

Positional Nystagmus of Central Origin

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Abstract

Audiometric, electrophysiologic, and radiographic findings for a 68-year-old male with an “imbalance” concern are presented. This paper has a two-fold purpose: (1) to present an unusual electronystagmography case study and (2) to highlight the importance of test conditions in lesion localization. The specific disease pathophysiology remains obscure. Repeated hearing tests documented a known hearing impairment with worsening word-recognition ability of the right ear. An initial electronystagmographic exam was normal except for a mild ageotropic direction-changing positional nystagmus with eyes open and fixed. No repeatable click-evoked auditory brainstem response waveforms could be collected. A magnetic resonance imaging of the brain documents diffuse ischemic white matter disease. A repeated vestibular examination some months later supports the initial findings. The case illustrates the importance of following diagnostic protocol, of repeated measures, and of using both a visual fixation and a nonfixation condition for select electronystagmographic subtests.

Key Words: Electronystagmography, gait, positional nystagmus

Abbreviations: ABR = auditory brainstem response; ENG = electronystagmography; MRI = magnetic resonance imaging; PAN = positional alcohol nystagmus; VNG = video electronystagmography

Sumario

Se presentan los hallazgos audiométricos, electrofisiológicos y radiográficos de un hombre de 68 años con una “preocupación” en cuánto a su equilibrio. Este artículo tiene un doble propósito: (1) presentar un estudio de caso electronistagmográfico inusual, y (2) destacar la importancia de las condiciones de la prueba en la localización de la lesión. La fisiopatología específica de la enfermedad en este caso sigue siendo oscura. Las pruebas auditivas repetidas documentaron un trastorno ya conocido de la audición, con una capacidad en deterioro del reconocimiento del lenguaje en el oído derecho. El examen electronistagmográfico inicial fue normal excepto por un nistagmo de posición ageotrópico leve de dirección cambiante, con los ojos abiertos y fijados. No se pudieron registrar ondas repetibles de respuesta auditiva del tallo cerebral evocadas por clic. Una resonancia magnética del cerebro documentó una enfermedad isquémica difusa de la sustancia blanca, desde el tallo cerebral a la corteza. Un nuevo examen vestibular, meses después, apoyó los hallazgos iniciales. El caso ilustra la importancia de seguir un protocolo diagnóstico, de las mediciones repetidas, y del uso tanto de condiciones visuales de fijación y de no fijación para seleccionar las sub-pruebas electronistagmográficas.

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Palabras Clave: Electronistagmografía, marcha, nistagmo de posición

Abreviaturas: ABR = respuesta auditiva del tallo cerebral; ENG = electronistagmografía; MRI = Imágenes por resonancia magnética; PAN = nistagmo alcohólico de posición; VNG = video-electronistagmografía

The following is a case presentation where electronystagmography (ENG) testing provided the primary suspicion for central pathology. Central pathology initially discovered by ENG testing is erratic and interesting. This paper has a two-fold purpose: to present an ENG single case study with an unusual nystagmus finding, whereupon only two positional ENG subtests were determined to be abnormal, and to stress the diagnostic importance of using visual fixation as a useful and beneficial test condition toward lesion localization.

Briefly, ENG examinations are commonly divided into three main subtests: tests of oculomotility, postural tests, and caloric irrigations of the ears. Postural tests are the focus of this paper. These are both static head and body positions and mobile maneuvers for the detection of abnormal nystagmus. The germinal work on this topic was first introduced to the audiologic community in Barber and Stockwell's original text on this subject matter in 1973 (see Barber and Wright, 1973; Barber, 1984). Positional nystagmus is the resulting nystagmus recorded during static or immobile clinical head positions (Brandt, 1993) and is currently differentiated from positioning nystagmus (e.g., the Dix-Hallpike maneuver). It is common practice to complete each positional ENG subtest and positioning maneuvers with constant mental alerting of the patient, avoiding eyelid closure with the use of Frenzel's lenses and using both no visual-fixation and visual-fixation conditions (Brandt, 1993). The number and complexity of static clinical head positions for ENG testing vary among clinical settings. Generally, a supine, a head hanging, and some type of head-lateral position are completed (Baloh, 1984).

