CHAPTER 18

Principles and Techniques of the Examination of Ocular Motility and Alignment

Mark S. Borchert

In the previous chapter, we discussed the anatomy and physiology of the major intracranial and extracranial structures that control eye movements. In this chapter, we discuss normal and abnormal monocular and binocular eye movements as they pertain to the techniques used in the examination of patients with disorders of ocular motility. The reader in further pursuit of the subjects considered in this chapter should consult the books by Nelson and Catalano (1), Leigh and Zee (2), and von Noorden (3).

HISTORY

A careful history should always precede a complete examination of the ocular motor system. Patients with ocular motor disorders may complain of a number of visual difficulties, including diplopia, visual confusion, blurred vision, and the vestibular symptoms of vertigo, oscillopsia, or tilt.

DIPLOPIA

Since misalignment of the visual axes causes the image of an object of interest to fall on noncorresponding parts of the two retinas, usually the fovea of one eye and the extrafoveal retina of the other eye, a sensory phenomenon occurs that is usually interpreted as *diplopia*, the visualization of an object in two different spatial locations. Depending on the nature of the misalignment, the diplopia may be horizontal, vertical, torsional, or a combination of these.

Diplopia that results from ocular misalignment disappears with either eye closed: it is a binocular phenomenon. Binocular diplopia is almost never caused by intraocular disease, although Burgess et al. reported a series of patients who developed binocular diplopia from the presence of a subretinal neovascular membrane in one eye (4). The pathophysiology of binocular diplopia with uniocular disease is unclear, but it may represent the establishment of rivalry between central and peripheral fusion mechanisms.

Diplopia that persists with one eye closed, *monocular diplopia*, is rarely caused by neurologic disease. In almost all cases, it is produced by local ocular phenomena, including uncorrected astigmatism or other refractive errors, corneal and iris abnormalities, cataract, and macular disease (5–10). Most patients with this type of monocular diplopia will recognize a difference in the intensity of the two images they

see. One image will be fairly clear, but the second image will be perceived as ''fuzzy'' and may be described as a ''ghost image'' that overlaps the clear image.

Rare cases of monocular diplopia and polyopia are occasionally reported in patients with central nervous system disease (11–15). Patients with ''cerebral polyopia'' usually do not complain of overlapping images and generally see each image with equal clarity. In addition, the monocular diplopia in these patients is always seen with both eyes (i.e., with either eye covered). Such patients usually have lesions in the parieto-occipital region. The mechanism of cerebral diplopia-polyopia is unknown. Bender postulated that an instability of fixation from occipital disease could produce rapid ocular excursions with consequent stimulation of retinal areas or ''competing maculae'' (11). Other evidence suggesting a role for the occipital lobe in the pathogenesis of cerebral diplopia or polyopia came from the work of Brindley and Lewin, who stimulated different sites in the visual cortex of a patient who was blind from glaucoma (16). The patient reported seeing two, three, or multiple points of light.

Monocular diplopia is occasionally described by patients after surgery to correct congenital strabismus (3,17). In such patients, it is believed that a portion of the extrafoveal retina in the previously deviated eye has been used as a ''fovea'' for many years. Once the true foveae of the two eyes are aligned, there is apparently a sensory conflict in the previously deviated eye between the true fovea and the portion of the retina that previously corresponded to the fovea of the opposite eye. Such patients may complain of monocular diplopia or binocular triplopia. These symptoms usually disappear with time.

Monocular diplopia may be a complaint of individuals with no evidence of ocular or cerebral disease. Such patients should not undergo extensive neurologic or neuroimaging evaluations.

Thus, in any patient complaining of ''diplopia,'' one should first determine if it is binocular or monocular. If the diplopia is monocular and the patient is otherwise healthy, the examiner may concentrate on ocular, rather than neurologic or myopathic, disorders that affect ocular alignment. In patients with binocular diplopia, the eyes are presumably misaligned, and the examiner should ascertain whether the diplopia is horizontal, vertical, or so forth; if it is better or worse in any particular direction of gaze; if it is different when viewing at distance or near; and if it is affected by head posture.

VISUAL CONFUSION

In patients with misalignment of the visual axes, the maculae of the two eyes are simultaneously viewing two different objects or areas. Occasionally, both macular images may be interpreted as existing at the same point in space. This sensory phenomenon is called *visual confusion*. Patients with visual confusion complain that the images of objects of interest are superimposed on inappropriate backgrounds.

BLURRED VISION

Misalignment of the visual axes does not always produce diplopia or visual confusion. In some patients, the images of an object seen by noncorresponding parts of the retina are so close together that the patient does not recognize diplopia but instead complains that the vision is blurred when both eyes are open. Similarly, some patients interpret visual confusion not as image superimposition but as simple ''blurred vision.'' Blurred vision that exists only with both eyes viewing is quite common in the early stages of an ocular motor nerve paresis. In such patients, the blurred vision clears completely if either eye is closed.

Blurred vision that resolves with one but not either eye closed usually suggests a primary visual sensory disturbance. Blurred vision that does not resolve with either eye closed also usually occurs from visual sensory disease but may also occur in some patients with disorders of saccades (e.g., saccadic oscillations such as ocular flutter; see Chapter 23) and in patients with impaired pursuit leading to disordered tracking.

VESTIBULAR SYMPTOMS: VERTIGO, OSCILLOPSIA, AND TILT

Patients with disorders that affect the vestibular system may complain of disequilibrium or unsteadiness, symptoms that reflect imbalance of vestibular tone. A common complaint of patients with vestibular imbalance is *vertigo*, the illusory sensation of motion of self or of the environment. Vertigo usually reflects a mismatch between vestibular, visual, and somatosensory inputs concerning the position or motion of one's body in space (see Chapter 17) (18). Although it is helpful to question patients with vertigo as to the direction of their vertiginous illusions, they are often uncertain because their vestibular sense indicates head rotation in one direction, whereas their eye movements (the slow phases of vestibular nystagmus) are producing visual image movements that connote rotation of the head in the opposite direction. It is best to evaluate the vestibular sense alone by asking the patient about the perceived direction of self-rotation with the eyes closed, thus eliminating conflicting visual stimuli.

Oscillopsia is an illusory to-and-fro movement of the environment that may be horizontal, vertical, torsional, or a combination of these directions. It is usually caused by an instability of fixation from mechanical or neurologic disorders (19–21). When oscillopsia is produced or accentuated by head movement, it is usually of vestibular origin. Oscillopsia is rarely present when ocular motor dysfunction is congenital.

A third group of vestibular symptoms include the perception of *tilts*—static rotations of the perceived world or the body. These complaints usually reflect a disturbance of the otolith organs, from either peripheral or central causes (see also Wallenberg's syndrome in Chapter 19) (22). When dealing with such patients, as with patients who complain of vertigo, the examiner should ask about the perception of the positions of the body with the eyes closed, to eliminate conflicting visual stimuli.

EXAMINATION

The examination of the ocular motor system generally consists of the assessment of (*a*) fixation and gaze-holding ability, (*b*) range of monocular and binocular eye movements, (*c*) ocular alignment, and (*d*) performance of versions (saccades, pursuit). In addition, depending upon the findings of the basic examination, it may be appropriate to test the vestibulo-ocular and optokinetic reflexes and to attempt mechanically to move the eyes using forced duction testing.

FIXATION AND GAZE-HOLDING ABILITY

Principles

In an awake individual, the eyes are never absolutely still. Fixation is interrupted by three distinctive types of miniature eye movements: (*a*) microsaccades, with an average amplitude of about 6 minutes of arc and a mean frequency of about 2 per second; (*b*) continuous microdrift at rates of less than 20 minutes of arc/second; and (*c*) microtremor, consisting of high frequency (40–60 Hz) oscillations of 5–30 seconds of arc (23–25). Square wave jerks—spontaneous, horizontal saccades of about 0.5°, followed about 200 msec later by a corrective saccade and occurring at a rate of less than 9 per minute—can also be observed during fixation in normal individuals. Herishanu and Sharpe suggested that in normal individuals, square wave jerks are sporadically enlarged microsaccades (26). St. Cyr and Fender (27) postulated that the spontaneous eye movements that occur during fixation correct minor fixation errors, whereas Ditchburn (28) believed that such movements may prevent images from fading on the retina. Kowler and Steinman, however, stated that these small saccades serve no useful purpose, since vision remains clear when they are voluntarily suppressed (29).

When no efforts are being made toward ocular fixation or accommodation, the eyes are said to be in a ''physiologic'' position of rest. With total ophthalmoplegia, there is usually a slight divergence of the visual axes, and this position usually also occurs during sleep, deep anesthesia, and death (30–35).

Technique

In patients complaining of intermittent diplopia, visual confusion, or strabismus, tests of sensory fusion (e.g., stereoacuity) and fixation should be performed before the eyes are dissociated by tests of monocular visual function (e.g., visual acuity, color vision, visual fields).

The initial part of the ocular motor examination should consist of a careful study of fixation (36). The patient should be instructed to focus on a distant target, and the eyes should be observed carefully. Asking the patient to describe the target can control attention. If strabismus is present, any preference for fixation with one eye should be noted. Constant or intermittent monocular and binocular eye movements, whether conjugate or dissociated, should be noted. Subtle degrees of abnormal fixation can often be easily detected during the ophthalmoscopic examination (37). The types of fixation abnormalities that may be observed are described in Chapters 19 and 23.

RANGE OF EYE MOVEMENTS

Principles

To discuss eye movements, it is necessary to have a frame of reference against which any movement may be quantified. Accordingly, the primary position of the eyes has been arbitrarily designated as that position from which all other ocular movements are initiated or measured (38,39).

It was once assumed that all ocular motions occurred around a fixed point in the orbit called the center of rotation. It has been shown, however, that there is no fixed center of rotation that does not move when the globe rotates and that the globe translates during every eye movement (40–41). Thus, horizontal movements rotate the center of the globe in a semicircle in the plane of eye rotation, called the *space centroid*. Nevertheless, for practical purposes, the globe can be considered to rotate around a fixed point that lays 13.5 mm posterior to the corneal apex and 1.6 mm nasal to the geometric center of the globe.

All movements of the globe around the hypothetical center of rotation can be analyzed in terms of a coordinate system with three axes perpendicular to each other and intersecting at the center of rotation (Fig. 18.1). These three axes, described by Fick in 1854, are called the *x*, *y*, and *z* axes of Fick (42). The *y* axis is equivalent to the visual axis; the *z* axis is vertical (around which the eye rotates horizontally); and the *x* axis is horizontal (around which the eye rotates vertically). These axes are stable with respect to a frontal plane, fixed in the skull, that corresponds roughly with the equatorial plane of the eye when it is directed straight ahead (Listing's plane) (43).

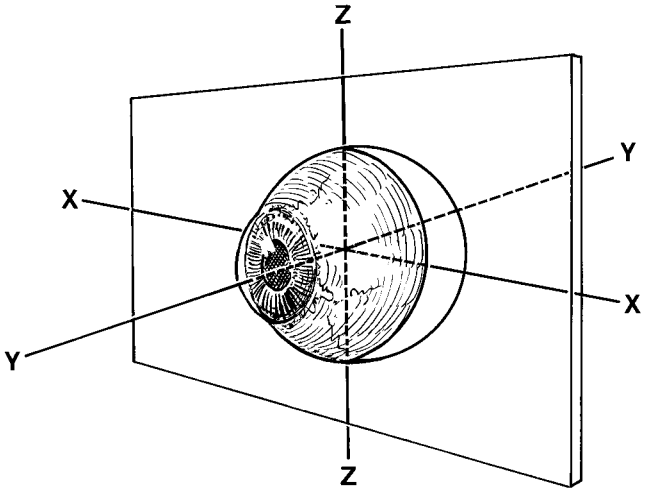

Figure 18.1. The axes of rotation of the eye. The *y* axis corresponds to the line of sight when the eye is in the primary position, looking straight ahead.

Rotations of both eyes (relative to one another) in the same direction are called *versions*. Rotations in opposite directions are called *vergences*. Only *convergence* (movement of the eyes toward one another) in the horizontal plane is volitionally significant. *Divergence* amplitudes (movement of the eyes away from one another) are small in normal individuals.

Rotations of either eye alone without attention to the movements of the other eye are called *ductions.* Horizontal rotation (rotation around the *z* axis of Fick) is termed *adduction* if the anterior pole of the eye is rotated nasally (i.e., inward, medially) and *abduction* if the anterior pole of the eye is rotated temporally (i.e., outward, laterally). Vertical rotation (around the *x* axis) is called *elevation* (or sursumduction) if the anterior pole of the eye rotates upward and *depression* (or deorsumduction) if it rotates downward.

Rotation during ductions or versions around either the horizontal or vertical axis places the eye in a secondary position of gaze. In achieving this position, there is no rotation of the globe around the *y* axis (i.e., there is no torsion).

The oblique positions of gaze are called tertiary positions. They are achieved by a simultaneous rotation around the horizontal and vertical axes, a movement that can be considered to occur around an oblique axis lying in Listing's plane. When an eye moves obliquely out of primary position, the vertical axis of the globe tilts with respect to the *x* and *z* axes of Fick; however, this tilt is considered ''false torsion'' since it does not represent a true rotation around the *y* axis but rather an apparent movement with respect to the planar coordinate system. For an excellent video demonstration of this phenomenon and its effect on visual perception, see *www.med.uwo.ca/physpharm/courses/llconsequencesweb/.* The amount of false torsion associated with any particular oblique position of gaze is constant, regardless of how the eye reaches that position (Donders' law) (44). Tertiary positions of gaze are thus positions of gaze associated with false torsion.

True ocular torsion is defined by the direction of the rotation around the *y* axis of Fick (i.e., the visual axis) relative to the nose. If the 12 o'clock region of the limbus rotates toward the nose, the movement is called *intorsion* (incycloduction; incyclotorsion). If the same area rotates away from the nose, the movement is called *extorsion* (excycloduction; excyclotorsion).

