerosis (38%), affecting a higher proportion of patients implanted with non-modiolushugging devices (44%) compared to patients implanted with modiolushugging devices (10%).[10] Programming details on the occurrence of FNS were available for 7 out of the 8 OI subjects summarized in Table 3; 5 subjects (70%) were affected by it. FNS has been postulated to be the result of deviant intracochlear current spread in dehiscent or otospongiotic bone because of low-impedance pathways, which give rise to an electrical field in the proximity of the facial nerve.[32] To avoid an unacceptable decrement in sound quality due to programming maneuvres such as inactivation of electrodes in order to correct FNS, some authors suggest fluoride treatment[33] or botulinum toxin[34] and even reimplantation using a device with modiolar facing contacts and perimodiolar position[35].

Average Electrode Voltage

The deviant intracochlear current spread in Case 3 has been briefly mentioned before, in a study on AEVs.[36] AEV amplitudes vary widely among subjects, partly because of insertion depth[37], but can still be considered to be a stable 'fingerprint' of the individual current spread within and outside the cochlea provided that stimulus and recording parameters are optimized. Normative data have been established, both for the Nucleus[37,38] and the Clarion device[39]. However, it has been shown that bipolar AEVs in patients with abnormal cochleae and/or abnormal electrode insertion are significantly deviant.[40] In the case of a well-isolated cochlea, AEV recordings decrease when stimulating more apically placed electrodes. The bipolar recordings from Case 3 do not decrease in the apical electrodes and further show phase-reversed AEVs for the basal electrodes. This deviant pattern was felt to be the result of the decalcified cochlea, as similar patterns were observed in patients with otosclerosis[41], although an erroneous location of the electrode array could not be excluded. No deviant AEVs were found in Cases 1 and 2 for either monopolar or various bipolar stimulation modes. The CT scans in these two subjects showed less demineralization compared to Case 3.

Spatial spread of neural excitation

In the present study, the neural excitation pattern was measured stimulating different masker and probe electrodes. In contrast to the study by Cohen et al.[42], in which the spread of neural excitation is described using NRT profiles obtained with masker and probe stimulus on the same electrode ('simple ECAP' method), the use of the 'advanced ECAP' method in our study revealed some effect of channel interactions, which may be recognized as a flat morphology of the response curve (e.g. electrode 11 in Case 1, electrodes 2 and 4 in Case 2).

The pattern of excitation is most likely affected by factors such as stimulus current level, neural survival, the presence of new bone formation or fibrous tissue, and the electrode-modiolar distance. The electrodes showing FNS showed inconsistent responses or even absent ECAPs, although this might be due to the fact that FNS appeared at stimulation levels below T-NRT. In Case 1, we did not find reproducible ECAPs for electrodes 15 to 18, that had to be deactivated in order to control for FNS. Because electrodes 14 and 19 did not cause FNS at C level, but led to some hearing sensations, these electrodes were switched on. Multicentre NRT data reported that ECAPs could be elicited in 96% of the cases.[43] It is obvious that this is not found in the OI patients. Nevertheless, the morphology of the spread of excitation patterns of those electrodes *without* FNS seems to be similar to those from other (non-OI) patients. However, a decreased spread of excitation at the electrodes positioned more deeply than 270°, as described previously by Cohen et al.[17], was not found in our cases.

Pitch estimation

Electrode discrimination experiments have shown that multichannel CIs exploit the tonotopic organisation of the cochlea[44,45] which enhances a better speech perception [19,46] The estimated pitch by Case 1 varied consistently for the electrodes that caused FNS Possibly, the spread of electrical current produced by these electrodes and the resulting nonauditory sensations made it more difficult to estimate the perceived pitch In Case 2 however, the estimated pitch during 4 subsequent measures varied less and overall, the tonotopy of the cochlea was well perceived despite FNS

Conclusions

Abnormal bone structure may evoke difficulties during CI surgery and postoperative rehabilitation and stimulation Specific observations in ear surgery on patients with OI have been reported, such as a thin external auditory canal skin, brittle scutum, sclerotic thickening of the cochlea, hyperplastic mucosa in the middle ear and persistent bleeding Most of these were encountered in the present patients and in the cases described in the literature undergoing cochlear implantation When aware and prepared for this, cochlear implantation can be a safe and feasible procedure in patients with OI Preoperative imaging is recommended to be fully informed on the morphology of the petrosal bone, preferably CT scanning and MRI In case of severe deformities on the CT scan, during counseling the possibility of misplacement and consequent disappointing results should be mentioned AEV values and ECAP reproducibility suggest a deviant current spread As a result of this, rehabilitation is often hindered by FNS requiring frequent refitting

References

1. Clark GM. Results: Speech perception with Cochlear Implants. 726-738, 2003b In: Cochlear Implants: Fundamentals and Applications. Beyer RT (ed), AIP Press
2. Clark GM Preoperative Selection 550-594, 2003a. In: Cochlear Implants: Fundamentals and Applications. Beyer RT (ed), AIP Press
3. Battmer RD, Gupta SP, Allum-Mecklenburg DJ et al. Factors influencing cochlear implant perceptual performance in 132 adults. *Ann Otol Rhinol Laryngol Suppl* 1995;166:185-187.
4 Blamey P, Arndt P, Bergeron F et al Factors affecting auditory performance of postlinguistically deaf adults using cochlear implants. *Audiol Neurotol* 1996;1 293-306.
5. Ross UH, Laszig R, Bornemann H et al Osteogenesis imperfecta: clinical symptoms and update findings in computed tomography and tympano-cochlear scintigraphy. *Acta Otolaryngol* 1993,113:620-624.
6. Sillence DO, Senn A, Danks DM. Genetic heterogeneity in osteogenesis imperfecta. *J Med Genet* 1979;16:101-116.
7. Nager GT Osteogenesis imperfecta of the temporal bone and its relation to otosclerosis *Ann Otol Rhinol Laryngol* 1988;97:585-593.
8. Tabor EK, Curtin HD, Hirsch BE et al Osteogenesis imperfecta tarda. appearance of the temporal bones at CT. *Radiology* 1990,175.181-183.
9 Ziych S, Berger R, Reisner K. MRI-visible pericochlear lesions in osteogenesis imperfecta type I. *Eur Radiol* 2000,10:1675-1677.
10. Rotteveel LJ, Proops DW, Ramsden RT et al. Cochlear implantation in 53 patients with otosclerosis: demographics, computed tomographic scanning, surgery, and complications. *Otol Neurotol* 2004,25:943-952.
11. Garretsen AJ, Cremers CW, Huygen PL. Hearing loss (in nonoperated ears) in relation to age in osteogenesis imperfecta type I *Ann Otol Rhinol Laryngol* 1997;106.575-582
12. Pedersen U. Osteogenesis imperfecta clinical features, hearing loss and stapedectomy. Biochemical, osteodensitometric, corneometric and histological aspects in comparison with otosclerosis. *Acta Otolaryngol Suppl* 1985;415:1-36.
13. Stewart EJ and O'Reilly BF. A clinical and audiological investigation of osteogenesis imperfecta. *Clin Otolaryngol Allied Sci* 1989,14:509-514.
14. Pedersen U. Hearing loss in patients with osteogenesis imperfecta. A clinical and audiological study of 201 patients. *Scand Audiol* 1984,13.67-74.
15. Abbas PJ, Brown CJ, Shallop JK et al: Summary of results using the nucleus CI24M implant to record the electrically evoked compound action potential. *Ear Hear* 1999;20 45-59
16. Dillier N, Lai WK, Almqvist B et al. Measurement of the electrically evoked compound action potential via a neural response telemetry system *Ann Otol Rhinol Laryngol* 2002;111:407-414.
17. Cohen LT, Saunders E, Richardson LM Spatial spread of neural excitation: comparison of compound action potential and forward-masking data in cochlear implant recipients. *Int J Audiol* 2004;43:346-355.
18. Eisen MD and Franck KH Electrode interaction in pediatric cochlear implant subjects. *J Assoc Res Otolaryngol.* 2005,6 160-170.
19. Busby PA and Clark GM Pitch estimation by early-deafened subjects using a multiple-electrode cochlear implant. *J Acoust Soc Am* 2000,107.547-558.
20. Bosman AJ. Speech perception by the hearing impaired. Thesis, Utrecht 1998
21. Szilvassy J, Jori J, Czigner J et al. Cochlear implantation in osteogenesis imperfecta. *Acta Otorhinolaryngol Belg* 1998;52:253-256
22. Huang TS, Yen PT, Liu SY. Cochlear implantation in a patient with osteogenesis imperfecta and otospongiosis. *Am J Otolaryngol* 1998,19.209-212
23 Migirov L, Henkin Y, Hildesheimer M et al. Cochlear implantation in a child with osteogenesis imperfecta *Int J Pediatr Otorhinolaryngol* 2003;67.677-680
24. Streubel SO and Lustig LR. Cochlear implantation in patients with osteogenesis imperfecta. *Otolaryngol Head Neck Surg* 2005;132:735-740.

25. D'Archambeau O, Parizel PM, Kockelkoren E et al. CT diagnosis and differential diagnosis of otodystrophic lesions of the temporal bone. *Eur J Radiol* 1990;11:22-30.
26. Garretsen AJ. Osteogenesis Imperfecta Type I, otological and clinical genetic aspects. Thesis, Radboud University Nijmegen, 1992.
27. Rotteveel LJ, Snik AF, Vermeulen AM et al. Three-year follow-up of children with postmeningitic deafness and partial cochlear implant insertion. *Clin Otolaryngol* 2005,30:242-248
28 Ramsden R, Bance M, Giles E et al. Cochlear implantation in otosclerosis: a unique positioning and programming problem *J Laryngol Otol* 1997,111.262-265
29 Kuurila K, Kentala E, Karjalainen S et al Vestibular dysfunction in adult patients with osteogenesis imperfecta *Am J Med Genet* 2003;120:350-358.
30. Handzel O, Burgess BJ, Nadol JB, Jr. Histopathology of the peripheral vestibular system after cochlear implantation in the human *Otol Neurotol* 2006,27:57-64.
31. Wilson BS. The future of cochlear implants. *Br J Audiol* 1997;31:205-225.
32. Bigelow DC, Kay DJ, Rafter KO et al. Facial nerve stimulation from cochlear implants. *Am J Otol* 1998,19:163-169.
33. Gold SR, Miller V, Kamerer DB et al. Fluoride treatment for facial nerve stimulation caused by cochlear implants in otosclerosis. *Otolaryngol Head Neck Surg* 1998,119:521-523.
34. Langman AW, Quigley SM, Heffernan JT et al. Use of botulinum toxin to prevent facial nerve stimulation following cochlear implantation *Ann Otol Rhinol Laryngol Suppl* 1995;166 426-428
35. Battmer RD, Pesch J, Joseph G et al. Eliminating facial nerve stimulation by reimplantation of a nucleus 24 contour implant system 2004, Conference Proceeding.
36. Mens LH and Mulder JJ. Averaged electrode voltages in users of the Clarion cochlear implant device. *Ann Otol Rhinol Laryngol* 2002;111:370-375.
37 Mens LH, Oostendorp T, van den Broek P. Identifying electrode failures with cochlear implant generated surface potentials. *Ear Hear* 1994b;15:330-338
38 Shallop JK. Objective electrophysiological measures from cochlear implant patients *Ear Hear* 1993;14:58-63.
39 Hughes ML, Brown CJ, Abbas PJ. Sensitivity and specificity of averaged electrode voltage measures in cochlear implant recipients. *Ear Hear* 2004;25:431-446.
40 Mens LH, Oostendorp T, van den Broek P. Cochlear implant generated surface potentials: current spread and side effects. *Ear Hear* 1994a;15.339-345.
41 Taitelbaum-Swead R, Kishon-Rabin L, Kaplan-Neeman R et al. Speech perception of children using Nucleus, Clarion or Med-El cochlear implants. *Int J Pediatr Otorhinolaryngol* 2005;69:1675-1683
42. Cohen LT, Richardson LM, Saunders E et al. Spatial spread of neural excitation in cochlear implant recipients comparison of improved ECAP method and psychophysical forward masking. *Hear Res* 2003,179:72-87.
43. Cafarelli DD, Dillier N, Lai WK et al Normative findings of electrically evoked compound action potential measurements using the neural response telemetry of the Nucleus CI24M cochlear implant system. *Audiol Neurotol* 2005,10.105-116.
44. Collins LM, Zwolan TA, Wakefield GH. Comparison of electrode discrimination, pitch ranking, and pitch scaling data in postlingually deafened adult cochlear implant subjects *J Acoust Soc Am* 1997;101:440-455
45. Tong YC and Clark GM. Percepts from scala tympani stimulation. *Ann N Y Acad Sci* 1983;405:264-267.
46. Donaldson GS and Nelson DA Place-pitch sensitivity and its relation to consonant recognition by cochlear implant listeners using the MPEAK and SPEAK speech processing strategies. *J Acoust Soc Am* 2000,107:1645-1658

