PART two

Introduction to Neuromuscular Anomalies of the Eyes
A deviation of the visual axes relative to each other is the most common sign in all neuromuscular anomalies of the eyes except for supranuclear affections. All neuromuscular anomalies of the eyes therefore are classified primarily on the basis of the properties and characteristics of the deviation, its direction, origin, temporal behavior, and various modifications imposed on it by the sensory system.

**Heterophoria and Heterotropia**

Proper alignment of the eyes is guaranteed by a normally functioning sensory and motor fusion mechanism. If sensory fusion is artificially suspended by excluding one eye from participating in vision, motor fusion is “frustrated,” as Chavasse put it, and a measurable relative deviation of the visual axes will appear in most patients. When the obstacle to sensory fusion is removed, motor fusion in most patients will return the visual axes to their proper relative positions. The relative deviations thus elicited are called heterophorias, a very useful term introduced by Stevens. Since heterophorias become evident only when normal cooperation of the eyes is disrupted, they may be defined as deviations kept latent by the fusion mechanism. Figure 8–1 shows the effect of fusion in controlling a large esodeviation that becomes manifest when fusion is disrupted with the translucent occluder of Spielmann.

In the absence of a properly functioning fusion mechanism, a more or less obvious deviation of one of the visual lines will be present. Such deviations, termed heterotropias, are manifest deviations not kept in check by fusion. The term heterophoria and related terms were formed from the Greek words heteros, “other,” “different from,” and phora, “bringing,” “carrying” (compare pherein, to bear, carry, a word from which so many medical and scientific words have been coined). Phora does not mean a tendency, even less so the word phoro from which Stevens states he derived his term. Stevens’ original definition of “heterophoria as a tendency of the visual lines to turn away from parallelism,” copied to this day in many texts, does not properly describe the phenomenon, as Lancaster pointed out.

In accordance with the foregoing definitions, all neuromuscular anomalies of the eyes can be separated into two main classes: latent deviations (heterophorias) and manifest deviations (heterotropias). Manifest deviations are known also by the generic name of strabismus, or squint.

According to Hirschberg the word strabismus originates from the Greek. Hippocrates used the word streblos, “turned,” “twisted,” when he talked about strabismic subjects and the word is
derived from the verb *strephein*, “to twist,” “to turn.” The Romans simply adopted the term *strabismus* into their language from whence it entered medical terminology. The proper Latin expressions were *paetus* and *luscus* which originally meant “one-eyed.” Neither of these terms or their derivatives are used in English but *luscus* survived in the French verb *loucher*, “to squint.” Whether the name of the famous Greek historian and geographer Strabo (“the squinter,” 66 BC–AD 24) had anything to do with the origin of *strabismus*, as has occasionally been claimed, is unlikely since Hippocrates had used the word 400 years earlier. Perhaps Strabo had strabismus and thereby got his name.

**Relative and Absolute Position of Rest**

The position assumed by the visual axes when fusion is suspended has been termed the *relative position of rest* of the eyes. This is an unfortunate term because ocular muscles are never “at rest” in a living, conscious person. It is known today from electromyographic evidence (see Chapter 6) that electrical activity is continuous in extraocular muscles when the eyes are steadily fixating. Indeed, even when fusion is interrupted, the deviation of the visual axes is not a passive but an active process, as shown by the increment and corresponding decrement in electrical activity of certain extraocular muscles. Long before electrophysiologic evidence became available, it was obvious that the eyes are never truly at rest in a waking person. Maintenance of the primary position, fixation, and proper alignment of the visual axes all require the presence of an actively supported tonus and a continuous shift in tonus of extraocular muscles (see p. 111). A differentiation was made therefore between the *relative, functional, or physiologic position of rest* assumed by the eyes when fusion was suspended and the *absolute position of rest* assumed by the eyes in death before the onset of rigor mortis, and in deep anesthesia. The absolute position of rest has also been termed *anatomical* or *static* because it is determined solely by anatomical and other mechanical factors. Spielmann introduced the term *fixation-free position* to describe the position of the eyes in the dark or when both eyes are covered with semiopaque occluders. This position is identical to what Lancaster called, less descriptively, the *static position*.

The term *relative position of rest* is an unnecessary one. Since binocular vision and fusion are not active when the vision of one eye is obstructed, it is best to say the particular position that the eyes assume under those conditions is the *fusion-free position*. Synonymous terms are the *heterophoric position* and the *dissociated position*.

