PERSPECTIVESon

Hearing and Hearing Disorders: Research and Diagnostics

SPECIAL INTEREST GROUP 6

AMERICAN SPEECH-LANGUAGE-HEARING ASSOCIATION

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Coordinator's Column

Jeanane M. Ferre

Welcome affiliates to the spring 2015 edition of SIG 6's *Perspectives on Hearing and Hearing Disorders: Research and Diagnostics.* As professionals in hearing and listening difficulties, we are often the first contact for clients experiencing difficulty listening in noise, following directions, remembering verbal messages, and/or using verbal information at work, at school, or in social settings. We administer behavioral and/or electrophysiological tests of hearing and middle ear function, discovering normal peripheral auditory function. So, now what? As audiologists—responsible for evaluating auditory function from outer ear to cortex—we must familiarize ourselves with those conditions that can create auditory complaints in the absence of demonstrable peripheral hearing impairment. Our first issue of 2015 offers you four papers discussing just such conditions. Jeanane Ferre provides an overview of central auditory processing assessment and intervention among listeners with normal peripheral function. The auditory sequalae associated with mild traumatic brain injury are discussed by Eric Hoover, Pam Souza, and Frederick Gallen. Helen Pryce discusses a fascinating syndrome—King-Kopetzky syndrome—that presents with normal hearing and auditory difficulties that may or may not be true auditory processing disorder; while Garreth Prendergast, Hannah Guest, and Christopher Plack examine relations among cochlear neuropathy, hidden hearing loss, and obscure auditory dysfunction.

SIG 6 appreciates our authors' contributions of time and expertise to bring you this *Perspectives* issue to you. *Perspectives* cannot happen without the work of an amazing team of volunteers: Editor, Sumit Dhar, Associate Editor, Ashley Harkrider, Editorial Reviewers, Marilyn Dille and Marc Brennan, and our Professional Development Manager, Peter Ivory. Our SIG 6 team is assisted by ASHA National Office staff, including Deborah Berndtson, our ex-officio and Victoria Davis, Production Editor.

Coming in the fall *Perspectives* issue, look for articles focusing on fluctuating hearing loss. Compelling readership numbers make it clear how much affiliates value *Perspectives* for access to cutting-edge information and interesting topics as well as professional development credits. Keep in mind that other opportunities for education and involvement abound. The coordinating committee (Chris Sanford, Susan Gordon-Hickey, Wafaa Kaf, Sumit, Peter, and myself) along with our SIG 6 liaison to the ASHA Convention Program Committee, Peter Ivory, have been busy since January working with the neuroanatomy/neurophysiology topic area and SIGs 7, 8, and 9 to bring you some exciting courses at this year's ASHA Convention in Denver. Log on to the SIG 6 website to share your ideas for future *Perspectives* topics and/or authors, express your concerns, engage in discussion on topics of interest to our community, or volunteer your time and expertise to the SIG 6 mission.

In April, the Coordinating Committee will be meeting at the National Office to work on additional opportunities for professional development, discuss upcoming *Perspectives* issues, monitor our work plan progress to date, and share information with ASHA staff working on issues important to audiologists and hearing scientists. We always need more volunteers to help with these efforts. If you're interested in becoming more involved, contact me at jmfphd@comcast.net or through the SIG 6 online ASHA community.

Auditory Dysfunction Beyond the 8th Nerve: Understanding Central Auditory Processing Disorders

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Financial Disclosure: Jeanane M. Ferre is an audiologist at a private practice.
Nonfinancial Disclosure: Jeanane M. Ferre has previously published in the subject area.

Abstract

Central auditory processing disorder (CAPD) refers to a deficit in the neural processing of auditory stimuli that can affect listening, language, and learning. Because CAPD manifests behavioral symptoms similar to those exhibited by listeners with peripheral hearing loss, it is important that the audiologist be prepared to "go beyond the 8th nerve" in the assessment process to evaluate central auditory skill sets and provide intervention as needed. Differential diagnosis of these disorders is accomplished using behavioral and electrophysiological tests that examine the array of auditory skills and integrity of the system from brainstem through the cortex. Test results are used to develop effective deficit-specific intervention plans designed to reduce/resolve the deficit and minimize the disorder's affect on the listener's life.

Central auditory processing disorder (CAPD) refers to a deficit in the perceptual (i.e., neural) processing of auditory stimuli and the neurobiological activity underlying that processing (American Speech-Language-Hearing Association [ASHA], 2005). Neuro-geographically, the central auditory system ranges from the cochlear nuclei through the cortex and includes the neural encoding of frequency and temporal cues by specific afferent centers as well as the binaural representation of those cues within the system. Both hearing loss and central auditory processing disorders can adversely affect communication, learning, and psychosocial wellness. Thus, the listening/hearing complaints of the listener with a CAPD are similar to those of individuals with peripheral hearing impairment. In fact, many disorders present behavioral characteristics similar to CAPD that may lead to similar functional listening difficulties. Disorders co-existing with and/or sharing symptoms of CAPD include neurocognitive disorders (e.g., attention deficit hyperactivity disorder—ADHD, executive function disorder), cognitive impairment (e.g., mental retardation), communication disorders (e.g., autistic spectrum disorder, Asperger's syndrome, language processing disorders, specific language impairment), social-emotional disturbance (e.g., behavior disorders), learning disability, and other sensory processing impairments (e.g., sensory integration disorder; ASHA, 2005; Bellis, 2006; Ferguson, Hall, Riley, & Moore, 2011; Keller, Tillery, & McFadden, 2006; Sharma, Purdy, & Kelly, 2009; Witton, 2010). Through differential diagnosis, the assessment team uncovers the nature of the underlying disorder contributing to a listener's functional challenges and maximizes the intervention for those challenges.

Differential diagnosis refers to the differentiation among two or more disorders that have similar symptoms and/or manifestations (ASHA, 2005; Bellis, 2006, 2014). Comprehensive differential evaluation of CAPD requires input from a variety of disciplines, including, but not limited to, audiology, speech-language pathology, neuropsychology, occupational therapy (OT), physical therapy (PT), education, and other related professions. The audiologist administers well-controlled tests, sensitive to dysfunction in the central auditory pathways, thus clarifying the *auditory* component, if present. Other professionals provide information regarding the extent to which there exist difficulties in other sensory processing skills and/or in higher-order cognitive, linguistic, or learning skills that may confound auditory test results or co-exist with a CAPD.

In general, the central auditory skill sets include auditory discrimination, binaural processing and temporal processing, and within these sets, specific skills have been identified. Sound localization and lateralization depend upon discrimination and binaural processing. Specific temporal processing includes temporal discrimination, ordering, integration, and masking. Auditory performance in the presence of competing signals requires dichotic listening, dependent upon binaural integration and separation. Some binaural tasks also tax discrimination (e.g., binaural fusion). Tests of auditory performance with degraded acoustic signals tax both auditory closure and discrimination skills. Finally auditory pattern recognition is associated with temporal and discrimination functions (ASHA, 1996, 2005; Bellis, 2003; Chermak & Musiek, 1997). With a goal of understanding the central auditory "lay of the land," as it were, a comprehensive assessment will include both formal and informal measures designed to assess all of these skills.

There is general agreement among audiologists that a battery of tests is essential for the differential diagnosis of a CAPD. While there is less agreement on exactly which specific tests should be included, there is agreement that a comprehensive diagnostic assessment should include verbal and nonverbal tasks as well as behavioral and electrophysiologic/electroacoustic measures. The central auditory evaluation may include the following elements:

- puretone air and bone conduction audiometry to examine peripheral hearing acuity;

- speech reception threshold and word recognition tests to establish baseline word recognition abilities;

- otoacoustic emissions, tympanometry, acoustic reflex, and reflex decay to rule out middle ear disorder, identify retrocochlear dysfunction, and/or differentiate a CAPD from auditory neuropathy;

- measures of brainstem level binaural interaction that may include binaural fusion, masking level difference, and/or assessment of localization to a sound in space;

- low pass filtered and/or time compressed speech tests to tax auditory discrimination;

- dichotic listening tests having varying linguistic loads to assess cortically-based binaural integration and separation skills;

- pitch and/or duration sequencing tasks, "global" measures providing information regarding the integrity of right-hemisphere-based pattern recognition, frequency or temporal discrimination, and interhemispheric transfer of function;

- electrophysiological assessment of the brainstem and/or cortex, e.g., ABR, cABR, MLR, MMN, P300 (Kraus & Hornickel, 2013; Schochat, Rabelo, Musiek, 2014);

- speech-in-noise tests that provide information regarding the listener's functional abilities in various listening situations.

Central auditory processing tasks are measures that, in general, examine how efficiently the auditory system operates by "overloading" or "overworking" it. Central auditory tests go beyond standard tests of hearing to examine how well the auditory system uses or interprets the information that the ear sends it. Results are compared to an age-matched peer group and performance profiles emerge that provide insights into the nature of the CAPD. As with any other clinical decision-making paradigm, it is the responsibility of the examiner to understand fully the nature and limitations of specific central auditory tests as well as the test-taking needs of the client, including chronological and/or developmental age. There remains on-going debate regarding the youngest age for which central auditory assessment is appropriate. While an in-depth discussion of this debate is beyond the scope of this paper, a brief discussion of central auditory skill development is appropriate.

As noted above, central auditory abilities include binaural interaction, auditory discrimination, auditory pattern recognition, dichotic listening, and interhemispheric integration.

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Neurodevelopmentally, these skills are present and measurable by 6 years of age, with some skills apparent at much younger ages than others (Bellis, 2003). Reliable diagnostic testing can be accomplished beginning at age 6 as there are versions of most central auditory tests with vocabulary and normative data available for children 6 years and older. For children between the ages of 4–6 years, an assessment including formal tests and informal screening indices examine the precursors of auditory processing. While not specifically diagnostic in nature, the "preschool" assessment provides information regarding a child's strengths and weaknesses with respect to developing auditory processing skills and can serve as a baseline for later assessments, as needed. To date, there are no *behavioral* central auditory tests appropriate for use with children under age four. By understanding the nature of the skills subserved by the peripheral and central auditory systems, we can assess development of the skills, draw inferences regarding impact of impaired or delayed skill development on the child's life, and develop deficit-specific intervention programs designed to reduce/resolve deficits and minimize adverse effects.

Binaural interaction refers to how well the two ears work together and reflects integrity of the auditory system at the low brainstem level. Binaural interaction assists the listener in localizing a sound in space and is important for attention, and listening in noise. Rudimentary binaural interaction ability is present by age two, as suggested by auditory brainstem testing.

Auditory discrimination is the ability to analyze fine differences in the speech spectra that contributes to recognition of running speech, recognition of speech in noise, phonemic/phonologic awareness, and language development. The ability is present at birth, becoming restricted to native language sounds by approximately 12 months of age, and becoming adult-like by age 10 (Bellis, 2003).

Temporal pattern recognition is the ability to identify and/or recognize auditory patterns (i.e., signals with more than two separate acoustic events). Pattern recognition depends on intact temporal resolution ability—the ability to "hear" where one sound ends and another begins—as well as intact right hemisphere function. Pattern recognition contributes to the listener's ability to recognize and process running speech as well as to perceive *intent* of a message, such as in sarcasm. Temporal resolution improves significantly from ages 3–5 years and is adult-like by age 10; while specific pattern recognition is apparent initially at approximately age 6 years and matures through age 12 (Bellis, 2003).

