Cochlear Implantation
in the
Compromised Cochlea

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COCHLEAR IMPLANTATION IN THE COMPROMISED COCHLEA

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Chapter 1

Introduction

1.1 Hearing, deafness and cochlear implants

Hearing is a result of sound vibrations transmitted down the ear canal, through the middle ear, to the inner ear. The inner ear is a snail-like structure of 2½ to 2¾ turns embedded in bone, the cochlea, that houses the sense organ of hearing, the organ of Corti (Figure 1). The cochlear turns are wound around the modiolus which contains the spiral ganglion cells of the cochlear nerves. The cochlear turns are composed of three spiral compartments: the scala media, scala vestibuli and scala tympani. The scala media contains the organ of Corti. It lies between the scala vestibuli and tympani. The organ of Corti rests on the basilar membrane. High frequency sound produce maximal vibrations of the basilar membrane at the beginning of the cochlear turn and low frequency sound 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 forward. Hair cell movement evokes action potentials in the cochlear nerve forming patterns of excitation. These patterns are transmitted to the higher brain centers of the auditory pathway where they are interpreted as sound and processed as pitch and loudness, as well as speech (Figure 2). There are some 20,000 hair cells required for normal hearing.

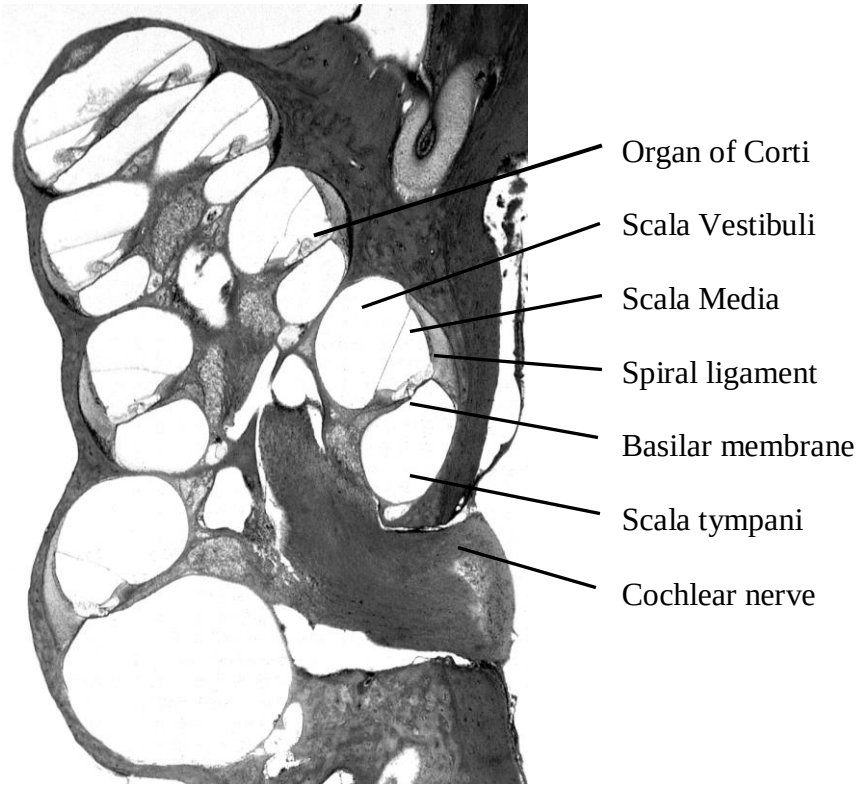

Figure 1. Cochlea of guinea-pig, 3½ turns.

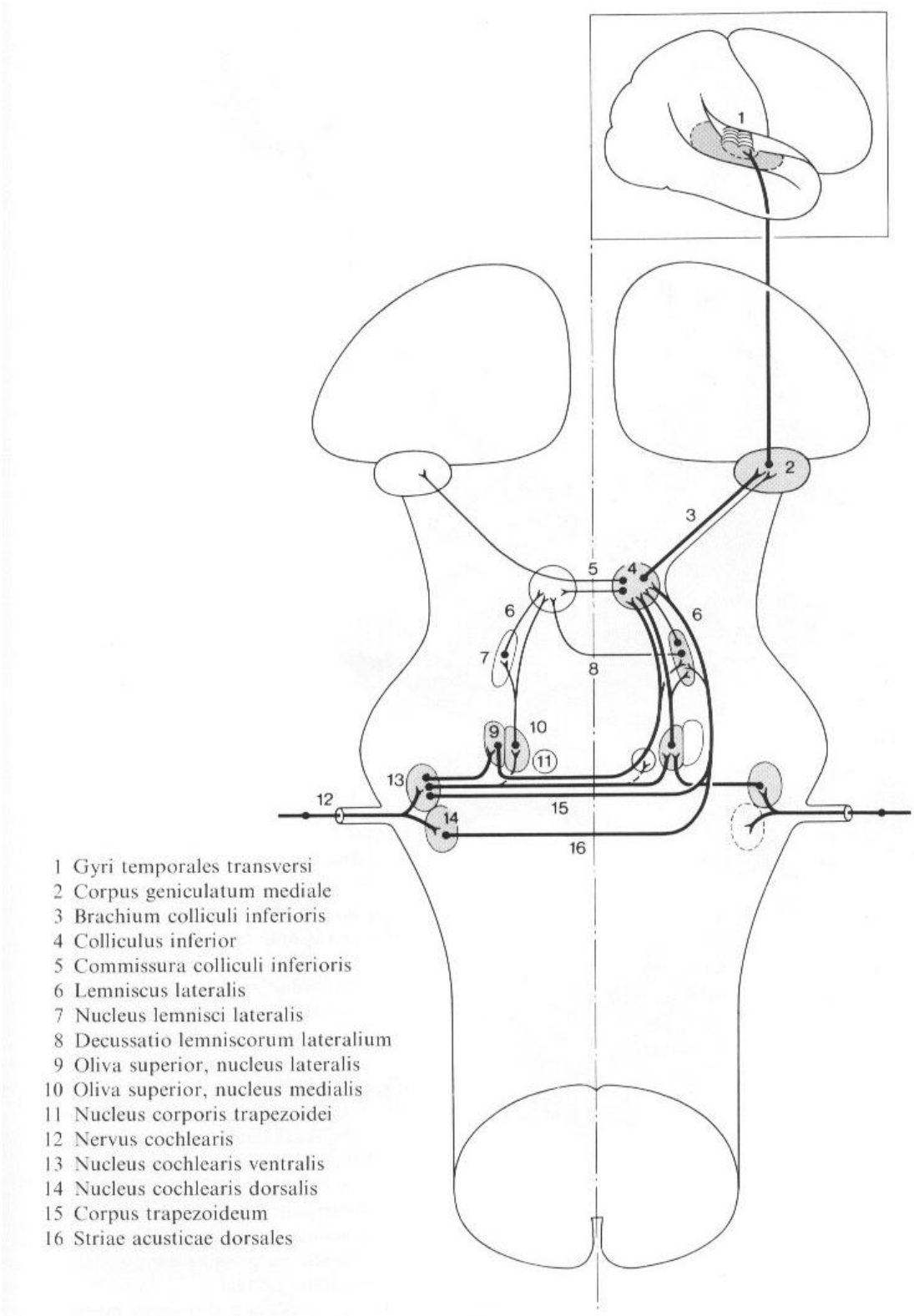

Figure 2. The central auditory pathway

When loss of hair cells occurs, the result is so-called sensorineural hearing loss (SNHL). SNHL is one of the most prevalent disabilities in the world.[1] When most of the hair cells are absent, amplification with conventional hearing aids will not help the person to hear

speech, as the auditory nerves leading to the brain centers can not be excited. A cochlear implant (CI) is an electronic device that bypasses the hair cells and provides information to the hearing centers of the central nervous system by electronically transforming acoustic vibrations into an electrical current that directly stimulates the hearing nerve. A CI can thus restore perception of sound, hearing, in severely to profoundly deaf people who do not benefit from conventional amplification and is also referred to as "the bionic ear".
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 which are sent to the speech processor. The speech processor transforms the input according to the processing strategy used. The information is then electromagnetically transmitted across the skin to an implanted receiver-stimulator, which decodes the transmitted signal and sends patterns of stimuli to intracochlearly placed electrodes (Figure 3).

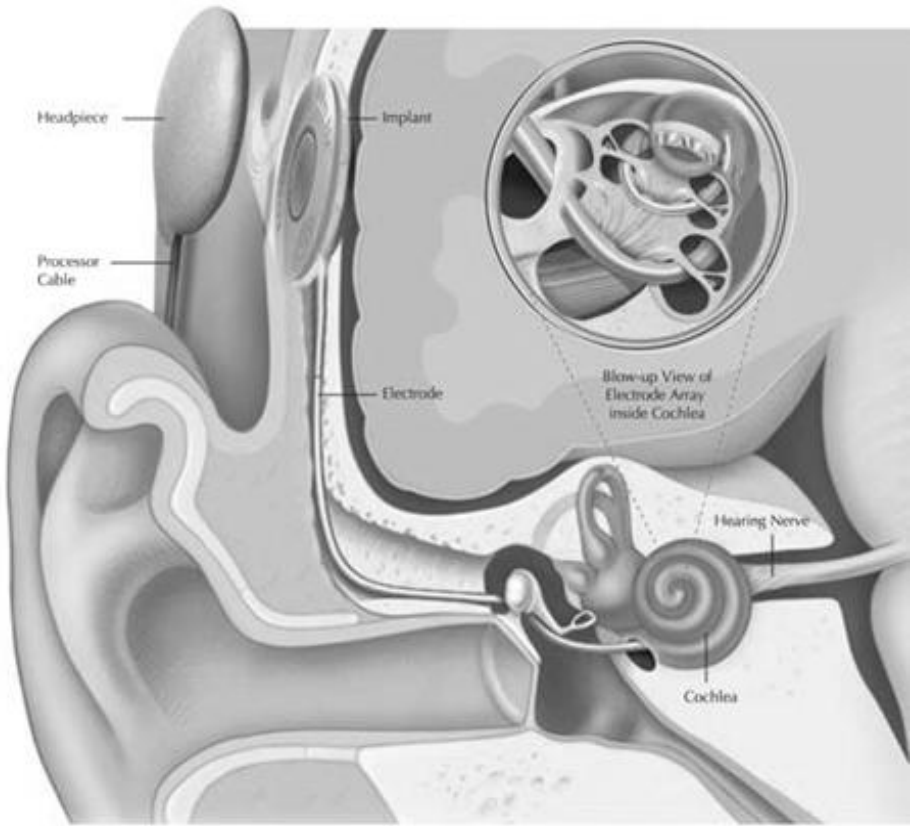

Figure 3. Intracochlear electrode array

1.2 History of Cochlear Implants

In 1790 the idea of using electrical energy to produce hearing sensation was first put into practice by Alessandro Volta: he put metal rods in 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 temporalis muscle, 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 to hear sounds like 'crickets'.

Despite the scepticism of many scientists in the 1960s about the practical possibility of a CI, as a result of extensive research, cochlear implantation has grown from a small number of isolated experimental studies, to a diverse discipline investigated by many. Few medical advances have required the integration of so many disciplines as the CI: it has been the result of research in surgical anatomy, pathology, biology, biophysics, neurophysiology, psychophysics, speech science, engineering, surgery, audiology, rehabilitation, education and quality of life studies. That cochlear implantation would become such an important otologic intervention could not have been foreseen.

From the 1960s, several types of CI devices have been developed. They can be classified into extracochlear and intracochlear systems and further classified into single-channel and multi-channel systems. Initially, it was feared that intracochlear placed devices would damage the refined hearing organ in the cochlea even more. Besides the possible consequences 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 in which the electrode was placed outside the cochlea, in the round window niche or on the promontory, were developed. 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 hearing nerve fibers connected to the different frequency regions of the brain. Moreover, electrode arrays with the right mechanical properties placed without excessive force did not cause any injury to the nerve fibers, 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 is delivered to a single electrode located in the scala tympani. With just one electrode, spectral information can not be transferred and eventually single-channel systems proved to provide too little information needed for open speech recognition.

In contrast, multiple-electrode stimulation takes advantage of the tonotopic organization of the cochlea. In 1978 the first postlingually deaf volunteer was implanted with the University of Melbourne's multiple-channel electrode array consisting of 20 electrodes.

Research not only aimed for a larger amount of electrodes in the cochlea but extended to optimize the intracochlear placement of the electrode array. Physiological data from animals[8] and modeling efforts[9] suggested that placement of the electrode array closer to the modiolus and spiral ganglion cells would result in a more localized current flow and a more effective stimulation of the neurons. This would provide better discrimination ability and speech understanding and a reduced power consumption. For this purpose, the University of Melbourne/Bionic Ear Institute developed the Contour array, the perimodiolar or 'modiolus-hugging' electrode array.[8,10] However, demonstrating their clinical efficacy has proven elusive: although some reports show better recognition scores of Contour™ users, other reports show no difference in performance between Contour™ and straight electrode users.[11]

Surgeons and researchers had to find ways to insert the electrode array as far as possible into an obliterated cochlea, as patients suffering from labyrinthitis ossificans were no longer excluded from cochlear implantation. In labyrinthitis ossificans the cochlear lumen is partly or completely filled with bone as a chronic stage in the healing of infection (for example after meningitis). Rather recently, a special implant called the Nucleus double array implant has been developed in collaboration with Cochlear Limited for those patients in whom despite attempts to create a new lumen in the ossified cochlea by drilling, the full length of the electrode array can not be inserted. The Nucleus double array implant features two separate electrode arrays containing 11 and 10 active electrodes, respectively, as well as a reference electrode located on the receiver-stimulator package. One electrode array can be placed into the drilled hole in the scala tympani of the basal turn, the other electrode array in the scala vestibuli of the second turn. Patients with a totally obliterated cochlea achieve significantly better auditory results as a result of an increased number of intracochlear electrodes.[12]

In order to achieve even better speech understanding by presenting the complex speech patterns to the nervous system by electrical stimulation, during years of research several speech processing strategies for the different CI systems were developed. The first wearable speech processor was developed in 1979.[13] In the 1980^{s} various relationships developed between research centers (University of Melbourne, House Ear Institute, Technical University of Vienna, University of California at San Francisco, University of Antwerp) and the industry (Cochlear Limited, 3M, Storz, Advanced Bionics, Philips, resp.) making the 1980^{s} the decade when CI research flourished and made great progress. Over the years, the development of new speech processing strategies showed an almost linear rise in CI patients' speech perception.

When the efficacy of cochlear implantation in adults was established, research extended to children. Early results showed that performance improved the younger the age at

implantation.[14] When implanting children under the age of 2, specific safety issues that have to be considered are the effects of drilling on head growth or head growth possibly resulting in electrode extraction in the long run, middle ear infection posing a risk of bacterial meningitis and the effect of electrical stimulation on a maturing nervous system. After several studies in the 1990[s] these issues proved to be no main cause of concern provided the electrode array entry point is properly sealed and implantation takes place in the absence of middle ear infection.[15,16]

1.3 Patient selection

When defining a criterion for implantation, one has to establish when the advantages of implantation outweigh the disadvantages. The preoperative evaluation of adults for cochlear implantation aims to select the patients who have a high probability of achieving better hearing opportunities than with their (appropriate and optimally fitted) hearing aids. For this purpose, preoperative speech perception and communication abilities need to be assessed carefully. Further, the candidate must have realistic expectations about the benefits and risks and have adequate help from family or social services to undertake the rehabilitation. When cochlear implantation was first introduced, it was provided only to the extreme cases, i.e. profoundly hearing-impaired adults who received no measurable benefit from conventional hearing aids. As a result of the positive outcomes, the criteria of candidacy became more liberal.

To be able to predict the benefit of cochlear implantation, the results of cochlear implant recipients have been analyzed extensively.[17-22] Several factors have been identified to influence the variance in performance among cochlear implant recipients. The general predictive factors that are common to the adult and the child are age when deafened, 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 indirectly influence a cochlear implant patients' performance by altering the anatomical, biophysical and biochemical properties of central nervous system (CNS), nerves and cochlea. Hereby, these indirect factors may also influence the electrode array's position in the cochlea and the number of active electrodes (Table 1).

Age at implantation correlates negatively with performance in adults only if the person is over 60 years[23] and in children if they have been born deaf or deafened early in life. The age when deafened determines whether a child is prelingually or postlingually deaf, the previous meaning the child has had little language experience. This has proven to be a negative factor, which can be overcome by early implantation which shortens the duration of deafness.[24] A negative correlation has been found for duration of deafness.[17,18] The

presence of some residual hearing[20], speech-reading ability[10] and duration of implant use[23] correlate positively with results.

The general predictive factors that specifically apply to children are language level and communication mode, mode of education after surgery, parental support, and delayed cognitive and motor milestones. Language development influences speech perception and vice versa and children do better if they were in auditory-oral communication programmes. In children with delayed cognitive and motor milestones learning takes longer and they do 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 a complex matter.

Table 1. Factors of influence on the variance in performance among cochlear implant recipients

Direct factors		Indirect factors
Device	speech processor characteristics; stimulator; electrode array	
Electrodes	position in the cochlea; number in use	aetiology of deafness (infection, ossification, demineralization, malformation, trauma, toxicity, genetics)
Cochlea	electrical properties; size; physical condition	aetiology of deafness (ossification, demineralization, malformation, trauma, toxicity, genetics) age at implantation (natural degeneration)
Nerves	spiral ganglion cell survival; spiral ganglion cell function	aetiology 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	aetiology 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 requires consultation from other specialists such as a psychologist, speech pathologist, neurologist, social worker or general physician (CI team). The audiologic consultation during the preoperative assessment consists of pure-tone audiometry, middle ear impedance testing and evaluation of speech perception and production and hearing aid evaluation. The otological consultation involves a medical history and standard ENT

examination, high-resolution computed tomography (HRCT) scan/ magnetic resonance imaging (MRI).

Imaging of the patient's temporal bone in the preoperative work-up for cochlear implantation enables the surgeon to study the morphology of the temporal bone as visualized by both high-resolution CT scans and MRI scans in order to be prepared for possible surgical difficulties. The positions of surgical landmarks such as the short incudal process, the vertical portion of the facial nerve, the oval and round window, the carotid canal and the sigmoid sinus can thus be studied (Figure 4).

