Spontaneous Nystagmus

Charles W. Stockwell, Ph.D.

SPONTANEOUS NYSTAGMUS has long been recognized as an important sign of vestibular dysfunction. This type of nystagmus is sometimes difficult to evaluate, so it seems worthwhile to review its characteristics and clinical significance and to provide some practical tips for interpreting it in the ENG tracing.

It should first be recognized that the term, "spontaneous nystagmus," is a misnomer. Spontaneous nystagmus does not arise spontaneously, but rather is caused by an asymmetry in the tonic activity of the vestibular system. This asymmetry mimics the asymmetry induced by natural head rotation, which increases activity on the side toward which the head is rotating and decreases it on the opposite side. Vestibular asymmetry generates a nystagmus with fast phases toward the side of increased activity, that is, a nystagmus with fast phases in the direction of presumed head rotation.

Characteristics

Spontaneous nystagmus has four characteristics that distinguish it from other types of nystagmus:

1. It is a horizontal-rotary nystagmus. ENG is insensitive to the rotary component of the nystagmus and records only the horizontal component, yielding a characteristic sawtooth waveform on the horizontal recording channel (Fig. 1). There is no significant vertical component, so little or no eye movement is seen on the vertical recording channel.

2. Spontaneous nystagmus is rarely purely vertical or purely rotary.
Figure 1. Spontaneous nystagmus in a patient with a recent right peripheral vestibular lesion. Bitemporal leads; upwards pen deflection denotes eye movement.

Vertical upbeat nystagmus usually denotes a lesion in the anterior vermis of the cerebellum. Vertical downbeat nystagmus usually denotes a lesion in the posterior fossa near the cranio-cervical junction. It is conceivable that vertical nystagmus could be due to peripheral vestibular lesions, but such lesions would have to selectively involve a single vertical semicircular canal in each labyrinth—a highly unlikely occurrence. The examiner should be aware that apparent vertical nystagmus, either upbeat or downbeat, detected in the ENG tracing only with eyes closed is nearly always an artifact caused by eye blinks. It should be confirmed with eyes open in total darkness or behind Frenzel's lenses before being designated as an abnormality.

Rotary nystagmus usually denotes a central vestibular lesion. It is conceivable that rotary nystagmus, like vertical nystagmus, could be due to a peripheral vestibular lesion, but such a lesion would have to selectively involve both vertical semicircular canals while sparing the horizontal canal of one labyrinth—also a highly unlikely occurrence.

(2) Spontaneous nystagmus is suppressed by visual fixation. If the nystagmus is relatively weak, it is completely abolished by fixation and present only when fixation is denied either by total darkness or by eye closure. If the nystagmus is strong, it may not be entirely abolished by visual fixation, but certainly is suppressed.

If visual fixation fails to adequately suppress spontaneous nystagmus, there is a lesion within the central nervous system involving pursuit and possibly other pathways responsible for fixation suppression of nystagmus. There is no firm criterion for abnormality of fixation suppression of spontaneous nystagmus, but many examiners apply the criterion for fixation suppression of caloric nystagmus, in which fixation suppression is judged abnormal if it fails to reduce nystagmus slow phase velocity by at least 40 percent. The comparison is usually made between eyes open and eyes closed with the eyes at center gaze in either case. However, when making this comparison, the examiner should be aware that nystagmus is sometimes inhibited by eye closure. If the patient displays an apparent failure of fixation suppression without a concomitant pursuit defect, this abnormality should be confirmed by measuring nystagmus with eyes open in darkness or behind Frenzel's lenses.

(3) Spontaneous nystagmus obeys Alexander's law, that is, it is strongest when the patient gazes in the direction of the fast phases of the nystagmus, weaker when he gazes straight ahead, and still weaker when he gazes in the direction of the slow phases. Alexander classified the strength of spontaneous nystagmus with visual fixation into three degrees—first degree, in which the nystagmus is only strong enough to be present when the patient gazes in the direction of the fast phases; second degree, in which the nystagmus is strong enough to be present when the patient gazes in the direction of the fast phases and also when he gazes straight ahead, and third degree, in which nystagmus is strong enough to be present when the patient gazes in the direction of the fast phases, straight ahead, and in the direction of the slow phases.