The absolute or anatomical position of the eyes in death is generally one of slight divergence and elevation, yet it does not attain the divergent angle of the orbital axes. The eyes may also be aligned in death. The position of the eyes in death is determined by the absence of nervous impulses to extraocular muscles. Curare or curare-like substances, which inhibit transmission at the neuromuscular junction, can be used to artificially reproduce this situation in normal subjects. Toselli did this and found that the eyes assumed a position of $8^\circ$ to $12^\circ$ of divergence and $3^\circ$ to $6^\circ$ of elevation, which is comparable to the position assumed by the eyes in general anesthesia. Using a linear mea-
Ocular Alignment

Ideally, the fusion-free position of the eyes should be such that the visual lines are parallel in distance fixation and have the proper convergence in near vision. This ideal, termed orthophoria, is infrequently realized; it is only approached more or less closely. Whenever fusion is suspended by some means, there is usually a deviation of the visual axes even though it may be too small to be measured by ordinary clinical means.

Orthophoria therefore is not a normal condition in the majority of people free from ocular symptoms. Consequently, many clinicians consider a certain amount of heterophoria to be normal. Moses, for example, stated that 1° to 2° of esophoria or 1° to 4° of exophoria in distance fixation should be considered physiologic. He went on to say that hyperphoria of 1½ of either eye nearly always produces symptoms; hence, only 0.5° of hyperphoria can be considered to be within the physiologic range. These values are selected on the basis of clinical significance. A clinically significant finding is one that may produce symptoms and may require treatment. It should be noted that the clinical significance of heterophorias depends not so much on their absolute values as on correlated findings, for example, the fusional amplitudes (see Chapter 4).

For a description of a complete alignment of the visual axes with the object of regard and thus of a desirable endstage of strabismus therapy, the term orthotropia appears most suitable. Orthotropia is present when the cover test is negative upon covering either eye in the absence of amblyopia. The latter qualification is necessary to exclude patients with amblyopia and eccentric fixation in whom the cover test may also be negative but in whom the visual axis of the amblyopic eye is not aligned with the object of regard (see Chapter 14). In Greek, orthos means "straight" or "correct" and, according to Lang, tropos means "turn" but also "direction." Thus orthotropia conveys the notion of a correct direction or position of the eyes. Another acceptable term is orthoposition, the position of the eyes in which the visual axes intersect at the fixation point under the influence of fusion. Both orthotropia and orthoposition may be used interchangeably to describe binocular alignment on a fixation target. The term orthophoria is not a good one since, as mentioned above, orthophoria is the exception and heterophoria is the rule in normal binocular vision. The terms straight-appearing eyes or straight eyes, which all too often seem to escape editorial scrutiny in the contemporary American literature, are to be avoided. They lack precision in describing the functional state of the patient since they encompass a whole spectrum of conditions that includes orthotropia, heterophoria of varying degrees and clinical significance, and even microtropia and heterotropia with a small angle.

Direction of Deviation

There are a variety of heterophoric or heterotrophic deviations (Fig. 8-2). If the visual axes converge, the condition is called esophoria or esotropia, and if they diverge it is known as exophoria or exotropia. Hyperphoria or hypertropia occurs if one visual line is higher than the other. It is present on the right if the right visual line is higher than that on the left and on the left if the left visual line is higher than that on the right.

One may also speak of a left hypophoria or hypotropia when the right visual line is the higher one and of a right hypophoria or hypotropia when the left visual line is the higher one. Since deviations of the visual lines are relative, the terms hypophoria and hypotropia may appear to be superfluous, but they are useful, especially in heterotropias, to indicate which eye is fixating. Thus right hypertropia indicates that the (lower) left eye is the one fixating, whereas left hypotropia indicates that the (higher) right eye is fixating. For
the vertical heterophorias or heterotropias, Bielschowsky\(^2\) also used the terms positive (right hyper-) or negative (left hyper-) vertical divergence (or deviation) in his American publications. These terms are still in common usage in German ophthalmology.

A misalignment of one or both eyes around the sagittal axis produces clockwise or counterclockwise rotations of the globe (cyclotropia). Since the direction of the deviation must be defined in each eye, the terms right or left excyclotropia or incyclotropia are used. Cyclodeviations are mostly manifest; hence, differentiation between cyclophoria and cyclotropia is difficult to justify on clinical grounds (see Chapter 18).

As already stated, deviations of the visual axes also frequently are referred to by the time-honored names of strabismus and squint. An esotropia is then a convergent strabismus; an exotropia, a divergent strabismus; a hypertropia, a vertical strabismus; and a cyclotropia, a torsional strabismus.

