Downbeat Nystagmus with a Pseudocycloid Waveform: Improvement with Base-Out Prisms

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Downbeat nystagmus in primary position and oscillopsia resulted from nutritional deficiency during prolonged intravenous therapy of a patient with hyperemesis gravidarum. Wide bandwidth infrared oculography demonstrated a pseudocycloid nystagmus waveform with an increasing-velocity exponential slow phase. Because the oscillopsia decreased and the nystagmus was damped with convergence, visual acuity improved with the addition of base-out prisms to each spectacle lens.


Downbeat nystagmus is a spontaneous vertical jerk nystagmus with fast phases beating downward with the eyes in primary position; it is usually attributed to abnormalities of the craniocervical junction [5], including Arnold-Chiari malformation and basilar invagination. Downbeat nystagmus has also been described in patients with idiopathic [23] as well as alcoholic [6] cerebellar degeneration, drug intoxication [2, 19], brainstem encephalitis [17], demyelinating disease [5, 13], magnesium depletion [16], vascular disease [5], and communicating hydrocephalus [15].

The slow phase of downbeat nystagmus previously has been defined as linear and thought to be caused by a unidirectional pursuit defect [20] or tonic imbalance in the central vestibular pathways [3]. Recently, however, Zee and colleagues [21] described downbeat nystagmus with increasing-velocity exponential slow phases in a patient with cerebellar dysfunction, a waveform heretofore described only in patients with congenital nystagmus (CN), in whom it has been restricted to the horizontal plane. These authors attributed the increasing-velocity exponential slow phases to excessive positive feedback in the hypothetical brainstem neural integrator. Acquired downbeat nystagmus in which exponentially increasing and decreasing and linear slow phases have been seen varying from beat to beat, and sometimes within beats, has also been reported [18].

We describe a patient with downbeat nystagmus characterized by pseudocycloid (PC) waveforms.

Case Report

A 29-year-old female pregnant with her third child was hospitalized during the seventh week of pregnancy because of intractible vomiting. Despite treatment with fluids, antiemetics, and high-calorie infusion, she lost 35 pounds over three months. About the twelfth week of pregnancy she complained of vertical oscillopsia, unsteadiness, and vertigo. Her husband noticed her eyes “bobbing up and down and turning in circles.”

A neurologist found marked torsional nystagmus with horizontal and vertical componentts present in all positions of gaze. The cranial nerves were otherwise normal. There was no significant deficit in power, sensation, or coordination, and deep tendon reflexes were present. She had postural hypotension and positioning vertigo. Vitamins, including thiamine, were added to the infusion, and the primary-position nystagmus disappeared. Although the hyperemesis improved initially, vomiting continued. She was discharged on a regimen of oral vitamin supplements and antiemetics during the fifteenth week of pregnancy. She still had vertical oscillopsia, disequilibrium, positioning vertigo, and symptoms of postural hypotension. Intermittent vomiting did not abate until after the birth of a healthy baby. Eight-and-a-half months after parturition she reported improvement in her mild disequilibrium, ataxia, and positioning vertigo but continued to be troubled by oscillopsia. She could improve her vision by tilting her head backward or by closing her left eye.
On examination she was alert, oriented, and intellectually intact. Corrected binocular visual acuity was 20/30-2 at distance. Color vision and pupillary reactions were normal, and confrontation visual fields were full. She was orthophoric and had a full range of eye movements. Downbeat nystagmus was present in primary position and in all other fields of gaze but diminished with downgaze and convergence. The amplitude was greater in the left eye. The remainder of the neuroophthalmological and neurological examination of this patient with Wernicke's encephalopathy was normal.

Investigations including roentgenography of the craniocervical junction and head computed tomographic scan were interpreted as normal.