Interhemispheric integration refers to the communication between the two hemispheres across the corpus callosum that contributes to our ability to process increasingly lengthy or complex speech, recognize competing auditory targets, follow directions, synthesize multiple targets as in phonologic processing, transition from task-to-task, and complete assignments in timely fashion. Integration contributes to overall processing speed/efficiency as well as to the ability to manage the breadth and depth of sensory information that we encounter daily. Corpus callosum (i.e., interhemispheric connections) development begins during the first year of life but is not fully mature until early adulthood and is highly variable in listeners under age six (Bellis, 2003).

Dichotic listening refers to the ability to process different information presented to each ear simultaneously. Dichotic listening assists listeners in processing multiple incoming acoustic targets (binaural integration) and/or to ignore target in presence of competing signal (binaural separation). Dichotic listening is present and measurable by age 5, remaining highly variable until age 8, and reaching adult-like values by age 12 (Bellis, 2003).

Central auditory processing disorders (CAPD) do not exist in a vacuum and many clients come to the evaluation with co-existing concerns including peripheral hearing impairment, especially among our aging clients, receptive and/or expressive speech-language issues (e.g., dyspraxia, poor receptive vocabulary, and aphasia), and/or neurocognitive or behavioral issues (e.g., TBI, cognitive impairment, attention deficit disorder, and emotional dysregulation or disorder). While these co-existing challenges do not necessarily exclude the individual from assessment,

consideration must be given to them as these factors may require an adjustment to the diagnostic protocol and/or confound test interpretation. Excellent discussions of considerations for diagnostic testing may be found in Bellis (2003); Geffner & Swain (2013), and Musiek & Chermak (2014).

Understanding central auditory processes (CAP) and ably administering a CAP test battery represents only half of the CAPD equation, the examiner must now turn attention to interpreting the results *and* developing a deficit-specific intervention plan. To that end, there exist CAP test profiling systems that assist the examiner in conceptualizing and clarifying test findings. While multiple models exist representing differing conceptualizations of CAPD, all share the notions that a multidisciplinary approach is needed for the assessment process, a test battery is needed for specific CAPD diagnosis, test scores should relate to functional needs, and intervention should be collaborative in order to be effective (ASHA, 2005; Bellis, 2003, 2014; Chermak, 2007; Ferre, 2006). Diagnostic interpretation using one such model is described here.

Bellis and Ferre (1999) outlined five central auditory processing test profiles that describe the nature of the disorder based on key central auditory test findings and measures of cognition, communication, and/or learning, and associated behavioral manifestations. The model is a theoretical framework in which individual test scores as well as inter- and intra-test patterns of performance are examined in order to relate central auditory test findings to both their presumed underlying neurophysiological bases and functional sequalae (ASHA, 2005; Bellis, 2003, 2006; Bellis & Ferre, 1999; Ferre, 2002, 2006). The model describes three primary central auditory deficit profiles characterized by presumed underlying site of dysfunction. Two secondary profiles yield unique patterns of results on central auditory tests; however, they may be described more appropriately as manifestations of supramodal or cognitive-linguistic disorders. For a comprehensive discussion of these deficit profiles, the reader is referred to Bellis (2003, 2006), and Bellis and Ferre (1999). For the purposes of this discussion, they are described briefly here.

Auditory decoding deficit is a deficit in auditory closure and related sound discrimination representing dysfunction in the primary (usually left) central auditory pathways. On central auditory tests, the profile is characterized by difficulty on tests of degraded speech tests (e.g., recognition of filtered or time-compressed targets) and/or measures of temporal discrimination (e.g., temporal gap detection). Binaural and/or right ear deficits may be observed on dichotic listening tests, especially those with relatively substantial linguistic demand (e.g., dichotic words versus dichotic digits). Poor discrimination means the listener's auditory system is working harder than that of a typical listener to extract the fine acoustic changes within the speech spectrum, even under optimal conditions. This difficulty places the listener at risk for listening difficulties when noise is present, in highly reverberant environments (e.g., arenas, restaurants, playgrounds, etc.) when extra visual and/or contextual cues are not available, or when listening to a soft-spoken speaker or one with a pronounced accent. As the acoustic or linguistic conditions deteriorate, more neural energy is expended to process the acoustic portions of the signal, leaving less energy for higher-order linguistic-cognitive processing. Processing inefficiency can result in fatigue and reduced listening comprehension. These behavioral listening difficulties are similar to those observed among listeners with peripheral hearing loss. Like the listener with peripheral hearing impairment, secondary psychosocial issues may arise including social withdrawal or depressive disorder. Auditory decoding deficit can create secondary difficulties in communication (e.g., vocabulary, syntax, semantics, and second language acquisition) and/or academic skills (e.g., reading decoding, spelling, notetaking, and/or direction following; Bellis & Ferre, 1999).

Integration deficit reflects deficient ability to recognize and use multisensory incoming cues quickly and efficiently, believed to be the result of inefficient communication across the corpus callosum. The profile is characterized on central auditory tests by excessive left ear suppression on dichotic listening tests combined with poor labelling but adequate imitation of tonal patterns (e.g., pitch sequencing test). Deficit in skills needed for information integration may affect listening comprehension, especially in groups, academics, and higher-level language processing.

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Deficits in other integrative skills (e.g., visual-motor, auditory-visual, etc.) are common with this profile (Bellis & Ferre, 1999; Ferre, 2002, 2006). As listening demands increase, the listener may become less tolerant of extraneous distraction. Fatigue may set in and listening attitude may deteriorate with the listener appearing inattentive or confused (Ferre, 2006, 2007). In the assessment process, impaired auditory integration should be differentiated from attention deficit, sensory dysregulation, executive dysfunction, or general anxiety or depression.

Impaired auditory pattern recognition, regardless of response mode (labeling or imitation) as well as excessive left ear suppression on dichotic listening tests suggests a prosodic deficit, a deficiency in using prosodic features of the signal, a predominantly right hemisphere function. Running speech can be conceptualized as a series of acoustic patterns to which specific meaning must be attached for comprehension to occur. As we speak, we often drop word endings and blur perceptual timing boundaries by failing to enunciate clearly (Picheny, Durlach, & Braida, 1985, 1986). In everyday communication, the listener must navigate between and among these rapidly changing acoustic patterns; analyzing, synthesizing, manipulating, and attaching meaning to them quickly and efficiently. A non-impaired listener is able to perceive the *ebb and flow* of these changing patterns in the speech stream and make sense of the signals even when they are disrupted. The listener with prosodic deficit has difficulty recognizing the acoustic contours (i.e., patterns) in the rapidly occurring speech stream and perceiving timing cues in running speech (e.g., pacing, segmentation, and rhythm cues). Prosodic deficit may manifest as inconsistent processing of rapid speech and/or difficulties listening in highly noisy or reverberant environments, when listening to unfamiliar vocabulary, an unfamiliar speaker, or to someone not speaking clearly. The listener may misperceive the intent of the message or perceive one that is very different from that which was spoken, resulting in miscommunication (Bellis, 2003).

Functional challenges may be seen in reading, spelling, direction following, note-taking, attention, working memory, and problem-solving. Ability to recognize and use other types of sensory patterns (e.g., visual and/or tactile) may be impaired. Communication problems of the listener with prosodic deficit can include difficulties in syntactic, semantic, pragmatic, and social language skills, including difficulty understanding sarcasm and recognizing and using nonverbal pragmatic language cues such as facial expressions, body language, and gestures (Bellis, 2006; Bellis & Ferre, 1999; Ferre, 2007).

As noted above, binaural processing, including binaural integration and separation, is a fundamental central auditory skill set. A primary CAPD can impair binaural processing through impaired discrimination, as in the decoding profile; impaired interhemispheric communication, as in the integration profile, or impaired right hemisphere function, as in the prosodic profile. However, poor performance on tests of binaural processing also may result from inefficient *intra*-hemispheric communication; the presumed underlying cause of associative deficit. A secondary central auditory processing test profile, this deficit is characterized by significant auditory-language processing difficulties, believed to be related to dysfunction in the communication between the primary (Heschl's gyrus) and secondary or auditory association (Wernicke's area) cortices of the dominant (usually left) hemisphere (Bellis & Ferre, 1999; Ferre, 2002). On central auditory tests, the ability to recognize degraded speech and temporal patterns is age appropriate with marked difficulty for one or both ears on dichotic listening tasks taxing binaural integration and separation skills. This intra-hemispheric deficit impacts language processing and the listener has difficulty attaching linguistic meaning to incoming acoustic signals quickly and efficiently (i.e., associating the auditory with language). In general, listeners with this auditory-language association deficit don't extract key words from running speech as well as their peers and appear not to *speak the same language* as their peers. The listener tends to take most statements literally and often sees ambiguity even in seemingly straightforward messages (Bellis, 2003, 2006; Bellis & Ferre, 1999; Ferre, 2006). Listening difficulties arise when vocabulary is unfamiliar; information is presented without sufficient contextual or visual cues, or when the message is linguistically ambiguous. Because this deficit is more accurately conceptualized as a language impairment, the daily living challenges for this listener center on issues of comprehension—reading or listening,

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specific language usage, and social/pragmatic communication (Bellis, 2003, 2006; Bellis & Ferre, 1999; Ferre, 2006).

As noted in the first paragraph of this paper, the "landscape" of CAPD is reflected in the action of the afferent auditory system. However, the "process" of processing is incomplete without considering the listener's ability to execute a response as evidenced by accurate and appropriate verbal or written responses or successful task completion. Certainly, impaired ability to discriminate, analyze, synthesize, and/or attach meaning to an incoming auditory signal can create disability in expression or execution skills. However, performance difficulties (e.g., poor direction following or task completion), may exist in the absence of receptive sensory or linguistic dysfunction. Disruptions associated with impaired expressive skills or difficulty executing a response may manifest on central auditory tests as an output-organization deficit profile. This secondary central auditory processing test profile is characterized by poor scores on tests requiring the reporting of multiple or precisely sequenced targets, with normal performance seen on single target and/or free recall tasks. Atypical crossed reflexes or otoacoustic emissions may be seen (Bellis, 2003). Difficulty in skills needed for information organization or recall may adversely affect planning, applied problem-solving, listening comprehension, direction following, spelling, verbal or written expression, word finding, and retrieval. Behaviorally, the listener may exhibit difficulty hearing in noise, be disorganized, impulsive, or present with issues in executive functioning. Although no specific neurophysiologic region of dysfunction is implicated by test findings, the central auditory test results and behavioral manifestations implicate the frontal and prefrontal cortices or efferent (i.e., motor) pathways as possible sites of dysfunction (Bellis, 2006; Bellis & Ferre, 1999; Ferre, 2002; Richard, 2001).

Central auditory processing profiles can occur singly or in combination, although one profile typically predominates. It is important to note that if the listener exhibits deficits in all auditory processes assessed or test results suggest the presence of more than two of the five functional deficit profiles, consideration should be given to the likelihood of global, higher-order, or bi-hemispheric dysfunction as the primary condition underlying reported or observed listening, language, or learning difficulties. In these cases, diagnosis of CAPD is not appropriate and referral should be made to related professionals (e.g., neuropsychology, neurology, and speech-language pathology) for additional assessment.

