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für Hals-, Nasen- und Ohrenheilkunde

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Clinical and audiological outcomes of cochlear implantation: A retrospective study of 55 cases

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Dedicated to my lovely wife and parents

Abbreviations

ABR Auditory Brainstem Response

AN/D Auditory Neuropathy/Dyssynchrony

BKB Bamford-Koval-Bench sentences

CI Cochlear Implant

CNC Consonant Nucleus Consonant

CSOM Chronic Suppurative Otitis Media

CT Computed Tomography

dB Decibels

ESP Early Speech Perception Test

FF Free Field

HA Hearing Aids

HINT Hearing in noise test

HL Hearing Level

LNT Lexical Neighborhood Test

OAE Otoacustic Emission

P Probability Levels

PE Pressure Equalization

PTA Pure Tone Audiometry

SDT Speech Discrimination Test

S\N signal/noise

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1 Introduction

Hearing loss poses an enormous blockage to the achievement and maintenance of effective communication skills. The awareness and the production of speech are highly dependent on the ability to process auditory information. Early identification of hearing loss is an important first step in managing the effects of hearing impairment. Once identified, the level of residual hearing, if any, must be determined and an appropriate sensory aid recommended. Conventional amplification is usually the first procedure of choice. If little or no benefit is realized with HA, CI becomes therapeutic options. Communication skills and needs must be assessed and a communication mode selected. A sophisticated multidisciplinary team approach that addresses the varied needs of the deaf recipient is required. Essential works of the aural/oral (re)habilitation program include listening skill development, speech therapy, speech-reading training, and language instruction. An absence or disturbance of cochlear hair cells causes most cases of deafness. This defect in normal cochlear function specifically, in the transduction of a mechanical acoustic signal into auditory nerve synaptic activity represents a broken link in the sometimes delicate chain that constitutes the human sense of hearing. The bipolar spiral ganglion neurons and their primary afferent dendrites remain intact, and they are available for direct electric stimulation by the CI. The processed signal is amplified and compressed to match the narrow electrical dynamic range of the ear. The typical response range of a deaf ear to electrical stimulation is on the order of only 10 to 20 dB, even less in the high frequencies. Transmission of the electrical signal across the skin from the external unit to the implanted electrode array is most commonly accomplished by the use of electromagnetic induction or radiofrequency transmission. The electric impulses directly depolarize the primary afferent neurons, thereby effectively bypassing the dysfunctional hair cells [1, 3].

CI are auditory prostheses designed to link an internal device that is interfaced with the cochlear nerve to an external device that uses a specific speech coding strategy to translate acoustic information into electric stimulation, and in this manner allow the transmission of acoustic information to the central auditory pathway. The tonotopic organization of the cochlea is emulated by orienting the

electrode contacts toward the modiolus within the scala tympani and assigning frequencies to specific electrodes along the length of the electrode array such that electric stimulation that corresponds with the highest pitches is delivered within the basal region of the cochlea, whereas electric stimulation that corresponds with the lowest pitches is delivered within the apical region of the cochlea [4].

Current technologic and scientific boundaries prohibit the artificial transduction of speech by utilizing the exact native cochlear patterns of synaptic activity at the level of each individual residual auditory nerve fiber that exists within the normal healthy inner ear. Even so, knowledge about these native patterns has aided the development of CI by allowing the processing of speech into new synthetic electronic codes that contain the key features of spoken sound. By utilizing these codes to systematically regulate the firing of intra-cochlear electrodes, it is possible to transmit the timing, frequency, and intensity of sound. Although relatively limited when compared with a normal cochlea and unable to exactly duplicate natural sounds, CI has nonetheless been shown to successfully represent acoustic signals as meaningful patterns of electrical activity in the central auditory pathway of properly selected individuals who are severely to profoundly deaf [4].

All kinds of device manufacturers use an external processor that encodes speech on the basis of the features that are critical for word understanding in normal listeners. Djourno and Eyries first described direct electrical excitation of the auditory nerve in 1957, since then, increasingly more sophisticated CI have been developed. The development and improvement of cochlear auditory prostheses have radically reshaped the management of children and adults with significant hearing loss. Rapid evolution in the candidacy criteria and the technology itself has resulted in large numbers of individuals who have benefited from implantation [2, 3].

Pre-lingually deafened children acquire speech and language through central plasticity resulting from stimulation by the auditory prosthesis. Some pre-lingually deafened adults are appropriate CI recipients, but they have more limited central

plasticity than what is required for auditory pathway development and processing. *Post-lingually* deafened children and adults, and those with significant hearing loss who gain marginal benefit from HA, are appropriate CI candidates.

Since 2002 initiated cochlear implantation in the Department of Otolaryngology, Head and Neck Surgery, University of Marburg, Germany, and every year the number of patients increases. The aim was to evaluate the clinical and audiological outcomes of CI patients with a follow up time of at least six months after the surgery. The study's design will be retrospective clinical study to patients underwent cochlear implantation.

This study aims:

- To evaluate Etiologies of the hearing loss, epidemiology (age, sex), geographical distribution, and radiological evaluations before and after the surgery.
- It would be concentrate to the duration time of operation, hospitalization days, applied devices and the complications during and after the surgery.
- To compare the PTA and SDT average before and after CI.

2 Patients and methods

2.1 Study's group

The main outcomes measures will be collected from CI cases charts and clinical reviews (55 CI cases in 47 patients) between August 2008 and April 2010 with audiological follow up time of at least six months after CI. The Surgical technique is a retroauricular incision with mastoidectomy and posterior tympanotomy using a round window cochleostomy.

2.2 Study's protocol

To accomplish the aim of the study, it has been developed an ordinary formula (look at Appendix page 58); every detail was recorded in it, and this form has been separated to: personal data, etiology and history, pre-operative evaluation, surgery, and post-operative measurements.

2.2.1 Personal data

Personal identification number, age (children, adults), sex (male, female), and the state of origin have been used to determine the epidemiology data and the geographic distribution of the patients.

2.2.2 Etiology and history

The etiology of the profound hearing loss divided to congenital and acquired sensorineural hearing loss. The pediatric patients (in particular congenital reason) have been examined to find out if they suffer from neuro-pathologic disorders (mental retardation or syndromes). In addition, it has been tried to specify the basic cause of the acquired disorder (meningitis, ototoxicity, presbycusis, etc).

The patients have been classified to pre-lingual or post-lingual hearing-impaired groups (short or long duration of deafness) according to international classification:

- Postlingually deafened adults and children. Patients who become deaf at or after age 5 years are generally classified as *postlingually* deafened. These patients have developed many or all aspects of spoken language before the onset of their deafness.

- Congenitally or early deafened children. Congenital or early acquired deafness is the most frequently encountered type of profound sensorineural hearing loss in children. They generally classified as *prelingually children.*

- Congenitally or early deafened adolescents and adults. When CI is considered in adolescence or young adulthood for a patient who has had little or no experience with sound because of congenital or early-onset deafness, caution must be exercised because this group has not demonstrated high levels of success with electrical stimulation of the auditory system [1]. They generally classified as *prelingually adults.*

Also, a concentration had been done at the patients, who use or not HA before the surgery and the interrupting time of using it, because it could have an influent to the outcomes. And it would be give attention if there is a chronic otitis media or not, because it may change the protocol of the surgery and the outcome.

2.2.3 Pre-operative evaluation

The details of diagnostic procedures have been deliberated before the surgery. It would be thought if there are otosclerosis, facial nerve malformations, congenital cochlear dysplasia and etc, therefore, every patient has imaged by CT-scan and MRI. The pre-operative hearing results with and without HA have been measured. Which were pure tone audiometery threshold [PTA] and has been established by the value of 500+1000+2000Hz in Hearing Level (HL) of dB divided on 3, speech discrimination test [SDT] which was Freiburg test and the number recognizing test. they have been used especially by pre-lingually groups and established by the highest percent of word's or number's production at the less HL of dB, auditory brainstem response [ABR] and concentrate to the dB

level of appearance of wave V and the present or not of prolonged I-V or III-V to guaranty that there was not a neural hearing loss, evoked otoacustic emission [OAE] to make out if the hearing loss is sensory or neural and free field testes [FF]. An especial concentration had been done to differentiate the results between the pre- and the post-lingual patient. The cohort as it mentioned before will divide to three groups dependent on international classification: post-lingually deafened patients, pre-lingually deafened children, and pre-lingually deafened adults.

2.2.4 Surgery

It has been ordered the operated ear to primary or re-implantation cases, which side has been implanted, and whether is a unilateral or bilateral CI. Also the duration of operation including the anesthesia time, facial monitoring preparation time, audiologic test time and finally the explanation time to the students, hospitalization days and applied devices have been taken in consideration. The complications of surgery: injury of facial nerve, corda tympani nerve, or the ear drum, or after the surgery: wound infection, tinnitus, vertigo, etc have been studied.

2.2.5 Post-operative measurements

Every patient has been underwent CT-scan or digital volume tomography [DVT] of the temporal bone after the surgery to confirm the location of electrode array. The next step of evaluation was hearing test results after surgery, which has been recorded after six months or more and the results before six months has not been considered. The results of SDT, PTA, FF testes have been matched up to the results before the surgery. Furthermore, the increasing of communication skills had been evaluated, especially if the post-lingual deaf patients can communicate with a telephone call or the pre-lingual deaf patients can produce more new words or numbers.

2.3 Statistical analysis

For the initial planning of the study timetable Microsoft-Excel 2007 of Windows Vista (Microsoft Corporation, One Microsoft Way, Redmond, WA 98052 USA) was used. For each case a printed form was prepared and labeled with the appropriate number. On this form all the information that we were looking for with details especially hearing results were recorded in a uniform way.

The statistical analysis was performed using Graph Pad Prism, and Microsoft-Excel (included Fisher and T testes) for the type of significance estimate and these testes are not only assuming the normal distribution or equal group variances, also these tests require that the Exact tests add-on module be installed.

The average of the audiologic tests and its diagrams was established with Excel program and also the distribution of audiologic and clinical results before and after the surgery was performed by Excel and Graph pad prism.

A *P* value of less than 0.05, which was calculated by Graph Pad Prism T test, was considered statistically significant and mentioned to it by symbol (★).

3 Results

3.1 Patient's data analysis

Forty-seven patients with CI have been separated to 15 children (32%) and 32 adults (68%). The average of the children age was 3.5 years, the youngest age was one year and the major age was 17 years. The mean age of the adult was 48 years, the smallest age was 18 years and the major age was 76 years. Twenty-five patients (53%) were male and 22 (47%) were female. Thirty-nine patients (82%) live in Hessen and 8 patients live in another germane state.

3.2 Etiology of hearing loss

The cohort (55 cases) consisted of 29 (53%) acquired and 26 (47%) congenital hearing loss cases. The reasons of the acquired hearing loss in 16 out of 29 cases distributed to; 12 presbycusis, 2 head trauma, 1 meningitis and 1 after chickenpox vaccine. The reasons of the residual 13 cases could not be identified. In another hand, 21 congenital cases were normal by neuro-pediatric exam, only 5 out of 26 cases had syndromic disease; one case had Bartter NKCC2 defect syndrome and 4 other cases had global development delay and mental retardation.

The cohort has been divided to three groups; 32 cases (58%) with post-lingual children and adults, 20 cases (36%) with pre-lingual children and 3 cases (6%) with pre-lingual adults. The incidence of using HA before the surgery was 98%. Only one case of post-lingual group has interrupted using HA for 3 years.

3.3 Pre-operative evaluation

3.3.1 Imaging studies of the temporal bone

The most cases (94%) did not illustrate any malformations. Only 3 cases had variations by temporal bone imaging. The first case emerged thick wall of the scala and a small cochlea without correlation to a syndrome. The second case showed cochlea dysplasia and the third case had radical cavity after cholesteatoma eradication.

3.3.2 Pre-operative audiologic results

The PTA threshold of the entire cohort and the groups had been illustrated in figure 1.

The PTA threshold without using HA has been utilized to 34 out of 55 cases. More than 50% of them were profound hearing loss or deafness and the average of PTA without HA was above 105 dB. The PTA threshold with using HA had been done to 8 out of 55 cases and the average of PTA with HA was 47 dB.

As it had been mentioned before, the cohort had been divided to three groups (see page 4). The PTA and FF tests had been identified of each group.

Thirty cases of the post-lingually group had been applied PTA threshold without using HA and the average of PTA without HA was more than 100 dB. Eight cases belongs to the post-lingually adults group had received PTA threshold with using HA and the average of PTA with HA was 47 dB. As well as the average of PTA without HA of 2 cases pre-lingually adults and of 2 cases pre-lingually children was above 115 dB.

The FF threshold with and without using HA had been utilized only to Pre-lingually children group. The mean PTA threshold without HA in FF of 10 cases pre-lingually children was more than 105 dB. As well as the mean PTA threshold with HA in FF of 15 cases pre-lingually children was above 75 dB.

All the groups had been given subjective and objective hearing tests to determine candidacy. The post-lingually group has been evaluated prevalently with PTA and SDT with and without HA. The pre-lingually children group has been assessment frequently with ABR, OAE, SDT and FF with and without HA. The pre-lingually adults group has been tried to cover with all the audiometric tests.

