CHAPTER 18

Cyclovertical Deviations

The diagnosis and management of cyclovertical deviations are special challenges to the ophthalmologist. There are several disorders that on first glance appear similar clinically but differ widely in etiology and management. As in no other aspect of strabismology, correct diagnosis is of utmost importance, since an operation performed on the basis of an erroneous interpretation of the underlying problem may cause disastrous and permanent consequences with respect to the patient’s binocular function. Once the correct diagnosis has been made, medical and surgical management of such deviations does not present any special problems, and the therapeutic results can be the most gratifying in the field of strabismus.

Cyclovertical deviations differ from horizontal deviations in several aspects. Sensorial adaptations in the form of amblyopia or anomalous retinal correspondence are noted far less frequently with this type of deviation than with horizontal strabismus. Comitance is rare, and the deviation is generally smaller in magnitude, yet the size of a cyclovertical deviation is not an indication of the extent of the problem caused for the patient. Although some patients with well-developed binocular functions often are able, by motor fusion, to overcome surprisingly large vertical deviations, the low fusional reserve in the vertical directions in most others precludes this compensatory mechanism. Consequently, a hyperdeviation of only $1^\circ$ or $2^\circ$ can cause diplopia or blurring of vision, especially during reading. Such small residual hyperdeviations following surgical alignment of horizontal strabismus are of special clinical significance, for they may present insurmountable obstacles to a functional cure.

The prevalence of cyclovertical deviations in association with horizontal deviations or as isolated anomalies is high. White and Brown observed that in patients with motility disorders, approximately half had isolated vertical anomalies and another third had combined horizontal and vertical muscle problems. Scobee found a vertical component in 43% of 457 patients with esotropia.

Bielschowsky classified cyclovertical deviations into five groups: (1) purely comitant vertical deviations, (2) vertical deviations of paretic origin, (3) deviations with unilateral overaction of the inferior oblique muscles, (4) dissociated vertical deviations, and (5) vertical deviations combined with features of several of the other groups. This classification is still of some usefulness today even though we have learned since Bielschowsky that elevation in adduction is not exclusively caused by an overacting inferior oblique muscle.

In this chapter nonparalytic and nonmechanical cyclodeviations are described. Paralytic deviations are discussed in Chapter 20, and hyperdeviations caused by mechanical factors (endocrine myopathy, congenital fibrosis, and orbital floor fractures) are described in Chapter 21.

Comitant Hyperdeviations

Etiology and Clinical Characteristics

Truly comitant hyperdeviations occur infrequently. To find a patient with a significant vertical devia-
tion of the same magnitude in all positions of gaze with either eye fixating and with the head tilted to either shoulder is indeed unusual. Anderson, in a survey of 600 patients with cyclovertical anomalies, was unable to find a single truly comitant deviation. Repeated measurements in the diagnostic positions of gaze in the majority of patients may reveal a paretic component or an apparently primary overaction of one or several cyclovertical muscles. The etiology of truly comitant deviations of a magnitude rarely exceeding a few prism diopters is not clear. At one point, some of the patients may have had a paretic deviation that became comitant with the passage of time (see Chapter 20). In others, an anomalous position of rest caused by anatomical or mechanical factors or abnormal innervation may be a causative mechanism.

### Therapy

The very nature of comitant cyclovertical deviations means that prisms are ideally suited for relief of the patient. They should be distributed evenly before the two eyes (base-down before the hypertropic eye), and the prescription should be based on the minimal prismatic power that provides comfortable single binocular vision. When performing surgery to correct a coexisting horizontal deviation, comitant hyperdeviations can be eliminated by lowering the horizontal muscle insertions of the hypertropic eye or raising the insertions of the hypotropic eye (see Chapter 26).

### Terminology

The lack of precise etiologic information about DVD is reflected by the plethora of terms in use at one time or another: anatopia, alternating hyperphoria or hypertropia, double hypertropia, occlusion hypertropia, alternating sursumduction, dissociated double hypertropia, dissociated alternating hyperphoria, and dissociated vertical divergence. To speak in this context of alternating, dissociated, double, or occlusion hyperphoria or hypertropia, as many authors (including Bielschowsky) have, is incorrect, because DVD is different from ordinary hyperdeviations. For instance, in a patient with right hypertropia, either the right eye is elevated when the left eye is fixating or the left eye is depressed when the right eye is fixating. On the other hand, in DVD, either eye elevates when the fellow eye is fixating. Alternating sursumduction, a term introduced by Lancaster and Swan emphasizes the monocular nature of the movement (a duction and not a version or vergence), and its use has become rather widespread. Nevertheless, this description is not completely accurate because the movements are not limited exclusively to sursumduction but contain substantial elements of excycloduction and sometimes abduction. Moreover, the deviation does not always alternate but may be restricted to one eye. For these reasons, we prefer the generic term dissociated vertical deviation, which carries no implications with regard to the etiology of the condition and for which the abbreviation DVD has become widely accepted. Although no great friends of medical abbreviations, we will use DVD during the remainder of this discussion.

### Dissociated Vertical Deviations

Dissociated vertical deviation (DVD) is among the most intriguing and least understood of all forms of strabismus. Even though the unique clinical features of this anomaly clearly distinguish it from other forms of vertical motor disturbance, the diagnosis may be difficult when associated with other forms of strabismus, especially with cyclovertical deviations. Although Bielschowsky credited Schweigger (1894), Stevens (1895), and Duane (1896) for the first reports of this entity, it was he who provided the first comprehensive description and minute clinical analysis of DVD.

### Clinical Characteristics

DVD is characterized by the spontaneous drifting of either eye upward when the patient is fatigued or daydreaming or when fusion is artificially interrupted by covering one eye (Fig. 18–1). When the elevated eye is covered, it may perform pendular, vertical movements. When the cover is removed, the elevated eye will move slowly downward and settle in the primary position.

