Indication and Surgical Consideration of Cochlear Implantation at Ramathibodi Hospital

Chanida Kanchanalarp MD, FICS*, Wichit Cheewaruangroj MD*,
Cheamchit Thawin MA*, Krisna Lertsukprasert MA*

** Department of Otolaryngology,Faculty of Medicine Ramathibodi Hospital, Mahidol University*

Objectives: *To demonstrate selection criteria for cochlear implant candidates as well as the outcome of quality of life (QoL) after cochlear implant surgery.*
Material and Method: *Retrospective review was performed of all cochlear implants at Ramathibodi Hospital. A total number of 33 cochlear implantations were performed during the period of 10 years from December 1995 to December 2005. Inclusion criteria were established and the audiological criteria were then evaluated using the electrical promontory stimulation test. The etiology of severe sensory neural hearing loss was detected. The CT scan and MRI of the inner ear were studied in the different causes of deafness.*
Results: *The main cause of deafness in the present study (16 adults and 17 children) was suffering from pregnancy rubella. The second one was familial congenital deafness. The CT scan studied in the rubella cases showed anatomical normal cochlea and the hereditary cause of deafness showed abnormal cochlea that caused a strong perilymphatic gusher in a 14 year- old boy. There were 16 cases of adolescent and adult patients who all had good response in the promontory stimulation test. The QoL post implantation was evaluated in regards to improvement in education and communication.*
Conclusion: *Multi-channel cochlear implantation in severe profound hearing loss patients could improve the hearing in both normal and abnormal cochlea, congenital rubella deafness and the familial cause of deafness. The outcomes of the QoL after surgery were better in hearing detection, speech perception, school performance, communication and return to work.*

Keywords: *Cochlear implant, Mondini, Congenital deafness*

J Med Assoc Thai 2006; 89 (8): 1171-7
Full text. e-Journal: *http://www.medassocthai.org/journal*

Multichannel intracochlear implants have been used to provide marked improvement in speech perception for postlingual deafened adults since 1986[1]. Profound hearing loss in children has serious consequences in the development of communication skills. This delayed speech and language development makes the assessment of children hearing difficult. This is in contrast to postlingual improvement that often can be demonstrated within a few months after implantation. The qualification of the benefits of implantation in the deaf children is further complicated by many factors affecting their development. These include age, education program, parental motivation, age at onset of hearing loss and etiology.

The objectives of the present study were to establish selection criteria and determine if postoperative speech perception results after cochlear implant were better than the preoperative results.

Background

A cochlear implant is an electronic prosthesis in the inner ear. With this device people who were born with normal hearing but later become deaf, for example, due to meningitis, or trauma, can hear sound and sometimes distinguish speech again. Therefore, they can have a better audiologic outcome and quality of life (QoL) than adults who remain deaf without a cochlear implant.

Recently, there have been 3 different cochlear implant manufacturers globally available: Cochlear

Correspondence to : Kanchanalarp C, Department of Otolaryngology,Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok 10400,Thailand.

Corporation (Australia) developed 22 channels at the beginning of the year 1986 and 24 channels in 1992; Medical Electronics (Austria) developed multi-channel high rate cochlear implant in 1994, and Advanced Bionics (USA) developed 16 channels in 1999.

A cochlear implant is made of a receiver, a head piece and a speech processor. The operation to insert the receiver and electrodes takes two to three hours. Four weeks after surgery, the implant will be activated. The head piece is worn behind the ear and contains a microphone to pick up sounds and a device to transmit them to the internal part of the implant.

A cochlear implant can help congenitally deaf children to acquire speech, to detect the environmental sound and to develop audioverbal communication skills with in one year. In the experience of the cochlear implant team at Ramathibodi Hospital, small children aged 2 years old can develop speech after implantation as early as 6 months and attend main stream kindergarten 2 years after the implantation.

Inclusion criteria of cochlear implantation at Ramathibodi Hospital

1. Severe profound bilateral hearing loss of at least 95 dB at 1000 Hz
2. Age more than 2 years old
3. Little or no benefit from hearing aid
4. Psychological and motivationally suitable
5. No anatomical contraindication to placing the implant in the cochlea (CT scan and MRI of cochlea)
6. No medical contraindication for the surgery and the anesthesia
7. Positive result to electrical promontory stimulation test in adults and adolescents to detect hearing perception of the auditory pathway and the brain
8. Financial resources
9. Familial support for rehabilitation
10. Programming and rehabilitation support from the implant center.

