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Communicative and Hearing Performance in Individuals with Cochlear Implants and Delayed Neuropsychomotor Development: A Longitudinal Analysis

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Abstract

Introduction Between 15% to 30% of individuals with bilateral prelingual sensorineural hearing loss present with associated disabilities. Cochlear implant (CI) is an alternative treatment that provides consistent access to environmental and speech sounds, which results in significant benefits regarding quality of life and auditory and language development.

Objectives To study the auditory and communicative performance of individuals with CI and delayed neuropsychomotor development after a minimum of five years using the device.

Methods A total of eight patients were included in the study. We collected the multidisciplinary clinical records of participants, as well as the answers for the questionnaires applied remotely, which included the Children with Cochlear Implants: Parental Perspectives (CCIPP), International Classification of Functioning, Disability and Health: Children and Youth Version (ICF-CY), and the Gross Motor Function Classification System (GMFCS).

Results We found that throughout the years of CI use, the auditory threshold means improved significantly in all tested frequencies, as did the speech detection threshold and the language and hearing results. Regarding parental perception, parents evaluated aspects related to their children's social relations to be positive, and had worse perceptions regarding aspects related to their education.

Conclusion We observed a progression in the participants' auditory and language skills throughout the years of CI use; even in the presence of other associated disabilities. Future multicentric studies with larger samples are needed to further the advancement of rehabilitation in patients with other associated disabilities.

Keywords
- hearing loss
- cochlear implantation
- cerebral palsy
- rehabilitation

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Introduction

Severe to profound bilateral sensorineural hearing loss is a comorbidity that affects more than 5% of individuals ($\sim$ 466 million people) globally.[1] Cochlear implant (CI) is an alternative treatment for these individuals, and it enables the development of auditory, language, communication, cognitive, and social skills.

Between 15% and 30% of individuals with bilateral prelingual sensorineural hearing loss present with associated disabilities, which include delayed motor, cognitive, and emotional development, usually diagnosed by an interdisciplinary team.[2,3] Previous studies [2] have reported that 60 out of 398 cases present with other associated deficiencies. Moreover, 8.33% of these cases have neuropsychomotor development delay (NPMD)/cerebral palsy. For these cases, the use of the CI provides consistent access to environmental and speech sounds, which results in significant benefits regarding quality of life and auditory and language development. However, these are usually inferior compared to patients without other associated disabilities.[3]

In another study,[2] with a sample of 96 individuals with CIs, 33% were identified with other associated disabilities, including developmental delay, NPMD/cerebral palsy, visual impairment, autism, and attention deficit disorder. After 12 months of use of the device, 52% of these individuals scored 5 on the Categories of Auditory Performance (CAP) index, with a median CAP score of 4 for the whole sample, while 96% of the individuals without other disabilities had CAP scores between 5 and 6.

Considering the findings in the literature, the clinical experience, and the degree of technical specialization of the CI services and team, the criteria for the indications of a CI, especially for individuals with NPMD, were established. One of the variables that have great importance for this indication is the individual's global rehabilitation with an interdisciplinary team associated with motivation from the family and the social environment, especially the school, as well as the development of a targeted and specific therapeutic plan for the individual.

Another important aspect in these individuals is the variability of clinical manifestations as regards to motor, sensory, perceptual, and cognitive characteristics, which results in limited activities that restrict learning and the development of sensory-perceptual and cognitive experiences, thereby affecting the individual's quality of life.[4–6]

Thus, the use of CI associated with the therapeutic process of specialized hearing habilitation and rehabilitation and specialized global sensorial rehabilitation results in better quality of life due to the possibility of developing auditory and oral language skills, sensorial integration, focusing on communication and higher levels of independence, social integration, and selfconfidence.[7–9]

In this sense, an increasing number of children with multiple disabilities undergo CI surgery. However, the available literature [2–4,7–9] that assesses the impact and benefit of the CI in the long term is limited. Therefore, it is extremely important to carry out new longitudinal studies in this field to better understand the resources, processes, and impact of CI on this population.

Thus, the present study described the auditory and communicative performance of individuals with CI and NPMD after at least five years of use of the device. In addition, the quality of life, motor development, functional performance, functionality in the activities of daily living, social participation, and the influence of environmental factors were described, as well as their relationship and progress with the CI.

Materials and Methods

The present was a retrospective and cross-sectional study approved by the institutional Ethics in Research Committee (under CAAE: 29801620.5.0000.5441). All subjects agreed to participate in the study by signing an informed consent form.

Casuistry

The eligibility criteria were as follows:

- Patients with symmetrical, sensorineural, profound, bilateral hearing loss (average thresholds in the frequencies of 500 Hz, 1,000 Hz, 2,000 Hz, and 4000 Hz for the classification of the degree of hearing loss);
- Subjects with at least five years of effective CI use;
- Patients diagnosed with NPMD without cognitive impairment;
- And those undergoing hearing habilitation and rehabilitation or therapeutic discharge.

Individuals with peripheral and/or central auditory pathway malformation were excluded.

A total of eight patients diagnosed with bilateral profound sensorineural hearing loss with NPMD and CI were included. The average age was 14 years, and most participants were male (75%, n = 6), attended a regular school (62.5%, n = 5), and were of a low socioeconomic level (50%, n = 4). Overall, two participants were diagnosed with microcephaly and autism spectrum disorder, respectively.

Of the 8 participants, 5 (62.5%) had unilateral CI, and 3 (37.5%) had bilateral sequential CI. The most recent CI had been implanted 20 months previously, while the least recent had been implanted for 61 months. The second CI had been activated for more than 11 years since they had the second surgery. On average, the patients had been using CIs for 11 years since the first CI surgery.

Currently, 5 (62.5%) patients are still undergoing speech therapy, with an average of 2 sessions per week. In total, 2 (25%) patients were discharged from speech therapy, and 1 (12.5%) could not undergo it because of social and financial limitations.