The amount of elevation when the eye is covered is variable, tending to increase after prolonged occlusion, and often differing between the two eyes. According to Bielschowsky several other features are present in most patients with
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DVD. These include excycloduction of the elevated eye and incycloduction of the fixating eye. As the elevated eye returns to the primary position it incycloducts. This torsional movement of the globe is often easily detected without magnification by the observer. Latent nystagmus, which often but not always has a cyclovertical component, may be associated with DVD.6

These additional symptoms justify consideration of DVDs as a syndrome. Observation of the iris pattern and the conjunctival vessels will nearly always reveal incycloduction as the elevated eye returns to the midline, indicating that it was excycloducted while in the dissociated position. The excycloduction of the elevating eye may be accompanied by a synchronous incycloduction of the fixating eye (cycloversion).44 Occasionally, excycloduction of each eye under cover or spontaneously and latent nystagmus may be the only manifestation of a dissociated deviation. In such cases we speak of a dissociated torsional deviation (DTD). In other cases, the full syndrome with its vertical component may involve one eye only while it manifests itself as an isolated excycloduction in the other.

Anderson7 and Lyle and Bridgeman69 drew attention to the association of a head tilt with DVD. The prevalence of anomalous head posture in this condition has been reported to range between 23%19 and 35%.5 Most authors reported the head to be tilted away from the eye with the larger vertical deviation19, 101 but the opposite has also been observed.5, 23 Passive tilting of the head toward the opposite of the side of the habitual posture increases the vertical deviation, which has led to the conclusion that the anomalous head posture decreases the magnitude and thus improves the motor control of the type of alternating hyperphoria investigated by these authors. De Decker and Dannheim de-Decker23 reported chin depression in patients with bilateral dissociated deviations. Surgical correction of DVD improves the head posture.23, 101

DVD occurs in patients with and without overaction of the inferior oblique muscles and may also be associated with overaction of the superior oblique muscles and an A-pattern exodeviation in downward gaze45, 71 (Fig. 18–2). The vertical angle of dissociated deviations is usually somewhat less in abduction than in adduction; however, it may also be larger in abduction.46 Latent nystagmus occurs in approximately half the patients with DVD and, in fact, is seldom encountered in the absence of this anomaly (see Anderson2, p.16).

If a photometric neutral filter wedge is placed before the fixating eye while the other eye is occluded and elevated, the eye behind the cover will make a gradual downward movement, and may even move below the primary position as the visual input to the fixating eye is progressively decreased by the filter wedge. When the wedge is moved from positions of greater to lesser filter

FIGURE 18–2. Dissociated vertical deviation with overaction of both superior oblique muscles. A, Left hypotropia with the right eye fixating and right hypotropia with the left eye fixating. B, Underaction of both inferior oblique and overaction of both superior oblique muscles.
density, the eye behind the cover will elevate. This intriguing observation was first reported by Bielschowsky and has become known as the Bielschowsky phenomenon. Bielschowsky explained the phenomenon that bears his name in the following manner: When the visual input to the fixating, say, the right eye is decreased by holding filters of increasing density before it, the effort to maintain fixation triggers an abnormal innervation to the elevators. The effort to maintain fixation with the right eye against this innervation elicits a compensatory innervation to the depressors. The left eye follows this innervation under cover and returns to the primary position or even below it.

A DVD may occasionally occur as an isolated phenomenon in patients in whom binocular functions are apparently normal, but is found often in association with infantile esotropia and less often with accommodative acquired esotropia, exotropia, and heterotropia of sensory origin. An association with Duane’s syndrome has also been described. The high prevalence of DVD in essential infantile esotropia has been discussed in Chapter 16 and it is of interest that a similarly high rate of occurrence has been reported in infantile exotropia. In spite of a careful search the condition is rarely diagnosed in infancy. In our experience the diagnosis is most commonly made between the ages of 2 and 5 years and often years after surgical alignment of the horizontal deviation. The age at surgical alignment could not be correlated with the manifestation of DVD.

DVD is usually bilateral and asymmetrical. Of the 170 cases observed by us in a group of 408 children with essential infantile esotropia, only 24 (14%) were unilateral and only 13 (9%) were symmetrical. However, unilateral occurrence is often observed in deeply amblyopic eyes and in sensory heterotropia. The commonly occurring asymmetry between the two eyes is reversed in the supine position with the head tilted back: the eye with a larger deviation in the upright position has a smaller one with the patient supine and the head tilted back. This observation suggests a possible effect of inputs from otolithic and possibly neck muscle sensors on the amplitude of a DVD.

An active suppression mechanism usually will eliminate diplopia in patients with a spontaneous DVD. Exceptions to this rule are rare but do occur as shown in Case 18–1.

CASE 18–1

This 26-year-old man has had crossed eyes since infancy. Surgery was performed on the eye muscles when he was 3 years of age. For the past 13 years he has experienced intermittent double vision. Examination showed corrected visual acuity of 6/4.5 OD and 6/6 OS. He wore prescription glasses to correct a mild compound myopic astigmatism. The patient had fairly pronounced latent nystagmus and an esotropia of 14° at near and distance fixation. In addition he had 18° right hypertropia with the OS fixating and 10° left hypertropia with the OD fixating. He exhibited characteristic incycloduction as each elevated eye took up fixation in primary position. A V pattern was absent, and there was no inhibitional palsy of the contralateral superior rectus when he fixated with the adducted elevated eye. As soon as the eyes were dissociated, the patient became aware of diplopia. The red-glass test established that the diplopia was in accord with the deviation; that is, the images were uncrossed and had a vertical component, the laterality of which depended on which eye was fixating. The diagnosis was residual esotropia with a DVD and absence of suppression. No treatment was advocated.

Campos and coworkers pointed out that suppression is not the only mechanism that accounts for absence of diplopia in this condition by showing that a binocular vertical perceptual adaptation may exist in these patients.

Diplopia can be elicited in most patients with a dark-red glass, and the amount of separation between the images is used to measure the amplitude of elevation of each eye (see Chapter 12). The fact that the patient will localize the red image below the fixation light, regardless of whether the red glass is held before the right or left eye, clearly differentiates a DVD from other forms of cyclovertical anomalies in which the red image is localized below or above the fixation light, depending on which eye fixates.

Measurement

An accurate quantitative assessment of DVD may be obtained provided visual acuity in each eye is sufficient to visualize the fixation target, using a modification of the prism and cover test. As the patient focuses on the fixation target at 6 m distance, the occluder is quickly shifted to the fixating eye, allowing the previously dissociated and elevated eye to take up fixation. The cover is then returned to the nonfixating eye. As the alternate cover test is continued, increasing amounts of
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base-down prisms are held under the occluder in front of the nonfixating eye until the downward fixation movement of that eye is neutralized. The procedure is then repeated with the fellow eye fixating.

