Literature Review of Autism and Deafness

Individuals with a dual-diagnosis of an autism spectrum disorder and hearing loss constitute a population presenting unique considerations for audiologists regarding assessment and intervention. This review explores the prevalence of co-morbid hearing loss and autism, characteristics of auditory processes in autistic individuals, behavioral and electrophysiological methods of evaluation, and clinical issues arising from the co-occurrence of autism and hearing loss.

Although anecdotal reports have noted an increased prevalence of hearing loss among the autistic population, there is little certainty pertaining to their accuracy. One inherent problem with the identification of the incidence of co-occurring autism and hearing loss is the ambiguity surrounding the diagnosis of autism itself. More recently, an increased awareness of a broader spectrum of autism diagnoses has led to a greater number of autism diagnoses, with estimates of .3-.6% (Rutter, 2005). At least one study, however, has pointed to a higher incidence of hearing loss in the autistic population compared to the general population. Rosenhall et al (1999) found incidence of permanent mild and permanent bilateral moderate to profound hearing loss of 7.9% and 3.5%, respectively, in a group of children diagnosed with autism spectrum disorder. In addition, a review of studies suggested the presence of autism in 5.3% of children with hearing loss (Hitoglou et al, 2010). Furthermore, the presence of hearing loss appeared to be evenly distributed across the spectrum of autism diagnoses, suggesting that no correlation exists between the occurrence of hearing loss and the severity of the autism diagnosis (Jure et al, 1991; Rosenhall et al, 1999).

Auditory dysfunction among autistic individuals has been found to include both sensorineural and conductive pathologies (Rosenhall et al, 1999). Another often-reported clinical presentation of autism is frequent hypersensitivity to sound. Experimental psychophysical evidence has also supported a decreased dynamic range and abnormal loudness growth in individuals with autism when compared to typically-developing age-matched controls (Khalfa et al, 2004). An interesting finding was noted by Bonnel et al (2003) regarding the enhanced sensitivity to pitch as measured by the superior performance of autistic individuals compared with typically-developing controls on tasks of pitch discrimination and categorization. Electrophysiological assessment also lends support to this notion, although this advantage was noted to disappear with regard to speech stimuli (Lepisto et al, 2008). Various electrophysiological measures have also been evaluated in an attempt to identify neurophysiological differences between typically-developing children and those with autism. Although their results have been generally insignificant, variable, or inconsistent, certain late components of cortical auditory evoked potentials have shown promise for the potential to explain underlying auditory processes in autistic individuals (Bomba & Pang, 2004; Tharpe et al, 2006).

A variety of instruments have been designed to screen for autism, including the Autism Screening Instrument for Educational Planning (ASI-EP), the screening tool for autism in two-year-olds (STAT), and social communication questionnaire (SCQ). One study using two components of the ASI-EP found the instrument incapable of distinguishing the behavior of deaf- and normal-hearing autistic children (Roper et al, 2003). Although the STAT has been shown to be highly sensitive and moderately specific for children as young as 14 months (Stone et al, 2008), its efficacy for distinguishing deaf/heard of hearing and normal-hearing autistic children has not been ascertained. Likewise, the SCQ has demonstrated high sensitivity for autism in young children, though its specificity has been variable (Allen et al, 2007; Wiggins et al, 2007). It should be noted that participants of these studies had been referred for early intervention services due to developmental concerns from the caregiver.

Because the disruption of language and communicative function that is often noted in individuals with autism may present similarly to deficits found in children with more severe hearing loss, the differentiation of these two populations is challenging. Even more difficulty arises with the need to separately identify individuals with co-occurring hearing loss and autism and those with a lone diagnosis of hearing loss. As is evident from the aforementioned studies, most of the existing research explores identifiable differences in auditory perception between autistic and typically-developing children. However, an innovative approach to distinguishing deaf/hard of hearing individuals from deaf/hard of hearing autistic individuals may be adapted from findings regarding the visual attention of autistic children. Landry and Bryson (2004) found significant and clinically-identifiable differences in the reaction time to disengage or shift visual attention from a fixating stimulus among groups of autistic children, children with Downs' syndrome, and typically-developing children.

