

Horizontal Pursuit Defect Nystagmus

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A teenage girl with a chronic large posterior fossa tumor had primary position right-beating nystagmus with a constant-velocity slow phase. Smooth pursuit was normal to the left but absent to the right. Vestibuloocular movements were present in both directions. The findings are analogous to downbeat and upbeat nystagmus, in which unidirectional pursuit palsies have been postulated as the pathogenic mechanism. This is the first reported case of a horizontal pursuit defect causing nystagmus.

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Ann Neurol 5:449-452, 1979

Zee and colleagues [14] determined that patients with primary position down-beating nystagmus have a unidirectional absence of pursuit in the downward direction. Vestibuloocular movements were intact, establishing the specificity of the pursuit abnormality. Zee and colleagues postulated that the downward pursuit deficit creates an imbalance, with unopposed upward pursuit tonus causing the eyes to drift upward (slow phases), which is corrected by saccades (fast phase). These workers thus identified a unidirectional pursuit defect as a mechanism for a type of jerk nystagmus. Major characteristics of the nystagmus were a linear (constant-velocity) slow phase and lack of substantive changes in darkness.

Subsequent reports [1, 6] demonstrated that upbeat nystagmus is also secondary to a pursuit abnormality; in these cases the impairment is in upward pursuit, leading to a linear tonic drift (slow phase downward). We present here the first case of a unilateral pursuit abnormality causing horizontal jerk nystagmus.

Case Report

An 18-year-old girl had been experiencing vertigo with nausea and vomiting since the age of 3 years. The episodes, lasting from one minute to several hours, were described as being precipitated by "nervousness." When these attacks were severe, she was unable to maintain the upright position. She had a lifelong history of nonprogressive impaired coordination in the left upper limb, facial asymmetry, and decreased hearing in the left ear.

Vital signs, general physical examination, and mental status were normal. Tandem gait could not be performed. The patient swayed in all directions when standing or sitting with eyes closed. Other findings were ataxia and difficulty with rapid alternating movements in the left upper and lower extremities, and diminished sensitivity to

pain over the left side. Tendon reflexes were slightly more active on the right; plantar responses were flexor on the left and neutral on the right. She had a mild peripheral left facial paresis, depressed left corneal reflex, and markedly impaired hearing in the left ear.

A right-beating, primary position, horizontal jerk nystagmus was present, which increased in intensity with gaze to the right. The nystagmus diminished with leftward gaze and was absent at about 30 degrees to the left. In more extreme leftward gaze, the nystagmus was variable and tended to dissociate between the two eyes. The left eye frequently beat downward, whereas the right eye usually had a rotary, leftward-jerking component. During vertical gaze the patient had right-beating nystagmus, the intensity of which depended on the relative horizontal deviation.

Audiometry revealed anacusis in the left ear with a normal stapedial reflex. Cerebrospinal fluid was under normal pressure with no cells and a protein concentration of 120 mg per milliliter.

CT scanning without contrast medium indicated large calcified densities in the left cerebellum and normal cerebellopontine and prepontine cisterns. There were large areas of contrast enhancement adjacent to the calcified region that involved the whole ventral aspect of the cerebellum extending anteriorly, probably into the brainstem. The enhancement abutted the anterolateral aspect of the fourth ventricle. The remainder of the examination was normal. The scan was identical to one performed elsewhere two years earlier.

The clinical impression, based on the lifelong stability of the patient's clinical state, the identical findings on CT scans two years apart, and a normal posterior fossa angiogram, was hamartoma or dermoid involving the ventral aspect of the left cerebellum with extension into the brainstem.

Methods

Eye movements were recorded using the infrared reflectance technique, with a recording system bandwidth of DC

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to 100 Hz. Velocity was computed by electronically differentiating the eye position signal. The patient was seated in a modified dental chair with chin rest and head brace during testing of saccades and smooth pursuit. Saccades were obtained by asking the patient to refixate between red light-emitting diodes located on a 1.14 m radius arc centered around the patient. The pursuit target was a small spot of light moving sinusoidally or with fixed velocity, back projected on a screen.

The sequence of testing was as follows: After calibration, the patient was asked to refixate between 0 degrees and targets placed every 5 degrees out to 30 degrees in each lateral direction. Pursuit was then tested with ramps (constant velocity) of 5 and 10 degrees per second to the left and right, and with sinusoids (pendular) of various frequencies. The vestibuloocular reflexes (VOR) were tested with the patient being rotated in a Barany chair.