Although often overlooked, intervention for CAPD is an integral part of the audiologist's role and responsibilities. A detailed discussion of specific intervention strategies is not within the scope of this paper. However, key points regarding intervention deserve a brief mention:

- Differential intervention is a balance of deficit-specific management and treatment strategies that derive logically from diagnostic test results and have solid neuroscientific foundations.

- Effective intervention is timely and collaborative and includes treatment goals that are measurable, functional, and consistent with the neurodevelopmental hierarchy of auditory and communicative-cognitive function.

- In treatment, formal and informal therapy techniques are used to reduce or resolve auditory deficits and to teach functional compensatory strategies. In management, compensatory strategies and environmental accommodations are implemented to minimize the impact of the disorder on the listener's day-to-day functioning.

- There is no *silver bullet* for treating CAPD. A program that may be effective for one listener may be ineffective for another based on the specific auditory skills affected and the impact of the disorder on the listener's life. Before implementing any treatment program, the audiologist should verify the specific needs of the listener with CAPD in the classroom, workplace, and at home. In addition, the nature of the processing deficit

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should be described as fully as possible, as not all treatment programs may be beneficial for all types of CAPD.

- Regardless of type of deficit and functional sequelae, treatment for CAPD typically includes both bottom-up therapy, designed to reduce the deficit, and top-down therapy, designed to minimize residual effects of the disorder (ASHA, 2005; Chermak & Musiek, 1997; Bellis, 2003; Ferre, 2002).

- Auditory skill development as well as success of recommended treatment and/or management strategies, or lack thereof, should be documented at periodic intervals and adjusted as needed to meet the listener's functional needs.

Central auditory processing disorder (CAPD) can affect adversely academic achievement, speaking and listening, life skills, and sense of self. Differential diagnosis of CAPD and related, often co-morbid disorders leads to differential intervention. In differential diagnosis, formal and informal assessment results are interpreted in order to clarify the nature of the deficit that affects a listener's life. In differential intervention, deficit-specific recommendations for management and treatment are developed that create positive outcomes for our clients.

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Competing Views on Abnormal Auditory Results After Mild Traumatic Brain Injury

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Abstract

*Traumatic brain injury affects the lives of millions of Americans. Within audiology, there is
general agreement that mild traumatic brain injury (MTBI) can result in long-term auditory
processing deficits. However, this agreement is not shared across disciplines. In this paper,
recent studies on the effects of MTBI on auditory function are reviewed in the context
of competing opinions on the interpretation of neurosensory deficits after MTBI. Three
hypotheses are presented that explain auditory test results as they relate to post-traumatic
stress disorder, subtle cognitive deficits resulting from MTBI, and physiological damage to
temporal processing in the auditory system.*

There is broad agreement within the audiology community that mild traumatic brain
injury (MTBI) commonly results in long-term auditory dysfunction (American Speech-Language-
Hearing Association [ASHA], 2005, 2007; American Academy of Audiology, 2010; Gallun, Lewis
et al., 2012; Myers, Henry, & Zaugg, 2008; Myers, Wilmington, Gallun, Henry, & Fausti, 2009;
Schairer, 2012; Vander Werff, 2012). The preceding list of review articles and clinical practice
guidelines reflect a shared view from within the field of audiology, and no publications exist that
promote the dissenting view that auditory complaints after MTBI have non-auditory causes (to the
authors' knowledge). The agreement within audiology, however, is not shared across disciplines. In
fact, the claim that any long-term behavioral deficits can occur as a result of MTBI is controversial
in the field of neuropsychology (e.g., Belanger, Curtiss, Demery, Lebowitz, & Vanderploeg, 2005;
Rohling et al., 2011). Why is there a disconnect between the opinions of audiology and other
disciplines? Is it that professionals outside our field have strong evidence against auditory
dysfunction, or is it that the data are unconvincing? In the following article we present the recent
literature on the effects of MTBI on the auditory system in the context of the ongoing debate in
clinical neuropsychology on the underlying causes of long-term neurosensory deficits.

The Centers for Disease Control estimate that in the United States, 1.4 million people die or seek medical care due to acute head injury each year (Langlois, Rutland-Brown, & Thomas, 2006). The majority of cases are mild, at least 75% (Finkelstein, Corso, & Miller, 2006), but the true number of mild cases is likely underestimated because only 47% of people who suffer sports-related injuries seek medical attention (McCrea, Hammeke, Olsen, Leo, & Guskiewicz, 2004). MTBI, including concussion, is defined by the *Diagnostic and Statistical Manual of Mental Disorders, 5th ed.* as a neurocognitive disorder resulting from an insult to the head causing a period of confusion or disorientation, loss of consciousness less than thirty minutes, and post-traumatic amnesia of any duration (American Psychiatric Association, 2013). Due to the lack of physical symptoms in the majority of cases, MTBI is typically diagnosed by patient interview. The primary concern in acute MTBI care is stabilizing the patient and verifying that there is no concern of sub-cranial hemorrhage, damage to the spinal column, and external injuries to the skull, eyes, or outer and middle ears (Saatman et al., 2008). Testing for cognitive or neurosensory deficits may or may not be performed depending on the severity of the injury and the physical assessment. Despite the fact that neurological changes have been reported to persist after MTBI, both in humans and animal models (Turner et al., 2014), behavioral deficits are difficult to measure beyond the first two months post-injury. This may be due to an absence of behavioral deficits or to the sensitivity of the cognitive tests currently in use (e.g., Lux, 2007).

Although the effects may be difficult to quantify, many people report suffering symptoms after MTBI beyond the two month recovery window. These symptoms are collectively called post-concussive syndrome (PCS; R.C. Hall, Hall, & Chapman, 2005). PCS is thought to be related to post-traumatic stress disorder (PTSD), due to the overlap in PTSD and PCS symptoms (Hoge et al., 2008; Schneiderman, Braver, & Kang, 2008), and the fact that treatment for PTSD is an effective treatment for PCS (Mittenberg, Tremont, Zielinski, Fichera, & Rayls, 1996). The theory is that headaches, memory loss, sleep disturbances, hyperacusis, and the many other possible PCS symptoms are manifestations of PTSD, an anxiety disorder that developed as a result of the traumatic event. Cognitive behavioral therapy is used to address the underlying anxiety disorder and PCS symptoms. This means that auditory complaints, including "difficulty in noise," – are not referred to audiology, but instead treated using cognitive behavioral therapy administered by a neuropsychologist or speech pathologist (Management of Concussion/mTBI Working Group, 2009).

Referral to audiology may only occur when the injury is thought to affect pure-tone thresholds. Damage to the auditory system is a critical component in acute and long-term MTBI care, particularly in blast injuries. The initial evaluation of a patient who suffered a blast-related or impact MTBI includes examination of the outer and middle ear and the temporal bone (Saatman et al., 2008). Tympanic membrane perforation, ossicular chain discontinuity, and temporal bone fracture are common indicators of more significant injuries (Myers et al., 2009; Fausti, Wilmington, Gallun, Myers, & Henry, 2009). However, sensorineural hearing loss is rare in the absence of transverse fracture of the temporal bone (Bergemalm, 2003; Browning, Swan, & Gatehouse, 1982). Patients who suffer MTBI are often not referred to audiology, even when they have specific auditory complaints. In order to understand why, it is important to consider the MTBI patient from the perspective of the clinician leading post-acute care, typically a neuropsychologist.

MTBI From the Neuropsychologists' Perspective

Neuropsychology begins behavioral assessment once the patient is stabilized. A battery of tests of cognition and language may be performed, assessing attention, memory, processing speed, and other abilities typically affected by MTBI in the acute phase. At the top of the list of symptoms of concern after MTBI is headaches. Headaches are reported in 90% of cases of within one week of the injury and are treated with a combination of pharmaceuticals and non-pharmaceutical treatments (Mittenberg, Canyock, Condit, & Patton, 2001). Headaches typically recover quickly, leaving only 10–20% of cases with headaches after one month (Hartlage, Durant-Wilson, & Patch,

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2001). A much higher proportion of veterans continue have headaches after one month, up to 74% (Patil et al., 2011). Those with headaches that continue past the acute stage of MTBI are treated for PCS with a combination of pharmaceuticals and PTSD therapy to address the underlying cause.

While headaches are at the top of the list, hearing complaints are much further down. Other common MTBI symptoms include decreased concentration, memory problems, irritability, fatigue, dizziness, visual disturbances, judgment problems, depression, and anxiety (Langlois et al., 2006). Less common problems include insomnia, blurred vision, seizures, intolerance of stress, alcohol and drug abuse, behavioral inhibition, anxiety, grief, nightmares, lack of motivation, deceased sexual drive, anger, indecisiveness, excessive crying, and the list goes on. Eighty to 100% of cases experience symptoms, typically within the first week, and 85% of cases recover within the first week after the injury (McCrea et al., 2009). Ten to 25% report symptoms after two months, and it is these cases that are treated for PCS. Persistent symptoms and those that present more than one week after the injury are thought to be related to PTSD and treated with cognitive-behavioral therapy in conjunction with ad hoc interventions specific to the presentation. Hearing loss may be a low priority until other symptoms are in recovery.

At present, it is common for patients with auditory complaints to be treated with cognitive behavioral therapy. Many of those with auditory complaints are never referred to audiology (65% in Oleksiak et al., 2012). Guidelines for clinical management in the *VA/DoD Clinical Practice Guideline* (Management of Concussion/mTBI Working Group, 2009) lists referral to otology for physical examination of the tympanic membrane and temporal bone. Referral to audiology is recommended only when the source of hearing difficulty is not apparent. Presumably this limits the role of audiology to the treatment of sensorineural hearing loss, although in practice it is likely that a wider array of cases are referred to audiology. Auditory processing deficits are not mentioned. If auditory complaints were an uncommon subset of symptoms in veterans with MTBI, it would make sense that other concerns take precedent over audition. However, recent evidence suggests that auditory complaints are, in fact, very common. In a chart review of veterans diagnosed with MTBI, Oleksiak and colleagues (2012) showed that 89% of MTBI resulting from blast resulted in auditory sequelae. Data from a recent study completed by the authors were consistent with this high estimate in civilians with MTBI (85%; Hoover, Souza, & Gallun, 2014). Following existing clinical practice guidelines would result in a failure to address the hearing difficulties of many people with MTBI. However, many veterans have access to audiological care because auditory deficits are the most common service-connected disability and the Veterans Health Administration has greater resources devoted to hearing healthcare than civilian health systems. It is likely that clinical protocols in civilian clinics refer a smaller proportion of MTBI patients to audiology, and these patients.

Auditory Complaints After MTBI

The effects of MTBI on audition extend from the outer ear to the central auditory system. The literature on this topic includes numerous case studies and experiments that have been reviewed recently (Myers et al., 2008; Schairer, 2012; Vander Werff, 2012). Not included in these reviews are several recent studies of MTBI in veterans and non-veterans (Gallun, Diedesch, et al., 2012; Hoover et al., 2014; Oleksiak et al., 2012). These studies provide evidence that is consistent with physiological damage to the auditory system in cases of blast-related and impact MTBI, even in the absence of elevated pure-tone thresholds.

