Clinical Characteristics of Neuromuscular Anomalies of the Eye
Esodeviations are caused by innervational or mechanical factors or a combination of both. The various theories for the etiology of strabismus are reviewed in Chapter 9. As is true with other forms of strabismus, an esodeviation may be controlled by fusional divergence (esophoria), intermittently controlled (intermittent esotropia), or manifest (esotropia). In addition to the differences in etiology, other variable characteristics of esodeviations include their state of comitance, the presence of sensorial adaptations, the age of the patient at the onset, the mode of onset, the size of the angle of strabismus, and the state of fixation behavior (unilateral or alternating). Thus, esodeviations are difficult to classify and are never entirely accurately classified, since the various characteristics may overlap in a single group of esotropes. For instance, it is an accepted fact that the characteristics of infantile esotropia are fairly uniform in most patients and are different in those with accommodative esotropia. Yet, accommodative factors may become superimposed in patients with essential infantile esotropia. These reservations notwithstanding, we have found the classification of esodeviations shown in Table 16–1 to be useful for clinical purposes and as a guideline for the student.

Not all forms of esodeviations listed in Table 16–1 are discussed in this chapter. Esotropia associated with A and V patterns is covered in Chapter 19, and the reader is referred to Chapter 20 for a discussion of paralytic esotropia, to Chapter 21 for descriptions of cyclic esotropia and esotropia caused by entrapment of the medial rectus muscle or as part of the Duane retraction syndrome, to Chapter 22 for divergence insufficiency, and to Chapter 23 for a discussion of the nystagmus blockage syndrome.

Esophoria and Intermittent Esotropia

Etiology

The etiology of latent or intermittent deviations is not qualitatively different from that of manifest deviations (see Chapter 9). Accommodative, non-accommodative, and other innervational (dynamic) factors may be involved.

Clinical Signs

As pointed out in Chapter 8, true orthophoria (i.e., the absence of heterophoria at any fixation distance and in any gaze position) is a rarity. Esophoria of a small degree is in fact a common finding in a normal population. Scobee stated that the average normal degree of heterophoria at infinity, as determined with a Maddox rod, is 1.4 and also noted reduced stereoaucity in patients with intermittent esotropia, exotropia, and esophoria. In our experience, this symptom is frequently associated with the intermittency of a deviation, but it is rarely a prominent clinical symptom in patients with a well-compensated heterophoria. The signs and symptoms of heterophoria are discussed in Chapter 10.
TABLE 16–1. Classification of Esodeviations

<table>
<thead>
<tr>
<th>I. Comitant esodeviations</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Accommodative esotropia</td>
</tr>
<tr>
<td>1. Refractive accommodative esotropia (normal AC/A)</td>
</tr>
<tr>
<td>2. Nonrefractive accommodative esotropia (high AC/A)</td>
</tr>
<tr>
<td>3. Hypoaccomodative esotropia (reduced NPA)</td>
</tr>
<tr>
<td>4. Partially accommodative esotropia</td>
</tr>
<tr>
<td>B. Nonaccommodative esotropia</td>
</tr>
<tr>
<td>1. Infantile esotropia</td>
</tr>
<tr>
<td>2. Nonaccommodative convergence excess (normal AC/A)</td>
</tr>
<tr>
<td>3. Acquired (basic) esotropia</td>
</tr>
<tr>
<td>4. Acute-onset esotropia</td>
</tr>
<tr>
<td>5. Divergence insufficiency or paralysis*</td>
</tr>
<tr>
<td>6. Cyclic esotropia*</td>
</tr>
<tr>
<td>7. Recurrent esotropia</td>
</tr>
<tr>
<td>C. Microtropia</td>
</tr>
<tr>
<td>1. Primary microtropia</td>
</tr>
<tr>
<td>2. Secondary microtropia</td>
</tr>
<tr>
<td>D. Nystagmus &quot;blockage&quot; syndrome*</td>
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<table>
<thead>
<tr>
<th>II. Incomitant esodeviations</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Paralytic</td>
</tr>
<tr>
<td>B. Nonparalytic</td>
</tr>
<tr>
<td>1. A- and V-pattern esotropia</td>
</tr>
<tr>
<td>2. Retraction syndrome</td>
</tr>
<tr>
<td>3. Mechanical-restrictive esodeviations</td>
</tr>
<tr>
<td>a. Congenital fibrosis of extraocular muscles</td>
</tr>
<tr>
<td>b. Acquired restriction (endocrine myopathy, trauma to orbital wall, excessive resection of medial rectus muscle(s), myositis, strabismus fixus)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>III. Secondary esodeviations</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Sensory</td>
</tr>
<tr>
<td>B. Consecutive</td>
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</tbody>
</table>

AC/A, accommodative convergence/accommodation ratio; NPA, reduced near point of accommodation.
*Forms of esotropia discussed in other chapters of this book.

Symptoms

Unless a heterophoria is intermittent, in which case the patient may be aware of periodic double vision, the symptoms are mainly asthenopic (see Chapter 10) and related to visual demands made on the eyes. In other words, asthenopic complaints occurring in the morning or after periods of rest are rarely caused by heterophorias. Whether a heterophoria becomes symptomatic or is well tolerated by the patient depends largely on the fusional reserve (i.e., in the case of esophoria, on the amplitude of fusional divergence). The point has been made (see p. 206) that the heterophoric position must be taken into account when measuring fusional vergences with rotary prisms.

Sensorial Adaptation

Sensorial adaptation is not difficult to explain in heterophoric subjects with intermittent deviations. Suppression or anomalous retinal correspondence develops in young patients as an adaptation to diplopia that is present during periods when the ocular deviation is manifest.

Flynn and coworkers reported on foveal suppression under binocular conditions of viewing in patients with heterophorias and intermittent heterotropias. Using entoptic images (Haidinger’s brushes and afterimages), they demonstrated in their patients not only foveal suppression but also what must be interpreted as instability of foveal visual directions. In fact, in several instances a minute angle of anomalous retinal correspondence was demonstrated. These authors emphasized that suppression in heterophoria may present a real obstacle to a functional cure. Stangler’s observation that heterophoric patients were unsuccessful in superimposing a vertical afterimage produced in one eye on a real object fixated by the other eye also indicates an instability of foveal visual directions in this condition.

The question must be asked, Why do heterophoric subjects develop suppression or even anomalous retinal correspondence if the deviation is controlled by fusion, and what is the purpose of sensorial adaptation under such circumstances? There are two possible answers. First, the deviation in such patients at times and under certain circumstances may become manifest (intermittent strabismus), making sensorial adaptation necessary to avoid diplopia. If fusion is artificially disrupted, as shown in the tests used by Flynn and coworkers and by Stangler, suppression and anomalous retinal correspondence will become manifest. Second, in heterophoric patients with suppression, an intermittent heterotropia may be on the verge of developing. It is possible that suppression may then prevail to avoid foveal diplopia. If fusion is artificially disrupted, as shown in the tests used by Flynn and coworkers and by Stangler, suppression and anomalous retinal correspondence will become manifest. Second, in heterophoric patients with suppression, an intermittent heterotropia may be on the verge of developing. It is possible that suppression may then prevail to avoid foveal diplopia, and fusion is maintained by peripheral retinal stimulation only. We believe that deficient stereopsis in heterophoric patients may be explained on the basis of this suppression since it is known that foveal suppression of one eye under binocular conditions of viewing will cause a decrease in stereacuity. Shippman and Cohen suggested that in patients with esophoria, stereacuity is better with uncrossed than with crossed disparity as determined by means of the Wirt stereotest.

Diagnosis

The diagnosis of heterophoria is discussed in Chapter 12, and only a few additional comments
need to be made here. The clinician must be aware that heterophorias can be measured with only approximate accuracy. For instance, the Maddox rod test measures the basic deviations when fusion is disrupted, but it does not determine that part of the deviation caused by dynamic factors such as accommodative convergence. Thus, in a symptomatic patient with esophoria, the deviation must be measured with the prism cover test at 33 cm while the patient reads letters of an appropriate small size at random. In fact, failure to control accommodation at near fixation may lead to an erroneous diagnosis, as illustrated by the following case.

**CASE 16–1**

A 27-year-old woman who had suffered a mild concussion in an automobile accident 6 months earlier complained of intermittent diplopia at distance and blurred vision when trying to read and related these complaints to the injury. Litigation of this case involving a considerable sum of money was pending. Examination by another ophthalmologist had revealed a visual acuity of 6/6 OU and entirely normal ocular findings except for an intermittent esotropia of 20° at distance fixation. She was reported to have orthophoria at near fixation. Ductions and versions were normal. A diagnosis of “divergence paralysis,” probably related to the trauma, was made. An extensive radiographic survey of the skull and a neurologic examination were ordered. When this patient was seen by us in consultation, the same measurements (20° intermittent esotropia) were obtained at distance fixation. At near fixation the alternate cover test failed to reveal a shift; however, the patient indicated that the fixation target held at 33 cm before her eyes appeared blurred. When she was asked to identify small letters at that distance, vision suddenly cleared and an intermittent esotropia of 25° appeared. On further questioning, the patient admitted that she sometimes saw double at both distance and near fixation and that she had learned to avoid diplopia at near vision by “relaxing her eyes.” The diagnosis of divergence paralysis could no longer be supported. We believed that the patient had an esophoria and that the deviation at near was initially missed because the patient was not accommodating on the fixation target. She was treated with base-out prisms, which almost instantly eliminated her complaint, and a resection of both lateral rectus muscles eventually brought a good result.

Case 16–1 illustrates that some patients prefer to see blurred and single by relaxing their accommodation rather than sharp and double. When measuring heterophoria with the prism and cover test, it is important to repeatedly cover each eye for several seconds and to switch the cover rapidly from eye to eye to suspend completely the influence of fusional vergence (see Chapter 12). This technique will often reveal larger amounts of basic esodeviation than originally suspected. In doubtful cases, prolonged monocular occlusion for days or even weeks has been advocated to disclose heterophorias that are not at once evident during the alternate cover test. It is erroneous to assume that momentary disruption of fusion by covering each eye in a rapid fashion will totally exclude a strong innervational tonus such as the one elicited by the compulsion to fuse.

Once the type and size of a heterophoria have been determined, the patient’s ability to cope with an ocular imbalance must be evaluated by measuring fusional amplitudes (see Chapter 12).

**Therapy**

The principle of treating esophoria and intermittent esotropia is the same as for all other forms of latent and intermittent deviations, that is, to create conditions that will allow the patient to enjoy comfortable and functionally complete binocular vision. Depending on the individual case, this goal can be approached by using one of several modes of therapy. Before discussing these methods, we would again like to stress that esophoria per se requires no therapy unless asthenopia or evidence of deterioration of binocular functions also is present.

A symptomatic esophoric patient in whom re-fraction reveals a significant amount of hypermetropia (at least +1.25 D sph) is treated by full correction of the hypermetropic refractive error in the same manner as in an esotropic patient. Patients with a high accommodative convergence/accommodation (AC/A) ratio and a symptomatic esophoria without hypermetropia may be considered for bifocal lenses or miotics. For details regarding this mode of therapy, see Chapter 24.

Prisms base-out may be helpful as a “crutch” in regaining visual comfort in patients with nonaccommodative esophoria. When prescribing prisms for esophoria, however, one must clearly understand that this mode of therapy does not cure the ocular imbalance; it only creates temporary conditions that enable the patient to cope more comfortably with the deviation. Correction of only one half to one third of the angle of deviation
with prisms is advisable to prevent total inactivity of the fusional divergence mechanism. Since partial or complete correction of esophoria with prisms will place fewer or no demands on fusional divergence, such patients may become increasingly dependent on their prisms. The use of correcting prisms, particularly in patients with esodeviations of dynamic origin, will result eventually in an increase of the esophoria, and prisms of increasing power will be required to create visual comfort. These objections notwithstanding, there is a place for prismatic correction in elderly patients with symptomatic esophoria who do not respond to orthoptics. In younger patients, we use prisms occasionally in those in whom fusion must be maintained until surgery can be performed (prismatic orthophorization).

Surgery should be considered only when the size of the deviation in a patient with esophoria or intermittent esotropia falls within the range (at least \(12^\circ\)) that can be corrected without fear of overcorrection. Prerequisites for planning surgery are stability of the deviation after full correction of the hypermetropic refractive error and the presence of muscular asthenopia. When the decision has been made to operate, the surgeon should determine the extent of surgery necessary and select the muscles to be operated on in the usual manner and as outlined in Chapter 26.

On numerous occasions, we have witnessed unwarranted timidity in the surgical approach to latent or intermittent esodeviations (or exodeviation, for that matter), apparently based on the misconception that such patients have “just” a heterophoria (as opposed to a heterotropia), and therefore lesser amounts of surgery are required. This attitude is erroneous, of course, and causes undercorrections that, because of their small size, may be difficult to control by additional surgery. The amount of surgery must be aimed at the basic deviation and on the goal to align the eyes, regardless of whether it is a latent, intermittent, or manifest deviation! It is preferable to establish a secondary exophoria rather than be left with a residual esophoria. Convergence fusional movements and also voluntary convergence are more effective than the divergence mechanism in keeping such a residual heterophoria in check.

A conservative approach is indicated when considering surgery for esophoric patients beyond the age of 50 years. In younger patients, small surgical overcorrections present no problem and usually are easily compensated for by fusional convergence. On the other hand, a consecutive esodeviation, regardless of how small, can cause considerable and often insurmountable difficulties in older persons. The elasticity of the fusional apparatus in overcoming motor obstacles in binocular vision tends to decline with advancing age. Experience has taught us to treat such patients with prisms; surgery should be contemplated only reluctantly and after all other therapeutic possibilities have been exhausted.

### Accommodative Esotropia

An esotropia caused by an increased accommodative effort or an abnormally high AC/A ratio is referred to as “accommodative” esotropia. However, several subgroups of accommodative esotropia exist and must be clearly differentiated as each requires different clinical management.

#### Refractive Accommodative Esotropia (Normal AC/A Ratio)

**Definition**

Refractive accommodative esotropia is defined as an esotropia that is restored to orthotropia at all fixation distances and in all gaze positions by optical correction of the underlying hypermetropic refractive error.

**Etiology**

The relationship between accommodation and convergence and the role of an uncorrected hypermetropic refractive error in causing a comitant esodeviation was discussed earlier (see p. 139). At this juncture it is useful to summarize the etiologic components and to mention why some patients with uncorrected hypermetropia do and others do not develop esotropia (Fig. 16–1). Most patients with uncorrected hypermetropia will attempt to clear the image blur by increasing accommodative effort that will, in turn, cause excessive accommodative convergence. If fusional divergence is insufficient to compensate for this impulse to converge the eyes (Fig. 16–1A) and in the presence of a normal or high AC/A ratio, esotropia will develop. If fusional divergence amplitudes are sufficient to cope with the induced esodeviation, an esophoria will be produced (Fig. 16–1B). In the presence of a low or flat AC/A

ratio, the patient may remain orthotropic since the convergence induced by excessive accommodation is normal or even subnormal\(^{209}\) (Fig. 16–1C). Finally, some patients with uncorrected high hypermetropia may remain orthotropic because they prefer blurred vision over the constant effort to accommodate excessively. Such patients may develop a mild form of pattern deprivation amblyopia in both eyes (ametropic amblyopia; see p. 252) or an accommodative deficiency with a reduced near point of accommodation,\(^{209}\) or both (Fig. 16–1D). Whether the visual system reacts with esotropia and clear vision or with orthophoria and blurred vision may depend less on the degree of hypermetropia than on the child’s personality. It is our impression, which needs to be substantiated by an appropriate study, that the former group often encompasses fastidious and exacting children and that the latter group is more relaxed and easygoing, as highlighted by Case 16–2.

