Critical Review: The efficacy of hearing amplification on speech perception results when used as a habilitation method for children with Auditory Neuropathy Spectrum Disorder.

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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 cannot be interpreted due to its abnormalities. The concern is to get the best treatment possible to ensure maximum language exposure to developing infants (Ngo, 2006).

Some of the controversy associated with hearing amplification as a treatment method centers around the disrupted temporal resolution associated with ANSD. There is concern regarding whether hearing amplification will simply further distort speech perception by amplifying background noise and an already distorted signal. Concern over possible cochlear damage created by increased sound levels associated with hearing aids is often a reason for clinicians delaying hearing amplification until after the patient's otoacoustic emissions have disappeared (Zeng, 2006). The mechanism responsible for OAE deterioration is not currently known, but the concern over outer hair cell (OHC) damage, from over-amplifying these children, is enough to delay amplification. Improvements in speech perception have, however been found in children with ANsd after being aided binaurally (Rance, 2005).

Objective

The primary objective of this review is to critically evaluate the existing literature regarding the success of hearing amplification for improving speech perception in children with Auditory Neuropathy Spectrum Disorder.

Methods

Search Strategy

Computerized databases including PubMed, Medline, CINAHL, and SCOPUS were searched using the following search strategy: [(auditory) OR (hearing) AND (dyssynchrony) OR (neuropathy) OR (ANSD) OR (AN/AD) AND (speech) AND (psychoacoustics) OR (amplification) OR (suprathreshold)].

The search was limited to articles written in English. Reference lists in acquired sources were also reviewed to find additional related journal articles.

Selection Criteria

Studies selected for inclusion in this critical review were required to investigate the improvements in speech perception measures in children with ANSD using hearing amplification. No limits were set on the age of the children when aided, primary communication method, the research methods, or type of speech perception measure used.

Data Collection

Results of the literature search produced the following types of studies consistent with the above mentioned selection criteria: case series study (3), nonrandomized clinical trial (2), case-control study (2), and a retrospective single group study. As determined by Archibald's Experimental Design – Decision Tree, four studies had an evidence level of 3 (Delentre et al, Lee et al, Rance et al (1999), and Raveh et al), two had an evidence level of 2a+ (Rance et al (2009) and Rance et al (2002)), and two had an evidence level of 2b (Rance et al (2008) and Rance et al (2007)).

Results

Retrospective Case-Series Pre- and Post-Test

Delentre et al. (1999) provided results for two children who presented with absent or severely distorted auditory brainstem responses (ABRs) with preserved cochlear microphonics (CMs) and a loss of otoacoustic emissions (OAEs), identified in infancy.

The first child was born prematurely at 28 weeks and suffered from serious neonatal complications, including severe respiratory distress syndrome and hyperbilirubinemia. At 3 years of age, behavioural thresholds were obtained in the moderate range, as measured in the free-field.

The second child was also born prematurely at 30 weeks and suffered from hyperbilirubinemia and exhibited several autistic traits. Due to the neurological deficits and autistic behaviour, behavioural testing was not easily obtained and amplification was not pursued, even with his reported threshold enhancement in hearing instrument trials.

For the first child, amplification was withheld until 4 years of age upon loss of her otoacoustic emissions (OAEs) due to concerns over OHC damage. The prescriptive method used and information about her hearing instruments and fitting are not disclosed so appropriateness of fitting and hearing instruments cannot be assumed. It was noted that her language began to develop rapidly following the hearing instrument fitting and aided open-set speech perception scores (phoneme identification and word recognition) showed significant improvement when measured, as compared to unaided, at ages 6 and 7 years. At 6 years of age, the child showed improvement from 0% in the unaided condition to 80% in the binaural aided condition and at age 7, from 0% in the unaided condition to 95% in the binaural condition. Closed-set speech perception was also tested using the Khomsi test, designed for French-speaking individuals to assess spoken language comprehension. Following one year of hearing instrument use (at age 5), the child gave no score, failing the test. She was again assessed using this test at age 7, the child's results indicated her speech comprehension was at the level of a 5 year old. The results indicate that conventional amplification can benefit at least some cases of ANSD.

Retrospective Case-Series (post-test only)

Lee et al. (2001) provided results for two children attending a school for the deaf who presented with transient otoacoustic emissions with absent or severely distorted ABRs.

The first child was 12 years old and had been identified with severe hearing loss at one year of age, with no ANSD risk factors, and was fit with amplification shortly after. Cantonese speech discrimination scores were found to be poor in both the unaided and aided conditions at 20% and 30%, respectively. The child's mother reported that he disliked wearing his hearing aids and complained that they were too loud.

