Special Types of Strabismus

Special forms of strabismus exist that, because of their unusual features, deserve discussion in a separate chapter. Some of these forms are caused by structural anomalies of the extraocular muscles or adjacent tissues. Their management varies according to their etiologic basis, and thus treatment often differs from that of ordinary types of comitant or noncomitant strabismus. For this reason, a discussion on treatment is included in this chapter after the description of the clinical findings for each of these special forms.

Retraction Syndrome (Duane Syndrome)

During the late nineteenth century, several papers were published that drew attention to a syndrome consisting of marked limitation or absence of abduction, restriction of adduction, retraction of the globe, and narrowing of the palpebral fissure on adduction. These manifestations were frequently associated with elevation or depression of the globe in adduction. Heuck was the first to describe retraction of the globe in a patient with severe limitation of ocular motility. Stilling, Türk, Bahr, Sinclair, Wolff, and others provided detailed descriptions of this syndrome, and when Alexander Duane published his paper in 1905, 54 cases were available to him for analysis. Even though Duane never claimed priority for discovery of this entity, in the United States his name has become attached to the retraction syndrome. In the European literature the syndrome is referred to, perhaps more appropriately, as the Stilling-Türk-Duane retraction syndrome.

Since these early descriptions, literally hundreds of papers dealing with the retraction syndrome have been published, which attests to the fact that this anomaly of ocular motility is by no means rare. The literature has been reviewed by various authors.

Laterality and Sex Distribution

There is general agreement that Duane syndrome is a more common occurrence in the left eye than in the right eye and also more common in females. Bilateral involvement is less common than unilateral occurrence. Only minor variations among the study results of different authors existed in large recent surveys. These data are summarized in Table 21–1, which has been modified from a survey of the literature on Duane syndrome by DeRespinis and coworkers.

<p>| TABLE 21–1. Sex Distribution and Laterality in 835 Patients with Duane Syndrome |
|----------------------------------|-------|-------|-------|-------|-------|</p>
<table>
<thead>
<tr>
<th></th>
<th>Female</th>
<th>Male</th>
<th>Left Eye</th>
<th>Right Eye</th>
<th>Bilateral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female</td>
<td>58%</td>
<td>42%</td>
<td>59%</td>
<td>23%</td>
<td>18%</td>
</tr>
</tbody>
</table>

Etiology

STRUCTURAL ANOMALIES. The etiology of the retraction syndrome has intrigued ophthalmologists for the past 90 years. In the older literature, most authors favored the view that congenital structural anomalies were the cause of the retraction phenomenon. Heuck,165 while performing surgery on one of his patients, found a posterior insertion of the medial rectus muscle, which he thought to be the cause of retraction of the globe on adduction. Bahr,18 Apple,8 and Bielschowsky27 shared this view, that a posteriorly inserted medial rectus muscle may act as a retractor bulbi.

We have observed a band that originated in the orbital apex and inserted 6 mm behind the medial rectus muscle in a patient with Duane syndrome, type I.261 Türk377 believed that fixation of the globe by a nonelastic lateral rectus muscle was the cause of retraction on adduction. Many other authors, who during surgery found a fibrotic nonelastic lateral rectus muscle or restricting fibrous bands beneath the muscle insertion, agreed with this view.14, 208, 233, 258, 347

The abnormal vertical eye movements that frequently occur with adduction were blamed in the older literature, on oblique overaction. T. Duane and coworkers98 believed that compensatory oblique overaction replaces the defective abducent action of the lateral rectus muscle. Parker280 thought that the vertical movement in adduction is caused by overaction of the vertical rectus muscles, which act as adductors if the medial rectus muscle is weak. Scott332 and Souza-Dias346 offered an entirely different explanation for this phenomenon, one based on co-contraction of the horizontal muscles (see p. 464). As a matter of historical interest, the curious theory of Wolff403 should be mentioned. He blamed elevation or depression in adduction on the attachment of the globe to the optic nerve, which would cause resistance in the plane of retraction and pull the eye in a vertical direction.

Many investigators have reported finding an abnormal lateral rectus muscle during surgery, but only a few histologic reports are available in the literature. Krüger208 reported degenerative changes in the lateral rectus muscle with an increase of fibrous tissue.

PARADOXICAL INNERVATION. Evidence accumulated from electromyographic studies has indicated that an innervational mechanism rather than anatomical abnormalities may be responsible for most cases of the retraction syndrome. Breinin39 was first to describe paradoxical electrical behavior of the lateral rectus muscle in such patients, that is, absence of electrical activity in this muscle on abduction and active electric potentials on adduction. He evolved the theory, now shared by many others, that this anomalous co-contraction of the medial and lateral rectus muscles is the cause for retraction of the globe on adduction. Paradoxical electromyographic activity of the medial rectus muscle in patients with Duane syndrome, type I has also been reported.318 Gross and coworkers341 reported absence of retraction in a patient with Duane syndrome, type II with electromyographically proven co-firing of medial and lateral rectus muscles on attempted adduction. These authors concluded from their findings that co-contraction of opposing muscles alone may not suffice to cause the retraction but that additional mechanical factors may play a role.

Breinin explained the abnormal activity of the lateral rectus muscle on adduction as an abnormally sensitized stretch reflex, a theory that cannot be upheld in view of the findings of Blodi and coworkers,29 who demonstrated co-contraction in an unstretched lateral rectus muscle detached from the globe during surgery. Breinin’s findings of paradoxical innervation were confirmed and enlarged on by many other investigations78, 173, 276, 325, 333 (Fig. 21–1). Strachan and Brown364 in a series of patients with Duane syndrome, demonstrated a spectrum of abnormal electrical activity in the lateral rectus muscle ranging from paradoxical innervation to subnormal firing in abduction. Abnormal synergistic innervation between the medial rectus muscle and the superior and inferior rectus or oblique muscles also was demonstrated electromyographically, which may explain the vertical deviation in adduction observed in some patients with this condition.276, 325 although other explanations have been offered. Narrowing of the palpebral fissure on adduction is usually interpreted as a passive adjustment of the lids to the retracting globe. Other explanations for the eyelid findings include decreased innervation to the levator palpebrae on adduction110 and a reorganization of the central oculomotor pathways.250
The currently favored theory is that in most instances the retraction syndrome is an innerva-
tional disturbance of brain stem origin rather than an anomaly caused by structural anomalies of the
muscles. A case of acquired bilateral retraction syndrome in a 23-year-old woman with a brain
stem tumor\(^{265}\) and another of a patient in whom co-contraction developed in both horizontal rectus
muscles in adduction and downward gaze after a head injury\(^{333}\) lend credence to this hypothesis.

Hoyt and Nachtigaller\(^{169}\) proposed that ocular retraction during adduction, as well as the electro-
myographic findings of synergistic innervation of medial and lateral rectus muscles, could be ex-
plained on the basis of substitute innervation of a paretic lateral rectus muscle by an extra branch
from the oculomotor nerve.\(^{42}\)

Matteucci\(^{232}\) described absence of the ipsilateral abducens nerve and hypoplasia of its nucleus in a
well-documented case of Duane syndrome. However, he did not discuss innervation of the lateral
rectus muscle, nor did he trace the terminal branch of the oculomotor nerve, as has been done in more
recent studies.\(^{372}\) More detailed clinicopathologic evidence of central nervous system anomalies in
patients with the retraction syndrome was provided by two important studies from the Wilmer
Institute at Johns Hopkins Hospital. In a patient with bilateral Duane syndrome, both abducens nu-
clei and nerves were absent from the brain stem and the lateral rectus muscles were partially inner-
vated by branches from the oculomotor nerves.\(^{167}\)

In a second patient with unilateral Duane syn-
drome, these same findings were limited to the involved side.\(^{245}\) The findings were confirmed by
additional reports. Mulhern and coworkers\(^{257}\) described absence of the right abducens nucleus in
a patient with Duane syndrome, type III of the right eye and Parsa and coworkers\(^{286}\) described
absence of the abducens nerve on magnetic resonance imaging (MRI) in Duane syndrome, type I.

According to Saad and coworkers\(^{319}\) the clinical features of Duane syndrome may be explained by
a failure to differentiate and to displace the abducens nucleus from the oculomotor nucleus in the
human embryo of day 21 to day 26.

Clinicopathologic evidence, the number of pa-
tients with acquired retraction syndrome and brain
tumor or trauma,\(^{270, 333}\) or on the basis of a nonspe-
cific vasculitis,\(^{6}\) the frequent association of the
gustolacrimal reflex (crocodile tears),\(^{78, 247, 304}\)
anomalies of the vestibulo-ocular reflex, optoki-
netic nystagmus,\(^{136}\) and auditory evoked re-
sponses\(^{186}\) clearly place the seat of this anomaly
in the brain stem.

**HEREDITY.** Hereditary patterns of Duane syn-
drome were mentioned by many investigators, and
familial occurrence with dominant inheritance pat-
terns\(^{66, 118, 200, 377, 385, 404}\) and Duane syndrome in
monozygotic twins\(^{235, 315}\) have been described. A
gene responsible for Duane syndrome and a domi-
nant form of hydrocephalus has been identified
and is located close to a gene causing theranchio-oto-renal syndrome.\(^{384}\) Other studies
have identified abnormalities in chromosomes in
patients with Duane syndrome\(^{5, 55, 64, 81, 384}\)
EMBRYOPATHY. In view of the high prevalence of ocular or systemic malformations associated with Duane syndrome, Cross and Pfaffenbach proposed the intriguing theory that a common teratogenic stimulus at 8 weeks of gestation may be of etiologic significance in sporadic cases of Duane syndrome—a thought that had been expressed earlier by Krüger. Duane syndrome has been reported in connection with the fetal alcohol syndrome, which suggests damage to the developing abducens nuclei in the middle of the first gestational trimester. A case of Duane syndrome was reported in a patient with a giant aneurysm of the vertebral basilar arterial junction. The authors speculated that Duane syndrome may be caused by vascular hypofunction during the fourth to fifth week of embryogenesis. Convincing evidence that the etiology of the retraction syndrome may be teratogenic in nature is provided by the finding that it occurs frequently in patients afflicted with the thalidomide syndrome. At this time it appears that several etiologic factors may be involved in the retraction syndrome, and it is doubtful that a single mechanism is responsible for this disturbance of ocular motility. The reported anatomical changes involving the horizontal rectus muscles imply a peripheral structural etiology in some cases, and such findings cannot be ignored. On the other hand, the electromyographic evidence of paradoxical innervation of the lateral rectus muscle and the clinicopathologic correlations cited above are equally convincing. One must also consider the possibility that some of the anatomical anomalies observed during surgery could be secondary to a primary innervational anomaly. For instance, we have been impressed by the frequent finding of a tight lateral rectus muscle at the time of surgery in Duane syndrome, types I and III. The possibility must be entertained that this muscle loses its elasticity from lack of contraction, which would contribute to the retraction of the globe on attempted adduction. T. Duane and coworkers reported narrowing of the palpebral fissure and retraction of the globe in abduction following fracture of the medial orbital wall with entrapment of the medial rectus muscle. We have seen retraction on attempted adduction after massive conjunctival scarring following removal of a dermolioma from the temporal aspect of the bulbar conjunctiva of the left eye (Fig. 21–2). Thus, retraction can be explained on a mechanical basis alone to which an innervational factor may be added in cases of co-contraction.