Findings from the numerous studies aiming to identify behaviors and neurophysiological differences aiming to distinguish autistic populations are variable; however, many of the conclusions point to the single idea that effective assessment should include a batter of multiple evaluative measures. In summary, the general lack of experimental evidence regarding the dual diagnosis of hearing loss and autism illustrates both the uncertainty with current clinical practices and the glaring need for focused research in this realm.

REFERENCES

Allen, C.W., Silove, N., Williams, K., & Hutchins, P. (2007). Validity of the social communication questionnaire in assessing risk of autism in preschool children with developmental problems. *Journal of Autism and Developmental Disorders*, 37, 1272-1278.

The authors examined the sensitivity and specificity of the social communication questionnaire for autism spectrum disorders for children aged 2-6. The SCQ was completed by 81 parents of children who were considered at high risk for developmental problems. Using a cut-off score of 11, the sensitivity and specificity for children aged 2-6 were 93% and 58%, respectively. For children aged 3-5, the authors noted a sensitivity of 100% and specificity of 62%. The authors concluded that the low specificity precludes the use of the SCQ as a diagnostic tool, but it may be employed as an assistive tool for identifying children who may require a more comprehensive autism assessment.

Bomba, M.D. & Pang, E.W. (2004). Cortical auditory evoked potentials in autism: a review. *International Journal of Psychophysiology*, 53, 161-169.

The authors provide a review of research investigating the role of auditory-evoked potentials in the study and assessment of autism and discuss the implications for future research. Notable conclusions of the review suggest that great variability in findings from electrophysiological studies preclude definitive differential diagnosis solely from the use of such tests. However, further exploration of electrophysiological measures, particularly the late waveform components, combined with an increased understanding of the spectrum of autism diagnoses may provide critical information about the auditory processing characteristics of autistic individuals.

Bonnel, A., Mottron, L., Peretz, I., Trudel, M., Gallun, E., & Bonnel, A.M. (2003). Enhanced pitch sensitivity in individuals with autism: a signal detection analysis. *Journal of Cognitive Neuroscience*, 15(2), 226-235.

The study employed a signal detection methodology to compare the performance of autistic individuals with typically-developing controls for tasks of pitch discrimination and pitch categorization. In the pitch discrimination task, the participants were asked to identify the presentation of a pair of tones as being identical or different. In the pitch categorization task, participants judged the pitch of presented tones to be high or low with respect to previously-presented tones. Results of the study indicated that the performance of autistic individuals was significantly superior on both pitch processing tasks. Furthermore, there was no difference between their performances for each task, while the typically-developing group performed more poorly for categorization than discrimination. The authors conclude that autistic individuals possess superior pitch processing abilities and hypothesize that the underlying memory processes used by these individuals may be similar regardless of the task.

Filipek, P.A., Accardo, P.J., Ashwal, S., Baranek, G.T., Cook, Jr., E.H., Dawson, G., et al (2000). Practice parameter: Screening and diagnosis of autism. *Neurology*, 55, 468-479.

The article discusses the current criteria for identifying and diagnosing autism spectrum disorders and the empirical evidence in which they are based. The authors also provide recommendations for a dual process approach of routine developmental observation to identify children at-risk for any developmental disorders and those specifically at-risk for autism spectrum disordes and, secondly, to identify aspects distinguishing autism from other developmental disorders.

Gravel, J.S., Dunn, M., Lee, W.W., & Ellis, M.A. (2006). Peripheral audition of children on the autistic spectrum. *Ear & Hearing*, 27(3), 299-312.

Hitoglou, M., Ververi, A., Antoniadis, A., & Zafeiriou, D.I. (2010). Childhood autism and auditory system abnormalities. *Pediatric Neurology*, 42(5), 309-314.

The authors review the occurrence of auditory dysfunction among children with autism. In addition to discussing prevalence of hearing loss and notable auditory characteristics, the review explores the results of studies of the underlying neurophysiologic processes of audition among autistic individuals.

Jure, R., Rapin, I., & Tuchman, R.F. (1991). Hearing-impaired autistic children. *Developmental Medicine & Child Neurology*, 33(12), 1062-1072.

