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DOI: 10.1177/1756285612465920

Posted at the Zurich Open Repository and Archive, University of Zurich
ZORA URL: http://doi.org/10.5167/uzh-80080
Accepted Version

Originally published at:
Clinical diagnosis of bilateral vestibular loss: three simple bedside tests

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Key words: Bilateral vestibular loss, head impulse test, dynamic visual acuity, vertigo

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Abstract

Bilateral vestibular loss (BVL) may present with or without vertigo and hearing loss. Amongst the causes of BVL are vestibulotoxic antibiotics, autoimmune ear diseases, Menière’s disease, and meningitis. Clinical diagnosis of BVL is based on the result of three simple bedside tests: a positive head impulse test (HIT), reduced dynamic visual acuity (DVA) and a positive Romberg test on foam rubber. With these signs, diagnosis of severe BVL is usually straightforward to establish.
Patients with bilateral vestibular loss (BVL) may present with or without vertigo and hearing loss. They usually complain about oscillopsia during head movements and about unsteadiness, especially while walking in the dark [Dandy, 1941, J.C., 1952]. Common causes of BVL are vestibulotoxic antibiotics (especially gentamicin) - even after short periods of administration – autoimmune ear diseases such as Cogan’s Syndrome, Menière’s disease and meningitis [Zingler et al., 2007]. BVL may also be associated with multiple system atrophy, neurofibromatosis type 2, hereditary ataxias such as Friedreich’s ataxia [Fahey et al., 2008], spinocerebellar ataxia [Gordon et al., 2003], and episodic ataxia type 2. A combination of vestibular loss, peripheral neuropathy and bilateral vestibular areflexia is known as CANVAS syndrome [Szmulewicz et al., 2011].

Clinical diagnosis of BVL is based on the result of three simple bedside tests: a positive head impulse test (HIT) [Halmagyi et al., 1988], reduced dynamic visual acuity (DVA) during head shaking [Demer et al., 1994], and a positive Romberg test on foam rubber [Halmagyi et al., 1994]. With these three clinical signs, diagnosis of severe BVL is usually straightforward to establish but mild BVL remains a diagnostic challenge, as clinical tests might not be conclusive and data from laboratory tests, including caloric irrigation, rotational chair and video head impulse testing, show considerable overlap between patients and normal subjects [Weber et al., 2009]. Vestibular outcome seems to be independent of age, gender, time course of manifestation and severity of BV [Zingler et al., 2008]. Roughly 80% of patients with BVL do not improve and thus, the prognosis seems less favourable than previously assumed [Zingler et al., 2008].

**Head impulse test**

Due to its short latency (about 10 milliseconds), the vestibulo-ocular reflex (VOR) keeps the eyes on target during head movements with equal eye rotations opposite to the direction of head rotations [Aw et al., 1996]. The image is efficiently stabilized on the retina as long as eye and head velocities are equal. In BVL, the VOR is impaired or absent bilaterally and gaze is only stabilized by visual reflexes, which react with considerable delay, i.e. a latency of 100 milliseconds or more [Viirre et al., 1996].

To perform a HIT, the examiner stands in front of the patient seated at the bedside or on a chair. The examiner asks the patient to focus a target (e.g. the examiner’s tip of the nose). The examiner then manually delivers brisk, passive horizontal head rotations of approximately 10-20 degrees amplitude. Normally, the patient’s eyes keep focusing the examiner’s nose. Instead, in a patient with BVL, where the VOR is impaired or absent, the eyes drift off the target to the side to which the head is rotated, so that the patient has to make a catch-up saccade to move them back to focus the examiner’s nose. Overt saccades after head rotation are the telltale sign of vestibular loss, whereas covert saccades during head rotation remain imperceptible to the naked eye [Weber et al., 2008]. While catch-up saccades appear with head impulses to either side in BVL, they indicate the affected side in patients with unilateral vestibular loss (UVL) whereas the patient’s eyes keep focusing the examiner’s nose when the head is rotated to the healthy side.