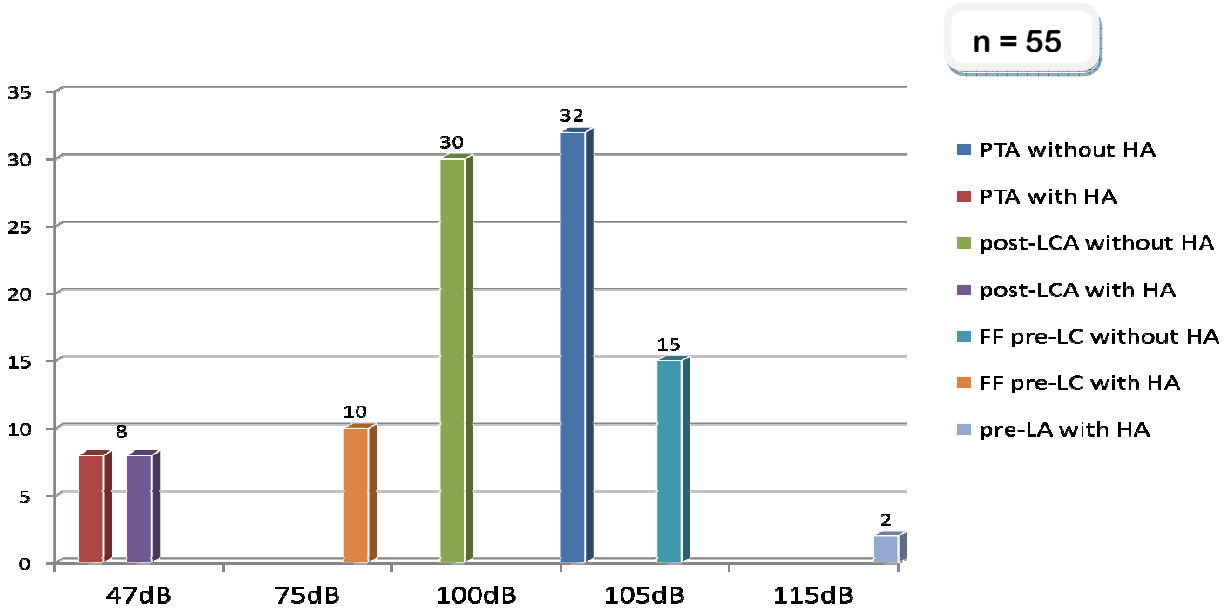

Figure 1 The mean PTA threshold pre surgery

The SDT with and without HA has been measured up to 47 out of 55 cases pre-operatively and all the next counted results are illustrated in figure 2.

The most cases (95%) have been responded of the SDT with and without HA at 0-40% of words producing. The average of the SDT without HA was 16% of words producing and with HA was 28% words producing.

The SDT with and without HA of the three groups of candidates has distributed from 0% to more than 80% of words producing. The average of SDT of the post-lingually group without HA was 15% of words producing and with HA was 24% of words producing. The average SDT of pre-lingually children group with using amplification (the best fitting condition) was 20% of words producing. The only one case of post-lingually group, which had interrupted using HA for 3 years, had been responded to SDT with HA at 30% of words.

The 2 out of 3 cases (one patient) of pre-lingually adults had been responded of the SDT with and without HA and they produced only 10% of words.

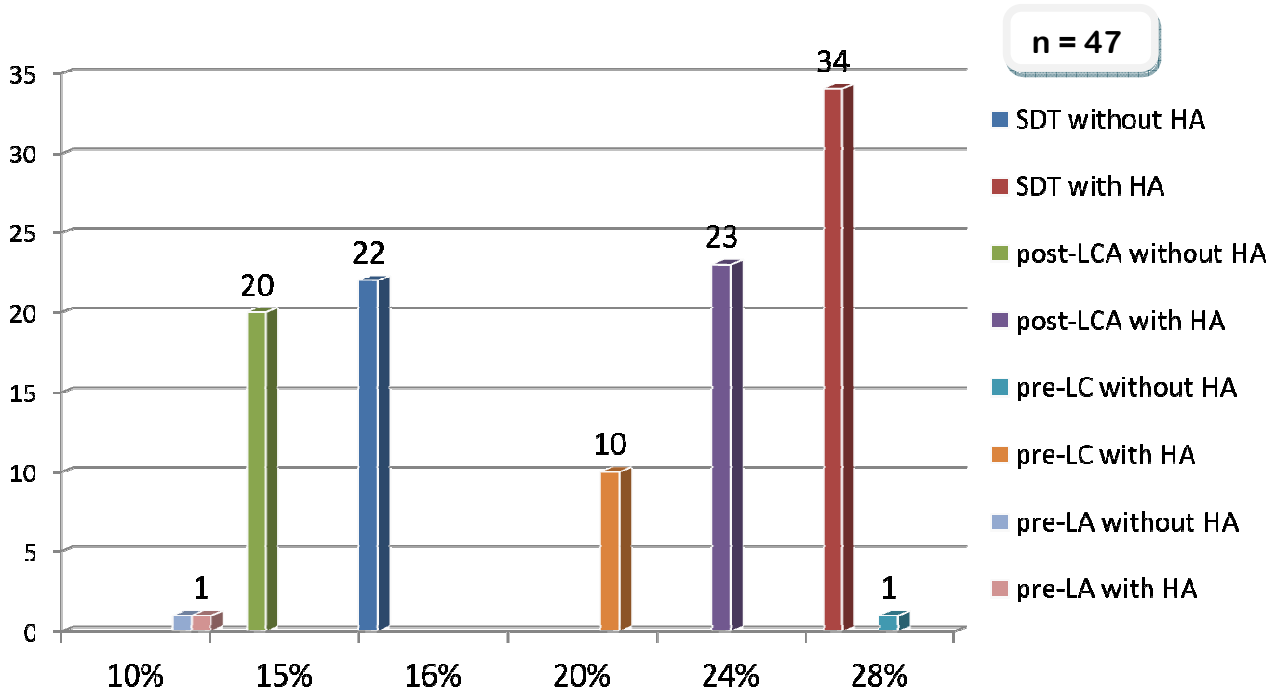

Figure 2 The mean percentage of words in SDT pre surgery

3.4 Surgery

The left ear had the better incidence (57%) of CI than the right ear which had incidence 43% of the entire cohort.

The fifty-five cases distributed to 22 bilateral CI (40%) and 33 unilateral CI (60%). There was no difference in the incidence of the bilateral cases between the adults (50%) and the children (50%). The primary CI was dominantly (90%) in contrast of 10% revision CI.

The device of cochlear has three types in the present study; 78% cases received Nucleus (Australia), 18% cases received MedEl (Austria) and 4% received Hires90k (USA). Additionally, there was no relation between the age of the patient and the art of used device. The use of Hires90k was only for the revision CI and the most use of MedEl was saved to the patients with remnant hearing.

The mean time of CI operation was 3.45 hours and it includes the anesthesia time, the audiologic test at the end of surgery, the preparation time of facial monitoring, and the clarification of CI surgical steps to the students. Thirty-eight CI cases (69%) lasted 3.5 - 4.5 hours. Eleven cases were early ended in 3 - 3.5

hours. Only one case had been completed in 9 hours, which was long approach with radical mastoidectomy plus abdominal fat obliteration with oversewing the external auditory canal.

Thirty-two cases (58%) had spent 5 days in the hospital after the surgery. Three cases were early discharged after 4 days. Only one case has expended 10 days, which had severe vertigo and disturbance in the heart vascular system.

3.5 Post-operative measurements

CT scan or DVT of temporal bone had been taken place to all the patients after the surgery. All the array of the CI device was remarked in the right place in the cochlear, especially in the first turn of the cochlear basis. Even the cases of suspected dysplasia of cochlear, the scala tympani were patent and had been received MedEl test device before the implantation.

The PTA and SDT results of the CI cases after six months of the surgery have been analyzed and compared with the results with and without HA before the surgery.

The following expected results of PTA threshold after CI of all the cohort and the three groups has been illustrated in figure 3.

The threshold of PTA of 47 cases after CI has been estimated and the average was 44 dB. More than half of the cases had threshold less than 50 dB.

More than 68% of the post-lingually patients and the pre-lingually children had PTA threshold in range 20-50 dB. The mean PTA threshold of the post-lingually group was 43 dB. As well as, the mean PTA threshold of the pre-lingually children group was 44 dB. It is noticeable that the PTA threshold after CI of the 3 cases pre-lingually adults group was between 60-70 dB.

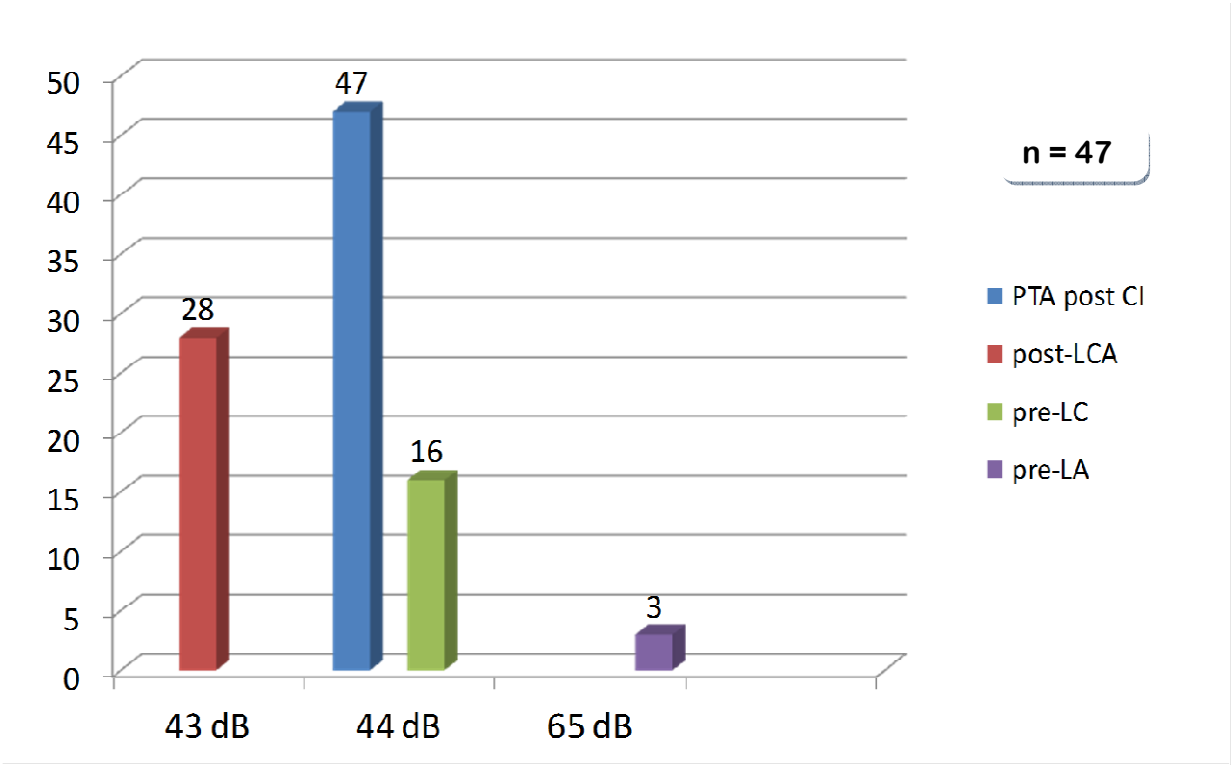

Figure 3 The mean PTA threshold post surgery

The important test after the surgery is SDT while it gave the benefit of CI. All the predicted results below of SDT after CI of all groups are illustrated in figure 4.

Twenty-four out of 55 cases (44%) had responded after 6 months by producing words. More than half of them after 6 months could give more than 60% of the words. The average of SDT post-CI was 62% of words producing.

The SDT had been measured up to the three candidate's groups. The average of SDT post-CI of the post-lingually group was 60% of words producing. The average SDT post-CI of the pre-lingually children group was 63% of words producing. One of the three cases of pre-lingually adults could produce 70-80% of words.

Also one case of the post-lingually group, which had interrupted using the HA for 3 years, could increase producing words from 30% of words by 90 dB HL with HA to 70% of words by 80 dB HL post-CI.

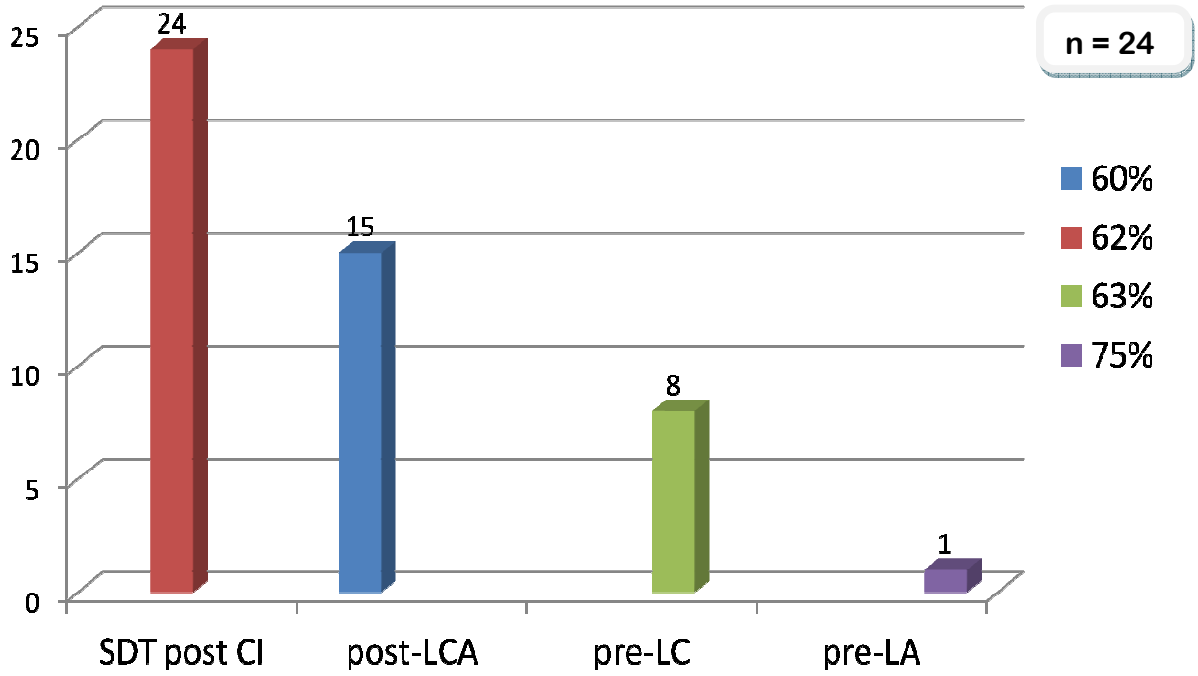

Figure 4 The percentage of SDT of words after surgery

Even the SDT by numbers has been estimated to 24 out of 55 cases (44%) after six months. More than 90% of them could produce 80-100% of numbers by 65-80 dB HL. The average of SDT post-CI was 92% of numbers producing. There is a remarkable incidence (83%) of the pre-lingually children and adults who could produce 90-100% of numbers of SDT.

