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Is very early hearing assessment always reliable in selecting patients for cochlear implants? A case series study

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ABSTRACT

Introduction: This study concerns a case series of 23 infants with a diagnosis of severe-to-profound hearing loss at 3 months old, who significantly improved (even reaching a normal auditory threshold) within their first year of life.

Methods: All infants were routinely followed up with audiological tests every 2 months after being fitted with hearing aids as necessary. A reliable consistency between the various test findings (DPOAE, ABR, behavioral responses, CAEP and ECoG) clearly emerged in most cases during the follow-up, albeit at different times after birth.

Results: The series of infants included 7 cases of severe prematurity, 6 of cerebral or complex syndromic malformations, 5 healthy infants, 2 with asymptomatic congenital CMV infection, and 1 case each of hyperbilirubinemia, hypoxia, and sepsis.

All term-born infants showed a significant improvement over their initial hearing threshold by 6 months of age, while in most of those born prematurely the first signs of threshold amelioration occurred beyond 70 weeks of gestational age, and even beyond 85 weeks in one case.

Conclusions: Cochlear implantation (CI) should only be considered after a period of auditory stimulation and follow-up with electrophysiological and behavioral tests, and an accurate analysis of their correlation. In our opinion, CI can be performed after a period of 8 months in all term-born infants with persistent severe-to-profound hearing loss without risk of diagnostic error, whereas the follow-up for severely preterm infants should extend to at least 80 weeks of gestational age.

1. Introduction

A child with congenital severe-to-profound hearing impairment should receive conventional hearing amplification as soon as possible, followed by cochlear implantation (CI) within the first year of life. This approach is believed to enable the maximal development of their communication skills, social exchange and cognitive abilities [1]. Unfortunately, very early audiological diagnoses are not always reliable, however (even when they are established at tertiary centers), and not all children identified as suffering from sensorineural hearing loss (SNHL) will have permanently impaired hearing thresholds. The absence of any electrophysiological or behavioral reaction to sound does not always reflect SNHL [2–4]. Many factors can affect hearing pathway maturation and the capacity to respond to a given threshold, including prematurity, neurological and metabolic diseases, hyperbilirubinemia, hypercholesterolemia, hypoxia, head trauma at birth, and the administration of certain antibiotics and diuretics at neonatal intensive care units [5,6]. It has also been repeatedly demonstrated that there is a strong association between these risk factors (especially hyperbilirubinemia and hypoxia) and auditory neuropathy/dys-synchrony (AN/AD) [7]. A complete or partial recovery of auditory brainstem responses (ABR) has frequently been reported in high-risk infants [8,9] and pediatric patients, but the most typical examples of ABR reversibility are probably those described in extremely and severely premature neonates (born at less than 31 weeks of gestation) [10]. The morphological and physiological development of the auditory pathway in postnatal life is characterized mainly by a continuing synaptogenesis and myelination of the nerve fibers, which begin to develop in the final stages of intrauterine life [11]. Taking these ontological aspects of the auditory pathway into account, variations in latencies and amplitudes of ABR waves in newborn and lactating infants can clearly coincide with the process of auditory maturation, as well as with possible impairments of auditory pathway components

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(from the cochlea to the cerebral cortex) [12]. Reversible abnormalities are observed in healthy infants, and the absence of risk factors and diseases in such cases makes it difficult to explain the mechanisms behind their impaired maturation.

The aim of this study was to describe a case series of 23 infants with an initial diagnosis of severe-to-profound hypoacusia, who showed a significant improvement (and even a normalization of their auditory threshold) during the first year of life, making CI unnecessary.

2. Material and methods

We reviewed the medical records on 438 infants seen at the pediatric audiology center of the Padova University ENT Department between January 2010 and December 2013. All the infants were under 6 months old and were referred to us with an initial diagnosis of bilateral or unilateral SNHL for a tertiary-level audiological diagnosis and, where necessary, for CI.

All infants underwent audiological follow-up every 2 months, and joined a program for the fitting of hearing aids. The audiological assessment was personalized and based on a combination of objective and subjective audiometric findings (Table 1), with regular cross-checks [13]. For the purpose of this study, we selected those infants in our series who showed signs of a significant improvement, and even a normalization of their auditory behavior for their developmental age, that enabled CI to be postponed or avoided.