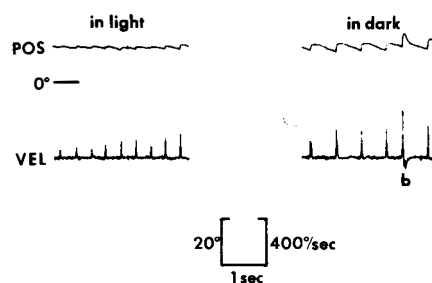
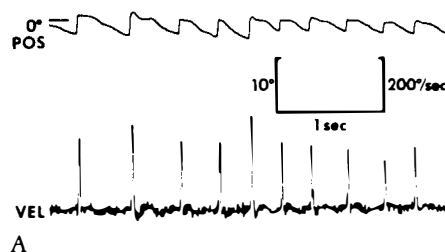
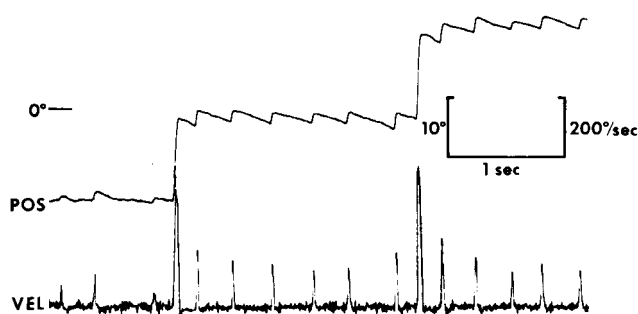
Results

Right-beating nystagmus, identical in the two eyes, was present in primary position and was increased in amplitude with rightward gaze (Fig 1). The slow phases were linear. Frequency was related to gaze, with values of 2.1 Hz at 20 degrees left, 2.5 Hz at 0 degrees, and 2.8 Hz at 20 degrees right. There were occasional examples of flattening of the waveform between the fast and slow phases (Fig 2A), indicating that the eyes were stationary. Convergence abolished the nystagmus, but square-wave jerks intervened to interrupt fixation. In darkness, the frequency of nystagmus decreased and amplitude increased, so the slow-phase velocity remained unchanged (Fig 2B).

Pursuit of the constant-velocity ramps was absent to the right (Fig 3A), where the patient made a series of staircase saccades. Identical findings occurred with sinusoidal tracking (Fig 3C).

Pursuit to the left was present. Tracking a 5 degrees per second ramp to the left resulted in a pursuit eye movement faster than the target (Fig 3B, left) necessitating rightward back-up saccades. When the

Fig 1. Increase in amplitude and frequency with increasing rightward gaze, going from 15 degrees left to 15 degrees right. (In this and succeeding figures, deflections up are rightward movements, deflections left are downward: POS = position, VEL = velocity.)



B

Fig 2. (A) Modification of jerk nystagmus waveform, showing 100- to 150-msec periods without eye movement (flattened segments of POS trace). (B) Effect of darkness: nystagmus frequency is decreased and amplitude is increased, leaving slow-phase velocity unchanged. (b = blink.)

patient was pursuing a 10 degree per second ramp (Fig. 3B, right) the initial eye velocity was approximately 25 degrees per second; this caused her eyes to overtake and pass the target at a point approximately 200 msec after the onset of target movement (point *t* of Fig 3B, right). Although her eyes were decelerating, their velocity was never reduced to that of the target and therefore continued to increase the amount by which they led the target. Back-up saccades did not occur. We did not test at ramps greater than 10 degrees per second, and we cannot compare eye and target velocities for her sinusoidal tracking.

Optokinetic stimuli to the patient's right produced no response, and the ongoing right-beating nystagmus did not change. Targets moving to the patient's left increased the amplitude of the ongoing nystagmus (Fig 4), reflecting summation with the optokinetic nystagmus.

A vestibuloocular reflex in darkness was present in both directions (Fig 5A). With the patient fixating on a stationary object (Fig 5B), the VOR was also present in both directions, although the rightward gain was somewhat reduced. In an attempt to suppress the VOR, the patient was asked to fixate a target rotating with her. Rotation to the left showed good fixation, whereas rotation to the right produced a brisk nystagmus (Fig 5C), indicating inability to suppress the leftward vestibuloocular movement of the eyes.

