Providing support to school children with hyperacusis

Abstract

Hyperacusis is a condition associated with irritation and discomfort from environmental sounds. A person with hyperacusis is bothered by sounds that are generally acceptable to others. Hyperacusis has been reported with people various pathologies as well as for people with no hearing loss or health condition. It is a common condition in children, especially children with Williams Syndrome, autistic spectrum disorder and tinnitus. The exact mechanisms underlying hyperacusis are unknown. However, it has been hypothesised that it may be a consequence of abnormally high gain applied by the central auditory system. Adverse emotional and behavioural reactions to the various sounds in the classroom may be experienced by school age children with hyperacusis. Audiological assessment and management of hyperacusis are briefly discussed in this article.

Key words

■ Hyperacusis ■ Decreased sound tolerance
■ Hearing impairment ■ Uncomfortable loudness levels
■ Audiological assessment ■ Audiological management

Hyperacusis is described as abnormally sensitive hearing in which normally tolerable sounds are perceived as excessively loud (Stach, 1997). It is not associated with unusually low (good) thresholds for detecting sounds. In other words, the absolute threshold, as measured by the pure-tone audiogram, does not indicate hypersensitivity to very weak sounds. Rather, hyperacusis is manifested by aversive reactions to sounds that are clearly audible but would not normally lead to aversive reactions in most people. Several authors distinguish between hyperacusis (hypersensitivity to sound in general), misophonia (dislike of certain sounds) and phonophobia (fear of certain sounds) (Jastreboff and Jastreboff, 2006; Zarchi et al, 2010). However, the distinction of hyperacusis from phonophobia and misophonia is not well established in the literature (Andersson et al, 2002).

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Hyperacusis has been reported for patients with various pathologies or symptoms (e.g. facial paralysis, head trauma, acute acoustic trauma, tinnitus, stapedectomy, Ramsay Hunt syndrome, recruitment of loudness, depression, anxiety, Williams syndrome, learning disabilities, stuttering, spinal problems, Addison's disease, Lyme disease, and migraine) as well as for normally hearing people with no health condition (Katzenell and Segal, 2001). Hyperacusis can begin at any age (from less than 1 year of age to old age) (Gothelf et al, 2006; Zarchi et al, 2010). Hyperacusis can have a great impact on the daily life of the sufferers, and preclude them from social interactions and daily activities (Anari et al, 1999). Adverse emotional and behavioural reactions to various sounds in the classroom can be experienced by school age children with hyperacusis and are illustrated in the case study provided by Nicole Lowe, autistic spectrum disorder resource provision manager at Robert May's School (*Table 1*).

Prevalence of hyperacusis

The prevalence of hyperacusis in adults is estimated to be between 7% and 23% of the population (Jastreboff and Jastreboff, 2000; Andersson et al, 2002). The prevalence of hyperacusis in children is estimated to be between 12% and 27% (Klein et al, 1990; Khalfa et al, 2004). However, research has shown that hyperacusis is much more common in children with Williams syndrome, autistic spectrum disorder (ASD), and tinnitus than in the population as a whole.

Williams syndrome is a genetic condition characterized by distinct facial features, growth deficiency, aortic stenosis, and intellectual disability. The prevalence of hyperacusis in children with Williams syndrome is estimated to be between 84% and 100% (Gothelf et al, 2006; Klein et al, 1990). ASD is characterized by widespread abnormalities of social interactions and communication, as well as restricted interests. The prevalence of hyperacusis in children with ASD is estimated to be about 63% (Khalfa et al, 2004). Tinnitus is the perception of sound in the ears or the head that does not have an external source. Some degree of hyperacusis can be found in 40% of tinnitus patients (Jastreboff and Jastreboff, 2000). Tinnitus is reported to be present in 50% of children with hyperacusis (Coelho et al, 2007).

Table 2 lists the most common sounds that cause irritation and discomfort for children with hyperacusis. These sounds are usually well tolerated by children and adults without hyperacusis. However, children with

hyperacusis may have severe emotional and behavioural reactions when exposed to these sounds. *Table 3* lists reactions to sound in children with hyperacusis, as reported by various authors.

The aim of this paper is to briefly review the mechanisms, assessment and management of hyperacusis.