Chapter 7

Cochlear implantation in the compromised cochlea

Summary and conclusions

7.1 Results of cochlear implantation

Multi-channel intracochlear CI systems are nowadays widely used; there are more than 100,000 implant users worldwide. That electrical stimulation of the impaired auditory pathway of a deaf person can lead to speech understanding is quite amazing. A CI can even be argued to be the most successful neural prosthesis.

The outcomes of cochlear implantation have been evaluated by numerous groups using varying measures to assess the outcome results. Speech perception tests are widely used in children and adults to evaluate cochlear implantation.[1-6] Other means of assessing the results of cochlear implantation are measurements of changes of voice and articulation, speech production, vocabulary development, receptive and expressive language skills, narrative abilities, educational placement, academic and/or occupational status, and literacy outcomes. Further, objective electrophysiological measurements such as auditory evoked cortical potentials are used to evaluate the benefit of cochlear implantation.[7] The last years, research has also focussed on the influence of cochlear implantation on the patients' quality of life.[8]

A problem with speech perception testing in children is the variability of linguistic abilities.[9,10] Usually, repeated speech perception measurements are performed in a single-subject design. In this way perceptual performance can be monitored as a function of duration of implant use. During the follow-up of a child with a CI, because of increased experience and maturing speech development, basal speech tests might show ceiling scores and more difficult tests floor scores. To deal with this problem, at the CI centre Nijmegen/Viataal the children are subjected to a test battery which quantifies speech perception on different levels of discrimination, suprasegmental identification, word identification and open-set word recognition tests. This test battery has been administered to a large group of profoundly and severely hearing-impaired children with binaural powerful conventional hearing aids whose hearing loss ranged from 50 to 130 dB HL PTA for reference purposes. The established relations between the test scores and the hearing loss in that reference group were used in reverse to express the scores of a child with a CI in one single measure, called the "overall Equivalent Hearing Loss" (EHL). Analysis of EHL values in **Chapter 2** showed that congenitally, prelingually and postlingually deaf children all benefit from their cochlear implant for speech perception tasks, but performance varied greatly. During the first 2 years after implantation, postlingually deaf children showed the fastest rate of improvement. After 3 years of implant use, the early implanted prelingually deaf children and congenitally deaf children implanted under the age of 6 years caught up with the postlingually deaf children. Prelingually deaf children implanted after a relatively long duration of deafness tended to show poorer performance than those with a shorter duration. After early implantation, the levels of performance that

were eventually achieved differed no more than 10 dB, irrespective of whether the onset of deafness was prelingual or postlingual. Performance of congenitally deaf children implanted after the age of 6 years was poorer and progress was slower. In congenitally deaf children, duration of deafness played a major role in speech perception performance, whereas in children with acquired deafness, communication was a major factor. Thus, the earlier a deaf child is implanted, the better his or her speech perception performance after 3 years of CI use. This is in accordance with other studies.[6 11-14] In recent years, the evidence-based opinion that early implantation results in better speech, language and listening outcomes resulted in a decline of the typical age at which children receive a cochlear implant. This opinion coincides with observations that humans seem to be better at learning speech and language when they are young than when they are older; there is a special time in development, termed either *critical period* or *sensitive period*, during which speech and language are learned efficiently. Commonly, *sensitive periods* are defined as a gradual time in development in which the organism is particularly responsive to experience based on an 'age-related plasticity', whereas a *critical period* is viewed as a rather fixed time window in development in which experience, or the absence of experience, results in a complete irreversible change in the brain.[15] When handling issues on speech and language development, although not scientifically based, we prefer using the term *sensitive period*. Physiological animal model experiments have indicated that the auditory system has considerable age-related plasticity.[16] This plasticity is reflected in the ability of the human auditory system to adapt to the novel stimulation delivered by the CI, which becomes obvious when documenting the performance of a CI patient. Patient's performance thus is related to age at implantation, or duration of auditory deprivation. Our data suggest that the sensitive period ends somewhere around the age of 6 years. Harrison et al.[17] could not detect a clear universal age or define a critical period during which cochlear implantation provides a clearly superior performance.

Due to the conservative inclusion criteria for cochlear implantation in the past and the more and more improved CI systems and experienced CI rehabilitation programmes the data presented in Chapter 2 are not representative for the children implanted in more recent years. However, these conservative inclusion criteria resulted in a rather homogenous study group concerning factors such as intelligence and amount of parental support. Communication mode at that time was not individually determined, but depended on the school to be predominantly oral-aural or solely signs based. The homogenous study group enabled us to study the effects of various variables on speech perception. In contrast, the present children enrolled in the CI rehabilitation program form a very heterogenous group concerning intelligence, parental support and educational placement due to the less strict inclusion criteria in which these variables cannot be tested.

7.2 Cochlear implantation in the compromised cochlea

With the demonstrated benefit of cochlear implantation in patients who suffered from profound sensorineural hearing loss with little benefit from conventional amplification, the indication for cochlear implantation has broadened considerably. This includes implantation in morphologically changed, compromised cochleae such as in congenital malformed inner ears, post-meningitis ossified cochleae and extensive cochlear otosclerosis. Consequently, the surgical challenge has increased and revision surgery has become more frequent.[18] Other changes in candidacy which have surgical implications are: the age of the candidate[19-21], presence of residual hearing[22] and multihandicapted patients[23,24]. New devices have contributed to expansion, surgical techniques have been modified and become more reliable. Complications have diminished still further from their previously already low and acceptable level. Although the improvement of CI performance noted in the past decade is usually attributed to technical innovation, it may also be caused in part by favorable characteristics of CI recipients such as shorter duration of deafness, more residual hearing, or younger age.

It is important to report on the difficult surgical cases and share the complications that have occurred so that other professionals might learn from the described experiences. This enables surgeons to be prepared for special circumstances, which must be discussed with the patients and their families in advance. Extreme cases of compromised cochleae are rare and it is therefor recommended to perform the surgical procedure in experienced CI centres.

7.2.1 Cochlear implantation in the pediatric compromised cochlea

Chapter 3 describes the results of 7 children with postmeningitic deafness and partial insertion of the Nucleus electrode array due to ossification of the cochlea and of 18 children with postmeningitic deafness and full insertion of the electrode array.

In 10 children, during surgery the preoperatively identified ossification on CT scan could be confirmed (sensitivity 53%). In 9 children, no ossification was visible on the CT scan, but was indeed encountered during surgery (false negative rate 47%): despite normal cochlear appearance on CT scans, the presence of ossification must be expected in a child with postmeningitic deafness, thus additional MRI is mandatory.

Both groups of children were evaluated with the same battery of speech perception tests, which can be reduced into an EHL value as described above. Three years after implantation, the children with partial insertion showed slower progress and they reached a relatively poor EHL plateau score. Patients with partial insertion do benefit from a CI, although less than patients with complete insertion. This implies that postmeningitic deaf children should receive a CI soon after the infection before ossification of the cochlea occurs. Nowadays, there is nation-wide consensus between ENT specialists and

pediatricians on early evaluation by audiometry and in case of hearing loss referral to an ENT specialist in children diagnosed with bacterial meningitis [25] In addition to the reduced number of electrodes, there are other explanations for poor speech perception when severe ossification leads to partial insertion, suboptimal modiolus-array proximity and a less favourable (broadly spread) electrical current in a drilled tunnel might negatively influence CI benefit in children with postmeningitic deafness

Patients with severe inner ear malformations are expected to perform less than patients with normal developed cochlea because of the likelihood of a decreased number of spiral ganglion cells associated with cochlear malformation, and because of the more complex surgical challenges in such malformed ears [26] In congenital malformations of the inner ear, abnormalities of the sensory epithelium are often associated with relatively poor development of neural elements Schmidt[27] found an average spiral ganglion cell count of 11,500 in Mondini's dysplasia compared to cell counts in the mid-20,000 range in otosclerosis or ototoxicity and ganglion cell counts of approximately 33,000 in normal-hearing persons Fortunately, temporal bone studies learned that benefit from cochlear implantation can occur in patients with as few as 3300 ganglion cells [28] To study the surgical aspects and performance outcome of cochlear implantation in children with malformed inner ears, in **Chapter 4** a clinical and audiometric evaluation is presented of 13 CI patients who had a variety of inner ear malformations Viewing the patients from this study and patients from a review of the literature concerning cochlear implantation in children with malformed inner ears including severe cochlear malformations, the occurrence of an aberrant facial nerve was 17%, which rises to 27% if one reviews the surgical findings in children with severe malformed cochleae like a common cavity or a severe cochlear hypoplasia In all 13 presented patients a complete insertion of all active electrodes was accomplished At 1 year of follow-up, for most children the open-set phoneme score could be measured Some patients however had limited language abilities and did not have an open speech perception yet, possibly due to young age, long duration of deafness or short follow-up However, they did demonstrate closed set speech perception, or at least an increased awareness of environmental sounds Generally, in patients with mild cochlear deformities, full insertion of the electrode array is possible and results can be obtained comparable to those obtained in profoundly deaf patients with normal cochleae[29], whereas patients with severe inner ear malformations are expected to perform less than patients with normal cochleae because of the likelihood of a decreased number of spiral ganglion cells and recurrent meningitis, and because the more complex surgical challenges [30] Although the result of cochlear implantation may be promising, as in our patient with a common cavity, during preoperative counselling, the child's parents should be informed that the result is uncertain

