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Effect of age at cochlear implantation on auditory and speech development of children with auditory neuropathy spectrum disorder

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ABSTRACT

Objective: To evaluate the auditory and speech abilities in children with auditory neuropathy spectrum disorder (ANSD) after cochlear implantation (CI) and determine the role of age at implantation.
Methods: Ten children participated in this retrospective case series study. All children had evidence of ANSD. All subjects had no cochlear nerve deficiency on magnetic resonance imaging and had used the cochlear implants for a period of 12–84 months. We divided our children into two groups: children who underwent implantation before 24 months of age and children who underwent implantation after 24 months of age. Their auditory and speech abilities were evaluated using the following: behavioral audiometry, the Categories of Auditory Performance (CAP), the Meaningful Auditory Integration Scale (MAIS), the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS), the Standard-Chinese version of the Monosyllabic Lexical Neighborhood Test (LNT), the Multisyllabic Lexical Neighborhood Test (MLNT), the Speech Intelligibility Rating (SIR) and the Meaningful Use of Speech Scale (MUSS).
Results: All children showed progress in their auditory and language abilities. The 4-frequency average hearing level (HL) (500 Hz, 1000 Hz, 2000 Hz and 4000 Hz) of aided hearing thresholds ranged from 17.5 to 57.5 dB HL. All children developed time-related auditory perception and speech skills. Scores of children with ANSD who received cochlear implants before 24 months tended to be better than those of children who received cochlear implants after 24 months. Seven children completed the Mandarin Lexical Neighborhood Test. Approximately half of the children showed improved open-set speech recognition.
Conclusion: Cochlear implantation is helpful for children with ANSD and may be a good optional treatment for many ANSD children. In addition, children with ANSD fitted with cochlear implants before 24 months tended to acquire auditory and speech skills better than children fitted with cochlear implants after 24 months.

1. Introduction

Auditory neuropathy spectrum disorder (ANSD) is a relatively new term that was adopted by the panel of the International Newborn Hearing Screening Conference in Como, Italy, in 2008. The disorder is thought to be a type of hearing impairment in which outer hair cell function is normal but afferent neural conduction in the auditory system is disordered. "Auditory neuropathy" and "auditory dys-synchrony" are terms that have been used to describe this disorder.

Starr et al. and Kaga et al. proposed auditory neuropathy (AN) in 1996 [1,2]. Starr et al. reported 10 patients with specific hearing impairments. In all of the patients, the preservation of otoacoustic emissions (OAEs) and cochlear microphonics (CMs) demonstrated that cochlear outer hair cell function was normal, whereas absent or severely abnormal auditory brainstem potentials showed evidence of abnormal auditory pathway function. Kaga et al. reported two patients who also lacked auditory brainstem

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responses (ABR) but exhibited almost normal otoacoustic emissions. The cause of the auditory neuropathy was suggested to be an abnormality of the VIII nerve.

Because the constellation of test results defining this disorder does not provide direct evidence of abnormal auditory nerve function or ''neuropathy,'' a term was necessary that includes more data [3–5]. Berlin and colleagues thus proposed the term ''auditory dys-synchrony'' [4].

An increasing number of studies have indicated that the mechanism may involve a disorder of the inner hair cells in the cochlea, disorder of the nerve synapse at the auditory nerve, or an auditory nerve lesion [3,6–10]. Thus, ANSD appears to be the best term to reflect the various sites of pathology.

ANSD patients differ greatly from one another in their ability to use temporal cues. The audiometric results of ANSD patients vary greatly from normal hearing to severe hearing loss. These patients experience great difficulty in understanding speech, particularly in the presence of noise [1,11,12].

ANSD in children should never be considered as a rare form of hearing loss. Studies suggest that ANSD is involved in 2.4–15% of permanent childhood hearing loss [10,13,14]. Vlastarakos et al. recently performed a systematic review that indicated that ANSD accounts for approximately 8% of newly diagnosed cases of hearing-impaired children each year [15].

In 2006, several cochlear nerve deficiency (CND) cases were reported in patients with ANSD [9]. Since then, the reported prevalence of CND in children with ANSD has been between 18% and 28% [9,16]. CND may be observed in association with congenital or acquired sensorineural hearing loss (SNHL) and may be important in the assessment of patients for cochlear implantation [17]. Thus, these factors can be applied to ANSD patients.

