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Critical Review: The Effect of Cochlear Implantation on Speech Perception and Neural Integrity for Children with Auditory Neuropathy/Dyssynchrony Compared to Children with Sensory Hearing Loss

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This critical review examines the speech perception and electrophysiological outcomes of cochlear implanted children who have been identified with Auditory Neuropathy/Dyssynchrony compared to cochlear implanted children with a sensory hearing loss. Study designs include: case study and non-randomized clinical trials. Overall, research supports the idea that cochlear implants can provide benefit in terms of improved speech perception and neural integrity for implanted children with Auditory Neuropathy/Dyssynchrony. However, a concrete statement cannot be made regarding the ability of these children to perform on speech perception measures compared to implanted children with a sensory hearing loss. This is due to the limited availability of research, small sample sizes, and lack of statistically significant calculations of results.

Introduction

Auditory Neuropathy/Dyssynchrony Disorder (AN/AD) is a type of hearing impairment in which outer hair cell function in the cochlea is preserved, however neural synchrony of the auditory nerve is impaired. Clinical presentations of AN/AD generally include the presence of otoacoustic emissions, abnormal auditory nerve activity, absent middle ear muscle reflexes, and various degrees of hearing loss. The underlying physiological mechanisms of AN/AD are difficult to determine in any given individual. The proposed sites of lesion include the cochlear inner hair cells, the synapse between the inner hair cells and the auditory nerve, and/or the auditory nerve itself. In many individuals poorer speech perception than what the pure tone audiogram would suggest is observed. This is thought to be due to temporal encoding problems as a result of auditory nerve dyssynchrony. When speech signals are presented at an audible level difficulty is still observed which results in limited use with conventional amplification in numerous patients (Rance & Barker, 2008).

Treatment for children with AN/AD is challenging considering the multiple possible etiologies that could be responsible for the clinical presentation of this disorder (Trautwein et al., 2000). Cochlear implantation for patients with severe speech processing difficulties who do not benefit from traditional amplification is currently used as a treatment option Rance & Barker, 2008). Traditional amplification is often an unsuccessful treatment in many, but not all instances. Some clinicians hesitate to recommend implantation for children with AN/AD because they assume that electrical stimulation of the auditory nerve will cause the same inferior result as acoustical stimulation. In many cases these children show a benefit in terms of speech perception from cochlear implantation with only a few cases experiencing poor results. It is useful to note that some children also demonstrate more synchronous ABR recordings post implantation. Different implantation outcomes support the notion that there are multiple possible etiologies responsible for AN/AD (Buss et al., 2002).

Previously, few studies with sufficient sample sizes have been published due to the small number of children with AN/AD who have received implants. This is a relatively new and important area of research that is essential in order to provide a basis for habilitation recommendations (Buss et al., 2002). Comparing implanted AN/AD children to implanted children with a sensory loss could reveal whether or not AN/AD children will achieve the same success with an implant and/or if they require a different customized habilitation plan.

Objectives

The primary objective of this review is to critically evaluate the existing literature regarding the benefits of a cochlear implant in terms of speech perception and neural integrity in children with Auditory Neuropathy/Dyssynchrony (AN/AD) compared to children with a sensory hearing loss.

Methods

<u>Search Strategy</u>

Computerized databases included PubMed and SCOPUS using the following search strategy: (Auditory Neuropathy) AND (Cochlear Implant) AND (Sensorineural Hearing Loss) AND (Children) OR (Auditory Neuropathy Dyssynchrony) AND (Cochlear Implant) AND (Children)

<u>Selection Criteria</u>

Studies selected for inclusion in this critical review were required to investigate outcomes in speech perception and/or electrophysiological measurements of neural integrity in patients with AN/AD and sensory hearing loss using a cochlear implant. This review is directed towards the pediatric population, therefore, studies which included individuals implanted as adults were excluded.

Studies which did not compare the results of children with AN/AD to children with sensory hearing loss were also excluded. No limitations were placed on the type of outcome measures that were used to determine speech perception or neural performance.

Data Collection

Results in the literature search yielded three articles that were congruent with the selection criteria above: case study (1) and non-randomized clinical trials (2).

Results and Discussion

Three studies have been conducted that compare speech perception and/or electrophysiological benefits in cochlear implanted children with Auditory Neuropathy/Dyssynchrony (AN/AD) versus cochlear implanted children with a sensory hearing loss.

Buss et al. (2002) studied four children identified with AN/AD who had received a Clarion cochlear implant unilaterally through the Carolina Children's Communicative Disorders Program at the University of North Carolina. The first two subjects (S1 and S2) were implanted at approximately 2 yrs of age and used oral communication, whereas the remaining two (S3 and S4) were implanted at approximately 5.5 yrs of age and used manual and cued speech respectively. The decision to implant the children in this study was made after an unsuccessful trial period with amplification. All of the children were diagnosed with AN/AD by the presence of a cochlear microphonic or otoacoustic emissions and the absence of a synchronized auditory brainstem response pre operatively.