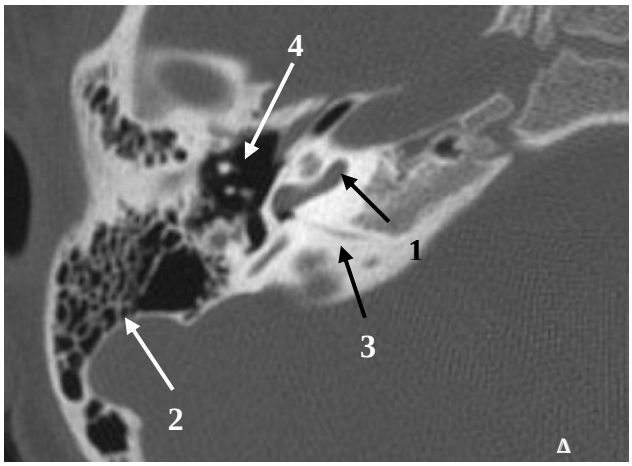

Figure 4A. Axial view of normal petrosal bone
1. Basal turn of the cochlea; 2. Mastoid air cells; 3. cochlear aquaduct; 4. Tympanum

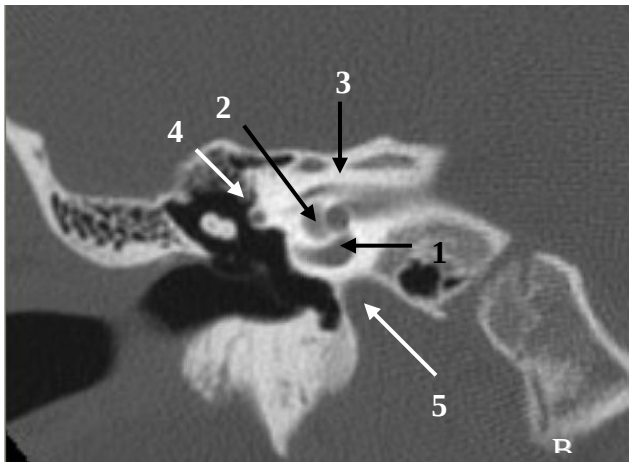

Figure 4B. Coronal view of normal petrosal bone
1. Basal turn of the cochlea; 2. Middle turn of the cochlea; 3. Labyrinthine segment of facial nerve; 4. Tympanic segment of facial nerve; 5. Jugular bulb

Variations in morphology may be encountered, but also abnormalities due to previous surgery and pathological alterations caused by ossification (in meningitis and oto-sclerosis), spongiosis (in otosclerosis, Osteogenesis Imperfecta and M.Paget) and congenital malformations. The degree of ossification must be taken into account when considering which ear to implant. The surgeon can arrange to have a double array implant available in the operation theatre. Preoperative awareness of the morphology is especially important in congenital malformation of the inner ear: when a well-defined modiolus is present, the insertion of a modiolus-hugging device is preferred. If not, the nerves may lie peripherally in the cavity so that a straight array is preferred. The location of the facial nerve must be noted because of a high percentage of aberrant courses 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 in case of a normal cochlear nerve canal.[28] *Early* cochlear

obliterarion and central causes of hearing loss are best shown on MRI.[29] Bettman et al.[25] proposed a CT imaging protocol for preoperative scanning of the temporal bone in CI candidates, while others[30,31] prefer the information provided by MRI in the decision making of cochlear implantation. Recent research by Trimble et al.[32] showed that although there is overlap between the imaging modalities HRCT and MR in the type of abnormalities detected, preoperative dual-modality imaging with HRCT and MR of the petrous bone and MRI of the brain in paediatric CI candidates detects abnormalities related to deafness, which would not have been found using either modality alone. They present an algorithm for selective use of either imaging modality, using the patient risk factors identified in the study.

Table 2. Key points for preoperative imaging studies.

Modifying surgical strategies or implant device	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

1.4 Surgical implantation procedure

The surgical procedure of cochlear implantation is similar in children and adults. Several modifications of skin incisions and approaches to the middle ear and cochlea have been developed.[33-35] At the Nijmegen/Viataal CI program the most common applied method is used in which the middle ear is reached via a mastoidectomy and facial recess approach and 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 syringe and needle containing methylene blue. A single flap is created containing skin as well as superficial and deep fascia. The deeper periostal flap is incised at a different location. When the landmarks of the mastoid bone are exposed, a well is drilled for placing the receiver-stimulator. Then mastoidectomy and the facial recess approach are used to gain access to the middle ear and cochlea (Figure 5).

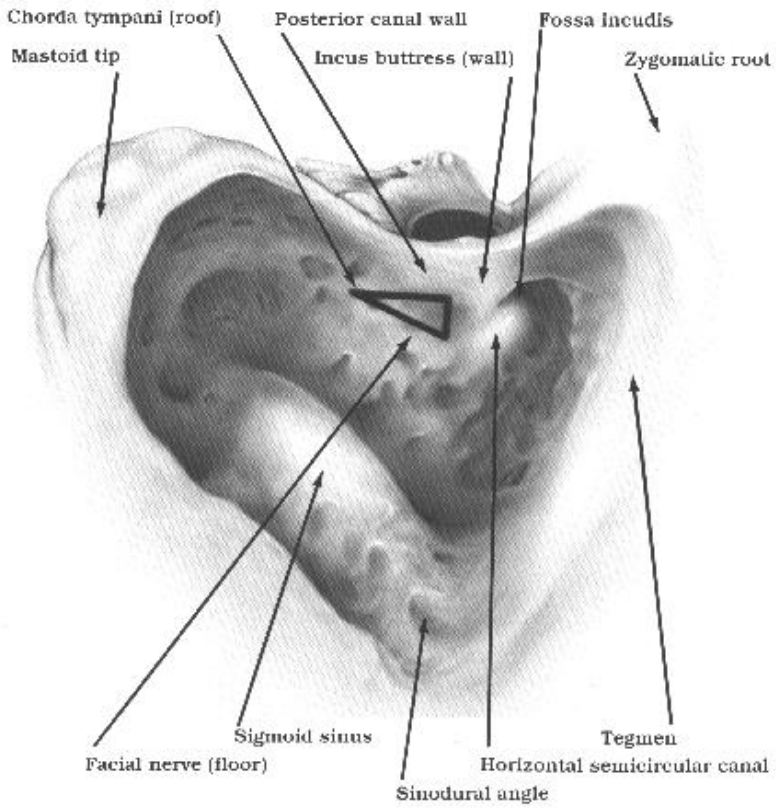

Figure 5. Mastoidectomy and site of the posterior tympanotomy.

The medial wall of the middle ear, the tympanum, presents a round eminence called the promontory which is the bulge of the basal turn of the cochlea. It 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 which overlies the scala tympani of the basal turn. The cochleostomy is made just anteroinferior to the round window to allow insertion of the electrode array into the scala tympani of the basal turn (Figure 6).

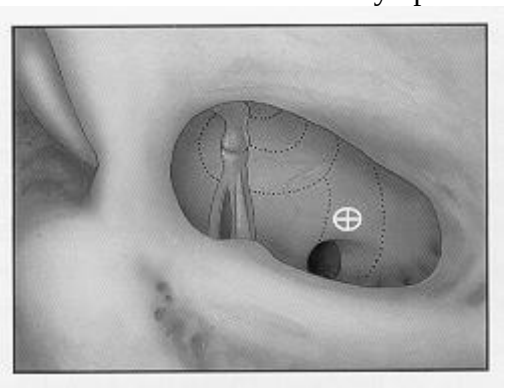

Figure 6. Cochleostomy

When resistance is felt or buckling of the array is seen during the insertion, it is important not to continue: a forceful insertion may damage the inner ear or electrodes. The location of the cochlear turns in relation to the medial wall of the tympanum are of particular importance in case of ossification of the cochlear lumen, for which drilling is necessary to

create a new lumen and make (partial) insertion possible. In case of severe ossification, the use of the double array implant might be indicated. The first cochleostomy is routinely made anterior to the round window and provides access to the basal turn (both the scala tympani and the scala vestibuli). Possible present connective tissue and bone can be removed until the anterior wall of the basal turn is approached. A second cochleostomy is performed at the second turn caudal of the cochleariform process and 2 mm anterior of the oval window after removal of the incus. Newly formed tissue should also be removed if necessary. The two electrode carriers are then placed into the scala tympani of the basal and the scala vestibuli of the second turn, respectively. The remaining surgical procedure is identical with that used for cochlear implantation in patients without obliterated cochlea's. In congenital malformation of the inner ear, the position of the facial nerve and cochlear windows may help locate the scalae so that a safe insertion can be performed.
The cochleostomy must be properly sealed, completely circumferential using fascial autograft or pericranium, to prevent middle ear infection spreading to the inner ear which could lead to labyrinthitis and even meningitis. To prevent extrusion of the electrode array from the cochlea during growth, it is fixed in the fossa incudis, because the distance from there to the round window does not change after birth.
Before sealing the cochleostomy, some CI centers perform intra-operative 3D rotational X-ray to be certain the electrode array is in the right position.[36] However, in patients with normal petrosal bones on the preoperative CT scan and in whom the surgical procedure is uneventful, in most CI centers intraoperative neural response measurements (NRT) are considered to be sufficient. Even routinely performing a postoperative X-ray (Stenvers) to be informed on electrode array position[37] is a practice that some CI centers abandon: imaging might only be indicated in those patients with abnormal postoperative clinical condition or abnormal implant electrophysical measurements.

1.5 Rehabilitation and Results

After recovery from the CI surgery, the speech processor is fitted. Threshold levels (T level) and maximum comfortable levels (C levels) for electrical stimulation on each electrode must be established and programmed into the patient's speech processor, called the MAP, to optimize the speech signal presented. The current levels between the T and C levels cover the neuronal dynamic range. The frequency boundaries for the electrode to be stimulated are also set to determine the pitch range per electrode. The MAP can be reprogrammed should the patient not be content with the sound perceived. The patient attends training sessions in how to interpret the sensations created by electrical stimulation. In adults, the rehabilitation is mainly focused on the development of speech recognition. In children, besides the development of speech perception, it involves the

development of speech production, receptive language and expressive language. The speech material used in auditory training is age appropriate and may be specific speech tokens such as vowels and consonants, or sentences and words. These training exercises can also be used to asses the performance of the patient.
During the rehabilitation, predictive factors for good outcomes, as described earlier, must be considered. For example, patients with a long duration of deafness are more likely to require long periods of rehabilitation for adequate speech perception. Medical conditions of the central nervous system can influence learning abilities. Children with developmental delay and learning disorders might have poorer speech perception and develop at a slower rate.[38] The age when deafness occurred is of importance since there is a 'most sensitive period' for the development of language within the first years of life. Fortunately, early results on cochlear implantation in young children suggested that both children born deaf and those deafened early in life could achieve similar benefits to adults who had been postlingually deaf and most of these children could get open-set speech recognition.[39,40]

1.6 The Nijmegen/Viataal CI programme

History
The first adult implantation procedure in the Netherlands was performed in Utrecht in 1985, followed by Nijmegen in 1987. Initially, implantation in adults was restricted to a government sponsored study of 20 patients. The positive findings in adults[41,42] led to the approval of cochlear implantation by the Dutch minister of health in 1997, after which the number of adult patients implanted each year rose (Figure 7). The first implantation of a child in 1989 was performed at the Radboud University Medical Centre in Nijmegen. A second clinical study on cochlear implantation in children was conducted from 1993-1996.[43] The first congenitally deaf child was implanted in 1990 at 13 years of age. Implantation in a congenitally malformed cochlea (Mondini's dysplasia) first took place in 1994 in a 7-year-old congenitally deaf girl. In 2001, the first Nucleus Double array cochlear implant was used in a postmeningitic deafened 6-year-old girl with ossified cochleae. At the end of 1999, the Dutch minister of Health approved cochlear implantation for prelingually deaf children.
For the rehabilitation of implanted adults and children, the Radboud University Nijmegen Medical Centre works in close collaboration with Viataal, the former Institute for the Deaf at St. Michielsgestel and together they form the Nijmegen/Viataal CI Centre. At this CI centre, at the end of 2006, 650 implantation procedures had been carried out in 316 adults and 285 children, including bilateral and reimplantation procedures.
In order to stay experienced in the various CI systems produced by the industry the Nijmegen/Viataal CI centre implants devices of more than one manufacturer. This enables

to make the best choice for a particular patient based on audiologic criteria as well as handling. Table 3 shows the devices used and their characteristics.

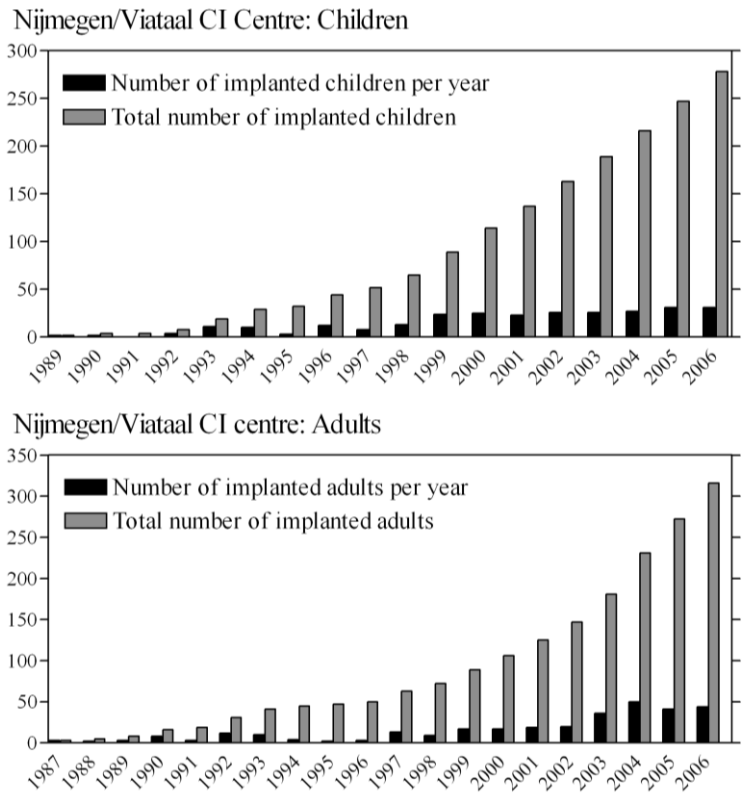

Figure 7. Number of patients implanted at the Nijmegen/Viataal CI centre.

Patient selection: work-up procedure

As described earlier, determining whether or not a deaf patient is a suitable candidate for cochlear implantation is a complex procedure. As the experience of Nijmegen/Viataal CI team throughout the years grew and cochlear implantation proved to be beneficial, the criteria for cochlear implantation (for example duration of deafness, amount of residual hearing and presence of additional handicaps) became less strict. This trend can be observed in CI teams and researchers worldwide. Patients that had been advised against implantation years before, were considered suitable candidates later. For example, during counseling of a prelingually deaf adult, despite the long duration of deafness, a thorough evaluation is done of the possible surplus value of cochlear implantation for that individual. This surplus value is not only based on the expected speech perception achieved after implantation, but also on the acquirement of perception of sound alone and quality of life. This can even lead to the implantation of patients with psychomotor retardation, which in Nijmegen was first performed in 2002.

Table 3. CI devices used in the Nijmegen/Viataal CI programme since 1987

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

Postoperative performance is influenced by various factors as described in paragraph 3 and during the work-up period standard evaluation of these factors is done by medical (medical history, ENT physical exam, MRI, CT scanning, ENG), audiological (pure tone audiogram, speech perception, BAER) and psychological tests, as well as speech-language tests in children. When considered necessary, supplementary measurements such as Auditory Steady State Responses (ASSR), electro cochleography (ECoG) and round window stimulation are performed.

Postoperative Evaluation protocol

In adults, 3 weeks after the implantation procedure, the speech processor is fitted ('week 0'). The necessary equipment is provided and checked. T and C levels are established and programmed into the speech processor (the MAP). In the first year post-implantation there are several fitting sessions. Audiometrical data are collected 5 weeks ('week 5') and 1 year ('week 52') after the first fitting session. In week 5 free field audiometry with CI and, in case of preoperative residual hearing, free field audiometry without CI is performed. One year postoperatively, besides free field PTA, word discrimination (AN spondee test) and speech perception in quiet and noise (word and phoneme scores on the open set NVA monosyllable word lists) are measured. At Viataal, the rehabilitation therapist assesses quality of life using the Nijmegen CI Questionnaires (NCIQ). After the first year of follow-up, annual equipment check-ups and measurements of PTA, word discrimination and speech perception are performed by the audiologist alternatively at Nijmegen and Viataal.

In children, rehabilitation takes place at Viataal and starts 3 to 6 weeks after the implantation surgery. One week after the first fitting session in which the CI is activated and the child's hearing sensations produced by the implant are assessed, an extensive week of fitting and stimulating lead by the audiologist and rehabilitation therapist takes place during which the child and it's parents can stay at the Guest house of Viataal. At 1-, 3-,6-, 9-, and 12-months follow-up the child's auditory development is monitored by audiometry, speech perception and speech production tests. The rehabilitation therapist further makes regular home visits and, in case of school going children, visits the school where the child is educated. After 1-year of follow-up, annual measurements are performed with special attention to the development of spoken language.

Nijmegen/Viataal CI centre: Results

The speech perception scores at 1-year follow-up of 213 adult CI patients enrolled at the Nijmegen/Viataal CI programme implanted with different CI systems are shown in figure 8. The average speech perception scores one year after implantation gradually improves through the years, although a wide variation in performance persists.

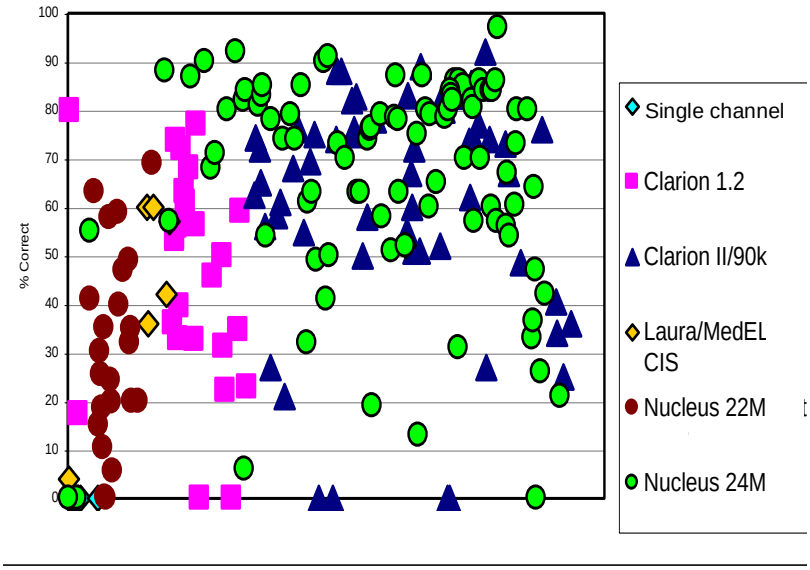

Figure 8. Monosyllable word recognition at 1-year follow-up

1.7 Aim of the thesis

Ever since cochlear implantation has first been introduced, efforts have been made to explain the variability in outcomes of CI patients in order to improve the results and to be able to select those patients who are expected to benefit from cochlear implantation. As described earlier, several factors have been recognized to influence the performance of a patient with CI. When the influence of factors such as the age of implantation and duration of deafness became apparent, 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. During this time, the criteria for implantation were still rather strict. This resulted in a fairly homogenous group of children. In order to deal with the 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 the different speech perception results were reduced into one measure: the "equivalent hearing loss"(EHL).[44] This outcome measure refers to the performance of a reference group of severely and profoundly hearing impaired children with conventional hearing aids. An attempt was made to explain the variability in long term speech perception performance of these implanted children by several factors, such as the age at implantation, duration of deafness and communication mode.

Since the criteria for cochlear implantation have been relaxed, the teams of CI centers are more and more confronted with patients deafened by a varying etiology that were once

considered a contraindication for cochlear implantation. Etiology of deafness is another predictive factor which has been recognized to more or less influence the performance of a patient with CI.[21,23] Disease of the cochlea can cause SNHL by several histopathological changes and might affect auditory performance of a patient with a CI via factors, such as the number and position of active electrodes, the electrical properties of the temporal bone, ganglion cell and central neural survival or function. Especially diseases that alter the morphology of the cochlea and otic capsule may compromise cochlear implantation.

Meningitis is an important disease in cochlear implantation, as it may cause deafness possibly requiring cochlear implantation, as well be a rare complication of cochlear implantation (device-related meningitis). Infection can spread from the meninges to the cochlea through the cochlear aqueduct and several foramina in the osseus labyrinth causing labyrinthitis, but also in the reverse direction when the infection originates in the middle and inner ears. When labyrinthitis occurs, new bone formation (meningogenic labyrinthitis ossificans) is part of the healing phase after inflammation and stimulable spiral ganglion cells may be lost[45] which might negatively influence outcomes with a CI. Further, the surgical implantation procedure of an obliterated cochlea may require special techniques such as various degrees of drilling[46,47], alternative placement of the electrode array in the scala vestibuli or extracochlearly[48,49] and 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 a partial insertion on the postoperative performance with CI, in Chapter 3 the outcome of children with postmeningitic deafness and partial insertion of the electrode array due to ossification of the cochlea is explored and compared to that of children with postmeningitic deafness and full insertion of the electrode array.

