

Long-term Outcomes in Down Syndrome Children After Cochlear Implantation: Particular Issues and Considerations

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Objective: The aim of the study was to analyze the long-term outcomes after cochlear implantation in deaf children with Down syndrome (DS) regarding age at the first implantation and refer the results to preoperative radiological findings as well as postoperative auditory and speech performance. Additionally, the influence of the age at implantation and duration of CI use on postoperative hearing and language skills were closely analyzed in children with DS.

Study Design: Retrospective analysis.

Setting: Referral center (Cochlear Implant Center).

Materials and Methods: Nine children with Down syndrome were compared with 220 pediatric patients without additional mental disorders or genetic mutations. Patients were divided into four categories depending on the age of the first implantation: CAT1 (0–3 yr), CAT2 (4–5 yr), CAT3 (6–7 yr), and CAT4 (8–17 yr). The auditory performance was assessed with the meaningful auditory integration scales (MAIS) and categories of auditory performance (CAP) scales. The speech and language development were further evaluated with meaningful use of speech scale (MUSS) and speech intelligibility rating (SIR). The postoperative speech skills were analyzed and compared between the study group and the reference group by using nonparametric statistical tests. Anatomic abnormalities of the inner ear were examined using magnetic resonance imaging (MRI) and

high-resolution computed tomography of the temporal bones (HRCT).

Results: The mean follow-up time was 14.9 years (range, 13.1–18.3 yr). Patients with DS received a multichannel implant at a mean age of 75.3 months (SD 27.9; ranging from 21 to 127 mo) and 220 non-syndromic children from reference group at a mean age of 51.4 months (SD 34.2; ranging from 9 to 167 mo). The intraoperative neural response was present in all cases. The auditory and speech performance improved in each DS child. The postoperative mean CAP and SIR scores were 4.4 (SD 0.8) and 3.2 (SD 0.6), respectively. The average of scores in MUSS and MAIS/IT-MAIS scales was 59.8% (SD 0.1) and 76.9% (SD 0.1), respectively. Gathered data indicates that children with DS implanted with CI at a younger age (<6 years of age) benefited from the CI more than children implanted later in life, similarly in a control group. There were additional anomalies of the temporal bone, external, middle, or inner ear observed in 90% of DS children, basing on MRI or HRCT.

Conclusions: The early cochlear implantation in children with DS is a similarly useful method in treating severe to profound sensorineural hearing loss (SNHL) as in non-syndromic patients, although the development of speech skills present differently. Due to a higher prevalence of ear and temporal bone malformations, detailed diagnostic imaging should be taken into account before the CI qualification. Better postoperative outcomes may be achieved through comprehensive care from parents/guardians and speech therapists thanks to intensive and systematic rehabilitation. **Key Words:** Additional disabilities—Auditory performance—Cochlear implantation—Down syndrome—Outcomes cochlear implant—Speech perception—Speech recognition.

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Down syndrome (DS), a genetic disorder resulting from an anomaly of 21 chromosome is the most common form of mental retardation disorder, with an incidence of approximately 1 in every 800 to 1000 live births (1). Out of 200,000 individuals with DS in Europe and 2 million worldwide (2), 45 to 90% suffer from various types of hearing disorders. In contrast, hearing loss (HL) occurs in only 2.5% of general society (1,3,4) and only 9% of the

mentally-delayed population (5). More than 80% of children with DS demonstrate conductive hearing loss (6). The prevalence of sensorineural hearing loss (SNHL) ranges from 4 to 20%, depending on the adopted criteria (4,6–8). It is hypothesized that the progressive nature of SNHL in individuals with DS is conditioned by abnormalities of the cochlea, inner auditory canal, cochleovestibular nerves, or lateral semicircular canal. Some studies considered that the most common cause of SNHL in patients with DS is internal auditory canal stenosis (9).

Hearing loss in children with Down syndrome is an additional disability associated with long-term difficulties in speech and language acquisition, poor academic performance, and behavioral or social integration problems. In the case of normal hearing, speech development depends on the level of the child's intellectual impairment. Children with mild mental retardation usually develop an adequate amount of speech to drive a simple conversation with others. In cases of a severe degree of intellectual disability accompanied by hearing disorders, children are incapable of communicating, and leading an independent life becomes impossible. Moreover, the presence of autistic spectrum may delay the diagnosis of HL and disrupt the stratification of its severity. The autistic spectrum in combination with progressing HL is rated as 7% (5,10). Therefore, early implementation of adequate treatment in the hearing-impaired population with DS plays an essential role in ensuring proper language development and preventing the accumulation of disadvantages at a later age. In the time since cochlear implants (CIs) were approved by the Food and Drug Administration (FDA), treatment of severe-to-profound SNHL has become more successful. Indications for CIs are widely known, but there is no consensus about implanting them to disabled children.

