Case Report

Indication process of cochlear implant for a child with cerebral palsy: a case report

Processo de indicação do implante coclear em uma criança com paralisia cerebral: estudo de caso

Maria Jaquelini Dias dos Santos[1], Maria Cecília Bevilacqua[2], Adriane Lima Mortari Moret[2], Dionísia Aparecida Cusin Lamônica[2], Orozimbo Alves Costa[2], Elisabete Honda Yamaguti[3]

ABSTRACT

Cerebral palsy (CP) is the most common motor disorder in childhood, and is usually associated with other disabilities, including hearing impairment. Studies regarding the indication of cochlear implant (CI) for children with cerebral palsy and hearing impairment are restricted internationally, and, in Brazil, non-existent. Therefore, the purpose of this study was to describe the process of pre-surgical assessment of a child with CP who was a candidate for cochlear implantation. The study was conducted with a boy, 2 years and 6 months old, with CP and bilateral hearing impairment. The pre-surgical protocol followed the procedures already used at the Hospital for Rehabilitation of Craniofacial Anomalies, including: speech-language pathology and audiological, neurological, otorhinolaryngological, and psychological evaluations through clinical observations, questionnaires, and imaging findings. The patient was diagnosed with profound and bilateral auditory neuropathy spectrum disorder, moderate quadriparesis athetoid, global developmental delay (except in visual function), presence of intentional communicative behavior, good social contact, absence of intellectual impairments or other additional disabilities within the clinical framework of CP, and a family integrated to the treatment. Based on the results obtained, the CI was recommended to this patient, who is currently enrolled in a systematic monitoring program. This study highlights the importance of the pre-surgical protocol for children with CP who are candidates for CI. Through assessment tools directed to the global aspects of development, one can obtain specific information that improve parent counseling regarding the child's prognosis and make it possible to trace actual hearing rehabilitation goals.

Keywords: Cochlear implant, Cerebral Palsy, Hearing Loss, Child, Assessment, Developmental disabilities.

INTRODUCTION

Cerebral palsy (CP) is defined as sequela of a non evolutionary etiological based cerebral injury, with varying signs and symptoms. It is characterized by being the most common motor disorder in childhood[1-3] and it may present associated changes in different functions: visual, mental, emotional and hearing[1]. At least 12% of children with CP present sensorineural hearing loss[2].

Internationally, the cochlear implant (CI) has been highlighted by its potential for habilitation and rehabilitation of both auditory skills and oral language[4-6]. For children only suffering from hearing loss, several studies in national and international literature report excellent results. However, for children with additional disabilities with hearing loss, especially CP, indication for CI use are still little studied[2,3,5,7,8].

Several aspects are weighed when in addition to hearing loss, the child presents other conditions. Each disability in addition to deafness will feature a distinguished clinical picture, which will influence from the diagnostic conduct up to the rehabilitation of the hearing impairment. The hearing results will depend, among other factors, on the child's potential in relation to their overall development.

International studies on the CI in children with CP are restricted[2,3,5,7,8], and at the national level, these are non-existent. Thus, the purpose of this study is to describe the process of pre-surgical assessment of a child with CP, candidate for the CI.

Study conducted at the Audiological Research Center, Hospital for Rehabilitation of Craniofacial Anomalies, Universidade de São Paulo – USP – Bauru (SP), Brazil.
(1) Graduate Program (Masters degree) in Speech-Language Pathology and Audiology, Department of Speech-Language Pathology and Audiology, Bauru School of Dentistry, Universidade de São Paulo – USP – Bauru (SP), Brazil.
(2) Department of Speech-Language Pathology and Audiology, Bauru School of Dentistry, Universidade de São Paulo – USP – Bauru (SP), Brazil.
(3) Audiological Research Center, Hospital for Rehabilitation of Craniofacial Anomalies, Universidade de São Paulo – USP – Bauru (SP), Brazil.
Correspondence address: Maria Jaquelini Dias dos Santos. R. Sílvio Marchione, 320, Vila Universitária, Bauru (SP), Brasil, CEP: 17012-900. E-mail: mariajaquelini@usp.com
Received: 10/1/2010; **Accepted:** 3/31/2011

CASE REPORT PRESENTATION

This study was approved by the Research Ethics Committee of the Hospital for Rehabilitation of Craniofacial Anomalies of the Universidade de São Paulo (USP), process number 041/2008, with grants from the National Council for Scientific and Technological Development (CNPq), process number 116125/2008 0.