Sensitivity of bedside HIT is adequate and therefore clinically useful in the hands of both neuro-otological experts and non-experts [Jorns-Haderli et al., 2007]. In a group of subjects with UVL and
BVL, its sensitivity was significantly lower (63% vs 72%) and its specificity was significantly higher for experts than non-experts (78% vs 64%) when a quantitative HIT measurement with scleral search coils was used as a reference [Jorns-Haderli et al., 2007]. In other words, neuro-otological experts were inclined to trade off sensitivity for specificity of the head impulse test, in order to avoid false-positive results. In another study, the accuracy of the bedside HIT for identifying BVL was comparable with 84% sensitivity and 82% specificity [Schubert et al., 2004]. The bedside HIT can identify patients with severe and moderate BVL. However, identification of mild BVL remains a challenge and covert saccades may conceal BVL even in patients with total vestibular loss [Weber et al., 2009]. In cases of an inconclusive bedside test, it has now become practical to measure HIT with high-speed video goggles in order to quantify the deficit of the vestibulo-ocular reflex (VOR) and detect covert saccades [Macdougall et al., 2009].

**Dynamic visual acuity**

Dynamic visual acuity (DVA) assesses a subject's ability to perceive objects accurately while the head is passively moved. To measure DVA clinically, the examiner oscillates the patient's head horizontally or vertically at 0.5 to 2 Hz and asks the patient to read optotypes on a visual acuity chart [Longridge et al., 1984, Demer et al., 1994]. In computerized DVA, optotypes are usually displayed during directional head impulses above a velocity threshold of 120-150°/s (add Herdman 1998 and Vital 2010). In patients with BVL, the VOR is no longer able to stabilize gaze, while the movements are too fast for the smooth pursuit system to keep the eyes on target. As a consequence, visual acuity decreases compared with what the patient is able to read when the head remains still. In healthy subjects, visual acuity may decline by one or two lines on the optotype chart. A decline of more than two lines is considered abnormal [Sargent et al., 1997, Fife et al., 2000], but severely affected BVL patients may show a decline of five or more lines. Subjects with UVL may also have abnormal DVA, especially at higher head oscillation frequencies (add Dannenbaum E 2009). DVA based on directional head impulses also indicate the side of the lesion in UVL (add Herdman 1998 and Vital 2010). DVA might display false-negative results when other mechanisms such as augmented cervico-ocular and visual reflexes compensate at least partially for the retinal instability during head movements [Chambers et al., 1985, Vital et al., 2010]. However, computerized DVA testing has good sensitivity (94.5%) and specificity (95.2%) in subjects with uni- and bilateral vestibular loss [Herdman et al., 1998].

**Romberg test on rubber foam**

Postural control depends on visual, proprioceptive, and vestibular input. In patients with BVL, postural control is impaired due to a loss of vestibulo-spinal reflexes [Horak et al., 2002]. A simple way of diagnosing ataxia is the Romberg test [Khasnis et al., 2003]: The examiner observes postural stability with the patient placing his feet together, initially with eyes open and then with the eyes closed. The Romberg test is positive when a patient is able to stand with feet together and eyes open, but sways or falls with eyes closed [Lanska et al., 2000]. However, this test is not specific for vestibular loss and does not help to distinguish between UVL and BVL, but also detects cerebellar and proprioceptive impairment. The test's sensitivity can be increased by having the patient stand on a foam rubber, which disrupts proprioceptive inputs [Shumway-Cook et al., 1986, Lanska et al., 2000]. Foam
posturography with eyes closed proved to be very sensitive (up to 79%) and specific (up to 80%) to detect patients with both UVL and BVL [Fujimoto et al., 2009].

Acknowledgements

This research was supported by the Betty and David Koetser Foundation for Brain Research.

Disclosure

Dr. K.P. Weber acts as an unpaid consultant and has received funding for travel from GN Otometrics.
Legends

**Figure 1:** Head impulse test. While the patient is asked to fixate a target, the examiner briskly rotates the patient’s head to the right or left and observes his eye movements. (A-C) In a healthy subject, the vestibulo-ocular reflex (VOR) will keep the eyes on the target. (D-F) In a patient with BVL (re-enacted scene), the eyes will move with the head during head impulses to both sides, so that the patient will have to make catch-up saccades after the head movements to bring the eyes back on target.

**Figure 2:** Dynamic visual acuity. (A) The examiner oscillates the patient’s head at about 2 Hz in the horizontal or vertical plane. (B) The patient is asked to read the optotypes on a visual acuity chart while the head is moving. A patient with BVL will typically loose three or more lines compared to static visual acuity.

**Figure 3:** Romberg test on rubber foam. The patient is asked to stand on a mat of rubber foam with his feet together and eyes closed. Without visual and disrupted proprioceptive input, a patient with total BVL will fall off the mat. The examiner must be ready to prevent the fall.
References