Mechanisms of hyperacusis

The mechanisms underlying hyperacusis are unknown. There are several theories that attempt to explain this condition. Hyperacusis sometimes occurs in people with hearing loss. For such cases, it has been hypothesised that the central auditory system provides abnormally high gain (amplification) to signals arriving from the cochlea, in order to compensate for the reduced input from the cochlea (Hall, 1998). Consistent with this idea, Formby et al (2003) showed that people reported sounds as being louder following 2 weeks of wearing bilateral earplugs (once the earplugs were removed). In a more recent study, Munro and Blount (2009) compared acoustic reflex thresholds (ART) for the two ears of adult listeners after use of a unilateral earplug. The ART is the lowest level of a stimulus at which the reflexive contraction of the middle ear muscles can be detected. Munro and Blount (2009) reported that ARTs were decreased in the ear that had been plugged for seven days, indicating increased sensitivity to sound.

The perceptual consequences of reduced ARTs and greater loudness are not clear. The increase in loudness after 14 days of ear plugging reported by Formby et al (2003) was equivalent to an increase in sound level of about 6 to 8 dB. The mean reduction in ARTs after 7 days of plugging of one ear was about 5 dB at 2 kHz and 7 dB at 4 kHz (Munro and Blount, 2009). It is not clear whether these changes are sufficient to account for the problems experienced by people with hyperacusis. In addition, there is not a one-to-one relationship between hyperacusis and hearing loss; many people with normal hearing have hyperacusis and many people with hearing loss do not experience hyperacusis. Nevertheless, the studies cited above are consistent with the idea that if the auditory system is somehow disturbed, this can lead to increased

Table 1. Case study

The student in this study is now in Year 10 and the difficulties he has encountered have remained largely unchanged despite the support and changes made to accommodate his needs. He has ASD and hyperacusis.

Everyday noises such as chairs on the ceiling above the room he is in, students laughing, and coughing or sneezing cause an immediate adverse reaction, similar to the fight/flight response. The student is particularly sensitive to the noise made by girls due to the pitch of their voices.

His usual response to sudden noises is to hit his head with his fist and if the noise continues, to scream and run away crying. At times he has thrown himself to the floor. He puts his fingers in his ears most of the time, which has resulted in the need for physiotherapy to help his posture and pain in his back. He is verbally aggressive towards both students and staff and has recently become physically aggressive.

The effect on the other students is that he often disturbs their learning by shouting at them to be quiet and he has disturbed the concentration of students in recent exams.

In an effort to ameliorate the effects of noise, the student leaves after the school bell has rung and arrives in class once the other students have settled in their places. He leaves early to avoid the busy corridors. He is able to work in the resource centre where it is quiet at any time in the day and completes all exams in there as well. He wears ear defenders when doing practical work in science and catering. The student is unable to attend lessons where he anticipates that there may be noise, and so he was removed from classes in music and drama and has followed an individual programme for physical education.

Table 2. Examples of common noises that cause irritation

Most common noises that cause irritation for children with hyperacusis	Reference
Dog bark, laughter	Hall, 1998
Screams, whistle, thunder, firecracker, truck, classroom noise, mixer, motorcycle, balloons, ambulance, school bell, dogs, car, musical instruments	Coelho et al, 2007
Electric drill	Zarchi et al, 2010
Vacuum cleaner, telephone, lawnmower, loud music, loud crowds	O'Reilly et al, 2000
Electric machines, thunder, bursting balloons, fireworks, siren, shouting, loud music	Gothelf et al, 2006

Table 3. Examples of reactions to sound

Reactions to sound in children with hyperacusis	Reference
Crying and escaping from sound source	Gothelf et al, 2006
Covering ears with both hands, crying and whining, making statements like 'it hurts my ears', cringing, hitting, throwing items, tearing books, and breaking pencils	O'Reilly et al, 2000
Making statements such as: 'when I hear the sound of an electric drill, I feel as if it is drilling into my body.'	Zarchi et al, 2010
Covering ears with their hands	Coelho et al, 2007
Crying and exaggerated startle response	Gothelf et al, 2006

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An example of a common reaction to sound from children with hyperacusis is covering the ears with both hands.

central gain, resulting in increased loudness of mid-level sounds and reduced ARTs, both of which could contribute to the symptoms of hyperacusis.