It is important to mention that only 24 out of 55 cases could success the SDT after the surgery, because of the short time (after six months) of phonetic rehabilitation. Regard to many centers, they maintain that some patients, especially pre-lingually children and adults, need about 1-2 years of phonetic rehabilitation to success more than 80% score of the SDT.

The PTA threshold had been evaluated between 36 cases of pre-CI and 47 cases of post-CI and the following detected results are illustrated in figure 5.

There is a considerable improvement in the PTA threshold. The range was pre-surgery 70 -120 dB and it becomes post-surgery 20 - 60 dB. The mean PTA threshold was 105 dB without HA and 47 dB with HA, and it has turned into 44 dB after CI.

By using the t test, it shows a significant variance between PTA threshold pre-CI and post-CI. The *P* value was less than 0.0001.

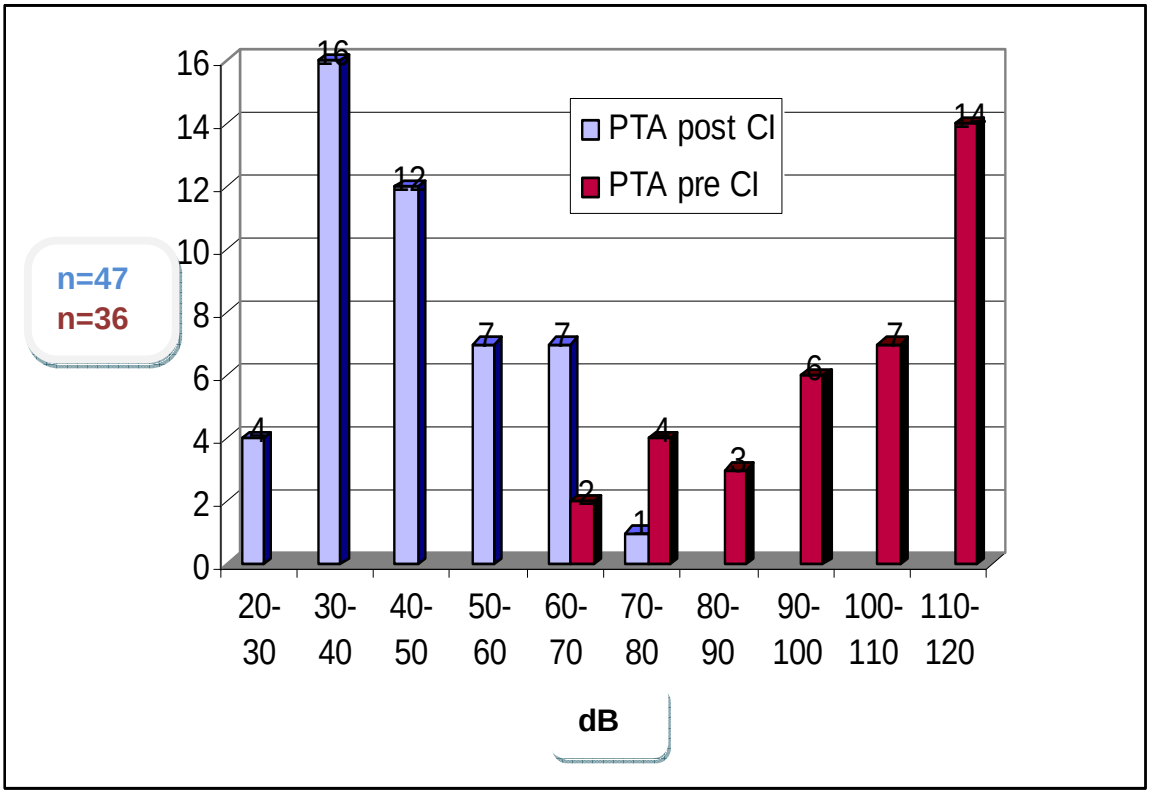

Figure 5 PTA Comparing between pre- and post- CI

The same distribution was predicted in PTA threshold of the three candidate's group between pre-CI and post-CI cases. All the counted results are illustrated in figure 6.

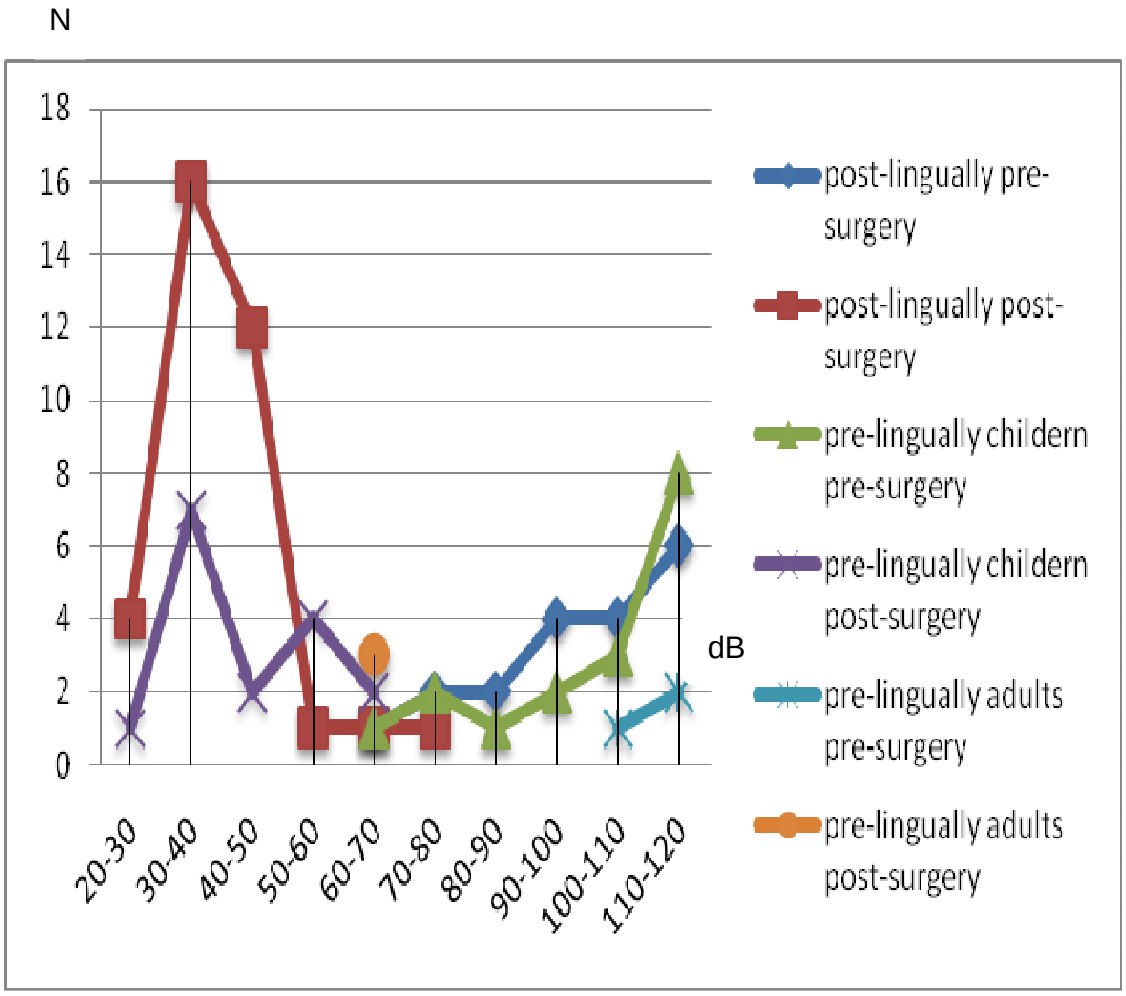

Figure 6 PTA threshold distribution of the cohort between pre-CI and post- CI (n = 47 pre-CI / 36 post-CI)

The average PTA of post-lingually group decreased from above 100 dB pre-CI to 43 dB post-CI. Also the mean average PTA of pre-lingually children group reduced from above 115 dB without HA, 105 dB without HA in FF and 75 dB with HA in FF to 44 dB post-CI.

It is important to mention that there is significant improvement of the mean average PTA threshold of the 3 cases pre-lingually adults group, which decreased from above 115 dB without HA to 60 -70 dB post-CI.

There is a considerable difference of the PTA threshold in t test between pre-CI and post-CI of the post-lingually and of pre-lingually children groups.By applying Prism Graph t test, the *P* value could be calculated and it presents in figure 7.

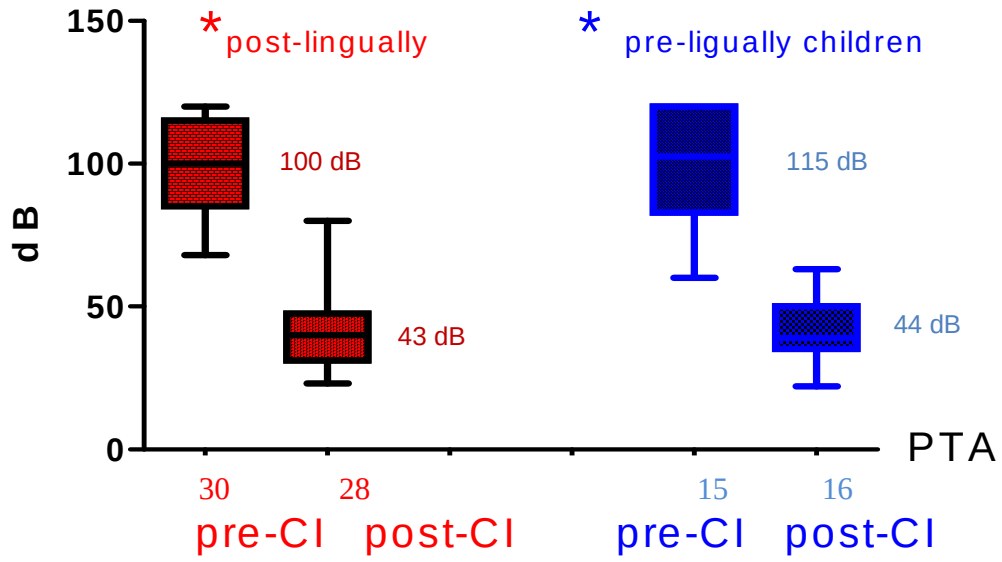

Figure 7 Correlation between PTA threshold pre-CI and post-CI of (post-lingually, pre-lingually children) groups using Prism Graph t test with *P* value * (*P*=< 0.0001, *P*=< 0.0001)

The comparison of SDT between 34 cases with and without using HA and 25 cases using CI has been demonstrated in figure 8.

There is a good improvement in producing and understanding the words by post-CI patients. The average of SDT improved from 28% of words with use HA and 16% of words without use HA to 62% of words by using CI.

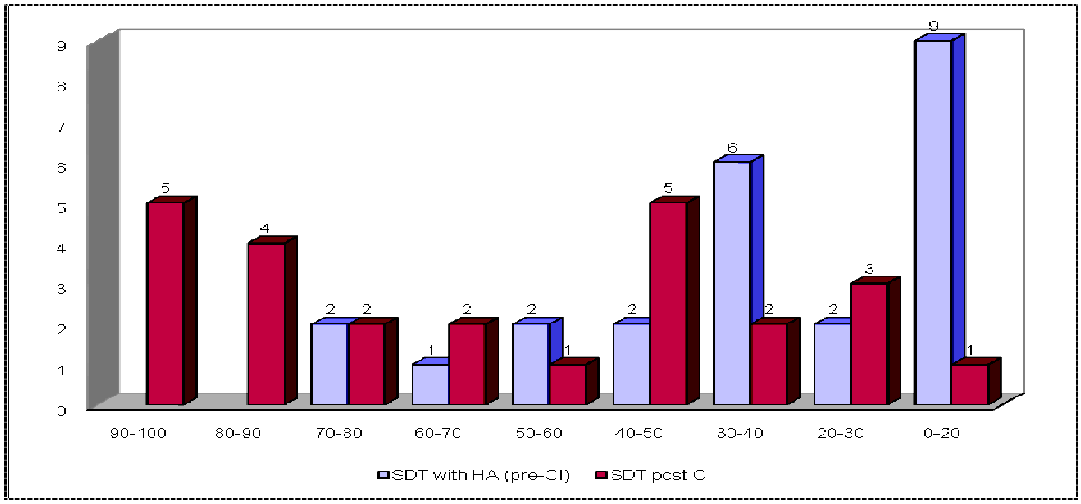

Figure 8 Comparing SDT between pre-CI with HA and post- CI (n=24 with HA / 25 Post-CI)

By applying Prism Graph t test to 24 pre-CI cases with use HA and 25 cases post-CI, there is significant difference in variation and *P* value was less than 0.0001. The previous results are confirmed below in figure 9.

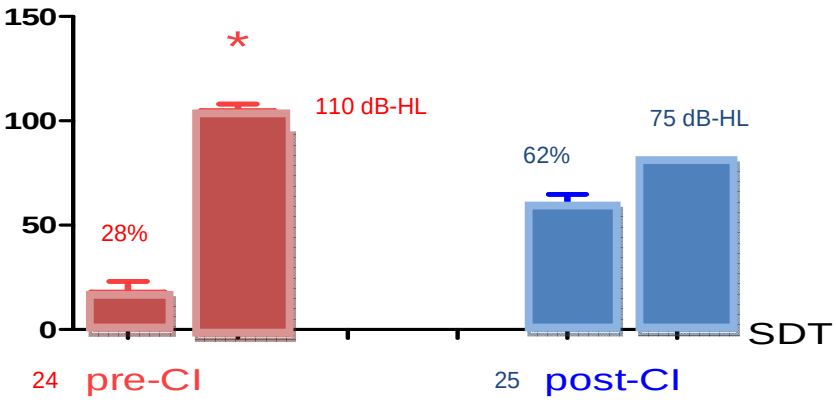

Figure 9 Correlation between SDT pre- and post- CI using Prism Graph t test with positive *P* value *P* = < 0.0001 *

The SDT between with HA and post-CI of the post-lingually group and the pre-lingually children group has been established in figure 10.

The average SDT of the post-lingually group increased from 24% of words producing with use HA and 15% of words producing without use HA to 60% of

words producing by using CI. Also the average SDT of pre-lingually children group moved up from 20% of words producing with and without use HA to 63% of words producing by use CI. One case of pre-lingually adults had improved the SDT from 10% of words producing with and without HA to 70-80% of words producing. There is a large raise of producing words especially by the children otherwise the short time of phonetic rehabilitation.