The terms convergent and divergent strabismus, which are widely used in the continental European literature, have not become popular in this country. This is rather fortunate since convergent and divergent could easily be misunderstood to mean that the convergence or divergence mechanism is implicated. Although this may well be so in some forms of horizontal strabismus, it certainly does not hold true for others. In American usage the term strabismus generally is understood to be synonymous with heterotropia. In British and continental European usage, the word includes both heterotropias and heterophorias. To differentiate between the two, the expressions manifest strabismus and latent strabismus are used. To encompass
both the heterophoric and heterotropic forms, such terms as esodeviation and exodeviation are appropriate.

A person may manifest a heterophoric or heterotropic deviation that combines two or more of the various directions mentioned. He or she may then have an esohyperdeviation, an exohyperdeviation, or a cyclovertical deviation.

**Comitance and Incomitance**

Strabismus may occur in one of two major forms: it is either comitant or incomitant. In comitant strabismus, the deviation is, within physiologic limits and for a given fixation distance, the same in all directions of gaze. In incomitant strabismus, one or more extraocular muscles show signs of underaction or paralysis. The deviation therefore varies in different directions of gaze but is larger when the eyes are turned in the direction of action of the underacting or paralytic muscle. Further differentiation between these two forms of strabismus is discussed in Chapter 20.

The term comitant originally had the form concomitant, derived from the late Latin concomitor, meaning “I attend,” “I accompany,” which implies that despite the deviation, one eye accompanies the other in all its excursions (compare the German Begleitschielen, concomitant squint). Duane suggested comitant, a form which is universally accepted in the American literature and not without linguistic justification.

We speak of incomitance when the angle of deviation changes in different positions of gaze. Incomitance may be caused by innervational factors (paralytic strabismus) or mechanical-restrictive factors.

**Constancy of Deviation**

A manifest deviation need not be present at all times. In some patients the fusion mechanism is inadequate to keep the eyes properly aligned under any circumstances. One then speaks of a constant deviation (constant strabismus, constant esotropia, constant exotropia). In other patients the fusion mechanism functions well in some but not in all circumstances. The deviation then is manifest only at certain times, when the patient awakes from a nap or is tired, ill, under stress, or in particular test situations. An intermittent deviation (intermittent strabismus, intermittent esotropia, intermittent exotropia) is then said to be present. Even though variations may exist between different teaching hospitals, the symbols listed in Table 8–1 are fairly uniformly used for abbreviation of strabismus forms in charts and orthoptic records.

Some patients display a heterophoric deviation for one fixation distance and a heterotropic deviation for another fixation distance (e.g., esotropic in near vision but esophoric in distance vision). Patients with a paralyzed muscle may be heterotropic in one direction of gaze but heterophoric in the opposite direction. Also, patients with an A or V pattern of fixation (see Chapter 19) may be heterotropic in the position of maximum deviation and heterophoric in the position of minimum deviation. In British usage, following the lead of Chavasse, this behavior is termed periodic strabismus, meaning that during the period when the eyes are in a certain position a manifest strabismus occurs. The term is not well chosen, since the word “periodic” generally refers to divisions of time. In these cases the essence is not the time during which a certain position of the eyes is assumed, nor is there any periodicity. It is the position of the fixation object in space that determines the sensorimotor response.

Cases of cyclic heterotropia (see Chapter 21) represent an interesting variant of the intermittent type of deviation. In these patients a manifest deviation appears at regular intervals (e.g., every other day). At the time when the eyes appear to be straight, no latent deviation comparable in amount to the manifest deviation can be measured.

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**TABLE 8–1. Common Abbreviations**

<table>
<thead>
<tr>
<th>Heterophoria</th>
<th>Heterotropia</th>
<th>Intermittent</th>
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<tbody>
<tr>
<td><strong>Near</strong></td>
<td><strong>Distance</strong></td>
<td><strong>Near</strong></td>
</tr>
<tr>
<td>Esodeviation</td>
<td>E’</td>
<td>E</td>
</tr>
<tr>
<td>Exodeviation</td>
<td>X’</td>
<td>X</td>
</tr>
<tr>
<td>Right hyperdeviation</td>
<td>RH’</td>
<td>RH</td>
</tr>
<tr>
<td>Left hyperdeviation</td>
<td>LH’</td>
<td>LH</td>
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By classifying these patients with the general group of intermittent strabismus, we do not imply that the mechanism is the same as that in ordinary intermittent esotropia or exotropia.

State of Vergence Systems

The role of accommodative convergence in determining the relative position of the visual axes has been discussed in Chapter 5. Its role in the etiology of esotropia and in the clinical picture of heterotropias is examined in later chapters (see Chapters 9 and 16). In this chapter mention will be made only that a further classification distinguishes accommodative esotropia from nonaccommodative esotropia. In accommodative esotropia the act of accommodation has a major influence on the deviation, whereas in nonaccommodative esotropia it does not.

