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# Hyperacusis in children: A clinical profile

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ABSTRACT

*Objective:* Hyperacusis is commonly seen in clinical paediatric practice and can be distressing for the children and their families. This paper looks at the clinical profile of children seen for hyperacusis in a paediatric audiology service and reviews the possible underlying mechanisms.

*Methods:* Retrospective study of case notes of 61 children with troublesome hyperacusis seen in the paediatric audiology service, looking at their clinical presentation and presence of other medical conditions.

*Results:* Hyperacusis was the main presenting complaint in more than half of the cases (n = 31, 51%). The commonest age at presentation with this problem was 3–4 years (n = 33, 54%).

Hearing was normal in the majority of these children (n = 41, 67%). An active middle ear problem was observed in nearly half (n = 29, 48%) of all the children, of which glue ear (otitis media with effusion, OME) was the commonest. Presence of a neurodevelopmental condition was found in almost half (n = 28, 46%) of these patients of which autistic spectrum disorder was the commonest (8/61, 13%). In nearly one-fourth of the children (23%), presence of both middle ear problems and neurodevelopmental was noted. Tinnitus was an accompanying symptom reported in 11% of all the patients.

*Conclusion:* Hyperacusis may commonly present at a very young age. Awareness of different clinical presentations, presence of other medical conditions and possible underlying pathomechanisms in children with hyperacusis can be helpful for clinicians in informing prognosis, counselling and in individualising management plan.

## 1. Introduction

Sensitivity to sounds, or hyperacusis, is common in children [1–5]. While for most children, this is transient and intervention is not sought, for some children, it can be a significant problem with a marked impact on the activities of the child and the family as a whole. Various terms have been used to describe the different ways in which sensitivity to sounds may present e.g. hyperacusis, phonophobia and misophonia. There is currently no universally agreed definition of hyperacusis. Phillips & Carr [6] defined hyperacusis as a disturbed loudness function remarking that the range of different reported definitions or descriptions used may describe different sensations, likely with different underlying mechanisms and aetiologies. Jastreboff & Jastreboff [7] defined hyperacusis as a decreased sound tolerance where there is a negative reaction to the physical characteristics of a sound. Katzenell & Segal [8] defined hyperacusis as an 'increased sensitivity to sound in levels that would not trouble a normal individual'. Phonophobia and misophonia are specific reactions to sounds. Phonophobia is a fear of sounds, often with anticipatory anxiety at the thought or sight of the object which generates the sound. The term misophonia [7] describes the dislike or, in some cases, distress caused by specific patterns of sound eg. chewing, pen clicking, rather than intolerance to the loudness of the sound.

It is difficult to determine the prevalence of hyperacusis in children because studies have used different criteria to determine and quantify hyperacusis in children e.g. patient/parent interviews, questionnaires and loudness discomfort levels. Overall the reported prevalence of hyperacusis in the general paediatric population varies from 3.2% to 17.1% [1]. Coelho et al., in a study on normally developing children, reported that, while 42% were annoyed or bothered by sounds, only 3.2% had troublesome hyperacusis of which 9% also had phonophobia [4]. Studies involving children with autistic spectrum disorder report a prevalence of hyperacusis between 18 and 63% [9,10]. Higher prevalence rates of up to 95% have been found in children with Williams' syndrome [11].

Hyperacusis can have a significant impact on the activities of both the child and family [12]. Activities may be limited or avoided e.g. the family may only go to certain places with the child or go at certain times when it is likely to be quiet. At home, family activities may be rearranged e.g. vacuuming when the child is away. The avoidance behaviour may include places and situations where troublesome sounds might be present e.g. supermarkets, public toilets with hand dryers.

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## 2. Aim

This paper looks at the clinical profile of children with troublesome hyperacusis seen in our paediatric audiology service and explores possible mechanisms underlying the hyperacusis.