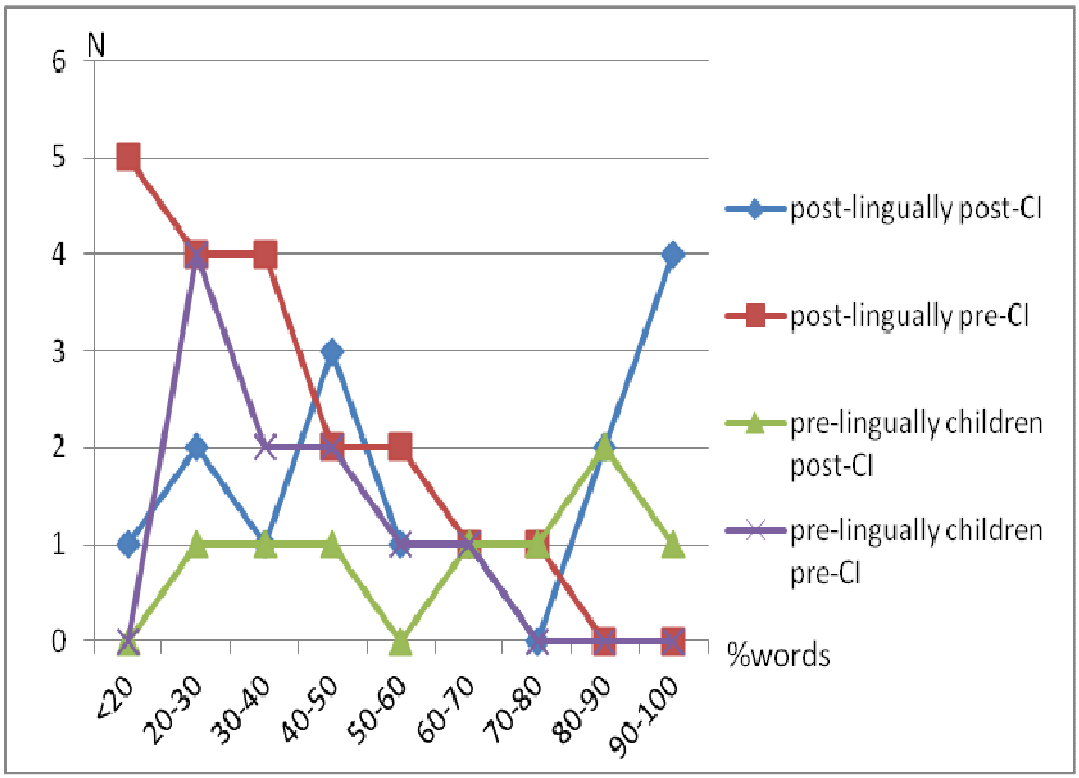

Figure 10 Comparison SDT of (post-lingually, pre-lingually children) group between pre- and post- CI (n = 33 pre-CI / 24 post-CI)

By utilizing Prism Graph t test to the SDT pre-CI and post-CI of the post-lingually group and of the pre-lingually children group, there was considerable difference in variation. The *P* value was less than 0.05. These results are utilized below in figure 11.

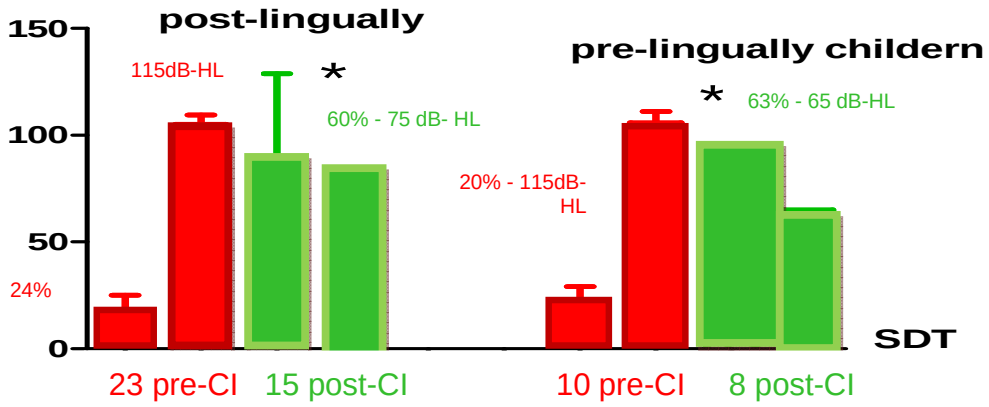

Figure 11 Correlation between SDT pre- and post- CI of (post-lingually, pre-lingually children) groups using Prism Graph t test with *P* value <0.05 *

3.6 Complications

Most of the patients (98%) had not any complications during the surgery. Only one case had injury of the corda tympani nerve, which was unnoticeable from the patient by asking him if there was any deference of tasting and that was thought related to the other healthy side. On the other hand, 38 out of 55 cases (69%) did not have any complication after the surgery. Seven cases (13%) had vestibular symptoms (vertigo, vomiting) but it was self limited after few days which associated with anti vomiting drugs. One case (2%) had tinnitus, which takes one week until using the external speech process to disappear. One case (2%) had fever, which had been treated with paracetamol and diclofenac beside antibiotic, and dropped down after 2 days. One case (2%) had cough, which was an influence from the anesthesia's drugs and stopped in the same day. One case (2%) had otalagia, which had been got care of it with analgesic drugs and gone after 4 days. One case (2%) had swelling in the temporal region, which take more than one weak associated with dressing the head to disappear. Five cases (9%) had been needed to treat with additional surgery; 3 wound infections later than 2 weeks from the surgery had needed to wound's debridement and linked with antibiotic, 1 acute otitis media after acute grippe and treated with ventilation T tube and antibiotic and lastly 1 acute mastoiditis occurred after 3 months of surgery because of acute otitis media and diagnosed by CT, which needed to mastiodectomy and antibiotic.

4 Discussion

4.1 Etiology

4.1.1 Genetic hearing loss

The etiology of the hearing loss is an important consideration. Of the genetic causes, several hundred forms of syndromic hearing loss have been identified, and the list of nonsyndromic loci now exceeds 65 [5]. Profound congenital deafness occurs in approximately 1 in every 1000 children, and roughly 60% of these cases are hereditary [6]. Although there are more than 400 genetic syndromes that include hearing loss, most syndromic deafness is confined to a very limited number of syndromes [8]. There are only two common autosomal-recessive forms of syndromic deafness: Pendred syndrome (deafness, wide vestibular aqueduct, and thyroid dysfunction) and Usher syndrome (deafness, blindness due to retinitis pigmentosa, with or without vestibular dysfunction) [3]. The precise etiology for the deafness cannot always be determined but is identified whenever possible; however, stimulable auditory neural elements are nearly always present regardless of cause of deafness [9]. Two exceptions are the Michel deformity, in which there is congenital agenesis of the cochlea, and the small internal auditory canal syndrome, in which the cochlear nerve may be congenitally absent. In the present study, the syndromatic children were (6%); one case of Bartter NKCC2 defect syndrome and 4 cases (2 bilateral patients) of global development delay and mental retardation.

Relatively recently, the diagnosis of auditory neuropathy/dyssynchrony (AN/D) has been specified as a hearing disorder in which normal cochlear outer hair cell function is found in conjunction with absent or abnormal auditory neural responses; this is analytic of poor neural synchrony [10].

4.1.2 Acquired deafness

In young children, many acquired forms of deafness cannot be easily differentiated from genetic deafness. Prenatal infection with the TORCH organisms (toxoplasmosis, other [syphilis], rubella, cytomegalovirus, and herpes)

is commonly associated with deafness. Prematurity and low birth weight, low Apgar scores, and hyperbilirubinemia can all be associated with deafness autoimmune inner ear disease [3]. There are many inherited or acquired diseases that affect the temporal bone that can produce hearing loss significant enough to require CI. Examples of these disease processes include otosclerosis, Paget's disease, Camurati-Engelmann disease [18], and meningitis with resultant labyrinthitis ossificans. A final consideration is related to temporal bone trauma. Although rare, bilateral temporal bone fractures that result in deafness can be rehabilitated with CI. Early implantation should be performed to avoid cochlear fibrosis. Patients with active chronic ear disease processes, however, are better served with initial conventional otologic treatment with separate additional procedures as needed [3]. Only 16 out of 29 acquired cases (55%) in the present study was determined and most of them (12 cases) was presbycusis (90%), and 10% divided to; 1 meningitis, 2 head trauma, and 1 after chicken box vaccine. The reasons of remain 13 cases (45%) could not be identified.

4.2 Patient evaluation

4.2.1 Otologic evaluation

The medical evaluation begins with a detailed collection of the patient's history followed by a physical examination. The otologic history includes age of onset of hearing loss, progression of the hearing loss, bilaterality of the hearing loss, risk factors for hearing loss (e.g., noise exposure ototoxicity, trauma), and history of ear disease and surgery. History of vestibular dysfunction includes delayed age of walking, difficulty with riding a bicycle, or difficulty maintaining balance while walking with eyes closed or in the dark. A vestibular evaluation, including at least electronystagmography and caloric testing, should be obtained if there is a suspicion of a unilateral or bilateral vestibular hypofunction [4].

A detailed family history is important, including the age of onset, the severity of the hearing loss, and the rate of progression, which had considered in the present study and noted in the study's formula.

For adult implant recipients, an intact tympanic membrane is preferred. Accordingly, those patients with a tympanic membrane perforation, a chronic draining ear, or cholesteatoma often require other surgical procedures prior to implantation [9]. CI was primarily viewed as contraindicated in young children with chronic suppurative otitis media (CSOM) because of the potential risk of infection [19]. Some surgeons advocate a two-stage surgical approach. The first surgery involves a radical mastoidectomy (if not already performed), Eustachian tube obliteration, and mastoid cavity obliteration with oversewing of the ear canal. The second procedure is CI and performed usually 2 to 6 months after obliteration [22]. In the present study one case had been operated in the first step to eradicate the cholesteatoma associated with a canal wall down approach. In a second step was carried out the CI approach with fat obliteration and oversewing the external canal.

Other otologic conditions that merit special attention in the process of surgical planning include otosclerosis and congenital cochlear dysplasia. Patients with otosclerosis are likely to be at a higher risk of unwanted facial nerve stimulation

due to coexistent demineralization of the surrounding bone. For patients with known cochlear dysplasia, unusual surgical anatomy and a higher incidence of CSF leak should be anticipated. Preoperative imaging is very useful in avoiding complications [4]. Regardless of the management protocol, all patients currently receive selected antimicrobial prophylaxis immediately before implantation and cortisone during the surgery [3].

In pediatric patients, it is important to make certain if there is a history of recurrent ear infections, pressure equalization (PE) tube placement, or other otologic surgeries. For patients with a chronic middle ear effusion or recurrent acute otitis media, myringotomy with PE tube placement may be considered. Because children can be implant recipients at a very young age, there is a high likelihood of undergoing an episode of AOM after implantation. These infections should be treated quickly with broad-spectrum antibiotics. Curiously, it has been documented that an ear with a cochlear implant is less likely to develop otitis media than the contralateral ear, probably due to the fact that a mastoidectomy is performed as a part of the implantation [4].

The pediatric patients should inspect by pediatric physician to find out if they suffer from neuro-pathologic disorders and the psychological testing is performed to identify subjects who have organic brain dysfunction, mental retardation, undetected psychosis or unrealistic expectations [1].

4.2.2 Imaging

Radiological evaluation of the cochlea is performed to determine whether the cochlea is present and patent and to identify congenital deformities of the cochlea. High-resolution, thin-section computed tomographic (CT) scanning of the cochlea remains the imaging technique of choice [27]. Intracochlear bone formation resulting from labyrinthitis ossificans can usually be demonstrated by CT scanning. However, when soft tissue obliteration occurs following sclerosing labyrinthitis, CT may not image the obstruction. In these cases, T2-weighted magnetic resonance imaging (MRI) is an effective procedure providing additional information regarding cochlear patency. Intracochlear ossification is not a

contraindication to CI but can limit the type and insertion depth of the electrode array that can be introduced into the cochlea. Congenital malformations of the cochlea are likewise not contraindications to CI. Cochlear dysplasia has been reported to occur in approximately 20% of children with congenital sensorineural hearing loss [28]. In the present study had been observed one case of cochlea dysplasia. Several reports of successful implantations in children with inner ear malformations have been published [29, 30]. A CSF gusher was reported in several patients, and also in this study one case was noted. Temporal bone dysplasia also may be associated with an anomalous facial nerve, which may increase the surgical risk [1]. When deafness is a result of meningitis, special attention is required preoperatively to find out for the possibility of cochlear ossification [4].

4.2.3 Classification of cochlear implant recipients

CI recipients can be divided into three main categories. Significantly different performance outcomes can be anticipated:

- Postlingually deafened adults and children. Patients who become deaf at or after age 5 years are generally classified as postlingually deafened. These patients have developed many or all aspects of spoken language before the onset of their deafness, and they were 32 cases (58%) in the present study.

- Congenitally or early deafened children. Congenital or early acquired deafness is the most frequently encountered type of profound sensorineural hearing loss in children. The achievement of oral communication skills can be a difficult process for these children. They were 20 cases (36%) in the present study.

- Congenitally or early deafened adolescents and adults. When CI is considered in adolescence or young adulthood for a patient who has had little or no experience with sound because of congenital or early-onset deafness, caution must be exercised because this group has not

demonstrated high levels of success with electrical stimulation of the auditory system [1]. This group includes 3 cases (6%) in the present study. Also the concentration had been focused to be sure if they have or not a benefit of the CI.

4.3 Evaluation of adult cochlear implant candidates

The benefits of CI have increased considerably over the last two decades due to changes in technology and expanded candidate criteria. Consideration for CI adults still requires careful assessment to: determine preimplant HA fitting and performance, compare a candidate's preimplant performance with that of current implant recipients, provide a recommendation for or against CI, select an ear for implantation and determine appropriate expectations that will guide the counseling of prospective patients, which is critical for user satisfaction [3].