Implant Center

The implant center must include at least 2 ear surgeons, 2 audiologists, 2 speech pathologists, 2 special teachers, one psychologist, one special radiologist,one social worker and a team coordinator.

Material and Method

From December 1995 to November 2005, there were 33 cases whose ages ranged from 2 2/12 to 57 yrs; the ratio of male to female was 6:5 (Table 1, 2). There were 11 Nucleus devices and 22 Med-El devices. All of these cases were evaluated by audiological assessment as follow: Baby screening test with Oto-acoustic Emission (OAE), pure tone audiogram, speech discrimination, tympanogram and stapedial reflex, auditory brain evoked potential response, hearing aid evaluation after hearing aid fitting for 6 months to 1 year, electrical promontory stimulation test in adolescent and adults, electronystagmography with caloric test in adult cases. Preoperative radiologic evaluation in every case should be a computed scan of the temporal bone to identify the structure of the inner ear, middle ear and mastoid air cells, especially cochlea and vestibular apparatus. The routine chest radiography for the anaesthetist was done. The etiology of deafness was evaluated by the history of pregnancy such as viral infection, pregnancy bleeding, prematurity and neonatal asphyxia, hyperbilirubinemia, fever with convulsion or rubella in childhood, and in adults with noise exposure or systemic diseases such as diabetes or viral infection.

In the cases of congenital abnormal cochlea, they would be requested for special MRI to identify cochlea lumen and for the surgeon to prepare the size of the electronic device. There are 2 different sizes of device in the Med-El brand that could be used, standard device for normal cochlea and short device for abnormal cochlea.

Results

There were 16 cases of adolescents and adults who had the promontory stimulation test performed (Table 1). They had good responses in the test of the hearing perception and the sound differentiation. Only one case where mastoiditis was the cause of deafness had minimum response for the hearing perception but had good outcome after surgery. The etiology of deafness in the first group of 5 was congenital pregnancy rubella and had normal cochlea with 2 turns of full insertion. They have been operated on at age 10, 19, 19, 36, 36 years old. The 10- year-old-boy is now in class 5 mainstream with good communication. The 19-year-old-male finished his studies in a college and had good verbal communication and is now working in a jewelry shop owned by his father. The 19-year-old-female finished deaf school. She could hear very well with good communication but she prefers to use sign language with her deaf boy friend. The 36-year-old-male enjoys hearing and watching television at home with his family. The 36-year-old-female has started learning to write and she enjoys hearing music and talking with her family at home.

Table 1. List of the patients with promontory stimulation test positive and showed the cause of deafness

Name	Age (yr)	Operative year	Cochlea in CT	Cause of deafness
1. SJ	43	Dec 1995	normal	Noise induced
2. PA	36	Jul 1999	normal	TB & Rubella
3. TP	14	Aug 2000	Mondini	Familial deafness
4. JS	32	Jul 2001	normal	Progressive deafness
5. JW	48	Mar 2004	normal	Diabetic deafness
6. PJ	16	Apr 2004	normal	Pregnancy fever
7. KS	54	Jun 2004	normal	Lt. Acoustic tumor
8. CC	19	Aug 2004	normal	Rubella deafness
9. NK	16	Oct 2004	normal	Pregnancy fever
10. GM	36	Nov 2004	normal	Rubella deafness
11. AT	10	Nov 2004	normal	Rubella deafness
12. TN	19	Jan 2005	normal	Rubella deafness
13. LS	14	Apr 2005	normal	Unknown deafness
14. TP	16	May 2005	normal	Premature
15. CS	57	Jul 2005	normal	Chronic mastoiditis
16. NP	39	Nov 2005	normal	Progressive deafness

Table 2. List of patients without promontory stimulation test and the cause of deafness