Procedures

The multidisciplinary clinical records of each participant were collected, including social, psychological, and medical data, as well as the results of a speech perception and language performance. The information collected included the following:

a) Free-field audiometry: thresholds of 500 Hz, 1,000 Hz, 2,000 Hz, and 4,000 Hz with the CI, using the warble stimulus and speaker at a distance of 1 m from the patient, at 90° azimuth, and the speech detection threshold (SDT) in the same conditions.
b) Score on the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS);[10]
c) Results of the Meaningful Use of Speech Scale (MUSS)[11] questionnaire;
d) Score on the CAP;[12]
e) Category of Language (Score on the Expressive Language Category, used in internal protocols, such as: category 1 - child does not speak and may present indistinct vocalization; category 2 - child speaks only few words; category 3 - child makes simple sentences; category 4 - child makes complex sentences; category 5 - child is fluent in oral language); [13]
f) Score on the Glendonald Auditory Screening Procedure (GASP), a procedure to assess speech perception in profoundly hearing-impaired children from the age of 5 years.[14] For the present research, only the last 3 tests (4, 5, and 6) were considered, and their analyses were performed using percentages;
g) List of disyllabic words to assess the perception of speech sounds;[15]
h) Score on the sentence recognition test in silence and the signal-to-noise ratio (SNR) of $+10$ dB;[16]
i) Psychological assessment: a survey of data from the clinical records was performed, with a global assessment of the patient, especially in the last evaluation;
j) Socioeconomic evaluation [17]: a survey of the clinical records referring to the institutional protocols, which consists of the users' living conditions, the need for implementation and services, average family income, and parents academic degree.

In addition to these data, the researcher applied the following questionnaires remotely (via telephone call):

- Children with Cochlear Implants: Parental Perspectives (CCIPP);[18,19]
- International Classification of Functioning, Disability and Health: Children and Youth Version– (ICF-CY);[20] and
- Gross Motor Function Classification System (GMFCS).[21,22]

Statistical Analysis

Descriptive and inferential statistics were used. The results were presented in tables and graphs. The data included auditory and language performance after three months, six months, one and two years of CI use, as well as the last return to the service.

The Shapiro-Wilk normality test was used. The Friedman test and the Dunn post-hoc test were applied to check for significant differences in the performance of patients on free-field audiometry at each frequency at different periods. They were also applied for the IT-MAIS, MUSS, GASP, and recognition of dissyllabic words tests, the CAP, and Expressive Language Categories (ELCs) over time of use of the device.[23]

The Spearman correlation coefficient was applied to verify the correlation between the results obtained in the following evaluations: CCIPP and the Activity and Participation and Environmental Factors domains of the ICF-CY with the results of the free-field audiometry obtained in the last return, IT-MAIS, MUSS, auditory recognition of words and phonemes, recognition of sentences in silence and noise, GASP, CAP and GMFCS, and the subscales of the Children with Cochlear Implant Category of Language over time of use of the device (data from the last return), as well as age at activation of the first CI, speech-language therapy, actual age, time of use of the CI, socioeconomic level, and other associated disabilities.

The significance level adopted was of 0.05.

Results

In the present study, the use of CI over the years improved the mean hearing thresholds in all tested frequencies (0.5 Hz, 1 Hz, 2 Hz, and 4 kHz), as well as the voice/speech detection threshold (SDT) obtained on free-field audiometry. When the threshold was compared at different intervals of use of the device, we found a significant difference between the mean thresholds obtained after three months use and those obtained at the last appointment for 500 Hz, 1,000 Hz, and 2,000 Hz and the SDT (Friedman test: $p < 0.001$; Dunn post-hoc test: $p < 0.05$). No significant difference was found regarding the hearing thresholds obtained at six months and at one and two years of CI use, compared with the last appointment (►**Fig. 1**).

The same results were found in the evaluations of the auditory and language skills throughout the years of CI use performed in the follow-up (►**Table 1**). On average, patients with NPMD and CI achieved progressive improvement and better results compared with the performance at the moment of activation and the performance at the last return in relation to the auditory skills assessed by the IT-MAIS, word and phoneme recognition, and sentence recognition in silence and in noise, as well as in relation to language skills (MUSS), with the difference being statistically significant over time for this group (Friedman test: $p < 0.001$). The Dunn post-hoc test ($p < 0.05$) showed that, in the IT-MAIS questionnaire, there was a significant difference regarding the results obtained at three months, one year, and two years of CI use and at the last return. As for the MUSS questionnaire, the difference was significant in terms of the results obtained at three and six months and two years of use of the device use and the last return.

The Dunn post-hoc did not signal in which interval of use the difference was statistically significant for word and phoneme recognition tests due to the sample size. However, a statistically significant difference was shown by the Friedman Test in these two evaluations.

We observed that 62,5% ($n = 5$) of the participants who had been using the device for two years reached a maximum score in the vowel discrimination test and vowel extension test of the GASP (►**Fig. 2**). Analyzing the results for auditory recognition in close-set and listening comprehension tests,

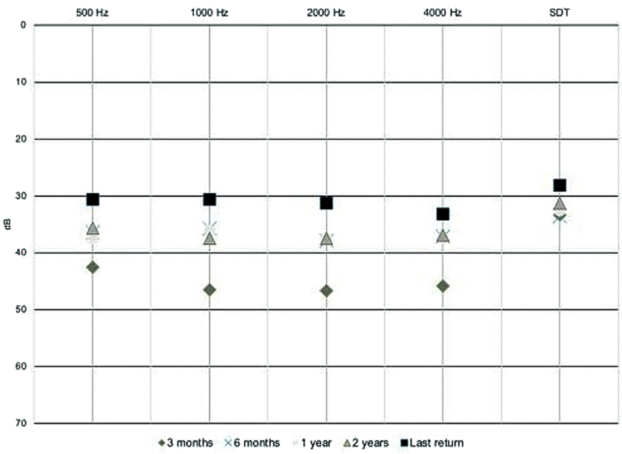


Fig. 1 Comparison of hearing thresholds (mean frequency) in a free field after three months, six months, and one and two years of CI use, and the results at the last appointment ($n = 8$).

we observed that, after two years of CI use and in the last appointment, some participants reached the highest score (100%) and others maintained a stable performance. In the statistical analysis, we found a significant difference (Friedman test: $p < 0.001$) in the results of the three tests over time, but the Dunn post-hoc test ($p < 0.05$) did not detect for which periods the difference was statistically significant due to the size of the analyzed sample.