Etiology

Of the numerous theories advanced to explain the mechanism of this intriguing anomaly in the past, only a few will be mentioned in this chapter. Elastic preponderance of the elevator or the depressor muscles\(^{102}\); paretic factors\(^{25}\) especially bilateral paresis of the depressor muscles\(^{104}\), p. 183; and imbalances between the amount of innervation originating from each vestibular organ\(^{87}\) have been cited as causes in the older literature. For other explanations, see White,\(^{127}\) Verhoeff,\(^{125}\) Posner,\(^{91}\) Crane,\(^{19}\) Helveston,\(^{46}\) and Houtman and coworkers.\(^{53}\) It has even been reported that DVD may be caused by an abnormal visual pathway routing similar to that described in albinism\(^{35}\) (see Chapter 9). However, as one may have expected, this finding could not be reproduced.\(^{3, 10, 62, 135}\)

The results of more recent investigations\(^{11, 15, 41, 93, 94, 133}\) are in basic agreement with what Bielschowsky\(^6\) so lucidly described in 1931 and in his later publications\(^7, 8\): DVD is caused by a vertical vergence signal that elevates the occluded eye and would depress the fixating eye if it were not for a simultaneous supraversion impulse that cancels the innervation to depress the fixating eye while at the same time, according to Hering’s law, increasing the innervation flowing to the elevators of the occluded eye. Bielschowsky arrived at this explanation by meticulous clinical observation, sound reasoning, and without the benefit of modern search coil recording techniques that have essentially confirmed the validity of this innervation pattern and sequence\(^{41, 93, 135}\).

The origin of the vertical vergence innervation is still a matter of dispute. Bielschowsky\(^6\) suspected an alternating and intermittent excitation of both subcortical centers that govern vertically divergent eye movements. He cited as examples for such movements and support for the existence of such centers skew deviation and seesaw nystagmus and felt that the unilaterality of the condition in some cases is “based on the coincidence of the voluntary fixation impulse with the involuntary action of one of the vertical divergence centers.”\(^8\), p. 35) The reason for this abnormal excitation of hypothetical vertical divergence centers remains unknown. There is no question, however, that the impulse for a DVD must originate in the fixating eye. Bielschowsky\(^8\), p. 36 emphasized the need to differentiate between a hyperdeviation based on anatomical conditions, that is, an anomalous position of rest, and the dissociated deviations of innervational origin. Spielmann\(^{116}\) convincingly confirmed this difference by showing that DVD does not occur when fixation is prevented by covering both eyes with translucent occluders (Fig. 18–3). Spielmann\(^{111-113}\) assumed that DVD is caused by an imbalance of binocular stimulation. Although this may explain the frequent occurrence of DVD in essential infantile esotropia and the occasional occurrence with sensory heterotropias, it does not account for DVD in patients with otherwise normal binocular functions.

Several additional explanations were proposed in recent years. From the direction of the cyclorotation of the elevating eye (extorsion) and the fixating eye (intorsion) several investigators\(^{31, 41, 93}\) have concluded that the vertical vergence movement must be predominantly mediated by the oblique muscles because the vertical rectus muscle would produce a cyclorotation in the opposite direction. Guyton\(^41\) and Cheeseman and Guyton\(^15\) believe that this oblique muscle–generated cycloversion is a purposeful eye movement that damps latent cyclovertical nystagmus to improve visual acuity. The accompanying elevation of the nonfixating eye, the DVD, is seen as an unavoidable

![Figure 18-3](image-url). Combined vertical and horizontal dissociated deviation in the right eye (A) and predominantly vertical dissociated deviation in the left eye (B). C. Absence of dissociated vertical deviation in the fixation-free position when both eyes are covered with translucent Spielmann occluders. For details, see text.
and undesirable byproduct of this nystagmus damping mechanism (see Chapter 23). There are a number of observations that are difficult to reconcile with this theory, which is based on the concept of the inferior oblique muscle being the primary elevator in vertical vergences. In DVD the dissociated eye elevates not only in adduction but also in primary position and abduction (Fig. 18–4). In fact, in some cases the elevation in abduction is greater than in adduction. Clearly, these are gaze positions in which the inferior oblique muscle has little or no elevating power and the superior rectus muscle must be the principal elevator. While it is indisputable that excycloduction of an elevating eye can only be caused by the inferior oblique muscle, it does not inescapably follow that this muscle is also the predominant elevator. One must also consider the possibility that both the superior rectus and inferior oblique muscles co-contract during elevation but that the stronger excyclotorsional effect of the inferior oblique overrides the weaker incyclotorsional effect of the superior rectus muscle. Moreover, in our experience and that of others a DVD continues unabated after a myectomy of the ipsilateral inferior oblique muscle. Also, a nystagmus dampening purpose of the vertical vergence is difficult to accept in view of the fact that a latent cyclovertical nystagmus is not a consistent feature of DVD. Finally, if DVD were elicited to dampen a latent nystagmus we would expect the nonfixating eye of a patient with DVD to elevate each time a patient with latent nystagmus reads his or her threshold acuity line on the office chart. Not only is this not the case but, on the contrary, DVD manifests itself typically when patients are daydreaming and uninvolved in active visual activities.

Van Rijn and coworkers felt that DVD represents a form of asymmetrical vertical heterophoria and could be considered as enhancement of a phenomenon that is present in normal subjects as well. They showed in patients with alternating hyperphoria, who have a right hyperphoria with the left eye fixating and a left hyperphoria with the right eye fixating, asymmetries of the vertical heterophoria angles, depending on which eye was fixating. It is true that alternating hyperphoria, which, incidentally, is an extremely rare clinical finding, bears a superficial resemblance to DVD. However, it is debatable whether these conditions are as closely related as assumed by these authors. A vertical heterophoria becomes manifest as soon as fusion is disrupted and the eye drifts into its anomalous position of rest. In DVD, the vertical movement is caused by an active vergence innervation and fusion is not a factor in controlling the deviation because it may occur in patients without the ability to fuse.