The present study aimed to review long-term outcomes concerning postoperative auditory and speech performance of deaf children with DS, who were implanted at the Clarós Clinic Cochlear Implant Center in Barcelona. By presenting our experience, our team wants to demonstrate the complexities of diagnosis, the difficulties with rehabilitation, and the problems with achieving useful social integration by impaired children.

MATERIALS AND METHODS

After obtaining the necessary authorizations to guarantee patient confidentiality, a retrospective analytical study was conducted on children with DS who were implanted with multichannel cochlear implants at the Clarós Clinic in Barcelona. Records of patients eligible for cochlear implantation since 1993 were identified. Patients were divided into four categories, depending on the age at the first implantation. Postoperative results were analyzed and compared between children with Down syndrome and those without any additional disabilities—already published in Clarós et al. study (11).

Study Group

We identified 11 cases of deaf children with DS. One individual was not implanted due to parental refusal. Another

was disqualified due to bilateral absence of cochlear nerves. Therefore, the study group consisted of nine children with DS who were diagnosed with severe-to-profound hearing loss. These nine children made up 1.7% of the pediatric population recommended for CI implantation in our clinic. Various studies have proven that the age at which a patient undergoes CI surgery plays an essential role in achieving desired postoperative outcomes (12–14). Therefore, study group participants were evaluated by age categories. Due to the frequent occurrence of intellectual disorders, each study group participant was assessed by a well-qualified psychologist to evaluate his/her mental condition. The study group was then compared with the reference group described below.

Reference Group

The reference group was selected from a pediatric population comprised of 220 subjects (55.3% men), with a mean age-at-implantation of 51.4 months (SD 34.2; range, 9–167 mo). The right ear was most commonly implanted, in 84.2% patients. Deafness was congenital in 75.8% of individuals, with documented genetic etiology (e.g., connexin 26 mutation, biotinidase deficiency) in 17.5% of cases. Preoperative radiological findings included: enlarged vestibular aqueduct (3.2%), cochlear incomplete partition type 1 including cystic vestibular anomaly (3.2%), cochlear incomplete partition type 2 including Mondini dysplasia (2.7%), cochlear hypoplasia (2.3%), semicircular canal dehiscence (3.6%), and other inner ear malformations (5.9%). Due to high risk of central nervous system failure, which may negatively affect postoperative results, exclusion criteria were: gestational infections (e.g., Citomegalovirus, Herpes simplex virus, Human immunodeficiency virus, Varicella zoster virus, influenza, rubella, or toxoplasmosis), encephalitis, poliomyelitis, early-onset neurological disorders (e.g., Attention deficit hyperactivity disorder), multiple sclerosis, adrenoleukodystrophy, brain tumors, autism, IQ less than 69, and other genetic disorders.

Medical Data

Recorded data included age, sex, duration of deafness, clinical and radiological features, implant device model, unusual surgical incidents, and postoperative results. Table 1 contains detailed information about cochlear implantation.

Before implantation, all of the children had documented severe-to-profound hearing loss (HL), and had failed an appropriate hearing-aid trial (i.e., they used a hearing aid for a minimum of 3 months). Hearing disorders were investigated with otoacoustic emissions (OAEs), auditory brainstem response (ABR), or auditory steady state response (ASSR). Children presenting otitis media with effusion (OME) underwent medical treatment or surgical procedures, e.g., adenoidectomy, myringotomy, or ventilation tubes insertion, before the audiological examination (ABR, OAE, or ASSR), so as not to disturb hearing test results. Due to the reluctance of some patients, some of these tests were performed while the child was sedated or under general anesthesia. When possible, the audiological assessment was completed using behavioral and tonal audiometry with and without hearing aids (15).