In the early days of paediatric 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 been deaf since birth. 20-30% of all cases of congenital profound hearing loss have bony abnormalities of the labyrinth.[50,51] Malformations of the cochlea are classified by Jackler et al.[50] based on embryonic life and may vary from total aplasia, severe cochlear hypoplasia, mild cochlear hypoplasia, common cavity, severe incomplete partition, mild incomplete partition to a subnormal cochlea which doesn't reach a full 2.5 turn. Besides a variable functioning and possibly tonotopically disorganized cochlear neuroepithelium, some inner ear malformations are associated with aplasia or hypoplasia of the cochlear nerve. In these cases, where there are no or at least insufficient nerve fibers, cochlear implants will be of no benefit.[52] Another challenge for the implantation team is that the surgical implantation procedure may be hindered by the presence of an anomalous facial nerve and the occurrence of cerebrospinal fluid (CSF) gusher.[53,54] Also postoperatively,

during activation and programming, specific problems can occur.[53,55,56] In Chapter 4 the surgical aspects and performance outcome of cochlear implantation in children with malformed inner ears are described.

In the adult CI population, over the past years, an increasing number of patients who received a CI have been diagnosed with otosclerosis (7 to 9.5%).[57] Otosclerosis is a heritable disease that only affects the bony structure of the temporal bone. In the active phase (otospongiosis), the normal lamellar bone is resorbed and is replaced by thick, irregular bone in the normal middle layer of the otic capsule (otosclerosis).[58] The subsequent hearing loss can be conductive, which is most commonly caused by stapes fixation, or sensorineural in the case of cochlear involvement. SNHL in otosclerosis is thought to be the result of narrowing of the cochlear lumen with distortion of the basilar membrane[59] or caused by lytic enzymes that are released into the perilymph from otosclerotic foci.[60,61] Histological studies have shown that otosclerosis has a relatively small effect on spiral ganglion cell survival compared to other causes of deafness.[62] If treatment such as stapedectomy or stapedotomy fails, the patient may become a candidate for CI surgery. The surgical implantation procedure might be hindered by obliteration of the round window or basal turn. Further, the otic capsule might be softened by otospongiosis so that an electrode array that is pushed forward easily penetrates. In order to evaluate the feasibility of cochlear implantation in patients deafened by otosclerosis, in Chapter 5 a group of 53 otosclerosis patients with a CI was evaluated for special features that had occurred during surgery and rehabilitation.

Another heritable bone disease which can cause severe SNHL is Osteogenesis Imperfecta (OI). OI is a heterogeneous disease of the connective tissue caused by a defective gene that is responsible for the production of collagen type I, leading to defective bone matrix and connective tissue. Hearing loss affects 35-60% of the patients with OI, most often in the form of the conductive or mixed type.[63] 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.[64,65] As the hearing loss will progress to deafness in 2-11% of OI patients[65,66], cochlear implantation may become the only remaining treatment option in some patients.
At the Nijmegen/Viataal CI Centre, three patients with OI have been enrolled in the CI rehabilitation program. In Chapter 6 the specific problems encountered during surgery and rehabilitation after cochlear implantation of these patients are described. To evaluate the

effect of the possible lower electrical resistance of an affected temporal bone, psychoacoustical, electrical and electrophysiological measures were performed.

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Chapter 2

Cochlear implantation in children

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

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(Submitted)

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. Furthermore, device type and number of active electrodes was also rather uniform.

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 duration of deafness, whereas communication mode was less significant. In the children with acquired deafness, communication mode and age at onset of deafness explained 71% of the variance, whereas duration of deafness was not a significant factor.

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, 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, e.g. 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 with 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 duration of deafness, age at onset, cause of hearing loss and communication mode. During the late nineties, inclusion criteria changed with respect to e.g. residual hearing, cognitive ability and age at implantation. 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, deveice type, support at home). The present study investigated this relation in 67 consecutive children implanted 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 implantation. After 3 years, many of the children were mainstreamed or placed at special schools for children with hearing impairment (not deaf). In the majority of children, the

communication mode remained the same throughout the 3-year study period, so it 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 University Medical Centre St Radboud 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 non-verbal intelligence.

Group demographics are shown in Table 1. Thirty-five children were born deaf with aetiologies 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. The children were grouped by age at implantation 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] 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. 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 post-operative 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 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).

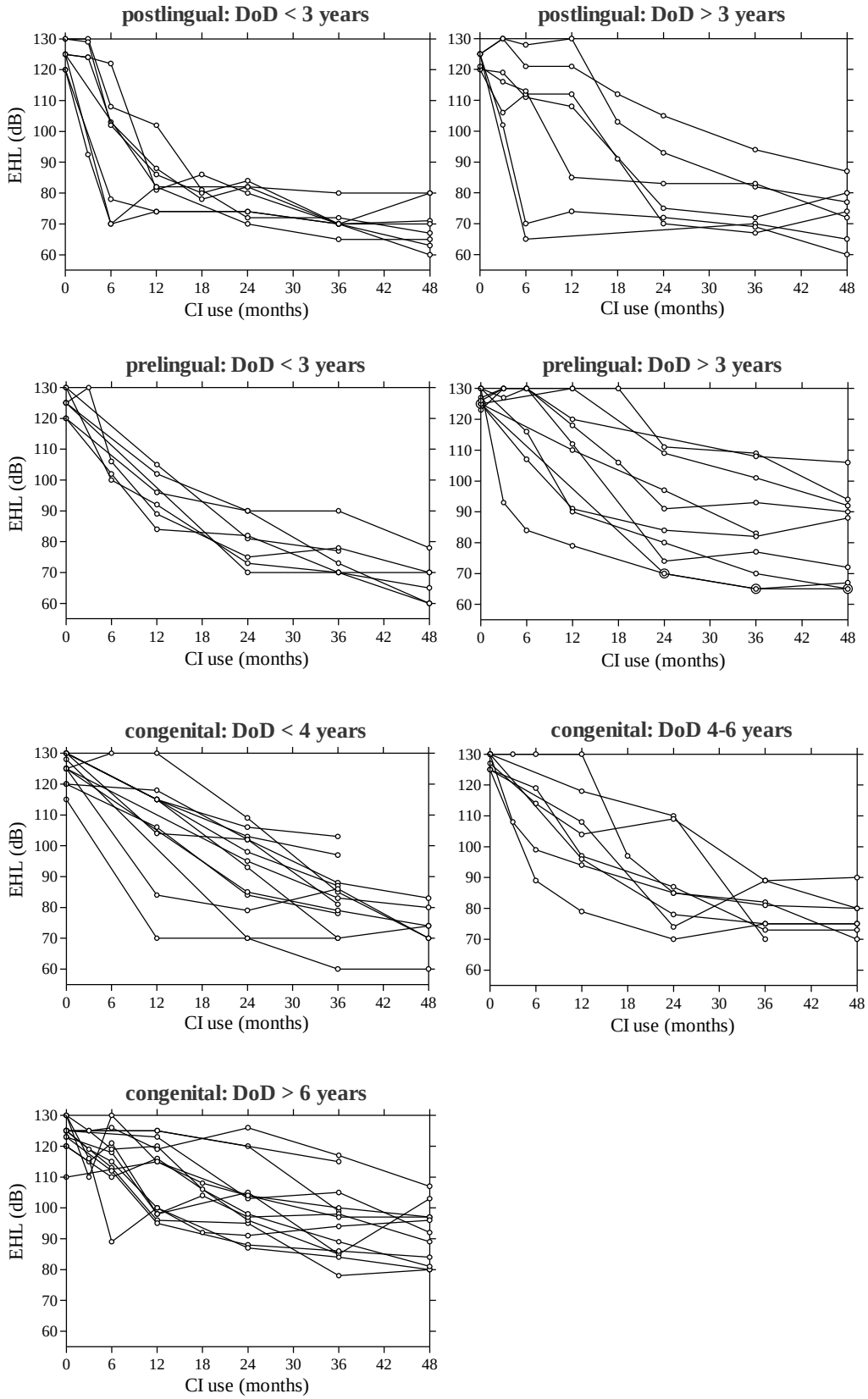

Figure 1. Individual longitudinal data of EHL in all subgroups. DoD= duration of deafness

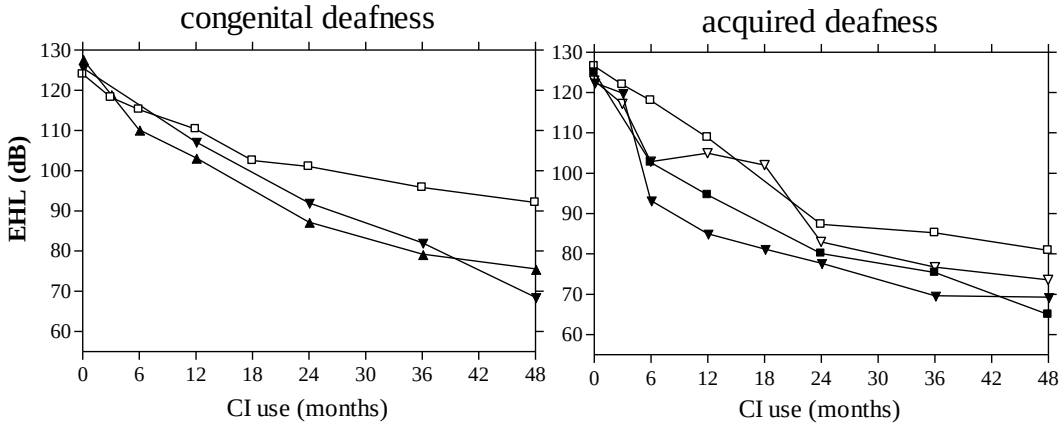

Figure 2. Longitudinal analysis of the mean EHL in prelingually and postlingually deaf children (right) and congenitally deaf children (left)

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.

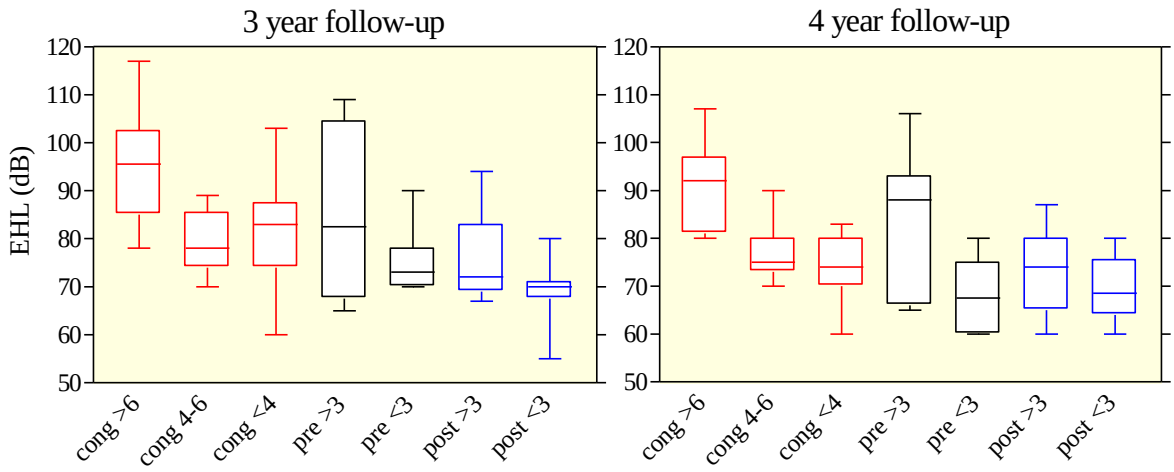

Figure 3a Figure 3b

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 (n=14, n=11);
Cong 4-6 = congenitally deaf children, 4 to 6 years old at implantation (n=8, n=7);
Cong < 4 = congenitally deaf children, younger than 4 years at implantation (n=13, n=7);
Pre > 3 = prelingually deaf children, duration of deafness longer than 3 years (n=10, n=9);
Pre < 3 = prelingually deaf children, duration of deafness less than 3 years (n=7, n=6);
Post > 3 = postlingually deaf children, duration of deafness longer than 3 years (n=7, n=7);
Post < 3 = postlingually deaf children, duration of deafness less than 3 years (n=8, n=8)

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.

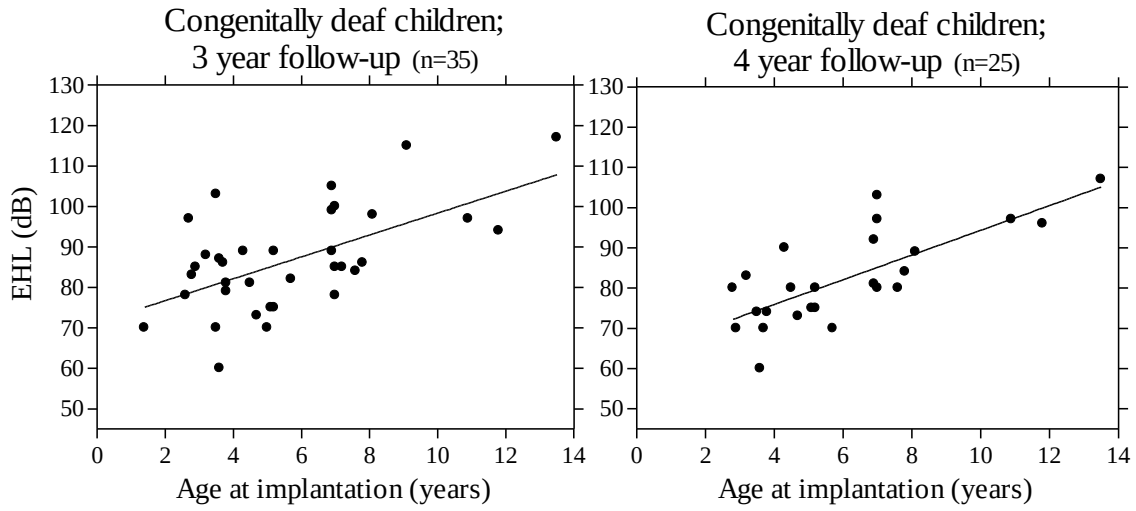

Figure 4. Equivalent Hearing Loss obtained at 3 and 4 years of follow-up as a function of the age at implantation for the group of congenitally deaf children

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.

Conclusion

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.

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Chapter 3

Cochlear implantation in the postmeningitic ossified cochlea

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

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Clinical 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: Three years after implantation, speech perception in the partial insertion children was poorer than that in the control group. 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 as many as 70% of cases with bacterial meningitis and profound hearing loss. The frequency and severity of ossification varies according to the causative organism.[2] Usually, neo-ossification is most marked in the scala tympani of the basal turn of the cochlea, while 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 was once considered to be a contraindication for cochlear implantation, nowadays many centres implant these patients routinely.
Meningogenic labyrinthitis ossificans poses two concerns in cochlear implantation: firstly, the loss of stimulable spiral ganglion cells and secondly, the technical difficulties of dealing with surgery of an obliterated cochlea. Histopathological temporal bone studies on the relation between the severity of labyrinthitis ossificans and ganglion cell survival are inconclusive. Hinojosa et al[5] did not find such a relationship and they observed large variability in ganglion cell populations in temporal bones of deaf subjects with and without ossification. Others, however, found a tendency towards substantial spiral cell loss in middle and apical turns affected by ossification.[5] Surgical techniques to deal with obstruction of the cochlea include 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 children with profound postmeningitic deafness who received a cochlear implant (CI).[11,12] Significant improvements were found in all areas of speech perception and production compared to their pre-operative performance with conventional hearing aids. Electrical stimulation of the auditory nerve was feasible despite the presence of new bone formation. This lead to

comparable results between implanted patients with ossified cochleae and patients with patent cochleae.[2,7,15] The degree of ossification did not appear to affect speech perception performance.[13] However, if the electrode array can only partially be inserted due to extensive cochlear ossification, the results can be less favourable.[7] Surgical procedure, placement and the number of electrodes in use seem to affect the patient's performance. 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] Many factors other than the number of electrodes are known to influence a child's performance with his or her CI, such as 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 at the University Medical Centre St. Radboud in Nijmegen between 1990 and 1998. Inclusion criteria are listed in Table 1. Twenty-five consecutive children were selected to take part in this study. The infection was caused by Streptococcus pneumoniae in 17, by Neisseria meningitidis in one and by Haemophilus influenzae in four patients. In two children the pathogen could not be demonstrated.

Seven children had partial insertion of the Nucleus multichannel CI electrode array. Partial insertion was defined as electrodes visible outside the cochlea during surgery, because it was impossible to insert all 22 electrodes through the cochleostomy.

Table 1. Inclusion criteria

Residual hearing	Profoundly deaf with hearing thresholds at 1, 2 and 4 kHz that exceed 95 dB HL and 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

Mean age at onset of deafness in this partial-insertion group was 1.8 years; mean age at implantation was 5.5 years; mean duration of deafness was 3.7 years. Duration of deafness was defined as the time between onset of deafness and implantation.
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; mean duration of deafness was 2.9 years. As the duration of deafness influences speech perception,[14] these children were divided into two groups according to whether the duration of deafness was longer or 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 preoperatively using computed tomography (CT) scanning. These scans were reviewed retrospectively and compared to the degree of cochlear ossification observed at surgery.
If it was impossible, during surgery, to locate a lumen in the scala tympani or the scala vestibuli that would allow insertion of all the electrodes, limited drilling of 6 to 8 mm was performed in an anteromedial direction in the obliterated basal turn along the scala tympani until a lumen became visible, or alternatively, until a new channel had been created.
All the children received the Nucleus 22 or 24 CI. The speech processors were programmed four to six weeks after surgery. During the rehabilitation period, testing took place and readjustments were made to the programming parameters to improve each child's performance. Almost all the children were using the MPEAK or SPEAK strategy. Four children were using the ACE strategy: three children in the control group (C16, C17 and C18) in their 3-year follow-up assessment and one child in the partial-insertion group (S1) in his second and third years of follow-up.

Performance
Aided sound-field thresholds were measured using narrow band noise with central frequencies from 0.25 to 4 kHz.
The speech perception data were analysed and computed into one single measure, called the 'equivalent hearing loss' (EHL).[15] To do so, 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 test scores and these children's degree of hearing loss were established with statistical procedures. These relations were used to transform the scores of subjects with a cochlear implant into an EHL value. EHL values vary between 50 and 130 dB hearing level (HL). This measure

can be used to summarise 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 supra-segmental 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). Test battery and procedures have been reported in detail.[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 identified preoperatively 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 out of seven children with partial insertion. In the remaining child, ossification seemed absent in one ear, but severe in the other ear. However, the quality of the CT scan was poor due to movement artefact.

Table 2. Group characteristics

Subject	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
S1	2.7	S. pneumoniae	3.5	6.2	Total	8	8
S2	2.6	Unknown	3.8	6.4	Total	10	Non user
S3	0.6	S. pneumoniae	8.6	9.3	Total	10	13
S4	0.4	H. influenzae	3.3	3.7	Total	13	11
S5	2.6	S. pneumoniae	3.0	5.6	None	13	13
S6	3.3	H. influenzae	1.8	5.2	Total	12	13
S7	0.4	S. pneumoniae	1.8	2.3	Partial	18	17
C1	2.7	S. pneumoniae	2.4	5.1	None	27	22
C2	2.7	S. pneumoniae	2.5	5.2	Partial	32	18
C3	2.5	S. pneumoniae	3.5	6.0	Partial	32	20
C4	2.6	S. pneumoniae	3.8	6.4	Partial	27	19
C5	2.9	H. influenzae	2.3	5.2	Partial	27	22
C6	2.3	H. influenzae	3.2	5.6	Partial	32	21
C7	3.0	H. influenzae	1.3	4.3	Partial	32	22
C8	0.2	S. pneumoniae	5.9	6.2	Partial	32	22
C9	1.8	S. pneumoniae	4.0	5.7	None	32	21
C10	1.4	S. pneumoniae	1.9	3.3	Partial	26	20
C11	1.7	S. pneumoniae	4.1	5.8	None	32	20
C12	0.9	N. meningitidis	2.0	2.9	None	27	20
C13	1.4	S. pneumoniae	2.2	3.6	None	29	20
C14	0.8	Unknown	3.1	3.9	None	22	20
C15	0.0	S. pneumoniae	3.2	3.3	None	26	20
C16	0.3	S. pneumoniae	1.9	2.2	Partial	30	20
C17	0.5	S. pneumoniae	2.4	2.9	None	32	20
C18	0.2	S. pneumoniae	2.4	2.5	Partial	32	20

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

Table 3. Comparison of group characteristics

	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

Some degree of ossification was present in 19 of the 25 children. In the control group with full electrode insertion, 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 for 1 to 6 mm into newly formed bone before the natural lumen in the scala tympani was reached (Table 5). There were no cases of scala vestibuli insertions, total drill out or circum-modiolar insertions.

