Residual Symptoms After Surgery for Unilateral Congenital Superior Oblique Palsy

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ABSTRACT

Purpose: To establish the surgical results and residual symptoms in 48 cases with unilateral congenital superior oblique muscle palsy that had surgical intervention to the vertical muscles alone.

Methods: Myectomy and concomitant disinsertion of the inferior oblique (IO) muscle was performed in 38 cases and myectomy and concomitant IO disinsertion and recession of the superior rectus muscle in the ipsilateral eye was performed in 10 cases. The preoperative and postoperative vertical deviation values and surgical results were compared.

Results: Of the patients who had myectomy and concomitant IO disinsertion, 74% achieved an “excellent” result, 21% a “good” result, and 5% a “poor” result postoperatively. The difference in deviation between preoperative and postoperative values was statistically significant ($P < .001$). Of the patients who had myectomy and concomitant inferior oblique disinsertion and ipsilateral superior rectus recession, 50% achieved an “excellent” result, 20% a “good” result, and 30% a “poor” result postoperatively. The difference in deviation between preoperative and postoperative values was statistically significant ($P < .001$).

Conclusion: Both procedures are effective and successful in patients with superior oblique muscle palsy, but a secondary surgery may be required.

INTRODUCTION

Forty percent of superior oblique muscle palsy is congenital, although some cases may not be clinically evident in childhood.¹ The hypertropia of the paretic eye that increases on ipsilateral head tilt and decreases on contralateral head tilt is observed in patients with a unilateral superior oblique muscle palsy.² In addition, excyclotropia may develop in the affected eye.³ There is a typical compensatory head posture characterized by a tilt to the contralateral side in these patients.² The purpose of the compensatory head tilt is considered to be this decrease in the hypertropia by many authorities.⁴ Adoption of the abnormal head posture usually develops to place the eyes in the field of gaze to maintain binocular single vision. Long-standing abnormal head posture due to extraocular muscle paresis may cause scoliosis.⁵

Superior oblique muscle palsy may result in weaknesses ranging from mild to severe. The anatomic course and attachment of the superior oblique muscle may vary, as may the location and effect of the trochlea within the orbit. Because secondary changes may develop with time, it is difficult to determine whether there is a long-standing, generally congenital superior oblique muscle palsy that has
become decompensated and clinically evident recently or a more recently acquired superior oblique muscle palsy.\textsuperscript{6}

Vertical or torsional diplopia, significant abnormal head posture, or asthenopic symptoms are indications for surgery. The aim of the treatment is mainly to achieve fusion in primary position and down gaze and to eliminate an abnormal head posture.\textsuperscript{7} Weakening of the overacting ipsilateral inferior oblique muscle, including disinsertion, myotomy, recession, and denervation, is the most common treatment.\textsuperscript{8}

The aim of this study is to establish residual symptoms and surgical results of patients whose vertical muscles were operated on with the diagnosis of unilateral congenital superior oblique muscle palsy.

**PATIENTS AND METHODS**

This study includes 48 cases who were operated on with the diagnosis of unilateral congenital superior oblique muscle palsy at Dicle University Faculty of Medicine, Ophthalmology Department between May 2004 and April 2010. All of the patients had surgical intervention to the vertical muscles alone.

The patients were evaluated according to age, gender, etiology, and the affected eye. The visual acuity, abnormal head posture, amblyopia, fusion, stereopsis, and muscle functions were examined. Deviation values were determined according to alternate occlusion test with prism in near and far fixation. The Worth 4-dot test was used for fusion and the Titmus test was used for stereopsis. The ex-cycloptropia values were detected by double Maddox rod test. The cases with amblyopia were treated with occlusion therapy.

