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Outcomes of visually impaired patients who received cochlear implantations

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ABSTRACT

Objective: Patients with multiple sensory deficits, including hearing loss and visual impairment, present a unique problem. We evaluated the clinical outcomes of cochlear implantation in patients with severe to profound sensorineural hearing loss and visual impairment.
Methods: We retrospectively reviewed eight patients with severe sensorineural hearing loss and visual impairment who underwent cochlear implantation at our institution between 1993 and 2014. The follow-up period was between 2 and 20 years. We evaluated the case histories, etiologies of hearing loss and visual impairment, pre- and postoperative pure-tone thresholds, speech perception rates after CI using the Japanese CD speech discrimination scoring system (CI-2004 test) for words and sentences, and pre- and postoperative communication means. Postoperative speech discrimination scores were compared between patients with and without visual impairment who underwent cochlear implantation.
Results: The outcomes of cochlear implantation were good in all patients, with seven showing the ability to hold a conversation with others. The average proportion of correct answers for words and sentences in the CI-2004 test was 72.3 ± 19.1% and 86.0 ± 16.1%, respectively, for the patients with visual impairment and 62.1 ± 21.7% and 78.5 ± 20.9%, respectively, for those without visual impairment (based on auditory senses only). There were no significant differences in results between the patients with and without visual impairment.
Conclusions: Cochlear implantation is important for the rehabilitation of patients with severe auditory loss and visual impairment. Medical staff members require additional skills to perform auditory evaluations and rehabilitate patients with multiple sensory deficits.

1. Introduction

Cochlear implantation (CI) has become an established method for the auditory rehabilitation of qualified patients with severe to profound sensorineural hearing loss (SNHL), and it is widely accepted as a safe procedure [1,2]. Dual-sensory impairment or SNHL with visual impairment is a relatively rare condition, and affected patients present a unique problem, have limited options for communication, and mainly rely on tactile stimulation [3]. These patients cannot depend on other individuals who are willing to learn how to communicate with them, so they learn special finger spelling, hand writing, and tactile communication methods. The magnitude of the disability caused by dual-sensory impairment presents great difficulties for individuals with normal hearing and sight [4]. In the present study, we evaluated the outcomes of CI in eight adult patients with severe to profound SNHL and severe visual impairment.

2. Materials and methods

The records of 270 patients who participated in the adult and pediatric cochlear implant programs at our institutions from 1993 to 2014 were retrospectively reviewed. Eight adult patients with SNHL and significant visual impairment were identified (Table 1). The age range of the patients was 57–82 years (average, 65.6 ± 8.9 years), while their duration of deafness ranged from 1 to 46 years. The follow-up period was between 2 and 20 years. The causes of deafness and visual impairment in these patients are shown in Table 1. The patients' visual acuities as measured by Japanese ophthalmologists and their Snellen notations converted from decimal notations are shown in Table 2. Four patients received a CI24 and one

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Table 1
Subject characteristics.

Patient no.	Age at CI	Gender	Causes of deafness (onset years)	Causes of visual impairment (onset years)	Implant device	Sound processor (type)
1	77	F	Idiopathic progressed (60s)	High fever (12)	CI24RE	N5 (ear hunging)
2	59	M	Sudden deafness (29) Menier's disease (36)	Glaucoma, cataract (late 20s)	CI22M	Spectra (box)
3	57	M	Idiopathic progressed (50)	Retinitis pigmentosa (30s)	CI24RE	Freedom (box/ear hunging)
4	82	F	Sudden deafness (60 on right, 81 on left)	Retinochoroidal atrophy (40s)	COMBI40+	OPUS (box)
5	61	F	Idiopathic progressed (55)	Unknown (10s)	N22	MSP (box)
6	58	F	Idiopathic progressed (56)	Injury (51)	M24	SPrint (box)
7	70	F	Idiopathic progressed (67)	Congenital	CI24R(CS)	ESPrint 3G (ear hunging)
8	61	F	Idiopathic progressed (15)	Glaucoma, cataract (50s)	CI24M	3G (box)

Table 2
Pre- and post-operative evaluations of the communication means.

