cord, which nerves, which muscles? Concentrate first on learning the tests that are most diagnostic of common lesions and fill in more details as you see more patients.

A Note to the Sophomore. By now you have probably noted some discrepancies between the sequence of examination taught in your school and the one offered here. In a little while, you will realize that there are some potential conflicts even between the latter and the outline presented in Chapter 4 (e.g., many of the cranial nerves were already examined earlier *en passant*). Even within this chapter there are inevitably arbitrary divisions. For instance, the vestibular and cerebellar systems really should go together. The cerebellum and the posterior (dorsal) columns of the spinal cord might be considered together. But in fact, signs of disease in these three systems are actually in three different sections of the work. Similarly, some of the tests of gaze are not considered with the skilled movements but rather with the cranial nerves whose testing might first reveal such gaze abnormalities. The principle of medicine is clear in all these situations: If you have already learned one order of examination, stick with it. Consistency is more important than the specific virtues of any necessarily arbitrary linear system of organization.

You have also noticed that much of the neurologic examination is performed while doing the musculoskeletal examination. It is not practical to have the patient constantly shift from one position to another. You organize the examination by body region and position, whereas your thinking (and certainly this book) may be organized by the system.

Dr Michael Schlitt of Renton, Washington, states that the most useful thing he learned was to start at one end and move toward the other. It is his preference, when doing a complete neurologic examination, to start with the feet and move north—although he does not record the examination in that order. Physicians more commonly start at the head, in this author's experience, but each one develops a routine that becomes familiar and efficient.

At the beginning of a neurologic examination, Dr Lawrence Huntoon of Lake View, New York, places all his instruments in order on the table and puts each one away after using it. This is comparable to that of a pilot going through a very specific checklist each and every time before taking off.

Cranial Nerves

Cranial Nerve I

The testing of the first cranial nerve is discussed under the regional examination of the nose in Chapter 12. Although performed regionally, the results are recorded under the neurologic examination in the case record.

Cranial Nerves II and III

The testing of vision, including related pupillary signs, is presented in detail in Chapter 10.

Cranial Nerves III, IV, and VI

Pupil (Cranial Nerve III)

Horner syndrome, or the sympathetic-ocular syndrome, consists of anisocoria (with the homolateral pupil smaller), enophthalmos, ptosis, and anhidrosis with decreased pilomotor response. It is frequently seen in Pancoast syndrome (see Chapter 10).

Anisocoria is also discussed in detail in Chapter 10.

Extraocular Muscles (Cranial Nerves III, IV, and VI)

A Difficult Self-test

The extraocular muscles are discussed in Chapter 10, but it is time for a review. The next few cases are difficult but very revealing. Go slowly through the questions that follow. Make good clinical observations and reason carefully from your data.

The answers to the questions will be found at the end of this section so that you will not inadvertently glance at them.

1. Look at Gregory (Fig. 26-1). His eyes are in the primary position. (We refer to the eyes as being in the primary position when the patient is following instructions to look directly at the examiner.) What is wrong with Gregory? Write down your full answer.
2. Moses (Fig. 26-2A) is apparently being seen in the dermatology clinic for a keratotic problem. For the time being, ignore that. (a) If the photograph was taken with Moses looking at you in the primary position, what is the most likely diagnosis? (b) Before turning to the end of this section for the answer, what diagnosis would you make if the picture was taken while Moses was watching your finger move from the midline to his left (your right)?
3. What is wrong with the virgin in Fig. 26-3A? She is not exactly in the primary position, but rather she is gazing down toward your right hand. However, if you ask her to look at you, she will do so but without moving her head. (If you have got the correct answer by now, either you have studied Chapter 10 or you should be writing books like this instead of reading them.)

FIGURE 26-1 Self-test (see text). (Saint Gregory, by Michelangelo, from the Piccolomini altar.)

FIGURE 26-2 Self-test (see text). (Monument of Pope Julius II. Moses, by Michelangelo.)

4. Look at the eyes in Fig. 26-4. (a) If this is the primary position, what are the *two* most likely diagnoses? (b) If you were checking the extraocular movements, and the patient was supposed to be looking to his right (to your left), what would be the two most likely diagnoses?

Answers. To answer these questions, you need to know the cardinal directions of gaze, shown in Fig. 26-5A. These are simplified, excluding the components of external and internal rotation. A more complete picture is given in Fig. 26-5B.

1. If you said oxycephaly, you are 100% wrong; that is just his hat. Go back and look at the eyes. Gregory has a left third cranial nerve oculomotor palsy. Any other answer is wrong. This cannot be a coincidental palsy of the individual muscles innervated by cranial nerve III on the left side.

Why not? (The answer to this question explains why all other explanations are incorrect.) Go back and look at the picture again.

Did you notice the ptosis on the left but not the right? This clearly implicates cranial nerve III. If Gregory were suffering only

FIGURE 26-3 Self-test (see text). The Bielschowsky sign. (The Medici Virgin, by Michelangelo.)

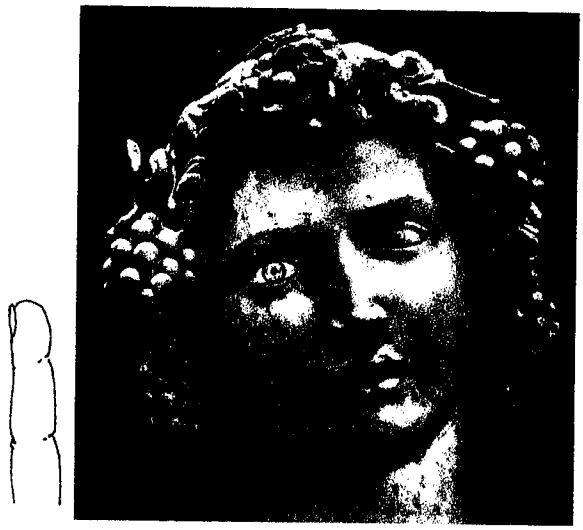


FIGURE 26-4 Self-test (see text). (Bacchus, by Michelangelo.)

from disease of the individual oculomotor muscles (as unlikely as William of Occam[1] would consider that), there would be no ptosis.

The left eye looks laterally because the lateral rectus, which is innervated by cranial nerve VI, is unopposed by the medial rectus, which is innervated by cranial nerve III.

The superior oblique muscle, which is innervated by the trochlear nerve, is unaffected. Although it is used for turning the eye down and in, it is not of much use without the superior rectus, inferior rectus, and inferior oblique muscles, all of which are lost here, being innervated by cranial nerve III.

2. (a) If the appearance in Fig. 26-2A occurs in the primary position, Moses has a left medial rectus muscle paralysis. Again, the unopposed lateral rectus can pull the eyeball laterally when it is not opposed by the medial rectus. (If you got that wrong, go back and do both questions over before reading on because you probably got the second one wrong also.) *Note*: This is probably not caused by an oculomotor nerve lesion as there is no ptosis. There is also no pupillary dilatation, but in the oculomotor nerve palsy secondary to a metabolic disorder such as diabetes (in contrast to structural lesions such as a posterior communicating artery aneurysm), the pupil is generally spared.

(b) If the appearance in Fig. 26-2A is seen while Moses is following your finger to his left, the single lesion most likely to explain the finding is a right medial rectus palsy because he is unable to pull the right eyeball around so that it will point toward your finger, although the left has moved normally.

This case illustrates why one must always describe the examination carefully: The same appearance can indicate disease in three different places, depending upon what instructions the patient is following when the appearance is observed.

Yes, *three* different places. There is one other lesion that could produce the appearance of Fig. 26-2A while gazing to the left (your right). But you could only detect it by testing the cardinal directions of gaze. What if you saw the appearance in Fig. 26-2B when Moses gazed to his right (your left)?

It now appears that each lateral rectus can work. A patient with these findings would look normal when asked to look directly at the examiner because each medial rectus also works. This, then, is a disturbance of conjugate gaze called *bilateral internuclear ophthalmoplegia*. (This is to be distinguished from *internal* ophthalmoplegia, which refers to paralysis of the pupil, and is usually used in distinction to *external* ophthalmoplegia or impaired mobility of the eye.) *Unilateral* internuclear ophthalmoplegia occurs with gaze palsy only to one side.

Internuclear ophthalmoplegia is important to know about for two reasons: (a) If you are not aware of it, you can miss it. (b) It is, like other disorders of conjugate gaze, not a peripheral muscle or nerve sign but a central nervous system sign. Bilateral internuclear ophthalmoplegia (Moses, Fig. 26-2A, B) is often seen in multiple sclerosis (MS); unilateral internuclear ophthalmoplegia often occurs after vascular accidents to the medial longitudinal fasciculus (MLF). Other causes of internuclear ophthalmoplegia include infections, cervical involvement in rheumatoid arthritis (Menezes et al., 1985), paraneoplastic disease, and other degenerative, nutritional, or metabolic processes (Brazis and Lee, 1999).

Neuroanatomy Review. Internuclear ophthalmoplegia is caused by a lesion in the MLF, which connects the abducens nucleus on one side with the oculomotor nucleus on the other. A unilateral MLF lesion causes inability to adduct the ipsilateral eye and also a monocular nystagmus of the contralateral eye when the patient looks *away* from the side of the lesion. *An MLF lesion is distinguished from a lesion of the medial rectus by the patient's ability to converge.* The corticobulbar fibers subserving convergence do not travel in the MLF.

3. This patient has a right trochlear nerve palsy or a paresis of the muscle that it innervates, the right superior oblique. The patient's head is tilted away from the lesion or in the direction in which the affected muscle moves the eye so as to minimize the double vision. This is the natural position of about 50% of the patients with this exceedingly rare problem (Younge and Sutula, 1977). (The head tilt may be documented in childhood and adult photographs.) Even if you did notice the head position, you would have to make the definitive diagnosis by going through the six cardinal motions of gaze. If you ask the patient, you will probably discover that she finds it somewhat difficult to walk down stairs, as diplopia is maximal on looking down.

[1] William of Occam was a philosopher whose "razor" was a logical device used to cut through intellectual problems that have multiple solutions. Basically, it said that the most likely solution is the simplest. Or, a single explanation that explains all findings is more likely to be true than multiple explanations for the same things. In medicine, this means that the least complex diagnosis is most likely to be correct. Stated another way, it tells us never to make two diagnoses when one would explain all the findings.

Now that our technology has developed to the point that we are able to keep patients alive with their diseases (and the attendant signs and symptoms) for prolonged periods, the rule must be modified. It should state that signs and symptoms not yet explained by previous known diagnoses are best explained by a single additional diagnosis, as opposed to two or more additional diagnoses. Most good clinicians understand this intuitively.

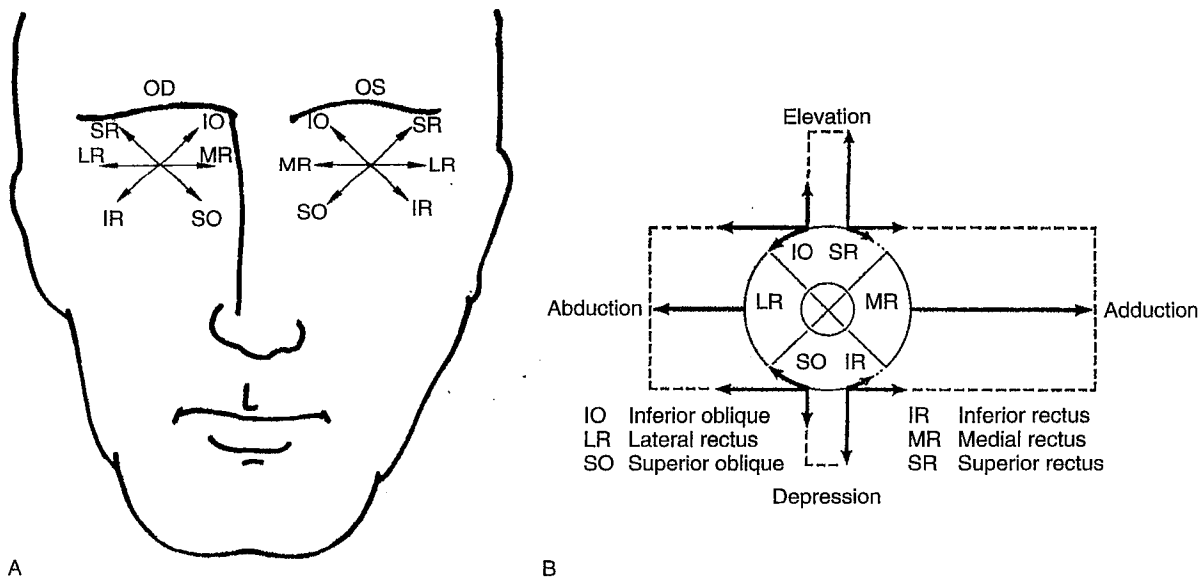


FIGURE 26-5 **A:** Cardinal directions of gaze and functions of the extraocular muscles. (From Younge BR, Sutula F. Analysis of trochlear nerve palsies: diagnosis, etiology, and treatment. *Mayo Clin Proc.* 1977;52:11–18, with permission.) A suggested mnemonic: IOUi (I owe you one: the inferior oblique pulls the globe up when turned in). **B:** Scheme to illustrate the action of the extraocular muscles. Note that only the medial and lateral recti operate in a single plane. Each of the other muscles have three components of action: adduction or abduction; depression or elevation; internal or external rotation. The actions are influenced by the position of the eyeball. When the globe is rotated outward 23 degrees, the superior rectus is a pure elevator, and the inferior rectus a pure depressor. The more the globe is turned inward, the more these muscles act as internal and external rotators. When the eye is turned inward, the inferior and superior oblique act as an elevator and a depressor, respectively. The various muscles act in concert with each other so that their conflicting tendencies normally cancel out to give a harmonious result; for example, when the two obliques aid the lateral rectus in abduction, their vertical and rotatory forces cancel each other out. IO, inferior oblique; MR, medial rectus; SO, superior oblique; SR, superior rectus; LR, lateral rectus; IR, inferior rectus. (Modified from Brain L, Walton JN. *Brain's Diseases of the Nervous System.* London: Oxford University Press; 1969, with permission.)

The cardinal positions of gaze (Fig. 26-5) would reveal the lesion as follows: When the patient looked straight ahead, the right globe would show some slight but definite upward deviation. This would implicate either the right superior oblique or right inferior rectus (the only two muscles that pull the right globe down). The upward deviation of the right globe would increase when the patient looked to the left (inward) but would decrease when she looked to the right (outward). This maneuver would exclude the right inferior rectus and would implicate the right superior oblique. (See the legend to Fig. 26-5B.)

The girl in Fig. 26-3B is the virgin's twin sister. She also has a cranial nerve IV (or superior oblique muscle) paralysis but on the left side. When she came in, her head was tilted toward her right. The present figure shows what happens when you tilt her head (examiner's hands not shown) out of its natural position, toward the side of the lesion. The sign is very subtle but definite. Look carefully at the illustration and describe what has happened.

The left pupil and iris have floated up. Notice that you can now see a little bit of sclera and all of the limbus on the left, but not on the right.

4. (a) In the primary position (i.e., if the left eye is the abnormal one), this would be a left lateral rectus muscle paralysis or a left cranial nerve VI lesion. The left eye is turned inward because the sixth cranial nerve innervates only the lateral rectus, and all of the other unopposed muscles, especially the medial rectus, are pulling the eyeball in.

(b) If, on the other hand, you were testing the extraocular movements (with your finger positioned as shown), this would probably be a right lateral rectus paralysis (or a right cranial nerve VI lesion) because the patient is unable to bring his right eyeball laterally.

Cranial nerve VI is the most common cranial nerve to be afflicted, presumably because it travels the longest course.

An Anatomy Review

For the Guru. If Fig. 26-5 is to be less of a mystery, you need to be able to visualize the action of the extraocular muscles rather than simply memorize the cardinal directions of gaze. To this end, you need to know the muscle insertions with respect to the equator and axis of the eyeball. Study Fig. 26-6; you may need to move the book around to help visualize the globe from various directions. It also helps to know that whereas all of the extraocular muscles have their origin from the common tendinous ring at the back of the orbit, the tendon of superior oblique passes through the trochlea in the medial side of the roof of the orbit before inserting on the globe, changing the direction of the muscle pull by 60 to 90 degrees.

Double Images

Thus far, we have concentrated on how the patient's eyes look to the examiner. From the appearance of the images to the patient, as with the red glass test (*vide infra*), one can also tell which muscle is weak, even if the palsy is so slight that the examiner does not perceive a defect in the ocular movements. If colored glass is not available, an intelligent and cooperative patient can usually distinguish the images by noticing which disappears when each eye is covered completely (Brain and Walton, 1969).