Positional nystagmus is known to be present in normal subjects (Barber and Wright, 1973; Schneider et al, 2004), may be

induced by alcohol intake: positional alcohol nystagmus phase I and II (PAN I and II). Classifications have been devised to assist the clinician with differential diagnosis (Aschan, 1961). Common choices toward this end are determinations of the direction and character of the recorded nystagmus as vertically or horizontally beating nystagmus, direction-changing or direction-fixed nystagmus, persistent or transitory character, peripheral or central origin, and signs indicating the laterality of pathology (Barber, 1984; Brandt, 1993). Additionally, assessing how the character of the positional nystagmus changes with visual fixation on a target is common practice to assist in differentiating central from peripheral pathology. Fixation upon a visual target acts to suppress or eliminate peripheral positional nystagmus (e.g., PAN I and II, spontaneous nystagmus) and exacerbate central positional nystagmus (Brandt, 1993).

In this case a direction-changing positional nystagmus is present only during two lateral head positions and then only when the eyes are fixed on a visual target. The test techniques described above were applied in this case. Direction-changing positional nystagmus has a low incidence (<4%) resulting in limited disease localization capabilities. It is a noteworthy communication for the audiologic community but, as in this case, does not provide specific disease pathophysiology information. Detection of direction-changing positional nystagmus does not generally assist in the lateralization or localization of a vestibular system disease or disorder (Lin et al, 1986). Other general characteristics of nystagmus that are known to assist in determining peripheral from central pathology are shown in Table 1.

CASE PRESENTATION

Requests to rule out vestibular system disease and disorders in this clinical setting begin with an initial consultation that includes an audiometric examination. An assessment of the external ears, ear canals, and tympanic membranes, routine audiometrics, evaluation of the patient's candidacy for ENG testing, and gross or bedside balance examinations are completed to observe obvious signs or symptoms of vestibular system disease or disorder. A formal case history is also completed at the time of this initial examination. Pure-tone audiometrics are tested at this time in the event that auditory brainstem response (ABR) testing is completed. This clinic protocol often eliminates the need for further testing. It minimizes complications from external ear aberrations and/or blindness. Finally, it provides the patient with written instructions for ENG testing and allows the clinician to gain a verbal confirmation that the patient understands.

The patient of interest was a 68-year-old Caucasian male who was initially seen by his primary care physician; he was then evaluated by our ear, nose, and throat (ENT) colleagues and then consulted to audiology for ENG testing. This was all completed within six week's time. The patient was known to the audiology clinic by a previous consultation for hearing aids and related visits. Medical attention was requested due to a reported decrease in hearing ability, a complaint of constant imbalance, and the recent disturbance of gait. Additional medical conditions included unspecified obesity, unspecified hypertension, corrected presbyopia, mild to profound sensorineural hearing loss, and hypothyroidism.

The audiologic examination documented the following: normal appearing pinnae, otoscopically clear external auditory ear canals, normal but "cloudy appearing" tympanic membranes, normal acoustic immittance measures (type "A" tympanograms) with crossed and uncrossed acoustic reflexes recorded at normal levels (90 dB HL for the right ear and 80 dB HL for the left ear) and without acoustic reflex decay of either ear. Pure-tone audiometrics were without any (less than a 5 dB HL) change at any given frequency as compared to a prior examination 14 months earlier. The result of pure-tone audiometrics was a symmetrical mild (30–35 dB HL at 250 and 500 Hz) to profound (90–95 dB HL at 4 kHz and 8 kHz) bilateral sensorineural hypacusis presumed to be a noise-induced hearing impairment given his history of unprotected military noise exposure. Speech reception thresholds were bilaterally identical to the previous examination. Measures of word-recognition ability (CD-recorded NU #6 word lists, presented at 40 dB sensation level in both examinations) were decreased from 80% correct to 56% correct for the right ear while these same measures for the left ear were without change (80% correct). A central auditory aberration was suspected due to the unilaterally decreased word-recognition ability. No additional audiometric tests were performed.

