

Superior Oblique Myokymia

Quantitative Characteristics of the Eye Movements in Three Patients

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• **Using the magnetic search coil technique, we measured horizontal, vertical, and torsional rotations of both eyes of two patients with idiopathic superior oblique myokymia, and of the affected eye in a third patient. Superior oblique myokymia was strictly monocular and consisted of an initial intorsion and depression of the affected eye and subsequent oscillations with torsional and vertical components. The peak-to-peak torsional and vertical amplitudes of the oscillations were less than 1°, but peak velocities frequently exceeded 4°/sec in both planes. Fourier analysis indicated two features: (1) a broad range of frequencies up to about 50 Hz, indicating irregular oscillations; and (2) a superimposed larger-amplitude oscillation in the range from 1.5 to 6 Hz. Taken with electromyographic data from other studies, these results indicate that superior oblique myokymia reflects spontaneous discharge of trochlear motor neurons that have undergone regenerative changes.**

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Superior oblique myokymia (SOM) was first described in 1906,¹ but clinicians became generally aware of the disorder following the description by Hoyt and Keane in 1970.² Typical symptoms include monocular blurring

of vision or tremorous sensations in the eye.^{2,4} Patients will variably admit to vertical or torsional diplopia and vertical or torsional oscillopsia. Attacks usually last less than 10 seconds, but may occur many times per day. The attacks may be brought on by looking downward, by tilting the head toward the side of the affected eye, and by blinking.

The eye movements of SOM are often difficult to appreciate on gross examination, although they are usually apparent during examination with the ophthalmoscope or slit lamp. They consist of spasms of cyclotorsional and vertical movements. Measurements of the movements of SOM using the magnetic search coil technique^{5,6} have indicated an initial intorsion and depression of the affected eye, followed by irregular oscillations of small amplitude. The frequency of these oscillations varies; some resemble jerk nystagmus at frequencies of 2 to 6 Hz, but superimposed on these oscillations are high-frequency movements that have not been systematically characterized. The majority of patients with SOM have no underlying disease, although cases have been reported following trochlear nerve palsy, head injury, and possible demyelination or brain stem stroke and with cerebellar tumor.^{2,7}

We report herein the quantitative characteristics of SOM in three patients and relate certain features, especially the frequency of their oscillations, to the results of electromyographic studies and the possible etiology of this condition.

REPORT OF CASES

CASE 1.—A 60-year-old woman complained of constant "fluttering" of her right eye of 3 years' duration that was severe

enough to impair her vision prior to her examination in February 1990. She estimated that symptoms occurred at least 100 times per day, with a duration from 1 second to 1 minute. Aside from a history of "sarcoidosis," diagnosed with skin biopsy 10 years previously, but without other systemic symptoms, she was in good general and ocular health, except for mild anxiety, for which she took 0.5 mg of alprazolam three times per day.

Slit-lamp examination showed the classic intorsional movements of SOM. No vertical phoria was found. The patient received 20 mg of propranolol hydrochloride twice per day. This medication had no effect on her eye movement symptoms, but she said it "slowed her down too much," and therefore was discontinued. Timolol maleate eye drops (0.5%) were administered twice per day to the right eye, but they also had no effect and were discontinued. Three months later, the eye movements spontaneously resolved, but recurred after a further 6 months.

CASE 2.—A 69-year-old woman developed the sudden onset of a "vibration" affecting the left eye on February 25, 1990. It occurred many times each day, and seemed worse when reading and later in the evening. Her medical history was positive for fever and "inflammation of the brain" at age 18 years, for which she was hospitalized for 3 weeks. The patient had had a mastectomy for breast cancer at age 36 years, with subsequent surgeries at ages 52 and 62 years for local recurrences. Twelve years before examination, she suffered a myocardial infarction and has had mild congestive heart failure and angina since. At age 42 years, she was in an automobile accident and hit the left side of her forehead, resulting in loss of consciousness for about 1 hour. The patient never experienced diplopia. Treatment included diltiazem hydrochloride, digoxin, tamoxifen citrate, furosemide, and potassium chloride.