People experiencing hyperacusis typically have negative emotional associations with environmental noises or sound in general. These may sometimes arise as a result of an unpleasant event or experience occurring at the same time as or associated with sound, but such an event cannot always be identified. Once an association is set up, exposure to environmental or social noises can result in an over-activation of the limbic and autonomic nervous systems, causing a 'fight or flight' response which is itself unpleasant and reinforces the negative associations with sound (Jastreboff and Jastreboff, 2000).

Results reported by Hwang et al (2009) are consistent with this theory. They compared the brain activation of three patients with hyperacusis with that of three healthy subjects with no hyperacusis, using functional magnetic resonance imaging (fMRI). The pattern of activation in response to white noise presented binaurally was clearly different between participants with and without hyperacusis. For people with no hyperacusis, the activation was mainly in the right superior temporal gyrus, as expected, since the temporal lobes contain the primary auditory cortex. However, for subjects with hyperacusis, activation was also observed in the frontal lobes and occipital lobes, indicating that systems outside the auditory pathway were activated by sound.

From the psychological perspective, the emotional reaction to sound in people with hyperacusis is related to the negative thoughts that are evoked automatically by environmental sounds (Andersson et al, 2005). These automatic thoughts arise with little awareness as to why they arise; they are perceived as strong effects that are not under direct control (Henry and Wilson, 2001). Several authors have acknowledged the role of negative thoughts about environmental sounds in the development of hyperacusis (Khalfa et al, 2002; Nelting et al, 2002). The role of negative thoughts in development of anxiety, depression and social phobia is well established (Kim, 2005; Kercher et al, 2009). As people with hyperacusis often present with symptoms of anxiety, depression and phobia (Goebel and Floetzinger, 2008), the role of negative thoughts in development of the disorder is very likely. The main point here is that the emotional response to sound (getting annoyed, frightened, or irritated) is a result of the content of the automatic thought, not the acoustical characteristics of the sound itself. Also, the sounds that are most effective in evoking emotional responses can vary markedly across individuals. Two people with hyperacusis may present completely opposite reactions in response to a given sound (e.g. loud music). One may experience irritation and discomfort while the other experiences a more neutral or even positive mood state.

Audiological assessment and management of hyperacusis

Many clinicians have used lowered uncomfortable loudness levels (ULLs) as a diagnostic criterion for hyperacusis. In order to measure the ULLs, the patient wears headphones and the tester presents sounds (usually sinusoidal tones) to one ear and gradually makes them more intense. The patient is asked to press a button when the sound becomes uncomfortably loud. The level at which the sound is first reported as being uncomfortably loud is taken as the ULL. When the average ULL at 0.5, 1, 2, and 4 kHz is less than 80 dB hearing level (HL), this is taken as indicating abnormally decreased tolerance to sound (Sherlock and Formby, 2005). Despite the relatively good test-retest repeatability of ULL measurements, ULLs can vary markedly across individuals without hyperacusis, and the results can be influenced by the exact instructions given. Thus, it remains unclear whether ULLs provide a valid or sufficient method for diagnosing hyperacusis.

It is generally believed that the proper management of hyperacusis requires information regarding the psychosocial aspects of sound intolerance (Aazh, 2009). According to the biopsychosocial model of health-care assessment (Havelka et al, 2009), clinicians should take into consideration the biological, psychological and social aspects of a health condition. If the wider psychosocial and social aspects of hyperacusis were ignored, and the clinician relied solely on estimates of ULLs, the clinician would have little to offer in guiding the rehabilitation process. More comprehensive assessment of hyperacusis is possible through self-report validated questionnaires (e.g. Hyperacusis Questionnaire (Khalfa et al, 2002), Geräuschüberempfindlichkeit (Nelting et al, 2002)).

Clinical management of hyperacusis was uncommon until the end of the nineties, when Jastreboff and Hazell developed tinnitus and hyperacusis retraining therapy (TRT: Jastreboff and Hazell, 1993). TRT is based on a neurophysiological model proposed by Jastreboff (1990), and is aimed at removing negative emotional associations with sound. The goal is to achieve this through retraining counselling and sound therapy.