## 3. Methods

A retrospective case review was conducted of 61 patients presenting with hyperacusis in the Paediatric Audiology department at Halliwell Health and Children's centre between Nov 2008–Dec 2013. All children seen for hyperacusis in our service are routinely asked by clinicians if they were troubled/bothered by these symptoms, which noises were particularly troublesome and how they react to them.

In this study we defined hyperacusis as an increased sound sensitivity and troublesome hyperacusis where the reaction to the troublesome sound had a negative impact on the child's behaviour or daily living.

### 3.1. Ethics approval

As the study was a retrospective case review with no identifiable patient information being used, we were advised by the local research authority that ethical approval was not needed.

### 3.2. Identification of participants

Patients were identified through clinic letters in the local electronic data base using the search terms 'hypersensitivity to sounds', 'sensitivity to sounds' and 'hyperacusis' (the usual terms used by clinicians in our service for this complaint). We then looked at case notes of all the identified patients who had presented with these symptoms for further details.

### 3.3. Inclusion criteria

Children, or parents of children, who reported hyperacusis as troublesome/bothersome and affecting the child's behaviour or activities were included in the study. All included children completed an age/ability appropriate hearing assessment.

### 3.4. Data collection

Information collected from case notes of each patient was recorded on a Microsoft excel spreadsheet based on the following criteria:

- age of the child at presentation
- gender
- new or follow-up patient
- reason for referral
- presenting symptoms
- which sounds were particularly troublesome
- reported reaction to troublesome sounds
- presence of additional medical conditions
- presence/absence of hearing loss on audiological assessment which included age/ability appropriate audiometry and tympanometry

The type of hearing assessment used depended on the age and developmental level of the child (visual reinforcement audiometry, performance test, play audiometry, pure tone audiometry) for four test frequencies- 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz using hearing loss descriptor in accordance with recommendations of British Society of Audiology [13].

- Normal hearing ≤20 dB HL
- Mild hearing loss 21–40 dB HL
- Moderate hearing loss 41–70 dB HL
- Severe hearing loss 71–95 dB HL
- Profound hearing loss > 95 dB HL

Where indicated, and possible, bone conduction thresholds were obtained to differentiate sensorineural and conductive hearing loss. In those children where tests were done in the free field, the average of the result of four frequencies in the free field was recorded when using hearing level descriptor. As some children, particularly those with additional needs, required several visits to complete the hearing assessment, test results that were complete from either the first and/or second consultation were taken and the average of the four frequencies in each ear was recorded. Otoscopy and tympanometry findings were recorded for all the ears. If the tympanogram trace was flat bilaterally with normal ear canal volume and raised hearing thresholds recorded in the free field on more than two consecutive tests done in an interval of three months, the outcome was recorded as positive for bilateral otitis media with effusion (OME).

## 4. Results

More than half of the 61 children (n = 31, 51%) were primarily referred for symptoms of hyperacusis. For the others, hyperacusis was identified as part of the audiology history in 25 (41%) patients who were referred for other problems and reported as a new concern in 5(8%) children who were already under our care.

Commonest age at presentation was 3–4 years (n = 33, 54%), age range being 1–15 years (Fig. 1). The mean age at presentations was 4.93 years (SD 2.57). The male (n = 38) to female (n = 23) ratio was 1.65 to 1 (p = 0.07).

In our cohort of children, the commonest troublesome sounds included vacuum cleaners, hair/hand dryers and sirens. The range and frequency of troublesome sounds are reported in Table 1. Some children reported being troubled by more than one sound (Fig. 2). The responses to the troublesome sounds were variable. The commonest response was to cover the ears (n = 43, 70%). Twenty-seven children (44%) became very distressed, reported crying or screaming in response to the troublesome sound. Pain in the ears or head was reported by 5(8%) of children. Three children (5%) reportedly ran away or hid in response. Two children became aggressive, for example, one boy would hit his sister when bothered by noises. One child had urinary incontinence in response and one child reportedly reacted by grinding his teeth.

