Section Ⅰ

Initial Clinical Evaluation

Chapter 1

Clinical Neurologic Evaluation

one

Overview and Basic Tenets

The neurologic examination begins the moment the patients get out of their seat to be greeted, the character of their smile or lack thereof, and subsequently as they walk to enter the neurologist's office. An excellent opportunity to judge the patient's language function and cognitive abilities occurs during the acquisition of the patient's history. Concurrently, the neurologist is always attuned to carefully making observations in order to identify various clinical signs. Some are overt movements (tremors, restlessness, dystonia or dyskinesia); others are subtler, e.g., vitiligo, implying a potential for a neurologic autoimmune disorder. Equally important may be the lack of normal movements, as seen in patients with Parkinson disease. By the time the neurologist completes the examination, she or he must be able to categorize and organize these historical and examination findings into a carefully structured diagnostic formulation.

The subsequent definition of the formal examination may be subdivided into a few major sections. Speech and language are assessed during the history taking. The cognitive part of the examination is often clearly defined with the initial history and often does not require formal mental status testing. However there are a number of clinical neurologic settings where this evaluation is very time consuming and complicated; Chapter 2 is dedicated to this aspect of the patient evaluation. However, when there is no clinical suspicion of either a cognitive or language dysfunction, these more formal testing modalities are not specifically required.

Here the multisystem neurologic examination provides a careful basis for most essential clinical evaluations. Neurologists in training and their colleagues in practice cannot expect to test all possible cognitive elements in each patient that they evaluate. Certain basic elements are required; most of these are readily observable or elicited during initial clinical evaluation. These include documentation of language function, affect, concentration, orientation, and memory. When concerned about the patient's cognitive abilities, the neurologist must elicit evidence of an apraxia or agnosia and test organizational skills. Once language and cognitive functions are assessed, the neurologist dedicates the remaining portion of the exam to the examination of many functions. These include visual fields, cranial nerves (CNs) (Fig. 1-1), muscle strength, muscle stretch reflexes (MSRs), plantar stimulation, coordination, gait and equilibrium, as well as sensory modalities. These should routinely be examined in an organized rote fashion in order not to overlook an important part of the examination. The patient's general health, nutritional status, and cardiac function, including the presence or absence of significant arrhythmia, heart murmur, hypertension, or signs of congestive failure, should be noted. If the patient is encephalopathic, it is important to search for subtle signs of infectious, hepatic, renal, or pulmonary disease.

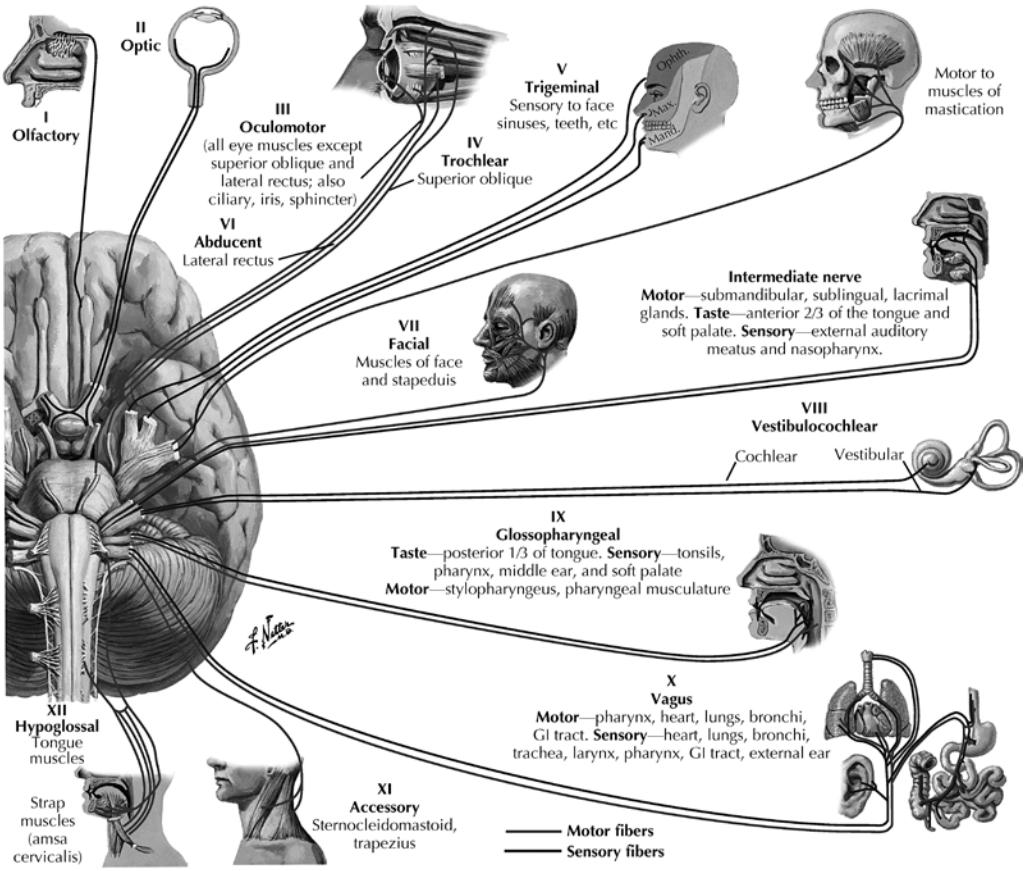


Figure 1-1 Cranial Nerves: Distribution of Motor and Sensory Fibers.

CRANIAL NERVES: AN INTRODUCTION

The 12 CNs subserve multiple types of neurologic function (Fig. 1-1). The cranial nerves are formed by afferent sensory fibers, motor efferent fibers, or mixed fibers traveling to and from brainstem nuclei (Fig. 1-2A and B).

The special senses are represented by all or part of the function of five different CNs, namely, olfaction, the olfactory (Ⅰ); vision, the optic (Ⅱ); taste, the facial (Ⅶ) as well as the glossopharyngeal (Ⅸ); hearing as well as vestibular function, the cochlear and vestibular (Ⅷ) nerves. Another three CNs are directly responsible for the coordinated, synchronous, and complex movements of both eyes; these include CNs Ⅲ (oculomotor), Ⅳ (trochlear), and Ⅵ (abducens). Cranial nerve Ⅶ is the primary CN responsible for facial expression, which is important for setting the outward signs of the patient's psyche's representation to his family and close associates, or signs of paralysis from a brain or cranial nerve lesion. Facial sensation is subserved primarily by the trigeminal nerve (Ⅴ); however, it is a mixed nerve also providing primary motor contributions to mastication. The ability to eat and drink depends on CNs Ⅸ (glossopharyngeal), Ⅹ (vagus), and Ⅻ (hypoglossal). The hypoglossal and recurrent laryngeal nerves are also important to the mechanical function of speech. Last, CN-Ⅺ, the accessory, contains both cranial and spinal nerve roots that provide motor innervation to the large muscles of the neck and shoulder.