In the partial-insertion group, the basal turn was totally obliterated in five children. Drilling for up to 8 mm still did not reach the lumen. 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, the reason for partial insertion was ossification of the apical turns. In one of these cases, some basal ossification was also present, which required limited drilling. Table 2 shows the number of electrodes inserted. In the 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 5. Causative organism and degree of ossification in the partial-insertion group 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. 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.

Speech perception scores of all individuals, expressed in EHL, 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.

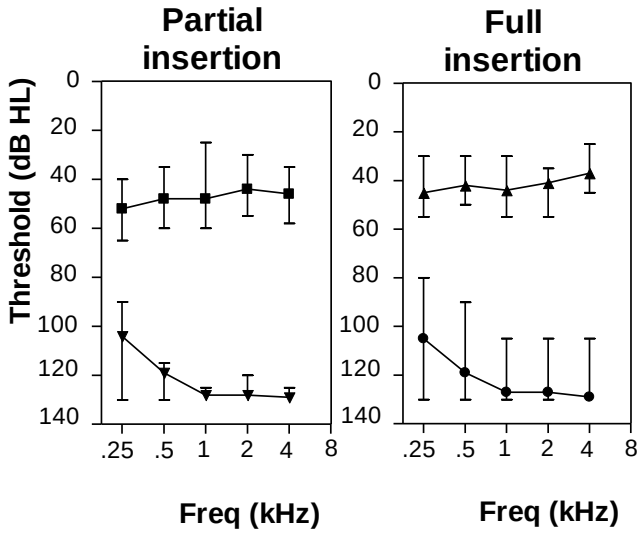

Figure 1. Unaided and aided thresholds

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)

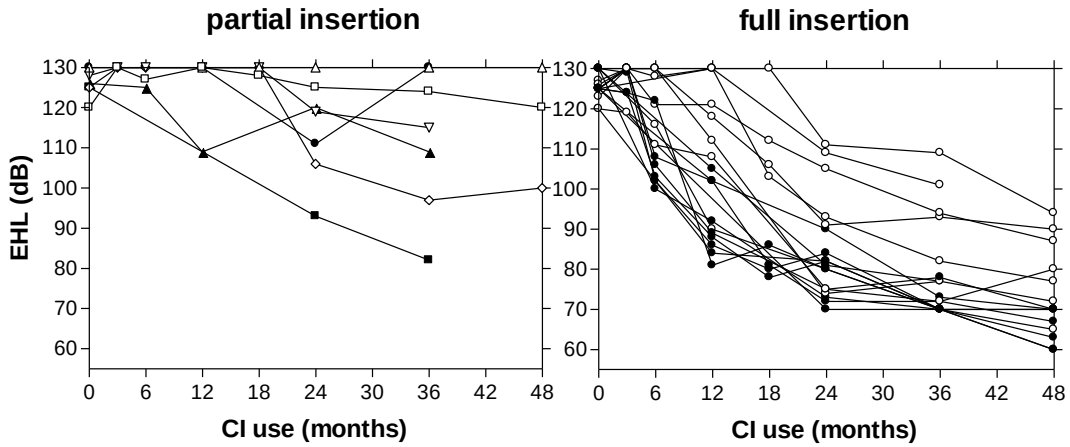

Figure 2. Speech perception expressed in EHL scores as a function of follow-up
Left: children with partial insertion of the electrode array. The symbols refer to the different subjects: □ S1, Δ S2, ▽S3, ◇S4, ● S5, ▲ S6, ■ S7
Right: control group with full insertion. Data from children with duration of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

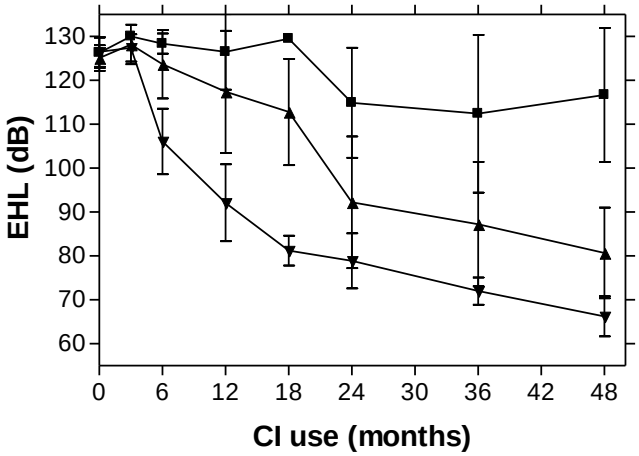

Figure 3. EHL group means and standard deviations as a function of follow-up
The symbols refer to: ■ = partial-insertion group (n=7); ▲= full-insertion control group whose duration of deafness was longer than 3 years (n=8); ▼= full-insertion control group whose duration of deafness was shorter than 3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

During the first 18 months post-implantation, most subjects with partial insertion made little or no progress. After one year of CI use, patient S3 became a non-user (discussed further on): 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. For patients S2, S5 and S6 the open-set speech recognition tests were too difficult: they had zero scores.

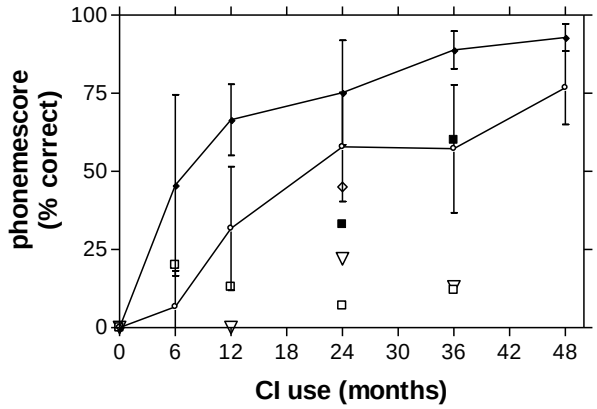

Figure 4. Individual phoneme scores of the children with partial insertion of the electrode array as a function of follow-up and the mean scores of the 2 subgroups with full insertion
The symbols refer to: ♦ = mean (with standard deviation) of the full-insertion control group whose duration of deafness 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

Owing to the limited number of patients with partial insertion, multivariate analysis to determine the influence of the number of active electrodes on speech perception performance could not be carried out. Patient S7 has the highest number of active electrodes and the best EHL score. Although 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 means higher speech perception scores.

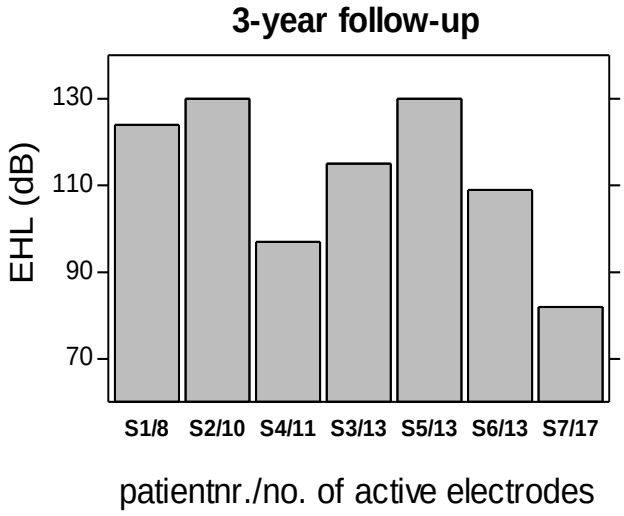

Figure 5. Number of active electrodes in the partial-insertion group (S1-7) versus the EHL at 3-year follow-up

Discussion

Meningitis and deafness

In the present study, 76% of the children with postmeningitic deafness had some degree of ossification, which is in accordance with the 70% mentioned in the literature.[34] 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[35] and Becker et al[36] 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[37] 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. Young et al[38] reported that temporal bone tomography predicted cochlear ossification in 50% of the children found to have ossification intra-operatively. The same rate was found in the present study, but others observed higher sensitivity (100%)[39]. The low sensitivity 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. The wide range in accuracy of CT scanning to detect cochlear ossification might be explained by the use of more or less advanced CT techniques. Nowadays, the imaging modality of choice to evaluate cochlear patency in patients with postmeningitic deafness is MRI, which can detect the presence or absence of fluid in the cochlear coil and visualize fibrosis.[40]

Auditory performance

The purpose of this study was to compare the performance of children with partial electrode insertion to 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.[41] 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 three years of implant use, the speech perception skills of the children with partial insertion showed consistent but slow improvement. Due to 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 one 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. Nevertheless, 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 as described by Mens et al[42] showed no abnormalities. 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. Therefore, on the basis of the integrity measurements, we concluded that the implant was working. 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 a limited number of other groups. Kemink et al[43] did not find any obvious differences in scores on selected speech perception tests over a 6 to 18-month follow-up period between five children with partial insertion and matched controls with full insertion. Kirk et al[44] drew a similar conclusion after comparing the results of five children with partial insertion to age-matched control subjects with full electrode insertion 18 months after the device had been fitted. Long-term results (4 to 5 years) were only reported in 2 of these children with partial insertion: their performance was similar to that of the control group with full insertion. Moreover, their open-set speech perception continued to develop in the long-term. In these studies, the shorter follow-up period and longer duration of deafness in the control groups might explain the discrepancy with the present results: 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 one year of implant use. Mean duration of deafness in the control group in Kirk's study[45] was 4.1 years, compared to five years in the study by Kemink.[46]

Cohen et al[47] found poor speech perception results in most of their cases (adults and children), while Beiter et al[48] concluded from their experiments that the patients (adults and children) with partial insertion of the electrode array benefited from a CI, although not to the same degree as the patients with complete insertion, despite the fact that some of their patients with partial insertion demonstrated open-set word recognition after two years of implant use. The present study results confirmed these findings. Rauch et al[49] observed poor performance in patients with complete ossification that required total drill-out procedures (radical cochleotomy according to Gantz[50]). 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[51] 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 play an additional role in performance variability.

In addition to 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, as mentioned above, the integrity of surviving spiral ganglion cells and auditory nerves is a major concern in determining CI benefit, 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 even these patients became successful CI users.[52] Unfortunately, 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 EABRs and neural response telemetry (NRT) measurements may be helpful to predict neural integrity, the survival rate of spiral ganglion cells and the functional prognosis of cochlear implantation.[53] However, these measurements are still under evaluation and have not yet become fully implemented in CI assessment.

Conclusion

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.

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Chapter 4

Cochlear implantation in the malformed cochlea

Congenital malformation of the inner ear and pediatric cochlear implantation

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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. A cerebrospinal fluid gusher was encountered in two patients and an aberrant facial nerve in another, which did not lead to any complications. The 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 one 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 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 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 paediatric 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 last 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 which doesn't 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 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 total deafness underwent multichannel cochlear implantation at the cochlear implant (CI) centre of the Radboud University Nijmegen in close collaboration with the Institute

for the Deaf in St Michielsgestel. Ten of the 13 children with a follow-up of at least one year were matched with children who had received implants with a normal cochlea for variables that are known to have influence on the performance: age at implantation, duration of deafness and electrode insertion depth. In Table 1 and 2 the most important patient characteristics and surgical aspects are shown. Eleven children with malformed cochlea were considered deaf from birth, and were implanted at an average age of 4.0 years. Patient 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 were implanted 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 were implanted 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. Patient 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 total deafness.

The inner ear malformations were diagnosed with high resolution CT (HRCT) scanning.

Table 1. Patient characteristics and surgical aspects

Patient	Hearing Loss	Vestibular tests	AI (years)	cochlear malformation on CT scan	labyrinthine, IAC or vestibular aqueduct malformation on CT scan	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; AI = age at implantation; CT = computed tomographic; 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 HL	Vestibular tests	AI (years)	CT scan findings	MRI findings	Intra-, and postoperative complications
1b	Congenital	unknown	areflexia	5.1	No abnormalities		
2b	Postlingual [3.2]	unknown, progres	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 otorrhoe
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		

HL = hearing loss; progres = progressive; AI = age at implantation; CT = computed tomographic; MRI = magnetic resonance imaging; SSC = superior semicircular canal; OME = otitis media with effusion

For patients 9 to 13, this was supplemented with magnetic resonance imaging (MRI). The severity of the cochlear malformation was graded based on 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, 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] The reaction of the child that had undergone implantation to sound was commented on in case the perception scores could not be obtained because of a limited follow-up or young age.

Table 3. Pre- and postoperative performance

Patient	**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**
1a	9;0	110-NM-NM-NM	70-75-NM-NM	45-40-40-35	83% (5 yrs)	73% (8 yrs)	
2a	7;2	105-115-110-105	75-65-75-65	45-50-45-40	95% (5 yrs)	89% (6 yrs)	
3a	4;6	80-80-80-75	40-40-30-35	35-40-40-35	90% (1 yr)	88% (4 yrs)	
4a	4;7	115-NM-NM-NM	75-80-80-NM	40-40-35-40	82% (4 yrs)		
5a	4;0	100-NM-NM-NM	80-90-NM-NM	40-40-35-30	80% (3 yrs)		
6a	4;2	NM-NM-NM-NM	65-60-70-65	45-40-40-35	70% (3 yrs)		
7a	2;6	NM-NM-NM-NM	80-75-85-90	55-50-55-50			54% Erber (2 yrs)
8a	2;4	105-120-NM-NM	65-50-70-70	40-40-45-35	78% (2 yrs)		
9a	2;0	110-110-120->120	55-60-75-NM	55-55-65-45			75% Erber (2 yrs)
10a	2;0	NM-NM-NM-NM	95-95-NM-NM	50-40-45-45	40% (1 yr)		
11	1;1	90-105-110-NM	40-45-65-100	45-40-40-30	68% (1 yr)		
12	1;0	NM-NM-NM-NM	90-NM-NM-NM	55-55-55-55			Discriminates sounds
13	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 patient 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; CI = cochlear implant.

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

Patient	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
1b	6;9	NM-NM-NM-NM	100-NM-NM-NM	50-50-50-45	67% (6 yrs)		
2b	4;6	125-135-NM-NM	85->110-NM-NM	30-30-25-30	98% (4 yrs)		
3b	7;5	110-125-125->130	75-70-NM-NM	30-30-35-35	80% (4 yrs)	94% (5 yrs)	
4b	6;7	NM-90-NM-NM	60-55-70-NM	45-40-40-35	55% (3 yrs)	91% (5 yrs)	
5b	5;8	105-NM-NM-NM	85-90->90-NM	40-35-30-30	100% (4 yrs)	64% (5 yrs)	
6b	2;0	120-130-130-NM	95-100-95->100	40-35-35-35			100% Erber (2 yrs)
7b	2;10	NM-NM-NM-NM	100-110-110-NM	45-45-45-45	87% (2 yrs)		
8b	3;11	NM-100-115-120 *	NM-NM-65-85 *	45-45-40-40	90% (3 yrs)	73% (3 yrs)	
9b	4;1	NM-NM-NM-NM	75-75-80-NM	40-35-40-40	23% (3 yrs)		
10b	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, less usual words; 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. Patient 12 and 13 are known with CHARGE-association with typical findings including retarded 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 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 implanted 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 installed for another week. No complications occurred afterwards, nor in any other child.

Patient 10a is presented 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 which had been treated with intravenous antibiotics. HRCT-scanning showed bilateral common cavities, aplasia of the semicircular canals, and a fluid-filled mastoid in the left ear (Figure 1). The internal auditory canals were normal. MRI demonstrated the presence of the eighth nerve and a probable intact cribriform plate (Figure 2). An explorative tympanotomy was performed in the left ear, confirming a small leakage of CSF through the anterior portion of the oval window. The oval window was malformed and in a more inferior position than normal. The leakage was sealed with temporal fascia and tissue glue. As suspected on HRCT-

scanning an aberrant facial nerve was present, 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.

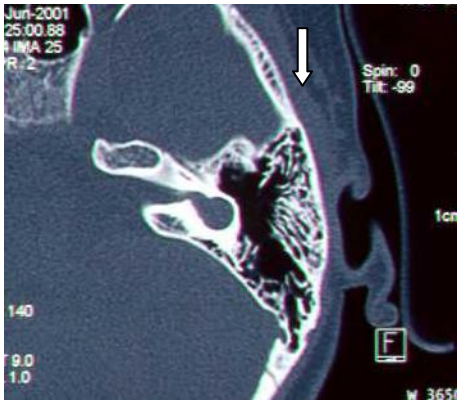

Figure 1. Patient 10a: transversal CT-image, left ear; common cavity, aplastic semicircular canals.

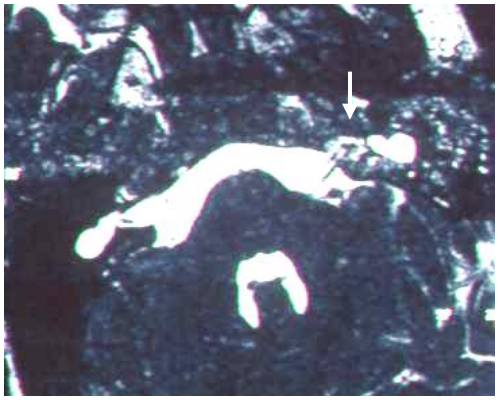

Figure 2. Patient 10a: transversal MRI-image; bilateral common cavities. There is an intact cribriform plate (arrow).

Audiometric results

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 T- and C-levels did not show abnormalities compared to 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 the T-levels approached the limits of the equipment. So, 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 which may be explained by age and follow-up (Table 3 and 4).

Speech perception scores obtained at the most recent audiometric session are shown in Table 3 and 4. At one 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 (patient 6b, 7a, 9a, 12, 13), possibly due to 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 the most important variable 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 one-year intervals postimplantation 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

Foremost important in the technical feasibility of cochlear implantation in profound deaf children with malformed cochlea is to determine whether there is sufficient cochlear lumen for electrode placement and to rule out eighth 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 eighth 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. less than 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 ABR, can give additional information on the neural pathway.[17,19] At our cochlear implant centre, MRI scanning is routinely performed in postmeningitic deaf children and in children with inner ear malformations.