Each child underwent magnetic resonance imaging (MRI) of the head, with further evaluation of the cochlea's and the internal auditory canal's (IAC) condition, with gadolinium-diethylenetriamine pentaacetic acid (DTPA) contrast, in T1, T2, and FLAIR sequences, and high-resolution computed tomography (HRCT) of the temporal bone. Both MRI and HRCT were performed during the same day, under general anesthesia, if necessary. In the

TABLE 1. Characteristics of cochlear implant recipients with DS

No	Sex ^a	Duration of CI Use (yr)	Duration of HA Use ^b (mo)	Age at CI (mo)	Ear Implanted	Brand of Cochlear Implant	Speech Processor	Number of Electrodes Activated	Intraoperative Neural Telemetry
1	M	14	80	82	Right	CI24R	ESPrIt 3G	22/22	Present
2	M	12	60	65	Right	CI24RE	ESPrIt 3G	22/22	Present
3	F	13	12	54	Left	CI24RE	ESPrIt 3G	22/22	Present
4	M	19	88	127 (I) ^c 169 (II)	Left (I) Left (II)	CII BE (I) CII BE (II)	Platinum BTE	8/8 (I) 16/16 (II)	Present (I) Present (II)
5	M	13	14	21	Left	CII BE	Platinum BTE	16/16	Present
6	F	12	50	70	Right	CI24RE	ESPrIt 3G	22/22	Present
7	F	12	13	78	Right	CI24RE	Freedom	22/22	Present
8	M	10	25	102	Right	CI24R	Freedom	21/22	Present
9	M	16	63	79 (I) 215 (II)	Right (I) Left (II)	CI24RE (I) CI24RE (II)	Freedom Freedom	21/22 (I) 22/22 (II)	Present (I) Present (II)

^aM—male, F—female.

^bDuration of hearing aids use before the cochlear implantation.

^cI—first cochlear implantation, II—second cochlear implantation. Patient No 4 underwent the re-implantation.

case of positive qualification for CI, children were vaccinated according to Center for Disease Control recommendations to prevent postoperative meningitis (16). In all patients, the cochlear implantation was performed by the same surgeon, with pre- and postoperative cardiac monitoring to control possible cardiac events that may occur during the surgery or CI mapping. Assessment of the electrode position was performed with radiograph in the operating theatre, after a telemetric measurement of the electrodes' impedance. Follow-up was done with the clinic's cochlear implantation team, psychologists, speech therapists, pediatricians, and the parents of implanted children. Psychological examination using the Wechsler scale was used to assess the study participants in terms of the degree of mental retardation: 1—mild (69–55 IQ), 2—moderate (54–35 IQ), 3—severe (34–20 IQ), 4—profound (<20 IQ).

Post-implantation Outcomes Assessment

To assess the outcomes, both groups (with DS and non-syndromic children) were divided at the first implantation into four categories, based on age: CAT1 (0–3 yr), CAT2 (4–5 yr), CAT3 (6–7 yr), and CAT4 (8–17 yr at CI surgery). The evaluation was performed with parental-validated questionnaires which contained meaningful auditory integration (MAIS)/infant-Toddler equivalent (IT-MAIS) and meaningful use of speech (MUSS) scales, as well as an audiological assessment using categories of auditory performance (CAP) and speech intelligibility rating (SIR) (13,17). The MAIS and the IT-MAIS questionnaires were used to measure auditory skills, including vocalization behavior, response to sounds, and the ability to derive the meaning of sounds. Specific scoring criteria were developed for each of 10 probes, and the overall score was obtained by adding the scores for each area. The maximum achievable result for these questionnaires was 40 points. The MUSS scale evaluates speech skills in the areas of vocal control, use of speech without gestures or signs, and communication strategies in daily situations. The maximum achievable result on the MUSS scale was 44 points. The parental evaluations of implanted children were done via phone calls or medical appointments once before the implantation, and every year after the surgery.

Finally, the same audiologist evaluated the CAP and SIR scores during medical visits before and after the CI implantation. The SIR, with scores ranging from 1 to 5, measures speech intelligibility, while the CAP is used to estimate the auditory performance (0–7 points). Interobserver reliability for both scales was formally validated (17).

Statistical Analysis

Statistical analysis was performed using R 3.5.1 - software (Free Software Foundation Inc., Boston, MA) with the nparLD package (18). All participants remained for the duration of the study, and there were no missing values. Statistical significance was reported at the alpha level of 0.05. Due to the small sample size, the results were analyzed with the nparLD package for R software, using a nonparametric test for sustainable measurements. During the calculations, we combined the CAT1 group with the CAT2 group (children implanted below the age of 6), and combined CAT3 with CAT4 (children implanted at ages 6 and above). Additional comparisons for time effects (i.e., testing if the changes stabilize at a certain point) were estimated by the Wilcoxon signed-rank test.

