Interpretation and Usefulness of Ocular Motility Testing

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There is much to be learned from the ocular motility test battery. The abnormalities that can be observed and their localization are summarized in Table 6–1. In the following text normal values for ocular motility testing are provided and common patterns of ocular motor abnormality receive comment. Most abnormalities will be illustrated, and can be compared with illustrations of normal ocular motor performance found in the preceding chapter. Unless otherwise noted, recordings of eye movements used for illustrations were made using an electro-oculography (EOG) system having a 0- to 40-Hz bandwidth.

SACCADES

Cerebellar disorders and degenerative disorders of the central nervous system can often be diagnosed through saccadic testing. The three saccadic parameters most relevant to clinicians are peak velocity, latency, and accuracy.

Disorders of Saccadic Velocity

Normal values for the velocity of 20° saccades are given in Table 6–2. Note that velocity is very sensitive to the method by which saccades are recorded. Normal saccadic velocity values obtained by infrared methods or scleral search coil recordings are usually higher than those obtained by means of EOG recordings.

Saccadic velocity is approximately proportional to saccadic amplitude for sizes between 5° and 20°. After amplitude reaches 20°, saccadic velocity undergoes a soft saturation with respect to further increases in amplitude. This pattern is seen on main sequences, which plot peak velocity against saccade size. The usual upper limit for saccadic velocity, no matter how big the saccade, is about 750°/sec. The author uses the function given in equation 6–1 for his limits of normal velocity. For the lower limit, the asymptote is set at 350°/sec. For the upper limit, the asymptote is 750°/sec. Saccadic amplitude is designated by E and saccadic velocity by V. Saccadic velocity cannot be altered voluntarily and is not affected substantially by age or gender.

\[ V = \text{asymptote} \times \left(1 - e^{-E/V} \right) \]

There are several pitfalls to be aware of in measuring saccadic velocities. Variability is appreciable and one is advised to acquire about 40 saccades varying in size between 10° and 40° to develop a reasonable main sequence. Calibration error is another common problem. The calibration error may be related to subtle factors that are not evident when the ocular motility test is read. For example, patients with ocular motor palsies may be unable to get one or both eyes to the inti-
TABLE 6-1.
Summary of Abnormalities Observed in the Oculomotor Tests*  
<table>
<thead>
<tr>
<th>Abnormality</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Saccade test</td>
<td>CNS or ocular disorder</td>
</tr>
<tr>
<td>Too fast saccades</td>
<td>CNS or ocular disorder</td>
</tr>
<tr>
<td>Asymmetrical velocity</td>
<td>CNS or ocular disorder</td>
</tr>
<tr>
<td>Prolonged latency</td>
<td>CNS or ocular disorder</td>
</tr>
<tr>
<td>Asymmetrical latency</td>
<td>CNS disorder</td>
</tr>
<tr>
<td>Dysmetria</td>
<td>CNS or ocular disorder</td>
</tr>
<tr>
<td>Pursuit test</td>
<td>Drug, CNS or visual disorder, nystagmus</td>
</tr>
<tr>
<td>Low gain pursuit</td>
<td>Cerebral nystagmus</td>
</tr>
<tr>
<td>Asymmetrical pursuit</td>
<td>Congenital nystagmus</td>
</tr>
<tr>
<td>Reversed pursuit</td>
<td>Pursuit disorder</td>
</tr>
<tr>
<td>OKN/OKAN test</td>
<td>Cerebral nystagmus</td>
</tr>
<tr>
<td>Low gain OKN</td>
<td>Pursuit disorder</td>
</tr>
<tr>
<td>Asymmetrical OKN</td>
<td>Pursuit disorder</td>
</tr>
<tr>
<td>Absent OKAN</td>
<td>Bilateral peripheral vestibular disorder</td>
</tr>
<tr>
<td>Asymmetrical OKAN</td>
<td>Peripheral vestibular disorder</td>
</tr>
<tr>
<td>Hyperactive OKAN</td>
<td>Mal de debarquement</td>
</tr>
<tr>
<td>Fixation</td>
<td>Pursuit disorder</td>
</tr>
<tr>
<td>Low fixation</td>
<td>CNS or ocular disorder</td>
</tr>
<tr>
<td>Suppression</td>
<td>CNS disorder</td>
</tr>
<tr>
<td>Gaze-evoked nystagmus</td>
<td>CNS disorder or congenital nystagmus</td>
</tr>
<tr>
<td>Refradnystagmus</td>
<td>CNS disorder</td>
</tr>
<tr>
<td>Bizarre nystagmus</td>
<td>CNS disorder</td>
</tr>
<tr>
<td>Square wave jerk</td>
<td>CNS disorder</td>
</tr>
</tbody>
</table>

* CNS, central nervous system; OKN, optokinetic; OKAN, optokinetik eye movements.

get. Patients with strabismus may alternate the eye that they view from, depending on the direction of gaze, and allow one eye to drift out away from the target. In these instances, measured saccadic velocities are wrong, because the calibration is inappropriate. Monocular record-

and single-eye viewing are essential to avoid error in these sorts of patients. In patients without problems of ocular alignment, evidence that the calibration is stable over several trials must be available before diagnosing abnormalities of saccades.

Slow Saccades

There are three types of disorders of saccadic velocity. Saccades may be too slow, too fast, or have substantially different velocities in one eye or direction than the other. Saccadic slowing is diagnosed when mean saccadic velocity for a particular amplitude is less than the lower fifth percentile of normal. Causes of slow saccades are listed in Table 6-3.