The second child was 11 years old and had been identified with a profound loss in his right ear and a moderate sensorineural hearing loss in his left ear at 17 months of age, with no ANSD risk factors. The child was fit with hearing instruments shortly after. Auditory neuropathy spectrum disorder was only suspected in this child's right ear. Cantonese speech discrimination scores were poor in both the unaided and aided conditions in the left ear at 20% and 10%, respectively. Word discrimination was not possible in the right ear. The child's mother reported that he very rarely wears his hearing instruments and that he also finds the sound too loud. The prescriptive method used and fit to targets, were not disclosed in this article for either child.

Between Groups Nonrandomized Clinical Trial

Rance et al. (2009) evaluated the receptive language and speech production abilities in a group of children with auditory neuropathy spectrum disorder (n=20), presenting with an absent or abnormal auditory ABR and present cochlear microphonics and/or OAEs. Ten of these children were fitted with a cochlear implant, either binaurally or monoaurally, and the remaining ten were fit with binaural behind-the-ear (BTE) hearing instruments, based on the NAL prescriptive method. The two subject groups were compared against each other, as well as to a control group of implanted children with sensorineural hearing loss (n=10), matched based on chronological age at assessment and age at implantation.

The receptive language skills are assessed with the Peabody Picture Vocabulary Test III A (PPV IIIA), giving a language quotient value for each child, and speech production was assessed using the Diagnostic Evaluation of Articulation and Phonology (DEAP), giving each child a percentage of phonemes correct score. To account for age related differences, an age-referenced error rate was calculated for each child.

The language quotient values were lower than expected for a normally developing child, but were equivalent across all subject groups. Receptive language acquisition was found to be positively correlated with speech perception ability for both the ANSD implanted group (r= 0.767, P=0.01) and SN implanted group (r=0.684, P=0.029), but not for the aided AN group (P=0.781). Reasonable speech production skills were found in all subject groups, with the percentage of phonemes correct score exceeding 70% in all but one implanted AN subject, with a one-way ANOVA showing no significant difference across subject groups. These results indicate that the aided children in this study performed as well their implanted peers.

Between Groups Nonrandomized Clinical Trial #2

Rance et al. (2008) evaluated speech perception skills in two groups of children with auditory neuropathy spectrum disorder, managed with either hearings aids (n=20) or cochlear implants (n=20). All of these children presented with an absent ABR and present cochlear microphonics and/or OAEs. The implanted children were fit with a cochlear implant, either binaurally or monaurally after discovery of a severe/profound hearing loss or limited success with conventional amplification. The aided children had been fit with BTE hearing instruments based on the NAL prescriptive method, with all but one child aided at ≤10 months of age. A cohort of implanted children with sensorieneural hearing loss was matched to the test groups based on chronological age and the age at implantation.

Open-set speech perception skills were assessed in the sound field and videotaped to be phonetically transcribed to obtain a percentage phonemes-correct score. The DEAP was used to assess speech production scores, with all children obtaining greater than 80%, indicating that speech perception scores were true measures, not affected by production difficulties.

The aided ANSD children showed some variability in their findings, but 8/10 performed in the range expected for those with sensorineural hearing loss. A one-way ANOVA was performed to show a significant group effect (F = 6.34, p+0.006) and a post hoc analysis (Tukey) demonstrated that the implanted SN group performed significantly above the two ANSD groups, but no difference was found between the two ANSD groups. The effects of other external factors (i.e. CNC phoneme score and age at assessment, age at device fitting, or duration of device use) were ruled out via a regression analysis.

Case-Control Study

Rance et al. (2007) examined speech reception and production skills in 12 ANSD children who were hearing instrument users. The ANSD subjects all presented with absent auditory brainstem responses with present cochlear microphonics and/or OAEs. All subjects were diagnosed by 32 months and the mean age of hearing instrument fitting was 9 months, with a range of 4 to 34 months. At the time of assessment, all of the subjects (except two) were consistent hearing instrument users and had been for over 4 years. A control group of 12 aided children with SN hearing loss were matched based on chronological age and 3-frequency average hearing level.

Receptive language was assessed using the Peabody Picture Vocabulary Test III A and speech production was assessed using the Diagnostic Evaluation of Articulation and Phonology. The study found that language reception skills were delayed for those with ANSD (as well as for the SN group) as compared to test norms, but not in correlation with their behavioural thresholds (r = -0.17, p = 0.59; r = -0.51, p = 0.09, respectively). A paired t-test was done to compare the AN and SN subject groups, but no significant difference was found (t = -0.58, p=0.57). The speech production abilities varied within the ANSD group, but no significant difference was found from the SN group when a paired t-test was conducted. These results indicate that aided children with ANSD can find success with conventional amplification, with speech reception and production skills equivalent to the SN control group.