Clinical Findings and Diagnosis

The retraction syndrome in its classic form is characterized by the following features:

1. Congenital onset (acquired forms are rare)
2. Severe limitation of abduction
3. Slight limitation of adduction
4. Globe retraction and narrowing of the palpebral fissure on adduction
5. Commonly associated elevation or depression in adduction

Owing to the lack of the patient’s cooperation the diagnosis may be difficult to make in infants. Retraction, elevation, or depression of the globe on adduction may not be detected until early childhood. We have seen several instances in which Duane syndrome, type I, was mistaken for essential infantile esotropia. Resection of the lateral rectus muscle in such patients will increase retraction of the globe on adduction and must be avoided. The differential diagnosis should also include congenital abducens paralysis even though in such cases the angle of esotropia in primary position is usually much larger than in the retraction syndrome.
Variations of this special form have been described. The first case of vertical retraction was reported by Böhm (1845) in a 40-year-old woman who from birth was unable to adduct or abduct her left eye. Abduction of the right eye caused elevation and retraction of the left eye, and on adduction of the right eye the left eye depressed. Brown reported retraction of both eyes in upward gaze and Khodadoust and von Noorden observed retraction on downward gaze in two siblings. It is likely that the cause of this retraction was fibrosis of the superior or inferior rectus muscles rather than co-contraction. However, retraction in upward gaze has also been reported in a patient with a classic bilateral Duane syndrome, type I. Pesando and coworkers reported a case of unilateral vertical retraction, and Osher and coworkers described acquired retraction of the globe on attempted gaze opposite the field of action of the involved muscles in patients with infiltrative myopathy caused by dysthyroid eye disease, inflammatory myositis, and neoplasms. Weinacht and coworkers showed lateral rectus muscle firing activity during upgaze and downgaze in a rare variant of a vertical retraction syndrome. Huber suggested the following useful classification to include most clinical variations in this entity:

**Duane I:** Marked limitation or complete absence of abduction; normal or only slightly defective adduction; narrowing of the palpebral fissure and retraction on adduction; widening of the palpebral fissure on attempted abduction (Fig. 21–3)

**Duane II:** Limitation or absence of adduction with exotropia of the affected eye; normal or slightly limited abduction; narrowing of the palpebral fissure and retraction of the globe on attempted adduction (Fig. 21–4)

**Duane III:** Combination of limitation or absence of both abduction and adduction; re-
FIGURE 21–5. Duane syndrome, type III, left eye. A, Face turn to the right and fused with the head position. B, 20° exotropia in primary position; imitation of adduction and abduction of the left eye with widening of the left palpebral fissure on levoversion and narrowing on dextroversion; upshoot of the left eye when attempting to adduct with the right eye elevated and downshoot when attempting to adduct in depression. C, Retraction of the globe on attempted adduction (lower photograph). (From Noorden GK von: Atlas of Strabismus, ed 4. St Louis, Mosby–Year Book, 1983.)

Type I is by far the most common (78%), followed in order of frequency by types III (15%) and II (7%), according to data from different authors and compiled by DeRespinis and coworkers. Not only is Huber’s classification the most useful one clinically, but it is supported by electromyographic documentation. Huber found paradoxical innervation in association with all three types of Duane syndrome. His work was confirmed by Maruo and coworkers who obtained electromyograms in 126 patients with the various types of this disorder. In those with type I the peak of innervation of the lateral rectus muscle occurs on adduction and the minimum on abduction. In those with type II the lateral rectus muscle fires maximally in abduction and adduction, and type III is characterized by simultaneous electrical activity of both the medial and lateral rectus muscles on adduction and abduction. Clear distinction between the different types is not always possible. For instance, a moderate limitation of abduction may be present in type II and other forms of “mixed” types may occur.

A bilateral case of Duane syndrome, type I is shown in Figure 21–6.

In the classic retraction syndrome, strabismus may or may not be present with the eyes in primary position. If strabismus is present, esotropia occurs more frequently than exotropia in patients with Duane syndrome, types I and III, and exotropia is a more frequent occurrence in those with type II. Many patients adopt a face turn to maintain single binocular vision. Complaints of diplopia are rare, except in the rare, acquired case and in view of the difficulties encountered in plotting suppression scotomas in such patients it has been suggested that the second image is ignored rather than suppressed.

Of special interest is the frequently associated upshoot and downshoot of the adducted eye, which at times causes a cosmetic problem of almost grotesque proportions; namely when the cornea of the adducted eye disappears from view (Figs.
Clinical Characteristics of Neuromuscular Anomalies of the Eye

At first glance, such patients appear to have increased overaction of the inferior or superior oblique muscles. Experience has taught us, however, that surgical weakening procedures on these muscles is entirely ineffectual. Scott\(^3\) pointed out that the high muscle tension caused by co-contraction of the horizontal rectus muscles or by structural tightness of the lateral rectus muscle when the medial rectus muscle contracts results in a vertical effect by allowing the muscles to slide over the globe ("bridle" or "leash effect") when contracting (see also Jampolsky\(^1\)). For instance, co-contraction of the medial and lateral rectus muscles, as occurs in adduction with Duane syndrome, type I, will increase the elevating effect of the horizontal muscles of a slightly elevated eye and cause upshoot, and the depressing effect of a slightly depressed eye will cause downshoot. Co-contraction of the horizontal muscles may enhance this effect not only in horizontal gaze but also when the eye elevates or depresses, as illustrated by Scott and Wong\(^3\) in their electromyographic studies. However, it has been shown by computed tomography (CT) scanning\(^3\) and MRI\(^8\) that it is actually not the muscles that slide over the surface of the globe (as had been suggested by Scott\(^2\)) but the globe that slips under the muscles, because the vertical displacement of the horizontal muscles during elevation and depression in relation to the orbital wall is negligible in most but not in all cases of Duane syndrome.\(^24\) It is because the horizontal rectus muscles maintain their vertical position with reference to the orbital walls that elevation of the eyes will move the center of rotation of the globe below the planes of the horizontal rectus muscles and depression will move this center above the muscle planes. This explains the bridle effect that occurs during co-contraction of these muscles when the eye is slightly elevated or depressed.

To control the change of the relationship between the muscle planes and the center of rotation of the globe Scott and Wong\(^3\) suggested retroequatorial fixation (posterior fixation suture) or a maximal recession of both horizontal rectus muscles (see also Souza-Dias\(^3\)). The results achieved with this surgery suggest that Scott’s and Souza-Dias’s theory is correct since we found posterior fixation sutures to be effective in decreasing the upshoot and downshoot in patients with Duane syndrome, types I and III\(^2\) (Fig. 21–7B). However, we have found that the application of posterior fixation may be technically difficult in these patients because of mechanical restriction of adduction. Recession of both horizontal muscles is equally effective and technically simpler to perform.\(^3\) Moreover, it has the added desirable

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**FIGURE 21–7.** A, Preoperative findings in bilateral Duane syndrome, type I, of both eyes with upshoot and downshoot of the left adducted eye. Note that this phenomenon occurs only when the fixating right abducted eye is slightly elevated or depressed. B, Postoperative findings after retroequatorial myopexy of the left medial (14 mm) and lateral rectus (17 mm) muscles. Note significant decrease of upshoot and downshoot of the adducted left eye.
effect of reducing retraction in adduction. It has also been suggested that the lateral rectus tendon be split into a Y configuration that increases the width of insertion and decreases the bridle effect.\textsuperscript{185, 311}

Much emphasis was placed in the past on the high prevalence of amblyopia and anisometropia in patients with Duane syndrome. This concern originated from the original paper of Duane\textsuperscript{96} and a report by Kirkham\textsuperscript{199} that amblyopia was present in 40\% of his patients. However, more recent studies have shown that the prevalence of anisometropia in Duane syndrome is no higher than in a normal population.\textsuperscript{232, 267, 310, 373}

Another feature of the retraction syndrome that deserves the special attention of the clinician is its frequent association with other ocular lesions and systemic congenital malformations. Among the ocular anomalies listed by Sachsenweger\textsuperscript{320}, p.283 are dysplasia of the iris stroma, pupillary anomalies, cataracts, heterochromia, persistent hyaloid arteries, choroidal colobomas, distichiasis, crocodile tears, microphthalmos, and many others. Numerous systemic anomalies have been described.\textsuperscript{99, 289, 320} Among these are Goldenhar’s syndrome,\textsuperscript{289, 290, 382} facial hemiatrophy,\textsuperscript{317} dystrophic defects such as the Klippel-Feil syndrome, arthrogryposis multiplex congenita,\textsuperscript{240} cervical spina bifida, cleft palate, sensorineural hearing deficits,\textsuperscript{198, 309} Chiari’s type I malformation,\textsuperscript{289, 410} deformities of the external ear, and anomalies of the limbs, feet, and hands.

These findings emphasize that a thorough ocular examination is mandatory in all patients with a retraction syndrome and that systemic malformations, especially hearing defects, must be ruled out by a general physical examination in each case.

\textbf{Therapy}

The results of surgical treatment of the retraction syndrome often have been disappointing. For this reason, we prefer not to operate when binocular vision is present with the eyes in primary position or if it can be maintained with only a slight head turn. Surgery is indicated only when there is a significant deviation in primary position or if the face turn is intolerable from a cosmetic or functional point of view. The patient must be informed that there is no surgical procedure that will restore normal ocular excursions in all gaze positions. When esotropia in the primary position causes a significant face turn in Duane syndrome, type I, we perform a 5-mm recession of the ipsilateral medial rectus muscle (see also Kaufmann and Mil-kowitz\textsuperscript{192}). To decrease retraction on adduction a recession of the lateral rectus muscle may be added.\textsuperscript{100, 222} The recessions must be asymmetrical to counteract the esotropia in primary position and the face turn.\textsuperscript{152}

Although these operations cannot be expected to improve motility in abduction in patients with Duane syndrome, type I, they are highly successful in eliminating the anomalous head posture.\textsuperscript{296} The patient should be informed that this operation is likely to further restrict ocular motility by decreasing adduction. A posterior fixation applied to the contralateral medial rectus muscle\textsuperscript{220, 327, 350} or both horizontal rectus muscles\textsuperscript{349} is said to improve comitance. We have not been impressed with the results of this combined surgical approach and no longer use it.

Several authors have recommended transposing the vertical rectus muscles to the insertion of the lateral rectus muscle in Duane syndrome, type I. Gobin,\textsuperscript{128} in reporting his results in 67 patients, noted an average improvement of abduction of 20\°, but limitation of adduction occurred in most of his patients. Molarte and Rosenbaum\textsuperscript{246} reported improvement of esotropia, abduction, head turn, and field of single vision. To avoid the complication of a surgically induced vertical strabismus, this procedure may be modified by putting the vertical muscles on adjustable sutures.\textsuperscript{214} Foster\textsuperscript{220} augmented the transposition by fixating the transposed muscles with nonabsorbable sutures 16 mm from the limbus and adjacent to the lateral rectus muscle insertion. Esotropia in primary position and face turn disappeared in most patients and he recorded the same improvement of abduction as Gobin did but, curiously, did not encounter an adduction deficit of the operated eye. One of us (E.C.) has observed fixation of the globe in abduction after the transposition procedure for Duane syndrome, type I.

Clearly, this approach, with or without scleral fixation of the transposed muscles, deserves further study since an isolated recession of the medial rectus muscle has no effect on the diplopia occurring when the involved eye attempts to abduct.

Resection of the lateral rectus muscle in Duane syndrome, type I should be avoided under any circumstances since this may increase retraction of the globe on adduction. In patients with an extreme head turn caused by Duane retraction syndrome, type I, de Decker\textsuperscript{87} recommended a
muscle transposition procedure on both eyes according to Kestenbaum.

For exotropia associated with the retraction syndrome, Papst and Stein recommended recession of the lateral rectus muscle of the involved eye or recession of the lateral rectus muscle and resection of the medial rectus muscle in the noninvolved eye. This approach has worked well in our hands.

A special therapeutic challenge exists in patients with Duane syndrome, type III (limitation of adduction and abduction), who are esotropic with the involved eye in abduction and exotropic with the involved eye in adduction and who maintain an anomalous head posture. Spielmann and coworkers reported good results with a posterior fixation suture, with or without recession of the horizontal rectus muscles of the sound eye in these cases. We mentioned earlier in this chapter the beneficial effect of recession of both horizontal rectus muscles on up- and downshoot of the adducted eye and on the retraction in Duane syndrome.

It is clear from the preceding paragraphs that no rigid rules exist regarding surgical treatment of the retraction syndrome and that an individualized approach taking into account coexisting horizontal or vertical deviations is necessary.

Brown Syndrome

In 1928 Jaensch described limitation of elevation of the adducted eye after a skiing accident in which the patient’s face struck the tip of a ski. The clinical picture resembled a paralysis of the inferior oblique muscle but the forced duction test showed resistance to elevation of the adducted eye. Jaensch suspected a traumatic adhesion between the trochlea and the globe anterior to or at the equator. Such an adhesion would not interfere with depression of the globe but would present an obstacle to elevation in adduction. Thus Jaensch was first to call attention to a pseudoparesis of the inferior oblique muscle as a result of acquired structural anomalies (see also Hass and Stein) involving the superior oblique tendon. In 1950 Brown described an identical anomaly of ocular motility which, unlike the case of Jaensch’s patient, occurred on a congenital basis. He suspected the existence of a congenitally short superior oblique tendon sheath in a patient who could not elevate the adducted eye and in whom elevation in adduction was restricted during the forced duction test. During surgery the sheath was isolated from the tendon, and tension on the sheath was demonstrated easily during passive elevation of the globe. When the sheath was severed, tension was no longer present.

Since Brown’s original descriptions in the 1950s, it has become clear, however, that there are many anomalies involving the superior oblique muscle, its tendon and surrounding tissue, or the trochlea that may contribute to a mechanical restriction of elevation of an adducted eye. For this reason the older term “superior tendon sheath syndrome” has been abandoned in favor of Brown syndrome although in light of the historical facts the name Jaensch-Brown syndrome has also been suggested. Brown syndrome consists of consistent and variable features which are listed in Table 21–2. A patient with Brown syndrome is shown in Figure 21–8.