It is difficult to test the effect of eye position on spontaneous nystagmus with visual fixation denied (either by total darkness or eye closure), since most patients have difficulty maintaining eccentric eye position in the absence of fixation. However, several investigators have shown that spontaneous nystagmus also obeys Alexander's law with fixation denied. It is strongest when the patient gazes in the direction of the fast phases, weaker when he gazes straight ahead, and still weaker when he gazes in the direction of the slow phases. If the nystagmus was not very strong to begin with, it may cease or even beat weakly in the opposite direction when the patient gazes in the direction of the slow phases.

Robinson et al. have proposed an explanation for the phenomenon of Alexander's law. They argue that a vestibular asymmetry, in addition to causing spontaneous nystagmus, also shortens the time constant of the neural integrator in the central nervous system that is responsible for holding eccentric gaze and induces a mild gaze-evoked nystagmus. Spontaneous nystagmus adds to the gaze-evoked nystagmus when the patient gazes in the direction of the fast phases of the spontaneous nystagmus and subtracts from it when he gazes in the opposite direction.

(4) Spontaneous nystagmus has been defined as nystagmus that is present when the patient is in the sitting position. It is rarely altered by a change in head position from sitting to supine, but it is often altered by position changes from supine to right-ear-undermost or left-ear-undermost, usually increasing in intensity with one ear undermost and decreasing...
or even reversing direction with the other ear undermost. When this happens, the examiner does not know whether to describe the nystagmus as spontaneous or positional, but it really does not matter because the clinical significance of spontaneous nystagmus is the same whether or not it is altered by head position. Presumably the sensitivity of spontaneous nystagmus to head position is due to asymmetrical otolithic input, but this is unproven.

The examiner should distinguish between spontaneous nystagmus altered by head position and true positional nystagmus. The most common of the latter is benign positional nystagmus, but there are other less common types, such as persistent positional nystagmus, which is nonlocalizing, and transient positional nystagmus, which has been described as horizontal canal benign positional nystagmus. The examiner may also evoke downbeating nystagmus when he places the patient in the supine position. Downbeating nystagmus is exacerbated when the patient is supine and if it is fairly weak, it may not be present when the patient is examined in the sitting position.

**Clinical Significance**

Any spontaneous nystagmus with visual fixation is abnormal. Stahle originally stated that any spontaneous nystagmus with fixation denied is also abnormal, but subsequent investigators have disproved this statement. Many normal individuals have weak spontaneous nystagmus with vision denied. Coats recommended that spontaneous nystagmus with vision denied and with the eyes at center gaze be designated as abnormal only if its intensity is at least 6-7 deg/sec. Most examiners now use this criterion.

The most common cause of abnormal spontaneous nystagmus is a sudden unilateral lesion of the labyrinth or vestibular nerve. Patients with such lesions typically present with a history of recent severe vertigo, a unilateral caloric weakness, and spontaneous nystagmus with fast phases toward the side opposite the weakness. The spontaneous nystagmus in such cases is due to the asymmetry caused by the sudden loss of tonic input from the damaged side. Vestibular compensation normally minimizes this asymmetry within a few days, but it often does not entirely abolish the asymmetry, so that spontaneous nystagmus with vision denied may persist for years following a peripheral vestibular lesion, although it is rarely strong enough to be designated as abnormal.

Abnormal spontaneous nystagmus in the absence of a recent unilateral peripheral vestibular lesion is uncommon. Most examiners regard it as a nonlocalizing sign of vestibular dysfunction.

Charles W. Stockwell is Director of the Vestibular Laboratory at Providence Hospital, Southfield, Michigan.

**References**