Name	Age (yr)	Operative year	Cochlea in CT	Cause of deafness
1. KS	4	Jul 1999	mild ossifican	Bacterial meningitis
2. NR	3 7/12	Oct 2000	normal	Rubella deafness
3. PN	2 2/12	Jan 2001	normal	Pregnancy bleeding
4. KS	4 6/12	Jun 2001	Mondini	Familial deafness
5. AC	2 9/12	Sep 2001	normal	Unknown deafness
6. SU	3 5/12	Feb 2002	Mondini	Prematurity
7. NT	8	Apr 2003	normal	Pregnancy vomiting
8. NL	2 9/12	Sep 2003	normal	Pregnancy bleeding
9. PJ	2 8/12	Sep 2003	normal	Congenital heart
10. DC	2 5/12	Feb 2004	Mondini	Familial deafness
11. SN	13 7/12	May 2004	normal	Unknown deafness
12. CR	7	Oct 2004	normal	Unknown deafness
13. TN	2 6/12	Nov 2004	Mondini	Unknown deafness
14. WS	11	Jan 2005	normal	Rubella & hyperbilirubinemia
15. JW	2 8/12	Jun 2005	normal	Unknown deafness
16. YN	2 11/12	Sep 2005	Mondini	Pregnancy fever
17. GJ	4	Oct 2005	normal	Unknown deafness

The second group of 6 had progressive hearing loss and they have been operated on when they were 43, 32, 48, 54, 57, 39 years old. The first 43-year-old-male with noise-induced hearing loss went back to work and EGAT refunded his surgery and device. The 32-year-old-female returned to work as a teacher. Her sister paid for the device. The 48-year-old-male with diabetes deafness went back to work and earns much more money for his bank and the bank paid for his surgery and device. The 54-year-old-male had an acoustic tumor in the left ear and progressive hearing loss on the right ear. His right ear was operated on and he went back to work as a banker. The bank paid for the surgery and his device. The 57-year-old-male had a right mastoidectomy and left chronic mastoiditis with progressive hearing loss. His left ear was operated with a good result. He works in his office selling construction materials. The 39-year-old-female had progressive hearing loss with antituberculotic drugs. After the cochlear implant on the right ear she went back to work

as a nurse in Khon Khan and received a refund for the device and surgery from the government. All patients in the second group could communicate by telephone after one year. The third group of 5 cases of congenital deafness had been operated on when they were 14, 16, 16, 14, 16 years old. The first 14-year-old-boy had perilymphatic gusher in the cochlecstomy that could seal with no complications. He is now studying in the university after 6 postoperative years. The 16-year-old-girl had been using a hearing aid since she was 2 years old and is now studying in a college after 2 post-operative years. She can also communicate well by telephone. The second 16 year-old-girl was learning in a deaf school and did need a longer time to start talking and understanding. Two years after surgery, she is now in normal school. The 14 year-old-girl with hearing aid use since she was 2 years can hear after one year of surgery and is very happy with good communication. She can learn now in the mainstream and use the telephone. The 16 year-old-boy had not used any hearing aid before surgery but after one year postoperation he can hear but does not like to talk. The CT scan and MRI showed one case of abnormal cochlea and vestibular apparatus that caused a strong gusher in the operation. It could be sealed with the connective tissue at the cochleostomy site. This was the case of familial hearing loss and Mondini cochlea where the post implant result was excellent. The patient was able to go to the mainstream school after 2 years and he will go to the university this year. The other two congenital rubella deafness underwent cochlear implantation at the age of 36 and 19 years old. They continued to use sign language and stayed at home with their family. For the 6 cases of progressive hearing loss,they were able to go back to work after implantation and used the telephone after one year. The adolescent cases that had used hearing aids before implantation could change the school after one year post implantation and could use telephone after 4 months to one year.

In the 17 cases of children who did not have the promontory stimulation test (Table 2), only the preoperative CT scan and MRI were evaluated. There were 6 abnormal cochlea cases such as 5 Mondini cochlea and one mild ossifican of cochlea.