True ocular torsion occurs only minimally during voluntary versions. Volitional head tilts are preceded by a brief torsion movement of the eye in the direction of the anticipated head tilt that disappears by the time the head reaches its desired position (45). Thereafter torsion in the opposite direction occurs to compensate for the head tilt (46). In this setting, the torsion movements are called *countertorsion* or *counterrolling*. Countertorsion has two components, dynamic and static. Dynamic countertorsion occurs during head tilt and reflects the semicircular canal-induced torsional vestibulo-ocular reflex (VOR) (47,48). Static countertorsion persists at a given angle of any head tilt, but the amount of rotation is minor compared with that which occurs from dynamic countertorsion (49–51). Static countertorsion reflects a tonic otolith-ocular reflex (52,53). Each utricle influences both eyes in both directions but primarily controls tilt to the contralateral side (54–56). In addition, abnormalities in static countertorsion that are found in patients with intracranial compressive lesions have been found to correspond in part to the extent to which they impinge on the utricular nerve and brain stem (56).

The entire concept of static countertorsion was challenged by Jampel, who believed that the anatomy of the oblique muscle tendon insertions prevents such torsion movements from occurring (57–61). Numerous other investigators using sophisticated magnetic search coil techniques subsequently confirmed that static countertorsion does exist (62,63).

Most investigators find that static countertorsion represents only about 10% of the total amount of torsion associated with any large head tilt (64–66). Dynamic and static torsion apparently work only within a small range to attempt to keep the sensory vertical raphes of each retina perpendicular to the horizon (67). It is suggested that countertorsion is a primitive compensatory adaptation of lateral-eyed animals to head movements in the roll plane (68). With evolution of the eyes to the frontal plane (to promote stereopsis) and development of upright posture, this movement is merely vestigial.

True torsion does occur as a normal component of voluntary convergence. Both eyes extort, and the extorsion is greater in downward convergence than in upward convergence (69). This may be a phylogenetically recent adaptation in frontal-eyed animals to prevent distortions in pitch stereopsis of vertical lines caused by false intorsion during downward convergence (68). (It is during downward gaze that accurate pitch stereopsis is most important for walking and reading.)

To discuss the independent action of any individual extraocular muscle or any pair of extraocular muscles is strictly a hypothetical convenience. In any actual rotation of the globe, all six muscles are affected and act as a single muscle unit with a single axis of rotation at any given moment (70). The complete muscle unit can produce an infinite variety of rotations consistent with Listing's and Donders' laws that, together, state that when the line of fixation passes from the primary to any other position, the angle of false torsion is the same as if the eye had arrived at this position by turning around a fixed axis perpendicular to the initial and final positions of the line of fixation. Listing's law has been modified to allow temporal rotation of Listing's plane to account for the necessary torsion during convergence (71). Nevertheless, some studies show that Listing's and Donders' laws are not precisely followed in that some true torsion does develop during eccentric gaze (72–74).

Demer et al. have shed light on the anatomic correlates of an extraocular muscle pulley system surrounding the globe roughly parallel to Listing's plane that anchors the functional origins of the muscles relative to one another and largely explains how Listing's and Donders' laws are obeyed (75–77). They have shown histologic and magnetic resonance imaging (MRI) evidence of orbital fibrous connective tissues that create this pulley system, allowing the extraocular muscles to function commutatively.

Within the concept of a single muscle unit, it nevertheless

seems acceptable to discuss the action of the extraocular muscles in the setting of individual antagonist pairs. Boeder emphasized that the two horizontal rectus muscles have only the primary action of either adduction (for the medial rectus) or abduction (for the lateral rectus) (70). The primary action of the two vertical rectus muscles is vertical eye movement (elevation for the superior rectus and depression for the inferior rectus), with both muscles additionally having secondary actions of adduction and torsion (intorsion for the superior rectus and extorsion for the inferior rectus). According to Boeder, torsion is the primary action of the two oblique muscles, with the superior oblique producing intorsion and the inferior oblique producing extorsion. The secondary actions of these muscles are abduction and vertical movement (depression for the superior oblique; elevation for the inferior oblique). For the oblique muscles, the secondary vertical actions are at least as great as the primary torsion effects.

Normal eye movements are binocular. Such movements are called *versions* if the movements of the two eyes are in the same direction. For practical purposes, the extraocular muscles of each eye work in pairs during such movements, with one muscle contracting (the *agonist*) and the other muscle relaxing (the *antagonist*). The three agonist–antagonist muscle pairs for each eye are the medial and lateral rectus muscles, the superior and inferior rectus muscles, and the superior and inferior oblique muscles. From experimentally produced ocular motor palsies, Sherrington proposed that whenever an agonist muscle receives a neural impulse to contract, an equivalent inhibitory impulse is sent to the motor neurons supplying the antagonist muscle so that it will relax (78). This is called Sherrington's law of reciprocal innervation.

For the eyes to move together to produce a horizontal version, the lateral rectus of one eye and the medial rectus of the opposite eye must contract together. These muscles constitute a *yoke pair.* The other two yoke pairs are (*a*) the superior rectus muscle of one eye and the inferior oblique muscle of the other eye and (*b*) the superior oblique muscle of one eye and the inferior rectus muscle of the other eye. Implicit in the concept of a yoke pair is the premise that such muscles receive equal innervation so that the eyes move together. This is the simplest statement of Hering's law of motor correspondence (79).

Techniques

When testing the range of ocular movement, the examiner should ask the patient to follow a target through the full range of movement, including the cardinal (or diagnostic) positions of gaze. The eyes are tested individually with one eye covered (ductions) and together with both eyes open (versions). The normal range of movements is fairly stable throughout life for all directions except upgaze. Normal abduction is usually 50°; adduction, 50°; and depression, 45° (3). Upward gaze decreases somewhat with advancing age. Chamberlain examined 367 ''normal'' individuals ranging in age from 5 to 94 years of age (80). He found that there was a progressive decrease in upward rotation of the eyes from 40° in patients 5–14 years of age to only 16° in patients 85–94 years of age. Thus, limitation of upward gaze in an older individual may simply be age-related and not necessarily a new, pathologic process.

When the range of motion is limited, it is necessary to determine whether the limitation is mechanical and, if not, whether the disturbance is supranuclear or peripheral.

Several tests may be used to determine whether a mechanical restriction of ocular motion is present. Mechanical limitation of motion (such as that seen in patients with thyroid ophthalmopathy or orbital floor fracture with entrapment) can be inferred if intraocular pressure increases substantially when the patient attempts to look in the direction of gaze limitation (81–84). The intraocular pressure measurements are most easily performed using a Tonopen or a pneumatic tonometer, although any similar instrument may be used (83).

Mechanical limitation of motion can more reliably be detected with forced duction (or traction) testing. In such tests, an attempt is made to move the eye forcibly in the direction(s) of gaze limitation (Fig. 18.2). As described by Jaensch, this test is performed as follows (85). The cornea is anesthetized using several drops of a topical anesthetic such as proparacaine or tetracaine hydrochloride. The conjunctiva is further anesthetized by holding a cotton swab or cotton-tipped applicator soaked with 5–10% cocaine against it for about 30 seconds. The conjunctiva is then grasped with a fine-toothed forceps near the limbus on the side opposite

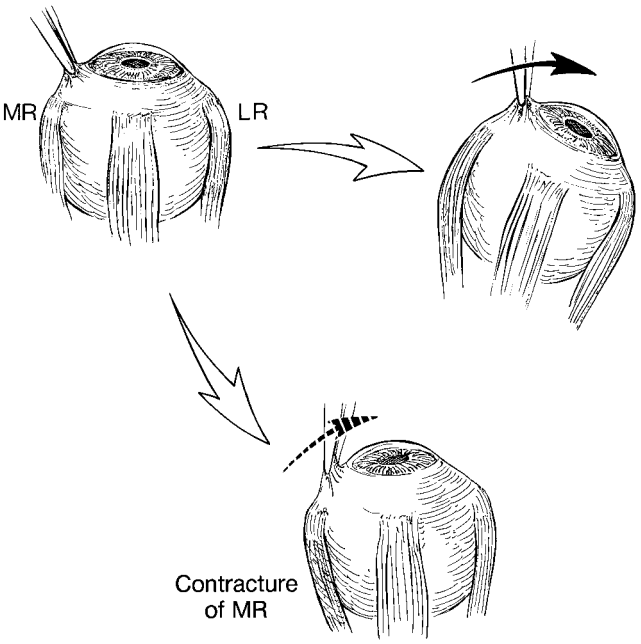

Figure 18.2. Forced duction testing. After the eye has been anesthetized with topical proparacaine and cocaine, the conjunctiva just posterior to the limbus is grasped with a fine-toothed forceps at a point opposite the direction of limitation. An attempt is then made to rotate the eye in the direction of limitation. If no mechanical limitation is present, the eye can be moved fully into the direction of limitation (*solid black arrow*). If mechanical limitation is present, the eye will resist attempts to rotate it into the field of limitation (*dashed black arrow*).

the direction in which the eye is to be moved. The patient is instructed to try to look in the direction of limitation, and an attempt is made to move the eye in that direction (i.e., opposite that in which mechanical restriction is suspected). If no resistance is encountered, the motility defect is not restrictive; however, if resistance is encountered, then mechanical restrictions exist (86–90). In some patients, particularly those who are cooperative and have substantial limitation of movement, the forced duction test can be performed simply by asking the patient to look in the direction of limitation and then attempting to move the eye by placing a cotton-tipped applicator stick against the eye on the opposite side just posterior to the limbus (91). Other investigators recommend using a suction device to perform and quantify forced duction testing (92–94).

Forced ductions can also be used to test restriction of the oblique muscles (95). For this test, the conjunctiva is grasped near the limbus at the 3 o'clock and 9 o'clock position with toothed forceps. Retropulsion of the globe is then applied, putting the oblique muscles on stretch. The eye is then moved from medially to laterally in an arc that follows the orbital rim while depressed (to test the inferior oblique muscle) or elevated (to test the superior oblique muscle). During this process, a distinct bump is encountered as the globe passes over the stretched oblique tendon or muscle. The resistance of this bump toward passage of the globe is an indication of the tightness of the muscle.

Often, particularly in children or when testing restriction of the oblique muscles, the forced duction test can be performed only under general anesthesia (Fig. 18.3). However, succinylcholine, which is often given to patients under general anesthesia, produces tonic contraction of the extraocular muscles, thereby altering the results of the forced duction test (96–98).

In addition to the forced duction test, mechanical determination of muscle force can be used to assess the function of apparently paretic muscles with contracture of their antagonists. An estimate of active muscle force present in patients with limitation of ocular motility can be made by stabilizing the anesthetized eye with a toothed forceps in a position near the limbus on the side of the limitation while the eye attempts to look into the field of the limitation (Fig. 18.4) (99,100). The presence of a tug on the forceps indicates that a contraction of the suspected paralytic muscle has occurred. The results of this ''forced generation test'' can even be quantified (101).

Nonrestrictive limitation of eye movements may occur from disease of supranuclear or infranuclear structures. Since the workup and management of the patient will vary considerably depending on the location of the lesion, supranuclear disorders must be distinguished from infranuclear disorders. From a practical standpoint, supranuclear disorders that cause abnormalities in the range of eye movements usually result from lesions of the cerebral hemispheres or the brain stem premotor structures. In such cases, stimulation of the vestibular apparatus can be used to assess the integrity of the peripheral ocular motor pathways either by oculocephalic testing (the doll's head maneuver) or by caloric testing (102–105).

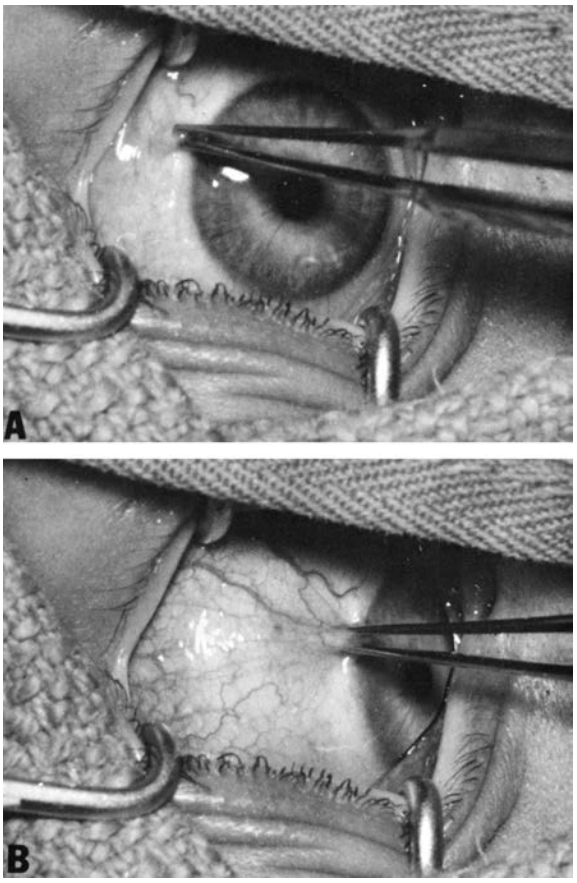

Figure 18.3. Forced duction testing in a patient under general anesthesia. *A,* The conjunctiva and episclera are grasped near the limbus with a fixation forceps. *B,* The eye is moved medially to test for mechanical restriction of adduction. The eye can be moved medially without difficulty. (From von Noorden G, Maumenee AE. Atlas of Strabismus. Ed 2. St Louis, CV Mosby, 1973 : 113.)

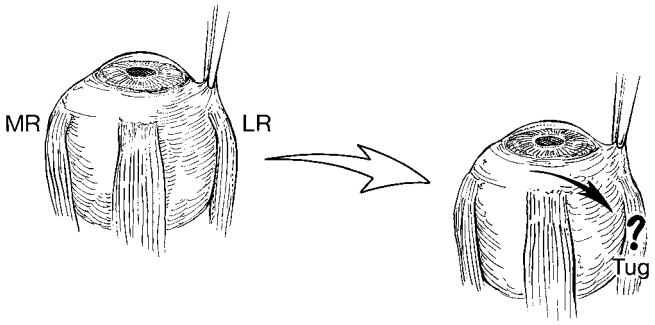

Figure 18.4. Estimation of active, generated muscle force. After the eye has been anesthetized with topical proparacaine and cocaine, the conjunctiva just posterior to the limbus is grasped with a fine-toothed forceps on the side of the limitation. The examiner then holds the eye while the patient attempts to look in the direction of limitation. If there is an intact nerve supply to the muscle that could move the eye into the field of limitation, the examiner will feel a tug on the forceps.