CASE 16–2

An 8-year-old girl and her 6-year-old brother were brought to our office for an eye examination. The girl had a history of a gradual onset of esotropia at the age of 3 years and had worn glasses since that time. The boy had no apparent strabismus, but had failed a school vision screening test. The girl had an esotropia of 35\(^{\circ}\) at near and distance fixation without glasses. She was wearing a hypermetropic correction of +5.00 sph in both eyes, which fully corrected her esotropia. Her corrected visual acuity was 6/6 in each eye. The boy had the same refractive error (confirmed by cycloplegic refraction in both children), but was orthophoric with and without glasses.
His best corrected visual acuity was 6/15 in each eye, which improved to 6/9 after wearing glasses for 6 weeks. His AC/A ratio, determined by the gradient method over a range of 6D was 0. The girl was a keen observer and asked numerous questions during the examination. The boy remained silent and rather passive. When we asked the mother to describe the most pertinent personality traits of her children, she replied, "She is the absolute perfectionist, meticulous in every respect. He is completely relaxed, quite sloppy, and does not care about a thing in the world."

Clinical Characteristics

As a rule, the onset of accommodative esotropia, whether refractive or nonrefractive, is between the ages of 2 and 3 years. The onset also may be delayed until adolescence or even adulthood, when it is often precipitated by a brief period of occlusion (see acute strabismus, p. 338). We have also seen children of 1 year or less with all the clinical features of accommodative esotropia, and Pollard\textsuperscript{237} reported two infants with hypermetropia in whom esotropia developed at 4 1/2 and 5 months of age (see also Coats and coworkers\textsuperscript{49} and Haver-tape and coworkers\textsuperscript{111}) and whose eyes became completely aligned following correction of the refractive error. Baker and Parks\textsuperscript{14} reported additional cases and pointed out that after initial control of esotropia by means of glasses, approximately 50% of these patients developed nonaccommodative esotropia. In such cases, surgical intervention may become necessary. These authors also reported that bifoveal fusion does not develop in patients with refractive accommodative esotropia of early onset (monofixation syndrome; see p. 341) and that their ocular deviation is similar to that in patients with essential infantile esotropia. Whether this sensory deficit can be related to how long the esotropia had been present before correction with glasses is unclear. The observations of these investigators show that the earlier viewpoint, that accommodation is inactive during infancy, can no longer be upheld. In fact, Haynes and coworkers\textsuperscript{113} showed that accommodation may reach the adult level by the fourth month of life.

When refractive accommodative esotropia is present, the ocular deviation is usually variable and larger at near than at distance fixation. The variability of the angle of deviation depends on the general state of the patient (alert or fatigued) and on the amount of accommodation exerted at a given moment. The evolution of accommodative esotropia usually is gradual, and most patients pass through a stage of intermittent strabismus. Asthenopic symptoms, complaints about intermittent diplopia, or closure of one eye when doing close work commonly occurs during development of the disease.

Therapy

The prognosis for restoration of normal binocular function in refractive accommodative esotropia is usually excellent if normal binocular functions existed before the onset of the deviation. Full correction of the hypermetropic refractive error, determined by cycloplegic refraction, is usually all that is required initially for rehabilitation. Patients who have never worn glasses may initially complain about blurring of vision with their optical correction. In this instance a brief period of atropinization to relax accommodation may be required before the glasses are tolerated. The cycloplegic refraction is repeated annually, and the glasses are adjusted when necessary. Although there is a tendency for hypermetropia to decrease as a child gets older the majority of patients require glasses well into adolescence\textsuperscript{283} and beyond.

If the distance deviation is reduced or eliminated by glasses and esotropia remains at near fixation, the AC/A ratio is higher than normal (nonrefractive accommodative esotropia), or the patient has a nonaccommodative convergence excess. If the glasses only partially reduce the angle of strabismus at near and distance fixation, then the strabismus is not purely refractive-accommodative in nature (partially accommodative esotropia).

Abraham\textsuperscript{1} has recommended that miotics be substituted for glasses in certain patients with refractive accommodative esotropia. We ordinarily do not advocate prolonged miotic therapy for this condition except in hyperactive or extremely uncooperative children for whom the incessant replacement of broken or lost spectacles imposes an unbearable financial burden on the parents. We have found it useful, however, to prescribe miotics in lieu of glasses for limited periods during the summer months for children who spend the holidays at the beach or long hours in or near a swimming pool. (For a general discussion of the
use and action of miotics in strabismus, see Chapter 24.)

Dyer advocates surgery in lieu of glasses or to reduce a strong correction. He stated that “at times the risk of a subsequent exodeviation is worthwhile when the patient can enjoy years of straight eyes without glasses or much weaker correction in the glasses.” This controversial philosophy never found a following in this country but has been reendorsed in recent years by Gobin, Béard and coworkers, and others in Europe. Under the influence of these authors it has become common practice in some countries to operate for fully refractive accommodative esotropia, that is, an esodeviation that is fully offset by the appropriate optical correction. Gobin stated that he no longer believes “that hypermetropia is the cause of convergent squint” and that “the accommodative component of the squint disappears” after surgical restoration of binocular vision. Among the reasons given by the proponents of surgery is that many patients corrected with glasses will show a progressive deterioration of binocular vision. It has also been claimed that cyclovertical incomitances such as A and V patterns and dysfunctions of the oblique muscles occur frequently in patients with refractive accommodative esotropia, present significant obstacles to fusion, and must be corrected by surgical desagittalization of the oblique muscles. However, no data have been presented to substantiate these claims. On the contrary, several independent studies have established that functional deterioration of fully refractive accommodative esotropia occurs infrequently and that the diagnoses of dysfunctions of the oblique muscles and of A and V patterns in these patients are caused by inadequate diagnostic and measurement techniques.

Moreover, Schiavi and coworkers have shown that inferior oblique overaction is not a common sign of refractive accommodative esotropia. When present, it does not necessarily herald a negative prognosis for preservation of normal binocularity through glasses. Inferior oblique overaction can develop after loss of alignment in some but not all the decompensated patients. No evidence can be found for a cause-effect relationship between oblique muscle dysfunction and loss of binocularity in refractive accommodative esotropia.

In view of the foregoing we conclude, therefore, that the case for surgery in this condition has not been proved. We reject this therapeutic approach, which is contrary to physiologic principles and clinical experience and may be harmful, as shown by the following case.

**CASE 16–3**

A 25-year-old female schoolteacher consulted us with severe asthenopic complaints. She had worn glasses since early childhood to correct for an esotropia, which was well controlled with spectacles correction. She gave no history of having suffered visual discomfort in the past except when taking her glasses off, which caused her to see double and forced her to close one eye. She went to a “strabismus center” 6 months ago where she was offered surgery as an alternative to her glasses. The patient was not advised of the possible unfavorable consequences of such an operation and enthusiastically agreed to have the procedure done. After muscle surgery her eyes were aligned and she no longer saw double without glasses. However, soon afterward she developed severe visual discomfort consisting of headaches, tearing, and nausea after reading without glasses for longer than 10 minutes. Her old glasses relieved these symptoms, but she had to close one eye since she now saw double with her spectacles. Because of her visual difficulties, she was unable to continue teaching school, an occupation that she had enjoyed very much. Her cycloplegic refraction was +4.75 sph in each eye. She was orthotropic without glasses and developed 18° exotropia at near and distance fixation with her glasses.

We predict that the woman in Case 16–3 is but one of numerous unhappy patients likely to populate the waiting rooms of ophthalmologists in the future if the practice of operating for esotropia that has already been fully corrected with glasses or contact lenses continues.

The use of excimer laser photokeratotomy and particularly LASIK (laser-assisted in situ keratomileusis) in the treatment of accommodative strabismus is being discussed with increasing frequency. At this stage of our knowledge and in view of the lack of long-term results with this and other keratorefractive procedures we are opposed to this treatment in the pediatric age group.

In the rare event of deterioration despite a satisfactory initial response to optical correction of the hypermetropia, a recession of both medial rectus muscles will restore fusion in most instances.
Nonrefractive Accommodative Esotropia (High AC/A Ratio)

Definition

Nonrefractive accommodative esotropia is defined as an esotropia greater at near than at distance fixation, unrelated to an uncorrected refractive error, and caused by an abnormally high AC/A ratio in the presence of a normal near point of accommodation.

Clinical Characteristics

Nonrefractive accommodative esotropia occurs in patients with emmetropia, hypermetropia, or myopia; however, moderate degrees of hypermetropia are encountered most frequently. The etiology is unrelated to the underlying refractive error but is closely linked with an abnormal synkinesis between accommodation and accommodative convergence—the effort to accommodate elicits an abnormally high accommodative convergence response. If motor fusion can cope with the increased convergence tonus at near fixation, an esophoria results. If motor fusion is insufficient, nonrefractive accommodative esotropia will become manifest. Unlike hypocommodative esotropia, discussed in the next section, the near point of accommodation is normal for the age of the patient.

Parks\(^2\) reported an abnormally high AC/A ratio in 46% of 897 children with comitant esotropia. The age of onset of accommodative esotropia in this series ranged from 8 months to 7 years, with an average of 30 months. However, Parks based the diagnosis of a high AC/A ratio on the heterophoria rather than on the gradient method. As pointed out earlier (see p. 91), the widely used and, in our opinion, inadequate heterophoria method to determine the AC/A ratio does not distinguish between a high AC/A ratio and nonaccommodative convergence excess. Thus, the patient group reported by Parks is not clearly defined. It is our clinical impression that most patients with nonrefractive accommodative esotropia present between the ages of 6 months and 3 years.

The diagnosis of nonrefractive accommodative esotropia is based on the presence of a significant esodeviation at near fixation on an accommodative fixation target (see Chapter 11) with the refractive error fully corrected and the presence of a high AC/A ratio as established with the gradient method to distinguish this condition from nonaccommodative convergence excess (see below). The necessity of measuring the angle of deviation at near fixation with accommodation fully controlled in patients with all types of strabismus, especially in this group of patients, cannot be overemphasized. The reason is that children with accommodative esotropia may manage to keep their eyes aligned at near fixation by accommodating only partially or not at all (see Case 16–1). The use of a fixation target that requires full accommodation to identify small details will eliminate this frequent cause of diagnostic error.

Confusion may arise when diagnosing an esotropia with a high AC/A ratio and an esotropia with a V pattern (see Chapter 19) if the angle of strabismus is measured at distance fixation with the eyes in primary position and at near fixation with the gaze lowered. In esotropia with the V pattern, the deviation increases characteristically only in downward gaze regardless of whether the patient fixates at near or distance. With an accommodative esotropia, the deviation will increase at near fixation regardless of the position of the eyes in which the angle of strabismus is measured.

Therapy

Since the near deviation is the primary obstacle to normal binocular vision in patients with nonrefractive accommodative esotropia, the conditions for treatment with bifocal lenses are ideal. For details regarding this therapy see Chapter 24.

Attempts have been made to substitute progressive lenses for bifocal lenses. However, unless the patient looks maximally downward, the add in the lower portion of the lens is too low and accommodation is still being employed in downward gaze. Children may not be inclined to look maximally downward during visual activities at near and although cosmetically preferable, we feel that progressive lenses should not be prescribed for the treatment of accommodative esotropia.

Long-acting anticholinesterase drops have also been advocated but because of side effects are sive lenses for bifocal lenses. However, unless the The principles of bifocal therapy are discussed in Chapter 24. The majority of patients with nonredation is still being employed in downward gaze. bifocals; however, we also have observed a slow deteriorating course of the disease.\(^2\) In such instances and after a patient has initially regained fusion and stereopsis at near fixation with bifocal
correction, the near deviation may increase without obvious cause, becoming first intermittent and eventually manifest in the course of several years.

Cycloplegic refraction performed at that point must exclude the possibility of an increase of the hypermetropic refractive error. In that case, stronger lenses should be prescribed. If this does not correct the deviation, then such patients respond well to a recession or posterior fixation of both medial rectus muscles or a combination of both.\textsuperscript{176, 234, 272, 275} The amount of surgery should be based on the near deviation without fear of causing an overcorrection at distance fixation. Most studies of the results achieved with either of these procedures do not distinguish between accommodative and nonaccommodative convergence excess (see below), making it somewhat difficult to evaluate such reports. We have been satisfied with the results of recession of both medial rectus muscles and no longer use retroequatorial myopexy for this condition.

**Hypoaccommodative Esotropia**

**Definition**

Hypoaccommodative esotropia is defined as an esotropia greater at near than at distance fixation, unrelated to an uncorrected hypermetropic refractive error and caused by excessive convergence from an increased accommodative effort to overcome a primary or secondary weakness of accommodation.

**Clinical Characteristics**

Costenbader\textsuperscript{52} drew attention to this special form of esotropia for which he suggested the descriptive term hypoaccommodative. This form is characterized by a small refractive error, a remote near point of accommodation, a small deviation at distance fixation, but a large esotropia at near fixation. He stated that routine testing of the near point of accommodation in strabismic patients revealed a surprisingly large number in whom the near point was recessed farther than one would expect from the patient’s age. According to Costenbader, such patients must exert an excessive accommodative effort to clear their vision at near and, in so doing, exhibit excessive and undesirable convergence. Clearly, this form of esotropia is accommodative, even though its mechanism differs from that discussed above in connection with refractive accommodative and nonrefractive accommodative esotropia.

Mühlendyck\textsuperscript{192, 193} confirmed Costenbader’s concept of hypoaccommodative esotropia and reported patients with reduced accommodative range, an esotropia at near fixation, asthenopia after prolonged periods of reading, and temporary blurring of vision after switching from near to distance vision. Mühlendyck pointed out that this condition may be one of the causes of reading difficulties in schoolchildren and recommended plus lenses for near vision. According to Mühlendyck, the prevalence of hypoaccommodative esotropia was 3.7% in 3929 patients with strabismus. With the exception of Mühlendyck’s work, hypoaccommodative esotropia has received no attention since Costenbader’s original description.

For many years we were skeptical of the existence of this entity until we became aware that children with an accommodative esotropia who had been treated with bifocals for a long time may have an abnormally low near point of accommodation.\textsuperscript{208} It may be argued that this accommodative weakness may have been caused by prolonged bifocal wear but, alerted by this finding, we since have identified children with an esotropia at near fixation who had a reduced near point of accommodation prior to bifocal therapy. Moreover, Mühlendyck and Goerd\textsuperscript{194} reported the near point of accommodation unchanged in children with hypoaccommodation after bifocal wear of 6 years or longer. On the basis of these observations we believe that Costenbader’s original concept needs to be reinvestigated because it may well deserve its proper place in a classification of esodeviations. The findings that not all children with a remote near point of accommodation become esotropic at near, that a reduced near point of accommodation may actually be associated with convergence insufficiency and exophoria,\textsuperscript{214, 215} and that the increased accommodative effort associated with beginning presbyopia rarely produces a manifest esodeviation do not necessarily present arguments against the validity of Costenbader’s theory.

**Partially Accommodative Esotropia**

**Definition**

An esotropia is partially accommodative when accommodative factors contribute to but do not account for the entire deviation.
Clinical Characteristics

Esodeviations of refractive or nonrefractive accommodative etiology do not always occur in their "pure" forms. A residual esotropia may exist despite full correction of a hypermetropic refractive error or prescription of bifocal lenses or miotics or both. As a matter of fact, the majority of patients with esotropia have a mixed type that is partially accommodative and partially nonaccommodative. This is especially so in essential infantile esotropia.123, 242

Few comments are necessary concerning this form of esotropia, but two points should be made. First, at times in a child with essential infantile esotropia an accommodative element becomes superimposed on the deviation as the child grows older, often accompanied by a larger hypermetropia than was first measured. Indeed, it may be the rule that the nonaccommodative element occurred early in infancy or was connatal, whereas the accommodative component is a later acquisition. Second, the presence of a nonaccommodative element should always raise the question that the basic refractive error is not fully corrected; cycloplegic refraction should be done to rule out this possibility.

Thus it appears that the nonaccommodative element in accommodative esotropia resists etiologic classification. In most instances the deviation is probably congenital with an accommodative element becoming superimposed as the child grows older, but in other cases a nonaccommodative element develops after initial alignment of the eyes with glasses or bifocal lenses. With our present knowledge, we can only postulate that increased convergence tonus or mechanical factors such as secondary contractures of the medial rectus muscles, conjunctiva, or Tenon's capsule may play a role.

