

Under-rated neuro-otological symptoms: Hoffman and Brookler 1978 revisited

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In 1978, Hoffman and Brookler published an article in *The Laryngoscope* to challenge prevailing views on the lack of diagnostic power of certain symptoms often reported by patients to neuro-otologists. Some of these 'under-rated neuro-otological symptoms' include complaints of non-rotational dizziness, blurred and double vision, and the development of visual motion hypersensitivity in patients with balance disorders. In this review, I revisit these visual symptoms in the light of new findings from our laboratory. Double vision due to skew eye deviation can indeed occur in peripheral vestibular disease when there is a large, acute peripheral imbalance of vestibular function. It is more frequent and severe in brain stem disease. In both cases, it is explained by disruption of the torsional vestibular ocular reflex. It is usually assumed that damage to the otolith underlies the emergence of skew diplopia, but recent evidence shows that the vertical canal system is likely to be partly responsible as well. The other 'under-rated symptom' revisited here is what patients describe as dizziness when watching moving objects or whilst walking in visually busy surroundings such as supermarkets. Recent work has shown that this 'visual vertigo' emerges in patients who, in addition to suffering from a vestibular disorder, have increased visual dependence. Visual dependence denotes subjects who preferentially use vision, as opposed to vestibular or proprioceptive input, for spatial orientation and postural control. We do not know as yet what makes some vestibular patients become extremely visually dependent. However, we have provided evidence for Hoffman and Brookler's impression that visually triggered complaints should not be summarily dismissed, as they often point to an underlying vestibular disorder.

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Excellent monographs on the subject of vertigo and vestibular disorders have appeared recently^{1,2}. For this reason, I will not review the whole subject of balance disorders, but rather concentrate on a few poorly understood vestibular symptoms which have been the subject of recent research. Indeed, this article was inspired by Hoffman and Brookler's paper in 1978, called *Underrated Neuro-otological Symptoms*³. This publication, which exudes clinical finesse, was structured around case

reports and set out to dispel the erroneous belief that certain symptoms reported by patients are not useful in the diagnosis of labyrinthine disorders. It shows that in the practice of medicine nothing replaces the combination of a good capacity for observation with the paying of careful attention to the patients' own description of their symptoms.

Some of these under-rated symptoms are complaints of dizziness, light-headedness or a floating sensation. Although presented in different ways, one of the myths in neuro-otology is that these are 'lesser' symptoms than 'proper' rotational vertigo. Whilst it is true that spinning vertigo usually implies disorder of the labyrinth or its immediate central connections, the reverse implication, that these underrated symptoms do not support the diagnosis of vestibular disease, is certainly not true. As Hoffman and Brookler indicate, anyone who has done a sufficient number of caloric tests knows that people use all sorts of terms to describe the vestibular sensation induced by caloric stimulation: faintness, rocking, staggering, light headedness, waviness as well as rotational vertigo, of course. One of my patients, clearly not bothered at all by the procedure, said 'it's like going to the pub but much cheaper'. The obvious conclusion in their paper was that: 'the complaint of dizziness, be it nondescript of rotatory vertigo, must be taken seriously and thoroughly investigated'.

In this paper, I will revisit the two underrated neuro-otological symptoms which, in Hoffman and Brookler's opinion, relate to the visual system.

Double vision and skew eye deviation in vestibular disease

The first neuro-otological visual symptom Hoffman and Brookler discussed is the vestibular patient who complains of blurred vision and, in extreme cases, of double vision. Whilst diplopia should always raise the possibility of brain stem involvement, Hoffman and Brookler quote Lord Brain's article of 1938⁴: 'it is important to remember that double vision may occur as a result of vestibular disorder, lest this symptom should be attributed to ophthalmoplegia and ascribed to a lesion of the nervous system. The two images are seen one above the other and the diplopia is doubtless due to skew deviation of the eyes, a disorder of ocular posture emanating from the labyrinth and sometimes occurring transitorily, as Cairns and I have shown after resection of the auditory nerve.' I suspect that this statement was basically ignored for some 40 years, between 1938 and 1978 to be precise.