In the oculocephalic test, the awake patient is asked to fixate a target straight ahead while the head (or the entire body) is rotated from side to side and up and down. A normal response consists of a conjugate eye deviation in the direction away from head or body rotation such that the eyes remain stable with respect to space despite the head movement. Asking the patient to read a Snellen chart during head or body rotation can demonstrate the remarkable integrity of this VOR. In patients with intact vestibular systems, there is no degradation of visual acuity, even with rotations of up to 40°/sec (106). Patients with vestibular disease have a rapid decline in this dynamic visual acuity with head rotation.

To perform the oculocephalic test in comatose patients, the eyelids are simply held open and the rotational head movements are performed. Chu et al. modified this procedure for patients with severe neck rigidity or injuries that prevent neck flexion and extension (107). These investigators place such patients on a stretcher with wheels. The stretcher is then sharply pushed in the direction of either the patient's head or feet. This maneuver does not produce a rotational stimulus but is simply a linear or translational movement that may stimulate otolith-ocular reflexes (which are minimal) and visual tracking. In fact, it probably functions as a full-field pursuit test, and similar results can be obtained using a slowly moving ''optokinetic'' tape or drum. Although these tests may not give exact information regarding the VOR, they nevertheless provide important information regarding ocular motility that may be otherwise impossible to obtain.

In unconscious patients, oculocephalic testing may be the most useful method of assessing eye movements. The VOR is often intact in such patients, whereas saccadic and pursuit eye movements are absent. Thus, rapid horizontal rotation of the head results in deviation of the eyes away from the direction of the head turn. The eyes then make an exponential drift back to primary position if the head rotation is maintained. Though not saccadic, this recentration of the eyes may be quite rapid, occurring with a time constant of less than 0.5 seconds in the most severe vegetative states (105). Normal responses during oculocephalic testing indicate that the nuclear and infranuclear ocular motor structures are intact and capable of being stimulated by an intact vestibular system. This test can also be used in patients with functional (nonorganic) limitation of gaze to show that a full range of eye movement can be elicited despite apparent gaze restriction during testing of voluntary eye movements (108,109).

Another way to stimulate the vestibular system is by caloric irrigation (102–104,110–115). In this test, performed in the light with the patient in a supine position, the external auditory canal is first inspected to make certain that the tympanic membrane is intact. The patient's head is flexed 30°. This places the lateral (horizontal) semicircular canals in a nearly vertical position, allowing the thermal stimulus to induce maximal convection currents in the endolymph. Up to 200 cc of warm (44°C) or cold (30°C) water is infused into the external canal using a small tube fitted onto a syringe (116). In the awake patient, a normal response consists of conjugate nystagmus, with the slow phase toward the side of cold water irrigation (or away from the side of warm water irrigation) and the fast phase away from the side of cold water irrigation (or toward the side of warm water irrigation). Temperature stimulation of the vestibular system causes a slow-phase movement followed by a quick refixation movement (saccade) resulting in nystagmus. If the induced nystagmus is consistently less when one ear is irrigated, regardless of the stimulus temperature, a peripheral vestibular disturbance is present on that side. If the nystagmus is consistently greater in one direction, regardless of which ear is stimulated, the patient has a directional preponderance of the vestibular system that may occur with central or peripheral vestibular lesions and is otherwise nonlocalizing (2).

The eye movements that occur during caloric irrigation can best be observed by placing Frenzel's spectacles on the patient (117). These spectacles eliminate patient fixation and provide magnification for the examiner; some models also provide illumination of the patient's eyes.

In practical terms, the caloric irrigation test is messy, uncomfortable for the awake patient, and usually useful only for detecting relatively gross asymmetry in vestibular function. Nevertheless, attempts have been made to quantify subtle vestibular dysfunction with this test. The duration of nystagmus after caloric irrigation seems to be a reproducible measure of vestibular function, and the slow-phase velocity of caloric stimulation nystagmus, as measured with electronystagmography, is a commonly used parameter for assessing vestibular function (118).

Itaya and Kitahara described an air caloric test that causes a continuous thermal change in the semicircular canals, thus avoiding the use of water (119). These inventors claimed that this test is more sensitive than water irrigation for detecting vestibular disorders.

In comatose patients with intact nuclear and infranuclear ocular motor structures and an intact vestibular system, a normal response is simply a tonic, conjugate ocular deviation toward the side of cold water irrigation and away from the side of warm water irrigation. There are no significant refixation movements, since all horizontal quick phases are generated by the paramedian pontine reticular formation (PPRF), which is not functioning in such patients. Absence of the VOR by either oculocephalic or caloric stimulation in comatose patients is consistently associated with poor outcome (120–122).

Caloric testing may be used to evaluate the integrity of vertical gaze by infusing warm or cold water simultaneously into both external auditory canals. A normal response in the awake individual is a conjugate jerk nystagmus with a slow phase that is upward when warm water is used and downward when cold water is used. A normal response in the comatose individual is a tonic, conjugate movement of the eyes upward (for warm water) or downward (for cold water). Although caloric testing is the best way to evaluate unilateral peripheral vestibular function, our experience with bilateral caloric irrigation suggests that it is of limited value in assessing the integrity of vertical gaze and that oculocephalic and rotation testing provides more accurate and reproducible results.

Caloric testing in patients with abolished vestibular func-

tion can occasionally induce nystagmus. This pseudocaloric nystagmus always beats away from the affected ear, regardless of whether cold or warm water is used for irrigation (123,124). It can thus be distinguished from caloric nystagmus that beats away from the irrigated ear when cold irrigation is used and toward the irrigated ear when warm irrigation is used. According to Becker et al., pseudocaloric nystagmus probably represents unmasking of a pre-existing vestibular nystagmus through tactile (caloric) stimulation (see Chapter 23) (124).

In some patients with paresis of upward gaze, Bell's phenomenon may be helpful in differentiating an infranuclear from a supranuclear lesion. Bell's phenomenon consists of outward and upward rolling of the eyes when forcible efforts are made to close the eyelids against resistance. It does not occur with blinks, and it is observed in only 50% of individuals during voluntary unrestrained lid closure (125). The presence of this movement in individuals who cannot voluntarily elevate their eyes usually indicates that brain stem pathways between the facial nerve nucleus and that portion of the oculomotor nucleus responsible for ocular elevation are intact, and thus that an upward gaze paresis is supranuclear in origin (126). However, an intact Bell's phenomenon may occur in patients with Guillain-Barré syndrome and was also demonstrated in a patient with complete ophthalmoplegia caused by myasthenia gravis (127,128). Absence of a Bell's phenomenon has less diagnostic usefulness, since about 10% of normal subjects do not have this fascio-ocular movement (129,130). A downward Bell's response is present in up to 8% of individuals (130).

OCULAR ALIGNMENT

Principles

When the eyes are not aligned on the same object, *strabismus* is present. The strabismus may be congenital or acquired and may be caused by central or peripheral dysfunction. In some individuals, particularly those with isolated congenital strabismus, the amount of ocular misalignment is unchanged regardless of the direction of gaze or of which eye is fixating the target. This type of strabismus is termed *comitant* or *concomitant*. On the other hand, when the amount of an ocular deviation changes in various directions of gaze, with either eye fixing, or both, the strabismus is said to be *incomitant* or *noncomitant*. Congenital comitant strabismus is occasionally associated with other neurologic dysfunction (131–134), and acquired comitant strabismus may rarely occur as a sign of intracranial disease (135–147). Most cases of acquired comitant strabismus appear in otherwise normal children and adults, as well as in persons with neurologic or systemic disease, from decompensation of a pre-existing phoria, or as a result of latent hypermetropia (3,148,149). Thus, most instances of neuropathic or myopathic strabismus are of the incomitant variety. The reasons for this incomitance are described in the next section.

Primary and Secondary Deviations

Any patient with a manifest deviation of one eye (heterotropia) will fixate a target with only one eye at a time. During viewing with one eye, the visual axis of the opposite (nonfixing) eye will be deviated a certain amount away from the target. Patients with a comitant strabismus have the same amount of deviation of the nonfixing eye regardless of the eye that is fixing or the field of gaze. Most patients with incomitant (and especially paralytic) strabismus tend to fix with the nonparetic eye if visual acuity is equal in the two eyes. In these patients, the deviation of the nonfixing eye is called the primary deviation. When such patients are forced to fix the same target with the paretic eye, the deviation that results, the secondary deviation, is always greater than the primary deviation (Fig. 18.5). The explanation for this phenomenon is related to the position of the eyes within the

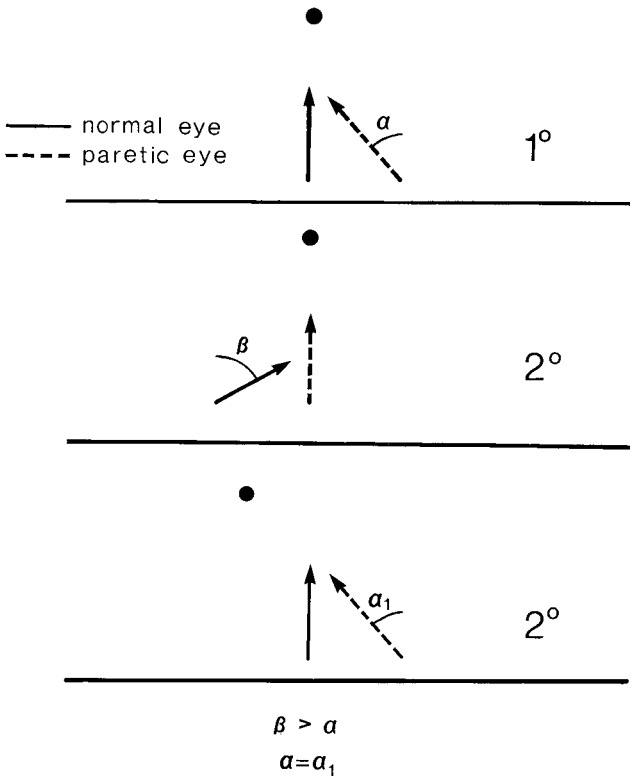

Figure 18.5. The principle of primary and secondary deviations. *Top,* When the normal eye fixes on a target directly ahead, the paretic eye deviates from the primary position by a certain amount (α). This is called the primary deviation. *Middle,* When the paretic eye fixes on a target in primary position, the normal eye also deviates from primary position by a certain amount (β), but this secondary deviation of the normal eye when the paretic eye is fixing is greater than the amount of deviation of the paretic eye when the normal eye is fixing ($\beta > \alpha$). *Bottom,* Although the common explanation of primary and secondary deviation is based on Hering's law of equal innervation to yoke muscles, Leigh and Zee (1991) suggested that the secondary deviation is greater than the primary deviation in paretic strabismus because when the paretic eye is fixing in primary position, it is forced further into its field of limitation. If the paretic eye were fixing on an object in the opposite direction, the deviation of the eye from primary position ($\alpha 1$) would be the same as if the normal eye were fixing on an object straight ahead ($\alpha = \alpha 1$). Thus, although Hering's law is maintained, the explanation for primary and secondary deviations is based upon the position of the eyes within the orbit and not upon which eye is fixing.

orbits (2). When a single muscle is paretic, the deviation between the two eyes is proportional to the difference between the forces generated by the paretic muscle and its normal yoke muscle. Furthermore, the amount of force contributed by each muscle toward holding the eye in a specific orbital position increases as the eye is moved into the direction of action of that muscle. Under normal circumstances, this force, thus obeying Hering's law, is equal for yoke pairs of muscles. However, as the eyes move into the direction of action of the paretic muscle, the difference in forces generated by the normal and paretic yoke muscles increases, thus increasing the deviation between the two eyes. When this change in deviation is tested as a function of orbital position, it is actually found to be independent of which eye is fixing. Thus, when the paretic eye is fixating a target, it is held in an orbital position further in the direction of action of the paretic muscle than when the nonparetic eye is fixating the same target. This results in a secondary deviation that is greater than the primary deviation simply because of the change in eye position toward the direction of action of the paretic muscle when the paretic eye is forced to take up fixation. The innervation to both muscles in the yoke pair is increased in that direction as predicted by Hering's law, and this increased innervation is more effective in the nonparetic muscle. However, the innervation is not increased any more than if the nonparetic eye is forced to take up fixation in eccentric gaze to achieve the same final orbital position.

Past-pointing and Disturbances of Egocentric Localization

Von Graefe first described anomalies of spatial localization that he referred to as *past-pointing* or *false orientation* in patients with paralytic strabismus (150). If a patient is asked to point at an object in the field of action of a paretic muscle while the paretic eye is fixating, the patient's finger will point beyond the object *toward* the field of action of the paretic muscle (Fig. 18.6). During this test, it is important that the hand be covered or that the patient point rapidly toward the object so as to avoid visual correction of the error of localization while the hand is still moving toward the object. Patients with accommodative esotropia exhibit a sim-

Figure 18.6. Past-pointing in a patient with a right sixth nerve paresis. *A,* In primary position, there is only a slight amount of past-pointing. *B,* In right gaze, however, the amount of past-pointing increases. The *white arrows* indicate the amount of past-pointing (the difference between the actual target location and the area in space to which the patient points).

ilar phenomenon while they have a manifest esotropia, pointing in the direction that the nonfixating eye is looking (151).