The impact on the child's home, school and social activities was significant for example, some children reported difficulties at school e.g. getting upset by school bell, avoiding lunch to escape the noisy dinner halls. Some families had to avoid parties because of the noisy crowd, for one child the family had to whisper 'happy birthday' song to him on his own birthday. For one child, avoidance of public bathrooms in case someone switched on the hand dryer led to toileting 'accidents'.

One family reported having to keep stopping on the road every time

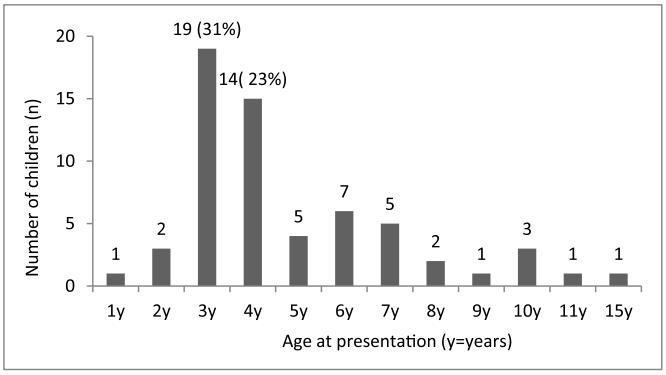


**Fig. 1.** Age at presentation (n = 61).

**Table 1**
Sounds reported as troublesome.

| Sounds (in order of frequency) | Number of children (%) |
|---|---|
| Vacuum cleaner | 28(45%) |
| Noisy crowd/children shouting | 22(36%) |
| Music related sounds | 17(28%) |
| Hair/hand dryer | 13(21%) |
| Siren/alarms | 9(15%) |
| Traffic/vehicle noise | 7 |
| Ringing noise/door bells | 5 |
| Washing machine | 5 |
| Clapping | 4 |
| Drilling noise | 4 |
| Dog barking | 3 |

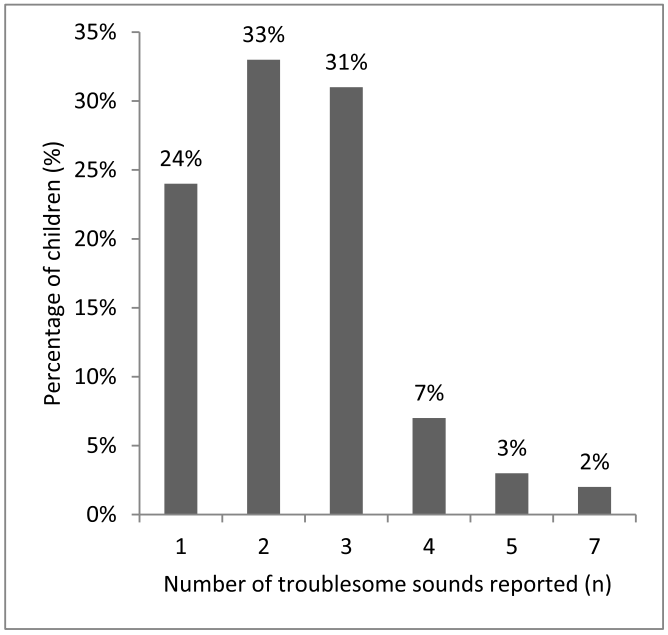


**Fig. 2.** Frequency of troublesome sounds.

a vehicle passed. The youngest child in the cohort was one year old and he would just freeze in response to loud noises.

