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<th>作者</th>
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Cochlear Implantation on Prelingually-Deafened Adults

Haruo Yoshida, M.D. 1), Yukihiko Kanda, M.D. 2), Ikue Miyamoto, M.D. 1), Tomomi Fukuda, M.D. 1), Haruo Takahashi, M.D. 1)

1) Division of Otorhinolaryngology, Department of Translational Medical Science, Nagasaki University Graduate School of Biomedical Sciences
2) Nagasaki Bell Hearing Center, Nagasaki, Japan

Correspondence / Reprint requests:

Haruo Yoshida, M.D.
Department of Otolaryngology, Nagasaki University Hospital
1-7-1 Sakamoto, Nagasaki 852-8501, Japan
Telephone: +81-95-849-7349, Fax: +81-95-849-7352
e-mail: y-haruo@f6.dion.ne.jp
ABSTRACT

Objective: To evaluate the validity of cochlear implantation (CI) on prelingually-deafened adults who have been trained by auditory-verbal/oral communication since childhood.

Methods: Preoperative and postoperative data was investigated regarding the rehabilitation, hearing level, and educational experience of eight prelingually-deafened adults. All eight patients were diagnosed with severe to profound sensorineural hearing loss (preoperative hearing levels were over 100dB). All used hearing aids (HA) before the age of two and were trained by auditory-verbal/oral communication since childhood. The average age of the patients at the time of their CI operations was 23.3 ranging from 18 to 29 years of age. The average postoperative observation time was 55.4 months ranging from 11 to 90 months.

Results: Improvement was achieved not only on the pure tone hearing threshold, but also in speech perception on tests using the Japanese video speech discrimination score (SDS) system. All of them now use CI very well in their daily lives and play important roles in society.

Conclusion: It was demonstrated that even prelingually-deafened adult patients could achieve considerable improvement through CI when they were trained well by auditory-verbal/oral communications since childhood. The indications of CI for prelingually-deafened adults must be determined carefully, but all of them do not have
to be rejected only because they are prelingually-deafened. In other words, CI could be recommended for prelingually-deafened adult patients if they received habilitation well with consistent auditory-verbal/oral training using well-fitted HAs.

Key Words: auditory-verbal/oral training, patient selection, and speech perception.
Introduction

It is now fully recognized that cochlear implantation (CI) is an effective treatment for patients with severe to profound sensorineural hearing loss (SNHL) [1-3]. Thousands of prelingually-deafened children and postlingually-deafened adults have shown excellent outcome in a wide range of measurements including hearing, speech, and language after CI. In previous reports, age at implantation and duration of deafness were pointed out as the most important factors for CI [2, 4], and children who received CI at an early age consistently performed better on all clinical tests than those who received CI at older age [5]. On the other hand, adolescents and adults with long-term prelingual deafness could achieve only limited postimplant improvement, and were not thought good candidates for CI [6-8]. However, a recent progress in CI technology has made it possible to considerably improve the quality levels of most CI [9-11], and the language perception after CI also can be improved through this technological progress for even prelingually-deafened patients [12, 13]. In addition, there is an increase in prelingually-deafened adult patients who were educated before CI by aurally-based educational programs, which is reported to improve the postimplantation audiological performance of patients with long-term prelingual deafness [14, 15].

In the present paper, we hypothesize that CI can be useful in some
prelingually-deafened adults if they received a good habilitation with auditory
everbal/oral manner, and report on our results of hearing status, word and speech
perception on eight prelingually-deafened adult patients, who have been trained
by auditory-verbal/oral communication since childhood and had CI.

**Materials and Methods**

The subjects were eight prelingually-deafened adult patients, which
correspond to 4.7% of totally 171 patients who underwent CI in Nagasaki
University Hospital since 1997. Detail of their clinical information is shown in
Table 1. Their ages at operation ranged from 18 to 29 with an average of 23.3
years, and the mean postoperative observation time ranged from 11 to 90
months with an average of 55.4 months. The results of the auditory brainstem
response (ABR) test before the age of two were unknown in case 4, but the other
cases showed 90dB or more from 7 months to 24 months, and all were
diagnosed as having severe to profound SNHL and started using hearing aids
(HAs) before the age of two. In the present study, we define term
“prelingually-deaf” as “severe or profound deafness with the onset before the
age of language development regardless of the presence or absence of language
development at that period”. All patients showed hearing loss over 90dB since
childhood, but there were no cases which clinically showed progressive hearing
loss. All attended ordinary schools with the exception of case 3 and 8 (school for the deaf). The education each received was of some variety, but all have been trained by auditory-verbal/oral communication since childhood. The causes of deafness were unknown in case 1, 4, 6, 7, and 8, and inner ear anomalies were found in case 5 and 7. In all cases, a full insertion of the intracochlear electrode array was obtained. A Nucleus CI 22 M (Cochlear Corp., Lane Cove, New South Wales, Australia) was implanted in case 2 and a Nucleus CI 24 M (Cochlear Corp.) was used in other cases. Their pre- and postoperative audiological data are shown in Table 2. Preoperative hearing level was over 100dB in all the cases, and two have a history of prematurity.