The etiology of congenital hearing loss in these cases were six of deafness of unknown cause, two of deafness with familial hearing loss, two of deafness of pregnancy bleeding, one of pregnancy severe vomiting, one of prematurity and hyperbilirubinemia, one of hyperbilirubinemia, two of pregnancy rubella, one of congenital abnormality of VSD, one of pregnancy fever and one of bacterial meningitis. The post-implantation results were good and all of these children could go to main stream school except for the only one case that had abnormal cochlea 1 turn and narrow acoustic canal. He still resided in the deaf school because he could not talk, but he still used the cochlear implant and had better communication than before implantation.The authors had an increasing rate of the cochlear implantation per year and showed the different devices and the re-implantation in the Fig. 1. There were 3 re-implantations of the Nucleus and one re-implantation of the Med-el due to sclerotic cochlea without MRI.

Discussion

In the present studies, routine ABR, audiogram, CT scan, MRI whenever necessary, stapedial reflex and tympanogram in every case were performed. The authors performed promontory stimulation test in 16 cases of adolescents and adult patients. They had demonstrated auditory perception on promontory stimulation test. The procedure was performed using local anesthesia in the adult patients, allowing for subjective assessment. All adult and adolescent patients reported auditory perception when tested in each ear. The authors have recommended a trial of hearing aid use for a predetermined time period and planned re-evaluation, keeping in mind the probable limited benefits with counseling of patients and families toward this end. Promontory stimulation test has been valuable, particularly in adults. The authors advised avoiding implantation in an adult ear that does not demonstrate auditory perception on promontory stimulation testing[2]. In the year 2004, one case of a 14 year-old girl in whom the authors had neither promontory stimulation test nor MRI, this cochlear implantation was not successful because of sclerotic cochlea, and therefore the authors had to use MRI and change the ear for re-implantation. Since then she is happy to use the device and could use the telephone after 6 months.

There were five cases of rubella deafness in the adult series, two cases that did not use a hearing aid before implantation and did not use the implant device after the operation because they loved to stay with the deaf community. The other ten cases of adolescent and adult patients who used a hearing aid before implantation enjoyed using the implant and changing to main stream education and work with better benefit than before implantation.

Cochlear implantation alone does not guarantee a profoundly deaf child a successful use of the

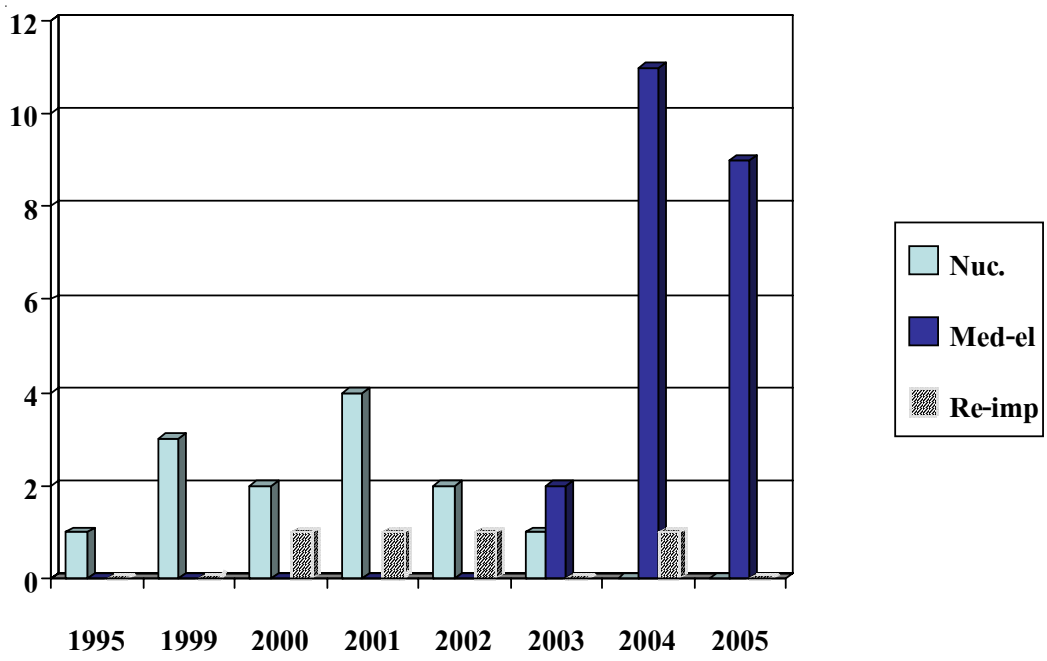

Fig. 1 Number of cases per year and the different devices and re-implantation cases

device. The implant function only enhances sound perception, but does not necessarily result in the accurate processing of the implant. Because the recognition of environment sounds is not passively acquired, the family must realize the exceptional time and effort to make the implanted device a successful communication aid[3].

