Hyperacusis in Autism Spectrum Disorders

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Contributor: Wafaa Kaf

Decreased Sound Tolerance Disorders (DSTD) are routinely observed in autism spectrum disorder (ASD). The most common types of DSTD are hyperacusis and misophonia. Hyperacusis is a class of decreased sound tolerance disorders in which a negative or incongruous reaction is triggered from exposure to sounds that are not described as threatening or uncomfortable by a neurotypical individual. These reactions are in response to general sounds, rather than specific sounds (such as chewing and sniffling), as would be the case with misophonia. Hyperacusis can affect an individual at various degrees depending on the severity. It can impact one's emotional wellbeing, sleep, concentration, and can cause anxiety.

Keywords: audiology ; auditory system ; decreased sound tolerance disorder ; hyperacusis ; misophonia ; noise sensitivity ; tinnitus ; autism spectrum disorder

1. Hyperacusis: Assessment and Prevalence

1.1. Assessment of Hyperacusis

Assessment of hyperacusis typically will involve extensive case history taking, pure tone audiometry, measurement of uncomfortable loudness levels (ULLs), and self-report questionnaires such as the hyperacusis questionnaire (HQ) [1][2][3]. However, due to limitations in obtaining accurate levels of loudness discomfort or sometimes hearing thresholds, particularly in severe cases of ASD, clinicians rely on behavioral observation strategies and case history. Generally, ULL provides the level above which tones become uncomfortably loud for a patient [2]. In patients with hyperacusis, ULLs will typically be lower than the average person with normal hearing and without hyperacusis. The average ULL for patients with normal hearing and without hyperacusis is around 100 dB hearing level (HL), and reports of ULLs for patients with hyperacusis have been reported to be around 60 to 85 dB HL [1][2].

Based on results obtained with adults, Aazh and Moore [4] proposed diagnostic criteria for hyperacusis based on the average ULL across 0.25, 0.5, 1, 2, 4 and 8 kHz for the ear with the lower average ULL, which is denoted ULLmin. They suggested that a value of ULLmin equal to or below 77 dB HL should be taken as indicating the presence of hyperacusis [4]. With this ULLmin criterion, 95% of adult patients diagnosed as having hyperacusis were found also to meet the criterion of a cut-off score on the Hyperacusis Questionnaire (HQ) [3] of 22 or more [4]. Interestingly, among children and adolescents seeking help for tinnitus and/or hyperacusis from an audiology clinic, the mean value of ULLmin was 64 dB HL (SD = 15, n = 34). [4]

1.2. Prevalence of Hyperacusis and Concomitant Diagnoses

A number of studies have analyzed the prevalence of hyperacusis in the general population [3][4]. These findings have ranged from reports of 3.2% up to 17.1% [3][4]. It has been found that hyperacusis often accompanies other medical conditions [4][5]. These diagnoses include a high number of psychiatric conditions such as post-traumatic stress disorder (PTSD), depression, and exhaustion syndrome as well as migraines, tinnitus, hearing loss, attention deficit hyperactivity disorder (ADHD), and autism spectrum disorder (ASD). In a retrospective study analyzing case notes of 61 children with hyperacusis, it was found that 28 of the children, or 46% of the sample, had a concomitant neurodevelopmental condition, with the most common diagnosis being autism spectrum disorder [5]. This is not surprising, as it is known that sound hypersensitivity is a common component of ASD.

A pilot study in 1995 reported a 40% prevalence of hyperacusis in the ASD population [6]. More recently, Demopoulos and Lewine analyzed the audiometric profiles of 60 autistic children and adolescents ages 5 through 18 [7]. Comprehensive assessment revealed 37% of the participants to have sound sensitivity in at least one ear [7]. Rosenhall et al., 1999, studied the auditory characteristics such as hearing loss and hyperacusis of 199 children and adolescents with ASD and reported 18% prevalence for hyperacusis in their study sample [8]. Danesh and colleagues conducted a study to evaluate the presence of tinnitus and hyperacusis in individuals between the ages of 4 and 42 diagnosed with Asperger's

Syndrome [9]. It should be noted that Asperger's Syndrome is a diagnosis that is no longer being used and has since become part of the ASD diagnosis [10]. Danesh, et al study used a home developed case history survey as well as the Tinnitus Reaction Questionnaire [11], Tinnitus Handicap Inventory [12], and Hyperacusis Questionnaire [4] to survey 55 participants. These questionnaires found that the ASD group had a much higher prevalence of both hyperacusis and tinnitus than the general population. Within this study group of ASD participants, 69% reported hyperacusis with an average Hyperacusis Questionnaire score of 20.7 [9]. In a recent metanalysis of hyperacusis in individuals with ASD, the researchers concluded that hyperacusis has a high prevalence across the life span of this population [10].

