Superior Oblique Myokymia: Preliminary Results of Surgical Treatment

Earl A. Palmer, M.D.
William T. Shults, M.D.
Portland, Oregon

Introduction

Superior oblique myokymia is a peculiar intermittent monocular cyclovertical nystagmus of unknown pathogenesis and apparently unassociated with any other ocular or systemic abnormalities. It typically causes a prominent visual disturbance that may be described as diplopia oscillopsia, to distinguish it from the oscillopsia that is seen in ordinary concomitant nystagmus. Treatment with carbamazepine (Tegretol) is often effective. Clonazepam, phenytoin, and baclofen may also have a therapeutic role in some cases. In some patients, however, medical treatment is ineffective or only partially effective. This fact, combined with the particularly annoying symptoms produced by this condition, has resulted in attempts to find a surgical treatment. Encouraged by brief reports of successful surgical treatment and other anecdotal favorable clinical experience (Personal Communication with D. Tucker, April 7, 1983), we carried out surgical treatment for three patients whose symptoms were unsatisfactorily controlled by medication.

Case Reports

PATIENT #1

A 29-year-old Caucasian policeman was referred for further evaluation of left superior oblique myokymia. He had been previously evaluated by a neurologist and had complained of a three-year history of “double vision” affected by head movements. He also gave a history of chronic headache. Propranolol was tried without effect. The patient stated that he had been having symptoms more than 50% of the time, although there had been intervals as long as two weeks during which he was free of symptoms. Initially, whenever his visual “twitching” developed, it caused nausea, but this symptom abated. Because of a 4 prism diopter esophoria, greater on gaze right, he had been given a 2 diopter base-out prism before the left eye, with –0.75 lenses.

General ocular examination was normal except for an early pterygium in the left eye. His uncorrected visual acuity was OD 20/15 –1 and OS 20/20 –1; stereoaucuity: 40 seconds of arc; and ocular deviation: 5 prism dipters intermittent left hypotropia associated with 2 prism dipters of esophoria. Versions were comitant (Figure 1). There was no torsion by double Maddox rod. The left hypotropia disappeared completely whenever myokymic twitching of the left eye would stop. During episodes of oscillopsia, he described incyclotorsion. After a complete blood count was obtained, carbamazepine 200 mg twice daily was prescribed. The only additional concurrent self-medications were multivitamins and Clonulor 200 mg twice daily for an old knee injury.

Carbamazepine caused a reduction in symptoms. After two weeks, the dose was doubled without further improvement, and over an additional two weeks, the symptoms gradually returned to nearly the degree of severity present prior to starting medication. The patient felt that he could not continue this medication on the job, and that the symptoms were disturbing while driving a patrol car. It was suggested that the patient try baclofen, but he declined and requested surgery, despite its unproven nature. The risks and uncertain benefits were fully discussed. Three and a half months after his first visit, the patient underwent an intrasheath superior oblique tenotomy and left inferior oblique myectomy under general anesthesia. For six weeks he was completely free of symptoms, except for diplopia on gaze up and right, consistent with tightness of the left superior oblique, and was pleased with the outcome. Then, six weeks following surgery, he noticed the gradual recurrence of myokymic oscillopsia. There was persistent limitation of elevation in adduction, but no vertical deviation in primary, right, left or downward gaze. On upward gaze there was a 12 prism diopter right hypertropia. No torsion was
present by double Maddox rod testing. Two and a half months following initial surgery, a left superior oblique tenectomy with removal of the sheath was performed. 

At his most recent evaluation, three and a half months following the second operation, there had been no recurrence of myokymia symptoms. He did have persistent diplopia on far downward gaze, especially bothersome on first arising in the morning, lasting about one hour. He has an 8 prism diopter left hypertropia on far downward gaze and 7 prism diopters left hypertropia on right gaze. On upward gaze there is no vertical deviation (Figure 2). Diplopia is absent in the usual directions of gaze, although the patient feels a “lag” on horizontal saccades. Double Maddox rod testing shows no torsional deviation. He particularly noted a reduced frequency of headaches and a reduced feeling of “tightness” in the eye except on downward gaze. The only inconvenient symptom occurs while lying down to watch a television set that is positioned toward the foot of his couch. He is quite happy with the outcome of surgery (Figure 3).