The range of tests used is shown in Fig. 3. Hearing was normal in the majority of these children (n = 41, 67%). A mild to moderate

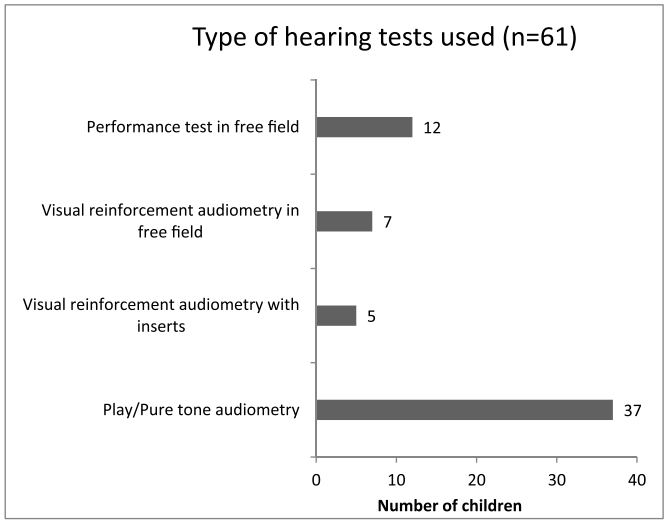


**Fig. 3.** Types of hearing tests used.

conductive hearing loss was noted in 20 (33%) children, mostly secondary to OME (otitis media with effusion) (16/20) (Fig. 4). Only one child had a bilateral mild sensorineural hearing loss. One child reported difficulties with listening despite normal peripheral hearing and was suspected to have auditory processing disorder (APD). While further assessments were planned to investigate APD, the child did not attend subsequent appointments.

An active middle ear problem was noted in 29(48%) of children with the commonest finding being was otitis media with effusion 23(38%). Of the remaining six children, two had a unilateral tympanic membrane perforation post grommet insertion and one had right sided chronic suppurative otitis media with right sided hyperacusis. Five additional children had a past history of otitis media with effusion for which they were previously seen in the department. Tinnitus was an accompanying symptom reported in 11% of the total children.

Neurodevelopmental conditions were observed in nearly half of the total children (n = 28, 46%) (Table 2). Autistic spectrum disorder was the commonest condition (8/61, 13%). Two children had significant sensory processing difficulties. An additional 6% of children had significant behavioural problems for which they were either under the Community Paediatrics Service or the Child and Adolescent Mental Health Service team. In nearly a quarter of the children (n = 14, 23%), middle ear problems and neurodevelopmental conditions were both noted to be present.

When existing medical conditions were explored across the age range, an active middle ear problem was much more common in the preschool age group. Also presence of both an active middle ear problem and a neurodevelopmental condition was much more commonly seen in the younger children. In the older children, presence of a neurodevelopmental condition was more commonly seen. Fig. 5 shows clinical conditions noted in relation to age.

## 5. Discussion

Hyperacusis is commonly seen in clinical practice in children. Children may present with this problem to different services e.g. Paediatrics, Audiology, ENT, Psychiatry and General Practice. Intervention should be considered when the condition impacts on the child's or the family's activities. Awareness of possible processes underlying a child's hyperacusis may help in predicting the likely course of the problem and the choice of management strategies. We looked at various mechanisms that could account for increased sensitivity to sounds in children.

Possible mechanisms for hyperacusis in children include

- Immature but normally developing auditory system
- Temporary auditory deprivation
- Disorder within the auditory system
- Disorder of sensory processing

As most of the children with hyperacusis fall in the pre-school group, it is likely that with maturation of the auditory system, the ability to process sounds improves with time and the sounds then cease to be troublesome. Part of the auditory maturation process is the development of the sensory gating process which inhibits 'distracting', or non-relevant auditory information. An inability to inhibit such information can cause difficulty in appropriately filtering and processing sounds. In studies investigating maturation of sensory processing mechanisms in children with normal development, one study reported that sensory gating matures around eight years of age [14]. Another study found that maturation of the auditory system continued until 13 years of age [15].