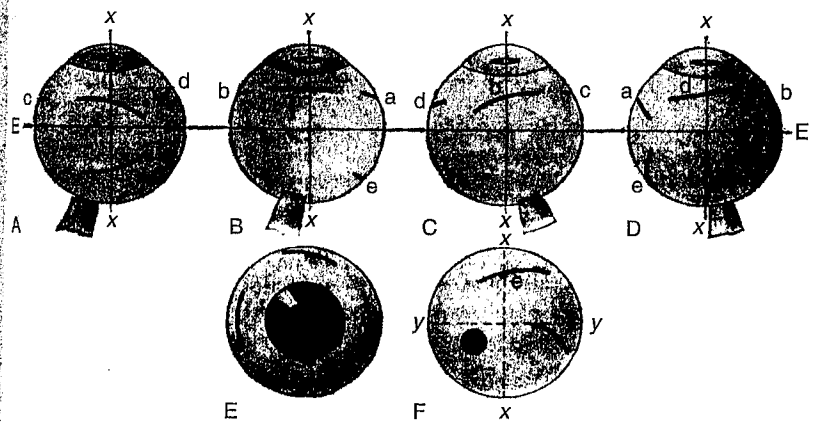


FIGURE 26-6 Insertions of extraocular muscles on the sclera of the right eye. **A:** View from above. **B:** View from nasal side. **C:** View from below. **D:** View from temporal side. **E:** View from the front. **F:** View from behind. a, superior rectus; b, inferior rectus; c, medial rectus; d, lateral rectus; e, superior oblique; f, inferior oblique; EE, equator; xx axis. In **F**, yy, horizontal meridian; xx, vertical meridian. (From Baker F. Eye. In: Buck AH, ed. *Reference Handbook of the Medical Sciences.* Vol. IV. New York: William Wood and Company; 1907:69, with permission.)

The Red Glass Test

A Method

1. Place a *red glass* (or cellophane) over the patient's right eye. Either eye could be used, but by convention it is generally the right.
2. Hold a flashlight at a distance of 1 m in each of the cardinal directions of gaze. Ask the patient to state the position of the red and the white images. Determine the direction in which the two images are farthest apart, and which one is more peripheral.
3. Plot the most deviant position of the false image—the more peripheral one—on the cardinal directions of gaze chart as in Fig. 26-7. Be sure to identify the correct eye from the color of the false image.

A Caveat. Be sure to keep the flashlight beyond the point of convergence. (Testing within the point of convergence is the most common cause of diplopia, with or without the red glass [S. Horenstein, *personal communication*, 1988], because neither eye can aim at the target, and a physiologic diplopia is thus induced. Try it on yourself: Hold your forefinger in front of your nose. As it is moved from say 12 in. away to 2 in. away, attend to how many images you see.)

Explanation

1. *The direction in which the separation between the images is maximum is the direction of action of the paretic muscle.* For example, if the greatest separation occurs on looking to the left, either the left lateral rectus or the right medial rectus is weak.
2. *The image formed by the paretic eye is the one projected most peripherally.* To understand why this is true, it may help to draw a picture. In the normal eye, the image falls on the macula; that is, the crosshairs are on the target. In the paretic eye, the image falls on the retina some distance from the macula. Thus, it will be less distinct (although the patient may not be able to perceive the difference) and will appear to come from the corresponding (opposite) visual field. For example, if the red image is farther to the left as the patient looks to the left, it is the right medial rectus that is not functioning properly. (The image falls on the left macula but on the temporal portion of the right retina, or in the right nasal visual field, to the patient's left.)

With the other muscles, the situation is much more complicated because each has three vectors of motion. The end result is that the false image caused by paresis of a single muscle is found in the quadrant of the visual field indicated in Fig. 26-7, which corresponds to the cardinal direction of gaze principally served by that muscle. With vertical diplopia secondary to a problem in a cranial nerve or the brain, the situation is very complex and may require referral to a specialist. A detailed exposition can be found on the Internet by searching for "Jeff Mann's EM Guidemaps, Diplopia."

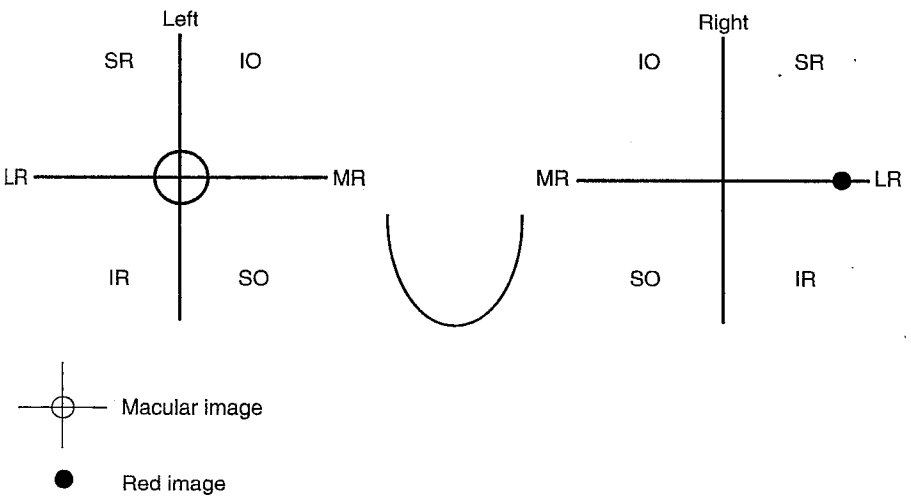


FIGURE 26-7 View through the patient's eyes. The open circle represents the macular image in the normal eye. The false image due to a paretic muscle would be seen within the indicated quadrant. The closed circle represents the false image seen by a patient with a paralysis of the right lateral rectus. (Courtesy of Dr Lawrence Huntoon, with permission.)

Dr Lawrence Huntoon states that three simple rules on diplopia due to cranial nerve palsies are generally accurate:

1. With a sixth nerve palsy, diplopia is greatest when looking to the affected side.
2. With a third nerve palsy, diplopia is greatest when looking up and to the opposite side.
3. With a fourth nerve palsy, diplopia is greatest when looking down and to the opposite side.

Case Report. A physician, on noting that "things just did not look right," ascertained that she had transient diplopia on gaze to the right (i.e., upon looking to the right, there were two images that gradually came together). Performing the red glass test on herself, with the red glass over the right eye, she observed that there were two images when the flashlight was held to the right, with the white image more peripheral, but there was only one image when it was held to the left. Having thereby persuaded herself that she was not imagining things, she made a sketch of her findings (Fig. 26-8) and consulted a neurologist.

Self-study Question. How would you interpret the test? (Answer is in Appendix 26.1.)

A Nonneurologic Cause of Diplopia or Ophthalmoplegia

There are numerous case reports of diplopia or complete ophthalmoplegia, with or without ptosis, in association with statin drug use (see Chapter 10) (Fraunfelder and Richards, 2008).

Paralysis of Conjugate Movements (Gaze)

Horizontal Gaze Palsy

When the patient is unable to shift his gaze to a particular direction, or when the eyes move together (conjugately) in an unexpected manner, the cause is inevitably central in origin. For instance, in acute cerebral cortical disease, especially involving a frontal lobe, the eyes conjugately gaze toward the side of the lesion and cannot, on command, move to the opposite side, although they will move together to the extent that they can move. On the other hand, with destructive lesions in the parietal lobe or below the cerebral cortex, as in the pons, the eyes tend to look away from the side of the lesion. The patient is unable to look in the other direction on command, though the motions that can occur are conjugate. In other words, pontine and parietal gaze palsies tend to "push" the eyes to the opposite side, whereas frontal gaze palsies tend to "pull" the eyes to the same side. With irritative lesions, the rules are reversed.

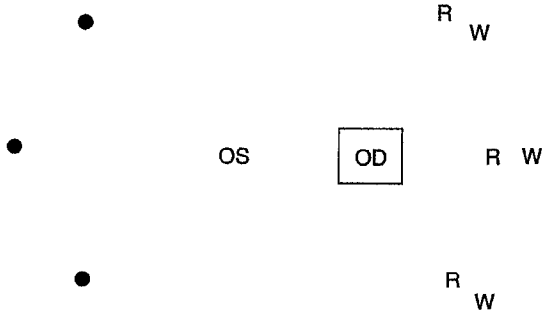


FIGURE 26-8 Diagram of results of red glass test from the chart of the patient described in the case study. R is red image, W is white image, and circles are for superimposed images. *Square* indicates red glass, which was placed over the right eye. OD, right eye, OS, left eye. The neurologist observed some nystagmus in the right eye on gaze to the right. See Appendix 26.1.

Gaze preference refers to conjugate deviation of a spontaneous nature that is inconstant. That is, the eyes can be made to cross the midline, as during caloric stimulation, thus ruling out brainstem disease, but the eyes customarily look at one and only one side.

A common case of a persistent gaze preference is a lesion in the visual cortex of the occipital lobe. A left homonymous hemianopsia causes a gaze preference to the right (L. Huntoon, *personal communication*, 1998).

If there were bilateral cortical infarcts involving the frontal areas controlling the initiation of horizontal gaze, a patient could lose the ability to track on command but would retain the ability to track by reflex. That is, the patient would be unable to follow the instruction "look to the right," but would be able to look to the right in the course of following the examiner's finger or sometimes the examiner's head if the latter was used as a tracking target. Metabolic disturbances can do this, and probably other lesions could interrupt the cortical projections to the midbrain centers controlling the eye movements. However, the most common cause of this phenomenon on a general medical ward is simply a patient who is too fatigued or confused to cooperate with the tracking or looking command, but who will reflexly follow the physician's head (as a tracking target) while staring into the physician's eyes.

Parinaud Syndrome

Parinaud syndrome is paralysis of conjugate vertical gaze due to damage near the posterior commissure. In some cases, downward conjugate gaze is preserved but upward conjugate gaze seems almost always to be lost. If this syndrome is found in a sexually precocious prepubertal boy, make a diagnosis of pinealoma.

An Eponymic Distinction. This Parinaud syndrome is not to be confused with Parinaud conjunctivitis, which is the same as Parinaud oculoglandular syndrome. The latter refers to a preauricular lymph node in combination with an ipsilateral (but unilateral) conjunctivitis (see Chapter 8). This is mentioned to point out the peculiar principle that a majority of clinical syndromes have been described by a small minority of physicians.

For the Advanced Student. Some authorities consider the neurologic Parinaud syndrome to be a triad including, in addition to supranuclear paralysis of upward conjugate gaze, defective convergence and (nonconstricted) pupils that react more briskly to accommodation than to light (Maciewicz, 1983). These latter two signs point to the same midline structures as the first, so the distinction between the two definitions is not always important.

For the Intern. Does the patient have some sort of myopathy or peripheral neuropathy that prevents the eyeballs from moving upward, or is he unable to look up *on command*? One trick is to have the patient track upward (e.g., ask him to follow your finger with his eyes). Some patients can track but cannot voluntarily look up. A second trick is to use the vertical version of the oculocephalic (doll's eye) reflex (sign, test), discussed later. If passive head tilting downward causes the globes to rotate superiorly (in relation to the skull) as the patient fixes his gaze on some object, one has demonstrated (a) an absence of a mysterious myopathy or

a combination of peripheral neuropathies inhibiting upward gaze and (b) that the projections from the midbrain loci controlling eye movements have become severed from their higher (frontal) projections.

For the Resident. Although Parinaud syndrome is most famous as a sign of pinealoma, hydrocephalus is probably its most common cause (Chattha and Delong, 1975).

For the Guru. What Parinaud actually described is a triad of disturbances of the three components of normal convergence. These signs taken together he referred to as "essential." Additionally, he recognized a second form called "combined," in which the "essential" form was combined with paralysis of elevation and/or depression (Parinaud, 1886). Thus, it is quite possible for someone to have Parinaud syndrome according to Parinaud's definition and not have a paralysis of upward conjugate gaze. The essential Parinaud syndrome is a peculiar triad, the individual features of which are quite inconstant:

1. Paralysis of convergence, indicated by the examiner's recognition that convergence does not occur and/or by a peculiar subjective diplopia of variable salience.
2. Paralysis of accommodation in one eye, both eyes, or neither eye (Maciewicz, 1983). (See Table 10.7.)
3. Pupillary reflexes exactly reverse to those of the Argyll Robertson pupil (Parinaud, 1886). Note that this can also occur in Wernicke encephalopathy (see Chapter 10).

In the same paper, Parinaud described a number of other gaze palsies. Most intriguing to the medical historian is the fact that not a single patient reported in Parinaud's original paper (Parinaud, 1886) had a pinealoma.

For the Scholar. By reading the paragraphs in this section in reverse sequence, much as an archaeologist examines the superimposed levels of civilization in a slit trench, from the oldest at the bottom to the newest at the top, one can observe the transmogrification of an original syndrome (Parinaud, 1883). Such distortions are euphemistically referred to as "the advance of modern science."

For the Clinician. Parinaud syndrome due to trauma may be distinguished from Parinaud syndrome due to pinealoma in that the former has a segmental palsy of the iris in response to light stimulation but the latter does not (Thompson, 1978).

Cerebellar Hemorrhage

The diagnosis of cerebellar hemorrhage is discussed here because of the historic importance attributed to its eye signs, especially the conjugate gaze palsies (Table 26.1). Note that the cerebellar hemorrhage has a wide variety of neurologic signs and symptoms by which it may be distinguished from other intracranial hemorrhages. The most common eye finding is an abducens nerve palsy, which also occurs with a number of other lesions.

Cerebellar hematoma often presents as a neurosurgical emergency. General physicians often have difficulty making the diagnosis and the appropriate referral. The presentation is frequently acute and characterized by the triad of headache, vomiting, and ataxia. Progression to gaze palsies, long-tract signs, and respiratory difficulties can occur rapidly. A cerebellar hematoma that is 3 cm or larger on computerized tomography almost always causes brainstem compression and requires immediate surgical evacuation. A large cerebellar infarct can mimic a hematoma and may also require prompt surgical decompression (M. Faria, *personal communication*, 1998).

The sign of "ocular bobbing" (referred to in Table 26.1) is abrupt spontaneous downward jerks of the eye with a slow return to the midposition *plus* paralysis of spontaneous and reflex horizontal eye movements (Bosch et al., 1975). It is actually a sign of compression of the pons and is not a "cerebellar sign" per se. It also may occur with obstructive hydrocephalus, pontine hemorrhages, and, occasionally, metabolic encephalopathy (L. Huntoon, *personal communication*, 1998).

In the mid-1960s, it was believed that one could, and must, make the emergent diagnosis of cerebellar hemorrhage based upon the eye findings. However, the eye signs are not as necessary to the diagnosis as once thought (Heiman and Satya-Murti, 1978).

Newer eye signs are not included in the table (e.g., spontaneous unilateral eye closure [Messert et al., 1976], which occurs because the patient develops an ipsilateral seventh nerve palsy due to displacement of the brainstem by the hematoma and then attempts to avoid diplopia by closing the only eyelid still under his control, the contralateral one. The eye on the side of the hematoma and seventh nerve palsy is the one that remains open).

For the Very Advanced Student. The maximal predictive value of a positive test is about 85% for decreased corneal reflex (*vide infra*),[2] about 67% for facial paresis, and about 71% for the presence of hypertension. The minimal predictive value of a negative test was less than 40% for any of the signs in the table. Even the old historic pearl, "Don't diagnose cerebellar hemorrhage in a nonhypertensive patient," breaks down. The minimal predictive value of a negative test for hypertension was only 40% (Rosenberg and Kaufman, 1976).

A Note on Terminology. We have invented two new statistical terms, the *maximal* predictive value of a positive test and the *minimal* predictive value of a negative test. These are defined as the predictive value of tests calculated from a series in which there were control subjects free of the disease being studied, but where the control subjects were probably not present in the same proportion as in real life. With the prevalence of the diseased subjects inflated, the apparent predictive value of a positive test can be considered the maximal predictive value of a positive test because it would decrease if the subjects without the disease were entered into the study in proportion appropriate to their prevalence. Conversely, the predictive value of the negative test defined in the artificial test population is the minimal predictive value of a negative test.

Cranial Nerve V: Motor

A Method for the Beginner

1. Place your hands over both of the patient's temporalis muscles and instruct the patient, "Grind your teeth." Normally, you should feel both of the temporalis muscles contracting. (Try it on your partner.)
2. Next slip your hands down to the masseter muscles at the lower posterior angle of the mandible. Again, instruct the patient to perform bruxism (teeth grinding) or simply ask him to chew.

[2] The corneal reflex will be impaired in all conditions wherein the facial muscles are paralyzed.

26.1 *Differential diagnosis of intracranial hemorrhages*

Finding	Cerebellar hemorrhage	Thalamic–subthalamic hemorrhage	Putaminal hemorrhage	Pontine hemorrhage	Ruptured aneurysm without intracerebral clot
Hemiplegia	No	Yes	Yes	Quadriplegia or bilateral motor signs	No
Size of pupils	Small, often	Small, often unequal	May be normal	Usually pinpoint	Variable
Pupillary reaction	Yes	Maybe	Yes	Maybe	Yes
Facial weakness	Ipsilateral peripheral mild	Contralateral central	Contralateral central	Contralateral, possibly	No
Sensory deficit	No	Yes	Yes	Yes	No
Conjugate gaze palsy	Common	Maybe	Yes	Yes	No
Side	Ipsilateral	Contralateral	Contralateral	Ipsilateral	—
Reversed by ice water?	No	Usually	Yes	No	—
Sixth nerve palsy	Yes	Yes	No	Yes	Maybe
Hemianopia	No	Yes, clears early	Maybe	Maybe	Maybe
Early inability to walk	Yes	No	No	Yes	No
Vomiting	Severe and repeated	Occasional	Occasional	Yes	Yes
Convulsion	No	No	Yes	No	Yes
Unconscious at presentation	Maybe	No	No	Yes	Often
Eyes deviate downward	No	Yes	No	No	No
Ocular bobbing	Yes	No	No	Yes	No
Preretinal hemorrhage	No	No	Occasional	No	Yes
Abrupt evolution	Often within hours	No	Occasional	No	Yes
Decerebrate posture	Bilateral, usually late	Unilateral	Unilateral	Unilateral	Late

From Brennan R, Bergland R. Acute cerebellar hemorrhage. *Neurology.* 1977;27:527–532; Fisher CM, Picard EH, Polak A, et al. Acute hypertensive cerebellar hemorrhage diagnosis and surgical treatment. *J New Ment Dis.* 1965;140:38–57; Plum F, Posner JB. *The Diagnosis of Stupor and Coma.* 3rd Ed. Philadelphia, PA: FA Davis Co; 1982; and Vincent F. Cerebellar hemorrhage. *Minn Med.* 1976;59:53–458, with permission.