The study was conducted in the Audiological Research Center of the Hospital for Rehabilitation of Craniofacial Anomalies. To begin the assessment process, the child's guardian signed the Term of Informed Consent, following Resolution MS/CNS/CNEP 196/96 of October 10th, 1996.

Characterization of the patient in the study

Boy, 2 years and 6 months old at the time of pre-surgical assessments. He was born preterm at 34 weeks, with a history of anoxia, jaundice, use of ototoxic drugs, and a stay in the Intensive Care Unit (ICU) for 15 days.

At about 10 months of age the patient was diagnosed with profound bilateral hearing impairment and it was possible to observe a delay in psychomotor development, with suspected cerebral palsy (CP). The audiological intervention occurred at 18 months of age, using a hearing aids, and systematic monitoring in auditory rehabilitation program, with a focus on oral communication in their city of origin.

The patient joined the hospital's cochlear implant program at 2 years of age, already adapted to the hearing aids and placed into a motor rehabilitation program since 12 months of age. He was also enrolled as a student at a regular infant school.

Pre-surgical protocol for evaluation of children with cerebral palsy

The institutional criteria used for the indication of CI were considered for this study[4]. The pre-surgical assessment was performed by an interdisciplinary team consisting of audiologists, physicians, psychologists and social workers. The audiological and language assessment included the following procedures: Visual Reinforcement Audiometry (VRA), Speech Detection Threshold (SDT), Impedanciometry (I), Transient and Distortion Product Evoked Otoacoustic Emissions (T/DP EOE), Brainstem Evoked Response Audiometry (BERA), Auditory Steady-State Response (ASSR); and implementation of the following questionnaires and scales: Infant Toddler Meaningful Auditory Infant Scale (IT-MAIS)[10], Meaningful Useful Speech Scale (MUSS)[6], Production Infant Scale Assessment (PRISE)[11], MacArthur Inventory: first words and gestures[12], Gesell and Amatruda Scale[13], Early Language Milestone Scale (ELM)[14]. After these procedures, hearing categories[4], Expressive Language[4], and classification of the predominant mode of communication[3] were assigned.

The medical assessments comprised both otolaryngology and neurology areas. Clinical and imaging examinations were held: computerized scan (CT) of the mastoid, magnetic resonance imaging (MRI) of the skull, posterior fossa and temporal regions.

The psychology team assessed the patient and his family. Combined with the structured psychological interview, classification in regards to the Family's Permeability Level and the Child's Cognitive Style were used[4].

RESULTS

The analysis of audiological findings in VRA, TE/DPOE, BAEP, ASSR, and impedanciometry (Table 1), indicated profound bilateral sensorioneural hearing loss, characterizing as auditory neuropathy spectrum disorder. Combined with the audiological results, the findings on MRI of the posterior fossa and temporal regions dismissed hypoplasia or agenesis of the auditory nerve. Such conditions if present, are contraindications to the CI.

Hearing loss presented by the participant in the study is not considered a contraindication to CI. The presence of profound or severe bilateral sensorineural hearing loss is one of the conditions for the individual to be a candidate for the CI.

In VRA performed with warble stimulus in free field, there was no significant difference between the thresholds with and without hearing aids (Table 1). The gain of 25 dB for the right ear and 20 dB for the left ear using the hearing aids was not sufficient to achieve the required hearing thresholds for the perception of speech sounds. The same was observed for speech detection threshold (SDT), in which the gain was equal in both ears: 30 dB (Table 1).

In addition to the gain in decibels, the communicative behavior of the individual to the world of sound with the hearing aids should be considered. In this study, the participant showed no significant change to the communicative behavior while using the hearing aids so that it would be kept as a treatment option for hearing impairment in question. However, it is important to highlight that the participant has made effective use of hearing aids since the age of 10 months, so he had the opportunity to receive auditory stimulation for about 20 months before receiving the CI.