**PATIENT #2**

This 31-year-old male was seen with a ten-year history of diplopic oscillopsia. Symptoms had been intermittent during the first year, and the condition spontaneously remitted for three to four years without recurrence. Recurrence was progressive, finally resulting in more than one year of daily symptoms. The patient had discovered that he could correct oscillopsia by elevating or depressing his head and also found that it could be triggered by certain directions of gaze. He gave a prior history of having seen a neurosurgeon and having had a negative arteriogram and CT scan. He was subsequently seen by one of us, who diagnosed superior
oblique myokymia. Carbamazepine was started at 100 mg twice daily, and later increased to 200 mg three times daily, but significant improvement did not occur.

There was a very fine torsional quivering of the right eye due to a small amplitude intorsional jerk nystagmus. An intermittent right hypertropia of 3 to 4 prism dipters could be measured in the primary position and on left gaze. The uncertain possibilities of surgical treatment were discussed with the patient, who elected to have another examination by a second neuro-ophthalmologist, who confirmed the diagnosis. Consideration of increasing the dose of carbamazepine or adding Clonopin was considered unrealistic, due to the patient's occupation. Therefore, a combined right superior oblique tenotomy and right inferior oblique myectomy were performed under general anesthesia.

For five days following surgery there had been no recurrence of oscillopsia, and the patient was orthophoric in primary position. He complained of persistent mild nausea and dizziness, which he compared to motion sickness. Diplopia on upward and downward gaze were present, with 3 prism dipters of right hypertropia on upward gaze and 5 prism dipters of right hypertropia on downward gaze. He remained orthophoric on direct right gaze, but exhibited a 4 prism dipter right hypertropia on left gaze. Double Maddox rod testing indicated no torsion. At near, there was a 4 prism dipter exophoria. Under normal conditions, he noticed diplopia only in the upper left quadrant of gaze.

Eleven days following surgery he experienced a recurrence of diplopic oscillopsia, which developed into a daily experience lasting 30 to 60 minutes.

Six weeks following surgery, he had a persistent 5 prism dipter right hypertropia on upward gaze and 2 prism dipter right hypertropia on downward gaze (Figure 4). This did not change on right or left head tilt. Versions indicated underac-
tion in the field of the right inferior oblique and resembled a Brown syndrome. He estimated that his overall symptoms were approximately 50% as bothersome as they were preoperatively. Even though the amplitude of his oscillopsia was reduced, he felt that the diplopia in gaze up and left was a sufficient problem to offset the overall benefit.

PATIENT #3

This 45-year-old woman was treated with carbamazepine for one year with partial control of symptoms of right superior oblique myokymia. Her diagnosis was made by one of us after a previous CT scan and normal neurologic consultation. Carbamazepine resulted in dampening the movement but not eliminating it. She believed that her symptoms were gradually worsening and was therefore considered for surgical treatment. She had first noticed intermittent quivering of vision about three and a half years earlier. Once it had begun, it soon became constant. Her oscillopsia and diplopia were most bothersome during automobile travel and, indeed, she compared her symptoms to the visual experience of looking in a rear view mirror on a bumpy road. Occasionally, the oscillopsia would stop and diplopia would persist for a time. She had found that by flexing her neck (producing upward gaze) she could reduce the amplitude of oscillopsia. With her eyes shut, she was unaware of any subjective movement. About six months prior to onset of the syndrome, she had bumped her head on a rain gutter and fallen to the ground, without apparent loss of consciousness. Interestingly, she felt that her symptoms were worse when she was "low on protein" and therefore carried nuts with her, to restore her protein level whenever necessary. There was no previous history of eye disease.

The right eye exhibited a rhythmic nystagmus with the rapid phase downward and towards intorsion. This was of larger amplitude than the typically described case, being grossly visible from two to three feet away. Although visual acuity without glasses was 20/25 in each eye, the right eye acuity degraded to 20/30 when the "jiggling" was fully manifest. Approximately 2 prism dipters of right hypertropia could be measured at times. Stereoaucity was 40 seconds of arc. Manifest refraction was plano bilarteral. The pupils were slightly asymmetric, with the right measuring 6 mm and the left 5.5 mm. Double Maddox rod indicated 5° of right exodeviation. Versions were comitant. Corneal sensitivity was equal and perhaps slightly reduced, so that she easily inhibited a blink reflex. Ocular tension and fundus examination were normal. After a full discussion of the uncertainty of surgical treatment, she accepted the risks. A combined right superior oblique tenectomy and right inferior oblique myectomy were performed under general anesthesia. On the first postoperative day, she was free of diplopia in primary position and on right and left gaze. There was slight limitation of vertical movement with the right eye in adduction.

At six weeks following surgery, the patient reported a significant reduction in her diplopic oscillopsia, although it was still present to a slight degree. She described the persistent jiggling as being "like a heat wave." She felt that this was tolerable, and a substantial improvement over her previous condition. Her oscillopsia was aggravated by drinking tea or coffee.

