**APD: A gold standard evidence-based approach**

Vasiliki (Vivian) Iliadou MD, PhD\(^1\), Gail D. Chermak PhD\(^2\), Doris-Eva Bamiou MD, PhD\(^3\), Frank E. Musiek PhD\(^4\)

1. Medical School, Aristotle University of Thessaloniki, Greece
2. Department of Speech and Hearing Sciences, Elson S. Floyd College of Medicine, Washington State University Health Sciences, Spokane, WA
3. Ear Institute, Faculty of Brain Sciences, University College London, London, United Kingdom
4. Neuroaudiology Lab, University of Arizona, Tucson, Arizona, USA.

Dr. Vasiliki (Vivian) Iliadou is an Associate Professor of Psychoacoustics at the Medical School Aristotle University of Thessaloniki, Greece. She has initiated the European APD Study Group and studies cognition measurement and APD.

Dr. Gail D. Chermak is Chair of the Department of Speech and Hearing Sciences at Washington State University. She is recognized internationally for her contributions to the diagnosis and treatment of auditory processing disorder.

Professor Doris-Eva Bamiou, UCL Ear Institute, has received the BAAP Pat Jobson Prize (2002), the RSM Edith Whetnall prize (2012) and the BSA Thomas Simm Littler Prize 2017 for promoting the field of APD.

Frank Musiek is a Professor in the Speech Language and Hearing Department at the University of Arizona. He has made notable contributions in NeuroAudiology (CAPD), functional neuroanatomy and auditory evoked potentials.

**Corresponding author:** Vasiliki (Vivian) Iliadou email: viliad@auth.gr  tel. 00306976805644

Clinical evaluation of hearing is often focused on hearing sensitivity despite evidence that depressed sensitivity alone does not reflect real, everyday listening and communication difficulties. Assessment
of auditory processing should be encouraged, in tandem with education of audiologists, as to the value and importance of more comprehensive evaluation.

**Hearing Evaluation Beyond the Pure Tone Audiogram**

The pure tone audiogram persists as the gold standard of hearing evaluation despite its limitations. While the audiogram is a primary tool for determining type, degree, and configuration of hearing loss, this measure does not provide information beyond hearing sensitivity to a limited range of frequencies (Musiek et al., 2017). It does not reveal the physiological state of the entire (peripheral and central) auditory system (the auditory nerve included), nor the complete physiological state of the cochlea. Auditory neuropathy provides a well-known example of the disconnection between the pure tone audiogram and auditory processing (Draper & Bamiou, 2009). The audiogram explains only a small part of the variance in speech understanding performance and self-reported hearing ability of neurologically normal older adults (Humes et al., 2013). Moreover, adults with central auditory nervous system (CANS) pathology (e.g., due to stroke, traumatic brain injury, degenerative disease) and children with auditory processing disorder (APD) and learning problems present difficulty understanding speech in noise or competing message environments despite normal pure tone audiograms. Notwithstanding these limitations, the audiogram may be the only test conducted for children and adults with auditory complaints, when the audiogram is normal. This is a matter of concern since failure to fully evaluate and identify the sources of the patient’s complaints precludes appropriate, effective, and efficient treatment and management (Chermak et al., 2017). It may also mislead the audiologist and patient into believing their hearing is entirely normal.

One of the primary reasons for this disconnect between research and practice seems to be the ongoing debate as to the interaction between cognition and auditory processing, with proponents on both sides of the debate arguing the neuroanatomical and neurophysiological source of APD. In this brief article, we provide evidence that attention is a term widely and “easily” misused to explain what might in fact be an APD, or even a hearing sensitivity impairment. This misuse of terminology stems
from the assumption that heterogeneity in a disorder undermines its reality. This assumption, however, is incorrect, in that it is incompatible with the nature of many disorders, and with clinical reality. As stated in the DSM-5: “the once plausible goal of identifying homogeneous populations for treatment and research resulted in narrow diagnostic categories that did not capture clinical reality, symptom heterogeneity within disorders, and significant sharing of symptoms across multiple disorders.” (DSM-5 introduction page 12, lines 31-34). Moreover, the DSM-5 notes that “…a too-rigid categorical system does not capture clinical experience or important scientific observations” and that “…the boundaries between many disorder “categories” are more fluid over the life course.” The DSM-5 recognizes that the fluidity of boundaries across disorders “should permit a more accurate description of patient presentations and increase the validity of a diagnosis” (DSM-5 page 5, 25-27, 29-30, 34-35). That APD may have diverse clinical presentations, overlapping symptoms, and comorbidities is thus consistent with the definition of disorders as described in DSM-5. It should also be noted that APD is one of a few neurodevelopmental disorders with a diagnosis based on standardized tests and not merely on symptoms or questionnaires (as is the case with attention deficit hyperactivity disorder (ADHD), autism, etc). Multiple sclerosis is also a good medical example as it can affect several systems but each of these systems involvement must be diagnosed and treated for best help for the patient.