Early auditory stimulation through a hearing aids, which happens before the CI referral, favors the process of maturation of the auditory system structures, because it sends sound information to a neural system in training. This information is stored and shall constitute the mnemonic repertoire of the patient to the sound world, which is nonetheless an important factor for the development of auditory skills, language and speech after the CI surgery and, consequently an important characteristic for the CI indication.

Criteria considered a contraindication to the CI[9] were not observed in this patient; they are: medical conditions which contraindicate the surgery; hearing impairment caused by agenesis of the cochlea, auditory nerve, or by central lesions; active infection of the middle ear, serious neurological conditions associated with hearing impairment.

In this study, although the PC was considered a neurological entity, the same was not considered an impediment to the use of CI, because the affected area, in this particular case, refers to the motor area.

The neurological assessment rated the PC as moderate quadriparesis athetoid with greater motor loss on the left side and

upper limbs. The neurological examination and psychological assessment ruled out cognitive and psychopathological conditions in association with CP that could interfere in the process of enabling the CI. Thus, it is believed that the use of CI can optimize receptive skills and promote integration of auditory stimuli, enabling the development of communication skills.

Thus, the results of the pre-surgical assessment were analyzed by the team, and considered favorable for the indication of CI surgery (Tables 1 and 2).

The indication of the CI was discussed with the patient's family and, after the family agreed with the surgery, the patient received the CI brand Advanced Bionics model HiRes 90k, with total insertion of electrodes in the left cochlea . The patient's age at surgery was 2 years and 7 months, the activation of the cochlear implant was performed one month after surgery and follow-up is being carried out systematically in the hospital.

DISCUSSION

Medical, audiological and language assessment in order to define the type and degree of hearing loss in this child differ

Table 1. Results of the pre-surgical assessment used for the indication of CI in children with cerebral palsy: hearing and language aspects

Evaluations	Results	
Audiological assessments	RE	LE
VRA (free field)		
Without hearing aids	95 dB	95 dB
With hearing aids	70 dB	75 dB
SDT (free field)		
Without hearing aids	85 dB	90 dB
With hearing aids	55 dB	60 dB
Tympanometry	Curve A – normal	Curve A – normal
Stapedial reflex	Absent	Absent
EOE		
Transient	Absent	Absent
Distortion product	Present	Present
BERA	Absent with cochlear microphonic	Absent with cochlear microphonic
ASSR	Present in 110 dB	Present in 110 dB
Questionnaires		
IT-MAIS	40%	
MUSS	27.5%	
PRISE	59%	
MacArthur Inventory	Area of dominance: actions and gestures	
Scales / Categories		
ELM Scale		
Receptive hearing function	38.5%*	
Receptive auditory function	33.4%*	
Visual function	100%*	
Gesell and Amatruda's scale		
Communicative behavior	18 months	
Gross motor behavior	12 months	
Fine motor behavior	10 months	
Language behavior	6 months	
Personal-social behavior	14 months	
Auditory category	0[1]	
Expressive language category	1[2]	
Mode of communication	2[3]	

* Quantitative analysis of items presented by the child in each function

Note: RE = right ear; LE = left ear; dB = decibel; VRA = visual reinforcement audiometry; VDT = verbal detection threshold; EOE = evoked otoacoustic emissions; BERA = Brainstem Evoked Response Audiometry; ASSR = Auditory steady-state response; [1] = does not detect speech; [2] = absence of speech or undifferentiated vocalizations; [3] = communication through gestures

Table 2. Pre-surgical assessment results used for indication of the cochlear implant in children with cerebral palsy: assessments by imaging, psychological and medical aspects

Assessments	Results
Psychological assessment	
Psychological interview	Absence of psychopathology in child and family
Family's permeability level	96.4%
Child's cognitive style	100%*
Medical clinical assessments	
Neurology	Without additional disabilities with cerebral palsy
	Moderate athetoid quadriparesis, greater motor loss on the left side
Otorhinolaryngology	No contraindications to the surgery
	Bilateral profound auditory neuropathy spectrum
Assessments by imaging	
Brain MRI	Increased bilateral frontal subarachnoid space and absence of ischemic lesions
CT of the mastoids	No abnormalities
RM of fossa and posterior temporal regions	No abnormalities

Note: CP = cerebral palsy; CI = cochlear implant; MRI = magnetic resonance imaging; CT = computerized tomography

from the existing in the standard protocol, due to the peculiarity of the evaluated child in having cerebral palsy. The battery of tests and procedures was crucial to dismissing central hearing loss. Central hearing loss is considered a contraindication to CI. In cases of extensive brain lesions, other cortical regions, in addition to the motor centers, can be affected. Thus, the imaging studies (MRI/CT) were considered a valuable and essential aid to the audiological diagnosis.

