"Inverse Latent" Macro Square-Wave Jerks and Macro Saccadic Oscillations

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A patient recovering from an acute encephalopathy demonstrated several ocular motor disturbances reflecting cerebellar and brainstem dysfunction. Two of these, macro square-wave jerks and macro saccadic oscillations, have never been reported in the same patient. The macro square-wave jerks disappeared with monocular viewing, a previously undescribed "inverse latent" characteristic.

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Various disturbances of eye movement result from lesions of the cerebellum and its connections within the brainstem. As summarized by Daroff [1, 2], these include skew deviation; nystagmus (gaze-evoked, upbeat, downbeat, positional, rapid horizontal bidirectional, rebound); ocular flutter; opsoclonus; ocular dysmetria; ocular myoclonus; asthenia of upward gaze; paresis of gaze and conjugate deviation; cogwheel (saccadic) pursuit movements; hypometric saccades; square-wave jerks; macro square-wave jerks; macro saccadic oscillations; and slow saccades. Macro square-wave jerks (MSWJs) and macro saccadic oscillations (MSOs) are two recently described signs [3, 4]. We report here the first patient documented to have both signs who, in addition, manifested a unique "inverse latent" characteristic to the MSWJ.

A 21-year-old white man developed occipital headache, intractable hiccups, mild right-sided numbness, and, shortly thereafter, horizontal double vision. Within two months the symptoms, except for diplopia, abated. The patient did not complain of oscillopsia. Neuroophthalmological examination at this time revealed a best corrected visual acuity of 20/20 in each eye and normal findings except for motility.

While fixing a distant light with both eyes open, he manifested large-amplitude conjugate horizontal oscillations that had the clinical appearance of MSWJs. When either eye was covered during distance fixation, the ocular oscillation stopped, but it resumed again with binocular viewing. Convergence also stopped the oscillation. On downward gaze he had intermittent downbeat nystagmus, most marked with the eyes deviated down and to the right. Optokinetic responses were diminished in all directions.

Quantitative heterophoria testing revealed a comitant 11 prism diopter esophoria at distance as well as vertical phorias indicating skew deviation. Neurological examination was otherwise normal, as was a CAT scan.

At this writing the patient is stable, and the future course will determine whether he has multiple sclerosis. This report is concerned primarily with the quantitative oculographic findings; the clinical details are being presented elsewhere [5].

Eye-Movement Recordings

Ocular motility was quantitatively recorded using infrared reflection and DC-coupled electronics with bandwidth of 100 Hz.

With the patient fixating binocularly at distance, leftward-going MSWJs were present which disappeared during convergence (Fig 1). Occasional square-wave jerks (SWJs) occurred during convergence. The velocities of adducting saccades were consistently slower than those of abduction bilaterally. Monocular viewing resulted in cessation of the MSWJs, but small-amplitude SWJs were present (Fig 2). The only exception was that the covered right eye had right-beating nystagmus in synchrony with the SWJ of the viewing left eye. Binocular viewing caused a return of the binocular MSWJ to the left.

Occasionally, bursts of saccadic oscillations occurred which increased and then decreased in amplitude while straddling a target (Fig 3). These conformed to the definition of macro saccadic oscillations. Figure 3 also shows MSWJs to the left of a target located 10 degrees to the right of midline, abduction overshoot, and slight adduction undershoot which, along with the consistently lower velocities of all adduction saccades, indicated bilateral internuclear ophthalmopareses.

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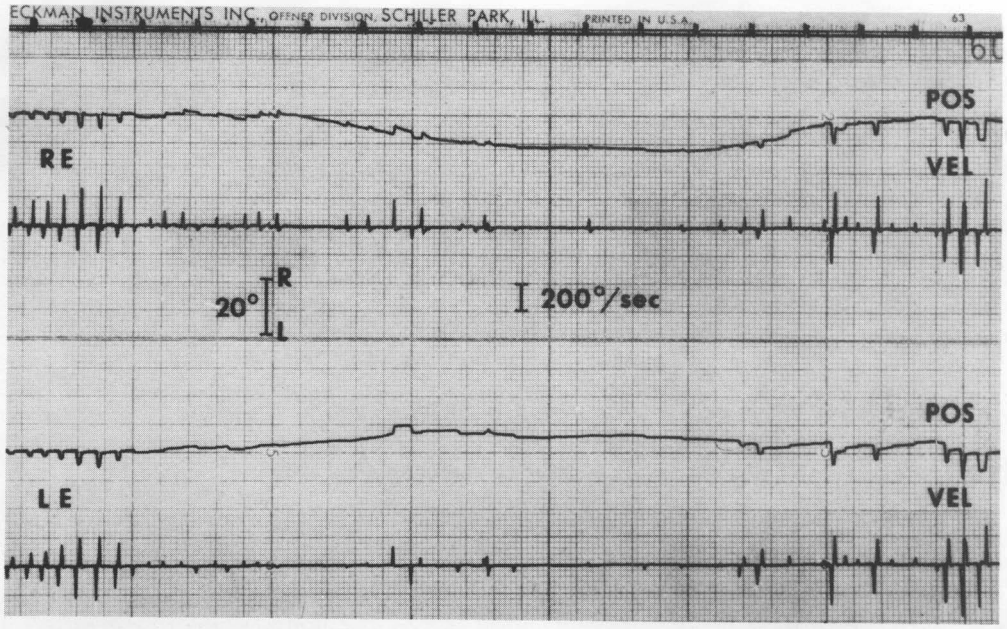

