filodiritto
APPROFONDISCI SEMPRE
Tribunale Bologna 24.07.2007, n.7770 - ISSN 2239-7752
Direttore responsabile: Antonio Zama

Management of Congenital Deafness in Children

20 Febbraio 2020

Cristian Martu, Roxana Serban, Alexandra Lungu, Bogdan Cobzeanu, Ioan Piftor, Lorenza Donea, Cristian Alexe

Abstract

Congenital and pre-lingual deafness has a significant impact on language acquisition and the educational pr ocess in children. The rehabilitation process must begin as soon as possible.

Restoration of profound sensorineural hearing loss is possible using cochlear implants which are recognize d worldwide as the standard treatment. Cochlear implantation in congenital deafness is the best when is perf ormed as early as possible. Late implantation in prelingual deafness does not lead to speech understanding a nd includes difficulties in auditory tasks. Brain plasticity seems to have an important role in language deve lopment in relation to hearing deprivation. To obtain a near to normal development of the congenital deaf c hild early intervention is mandatory and this is possible when coherent health policies exist for diagnosis a nd appropriate treatment. This can have a major impact on the development and social integration of the chi ld.

Table of Contents:

1. Introduction

> **Deafness is a serious sensory deficiency. Congenital and pre-ling ual deafness has important and unfavourable consequences on the psychological maturation of the child, on the evolution of lan guage acquisition, on the educational process as a whole and as w ell as on his/her subsequent social insertion [1, 2, 3]. Having a dec isive importance, early identification of hearing loss [4] will al low early rehabilitation treatments which consist in hearing aids recommendation [5] and inclusion of the child in speech rehabilit ation programs.**

Currently, in our country, the average age at which hearing impairment is diagnosed is usually after 4 years. **The goal of the rehabilitation process is to begin the treatment of deaf a child well before the age of 2 years and, in short term, it is desirable to be able to start this rehabilitative program before the age of 6 months** [6]. For this, it is necessary first of all to initiate an early detection program for deaf children – possibly by introducing a universal new born screening program nationwide.
To understand the need for early treatment of hearing loss, we will briefly discuss some basic notions of ana tomy and physiology that impose the particularities of the treatment of hearing loss in the child.

2. Anatomical factors

Anatomical factors have a huge implication in ear surgery in children that might benefit from implantable h earing aids. **The phylogenetic importance of the ear is demonstrated by the precocity of the appearanc e and the complexity of its development**. Middle ear: the tubo-timpanic recess from the middle ear deve lops from an endodermic diverticulum of the dorsal segment of the first pouch. The embryonic ossicles dev elop from the arches I and II (the maleus – from Meckel cartilage and the stapes and part of the incus are fo rmed from Reichert cartilage).

> **The musculo-ligament apparatus is developed from the mesench yme of the first and second arch. Inner ear has a double origin: ectodermal for the membranous labyrinth and mesodermal for the bone labyrinth. In the 4th week, from the ectoderm and the lateral part of the rombencephalus, the otic plate is formed from which the otic vesicle will develop.**

Neurosensory epithelium is formed under the induction of neuroblasts in the Scarpa and Corti nuc leus, activated by neural crests. The inner ear and the auditory nerve are functional from the 4th month of life. The auditory nucleus of the cerebral cortex is functional from the 7th month of intrauterine life. Te mporal bone grows continuously until the age of 18-20 years, but the most significant changes in the deve lopment, number and configuration of mastoid cells occur until the age of 2 years.

3. Functional factors

The external and the middle ear conduct the sound to the cochlea. **Lesions at this level produce conducti ve deafness** (9.55% of 1/1000 children with severe bilateral hearing loss) [7].

This type of hearing loss may benefit from surgical treatment (transtympanic tubes, tympanoplasties or implantable or conventional hearing aids). The inner ear (hair cells) – transforms the mechanical energy int o electrical impulses. The lesions at this level produce sensory hearing loss (approximately 2% of congenita l profound deafness cases are due to cochlear nerve deficiency) [8]. Sensorineural hearing loss is treated wit h hearing aids or cochlear implants and speech rehabilitation.

4. Brain plasticity and neural competition

Plasticity is the ability of the brain to organize itself according to the type of received stimuli. **Brain is the organ of behavior, the most important aspect of behavior being the learning process** (change in behavior as a result of experience).

