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Hands-on Course 3

Bedside examination of the vestibular and ocular motor system - Level 2

How to examine the ocular motor system

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Eye position

First, the motor range of each eye is tested separately (with the other eye covered) by having the patient visually fix upon a small visual target, e.g., the tip of a pen, that is moved from straight-head to the limits of maximal horizontal and vertical eccentricities. The same procedure is then repeated with binocular viewing to detect manifest strabismus (= tropia). Finally, the examiner alternately covers either eye using the palm or a paddle (alternating cover test). The examiner looks for movements of the eye that has just been uncovered. These movements are directed oppositely to the deviation of the eye, when it was still covered.

If strabismus is detected, the clinician determines whether the binocular misalignment is: (1) manifest or latent; (2) concomitant, paretic, or internuclear; and (3) horizontal or vertical.

Horizontal eye movements elicited by the alternating cover test are common in healthy subjects, indicating esophoria (eye moves temporally when uncovered) or exophoria (eye moves nasally when uncovered). Vertical eye movements elicited by the alternating cover test, however, are almost always pathological and are due to either a skew deviation or a trochlear nerve palsy.
In patients with acute vertigo and in the absence of a trochlear nerve palsy, a vertical deviation, either manifest or unmasked with the alternating cover test, indicates a brainstem stroke along the graviceptive pathways until proven otherwise (Kattah et al. 2009).

To distinguish a skew deviation from a trochlear nerve palsy, it is helpful to perform the alternating cover test with the patient in both the sitting and supine position. While in trochlear nerve palsy the vertical deviation of the eyes at gaze straight ahead is independent of the body position, skew deviation decreases when the patient is moved from upright to supine (Wong et al. 2011).

Skew deviation is usually associated with static binocular cyclorotation. While static binocular cyclorotation cannot be detected clinically, it can be suspected, however, in cases with additional spontaneous torsional nystagmus of both eyes. Static ocular cyclorotation can be measured by fundus photography. This technique allows distinguishing skew deviation from trochlear nerve palsy: the hypertropic eye is incyclorated in skew torsion, but excyclorated in trochlear nerve palsy. An additional head tilt is seen in both conditions and is toward the lower eye. The combination of skew torsion and head tilt toward the lower eye as part of the vestigial righting response is called the ocular tilt reaction (Westheimer and Blair 1975; Halmagyi et al. 1991), while patients with trochlear nerve palsy tilt their head inadvertently toward the unaffected side to reduce vertical diplopia by inducing ocular counterroll to the affected side (anti-Bielschowsky head position).
Spontaneous nystagmus

Normally, with gaze pointing straight ahead, the two eyes remain stable in their neutral position, i.e., in the center of the orbit. Disorders of the labyrinth, the vestibular nerve, and the central structures of the vestibular and ocular motor systems, however, may lead to drifting of the eyes with various horizontal, vertical, and torsional components. These ocular drifts away from the neutral position are typically interrupted by saccadic movements, so-called fast phases, in the opposite direction. Fast phases bring the eyes back close to their neutral position. Properties of this so-called spontaneous jerk nystagmus are important elements in the differential diagnosis of both acute and chronic vertigo. While spontaneous nystagmus with fast and slow phases can be due to disorders of both the vestibular and ocular motor systems, pendular spontaneous nystagmus is caused by disorders of ocular motor centers and pathways.

Acute spontaneous nystagmus

In patients with acute vertigo, the clinician must quickly form an opinion on whether the underlying lesion is neuroanatomically peripheral or central. “Peripheral” includes the vestibular labyrinth and the vestibular nerve, while “central” includes any brain structure that may, if lesioned or irritated, lead to vertigo. The suspicion of acute central vertigo will always lead to emergency procedures, potentially including stroke treatment, while peripheral acute vertigo is usually not due to a life-threatening condition (Tarnutzer et al. 2011).
An expeditious assessment of spontaneous nystagmus in patients with acute vertigo should answer the following questions:

1. Is the main direction of the nystagmus primarily horizontal, vertical or torsional?

2. In patients with horizontal spontaneous nystagmus, does the direction of nystagmus reverse when gaze is held in an eccentric position in the direction of the slow phases?

3. Does visual fixation suppress or enhance the spontaneous nystagmus?

After answering these three questions, the clinician can attribute the spontaneous nystagmus in a patient with acute vertigo to a peripheral-vestibular or central dysfunction. An exception is a lesion at the root entry zone of the vestibular nerve, which in absence of central neurologic signs may present like a peripheral vestibulopathy, hence the term vestibular pseudoneuritis (Dieterich 2002).