Again you should feel both masseters contract under your hands. (Try it on your partner.)

3. The simplest test is to have the patient open his mouth against resistance. The jaw will deviate toward the less powerful side (M. Schlitt, *personal communication*, 1999).

A Rounding Play

The most common cause of reported bilateral temporalis muscle failure is the examination of an edentulous patient (not realized to be such) by a senior medical student on a neurology elective. (A senior medical student who is already specializing in neurology apparently no longer needs to examine the mouth. Because the cranial nerves do not show up on a computerized tomographic (CT) scan, there is nothing that can be missed by not looking inside the mouth.)

Rather, the edentulous patient should be instructed to gum his lips or lower jaw. (Skip the temporalis muscle examination and go directly to the masseter examination because without real teeth, even this maneuver will not generate as good a contraction of the temporalis as of the masseter.)

Additionally, when examining the mouth, perform the following maneuvers:

1. Rotate the tongue blade within the mouth to a vertical position.
2. Instruct the patient to grasp the tongue blade between his teeth.
3. Then ask him to wiggle the grasped tongue blade.

I prefer this to the usual method of having the patient simply grasp the horizontal tongue blade between his clamped teeth and "hold it in place." By requiring the patient to hold a vertical tongue blade and wiggle it, we also check the internal and external pterygoid muscles. The former method tests only the masseter and temporalis.

Do not check the pterygoids in a patient with a weak masseter because the jaw may dislocate (S. Horenstein, *personal communication*, 1988).

For the Advanced Resident. Landry–Guillain–Barré–Strohl[3] syndrome tends to spare the masseter muscle, which is frequently involved in myasthenia gravis. This distinction is empiric but not scientific because we have no underlying basis for explaining it (see Chapter 1).

Cranial Nerve V: Sensory

Skin Sensation

Because the detailed examination takes the same amount of time as the screening examination, one might as well do the detailed examination. The technique is called double simultaneous stimulation (DSS).

A Method

1. Ask the patient to close his eyes and report which side(s) is (are) touched.
2. Start at the top and work down, being careful to perform testing within each of the three divisions of the trigeminal nerve. Within each division, touch both sides at once as well as one side only.

If the patient insists on watching you, stand in front of him and have him stare at your nose. Moving both your index fingers constantly so as to eliminate visual cuing, occasionally bring one or both fingers close enough to the face to touch the skin lightly.

Do not tap the skin because this stimulates several different branches of the sensory system. You may find it easier to perform the same test with cotton wisps instead of your fingers.

Interpretation. If the afflicted side does not perceive touch with DSS, although it may perceive touch when stimulated by itself, there is generally a cortical lesion. With peripheral disease, the subject will not perceive either a single or double stimulus.

You may wonder why one is sure to touch one side only in each of the three divisions if DSS is superior to single stimulation. First, the subject may have peripheral disease. But even with cortical disease, the single stimulation in each division must be randomly performed so as to break up the "halo" effect. That is, if the subject with the cortical disease always sees the fingers wiggling, even when they are not touching him, and he is always touched doubly, he may learn to report the sensation of DSS each time even when he is not experiencing it, thus defeating the purpose of the test.

Despite the verbosity of the description, DSS with control single-sided stimulation of all three divisions of the trigeminal nerve can be performed in 8 seconds.

Corneal Reflex

The sensory nerve V also supplies the corneal surface.

A Method

1. Moisten a soft piece of cotton or other material that will not scratch the cornea. The corner of a tissue works fine. This should of course be clean. It should not have been used on another patient, especially one with Jakob–Creutzfeldt disease, acquired immune deficiency syndrome (AIDS), or herpetic keratitis.
2. Touch the cornea. This sounds simple, but one must steer between the Scylla of the sclera or the eyelid and the Charybdis of the visual startle reflex. Have the patient look up while you approach the eye from the side so that the patient cannot see the wisp of cotton. When the wisp is over a portion of the cornea not covering the pupillary aperture, go straight in, observing both eyes for the blink.
3. Repeat on the other side.

If you touch the cornea of the right eye and neither eye blinks, what is your diagnosis? If only the left eye blinks, what is your diagnosis? (Write your answers down before looking in Appendix 26.2.)

An Alternate Method

The French method, which avoids the potential for contagion as well as the problems listed in step 2, is to take a straw or a syringe and squirt air at the cornea (S. Horenstein, *personal communication*, 1988).

Interpretation. The corneal reflex involves two different cranial nerves, V for the afferent limb and VII for the efferent limb, and not knowing this can get you into a pickle. This should remind you of the advertisement for the 57 varieties of Heinz pickles. This pickle (5×7) can remind you of the afferent limb (V) and the efferent limb (VII). Accordingly, we can progress to the study of the seventh cranial nerve with relish.

Compromise of cranial nerve IX in a patient with rheumatoid arthritis, manifested by facial paresthesias or loss of the corneal reflex, is suggestive of brainstem compression from "cranial settling" (vertical subluxation of the odontoid process), owing to involvement of upper cervical vertebrae (Bouchaud-Chabot and Lioté, 2002).

Corneomandibular Reflex

If corneal stimulation also produces a lateral deviation of the mandible, it is called the *corneomandibular reflex*. This is probably an associated movement more than a true reflex. It indicates supranuclear interruption of the corticotrigeminal tract on the side of the stimulated cornea.

Whereas the corneomandibular reflex is abnormal in the awake patient, its presence in a comatose patient indicates that the brainstem is intact. It is one of the "brainstem release" signs rarely used today.

The Perioral Reflex

A Method

Place a finger in the angle of the mouth and strike it to see the reflex closure. (Alternately, stroke the nasolabial fold.)

Interpretation. This reflex tests cranial nerve V as the afferent and tests cranial nerve VII as the efferent. Except in infants, there is normally no response. In an adult, the infantile response of contraction of the nearby facial muscles is a cortical release sign, signifying severe damage.

Cranial Nerve VII: Somatomotor Portion

Routinely, we test only the somatomotor portion of cranial nerve VII that supplies the muscles of facial expression.

[3] Strohl was actually the medical student who found the patients and presented them at neurology conference (Guillain et al., 1916).

Peripheral versus Central Palsy

A Method

1. Observe the patient for greater than normal facial asymmetry, including the inspection of the lateral palpebral commissures. Also look for flattening of a nasolabial fold.
2. Ask the patient to raise his eyebrows and look for symmetric wrinkling of the forehead.
3. Ask the patient to close his eyes tightly. Try to force the lids open with your thumb, rolling the skin against the supraorbital rim, *not* against the globe, in order to detect weakness of one eyelid.
4. Ask the patient to show his teeth (or gums) and to puff out his cheeks.

Interpretation. Central facial palsies are caused by cortical (upper motor neuron) lesions, and peripheral palsies are caused by lesions of the facial nerve or its brainstem nucleus (lower motor neurons). Note that "peripheral" as used here includes a portion of the central nervous system, the nucleus in the brainstem.

The upper third of the face, including the muscles of the forehead and the orbicularis oculi, is supplied by a nucleus that receives fibers from both sides of the cerebrum. The lower part of the face is supplied by a nucleus that receives fibers from only one side of the cerebral cortex. Therefore, if the patient experiences a supranuclear lesion (e.g., a lesion of the motor cortex or of its fibers descending to the nucleus) that produces a facial palsy, the forehead will be spared. Such a patient would be able to perform step 2 normally but not step 4. On the other hand, if the facial palsy is due to a lesion of the peripheral nerve or of the brainstem nucleus, the forehead and eyelids will also be paretic. (See "The Bells of Scotland," below.)

In the case record, be sure to describe the findings (e.g., "upper face (not) spared"); "peripheral" and "central" palsy are conclusions, not findings.

When localization of a lesion based upon loss of the upper third of the facial muscle is not completely compatible with the rest of the examination, the former should be abandoned because about 5% of the population does *not* have crossed innervation of the motor supply to the upper third of the facial nerve. In such patients, total facial hemiparesis cannot be used as evidence of nuclear (brainstem) disease. Of course, if the upper third is preserved, that is still evidence of supranuclear disease.

Emotional versus Volitional Pathways

Some patients with facial palsy are wrongly suspected of malingering because they cannot seem to move the facial muscles on the examiner's command but are capable of moving them if the examiner can make the patient laugh involuntarily. This retention of motor function in response to strong emotion is called *volitional palsy*. In contrast, patients with *emotional* (or mimetic) *palsy* can move their facial muscles on command but not during the spontaneous expression of strong emotion. They may be misdiagnosed as parkinsonian or schizophrenic. It is for the latter reason that the instructions on judging affect require the examiner to measure more than just facial appearance.

In the former instance, the patient has a lesion in the corticobulbar pathway between the motor cortex and the pons. In the latter, the lesion is in a more anterior frontopontomedullary connection.

A person who can smile but not whistle might also be accused of malingering. But this is exactly what is seen as an early manifestation of progressive muscular dystrophy of the facioscapulohumeral type (Landouzy–Dejerine), in which the seventh cranial nerve is intact but the orbicularis oris is asymmetrically involved (Perkoff and Tyler, 1953).

Bell Palsy

Bell palsy, shown in Fig. 26-9, is a subset of the peripheral variety of facial palsies, referring to lesions distal to the geniculate ganglion (DeJong, 1979).

Hunt syndrome, which is herpes of the geniculate ganglion, produces a peripheral facial palsy and herpetic vesicles on the eardrum. This situation is the only one in which it is useful to know that the eardrum, parts of the ear canal, and parts of the tragus are supplied by the somatosensory branch of cranial nerve VII. The actual distribution varies from person to person.

A Method

The best test[4] for Bell palsy is to ask the patient to close each eye, individually, in turn. In very mild cases, the patient will not be able to close the eye on the afflicted side without also closing the contralateral (normal) eye.

Two Caveats. If you do not know the history, merely observing the patient's facial appearance, especially around the eyes, can be misleading. With recovery, partial contractures may set in on the paralyzed side, making it appear to be the more contracted side. Hence, the normal side may appear comparatively paretic.

Similarly, a third nerve lesion can produce upper lid ptosis (mimicking the contractures mentioned above), but not the lower lid droop seen in cranial nerve VII lesions.

During an eye blink, the lower lid normally moves 2 to 5 mm horizontally and nasally, thereby producing a partial vacuum in the lacrimal system that helps to clear it and to wipe the tears away from the globe. However, in facial palsy, the lateral motion is lost, and in fact the lower lid does not move up much at all. This is best demonstrated with a camera, but the effects can be seen without it (Arrigg and Miller, 1985). The tears build up on the affected side. Because they form a stagnant pool, they can permit bacterial overgrowth and infection.

The Bell Phenomenon

Ordinarily, when one attempts to close the eye against a resistance by contraction of the orbicularis muscle of the eyelids, the globe rotates upward. This is not pathologic but rather is a normal synkinesia (associated movement). Hence, we speak of the "Bell phenomenon," not the "Bell sign." Usually, it is not noticed because the closing eye obscures the globe.

A Method

Ask a naive medical student to try to keep his eyes closed while you try to open them by opposing them with your thumbs. Press extra hard on one side only to keep that eye from closing, and you will see the Bell phenomenon. On the other side, you must let the eyelid win over your thumb, or your subject may visually fix on a spot as he concentrates on his task. Such visual fixation can eliminate the Bell phenomenon bilaterally.

[4] Because of the extremely large number of talented people who have worked in the field of neurology over the past century, we are blessed with a surfeit of neurologic tests for each lesion. Accordingly, we will often try to pick one simple test and ask the reader to consult DeJong for a fuller catalog.

FIGURE 26-9 This drawing of a Canadian (Ojibway) Indian mask called "Broken Nose" demonstrates the classic features of Bell palsy: loss of forehead wrinkling, deviation of the mouth to the nonparalyzed side, sagging of the paralyzed side with loss of the nasolabial crease, and the rounder eye suggesting lagophthalmos (failure of the upper eyelid to move down when the patient is asked to close his eye). (Drawing by William Snavely, with permission.)

Interpretation. The Bell phenomenon may be significant only when it is absent. (Such phenomena are often likened to Sherlock Holmes's barking dog.[5])

Bilateral absence occurs in (a) visual fixation by a neurologically intact person, (b) brainstem or lower motor neuron oculomotor disease, but not supranuclear oculomotor disease, (c) bilateral disease of the third cranial nerves (unlikely by Occam's razor), and (d) 15% of healthy persons (Adams and Victor, 1985).

The phenomenon is absent unilaterally in ipsilateral third cranial nerve disease (not seventh).

In Bell palsy, whenever the orbicularis muscle closes, it must do so against an effective "resistance." Because of this, it is particularly easy to observe the unmasked (normal) Bell phenomenon on the afflicted side (DeGowin, 1965; Delp and Manning, 1975). This has led to the unfortunate conclusion that the Bell phenomenon is a "sign" of Bell palsy. This convoluted reasoning is reinforced by the similarity in names and the unhappy practice of describing both palsy and phenomenon in the same physical diagnosis textbook passage. This emphasis on one situation in which the phenomenon is present has unfortunately detracted from its more salient use in those situations—outlined above—in which it is absent.

Because it is a normal synkinesia, the Bell phenomenon has been used by others to distinguish an oculomotor conversion reaction from "organic" pathology. (If the synkinesia occurs, the innervation of the eye muscles must be intact; therefore, the patient is "malingering" if unable to move his eyes on command.) However, there are problems with relying on this phenomenon alone because, if the phenomenon persists, the patient might actually have supranuclear disease.

The Bell phenomenon has also been confused in some texts with the Negro sign, the movement of the globe up and out when the patient looks up at the ceiling. The Negro sign is present in both central and peripheral facial palsies. The Negro sign, like the Bell phenomenon, is more pronounced on the side of the lesion.

Historic Note: The Bells of Scotland

Both Bell palsy and the Bell phenomenon were named for Sir Charles Bell (1774–1842). However, this was not the Bell whose clinical acumen so impressed the medical student Arthur Conan Doyle that he became the model for Sherlock Holmes (Doyle, 1958). The latter, Joseph Bell, was a scion of the other Scottish family of eminent surgeons, also named Bell.

[5] Just as Willie ("the Actor") Sutton never actually authored the words of Sutton's law (Chapter 27), so Holmes never referred to the dog as the "barking dog." In the adventure called *Silver Blaze*, Watson asks Holmes:

"Is there any point to which you would draw my attention?"

"To the curious incident of the dog in the night-time."

"The dog did nothing in the night-time."

"That is the curious incident." remarked Sherlock Holmes.

Later, Holmes explains, "...I had grasped the significance of the silence of the dog, for one true inference invariably suggests others. The Simpson incident had shown me that a dog was kept in the stables, and yet, though someone had been in and had fetched out a horse, he had not barked enough to arouse the two lads in the loft. Obviously the midnight visitor was someone whom the dog knew well."

Thus, this should be called the *silent dog*, not the barking dog (as Sutton's law should be named after William Dock, who actually described it).

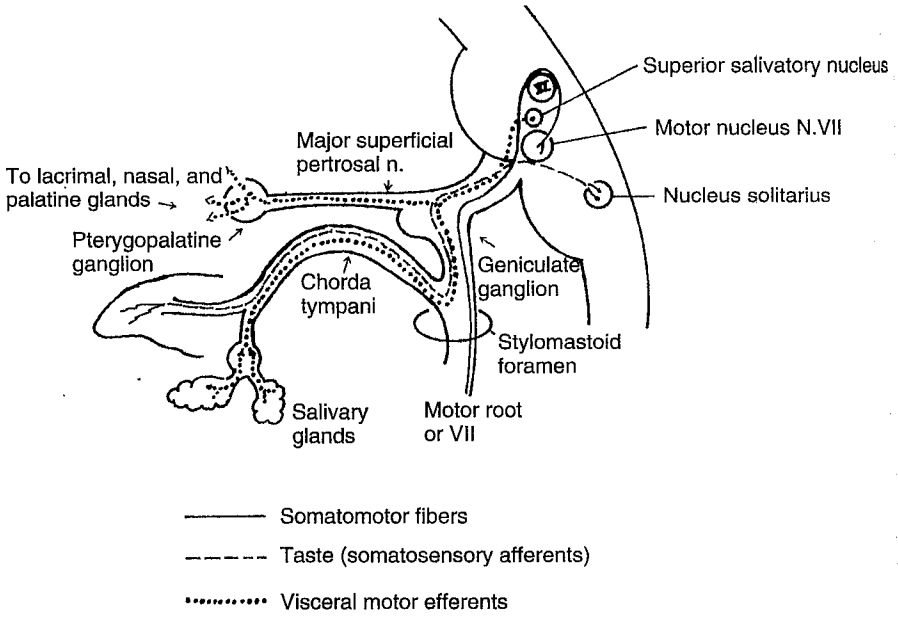


FIGURE 26-10 The branches of cranial nerve VII.