The use of Gesell and Amatruda's scale[13] and the ELM Scale[14] indicated to us the patient's state of development in other areas other than auditory and linguistic skills. In this case, finding global development delay, consistent with cerebral palsy, and lowered scores on the questionnaires was expected, which indeed was observed (Table 1). But in the field of visual function did the patient show no delay compared to his chronological age. In the PRISE questionnaire[11], which refers to the vocalizations and communication skills, the patient score above 50%, reaffirming its communicative potential.

Importantly, a group of children with CP and hearing loss will always be considered a heterogeneous group. The only similarity lies in the fact that they suffer from hearing loss and are candidates for the CI. In contrast, other characteristics of the motor and cognitive areas are distinct, which may present potential variables in the assessment process and, consequently, habilitation and rehabilitation with the CI. For this reason, this study reinforces the need to have a child with CP fully evaluate by an interdisciplinary team, considering the individual aspects of development of each child. In this study, the participant showed favorable characteristics to the indication of the CI.

Most international research encompassed children with different associated disabilities in the case study sample, including CP as one of them[2,3,5,7,8]. Only one research dealt exclusively with cochlear implants in children with cerebral palsy[2]. This study evaluated the perception and speech intelligibility of these children after a certain period of CI use. However, regarding the pre-surgical assessment process, there is no information.

Studies about cochlear implant in this population of children with CP, or with other additional disabilities with hearing loss, focus on the analysis of the benefits observed after the use of CI, in relation to the perception and production of speech. The assessment process prior to the indication of the CI is not studied in depth. Thus, it is possible to conclude that the assessments and criteria used for selection of children with multiple disabilities candidates for cochlear implants did not differ greatly from the protocols and criteria for children only suffering from hearing loss in different centers where the studies were conducted .

The criteria for the indication of CI for children with CP in this study were indistinguishable from the one already used for children only suffering from hearing impairment[9]. However, the process of pre-surgical assessment should present instruments directed to the global aspects of development and specific to each deficiency associated with hearing loss, in this case CP. This will be instrumental in establishing prognosis, as well as to conducting the process of guidance and family counseling, especially in regards to family expectations.

For children who have cerebral palsy or other additional disabilities with hearing loss, the pre-surgical performance on the different functional areas of development is valuable and necessary. It can simplify the obtaining of measures to quantify and qualify the progress after CI use, and probably generate discussion beyond the benefit of hearing and speaking for children with CP, in particular in regards to the quality of life.

FINAL COMMENTS

In this case study, CP was not considered a contraindication for the CI. The case study described all the necessary criteria for CI implantation. However, it is important to highlight the importance of an interdisciplinary team for the identification of the general state of health, as well as global levels of development of the child who is a candidate to the CI presented

in association with cerebral palsy or any other additional disability.

Directed and broad assessments during the process of indication of CI for this group of children assist in the decision of indicating or not indicating of CI, in addition to bringing useful information for therapeutic planning and fundamentally to enrich the guidance given to parents about the child's prognosis.

ACKNOWLEDGEMENTS

We would like to thank the National Council for Scientific and Technological Development (Conselho Nacional de Desenvolvimento Científico e Tecnológico – CNPq), for the Scientific Initiation scholarship granted for this research, under process number 116125/2008 0.