Disorders of the CNs can be confined to a single nerve such as the olfactory (from a closed-head injury, early Parkinson disease, or meningioma), trigeminal (tic douloureux), facial (Bell palsy), acoustic (schwannoma), and hypoglossal (carotid dissection). There is a subset of systemic disorders with the potential to infiltrate or seed the base of the brain and the brainstem at the points of exit of the various CNs from their intraaxial origins. These processes include leptomeningeal seeding of metastatic malignancies originating in the lung, breast, and stomach, as well as various lymphomas, or granulomatous processes such as sarcoidosis or tuberculosis, each leading to a clinical picture of multiple, sometimes disparate

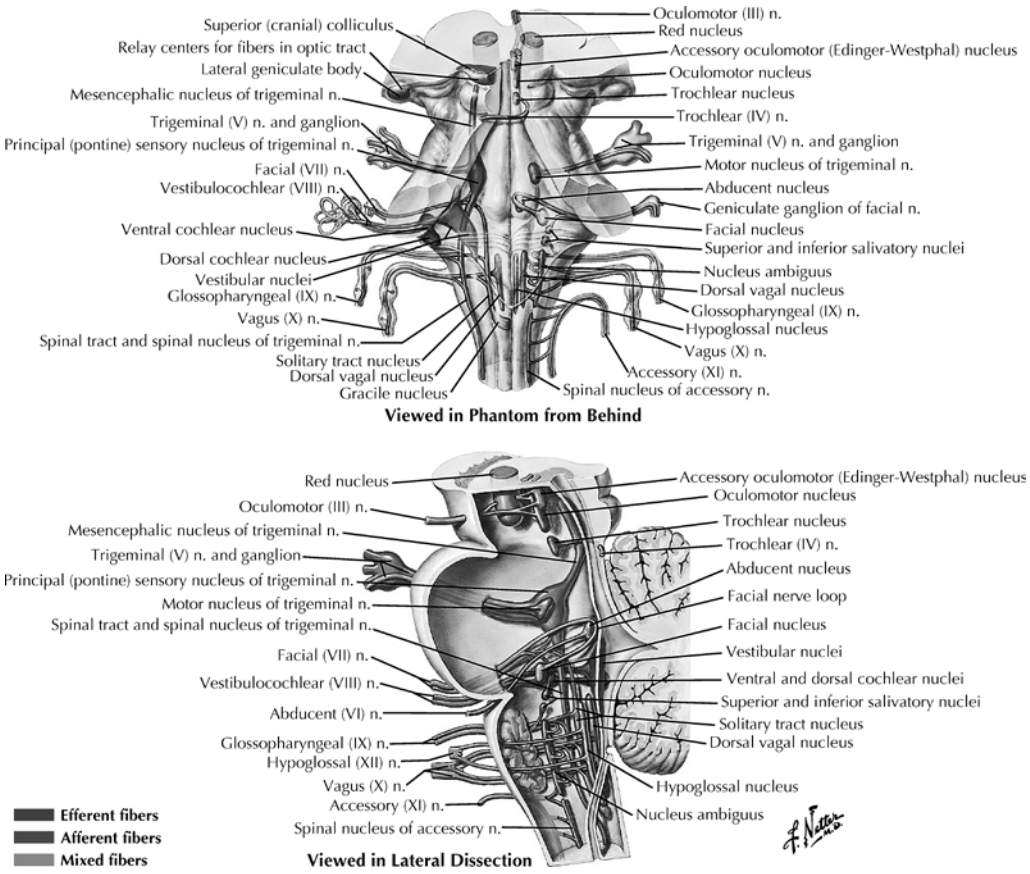


Figure 1-2 Cranial Nerves: Nerves and Nuclei.

cranial neuropathies. Many times, a stuttering onset occurs. The various symptoms are related to individual CNs. These typically develop within just weeks or no more than a few months.

Cranial nerve dysfunctions will commonly bring patients to medical attention for a number of clinical limitations. These include ophthalmic difficulties, such as diminished visual acuity or visual field deficits (optic nerve and peri-cavernous chiasm) and double vision, either horizontal, vertical, or skewed (oculomotor, trochlear, and abducens nerves). Other cranial nerve presentations include facial pain (trigeminal nerve), evolving facial weakness (facial nerve), difficulty swallowing (glossopharyngeal and vagus nerves), and slurred speech (hypoglossal nerves).

CRANIAL NERVE TESTING

I: Olfactory Nerve

The sense of smell is a very important primordial function that is much more finely tuned in other animal species. Here other mammals are able to seek out food, find their mates, and identify friend and foe alike because of their finely tuned olfactory brain. In the human, the loss of this function can still occasionally have very significant consequences primarily bearing on personal safety. If the human being cannot smell fires or burning food, their survival can be put at serious risk. The loss of smell also affects the pleasure of being able to taste, even though, as later noted, taste per se is primarily a function of cranial nerves Ⅶ and Ⅸ.

Olfactory nerve function testing is relevant despite its only occasional clinical involvement. This may be impaired after relatively uncomplicated head trauma and in individuals with various causes of frontal lobe dysfunction, especially an olfactory groove meningioma. Loss of olfaction is sometimes an early sign of Parkinson disease. Clinical evaluation of olfactory functions is straightforward. The examiner has the patient sniff and attempt to identify

familiar substances having specific odors (coffee beans, leaves of peppermint, lemon). Inability or reduced capacity to detect an odor is known as anosmia or hyposmia, respectively; inability to identify an odor correctly or smell distortion is described as parosmia or dysosmia. Bilateral olfactory nerve disturbance with total loss of smell, typically from head trauma, chronic upper airway infections, or medication, is usually a less ominous sign than unilateral loss, which raises the concern for a focal infiltrative or compressive lesion such as a frontal grove meningioma.

II: Optic Nerve

Of all the human sensations, the ability to see one's family and friends, to read, and appreciate the beauties of nature, it is difficult to imagine life without vision, something that is totally dependent on the second cranial nerve. Obviously many individuals, such as Helen Keller, have vigorously and successfully conquered the challenge of being blind; however, given the choice, vision is one of the most precious of all animal sensations. "Blurred" vision is a common but relatively nonspecific symptom that may relate to dysfunction anywhere along the visual pathway (Fig. 1-3). When examining optic nerve function, it is important to identify any concomitant ocular abnormalities such as proptosis, ptosis, scleral injection (congestion), tenderness, bruits, and pupillary changes.

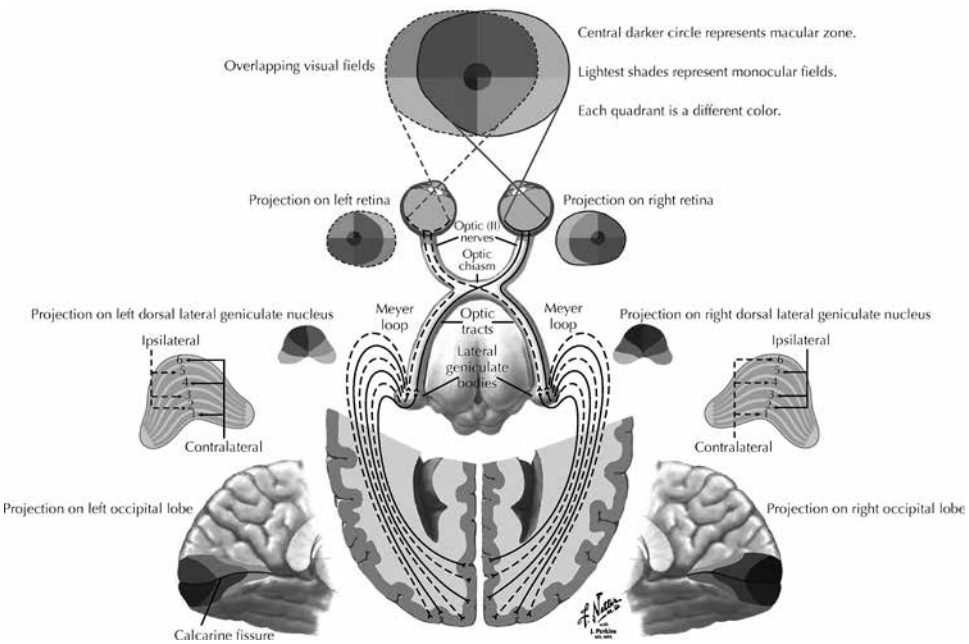


Figure 1-3 Visual Pathways: Retina to Occipital Cortex.