Incidence, Laterality, and Heredity

The incidence of Brown syndrome in a strabismic population is low; in a review of 2583 consecutive strabismus patients, Crosswell and Haldi encountered only six cases. The syndrome is usually unilateral but may occur in both eyes in about 10% of the cases. Familial occurrence has been described and mirror reversal was observed in monozygotic twins. Calceldra reviewed the literature on twin studies and reported the syndrome in dizygotic female twins. The syndrome may present in a congenital, acquired, constant, or intermittent form. Despite Brown’s original impression that the condition occurs more often in females and in the right eye, subsequent reports have failed to substantiate a sex or laterality predilection.

<table>
<thead>
<tr>
<th>Consistent Features</th>
<th>Variable Features</th>
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<tbody>
<tr>
<td>Absence of elevation in adduction</td>
<td>Mild limitation of elevation from primary position</td>
</tr>
<tr>
<td>Normal elevation in adduction</td>
<td>Downshoot in adduction</td>
</tr>
<tr>
<td>Forced ductions show severe mechanical restriction on attempts to elevate the adducted eye; no limitations of elevation in adduction</td>
<td>Widening of lid fissure</td>
</tr>
<tr>
<td>Hypotropia in primary position</td>
<td>Compensatory head posture</td>
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</table>
Associated Anomalies

Brown syndrome occurs, as a rule, as an isolated anomaly. However, exceptions have been reported, including an association with superior oblique palsy, dissociated vertical deviation, and contralateral inferior oblique overaction. A patient with Hurler-Scheie syndrome developed an acquired Brown syndrome, and other anomalies listed in the comprehensive review on Brown syndrome by Wilson and coworkers include crocodile tears, Marcus Gunn synkinetic movements, coloboma of the choroid, and congenital cardiac anomalies. The infrequent occurrence of associated anomalies raises the distinct possibility that these are coincidental findings rather than true associations.

Natural History

Spontaneous remission of an apparently congenital Brown syndrome was reported by several authors. Spontaneous resolution occurred in 6 of 60 patients and significant improvement in another 6 patients after an average follow-up of 46 months. Even more impressive are the results of a longitudinal long-term study by Gregersen and Rindziuski: of 10 patients with Brown syndrome diagnosed during the first 2 years of life and followed for an average of 13 years, 3 patients had complete recovery of normal ocular motility. The remaining patients had a decrease of the hypotropia and depression of the adducted eye. In this connection, it is of interest that this syndrome is infrequently observed in adults. Of the 126 patients described by Brown, only 14 were 13 years of age or older.

Etiology

As has been mentioned above and will be further discussed in the following paragraphs, mechanical limitation of elevation in adduction has more than one cause. For this reason Helveston suggested a generic description of the syndrome according to which the inability to elevate the adducted eye is due to a failure to increase the distance between the trochlea and the superior oblique tendon insertion. He further described the complex machinery consisting of muscle, tendon, and trochlea which is especially vulnerable to developmental and acquired defects.

ANOMALIES OF TENDON SHEATH. Brown described the mechanism as follows:

The tendon sheath of the superior oblique, according to Whitnall, is fixed to and terminates at the pulley. The effect of the two fixed points, the attachment of the sheath at the pulley and its attachment at the scleral insertion of the fused tendon and sheath, results, if the tendon is short, in restriction of elevation in the nasal field. The linear distance from the pulley to the insertion of the superior oblique muscle increases on adduction of the eye and decreases on abduction. Therefore if the tendon sheath is taut when the eye is in the primary position, adduction will be possible only as the eye is depressed. Normally this sheath acts as a check-ligament for the inferior oblique muscle.

Parks observed that the superior oblique tendon does not have a sheath at all and that the term “superior tendon sheath syndrome” introduced by
Brown is a misnomer. Parks reasoned that the tissue surrounding the tendon is actually a sleeve consisting of Tenon’s capsule and that this sleeve may have been mistaken by Brown for a sheath.

We do not argue that what has been described by Fink and by Berke as a sheath may not be a sheath after all. However, we cannot agree that this tissue, whatever its nature may be, is altogether blameless for causing restriction, as current authors would have it. Although we no longer use this method, surgical stripping of the sheath or pseudosheath, leaving the tendon proper intact, has relieved the restriction in some cases of Brown syndrome operated on by us. The normalization of the forced duction test after this tissue was dissected was often dramatic. Brown achieved full correction of the motility defect in 5 of 26 patients operated on in this manner.

Soon after Brown’s original description it became apparent, however, that stripping of the peritendinous tissue was not always effective in relieving the restriction of ocular motility.

Ophthalmologists had also become aware that the syndrome occurred in an acquired and intermittent form or resolved with the passage of time. These features are incompatible with a congenital anomaly of the tendon sheath. In 1973, therefore, Brown began to distinguish between a true and a simulated syndrome. The true syndrome includes only those patients who have a congenitally short anterior tendon sheath. A simulated sheath syndrome with identical clinical features that includes all acquired cases may be caused by other factors such as anomalies of the tendon itself or of the trochlea.

ANOMALIES OF TENDON OR TROCHLEA.

Thickening of the muscle entering the trochlea, trauma, or inflammatory changes involving the trochlea or the region adjacent to it; developmental structural anomalies of the trochlea; and congenital or iatrogenic shortening of the superior oblique tendon are some of the factors to be considered in the etiology of Brown syndrome.

TIGHT TENDON. Crawford proposed that the cause of a “true” Brown syndrome is a tight tendon and reported excellent surgical results after cutting the tendon just medial to the superior rectus muscle rather than merely cutting the sheath (see also Jakobi). Parks made similar observations and the uniformly good results reported after tenectomy of the superior oblique support the current view that most cases of constant congenital Brown syndrome are caused by an abnormal tightness of the muscle-tendon complex. Spontaneous limitation of elevation in adduction in the wake of an acquired superior oblique paralysis, presumably caused by a fibrotic tendon, has been discussed in Chapter 20.

IMPAIRED SLIPPAGE OF THE TENDON THROUGH THE TROCHLEA. Girard reported a patient with Brown syndrome in whom, on repeated attempts to elevate the eye in adduction, sudden release of the restriction occurred and full motility of the globe was restored. He stated that a congenital anomaly of the tendon may have been the cause of the restriction. Similar instances of intermittent Brown syndrome were reported by other authors, and we have observed patients with Brown syndrome who were unable to elevate the eye from a position of adduction, but who had no restriction of ocular motility when the eye was first elevated in adduction and then, while maintaining the elevation, moved nasalward into adduction. We have seen others who relieved vertical diplopia caused by Brown syndrome by applying digital pressure to or massaging the region of the trochlea. Leone followed a patient with apparently constant congenital Brown syndrome. After 15 years of observation the patient began to exercise elevation of the adducted eye and was eventually able to do so with an audible click. Roper Hall reviewed 18 cases of what he termed the superior oblique click syndrome (see also Folk et al.). Obviously, the mechanism of these intermittent forms of Brown syndrome must be an impairment of slippage of the tendon through the trochlea caused by a retrotrochlear thickening of the tendon or anomalies of the trochlea itself.

That impaired slippage of the tendon through the trochlea may also occur on an acquired basis was shown by Stein, who reported a patient in whom typical features of Brown syndrome developed after a blunt injury to the eye. Surgical exploration revealed a blood cyst in the sheath of the superior oblique tendon. After removal of this lesion, function returned to normal. Cysts in the reflected tendon were also observed by Helveston who reported resolution of Brown syndrome after their dissection.

Sandford-Smith suggested that hypertrophy and constriction of the trochlea and tendon sheath associated with localized swelling of the tendon (stenosing tenosynovitis) may cause Brown syndrome, and acquired Brown syndrome, usually
with spontaneous reversal, has been reported in children\textsuperscript{391} and adults\textsuperscript{20, 196, 341} as a manifestation of rheumatoid arthritis or pansinusitis.\textsuperscript{326} MRI in such patients shows fibrosis of the superior oblique tendon, apparently thought to be caused by chronic inflammation.\textsuperscript{371}

Beck and Hickling\textsuperscript{20} suggested treating such patients with \textit{local injections of corticosteroids}. Wright and coworkers\textsuperscript{409} reported negative findings on pathologic examination of the superior oblique tendon, trochlea, and superior oblique muscle in a patient with acquired inflammatory Brown syndrome and an audible click on elevating the adducted eye. We have followed several patients with acquired Brown syndrome. Among the more unusual ones were a 43-year-old man with episcleritis who developed a transient inability to elevate the adducted eye.\textsuperscript{262} Forced duction tests were positive, and spontaneous remission occurred after 2 weeks. Typical Brown syndrome developed in a second patient during her third month of pregnancy. No improvement of her condition occurred during 3 years of observation. Postpartum acquired Brown syndrome has also been reported\textsuperscript{65} and a comparison with the carpal tunnel syndrome comes to mind.

\textbf{DEVELOPMENTAL ANOMALIES OF THE TROCHLEA.} Helveston and coworkers\textsuperscript{158, 161} studied the anatomy and physiology of the human trochlea in cadaver specimens and described a bursa-like structure between the vascular sheath of the tendon and the trochlear saddle. They proposed that excess fluid accumulation or concretion in this bursa-like space could cause limitation of movement through the trochlear tunnel, causing an acquired Brown syndrome. Wilson and coworkers\textsuperscript{398} suggested that if the telescoping movement of the tendon described by Helveston and his group were interfered with by an intrinsic anomaly of the trochlea or the tendon, Brown syndrome would result.

Sevel,\textsuperscript{334} in a developmental study of the extraocular muscles, raised the interesting possibility that persistence of embryonic trabecular connections between the superior oblique tendon and the trochlea may account for Brown syndrome. Under normal circumstances, only fine remnants of these trabeculae remain and act as tethering strands to control and limit excursions of the tendon in the trochlea.\textsuperscript{335}

\textbf{ANOMALY OF THE SUPERIOR OBLIQUE MUSCLE.} Gradual onset of a pseudo-Brown syndrome occurred in a 62-year-old man who developed a metastasis from a carcinoma of the prostate to his superior oblique muscle.\textsuperscript{33}

\textbf{ANOMALIES OF INFERIOR OBLIQUE MUSCLE AND ADJACENT STRUCTURES.} Girard\textsuperscript{127} described a patient with all the features of Brown syndrome in whom dissection of the superior oblique tendon sheath had no effect; however, after a dense fibrous attachment was severed that extended from the insertion of the inferior oblique muscle to the lateral wall of the orbit, all resistance to passive elevation of the globe in adduction disappeared. Zipf and Trokel\textsuperscript{412} observed two patients with restricted elevation in aduction following a blow-out fracture of the orbital floor. Surgical exploration revealed incarceration of the inferior orbital tissue in the fracture site.

\textbf{PARADOXICAL INNERVATION.} Analogous to the findings in Duane retraction syndrome is the discovery of Papst and Stein,\textsuperscript{278} who, in two patients with Brown syndrome, demonstrated paradoxical innervation of the superior oblique muscle on attempts to elevate the eye. Feric-Swiwerth\textsuperscript{114} reported similar results in one patient in whom simultaneous electromyographic recordings were obtained from the superior and inferior oblique muscles. However, these findings could not be confirmed by Catford and Hart,\textsuperscript{62} who demonstrated electric silence on recording from the superior oblique muscle and maximal activity from the inferior oblique muscle in patients who attempted elevation in adduction. Moreover, if this mechanism would prevail in Brown syndrome, one would expect the forced duction test to become negative with the patient under anesthesia. This is never the case in patients with true Brown syndrome.

\textbf{POSTOPERATIVE: AFTER SUPERIOR OBLIQUE TUCKING.} Girard\textsuperscript{127} and Hervouet and Chevannes\textsuperscript{62} each described the appearance of Brown syndrome as a result of their tucking the superior oblique tendon. We have observed this complication in several patients after the tucking operation. Spontaneous improvement occurred in some but not all of them, and persistent diplopia in upward gaze may become a significant problem. Excessive tucking can be avoided by performing a forced duction test at the end of the procedure. The tuck must be undone when there is more than a moderate elastic resistance to elevation of the adducted eye. We believe that another cause of a postopera-
tive Brown syndrome is suturing the tucked tendon back to the sclera. Since we stopped doing this we have no longer encountered this complication.

