NORMAL MECHANISMS FOR GAZE STABILITY

In order for us to see an object best, its image must be held steady over the foveal region of the retina. Although the visual system can tolerate some motion of images on the retina (3), if this motion becomes excessive (more than about 5°/second for Snellen optotypes), vision declines. Furthermore, if the image is moved from the fovea to peripheral retina, it will be seen less clearly.

In healthy persons, three separate mechanisms work together to prevent deviation of the line of sight from the object of regard. The first is fixation, which has two distinct components: (a) the visual system’s ability to detect retinal image drift and program corrective eye movements; and (b) the suppression of unwanted saccades that would take the eye off target. The second mechanism is the vestibulo-ocular reflex, by which eye movements compensate for head perturbations at short latency and thus maintain clear vision during natural activities, especially locomotion. The third mechanism is the ability of the brain to hold the eye at an eccentric position in the orbit against the elastic pull of the suspensory ligaments and extraocular muscles, which tend to return it toward central position. For all three gaze-holding mechanisms to work effectively, their performance must be tuned by adaptive mechanisms that monitor the visual consequences of eye movements.

TYPES OF ABNORMAL EYE MOVEMENTS THAT DISRUPT STEADY FIXATION: NYSTAGMUS AND SACCADIC INTRUSIONS

The essential difference between nystagmus and saccadic intrusions lies in the initial eye movement that takes the line of sight off the object of regard. For nystagmus, it is a slow drift (or “slow phase”), as opposed to an inappropriate saccadic movement that intrudes on steady fixation. After the initial movement, corrective or other abnormal eye movements may follow. Thus, nystagmus may be defined as a repetitive, to-and-fro movement of the eyes that is initiated by a slow phase (drift). Saccadic intrusions, on the other hand, are rapid eye movements that take the eye off target. They include a spectrum of abnormal movements, ranging from single saccades to sustained saccadic oscillations.

DIFFERENCES BETWEEN PHYSIOLOGIC AND PATHOLOGIC NYSTAGMUS

It is important to realize that not all nystagmus is pathologic. Physiologic nystagmus preserves clear vision during self-rotation. Under most circumstances, for example during locomotion, head movements are small and the vestibulo-ocular reflex is able to generate eye movements that compensate for them. Consequently, the line of sight remains pointed at the object of regard. In response to large head or body rotations, however, the vestibulo-ocular reflex alone cannot preserve clear vision because the eyes are limited in their range of rotation. Thus, during sustained rotations, quick phases occur to reset the eyes into their working range: vestibular nystagmus. If rotation is sustained for several seconds, the vestibular afferents no longer accurately signal head rotation, and visually driven or optokinetic nystagmus takes over to stop excessive slip of stationary retinal images. Additional examples of physiologic nystagmus are arthrogenic and audiokinetic nystagmus (discussion following). In contrast to vestibular and optokinetic nystagmus, pathologic nystagmus causes excessive drift of stationary retinal images that degrades vision and may produce illusory motion of the seen world: oscillopsia (4–7). An exception is congenital nystagmus, which may be associated with normal visual acuity and which seldom causes oscillopsia (8).

Nystagmus, both physiologic and pathologic, may consist of alternating slow drifts (slow phases) in one direction and corrective, resetting saccades (quick phases) in the other: jerk nystagmus (Fig. 23.1A). Pathologic nystagmus may,

![Figure 23.1. Four common slow-phase waveforms of nystagmus. A. Constant velocity drift of the eyes. This occurs in nystagmus caused by peripheral or central vestibular disease and also with lesions of the cerebral hemisphere. The added quick-phases give a “saw-toothed” appearance. B. Drift of the eyes back from an eccentric orbital position toward the midline (gaze-evoked nystagmus). The drift shows a negative exponential time course, with decreasing velocity. This waveform reflects an unsustained eye position signal caused by a “leaky” neural integrator. C. Drift of the eyes away from the central position with a positive exponential time course (increasing velocity). This waveform suggests an unstable neural integrator and is usually encountered in congenital nystagmus. D. Pendular nystagmus, which is encountered as a type of congenital nystagmus and with acquired brainstem disease. (From Leigh RJ, Zee DS. The Neurology of Eye Movements. Ed 3. New York, Oxford University Press, 1999.)
however, also consist of smooth to-and-fro oscillations; pendular nystagmus (Fig. 23.1D). Conventionally, jerk nystagmus is described according to the direction of the quick phase. Thus, if the slow movement is drifting up, the nystagmus is called “downbeating”; if the slow movement is to the right, the nystagmus is “left-beating.” Although it is convenient to describe the frequency, amplitude, and direction of the quick phases of the nystagmus, it should be remembered that it is the slow phase that reflects the underlying abnormality.