When saccadic slowing is observed, drug ingestion should be the first consideration. Anticonvulsants, sedatives, and antidepressants are the most common culprits. Saccades can be slowed as much as 50% when subjects become drowsy. If the patient is wide awake and not taking a centrally acting medication, then the alternative diagnoses in Table 6-3 should be considered.

In some disorders, subtleties of the pattern of saccadic slowing will allow one to further narrow the list of diagnostic possibilities. One should try to judge whether the slowing involves all saccades, or just horizontal or vertical saccades. Metabolic conditions, such as drug ingestion and droxiness, cause changes in the central nervous system that are accomplished by slow

TABLE 6-2.
Peak Velocity of 20° Saccades in Normal Subjects

<table>
<thead>
<tr>
<th>Method</th>
<th>Velocity (°/sec)</th>
<th>Lower Limit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infrared (&lt;300 Hz)*</td>
<td>657 ± 78</td>
<td>491</td>
</tr>
<tr>
<td>Eye coil (60 Hz)</td>
<td>410 ± 100</td>
<td>210</td>
</tr>
<tr>
<td>Electro-oculography (35 Hz)</td>
<td>336 ± 42</td>
<td>252</td>
</tr>
<tr>
<td>Electro-oculography (15 Hz)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


†Han T. Unpublished data. Twenty normal subjects.


problematic conditions.
saccades, such as cerebellar degenerations, Huntington's chorea, and the chronic progressive external ophthalmoplegias also cause a similar global pattern of slowing. Figure 6–1, A shows a graph of slow saccades in a patient with a mitochondrial myopathy, which is one of the causes of chronic progressive external ophthalmoplegia.

On the other hand, several disorders affect vertical saccades and horizontal saccades differentially. Disorders that affect vertical saccades to a greater extent than horizontal saccades include disorders of the midbrain, such as progressive supranuclear palsy (PSP), and the ocular muscle involvement typical of thyroid disease. Figure 6–1, B shows an example of a patient with PSP in which only the larger horizontal saccades are abnormally slowed. Another helpful point that may assist in identification of PSP and related disorders is constriction of range. For example, in Figure 6–1, B, while the target displacement was as large as 40°, this patient showed a paucity of saccades greater than 30°. Examples of disorders that affect horizontal saccades to a greater extent than vertical saccades include focal lesions of the pons such as internuclear ophthalmoplegia, sixth nerve palsy, and disorders of the lateral and medial ocular muscles.

Ocular myasthenia may cause weakness of all ocular muscles, or be restricted to individual muscles. Thus, the
normally fast for their size. A rare cause of saccades that are too fast for their size are ocular disorders in which eye movement is restricted. A large saccade may be programmed centrally, but because the eye is brought up short by muscle contraction or rapid fatigue, a small saccade is made with the velocity appropriate to a bigger saccade. A clue here is that these patients never make saccades faster than the upper limit of normal for large saccades (about 750°/sec for recordings made with a 40-Hz bandwidth).

**Asymmetrical Saccadic Velocity**

Saccadic velocity, for a given amplitude, should be equal between eyes. Velocity should also be equal whether the eye is abducting or adducting. Saccadic velocity asymmetry then consists of significant inequality in velocity between eyes or directions. Of course, asymmetry between eyes can only be detected when monocular recordings are available. Unfortunately, the method by which velocity is measured can create an artifact of asymmetry. Studies done with EOG recordings suggest that adducting saccades are faster, while studies performed with infrared recordings suggest that abducting saccades are faster. The difference between the peak velocities of abducting and adducting 20° saccades reported by Fricker and Sanders is in a population of 40 normal subjects ranged from 70° to 200°sec (95% range, infra-red method). We recommend establishing one's own fifth percentile lower limit of normal, for the method in use locally.

The most frequent causes of asymmetrical velocity are listed in Table 6-5. There are several potential asymmetry patterns, the most common of which is normal abduction with slowed adduction. This occurs mainly in internuclear ophthalmoplegia (INO) (Fig 6-2). Internuclear ophthalmoplegic is due to a lesion in the medial longitudinal fasciculus, which connects the paramedian pontine reticular formation and
the oculomotor nucleus. The INO condition is most often found in patients with multiple sclerosis or cerebrovascular accidents involving the brain stem. The hallmark of INO is slowing of adducting saccades, accompanied by an overshoot of the abducting eye. The condition can be unilateral or bilateral. A reduction in adducting velocity into the abnormal range, accompanied by normal abducting velocity, for medium-size saccades (about 20°) should cause one to consider INO. In this case, one should also examine the position traces of each eye, as shown in Figure 6–2. The combination of an overshoot of the abducting eye, with significant slowing of the adducting eye occurring simultaneously, confirms the diagnosis of INO.

Normal adduction with slowed abduction occurs most commonly in patients with palsy of the sixth cranial nerve. One should look for substantial slowing for a medium-size saccade. Note that calibration error is common in this situation, as the patient with a sixth nerve palsy will often be unable to fixate the target with both eyes when looking in the direction of paresia.