**TRAUMA.** As mentioned above direct trauma to the trochlear region can cause restriction of elevation in adduction, often combined with restriction of depression. The “canine tooth syndrome” of Knapp\(^\text{201}\) falls into this category and numerous other cases of traumatic Brown syndrome have been reported.\(^\text{398}\) Surgical trauma has emerged as another cause of Brown syndrome. We have observed one case after cosmetic blepharoplasty and another patient developed the syndrome after a double-plate Molteno implant.\(^\text{93}\)

**SECONDARY TO PARALYSIS OF THE INFERIOR OBLIQUE MUSCLE.** There are those who believe that fibrosis of the sheath of the superior oblique tendon is the result of primary paralysis of the inferior oblique muscle.\(^\text{274, 380}\) This concept cannot be upheld in view of the postoperative finding of normal elevation in adduction, of a normal electromyographic innervational pattern of the inferior oblique muscle,\(^\text{40, 62}\) and of normal saccadic velocity in patients with Brown syndrome.\(^\text{238}\)

**Diagnosis and Differential Diagnosis**

The clinical manifestations of Brown syndrome are listed in Table 21–2. The direction of a head tilt, if present, and the position of the chin are similar as in paralysis of the inferior oblique muscle: the chin is lifted and the head is tilted toward the involved side. Chin elevation, as a rule, is more pronounced than the head inclination. Diplopia, a frequent complaint of patients with this syndrome, usually can be elicited when the involved eye is adducted. Diplopia in the primary position is often avoided by anomalous head posture. Suppression is rare, and we have observed only one patient who had ambylopia of the involved eye.

The differential diagnosis should include primarily a paralysis of the inferior oblique muscle. The positive forced duction test clearly differentiates Brown syndrome from a paralysis of the inferior oblique muscle. Furthermore, depression of the adducted eye, normal action of the contralateral superior rectus muscles, and a V pattern in upward gaze are features that are frequently found in association with Brown syndrome. On the other hand, in inferior oblique paralysis one may expect an A pattern because of loss of the abductive action of the paralyzed muscle in upward gaze.\(^\text{255}\)

The ultimate distinction between Brown syndrome and inferior oblique muscle paralysis rests with the outcome of the *forced duction* test. The ophthalmologist should be aware of other causes of restriction of elevation, such as congenital fibrosis of the inferior rectus muscle, endocrine orbitopathy, so-called double elevator paralysis, or fractures of the orbital floor. However, unlike in Brown syndrome, these conditions usually cause restriction of elevation from any gaze position and are not limited to restriction of elevation in adduction. Exceptions to this rule do occur and should be kept in mind; both an orbital floor fracture\(^\text{180, 412}\) and endocrine orbitopathy may occasionally simulate Brown syndrome.\(^\text{130, 174}\)

**Therapy**

When binocular vision is normal and comfortable with the eye in primary position and without an extreme anomalous head posture, we do not advocate surgery. Such patients may experience diplopia on attempts to elevate the involved eye in adduction, but they will learn to avoid this position of gaze. Downshoot in adduction alone does not present an indication for surgery. On the other hand, when the involved eye is hypotropic in primary position, when there is a significant anomalous head posture or, if traumatic in origin, does not spontaneously resolve, surgery should be considered in an attempt to restore binocular function in primary position. Another indication for surgery exists in the absence of binocularity in patients who habitually fixate with the involved eye and develop an anomalous head posture.

As mentioned, the results of dissecting and stripping the sheath while leaving the tendon intact, as originally advocated by Brown, have been unsatisfactory in the majority of cases. We perform a *complete tenectomy of the superior oblique muscle*,\(^\text{265, 353, 357}\) as advocated by Jacobi\(^\text{181}\) and by Crawford\(^\text{76}\) and Crawford and coworkers.\(^\text{77}\) The immediate result of this operation is often dramatic: the forced duction test becomes negative, and the eye elevates freely in adduction. However, in some patients the effect of surgery does not become fully apparent until several weeks or months later. In one half of 38 patients treated by us in this manner\(^\text{557}\) and followed for more than 1 year, the classic features of a *superior oblique paralysis* developed after tenectomy of the supe-
rior oblique. This problem has, in our hands, responded well to a subsequently performed recession of the contralateral inferior rectus muscle or myectomy or recession of the ipsilateral inferior oblique muscle but other authors have encountered complications arising from attempts to repair the iatrogenic superior oblique paralysis.

In the other half of our patients superior oblique paralysis did not occur, which is the reason why we do not perform a superior oblique tenectomy combined with recession of the inferior oblique muscle at the time of initial surgery, as was advocated by several authors. In our series there was no loss of sensory function as a result of surgery, which has been mentioned as a surgical complication of tenectomy by Wright and coworkers.

Wright introduced a silicone superior oblique tendon expander to lengthen the tendon rather than cutting it. He reported superior results with this method when compared with tenectomy. Others have also reported favorable results with the silicone expander technique in Brown syndrome. For reasons outlined in Chapter 26 we see no reason to abandon the superior oblique tenectomy, which in our hands has proved to be a very simple, safe, and effective procedure in treating Brown syndrome. In patients with acquired Brown syndrome a conservative approach is indicated unless a surgically amenable cause, such as a cyst or trauma to the trochlea, can be identified. As mentioned, spontaneous resolution is not infrequent and corticosteroid injections near the trochlea in patients with juvenile rheumatoid arthritis or other inflammatory conditions in this region are of possible benefit.

### Adherence Syndrome

Several investigators have described developmental anomalies of extraocular muscles in which the sheaths of the lateral rectus and inferior oblique muscles and those of the superior rectus and oblique muscles are adherent. Such anomalies are rare, but the ophthalmologist must be aware of their existence, for they may produce perplexing diagnostic problems.

Johnson described several variations of the adherence syndrome. Abnormal fascial connections between the lateral rectus and the inferior oblique muscles will cause pseudoparalysis of the lateral rectus muscle. After the insertion of the lateral rectus muscle is severed from the sclera, resistance to passive abduction will be apparent during the forced duction test.

Similarly, adherence between the superior rectus muscle and the tendon of the superior oblique muscle may cause pseudoparalysis of the superior rectus muscle. After the lateral rectus or the superior rectus muscle has been temporarily detached from the globe, Johnson recommends lysis of these adhesions. This is accomplished by forcefully rotating the eye medially with forceps for the lateral adherence syndrome or downward for the superior adherence syndrome.

Johnson’s description of the adherence syndrome leaves the impression that it occurs relatively often. Though we routinely use the forced duction test before, during, and after surgery, we have never observed a vertical adherence syndrome, and in only two patients have we seen the horizontal form. One of these patients had previous surgery of the lateral rectus muscle with resulting massive scar formation that extended posteriorly and involved both lateral rectus and inferior oblique muscles. In view of the scarcity of additional descriptions of this anomaly after Johnson’s initial report and the infrequency with which it is encountered in clinical practice, we believe its significance has been overemphasized.

Parks used the term adherence syndrome to describe a complication of myectomy of the inferior oblique muscle that he believed was caused by proliferation of fibrofatty tissue and reattachment of the proximal muscle stump to Tenon’s capsule. Patients with this syndrome had hypotropia in primary position, restricted elevation, and a positive traction test. Parks encountered this complication in 13% of patients in whom myectomy was performed at the insertion and in 26% with disinsertions of the inferior oblique muscles. One of us (G.K.v.N.) has performed myectomy of the inferior oblique as the preferred weakening procedure of this muscle for the past 40 years and has encountered this complication in only two instances. Clearly, variations in the surgical technique must be responsible for this difference in prevalence.

### Strabismus Fixus

#### Clinical Findings and Etiology

Strabismus fixus is a rare condition in which one or both eyes are anchored, as a rule, in a position...
of extreme adduction (Fig. 21–9). The involved eye is “fixed” in this position and cannot be moved, and the forced duction test will confirm the immobility of the eye.

Divergent forms of strabismus fixus not accompanied by ptosis or generalized fibrosis of the extraocular muscles are even more unusual\(^{125}\) (Fig. 21–10) and enlargement of the nasal field of vision has been described in a case of divergent strabismus fixus.\(^{330}\) Patients with bilateral strabismus fixus are severely handicapped. One eye usually is preferred for vision, and the immobility of the eyes necessitates an extreme degree of head turn for such patients to get around. Strabismus fixus may occasionally reach bizarre proportions as shown by Case 21–1.

**CASE 21–1**

A middle-aged man complained that the colored part of his left eye had “gone.” He remembered that the eye had always been turned toward the nose and that the iris had gradually disappeared from view. Examination showed absence of the cornea in the palpebral fissure (Fig. 21–11A) and a CT scan revealed that the eye had rotated inward approximately \(135^\circ\), almost exposing the optic nerve in the palpebral fissure (Fig. 21–11B). No treatment was advocated.

The condition generally is thought to be congenital and caused by fibrosis, which would explain the loss of elasticity of the medial rectus muscle. Acquired strabismus fixus was described by Villasecca\(^{383}\) and Martinez.\(^{230}\) Villasecca postulated that fibrosis of the medial rectus muscle is the consequence of contracture following a lateral rectus paralysis rather than a primary anomaly. Mechanical stretching and torsion of the optic nerve with strangulation of its blood supply has been reported to cause ocular ischemia, optic atrophy, and central artery occlusion in convergent strabismus fixus.\(^{228}\) Acquired strabismus fixus has also been reported in conjunction with amyloidosis\(^{336}\) and in patients with high myopia.\(^{26, 176, 306}\) An unspecified, progressive fibrosis, myopathy, or myositis is thought to be the cause of myopic strabismus fixus, which usually occurs in a convergent form and may reach extreme proportions.\(^{294}\)

**Therapy**

Treatment is surgical and, for the convergent form of strabismus fixus, consists of complete disinsertion of the medial rectus muscle(s). In addition, resection of the lateral rectus muscles and recession of the conjunctiva and Tenon’s capsule should be carried out. Even though abduction beyond the midline will not become possible after surgery, some cosmetic and functional improvement can be accomplished in this manner. Under optimal conditions, a small field of single binocular vision can be restored by surgically moving the eyes into primary position.
Strabismus in High Myopes

Hugonnier and Magnard (1969) were first to direct attention to restrictive motility disturbances in severe myopia which they believed was caused by an unspecified myositis. Since then it has become obvious that progressive strabismus occurring in high myopes may be due to more than one mechanism. One cause is a disproportion between the size of the orbit and the volume of an enlarged or elongated myopic globe. This may cause a generalized restriction of motility in several gaze directions and masquerade as endocrine orbitopathy, as shown by Case 21–2.89

Case 21–2

A 45-year-old woman wore lenticular spectacles for a myopia of OD −28.00 +2.50 × 125° and OS −28.00 +0.75 × 165°, which corrected her visual acuity to 6/15 OD and 6/12 OS. Nine years ago she noted diplopia on tilting her head to the left. A left inferior oblique myectomy was performed elsewhere without relieving her symptoms. She now complains about vertical diplopia in all gaze positions with an insidious onset. Her motility examination showed comitant left hypertropia of 10° which increased to 16° when looking down and to the left. Abduction was limited OD and elevation in adduction was limited OS (Fig. 21–12). Forced duction testing showed mechanical restriction of ocular motility in all peripheral gaze positions. Because of her ocular motility pattern, the results of the forced duction tests, and periorbital edema, a thyroid evaluation was performed that was essentially negative. Fundus examination showed vertically elongated and tilted optic nerve heads, peripapillary loss of pigmentation, and myopic retinal changes. A B-scan echogram demonstrated elongation of OS with a diffuse posterior, conical staphyloma. CT (Fig. 21–13A) showed marked enlargement and elongation of the globes, which were nearly filling the orbits, and diffuse posterior staphylomas. A coronal view of the orbit (Fig. 21–13B) showed a normal cross-sectional appearance of the extraocular muscles. Surgery consisted of a 4-mm recession of the left inferior rectus muscle, using an adjustable suture anchored to the globe by passage through a residual muscle stump to avoid perforation of the sclera. The patient has remained free of vertical diplopia.

We felt that the restriction of ocular motility was caused by contact between the elongated globes and the orbital walls and apices of the...
orbits. Bagolini and coworkers\textsuperscript{15} suggested that compression of the lateral rectus muscle against the lateral orbital wall by an enlarged myopic globe will cause esotropia. Depending on the degree of esotropia surgery may be indicated to facilitate fundus examination and allow the patient to wear contact lenses. The medial rectus muscle or muscles must be retroplaced maximally (up to 13 mm) and a hang-back suture is used to avoid having to place the suture through the sclera that far posteriorly.

