Hyperacusis and autism spectrum disorder

BY ALI A DANESH AND JESSICA KAPLAN

Several different auditory deficits have been found to be co-morbidities of ASD. This article reviews literature with respect to the relationship between hyperacusis and ASD.

Autism spectrum disorder (ASD) can be characterised as a neurodevelopmental condition that is marked by impairments in social interaction, communication, and behavioural challenges. In children with ASD, it has been documented that auditory deficits can be comorbid with their diagnosis [1]. These include hearing loss, tinnitus, auditory processing disorder, and hypersensitivity to increased levels of sounds [2]. Individuals who are diagnosed with ASD, in some cases, experience decreased sound tolerance (DST) disorder, often referred to as auditory hypersensitivity or hyperacusis. Hyperacusis is a known disorder in which individuals experience a reduced sound tolerance, with heightened complaints and emotional reactions to typical auditory stimuli, being perceived as too loud and uncomfortable, causing high levels of distress and irritability. In contrast, in neurotypical individuals, these sounds would not cause a negative reaction or a high level of discomfort. Individuals with hyperacusis typically have normal functioning auditory thresholds but have a reduced threshold for loudness discomfort levels (LDLs), leading sounds with moderate intensity to be perceived as very loud [3]. Those who have a diagnosis of ASD can experience these auditory stimuli at a higher level of discomfort. Hyperacusis can impose limitations on one's ability to communicate during social interactions and decrease a person's ability to comprehend speech in loud environments. More specifically, in individuals with ASD these symptoms become exacerbated and may hinder their ability to function [2].

Literature on the aetiology and pathophysiology of hyperacusis is still medically undefined and there is a sufficient lack of evidence-based recommendations on how to diagnosis and provide treatment in those with this disorder [1], particularly for individuals with ASD. Although there is information lacking, there is correlation between hyperacusis and the ASD population. As stated previously, individuals with autism have difficulty with social aspects of communication, language development, and behavioural challenges. Along with these factors, the ASD population also displays hyperreactivity to a variety of sensory stimuli which has been noted as a core characteristic in the *Diagnostic and Statistical Manual of Mental Disorders*, *Fifth Edition* (DSM-5) [4]. Studies have reported that the prevalence rate of DST in the ASD population is 86% [1,5,6]. Through an online survey study by Law and colleagues, it was documented that a prevalence of auditory hypersensitivity in a sample of 814 children with ASD was 77.6% to 86.6% [6]. When assessed using psychoacoustic tests and behavioural observations, prevalence rates tended to be lower. Rosenhall et al noted prevalence rates to be 18% based on the inability to tolerate sounds at 80 dB HL using broadband clicks [7], and Demopoulos and Lewine reported a hyperacusis prevalence rate of 37% when using speech loudness discomfort levels (LDLs) [8]. In a systematic review and meta-analysis by Williams and colleagues, it was observed that the prevalence of hyperacusis was 41.42% when assessed using an interview or questionnaire and 27.3% when assessed using an observational or objective measure [1]. The results of their study suggested that 2.4–3 million individuals with ASD in the United States experience symptoms of hyperacusis. The impacts of hyperacusis on this population is severe, reducing their engagement in nearly all daily activities including family, school, and social activities. Not only does it impact the individual child; it also places an increased burden on their caregivers, as well as placing limitations for future employment and quality of life in adulthood [1].

Neurophysiological research has provided insights into the atypical neural processing of auditory information in individuals with ASD. For example, a study by Ohmura and colleagues examined whether inner ear function could give reasons to hyperacusis in ASD through examination of stapedial reflex (SR) thresholds and distortion product otoacoustic emissions (DPOAE) [9]. Using a hyperacusis index related to SR thresholds and DPOAE suppressions in an ASD group and control group, the results showed that only SR thresholds were correlated with the hyperacusis index in both groups. The SR thresholds in the ASD group were found to be lower than in the control group. According to their results, the researchers suggested that hyperacusis in ASD can be explained by high sensitivity of inner ear function. Another study by McCullagh et al looked at the effects of *fragile X factor 1 (FMR1)*

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gene deletion in the auditory pathway in humans and animal models [10]. The study examined the auditory system in fragile X syndrome by looking at the fragile X factor protein (FMRP). The results exhibited FMRP deficiency effects on the auditory system at various stages on the auditory ascending pathway, beginning at the spiral ganglion and first nuclei in the brainstem and leading all the way to the auditory cortex. The involvement of multiple stages along the auditory pathway strongly supports the concept that fragile X syndrome is not only a cognitive condition but also changes the essential sensory information processing that is necessary for an individual to perceive, separate and localise sounds, which suggests that hypersensitivity in fragile X syndrome is due to FMRP deficiency. This study can provide further insight into the neurophysiological cause of hyperacusis/DST in the ASD population, as *FMR1* is the most prevalent monogenic cause of ASD. Additionally, Danesh and Kaf, investigated the DPOAE characteristics of children with autism compared to a control group [11]. The researchers suggest that hypersensitivity in the autistic population could be a result of cochlear dysfunction and abnormal efferent auditory pathways due to the lack of succinct contralateral suppression. The neurophysiological research supporting the underlying cause of DST and hyperacusis in individuals with ASD is continued to be widely explored but remains unknown and warrants further research.

Diagnosis of hyperacusis in the ASD population proposes significant challenges due to their underlying characteristics, as it is difficult for autistic individuals to verbally express their symptoms, thus relying on caregiver questionnaires and negative reactions to auditory stimuli to determine an accurate diagnosis. Scheerer et al utilised an altered version of the Auditory Sensitivity and Child Safety Questionnaire, which was given to caregivers of autistic children, that included questions relating to onset, development, negative reactions/behaviours, types of sound eliciting reactions, coping mechanisms to manage DST and impact of DST on the child’s quality of life [3]. Parent reports of 88 ASD children in this study exhibited that DST began between the ages of one year and 10 years old, with 87.5% of parents still reporting hypersensitivity to sound as a current issue.

Regarding treatment of hyperacusis in the ASD population, it is very limited and further research is needed. Danesh and colleagues reviewed three different treatment approaches for hyperacusis in ASD: (1) habituation training, (2) cognitive behavioural therapy, and (3) auditory integration training [12]. Habituation training essentially consists of desensitisation through retraining the emotional and non-classical auditory pathways to decrease fear response to sounds. According to the researchers, this approach is extremely effective in the reduction of negative reactions to sounds in individuals with ASD. Currently, there are no studies that demonstrate the efficacy of cognitive behavioural therapy and auditory integration therapy for individuals with ASD. Another study by Pfeiffer and colleagues suggests that noise-attenuated headphones may be a possible treatment method for reducing nervous system sympathetic reactivity for hyperacusis in individuals with autism [13]. However, the authors of this article question this approach, as avoiding sound can enhance the central gain in the brain and impact the intolerance to the sounds due to the lack of proper exposure.

In this review, we aimed to address the relationship that currently exists in literature of hyperacusis in the ASD population. It should be noted that there is much research to be done to further define the aetiology, neurophysiology, prevalence, diagnosis, and treatment for hyperacusis in this population as it currently remains undefined. Future research in these areas will provide resources to improve the quality of life of individuals with ASD as hyperacusis has significant effects on social aspects of their life.

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