Qualitative retrospective-longitudinal analysis of factors influencing the development of auditory and language skills in children with cochlear implants

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Abstract
Background: Auditory results of cochlear implant (CI) vary according to both the presence/absence and severity of the malformations and the clinical condition of the patient.
Methods: This retrospective-longitudinal study examined the trend of the hearing threshold, verbal perception and CI mapping parameters in 50 pediatric patients, who underwent CI surgery at an age ≤7 years over a 12-month period (from activation to 12 months post-IC). The sample (4 groups) was clustered according to etiology, presence or absence of inner ear dysplasia and/or associated disabilities; a second subdivision (3 groups) considered the time of CI intervention (< 18 months, 18-36 months, > 36 months).
Results: The whole cohort shows an overall improvement in both tonal audiometric threshold and verbal perception over time. The subdivision according to etiology shows better results in children characterized by a normo-developed cochlea and absence of associated disabilities. The subdivision according to the time of CI intervention shows a greater development of perceptual abilities in children implanted before 18 months of age. Electrophysiological parameters and CI stimulation levels show a standard trend over time in the different groups and higher C levels in the group of patients with congenital conditions.
Conclusion: Nowadays, anatomical malformations of the inner ear are no longer a contraindication for CI surgery. An interdisciplinary approach and detailed counseling regarding the expected results should be tailored according to both the etiology of the hearing loss and the time of the CI intervention.

Keywords: Cochlear implant, hearing loss, impedance, current levels, etiology

Introduction

Pediatric hearing loss affects 1 to 3 children per 1000 births (Hawley et al., 2017) and it is classified according to multiple criteria: site responsible for the hearing loss, its severity (mild, moderate, severe and profound), the age of onset (pre- to post-verbal), the duration of hearing deprivation (permanent hearing loss, transient), the course over time (stable, progressive) and the etiological factors.

Permanent pediatric hearing loss is classified into: congenital forms (present at birth and caused by exogenous or endogenous factors) and acquired forms (occurring after birth). In particular, in 20-35% (Chadha et al.,2009; Sennaroglu, 2010; Ha et al.,2012) of children, congenital hearing loss is due to anatomical malformations that can affect both the outer and middle ear as well as the inner ear, with particular involvement of the cochlea, the VIII cranial nerve, the semicircular canals, and the vestibular aqueduct: the above mentioned lesions usually leads to sensorineural hearing loss with consequent speech impairments. Therefore, the timing and appropriateness of identifying and taking charge of the baby are essential in order to set up a therapeutic-rehabilitative plan aimed at obtaining the best results desirable by means of hearing aids or, for severe to profound hypoacusis, cochlear implants (CIs). (Ronner, 2020)

The first CI surgery in pediatric subjects dates back to the mid-1980s (Luxford &

House, 1985). In particular, in 1983, the first application of CIs in an altered cochlea was documented, thus extending the indication for implantation also to children suffering from abnormalities of the cochleovestibular structures (e.g. malformations of the inner ear) considered until then a contraindication to the use of this device. (Luxford & House, 1985; Miyamoto et al., 1986; Silverstein et al.,1988; Slattery & Luxford,1995; Tucci et al.,1995; Turrini et al.,1997;Woolley et al.,1998)

Over the years, the growing experience with cochlear implants has made it possible to affirm that the outcome of CI in pediatric patients is influenced not only by factors such as the onset of hearing loss and the duration of hearing deprivation, but also by the age at which the cochlear implant was performed, the presence of any additional associated disabilities, and the cochleovestibular anatomy. Therefore, the age at which the implant is applied is one of the most important elements influencing the outcome achieved by pediatric patients. As proof of this, Ching & Dillon (2013) examined the influence of implantation age on the global perceptual-communicative outcome of 134 children rehabilitated with CIs and found that from the age of 6 months, there is a delay of ± 1/2 standard deviations in global outcomes for every 6 months delay at surgery (Ching et al.,2013). Similarly, Houston et al. (2012) also compared the perceptual-linguistic abilities of pre-school CI patients to a sample of normal hearing children, revealing that children implanted before the age of one year showed a similar outcome to that of the normal hearing group, whereas children implanted after the age of one year showed statistically worse results (Houston et al.,2012). Overall, there are numerous studies in the literature showing that early rehabilitation intervention leads to both better development of perceptual-communicative skills and better overall results.(Ching et al.,2013; Dettmann et al.,2016; Houston & Miyamoto,2010; Franchella et al.,2023)

In addition to this, the literature reports how pediatric patients with hearing deficits of non-genetic etiology obtain excellent performance with implantation if the cochlear and auditory nerve gross anatomy is preserved (Shi et al., 2019; Eisenman et al., 2001; Ozkan et al., 2021). Indeed, in the presence of dysplasia of the cochlea the outcomes with implantation vary according to the severity of the malformation (Papsin,2005; Brotto et al.,2021): indicatively excellent scores are reported in the case of enlarged vestibular aqueduct (EVA) (Park et al.,2017), incomplete partition (IP) type II (also called Mondini-type) malformations, partial aplasia of the semicircular canals; markedly limited benefits in the case of IP type I or III, cochlear hypoplasia, common cavity, or complete aplasia of the semicircular canals (Papsin,2005;Brotto et al.,2019a). Moreover, if these anomalies are associated with syndromic pictures, they lead to even poorer performance: for example, in patients suffering from CHARGE syndrome, the outcome might be negatively influenced by alterations in the VIII nerve(s), such as hypoplasia, and the presence of cognitive and/or neurological disorders (Young et al.,2017). This is also the case for all those with cochlear nerve deficiency (CND) (Peng et al.,2017; Brotto et al.,2019b), i.e. a cochlear nerve that is absent or smaller than normal. (Glastonbury et al.,2002)

Regarding the parameters of the CI, studies in the literature show that, compared to patients with normally developed cochlea (Zwolan et al.,2008), impedance values and T and C levels are similar for patients with Mondini Malformation (Li et al., 2004) or incomplete partition type II/EVA (Adunka et al.,2012). Whereas threshold levels are systematically increased for patients with severe inner ear malformations (e.g. cochlear hypoplasia, common cavity, etc.). (Papsin,2005; Kocabay et al.,2022; Zhao et al.,2003; Liu et al.,2022) The latter usually succeed in improving their tonal but not verbal discrimination following frequent changes in pulsewidth (PW) (Papsin,2005) or rate (Kocabay et al.,2022). Finally, in patients with CND the parameters of stimulation frequency and pulse duration, when looking at a general sample, are significantly higher than in control patients.(Wei et al.,2023) However, Wei et al. also report the need for some patients to reduce the rate or increase the PW or set a three-phasic stimulus (Wei et al.,2023), to manage the possible occurrence of facial nerve stimulation when higher current levels are required.