Another form of restrictive strabismus in high myopes has become known as the heavy eye syndrome. This syndrome consists of a slow and progressive development of esotropia and hypotropia. A typical case is depicted in Figure 21–14. Since the original description by Bagshaw\textsuperscript{16} numerous additional cases have been reported\textsuperscript{13, 16, 73, 154, 205, 216, 317, 368} and the results of neuroimaging studies have become available. Herzau and Ioannakis\textsuperscript{163} noted at the time of operation a downward displacement of the lateral rectus muscle in progressively myopic eyes with the heavy eye syndrome. They postulated that scleral ectasia in these patients causes a downslop of the muscle in relation to the globe. This change of muscle path gives the muscle a depressing effect at the cost of its physiologic action of abduction. Krzizok and coworkers\textsuperscript{209–211} confirmed this change of muscle path with MRI in 33 orbits and noted also that the lateral rectus muscles may not be the only muscle involved; in two patients with exotropia and hypotropia the medial rectus muscle path was displaced inferiorly. Scan echography showed no differences in muscle size between normal and myopic patients\textsuperscript{291}.

To normalize the muscle path Herzau and Ioannakis\textsuperscript{163} recommended adding a supratransposition to a resection of the lateral rectus in combination with a recession of the medial rectus muscle. In several of their cases this operation was augmented with scleral fixation of the transposed muscle with a silicone loop to counteract its tendency to slip back inferiorly (see also Krzizok et al\textsuperscript{212}). The results of muscle transposition for this condition have been disappointing in the hands of one of us (E.C.).

Fibrosis of the Extraocular Muscles

A rare congenital, familial, or sporadic anomaly involving fibrosis of most or all of the extraocular muscles was described by Heuck in 1879\textsuperscript{165} This condition is characterized by the following features:

1. Downward fixation of one or both eyes
2. Marked ptosis
3. Chin elevation
4. Perverted convergence movements on attempts to look upward or to either side
5. Familial or sporadic occurrence

Histologic examination of excised tissue from the extraocular muscles in patients with this condition reveals total replacement of muscle fibers by
fibrous elements. CT shows marked atrophy of the inferior rectus muscle. Assaf pointed out that these histologic anomalies may be secondary to an innervational disturbance rather than a primary ocular myopathy as the term fibrosis syndrome suggests and raises the possibility that a supranuclear disturbance may be present in these patients. Engle and coworkers described a family with chromosome 12–linked congenital fibrosis of the extraocular muscles and fixation of the eyes in downward gaze. Autopsy findings in one affected member showed absence of the superior division of the oculomotor nerve and its corresponding alpha neurons as well as histologic abnormalities of the levator palpebrae and superior rectus muscles. The other extraocular muscles showed abnormal mitochondrial clumping, indicating that muscles other than those innervated by the superior division of the third cranial nerve are also affected. This report suggests that congenital fibrosis of the extraocular muscles may be caused by an abnormality of the lower motor neuron system.

Familial occurrence has been described by many authors and other systemic congenital defects may be present in the affected and nonaffected family members. Gillies and coworkers described a family with dominantly inherited total absence of vertical ocular movements and found a reduction in size of cross sections of the extraocular muscles on CT scans with the superior rectus muscle particularly involved.

We treated a 21-year-old patient who had generalized fibrosis of the extraocular muscles associated with keratoconus and arthrogryposis. Her deceased father had a similar condition. The possibility that a vertical retraction syndrome may be caused by fibrosis of the vertical rectus muscles has been mentioned (see p. 462). Congenital ocular fibrosis associated with the Prader-Willi syndrome has been reported, as has the association with oculocutaneous albinism, synergistic divergence, jaw-winking.

We have been impressed by the high degree of hypermetropic astigmatism and amblyopia in these patients, an observation also made by Sugawara and coworkers. Harley and coworkers reviewed the literature pertaining to ocular fibrosis.

Figure 21–15 shows a family with ptosis and generalized fibrosis of the extraocular muscles. Even though an autosomal dominant pattern of inheritance may be present, the sporadic form is the most common form encountered in our practice.

In making the differential diagnosis the physician must include orbital floor fracture, endocrine myopathy, Brown syndrome, double elevator palsy, and chronic progressive external ophthalmoplegia, all of which are discussed in this chapter.

FIGURE 21–15. Congenital fibrosis of extraocular muscles. The mother (left) had frontalis suspension procedures to both upper lids. Note chin elevation of son and daughter caused by severe ptosis. Motility of the globes was severely restricted in all members of this family. (Courtesy of Dr. Robert M. Feibel, St. Louis, MO.)
Treatment consists of a complete tenotomy to release the tight inferior rectus muscle. The results are never completely satisfactory, and the best a surgeon can hope for is to improve the compensatory head posture. Some patients need ptosis surgery. Since Bell’s phenomenon is not present, special care must be taken to avoid postoperative exposure problems. We prefer frontalis suspension of the upper lids with a nonabsorbable suture that can be adjusted postoperatively rather than fascia lata suspension of the lid for these patients. The surgeon should strive for undercorrection of the ptosis and lifting the lids just enough to expose the pupils. Since it is seldom possible to predict the position of the eyes after tenotomy of the inferior rectus muscle, we prefer to perform ptosis surgery as a separate procedure.

Graves’ Endocrine Ophthalmopathy

Etiology

Graves’ ophthalmopathy is part of a multiorgan autoimmune inflammatory disease that may cause periorbital edema, enlargement of the extraocular muscles, proptosis, lid retraction, optic neuropathy, and secondary increase of intraocular pressure. Limitation of ocular motility, most commonly a restriction of elevation in one or both eyes, is a prominent feature of Graves’ disease. A variety of terms are used to describe this condition: exophthalmic ophthalmoplegia, endocrine ophthalmopathy, endocrine orbitopathy, endocrine myopathy, dysthyroid eye disease, infiltrative ophthalmopathy, dysthyroid myositis, and exophthalmic goiter. This plethora of terms reflects our lack of knowledge of the exact nature of the relation between disturbance of thyroid function and involvement of the extraocular muscles.

It is of historical interest that before reintroduction of the forced duction test by Dunnington and Berke in 1943, the opinion prevailed that the limitation of elevation is caused by toxic damage to the elevator muscles, the superior rectus in particular.229 This view was perpetuated until the mid-1960s,320 when it was replaced by the current concept that limitations of ocular motility are caused by the swelling and loss of elasticity of the extraocular muscles. Dunnington and Berke were the first to recognize that pseudoparalysis of the superior rectus muscle in exophthalmic patients is caused by a myopathy and loss of elasticity of the inferior rectus muscle.

Further confirmation of the primary myopathic nature of ocular motility disturbances in patients with Graves’ disease has been provided by electromyographic studies.172, 225, 241, 331 Most of the clinical features of the disease can be traced to an overproduction of glycosaminoglycans within the orbit and histologic examination shows an accumulation of this substance in the connective tissue components of the orbital fat and extraocular muscles.58 Edema and inflammation of the muscles contribute to their swelling and dysfunction. It has been shown by length-tension curves during surgery that in the early stages of the disease muscle dysfunction is caused by increased tension and reduced elasticity.338 In the late stage of the disease the muscle fibers are replaced by connective tissue. Environmental and immunogenic factors probably contribute to the development of Graves’ ophthalmopathy and the reader is referred to recent review articles17, 388 and recent texts58, 387 for detailed discussion of current theories.

Diagnosis and Clinical Findings

Graves’ ophthalmopathy occurs more commonly in women than in men and usually affects middle-aged persons. The onset of diplopia usually is insidious and related closely to the onset of exophthalmos. Exceptions to this rule occur, however, and Bixenman and von Noorden described two patients in whom diplopia and restriction of motility developed practically overnight. We have observed several patients in whom exophthalmos developed after the onset of the motility defect or did not develop at all. Inconspicuous periorbital edema in association with limitation of elevation of one eye may be the first symptom of an endocrine myopathy. Limitation of elevation is by far the most common defect of ocular motility, followed in order of frequency by limitation of horizontal and vertical gaze, caused by myopathy of the medial and superior rectus, respectively. The lateral rectus muscle is least commonly involved. Limitation of ocular motility frequently remains
unilateral or is asymmetrical when myopathy affects both eyes. Saccadic eye movements in euthyroid Graves’ disease were found to be less conjugate than in those of control subjects and differences existed also in velocity characteristics of normals and patients with this condition.\textsuperscript{405}

Compression of the orbital apex by the enlarged extraocular muscles, especially the medial rectus muscle, may cause congestion of the optic nerve, axonal death, and decrease in visual acuity.\textsuperscript{113} However, pressure from the swollen muscles is not the only cause of optic neuropathy, which may also occur in the absence of muscle swelling.\textsuperscript{7}

Limitation of ocular motility, most commonly a restriction of elevation in one or both eyes and the optic neuropathy, has been shown to correlate with extraocular muscle volume as determined by CT scan.\textsuperscript{145} The possibility that initial unilaterality of the condition may be followed by affection of the other eye and renewed diplopia should be pointed out to the patient to avoid disappointment with the initial surgical result at a later date.

An increase in intraocular pressure on upward gaze, suggesting tightness of the inferior rectus muscle, has been used for many years\textsuperscript{36} as a diagnostic tool in the early stages of the disease. However, the value of this test has been questioned since pressure increase on upward gaze occurred also in normals.\textsuperscript{351}

Retraction of the upper lid is one of the many manifestations of Graves’ disease. It is usually caused by increased sympathetic innervation, although a shortening of the levator aponeurosis has also been implicated.\textsuperscript{142} The effort to elevate the eye in the presence of a tight inferior rectus muscle may contribute substantially to lid retraction (Fig. 21–16).

The diagnosis of endocrine myopathy is confirmed by the forced duction test, which reveals restriction of passive movements of the globe, and by a thorough evaluation by an internist. The frequent association between Graves’ disease and myasthenia gravis should be kept in mind. If the restrictive component in a patient with suspected Graves’ disease does not adequately explain the ocular motility deficit, an estimation of the generated muscle force and a Tensilon (edrophonium chloride) test are mandatory.\textsuperscript{53}

In the differential diagnosis one must consider other causes of restricted globe motility and endocrine ophthalmopathy has been reported to masquerade as a superior oblique paresis\textsuperscript{252} and Brown syndrome.\textsuperscript{175}

A CT scan is of great value, especially in distinguishing endocrine myopathy from other pathologic changes of the orbit that restrict ocular motility. Trokel and Hilal\textsuperscript{375} and Patrinely and coworkers\textsuperscript{287} outlined the variations and differential diagnosis of muscle thickening as seen on CT scans. The dramatic fusiform swelling of the extraocular muscles in a patient with endocrine myopathy is shown in Figure 21–17. As can be
seen by comparing Figures 21–17 and 21–19, the swelling in endocrine myopathy is limited to the posterior aspects of the muscle, whereas in ocular myositis it involves the anterior portions as well. Considering that the distribution of immunocompetent cells is fairly homogeneous throughout the length of the muscle, the predilection for this posterior site remains a mystery.

The more frequent involvement of the inferior rectus muscle may be related to the anatomy of the lower portion of the orbit. J. E. Miller and coworkers expressed the view that since the inferior rectus and inferior oblique muscles are the only muscles in direct contact with each other, any inflammatory process would lead to a fibrous union between these two muscles and the ligament of Lockwood. After confirming through surgery the existence of such anomalous connections between these structures, Miller and coworkers further pointed out that a similar inflammatory process may occur in other extraocular muscles but is less likely to cause symptoms since such changes would go unrecognized in symmetrical involvement of yoke muscles.

The relation between the onset of myopathy and thyroid dysfunction needs further clarification. Limitation of ocular motility may occur at any point in the continuum now recognized as Graves’ disease. In such patients, evaluation by an internist may reveal hyperthyroidism, euthyroidism, or even hypothyroidism. Of 120 patients with Graves’ disease 90% had hyperthyroidism, 1% had primary hypothyroidism, 3% had Hashimoto’s disease, and 6% were euthyroid. It is not uncommon to find the disease in patients who have had previous transient undiagnosed episodes of hyperthyroidism or in those who had undergone thyroidectomy several years before examination. Thus the ocular myopathy does not necessarily reflect the state of thyroid activity. The role of the long-acting thyroid stimulant (LATS) in the pathogenesis of endocrine myopathy is poorly defined at this time.

Many otherwise excellent studies by thyroidologists suffer from lack of a thorough ophthalmologic evaluation of ocular motility defects. We believe, for instance, that considerable confusion has been created by failure to differentiate between general limitation of ocular motility in several or all directions of gaze, usually accompanied by marked exophthalmos, in the acute “wet” congestive phase of the disease and the severe restriction of elevation, abduction, or adduction that is a feature of its more chronic, noncongestive “dry” phase. In the wet phase of the disease, the limitation of eye movement may be caused merely by marked swelling and congestion of the retrobulbar orbital contents. In the dry phase, limited eye movement is caused by actual infiltration and enlargement of the muscles and subsequent loss of elasticity.