Therapy

Amblyopia must be eliminated by appropriate therapy (see Chapter 24), and a full hypermetropic correction should be prescribed. Bifocals or miotics, or a combination of both, are useless since the deviation is only reduced but not eradicated. If it is warranted by the size of the nonaccommodative angle of strabismus, which must be determined while the patient is wearing full correction, surgery should be performed to align the eyes. Conservatism is indicated in hypermetropes of +4.00D sph or more (see p. 324). We want to reemphasize that only the nonaccommodative component of the strabismus should be corrected surgically. Special care must be taken to explain this in great detail to parents, who otherwise may expect that glasses will not be required after the operation.

Nonaccommodative Esotropia

Essential Infantile Esotropia

Definition

We define infantile esotropia as a manifest esodeviation with an onset between birth and 6 months of age, and following the suggestion of the Huigoniers134, p.208 add the modifier essential to emphasize the obscure etiology of this condition and to distinguish it from other forms of esodeviation with an onset at about that time.203 Because infantile esotropia is commonly accompanied by a set of other clinical findings (Table 16–2) it is justified to speak of the essential infantile esotropia syndrome.167

Terminology, Prevalence, Etiology

We prefer the term essential infantile esotropia over the older term congenital esotropia but have no objection to using both terms interchangeably.

<table>
<thead>
<tr>
<th>TABLE 16–2. Clinical Characteristics of Essential Infantile Esotropia</th>
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<tbody>
<tr>
<td><strong>Consistent Findings</strong></td>
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<tr>
<td>Onset from birth–6 mo</td>
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<tr>
<td>Large angle (≥30°)</td>
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<tr>
<td>Stable angle which may increase with time</td>
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<tr>
<td>Initial alternation with crossed fixation</td>
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<tr>
<td>Occasionally also very early fixation preference</td>
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<tr>
<td>No clinically apparent CNS involvement</td>
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<tr>
<td>Asymmetrical optokinetic nystagmus</td>
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<tr>
<td><strong>Variable Findings</strong></td>
</tr>
<tr>
<td>Amblyopia</td>
</tr>
<tr>
<td>Apparently defective abduction</td>
</tr>
<tr>
<td>Apparently excessive adduction</td>
</tr>
<tr>
<td>Up- or downshoot on adduction</td>
</tr>
<tr>
<td>A or V pattern</td>
</tr>
<tr>
<td>DVD/DHD</td>
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<tr>
<td>Manifest-latent nystagmus</td>
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<tr>
<td>Manifest nystagmus (rare)</td>
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<tr>
<td>Anomalous head posture</td>
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<td>Heredity</td>
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CNS, central nervous system; DVD, dissociated horizontal deviation; DHD, dissociated horizontal deviation.
The reason for this preference, which has not gone unchallenged, is the following. *Congenital* is defined as “existing at or dating from birth.” The fact is that esotropia is rarely present at birth even though many parents may insist that this was the case. Comprehensive and independent studies of a total of 2200 newborns have shown that esotropia and, to a much lesser degree, esotropia occur commonly in the neonatal period but are, as a rule, transient and disappear by the age of 3 months. Children who eventually develop the essential infantile esotropia syndrome may actually have been exotropic or orthotropic at birth. A congenital onset according to the definition, if it occurs at all, must be exceedingly rare. It is of historical interest that the English surgeon Edward W. Duffin commented as early as 1840 that he had “not met with a single case of congenital strabismus, though in many instances the deformity has been reported to have supervened a few days after birth. Even children of parents who are affected with strabismus and in whom we might conclude it would be hereditary, do not exhibit any appearance of the deformity for months or perhaps years after birth.”

This should not distract from the probability that hereditary factors play a role in the etiology of this disorder. However, a hereditary component does not make a condition “congenital.” Esotropia with an onset after 6 months of age is referred to as *early acquired esotropia*.

The *prevalence* of essential infantile esotropia was once estimated to be 1% of the population. However, the recent comprehensive longitudinal studies of Helveston and his coworkers have established this number to be closer to 0.1%. Even with this reduced prevalence, essential infantile esotropia is the most common form of strabismus.

The *etiology* of essential infantile esotropia is unknown, and the various theories have been discussed in Chapter 9. We favor a hypothesis according to which various strabismogenic forces impinge on a sensorially normal but immature and therefore functionally imperfect visual system. A normally functioning vergence mechanism is capable of overcoming these forces; delayed development or a defect of the vergence system is not capable of overcoming these forces and esotropia ensues. This view is shared by other authors. This hypothesis is summarized in Figure 16–2. It must be emphasized, however, that this is merely a working hypothesis that may have to be modified or even abandoned as new information becomes available. Several reports, according to which normal binocular vision with random-dot stereopsis may occasionally be restored by early surgical alignment (even in some patients who do not undergo operation), support our assumption that there is no underlying congenital sensory defect preventing a functional cure in these patients.

**Differential Diagnosis**

Essential infantile esotropia is not the only form of esodeviation with an onset during the first 6 months of life. There are other conditions, some truly congenital, that is, present at birth, and others acquired during the first few months of life, like essential infantile esotropia. Among the congenital defects are bilateral abducens paralysis (Chapter 20), Duane syndrome type I, and Möbius syndrome (Chapter 21). Conditions acquired during the first few months of life may be sensory esotropia (see p. 345), refractive accommodative esotropia (see p. 314), the nystagmus compensation (blockage) syndrome (Chapter 23), or esotropia in association with other central nervous system manifestations, such as Down syndrome, albinism, cerebral palsy, mental retardation, and so on. The latter group deserves to be separated from essential infantile esotropia in otherwise normal children because the surgical outcome, in our experience and that of others, is less predictable.

**Clinical Characteristics**

Some disagreement exists among current authors as to the significance of the clinical characteristics and their prevalence in patients with essential infantile esotropia. These variations can sometimes be explained by differences in examination techniques or by geographic differences. Table 16–2 lists what we consider to be the most typical characteristics of this condition, some of which are more or less consistent; others are variable. Ciancia described a group of patients with essential infantile esotropia, latent nystagmus, a head turn toward the adducting eye, and apparently limited abduction in both eyes. This has been referred to as the *Ciancia syndrome*. Lang emphasized the frequent association between early-onset esotropia, dissociated vertical deviation, exycloduction of the nonfixating eye, and abnormal
head posture. In the European literature essential infantile esotropia is therefore frequently referred to as Lang syndrome. Both of these contributions are important because they focused attention on previously neglected aspects of essential infantile esotropia. It is questionable, however, whether these syndromes represent separate entities and we believe that they emphasize some of the more variable features of the infantile esotropia syndrome (see also Helveston118).

**AGE AT ONSET.** The age of a child at the onset of esotropia must be established by history during clinical examination since children rarely are brought to an ophthalmologist before 6 months of age. In evaluating the validity of the history given by the parents, one should remember that mothers often tend to overlook strabismus in their child because they do not want to believe their baby has a defect. It is characteristic for the mother to report that relatives, often the mother-in-law(!), or friends first remarked on the ocular deviation, although she herself had not noticed it. In other cases the onset actually may have occurred later in life than the history given by the parents indicates.

Costenbader14 noted the high prevalence of pseudostrabismus in a group of children with a primary diagnosis of strabismus (352 of 753 patients) and postulated that children at first may have pseudostrabismus that subsequently develops into an esotropia. In such cases, the history given by the parents indicates that the onset of esotropia was at birth rather than at a later date. In this group the prognosis for normalization of binocular functions may be significantly better than in patients with a true congenital esotropia since the former group of patients had an opportunity to acquire binocular single vision before the deviation occurred. If the reliability of the history obtained from the parents is doubtful, photographs

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of the patient are useful in determining whether strabismus was present at an earlier age than indicated by the parents. Careful investigation for other frequently associated features of essential infantile esotropia in most instances will allow one to arrive at the correct diagnosis, even in patients whose histories are doubtful or in whom a history cannot be obtained, such as adopted children.

**SIZE OF DEVIATION.** Unlike esotropia of later onset, essential infantile esotropia is usually characterized by a large deviation of 30° or more.\(^5^4, 5^7, 20^5\) Deviations of less than 30° also occur but are less common. The angle of deviation is usually quite stable if stability of the angle is defined as insignificant changes in its size in the course of the examination and on subsequent reexaminations. Exceptions do occur, especially in patients with smaller angles of deviation in whom spontaneous resolution of esotropia may occur\(^25\) and in those with the nystagmus blockage syndrome in whom the angle is quite variable (see Chapter 23). The concept of a stable angle in patients with essential infantile esotropia is not shared by all ophthalmologists.\(^6^9, 12^3, 16^5\) Clark and Noel\(^4^6\) and Hiles and coworkers\(^12^3\) described cases of large angle essential infantile esotropia with spontaneous regression of the angle over 3 years. Such events are rare, however, and Birch and coworkers\(^25\) reported stability of the angle in 66 children with essential infantile esotropia of 40° or more who were followed from infancy until the age of 4½ years or older.

As a rule, there is no significant difference in the angle at near and distance fixation, which indicates a normal AC/A ratio. The widely held concept that essential infantile esotropia is essentially nonaccommodative has been challenged by some authors.\(^5^0, 2^4^4, 2^4^5\) The ophthalmologist must be aware that an accommodative component may occasionally be superimposed upon the basic condition and require optical correction in case of an uncorrected hypermetropia or bifocals in case of a high AC/A ratio.

**REFRACTIVE ERRORS.** Costenbader,\(^5^4\) in a survey of 500 children with essential infantile esotropia, described the distribution of refractive errors: 5.6% myopes, 46.4% mild hyperopes (emmetropia to +2.00D sph), 41.8% moderate hyperopes (+2.25D to +5.00D sph), and 6.4% high hypermetropes (+5.25D sph, and more). It is of interest in this series of patients that the size of the deviation was unrelated to the size and type of refractive error. We have analyzed the refractive errors in 408 patients with the diagnosis of essential infantile esotropia who were treated at our clinic and found a distribution similar to that reported by Costenbader (Fig. 16–3).

The amount of hypermetropia present at the first examination may depend, of course, on the age at which the child is first seen, since numerous studies have shown that at 1 to 2 years of age

![FIGURE 16-3. Distribution of refractive errors (spherical equivalent) in 408 patients with essential infantile esotropia.](image-url)
both emmetropia and hypermetropia up to 3D can be considered as being within normal limits (see Molnar\textsuperscript{190} and many others; see also Chapter 7). The generally held view that this “physiologic” hypermetropia diminishes as the child grows older was challenged by Brown and Kronfeld.\textsuperscript{31} These authors monitored the refractive error in a group of children during the first 5 years of life and found either an increase of hypermetropia or no change. In a later study, Brown\textsuperscript{90} reported an increase of hypermetropia until the end of the seventh year.

Burian\textsuperscript{14} emphasized that in high hypermetropes (+ 4.00D sph or more) the esodeviation has a tendency to decrease with passage of time. In fact, 10% to 20% of these patients will eventually develop an esotropia. This observation justifies conservatism with respect to early surgical treatment in such patients (see also (Clark and Noel,\textsuperscript{46} Moore,\textsuperscript{191} and Stangler-Zuschrott\textsuperscript{274}).

**DUCTIONS AND VERSIONS.** Most children with essential infantile esotropia exhibit apparent defective abduction or excessive adduction or both. This is often mistaken for bilateral paresis or paralysis of the lateral rectus muscles. If amblyopia is present, the defective abduction often is more prominent in the amblyopic eye. If abduction is apparently restricted, one cannot be sure whether the child is unwilling or unable to abduct fully. Examination of ocular rotations in extreme positions of gaze is not easy in young infants. Even older children and some adults may find it difficult to move the eyes into extreme positions of levoversion or dextroversion. One reason why it may be difficult to get children with essential infantile esotropia to abduct fully when the fellow eye is conversed is manifest-latent nystagmus. Such patients habitually fixate with one eye in adduction, a position in which the nystagmus is least pronounced or even absent and visual acuity is at its best.

In our experience a true abducens paresis is very rare in early infancy. The vast majority of patients with infantile esotropia and apparently limited abduction are either unwilling to abduct fully or are unable to do so because of secondary contracture of the medial rectus muscle(s), conjunctive, or both. In the former condition the doll’s head maneuver or a few hours of occlusion of either eye will readily differentiate a true from a pseudoparesis of the lateral rectus muscle(s). In the latter, a forced duction test may become necessary to diagnose contracture. Recession of the tight medial rectus muscles in such patients will normalize the action of the seemingly deficient lateral rectus muscle.

The role of the nystagmus blockage syndrome in simulating an abducens paralysis is discussed in Chapter 23.

**AMBLYOPIA.** Amblyopia is a commonly associated factor in essential infantile esotropia. It was found in 35% of 408 patients with essential infantile esotropia treated in our clinic.\textsuperscript{205} Costenbader\textsuperscript{44} reported a prevalence of 41% in his series of 500 cases and Shauly and coworkers\textsuperscript{259} diagnosed amblyopia in 48% of their 103 patients. It is generally agreed that amblyopia, unless treated and cured early in life, is a severe obstacle to the return of normal binocular functions. Curiously, the prevalence of amblyopia in patients with essential infantile esotropia not operated on is much lower (14% to 19%).\textsuperscript{36, 97} The reason for this difference is not clear. However it is unlikely that a large angle esotropia protects a patient from amblyopia, as proposed by Good and coworkers,\textsuperscript{102} since there is no apparent relationship between amblyopia and the size of the deviation.\textsuperscript{211}

**ASSOCIATED VERTICAL DEVIATIONS.** To distinguish clearly between an elevation in adduction caused by an overacting inferior oblique and a dissociated vertical deviation may be difficult in infants. We suspect that many patients with essential infantile esotropia in whom a diagnosis of inferior oblique overaction was made in the past actually had a dissociated vertical deviation. The differential diagnosis between these conditions is discussed in Chapter 18. Attention has also been drawn to the fact that an apparent over- or under-action of an oblique muscle may be simulated by atopic muscle pulleys\textsuperscript{45} or by cyclotropia (see Chapter 18). It is for these and other reasons (see Chapter 18) that in recent years we have discouraged the indiscriminate use of the diagnostic label inferior or superior oblique overaction in patients with elevation or depression of the adducting eye and prefer the generic terms of elevation or “upshoot” in adduction and depression or “downshoot” in adduction instead.

Elevation or depression in adduction, often associated with a V or A pattern, and dissociated vertical or horizontal deviations are common components of the essential infantile esotropia syndrome. We found elevation in adduction in 68% of 408 cases.\textsuperscript{205} The point is often made that up-
Esodeviations

and downshoot in adduction and dissociated deviations are infrequently found in children with essential infantile esotropia who are under 1 year of age, and do not emerge until the horizontal deviation has been surgically corrected. This has certainly also been our impression. However, it must also be considered that because of the difficulties encountered in performing a complete motility analysis in infants these conditions are already present before surgical correction of the esotropia but are not diagnosed because of the masking effect of a large horizontal deviation. Campos observed that dissociated vertical deviations present before surgery may actually resolve after early horizontal alignment with chemodenervation.

The age at surgical correction of the esotropia is unrelated to the occurrence of dissociated vertical deviations. In fact, this condition occurred also in 60% of 113 patients with essential infantile esotropia who remained untreated until visual adulthood.

The prevalence of dissociated vertical deviation, which often has a horizontal component (see Chapter 18), in patients with essential infantile esotropia is high. We have diagnosed this condition in 51% of 408 patients with essential infantile esotropia. Other authors have reported an even higher prevalence, for example, Lang (90%), Parks (76%), Helveston (70% to 90%), and Calcutt and Murray (60%). A possible reason for these differences in the reported prevalence of dissociated vertical deviations is that some authors restrict the diagnosis to the presence of a manifest dissociated deviation whereas others include cases with a latent dissociated deviation, which can only be elicited by covering one eye.

Parks believes that a dissociated vertical deviation is indirect evidence for the onset of esotropia at the time of birth. We do not agree with this view since a dissociated vertical deviation is an entity sui generis. Although it occurs commonly in association with essential infantile esotropia, this form of deviation also may accompany acquired esotropia or exotropia (see Chapter 18). Even patients in whom no other form of strabismus is present may have this type of anomaly. Evidence is lacking to support the belief that the presence of this anomaly is proof of the congenital nature of an associated horizontal strabismus.

