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Auditory and language skills development after cochlear implantation in children with multiple disabilities

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Abstract

Background Cochlear implantation (CI) in children with additional disabilities can be a fundamental and supportive intervention. Although, there may be some positive impacts of CI on children with multiple disabilities such as better outcomes of communication skills, development, and quality of life, the families of those children complain from the post-implant habilitation efforts that considered as a burden.

Objective To investigate the outcomes of CI children with different co-disabilities through using the Meaningful Auditory Integration Scale (MAIS) and the Meaningful Use of Speech Scale (MUSS) as outcome measurement tools.

Methods The study sample comprised 25 hearing-impaired children with co-disability who received cochlear implantation. Age and gender-matched control group of 25 cochlear-implanted children without any other disability has been also included. The participants' auditory skills and speech outcomes were assessed using MAIS and MUSS tests.

Results There was a statistically significant difference in the different outcomes measure between the two groups. However, the outcomes of some multiple disabilities subgroups were comparable to the control group. Around 40% of the participants with co-disabilities experienced advancement in their methods of communication from behavior to oral mode.

Conclusion Cochlear-implanted children with multiple disabilities showed variable degrees of auditory and speech outcomes. The degree of benefits depends on the type of the co-disability. Long-term follow-up is recommended for those children.

Keywords Children with disabilities · Hearing impairment · Cochlear implants · Language development

Introduction

According to the 2018 report of WHO, hearing loss is estimated to be the fourth leading cause of disability globally that affects millions of people around the world [1]. It has been estimated that approximately 40–50% of children with hearing loss have an additional medical or developmental disability [2–4]. This additional disability may include, and not limited to, physical disabilities, visual impairment, developmental delay, cerebral palsy, autism spectrum disorders, attention-deficit hyperactivity, mental retardation, and learning disabilities. The coincidence of two or more disabilities early in the life of those hard of hearing children creates a more complex condition that needs more challenging approaches for management [4]. For instance, this group of children represents a challenge regarding proper auditory amplification and rehabilitation programs.

Cochlear implantation (CI) in children with additional disabilities can be a fundamental and supportive intervention [4]. In the early advances of cochlear implantation programs, children with co-disability apart from hearing loss were considered not candidates to receive cochlear implants. However, recently the candidacy criteria for pediatric cochlear implantation are getting more flexible to include some hard of hearing children with other disabilities and complex needs [5]. Although, there are some positive impacts of CI on children with multiple disability communication, development and quality of life, the families of those children

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complain from the post-implant habilitation efforts that are considered as a burden [6–8].

The main limiting factor for cochlear implantation centers to include children with multiple disabilities is the expected poor outcomes of the rehabilitation process. Nowadays, with the increased rehabilitation experience, some centers consider more children with multiple needs for assessment and enrollment in CI programs. Although some studies investigated the quality of life issues in cochlear-implanted children with other disabilities [8, 9], few data are available regarding the rehabilitation outcomes in terms of auditory and verbal communication skills of those children.

There are different outcome measurement tools for the auditory and verbal communicative skills that can be used to assess the abilities of the CI children following implantation and rehabilitation programs. The Meaningful Auditory Integration Scale (MAIS) and the Meaningful Use of Speech Scale (MUSS) are commonly used for evaluating rehabilitation outcomes of CI children. MAIS has been developed to assess auditory skills and behavior of CI children, while MUSS was established to assess the use of spoken language in CI children (Appendix A and Appendix B) [10, 11].

The main objective of this study is to investigate the outcomes of CI children with different co-disabilities through using the Meaningful Auditory Integration Scale (MAIS) and the Meaningful Use of Speech Scale (MUSS) as outcome measurement tools.

Subjects and methods

Participants

The Institutional Review Board at College of Medicine, King Saud University has approved the study under IRB Number E-14-1217. This is a case–control study that comprised 25 hearing-impaired children with co-disability who received cochlear implantation at our center. A comprehensive files review has been done to identify all children who had received a CI and had been diagnosed with additional disabilities. Age and gender-matched control group of 25 cochlear-implanted children without co-disabilities has been also included. The following inclusion criteria were applied to select the sample: (1) all children should be pre-lingual; (2) they had severe to profound sensorineural hearing loss; (3) all participants have used a hearing aid for a period ranged between 3 and 6 months before surgery; (4) CI had been used for at least 12 months; (5) all children underwent rehabilitation for at least 12 months using auditory-verbal therapy; and (6) documented diagnosis of at least one disability in addition to deafness. Table 1 shows the demographics of the study group with their associated disabilities.

