

Editorial

When May Cochlear Implant Not Work Even at a Very Young Age?

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Multichannel cochlear implant is the first neural prosthesis to bring electronic technology effectively and safely into a direct physiological relation with the central nervous system and human consciousness.¹

Among children, classical candidates to cochlear implantation are those presenting bilateral severe to profound hearing loss when amplification benefits are limited to the optimal auditory and language development.^{2,3}

Despite the remarkable contribution to the access of speech sounds, cochlear implant outcomes vary considerably depending on several factors.⁴ Sharma et al.⁵ (2002) suggested that a critical period of plasticity existed for developing brain connections, and early surgical intervention has been indicated ever since. Nevertheless, just 'opening' the doorway of the brain (the ears) to receive sounds may not be enough to allow their interpretation.⁶

Once the hearing loss is diagnosed, an early intervention program should begin to promote receptive and expressive language development. This program will require the child to be educated to maximally use hearing with the cochlear implant.^{1,7,8}

Listening presupposes an intentional activity. In listening, one is actively trying to focus on some sounds. In contrast, hearing may happen without necessarily paying attention or intention. It is possible to hear something even when you don't want to hear it and try not to hear it. The brain pathways used in 'listening' and simply hearing are different. When listening, in addition to paying attention to what we hear, it implies finding the pathways that lead to attributing representation to that sound, that could be a meaning or a feeling stored in memory. To hear, it is enough to detect, sound can pass through the ear and reach the temporal lobe, without connecting to symbolic or meaningful associations. When one really 'want' to listen, must pay attention, inten-

tion, and trigger association pathways to the 'end point' where there must have stored information, so that the input message can be compared to previous listening experiences, be understood and make sense. At the extreme, one may put all the attention and intention into listening to someone talking in Russian, while nothing that was heard was understood, if one does not have meaningful stored information. Thus, to understand, it is necessary that the brain has a stored arsenal to be able to interpret the sound.

In the first years of life, everyone who is born with normal hearing thresholds, may be able to hear 'everything' and will store auditory experiences associated with sensations, emotions, and daily experiences that, through repetition and routine, will allow the creation of a significant arsenal (Flexer, 2011).⁶ The 'meaningful arsenal' is achieved with emotional bonding among the family members (that will give the motivation to the intention to hear, or listening) and is the basis of language. Language will be transmitted as speech (oral language) when a connection among the ascending auditory pathways, meaningful association pathways and the descending motor efferent pathways are linked. The language is accumulated with the experiences that generate the processing of several stations, regardless of the way it will be expressed.

ASHA⁹ summarizes some expected landmarks in the auditory and speech development (►Table 1) in children with typical hearing. Although they may have thresholds within normal ranges, they only respond reflexively to loud sounds during the first three months of life. They begin to imitate speech sounds only after more than 7 months of repeated auditory experiences and motor maturity in the tongue and mouth, revealing the connection between the afferent auditory and the efferent speech motor pathways. They will point out body parts around 12 months old, after

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Table 1 Expected markers in the development of listening and speaking skills during the first 2 years of life⁹

Hearing and comprehension	Speech
Birth to 3 months	
Startles with loud sounds Be quiet or smile when spoken to	Makes pleasant sounds Smile when sees mom/dad
4 to 7 months	
Moves eyes in the direction of sounds Notices toys that make noise	Babbles different speech sounds Vocalizes while playing
7 to 12 months	
Turn and look in the direction of the sound source Starts responding to requests ('come here')	Babbling has both long and short sounds. Imitates different speech sounds
1 to 2 years	
Points to some body parts Follows simple orders (where's the shoe)	Says a new word every month Uses 1-2 word phrases and questions

taking at least 365 baths or showers, listening in a pleasant and playful environment with their parents and caregivers.

Hence, the construction of language is the result of countless experiences and situations stored and associated by synaptic connections, that are potentiated by emotions and hormones.¹⁰

As mentioned earlier, when the child is born with severe to profound hearing loss and conventional amplification hearing aids are not able to stimulate all the tonotopic regions of the cochlea and cochlear nerve with speech sounds, the cochlear implant will be indicated to allow access to the necessary acoustic cues. This may allow the development of auditory skills, the experience of representative situations, culminating in oral language.¹¹

Diagnosis at birth or in the first month is very important so that the necessary actions for intervention are taken in time, that is, before 3 to 6 months, to allow this children have the meaningful experiences almost at the same time as their hearing peers.⁷ The more adequate the auditory and linguistic development before the CI, the better the evolution after the surgery. As important as the diagnosis is the supporting and counseling of families. Families that are "paralyzed" in the diagnosis, not knowing how to deal with deaf children, without the adequate guidance might unconsciously becoming "mute families". One of the possible reactions or tendencies when dealing with someone that may not be hearing us, may be 'not talking'. "What is the point of talking", one may ask themselves, "if he/she won't listen to me". The importance of supporting families involved in the child's rehabilitation process has been reinforced and proven in numerous studies.^{4,10-16} Lind-Combs & Holt¹⁵ (2022) conducted a study with 62 dyads of children with typical hearing and with hearing loss using conventional hearing aids or cochlear implants, between 3 and 8 years of age. They found that the

vocabulary used by the family associated with a mental state (such as: I think that, I feel that) positively affected the inhibitory control of children using hearing devices. Hence, not only language development is impacted by family communication style, but the whole cognitive development.

The great contribution of rehabilitation is precisely to establish a partnership, strengthening guidance and advice to families of children with hearing loss to transform the home into a stimulating environment.