Lang commented on the common occurrence (65%) of excyclotropia of the nonfixating eye in his patients with infantile strabismus. Rather than considering excyclotropia of the nonfixating eye as an isolated associated anomaly in the essential infantile esotropia syndrome, we prefer to regard it as a component of the dissociated vertical deviation syndrome (see Chapter 18).

**NYSTAGMUS.** The clinical characteristics of the various congenital nystagmus forms, including the differentiation between manifest-latent and manifest nystagmus, are discussed in Chapter 23. We will consider nystagmus in this section only as it pertains to essential infantile esotropia.

**LATENT OR MANIFEST-LATENT NYSTAGMUS.** These types of congenital nystagmus occur commonly in essential infantile esotropia and must be distinguished from manifest congenital nystagmus, a less commonly associated oculomotor anomaly. Latent nystagmus is characterized by a nasally directed drift of the nonfixating eye, followed by a fast corrective saccade in the temporal direction. Upon changing fixation to the fellow eye the direction of the nystagmus reverses. True latent nystagmus that is present only with one eye occluded is rare and in most patients a manifest nystagmus, albeit of lesser amplitude, is present with both eyes open; hence the somewhat awkward term manifest-latent nystagmus.

We have suggested that infantile esotropia, when associated with manifest-latent nystagmus, may well represent a special subgroup within the essential infantile esotropia syndrome. In this connection a recent study is of interest according to which nystagmus when associated with infantile esotropia may increase the risk of requiring additional operations for overcorrection of residual deviation.

Since differentiation between manifest-latent and manifest nystagmus is often not possible on clinical grounds alone and reliable electronystagmographic recordings are difficult to obtain in small children, both conditions are easily confused. This may explain the large differences in the prevalence of latent nystagmus reported by different authors. For instance, Ciancia observed latent nystagmus that increased in abduction and decreased in adduction in 33% of patients with essential infantile esotropia, whereas Lang made this diagnosis in 43% (52%) of 82 patients. Electronystagmographic studies in a small number of patients have even indicated a prevalence of 95%. We have diagnosed nystagmus without the benefit of nystagmographic recordings in only 25% of 408 patients with essential infantile esotropia and
were able to recognize the latent variety in only 10%.\textsuperscript{205} As pointed out by Ciancia,\textsuperscript{42} many of these patients have an abnormal head posture and turn their face in the direction of the fixating eye. The nystagmus is less pronounced or even absent in adduction, and improvement of visual acuity in this position explains the anomalous head posture (see Chapter 23). Spielmann and Spielmann\textsuperscript{269} emphasized that this condition is not to be confused with the blocking of manifest congenital nystagmus by convergence.

Earlier studies had led some investigators to propose that a disturbance of coordination between vestibular and optic control of the oculomotor system may be of etiologic significance in causing latent nystagmus and dissociated vertical deviation.\textsuperscript{80, 81, 165} Doden and Adams\textsuperscript{81} described involuntary rhythmic, conjugate, pendular deviations of the eyes on vestibular testing in 23% of 150 strabismic subjects. They interpreted these anomalies as expressions of a central disturbance of coordination, possibly caused by lesions of the brain stem involving the vestibular nuclei and the substantia reticularis. Hoyt\textsuperscript{131} reported abnormalities of the vestibulo-ocular response in infantile esotropes without nystagmus. For more recent views of a causal relationship between manifest-latent nystagmus and optokinetic asymmetry (Kommerell\textsuperscript{155}) and a hypothesis linking latent nystagmus etiologically with infantile esotropia (Lang\textsuperscript{170}), see Chapter 9.

**MANIFEST NYSTAGMUS.** Most authors report that manifest nystagmus occurs less commonly with essential infantile esotropia than do latent and manifest-latent nystagmus.\textsuperscript{78, 205, 218} However, it is commonly encountered when essential infantile esotropia is associated with other conditions, such as Down syndrome, ocular albinism, cerebral palsy, and hydrocephalus. Under certain conditions patients learn to dampen the nystagmus by convergence, and this sustained convergence may cause a secondary esotropia (nystagmus blocking syndrome), which is discussed in Chapter 23. In this context it must be underscored that only the mechanism of this esotropia is distinctly different from essentially essential infantile esotropia. Other patients may dampen the nystagmus in a lateral gaze position.

**ASYMMETRICAL OPTOKINETIC NYSTAGMUS.** A common association between optokinetic asymmetry and essential infantile esotropia has been established.\textsuperscript{28, 79, 92, 154, 186, 289, 290} In normal subjects the optokinetic response elicited by a rotating nystagmus drum (Fig. 16–4) consists of a smooth pursuit movement in the direction of the moving stripes or pictures, followed by a corrective saccade in the opposite direction. This pursuit movement occurs with equal facility, regardless of whether the stripes move from a nasal to a temporal or from a temporal to a nasal direction.

However, in many esotropic patients this symmetry is grossly disturbed (Fig. 16–5) and pursuit movements are irregular or are difficult to elicit when the drum moves in a nasotemporal direction (optokinetic asymmetry). This phenomenon has been interpreted as a defect in visual motion processing: the visual cortex fails to acquire the ability to transmit temporally directed object motion to the nucleus of the optic tract (NOT), although not affecting object motion from the retina to the cortex.\textsuperscript{124, 154} However, this asymmetry is not a pathognomonic feature of essential infantile esotropia but occurs also in normal, visually immature infants.\textsuperscript{9, 125} in nonstrabismic patients with deficient binocular input because of anisometropia, and in various other forms of monocular visual deprivation early in life,\textsuperscript{105, 125, 178} after enucleation,\textsuperscript{246} and in the Duane type I syndrome.\textsuperscript{105} Its presence is merely evidence for disruption of binocular vision during visual immaturity before the age of 3 to 4 months, regardless of its cause. Normal and equal binocular visual input is required during infancy for maturation of the optokinetic reflex and the state of immaturity (asymmetry) persists in the absence of such input. Thus, optokinetic asymmetry must be considered a con-

![FIGURE 16-4. Use of a pediatric nystagmus drum to elicit optokinetic nystagmus in children. (Courtesy of Dr. D. Coats, Houston, Texas.)](image)
sequence of essential infantile esotropia rather than, as has been suggested, the manifestation of a primary motion-processing defect in the visual pathway. In support of a primary motion-processing defect in these patients it has been reported that their nonstrabismic first-degree relatives demonstrate significant motion-processing asymmetries. However, this finding could not be confirmed in a study performed by us.

Westfall and coworkers confirmed the existence of asymmetrical optokinetic nystagmus in patients with essential infantile esotropia. In some of these patients sensory fusion could be detected by means of dynamic random-dots visual evoked response (VER). These authors argued therefore that optokinetic asymmetry is not associated with a deficit in the cortical fusion facility, but rather with deficits in binocular pathways projecting to monocular optokinetic nystagmus centers. These deficits may be associated with abnormal processing subsequent to sensory fusion or with abnormal processing in motion pathways, which run parallel to sensory fusion pathways.

Optokinetic asymmetry is, with certain limitations, a useful clinical sign to date the onset of strabismus since it occurs more commonly in children with an onset before the age of 6 months than in those with a later onset (Fig. 16–6). Care must be taken, however, in interpreting optokinetic responses obtained with a nystagmus drum in children since the asymmetry may be subtle and not recognizable unless nystagmography can be performed.

As mentioned in Chapter 9 the common association between optokinetic asymmetry and latent nystagmus in essential infantile esotropia has led to speculations regarding a causal relationship. The reportedly high correlation between the severity of pursuit asymmetry and the intensity of latent nystagmus seems to be in accord with this hypothesis. In further support of it we would also expect an invariable linkage between optokinetic asymmetry and latent nystagmus. However, latent nystagmus is not consistently associated with asymmetry of the optokinetic response or with motion detection deficits.

**MOTION-PROCESSING DEFICITS.** The naso-temporal motion defect is not limited to the pursuit system and a similar response bias exists for motion processing, as shown by monocularly recorded VEPs (visual evoked potentials). As with optokinetic asymmetry this bias is a normal feature in infants but persists into adulthood in

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**FIGURE 16–5.** Binocular nystagmogram during optokinetic stimulation from nasal to temporal (upper tracings) and temporal to nasal (lower tracings) in a normal subject (left) and a patient with essential infantile esotropia (right). Note poor optokinetic response when the drum is moved in a naso-temporal direction in the esotropic patient. (From Noorden GK von: Current concepts of essential infantile esotropia (Bowman Lecture). Eye 2:343, 1988.)
FIGURE 16–6. Histogram showing the prevalence of clinical monocular optokinetic nystagmus (OKN) asymmetry in esotropic patients with various ages of onset of strabismus. The asterisks indicate statistically significant differences in prevalence between adjacent columns, and the error bars indicate 95% confidence intervals for mean prevalence. There was a significantly greater prevalence of asymmetry in esotropic patients with onset before the age of 6 months than in those with onset between 6 and 12 months, and these, in turn, had a significantly greater prevalence than those with later onset. (From Demer JL, Noorden GK von: Optokinetic asymmetry in esotropia. J Pediatr Ophthalmol Strabismus 25:286, 1988.)

infantile esotropia. The results of studies on motion perception (reviewed by Fawcett and coworkers89) are contradictory with regard to the presence of a deficit and the direction of monocular asymmetries in infantile esotropia. Kommerell and co-workers157 questioned whether optokinetic asymmetry and motion asymmetries are caused by the same central defect as they found no significant correlation between these disturbances. Fawcett and coworkers89 compared motion perception in stereoblind infantile esotropes and patients with acquired esotropia and normal stereopsis and found similar anomalies in both groups. These authors concluded that interruption of binocularity cannot be the underlying cause of abnormal motion processing in essential infantile esotropia.

ANOMALOUS HEAD POSTURE. Lang165 reported an anomalous head posture in 57 (70%) of 82 patients with essential infantile esotropia, and others have commented on the high rate of occurrence of this association.42 The head and face are said to be tilted toward the shoulder of the fixating eye.167 We cannot confirm this high rate of occurrence, which we have observed in only 6% of 408 patients with essential infantile esotropia.205 De Decker and Dannheim-de Decker73 observed a conspicuous anomalous head posture in only 2% of their patients and noted a head tilt toward the side of the fixating eye in most instances. However, patients with a head tilt toward the side of the nondominant eye often had a dissociated vertical deviation with strong unilateral preponderance. In some patients the anomalous head posture is associated with latent or manifest-latent nystagmus and the patient turns the head toward the side of the fixating eye (Ciancia syndrome). However, as pointed out by de Decker71 and in our experience as well, this correlation is by no means consistent.

Crone61 has stated that the torticollis compensates for an incyclotropia of the fixating eye, a view shared by other authors.4, 266 However, an incycloduction of the fixating eye is not a consistent feature of dissociated vertical deviations and one also wonders why a head tilt is not more common considering that dissociated vertical deviations occur at least in one half of all patients with essential infantile esotropia.205

The prevalence of various components of the essential infantile esotropia syndrome encountered in our patient population is summarized in Table 16–3.
Therapy

It is a tragic fallacy for parents of strabismic children to be told by their family physician that the problem will need no medical attention until the child reaches preschool age or that in time the eyes will straighten out spontaneously. The truth is that strabismus neglected in early childhood may cause severe, irreversible sensory anomalies and that secondary changes in the extraocular muscles, conjunctiva, and Tenon’s capsule that develop as a result of long-standing strabismus will make the results of surgical correction at a later age less predictable. It also behooves the primary care physician to remember that strabismus may develop secondary to reduced visual acuity and may well be the second most common sign of retinoblastoma.86

We insist therefore that every child whose eyes are not aligned by 3 months of age be given a complete ophthalmologic examination. Although treatment may not be possible or even necessary on the first visit, a baseline of clinical information can be established that will be helpful in the future management of the patient.

The treatment of choice for essential infantile esotropia is surgical alignment of the eyes. Non-surgical treatment is directed toward correction of a significant refractive error, elimination of amblyopia during the preoperative phase, and treatment of a residual angle of esotropia during the postoperative period.

**GOALS OF TREATMENT.** A cure of strabismus may be defined as a restoration of single binocular vision in the practical field of gaze, that is, orthotropia or asymptomatic heterophoria; normal visual acuity in each eye; normal stereoacuity on random-dot testing; normal retinal correspondence; and stable sensory (bifoveal) and motor fusion. The isolated case of Parks230 in which binfixation was restored, and the cases of Wright and coworkers303 in which normal random-dot stereopsis was achieved notwithstanding, there is universal agreement among strabismologists that complete restoration of normal binocular vision with normal random-dot stereopsis is unattainable in essential infantile esotropia except in the rarest of circumstances. However, this conclusion need not lead to capitulation before a seemingly incurable anomaly in which surgery produces at best an improved cosmetic appearance. Stereopsis, while essential for a certain, though limited number of occupations, is not indispensable in the presence of monocular clues for depth perception. Moreover, stable sensory and motor fusion may occur in the absence of stereopsis.

A cure is not absolute, but consists of several grades of subnormal or abnormal binocular vision. We must ask: What is the nature of these less than ideal forms of binocular vision? Of what functional benefit are they for the patient? How often do they occur in a group of surgically treated patients with essential infantile esotropia? Does their occurrence depend on the age at which the eyes are surgically aligned?

**NONSURGICAL TREATMENT.** We stated in the beginning of this chapter that hypermetropic refractive errors not exceeding 2D to 3D are physiologic variants in infants. The question arises with respect to essential infantile esotropia whether correction of a small hypermetropic refractive error is indicated. Essential infantile esotropia generally is nonaccommodative; that is, the AC/A ratio is usually normal, high hypermetropic refractive errors are rare (see Table 16–3), and little if any difference exists between the angle of deviation measured at distance and near fixation. However, as mentioned earlier, exceptions do occur, and refractive and nonrefractive accommodative esotropia can have their onset in early infancy.

We have observed on occasion a high AC/A ratio in patients with essential infantile esotropia without a refractive error. In such cases, the esotropia increases significantly at near fixation and little if any deviation is present at distance fixation. Before this diagnosis can be established in infants, one must, of course, rule out interference with steady fixation at distance through lack of attention during the measurement.

**TABLE 16–3.** Prevalence of Characteristics of Essential Infantile Esotropia* (N = 408)

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>n</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amblyopia</td>
<td>144</td>
<td>35</td>
</tr>
<tr>
<td>Anomalous head posture</td>
<td>26</td>
<td>6</td>
</tr>
<tr>
<td>Dissociated vertical deviation (DVD)</td>
<td>208</td>
<td>51</td>
</tr>
<tr>
<td>Overaction of inferior obliques (OAIO)</td>
<td>277</td>
<td>68</td>
</tr>
<tr>
<td>DVD and OAIO (combined)</td>
<td>171</td>
<td>42</td>
</tr>
<tr>
<td>Manifest nystagmus</td>
<td>62</td>
<td>15</td>
</tr>
<tr>
<td>Manifest-latent nystagmus</td>
<td>41</td>
<td>10</td>
</tr>
</tbody>
</table>

*Mean deviation at distance, 44° (range, 5°–100°); mean deviation at near, 49° (range, 10°–95°).

It has become our policy to correct all hypermetropic refractive errors in excess of +2.50D sph before considering surgery. In uncooperative infants, a trial of miotics may be considered in lieu of glasses. In most patients with essential infantile esotropia, however, correction of the “physiologic” low-degree hypermetropia has little, if any, effect on the deviation.