The explanation of past-pointing is controversial. One explanation is that the image of the target lies in an abnormal location relative to the fovea so that the patient incorrectly localizes the object into that field. This explanation, however, does not account for the past-pointing that is observed when the image of the target lies on the fovea of the paretic eye (152–154). Clearly nonvisual information about eye position is involved in spatial localization, and an argument has raged over the relative role of efferent command to the eye muscles versus proprioceptive afferent information from the eye muscles. In support of the efferent command theory, Helmholtz argued that past-pointing depends on the ''intensity of the effort of will'' that is sent to the paretic muscle (155). For a thorough description of the evidence for proprioceptive feedback from the muscles in spatial localization, see the review by Weir (156).

Head Turns and Tilts

Patients with strabismus commonly turn or tilt the head to minimize diplopia. Head turns are frequently associated with paresis of the horizontal extraocular muscles. Similarly, patients with vertical extraocular muscle paresis may carry their head flexed or extended. Most patients with such head turns adopt the particular posture to minimize or eliminate diplopia by moving the eyes away from the field of action of the paretic muscle; however, some patients adopt a head posture that actually increases the distance between the two images, allowing one of the images to be more easily ignored.

Head turns also occur in patients with congenital nystagmus. In such patients, keeping the eyes in an eccentric (null) position in the orbit by means of the head turn may result in reduction in the amplitude or frequency of the nystagmus.

Head tilts are most commonly observed with paresis of the oblique muscles, particularly the superior oblique. With an acquired superior oblique palsy, for example, the face is usually turned away from the paretic eye, the chin is down slightly, and the head is tilted toward the side opposite the paretic muscle. This permits fusion of images. Patients with congenital superior oblique palsy may adopt a similar head tilt or one in the opposite direction (i.e., toward the side of the paretic muscle) to more widely separate the images. Head tilts that occur from ocular causes often must be differentiated from nonocular torticollis.

Finally, some patients develop head turns that seem to be caused by central visual field defects and not by ocular misalignment (157). Such patients turn their heads toward the hemianopic field under both monocular and binocular conditions. The explanation for the head turn in these patients is unclear.

Techniques

Ocular alignment may be tested subjectively or objectively, depending on the circumstances under which the examination is performed and the physical and mental state of the patient.

Subjective Testing

When a patient is cooperative, subjective testing of diplopia reliably indicates the disparity between retinal images. The simplest subjective tests of ocular alignment use colored filters to dissociate the deviation and to emphasize and differentiate the images so that the patient and the observer can interpret them. A fixation light is used to provide the image. A red filter held over one eye suffices in most cases. However, the addition of a green filter over the opposite eye gives better results in children, in patients with a tendency to suppress or ignore one of the images, and in patients with a slight paresis and good fusion ability who can overcome their deviation. The use of complementary colored filters, one over each eye, produces maximum dissociation of images, since there is no part of the visible spectrum common to both eyes.

The red filter is always placed over the patient's right eye, and all questions posed to the patient relate to the relationship of the red image with respect to the white (or green) image. The patient is first asked if he or she sees one or two lights. If the patient sees two lights, he or she is then asked what color they are. After the appropriate answer, the patient is asked if the red light is to the right or left of the other light and if it is above or below the other light. The information thus received will be that the patient sees two lights, one red and one white or green, that they are crossed (each image is perceived on the side opposite the eye that is seeing it) or uncrossed (each image is perceived on the same side as the eye that is seeing it), and that the image relating to the right eye (red image) is higher or lower than the other image. The image of an object is always displaced in the *opposite* direction to the position of the eye (Fig. 18.7). Thus, if an eye is exotropic, the patient will have *crossed* diplopia,

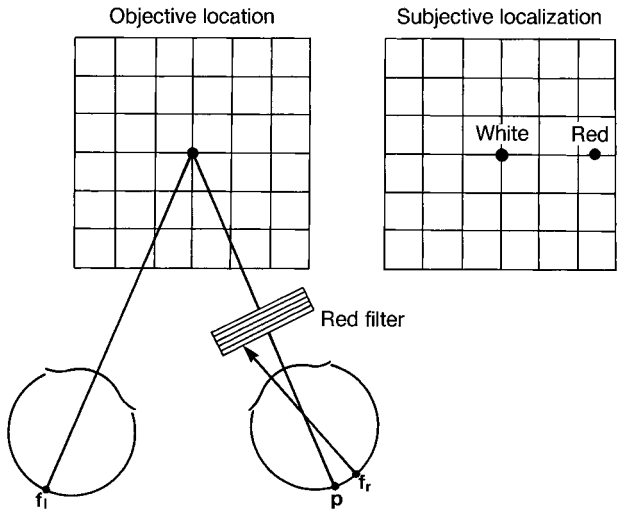

Figure 18.7. The principle of diplopia tests. A red filter is placed in front of the right eye, and the patient fixes a single light in the distance. If the eyes are misaligned, the light is imaged on the fovea of one eye (fl) and the nonfoveal retina (p) of the opposite eye. The patient thus sees two images, white and red, in different locations in space.

and (with a red filter over the right eye) the patient will see the red image to the *left* of the other image. Similarly, if the patient has an esotropia, the red image will be seen to the *right* of the other image (*uncrossed* diplopia). If the patient has a vertical deviation of the eyes, the eye that is higher will see the image of an object *below* that of the opposite eye.

Once the patient indicates that there is a clear separation of images when he or she is fixing on a light held straight ahead, the examiner can determine the area of maximum vertical separation, horizontal separation, or both, by having the patient look at a light held in the eight other cardinal positions of gaze (right, upper right, up, upper left, left, lower left, down, lower right).

In addition to the use of filters placed over one or both eyes, one can place a red Maddox rod over one eye and have the patient fixate on a white light (158). The ''rod'' is, in fact, a set of small half-cylinders aligned side by side in a frame in such a way that when the eye views a light through the cylinders, the image seen is that of a line perpendicular to the cylinder axis. Thus, if one views a white light with one eye covered by a red Maddox rod, the images will be those of a red line and a white light. The Maddox rod can be placed in such a manner as to produce a vertical, horizontal, or oblique line. Individuals who are orthophoric will see the line pass through the light. When the rod is oriented to produce the image of a vertical line, patients with a horizontal strabismus will see the line to the left or right of the light. When the rod is oriented so that a horizontal line image is produced, patients with a vertical strabismus will see the line above or below the light.

Torsional misalignment of the eyes (e.g., superior oblique palsy) can be tested with two Maddox rods, one over each eye (159). This is best performed using a trial lens frame. If both rods are oriented so as to produce a horizontal line image, an eye with torsional dysfunction will see the line as oblique rather than horizontal. The patient is then asked to rotate the rod until the line is perceived as horizontal. By this method, the amount of torsion can be measured and followed. Traditionally, one clear (or white) Maddox rod and one red Maddox rod are used for this test. However, Simons et al. showed that the subjective torsion more reliably localizes to the paretic eye if two red Maddox rods are used and if the room is dark during testing (160).

The precise methods of testing and documenting positions of gaze have been described in detail by numerous authors and are not discussed further in this chapter (2,3,161).

In addition to the tests described above, other subjective techniques may be used in which two test objects rather than one are presented to the patient in such a way that each object is viewed with only one eye (Fig. 18.8). The patient is then required to place the two objects in such a fashion that they appear to be superimposed. The objects appear superimposed only when their images fall on the fovea of each eye. Misalignment of the foveas results in the patient placing the objects in different locations in space. The eyes are differentiated and dissociated in various ways. Each eye may be presented with a different target, or complementary colors may be placed into the visual field, either directly or by projection, with each eye being provided with a corresponding colored filter. These haploscopic tests include the use of a major amblyoscope and the Hess screen and Lancaster red-green tests (162,163).

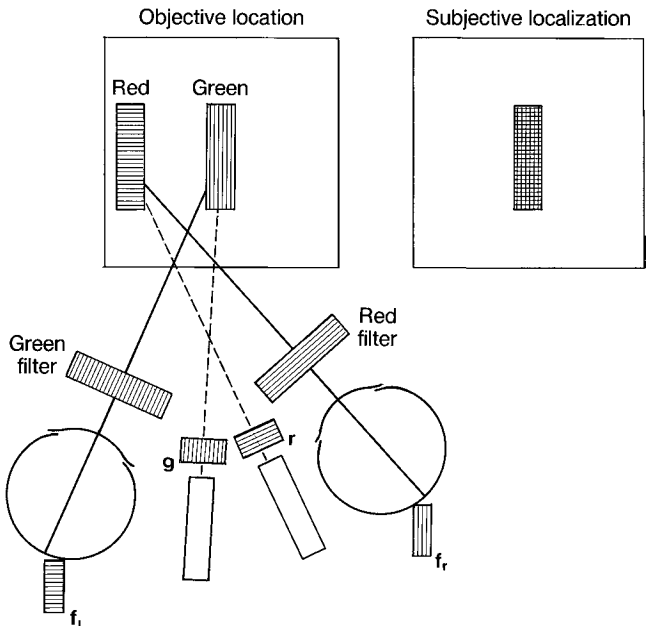

Figure 18.8. The principle of haploscopic tests. Red and green test objects are used, and the patient has a red filter placed in front of one eye and a green filter in front of the other eye.

In general, the Hess test uses a gray or black screen marked with a tangent scale on which red targets are projected or positioned where the tangent lines cross. A green target or light is superimposed subjectively on the red fixation target. Complementary red and green filters are worn to permit (and stimulate) binocular dissociation, thus revealing the ocular deviation in each position of fixation. The test is performed with the patient 0.5 meters from the screen.

The Lancaster test incorporates the same principles as the Hess screen but uses a two-dimensional grid rather than a tangent screen and is performed with the patient 1 or 2 meters from the grid.

Although the Hess and Lancaster tests use red and green colors to dissociate images, as in the more simple ''red glass'' test described above, they differ from that test in principle. In the red glass test, one white fixation light is used that can be seen by each eye through the red and green (if used) filters. In the Hess or Lancaster tests, instead of a white light, two different-colored test objects are used. These are projected red and green lines that can be perceived only by the fovea of the eye that is viewing through the corresponding colored filter. Thus, the eye that is viewing through the red filter (usually the right eye) sees only the red line, whereas the eye that is viewing through the green filter (usually the left eye) sees only the green line. Each fovea perceives the image from its respective target as being located straight ahead. This is macular-macular projection (confu-

sion). The binocular dissociation produced in this manner is sufficient to reveal even a minimal and well-controlled heterophoria. The patient, wearing the red filter over the right eye, is asked to superimpose the green line on the red line as each fixation position on the screen or grid is illuminated consecutively. The nonfixing left eye is deviated behind the green filter, but the patient merely guides the flashlight so that the green line is positioned where the deviated eye is pointing. At this point, the patient sees the red and green lights superimposed. The examiner, however, can see the deviation of the green line from the red line and can plot the position of the green line onto a chart. The test is then repeated with the red and green filters reversed, so that the left eye now fixates the red line, and the field of the deviating right eye is plotted. Not only is this test helpful in assessing the degree of deviation of both comitant and noncomitant strabismus at any given time, but also it may be used in patients with paralytic strabismus to detect subtle changes in ocular alignment that occur over time. It thus may help in the planning and evaluation of therapy for such patients (164). In addition, subjective torsion of the eyes can be detected when the patient rotates one line at an oblique angle relative to the other line. We believe that this test is the best way to subjectively determine the degree of ocular torsion present in one or both eyes. The interested reader should consult von Noorden (3), Burde (39), and Zee et al. (164) for excellent descriptions of these tests.

Objective Testing

The simplest objective method of determining ocular alignment is the use of a hand light to cast a reflection on the corneal surfaces of both eyes in the cardinal positions of gaze. If the images from the two corneas appear centered, then the visual axes are often correctly aligned. If the light reflexes are not centered, one can either estimate the amount of misalignment based on the apparent amount of decentration of the light reflex (with the fixation light held 33 cm from the patient, 1 mm of decentration equals 7° of ocular deviation) (165), or prisms can be placed over either of the eyes until the light reflexes appear centered. Krimsky recommended that the prisms be placed over the nonfixing eye (166), but von Noorden considered this technique less precise and more difficult than placing prisms over the fixing eye (3), and we agree; however, if the nonfixing eye is so eccentric and limited in its excursion that centration of the light reflex is impossible or requires excessive prism over the fixing eye, holding the prism over the fixing eye results in a measurement of the deviation only in eccentric gaze. As noted above, this is essentially the same as measuring the secondary, rather than the primary, deviation.

A number of conditions other than a heterotropia may cause decentration of the corneal light reflex and must be considered to interpret tests correctly based on centration of the light reflex. The angle kappa is defined as the angle between the visual line (line connecting the point of fixation with the fovea) and the pupillary axis (line through the center of the pupil perpendicular to the cornea). The angle is measured at the center of the pupil. It is called positive when the light reflex is displaced nasally and negative when the light reflex is displaced temporally. A positive angle kappa may simulate an exodeviation (Fig. 18.9), and a negative angle kappa may simulate an esodeviation. Conversely, strabismus may be less apparent when a large angle kappa is associated with esotropia or a large negative angle is associated with exotropia. Other ocular abnormalities that produce decentration of the corneal light reflex include eccentric fixation and ectopic macula (e.g., in patients with retinopathy of prematurity or other retinal disease with macular traction).