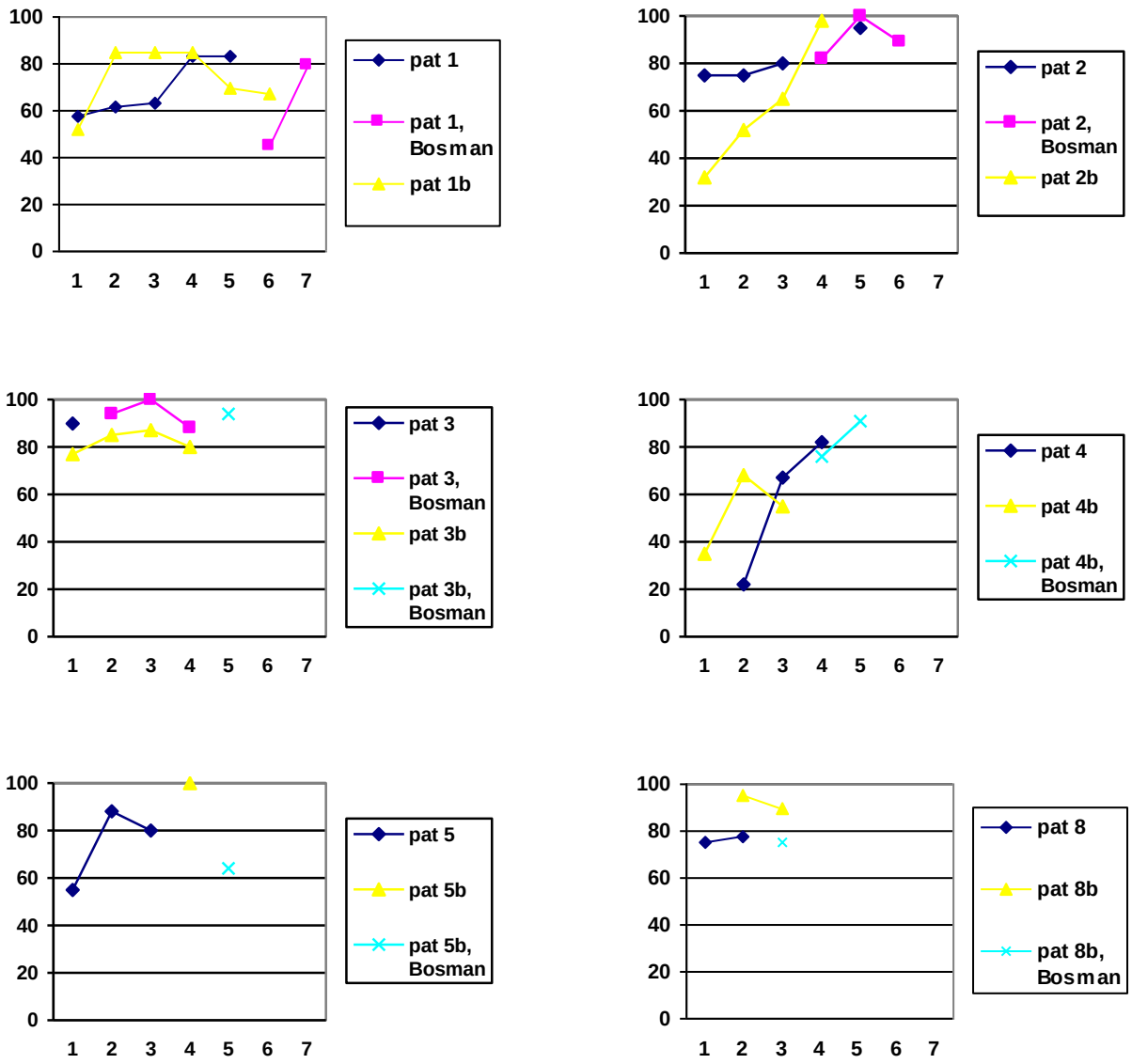

Figure 3. Open set speech perception (phoneme) scores (% correct) of 6 matched pairs.
GN = Gestel/Nijmegen open set phoneme perception test, Bosman = open set phoneme perception test. X-axis; years.

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[54], and noted more frequently in patients with a severe malformation like 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 described the transmastoid

labyrinthotomy approach to common cavity malformations to minimise the risk of injuring an aberrant facial nerve and to have better control of a possible CSF gusher.[8] 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- and MR-imaging 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, pre-operative CT-imaging 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-imaging demonstrated patulous communication in patients in whom CSF outflow did not occur.[12] 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 of 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 a postoperative meningitis.

In Table 5 the surgical results are given of twelve previous studies concerning cochlear implantation in children with malformed inner ears that have included patients with severe cochlear malformations like common cavities and sever cochlear hypoplasia. Including our own data, a total of 81 children with inner ear malformation is shown, including 23 ears with a common cavity deformity and 10 ears with cochlear hypoplasia (one patient was implanted bilaterally). In three of the 7 studies that include 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 present study, an aberrant facial nerve was found in 14 ears (17%), of which (at least) 9 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 gusher was 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 cochlear implant depends on the presence of spiral ganglion cells and cochlear nerve fibres. As the exact location of neural tissue within a severely malformed cochlea like 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 peroperative antero-posterior 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 an 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 Malfor-mation	Age (yrs) at implantation	Follow-up (months)	Thresholds with CI	closed set speech perception	open set speech perception	Comments
Jackler et al. (1987)	CC	5(right ear)*	12	73 dB HL to NM			All implants were single electrode.
	CH	7(left ear)*	2	53 to 68 dB HL			*patient became a non-user as a result of facial nerve
	CC	5	12	62 to 69 dB HL			stimulation. The other ear (left) was implanted 2 years
	CH	9	10	39 to 63 dB HL			later
Slattery and Luxford (1987)	CC	3.5	42°	55 to 60 dB HL	No		°Single electrode implant.
	CH	4.5	42	20 to 30 dB HL	Yes		
	CC	3	2	55 to 80 dB HL	NT		
Molter et al. (1993)	CC	4	10		39% MTS (W)		
Tucci et al. (1995)	CC	3.5	12	Range of all patients:	<10% MTS(W)	35% GASP(W)	Fluctuating thresholds: patients with abnormal cochleae
	CH	4	18	30 to 40 dB HL	70%	10%	may require frequent monitoring of psychophysical
	CH	4	18		<10%		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	3	87	20 dB HL SRT	Yes	56% PBK	Results in patients with inner ear malformations other
	CC	3	9	25 dB HL SRT	No	0%	than CC are comparable to those of other deaf children
	CC	4	8	15 dB HL SRT	No	0%	with CI
Weber et al. (1998)	CC	3.5	22		At 9 months	At 15 months	Slower rate of development compared to normal cochleae
	CC	3.5	7		-	-	
	CH	3.3	19		at 7 months	-	
	CH	4.1	37		at 8 months	-	
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	7.7	24		21% ESP	0% GASP(W)	No statistical difference between malformations and
	CC	3.8	24		8%	0%	controls.
	CC	2.1	24		8%	0%	No statistical difference between mild and severe
	CC	4.8	24		100%	75%	malformations (small numbers!).
	CH	8.5	6		29%	17%	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 table 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 cochlear implant vary enormously. Results after 24 months vary from no speech perception at all to 100% closed set word perception and 75% open set word perception. 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 cochleas 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 times were too small to allow comparison between degree of malformation and performance. 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 cochlea 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] A recent 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 six children with a follow-up of more than 2 years are in mainstream schools and one 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 two 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 implant in a common cavity[20] and tends to have an overall higher incidence in patients with 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 more electrodes have to be deactivated, the patient's performance may be decreased. 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 aetiology of deafness in the paediatric 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 of importance. In this study, own results and reports from the literature combined, the incidence of aberrant facial nerve in inner ear malformations was, after Hoffman[9], confirmed at 17%, which rises to 27% in severe cochlear malformations, and the incidence of CSF gusher at 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. Pre-operative HRCT- and MRI imaging and facial nerve monitoring are essential. Concomitant middle ear problems may lead to potentially hazardous situations as was shown in one of our patients. Per-operative imaging should be considered when implanting a malformed cochlea to rule out IAM insertion.

The performance of the 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 like age at implantation and mode of communication. The studies should provide detailed information concerning the anatomy of the inner ear. To obtain this knowledge is especially important in counselling the parents before implantation.

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5.1

Cochlear implantation in otosclerosis

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

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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, CT scans, surgical findings and complications, and the occurrence of postoperative facial nerve stimulation.
Study design: retrospective, multicentre study

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

Results: The CT imaging 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, surgically problematic insertion of the electrode array and facial nerve stimulation. In four patients revision surgery had to be performed. Thirty-eight percent (20 of 53) of 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.[55] In adults with acquired deafness, the aetiology 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. In order to acquire more insight into these matters, a multicentre study was undertaken. Thus, a relatively large number of cochlear implant 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. The type of hearing loss may be conductive (CHL) as in fenestral otosclerosis.[56] In addition there may be a progressive sensorineural hearing loss (SNHL) causing a mixed hearing loss pattern. In far advanced otosclerosis or retrofenestral otosclerosis pure SNHL may exist.[57]
Otosclerosis occurs more frequently in Caucasians and usually presents between the ages of 15 to 45 years.[58] 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.[58] 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.[59,60] Family linkage studies have identified regions on chromosome 15, 7 and 6 that can be ascribed to this disorder.[61] Based on the assumption that otosclerosis is an inherited collagen disorder, otosclerosis has historically been associated with other connective tissue disorders like osteogenesis imperfecta.[58,62] However, some clinico-genetic and histopathological studies showed that otosclerosis is not a localised form of osteogenesis imperfecta.[63,64]
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). Some suggest that the sclerotic phase may not be a healing process following the spongiotic phase, but that it is the first stage of the disease.[65] 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[66] or by narrowing of the cochlear lumen with distortion of the

basilar membrane.[67] Long-term follow-up studies suggest that about 10% of ears with otosclerosis and CHL also develop SNHL.[68,69]

High resolution computed tomography (HRCT) using sections at 1 mm increments is at present the imaging modality of choice for the assessment of bony images such as the osseous labyrinth, labyrinthine windows and cochlear capsule. In CT scanning resorption of bone results in areas of decreased density, lucent zones, which may give the impression that unusual canals and ducts exist. Abnormal sclerosis is sometimes difficult to evaluate because of the subtle changes. 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.[70] Sclerotic foci cause abnormal irregularity and narrowing of the cochlear turns, best evaluated at the basal turn.[71]

Management of patients with otosclerosis and severe or profound HL is stapedectomy or stapedotomy and hearing aid amplification. Further, there have been some studies that demonstrated that sodium fluoride reduces the rate of SNHL.[72,73] If treatment fails, the patient may become a candidate for cochlear implant (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 cochlear implant 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 four cochlear implant centres in the Netherlands and United Kingdom, University Hospital Birmingham, Manchester Royal Infirmary, University Medical Centre Nijmegen and University Medical Centre Utrecht, were searched for patients with otosclerosis. Sixty-one 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 author. Included in the study

were patients with either retrofenestral and/or fenestral otosclerotic lesions on CT scan, normal CT scans but 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, fifty-three 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 well as 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 pre-operative work-up 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).
The patients were divided in two groups: patients with duration of progressive hearing loss in their implanted ear less than 10 years before they eventually became profoundly deaf (defined as rapidly progressive hearing loss) and patients with duration of progressive hearing loss longer than 10 years (defined as slowly progressive hearing loss) (Figure 2).

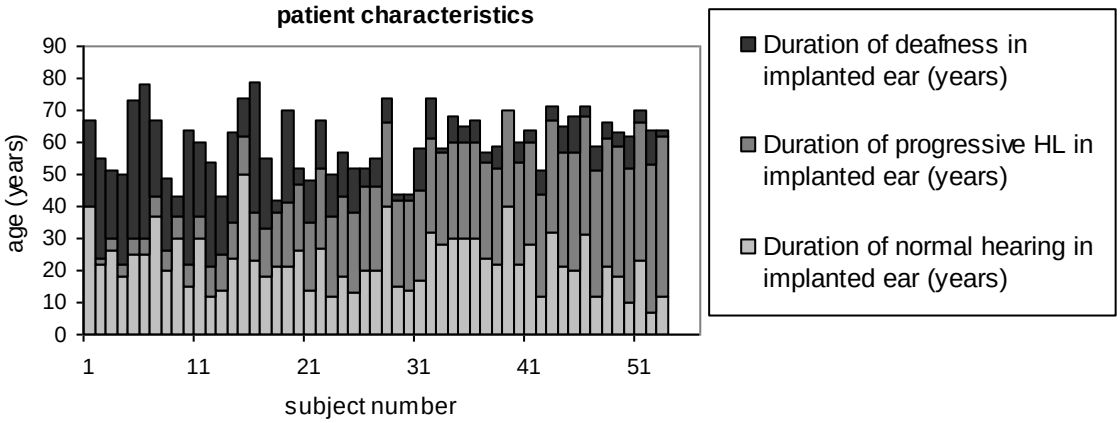

Figure 1. Patient characteristics

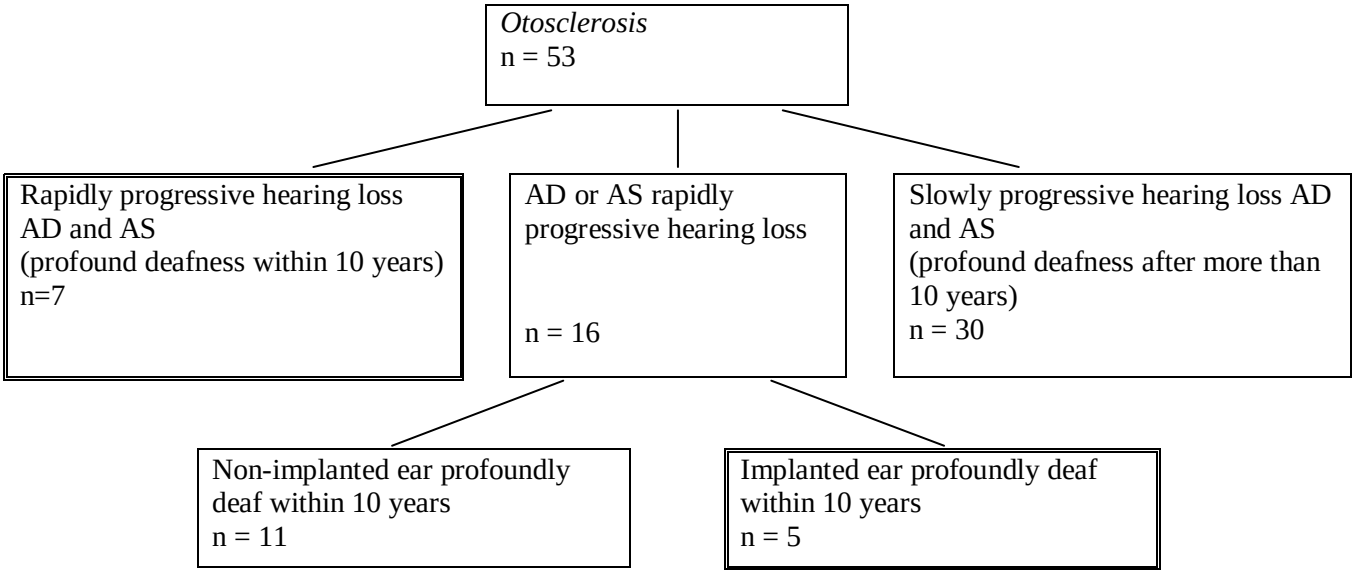

Figure 2. Numbers of patients with rapidly and slowly progressive hearing loss in the implanted and non-implanted ear. The boxes with double lines represent the patients who received an implant in an ear that was deafened within 10 years: the rapidly progressive deaf patient group (n=12)

As part of the standard evaluation for cochlear implantation of all cochlear implant 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 cochlear implant centre in 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. Following Lindsay's[74] 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 (mild or severe 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 (kappa 0.77). Most disagreement (4 of 5 ears) concerned stage 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, gender, surgical problems and the occurrence of FNS.

In all but one patient (implanted with UCH rnid Mk2 single channel device) multichannel implant systems were used. These comprised 6 Clarion devices (CI, CII, Enhanced Bipolar, HiFocus), Advanced Bionics, Sylmar, CA; 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

Of the 53 patients with profound hearing loss and a clinical diagnosis of otosclerosis, 12 patients received an implant in an ear that was deafened within 10 years (Figure 2). These 12 patients were compared to the 41 patients who received an implant in an ear that had suffered a progressive hearing loss for a period longer than 10 years. Group characteristics are summarized in Table 1. Both groups developed hearing loss at the same age (P=0.29) and were implanted around the same age (P=0.48). The patients with rapidly progressive hearing loss had longer duration of profound deafness (Figure 1).

Outcomes on the prevalence of a history of stapes surgery in both groups are similar. Eight (67%) of the patients implanted in rapidly deteriorated ears had previously undergone stapes surgery, the first surgical procedure took place at a mean age of 30 years, while 25 (61%) of the patients implanted in slowly deteriorated ears had undergone stapes surgery (χ^2=0.13; df=1; P=0.7) at a mean age of 33 years (Mann Whitney U = 73.5; P= 0.5). None of the ears deafened after stapes surgery (n=5) has later been implanted.

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). In the group with rapidly progressive hearing loss 43% had a positive family history (3 out of 7) and in the group with slowly progressive hearing loss 52% (16 out of 31). These results are not significantly different (χ^2=0.18; df=1; P=0.7). In both groups, missing data occurred equally (χ^2=1.4; df=1; P=0.2).

For both groups, the numbers of patients with a certain degree of tinnitus are registered in Table 1. Nine patients declared to suffer from tinnitus that was worse in one ear than in the other. In these cases of asymmetrical tinnitus, the tinnitus in the implanted ear has been used for analysis. Chi-square tests did not reveal any difference in the prevalence of tinnitus between the groups with rapidly or slowly progressive hearing loss.

Table 1. Characteristics of 12 patients with rapidly progressive hearing loss (rapid: <10 years between onset of hearing loss and onset of deafness) and of 41 patients with slowly progressive hearing loss (slow: >10 years between onset of hearing loss and onset of deafness)

Patient characteristics (in years)	rapid (n= 12)	slow (n= 41)
Mean age at onset HL implanted ear	25	22.2
Mean age at onset HL non-implanted ear	27.6	25.0
Mean duration of progressive HL implanted ear	5.2	28.9
Mean duration of progressive HL non-implanted ear	15.2	21.7
Mean age at onset deafness implanted ear	30.2	51.3*
Mean age at onset deafness non-implanted ear	40.2	44.7
Mean duration of deafness implanted ear	29	10.0
Mean total duration of hearing loss implanted ear	34.2	38.9
Mean age at implantation	59.3	61.2
Severity of tinnitus (no. of patients)	n=12	n=40
Generally absent	3	12
Occasional, not bothersome	6	21
Definite	3	7

Abbreviations: HL = hearing loss; * Significant difference

CT scanning

The results of the 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 3a) and 55 (52%) had retrofenestral lesions with or without fenestral involvement (type 2, Figures 3b and 3c). 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 3d). In such severe cases assessing fenestral involvement was found very difficult.

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 Type 2a: double ring effect Type 2b: narrowed basal turn Type 2c: double ring and narrowed basal turn	55 (52%) 26 (25%) 4 (4%) 25 (23%)
Type 3	severe retrofenestral (unrecognizable otic capsule), with or without fenestral involvement	27 (25%)

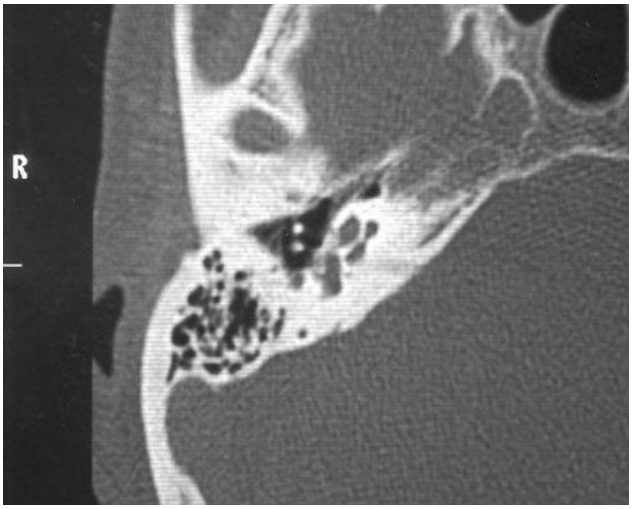

Figure 3a Anterofenestral focus and thickened footplate: type 1 (*case 35*)

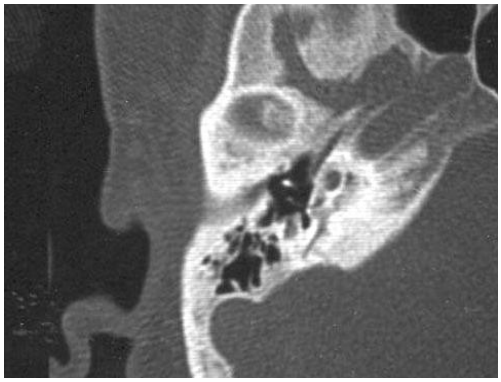

Figure 3c Double ring effect and a narrowed basal turn: type 2c (*case 29*)

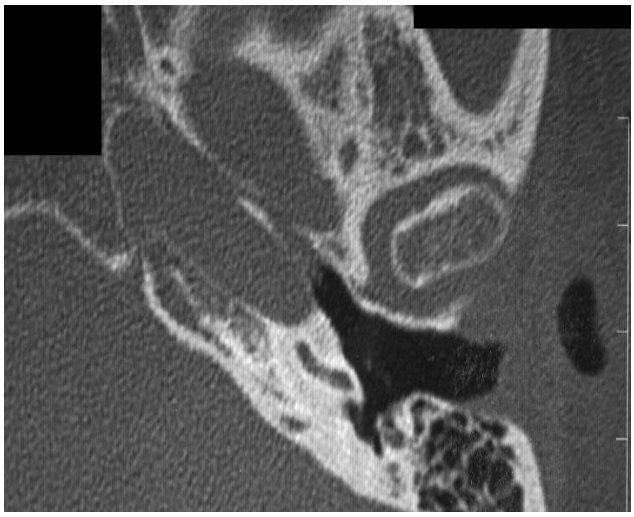

Figure 3b. Double ring effect or "halo" (hypodensity around the basal turn), no narrowing of the basal turn: type 2a (*case 41*)

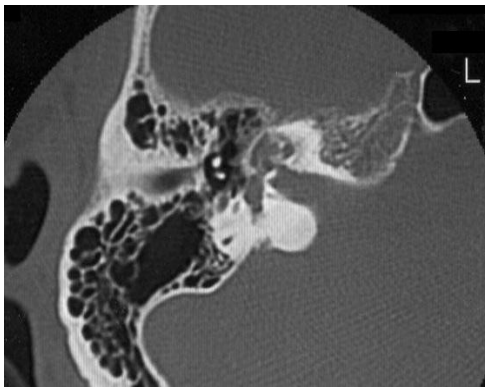

Figure 3d. Severe retrofenestral involvement of otosclerosis, no otic capsule recognizable: type 3 (*case 2*)

In 11 patients (20%) the severity of otosclerosis was asymmetrical: 3 patients had one ear without signs of otosclerosis, while 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 otosclerosis of the implanted ears as categorized in 3 types was not significantly different from the non-implanted ears (Figure 4a).