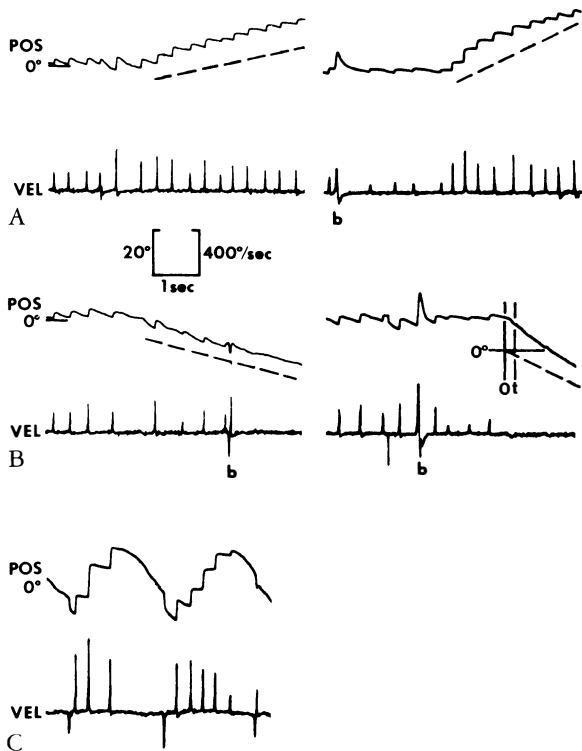


Fig 3. (A) Attempted ramp smooth pursuit to right at 5 degrees per second (left) and 10 degrees per second (right). Dashed lines indicate slope of target trajectory. Note complete lack of smooth movements to the right. (B) Ramp smooth pursuit to left at 5 degrees per second (left) and 10 degrees per second (right). Note the backward saccades in the 5 degrees per second pursuit. In the 10 degrees per second pursuit, the vertical O axis indicates time of stimulus onset: t indicates the point at which the patient's eyes caught the target. Although eye velocity exceeded 10 degrees per second and took the eyes past the target, no backward corrections are seen. (C) Attempted sinusoidal smooth pursuit demonstrated normal leftward pursuit and absent pursuit to the right. Tracking to the right was accomplished by saccades. (b = blink.)

Discussion

Our patient's horizontal right-beating nystagmus met the criteria for pursuit-defect nystagmus established by Zee et al [14]. The slow phase was linear and did not increase in velocity during darkness. Pursuit was absent in the direction opposite to the slow phase, but vestibuloocular movements were present in that direction.

A primary position horizontal jerk nystagmus with a linear slow phase could also be secondary to a vestibular lesion. Right-beating vestibular nystagmus would, however, be associated with impaired vestibuloocular movements in the rightward direction and normal pursuit in both directions. Other forms of primary position horizontal jerk nystagmus have exponential slow phases as distinct from the linear slope in pursuit-defect and vestibular nystagmus. The slow phase in congenital nystagmus usually

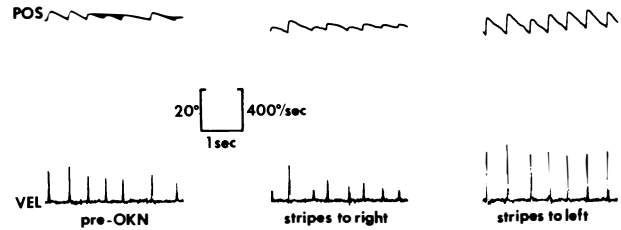


Fig 4. Optokinetic responses. Nystagmus during rightward stripe movement is the same as that with no stimulation: response to left-going stripes appears to be a summation of optokinetic nystagmus and ongoing nystagmus.

has an increasing-velocity exponential [4], whereas in manifest-latent nystagmus the slow phase is decreasing in velocity [8].

The momentary cessations of the nystagmus (see Fig 2A) are a feature characteristic of congenital nystagmus [4, 5], which we had not seen previously in other adult patients with posterior fossa disease.

The tumor in our patient has probably been present since birth or early infancy, implying that the developing nervous system may employ similar adaptation to congenital and early acquired nystagmus. Another finding also observed in congenital nystagmus was the abolition of the nystagmus during convergence. Caloric-induced nystagmus may be damped by convergence [7], but complete inhibition in pathological nystagmus has been reported only infrequently [3].