A dizziness questionnaire (adapted after Goodhill and Harris, 1979) was completed with the patient at the time of the audiologic consultation and indicated a constant imbalance with an onset of three to four months ago. Results included daily short-lived periodic episodes of lightheadedness upon rising to a standing position from a seated position. There were very brief bouts

Table 1. Differentiation between Spontaneous Nystagmus of Peripheral and Central Origin

	Appearance	Fixation	Gaze	Mechanism	Localization
Peripheral	Combined torsional, horizontal	Inhibited	Unidirectional (Alexander's Law)	Asymmetric loss of peripheral vestibular tone	Labyrinth or vestibular nerve
Central	Often pure vertical, horizontal, or torsional	Usually little effect	May change direction	Imbalance in central oculomotor tone; usually central vestibular, may be pursuit or OKN	CNS, usually brainstem or cerebellum

Adapted by permission from Baloh and Honrubia, 1990, table 5-2, p. 120.

of nausea without emesis associated with the lightheadedness. There was occasional staggering to the right side when walking. Further, there was clumsiness of the upper extremities with unintentional dropping of objects. The remainder of the dizziness questionnaire was devoid of any additional general health, neurological, optometric, ophthalmologic, or otologic concern. Based on the dizziness questionnaire information, a bilateral vestibular paresis or a right-sided unilateral vestibular paresis and orthostatic lightheadedness were the probable conclusions reported.

The requested ENG examination was scheduled and completed three weeks later. The patient was prepared to complete ENG testing by abstaining from items prior to examination: food and tobacco products the morning of the examination, alcoholic drinks for three days prior, and prescriptions to control dizziness and/or induce sedation for two weeks prior. He brought his eyeglasses and was as well rested as possible. The dizziness history was reviewed at the time of the ENG examination and reiterated a now four- to five-month history of orthostatic lightheadedness, constant imbalance, and gait disturbance.

Electrode placement, impedance, and patient cooperation were documented as "excellent" at the time of the examination. The test environment was an average medical center examination room without windows and with a comfortable temperature. Testing was completed on an electronic-powered ENG table with the lighting dimmed to a semidark condition. Commercially available two-channel ENG test equipment was used with bitemporal horizontal electrodes and vertical electrodes placed above and below the left eye with a ground electrode placed on the forehead. Frenzel lenses (rather than eye closure) were used when appropriate. Visual fixation was completed with the use of a dimly lit otoscope held lightly to the middle of one of the Frenzel lenses alternately over each eye.

Horizontal and vertical extraocular eye movements were found to be normal. These included calibration saccades, tests of vertical and horizontal gaze, horizontal saccades, and horizontal pursuit. A search for spontaneous nystagmus (sitting motionless as well as after a head shaking test with Frenzel's lenses) was negative: without nystagmus. Postural tests

included a head-hanging-right Dix-Hallpike maneuver, a supine to head-right maneuver, static positional tests that included body right, head right, supine, head-hanging-right and head-hanging. All of these head-right tests were repeated with the head left. Select static head position tests were simply monitored for up to two minutes after completing a positioning maneuver. There was an unusual positional nystagmus, described in detail below. Finally, alternate binaural bithermal open-water caloric irrigations of the ears with an examination of fixation suppression were completed. Caloric responsiveness (averaging 17 degrees/second maximum slow phase velocity) was used to determine a vestibular paresis (<8% right), a directional preponderance (<17% right), and a failure of fixation suppression (>80% suppression) that were all within normal limits.