Although few studies have shown evidence of elevated pure-tone thresholds after MTBI in the absence of temporal bone fracture, it is important to consider the possibility of a combination of central and peripheral dysfunction. Cochlear hearing loss is common after blast injury (Myers, et al., 2009), and in veterans who likely had comorbidities related to excessive noise exposure (Gallun, Lewis, et al., 2012; Oleksiak et al., 2012). It has been suggested that elevated pure-tone thresholds after MTBI may be missed due to a small, 10–15 dB shift that remains within normal

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limits (Myers et al., 2008). Suprathreshold tests of cochlear function, including acoustic reflexes and otoacoustic emissions, have shown long-term changes in some listeners after MTBI (reviewed in Myers et al., 2009). Even in cases that present with no change in pure-tone thresholds, suprathreshold deficits in peripheral encoding of sound, cochlear innervation, and strial health may contribute to auditory dysfunction.

Evidence from recent studies confirms the notion that MTBI causes physiological damage in the brainstem and auditory cortex. Oleksiak and colleagues (2012) found abnormal acoustic reflex thresholds in 18% of veterans with MTBI, consistent with previous reports of changes to acoustic reflexes and suppression of otoacoustic emissions (Ceranic, Prasher, Raglan, & Luxon, 1998; Nölle, Todt, Seidl, & Ernst, 2004). These results provide physiological evidence of impairment in the structures of the cochlea or lower brainstem. Further evidence of physiological dysfunction at higher centers in the auditory system was found in the decreased amplitude of P300 response in blast-injured veterans (Gallun, Diedesch, et al., 2012).

Behavioral data consistently show evidence of auditory dysfunction. Eighty to 100% of listeners report subjective hearing difficulty after MTBI (Hoover et al., 2014; Oleksiak et al., 2012), with a higher proportion reporting difficulty when the cause of the injury was blast exposure. Performance on tasks of speech understanding in noise, including tests for central auditory processing disorder (CAPD), were more likely to be abnormal than healthy controls (Gallun, Diedesch, et al., 2012; Hoover et al., 2014). Tests that were most likely to be abnormal in blast-exposed or MTBI listeners were the Words-in-Noise test (WIN; Wilson & Burks, 2005), Staggered Spondaic Words (Katz, 1998), and the Speech Spatial Release test (SSR; Gallun, Diedesch, Kampel, & Jakien, 2013). Of the small proportion of veterans with MTBI evaluated clinically for CAPD (16%), 100% were found to have abnormal results (Oleksiak et al., 2012). This was systematically tested by Gallun, Diedesch, and colleagues (2012), and blast-injured listeners were more likely to be abnormal on CAPD tests than healthy controls, with abnormal results reported in 3–39% of cases on the battery of tests included in the study and a greater proportion of 2 or more abnormal test results for individuals in the blast-injured group than controls. Many of the listeners in the study were never diagnosed with TBI and a diagnosis was not predictive of abnormal results. These results are consistent with previous studies that showed a higher prevalence of abnormal results on CAPD tests among listeners with MTBI relative to controls (Turgeon, Champoux, Lepore, Leclerc, & Ellemberg, 2011; Vander Werff, 2012). Behavioral test of central auditory function are consistent with auditory impairment, but the mechanism of the impairment remains controversial.

Possible Explanations of Auditory Dysfunction After MTBI

From the perspective of audiology, there is evidence that abnormal auditory test results after MTBI are due to physiological damage to the auditory system (discussed below). However, there are two alternate hypotheses: (a) that the abnormal results are associated with PCS and share an underlying cause of PTSD, and (b) that subtle cognitive deficits exist after MTBI that affect performance on challenging auditory tasks. The standard theory in neuropsychology is that PTSD is responsible for most or all symptoms that persist after MTBI, and cognitive-behavioral therapy should be used to target the impairment. Considering the similarities between cognitive-behavioral therapy and aural rehabilitation, this approach may provide positive results in many cases. However, even if auditory dysfunction were related to PTSD, there remains a potential benefit of audiological evaluation and treatment. The most qualified person to provide therapy targeting hearing difficulties is the audiologist. There is a growing body of evidence that neurosensory deficits commonly found after MTBI represent a different pattern from other PCS symptoms (reviewed in Hoffer et al., 2013). Auditory dysfunction, as well as visual, vestibular, and somatosensory symptoms, may have a separate underlying cause than the symptoms like headache and sleep disturbance associated with PTSD.

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The second non-auditory explanation for auditory complaints after MTBI and abnormal auditory test results is that they are due to domain-general cognitive deficits that affect performance on difficult auditory tasks. The standard battery of neuropsychological tests shows recovery within two months of MTBI, but studies have shown subtle deficits in working memory, attention, and processing speed that persist beyond the acute stage (e.g., Lux, 2007). It is likely that cognitive deficits resulting from more severe TBI affect auditory function.

However, it remains unclear whether the relatively minor cognitive effects of MTBI are sufficient to explain the common complaint of difficulty understanding speech and the behavioral results that have been found in numerous hearing studies. It is possible that CAPD and speech understanding in noise tests are highly sensitive to cognitive deficits, and that these tests are capable of revealing subtle long-term cognitive effects of MTBI. In order to test this relationship, cooperation between neuropsychology and audiology is required so that valid behavioral data can be collected on cognitive abilities and hearing abilities in the same individuals.

The auditory hypothesis is that auditory tests are abnormal because of physiological damage to the auditory system resulting from MTBI. This is supported by the growing body of evidence on neurosensory deficits after MTBI across other modalities (e.g., Lew et al., 2009). Physiological damage to the auditory sensory system resulting from MTBI is a likely culprit. Behavioral and physiological data are consistent with the mechanisms of damage in MTBI. Like more severe TBI, MTBI results in diffuse axonal damage, deafferentation, and neuronal loss (Gennarelli et al., 1982). The effect of such damage is decreased precision in the encoding of sound in the auditory system.

Degraded temporal precision in the auditory system is also consistent with the abnormal behavioral data after MTBI and blast-related injuries. Precise encoding of timing information is needed for speech tasks used to evaluate speech understanding in noise and CAPD. In speech, the use of the temporal fine structure helps the listener separate a single talker from other competing talkers (e.g., Drennan, Won, Dasika, & Rubinstein, 2007). Impaired performance on competing speech tasks such as the QuickSIN, the WIN test, and CAPD tests such as Staggered Spondaic Words and Dichotic Digits is consistent with degraded temporal precision in the auditory system. Gallun, Diedesch, and colleagues (2012) showed that listeners were more likely to be abnormal in these speech tests than controls not exposed to a blast (2012). These tests were also more likely to be abnormal in listeners with a history of sports-related MTBI (Turgeon et al., 2011). Recent evidence suggests that abnormal speech understanding in noise may be related specifically to deficits encoding temporal information with the high precision required for the use of temporal fine structure cues in speech and psychophysical tasks (Hoover et al., 2014). Data from numerous studies are consistent with degraded temporal processing in the auditory system after MTBI, and the mechanisms of injury in MTBI are consistent with degraded temporal precision in the auditory system.

Evidence of Auditory Dysfunction in the Absence of Cognitive Factors

In a recent study, we evaluated the hearing and cognitive function of people with long-term auditory complaints after MTBI (Hoover et al., 2014; Hoover, 2015). In each case, the listener with MTBI had more than one year pass since their most recent head injury. The listeners were asked questions about their hearing ability, and their responses to these questions were compared to objective speech understanding measures. A complete audiometric evaluation was performed, including speech understanding in quiet and in multi-talker noise. Using this data, we were able to determine whether listeners with auditory complaints actually performed worse than age- and audiogram-matched controls on objective speech tasks. To determine whether their impaired speech understanding was a consequence of domain-general cognitive deficits, listeners were evaluated using tests recommended in the National Institute of Health Common Data Elements Toolbox as most likely to be impaired after acute MTBI including attention, memory, and executive function (Maas et al., 2010). Finally, auditory psychophysics tasks including monaural and

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binaural temporal fine structure and frequency selectivity were completed to test the hypothesis that auditory dysfunction after MTBI is related to impaired temporal precision in the auditory system. Individual cases from this study revealed interesting insights into the potential role of audiology in the MTBI rehabilitation.

Figure 1 shows the pure-tone thresholds of a woman age 56 years who started experiencing hearing difficulties after a motor vehicle accident in 2003 that resulted in a diagnosis of MTBI. At the time she experienced a brief period of loss of consciousness and post-traumatic amnesia. Following the event she reported chronic joint and muscle pain, severe headaches, intermittent dizziness and nausea, tinnitus, difficulty understanding speech in noise, and impaired memory. The list of symptoms she reported were consistent with PCS, and she received evaluations from neurology and neuropsychology including multiple imaging and behavioral assessments. Included in the assessment were tests of memory, language, and other cognitive functions. Short-term memory deficits found in the acute stage recovered spontaneously within weeks of the incident, but she felt that the test results were not consistent with her perceived memory impairment. She was not referred to audiology and had never received a hearing evaluation prior to participating in our study. Her audiogram shows pure-tone thresholds within normal limits from 250 Hz through 8000 Hz in the left ear and in the right ear through 4000 Hz sloping to mild sensorineural hearing loss at 8000 Hz. There is a significant asymmetry in air-conduction thresholds of 15 dB at 6000 Hz, and 20 dB at 8000 Hz. At 250 Hz, a gap between air- and bone-conduction thresholds are consistent with a borderline conductive hearing loss in both ears.

Figure 1. Audiogram for a Case of Auditory Dysfunction After MTBI.
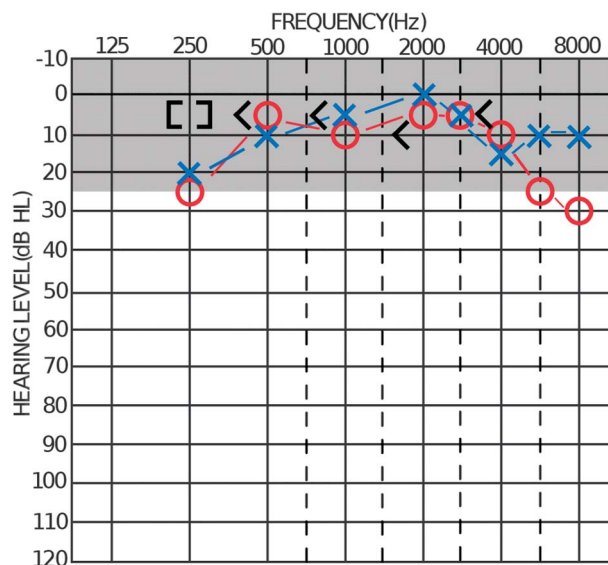
When asked if she had difficulty understanding speech in quiet, the patient answered, "no," but reported "yes," when asked if she had difficulty understanding speech in a noisy environment, a finding consistent with 85% of listeners with a history of MTBI (Hoover, 2015). Her speech understanding scores are shown in Table 1. In agreement with her self-assessment, speech understanding in noise scores were consistent with hearing impairment. These tests included QuickSIN (Killion, Niquette, Gudmundsen, Revit, & Banerjee, 2004), WIN (Wilson & Burks, 2005), and SSR (Gallun et al., 2013). Results of each of these tests were abnormal, indicating that she requires a greater signal to noise ratio than a typical listener with normal pure-tone thresholds and that her ability to benefit from a spatial separation of talkers is reduced. Her word recognition

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in quiet and QuickSIN scores are asymmetric, and the asymmetry is consistent with her audiogram with better performance in the right ear than the left. Given her age and pure-tone thresholds, her scores on the speech tests are worse than expected for someone with no retrocochlear lesion. In particular the WIN and SSR scores reveal substantial impairment, which may be due to peripheral or central auditory deficits. Furthermore, without audiometric data from before the MTBI it is impossible to definitively say that the auditory impairment was related to the injury.