As for the case of Mondini cochlea who had use hearing aid since 2 years old, had auditory perception in the promontory stimulation test at 14 years old, had the operation in the year 2000, he had a strong perilymphatic gusher where the authors were able to seal by the connective tissue in the cochleostomy hole. Postoperatively, he could use the device very successfully. He was able to attend and learn at university level and use the telephone. The use of cochlear implantation to treat patients with inner ear malformations such as Mondini dysplasia has been increasing and successful[4]. The post lingual deaf adult patient could go back to work after one month and could use the telephone after only 6 months postoperatively.

The 4 year-old child who had bacterial meningitis at two years old and subsequently had cochlear implantation at age 4, could hear and talk immediately only one month after the operation on the time of mapping and switching on the device. Post meningitis cases with no ossified cochlea received significant benefit from cochlear implantation[5]. The other congenital deaf small children who were at the age of 2 and 2/12 years to 4 and 6/12 years at the time of implantation could go to the kindergarten school and prepare for the mainstream school. Only one child who had the cochlear implantation at the time of 3 and 5/12 years could not attend the mainstream school because he could not talk. But he could understand conversation and use the device every day. This patient had a small internal acoustic meatus. The authors had to be careful to see the MRI result before operation whether there was an auditory nerve or not. Cochlear implantation is recognized as a valuable intervention with important implication for the acquisition of speech perception and verbal language in children with severe to profound hearing impairment. Auditory rehabilitation, language intervention and close co-ordination between parents, schools and the implant center are necessary to maximize efficacy[6]. Early identification of hearing aid use and language intervention and cochlear implantation by 2 years of age are positive predictors for language acquisition. Increased access to mainstream education and improvement in QoL are long term benefits that render cochlear implantation to be cost effective[7].

Conclusion

The present results with 10 years of experience in cochlear implantation showed the benefit in the QoL

in all of the children, adolescents and adults who had bilateral sensorineural hearing loss in both congenital cause and acquired cause. The authors detected that the rubella cause always showed anatomical normal cochlea, but the familial deafness showed abnormal Mondini cochlea. MRI in the familial deafness is very important for the preparation to surgery. The awareness in the case of anatomical abnormal cochlea would protect the serious complications such as the strong perilymphatic gusher that the authors could seal without lumbar puncture and also result in good hearing perception and communication in the school performance and at the university level. The promontory stimulation test that the authors used in the long time hearing loss in adult cases, could answer the question in the hearing perception ability after implantation. If the test had minimal response results, the authors would have predicted the result for the patient to know before surgery if it would be worthwhile to do. The authors have an increasing rate of cochlear implantation during the last 2 years, because the authors have had a good selection of patients along with good team screening audiologists and a team of rehabilitation that increased the outcome regarding the quality of life.

Acknowledgements

The authors wish to thank for the supervision and co-operation of experience of the following professors: GrahamClark (Australia), Andrew Van Hasselt (Hong Kong), Rudolf Haeusler (Switzerland), Manual Sainz (Spain), Joachim Mueller (Germany) and Wolf-Dieter Baumgartner (Austria). The help of Professor Amnuay Thithapandha regarding English editing is also gratefully acknowledged.