Current adult selection criteria in the most recent clinical trials include: (1) severe or profound hearing loss with a pure tone average of 70 dB hearing loss (HL); (2) use of appropriately fit HA or a trial with amplification; (3) aided scores on open-set sentence or words tests of <50%; (4) no evidence of central auditory lesions or lack of an auditory nerve; and (5) no evidence of contraindications for surgery in general or CI surgery in particular.

Additionally, CI centers generally recommend at least 1 to 3 months of HA use, which was in this study about 96% of cases using HA. Their mean average PTA without HA was above 105 dB and their SDT mean average was 16% words. Realistic expectations by the patient and family members and willingness to submit with follow-up procedure as defined by each CI center alone [3], which was at least 6 months after CI in the present study.

For adults, sound detection and speech perception abilities are assessed to determine candidacy. Preoperatively, patients are evaluated with a battery of measures while using and without using HA. Preoperative measures are also repeated after the implant for longitudinal monitoring of patient performance.

Preimplant audiologic tests include unaided and aided detection thresholds for pure tone and warble-tone stimuli, respectively. Unaided thresholds are obtained in each ear individually, and aided detection thresholds may be obtained monaurally as well as binaurally. Aided speech perception abilities are often assessed in both monaural and binaural conditions, depending on the use of amplification in each ear. Speech perception measures are conducted in the sound field (FF) and include open-set recorded presentations of words and sentences in quiet and, if appropriate, in noise. In the best-aided condition, the assessment of individual ears provides critical information for determining in which ear to place the implant for unilateral implantation. In addition, the best aided condition, whether it be either ear alone or both ears together, provides information about the candidate's maximum performance for comparison with CI performance. Word and sentence recognition tests are a set of compact disc recordings designed to provide word and sentence tests for the preimplant and postimplant evaluation of speech recognition, regardless of implant device. The Consonant-Nucleus-Consonant (CNC) Monosyllable Word Test [32] assesses single syllable word recognition. One CNC list contains 50 monosyllabic words presented in an open-set format, which was Freiburg test in German words in the present study. Clinical observations suggest that, when testing adults, scores on open-set word and sentence measures are more reflective of patient satisfaction with hearing aids and more useful for determining CI candidacy than unaided and/or aided detection thresholds [3]. In the Marburg hospital received the adults patients the same worldwide standard audiologic diagnostic protocol, and it was obvious in the results of the present study.

Traditionally individuals have received CI in one ear only; binaural implantation includes improvements in sound localization and listening in noise [39]. Specifically, studies have shown that binaural implants provide a "head shadow" effect for listening to speech in the presence of other noise or opposing speakers, this occurs because one ear is "shadowed" from the noise source when speech and noise come from different directions, thus allowing the ear with the better Sound/Noise ratio to do the listening. Other binaural advantages occur when information from both ears is combined to improve listening. Patients are

increasingly inquiring about the possibility of binaural implantation. Because results thus far are encouraging and because the majority of bilateral recipients indicate a strong preference for bilateral over unilateral implant use, it is possible that binaural implantation will become a part of the candidacy decision [3]. This tendency is obvious in the present study, which were the binaural 22 from 55 cases (44%).

The most common pre-implant factors that affect performance for adults include hearing experience (e.g., amount of residual hearing, length of profound hearing loss, hearing history for each ear), age at onset of profound hearing loss (particularly if before the age 3 years), age at implant (particularly if 75 years old or older), cognitive/central abilities, and motivation to hear. Post implant factors that contribute to performance levels may include length of CI use, stability of threshold and comfort levels used for device programming, and lifestyle. The need for auditory skills and social interaction in the environment can be more of an issue for those who are not in the work force or who live alone (often the elderly), because they have less practice listening. Two such factors are age at implantation and duration of deafness [43, 44, 45], specifically, patients who are implanted at a young age and have a shorter period of auditory deficiency are more likely to achieve good outcomes. Other factors that have been found to significantly correlate with adult outcomes include speech-reading ability [46, 47] and degree of residual hearing [46, 48].

CI teams have different philosophies about the selection of the ear for implantation. Some believe that the poorer ear should be chosen for implantation, whereas others consistently choose the better ear. Generally speaking, with a normally developed cochlea, some authors expect the ear with the shortest length of deafness, better acoustic detection thresholds, acoustic hearing at more frequencies, and better word recognition to be the better ear. The selection of worse ear to receive CI was carried out in the present study.

4.4 Evaluation of pediatric cochlear implant candidates

CI has been available for children between the ages of 2 and 17 years since 1990, nowadays the age begins at 1 year old. Originally, children who were candidates for CI typically had profound bilateral sensorineural hearing loss with pure tone average thresholds of 100 dB HL or greater, often with corner audiograms, which was above 115 dB in the present study. These children also showed aided sound-field thresholds well below the range of average conversational speech and typical speech detection thresholds at and above 60 dB HL, also was in the present study above 75 dB. As is the case with adults, consideration for CI still requires careful assessment to do the following: determine the preimplant fitting of HA and baseline performance, compare a candidate's preimplant performance with that of current implant users, provide a recommendation for or against CI, select an ear for implantation, and determine appropriate expectations that will guide the counseling of prospective families [3].

Generally speaking, the subject selection criteria include: (1) 12 months through 17 years old; which was equivalent in the present study, (2) profound sensorineural hearing loss (unaided pure tone average thresholds of 90 dB HL or greater); and was above 115 dB in the present study, (3) minimal benefit from hearing aids, which is defined as less than 20% to 30% on single-syllable word tests, so the same in the present study which was 20% with HA. For younger children, the lack of developmentally appropriate auditory landmark measured using parent report scales, (4) no evidence of central auditory lesions or lack of an auditory nerve, and (5) no evidence of contraindications for surgery in general or CI surgery in particular. Additionally, CI centers generally recommend at least 3 to 6 months of HA use unless cochlear ossification is noted or predictable but it is not always essential to carry out, when the family has a history of hearing loss; realistic expectations by family members; staffing in a post-operative rehabilitation program that supports the use of CI and the development of auditory skills; and motivation on the part of the family to comply with follow-up procedures as defined by each CI center alone [3], which was at least 6 months in the present study.

As with adults, children are assessed preoperatively with a battery of sound detection and speech perception measures while using or not HA. For children, speech perception measures assess a wide range of auditory skills, from sound detection to the recognition of words and sentences. Measures are selected that are developmentally appropriate for the child's age, language level, and auditory ability. Although the audiologic assessment will play a key role in candidacy, with children, other factors may influence the candidacy decision and/or postimplant outcome and, therefore, a multidisciplinary team approach is advised [3].

Before CI evaluation, most children will have an ABR test as an objective measure of the status of the peripheral and brainstem auditory system. With an ABR, acoustic click stimuli are presented to assess the auditory sensitivity of each ear. Children who are implant candidates typically have no response to acoustic stimuli at the limits of the testing equipment, thereby suggesting with reasonable accuracy significant hearing loss in the profound range. Another group of children that can present absent or abnormal ABR findings are those with auditory neuropathy, a condition that is characterized by abnormal neural function at the level of the inner hair cells or cochlear nerve but normal outer hair cell function [49]. In these cases of absent/abnormal ABR, a comparison of positive (condensation) and negative (rarefaction) polarity stimuli will show an inversion of the peaks of the cochlear microphonic. The cochlear microphonic appears as an early latency response on the ABR waveform and is indicative of outer hair cell function.

OAE testing can also be used as a measure of outer hair cell function. Because of the prevalence of children diagnosed with auditory neuropathy/dyssychrony [50] and because of the number of these children who have received cochlear implants [50], the current protocols for electrophysiologic assessment include OAE and ABR testing, because these measures are sensitive to cochlear and auditory nerve function, respectively.

Unaided detection thresholds for pure tone stimuli are obtained in individual ears using standard clinical procedures. Aided thresholds are obtained in the binaural condition and, if possible, the monaural condition. For young children who are

unable to participate in speech perception tasks, both unaided threshold testing and electrophysiologic measures become important criteria for cochlear implantation [3], as well in Marburg hospital the children patients received the identical standard audiologic diagnostic protocol and it was obvious in the results.

Tests of speech perception assess a range of skills that depend on the child's auditory abilities and language level. Closed-set measures include a small number of choices that are provided to the child either as objects or pictures (e.g., Early Speech Perception Test) [52]. Monosyllable, spondee, and/or trochee words are spoken with test alone (no visual cues), and the child is asked to select the object or picture that represents the stimulus. With open-set measures of word and sentence recognition, no choices are provided. The child repeats the words or sentences presented in quiet or in the presence of background noise. For children with vocabulary levels that approximate those of 5-year-old child, the Phonetically Balanced Kindergarten Test [54] can be administered; it includes 50 words and has been in clinical use for many years.

For children, the results of speech production assessments are good indicators of hearing history and of whether the child has learned to use his or her residual hearing. Language evaluations are also important, because the vital goal of cochlear implantation is effective communication. Results also are used to monitor either pre or post implant performance over time and to develop rehabilitation goals for educators, clinicians, and parents [3]. Differentiating the impact of deafness and CI from other disabilities or diagnoses such as developmental delay, autism, attention deficit disorder, or learning disabilities can be difficult. These issues are addressed in the pediatric psychological evaluation before the implant and influence the recommendation for or against cochlear implantation, provide guidance for counseling families, and assist with rehabilitative planning. A team effort is best started during the pre implant process and sets the stage for later communication between the individuals on the implant team and the child's educators and family. Early development of communication is important for a variety of reasons, including the confirmation of the child's test results and use of residual hearing, the discussion of areas of concern, the sharing of effective test-taking and rehabilitative strategies, the

setting of expectations, and the identification of post implantation rehabilitation sources and goals [3].

As with adults, there has been an increase in the number of centers involved with the bilateral cochlear implantation of children, primarily in Europe, especially Germany. Reports for children follow similar trends as those for adults, with improvements in the ability to recognize speech in noise and to localize a sound source. The ability to follow large spatial changes in speaker location is a critical skill for academic learning in the classroom setting, as is the ability to follow rapid changes between speakers in a smaller space (e.g., in a small group setting at school or during a conversation with multiple speakers at home) [3].

The most common pre-implant factors that affect performance for children include age at implantation; hearing experience (age at onset of profound hearing loss, amount of residual hearing, progressive nature of the hearing loss, aided levels, stability of HA use), training with amplification (in the case of some residual hearing), presence of other disabilities, and parent and family support. Furthermore, postimplant factors that contribute to performance levels include length of CI use, rehabilitative training, and family support. Communication mode is also a documented variable that affects postimplant outcome; this essentially means that children in programs and homes that focus on the development of spoken language perform at a higher level than children in programs without this emphasis [58].

For children, the selection of the ear for unilateral implantation follows the same logic as discussed earlier for adults. Because some centers encourage the use of a contralateral HA after the implant if at all possible, they select the ear for implantation that is least likely to benefit from amplification. When all things are equal, they select the right ear to capture the possible advantage of contralateral left-hemisphere specialization for speech recognition [59].

A unique group of individuals requiring careful consideration are those with hearing loss and other developmental and cognitive deficits. Historically, children with cerebral palsy or children with other conditions in addition to hearing loss were denied implantation. It is now clear, however, that many of these patients

are very good candidates. In fact, if a hearing disability can be reduced with a CI; other disabilities (eg, a learning disability) may become less pronounced or more manageable [4].

4.5 Cochlear implant systems

4.5.1 Hardware

Currently, three separate corporations manufacture multichannel implant systems that are commercially for use in both adults and children: (1) the Nucleus Contour system manufactured by the Cochlear Corporation (Sydney, Australia), (2) the Clarion system manufactured by the Advanced Bionics Corporation (Sylmar, California), and (3) a recently approved system manufactured by the Med-El Corporation (Innsbruck, Austria). All modern implant systems function by the use of the same basic components, including a microphone, a speech processor, and an implanted receiver-stimulator.

4.5.2 Microphone and receiver-stimulator

Sound is first detected by a microphone (usually worn on the ear) and converted into an analog electrical signal. This signal is then sent to an external processor where, according to one of a number of different processing strategies, it is transformed into an electronic code. This code, usually a digital signal at this point, is transmitted via radiofrequency through the skin by a transmitting coil that is held externally over the receiver-stimulator by a magnet. Ultimately, this code is translated by the receiver-stimulator into rapid electrical impulses distributed to electrodes on a coil implanted within the cochlea (figure 12).

4.5.3 External speech processors

Each manufacturer offers both body-worn and behind-the-ear processors, both of them have program switches, volume and/or sensitivity controls, batteries (rechargeable or alkaline), and accessories.

External processor wear options vary from one device to another, but they may include, for example, a remote battery that is pack worn off of the ear or a rechargeable battery pack that is worn on the processor at the ear. A variety of mechanisms exist (e.g., ear hooks, indicator lights) that perform functions such as alerting parents about a low battery or a disconnected headpiece. External auditory input sources can be connected to the processors, such as supporting microphones, telephone adaptors, tape recorders, television audio amplifiers, and FM systems.

4.5.4 Speech processing

The literature uses the term speech processing but this component may be more rightly termed *sound processing*, as the manipulations are not limited to speech only.

In fact, there is now a greater focus on enhancing the quality of all sound, and specifically an effort to improve music enjoyment. Processing speech and other sounds within a CI system is a complex process that is continually developing. No matter what strategy is employed, part of this process must include both

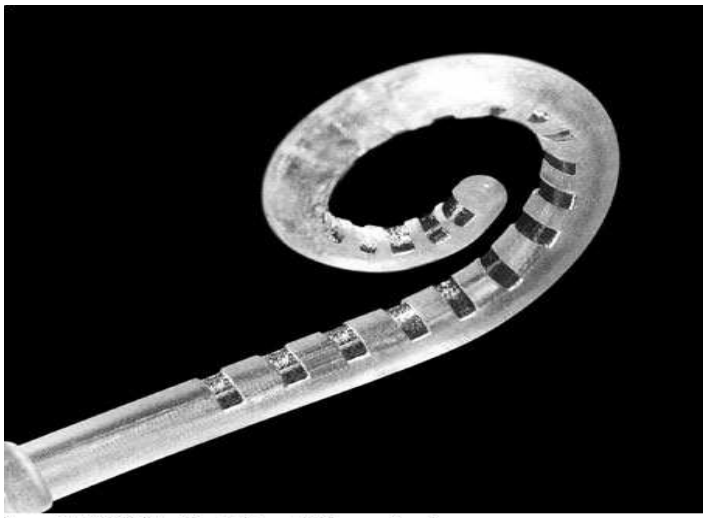

Figure 12 The Nucleus CI-24 curled electrode array

amplification (eg, gain control) and compression. Since the deaf ear responds to electrical stimulation with a dynamic response in the range of approximately 10–25 dB, processing must include compression of the signal to fit within this narrow range.