Table 1. Audiometry and Speech Summary Data.

Ear	PTA	SRT	WR	QuickSIN	WIN	SSR
Right	7 dB HL	10 dB HL	88%	2 dB	9.2 dB	2 dB
Left	5 dB HL	5 dB HL	100%	4 dB	10 dB	

Summary data from audiometry and speech tests for a listener with a history of MTBI. Speech understanding in noise shows greater impairment than would be expected given the listener's age and pure-tone thresholds, but an asymmetry in word recognition and QuickSIN scores suggests peripheral as well as central impairment. Results of psychophysical tasks were consistent with the hypothesis that the difficulty this listener reported were related to degraded temporal precision in the auditory system, but, like the speech data, a contribution of cochlear impairment could not be ruled out. Her thresholds were impaired relative to age- and audiogram-matched controls on tests of auditory temporal processing, consistent with degraded temporal processing, and spectral resolution. Impaired spectral resolution suggests cochlear hearing loss. One possible explanation for these results is that cognitive factors contributed to her poor performance on all the tests. However, her scores were within normal limits for all of the tests in the cognitive battery. This suggests that cognition was not a contributing factor in her difficulty in the speech tasks and the psychophysical tasks, and her auditory symptoms reflect impaired auditory physiology.

This case demonstrates the general difficulty in evaluating auditory dysfunction in listeners with MTBI. In an individual listener, it can be difficult to account for all of the factors that can contribute to perceived and objective impairment in a way that is satisfactorily explained by any single measure. That is why a battery of tests are used, and, as this case demonstrates, why hearing tests should be included in a battery of tests for long-term PCS symptoms.

Conclusion

Although there is general agreement within the field of audiology that auditory dysfunction after MTBI is related to deficits in auditory physiology, it is important to remember that this opinion is not widely held among other professionals who work with the MTBI population. Growing evidence of related neurosensory deficits provide support for the auditory hypothesis, but more research is needed to identify the contributions of PTSD, domain-general cognitive deficits, and auditory physiology in people with auditory complaints after MTBI. Furthermore, clinical research is needed to determine whether patients with PCS have their rehabilitation improved with the addition of audiological evaluation and treatment, or if the existing model is the most efficient use of healthcare resources.

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King-Kopetzky Syndrome? A Bio-Psychosocial Approach to Adult "APD"

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Abstract

This paper discusses the relationship between King-Kopetzky syndrome and auditory processing disorder (APD). King-Kopetzky syndrome is the term applied to adults who present for help in the presence of normal audiometric thresholds. These adults may or may not have an APD and this paper discusses the factors that influence help-seeking and what is known about audiological diagnosis and management.

Mr Smith has noticed increasing difficulty hearing in group situations. He is concerned that his hearing may be getting worse and seeks help from his doctor. The doctor refers him to an otolaryngologist for a medical inspection. No obvious cause or site of hearing disorder is apparent. Mr. Smith presents with normal audiometric thresholds. The otolaryngologist refers him on to the audiologist for further help.

Why do some people complain of hearing difficulties when their hearing thresholds are normal? Adults presenting with disabling hearing difficulties (despite apparently normal hearing thresholds) have been the subject of much research interest and clinical speculation over the years. This is a common occurrence accounting for an estimated 5–10% of all referrals to otolaryngology (Saunders & Haggard, 1989). These cases challenge audiologists to tolerate uncertainty about underpinning causes, best actions for investigation, and best treatment options.

This article will discuss how help seeking and symptom detection occur. It will identify the philosophical perspectives that underpin the research to date and examine what this information contributes to our clinical knowledge. It will consider the underlying factors that contribute to an individual seeking help for a hearing problem and what evidence we have about what really helps.

The most commonly applied clinical label for this experience of hearing difficulties with normal audiometric thresholds is auditory processing disorder (APD), but it has also been referred to as King-Kopetzky syndrome. King Kopetzky syndrome refers to a wide heterogeneity in presentations, symptoms, and underpinning factors. It refers exclusively to adult presentations. Some individuals who present for help may have an auditory impairment but not necessarily (Pryce, Metcalfe, Hall, & Claire, 2010).

Is There a Difference Between King-Kopetzky Syndrome and Auditory Processing Disorder?

There remains ambiguity about whether a disorder identified in children is the root cause of adult presentations with hearing difficulty. Simply because both groups might present for help with normal audiometric thresholds does not mean that the underlying basis can be assumed to be the same. The research literature is dominated by investigations of auditory processing in children, which is then considered to apply to adults as well (American Academy of Audiology [AAA], 2010; Moore, Rosen, Bamiou, Campbell, & Sirimanna, 2013). Studies focussed on adults only are often overlooked (not least because the APD literature does not reference the King-Kopetzky syndrome literature).The literature that deals exclusively with adult presentations is found under the term King-Kopetzky syndrome or obscure auditory dysfunction (OAD).

The bio-medical perspective is probably the dominant view held by both clinicians and patients alike (Wade, 2006). If someone is complaining of hearing difficulties, the most likely starting point for the health practitioner is to consider the possible biological causes of such difficulties and check all auditory functions (both peripheral and central) that could have become impaired. Therefore APD (AAA, 2010) is the most frequently used clinical label for such patients in audiology in the United States and UK. But is this label adequate? In such cases, adult patients frequently describe their childhood experience of hearing as normal, without any schooling or other difficulties, though there may be more complicated social factors which influence their decision to check their hearing.

There are implicit philosophical differences in perceived versus objectively observed disorders. It is not inevitable that all presentations with hearing difficulties must be dependent on processing impairment. This corresponds with a dualist separation of mind and body and a belief in the objective presence of an underlying pathology in all cases.

Yet this position is not consistent with what we know about help-seeking behaviours, symptoms perceptions, and the way individuals evaluate their health. Dualism is not a helpful model as mind/body interactions are demonstrated in most aspects of health and well-being (Balint, 1957). People seek help as a result of multiple factors including impairments in peripheral and central auditory pathways but also as a result of psychological and social factors (Cramer, 1999; Lazarus & Folkman, 1984; Moller-Leimkuhler, 2002; Shaw, 2001).

A "syndrome" is defined a set of symptoms that occur together and characterise a particular abnormality in function (Oxford Dictionaries, 2015). This terminology does not prejudge symptoms as dependent on impairment in processing. For example, a change of lifestyle to a new noisy and stressful listening environment may contribute to someone's decision to seek help (Pryce, 2003). It may be that their function is not innately impaired but the stress of managing communication in a difficult listening environment has led to concerns about their hearing (Borg & Stephens, 2003). In these cases a diagnosis as having an inherent APD may carry unintended consequences and even harms. There are increasingly recognised risks of pathologising and over-diagnosing health conditions which may be subject to social and environmental factors (Moynihan et al., 2013). Audiologists, in keeping with most other healthcare professionals are under increasing pressure to be mindful of the risk of over diagnosis (Moynihan et al., 2013). Clinicians are challenged to identify and tease out the scientific evidence from amongst professional preferences and patient desires. As in most areas of audiology it is difficult to see a consensus on one single way to proceed.

Arguably, for adults, it is entirely normal to experience changes in our auditory perception over time (as we age, for example). Our colleagues in medical sociology and health psychology might regard these biological changes as less relevant to clinical presentation. Medical sociologists might consider this as pursuing a socially sanctioned "sick role" (Parsons 1964) which relieves our patient, Mr. Smith, of social responsibility, elicits sympathy, and justifies communication

problems. A health psychologist might consider Mr. Smith as responding to a series of cognitions about his hearing performance. He is evaluating it according to individually held beliefs about normal and impaired hearing performance and the consequence of these (Leventhal, Nerenz, & Purse, 1984).

In summary our knowledge about our own health, including our hearing performance, is relative and varies according to our lay beliefs and expectations. We may seek help on the basis of our perceptions of illness and normality, rather than on the basis of objective signs alone (Lazarus & Folkman, 1984). As Figure 1 illustrates the clinical label King-Kopetzky syndrome encompasses several causal factors.

Figure 1. An Illustration of the Relationship Between King-Kopetzky Syndrome and Causal Factors.
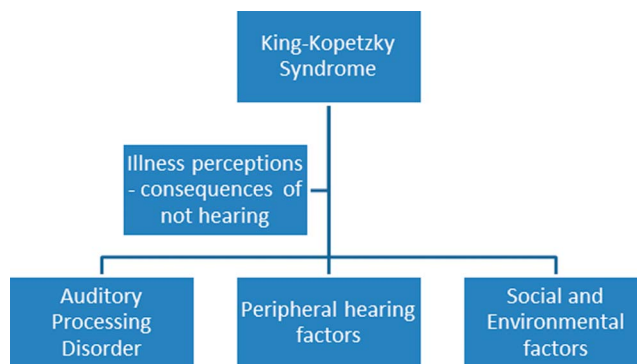
The World Health Organization (WHO, 2001) proposes an internationally agreed framework in which biological, psychological, and social factors are deeply interwoven in our experience of health and wellness. The risk of adopting a narrow, disease-led model of understanding this type of hearing problem is that we fail to recognize the human needs of the patient seeking help. We are in danger of over simplifying a complex problem into a purely biological cause.

King-Kopetzky syndrome is the proposed term for adults who seek help in the presence of normal audiometric thresholds. They may or may not have an APD. This definition recognizes that for some people the problem is due to anxiety and perception of performance. Zhao and Stephens (2000) identify seven sub-categories of King-Kopetzky syndrome:

1. Middle ear dysfunction

2. Mild cochlear pathology

3. Central/medial olivocochlear efferent system dysfunction

4. Purely psychological problem

5. Multiple auditory pathologies

6. Combined auditory dysfunction and psychological problems

7. Unknown

This heterogeneous model of difficulty has in recent years been adopted by the definitions of APD (BSA, 2011). The British Society of Audiology (BSA, 2011) Special Interest Group in APD now consider acquired APD in adults and children to exist in the presence of, or as a result of peripheral hearing loss, including transient and treated hearing loss; certainly the role of memory and attention is now considered important in "developmental" (or acquired) APD (Moore et al, 2013). Memory, attention and cognitive function can be compromised by numerous factors, including stress (Kim & Diamond). King-Kopetzky syndrome pre dates this revised interpretation of

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auditory processing disorder and none of the literature is referenced in the position statements on APD (e.g., AAA, 2010; BSA, 2011). This impacts the application of guideline to adult cases. There has been relatively little research into the effects of these hearing difficulties (Pryce, 2003; King & Stephens, 1992) or their treatment (Zhao, Stephens, Pryce, Zheng, & Bahgat, 2008).

