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Strabismus Surgery for Internuclear Ophthalmoplegia with Exotropia in Multiple Sclerosis

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Abstract

Background—Internuclear ophthalmoplegia (INO) is a disabling condition affecting up to 40% of patients with multiple sclerosis (MS). Management of bilateral internuclear ophthalmoplegia (BINO) with exotropia in MS has been controversial due to the uncertain course of MS. Diplopia associated with INO severely impacts the patient's quality of life, and therefore treatment should be considered.

Methods—Three patients (ages 28, 62, and 82 years) who had BINO with exotropia and disabling diplopia, secondary to MS, underwent bilateral medial rectus resections with either unilateral or bilateral adjustable lateral rectus recession(s). Alignment was measured pre- and post-operatively, and symptoms were recorded.

Results—Pre-operative exotropia ranged from 40 to 64 prism diopters (pd) for distance fixation and from 50 to 70 pd for near fixation. Pre-operatively all patients had diplopia at distance and near fixation. Immediately following surgery, patients were adjusted to 10 to 18 pd ET at distance fixation with the expectation of an exotropic drift. None of the patients had a tropia at distance and near fixation six months following surgery, with single vision in primary position and reading. Two patients had a two-year follow up examination; one required a small amount of base-in prism for comfortable reading.

Conclusions—Three patients who had BINO with exotropia secondary to MS all benefited from surgery. Surgery should be considered as an option for symptomatic patients who have BINO with exotropia due to MS.

Introduction

Multiple sclerosis (MS) is a disabling condition that affects around one million people worldwide and between 250,000–300,000 in the USA, with many patients experiencing their first symptom between the ages of 20 and 40 years (National Institute of Neurological Disorders and Stroke, National Institute of Health, Multiple Sclerosis: Hope through research. http://www.ninds.nih.gov/disorders/multiple_sclerosis/detail_multiple_sclerosis.htm#104853215, accessed June 7, 2008). Ocular manifestations are common, affecting up to 70% of all patients with MS at some point during their disease course,^{1, 2} including conditions such as optic neuritis, nystagmus, uveitis, and internuclear ophthalmoplegia (INO). Each of these

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ocular manifestations can have an impact on the patient's mobility, occupation, and overall quality of life.³ INO affects up to 41% of patients with MS,⁴ and when associated with an exotropia, patients often experience disabling diplopia, abnormal head postures, and poor appearance. Others have referred to Bilateral INO (BINO) with exotropia as "wall-eyed BINO" or WEBINO.

Reports of surgical intervention for BINO with exotropia in MS are few. Surgery seems to be rarely recommended because the natural history of a demyelinating condition is uncertain; therefore, surgical outcomes are thought to be unpredictable. Although Botulinum toxin injections and surgery have been reported as treatments to correct INO, the majority of these patients had INO secondary to vascular pathology.^{3, 5-8} It is possible that INO secondary to MS may behave differently from INO secondary to vascular causes, and it seems that surgeons are more reluctant to operate on patients with MS, due to uncertainty of clinical course. Murthy et al report four patients with INO secondary to MS treated with botulinum toxin, but, at their last reported examination, two patients were managed with occlusion, one was receiving maintenance injections, and one went on to receive surgery.⁵

In the present study, we present the results from surgical intervention in three patients who had BINO with exotropia secondary to MS.

Subjects and Methods

Institutional Review Board approval was obtained for this retrospective chart review study. The surgical database of one of the authors (JMH) yielded three patients with a diagnosis of BINO secondary to MS that had undergone surgery.

Patients

Three female patients ages 28, 62, and 82 years had secondary progressive MS for 3 to 41 years prior to surgery. All three patients had slowing of adducting saccades and abducting nystagmus, confirming the diagnosis of INO. No associated skew deviation was found in any of the three patients. Each patient had disabling diplopia with large angle exotropia. Pre-operative angle of deviation with prism alternate and cover test (PACT) ranged from 40 to 64 prism diopters (pd) exotropia at distance fixation and 50 to 70 pd exotropia at near fixation.