4.5.5 Internal receiver/stimulators and electrode designs

The Nucleus 3 System (CI24R (CS) with Contour electrode) internal receiver/-stimulator uses a flexible silicone housing that surrounds a titanium case (Figure 13). The Nucleus 24 Contour electrode uses a perimodiolar electrode design, and it is preformed to match to the modiolus. There is a style that is positioned within this electrode array that maintains the electrode in a straight configuration until its removal during surgery. The electrode array is curved and consists of 22 half-banded platinum electrodes that are variably spaced over 15 mm. Overall, the length of the electrode array distal to the first of three silicon marker rings is 24 mm; however, the electrode is designed to be inserted 22 mm, and a platinum band is present at this position to use as a guide for depth of insertion. Of all of the available electrodes on the market, this is the stiffest electrode and consequently, it is relatively easy to insert. The highest incidence (78%) of this device has been implanted in the present study.

The greatest disadvantage of this current electrode design is that, after the style has been removed, it cannot be replaced. This is problematic if the electrode insertion is difficult because of anatomic variations, in which case the backup device would be required [60]. The Nucleus device also has a second electrode design: a double electrode array to be used for the implantation of severely ossified cochlea. The configuration for this includes two electrode arrays, each with 11 contacts within a length of 8.5 mm. A depth gauge is used to determine whether the standard or double array is appropriate.

Figure 13 Nucleus Contour

The Advanced Bionics Corporation system includes the HiRes90K receiver/-stimulator and the HiFocus Ij electrode array. The receiver/stimulator uses a flexible silicone housing that surrounds a titanium case. The electrode is "banana-shaped" and curved toward the modiolus and consists of 16 contacts that are spaced every 1.1 mm over 17 mm. The diameter of the intracochlear portion ranges from 0.6 to 0.8 mm. Overall, the length of the electrode array inserted into the cochlea is 23 mm. The HiFocus Ij electrode system involves an insertion tube through which the insertion tool allows for advancement of the electrode array. Gentle pressure along a thumb-driven advancement mechanism is required to insert the electrode. Should errors occur during electrode insertion, the electrode is easily reloaded into the insertion tube, and additional electrode insertion attempts can be completed until electrode insertion is complete. But it is not common to apply in Marburg hospital, only 4% has been used in the present study, which was specified for only to its defect model.

The MED-EL C40+ system uses a receiver/stimulator that is housed in a ceramic case. The MED-EL system has three separate electrode designs. The standard electrode is the longest electrode available in the marketplace and has a conical design. Twelve pairs of electrode bands are distributed over the 31.5-mm

electrode array length. For cochlea that is partially ossified, a compressed electrode is also available; for severely ossified cochlea, a split electrode array is available. If ossification of the cochlea is encountered during the opening of the cochleostomy, the use of the MED-EL Insertion Test Device can be helpful for determining which of the various electrode options should be used. If the Insertion Test Device can be inserted to the small flanges that are present 17.8 mm from the tip, then the standard array should be used; if insertion is less than that, then the compressed array should be used. The C40+ compressed electrode (C40+S) is designed with the same number of electrode contacts (n = 12 pairs), but the total length of the electrode array is 18 mm as compared with 31.5 mm. For more severely ossified cochleas, the MED-EL split electrode design (C40+GB) has two compressed electrode arrays with five and seven pairs of electrode contacts, respectively. These electrode arrays are inserted via two cochleostomies. When the two arrays are in place, the electrode contacts provide more sites of potential stimulation than a single standard array that is incompletely inserted into the cochlea [3]. Also this model was reserved to use when the patients have a remnant hearing in the present study and the incidence was 18%.

4.6 Surgical implantation

CI in both children and adults requires meticulous attention to the delicate tissues and small dimensions. Skin incisions are designed to provide access to the mastoid process and coverage of the external portion of the implant package while preserving the blood supply of the post auricular skin. The incision has eliminated the need to develop a large post auricular flap. The inferior extent of the incision is made well posterior to the mastoid tip to preserve the branches of the post auricular artery. From here the incision is directed posterior-superiorly and then superiorly, with-out a superior-anterior limb. In children, the incision includes the temporalis muscle to give added thickness. A subperiosteal pocket is created for positioning the implant induction coil. A bone pocket well customized to the device being implanted is created, and the induction coil is fixed to the cortex with a fixation suture or periosteal flaps. Following

development of the skin incision, a mastoidectomy is performed. The horizontal semicircular canal is identified in the depths of the mastoid antrum, and the short process of the incus is identified in the fossa incudis. The facial recess is opened using the fossa incudis as an initial landmark. The facial recess is a triangular area bound by (a) the fossa incudis superiorly, (b) the chorda tympani nerve laterally and anteriorly, and (c) the facial nerve medially and posteriorly. The facial nerve can usually be visualized through the bone without exposing it. The round window niche is visualized through the facial recess about 2 mm inferior to the stapes. Occasionally, the round window niche is posteriorly positioned and is not well visualized through the facial recess or is obscured by ossification. Particularly in these situations, it is important not to be misdirected by hypo-tympanic air cells. Entry into the scala tympani is accomplished best through a cochleostomy created anterior and inferior to the annulus of the round window membrane. A small fenestra slightly larger than the electrode to be implanted (usually 0.5 mm) is developed. A small diamond bur is used to “blue line” the endosteum of the scala tympani and the endosteal membrane is removed by using small picks. This approach bypasses the hook area of the scala tympani, allowing direct insertion of the active electrode array. After insertion of the active electrode array, the cochleostomy area is sealed with small pieces of fascia. Generally this approach was organized to all the patients in the present study but rarely there was a difficulty to define the round window (small space by posterior tympanotomy), so it has been made a cochleostomy at the promontory.

4.7 Special surgical considerations

4.7.1 Cochlear dysplasia

In cases of cochlear dysplasia, a CSF gusher may be encountered on fenestrating the cochlea while performing the cochleostomy. The flow of CSF has been successfully controlled by entering the cochlea through a small fenestra, allowing the CSF reservoir to drain off, inserting the electrode into the cochleostomy, and tightly packing the electrode at the cochleostomy with fascia. It is postulated that the source of the leak is through the lateral end of the internal

auditory canal. In severe dysplasia cases with a common cavity deformity, the electrode array may be inserted directly by a trans-mastoid labyrinthotomy approach. The otic capsule is opened posterosuperior to the second genu of the facial nerve, and the common cavity is entered. Several patients have been treated in this way with no vestibular side effects [64]. Finally, one case had CSF gusher in the present study, which had cochlear dysplasia and treated by performing a cochleostomy with locked packing.

4.7.2 Aberrant facial nerve

In patients who have malformations of the labyrinth, and occasionally in patients with otherwise normal anatomy, the facial nerve may follow an aberrant course. Although not all aberrant facial nerves impact CI surgery, those that do must be recognized and dealt with effectively. Two anomalous courses of the facial nerve that place it at risk are the laterally and anteriorly displaced vertical portion of the facial nerve and a facial nerve that courses over the promontory or anterior to the round window [65]. For the safety of the facial nerve, it has been used a facial nerve monitoring as a routine in the present study.

4.7.3 Intracochlear ossification

Ossification at the round window is common in patients after meningitis and has been encountered in approximately one half of the children whose cause of deafness was meningitis who have received a CI at some centers in USA. In these patients, a cochleostomy is developed anterior to the round window and the new bone is drilled until an open scala is entered. A full electrode insertion can then be accomplished. Less frequently, labyrinthitis ossificans with extensive intracochlear bone formation may occur with complete obliteration of the scala tympani. In these cases, it is better to drill to open the basal turn of the cochlea and create a tunnel approximately 6 mm deep and partially insert a Nucleus electrode. More recently, a specially designed split electrode developed by the Med-El Corporation has been used wherein one branch of the electrode array is placed in the tunnel described above and the second active electrode is inserted into an additional cochleostomy developed just anterior to the oval

window. Finally, only one case had meningitis in the present study, but there was no ossification.

4.7.4 Surgery time

The operation time was included; the anesthesia time (about 45 minute), furthermore, the time of operation connected well with the art of the operation, the device itself, the audiologic test at the end of operation (15 minute), the preparation time of facial monitoring, the clarification of the CI surgical steps to the students, and the general state of the patient (the anatomy). So the mean average of the surgery time in the present study was 3.45 hours, the longest operate was 9 hours, because the patient had received canal down approach with abdominal fat obliteration and sew up the external auditory meatus. In medizinische Hochschule Hannover (MHH) the average time is 2 hours, but they do 5 CI operations daily and they don't use facial monitoring.

4.8 Intraoperative and postoperative complications

CI requires a surgical procedure under general anesthesia and therefore carries some risk. In particular, risks such as those encountered when removing a cholesteatoma or performing any surgery for chronic ear pathology do exist, including wound infection, facial nerve injury, taste disturbance, tinnitus, and balance problems. Overall, the complication rate of cochlear implantation has been reported as being 5–10% [4], 16% [77], 9.1% [80], 7% [79], and 9% in the present study.

A postoperative wound infection can usually be adequately treated with local wound care and antibiotics, but due to the presence of inserted foreign body, explantation of the device is occasionally required. There was not any cases required explantation in the present study, but there was reported with 1.4% in France [77]. Wound or skin breakdown can occur with an acute infection or may be related to excessive pressure of the magnet over the implant. It is important for patients to monitor the condition of the skin between the magnet and the implant device; the magnet strength can be adjusted to account for skin

thickness. Three cases in the present study had suffered from wound infections (5.4%) and with wound debridement treated; also the same incidence (5.6%) was reported at 2008 in France [77].

Facial nerve injury has been reported as well, which perhaps should not be surprising due to the wide array of aberrant anatomy potentially encountered in this unique patient population. The expectation of an abnormal nerve location and the use of intraoperative facial nerve monitoring should result in very few cases of temporary or permanent nerve injury. Fortunately, there was not any facial injury in the present study, but it has been reported one case in China [79], and also one case (0.2%) in France [77]. The corda tympani injury was reported as being 15-22% in New Zealand [78], but it was one case (2%) unilateral in the present study, which was unnoticeable from the patient and that was related to the other healthy side.

Patients need to understand that the residual hearing in the ear with the implant is likely to be lost and that a hearing aid will be of no benefit. Cochlear trauma from device insertion not only results in a loss of hearing, but it also may lead to make worse tinnitus. When encountered in this setting, tinnitus will typically lessen in time and often markedly improves following device programming. In the present study; one case had a tinnitus and was self limited after using the CI. Also it has been reported that CI had positive effect on tinnitus and could also induce its partial or total suppression in the contralateral ear [82].

Violation of the restrictions of the inner ear may also result in vestibular dysfunction with temporary balance problems and has been reported as 7% in China [79]. However, permanent balance difficulty has, in rare cases, been reported as well [4]. Accordingly, if the patient is at all suspected of having contralateral vestibular dysfunction, a preoperative ENG should be considered. In the present study there were six cases (10%) had self limited vestibular symptoms, and one case (2%) had spent 10 days in the hospital until it had cured from vertigo and its general heart vascular system, also the mean average of hospitality was 5 days.

Although the implanted device has no moving parts to wear out, there are still instances of electronic malfunction or failure due to trauma. Mechanically spoiled devices can usually be replaced with good results. Two cases in the present study (4%) have been exchanged because of device's deficiency and it has been reported as being 7.2% in France [77].

The risk of meningitis in implant recipients is being inspected. Patients with inner ear malformations have a higher risk of meningitis pre and post operatively unrelated to the CI. The role of the electrode design and its impact on the risk of meningitis is under investigation. It is wise for adult and pediatric implant recipients to receive the available pneumococcal vaccine; additionally, children should be vaccinated against hemophilus.

4.9 Assessment of outcomes

After the patient has healed from surgery, usually in 2–4 weeks, the device hardware is fully engaged and programmed. The initial programming is often done over 2–3 days. There are a countless of variables that can be adjusted to improve the sound quality. After the first day, most adults will report that speech sounds like static or voices sound either like "Donald Duck" or metallic in character. Amazingly, without any changes to the device, over the next 24 hours the sound quality improves. The brain somehow manages to adapt to the signal. This learning by the brain occurs mostly within the first 3–6 months, after which the rate of improvement in sound quality slows. Most adults will have programming meetings 4–6 times in the first year and then annually or as needed. Children (particularly infants) are more difficult to program because of the lack of a consistent feed-back response regarding volume and clarity. Objective intraoperative measurements (e.g., Neural Response Telemetry) are helpful in estimating hearing thresholds and comfort levels. It is obviously very important to not provide too much gain. Children are seen more frequently for programming. Programming is critical to the success of the device and experienced audiologists are able to achieve better outcomes than less experienced audiologists.

Rarely in medicine is there a procedure that has such a profoundly positive impact on the quality of life. Successful CI is extremely rewarding for implant team members and patients equally. Multiple factors have been shown to have an outlook on the degree of benefit obtained from implantation (Table 1) [4].

4.9.1 Outcome expectations for adults

Almost all patients demonstrate improved sound detection with their CI as compared with their preoperative performance with HA, and this is especially evident in the high-frequency range, average postoperative sound field detection thresholds for warble-tone stimuli are approximately 25 to 30 dB HL for frequencies between 250 and 4000 Hz [68].

In the present study, the mean average of PTA of post-lingually adults has increased from obove100 dB without HA and 47 dB with HA to 43 dB of post-CI. In a recent study of 78 adult CI users (26 each with the Clarion, Nucleus, and MED-EL devices), the average CNC word scores at 70, 60, and 50 dB SPL were 42%, 39%, and 24%, respectively [68], in this same group of subjects, the mean HINT scores at 70, 60, and 50 dB SPL were 72%, 73%, and 57%, respectively, comparatively in the present study, the mean average of SDT was increased from 24% words with HA and 15% words without HA to 60% words of post CI by 60-80 dB, and also the mean average of SDT numbers was increased to more than 90%. These results represent average performance; however, there was a great deal of variation in scores for individuals, ranging from 0% to 100% for most measures. In general, patients perform poorer on single-syllable word tests as compared with sentence tests or numbers, and poorer in the presence of noise than quiet. There are many CI users who are able to understand sentences without lip reading cues and, therefore, can converse on the telephone.