Some authors have speculated that DVD is a manifestation of atavistic oculomotor reflexes that are present in birds and fish. Crone considered that DVD may represent a phylogenetic residuum of monocular vertical movements present in birds and inhibited in normal humans. Brodsky suggested that DVD is a primitive dorsal light reflex in which asymmetrical visual input to the eyes evokes a vertical divergence movement. In lateral-eyed animals this reflex serves as a primitive visual-vestibular righting response. Suppressed in normal humans, it is thought to manifest itself when early-onset strabismus precludes normal binocular development. Can the stimulus for the dorsal reflex in fish be compared to the stimulus situation in a human strabismic infant? A fish illuminated from one side depresses the eye ipsilateral to the light source and elevates the contralateral eye. The stimulus for this reflex eye movement, which does indeed resemble a vertical vergence response, is a difference in illu-

![FIGURE 18-4. Dissociated vertical deviation in different gaze positions. A, In the case of a dissociated vertical deviation of the left eye, elevation occurs in adduction, primary position, and, to a lesser degree, in abduction. B, When the elevated adducted left eye takes up fixation, the covered right eye will be elevated to an equal degree. (From Noorden GK von: Atlas of Strabismus, ed 4. St Louis, Mosby-Year Book, 1983.)](image-url)
mination between the two eyes. However, such differences do not occur in strabismus, as each eye receives the same amount of light. In strabismus, the asymmetry of visual input that disrupts binocularity is caused instead by the incongruity of the retinal images formed in each eye.

To summarize, it is fair to state that despite numerous attempts to clarify it, the etiology of DVD is still obscure. Bielschowsky’s original explanation of this form of strabismus, confirmed by modern eye movement recording techniques, has established indisputably that DVD is a verticalvergence eye movement. However, the stimulus for this movement and its relationship to various forms of strabismus, especially to essential infantile esotropia, have yet to be convincingly identified.

**Differential Diagnosis**

Even though the pattern of the deviation and the results of the red-glass test are clearly different in DVD from those in other cyclovertical anomalies, clinicians sometimes confuse this condition with upshoot in adduction caused by overaction of the inferior oblique muscles (see p. 386). To be sure, overaction of the inferior oblique muscle may occur in patients with DVD, but such overaction cannot be held responsible for this anomaly. Several clinical findings clearly distinguish DVD from overaction of the inferior oblique muscle.

*First*, in DVD the covered eye becomes elevated in abduction, primary position, and adduction (Fig. 18–4). Conversely, with overaction of the inferior oblique muscles, each eye becomes elevated primarily in adduction and never in abduction unless there is coexisting contracture of the ipsilateral superior rectus muscle. Unlike DVD, overaction of the inferior obliques is commonly associated with a V-pattern esotropia. The reason for elevation in adduction in a DVD is that the adducted eye becomes occluded by the nasal bridge and fusion is suspended. In children under the age of 2 to 3 years the nasal bridge has not yet fully developed and the upshoot in adduction is rarely seen.

*Second*, DVD is found also in patients in whom there is no noticeable overaction of the inferior oblique muscles and actually occurs frequently in those with underacting inferior oblique and overacting superior oblique muscles\(^{45, 71}\) (see Fig. 18–1).

*Third*, when a patient with an overacting inferior oblique muscle fixates with the involved eye in the field of action of that muscle (elevation and adduction), the contralateral superior rectus muscle will underact (see Fig. 20–1). This apparent paresis of the superior rectus muscle has been discussed under inhibitional palsy (Chapter 20). Conversely, in patients with DVD who are tested in the same manner, underaction of the contralateral yoke muscle does not occur (Fig. 18–4, B).

*Fourth*, in patients with inferior oblique overaction, the speed of the refixation movement of the eye after covering the fellow eye is rapid (200° to 400°/s) compared with the much slower infraduction movements in patients with DVD, which are usually between 10° and 200°/s.\(^{46}\)

*Fifth*, the characteristic slow, tonic incycloduction of the eye as it returns from the dissociated to the primary position cannot be observed with equal facility when overaction of the inferior oblique is present. In that case refixation after covering the fixating eye is also accompanied by incycloduction but this movement is so fast that it often escapes observation.

The differential diagnosis between these two conditions is summarized in Table 18–1 in which additional distinguishing findings are listed. Clear distinction between them is clinically important; for example, a recession or myectomy of the inferior oblique will have no effect on upshoot in adduction if the patient actually has a DVD.

When associated with comitant or paretic cyclovertical anomalies the diagnosis of DVD is more difficult. When evaluating such patients, one must take into account the starting position of each eye before the cover is applied. For instance, if a right hypertropia is associated with a DVD, the right eye will become further elevated under the cover, and the fellow left hypotropic eye when covered will move upward the same amount but may only reach the midline, since it began its movement from a depressed position. Careful observation of each eye before, during, and after the cover has been applied is essential to detecting a dissociated vertical component in a patient with a comitant or paretic cyclovertical deviation.

**Therapy**

During the first half of the twentieth century clinicians took a rather passive attitude toward treatment of DVD. This conservatism probably finds its roots in Bielschowsky’s teachings that this
TABLE 18–1. Differential Diagnosis: Dissociated Vertical Deviation vs. Inferior Oblique Overaction

<table>
<thead>
<tr>
<th>Dissociated Vertical Deviation</th>
<th>Inferior Oblique Overaction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Elevation</td>
<td>From primary position, adduction and abduction</td>
</tr>
<tr>
<td>Superior oblique action</td>
<td>May overact</td>
</tr>
<tr>
<td>V pattern</td>
<td>Absent</td>
</tr>
<tr>
<td>Pseudoparesis of contralateral superior rectus</td>
<td>Absent</td>
</tr>
<tr>
<td>Incycloduction on refixation</td>
<td>Present</td>
</tr>
<tr>
<td>Saccadic velocity of refixation movement</td>
<td>10°–200°/s</td>
</tr>
<tr>
<td>Latent nystagmus</td>
<td>Often present</td>
</tr>
<tr>
<td>Bielschowsky phenomenon</td>
<td>Often present</td>
</tr>
</tbody>
</table>

condition does not lend itself to optical or operative therapy as do comitant or paretic deviations. Bielschowsky cited cases of torsional diplopia produced by surgery on the superior rectus muscles, and stated that therapy, if contemplated at all, should be directed toward strengthening the fusional mechanism.

Patients with DVD usually are asymptomatic, and complaints of diplopia have been an infrequent problem in our experience; however, when the patient is daydreaming or tired, the elevated position of either eye may become conspicuous and a source of embarrassment to the patient. In that case surgery may be considered. We have not observed DVD in adults as often as we have in children, and were, perhaps erroneously, under the impression that this disorder tends to improve with time. However, Harcourt and coworkers followed 100 patients with DVD for as long as 7.3 years and found no significant decrease in the deviation during this period of observation.