The Paradox of Help-Seeking

Why do these patients seek help when so many with measurable and disabling hearing losses do not? One of the main curiosities in this area is that these patients seek help with relatively small changes in function. If we accept current estimates, then on average someone will experience hearing difficulties which mask participation in conversation and limit their function for ten to twenty years before they seek help with it (Carson 2000; Getty & Hétu, 1994; Kyle, Jones, & Wood, 1985; van den Brink, Wit, Kempen, & van Heuvelen, 1996; Watson & Crowther, 1989). We hear much of the stigma of hearing loss and the barriers to help-seeking (Wallhagen, 2010). Regardless of where a lesion might actually lie, the disorder (if indeed one can be identified) does not accurately predict the perceived limit to function and participation. The answer appears to come from models of illness perception, well-understood in health psychology. In order to identify ill health, people are thought to compare their symptoms with prototypical symptom sets (Bishop & Converse, 1986). Illness perceptions also guide cognitive appraisals of the symptom sets (Leventhal 1990). In other words a person compares their bodily "signs," in this case, how well they appear to hear in a particular environment with their expectation of how well they think they should hear based on their perception of those around them, what they consider to be "normal" for the environment and noise level, people their age and in their position etc. So those who seek help with hearing symptoms do so on the basis of internally held schema about what is "normal".

Where Does "King-Kopetzky" Come From?

Samuel Kopetzky described the problems of hearing in radio operators returning from the Second World War. He conceptualized these cases as having "loss of capacity for discriminative listening" (Kopetzky 1948; Saunders & Haggard, 1989). These individuals had spent many hours endeavouring to decipher enemy signals over radio. They later presented with concerns that their hearing was affected. Kopetzky notes that there was considerable anxiety around this evaluation of their hearing. Air Vice Marshal Peter King (1954) developed this work with a fuller description of possible causes in 1954. The term King-Kopetzky syndrome was coined by Ron Hinchcliffe in 1992. Hinchcliffe reviewed what was known about such hearing difficulties and concluded that these were manifestations of "auditory stress disorder". He includes the following early description, "He was a worried, tense man extremely anxious lest his defect should be the cause of a disaster, particularly when told that his hearing was normal" (Hinchcliffe, 1992).

The Psychological Dimension

One of the difficulties in interpreting APD data is that help-seekers and non help-seekers are rarely compared. Help-seeking is key to the clinical categorisation and diagnosis. Therefore, it is important to consider a comparison between those who do and do not seek help with the same symptoms.

Investigations of King-Kopetzky syndrome have included psychological appraisal. Stephens et al. (2003) identified that self-rated participation restriction varied with cognitive appraisal. Likewise cognitive appraisal was important in describing the maintenance of hearing difficulties (Pryce, 2003). The Crown-Crisp Experiential Index (Crown & Crisp, 1956) has been used to examine psychoneurotic traits in those with hearing difficulties and normal audiometry and those with chronic pain. This index comprises items measuring levels of anxiety, phobic anxiety, obsessionality, somatic anxiety, depression, and hysteria. When compared with control subjects without any reported pathology, more phobic and free floating anxiety were noted in those with hearing difficulties. When compared to chronic pelvic pain the psychoneurotic traits were similar

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but hearing performance (measured as speech in noise) was worse (Saunders & Haggard 1993). The examination of psychological traits has suggested that this group of patients experience higher rates of anxiety. The direction of causality between anxiety and hearing difficulty is debateable. To aid interpretation, qualitative work has modelled how anxiety is experienced and how it links to hearing difficulties. This suggests that anxiety has a role in maintaining awareness of symptoms by sensitising individuals to particular communication environments or partners (Pryce, 2003, 2006).

Patients describe in detail how the way they interpret their symptoms determines the likelihood of future communication breakdown (Pryce, 2006). The role of symptom appraisal was examined through a large scale multivariable analysis in which dichotic listening, speech-in-noise, and frequency resolution tests and illness perceptions were examined to identify which factors predicted help-seeking (Pryce et al., 2010). This study revealed that the factor that predicted help-seeking was the belief in the consequences of having a hearing difficulty. In other words help-seeking behaviour was better predicted by a psychological factor in the impact of the symptoms than any audiological function or impairment (Pryce et al., 2010).

Wider Context—How Common is a Psychological Component?

Medically unexplained illness in which psychological components co-exist with physical signs are common in healthcare (Pennebaker, 1982; Salmon, 2000). A review of neurological symptoms found that 26% were medically unexplained (Perkin, 1989).

This is not an "either/ or" situation. There are frequent examples in the literature where health conditions have a biological underpinning which is exacerbated by emotional response. In particular, anxiety has a role in starting and maintaining symptoms. This has been identified in abdominal pain, headaches, back ache, and benign palpitations (Mayou, 1991. In otolaryngology such amplification occurs in vertigo (Hallam & Stephens, 1985), globus pharyngis (Deary et al., 1989), and tinnitus (McKenna, Hallam, & Hinchcliffe, 1991).

In a small study detailing the patient perception of the way hearing difficulties in King-Kopetzky syndrome arose, emotional distress was highlighted as a factor in both the start and maintenance of symptoms. For some this was a clear incidence of trauma preceding the symptoms (i.e., a car accident, divorce, death of a loved one or collapse of a business; Pryce, 2003). Study participants revealed that such traumas were part of their own personal history and interpretation of their symptoms. This study also highlighted that the anxiety experienced during communication would predispose the individual to further difficulty in communication. What is more this "vicious circle" could be reversed when the communication partner was deemed to be understanding of the communication difficulties, the difficulties themselves reduced (Pryce, 2003, 2006).

The presence of psychological components in the sensitivity to underlying auditory processing disorders mean that for the patient their day to day experience of hearing difficulties varies according to social and psychological factors. Hearing and communication are interactive processes occurring in an ecological context influenced by intra personal, interpersonal social, and cultural factors (Borg & Stephens, 2003).

So What Helps?

Coping in King-Kopetzky syndrome was examined in detailed qualitative work. This study explored the individual cognitive and behavioural attempts to manage stress cause by hearing symptoms (Pryce, 2006). The process of coping relied on conceptualising the symptoms and adopting strategies to manage day to day communication. The strategies identified were:

- Concentrating to piece together the communicative message
- Bluffing
- Avoiding communication

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- Asking for repetition

- Lip-reading

- Directing speakers to adjust their behaviour

- Changing the environment

- Disclosing their difficulty

- Humour

The strategies are similar to those adopted by people with other forms of hearing loss. What is interesting here is that the clinical encounter could prompt the use of strategies and reduce stress when employing strategies. This was in part because the clinical encounter assisted validation of the symptoms (Pryce & Wainwright, 2008). The characteristics of clinical encounters determined outcomes in either increasing or reducing anxiety about symptoms. In particular, encounters that were deemed to be dismissive were unhelpful and raised anxiety. This was typically the case when clinicians sought to provide brief reassurances that test results were normal. According to a participant reported in Pryce and Wainwright (2008), "they say your hearing is normal and they don't consider there's a problem. They're not interested".

One of the most helpful things to emerge from the BSA (2011) and AAA (2010) has been the recognition of APD as a worthwhile and legitimate presentation along with some guidance as to clinical action. It is hoped that such guidance will encourage and increase helpful and purposeful encounters for patients. However the audiologist should remain vigilant about the potential for testing to cause anxiety and to use test results to tailor further interventions to the individual. The most helpful part of the clinical encounter that patients reported was the sense of being understood and being given an explanation for the difficulties in hearing (Pryce & Wainwright, 2008). Participants reported this assisted both emotion focussed coping and practical uptake of strategies:

> I realized there were other people that were the same and that actually it wasn't a really negative thing about me. It was just me....I've got more courage now to say "I didn't hear you because" rather than saying nothing before.

(Participant T3 reported in Pryce & Wainwright 2008)

So What Sort of Assessment?

The BSA (2011) guidance for APD practice proposes a full case history be taken, supplemented with questionnaire measures. One of the challenges is that the questionnaire measures are developed for use with children and do not easily apply to adults. However, a full case history including information about the context in which difficulties occur can be very helpful. It is also useful to directly explore issues around stress in listening (Saunders & Haggard, 1989; Pryce, 2006). The AAA guidance for (C)APD suggests including an evaluation of:

- auditory and/or communication difficulties experienced by the individual

- family history of hearing loss and/or central auditory processing deficits

- medical history, including birth, otologic and neurologic history, general health history, and medications

- speech and language development and behaviors

- educational history and/or work history

- existence of any known comorbid conditions, including cognitive, intellectual, and/or medical disorders

- social development

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- linguistic and cultural background
- prior and/or current therapy for any cognitive, linguistic, or sensory disorder or disability (AAA, 2010).

The guidelines propose that further testing should be offered to those with a history of significant history of otitis media, neurological disease or disorder. Patients presenting with seizure disorders and hyperbilirubinemia should also be considered for testing (AAA, 2010).

Test packages such as the SCAN packages (Dawes & Bishop, 2007; Keith, 2000) are freely available to audiologists. These tests assess ability to detect speech-in-noise and attend to information for each ear individually (Pryce et al., 2010). However the reliability of tests is still debated as they are subject to learning effects (Domitz & Schow, 2000), and cannot discriminate between pre-cognitive and linguistic factors (Moore, Rosen, Bamiou, Campbell, & Sirimanna, 2013; Pryce et al., 2010).

These tests require a clear sense of purpose when applied in the clinic. Their application should be in an ethical context where information will be used to benefit an individual in a way that outweighs the associated risks of enhancing anxiety about performance and function further. One way to consider this is to focus efforts on validating and supporting patients to make their choices about hearing management. The act of validation and labelling the condition enhance coping by supporting illness coherence (Pryce, 2003, 2006; Pryce et al., 2010). It also has potential to inform both audiologist and patient and facilitate better informed choices about care and management.

Treatment

Borg and Stephens (2003) conceptualize the experience of King-Kopetzky syndrome as a disruption between an individual and their environment. Therefore it is important to examine how strategies can reduce this disruption. Patient behavioural change is required for most rehabilitation in hearing loss and King-Kopetzky syndrome is no different. In this case examining the environment and adapting it where possible is particularly important to improve speech-in-noise perception. Relatively simple strategies such as turning off background noise where possible can be extremely helpful (Zhao et al., 2008). There is limited evidence that auditory training may have an effect on day to day function (Bronus et al., 2011).

To examine complex interventions such a therapeutic interventions for King-Kopetzky syndrome fully the UK Medical Research Council suggests that we develop clear, theoretical understanding of the way interventions might work. In this case the theoretical work has been conducted through qualitative approaches, based on grounded theory. Through this approach there are clear descriptions of how clinical interventions do and do not assist coping as the patient perceives it. Above all, the most valuable intervention reported in interviews accounts about what aids coping is the act of being listened to and being taken seriously (Pryce & Wainwright 2008; Zhao et al., 2008). This is helpful in developing coping by supporting the use of strategies to manage difficult situations and to build confidence in encouraging speakers to slow down, move away from background noise, switch room etc. (Pryce, 2006; Pryce & Wainwright 2008). This apparently simple use of counselling and problem solving therapy is effective in other medically unexplained conditions such as fibromyalgia, chronic fatigue syndrome, etc. (Hassett & Gevirtz, 2009) so it is perhaps not so surprising this works in this instance.