The aim of this study is to compare CI fitting parameters, auditory abilities and perceptual outcomes pre- and post-IC in children with and without malformations about the type of malformation, the presence or absence of associated disabilities and the timing of cochlear implantation.

Materials and Methods

The present retrospective cohort study, conducted at the Otorhinolaryngology Unit of the Azienda Ospedale-University of Padua, aims to examine the trend of the hearing threshold, verbal perception and CI mapping parameters in pediatric patients undergoing cochlear implantation (CI) surgery at the age of ≤ 7 years over 12-month (from activation to 12 months post-IC).

Sample

The final sample obtained consists of a total of 50 patients, homogeneous with respect to the brand of CI (i.e.: Cochlear™), the type of array (perimodiolar) and external processor model (CP1000) but characterized by a broad and distinct etiology of deafness. Therefore, the sample was divided into the following four groups according to: etiology, presence or absence of inner ear dysplasia (inner ear abnormalities) in relation to the severity of the dysplasia, presence or absence of associated disabilities.(Papsin,2005;Peng et al.,2017;Buchman et al.,2004; Isaiah et al.,2017; Birman et al.,2016;Yoshida et al.,2017a)

1. Group A with the absence of inner ear dysplasia and associated disabilities (21 patients): hearing loss due to infectious causes (Cytomegalovirus, Meningitis) and mutation of the gene connexin 26 (Cx26);
2. Group B with presence of inner ear dysplasia or associated disabilities (8 patients): hearing loss due to syndromic causes (Waardenburg's Sdr. type I, Down's Sdr., EVA Sdr. and Mondini malformation (EVA plus IP2)), prematurity at birth associated with cerebral hemorrhage, ototoxic drugs associated with variants of the mutation of the gene connexin 26 (included in this group as they are associated with the presence of an overall developmental delay);
3. Group C with the presence of severe inner ear dysplasia and/or severe associated disabilities (13 patients): hearing loss due to syndromic causes (Sdr. of Charge, Sdr. of Waardenburg type IV), malformations of the inner ear (rudimentary otocyst, common cavity), infectious causes (CMV) and prematurity at birth with severe neurological complications and associated disabilities;
4. Group D with unknown etiology and presence of associated disabilities (8 patients).

Then, this sample was further subdivided according to the age of CI intervention:

1. < 18 months (24 patients): 12 patients belonging to group A, 5 patients belonging to group B, 1 patient belonging to group C, 6 patients belonging to group D
2. 18 - 36 months (14 patients): 4 patients belonging to group A, 2 patients belonging to group B, 7 patients belonging to group C, 1 patient belonging to group D
3. > 36 months (12 patients): 5 patients belonging to group A, 1 patient belonging to group B, 5 patients belonging to group C, 1 patient belonging to group D.

In addition, exclusion criteria were the following: presence of abnormalities in the insertion of the array (i.e. partial insertion of the electrode cable) and one or more electrodes off due to open circuits or short-circuits.

Data collection

Each subject included in the study, after cochlear implant surgery and subsequent activation, was included in the audiological follow-up with controls established at 1, 3, 6 and 12 months after activation. At every control, each patient underwent both a free-field pure tone audiometric examination with CI, by means of sending warble tone (modulated signal) determining the tone threshold for the frequencies 500-1000-2000 Hz and calculating the respective PTA (pure tone average) either by logopaedic assessment, evaluating the different levels of auditory perception according to the scheme proposed by Erber (Erber,1982) (from the detection of simple verbal sounds to the recognition of words in an open list), in order to monitor the trend and any progress of each subject over the first year of activation of the CI. Subsequently, the audiometric data obtained were compared to two important parameters of cochlear implant function, electrical impedance (measurement of electrode function) and C-level (maximum level of electrical current that allows prolonged and comfortable listening over time), evaluating the values recorded according to the etiology and time of CI intervention in each study group. Moreover, the electrodes of the array were divided into three groups (apical, middle and basal electrodes), according to their position within the cochlea, in order to better characterize the C-level trends of the entire cochlear partition. Specifically, the apical electrodes (22-15 el.) are located in the apical gyrus of the cochlea, the middle electrodes (14-8 el.) in the middle gyrus, and the basal electrodes (7-1 el.) in the basal gyrus. Finally, based on this distribution, the average of C-level was calculated.

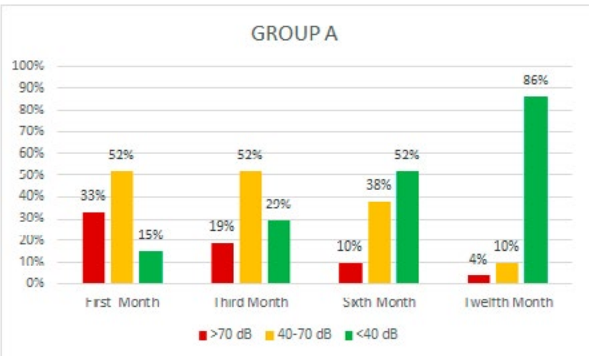

Figure 1 - PTA trend in group A

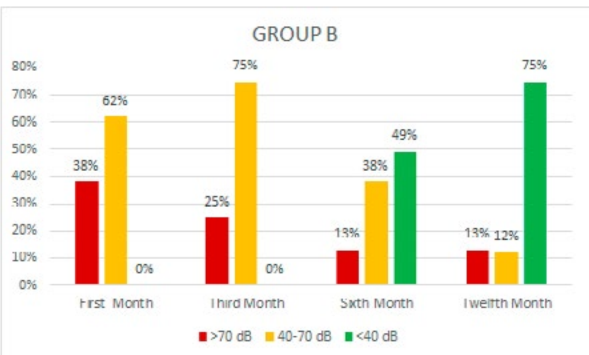

Figure 2 - PTA trend in group B

Data analysis

A qualitative analysis using various types of graphs (histogram graph, line graph) was performed to assess and analyze the trend over 1 year after the activation of the CI of the various parameters investigated in the sample: both for the breakdown according to etiology and respective expected outcome, and for the breakdown according to the time of CI intervention.

Results

The figures below (figure 1, 2, 3, 4) show the development of PTA over a 1-year period, respectively at 1, 3, 6 and 12 months post activation of the CI, in relation to the etiology and the respective expected outcome.