Surgical procedures preferred by various authors are (1) recession of the superior combined with resection of the inferior rectus muscles, (2) resection of the inferior recti, (3) retroequatorial myopexy (posterior fixation) of the superior recti combined with a recession of these muscles, (4) unconventionally large recessions (7 to 10 mm) of the superior recti, and (5) anterior displacement of the inferior oblique insertion, which may be combined with superior rectus recession.

Our initial enthusiasm for using a conventional (4 to 5 mm) recession of the superior recti combined with a retroequatorial myopexy 12 to 15 mm behind the original insertion has waned because of many recurrences occurring as late as several years after an initial satisfactory result. Currently, we prefer 7- to 9-mm recessions of the superior recti and vary the amount of surgery in the two eyes when the deviation is asymmetrical. This approach has yielded a cure or significant improvement (defined as a cosmetically insignificant residual angle) in 23 (72%) of 32 patients after a follow-up of at least 3 years. Contrary to our earlier concern that such an extensive weakening procedure would produce a paresis of the superior rectus, we have not observed this complication. The effectiveness of superior rectus muscle recession in terms of correction of a deviation in prism dipters per millimeter of recession in other forms of vertical strabismus and the lesser effect of this procedure in a DVD of the same magnitude emphasize the unique position of this anomaly among other forms of strabismus.

Several authors have in recent years reported good results with anterior displacement of the inferior oblique muscle and this treatment has become the procedure of choice for some. However, we prefer recession of the superior rectus muscle(s), which is less likely to cause some of the complications reported after surgery on the inferior oblique insertion (see Chapter 26).

The question has been debated whether surgery should always be performed in both eyes even though the deviation may be present preoperatively only in one eye. It is not uncommon and quite disappointing to have a patient return after surgery in one eye with a DVD in the fellow eye. However, since in our experience this does not happen in every instance of asymmetrical occurrence we operate on both eyes only when a deviation can be diagnosed preoperatively in both eyes.

Recurrences are not uncommon even after unconventionally large recessions of the superior recti and require additional surgery consisting of a 4- to 5-mm resection of the inferior rectus muscle. Full correction or improvement, as defined above, can in our experience be achieved in 92% of the patients after this operation.
Although in most patients with a conspicuous DVD surgery is the recommended treatment, the possible effectiveness of a conservative approach should not be ignored. This is especially true in patients with asymmetrical involvement or those accustomed to wearing glasses. For example, a patient without binocular vision (after horizontal surgical alignment in infantile esotropia) may exhibit a significant DVD of the left eye when fixating with the right eye, but only an insignificant deviation of the right eye may occur with the left eye fixating. A slight optical blur induced by increasing the power of the lens over the right eye (+2.00 sph usually is sufficient) or a contact lens (see Case 24–1, p 538) will switch fixation preference to the left eye, and the DVD is no longer a cosmetic problem. Simon and coworkers have110 confirmed the efficacy of this approach in selected cases but have used atropine rather than optical penalization.

**Dissociated Horizontal Deviations**

It has only recently been recognized that DVD may also have a horizontal component. Raab92 mentioned in 1970 that the vertical movement in DVD may be accompanied by abduction. Little attention was paid to this observation until the term **dissociated horizontal deviations (DHDs)** became established in the literature.30, 113, 130, 134

The condition is characterized by intermittent, asymmetrical abduction and elevation of the dissociated eye (see Fig. 18–2A). Occasionally, DHD occurs in an isolated form, not accompanied by a vertical deviation. As with DVD, latent nystagmus and excyclotropia of the deviated eye are frequently associated findings. Interestingly, and perhaps significantly, Wilson and coworkers132 found that only two of six patients with a prominent DHD had a history of essential infantile esotropia. The remainder had accommodative esotropia, a condition that is only infrequently associated with DVD. These authors also reported that when DHD is associated with esotropia the patient may become exotropic during periods of visual inattention.

In an earlier report Wilson and McClatchey130 had pointed out that unlike in ordinary intermittent exotropia, the alternate cover test reveals less exodeviation than when the eye abducts spontaneously or under cover, that the fixating eye may adduct during attempts to neutralize the horizontal deviation with base-in prisms, and that the exodeviation may be strictly unilateral. Moreover, they described what is similar to the Bielschowsky phenomenon in DVD: when neutral density filters are placed before the fixating eye the abducted dissociated eye returns to the primary position and may even adduct. Since we became aware of this condition we have observed several patients with a pure dissociated horizontal deviation in one eye and a pure DVD in the fellow eye.

For dissociated exodeviations a 5- to 7-mm recession of the lateral rectus muscle of the involved eye is recommended when the size of the deviation is such that the patient or his or her parents desire correction. Satisfactory results have been reported using this approach.131

This may be combined with recession of the superior rectus when associated with a vertical component. In patients whose dissociated deviation is predominantly vertical with only a small horizontal component, we have found a large recession of the superior rectus is usually sufficient to correct both problems.

Spielmann115 described **dissociated esodeviations** that occur when either eye is covered with the semiopaque occluder. When both eyes are occluded (fixation-free position) the eyes remain aligned, which distinguishes this condition from esophoria. In our experience dissociated esodeviations occur much less frequently than dissociated exodeviations. If the deviation becomes intermittent and a cosmetic consideration, a posterior fixation of the medial rectus muscle of the involved eye 14 mm behind its insertion is effective. Observation rather than surgery has been advocated when the esotropia changes to exotropia during visual inattention.132

In view of the great clinical similarity of dissociated vertical, torsional, and horizontal deviations, we agree with those who consider these strabismus forms as variations on the same theme rather than as different entities sui generis.114, 134

**Elevation in Adduction (Strabismus Sursoadductorius)**

**Clinical Characteristics**

When examining the versions, one may find elevation of an eye as it moves toward adduction (Fig.
18–5). Once in a position of maximal elevation in adduction, the eye will be elevated further than a normal eye. This anomaly may be unilateral or bilateral and has been termed strabismus surosaicductorius or, if the opposite situation occurs, that is, the eye shows depression in adduction, strabismus deorsosaicductorius. These Latin terms have never become popular in the English strabismologic literature where they are used in their translated forms, elevation (or upshoot) in adduction and depression (or downshoot) in adduction. Upshoot in adduction in its bilateral form is characterized by left hypertropia in dextroversion and right hypertropia in levoversion (double or alternating hypertropia), but a vertical deviation is present infrequently in primary position. Upshoot in adduction is an isolated phenomenon or occurs with esotropia or exotropia, often associated with a V pattern (see Chapter 19). It is frequently observed in infantile esotropia. It is often automatically and erroneously assumed that elevation in adduction is caused by inferior oblique overaction, that is, by excessive innervation of that muscle. While this is often the case, we shall see that there are other causes for this condition.

