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Deborah I. Friedman and Louis F. Dell'Ossino
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CDR may result from several factors: (1) it is clinically based (i.e., independent of psychometric test scores); (2) the six categories used for rating dementia severity are directly linked to validated clinical diagnostic criteria for AD; (3) it has high inter-rater reliability for physicians and nonphysicians; and (4) an expanded and more quantitative version of the scale can be achieved by summing the ratings in each of the six categories to provide the Sum of Boxes. The CDR has been modified slightly over the years, as experience permitted resolution of ambiguities. These refinements primarily have been to sharpen the distinction between specific severity levels within a category. The first revision separated Community Affairs box scores 2 and 3, eliminated the possibility that Home and Hobbies could be “well maintained” for box score 0.5, and removed vague modifiers (“if any” from box score 0.5 in Community Affairs and “occasional” from box score 1 in Personal Care). The next revision distinguished Orientation box scores 0 and 0.5; other changes were slightly reworded descriptions for Orientation box scores 1 and 2 and for Judgment and Problem Solving box score 1, deletion of the modifier “may still” from the box score 1 description of normal appearance in Community Affairs, and substitution of “slight” for “mild” or “only doubtful” to describe 0.5 impairment for Memory, Judgment and Problem Solving, and Community Affairs.

A new version of the CDR more appropriately uses information regarding performance of financial transactions for rating Judgment and Problem Solving rather than Community Affairs. The new version is presented here (Table) for interested readers, along with improved clinical scoring rules for the global CDR (material added to the original rules is shown in italics).

From the Departments of Neurology and Pathology (Neuropathology) and the Alzheimer’s Disease Research Center, Washington University School of Medicine, St. Louis, MO.

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Address correspondence and reprint requests to Dr. John C. Morris, Memory and Aging Project, Washington University School of Medicine, 660 South Euclid Avenue, P.O. Box 8111, St. Louis, MO 63110.

References


‘Reappearance’ of congenital nystagmus after minor head trauma

Deborah I. Friedman, MD, and Louis F. Dell’Osso, PhD

Congenital nystagmus (CN) is typically horizontal, conjugate, and is noticed at birth or in early infancy. Although the appearance of CN does not usually change much throughout life, fluctuations in the amplitude of the nystagmus are common. We report a patient with congenital nystagmus that was first noticed at age 6, “resolved” at age 7, and became clinically apparent again after a minor closed-head injury at age twelve.

Case report. A 12-year-old boy was evaluated for reappearance of nystagmus after a minor head injury. His parents first noticed a head-tilt when he was 2 to 3 years old. Ophthalmic evaluation at age 5 showed no clinical evidence of nystagmus. Pendular nystagmus with excellent visual acuity (20/20 OU) and normal fusional amplitudes were found at age six. CT of the brain was reportedly normal except for sinus disease. The nystagmus was no longer visible at age seven. In the interim, he was a honor student and active in sports. Two months before his examination, he was struck in the back of the head with a baseball. There was no loss of consciousness. He was brought to the hospital for evaluation. Nystagmus was noted by the ambulance driver, and it subsequently persisted. There were no other neurologic sequelae from the head injury.

Since the injury, the patient needed to position his head to see clearly, and had difficulty catching a baseball. His parents noticed an intermittent head-turn while he was doing close work.

The patient had asthma, but no other medical, ophthalmologic, or neurologic conditions. Several family members had strabismus; there was no known nystagmus in the family.

Neuro-ophthalmic examination revealed normal visual acuity, color vision (Hardy-Rand-Ritter plates), pupils, visual fields, and intraocular pressures. The extraocular movements were full. The eye movements were videotaped and reviewed. Horizontal pendular-appearing nystagmus was present in primary gaze. The amplitude was variable and, at times, no nystagmus was visible. In lateral horizontal gaze, the nystagmus had a jerk component, beating in the direction of gaze. No definite roll position was present in primary gaze. The amplitude of the nystagmus seemed greater at distance than at near, and there was poor generation of horizontal optokinetic nystagmus (OKN). “Inversion” OKN was not observed. There was no observable change in the nystagmus with monocular occlusion, and no head-tilt, head-nodding, or head oscillation was seen. The neuro-ophthalmologic examination was normal.

Eye movement recordings. Eye movements were recorded using the infrared reflection technique. Full-system bandwidth was direct current to 100 Hz. The predominant waveform, jerk nystagmus with extended foveation, allowed foveation periods from 50 to 400 msec. In the figure, A and B show the position and velocity records during 4 seconds of fixation on a 0.2° diameter light-emitting diode. Note the periods of extended foveation despite the frequent bias reversals. In the figure, C shows the phase planes of both eyes during this 4-second interval, also indicating well-developed foveation within the foveation window. The 0.5° window has been extended ±0.1° to allow for the 0.1° radius of the target. The foveating saccades from both directions bring the target within this effective foveation window of ±0.6° and ±4°/sec. Pendular waveforms were not observed during the recording. There was a broad neutral zone with frequent bias reversals within ±20° of primary gaze. Convergence had a minimal damping effect on the nystagmus. Smooth pursuit was normal at low velocities, but of low gain at velocities of 20°/sec and above. The amplitude of the nystagmus was variable throughout the record, and ceased completely with inattention.

There was no evidence of a superimposed acquired nystagmus.