►**Fig. 3** shows the results of the patients, medians, and 95% confidence intervals obtained in the CAP and ELC. Some patients reached categories 6 or 5 on the CAP in their last appointment. However, other participants only reached category 1 on the CAP and ELC. A statistically significant difference was found between the performance after three months of use and that at the last appointment (Friedman test: $p < 0.001$; Dunn post-hoc test: $p < 0.05$) in the two assessments. This indicates that there was a significant improvement in the language and hearing results throughout the years of CI use among these participants.

Concerning the different levels of skills and limitations in gross motor function measured by the GMFCS, the following results were obtained: 6 (75%) participants were classified as level I (walks without limitations), 1 (12.5%), level II (walks with limitations), and 1 (12.5%), as level III (walks using a manual mobility device). Regarding the parental perception of quality of life, the aspects related to the children's social relationships were reported to be positive, whereas the perceptions were worse for the aspects related to their education (►**Fig. 4**).

Regarding the body functions domain of the ICF-CY, 15 items were considered in the evaluation of the participants in the present study (►**Table 2**). The items that were most often qualified with a disability (mild, moderate, severe, or total) were as follows: b117—intellectual functions (87.5%), b164—higher cognitive functions (75%), b235—vestibular function (75%), and b765—involuntary movements.

In the activities and participation domain of the ICF-CY, 26 items were qualified. We identified that the items with which the participants had more difficulty (mild, moderate, severe, or total) were as follows: d140—learning to read (75%), d145—learning to write (100%), d150—learning to calculate (87.5%), d310—communicating with - receiving - spoken messages (75%), d330—speaking (62.5%), d440—fine hand use (62.5%), d450—walking (75%), d720—complex interpersonal interactions (75%), and d820—school education (75%) (►**Table 2**).

For the environmental factors (►**Table 3**), the use of CI (e125—products and technology for communication), family (e310—immediate family), and access to health services (e580—health services, systems, and policies) were considered environmental facilitators for most of the participants. By contrast, friends (e320—friends) were qualified neither as a barrier nor as a facilitator, and background noise (e250—sound) was an important environmental barrier in the present study.

The Spearman correlation test was applied to verify whether the results obtained in the CCIPP, ICF-CY, and GMFCS correlated with the results obtained in the assessments performed in the service during the follow-up (►**Table 4**). No significant correlations were found between the GMFCS, and the body functions, and environmental factors domains of the ICF-CY with the selected variables. These results were not included in the table. Variables such as age at activation of the first CI, speech therapy, IT-MAIS, and recognition of sentences in silence and noise did not correlate with the evaluations. Therefore, the results were deleted from the table. Most of the correlations found were considered moderate and strong, as shown in ►**Table 4**.

Discussion

Due to the variability in clinical findings and comorbidities involved, NPMD encompasses different cases. Therefore, depending on the case, after a thorough and safe evaluation of the diagnosis, it is possible to define the best treatment alternative with an interdisciplinary team.

We found that 75% ($n = 6$) of the participants in the present study received the first implant before 42 months of age. This is, therefore, a positive aspect considering the findings in the literature for auditory and language development given the period of neuronal plasticity.[24]

Cejas et al.[25] found that, compared with CI use in children without disabilities other than hearing loss, CI use in children with multiple disabilities was beneficial, especially if the device was used early, considering other variables such as auditory-verbal therapy, effective use, and a family who stimulate their child daily, but at a different time that when the cochlear implant was effectively used.

It is important to emphasize the interdisciplinary criteria for the indication of a CI. Two cases were implanted later, at 60 and 61 months, due to diagnostic delays. The first participant had auditory neuropathy spectrum disorder, which initially showed improvement with conventional amplification. It then progressed to hearing loss and limited speech perception performance. The second participant had a late surgery as he was diagnosed with microcephaly; however, there was no cognitive delay in the psychological assessment.

Table 1 Results of the evaluations after three months, six months, one year, two years, and at the last appointment ($n = 8$)

Evaluation		Average	Minimum	Maximum	Standard deviation	*p*-value
	3 months	66.55	52.25	85.25	12.21	
	6 months	82.92	57.50	92.50	12.79	
Infant-Toddler Meaningful Auditory Integration Scale	1 year	90.71	77.50	100.00	10.18	0.0002
	2 years	90.00	60.00	100.00	14.65	
	Last appointment	92.86	50.00	100.00	18.90	
	3 months	10.50	2.50	17.50	5.42	
	6 months	20.00	7.50	32.50	11.40	
Meaningful Use of Speech Scale	1 year	30.00	10.00	72.50	22.36	< 0.001
	2 years	46.43	20.00	85.00	26.41	
	Last appointment	57.50	0.00	100.00	50.58	
	3 months	0.00	0.00	0.00	0.00	
	6 months	0.00	0.00	0.00	0.00	
Auditory recognition of phonemes (%)	1 year	0.00	0.00	0.00	0.00	0.0014
	2 years	21.56	0.00	90.00	39.98	
	Last appointment	46.53	0.00	100.00	44.88	
	3 months	0.00	0.00	0.00	0.00	
	6 months	0.00	0.00	0.00	0.00	
Auditory recognition of words (%)	1 year	0.00	0.00	0.00	0.00	0.0014
	2 years	14.38	0.00	75.00	28.21	
	Last appointment	42.50	0.00	100.00	42.17	
Sentences in silence	Last appointment	25.00	0.00	100.00	46.29	–
Sentences in noise with signal-to-noise ratio +10 dB	Last appointment	23.38	0.00	100.00	43.42	–

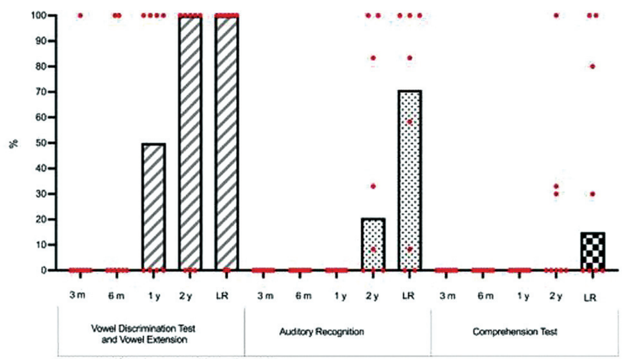
As for the audibility after the CI, the results showed a statistically significant progressive evolution in the auditory thresholds in the free field at all the tested frequencies (0.5 Hz, 1 Hz, 2 Hz, and 4 kHz), and for the voice/speech detection threshold, both after 3 months of CI use and at the last appointment. When the 3 months' use was compared with the last assessment, the auditory thresholds in the free field showed a statistically significant improvement at all tested frequencies (0.5 Hz, 1 Hz, 2 Hz, and 4 kHz) and for the SDT.