The issue has been re-examined by Riordan-Eva *et al*⁵ in a robust study including 18 patients who underwent vestibular nerve section for intractable vertigo or acoustic neuroma. Patients were assessed pre- and postoperatively ophthalmologically and with measurements of the subjective visual vertical (*i.e.* the task of aligning a luminous straight line

in the dark to the perceived gravitational vertical). It was found that 5 patients developed an ocular skew deviation (that is, a vertical squint due to a supranuclear, vertical, disconjugate misalignment of the eyes). Only three reported frank diplopia, lasting 1 day to 6 months. There was an association between large changes in ocular torsional position (ocular tilt) and tilts of the visual vertical. In turn, this was associated with lesser degrees of canal paresis on pre-operative caloric assessment. The results indicate that, as expected, the larger the vestibular imbalance produced by the surgery, the larger the tilts in ocular position and, consequently, in subjective visual vertical. They also suggest that the presence of vertical skew deviation is dependent on the presence of a large torsional change. Although prevalent wisdom dictates that such ocular skew and torsional changes are due to the acute asymmetry induced in the otolith control of eye position, alternative sources such as asymmetry in vertical canal function were mentioned.

It may be helpful to examine why the vestibular system needs to be involved in the control of vertical ocular conjugacy in the first place. Figure 1 is taken from the work of Lopez *et al*.⁸ Imagine that you tilt your head slowly towards your right shoulder. As you do so, you begin to lose good visual contact with the vertically oriented visual world (e.g. try to read this article with your head tilted maximally towards the right or left shoulder). *Prima facie*, a good compensatory vestibulo-ocular mechanism would be to counter-rotate the eyes conjugately in the opposite direction to the head tilt and to produce a disconjugate skew deviation so that both eyes lie parallel to the horizon as we tilt (see Fig. 1). Although such mechanism is present

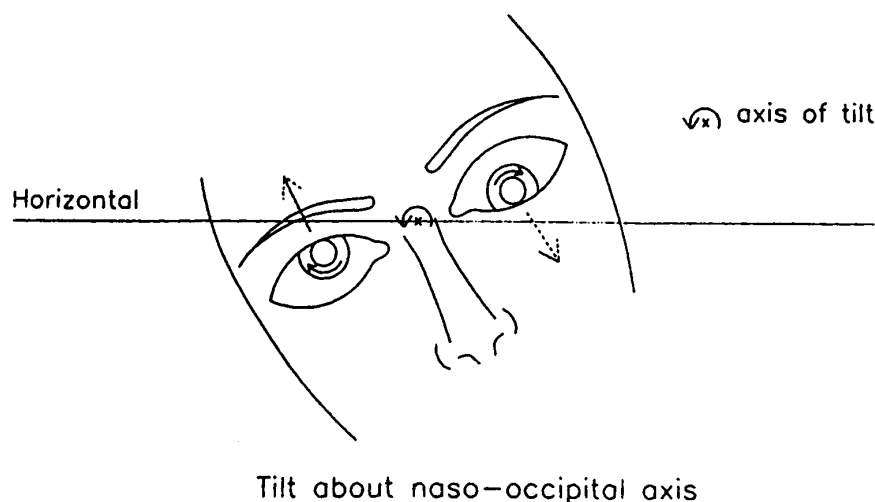


Fig. 1 Diagram showing the compensatory eye movements which could be expected during a right ear down head tilt. The vertical ocular disconjugacy induced is usually called a skew deviation. From Lopez *et al*⁸ with permission.

in animals with laterally placed eyes, like the rabbit, its existence and underlying mechanisms in man are not entirely clear.

Lopez *et al*⁶ examined patients with spontaneous torsional nystagmus and identified the site of lesion in the contralateral vestibular nuclei, in the pontomedullary junction. In addition to this topographic finding, the

