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Chapter 32

Functional auditory disorders

D.M. BAGULEY[1]*, T.E. COPE[2], AND D.J. McFERRAN[3]
[1]*Department of Audiology, Cambridge University Hospitals, Cambridge, UK*
[2]*Department of Neurology, Cambridge University Hospitals, Cambridge, UK*
[3]*Department of Otolaryngology, Colchester Hospital University, Colchester, UK*

Abstract

There are a number of auditory symptom syndromes that can develop without an organic basis. Some of these, such as nonorganic hearing loss, affect populations similar to those presenting with functional somatosensory and motor symptoms, while others, such as musical hallucination, affect populations with a significantly different demographic and require different treatment strategies. Many of these conditions owe their origin to measurably abnormal peripheral sensory pathology or brain network activity, but their pathological impact is often due, at least in part, to overamplification of the salience of these phenomena. For each syndrome, this chapter briefly outlines a definition, demographics, investigations, putative mechanisms, and treatment strategies. Consideration is given to what extent they can be considered to have a functional basis. Treatments are in many cases pragmatic and rudimentary, needing more work to be done in integrating insights from behavioral and cognitive psychology to auditory neuroscience. The audiology literature has historically equated the term functional with malingering, although this perception is, thankfully, slowly changing. These disorders transcend the disciplines of audiology, otorhinolaryngology, neurology and psychiatry, and a multidisciplinary approach is often rewarding.

INTRODUCTION

This chapter outlines the disparate collection of auditory symptoms that can be considered to have a functional basis, at least in some cases. They transcend the disciplines of audiology, otorhinolaryngology, neurology, and psychiatry. Many of these conditions owe their origin to measurably abnormal peripheral sensory pathology or brain network activity, but their pathological impact is often due, at least in part, to overamplification of the salience of these phenomena.

Some of the conditions we describe, such as nonorganic hearing loss (NOHL), appear to affect a similar demographic and are amenable to similar psychological interventions to those functional disorders affecting motor or somatosensory systems that are commonly encountered in neurology clinics. Others, such as musical hallucination (MH), affect strikingly different population groups. As in those functional disorders affecting motor and sensory symptoms, it has only been in relatively recent years that we have come to recognize these conditions as truly disabling.

NONORGANIC HEARING LOSS

Definition

The situation in which patients may behave as if they have a significant hearing loss, both in general communication and on pure-tone audiometry, that is not borne out by specialized or objective testing, has a varied terminology. The descriptors malingering and feigning have been used clinically, possibly deriving from early reports of such behavior in wartime (Peck, 2012), and assume intentionality. The term psychogenic hearing loss makes an assumption that this is an exclusively

*Correspondence to: David M. Baguley, Audiology (94), Cambridge University Hospitals, Hills Road, Cambridge CB2 0QQ, UK. Tel: +44-1223-217594, E-mail: dmb29@cam.ac.uk

psychological disorder. Functional hearing loss and NOHL are less loaded, and thus are preferable.

Demographics

Information regarding the epidemiology of NOHL is extremely sparse. There have been estimates that up to 30% of individuals claiming compensation for noise-induced hearing loss have some nonorganic component to their complaint (Peck, 2012). The presentation is familiar to all audiologists and otolaryngologists, but no systematic study of prevalence and incidence has been undertaken.

The patient population with NOHL is varied, but there are some classic presentations. Teenagers, said to be more often girls than boys (Peck, 2012), may claim poor hearing but demonstrate inconsistent extent of hearing loss. Individuals claiming compensation for industrial or accidentally caused hearing loss may wish the extent of the loss to appear greater than it is. Some adults may receive secondary gain from being thought to have a hearing loss, whether in terms of support from family/friends, from the state, or greater respect and identity; recognition of this gain might not be conscious.

Investigations

Pure-tone audiometry is required in any consideration of hearing loss but it should be remembered that the audiogram is a threshold test of signal detection and does not adequately represent real-world hearing abilities. In NOHL an audiologist can use specific techniques to obtain audiometric thresholds: these may include presenting the stimuli at random, nonpredictable intensities, or only presenting at low intensities.

Specialized behavioral tests exist, the most common example being the Stenger test, which exploits the principle that if stimuli are presented simultaneously to each ear, only the louder will be perceived. A tone is presented at 10 dB above audiometric threshold to one ear, and 20 dB below the admitted threshold in the other, this latter tone then being raised in steps until the patient ceases responding, that point approximating the threshold in that ear.

