Recovery Nystagmus Revisited

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Abstract

Recovery nystagmus (RN) describes a spontaneous nystagmus with a fast-phase beating toward the ipsilesional ear. The mechanisms underlying RN implicate central vestibular system compensation processes. The presence of RN is significant because it implies that function has returned from the affected peripheral vestibular system. A case is described where RN was recorded. The processes underlying RN are described.

Key Words: Central vestibular compensation, nystagmus, recovery nystagmus, vestibular system

Abbreviations: ABR = auditory brainstem response, IN = irritative nystagmus, MRI = magnetic resonance imaging, MVN = medial vestibular nucleus, RN = recovery nystagmus, SN = spontaneous nystagmus, SOT = sensory organization test, VOR = vestibuloculomotor reflex

It is not uncommon to encounter a patient who demonstrates a unilateral weakness on caloric testing and also a nystagmus with a fast phase that beats in the direction of the unilateral weakness. This nystagmus may be interpreted as a positional nystagmus if it occurs when the patient is placed in a position other than sitting (e.g., supine or lateral position). Conversely, the eye movement is a spontaneous nystagmus (SN) if it is present with eyes closed in the sitting position (or with eyes open and is accentuated by the loss of visual fixation). Often, a SN persists throughout positional testing. The finding of a SN that beats toward the affected ear has been interpreted in at least two ways, one implicating the peripheral vestibular system and the second implicating the compensation processes of the central vestibular system. In the first explanation, a disease process on the affected side causes the peripheral vestibular system to generate a greater amount of tonic resting electrical activity than normal. This tonic resting activity exceeds that on the unaffected side. The result is an “irritative” nystagmus (nystagmus resulting from increased tonic outflow from the diseased end organ system) that beats toward the affected ear. The second explanation for this finding implicates the process of central vestibular system compensation for reductions in peripheral vestibular system function and has been termed “recovery nystagmus” (RN). We believe that the following case study is an example of RN.

CASE REPORT

The patient was a 47-year-old female who presented to her primary care physician 1 month prior to her otolaryngology evaluation with a 1-week history of pain on the left side of her face and a purulent nasal discharge. She was subsequently treated with antibiotics and nasal decongestant spray without relief. The patient presented to the otolaryngology clinic 1 month later with a 3-week complaint of acute hearing loss in the left ear and a feeling of being off balance. She denied spinning vertigo, lightheadedness, otalgia, tinnitus, aural fullness, aural discharge, and recent trauma.
Audiometric testing (Fig. 1) showed the patient to have a mild, flat sensorineural hearing loss on the left side and normal hearing sensitivity on the right side. Word recognition testing (NU-6) conducted at +40 dB SL and at 90 dB HL revealed 12 percent and 0 percent word recognition ability, respectively, on the left side. A word recognition score of 92 percent was obtained on the right side. Tympanometry revealed bilateral Type A tympanograms. Stapedial reflexes were normal (ipsilateral reflex) or slightly elevated (contralateral reflex) with the driving stimulus presented to the right ear. Stapedial reflexes were absent for ipsilateral and contralateral stimuli with the driving stimulus presented to the left ear. Stapedial reflexes were absent for ipsilateral and contralateral stimuli with the driving stimulus presented to the left ear. Stapedial reflex decay could not be tested for the left ear.

A balance function test was performed. Disposable silver-silver chloride electrodes were placed at the outer canthus of each eye, above and below the right eye and at Fp1 (ground). Results of saccadic subsystem testing (Fig. 3) served as ground. The bioelectrical activity was amplified, filtered 100 to 2000 Hz, and signal averaged (minimum 2000 samples per block). A minimum of two blocks were obtained for each ear so that waveform reproducibility could be assessed. Stimuli were 100-µsec duration, unfiltered clicks presented at a rate of 21.3 Hz and at an intensity of 85 dB nHL. Both condensation and rarefaction polarity stimuli were presented. Only responses obtained with the better of the two polarities (rarefaction polarity) are depicted in Figure 2. Wave I only was present with stimuli presented to the left ear. A wave I–V latency prolongation (5.63 msec) was observed with stimuli presented to the right ear. The source of the latency prolongation was the wave III–V segment (3.30 msec) of the wave I–V interwave interval. These findings were consistent with (1) impaired transmission of electrical signals through the proximal segment of the VIIIth nerve on the left side and (2) a disorder affecting neural conduction through the rostral pontine auditory pathway on the left side.
Results of horizontal pursuit subsystem testing. Note that the patient was incapable of generating smooth pursuit eye movements in either left or right directions.

Figure 2 Results of ABR test. An ABR showing a delay in the wave III–V interwave interval was recorded following right ear stimulation (bottom tracing of top box). Only wave I could be recorded following stimulation of the left ear.

Figure 3 Results of saccadic subsystem testing. Notice evidence of hypermetria for leftward eye excursions.

Figure 4 Results of horizontal pursuit subsystem testing. Note that the patient was incapable of generating smooth pursuit eye movements in either left or right directions.