One third of the children in our cohort had a hearing loss, mainly conductive. Hyperacusis related to temporary hearing loss, for example due to otitis media with effusion, may involve a number of mechanisms. Sun Wei et al. [16], in a study on rats, observed that conductive hearing

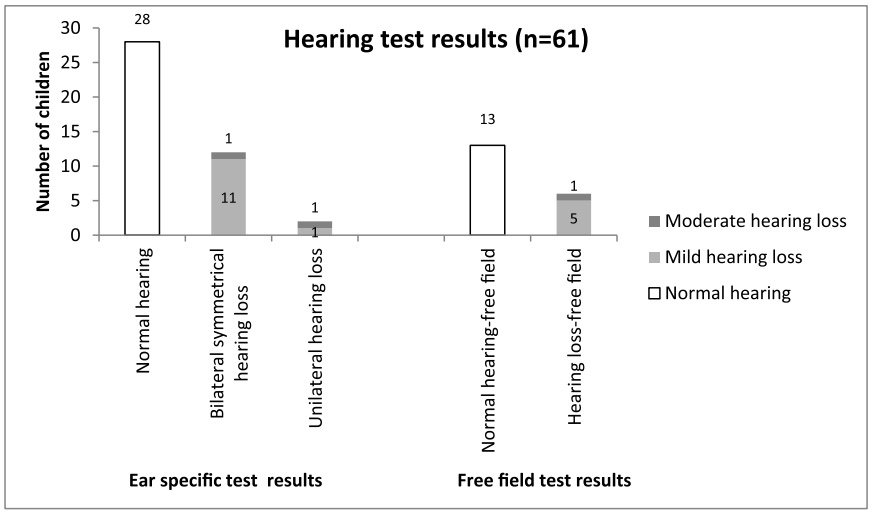


**Fig. 4.** Results of hearing assessments.

**Table 2**
Neurodevelopmental conditions.

| Neurodevelopmental problems noted | Number of children (28/61) 46% |
|---|---|
| Autistic Spectrum Disorder | 8 |
| Cerebral Palsy | 3 |
| Attention Deficit Hyperactivity Disorder | 2 |
| Down's Syndrome | 2 |
| Prematurity and post-haemorrhagic hydrocephalus | 2 |
| Epilepsy | 2 |
| Sensory processing problems | 2 |
| Dyspraxia | 1 |
| Klinefelter's syndrome | 1 |
| Microcephaly, severe learning difficulties | 1 |
| Williams syndrome | 1 |
| Leigh's syndrome | 1 |
| Microdeletion 15q13.3 | 1 |
| Possible Syndrome (facial dysmorphism, developmental delay, cardiac defect) | 1 |

loss at an early age impaired sound tolerance by reducing GABA inhibition in the inferior colliculus. Auditory deprivation can also impact on the processing of sounds in the auditory brainstem. There have been several human studies which show that temporary auditory deprivation may result in lower acoustic reflex thresholds and a temporary increase in the central auditory gain [17–19]. This increase in the central auditory gain has the effect of 'turning up' the subjective loudness of sounds which is particularly apparent when the auditory obstruction has resolved. This effect can be noted in children who have had ventilation tubes inserted for chronic otitis media with effusion. These children often report that sounds are loud post-operatively. One study post insertion of ventilation tubes reported hyperacusis of varying severity in 47% of children lasting between 2 and 40 days [20].

For some children, the disorder in auditory system may be permanent rather than temporary. Autistic spectrum disorder (ASD) was the commonest associated neurodevelopmental condition in our cohort. Children with autistic spectrum disorder are reported to have disordered loudness processing with a restricted dynamic range of perception, an increased subjective perception and reduced tolerance of loudness [9]. This auditory 'hypersensitivity' was noted not only with loud sounds but also with sounds considered to be of moderate sound intensity. For some children, there may be the possibility that a medical condition, eg diabetes mellitus, may have an impact on central auditory mechanisms due to pathological changes in medial olivocochlear myelinated fibres [21].