Objective (physiological) hearing testing, in the form of auditory evoked potentials and otoacoustic emission testing, has largely replaced the behavioral testing approaches. It does require patient compliance for extended test periods, but can glean ear- and frequency-specific physiological thresholds. Cortical auditory evoked potentials require the patient to be alert, and are attenuated with general anesthesia (Simpson et al., 2002). Auditory brainstem responses (ABRs) persist under general anesthesia and natural sleep, and frequency-specific test protocols are now in use, rather than the traditional wide frequency band click stimuli.

Is it functional?

Austen and Lynch (2004) proposed a model that considered the following factors: motivating factors that may be observed, the type of gain (including financial or role), the degree of intention, and the consistency of response to testing.

They hypothesized that three categories of disorder occur: malingering, factitious, and conversion. It was considered that individuals might move between these categories, and that management strategies would be different for each. This model has been influential and widely adopted, despite retaining pejorative vocabulary, but no further systematic research has been undertaken.

It is almost tautologic but nonetheless important to indicate that NOHL is functional in some cases, though in others there may be intentional aspects to the behaviors.

Treatment strategies

There are indications that NOHL may be a marker for psychological problems (Austen and Lynch, 2004), so the approach to treatment has to include vigilance to such issues, but to avoid overreaction. The present consensus is to avoid confrontation, and to create an expectation of recovery. Where there is secondary gain, psychology support to meet those needs in other forms may be of use, and where conversion disorder is evident, psychiatric support should be sought.

DISORDERS OF AUDITORY PROCESSING

Definition

Auditory processing disorder (APD) encompasses a range of developmental and acquired disorders that affect auditory analysis and cannot be directly explained by structural pathology in the brain or cochlea or generalized cognitive deficit. Patients typically have normal auditory threshold sensitivity but have difficulty identifying speech (Keith, 2000) and/or nonspeech sounds (Rosen, 2005; Moore, 2006). The usual presenting complaint is an impaired ability to hear speech in background noise in comparison to their peers. APD may coexist with peripheral hearing loss, complicating diagnosis (Moore et al., 2013). The auditory discrimination difficulties are especially marked in challenging listening environments, when target sounds are brief, masked, or degraded (ASHA, 1996; Jerger and Musiek, 2000). The diagnosis encompasses a number of overlapping clinical syndromes (Jerger and Musiek, 2000; Hind, 2006), and its underlying pathological basis is poorly understood. Of

those children complaining of symptoms consistent with APD, only around 5% have an underlying structural or other obvious neurological cause (Chermak and Musiek, 1997).

Demographics

There remains debate about whether the diagnosis of APD should be confined to that small group of children who have difficulties restricted to the processing of complex sounds, or whether it can be invoked for individuals with multimodal perceptual processing difficulties (Cacace and McFarland, 2005). When defined broadly, APDs are a common group of conditions, affecting up to 7% of children (Bamiou et al., 2001), but only a small proportion of this group will seek a medical opinion.

Investigations

In APD pure-tone audiometry is often, but not always, normal. ABRs can be helpful in disambiguating APD from its key differential diagnosis, auditory neuropathy / dyssynchrony, in which the presenting symptom may also be problems with complex sound processing. In APD one would expect ABRs to mirror pure-tone audiometry and outer hair cell function (i.e., to be normal if peripheral function is normal), while in auditory neuropathy ABRs are significantly disrupted (Starr et al., 1996; Berlin et al., 2003).

Middle latency responses and cortical responses to unexpected stimuli (mismatch negativity: Garrido et al., 2009) are rarely measured in clinical practice (Emanuel et al., 2011). While some authorities have proposed that these responses, which are generated higher in the auditory pathway, might provide objective evidence of a processing abnormality (Sharma et al., 2006), their sensitivity and specificity are not well established.

Auditory psychophysical tests form the cornerstone of assessment for APD, but no gold-standard test battery has been established (Moore et al., 2013). The most commonly applied battery of tests is called SCAN (Bensimon et al., 2009), and versions exist for children under 12 (SCAN-C: Keith, 2000), as well as adolescents and adults (SCAN-A: Keith, 1995). It comprises four subtests assessing the perception of: (1) words presented monaurally in background noise; (2) acoustically degraded single words; (3) dichotic single words; and (4) sentences.

It is heavily linguistically based, and population norms vary significantly between countries and ethnic background (Dawes and Bishop, 2007). Because of this, many centers supplement SCAN with nonlinguistic tests of auditory processing, commonly assessing performance in temporal and pitch processing, as well as sequence analysis and binaural integration. It is also important to assess global cognitive function with a battery of standard neuropsychological tests.