> **The brain is responsable for our ability to hear and also for our a bility to acquire language, social communication, appreciation of music, etc. Brain development is achieved progressively, with pre cise temporal sequences characteristic of each neural entity and e ach system, through neuronal connections in the brain and ne uronal differentiation.**

In recent years, many studies have brought to light a series of notions that have improved our unde rstanding of brain physiology, including: brain plasticity and neuronal competition. In children, audito ry deprivation leads to delayed or abnormal development of the central auditory pathways, particularly if de privation occurs during a sensitive period, or an established time window of approximately 3.5 years durin g which alterations in sensory input (e.g. deafness or hearing loss) can lead to profound and long-term imp acts on the brain. [9, 10]. **If auditory deprivation continues beyond the sensitive period, the connectivit y within the auditory system is disrupted resulting in significant deficits in brain and behavior** (e.g. working memory, executive functioning etc.) [11]. Cross-modal re-organization by vision is one form of the changes that take place in cortico-cortico connectivity and was observed in congenital or pre- lingua l onset of deafness. [12, 13]

Restoration of profound hearing loss is possible using cochlear implants both in acquired [14] and congenita l deafness.

> **Cochlear implants are recognized worldwide as the standard trea tment for profound hearing loss. It is important, during the surge ry, to preserve as much as possible the delicate structures of the c ochleea for the future therapies [15, 16].**

The outcome of implantation in congenital deafness is the best when the cochlear implantation is performe d as early as possible, ideally in the first 18 month of life [17].

Clinical outcomes differentiate early and late implantations with regard to auditory performance. Ver y late implantation (in teen ages) in prelingually deaf does not lead to speech understanding and includes dif ficulties in more complex auditory tasks. Even after years of experience, performance is influenced by impl antation age [18]. Early implanted children perform significantly better than later-implanted children. Late i mplanted subjects will not develop the ability to differentiate speech but they will still benefit from cochlea r implant by the awareness of sound.

The fore-mentioned theories have been proved both in animals and in humans. **Immediately afte r birth, the printing period begins. Hearing deprivation was experienced on birds and rare cases of human without speech exposure** – for example, Aveyron boy case study (a child who was captured by hunters after surviving about 12-13 years in a wolf-family and although he had a normal hearing the lack of exposure to speech made him, mental and psychological, equivalent of someone born deaf) [19] suggest that early childhood exposure to speech results in better language development.

The psycho-social integration of the hearing-impaired child therefore necessitates the reintroduction of the individual into the sound environment. Despite an early intervention – before developmental pe riod, the brain changes induced by complete absence of hearing from birth are difficult to totaly reverse de spite years-long sensory experience following therapy of the defect [20]. To obtain a near to normal develop ment of the congenital deaf child early intervention is mandatory. This is possible where coherent health pol icies exist: universal hearing loss screening, comprehensive genetic tests followed by genetic counseling wh enever necessary, early diagnosis and treatment (conventional hearing aids or/and cochlear implants) – up to age of 9 months, speech therapy, follow-up of the child. An important factor that contributes negatively to the early cochlear implantation of children is parental refusal [21, 22].

5. Management of congenital profound deaf child

There are some general principles in management of congenital profound deaf child:

1. The first principle is that of an early diagnosis. Ideally the congenital hearing loss has to be diagnosed a t birth. During the first few months of life there are several factors to consider: narrow external auditory can al, behavior, cyrcadian rhythm, audiometric curve inaccuracy (after 6 months of age the association of beha vioral audiometry with auditory evoked potentials allows a more accurate diagnosis).

2. Using a battery of audiological tests. They can establish a precise diagnosis for each ear.

Clinical evaluation, evoked auditory potentials, auditory steady state response, behavioral audiometry are required [22].

3. Early treatment - consisting in conventional hearing aids or/and cochlear implants.

4. Hearing rehabilitation must be associated with speech ththerapy.

5. Continuous monitoring of the child and his/her family by the cochlear implant team.

6. Conclusions

> **In conclusion it is important to emphazise the huge impact of ear ly diagnosis and treatment (best before the first year of life) of c ongenitaly deaf child on his/her language development as well a s on academic and social future life. A coherent health policy is m andatory to obtain this goal with an immense impact on the enti re life of the children.**

Contributo selezionato da Filodiritto tra quelli pubblicati nei Proceedings “National ENT, Head and Neck Surgery Conference - 2018”
Per acquistare i Proceedings clicca qui.

Contribution selected by Filodiritto among those published in the Proceedings “National ENT, Head and Neck Surgery Conference - 2018”
To buy the Proceedings click here.