Speech perception outcomes were measured by the Paden-Brown test. This standard test assesses the kinds of errors children with poor auditory language exposure might make in producing speech. The children were scored based on type of error: number of syllables, stress pattern across syllables, initial consonant, final consonant, inclusion of appropriate vowel, production of diphthong, consonant manner, consonant place, and consonant voicing. The results of this test were compared to the results of implanted children with a sensory hearing loss who were matched for age and duration of implantation (n=33). The Paden-Brown test was given to all the children by same Speech-Language Pathologist, however it is unknown if the tester was blind to the subject groups

Neural integrity was evaluated by delivering 75 μs biphasic pluses at 21.2 pulses/s to electrodes 1, 4, or 8 in the implanted device to obtain an EABR measure. EABR data were collected using a Nicolet Biomedical system in combination with an Advanced Bionics speech processor to deliver the stimulus. Acoustic reflexes were measured contralateral to the implanted ear using a 226 Hz probe tone.

The children were grouped by age for comparison. 2-4 years of age for subjects 1 and 2, and 4-6 years of age for subjects 3 and 4. The results of the Paden-Brown test indicated that for all measures subjects 1, 2 and 4 were within or just above a one standard deviation confidence interval of the mean for implanted children with a sensory hearing loss. Half the the subjects performed better than the mean for implanted children with sensory hearing loss. Subject 3 fell below one standard deviation below the mean on two of the nine measures which could have been linked to the continued use of manual speech for communication. Wave 5 in the EABR recordings for all patients were present on at least two of the three electrodes tested. There was some variability in latency and morphology of wave 5 between subjects however this variability was also observed in implanted children who were not identified with AN/AD. At least one acoustic reflex was recorded for all four subjects indicating improvement in neural integrity post implantation.

This study was conducted with a very small sample size (n=4) compared to that of the control group (n=33). The data obtained for the control group were most likely representative of their population however the same conclusion cannot be made regarding the AN/AD group. In this study the researchers did not account for possible speech production or vocabulary difficulties as possible confounds of the results of the Paden-Brown test. It is not indicated what type of auditory training either the controls or the subjects had post implantation which could affect the test results. Statistically significant differences between the control and the subject groups were not analyzed. Performance was gauged by comparing the subjects to the mean of the control group and discussing whether or not they were within one confidence interval (1 SD) of the mean.

Rance and Barker (2008) studied 20 children with an AN/AD related hearing loss who demonstrated an absent auditory brainstem response and present cochlear microphonics or otoacoustic emissions. Half of these children were fitted with a Nucleus cochlear implant either monaurally or binaurally following limited success with amplification. All but one child demonstrated an electrical auditory brainstem response within normal limits post implantation. This group had a mean implantation age of 33.3 ± 16.9 months with a mean age at testing of 89.6 ± 42.1 months. The other 10 children demonstrated some success with amplification and were fitted with BTE hearing instruments bilaterally from an early age. These children were selected to match the implanted AN/AD group for age at assessment at 94.2 ± 57 months. Both of these groups were compared to a third group of 37 implanted children with sensory hearing loss. They were also matched to the AN/AD implanted group for age of implantation and age at assessment.

Speech perception outcomes were examined using pre-recorded consonant-nucleus-consonant (CNC) phonemically balanced words presented at 70 dB SPL at 1m from the speaker. The children were asked to repeat the test items which were later transcribed to estimate a percent correct phoneme score. Experimenter bias was not controlled for; it is unknown if all data was transcribed by the same individual, or if the transcriber (s) were blind to the subject groups. Each child had a phoneme repertoire greater than 80% as indicated by the Diagnostic Evaluation of Articulation and Phonology test. This ensured that difficulties in speech production were not likely to confound the results.

A one way analysis of variance of speech perception results indicated that implanted children with sensory hearing loss scored statistically significantly better than the implanted AN/AD group and the amplified AN/AD group ($F2, 28=8.07$ $p = 0.002$). However, it is important to note that children with AN/AD still demonstrated improved open speech perception post implantation. The results for the aided AN/AD group were variable but not significantly different from the implanted AN/AD group. Regression analysis revealed no relationship between percent correct phoneme score and average hearing loss. Regression analysis also demonstrated a lack of correlation between phoneme score, age at assessment, age at implantation, or duration of implantation use for any of the subject groups.