RESULTS

Cochlear Implantation Data

During the 25 years considered in our analysis, 935 implantations of CI were recorded, 70% of which were in children. We focused on nine children with DS who received their first multichannel cochlear implant at an average age of 75.3 months (SD 27.9, range, 21–127 mo). There were six men and three women. Five patients were implanted on the right ear, three on the left ear and one patient received implants on both ears. There were no complications during or after the CI surgeries, except for one case that required re-implantation. This was due to device failure caused by a loss of hermetic seal and corrosion of the resistive film material. Table 1 provides detailed information about the cochlear implant recipients.

In six cases, the cochlear implant surgery was performed with cortical mastoidectomy, followed by posterior tympanostomy and cochleostomy, from the postaural approach. In three cases, due to the small cavity of the middle ear and poor pneumatization of the mastoid, the canal wall down technique with temporal muscle obliteration was performed. This technique was used to avoid inadvertent damage of the facial nerve or vestibulum. There were no cardiac events during the cochlear device programming. The postoperative follow-up ranged from 13.1 to 18.3 years (mean 14.9, SD 1.5).

Radiological Evaluation

The radiological evaluation revealed more anomalies in the group with DS than in the non-syndromic population. Out of 90% of a study group, we reported the narrowed external auditory canal (EAC) with residual cerumen (40%), poor pneumatization of the mastoid (30%), small cavity of the middle ear (30%), cochlear nerve absence (unilateral 10%; bilateral 10%), enlarged vestibular aqueduct (EVA, 10%), semicircular canal dehiscence (SCCD; 10%), incomplete partition of the cochlea type 1 and 2 (IP-1 and IP-2; 20%), and narrowed internal auditory canal (IAC; 10%).

Psychological Assessment

Psychological tests conducted by a qualified psychologist showed that the children with DS included in the study had confirmed mental retardation at a mild or moderate level (defined by an IQ range of 44–69 [SD 51.4]). The assessments were conducted using the Wechsler Intelligence Scale for Children (WISC).

Audiological Assessment

Before the CI, objective tests showed no otoacoustic emissions (OAE), ASSR more than 100 dB, and no response in EABR at 95 dB. The 4-frequency average hearing level (500, 1000, 2000, 4000 Hz) of the aided hearing thresholds before the procedure was 105.6 dB (SD 6.8). The average age of hearing loss (HL) detection was 24.8 months (SD 11.1, range, 5–41 mo). The average duration of hearing aid (HA) use was 45 months (SD 28.1, range, 12–88 mo). After the CI, children with DS progressed in their auditory abilities. The postoperative average hearing level, assessed with free field audiometry, was 26.1 dB (SD 7.7, range, 20–45 dB).

Outcomes Assessment

The rehabilitation program consisted of three audiological sessions per week with the same professional therapist, and included documentation of progress. Out of all patients with DS, 89% participated in sessions regularly, and 100% were full cochlear implant users who had strong commitment and support from their parents/guardians. The post-implantation outcomes comparison between children with DS and non-syndromic group is presented in Table 2.

The CAP, SIR, and MUSS results were compared between the group with DS ($n=9$) and non-syndromic group ($n=220$). MAIS scores were unavailable for the non-syndromic children, so they were not included in the comparison. The Mann-Whitney U test was used because of the non-normal distribution of the data. Children with DS achieved significantly lower scores than the non-syndromic patients in all three tests. The results for children with DS present as follow: CAP (4.4, 95%, SD 0.9, $p=0.02$, $U=542.5$); SIR (3.2, 95%, SD 0.7, $p=0.03$, $U=579$); MUSS (60%, 95%, SD 0.1, $p=0.03$, $U=576.5$).

Next, we analyzed the results in children with DS, accounting for age at first implantation. We split the participants into two categories: 1) patients implanted before age 6 (CAT1 and CAT2; $n=4$), and 2) those implanted at ages 6+ (CAT3 and CAT4; $n=5$). We compared the results of CAP, SIR, MAIS/IT-MAIS, and MUSS scales for these two categories at five time points: before the surgery (T00), one (T01), five (T05), and 10 (T10) years after the CI, and finally at the last available follow-up (T11).

The CAP scale analysis shows significant effects of time, age group, and interaction effect ($p < 0.05$). As visible in Figure 1, the group implanted at a younger age achieved better and faster results than patients who were implanted at an older age. The Wilcoxon signed-rank test indicates that improvement stabilized around T05, so after T05, there is no significant time effect ($p > 0.05$). The SIR measurement analysis shows a significant effect of time only ($p < 0.001$). Similarly to the CAP results, patients' scores did not improve significantly after T05 ($p > 0.05$) (Fig. 2). The MAIS/IT-MAIS scale analysis shows significant effects of time, age group, and interaction effect ($p < 0.001$). The group implanted at a younger age showed a faster improvement in scores when compared with their older peers. Time effect is not significant after T10 ($p > 0.05$) (Fig. 3). The MUSS analysis shows significant effects of time and interaction ($p < 0.001$). Time effect is not significant after T10 ($p > 0.05$) (Fig. 4).