Is it functional?

A particular diagnostic challenge is the disambiguation of APD from more general deficits of attentional processes such as attention deficit hyperactivity disorder and, indeed, these conditions frequently coexist (Riccio et al., 1994). The interplay of attention and auditory processing is complex and variable. Performance on objective tests of auditory processing is strongly affected by inattention, especially in young children (Moore et al., 2008). Some have argued that this process underlies the majority of deficits in APD, while others point to cases where auditory processing is impaired despite intense concentration by the listener. As such, a functional component is present in many cases of APD.

Treatment strategies

Intervention in APD focuses on auditory training in combination with compensatory environmental and behavioral modification. There is no universally agreed strategy for auditory training, and both computer-based and face-to-face strategies are employed, but it is generally more effective if it involves audiovisual integration of meaningful sound stimuli (Loo et al., 2010). There is preliminary evidence that these methods modestly improve both behavioral performance and neural encoding of auditory information (Russo et al., 2005), especially if the deficit affects only one ear (Moncrieff and Wertz, 2008). For school- and university-age patients, educational support, sound reinforcement, and personal FM systems (frequency-modulated radio devices that allow teachers to communicate directly with pupils) can be of benefit. In general, APD improves with age, but it can persist into adulthood or develop *de novo* in elderly individuals (Cooper and Gates, 1991).

TINNITUS

Definition

Tinnitus is a common symptom that is surprisingly difficult to define unambiguously. One regularly used definition is that tinnitus is the conscious perception of an auditory sensation in the absence of a corresponding external stimulus. This definition could include the auditory hallucinations of psychotic illness but in practice these are excluded. Other symptoms that comply with this definition and are sometimes seen as subtypes of tinnitus include musical hallucination (see below) and pulsatile tinnitus. The latter is a rhythmic percept and, if synchronous with the heart beat, a vascular origin is likely; if asynchronous, myoclonus of the middle-ear

muscles or palatal muscles is likely. The sound of pulsatile tinnitus can occasionally be perceived by external measuring equipment, in which case it is designated as objective. Most tinnitus, however, can only be heard by the sufferer and is classified as subjective. Commonly perceived sounds include buzzing, ringing, whistling or humming, and the sensation may be localized in one or both ears, or inside the head. A small number of people perceive tinnitus as an external sound.

Demographics

Epidemiological studies have estimated the prevalence of tinnitus in Europe, Asia, Africa, and North America, mostly producing rates between 10 and 15% of the adult population (Baguley et al., 2013). The largest study so far was undertaken in the UK as part of the National Study of Hearing in England (Davis and El Rafaie, 2000). With a study population of 48 313, this gave a tinnitus prevalence of 10.1%, with 2.8% describing the tinnitus as at least moderately annoying and 0.5% reporting that it had a severe effect on their ability to lead a normal life.

Prevalence in men and women is broadly similar. Tinnitus prevalence increases with age up to the seventh decade of life, with some studies showing the prevalence continuing to rise beyond that point, whereas others demonstrate a plateau or even a decline (see Baguley et al., 2013 for review). It is unusual for children to spontaneously complain about tinnitus but, when appropriate questions are asked, the prevalence of tinnitus experience in childhood seems similar to that in adulthood. Tinnitus is more common in people who have had significant noise exposure, and more common in those with hearing loss. However, the relationship between hearing loss and tinnitus is controversial: it is possible to have tinnitus with a normal audiogram and the degree of any hearing loss correlates poorly with tinnitus impact. There are few longitudinal incidence studies and it is difficult to draw conclusions, though one study demonstrated that tinnitus severity generally lessens rather than worsens with time (Nondahl et al., 2010). Tinnitus is seen as part of several otological diseases, including otosclerosis, Ménière's disease, and tumors of the cerebellopontine angle. Tinnitus has several comorbidities: the most common are disorders of loudness perception (discussed below) and other forms of impaired sound tolerance; anxiety, depression, and temporomandibular joint dysfunction are also seen in association.

Investigations

Audiometry is the main (and often the only) investigation required for tinnitus patients. A pure-tone audiogram aids otological diagnosis, determines those who need further investigations, and helps to direct subsequent management strategies. Measuring middle-ear function using tympanometry is often useful, as many patients with tinnitus complain of a feeling of aural fullness or blockage; normal tympanometry results exclude a diagnosis of eustachian tube dysfunction at the time of testing. Audiometric equipment can be used to try and match the pitch and loudness of tinnitus (Cope et al., 2011), but this is time consuming and does not usually help with treatment. Patients with unilateral or asymmetric tinnitus, an asymmetric audiogram, or associated unexplained neurological symptoms should undergo magnetic resonance imaging (MRI).