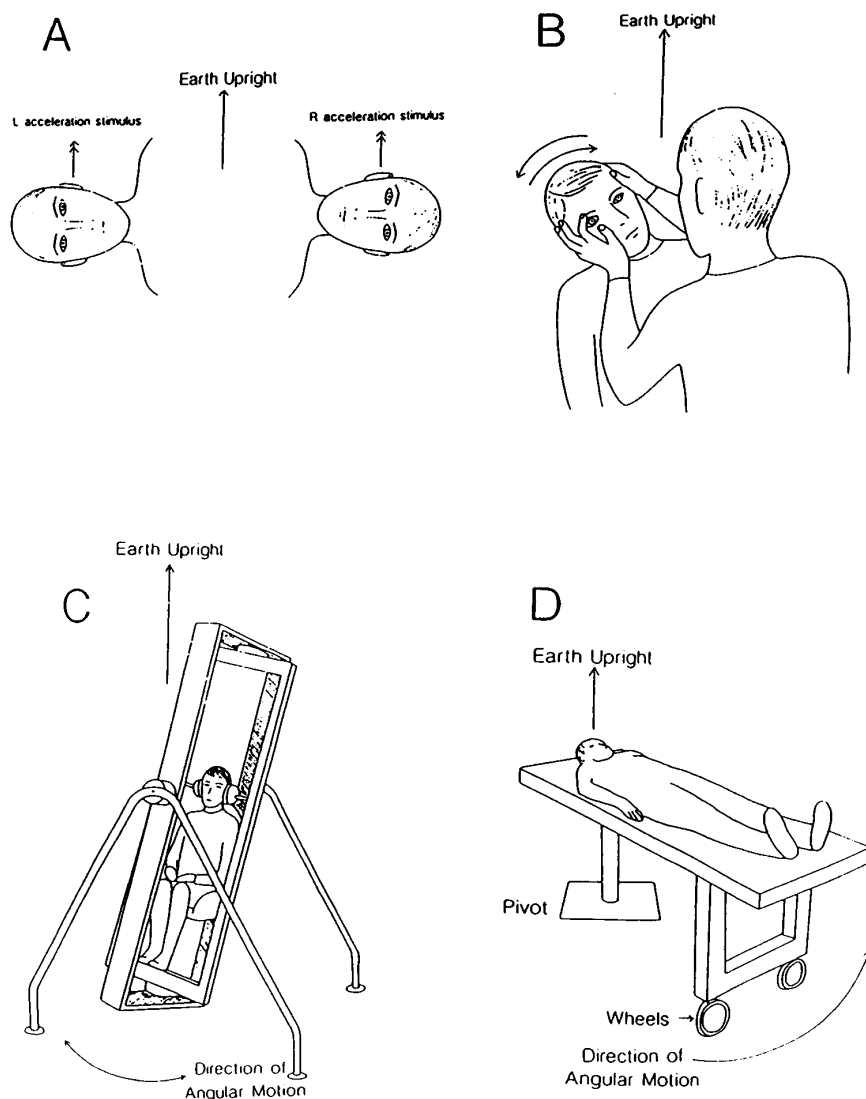


Fig. 2 Diagram showing manoeuvres able to induce torsional eye movements and, therefore, capable of modulating a pathological torsional nystagmus. In (A), a simple head tilt to right or left produces a change in the direction of action of the gravitational vector on the otoliths. In (B), the clinical manoeuvre used to induce ocular counter-rolling activates both otolith and vertical semicircular canals; the gymbals system shown in (C) is similar. In (D), only the vertical canals are activated during rotation since the head is not re-oriented with respect to gravity. From Lopez *et al*⁶ with permission from *Brain*.

authors examined the physiological modulation of the nystagmus during predominantly otolith manoeuvres (static tilt) or predominantly vertical semicircular canal manoeuvres (head rolling movements; Fig. 2). Since the nystagmus was mainly modulated by the latter manoeuvres, the authors concluded that the pathophysiological basis of torsional nystagmus is a functional asymmetry in the central projections of the semicircular canal system. But it was also noted that many of the patients had ocular skew deviations as well. The presence of skew deviations in patients with a disorder of the central semicircular canal system (rather than of the otolithic gravitational system) was puzzling. For this reason, it was decided to investigate the normal physiological basis for this phenomenon, namely whether the normal vertical canal system participates in the control of ocular vertical alignment.

In a series of experiments by Jauregui-Renaud *et al*^{7,8}, normal subjects were whole-body rotated in roll (*i.e.* about the visual axis) with the rotational axis placed either earth-horizontal (subjects upright; Fig. 2C) or earth-vertical (subjects supine; Fig. 2D). In the first condition, the compensatory vestibulo-ocular response is mediated by the otoliths, as they continuously change orientation with respect to the gravitational field, and by the semicircular canals. In the second condition, since the otolith do not undergo re-orientation with respect to gravity, the response is only mediated by the vertical canals. For these experiments, eye movements have to be recorded not with electro-oculography (EOG, ENG) but with techniques able to measure vertical, horizontal and torsional movements, such as the scleral coil technique or video-oculography (3-D VOG). The results showed clearly that a dynamic skew deviation occurs during the roll oscillation and that there was no difference in the magnitude of the skew when subjects were oscillated supine or upright. The conclusion is that the vertical semicircular canal system exerts a dominant influence on vertical, divergent ocular movements. Therefore, lesions to this system, central or peripheral, have the potential to produce pathological skew deviations in man and, consequently, diplopia. In further studies, it was shown that, during whole body velocity steps in the roll plane, the magnitude of the physiological skew deviation decays with a time constant of approximately 5 s⁹. This time constant is identical to the time constant of the vertical semicircular canal system, measured by eye movement⁹ or psychophysical¹⁰ techniques, further indication of the prominent role of the vertical canal system in the origin of the dynamic skew eye deviation.