The most precise objective methods of measuring ocular alignment are the cover tests (153,167,168). Although these tests require that the patient be able to fixate a target with either eye, they generally require less cooperation than do

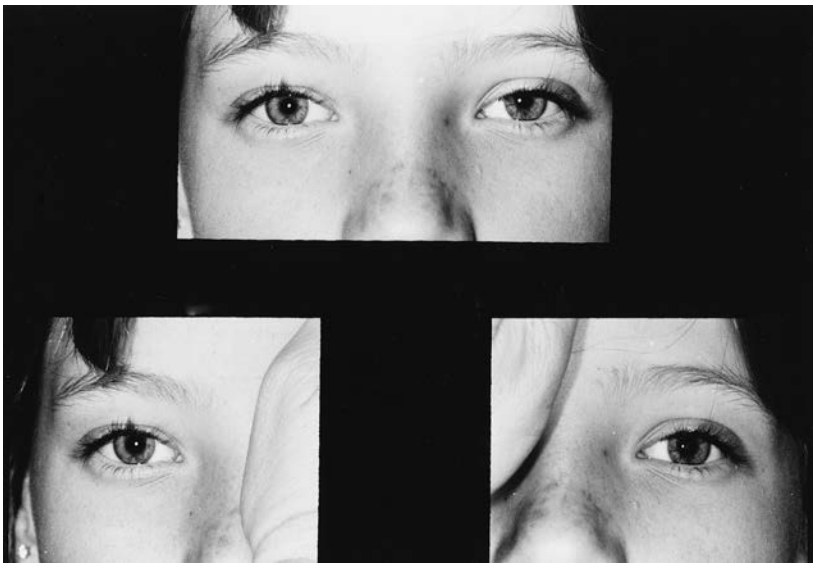

Figure 18.9. Positive angle kappa with the corneal light reflex test. With both eyes open (*top*), the eyes appear exotropic because the light reflection is decentered nasally in the left eye. The reflex remains centered in the right eye when the left eye is covered (*bottom left*). When the right eye is covered (*bottom right*), there is no shift of fixation, and the light reflection remains nasally displaced.

the subjective tests described above. The three types of cover tests used by most clinicians are the single cover test, the cover/uncover test, and the alternate cover (cross-cover) test.

In the single cover test, the patient fixates an accommodative target at 33 cm (near target) or 6 meters (distant target). An opaque occluder is placed in front of one eye, and the examiner observes the opposite eye to see whether it moves to take up fixation of the target (Fig. 18.10). If movement is observed, its direction and speed should be noted. The test is then repeated on the opposite eye. If the patient has a manifest ocular deviation (heterotropia), the previously nonfixing eye will be observed to change position to take up fixation when the fixing eye is covered. On the other hand, when the nonfixing eye is covered, no movement of the fixing eye will be observed (since it is already fixing on the target). This test is usually performed with the patient fixing in primary position and with the eyes in the other cardinal positions of gaze. In our experience, movements of as little as 1° can be easily observed.

In the cover/uncover test, the patient fixates on an accommodative target, and one eye is occluded. The behavior of that eye is then observed as the cover is removed (Fig. 18.10). The direction of any deviation and the speed and rate of recovery to binocular fixation are noted. If no movement of the uncovered eye is observed when the cover/uncover test is performed, an alternate cover test may be used to detect a latent deviation of the eyes (heterophoria). Instead of occluding one eye and then taking the occluder away, first one eye and then the other are alternately occluded. The cover should remain in front of each eye long enough to allow the patient to take up fixation with the uncovered eye. This test prevents fusion and dissociates the visual axes. Any movement of either uncovered eye suggests that although the eyes are straight during binocular viewing, loss of fusion (i.e., by the alternate occlusion of the two eyes) results in a deviation of whichever eye is covered (Fig. 18.11).

The importance of distinguishing between heterophorias and heterotropias cannot be overemphasized, since patients with heterophorias have binocular central fusion, whereas patients with heterotropias do not.

If either the cover/uncover or alternate cover test detects evidence of ocular misalignment, prisms can be used to neutralize the movement and thereby measure the deviation, whether it is a heterotropia or heterophoria. Prisms are placed in front of either eye such that the apex of the prism is oriented in the direction of the deviation, and the prism strength is altered until the deviation is no longer observed. The technique for measuring the amount of strabismus with ophthalmic prisms was beautifully described by Thompson and Guyton, and their manuscript should be reviewed by the physician or technician planning to perform such measurements (169).

When there is a vertical ocular deviation, it is often helpful to perform cover tests with the head tilted first toward one side and then toward the other (170,171). The patient is instructed to maintain fixation on a distant target, and the position of the eyes relative to each other is measured. The patient is then instructed to tilt the head to one side while maintaining fixation on the target, and the eyes are again

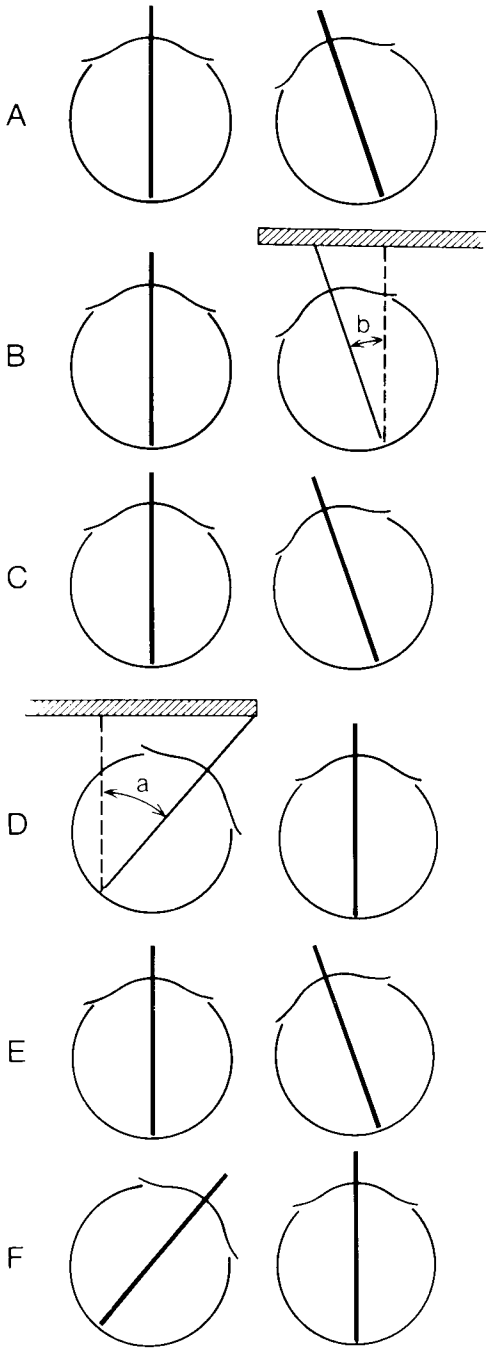

Figure 18.10. The single cover and cover/uncover tests. In both tests, one eye is covered at a time; however, in the single cover test, one eye is covered and the opposite eye is observed. In the cover/uncover test, one eye is covered and the behavior of that eye is observed when the cover is removed. *A,* Initially, with both eyes viewing, the left eye is fixating the target and the right eye is esotropic. When the right eye is covered (*B*), no movement of the left (uncovered) eye is observed, nor is any movement of the right eye observed when the cover is removed (*C*). *D,* When the left eye is covered, the right eye moves outward to take up fixation. The deviation of the normal eye under cover (the secondary deviation, a) is greater than that of the paretic eye under cover (primary deviation, b). When the cover is removed, either the left eye again takes up fixation (E) or the paretic right eye continues to fixate (F).

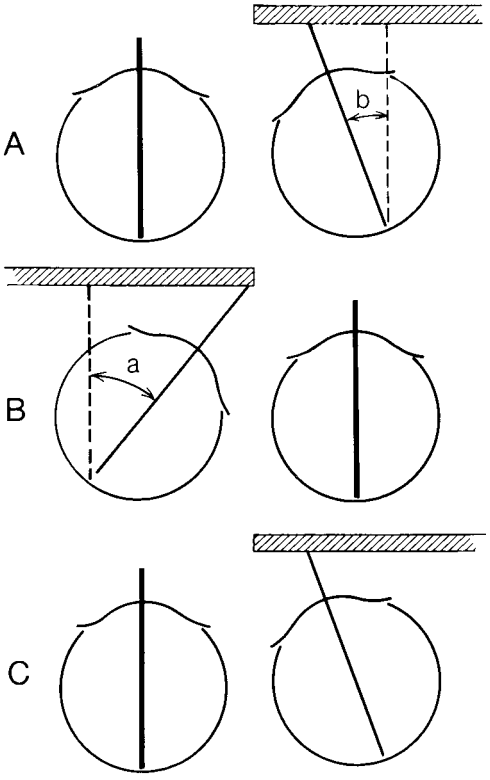

Figure 18.11. The alternate cover test. This test prevents fusional vergence and thus tests both phorias and tropias but does not differentiate between them. In this test, the cover is quickly moved from one eye to the other, and any movement of either eye is noted. In this example, there is an esodeviation.

examined to see whether one eye has moved higher than the other. Measurements are made with the head tilted to each side.

This test is useful primarily in the diagnosis of superior oblique palsy, since patients with this disorder consistently show a hypertropia of the damaged eye when the head is tilted toward the affected side (159). It has been suggested that the physiologic basis of this test lies in the fact that the vertical eye muscles are not driven in their usual yoked pairs when the head is tilted. Instead, the vestibular apparatus produces compensatory torsion of the eyes by co-innervation of the ipsilateral (to the side of the tilt) superior oblique and superior rectus muscles, producing intorsion, and of the contralateral inferior oblique and inferior rectus muscles, producing extorsion. When a patient with a trochlear nerve palsy tilts the head toward the affected side, intorsion of that eye should occur to keep the vertical meridian perpendicular to the horizon. As noted above, this intorsion is usually about 10° and is produced by the otolith-ocular reflex, resulting in synergistic contractions of the superior rectus and superior oblique muscles. If, however, the superior oblique muscle is paretic, its secondary actions, one of which is depression, are also impaired. The superior rectus muscle is therefore the only means by which the eye is intorted, and its main action, elevation of the eye, is unopposed. This mechanism is contested by authors who find no relationship between the amount of hypertropia and the amount of countertorsion with the head tilt test (172).

The head tilt test is best used as part of a three-step diagnostic test first described by Haagedorn (173) and popularized by Parks (174) and Hardesty (175). This test is used to isolate the affected paretic muscle in concomitant or incomitant vertical deviations. It is useful only if the paresis is of a single cyclovertical muscle (176). The steps are as follows:

1. The presence of a vertical heterotropia is determined in primary position. Depending on the eye that is hypertropic, one of four muscles may be paretic: the ipsilateral inferior rectus, the ipsilateral superior oblique, the contralateral superior rectus, or the contralateral inferior oblique. Thus, if the patient has a right hypertropia, the muscles that may be paretic are the right superior oblique, the right inferior rectus, the left superior rectus, or the left inferior oblique.
2. Whether the hypertropia increases in right or left horizontal gaze is determined. This reduces the potential paretic muscles to two. Thus, in a patient with a right hypertropia, if the deviation increases in right gaze, the affected muscles can only be the right inferior rectus (which has its maximum vertical action when the eye is in an abducted position) or the left inferior oblique (which has its maximum vertical action when the eye is in an adducted position). If the deviation increases in left gaze, the affected muscles could be either the right superior oblique or the left superior rectus.
3. The differential diagnosis between the two muscles, one in each eye, that are potentially responsible for the vertical heterotropia, is now made using the head tilt test as described above. Thus, if the patient has a right hypertropia that increases in left gaze, an increase in the hypertropia when the head is tilted to the right side indicates a paretic superior oblique muscle.

Although we believe the three-step test is extremely useful in patients with presumed trochlear nerve palsies (159,177), we and others find that its reliability in the diagnosis of pareses of other vertical muscles is questionable (178). Furthermore, both restrictive ophthalmoplegia and myasthenia gravis can mimic superior oblique palsy using the three-step test (179).

A final objective method of determining ocular torsion is direct observation of the ocular fundus (180–182). The normal fovea is generally located about 7° below the center of the optic disc (range 0–16°) or along a horizontal line originating from a point between the middle and lower thirds of the optic disc (183). When an eye is extorted, the foveal reflex rotates below this plane, whereas intorsion results in upward rotation of the reflex (Fig. 18.12). The amount of torsion can be determined using a variety of ophthalmoscopic and photographic methods (180–186).

PERFORMANCE OF VERSIONS

Versions may be tested by examining the saccadic, pursuit, vestibular, and optokinetic systems.

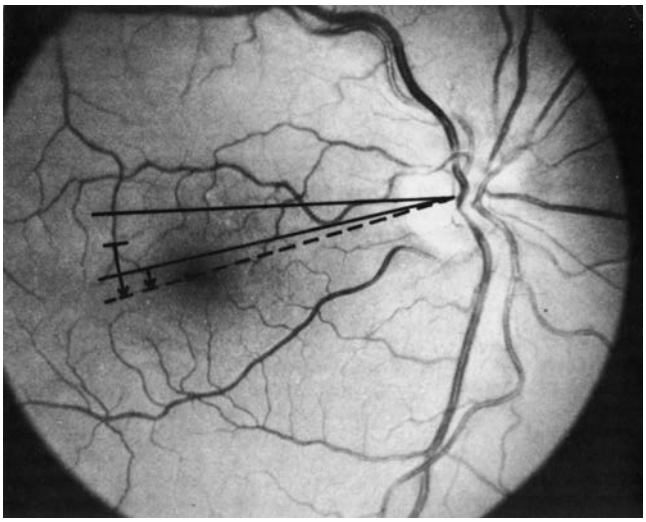

Figure 18.12. Torsion of the ocular fundus. Photograph of an extorted right fundus, direct view, with a *dashed line* drawn from the center of the optic disc through the fovea. The normal angular range of the fovea from the center of the disc is shown by the two *solid lines.* The amount of extorsion can be measured in degrees from the center of the normal range (*long arrow*) or from the limit of the normal range (*short arrow*). (Courtesy of Dr. David L Guyton.)

Clinical Examination of Saccades

Saccadic eye movements are examined clinically by instructing the patient to alternately fixate upon two targets, usually the examiner's finger and nose. Saccades in each direction can be examined in each field of gaze in both the horizontal and vertical planes. The examiner must determine whether the saccades are promptly initiated, of normal velocity, and accurate.

Saccadic latencies can be appreciated by noting the time it takes the patient to initiate the saccade. Abnormal voluntary saccadic velocities may be accentuated by using a drum or hand-held tape with repetitive patterns. Rotating the drum or passing the tape horizontally and vertically across the patient's visual field stimulates the optokinetic system to produce nystagmus (see Chapter 17). Abnormal saccadic velocities, particularly slowing, thus become more evident when the patient is forced to make multiple, repetitive saccades to refixate the passing targets. Altered saccadic velocities that occur in only one plane of movement can also be appreciated by using obliquely placed targets to stimulate oblique saccades. In such patients, the normal-velocity component of the saccade (e.g., the horizontal) will be completed before the other component (e.g., the vertical), so that the trajectory appears L-shaped.