Trautwein et al. (2000) examined the case study of a child with AN/AD who, after 16 months of amplification, had little auditory-oral language skills. This child was implanted with a Nucleus CI24M at 3 yrs and 3 months of age with follow up testing occurring one year later. Prior to implantation the patient had no recordable auditory brainstem response. To measure neural integrity post implantation an electrically evoked compound action potential (CAP) was used. The subject demonstrated robust CAPs in response to electrical stimulation with no decrease in amplitude with a faster click rate. This suggests that some neural synchrony and therefore temporal encoding was present post implantation.

Speech perception data were collected on three measures for this child. Results of these tests were then compared to ten other children who had received either a Nucleus 22 or a Nucleus CI24M cochlear implant but did not have an AN/AD related hearing loss.

The Ling Six Sounds Test assesses how well the patient can aurally discriminate between /a, u, i, s, sh, m/ without any visual cues. This test was given to all test subjects before implantation as well as one year post implantation. The child with AN/AD's scores improved post implantation to where he could discriminate between the phonemes accurately. This was comparable to most of the other implanted children with a sensory hearing loss. Whether or not this improvement is statistically significant is not discussed. Another speech measure used in this study was the Early Speech Perception Test. This test is a tool used to evaluate children with limited vocabulary and poor language skills. The results of three sets of stimuli presented in a closed set leads to the placement of each child into four speech perception categories: 1 = no pattern perception, 2 = pattern perception, 3 = some word identification and 4 = consistent word identification. Prior to implantation all children were evaluated as being in category 1. One year post implantation the child with AN/AD improved into category 4. Results for the implanted children with sensory loss were variable with one child remaining in category 1, and others reaching category 4

The final speech perception measure, the Test of Auditory Comprehension tests auditory discrimination using closed sets recorded on a tape. This test assesses supra-segmental discrimination, memory-sequencing abilities, auditory comprehension, and auditory figure ground abilities. This measure was only recorded one year post implantation. The child with AN/AD scored at the third subset indicating that he had the ability to decipher stereotypic messages. Results for the implanted children with a sensory loss were variable from zero subsets to five subsets passed.

Overall, the child with AN/AD had improved speech perception results from pre to post implantation and was within the range of results obtained from ten implanted children with sensory hearing loss. However due to the nature of this study (case study) general conclusions about implanted children with AN/AD compared to implanted children with sensory hearing loss cannot be made based on these speech measures. The child with AN/AD scored better on some measures and worse on others when compared to children with a sensory hearing loss. Tests of statistical significance based on the results obtained in this study were not performed.

Conclusions and Clinical Recommendations

Previous research on both pediatric and adult populations with AN/AD has shown that improvement in speech perception and neural integrity has been noted in the majority of cases post cochlear implantation. However, benefit is not observed in every single case. It is difficult to predict who will benefit and who will not due to the complicated etiology of this disorder (Rance and Barker, 2008).

Although the available literature indicates that overall, children with Auditory Neuropathy/ Dyssynchrony demonstrated benefit from cochlear implantation similar to children with a sensory hearing loss, there are some limitations to this research. All of the studies discussed above agree that a restoration of neural integrity and temporal encoding which is essential for speech perception and language development occurred in implanted children with AN/AD. This indicates that electrical stimulation from a cochlear implant can result in synchronous firing of the auditory nerve in children with AN/AD. Buss et al. (2002) and Trautwein et al. (2000) found that children with AN/AD and children with a sensory loss performed similarly on speech perception measures post cochlear implantation. However, based on Rance and Barker's (2008) study, children with AN/AD may not perform at the same level on speech production measures as children with a sensory hearing loss. Given the rarity of the condition and the ethics surrounding denying a viable treatment, children in the above studies were not randomly assigned to treatment groups

Cochlear implantation may not be a suitable recommendation for every child diagnosed with Auditory Neuropathy Dyssynchrony. Rance and Barker (2008) concluded that open set speech perception can be observed in some amplified children with auditory neuropathy. Therefore a trial with amplification should be performed and candidacy for implantation considered only after the child does not demonstrate any benefit in speech perception and language development over time.

Based on the available research a concrete statement cannot be made regarding the ability of implanted AN/AD children to perform on speech perception measures compared to implanted children with a sensory hearing loss. This is due to the limited availability of research, small sample sizes, absence of a uniform speech perception measure, and the lack of statistically significant calculations of the results. Based on Rance and Barker's (2008) study, we might expect implanted children with AN/AD to require more rehabilitation and training to reach the same level of function with a cochlear implant as children with a sensory hearing loss. This has clinical implications for determining candidacy for implantation and for counseling children's families regarding rehabilitation plans and expectations. Further research is needed to determine how much and what type of training implanted children with AN/AD might need to perform similarly to implanted children with a sensory hearing loss; and if it is possible for this level to be attained. This research would require larger sample sizes and a more controlled post implantation rehabilitation program.

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