Figure 5 Results of test for spontaneous nystagmus. Top: horizontal and vertical eye position record with patient sitting, eyes open, and fixating gaze at center on a point 4 feet in front of her. Bottom: horizontal and vertical eye position record with eyes closed. Notice left-beating SN.
Figure 6 Results of testing for positional nystagmus. Notice that the left-beating SN predominates throughout positional testing.

percent left UW since the observed nystagmus represented the left-beating SN only.

Figure 7 Results of caloric testing (uncorrected for presence of SN). Notice that the SN is present during left cool and warm caloric irrigations.

Figure 8 Results of rotary chair testing (0.01–0.32 Hz). The patient demonstrates a phase lead at 0.01 Hz and a right asymmetry. That is, the peak right slow phase eye velocity (i.e., left-beating nystagmus) was significantly greater than the peak left slow phase eye velocity (i.e., right-beating nystagmus). VOR gains are within normal limits throughout the frequency range tested.

Rotary chair testing (Fig. 8) showed the patient to have an abnormal phase lead at 0.01 Hz. Further, the patient demonstrated a right vestibulocular reflex (VOR) asymmetry. That is, the right peak slow phase velocity (i.e., left-beating nystagmus) was of greater magnitude than the left peak slow phase velocity (i.e., right-beating nystagmus). This occurred due to the pre-existing left-beating SN.

Computerized dynamic posturography showed the patient to fall consistently on condition 5 (absent vision, sway-referenced support) of the Equitest (NeuroCom International, Inc.) sensory organization test (SOT) and to be abnormally unsteady on condition 6 (sway-referenced vision and sway-referenced support). These abnormalities (shown in Fig. 9) resulted in a SOT composite score (58 points) that was below the normal upper limits.

Figure 9 Results of sensory organization testing on platform posturography. The patient shows a composite score (58) that is outside normal limits. The composite score is abnormal because the patient falls on condition 5 (eyes closed, sway-referenced platform), where she must rely only on intact vestibular system function.
A gadolinium contrast (GdTPA) T1- and T2-weighted MRI of the head and cerebello-pontine angle was performed 2 weeks after the initial ENT consultation (see Figs. 10A, B, and C). The examination revealed a large extra-axial mass encompassing 3.8 cm in anterior-posterior diameter, 4.0 cm in transverse diameter, and 5.0 cm in superior-inferior diameter. The tumor extended from the left tentorium into the left cerebellopontine angle cistern and left internal auditory canal with mass effect on the brain stem. Signal characteristics were compatible with meningioma. One month later, the patient underwent a supra and infratentorial craniotomy with transpetrous resection of the tumor. Facial nerve monitoring was performed. The tumor was diagnosed as a meningioma. Postoperatively, the patient has a mixed hearing loss (bone-conduction thresholds that are within normal limits) and normal facial nerve function on the left side.

**COMMENT**

This patient presented with balance function test results that were compatible with a left-sided peripheral (i.e., end organ and/or VIIIth nerve) vestibular system disorder (left UW). In fact, the loss of vestibular function on the left side was profound for low stimulus frequencies (as evidenced by the 100 percent UW on caloric testing and phase lead and low VOR gain at the lowest rotational frequency) and within normal limits for higher stimulus frequencies (e.g., as shown by the normal VOR function at higher rotational chair frequencies). The saccadic abnormalities were consistent with a left-sided disorder affecting the generator of horizontal saccades (cerebellar vermis and the paramedian pontine reticular formation/parabulbar region of the pons). Taken together with word recognition test results (i.e., inordinately poor word recognition ability), stapedial reflex test results, and ABR abnormalities, the results of audiovestibular neurodiagnostic testing argued strongly for a mass lesion that compressed the left VIIIth nerve and was large enough to shift the pons contralaterally (explaining the contralateral ABR abnormalities). What was not clear was how a left-sided peripheral vestibular system disorder could result in a left-beating SN. Normally, SN shows a fast phase that beats in the direction of the healthy end organ. In this case, the patient demonstrated a SN that beat in the direction of the impaired peripheral vestibular system.
In a healthy peripheral vestibular system, there are approximately 1 million discharges per second flowing toward the vestibular nucleus from the 18,000 single nerve fibers in the vestibular division of the VIIIth nerve. A SN occurs when the tonic resting activity in one peripheral vestibular system exceeds that of the other. It is not known precisely how great the magnitude of difference in tonic activity is required for SN to be generated. There are two possible explanations for a SN that beats toward an impaired peripheral vestibular system. An "ipsilesional" SN (SN with fast phase directed toward the impaired ear) may occur when the effects of the disorder result in increased neural firing over that normally seen at rest. This has been termed "irritative nystagmus" (IN) and, though rare, occurs in the early stages of a Meniere's disease attack prior to membrane rupture (Matsuzaki and Kamei, 1995). It is hypothesized that increasing fluid pressure on the affected side mechanically deflects the horizontal semicircular canal cupulae and results in chronic depolarization at the level of the hair cell. Despite the fact that IN has been described in the literature, it is observed rarely in the clinic (seldom seen by the primary author in that setting). A slightly more common occurrence is the appearance of RN (Stenger, 1959; Jung and Kornhuber, 1964; Kornhuber, 1974; McClure and Lycett, 1978). RN is believed to occur as an artifact of the process of central vestibular compensation and is described below.