Patient no.	Visual acuity		Average of preoperative Pure-tone thresholds (dB)	Preoperative communication means	Postoperative communication means	Using a phone
	Right	Left				
1	0	0	110/110	Finger braille Handwritten letters	Voice conversation	Possible
2	20/1000	0	110/110	Bold and bigger letters (visual field center part only)	Voice conversation	Possible
3	20/1000	20/1000	105/110	Bold and bigger letters (visual field center part only)	Voice conversation	Possible
4	20/1000	HM	110/110	Bold and bigger letters	Voice conversation	Possible
5	20/1000	20/800	105/105	Bold and bigger letters	Voice conversation	Impossible
6	20/800	0	110/105	Bold and bigger letters	Environmental sound	Impossible
7	20/800	20/1000	130/130	Bold and bigger letters	Voice conversation	Possible
8	20/50	0	110/110	Bigger letters	Voice conversation	Possible

each received a CI22, Nucleus 22, Nucleus 24, and COMBI40+. All electrodes were inserted into the cochlea without any intraoperative problems. In addition, 90 adult patients, over fifty years of age (average, 62.0 ± 8.4 years; range, 50–84 years, 25 men 65 women), without visual impairment, who underwent CI at our institution during the same period and are currently alive, were included as a control group. The duration of deafness ranged from 4 months to 46 years, with a follow-up period of 1 to 27 years.

We evaluated the following: (1) etiology of hearing loss and visual impairment, (2) pre- and postoperative pure-tone thresholds, (3) speech perception rates after CI using the Japanese CD speech discrimination scoring system (CI-2004 test) for words and sentences, and (4) pre- and postoperative communication means. Briefly, pure-tone thresholds and the average frequency (±standard deviation) at 250, 500, 1000, 2000, and 4000 Hz were measured after CI in the eight patients with visual impairment. The proportion of correct answers for words (70 dB) and sentences (70 dB) in the Japanese CI-2004 test (speech discrimination scores) were compared between patients with and without visual impairment. Student's *t*-test was used to evaluate significant differences in the average frequency and CI-2004 test results between the two groups. Data are expressed as means ± standard deviations. *P*-values of <0.05 were considered to indicate statistical significance.

3. Results

3.1. Case histories

3.1.1. Case 1

A 77-year-old woman lost her sight after she developed high fever (details unknown) at the age of 12 years. She developed SHNL in her 60s and lost her hearing at 74 years of age. Her husband was also blind with moderate hearing loss. She desired relief from her auditory impairment and the ability to communicate with her husband. Before implantation, her only means of communication were finger braille and handwritten letters. She received a Nucleus CI24RE device and immediately acquired effortless speech tracking abilities. She was able to converse without finger braille and handwritten letters, could hold a conversation with others on the telephone, and could enjoy listening to radio or television.

3.1.2. Case 2

A 59-year-old man with visual impairment due to bilateral glaucoma and cataracts since his late 20s was diagnosed with hearing loss after experiencing sudden deafness in his left ear at 29 years of age. At 36 years of age, he developed Meniere's disease in the right ear. He was barely able to read bold, large letters. He received a Nucleus CI22M device, and his response to the rehabilitation program has been excellent. He is doing well with regard to hearing approximately 18 years after CI.

3.1.3. Case 3

A 57-year-old man developed visual impairment in his 30s because of retinitis pigmentosa. His hearing level had decreased since he was 10 years of age, and he began using hearing aids at 13 years of age. His hearing loss progressed, and he became completely deaf 7 years before CI. Although he was suspected to be Usher syndrome, he did not hope to undergo genetic testing. He received a Nucleus CI24RE device and reported subjective improvement in communication as early as 4 weeks after activation. Six months after activation, his CI-2004 score was 81% for sentences, and approximately 6 years later, he was able to communicate face-to-face with family and friends.

3.1.4. Case 4

An 82-year-old woman developed visual impairment in her 40s because of retinochoroidal atrophy. She developed sudden deafness in her right ear at 60 years of age and in her left ear at 81 years of age. She scored 0% in the speech discrimination test using closed-set sentences in the best-aided condition. Subsequently, she received a COMBI40+ device and scored 84% for words in the CI-2004 test. She is now 87 years old, fit, able to communicate even with unfamiliar speakers without difficulty, and able to use the telephone.

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