7 2 2 Cochlear implantation in the adult compromised cochlea

In the adult CI population 7 to 9 5% of patients who received a CI have been diagnosed with otosclerosis [31] The majority of the 53 patients with otosclerosis retrieved from 4 different CI centres described in **Chapter 5** had a preoperative CT scan demonstrating retrofenestral (cochlear) otosclerotic lesions, which had a tendency towards being more extensive in patients with rapidly progressive hearing loss, surgically problematic insertion of the electrode array and facial nerve stimulation In four patients revision surgery had to be performed A very high proportion of patients (38%) experienced facial nerve stimulation mainly caused by the distal electrodes

There was wide variability in the speech perception results Poor and good performers did not differ in age at onset of hearing loss, duration of hearing loss rate of progression, age at onset of deafness, or duration of deafness Better performance however was related to less severe signs of otosclerosis on CT scan, full insertion of the electrode array and little or no facial nerve stimulation One indirect disease-related factor, the number of active electrodes, appeared to be the most important determinant of the outcome This is in agreement with our findings in postmeningitic deaf children as described in Chapter 3, the full-insertion group, with an average of 20 active electrodes, had significantly better speech perception than partial-insertion subjects in whom 8 to 13 electrodes had been implanted

In **Chapter 6** the surgical procedure and rehabilitation after cochlear implantation of 3 patients with severe sensorineural hearing loss due to Osteogenesis Imperfecta are described The diagnosis Osteogenesis Imperfecta could be confirmed by the clinical features described in the medical charts and imaging in all 3 cases corresponded with the diagnosis Most of the specific observations in ear surgery on patients with Osteogenesis Imperfecta, such as brittle scutum, sclerotic thickening of the cochlea, hyperplastic mucosa in the middle ear and persistent bleeding, were encountered In Case 3, with severe deformities on the CT scan, misplacement of the electrode array into the horizontal semicircular canal occurred The possibility of misplacement of the electrode array in an otospongiotic otic capsule has also been described in patients with otosclerosis [32,33]

In all 3 cases, programming was hindered by non-auditory stimulation Even after reimplantation, nonauditory sensations lead to Case 3 becoming a nonuser Averaged electrode voltages (AEVs) in Case 3 were deviant in accordance with an abnormally conductive otic capsule Spatial spread of neural excitation responses in Cases 1 and 2 suggested intracochlear channel interaction for several electrodes, often in combination with facial nerve stimulation (FNS) In Case 1, the estimated pitch of the electrodes that caused FNS varied consistently Nevertheless, after 1-year follow up, open-set phoneme scores as high as 81% and 78% were reached in Cases 1 and 2, respectively

Future research

In the last two decades, research and development in cochlear implantation enforce collaboration of various disciplines including physicians, engineers, and scientists and has resulted in new implant designs, electrode array configurations, specialized soft ware and lower power consumption. Refinements in speech coding algorithms have led to a tremendous rise in CI patients' speech perception scores. Further development is aimed to achieve even higher resolution without compromising in power consumption. A more focused stimulation may lead to a decrease in negative side effects such as facial nerve stimulation, which particularly is important for patients with otosclerosis.

As mentioned above, the CI can be seen as the most successful neural prosthesis. The auditory brainstem implant (ABI) is a modification of the CI for patients who cannot be fitted with CIs because of the presence of severely compromised cochlea or cochlear nerve malfunction, in which the electrode array is placed directly onto the brainstem. Initially it involved patients with neurofibromatosis type 2 who had bilateral tumours in the cerebello-pontine angle. Only a small percentage of these ABI recipients have proven capable of identifying words. More recently, the ABI was applied to a series of non-tumour patients who had compromised cochleae or cochlear nerve aplasia, and a significant number of these patients was capable of understanding speech at a level comparable to that of most successful CI users, including conversational telephone use.[34] Although these results should be considered preliminary, in patients with ossified cochlea, severe cochlear malformation, otosclerosis and Osteogenesis Imperfecta, despite the presence of an excitable cochlear nerve, in some cases results might be better with an ABI. Better results with a CI depend on good electrode array position and number of active electrodes, which in severe cochlear ossification, otosclerosis and Osteogenesis Imperfecta might not be able to achieve. The promising results in non-tumour patients, in contrast to the patients with neurofibromatosis type 2, possibly are a reflection of the absence of cerebello-pontine or brainstem pathology.[35] Depending on future developments of the ABI system, in such cases of compromised cochleae, implantation of an ABI may be preferred over cochlear implantation.[36]

Concerning future developments in general, at present, various CI centres are conducting clinical studies in order to optimize bimodal[37] and bilateral fitting[38] of CIs and to explore the simultaneous use of acoustical and electrical stimuli in one ear in patients suffering from high frequency deafness.[39] Further challenges lie in the development of a totally implantable cochlear prosthesis[40] and the use of intracochlear nerve growth factors to block neuronal death and even lead to repair or regeneration of neurons. Future research also entails exploring auditory physiology using the CI, which in turn can lead to improved restoration of hearing with CIs. Studies of hearing in children with CIs form an ideal opportunity to explore age-related plasticity or critical periods in auditory development.[17]

7.4 Conclusions

7.4.1 Children

Congenitally, prelingually and postlingually deaf children all derived benefit from their CI for speech perception tasks, but performance varied greatly. After early implantation, the levels of performance that were eventually achieved differed much less, irrespective of whether the onset of deafness was prelingual or postlingual. In congenitally deaf children, duration of deafness played a major role in speech perception performance, whereas in children with acquired deafness, communication mode (aural-oral or sign based) was a major factor.

7.4.2 The postmeningitic ossified cochlea

Patients with partial insertion of the electrode array benefit from a CI, although to a lesser extent than patients with complete insertion.

7.4.3 The congenitally malformed cochlea

CI surgery in children with malformed inner ears may be more difficult as a result of the abnormal anatomy of the temporal bone, the possibility of an aberrant course of the facial nerve (17%), and the occurrence of cerebrospinal fluid gusher. However, the surgical procedure is considered feasible. Although the result of cochlear implantation in congenital malformation may be promising, speech perception scores vary considerately, especially in patients with severe malformations.

7.4.4 The cochlea in otosclerosis

Most of the preoperative CT scans of patients with otosclerosis referred for cochlear implantation demonstrated retrofenestral (cochlear) otosclerotic lesions, which had a tendency towards being more extensive in patients with rapidly progressive hearing loss, surgically problematic insertion of the electrode array and facial nerve stimulation. Revision surgery was mandatory in 4 patients. Facial nerve stimulation occurred in 38% of the patients.

Speech perception results showed wide variability. Better performance was related to less severe signs of otosclerosis on CT scan, full insertion of the electrode array and little or no facial nerve stimulation. The number of active electrodes appeared to be the most important determinant of the outcome.

7.4.5 The cochlea in Osteogenesis Imperfecta

When aware and prepared for the specific abnormalities of the temporal bone in Osteogenesis Imperfecta, cochlear implantation can be a safe and feasible procedure.

Preoperative imaging is recommended to be fully informed on the morphology of the petrosal bone. In case of severe deformities on the CT scan, during counselling the possibility of misplacement should be mentioned. Rehabilitation is often hindered by facial nerve stimulation requiring frequent refitting. Despite the electrophysiological changes, 2 of the 3 implanted patients had high phoneme scores.

References

1. Taitelbaum-Swead R, Kishon-Rabin L, Kaplan-Neeman R, Muchnik C, Kronenberg J, Hildesheimer M. Speech perception of children using Nucleus, Clarion or Med-El cochlear implants *Int J Pediatr Otorhinolaryngol* 2005,69 1675-83.
2. Beadle EA, McKinley DJ, Nikolopoulos TP, Brough J, O'Donoghue GM, Archbold SM. Long-Term Functional Outcomes and Academic-Occupational Status in Implanted Children After 10 to 14 Years of Cochlear Implant Use. *Otol Neurotol* 2005,26:1152-60
3. Staller S, Parkinson A, Arcaroli J, Arndt P. Pediatric outcomes with the nucleus 24 contour: North American clinical trial. *Ann Otol Rhinol Laryngol Suppl* 2002;189:56-61
4. Oh SH, Kim CS, Kang EJ, Lee DS, Lee HJ, Chang SO, Ahn SH, Hwang CH, Park HJ, Koo JW. Speech perception after cochlear implantation over a 4-year time period *Acta Otolaryngol* 2003;123:148-53.
5. Houston DM, Pisoni DB, Kirk KI, Ying EA, Miyamoto RT. Speech perception skills of deaf infants following cochlear implantation: a first report. *Int J Pediatr Otorhinolaryngol* 2003;67:479-95
6. Bassim MK, Buss E, Clark MS, Kolln KA, Pillsbury CH, Pillsbury HC, III, Buchman CA MED-EL Combi40+ cochlear implantation in adults. *Laryngoscope* 2005;115:1568-73.
7. Beynon AJ, Snik AF, van den Broek P. Evaluation of cochlear implant benefit with auditory cortical evoked potentials. *Int J Audiol* 2002;41:429-35
8 Damen GW, Beynon AJ, Krabbe PF, Mulder JJ, Mylanus EA Cochlear implantation and quality of life in postlingually deaf adults: long-term follow-up. *Otolaryngol Head Neck Surg* 2007;136.597-604.
9. Allum JH, Greisiger R, Straubhaar S, Carpenter MG Auditory perception and speech identification in children with cochlear implants tested with the EARS protocol. *Br J Audiol* 2000;34:293-303.
10 Loundon N, Busquet D, Roger G, Moatti L, Garabedian EN. Audiophonological results after cochlear implantation in 40 congenitally deaf patients: preliminary results. *Int J Pediatr Otorhinolaryngol* 2000,56:9-21.
11. El-Hakim H, Abdolell M, Mount RJ, Papsin BC, Harrison RV. Influence of age at implantation and of residual hearing on speech outcome measures after cochlear implantation. binary partitioning analysis. *Ann Otol Rhinol Laryngol Suppl* 2002;189:102-8.
12. Fryauf-Bertschy H, Tyler RS, Kelsay DM, Gantz BJ, Woodworth GG. Cochlear implant use by prelingually deafened children: the influences of age at implant and length of device use. *J Speech Lang Hear Res* 1997;40:183-99.
13. Lesinski A, Battmer RD, Bertram B, Lenarz T. Appropriate age for cochlear implantation in children--experience since 1986 with 359 implanted children. *Adv Otorhinolaryngol* 1997;52:214-7
14. Snik AF, Makhdoum MJ, Vermeulen AM, Brokx JP, van den BP. The relation between age at the time of cochlear implantation and long- term speech perception abilities in congenitally deaf subjects. *Int J Pediatr Otorhinolaryngol* 1997;41 121-31.
15. Tyler RS, Teagle HF, Kelsay DM, Gantz BJ, Woodworth GG, Parkinson AJ. Speech perception by prelingually deaf children after six years of Cochlear implant use: effects of age at implantation *Ann Otol Rhinol Laryngol Suppl* 2000;185:82-4.