Surgical Intervention

Each patient underwent bilateral medial rectus resection with either unilateral or bilateral adjustable lateral rectus recession. In each case, post-operative adjustment was performed later the same day.

Case One

Case one was 28 years old at the time of surgery and had secondary progressive MS associated with poor mobility due to lower limb weakness for three years. She had a one-year history of worsening diplopia and, when referred, had diplopia in all positions of gaze and was occluding one eye. Her pre-operative alignment by PACT was 64 prism diopters (pd) exotropia at distance fixation and 64 pd exotropia at near fixation. She underwent a bilateral medial rectus resection of 7 mm and a bilateral lateral rectus recession of 7 mm on adjustable sutures. Following surgery, the immediate pre-adjustment alignment was 20 pd esotropia at distance fixation and 6 pd esotropia at near fixation. At adjustment, the lateral rectus muscle was advanced 2 mm, giving a final alignment of 14 pd esotropia for distance fixation and orthotropia for near fixation.

The distance esotropia resolved over three months, and the patient was essentially diplopia-free for 12 months. At 12 months after surgery, the patient had a well controlled intermittent exotropia of 6 pd at distance fixation and 14pd at near fixation. At 18 months after surgery, mild symptoms of intermittent diplopia recurred, and the patient was therefore fitted with a 10 pd base-in prism. She remained stable in this small amount of prism for seven years, but then developed a slow recurrence of the BINO and exotropia with increasing diplopia and underwent further surgery.

At the time of the second surgery, seven years after the first surgery, pre-operative measurements were 50 pd exotropia at near fixation and 30 pd exotropia at distance fixation with PACT. Bilateral medial rectus resection was performed. The left medial rectus was resected 8 mm on an adjustable suture, and the right medial rectus was resected 6 mm with 2 mm advancement on an adjustable suture, since the right medial rectus was found inserted 2 mm back from the original insertion. The immediate pre-adjustment post-operative alignment was 18 pd esotropia at distance fixation with orthotropia at near fixation. This moderate esotropia was felt to be desirable, and so the sutures were tied. Three months following this second surgery, she was orthophoric for distance and near fixation with no symptoms of diplopia. Further follow-up continues.

Case Two

Case two was 62 years old at time of surgery, and had been diagnosed with secondary progressive MS for 19 years. She had been seen previously with other ocular manifestations of MS, including a left optic neuritis, left homonymous hemianopia, and a mild posterior uveitis. She had been having problems with intermittent diplopia since initial diagnosis, and was diagnosed with BINO eight years prior to presentation. Pre-operative alignment with PACT measured 50 pd exotropia at near fixation and 40 pd exotropia at distance fixation.

Surgery was performed, consisting of bilateral medial rectus resections of 6.5 mm and an 8 mm recession of the left lateral rectus on an adjustable suture. Following suture adjustment, she had 10 pd esotropia at distance fixation and 2 pd exophoria for near fixation. The moderate distance esotropia was planned, expecting an exotropic drift.

The distance esotropia resolved over three months, and the patient had marked improvement of her symptoms of diplopia at both distance and near fixation. Twelve months following surgery, the patient was asymptomatic with a 12 pd exophoria for distance fixation and orthophoria for near fixation. At two years follow up this patient was still asymptomatic, with no diplopia, and measured 8 pd exophoria for distance fixation and 4 pd of exophoria for near fixation.

Case Three

Case three was 82 years old at the time of surgery, and had been diagnosed with MS 41 years earlier. She had secondary progressive MS associated with severe mobility problems which had left her wheelchair bound. She reported intermittent wandering of her eyes for 10 years, with intermittent diplopia for distance and near fixation. Pre-operative alignment with PACT measured 50 pd XT at distance fixation and 70 pd at near fixation.