The most commonly found pathology in association with tinnitus is a vestibular schwannoma, which can be missed by computed tomography. For those with variants of tinnitus, particularly pulsatile tinnitus, more complex investigative algorithms are required. Tinnitus-specific questionnaires can help to assess the impact of tinnitus, the present instrument of choice being the Tinnitus Functional Index (Meikle et al., 2012).

There is an association between tinnitus and anxiety and depression, and in assessing a patient with tinnitus, some awareness of these symptoms and assessment of severity is indicated.

Is it functional?

There are subtypes of tinnitus, particularly pulsatile tinnitus, in which the symptom is associated with real noise of vascular or muscular origin. Such cases have hitherto generally not been regarded as functional. Recent observational studies, however, have suggested that at least some cases of both middle-ear myoclonus and palatal myoclonus may have a functional basis and hence any associated tinnitus could be regarded as functional (Stamelou et al., 2012; Ellenstein et al., 2013). For the majority of cases of tinnitus the percept is nonpulsatile and not associated with any internal sound source. Such tinnitus has paradoxes: tinnitus is possible with normal audiometry; most people with hearing loss do not have tinnitus; the extent of any hearing loss does not correlate well with tinnitus severity; tinnitus can occur even after the auditory nerve has been severed. But can tinnitus occur with a completely normal peripheral auditory system? Even those patients who have a normal pure-tone audiogram may have subtle cochlear deficits, and using tools such as extended-range audiometry, threshold-equalizing noise testing, or distortion product otoacoustic emission testing supports the view that the patients with tinnitus are more likely to have defects within the cochlea than audiometrically matched non-tinnitus controls (Weisz et al., 2006; Fabijańska et al., 2012).

There are, however, some tinnitus patients who appear to have tinnitus in the presence of normal cochlear function: a study that used extended-range audiometry to

investigate 18 people who had tinnitus with a normal conventional audiogram found that 6 of the 18 subjects had high-frequency hearing as good as or better than the control group (Shim et al., 2009). Certainly, aural pathology on its own cannot fully explain symptom variability, giving rise to a theory that the pathophysiology of tinnitus is a two-stage process with an ignition occurring anywhere in the auditory system and then a process of promotion within the central auditory system (Baguley, 2006). Suggestions regarding the central mechanisms include the sequelae of deafferentation, namely increased spontaneous neural firing, cortical map reorganization, or increased neural synchrony. Overall, the evidence suggests an experience-dependent abnormal central analysis of peripheral information, remarkably concordant with evolving views of functional motor and somatosensory phenomena (Edwards et al., 2012).

Treatment strategies

In a small proportion of cases a specific treatment may be possible: examples include stapedectomy for people with tinnitus in association with otosclerosis, or embolization of an arteriovenous fistula that has caused pulsatile tinnitus. For the vast majority of cases of tinnitus there is no specific curative treatment, and management is largely supportive and empirical. Explanation, reassurance, and education may be all that is required. Correcting any associated hearing loss with hearing aids is anecdotally helpful, even if the hearing loss is mild and not causing significant communication problems (Sereda et al., 2015). Sound therapy is often utilized – either adding low-level sound to the patient's environment or giving the patient a wide-band, ear-level sound generator (masker) to wear. There are also combination devices that marry a sound generator to a hearing aid. Despite being widely used there is little scientific evidence to support sound therapy in tinnitus management (Hobson et al., 2012). Relaxation training may be offered, particularly to those who report that their tinnitus increases when stressed.

Psychological treatments have the best evidence base for effective tinnitus management, particularly cognitive-behavioral therapy (CBT) (Martinez-Devesa et al., 2010; Hesser et al., 2011), though recently mindfulness meditation (Philippot et al., 2012) and acceptance and commitment therapy (Westin et al., 2011) have also been positively assessed. Several protocols have been devised that use a range of the above modalities in combination in a structured framework. These include tinnitus retraining therapy (Jastreboff and Hazell, 1993), progressive audiologic tinnitus management (Myers et al., 2014), and tinnitus activities treatment (Tyler et al., 2007): there is limited evidence that this approach is helpful. Many drugs have been investigated, but none is currently recommended for the treatment of tinnitus. Similarly, although several other treatment modalities such as repetitive transcranial magnetic stimulation, transcranial direct current brain stimulation, and low-level laser therapy have been studied experimentally, none is in widespread clinical usage.