Rehabilitation program

All children enrolled in the study have attended the auditory-verbal therapy sessions regularly and parents were given a home-program to apply with their children that was followed up with the speech pathologist. Children in the disability group received rehabilitation therapy for a mean duration of 30.92 (±8.53) months, while the mean duration of rehabilitation in the control group was 30.51 (±8.04) months.

Auditory skills outcomes

Meaningful Auditory Integration Scale (MAIS) was used to assess the children auditory skills. This is a 10-questions tool in which parents are asked to report to the best of their knowledge the behavior and reactions of their children in response to noise and their ability to sustain using the cochlear implant device on throughout the day. A maximum of 40 points is obtainable under the scoring system. Each of the ten questions is scored on a five-point scale from 0 (lowest) to 4 (highest). There are three divisions in scoring MAIS including reliance on using the device (item 1–2), alerting to sounds (item 3–6), and deriving meaning from sound (item 7–10). The general score is obtained by adding the scores for each area. In addition to using the MAIS in evaluating the auditory skills outcomes from the parents' perspective, assessment of hearing was done by comparing the aided hearing threshold results between the patients' group and the control subjects.

Language and speech outcomes

Meaningful Use of Speech Scale (MUSS) was used to assess the speech outcomes of the participants. Similar to MIAS, MUSS is also a 10-item questionnaire in which the parents rate speech abilities of their children on a scale from 0 (lowest) to 4 (highest). There are 3 divisions in scoring MUSS including the sound generation (item 1–3), the communication capability (item 4–7), and communication skills (item 8–10). The general score is obtained by adding the scores for each area.

Statistical analysis

This study included both qualitative and quantitative variables. Quantitative variables were represented by mean and standard deviation, while qualitative data were represented by proportions. According to the normality test, our data were not normally distributed and non-parametric statistical analyses were applied. The Mann–Whitney test was used to examine differences between groups with a level of

Table 1 Descriptive characteristics of the disability group

N	Disability	Gender	Age	Age at implant	Hearing age	Communication mode		Laterality of surgery
						Pre-implant	Post-implant	
1	Usher	M	9.9	2.9	7.0	Behavior	Behavior	Bilateral
2	Usher	M	7.2	5.0	2.2	Behavior	Behavior	Unilateral
3	Usher	M	7.1	1.7	5.4	Behavior	Oral	Bilateral
4	Usher	F	9.7	8.1	1.6	Behavior	Oral	Bilateral
5	Usher	M	12.5	10.9	1.6	Behavior	Oral	Bilateral
6	Usher	F	14.6	6.3	8.4	Behavior	Oral	Unilateral
7	ASD	M	7.9	3.6	4.3	Behavior	Oral	Unilateral
8	ASD	F	6.5	2.1	4.3	Behavior	Behavior	Bilateral
9	ASD	M	8.7	4.3	4.5	Behavior	Behavior	Bilateral
10	ASD	M	7.3	2.8	4.5	Behavior	Behavior	Bilateral
11	ASD	M	5.2	4.1	1.1	Behavior	Behavior	Unilateral
12	ASD	M	10.5	5.9	4.7	Behavior	Behavior	Bilateral
13	ASD	F	7.9	4.1	3.8	Behavior	Behavior	Unilateral
14	ASD	F	8.9	2.0	6.9	Behavior	Behavior	Unilateral
15	ASD	M	4.8	1.4	3.3	Behavior	Behavior	Bilateral
16	GDD	M	2.9	1.7	1.2	Behavior	Behavior	Unilateral
17	GDD	M	2.3	1.4	1.0	Behavior	Behavior	Bilateral
18	GDD	F	3.2	1.0	2.2	Behavior	Behavior	Unilateral
19	GDD	F	6.0	4.3	1.7	Behavior	Behavior	Unilateral
20	GDD	M	13.8	5.7	8.1	Behavior	Oral	Unilateral
21	GDD	F	4.8	0.4	4.5	Behavior	Oral	Bilateral
22	ADHD	F	11.5	4.3	7.3	Behavior	Oral	Unilateral
23	ADHD	M	3.7	2.4	1.3	Behavior	Behavior	Bilateral
24	ADHD	F	8.3	2.5	5.8	Behavior	Oral	Bilateral
25	ADHD	M	6.3	2.4	3.9	Behavior	Oral	Bilateral

ASD autism spectrum disorder, *GDD* global developmental delay, *ADHD* attention-deficit hyperactivity disorder

significance at 0.05. Cross tabulation and χ^2 were used to compare proportions of qualitative variables. The Statistical Package for the Social Sciences, version 17 (SPSS Inc., Chicago, Ill., USA) was used for all statistical analysis.