16 Tomblin JB, Barker BA, Hubbs S. Developmental constraints on language development in children with cochlear implants. *Int J Audiol* 2007,46 512-23
17. Harrison RV, Gordon KA, Mount RJ. Is there a critical period for cochlear implantation in congenitally deaf children? Analyses of hearing and speech perception performance after implantation. *Dev Psychobiol* 2005;46.252-61.
18. Hoffman RA, Cohen NL. Complications of cochlear implant surgery. *Ann Otol Rhinol Laryngol Suppl* 1995;166:420-2.
19 Balkany TJ, Hodges AV, Eshraghi AA, Butts S, Bricker K, Lingvai J, Polak M, King J. Cochlear implants in children--a review. *Acta Otolaryngol* 2002,122.356-62.
20. van den Broek P, Cohen N, O'Donoghue G, Frayssc B, Laszig R, Offeciers E. Cochlear implantation in children *Int J Pediatr Otorhinolaryngol* 1995;32 Suppl:S217-23
21. Hehar SS, Nikolopoulos TP, Gibbin KP, O'Donoghue GM. Surgery and functional outcomes in deaf children receiving cochlear implants before age 2 years. *Arch Otolaryngol Head Neck Surg* 2002;128:11-4.
22. Barbara M, Mancini P, Mattioni A, Monini S, Ballantyne D, Filipo R. Residual hearing after cochlear implantation. *Adv Otorhinolaryngol* 2000;57.385-8.
23. Saeed SR, Ramsden RT, Axon PR. Cochlear implantation in the deaf-blind. *Am J Otol* 1998;19:774-7.
24. Waltzman SB, Scalchunes V, Cohen NL. Performance of multiply handicapped children using cochlear implants. *Am J Otol* 2000;21:329-35.
25. Merkus P, van Furth AM, Goverts ST, Suer M, Smits CF, Smit C. [Postmeningitis deafness in young children. action warranted before obliteration of the cochlea] *Ned Tijdschr Geneeskd* 2007;151.1209-13.
26. Eisenman DJ, Ashbaugh C, Zwolan TA, Arts HA, Telian SA. Implantation of the malformed cochlea. *Otol Neurotol* 2001;22·834-41.
27. Schmidt JM. Cochlear neuronal populations in developmental defects of the inner ear. Implications for cochlear implantation *Acta Otolaryngol* 1985;99:14-20.
28. Linthicum FH, Jr., Fayad J, Otto SR, Galey FR, House WF. Cochlear implant histopathology. *Am J Otol* 1991;12·245-311
29. Graham JM, Phelps PD, Michaels L. Congenital malformations of the ear and cochlear implantation in children· review and temporal bone report of common cavity. *J Laryngol Otol Suppl* 2000;25.1-14.
30. Eisenman DJ, Ashbaugh C, Zwolan TA, Arts HA, Telian SA. Implantation of the malformed cochlea. *Otol Neurotol* 2001;22·834-41.
31. Woolford TJ, Roberts GR, Hartley C, Ramsden RT. Etiology of hearing loss and cochlear computed tomography· findings in preimplant assessment. *Ann Otol Rhinol Laryngol Suppl* 1995;166:201-6.
32. Rotteveel LJ, Proops DW, Ramsden RT, Saeed SR, van Olphen AF, Mylanus EA. Cochlear implantation in 53 patients with otosclerosis: demographics, computed tomographic scanning, surgery, and complications. *Otol Neurotol* 2004;25 943-52.
33. Ramsden R, Bance M, Giles E, Mawman D. Cochlear implantation in otosclerosis· a unique positioning and programming problem. *J Larvngol Otol* 1997;111·262-5
34. Colletti V, Carner M, Miorelli V, Guida M, Colletti L, Fiorino F. Auditory brainstem implant (ABI): new frontiers in adults and children *Otolarvngol Head Neck Surg* 2005;133·126-38
35. Colletti V. Auditory outcomes in tumor vs. nontumor patients fitted with auditory brainstem implants *Adv Otorhinolaryngol* 2006,64:167-85
36. Colletti V, Fiorino FG, Carner M, Miorelli V, Guida M, Colletti L. Auditory brainstem implant as a salvage treatment after unsuccessful cochlear implantation. *Otol Neurotol* 2004;25:485-96.
37. Morera C, Manrique M, Ramos A, Garcia-Ibanez L, Cavalle L, Huarte A, Castillo C, Estrada E. Advantages of binaural hearing provided through bimodal stimulation via a cochlear implant and a conventional hearing aid: a 6-month comparative study. *Acta Otolaryngol* 2005;125:596-606.

38. Galvin KL, Mok M, Dowell RC. Perceptual benefit and functional outcomes for children using sequential bilateral cochlear implants. *Ear Hear* 2007;28 470-82.
39. Novak MA, Black JM, Koch DB. Standard cochlear implantation of adults with residual low-frequency hearing: implications for combined electro-acoustic stimulation *Otol Neurotol* 2007;28:609-14.
40. Cohen N. The totally implantable cochlear implant. *Ear Hear* 2007;28:100S-1S.
41. Yamagata T, Miller JM, Ulfendahl M, Olivius NP, Altschuler RA, Pyykko I, Bredberg G. Delayed neurotrophic treatment preserves nerve survival and electrophysiological responsiveness in neomycin-deafened guinea pigs *J Neurosci Res* 2004,78:75-86.

Chapter 8

Cochleaire implantatie in de gecompromitteerde cochlea

Samenvatting en conclusies

8.1 Resultaten van cochleaire implantatie

CI systemen worden tegenwoordig op grote schaal toegepast; wereldwijd zijn er meer dan 100.000 geïmplanteerde patiënten. Men zou kunnen stellen dat het CI de meest succesvolle neurale prothese is. Het blijft verbazingwekkend dat elektrische stimulatie van het pathologische auditieve systeem van een dove persoon kan leiden tot spraakherkenning De resultaten van cochleaire implantatie zijn geëvalueerd door verschillende onderzoeksgroepen met variërende methoden om de uitkomsten te beoordelen. Bij kinderen en volwassenen worden vaak spraakverstaan testen gebruikt om het resultaat van de implantatie te beoordelen.[1-6] Andere methoden zijn het meten van veranderingen van stem en articulatie, spraakproductie, ontwikkeling van de woordenschat, cognitieve en expressieve taalvaardigheden, onderwijs en beroepskeuze en algemene geletterdheid. Ook worden objectieve elektrofysiologische metingen, zoals auditief opgewekte corticale potentialen, gebruikt om de resultaten van cochleaire implantatie te onderzoeken.[7] De laatste jaren heeft het onderzoek zich bovendien gericht op de invloed van cochleaire implantatie op de kwaliteit van leven van de patiënt.[8]

Een probleem bij het testen van het spraakverstaan bij kinderen is de variatie in taalvaardigheid.[9,10] Het is gebruikelijk na implantatie halfjaarlijks of jaarlijks spraakverstaan metingen af te nemen. Op deze manier kan de mate van spraakverstaan gemeten worden als een functie van de duur van het gebruik van het implantaat. Door toenemende ervaring en ontwikkeling in spraakverstaan kunnen er tijdens de follow-up van een kind met een CI 'plafond scores' voorkomen bij het afnemen van makkelijkere testen en zogenaamde 'bodem scores' op de moeilijkere testen. Om dit probleem op te lossen, is in het CI centrum Nijmegen/Viataal een testbatterij ontwikkeld waarin de diverse aspecten van spraakverstaan worden geëvalueerd; van zeer basale vaardigheden zoals discriminatie van klinkers en vaststellen van het aantal lettergrepen in een woord (suprasegmentele identificatie) tot het verstaan van woorden en fonemen. Deze testbatterij is eerst afgenomen bij een referentie groep die bestond uit een groot aantal slechthorende kinderen die ervaren gebruikers waren van conventionele hoortoestellen en van wie het gehoorverlies varieerde van 50 tot 130 dB HL. De relatie tussen de behaalde testscores en het gehoorverlies van de referentiegroep werd gebruikt om de testscores van een kind met een CI in één enkel getal uit te drukken. Dit getal werd het gemiddelde "Equivalent Hearing Loss" genoemd (EHL). Een EHL van 80 dB HL betekent dat het kind met een CI op dat moment even goed scoort als kinderen uit de referentiegroep met een verlies van 80 dB HL met hun conventionele hoortoestellen.
Analyse van EHL waardes in **Hoofdstuk 2** toont aan dat congenitaal, prelinguaal en postlinguaal dove kinderen allen profijt hebben van hun CI bij spraakverstaan testen, maar

dat de prestaties bijzonder uiteenlopen. Gedurende de eerste twee jaren na implantatie toonden postlinguaal dove kinderen de snelste vooruitgang. Na drie jaar gebruik van het implantaat, haalden de vroeg geïmplanteerde prelinguaal en congenitaal dove kinderen (geïmplanteerd voor de leeftijd van zes jaar) de achterstand in. Prelinguaal dove kinderen die het implantaat kregen na een lange duur van doofheid presteerden minder dan diegenen die vroeg geïmplanteerd werden. Na vroege implantatie varieerden de prestaties uiteindelijk niet meer dan 10 dB, ongeacht of het ontstaan van de doofheid prelinguaal of postlinguaal was. De prestaties van congenitaal dove kinderen die na hun zesde jaar een implantaat kregen waren beduidend minder en de vooruitgang was langzamer. Bij de congenitaal dove kinderen speelde de duur van hun doofheid een kritieke rol bij de mate van spraakverstaan, terwijl bij kinderen met een verworven doofheid vooral de manier van communicatie van belang was Dus, hoe eerder een kind geïmplanteerd wordt, des te beter zal zijn of haar spraakherkenning zijn na drie jaar gebruik van het CI. Dit komt overeen met andere studieresultaten.[11-15] In de afgelopen jaren heeft de onderbouwde mening dat vroege implantatie resulteert in betere spraak-, taal- en luistervaardigheden geresulteerd in een verlaging van de gemiddelde leeftijd waarop kinderen een CI krijgen. Deze mening wordt ondersteund door onderzoeken waarin is aangetoond dat mensen beter in staat zijn om spraak en taal te leren op jonge leeftijd, dan wanneer zij ouder zijn; er is een specifiek moment in de ontwikkeling, genaamd *kritische periode* of *sensitieve periode*, waarin men spraak en taal het meest efficiënt leert. De *sensitieve periode* wordt veelal gedefinieerd als een geleidelijke tijd in de ontwikkeling waarin het organisme uitzonderlijk gevoelig is voor bepaalde ervaringen. Deze gevoeligheid is gebaseerd op de zogenaamde 'leeftijdsafhankelijke plasticiteit'. De *kritische periode* daarentegen wordt gezien als een vastomlijnde tijdspanne in de ontwikkeling, waarin ervaringen, of het gebrek hieraan, resulteren in een compleet onomkeerbare verandering in de hersenen.[16] Hoewel dit niet wetenschappelijk gestoeld is, gebruiken wij bij voorkeur de term *sensitieve periode*.