Positioning and positional test findings read as, "a consistent and repeatable ageotropic positional nystagmus with eyes open and fixed upon a visual target" (see Figure 1). This nystagmus was consistently present as long as the eyes were fixed on a visual target and averaged 5 degrees per second. There was no reported vertigo; in fact, when questioned during the recorded nystagmus, the patient reported that he felt fine: "not dizzy at all." Mental alerting tasks were used during positioning and positional tests to optimize nystagmus identification. These tasks consisted of continuous counting, backwards, voicing every third number, from 100. Figure 1a depicts a part of the positional (static head position) testing some 10 to 25 seconds into the head-left position for a second time (without the supine to head-left maneuver). In this position, the patient's torso is supine, and the neck and head are rotated to the left with the left ear as the undermost ear. The tracings in Figure 1a reveal a right-beating horizontal nystagmus when the eyes are open and fixed on a visual target, and there is no nystagmus evidenced when visual fixation is eliminated. Figure 1b depicts the head-hanging-right position (of the first head-right Dix-Hallpike maneuver) approximately two minutes and seventeen seconds after the maneuver (thus, positional) such that the head is being held in the static head-hanging-right position. What is observed is a left-beating horizontal nystagmus again only when a visual fixation on a target is

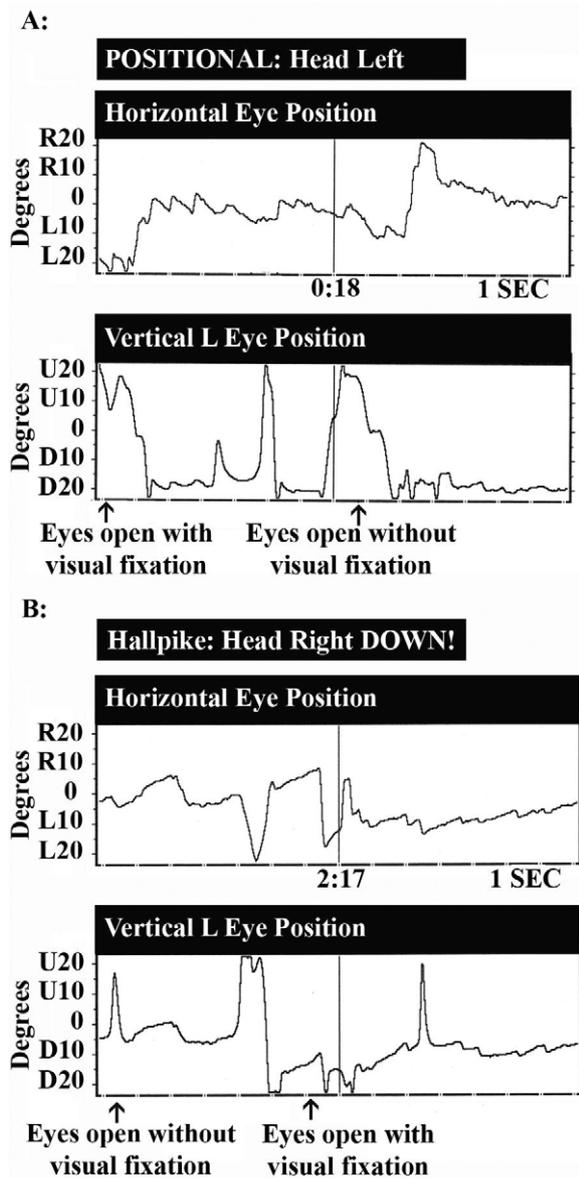


Figure 1. Horizontal nystagmus (upper traces): (A) horizontal nystagmus found 10 seconds into the static head-left position; (B) horizontal nystagmus found approximately two minutes and 17 seconds after the head-right Dix-Hallpike maneuver.

provided. A nonlateralizing central vestibular disorder was suspect.

Due to unilaterally decreased word-recognition scores of the right ear on a follow-up audiometric examination and in an attempt to rule out central pathology due to these vestibular test findings, auditory brainstem response (ABR) tests were completed. The patient was seated and then

reclined in an electronically powered chair with the head supported in the same semidark examination room as above. A conventional two-channel ABR test was completed with commercially available equipment using a click stimulus at a slow stimulus rate and at high intensity. For these tests, click intensities ranged from 70 to 90 dB nHL and click rates ranged from 7.7 to 27.7 clicks per second. Noninverting electrodes were attached to abraded skin within each external ear canal (tip-trodes) and referenced to the forehead with ground placed on the nasion. Stimulus repetition rate, stimulus intensity, and the bandwidth of biologic filters were all manipulated to enhance waveform repeatability and waveform identification confidence without success. The degree of hearing impairment in the high frequencies was reported as the reason for our inability to record a repeatable and measurable ABR result rendering ABR testing ineffectual. No additional electrophysiologic tests could be completed at that time.