Results

A total of 50 cochlear-implanted children have been included in the study. The characteristics of the study group are demonstrated in Table 1. There were 25 children in the disability group and 25 children served as control. The mean age of the disability group was 7.7 years (±3.29), while that of the control group was 6.7 years (±2.84). Fifty-six percent of the disability group received bilateral CI, whereas 44% of the control group had bilateral CI. Both groups were matched regarding the age at implant and hearing age. Both groups were matched regarding age, gender, hearing age, age at implant, and laterality of CI surgery (Table 2).

The disability group was subdivided according to the type of the disability into four subgroups: children with Usher

Table 2 Comparison between disability and control group regarding demographic data and CI surgery-related characteristics

	Patients (*N*=25)	Control (*N*=25)	*P*
Age mean (SD)	7.7 (3.29)	6.7 (2.84)	0.3
Gender			
Male *N* (%)	15 (60%)	12 (48%)	
Female *N* (%)	10 (40%)	13 (52%)	0.39
Age at implant mean (SD)	3.65 (2.40)	3.42 (1.77)	0.69
Hearing age mean (SD)	4.02 (2.30)	3.33 (0.81)	0.24
Laterality of surgery			
Bilateral *N* (%)	14 (56%)	11 (44%)	
Unilateral *N* (%)	11 (44%)	14 (56%)	0.57
Pre-implantation communication mode			
Behavioral *N* (%)	25 (100%)	25 (100%)	
Oral *N* (%)	0 (0%)	0 (0%)	
Post-implantation communication mode			
Behavioral *N* (%)	15 (60%)	1 (4%)	
Oral *N* (%)	10 (40%)	24 (96%)	0.0001

syndrome ($N=6$), children with global developmental delay ($N=9$), children with autism spectrum disorder (ASD) ($N=6$), and children with attention-deficit hyperactive disorder (ADHD) ($N=4$).

Communication mode

Communication mode outcomes are demonstrated in Table 1. Communication skills were investigated pre- and post-implantation. (This was done through the speech therapist team during rehabilitation sessions.) Families described in great details their child's individual pre- and post-implantation communication skills. Skills included either the child is using behavior mode or oral mode in communication. All participants in both groups were using behavior communication mode before cochlear implantation. Post-implantation, 15 out of 25 children in the case group made communication progress. In the control group, all the children except one were advanced to oral mode. Of the six children with Usher syndrome, four advanced to oral communication mode. Three out of the four attention-deficit hyperactive disorder (ADHD) used oral mode post-implantation. In children with autism spectrum disorder subgroup, eight participants out of nine remained using the behavior communication mode. Two out of sex of the children with developmental delay were advanced to oral mode. However, by comparing the overall communication mode between the study group and the control group, there was a statistically significant difference in favor of the control group regarding developing oral communication (Table 2).

Auditory (hearing) evaluation

Aided thresholds have been estimated for all the participants. In general, there was a statistically significant difference between the two study groups. The mean aided threshold for case and control groups was 40.89 dB and 28 dB, respectively. When comparing each case subgroup with the control group, there was no statistically significant difference except with the autism subgroup ($p=0.0001$) (Table 3).

Auditory skills outcomes

Auditory skills outcomes were measured through MAIS questionnaire. There was a statistically significant difference between the two groups ($P=0.0001$). When comparing each subgroup with the control group, it was found that only the Usher syndrome subgroup has obtained comparable auditory skills level as the control group. The autism spectrum disorder subgroup experienced the worst MAIS score compared to the control group (Table 3).

Language and speech outcomes

MUSS was used to assess the language and speech skills for all participants. A significantly statistical difference was obtained between the two groups. The average scale was 29.88 and 13.52 for the control and the study groups, respectively. Again, it was found that there was no statistical difference between the Usher syndrome subgroup and the control group (Table 3).