**Therapy**

A discussion of the roles of systemic corticosteroids, immunosuppressive therapy, orbital radiation, or surgical decompression is beyond the scope of this book, and the reader is referred to the pertinent literature. We shall be concerned here only with treatment of strabismus caused by the myopathy.

Once the diagnosis of endocrine ocular myopathy has been established, it is advisable to await normalization of the endocrine imbalance by medical therapy before considering surgical correction. Spontaneous recovery of restricted ocular motility has been reported, but is rare in our experience. We prefer to observe such patients for at least 6 months to establish the stationary nature of the disease. On many occasions we have noted that during this observation period other muscles or muscle groups become involved so that a change in the surgical approach became necessary. Prismatic correction of vertical or horizontal deviations may be beneficial during this waiting period or unilateral complete or segmental occlusion be required when the deviation exceeds the amount correctable with prisms.

Other authors have advocated early intervention in patients who are severely disabled on account of their abnormal head position.

Systemic treatment directed at normalization of thyroid function in patients with hyperthyroidism, in our experience, has not been effective in eliminating the restriction of ocular motility. Brown and coworkers reported improvement in ocular motility in some patients following high dosages of corticosteroids. However, patients so treated suffered a severe, acute form of infiltrative Graves’ disease, and in their paper these workers made no distinction between the restriction of ocular motility caused by acute orbital congestion and that caused by myopathy. Even though steroids are known to dramatically improve the congestive phase of the disease, they have, in our
experience, no significant effect on the chronic form of endocrine myopathy.

Once the deviation has stabilized, surgery becomes the treatment of choice. The aim of surgery is to restore single binocular vision in those gaze positions that are functionally important to the patient. When elevation is limited, a 4- to 7-mm recession of the inferior rectus muscle is indicated.23 The amount of recession should exceed what is ordinarily recommended for vertical deviations (see Chapter 26). When adjustable sutures are used one should realize that an undereffect in the immediate postoperative phase may be only temporary and that a residual angle of strabismus tends to decrease further with time.109 Along the same line, it has been noted that overcorrections after initial postoperative alignment developed spontaneously in 5 of 12 patients.174 For these and other reasons discussed in the following paragraph, a slight undercorrection appears desirable. Adjustable sutures are well suited to deal with the relatively unpredictable results of a conventional recession in this condition but special care must be taken to anchor the sutures well in the sclera since the only two patients one of us (G.K.v.N.) has encountered in whom adjustable sutures became undone (3 and 10 days after surgery) had Graves’ ophthalmopathy. Helveston has observed the same complication in this condition.157

When recessing the inferior rectus muscle in patients with Graves’ ophthalmopathy the surgeon must be aware that while this operation in most instances restores single vision in primary position, it may cause a postoperative hypertropia in downward gaze that causes double vision. Depending on the patient’s occupational needs, an undercorrection should be strived for. A secretary or accountant, for instance, is better off with a mild residual chin elevation in primary position and fusion in downward gaze than with a normal head posture but diplopia during reading. If this complication is unavoidable, the appropriate surgery to restore fusion in downward gaze must be performed on the fellow eye in a second procedure. Another potential problem the patient should be advised of prior to surgery is asthenopia during near work. Progressive bifocal lenses may no longer work and a conventional bifocal segment may have to be repositioned.

When the inferior rectus muscle is recessed, freeing the muscle completely by sharp dissection from Lockwood’s ligament is not always possible because the extraordinary tightness of the muscle prior to its detachment prevents the necessary elevation of the globe. Postoperative retraction of the lower lid in a patient whose lid fissures are already abnormally wide may create a cosmetic problem. A lateral tarsorrhaphy at a later date may improve the appearance. Occasionally, the markedly depressed position of the globe, swelling of the muscle, and tightness of the tissues will make it technically impossible to insert a muscle hook under the insertion. In such instances, we have performed a complete tenotomy of the muscle at the insertion but obtained undesirable overcorrection in 2 of 19 patients so treated. Several muscle hooks with a groove to guide the tip of a knife during tenotomy have been designed but we have found that if a muscle hook can be inserted at all, a recession or retroplacement on a hang-back suture can also be performed and is preferable to a free tenotomy.

In patients with long-standing depression of the globe, the conjunctiva and Tenon’s capsule may shrink. To restore normal ocular motility, we perform a generous recession of the conjunctiva and Tenon’s capsule rather than force these back to the original incision during wound closure (see Chapter 26). For limitation of abduction, a similarly large recession of the medial rectus muscle is recommended, and other restrictions of ocular motility associated with an endocrine myopathy should be treated in an analogous manner. Because of the many variables involved, attempts to create dose-response curves for recessions in Graves’ ophthalmopathy have been unsuccessful.31 Some surgeons advocate that surgery on these patients be performed under topical anesthesia.31 Since it cannot be anticipated how much discomfort will be caused by excessive pull on the muscle hook, we prefer general anesthesia. It is of interest, however, that the oculocardiac reflex in patients with Graves’ ophthalmopathy has been reported to be markedly reduced or absent.31, 256

Injection of the inferior rectus muscle with botulinum toxin (Botox) is discussed in Chapter 25.

Most patients with Graves’ ophthalmopathy are severely incapacitated for many months by vertical diplopia. Restoration of single binocular vision, at least in primary position and in downward gaze, often can be accomplished by surgery. The functional results usually are excellent and a source of gratification for both patient and surgeon (see Fig. 21–16B). It must be mentioned, however,
Acute Orbital Myositis

Acute orbital myositis belongs to a subgroup of nonspecific orbital pseudotumors, and its etiology remains obscure, although immunologic mechanisms have been postulated.52, 253, 343 Symptoms include acute onset of unilateral orbital pain which may increase with eye movements, ptosis, conjunctival injection over the muscle insertion, proptosis, and diplopia.395 Unlike Graves’ ophthalmopathy, which shows a predilection for certain tissues and involvement of the posterior part of the muscle belly, inflammatory pseudotumor may involve any or all orbital tissues and the entire length of the muscle. Limitation of ocular motility is a common finding and may consist of restriction of ductions in the field of action of the involved muscle, distinguishing it from Graves’ ophthalmopathy in which motility is restricted in the field opposite that of the involved muscle. Weinstein and coworkers395 reported 21 patients with this condition, four with histories of ocular or systemic autoimmune disease. Treatment consists of systemic corticosteroids, supplemented if necessary by radiation therapy. Surgery may occasionally be required in patients with persistent diplopia who may benefit also from a diagnostic botulinum toxin injection to predict the outcome of an operation.24 Stidham and coworkers562 described isolated myositis of the superior oblique muscle. Recurrences have been reported in 50% of the cases248 and have been associated with lack of responses to systemic corticosteroids or nonsteroidal anti-inflammatory agents.227

With Case 21–3 we describe a typical patient.

CASE 21–3

An 8-year-old boy presented to our emergency room with bilateral lid swelling and proptosis of OS of 8 days’ duration. The patient had been using sulfacetamide sodium (Bleph-10) and naphazoline hydrochloride and antazoline phosphate (Vasocon-A) for the previous week without improvement. The patient complained about double vision and pain in OS. The mother stated that the proptosis had increased over the last 2 days with frequent episodes of nausea and vomiting. The history for trauma was negative and the child had been lethargic and had a low-grade fever for the past week. On examination the uncorrected visual acuity was 6/6 in OU. Exophthalmometry (Hertel) showed 18 mm OD and 21 mm OS. There was decreased retropulsion and pain on retropulsion OS. An exotropia of 30° and a left hypertropia of 10° was present at near and distance fixation. There was generalized conjunctival hyperemia, especially over the left medial rectus muscle. OS would not adduct (Fig. 21–18). CT showed swelling of the horizontal rectus muscles OU with maximal swelling of the left medial rectus muscle (Fig. 21–19A). The vertical recti were also involved (Fig. 21–19B). The diagnosis of acute myositis was made and the patient was placed on intravenous dexamethasone (Decadron, 15 mg every 6 hours). Improvement was rapid and dramatic; the exophthalmos, the adduction deficit in the left eye, and the pain subsided within several days.

Cyclic Heterotropia

Clinical Findings and Etiology

A rare but most intriguing form of strabismus apparently depends on a regular clock mechanism that usually follows a 48-hour rhythm; that is, a 24-hour period of normal binocular vision is followed by 24 hours of manifest heterotropia. The first two cases of alternate-day esotropia were described by Bühm in 1845.32 In 1905 Worth404 mentioned records in his possession of periodic strabismus that occurred every second day. There was no further mention of this form of strabismus in the literature until the roundtable discussion at the Second Strabismus Symposium of the New Orleans Academy of Ophthalmology in 1958 during which Burian59 related a similar case. Since that time, many cases have been reported and are referred to as circadian, periodic, alternate-day, or clock-mechanism esotropia.5, 49, 54, 63, 75, 121, 156, 254, 307, 314, 329, 399, 400 Costenbader and Mousel75 observed only three cases of cyclic heterotropia in 3500 strabismic patients.

In most patients with cyclic heterotropia, certain characteristics are evident. The onset may be in early infancy, but the condition usually becomes apparent during early childhood. On “straight” days no anomalies of binocular vision are observed; heterophorias and refractive errors, if at all present, are slight. However, cyclic esotropia has also been described in a monocular patient.
whose seeing eye adducted every second day.\textsuperscript{308} Thus, the presence of normal binocular function is not a prerequisite of the development of cyclic strabismus. On strabismic days, a large angle esotropia, often as large as $40^\circ$ to $50^\circ$, will appear. These measurements are consistent on subsequent examinations. On days when strabismus is present, sensory anomalies often are found, diplopia is infrequent, and fusional amplitudes determined with the amblyoscope are defective or absent.

The cyclic nature of the strabismus may last from 4 months to several years, after which the cycle breaks and esotropia becomes constant. A change in the cycle has also been reported after traveling rapidly through different time zones.\textsuperscript{239} The 48-hour cycle is encountered most commonly, but 72-hour\textsuperscript{49} and 96-hour cycles\textsuperscript{75} have been reported. Consecutive cyclic esotropia has been reported following surgery for intermittent exotropia, and the only three patients with cyclic esotropia that we have seen and treated each had a history of surgery for intermittent exotropia (see also Muchnik and coworkers\textsuperscript{254} and Uemura and coworkers\textsuperscript{379}).

In most reported instances cyclic esotropia occurred in childhood, although a sudden onset of this condition has also been reported in adults.\textsuperscript{71, 356, 376} Curiously, in one case cyclic esotropia developed after unioocular traumatic aphakia and was corrected by a secondary intraocular lens implantation.\textsuperscript{71}

While cyclic strabismus mostly occurs as esotropia, cyclic exotropia has also been reported.\textsuperscript{1} The most unusual case of cyclic strabismus was reported by Windsor and Berg,\textsuperscript{399} who observed a cyclic left superior oblique paresis in a 10-year-old boy following an injury to the trochlear region of the left eye. Left hypertropia with vertical diplopia accompanied by a positive Bielschowsky’s head tilt test for a left superior oblique paresis was present only on every second day. On alternate days, binocular vision was normal and the head tilt test was negative. After phenobarbital medication was given, the left hypertropia became constant, but when the drug was discontinued, the cycle returned. When the patient was kept awake all night, a spontaneous switch in the cycle occurred from 5 to 6 AM.

The mechanism of this extraordinary form of strabismus is obscure. Pillai and Dhand\textsuperscript{292} reported cyclic esotropia in association with central nervous system lesions. In one case the condition occurred after removal of a third ventricle astrocytoma, and in the other it developed with the advent of an epileptiform disorder. Cyclic esotropia developed in one instance 1 year after recovery from a traumatic abducens palsy after closed head surgery.
In some instances, features possibly related to a clock mechanism elsewhere in the body were noted. Böhm, in the first reported case, mentioned that in his patient the strabismus eventually disappeared but recurred whenever the child became upset. Roper Hall and Yapp observed behavioral changes on the squinting day, and in two of their patients abnormal electroencephalograms showed a change alternating from day to day. One patient experienced frequency of micturition only on the squinting day. Friendly and coworkers monitored psychological and physical functions in a patient with alternate-day esotropia and were unable to detect any concurrent cyclic changes. Richter, a world authority on biological clock mechanisms, pointed out that 24- to 96-hour cycles, as found with this entity, are not unique. Other periodic biological phenomena include sweating, salivation, body temperature, and pulse rate; a cyclic pattern occurs with almost every form of abnormal behavior in psychiatric patients. Richter’s research with rats and monkeys indicates the presence of a biological clock mechanism that keeps time with extraordinary accuracy and is entirely independent of all internal and external disturbances.