In the parental assessments, the improvement in auditory and speech skills after the CI implantation was recorded. Average results in MAIS/IT-MAIS scales were 76.9% (SD 0.1). The MUSS questionnaire application

TABLE 2. Mean results after CI regarding age at the first implantation in a group with DS and non-syndromic group

Category	CAT 1		CAT 2		CAT 3		CAT 4	
	0–3 years		4–5 years		6–7 years		8–17 years	
Age at CI	0–3 years		4–5 years		6–7 years		8–17 years	
Group size	DS ^a (n=1)	NS ^b (n=86)	DS (n=3)	NS (n=47)	DS (n=3)	NS (n=26)	DS (n=2)	NS (n=61)
CAP	6	6.2	4.7	5.4	4.3	5	3.5	4.1
SIR	4	4.3	3.3	4	3.3	3.7	2.5	2.9
MUSS (%)	68.2%	68.4%	62.9%	66.4%	59.9%	63.2%	51.1%	61.4%

^aDS—children with Down syndrome.

^bNS—non-syndromic children.

CAP indicates categories of auditory performance; MUSS, meaningful use of speech scale; SIR, speech intelligibility rating.

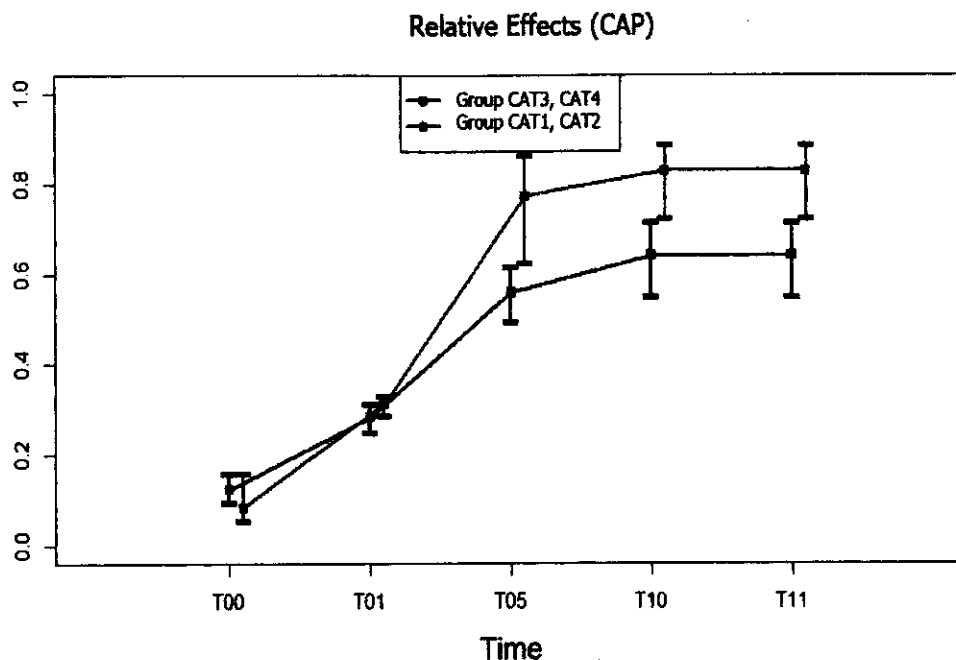


FIG. 1. Relative effects of CAP variable in children implanted before 6 years of age (CAT1, CAT2) and after 6 years of age (CAT3, CAT4). "Group CAT1, CAT2" outperformed older children in T01 in auditory performance. After 5 years from cochlear implantation (T05), the younger group obtained significantly higher results ($p < 0.05$) in CAP scores, and CAP variable changes stabilized in both groups. CAP indicates includes categories of auditory performance.

revealed a mean score of 59.8% (SD 0.1). Children with DS developed better vocalization and use of speech, became able to communicate with other people, respond to traffic signals, and even use the telephone. Abilities to

recognize and distinguish sounds were also acquired. Therefore, it could be concluded that cochlear implantation benefited deaf children with DS in general, and results corresponded with the age of implantation.