The criteria used in diagnosis of congenital superior oblique muscle palsy were: a history of ocular torticollis since early childhood, increased vertical fusional amplitudes and absence of torsional diplopia, hypertropia increasing by adduction of the paralytic eye, hypofunction of the superior oblique muscle and hyperfunction of its antagonist inferior oblique muscle, hypertropia increasing with head tilt toward the paralytic side (positive Bielschowsky head tilt test), extorsion less than 15° with Maddox rod, and lack of V-pattern deviation. Prior history of trauma to the eye, head, neck, or shoulder area, previous eye muscle surgery, orbital lesion, additional eye muscle palsy, significant horizontal strabismus, and other ocular disease were the criteria of exclusion. Cases with the absence or weakening of the tendon that was detected by intraoperative traction test were also excluded.

The ocular motility was assessed with a subjective grading (0 to 4) of underaction (-) or overaction (+) of the cyclovertical muscles. The deviation was measured in primary gaze and the cardinal positions of gaze, with or without the abnormal head posture, at both 33 cm and 6 m.\textsuperscript{8}

A 5-mm myectomy and concomitant disinsertion of the ipsilateral inferior oblique muscle was performed in 38 cases and 5-mm myectomy and concomitant inferior oblique disinsertion and recession of the superior rectus muscle in the ipsilateral eye was performed in 10 cases. Patients underwent a fornix-based 5-mm myectomy and concomitant unilateral inferior oblique disinsertion by one surgeon. The postoperative vertical deviation was accepted as “excellent” (0 to 3 prism diopters [PD]), “good” (4 to 7 PD), and “poor” (> 7 PD).\textsuperscript{9}

The value of vertical deviation was considered to decide surgical options. A 5-mm myectomy and concomitant disinsertion to ipsilateral inferior oblique muscle was performed in cases with a vertical deviation in primary gaze position of less than 25 PD, but 5-mm myectomy and concomitant inferior oblique disinsertion and recession to ipsilateral superior rectus muscle was performed in cases in which vertical deviation was 25 PD or greater.

All subjects were followed up for a minimum of 6 months postoperatively. The protocol in the postoperative period was control visits at week 1, months 1, 3, and 6, and at 3-month intervals thereafter. The paired sample \( t \) test was used in statistical analysis. A \( P \) value of less than .05 was considered statistically significant.

**RESULTS**

Thirty of the patients were female and 18 were male. Children with a mean age at operation of 56.7 ± 48.6 months (range: 36 to 112 months) were recruited, with a mean follow-up of 8.3 ± 5.2 months (range: 6 to 15 months).

The complaints at the time of admission to our clinics were abnormal head posture in 32 patients (67%), vertical deviation in 11 patients (23%), and diplopia in 5 patients (10%). Among these unilateral cases, 26 (54%) had left and 22 (46%) had right superior oblique muscle palsy. Facial asymmetry developed in 28 patients (58%) with congenital su-
perior oblique muscle palsy. Twenty-seven (56%) of the patients had a history of parental consanguinity. Six patients (13%) had physiotherapy before ophthalmology consultations. An orthopedic procedure had been planned in 3 (6%) patients previously. This could be a reason for having an ophthalmology consultation in any patient with torticollis in whom there is any question of an eye muscle imbalance.

Thirty-eight patients who had myectomy and concomitant inferior oblique disinsertion had preoperative values as follows: near deviation of 10.7 ± 5.8 PD (range: 8 to 24 PD) and far deviation of 11.4 ± 6.9 PD (range: 9 to 24 PD). Their postoperative values were near deviation of 1.8 ± 3.3 PD (range: 0 to 8 PD) and far deviation of 1.8 ± 3.7 PD (range: 0 to 8 PD) (Table 1). The difference of deviation between preoperative and postoperative values was statistically significant \( (P < .001) \). The patients included in this study were classified according to the criteria of residual vertical deviation as outlined earlier, 5 (50%) patients achieved an "excellent" (0 to 3 PD) postoperative result, 2 (20%) a "good" result (4 to 7 PD), and 3 (30%) a "poor" result (> 7 PD). The mean preoperative score of inferior oblique overaction was +3.36 (range: +3 to +4). Inferior oblique overaction diminished in 6 patients, 3 patients had +1.0 overaction in adduction of the affected eye, and 1 patient had +2.0 overaction in adduction of the affected eye.

