

## Cochlear implantation in prelingually deaf persons with additional disability

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### Abstract

**Objectives:** We aimed to identify the frequency with which the following conditions were present as a second disability in cochlear-implanted, prelingually deaf persons: mild and moderate mental retardation; learning disability; attention deficit/hyperactivity disorder; cerebral palsy; congenital blindness; and autism. We also aimed to document the development of auditory perception in patients having one of these additional disabilities.

**Study design:** A retrospective study was designed to pursue the above aims.

**Methods:** We examined the records of 398 cochlear-implanted, prelingually deaf patients who had received a cochlear implant at least one year previously. Patients were selected who showed a delay in motor, cognitive or emotional development. The selected cases were referred for psychological evaluation in order to identify patients with additional disabilities. We then compared these patients' auditory perception prior to and one year following cochlear implantation.

**Results:** A total of 60 (15 per cent) cochlear-implanted, prelingually deaf patients were diagnosed with additional disabilities. These were classified as: mild mental retardation in eight cases (13.33 per cent); moderate mental retardation in five (8.33 per cent); learning disability in 20 (33.33 per cent); attention deficit/hyperactivity disorder in 15 (25 per cent); cerebral palsy in five (8.33); congenital blindness in three (5 per cent); and autism in four (6.66 per cent). All patients showed significant development in speech perception, except for autistic and congenitally deaf-blind patients.

**Conclusion:** Although cochlear implantation is not contraindicated in prelingually deaf persons with additional disabilities, congenitally deaf-blind and autistic patients showed limited development in auditory perception as a main outcome of cochlear implantation. These patients require unique rehabilitation in order to achieve more auditory development.

**Key words:** Cochlear Implants; Outcome Assessment; Language Development Disorders; Rehabilitation

### Introduction

The terms 'additional disability' and 'multiple disabilities' have been used to describe more than one disability occurring in an individual. The combined effect of hearing loss and another, concurrent disability presents a unique and complex problem. Multiple disabilities create a pattern of problems which differs from that usually associated with any one disability alone.

The frequency of other disabilities in addition to hearing loss is approximately three times as great (30.2 per cent) in the deaf or hard of hearing population as in the general school population. Some of this may be explained by the varying causes of hearing loss. Some of the current documented aetiologies of childhood deafness include maternal rubella (2 per cent), prematurity (5 per cent), cytomegalovirus (1 per cent) and meningitis (9 per cent).<sup>1</sup> It is

logical to assume that a population demonstrating hearing loss would be at higher risk of additional disabilities, since the previously mentioned aetiologies are also known to be associated with neurological effects.

Additional disabilities may include mental retardation, learning disabilities, attention deficit/hyperactivity disorder, visual impairment, cerebral palsy, orthopaedic involvement or other physical disabilities. The three additional disabilities most reported in deaf or hard of hearing children are learning disabilities, intellectual disabilities, and emotional or behavioural disabilities.<sup>2</sup>

Providing this group with cochlear implants can result in substantial benefit for both the child and the parents. Improvement in open-set speech recognition skills has been documented in congenitally and prelingually deaf children implanted at a young

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age, when no additional disability exists.<sup>3</sup> Nevertheless, prelingually deaf patients with additional disabilities are not contraindicated for cochlear implantation, although not all are suitable.<sup>4</sup> Some studies indicated poor cochlear implantation outcomes in some deaf cases with additional disabilities.<sup>5,6</sup> Cochlear-implanted individuals with additional disabilities may also be distractable, hyperactive, have a short attention span and/or memory problems, and need ancillary services (such as physical or occupational therapy) in addition to the regular rehabilitation given to cases without additional disability.

The cochlear implantation programme begun in Iran in 1991 has grown rapidly. So far, over 1000 patients have been implanted at the Iran Cochlear Implant Center, including 896 prelingually deaf children. According to the centre's diagnostic protocol (see Table I), having an additional disability is not usually considered a contraindication for cochlear implantation in children.

This study had two aims. The first was to identify the number of cochlear-implanted, prelingually deaf patients who had a second disability (including mild and moderate mental retardation, learning disability, attention deficit/hyperactivity disorder, motor delay, and autism). The second was to document the development of auditory perception in patients with one of the above-mentioned additional disabilities.

## Methods and materials

We performed a retrospective study on 398 cochlear-implanted, prelingually deaf patients with profound hearing loss (either congenital or with onset within the first year of life), in order to identify the frequency of additional disability and to evaluate auditory perception in these cases. All patients received cochlear implants between 1991 and 2002 at the Iran Cochlear Implant Center. Their ages at implantation ranged from 15 to 240 months (mean = 68 months; standard deviation (SD) = 32.2 months). The devices used for implants were 131 Med-El combi40+ (Med-El Company, Innsbruck, Austria) and the 267 Nucleus (CI 22 & CI 24) (Cochlear Ltd., NSW, Australia). Patients received auditory verbal therapy sessions twice a week for a minimum of one year following cochlear implantation.