The view that essential infantile esotropia in most instances is nonaccommodative was challenged by Réthy and Gál.245 Réthy,244 and Kettesy.150 These authors rejected the concept of congenital nonaccommodative esotropia and claimed that in a high percentage of cases the deviation is accommodative in origin. Réthy noted that after correction of the full hypermetropic refractive error in esotropic children and repetition of cycloplegic refraction a month or two later, retinoscopy revealed a higher refractive error than was found on the first examination. After increasing the prescription and overcorrecting the hypermetropia by 0.5D to 1.0D, he noted reduction of the angle of deviation. Such patients were atropinized to facilitate acceptance of the overcorrection. When the procedure was repeated several times, latent hypermetropia became increasingly manifest and was corrected or overcorrected until the accommodative effort was decreased to the point where the associated accommodative convergence no longer caused esotropia. Réthy claimed that unless this therapy is instituted in early infancy, the increased accommodative tonus and the associated increased accommodative convergence will stabilize and become refractory to belated therapeutic measures. According to this author, such cases are then erroneously referred to by strabismologists as “nonaccommodative.” Réthy claimed that surgery can be avoided in 90% [sic] of esotropic patients if his method of treatment is followed.244

We are in full agreement with Réthy and Kettesy concerning the validity of Donders’ theory whenever it is applicable. We also stress the often neglected need for full correction of a hypermetropic refractive error and for frequent refractions in patients with accommodative esotropia, in view of the findings of Brown and Kronfeld35 and those of Brown.30 Furthermore, the effectiveness of atropinization in causing complete cycloplegia varies in different patients, and latent hypermetropia may initially go undetected66 and become only gradually manifest as corrective lenses are worn for some time. On the other hand, the angle of deviation in essential infantile esotropia as a rule is not consistent with the excess of accommodation required to overcome hypermetropia. In most instances there is no relationship between the angle of strabismus and the size or type of refractive error in essential infantile esotropia.54 The deviation usually is considerably greater than it would be if the excess convergence were related to the increased accommodative effort. Moreover, a prospective study by Ingram and coworkers140 has shown that a prophylactic correction of hypermetropic refractive errors in excess of 2.00D did not prevent development of strabismus. Thus the extension of Donders’ doctrine by Réthy and Kettesy to include the vast majority of patients with essential infantile esotropia is not justified in our opinion, which should not detract from the fact that the theory of Donders remains the best substantiated explanation for accommodative strabismus (see Chapter 9).

Amblyopia should be treated rigorously before and not, as advocated by some authors,162, 294 after surgery for the following reasons: (1) The earlier in life the treatment is begun, the shorter the duration of treatment. (2) The diagnosis of amblyopia and the monitoring of the fixation preference during treatment are more difficult once the eyes are aligned or nearly aligned by surgery than in the presence of a large angle esodeviation.37, 207 (3) Once the eyes are aligned some parents may be lulled into thinking that all problems are over and become negligent in keeping their follow-up appointments. We have repeatedly seen children with deep amblyopia who had early surgery and who, in spite of our instructions, did not return to our office until years later. (4) The outcome of surgery is less favorable in patients who remain amblyopic at the time of surgery.148, 258, 259 The time for surgery has come when the child alternates freely or can hold fixation with the formerly amblyopic eye through a blink.

Some investigators have recommended the use of prisms in the preoperative treatment of essential infantile esotropia.77, 292 However, this treatment has never become popular and is not used by us. Prismatic therapy and the prism adaptation test are discussed in Chapter 24.

**SURGICAL TREATMENT**

**TIMING OF SURGERY AND RESULTS.** Much discussion has centered on the optimal time at which to operate on children with essential infantile esotropia. Several schools of thought have
evolved, some advocating surgery as early as 3 months and others as late as 4 years of age. Fifty years ago an operation at 4 or 5 years of age was considered early and in most instances surgery was not contemplated until the child was ready for school. There has been a general tendency among ophthalmologists to operate on children younger than was customary then. Improved safety of anesthetic procedures has reduced the surgical risk to an almost negligible minimum. A steadily growing group of surgeons now believe that an operation for essential infantile esotropia is advisable before completion of the first 24 months of life and some prefer to complete surgery during the first 12 months or even earlier. The arguments for this reasoning are that early surgical treatment provides a better chance for functional improvement, is desirable for psychological reasons, and that secondary changes occur in the extraocular muscles, the conjunctiva, and Tenon’s capsule—all of which make a correction at a later date more difficult and less predictable.

Early surgery was pioneered by the late Frank Costenbader who in 1958 stated: I feel strongly that we should, first, regain and maintain vision in each eye from early childhood, and, second, regain binocular alignment as early in infancy and childhood as possible and to maintain it thereafter.53, p. 331

Costenbader’s plea for early surgical alignment of the eyes was undoubtedly influenced by Chavasse, who, in turn, was influenced by the Pavlovian thinking of his time. Chavasse stated that the opportunity for early single binocular vision is of paramount importance for development of normal binocular reflexes. Sporadic reports in the literature of normal or near-normal random-dot stereopsis after surgical alignment before 6 months of age seemed to support this view.53, 138, 303

Other eye surgeons advocated operating when the child is about 2 years of age or even older. Many clinicians believe that examination during early infancy cannot be sufficiently complete for careful surgical planning; that associated vertical anomalies, including the A and V patterns, overaction of the inferior oblique muscles, or dissociated vertical deviations may be overlooked; and that the deviation at distance fixation cannot be evaluated reliably before 2 years of age.

The older literature is replete with a pessimistic outlook regarding the functional outcome following surgery in patients with infantile strabismus that is based on Worth’s assumption that a congenital defect of the fusion faculty is the cause of squint. Indeed, the view was common that normal binocular functions are obtained rarely, if ever, when the deviation dates from birth. The beneficial effect of surgical alignment of the eyes by the age of 24 months was first discussed in the frequently quoted paper by Ing. Other studies came to the same conclusion. The major problem in evaluating much of this work involves the interpretation of tests used to determine the presence of binocularity. We find it erroneous to assume, as is frequently done, that gross stereopsis, a positive Worth four-dot test, or visibility of the two stripes during the Bagolini striated glasses test are indicators of fusion when in fact any or all of these responses can be elicited when a manifest residual esodeviation is present along with anomalous retinal correspondence. The binocular cooperation on the basis of anomalous retinal correspondence between the fovea of the fixating eye and a peripheral retinal area in the eye with a residual eso- or exodeviation is functionally not equivalent to stable normal binocular vision at all fixation distances with fusional amplitudes! Unless a clear distinction is made between normal and anomalous binocular vision in evaluating the results of patients operated on and aligned at different ages, no conclusions regarding the therapeutic superiority of operating at an early age can be drawn.

A second problem with many studies on the results of surgery in essential infantile esotropia is that contemporary authors, too numerous to cite here, consider a residual deviation of 10° a satisfactory surgical outcome. Must it be emphasized that orthotropia, eso- and exophoria, intermittent heterophoria, and eso- and exotropias of 10° are functionally not the same? If no distinction is made on the basis of the cover and the cover-uncover tests that this residual deviation is a heterophoria or a heterotropia at all fixation distances, the criterion of 10° for surgical success is misleading and therefore useless. At best, it tells whether the patient has been cosmetically improved by the surgery.

A third problem with many of the older studies is that no distinction is made between the age at the first operation and the age at which alignment was accomplished. Clearly, only the latter is relevant to this discussion.
In an effort to improve communication between clinical investigators and with the shortcomings, mentioned above, of previously used outcome criteria in mind, we suggested the following classification of the results of surgery in essential infantile esotropia: (1) subnormal binocular vision, (2) microtropia, (3) small angle esotropia or exotropia, and (4) large angle esotropia or exotropia. Some of the clinical features of these conditions are summarized in Table 16–4, and the tests employed for their diagnosis have been discussed in Chapter 12 (see also von Noorden). Table 16–4 has been modified from previous editions in response to questions raised regarding its clarity.

We consider subnormal binocular vision, a term introduced by Lyle and Foley and also used by de Decker and Haase, as an optimal treatment outcome and have never seen a result better than that in infantile esotropia. Unlike in microtropia, the patient fixates centrally, has normal visual acuity in each eye, and behaves in all other respects like someone with normal binocular vision, except for reduced stereopsis and a foveal suppression scotoma in one eye that is only present under binocular conditions. As will be discussed later in this chapter, microtropes are often mildly amblyopic with parafoveolar fixation and have anomalous retinal correspondence with identity of the angle of anomaly and the degree of fixation eccentricity. Microtropes with foveolar fixation in each eye may be difficult to distinguish from patients with subnormal binocular vision and we readily admit that this is a transitional zone in the classification of outcomes presented here. Clearly, however, a microtropia because of the amblyopia is less advantageous from a functional point of view than is subnormal binocular vision and must therefore be considered a less than optimal but acceptable outcome.

Subnormal binocular vision and microtropia are often thought of as favoring motor stability, that is, being protective against a recurrence of strabismus. However, several studies have shown that whereas the stability of alignment is significantly better when these conditions are present as opposed to when they are not, deterioration does occur. Residual small angle eso- or exotropias of less than 20 do not interfere with the normal appearance in most patients and in this case require no further treatment, except for a still existing or recurrent amblyopia. As in microtropia we speak of a less than optimal but still acceptable outcome when a residual but small deviation remains. Residual esotropia or consecutive exotropia requiring surgery is clearly an unacceptable outcome.

Using these criteria we have analyzed results of surgical attempts to align the eyes to an orthotropic position as closely as possible by one or multiple operations in 358 patients with a documented onset of esotropia before the age of 6 months. Figure 16–7 lists the prevalence of the various functional endstages of therapy according to whether surgical treatment was completed between 4 months and 2 years, 2 to 4 years, or older than 4 years after a mean follow-up of 39 months. This analysis shows that as age at completion of surgical therapy increases, the probability of an optimal outcome (subnormal binocular vision) decreases. Increasing age at the completion of treat-

<table>
<thead>
<tr>
<th>Subnormal Binocular Vision</th>
<th>Microtropia</th>
<th>Small Angle ET/XT (&lt;20°)</th>
<th>Large Angle ET/XT (&gt;20°)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Orthotropia or asymptomatic heterophoria</td>
<td>Inconspicuous shift or no shift on cover test</td>
<td>Appearance improved; most parents are happy with outcome</td>
<td>Conspicuous residual angle</td>
</tr>
<tr>
<td>Normal VA in each eye</td>
<td>Mild amblyopia is common</td>
<td></td>
<td></td>
</tr>
<tr>
<td>NRC</td>
<td>Usually ARC</td>
<td>ARC is common</td>
<td>ARC or suppression</td>
</tr>
<tr>
<td>Fusion with amplitudes</td>
<td>Anomalous fusion on the basis of ARC</td>
<td></td>
<td>No fusion</td>
</tr>
<tr>
<td>Reduced stereopsis</td>
<td>Stereopsis reduced or absent</td>
<td></td>
<td>Absence of stereopsis</td>
</tr>
<tr>
<td>Stable alignment</td>
<td>Some stability</td>
<td>Less stability</td>
<td>May be unstable</td>
</tr>
<tr>
<td>No further treatment</td>
<td>No treatment except for amblyopia</td>
<td></td>
<td>Additional surgery may be required</td>
</tr>
<tr>
<td>Optimal outcome</td>
<td>Less than optimal but still acceptable</td>
<td></td>
<td>Unacceptable outcome</td>
</tr>
</tbody>
</table>

ARC, abnormal retinal correspondence; ET, esotropia; NRC, normal retinal correspondence; VA, visual acuity; XT, exotropia.
ment tends to move the patient away from subnormal binocular vision and into the functionally inferior microtropia and small angle esotropia or exotropia group. These data, although based on evaluation criteria quite different from those used by previous investigators, are in support of the current view that surgery completed before the age of 2 years yields superior results. They have been confirmed by Shauly and coworkers who have adopted our classification of surgical results. However, they are at variance with the admonition that unless surgery is completed before that time, a functionally useful form of binocular vision cannot be expected. In fact, many of our patients achieved such results when surgical treatment was concluded after the age of 2 or even 4 years. De Decker has confirmed these findings.

Although these results are far from perfect, they are not nearly as dismal as one must conclude from the older literature, especially since we have learned to consider anomalous correspondence in a more favorable light (see Chapter 13). Some form of binocular cooperation ranging from near-normal (subnormal binocular vision) to anomalous (microtropia and small angle esotropia or exotropia) existed in 66% of our patients. To the mother who asks, “Is this operation done for cosmetic reasons or will my child ever learn to use the eyes together?” we can answer, “There is an above-average chance for some form of binocular cooperation after surgery, although normal depth perception cannot be expected.”

Until such time that new information becomes available that may require modification of this approach we advocate surgery when the following criteria are met: (1) demonstration of a stable and sufficiently large deviation, (2) absence of an accommodative factor, (3) alternating fixation behavior after treatment of amblyopia, and (4) identification of the nature of associated vertical deviations or of vertical incomitance (A or V patterns). As soon as this information is unequivocally available, we see no reason to delay surgery. In addition to functional and psychological benefits (see Chapter 10) there is some evidence that the child’s motor development improves after surgery. Although controlled studies are needed to validate this point, it is certainly astonishing, and perhaps more than just a coincidence, how often parents will report spontaneously during postoperative visits that the child stumbles and runs into walls less often. In this connection it may be of interest that expansion of the binocular field of vision has been reported in adults after surgery for esotropia.

The completeness with which all necessary preoperative information can be obtained depends largely on the cooperation of the child as well as the patience and understanding of the examiner. In some patients, these data can be obtained by 6 months of age or earlier, whereas in others the

surgeon may be forced to procrastinate until the child is 2 years of age or older.

Of special significance with regard to outcome expectations is the fact that normal stereoscopic vision remains such an elusive therapeutic goal in the overwhelming majority of patients with essential infantile esotropia. In view of the extraordinary sensitivity to conflicting visual input of cortical binocular neurons mediating stereopsis in infant kittens\textsuperscript{132} or infant monkeys,\textsuperscript{58–60} the question arose whether surgery even at the age of 6 months may be too late to preserve stereopsis. Several recent studies have addressed this point and the results are controversial. Birch and coworkers\textsuperscript{25} reported better random-dot stereopsis in children operated upon between the ages of 5 and 12 months than in another group who had surgery at 13 to 18 months. Wright and coworkers\textsuperscript{103} operated on seven patients between the ages of 13 and 19 weeks and achieved normal stereopsis (Titmus test) in two cases. However, Ing\textsuperscript{138} examined 16 patients who were surgically aligned by other ophthalmologists at a mean age of 4.2 months but was able to identify only one patient with refined stereoscopic discrimination of 40 seconds of arc (Titmus test) and 20 seconds of arc (Randot test). Likewise, Helveston and coworkers\textsuperscript{122} who operated on 10 patients aged 4 months found stereopsis of 140 seconds (Titmus test) in only one instance.

We do not believe that the evidence thus far presented in favor of operating before the age of 6 months is sufficient to advocate this approach on a general basis. Long-term follow up studies are essential to improve our knowledge as to the functional benefits to be derived from operating that early in life. The longitudinal study of patients thus treated by Helveston and coworkers\textsuperscript{122} showed subsequent deterioration after initial ocular alignment in many instances.

The observations that normal or near-normal stereopsis can be restored at all in isolated instances after surgical alignment before the age of 6 months is of interest with regard to the etiology of infantile esotropia. Stereopsis has been demonstrated by objective means, at least transiently, in esotropic infants corrected with prisms\textsuperscript{189} or immediately after surgical alignment and before a residual strabismus established itself.\textsuperscript{5, 240} On the other hand, we have learned that artificial strabismus produced in infant monkeys for as briefly as 1 week decimates the number of striate neurons that normally receive input from both eyes\textsuperscript{12, 58} and that are linked to stereopsis.\textsuperscript{15, 59, 60} This loss of binocular neurons is irreversible in monkeys even after realignment of the eyes and subsequent exposure to a normal visual environment for as long as 2 years. In view of the great functional and anatomical similarity of the visual system in monkeys and humans it is reasonable to conclude that defective stereopsis in infantile esotropes, similar to optokinetic asymmetry, is an irreversible consequence of strabismus early in infancy rather than, as has been stated, the manifestation of a genetic sensory defect\textsuperscript{232} in the afferent visual pathway that precludes complete functional recovery.

Presuming that the sensory substrate before onset of the esotropia was normal, we must ask why only such a small number of patients recover normal or near-normal stereopsis in spite of very early surgery? Among the possibilities to be considered are that surgery as early as 3 to 4 months was already too late and that the destruction of cortical binocularity had already occurred before alignment was accomplished. The minimal duration of incongruous visual stimulation necessary to permanently impair cortical binocular cells is unknown in humans, and interindividual differences in susceptibility may well exist. Second, the degree of functional recovery may depend on the duration of a brief period of normal binocular visual input or of intermittency of the deviation prior to the onset of constant esotropia. We know that essential infantile esotropia has its onset usually during the first 3 months of life and is rarely, if ever, present at birth (see p. 321). Thus, a brief period or, in the case of early intermittency, period of normal binocular stimulation may well have been present prior to the onset of esotropia and stabilized binocular connections to the point where they may be recoverable after alignment. Third, a residual angle of esotropia after surgery may preclude normal stereopsis.