Several other patterns occur frequently in patients with cerebrovascular disease or demyelinating disease involving the brain stem. Preserved abduction in one eye, combined with slowing of all
other horizontal motion in both eyes, occurs in the one-and-a-half syndrome. Reduced speed of adduction in one eye combined with reduced abduction in the other eye occurs in conjugate gaze palsies. Skewing of all horizontal saccades, combined with normal vertical saccades occurs in bilateral pontine lesions which affect the burst cells, such as pontine hemorhage. The interested reader can find more detail about these conditions in Leigh and Zee.53

Disorders of Saccadic Latency

Saccadic latencies are calculated from the difference in time between target displacement and the onset of the first saccade toward the new target position. In Chapter 5, several paradigms to elicit saccades were presented which differed mainly in their effects on saccadic latency. These included the random, express saccade, and antisaccade paradigms. At this writing, only the random paradigm is being used clinically. In this simple procedure, the target changes position at unpredictable times, to unpredictable positions.

Representative normal values for latencies are given in Table 6-6. Normal saccadic latencies are independent of target amplitude and are insensitive to the method used to record eye movements, but vary according to target luminance, size, and contrast (whether the target is visual, auditory, or both) and the predictability of the target. Thus it is best to establish normal values specific to one's own laboratory unless one is using commercial equipment within an environmentally controlled booth.

There are several pitfalls to consider when measuring latency. Latencies are relevant only when the timing of target motion is unpredictable. That is, patients may anticipate predictable targets, producing a latency that is impossibly short or even negative. Latency may also be reduced by input from nonvisual senses, such as noises associated with target displacement. Saccadic latency is strongly affected by visual acuity, and delayed latencies are common in persons with cataracts or other disorders that reduce vision. Latency decreases about 15 ms per logarithmic unit of luminance above foveal threshold.44 Thus a bright target is essential. A small laser produces an extremely bright target which is ideal for this purpose. If one is using a light-emitting diode based stimulator such as a light bar, it may be helpful to test in dim lighting to improve contrast and minimize effects of visual acuity.

Prolonged and Reduced Saccadic Latencies

A general prolongation of saccadic latency is an average latency greater than 400 ms. While general prolongation is associated with certain disease processes, as outlined in Table 6-7, in most instances this finding has no diagnostic significance because saccadic latencies are sensitive to the mental state of the subject. Uncooperative patients may simply produce erratic or prolonged saccadic movements.

There are no disease processes that cause a general shortening of latency; accordingly, this finding is always related to technical error, anticipation, or lack of cooperation. Lack of cooperation can cause the appearance of a general shortening.

TABLE 6-6. Normal Latencies of 20° Saccades

<table>
<thead>
<tr>
<th>Angle</th>
<th>Normal Latency (ms)</th>
</tr>
</thead>
<tbody>
<tr>
<td>225°</td>
<td>2.5 ± 0.5</td>
</tr>
<tr>
<td>275°</td>
<td>7.4 ± 1.7</td>
</tr>
</tbody>
</table>

*Data from Abe et al.*

TABLE 6-7. Disorders of Saccadic Latency

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>General prolongation of latency</td>
<td>Insufficiency of saccades</td>
</tr>
<tr>
<td>Basal ganglia disorders</td>
<td>General shortening of latency</td>
</tr>
<tr>
<td>Anticipation</td>
<td>Extraneous saccades</td>
</tr>
<tr>
<td>Asymmetry in latency</td>
<td>Visual field cut</td>
</tr>
<tr>
<td>Hemi-inattention</td>
<td></td>
</tr>
</tbody>
</table>
Asymmetrical Saccadic Latencies

On the other hand, asymmetry in latency between saccades into one or the other hemifield is useful clinically, as it may indicate the presence of a lesion involving parietal or occipital cortex. Figure 6–3 shows an example of such a patient. What is helpful in this instance is that saccades in one direction provide a control for saccades in the opposite direction. This pattern is frequent in patients who have cerebrovascular accidents. Patients with occipital lesions may not see targets in the blind parts of their fields and may produce a "staircase" of searching saccades, the first of which has a prolonged latency. Patients with parietal lobe lesions have intense attention to the side of their lesion, and may produce no saccade at all or make saccades with prolonged latency to that side. 75

Disorders of Saccadic Accuracy

Causes of the four most common patterns of saccadic inaccuracy—overshoot dysmetria, undershoot dysmetria, glissades, and pulsions—are listed in Table 6–8. These disorders are caused both by ocular disorders and central nervous system disorders.

There are several pitfalls to be aware of when considering the diagnosis of dysmetria. Bink artifact is the most troublesome because many subjects blink with every saccade, unless otherwise instructed. Bink artifact can be easily seen in Figures 6–4 and 6–6 where there are brief deflections in the vertical trace, lasting about 200 ms, accompanied by synchronous deflections in the horizontal traces. Blinks contribute a technical artifac due to interactions with the EOG and infrared methods of measuring eye movements. Only the magnetic scleral eye coil technique of measuring eye movements is immune to blink artifact. EOG recordings are mainly affected in the vertical lead, but in infrared recordings, both the horizontal and vertical components are affected. When EOG recordings are used, it is quite common for the direction of blink artifact to differ between each eye, or for blink artifact to be strong in one eye and absent in the other. These problems are usually related to errors in electrode placement. Blinks are also accompanied by a small eye movement, 79 and also may interact centrally with saccades causing overshoot. 79 Bink artifact is best avoided.
by having a vertical lead recording available, which allows one to ignore saccades with superimposed blinks, and by instructing the patient to avoid blinking during the testing. When a vertical lead is not available, such as in Figures 6–2 and 6–7, it is quite difficult to be sure that a saccade of unusual configuration is truly aberrant, and one may have to fall back on direct visual inspection of the patient.