These findings prove that, with adequate programming, the CI can promote consistent access to environmental and speech sounds, considering the improvement in thresholds observed. The improvement in the thresholds presented after three months of CI use and the last appointment illustrates the importance of the fine adjustments in the programming performed at each follow-up. This enables the detection of low-intensity sounds and the perception of spectral details, which are prerequisites for the development of auditory, language, and communication skills.[26,27]

Another important correlation was observed regarding the speech perception protocols at each follow-up, in which there was a progressive improvement in hearing and language skills with an increase in the duration of CI use. This is consistent with the clinical reasoning that CI provides audibility to environmental and speech sounds, maintaining the perception of the different intensities and frequencies. Thus, respecting all the variables involved, it is possible to obtain a significant improvement in speech perception.

This clinical reasoning can be confirmed by analyzing the results obtained in the speech perception tests. For the IT-MAIS, the difference was significant when the results obtained at three months of CI use were compared, with those at one year, two years, and at the last appointment. These results corroborate with those observed in the study by Alvarenga et al.,[28] in which the score on the IT-MAIS increased significantly according to the duration of the CI use.

In the present study, we chose to analyze the GASP scores on tests 4, 5, and 6, as these represent the most advanced hearing skills, which are gradually developed. Most of the



Subtitle: m: month; y: year; LR: last return

Fig. 2 Median and individual results in vowel discrimination and vowel extension discrimination, auditory recognition, and auditory comprehension on the GASP at three months, six months, one year, two years, and at the last appointment ($n = 8$).

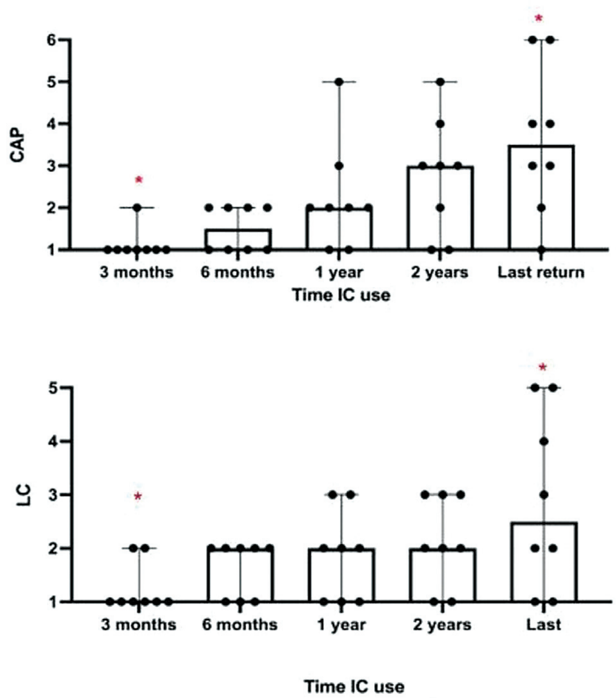
participants were able to reach the maximum score in the vowel discrimination test after two years of activation. This illustrates the ability to recognize extension patterns and differentiate segmental and suprasegmental speech characteristics, which is an important skill in the acquisition of language, phonetic, and phonological repertoire for the development of more advanced auditory skills.

However, a variety of findings regarding the results of the tests of recognition and auditory comprehension was observed, since some patients reached the maximum score, and others were unable to perform the tests even at the last appointment. Silva et al.[29] found that only children after 11 months of use of cochlear implant were able to take test 5 of the GASP. Nonetheless, this did not occur in most children. They[29] also found that there was a greater variability in results in the first year of CI use and after one year of hearing age. The evolution curve of the responses was increasingly evident and more regular; this was also observed in the present study.

This progressive improvement was noted with the results of word recognition tests that started after two years of use of the device, and sentence recognition, which started at the last appointment for some of the participants. The results of the speech perception tests are consistent with those obtained following the evolution and progressive improvement of the auditory performance categories.

The same thing can be observed regarding the language measured by the MUSS and ELCs. For the MUSS, the difference was significant regarding the results obtained at three and six months of CI use compared with those after two years of use and at the last appointment. For the ELCs, there was a progressive improvement in the auditory performance categories. Thus, we found that these participants developed advanced auditory skills, which are called an open set. Thus, from the variability of the cases, we can state there was a significant improvement in language and auditory skills throughout the years of CI use of the CI, which is in agreement with other studies[30] performed in children with hearing loss who presented with NPMD.

Considering the variability of these findings, several factors can influence CI use, such as effective use, maintenance of the device, family background, sensory deprivation time, speech therapy, among other factors with a long-term impact. Thus, it should be noted that the participant who did not show improvement in hearing and language skills (CAP 1 and ELCs 1) did not effectively use the speech processor in the first year. Three months after activation, the speech processor broke, and the patient presented at the six-month appointment after activation without the processor because



Subtitle: Statistically significant difference (*) ($p < 0.001$)

Fig. 3 Patients performance, median, and 95% confidence interval obtained on the CAP and ELC, at three months, six months, one year, and two years of CI use, and the results at the last appointment ($n = 8$).

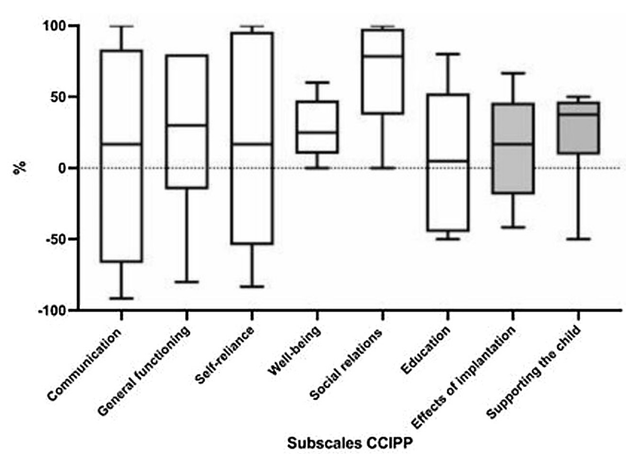


Fig. 4 Means, medians, and the minimum and maximum values obtained regarding parental perceptions in the subscales of the CCIPP represented by box plots. The higher the average, the more positive the parental perceptions.