Fig 1. Binocular position (POS) *and velocity* (VEL) *recordings of macro square-wave jerks to the left of target with the patient fixating binocularly at distance. They disappeared upon convergence and reappeared with divergence. (Timing marks at the top indicate 1-second intervals in this and subsequent figures.)*

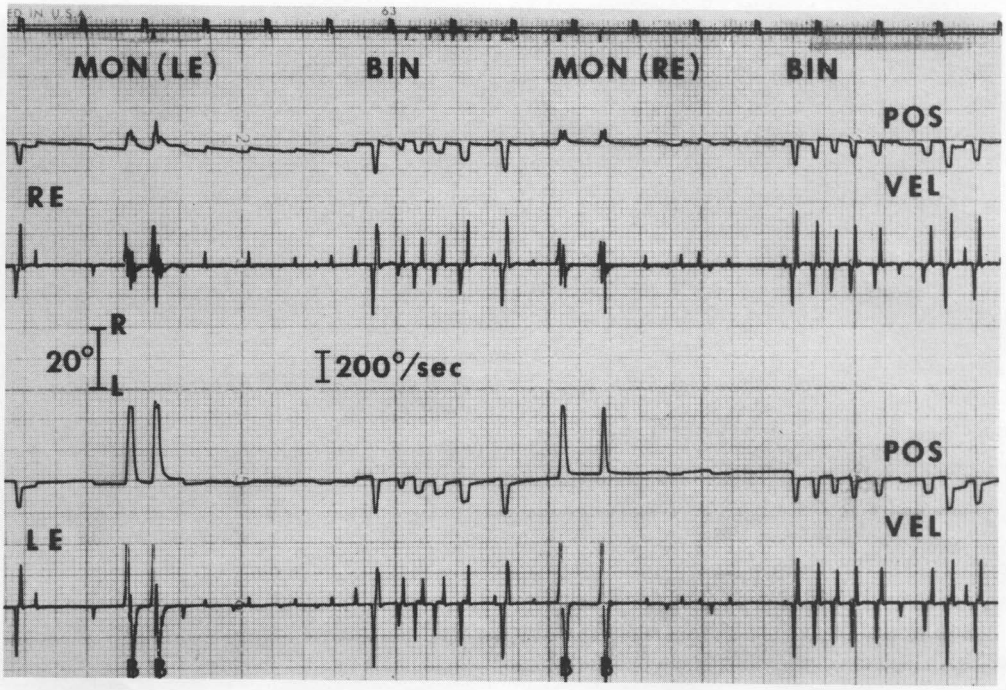

Fig 2. Binocular position (POS) *and velocity* (VEL) *recordings of macro square-wave jerks present with binocular* (BIN) *distance fixation but absent with monocular left eye* [MON(LE)] *and right eye* [MON(RE)] *fixation. Blinks are labeled* B. *With fixation, the left eye developed small-amplitude square-wave jerks in synchrony with right-beating nystagmus in the right eye, which had deviated into an esophoric position under cover. Bilateral small-amplitude square-wave jerks occurred with right eye fixation. Adduction saccades were consistently slower.*