The authors reviewed audiological information for a group of 46 children diagnosed with both autism and hearing loss. The authors concluded that while the severity of the autism diagnosis was related to the cognitive dysfunction, it was unrelated to the severity of the hearing loss. Furthermore, the review noted that almost 25% of the children remained undiagnosed with autism for a period of at least 4 years.

Khalfa, S., Bruneau, N., Roge, B., Georgieff, N., Veuillet, E., Adrien, J.L., Barthelemy, C., & Collet, L. (2001). Peripheral auditory asymmetry in infantile autism. *European Journal of Neuroscience*, 13, 628-632.

The study explored the contralateral suppression of otoacoustic emissions in individuals with autism and age- and gender-matched typically-developing individuals. Suppression of otoacoustic emissions is hypothesized to result from activity of the efferent medial-olivo-cochlear pathway and is thought to be involved in the filtering of background noise. Results of the study demonstrated differing patterns of asymmetry between groups as well as an age-related decrease in otoacoustic emission amplitude among autistic individuals. The authors conclude that the findings may suggest involvement of higher-level processes as well as an age-related decrease in outer hair cell function among autistic individuals.

Khalfa, S., Bruneau, N., Roge, B., Georgieff, N., Veuillet, E., Adrien, J.L., Barthelemy, C., & Collet, L. (2004). Increased perception of loudness in autism. *Hearing Research*, 198, 87-92.

The authors investigated previously-noted findings of increased sensitivity to sound among autistic individuals. The study tested the dynamic range for pure tones of varying frequencies and also measured loudness growth function of a 1000 Hz tone using a categoral loudness scaling procedure. Trends of loudness perception in autistic individuals was compared with those of typically-developing controls. Results demonstrated a reduced dynamic range and a steeper loudness growth function in autistic individuals, suggesting an increased perception of loudness in these individuals.

Landry, R. & Bryson, S.E. (2004). Impaired disengagement of attention in young children with autism. *Journal of Child Psychology and Psychiatry*, 45(6), 1115-1122.

The study examined the disengagement and shift of visual attention in young children with autism. The authors compared the performance of autistic children with a group of typically-developing children and a group of children with Downs' syndrome on two tasks. In the first task (Shift), a visual stimulus was first presented in front of the child before being turned off and followed immediately by a visual stimulus presented in the child's peripheral visual field. In the second task (Disengage), the central fixation stimulus remained present as the secondary peripheral stimulus was presented. The latency for visual tracking of the peripheral stimulus was the dependent measure. The results demonstated no significant differences among the groups for the shift trials; however, reaction time latency for the disengagement task was significantly longer for the autistic children than for either the typically-developing group or Downs' syndrome group. These findings implicate a possible role for the assessment of disengagement of visual attention in differentiating children with autism spectrum disorders from their peers.

Lepisto, T., Kajander, M., Vanhala, R., Alku, P., Huotilainen, M., Naatanen, R., & Kujala, T. (2008). The perception of invariant speech features in children with autism. *Biological Psychology*, 77, 25-31.

The authors implemented the mismatch negativity response (MMN) as an electrophysiological measure of auditory processing of phoneme-category changes and pitch changes to speech stimuli with constant- and varying-feature standard/deviant stimuli in children with autism and typically-developing controls. Results demonstrated that although children with autism had robust MMN responses for both pitch and phoneme-category changes, the MMN response was significantly diminished when pitch variation of phoneme-category changes was introduced. The authors conclude that the enhanced pitch processing of the autistic children may produce a deleterious effect when stimuli are speech-like in nature.

Roper, L., Arnold, P., & Monteiro, B. (2003). Co-occurrence of autism and deafness: diagnostic considerations. *Autism*, 7(3), 245-253.