@1: Main direction of nystagmus

Spontaneous nystagmus is almost never purely horizontal, vertical, or even torsional. Directional components in addition to the main nystagmus component result from the coordinate system of the neural signal that causes the nystagmus. They also depend on whether the clinician chooses to describe the nystagmus in a head-fixed or eye-fixed reference system. For instance, in an eye-fixed reference system, spontaneous nystagmus resulting from an acute superior vestibular neuritis may appear more horizontal-torsional at gaze directed to the contralesional side and more horizontal-vertical at gaze directed to the ipsilesional side. In contrast, the very same nystagmus, when described in a head-fixed reference system, shows
little dependence of its direction on eye position. Most examiners use an eye-fixed reference system.

Another factor to consider in observing the different directional components of spontaneous nystagmus is the effect of visual fixation suppression. As the horizontal and vertical components of eye movements are more suppressed by visual fixation than the torsional component, the relative magnitude of the torsional component increases when the patient attempts to fix the gaze on a visual target.

**Horizontal spontaneous nystagmus**

Horizontal spontaneous nystagmus in patients with acute vertigo can be of peripheral-vestibular or of central origin. Only the identification of additional signs allows horizontal spontaneous nystagmus to be attributed to a peripheral or central pathomechanism.

**Vertical spontaneous nystagmus**

Vertical spontaneous nystagmus in patients with acute vertigo is practically always of central origin and either downbeat (upward drift) or upbeat (downward drift). In general, downbeat nystagmus is a chronic sign, whereas upbeat nystagmus appears acutely and most often results from a lesion of ocular motor pathways or centers within the brainstem (Pierrot-Deseilligny and Milea 2005; Kim et al. 2006). Many times, up-beat spontaneous nystagmus disappears over time.

**Torsional spontaneous nystagmus**

Torsional spontaneous nystagmus in patients with acute vertigo is frequently associated with vertical misalignment of the eyes, so-called skew deviation, with the upper poles of the eyes beating in the direction of the
hypertropic eye. Additionally, the eyes are statically cyclorotated with their upper poles in the direction of the hypotropic eye (Brandt and Dieterich 1993). Skew deviation with static binocular cyclorotation, so-called skew torsion, with or without spontaneous torsional nystagmus, may be associated with head tilt in the direction of the hypotropic eye, so-called ocular tilt reaction (Westheimer and Blair 1975; Halmagyi et al. 1991). Torsional spontaneous nystagmus, skew torsion, or ocular tilt reaction in patients with acute vertigo are usually due to a brainstem lesion.

@2: Directional reversal of horizontal spontaneous nystagmus at eccentric gaze

The examiner looks for a horizontal gaze-evoked nystagmus at right and left gaze, in addition to the underlying horizontal spontaneous nystagmus. For not yet completely understood reasons, the drift velocity of spontaneous nystagmus typically increases at gaze increasingly held in the direction of the fast phases (Robinson et al. 1984; Bockisch and Hegemann 2008). This pattern is called Alexander’s law and in the case of horizontal spontaneous nystagmus points to a peripheral dysfunction that causes acute vertigo. If, however, the horizontal nystagmus changes its direction, when the patients holds gaze eccentrically in the horizontal direction of the slow phases, this additional gaze-evoked nystagmus indicates a lesion in the brainstem or cerebellum (Kattah et al. 2009).

@3: Suppression or enhancement of spontaneous nystagmus by visual fixation

Spontaneous nystagmus caused by an acute peripheral vestibular asymmetry usually can be suppressed by visual fixation. This is best evaluated by comparing the nystagmus during fixation with nystagmus with removed fixation, e.g., by the patient wearing Frenzel goggles,
which prevent visual fixation. Visual fixation suppresses only the horizontal and vertical components of nystagmus; therefore a torsional component becomes more prominent. Lacking visual fixation suppression of spontaneous nystagmus points toward a central lesion (Cnyrim et al. 2008), but the presence of visual fixation suppression does not exclude a central lesion (An et al. 2014). Increasing nystagmus during attempted visual fixation is typical for infantile nystagmus syndromes.

**Chronic spontaneous nystagmus**

Chronic spontaneous nystagmus is usually due to a central vestibular or ocular motor dysfunction. The clinician should try to answer the following questions:

1. Is the spontaneous nystagmus jerk or pendular?
2. What is the direction of the spontaneous nystagmus? Does it change over time?
3. Is the spontaneous nystagmus monocular or binocular?
4. Is the spontaneous nystagmus conjugate or disconjugate?
5. Is the spontaneous nystagmus evoked by covering one eye?
6. From the history: is the nystagmus infantile or acquired?