A more subtle pitfall relates to calibration error. Certain commercial electro- nystagmography systems calculate metrics by comparing the actual saccade displacement to the target displacement. In this situation, an incorrect calibration can cause a numerical dysmetria, which is an artifact of the calibration error. This mistake can easily be detected by inspecting the eye position traces, as true dysmetria is always accompanied by corrective saccades.

**Overshoot Dysmetria**

In overshoot dysmetria, the initial horizontal saccade is too large and the corrective saccade occurs in the opposite direction to the target displacement. Figure 6–4 shows recordings of overshoot dysmetria in a patient with a cerebellar lesion.

Overshoot dysmetria is not always abnormal. In normal subjects, transient overshoot dysmetria is common in saccades directed toward a primary position, in saccades less than about 10° in size, and saccades made to a stimulus appearing in a novel location. Normal subjects will, however, readjust their saccades to a predictable target location and, after several refixations to the same place, stop producing overshoots. Overshoot dysmetria is abnormal when it is frequent (at least 90% of the time), of significant size (greater than 2°), and when it occurs in centrifugal saccades larger than 20°. While numerical criteria for overshoot are available, we do not feel these are necessary, as the diagnosis is usually obvious from inspection. Enduring overshoot dysmetria is a classic sign of a cerebellar lesion. It also can occur in the abducting eye in internuclear ophthalmoplegia, in patients with visual field disturbances, and in the stronger eye of a habitual paretic-eye fixator.
Undershoot Dysmetria

In undershoot dysmetria, the initial saccade is too small and the corrective saccade continues onward toward the target. Undershoot dysmetria does not carry the same pathologic connotation as does overshoot dysmetria, as undershoot is common in normal subjects. Normal subjects will show about 1° to 2° of undershoot for 20° and larger target displacements. Constant and significant (first saccade < 50% of target displacement) undershooting is suggestive of a basal ganglia disorder such as Parkinson's disease or PSP. Figure 6-5 is a graph of dysmetric saccades produced by a patient with PSP. Patients with visual field deficits may also produce inaccurate saccades, but undershooting is the more common pattern, as in this way a hemianoptic patient can hit the target into his or her seeing field. Patients with poor vision, such as those with cataract, may simply be guessing as to new target location, and can produce undershoot or overshoot patterns.

Pulsion

The term "pulsion" is applied to vertical saccades that are pulled to the right or left, requiring a horizontal corrective saccade to fixate the target. Both upward and downward saccades are pulled in the same horizontal direction. Pulsion towards the side of lesion, or "ipsipulsion," occurs after infarcts in the distribution of the posterior inferior cerebellar artery. Pulsion away from the side of lesion, or "contrapulsion," may occur after infarcts in the distribution of the superior cerebellar artery. Most clinical laboratoroies do not attempt to record pulsion.

Glissades

The term "glissade" designates a saccade that does not end crisply, but rather glides to its end point. "Onward glissades" occur when the eye continues to glide in the same direction as the faster part of the saccade; "backward" glissades occur when the eye drifts in the opposite direction to the main saccadic movement. Figure 6-6 illustrates backward glissades in a patient with myasthenia gravis. Glissades occur in conditions in which the brain stem miscalculates the "pulse" of oculomotor activity needed to get the eye to new position or the "step" of innervation needed to hold the eye in place against elastic restoring forces. Thus, glissades are often said to be due to a "pulse-step mismatch." Patients having rapid changes in oculomotor function, such as ocular myasthenics, are particularly prone to developing a glissadic pattern, because the amount of neural firing required to obtain a given eye position and to hold

![Diagram of eye movements with progressive supranuclear palsy](image)
It there against elastic restoring forces is constantly varying. Myasthenics also may demonstrate a briefer drift called "quiver," an example of which is shown in Figure 6-7. Patients with cerebellar lesions may produce glissades because they are unable to adjust their pulse-step ratio. Patients with internuclear ophthalmoplegia show onward prolonged glissades in the adducting eye, and briefer backward glissades in the abducting eye (see Fig 6-2).

The main pitfall to consider when trying to decide if a patient has glissades is the adequacy of head stabilization. If the head is free to move and does so during a saccade, the eye component of a combined head-eye saccade may resemble a glissade. Infrared recordings also have a special problem as they may show a glissade-like artifact related to changes in eyelid position than accompany saccades.

**Pursuit**

Both the sinusoidal and the triangular wave pursuit stimuli are used for clinical testing. Sinusoidal stimuli are appropriate for detecting asymmetrical disturbances of pursuit, and triangular wave stimuli are used to detect pursuit which is better in one direction than the other. Pursuit gain, which is the ratio of eye velocity to target velocity, is affected by target velocity, acceleration, and frequency. For the sinusoidal pursuit stimulus, these three stimulus parameters are mutually interdependent, as discussed in Chapter 5. For the triangular wave pursuit stimulus, velocity is constant, and acceleration appears as periodic pulses. Accordingly, frequency and velocity can be varied independently of acceleration. Unfortunately, perfect tracking of the triangular wave stimulus is impossible because of the abrupt accelerations at turn-around time.