2. Hyperacusis and ASD

2.1. Autism Spectrum Disorder and Hyperacusis

Autism spectrum disorder (ASD) is a complex neurological and developmental condition that is characterized by a number of differences in the sensory, behavioral, language, and social domains [13][14]. ASD is referred to as a spectrum disorder because presentations of this diagnosis vary by individual. Most commonly discussed presentations include language delays, social/pragmatic differences, rigid and restricted interests and behaviors [14]. More recently, differences in attention and perceptual and sensory processing have been found to be central components to ASD as more research has found the role these differences play in communication [15]. Research on sensory profiles of autistic individuals has suggested increased sensory discrimination skills as well as increased distractibility [16]. Theories reviewed by Remington and Fairnie include the idea that autistic individuals have an increased perceptual capacity allowing them to process an increased amount of cognitive information as compared to neurotypical peers [17]. This includes visual information as well as auditory stimuli.

It is important to note the semantic preferences in the autism community have changed over the years as the neurodiversity movement has become increasingly widespread. Research demonstrates that many members of the autistic community most commonly prefer use of identity first language (autistic person), whereas allies and professionals working were more likely to use person first language (person with autism) [18]. It is important for professionals to understand and acknowledge the preferences of autistic individuals in order to help to facilitate a positive and inclusive environment.

Across a variety of sensory domains including auditory processing, autistic individuals may demonstrate hyper-reactivity or hypo-reactivity. It has been reported that hypersensitivity is perceived to be more of a problem impacting daily life than hyporeactivity [19]. Auditory processing differences in autistic individuals have been noted regarding sensory perception and processing of auditory stimuli. Perceptual differences have been found including superior perception of pitch, superior identification of musical notes, and superior local processing of auditory stimuli as compared to neurotypical adults [20][21]. Autistic individuals have been found to have intact global processing of auditory stimuli and reduced global interference, meaning they best process auditory stimuli at the musical note level as compared to processing a melody. In addition to enhanced abilities and differing levels of processing, autistic individuals also have been found to have difficulties with regard to auditory processing. Recent research by Vlaskamp and colleagues in 2017 has noted that autistic children demonstrate reduced mismatch negativity, suggesting that autistic children are less able to automatically encode deviant sounds [20]. In fact, auditory hypersensitivity is often a key indicator of ASD [19][21]. Hypersensitivity to auditory stimuli is exacerbated in the ASD population, resulting in sensory-based reactions such as covering one's ears, crying, or running away [21][22][23]. Such reactions may be perceived as pragmatically atypical may adversely impact social and academic function. A summary of the prevalence of hyperacusis in ASD population from four studies is shown in **Table 1**. A quick glance on **Table 1** shows that these percentages are more than double the prevalence of the general population.

Table 1. ASD Characteristics and Prevalence of Hyperacusis.

Participants	Age (Years)	Severity of ASD	Tool to Assess Hyperacusis	Hyperacusis%	
Rimland & Edelson (1995) [6]	**17 (11 males) (6 females)**	**4–21**	**Unspecified**	**Hearing Sensitivity Questionnaire was given to parents**	**Mild—53% Moderate—24% Strong—18%**
Rosenhall et al., (1999) [8]	**199 (153 males) (46 females)**	**Children Adolescents**	**Autism**	**Unspecified**	**18%**

Participants	Age (Years)	Severity of ASD	Tool to Assess Hyperacusis	Hyperacusis%	
Demopoulos & Lewine (2016) [7]	60 (48 males) (12 females)	5–18	High-low functioning	Unspecified	37%
Danesh et al., (2015) [9]	55 (46 males) (9 females)	4–42	High-functioning (Asperger)	Hyperacusis Questionnaire	69%