There has been suggestion that auditory training packages might help individuals to function better by retraining their discrimination of sound. The theoretical basis for this is not completely clear and findings are mixed (Moore et al., 2013). The practice of repeatedly listening to sound contrasts is believed to drive the development of more efficient neuronal pathways, thereby improving auditory processing (Bronus et al., 2011). However there is a lack of neurophysiological studies to examine this action (Bronus et al., 2011). Moreover there is a

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dearth of studies demonstrating more than a modest effect size for the benefits of training on task (Bronus et al., 2011).

The potential for communication training programmes, which include an educational and behavioural component such as active communication education also deserve further investigation with this population. These programmes work on problem solving and application of strategies to manage difficult situations and have demonstrated effective improvement in communication function, hearing handicap, and well-being in adults with hearing loss (Hickson, Worrall, & Scarinci, 2007).

It is likely that such approaches will trigger coping adaptations which in turn can affect hearing performance. A study into the effects of mindfulness therapy identified improvements in coping and even reports of improvements in hearing performance suggesting that coping responses can moderate function (Sadlier & Stephens, 2009).

As with other hearing conditions, it makes sense to consider how clinicians can enact shared decision-making with this population, especially where treatment is dependent on coping skills and behavioural adaptations. Presenting patients with clear choices in interventions and outlining the advantages and disadvantages of different course of action will be important here (Pryce & Hall, 2014).

Conclusions

King-Kopetzky syndrome represents a fascinating experience of mis-hearing based on multiple factors, which vary in emphasis from individual to individual. The role of APD as a sub component to the presentation is interesting and more that is learned about this component will help with better patient counselling and guidance. Evidence from qualitative investigations suggests that the broader, non-biological factors that lead to clinical help-seeking should be considered alongside audiological factors in clinical history taking, counselling, and problem solving. This area of work highlights the complexity of the audiological role in addressing psychological and social needs of patients alongside the biological needs. It also highlights the needs for audiologists to engage in counselling and communication training approaches beyond the traditional technical investigations.

It reminds us that healthcare involves the toleration of uncertainties about causation and remediation. Our role is to support the whole person, including providing reassurance about the consequences of not hearing, arming them with practical strategies to repair conversations, informing them about their function and potential and, above all, listening and bearing witness to their experience.

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The Relation Between Cochlear Neuropathy, Hidden Hearing Loss and Obscure Auditory Dysfunction

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Abstract

Sensorineural hearing loss is typically defined by threshold elevation, often resulting from dysfunction of cochlear outer hair cells. Damage can occur as a result of ageing and noise exposure, reducing auditory sensitivity, and allowing detection using pure tone audiometry, the cornerstone of audiological testing. Yet the audiogram is limited in its ability to predict perceptual performance, especially for speech perception in noise. The deficits reported in some audiometrically normal listeners provide stark examples of this insensitivity. Recent research in animals has revealed that widespread loss of auditory nerve fibers can take place without hair cell loss or threshold elevation, as a result of either noise exposure or ageing. This pattern of damage may manifest as "hidden hearing loss" in humans, leading to perceptual deficits without affecting the audiogram. A great deal of interest is currently focused on this condition and its putative effects, which are expected to impact listening in complex auditory environments. Much work remains to be done in establishing the existence, causes, effects, and clinical relevance of hidden hearing loss in humans. However, forthcoming research findings are likely to be pivotal and may demand that we reassess our diagnostic methods, management strategies, and fundamental understanding of hearing loss.

Some individuals report significant difficulties understanding speech in noisy environments despite having normal audiometric thresholds. This clinical presentation is commonly denoted "obscure auditory dysfunction" (OAD; Saunders & Haggard, 1989), though some consider "auditory processing disorder" (APD), an apt and preferable term (British Society of Audiology APD Special Interest Group, 2011). This article will use the term OAD, which avoids mechanistic assumptions and specifies listening difficulties in noise, though implications for APD will be briefly considered.

Amongst patients referred to audiology services for listening difficulties, clinically normal audiograms occur in an estimated 2 to 8% of patients (Hind et al., 2011; Stephens, Zhao, & Kennedy, 2003), though clinical data may underestimate population prevalence. A UK study of

~2500 individuals found that 26% of adults reported great difficulty hearing speech in noise, whilst only 16% demonstrated clinically abnormal audiometric sensitivity (Davis, 1989). A variety of factors are thought to motivate clinical presentations of OAD, including psychological, central, cognitive, and sensory factors (Saunders & Haggard, 1992; Zhao & Stephens, 2007), and knowledge of underlying pathologies and their relation to one another remains incomplete. However, as will become clear, recent scientific findings from both human and animal studies suggest that damage to the auditory nerve (AN) could underlie some cases of OAD.

Animal Evidence of Cochlear Neuropathy

The classic view of sensorineural hearing loss holds that damage occurs primarily to the hair cells of the cochlea, resulting chiefly from the ageing process and from exposure to loud sounds. Cochlear outer hair cells (OHCs), which amplify the motion of the basilar membrane, are thought to be particularly vulnerable. Loss of OHC function reduces the effectiveness of the cochlear amplifier, resulting in decreased sensitivity to quiet sounds and hence to elevated audiometric thresholds. However, a very different type of hearing loss was recently shown to be dominant in seminal work on noise exposure using a mouse model.

Kujawa and Liberman (2009) exposed mice to 2 hours of 100 dB SPL noise (8–16 kHz), which resulted in only temporary threshold shifts. Otoacoustic emissions (OAEs) and auditory brainstem responses (ABRs) at low sound levels returned to normal several weeks post-exposure, implying no permanent effects on OHC function. However, ABR wave I amplitudes, which reflect AN activity, were permanently reduced in response to medium and high level sounds, and histological investigations revealed that up to half of the synapses between inner hair cells (IHCs) and AN fibers had been lost in high frequency regions. This synaptic loss is thought to have resulted from glutamate excitotoxicity (Pujol & Puel, 1999) and was followed by gradual degeneration of AN fibers, despite the survival of cochlear hair cells. Termed "cochlear neuropathy," this pattern of damage represents a new variety of sensorineural hearing loss, distinct from hair cell loss and not necessarily reflected by permanent threshold elevation.

The findings of widespread neuronal loss and normal cochlear sensitivity seem contradictory, yet may be explained by a specific pattern of neural damage. IHCs synapse with many AN fibers, which have a range of spontaneous firing rates. A subsequent study by Furman, Kujawa, and Liberman (2013) demonstrated noise-induced cochlear neuropathy in another species, the guinea pig, and found that neuropathy was selective for fibers with low and medium spontaneous rates. These fibers have relatively high response thresholds and contribute little to the detection of low-level sounds, but may be important in the coding of auditory inputs at higher levels (Bharadwaj, Verhulst, Shaheen, Liberman, & Shinn-Cunningham, 2014).

Maison, Usubuchi, and Liberman (2013) showed that cochlear neuropathy in mice can result from exposure to more moderate sound levels (84 dB SPL) over longer durations and is exacerbated by loss of medial olivocochlear feedback. Similar effects have been found as a result of ageing, in animals not exposed to high sound levels. Microscopic examination of the inner ears of mice by Sergeyenko, Lall, Liberman, and Kujawa (2013) revealed a progressive loss of cochlear synapses throughout the lifespan, well in advance of threshold changes or OHC loss, with neuronal loss following. Previous AN recordings from ageing gerbils indicate declining proportions of fibers with low spontaneous rates (Schmiedt, Mills, & Boettcher, 1996). Taken together, these findings suggest that noise-induced and age-related cochlear neuropathy may share the same underlying mechanisms and effects, with noise exposure hastening the onset and progression of neuronal loss that occurs naturally as part of the ageing process.

Human Evidence Consistent With Cochlear Neuropathy

The term "hidden hearing loss" (HHL), coined by Schaette and McAlpine (2011), is increasingly prominent in literature relating to cochlear neuropathy. The term's usage has sometimes been

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inexact, referring to suprathreshold deficits of uncertain cause. Contrary to this practice, we define HHL as cochlear neuropathy that is undetectable by pure tone audiometry and that leads to perceptual effects. If those perceptual effects include impaired speech perception in noise, as proposed by Kujawa and Liberman (2009), then HHL may explain some cases of OAD.

Relevant research in humans is in its infancy and is yet to yield conclusive evidence of noise-induced or age-related HHL. However, a variety of indirect evidence has accumulated. As we shall see, noise exposure and ageing have been linked to a range of perceptual and electrophysiological abnormalities, which are consistent with HHL and its expected effects on auditory processing. Kujawa and Liberman (2009) proposed that cochlear neuropathy should degrade the neural encoding of sound in low signal-to-noise ratio conditions. Lopez-Poveda and Barrios (2013) used a computer model to investigate the effects of deafferentation on the neural representation of speech. According to their simulation, diffuse neuronal loss leads to a "noisier" representation of the signal in the AN, which impairs speech perception in noise more than in quiet. Bharadwaj et al. (2014) proposed that cochlear neuropathy should impair the temporal representation of auditory inputs. In particular, they suggested that the selective loss of high threshold AN fibers degrades the encoding of the slowly varying temporal envelope, especially at high overall input levels.

Multiple studies demonstrate associations between noise exposure and suprathreshold perceptual deficits in individuals with normal or near-normal audiograms (see Plack, Barker, & Prendergast, 2014 for a review). Many of the findings are consistent with an underlying noise-induced HHL but as yet offer no conclusive evidence. For example, neuroplastic changes at higher levels of the central auditory system may play a role in the deleterious effects of noise exposure (Gourevitch, Edeline, Occelli, & Eggermont, 2014). It is important to note that Stephens et al. (2003) found no association between OAD and history of noise exposure. However, this cannot be taken as evidence that HHL is not a cause of OAD, since both OAD and cochlear neuropathy are likely to be multifactorial conditions.

Clearer substantiation of HHL requires objective and non-invasive measures of neuronal function in order to detect alterations in processing and establish the relation to underlying neuropathy. Promising electrophysiological techniques include the ABR at suprathreshold levels, as used in animal studies of cochlear neuropathy, and the frequency-following response (FFR)—a scalp-recorded potential reflecting phase-locked neural activity in the rostral brainstem (Krishnan, 2006). The FFR, at least at low frequencies, is not thought to reflect AN activity directly, but provides a useful measure of temporal precision in the brainstem and is likely to be sensitive to coding deficits earlier in the auditory pathway, including those due to cochlear neuropathy. Preliminary data suggest that the FFR may be sensitive to alterations in neural coding related to noise-exposure (Barker, Hopkins, Baker, & Plack, 2014).

The earliest interpretation of human electrophysiological data in relation to HHL involved individuals with and without tinnitus (the perception of a sound in the absence of an acoustic source). Schaette and McAlpine (2011) measured ABRs to high-level stimuli in two audiometrically matched groups of normal-hearing individuals, with one group containing individuals who reported tinnitus. They found that the tinnitus group had reduced wave I amplitudes at high levels, indicating reduced AN activity. This finding parallels the diminished wave I amplitudes seen in the animal model of noise-induced cochlear neuropathy and suggests that deafferentation may account for some cases of tinnitus. It is tempting to speculate that noise exposure might have played a causal role, though no measures of noise exposure were reported. Amplitude reductions were not seen in later peaks of the ABR, which the authors interpreted as evidence for compensation at the level of the auditory brainstem. This pattern of results, which was corroborated by Gu, Herrmann, Levine, and Melcher (2012), suggests not only that reduced AN activity can trigger tinnitus but that it does so by means of a "neural gain" mechanism. Such a compensatory mechanism may mitigate the extent to which an altered AN response affects perception, but the interaction of central gain mechanisms and cochlear neuropathy requires more thorough investigation.