Retraining counselling is a crucial part of TRT; it teaches patients the neurophysiology of the auditory system, components of the Jastreboff neurophysiological model, the interactions between the limbic system, the autonomic nervous system and the auditory pathway, as well as the basis of classical conditioning. This helps the individual to understand how one's emotional reaction to sound affects the perception of sound. Sound therapy is assumed to facilitate the therapy by desensitising the auditory system through the use of wearable wideband noise generators. There are several research studies in the literature supporting the effectiveness of TRT in the management of hyperacusis (Jastreboff and Jastreboff, 2000; Jastreboff and Jastreboff, 2003).

Given the 60% comorbidity of chronic hyperacusis with psychiatric symptoms (Goebel and Floetzinger, 2008), it has been suggested that it may be beneficial to combine TRT with cognitive behavioural therapy (CBT) (Martinez-Devesa et al, 2010), which is a psychological intervention (Andersson et al, 2005; Goebel and Floetzinger, 2008). CBT is effective in reducing anxiety symptoms through helping the individual to identify, challenge and modify their negative automatic thoughts (Hunot et al, 2007). By modifying the thoughts, the emotional reaction to sound is reduced, which promotes recovery from hyperacusis. The main CBT interventions used for management of hyperacusis include cognitive restructuring, attention control, imagery training, relaxation techniques, and graded exposure.

Do earplugs help in the management of hyperacusis?

It has been reported that some people experiencing hyperacusis use earplugs to reduce exposure to day-to-day social noises (Khalfa et al, 2002; Nelting et al, 2002). Use of earplugs has also been recommended for children with hyperacusis when exposed to noisy environments (O'Reilly et al, 2000; Zarchi et al, 2010). However, this may not be a sensible procedure, as the use of earplugs increases the sensitivity of the ear (Formby et al, 2003; Munro and Blount, 2009), and hence could potentially worsen hyperacusis when the earplugs are removed. In addition, it is possible that the reduced loudness of environmental noises offered by the use of earplugs reinforces the association between the avoidance of sound and lack of distress and hence maintains the underlying fears and concerns. An alternative almost opposite approach is a gradual exposure to feared situations (environmental sounds) in a controlled and safe manner, sometimes referred to as desensitization or graded exposure. This can have a therapeutic effect for patients with hyperacusis (Andersson, 2002). Hearing protection devices should only be used where recommended by the health and safety guidelines.

Conclusions

Hyperacusis is a common condition in children, especially children with Williams Syndrome, ASD, and tinnitus. It is, therefore, important that school nurses are aware of the assessment and management of the condition so that they can provide adequate support and information to the children, their families and relevant school employees. Audiological management of hyperacusis involves educational counselling, sound therapy and cognitive behavioural therapy. Use of earplugs is not recommended, as it can increase the sensitivity of the ear when the earplugs are removed and it reinforces safety-seeking behaviour, which could maintain the condition. **BJSN**

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Key Points

- Children with hyperacusis may have severe emotional and behavioural reactions in noisy environments.
- The use of earplugs is not recommended as it may reinforce maladaptive avoidance of sounds and may increases sensitivity to sounds when the earplugs are removed. Hearing protection devices should only be used when recommended by the health and safety guidelines.
- School nurses have an important role to play in identifying the problem and providing adequate support and information during the management process.

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Further information

Hyperacusis factsheet

Deafness Research UK (2006) Hyperacusis. www.deafnessresearch.org.uk/factsheets/hyperacusis.pdf (accessed 3 May 2011)

Deafness Research UK

Almost 9 million people in the UK, one in 7 of the population, experience deafness or have a significant hearing difficulty.

Deafness Research UK is a charity dedicated to funding medical research and provides support and information on hearing loss and deafness.

The website provides a wealth of information for professionals and patients on hearing loss, deafness and related conditions, such as tinnitus.

The publication section provides comprehensive factsheets on glue ear including general information, a guide for teachers and communication tips for helping a child.

The freephone helpline is backed up by a team of medical and scientific experts and is available to answer any questions on hearing loss.

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