There are three strategies, including hearing aids, cochlear implants (CIs) and FM systems, that could benefit ANSD patients. Recently, several articles proposed that CI may result in better success rates in ANSD patients [14,18,19]. For ANSD children with profound hearing loss, CIs are more effective.

The purpose of this study was to evaluate the auditory and language abilities after cochlear implantation in children with ANSD excluding CND and to determine the role of age at implantation to enrich the available clinical data.

2. Materials and methods

2.1. Subjects

This study included 10 children. All children were implanted in the Otorhinolaryngology Department of TongRen Hospital under the same pediatric cochlear implant program. The children all had clinical evidence of ANSD, and all fit the following criteria: (1) OAEs and/or CMs; (2) a severely abnormal auditory brainstem response (ABR), which was defined as absent waveforms or absent responses at the maximum output level of 100 dB normal hearing level; (3) no cochlear nerve deficiency on magnetic resonance imaging (MRI) and computed tomography (CT) and (4) bilateral profound hearing loss. Of these children, 8 were male, and 2 were female. The mean age at implantation was 35.5 ± 26.2 months (range 11–86 months), and the mean duration of use was 45.6 ± 23.9 months (range 12–84 months).

An audiologic assessment, speech therapy and psychological evaluations were performed on all children. The CIs in children were programmed using the speech processing strategy recommended at the time and upgraded with new encoding strategies as they became available.

We divided the children into two groups: children who underwent implantation before 24 months of age and children who underwent implantation after 24 months of age.

2.2. Evaluation tests

The development of auditory perception and speech production skills was assessed using the following test batteries: the Categories of Auditory Performance (CAP) [20], the Meaningful Auditory Integration Scale (MAIS) [21], the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS) [22], the Standard-Chinese version of the Monosyllabic Lexical Neighborhood Test (LNT), the Multisyllabic Lexical Neighborhood Test (MLNT) [23], the Speech Intelligibility Rating (SIR) [24] and the Meaningful Use of Speech Scale (MUSS).

The CAP ranks the auditory receptive ability into eight hierarchical categories from 'no awareness of the environment' to the 'use of the telephone with known users' and is intended to reflect the real-life progress of children in their auditory abilities.

The MAIS is a test battery administered through parental interviews to assess the early prelingual auditory development of children aged 3 years and older. The scale comprises 10 items, and each item is scored on a scale of 0–4. The MAIS is intended to address the parents' evaluation of their child's hearing capabilities. The first 2 items evaluate the child's acceptance of and reliance on the device. The next 4 items assess the child's detection of and spontaneous response to his/her own name and to environmental sounds and signals. The final 4 items assess the child's ability to recognize and discriminate sounds, such as the differences between two speakers' voices, differences between speech and non-speech sounds and differences in vocal tones that convey emotions.

The IT-MAIS is used in young children from infancy to 2 or 3 years of age and is administered and scored in the same way as the MAIS. However, the first 2 items of the scale have been changed. These two items relate to the child's vocal behavior rather than the child's acceptance of and reliance on the device. The remaining items are identical to those in the MAIS.

The Standard-Chinese version of the LNT and the MLNT are used to examine the open-set speech recognition abilities of children. They consist of monosyllabic word lists and disyllabic word lists. Both monosyllabic and disyllabic word lists are further divided into easy and hard lists.

The SIR is an ordinal scale to rank the child's spontaneous speech into five categories. It is a measure of speech production in real-life situations with high reliability and time-effectiveness.

The MUSS is a parental reporting scale used to determine the frequency of speech use in the child's daily behavior. It consists of 10 questions.

To perform behavioral audiometry, the Standard-Chinese version of the LNT, and the MLNT, test stimuli were presented using a loudspeaker situated at a distance of 1 m and producing an average presentation sound pressure of 70 dB. Each child was tested under normal listening conditions with the cochlear implant.

2.3. Statistics

All results were analyzed using the Statistical Package for Social Sciences (SPSS) 17.0 (SPSS Inc., Chicago, IL, USA) and R 3.0.2 (R Development Core Team, Vienna, Austria). The mean, standard deviation (SD) and minimum–maximum values are presented as descriptive statistics. Statistical comparisons were made using the Wilcoxon signed rank test. A p-value of <0.05 was determined to be statistically significant.

3. Results

All children progressed in their auditory and speech abilities. The 4-frequency average hearing level (HL) (500 Hz, 1000 Hz,