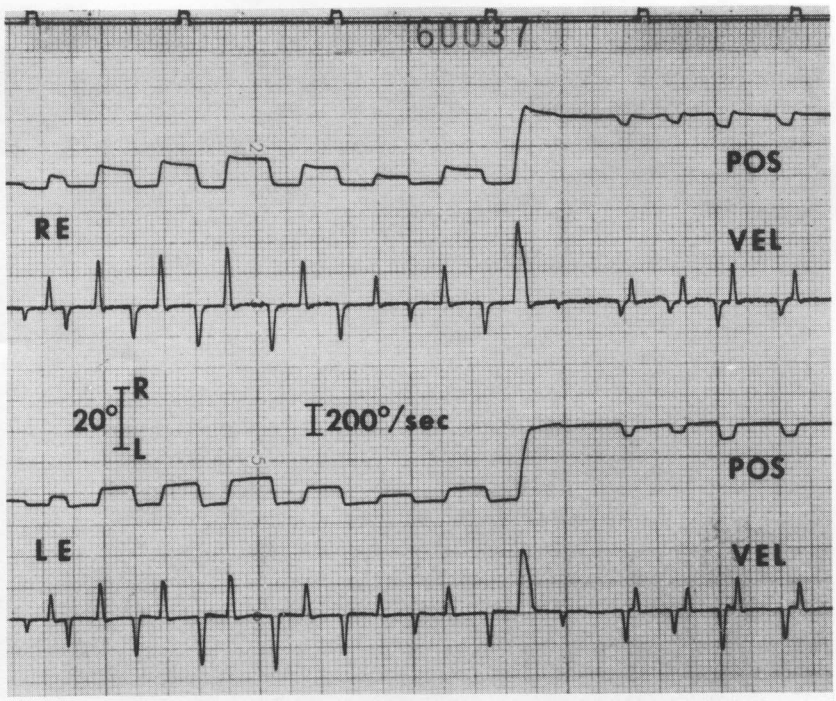

Fig 3. Binocular position (POS) *and velocity* (VEL) *recordings of macro saccadic oscillations with the patient fixating 10 degrees to his left, macro square-wave jerks while fixating 10 degrees to his right, and bilateral internuclear ophthalmopareses (the latter were inapparent clinically but are revealed by the higher velocities of abducting saccades compared to those of adduction).*

Discussion

The patient demonstrated a number of eye-movement problems indicative of brainstem or cerebellar dysfunction: skew deviation, gaze-evoked downbeating nystagmus, bilateral internuclear ophthalmopareses, SWJs, MSWJs with "inverse latent" characteristics, and MSOs. The possible nosological confusion concerning MSWJ and MSO and the unique monocular suppression of the MSWJ prompts limitation of our discussion to these entities.

Although SWJ, MSWJ, and MSO are all "cerebellar" eye signs by Daroff's categorization [1, 2], the difficulty of distinguishing pure cerebellar lesions from those of cerebellar pathways in the brainstem and the frequent coexistence of brainstem and cerebellar dysfunction [1, 2, 6] prompt the designation *cerebellar system*. SWJ and MSWJ have been discussed in the English-language literature only recently [3].

Square-wave jerks (Gegenrücke) are usually small-amplitude (½ to 3 degrees), conjugate saccadic eye movements that move the eyes away from fixation. After a latent period of approximately 200 msec (the visual reaction time), the eyes return to the target. SWJs have a maximum frequency of approximately 2 Hz. They can occur in normal persons with closed lids but, if present with eyes open during fixation, represent a pathological eye sign that is suggestive [6, 7] but not diagnostic [8, 9] of cerebellar disease. SWJs are a subtle disturbance easily missed clinically but obvious with eye-movement recordings.

Macro square-wave jerks [3] are usually larger-amplitude SWJs which are fixation dependent and also have a frequency of approximately 2 Hz. Both eyes suddenly and conjugately move off target with a saccade. After a latent period of approximately 80 msec a non-visually-evoked reflex saccade brings them back on target. The MSWJs in our patient might be confused clinically with a slow ocular flutter [10], but since the oscillations are flat topped (square), the recordings made the distinction.

Our patient showed classic MSWJs that were conjugate, 2 to 3 Hz, and of approximately 4 to 10 degrees amplitude with a 50 to 150 msec latent period between the initiating (leftward) saccade and refixation (rightward) saccade. The MSWJs manifested a unique attribute that has not been previously reported: present with binocular fixation at distance, they stopped when either eye was covered. Since *latent nystagmus* is the accepted term for a nystagmus absent with binocular fixation and present only when one eye is covered [11], the phrase *inverse latent* seems appropriate in our patient, as it emphasizes the feature of suppression by monocular fixation.

The patient also had MSOs, described by Selhorst et al [4] in 4 patients with cerebellar disease. Unlike MSWJs, MSOs increase and damp in amplitude, by-

Characteristics of Saccadic Instabilities

Variable	SWJ	MSWJ	MSO
Amplitude (degrees)	1/2–3,[a] constant	4–30, variable	1–30, increasing then decreasing
Time course	Sporadic	Bursts	Bursts
Latency (msec)	200	50–150	200
Foveation	Yes	Yes	No
Presence in darkness	Yes	Yes	No

[a]Occasionally can be up to 10 degrees.

SWJ = square-wave jerk; MSWJ = macro square-wave jerk; MSO = macro saccadic oscillation.

pass the fixation point with each saccade, and are not present in darkness. Another difference between the wave forms is the shorter latency of the return saccades in the MSWJ. Our patient's MSWJs and MSOs point out the lower end of their amplitude ranges. Salient features of SWJ, MSWJ, and MSO are listed in the Table. As is apparent, amplitude alone cannot invariably distinguish MSO from MSWJ; the waveforms are distinctive, however.

The other eye-movement abnormalities manifested by our patient warrant brief discussion. Skew deviation reflects brainstem or cerebellar disease and cannot be further localized [12]. Clinically the bilateral internuclear ophthalmopareses were masked by the saccadic instabilities and were revealed only during eye-movement recordings. Of note is the previous report of a patient with multiple sclerosis who also had both MSWJs and bilateral internuclear ophthalmopareses [3]. Downbeating nystagmus in the primary position suggests dysfunction at the level of the cervicomedullary junction [2] but probably can occur with parenchymal cerebellar disease [13]. Gaze-evoked downbeat nystagmus is probably less specific.

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