Referrences

1. Dawson PW, Blamey PJ, Rawland LC, Dettman SJ, Clark GM, Busby PA. Cochlear implantations in children, adolescents and prelinguistically deafness adults: speech perception. J Speech Hearing Research 1992; 35: 401-7.
2. Mason JC, Michael AD, Stevens C, Ruth RA, Hashisaki GT. Cochlear implantation in patients with auditory neuropathy of varied etiologies. Laryngoscope 2003; 113: 45-9.
3. Kutz W, Wright C, Krull KR, Manolidis S. Neuropsychological testing in the screening for cochlear implant candicacy. Laryngoscope 2003; 113: 57-63.
4. Daneshi A, Hassanzadeh S, Abasalipour P, Emamdjomeh H, Farhdi M. Cochlear implantation in Mondini dysplasia. ORL J Otorhinolaryngol Relat Spec 2003; 65: 39-44.
5. El-Kashlan HK, Ashbaugh C, Zwolan T, Telian SA. Cochlear implantation in prelingually deaf children with ossified cochlea. Otol Neurotol 2003; 24: 596-600.
6. Francis HW, Niparko JK. Cochlear implantation update. Pediatr Clin North Am 2003; 50:341-61.
7. Kanchanalarp C, Cheewaruangroj W, Kasemsuwan L, Thawin C, Sriwanyong S. Pediatric cochlear implantation: experience in Thai patients. J Med Assoc Thai 2005; 88: 484-91.

การคัดเลือกผู้ป่วยเพื่อผ่าตัดประสาทหูเทียมในโรงพยาบาลรามาธิบดี

ชนิดา กาญจนลาภ, วิชิต ชีวเรืองโรจน์, เจียมจิต ถวิล, กฤษณา เลิศสุขประเสริฐ

วัตถุประสงค์: *เพื่อรายงานถึงการคัดเลือกและการพิจารณาผู้ป่วยในการผ่าตัดประสาทหูเทียมโดยละเอียด สาเหตุของหูหนวกในผู้ป่วยที่มาผ่าตัดในโรงพยาบาลและคุณภาพชีวิตหลังการผ่าตัดประสาทหูเทียม*

วัสดุและวิธีการ: *ระหว่างเดือนธันวาคม พ.ศ. 2538 ถึง ธันวาคม พ.ศ. 2548 ผู้ป่วย 33 คน ได้รับการผ่าตัดประสาทหูเทียมในโรงพยาบาลรามาธิบดี การประเมินการได้ยินก่อนผ่าตัด และการตรวจกระตุ้นด้วยไฟฟ้าที่โพรมอนทารี่ในเด็กโตและในผู้ใหญ่ การตรวจคอมพิวเตอร์เอกซเรย์ของหูชั้นในในผู้ป่วยประสาทหูพิการทุกคน*

ผลการศึกษา: *สาเหตุของประสาทหูพิการอันดับแรกในผู้ใหญ่ 16 คนและเด็ก 17 คนพบเป็นหัดเยอรมัน และสาเหตุอันดับสองเป็นหูหนวกจากกรรมพันธุ์ ในการศึกษาคอมพิวเตอร์เอกซเรย์พบว่าในรายที่เป็นหัดเยอรมัน มักมีกระดูกหอยโข่งปกติส่วนรายที่เป็นจากกรรมพันธุ์นั้นมักมีกระดูกหอยโข่งผิดปกติและมีน้ำไขสันหลังพุ่งออกมาอย่างแรงขณะเจาะเข้ากระดูกหอยโข่ง 1 ราย ในเด็กอายุ 14 ปี ในเด็กโตและผู้ใหญ่ทั้งหมด 16 ราย ได้รับการตรวจกระตุ้นไฟฟ้าที่กระดูกโพรมอนทารี่พบว่าได้ผลตอบสนองในเกณฑ์ดี คุณภาพชีวิตหลังผ่าตัดดีขึ้นมากทั้งการศึกษาและการสื่อสาร*

สรุป: *การผ่าตัดประสาทหูเทียมในผู้ที่มีประสาทหูพิการทั้งสองข้างนั้นสามารถช่วยให้ได้ยินดีขึ้นทั้งคนที่มีกระดูกหอยโข่งปกติและผิดปกติ ในรายประสาทหูพิการจากหัดเยอรมันและจากสาเหตุทางกรรมพันธุ์ ผลลัพธ์ที่ได้เกี่ยวกับคุณภาพชีวิตหลังผ่าตัดดีกว่าก่อนผ่าตัดในด้านการได้ยิน การพูด การเปลี่ยนโรงเรียน การสื่อสาร และการกลับเข้าทำงานได้ตามปกติ*