Bilateral medial rectus resection of 7 mm with a left lateral rectus recession of 8 mm on an adjustable suture was performed. Pre-adjustment immediate post-operative alignment was 25 pd esotropia at distance fixation and 14 pd esotropia at near fixation. The left lateral rectus was advanced 4 mm, leaving a post-adjustment alignment of 14 pd esotropia at distance fixation and 4 pd esotropia at near fixation.

The planned immediate post-operative esotropia resolved over the 6 weeks following surgery, and the patient became diplopia free at near fixation. At six months post-operatively, the patient remained asymptomatic. Measurements at this time were 3 pd esotropia at distance fixation and 2 pd exophoria at near fixation. The small angle esotropia at distance was asymptomatic and associated with central suppression.

Discussion

In our small case series we have described three patients who had a marked improvement in alignment and function following surgery for BINO with exotropia, regaining the ability to read comfortably. One patient had two surgeries seven years apart, and had sustained good visual function during the seven years between surgeries, with the aide of a small amount of prism.

Multiple sclerosis is a multisystem disorder, and visual quality of life is often overlooked when treating motor and other sensory disability.⁹ Diplopia was the main complaint in our group of patients, and the initial referral from neurology to the ophthalmic service was for treatment options for this symptom. Previously, patients with BINO of vascular etiology have been reported to have improved convergence to some extent following surgical alignment,⁶ possibly by enabling utilization of neurologically-preserved convergence. Nevertheless, patients who have BINO with exotropia may have compromised convergence.¹⁰ All of the patients in our series had a large angle exotropia which they were unable to comfortably overcome by convergence. By surgically reducing the exotropia, we subsequently reduced the amount of convergence effort needed to fuse, allowing each patient to read comfortably. Our approach to BINO with exotropia was similar to that reported recently by Roper-Hall et al,⁸ but they performed mainly unilateral resect-recess procedures, and, when bilateral, only performed lateral rectus recessions, expecting an undercorrection in some patients and anticipating a second surgery, where we describe an approach designed to be effective in one procedure. Roper-Hall et al also reserved their surgery for patients with vascular and traumatic causes, where we address BINO with exotropia in patients specifically with MS.

All of our patients had surgery using adjustable sutures, which enabled fine tuning of the alignment in the immediate post-operative period. Our target angle was 10 to 18 pd esotropia in the distance, assuming that there would be a postoperative exotropic drift. Indeed, in all three patients the distance esotropia reduced to zero or a very small angle within three months. The larger value of 18 pd was chosen for the second surgery of Case One, due to the recurrence of the condition, albeit a slow recurrence. The optimum post adjustment alignment is not known, but in convergence insufficiency exotropia, where the near angle of exotropia is also greater than the distance angle, Choi et al¹¹ report good long term results when the patients are set with an initial post adjustment distance esotropia of 10 to 20 prism diopters following medial rectus resection. We therefore suggest using adjustable sutures to establish an initial moderate esotropic overcorrection in patients with BINO with exotropia.

Botulinum toxin has been used previously for BINO with exotropia in patients with multiple sclerosis,^{3, 5, 7} but the numbers are few and the long term outcomes in this group of patients were suboptimal with many patients, requiring maintenance injections, prisms, occlusion, or surgery.

Orthoptic treatment options for BINO with exotropia include the use of prisms, but this option is only suitable for patients with smaller deviations, and in our series the exotropic deviations were too large to be treated with prisms. Nevertheless, prism was used postoperatively in the first case one year following the initial surgery due to a slight recurrence of the exotropia. The

use of prism in this case maintained comfortable reading for several years, until the angle of deviation increased to the point of needing additional surgery.

We suggest that surgery should be considered earlier in patients who have binocular internuclear ophthalmoplegia with exotropia secondary to multiple sclerosis. In the past, surgery may have been avoided in this group of patients, but we have provided evidence of restoration of function and improvement of appearance following surgery for this condition. Surgery for binocular internuclear ophthalmoplegia with exotropia in multiple sclerosis can significantly improve quality of life by reducing symptoms of diplopia and enabling comfortable reading, and should be offered to many more patients with this condition.

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