Returning to 'under-rated neuro-otological symptoms', it is clear that double vision can occur in peripheral vestibular disease, as a result of acute disruption to otolith and vertical canal mechanisms controlling vertico-torsional ocular alignment. Good clinical practice still dictates, however, that care should be exercised before attributing diplopia to labyrinthine disease. Furthermore, clinically obvious skew ocular deviations must be

considered as secondary to CNS lesions disrupting central otolith and/or vertical canal pathways. In this regard, lesions in the vicinity of the vestibular nuclei in the pontomedullary junction usually produce hypodeviation of the ipsilateral eye (e.g. a left medullary lesion produces a skew eye deviation in which the left eye is lower than the right) and mid-brain lesions induce skew deviations with the ipsilateral eye uppermost¹¹. When this rule of thumb does not explain the clinical findings, other mechanisms may be at work. For instance, patients may have paroxysmal skew deviations due to 'irritative' lesions of the brain stem which produce an abnormal increase of activity in saccadic-related areas¹². Since the brain stem saccadic centres are also responsible for the generation of the fast phases of nystagmus, such skew deviations can be considered to arise from abnormally overactive quick components of vertico-torsional nystagmus. Also, cerebellar lesions (e.g. in the uvula) can produce disinhibition and instability in the vestibular vertico-torsional system¹³, leading to paroxysmal nystagmus with skew deviation and disabling oscillopsia (moving images) and diplopia (double vision).

Visual influences on vestibular symptoms

The other under-rated visual neuro-otological symptom discussed by Hoffman and Brookler is exemplified by their 'case 6', a woman with a cerebello-pontine angle lesion who presented with intolerance to visual motion.

Indeed, many patients state that their dizziness or unsteadiness is triggered or increased in surroundings with profuse visual motion or repetitive visual patterns. Patients may dislike traffic, moving crowds, supermarket aisles, watching car chases in movies, ironing striped shirts, or driving on motorways, with many patients displaying several of these triggers. Undoubtedly, in some of these patients the diagnosis is one of anxiety, phobia or panic. But this is not true for all of them, particularly when, as in patient 6 of Hoffman and Brookler, such symptoms develop after a vestibular insult. The origin of these symptoms, termed 'space and motion discomfort'¹⁴, 'visuo-vestibular mismatch'¹⁵, 'visual vertigo syndrome'¹⁶ or 'motorist disorientation syndrome'¹⁷ by different authors, has been recently investigated by Guerraz *et al*¹⁸.

To begin with, it should be remembered that, as soon as the vestibular system is damaged, a neural process of recovery called vestibular compensation gets under way. Guerraz *et al*¹⁸ speculated that, if the process of compensation from vestibular lesions is dependent on alternative sources of sensory information (visual, proprioceptive), individual differences in the functional status of these systems should have a critical influence on the clinical outcome of a vestibular disorder. Since most of these patients do