Nystagmus may occur in any plane, although it is often predominantly horizontal, vertical, or torsional. Physiologic nystagmus is essentially conjugate. Pathologic nystagmus, on the other hand, may have different amplitudes in the two eyes (dissociated nystagmus); it may go in different directions leading to different trajectories of nystagmus in the two eyes; or may have different temporal properties, i.e., phase shift between the two eyes, leading to movements that are sometimes in opposite directions (disconjugate nystagmus).

METHODS OF OBSERVING, ELICITING, AND RECORDING NYSTAGMUS

It is often possible to diagnose the cause of nystagmus through careful history and systematic examination of the patient (9,10). History should include duration of nystagmus, whether it interferes with vision and causes oscillopsia, and accompanying neurological symptoms. The physician should also determine if nystagmus and attendant visual symptoms are worse with viewing far or near objects, with patient motion, or with different gaze angles (e.g., worse on right gaze). If the patient habitually tilts or turns the head, the physician should determine whether or not these features are evident on old photographs.

Before assessing eye movements, the physician must examine the visual system, looking for signs of optic nerve demyelination or malformation, or ocular albinism which often suggests the diagnosis. The stability of fixation should be assessed with the eyes close to central position, viewing near and far targets, and at eccentric gaze angles. It is often useful to record the direction and amplitude of nystagmus for each of the cardinal gaze positions. If the patient has a head turn or tilt, the eyes should be observed in various directions of gaze when the head is in that position as well as when the head is held straight. During fixation, each eye should be occluded in turn to check for latent nystagmus. The presence of pseudonystagmus and oscillopsia in patients with head tremor who have lost their vestibulo-ocular reflex must be differentiated from true nystagmus.

Subtle forms of nystagmus, due to low amplitude or inconsistent presence, require prolonged observation over 2–3 minutes. Low amplitude nystagmus may be detected only by viewing the patient’s retina with an ophthalmoscope (11). (Note, however, that the direction of horizontal or vertical nystagmus is inverted when viewed through the ophthalmoscope.) The effect of removal of fixation should always be determined. Nystagmus caused by peripheral vestibular imbalance may be apparent only under these circumstances. Removal of fixation is often achieved by eyelid closure; nystagmus is then evaluated by recording eye movements, by palpating the globes, or by auscultation with a stethoscope. Lid closure itself may affect nystagmus, however, and it is better to evaluate the effects of removing fixation with the eyelids open. Several clinical methods are available, such as Frenzel goggles which consist of 10- to 20-diopter spherical convex lenses placed in a frame that has its own light source. The goggles defocus the patient’s vision, thus preventing fixation of objects, and also provide the examiner with a magnified, illuminated view of the patient’s eyes. An alternative is to use two high-plus spherical lenses from a trial case, or to determine the effect of transiently covering the viewing eye during ophthalmoscopy in an otherwise dark room.

Evaluation of nystagmus is incomplete without a systematic examination of each functional class of eye movements (vestibular, optokinetic, smooth-pursuit, saccades, vergence) and their effect on the nystagmus, since different forms of nystagmus can be directly attributed to abnormalities of some of these movements. Physiological optokinetic nystagmus occurs during self-rotation, but it can be elicited at the bedside using a small drum or tape with alternating black and white lines, although larger displays are more effective in patients with voluntary gaze palsies. The slow phases represent visual tracking, including smooth pursuit; the resetting quick phases are saccadic in origin (12). In children and patients with impaired voluntary gaze, an optokinetic stimulus often provides useful information about both pursuit and saccadic systems (13–17). Vestibular nystagmus can be conveniently induced by rotating the patient in a swivel office chair for 30 seconds and then stopping: post-rotational nystagmus and vertigo are induced, which may help patients identify the nature of any paroxysmal attacks of dizziness. Caloric and other forms of induced vestibular nystagmus are described below.

It is often helpful to measure the nystagmus waveform because the shape of the slow phase often provides a pathological signature of the underlying disorder (18,19). To properly characterize nystagmus, it is important to measure eye position and velocity, as well as target position, during attempted fixation at different gaze angles, in darkness, and during vestibular, optokinetic, saccadic, pursuit, and vergence movements. Common slow-phase waveforms of nystagmus are shown in Figure 23.1.

Conventionally, nystagmus is measured in terms of its amplitude, frequency, and their product: intensity. However, visual symptoms caused by nystagmus usually correlate best with the speed of the slow phase and displacement of the image of the object of regard from the fovea (7).

There are many different methods now available for recording eye movements, and these are discussed more fully elsewhere (20,21). Because many patients with nystagmus cannot accurately point their eyes at visual targets, precise measurement is best achieved with the magnetic search coil technique (Fig. 23.2), since the contact lens that the patient wears can be precalibrated on a protractor-gimbal device. In addition, this is the only technique that permits precise measurement of horizontal, vertical, and torsional oscilla-