REFERENCES

1. Zaidman-Zait, A., Dotan, A. (2017). Everyday Stressors in Deaf and Hard of Hearing Adolescents: Th e Role of Coping and Pragmatics. J Deaf Stud Deaf Educ 22(3), pp. 257-268.
2. Schick, B., Skalicky, A., Edwards,T., *et al*., (2013). School placement and perceived quality of life in y outh who are deaf or hard of hearing . J Deaf Stud Deaf Educ 18(1), pp. 47-61.
3. Punch, R. (2016). Employment and Adults Who Are Deaf or Hard of Hearing: Current Status and Expe riences of Barriers, Accommodations, and Stress in the Workplace. Am Ann Deaf. Summer 161(3), pp. 384 -97.
4. Downs, M.P., Yoshinaga-Itano, C. (1999). The efficacy of early identification and intervention for chil dren with hearing impairment. Pediatric Clinics of North America 46(1), pp. 79-87.
5. Morgon, A.H. (1998). Appareillage de l’enfant sourd porteur d’une surdite de perception-Suppleance i nstrumentale de la surdite: les aides auditives, pp. 203-216.
6. Netten, A.P., Rieffe, C., Theunissen, S.C., *et al*., (2015). Early identification: Language skills and so cialfunctioning in deaf and hard of hearing preschool children. DECIBEL Collaborative study group. Int J Pediatr Otorhinolaryngol 79(12), pp. 2221-6.
7. Stewart, J.M., Downs, M.P. (1993). Congenital conductive hearing loss: the need for early identificatio n and intervention. Pediatrics 91(2), pp. 355-9.
8. Freeman, S.R., Sennaroglu, L. (2018). Management of Cochlear Nerve Hypoplasia and Aplasia. Adv Otorhinolaryngol 81, pp. 81-92.
9. Kral, A., Sharma, A. (2012). Developmental neuroplasticity after cochlear implantation. Trends Neuros ci 35(2), pp. 111-22.

10. Sharma, A., Nash, A., Dorman, M. (2009). Cortical development, plasticity and reorganization in childr en with cochlear implants. J. Commun. Disord 42, pp. 272-279.
11. Kral, A., Kronenberger, W.G., Pisoni, D.B., *et al*., (2016). Neurocognitive factors in sensory restoratio n of early deafness: a connectome model. Lancet Neurol 15(6), pp. 610-21.
12. Neville, H.J., Lawson, D. (1987). Attention to central and peripheral visual space in a movement detecti on task. III. Separate effects of auditory deprivation and acquisition of a visual language. Brain Res 405(2), pp. 284-94.
13. Lomber, S.G., Meredith, M.A., Kral, A. (2010). Cross-modal plasticity in specific auditory cortices und erlies visual compensations in the deaf. Nat. Neurosci. 13, pp. 1421-1427.
14. Cavaleriu, B., Martu, D., Hritcu, L., Manolache, O., Radulescu, L. (2015). Idiopathic sudden hearing lo ss: Oxidative status before and after corticoid treatment. January Archives of Biological Sciences 67, pp. 106-106.
15. FDA-Regulated Study of Cord Blood Stem Cells to Treat Acquired Hearing Loss Launches www.cordb lood.com/hearingloss (accesed 15.07.2018)
16. Martu, C., Georgescu M.G., Martu, I., *et al*., (2016). Utility of Drug Loaded Nanoparticles in the Treat ment of Inner Ear Pathology. Materiale Plastice 53 (2), pp. 321-325.
17. Vavatzanidis, N.K., Mu?rbe, D., Friederici, A., *et al*., (2018). Establishing a mental lexicon with coch lear implants: an ERP study with young children Scientific Reports. Nature 8, p. 910.
18. Geers, A.E., Sedey, A.L. (2011). Language and verbal reasoning skills in adolescents with 10 or more y ears of cochlear implant experience. Ear Hear 32, pp. 39S-48S.
19. Cayea, W. (2006) Feral child: the legacy of the wild boy of Aveyron in the domains of language acquis ition and deaf education. Master Thesis submitted to National technical Institute for the Deaf in Rochester Institute of Technology. Rochester, New York accessedonline15.07.2018 http://scholarworks.rit.edu/cgi/vie wcontent.cgi?article=5163&context=theses.
20. Kral, A. (2013). Neuroscience Auditory critical periods: A review from system's perspective. Ne uroscience 247, pp. 117-133.
21. Radulescu, L., Martu, D. (2007). Do we need an ethics committee in order to make decisions regarding the cochlear implant? Revista Romana de Bioetica 5(2): pp. 27-32.
22. Byrd, S., Shuman, A.G., Kileny, S., *et al*., (2011). The right not to hear: the ethics of parental refusal o f hearing rehabilitation. Laryngoscope. 121(8), pp. 1800-4.
23. Courtat, P., Peytral, C., Elbaz, P. (1994). Exploration fonctionnell en ORL, Paris, Masson, pp. 31 -56.

TAG: *childhood, Congenital Deafness, proceedings*