On slit-lamp examination, movements typical of SOM of the right eye were noted. The patient developed vertical diplopia on extreme right gaze and on left head tilt. In

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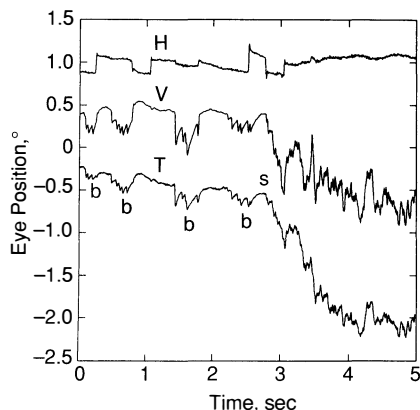


Fig 1.—Record of typical attack of superior oblique myokymia (SOM) from the right eye of patient 1. Horizontal (H), vertical (V), and torsional (T) gaze records are shown. Upward deflections indicate rightward, upward, and clockwise rotations from the point of view of the subject. The starting positions of H, V, and T have been offset for clarity of display. The patient blinked (b) four times, and, starting at about 3 seconds (s), an attack of SOM was induced, consisting of depression and intorsion of the eye with superimposed high-frequency oscillations.

primary position, there was a left hyperphoria of 2 prism diopters (D) that changed to 4 D of left hypertropia on right gaze and 3 D of left hypertropia on left head tilt.

Because of the history of breast cancer, magnetic resonance imaging was performed, and it showed mild, generalized, cerebral atrophy and enlargement of the diploic space of the skull; no metastases or lesions along the course of the fourth cranial nerve were identified. Skull roentgenograms and a bone scan showed no metastatic disease.

The patient was treated with timolol maleate eye drops and had some improvement, but not resolution, of her symptoms. Because of mild shortness of breath, therapy was switched to betaxolol hydrochloride eye drops (a β -1-selective agent) twice per day beginning April 30, 1990. By mid-July, the patient's symptoms disappeared completely and the betaxolol therapy was discontinued. As of August 24, 1990, she continued to be asymptomatic.

CASE 3.—A 50-year-old woman had intermittent monocular vertical oscillopsia affecting the left eye that began 3 months before her examination in February 1988. She denied having diplopia or other neurologic, ocular, or systemic symptoms or diseases except for mild osteoarthritis. There was no history of head trauma. Drinking coffee or smoking had no effect on her ocular symptoms.

The results of the neuro-ophthalmologic examination were normal except for episodic intorsional movements of the left eye seen during slit-lamp examination. There was a 1-D left hyperphoria in primary position measured with the Maddox rod, but the results of a Bielschowsky head tilt test were negative. Baclofen (5 mg) three times

Stability of Gaze in Patients and Control Subjects*							
Patient No.	Eye	SD of Position,°			SD of Velocity,°/sec		
		TPOS	VPOS	HPOS	TVEL	VVEL	HVEL
1	Right†	0.12	0.16	0.07	3.79	4.91	2.65
	Left	0.07	0.06	0.06	1.83	1.63	2.32
2	Right	0.09	0.05	0.08	1.51	1.46	1.41
	Left†	0.28	0.14	0.11	3.88	2.70	1.72
3	Left†	0.07	0.06	0.06	1.86	1.13	1.23
Control subjects	Minimum‡	0.03	0.03	0.02	1.55	0.76	0.85
	Maximum‡	0.10	0.05	0.08	2.11	1.39	1.68

* TPOS indicates torsional gaze position; VPOS, vertical gaze position; HPOS, horizontal gaze position; TVEL, torsional gaze velocity; VVEL, vertical gaze velocity; and HVEL, horizontal gaze velocity.

† Eye affected by superior oblique myokymia.

‡ For control subjects, the minimum and maximum SD values from six eyes are given.

per day had no effect on her symptoms. When contacted for follow-up 2 years later, the patient's symptoms had resolved.

MATERIALS AND METHODS

Horizontal, vertical, and torsional rotations of both eyes (patients 1 and 2) or the left eye (patient 3) were recorded using 2-m magnetic field coils (CNC Engineering, Seattle, Wash) and search coils consisting of Silastic scleral annuli (Skalar, Delft, the Netherlands). Search coils were precalibrated by clamping them to a protractor device and measuring changes in voltage induced by known rotations in three planes. The system was 98.5% linear over the operating range of plus or minus 20° in all three planes, and, for the amplifier settings used, the SD of the noise of the system was less than 0.02°.