Etiology

OVERACTION OF THE INFERIOR OBLIQUE MUSCLE. It has been customary to distinguish between primary and secondary overactions of this muscle. Primary overaction of the inferior oblique muscle in which there is no evidence for a past or present ipsilateral superior oblique paralysis or paresis is difficult to explain. A V-pattern type of strabismus is often present in such patients and, typically, the Bielschowsky head tilt test is negative. This condition occurs frequently in essential infantile esotropia.

The explanations given for apparent primary overaction in the older literature are vague, to say the least. Duane,25 for instance, suggested that there is normally an upshoot of the adducted eye because of the greater mechanical advantage of the inferior oblique muscle of the adducted eye over the superior rectus muscle of the abducted eye. It is of interest in this connection that Lisch and Simonsz66 have reported in normal subjects up- and downshoot in adduction after prolonged monocular patching. This may suggest that there is a natural tendency for elevation and depression in adduction to occur but that under normal conditions such eye movements are controlled by fusion.66 Scobee104, p.378 agreed that overaction of the inferior oblique muscle is normal because of the increased impulse required by the mechanically disadvantaged superior rectus muscle. This strong impulse is communicated to its yoke muscle, the inferior oblique of the adducted eye hidden behind the nose. He also stated that the elevating action of the inferior oblique muscle in adduction is greater than the depressing action of the superior oblique muscle. Therefore there would be an imbalance if the eyes should be dissociated by the nose, and the result would be an upshoot of the adducted eye. Lancaster65 agreed with this view. Guibor40 suggested that inferior oblique overactions could be caused by a synkinesis of that muscle with the ipsilateral medial rectus muscle owing to an impulse spread within the central nervous system.

None of these older explanations are convincing and it remains quite doubtful whether a true primary overaction of an oblique muscle on an innervational basis exists at all. The discovery of muscle pulleys (see below) has directed our attention to other etiologic possibilities for this apparent overaction. We are in agreement with Clark and coworkers16 who lamented the use of diagnosis-laden terms for ocular motility disorders except in cases where the etiology is clear. For this reason and because there are several causes for elevation in adduction that are unrelated to excessive inferior oblique muscle contraction, we recommend that the terms primary overaction of the inferior oblique (or, for that matter, of the superior oblique muscle) should be abandoned in favor of the more generic elevation (or upshoot) in adduction or depression (or downshoot) in adduction.

Secondary overaction of the inferior oblique

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**FIGURE 18–5.** Elevation in adduction. Marked left hypertropia in dextroversion and right hypertropia in levoversion. No vertical deviation in primary position.
muscle is easier to understand and is caused by paresis or paralysis of the ipsilateral superior oblique muscle or by paresis or paralysis of the contralateral superior rectus muscle when the patient fixates with the paretic eye. In the latter situation the upshoot in adduction is actually caused by increased innervation flowing to the inferior oblique muscle, according to Hering’s law. However, in the former condition the upshoot in adduction is not caused by excessive innervation of the inferior oblique muscle but by a lack of tonus of its paralyzed antagonist. In this situation a normal innervational impulse will suffice to cause the eye to overshoot in the field of action of the inferior oblique.

A similar situation exists when the balance of forces between superior and inferior oblique muscles is offset by anatomical rather than innervational causes, as in plagiocephaly (see Chapter 19). Here, the recessed trochlea has changed the plane of the superior oblique tendon, which places the inferior oblique at a functional advantage over the superior oblique, which causes an upshoot in adduction.

In the following discussion we shall see that elevation in adduction may be caused by a number of other factors.

HETEROTOPIA OF RECTUS MUSCLE PULLEYS. Recent work by Clark and coworkers has provided evidence that what appears as primary overaction of an oblique muscle may actually be a secondary overaction of both elevators caused by heterotopia of muscle pulleys (see Chapter 3). Because of the significance of these findings with respect to the etiology of A and V patterns they will be discussed in Chapter 19

OCULAR AND ORBITAL TORSION. For a discussion of the roles of ocular and orbital torsion in producing upshoot in adduction, the reader is directed to Chapter 19, where this mechanism is discussed in connection with the etiology of A- and V-pattern strabismus. It will suffice to say here that excyclotropia of an eye may cause elevation in adduction and depression in abduction in the absence of increased innervation of the oblique muscles (Fig. 18–6). When the eye is excyclotorted the medial rectus muscle will no longer act as a pure adductor but will gain elevating action as well. Thus, medial rectus contraction will not only adduct but will also elevate the eye (upshoot) under these circumstances. Likewise, the lateral rectus muscle will no longer be a pure abductor but abduct and depress the eye (downshoot). The elevation in adduction is usually more prominent than the depression in abduction, which may even be absent. This may be due to structural differences between the medial and lateral aspects of the orbit. It is of historical interest in this connection that Bielschowsky documented a case of an exotropic patient in whom a right hypertropia caused by an apparently overacting inferior oblique muscle disappeared after merely advancing and lowering the insertion of the medial rectus muscle.

DUANE SYNDROME. Another cause of elevation in adduction, unrelated to inferior oblique overaction, is the result of co-contraction of the horizontal rectus muscle in Duane’s syndrome (see Chapter 21).

DISSOCIATED VERTICAL DEVIATION. Elevation in adduction caused by DVD when fusion is interrupted by the nasal bridge has been mentioned above.

Therapy

When elevation in adduction is caused by an overacting inferior oblique muscle, treatment, when indicated, is surgical and should consist of a weakening procedure on that muscle. In view of the different etiologies for upshoot in adduction discussed above, Spielmann warned against the indiscriminate use of inferior oblique weakening procedures for this condition. It seldom presents a cosmetic problem, considering that the eyes rarely move from primary position more than 15° to either side under casual conditions of seeing. Surgery is done mostly for functional reasons, that is, when the hypertropia in adduction presents an obstacle to fusion in lateral gaze or a V pattern exists that disrupts fusion in upward (V exotropia) or downward (V esotropia) gaze.