Table 2 Distribution of the frequencies of the categories of the ICF-CY for the body functions and activities and participation domains ($n = 8$)

		Some degree of difficulty			
Body functions	**No disability n (%)**	**Mild disability n (%)**	**Moderate disability n (%)**	**Severe disability n (%)**	**Total disability n (%)**
b110–State of consciousness	7 (87.50)	1 (12.50)	–	–	–
b114–Orientation functions	6 (75.00)	–	1 (12.50)	–	1 (12.50)
b117–Intellectual functions	1 (12.50)	3 (37.50)	1 (12.50)	2 (25.00)	1 (12.50)
b140–Attention functions	4 (50.00)	1 (12.50)	2 (25.00)	1 (12.50)	–
b144–Memory functions	4 (50.00)	1 (12.50)	2 (25.00)	1 (12.50)	–
b152–Emotional functions	7 (87.50)	1 (12.50)	–	–	–
b164–Higher-level cognitive functions	2 (25.00)	2 (25.00)	2 (25.00)	2 (25.00)	–
b167–Mental function of language	6 (75.00)	–	2 (25.00)	–	–
b210–Seeing functions	5 (62.50)	1 (12.50)	1 (12.50)	1 (12.50)	–
b230–Hearing functions	–	–	–	–	8 (100.00)
b235–Vestibular functions	2 (25.00)	4 (50.00)	1 (12.50)	–	1 (12.50)
b710–Mobility of joint functions	4 (50.00)	2 (25.00)	2 (25.00)	–	–
b730–Muscle power functions	6 (75.00)	1 (12.50)	1 (12.50)	–	–
b735–Muscle tone functions	4 (50.00)	3 (37.50)	–	–	1 (12.50)
b765–Involuntary movement functions	2 (25.00)	4 (50.00)	–	1 (12.50)	1 (12.50)
		Some degree of difficulty			
Activities and participation	**No difficulty n (%)**	**Slight difficulties n (%)**	**Moderate difficulties n (%)**	**Severe difficulties n (%)**	**Total difficulties n (%)**
d110–Watching	4 (50.00)	–	1 (12.50)	1 (12.50)	2 (25.00)
d115–Listening	4 (50.00)	1 (12.50)	1 (12.50)	1 (12.50)	1 (12.50)
d140–Learning to read	2 (25.00)	–	2 (25.00)	1 (12.50)	3 (37.50)
d145–Learning to write	–	2 (25.00)	1 (12.50)	2 (25.00)	3 (37.50)
d150–Learning to calculate	1 (12.50)	1 (12.50)	1 (12.50)	1 (12.50)	4 (50.00)
d175–Solving problems	4 (50.00)	1 (12.50)	2 (25.00)	–	1 (12.50)
d210–Undertaking a single task	6 (75.00)	1 (12.50)	1 (12.50)	–	–
d220–Undertaking multiple tasks	6 (75.00)	–	–	–	2 (25.00)
d310–Communicating with - receiving - spoken messages	2 (25.00)	1 (12.50)	1 (12.50)	2 (25.00)	2 (25.00)

(*Continued*)

Table 2 (*Continued*)

		Some degree of difficulty			
Body functions	No disability n (%)	Mild disability n (%)	Moderate disability n (%)	Severe disability n (%)	Total disability n (%)
d315–Communicating with - receiving - non-verbal messages	5 (62.50)	1 (12.50)	2 (25.00)	–	–
d330–Speaking	3 (37.50)	1 (12.50)	2 (25.00)	2 (25.00)	–
d335–Producing non-verbal messages	6 (75.00)	1 (12.50)	1 (12.50)	1 (12.50)	–
d350–Conversation	5 (62.50)	1 (12.50)	–	1 (12.50)	1 (12.50)
d440–Fine hand use	3 (37.50)	1 (12.50)	3 (37.50)	1 (12.50)	–
d450–Walking	2 (25.00)	4 (50.00)	–	1 (12.50)	1 (12.50)
d465–Moving around using equipment	5 (62.50)	1 (12.50)	–	–	2 (25.00)
d510–Washing oneself	6 (75.00)	–	1 (12.50)	–	1 (12.50)
d520–Caring for body parts	4 (50.00)		2 (25.00)	1 (12.50)	1 (12.50)
d540–Dressing	5 (62.50)	2 (25.00)	–	1 (12.50)	–
d550–Eating	6 (75.00)	–	1 (12.50)	1 (12.50)	–
d560–Drinking	7 (87.50)	–	–	1 (12.50)	–
d710–Basic interpersonal interactions	6 (75.00)	–	–	2 (25.00)	–
d720–Complex interpersonal interactions	2 (25.00)	2 (25.00)	1 (12.50)	3 (37.50)	–
d760–Family relationships	7 (87.50)	–	–	1 (12.50)	–
d810–Informal education	6 (75.00)	–	1 (12.50)	–	1 (12.50)
d820–School education	2 (25.00)	–	2 (25.00)	2 (25.00)	2 (25.00)
d910–Community life	6 (75.00)	–	–	–	1 (12.50)

Abbreviation: ICF-CY, International Classification of Functioning, Disability, and Health: for Children and Youth Version.

it had been stolen. Therefore, the effective evaluations of CI use by the patient only occurred after one year of the activation. In addition, this same participant was also diagnosed with autism spectrum disorder. The literature[2] points out greater difficulties in the development of auditory, language, and communication skills in such cases. Steven et al.[31] reported that, when there is mild cognitive impairment, the results are positive in terms of the acquisition of auditory and language skills, but, in cases of more severe impairment, the results are worse.