With their heads restrained, the patients attempted steady, binocular fixation of a laser spot projected onto a tangent screen; this fixation target subtended 0.3° at a viewing distance of 1.3 m. In addition, patients made horizontal and vertical saccades between fixed-target locations, pursued a small moving target at constant horizontal or vertical velocities of up to 20°/sec, and fixed on a small object held by the examiner at a distance of 20 cm. For patients 1 and 2, the eye coil from the nonaffected eye was removed during the session and taped to the patient's head so that the stability of gaze could be measured during active yaw, pitch, or roll movements of the head. Data were filtered (bandwidth, 0 to 90 Hz) prior to digitization at 200 Hz. Computer analysis was performed using interactive programs written in the ASYST language (Keithley Asyst, Rochester, NY).⁸ Epochs of 3 seconds or more of typical SOM were analyzed to determine the SD of position and velocity in each plane in each eye. A 512-point fast Fourier transform was performed on typical episodes of SOM from each patient. For patients 1 and 2, a fast Fourier transform was also carried out on corresponding time segments from the unaffected eye.

Measurements of the stability of horizontal, vertical, and torsional gaze were also measured from four male control subjects (age range, 26 to 44 years). Three of these control subjects had myopia and one had emmetropia; none wore their spectacle cor-

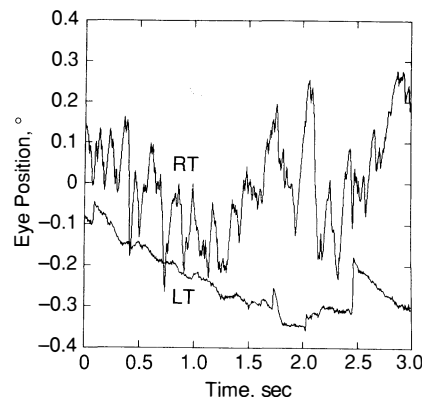


Fig 2.—A comparison of the torsional position of the right (RT) and left (LT) eyes of patient 1 during an attack of superior oblique myokymia. Upward deflections indicate clockwise rotations from the point of view of the subject. Note the high-frequency, small-amplitude oscillations of the right eye; these are absent from the left eye, which shows the typical drift and nystagmus encountered in normal subjects in the torsional plane.⁹

rections during measurements, but all easily fixed on the laser spot for 15-second intervals. Epochs of 3 seconds that were free of saccades and blinks were analyzed from each eye of each subject to provide a normative database.

RESULTS

In each patient, the onset of visual symptoms was synchronous with the onset of the recorded eye movement abnormalities. Patients were able to precipitate their visual symptoms by looking down and back up, by head tilt, or by blinking. In each patient, the onset of SOM was characterized by an intorsional and downward deviation of the affected eye; superimposed were oscillations that persisted for up to 10 seconds (Fig 1).

Measured amplitudes and velocities of these oscillations are summarized in the Table; control values obtained from recordings of the unaffected eye in

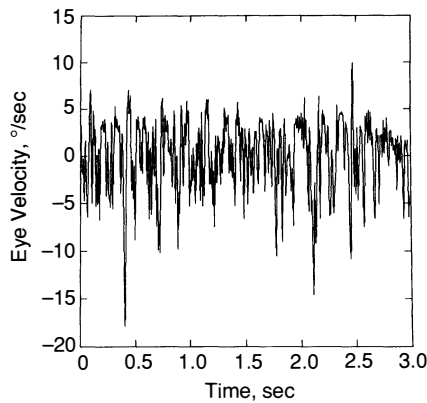


Fig 3.—A record of the velocity of torsional rotations of the right eye of patient 1 during an attack of superior oblique myokymia. Upward deflections indicate clockwise rotations from the point of view of the subject. Eye velocity often exceeded $4^\circ/\text{sec}$, especially intorsionally.