Dierexperimenten hebben reeds aangetoond dat het auditieve systeem een hoge mate van plasticiteit bezit.[17] Bij de mens uit deze plasticiteit zich in het vermogen van het auditieve system zich aan te passen aan de nieuwe, door het CI aangeboden stimuli. Dat veel patiënten tot een goed spraakverstaan kunnen komen, duidt op een goed aanpassingsvermogen van het auditieve systeem. Dit vermogen kan na een langdurige auditieve deprivatie afnemen, waardoor de resultaten van cochleaire implantatie afhankelijk zijn van de leeftijd waarop de implantatie plaatsvindt. Uit onze data kunnen we afleiden dat de sensitieve periode rond het 6de levensjaar eindigt. Een scherpere bovenste leeftijdsgrens of periode waarin implantatie het meest succesvol is heeft eerder onderzoek niet aan kunnen tonen.[17]

Door de conservatieve inclusie criteria voor cochleaire implantatie in het verleden, de ontwikkeling van steeds betere CI systemen en de toegenomen expertise van de CI teams

zijn de data gepresenteerd in Hoofdstuk 2 niet representatief voor de huidige paediatrische CI populatie Echter, de conservatieve inclusie criteria van die tijd hebben geresulteerd in een behoorlijk homogene groep geïmplanteerde kinderen met vergelijkbare intelligentie en mate van steun geboden door de ouders De manier van communiceren werd niet aan het individuele kind aangepast maar was afhankelijk van de school waarop het kind geplaatst was, het betrof dan een voornamelijk oraal-aurale communicatie of een voornamelijk op gebarentaal gebaseerde communicatie Deze homogeniteit biedt ons de mogelijkheid om lange termijn resultaten van deze groep kinderen op een aantal variabelen te onderzoeken De meer recent geïmplanteerde kinderen vormen door het versoepelen van de inclusie criteria een meer heterogene groep met uiteenlopende intelligentie, steun van ouders en schoolplaatsing waardoor het effect van specifieke variabelen op het spraakverstaan niet meer betrouwbaar onderzocht kunnen worden

8.2 Cochleaire implantatie in de gecompromitteerde cochlea

Sinds de voordelen van cochleaire implantatie voor dove patiënten die geen baat hebben van een conventioneel hoortoestel duidelijk zijn aangetoond, zijn de indicaties voor cochleaire implantatie flink uitgebreid Tegenwoordig worden ook patiënten met morfologische veranderde, gecompromitteerde cochleae geïmplanteerd, zoals het geval is in congenitale malformaties van het binnenoor, geossificeerde cochleae na meningitis en vergevorderde otosclerose Het gevolg hiervan is dat de chirurgische procedure ingewikkelder is geworden en revisie chirurgie vaker nodig is gebleken [18] Ook het feit dat er patiënten op heel jonge en hoge leeftijd,[19-21] met restgehoor[22] en meerdere handicaps[23 24] worden geopereerd heeft consequenties voor de chirurgische procedure Deze ontwikkeling heeft tot gemodificeerde implantaten en chirurgische technieken geleid De incidentie van complicaties van de operatie is nog verder gedaald Niet alleen deze verbeterde technieken hebben bijgedragen aan de steeds toenemende resultaten van patiënten met een CI, maar ook de karakteristieken van de huidige CI patiënt zoals een kortere duur van doofheid door snellere implantatie, aanwezigheid van restgehoor en een jongere leeftijd

Het is van belang te rapporteren over de moeilijke chirurgische casus en de voorgekomen complicaties te bespreken zodat deskundigen van elkaars ervaringen kunnen leren Zo kan een chirurg voorbereid zijn op bijzondere omstandigheden, die ook preoperatief met de patiënt en zijn/haar familie besproken moeten worden De incidentie van patiënten met ernstig gecompromitteerde cochleae is laag, zodat aanbevolen wordt de implantatie procedure plaats te laten vinden in ervaren CI centra

8.2.1 Cochleaire implantatie in de gecompromitteerde cochlea van het kind

In **Hoofdstuk 3** worden de resultaten beschreven van 7 kinderen, doof geworden na meningitis, met partiële insertie van de Nucleus elektrode array ten gevolge van ossificatie van de cochlea, en van 18 kinderen doof geworden na meningitis met een volledige insertie van de elektrode array in de cochlea. Bij 10 kinderen was er sprake van ossificatie op de CT scan die ook tijdens de operatie gevonden werd (sensitiviteit 53%). Bij 9 kinderen echter was er geen ossificatie zichtbaar op de CT scan, terwijl die tijdens de operatie wel gevonden werd (fout negatieve ratio 47%); ondanks een normale cochlea op de CT scan van een kind met postmeningitis doofheid moet men dus bedacht zijn op het aantreffen van ossificatie, aanvullend onderzoek in de vorm van een MRI scan is in deze gevallen aangewezen.

Beide groepen kinderen werden met dezelfde testbatterij getest die gereduceerd kan worden in een EHL waarde zoals eerder beschreven. Drie jaar na implantatie hadden de kinderen met partiële insertie een tragere vooruitgang en bereikten ze een minder goede plateau score. Patiënten met partiële insertie hebben profijt van een CI, maar minder dan patiënten met volledige insertie van de elektrode array. Dit pleit voor een snelle implantatie van kinderen die doof geworden zijn door meningitis, nog voor ossificatie plaats vindt. Tegenwoordig is er nationale consensus tussen KNO-artsen en kinderartsen over het vroeg uitvoeren van audiometrie bij kinderen met bacteriële meningitis en in geval van hoorverlies doorverwijzen naar een KNO-arts.[25]

Naast het gereduceerde aantal elektrodes zijn er nog andere factoren die een rol kunnen spelen in de matigere prestaties van patiënten met partiële insertie ten gevolge van ossificatie; mogelijk hebben ook een suboptimale afstand tussen de elektrode array en de modiolus en een meer verspreidde elektrische stroom in de uitgeboorde tunnel een negatieve invloed.

Aangezien cochleaire malformaties geassocieerd zijn met een afgenomen hoeveelheid spirale ganglion cellen en een vaak complexere chirurgische procedure[26] wordt van patiënten met ernstige malformaties van het binnenoor een slechter resultaat van de cochleaire implantatie verwacht dan van patiënten met normaal ontwikkelde cochleae. De neurale elementen zijn vaak onderontwikkeld. Schmidt[27] vond een gemiddelde hoeveelheid spirale ganglioncellen van 11,500 bij dysplasie van Mondini, vergeleken met rond de 20,000 spirale ganglioncellen bij otosclerose en ototoxiciteit, en ongeveer 33,000 bij normaal horende personen. Gelukkig is uit studies van het os temporale gebleken dat slechts 3300 ganglion cellen nodig zijn voor een goed resultaat van cochleaire implantatie.[28] Om de chirurgische aspecten en resultaten van cochleaire implantatie van kinderen met cochleaire malformaties te onderzoeken zijn in **Hoofdstuk 4** de klinische en audiometrische gegevens beschreven van 13 patientjes met variërende binnenoor

malformaties. Bij deze 13 patientjes samen met de in de literatuur gerapporteerde geïmplanteerde kinderen met cochleaire malformaties kwam een afwijkend verloop van de nervus facialis bij 17% van de kinderen voor. Wanneer alleen gekeken wordt naar kinderen met ernstige cochleaire malformaties zoals de 'common cavity' of ernstige cochleaire hypoplasie komt een aberrant verloop van de nervus facialis zelfs bij 27% voor.
Bij alle 13 kinderen kon de elektrode array volledig geïnsereerd worden in de cochlea. Na 1 jaar follow-up, waren de meeste kinderen in staat deel te nemen aan open spraakverstaan testen. Enkelen echter hadden slechts een beperkte taalvaardigheid en nog geen open spraakverstaan, mogelijk door de jonge leeftijd, een lange duur van doofheid of de korte follow-up. Zij konden echter wel deelnemen aan gesloten spraakverstaan testen, en toonden tenminste een toegenomen gewaarwording van omgevingsgeluiden.
In het algemeen kan bij patiënten met milde cochleaire malformaties een volledige insertie bereikt worden en zijn de resultaten vergelijkbaar met patiënten met normale cochleae[29]; patiënten met ernstige cochleaire malformaties daarentegen presteren naar verwachting minder goed dan patiënten met normaal gevormde cochleae doordat er bij hen vaak sprake is van een verminderd aantal ganglioncellen, recidiverende meningitiden en een moeizamere chirurgische procedure.[30] Hoewel de resultaten van cochleaire implantatie veelbelovend zijn is het tijdens de preoperatieve counseling van een patiënt met een cochleaire malformatie van belang het kind en zijn ouders goed te informeren over de onzekere resultaten van met name de ernstige malformaties

8.2.2 Cochleaire implantatie in de gecompromitteerde cochlea van de volwassene

Zeven tot 9.5% van de volwassen CI populatie is doof geworden ten gevolge van otosclerose.[31] De databases van 4 CI centra leverden 53 patiënten met otosclerose op die beschreven zijn in **Hoofdstuk 5**. De meerderheid van deze patiënten had afwijkingen op de CT scan, zoals retrofenestrale (cochleaire) otosclerotische haarden. De CT afwijkingen waren uitgebreider bij patiënten met een snel progressieve slechthorend, een moeizamere chirurgische procedure met een problematisch verlopen insertie van de elektrode array en tijdens de revalidatie nervus facialis stimulatie door activatie van het implantaat. Bij 4 patiënten was revisie chirurgie noodzakelijk geweest. Bij een groot aantal patiënten was sprake van nervus facialis stimulatie (38%) dat meestal door de distale elektrodes werd veroorzaakt.
De spraakverstaan scores varieerden in hoge mate. Vergeleken met de patiënten met goede scores, verschilden de patiënten met matige scores niet qua leeftijd van ontstaan van slechthorendheid, duur van de slechthorendheid, mate van progressie, leeftijd van ontstaan van doofheid of duur van de doofheid. Een goed spraakverstaan was gerelateerd aan minder afwijkingen op de CT scan, volledige insertie van de elektrode array en geen tot nauwelijks optredende nervus facialis stimulatie. Het aantal actieve elektrodes, dat indirect

aan de ziekte is gerelateerd, bleek de meest bepalende factor voor het resultaat met de CI. Dit is komt overeen met de resultaten beschreven in Hoofdstuk 3 waarin de groep met volledige insertie significant beter scoorde dan de groep met partiële insertie.

In **Hoofdstuk 6** worden de resultaten beschreven van de cochleaire implantatie van 3 patiënten met ernstige perceptieve slechthorendheid ten gevolge van Osteogenesis Imperfecta. De diagnose Osteogenesis Imperfecta was gesteld op basis van de klinische bevindingen waarbij ook de beeldvorming van alle 3 de patiënten overeenkwam met de diagnose.
De typische bevindingen tijdens oorchirurgie van patiënten met Osteogenesis Imperfecta, zoals een broos scutum, een sclerotisch verdikte cochlea en een hyperplastische middenoor mucosa met persisterend bloeden werden ook bij deze patiënten aangetroffen. Bij Casus 3, waarvan de CT van het os petrosum ernstige afwijkingen had laten zien, kwam de elektrode in het horizontale semicirculaire kanaal terecht. Dat de elektrode array buiten de cochlea kan 'doorschieten' in een otospongiotisch os petrosum is ook al beschreven bij patiënten met otosclerose.[32,33]
Bij alle 3 de patiënten werd de revalidatie gehinderd door het opwekken van non-auditieve sensaties Zelfs na reïmplantatie leidden deze sensaties in Casus 3 tot het uiteindelijk staken van gebruik van het implantaat. Bij Casus 3 werden afwijkende 'averaged electrode voltages (AEVs)' gevonden die passen bij een abnormale geleiding van het bot. De 'Spatial spread of neural excitation responses' van Casus 1 en 2 duidden op interactie tussen intracochleaire kanalen, die vaak samen voorkwam met nervus facialis stimulatie. De geschatte toonhoogte van Casus 1 varieerde met name voor de elektrodes die ook nervus facialis stimulatie veroorzaakten. Desondanks behaalden Casus 1 en 2 na 1 jaar follow-up goede foneem scores van respectievelijk 81% en 78%.