**Methods**

Vertical eye movement recordings were made by using infrared oculography with a system bandwidth (position and velocity) of DC to 100 Hz (Biometric Model-200 and a Beckman Type R rectilinear Dynograph). It was necessary to modify this technique for recording vertical eye movements. In horizontal recordings, the phototransistors are aimed at the limbus on the left and right of each eye. This positioning is impossible in the vertical plane because the iris-scleral border is obscured by the eyelids. To record vertical movements, the patient's lower lids were slightly retracted with adhesive tape, leaving the entire lower portion of the iris visible but still permitting the patient to blink. The optoelectronic assembly was positioned well below the center of the pupil, with the phototransistors angled sharply toward the center of the lower margin of the iris. The outputs of the transistors were summed rather than used differentially. With this technique, linearity was obtained over a range of approximately \( \pm 10^\circ \) vertically. The patient was seated with head brace and chin rest at the center of an arc with a radius of 1.14 m. Targets were red light-emitting diodes mounted on the vertically oriented arc. Blink artifacts were detected by vertically placed surface electrodes. The eyes were recorded during fixation at 0 and 5° above and below primary position as well as during convergence.

**Results**

**Eye Movements**

An example of the patient's nystagmus in primary position is illustrated in Figure 1. The nystagmus beats are characterized by accelerating upward drifts off target followed by downward braking saccades \[11\] of insufficient amplitude for target refoveation. The saccades, therefore, are followed by decelerating slow eye movements which bring the eyes back on target. The direction of this nystagmus is the direction of the corrective saccade, despite its small amplitude. In some cases the small braking saccade is not clearly evident from the position record alone but is shown clearly by a velocity tracing. At times, the amount of curvature (acceleration phases) of the slow phases varied from beat to beat; occasional runs of beats were virtually linear. The frequency of the beats in primary position was 2 Hz. The nystagmus amplitude varied from one beat to another and was greater in the left eye. It increased slightly in amplitude on upgaze and decreased on downgaze (Fig 2). In downgaze the intensity of the nystagmus (frequency \( \times \) amplitude) was greatly reduced; the amplitude was affected more than the frequency. Convergence damped the nystagmus (Fig 3).

**Visual Acuity**

When we added 7 diopter base-out prisms and \(-1.00\) diopter spherical lenses to the patient's refractive correction for distance, her binocular acuity improved to 20/20-3 and the nystagmus and oscillopsia disappeared. The \(-1.00\) diopter spherical lenses are necessary to
nullify the accommodation produced by the prs-induced convergence; this preserves clear vision at distance.

Discussion

This patient with Wernicke’s encephalopathy acquired vertical jerk nystagmus with PC waveforms that were damped with convergence. Base-out converging prisms induced both a net improvement in her visual acuity and the disappearance of oscillopsia.

Accurate eye movement recordings of jerk nystagmus have identified three different slow-phase waveforms: increasing-velocity exponential, decreasing-velocity exponential, and linear. Different pathophysiological mechanisms have been postulated to explain each [8]. For example, a high-gain instability of the slow eye movement subsystem may be responsible for all varieties of waveforms in CN; the jerk types of CN have increasing-velocity exponential waveforms. Defective tonic ocular motor neuron activity results in gaze-evoked nystagmus with decreasing-velocity exponential slow phases (gaze-paretic) [1]. An imbalance in tonic vestibular innervation causes vestibular nystagmus, which has jerk waveforms with linear slow phases.

Downbeat nystagmus is the term originally used to describe nystagmus with linear upward slow phases and downward fast phases when the eyes are in primary position. Although its mechanism is not yet known, several theories have been proposed. Zee and co-workers [20] postulated a selective deficit in the vertical smooth pursuit system. Baloh and Spooner [3] suggested that downbeat nystagmus indicated a dysfunction in the floccular connections to vestibular neurons or vestibuloculomotor pathways. Thus, a tonic imbalance either in the pursuit system or between the tonic vestibular inputs from each labyrinth may cause a sawtooth pattern of nystagmus—one with constant-velocity (linear) slow phases.

