



## Response: Proposed Diagnostic Criteria for Definite Isolated Otolith Dysfunction

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Dear Editor,

We would like to thank Kenneth Chua Wei De and his colleagues for their valuable comments on, and critique of, our new definition of definite isolated otolith dysfunction (iOD) [1]. As iOD is an emerging concept, it is essential to collect expert opinions when refining the diagnostic criteria. We hope that open discussion and ongoing research will soon yield diagnostic criteria that can achieve international consensus. Here, we reply to the points made by the cited authors and comments that we have received from other experts.

1) We agree that, “the pathognomonic otolith symptoms are not well understood yet and may be easily confused.” All of non-spinning, translation, tilt, floating, flipping-over, swaying, or rocking may not be perfect in defining the symptoms of iOD patients. Although iOD symptoms differ from those of typical, peripheral vestibular disorders [2], symptomatic descriptions can mean different things to patients, generalists, physicians, and even otologists [3]. We are especially concerned about the symptomatic overlap between iOD and persistent postural-perceptual dizziness (PPPD) defined by the Barany Society in 2017 [4]. PPPD symptoms may include false or distorted sensations of swaying, rocking, bobbing, or bouncing of oneself (internal non-spinning vertigo) or similar sensation of movements of the surroundings (external non-spinning vertigo) [4]. To overcome the unclear

definitions of symptoms, our definition of definite iOD requires laboratory findings indicating utricular and/or saccular dysfunction. We believe that such an objective requirement will allow clinicians and researchers to differentiate PPPD from definite iOD.

2) It is a true that diagnosis of iOD secondary to (co-existing) Meniere’s disease (MD) and vestibular migraine (VM) can be challenging. If a patient has another, active, well-defined vestibular disorder, it may be safe to focus on the principal diagnosis because the dizziness may not be caused by otolith dysfunction alone (which is not “isolated” in such patients). We now include the text: “Cannot be explained by another disease or disorder” in our definition. This is a stronger statement than that included in the definition of definite MD by the Barany Society [5], which reads, “Not better accounted for by another vestibular diagnosis.” As is the case for patients with definite MD, the diagnosis that better explains the dizziness should be selected when two diagnoses compete or co-exist. In a case of definite iOD, any other active vestibular disorder should be the subject of the principal diagnosis, regardless of how effectively iOD explains the dizziness. This is because iOD remains a controversial concept; no strong diagnostic consensus has yet been attained. Also, it is less likely that the cause of dizziness is “isolated” otolith dysfunction in such patients.

However, it should be noted that definite iOD can be diagnosed when the underlying MD and/or VM are/is no longer active. In such cases, the inactive (past) vestibular disorder may be the cause of the present definite iOD (we describe this as “secondary definite iOD”).

3) It is true that, “abnormal VEMP may not always suggest a peripheral pathology.” Latency abnormality is more often detected in patients with central disorders such as multiple sclerosis [6], vestibular schwannoma [7], stroke [8,9], or migraine [10]. A definite iOD diagnosis requires proof of “abnormal otolith function test: cVEMP, oVEMP, or subjec-

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**Table 1.** Proposed diagnostic criteria for isolated otolith dysfunction (iOD)

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**Definite iOD**

- A. Laboratory findings that indicate OD but normal semicircular canal function.
  - 1. OD proven using the cVEMP and/or oVEMP tests.
  - 2. Normal caloric and vHIT test results (for all vertical and horizontal canals).
- B. Symptoms that indicate OD.
  - Non-spinning, translation, tilt, floating, or flipping-over.
- C. Cannot be explained by another disease or disorder.

**Probable iOD**

Laboratory-based probable iOD

- A. Laboratory findings that indicate OD but normal semicircular canal function.
  - 1. OD proven by cVEMP and/or oVEMP and/or SVV testing.
  - 2. Normal caloric and vHIT test results.
- B. Cannot be explained by another disease or disorder.

Symptom-based probable iOD

- A. Symptoms that indicate OD.
  - Non-spinning, translation, tilt, floating, or flipping-over.
- B. Cannot be explained by another disease or disorder.

**Additional descriptions (not mandatory)**

Idiopathic definite/probable iOD.

- When the etiology of iOD cannot be identified.

Secondary definite/probable iOD.

- When a causal relationship with another disorder is identified.

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cVEMP: cervical vestibular myogenic potential, oVEMP: ocular vestibular myogenic potential, vHIT: video head impulse test, SVV: subjective visual vertical

tive visual vertical (SVV)” [1]. The latency abnormality of VEMP is indicative of a central disorder, not a proof of abnormal otolith function. We have refined our definition of definite iOD to prevent such confusion. Table 4 of our original proposal referred to: “abnormal cVEMP, oVEMP, or SVV” [1]. To reduce confusion, we have changed this to read: “OD proven by cVEMP, oVEMP, or SVV”.

Another expert commented that the SVV test might not be optimal in terms of proving otolith dysfunction because the SVV may be affected by central nervous system conditions (including those of the vestibular cortex). Thus, an abnormal SVV may not prove abnormal otolith function in certain patients. SVV may serve as an indicator of probable iOD, but is not the best vestibular evaluation tool. To reflect this fact, we have changed the definition of definite iOD to: “OD proven by cVEMP or oVEMP”.

4) We agree that consideration of “whether iOD is of a primary idiopathic cause or secondary” is not mandatory when any causal or co-incidental relationship between iOD and a co-existent disorder is unclear.

5) Indeed, no management plan specific for iOD exists. We hope that our effort to establish an acceptable definition of iOD will lead to creation of such a plan in the future.

6) It is important to clarify that the video head impulse

tests must be normal for both the vertical and horizontal canals. iOD can be diagnosed only when all other vestibular organs are normal.

Given the valuable comments that we received, we have revised our iOD definition (Table 1). According to the International Classification of Vestibular Disorders (ICVD) [11], iOD may be a Layer III-A entity (Disorders & Diseases). Our definition reflects all new ideas suggested up to October 15, 2020. Additional discussion will improve the definition. Also, the Barany society and the ICVD authorities must be approached for comment.

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**Conflicts of interest**

The authors have no financial conflicts of interest.

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