What’s In A Name?

Discussing client needs in an audiology clinic, including cases of APD, involves consideration of the contributions of interdisciplinary teams. This is most appropriate given the comorbidities often present with APD, and standard practice for several neurodevelopmental disorders, in order to ensure
that all pertinent components of an individual’s deficits are addressed. This does not undermine the diagnostic entity of APD. The diagnostic label APD is dismissed by some clinicians, some of whom, interestingly, may be comfortable using the term “hidden hearing loss,” for which the evidence in humans is limited, inconsistent, and some would assert speculative at this time. If you ask why this is the case, the answer probably is that a rather precise anatomical issue is suggested as the source of hidden hearing or cochlear synaptopathy (i.e., the loss of afferent fiber communication at the inner hair cell ribbon synapse). However, permanent behavioral thresholds shifts are not observed until 80-90% of the synapses or inner hair cells are lost (Liberman & Kujawa, 2017) and cell bodies of the affected cochlear neurons can survive for decades despite their loss of functional connection to the hair cells, which means the inner hair cells can survive long periods of time as well. Moreover, as stated above, the evidence for hidden hearing loss in humans is limited and inconsistent at this time.

As argued by Musiek et al (2018), the most likely cause of the primary symptom presumed to reflect ‘hidden hearing loss’---speech recognition in noise difficulties despite normal pure tone thresholds---is APD.

Sensitive and specific tests of APD to evaluate specific components of auditory processing and underlying function of the central auditory nervous system (CANS) provide the clinical tools to examine the sources of patients’ “hearing” complaints, especially related to speech recognition in noise difficulties. We caution clinicians not to be tempted to adopt a diagnostic label—hidden hearing loss- for which there is limited evidence in humans and for which there is no clinically proven means of directly assessing (only inferring) the underlying cochlear synaptopathy. We caution clinicians not to reject APD due to its heterogeneity, a feature it shares with many other disorders, and the need, therefore, for interdisciplinary evaluation by experienced and knowledgeable clinicians. Clinical profiles may not be simple, but that must not deter us from fully examining the underlying components. Evaluation and diagnosis of APD requires a battery of efficient auditory processing tests to examine
the patient’s complaints and symptoms, interpreted in the context of a comprehensive history (including medical issues, past and present) (Chermak et al, 2017).

**Gold Standards for APD and Hearing Loss**

The gold standard for the diagnosis of a disorder is defined as the best available evidence-based methodology for diagnosing the disorder. Current clinical practice guidelines depend on the use of test batteries due to the inherent complexity of any given disorder. This enables more accurate diagnosis. The auditory processing test battery approach (AAA, 2010; European Consensus, 2017) is the best available gold standard approach to APD diagnosis. This is based on the best available evidence as presented in numerous published research papers, as well as clinical practice experience. Moreover, there are several parallels between this APD gold standard and the use of the pure tone audiogram as the gold standard for the diagnosis of hearing loss. Among common characteristics are: (1) the subjective nature of testing, (2) results dependent on the clinician’s ability to elicit behavioral responses that reflect an individual’s true auditory ability, and (3) the need for the test results to be interpreted within the context of medical history to arrive at a conclusion regarding “hearing” status. Pure tone audiometry results may not always agree with objective (i.e., electroacoustic or electrophysiological) audiological tests, yet this does not lead to questioning the pure tone audiogram as a gold standard approach to determining hearing sensitivity and hearing loss. The same logic and standard apply to clinically used auditory processing testing. In addition, all behavioral tests impose a degree of cognitive load, including the pure tone audiogram, including attention, executive function and working memory (Iliadou et al, 2018). However, the extent of cognitive influence on audiometric tests, including those of central auditory processing, is limited such that cognition does not solely drive central auditory processing performance. Ironically, the influence of an auditory deficit on measured cognition often is ignored.
Is it valid to evaluate suspected APD individuals when researching for APD?