MUSICAL HALLUCINATION (MH)

Definition

Hallucination is the experience of a percept without a causal external stimulus. MH is therefore more than simply having a tune "stuck in your head" (an earworm), as it must have a compelling sense of reality. Indeed, patients commonly present to our services having first erroneously complained to police or local council services about their neighbors' antisocial music playing, and some still believe the source to be external when assessed in clinic. MH is typically experienced as short fragments of simple melodies – often from music heard regularly and familiar from youth, and especially from hymns and carols (Griffiths, 2000; Warner and Aziz, 2005). Lyrics may or may not be heard, but it is phenomenologically and demographically distinct from verbal hallucination (in which voices are heard) and has different neural correlates (Izumi et al., 2002).

Demographics

MH is much less common than tinnitus (described above). To date there have been no robust prevalence studies in large unselected populations. Amongst 3678 general psychiatric admissions, only 0.16% reported MHs (Fukunishi et al., 1998); this comprised 6 cases, of whom 5 were female, 3 were hearing-aid users, and 3 elderly. In the at-risk group of elderly individuals with hearing impairment, prevalence in small samples ranges from 0.8% (Cole et al., 2002) to 3.6% (Teunisse and Olde Rikkert, 2012). A salutary lesson on the subjectivity of survey questions comes from Goycoolea et al. (2007), who report "spontaneous musical sensations" in 39.4% of a group of 150 otolaryngologists and 97% of a group of 100 musicians.

There is a female preponderance of approximately 3:1, even accounting for the fact that women live to an older age and are more likely to live alone in old age (Cope and Baguley, 2009). Socially isolated individuals with hearing loss are more likely to be affected. MH is more common in those over 60, but there is no apparent increase in risk beyond this (Berrios, 1990); it is unclear to what extent age is an independent factor, and how much it is merely co-associated with hearing loss,

vascular and neurological pathologies, social isolation, and pharmacological treatment.

Investigations

There is an association between MH and hearing loss, and initial investigation should always include measurement of audiometric thresholds. While organic brain pathology in a number of regions can give rise to MH, and some form of brain imaging should be performed to rule out a structural lesion, this is normally unrevealing. Paroxysmal MH as a primary manifestation of epilepsy is very rare (Couper, 1994), and electroencephalography should only be performed if there are other grounds for clinical suspicion. More intensive investigation for organic brain disease should be triggered by transient visual disturbances, disabling dizziness, severe headache, abnormal speech or neurological examination, or an audible carotid bruit.

A mental state examination should be performed, primarily assessing mood. Depression is common in MH, affecting around a third of elderly sufferers (Aizenberg et al., 1987). This should be probed for in the history, and treatment of depression can often improve MH. It is unclear whether this occurs simply through reducing social isolation or whether the interplay is more complex. MH is also common in patients with obsessive compulsive disorder (OCD): patients with OCD have an approximately 40% lifetime risk of experiencing MH, although most patients with MH do not have OCD. MH is, however, exceedingly rare in patients with schizophrenia and related psychoses (Hermesh et al., 2004).

Is it functional?

An attractive model for the pathogenesis of MH relies on the concept of peripheral and central "disinhibition" (Griffiths, 2000). Reduced auditory inputs (due to hearing loss and social isolation) combine with reduced inhibition from higher centers to increase the "gain" of association auditory cortex. This leads to an increasing tendency to interpret "system noise" as musical, and imagined musical imagery (earworms) as perceptually salient. Mechanistically, this can be understood within a "predictive coding" framework (Kumar et al., 2014a); as the sensory signal becomes degraded and prior expectations become more precise, perceptual inference is abnormally shifted. Although the underlying causes differ, as noted above, this mechanism bears a striking resemblance to the evolving understanding of functional somatosensory and motor phenomena (Edwards et al., 2012).

Treatment strategies

Although the framework for understanding MH is similar to that of functional sensory and motor phenomena, it has a number of underlying drivers that should be addressed before psychological therapies are considered. Specifically, hearing loss should be corrected and, if possible, social isolation and low mood addressed. It should be stressed that MH is not a sign of dementia or psychosis, as this is a common concern amongst patients, and a thorough explanation of symptoms should be provided. If an underlying cause is suspected, treatment of this can often resolve MH, but beyond this, there is little consensus regarding optimal therapy. While the literature most commonly reports pharmacological success with antipsychotics and anticholinesterases, before embarking on this course it should be borne in mind that MH is often not particularly distressing and, after reassurance that it does not signify more concerning pathology, patients are often happy to coexist with their musical experiences.