Detailed examination at six weeks following surgery (Figure 5) indicated a 6 prism dipter intermittent right hypertropia that reduced to 2 prism dipters on right gaze and increased to 10 prism dipters on left gaze. On downward gaze this became 18 prism dipters of right hypertropia, and it reduced to zero on upward gaze. Hypertropia was greater on right tilt than on left tilt. Versions indicated underaction in the field of both right obliques, and weakness of depression of the right eye (Figure 6). Observing the movement of the right eye disclosed that the amplitude of jiggling was very small and far more difficult to see than it had been preoperatively.

Eleven weeks following surgery, she reported that her occurrences of visual "shimmery" had perhaps increased, and were now occurring around 60% of the time. She noted that looking left produced ocular jiggling, but felt that she could look increasingly farther to the left before experiencing diplopia. She continues to have diplopia when trying to read, but has chosen to wait before trying prism glasses. She notes diplopia on tilting her head toward the right or on looking down and left. Although she is disappointed that she did not have perfect relief of her symptom, she stated that she was glad to no longer be dependent on medications to maintain reasonable visual comfort.

Discussion

Previous reports of surgical treatment have been somewhat cursory, but sufficiently optimistic to encourage further surgical efforts for suitable patients. Hoyt and Keane reported that one of their patients, a 24-year-old woman, underwent an intrasheath superior oblique tenotomy and recession of the inferior oblique, had postoperative diplopia only in the extremes of vertical gaze at two months following surgery, and six months later was asymptomatic.1 Susac and colleagues reported excellent results in one case treated with superior oblique tenotomy combined with inferior oblique recession.2 Kommerell and Schaubele performed a superior oblique tenotomy, producing a palsy that required further surgery.5 In a review of superior oblique
myokymia, two additional surgical cases are noted, one successfully treated by superior oblique tenectomy combined with inferior oblique myectomy, and the other responding transiently to superior oblique tenectomy.⁴

Although Hoyt and Keane remarked that superior oblique myokymia may not be as rare as would be inferred from the paucity of reports in the literature,¹ it is undoubtedly still rather rarely encountered in primary care or in a secondary level referral ophthalmic practice. Many of these cases are evaluated by a neurologist along the way. Some patients choose not to see a physician since symptoms are mild; others see a physician who cannot explain the symptoms and conclude there is nothing to be done for it. Most recognized cases are probably not suitable for surgical consideration due to the self-limited character of the disease, a favorable response to medication, or relatively mild and tolerable symptoms. A neuro-ophthalmologist receiving referrals from related specialists will probably encounter this condition repeatedly.

Medical treatment with carbamazepine is often effective and yet not without a certain small risk of aplastic anemia. This risk may indicate blood counts as often as every week during the first three months of therapy and monthly thereafter for at least two to three years. Patients need to be made aware of this risk and report suspicious signs and symptoms to the physician. Because of unknown teratogenic potential, caution is urged in the use of carbamazepine for women of child-bearing age. In addition, there is a potential hazard in operating machinery, including an automobile, or performing other dangerous tasks.⁸ This arises, in part, from an occasional toxic ataxia syndrome, consisting of nystagmus, confusion, drowsiness, and ataxia. Ocular side effects can include reduced accommodation, and patients have experienced visual hallucinations. The drug may potentiate the effects of alcohol and barbiturates.⁹ Other reported side effects are dizziness, gastrointestinal disturbances, urinary distress, and allergic reactions.¹⁰ It should be noted that the benefit from, and the need for, routinely ordering extensive serial hematologic studies for patients taking carbamazepine has been seriously questioned.¹¹

None of the three patients in the present report wished to
continue taking carbamazepine, even though numerous side effects were neither especially emphasized to them nor experienced by them. Their decision is, of course, also partially due to the inadequate response to medication experienced by each of these patients.

Conclusion

The presently reported surgical case histories indicate that surgical operation for this condition is not always curative and has some undesired consequences. The only one of these three who is completely satisfied with the result underwent two procedures. It would seem realistic in view of the data currently available that if surgery is to be offered, it be recommended to selected patients who are advised that it will “probably” require two procedures and “possibly three or more” procedures.

Acknowledgments

These patients were referred by William N. Hawks, Jr., M.D., Chelsea, Michigan; Russell J. Leavitt, M.D., Grants Pass, Oregon; and Darrell R. Lumaco, M.D., Hillsboro, Oregon. Postoperative care of one patient was assisted by A. Richard Johnson, M.D., Yreka, California.

References