8.3 Toekomstig onderzoek

De afgelopen 2 decades heeft onderzoek van verschillende disciplines zoals artsen, natuurkundigen en wetenschappers in het veld van de cochleaire implantatie geleid tot de ontwikkeling van nieuw vormgegeven implantaten en elektrode arrays, gespecialiseerde software en verminderd stroomverbruik. Voorts hebben verfijningen van spraak coderings algorithmes geleid tot een grote vooruitgang in de spraakverstaan scores van CI patiënten. Met verder onderzoek wordt naar een nog hogere resolutie gestreefd zonder toename in stroom verbruik. Elektrische stimulatie die meer gericht en minder gespreid is kan nervus facialis stimulatie verminderen, dat vooral voor patiënten met otosclerose van belang is.

Zoals eerder gemeld, kan het CI beschouwd worden als de meest succesvolle neurale prothese Het hersenstam implantaat (auditory brainstem implant, ABI) is een modificatie van het CI voor patienten die door de aanwezigheid van ernstig gecompromitteerde cochleae of disfunctionele nervus cochlearis niet geimplanteerd kunnen worden met een CI De elektrodes van het ABI worden direct tegen de hersenstam geplaatst Aanvankelijk werden ABI's geplaatst bij patienten met neurofibromatose type 2, die bilateraal tumoren in de brughoek hadden Slechts een klein gedeelde van deze patienten was in staat woorden te herkennen Recent zijn ABI's geimplanteerd in een serie patienten zonder tumoren maar met gecompromitteerde cochleae of aplasie van de nervus cochlearis Een significant gedeelte van deze patienten bleek wel degelijk in staat tot spraakverstaan op een niveau gelijkwaardig aan dat van de meeste succesvolle CI patienten, waaronder het gebruik van de telefoon [34] Hoewel dit voorlopige resultaten zijn, kunnen bij patienten met geossificeerde cochleae, vergevorderde otosclerose en Osteogenesis Imperfecta in sommige gevallen ondanks de aanwezigheid van een normaal stimuleerbare nervus cochlearis de resultaten beter zijn met een ABI Goede resultaten met een CI hangen af van een goede positie van de elektrode array en een groot aantal actieve elektrodes, hetgeen bij ernstige cochleaire ossificatie, otosclerose en Osteogenesis Imperfecta soms niet bereikt kan worden De veelbelovende resultaten van hersenstam implantatie bij patienten zonder tumoren, in tegenstelling tot die van patienten met neurofibromatose type 2, worden mogelijk verklaard door de afwezigheid van pathologie in de brughoek of hersenstam [35] Afhankelijk van de toekomstige ontwikkelingen van hersenstam implantatie kan in voornoemde gevallen van gecompromitteerde cochleae een ABI de voorkeur hebben boven cochleaire implantatie [36]

Ontwikkelingen op andere gebieden door diverse CI teams beogen het optimaliseren van bimodale[37] en bilaterale[38] aanpassingen van cochleaire implantaten Ook worden de mogelijkheden onderzocht voor simultaan gebruik van zowel akoestische als elektrische stimulatie van één oor bij patienten met hoge tonen verlies [39] Naast het verbeteren van de resultaten van cochleaire implantatie bij patienten met steeds uitgebreidere indicaties liggen er nog volop wetenschappelijke uitdagingen in het verschiet, zoals de ontwikkeling van een volledig implanteerbare cochleaire prothese[40] en het gebruik van intracochleaire neuronale groeifactoren om verdere neuronen verval te blokkeren en eventueel zelfs te leiden tot herstel ervan [41] Het CI leent zich ook uitstekend voor de exploratie van het auditieve systeem, hetgeen weer kan leiden tot een verbeterd herstel van gehoor met behulp van CIs Verder bieden studies naar het gehoor van kinderen met een CI een ideale mogelijkheid om de leeftijdsgebonden plasticiteit of sensitieve periode in de auditieve ontwikkeling te onderzoeken [17]

8.4 Conclusies

8.4.1 Kinderen

Zowel congenitaal, als prelinguaal en postlinguaal dove kinderen hebben profijt van een CI, hoewel de mate van profijt erg kan variëren. Na implantatie op jonge leeftijd behaalden de kinderen echter vergelijkbare scores, waarbij dit onafhankelijk was van een prelinguaal of postlinguaal ontstaan van de doofheid. Bij de congenitaal dove kinderen speelde met name de duur van de doofheid een belangrijke rol in de mate van spraakverstaan met CI, terwijl bij de kinderen met verworven doofheid vooral de manier van communiceren (orale versus gebaren taal) een rol speelde.

8.4.2 De cochlea met ossificatie na meningitis

Patiënten met partiële insertie van de elektrode array hebben profijt van een CI, hoewel minder dan patiënten met een volledige insertie van de elektrode array.

8.4.3 De cochlea met congenitale malformatie

De chirurgische implantatie procedure kan bij kinderen met congenitale malformaties van het binnenoor bemoeilijkt worden door de abnormale anatomie van het os petrosum, een mogelijk aberrant verloop van de nervus facialis (17%) en het voorkomen van liquor Gusher. Desondanks is cochleaire implantatie zeker haalbaar. De resultaten die patiënten met congenitale malformaties van het binnenoor behalen met een CI zijn veelbelovend, hoewel de spraakverstaan scores met name bij patiënten met de ernstigere malformaties erg variëren.

8.4.4 De cochlea met otosclerose

Het merendeel van de preoperatieve CT scans van patiënten met otosclerose die verwezen waren voor cochleaire implantatie liet aanwijzingen zien voor retrofenestrale (cochleaire) otosclerose. Deze afwijkingen waren veelal uitgebreider bij patiënten die een snel progressief verloop van het hoorverlies hadden, een problematischer verlopen implantatie procedure hadden met moeizame insertie van de electrode array en postoperatief nervus facialis stimulatie ondervonden door activatie van het CI. Revisie chirurgie was noodzakelijk bij 4 van de 53 patiënten. Nervus facialis stimulatie kwam bij 38% van de otosclerose patiënten voor.

Er was een grote variatie in spraakverstaan scores. Een beter resultaat was geassocieerd met minder afwijkingen op de CT scan, volledige insertie van de elektrode array en afwezigheid van nervus facialis stimulatie. De meest bepalende factor met betrekking tot het eindresultaat bleek het aantal actieve elektrodes te zijn.

8.4.5 De cochlea met Osteogenesis Imperfecta

Wanneer men bewust is van de specifieke veranderingen van het os temporale van patiënten met Osteogenesis Imperfecta en hierop voorbereid is, kan de chirurgische implantatie veilig en haalbaar zijn. Met behulp van beeldvorming kan men preoperatief goed geïnformeerd worden over de morfologie van het os petrosum. Indien ernstige afwijkingen op de CT scan te zien zijn, moet tijdens de preoperatieve counseling de mogelijkheid besproken worden dat de elektrode array buiten de cochlea kan 'doorschieten' in het otospongiotische os petrosum.

De revalidatie wordt vaak gehinderd door het voorkomen van nervus facialis stimulatie waardoor regelmatig afstellen van de spraakprocessor noodzakelijk kan zijn. Twee van de 3 geïmplanteerde patiënten met Osteogenesis Imperfecta hadden, ondanks de gemeten elektrofysiologische veranderingen, hoge spraakverstaan scores.

Referenties

1. Taitelbaum-Swead R, Kishon-Rabin L, Kaplan-Neeman R, Muchnik C, Kronenberg J, Hildesheimer M. Speech perception of children using Nucleus, Clarion or Med-El cochlear implants. *Int J Pediatr Otorhinolaryngol* 2005;69:1675-83
2. Beadle EA, McKinley DJ, Nikolopoulos TP, Brough J, O'Donoghue GM, Archbold SM. Long-Term Functional Outcomes and Academic-Occupational Status in Implanted Children After 10 to 14 Years of Cochlear Implant Use. *Otol Neurotol* 2005,26:1152-60.
3. Staller S, Parkinson A, Arcaroli J, Arndt P Pediatric outcomes with the nucleus 24 contour. North American clinical trial. *Ann Otol Rhinol Laryngol Suppl* 2002,189:56-61.
4. Oh SH, Kim CS, Kang EJ, Lee DS, Lee HJ, Chang SO, Ahn SH, Hwang CH, Park HJ, Koo JW Speech perception after cochlear implantation over a 4-year time period. *Acta Otolaryngol* 2003;123 148-53
5. Houston DM, Pisoni DB, Kirk KI, Ying EA, Miyamoto RT. Speech perception skills of deaf infants following cochlear implantation a first report. *Int J Pediatr Otorhinolaryngol* 2003;67:479-95.
6. Bassim MK, Buss E, Clark MS, Kolln KA, Pillsbury CH, Pillsbury HC, III, Buchman CA. MED-EL Combi40+ cochlear implantation in adults *Laryngoscope* 2005;115.1568-73
7. Beynon AJ, Snik AF, van den BP. Evaluation of cochlear implant benefit with auditory cortical evoked potentials. *Int J Audiol* 2002;41:429-35.
8. Damen GW, Beynon AJ, Krabbe PF, Mulder JJ, Mylanus EA. Cochlear implantation and quality of life in postlingually deaf adults: long-term follow-up. *Otolaryngol Head Neck Surg* 2007;136.597-604.
9. Allum JH, Greisiger R, Straubhaar S, Carpenter MG. Auditory perception and speech identification in children with cochlear implants tested with the EARS protocol. *Br J Audiol* 2000;34 293-303.
10. Loundon N, Busquet D, Roger G, Moatti L, Garabedian EN. Audiophonological results after cochlear implantation in 40 congenitally deaf patients: preliminary results. *Int J Pediatr Otorhinolaryngol* 2000;56:9-21.
11. El-Hakim H, Abdolell M, Mount RJ, Papsin BC, Harrison RV. Influence of age at implantation and of residual hearing on speech outcome measures after cochlear implantation. binary partitioning analysis. *Ann Otol Rhinol Laryngol Suppl* 2002;189:102-8.
12 Fryauf-Bertschy H, Tyler RS, Kelsay DM, Gantz BJ, Woodworth GG. Cochlear implant use by prelingually deafened children· the influences of age at implant and length of device use. *J Speech Lang Hear Res* 1997;40:183-99.

