Cr ic Re e he eff c cy of he r ng p f c on on speech percep on res s hen sed s h r on e hod for ch dren h A d ory Ne rop hy pec r D sorder

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This critical review examines the efficacy of hearing amplification on speech perception results when used as a habilitation method for children with Auditory Neuropathy Spectrum Disorder in eight studies. Study designs include 3 case series studies, 2 nonrandomized clinical trials, 2 case-control studies, and a retrospective single group study. Overall, the evidence provided by these studies is inconclusive in providing support for the use of hearing amplification as a primary treatment for children with Auditory Neuropathy Spectrum Disorder. This is due to the limited sample sizes, limited related research available, and biases in the populations selected. Further research should address these problems as well as appropriate ages for fitting and settings of hearing amplification.

Introduction

Auditory Neuropathy Spectrum Disorder (ANSD) involves a collection of conditions with a common diagnostic profile. ANSD is clinically diagnosed by an absent/abnormal auditory brainstem response (ABR) due to neural dysfunction at the level of cranial nerve VIII, in the presence of outer hair cell function, indicated by present otoacoustic emissions (OAEs) and/or cochlear microphonics (CM). Patients with ANSD present with a multitude and variety of features, including fluctuating or permanent hearing loss, speech reception scores that are much lower than would be expected from the individual's pure-tone air conduction results, absent acoustic reflexes, and normal radiological findings. This variability in the presentation of ANSD makes it difficult for an accurate measure of incidence of the population, with reports varying from 1.83%, to as high as 11% of the hearing impaired population (Kumar, 2006). These rates are likely to increase due to improved detection methods in infant hearing screening programs and as strategies for caring for premature and low-birth weight babies improve. Some of the variability in presentation of those with ANSD is thought to stem from the site-of-lesion associated with the dysfunction. The sites-of-lesion are thought to be either the inner hair cells, the tectorial membrane, the synapse between the inner hair cells and the auditory nerve, or some combination of these (Rance, 2005; Santarelli, 2002).

There is significant clinical dilemma associated with the treatment of Auditory Neuropathy Spectrum Disorder, thought to be due to the differing sites of lesion and range of auditory perceptual abnormalities. Some patients have found success with hearing amplification, some with cochlear implants, while others were only successful with manual forms of communication. This issue is especially critical for infants, as language development is of utmost importance. No reliable behavioural measurements can be obtained until about 6 months of age, due to immaturity, and a threshold ABR

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Rance et al. (2002) assessed speech perception and production in the aided and unaided conditions in children with auditory neuropathy spectrum disorder (n= 18), presenting with an absent ABR waves, elevated pure-tone or speech thresholds, and present cochlear microphonics and/or OAEs. A control group of 18 children with SN hearing loss were matched based on chronologic age and pure-tone audiogram. All subjects had been diagnosed in infancy and had been fit with amplification by 12 months, with the exception of 3 who were fit by 24 months. All had been consistent hearing instrument users for at least 12 months at the time of assessment with BTE aids fit to target using the NAL prescriptive method.

Only 15 of the children in the ANSD group were able to complete speech perception testing, due to immaturity (n=2) or physical disabilities (n=1). In the unaided condition, all AN children displayed poor open-set speech perception abilities, with improvement in the aided condition in 8/15 cases, with a mean PBK difference score (aided – unaided) of 56.8% in these cases. No correlation was found between the aided PBK score and the age at assessment ($r = 0.13$, $p = 0.18$), between age at hearing instrument fitting and

remaining aided children, as they're not an unbiased sample of the ANSD population.

There were concomitant issues in many of the ANSD subjects that excluded them from speech perception testing, or affected the results. Some issues, such as prematurity or mental retardation, excluded subjects from speech perception testing, while other studies had issues with compliance, which also decreased their sample sizes. Differing sites of lesion and associated risk factors (i.e. hyperbilirubinemia, ototoxicity, consanguinity, etc.) could also have increased the variability of the results somewhat. These issues could affect the results by introducing additional complications that may affect the testing procedure, by decreasing the matching reliability across subject groups or by not fairly representing the general population. (Delentre et al (1999), Rance et al (2002), Rance et al (1999), Raveh et al (2006)).

More information is needed on the ANSD (aided, implanted) and SNHL control populations before a treatment measure is selected to determine proper matching between the groups. This is not the focus of the majority of the studies. However, when Rance et al. (2007) did disclose whether oral or total communication was the primary communication method for the child, differences were found. Speech perception results would naturally be affected by the mode of communication used by the child, which may affect some of the studies' findings when not controlled for. Rehabilitation measures and support available were also not controlled for in these studies, but could affect a child's success.

Conclusion and Clinical Implications

It is not clear from the literature what clinical changes can or should be made when analyzing a treatment method for a patient with ANSD, as many of the include56603(m)17.497re ti hi or.64358(n)56603(i)0.3

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