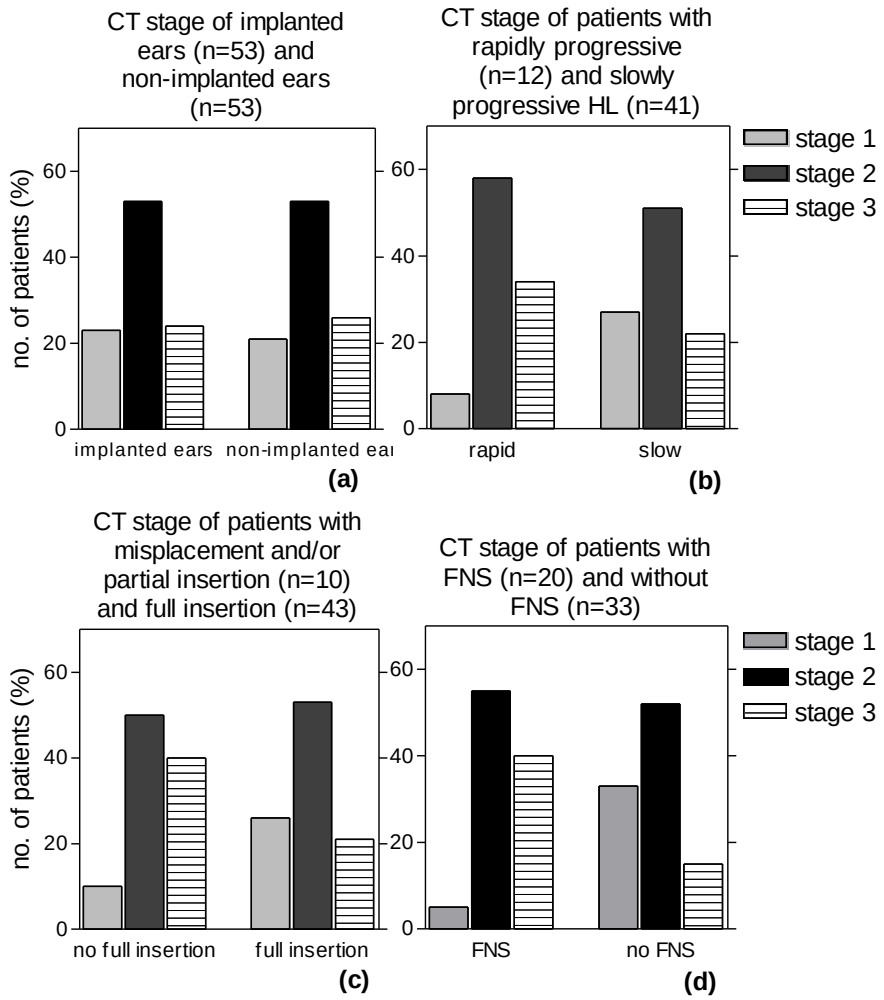

Figure 4. Comparisons of CT stages between different groups

Figure 4b depicts the distribution of CT findings in the groups of patients with rapidly and slowly progressive hearing loss in the implanted ear. Although the percentage of patients with non-affected ears plus type 1 ears, seems higher in the group with slowly progressive hearing loss, this difference is not significant (χ^2=1.8; df=1; P=0.18). Neither was there any difference between both groups in the prevalence of type 2 and 3. The comparison of CT findings of ears with rapidly and slowly progressive hearing loss (30 respectively 76 ears) gave similar results. 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.

For the whole group of patients no significant correlation was found between the extent of otosclerosis on CT scan (type 1, 2 and 3) and the total duration of hearing loss (time between onset of hearing loss and time of implantation), (Spearman r=0.08, P=0.57). The extent of otosclerosis on CT scan 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 age at implantation nor gender (Table 3).

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

	AOHL	AOD	DoHL	DoD	AImp	gender	stapes
Spearman r	-0.30	-0.34	-0.12	0.20	-0.26	-0.08	-0.07
P value (two-tailed)	0.03	0.01	0.39	0.16	0.06	0.58	0.60
significant	yes	yes	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; 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 program. 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. The other 3 revision cases will be discussed in more detail below.

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.[75] In all but two of the patients (*case 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 5). 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 1 partial insertion and two misplacements. In Figure 4c the CT scans of patients with partial insertion and/or misplacement are compared to the scans of patients with full insertion. Cochlear pathology (type 2 and 3) seems more extensive in the group with partial insertion and/or misplacement. Chi square tests however, do not show any significant differences in prevalence of type 1, 2 or 3 between both groups.

Table 4. Partial insertions (partial), misplacements (misplaced) of electrode (e) arrays and revision surgeries in 10 patients. c.l. = contralateral

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

Few other complications occurred during surgery. In one patient (*case 3*) due to ossification the scala tympani could not easily be identified and the anterior vertical semicircular canal was opened instead, before a correct cochleostomy eventually took place. Two Clarion positioners were partially inserted; a postoperative CT scan showed a bent-over tip of the electrode array in one. In one patient (*case 7*) an oval window fistula was noted and closed with muscle. Postoperative complications were not seen.

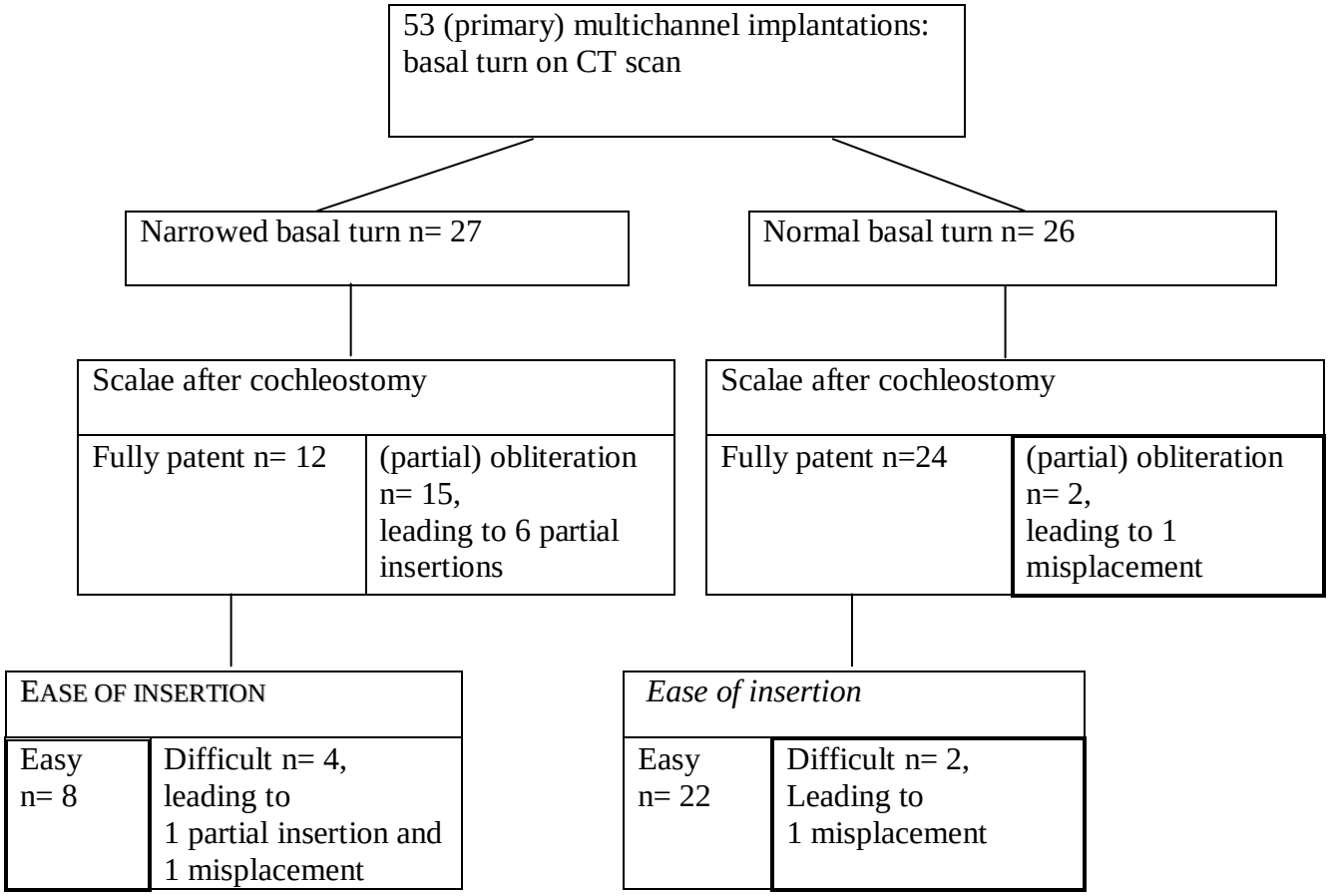

Figure 5. The presence or absence of a narrowed basal turn on CT scan, confirmed at surgery by observation after cochleostomy and ease of electrode array insertion. The boxes with bold lines represent the patients with false negative scan results (n=4), boxes with double lines represent the patients with false positive scan results (n=8).

Revision surgery 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.

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. ENG showed areflexia AD and hyporeflexia AS. Because her hearing did not improve after the stapedectomy, two 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. 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 was re-operated: the array was taken out and by drilling the cochleostomy was widened. A heavily obstructed basal turn emerged. No natural lumen could be reached by drilling and four electrodes were placed in the drilled canal. Peroperatively 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 two and a half 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 AS by the age of 52 and AD 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: due to 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 years, range 0.5-13 years), 20 patients (38%) developed FNS when the implant was activated at various time intervals. Two patients were implanted with a Clarion device; the remaining patients all had a Nucleus device. Five patients had partial insertions of the electrode array. The electrodes causing facial nerve stimulation (FNS) in the patients with full insertion of the active electrodes of a Nucleus electrode array are shown in Figure 6. When comparing the CT scans of patients with FNS to those of patients without FNS (Figure 4d), type 1 otosclerosis is 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, whether or not temporarily. In one of the cochlear implant 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 eight electrodes remained active. In the two years of fluoride treatment the FNS remained stable. The other patient stopped using fluoride because the side effects of the treatment.

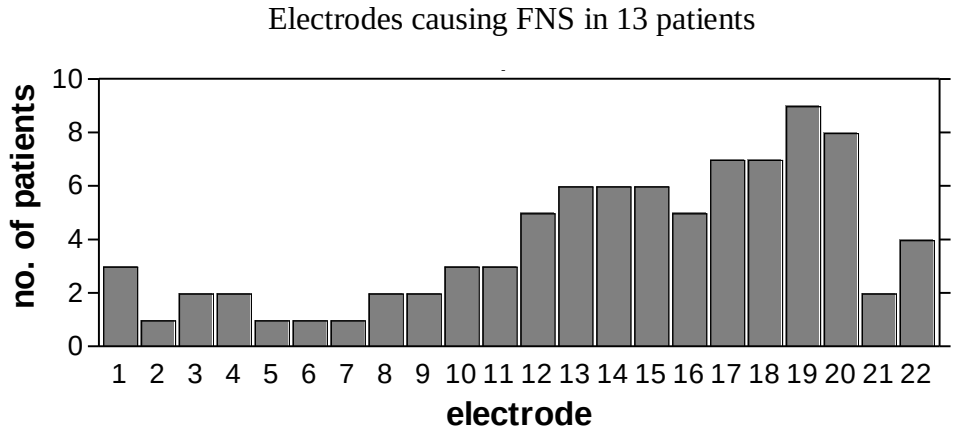

Figure 6. Electrodes causing facial nerve stimulation (FNS) in 13 patients with a multichannel Nucleus device with a complete insertion of the electrode array

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 four implant centres up to the end of 2002 (n=788), 6.7 per cent has otosclerosis as the aetiology of deafness. So, otosclerosis is not a rare indication for cochlear implantation, although far advanced otosclerosis was once considered a contra indication for cochlear implantation.[76]

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.[77] In this group of otosclerosis patients who eventually received a CI, 38% was female, and the mean age at onset of hearing loss in the implanted ears was 23.6 years (in non-implanted ears 26.3 years). Thus, a female dominance in this selected group of otosclerosis patients is no longer present. In accordance to the eight otosclerosis patients receiving a CI in the study by Ruckenstein et al.[78] 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).[77] In the present study 50% of the patients had a positive family history. Possibly when data on family history are systematically collected such as 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 asymmetrical. 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, to distinguish 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 less than 1 mm)[79], or the chemical process in otosclerotic lesions damaged neural elements in the cochlea. Patients may have SNHL before otosclerosis becomes apparent on CT images and bilateral hearing loss may be present with unilateral or asymmetric CT findings.[80] 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.[81]

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[82] 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.[83] As shown in Figure 4b, the duration of progressive hearing loss, rapidly or slowly progressive, 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.[84] 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 six 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.[85] in one of eight otosclerosis patients a drill-out of the basal turn had to be performed. No partial insertions occurred. Despite ossified scala tympani in two patients, full insertions were achieved by insertion of the scala vestibuli. In the present study, in 17 (32%) patients a partially or complete obliteration of the basal turn was observed during surgery which 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.[86] Obstruction of the scalae may not be evident on CT scan, especially at the more apical turns. Magnetic resonance imaging (MRI) has been useful in the assessment of the membranous labyrinth with it's neural elements and of the cochlear lumen prior 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.[87]

Revision surgery has been necessary in four patients of which one patient unfortunately had three operations after the primary implantation. Two of the three patients with a misplaced electrode array were revised. 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.[88] 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 cochlear implant population.

Facial nerve stimulation

Facial nerve stimulation after cochlear implantation has been reported with a variable incidence. FNS may be a serious problem even leading to explantation.[89] The incidence of FNS in the general CI population as reported in the literature varies from 0.9% [90], 3% [91] to 14.6% [92] and is more frequent in patients with otosclerosis and otosyphilis.[93-97] 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. Since 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.[98] Both mechanisms are probable in otosclerosis.[99-101] Sometimes electrodes positioned at the round window are responsible for FNS at the tympanic segment or even vertical segment of the facial nerve.[102] 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.[103] Bigelow et al.[104] 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 seven patients with FNS the electrodes causing stimulation most frequently (in more than 2 out of 7 (>29%) patients) ranged from electrode 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 electrode 12 to 20 (Figure 6). The difference is explained by the fact that in the present study calculations were based on the number of inserted electrodes plus a variable number of 0-10 supporting rings measured from the cochleostomy, whereas in the previous study in all cases all 10 supporting rings were included in the number of inserted electrodes. 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.

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 pathology than in patients without FNS, although the difference was not statistically significant. In the preoperative counselling 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 cochlear implants 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. 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. 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 percentage of patients was confronted with facial nerve stimulation mainly caused by the distal electrodes which must be discussed with patients preoperatively.

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Chapter 5.2

Cochlear implantation in otosclerosis

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

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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.

Study design: retrospective, multicentre study

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 aetiology of deafness (5;8-10). The relation between speech perception scores and the aetiology of deafness was reported to be weak (8). However, aetiology 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 counselling purposes.

In order to draw firm conclusions, a sufficiently large number of patients must be available. In the present study, a retrospective multi-centre 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 cochlear implant 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 four 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 four 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 (Nucleus 22 and Clarion I) and the more recent devices (Nucleus 24 and Clarion II) between the four centres (Kruskal-Wallis test, P=0.52).

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 four centres (Kruskal-Wallis test, P=0.67).

Table 1. Number of implanted devices per cochlear implant centre

Device type	Nijmegen (13 patients)	Utrecht (9 patients)	Birmingham (17 patients)	Manchester (14 patients)
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

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 four 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 four 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 four CI centres were compared with respect to demographics and pre-surgical audiological characteristics, CT scan results and the types of implants used. For further analysis, the patients were grouped into poor and good performers, based on 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 ‘good’ 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 four 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 four centres were considered to be largely comparable.

Preoperative evaluation data

At all four 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 four 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 four 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 four centres (Kruskal-Wallis tests; P=0.87).

Table 3. Patient characteristics of implanted ears per cochlear implant 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 = mean age at onset of progressive hearing loss (years); DoHL = mean duration of progressive hearing loss (years); A@OD = mean age at onset of deafness (years); DoD = mean duration of deafness (years); A@Impl = mean age at implantation (years); 25% = 25th percentile; M = median; 75% = 75th percentile; P-value on Kruskal-Wallis test (Sign<0.05)

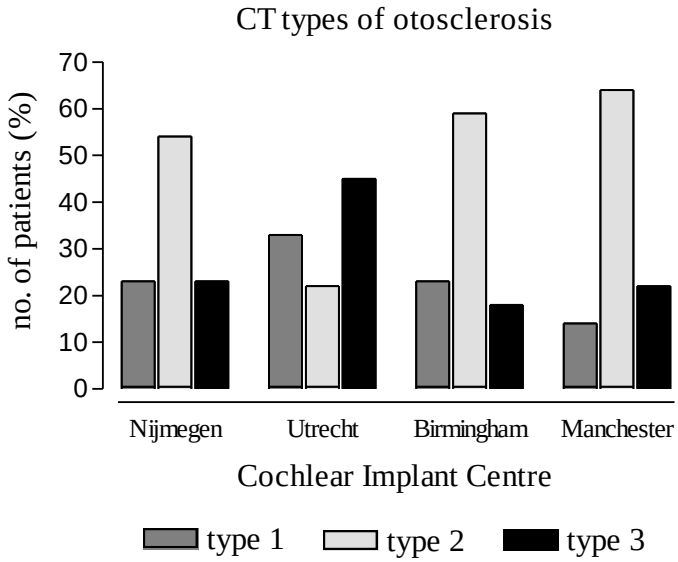

Figure 1. Severity of otosclerosis as categorized in 3 types per Cochlear Implant Centre

Categorization according to patient performance

Phoneme scores

Phoneme scores were available from 19 out of the 31 English-speaking patients on the AB monosyllable test and from 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 "good performance subgroup" (group 1) or the "poor performing subgroup" (group 2). Patients with a phoneme score at 9 months follow-up of higher than 40% (n=24) were categorized as good performers; 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).