Visual acuity is screened using a standard Snellen vision chart that is held 14 inches from the eye. Screening must be performed in proper light as well as to the patient's refractive advantage using corrective lenses or a pinhole when indicated.

A careful visual field evaluation is the other important means to assess visual function. These tests are complementary, one testing central resolution at the retinal level and the other to evaluate peripheral visual field defects secondary to lesions at the levels of the optic chiasm, optic tracts, and occipital cortex. Visual fields are evaluated by having the patient sit comfortably facing the examiner at a similar eye level. First, each eye is tested independently. The patient is asked to look straight at the examiner's nose. The examiner extends an arm laterally, equidistant from himself and the patient, and asks the patient to differentiate between one and two fingers. The patient's attention must always be directed back to the examiner as most patients will reflexively look laterally at the fingers. This will require repeated testing. Each quadrant of vision is evaluated separately. After individual testing, both eyes are tested simultaneously for visual neglect, as may occur with right hemispheric lesions. Progressively complex perimetric devices have the advantage of providing valuable data on the health of the visual

system.

When one examines the pupils, their shape and size need to be recorded. A side-to-side difference of no more than 1 mm in otherwise round pupils is acceptable as a normal variant. Pupillary responses are tested with a bright flashlight and are primarily mediated by the autonomic innervation of the eye (Fig. 1-4). A normal pupil reacts to light stimulus by constricting with the contralateral constriction of the unstimulated pupil as well. These responses are called the *direct and consensual reactions*, respectively, and are mediated through parasympathetic innervation to the pupillary sphincter from the Edinger-Westphal nucleus along the oculomotor nerve. The pupils also constrict when shifting focus from a far to a near object (*accommodation*) and during convergence of the eyes, as when patients are asked to look at their nose.

The sympathetic innervation of the pupillary

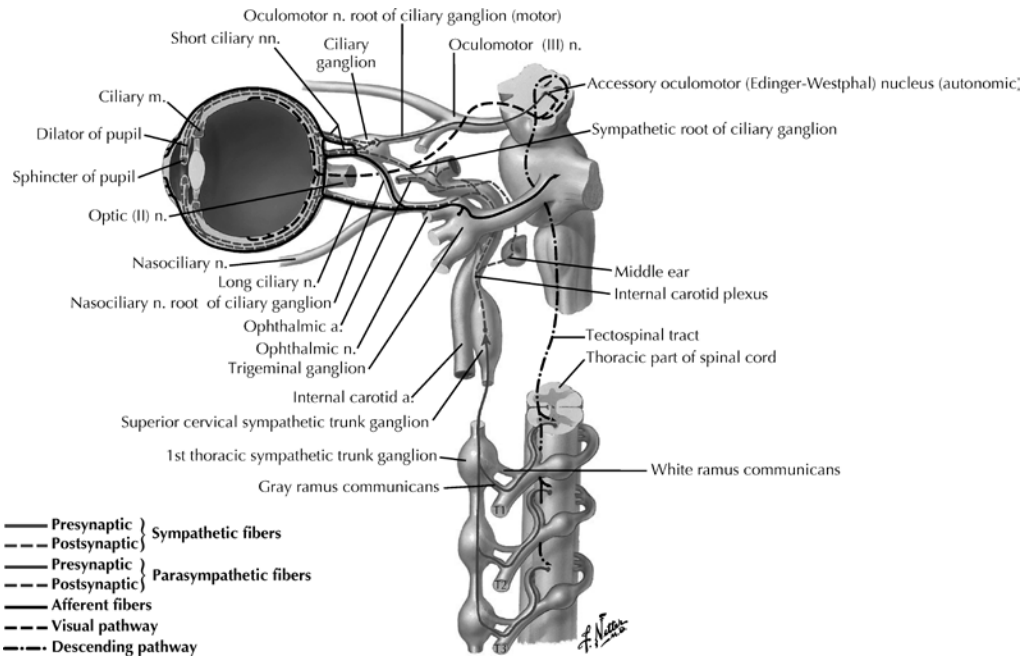


Figure 1-4 Autonomic Innervation of Eye.

dilator muscle involves a multisynaptic pathway with fibers ultimately reaching intracranially along the course of the internal carotid artery. Branches innervate the eye after traveling through the long and short ciliary nerves. The *ciliospinal reflex* is potentially useful when evaluating comatose patients. In this setting, if the examiner pinches the patient's neck, the ipsilateral pupil should transiently dilate. This provides a means to test the integrity of ipsilateral neuropathways to midbrain structures.

The short ciliary nerve, supplying parasympathetic inputs to the pupil, may be damaged by various forms of trauma. This results in a unilateral dilated pupil with preservation of other third nerve function. Significant unilateral pupillary abnormalities are usually related to innervation changes in pupillary muscles.

A number of pathophysiologic mechanisms lead to mydriasis (pupillary dilatation) (Table 1-1). Atropine-like eye drops, often used for their ability to produce pupillary dilation, inadvertent ocular application of certain nebulized bronchodilators, and placement of a scopolamine anti-motion patch with inadvertent leak into the conjunctiva are occasionally overlooked as potential causes for an otherwise asymptomatic, dilated, poorly reactive pupil. Other medications may also lead to certain atypical light reactions. The presence of bilateral dilated pupils, in an otherwise neurologically intact patient, is unlikely to reflect significant neuropathology. In contrast, the presence of prominent pupillary constriction most likely reflects the use of narcotic analogs or parasympathomimetic drugs, such as those typically used to treat glaucoma.

Table 1-1 Pupillary Abnormalities

	Argyll Robertson	Horner	Holmes Adie
Response to light	None	Yes	None
Other responses	Brisk reaction to near stimulus Converge	Normal	Tonic reaction to near stimulus Accommodation
Margins	Irregular	Regular	Regular
Associated changes	Iris depigmentation	Ptosis	Loss of MSR
Causes	Tabes dorsalis	Carotid dissection Carotid aneurysm Pancoast tumor Syringomyelia	Ciliary ganglion
Anatomy	Unknown (tectum of midbrain likely)	Loss of sympathetic	Loss of parasympathetic

MSR, Muscle stretch reflex.