In apparently unilateral overaction of the inferior oblique muscle, a careful search should always be made in the fellow eye. After myectomy or recession of an overacting inferior oblique muscle, it is not unusual for overaction in the fellow eye to become manifest.

Depression in Adduction (Strabismus Deorsoadductorius)

As mentioned in the preceding paragraph in connection with elevation, a depression in adduction
(Fig. 18–7) may have more than one cause. Again, we must distinguish between primary and secondary forms. Secondary overaction is well understood and may occur on the basis of paresis or paralysis of the ipsilateral inferior oblique muscle or of the contralateral inferior rectus muscle. Another cause of secondary overaction is contracture of the contralateral superior rectus muscle, which is occasionally seen in conjunction with longstanding paralysis of the contralateral superior oblique muscle (see p. 435). In view of the relative frequency of superior oblique paralyses when compared with paralysis of the inferior oblique and inferior rectus muscles, it is not surprising that depression in adduction occurs less frequently than elevation.

As in the case of so-called primary overaction of the inferior oblique muscle the etiology of
apparently primary overaction of the superior oblique is obscure and, analogously to the former, we prefer the more generic term depression in adduction for this condition. The recent findings of heterotopic muscle pulleys to explain a downshoot on adduction on a mechanical basis has been mentioned (see p. 387). Ocular or orbital incyclotorsion may also cause depression in adduction, similar to the elevation produced by excyclotorsion. Duane’s syndrome with co-contraction of the horizontal rectus muscles and Brown’s syndrome are other causes.

**Cyclovertical Deviations**

In cyclotropia the eyes are misaligned around the anteroposterior axis either as an isolated disturbance of ocular motility or, more frequently, in association with any other form of strabismus. In most instances, cyclodeviations are caused by an imbalance between the muscle pair affecting intorsion (superior oblique and superior rectus muscles) and the muscle pair producing extorsion of the globe (inferior oblique and inferior rectus muscles). Consequently, such deviations are associated almost invariably with paretic or paralytic cyclovertical muscle problems, particularly those caused by dysfunction of the oblique muscles. On the other hand, cyclodeviations also occur in association with DVD, in the A and V patterns of strabismus without an obvious paretic component, in endocrine ophthalmopathy, myasthenia gravis, plagiocephaly, after surgery for retinal detachment, and in heterotopia of the macula, secondary to retinal traction.

In recent years iatrogenic cyclodeviations have been produced surgically as a consequence of macular rotation for age-related macular degeneration.

**Diagnosis**

The diagnosis of cyclodeviations is discussed in Chapter 12.

**Clinical Characteristics**

No studies are available that reflect the prevalence of cyclodeviations. Most ophthalmologists do not routinely test for such anomalies unless the patient specifically complains about torsional diplopia. In the absence of a cyclovertical muscle imbalance such complaints are easily misinterpreted, as pointed out by Kushner. The results of fundus photography (see Fig. 18–6), the Maddox double rod test, and scotometry show that cyclodeviations occur with great regularity and frequency with any disturbance of the oblique and, to a somewhat lesser degree, vertical rectus muscles. Curiously, however, with the exception of paretic conditions of recent onset, particularly traumatic unilateral or bilateral superior oblique paralysis, symptoms related to cyclotropia—such as torsional diplopia, dizziness, and difficulties in negotiating stairways, steps, and street curbs—are seldom encountered in clinical practice.

There are several reasons why patients with cyclodeviations are commonly asymptomatic. First, we must consider that cyclodeviations remain compensated for by cyclofusion through cyclovergences. In such patients the Maddox rods (see Chapter 12) will show various degrees of cyclotropia. However, when tested with Bagolini lenses and the coexisting vertical or horizontal deviations are prismatically corrected, cyclotropia will be absent. This discrepancy in testing results is explained by the fact that Maddox rods disrupt fusion, whereas Bagolini lenses do not. Ruttum and von Noorden pointed out that whereas the Maddox test is of value in substantiating and measuring cyclotropia, it addresses the position of the eye only under dissociated viewing conditions. The Bagolini test result, on the other hand, predicts how a patient will handle a cycloptropia by cyclofusion when coexisting vertical and horizontal deviations are surgically eliminated.

The question whether cyclofusion occurs purely on a sensory basis or has a motor component has been discussed in Chapter 4. It has been claimed in the literature that in cyclophoria the involved eye realigns itself around its anteroposterior axis under the influence of cyclofusion. However, we have been unable to ascertain the presence of such a corrective cycloduction by direct observation (see also Jampel and coworkers) or by comparison of fundus photographs taken under monocular and binocular viewing conditions. Likewise, Locke found no change in the position of the vertically displaced blind spot of cyclotropic patients when perimetry was performed under monocular and binocular conditions. On the other hand, Herzau and Joos noted variations in position of the blind spot during monocular and binocular perimetry on the phase difference haploscope in patients with cyclovertical
strabismus and concluded that cyclofusional movements must exist after all (see also Kolling60). However, the velocity of such movements is slow and their amplitudes are small so that they may easily escape detection with the naked eye.49

**SENSORIAL ADAPTATIONS.** Many patients are unaware of image tilting because of suppression, anomalous retinal correspondence,4 or, in rare instances, a compensatory anomalous head posture.85 However, these mechanisms do not explain the common and puzzling finding that a patient whose eye is found to be rotated around the anteroposterior axis on ophthalmoscopy or fundus photography (see Chapter 12) fails to see a tilted visual environment when the nonparalyzed eye is occluded. The reason for this frequent finding, for instance, in patients with congenital superior oblique palsy, is that adaptations have developed that are quite unique to cyclodeviations.

The older literature contains references to the fact that the spatial response of retinal elements can be reordered along new vertical and horizontal meridians,50, 51 and the famous case of Sachs and Meller100 is cited often in this connection. Ruttum and von Noorden98 and Olivier and von Noorden89 reinvestigated this phenomenon and confirmed that a spatial reorientation of the horizontal and vertical retinal meridians occurs in certain patients with congenital or early acquired cyclodeviations. This spatial adaptation compensates for the image tilt that would otherwise be perceived (Fig. 18–8). It explains why patients with objective cyclotropia may temporarily note a tilting of the environment in the opposite direction after surgical correction of the cyclotropia before normal, innate spatial orientation of the retinal meridians reestablishes itself.78, 83 The practical implication of this finding in connection with postoperative adjustment of a Harada-Ito procedure is mentioned at the end of this chapter.