For other children, the problem may lie with sensory rather than auditory processing. They may have difficulty in regulating or integrating sensory information (visual, touch, sounds, smells, proprioception) which can lead to patterns of hyper-sensitivity to sensory stimuli or a 'sensory overload' effect. This may be due to Sensory Over-Responsivity (SOR), a sensory modulation disorder manifested by behavioural responses that are faster, longer or more intense compared to peers [22]. Normally developing children may display one or two such

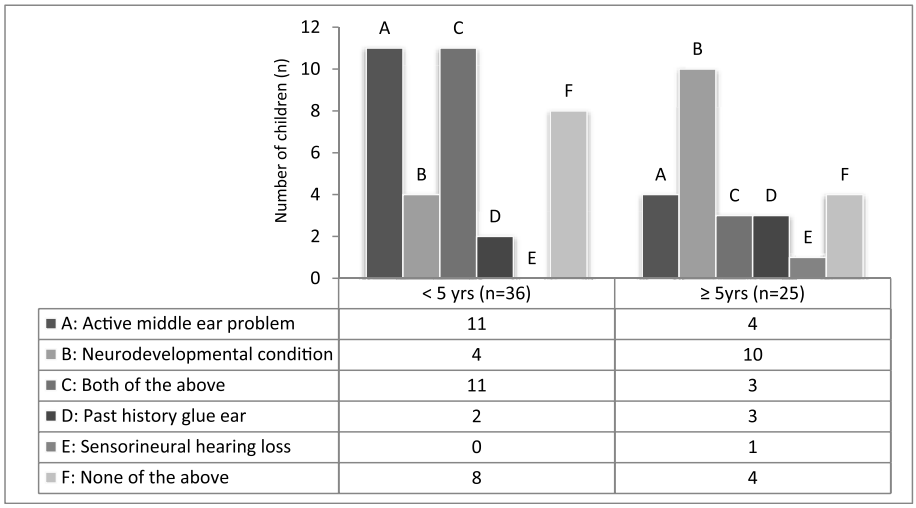


**Fig. 5.** Associated medical conditions in relation to age (n = 61).

symptoms. Ben-Sasson & Carter [23] reported that SOR interfered with routine activities in 5–16% of school age children. SOR has been reported in children with neurodevelopmental disorders e.g. Attention Deficit Hyperactivity Disorder, Fragile X Syndrome and ASD [24–28]. In children with autism, poor inhibitory sensory processing control [29] and difficulties with attention control [30] have been suggested as mechanisms. Adamson et al. [31] reported that autistic children, independent of age, also had abnormal sensory reactivity with greatest difficulty in auditory filtering. Our case review showed sensitivity to sounds in children with neuro-developmental disorders across the age range. Two children in our cohort reported difficulties with multiple sensory stimuli.

We had one child with William's syndrome in this cohort, a condition known to be associated with a high prevalence of hyperacusis [11,32]. Theories proposed for hyperacusis in this population include a dysfunctional auditory efferent system, lack of acoustic reflexes and reported hyperactive central auditory processing [32–34]. There is also a high rate of otitis media with effusion reported in children with William's syndrome (65%) [11].

One of the strengths of this study is that we looked at the clinical profile of children with troublesome hyperacusis across a wide age range including preschool children who have not previously been studied. A limitation of a retrospective approach however is that not all children seen with troublesome hyperacusis may have been captured in this study and symptoms of hyperacusis and/or any associated problems may not have been consistently explored or documented during the clinic consultation.

It is important to take a thorough history when assessing a child with hyperacusis. History should include information about the troublesome sounds and situations, how the child reacts and how this impacts on the child and the family. It can be helpful for the family to keep a ‘sound diary’ to explore this. It is important to look at any safety behaviours used by the child and the family, as these can make it difficult for the child to habituate to the troublesome sounds and/or perpetuate the problem. A common safety behaviour is the brisk removal of the child from the situation once the distress is noted. This denies the child the opportunity to habituate to the sounds or the situation. It is more helpful to prepare the child for the experience, perhaps by gradual exposure or by giving the child cues about the order of events. Another common unhelpful safety behaviour is the use of ear defenders. This can exacerbate hyperacusis as the resulting increased central auditory gain leads to an increased perception of loudness of sound. The use of ear defenders must therefore be discouraged, unless in a very loud environment when everyone should use them.