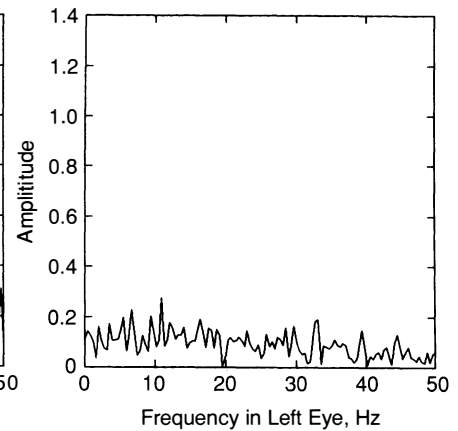
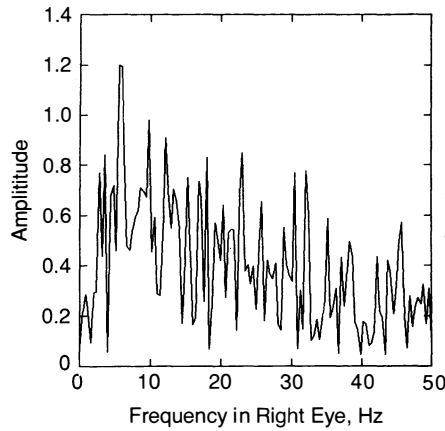


Fig 4.—Comparison of Fourier transforms of torsional movements of the right and left eyes of patient 1 using data recorded during an attack of superior oblique myokymia. The ordinate scales represent the relative amplitudes of the Fourier coefficients and are similar for both eyes. Note that the right eye shows a predominant frequency of 5.6 Hz, but that smaller component frequencies are evident up to 50 Hz. No comparable findings are present in data from the left eye.

patients 1 and 2 and from the control subjects are also included for comparison. Binocular recordings in patients 1 and 2 confirmed that SOM is a monocular phenomenon (Fig 2). The amplitude of the initial torsional deviation of the eye that heralded each attack was typically 1.5° , 1.5° , and 0.5° for patients 1, 2, and 3, respectively. The amplitude of the vertical component of the initial deviation tended to be smaller (Fig 1). The subsequent oscillations, superimposed on the deviation of the eye, were of small amplitude, less than 1° peak to peak. For patients 1 and 2, the SDs of the torsional and vertical deviations were only slightly greater than those of patients' unaffected eyes or values from control subjects. The SD of the amplitude of the torsional and vertical components during SOM in patient 3 was similar to that of the control subjects. No perturbation of the affected eyes in the horizontal plane was detected.

In patients 1 and 2, the velocity of SOM movements in the torsional and vertical planes (Fig 3) was higher than the corresponding values for the unaffected eye or measurements from the control subjects (Table 1). In patient 3, the SD of eye velocity was similar to that of normal subjects in the torsional and vertical planes; however, intorsional velocities ranged above $4^\circ/\text{sec}$. In all three patients, the velocity of intorsional eye rotations was greater than extorsional rotations.

Analysis of the frequency of the oscillations of SOM revealed two basic features. Each patient showed irregular, fine oscillations, ranging up to about 50 Hz, that were present

throughout the periods of SOM. In patients 1 and 2, superimposed jerk-like movements, directed intorsionally and downward, were superimposed (Figs 1 and 3). Figure 4 compares the relative amplitudes of the different frequencies of oscillation in the affected and unaffected eyes of patient 1 with the same episode of SOM shown in Figs 2 and 3. It is evident that although components of the oscillation are present up to 50 Hz, there is a peak at 5.6 Hz that corresponds to the frequency of the "jerk" movements. For patient 2, the corresponding "jerk" frequency was 1.5 Hz. Patient 3 showed no "jerk" movements, but only irregular fine oscillations. The results of Fourier analysis from the unaffected eyes of patients 1 and 2 were similar to the results from control subjects.

Measurement of saccades, smooth pursuit, and gaze stability during head rotations showed no abnormalities other than those due to the superimposed SOM.

COMMENT

The present measurements confirm that SOM is a paroxysmal monocular oscillation consisting of cyclotorsional and vertical movements. Although horizontal movements might be expected, given the tertiary (abducting) action of the superior oblique muscle, no such movements could be reliably identified.