Unilateral Hyperacusis

Unilateral hyperacusis may be caused by a palsy of the stapedius muscle, which normally functions to dampen the oscillations of the ear ossicles. For detecting this finding, one may use the tape recorder and earpiece described for interviewing patients who are hard of hearing (see Chapter 2). The two sides may be compared by noting the volume setting that elicits the acoustic reflex (a startle, wince, grimace, or blink in response to a sound sufficiently loud to be unpleasant). Pure tones and white noise appear to be the best discriminators, not music (Johns, 1986).

Cranial Nerve VII: Visceral Sensory

Taste is more complicated than smell because the anterior two thirds of the tongue is innervated by cranial nerve VII, the back of the tongue by cranial nerve IX, and the epiglottal taste buds by cranial nerve X. It is difficult to detect unilateral lesions or lesions affecting individual cranial nerves VII, IX, or X because of diffusion of the test substance or overlap of innervation (L. Huntoon, *personal communication*, 1998). Testing with sugar solution is very difficult because it tends to roll across the midline. Some examiners use only salt on the side of the tongue (S. Horenstein, *personal communication*, 1988).

Tests of taste are of little use in the routine neurologic examination but are included for use in patients with relevant problems.

A Method

The following technique may be adapted for use in lieu of a formal taste sensation laboratory:

1. Make up 0.25% aqueous solutions of table salt, table sugar, quinine (about one-half tsp per liter) and about 1 tsp per cup of kitchen vinegar. These solutions should be strong enough for an older person (who will have fewer taste buds) to taste. Remember, you are not testing for threshold but for total absence of taste.[6]
2. Write on a piece of paper the words "sweet," "salty," "sour," "bitter," and "plain water." (If the patient is unable to indicate his choice by pointing to one of the words, you may have trouble with the test. (a) If the patient is allowed to open his mouth to speak his answer, the solutions will run from one side to the other and from front to back, so the loss of taste on one side or the other, or in the area supplied by a specific cranial nerve, may be missed; (b) If the patient is allowed simply to signal "yes," the vinegar may stimulate cranial nerve V in the mouth and nose, and this sensation may be reported as a "positive" response even though the patient is not experiencing taste.)
3. Have the patient protrude his tongue, and paint on the solutions one at a time, in one place at a time, and on only one side at a time. To test cranial nerve VII, use the salt and sugar solutions on the anterior tongue. To test cranial nerves VII and IX, paint the sour solution (the vinegar) on the lateral surface, anteriorly for VII, and posteriorly for IX. To test cranial nerve IX, paint the bitter quinine solution posteriorly but do not let the patient swallow. (He will not be able to as long as his tongue is sticking out.) The quinine solution should be used last.
4. With the tongue still out, ask the patient to make a response.
5. After each response, let the patient rinse with some water, which should be expectorated, not swallowed.

Interpretation

Unilateral impairment of taste on the anterior two thirds of the tongue signifies a lesion of cranial nerve VII, located above its exit from the stylomastoid foramen, either in the portion that passes through the auditory canal (the chorda tympani) or in the portion that proceeds to the geniculate ganglion. Contrariwise, supranuclear lesions are not accompanied by alteration in taste (Fig. 26-10).

Causes of bilateral decrease or loss of taste are listed in Table 26.2.

Complaints of dysgeusia (as in gold therapy) or hypogeusia may actually have normal taste tests but abnormal olfactory nerve testing. In fact, olfactory problems are the most common cause of dysgeusia, a problem that seems especially prominent in otherwise normal postmenopausal women (M. Schlitt, *personal communication*, 1999).

[6] The median detection threshold/median recognition threshold, in mmol per L, for healthy subjects, is as follows: sodium chloride 12/30; sucrose 12/30; hydrochloric acid 3/6; urea 120/150 (Henkin et al., 1971).

26.2 *Causes of bilateral loss of taste or hypogeusia (decreased taste)*

Neurologic disease
Familial dysautonomia
Local afflictions
Facial hypoplasia
Sjögren syndrome
After radiation therapy
Laryngectomy
Drug therapy
Steroids
Diuretics
Aspirin
Nutritional deficiencies
Niacin deficiency
Zinc deficiency
Endocrine conditions
Cushing syndrome
Hypothyroidism
Diabetes mellitus (abnormal taste of glucose only)
Pseudohypoparathyroidism
Infectious diseases
Dengue fever
Influenzalike infections
Other systemic conditions
Sarcoidosis
Cancer
Chronic renal failure
Cirrhosis of the liver
Burns
Hypertension (abnormal taste of salt only)

From Clee MD, Burrow L, Delaney P, et al. Taste and smell in disease. *N Engl J Med.* 1983;309:1062–1063; Schiffman SS. Taste and smell in disease. *N Engl J Med.* 1983;308:1275–1279; and Wechsler IS. *Clinical Neurology.* 9th Ed. Philadelphia, PA: W. B. Saunders; 1963, with permission.

Cranial Nerve VII: Visceral Motor

The visceral motor branches of cranial nerve VII supply the salivary (Chapter 13) and lacrimal glands (see the Schirmer test, Chapter 10). Unilateral dryness of the eyes and mouth could result from a lesion of this nerve. If bilateral, the problem is probably end-organ damage as in Sjögren syndrome.

Cranial Nerve VIII

The auditory portion of cranial nerve VIII has already been tested with the Rinne, Weber, and Schwabach tests during the examination of the ear (see Chapter 11). And you will of course have already checked the ear for cerumen.

The simplest screening test, which has a fairly high sensitivity, is simply to rub one's fingers together or a few strands of hair between one's fingers, near the patient's ear, and compare the two sides (M. Schlitt, *personal communication*, 1999).

One aspect of the vestibular portion of cranial nerve VIII was tested during the examination of the extraocular movements, when the presence or absence of nystagmus was noted.

The findings from these two examinations might have been recorded under the respective regional examinations.

Additionally, screening tests for vestibular function, the Bárány test, and the caloric response are discussed later because the vestibular system is most easily tested as a system.

The Arbit Hearing Test

A device constructed of a stethoscope, a tuning fork (512 Hz), and a suction cup or feeding nipple (designed to attach the tuning fork to the stethoscope diaphragm) has been described for improved bedside testing of hearing (Arbit, 1977). One also needs a device such as a hemostat for clamping (occluding) the stethoscope tubing (Fig. 26-11).

A Method

1. Preinstruct the patient that he is to indicate when he is no longer able to hear the sound of the tuning fork.
2. Place the earpieces of the stethoscope in the patient's ears.
3. Strike the tuning fork, which is attached to the stethoscope diaphragm by the feeding nipple.
4. Place your ear close to the tuning fork and listen while watching the patient for the preestablished signal.

Hearing is said to be normal if the patient hears the sound at least 15 seconds longer than the examiner.

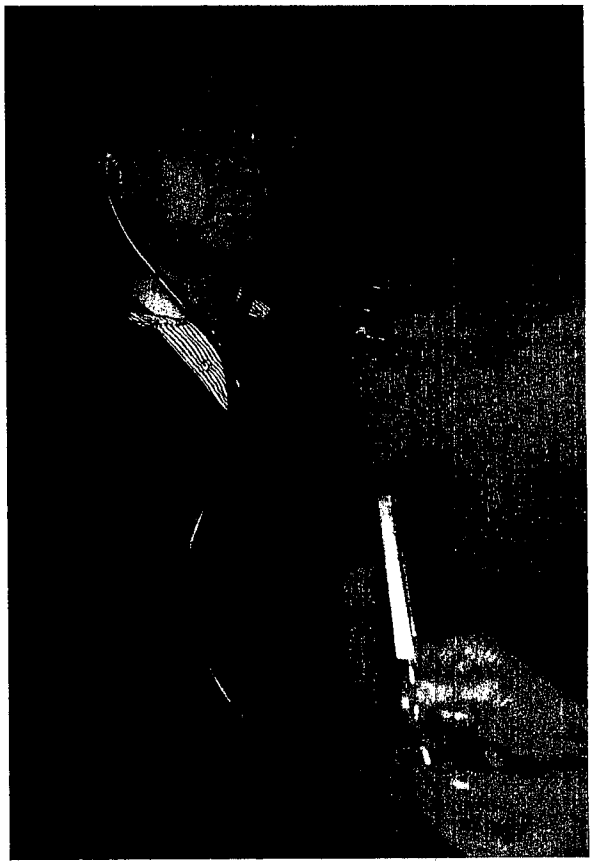


FIGURE 26-11 Set-up for Arbit test to check hearing in one ear at a time. The examiner needs to put her ear close to the tuning fork.

Comment. Although this test seems to work as well or better than the contemporary "customary" examination, it has not been compared with the Rinne and Weber tests.

A Unilateral Method. Test each ear individually by clamping the tubing leading to one ear. The patient again signals when he no longer hears the sound (in the ear served by the nonoccluded tubing). At that instant, unclamp the tubing and ask the patient if he now hears the sound. If he does, you have identified a definite hearing difference between the two ears.

Loudness Recruitment

The device described above can also be used to detect loudness recruitment by comparing the two ears at different tuning fork intensities (for details, *vide infra*).

If you could carry around an audiometer in your pocket and use it as an independent variable for testing your diagnosed patients with unilateral deafness, you would soon make an interesting discovery: Patients with unilateral sensorineural deafness due to the involvement of the hair cells of the organ of Corti would show very strange audiograms (Fig. 26-12).

The schematic audiogram demonstrating recruitment (*solid circles*) would not be seen in non–end-organ deafness (*squares*). Looking at the circles, we see that the infirm ear has a very high threshold. The difference between the good ear and the infirm ear decreases at higher decibels (louder sound). In fact, at very high amplitudes (sound intensities), the good ear and the bad ear hear equally well! The bad ear is thus said to have "recruited" something that permits it to be equal to the good ear at very suprathreshold intensities.

For the Attending. Recruitment is almost never found in sensorineural deafness due to involvement of the cochlear nerve (VIII) (i.e., its presence implies disease of the end organ rather than that of the communicating neural tissue). The exception has been the report of this phenomenon in a few cases of acoustic neurinoma (cerebellopontine angle tumor) (Alpers and Manchall, 1971). In a general medical practice, recruitment most often is a sign of Ménière disease.

A Bedside Test for Unilateral Recruitment

A Method

Hit the tuning fork very lightly, and rapidly present it to each ear in sequence. Repeat several times, each time with a lighter touch on the tuning fork, varying the ear that is stimulated first. The purpose is to search for evidence that threshold or near-threshold sound is significantly different between the two ears. This point is operationally defined as the softest touch on the tuning fork that will permit an ear to perceive the sound as softer (than the opposite ear) regardless of the sequence of presentation.

Next, hit the tuning fork as hard as you can and quickly present it to each ear. If the subject now reports that the infirm ear hears just as well as—or sometimes better than— the good ear, recruitment is said to be present (Chandler, 1958).

This test was developed by Chandler, who noticed that the patient who has recruitment will often give visual signals of discomfort when the maximally activated tuning fork is presented to the infirm ear.

Of course, the patient with nonrecruiting sensorineural deafness, or one with a conductive deafness, will continue to report that the sound is much louder in the "good" ear, no matter how hard you hit the fork.

Obviously, the Chandler test for recruitment can be modified as by Arbit in that the clamped and unclamped tubing permits the

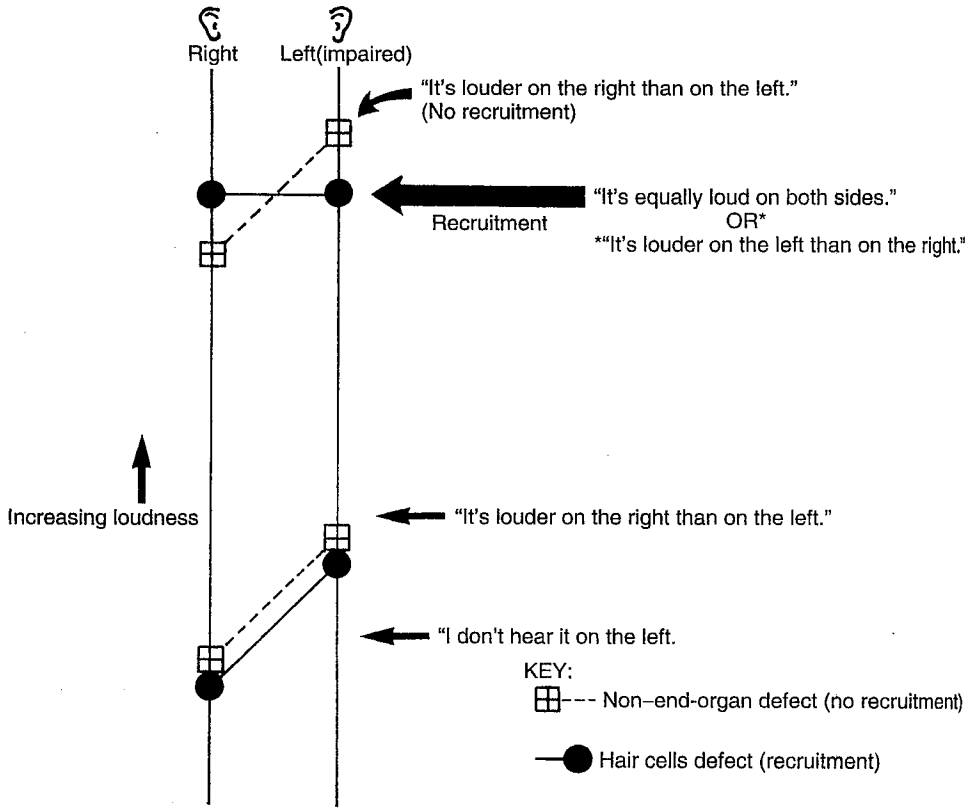


FIGURE 26-12 Loudness recruitment thresholds (see text).

same tuning fork to be presented to each ear in sequence without actually moving the fork around the patient's head.

For the Guru. We need to acknowledge that the distinction between sensorineural and conduction deafness derives from 19th-century concepts. They were based upon tuning fork tests and therefore are still convenient. However, with increasing information about the cochlear portion of the ear, exceptions to the old concepts have arisen (Goodhill, 1979).

More on Ménière Disease

Taking the History

The classic tetrad of Ménière disease is fluctuating hearing loss, episodic vertigo typically lasting 1 to 5 hours, tinnitus, and aural fullness. The tinnitus is often described as a low-frequency roaring. The aural fullness may be triggered by high-salt meals or caffeine, although the patient may not have noticed the connection. Thus, a low-salt diet may be helpful. The classic tetrad does not always exist at initial presentation.

Ask the patient: "Does the vertigo start 'right out of the blue' when you are seated and doing nothing, or does it come on with sudden movement of your head or body?" With Ménière disease, the vertigo seems to occur unprovoked, whereas with benign paroxysmal positional vertigo (BPPV), it tends to be brought on by rolling over in bed, lifting dishes to the top shelf, turning to check traffic, or picking something up.

Also ask: "Is the spinning fast or slow?" Patients with Ménière disease or BPPV generally spin fast, whereas migraineurs and patients with anxiety spin slowly.

Low-Frequency Hearing Loss

Ménière disease should not be diagnosed without vestibular signs (*vide infra*) and low-frequency hearing loss. One way to detect the latter is to see whether the patient can hear the dial tone on the telephone. This test works all over the world, except in Vienna, where the dial tone is the 440–Hz A to which the Vienna Philharmonic tunes up (S. Horenstein, *personal communication*, 1988). Low-frequency tuning forks (such as 128 Hz) can be used to demonstrate this at the bedside.

Patients may describe *diplacusis*—a distortion of pitch or perception of pitch difference from side to side. Check for this by striking the tuning fork, presenting it to alternate sides, and asking whether the "pitch" (not volume or timbre) is the same in both ears. Some patients will not be able to understand this concept, but many perform the test very well. The pitch in the infirm ear will generally be described as "lower."

Syndrome of Acoustic Neuroma (Vestibular Schwannoma)

The first major neurologic abnormality is auditory, although the tumor arises from the vestibular branch of the cranial nerve VIII (Patten, 1996). Hearing loss is found in 98% of these patients, tinnitus in 70%, disequilibrium in 67%, and nystagmus in 26% (Harner and Laws, 1983).

On vestibular testing (see later in this chapter), more than 80% of patients will have at least some decrease on one side.

An eighth-nerve tumor, especially if bilateral, suggests the diagnosis of neurofibromatosis type 2 (Karnes, 1998).