Adults and children

1. Shorter duration of deafness
2. Better preoperative word or sentence recognition (or both)
3. Lip reading ability higher intelligence quotient (I.Q.)
4. Better preoperative residual hearing Optimized implant technology and processing strategy
5. Cause of deafness (eg, meningitis associated with poor outcomes)
6. Intact, nonossified cochlea

Additional factors in children

1. Younger age at implantation
2. Motivated family assistance
3. Oral preoperative education
4. Oral education rehabilitation program as opposed to total communication

Table1 Factors Generally Associated with Better Outcomes in CI

Although the primary objective of speech coding strategies is the perception of speech, some patients also enjoy music [3]. The majority of post-lingually deafened adults demonstrate significant preoperative to postoperative improvements on open-set speech perception measures, often as early as 1 month after the implant. As compared with post-lingual adults, some pre-lingually deafened adults (defined as having the onset of profound or severe to profound hearing loss at less than 3 to 6 years of age, depending on the respective study) demonstrate open-set speech recognition, although the percentage is smaller, and often the length of device use needed to achieve this is longer, and there was attained it by one case of pre-lingually adults in the present study, who had decreased the mean average of PTA threshold from above 115 dB without HA to 62 dB of post-CI and also increased the words and numbers of SDT mean average respectively from 0-20% with HA to 70-80% of words and above 90% of numbers post-CI. Although the average postoperative scores for individuals with pre-lingually hearing loss are generally lower than those with post-lingually hearing loss, there have been significant preoperative to postoperative improvements in speech perception reported for this group [69]. Therefore, adults with the pre-lingually onset of severe to profound hearing loss may be appropriate candidates for CI. Providing that older patients are enjoying relatively good health, there presently is no upper age limit for CI. Audiologic results for CI users between the ages of 65 and 80 years indicate significant improvements for both pre operation and postoperative comparisons [70,71] and for varied speech stimulus presentation levels[72], in the present study the major age was 76 years old. Although increased age is not a contraindication for CI candidacy, it will be important to study the effects of aging on implant performance and to determine whether additional pre implant central auditory assessment.

4.9.2 Outcome expectations for children

Auditory detection levels with a CI are expected to be similar to those for adults, which are approximately 25 dB HL for frequencies 250 to 4000 Hz. These detection levels allow access to information that is important for the development of auditory skills and communication. As with adults, when determining

expectations, it is important to stay informed of the average and the range of pediatric CI performance, the average of PTA of children after CI in the present study was 44 dB, which was above 115 dB without HA. In a publication by Geers and colleagues[73], the results of 181 pre-lingually deaf children, who received implants before the age of 5 years and who had used their CI for an average of 5 years were reported for the outcome areas of speech perception, speech production, spoken language, total language, and reading, the average scores reported for several measures were as follows: ESP-spondee 85%, ESP-monosyllable 79%, LNT-easy 48%, LNT-hard 44%, and BKB sentences 57%. Children who were good speech perceivers were also the children who exhibited superior performance for measures of speech intelligibility, language, and reading. Half of the children were enrolled in oral communication programs, and the other half were enrolled in programs that involved total communication. Those children enrolled in educational environments that emphasized auditory and spoken language development had the highest scores for speech perception, speech production, and language measures. In addition, in the present study the pre-lingually children had average of SDT after 6 months 63% of words and by numbers more than 90%, which was 20% of words with HA. Studies conducted with children indicate that earlier implantation is associated with higher performance for a given time period after the implant [74], that pre implant unaided residual hearing influences performance and the development of speech perception skills after the implant [75], and that there is a steady increase in performance over time that does not plateau during the first 3 to 5 years of implant use [76]. Generally, children who receive their implants at an older age require more time to reach their potential with the device than those who receive them at younger ages [3]. In addition, for children with the progressive or sudden onset of hearing loss, there is an expectation of excellent progress with CI and achievement of these skills with a shorter duration of CI use. Likewise, for children with some residual hearing before the implant, also an expectation of higher levels of performance in relatively shorter periods of time [3].

5 Conclusion

CI are auditory prostheses designed to link an internal device, which is interfaced with the cochlear nerve, to an external device, which uses a specific speech coding strategy to translate acoustic information into electric stimulation. This allows the transmission of acoustic information to the central auditory pathway. A sophisticated multidisciplinary team approach that addresses the varied needs of the deaf recipients is required. The essential works of the aural/oral (re)habilitation program include listening skill development, speech therapy, speech-reading training and language instruction.

This study aims to evaluate the etiology and epidemiology of hearing loss in 55 CI cases. Surgical techniques and audiometric and radiological results were assessed preoperatively and postoperatively with a minimum follow up time of six months.

The acquired and congenital hearing loss incidences were almost equal in our group of patients. All arrays of CI postoperatively were in the first turn of the cochlear basis, and the dominant used device was Cochlear Nucleus (Australia). CI is considered as an oto-surgical procedure with a low risk and low complication rate compared with other surgical techniques.

Moreover, the audiological protocol was performed like an international classification to achieve the exact indication of CI, and there is a considerable improvement in the average of PTA threshold and the average of SDT records post-CI. Similar results of hearing improvement were published by other authors.

Finally, it is important to note that the present study is retrospective. Further prospective trials are recommended to investigate the SDT two years after obtaining a phonetic rehabilitation, especially to pre-lingually deaf children and adult groups.

6 Zusammenfassung

Cochlear Implantat ist eine auditive Prothese bestehend aus zwei Teilen; dem Elektrodenträger, der mit dem Nervus cochlearis verbunden wird, und einem externen Gerät. Dieses Gerät verwendet eine Sprachkodierung, um die akustische Information in elektrische Stimulation umzuwandeln. Auf diese Weise werden die akustischen Informationen in die zentrale Hörbahn übertragen. Ein interdisziplinäres Team ist erforderlich, um den unterschiedlichen Bedürfnissen der gehörlosen Empfänger gerecht zu werden. Die nötigen akustischen oder sprachlichen Rehabilitationsprogramme bestehen aus dem Aufbau der Hörfähigkeit, Logopädie, Sprach-Lese-Training und dem Sprechunterricht.

Die vorliegende Arbeit zielt darauf ab, epidemiologische und ätiologische Daten von Patienten mit Hörverlusten in 55 Fällen zu analysieren. Die chirurgischen, audiometrischen und radiologischen Untersuchungen wurden vor und mindestens sechs Monate nach der CI ermittelt. Zusätzlich wurde das audiologische Protokoll gemäß einer internationalen Klassifizierung durchgeführt, um die genauen Indikationen für ein CI festzustellen.

Die Anzahl der erworbenen und kongenitalen Schwerhörigkeiten der untersuchten Patienten zeigte eine ähnliche Verteilung. Postoperativ wurde die korrekte Lage der Elektroden in der ersten Windung der Cochlea radiologisch bestätigt. Das am häufigsten verwendete CI-Gerät in der vorliegenden Arbeit war Nucleus (Australien). Die Ergebnisse der durchgeführten Untersuchung zeigten, dass die Cochlear Implantation mit einer geringen Komplikationsrate einhergeht. Die audiometrischen Analysen der Ergebnisse unter Berücksichtigung der PTA-Schwelle und SDT zeigten eine signifikante Hörverbesserung nach CI.

Schließlich ist es wichtig anzumerken, dass es sich bei der vorliegenden Untersuchung um eine retrospektive Arbeit handelt. Weitere prospektive Untersuchungen sind erforderlich, um den SDT vor allem bei prälingual Ertaubten nach zwei oder mehr Jahren phonetischer Rehabilitation zu ermitteln.

7 References

1. **James B, Snow Jr, P Ashley Wackym.** Ballenger's, Otorhinolaryngology, Head and Neck Surgery. Cochlear implant. 17th edition. Bc Decker. 2008, 23:476-488.

2. **Djourno A, Eyries C.** Prothèse auditive par excitation électrique a distance du nerf sensorial. *Presse Medicale. 1957,* 35:14-17.

3. **Charles W Cummings, John M Fredrickson, Lee A Harker, et al.** Cummings Otolaryngology, Head & Neck Surgery. Cochlear implant. 4th edition. Mosby, St. Louis. 2005, 4:158-173.

4. **Anil Lalwani.** Current Diagnosis and Treatment in Otolaryngology, Head & Neck Surgery. Cochlerar Implants. McGraw-Hill. 2008, 70:1-38.

5. **Van Camp G, Smith RJH.** Hereditary hearing loss. *J Pediatric Otorhinolarygology. 2005,* 69:449-455.

6. **Morton NE.** Genetic epidemiology of hearing impairment. *Ann N Y Acad Sci. 1991,* 630:16-17.

7. **McGuirt WT, Smith RJ.** Connexin 26 as a cause of hereditary hearing loss. *Am J Audiol. 1999,* 8:93-94.

8. **Gorlin RJ, Toriello HV, Cohen MM.** Hereditary hearing loss and its syndromes. *European J Human Genetics. 2003,* 11:185-188.

9. **Hinojosa R, Marion M.** Histopathology of profound sensorineural deafness. *Ann N Y Acad Sci. 1983,* 405:459-84.

10. **Starr A.** Auditory neuropathy. *Brain. 1996,* 119:741-742.

11. **Davis H, Hirsh SK.** A slow brainstem response for low-frequency audiometry. *Audiology. 1979,* 18:445-446.

12. **Kraus N.** Absent auditory brain stem response. peripheral hearing loss of brain stem dysfunction. *Laryngoscope. 1984,* 94:400-401.

13. **Starr A.** The neurology of auditory neuropathy. Sininger Y, Starr A, ed. Auditory neuropathy, San Diego. *Singular. 2001,* 37:201-210.

14. **Gorga MP.** Case of recurrent, reversible sudden sensorineural hearing loss in a child. *J Am Acad Audiol. 1995,* 6:163-164.

15. **Starr A.** Transient deafness due to temperature-sensitive auditory neuropathy. *Ear Hearing. 1998,* 19:169-170.

16. **Sininger Y, Oba S.** Patients with auditory neuropathy. Who are they and what can they hear. Sininger Y, Starr A, ed. Auditory neuropathy. San Francisco. *Singular. 2001,* 15:501-503.

17. **Trautwein PG, Sininger YS, Nelson R.** Cochlear implantation of auditory neuropathy. *J Am Acad Audiol. 2000,* 11:309-310.

18. **Friedland DR.** Cochlear implantation for auditory rehabilitation in Camurati-Engelmann disease (hereditary diaphyseal dysplasia). *Ann Otol Rhinol Laryngol. 2000,* 109:160-161.

19. **Belal Jr A.** Contraindications to cochlear implantation. *Am J Otol. 1986,* 7:172-173.

20. **Kelsall DC, Shallop JK, Burnelli T.** Cochlear implantation in the elderly. *Am J Otol. 1995,* 16:609-610.

21. **House WF, Luxford WM, Courtney B.** Otitis media in children following the cochlear implant. *Ear Hearing. 1985,* 6:24-25.

22. **Gray RF, Irving RM.** Cochlear implants in chronic suppurative otitis media. *Am J Otol. 1995,* 16:682-683.

23. **Axon PR.** Cochlear implantation in the presence of chronic suppurative otitis media. *J Laryngol Otol. 1997,* 111:228-229.

24. **El-Kashlan HK, Arts HA, Telian SA.** Cochlear implantation in chronic suppurative otitis media. *Otol Neurotol. 2002,* 23:53-54.

25. **Hamzavi J.** Radical cavities and cochlear implantation. *Acta Otolaryngol. 2001,* 121:607-608.

26. **Luntz M.** Cochlear implantation in healthy and otitis-prone children. A prospective study. *Laryngoscope. 2001,* 111:1614-1615.

27. **Yune HY, Miyamoto RT.** Medical imaging in cochlear implant candidates. *Am J Otol. 1991,* 12:7–11.

28. **Jensen J.** Tomography of the inner ear in deaf children. Radiological demonstration of two cases with the Mondini malformation. *J Laryngol Otol. 1967,* 81:27–35.

29. **Mangabeira-Albernaz PL.** The Mondini dysplasia. from early diagnosis to cochlear implant. *Acta Otolaryngol. 1983,* 95:627–631.

30. **Tucci DL, Telian SA.** Cochlear implantation in patients with cochlear malformations. *Arch Otolaryngol Head Neck Surg. 1995,* 121:833–838.

31. **Schuknecht HF.** Mondini dysplasia. A clinical and pathological study. *Ann Otol Rhinol Laryngol. 1980,* 65:1–23.

32. **Peterson G, Lehiste I.** Revised CNC lists for auditory tests*. J Speech Hear Dis. 1962,* 27:62-63.

33. **Tillman T, Carhart R, Wilbur L.** A test for speech discrimination composed of CNC monosyllabic words. *J Speech Hearing Reserch. 1995,* 38:490-502.

34. **Nilsson M, Soli S, Sullivan JA.** Development of the Hearing in Noise Test for measurement of speech reception thresholds in quiet and in noise. *J Acoust Soc. 1994,* 95:1085-1086.

35. **Bamford J, Wilson I.** Methodological considerations and practical aspects of the BKB sentence lists. Bench J, Bamford JM, ed. Speech-hearing tests and the spoken language of hearing-impaired children. *Science Direct. 2008,* 106:707-729.

36. **Pearsons KS, Bennett RL, Fidell S.** Speech levels in various environments, Bolt Beranek and Newman Report. *J Singular. 2001,* 350:15-30.

37. **Skinner MW.** Speech recognition at simulated soft, conversational, and raised to loud vocal efforts by adults with cochlear implants. *J Acoust Soc. 1997,* 101:3766-3767.

38. **Firszt JB, Holden LK, Skinner MW.** Recognition of speech presented at soft to loud levels by adult cochlear implant recipients of three cochlear implant systems. *Ear Hearing. 2004,* 25:375-387.

39. **Wilson BS.** Cochlear implants. Some likely next steps. *Ann Rev Biomed Eng. 2003,* 5:207-208.