1st month: PTA > 70 dB HL (33% pt.), PTA = 40-70 dB HL (52% pt.), PTA < 40 dB HL (15% pt.)
3rd month: PTA > 70 dB HL (19% pt.), PTA = 40-70 dB HL (52% pt.), PTA < 40 dB HL (29% pt.)
6th month: PTA > 70 dB HL (10% pt.), PTA = 40-70 dB HL (38% pt.), PTA < 40 dB HL (52% pt.)
12th month: PTA > 70 dB HL (4% pt.), PTA = 40-70 dB HL (10% pt.), PTA < 40 dB HL (86% pt.)

1st month: PTA > 70 dB HL (38% pt.), PTA = 40-70 dB HL (62% pt.), PTA < 40 dB HL (0% pt.)
3rd month: PTA > 70 dB HL (25% pt.), PTA = 40-70 dB HL (75% pt.), PTA < 40 dB HL (0% pt.)
6th month: PTA > 70 dB HL (13% pt.), PTA = 40-70 dB HL (38% pt.), PTA < 40 dB HL (49% pt.)
12th month: PTA > 70 dB HL (13% pt.), PTA = 40-70 dB HL (12% pt.), PTA < 40 dB HL (75% pt.)

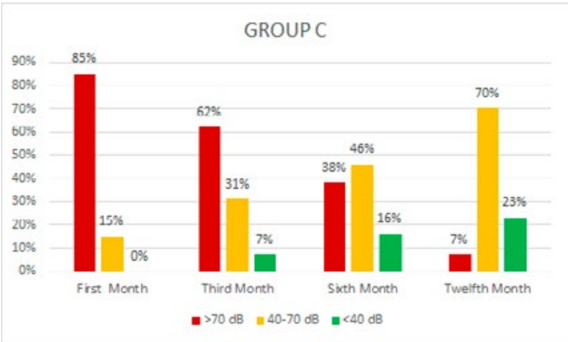

Figure 3 - PTA trend in group C

1st month: PTA > 70 dB HL (85% pt.), PTA = 40-70 dB HL (15% pt.), PTA < 40 dB HL (0% pt.)
3rd month: PTA > 70 dB HL (62% pt.), PTA = 40-70 dB HL (31% pt.), PTA < 40 dB HL (7% pt.)
6th month: PTA > 70 dB HL (38% pt.), PTA = 40-70 dB HL (46% pt.), PTA < 40 dB HL (16% pt.)
12th month: PTA > 70 dB HL (7% pt.), PTA = 40-70 dB HL (70% pt.), PTA < 40 dB HL (23% pt.)

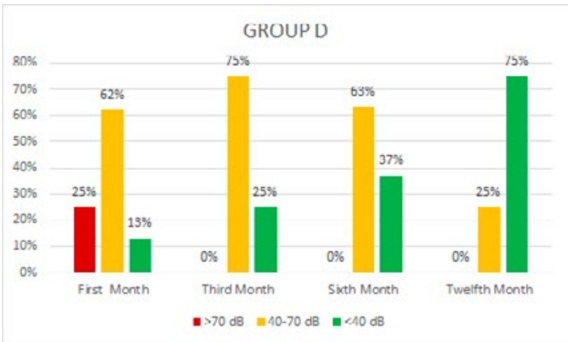

Figure 4 - PTA trend in group D

1st month: PTA > 70 dB HL (25% pt.), PTA = 40-70 dB HL (62% pt.), PTA < 40 dB HL (13% pt.)
3rd month: PTA > 70 dB HL (0% pt.), PTA = 40-70 dB HL (75% pt.), PTA < 40 dB HL (25% pt.)
6th month: PTA > 70 dB HL (0% pt.), PTA = 40-70 dB HL (63% pt.), PTA < 40 dB HL (37% pt.)
12th month: PTA > 70 dB HL (0% pt.), PTA = 40-70 dB HL (25% pt.), PTA < 40 dB HL (75% pt.)

Figure 5 shows the average PTA values of each of the previous 4 groups at 1, 3, 6 and 12 months post activation of the CI, respectively.

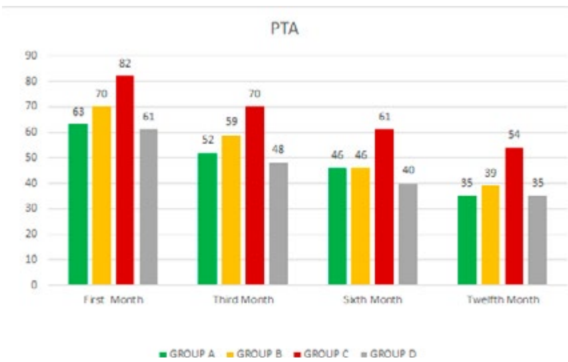

Figure 5 - Average PTA trend over time

1st month: average PTA = 63 dB HL ±17 dB (dB) (group A), average PTA = 70 dB HL ±13 dB (group B), average PTA = 82 dB HL ±10 dB (group C), average PTA = 61 dB HL ± 16 dB (group D)
3rd month: average PTA = 52 dB HL ±18 dB (group A), average PTA = 59 dB HL ±14 dB (group B), average PTA = 70 dB HL ±16 dB (group C), average PTA = 48 dB HL ±8 dB (group D)
6th month: average PTA = 46 dB HL ±20 dB (group A), average PTA = 46 dB HL ±15 dB (group B), average PTA = 61 dB HL ±16 dB (group C), average PTA = 40 dB HL ±4 dB (group D)
12th month: average PTA = 35 dB HL ±10 dB (group A), average PTA = 39 dB HL ±17 dB (group B), average PTA = 54 dB HL ±16 dB (group C), average PTA = 35 dB HL dB (group D)

The figures below (figures 6, 7, 8) show the development of PTA over 1 year, respectively at 1, 3, 6 and 12 months post activation of the CI, about the time of CI intervention.

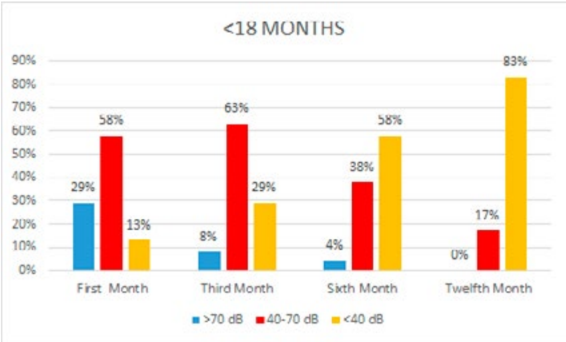

Figure 6 - PTA trend in the < 18 months group

1st month: PTA > 70 dB HL (29% pt.), PTA = 40-70 dB HL (58% pt.), PTA < 40 dB HL (13% pt.)
3rd month: PTA > 70 dB HL (8% pt.), PTA = 40-70 dB HL (63% pt.), PTA < 40 dB HL (29% pt.)
6th month: PTA > 70 dB HL (4% pt.), PTA = 40-70 dB HL (38% pt.), PTA < 40 dB HL (58% pt.)
12th month: PTA > 70 dB HL (0% pt.), PTA = 40-70 dB HL (17% pt.), PTA < 40 dB HL (83% pt.)