The study investigated the efficacy of two screening instruments and a non-standardized parental questionnaire for distinguishing deaf autistic, normal-hearing autistic, and deaf learning-disabled individuals. The Autism Behavior Checklist (ABC) and Interaction Assessment (IA), two components of the Autism Screening Instrument for Educational Planning, were administered to participants of the study, and the parental questionnaire was distributed to parents. Although scoring of the ABC and IA resulted in differences between the deaf learning-disabled group and normal-hearing autistic group and the deaf learning-disabled group and deaf autistic group, no significant differences existed between the deaf and normal-hearing autistic groups. Questionnaire results indicated that parents of deaf autistic children suspected developmental problems at an earlier age than parents of hearing autistic children, and deaf autistic children were diagnosed at a later age than hearing autistic children.

Rosenhall, U., Nordin, V., Sandstrom, M., Ahlsen, G., & Gillberg, C. (1999). Autism and hearing loss. *Journal of Autism and Developmental Disorders*, 29(5), 349-357.

The authors investigated the prevalence of hearing loss among a group of 199 children and adolescents diagnosed with an autistic spectrum disorder. Findings of the study demonstrated the prevalence of permanent mild hearing loss and permanent bilateral moderate-profound hearing loss to be 7.9% and 3.5%, respectively. Furthermore, the occurrence of hearing loss was evenly-distributed among the spectrum of low- to high-intellectually functioning individuals, suggesting that the presence of hearing loss is unrelated to the severity of the autistic disorder.

Rutter, M. (2005). Incidence of autism spectrum disorders: changes over time and their meaning. *Acta Paediatrica*, 94(1), 2-15.

The author provides statistical data regarding the incidence of autism spectrum disorders and discusses the implications and possible reasons for the increase in incidence rates over time that have been noted by other studies. The author concludes that the prevalence of autism is likely 30-60 per 10,000 and that the increased prevalence is largely a result of improved methods of assessment as well as the recognition of a broader spectrum of autism.

Stone, W.L., McMahon, C.R., & Henderson, L.M. (2008). Use of the screening tool for autism in two-year-olds (STAT) for children under 24 months: an explanatory study. *Autism*, 12(5), 557-573.

The authors explored the utility of the STAT as a screening tool for children aged 12 to 23 months. The 71 children participating in the study either had an older sibling with autism spectrum diagnosis or were referred due to concerns of possible autism. Using a modified cut-off score, the findings of the study noted a sensitivity of .95 and specificity of .73 for the sample of children. False positives were also noted to be higher for 12- and 13-month-olds and for

children who were later diagnosed with another developmental disorder. The authors concluded that the STAT provides clinical value for assessing children as young as 14 months, although considerations should be made to the modification of the diagnostic criterion cut-off.

Tharpe, A.M., Bess, F.H., Sladen, D.P., Schissel, H., Couch, S., & Schery, T. (2006). Auditory characteristics of children with autism. *Ear & Hearing*, 24(7), 430-441.

The study investigated the auditory characteristics of typically-developing children and children diagnosed with autism as measured by behavioral and physiologic testing. Results indicated no group differences with regard to physiologic tests (auditory brainstem response, distortion product otoacoustic emissions, and acoustic reflex). However, the authors noted significant differences for behavioral test measures. While all typically-developing children were within normal limits for pure-tone thresholds, approximately half of the autistic children presented with elevated pure-tone thresholds greater than 20 dB HL. Furthermore, test-retest reliability for behavioral thresholds for the typically-developing children was generally 10 dB or less, while that of the autistic children was usually 15 dB or greater.

Wiggins, L.D., Bakeman, R., Adamson, L.B., & Robins, D.L. (2007). Utility of the social communication questionnaire in screening for autism in children referred for early intervention. *Focus on Autism and Other Developmental Disabilities*, 22(1), 33-38.

The authors examined the validity of the social communication questionnaire (SCQ) as a screening instrument for autism spectrum disorders in children younger than 4 years old. Participants in the study were 37 children aged 17 to 45 months who were classified into either an autism spectrum disorder group of developmental disabilities group. Using the recommended cut-off score (15), the study noted the SCQ to have a sensitivity of .47 and specificity of .89. The maximum rates for both sensitivity and specificity (.89 and .89) were achieved with a cut-off score of 11. The authors concluded that use of the SCQ with the modified cut-off score of 11 holds clinical value in identifying younger children who are at-risk for a diagnosis of autistic spectrum disorders.