Discussion

In recent years, the number of hearing-impaired children with co-disabilities undergoing cochlear implantation surgery is increasing. Considering the few publications in this aspect, the outcomes of those children regarding both the auditory and communicative skills are not well documented

Table 3 Comparison between the disability and control groups regarding auditory and speech outcomes post-implantation

Groups	Hearing age			Age at implant			Aided threshold			MAIS			MUSS		
	Mean	SD	*P*	Mean	SD	*P*	Mean	SD	*P*	Mean	SD	*P*	Mean	SD	*P*
Usher ($N=6$)	4.36	2.97	0.88	5.81	3.38	0.14	33.54	10.53	0.77	38.5	3.67	0.99	25.83	11.03	0.88
Control ($N=25$)	3.33	1.78		3.42	1.77		28.00	4.77		40.00	0.01		29.88	7.61	
GDD ($N=9$)	3.11	2.74	0.99	2.41	2.09	0.68	39.58	15.50	0.11	23.83	14.74	0.006	12.5	10.11	0.001
Control ($N=25$)	3.33	1.78		3.42	1.77		28.00	4.77		40.00	0.01		29.88	7.61	
ASD ($N=6$)	4.15	1.51	0.90	3.36	1.41	0.98	49.86	13.23	0.0001	18.88	15.43	0.001	7.33	7.56	0.0001
Control ($N=25$)	3.33	1.78		3.42	1.77		28.00	4.77		40.00	0.01		29.88	7.61	
ADHD ($N=4$)	4.57	2.58	0.87	2.90	0.93	0.99	33.68	1.02	0.85	24.75	10.40	0.04	10.5	4.43	0.003
Control ($N=25$)	3.33	1.78		3.42	1.77		28.00	4.77		40.00	0.01		29.88	7.61	
Total													13.52 29.88	11.05 7.61	0.0001
Patients ($N=25$)	4.02	2.30	0.24	3.65	2.40	0.69	40.89	3.58	0.0001	25.72	14.15	0.0001			
Control ($N=25$)	3.33	1.78		3.42	1.77		28.00	4.77		40.00	0.01				

ASD autism spectrum disorder, *GDD* global developmental delay, *ADHD* attention-deficit hyperactivity disorder

[5]. No one can ignore the difficulty of cochlear implantation surgery decision in children with multiple disabilities considering the prediction of the outcome following the surgery. Understanding the needs of those children and their family expectations about the surgical outcomes is essential to be acknowledged for providing better support for this special group of hearing-impaired population [12].

The aim of this study was to investigate the outcomes of CI surgery in a group of hearing-impaired children with different co-disabilities in comparison to a control group of hearing-impaired children without additional disabilities. The MAIS and MUSS in addition to aided hearing assessment were used as measurement tools for the surgical outcomes in both groups. Both groups were matched regarding gender, age at implant, hearing age and the duration of rehabilitation they received following implantation. The outcomes of the CI surgery were significantly higher in the control group compared to the study subjects considering the aided hearing threshold, MAIS, and MUSS. However, the responses of the children within the study group varied according to the specific disability they have.

Usher syndrome group

Usher syndrome is an autosomal recessive inherited disorder that is considered the most common cause of genetically associated deafness and blindness. Affected subjects suffer from congenital sensorineural hearing loss and progressive retinitis pigmentosa [13, 14]. There are three subtypes of this syndrome that varies in their clinical presentations. However, the subjects in our study group were all of subtype I in which children present with congenital profound sensorineural hearing loss and gradual progressive visual impairment that starts at late childhood. Usher group demonstrated the most favorable outcomes compared to other disability groups at all levels including the auditory skills, communicative skills, and the measured aided response that were comparable to the results of the control group. These results can be explained by the type of the disability those children have at the age of implant, as all of the children in this group had only the hearing impairment disability, while most of those subjects demonstrated almost normal visual abilities and had not developed visual impairment yet. Thus their results were comparable to the control subjects. These results were in accordance with other findings of similar studies such as that of El-kashalan et al. [15] and Al-Zhrani et al. [16] who reported good outcomes of Usher syndrome children following cochlear implantation. Similar favorable results were reported by other studies investigated the post-implantation outcomes in Usher syndrome children [17, 18]. This finding signifies the importance of having early CI surgery for children with Usher syndrome to ensure having better outcome results before the development of visual impairment that might hinder the rehabilitation process.

Global developmental delay group

Global developmental delay is commonly described as a subset of developmental disabilities in two or more of the following developmental domains; gross/fine motor, speech/language, cognition, social/personal, and activities of daily living [19]. In our study group of children with developmental delay, there was a significant delay in cognitive and motor abilities compared to the control subjects. Accordingly, their post-implantation outcomes were significantly lower than that of the control subjects. These results were in agreement with the results reported by Edwards et al. [20] who found poor speech outcomes post-implantation for a small group of children with a significant developmental delay compared to their normally developed implanted peers. Similar results of poor outcomes following cochlear implantation for children with developmental delay were reported also by other related studies [21, 22]. These poor outcome results can be explained by the poor cognitive and motor abilities this group of children had that hindered their progress following implantation.