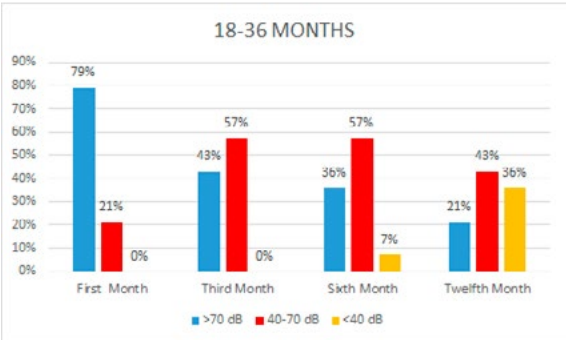

Figure 7 - PTA trend in the 18-36 months group

1 month: PTA > 70 dB HL (79% pt.), PTA = 40-70 dB HL (21% pt.), PTA < 40 dB HL (0% pt.)
3rd month: PTA > 70 dB HL (43% pt.), PTA = 40-70 dB HL (57% pt.), PTA < 40 dB HL (0% pt.)
6th month: PTA > 70 dB HL (36% pt.), PTA = 40-70 dB HL (57% pt.), PTA < 40 dB HL (7% pt.)
12th month: PTA > 70 dB HL (21% pt.), PTA = 40-70 dB HL (43% pt.), PTA < 40 dB HL (36% pt.)

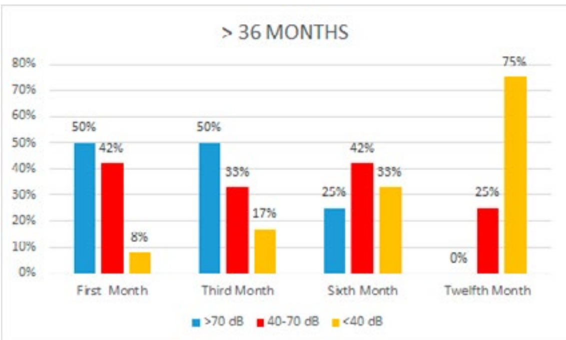

Figure 8 - PTA trend in the > 36 months group

1st month: PTA > 70 dB HL (50% pt.), PTA = 40-70 dB HL (42% pt.), PTA < 40 dB HL (8% pt.)
3rd month: PTA > 70 dB HL (50% pt.), PTA = 40-70 dB HL (33% pt.), PTA < 40 dB HL (17% pt.)
6th month: PTA > 70 dB HL (25% pt.), PTA = 40-70 dB HL (42% pt.), PTA < 40 dB HL (33% pt.)
12th month: PTA > 70 dB HL (0% pt.), PTA = 40-70 dB HL (25% pt.), PTA < 40 dB HL (75% pt.)

Figure 9 shows the average PTA values of each of the previous 3 groups at 1, 3, 6 and 12 months post activation of the CI, respectively.

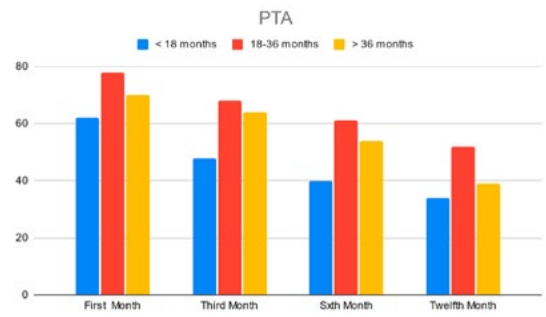

Figure 9 - Average PTA trend over time

1st month: average PTA = 62 dB HL ±16 dB (group < 18 months), average PTA = 78 dB HL ±12 dB (group 18-36 months), average PTA = 70 dB HL ±19 dB (group > 36 months)
3rd month: average PTA = 48 dB HL ±13 dB (group < 18 months), average PTA = 68 dB HL ±15 dB (group 18-36 months), average PTA = 64 dB HL ±21 dB (group > 36 months)
6th month: average PTA = 40 dB HL ±11 dB (group < 18 months), average PTA = 61 dB HL ±19 dB (group 18-36 months), average PTA = 54 dB HL ±20 dB (group > 36 months)
12th month: average PTA = 34 dB HL ±9 dB (group < 18 months), average PTA = 52 dB HL ±19 dB (group 18-36 months), average PTA = 39 dB HL ±13 dB (group > 36 months)

The figures below (figures 10, 11, 12, 13) show the development of the verbal perception scores for 1 year, respectively at 1, 3, 6 and 12 months post activation of the CI, concerning the etiology and the respective intended outcome.

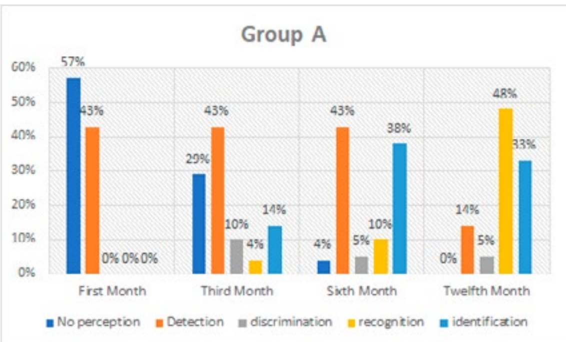

Figure 10 - Verbal perception trend in group A

1st month: no perception (57% pt.), detection (43% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
3rd month: no perception (29% pt.), detection (43% pt.), discrimination (10% pt.), recognition (4% pt.), identification (14% pt.)
6th month: no perception (4% pt.), detection (43% pt.), discrimination (5% pt.), recognition (10% pt.), identification (38% pt.)
12th month: no perception (0% pt.), detection (14% pt.), discrimination (5% pt.), recognition (48% pt.), identification (33% pt.)

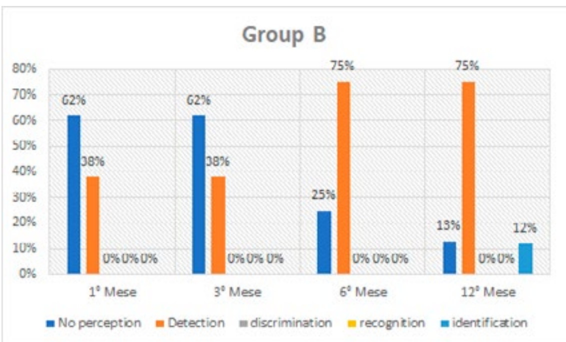

Figure 11 - Verbal perception trend in group B

1st month: no perception (62% pt.), detection (38% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
3rd month: no perception (62% pt.), detection (38% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
6th month: no perception (25% pt.), detection (75% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
12th month: no perception (13% pt.), detection (75% pt.), discrimination (0% pt.), recognition (0% pt.), identification (12% pt.)

