Hyperacusis: major research questions

Background

The term “hyperacusis” is used to describe the experience of everyday sounds being perceived as intense and overwhelming. Other terminology that is used in this regard includes “decreased” or “reduced sound tolerance”: An Internet patient forum (www.hyperacusis.net [23]) uses the variant “collapsed sound tolerance.” While there is undoubtedly an emotional and psychological component to hyperacusis [25] (not least since becoming apprehensive about sound exposure is an obvious corollary to perceiving that sound as intense), hyperacusis is a subjective self-reported symptom of some physiological change in the central auditory system such as increased gain [3], such that even when sound is of a moderate intensity it is perceived as loud and intrusive. Hyperacusis is almost exclusively bilateral, and the presentation of unilateral hyperacusis is confined to unilateral triggers such as an acoustic shock [30] or a specific unilateral neural lesion [7].

Epidemiology and natural history

Interest in hyperacusis from both clinicians and researchers is gathering pace, and the numbers of peer-reviewed scientific papers published on the topic of hyperacusis in the past four decades has increased on an annual basis (Fig. 1). Despite this burgeoning attention to symptoms of decreased sound tolerance, fundamental questions remain. The purpose of the present article is to describe and delineate several of these questions, with the aim of supporting research efforts to gather evidence on hyperacusis.

Fig. 1  Papers with hyperacusis as a major topic by year (adapted from [5])
whether they, “ever experience over-sensitivity or distress to particular sounds?” This equates to one child in every typical UK classroom (about 30 children). Risk factors included male gender, higher maternal education level, and readmission to hospital in the first 4 weeks of life.

The situation is much the same regarding the prevalence of hyperacusis in adults, and some basic information about the epidemiology of hyperacusis in adults is not yet available. Paulin and colleagues [34] investigated hyperacusis in a sub-study of the Västerbotten Environmental Health Study in Sweden. Of 8520 adults contacted from the general population, 3406 (40.6%) consented to participation in the study, and it is possible that hyperacusis is over-represented as a result of the low response rate. Of the responders, 9.2% self-identified as having hyperacusis, saying “yes” to: “Do you have a hard time tolerating everyday sounds that you believe most other people can tolerate?”. 1.9% had been diagnosed with sound intolerance by a physician (there was unspecified overlap between the groups). The length of history was not reported.

For some people, hyperacusis is a long-term condition

A question that often arises when counseling a patient with hyperacusis is that of the natural history of the condition. As with epidemiology, basic information is not yet available in this regard, and presently it is not possible to be certain about the future trajectory of a person with hyperacusis. It is evident from patient forums that for some people hyperacusis is a long-term condition, and that for some it is marked by exacerbation and pain as the important characteristics of their experience, suggesting loudness, annoyance, fear, and pain as the important characteristics. While in clinical practice it may not be easy to disambiguate these categories, drawing attention to the experience of sound-evoked pain is of interest. Recent physiology research [15] has identified a population of fibers in the cochlear nerve that appear to be involved in pain perception, perhaps as a warning of cochlear injury. The possibility that these type II unmyelinated fibers are involved in hyperacusis is a potentially important topic for research.

Mechanisms

Although there is a consensus building that hyperacusis is underpinned by an aberrant increase in central auditory gain [4, 29, 44] (whereby “neural activity from more central auditory structures is paradoxically increased at suprathreshold intensities” —4, p1), further and more detailed information is not yet available. In part this is due to the lack of a satisfactory animal model of hyperacusis [12], but it is also the case that several aspects of mechanisms of loudness perception remain obscure [14]. Moreover, the terminology used by the auditory neuroscience community regarding decreased sound tolerance is variable and nonspecific (Table 1).

One potential way forward would be for the auditory neuroscience community to reach a consensus on the terminology and definitions regarding hyperacusis, and then to undertake specific projects detailing how the increased central auditory gain originates, and then persists.
Association with tinnitus

Common mechanisms of hyperacusis and tinnitus have been proposed [23] because they commonly occur together (Table 2).

While there are several studies detailing hyperacusis in persons with a primary complaint of tinnitus, there is less information about tinnitus in persons with a primary complaint of hyperacusis. Anari and colleagues [3] studied 100 adult patients with a primary complaint of hyperacusis, finding that 86% experienced tinnitus, although the severity and impact of tinnitus were not reported.

What is also missing from the literature is information regarding the severity of hyperacusis in a person with a primary complaint of tinnitus, and vice versa. This would be useful when designing interventions that either have to address both symptoms if severe, or focusing on one or other, with a secondary and less severe symptom not requiring direct intervention.

Tinnitus and hyperacusis can be exacerbated by anxiety and stress

Some aspects of the experiences of people with tinnitus, hyperacusis, or both, are convergent. Both tinnitus and hyperacusis can be exacerbated by anxiety and stress, and in each there is an increased incidence of depression. Treatments for each symptom are emerging that utilize elements of cognitive behavioral therapy (CBT) [8, 26], and these can be combined with sound-based therapy.

There are also several aspects of tinnitus and hyperacusis that are markedly divergent, however. Some of these are illustrated in Table 3. This provides further opportunities for clinical research. The areas of divergence are sufficient for one to consider that hyperacusis and tinnitus are quite distinct phenomena, and while both may involve maladaptive change in the central auditory system, the specific mechanisms and manifestations of these changes may be separate, although they may occur together.