HORNER SYNDROME

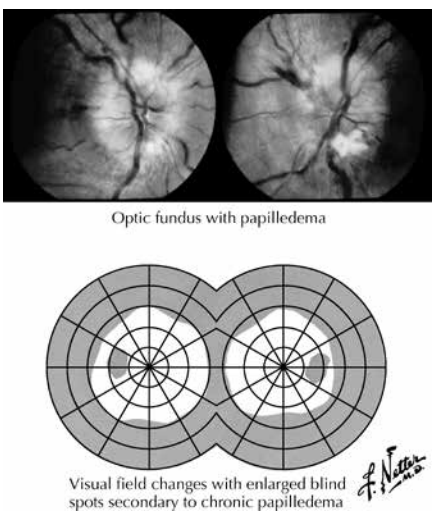
The classic findings include miosis (pupillary constriction), subtle ptosis, and an ipsilateral loss of facial sweating. Here the constricted pupil develops secondary to interference with the sympathetic nerves at one of many different levels along its long intramedullary (brain and spinal cord) and complicated extracranial course.

Sympathetic efferent fibers originate within the hypothalamus and traverse the brainstem and cervical spinal cord, then exit the upper thoracic levels and course rostrally to reach the superior cervical ganglia (Fig. 1-4). Subsequently, these sympathetic fibers track with the carotid artery within the neck to reenter the cranium and subsequently reach their destination innervating the eye's pupillodilator musculature. Typically, patients with Horner syndrome have an ipsilateral loss of sweating in the face (anhidrosis), a constricted pupil (miosis), and an upper lid droop from loss of innervation to Muller's muscle, a small smooth muscle lid elevator (ptosis). The levator palpebra superioris, a striated muscle innervated by the oculomotor nerve CN-III, is not affected (Fig. 1-5).

Interruption of the sympathetic fibers outside the brain causes ipsilateral ptosis, anhidrosis, and miosis without abnormal ocular mobility.

Figure 1-5 Right Horner Syndrome.

Papilledema is characterized by elevation and blurring of the optic disk, absence of venous pulsations, and hemorrhages adjacent to and on the disk (Fig. 1-6). The finding of papilledema indicates increased intracranial pressure of any cause, including brain tumors, subarachnoid hemorrhage, metabolic processes, pseudotumor cerebri, and venous sinus thrombosis.



Optic fundus with papilledema

Visual field changes with enlarged blind spots secondary to chronic papilledema

Figure 1-6 Effects of Increased Intracranial Pressure on Optic Disk and Visual Fields.

III, IV, VI: Oculomotor, Trochlear, and Abducens Nerves

Our ability to acutely focus our eyes on an object of interest depends on being able to move the eyes together in a conjugate fashion; this requires three related cranial nerves that take their origin from various juxta midline midbrain and pontine nuclei. These provide us with the ability to astutely focus on an object of interest without concomitantly moving our head. Whether it is a detective watching a suspect or a teenager taking a furtive glance at a new classmate, these cranial nerves provide us with a broad sweep of very finely tuned motor function. There is no other group of

muscles that are so finely innervated as these. Their innervation ratio is approximately 20 : 1 in contrast to those of large muscles of the extremities with ratios between 400 and 2000 to 1. Certainly, this accounts for the fact that one of the earliest clinical manifestations of myasthenia gravis relates to the extraocular muscles (EOMs), where the interruption of just a few neuromuscular junctions affects the finely harmonized EOM function, leading to a skewed operation and thus double vision.

In order to identify isolated EOM dysfunction, it is most accurate to test each eye individually describing the observed specific loss of EOM function. For example, when the eye cannot be turned laterally, the condition is labeled as an *abduction paresis*, as opposed to CN-VI palsy. This is because the responsible lesion can be at any one of three sites, namely, cranial nerve, neuromuscular junction, or muscle per se. A more detailed assessment of these cranial nerves is available in Section II, Chapter 5.

The medial longitudinal fasciculus (MLF) is responsible for controlling EOM function because it provides a means to modify central horizontal conjugate gaze circuits. The medial longitudinal fasciculus connects CN-III on one side and CN-VI on the opposite side. Understanding the circuit of horizontal conjugate gaze helps clinicians appreciate the relation between the frontal eye fields and the influence it exerts on horizontal conjugate gaze (Fig. 1-6) as well the reflex relation between the ocular and vestibular systems (Fig. 1-7).

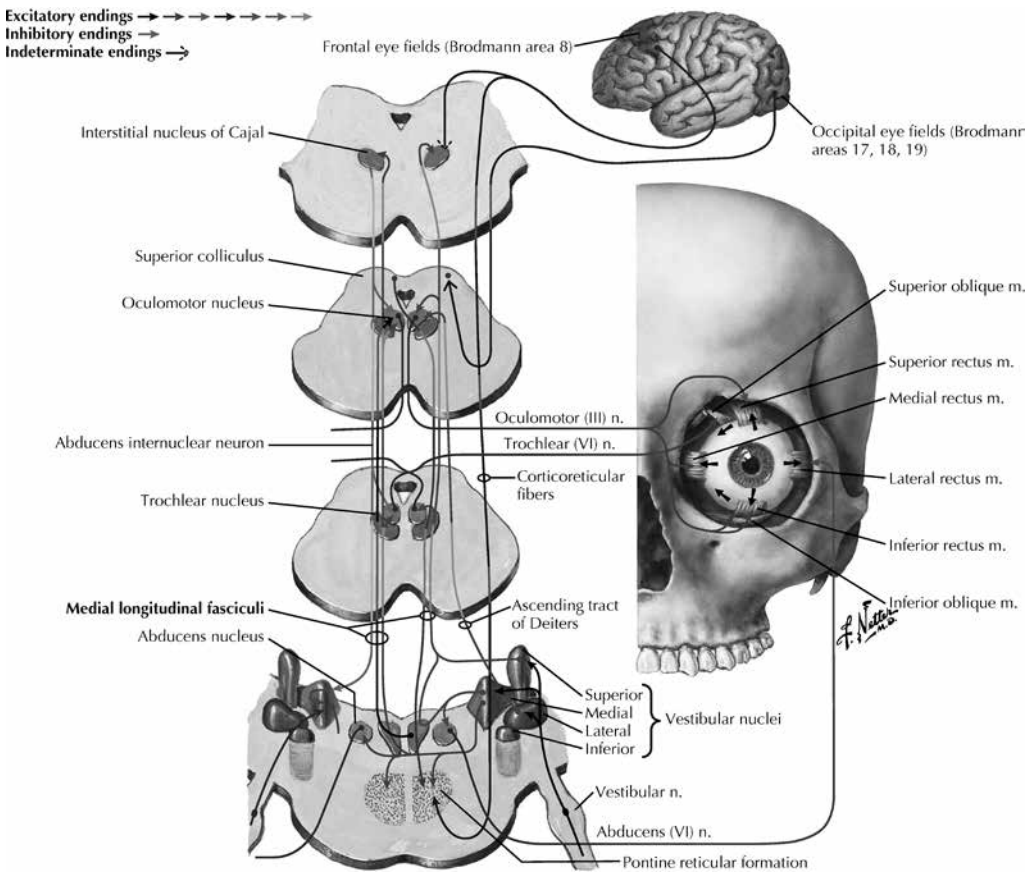


Figure 1-7 Control of Eye Movements.