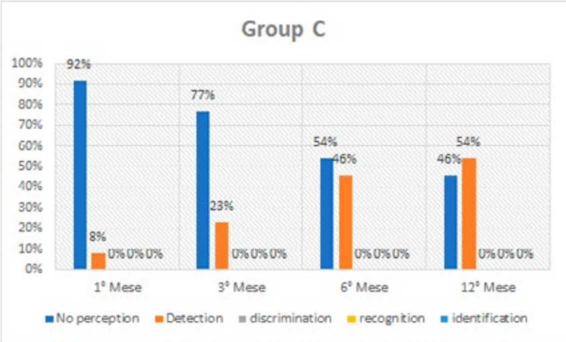

Figure 12 - Verbal perception trend in group C

1st month: no perception (92% pt.), detection (8% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
3rd month: no perception (77% pt.), detection (23% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
6th month: no perception (54% pt.), detection (46% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
12th month: no perception (46% pt.), detection (54% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)

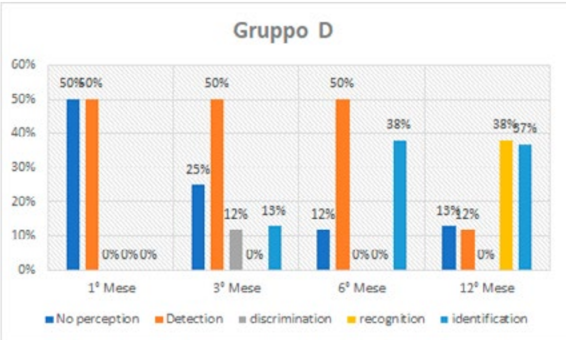

Figure 13 - Verbal perception trend in group D

1st month: no perception (50% pt.), detection (60% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
3rd month: no perception (25% pt.), detection (50% pt.), discrimination (12% pt.), recognition (0% pt.), identification (13% pt.)
6th month: no perception (12% pt.), detection (50% pt.), discrimination (0% pt.), recognition (0% pt.), identification (38% pt.)
12th month: no perception (13% pt.), detection (12% pt.), discrimination (0% pt.), recognition (38% pt.), identification (37% pt.)

The figures below (figures 14, 15, 16) show the development of verbal perception scores over a 1-year period, at 1, 3, 6 and 12 months post-CI activation, respectively, in relation to the time of CI intervention.

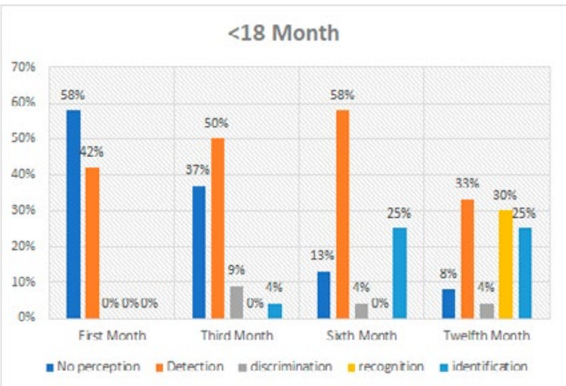

Figure 14 - Verbal perception trend in the group < 18 months

1st month: no perception (58% pt.), detection (42% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
3rd month: no perception (37% pt.), detection (50% pt.), discrimination (9% pt.), recognition (0% pt.), identification (4% pt.)
6th month: no perception (13% pt.), detection (58% pt.), discrimination (4% pt.), recognition (0% pt.), identification (25% pt.)
12th month: no perception (8% pt.), detection (33% pt.), discrimination (4% pt.), recognition (30% pt.), identification (25% pt.)

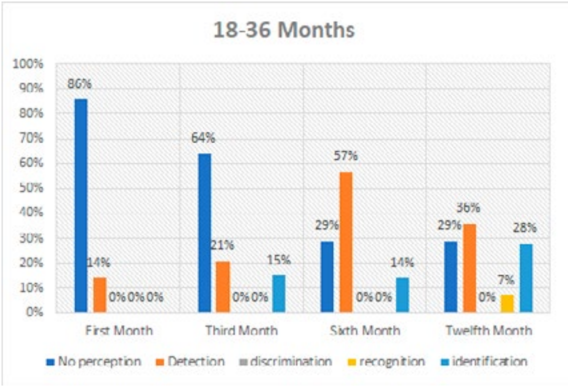

Figure 15 - Verbal perception trend in the group 18-36 months

1st month: no perception (86% pt.), detection (14% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
3rd month: no perception (64% pt.), detection (21% pt.), discrimination (0% pt.), recognition (0% pt.), identification (15% pt.)
6th month: no perception (29% pt.), detection (57% pt.), discrimination (0% pt.), recognition (0% pt.), identification (14% pt.)
12th month: no perception (29% pt.), detection (36% pt.), discrimination (0% pt.), recognition (7% pt.), identification (28% pt.)

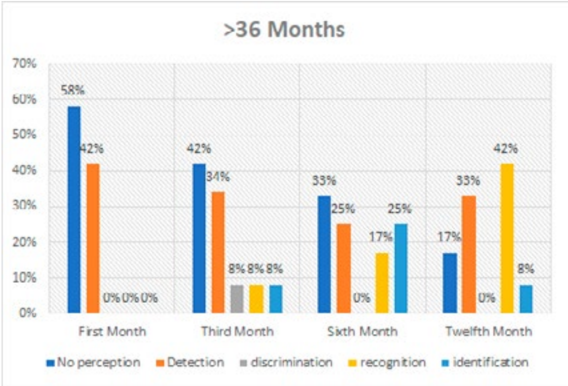

Figure 16 - Verbal perception trend in the group > 36 months

1st month: no perception (58% pt.), detection (42% pt.), discrimination (0% pt.), recognition (0% pt.), identification (0% pt.)
3rd month: no perception (42% pt.), detection (34% pt.), discrimination (8% pt.), recognition (8% pt.), identification (8% pt.)
6th month: no perception (33% pt.), detection (25% pt.), discrimination (0% pt.), recognition (17% pt.), identification (25% pt.)
12th month: no perception (17% pt.), detection (33% pt.), discrimination (0% pt.), recognition (42% pt.), identification (8% pt.)