40. **Müller J, Schon F, Helms J.** Speech understanding in quiet and noise in bilateral users of the MED-EL Combi 40/40+ cochlear implant system. *Ear Hearing. 2002,* 23:198-199.

41. **Schleich P, Nopp P, D'Haese.** Head shadow, squelch and summation effects in bilateral users of the MED-EL Combi 40/40+ cochlear implant system. *Ear Hearing. 2004,* 25:197-204.

42. **Wilson BS, Lawson DS, Finley CC, et al.** Importance of patient and processor variables in determining outcomes with cochlear implants. *J Speech Hear Res. 1993,* 36:373–379.

43. **Geier LL, Fisher LM, Barker EJ, et al.** The effect of long-term deafness on speech recognition in postlingually deafened adult Clarion cochlear implant users*. Ann Otol Rhinol Laryngol. 1999,* 108:80-83.

44. **Blamey PJ, Pyman BC, Clark GM, et al.** Factors predicting postoperative sentence scores in postlingually deaf adult cochlear implant patients. *Ann Otol Rhinol Laryngol. 1992,* 101:342–348.

45. **Shipp D, Nedzelski J, Chen J, et al.** Prognostic indicators of speech recognition performance in postlinguistically deafened adult cochlear implant user. *Adv Otorhinolaryngol. 1997,* 52:74–77.

46. **Gantz B, Tyler RS, Abbas PJ, et al.** Evaluation of five different cochlear implant designs. Audiologic assessment and predictors of performance. *Laryngoscope. 1988,* 98:1100-1106.

47. **Cohen NL, Waltzman SB, Fisher SG, et al.** A prospective, randomized study of cochlear implants. The Department of Veterans Affairs Cochlear Implant Study Group. *N Engl J Med. 1993,* 328:233–237.

48. **Rubinstein JT, Miller CA.** How do cochlear prostheses work. *Curr Opin Neurobiol. 1999,* 9:399-404.

49. **Starr A.** Auditory neuropathy. *Brain. 1996,* 119:741-742.

50. **Sininger Y, Oba S.** Patients with auditory neuropathy. Who are they and what can they hear. Sininger Y, Starr A, ed. Auditory neuropathy. *San Francisco. Singular. 2001,* 15.

51. **Peterson A.** Outcomes of cochlear implantation in children with auditory neuropathy. *J Am Acad Audiol. 2003,* 14:188-189.

52. **Moog JS, Geers AE.** Early Speech Perception Test. St Louis, Mosby. Central Institute for the Deaf, 1990.

53. **Kirk KI, Pisoni DB, Osberger MJ.** Lexical effects of spoken word recognition by pediatric cochlear implant users. *Ear Hearing. 1995,* 16:470-471.

54. **Haskins HA.** A phonetically balanced test of speech discrimination for children, Unpublished Master's thesis, Evanston, Illinois, Northwestern University, 1949.

55. **Bamford J, Wilson I.** Methodological considerations and practical aspects of the BKB sentence lists. Bench J, Bamford JM, ed. Speech-hearing tests and the spoken language of hearing-impaired children. London. *Ear Hearing. 2007, 2*8:131-132.

56. **Nilsson M, Soli S, Sullivan JA.** Development of the Hearing in Noise Test for measurement of speech reception thresholds in quiet and in noise. *J Acoust Soc. 1994,* 95:1085-1086.

57. **Robbins AM, Renshaw JJ, Berry SW.** Evaluating meaningful auditory integration in profoundly hearing-impaired children. *J Oto. 1991,* 12:151-152.

58. **Geers A, Brenner C, Davidson L.** Factors associated with development of speech perception skills in children implanted by age five. *Ear Hearing. 2003,* 24:24-25.

59. **Geshwind N, Levitsky W.** Human brain. Left-right asymmetries in temporal speech region. *Science. 1968,* 161:186-187.

60. **Wackym PA, Michel MA, Prost RW.** Electrophysiological effects of placing cochlear implant electrodes in a perimodiolar position in young children. *Laryngoscope. 2004,* 114:1355-1361.

61. **Baumgartner W.** MRI in 30 cochlear implanted patients. *J OtolNeurotol. 2001,* 22:818-819.

62. **Wilson BS.** Better speech recognition with cochlear implants. *Nature. 1991,* 352:236-237.

63. **Vandali AE.** Speech perception as a function of electrical stimulation rate. Using the Nucleus 24 cochlear implant system*. Ear Hearing. 2000,* 21:608-609.

64. **McElveen JT, Carrasco VN, Miyamoto RT, et al.** Cochlear implantation in common cavity malformations using a transmastoid labyrinthotomy approach. *Laryngoscope. 1997,* 107:1032-1036.

65. **Miyamoto RT, Kaiser AR.** Facial nerve anomalies in cochlear implantation. *J Otorhinolaryngol. 2000,* 57:131–133.

66. **Gantz BJ,McCabe BF, Tyler RS.** Use of multichannel cochlear implants in obstructed and obliterated cochleas.*J Otolaryngol Head Neck Surg. 1988,* 98: 72–81*.*

67. **Steenerson RL, Gary LB,Wynens MS.** Scala vestibule cochlear implantations for labyrinthine ossification. *J Otol. 1990,* 11:360–363.

68. **Firszt JB, Holden LK, Skinner MW.** Recognition of speech presented at soft to loud levels by adult cochlear implant recipients of three cochlear implant systems. *Ear Hearing. 2004,* 25:375-387.

69. **Zierhofer CM, Hochmair IJ, Hochmair ES.** The advance Combi 40+ cochlear implant. *J Otol. 1997,* 18:37-38.

70. **Kelsall DC, Shallop JK, Burnelli T.** Cochlear implantation in the elderly. *J Otol. 1995,* 16:609-610.

71. **Waltzman S, Cohen N, Shapiro W.** The benefits of cochlear implantation in the geriatric population. *Otolaryngol Head Neck Surg. 1995,* 108:329-330.

72. **Firszt JB, Holden LK, Skinner MW.** Recognition of speech presented at soft to loud levels by adult cochlear implant recipients of three cochlear implant systems. *Ear Hearing. 2004,* 25:375-387.

73. **Geers A, Brenner C, Davidson L.** Factors associated with development of speech perception skills in children implanted by age five. *Ear Hearing. 2003,* 24:24-25.

74. **Fryauf-Bertschy H.** Cochlear implant use by prelingually deafened. The influences of age at implant and length of device use. *J Speech Lang Hear Res. 1997,* 40:183-184.

75. **Zwolan TA.** Cochlear implantation of children with minimal open-set speech recognition. *Ear Hearing. 1997,* 18:240-241.

76. **Miyamoto RT.** Variables affecting implant performance in children. *Laryngoscope. 1994,* 104:1120-1121.

77. **Venail F, Sicard M, Piron JP, et al.** Reliability and complications of 500 consecutive cochlear implantations. *J Otolaryngol HNSurg. 2008,* 134:1276-1281.

78. **Mcmanus L J, Stringer M D, Dawes P J D.** Iatrogenic injury of the corda tympani: a systemic rewiew*. J Laryngology. 2012***,** 126: 8-14.

79. **Qui J, Chen Y, Tan P, et al.** Complications and clinical analysis of 416 consecutive cochlear implantations. *J Pediatr Otorhinolaryngol. 2011,* 75: 1143-1146.

80. **Ciorba A, Bovo R, Trevisi P, et al.** Postoperative complications in cochlear implants: a retrospective analysis of 438 consecutive cases*. J Otorhinolaryngol. 2011,* 1:1-2.

81. **Stamatiou G A, Kyrodimos E, Sismanis A.** Complications of cochlear implantation in adults. *J Laryngol. 2011,* 120:428-432.

82. **Yonehara E, Mezzalira R, Porto P R C, et al.** Can Cochlear Implants Decrease Tinnitus. *J Inter Tinnitus. 2005,* 105:173-174.

83. **Kurzweg T, Teymoortash A, Dalchow CV, et al.** The value of DVT in assessing the position of cochlear implant in temporal bone specimens. *Ear and Hearing. 2010,* 31:413-419.

8 Appendix

8.1 Study's formula:

1-Personal Identification:

- **Name:**
- **PID:**
- **Age:** O adult O children
- **Sex:** O male O female
- **State:** O Hessen O others

2-Etiology and History:

- **Congenital :** O

Pediatric neurological exam for prelingual deaf patients**:**
□ normal
□ pathologic finding:

O mental retardation O Syndrome ----------

O others ----------

- **Acquired :** O

□ meningitis □ ototoxicity
□ presbycusis □ noise-induced\acoustic trauma
□ head trauma □ others ----------

- **Time of Deafness or profound sensorineural hearing loss:**

O prelingually deafened children (congenitally or early deafened children)

O prelingually deafened adults (congenitally or early deafened adolescents and adults)

O postlingually deafened adults and children

- **Did the deaf patients use the hearing aids? :**

O yes O no

- **Did the deaf patients interrupt using hearing aids before the surgery and how long?**

O yes : ---------- years O no

- **Chronic otitis media history:**

O yes O no

3-Pre Operative diagnosis:

- **CT-scan and MRI:**

O normal

O pathologic finding:

☐ otosclerosis ☐ cochlear dysplasia

☐ Facial Nerve malformation ☐ common cave cochlea

☐ narrow Internal Acoustic Canal ☐ others ----------

- **Hearing Tests:**

Pre operative hearing results for postlingual deaf patients:

PTA threshold without hearing aids ☐ dB

PTA threshold with hearing aids ☐ dB

Speech discrimination test without hearing aids ☐ %, ☐ dB

Speech discrimination test with hearing aids ☐ %, ☐ dB

Impedance: ☐ A ☐ B ☐ C

Stapes reflex: ☐ found ☐ not found

Pre operative hearing results for prelingual deaf patients:

ABR: wave **V** ☐ dB

prolonged **I-V** or **III-V :** O yes O no

Evoked OAE: ☐ normal (30db or less) ☐ not normal (up 45db)

Impedance: ☐ A ☐ B ☐ C

Stapes reflex: ☐ found ☐ not found

4-The Surgery:

- **Operated ear:** O right O left / O unilateral O bilateral / O primary O re implant
- **The Approach:** O Round window O Cochleostomy
- **Devise:** O MedEL O Nucleus Cochlear
- **Operation Time:** ----- hours

5-Post operative measurements:

- **CT-scan or DVT:** the location of electrode array

  □ Right position □ scala tympani □ scala vestibuli

- **Hearing test results after surgery:**

Post lingual deaf patients can understand a telephone call: O yes O no

Pre lingual deaf patient's parents satisfied with hearing and communication results: O yes O no

PTA threshold (free field) for post lingual deaf patients ☐ dB

Speech discrimination test (free field) for post lingual deaf patient ☐ %, ☐ dB

ABR results for prelingual deaf patients:

- **Time of hospitalization:** ☐ days

6-Complication:

- **Complication of surgery:**

O not found O found:

☐ injury of Facial Nerve ☐ injury of Corda tympani

☐ injury of ear drum ☐ others ----------

- **Complication after surgery:**

O not found O found:

☐ tinnitus ☐ vertigo

☐ wound infection ☐ extrusion

☐others ----------

8.2 Curriculum vitae

Name	Khayat
Surname	Wehab
Date of birth	27.06.1975
Place of birth	Aleppo,Syria
Marital status	Married to Raghda Seddik
Nationality	Syrian
Parents	Nehad Khayat
	Salwa Sakal

Education

1981-1986	Eben zaidon Primary school, Aleppo, Syria
1987-1992	Al kendi secondary school, Aleppo, Syria
1993	Graduation

Studies

1993-1999	Medical school at University Aleppo, Syria
1999	Medical examination, Full license to practice Medicine
1999-2003	Residency at the Department of Otorhinolarygology, Head and Neck Surgery, University Hospital, Aleppo, Syria
2004	Full license of ENT Specialist
2004-2008	Lecturer at the Department of Otorhinolarygology, Head and Neck Surgery, University of Aleppo, Syria
2009	Attendance to the Department of Otorhinolarygology, Head and Neck Surgery, Marburg, and start with doctor thesis.

8.3 Academic teachers and lecturers

In **Aleppo, Syria**

Prof.R.Asfari,
Prof.N.Akil,
Prof.H.Kyali,
Prof.R.zerz,
Prof.F.Eysa,
Prof .A.Knama,
Prof.A.Serio,
Prof.B.Halabi,
Prof.Khory,
Prof.R.Zahrawii,
Prof.A.Hasn,
Prof.S.Ashkhanian,
Prof.M.Tasabihjii.

In **Marburg**
Prof.A.Teymoortash,
Prof.J.A.Werner.

8.4 Acknowledgment

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I am also grateful to all my colleagues in ENT Hospital, Marburg.

8.5 Sworn declaration

I solemnly declare that I have submitted this Medical doctor thesis entitled „ Clinical and audiological outcomes of cochlear implantation: A retrospective study of 55 cases" only to the Faculty of Medicine, Philipps University in Marburg for the doctoral examination. This doctoral work was performed at the department of Otorhinolaryngology, Head and Neck Surgery, led by Professor. Dr. J.A. Werner and was supervised by Prof. Dr. A. Teymoortash.

I have neither applied to be admitted for doctoral studies outside Philipps University, Marburg nor have I submitted any other work as a dissertation to any other institution within or outside Germany.

Ehrenwörtliche Erklärung

Ich erkläre ehrenwörtlich, dass ich die dem Fachbereich Medizin Marburg zur Promotionsprüfung eingereichte Arbeit mit dem Titel „Clinical and audiological outcomes of cochlear implantation: A retrospective study of 55 cases" im Medizinischen Zentrum für Hals-, Nasen-, Ohrenheilkunde unter der Leitung von Herrn Prof. Dr. A. Teymoortash mit Unterstützung durch Herrn Prof. J.A. Werner ohne sonstige Hilfe selbst durchgeführt und bei der Abfassung der Arbeit keine andere als die in der Dissertation angeführten Hilfsmittel benutzt habe.

Ich habe bisher an keinem in- und ausländischen Medizinischen Fachbereich ein Gesuch um Zulassung zur Promotion eingereicht, noch die vorliegende oder eine andere Arbeit als Dissertation vorgelegt.

Die veröffentlichung der Arbeit ist vorgesehen

Mit freundlichen Grüssen

Marburg, den 01.08.2012

Wehab Khayat