A number of controversial positions regarding the relationship between auditory processing and cognition are the result of research design weaknesses, most notably poorly defined participants. Many of the conclusions reached in these studies are based on reports that included participants suspected of APD and/or defined on the basis of parent or teacher reports rather than diagnosed based on test evidence. Conclusions drawn based on the performance of poorly defined participant samples pose significant threats to the validity of the research and cannot be relied upon, because one cannot be sure whether the participants in the study presented any type of true auditory deficit, while these participants may also have had a wide range of other unidentified issues (Chermak, Musiek, & Weihing, 2017). Inflated cognition effects can be obtained when participants with unknown cognitive status are included in study samples. The relationship between central auditory tests (or any behavioral measure) and cognitive measures can appear to be stronger than actually is the case when participants with low cognition are not excluded from the analysis (Brenneman et al., 2017). Next, we present some of the emerging evidence indicating a disconnection between auditory processing and cognition.

Cognition and Audition

It is worthwhile to compare the interaction between APD and hearing impairment and the interaction between APD and cognition. Hearing (hearing sensitivity and auditory processing) facilitates children’s and adults’ development of cognitive abilities. Adults with hearing impairment use a disproportionate amount of their cognitive reservoir to support listening (i.e. perceiving auditory information), as this is particularly challenging given their sensory deficit (Pichora-Fuller et al., 2016). This might translate into decreased capacity of higher cognitive abilities (i.e. short-term and working
memory, focused and sustained attention). Perhaps as a compensatory mechanism, the role of working memory in support of listening increases as the signal is more degraded by background noise. The assessment of cognitive capacity in hard of hearing individuals is influenced by their sensory deficit, as the most common approach to cognitive evaluation is through the auditory channel (i.e. oral instruction, repetition or reordering of auditory stimuli). Since audibility does not guarantee normal auditory processing, hearing sensitivity and auditory processing must be evaluated before cognitive testing takes place to ensure accurate results (Davies, 2017). If this is not feasible, one must insist that a thorough audiological diagnostic evaluation follow initial cognitive testing (Iliadou et al, 2018). Studies with children diagnosed with APD show that only a subgroup presents with attention deficits, mostly focused auditory attention, not necessarily global attention deficits (Stavrinos et al, 2018). This indicates that the interaction between cognition and hearing (auditory processing included) is complex. In a recent study in children diagnosed with APD, a dissociation was seen between attention and rhythm perception demonstrating that attention is not the source of children with APD’s difficulties processing a simple auditory rhythm (Sidiras et al under review). The ability to process rhythm by children with normal auditory processing ability was very much influenced by their attention. This indicates that extrapolating correlations found in typically developing children with children with APD is not a valid approach. Quoting Phillips (1990): “Following from the modern spotlight metaphor of selective attention, it is perhaps more likely that a deficit in selective attention is secondary to the deprivation of attentional processes from their normal neural representations of the acoustic signal.”

**Summary & Conclusions**

APD is a multifactorial presentation and complex disorder, with no single contributing factor able to account either entirely or even for the majority of the clinical profile in patient cohorts – and certainly not in the individual patient. Some might find the single, one test gold standard approach attractive
in organizing one’s practice; however, such an approach is not likely to maximize the accuracy of differential diagnosis, nor promote effective and efficient management for the individual with APD. Clinicians are well aware that a “cookie cutter” procrustean approach does not fit real patients, and that consideration of each individual’s unique characteristics should drive the choice of tests/assessments, overall diagnostic approach, clinical decision making, and intervention. Clinicians must be cognizant of system interactions, including the influence of hearing sensitivity and auditory processing on cognition, especially if cognition is measured exclusively through the auditory modality as well as the limitations of a standard, peripheral hearing evaluation to reveal more central auditory involvement. These understandings will lead to more comprehensive and accurate differential diagnoses and targeted and efficient interventions.