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Responses to Drs. Kaski and Bronstein and Dr. Crane

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Dear Dr. Niparko:

We appreciate the thoughtful comments on our manuscript from Drs. Bronstein and Crane. They raise important points in opposition to the notion that vestibular atelectasis (VA) could be the underlying cause for the syndrome of bilateral vestibular hypofunction combined with sound or pressure-evoked vertigo, and we hope to address their concerns here.

We explained what we might suppose would happen to labyrinthine transduction with atelectasis of the membranous labyrinth, but a concern was that the clinical histories provided by Merchant and Schuknecht (1988) did not note any sound- or pressure-induced vertigo in their cases. The methods of Merchant and Schuknecht (1988) indicate that histories in the cases of pathological VA were taken retrospectively from charts and only from the histories of present illness at the time of terminal hospitalization, purposely excluding complaints in the past medical history or review of systems. Furthermore, in the four patients presented in our study, sound- or pressure-induced vertigo was *not*, in fact, the chief complaint in any case. Rather, their presenting symptoms were dominated by

complaints of imbalance and oscillopsia, primarily reflecting the impact of bilateral vestibular hypofunction. In 2/4 cases, the clinical history indicated an evolving syndrome in which initial complaints of episodic vertigo gave way later to imbalance and oscillopsia. This suggests that the underlying pathology – be it VA or otherwise – might evolve over time, and that sound- or pressure-induced vertigo could be a phenomenon that dominates only early in the clinical evolution.

Superior canal dehiscence syndrome has taught us that a sizable fraction of patients who have sound- or pressure-induced vertigo learn to avoid provoking stimuli. Instead they may present with chief complaints of symptoms they cannot control: autophony, pulsatile tinnitus, constant imbalance from dural pulsations into the dehiscent canal, etc. Without careful and specific questioning about both past and present symptoms, sound- or pressure-induced vertigo might not be recognized, and the clinical histories in Merchant and Schuknecht's cases might simply not have ascertained past complaints of sound- or pressure-induced vertigo.

Dr. Crane asks, "Is it possible that unilateral VA could be a source of sound evoked nystagmus in a subset of patients with conditions such as the near-dehiscence of the superior semicircular canal?" We considered this possibility, but rejected it because we found that patients with near-dehiscence of the superior canal have physiological findings consistent with a 3rd mobile window syndrome, including elevated oVEMP amplitudes, reduced cVEMP thresholds and bone-conduction hyperacusis (Ward et al 2013). Furthermore, we have not observed vestibular hypofunction in patients with near-dehiscence.

Both Dr. Crane and Dr. Bronstein point out that most of the VA cases reported by Merchant and Schuknecht had unilateral findings, not bilateral as would be expected from the clinical findings in our patients. We agree with Dr. Crane that unilateral VA may be relatively asymptomatic because unilateral hypofunction could be compensated using signals from the unaffected side. In addition, in only some cases – and perhaps in only the most severe - VA might lead to collapse of the membranous labyrinth in such a way that the saccule or utricle would contact the vestibular face of the stapes footplate, coupling residual hair cell function to sound or pressure stimuli. In the majority of cases of VA, collapse of the membranous labyrinth might only have the effect of hydrodynamic dampening, so that responses to head rotation and caloric stimulation of the canals would appear reduced, giving only a picture of idiopathic bilateral vestibular hypofunction.

Our proposal that the syndrome of bilateral vestibular hypofunction combined with sound or pressure-evoked vertigo might be due to VA was our attempt to explain this rare—but we now note repeatedly—observed phenomenon using known observations from decades of careful temporal bone histopathological studies. At present, this is only a working hypothesis because we have no micro-anatomical data to determine if VA is present in our living patients. Nevertheless, this is a starting point for critical inquiry and further investigations. We hope that advances in imaging of the fluids of the living inner ear that have demonstrated hydrops might soon be harnessed to also probe for VA.

Sincerely,

John Carey, MD Bryan Ward, MD

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