Disorders of saccadic accuracy (e.g., saccadic dysmetria) can be inferred from the direction and size of corrective saccades that the patient must make to ultimately acquire the fixation target. Since saccades as small as ½° can be easily identified during clinical observation, minimal degrees of saccadic dysmetria can be easily appreciated during the clinical examination. Normal individuals may undershoot a target by a few degrees when refixations are large. Similarly, such individuals may overshoot a target during centripetal, and especially downward, saccades. This dysmetria is transient, gradually disappearing during repetitive refixations between the same targets.

When a saccadic abnormality is detected during the clinical examination, the examiner must attempt to localize the disturbance within the hierarchical organization of the saccadic eye movement system (see Chapters 17 and 19). The first step in localization is to establish whether the disease process affects reflexive saccades. Quick phases can be examined by spinning the patient in a swivel chair to elicit vestibular and optokinetic nystagmus. Next, an attempt should be made to determine whether saccades can be performed without visual targets or in response to auditory targets, by asking the patient to refixate under closed lids. The eye movements thus generated can be observed, palpated, and even heard (with a stethoscope applied to the lids).

During the evaluation of saccadic eye movements, it is often helpful to observe gaze changes when the patient makes a combined eye/head movement to see whether an accompanying head movement can facilitate the production of a saccade. This strategy is used by some patients with ocular motor apraxia (see Chapter 19). The effect of blinks should also be noted, since they may facilitate both the ability to initiate saccades and the subsequent saccadic velocity. Finally, asking the patient to repetitively refixate between two targets may reveal fatigue of saccadic eye movements.

Clinical Examination of Pursuit

Patients with isolated deficiency of smooth pursuit do not usually complain of visual symptoms, since they can track moving objects with a series of saccades. Only very demanding tracking tasks (e.g., playing tennis, handball, or baseball) may cause patients with impaired pursuit to report difficulties. The vision of normal subjects deteriorates during tracking of targets moving at high frequencies, however, so that even complaints of inability to track fast-moving objects may not signify a disorder of smooth pursuit (187).

To test the pursuit system, the patient should be asked to track a small target, such as a pencil tip held a meter or more from the eyes, with the head held still. The target should initially be moved at a low, uniform velocity. Pursuit movements that do not match the target velocity result in corrective saccades. If these are ''catch-up'' saccades, then the pursuit gain is low. If pursuit gain is too high, then ''back-up'' saccades are observed. Small children, uncooperative patients, or individuals thought to have nonorganic blindness may be tested with a slowly rotating large mirror held before their eyes.

Although hand-held drums or tapes with repetitive targets do not truly test the optokinetic system (see Chapter 17), they do stimulate pursuit and may be useful in the detection of pursuit asymmetries and other abnormalities of the pursuit system.

The VOR generates eye movements that compensate for angular displacement of the head and maintain the visual axes ''on target.'' If one observes a slowly moving target

by moving the head, so that the target remains stationary relative to the head, the eye movements generated by the VOR are inappropriate and must be suppressed. The ability of a patient to suppress (or cancel) the VOR can be evaluated by using a central fixation target that moves in the same direction and at the same velocity as the head (37,188). Patients often do this best by fixating on their thumbnail with their arm outstretched while being rotated in the examination chair. Those who have limb muscle weakness can be rotated in a wheelchair while fixating a target that rotates with the chair (189). When suppression of the VOR and pursuit are compared in normal human subjects, similar frequency response curves are obtained, leading to the hypothesis that suppression of the VOR depends directly on information derived from the smooth pursuit system (190,191). This hypothesis is supported by the clinical observation that patients with impaired smooth pursuit also have abnormal suppression of the VOR (192–194). The evaluation of VOR suppression is thus another way to test the integrity of the pursuit system. Deficits in VOR suppression, however, are nonlocalizing, since they may occur with either cerebral or cerebellar disease (195–197).

In some patients, it is difficult to test smooth pursuit because of spontaneous nystagmus; however, in some of these patients, the nystagmus is less prominent in a specific position of gaze (the null point; see Chapter 23). In these patients, cancellation of the VOR during head rotation can be tested with the eyes fixing on a target in this position. As with pursuit, the head rotation should be gentle at first. In patients with inadequate cancellation, the eyes are continually taken off target by the intact VOR, and corrective saccades therefore occur. An asymmetric deficit may imply a pursuit imbalance provided that the VOR is intact and symmetric: deficient cancellation of the VOR on rotation to one side corresponds to a low pursuit gain to that side. Furthermore, when there is a discrepancy between the performance of smooth pursuit and cancellation of the VOR (e.g., poor pursuit but good cancellation), one should suspect an inadequate or asymmetric VOR.

Clinical Examination of the Vestibular and Optokinetic Systems

We previously described the clinical methods used to test the vestibular system (oculocephalic and caloric testing), and we will not repeat them here. Although the optokinetic system can be tested in the laboratory (discussed later), the system cannot be tested as part of a routine clinical examination.

QUANTITATIVE ANALYSIS OF EYE MOVEMENTS

Voluntary Eye Movements

Most disturbances of ocular motility and alignment can be detected during a standard clinical examination; however, performing a quantitative analysis of eye movements may more accurately reveal subtle abnormalities of the pursuit, saccadic, optokinetic, and vestibulo-ocular systems. The most common methods used to record eye movements are electro-oculography and infrared oculography (198–207). These techniques may be used to distinguish myopathic (restrictive) from neuropathic conditions that affect ocular motility and to determine the presence or absence of improvement of ocular motor function (208–212). The value of vertical saccadic velocity or amplitude determinations in adduction versus abduction to identify and monitor superior oblique muscle dysfunction remains controversial (213–215).

Although electro-oculography can yield reasonable recordings of horizontal eye movements, vertical measurements with this technique are affected by eyelid artifacts and nonlinearities (206). Changes in illumination and skin resistance also affect the readings with this method. Infrared oculography provides higher-resolution measurements of both horizontal and vertical eye movements, but over a limited range, especially vertically. In addition, the signal is lost when the eyes are closed. Finally, neither electro-oculography nor infrared oculography measures ocular torsion.

The magnetic field-search coil method, which uses coils embedded in a silicone rubber ring that adheres to the sclera by suction, overcomes most of the problems that limit both electro-oculography and infrared oculography (62,216). Further advances in this system using a digital microprocessor enable this technique to be used with great accuracy to measure virtually all types of normal and abnormal eye movements (206,207).

Vestibulo-Ocular Reflex

As explained above, the oculocephalic and caloric tests that are performed in patients with apparent limitation of eye movement primarily test the function of the semicircular canals of the vestibular system. More extensive testing is usually directed toward determining gain, phase, and balance. Rotation tests give more accurate and reproducible results than do caloric tests, although the mental state of the patient while in darkness may influence the results (217). The gain of the VOR may be obtained by measuring the peak eye velocity in response to a velocity step (e.g., sudden sustained rotation at 60°/sec) in darkness. This is usually done in vestibular laboratories equipped with servo-controlled chairs and eye monitoring equipment, although portable systems for this purpose are available (217,218).

Although the otolith organs respond to linear acceleration, tests of their function are rarely performed. Several investigators have suggested that muscle responses that occur less than 100 ms after release into free-fall are part of a normal startle reflex that originates in the otoliths (219–221). Using this theory, Halmagyi and Gresty developed a technique for measuring otolith integrity by recording eye blink reflexes that occur following sudden free-falls (222). Such testing may confirm the presence of otolith function in patients with impaired semicircular canal function or show impaired otolith function in patients with normal semicircular canal function.

Optokinetic System

The hand-held ''optokinetic'' drums or tapes that are used to elicit smooth movements primarily test the pursuit system.

True optokinetic testing requires a stimulus that fills the field of vision. A common technique is to have the patient sit inside a large, patterned optokinetic drum that is rotated around the patient. A true optokinetic stimulus induces a sensation of self-rotation (223). Another method of eliciting a true optokinetic response is rotation of an individual at a constant velocity in the light for over 1 minute. The sustained nystagmus that results is caused by purely visual stimuli (the vestibular response having died away); however, it is still difficult to separate its pursuit and optokinetic components.

REFERENCES

1. Nelson LB, Catalano RA. Atlas of Ocular Motility. Philadelphia: WB Saunders, 1989.
2. Leigh RJ, Zee DS. The Neurology of Eye Movements. Ed 3. Philadelphia: FA Davis, 1999.
3. von Noorden GK. Binocular Vision and Ocular Motility. Ed 5. St Louis: CV Mosby, 1996.
4. Burgess D, Roper-Hall G, Burde RM. Binocular diplopia associated with subretinal neovascular membranes. Arch Ophthalmol 1980;98:311–317.
5. Fincham EF. Monocular diplopia. Br J Ophthalmol 1963;47:705–712.
6. Ho PC, Elliott JH, O'Day DM. Multirefractile cataract. Invest Ophthalmol Vis Sci (Suppl) 1981;20:50.
7. Amos JF. Diagnosis and management of monocular diplopia. J Am Optom Assoc 1982;53:101–115.
8. Hirst LW, Miller NR, Johnson RT. Monocular polyopia. Arch Neurol 1983;40: 756–757.
9. Coffeen P, Guyton DL. Monocular diplopia accompanying ordinary refractive errors. Am J Ophthalmol 1988;105:451–459.
10. Woods RL, Bradley A, Atchison DA. Monocular diplopia caused by ocular aberrations and hyperopic defocus. Vision Res 1996;36:3597–3606.
11. Bender MB. Polyopia and monocular diplopia of cerebral origin. Arch Neurol 1945;54:323–338.
12. Kinsbourne M, Warrington EK. A study of visual perseveration. J Neurol Neurosurg Psychiatr 1963;26:468–475.
13. Meadows JC. Observations on a case of monocular diplopia of cerebral origin. J Neurol Sci 1973;18:249–253.
14. Safran AB, Kline LB, Glaser JS, et al. Television-induced formed visual hallucinations and cerebral diplopia. Br J Ophthalmol 1981;65:707–711.
15. Kasmann B, Ruprecht KW. Eclamptogenic Gerstmann's syndrome in combination with cortical agnosia and cortical diplopia. Germ J Ophthalmol 1995;4: 234–238.
16. Brindley GS, Lewin WS. The sensations produced by electrical stimulation of the visual cortex. J Physiol (Lond) 1968;196:479–493.
17. Cass EE. Monocular diplopia occurring in cases of squint. Br J Ophthalmol 1941;25:565–577.
18. Brandt T, Daroff RB. The multisensory physiological and pathological vertigo syndromes. Ann Neurol 1980;7:195–203.
19. Knight RT, St. John JN, Nakada T. Chewing oscillopsia: a case of voluntary visual illusions of movement. Arch Neurol 1984;41:95–96.
20. Bender MB. Oscillopsia. Arch Neurol 1965;13:204–213.
21. Gresty MA, Hess K, Leech J. Disorders of the vestibulo-ocular reflex producing oscillopsia and mechanisms compensating for loss of labyrinthine function. Brain 1977;100:693–716.
22. Ropper AH. Illusion of tilting of the visual environment: report of five cases. J Clin Neuroophthalmol 1983;3:147–151.
23. Steinman RM, Haddad GM, Skavenski AA, et al. Miniature eye movement: the pattern of saccades made by man during maintained fixation may be a refined but useless motor habit. Science 1973;181:810–819.
24. Winterson BJ, Collewijn H. Microsaccades during finely guided visuomotor tasks. Vis Res 1976;16:1387–1390.
25. Ciuffreda KJ, Kenyon RV, Stark L. Fixational eye movements in amblyopia and strabismus. J Am Optom Assoc 1979;50:1251.
26. Herishanu YO, Sharpe JA. Normal square wave jerks. Invest Ophthalmol Vis Sci 1981;20:268–272.
27. St Cyr GJ, Fender DH. The interplay of drifts and flicks in binocular fixation. Vision Res 1969;9:245–265.
28. Ditchburn RW. The function of small saccades. Vision Res 1980;20:271–272.
29. Kowler E, Steinman RM. Small saccades serve no useful purpose [reply to a letter]. Vision Res 1980;20:273–276.
30. Grut EH. A contribution to the pathogeny of concomitant squinting. Trans Ophthalmol Soc UK 1890;10:1–41.
31. Breinin GW. The position of rest during anesthesia and sleep. Arch Ophthalmol 1957;57:323–326.
32. Worth C, Chavasse B. Squint. 9th ed. London, Bailliere, Tindal, and Cox, 1959: 714.
33. Abraham SV. The basic position of rest: convergence and divergence. J Pediatr Ophthalmol 1964;1:9–24.
34. Jacobs L, Feldman M, Bender MB. Eye movements during sleep: I. The pattern in the normal human. Arch Neurol 1971;25:151–159.
35. Duke-Elder S, Wybar KC. Ocular motility and strabismus. In: Duke-Elder S, ed. System of Ophthalmology. Vol 6. St Louis, CV Mosby, 1973:96.
36. Hayashi S. Studies of the fixation movements of the eye ball. Jpn J Ophthalmol 1960;4:234–247.
37. Zee DS. Ophthalmoscopy in examination of patients with vestibular disorders. Ann Neurol 1978;3:373–374.
38. Scobee RG. The Oculorotary Muscles. 2nd ed. St Louis, CV Mosby, 1952.
39. Burde RM. The extraocular muscles: anatomy, physiology, and pharmacology. In: Moses RA, ed. Adler's Physiology of the Eye. 7th ed. St Louis, CV Mosby, 1981.
40. Park R, Park G. The center of ocular rotation in the horizontal plane. Am J Physiol 1933;104:545–552.
41. Verrijp C. Movements of the eyes. In: Behrens C, ed. The Eye and Its Diseases. Philadelphia, WB Saunders, 1949.
42. Fick A. Die Bewegungen des menschlichen Augapfels. Henle und Pfeufer, Ztschr 4:101, 1854.
43. Listing JB. In: Ruete CG, ed. Lehrbuch der Ophthalmologie für Aerzte and Studierende. 2nd ed, Vol 1. Braunschweig, Friedr Vieweg & Sohn GmbH, 1855; 37ff.
44. Donders FC. Beitrag zur lehre von den bewegungen des menschlichen auges. Holländ Beitr Anat Physiol Wiss 1848;1:104, 384.
45. Tweed D, Haslwanter T, Fetter M. Optimizing gaze control in three dimensions. Science 1998;281:1363–1366.
46. Cohen B. The vestibulo-ocular reflex arc. In: Kornhuber HH, ed. Handbook of Sensory Physiology. Vol 6. Berlin, Springer-Verlag, 1974:477–540.
47. Leigh RJ, Brandt T. A re-evaluation of the vestibulo-ocular reflex: new ideas of its purpose, properties, neural substrate, and disorders. Neurology 1993;43: 1288–1295.
48. Dieterich M, Brandt T. Vestibulo-ocular reflex. Curr Opin Neurol 1995;8:83–88.
49. Nelson JR, Cope D. The otoliths and the ocular countertorsion reflex. Arch Otolaryngol 1971;94:40–50.
50. Nelson JR, House WF. Ocular countertorsion as an indicator of otolith function: effects of unilateral vestibular lesions. Trans Am Acad Ophthalmol Otolaryngol 1971;75:1313–1321.
51. Woellner RC, Graybiel A. Counterrolling of the eyes and its dependence on the magnitude of gravitational or inertial force acting laterally on the body. J Appl Physiol 1959;14:632–634.
52. Uemura T, Cohen B. Effects of vestibular nuclei lesions on vestibulo-ocular reflexes and posture in monkeys. Acta Otolaryngol (Suppl) 1973;315:1–71.
53. Suzuki J, Cohen B, Bender MB. Compensatory eye movements induced by vertical semicircular canal stimulation. Exp Neurol 1964;9:137–160.
54. Diamond SG, Markham CH, Simpson NE, et al. Binocular counterrolling in humans during dynamic rotation. Acta Otolaryngol 1979;87:490–498.
55. Diamond SG, Markham CH. Binocular counterrolling in humans with unilateral labyrinthectomy and in normal controls. Ann NY Acad Sci 1981;374:69–79.
56. Diamond SG, Markham CH. Ocular counterrolling as an indicator of vestibular otolith function. Neurology 1983;33:1460–1469.
57. Jampel RS. The action of the superior oblique muscle. Arch Ophthalmol 1966; 75:535–544.
58. Jampel RS. The fundamental principle of the action of the oblique ocular muscles. Am J Ophthalmol 1970;69:623–638.
59. Jampel RS. Ocular torsion and the function of the vertical extraocular muscles. Am J Ophthalmol 1975;79:292–304.
60. Jampel RS. Ocular torsion and the law of the primary retinal meridians. In: Glaser JS, ed. Neuro-ophthalmology. Vol 10. St Louis, CV Mosby, 1980:201–216.
61. Jampel RS. Ocular torsion and the primary retinal meridians. Am J Ophthalmol 1981;91:14–24.
62. Collewijn H, Van der Steen J, Ferman L, et al. Human ocular counterroll: assessment of static and dynamic properties from electromagnetic scleral coil recordings. Exp Brain Res 1985;59:185–196.
63. Vogel R, Thümler R, von Baumgarten RJ. Ocular counterrolling. Some practical considerations of a new evaluation method for diagnostic purposes. Acta Otolaryngol (Stockh) 1986;102:457–462.
64. Krejcova H, Highstein S, Cohen B. Labyrinthine and extra-labyrinthine effects on ocular counter-rolling. Acta Otolaryngol 1971;72:165–171.
65. Linwong M, Herman SJ. Cycloduction of the eyes with head tilt. Arch Ophthalmol 1971;85:570–573.
66. Miller EF, Graybiel A. Effect of gravitoinertial force on ocular counter-rolling. J Appl Physiol 1971;35:697–700.
67. Horn G, Stechler G, Hill RM. Receptive fields of units in the visual cortex of the cat in the presence or absence of bodily tilt. Exp Brain Res 1972;15:113–132.
68. Brodsky MC. Do you really need your oblique muscles? Arch Ophthalmol 2002; 120:820–828.
69. Hering E. In: Bridgeman B, Stark L, eds. The Theory of Binocular Vision. New York, Plenum Press, 1977:129–145.
70. Boeder P. The cooperation of extraocular muscles. Am J Ophthalmol 1961;51: 469–481.