Zee and co-workers [21] described for the first time a patient with cerebellar dysfunction who showed downbeat nystagmus with increasing-velocity exponential slow-phase waveforms; this pattern has also been observed in a patient with Arnold-Chiari malformation [14]. Similarly, in flocculectomized monkeys the downbeat nystagmus waveforms have had either exponentially decreasing or exponentially increasing slow phases [22]. Zee and co-workers [21, 22] postulated on the basis of computer simulation that this nystagmus arises from a brainstem neural integrator that has become leaky (causing decreasing-velocity slow phases) or unstable (causing increasing-velocity slow phases) because of cerebellar dysfunction. They suggest that the flocculus controls the time constant and stability of the brainstem ocular motor integrators.

Jerk nystagmus with PC waveforms is one of the many types exhibited by patients with horizontal CN [10]. This waveform is reported here for the first time in a patient with acquired nystagmus. Many subtleties of these waveforms can be revealed only through accurate ocular motility recordings, being impossible to see clinically. The most nearly imperceptible are the small braking saccades [11]. In all forms of acquired jerk nystagmus previously described, braking saccades were always foxtailing saccades. In PC waveforms, braking saccades are insufficient to refoveate the target fully. They are, therefore, followed by decelerating slow eye movements that bring the eyes back on target. Thus, PC waveforms are the result of an accelerating drift off target (typical of all waveforms exhibited by patients with CN) stopped by a small braking saccade which is followed by a slow eye movement that moves the eyes back on target. Because of the rapid acceleration of the initial drift off target (slow phase) and the refoveating slow eye movement that follows the braking saccade (fast phase), this waveform is often clinically confused with pendular nystagmus.

Horizontal nystagmus is an early and common finding in Wernicke-Korsakoff syndrome. Vertical nystagmus is less frequently observed and not well characterized either clinically or oculographically; when present, it is usually gaze-evoked. Zumstein and Meienberg [24] recently reported a patient with Wernicke’s encephalopathy and upbeat nystagmus. The waveforms had decreasing-velocity exponential slow phases. Previous reports of downbeat nystagmus in patients with a presumed diagnosis of Wernicke’s encephalopathy lack ocular motility recordings, precluding comparative discussion of waveforms. We have recorded a patient with an Arnold-Chiari malformation and downbeat nystagmus with similar PC waveform characteristics. These data suggest that downbeat nystagmus with PC waveforms is not pathognomonic of Wernicke’s encephalopathy but may relate to the site of the lesion.

The most common causes of downbeat nystagmus (posterior fossa malformations, tumors, multiple sclerosis, spinocerebellar degeneration, brainstem infarction) frequently involve more than one area of the

Fig 3. Effect of convergence on nystagmus. (A) Primary position, at long distance; (B) at near distance. (Abbreviations as in Figure 1.)
neuraxis, which may be the reason for the different slow-phase waveforms described. The recent discovery of the multiple waveforms reflects the use of highly accurate eye movement recording techniques and the current interest in analyzing different waveforms in an effort to understand the underlying mechanisms.

CN may be damped by convergence. This effect of convergence on downbeat nystagmus has not been reported, although Cox and co-workers [7] described upbeat nystagmus changing to downbeating with convergence in a patient with a putative diagnosis of Wernicke-Korsakoff syndrome, and Carl and associates [4] reported five patients in whom convergence enhanced the upward slow phases. Daroff and colleagues [9] described the convergence-induced cessation of an acquired, gaze-evoked horizontal nystagmus.

Because the reduction in visual acuity caused by nystagmus is proportional to the intensity of the movement, our patient's vision improved with suppression of her nystagmus by converging (base-out) prisms. Prism therapy has been used to reduce CN by exploiting the gaze angle or convergence angle, or both, that produced minimal nystagmus [12].

Our patient complained of oscillopsia (an illusory movement of the environment), as do most patients with acquired nystagmus. The nystagmus (as shown in Figure 2) increased with upgaze and diminished with downgaze; thus, our patient tilted her head backward when viewing targets straight ahead, thereby increasing her visual acuity and reducing her oscillopsia. With downgaze, the braking saccades became smaller and foveation time increased; this was the predominant reason for the patient's preferred backward head tilt.

References