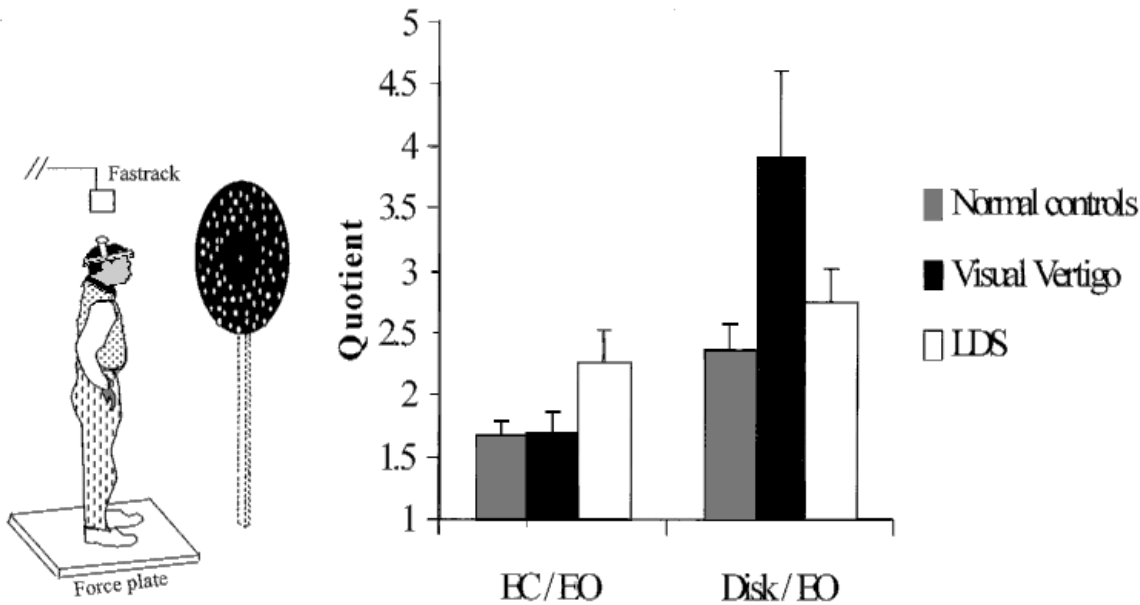


Fig. 3 Diagram illustrating the large rotating disk used to destabilise posture, whilst recording head sway with an electro-magnetic device (fastrak) and force platform posturography (Left). The body sway results show sway path expressed as ratios between two conditions in normal control subjects, bilateral labyrinthine defective subjects (LDS) and patients with visually induced dizziness (visual vertigo). The eyes closed/eyes open ratio (EC/EO), or Romberg quotient, represents the stabilising power of vision, *i.e.* how unstable subjects become with eyes closed relative to eyes in a stationary environment. The Disk/EO ratio indicates how unstable subjects become when viewing the rotating disk in comparison with eyes open in the stationary environment (the destabilising power of vision). Note that the visual vertigo patients are selectively unstable during disk rotation. Reproduced with permission from *Current Opinion in Neurology*²⁰.

not have clinically obvious visual or somatosensory disorders, Guerraz *et al*¹⁸ explored the possibility that minor idiosyncratic differences present in normal people could be the underlying cause. Specifically, it was decided to investigate if patients with visual vertigo were 'visually dependent'.

In essence, a visually dependent person is someone who relies more on vision than on gravito-inertial (vestibulo-proprioceptive) cues for spatial orientation. Visually independent people do just the opposite and can quite happily disregard misleading visual information. Visually dependent and independent people represent the two ends of a continuum in the normal population. However, a patient with a vestibular disorder **and** visual dependency is more likely to be made dizzy by excessive or disorienting visual stimuli than a visually independent subject.

In order to examine the role of visual dependence, a group of patients with dizziness triggered by visual stimuli (visual vertigo) were probed with psychophysical and postural tasks of the type shown in Figure 3. Essentially, the tests measured how much a large field rotating disk (Fig. 3)

and a statically tilted luminous frame (not shown) can alter the perception of verticality whilst seated, and postural balance whilst standing up. Questionnaires were used to measure spontaneous dizziness and autonomic symptoms as well as handicap and trait anxiety levels. In addition to visual vertigo patients, two control groups were tested – a normal control group, and a group of bilateral labyrinthine defective subjects with absence of vestibular function. The latter were included as a positive control group since labyrinthine defective subjects are by definition visually dependent.

The main results of this study were: (i) the majority of the visual vertigo patients were thought to have a peripheral vestibular disorder; (ii) levels of anxiety were similar in the two patient groups (visual vertigo and labyrinthine defective); and (iii) visual vertigo patients had abnormally large perceptual and postural responses to the tilted frame and the rotating disk, *i.e.* they were visually dependent. Furthermore, when the postural sway induced by the rotating disk was expressed relative to the static baseline sway, the visual vertigo patients had significantly larger responses than those in the labyrinthine defective group (Fig. 3).

Conclusions and key points for clinical practice

- The findings support the view that patients whose dizzy symptoms are precipitated by disorienting visual surroundings are likely to have suffered a vestibular episode **and** be visually dependent.
- This combination should be highly debilitating for visual vertigo patients in disorienting visual environments when both visual and vestibular signals are unreliable.
- Clinical experience indicates that vestibular rehabilitation including repetitive optokinetic stimulation can be extremely beneficial in these patients. A formal trial has just been completed by Pavlou *et al*⁹ with very encouraging results.
- The fact that a patient may have additional anxiety or phobic symptoms should not prompt the clinician to think that **all** symptoms in that patient are psychological. Not surprisingly, I also agree with Hoffman and Brookler in this matter.

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