Cranial nerves V and VII may also be involved: 26% to 29% of the patients have facial numbness, and 10% to 12% have facial weakness. Either dysgeusia or lingual numbness from involvement of chorda tympani fibers is found in 6%. An abnormal corneal reflex is found in 33%.

It is essential to check the corneal reflex because loss or depression of this reflex is the most consistent early sign. The fifth nerve is lifted up by the tumor and the afferent fibers for the corneal reflex seem especially sensitive to such distortion. Numbness over the face may appear later (Patten, 1996).

Only about 10% have diplopia, and abnormal eye movements are found in 11% (owing to involvement of cranial nerves III, IV, and VI).

Cranial nerves I, II, IX, X, XI, or XII are collectively and individually involved, but rarely. (Rarely means <1% of the time.) Abnormalities of these cranial nerves, especially in a discontinuous fashion (i.e., not in numeric sequence), should suggest an alternative diagnosis.

As with other retrocochlear diseases, there is a disproportionate impairment in speech discrimination, as opposed to pure-tone audiometry.

With present imaging techniques, especially gadolinium-enhanced magnetic resonance imaging (MRI) and temporal bone scans, acoustic neuroma is diagnosed so early that the classic picture is no longer seen, just as brain tumors no longer present with papilledema (S. Horenstein, *personal communication*, 1988).

The acoustic nerve tumor is the most common type to occur in the cerebellopontine angle. Other tumors and lesions involving this area include meningioma; glomus jugulare tumor; cholesteatoma; eccentric pontine glioma; neuromas of cranial nerve V, VII, or IX; chordoma; astrocytoma of cranial nerve VIII; arteriovenous malformation; aneurysm of the intrapetrous carotid artery; chondrosarcoma; nasopharyngeal carcinoma; and metastatic deposits of carcinoma or lymphoma. Early involvement of cranial nerve VII suggests one of the alternate types, being quite rare in acoustic neuroma (Patten, 1996).

A Superspecialist Uses Tuning Forks

Dr Michael J. A. Robb of Phoenix, Arizona, a neuro-otologist, writes that he uses tuning forks ranging in frequency from 128 to 4,096 Hz at the bedside. Hearing can be checked using the 256 to 512 Hz forks for the Weber, Rinne, and Schwabach tests (see Chapter 11), and air conduction thresholds over five octaves can be checked quickly with the 128 to 4,096 forks, provided that the examiner has normal hearing and is willing to practice. Dr Robb reports that he can predict the shape and degree of hearing loss over five octaves to within 15 dB using six different tuning forks. The tuning forks also provide clues to hyperacusis, misophonia, migraine-related phonophobia, and diplacusis (M.J.A. Robb, *personal communication*, 2004). Formal audiometric testing should also be obtained. If done during symptomatic and asymptomatic periods, this is especially helpful for demonstrating the fluctuating hearing loss that makes Ménière disease more likely.

Cranial Nerves IX and X

Where accessible to the physical diagnostician, cranial nerves IX and X overlap so much that they can be considered together.

A Method for the Beginner

Examine the palate to see whether the uvula is in the midline position. In asymptomatic patients, it usually is.

Also perform a gag reflex at the conclusion of the examination of the throat. Simply let your tongue depressor slide back toward the posterior third of the tongue. In all likelihood, you will have already produced many gag reflexes inadvertently by the time you are polished enough to produce one intentionally. Accordingly, by the time you do that, you will know what it looks like.

The Uvula in Pathologic Conditions

A great deal can be learned from the position of the uvula in pathologic conditions. In Fig. 26-13, each row refers to a pathologic condition, except for the top row, which illustrates the normal. Each column represents a condition of examination. The figures in the left-hand column are the palate and uvula as observed at rest. The second column is the result of following the command to say "ahh."

If the individual has palsy, the uvula will be deviated toward the good (healthy) side. Please note that the uvula may not exactly point to the good side (as shown in the figure) but rather may seem to be hanging straight down on the good side of the midline.

In some cases of partial palsy, the uvula will hang in the middle, but on induced phonation ("ahhh") will move toward the good side. Again, in actual practice the whole area moves. The uvula again does not really point at the good side as shown in the figure, which is intended as a mnemonic.

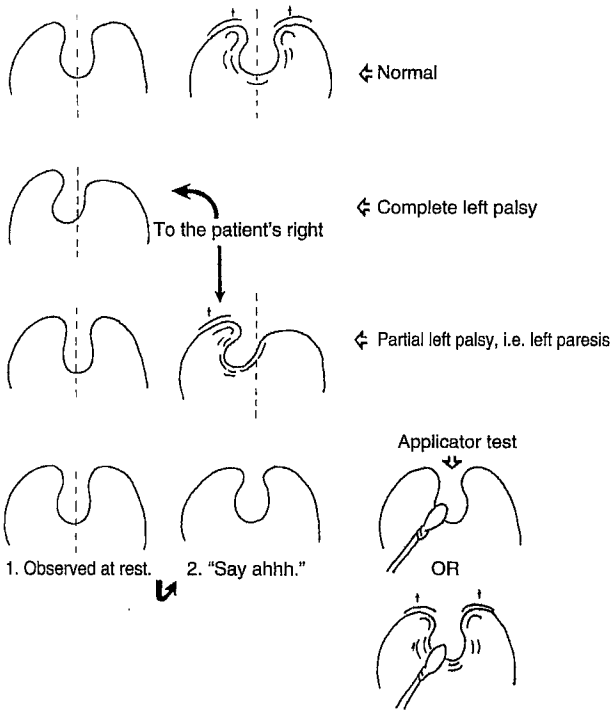


FIGURE 26-13 The uvula in various pathologic conditions. **Left column:** At rest. **Middle column:** On phonation ("ahhh"). **Right column:** Applicator test. **First row:** Normal. **Second row:** Complete left palsy. **Third row:** Partial left palsy (i.e., left paresis). **Fourth row:** Lower motor neuron disease of cranial nerves IX and X. **Fifth row:** Upper motor neuron disease of cranial nerves IX and X.

Sometimes the uvula is in the midline but does not move on phonation. Here we must be dealing with a bilateral paralysis. But does it result from upper or lower motor neuron disease? To find out, simply touch a cotton applicator to the uvula. If it does not move, one is obviously dealing with bilateral lower motor neuron palsy. If, as in the bottom panel, it does respond (bilaterally) to direct stimulation, one is dealing with bilateral upper motor neuron palsy.

Notice that we have not said whether IX, X, or both cranial nerves were involved in this palsy. This is because they usually go together. The most common causes seen in practice are involvement of the IX and X cranial nerves in brainstem disease. Sometimes, there is unilateral involvement due to the syndrome of the jugular foramen. In this instance, one also sees homolateral involvement of XI in addition to IX and X (syndrome of Vernet, *vide infra*).

Distinguishing Lesions of IX and X

If one wishes to separate lesions of IX and X, one is obliged to find a testable function served by one but not the other.

Although cranial nerve IX does supply taste to the posterior third of the tongue, it should be noted that cranial nerve X supplies the taste buds even further back. Given the vagaries of testing for taste in general, and without touching the anterior two thirds of the tongue in particular, the distinction can probably not be made on these grounds.

However, cranial nerve X does have some accessible and testable functions. Old-time neurologists used to perform the *oculocardiac reflex*. This was done by pressing on the eyeball and observing the slowing of the heart. If the visceral motor branches of X were interrupted, no such slowing would take place. (However, interruption of the afferent fibers of V also leads to loss of the reflex.) If one wishes to slow the heart rate without using drugs, this method may be safer than carotid massage (see Chapter 18). Demonstrating the absence of this reflex is required by some before declaring brain death.

Cranial nerve X also has somatosensory branches that supply the anterior aspect of the tragus, sometimes the posterior aspect of the tragus, sometimes a little spot of skin on the back of the ear, and sometimes the external auditory canal. Unfortunately, IX also supplies these areas in some persons. The best place to search for an anesthetic spot is on the tragus. If you find it, you have fair evidence of involvement of cranial nerve X.

Finally, X supplies somatomotor branches to the vocal cords.

Vocal Cord Paralysis

For the Attending. The following discussion presumes that you have performed indirect laryngoscopy or have consulted a laryngoscopist.

Total bilateral loss of cranial nerve X is usually associated with a fatal outcome, although the loss of X itself is not fatal, because both sides of the medulla oblongata must be damaged for a long distance in order to destroy the entire nucleus of X. A person can live with bilateral paralysis of the recurrent laryngeal branches of X, although he will have dyspnea and impaired phonation. If there is bilateral disease of the adductor branches, there will be impaired phonation but no respiratory difficulties. If there is bilateral paralysis of the abductor branches, there will be no difficulties with phonation

to speak of, but there will be severe dyspnea. The last is known as Gerhardt syndrome.

Unilateral laryngeal nerve palsies are a little more common. Unilateral recurrent laryngeal nerve palsy is associated with a raspy voice that easily fatigues. Its differential diagnosis is to some degree dependent on the laterality. If the left recurrent laryngeal nerve is affected, one should think of aortic aneurysm, trauma (e.g., thyroidectomy), mitral stenosis, or other causes of left pulmonary artery enlargement. If the right recurrent laryngeal nerve is involved, one should look for disease of the right pulmonary apex. Recurrent laryngeal nerve palsy of either side may be caused by pericarditis, goiter (Hamburger, 1986), tumors, and (rarely) tabes dorsalis. The most common cause of recurrent laryngeal nerve palsy is postsurgical, with operations on the anterior half of the neck, especially in the 20% of individuals who have an anomalous recurrent laryngeal nerve (M. Schlitt, *personal communication*, 1999). Unilateral superior laryngeal nerve palsy is usually the result of trauma.

Vocal cord immobility in patients with rheumatoid arthritis has been assumed to result from cricoarytenoid arthritis with joint fixation. Cervicomedullary compression (*vide infra*) should also be considered (Thompson et al., 1998).

Cranial Nerve X in Combination with Other Cranial Nerve Lesions

All of these are homolateral syndromes. When nerves IX, X, and XI are involved together, that is the syndrome of Vernet and indicates disease at the jugular foramen, if unilateral. The syndrome of Vernet plus involvement of cranial nerve XII is Collet–Sicard syndrome, due to the involvement of the posterior laterocondylar space with tumors of the parotid gland or carotid gland, tuberculous adenitis, lymphoma, or secondary tumors. The additional involvement of the sympathetics is known as the syndrome of Villaret and indicates that the lesion is in the retroparotid space, if unilateral. However, if only nerves X and XI are involved, that is the syndrome of Schmidt and should be attributed to disease of the central nervous system (rather than the peripheral nerves) until proved otherwise.

In a patient with rheumatoid arthritis, symptoms or signs related to cranial nerves IX-XII should trigger concerns about brainstem compression from "cranial settling" (*vide supra*). Sudden death can result (Menezes et al., 1985; Bouchaud-Chabot and Lioté, 2002).

Cranial Nerve XI

The trapezius muscles may be tested by having the patient shrug his shoulders. The sternomastoid muscles may be tested by having the patient rotate his head away from the side being tested (when facing you) against the resistance of the examiner's hand placed on the lateral aspect of the mandible opposite the side being tested (Fig. 26-14).

1. The supranuclear control of the spinal part of the nucleus of cranial nerve XI is unusual in that the hemisphere exerts control over the sternomastoid muscle on the same side and the trapezius on the opposite side. The functional importance is obvious: the right hemisphere activates the left limbs and also the sternomastoid that turns the head toward rather than away from the left. In patients experiencing a focal motor seizure, the head turns toward the convulsing limbs, as would be expected from this innervation pattern (Patten, 1996).

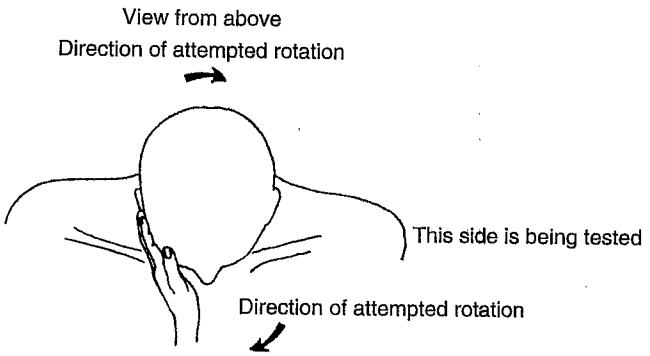


FIGURE 26-14 Testing the sternocleidomastoid. (See text.)

2. The function of the spinal accessory nerve may also be impaired by neck lesions that cause entrapment.
3. The most common cause of nerve XI palsies is surgical mishap in the posterior triangle of the neck (M. Schlitt, *personal communication*, 1999).
4. *Weakness of neck forward flexion is found in 100% of myopathies of any cause.* Do not test the side-to-side or rotational strength. Instead, tell the patient to put the chin on the chest and keep it there while you try to extend the neck (forcing the head up and back). The absence of sternomastoid weakness as demonstrated by this maneuver calls into question the diagnosis of myopathy (S. Horenstein, *personal communication*, 1988).

Cranial Nerve XII

A Method

Instruct the patient to stick his tongue out at you and then, as you lightly hold his mandible, ask him to point the tongue first to the right and then to the left.

In the *acute* stage of lower motor neuron (peripheral or nuclear) hypoglossal paralysis, the tongue points toward the paralyzed side; the paralyzed side may *appear* higher and more voluminous (the Dinkler sign, Fig. 26-15). This may also be seen in *acute* supranuclear hypoglossal palsies.

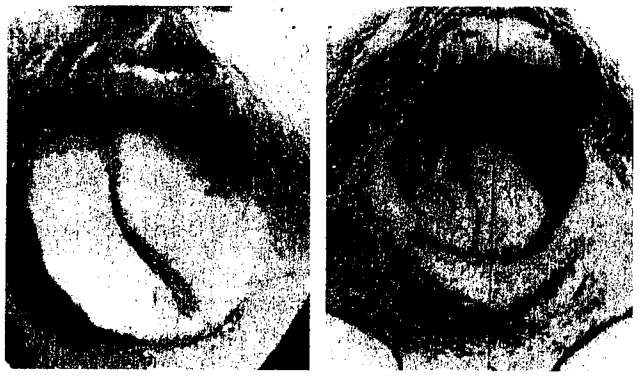
The Dinkler sign is not seen in acute malingering or acute conversion reactions.

A false-positive Dinkler sign may be seen in facial nerve palsy. The tongue appears to be pointing to the side of the facial nerve lesion on casual inspection, but closer examination will reveal that the tongue is pointing straight out but the patient is unable to open his mouth completely on the affected side.

The Dinkler sign associated with pain is now usually the result of carcinomatous involvement of the peripheral nerve XII. It may also be caused by basilar meningeal carcinomatosis, usually from breast cancer in women and prostate or lung cancer in men.

An uncomplicated *chronic* unilateral cranial nerve XII palsy will not usually "point to the bad side" when the patient first protrudes his tongue. The tongue will usually come straight out of the mouth, at which time you should inspect and palpate it for atrophy of the diseased side. (The bulging deformity of the Dinkler sign will not be seen if atrophy has supervened.)

But the patient with chronic hypoglossal nerve palsy (just like the patient with acute hypoglossal nerve palsy) will not be able to



A

B

FIGURE 26-15 **Top:** When a patient with the Dinkler sign sticks his tongue out, it points to the bad side. Also, the acutely paralyzed left side seems to be bunched up (the Dinkler sign), while the normal right side is relatively flat. When the patient puts his tongue back in his mouth, both sides are flat. Notice also that the tongue goes in on the right side—that is, it is pulled back to the good side by the good muscle. **Bottom:** This patient is faking an acute hypoglossal nerve palsy. Notice that his tongue is quite flat, in contrast to the photographs on top. (From Alexander L. The neurologic examination. *Pullen's Medical Diagnosis.* Philadelphia, PA: W. B. Saunders; 1950, with permission.)

move his protruded tongue to the normal side of his mouth. Be aware that if commanded to point the tongue to the good side, some such patients will move their mandible and facial muscles so as to bring the normal angle of the mouth over to the midline tongue. (If the tongue cannot come to the angle of the mouth, the mouth will come to the tongue. That is why you *lightly* hold the mandible.)

If there is ipsilateral basal occipital bone tenderness in the region of the hypoglossal canal (through which nerve XII exits the skull), one is most likely dealing with neoplasia (especially prostatic in the male), even if the patient has already survived long enough to have chronic hypoglossal nerve palsy with hemiatrophy.

The (relaxed) tongue can be examined for fasciculations at rest (*vide infra*) as well as for myotonia (see Chapter 13). These findings, of course, have no specific significance related to cranial nerve XII.

For tongue signs of chorea, see later in this chapter.

The Cranial Nerve Examination and Differential Diagnosis

The importance of the cranial nerve examination is shown by the fact that this portion of the examination occupies almost one fourth of an excellent textbook on neurologic differential diagnosis (Patten, 1996). Careful attention to the cranial nerves, especially in elderly patients with comorbid conditions, can help prevent dangerous misdiagnoses in this age of overreliance on sophisticated neuroimaging and aggressive therapy such as thrombolysis. For example, two elderly patients with myasthenia gravis were recently given an initial diagnosis of stroke by a neurologist, with CT findings considered to be confirmatory. History and findings of ptosis, dysarthria, difficulty in handling secretions, diplopia, tongue weakness, and neck flexor weakness eventually led to the correct diagnosis (Kleiner-Fisman and Kott, 1998).