In addition to defective stereopsis, a second and perhaps etiologically related residual sensory defect persists even in patients with optimal or desirable surgical outcome. This consists of a foveal suppression scotoma in one eye of a patient with subnormal binocular vision and microstrabismus. This scotoma, to which Parks\textsuperscript{226} and von Noorden and coworkers\textsuperscript{216} drew attention, measures 2\textdegree{} or less in diameter and may be present only under binocular conditions of seeing. It usually occurs in the nondominant eye but may switch rapidly from one eye to the other (alternating
foveal suppression). The scotoma is diagnosed with the 4° base-out prism test (see p. 218) or by a decrease of visual acuity in one eye when acuity is determined under binocular conditions of seeing with a polarized projected chart. Thus postoperative alignment in patients with essential infantile esotropia is maintained by peripheral fusion alone and is not dependent on normal bifoveal interaction. Few will disagree that this state of binocular cooperation, although not perfect, is of functional benefit to the patient.

Although an etiologic and pathophysiologic relationship may exist between a binocular foveal suppression scotoma and reduced stereoaucity, to use the degree of stereoaucity as an indicator for suppression of bifoveal fusion, as proposed by Parks, is not justified. Normal stereoaucity (15 to 60 seconds of arc on any of the random-dot stereograms) is indisputably the hallmark of normal binocular function. On the other hand, subnormal stereoaucity or even stereoblindness can be present in orthotropic subjects with stable fusion. The mere reduction of stereoaucity alone is insufficient proof of foveal suppression.

In summarizing the ongoing discussion as to the optimal age at which to operate, it is fair to state this has yet to be established. Normal stereopsis has been restored only in isolated cases after surgical alignment prior to the sixth month of life. There is evidence from many independent studies that surgery completed before the second year of life improves the chances for recovery of limited binocularity. However, it has also been shown that such recovery may occur if surgery is delayed up to or beyond the age of 4 years. Strabismologists are awaiting eagerly the outcomes of two ongoing prospective multicenter trials in the United States and in Europe that address this important issue.

**TYPE OF OPERATION.** Our surgical approach to treatment of essential infantile esotropia has undergone periodic changes over the years. Initially, we favored a recession-resection operation on the nondominant eye, combined with inferior oblique myectomies, if indicated, to be followed, if necessary, by a recession-resection on the fellow eye. The amount of surgery varied according to the size of the deviation and on the basis of observations made during examination of the ductions of the eyes and ranged from 3- to 5-mm recessions and 5- to 8-mm resections. However, the number of reoperations required to gain or maintain alignment was discouraging. In 1972 we reported that an average of 2.1 operations per patient was required to align the eyes. Ing and coworkers, who used 3- to 5-mm recessions of both medial recti, required as many as 2.6 operations per patient to achieve this goal. In recent years we have changed our method and now employ recessions of the medial recti ranging from 5 to 8 mm, provided the deviation measures 30° or more at near fixation. This more aggressive approach has decreased the need for additional surgery and, contrary to our initial concern, does not cause limitation of adduction. Initial reports show that by doing these unconventionally large recessions of both medial recti, 73% to 84% of the patients are successfully aligned with one operation. In the presence of a significantly large residual deviation, we resect both lateral rectus muscles in a second procedure. It has been advocated that the conventional amount of surgery in myopia be increased because of a higher percentage of unacceptable undercorrections.

At this time we use a unilateral recession-resection operation on the nondominant eye frequently to treat essential infantile esotropia and only in patients who have failed to respond to amblyopia treatment.

The use of posterior fixation sutures in lieu of large bimedial recessions of the medial rectus muscles in the treatment of essential infantile esotropia is enjoying greater popularity in Europe, especially in Germany, than elsewhere. This approach is preferred by some because it is said to reduce the prevalence of consecutive exotropia. Seventy-five percent of patients thus operated on achieve an alignment between 2° exodeviation and 10° esodeviation, which is comparable to what can be accomplished with large recession of both medial rectus muscles (see above).

**POSTOPERATIVE TREATMENT**

Postoperative care should be concerned primarily with the prevention of strabismic amblyopia (see Chapter 24) and correction of a hypermetropic refractive error. Since most postoperative patients have a small angle esotropia and a deep-seated anomalous retinal correspondence, subjective complaints about diplopia or other types of visual discomfort are practically never encountered. Although from time to time the suggestion has been made, with varying degrees of enthusi-
asm, that such patients should be treated orthoptically, we have not found this treatment of much value (see Chapter 24).

CHEMODENERVATION

Injection of the extraocular muscles with botulinum toxin, type A (Botox) has been suggested as a viable alternative in the treatment of infantile esotropia. For further discussion, see Chapter 25.

Nonaccommodative Convergence Excess Esotropia (Normal AC/A Ratio)

Definition

Nonaccommodative convergence excess esotropia is defined as an esotropia that is larger at near (at least 15°) than at distance fixation in an optically fully corrected patient whose AC/A ratio is normal when determined with the gradient method.

Clinical Characteristics

As is the case in accommodative esotropia, the onset is early in life, occurring as a rule between 2 and 3 years of age, but we have also seen patients in whom the onset was shortly after birth. Such patients are characteristically orthotropic or have a small angle esotropia at distance fixation and a larger esotropia (20° to 40°) at near fixation. In contradistinction to esotropia with a high AC/A ratio, however, relaxation of accommodation by bifocals or its facilitation by miotics has little if any effect on the near deviation. The AC/A ratio, if determined with the gradient method, may be normal or abnormally low. This condition differs from the “hypoaccommodative” esotropia of Costenbader (see above) inasmuch as the near point of accommodation is within the normal range. Obviously, excessive convergence in such patients must occur on a basis other than accommodation, perhaps from tonic innervation, which is the reason we suggested the term nonaccommodative convergence excess for this entity.

Clearly, an abnormal distance-near relationship in the angle of esotropia is not always caused, as has been assumed, by a high AC/A ratio. The widespread and, in our opinion, unsound practice of determining the AC/A ratio by comparing the distance and near deviation (heterophoria method; see Chapter 5) is likely to miss the diagnosis of nonaccommodative convergence excess and to subject such patients to bifocal therapy to which they will not respond. Case 16–4 illustrates the features of this form of esotropia.

CASE 16–4

A 5½-year-old girl was first noted to intermittently cross her eyes when she was 2 years of age. Amblyopia of OD was diagnosed by her local ophthalmologist, and she responded well to occlusion treatment begun at 4 years of age. On examination, her uncorrected visual acuity was 20/40 + 1 OD and 20/25 + 2 OS. The prism and cover test showed 12° esotropia at distance and 30° at near. When the measurement was repeated on several occasions with the patient looking through +3.00 spherical lenses, the near deviation still measured 22°. The patient manifested a slight A pattern with minimal overaction of both superior oblique muscles. Cycloplegic refraction indicated the presence of mild hypermetropia of +0.75 sph OD and +1.00 sph OS. The remainder of the examination, including the fundus examination, was normal. Clearly, the persistent increased near deviation after relaxation of accommodation with +3.00 spherical lenses must have been caused by factors other than accommodative convergence.

Treatment

Since bifocals or miotics are ineffective in controlling the deviation at near, surgery must be considered for the nonaccommodative element of the anomaly. In our hands, a conventional recession procedure of both medial rectus muscles (4 to 5 mm) alone or combined with posterior fixation sutures has been surprisingly ineffective in significantly reducing the near deviation. In fact, this combined operation performed in the patient described in Case 16–4 reduced the near deviation from 30° to 25° esotropia! We feel that unconventionally large recessions of both medial rectus muscles (5 to 8 mm) may be more effective, but more clinical experience must be accumulated before recommendations regarding the most effective management can be made.

Acquired or Basic Esotropia

Definition

We define acquired nonaccommodative esotropia as a comitant esotropia with a gradual onset after 6 months of age but usually limited to childhood and a near deviation that approximately equals the
distance deviation. Unlike in refractive accommodative esotropia a significant uncorrected hypermetropic refractive error is absent and unlike in nonrefractive accommodative esotropia the AC/A ratio is normal.

Clinical Characteristics

Costenbader referred to this type of deviation as “acquired tonic esotropia,” and the Hugonniers called it “essential esotropia of late onset.” At the onset the angle of strabismus generally is smaller than in patients with essential infantile esotropia, but the angle tends to increase to a magnitude of 30° to 70°. Since the eyes usually straighten out or even become divergent under general anesthesia and since the forced duction tests are, as a rule, negative, we are inclined to implicate an innervational anomaly rather than mechanical factors as the cause of this form of strabismus. Because parents frequently associate onset of the deviation with injury, illness, or emotional upset of the child, Costenbader postulated that such patients have an excessive convergence tonus that is controlled initially by fusional divergence but is disrupted easily by exogenous factors.

The clinician should always keep in mind the possibility of an underlying lesion or malformation in the central nervous system in a young patient with acquired nonaccommodative esotropia, the onset of which need not always be acute. Many of these patients receive treatment for their esotropia and some even undergo strabismus surgery before the correct diagnosis, which may include a brain tumor or other life-threatening condition, is made. It behooves the ophthalmologist to search for signs of increased intracranial pressure in all patients with an acquired esotropia. Special vigilance is called for when the esodeviation is greater at distance than at near fixation (divergence paralysis; see Chapter 22), which has been described in a tumor of the corpus callosum and in Arnold-Chiari malformation.

The importance of a fundus examination to rule out papilledema or optic atrophy in every patient with strabismus is convincingly demonstrated by the case report of a patient with a gradually acquired esotropia who turned out to have an underlying life-threatening condition.

CASE 16-5

A 5-year-old boy who had gradually developed esotropia 6 months before our seeing him was referred for treatment of amblyopia. The referring physician had performed a cycloplegic refraction (+3.50 sph OU) and prescribed glasses. On examination, the best corrected visual acuity was 6/30 OD and 6/9 OS. The prism cover test showed a comitant esotropia of 40° at distance and 50° at near fixation. Examination of the ductions and versions revealed minimal underaction of the right lateral rectus muscle and minimal overaction of the right superior oblique muscle. The patient suppressed OD at near and distance with the Worth four-dot test. Fundus examination showed massive choking of the optic nerve head in both eyes. Computed tomography revealed a midline posterior fossa tumor, which was successfully removed 2 days later and identified as an astrocytoma. The papilledema receded postoperatively, and visual acuity OD improved to 6/9 after 1 month of occlusion therapy. The esotropia remained unchanged, however, and 4 months after brain surgery the right medial rectus muscle was recessed 4 mm and the right lateral rectus muscle resected 7 mm. Six months after muscle surgery the patient had a best corrected visual acuity of 6/9 OD and 6/8 OS. The patient was orthophoric at near and distance fixation, and he had 60 seconds of arc stereoaucity while wearing his glasses. Without glasses he had a residual esotropia of 4° at distance and 10° at near fixation. Fundus examination revealed postpapillitic gliosis but no atrophic discoloration of the nerve heads.

Therapy

Therapy consisting of elimination of amblyopia followed by surgical correction should be started as soon as possible after the onset of the deviation. Since a period of normal binocular vision has existed for at least 6 months or longer before the onset of the disease, the prognosis for normalization of binocular functions is better than in those with essential infantile esotropia, provided treatment is started without delay. Lang reported that if onset occurs after the age of 1½ years a complete cure as defined by orthotropia and random-dot stereopsis becomes possible after surgical alignment and he referred to this form of strabismus as normosensorial late-onset esotropia. Dankner and coworkers reported that a consecutive exotropia in these patients during the immediate postoperative period resulted in a higher incidence of fusion than in those who were initially orthophoric or undercorrected.
Esotropia in Myopia

It is well established that myopia is present in 3% to 5% of patients with nonaccommodative esotropia, and most clinical characteristics of this esotropia are no different from those associated with emmetropia or hypermetropia. There are, however, two special forms of esotropia occurring with myopia that, in view of their unusual features, deserve separate discussion. Von Graefe\textsuperscript{103} recognized the first type, and Bielschowsky\textsuperscript{20} later described it in detail. The esotropia is accompanied by diplopia first only at distance and eventually also at near fixation, mild limitation of abduction in both eyes, and normal adduction. It occurs predominantly in young myopic adults. The explanation given by von Graefe and Bielschowsky for the etiology of this entity is speculative. Bielschowsky advised resection and advancement of both lateral rectus muscles to treat these patients. This condition must be very rare since we have encountered it only once in 35 years of a practice, in a 19-year-old Asian man who had a myopia of $-6D$ in both eyes.

The second type is caused by restrictive factors and will be discussed in Chapter 21.

The treatment of \textit{unrestrictive esotropia} with myopia is not different from the treatment of hypermetropic esotropes. However, special precautions are in order in a patient with a thin sclera. To avoid any scleral suturing we have successfully treated a highly myopic esotropic patient with a history of retinal detachment by performing a marginal myotomy of the medial rectus and a resection followed by end-to-end suturing of the lateral rectus muscle.\textsuperscript{201} Coats and Paysse\textsuperscript{48} reported a technical modification of the classic resection and resection procedure to avoid scleral sutures in such cases.

Acute Acquired Comitant Esotropia

The onset of acute acquired comitant strabismus is always an alarming event for both the patient and the physician. In young children and infants the acute mode of onset can rarely be determined with certainty and voluntary closure of one eye may often be the only sign. In older children or adults with acute strabismus, sudden diplopia is of immediate concern and the onset of the disease often can be traced to a precise hour of a particular day. In patients with acute strabismus, regardless of how obvious the etiology might be, an especially careful motility analysis is always necessary to rule out a paretic deviation.

Unilateral or bilateral paresis of the abducens nerve, commonly the first manifestation of a central nervous system disorder or of a medical problem, may cause an acute esotropia with an angle greater at distance than at near fixation (see also divergence paralysis, Chapter 22). This deviation may quickly become comitant, in which case it will be difficult to recognize the paretic element. Thus any acute esotropia with the prominent complaint of diplopia of sudden onset calls for increased vigilance and may require a neurologic evaluation even though its cause may be quite harmless.

We distinguish three forms of acute comitant strabismus: (1) that occurring after artificial interruption of binocular vision; (2) that occurring without interruption of binocular vision as a result of a decompensated esophoria; and (3) that caused by an intracranial pathologic process.

Acute Strabismus After Artificial Interruption of Fusion

By far the most commonly encountered form of acute strabismus in clinical practice is that which occurs after temporary occlusion of one eye in patients with no previous history of disturbance of binocular vision or in the course of treatment of amblyopia in patients without strabismus (anisometropic amblyopia). When the patch is removed, the occluded eye will be in an esotropic position or, in adults with large angle exophoria, occasionally in an exotropic position. This disturbing event has been reported when one eye has been bandaged for several days, after a perforating corneal injury, after excision of a chalazion, or as in Case 16–6, after swelling of the lids following blunt trauma. Swan\textsuperscript{278} reported a group of patients in whom a large angle esotropia developed in the course of occlusion therapy for amblyopia. In several of them, surgery was necessary to straighten the eyes.

CASE 16–6

\begin{tabular}{|l|}
\hline
\textbf{Age}: 5 years \\
\textbf{March 17, 1971} \\
Routine eye examination, no visual complaints \\
\hline
\end{tabular}
The child in Case 16–6 had an esophoria that decompensated and became manifest after artificial disruption of fusion. The deviation was readily controlled with glasses, and eventually fusional amplitudes recovered sufficiently to control the deviation without glasses.

Occlusion of one eye presents an obstacle to binocular vision since it disrupts fusion. Once fusion is disrupted and the compensatory mechanism is thus suspended, a formerly latent esodeviation will become manifest. In some patients, correction of the underlying refractive error will straighten the eyes. In others, the deviation is of a temporary nature and will disappear spontaneously. In another group, surgery may be indicated. The prognosis for restoration of normal binocular vision is excellent, although improvement is not always so spontaneous, and in some patients surgery may become necessary. It is prudent to perform a refraction before considering patching one eye for whatever reason. In the presence of a significant uncorrected hypermetropic refractive error, the patient should be warned that an esotropia may ensue from wearing the patch.