RESUMO

A paralisia cerebral (PC) é o transtorno motor mais frequente na infância, podendo apresentar outras deficiências associadas, inclusive a deficiência auditiva. Estudos sobre a indicação do implante coclear (IC) em crianças com PC e deficiência auditiva são restritos internacionalmente, e no Brasil, inexistentes. Desta maneira, o objetivo do estudo foi descrever o processo de avaliação pré-cirúrgica de uma criança com PC candidata ao IC. O estudo foi realizado com um menino, com 2 anos e 6 meses de idade, com PC e deficiência auditiva bilateral. O protocolo pré-cirúrgico seguiu os procedimentos já utilizados no Hospital de Reabilitação de Anomalias Craniofaciais, compreendendo: avaliações fonoaudiológicas, neurológica, otorrinolaringológica e psicológica, por meio de observações clinicas, questionários, e exames radiológicos. O paciente foi diagnosticado com espectro da neuropatia auditiva de grau profundo bilateral, quadriparesia atetóide de grau moderado, atraso global do desenvolvimento, exceto na função visual, comportamento comunicativo intencional presente, bom contato social, ausência de comprometimentos intelectuais ou outras deficiências associadas ao quadro da PC e família integrada ao tratamento. De acordo com os resultados obtidos, o paciente foi considerado apto para cirurgia de IC, e permanece em acompanhamento sistemático no respectivo programa. Ressalta-se a importância de uma etapa pré-cirúrgica ampla e estruturada para crianças com paralisia cerebral e candidatas ao IC. Por meio de instrumentos de avaliação direcionados aos aspectos globais do desenvolvimento, é possível obter informações específicas que enriquecem as orientações dadas aos pais sobre o prognóstico da criança e possibilitam traçar metas reais de reabilitação auditiva.

Descritores: Implante coclear; Paralisia cerebral; Perda auditiva; Criança; Avaliação, Deficiências do desenvolvimento

REFERENCES

1. Lamônica DA. Estimulação da linguagem em crianças com paralisia cerebral. São Paulo: Pulso; 2008. Estimulação da linguagem: aspectos teóricos e práticos; p. 163-77.
2. Bacciu A, Pasanisi E, Vincenti V, Ormitti F, Di Lella F, Guida M, et al. Cochlear implantation in children with cerebral palsy. A preliminary report. Int J Pediatr Otorhinolaryngol. 2009;73(5):717-21.
3. Berrettini S, Forli F, Genovese E, Santarelli R, Arslan E, Chilosi AM, et al. Cochlear implantation in deaf children with associated disabilities: challenges and outcomes. Int J Audiol. 2008;47(4):199-208.
4. Moret AL, Bevilacqua MC, Costa OC. Implante coclear: audição e linguagem em crianças deficientes auditivas pré-linguais. Pro-Fono. 2007;19(3):295-304.
5. Wiley S, Jahnke M, Meinzen-Derr J, Choo D. Perceived qualitative benefits of cochlear implants in children with multi-handicaps. Int J Pediatr Otorhinolaryngol. 2005;69(6):791-8.
6. Nascimento LT. Uma proposta de avaliação da linguagem oral [monografia]. Bauru: Universidade de São Paulo, Hospital de Reabilitação de Anomalias Craniofaciais; 1997.
7. Daneshi A, Hassanzadeh S. Cochlear implantation in prelingually deaf persons with additional disability. J Laryngol Otol. 2007;121(7):635-8.
8. Waltzman SB, Scalchunes V, Cohen NL. Performance of multiply handicapped children using cochlear implants. Am J Otol. 2000;21(3):329-35.
9. Bevilacqua MC, Moret AL. Reabilitação e implante coclear. In: Lopes Filho O. Tratado de fonoaudiologia. Ribeirão Preto: Tecmedd; 2005. p. 423-36.
10. Castiquini EA, Bevilcqua MC. Escala de integração auditiva significativa: procedimento adaptado para a avaliação da percepção da fala. Rev Soc Bras Fonoaudiol. 2000;4(6):51-60.
11. Kishon-Rabin L, Taitelbaum-Swead R, Ezrati-Vinacour R, Hildesheimer M. Prelexical vocalization in normal hearing and hearing-impaired infants before and after cochlear implantation and its relation to early auditory skills. Ear Hear. 2005;26(4 Suppl):17S–29S.
12. Padovani CM, Teixeira ER. Using the MacArthur Comunicative Development Inventories (CDI'S) to assess the lexical development of cochlear implanted children. Pro-Fono. 2004;16(2):217-24.
13. Gesell A. Gesell e Amatruda: diagnóstico do desenvolvimento: avaliação e tratamento do desenvolvimento neuropsicológico do lactente e da criança pequena, o normal e o patológico. 3a ed. São Paulo: Atheneu; 2000.
14. Coplan J. Early Language Milestone Scale. Austin: Pro-Ed; 1993.