Figures 17 and 18 show the trend of the average electrical impedance values over 1 year, respectively at 1, 3, 6 and 12 months post CI activation, with both the etiology and the respective expected outcome and the time of CI intervention.

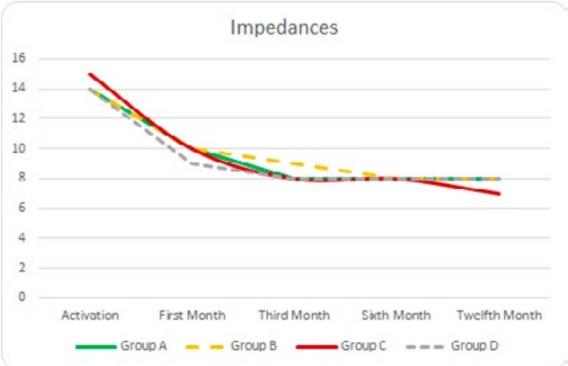

Figure 17 - Average electrical impedance trend to etiology and respective expected outcome

Group A: activation 14 kΩ ±1 kΩ , 1st month 10 kΩ ±2 kΩ , 3rd month 8 kΩ ±1 kΩ , 6th month 8 kΩ ±1 kΩ , 12th month 8 kΩ ±1 kΩ .
Group B: activation 14 kΩ ±1 kΩ , 1st month 10 kΩ ±2 kΩ , 3rd month 9 kΩ ±1 kΩ , 6th month 8 kΩ ±2 kΩ , 12th month 8 kΩ ±1 kΩ .
Group C: activation 15 kΩ ±1 kΩ , 1st month 10 kΩ ±2 kΩ, 3rd month 8 kΩ ±1 kΩ, 6th month 8 kΩ ±1 kΩ, 12th month 7 kΩ ±1 kΩ.
Group D: activation 14 kΩ ±1 kΩ, 1st month 9 kΩ ±2 kΩ, 3rd month 8 kΩ ±1 kΩ, 6th month 8 kΩ ±1 kΩ, 12th month 8 kΩ ±1 kΩ

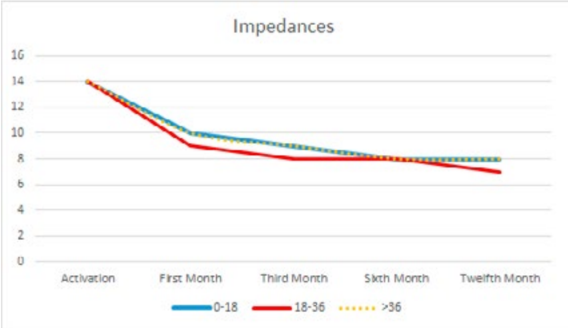

Figure 18 - Average electrical impedance trend to the time of IC intervention

Group < 18 months: activation 14 kΩ ±1 kΩ, 1st month 10 kΩ ±2 kΩ, 3rd month 9 kΩ ±1 kΩ, 6th month 8 kΩ ±1 kΩ, 12th month 8 kΩ ±1 kΩ.
Group 18-36 months: activation 14 kΩ ±1 kΩ, 1st month 9 kΩ ±1 kΩ, 3rd month 8 kΩ ±1 kΩ, 6th month 8 kΩ ±1 kΩ, 12th month 7 kΩ ±1 kΩ.
Group > 36 months: activation 14 kΩ ±1 kΩ, 1st month 10 kΩ ±2 kΩ, 3rd month 9 kΩ ±1 kΩ, 6th month 8 kΩ ±1 kΩ, 12th month 8 kΩ ±1 kΩ.

The figures below show the trend of the average values of the apical, mean and basal electrode C-current levels for 1 year, respectively at 1, 3, 6 and 12 months post CI activation, about both the etiology and the respective expected outcome (figures 19, 20, 21) and the time of CI intervention (figures 22, 23, 24).

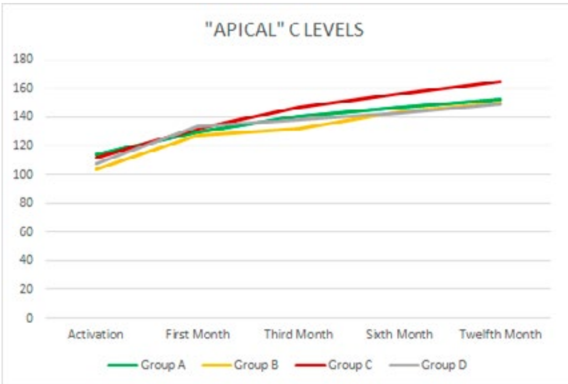

Figure 19 - Average trend in C el. apical levels

Group A: activation 114 CL ±21 CL, 1st month 130 CL ±21 CL, 3rd month 141 CL ±21 CL, 6th month 147 CL ±22 CL, 12th month 152 CL ±19 CL.
Group B: activation 104 CL ±24 CL, 1st month 127 CL ±21 CL, 3rd month 132 CL ±14 CL, 6th month 144 CL ±14 CL, 12th month 150 CL ±13 CL.
Group C: activation 112 CL ±18 CL, 1st month 132 CL ±21 CL, 3rd month 147 CL ±17 CL, 6th month 156 CL ±15 CL, 12th month 165 CL ±15 CL.
Group D: activation 108 CL ±25 CL, 1st month 134 CL ±21 CL, 3rd month 138 CL ±17 CL, 6th month 143 CL ±14 CL, 12th month 149 CL ±13 CL.

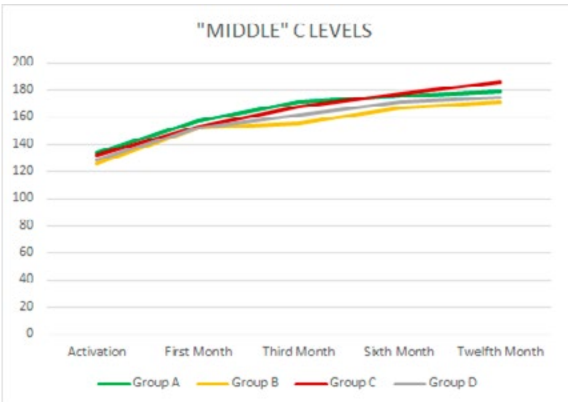

Figure 20 - Average trend in C el. middle levels

Group A: activation 134 CL ±24 CL, 1st month 157 CL ±21 CL, 3rd month 171 CL ±19 CL, 6th month 176 CL ±16 CL, 12th month 179 CL ±13 CL.
Group B: activation 126 CL ±20 CL, 1st month 152 CL ±19 CL, 3rd month 156 CL ±15 CL, 6th month 167 CL ±15 CL, 12th month 171 CL ±12 CL.
Group C: activation 132 CL ±15 CL, 1st month 153 CL ±21 CL, 3rd month 168 CL ±14 CL, 6th month 177 CL ±19 CL, 12th month 186 CL ±16 CL.
Group D: activation 129 CL ±22 CL, 1st month 152 CL ±19 CL, 3rd month 162 CL ±12 CL, 6th month 171 CL ±11 CL, 12th month 175 CL ±10 CL.