Registration of smooth pursuit is of minor diagnostic utility, because disturbances of pursuit are usually nonspecific. Pursuit performance is strongly affected by attention, and inattentive or uncooperative subjects can perform poorly without having any significant central lesion. Another source of difficulty is that the lack of a standard pursuit paradigm asso...
FIG 6–7. Quiver in a patient with myasthenia gravis. These data were obtained with the sidereal eye coil, after injection of edrophonium (Tensilon).

ciated with a well-defined normal data set. Simple sinusoidal pursuit paradigms can be characterized by pairs of three variables (frequency, amplitude, and peak velocity), and pursuit tracking performance is a function of all three variables. Most laboratories have used idiosyncratic combinations of paradigm variables, which has resulted in the generation of many small normal data sets that cannot be compared to others. There is considerable variability even when the paradigm variables are similar. This variability may be related to factors that are difficult to quantify, such as the degree of alertness present in subjects or the visibility of the pursuit target. Pursuit is easily disrupted by common centrally acting medications such as anticonvulsants, minor tranquilizers, and preparations used for sleep. Finally, it is also clear that pursuit performance declines with age.24

**Normal Limits for Smooth Pursuit**

Figure 6–8 summarizes the lower limits of normal sinusoidal pursuit gain from a commonly used commercial electr

**Symmetrical Disturbances of Pursuit**

Symmetrical reduction of smooth pursuit is encountered frequently. Table 6–9 lists the most common causes of reduced pursuit gain. For the reasons advanced earlier, one should be conservative when diagnosing abnormalities of pursuit. Clinically, it is adequate to classify patients with symmetrical pursuit into those with perfect pursuit, those with moderately impaired pursuit, and those with no pursuit at all. This classification can usually be done by eye from the position trace, when one uses a reasonable sinusoidal stimulus (for example, 0.5 Hz, ± 20° amplitude).

Those with perfect or near perfect
pursuit, as judged from the lack of catch-up saccades, or from pursuit gains greater than 0.8, are considered normal. Persons with some, but not perfect pursuit are in a grey zone. Typically they have pursuit gains greater than 0.2 but less than 0.8. Such moderately impaired pursuit tracking might be related to inattention or to medication, to an underlying central nervous system disorder, or to advanced age.

Persons with no pursuit at all, operationally defined as pursuit gain less than 0.2, are the most important patients to be identified, because they will nearly always have a central nervous system disturbance. Figure 6–9 shows an example taken from a patient with a cerebellar degeneration. A staircase of saccades must be present, which indicates that the patient is attempting to track. Rarely, pursuit gain greater than 1.0 is noted. This is recognized by the occurrence of "backup" saccades (saccades directed against target motion). If backup saccades are not present, one will inevitably find a technical error. Pursuit gain that is truly greater than 1.0 occurs most frequently in patients with a form of congenital nystagmus called "latent nystagmus," during triangular wave pursuit. Some normal subjects can also track with gains slightly greater than 1.0.

Asymmetrical Pursuit

Pursuit which is significantly worse in one direction than another is termed asymmetrical. While rare, asymmetrical pursuit is more often of clinical utility than is symmetrically reduced pursuit, because it is a specific indicator for a central nervous system disorder. One can easily detect pursuit asymmetry if a plot is available in which there is an indication of mean gain and the standard

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**TABLE 6–9.**

<table>
<thead>
<tr>
<th>Disorders of Smooth Pursuit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Advanced age</td>
</tr>
<tr>
<td>Brain stem disorders</td>
</tr>
<tr>
<td>Cerebellar disorders</td>
</tr>
<tr>
<td>Cerebral cortical disturbances</td>
</tr>
<tr>
<td>Congenital nystagmus</td>
</tr>
<tr>
<td>Drug ingestion</td>
</tr>
<tr>
<td>Inattention</td>
</tr>
<tr>
<td>Visual disorders</td>
</tr>
</tbody>
</table>

**FIG 6–9.** Reduced gain pursuit and staircase of saccades indicating that patient is attempting to follow target. This patient had a vermis cerebellar degeneration.
deviation in each direction. One must use a stimulus in which velocity is constant, such as the triangular wave paradigm, in order to be able to compare rightward and leftward gain.

There are several causes of asymmetrical pursuit (Table 6–10). Patients with acute parietal or frontal lesions may transiently exhibit better pursuit directed contralateral to their lesion. Pursuit asymmetry resulting from a cortical injury typically persists for only several weeks.

Unidirectional spontaneous nystagmus may be superimposed on pursuit and cause asymmetry. Spontaneous nystagmus due to peripheral vestibular lesions, when severe, may not affect pursuit at all, but when it is strong (for example, 20°/sec in the dark), it may overwhelm the pursuit system. Spontaneous nystagmus resulting from central lesions may go uncorrected by the pursuit system and result in a pronounced asymmetry pattern. These patients present with a spontaneous nystagmus that is poorly suppressed by fixation, reduced and asymmetrical pursuit, and gaze-evoked nystagmus. An example is shown in Figure 6–10. In these instances it is helpful to measure pursuit gain only around regions where the eye is crossing primary position, as in this way the effects of gaze can be eliminated.

In patients with a form of congenital nystagmus called latent nystagmus, a pursuit asymmetry can be recorded which alternates direction according to the viewing eye. These patients usually have a history of amblyopia. As no pursuit asymmetry is seen by nystagmus may be seen with both eyes viewing, this condition can cause confusion if the patient alternates fixation during the oculomotor battery.