V: Trigeminal Nerve

Our ability to perceive various stimuli applied to the face depends almost entirely on this nerve; whether as a warning to protect oneself from subzero cold, something potentially threatening to our eyesight, or the pleasurable sensation from the kiss of a beloved one, all forms of sensations applied to the face are tracked to our brain through the trigeminal nerve (Fig. 1-8). The primary sensory portion of this nerve has three divisions, ophthalmic, maxillary, and mandibular;

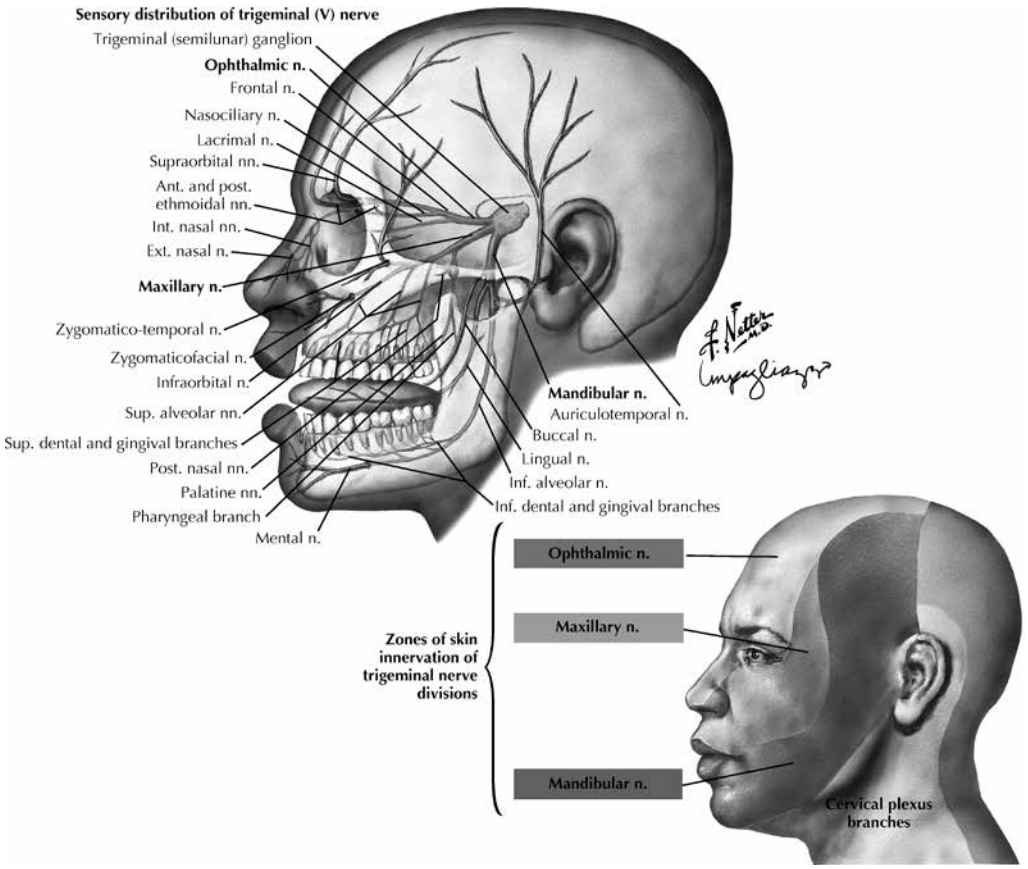


Figure 1-8 Trigeminal Nerve Neuralgia.

they respectively supply approximately one third of the face from top to bottom, as well as the anterior aspects of the scalp. The angle of the jaw is spared within the trigeminal mandibular division territory. This provides an important landmark to differentiate patients with conversion disorders or obvious secondary gain as they are not anatomically sophisticated and will report they have lost sensation in this.

The clinical testing of trigeminal nerve function includes both appreciation of a wisp of cotton and a sharp object on the facial skin per se as well as the corneal reflex. To evaluate the broad spectrum of facial sensation, that is, touch, pain, and temperature, the examiner uses a cotton wisp; the tip of a new, previously unused safety pin; and the cold handle of a tuning fork. In a symmetric fashion, the physician asks whether the patient can perceive each stimulus in the three major divisions of the trigeminal nerve supplying the face.

The *corneal reflex* depends on afferents from the first division of the trigeminal nerve combined with facial nerve efferents. This is also best tested using a wisp of cotton approaching the patient from the side while she or he looks away. Normally, both eyelids close when the cornea on one side is stimulated; this is because this reflex involves multisynaptic brainstem pathways.

Lastly, there is a primary motor portion that is part of the trigeminal nerve. It primarily supplies the muscles of mastication. It is best assessed by having the patient bite down and try to open the mouth against resistance.

VII: Facial Nerve

Facial expression is one of our very important innate human attributes allowing one to demonstrate a very broad spectrum of human emotions, especially happiness and sorrow; these are primarily dependent on the facial nerve (Fig. 1-9). The motor functions of CN-VII are tested by asking patients to wrinkle their forehead, close their eyes, and smile. Whistling and puffing up the cheeks are other techniques to test for subtle weakness. When unilateral peripheral weakness affects the facial nerve after it leaves the brainstem, the face may look "ironed out," and when the patient smiles, the contralateral healthy facial muscle pulls up the

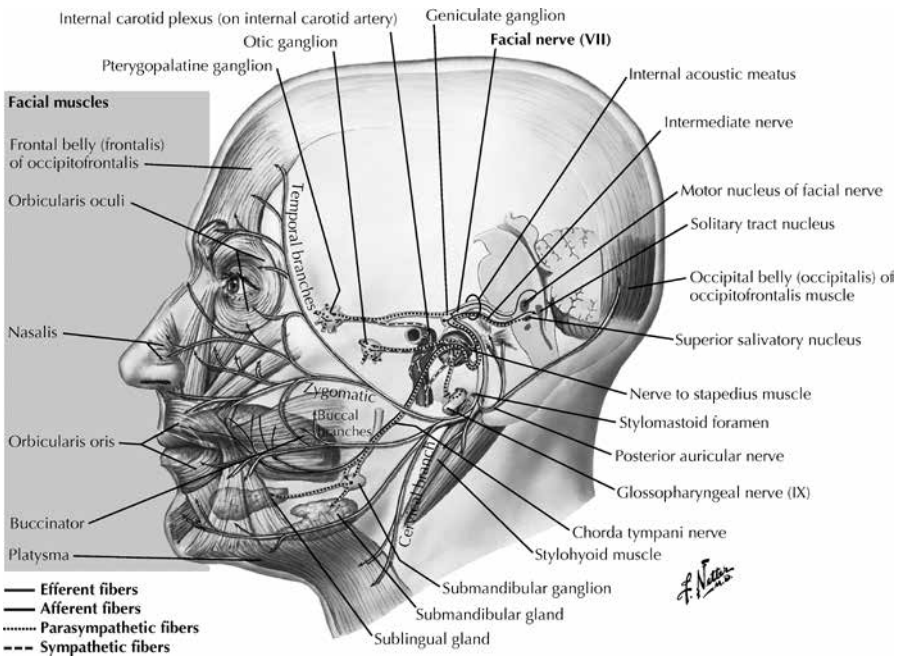


Figure 1-9 Facial Nerve With Its Muscle Innervation.

opposite half of the mouth while the affected side remains motionless. Patients often cannot keep water in their mouths, and saliva may constantly drip from the paralyzed side. With peripheral CN-Ⅶ palsies, patients are also unable to close their ipsilateral eye or wrinkle their foreheads on the affected side. However, although the lid cannot close, the eyeball rolls up into the head, removing the pupil from observation. This is known as the Bell phenomena.