An amelioration was considered reliable when one or more of the following conditions were satisfied: (i) the second or subsequent ABR (in one ear at least) revealed a consistently better replicability, a better trace morphology, and lower latencies; (ii) the psychoacoustic threshold improved by 20 dB or more vis-à-vis the previous ABR threshold; (iii) a significant improvement emerged from parental reports and questionnaires (PEACH) [14], associated with a clinical and behavioral picture judged to be normal for the child's developmental age in response to vocal stimuli (Ling-6 sounds) presented at a mild intensity. A reliable consistency between the various tests findings, according to the Jerger & Hayes cross-check principle [15], emerged clearly in most cases during the follow-up, albeit at different times after birth. Total consistency between behavioral and electrophysiological test findings was not an absolute requirement for defining amelioration, however. Lengthy periods in which there were inconsistencies between behavioral and electrophysiological responses were seen in several cases, due mainly to AN/AD. To avoid any possible biases, infants who revealed steeply rising or falling thresholds during the follow-up were ruled out, and so were those with middle ear dysfunction, as assessed on 1 kHz or 250 Hz probe tone tympanograms, depending on the child's age. The study has been approved by the Regional Committees for Ethical Medical and Health Research.

3. Results

In our series of 438 infants, 367 (83.8%) were not implanted promptly; they were fitted instead with hearing aids as necessary and followed up routinely for at least a year. Six infants (1.4%) were lost to follow-up. Sixty-five (14.8%) had a CI within 8 months of life in the light of a highly reliable diagnosis; these implants were unilateral in 42 cases (64.6%) and bilateral in 23 (35.4%). One premature newborn was implanted at 2 months of corrected age due to ongoing cochlear ossification secondary to neonatal meningitis. None of the 42 infants treated with early unilateral implants experienced any amelioration of their contralateral threshold after a mean follow-up of 21.7 months (range 2.1–47.3 months).

In the group of 367 infants who were monitored but not implanted, we identified 23 cases (6.2%) whose hearing threshold improved significantly during their first year of life. Nineteen of these 23 infants underwent MRI during this period, and none had cochlear nerve hypoplasia. Tables 2 and 3 summarize the evolution of the reversible hearing loss in the 23 untreated cases. Table 2 contains data on 7 premature infants whose hearing improvement is given in terms of weeks of gestation. Table 3 provides data on 16

Table 1
Audiological assessment parameters.

Anamnesis and objective exam	Family history, prenatal, perinatal, postnatal findings Otoscopy	
Electrophysiology	Auditory brainstem responses (ABR)	Broadband click stimuli of 100 ms; polarity: alternating; repetition rate: 21 p/s; contralateral masking
	Auditory steady state respones (ASSR)	Pure-tone stimuli, varying frequency and amplitude
	Electrocochleography (ECoG)	Extratympanic (foam ear plug electrode), or transtympanic (sedation or general anesthesia). Click stimulus: alternate or rarefaction/condensation; 11, 21, 30 p/s. Contralateral masking
	Cortical auditory evoked potential (CAEP)	Unaided free field presentation. Stimulus: speech sounds (/m/, /g/, /t/) at 65 dB SPL
OAE	DPOAE – distorsion product otoacoustic emissions	Tone pairs 2F1–F2. Four stimulation frequencies in the 1000–6000 Hz range
	TEOAE – transient evoked otoacustic emissions	Click stimulus
Acoustic immittance	Physical volume of the canal and dynamic tympanogram Ipsilateral and contralateral stapedius reflex	Test sound of 1 kHz (or 226 Hz in older children)
Behavioral	BOA behavioral observation audiometry	Warble stimulus/narrow band noise stimulus
	VRA visual reinforced audiometry	Warble stimulus/narrow band noise stimulus at 90° azimuth
Imaging	Cerebral magnetic resonance (MRI) Inner ear MRI	Sedation or general anesthesia, with/without gadolinium
	Temporal bone computed tomography (CT)	High-resolution computed tomography (HRCT) or cone beam computed tomography (CBCT)
Genetic molecular evaluation	Selective gene evaluation or comprehensive genetic screening platforms	
Speech perception and early language development	Early vocalization, speech and language tests Parental questionnaires, parental reports	NEAP, PRICE, MacArthur's language development questionnaire, Ling 6 sound test, MAIS (Italian versions), PEACH