71. Mok D, Ro A, Cadera W, Crawford JD, Vilis T. Rotation of Listing's plane during vergences. Vision Res 1992;32:2055–2064.
72. Ferman L, Collewijn H, Van den Berg AV. A direct test of Listing's law: I. Human ocular torsion measured in static tertiary positions. Vision Res 1987;27: 929–938.
73. Ferman L, Collewijn H, Van den Berg AV. A direct test of Listing's law: II. Human ocular torsion measured under dynamic conditions. Vision Res 1987; 27:939–951.
74. Tweed D, Vilis T. Geometric relations of eye position and velocity vectors during saccades. Vis Res 1990;30:111–127.
75. Demer JL, Miller JM, Poukens V, et al. Evidence for fibromuscular pulleys of the recti extraocular muscles. Invest Ophthalmol Vis Sci 1995;36:1125–1136.
76. Kono R, Poukens V, Demer JL. Quantitative analysis of the structure of the human extraocular muscle pulley system. Invest Ophthalmol Vis Sci 2002;43: 2923–2932.
77. Demer JL, Oh SY, Clarke RA, Poukens V. Evidence for a pulley of the inferior oblique muscle. Invest Ophthalmol Vis Sci 2003;44:3856–3865.
78. Sherrington CS. Experimental note on two movements of the eyes. J Physiol (Lond) 1894;17:27.
79. Hering E. Die Lehre von Binocularen Sehen. Leipzig. Wilhelm Engelmann, 1868.
80. Chamberlain W. Restriction in upward gaze with advancing age. Am J Ophthalmol 1971;71:341–346.
81. Zappia RJ, Winkelman JZ, Gay AJ. Intraocular pressure changes in normal subjects and the adhesive muscle syndrome. Am J Ophthalmol 1971;71: 880–883.
82. Helveston E. The clinical applications of passive ductions and generated muscle force. Ophthalmology 1979;86:30.
83. Saunders RA, Helveston EM, Ellis FD. Differential intraocular pressure in strabismus diagnosis. Ophthalmology 1981;87:59–70
84. Reader AL III. Normal variations of intraocular pressure on vertical gaze. Ophthalmology 1982;89:1084–1087.
85. Jaensch PA. Paresen der schrúgen Heber. Albrecht von Graefes Arch Klin Exp Ophthalmol 1929;121:113–125.
86. Wolff J. The occurrence of retraction movements of the eyeball together with congenital defects in the external ocular muscles. Arch Ophthalmol 1900;29: 297–309.
87. Dunnington JH, Berke RN. Exophthalmos due to chronic orbital myositis. Arch Ophthalmol 1943;30:446–466.
88. Duane TD, Schatz HJ, Caputo AR. Pseudo-Duane's retraction syndrome. Trans Am Ophthalmol Soc 1976;74:121–132.
89. Goldstein JH, Sachs DB. Bilateral Duane's syndrome. J Pediatr Ophthalmol 1977;14:12–17.
90. Oei TH, Verhagen WIM, Horsten GPM. Forced duction test in clinical practice. Ophthalmologica 1983;186:87–90.
91. Smith JL. The office forced duction test. Ophthalmic Surg 1975;6:62–63.
92. Stephens KF, Reinecke RD. Quantitative forced duction. Trans Am Acad Ophthalmol Otolaryngol 1967;71:324–329.
93. Knox DL. Suction cup forced duction. Am J Ophthalmol 1974;77:762–763.
94. Miller FS III, Kalina RE. Suction cup for forced duction testing. Am J Ophthalmol 1984;97:390–391.
95. Guyton DL. Exaggerated traction test for the oblique muscles. Ophthalmology 1981;88:1035–1040.
96. Jampolsky A. Strabismus: surgical overcorrections. Highlights Ophthalmol 1965;8:78–79.
97. Katz RL, Eakins KE. Pharmacologic studies of extraocular muscles. Invest Ophthalmol 1967;6:261–268.
98. Smith RB. Succinylcholine and the forced duction test. Ophthalmic Surg 1974; 5:53–55.
99. Scott AB. Active force tests in lateral rectus paralysis. Arch Ophthalmol 1971; 85:397–404.
100. Scott AB. Extraocular muscle forces in strabismus. In: Bach-y-Rita P, Collins CC, Hyde JE, eds. The Control of Eye Movements. New York, Academic Press, 1971:327–342.
101. Scott AB, Collins CC, O'Meara DM. A forceps to measure strabismus forces. Arch Ophthalmol 1972;88:330–333.
102. Bender MB, Bergman PS, Nathanson M. Ocular movements on passive head turning and caloric stimulation in comatose patients. Trans Am Neurol Assoc 1955;80:184–185.
103. Nathanson M, Bergman PS, Anderson PJ. Significance of oculocephalic and caloric responses in the unconscious patient. Neurology 1957;7:829–832.
104. Fisher CM. Neurological examination of the comatose patient. Acta Neurol Scand 1969;45(Suppl 36):1–56.
105. Leigh RJ, Hanley DF, Munschauer FE III, et al. Eye movements induced by head rotation in unresponsive patients. Ann Neurol 1984;15:465–473.
106. Demer JL, Honrubia V, Baloh RW. Dynamic visual acuity: a test for oscillopsia and vestibulo-ocular reflex function. Am J Otol 1994;15:340–347.
107. Chu FC, Reingold D, Cogan DG. A maneuver to elicit vertical dolls' eye movements. Am J Ophthalmol 1979;87:724.
108. Griffin JF, Wray SH, Anderson DP. Misdiagnosis of spasm of the near reflex. Neurology 1976;26:1018–1020.
109. Troost BT, Troost EG. Functional paralysis of horizontal gaze. Neurology 1979; 29:82–85.
110. Klingon GH. Caloric stimulation in localization of brainstem lesions in the comatose patient. Arch Neurol 1952;68:233–235.
111. Vaernet K. Caloric vestibular reactions in transtentorial herniation of the brainstem. Neurology 1957;7:833–836.
112. Blegvard B. Caloric vestibular reactions in transtentorial herniation of the brainstem. Acta Psychiatr Neurol Scand 1962;30:187–196.
113. Rodriguez Barrios R, Mottinelli MD, Medoc J. The study of ocular motility in the comatose patient. J Neurol Sci 1966;3:183–206.
114. Johnkees LBW. The caloric test and its value in evaluation of the patient with vertigo. Otolaryngol Clin North Am 1973;6:73–93.
115. Eviatar A. A critical look at the ''cold calorics.'' Arch Otolaryngol 1974;89: 361–365.
116. Plum F, Posner JB. The Diagnosis of Stupor and Coma. 3rd ed. Philadelphia, FA Davis, 1980.
117. Frenzel H. Spontan- und Provokations-nystagmus als Krankheitssymptom: Ein Leitfaden für seine Beobachtung, Aufzeichnung, und Formanalyse. Berlin, Springer-Verlag, 1955.
118. Luxon L. Comparison of assessment of caloric nystagmus by observation of suration and by electronystagmographic measurement of slow-phase velocity. Br J Audiol 1995;29:107–115.
119. Itaya T, Kitahara M. Air caloric test with continuous thermal change in patients with vestibular disorders. Acta Oto-Laryngol (Suppl) 1995;519:184–187.
120. Facco E, Zuccarello M, Pittoni G, et al. Early outcome prediction in severe head injury: Comparison between children and adults. Childs Nerv Sys 1986;2:67–71.
121. Kennedy CR, Duffy SW, Smith R, et al. Clinical predictors of outcome of encephalitis. Arch Dis Child 1987;62:1156–1162.
122. Müller-Jensen A, Neunzig HP, Emskotter T. Outcome prediction in comatose patients: signficance of reflex eye movement analysis. J Neurol Neurosurg Psychiatr 1987;50:389–392.
123. Griesen O. Pseudocaloric nystagmus. Acta Otolaryngol 1972;73:341–343.
124. Becker GD, Davis JL, Parell GJ. Pseudocaloric nystagmus. Arch Neurol 1978; 35:93–94.
125. Collewijn H, van der Steen J, Steinman RM. Human eye movements associated with blinks and prolonged eyelid closure. J Neurophysiol 1985;54:11–27.
126. Jampel RS, Haidt SJ. Bell's phenomenon and acute idiopathic polyneuritis. Am J Ophthalmol 1972;74:145–153.
127. Ropper AH. The CNS in Guillain-Barré syndrome. Arch Neurol 1983;40: 397–398.
128. Miller NR, Griffin J, Cornblath D, et al. Intact Bell's phenomenon in a patient with myasthenia gravis and upward gaze paresis. Arch Ophthalmol 1989;107: 1117.
129. Hall A. The origin and purposes of blinking. Br J Ophthalmol 1945;29:455–467.
130. Francis IC, Loughhead JA. Bell's phenomenon. A study of 508 patients. Austral J Ophthalmol 1984;12:15–21.
131. Guibor GP. Some eye defects seen in cerebral palsy, with some statistics. Am J Phys Med 1953;32:342–347.
132. Frandsen AD. Occurrence of squint. Acta Ophthalmol (Suppl) 1960;62:9–158.
133. Hiles DA, Wallar PH, McFarlane F. Current concepts in the management of strabismus in children with cerebral palsy. Ann Ophthalmol 1975;7:789–798.
134. Hoyt CS. Abnormalities of the vestibulo-ocular response in congenital esotropia. Am J Ophthalmol 1982;93:704–708.
135. Anderson WD, Lubow M. Astrocytoma of the corpus callosum presenting with acute comitant esotropia. Am J Ophthalmol 1970;69:594–598.
136. Wolter JR, Kindt GW, Waldman J. Concomitant exotropia: following Reye's syndrome. J Pediatr Ophthalmol 1975;12:162–164.
137. Goldstein JH, Wolintz AH, Stein SC. Concomitant strabismus as a sign of intracranial disease. Ann Ophthalmol 1983;15:53–55.
138. Bixenman WW, Laguna JF. Acquired esotropia as initial manifestation of Arnold-Chiari malformation. J Pediatr Ophthalmol Strabismus 1987;24:83–86.
139. Vollrath-Junger C, Lang J. Akuter Strabismus convergens bei erhöhtem Hirndruck. Klin Monatsbl Augenheilkd 1987;190:359–362.
140. Slavin ML. Transient comitant esotropia in a child with migraine. Am J Ophthalmol 1989;107:190–192.
141. Williams AS, Hoyt CS. Acute comitant esotropia in children with brain tumors. Arch Ophthalmol 1989;107:376–378.
142. Astle WF, Miller SJ. Acute comitant esotropia: a sign of intracranial disease. Can J Ophthalmol 1994;29:151–154.
143. Akman A, Dayanir V, Sanaç AS, et al. Acquired esotropia as presenting sign of cranio-cervical junction anomalies. Neuroophthalmology 1995;15:311–314.
144. Hoyt CS, Good WV. Acute onset concomitant esotropia: when is it a sign of serious neurological disease? Br J Ophthalmol 1995;79:498–501.
145. Lewis AR, Kline LB, Sharpe JA. Acquired esotropia due to Arnold-Chiari I malformation. J Neuroophthalmol 1996;16:49–54.
146. Macpherson H, De Becker I, MacNeill JR. Beware: armed and dangerous—acquired non-accommodative esotropia. Am Orthop J 1996;46:44–56.
147. Simon JW, Waldman JB, Couture KC. Cerebellar astrocytoma manifesting as isolated, comitant esotropia in childhood. Am J Ophthalmol 1996;121:584–586.
148. Hiatt RL, Boyer SL, Cope-Troupe C. Decompensated strabismus. Ann Ophthalmol 1979;11:1581–1588.