The laterality of bulbar signs as compared with limb signs helps to localize the lesion to the brainstem (ipsilateral signs) or higher (contralateral signs).

Skilled Acts

Aphasia and Other Disorders of Speech

> How could we arrest scientific and industrial progress? By closing down, or by controlling, laboratories for research, by suppressing and controlling scientific periodicals and other means of discussion, by suppressing scientific congresses and other conferences, by suppressing universities and other schools, by suppressing books, the printing press, writing, and, in the end, speaking. All these things that indeed might be suppressed (or controlled) are social institutions. Language is a social institution without which scientific progress is unthinkable because without it there can be neither science nor a growing and progressive tradition (Popper, 1964, p. 154).

Aphasia is a word coined by Trousseau, meaning loss of speech. Technically, most patients categorized as aphasic have some speech and thus are actually dysphasic, but the question of the extent of the abnormality is much less important than two other questions: (a) Does the patient actually have an abnormality of speech itself (i.e., word selection and understanding) or of something else (e.g., the mechanism for articulation) that is revealed in the process of speaking? (b) For those patients who truly do have aphasia, what is the specific type?

What Aphasia Is Not

Aphasia is not mutism, although patients with global aphasia may be mute. Patients with mutism are unable to make sounds, but language function is retained, as may be demonstrated (in literate patients) by reading and writing, two functions that are almost always impaired in the true aphasic.

One method of testing is to show the mute patient the following message, printed in large block letters: "Put your left thumb on your right cheek." Some aphasic patients will not be able to follow this written instruction but all literate mute patients can. Some patients with mutism can hear and thus can follow spoken instructions or respond appropriately to yes-or-no questions and questions permitting small number responses to be given by holding

up fingers. Usually, if one understands the distinction between aphasia and mutism, the diagnosis of mutism will have been made long before reaching the formal neurologic examination.

Aphasia is also not dysarthria. Dysarthria is the inability to articulate clearly, typically because of brainstem or posterior fossa lesions. A dysarthric patient does get the approximate sounds in the correct order in the correct words in the correct arrangement. Likewise, aphasia is not a dysphonia, a difficulty in phonation due to cranial nerve palsies involving VII, IX, X, or XII, or an intrinsic disease of one of the end organs of sound-making such as the nasopharynx or tongue. If one remembers the distinction between dysarthria or dysphonia and aphasia, one will usually have made the diagnosis early in the interview. If there is a question, remember that language function is intact in dysarthria and dysphonia, so the patient will be able to respond to written instructions, make nonverbal responses, and write linguistically coherent material, as described above in mutism.

Good repeat-after-me test phrases for dysarthria or dysphonia (but not always for aphasia, *vide infra*) are, "Methodist Episcopalian," "around the rock the ragged rascal ran," and "Peter Piper picked a peck of pickled peppers."

Aphasia is also not cerebellar speech, which is a dysarthria.

Some apparent abnormalities of speech that are actually abnormalities of mental status may also be detected during the interview. These abnormalities could conceivably be confusing to the sophomore who did not know the correct definitions of these types of speech. *Word salad* refers to a verbal output characteristic of some (but not all) schizophrenic patients. It sounds jumbled up, like a salad, in the sense that a salad is a mixture of small amounts of many things with no predominant theme. Such patients are quite fluent, but the content of their speech makes no sense. Word salad can be aphasia; however, with the schizophrenic patient, it is the thought content that is abnormal, as reflected perfectly in the language that is produced. Fortunately for the sophomore, the schizophrenic patients who produce word salad usually do so in the context of so many other peculiar actions and mannerisms that the diagnosis is not long in doubt (see later in this chapter for diagnostic criteria).

Manic patients may also demonstrate their disease through language. *Pressure of speech* refers to an accelerated rate of speech and a relative inability to be quiet; it is therefore almost never confused with aphasia. *Flight of ideas* is just that. The main content of the speech is difficult to follow, not because of language problems but because the main subject of the monologue changes so rapidly. *Tangentiality and circumstantiality* (see Chapter 2, footnote 1) could also be confused with aphasia by the jejune. However, within the context of the other abnormalities of mania (*vide infra*), the distinction should be obvious.

Depressed patients may produce a small quantity of speech, slowly and sometimes softly, but the grammar, syntax, and word choice are all normal.

Caveats

The student must realize that the perfect correlation between anatomic localization and abnormality of function that is seen in other areas, say with valvular heart disease or spinal cord disease, does not always exist for aphasia. Most patients will not present with one of the "pure culture" clinical pictures about to be described. The clinical picture resulting from etiologies as diverse as metabolic encephalopathies or brain tumors or Alzheimer disease may vary from day to day in the same patient, even though the anatomic substrate is presumably unchanged. Nevertheless, there are still useful concepts and methods of examination. Anatomic correlation also improves with time spent with the patient and the sophistication of the examiner.

A Method

The examination for aphasia and other disorders of speech (broadly conceived) is carried out in two parts. The first consists of observations of the usual doctor–patient conversation. Look for fluency (the rate of speech) and the latency to beginning speech (S. Horenstein, *personal communication*, 1988). The second, which is used to classify aphasias, is based upon the patient's response to specific instructions. The specific abstractions from conversations and the specific maneuvers including test phrases are presented throughout this section *ad seriatim*.

The first part of the examination is not in itself sufficient to rule out aphasia; active testing is necessary.

Fluent versus Nonfluent Aphasias

Having determined that the patient does truly have aphasia, one next wishes to determine whether it is fluent or nonfluent. (Some characteristics of various aphasias are summarized in Table 26.3.)

1. The nonfluent (also referred to as Broca, motor, frontal, or expressive) aphasia is characterized by spontaneous speech that is slow, labored, increased in latency, often mumbled, and lacking grammatical niceties such as definite articles or appropriate suffixes. It resembles what one composed in the days of 10 words per telegram.

   The nonfluent aphasia signifies a lesion in the Broca area (Fig. 26-16).

   The patient with bilateral frontal lobe isolation described below may at first appear to have a nonfluent aphasia.
2. Fluent aphasias are characterized by speech of normal grammar and at least normal speed but inability to use the correct words. If asked what class of objects comprises a Ford, a Cadillac, and a Toyota, the patient may use an indefinite ("you know, the thing"), an operational definition ("what you come to work in"), a paraphasia ("a horse"), or a paranymy ("a bar"). (Paraphasia has a better prognosis than paranymy. For example, if someone means to say "my sister," but says "my niece," he is at least within the correct class of objects; whereas the paranymy "my blister" is not even close.)

   (a) *Conduction Aphasia.* To test for conduction aphasia, ask *the patient to say, "No ifs, ands, or buts, please." Patients with* conduction aphasia understand the directions to repeat the test phrase because their comprehension is intact. But their attempted repetition is impaired, and they will stumble over this particular test phrase. Oddly enough, some of those who cannot do this test phrase can easily repeat some of the longer, seemingly more difficult phrases that have been used. In fact, I have had patients with large cerebral metastases in the area marked C in Fig. 26-16 who could initiate and maintain long goal-oriented

26.3 Some common kinds of aphasia

Aphasia type	Spontaneous speech	Comprehension	Repetition	Associated findings
Broca ("motor") aphasia	Nonfluent	Intact		Paresis of right face and arm
Conduction aphasia	Fluent	Intact	Impaired	
Isolation aphasia	Fluent	Impaired	Intact	
Wernicke ("receptive") aphasia	Fluent	Impaired	Impaired	Right superior quadrantanopsia
Nominal aphasia	Fluent	Intact	Intact	

From Geschwind N. Current concepts: Aphasia. *N Engl J Med.* 1971;284:654–656, with permission.

discussions (such as those concerned with obtaining cigarette privileges) without any evidence of error in their spontaneous speech but who could not repeat this phrase with any amount of practice.

(b) *Isolation Aphasia.* Patients with isolation aphasia are like parrots in that they can repeat any test phrase but do not understand what they are repeating. This ability may sometimes fool the examiner, but the aphasia can be detected by using a test phrase with a command in it, such as "put your left thumb on your right ear."

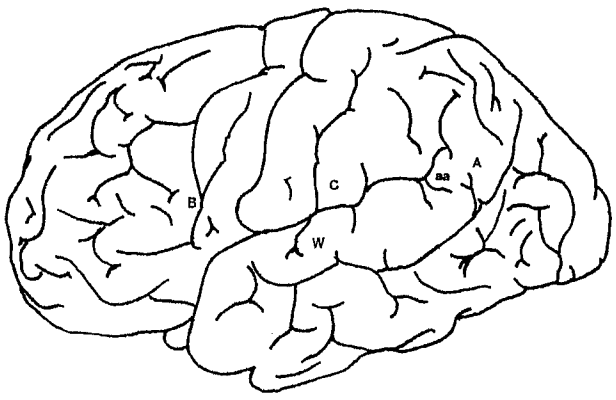


FIGURE 26-16 The speech areas. *B*, the Broca area, where lesions can produce a nonfluent aphasia. (Note the proximity to the motor area; you can see why afflictions of the Broca area are likely to be accompanied by hemiplegia.) *C*, the general area where lesions produce a conduction aphasia, a fluent aphasia in which the patient will attempt to follow the examiner's instructions, which he does understand. But he will not be able to repeat the test phrase, "No ifs, ands, or buts, please." Actually, conduction aphasia may occur with lesions in the C-shaped loop between B and W. *W*, the Wernicke area, where lesions may produce a fluent aphasia, in which the patient can neither understand nor respond to verbal repetition or other tasks. In the unhappy circumstance that both Wernicke and Broca areas are afflicted, the patient will have global (total) aphasia. *A*, the old "alexic" area where a lesion may leave competence in the spoken language but incompetence in selecting specific words or in writing, or Gerstmann syndrome. Region *aa* is one area associated with what was formerly called *amnestic aphasia*, or anomic aphasia in the old classification. Regions *A* and *aa* may overlap.

These patients have lesions that surround but do not involve either the Broca area or the Wernicke area. Isolation aphasia is very rare.

(c) *Wernicke (Receptive, Sensory, or Temporal) Aphasia.* Patients with Wernicke aphasia are unable to comprehend the instructions and are also unable to repeat the instructions. Thus, they fail to produce any test sentence on command.

Additionally, if given verbal instructions and asked to nod yes or no as to whether they understand the instructions, patients with impaired comprehension (as well as those with isolation aphasia) do *not* nod affirmatively. They just stare at the examiner. It is easy to see why this was formerly called *receptive aphasia* or "word deafness," although such patients are not deaf. (This can be demonstrated by continuing to speak as you walk in a circle around the patient and observe his head movements.)

(Another way of testing for comprehension is to give verbal instructions or instructions printed on a piece of paper—for example, "Tear this piece of paper in four parts; given me one, *put one on the table, and keep two for yourself*, one in each hand." Such instructions require no use of speech in the response. If the patient cannot comprehend, he will not be able to follow the written instructions.)

These patients have lesions in the classic Wernicke area as shown in Fig. 26-16.

(d) *Nominal Aphasia.* This type of aphasia is most often missed because the patient has intact comprehension and repetition and so passes all the tests above. But, like all aphasic patients, the patient with nominal aphasia has trouble naming objects. Sometimes, a rather long list of objects must be presented. Some neurologists use objects of increasing rarity, such as a wristwatch, the strap, the buckle, and, finally, the part of the buckle that fits into the leather hole. Hardly anyone knows the name of the latter (the tongue of the buckle). Thus, it permits testing for neologism: "Make up a word to describe this." (Aphasic patients cannot make up words.) "Pin" would be a good neologism. "Din" would be paranymy.

These patients may have lesions located at the angular gyrus or the adjacent portion of the temporal lobe or sometimes at

other places (Fig. 26-16). Sometimes there is no focal lesion but rather a metabolic encephalopathy. Occasionally, this type of aphasia occurs during recovery from one of the other types of aphasia. Additionally, naming problems are seen in the dementia of Alzheimer disease, in Huntington disease, and, to a lesser extent, in Parkinson disease (Frank et al., 1996).

You can see why the classification of aphasias has occupied the best minds of neurology for a long time, with a perfect solution not yet in sight.

Cerebral Dominance

Much has been made about determining handedness in the aphasic patient who has suffered a stroke because more than 99% of right-handed persons have their language function in the left cerebral cortex (Baker and Joynt, 1986; DeJong, 1979). It is said that about 50% of practicing lefthanders also have their language function in the left cortex, and a small percentage of these sinistrals may even have bilateral representation. It is also said that essentially all human beings are left-hemisphere dominant or perhaps codominant (M. Schlitt, *personal communication*, 1999). In any case, the value of determining handedness, if the patient cannot tell, may be purely academic.

There is some evidence that with nonalphabetic languages like Chinese, language function may reside in the right cortex of a right-handed person, although if such persons learn English or another alphabetic language, the language function for the alphabetic language resides in the left cortex of a right-handed person. To show the severe limitation of all the "rules," we have the situation of the Japanese language, which includes both alphabetic and morphogrammatic letters (the latter called *kanji*). These letters are learned in school at the same time and are used together in many kinds of printed matter. Yet, when a 32-year-old dextral Japanese woman suffered a proved hemorrhage of the left posterior inferior temporal gyrus, she lost her ability to read the kanji but retained her ability to read the alphabetic letters and words (Kawamura et al., 1987).

Agnosias

The agnosias are disorders in which the patient cannot recognize common objects nor tell their meaning.

Usually, only astereognosis is specifically tested for. (Stereognosis is the power of judging the form of an object by touch.) Common objects are placed in the patient's hand without allowing him to see them and he is asked to identify them. Examples include pens, keys, and coins. It is not necessary for the patient to be able to tell the exact denomination of the coin.

Apraxias

Apraxia is the inability to perform a task despite intact functional areas, and by tradition it refers to nonverbal tasks—for example, getting dressed.

There are two kinds of apraxia: ideational or ideomotor (due to posterior hemispheric lesions) and kinetic (due to frontal lesions). To distinguish them, take out a wooden matchbox and ask the patient to strike a light. Patients with ideational apraxia will not be able to slide the box open. (They cannot dial a telephone either.) They may rip it open to get to the matches inside. Patients with kinetic apraxia will go to strike the match but will hold it wrong so that it does not light or so that it breaks (S. Horenstein, *personal communication*, 1988). A patient with ideomotor apraxia may be able to perform a task spontaneously in certain contexts even though unable to do it on command.

A progressive deterioration of cognitive function, resulting in aphasia, agnosia, and apraxia, is seen in Alzheimer disease in the absence of prominent motor signs (Geldmacher and Whitehouse, 1997).

For further descriptions of agnosia and apraxia, see the section on "Parietal Lobe Syndromes."

Meningitis

Stiff Neck

A stiff neck that is resistant to passive motion may be the first physical sign of meningitis.

This sign, however, can also be present in rheumatoid arthritis, cervical osteoarthritis, polymyalgia rheumatica, and a wide variety of other cervical afflictions of the bones, joints, ligaments, and muscles of the cervical area. Thus, if the neck is supple, one can (usually) be reassured of the absence of meningitis—*except in the elderly, who may present with confusion and develop nuchal rigidity only when moribund*. But if the neck is stiff, the problem has still not been localized to the meninges. These days, the term "meningismus" may be used in neurology to signify a stiff neck from any cause.

Do not check for meningitis by flexing the neck in an unconscious patient unless head and neck trauma has been excluded. Additionally, be aware that meningismus can be a sign of a posterior fossa mass lesion, and flexion of the neck could provoke a foramen magnum herniation (M. Schlitt, *personal communication*, 1999).

The Kernig–Lasègue and Brudzinski Signs

History

Lasègue wrote a classic article on sciatica in 1864 without mentioning limitation of straight-leg raising. It is apparent that the authors of leading textbooks of neurology, which cited this article as the source of the Lasègue sign, had never read it (Wartenberg, 1956). The Kernig–Lasègue sign was first described in 1880 by a Yugoslav physician, Lazarevic (Clain, 1973). He has never received credit for it. His sign is known by two other names to two different groups of specialists who use it for different purposes. (Having encouraged the student to describe something original in the hope of eponymous immortality, it must now be recalled how many times the original innovator's name is lost to posterity. As Shaw noted, virtue is its own punishment.)

The next year, 1881, the sign was published by Forst, in French, in his MD dissertation. Forst credited his teacher Lasègue with having taught him the sign (Wartenberg, 1950).

In 1882, Kernig published the same test in a Russian journal, and he did it again in German in 1884. Up until this point, it is not clear that any of the authors knew of each other's work or that they realized they were all describing essentially the same sign.

jars. The glabella tap sign has a positive likelihood ratio (LR) of 4.5 and a negative LR of 0.13. A softer voice has a positive LR of 3.4 and a negative LR of 0.45. Various LRs have been tabulated (Rao et al., 2003). Not all symptoms and signs of parkinsonism have been studied from an "evidence-based" statistical perspective. Cogwheel rigidity, for example, is not included by Rao et al.