Convergence is more active in near fixation and divergence in distance fixation. On this basis Duane6 developed his classification of the comitant motor anomalies. If an esodeviation is greater at near than at distance, one may speak of a convergence excess type; if an exodeviation is greater at near than at distance fixation, then it is referred to as a convergence insufficiency type. If an exodeviation is greater at distance than at near, there is a divergence excess type; if an esodeviation is greater at distance than at near, there is a divergence insufficiency type. We find this classification useful provided it is clearly understood that this terminology is to be used only in a descriptive sense, that is, without etiologic implications. This classification is described in more detail in Chapter 17.

Type of Fixation

The use made of the eyes for fixation is another important criterion for classifying heterotropias. One distinguishes unilateral heterotropias (e.g., right esotropia or exotropia), in which the patient habitually uses one eye for fixation, from alternating heterotropias, in which the patient fixes with either eye. A whole spectrum of fixation habits exists, ranging from extreme unilaterality to free random alternation. The term nonalternating strabismus is preferable to “unilateral strabismus.” Unilateral is the contrary of bilateral. It would seem inappropriate to define an alternating heterotropia as “bilateral strabismus.” The latter term should be reserved for those rare cases in which both eyes are deviated from the primary position (e.g., skew deviations, myogenic or mechanical strabismus).

The term monofixation introduced by Parks20 to describe what Lang12 referred to as microstrabismus or microtropia (see Chapter 16) is somewhat ambiguous. It implies that only one eye is fixating. However, that is also the case in other forms of strabismus. Monofixation could also be interpreted as lack of alternation.

Time of Onset of Deviation

A deviation noted at birth or in the first months of life is termed congenital (connatal). Because the onset is difficult to document at that age (see Chapter 16), the term congenital has been replaced by or is used synonymously with infantile, which includes all forms of strabismus with an onset during the first 6 months of life. If the deviation arises after that age, it is called acquired. A variant of the acquired form is acute esotropia (see Chapter 16). These forms are also spoken of as primary heterotropia. The meaning of the term secondary heterotropia is not quite uniform. In general it refers to a deviation that results from some known cause such as a sight-impairing disease or trauma of one eye (sensory heterotropia), or after surgical overcorrection (consecutive heterotropia). Occasionally an esotropic eye may change spontaneously into an exodeviation, in which case the term consecutive deviation is also used.

Paralytic Strabismus

Paralysis and Paresis

If the action of a muscle or a group of muscles is completely abolished, this condition is a paralysis or palsy; if the action of the muscle or muscles is impaired but not abolished, this is a paresis. It is not always possible to distinguish on clinical grounds between paresis and paralysis since an apparently paralysed muscle may occasionally regain some function after surgery or Botox (botulinum toxin, type A) injection of its antagonist. Inability to move an eye in a certain gaze direction does not automatically imply that the muscle involved is paralyzed, as mechanical factors may
also impede ocular motility. In that case the term paralysis, which should be limited to an innervational cause of restricted motility, is inappropriate.

Muscles Affected

The terms N III, N IV, and N VI paralysis refer to paralyses of muscles supplied by those cranial nerves. If all extraocular muscles supplied by the third cranial nerve are paralyzed, the paralysis is termed a complete oculomotor palsy; if one or more extraocular muscles are spared, the oculomotor palsy is partial. If all extraocular muscles are paralyzed, the condition is called an external ophthalmoplegia. If the intrinsic ocular muscles are paralytic, one speaks of an internal ophthalmoplegia. If both the extrinsic and intrinsic ocular muscles are affected, then complete ophthalmoplegia occurs.

Duration and Cause

The characteristics of a paralytic strabismus vary with time (see Chapter 20). One must therefore separate the cases of paralytic strabismus according to duration of the condition and time of onset. In these cases, as in comitant heterotropias, the paralysis may be congenital or acquired. Acquired paralytic strabismus, in turn, is acute or gradual, and may be caused by trauma, infection, inflammation, vascular conditions, tumors, or degenerative processes.

Seat of Lesion

Depending on the seat of the lesion, neurogenic paralyses of extraocular muscles may be supranuclear, nuclear, or infranuclear in origin. Myogenic paralyses result from diseased states of the muscles themselves.

Mechanical-Restrictive Strabismus

Structural alteration of the muscle itself or of its antagonist may limit its ability to function normally. In that case we speak of mechanical (also structural or restrictive) strabismus as opposed to innervational strabismus.

Orbital Strabismus

Any ocular misalignment caused by anomalies of the orbit or of the face (craniofacial dysostoses, plagiocephaly, hydrocephalus, heterotopia of muscle pulleys, and so forth) may be referred to as orbital strabismus.

REFERENCES