Following the audiologic, ENG, and ABR assessments, it was suggested to ENT that central pathology would need to be ruled out via radiographic study. A magnetic resonance imaging (MRI) of the brain with specific attention to the brainstem, internal auditory canals, and cerebellum was ordered as well as a neurology consultation. The neurology examination noted no obvious focal neurological deficits. Some questionable orthostatic lightheadedness was noted, which was noted without significant blood pressure/pulse aberrations. The MRI examination was completed with 3 mm axial and coronal images with and without contrast media. Reported findings include mild early frontal parietal atrophy, tiny scattered areas of white matter ischemic disease mainly involving the left para-ventricular region and a "focal event" within "right-sided midlevel pontine structures." The MRI examination was negative for cerebral edema, mass effect, hemorrhage, ventricular system, and cerebellar aberrations. The MRI, however, has no diffusion weighted images (sensitive for acute events), and therefore any reference to a "focal event" can not be substantiated. A second reading of this scan suggests that there are multiple scattered lesions from medullary and pontine (adjacent to the cerebellar peduncle) to cortical levels. The

lesions are ischemic in nature, presumably due to small vessel cerebrovascular disease. A referral to the Physical Medicine and Rehabilitation Service was made for a fall prevention evaluation and therapy. A follow-up video electronystagmography (VNG) was completed by another clinician at this same facility and with VNG equipment 23 months later because poor balance complaints persisted. This examination produced the same results described above but with higher maximum slow phase velocities for the caloric-induced nystagmus (average 56 degrees per second) and without evidence of torsional or rotary eye movements.

DISCUSSION

The patient of interest described the following new medical concerns to his primary care physician: a recent decrease in hearing ability, a generalized imbalance, and a gait disturbance. He was referred to our ENT colleagues, who requested ENG testing. The patient's presenting history as above, especially the new onset gait dysfunction, is not commonly that of a person suffering from a peripheral vestibular dysfunction as there was no true vertigo reported (Whitman et al, 2001). Serial audiometrics document a decreased word-recognition ability of the right ear as compared to an examination completed some 14 months earlier. The requested ENG testing was completed following the audiometric examination due to imbalance and documented an unusual direction-changing ageotropic positional nystagmus, presumably of central origin. ABR testing was completed due to the depressed right ear speech audiometry result and abnormal ENG findings. Despite multiple stimulus and filter manipulations, no repeatable ABR waveform could be produced stimulating either ear. This result was first felt to be due to the profound degree of bilateral high-frequency hearing impairment. Afterthoughts consider that the poor ABR repeatability may have been disturbed by the diffuse and multiple lesions evident within the brainstem by MRI scan. This remains questionable as no consistent compound action potential or cochlear microphonic could be recorded bilaterally. Otoacoustic emission, electrocochleography, Rotary chair, and posturographic tests were not available at that time. A follow-up VNG

testing some 23 months later, completed at the same facility, resulted in the identical findings (not worse) with a more standardized visual fixation target and without video or graphic evidence of torsional or rotatory eye movements. These repeated measures reduce the possibilities of these results being artifact (eye blink, square wave jerks) or technical error (Bell's phenomenon).