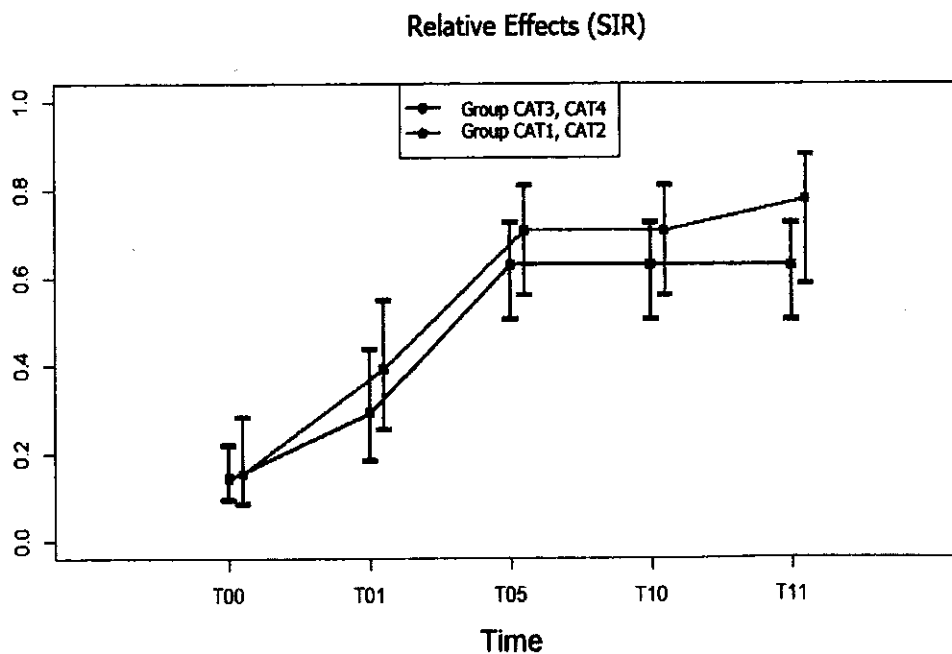


FIG. 2. Relative effects of SIR variable in children implanted before 6 years of age (CAT1, CAT2) and after 6 years of age (CAT3, CAT4). "Group CAT1, CAT2" and "Group CAT3, CAT4" were developing similarly, although younger children obtained higher scores in SIR in each measured period. The greatest improvement was recorded during the first 5 years of observation in both groups ($p < 0.001$). SIR indicates speech intelligibility rating.

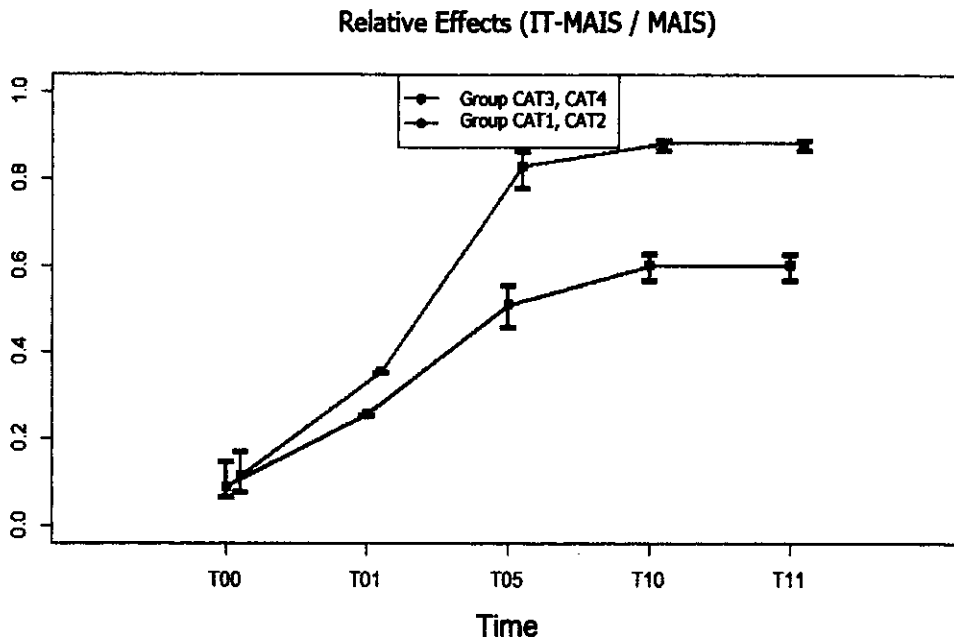


FIG. 3. Relative effects of MAIS/IT-MAIS variables in children implanted before 6 years of age (CAT1, CAT2) and after 6 years of age (CAT3, CAT4). "Group CAT1, CAT2" achieved significantly higher and faster post-implantation outcomes performance in recognizing and responding to sounds in comparison to "Group CAT3, CAT4" ($p < 0.001$). No changes in variable were observed after 10 years from the CI in both groups. CI indicates cochlear implants; MAIS/IT-MAIS, meaningful auditory integration/infant-toddler equivalent.