In addition, there is another motor branch of the facial nerve; this innervates the stapedius muscle. It helps to modulate the vibration of the tympanic membrane and dampens sounds. When this part of the facial nerve is affected, the patient notes hyperacusis. This is an increased, often unpleasant perception of sound when listening to a phone with the ipsilateral ear.

Lastly, the facial nerve has a few other functions. These include prominent autonomic function, sending parasympathetic fibers to both the lacrimal and the salivary glands. It also subserves the important function of taste, another function providing both safety from rancid food and pleasure from a delightful wine. There is also a tiny degree of routine skin sensation represented for portions of the ear.

VIII: Cochlear and Vestibular Nerves

Beyond the simple test of being able to hear at all, more sophisticated clinical evaluation of CN-Ⅷ is often challenging for the neurologist. Fortunately our otolaryngologic colleagues are able to precisely measure the appreciation of specific auditory frequencies in a very sophisticated manner. Barring the availability of these formal audiometric evaluations, simple office-based hearing tests sometimes help us demonstrate diagnostically useful asymmetries. Using a standard tuning fork, it is possible to differentiate between *nerve (perceptive) deafness* caused by cochlear nerve damage and that caused by *middle ear (conduction) deafness* with two different applications of the standard tuning fork. We are able to test both air and bone conduction.

Initially a vibrating tuning fork is placed on the vertex of the skull, *Weber test*, allowing bone conduction to be assessed. Here the patient is asked to decide whether one ear perceives the sound created by the vibration better than the other (Fig. 1-10). If the patient has nerve deafness, the vibrations are still appreciated more in the normal ear. In contrast, with conduction deafness, the vibrations are better appreciated in the abnormal ear.

The *Rinne test* is carried out by placing this vibrating instrument on the mastoid process of the skull. Here the patient is asked to identify the presence of sound. As the vibrations of the tuning fork diminish, eventually the patient is unable to appreciate the sound. At that instant,

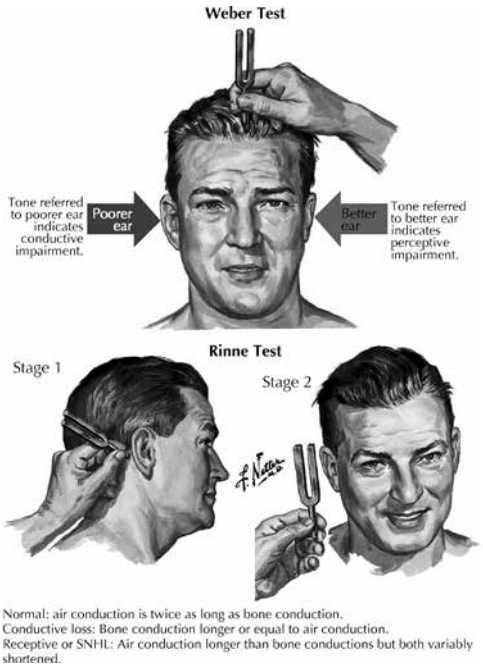


Figure 1-10 Auditory Nerve Testing: Weber and Rinne Testing.

the instrument is moved close to the external ear canal to evaluate air conduction. If the individual has normal hearing, air conduction is longer than bone conduction. When a patient has nerve (perceptive) deafness, both bone and air conductions are diminished, but air conduction is still better than bone conduction. In contrast with *conduction deafness*, secondary to middle ear pathology, these findings are reversed. Here, when the patient's bony conduction has ceased, air conduction is limited by the intrinsic disorder within the middle ear. Therefore, the sound can no longer be heard; that is, it cannot pass through the mechanoreceptors that amplify the sound and thus cannot reach the auditory nerve per se.

IX, X, XI: Glossopharyngeal, Vagus, and Accessory Nerves

The most common complaints related to glossopharyngeal-vagal system dysfunction include swallowing difficulties (dysphagia) and changes in voice (dysphonia). A patient with a glossopharyngeal nerve paresis presents with flattening of the palate on the affected side. When the patient is asked to produce a sound, the uvula is drawn to the unaffected side (Fig. 1-11). Indirect mirror examination of the vocal cords may demonstrate paralysis of the ipsilateral cord. The traditional test for gag reflex, placing a tongue depressor on the posterior pharynx, is of equivocal significance at best, because the gag response varies significantly and patients evidence wide varieties of tolerance to this stimulus. Preservation of swallowing reflexes is best tested by giving the

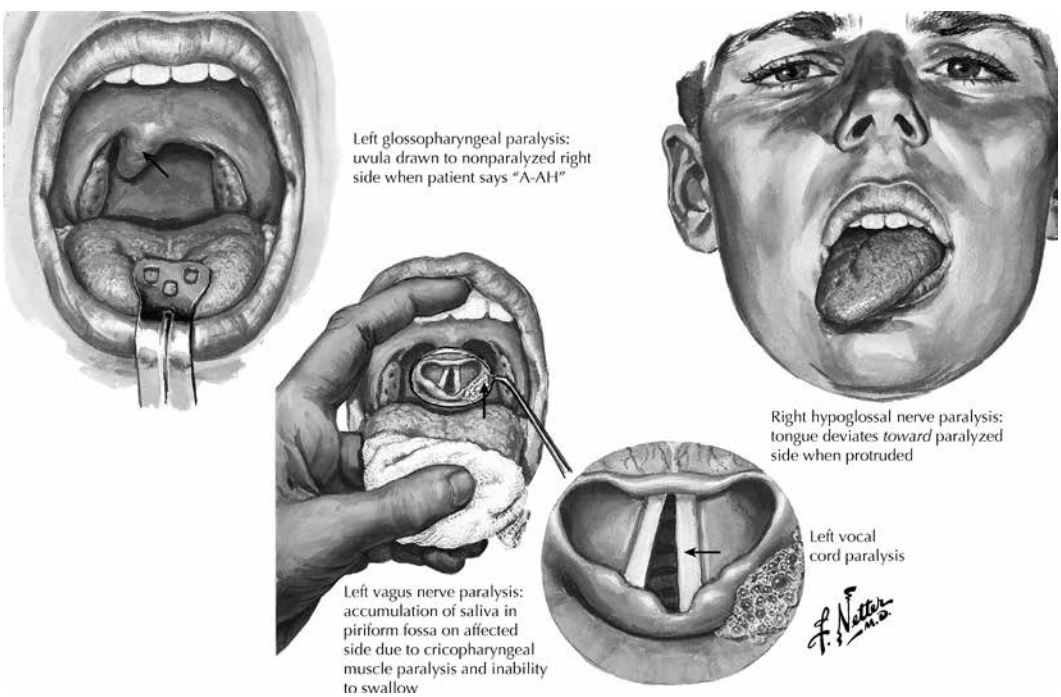


Figure 1-11 Uvula, Tongue, and Vocal Cord Weakness.

patient 30 mL of fluid to drink through a straw while seated at 90°. Patients with compromised swallowing reflexes develop a "wet cough" and regurgitate fluids through their nose. Intracranial or proximal spinal accessory nerve damage limits the ability to turn the head to the opposite side (weakness of the ipsilateral sternocleidomastoid muscles and trapezius muscle). More distal accessory nerve damage is most commonly seen following surgical misadventures during biopsying a lymph node from the posterior triangle of the neck, sparing the sternocleidomastoid but affecting the trapezius, causing dysfunction and winging of the scapula.