Drug-Induced Movement Disorders

Obviously, any drug that can block striatal dopaminergic receptors can result in an imbalance between the dopaminergic and cholinergic systems in such a way that the dopaminergic system is relatively defective. Thus, dopaminergic blockers, including but not limited to the phenothiazines, can acutely produce a picture that mimics parkinsonism. The cure is obviously to inhibit the cholinergic system by the administration of an anticholinergic drug.

Technically, the above movement disorder could be called a *dyskinesia*, which simply means an abnormality of movement. However, dyskinesia has been adopted for another movement disorder, *tardive dyskinesia*, which is biochemically the opposite of drug-induced parkinsonism. It tends to come on in patients who have been on chronic dopaminergic blockade, which is being withdrawn or diminished. (Sometimes, the dosage has not been changed but the dopaminergic system just seems to age and to exhibit tachyphylaxis. Often, these patients are also receiving chronic anticholinergic medication.) At this point, tics and dyskinesias of various types suddenly emerge. In this case, the treatment is to increase dopaminergic blockade (the "hair-of-the-dog-that-bit-you" approach) and/or to increase cholinergic activity—the exact opposite of the approach to the drug-induced parkinsonism discussed above.

Because these two movement disorders have exactly opposite biochemical causes and treatments, a word about nomenclature is in order. "Tardive" means late or after a long period of treatment. It is used to highlight the fact that tardive dyskinesia (which can almost be thought of as dopaminergic-blocker withdrawal) comes on after a long period of time.

Similarly, "dyskinesia" can and should be modified by indicating the muscle groups involved. The classic flycatcher dyskinesia, wherein the tongue darts in and out of the mouth like that of a frog catching flies, should be called a *buccolingual dyskinesia*; if it is a *tardive* dyskinesia, this word should be added to the description.

"Dyskinesia" by itself does not imply a specific etiology and it should not imply only a facial or lingual dyskinesia. For instance, some patients with tardive dyskinesia can present with chest, abdominal wall, or diaphragmatic spasms.

In any patient presenting with a movement disorder, a careful drug history is mandatory. A number of drug-induced movement disorders are summarized in Table 26.6. Consider prescription drugs, illicit drugs, and potential interactions. Cocaine use is a major risk factor for the development of acute neuroleptic-induced acute dystonia (van Harten et al., 1998). The "crack dance," well known to drug abusers, is seldom reported to physicians (Diederich and Goetz, 1998).

Not all dyskinesias in patients receiving drug therapy are caused by the medication. A review of case records of neuroleptic-naive patients showed abnormal involuntary movements in up to 28% of schizophrenic patients. Definite dyskinesias were described in 14.9% of 94 schizophrenic patients compared with only 1.7% of nonschizophrenic patients. Oral-facial movements were the most common type, being seen in 19% (Fenton et al., 1997).

Coordination

Posterior Column Signs (or Signs Potentially Related to the Posterior Columns)

The posterior columns carry the tracts conducting vibratory and position sense up to the higher centers. Therefore, an examination of the posterior columns' integrity really begins with the peripheral examination of vibratory and position (proprioception) sensations (*vide infra*).

Other tests of the posterior columns, especially of proprioception, may also test cerebellar and vestibular functions. A general principle is that if the patient can perform the task with his eyes open (visual cues available) but not with his eyes closed, it is a sign of impaired proprioception (posterior column disease); but if the individual cannot perform the test even with his eyes open, it is a sign of vestibular-cerebellar dysfunction.

The importance of position sense, taken for granted, is only appreciated if it is lost; see the tale of the "disembodied lady," who could function only by visual cues. A severe sensory neuropathy of this type may be caused by poisoning with enormous quantities of pyridoxine (Sacks, 1985).

The Romberg Test[11]

A Method

1. Have the patient stand with his feet together and with his arms at his sides.
2. Stand nearby, not touching the patient but ready to catch him if he falls.
3. Observe the patient for about 20 seconds.
4. Tell the patient to close his eyes. Continue the test for another 30 seconds.

Interpretation. As noted above, the ability to maintain posture only with the eyes open is evidence of impaired proprioception. But if the patient falls even with his eyes open, the test is positive for cerebellar and vestibular disease. (It is stated that if the patient falls repeatedly to one side, that is a sign of ipsilateral cerebellar disease; however, in the course of the required repeated testing, many patients learn that they will fall to that side and thus sometimes overcompensate by going to the other side in a lurch. Accordingly, it is suggested that if cerebellar disease is suspected on the basis of the Romberg test, the examiner should immediately proceed to some of the more specific and sensitive cerebellar signs described below.)

False Positives

Malingerers will also fall but they will first flex their knees and engage in other activities to lower the center of gravity and decrease the painful impact. When they learn that they will be caught and

[11] Romberg wrote the first neurology textbook. Prior to his work, there was only Morgagni's notebook.

protected, they may bend into the protecting examiner from the waist, keeping the legs straight. However, if they are not certain that they will be caught, they may simply collapse in a heap, straight down and not to one side or the other.

False Negative

The Romberg test was previously known to Morgagni. Romberg reinvented it because his practice as a neurologist was greatly concerned with tabes dorsalis. Nowadays we do not see much tabes, but we see a lot of diabetic neuropathy. This leads to an important false negative: The modern diabetic may have lost position sense in his toes but still have position sense at the ankles and thus be able to perform the Romberg test perfectly.

Cerebellar Signs or Predominantly Cerebellar Signs or Potentially Cerebellar Signs

Also see Chapter 10 and earlier in this chapter for nystagmus, the section on "Gait" in this chapter, Table 26.1 and accompanying discussion for the eye signs of acute cerebellar hemorrhage, and "Freebies" at the end of this section.

Dysdiadokokinesia

Dysdiadokokinesia is clumsiness in performing rapid alternating movements, such as alternating pronation and supination of the hands, or difficulty with the tongue movements required to pronounce "k," "t," or "cu."

A Method

1. Tell the patient to imitate you, and begin by rapidly (150 times per minute) patting your own chest with both hands, using, say, the radial side of each hand.
2. While watching the patient's imitation, increase the speed, looking for differences in rate, range, amplitude, and direction between the patient's two sides (a very sensitive test of unilateral cerebellar disease in itself).
3. Next, begin alternating the side of the hand striking the chest, going to the ulnar side for 10 beats and then back to the radial side for 5 beats and finally alternating sides on each blow. Watch the patient to see if one side "lags behind" or falls out of rhythm. Of course, any limb or set of alternating movements may be used (though forearm rotation is generally best) but the test should always be done bilaterally.

Another Method

Ask the patient to repeat "Topeka" or "Katy, K-Katy" as fast as possible.

Interpretation. Abnormalities of rate, range, amplitude, and direction occur only in cerebellar disease, so the predictive value of a properly performed positive test is quite high.

Sometimes it is easier to make a determination by listening, not just to "Topeka" but also to the hand-patting test (steps 1 and 2 above). Are rhythm and amplitude maintained, or do you hear a ritardo and diminuendo?

False Negatives

Wiener and Nathanson (1976–1977) note that many physicians miss unilateral cerebellar disease through not having the patient perform with both hands at the same time. Such a failure obviously deprives the physician of his control side.

False Positives

A false-positive test should be considered a possibility any time the test for dysdiadokokinesia is positive and other cerebellar tests are negative. This can occur in the nondominant hand, which is often less skilled at fine motor performance than the dominant side. It can also occur with frontal lesions that produce apraxia, owing to the fact that the cerebellum, whose posterior lobe receives its major projections from the frontal lobe, is thus rendered "deaf" (i.e., suffers sensory deprivation).

The Heel-to-Knee-to-Shin Test

A Method

1. Tell the supine patient to place one of his heels on the opposite knee and then to slide the heel all the way down the shin to the big toe.
2. Repeat with the other heel.

Interpretation. Patients with cerebellar disease cannot do the heel-to-knee-to-shin test. If the cerebellar disease is unilateral, the impairment will only be seen with the ipsilateral heel.

As with many of the vestibulocerebellar tests, the heel-to-knee-to-shin test may be converted to a proprioception-posterior columns test by having the patient also perform it with his eyes closed. Patients with cerebellar disease will have an abnormal test with the eyes closed or open; but patients with proprioception-posterior columns disease will be markedly improved with the eyes open.

Norman Bass Razor. Repeat the heel-to-knee-to-shin test on both sides, insisting that the heel be *exactly* 1 in. above the shin. The induced "locking" of the hip joint actually improves the performance of this step in patients with cerebellar ataxia but does not improve the performance of the test in persons with other causes of poor performance such as malingering.

The Finger-to-Nose Test

A Method

1. Hold your finger in front of the patient's face, far enough away that he must fully extend his arm to touch it, and instruct him, "Touch my finger with your right forefinger."
2. After he does this, say, "Now touch your nose."
3. Repeat several times with your finger in the same place.
4. While the patient is moving his finger back to his nose, relocate your own target finger in a different quadrant.
5. After he touches his nose, say, "Now touch my finger again."
6. Repeat steps 4 and 5, randomly varying the target, each time causing the patient to extend his arm fully. Increase the speed slightly and do about five trials.
7. Repeat the whole procedure with the patient's other hand.

Interpretation. Look for *cerebellar ataxia*, which will be manifested by a jerky, asynergic pointing at the target finger of the examiner. Also look for *dysmetria* (abnormal movement/judgment of distance), which will be manifested by missing the target through lateral displacement at termination of the movement.

Dysmetria is found not only in cerebellar disease but also in upper-extremity proprioceptive disease (which is somewhat rare, occurring in tabes dorsalis and untreated pernicious anemia) and in vestibular disease.

But if upper-extremity position sense is intact and there are no vestibular signs elicitable, then this is an excellent test for cerebellar disease. In that case, if the abnormalities noted above are found only on one side, then the patient has ipsilateral hemispheric cerebellar disease. (If the signs are abnormal *bilaterally*, the patient has either bilateral cerebellar hemispheric disease or disease of the midline cerebellar structures.)

In unilateral disease, if the test is done rapidly with alternate hands, the "bad" hand will drift or drop toward the floor during the "good" hand's turn (while the "bad" hand is waiting its turn).

A Different Response. Overpointing can be a sign of loss of depth perception (e.g., in parietal lobe dysfunction; see later in this chapter).

Because the subject must be allowed to look at the target finger, the test in its present form does not lend itself to repetition without visual cues; therefore, an alternate method is described to permit the differentiation of cerebellar and vestibular disease.

An Alternate Method

1. Have the patient extend his arm laterally. The movements here are initiated with the patient looking straight ahead and therefore lacking visual cues.
2. With flexion permitted only at the elbow joint, ask him to place his forefinger on the tip of his nose.

This test could be modified to permit visual cues by having the patient rotate his head toward the side of the tested forefinger.

Truncal Ataxia

Whereas limb ataxia is a sign of unilateral cerebellar hemisphere disease, truncal ataxia is a sign of disease of the posterior vermis and archeocerebellum. It is not necessary to get the patient to stand up for this, an important point because many of the patients so afflicted are unable to get out of bed. Even if the patient can simply be made to sit up in bed, patient observation will often be rewarded within 30 seconds by a torso that sways like a palm tree in a gusting wind.

Alternately, if the patient can sit on the side of the bed, sit down next to him. Healthy patients will adjust their position; patients with truncal ataxia will not. This may be the first or only sign to occur in children with posterior midline tumors (S. Horenstein, *personal communication*, 1988).

The Rebound Phenomenon of Stewart and Holmes

A Method

1. Have the patient flex his arm at the elbow, and attempt to extend it against the examiner's resistance, as during the testing of triceps strength.
2. Suddenly release the resistance.

Interpretation. If the arm goes into *complete* extension, with a force jerking the patient upward and forward, the test is positive. This is, of course, just one more test of the *impaired reciprocal inhibition* seen in cerebellar disease because normally one can stop the contraction of the extensors and initiate the contraction of the antagonistic flexors prior to reaching full extension.

Other authors' instructions to the contrary, this test should not be done in the opposite direction (as during testing of the biceps' motor strength) as the patient with cerebellar disease may strike himself in the face. Although this has been used as a test for malingering (because the malingerer has normal reciprocal inhibition and so will not strike his own face), it is a test for *cerebellar* malingering only. Contrary to some opinion, the absence of flexion rebound in a patient with simple asthenia of that upper extremity does not prove malingering. It simply proves that the patient has no detectable cerebellar disease.

Freebies

There are other cerebellar signs that may be casually noticed during the examination of the patient but that are not usually specifically sought.

1. *Cerebellar speech* is irregular, jerky, and briefly monotonous and slow, with sudden punctuations by explosive increases in rate and volume, as if a poorly tuned motorcycle were speaking.
2. After the patellar jerk has been elicited, the leg will continue to swing back and forth several times before it comes to rest. (*Before* eliciting the knee jerk, make sure there is room for the leg to swing back, should this sign be present.) This is called the *pendular knee jerk*. (The test also works with the triceps jerk.) It is the best test for hypotonia, which may also be noted by the experienced examiner during muscle testing.
3. Eye signs of cerebellar hemorrhage have been listed in Table 26.1.

The cerebellar signs are intellectually appealing for two reasons. First, they are easy to learn to perform; and second, when one of them is present, others usually will be found. (This is true for clinicians beginning with the patient and progressing toward the diagnosis. If one begins with the CT scan and then tries to reason back to the physical, one can occasionally find a cerebellar metastasis that has not yet caused any physical findings.)

Vestibular Signs and Predominantly Vestibular Signs

The Oculocephalic ("Doll's Eye") Reflex

Also called *Bielschowsky doll's-head eye reflex* (the original term), the doll's head, the doll's eye sign or maneuver, or the oculocephalic reflex, sign, or maneuver, this is a useful test in comatose patients in whom there is a question as to whether the midbrain and its reflexes have been preserved.

In a comatose patient over 6 months of age with an intact midbrain and vestibular reflexes, the eyes will maintain the original direction of gaze as the patient's head is passively turned. This means that the eyes are fixed on the same point in the room as the patient's head is moved, which means that the globes move within the head proportionally to the movement of the head but in a direction opposite to the passive motion of the head. If you stare at the right edge of this page as you rotate your head to the left, you are maintaining the "original direction of gaze."

Interpretation. The slow rotation activates the vestibular apparatus, which provides input to the midbrain, which instructs the extraocular muscles to move the globes so as to maintain the original direction of gaze, as if the patient were still focusing on an object. But if there is midbrain damage or dysfunction (specifically in the

area of the MLF from the horizontal gaze center in the abducens nucleus in the lower pons to the oculomotor nucleus in the midbrain), these reflexes are lost, and passive rotation of the head will not result in movement of the globes to maintain the original direction of gaze. Rather, the eyes will continue to stare in whatever direction the head is pointed. It is as if they were painted onto the skull.

Caveats. Because the doll's eye test involves the passive rotation of the skull in a patient who is unconscious, we must remember to assure ourselves of the integrity of the cervical vertebrae before performing this test. Thus, this maneuver is generally neither useful nor advisable in the evaluation of patients in the emergency room (M. Faria, *personal communication*, 1998). If one cannot be assured of the absence of a neck injury, one should proceed directly to caloric testing.

This test is generally done only in comatose patients. Patients who are awake will have doll's eyes, or not, depending on whether they have fixed their gaze or are looking wherever you turn their head.

For the Advanced Student. The oculocephalic reflex also operates in the vertical direction ("setting sun sign") and it has been so used in the study of paralysis of upward voluntary gaze (see earlier in this chapter).

A Note on Recording the Findings. A report that the "doll's eye test" is "positive" or "negative" may be misunderstood. The term refers to dolls with mobile, weighted eyes that maintained their position as the doll's head was turned. This author remembers thinking, during a demonstration of an intact oculocephalic reflex: "But dolls' eyes don't do that." All her dolls had had fixed or painted-on eyes.

The shorthand terminology that is least confusing to this author is to say "oculocephalic reflexes intact (or preserved) bilaterally" (meaning that the eyes maintained their original direction of gaze on lateral rotation of the head to one side and the other) or to describe what happens if this is not the case (such as "the position of the eyes remained fixed with respect to the head on rotation to either side").

Perhaps it is easiest to remember that dead people's eyes do not move. The usual reason for testing the oculocephalic reflex is not to test for an obscure lesion in the medulla or midbrain or connections in between but to check for brain death (*vide infra*).

The Vestibular Finger-to-Nose Test

This is a test of upper-extremity proprioception of the cerebellum and of the vestibular apparatus. Because upper-extremity proprioception is usually normal (as can be independently determined; see position sense testing presented later in this chapter), this is used as a test for the cerebellum and the vestibular apparatus. For the latter, the test is used in conjunction with one of the vestibular stimulation tests (calorics and the various motion tests, *vide infra*).

A Method

1. Have the patient touch your fingertip, as in the original version of this test presented earlier in this chapter.
2. After he touches your finger, have the patient touch his nose as before.
3. Tell him to close his eyes.
4. Immediately ask him to touch your (unmoved) finger again.

Interpretation:

1. With cerebellar disease, the patient will probably show a cerebellar intention tremor.
2. If the lesion is not unilateral, the patient will "past point" on either side (randomly) an equal number of times. With unilateral vestibular or cerebellar disease, the patient will past point only to one side.

Spontaneous Nystagmus

Before performing the tests for induced nystagmus (*vide infra*), check for baseline nystagmus (review Chapter 10 for a description of various types of nystagmus and for additional comments).[12] In the awake patient, nystagmus has two components: the fast (jerk) cerebral component and the slow vestibular component.