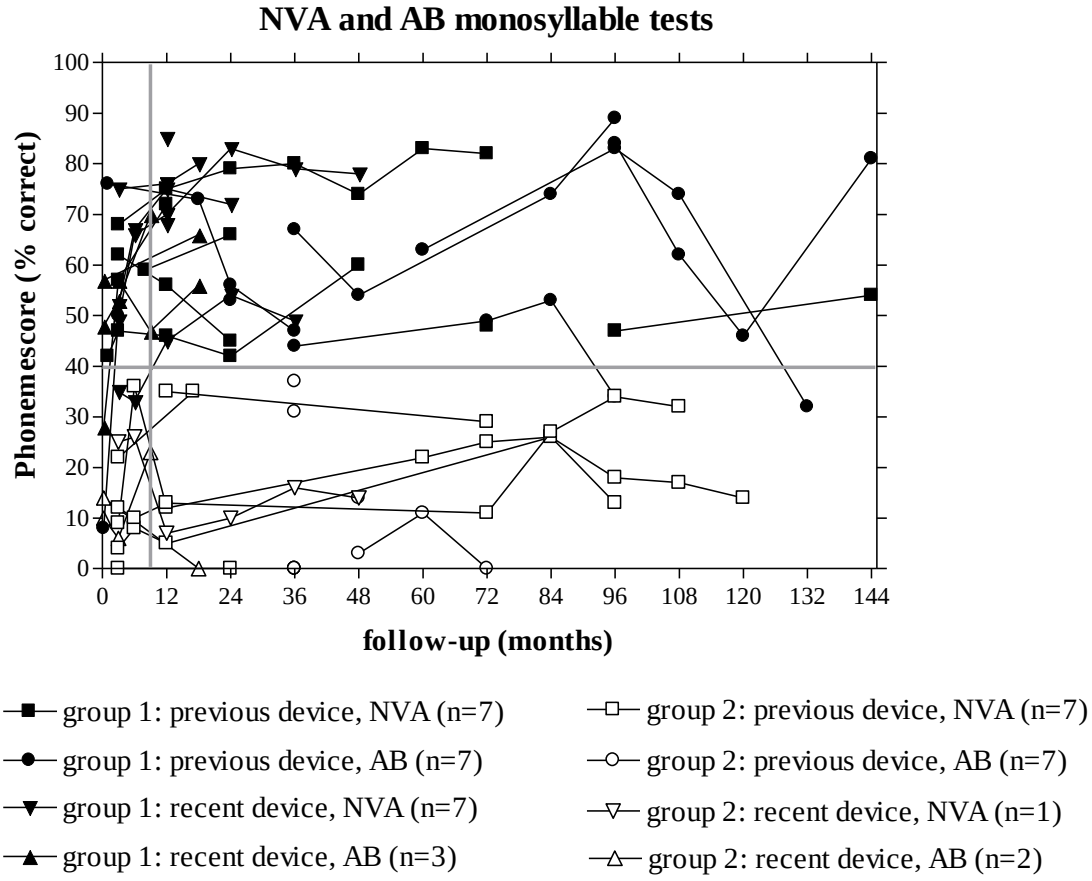

Figure 2. Phoneme scores of 19 English-speaking patients on the AB monosyllable test and of 22 Dutch-speaking patients on the NVA monosyllable test

Group 1 and 2 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 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 good 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 (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, but again statistical significance was not reached in these groups (Fisher's exact test, P= 0.14, 0.10 and 0.06, respectively).

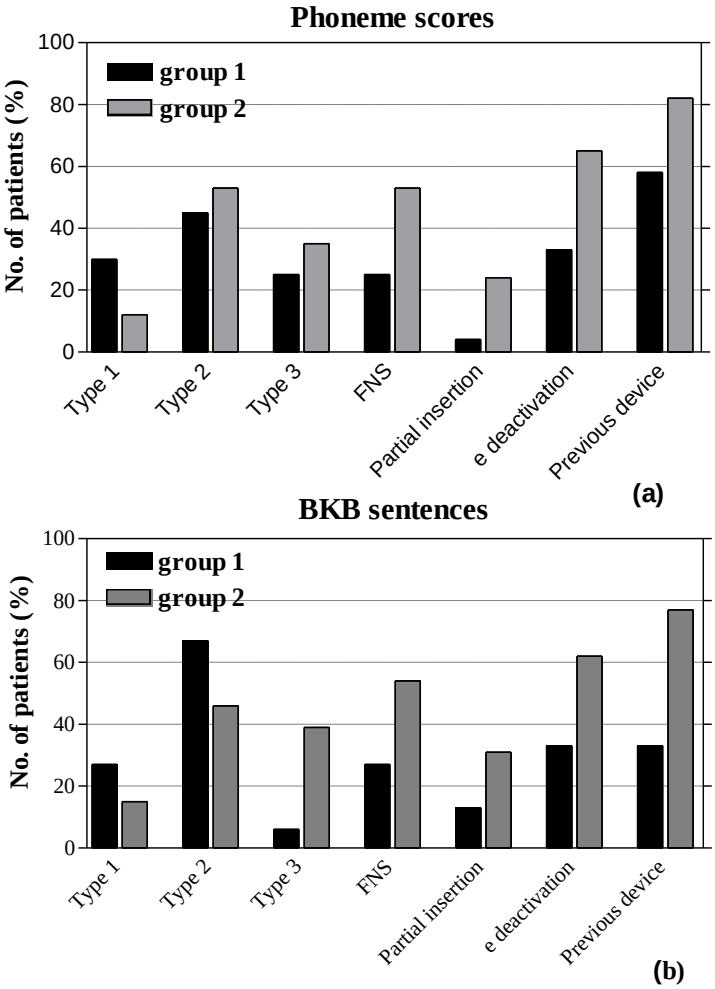

Figure 3. Comparisons between the good (group 1) and poor (group 2) performers on NVA and AB monosyllable test results (Figure 3a) and between the good (group 1) and poor (group 2) performers on BKB sentences test results (Figure 3b)

BKB sentences test scores

BKB sentences test scores had been obtained from 28 English-speaking patients. Data from the two 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 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).

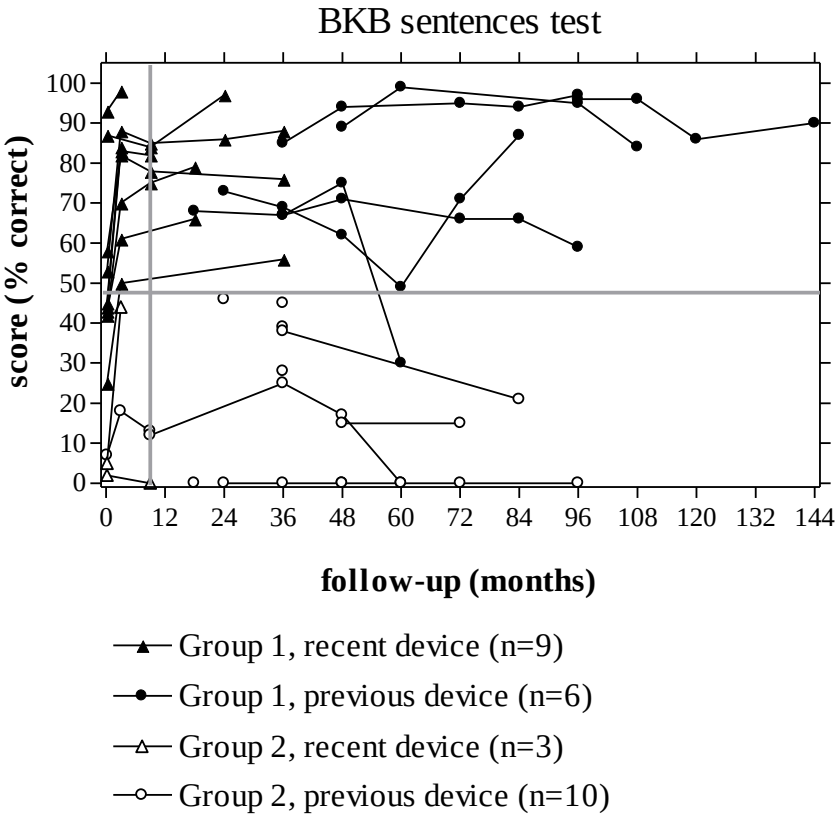

Figure 4. Scores on the BKB sentences test of 28 English-speaking patients

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 old 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 four different CI centres. Similarities in demographic data, pre-operative CT scans and audiological data meant that the patients from the four different centres could be considered as one group. The pre-operative audiological data reflected that all four 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 good and poor performers. No differences were found in demographic factors between the poor en good 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 good performers comprised factors directly related to the disease (extent of otosclerosis on the CT scan, non-auditory 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 good 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.

Conclusion

A previous paper showed that 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 percentage of patients was confronted with facial nerve stimulation 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 counselling, 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.

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Chapter 6

Cochlear implantation in Osteogenesis Imperfecta

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

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Summary

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

Methods: 3 patients with OI were retrieved from the 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 OI, 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 non-auditory stimulation. Even after reimplantation, non-auditory sensations lead to case 3 becoming a non-user. 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 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 OI, 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 FNS requiring frequent refitting. Despite the electrophysiological changes, Case 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 [Clark, 2003b]. Numerous factors have been related to postoperative performance as reflected in speech perception tests [Clark, 2003a]. 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. Aetiology of deafness has also been recognized as a factor of influence on performance [Battmer et al., 1995; Blamey et al., 1996]. 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 aetiology.

At the Nijmegen Cochlear Implant Centre, 3 patients with a rare bone disease, Osteogenesis Imperfecta (OI), have been enrolled in the CI rehabilitation program. 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 20.000 subjects is maintained by a high rate of new mutations in most families [Ross et al., 1993]. 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. [1979], 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 [Nager, 1988].

Table 1. Classification of Osteogenesis Imperfecta

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

* = not applicable because of intra-uterine 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 [Tabor et al., 1990]. 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 [Ross et al., 1993; Tabor et al., 1990]. 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 [Ziyeh et al., 2000]. 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 [Rotteveel et al., 2004]. 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 [Garretsen et al., 1997]. 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 [Pedersen, 1985; Tabor et al., 1990]. Pure SNHL in OI is rare (10%) [Pedersen, 1985]. The hearing loss usually begins in the late teens, with the sensorineural component appearing in the third decade [Stewart and O'Reilly, 1989]. It gradually leads to profound deafness, tinnitus, and vertigo by the end of the forth to fifth decade [Pedersen, 1985]. As the hearing loss will progress to deafness in a varying reported amount (2% [Garretsen et al., 1997] - 11% [Pedersen, 1984]) 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 non-acoustic nerve stimulation like facial nerve stimulation (FNS) and a lesser frequency specificity from multichannel stimulation. To evaluate this, objective electrical and electro-physiological 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 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 T1- and T2-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) [Rotteveel et al., 2004]. 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.

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

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

Hypertrophic or demineralised 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

Neural response telemetry (NRT) has been widely used intra- and postoperatively to measure electrically evoked compound action potentials (ECAPs) in CI subjects [Abbas et al., 1999; Dillier et al., 2002] using a 'masker electrode /probe electrode -stimulus paradigm' stimulating the *same* electrode. The ECAP threshold measured by NRT is referred to as T-NRT [Dillier et al., 2002].

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 [Cohen et al., 2004; Eisen and Franck, 2005]. 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 [2000]. 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 [Bosman, 1998].

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.

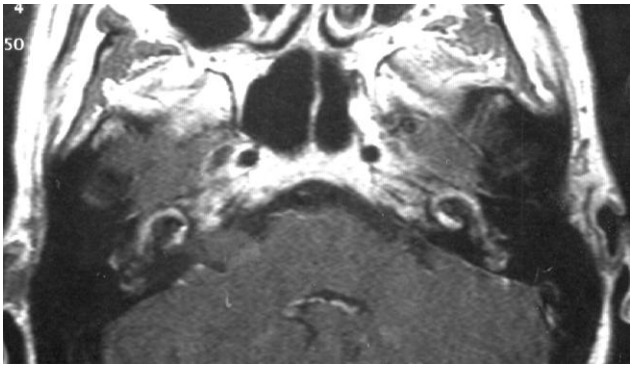

Figure 1a. Preoperative MRI (T1 weighed with contrast) of the petrosal bone (transversal view): pericochlear enhancement

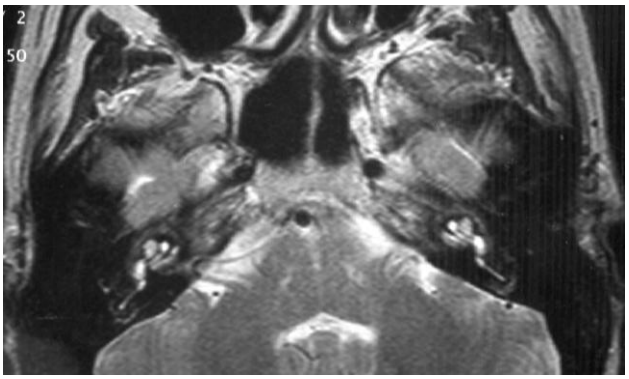

Figure 1b. Preoperative MRI (T2 weighed) of the petrosal bone (transversal view): cochlea and labyrinth are both patent, on the right more eminent than the left side. There is a pericochlear high signal.

Figure 1. Imaging in case 1

The 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. 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 the 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.

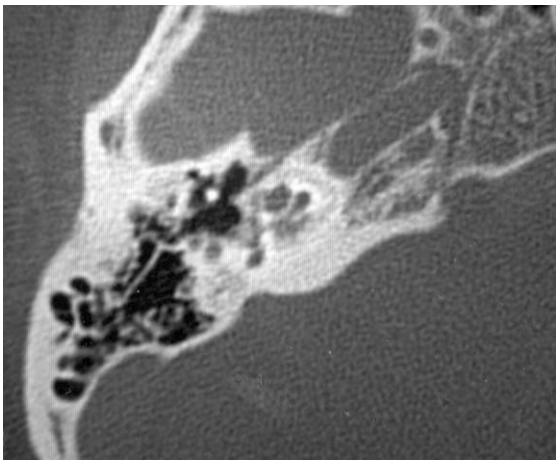

Figure 2. Imaging in case 2; preoperative CT scan right ear (transversal view): type 2c (pericochlear lucency -double ring effect- and narrowed basal turn)

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
Szilvássy 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, e15-18 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 1 / clinical diagnosis	Progressive mixed HL, 2 years	Fenestral abnormalities and pericochlear lucencies ADS; 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 1 / 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 C1 array.	FNS and other, severe discomfort for which all e's switched off	-

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 half to three-quarters 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 scan demonstrated a loss of architecture of the cochlea, demineralization but patent scalae of the cochlea, especially at the right ear (Figure 3a, Table 3).

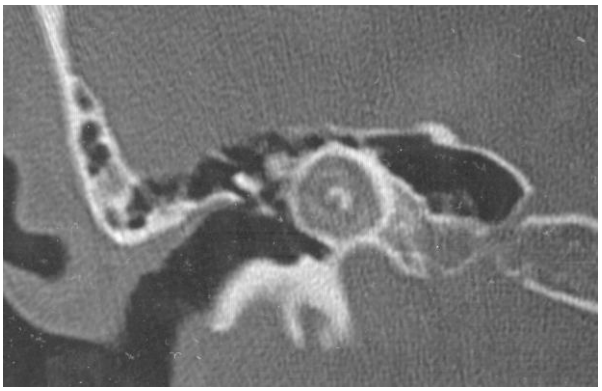

Figure 3a. Preoperative CT scan right ear (coronal view): type 3 (unrecognizable otic capsule)

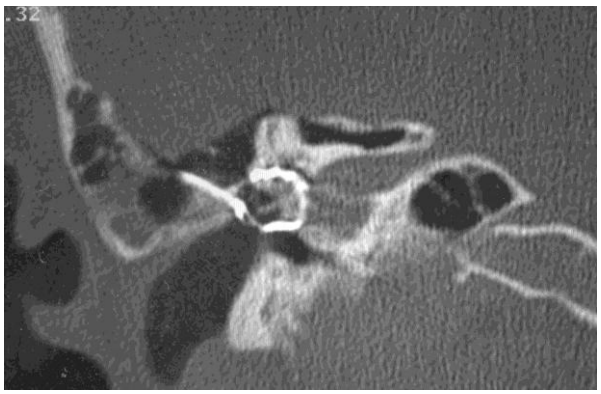

Figure 3b Postoperative CT scan right ear (coronal view): the electrode array is in the lateral semicircular canal

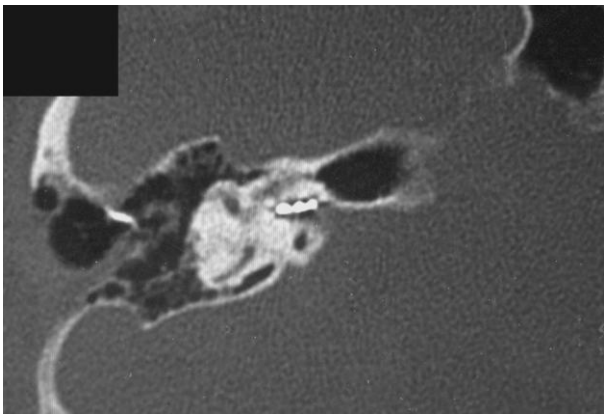

Figure 3c. Postoperative CT scan right ear (coronal view): the electrode array penetrates the internal auditory canal

Figure 3. Imaging in case 3

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 hyperaemic. 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 (electrode 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. Non-auditory 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 three months time the number of usable electrodes was reduced to two, despite frequent refitting. Eventually, this patient became a non-user. 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. Electrode 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 from 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.