XII: Hypoglossal Nerve

Damage to the hypoglossal nucleus or its nerve produces tongue atrophy and fasciculations The fasciculations usually are seen best on the lateral aspects of the tongue. If the nerve damage is unilateral, the tongue often deviates to that side (Fig. 1-11). Two means to test for subtle weakness include asking the patient to push against a tongue depressor held by the examiner and having the patient push the tongue into the cheek.

CEREBELLAR DYSFUNCTION

Evaluation of posture and gait provides the opportunity to observe the most dramatic clinical manifestations of cerebellar dysfunction. The patient presenting with *midline cerebellar lesions* affecting the vermis characteristically assumes a broad-based stance when walking that typically mimics an inebriated individual. At the extreme, these individuals are unable to maintain a stance. In contrast, when there is a *cerebellar hemisphere problem*, the patient has a tendency to veer to the affected side. With midline lesions, gait is usually unchanged whether the eyes are open or closed, suggesting that this is not the result of disruption of proprioceptive inputs. Patients with unilateral lesions are often able to compensate with their eyes open but deteriorate when they lose visual inputs.

Loss of limb coordination is the result of cerebellar inability to calculate inputs from different joints and muscles and coordinate them into smooth movements. This abnormality is best observed by testing *finger-to-nose* and *heel-to-shin* movements and making bilateral comparisons. When performing the finger-to-nose test, the examiner provides his or her finger as the target; it is sequentially moved to different locations. The patient in turn keeps the arm extended and tries to touch the examiner's finger at each location. When unilateral cerebellar dysfunction is present, the patient *overshoots* the target, so-called *past pointing*. It is important not to misinterpret such findings as always of cerebellar origin, as patients with focal motor or sensory cerebral cortex lesions may present with mild arm weakness and proprioceptive sensory loss affecting that limb. In this setting, a degree of focal limb dysmetria may develop; this is sometimes difficult to distinguish from primary cerebellar dysfunction. One clinical means to distinguish cerebellar from cerebral cortical dysfunction is that the patient with cerebellar hemisphere lesions will have these movements improve after a few trials. In contrast, with cerebral cortical dysmetria, repeated trials only lead to further deterioration in the attempted action.

Dysdiadochokinesia is a sign of cerebellar dysfunction that occurs when the patient is asked to rapidly change hand or finger movements, that is, alternating between palms up and palm down. Patients with cerebellar dysfunction typically have difficulties switching and maintaining smooth, rapid, alternating movements.

Tremor, *nystagmus*, and *hypotonia* are other important indications of potential cerebellar dysfunction. *Tremors* may develop from any lesion that affects the cerebellar efferent fibers via the superior cerebellar peduncle. This is characterized by coarse, irregular movement. *Nystagmus* may occur with unilateral cerebellar disease; the nystagmus is most prominent on looking to the affected side. *Hypotonia* may be present but is often difficult to document. This is best observed when testing a patient's muscle stretch reflexes at the quadriceps tendon knee jerk. Here, the normal "check" does not occur after the initial movement, so the leg on the affected side swings back and forth a few times after the initial patellar tendon percussion.

GAIT EVALUATION

Whenever possible, the neurologic clinician is encouraged to personally greet the patient, watching them arise from their chair and initiate their gate. Next, before moving to the examination room the patient needs to be observed walking in

the hallway. On occasion it is important to observe the patient on stairs particularly if there is a query about proximal weakness. A smooth gait requires multiple inputs from the cerebellum and primary motor and sensory systems. Gait disorders provide a very broad differential diagnostic challenge that results from lesions in any part of the neuraxis (Fig. 1-12).

Frontal lobe (Fig. 1-12, D) processes including

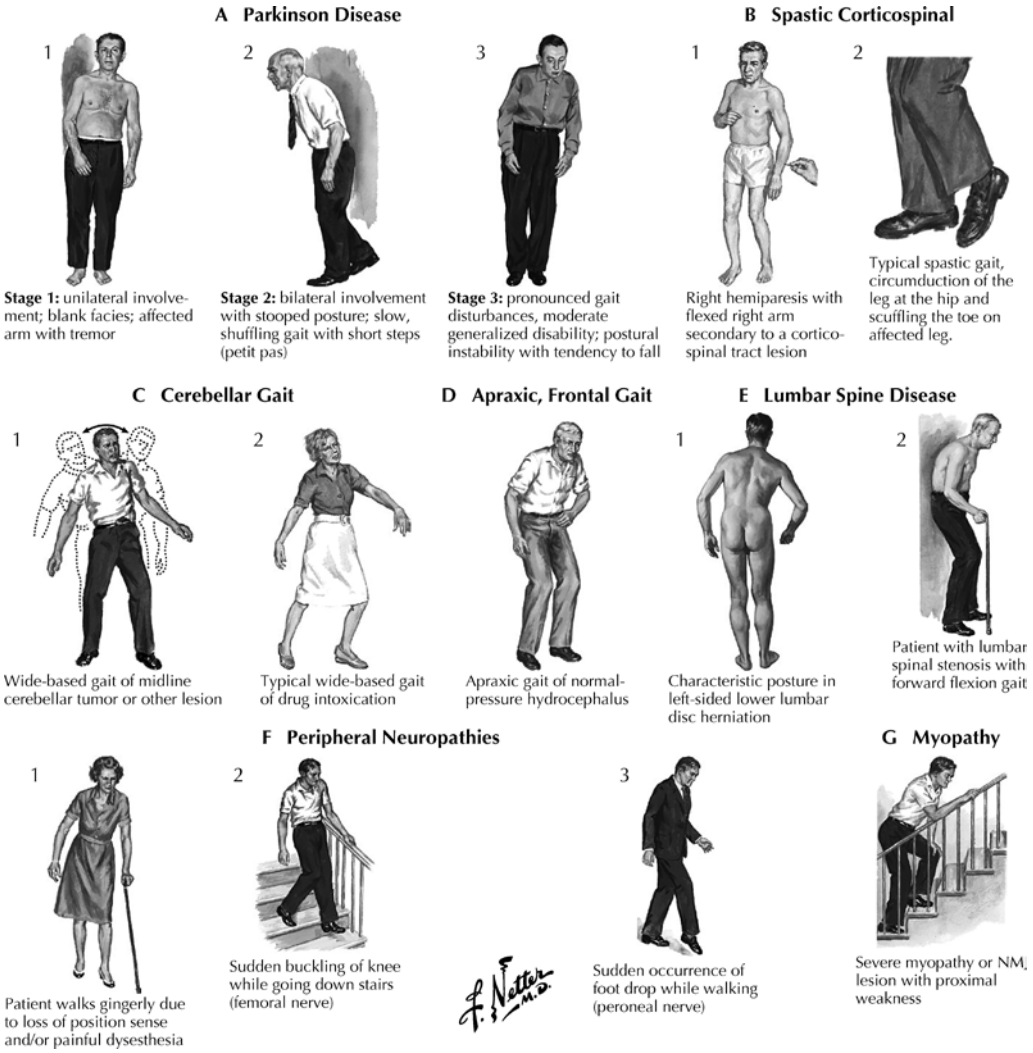


Figure 1-12 Gait Disorder Characteristics and Etiology.

tumors and normal-pressure hydrocephalus lead to apraxia, spasticity, and leg weakness. *Spasticity* per se is a nonspecific marker of corticospinal tract disorders that may arise with various neurologic lesions between the frontal lobe and the distal spinal cord (Fig. 1-13). Various *neurodegenerative* conditions, particularly those affecting the basal ganglia, such as *Parkinson disease* (Fig. 1-12, A1–3), are some of the most common causes of gait difficulties. These are typically manifested by slowness initiating gait, small steps, and eventually gait festination, wherein once patients begin to accelerate their walking, they take increasingly more rapid but paradoxically smaller steps. There is an innate, almost wax-like rigidity to their stooped body carriage, including the frozen posture of one or both arms that usually lack the normal arm swing. Very occasionally, a change in posture from the seated position to attempted gait will be manifested by a dystonic posturing, which may be indicative of another genetic disorder, dystonia musculorum deformans or paroxysmal choreoathetosis.