Results

Case 1 has used high-power box HAs in both ears since the age of one-and-a-half, and attended an ordinary elementary school where she received habilitation of auditory-verbal/oral throughout high school. After high school graduation, she worked as a care worker, but could not to hear soft voices of her patients and electronic sounds such as nurse calls during work. Preoperative imaging studies showed no abnormal findings in either CT or MR, and her cochlear nerve was observed as normal without hypoplasia. In spite of profound hearing loss, her articulation was not distorted much, probably due to the habilitation with auditory-verbal manner since the age of six. Her mean
hearing level was 100.0 dB on the right and 98.7 dB on the left at the age of 4 (Fig 1), and slowly progressed and became 105.0 dB on the right and 105.2 dB on the left (Fig 2 and Table 2). The hearing level with the BTE type HAs on the both ears was 52.5 dB on the average around the low tone, but high tones were scale-out. She received CI on the left side (better hearing side) in September, 2002, and all the active electrodes were inserted. Her mean hearing threshold with CI is 28.7 dB at present, and it is better than the preoperative level (52.5 dB) with HAs on both sides. Her Japanese video speech discrimination score (SDS) also improved from 43% to 73% with auditory and visual (A+V), and from 10% to 47% with auditory only (A). She can hear many environmental sounds such as a nurse call, ventilation fan, microwave range, and voice of the patients through the curtain, which could not be heard before surgery. She now plays an important part in society as a care worker. In comparison, apparent improvements were observed not only in pure-tone hearing level, but also in SDS (A and A+V) and speech perception rates in all the cases after CI as shown in Table 2. The preoperative articulation of all cases was distorted, but slightly improvement of articulation was recognized in case 1, 3-5, and 8 after CI. These improvements in patients’ communication ability also lead to improvements in the quality of life for each. In case 2, improvement permitted her hearing with only CI in her daily life, and she works together with hearing
people. Case 3 and 4 now work in normal hearing communities. Case 5 graduated from one of the most famous national universities in Japan, and now works as an engineer in hearing society. Case 6-8 talk to everyone more easily than before, and now working as a kimono tailor, an office lady, and a computer engineer, respectively.
Discussion

There have been many reports about the audiologic outcome of postlingually-deafened adults and prelingually-deafened young children, as well as the reports about CI of prelingually-deafened adults [6-8, 12-15]. Earlier reports of selected prelingually deafened patients using the older speech processing strategies showed less improvement after CI [6-8]. After these reports, Manrique et al. [16] followed 98 prelingually-deafened patients for one to six years after CI, and reported that post CI performance is inversely related to the duration of deafness before CI, and that patients implanted after 11 years of age showed virtually no open-set speech-perception understanding. Snik et al. [17] investigated 12 congenitally-deaf patients who received CI from four to 33 years of age, and concluded that CI during or after puberty offers only limited benefit. Thus, many of the prelingually-deafened adults have not been considered good candidates for CI because of their long duration of deafness. Although the average performance was still below the findings published for postlingually-deaf adult patients or prelingually-deaf children with short-term deafness, recent reports [12, 13, 18-20] mentioned several implanted prelingually-deaf patients with moderate improvement in speech understanding as measured by a variety of clinical tests.
In the present study, the pure-tone hearing threshold with CI was better than that with HA before operation, and improvement in speech understanding was also found apparent in all eight cases. We speculate two important factors for such good performances with CI in prelingually-deafened adults. One is their educational environment, in other words, the fact that they had been educated mainly by auditory-verbal/oral manner. This was suggested from the evidence of the aurally-based educational programs before (with HAs) and after CI could reduce the cortical colonization phenomenon and potentially improves postimplant audiological performance of patients with long-term prelingual deafness [14, 21]. The other is to start its education as early as possible with well-fitted HAs which are also indispensable to receive promising habilitation as the concept of auditory-verbal/oral manner.