Within hours, electrical activity measured within the contralesional MVN (right side in this example) will be reduced dramatically, although not completely (see Fig. 11, condition 2; see Fig. 12, condition 3). During this period, a human subject may show a contralesional nystagmus with eyes open (e.g., a first- or second-degree SN) or only behind closed lids. The processes responsible for the down-regulation of MVN activity (Curthoys and Halmagyi, 1996) on the contralesional side include:

- **Synaptogenesis.** Healthy axons sprout into synaptic contacts vacated by deafferented, degenerating neurons. The process of synaptogenesis is too slow to account for the speed of compensation in humans (i.e., axonal sprouting occurs within 5 days and return of neural activity in the guinea pig occurs in 52 hours).
- **Denervation sensitivity.** Neurons in MVN on the damaged side that have been deprived of tonic peripheral input become more sensitive than normal to transmitter substances normally released by the afferents connected to the impaired peripheral vestibular system. This process also requires a time course of days as opposed to hours.
Adaptation. In this example, the contralateral side Type I neurons adapt slowly to the increase in activity that occurs due to the loss of inhibitory control over them by the ipsilesional MVN. This results in a progressive diminution in commissural inhibition from these neurons to cells in ipsilesional MVN.

Commissural efficacy. Unilateral peripheral damage results in a change in the synaptic efficacy of commissural connections between MVNs.

Cerebellar shutdown. Midline cerebellar structures respond to the resting imbalance by altering the tonic inhibition to MVN cells, thus rebalancing the system.

Gaze changes. The change in gaze deviation that occurs due to the sudden unilateral loss of vestibular function forces the contralateral VN to increase its activity to recalibrate the gaze system (i.e., restore gaze to center).

Spinal input. Changes in posture resulting from the unilateral loss of vestibular system function are routed through the spino-vestibular pathway to the vestibular nuclei.

It is probable that central vestibular compensation represents a “blend” of any one or all of the above processes. The result is that even following a complete loss of one vestibular end organ, within weeks the patient will be able to ambulate with little difficulty (assuming the circuitry underlying central vestibular compensation is intact).

If the injury to the peripheral vestibular system is not complete, then restoration of function on the ipsilesional side (however slight) following suppression of function of the contralateral
MVN by the central nervous system will result in a transient imbalance where resting tonic activity in the ipsilesional MVN is greater than in the contralesional MVN (see Fig. 11, condition 5; see Fig. 12, condition 4). In this case, a SN beating toward the injured vestibular system may be recorded. This nystagmus was originally termed Erholungsnystagmus (recovery nystagmus) by Stenger (1959). Jung and Kornhuber (1964) described patients with RN as having regained caloric responses despite the presence of RN beating toward the injured system. The authors reported that Stenger (1959) observed RN in patients within 3 months of vascular or inflammatory lesions. RN has been reported in patients following Meniere's attacks. The observation of RN is important because it denotes recovery of function on the affected side (McClure et al., 1981; Parnes and McClure, 1990).

If the injury to the peripheral vestibular system is permanent but the rebalancing of the tonic activity by the central compensating mechanisms is imperfect, it is possible in the early stages of compensation for tonic activity in the ipsilesional MVN to be greater than that of the contralesional MVN (see Fig. 11, condition 5; see Fig. 12, condition 4).

Eventually, if central vestibular system compensation is complete, the patient with unilateral temporary or permanent loss of peripheral vestibular system function will not demonstrate nystagmus at rest (see Fig. 12, condition 5).

In the present instance, it is possible that either of these mechanisms may have occurred. It has been reported by Bederson et al. (1991) that when patients with acoustic tumors are followed over an average of 2 years with serial MRI scans that 6 percent of the patients showed regression in tumor size. Additionally, 40 percent of their patients showed no tumor growth over the same period. It is possible that this large tumor may have decreased in size or that the contents of the posterior fossa may have shifted slightly to accommodate better the mass. Either of these actions could have reduced slightly the effect of
the tumor on the VIIIth nerve, which is considered functionally to be part of the peripheral vestibular system. Although the caloric examination showed a complete loss of function on the left side, the finding of normal gain and VOR phase values for middle and higher rotational chair frequencies with marginal asymmetries suggests that residual function exists in the peripheral vestibular system on the left side. However, it should be noted that our explanations for how function could have been restored on the affected side represent speculations. That is, we do not have serial MRI scans documenting a reduction in tumor size or shifting of the brain stem. This was a practical limitation of the present case report.

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REFERENCES