Chronic horizontal spontaneous nystagmus due to a peripheral deficit (e.g., vestibular neuritis) or irritation (e.g., Menière’s disease) is usually transient and, after days, can only be seen under Frenzel glasses. Head shaking or mastoid vibration can unmask a peripheral or central asymmetry. This provoked nystagmus, i.e., after head shaking or during vibration, is also best seen under Frenzel glasses.
Downbeat spontaneous nystagmus is usually due to an impairment of the cerebellar flocculus (Zee et al. 1981). A typical feature of cerebellar downbeat nystagmus is an increase of drift velocity and hence nystagmus intensity with horizontal eccentric gaze. In case of an additional horizontal gaze-evoked nystagmus, the nystagmus direction becomes oblique, i.e., vertical-horizontal, at horizontal gaze eccentricities. Downbeat nystagmus frequently obeys Alexander’s law, i.e., upward drift and therefore nystagmus intensity increases with downgaze. In a minority of cases, however, Alexander’s law of downbeat nystagmus is reversed (Straumann et al. 2000).

Seesaw nystagmus can be pendular or jerk. While one eye moves upward and incyclorotates (upper pole moving in the contralateral direction), the other eye moves downward and excyclorotates. During the second half of the cycle, this pattern is mirrored between the two eyes.

Acquired pendular nystagmus is horizontal, vertical, oblique, elliptic, or circular. Nystagmus amplitude can be different between the two eyes and there are also monocular forms of pendular nystagmus. If the palate moves in synchrony with the eyes, this combination is called oculopalatal tremor or myoclonus.

Periodic alternating nystagmus is a horizontal spontaneous jerk nystagmus that changes its direction every 90-120 seconds. Typically, the nystagmus becomes weaker before changing its direction.

The nystagmus in infantile nystagmus syndrome, formerly called congenital nystagmus, is almost always horizontal, sometimes with a small torsional component. Eye movements are conjugate and may change.
direction, amplitude, and morphology, but not frequency, as a function of
gaze direction. Usually, there is a gaze direction with minimal nystagmus
amplitude, the so-called null zone. Infantile nystagmus syndrome often
includes head tremor. Nystagmus and head tremor increase with visual
fixation and under emotional stress. A variant of the infantile nystagmus
syndrome is latent nystagmus, which is a spontaneous horizontal nys-
tagmus that appears in both eyes when one eye is covered and always
beats toward the uncovered eye.

**Saccadic intrusions and oscillations**

Besides spontaneous nystagmus, visual fixation can be interrupted by
saccadic intrusions and oscillations. These include square-wave jerks,
ocular flutter, and opsoclonus. Myokymia of the superior oblique extra-
ocular muscles manifests as monocular vertical-torsional oscillation.

**Gaze-evoked nystagmus**

Nystagmus beating in the direction of gaze at horizontal or vertical gaze
eccentricities, so-called gaze-evoked nystagmus, is a sign of impaired eye-
velocity-to-position integration in the brainstem and cerebellum. Gaze-
evoked nystagmus must be differentiated from spontaneous nystagmus
obeying Alexander’s law.

Dissociated gaze-evoked nystagmus with smaller and slower fast phases of
the adducting eye, typically together with an adduction deficit of the
same eye, suggests ipsilateral internuclear ophthalmoplegia. The adduc-
tion deficit may disappear or become minimal during convergence.
Smooth pursuit

With the patient visually fixating on the tip of the nose of the examiner, passive horizontal head oscillations at 0.5-1 Hz evoke redundant activity of the vestibulo-ocular reflex and the smooth-pursuit system; if the patient cannot keep the eyes on the target, both mechanisms are deficient.

Ocular smooth pursuit alone is tested by having the patient follow with the eyes a moving flashlight at about 20°/second, first in the horizontal, then in the vertical direction. Deficient smooth pursuit is compensated by saccadic movements that follow the visual target.

Fixation suppression of the vestibulo-ocular reflex

To assess the smooth-pursuit system with the eyes centered and not moving, e.g., when additional gaze-evoked nystagmus interferes with smooth-pursuit eye movements in eccentric positions, visual suppression of the vestibulo-ocular reflex is tested. Provided the vestibulo-ocular reflex is functioning, the inability to suppress nystagmus during head oscillation and simultaneous visual fixation of a head-fixed target demonstrates a deficient smooth-pursuit system.

Saccades

Saccades are tested in the horizontal, vertical, and occasionally oblique directions. The patient, with the head not moving, changes gaze direction between the examiner’s tip of the nose and an eccentric target. Saccades are elicited by verbal command. The examiner assesses the latency, velocity, and accuracy of saccades.
Optokinetic nystagmus

Optokinetic nystagmus is elicited by a visual pattern on a hand-held rotating drum (optokinetic drum). This relatively small stimulus mainly activates the smooth-pursuit system (not the optokinetic system, as the name wrongly suggests). This test allows an efficient assessment of the conjugacy of pursuit eye movements (slow phases of nystagmus) and saccades (fast phases of nystagmus). Patients with visual vertigo standing on foam rubber are sometimes pulled in the direction of the optokinetic drum rotation or show increased body sway in the plane of stimulation.
References