TABLE I

PRE-OPERATIVE DIAGNOSTIC PROTOCOL

Medical history & examination
Behavioural audiometry
Objective audiometry
Evoked auditory brainstem response
Computed tomography scan
Promontory stimulation test
Pedagogic evaluation
Use of hearing aid for at least 6 months

Before cochlear implantation, brainstem evoked response audiometry testing by click stimuli (up to 110 dB HL) elicited no spikes in any patient.

All 398 cases were examined to identify any additional disabilities. The identification of additional disability was accomplished in two stages: screening and comprehensive psychological assessment.

### Screening for additional disabilities

We screened only those cochlear-implanted patients who fulfilled the following criteria: (1) delay in motor, cognitive and social-emotional domains (based on the documented results of pre-implantation developmental assessment); and/or (2) suggestion in therapists' reports of the possibility of delay in one of the above-mentioned domains.

### Comprehensive psychological assessment

Based on screening results, subjects with actual or suspected delay in the motor, cognitive or emotional domains were evaluated by a psychologist experienced in the psychology and education of exceptional children (SH). Identification of additional disability was based on the criteria of the *Diagnostic and Statistical Manual of Mental Disorders*, fourth revision, and the *International Statistical Classification of Disease and Related Health Problems*, 10th revision.

The revised Leiter international performance scale was used to assess intelligence, attention deficit/hyperactivity disorder and learning disability. This scale is a nonverbal intelligence test originally designed for deaf persons but often used with patients who cannot communicate verbally. The scale consists of two components: (1) visualisation and reasoning domains for measuring intelligence quotient, and (2) attention and memory domains to distinguish children with attention deficit/hyperactivity disorder, learning disability or neuropsychological impairment. The reliability of this test, based on internal consistency (Cronbach's Alpha) and test-retest, is  $\alpha = 0.83$  and  $\alpha = 0.85$ , respectively. The concurrent validity using Wechsler's Intelligence Scale for Children, The Third Revision (WISC-III) (in the attention composite, of which the attention sustain is one of two subtests) is 0.83.<sup>7</sup>

### Auditory perception

The Persian auditory perception test for the hearing impaired<sup>8</sup> was utilised to evaluate the auditory perception of cochlear-implanted subjects with additional disability. This test consists of 50 items, ranked in three levels based on degree of difficulty. The first level has 16 items evaluating auditory awareness, duration, intensity, pitch identification, and identification of words and sentences through suprasegmental information. The second level has 22 items evaluating vowel and consonant perception by segmental information and identification of phonemes, words and phrases, using segmental information in a closed set. The third level has 12 items evaluating comprehension in closed and open sets.

TABLE II

ADDITIONAL DISABILITIES IN COCHLEAR-IMPLANTED CHILDREN\*

Second disability	<i>n</i>	%	Boy	Girl
Mild mental retardation	8	13.33	5	3
Moderate mental retardation	5	8.33	3	2
Learning disability	20	33.33	11	9
Attention deficit/ hyperactivity disorder	15	25	9	6
Cerebral palsy	5	8.33	3	2
Congenital blindness	3	5	2	1
Autism	4	6.66	2	2
Total	60	100	35	25

\**n* = 55

The total possible score for the test is 100. The test's reliability and validity are accepted: reliability, based on the split half method with Spearman Brown formula and test-retest, is  $\alpha = 0.96$  and  $\alpha = 0.97$ , respectively; internal consistency, by the Kuder Richardson formula, is  $\alpha = 0.95$ . The construct validity of the test is  $R = 0.83$ . Items in the test were presented by a male speaker in a controlled live voice mode at an average presentation level of 70 dB Sound Pressure Level (SPL) in a soundproof room. The test was administered prior to implantation and then at three-month intervals after the device had been switched on.

In view of the small numbers in some of the second disability groups, we used the non-parametric Wilcoxon matched pairs signed-ranked test to compare the auditory perception results of cochlear-implanted children with additional disability, before and after implantation.

## Results

Evaluation of the cochlear-implanted, prelingually deaf patients with additional disabilities presented a considerable challenge. Difficulties were encountered because of limited vocabulary, short span of auditory memory, cognitive immaturity, short attention span, lack of concentration and poor cooperation with the tester over an extended period of time. However, comprehensive psychological assessment diagnosed additional disability in a total of 60 (15 per cent) of the cochlear-implanted, prelingually deaf patients. This group comprised 35 (53.8 per cent) males and 25 (46.2 per cent) females. The disability was classified as: mild mental retardation in eight (13.33 per cent) cases; moderate mental

retardation in five (8.33 per cent); learning disability in 20 (33.33 per cent); attention deficit/hyperactivity disorder in 15 (25 per cent); cerebral palsy in five (8.33); congenital blindness in three (5 per cent); and autism in four (6.66 per cent) (Table II).