The other patient with limited auditory and language development had microcephaly. This condition is responsible

Table 3 Distribution of the frequency of the categories of the ICF-CY as a barrier or facilitator for the environmental factors domain ($n = 8$)

	Neither a barrier nor a facilitator n (%)	Barrier n (%)	Facilitator n (%)
e125–Products and technology for communication	2 (25.00)	1 (12.50)	5 (62.50)
e250–Sound	2 (25.00)	3 (37.50)	3 (37.50)
e310–Immediate family	3 (37.50)	1 (12.50)	4 (50.00)
e320–Friends	5 (62.50)	–	3 (37.50)
e580–Health services, systems, and policies	2 (25.00)	1 (12.50)	5 (62.50)

Abbreviation: ICF-CY, International Classification of Functioning, Disability, and Health: for Children and Youth Version.

Table 4 Spearman correlation regarding the subscales of the CCIPP questionnaire, ICF-CY domains, GMFCS, MUSS, and variables pertaining to the patients ($n = 8$)

	Communication (CCIPP) *p*; *rho*	General functionality (CCIPP) *p*; *rho*	Self-confidence (CCIPP) *p*; *rho*	Wellbeing and happiness (CCIPP) *p*; *rho*	Social relationships (CCIPP) *p*; *rho*	Education (CCIPP) *p*; *rho*	Effects of CI (CCIPP) *p*; *rho*	Child support (CCIPP) *p*; *rho*	Activities and participation *p*; *rho*
Current age of the participant	–	–	–	–	–	–	–	0.0333; 0.7478	–
Use of CI in months (first surgery/activation)	–	–	–	0.0066; - 0.8795	–	–	0.0167; - 0.8263	–	0.0179; 0.8505
Thresholds for warble stimulus in free-field audiometry at the last appointment (dB)	–	–	–	–	–	0.0298; -0.7690	–	–	–
Thresholds for speech stimulus in free-field audiometry at the last appointment (dB)	–	–	–	–	0.0369; - 0.7708	–	–	–	–
MUSS	0.0232; 0.8025	0.0071; 0.9000	–	–	–	0.0402; 0.7531	–	–	–
Auditory performance category	0.0103; 0.8589	0.0202; 0.8199	0.0157; 0.8221	–	–	0.0190; 0.8160	0.0095; 0.6587	0.0214; 0.5758	0.0238; -0.8399
Language category	0.0048; 0.9141	0.0006; 0.9690	–	–	–	0.0123; 0.8528	0.0532; 0.8607	–	–
Auditory recognition of phonemes (Delgado list)	0.0345; 0.7655	0.0039; 0.9250	–	–	–	0.0152; 0.8396	–	–	–
Auditory recognition of words (Delgado list)	0.0101 0.8643	0.0021; 0.9500	–	–	–	0.0080; 0.8766	–	–	–
Socioeconomic level	–	–	–	–	–	–	–	0.0429; -0.7329	–
Other associated disabilities	–	0.036; - 0.775	–	–	–	–	–	–	–

Abbreviation: CCIPP, Children with Cochlear Implants: Parental Perspectives; CI, cochlear implant; GMFCS, Gross Motor Function Classification System; ICF-CY, International Classification of Functioning, Disability, and Health: for Children and Youth Version; MUSS, Meaningful Use of Speech Scales.

for 65% of children diagnosed with intellectual disabilities or delayed neurological development. In most cases, these children require an interdisciplinary team for rehabilitation as well as special education in school.[32] This participant communicated using Brazilian Sign Language (Língua Brasileira de Sinais, LIBRAS, in Portuguese) and attends a special school in which he learns this form of communication. In addition, another important variable, considering the importance of the period of brain plasticity indicated in the literature, was the age at implantation, which was of 61 months.[24]

Therefore, it should be noted that even though there was variability in the clinical findings in the present study, it is very important to highlight that the best rehabilitation alternatives have to be discussed and selected with the family at each stage of diagnosis and treatment. Reevaluations o the therapeutic plan are fundamental for the longitudinal monitoring of these cases, first involving the patient-centered approach, defining what is best for each case depending on the responses presented to each therapeutic option performed.

Therefore, the family must play an important role in choosing the treatment. Regarding the quality of life of the patient and their family, in terms of functionality, important findings were observed.

When asked about quality of life, their children's social relationships were the most valued aspect, whereas aspects related to education had the worst evaluations. This complaint changed according to the average age of the participants, which was 14 years. In this age group, parents have already shaped their expectations regarding their children's speech. However, they have more expectations related to their children's school activities, such as reading and writing. On the ICF-CY, parents also pointed out the issue of education as one of their children's greatest difficulties: learning to read (75%, n = 6), learning to write (100%, n = 8), learning to calculate (87.5%, n = 7), and school education (75%,n = 6).

Regarding the body functions domain of the ICF-CY, reinforcing the NPMD in these participants, we observed that the items most frequently associated with disabilities were intellectual functions (87.5%, n = 7) and vestibular function (75%, n = 6), the latter being considered within gait changes.

In the activities and participation domain, in addition to the items related to education, the parents described as the greatest difficulties the reception of verbalmessages (75%, n = 6), justified by the hearing loss; speech (62.5%, n = 5), which may be related to expectations regarding the CI; fine use of hands (62.5%, n = 5), which justified the difficulty in writing, being a remnant of NPMD; and walking (75%, n = 6). In this item, despite most of the participants (75%, n = 6) being classified in levelI in the system of classification of gross motor function, the parents scored the difficulty in walking related to balance and gait.

In relation to environmental factors, CI use (e125 –products and technologies for communication) was classified as a facilitator for most parents (62.5%, n = 5). However, 12.5%, (n = 1) classified it as a barrier; this is justified by the difficulty in obtaining the CI and access to specialized quality service, often requiring these families to move from their cities of origin. For 50%, (n = 4) of the interviewed parents, the family (e310 – immediate family) was considered a facilitator, demonstrating the importance of having strong support networks, especially with children with disabilities. Background noise (e250 –sound) was classified as an important environmental barrier, as itmakes it difficult for these users to communicate and further increases their auditory effort.

In the correlation test (►**Table 4**), moderate and strong correlations regarding the instruments applied were observed. The better the results on the MUSS, CAP, LCs, and list of words to assess the perception of speech sounds, the better the parents evaluated the communication, general functionality, and education of their children. In addition, the better the results on the CAP, the better the parents classified the effects of the CI, self-confidence, support for their children, and their activities and participation. The ELCs correlated with better perceptions related to the effects of the CI. Byun et al.[33] also observed that these children may have benefits in auditory perception, speech production, and language capacity equivalent to those of their peers whose only disability is hearing loss when surgery is performed within a critical period.