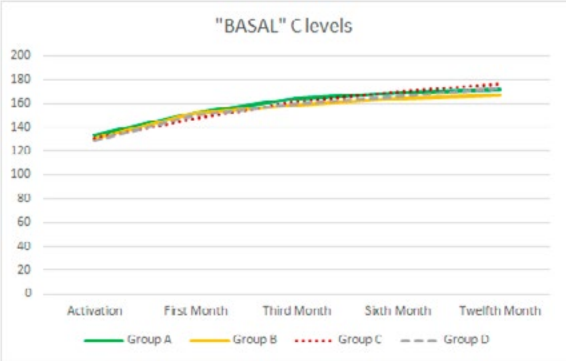

Figure 21 - Average trend in C el. basal levels

Group A: activation 134 CL ±23 CL, 1st month 152 CL ±18 CL, 3rd month 164 CL ±19 CL, 6th month 169 CL ±14 CL, 12th month 172 CL ±13 CL.
Group B: activation 131 CL ±19 CL, 1st month 152 CL ±13 CL, 3rd month 159 CL ±10 CL, 6th month 164 CL ±14 CL, 12th month 167 CL ±14 CL.
Group C: activation 131 CL ±16 CL, 1st month 148 CL ±20 CL, 3rd month 162 CL ±15 CL, 6th month 170 CL ±17 CL, 12th month 177 CL ±13 CL.
Group D: activation 129 CL ±20 CL, 1st month 150 CL ±20 CL, 3rd month 160 CL ±9 CL, 6th month 166 CL ±11 CL, 12th month 173 CL ±6 CL.

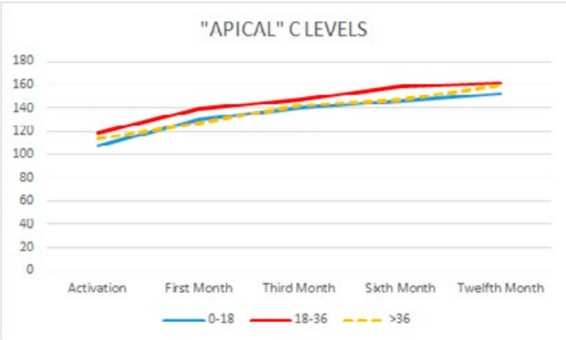

Figure 22 - Average trend in C el. apical levels

Group < 18 months: activation 107 CL ±21 CL, 1st month 130 CL ±19 CL, 3rd month 140 CL ±16 CL, 6th month 146 CL ±16 CL, 12th month 152 CL ±15 CL.
Group 18-36 months: activation 119 CL ±18 CL, 1st month 139 CL ±21 CL, 3rd month 147 CL ±21 CL, 6th month 158 CL ±21 CL, 12th month 161 CL ±20 CL.
Group > 36 months: activation 114 CL ±23 CL, 1st month 127 CL ±20 CL, 3rd month 142 CL ±20 CL, 6th month 147 CL ±16 CL, 12th month 159 CL ±19 CL.

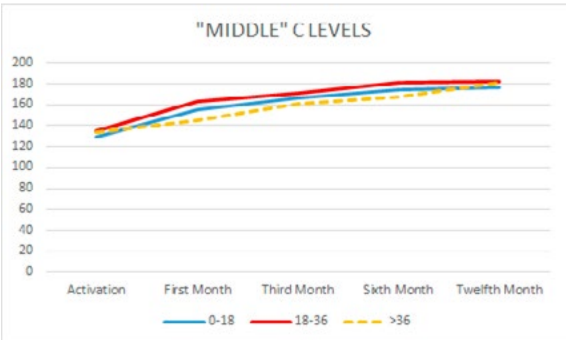

Figure 23 - Average trend in C el. middle levels

Group < 18 months: activation 129 CL ±20 CL, 1st month 155 CL ±19 CL, 3rd month 167 CL ±16 CL, 6th month 174 CL ±14 CL, 12th month 177 CL ±12 CL.
Group 18-36 months: activation 135 CL ±24 CL, 1st month 163 CL ±18 CL, 3rd month 171 CL ±19 CL, 6th month 181 CL ±20 CL, 12th month 182 CL ±18 CL.
Group > 36 months: activation 134 CL ±19 CL, 1st month 145 CL ±19 CL, 3rd month 161 CL ±16 CL, 6th month 168 CL ±12 CL, 12th month 181 CL ±12 CL.

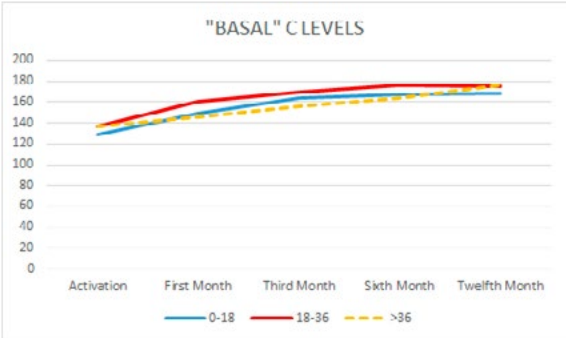

Figure 24 - Average trend in C el. basal levels

Group < 18 months: activation 129 CL ±17CL, 1st month 149 CL ±14 CL, 3rd month 164 CL ±11 CL, 6th month 167 CL ±12 CL, 12th month 168 CL ±13 CL.
Group 18-36 months: activation 137 CL ±15 CL, 1st month 160 CL ±9 CL, 3rd month 169 CL ±10 CL, 6th month 176 CL ±15 CL, 12th month 175 CL ±13 CL.
Group > 36 months: activation 137 CL ±24 CL, 1st month 146 CL ±22 CL, 3rd month 156 CL ±18 CL, 6th month 164 CL ±14 CL, 12th month 176 CL ±13 CL.

Discussion

Early diagnosis and timely intervention are essential elements in promoting language development in children with hearing loss. In particular, in these patients, factors such as etiology and age of application of the rehabilitation device are crucial for successful hearing rehabilitation.

