A PATIENT THAT CHANGED MY PRACTICE

Benign paroxysmal positional vertigo is sometimes not so benign

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Volker Henn, Günter Baumgartner and Felix Jerusalem, my teachers at medical school and during my first year of neurology residency in Zurich in 1978, taught me about the approach to and examination of the ‘dizzy patient’. They taught me that some types of vertigo occur only in certain positions of the head or body, or during certain changes of position, and that examination of a dizzy patient is never complete without having performed the Hallpike manoeuvre (Fig. 1), best with Frenzel goggles (glasses consisting of +30 lenses mounted in a frame that contains a light source so the patient’s eyes are easily seen by the examiner and also the patient cannot focus his eyes on an object that abolishes visual suppression of nystagmus). I eagerly read many articles about positioning vertigo, mostly referred to as ‘benign positional vertigo’ – a shift of position provokes vertigo and transient nystagmus. Typically, in the Hallpike manoeuvre, the patient is moved from the sitting to the supine position with the head 30° downward and turned to the left or right. Nystagmus appears after a latency of a few seconds, increases in intensity over another few seconds, then diminishes and disappears. The nystagmus is mainly rotatory, clockwise when the head is down and to the left and anti-clockwise when the head is down and to the right. If the patient looks toward the floor, the nystagmus becomes purely rotatory. If the patient looks at his own nose (away from the floor), it beats upwards (Mumenthaler & Mattle 2004). The patient simultaneously experiences intense rotatory vertigo, perhaps accompanied by nausea.

According to the fascinating animal experiments by Harold Schuknecht from Boston, the posterior semicircular canal on the side the head is turned while down, is involved in the generation of the vertigo. For anatomical reasons, detritus, such as particles shed by the otolith membrane, tend to land in the posterior semicircular canal. They may be caught on the cupula and so cause positioning vertigo due to excessive loading of the cupula. This is called ‘cupulolithiasis’ and was illustrated by Schuknecht with a post-mortem image of the posterior semicircular canal

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Figure 1 Technique to induce paroxysmal positional vertigo (the Hallpike manoeuvre). The patient is taken rapidly from the sitting to the head hanging position while the head is rotated simultaneously 60° to the side. (a) Nystagmus appears after a latency of a few seconds, increases in intensity over another few seconds, then diminishes and disappears. (b) The nystagmus is mainly rotatory, (c) clockwise when the head is down and to the left and (d) anti-clockwise when the head is down and to the right. Illustration from Marco Mumenthaler & Heinrich Mattle. Grundkurs Neurologie, Thieme Verlag, Stuttgart, 2002, with permission.
cupula of a patient who had died for another reason (Schuknecht 1974).

I did not question this pathophysiological explanation for years, even though positioning nystagmus generally fatigues rapidly and can usually only be elicited if the patient is allowed to rest for a while before the test is performed for the first time, or repeated. But if the pathogenesis of excessive loading of the cupula was true, then the vertigo should not habituate and also it should remain the same with every head positioning manoeuvre. It was Thomas Brandt, at that time in Essen, who pointed out this discrepancy between habituation of positioning vertigo and the notion of cupulolithiasis. His explanation was that any detritus is actually floating freely in the endolymph of the posterior canal, and he coined the term ‘canalolithiasis’. Most patients with benign positional vertigo have canalolithiasis and only a few have cupulolithiasis.

By the time I had learned the difference between canalolithiasis and cupulolithiasis I had moved to Bern, and Brandt and Daroff had already published their seminal paper on ‘physical therapy for benign paroxysmal positional vertigo’ (Brandt & Daroff 1980). Whenever I saw a patient with benign paroxysmal positional vertigo I recommended the Brandt-Daroff manoeuvre. Some patients were lucky to lose their vertigo, some patients the manoeuvre did not help. Nevertheless, I eagerly continued to read Brandt’s publications and his monograph on vertigo became my ‘dizzy bible’ (Brandt 1999). He also emphasized what my Zurich teachers had always taught, that positioning vertigo can also be ‘central’ and not benign, but I never saw a patient with central positioning vertigo.

Sometime in the 1990s I heard of the Semont and Epley manoeuvres at a meeting, and one of the speakers claimed a 100% success rate for the Epley manoeuvre for benign paroxysmal positional vertigo (Fig. 2). These manoeuvres are used to flush the detritus out of the posterior semicircular canal and relieve vertigo immediately. Having carefully studied Epley’s original description I started to apply his manoeuvre, and it usually worked – at follow-up a few days later most patients were relieved of their vertigo (Epley 1992).

One day I found a patient who had positioning vertigo with linear horizontally beating nystagmus toward the undermost ear. From Robert Baloh’s publication I had learned that this is the result of canalolithiasis of the hori-
horizontal semicircular canal, a condition which had been described in 1985 by McClure (Baloh et al. 1993; McClure 1985). When the detritus in canalolithiasis is freely floating in the horizontal canal, the direction of the nystagmus is toward the undermost ear. Unlike in canalolithiasis, the detritus in cupulolithiasis is fixed to the cupula, and in cupulolithiasis of the horizontal semicircular canal the nystagmus beats to the uppermost ear.

On a later occasion, when I was already behind schedule with my patients, I saw a 32-year-old woman who complained of positioning vertigo. Her neurological findings were normal except for rotatory, anti-clockwise nystagmus and intense vertigo and nausea when the head was positioned down and to the left. I performed an Epley manoeuvre and, unlike other patients, she still had vertigo when she left the room. I had an uneasy feeling that I could not explain. I wondered about ordering an MR scan if the vertigo had not gone at follow-up. However, she did not come back for follow-up as agreed. Instead she had been brought to our Department of Neurosurgery with a tight posterior fossa because of a giant medulloblastoma and she died a few days later. Only then did I realize that I had not listened to her complaint of a mildly aching pressure in the occiput. Instead, I had focused all my attention on the vertigo. Looking at her records I also realized that her nystagmus had been rotatory anti-clockwise when the head was down and to the left. It should have been rotatory clockwise with benign paroxysmal positional vertigo of the left anterior semicircular canal. I swore to myself that whenever I saw another patient with benign paroxysmal positional vertigo combined with only a tiny trace of an atypical feature I would perform an MR scan immediately.

Last year a retired professor of my university referred his wife because of vertigo. Whenever she looked up or turned to her right when lying in bed, severe vertigo occurred for a few seconds. When lying down she experienced a sensation of being pulled further backwards and downwards but without rotatory vertigo. Her vertigo was purely linear, like being in an elevator accelerating rapidly downwards. Examination with Frenzel goggles demonstrated rotatory and at the same time downbeating nystagmus when her head was positioned down and to the right with the Hallpike manoeuvre. When she sat up the nystagmus was upbeating. As logical as I like to think after my big diagnostic error with the medulloblastoma, I did not order an MR. I consulted Thomas Brandt’s book on vertigo where I found the solution. She had canalolithiasis of her left anterior semicircular canal. It is not clear, which manoeuvre is best to flush the detritus out of the anterior canal but I performed an Epley manoeuvre anyway, and a few days later I received a bottle of Burgundy wine.

REFERENCES