Idiopathic Superior Oblique Palsies in Children

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ABSTRACT
The records of 110 patients with superior oblique palsy seen at the Boston Children's Hospital between 1966 and 1988 were reviewed. Patients with identifiable orbital or neurological disorders affecting the fourth cranial nerve or with a history of head trauma preceding their superior oblique palsy were excluded from analysis. Patients were also excluded because of prior surgery elsewhere or insufficient diagnostic information. The remaining 63 patients were considered to have idiopathic superior oblique palsies. All 63 patients presented with unilateral palsies. All had incomitant hypertropia of the affected eye, greater on gaze to the contralateral side and increased by ipsilateral head tilt. Left eyes were affected in 41 patients. The age at first ophthalmological examination was spread over the first 20 years, with heavier representation in the first 5 years and decreasing numbers in each succeeding 5-year interval. Only one patient was found to have a masked bilateral superior oblique palsy following surgery for an apparent unilateral palsy. A distinction is made between unmasking a bilateral superior oblique palsy and surgical overcorrection of a unilateral palsy. The predominant unilaterality of idiopathic superior oblique palsies is in contrast to the reported frequency of bilaterality in traumatic cases.

INTRODUCTION
Uncertainty about the frequency of bilateral involvement in nontraumatic superior oblique palsies prompted a review of the cases that had been seen at the Boston Children's Hospital over a period of slightly more than 20 years. Those fourth nerve palsies associated with recognizable orbital or neurological disorders were excluded from consideration. A developmental origin of the remaining palsies was presumed, although no anatomical data was available in support of this inference. Since the actual time of onset of the vertical deviation was only imprecisely established by history, no clear distinction could be made between congenital and acquired palsies, and all were grouped together under the designation idiopathic.

PATIENTS AND METHODS
The records of 110 patients with superior oblique paralysis seen at the Boston Children's Hospital between 1966 and 1988 were reviewed. Twenty-five patients had identifiable orbital or neurological disorders affecting the fourth cranial nerve (Table) and were excluded from further consideration. Fifteen patients had a clear history of head trauma preceding their fourth nerve palsy, and these too were set aside. Seven additional patients were excluded from analysis because of prior surgery elsewhere or lack of a recorded Bielschowsky head tilt test. The remaining 63 patients were considered to have idiopathic superior oblique palsies. All had an incomitant hypertropia of the affected eye, greater on horizontal gaze to the opposite side and increased by head tilt to the same side. Most had some underaction of the affected superior oblique muscle on ocular versions, combined with more obvious overaction of the ipsilateral inferior oblique. Patients with V-pattern esotropia and primary overaction of the inferior oblique muscles unassociated with a positive Bielschowsky head tilt test were not included in this study. The vertical deviation in the primary position, right and left gaze, and with head tilt right and left was measured, when possible, by prism and alternate cover testing. Krimsy estimates were sometimes used in younger children. Ocular versions were assessed by observation as the patient followed visual stimuli into the cardinal positions of gaze. Overactions or underactions of the oblique muscles were graded on a 1 to 3 scale, but this quantitation was considered only approximate because of the multiplicity of examiners. A spontaneous head tilt to
the opposite shoulder was observed in many patients, however it was not considered essential for the diagnosis.

Age at the time of first ophthalmological examination was noted and is presented in Fig 1. Although signs and symptoms of the superior oblique paralysis preceded this age by varying amounts, it was not possible to estimate the time of onset with accuracy. The results of subjective visual acuity testing and cycloplegic refractions were recorded as they became available during initial and follow-up examinations.

The type and number of surgical procedures performed to correct the superior oblique palsy were recorded. This compilation is complete for 40 patients, whereas other patients moved and were referred elsewhere for surgery, and still others have been lost to follow-up. Particular attention was paid to evidence for bilateral superior oblique palsy, both pre- and postoperatively. Preoperatively this would be indicated by right hypertropia in left gaze and left hypertropia in right gaze and a positive Bielschowsky test to both shoulders. Surgically unmasked bilateral involvement was looked for in the form of a postoperative incomitant hypertropia of the eye that had previously seemed to be unaffected, a reverse of the preoperative spontaneous head tilt and Bielschowsky test, and newly evident imbalance of the oblique muscles on version testing.

Sensory evidence for binocular vision with Worth lights and stereopsis with polaroid or random dot stereotests was counted as being present, whether it was documented before or after surgery. Information about family history of strabismus in siblings and parents was available in 49 of 63 records. The 63 patients with idiopathic superior oblique palsy were in good general health except for the following conditions that were felt to be coincidental: dysplastic kidney (1 patient), Hirschsprung’s disease (1), mild microcephaly (1), seizure disorder (1), neurofibromatosis without other cranial nerve involvement (2), Down syndrome (1), gastroschisis (1), asthma (2).