149. Legmann Simon A, Borchert M. Etiology and prognosis of acute, late-onset esotropia. Ophthalmology 1997;104:1348–1352.
150. von Graefe A. Beiträge zur physiologie und pathologie der schiefen augenmuskeln. Albrecht von Graefes Arch Klin Exp Ophthalmol 1854;1:1–81.
151. Weir CR, Cleary M, Parks S, Dutton GN. Spatial localization in esotropia: does extraretinal eye position information change? Invest Ophthalmol Vis Sci 2000; 41:3782–3786.
152. von Noorden GK, Awaya S, Romano PE. Past pointing in paralytic strabismus. Trans Am Ophthalmol Soc 1970;68:72–85.
153. von Noorden GK, Awaya S, Romano PE. Past pointing in paralytic strabismus. Am J Ophthalmol 1971;71:27–33.
154. Perenin MT, Jeannerod M, Prablanc C. Spatial localisation with paralysed eye muscles. Ophthalmologica 1977;175:206–214.
155. Helmholtz H. Treatise on Physiological Optics. Vol 3 [Southall JPC, translator]. New York, Dover Publications, 1962.
156. Weir CR. Spatial localization: does extraocular muscle proprioception play a role? Graefes Arch Clin Exp Ophthalmol 2000;238:868–873.
157. Paysse EA, Coates DK. Anomalous head posture with early-onset homonymous hemianopia. J AAPOS 1997;1:209–213.
158. Maddox EE. A new test for heterophoria. Ophthalmol Rev 1890;9:129–133.
159. Trobe JD. Cyclodeviation in acquired vertical strabismus. Arch Ophthalmol 1984;102:717–20.
160. Simons K, Arnoldi K, Brown MH. Color dissociation artifacts in double Maddox rod cyclodeviation testing. Ophthalmology 1994;101:1897–1901.
161. Roper-Hall G. Methods and theories of testing extraocular movements. Int Ophthalmol Clin 1978;18:1–18.
162. Hess WR. Ein einfaches messendes Verfahren zur Motilitätsprufung der Augen. Z Augenheilkd 1916;35:201–219.
163. Lancaster WB. Detecting, measuring, plotting and interpreting ocular deviations. Trans Sect Ophthalmol AMA, 1939:78–94.
164. Zee DS, Chu FC, Optican LM, et al. Graphic analysis of paralytic strabismus with the Lancaster red-green test. Am J Ophthalmol 1984;97:587–592.
165. Hirschberg J. Über die messung des schieldgrades und die dosierung der schieloperation. Zentrabl Prakt Augenheilkd 1885;8:325.
166. Krimsky E. The Management of Binocular Imbalance. Philadelphia, Lea & Febiger, 1948:175ff.
167. Kornder LD, Nursey JN, Pratt-Johnson JA, et al. Detection of manifest strabismus in young children: I. A prospective study. Am J Ophthalmol 1974;77: 207–210.
168. Kornder LD, Nursey JN, Pratt-Johnson JA, et al. Detection of manifest strabismus in young children: II. A retrospective study. Am J Ophthalmol 1974;77: 211–214.
169. Thompson JT, Guyton DL. Ophthalmic prisms: measurement errors and how to minimize them. Ophthalmology 1983;90:204–210.
170. Bielschowsky A. Lectures on Motor Anomalies of the Eyes. Hanover, Dartmouth Publications, 1940:88.
171. Urist MJ. Head tilt in vertical muscle paresis. Am J Ophthalmol 1970;69: 440–442.
172. Ohtsuki H, Kishimoto F, Kobashi R, et al. Motor adaptation in the Bielschowsky head-tilt test in cases of superior oblique palsy. Nippon Ganka Gakkai Zashi [Acta Soc Ophthalmol Jpn] 1992;96:1055–1060.
173. Haagedorn A. A new diagnostic motility scheme. Am J Ophthalmol 1942;25: 726–728.
174. Parks MM. Isolated cyclovertical muscle palsy. Arch Ophthalmol 1958;60: 1027–1035.
175. Hardesty HH. Diagnosis of paretic vertical rotators. Am J Ophthalmol 1963;56: 811–816.
176. Kushner BJ. Errors in the three-step test in the diagnosis of vertical strabismus. Ophthalmology 1989;96:127–132.
177. Sydnor CF, Seaber JH, Buckley EG. Traumatic superior oblique palsies. Ophthalmology 1982;89:134–138.
178. von Noorden GK, Hansell R. Clinical characteristics and treatment of isolated inferior rectus paralysis. Ophthalmology 1991;98:253–257.
179. Moster ML, Bosley TM, Slavin ML, et al. Thyroid ophthalmopathy presenting as superior oblique paresis. J Clin Neuroophthalmol 1992;12:94–97.
180. Guyton DL. Clinical assessment of ocular torsion. Am Orthopt J 1983;33:7–15.
181. Guyton DL, Weingarten P. Sensory torsion as the cause of primary oblique muscle overaction/underaction and A- and V-pattern strabismus. Binoc Vision Eye Muscle Qtrly 1994;9:209–236.
182. Guyton DL. Strabismus surgery decisions based on torsion findings. In: Long AD, ed. Anterior Segment and Strabismus Surgery. Amsterdam, Kugler, 1996: 129–137.
183. Madigan WP Jr, Katz NN. Ocular torsion: direct measurement with indirect ophthalmoscope and protractor. J Pediatr Ophthalmol Strabismus 1992;29: 171–174.
184. Morton GV, Lucchese N, Kushner BJ. The role of fundoscopy and fundus photography in strabismus diagnosis. Ophthalmology 1983;90:1186–1191.
185. Ruttum M, von Noorden GK. Adaptation to tilting of the environment in cyclotropia. Am J Ophthalmol 1983;96:229–237.
186. De Ancos E, Klainguti G. An objective measure of ocular torsion: a new indirect ophthalmoscopy lens. Klin Monatsbl Augeneheilkd 1994;204:360–362.
187. Benson AJ, Barnes GR. Vision during angular oscillation: the dynamic interaction of visual and vestibular mechanisms. Aviat Space Environ Med 1978;49: 340–345.
188. Barnes GR, Benson AJ, Prior ARJ. Visual-vestibular interaction in the control of eye movement. Aviat Space Environ Med 1978;49:557–564.
189. Zee DS. Suppression of vestibular nystagmus. Ann Neurol 1977;1:207.
190. Chambers BR, Gresty MA. Effects of fixation and optokinetic stimulation on vestibulo-ocular reflex suppression. J Neurol Neurosurg Psychiatr 1982;45: 998–1004.
191. Barnes GR, Grealy MA. Predictive mechanisms of head-eye coordination and vestibulo-ocular reflex suppression in humans. J Vestib Res 1992;2:193–212.
192. Dichgans J. Optokinetic nystagmus as dependent on the retinal periphery via the vestibular nucleus. In: Baker R, Berthoz A, eds. Control of Gaze by Brain Stem Neurons. New York, Elsevier, 1977:261–267.
193. Halmagyi GM, Gresty MA. Clinical signs of visual-vestibular interaction. J Neurol Neurosurg Psychiatr 1979;42:934–939.
194. Buttner U, Grundei T. Gaze-evoked nystagmus and smooth pursuit deficits: their relationship studied in 52 patients. J Neurol 1995;242:384–389.
195. Waterson JA, Barnes GR, Grealy MA. A quantitative study of eye and head movements during smooth pursuit in patients with cerebellar disease. Brain 1992; 115:1343–1358.
196. Catz A, Ron S, Solzi P, Korczyn AD. Vestibulo-ocular reflex suppression following hemispheric stroke. Scand J Rehab Med 1993;25:149–152.
197. Fetter M, Klockgether T, Schulz JB, et al. Oculomotor abnormalities and MRI findings in idiopathic cerebellar ataxia. J Neurol 1994;241:234–241.
198. Baloh RW, Sills AW, Kumley WE, et al. Quantitative measurement of saccade amplitude, duration, and velocity. Neurology 1975;25:1065–1070.
199. Collewijn H, Van der Mark F, Jansen TC. Precise recording of human eye movements. Vision Res 1975;15:447–450.
200. Young LR, Sheena D. Survey of eye movement recording models. Behav Res Method Instrum 1975;7:397–429.
201. Baloh RW, Kumley WE, Sills AW, et al. Quantitative measurement of smooth pursuit eye movements. Ann Otolaryngol 1976;85:111–119.
202. Dell'Osso LF, Daroff RB. Eye movement characteristics and recording techniques. In: Glaser JS, ed. Neuro-ophthalmology. Hagerstown, Harper and Row, 1978:185–198.
203. Robinson DA. The control of eye movements. In: Brooks VB, ed. Handbook of Physiology. Vol 2. Bethesda, American Physiology Society, 1981:1275–1320.
204. Yee RD. Eye movement recording as a clinical tool. Ophthalmology 1983;90: 211–222.
205. Eizenman M, Frecker RC, Hallett PE. Precise non-contacting measurement of eye movements using the corneal reflex. Vis Res 1984;24:167–174.
206. Bechert K, Koenig E. A search coil system with automatic field stabilization, calibration, and geometric processing for eye movement recordings in humans. Neuroophthalmology 1996;16:163–170.
207. Koenig E, Westermann H, Jäger K, et al. A new multiaxis rotating chair for oculomotor and vestibular function testing in humans. Neuroophthalmology 1996;16:157–162.
208. Metz HS, Scott AB, O'Meara DM, et al. Ocular saccades in lateral rectus palsy. Arch Ophthalmol 1970;84:453–460.
209. Metz HS. III nerve palsy: I. Saccadic velocity studies. Ann Ophthalmol 1973; 5:526–528.
210. Metz HS, Scott WE, Madison E, et al. Saccadic velocity and active force studies in blowout fractures of the orbit. Am J Ophthalmol 1974;78:665–670.
211. Zee DS, Yee RD. Abnormal saccades in paralytic strabismus. Am J Ophthalmol 1977;83:112–114.
212. Metz HS. Saccades with limited downward gaze. Arch Ophthalmol 1980;98: 2204–2205.
213. Stathacopoulos RA, Yee RD, Bateman JB. Vertical saccades in superior oblique palsy. Inv Ophthalmol Vis Sci 1991;32:1938–1943.
214. Tian S, Lennerstrand G. Vertical saccadic velocity and force development in superior oblique palsy. Vis Res 1994;34:1785–1798.
215. Lewis RF, Zee DJ, Repka MX, et al. Regulation of static and dynamic ocular alignment in patients with trochlear nerve paresis. Vision Res 1995;35: 3255–3264.
216. Ferman L, Collewijn H, Jansen TC, et al. Human gaze stability in the horizontal, vertical and torsional direction during voluntary head movements, evaluated with a three-dimensional scleral induction coil. Vision Res 1987;27:811–828.
217. Saadat D, Oleary DP, Pulec JL, et al. Comparison of vestibular autorotation and caloric testing. Otolaryngol Head Neck Surg 1995;113:215–222.
218. Goebel JA, Hanson JM, Langhofer LR, et al. Head-shake vestibulo-ocular reflex testing: comparison of results with rotational chair testing. Otolaryngol Head Neck Surg 1995;112:203–209.
219. Greenwood R, Hopkins A. Muscle responses during sudden falls in man. J Physiol (Lond) 1976;254:507–518.
220. Greenwood R, Hopkins A. Landing from an unexpected fall and a voluntary step. Brain 1976;99:375–386.
221. Watt DGD. Responses of cats to sudden falls: an otolith-originating reflex assisting landing. J Neurophysiol 1976;39:257–265.
222. Halmagyi GM, Gresty MA. Eye blink reflexes to sudden free falls: a clinical test of otolith function. J Neurol Neurosurg Psychiatr 1983;46:844–847.
223. Dichgans J, Von Reutern GM, Rommelt U. Impaired suppression of vestibular nystagmus by fixation in cerebellar and noncerebellar patients. Arch Psychiatr Nervenkr 1978;226:183–199.