Both the torsional and vertical components of SOM are of small amplitude. In comparing SOM with movements of the unaffected eye, it should be noted that the stability of torsional gaze in normal subjects is less precise

than that of the horizontal or vertical systems, as shown by our normal subjects and in the results of studies from other laboratories.* Thus, the SD of eye position in the torsional plane was only a fraction of a degree greater than that of the unaffected eye in patients 1 and 2, and the SD of the amplitude of the movements in patient 3 was similar to that of control subjects. The amplitudes of the vertical components of SOM were also quite small, although these were more likely to produce visual symptoms, such as diplopia or oscillopsia. Overall, it is not surprising that movements of SOM can be missed if not specifically looked for with slit-lamp examination or ophthalmoscopy.

On the other hand, the velocities of the torsional and vertical components of SOM in patients 1 and 2 were greater than those in normal subjects, reflecting the high-frequency components of these oscillations. Moreover, in patient 3, intorsional velocities exceeded $4^\circ/\text{sec}$. Thus, although the amplitude of these oscillations causes only a small displacement of the image of regard from the center of the fovea, the range of velocities of SOM, at least in the vertical plane, is outside the limits necessary for clear and stable vision (about $4^\circ/\text{sec}$). This excessive motion of images may cause oscillopsia.¹⁰

By performing a Fourier transform of the eye movements of SOM, we confirmed that two characteristics are often present: one is a low-amplitude, irregular oscillation with frequencies ranging up to 50 Hz, and the other is a large-amplitude "jerk" waveform with frequencies ranging from 1.5 to 6 Hz.

These findings have been commented on by other investigators, but not formally analyzed, to our knowledge.^{5,6}

How can the frequencies of the oscillations in SOM be related to the results of electromyographic studies of this condition? Electromyographic recordings from superior oblique muscles affected by SOM have revealed the presence of some fibers that either discharge spontaneously or persistently after contraction of the muscle.^{2,11,12} These muscle potentials were abnormal because they were of long duration (greater than 2 milliseconds) and increased amplitude and were polyphasic. Their rate of spontaneous discharge was approximately 45 Hz. Spontaneous unit activity was only silent with large saccades in the "off" (upward) direction, and was less affected by vestibular eye movements. Some units showed an irregular discharge following muscle contraction before subsiding to a regular discharge of 35 Hz. Simultaneous recordings from the inferior oblique muscle during episodes of SOM were normal. Taken together, evidence from electromyographic findings has been interpreted as indicating damage to neurons of the trochlear nerve, with subsequent regeneration of axons.^{2,11,12}

Experimental lesions of the trochlear nerve have demonstrated a considerable capacity for regeneration. It has been shown that if some of the trochlear motoneurons die after a nerve injury, the surviving motoneurons increase their number of axons to hold their number constant (in cat, at approximately 1000 axons per trochlear nerve).¹³ Such regeneration occurred only if neuronal cell death was less than 70% of the original population of trochlear motoneurons. Superior oblique myokymia has only rarely been reported to be preceded by trochlear nerve palsy.^{2,7} It is sometimes associated with other neurologic diseases.^{2,5}

Why is SOM associated with fourth nerve palsy so uncommonly if SOM reflects damage and regeneration of the fourth cranial nerve? One possibility is that *mild* damage to the trochlear nerve could trigger the mechanism for maintaining a constant number of axons in the nerve; regeneration of axons might lead to SOM. If SOM is due to axonal regeneration following injury (as the results of electromyographic studies suggest), then only those patients with incomplete injury would be expected to develop it. On the other hand, more severe injury to the fourth cranial nerve might prevent this repair

mechanism from taking effect.

Superior oblique myokymia shows properties that are dynamically distinct from those reported for ocular neuromyotonia, which is characterized by sustained contraction of the extraocular muscles following gaze deviation in individuals who have received radiation to the orbit and parasellar region.^{14,15}

Finally, there are no dependable treatments for SOM, although individual patients have been reported to respond to a number of drugs (ie, carbamazepine, baclofen, and β -blockers administered systemically or topically),^{3,16,17} and occasional patients have responded to surgery.¹⁸ We tried oral propranolol and topical timolol therapy in patient 1 without salutary effect. Patient 2 subjectively responded to topical betaxolol. The mechanism for improvement of SOM with β -blockers is unclear,¹⁷ especially in the case of timolol and betaxolol, which do not have membrane-stabilizing effects.

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