Even in normal subjects there exists a certain degree of spatial adaptability of the vertical and horizontal retinal meridians and their central connection as demonstrated in the famous “tilt after-effect” experiment of Vernon126 and Gibson and Radner.37 This experiment may be easily repeated by the interested reader: monocular observation for a few minutes of a fixation mark bisecting a line inclined 45° will cause a subsequently viewed vertical line to appear inclined in the opposite direction. Adaptation to a tilted environment probably has a neurophysiologic basis in terms of a change in orientation tuning of the striate cortical neurons12 and suggests a certain degree of cortical plasticity even in visually mature adults.

**PSYCHOLOGICAL ADAPTATION.** Some cyclovertical patients may be unaware of a tilted environment because of empirical spatial clues. Experience has taught us that familiar objects such as doors, windows, houses, and trees have a consistent vertical or horizontal orientation in physical space. Such spatial clues from an orderly visual environment are used to correct for image tilting.50 As soon as this normal frame of visual reference is no longer available, for instance, in complete darkness, these patients become aware of cycloptropia. Ruttum and von Noorden98 confirmed this by measuring the so-called subjective horizontal in cyclovertical subjects. The subjective horizontal is defined as a subject’s perception of a horizontal plane as opposed to its actual position in physical space. Its determination was once a popular diagnostic procedure in the diagnosis of cyclovertical
strabismus. Asymptomatic patients with cyclotropia as diagnosed by fundus photography and Maddox rods may perceive a faintly illuminated horizontal line as tilted when no other visual clues are available. The use of cylindrical lenses with their axis placed so as to offset the cyclotropia has been advocated, but the value of this therapy is highly questionable. The treatment of symptomatic cyclotropia is surgical. When the action of the oblique muscles is abnormal, cyclotropia usually occurs in association with a clinically significant hyperdeviation; thus the choice of muscles on which to operate presents no difficulties, since elimination of the hyperdeviation also will correct the cyclodeviation. For instance, in a patient with paralysis of the homolateral superior oblique muscle, excyclotropia caused by unopposed action of the inferior oblique muscle can be eliminated by a weakening procedure on the inferior oblique muscle that will correct both the hypertropia and the cyclodeviation. Likewise, if the inferior oblique muscles are not overacting and the vertical deviation occurs only in the field of action of the paretic muscle, tucking of the tendon of the paretic superior oblique muscle is similarly effective in eliminating the hypertropia and the excyclotropia.

Management of isolated cyclodeviations in patients without a significant associated vertical deviation presents a special problem. The most frequent cause for an isolated symptomatic cyclotropia is a residual excyclotropia after traumatic trochlear paralysis. A conventional weakening or strengthening procedure on offending cyclovertical muscles may correct the cyclodeviation in such cases, but it will also produce an undesired vertical effect. A procedure is required that affects the cyclodeviation exclusively. This requirement is met by several operations. The advancement and lateralization of the superior oblique tendon for excyclotropia according to Harada and Ito43 (see Chapter 26) and its many variations has become firmly established in our surgical armamentarium. However, this procedure cannot be performed when the superior oblique tendon is congenitally absent or has been previously tenotomized. In that case nasal transposition of the inferior rectus muscle is an effective surgical alternative to correct excyclotropia in downward gaze. When excyclotropia is also present in primary position we add a temporal transposition of the superior rectus muscle. For incyclotropia, which occurs much less frequently than excyclotropia, the inferior rectus is shifted templeward and the superior rectus nasalward. In our hands, the average effect of these operations in terms of rotating the eye around the anteroposterior axis is 10°, ranging from 8° to 12°. Ohmi and coworkers48 reported similar results. Other procedures to correct cyclotropia without producing vertical or horizontal strabismus include slanting of the insertion of all rectus muscles, vertical transposition of the horizontal rectus muscles, and transpositions of the anterior aspects of the inferior and superior oblique tendons.18 The surgical technique for these procedures, as well as surgical induction of cyclotropia to counteract a compensatory head tilt in patients with congenital nystagmus, is discussed in Chapters 23 and 26.

An overcorrection after surgery for cyclotropia (e.g., excyclotropia changing to incyclotropia) usually is only temporary and can be explained by the persistence of sensory adaptation to image tilting. The surgeon should keep this in mind and not be too hasty in planning a reoperation or adjusting the sutures on the first postoperative day if adjustable sutures are used for the Harada-Ito procedure. In fact, a slight overcorrection after the Harada-Ito procedure is desirable since the effect of surgery tends to decrease with time.

A special challenge exists in patients who had vertical macular translocation for improvement of visual acuity of an eye with age-related macular degeneration. This operation invariably causes horizontal and vertical strabismus in addition to
cycloptropia. The magnitude of this iatrogenic incycloptropia is formidable indeed if compared to what is usually encountered in cyclovertical strabismus and may range from as much as 33 degrees26, 29, 36 to 45 degrees.106 It is all the more surprising that spontaneous adaptations have been reported in such patients106, 107 but most will complain about an intolerable shift of the visual environment. Conventional surgery consisting of an advancement and transposition of the inferior oblique and a recession of the superior oblique muscle, as advocated by Conrad and de Decker28 and as employed by others,36 or horizontal transposition as used by us, does not suffice to correct a cyclodeivation of this magnitude. Eckardt and Eckardt,28, 29 recently recommended adding to the surgery on the oblique muscles (according to Conrad and de Decker18) a transposition of strips from two or from all four rectus muscles to the insertion of the adjoining rectus muscles, similar to a Humelshiem procedure. This nearly doubled the cyclorotational effect of the operation to a total of 30° to 40°, which about compensated for the cycloptropia produced by macular translocation. Freedman and coworkers36 advocated combined superior oblique muscle recession and inferior oblique advancement for incycloptropia following macular translocation. A rotation of only a retinal flap rather than the entire retina apparently causes less of a postoperative image tilt, since patients operated on in this fashion adjusted to the tilt without muscle surgery.24, 75 Perhaps this modification, if proved to be as effective as total retinal rotation, will eventually eliminate the need for muscle surgery in conjunction with or following macular translocation.

REFERENCES

Clinical Characteristics of Neuromuscular Anomalies of the Eye