LOW-FREQUENCY NOISE COMPLAINT

Definition

A small number of individuals have a persistent complaint of low-frequency noise (LFN) in their environment (usually the home), causing them severe physical and emotional distress. In comparison with patients with tinnitus, those with LFN complaint are insistent that the source is external rather than internal. Complainants tend to describe humming or rumbling, often accompanied by a feeling of pressure on the ears or vibration in the body – a common descriptor would be that of a "distant engine." When acoustic measurements are undertaken, an LFN signal can be identified in only 30% of cases (and in such cases there is hope of a noise control solution), leaving uncertainty about the etiology of the LFN perception in the majority of cases (Moorhouse et al., 2005). The incidence of LFN in homes where there is no LFN complaint is unknown.

The distress of the LFN complainant can be severe and on occasion debilitating. Physical agitation can be marked, as can the emotional reaction, including fear and aversion to the home environment, and a component of hypervigilance to LFN can be observed. The LFN is perceived to be worse at night, and may lead to insomnia, and affected individuals may resort to sleeping in a car away from their property. One individual is usually affected in a household, but in some cases a partner or relative may develop some awareness of LFN at some later date. In rare circumstances many members of a community may complain of LFN (Pedersen et al., 2008). Complainants often have a definite belief about the source of the LFN, and this may include a component of the agent causing the LFN (e.g., a company or local council) being dismissive or antagonistic. Suggestions that there may be a tinnitus component to the percept may be met with dismay and disbelief. Reports of this

phenomenon have been noted in the UK (Tempest, 1989), the Netherlands (Oud, 2012), Denmark (Møller and Lydolf, 2002), and Sweden (Persson and Rylander, 1988).

Demographics

Data on the epidemiology of LFN complaint are very sparse. Surveys of complainants have indicated that this phenomenon is more likely to be reported in middle age, with a mean age of 55 reported (Pedersen et al., 2008). Two-thirds of LFN complainants are female (Leventhall, 2003).

Investigations

There are two aspects to the investigation of LFN complaint: that of the individual, and of the environment. Regarding the individual, otoscopic and audiometric examination is required, as is a careful history for tinnitus, hyperacusis, and for anxiety and depression both prior to, and associated with, the LFN. An assessment should be made of the risk the individual represents to him- or herself (e.g., self-harm, or physical harm due to sleeping outside the home) and to any others that the patient believes are involved in generating the noise or disinclined to make it cease.

Testing low-frequency hearing thresholds (e.g., below 250 Hz) is not feasible in most clinical environments. When it has been performed in laboratory conditions, LFN complainants are found to have normal LF thresholds, but when asked to set acceptable levels, do so at a lower intensity than noncomplainers (Leventhall et al., 2008).

A structured protocol for the environmental investigation in an LFN complaint is available (Moorhouse et al., 2005), produced in the UK to support environmental health officers involved in such cases and ensure definitive investigation. Appropriate specialist recording equipment is needed, and should run through several nights. When no LFN source is identified, complainants often opine that the recording was done on an occasion when the LFN was unexpectedly absent.

Is it functional?

In a LFN complaint where no noise source is found, there are at least two possibilities:

1. that the individual has an LF tinnitus, and mistakenly attributes this to an external source
2. that the individual has become sensitized to environmental LF sound, and is experiencing some form of LF hyperacusis.

Understanding of the mechanisms by which the distress has arisen rests upon the more classic presentations of tinnitus and hyperacusis, specifically the links between the auditory brain and systems of learning, vigilance, and threat reaction.

An alternative model of heightened awareness of LFN was proposed by Salt and Hullar (2010), who contended that outer hair cells in the cochlea may be activated by low-frequency sound at subthreshold levels, and that in certain conditions an individual may become aware of that stimulus. Whilst this theory has not been substantiated with physiological evidence, the possibility that LFN complainants may be experiencing a psychophysical phenomenon rather than a heightened or overvigilant response to environment sound should not be discounted.

Treatment strategies

Acoustic masking of LFN is not feasible as most masking devices have little output below 250 Hz. Informational masking (i.e., utilizing an alternative sound that has properties that capture the attention of the auditory brain) may have a role, specifically the use of rain/ocean-type environmental sounds at the bedside throughout the night. Hearing aids may fulfill this function during the day.