![FIGURE 1: Age in years at the time of first ophthalmological examination for idiopathic fourth nerve palsy.](image)

**TABLE**

<table>
<thead>
<tr>
<th>Orbital or Intracranial Pathology Causing Fourth Nerve Palsy</th>
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<tbody>
<tr>
<td>5  Rhabdomyosarcoma of orbit or nasopharynx</td>
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<tr>
<td>2  Brain stem glioma</td>
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<tr>
<td>1  Orbital stick injury</td>
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<tr>
<td>2  Cerebral artery aneurysm</td>
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<tr>
<td>1  Neurofibromatosis with multiple cranial nerve palsies</td>
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<td>5  Tumor or pseudotumor cerebri</td>
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<tr>
<td>5  Meningitis, encephalitis, or cerebral abscess</td>
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<td>1  Polyneuritis</td>
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<tr>
<td>1  Guillain-Barre syndrome</td>
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<td>1  Congenital myasthenia</td>
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<td>1  Seizure disorder with transient fourth nerve palsy</td>
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**RESULTS**

All 63 patients with idiopathic superior oblique palsies presented with a unilateral palsy. In some patients the hypertropia disappeared on gaze to the ipsilateral side, but in no case did it reverse, nor did the Bielschowsky head tilt test cause a reversal of the hypertropia with tilt to the contralateral side. The vertical deviation was invariably greatest with gaze to the side opposite the palsy or with head tilt toward the ipsilateral shoulder. Forty-eight of the 63 patients had an abnormal spontaneous head position, usually with the head tilted and the face somewhat turned to the opposite shoulder. In those patients without a head tilt a unilateral vertical deviation was a common initial complaint, and among older children intermittent diplopia was occasionally noted. No patient complained of image tilt.

In 41 of the 63 patients the left eye was paretic, a preponderance that is statistically significant at the 0.05 level. There were 40 males and 23 females in the group, a distribution which is not statistically different from that of the general population in the United States under the age of 20 years (U.S. Census Bureau data from 1980 census gives 51.1% males and 48.9% females in this age group).

The time of first ophthalmological examination was spread over the first 20 years of life (Fig 1). The largest proportion presented in the first 5 years (48%), and decreasing numbers had their first examination in subsequent 5-year intervals: 5-10 years, 24%; 10-15 years, 18%; 15-20 years, 10%. This distribution is, of course, affected by the fact that patients were seen at a children’s hospital where few individuals over 20 years of age are examined.

No patient had strabismic amblyopia. No particular pattern or asymmetry of refractive errors could be identified. Among the 40 patients in whom responses to Worth lights or stereopsis testing were recorded, 28 demonstrated fusion with Worth lights, 25 had some level of stereopsis, 37 had either fusion or stereopsis or both, and three had no binocularity. Those without binocularity had a horizontal deviation in addition to their vertical strabismus and had an alternating fixation pattern.
IDIOPATHIC SUPERIOR OBLIQUE PALSIES

The vertical deviation in the primary position in 50 patients for whom a quantitative deviation in prism dipters was recorded is presented in Fig 2. Both the mean and the median vertical deviations were 15 prism dipters. Sixty-two of 63 patients showed some overaction of the ipsilateral inferior oblique muscle by observation of ocular versions. Fifty-two patients demonstrated some underaction of the affected superior oblique muscle, but this was usually more difficult to recognize than the inferior oblique overaction. Thirteen of the 63 patients had overaction of the contralateral superior oblique.

Surgery was performed on 40 of the 63 patients at Boston Children's Hospital. Sixteen patients had no surgery, and seven were referred elsewhere for operation because of convenience or a family relocation. All 40 patients who had surgery at Children's Hospital had an ipsilateral inferior oblique myectomy. Four had an additional recession of the contralateral inferior rectus, two a recession of the ipsilateral superior rectus, and two a tenotomy of the contralateral superior oblique. Four patients required a repeat myectomy of the ipsilateral inferior oblique, and four required horizontal rectus surgery for a coexisting horizontal deviation.

Only one patient was found to have weakness of the opposite superior oblique following surgery for what initially appeared to be an isolated left superior oblique palsy. In this patient, approximately 1 year after a left inferior oblique myectomy had been performed, a right hypertropia developed, the original spontaneous right head tilt was replaced by a left head tilt, the Bielschowsky test that had been positive to the left became positive to the right, and the right hypertropia was noted to increase on left gaze. A myectomy of the right inferior oblique was performed, eliminating the hypertropia and the head tilt. Two other patients developed significant overcorrections of their left superior oblique palsies, one after myectomy of the ipsilateral inferior oblique and a sequential recession of the contralateral inferior rectus, the other after the same surgery plus recession of the ipsilateral superior rectus. Although the hypertropia switched to the right eye, neither patient developed a positive Bielschowsky test to the right shoulder, nor did the right hypertropia show incomitance consistent with right oblique muscle imbalance. Both patients underwent further surgery to eliminate the overcorrection, and both continued to exhibit a slightly positive Bielschowsky test with forced head tilt to the left.