In the present study, the ABR or play audiometry tests showed severe to profound SNHL and started using hearing aids (HAs) before the age of two in all cases. After that, all cases had serial hearing tests at our institution, other hospitals, and deaf schools, but no cases showed progressive hearing loss clinically. In Nagasaki, medical care by otolaryngologists and education in deaf schools has a good history of strong cooperation over the past 30 years. The basic concept of our auditory-verbal/oral habilitation is for teachers and parents to communicate over the level of 70-80 dBSPL in proximity close to
well-fitted HAs at school and at home. Furthermore, this is well informed to the family of the patients. In the current study, all patients had hearing loss over 90 dB since childhood, however, as their aided hearing level was controlled from 50 dBHL to 60 dBHL, they could learn their mother language by using auditory-verbal/oral manner with 70-80 dBSPL voice sound in a proximity close to HAs. The mother of case 1 had been had strong and tight support and education since case 1 was diagnosed as severe SNHL. Other cases also supported by their family, otolaryngologists, and staff in deaf school, to which they periodic went in case they usually went to ordinary schools. We consider that these entire environment lead to establish a good language skill and hearing preoperatively, and they can become good candidates for CI and show good CI results in spite of their prelingual deafness.

Therefore, the indications of CI for prelingually deafened adults must be determined carefully and age at CI is actually the most important factor for good outcome as previous reports, but all of them do not have to be rejected only because they are prelingually-deafened. The establishment of good aided hearing level as well as recognizable speech discrimination scores by early adapted HA and intensive auditory-verbal/oral communications are first considerations for good prognosis of CI in prelingually-deafened adult patients. We suggest CI can be recommended in some of prelingually-deafened adult
patients if they received such good habilitation with consistent auditory verbal/oral training using well-fitted HAs.
References


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Legends

Figure 1
Pure tone audiograms at the age of 4 of case 1. ○–○, ×–×: unaided threshold.

Figure 2
Fig. 2
Table 1. Clinical information of the 6 cases

<table>
<thead>
<tr>
<th>Number of cases</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
<th>8</th>
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<tr>
<td>Sex</td>
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<td>Female</td>
<td>Male</td>
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<td>Unknown</td>
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<td>Post CI (months)</td>
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<td>90</td>
<td>82</td>
<td>79</td>
<td>75</td>
<td>37</td>
<td>16</td>
<td>11</td>
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<tr>
<td>Age starting HA (months)</td>
<td>24</td>
<td>12</td>
<td>24</td>
<td>12</td>
<td>18</td>
<td>14</td>
<td>18</td>
<td>18</td>
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<tr>
<td>Education</td>
<td>Normal</td>
<td>Normal</td>
<td>Deaf school</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Deaf school</td>
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<tr>
<td>Age at CI (years)</td>
<td>29</td>
<td>19</td>
<td>18</td>
<td>24</td>
<td>24</td>
<td>22</td>
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CI: cochlear implantation, HA: hearing aid, ASCC: anterior semicircular canal, LVAS: large vestibular aqueduct syndrome
Table 2. Details of the audiological outcome of the 6 cases

<table>
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<th>Number of cases</th>
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<tr>
<td>Rt-hearing level (mean) (dB)</td>
<td>105.0</td>
<td>105.0</td>
<td>105.0</td>
<td>105.0</td>
<td>103.3</td>
<td>102.5</td>
<td>105.0</td>
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<tr>
<td>Lt-hearing level (mean) (dB)</td>
<td>105.0</td>
<td>103.8</td>
<td>105.0</td>
<td>105.0</td>
<td>105.0</td>
<td>105.0</td>
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<tr>
<td>Aided hearing level (mean) (dB)</td>
<td>52.5</td>
<td>51.7</td>
<td>63.3</td>
<td>45.0</td>
<td>65.0</td>
<td>58.7</td>
<td>60.0</td>
<td>61.3</td>
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<td>SDS (A+V) (%)</td>
<td>43</td>
<td>53</td>
<td>56</td>
<td>53</td>
<td>63</td>
<td>57</td>
<td>43</td>
<td>23</td>
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<tr>
<td>SDS (A) (%)</td>
<td>10</td>
<td>8</td>
<td>3</td>
<td>8</td>
<td>10</td>
<td>13</td>
<td>10</td>
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<td></td>
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<tr>
<td>Hearing level with CI (mean)(dB)</td>
<td>28.7</td>
<td>38.3</td>
<td>31.7</td>
<td>38.3</td>
<td>31.7</td>
<td>31.2</td>
<td>26.3</td>
<td>28.8</td>
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<td>SDS (A+V) (%)</td>
<td>73</td>
<td>86</td>
<td>86</td>
<td>90</td>
<td>70</td>
<td>67</td>
<td>47</td>
<td>53</td>
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<tr>
<td>SDS (A) (%)</td>
<td>47</td>
<td>50</td>
<td>46</td>
<td>50</td>
<td>28</td>
<td>50</td>
<td>10</td>
<td>37</td>
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<tr>
<td>Speech perception rates (%)</td>
<td>90</td>
<td>60</td>
<td>66</td>
<td>60</td>
<td>84</td>
<td>90</td>
<td>95</td>
<td>85</td>
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</table>

(A only open set-sentence without HAs)

CI: cochlear implantation, A: auditory, V: visual, HAs: hearing aids