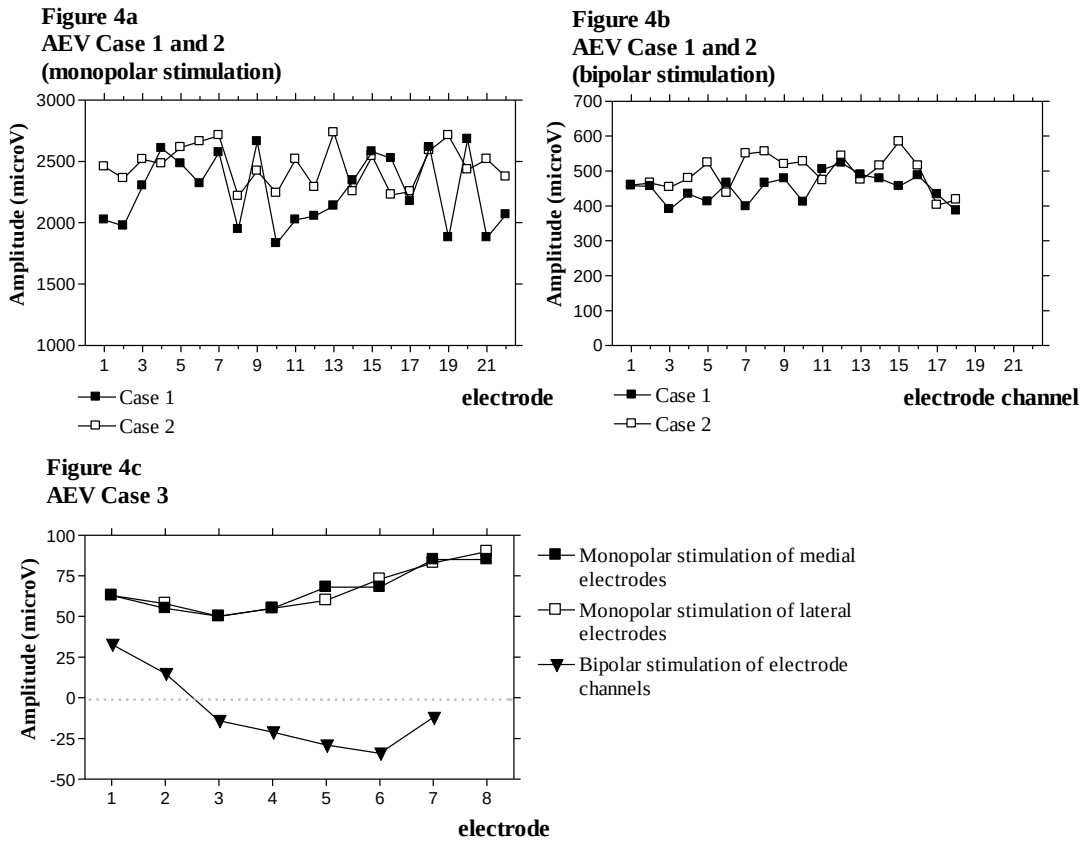

Figure 4. AEV measurements obtained with monopolar (MP) and bipolar stimulation

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 (electrode 4, 11 and 22, respectively). Stimulating electrode 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

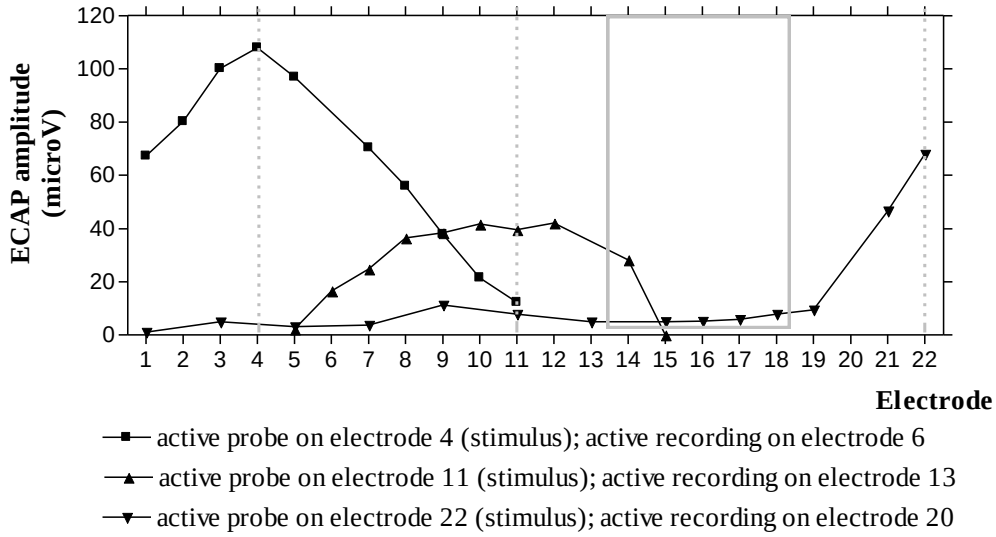

Figure 5b:
Spatial Spread of Neural Excitation in Case 2

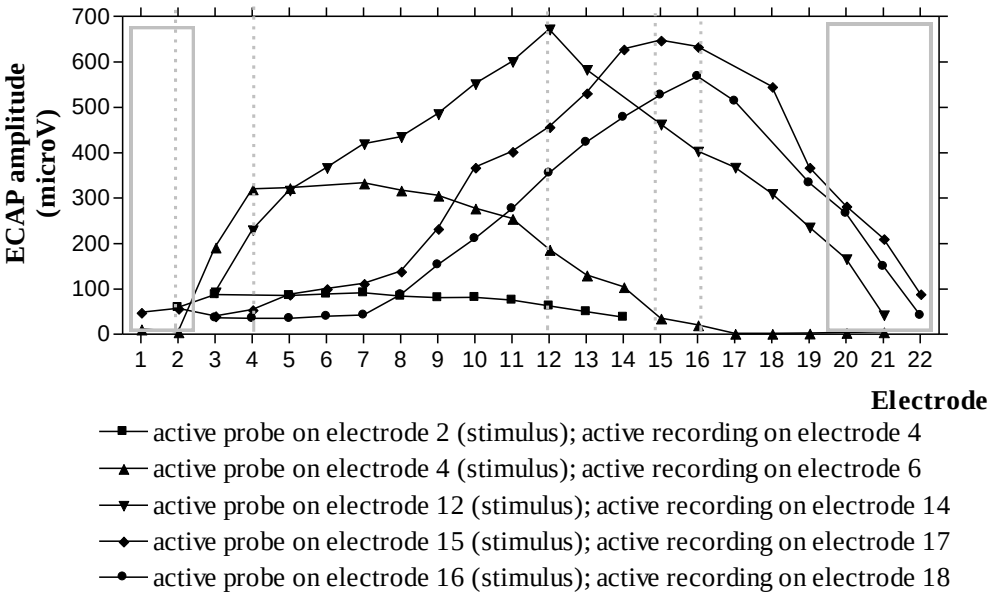

Figure 5. Spatial spread of neural excitation.
Stimulus electrodes are indicated with vertical dashed lines; electrodes lacking an ECAP amplitude measurement are the electrodes on which the active recording took place; the masker active electrode varied; areas of electrodes that had no reproducible ECAPs 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

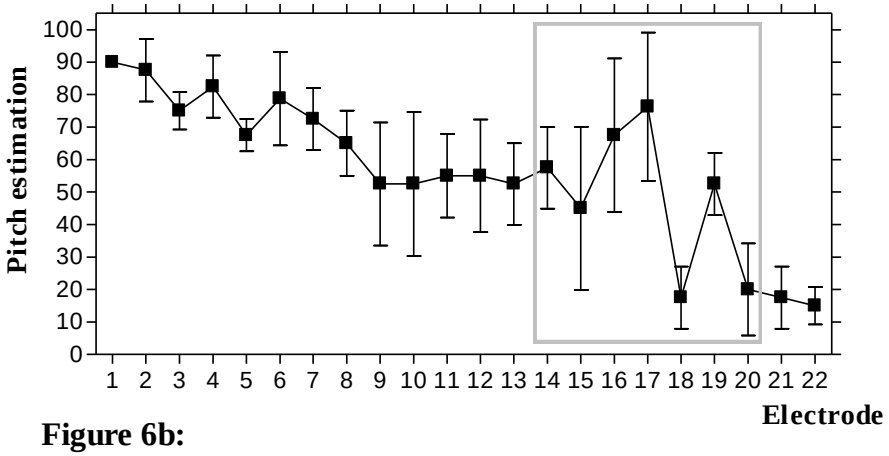

Figure 6b:
Mean and SD of estimated pitch in Case 2

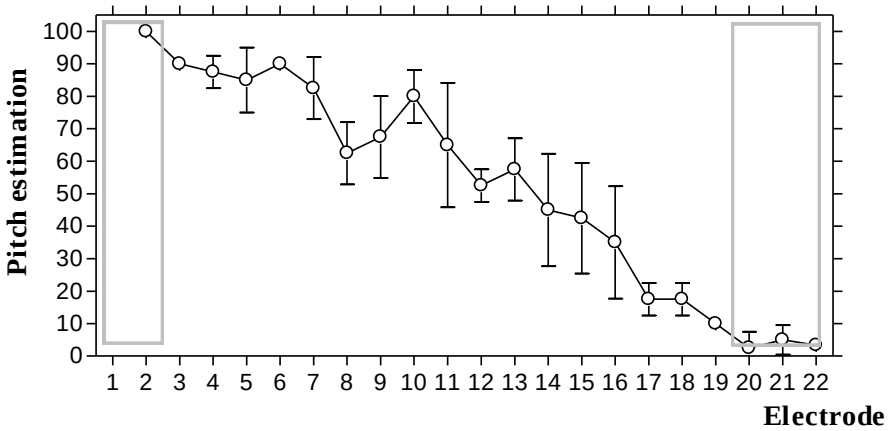

Figure 6. Pitch estimation.
The behavioural responses were obtained using a pitch estimation experiment on a 100-point scale. Each electrode was measured 4 times. Mean scores and standard deviation of these 4 trials are shown. Electrodes that caused FNS are indicated in the grey squares.

After one 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.

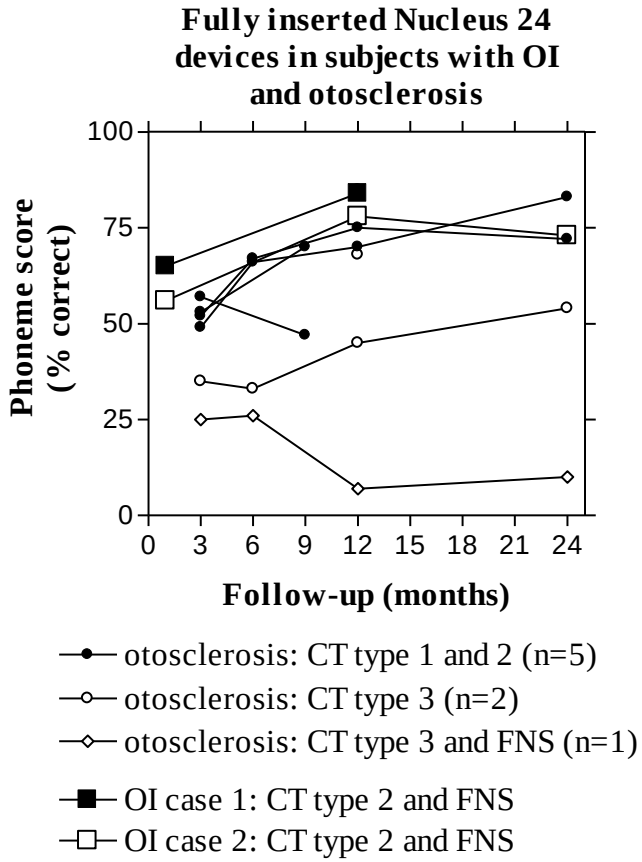

Figure 7. Phoneme scores in 2 patients with OI and 8 patients with otosclerosis implanted with a Nucleus 24 device.

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.[1990] 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 [Ross et al., 1993; Tabor et al., 1990], 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 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.(Thesis, 1992) 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 [Huang et al., 1998; Migirov et al., 2003; Streubel and Lustig, 2005; Szilvassy et al., 1998]. 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 [Migirov et al., 2003]. Previously, the findings on the CT scan in OI patients had proven not to be correlated to the severity of the hearing loss [Ross et al., 1993]. 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 [Streubel and Lustig, 2005]. 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 [Rotteveel et al., 2005]. A preoperative MRI scan can be helpful in predicting cochlear patency and determining which ear to implant. In none of the present OI patients nor in the patients reported in the literature obliteration of the basal turn was 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 [Rotteveel et al., 2004], mastoid cavity or internal meatus, or it might penetrate a newly formed osteolytic cavity in the otic capsule [Ramsden et al., 1997].

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 [Kuurila et al., 2003]. 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 [Handzel et al., 2006]. 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.[2005].

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 [1997] 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. Non-auditory 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%) [Rotteveel et al., 2004]. 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 nerve in dehiscent or otospongiotic bone due to low-impedance pathways, which give rise to an electrical field in the proximity of the facial

nerve [Bigelow et al., 1998]. To avoid an unacceptable decrement in sound quality due to programming manoeuvres such as inactivation of electrodes in order to correct FNS, some authors suggest fluoride treatment [Gold et al., 1998] or botulinum toxin [Langman et al., 1995] and even reimplantation using a device with modiolar facing contacts and perimodiolar position [Battmer et al., 2004].

Average Electrode Voltage

The deviant intracochlear current spread in case 3 has been briefly mentioned before, in a study on AEVs [Mens and Mulder, 2002]. AEV amplitudes vary widely among subjects, partly because of insertion depth [Mens et al., 1994b], 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 [Mens et al., 1994b; Shallop, 1993] and the Clarion device [Hughes et al., 2004]. However, it has been shown that bipolar AEVs in patients with abnormal cochlea's and/or abnormal electrode insertion are significantly deviant [Mens et al., 1994a]. In 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 [Taitelbaum-Swead et al., 2005], 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.[2003], 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 which had to be deactivated in order to control for FNS. Since electrodes 14 and 19 did not cause FNS at C-level, but led to some hearing sensations, these electrodes were switched on. Multi-centre NRT data reported that ECAPs could be elicited in 96% of the cases [Cafarelli et al., 2005]. 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 degrees, as described previously by Cohen et al.[2004], was not found in our cases.

Pitch estimation

Electrode discrimination experiments have shown that multichannel CIs exploit the tonotopic organisation of the cochlea [Collins et al., 1997; Tong and Clark, 1983] which enhances a better speech perception [Busby and Clark, 2000; Donaldson and Nelson, 2000]. 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 non-auditory 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 cochlear implant 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 counselling 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.

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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.[20,105-109] 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.[110] The last years, research has also focussed on the influence of cochlear implantation on the patients quality of life.[111]

A problem with speech perception testing in children is the variability of linguistic abilities.[112,113] 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.[6,7,114-116] 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.[117] 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.[118] 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.[119] 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 cochleas such as in congenital malformed inner ears, post-meningitis ossified cochlea's and extensive cochlear otosclerosis. Consequently, the surgical challenge has increased and revision surgery has become more frequent.[120] Other changes in candidacy which have surgical implications are: the age of the candidate[121-123], presence of residual hearing[124] and multihandicapted patients.[125,126] 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.

7.2.1 Cochlear implantation in the paediatric 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 cochlear implant, although less than patients with complete insertion. This implies that postmeningitic deaf children should receive a cochlear implant soon after the infection before ossification of the cochlea occurs. Nowadays, there is nation-wide consensus between ENT specialists and paediatricians on early evaluation by audiometry and in case of hearing loss referral to an ENT specialist in children diagnosed with bacterial

meningitis.[127] 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.[128] In congenital malformations of the inner ear, abnormalities of the sensory epithelium are often associated with relatively poor development of neural elements. Schmidt [129] 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.[130] 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 cochlear implant 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 one 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[131], while 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 and recurrent meningitis, and because the more complex surgical challenges.[132] 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.[133] The majority of the 53 patients with otosclerosis retrieved from four 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.[134,135]

In all 3 cases, programming was hindered by non-auditory stimulation. Even after reimplantation, non-auditory sensations lead to case 3 becoming a non-user. 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.

7.3 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 cochlear implant 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 cochlear implants 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 cochlea's 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 cochlear implant users, including effortless conversational telephone use.[136] Although these results should be considered preliminary, in patients with ossified cochlea, 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.[137] Depending on future developments of the ABI system, in such cases of compromised cochlea's, implantation of an ABI may be preferred over cochlear implantation.[138]

Concerning future developments in general, at present, various CI centers are conducting clinical studies in order to optimize bimodal[139] and bilateral fitting[140] of cochlear implants and to explore the simultaneous use of acoustical and electrical stimuli in one ear in patients suffering from high frequency deafness.[141] Besides optimizing the results of cochlear implantation for patients with even extended indications, further challenges lie in the development of a totally implantable cochlear prosthesis[142] and the use of intracochlear

nerve growth factors (neurotrophins) to block neuronal death and even lead to repair or regeneration of neurons.[143] Future research also entails exploring auditory physiology using the CI, which in turn can lead to improved restoration of hearing with cochlear implants. Further, studies of hearing in children with cochlear implants form an ideal opportunity to explore age-related plasticity or critical periods in auditory development.[144]

7.4 Conclusions

7.4.1 Children

Congenitally, prelingually and postlingually deaf children all derived benefit from their cochlear implant 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 cochlear implant, although to a lesser extent than patients with complete insertion.

7.4.3 The congenitally malformed cochlea

Cochlear implant 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.

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Chapter 8

Cochleaire implantatie in de gecompromitteerde cochlea

Samenvatting en conclusies

8.1 Resultaten van cochleaire implantatie

Meerkanaals intracochleaire 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.[20,105,145-148] Andere methoden zijn het meten van veranderingen van stem en articulatie, spraakproductie, ontwikkeling van vocabulaire, 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.[149] De laatste jaren heeft het onderzoek zich bovendien gericht op de invloed van cochleaire implantatie op de kwaliteit van leven van de patiënt.[150]

Een probleem bij het testen van het spraakverstaan bij kinderen is de variatie in taalvaardigheid.[113,151] 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 PTA. 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 cochleaire implantaat 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 10dB, 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.[6,7,115,152,153] 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.[154] 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.[155] Deze plasticiteit uit zich in het vermogen van het auditieve system zich aan te passen aan de nieuw aangeboden stimuli aangeboden het CI. 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 leeftijdsgrens of periode waarin implantatie het meest succesvol is heeft eerder onderzoek niet aan kunnen tonen.[156]

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 deze groep kinderen op een aantal variabelen te onderzoeken. De meer recent geïmplanteerde kinderen vormen door het versoepelen van de inclusie criteria een veel 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 cochlea's geïmplanteerd, zoals het geval is in congenitale malformaties van het binnenoor, geossificeerde cochlea's na meningitis en vergevorderde otosclerose. Het gevolg hiervan is dat de chirurgische procedure ingewikkelder is geworden en revisie chirurgie vaker nodig is gebleken.[157] Ook het feit dat er patiënten op heel jonge en hoge leeftijd[158-160], met restgehoor[124] en meerdere handicaps[161,162] worden geopereerd heeft consequenties voor de chirurgische procedure. Deze ontwikkeling heeft tot gemodificeerde implantaten en chirurgische technieken geleid. Complicaties van de operatie zijn tot een nog lager voorkomen 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.

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 beducht 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.[163]
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[164] wordt van patiënten met ernstige malformaties van het binnenoor een slechter resultaat van de cochleaire implantatie verwacht dan van patiënten met normaal ontwikkelde cochlea's. De neurale elementen zijn vaak onderontwikkeld. Schmidt[165] 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.[166]
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 aberrant 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 cochlea's[131]; patiënten met ernstige cochleaire malformaties daarentegen presteren naar verwachting minder goed dan patiënten met normaal gevormde cochlea's doordat er bij hen vaak sprake is van een verminderd aantal ganglioncellen, recidiverende meningitiden en een moeizamere chirurgische procedure.[167] 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.[168] 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.[169,170]
Bij alle 3 de patiënten werd de revalidatie gehinderd door het voorkomen van non-auditieve stimulatie. Zelfs na re-implantatie leidden deze non-auditieve 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 enorme vooruitgang in de spraakverstaan scores van CI patiënten. Met verder onderzoek wordt naar een nog hogere resolutie gestreefd zonder toename in batterij 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 patiënten die door de aanwezigheid van ernstig gecompromitteerde cochlea's of disfunctionele nervus cochlearis niet geïmplanteerd kunnen worden met een CI. De elektrodes van het ABI worden direct tegen de hersenstam geplaatst. Aanvankelijk

werden ABI's geplaatst bij patiënten met neurofibromatose type 2 die bilateraal tumoren in de brughoek hadden. Slechts een klein gedeelde van deze patiënten was in staat woorden te herkennen. Recent zijn ABI's geïmplanteerd in een serie patiënten zonder tumoren maar met gecompromitteerde cochlea's of aplasie van de nervus cochlearis. Een significant gedeelte van deze patiënten bleek wel degelijk in staat tot spraakverstaan op een niveau gelijkwaardig aan dat van de meeste succesvolle CI patiënten, waaronder het moeiteloos gebruiken van de telefoon.[171] Hoewel dit voorlopige resultaten zijn, kunnen bij patiënten met geossificeerde cochlea's, 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 patiënten zonder tumoren, in tegenstelling tot die van patiënten met neurofibromatose type 2, worden mogelijk verklaard door de afwezigheid van pathologie in de brughoek of hersenstam.[137] Afhankelijk van de toekomstige ontwikkelingen van hersenstam implantatie kan in voornoemde gevallen van gecompromitteerde cochlea's een ABI de voorkeur hebben boven cochleaire implantatie.[172]

Ontwikkelingen op andere gebieden door diverse CI teams beogen het optimaliseren van bimodale[173]en bilaterale[174] aanpassingen van cochleaire implantaten. Ook worden de mogelijkheden onderzocht voor simultaan gebruik van zowel akoestische als elektrische stimulatie van één oor bij patiënten met hoge tonen verlies.[175] Naast het verbeteren van de resultaten van cochleaire implantatie bij patiënten met steeds uitgebreidere indicaties liggen er nog volop wetenschappelijke uitdagingen in het verschiet zoals de ontwikkeling van een volledig implanteerbare cochleaire prothese[176] en het gebruik van intracochleaire neuronale groeifactoren (neurotrofines) om verdere neuronen verval te blokkeren en eventueel zelfs te leiden tot herstel ervan.[177] 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 cochleaire implantaten. Verder bieden studies naar het gehoor van kinderen met een CI een ideale mogelijkheid om de leeftijdsgebonden plasticiteit of kritische periode in de auditieve ontwikkeling te onderzoeken.[178]

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 afregelen noodzakelijk kan zijn. Twee van de 3 geïmplanteerde patiënten met Osteogenesis Imperfecta hadden, ondanks de gemeten elektrofysiologische veranderingen, hoge foneemscores.

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