Now, in regard to the direction-changing positional nystagmus of central origin, this is often described as a tonic imbalance within the central processors of the vestibular system. Specifically, this case presentation can not elucidate further because of the diffuse nature of the lesions. These diffuse lesions may include multiple sclerosis, disorders and/or diseases of the posterior fossa and the lateral brainstem. Other focal lesions may be considered as well, such as tumor, infarction, and hematoma (Leigh and Zee, 1999). Gaze holding and fixation capabilities are mediated with contributions from many cortical, midbrain, and brainstem nuclei: the nucleus prepositus hypoglossi, the medial vestibular nucleus, and the ocular motorneurons (cranial nerve nucleus III, IV, and VI) are known brainstem participants. This particular case presentation describes a presumed central disorder or tonic imbalance of fixation visual system due to diffuse and multiple ischemic lesions. That is, visual fixation on a target only in a lateral head and recumbent body position produced a mild ageotropic nystagmus. Unfortunately, due to the diffuse nature of the lesions, the lack of any verifiable focal lesions at brainstem levels, the poor quality of the MRI scan, and the noted variability in reading this scan, no specific cause and effect relationship between these intra-axial lesions and the resulting nystagmus can be developed.

Interestingly, other clinical measures of both the pursuit and fixation systems remain within normal limits in this case. Horizontal and vertical gaze testing was completed at 20 and 15 degrees of visual angle. This testing was maintained for a 20 second duration. This testing was completed normally, without nystagmus and without any central drifting of the eyes. Horizontal pursuit that shares similar fixation system neuronal pathways tested at target frequencies of 0.2 to 0.7 Hertz had normal and symmetrical velocity gains of 0.9 to 0.5 respectively. Visual fixation

suppression for all four caloric irrigations was normal with 100% suppression for the cool irrigations and >80% suppression for the warms. There is no evidence of any positioning effects as the static head positions were comfortably maintained without movement over several minutes, repeatedly, and the nystagmus only appears when the eyes are allowed to fix on a lighted object. Finally, there was no evidence of any peripheral vestibular system concern (unilateral paresis, PAN I, PAN II, and no spontaneous nystagmus) as there was no complaint of vertigo. There was no classic spontaneous nystagmus, and caloric studies were normal. Again this direction-changing nystagmus only appears and changes direction in these two positions with visual fixation. The provocative head position and resulting nystagmus bring the suggestion that this is a nystagmus perhaps related to cervical compression of the vertebral arteries. Arguments against this condition include the autoregulation of the central circulatory system, the lack of acute (without fixation) nystagmus, the lack of vertigo, and the central nature of this direction-changing horizontal nystagmus (see Table 1).

There is often an irresistible urge to use a presumed audiometric indicator of central pathology, the lack of repeatability in ABR testing, and the central character of the pathologic nystagmus to support the definitive MRI evidenced ischemic white matter disease also known as Binwanger's disease (and vice versa). Other health factors in this case include an unspecified obesity, unspecified hypertension, and hypothyroidism. These vascular and metabolic factors may suggest a predisposition for transient vertebrobasilar insufficiency and orthostatic lightheadedness. These same factors are not associated with true vertigo. Finally, it is further tempting to suggest that the increased caloric responsiveness found upon the follow-up ENG is related to the progression of this disease process and decreased central regulation of the vestibule-ocular reflex.

Now, with regard to test technique, most mild positional nystagmus is commonly considered "a normal variant" (Barber and Wright, 1973; Schneider et al, 2004). The patient would have been declared disorder-free had positional tests and/or the use of visual fixation conditions been omitted, given

that the remainder of this patient's ENG examination was normal. Indeed, it may be argued that positional and positioning testing could have been eliminated altogether because there was no historical evidence of postural vertigo. The case described here emphasizes the importance of maintaining and completing sound clinical testing protocols, specifically, postural tests with the patient's eyes open with adequate mental alerting and using both visual fixation and a non-visual fixation test condition.

The patient presented here was lost to follow as he suffered a rapid decline in cognition, general health, and mobility. The patient's cognitive abilities quickly declined, and his combative behaviors increased to a point where further behavioral testing was not possible. Although there is no cure for Binswanger's disease (see Pantoni and Garcia, 1995) and no recovery is possible, additional medical attention is being provided toward this patient's vascular and circulatory system concerns. Fall prevention measures and therapy will likely prevent bodily injury, and the patient's quality of life will be maximally maintained. He currently resides in a full-service nursing home under the care and direction of his primary care physician, the fall prevention team, and the geriatric psychiatry services of our medical center.

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