How to measure hyperacusis

Several methods exist that attempt to measure hyperacusis. There are techniques for the determination of the loudest sound an individual can tolerate, or is comfortable with, and these include loudness discomfort levels and loudness scaling techniques [1, 31]. The limitations of such procedures are substantial, however, with marked interobserver and test–retest variability [39]. The use of pure-tone stimuli rather than the environmental sounds involved in the lived experience of a person with hyperacusis also limits how generalizable the measure is to real-world difficulties. Unless performed with great care, exposing an individual to sounds at or close to an intensity that evokes discomfort and pain
Hyperacusis

Somatic modulation is rare. Validation of self-help unknown, maybe very limited. Almost exclusively bilateral.

Format

Percept is vivid and salient.

Table 3

| Tinnitus | Hyperacusis |
|----------|-------------|
| Often unilateral, or highly lateralized | Almost exclusively bilateral |
| Somatic modulation is common | Somatic modulation is rare |
| Often intermittent | Rarely intermittent |
| Percept can be formless or primitive | Percept is vivid and salient |
| Self-help can be very effective | Impact of self-help unknown, may be very limited |

Table 4

| Name | Authors (date) | Format | Validation population | Languages available |
|------|----------------|--------|-----------------------|---------------------|
| Geräuschüberempfindlichkeit (GÜF) | Nelting et al. (2002) [32] | 27-item self-report | N = 226 with hyperacusis | German, English (Blasing et al., 2010) [6] |
| Hyperacusis Questionnaire (HQ) | Khalifa et al. (2002) [28] | 12-item self-report | N = 201 general adult population | French, English |
| Multiple Activity Scale for Hyperacusis (MASH) | Dauman and Bous- cau-Faure (2005) [10] | 15-item clinician-led questionnaire | N = 249 adults with tinnitus (79% also had hyperacusis) | English |

The Multiple-Activity Scale for Hyperacusis (MASH; [10]) was developed to assess in which life situations a person is limited by hyperacusis, how annoyed they are by it, how much speech understanding is affected, and how severe it is at different times. It was validated in a tinnitus population. This modified version is yet to be validated in a new tinnitus participant cohort. The use of sound therapy is widespread, and there are two general approaches, both utilizing wide-band noise. The first is to introduce the sound at a quiet and unchallenging level, and then to gradually increase the intensity over a matter of weeks, with the suggestion that this is similar to a graduated exposure program that might be used for desensitization [25]. Alternatively, one might introduce the sound at a quiet and comfortable level and maintain that intensity, the proposal being that the gain of the auditory system is somehow “recalibrated” by that signal. While there are patient self-help reports indicating that pink noise, for example, may be more beneficial than white noise [24], randomized controlled trials (RCT) of these and other sound-based approaches are not yet available.

Another approach used for hyperacusis treatment is CBT. An RCT for CBT in hyperacusis indicated benefit and improvement in measures of sound tolerance [27]. In the case of tinnitus, combining sound-based therapy with elements of CBT has been demonstrated to be beneficial [8], and on the face of it, such combination therapy might also be effective for hyperacusis.

In the case of sound-evoked otalgia, in which pain-sensitive pathways in the cochlear nerve have been implicated, some form of analgesia might be effective. Intratympanic lidocaine has been trialed for tinnitus [9], but the benefits were minimal and the acute side effect of violent vertigo was said to be debilitating. Any effect on hyperacusis, or sound-evoked otalgia, has not been reported.

Outlook

In this paper we have described several areas where important information is lacking regarding hyperacusis (summary in Table 5). Clinicians and researchers are encouraged to collaborate and undertake work in this area, with the aim of increasing knowledge and ultimately improving the care of patients who experience hyperacusis. Such collaborative and sustained effort is proving of benefit in the adjacent field of tinnitus [16, 18–20]. In the case of tinnitus, and more recently mild-to-moderate hearing loss, lis-
The field of hyperacusis is young and evolving. There are still several areas where hyperacusis can have a marked negative impact on quality of life. There are still several areas where important information is lacking regarding hyperacusis. Clinicians and researchers are encouraged to collaborate so as to increase knowledge and ultimately improve the care of patients with hyperacusis. The field of hyperacusis is young and there is a need for capacity building in this challenging yet fascinating area.

**Compliance with ethical guidelines**

**Conflict of interest.** D. M. Baguley and D. J. Hoare declare that they have no competing interests.

This article does not contain any studies with human participants or animals performed by any of the authors.

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**Practical conclusion**

- Hyperacusis can have a marked negative impact on quality of life.
- There are still several areas where important information is lacking regarding hyperacusis.
- Clinicians and researchers are encouraged to collaborate so as to increase knowledge and ultimately improve the care of patients with hyperacusis.
- The field of hyperacusis is young and there is a need for capacity building in this challenging yet fascinating area.

**Table 5** Major research questions in hyperacusis

| Question                                                                 |
|--------------------------------------------------------------------------|
| What is the prevalence of hyperacusis in adults and children?            |
| What are the risk factors associated with hyperacusis?                   |
| What is the natural history of hyperacusis?                              |
| How is “pain hyperacusis” perceived?                                     |
| What mechanisms are involved in hyperacusis?                            |
| What is the relationship between hyperacusis and tinnitus?              |
| Can a questionnaire be developed that accurately measures the impact of hyperacusis and can be used as a treatment outcome measure? |
| What treatments, alone or in combination, are effective for hyperacusis? |

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