In this regard, a positive factor observed in the present study was that 62.5% (n = 5) of the patients are currently undergoing speech therapy, and 25% (n = 2) have already been discharged. Speech language therapy is one of the most important variables involved in the prognosis of prelingual children with CI, for auditory, language, and communication development. If the intervention occurs in the first years of life, the auditory perception improves. Consequently, the initial linguistic activities, formation of statements, phonemic diversity, and phonetic patterns improve, and, therefore, the better is the speech development.[34,35]

Lower thresholds in the free field correlated directly with the parents' better perception of education; thus, better thresholds possibly mean less hearing effort, especially in the school environment. The same is true for the voice-detection threshold: better thresholds are related to better perceptions regarding the social relationships of the participants.

Some negative correlations were observed, such as, the longer the use of the CI, the worse the parents' perception of its effect and the child's well-being and happiness. It is believed that this result may be related to the expectations that parents have with the use of the CI and how they change in the long term. The parents become more realistic about the prognosis and alternatives for rehabilitation in the long term.

Another negative correlation was that the lower the socioeconomic level, the better the parents' perception of child support, which differs from the literature[36] reports that children who needed more support were those with lower socioeconomic status.

Studies[37,38] have shown that the CI also helps in the development of cognitive skills (general development). It has been observed that children with hearing loss show improvements in their understanding, concentration, sequential processing, and working memory, with almost normal values. The

CI not only restores auditory function, but also plays an important role in cognitive development.[37,38]

Due to the limitation in the sample size of the present study, new studies with larger and multicentric samples are required, because the results of the present sample varied according to the commitment of each patient regarding CI use. Therefore, new studies are needed to make new advances in hearing rehabilitation in patients with other associated disabilities.

Conclusion

With the present study, we could observe a progression in auditory and language skills throughout the years of CI use: that is, even with other associated disabilities, the patients were able to benefit from the use of the device in the long term. Advanced auditory skills, as well as improvement and possibility of communicative independence in activities of daily living, improved over time, which directly interferes with the quality of life and social relationships of the patient.

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Conflict of Interests
The authors have no conflict of interests to declare.

References

1 United Nations News [Internet]. WHO warns that hearing loss could affect more than 900 million by 2050; 2020 Mar 3 [cited 2020 Nov 6]. Available from: https://news.un.org/pt/story/2020/03/1705931

2 Daneshi A, Hassanzadeh S. Cochlear implantation in prelingually deaf persons with additional disability. J Laryngol Otol 2007;121(07):635–638. https://pubmed.ncbi.nlm.nih.gov/17147840/ cited 2020 Nov 6 [Internet]

3 Birman CS, Elliott EJ, Gibson WPR. Pediatric cochlear implants: additional disabilities prevalence, risk factors, and effect on language outcomes. Otol Neurotol 2012;33(08):1347–1352. https://pubmed.ncbi.nlm.nih.gov/22975903/ cited 2020 Nov 6 [Internet]

4 Edwards LC. Children with cochlear implants and complex needs: a review of outcome research and psychological practice. J Deaf Stud Deaf Educ 2007;12(03):258–268. https://pubmed.ncbi.nlm.nih.gov/17493953/ cited 2020 May 26 [Internet]

5 Rosenbaum P, Paneth N, Leviton A, et al. A report: the definition and classification of cerebral palsy April 2006. Dev Med Child Neurol Suppl 2007;109:8–14. https://pubmed.ncbi.nlm.nih.gov/17370477/ cited 2020 May 26 [Internet]

6 Brasil. Ministério da Saúde. Secretaria de Atenção à Saúde, Departamento de Ações Programáticas Estratégicas. Diretrizes de atenção à pessoa com paralisia cerebral. Ministério da Saúde. 1st ed. Brasília2013 [cited 2020 Nov 7]. Available from: http://bvsms.saude.gov.br/bvs/publicacoes/diretrizes_atencao_paralisia_cerebral.pdf

7 Bacciu A, Pasanisi E, Vincenti V, et al. Cochlear implantation in children with cerebral palsy. A preliminary report. Int J Pediatr Otorhinolaryngol 2009;73(05):717–721. https://pubmed.ncbi.nlm.nih.gov/19201488/ cited 2020 Nov 6 [Internet]

8 Lachowska M, Rózycka J, Łukaszewicz Z, Konecka A, Niemczyk K. [Auditory skills in multi-handicapped children with cochlear implants]. Otolaryngol Pol 2010;64(07):22–26. https://pubmed.ncbi.nlm.nih.gov/21171306/ cited 2020 Nov 6 [Internet]

9 Berrettini S, Forli F, Genovese E, et al. Cochlear implantation in deaf children with associated disabilities: challenges and outcomes. Int J Audiol 2008;47(04):199–208 cited 2020 Nov 6. Doi: 10.1080/14992020701870197 [Internet]

10 Castiquini EAT, Bevilacqua MC. Scale of significant auditory integration: adapted procedure for the assessment of speech perception. Rev Soc Bras Fonoaudiol 2000;4(06):51–60. Available from https://repositorio.usp.br/item/001110055 cited 2020 Nov 6 [Internet]

11 Nascimento LT. Uma proposta de avaliação da linguagem oral [Monografia]. Bauru: Especialização em Audiologia, Universidade de São Paulo; 1997

12 Geers A. Techniques for Assessing Auditory Speech Perception and Lipreading Enhancement in Young Deaf Children. Volta Review 1994;96(05):85–96. Available from https://eric.ed.gov/?id=EJ505099 cited 2020 Nov 6 [Internet]

13 Bevilacqua MC, Delgado EM, Moret AL. Clinical case studies of children from the Educational Center for the Hearing Impaired (CEDAU), Proceedings of the International Audiology Meeting. ; March 30th to April 2nd; Bauru, Brazil1996

14 Bevilacqua MC, Tech EA. A method for assessing speech perception in deaf children—more than five years. Marchesan IQ, Zorzi JL, Gomes ICDeditors. Topics in speech therapy. São Paulo: Lovise; 1996:411–433

15 Delgado EMC, Bevilacqua MC. List of words to evaluated the speech perception in deaf children. Pro Fono 1999;11(01):59–64. Available from https://repositorio.usp.br/item/001224091 cited 2020 Nov 6 [Internet]