Discussion. As is typical of persons with CN, our patient did not experience oscillopsia. His well-developed foveation periods allowed him to enjoy excellent visual acuity most of the time. High-acuity foveation periods require that the eye position and target position coincide with minimal retinal slip. During smooth pursuit, the ability to match eye position to target position during foveation periods declines with increasing target velocity. In our patient, low pursuit gain with pursuit movements of 20°/sec or more accounted for his intermittent blurred vision, especially when trying to fixate on targets moving rapidly across his visual field. Although his nystagmus was not apparent in infancy, the presence of excellent vision and lack of oscillopsia suggested that the nystagmus was not an acquired type; the recorded CN waveforms confirmed this.

Our patient also demonstrated several atypical clinical features of CN: his nystagmus was not noted until age 5, his waxing and waning course was unusual, and his nystagmus reemerged after minor head trauma. The amplitude of CN can vary with many factors, including head position, eye position, febrile illness, and anxiety. Nystagmus with typical CN waveforms has developed in adolescence or adulthood without any
Figure. Records of (A) position (pos) and (B) velocity (vel) of both the right (RE) and left (LE) eyes during 4 seconds of fixation on a 0.2° light-emitting diode (LED). The dashed lines indicate the extent of the fovea and extended fovea (i.e., the radius of the LED added). The phase planes of both eyes are shown in (C), along with the foveation window (±0.5° by ±4°/sec) and effective foveation window (±0.6° by ±4°/sec). These windows are shown by solid lines; because the phase plane completely filled the foveation window, making it difficult to illustrate their extent, these data have been removed from the windows for clarity. All foveating saccades from both directions bring the eyes into the foveation windows.
identifiable trauma or neurologic disease. Those few patients with late-emerging CN previously probably had small-amplitude CN that was not clinically apparent, but would have been detectable with ophthalmoscopic examination.

Despite its name, CN can appear in early infancy as well as at birth. Thus, the term "congenital" refers to a congenital pre-disposition for ocular motor instability rather than the exact time of manifestation. Attempting to rename CN to "infantile nystagmus" is counterproductive because it emphasizes time of onset rather than the underlying mechanism, and since time of onset can extend to adulthood, the term "infantile" is overly restrictive. In agreement with previous usage, we ally at time

Although our patient's nystagmus appeared to be pendular on clinical examination, eye movement recordings demonstrated only jerk nystagmus (usually with extended saccadic periods). The frequent bias reversals of unidirectional jerk waveforms can have the clinical appearance of a pendular oscillation. Since the clinical examination can be misleading, the diagnosis of pendular CN should be made cautiously when eye movement recordings are unavailable.

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From the Departments of Neurology and Ophthalmology (Dr. Friedman), State University of New York Health Science Center, Syracuse, NY; and the Ocular Motor Neurophysiology Lab, Veterans Administration Medical Center, and Department of Neurology (Dr. Dell'Oeso), Case Western Reserve University, Cleveland, OH.

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Address correspondence and reprint requests to Dr. Deborah I. Friedman, Department of Neurology, SUNY Health Science Center, 750 East Adams Street, Syracuse, NY 13210.

References


Episodic changes in T-cell frequencies to myelin basic protein in patients with multiple sclerosis

A.A. Vandenbark, PhD; D.N. Bourdette, MD; R. Whitham, MD; Y.K. Chou, PhD; G.A. Hashim, PhD; and H. Offer, DSc

Although T-cell response to myelin antigens may contribute to inflammation and demyelination in MS, there are many discrepant findings in the literature. A possible explanation lies in episodic responses that we detected by longitudinal evaluations of T-cell frequencies, which we now report.

Methods and results. Using the limiting-dilution assay over a period of 17 months, we estimated the frequency of peripheral T cells specific for myelin basic protein (BP) and other antigens in 12 patients with progressive MS. These patients included nine men and three women with an average age of 52 years, average duration of MS 18 years, and average disability 6.0 Kurtzke Disability Status Score (KDDS) units. Data from one to four assays per month per patient were pooled and analyzed by chi-square minimization to determine the mean frequency ±95% confidence interval for each month. Frequency estimates obtained within 1 week after symptomatic treatment with corticosteroids were diminished and were not included in the results, but estimates obtained within 2 weeks returned to previous levels and were included.

The composite estimated frequency ±SE shown in the figure demonstrates several striking features. For the period of January 1991 to May 1991, the mean frequency remained at a baseline level between 1 and 2 BP-specific T cells per million blood mononuclear cells. However, during the period of June 1991 to October 1991, there was a dramatic and significant rise in the BP-specific T-cell frequency in all of the patients, with the mean frequency being approximately 7 per million. Subsequently, the BP-specific T-cell frequency declined to baseline. The frequency at the peak of response was similar to that reported in our previous study (6.2 per million), in which blood samples from many of the same patients and others were obtained approximately 1 year before (August 1990 to October 1990). Individually, during the period of June 1991 to October 1991, 10 of the 12 patients reached a maximal frequency of 10 BP-specific T cells per million blood mononuclear cells, which approximates the frequency of BP-specific T cells measured by the same method in the blood of paralyzed rats with experimental allergic encephalomyelitis (EAE) (11 per million). In two patients, the maximal frequencies did not exceed five BP-specific T cells per million, raising speculation that in a subgroup of MS patients, like in some mouse strains with EAE, other myelin antigens such as proteolipid protein may be the predominant encephalitogen.

Simultaneous frequency analyses of herpes simplex virus (HSV)-specific T cells showed a nonepisodic pattern that