**Acute Esotropia Without Preceding Disruption of Fusion (Burian-Franceschetti Type)**

This form of strabismus is characterized by an acute onset with diplopia, a relatively large angle esotropia, absence of signs of paralysis, and a good potential for binocular cooperation. The refractive error, as a rule, is insignificant and the accommodative element is minimal. Disruption of fusion is not an etiologic factor, and in most instances the deviation apparently occurs spontaneously. However, in some patients a debilitating illness or physical or emotional stress may precede the onset of the deviation.

This form of acute strabismus was first described by Burian, who reported on four patients ranging in age from 11 to 72 years who had esotropia of acute onset and diplopia. All had low hypermetropic refractive errors, and the angle of strabismus ranged from 20° to 60°. In all patients there was good binocular cooperation with the angle of strabismus corrected, and the functional results following surgery were excellent. Additional cases were reported by Franceschetti and Bischler, and this form of strabismus has become associated with Franceschetti’s name in the European literature. Several additional cases have been reported, and the literature was reviewed by Burian and Miller. It appears that such patients have an asymptomatic esophoria with only a slim reserve of fusional amplitude that maintains alignment of the eyes over the years but that may become lost under the influence of physical or emotional strain. In addition to a favorable outcome after surgical management of this condition good results have also been obtained after chemodenervation. We feel that surgical correction of acute-onset comitant esotropia in children under 5 years of age who are neurologically normal should not be delayed for longer than a few months to avoid the development of suppression and amblyopia. In visually mature children and adults this risk no longer exists and a longer delay of surgery is tolerated.

**Acute Esotropia of Neurologic Origin**

This potentially threatening event is fortunately rare but should always be kept in mind when
encountering acute-onset comitant esotropia. As mentioned earlier in this chapter, a comitant esotropia with a gradual onset may occur in conjunction with Arnold-Chiari malformation,147, 263, 264, 299 intracranial astrocytoma, and other brain tumors,107, 295 but the onset may also be sudden in any of these conditions. In the case of craniocervical junction anomalies, suboccipital decompression should precede strabismus surgery since otherwise recurrence of the esotropia is common.297 Unlike in the two former forms of acute esotropia, the functional results after surgical alignment of the eyes are not always favorable in acute esotropia of neurologic origin.43, 297

Unless the cause of acute-onset esotropia is obvious, such as after artificial interruption of binocular vision or uncorrected hypermetropia, an underlying neurologic condition should always be considered. Hoyt suggests that the presence of nystagmus in such patients or failure to restore normal binocular vision with surgery should be sufficient cause to proceed with a neurologic evaluation.

Microtropia

Ultrasmall angles of strabismus may escape diagnosis by ordinary methods of examination and are frequently overlooked. The cover test may be negative, or the fixation movement of the deviated eye may be absent or so small that it defies detection by the examiner when the sound eye is covered. Since amblyopia is a regular feature of microtropia, such patients often are subjected to an extensive, costly, and quite unnecessary neurologic evaluation in an effort to establish the cause of reduced visual acuity in one eye.127

Thus microtropias are of considerable clinical significance. In view of the confusion with respect to the terminology and clinical characteristics of microtropias, a discussion of this entity must be quite detailed.

The ophthalmic literature has become redundant with descriptions of numerous forms of ultrasmall angles of strabismus. Many authors have introduced their own definitions and terms for what often appear to be similar, overlapping, or even identical clinical entities. Parks rightfully comments on the monstrous semantic structure that has evolved, including, among others, the terms retinal slip, fixation disparity, fusion disparity, retinal flicker, monofixational esophoria, monofixational syndrome, strabismus spurious, microtropia unilateralis anomoluo-fusionalis, microstrabismus, and minisquint. At the present stage of our knowledge, it may be difficult and even impossible to bring order into this system. Before attempting to do so, a brief historical review of ultrasmall angle deviations is in order.

Historical Review

Irvine reported detailed studies on 16 apparently nonstrabismic, anisometropic amblyopes in whom the 4° base-out test elicited positive scotoma responses and in whom close observation of the corneal reflex revealed “only reasonably good fixation.” This combination of amblyopia, anisometropia, and unsteady or possibly nonfoveal fixation may have been one of the first descriptions of what is currently recognized as microstrabismus.

Irvine’s study was followed by several reports on small angle deviations characterized by foveal suppression of the deviated eye and normal or near-normal peripheral fusional amplitudes. These forms were referred to as “retinal slip” by Pugh, “esophoria with fixation disparity” by Gittoes-Davies, “flicker cases” by Bryer, and “fusion disparity” by Jampolsky. The term fixation disparity entered the discussion of small angle strabismus, adding further to the confusion in terminology. Jampolsky, who uses the term fixation disparity interchangeably with fusion disparity, defines this as a heterophoria in which there is no exact bifoveal fixation. In his opinion, fusion (fixation) disparity occupies an intermediate state between heterophoria and heterotropia. Cranford pointed out that gradual transitions exist between orthophoria and microstrabismus; he considers fixation disparity a manifestation of abnormal binocular vision. Ogle and coworkers had previously used the term fixation disparity to describe a long-known minute maladjustment of the visual axis ranging in magnitude from several minutes to maximally 20 minutes of arc. It occurs in subjects with heterotropia but also in those with normal binocular functions, equal visual acuity in each eye, and absence of suppression scotomas.

Parks and Eustis and Parks applied the
term monofixational phoria to patients with esodeviations in whom the angle of strabismus was larger on the alternate cover than on the cover test. Parks believed that the deviation in this group was kept partially latent by peripheral fusion and that macular suppression, normal retinal correspondence, mild degrees of amblyopia, gross stereopsis, and normal fusional vergences were other features of this entity. Peripheral fusion with normal retinal correspondence was thought to be possible in such cases on the basis of a “stretched Panum’s area.”

In a later paper, Parks presented his most recent thinking on the monofixational syndrome—to replace all terminology previously introduced by him. Patients with this syndrome are those in whom a “macular” scotoma in one eye (monofixation), good peripheral fusion with fusional amplitudes, and gross stereopsis are present consistently. Variable features associated with the monofixational syndrome are a history of strabismus, anisometropia, organic unilateral macular lesions, amblyopia, nonfoveal fixation, orthophoria, small angle heterotropia, and possibly a deviation that is larger on the alternate cover test than on the cover test. The monofixational syndrome may occur (1) primarily because of inability to fuse similar macular images, (2) secondary to treatment of large angle strabismus, (3) secondary to anisometropia, and (4) secondary to a unilateral macular lesion.

Lang introduced the terms microstrabismus and microtropia to describe small angle heterotropias of less than 5° associated with harmonious anomalous retinal correspondence, partial stereopsis, and mild amblyopia. He had reported this form of heterotropia earlier in conjunction with an inconspicuous angle of the deviating eye and summarized his studies on microtropia in a monograph. According to Lang’s concept, microtropia occurs in a primary and consecutive form. The primary form may remain constant during life, or the angle of heterotropia may increase (decompensating primary microtropia). Lang recently proposed that primary microtropia may be related to the strong dominance of the fixating eye. The nondominant eye fails to track precisely in unison with the dominant eye, the binocular connection is loosened, and microstrabismus with anomalous correspondence develops. Consecutive microtropias are caused by surgical or optical correction of a large angle heterotropia and are a common finding after surgical alignment of essential infantile esotropia.

Three types of microstrabismus can be differentiated according to the fixation behavior: (1) central fixation; (2) eccentric fixation and anomalous retinal correspondence, the angle of the anomaly being larger than the degree of eccentricity of fixation; and (3) an angle of anomaly identical to the degree of eccentricity of the monocular fixation (microtropia with identity). Lang thus includes patients in whom the cover test is positive, (1) and (2), and those in whom it is negative, (3). Lang also stressed the frequent occurrence of familial microtropia and drew the interesting but unsupported conclusion that anomalous correspondence in these patients is a primary and hereditary defect, but revised this opinion recently. Holland and Richter had previously discussed this possibility in connection with small angle deviations. Cantolino and von Noorden reported an uncommonly high prevalence of sensory, motor, and refractive anomalies in families with microtropus propositi, but rejected the concept that microtropia is a primary congenital defect. Rather, they believe that microtropia is the result of multiple and independently inherited refractive, sensory, or motor anomalies.

Helveston and von Noorden questioned whether some of the strabismus forms that Lang had placed in the category of microtropia are sufficiently specific or different from those long known and accepted by ophthalmologists as “small angle deviations” to deserve a special classification. They suggested that the term microtropia be reserved to describe a unique form of sensorial adaptation in which the cover test is negative and amblyopia, eccentric fixation, and harmonious anomalous retinal correspondence are present (type 3 of Lang). In these cases, the angle of heterotropia equals the distance between the fovea and the area of eccentric fixation. The eccentric area is used for binocular as well as for monococular fixation; therefore the cover test will be negative because a fixation movement is not required when the fixating eye is covered. Peripheral fusion with fusional amplitudes and gross stereopsis are usually present. The functional completeness of sensorial adaptation in microtropia, as defined by Helveston and von Noorden, is emphasized further by their finding that heterophorias may be present in a direction opposite that of microtropia (exophoria with microesotropia). Cüppers had previously pointed out that there
are patients with eccentric fixation in whom the degree of eccentricity of fixation under monocular conditions and the angle of anomaly under binocular conditions are identical. Holland described cases of amblyopia, an inconspicuous angle of esotropia, and anomalous retinal correspondence that were identical to those currently being classified as microtropia.

The high incidence of anisometropia in patients with microtropia suggests a possible etiologic relationship. Von Noorden assumed that, unlike other forms of strabismus in which suppression occurs secondary to the motor anomaly, microstrabismus may develop secondary to a foveal scotoma caused by uncorrected anisometropia during early infancy. With foveal function thus diminished early in life and before the fixation reflex is fully developed, he postulated that the fixation reflex may become adjusted to extrafoveal retinal elements having a higher visual function than the fovea. Such an event may lead eventually to eccentric fixation under monocular conditions and to anomalous retinal correspondence under binocular conditions.

Lang argued against Helveston and von Noorden’s restricting the definition of microtropia. He pointed out that although fixation may be eccentric in persons having microtropia, the degree of eccentricity need not necessarily coincide with the angle of anomaly. Thus he continues to group together patients with central fixation (positive cover test), eccentric fixation without identity with the angle of anomaly (positive cover test), and those in whom identity exists (negative cover test).

Epstein and Tredici pointed out that microtropias do not occur exclusively with esodeviations but that there are also microexotropias that can be diagnosed only by using the 4 test base-in.

Current Concepts and Clinical Significance

From the foregoing reports and observations, it is obvious that a large spectrum of strabismus forms exist with inconspicuously small angles and various degrees of sensorial adaptations. Whether additional attempts to further categorize such deviations are clinically useful is debatable. On theoretical grounds, one could conceive of a spectrum that ranges from normal binocular vision with bifixation at one end, to fixation disparity as defined by Ogle and coworkers, the various manifestations of microtropia, and small-angle esotropia at the other end. (See also de Decker and Haase.) However, to force biological phenomena into an orderly and rigid scheme that satisfies the human intellect is not always possible. A certain amount of overlap and variance is more in accordance with nature’s ways and will defy all such attempts. We agree with Crone that it is more important to analyze the binocular mechanism in each patient than to set up artificial barriers by a multiplicity of terms and classifications.

For these reasons, none of the definitions and classifications of ultrasmall angles of strabismus currently in use is without flaw. For instance, the monofixational syndrome of Parks includes patients without manifest strabismus and with no sensory anomalies other than a unilateral foveal scotoma. Strictly speaking, this latter entity would have no place in a discussion of strabismus were it not for the fact that, in patients with essential infantile esotropia, unilateral foveal suppression under binocular conditions occurs invariably as an end state after complete surgical alignment is achieved. The term monofixation is somewhat misleading since fixation may occur with each fovea in spite of the fact that a foveal suppression scotoma may be present. Also, the term fixation refers to seemingly steady maintenance of the image of the object of attention on the fovea, that is, to a motor rather than to a sensory process. Lang includes heterotropias as large as 5 in his classification of microtropia, although there are no obvious pathophysiologic or clinical differences between an esotropia of 5 or, say, 10. Thus the borderline between microtropia as defined by Lang and a small angle esotropia is poorly defined. A small angle esotropia is said to be present when the deviation is cosmetically noticeable but not disfiguring. Whether strabismus presents a cosmetic problem depends, among other factors, on the facial configuration of the patient since a certain angle of strabismus can be inconspicuous in one patient and cause a significant cosmetic disfigurement in another. It is therefore somewhat arbitrary when Lang refers to small angle esotropia when the deviation is between 5 and 12 and large angle strabismus when the deviation exceeds 12.

The group singled out by Helveston and von Noorden is unique in regard to completeness of sensorial adaptation, but the relationship between the degree of eccentricity of unilateral fixation and the angle of anomaly under binocular conditions cannot always be established unequivocally.
Anisometropia, though often associated with microtropia as defined by these and other authors, is not a consistent finding and anisometric amblyopia may occur without microtropia. On the other hand, the anisometropia may no longer be present when the diagnosis of microtropia is made. The patient material published by Lang and noted in Case 16–7 (see below), and several other patients with primary microtropia and emmetropia observed by us add support to Lang’s contention that primary microtropia, unrelated to anisometropia, is a distinct entity. In view of the clinical importance of ultrasmall heterotropia and the confusion in the literature to which almost everyone writing on this subject has added a share, the following synthesis appears useful.

In addition to large and small angle esodeviations with specific sensory and motor characteristics, manifest esodeviations with inconspicuously small angles also exist. We have adopted Lang’s microstrabismus or microtropia as an appropriate term to describe these deviations. Consistent findings in such patients are amblyopia; abnormal retinal correspondence (as determined with the Bagolini striated glasses or the foveo-foveal test of Cuppers); relative scotoma on the fovea or, in the case of parafoveal fixation, the fixation point of the deviated eye (as determined with the base-out or base-in prism tests, the Bagolini test, or with binocular perimetry); normal or near-normal peripheral fusion with amplitudes; and defective stereoaucity (see Table 16–4). Variable findings include the size of the deviation, foveal or nonfoveal fixation behavior, identity between the degree of eccentric fixation and the angle of anomaly, the presence or absence of anisometropia, and positive or negative cover test results.

Microtropia is a stable condition in most patients but not a guarantee against subsequent deterioration into larger deviation as shown by many studies. A microtropia with identity from nonstrabismic abnormalities causing decreased visual acuity in one eye. A cycloplegic refraction should be carried out at the beginning of such an examination since microtropia occurs frequently with anisotropic amblyopia. Examination of the fixation pattern (see Chapter 15) will establish whether foveolar or parafoveolar fixation is present. The finding of nonfoveal fixation in the amblyopic eye clearly establishes the diagnosis of microtropia (Fig. 16–8). Identification of microtropia is more difficult in isometric patients and in those with minute degrees of fixation anomalies, for the mere presence of a fixation spot scotoma, diagnosed with the Bagolini striated glasses (see p. 228), polarized visual acuity charts, or the base-out (see p. 218) or base-in prism cover test does not establish unequivocally whether the underlying cause is functional, as in microtropia, or organic. Likewise, stereoaucity is reduced not only with functional amblyopia but also when foveal function is reduced by organic lesions. In such patients, the foveo-foveal test of Cuppers (see p. 230) may be helpful (see Chapter 15). The finding of a minute angle of anomalous retinal correspondence clearly identifies the patient as having microtropia, even if the results of the cover test are negative or the amplitude of the fixation movement of the amblyopic eye is too small to detect when the sound eye is covered. Fusional amplitudes (on the basis of anomalous retinal correspondence) can be elicited with rotary prisms or on the amblyoscope and recordings of binocular VEPs are consistent with the presence of peripheral fusion.