A secondary operation was planned 6 months after the first operation in 2 eyes; 2 patients in whom abnormal head posture was found to persist and 1 patient who complained of diplopia. The preoperative surgical plan was to re-explore the inferior oblique muscle in all cases. In 3 eyes, residual posterior fibers were not identified and recession of the inferior rectus muscle of the contralateral eye was performed. Two patients achieved an "excellent" (0 to 3 PD) postoperative result and 1 a "good" result (4 PD) (Table 2).

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<th>Type of Surgery</th>
<th>No. of Cases (N = 48)</th>
<th>Preoperative Near Deviation (PD)</th>
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<td>Myectomy and concomitant IO disinsertion</td>
<td>38</td>
<td>10.7 ± 5.8 (8–24)</td>
<td>11.4 ± 6.9 (9–24)</td>
<td>1.8 ± 3.3 (0–8)</td>
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<td>Myectomy and concomitant IO disinsertion and SR recession</td>
<td>10</td>
<td>20.5 ± 10.7 (25–32)</td>
<td>22.5 ± 11.7 (25–33)</td>
<td>2.5 ± 3.8 (0–10)</td>
<td>2.1 ± 4.2 (0–10)</td>
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SD = standard deviation; PD = prism diopters; IO = inferior oblique muscle; SR = superior rectus muscle.
Contralateral inferior oblique hyperfunction was detected at the final postoperative visit of 4 (11%) of 38 patients who had myectomy and concomitant inferior oblique disinsertion. Mild (-1) inferior oblique hypofunction was found in 1 (3%) of 38 patients who had myectomy and concomitant inferior oblique disinsertion. Contralateral inferior oblique hyperfunction developed in 1 (10%) of 10 patients in whom myectomy and concomitant inferior oblique disinsertion and ipsilateral rectus recession were performed. Restrictive complications such as adherence syndrome defined with hypotropia, restriction of elevation, perforation of sclera, vortex vein injury, and macular damage were not detected in either group. The 4 patients with myectomy and concomitant inferior oblique disinsertion who developed contralateral inferior oblique hyperfunction may represent a masked bilateral superior oblique palsy in which superior oblique palsy in the opposite eye appears after surgically correcting one side. The same could be said about one patient who developed the same after myectomy and concomitant inferior oblique disinsertion and ipsilateral superior rectus recession.

Double Maddox rod test could be performed in 30 (63%) patients and excyclotropia was detected in 20 (42%) patients. The range of excyclotropia values was between 4° and 14°. Fusion was detected in 34 (71%) patients by Worth 4-dot test. Stereopsis was established in 28 (58%) patients by Titmus test, and it was measured between 400 and 40 seconds of arc. Stereopsis was not detected in the preoperative examinations of 20 patients, but developed (200 to 400 seconds of arc) in 8 (17%) patients during the postoperative period. Abnormal head posture completely resolved postoperatively in all cases that had abnormal head posture preoperatively. None of the patients had residual diplopia in primary gaze and reading positions postoperatively.

**DISCUSSION**

The superior oblique is a challenging cyclovertical muscle. It is difficult to classify the disorders of this muscle, so it has variable anatomic location and strength of action. Superior oblique dysfunction will cause complex adaptive mechanisms leading to secondary changes such as further complications of other muscles. The most important cause of ocular torticollis is superior oblique muscle palsy. The patient will develop compensatory head posture to place the eyes in a particular field of gaze to obtain binocular single vision. Recent studies demonstrated that the effect of a compensatory head posture in patients with unilateral superior oblique muscle palsy is to minimize the vertical deviation rather than to decrease ocular torsion. A paradoxical posture such as opposite to the usual abnormal head posture expected for maximal separation of the diplopia was observed in a small group of patients (3.3%). Mitchell examined 630 patients with ocular torticollis and found 52.4% had incomitance, 19% had nystagmus, 10.9% had congenital esotropia with ocular posture, and 4.3% had torticollis permitting foveal fixation. Lau et al. did not find any apparent cause in 9.0% of the patients.