Cyclic strabismus, even though intermittent, differs profoundly from other forms of intermittent strabismus in that a significant latent deviation is absent on the nonsquinting day. Moreover, the abnormality is not associated with factors such as fatigue, accommodation, or disruption of fusion that convert an ordinary intermittent strabismus from its latent to its manifest form. The mystery of cyclic heterotropia is compounded by the fact that some reported patients have strong family histories of manifest strabismus or other anomalies of binocular vision.

Another rare cyclic form of strabismus, observed in association with periodic alternating nystagmus or periodic alternating gaze deviation, is periodic alternating esotropia. Hamed and Sibiger described this condition as an esotropia maintained in one eye for 1 to 2 minutes while the fellow eye fixates in abduction and the face is turned toward the side of the esotropic eye. This is followed by a phase of normalcy in which the eyes are aligned, after which the previously fixating eye becomes esotropic, the fellow eye fixates in abduction, and the direction of the face turns reverses. The authors reported this phenomenon in a 9-month-old girl with developmental delay and cerebellar atrophy.

**Therapy**

In considering treatment of cyclic strabismus the ophthalmologist faces two basic questions: Are these patients basically strabismic and, by some unknown mechanism, capable of maintaining normal binocular vision on alternate days? Or do these patients usually have normal binocular vision but develop strabismus on alternate days because of external stress or some unknown psychomotor disorder? The natural history of the disease (the clock mechanism eventually “breaks” and strabismus becomes constant on an everyday basis) and the results of surgical treatment seem to favor the first hypothesis. Surgery based on the full amount of heterotropia as it occurs on the day of squinting has been eminently successful in permanently curing this condition and in reestablishing normal binocular functions. If one were to accept the second hypothesis, one would expect a significant surgical overcorrection to appear on the “straight” days, but this has not been the case in patients who have undergone surgery.

**Acquired Motor Fusion Deficiency**

Acquired motor fusion deficiency is an infrequent disturbance of both fusional convergence and divergence that occurs after closed head trauma, after a cerebrovascular accident, as a result of intracranial tumors, and after brain surgery. The first description was by Jaensch in 1935 and damage to the midbrain has been postulated as a cause.

Acquired motor fusion deficiency may be associated with a decreased range of accommodation. Such patients complain of severe asthenopia, intractable diplopia, and inability to maintain single vision for any length of time. Post-traumatic fusion deficiency may follow surgical correction of paralytic strabismus of traumatic origin. In spite of perfect ocular alignment, diplopia persists and may be crossed at one moment and uncrossed or vertical at another. Examination will reveal marked decreased, or complete absence of, fusional amplitudes, that is, motor fusion. In contrast, sensory fusion and stereopsis are intact during the brief moments that such patients are able to superimpose the double images. It is reasonable to assume that a lesion in the midbrain accounts for this problem.
Acquired motor fusion deficiency after trauma must be distinguished from loss of motor fusion in adult patients caused by a unilateral cataract or uncorrected unilateral aphakia of long standing. In such patients motor fusion has become weakened from lack of binocular sensory input and usually returns once the obstacle to binocular vision has been removed. Motor fusion deficiency must also be differentiated from combined convergence and accommodation deficiency (see p. 503) since the impairment is not limited to convergence but affects divergence and sursumvergence as well.

A defect of motor fusion may be mistaken for malingering in a patient who complains of transient diplopia without an identifiable defect of ocular motility and seeks compensation for a work-related accident. Determination of fusional amplitudes with a rotary prism will quickly distinguish between a malingerer in whom motor fusion can be readily elicited and a patient with an organic motor fusion defect with decreased or absent fusional amplitudes. Ophthalmologists must become more aware of this condition, which is frequently overlooked, and recognize it as a legitimate post-traumatic disability.

There is no effective therapy. Prisms are useless because of the constant need for readjustments of the power and the direction of the prism base. Spontaneous improvement has been reported but is infrequent in our experience. An occluding scleral contact lens may be all one can offer to such patients.

Fracture of the Orbital Floor

Clinical Findings and Etiology

An exhaustive coverage of the subject of orbital fractures would exceed the scope of this book. However, because diplopia is often a prominent manifestation of this injury and its management clearly belongs in the realm of those concerned with ocular motility disorders, a brief discussion of this topic is justified.

The mechanism of orbital floor fractures and their clinical symptomatology have been described sporadically since 1889 but not until 1957, when Smith and Regan published their now classic paper, did the concept of an orbital “blow-out” become recognized. These authors demonstrated in cadavers that posterior impaction of the globe may cause a blow-out of the thin orbital floor as a result of sudden pressure on the bony area of least resistance. Orbital contents such as fat, fascia, the inferior rectus and oblique muscles, or, in some instances, the entire globe may prolapse into the maxillary antrum or part of these tissues may become incarcerated in a linear crack in the orbital floor.

The concept of a blow-out mechanism has been questioned since orbital floor fractures have been observed after the eye had been enucleated. A second mechanism for orbital floor fractures, which is independent of increased intraorbital pressure, has been proposed and is caused by the buckling effect of a severe blow to the inferior orbital rim (blow-in fracture). A recent study based on interference holography has identified several points along the orbital rim that on contact cause deformation of the bony orbit. Regardless of where the stress was applied the maximal deformation occurred on the medial aspect of the orbital floor, which also happens to be the area where most clinically diagnosed fractures occur.

The principal clinical manifestations after a recent fracture of the orbital floor usually are marked swelling and ecchymosis of the lids and periorbital soft tissue. Epistaxis may take place on the affected side. Proptosis commonly occurs during the immediate post-traumatic phase, even though in some patients a large defect of the orbital floor may cause enophthalmos. The presence of subcutaneous emphysema with crepitus is an indication of injury to the medial orbital wall. An associated malar fracture will produce anesthesia of the skin region supplied by the infraorbital nerve. Marked limitation of eye movements, particularly elevation, depression, or both, are common findings (Fig. 21–20); once the swelling of the lid has subsided and the eye can be opened, the patient will complain of diplopia. Limitation of elevation is more likely to be caused by an anterior and of depression by a posterior fracture site (Fig. 21–21).

The forced duction test will demonstrate limitation of passive elevation when the structures surrounding the inferior muscles or the muscles themselves are incarcerated. The clinician must be aware, however, that limitations of passive ductions also may occur from intraorbital hemorrhages or edema, especially in the immediate post-traumatic phase (see below).

Of the various methods of radiologic examination, CT scans with coronal views have emerged as a most accurate technique for demonstrating...
defects in the bony structure of the orbital floor (Fig. 21–22). If the patient cannot be positioned for CT scans, conventional tomography is advocated. When evaluating cloudiness of the antrum, it is important to differentiate between herniated orbital tissue and hemorrhage associated with the injured maxillary periosteum or mucosa with radiographic clouding (pseudoprolapse) of the antrum in the presence of an intact orbital floor, as shown in Figure 21–23.

Serious associated ocular injuries, led in frequency of occurrence by retinal edema and hyphema, were present in 50 of 159 patients evaluated at the Wilmer Institute. Careful ophthalmologic examination is absolutely essential in all patients with orbital injury, and they should never be treated, as sometimes happens, by otolaryngologists or plastic surgeons without ophthalmologic evaluation.

**Therapy**

In the past, most authors stressed the need for early surgical repair of the orbital floor to prevent
late diplopia and enophthalmos and to avoid technical difficulties from scar formation and fibrosis if surgery is delayed. The questions arose as to whether all patients with radiographic evidence of an orbital floor fracture require surgical repair and how soon after the injury such repair should be attempted. Studies of the natural history of orbital floor fractures not surgically treated have shown clearly that not all patients require surgery. Patients with orbital floor fractures who initially have no diplopia or in whom diplopia disappears within 14 days after injury should not undergo surgery.

The fact that diplopia after orbital injury is not always caused by incarceration of orbital tissues in the fracture site must be taken into consideration. Limitation of eye movement can be caused also by contusion of one or more extraocular muscles or their nerves. Of 40 blow-out fracture patients studied by Wojno, 7 had motility defects consistent with paralysis of one of the extraocular muscles or cranial nerves. Restriction of ocular motility may also occur because of edema or hemorrhage within the orbit. In such cases, alleviation of the limitation of ocular motility and decrease of double vision can be expected soon after injury. Such changes may be subtle at first; therefore we recommend careful charting of diplopia fields in following these patients.

These factors also must be considered when evaluating the results of the forced duction test. A positive test result in the early post-traumatic phase may be unrelated to actual incarceration of tissue. Only several days after the injury, when the initial effects of trauma have subsided, will the forced duction test become more reliable in the diagnosis of a blow-out fracture. When the patient is seen first in the emergency room and the diagnosis of an orbital floor fracture has been confirmed by a CT scan, one of us (E.C.) places a traction suture through the inferior rectus muscle insertion to fixate the eye in a position of elevation for about a week. He has found that surgery may be avoided with this method in many cases.

Surgical repair has been advocated even in the absence of diplopia to prevent development of enophthalmos. Enophthalmos occurs rarely in the immediate post-traumatic phase, and if it does occur, usually signifies a massive defect in the orbital floor that must be repaired without much delay. Measurement of the orbital volume by CT within 20 days after injury may identify those patients at risk for late enophthalmos.

In most instances enophthalmos does not develop until months after the injury. Its etiology may be unrelated to actual loss of orbital tissue into the maxillary antrum but rather caused by a contusion injury with subsequent shrinkage of the orbital fat tissue, possibly from interference with its blood supply. Clearly, surgical repair of the fracture would not prevent this complication, and Emery and coworkers have shown that the occurrence of enophthalmos is unrelated to whether the fracture was repaired or not. Once it has been established that there is no improvement in a clinically significant defect of ocular motility, the radiographic findings are positive for a fracture, and the results of the forced duction test indicate incarceration of orbital tissue, surgery should be delayed no longer. Exploration of the orbital floor is performed under general anesthesia. The surgical technique that we use has been described and illustrated by Goldberg, and only a brief summary of the procedure follows.

The infraorbital rim is exposed through a curvilinear incision through the fold of the lower lid. The periosteum is incised slightly below the orbital rim and elevated posteriorly to expose the fracture site. Special care must be exercised not to confuse the infraorbital groove with a fracture line or to mistake the infraorbital nerve for incarcerated tissue. Once the limits of the fracture have been identified, the herniated tissue is gently ex-
tracted from the defect in the bony orbital floor. Occasionally, difficulties are encountered in freeing all the incarcerated tissues when using the orbital approach. Should this occur, combined manipulation of the tissue from above and below, through a transoral Caldwell-Luc approach, is recommended. It is advisable therefore to have an otolaryngologist standing by to join the operating team and perform the Caldwell-Luc procedure if this complication occurs. The fractured floor components are elevated and replaced in their normal position. When a large bony defect is encountered, a piece of 0.3 mm Supramid sheath should be sutured to the bone or periosteum to seal the orbital floor and prevent migration or extrusion. The periosteum is closed with 3-0 chromic gut and the skin wound with interrupted 6-0 silk sutures.

The most serious complication after repair of orbital floor fractures is loss of light perception as a result of postoperative orbital hemorrhage, occlusion of the central retinal artery, or damage to the optic nerve during surgery. Less serious postoperative complications include extrusion of the implant, ectropion of the lower lid, and persistent diplopia. The diagnosis and management of these and other complications have been discussed elsewhere.259

As mentioned above, some restriction of ocular motility as a result of tissue swelling is commonly encountered during the immediate postoperative period. If diplopia in primary position or in downward gaze persists for as long as several months after surgery, the defect must be reevaluated for further surgical treatment.

Ocular motility after repair of an orbital floor fracture may be limited because of incomplete liberation of incarcerated tissue (i.e., the fracture may have extended farther posteriorly than was apparent at the time of surgery) or by post-traumatic fibrosis, loss of elasticity, or paralysis of the inferior extraocular muscles. Diplopia in upward gaze usually can be ignored but should be corrected if it occurs in primary position or depression and the patient has a significant chin elevation. If the forced duction test reveals mechanical limitation of elevation, we first recess the ipsilateral inferior rectus and use adjustable sutures in adults. The contralateral superior rectus may be recessed at the same or at a subsequent surgical session to counteract the secondary hypertropia. In the case of a traumatic inferior rectus palsy, depression of the eye will be limited, and resection of the paralyzed muscle combined with recession of its antagonist is usually effective in eliminating double vision in downward gaze.263, 381

Fracture of the Medial Orbital Wall

A less well-known sequela of orbital trauma is a fracture of the medial orbital wall as an isolated lesion or accompanying a fracture of the floor.94, 98, 397 Adduction will be limited and this limitation can reach severe degrees as shown by the tragic Case 21–4.