If there is a destructive lesion of one vestibular nerve or labyrinthine apparatus, there may be a spontaneous nystagmus in which the slow component looks at the diseased side. This simple observation has saved a lot of people a lot of time.


If the fast component is always in the same direction (regardless of whether the eyes are looking to the left or the right), the lesion is labyrinthine. If the fast component changes direction with changing direction of gaze, always showing the fast component in the direction of gaze, then the problem is not labyrinthine but in the brainstem (including drug intoxications).


Vertical nystagmus always signifies a lesion in the brain. It is usually secondary to a midline lesion or to Wernicke encephalopathy. It can be a sign of acute beriberi, usually precipitated by administering glucose to a thiamine-deficient alcoholic. This requires urgent administration of intravenous thiamine. It is better to avoid this emergency by generous use of thiamine in all high-risk patients.

Upbeat vertical nystagmus in the primary position, accentuated by upward gaze, points to a lesion in the anterior cerebellar vermis or the medulla, as in the medial medullary syndrome. This syndrome results from vascular occlusion; two cases secondary to fibrocartilaginous embolism to the anterior spinal artery and its branches, presumably from the nucleus pulposus of an intervertebral disc, have been described (Kase et al., 1983).

If nystagmus is present only and always together with vertigo, then the patient has vestibular disease, and thus may have Ménière disease. However, if the nystagmus and vertigo are dissociated, Ménière disease is excluded because the lesion must be in the brainstem (S. Horenstein, *personal communication*, 1988). With Ménière syndrome, there is no nystagmus between spells with eyes open but it may be present with eyes closed (Alford, 1972).

Nystagmus on rapid rising results from disease in the posterior semicircular canals, not the brainstem. Thus, it is important to make the physical examination appropriate to the history. If the patient complains of vertigo (not light-headedness) on rapid rising, have him stand up quickly and check whether nystagmus occurs at the same time (S. Horenstein, *personal communication*, 1988).

Rotatory nystagmus is a classic sign of BPPV (see discussion of Dix–Hallpike maneuver below).

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[12] Also see Chapter 4 for instructions about the *naming* of nystagmus. In this chapter, the convention of neurology tests will be followed when possible: The nystagmus is named for the fast component.

Downbeat nystagmus is presumptive evidence of dysfunction at the lower end of the brainstem or cerebellum. Structural causes include platybasia and the Arnold–Chiari malformation. Metabolic causes include magnesium depletion, lithium intoxication, alcoholic cerebellar degeneration, Wernicke encephalopathy, anoxia, and anticonvulsant toxicity (phenytoin or carbamazepine). Inflammatory causes include encephalitis and syphilis. Other causes include hereditary and paraneoplastic cerebellar degeneration, encephalitis, brainstem infarction or vertebrobasilar insufficiency, brainstem compression from cervical involvement in rheumatoid arthritis (Menezes et al., 1985), MS, and bilateral internuclear ophthalmoplegia (Alpert, 1978; Chrousos et al., 1987; Cogan, 1968).

Monocular nystagmus almost always signifies pontomedullary disease.

In *seesaw nystagmus*, one eye goes up and intorts as the other eye goes down and extorts, most prominently when the patient is fixating. It is usually associated with severe visual field defects. The lesion usually is in the rostral part of the mesencephalon, probably involving the interstitial nucleus of Cajal (L. Huntoon, *personal communication*, 2004).

Caloric Testing (Oculovestibular Reflexes)

To test the integrity of the labyrinthine apparatus, the midbrain, and the oculomotor efferents, heat and cold are used to induce thermal currents in the vestibular endolymph. These in turn produce nystagmus, provided that the neural tissue specified above is intact.

A Method

1. If the patient is supine, tip his head forward about 30 degrees. This ensures that you get maximal stimulation of the horizontal canals and so produce horizontal nystagmus.

   For the Conscious Patient. If the patient is sitting, tip his head back about 60 degrees. If you leave his head upright, you will get maximal stimulation of the vertical canals, which produces rotatory nystagmus. Having the conscious patient wear a pair of strong convex lenses prevents him from accommodating to the stimulus by visual fixation, yet still permits the examiner to see the nystagmus. Alternately, Dr Broadwater of Missouri simply has the patient close his eyes; nystagmus can be detected from the corneal bulges in the lightly closed lids.

   This test may cause the conscious patient to vomit. Be sure to have a bucket close by.

   Cerebellar nystagmus, unlike caloric nystagmus, is not position dependent.
2. Pour 100 mL of very cold water in one of the external canals after you have assured yourself that there is no perforation of the tympanic membrane and that the canal is free of cerumen. If cerumen keeps the cold water from reaching the tympanic membrane and setting up a current in the endolymph, the result will be a false-negative test.

   Comment. The purpose of the ice water is not to give the patient frostbite of the tragus but to set up a thermal current in the endolymph. If you use cool water (20°C), you can eventually get the same effect but it will require about a quart of water. With ice water, you can get a nice effect with only 5 to 30 mL, starting in about 20 to 30 seconds and lasting about 1 minute.

   The same thing goes for warm water, should you care to use it. You can use a quart of 50°C water or smaller volumes of hotter water. The fact that this test is usually performed on unconscious patients should not encourage you to scald the tympanic membrane; test the temperature against the back of your hand.
3. *Look for nystagmus in the conscious patient or deviation of the eyes in the unconscious patient.* In the conscious patient, the cold-induced slow component comes toward the stimulated ear and the fast jerky compensatory cerebral component, for which the nystagmus is named, moves opposite. With warm water, the directions are reversed. (Thus, the mnemonic COWS = cold opposite, warm same.)

Interpretation:

1. In unconscious patients, there can be no nystagmus because there is no fast cerebral component. *The unopposed slow cerebellar/vestibular component appears as tonic deviation toward the cold side.* If both sides are stimulated with cold, the eyes look downward (Plum and Posner, 1972).
2. In conscious patients, the absence of thermally induced nystagmus on just one side means that a destructive lesion (labyrinth or vestibular nerve) is ipsilateral. The absence of an inducible response bilaterally means that there is bilateral disease, which by Occam's razor is probably in the brainstem.
3. In conscious patients, a trick for amplifying the nystagmus so that it is more easily seen is to have the patient look to the side away from the cold-stimulated ear rather than straight ahead.

   Lateral past pointing and falling, which can be checked for at the same time, are, however, still *toward* the side of the cold irrigation.
4. Some lesions such as expanding tumors, abscesses, vascular aneurysms, and so forth, initially go through an irritative phase before they become destructive. At this point, there is simply an exaggeration of all normal responses. For instance, cold stimulation of the diseased side will produce nystagmus (fast component to the healthy side), which can be accentuated on lateral gaze to the healthy side. There is also falling and past pointing to the afflicted side. This phase of the disease does not usually last long but can occasionally be confusing if one does not know about it.

Fortunately, electronystagmograms have now generally replaced caloric testing, which is miserably uncomfortable.

The Bárány Chair Test

If you can find an old-fashioned barbershop chair with a headrest and a spinning base, you can try this one. (A Veterans Administration hospital may have a barbershop with such a chair.) A desk swivel chair with arms could also be used.

A Method

1. Instruct the patient to close his eyes and place his head against the headrest, which is tilted 60 degrees backward (from vertical) or 30 degrees forward (from horizontal).
2. Spin the chair to the patient's right 10 times, as fast as you can, and abruptly stop it with the patient facing you.
3. Tell the patient to open his eyes.

4. Normally, the patient will have nystagmus with the slow component going to the right (the direction in which the patient was spun).
5. If the patient attempts to stand, he will fall, or more commonly will lean to his right while seated.
6. Hold a finger up directly in front of the patient and ask him to touch it with his forefinger. Normally, he will past point to his right.
7. Repeat, spinning the chair to the patient's left.

Interpretation:

1. Destructive lesions cause the absence of the normally elicited responses (steps 4 to 6 above).
2. If only some of the responses are lost, the patient may have an intramedullary lesion.
3. Various other interpretations of these tests have been offered without any good data from large series. The availability of computerized tomography and nuclear MRI increases the opportunity for definitive analysis of all patterns of response, but at the same time has decreased performance of these clinical tests for cerebellar and midbrain lesions, which can now be imaged *in vivo*.

The Dix–Hallpike Maneuver

This test, which has also been called the *Bárány or Nylen–Bárány maneuver*, is a unilateral vestibular stimulation test for BPPV. It should be done routinely in all patients complaining of vertigo. BPPV is the most common and the most easily treatable cause of dizzy spells.

A Method

1. Have someone stand on each side of the examining table when you perform the maneuver. Some patients develop severe vertigo, lose their bearings, and can fall off the table. It is prudent to have a bucket nearby as patients occasionally vomit.
2. Have the patient seated so that when he lies down, his head will extend over the end of the examining table.
3. Explain to the patient what is going to be done and that any dizziness will only last a few seconds.
4. Maintain control of the patient's head to be sure the maneuver is performed optimally so as to provide maximal stimulation. Place one hand on top of the head and one hand under the chin.
5. On the count of "one, two, three," have the patient go quickly from the sitting to the supine position, as you quickly turn his head toward you, stimulating the posterior semicircular canal. Tell the patient, "Keep your eyes open and look directly at me." The natural tendency to close one's eyes at the sudden onset of vertigo could cause you to miss the rotatory nystagmus that may only last a few seconds.
6. Observe the patient's eyes for at least 15 seconds to see whether nystagmus is induced. Be attentive because the nystagmus may last only a few (from 2 to 30) seconds.
7. Slowly bring the patient to a sitting position, with the head still rotated, and check for nystagmus again.
8. Repeat, with the head rotated to the opposite direction. (The "down" ear is the one being tested.) If the patient can tell you which side down causes the vertigo, check the opposite side first to minimize nausea.

Interpretation. Whichever down side produces nystagmus is the "affected" side causing the positional vertigo.

With labyrinthine disease, there is a 4- to 5-second latent period between the movement and the onset of nystagmus. The nystagmus is primarily torsional but may have some vertical components. The results are prominent and dramatic. In some cases, the test is strongly positive only the first time that it is performed, with the response fatiguing rapidly.

The test does not always induce nystagmus in BPPV; the sensitivity is reportedly from 50% to 78%. A positive test combined with a history of either vertigo or vomiting gives an LR of 7.6 for a peripheral, nonemergent form of vertigo. If one or more of these features are lacking in a "dizzy" patient, the LR for peripheral, nonemergent vertigo as opposed to a potentially emergent and/or central cause is 0.6 (Froehling et al., 1994).

In brainstem disease, there is no latent period, and the nystagmus may be purely vertical.

Cerebellar infarction could masquerade as a peripheral vestibular lesion because vertigo and severe imbalance may be the only presenting features. If this lesion is suspected, neuroimaging is required on an urgent basis. Because the distinction is so critical, patients should try to stand and walk even if it is very uncomfortable. Patients with a peripheral lesion should be able to walk during the acute phase, whereas those with a central lesion often cannot stand to take a single step without falling (Baloh, 1998a,b).

Semeiophysiology. The stereotypical quality of the response gives a clue to the origin of the disorder. BPPV results from the free movement of particulate debris (calcium carbonate crystals called *otoconia*) in the semicircular canals, almost always in the posterior portion. The particles may dislodge, either spontaneously or as a result of head trauma, from a membrane in the utricle. Their weight helps the body sense gravity. The latency before the onset of nystagmus results from the time required for the particles to start moving under the influence of gravity. Nystagmus ceases when the particles come to rest. This mechanism suggests the treatment, the Epley maneuver.

The Epley (Canalith Repositioning) Maneuver. If the Dix–Hallpike maneuver produces a positive result, some physicians immediately undertake the Epley maneuver, which rotates the patient's head so that the loose particles slide out of the posterior semicircular canal into the utricle. If the vertigo occurs with the right ear facing downward, the physician waits for the vertigo to stop, then rotates the patient's head to the left, with the right ear facing upward. After holding the head in that position for 30 seconds, the physician asks the patient to roll over onto his left side.

Meanwhile, the physician rotates the patient's head to the left until the nose points toward the floor and holds that position for 30 seconds. Finally, the physician lifts the patient into a sitting position with the head still facing left. For vertigo occurring with the left ear down, carry out the same maneuver with appropriate modifications. If necessary, the maneuver can be repeated several times until the nystagmus is gone.

Usually, if the diagnosis is correct the maneuver produces a dramatic cure. To prevent recurrence, the patient is cautioned not to lie flat for a few days, or not to bend down, tilt the head backward,

make quick head-turning movements, or sleep on the affected side.

Dr Lawrence Huntoon teaches patients and family members how to do this maneuver themselves. It is important to take precautions against patients falling off the table.

A number of variations on the method have been described.

Head Thrust Test

This test assesses the vestibulo-ocular reflex (VOR), which is checked in comatose patients with the doll's eye or caloric test.

A Method

1. Stand in front of the patient and grasp his head in both hands.
2. Instruct the patient to look at your nose.
3. Quickly move the patient's head 5 to 10 degrees to one side and observe the eye movements.
4. Repeat for the other side.

Interpretation. A normal response is for the patient's eyes to stay on target (the examiner's nose). When there is a lesion of the VOR on one side, a corrective eye movement (i.e., a corrective "saccade") back to the examiner's nose is seen after the head is moved *toward* the affected side. These features can be appreciated even when spontaneous nystagmus is present. A patient with unidirectional nystagmus, a positive head thrust in the direction opposite the fast phase of nystagmus, and no other neurologic features can be diagnosed with vestibular neuritis, a peripheral lesion, with a high level of certainty (Kerber 2009).

A Note on Vertigo

Dizziness is the third most frequent complaint in a study of patients presenting at a general internal medicine outpatient clinic. Vertigo was the most frequent category of dizziness (Froehling et al., 1994).

It is important to distinguish peripheral causes of vertigo (the labyrinth or eighth cranial nerve) from central nervous system causes.

The differential diagnosis of central causes includes vertebrobasilar ischemia, cerebellar infarction or hemorrhage, brain tumors, MS, or migraine. A careful neurologic examination is indicated.

Nonbenign peripheral causes include cholesteatoma and acoustic neuroma. Do not neglect to examine the ear and check the hearing (also see Chapter 11).

Motor Examination

Testing of muscle strength has been described in detail in Chapter 25.

Intercostal Muscle Innervation Test

A quick way to determine motor function in the thoracic spinal levels is to place your hands laterally on the patient's rib cage, with your fingers between the ribs, and ask the patient to breathe deeply. You may need to move your hands cranially or caudally a few times to find the lowest working level. If a pair of ribs separates, there is some motor function; otherwise, there is not. False positives are possible from sternocleidomastoid or other muscles. In a complete T-6 paraplegic, Dr Edward J. Harshman, who developed this concept, reports being able to find a sharp line between working and nonworking intercostals. This method is more objective than the well-known sensory level for spinal cord injury. It also predicts sitting balance in stroke patients (E.J. Harshman, *personal communication*, 2004).

Fibrillation and Fasciculation

Though involving the muscles, these phenomena are signs of neurologic disease. In *fibrillation*, individual muscle fibers are firing spontaneously because they have lost their innervation. A *fasciculation* involves the entire motor unit: the nerve cell, the axon, and all the motor fibers that the unit innervates. Fibrillations are regular. On electromyogram (EMG) needle recordings, fibrillations sound like a ticking clock. Fasciculations, on the other hand, are very irregular and sound like a thump in the night (L. Huntoon, *personal communication*, 2004).

Fibrillations cannot be seen. Fasciculations in the tongue may cause it to resemble a bag of worms. Do not have the patient protrude the tongue; just have him open his mouth.

Fasciculations may be seen from time to time as muscle twitches in healthy persons. They can be caused by compression on a nerve root or irritation in an anterior horn cell.

Pernicious fasciculations have the following four characteristics:

1. They are stereotyped, that is, they always affect the same muscle fibers.
2. They are present at rest.
3. They may even be present at sleep.
4. They are perfectly rhythmic or periodic.

The patient may also have atrophy and/or weakness in the same muscles. Pernicious fasciculations may be seen in amyotrophic lateral sclerosis (ALS), syringomyelia, bulbar palsy, and progressive spinal muscle atrophy of whatever eponym.

A Note on Muscle Tone and Upper and Lower Motor Neuron Disease

A spastic paralysis, associated with muscle hypertonicity and exaggerated deep tendon reflexes, results from lesions of the brain or from interruption of the descending tracts. In contrast, lower motor neuron or peripheral nerve lesions produce a flaccid paralysis.

A spinal cord compression may give lower motor neuron signs in the muscles innervated by the compressed segment plus upper motor neuron signs on one or both sides below the level of the compression (Brain and Walton, 1969).

A combination of upper and lower motor neuron signs also occurs in ALS (or Lou Gehrig disease). The classic presentation involves fasciculations (as in the tongue) and muscle wasting (as in the intrinsic muscles of the hand), with increased reflexes and weakness in a pyramidal distribution (*vide infra*) and extensor plantar responses (*vide infra*).

Assessing Muscle Tone

As muscle tone is of practical importance in neurologic diagnosis, pendulousness of the legs was described as a simple and reliable office test (Wartenberg, 1951).