2.2. Correlates of Hyperacusis in the ASD Population

A number of proposed causes of hyperacusis have been introduced, but it is important to consider that correlates of hyperacusis may differ across individual cases of ASD and hyperacusis given the extreme level of variability in the makeup of the brain depending on the level of severity of the ASD. Smith, Storti, Lukose, and Kulesza [24] reported imaging studies that demonstrated cerebellar and brainstem hypoplasia in the ASD population compared to age matched people with neurotypical development, including hypoplasia of the facial nucleus and superior olivary complex. Another interesting study highlighting anatomical causes of hyperacusis in the ASD population noted that 29% of autistic people and hyperacusis were found to have superior semicircular canal dehiscence as demonstrated by computerized tomography imaging [25]. Due to this significant percentage of superior canal dehiscence within this specific population, this study was then elaborated to find that vestibular evoked myogenic potential (VEMP) demonstrated diagnostic ability to differentiate between hyperacusis due to superior canal dehiscence and dehiscence due to bone immaturity in autistic children [26]. A 2013 study found a delayed response of stapedial acoustic reflex in the ASD group, asserting that autistic patients can be identified using this measure [27]. Another recent study assessed the correlation of loudness tolerance with the stapedial reflex threshold with contralateral suppression of distortion product otoacoustic emissions (DPOAEs), as contralateral suppression of DPOAEs is typically increased in patients with hyperacusis [28]. Results indicated that stapedial reflex was lower in the ASD group and was significantly correlated with loudness tolerance in both the ASD and control groups. This is supported by previous findings by Danesh and Kaf [29], in which DPOAEs were found to have lower amplitudes in absence of noise in the ASD group, and with contralateral noise, the suppression effect was weaker in the ASD group indicating both cochlear and efferent system lesion. Kaf and Danesh [30] also studied contralateral suppression of DPOAEs to broad band noise in 18 autistic children with the primary diagnosis of Asperger's syndrome and 18 control group participants. Results showed no significant differences between groups for both DPOAE response signal to noise ratio with and without contralateral noise. The lack of significant differences may be because the study group was on the high-functioning end of the autism spectrum (e.g., Asperger). Kaf and Danesh [30] also suggested that the generation of hypersensitivity to sounds in high-functioning autistic children may be due to abnormal neural connections at proximal structures to the medical olivary complex, such as the temporal lobe, limbic system and autonomic nervous system. Several neuroimaging studies have shown abnormal connectivity in the brain of children with ASD such as enlarged brain volume, 10% enlargement of the white matter of the temporal lobe and corpus callosum and decreased neural cell size in ASD [31][32]. These brain abnormalities are age-specific, with faster growth in childhood [33] as well as gender specific [34]. Thus, differences in the study participants' ages and gender may explain lack of significant findings or contradictory findings in autistic children.

Other causes of hyperacusis within the ASD population have been attributed to reduced strength of the efferent pathway of the auditory system. Knowing that medial olivocochlear (MOC) reflex is stronger than average in neurotypical adults with hyperacusis, Wilson, Sadler, Hancock, Guinan, and Lichtenhan [35] assessed the MOC efferent reflex in autistic children using transient evoked OAE (TEOAE). Results indicated that the group of autistic participants with severe hyperacusis had MOC reflexes that were twice as strong as groups of neurotypical participants and groups of autistic participants without severe hyperacusis. Research by Ida-Eto, Hara, Ohkawara, and Narita suggested that auditory hypersensitivity in ASD may be linked to impairment of inhibitory processing of the auditory system [36]. This study found decreased expression of protein markers for inhibitory neurons (i.e., immunoreactivity) of the superior olivary complex secondary to decreased size of the medial nucleus of the trapezoid body; however, it is important to note that this was found using ASD model rats with prenatal exposure to thalidomide as opposed to transgenic animal models or autistic human subjects.

In addition to possible anatomical and physiological causes of hyperacusis, outside factors possibly contributing to hyperacusis in the autism population have been found in the literature, including a recent case study of an autistic 11 year-old boy, whose hyperacusis worsened when he was taking risperidone to treat hyperactivity and behavioral problems

[37]. However, a contrasting case study noted an autistic five and a half year-old girl whose hyperacusis, according to parent report, alleviated after taking the same medication [38]. Knowing the heightening effects hyperacusis has on existing sensory-based responses in ASD, more research is needed on the possible effects of supplements or medications and the role they play in hyperacusis of autistic people.

Genetics may also play a role in hyperacusis within the ASD population. Mertcati and colleagues [39] affirmed that contactin genes CNTN5 (contactin 5 is a protein coding gene) and CNTN6 (contactin 6 is also a protein coding gene) for neuronal cell adhesion molecules which promote neurite outgrowth in the sensory-motor pathways have been reported in autistic people. Reports included an autistic girl who had 5 copies of the CNTN5 gene, as well as multiple cases of deletion or mutation of the CNTN6 gene in other autistic individuals. Clinical investigation of patients carrying these CNTN5 and CNTN6 variants demonstrated the presence of hypersensitivity to sounds, and it was found that they had changes in wave latency of their ABR within the auditory pathway [33].

As previously noted, Remington and Fairnie directly assessed auditory capacity in autistic individuals [17]. They used behavioral experiments to examine the auditory processing profile of autistic individuals, with findings suggesting that autistic individuals have an increased auditory perceptual capacity compared to neurotypical individuals, which may result in sensory overload. This suggests that increased processing capacity may be the reason why autistic individuals are increasingly predisposed to hyperacusis. The authors proposed the reframing of perceptual processing of autistic individuals in terms of increased capacity as opposed to a deficit in filtering or processing sounds, in order to best develop interventions to alleviate distress in response to sensory stimuli including hyperacusis [17].

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