CASE 21–4

A 26-year-old woman without previous ocular complaints underwent a right ethmoidectomy through a transnasal approach. Upon awakening she noted that she could not see with her OD. Upon examination 10 days after surgery, visual acuity OD was found to be reduced to finger counting at 1 m, her acuity OS was 6/6. OS was in a position of abduction and could not be moved into primary or any other gaze position (Fig. 21–24). The pupil of OD was dilated and fixed. The forced duction test showed that the eye could not be moved at all by passive force. CT showed a large defect of the right medial orbital wall with incarceration of the optic nerve and medial rectus muscle in the ethmoidal sinus (Fig. 21–25). Apparently, the orbit had been entered during the ethmoidectomy and these orbital tissues had been pulled into the ethmoidal sinus. A surgical attempt to free this restriction was unsuccessful. Within a matter of several days of observation visual acuity OD decreased to no light perception. A second attempt by another surgeon to mobilize the eye

FIGURE 21–24. Case 21–4. Note immobility of the right eye, which is fixed in a position of abduction as the left eye adducts (A), fixates in primary position (B), and abducts (C).
Several similar cases have been reported.  

Superior Oblique Myokymia

Clinical Findings and Etiology

Episodic nystagmoid intorsion and depression of one eye, accompanied by visual shimmer and oscillopsia, although mentioned by Alexander Duane in 1906 and, more recently, by Clark, was not sufficiently recognized as a distinct clinical entity until Hoyt and Keane’s classic description of benign superior oblique myokymia. The onset of this condition is, as a rule, in adulthood, and the symptoms are most annoying to the patient. The diagnosis is often missed by the primary physician, and we have seen several cases diagnosed elsewhere as having a functional disorder. However, careful examination, with the slit lamp if necessary, will reveal high-frequency and low-amplitude torsional and vertical oscillations of the affected eye during an attack. Each episode may last from 20 seconds to up to several minutes, with recurrences at irregular intervals, usually several times each day. In some patients the attacks are precipitated by downward gaze or by physical activity. One of our patients reported episodes of superior oblique myokymia during sexual intercourse and while lifting weights. Duration and frequency of these episodes tend to increase with time and long-term observation has established the chronicity of the condition.

Superior oblique myokymia is usually benign and in most instances not accompanied by other conditions. However, association with a posterior fossa tumor has been described and the myokymia was the only neurologic sign. In another instance myokymia developed in a patient with a dural arteriovenous fistula. Myokymia has also been reported to occur months and years after acquired superior oblique paralysis, perhaps as a manifestation of a postdenervation phenomenon. This possibility is further supported by the results of MRI in two patients which showed the cross-sectional area of the superior oblique muscle to be smaller than normal. On the basis of electromyographic recordings from the ipsilateral superior oblique muscle, a nuclear disorder or a supranuclear response to peripheral injury of the trochlear nerve has been implicated. An analysis of eye movements with the search coil technique taken with electromyographic evidence from other studies led Leigh and coworkers to propose that myokymia reflects spontaneous discharge of trochlear neurons that have undergone regenerative changes. The observation of myokymia developing after superior oblique paralysis and the MRI findings seem to support this theory.

Therapy

Medical treatment with carbamazepine (Tegretol), phenytoin, clonazepam, baclofen, and propranolol hydrochloride or the topical use of a beta blocker has been reported to be successful. We have been unimpressed with medical treatment in controlling the symptoms on a long-term basis in the five patients we have treated. Similar conclusions were reached in a survey of 16 patients followed at the Wilmer Institute. Prolonged use of carbamazepine is not without risks and requires regular monitoring of the blood count. Drowsiness, dizziness, a potential teratogenic effect during pregnancy, and incompatibility with alcohol and barbiturates are but some of the reasons why in our experience most patients, given the alternative, eventually opt for surgery.

Surgical treatment consists of a tenotomy or tenectomy of the superior oblique tendon, usually combined with myectomy of the ipsilateral inferior oblique. One patient responded...
well to a nasal recession of the anterior portion of the superior oblique tendon. Microvascular decompression of the trochlear nerve at the root exit zone has also been reported to be successful but appears to us as an unnecessarily complex procedure, considering the ease with which surgery on both oblique muscles can be performed. Several authors have stressed that more than one operation may be necessary, and in one instance a superior oblique myectomy with trochlectomy had to be performed after failure of superior oblique tenectomy to adequately eliminate symptoms.

We are inclined to blame the postoperative recurrences of oscillopsia reported in the literature on an incomplete transection of the tendon and its sheath or on adhesions between the proximal stump of the tendon and the superonasal aspect of the globe. The first complication can be avoided by adequate surgical exposure and meticulous operating technique (see Chapter 26), the second by removing a large segment of the tendon and sheath.

Ocular Myasthenia Gravis

Clinical Findings

Myasthenia gravis is of interest primarily to the neuro-ophthalmologist. Several reviews are available in the recent literature. Isolated case descriptions can be found in the literature as early as 1672 but the first detailed report of three patients was that by Erb in 1879 whose name has remained associated with the disease. A brief discussion of ocular myasthenia in this chapter is in order because of the ocular motility disturbances frequently associated with this disease.

Myasthenia is an acquired autoimmune disease, affecting synaptic transmission across the neuromuscular junction in which the number of available acetylcholine receptors is decreased. A close relationship exists between myasthenia gravis and hyperplasia of the thymus, and thymectomy is followed occasionally by a dramatic remission of symptoms. A pure ocular form of the disease exists, but more frequently other muscles, especially those involved in mastication and breathing, become affected. The specific characteristics of extraocular muscles, rapid contraction rate, and high firing frequency of the fast twitch fibers (see Chapter 6) make the extraocular muscles especially vulnerable to the disease. Diplopia and ptosis are the first symptoms in half the cases. The first symptoms may be preceded by an emotional upset, upper respiratory infections, or pregnancy. These symptoms may be absent or less pronounced on awakening, then progress dramatically with increased muscular activity. Ptosis is often asymmetrical, may be completely unilateral, and infrequently involves both eyes to the same degree. In most instances the motility disturbances are fleeting, may affect any or all of the extraocular muscles, range from slight paresis to complete gaze paralysis, and may simulate paralyses of cranial nerves III, IV, and VI. Paresis of upward gaze is an early manifestation of the disease. Cogan’s lid twitch sign is a helpful diagnostic sign in the early stages of the disease: after rapidly and repeatedly looking from downward gaze into the primary position, the upper lid(s) will elevate excessively for a brief moment and may twitch several times. Weakness of adduction of either eye and nystagmus of the abducting eye may mimic an internuclear ophthalmoplegia and one of us (G.K.v.N.) had the privilege to examine, together with the late Dr. Frank Walsh, one of the first recognized cases of pseudo-internuclear ophthalmoplegia in association with myasthenia that was later published by Glaser.

The disease is more prevalent in women than in men (3:1) although the ocular form affects men more frequently, especially after the age of 40 years. As a rule the first symptoms occur between the second and fourth decades of life. However, Walsh and Hoyt have commented that the onset of myasthenia gravis may occur as early as the first year or as late as the seventh decade of life. We have observed children between the ages of 2 and 4 years who developed the disease and occurrence in children younger than that has been described.

Diagnosis

The diagnosis of myasthenia gravis is based on demonstration of easy muscular fatigability and its rapid relief by systemic administration of an anticholinesterase agent such as edrophonium chloride (Tensilon). The improvement of ptosis is often more dramatic than the improvement of ocular motility, which may be subtle and of very short duration. We prefer to administer the Tensilon test with the patient seated before a deviometer while
the strabismus angle is being measured by an orthoptist. Coll and Demer\textsuperscript{72} advocated performing the test while the angle of strabismus is being monitored on a Hess screen, which seems to be an even more accurate method to evaluate ocular motility during the injection.

We use a 10-mg/mL solution of Tensilon intravenously. A second syringe containing 0.5 to 1.0 mg of atropine to counteract side effects should be readily available. Initially, a 1- to 2-mg test dose is given and the patient is observed for the development of hypotension, bradycardia, or arrhythmia. Fortunately, such side effects occur infrequently. The injection is then continued in additional increments of 1 to 2 mg every 60 seconds until a positive response is obtained or the syringe is empty.

If the test result is ambiguous a determination of circulating antiacetylcholine receptor antibodies may be helpful in establishing the diagnosis. These antibodies have been found in up to 87% of patients with the disease.\textsuperscript{394} Electromyography is useful as an auxiliary diagnostic test since it shows a characteristic myopathic pattern (Fig. 21–26). This test is helpful in differentiating the ocular involvement of myasthenia gravis from a peripheral neurogenic paresis.\textsuperscript{38, 170, 277, 369}

**Therapy**

Treatment is directed toward providing the patient with symptomatic relief from double vision or obstruction of vision by a drooping upper lid and should be initiated and supervised by a neurologist. In our experience, cholinesterase inhibitors (pyridostigmine bromide, Mestinon) are rarely successful in completely controlling ptosis and diplopia in the ocular form of the disease. Oral corticosteroids are often more effective in alleviating ocular myasthenia. If medical treatment is unsuccessful, the clinician may have to resort to occluding one eye to eliminate double vision or to keeping the lid(s) elevated by a ptosis crutch.

As a rule, surgery is not indicated for this condition. Exceptions occur, however, in the instance of a myasthenic patient with long-term and stable paresis or paralysis of a particular muscle or muscle group or stable comitant strabismus who does not respond to medical therapy. We have been able to eliminate diplopia by muscle surgery in several such patients after ascertaining the stability of the condition during prolonged periods of observation. Others have also obtained satisfactory results with surgery in selected cases.\textsuperscript{2, 85, 266}

**Chronic Progressive External Ophthalmoplegia (Ocular Myopathy of von Graefe)**

**Clinical Findings and Etiology**

Chronic progressive external ophthalmoplegia (CPEO) is a rare and, as its name implies, progressive disorder that affects ocular motility and the function of the levator palpebrae muscle. This condition also belongs in the realm of neuroophthalmology, and the reader is referred to the standard texts for detailed information. CPEO was first described by von Graefe\textsuperscript{138} and is characterized by bilateral ptosis and decreasing motility of the eyes in all directions of gaze. Its etiology is still disputed, and there has been extensive discussion as to whether this disease is caused by a central lesion involving the ocular motor nuclei\textsuperscript{43, 84} or by a primary myopathy of the extraocular muscles similar to muscular dystrophy.\textsuperscript{397} Histologic evidence has been presented to support both hypotheses. Electromyographic data, on the other hand, clearly indicate a myopathy of the extraocular muscles.\textsuperscript{40, 369} The frequent association of chronic progressive external ophthalmoplegia

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**FIGURE 21–26.** Electromyogram from extraocular muscle in myasthenia gravis. Decreased frequency and amplitude of action potentials after prolonged voluntary innervation (upper tracing); activation of numerous motor units and increase of discharge frequency after intravenous injection of 10 mg edrophonium chloride (Tensilon) (lower tracing). (From Huber A: Die peripheren Augenmuskellähmungen. Ber Dtsch Ophthalmol Ges 67:26, 1966.)
Clinical Characteristics of Neuromuscular Anomalies of the Eye

With pigmentary and other forms of retinal degeneration has led Thorson and Bell to postulate that this disease may be considered abiotrophic. Recent advances in molecular genetics (for a review of the literature, see Wallace et al.) have identified CPEO as a mitochondrial cytopathy, especially in highly oxidative tissue such as muscle, brain, and heart.

The onset is usually before 30 years of age, and in some cases the disease occurs early in childhood. Both sexes are equally involved, and familial occurrence is frequent. Ptosis, which as a rule is bilateral, is often the first symptom, followed by slowly progressive limitation of ocular motility with a predominant involvement of the elevating muscles (Fig. 21–27). In the extreme case, both eyes may become “frozen.” In the advanced stage, complete ptosis may force the patient’s chin to be maximally elevated to allow vision. In patients with ocular myopathy, unlike those with myasthenia gravis, there are no remissions and anticholinesterase agents have no effect on muscular function. Complaints about diplopia are, curiously, rare, even though a severe disturbance of ocular motility may be present. In addition to involvement of the extraocular and levator muscles, the orbicularis and other facial muscles may become affected, especially those used in mastication. Atrophy of the extraocular muscles, especially a decrease of muscle thickness, can be demonstrated on CT scans.

Therapy

In advanced disease a ptosis crutch may help the patient; however, a lid suspension procedure often becomes necessary. Special care must be taken to protect the cornea during sleep by taping the lids at night. Prisms may be helpful in eliminating diplopia in some patients, and surgical alignment of the eyes has been reported to create satisfactory results. We have no personal experience with surgery on the extraocular muscles for this condition.

REFERENCES

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408. Wright KW, Min BM, Park C: Comparison of superior...