Electrophysiological findings, possibly reflecting cochlear neuropathy, have also been related to performance on complex discrimination tasks. FFR synchronization strength to speech stimuli in audiometrically normal listeners has been shown to relate to speech recognition in noise (Song, Skoe, Banai, & Kraus, 2011) and to performance on an auditory selective attention task involving competing voices and spatial cues (Ruggles, Bharadwaj, & Shinn-Cunningham, 2011). The FFR is also predictive of performance on simpler discrimination tasks (e.g., pure tone frequency discrimination) even when audiometric threshold and age have been controlled (Marmel et al., 2013). More recently, Bharadwaj, Masud, Verhulst, Mehraei, and Shinn-Cunningham (2015) designed an FFR measure to be specifically sensitive to the hypothesized effects of cochlear neuropathy. They used stimuli with high sound levels and shallow modulations to challenge temporal coding and emphasize the contribution of high-threshold fibers. This electrophysiological measure correlated with behavioral measures of temporal coding and both types of measure were highly predictive of auditory selective attention. The authors concluded that young, audiometrically normal people vary markedly in their ability to analyze complex auditory scenes and that temporal coding deficits consistent with cochlear neuropathy may play a significant role in this variation.

A wide variety of data provides support for age-related HHL, including histological evidence. Makary, Shin, Kujawa, Liberman, and Merchant (2011) counted spiral ganglion cells (SGCs) in individuals with full sets of cochlear hair cells. SGC counts declined throughout the lifespan, preceding audiometric changes, and the rate of loss closely paralleled the rate found in mice when normalized by mean lifespan. The SGC loss may underestimate synaptic loss, since synaptopathy significantly preceded SGC loss in the mouse model (Sergeyenko, Lall, Liberman, & Kujawa, 2013). Though the overall pattern was of steady decline, some teenagers had lost over a quarter of their SGCs while some older individuals had very little loss. Though speculative, it is possible that these radically different rates of loss were attributable to differences in lifetime noise exposure. Such a study, which relates SGC counts to noise exposure, would aid our understanding of the causes of the cochlear neuropathy underlying HHL and may inform decisions relating to preventing and managing HHL.

Ageing is well known to be associated with deficits in auditory perception, many of which are consistent with the expected effects of HHL. Impaired speech perception in noise is frequently reported, even in the absence of threshold elevation (e.g., Dubno, Dirks, & Morgan 1984; Kim, Frisina, Mapes, Hickman, & Frisina, 2006). Declining cognition is thought to be a contributory factor, but degraded temporal processing is likely to play an additional role (Füllgrabe, Moore, & Stone, 2014). Age-related declines in temporal processing have been demonstrated independent of audiometric threshold, both in behavioral (Füllgrabe, Moore, & Stone, 2014; King, Hopkins, & Plack, 2014) and electrophysiological research using the FFR (Bones & Plack, 2015; Clinard & Tremblay, 2013; Marmel et al., 2013). However, Anderson, Parbery-Clark, White-Schwoch, and Kraus (2013) note that mechanisms other than AN deafferentation could account for altered temporal processing in older adults, such as a lack of inhibition or an increase in the variability of the neural response. Similarly, ABR amplitudes have been shown to diminish with age, largely independent of changes in the audiogram (Konrad-Martin et al., 2012). In particular, reductions were recorded in wave I, suggestive of cochlear neuropathy, though findings were also consistent with desynchronization of AN activity.

From Current Evidence to Clinical Significance

It has long been clear that pure tone audiometry is limited in its ability to predict everyday perceptual performance and to identify auditory pathology. Yet our understanding of the pathologies concealed beyond the audiogram remains unclear and incomplete. Cochlear neuropathy is a model of underlying pathology that has the capacity to explain some of the suprathreshold perceptual deficits reported in the literature, since it allows for widespread cochlear damage with normal audiometric findings. However, many questions must be answered if we are to establish HHL as a cause of hearing dysfunction in humans and to understand its contribution to OAD.

The animal evidence is compelling. In small mammals, cochlear neuropathy occurs as part of the natural ageing process and can also be induced by noise exposure, without OHC loss or permanent threshold elevation. However, it is by no means certain that comparable processes take place in the human ear, especially given the interspecies differences in the animal data. Studies that have induced cochlear neuropathy in guinea pigs rather than mice have reported much greater survival of SGCs (Lin, Furman, Kujawa, & Liberman, 2011) and widespread synaptic recovery (Liu et al., 2012). It may be that mammals differ widely in their susceptibility to cochlear neuronal loss. Additionally, animal research has largely neglected investigation of perceptual effects. These are required in order to substantiate HHL as we define it, and may play a key role in future HHL research by allowing the demonstration of parallel effects in animals and humans.

Current evidence supporting noise-induced HHL is indirect, mainly consisting of noise-related perceptual deficits in humans with normal or near normal hearing, which are generally consistent with HHL. Preliminary electrophysiological data has also associated noise exposure with temporal coding deficits and ongoing research is likely to corroborate or refute noise-induced HHL in the near future. As regards ageing, researchers have long puzzled over the declines observed in speech perception and temporal coding, which outpace changes in the audiogram. Age-related HHL is a plausible contributor to such suprathreshold deficits, particularly given the evidence of SGC loss in ageing humans and its correspondence to the animal model of cochlear neuropathy.

Clearly there is much work to be done in determining the actuality, causes and effects of HHL. Due to the constraints on invasive measures in humans, it may be most practical to devise paradigms that combine animal and human data to form a chain of evidence from histological findings all the way through to high-level perceptual effects. Electrophysiological measures will surely play a key role, in light of the proven sensitivity of such measures to cochlear neuropathy and the potential for parallel application in human and animal species.

Additional research will be required to establish a role for HHL in OAD, which is thought to reflect heterogeneous underlying factors. The effects of cochlear neuropathy on speech perception in noise must be shown not only to exist but also to be clinically significant. The existing evidence discussed above, relating neural temporal coding to speech discrimination, is derived from participants without known perceptual deficits. It may be that temporal coding is further degraded in some individuals with OAD, or it may be that this mechanism contributes little to their clinical presentation. There is a need for carefully designed research with this patient group, establishing the role of HHL relative to others factors such as cognition, personality, and subtle hair cell dysfunction.

Finally, it is important to note that the potential implications of HHL extend beyond hearing dysfunction with normal hearing thresholds. Cases of OAD and tinnitus with normal audiograms might represent particularly dramatic manifestations of HHL, but cochlear neuropathy may also be present in individuals with audiometric hearing loss. There are likely to be significant challenges in trying to determine the prevalence of HHL in clinically hearing impaired listeners and its contribution to their perceptual deficits. A complex mixture of elevated audiometric thresholds combined with a degree of cochlear neuropathy could explain why signal amplification does not fully restore speech perception in noise and why it is difficult to predict speech perception defects based on the audibility of the individual. There are potentially complex interactions at work; however, these questions should not be neglected and their answers may ultimately entail substantial revision of our understanding, assessment, and management of hearing loss.

Clinical Implications

Given the current state of HHL research, in-depth discussion of clinical implications is rather premature. Nevertheless, these ramifications deserve some consideration at this stage, given their potential importance for hearing health. If HHL is found to be a significant source of

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hearing dysfunction, then it must be remodeled as an auditory condition that can be detected, quantified, managed, and ideally prevented.

By definition, a "hidden" pathology demands new diagnostic techniques. Cochlear neuropathy eludes not only audiometry but also sensitive measures of auditory function such as OAEs and ABR thresholds (as currently the main clinical utility of an ABR recording is obtaining one at threshold). Current research into HHL aims not only to elucidate the condition but also to lay the groundwork for practical diagnostic tools. Particular promise may lie in applying electrophysiological measures such as the ABR and FFR at suprathreshold levels, allowing objective assessment of neural function early in the auditory pathway, free from the confounds of attention and cognition. It may be that the most sensitive and specific applications involve the comparison of information across different frequencies and sound levels, to draw out the effects of selective neuronal loss.

Arguably the most important implications of the existence of HHL concern noise exposure and hearing health protection. The potential dangers of high sound levels are recognized and addressed by evidence-based regulations on noise exposure in the workplace (United States Department of Labor, n.d.), as well as clinical recommendations and public health advice. Yet all rest on the assumption that exposures insufficient to cause permanent threshold shifts are not significantly damaging to long-term hearing health (May, 2000). Further research into HHL may show this premise to be unfounded. In particular, exposures considered moderate may in fact have the potential to cause permanent damage with disabling effects (Maison, Usubuchi, & Liberman, 2013), calling for the revision of guidance and legislation in order to best protect against hearing dysfunction. The diagnostic tools discussed above may play a role in this endeavor, by allowing early identification of susceptible individuals.

In the long term, a diagnosis of HHL may lead to direct treatment of neural damage. Advances in research with human and animal stem cells suggest that therapy to restore functional auditory neurons may one day be possible (Chen et al., 2012; Nayagam et al., 2013; Needham, Hyakumura, Gunewardene, Dottori, & Nayagam, 2014). In a similar vein, neurotrophins have been shown to promote regrowth of peripheral processes of auditory neurons and reduction of SGC loss (Glueckert et al., 2008; Wan et al., 2014).

In the more immediate future, insight into HHL should guide selection of management options for affected individuals and the development of new therapies. For cases of OAD attributable to HHL, the diagnosis of underlying pathology would indicate auditory rehabilitation, as opposed to (or in addition to) management of psychological or cognitive factors (Zhao & Stephens, 2007). Patients should be offered hearing and communication advice appropriate to AN dysfunction and consequent perceptual deficits. In addition, the identification of abnormal auditory processing might mean a place for appropriate auditory training (Moore, 2011), ideally developed to address coding deficits early in the auditory pathway. Lastly, technological advances might allow a role for hearing assistive technology. For example, "bilateral beamforming" hearing aids offer more enhanced directivity and greater improvements in signal-to-noise ratio (SNR) than conventional aids (Picou, Aspell, & Ricketts, 2014). These and other innovative signal processing strategies may be of benefit in offsetting the expected effects of HHL on neural SNR and temporal processing.

Finally, if HHL is found to underlie some cases of OAD, then clinical understanding of the condition will be aided by careful delineation of relevant terminology and of the relation of HHL to other conditions. Patients diagnosed with HHL could no longer be classified as having an "obscure" condition, though it would remain "hidden" to conventional audiometry. The condition would arguably constitute a variety of APD, though not of OAD, and could also be considered a form of sub-clinical auditory neuropathy, since the latter condition is defined by more severely disrupted neural function. Efforts to understand these correspondences may yield benefits in both research and clinical practice.

The next few years of research into HHL are likely to be pivotal in determining the prevalence, causes, and effects of cochlear neuropathy, not only in listeners with normal audiometric thresholds but in the wider human population. Defining the extent of the problem and developing sensitive and specific diagnostic methods are likely to be of great clinical importance. In summary, forthcoming HHL findings may demand a comprehensive overhaul of the classic account of sensorineural hearing loss and our understanding of what can be regarded as "normal" hearing.

Acknowledgments

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