Autism spectrum disorder (ASD) group

Autism spectrum disorders (ASD) is considered one of the most challenging disabilities a cochlear implant team could face for taking a decision whether to implant or not. One of the main challenging aspects in this disorder is the difficulty of early diagnosis. Accordingly, a child can be implanted then diagnosed with ASD later on after implantation. Unfortunately, due to their poor communicative and social skills in addition to the sensory integration problems they could have, the outcomes of cochlear implantation in this group of children is not favorable regarding communication and speaking abilities. Because of that, in the past, those subjects were considered not candidates for receiving cochlear implantation surgery [5].

Not surprisingly, the ASD group in our study had the poorest outcome results amongst the disability group compared to the control subjects regarding the communicative and auditory skills outcomes. These poor outcome results were a common finding as well in the study of Donaldson et al. [23] who reported difficulty in assessing the speech and language outcomes in their study group of ASD children due to the developmental problems they have. However, they reported in their study positive responses from the parents of those children regarding some benefits they noticed on their children following implantation including awareness to environmental sounds, reaction to music, and improved eye contact. Thus it is important to explain to the family of

those patients the realistic outcomes of the surgery that may include improvement in some aspects of quality of life rather than improvement in the oral or communicative abilities.

Attention-deficit/hyperactivity disorder (ADHD) group

Attention-deficit/hyperactivity disorder (ADHD) is defined as one of the most common neurodevelopmental disorders that is characterized by a persistent pattern of inattention and/or hyperactivity–impulsivity that may interfere with language acquisition and development [24]. Favorable outcome results in regards to speech perception and language abilities of children with ADHD who received CI were reported by other studies [8, 25, 26].

In our study group, three out of four children with ADHD achieved oral communication post-implantation reflecting evident benefit from implantation. However, the results of the MUSS compared to the control subjects did not reach a significant level. This can be explained by the inattentive problem and central cognitive deficits those children have that can hinder their responses in the rehabilitation program. Accordingly, it has been recommended that those children be identified properly pre-implantation and to start behavioral modification program as early as possible to avoid obstacles in the rehabilitation process following implantation [27].

Conclusion

Hearing-impaired children with co-disabilities can demonstrate some benefits from receiving cochlear implantation surgery including not only the auditory skills but also the communicative and language acquisition skills. However, those benefits are limited and cannot be compared to the outcomes obtained in hearing-impaired children without additional disabilities who significantly gain better results on both auditory and communicative abilities following implantation. Accordingly, cochlear implantation decision for children with multiple disabilities should be cautiously considered in this group of patients and should be studied on an individual basis according to the potential excepted benefits of the surgery for each case.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

Appendix A

Meaningful Auditory Integration Scale (MAIS)

1. Score item 1a if the child is younger than age 5 and item 1b if the child is older than age 5 and 1c unable to score 1a and 1b

   a. Does the child wear the device all waking hours without resistance?
   b. Does the child ask to have his or her device put on or put it on him/herself without being told
   c. Is the child's vocal behavior affected while wearing hearing his/her aids?

2. Score item 2b if unable to score 2a

   a. Does the child report and/or appear upset if his/her device is non-functioning for any reason
   b. Does the child produce well-formed syllables and syllable sequences that are recognized as "speech"?

3. Does the child spontaneously respond to his/her name in quiet when called (hearing-only), with no visual clues?
4. Does the child spontaneously respond to his/her name in the presence of background noise when called (hearing-only), with no visual clues?
5. Does the child spontaneously alert to environmental sounds (doorbell, telephone) in the home without being prompted or told to listen?
6. Does the child spontaneously alert to environmental sounds in new environments?
7. Does the child discriminate auditory signals that are part of his daily routine?
8. Does the child discriminate two speakers by listening only?
9. Does the child spontaneously know the difference between speech and non-speech stimuli?

10. Does the child spontaneously associate vocal tone with its meaning based on listening only?

Appendix B

Meaningful Use of Speech Scale (MUSS)

1. Vocalizes during communication interactions.
2. Uses speech to attract other's attention.
3. Vocalizations vary with content and intent of messages.
4. Is willing to use speech primarily to communicate with familiar people on known topics.
5. Is willing to use speech primarily to communicate with unfamiliar people on known topics.
6. Is willing to use speech primarily to communicate with familiar people on novel topics.
7. Is willing to use speech primarily to communicate with unfamiliar people on novel topics.
8. Produces messages understood by people familiar with his speech.
9. Produces messages understood by people unfamiliar with his speech.
10. Uses appropriate repair and clarification strategies.

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