Unilateral superior oblique muscle palsy was diagnosed by the three-step test plus the presence of an excyclotropia (either subjectively seen with the double Maddox rod test or objectively seen on funduscopy), the absence of reversal of the hypertropia in any of the nine diagnostic fields of gaze or head tilt right and left, and the absence of

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PD = prism diopters.
dissociated vertical divergence. Secondary changes and muscle contracture take place more often in an older child at the time of operation. The presence of torticollis is the cause of shortening and flattening of the hemiface to which the head inclined. The history of a long-standing compensatory head posture in a congenital superior oblique muscle palsy can be documented as the old photographs of the patient are reviewed or when there is facial asymmetry, which is common in congenital cases. Lau et al. reported the ratio of 50% facial asymmetry in patients with congenital superior oblique muscle palsy. Twenty-eight (58%) patients were detected to have facial asymmetry in our study.

Huge vertical deviation is common in congenital superior oblique muscle palsy. In addition to the differences between excyclotropia and vertical deviation and the incomitance pattern, the degree of strabismus depends on the grade of the palsy and the anatomy of the patient. Individual variability of the pulling directions of both the superior oblique tendon and the inferior oblique muscle has already been established by post-mortem studies. Many variable techniques have been recommended for the initial treatment for congenital superior oblique muscle palsy. Toosi and Von Noorden reported myectomy as the primary procedure. Saunders recommended superior oblique tendon tucking with ipsilateral inferior oblique myectomy as safe and effective in patients with vertical deviation of greater than 30 PD. Chang et al. suggested inferior oblique anterior transposition, Mulvihill et al. reported inferior oblique disinsertion and Duranoglu recommended inferior oblique disinsertion–resection–tucking techniques for superior oblique muscle palsy treatment.

Lau et al. reported their series of 32 patients diagnosed as having congenital superior oblique muscle palsy. They performed ipsilateral inferior oblique recession, contralateral inferior rectus recession, or both. The mean preoperative vertical deviation in primary gaze was 14.3 ± 5.7 PD and postoperative vertical deviation was 3.5 ± 5.5 PD. The mean difference between the preoperative and postoperative vertical deviation was 10.7 PD.

Duranoglu reported the results of 31 patients with unilateral long-standing superior oblique muscle palsy who underwent “disinsertion-resection and tucking of the inferior oblique muscle.” The preoperative mean inferior oblique overaction was graduated at 3° and the mean vertical deviation was 15.9 PD; 83.8% had a satisfactory outcome (defined as improvement of torticollis or recovery of diplopia in primary gaze). Elevation deficit, abnormal eyelid position, pupillary abnormality, or hypotropia in primary position were not detected in any patients. Possible reasons for failure of inferior oblique weakening surgery include a structural abnormality, missed posterior fibers, or inadequate superior oblique muscle force.

Simons et al. found that 60% of 123 cases of superior oblique muscle palsy had an “excellent” result for vertical deviation postoperatively. Von Noorden et al. reported a surgical success rate of 89% in 96 cases. However, their group only used head tilt as a measure of abnormal head posture, without considering the face turn or chin position.

Davis et al. reported a series of 37 patients who remained symptomatic after an initial inferior oblique weakening procedure for superior oblique muscle palsy. Twenty-four percent remained symptomatic after their second surgery. As a result they suggested a convenient step-wise management for patients with failed initial ipsilateral inferior oblique weakening procedure. Although surgery to treat unilateral superior oblique muscle palsy usually involves oblique muscle surgery, the motility pattern may indicate that rectus muscle surgery alone may be preferable, especially when previous oblique muscle surgery already has been performed.

Our study results show that myectomy and concomitant inferior oblique disinsertion or myectomy and concomitant inferior oblique disinsertion and superior rectus recession of ipsilateral eye are effective, convenient, reliable, and successful methods in patients with unilateral congenital superior oblique muscle palsy.

REFERENCES
7. Saunders RA. Treatment of superior oblique palsy with superior oblique tendon tuck and inferior oblique muscle myectomy. Oph-