TABLE 6–10.
Causes of Asymmetrical Pursuit:

Acute parietal lobe disorder
Acute frontal lobe disorder
Superimposed nystagmus

FIG 6–10. Asymmetrical pursuit. This recording is from a patient after removal of a posterior fossa meningoma. A complex pattern of nystagmus is shown which is related to the continuation of spontaneous nystagmus, gaze-evoked nystagmus, and diminished pursuit tracking. On the bottom plot, hatched bars indicate pursuit gain for triangular wave tracking, and filled bars indicate gain of optokinetic nystagmus (60°/sec constant velocity). Eye movements were recorded with the infrared technique.

Reversed Pursuit

In certain patients with congenital nystagmus, the eyes will make saccades in the direction of target motion and slow, smooth movements against target motion. Some authors prefer to avoid using the term "reversed pursuit" under these circumstances because eye velocity is not proportional to target velocity in these patients.1

Miscellaneous Pursuit Abnormalities

In patients with poor peripheral vision, such as those with retinal pigmentary degenerations, from time to time the eyes may "get lost" during tracking, showing a characteristic pattern of searching saccades. However, because the patient can find the target intermittently, numerical figures for tracking may be normal. Figure 6–11 shows an example of such a case. The term "disor-
OPTOKINETIC NYSTAGMUS AND OPTOKINETIC AFTERNYSTAGMUS

Disorders of Optokinetic Nystagmus

Optokinetic nystagmus (OKN), like pursuit, has only minor diagnostic utility. Although OKN is more specific than pursuit, as it is not as affected by inattention and medication as is pursuit, it is also less sensitive. Presumably the relative lack of sensitivity of OKN to unilateral and central disorders occurs because OKN is the sum of two tracking mechanisms, namely, the smooth pursuit system, which uses foveal vision, and a separate tracking system, which uses both foveal and extrafoveal vision.

**Normal values for OKN gain are similar to those given for pursuit gain, or slightly greater; but OKN gain is less strongly reduced at high frequencies.**

While normal values are available for OKN phase, it is uncertain whether or not phase is affected by disease. Practically, OKN is best evaluated by comparing it to smooth pursuit, using the normal values developed for pursuit.

There are several pitfalls unique to optokinetic testing. While less sensitive to attention than pursuit, because OKN is disturbing to some patients, there may be an active attempt made to suppress OKN by fixating on a nonmoving object in the room. This pattern is easily recognized because these persons are generally otherwise healthy individuals, and because their initial responses are robust. Also as discussed in Chapter 5, many commercial optokinetic simulators are actually devices which elicit smooth pursuit. If one is using such a device, the diagnostic points listed later, which depend on noticing differences between pursuit and optokinetic responses, do not apply.

**Symmetrically Reduced OKN Gain**

Table 6–11 lists causes of OKN abnormalities. There are three specific patterns of abnormal OKN, the first of which is symmetrically reduced OKN gain. Reduced OKN occurs in visual disorders, in pursuit system disorders, and in disorders of fast phases. While smooth pursuit is most affected by visual acuity, which represents foveal vision, OKN is pro-

<table>
<thead>
<tr>
<th>TABLE 6–11. Causes of Abnormal Optokinetic Nystagmus</th>
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<tbody>
<tr>
<td>Visual disorder:</td>
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<tr>
<td>Pursuit system disorder</td>
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<tr>
<td>Fast-phase disorder</td>
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<tr>
<td>Superimposed nystagmus</td>
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<tr>
<td>Congenital nystagmus</td>
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ganized pursuit" is sometimes applied to severely abnormal pursuit falling into one of the categories mentioned earlier. Disconjugate eye movements may occur rarely during pursuit. In most cases it is not necessary to scrutinize the pursuit trace, because the same underlying disorders that cause disconjugate pursuit, also cause disconjugate saccades.
duced both by foveal and extrfoveal vision, and thus may persist even when visual acuity is poor. In disorders which selectively affect foveal vision, a slow buildup of OKN may occur to a constant velocity stimulus. In disorders which spare foveal vision but abolish peripheral vision, such as extremely severe retinal pigmentary degenerations, no buildup of OKN is seen. Another context in which pursuit is normal but OKN is symmetrically reduced are patients with fast-phase disorders. The most common clinical disorder of this type is PSP, a degenerative disorder of the brainstem, in which saccades are slowed and difficult to initiate. Accordingly, patients with PSP may have normal pursuit to a sinusoidal or triangular wave target, but poor OKN to a drum moving at constant velocity because their OKN "hangs up" in the orbit (Fig 6-12). In other words, the eyes deviate to the orbital edge and just stay there, instead of undergoing periodic resetting quick phases which bring the eye back to the center. These patients show a similar disorder of vestibular fast phases, and get "hung-up" when rotated at constant velocity. In the later stages of PSP, both pursuit and OKN are lost.

Asymmetrical OKN Gain

Asymmetrical OKN is not as helpful for diagnosis of central nervous system disorders as asymmetrical pursuit, mainly because it occurs so infrequently. Presumably asymmetrical OKN is uncommon because it requires lesions in two tracking systems: foveal and extrfoveal. Only a minor asymmetry of OKN appears following complete unilateral peripheral vestibular lesions. Asymmetrical OKN also appears briefly following unilateral parietooccipital lesions.

Reversal of Optokinetic Nystagmus

Reversal or inverted OKN occurs in patients with congenital nystagmus, which is discussed under the heading of fixation. In these patients, the nystagmus beats in the direction of stripe movement. However, the slow-phase velocity of the nystagmus does not scale with the stimulus speed.