Six patients have developed a slightly positive Bielschowsky test to the side opposite the original superior oblique palsy after myectomy of the ipsilateral inferior oblique, without a noticeable hypertropia in the primary position. One of these patients also had a tenotomy of an overacting contralateral superior oblique, and the surgical weakening of this superior oblique could account for the reversed Bielschowsky test. In the other five patients surgical weakening of the inferior oblique itself could explain the positive Bielschowsky test to the contralateral side. In none of these patients has imbalance of the contralateral obliques become apparent, nor is there an increasing vertical deviation in the field of action of these muscles.

Of the 40 patients whose surgery was performed at Children's Hospital, 32 had only a small or no residual head tilt following their last surgery. Six patients had a noticeable residual head tilt, and for two patients there was incomplete follow-up data in the record. Nine patients had greater than 5 prism dipters of residual hypertropia in the primary position, but for various reasons no further surgery was done.

Among the 49 patients for whom data on a family history of strabismus was recorded, 12 patients (24%) were noted to have at least one sibling or parent with strabismus. Two of the patients with left superior oblique palsies were sisters, and the father of one male patient with a right superior oblique palsy was known also to have a right superior oblique palsy (not included in this study). Five patients in the study had twin siblings, one fraternal and four considered to be identical. None of the identical siblings was known to have a superior oblique palsy.

DISCUSSION

The term idiopathic superior oblique palsy has been used to distinguish this condition from traumatic fourth cranial nerve palsies and fourth nerve palsies resulting from orbital or intracranial disease of recognizable nature. It is reasonable to assume that the idiopathic palsies are developmental in origin, although no anatomical evidence in support of this etiology is known to the author. The presentation of idiopathic palsies is often in the early years, but over half of them come to the ophthalmologist for the first time after the age of 5 years. Unrecognized or unreported trauma is a possible cause in some instances.

The great preponderance of unilateral cases in the idiopathic group is confirmed by Helveston and Ellis. VonNoorden, Murray, and Wong found seven bilateral cases in 107 patients with superior oblique palsy that dated back to infancy and only one bilateral case in a group.
of 63 idiopathic superior oblique palsies that were later in onset but not traumatic in origin. The latter authors mentioned that bilateral palsies are frequently asymmetrical in presentation and may appear to be unilateral prior to surgery. The unmasking of a bilateral superior oblique palsy after initial surgery for a unilateral palsy has been mentioned frequently in the literature, especially with reference to traumatic cases among which the prevalence of bilateraltery may reach 20%. A few unmasked idiopathic bilateral cases, similar to the one in our series, have been reported. Often it has been left unclear how frequent these cases were in the total population of idiopathic fourth nerve palsies. A distinction should be made between unmasked bilateral cases and surgical overcorrection of unilateral palsies. In the latter instance there is reversal of the hypertropia in the primary position but no sign of superior oblique weakness in the fellow eye. With regard to the five patients in this series who had slight reversal of the Bielschowsky test following ipsilateral inferior oblique myectomy, it might be postulated that this represents a subtle unmasking of a bilateral superior oblique palsy. Against this possibility is their failure to exhibit imbalance of the contralateral obliques or an increasing vertical deviation in the field of action of these muscles. These latter two signs were the first indication of an unmasked bilateral palsy in Kraft and Scott's series.

In addition to being predominantly unilateral, idiopathic superior oblique palsies appear to be significantly more frequent in left eyes than in right. The developmental significance of this laterality is not at present clear. A similar predilection for left eye involvement is seen in the Duane syndrome, a condition which is also predominantly unilateral and due to an anomaly of development of cranial nerves. In Duane syndrome, as in idiopathic superior oblique palsy, there may be a family history of similar involvement, although most instances are sporadic.

Amblyopia appears to be a matter of little concern in superior oblique palsies. Presumably this is due to the fact that most patients can achieve at least intermittent fusion with a combination of favorable head position and large fusional vergences. The exceptions to this rule are those patients with significant horizontal deviations in addition to their superior oblique palsies. Breaks of fusion lead to vertical diplopia in some older patients, but not to torsional symptoms, a fact suggesting an early onset for the underlying deviation.

Strabismus surgery was generally helpful in this group of patients. Weakening of the ipsilateral inferior oblique muscle was the most common procedure employed because overaction of this muscle was virtually universal and myectomy has been shown to be effective in reducing the deviation.

Surgical tucking of the affected superior oblique was not performed, although it has been used with satisfaction by some surgeons. A surprisingly large proportion of patients had no surgery, either because their deviation was small and no surgery was suggested, or because the parents were not troubled by the relatively modest head tilt that was used to control the vertical deviation. For these same reasons small surgical undercorrections seemed to be tolerated well by a number of patients.

REFERENCES