DISCUSSION

Cochlear implantation is approved worldwide as an effective method in the rehabilitation of deaf children (19-22), but only few studies have evaluated its efficacy in a population with DS (23-25). Therefore, our study

added to the evidence that individuals with DS may obtain a satisfactory improvement in auditory and speech skills after implantation. We reported better post-implantation outcomes in younger patients, and this phenomenon may be dependent on the brain plasticity occurring in early adolescence (26,27). Some studies documented the

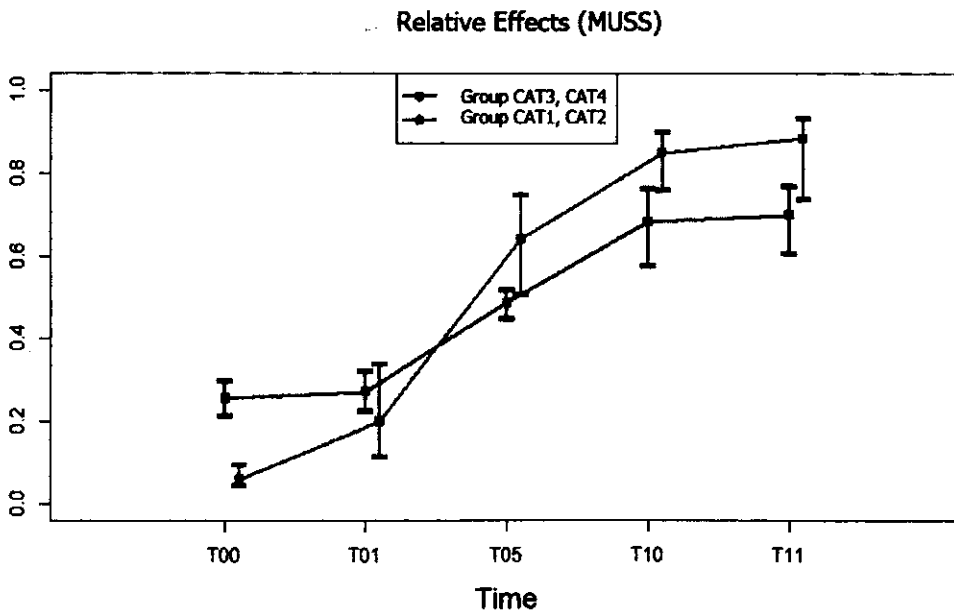


FIG. 4. Relative effects of MUSS variable in children implanted before 6 years of age (CAT1, CAT2) and after 6 years of age (CAT3, CAT4). "Group CAT1, CAT2" with the poorer speech performance at the beginning of the observation outperformed "Group CAT3, CAT4" between T01 and T05. No variable changes were observed after 10 years from implantation. MUSS indicates meaningful use of speech scale.

presence of a sensitive period, ending around 3.5 years of age, during which the central auditory pathways present maximal neural plasticity (28,29). In contrast, congenitally deaf children implanted after 7 years of age presented incomplete maturation of their electrically-evoked cortical potentials, including latencies that are longer than age-matched controls (30). This phenomenon may explain the poorer response after the CI in the patients who received implant at a later age. In a study conducted by Richter et al. (31), the age at implantation was presented as the most important prognostic factor. Additionally, Kileny et al. (32) demonstrated the influence of age at implantation and duration of CI use on speech development, and proved that children implanted between the ages of 12 and 36 months outperformed children implanted between the ages of 37 and 60 months. In our study, we observed a similar effect of age at implantation and duration of CI use on the results. We noticed that children with DS implanted under the age of 6 years old had worse results at the beginning of the study, but over time they outperformed the children implanted at a later age. This was, in particular, observed by analyzing the results of the SIR and MUSS scales. It is known that younger children with DS are characterized by a less developed use of oral skills in social life. However, after 10 years of follow-up, it can be observed that children with DS who received the CI at a younger age have acquired better communication skills, more pronounced speech, and better vocalization compared with patients who were implanted at a later age. Developmental milestones also support the beneficial effects of early implantation. The presence of a critical period for language development in the pediatric population is well documented, and the consensus is that children have the best opportunity to learn language during their first 5 years of life (32). Many studies support the concept of lowering the age criterion to qualify for cochlear implantation (28–35). Nowadays, it is recommended in many publications to perform the CI at the age of 1 year (33,34) or even earlier (35).