Information should be obtained about the child's otological, audiological, medical and developmental history. Also it is helpful to explore if there are any other sensory difficulties, as hyperacusis may be just one of several experienced sensory difficulties. Selection of audiological assessments is dependent on the child's age and ability and can include appropriate behavioural audiometry, tympanometry and otoacoustic emissions. The use of stapedial reflexes and loudness discomfort levels is more likely to lead to distress than significant information to influence management. A paediatric assessment may be required if there is possibility of an associated or co-existing neurodevelopmental condition.

It is helpful to involve the child in the management process. The explanation and management strategies should be age and ability dependent. For children with normal auditory development, an explanation to the child and parents/carers, with reassurance that the problem will resolve over time, may be all that is needed. Most children with hyperacusis are comfortable with very loud sounds that they generate but have difficulty with sounds from other sources over which they have no control. Simple intervention strategies can include enabling the child to have some control over the troublesome sound e.g. assisting with the vacuuming. Strategies can also focus on an enjoyable element of the sound e.g. the ice cream flavours rather than the sound of the ice cream van. In situations where there are multiple stimuli e.g. the sights, sounds and smells of a children's party, the child could be encouraged to focus on a specific aspect of the situation, e.g. an individual child or activity, to reduce the overwhelming impact, or ‘sensory overload’, of the experience.

Some children may demonstrate anticipatory anxiety to a sound or a situation, for example becoming distressed at the sight of the vacuum cleaner in anticipation of its sound, or, at the sight of a balloon in case it pops. Here, behavioural desensitisation strategies may be helpful e.g. introducing objects/vacuum cleaner in a positive manner, perhaps through a storyboard, and/or gradually building up a tolerance to its presence and sound. Where the hyperacusis is a problem at school, it can be helpful to involve school staff to ensure that a consistent approach is followed across home and school. Some children may benefit from referral to a paediatric psychology or mental health service for desensitisation strategies, particularly where phobias or panic attacks are part of the response. Others, particularly those with developmental difficulties, may benefit from a combined behavioural and auditory approach. Auditory desensitisation strategies may involve sound therapy [35] or devices e.g. sound generators, to help decrease the impact of the troublesome sounds while the behavioural strategies are implemented.

## 6. Conclusion

Hyperacusis may commonly present at a very young age in children. It can be distressing for some children and their families. Exploration of the nature and impact of hyperacusis and any safety behaviours can help in the development of a management plan to lessen the associated distress and improve the child's quality of life.

Developmental immaturity of sensory processing and correlation with middle ear related problems are possible underlying mechanisms in younger children that resolve as they grow older. In children with neurodevelopmental conditions, the mechanisms may include abnormal sensory processing and may explain longer persistence of the symptoms. These children benefit from a multimodal management approach. Multidisciplinary input from paediatric audiology, paediatrics, psychology services and school may be beneficial in addressing the varied impact of the hyperacusis on some children and in managing associated conditions.

Awareness of the different presentations, age at presentation and clinical profile of children can help clinicians in determining the likely prognosis and individualising the management plan. Further studies are needed in this area to establish more clarity in the mechanisms underlying hyperacusis in children and in the development of management strategies.

## Licence statement



## Funding statement

Funding was not required.

## Contributors

SM-study design, data acquisition and analysis, literature review, revision of the manuscript.

VK- conception, study design, data analysis, important intellectual content, literature review, revision of manuscript.

## Competing interests

None.

## Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.ijporl.2018.01.004.

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