The sample of this study, characterized by a total of 50 patients differing in both the etiology of hearing loss and time of CI surgery, was subjected to a qualitative analysis comparing: cochlear implant adjustment parameters, audiometric thresholds and speech-language results. As a result of the findings, the best audiometric thresholds are represented by patients without malformations or disabilities (with particular reference to groups A and D), while the worst audiometric thresholds are represented by patients with associated malformations or disabilities, despite presenting an overall, albeit small, threshold improvement. Therefore, it can be seen that cochlear implantation provides better results in patients with a normally developed cochlea and no associated disability.

The logopaedic results, in agreement with what was found in tonal audiometry for the different groups analyzed, are better in group A and worse in groups B and C though with progressive improvements over time. Overall, as expected, group C presents the worst results for each parameter analyzed: over time, however, effective stimulation by the CI is evident following the gradual improvement in the tonal threshold, despite the poor scores obtained during the speech assessment (detection 54% of patients and absence of perception 46% of patients), probably due to the presence of inner ear malformations and comorbidities (affecting 62% of the subjects in this group). Group B, on the other hand, presents an overall positive trend but lower than expected based on the results obtained during the audiometric assessment (absence of perception 13% of patients, detection 75% of patients, identification 12% of patients): this finding could also be attributed, in addition to the fact that malformation pictures were found in these patients, to the presence of various types of difficulties in the psychomotor development of 50% of the subjects included in this group. In particular, the results obtained in groups B and C show how the presence of inner ear dysplasia (type of inner ear abnormality) significantly influences the perceptual-communicative outcome of CI patients, in agreement with what is reported in the literature. (Papsin,2005;Buchman et al.,2004; Isaiah et al.,2017; Birman et al.,2016)

Lastly, the cochlear implant adjustment parameters show: a uniform impedance with a standard trend over time, regardless of the etiology of the hearing impairment and the age of application of the CI, and a slightly higher C level, but within normal limits, in the group characterized by malformations, as reported in the literature (Zwolan et al.,2008).

In light of the findings, the results obtained confirm what was expected (Papsin,2005;Park et al.,2017;Young et al.,2017;Bayrak et al.,2017; Broomfield et al.,2013; Hoey et al.,2017;Yoshida et al.,2017b): reduced tonal thresholds and speech-language results for the subjects belonging to group C with the need to increase the cochlear implant C levels (Papsin,2005;Zwolan et al.,2008;Kocabay et al.,2022; Zhao et al.,2003; Liu et al.,2022). Nevertheless, compared to the baseline, there is an overall improvement in tonal thresholds and speech-language results for each group analyzed.

Then, the study sample was analyzed according to the time of cochlear implant surgery. This analysis revealed the following: the group 18 - 36 months is the worst in terms of performance, contrary to what was expected (this finding could be affected by the numerical inhomogeneity of the sample in this group), while the group < 18 months is the best in terms of the development of perceptual abilities, in agreement with what was expected. Furthermore, the latter finding is corroborated both by a study conducted by Ching & Dillon (2013)(Ching et al.,2013), which demonstrates better and greater development of perceptual and communicative skills in children implanted early, and by a study conducted by Houston & Miyamoto (2010) (Houston & Miyamoto,2010), which shows good speech scores in children implanted within the age of one year, similar to those obtained by normally hearing children, and poor speech scores in children implanted after the age of one year. (Ching et al.,2013;Dettmann et al.,2016;Houston & Miyamoto,2010) Concerning the latter study, the analyses carried out on the group of patients implanted after the age of 36 months reveal discrete results (recognition of 42% of patients and identification of 8% of patients at 12 months after activation of the CI), thus demonstrating the possibility of obtaining satisfactory performance even in subjects undergoing cochlear implantation late in life.

Finally, the cochlear implant adjustment parameters show: a uniform impedance with a standard trend over time, characterized by an initial decrease in the time between activation and the 1st month post-IC and a subsequent stabilization from the 3rd month until the 12th month post-IC as previously reported,(Brotto et al.,2022) for all three groups and a higher C level for the 18 - 36 month group which includes the largest number of patients from whom a poor outcome is expected. In particular, the group < 18 months shows higher stimulation levels for the basal and middle electrodes than the group > 36 months. According to our results, the C levels measured in our cohort are lower than those reported in the literature by Zwolan et al. (Zwolan et al.,2008). These results may be due to the structural characteristics of the implants considered since the previous study collected one hundred-three children who received a Nucleus device (CI24RCS, 74; CI24RCA, 25; CI24RST, 3; Freedom, 1) (Zwolan et al.,2008) while we considered patients with more recent surgeries and consequently more recent implants. Also, the present cohort considered only patients with perimodiolar electrodes and this may explain the reduced current levels necessary to obtain an optimal outcome when compared to the previous study.

The stability of impedance levels previously reported by Brotto et al. (Brotto et al.,2022), up to 10 years, is confirmed by the present study, even if the group considered is smaller and heterogeneous in terms of etiology. Moreover, the confirmed trend of impedance levels being reduced after activation seems to be related to the activity of the implant: indeed, the electrical stimulation seems to destroy the accumulation of proteins and other substances present on the surface of the array in the post-surgical period. All the above mentioned considerations about the impedance levels and the current levels required should be considered preliminary and future studies will be necessary to better specify if the etiology may have an impact on these parameters. A closer look, considering shorter intervals of time, might reveal possible variations, especially for patients with inner ear malformations.

Moreover, the present cohort is too limited to express useful results in terms of the values of these parameters for patients with etiologies that imply a sub-optimal auditory outcome, and this is particularly true for patients with malformations, in which the morphology of the inner ear, the surgical technique, and in the end the positioning of the array may have a strong impact on these current-related parameters.

The results obtained from the present study are influenced by the following limitations: the size and heterogeneity of the sample and its subdivision according to both the etiology of the hearing loss and the time of CI intervention. This entailed a difficult statistical comparison of the results with the consequent impossibility of identifying statistically significant differences, allowing only a qualitative analysis of the sample.

Conclusion

In recent years, thanks to the advances in surgical techniques and prosthetic devices, inner ear anomalies are no longer an insurmountable problem from an audiological point of view.

In most of the participants included in the present study, the chosen rehabilitation device restored auditory sensitivity. Nevertheless, performance about language development varies greatly, which is why it cannot be easily compared with hearing-impaired individuals of the same age who have no associated malformations or disabilities.

The above-mentioned results emphasize the importance of an interdisciplinary approach to ensure the best patient care; at the same time, detailed counseling is necessary to inform the patients' families about the expected outcomes of the rehabilitation intervention according to the etiology of the hearing loss and the time of CI intervention.

Future studies considering a larger, more homogeneous sample of patients and assessing possible additional parameters will reveal even more about these still open issues.

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