Extension of indications for cochlear implantation, including extremely young children, is associated with an increased risk of developing mastoiditis, acute otitis media or even intracranial infection, which may spread through the canal performed during the cochleostomy. Children with DS are more likely to develop such complications due to additional anatomical anomalies, including “glue ear” or immunodeficiency (6,36). Luntz et al. (37,38) recommended the continuous use of ventilating tubes in pediatric candidates to the CI with exudative otitis media in pediatric candidates. In our study, 40% of patients with DS underwent ventilation tubes insertion before the implantation. The implementation of proper treatment may prevent or reduce post-operative complications.

Long-term use of CI was confirmed by Geers et al. (39) to be another prognostic factor which significantly improves speech perception skills. This is consistent with

our study, where the increase in CAP, SIR, MAIS, and MUSS scores were higher when the duration of the CI use was longer. We noticed that after 5 years of follow-up, there was a significant difference between children implanted at a younger age and children implanted at an older age, in the following categories: recognizing and responding to speech sounds, demonstrating the ability to discriminate spontaneously between two speakers, and associating vocal tones. Our results may refer to the Hans et al. (23) observation of children with DS, where the advantages from cochlear implants were higher when there was more extended CI use; and to the Bakhshaei et al. (40) study, where the highest rates in the SIR scale were achieved during the first 3 years of CI use.

After a review of the literature, it can be concluded that cognitive impairment is another significant predictor of post-implantation outcomes (41). Some authors believe that mental retardation should be a contraindication for cochlear implantation, and that disabled children will not achieve satisfactory results (42). It was documented that benefits resulting from the CI are higher among deaf children without any additional disabilities, and our study also confirmed this statement. However, even though the patients with DS achieved a lower level of speech perception skills than their non-DS peers, the individuals with DS gained satisfactory results regarding socialization and interest in the environment, and achieved an overall improvement in the quality of life. Language acquisition is the result of the process of social integration, involving shared activities, surrounding stimuli or parental quality and intensity of using language (43). It is worth mentioning that typical postoperative audio-oral implementation may be insufficient for patients with DS, considering that about 17.6% of children with DS aged less than 20 years present problems with concentration, inadequate attention, hyperactivity disorder (6.1%), conduct/oppositional disorder (5.4%), or aggressive behavior (6.5%) (44). These observations may lead us to certify that implanted children with DS should receive more intense and comprehensive rehabilitation. Undoubtedly, postoperative care over implanted patients with DS is a challenge for both parents and speech-therapists.

Another challenge is choosing an appropriate surgical technique for various anatomical anomalies, to avoid unnecessary complications (45). Some studies highlighted the necessity of a detailed radiological examination in patients with genetic mutations, due to a higher risk of unexpected anatomical malformations (9,46). The temporal bone study of patients with Down syndrome conducted by Bilgin et al. (46) revealed the most common abnormalities of the inner ear, such as: malformed bone islands of lateral semi-circular canal (LSCC), narrow internal auditory canals (IACs), cochlear nerve canal stenoses, semicircular canal dehiscence (SCCD), and enlarged vestibular aqueduct (EVA). Intrapiromkul et al. (9) also confirmed the malformed (<3 mm) bone

island of LSCC to be the most common inner ear deformation. These results are comparable to our research, in which 90% of children with DS presented similar otological abnormalities. Taking into account our experience and the results from presented studies, both MRI and HRCT of temporal bone should be obligatorily performed before the cochlear implantation.

CONCLUSIONS

Cochlear implantation is an advantageous procedure for deaf children with Down syndrome. Despite some difficulties this method significantly increases the ability to understand and articulate speech, especially when patients are implanted in their early childhood. Therefore, we should be particularly thorough when diagnosing hearing disorders, and detect them as early as possible to implement appropriate treatment. Due to the frequent occurrence of ear malformations, imaging tests should be thoroughly analyzed to avoid complications and unsuccessful implementation of the CI device. Hearing-impaired children with DS need unique and comprehensive care before implantation, as well as further rehabilitation after implantation, to obtain beneficial results. In our opinion, various disabilities related to DS should not be considered as a contraindication for cochlear implantation, but a challenge to be addressed by the entire cochlear team.

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