13. Lesinski A, Battmer RD, Bertram B, Lenarz T Appropriate age for cochlear implantation in children--experience since 1986 with 359 implanted children *Adv Otorhinolaryngol* 1997;52:214-7.
14. Snik AF, Makhdoum MJ, Vermeulen AM, Brokx JP, van den Broek P. The relation between age at the time of cochlear implantation and long- term speech perception abilities in congenitally deaf subjects. *Int J Pediatr Otorhinolaryngol* 1997,41·121-31
15. Tyler RS, Teagle HF, Kelsay DM, Gantz BJ, Woodworth GG, Parkinson AJ. Speech perception by prelingually deaf children after six years of Cochlear implant use: effects of age at implantation. *Ann Otol Rhinol Laryngol Suppl* 2000,185.82-4.
16. Tomblin JB, Barker BA, Hubbs S. Developmental constraints on language development in children with cochlear implants. *Int J Audiol* 2007;46:512-23.
17. Harrison RV, Gordon KA, Mount RJ. Is there a critical period for cochlear implantation in congenitally deaf children? Analyses of hearing and speech perception performance after implantation. *Dev Psychobiol* 2005;46:252-61.
18. Hoffman RA, Cohen NL Complications of cochlear implant surgery *Ann Otol Rhinol Laryngol Suppl* 1995;166:420-2.
19. Balkany TJ, Hodges AV, Eshraghi AA, Butts S, Bricker K, Lingvai J, Polak M, King J Cochlear implants in children--a review. *Acta Otolaryngol* 2002;122·356-62
20. van den Broek P, Cohen N, O'Donoghue G, Frayssc B, Laszig R, Offeciers E Cochlear implantation in children. *Int J Pediatr Otorhinolaryngol* 1995;32 Suppl·S217-S223.
21. Hehar SS, Nikolopoulos TP, Gibbin KP, O'Donoghue GM Surgery and functional outcomes in deaf children receiving cochlear implants before age 2 years. *Arch Otolaryngol Head Neck Surg* 2002;128·11-4
22 Barbara M, Mancini P, Mattioni A, Monini S, Ballantyne D, Filipo R. Residual hearing after cochlear implantation. *Adv Otorhinolaryngol* 2000,57:385-8.
23. Saeed SR, Ramsden RT, Axon PR Cochlear implantation in the deaf-blind. *Am J Otol* 1998;19:774-7.
24. Waltzman SB, Scalchunes V, Cohen NL. Performance of multiply handicapped children using cochlear implants. *Am J Otol* 2000;21.329-35
25. Merkus P, van Furth AM, Goverts ST, Suer M, Smits CF, Smit C. Postmeningitis deafness in young children: action warranted before obliteration of the cochlea *Ned Tijdschr Geneeskd* 2007;151:1209-13
26. Eisenman DJ, Ashbaugh C, Zwolan TA, Arts HA, Telian SA Implantation of the malformed cochlea. *Otol Neurotol* 2001;22·834-41
27. Schmidt JM Cochlear neuronal populations in developmental defects of the inner ear. Implications for cochlear implantation. *Acta Otolaryngol* 1985;99·14-20.
28. Linthicum FH, Jr, Fayad J, Otto SR, Galey FR, House WF Cochlear implant histopathology *Am J Otol* 1991,12·245-311
29. Graham JM, Phelps PD, Michaels L Congenital malformations of the ear and cochlear implantation in children: review and temporal bone report of common cavity. *J Laryngol Otol Suppl* 2000;25·1-14
30. Eisenman DJ, Ashbaugh C, Zwolan TA, Arts HA, Telian SA. Implantation of the malformed cochlea. *Otol Neurotol* 2001;22.834-41.
31. Woolford TJ, Roberts GR, Hartley C, Ramsden RT. Etiology of hearing loss and cochlear computed tomography: findings in preimplant assessment. *Ann Otol Rhinol Laryngol Suppl* 1995;166:201-6.
32. Rotteveel LJ, Proops DW, Ramsden RT, Saeed SR, van Olphen AF, Mylanus EA. Cochlear implantation in 53 patients with otosclerosis: demographics, computed tomographic scanning, surgery, and complications. *Otol Neurotol* 2004,25 943-52.
33. Ramsden R, Bance M, Giles E, Mawman D. Cochlear implantation in otosclerosis· a unique positioning and programming problem *J Laryngol Otol* 1997,111 262-5.
34. Colletti V, Carner M, Miorelli V, Guida M, Colletti L, Fiorino F. Auditory brainstem implant (ABI): new frontiers in adults and children *Otolaryngol Head Neck Surg* 2005;133:126-38.

35 Colletti V Auditory outcomes in tumor vs nontumor patients fitted with auditory brainstem implants *Adv Otorhinolaryngol* 2006,64 167-85
36 Colletti V, Fiorino FG, Carner M, Miorelli V, Guida M, Colletti L Auditory brainstem implant as a salvage treatment after unsuccessful cochlear implantation *Otol Neurotol* 2004,25 485-96
37 Morera C, Manrique M, Ramos A, Garcia-Ibanez L, Cavalle L, Huarte A, Castillo C, Estrada E Advantages of binaural hearing provided through bimodal stimulation via a cochlear implant and a conventional hearing aid a 6-month comparative study *Acta Otolaryngol* 2005,125 596-606
38. Galvin KL, Mok M, Dowell RC Perceptual benefit and functional outcomes for children using sequential bilateral cochlear implants *Ear Hear* 2007,28.470-82
39 Novak MA, Black JM, Koch DB Standard cochlear implantation of adults with residual low-frequency hearing implications for combined electro-acoustic stimulation *Otol Neurotol* 2007,28.609-14.
40 Cohen N The totally implantable cochlear implant *Ear Hear* 2007;28 100S-1S
41 Yamagata T, Miller JM, Ulfendahl M, Olivius NP, Altschuler RA, Pyykko I, Bredberg G Delayed neurotrophic treatment preserves nerve survival and electrophysiological responsiveness in neomycin-deafened guinea pigs *J Neurosci Res* 2004,78 75-86

Dankwoord

Dit proefschrift is tot stand gekomen door de medewerking van verscheidene professionals en ook betrokken niet-professionals, waar ik in de afgelopen jaren graag gebruik van heb gemaakt en die ik hierbij voor hun bijdrage wil bedanken.

Prof. dr. ir. A.F.M. Snik, beste Ad, het was erg prettig met je samen te werken. De rust die je uitstraalt en de altijd nuttige adviezen waren zeer welkom, net als het feit dat je nooit over deadlines bent begonnen. Bij jou kon ik terecht met mijn (verschillende) statistische berekeningen, waarbij je A4tjes vol hebt getekend met tabellen, formules en klokken. Als je in mijn ellenlange lappen tekst weer de helft had weggestreept, werd het inderdaad leesbaarder, hoewel menig editor de stukken dan nog te lang vond. Eén en ander heeft geresulteerd in dit, naar ik mag zeggen, best fraaie boekje! Dank daarvoor!!

Prof. dr. C.W.R.J. Cremers, om in uw metaforen te spreken: u was er om mijn onderzoek op de rails te zetten en vorm te geven, en toen het eindstation naderde was u er weer om het veilig binnen te loodsen. Uw uitgebreide ervaring als promotor heeft mij zeer geholpen.

Dr. E.A.M. Mylanus, beste Emmanuel, jouw energie lijkt onuitputtelijk en werkt aanstekelijk! Je professionele opmerkingen bij de verschillende artikelen en het feit dat je altijd bereid was tot overleg, maakten het er zeker een stuk gemakkelijker op. Dat je ook altijd in bent voor een biertje, maakte het er daarnaast ook nog eens gezelliger op!

Diverse leden van het Nijmegen/Vlaataal CI team hebben geholpen met de dataverzameling en gaven enthousiast de benodigde uitleg, waarbij ik speciaal wil bedanken Lucas Mens, Andy Beynon, AnnekeVermeulen, Esther Dekkers, Wendy Huinck en Rens Leeuw.

Prof. R.T. Ramsden and CI team Manchester, Mr. D.W. Proops and CI team Birmingham, thank you for giving me the opportunity to collect data from your CI centres: I have enjoyed our cooperation very much and am proud of the resulting two papers on cochlear implantation in patients with otosclerosis.

Dankzij de medewerking van Dr. A.F. van Olphen hadden we beschikking over de data van de Utrechtse CI patiënten met otosclerose: Diewertje Plancken bedankt voor je vriendelijke ontvangst en je hulp met de Utrechtse database.

Diny Helsper. Voor een digibeet is hulp bij de lay-out onontbeerlijk (al delen we denk ik inmiddels allebei een aversie tegen reference manager), hartelijk dank hiervoor.

Prof dr K Graamans en stafleden van de afdeling KNO Tijdens de opleiding worden omstandigheden gecreeerd die het promotie onderzoek van ons arts-assistenten mogelijk maakt zelfs mensen met wat minder discipline en meer perfectionisme lukt het zo om de eindstreep te halen!

(Oud-) arts-assistenten KNO Het is al vele malen gezegd maar het blijft ook nu met 'de nieuwe generatie'een feit we hebben een gezellige en hechte groep collega's waardoor het niet alleen goed samenwerken is, maar er binnen en buiten het ziekenhuis ook de nodige lol te beleven valt Vroeg opstaan is niet leuk, maar als om 8 uur de eerste grappen alweer worden gemaakt, kan de dag weer beginnen!

Vrienden en vriendinnetjes Ik heb maar zelden een etentje, shop-date, avondje stappen of weekendje-weg laten schieten voor mijn onderzoek ik had ze ook niet willen missen! Ik hoop dat er in de toekomst nog vele zullen volgen, maar dan zonder schuldgevoel!

Paranimfen Annemieke de Greef en Caroline Andeweg Miek, lieve zus en vriendin, eigenlijk delen we alles, dus ook deze promotie Caro, geneeskunde vriendinnetje van het eerste uur en vaste vakantie partner in onze studententijd, gelukkig bleef jij voor je opleiding Chirurgie ook in Nijmegen waardoor we als vanzelfsprekend onze vriendschap konden voortzetten Ik vind jullie een goed team aan mijn zijde op 9 juni!