Disorders of Optokinetic Afternystagmus

Optokinetic afternystagmus (OKAN) is the nystagmus that follows a constant velocity optokinetic stimulation, after the lights have been turned off. It is a weak response in humans, generally decaying from an initial value of about 10°/second to zero, over about 15 seconds. A good normal data base of normal subjects exists for the study of OKAN. OKAN is characterized by three parameters: initial velocity, the time constant of decay, and the slow-cumulative eye position (SCEP). The most useful of these parameters is the SCEP. The lower limit of normal for SCEP used in the author's laboratory is 40°.

The major pitfall to be aware of when attempting to use OKAN for clinical diagnosis is that OKAN varies substantially in the same individual from trial to trial. Averaging can be used to overcome this problem.

Conditions that may result in abnormal OKAN are listed in Table 6-12.

Symmetrical Reduction of OKAN

There are three abnormal patterns to OKAN: complete loss, significant asymmetry, and hyperactive OKAN. Complete
TABLE 6—12.
Causes of Abnormal Optokinetic Afternystagmus
Optokinetic nystagmus disorders
Peripheral vestibular lesions
Central vestibular lesions
Mal de debarquement syndrome

loss of OKAN, or bilateral reduction of the SCEP to less than 40°, occurs very commonly in patients with bilateral vestibular loss. Figure 6—13 shows an example from a patient with antibiotic-induced ototoxicity. Optokinetic afternystagmus can also be lost in central lesions that affect vestibular connections.

Asymmetrical Reduction of OKAN
Asymmetry of OKAN occurs in patients with unilateral vestibular loss (Fig 6—14). A stronger response is found for drum rotation toward the side of lesion. Asymmetrical OKAN also occurs in many subjects who otherwise test as normal, for uncertain reasons. Because of this normal variability, a significant directional preponderance in OKAN occurs in only about half of patients with complete unilateral vestibular loss.

FIG 6—13.
Reduced optokinetic afternystagmus (OKAN) in a patient with unilateral loss of vestibular function resulting from antibiotic ototoxicity. The cross-hatched area indicates remaining OKAN.

Hyperactive OKAN
Abnormally increased OKAN may be found in patients with "mal de debarquement," a condition in which the vestibular system is overactive, and causes a prolonged "land sickness."

FIXATION
The utility of the fixation test is related to identification of central disorders. Five abnormalities of fixation will be discussed, namely impaired fixation suppression, gaze-evoked nystagmus, re-bound nystagmus, congenital nystagmus, and square wave jerks.

Impaired Fixation Suppression
The diagnosis of impaired fixation suppression is made by observing the effect upon an ongoing nystagmus of asking the subject to fix his or her eyes on a clearly visible target, upon any ongoing nystagmus. The fixation index is the ratio of nystagmus peak slow-phase velocity with fixation to nystagmus intensity without fixation removed (for example, complete darkness or subject's eyes closed).

For nystagmus induced by caloric input, Takemori and Cohen found the normal mean fixation index to be 48 ± 10%. However, it is questionable whether this value is an appropriate normal value for several reasons. It seems likely that fixation index should increase with age, as the pursuit declines with age, but fixation data by age of subject are not available. Similarly, fixation suppression is probably affected by the many other variables that affect pursuit. It also seems likely that the efficiency of fixation is related to the intensity of nystagmus one is attempting to suppress.

Peripheral vestibular nystagmus is usually well suppressed by fixation. Congenital nystagmus, which is to be discussed later, and many varieties of central nystagmus are unaffected by or even increased by fixation. Nystagmus
which is increased by fixation is called "fixation nystagmus."

**Gaze-Evoked Nystagmus**

A second point of information to be gained from the fixation test is the adequacy of gaze holding, as impaired gaze holding may indicate the presence of a central lesion. Gaze-evoked nystagmus is a drift of the eye which is only present for certain directions of gaze. When EEG recordings are used, any persistant nystagmus for ocular displacements of 30° or less are considered abnormal. When using infrared recordings, small amounts of weak (0.5°/sec to 3.0°/sec) gaze-evoked nystagmus may be recorded in normal subjects.

Causes of gaze-evoked nystagmus are listed in Table 6–13. Several distinct patterns of gaze-evoked nystagmus can be identified by scrutiny of the eye position trace. The most common variety consists of a drift towards the center of the orbit, interspersed by corrective on-going saccades attempting to acquire a target which has drifted off the locus (Fig 6–15). In this situation, the initial rate at which the eye drifts is directly proportional to how far the eye is from center, because elastic restoring forces are proportional to displacement. Accordingly, as the eye approaches center, the rate of drift decreases, accounting for the characteristic decreasing exponential trajectory of ocular drift. The decreasing exponential pattern may be difficult to appreciate if the patient makes frequent saccades to the target, and one must look for a slow phase in which the patient allowed his or her eye to drift close to the center. Gaze-evoked nystagmus on lateral gaze and upward gaze is common, whereas gaze-evoked nystagmus on downward gaze is infrequent. Certain patients with congenital nystagmus...
Gaze-evoked nystagmus in a patient with a cerebellar degeneration. This patient also had a downbeat nystagmus (see vertical lead), which increased on left gaze.

FIG 6–15.

Gaze-evoked Nystagmus in a patient with a cerebellar degeneration. This patient also had a downbeat nystagmus (see vertical lead), which increased on left gaze.

Gaze-evoked nystagmus or with acquired central nystagmus varieties have increasing exponential velocity patterns. More will be said about this shortly under the heading "Congenital Nystagmus."