Three studies have investigated the benefits of CBT-like interventions. Leventhall et al. (2008) evaluated sessions in a small group ($n=9$) involving information, imaginal exposure exercises, and relaxation, delivered by an experienced psychotherapist, with moderate benefits. Similar material was used in book and online formats in a larger study ($n=27$ completers from a group of $n=46$ who agreed to participate), with similar results, though no intention-to-treat analysis was undertaken (Leventhall, 2009). Moorhouse et al. (2015) trialed a treatment protocol delivered by audiologists including information, attentional masking and relaxation – benefits were positive but modest. No data regarding long-term benefit are available. We are unaware of studies evaluating the efficacy of CBT in LFN complaint.

DISORDERS OF LOUDNESS PERCEPTION

Definition

Disorders of sound tolerance fall into two categories: dislike of sound above a certain intensity and dislike of particular sounds irrespective of their level. Terminology is confusing and still developing. Hyperacusis is a word used both as a blanket term to cover all types of impaired sound tolerance and to define a specific subtype. When used in the specific instance, hyperacusis refers to a dislike of all sounds above a certain level. Recruitment is a condition seen in association with significant sensorineural hearing loss in which rising sound intensity causes a greater than normal rise in perceived loudness.

Misophonia is a strong dislike of certain sounds, irrespective of their level, and is discussed separately. Phonophobia is a variant of misophonia in which the dominant emotion is fear. These definitions have recently been challenged and a new classification of loudness hyperacusis, annoyance hyperacusis, fear hyperacusis, and pain hyperacusis has been proposed (Tyler et al., 2014). Different types of impaired sound tolerance may coexist in the same patient.

Demographics

There are very few epidemiological studies regarding impaired loudness tolerance. Studies in Sweden (Andersson et al., 2002) and Poland (Fabijanska et al., 1999) showed a hyperacusis prevalence of 8.6% and 15.2% of their adult populations respectively. Neither of these studies attempted to assess the severity or impact of the symptom. A study in Brazil (Coelho et al., 2007) found a hyperacusis prevalence of 3.2% in the pediatric population. There is a strong comorbidity of hyperacusis and tinnitus: 40% of people with tinnitus as their main complaint report some degree of hyperacusis, whereas 86% of those who present with hyperacusis will also describe tinnitus.

Investigations

The investigation of patients with impaired sound tolerance is largely the same as for patients with tinnitus (see above), with a few additional caveats. Tympanometry, particularly when used to estimate stapedial reflex thresholds, involves significant sound levels and MRI scanning is notoriously noisy. Patients with impaired sound tolerance need careful counseling prior to such tests and if necessary the test should be deferred or a quieter alternative sought. Loudness tolerance can be estimated using standard audiologic equipment to measure the loudest sounds that the patient can tolerate at particular frequencies (loudness discomfort levels). Because of the sound levels involved, such tests run significant risk of distressing the patient and any clinical benefit is usually outweighed by the risk of losing the patient's trust.

Is it functional?

Hyperacusis is occasionally associated with facial nerve palsies which cause loss of the ear's protective stapedial reflex. In such cases, because the symptom is associated with a demonstrable lesion, it cannot be regarded as functional. The vast majority of cases of hyperacusis, however, are not associated with structural pathology. Although various pathophysiological mechanisms have been suggested, the cause remains unknown. Some theories are directed at the auditory periphery but many focus on the central auditory system, proposing similar mechanisms to those seen in tinnitus (see above). It therefore seems likely that impaired loudness tolerance has a functional basis in at least a proportion of cases.

Treatment strategies

Education and reassurance are important treatment components. In particular, many patients with significant hyperacusis protect themselves from sound by seeking quiet environments or by wearing sound-attenuating devices. Although this seems sensible, it is hypothesized to result in increased central auditory gain, which exacerbates the problem. Careful reintroduction of sound is one of the mainstays of hyperacusis treatment. Sound therapy can be used to improve sound tolerance using continuous low-level sound in a technique called recalibration or by slowly increasing sound in a technique called desensitization. As with tinnitus, protocols been developed that use several treatment modalities in a structured way. These include tinnitus retraining therapy (see section on tinnitus, above) and hyperacusis activities treatment (Tyler et al., 2009). Psychological treatments, particularly CBT, have been tried with some benefit (Jüris et al., 2014).