Familie Miek & Jasper, Joris & Dorien, het 'front', ik had me geen betere broer en zus kunnen wensen, en ook met 'de aanhang' vormen we een hecht geheel Dank voor alle steun Lieve Omi, bijna 90 en nooit vergeet u te informeren naar mijn werk en mijn onderzoek, ik ben er trots op dat u mijn oma bent! Lieve papa en mama, het valt niet in woorden uit te drukken hoe waardevol ik jullie onvoorwaardelijke steun en liefde vind Bij jullie vind ik altijd een warm en gastvrij thuis, waarbij ik ook op het professionele vlak mijn hart kan luchten Papa krijgt het toch altijd weer voor elkaar dat ik de zaken weer nuchter kan bekijken, hetgeen ook mijn bloeddruk ten goede komt Zonder jullie was ik niet waar ik nu ben

Lieve Vincent, Viçenti, je belofte om als pimpnimf op te treden (in plaats van paranimf) heb je ruimschoots waargemaakt de kaft van dit boekje heb je super gepimpt!
Een weekendrelatie en het laatste jaar van een promotie, dat is geen gelukkige combinatie gelukkig zijn wij dat wel! Ik bedenk me steeds weer, jij *past* goed bij mij

Curriculum Vitae

Liselotte Rotteveel werd op 16 februari 1976 geboren te Nijmegen. In 1994 behaalde zij het VWO diploma aan het Stedelijk Gymnasium te Nijmegen en startte de studie Geneeskunde aan de Radboud Universiteit Nijmegen. In het kader van een uitwisselingsprogramma voerde zij haar eerste coassistentschap uit op de afdeling Gynaecologie & Obstetrie in het Assaf Harofeh Medical Centre, Tel Aviv, Israel. Haar wetenschappelijke stage aan het eind van haar coschappen verliep onder begeleiding van de afdelingen revalidatie geneeskunde en Keel-, Neus- en Oorheelkunde van de Radboud Universiteit Nijmegen, Medisch Centrum en was onderdeel van het promotie onderzoek van Dr. P.H. Jongerius. In 2001 behaalde zij het artsexamen waarna ze 9 maanden werkzaam was als poortarts op de Spoedeisende Hulp van het Carolus Liduina Ziekenhuis te 's-Hertogenbosch. Nadat zij in april en mei 2002 korte tijd als AGNIO voor de chirurgische discipline in het Antoni van Leeuwenhoek Ziekenhuis te Amsterdam werkzaam was geweest, werd zij aangesteld als arts-onderzoeker op de afdeling Keel-, Neus- en Oorheelkunde van de Radboud Universiteit Nijmegen, Medisch Centrum voor het onderzoek dat ten grondslag ligt aan dit proefschrift, onder leiding van Prof. dr. ir. A.F.M. Snik en Prof. dr. C.W.R.J. Cremers. Sinds januari 2004 is zij in opleiding tot Keel-Neus- en Oorarts aan Radboud Universiteit Nijmegen, Medisch Centrum (opleider Prof. dr. K. Graamans)

Abbreviations

ABI	auditory brainstem implant
AB test	open-set Arthur Boothroyd monosyllables test: phoneme scores
AN test	Antwerpen Nijmegen test: word scores
AEVs	Averaged Electrode Voltages
BKB test	Bamford-Kowal-Bench sentences test
BEAR	Brainstem Evoked Auditory Response
CI	Cochlear Implant
C level	Most Comfortable Loudness level
CSF	Cerebro Spinal Fluid
CT	Computed Tomography
CVC test	Consonant-Vowel-Consonant test
dB	Decibel
ECAPs	evoked compound action potentials
ECoG	Electrocochleography
EHL	Equivalent Hearing Loss
ENT	Ear Nose and Throat
FNS	Facial Nerve Stimulation
GN test	Gestel Nijmegen speech perception test
HL	Hearing Level
HRCT	High Resolution Computed tomography
NCIQ	Nijmegen Cochlear Implant Questionnaire
NRT	Neural Response Telemetry
NVA test	'Nederlandse Vereniging van Audiologie' open monosyllable tests
OI	Osteogenesis Imperfecta
PTA	Pure Tone Average (average hearing loss at 0.5, 1 and 2 kHz)
SPL	Sound Pressure Level
SRT	Speech Reception Threshold
T level	Behavioral Threshold level
U level	Uncomfortable Loudness level

List of publications

Rotteveel LJC, Jongenius PH, van Limbeek J, van den Hoogen FJA Salivation in healthy schoolchildren *Int J Peadiatr Otorhinolaryngol* 2004,68 767-774

Mylanus EAM, Rotteveel LJC, RL Leeuw RL Congenital Malformation of the Inner Ear and Pediatric Cochlear Implantation *Otol Neurotol* 2004,25 308-317

Rotteveel LJC, Proops DW, Ramsden RT, Saeed SR, van Olphen AF, Mylanus EAM Cochlear implantation in 53 patients with otosclerosis demographics, CT scanning, surgery and complications *Otol Neurotol* 2004,25 943-952

Rotteveel LJC, Snik AFM, Vermeulen AM, Mylanus EAM Three year follow-up of children with postmeningitic deafness and partial cochlear implant insertion *Clin Otolaryngol* 2005,30 242-248

Ramsden RT, Rotteveel LJC, Proops DW, Saeed SR, van Olphen AF, Mylanus EAM Cochlear implantation in otosclerotic deafness *Adv Otorhinolaryngol* 2007,65 328-34

Rotteveel LJC, Beynon AJ, Mens LHM, Mulder JJ, Snik AFM, Mylanus EAM Cochlear implantation in 3 patients with Osteogenesis Imperfecta imaging, surgery and programming issues *Audiol Neurootol* 2008,13 73-85

Rotteveel LJC, Snik AFM, Cooper HR, Mawman DJ, van Olphen AF, Mylanus EAM Speech perception after cochlear implantation in 53 patients with otosclerosis multicentre results *Submitted* 2008

Rotteveel LJC, Snik AFM, Vermeulen AM, Cremers CWRJ, Mylanus EAM Speech perception in congenitally, prelingually and postlingually deaf children expressed in an Equivalent Hearing Loss value *Submitted* 2008

STELLINGEN

behorende bij het proefschrift

Cochlear Implantation in the Compromised Cochlea

1 Zowel congenitaal, prelinguaal als postlinguaal dove kinderen hebben profijt van een CI De belangrijkste voorspellende factor is bij congenitaal dove kinderen de duur van doofheid en bij kinderen met verworven doofheid de manier van communiceren (*Dit proefschrift*)

2 Patiënten met partiële insertie van de elektrode array hebben toch profijt van een CI, hoewel minder dan patiënten met een volledige insertie (*Dit proefschrift*)

3 Ondanks de abnormale anatomie van het os petrosum, een mogelijk afwijkend verloop van de nervus facialis en het voorkomen van gusher, is de chirurgische implantatie procedure bij kinderen met congenitale malformaties van het binnenoor haalbaar (*Dit proefschrift*)

4 De resultaten die patiënten met congenitale malformaties van het binnenoor behalen met een CI zijn veelbelovend, hoewel met name bij patiënten met de ernstigere malformaties de spraakverstaan scores erg variëren (*Dit proefschrift*)

5 Stimulatie van de nervus facialis door activatie van het CI komt bij 38% van de CI patiënten met otosclerose voor en wordt met name veroorzaakt door de distale electrodes (*Dit proefschrift*)

6 CI patiënten met otosclerose met geringe afwijkingen op de CT scan, volledige insertie van de elektrode array en afwezigheid van nervus facialis stimulatie hebben een beter spraakverstaan dan de overige CI patienten met otosclerose De meest bepalende factor met betrekking tot het eindresultaat is het aantal actieve elektrodes (*Dit proefschrift*)

7 Kennis van de specifieke veranderingen van het os temporale van patiénten met Osteogenesis Imperfecta maakt de chirurgische implantatie veilig en haalbaar (*Dit proefschrift*)

8 Much is to be gained by sharing the details of difficult surgical cases rather than conceal the warts behind a thick layer of cosmetics (*Prof R T Ramsden*)

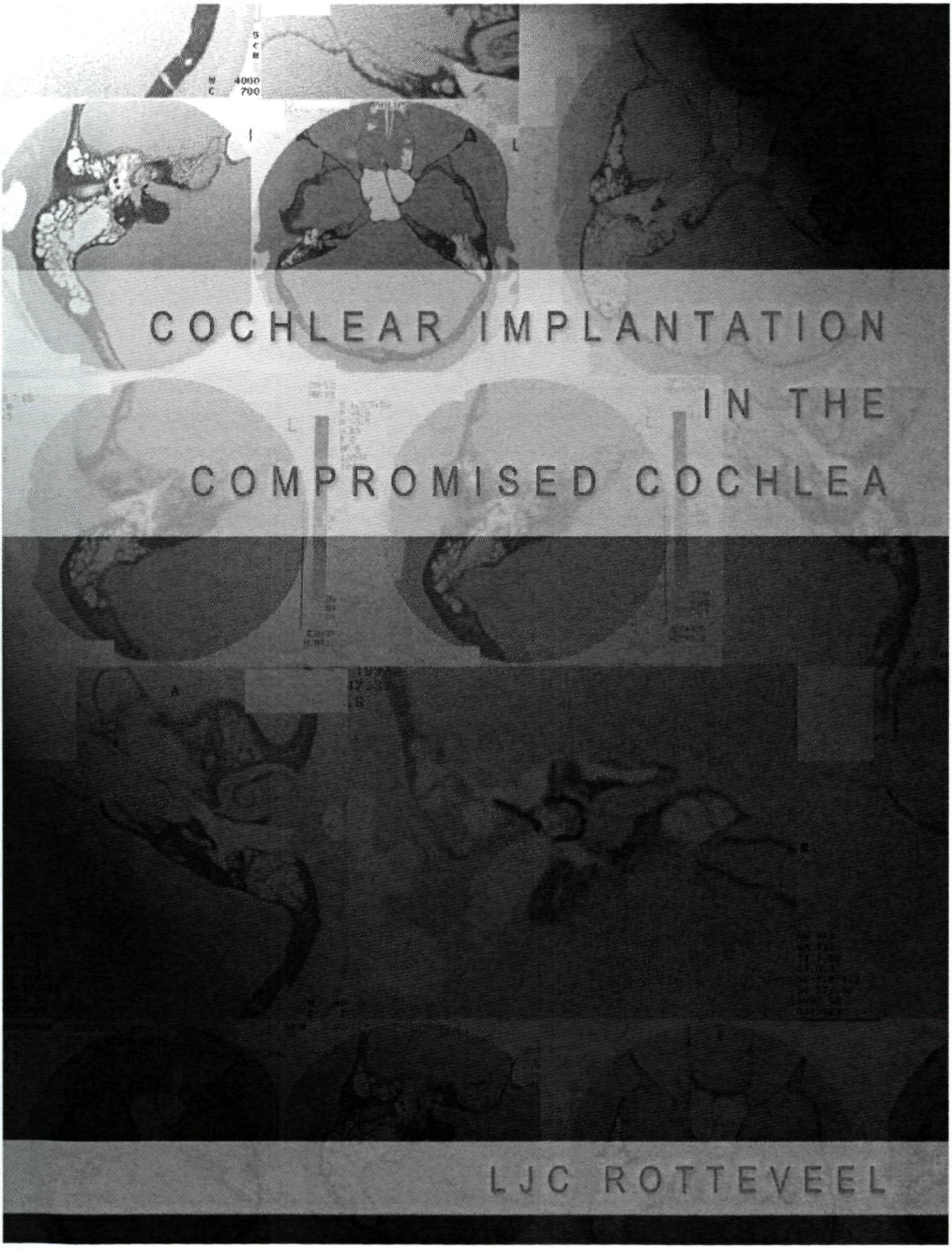
9 De 'watten-methode' is een betrouwbare test om de speekselkliersecretie te meten bij kinderen

10 Intraglandulaire Botuline toxine injecties bij kinderen met cerebrale parese zijn een effectieve therapie tegen kwijlen (*Dr P H Jongerius*)

11 Als je met computers werkt gaat alles automatisch, maar niets vanzelf

12 Het duurt langer, om iets kort te formuleren

13 Een spelfout in een proefschrift is minder pijnlijk dan een spelfout in een tatoeage


COCHLEAR IMPLANTATION
IN THE
COMPROMISED COCHLEA
LJC ROTTEVEEL