Thus, in addition to the the age at diagnosis and intervention with conventional hearing aids before 3 to 6 months of age, adequate and desired oral language development after early cochlear implantation will only happen when:

- There is adequate family involvement and communication,
- Residual hearing is effectively harnessed with hearing aids before the CI, while the greater the degree of hearing loss, the shorter the deprivation time should be,
- Etiology allows proper insertion of electrodes and sufficient and adequate neural density of ganglion cells for electrical stimulation
- The Map (programming) in the speech processor is adequate to allow optimized access to speech sounds and features
- There are no other associated handicaps
- Appropriate school and rehabilitation quality
- Adequate nonverbal intelligence
- And CI usage during all the waking hours, that is to say an average of 10 hours a day.

Pianesi et al.¹⁷ (2016) warned that the risk of lack of oral language development can be identified in the first 6 months after CI, due to slow development of auditory skills identified by IT-MAIS. Silva-Comerlatto¹⁸ has also studied 230 Brazilian kids with severe to profound hearing loss, and established the landmarks for auditory and oral language after cochlear implants. Therefore, children who do not demonstrate development in the first 6 months after CI should be considered as 'red flags' and actions such as the verification of the intervention (rehabilitation and family communication), speech processor programming and hours of use should be investigated.

Robbins¹⁹ (2005) proposed a list of Red flags for both groups of children implanted early and late in life, up to 12 months after CI, that may also serve as markers for taking actions after cochlear implantation. Our proposal is that, if there is no expected evolution, a list of reasons may be reviewed to allow the identification of the aspects that may be driven (**figure 1**).

In summary

Cochlear implant is a remarkable solution to open the doorway to the brain,⁶ but the factors that influence the development of oral language, that is, auditory and communicative skills in severe to profound deaf children, transcend the age of the cochlear implant surgery.

Furthermore, we must have in mind that in very early cochlear implantation, many other additional handicap diagnoses may appear later, after the surgery, including Autism Spectral Disorders.^{20,21}

Checklist for identifying variables that may affect CI outcomes

Name: _____ date: _____

Age: _____ Age at implantation: _____ Surgery (CI) date: _____

Issues/complaint: _____

Etiology	<input type="checkbox"/>	Unknown, no impact on cochlear anatomy or nerve density	<input type="checkbox"/>	Meningitis, Otosclerosis, CND/CMF, other infectious diseases
Pre op speech&language evaluation	<input type="checkbox"/>	Adequate, no identifiable issues	<input type="checkbox"/>	Failures in communicative intention, weak attention, or speaker's face or to nonverbal cues from the speaker or environment, lack of prelinguistic landmarks have already been identified
Pre op psychological evaluation	<input type="checkbox"/>	Adequate, no identifiable issues	<input type="checkbox"/>	Issues related to the patient's family or cognitive-emotional/attentional aspects were identified
Neurological or developmental evaluation	<input type="checkbox"/>	Adequate, no identifiable issues	<input type="checkbox"/>	Other deficiencies or handicaps were identified
Post op Imaging	<input type="checkbox"/>	Adequate (complete insertion, no tip foldover or translocations to scala vestibuli)	<input type="checkbox"/>	Partial insertion, false pathway, other bad positioning
Intraop impedances	<input type="checkbox"/>	Adequate for all electrodes, including grounds	<input type="checkbox"/>	Shorts or open circuits in some electrodes
Intraoperative neural response	<input type="checkbox"/>	Present	<input type="checkbox"/>	Absent
SOE intraop	<input type="checkbox"/>	Adequate at all tested electrodes	<input type="checkbox"/>	Too wide, double peaked or not available
Free field thresholds with CI	<input type="checkbox"/>	25 - 30 dBHL	<input type="checkbox"/>	> 35 dBHL/does not show detection or attention to sound/is not conditioned to respond
Speech processor fitting (MAP)	<input type="checkbox"/>	Reliable levels	<input type="checkbox"/>	Doubtful levels (patient does not respond well)
Postoperative neural response	<input type="checkbox"/>	Present	<input type="checkbox"/>	Absent
Postoperative Impedances	<input type="checkbox"/>	Adequate for all electrodes, including grounds	<input type="checkbox"/>	Shorts or open circuits in some electrodes, fluctuating or varying across fitting sessions
Daily speech processor use	<input type="checkbox"/>	Systematic (+ 8 to 10 hours/day)	<input type="checkbox"/>	Assystematic
Speech processor maintenance	<input type="checkbox"/>	Adequate	<input type="checkbox"/>	Poor maintenance, frequent problems, long 'out of the air' periods
IT-MAIS at 6 months post CI	<input type="checkbox"/>	>60%	<input type="checkbox"/>	Evolution unchanged or <60%
Speech Recognition/Auditory Skills	<input type="checkbox"/>	Progress, expected evolution for the case	<input type="checkbox"/>	No progress or falling behind expectations
Rehabilitation	<input type="checkbox"/>	Good partnership with the Speech/Language therapist, with frequent reports promoting full participation of the family	<input type="checkbox"/>	Inadequate or insufficient. Not at all
Family/Parents	<input type="checkbox"/>	Show commitment with the rehabilitation process, in therapy and at home (Moeller > 4) ¹³	<input type="checkbox"/>	Do not adhere to/participate in the process, do not accept the diagnosis, or follows the suggestions of the Rehab team (Moeller <3) ¹³
Falls or head trauma	<input type="checkbox"/>	Absent	<input type="checkbox"/>	Present

Fig. 1 Checklist of reasons for the development below expectations. Legend: SOE. Spread of excitation; Rehab. Rehabilitation

Considering the individuality and characteristics of each child, it is the responsibility as the multidisciplinary team, including pediatricians, otorhinolaryngologists, speech therapists, social workers, psychologists, to offer support and the best possible guidance to the children and their families.

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