A microtropia should always be suspected in unilateral decrease of visual acuity for which no organic cause can be found in patients without apparent strabismus or a history of such and without significant refractive errors or anisometropia. Extensive neuro-opthalmologic evaluations and parental fears of an intracranial lesion can be avoided by making the correct diagnosis, as shown in Case 16–7.
FIGURE 16-8. Microtropia with identity. A, Eyes appear straight. Mild amblyopia OD. The central suppression scotoma OD has led to parafoveal fixation. B, Cover test fails to reveal a fixation movement. OD continues to fixate with the same extrafoveal elements used for fixation when both eyes were open. C, Visuscope reveals fixation OD 2° to 3° nasal to and 1° below the foveola. (From Noorden GK von: Atlas of Strabismus, ed 4. St Louis, Mosby-Year Book, 1983.)

computed tomography scan of the skull. There was no history of strabismus. The medical history was negative. One maternal aunt had strabismus in childhood for which surgery had been performed. On examination visual acuity was 6/6 OD and 6/60 OS when tested with Snellen letters. The cover and cover-uncover tests were negative at near and distance fixation. The 4° prism test gave a scotoma response OS. Cycloplegic refraction was +1.00 sph OU. Anterior segments and fundi were normal. Examination of the fixation behavior by direct ophthalmoscopy showed steady foveolar fixation OD and unsteady parafoveal fixation with an area 2° above and nasal to the foveola OS. There was anomalous retinal correspondence with the Bagolini and foveofoveal test of Coppers. The patient had no stereopsis on random-dot tests, and his fusional amplitudes were normal. We diagnosed a primary microtropia and since the child had never been treated and visual acuity OS was very low, ordered total occlusion of OD for 2 months. The patient returned 2½ months later with a visual acuity of 6/9 OD and 6/15 OS. Continued occlusion treatment for another 4½ months resulted in a final visual acuity of 6½ OS.

Therapy
Microtropia in the older child or adult does not require therapy. On the contrary, we feel that treatment in such patients is ill advised, for elimination of the central scotoma may cause intractable diplopia. Such patients have comfortable and nearly normal binocular vision with good peripheral fusional amplitudes. In young children up to 6 years
of age, however, attempts should be made to treat the amblyopia. If significant anisometropia is present, we occlude the fixing eye and prescribe the full refractive correction. We have observed many patients in whom the microtropia disappeared under energetic occlusion therapy. Fixation of the amblyopic eye changed from parafoveal to central and steady, visual acuity reached a level of 6/6, retinal correspondence became normal, and stereoaucity improved from 100 to 40 seconds of arc. Other authors have made similar observations. The fact that microtropia, if diagnosed and treated in the young child, can be cured refutes the concept of an underlying primary congenital defect of retinal correspondence, as proposed by several authors.

**Recurrent Esotropia**

An unusual form of esotropia which recurs relentlessly to the same angle despite multiple operations but is fortunately rare was identified by von Noorden and Munoz in 19 of 3000 patients who underwent surgery for esotropia of the essential infantile type or with an onset in early childhood. Among the factors that could conceivably cause such a condition one must consider an increase of uncorrected hypermetropia, a deep-seated anomalous retinal correspondence, nystagmus blockage by convergence, an unstable AC/A ratio, or a blind spot syndrome. None of these factors could be implicated in the patients we studied and the cause of recurrent esotropia for which we used the clinical jargon “malignant” esotropia remains unknown.

**Secondary Esotropia**

**Sensory Esotropia**

Etiology and Clinical Characteristics

Reduced visual acuity in one eye presents a severe obstacle to sensory fusion and in fact may abolish the fusion mechanism altogether. The ensuing strabismus is the direct consequence of a primary sensory deficit, and in such cases the term sensory heterotropia is used. Obviously, the origins of sensory esotropia are numerous, limited only by the number of pathologic conditions that can affect visual acuity in one eye. The most common causes are anisometropia, injuries, corneal opacities, congenital or traumatic unilateral cataracts, macular lesions, and optic atrophy.

In the past it was thought that whether or not a sensory esotropia or exotropia developed depended on the age of the patient at the time of visual acuity decrease in one eye. For instance, Chavasse stated that eyes that are congenitally blind or have lost vision shortly after birth diverge. Hamburger, on the other hand, wrote that most eyes with congenital unilateral blindness or severe visual impairment in early childhood converge. There is similar disagreement in the literature as to the direction of strabismus when the onset of visual impairment occurs during later childhood or adolescence. We have analyzed the records of 121 patients with sensory heterotropia and have encountered esotropia and exotropia of almost equal frequency when the onset of visual impairment occurred at birth or between birth and 5 years of age. Exotropia predominated in older children and adults (Fig. 16–9), and there was no correlation between the degree of visual impairment and the development of esotropia or exotropia. Similar observations were reported by Bielschowsky and by Broendstrup. We also encountered a strikingly high prevalence of overacting inferior and superior oblique muscles in patients with sensory esodeviations or exodeviations. This association, which has also been noted by other investigators, does not have a satisfactory explanation at this time. A prevalence of dissociated vertical deviations in 12.5% of patients with sensory heterotropias may be explained on the basis of loss of fusion.

Several authors have commented on the frequent association between unilateral visual loss from birth or with onset in early infancy, with esotropia, manifest latent nystagmus with a null in adduction, fixation preference in adduction, and a head turn toward the side of the fixing eye. Spielmann coined the term functional monopthalmic syndrome for this entity and suggested that these signs, together with optokinetic asymmetry, are manifestations of optomotor immaturity from lack of normal binocular input during early infancy.

It is not entirely clear why some patients become esotropic and others exotropic when they lose sight in one eye. Bielschowsky explained the increased incidence of sensory esotropia with advancing age as a gradual change of topographic-anatomical orbital factors in adolescence, favoring
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divergence rather than convergence. This explanation is difficult to reconcile with the fact that the orbital axes actually converge slightly between birth and adulthood. The notion that the visually impaired eye, suspended from fusional innervation, drifts into a relative position of rest determined by anatomical factors also does not explain why this position should be one of esotropia in one patient and of exotropia in another in the same age group.

Worth speculated that the direction of a sensory heterotropia is determined by the refractive error of the sound eye; that is, the blind eye will diverge if the sound eye is ametropic or myopic and will converge if the sound eye is hypermetropic. This assumption was not supported by our data, which showed an equal distribution of refractive errors in various patient groups. Possibly, various degrees of tonic convergence during early childhood and perhaps less forceful tonic convergence during adulthood contribute to the direction of a sensory heterotropia. Sensory esotropia is usually comitant; however, we have examined patients with a long-standing sensory esotropia in whom limitation of abduction and excessive adduction were present. Forcedduction tests in such patients reveal restriction of passive abduction, a finding that must be interpreted as evidence for contracture of the medial rectus or the conjunctiva, or both, and Tenon’s capsule.

The clinician must never forget that any type of esotropia, whether diagnosed early in life or at a later stage, may be sensory and could be the first clinical sign of poor visual acuity or even blindness in one eye. For this reason, we consider examination of the entire globe to be an absolutely essential part of the evaluation of all strabismic patients, regardless of how transparent the clinical situation appears. For instance, Costenbader and O’Rourke described a number of children with optic atrophy in whom the chief complaint when first seen was strabismus. Ellsworth reported esotropia to be the second most common presenting sign of retinoblastoma.

Therapy

Treatment usually is directed toward improving the cosmetic appearance by means of surgical correction since, in most instances, the very nature of sensory esotropia precludes restoration of binocular function. An exception to this is children with unilateral congenital or traumatic cataracts of postnatal development. In such patients, the grad-
ual onset of an esotropia heralds disruption of fusion, and cataract surgery should be performed without delay, followed by contact lens correction, occlusion treatment for the amblyopia, and eventually by strabismus surgery. The longer the deviation is allowed to persist, the less likelihood there is of binocular vision being restored after successful cataract surgery, especially in adults with acquired cataracts.286

When the patient is blind in one eye and therapy is aimed only at improving the cosmetic appearance, a base-out prism before the blind eye may be tried to make the deviation seem less obvious. However, most patients require surgery, and an operation should not be discouraged because of the remote chance that the eye may eventually straighten spontaneously or even become exotropic. If that occurs, additional surgery can be performed. There is no need for a patient to go through adolescence with a severe cosmetic handicap that will invariably have a negative psychological effect. In the presence of a head turn toward the side of the fixating eye (functional monophthalmic syndrome of Spielmann266–268), surgery must be performed on the normal eye to normalize the head position.

Depending on the size of the deviation, we prefer to operate on the deviated eye and to perform a recession of the medial rectus muscle which may be combined with resection of the lateral rectus muscle and with an inferior oblique myectomy if this muscle is found to be overacting. An esotropia that is present only at near fixation responds well to posterior fixation of the medial rectus muscle at least 13 mm behind its insertion. If the forced duction tests are positive, a bare scleral recession of the nasal conjunctiva and Tenon’s capsule should be carried out. The surgical result in sensory esotropia is less predictable than when visual acuity is normal in each eye, and adjustable sutures are helpful in improving the alignment postoperatively. Even though surgical alignment of a sensory deviation may create a stable result in many patients,84 the esotropia may recur or a consecutive exotropia may develop years after the first operation. The surgeon is advised to inform patients of this possibility.

**Consecutive Esotropia**

Consecutive esotropia occurs almost exclusively iatrogenically after surgical overcorrection of an exodeviation. This complication and its management are discussed in Chapter 17. A spontaneous consecutive esotropia, that is, a change from exotropia into esotropia without exogenous mechanical factors or an acquired paralytic component, is a most extraordinary occurrence indeed. To our knowledge only one case has been reported.91

**Management of Surgical Overcorrections**

The etiology, management, and prevention of overcorrections after strabismus surgery are discussed in Chapter 26. Overcorrections can sometimes be related to inadequate diagnosis and subsequent inappropriate surgical procedures. In the case of an esotropic patient, this applies specifically to disregard of an accommodative element, a high hypermetropic error, or vertical incomitance (A or V pattern). In other instances they occur without obvious cause and in spite of an appropriate amount of surgery.

The prevalence of consecutive exotropia is surprisingly low and, according to several large surveys, ranges between only 2% and 8% of all esotropes on whom surgery was performed.50, 300 Our own experience is in accordance with these observations.216 These figures contrast sharply with the reported incidence of undercorrections.205, 259

Except for large overcorrections with severe restriction of ocular motility, in which case disinsertion of a muscle must be considered (see Chapter 26), the treatment of consecutive exotropia is one of watchful waiting. From a functional point of view, recent data show that a surgical overcorrection actually may be more beneficial than an undercorrection. The effect of strabismus surgery on sensory adaptations, especially on the normalization of retinal correspondence, is well-known.151, 287 It seems that this effect can be enhanced if consecutive exotropia is allowed to persist for some time; this has led some ophthalmologists to strive intentionally for an overcorrection.57, 75, 133, 142, 252, 300 Even though a consecutive exotropia is far from being universally accepted as a desirable outcome of surgery for esotropia, these data show that waiting does no harm and may even be beneficial before a reoperation is considered.

The nonsurgical management of consecutive esotropia consists mainly of reduction of the spectacles correction if the patient is hypermetropic.
Although such measures may temporarily straighten the eyes, they do not eliminate the basic problem and may lead to accommodative asthenopia in older children, depending on the degree of undercorrection. Prisms base-in may be considered in older patients to eliminate diplopia.

Alternate occlusion, immediately after surgery, is sometimes effective in reducing the exotropia provided that ocular motility is normal, that is, the exotropia is not caused by excessive recession of the medial rectus muscles. We have used this approach for many years and have been moderately successful, particularly in patients without hypermetropia. As a rule, consecutive exotropia decreases with time, and should reoperation become necessary we prefer to wait at least 6 months before proceeding with it. The reader is referred to Chapter 26 for other aspects of the surgical management of overcorrections.

**Esotropia Associated with Vertical Deviations**

Hyperdeviations frequently are found in association with esotropia. Symptoms in each patient must be analyzed carefully since the clinical neglect of associated vertical deviations may severely jeopardize attempts to restore binocular vision. The clinical manifestations of vertical deviations are numerous. They may be comitant in all directions of gaze, incomitant and with all other characteristics of a paretic deviation, absent in primary position, manifest in lateral gaze only, or present as a dissociated vertical deviation. In each instance, one must consider whether the hyperdeviation is primary or secondary in relation to the underlying esodeviation.

**Clinical Characteristics and Diagnosis**

A small angle comitant hypertropia of not more than $3^\circ$ occurs in 50% of patients with constant esotropia and in 25% of all those with heterotropia.$^{254}$ Ductions and versions may be essentially normal with no evidence of a paretic cyclovertical muscle. The etiology of this deviation is unknown; however, during the prism and cover test, the physician must rule out artifacts induced by oblique positioning of the prism.

Incomitant associated hyperdeviations can be placed in two categories: (1) those caused by paresis of one of the cyclovertical muscles and (2) those caused by primary or secondary overaction of one or both inferior or superior oblique muscles. A deviation of the first type is greatest when fixating with the paretic eye in the field of action of the paretic muscle and will exhibit all characteristics consistent with a cyclovertical paresis (see Chapter 18). The degree of esotropia often is small, and in such instances the vertical deviation is primary while the esotropia is secondary to disruption of fusion by the hypertropia. We pointed out earlier in this chapter that during childhood an esodeviation is a common response to interruption of fusion.

Deviations of the second type are characterized by overaction of one or both inferior oblique muscles, usually associated with a V pattern (see Chapter 19). Such patients exhibit the characteristic elevation of the adducted eye (strabismus sursoaductorius; see Chapter 18), and when the involvement is bilateral, they have a large right hypertropia in levoversion and a large left hypertropia in dextroversion. With the eyes in primary position, the hypertropia may be small or nonexistent. Overaction of the superior oblique muscles, usually associated with an A pattern, is less common in esotropes. Overaction of one or both inferior or superior oblique muscles may be secondary to weakness of their ipsilateral antagonists, or apparently primary if dysfunction of the antagonists cannot be established, in which case the generic term elevation or depression in adduction is preferable. As pointed out earlier in this chapter apparent overaction of the inferior obliques, encountered frequently in patients with essential infantile esotropia, must be distinguished from a dissociated deviation.

The etiology and differential diagnosis of elevation in adduction is discussed in Chapter 18. In this chapter it is necessary to say only that some investigators have interpreted the apparently primary overaction of the inferior oblique muscles in patients with horizontal strabismus as being secondary to the horizontal deviation since the oblique dysfunction may disappear after horizontal surgery.$^{21, 51}$

Brief mention must be made of the hyperdeviations that occur, often to the great chagrin of the surgeon, after surgery for esotropia has been performed. In such cases, it is commonly believed that while reinserting one or both of the horizontal rectus muscles the surgeon inadvertently selected a new site either above or below the horizontal
meridian of the globe. Foster and Pemberton, however, reported that purposely raising or lowering the insertion of the horizontal rectus muscles produces only a relatively small hyperdeviation (up to 11°). Many surgeons actually use this effect of vertical transposition of the horizontal muscles to correct small degrees of associated hyperdeviation (see Chapter 25).

Scobee also mentioned the possibility that hypertropia, when occurring postoperatively, actually may have been present before surgery but was not apparent on routine examination. He postulated that a hyperdeviation in association with an esodeviation is a manifestation of mechanical superiority of the inferior over the superior oblique muscle when the eye is in extreme adduction and cannot fixate, the fixation object being hidden by the nose. Against this view one may argue that the inferior or superior oblique muscles do not necessarily overact when the fixation object is no longer visible.

Finally, when large degrees of hypertropia occur postoperatively, consideration must always be given to the fact that surgery may have been performed erroneously on a vertical rather than on a horizontal muscle. How to avoid this very perturbing but by no means unprecedented complication is discussed in Chapter 26.

**Therapy**

In patients with primary paretic vertical deviations in whom esotropia develops secondary to disruption of fusion, therapy is nonsurgical (prisms) or surgical, depending on the amount and type of deviation (see Chapter 20). In such cases, we prefer first to correct the vertical deviation and after surgery to reevaluate the need for additional correction of the horizontal deviation.

In patients in whom one or both inferior oblique muscles are overacting, myectomy of the oblique(s) is combined with horizontal muscle surgery (see also therapy of V esotropia, Chapter 19).

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