MISOPHONIA

Definition

Misophonia is a disorder of the emotional processing of specific sounds, and can be literally translated as "hatred of sound" (Jastreboff and Jastreboff, 2001). Background sounds that would be generally described as perhaps mildly irritating, such as eating, noisy breathing, and typing, produce a strong sense of anger, and either aggressive or aversive behavior in sufferers (Schroder et al., 2013). This effect is distinguished from hyperacusis in that it is restricted to particular sounds, individual to each sufferer, and does not relate to the spectral properties of the auditory stimulus that contribute to the general unpleasantness of sounds such as nails on a chalkboard (Kumar et al., 2012), although these conditions can coexist (Jastreboff and Jastreboff, 2015). It should be emphasized that the dominant emotion is almost always anger and therefore misophonia is not a true phobia, but many sufferers will adopt avoidance behaviors for situations where trigger sounds might occur.

Demographics

Age of onset is variable, but symptoms often emerge in childhood or adolescence and persist into adulthood (Kumar et al., 2014b), with an average latency before formal diagnosis of 25 years. This is likely to be an

underestimate, as it only records that minority of sufferers seeking medical assessment. Males and females appear equally likely to be affected. Approximately 50% of sufferers have anankastic (obsessive compulsive) personality traits, but true OCD and other psychiatric comorbidities are rare (Schroder et al., 2013).

Investigations

There are no reports of misophonia arising as a result of organic brain pathology, so unless there are other grounds for clinical suspicion, routine brain imaging is not necessary. Mental state examination should be performed as the detection of psychiatric comorbidities or personality traits can be useful in tailoring the treatment approach.

Is it functional?

The exact mechanisms of misophonia are unclear, and are a topic of ongoing investigation (Kumar et al., 2014b). Trigger sounds are associated with abnormal autonomic effects that do not generalize to visual stimuli (such as a video of chewing gum) (Edelstein et al., 2013). Current conceptualizations rely on repetitive minor annoyances associated with sounds and culminating in pathological emotional valence (LeDoux, 2000), but evidence of this from the patient history is rarely present (Schroder et al., 2013).

Treatment strategies

There are no randomized controlled trials of treatment methods. Explanation and validation of the condition can be helpful. Pragmatic strategies such as using noise-canceling headphones at mealtimes can be employed. Beyond this, currently employed strategies involve habituation and retraining therapies similar to those used in tinnitus and hyperacusis (Jastreboff and Jastreboff, 2015), as well as CBT and other psychological interventions.

ACOUSTIC SHOCK

Definition

Acoustic shock (also known as acoustic shock syndrome or acoustic shock disorder) describes a group of symptoms seen in people who have been exposed to sudden unexpected sounds. Initially recognized in people working in call centers using headsets, the symptom cluster has also been seen following exposure to a variety of other sound sources, particularly when the causative sound is generated close to the ear. The commonest symptom reported is pain in or close to the ear, followed by tinnitus, hyperacusis, balance disturbance, hypervigilance, and sleep disturbance (Milhinch, 2002). The level of the causative sound seems relatively unimportant compared to its rise time: it seems to be the suddenness rather than the loudness that is the issue. Hearing loss occurs in fewer than 1 in 5 people with acoustic shock and when it does happen it does not necessarily have the characteristics of noise-induced hearing loss.

Demographics

There are no reliable epidemiological data regarding acoustic shock. Initial reports were from Australia, Denmark, and the UK, but anecdotally the symptom is recognized globally.

Investigations

The investigation of a patient with suspected acoustic shock is the same as for patients with tinnitus or disorders of loudness perception (see above).

Is it functional?

Suggestions for the pathophysiology of acoustic shock include cochlear damage, tonic contraction of the tensor tympani muscle (Westcott, 2006), or psychological mechanisms. In many cases there is no measurable deficit within the peripheral auditory system, suggesting that a functional origin is likely. There have, however, been suggestions that at least some cases are attributable to malingering (Hooper, 2014).

Treatment strategies

Treatment is largely the same as for disorders of loudness perception (see above). If the acoustic shock occurred in a call center environment, adjustments to the patient's job may be required. Electronic devices to try and suppress causative sounds and limit overall sound exposure while maintaining speech clarity have been developed for telecommunications equipment within call centers.

CONCLUSION

There are a number of auditory symptoms that appear to have a functional component, some of which affect similar populations to those with functional neurological symptoms, whilst others affect very specific populations (notably MH and misophonia). Understanding in this area is emergent, and treatments are in many cases pragmatic and rudimentary, needing more work to be done in integrating insights from behavioral and cognitive psychology to auditory neuroscience. The audiology literature has historically equated the term functional with malingering, and more work needs to be done in developing interest and expertise in these conditions.

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