The inability of our patient to suppress her VOR to the left while fixating an object moving with her (Fig 5C) confirms previous observations that such suppression is related to pursuit [9, 11, 13, 15]. The normal leftward pursuit could suppress the rightward VOR, but the leftward VOR was unopposed because of the absent rightward pursuit.

The precise anatomical extent of the lesion in our patient is unknown. The CT scan indicated major involvement of the ventral aspect of the left cerebellar hemisphere, certainly sufficient to explain the left hemiataxia. The left facial weakness and deafness could be secondary to extraaxial involvement, but the normally appearing cerebellopontine cistern on the CT scan suggests intrinsic left pontine involvement, which could also account for the right hyperreflexia. The sensory loss over the patient's entire left side is best explained by a right-sided brainstem lesion.

Unilateral cerebellar hemispherectomy totally abolishes pursuit in the ipsilateral direction [12]. In our patient, the pursuit defect was contralateral to the cerebellar lesion. Pursuit is generated ipsilaterally within the pons. Although the major brainstem abnormality in our patient was on the left side, the rightward pursuit palsy had to be secondary to a right-sided brainstem lesion as was postulated to explain the left-sided sensory defect. This explanation

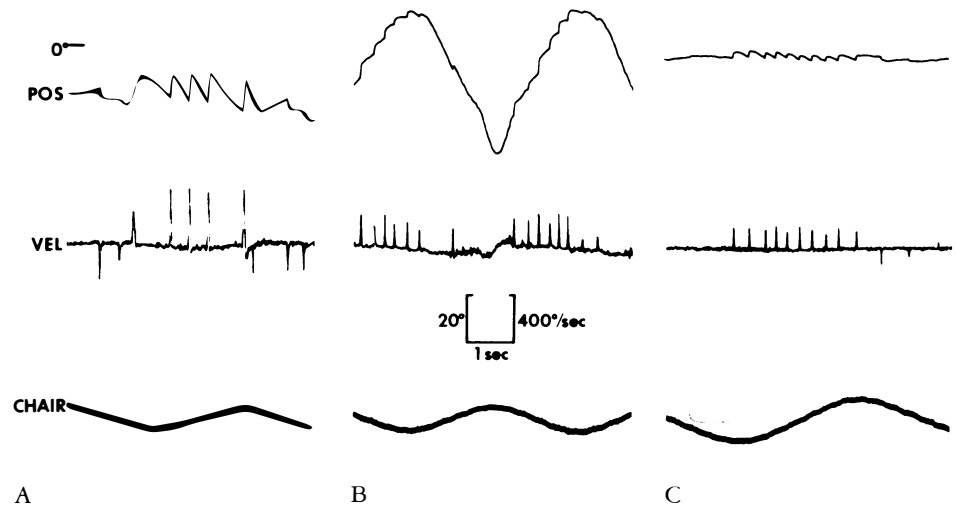


Fig 5. (A) Vestibuloocular reflex (VOR) during sinusoidal rotation in darkness while performing mental arithmetic. (B) VOR during sinusoidal rotation while fixating on stationary subject. Note smooth movements in both directions, in contrast to pursuit responses. (C) VOR suppression while fixating on object turning with patient. Note suppressed VOR during leftward movements only.

is admittedly speculative, but the purpose of this report is not to establish a clinicopathological correlation but rather to describe a new mechanism for horizontal jerk nystagmus.

The prototype lesion for horizontal pursuit asymmetry is posterior cerebral hemispheric disease [2], in which pursuit is only impaired ipsilateral to the lesion but there is neither primary position nor gaze-evoked nystagmus. Even with total cerebral hemispherectomy, the ipsilateral pursuit defect is only a decreased gain and not abolition [10] as with cerebellar ablation. Either the unilateral pursuit defect must be total for nystagmus to develop or, alternatively, cerebral disease may not cause nystagmus when the brainstem and cerebellum are intact. Indeed, there are no examples of chronic sustained nystagmus secondary to cerebral hemispheric disease. Perhaps both explanations are valid in that the reported examples of pursuit-defect nystagmus in the vertical plane [1, 6, 14] had profound deficits with little or no pursuit in the direction of the nystagmus fast phases.

The observation that our patient's "normal" leftward pursuit was faster than the target was analogous to the findings of Zee et al [14] with upward pursuit in their patients with downbeat nystagmus. We did not study leftward pursuit in sufficient quantitative detail to warrant further discussion of this curious phenomenon.

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