Two factors may contribute to the amount of gaze-evoked nystagmus found in individual patients. The first relates to the pattern of neural firing associated with maintenance of eye position against elastic restoring forces. Central disorders, particularly those involving the cerebellum, can disrupt the neural "step" of firing, which holds the eye in place against elastic forces, and cause centripetal drift. A second consideration relates to how proficiently the patient can use visual tracking mechanisms such as pursuit or optokinetic responses to offset and eliminate drift, even though it is self-generated.

Gaze-evoked nystagmus which is greater when the subject is looking in one direction than in the other occurs in several situations. In vestibular disorders, when gaze-evoked nystagmus is combined with a spontaneous nystagmus, they add when gazing toward the fast phase of the spontaneous nystagmus and subtract when the gaze is toward the opposite direction. This often results in the pattern of a greater overall nystagmus when gazing towards the fast-phase direction of the spontaneous nystagmus. This common clinical pattern is called "Alexander's law," and occurs in patients with peripheral and in some patients with central vestibular imbalance (Fig 6–16). Brown's nystagmus, which occurs in patients with cerebellar lesions, refers to asymmetrical nystagmus in which there is little or no spontaneous nystagmus in a primary position, but an asymmetry exists at the extremes of lateral gaze (Fig 6–17). Patients with NO often exhibit a disconjugate gaze-evoked nystagmus in which the abducting eye exhibits a more prominent nystagmus than the adducting eye (see Fig 6–2).

Rebound Nystagmus

Rebound nystagmus is a primary position nystagmus that is provoked by prolonged eccentric gaze holding. It appears after the eyes are returned to primary position. An example is shown in Figure 6–18. An abnormal amount of rebound consists of at least three beats of clear nystagmus, with the slow-phases directed toward the previous position of gaze. Rebound after gaze holding for periods more prolonged than 30 seconds, or for eccentricities larger than about 45° is of uncertain significance, as healthy subjects may exhibit rebound under such circumstances. Vertical rebound nystagmus is rare.

Rebound nystagmus is always pathologic and is related to brain stem or cerebellar disease. Accordingly, if an unusually large gaze-evoked nystagmus is observed, one should automatically look for rebound nystagmus. On the other hand, gaze-evoked nystagmus without
rebound is usually of little significance. Rebound is always associated with poor pursuit.

**Congenital Nystagmus**

Congenital nystagmus is a term applied to a diverse group of abnormal eye movements that are noted at birth or shortly thereafter. Congenital nystagmus is included under the category of disorders of fixation because it can frequently present as a severe gaze-evoked nystagmus, and because it is often increased by attempts at fixation. Figure 6–19 shows an example of congenital nystagmus in a patient with albinism, showing rounding of slow phases, with convexity in the direction of gaze. Such “increasing exponential velocity profiles” are typical of congenital nystagmus. No special procedure is required to elicit congenital nystagmus other than that described for registration of gaze-evoked nystagmus.

There are several types of acquired nystagmus that appear similar to congenital nystagmus. Nystagmus of the blind is a constantly present nystagmus that may undergo periodic changes in direction. Spasmus nutans consists of a pendular, dysconjugate nystagmus accompanied by head-nodding, which occurs in children. Similar acquired pendular nystagmus in adults can be caused by multiple sclerosis, and follow brain stem infarctions. Occasional central nystagmus patterns, such as those related to Wernicke’s encephalopathy, may have increasing exponential velocity profiles similar to those seen in some forms of congenital nystagmus.

One must be cautious when using infrared oculography for registration of congenital nystagmus and gaze-evoked nystagmus, because artifact due to transducer nonlinearity can cause an over-
dinary gaze-evoked nystagmus to resemble the increasing exponential pattern described earlier. Also, care must be taken that an unusually intense gaze-evoked nystagmus is not mistaken for congenital nystagmus.

**Square Wave Jerks**

The last point of information to be derived from the fixation test relates to square wave jerks. These are inappropriate saccades that take the eye off the target, followed by a nearly normal inter-saccadic interval (approximately 200

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<th>SES</th>
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**Vertical**

<table>
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<th>TARGET</th>
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**FIG 6-18.** Congenital nystagmus in a patient with abinism.

**TABLE 6-14.**

<table>
<thead>
<tr>
<th>Causes of Square Wave Jerks</th>
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<tbody>
<tr>
<td>Cerebral lesions</td>
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<tr>
<td>Cerebellar disorders</td>
</tr>
<tr>
<td>Basal ganglia disorders</td>
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<tr>
<td>Cholinergic depression</td>
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<td>Strabismus</td>
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<td>Normal aging</td>
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</table>
corded in dark without visual fixation, and 3.4 per minute when recorded with the subject's eyes closed.

The clinical utility of square wave jerks is to point toward the possibility of a central disorder. In young normal persons, square wave jerks occur infrequently. Accordingly, when they are found frequently in a young patient (more than 1 per second), the possibility of a cerebellar disorder should be considered. In the elderly, square wave jerks are common and are rarely of significance. However, in certain conditions such as progressive supranuclear palsy (see Fig 6–20), the diagnosis cannot be made without finding frequent square wave jerks.38

REFERENCES

FIG 6–20. Square wave jerks in a patient with progressive supranuclear palsy. Frequent small square waves are seen superimposed upon a saccade sequence. The initial large horizontal movements are between <20°.