Mixed (Between & Within) Nonrandomized Clinical Trial

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 PBK score ($r = 0.01$, $p = 0.78$), or between aided articulation index ratio and PBK score ($r = 0.14$, $p = 0.18$). In the 50% of subjects in whom improvement was seen, there was not a significant difference in speech perception scores from the SN group, although no statistical comparisons were reported. These results indicate that at least some children with ANSD can benefit from conventional amplification.

Retrospective Single Group Study

Rance et al. (1999) provided a retrospective overview of 20 cases of infants with ANSD who demonstrated cochlear microphonics in the absence of ABRs. Fifteen of these cases had been fit with binaural BTE hearing instruments, using the NAL prescriptive method, and had been consistent users for over 12 months at the time of speech perception testing. Of the fifteen, only 8 were able to complete speech perception testing in both the aided and unaided conditions, due to immaturity or generalized under-development.

Of the eight, four children showed significant improvement in the aided condition over the unaided condition ($p < 0.01$). The other 4 children scored equally poor in both test conditions, suggesting no amplification benefit. A nonparametric, Mann-Whitney) test was performed to assess the relationship between unaided pure-tone thresholds and hearing aid benefit, but showed no significant difference between the children who showed improvement and those who did not. Therefore, the results show that some children with ANSD can derive benefit from amplification, but this benefit cannot be predicted from behavioural thresholds.

Retrospective Case-Series Study

Raveh et al. (2006) provide retrospective case information for 19 children with auditory neuropathy, as diagnosed by absent or distorted ABRs with present OAEs, who were fitted with hearing amplification. Mean age at diagnosis was 13 months and mean age at hearing instrument fitting was not disclosed.

Of the 19 fit with hearing aids, only one displayed very good auditory and speech performance when aided. Fourteen of the children did not obtain performance improvement in the aided condition and four are still in the rehabilitation process or have failed to comply with the treatment protocol. The speech perception assessment protocols were not disclosed. It was reported that these subjects had difficulties understanding speech, poor word recognition scores, and in older children, low discrimination levels or a severe drop in discrimination level in the presence of background noise was found. The researchers conclude that conventional amplification is not a successful habilitation method due to the poor word recognition abilities found in those with auditory neuropathy.

Discussion

Overall there are few studies directly examining the success of hearing amplification in the treatment of children with ANSD. Most that do exist have small sample sizes and are retrospective in nature. The results are mixed, with six out of eight studies finding moderate success in the treatment of ANSD with hearing amplification (50-55% of subjects) and two studies finding little to no improvement in speech perception measures following habilitation with hearing aids.

The small sample sizes and retrospective nature of these studies decrease the ability to draw generalizable conclusions from this limited evidence. However, a small sample size can be partially justified due to the small population of children with ANSD that are aided.

Many of the studies with larger success rates (Rance et al. (2009)(2008)(2007)(2002)(1999)) only included the higher functioning aid users in their amplification population, with the poorer ones being implanted. At this time, a trial with hearing amplification is mandatory when determining cochlear implant candidature. In the above studies, those that performed poorly with hearing amplification, as determined by monosyllabic word testing results (<40% in aided condition), were automatically implanted. This can create a bias towards improvement in aided speech perception results in the

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 included studies were not designed to directly assess this issue. Therefore the ability to obtain a true measure of the success of aiding children with ANSD cannot be obtained from this review.

A treatment option other than irreversible cochlear implants or manual communication is the reason for examining hearing amplification success in children with ANSD.

The issue that needs to be taken into consideration by clinicians is at what age it is appropriate to begin habilitation for these children. Hearing thresholds are not predictable by ABR or ASSR in those with ANSD, so later fittings often result once behavioural thresholds can be obtained, increasing the amount of time possibly spent without language input in this sensitive period for acquisition of language. Concern over OHC damage, resulting from fitting hearing amplification when OAEs are still present, is often a cause of delay in clinicians when fitting children with amplification. These age-sensitive issues need to be assessed in future research in order to establish a more efficient treatment protocol for these children.

Future studies should be performed with larger sample sizes and unbiased hearing instrument populations. Future research should also evaluate more appropriate hearing amplification or cochlear implantation criteria for these children to ensure access to language at an earlier age.

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