16 Oliveira ST. Assessment of speech perception using everyday sentences [dissertation]. São Paulo: PUC, 1992

17 Graciano MIG. Socioeconomic study: a technical-operative instrument. Proceedings of the Congenital Labiopalatine Anomalies Course. ; Bauru, University of São Paulo, Hospital for the Rehabilitation of Craniofacial Anomalies, São Paulo; 2013

18 Archbold SM, Lutman ME, Gregory S, O'Neill C, Nikolopoulos TP. Parents and their deaf child: their perceptions three years after cochlear implantation. Deaf Educ Int 2002

19 Fortunato-Tavares T, Befi-Lopes D, Bento RF, Andrade CR. Children with cochlear implants: communication skills and quality of life. Rev Bras Otorrinolaringol (Engl Ed) 2012;78(01):15–25. https://www.scielo.br/j/bjorl/a/HkHsm6vPFxMjkrRHqMwhKcg/?lang=en cited 2020 Nov 7 [Internet]

20 World Health Organization. CIF-CJ: Classificação Internacional de Funcionalidade, Incapacidade e Saúde, Versão para Crianças e Jovens. Edusp; 2011

21 Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Dev Med Child Neurol 1997;39(04):214–223. https://www.ncbi.nlm.nih.gov/pubmed/9183258 cited 2020 Nov 6 [Internet]

22 Silva DBR, Pfeifer LI, Funayama CAR. GMFCS - E&R: Gross motor function classification system - expanded and revised [Internet]. 2010 [cited 2020 Nov 6]; Available from: https://repositorio.usp.br/item/002175082

23 Vieira S. Introdução à Bioestatística. Elsevier Brasil; 2015

24 Sharma A, Dorman MF, Spahr AJ. A sensitive period for the development of the central auditory system in children with cochlear implants: implications for age of implantation. Ear Hear 2002;23(06):532–539 cited 2020 Nov 6. Doi: 10.1097/00003446-200212000-00004 [Internet]

25 Cejas I, Hoffman MF, Quittner AL. Outcomes and benefits of pediatric cochlear implantation in children with additional disabilities: a review and report of family influences on outcomes. Pediatric Health Med Ther 2015;6:45–63 cited 2020 Nov 8. Doi: 10.2147/phmt.s65797 [Internet]

26 Fernandes NF, Yamaguti EH, Morettin M, Costa OA. Speech perception in users of hearing aid with auditory neuropathy

spectrum disorder. CoDAS 2016;28(01):22–26 cited 2020 Nov 8. Doi: 10.1590/231 7-1782/20162014157 [Internet]

27 Hoshino ACH, da Cruz DR, Goffi-Gomez MVS, et al. Audiometric evolution in multichannel cochlear implant users. Rev CEFAC 2013;15(02):297–304 Available from https://www.scielo.br/scielo.php?script=sci_arttext&pid=S1516184620130002000005 cited 2020 Nov 6 [Internet]

28 Alvarenga Kde F, Vicente LC, Lopes RCF, et al. Development of P1 cortical auditory evoked potential in children with sensorineural hearing loss after cochlear implantation: longitudinal study. CoDAS 2013;25(06):521–526. https://www.scielo.br/scielo.php?pid=S2317178220130006005 21&script=sci_arttext&tlng=pt cited 2020Nov7 [Internet]

29 Silva BCS, Moret ALM, Silva LTDN, Costa OAD, Alvarenga KF, Silva-Comerlatto MPD. Glendonald Auditory Screening Procedure (GASP): clinical markers of the development of auditory recognition and comprehension abilities in children using cochlear implants. CoDAS 2019;31(04):e20180142

30 dos Santos MJD, Lamônica DAC, Ribeiro Mde M, McCracken W, Silva LT, Costa OA. Outcomes of cochlear implanted children with cerebral palsy: A holistic approach. Int J Pediatr Otorhinolaryngol 2015;79(07):1090–1095. https://pubmed.ncbi.nlm.nih.gov/25977237/ cited 2020 Nov 6 [Internet]

31 Steven RA, Green KMJ, Broomfield SJ, Henderson LA, Ramsden RT, Bruce IA. Cochlear implantation in children with cerebral palsy. Int J Pediatr Otorhinolaryngol 2011;75(11):1427–1430. https://pubmed.ncbi.nlm.nih.gov/21893352/ cited 2020Nov6 [Internet]

32 von der Hagen M, Pivarcsi M, Liebe J, et al. Diagnostic approach to microcephaly in childhood: a two-center study and review of the literature. Dev Med Child Neurol 2014;56(08):732–741. https://pubmed.ncbi.nlm.nih.gov/24617602/ cited 2020 Nov 6 [Internet]

33 Byun H, Moon IJ, Kim EY, et al. Performance after timely cochlear implantation in prelingually deaf children with cerebral palsy. Int J Pediatr Otorhinolaryngol 2013;77(06):1013–1018. https://pubmed.ncbi.nlm.nih.gov/23639338/ cited 2020 Nov 6 [Internet]

34 Moeller MP. Early intervention and language development in children who are deaf and hard of hearing. Pediatrics 2000;106 (03):E43. https://www.ncbi.nlm.nih.gov/pubmed/10969127 cited 2019 Oct 14 [Internet]

35 Padovani CMCA, Teixeira ER. From babbling to speech - reflections on the importance of initial linguistic activities and the development of oral language in children with hearing impairment. Distúrb Comun 2005;17(01): Available from https://revistas.pucsp.br/dic/article/view/11681 cited 2020 Nov 6 [Internet]

36 Spinazola Cde CAzevedo TL de, Gualda DS, Cia F. Correlation between socioeconomic status, needs, social support and family resources of mothers of children with physical disabilities, Down syndrome and autism. Rev Educ Espec 2018;31(62):697–712

37 Shin M-S, Kim S-K, Kim S-S, Park M-H, Kim C-S, Oh S-H. Comparison of cognitive function in deaf children between before and after cochlear implant. Ear Hear 2007;28(02, Suppl)22S–28S

38 Pantelemon C, Necula V, Berghe A-S, et al. Neurodevelopmental Aspects and Cortical Auditory Maturation in Children with Cochlear Implants. Medicina 2020;56(07):