Table III shows the auditory perception test results in the cochlear-implanted patients with additional disabilities, both before cochlear implantation and one year after device activation. Auditory perception improved in each of the seven groups of patients; nevertheless, much diversity can be seen. Compared with the other groups, subjects with cerebral palsy showed the maximum improvement in auditory perception (mean = 61.00; SD = 15.66), whereas those with autism showed the minimum improvement (mean = 11.00; SD = 2.58). The Wilcoxon matched pairs signed-ranked test was used to identify the significance of differences between the mean scores of auditory perception before and one year following cochlear implantation. The differences were significant in subjects with mild mental retardation ( $p < 0.012$ ), moderate mental retardation ( $p < 0.043$ ), learning disability ( $p < 0.001$ ), attention deficit/hyperactivity disorder ( $p < 0.000$ ) and cerebral palsy ( $p < 0.043$ ). However, such differences were not significant in patients with congenital blindness ( $p < 0.102$ ) and autism ( $p < 0.068$ ).

## Discussion

We retrospectively identified the frequency with which mild and moderate mental retardation, learning disability, attention deficit/hyperactivity disorder, cerebral palsy, and autism were present as a second disability in cochlear-implanted, prelingually deaf patients. We also assessed the auditory perception in these children.

In our study, the frequency of other disabilities in addition to hearing impairment was 15.07 per cent. Other authors have found a frequency of 30.2 per cent.<sup>1</sup> This considerable difference might be due to the fact that the latter result was for subjects of school age; the diagnosis of additional disabilities (specifically learning disabilities) is much more feasible at that age.<sup>9,10</sup> In addition, we excluded children with orthopaedic abnormalities, while the latter study did not.

It should be noted that severe mental retardation was considered a contraindication for cochlear implantation in our programme.

TABLE III

AUDITORY PERCEPTION RESULTS BEFORE AND ONE YEAR AFTER COCHLEAR IMPLANTATION IN CHILDREN WITH ADDITIONAL DISABILITY

Second disability	<i>n</i>	Pre-CI (mean (SD))	Post-CI (mean (SD))	<i>p</i>
Mild mental retardation	8	2.750 (1.832)	45.000 (23.299)	0.012
Moderate mental retardation	5	2.800 (2.588)	37.200 (18.952)	0.043
Attention deficit/hyperactivity disorder	15	2.466 (1.641)	40.800 (17.271)	0.001
Learning disability	20	3.150 (1.565)	31.250 (17.314)	0.000
Cerebral palsy	5	3.200 (2.167)	61.00 (15.668)	0.043
Congenital blindness	3	1.000 (1.000)	15.000 (2.645)	0.102
Autism	4	0.500 (0.577)	11.000 (2.582)	0.068

Pre-CI = before cochlear implantation; post-CI = one year after cochlear implantation; SD = standard deviation

Deaf children with additional disabilities differ enormously. This creates a very challenging problem in terms of evaluation, planning and implementing rehabilitation.<sup>4–6</sup> There are a variety of criteria for identifying deaf persons with additional disabilities. Much of the diagnosis procedure is not substantially different from that for deaf or hard of hearing individuals without additional disabilities. Developmental checklists may indicate problems in areas such as motor skills, self help and social skills. Parents' reports of their child's development and difficulties can often be a clue.

- **This retrospective study investigated the frequency of some common second disabilities in cochlear-implanted, prelingually deaf persons, and also evaluated their improvement in auditory perception following cochlear implantation**
- **The records of 398 patients who had undergone cochlear implantation at least one year previously were examined in order to identify patients with delayed motor, cognitive or emotional development**
- **Congenitally deaf-blind and autistic patients had limited development in auditory perception as the main outcome of cochlear implantation. These patients require unique rehabilitation for optimum development**

Many deaf patients with additional disabilities often experience a great deal of failure, due both to their disabilities and to the limited number of therapists trained to work with them. Persons with additional disabilities may dislike rehabilitation sessions for this reason, and this may cause behaviour problems.

### Conclusion

Although cochlear implantation is not contraindicated in prelingually deaf persons with additional disabilities, not all such patients are considered good candidates. In our study, congenitally deaf-blind and autistic children showed limited

development in auditory perception, as a main outcome of cochlear implantation.

Deaf patients with additional disabilities require unique rehabilitation. The team responsible for planning such rehabilitation for cochlear-implanted, prelingually deaf persons with additional disabilities may include parents, special education experts, speech pathologists, occupational therapists and audiologists.

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