Cerebellar disorders related to midline anterior cerebellar *vermis* lesions or various *heredofamilial spinocerebellar* entities lead to a broad-base gait

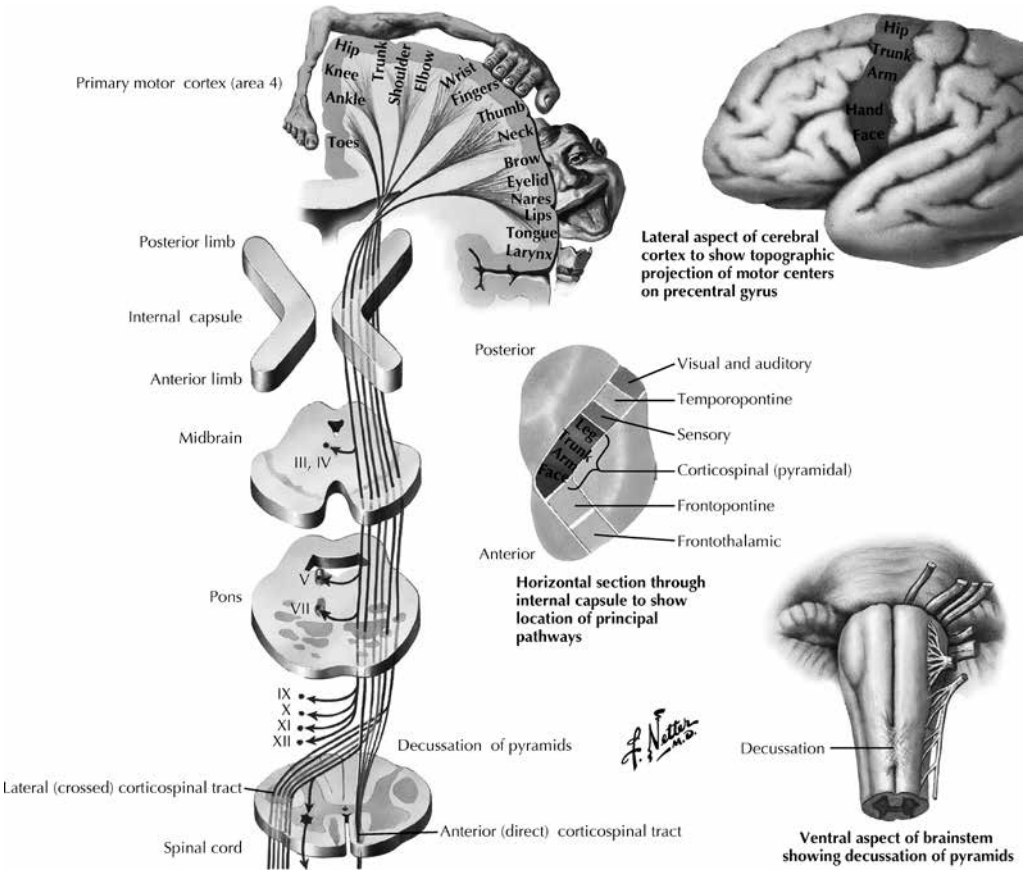


Figure 1-13 Pyramidal System, Corticospinal Tract. Gait Disorders Can Arise From Interruption of These Pathways at Any Level.

ataxia (Fig. 1-12,C1–2). The patient is asked to walk in tandem, with one foot in front of the other. It is an effective means to elicit a subtle disequilibrium often related to midline cerebellar dysfunction such as with simple entities, including alcohol intoxication.

Myelopathies with posterior column dysfunction, such as vitamin B_{12} deficiency, present with loss of proprioception function. These particularly affect the patient's gait in dark environments, as do some of the *peripheral neuropathies, especially those with a primary sensory ganglionopathy* (Fig. 1-12, F1). Testing for the presence of a Romberg sign is an excellent clinical marker for these disorders. Here patients are asked to stand in place with their eyes open, gain their equilibrium, and then close their eyes. Individuals with various proprioceptive disorders are unable to maintain their balance when visual clues are withdrawn; such a condition is referred to as a positive Romberg sign. One of the earliest signs, and at times a prominent sign of a myopathy, is needing to push off the arms of a chair when arising to walk. When these individuals do walk their gait may be a broad-based gait mimicking an anterior cerebellar lesion. When viewed from the side the curve of their low back is accentuated, i.e. hyperlordotic. Both the wide base and the hyperlordosis are representative of weakness of the most proximal muscle groups—the iliopsoas, quadriceps, and glutei—as well as the paraspinal axial musculature.

ABNORMAL ADVENTITIOUS MOVEMENTS

Neurologists are frequently consulted to evaluate various adventitious movements, including tremors, chorea, dyskinesias, and ballismus. The most common movement disorder encountered in the office is "essential tremor," usually a "benign" hereditary condition that generally

does not herald a progressive neurodegenerative process. These patients often seek medical attention because they are concerned that their tremors are a sign of Parkinson disease. Therefore, differentiating between different types of tremors is a common and important concern. An essential tremor characteristically occurs during certain voluntary actions, such as when bringing a cup of coffee to the mouth. In contrast, with classic Parkinson disease, the pill-rolling tremor is primarily evident at rest and when the patient is seated or walking and disappears with the spontaneous use of the extremity. A subtle fidgeting may represent the earliest sign of Huntington or Sydenham chorea. Very rarely a patient will present with a more energetic, purposeless, wing beating movement of an extremity referred to as hemiballismus. A full discussion of movement disorders and their presentation is found in Section.

MUSCLE STRENGTH EVALUATION

Partial limb weakness is referred to as *monoparesis*. Total limb paralysis is referred to as *monoplegia*. Unilateral weakness of the limbs is referred to as hemiparesis or *hemiplegia*. Paraparesis refers to involvement of both legs; if no motor function remains, this is considered *paraplegia*. Similarly, *quadriplegia* relates to total paralysis of all 4 extremities.

Grading Weakness

The traditional, most widely used British system for quantifying degrees of weakness is based on a scoring range of 0 to 5, with 5 being normal. The extremes of grading are easy to understand, although the subtle grading between 4 and 5 (i.e., >4, 4, >4, or <5) may be slightly different depending on the examiner's own strength (Table 1-2). Other systems judges the patient to have mild (<1), moderate (<2), severe (<3), or total paralysis (<4) strength, and this grading is viewed by some of us to be a simpler and more reproducible methodology. When testing individual muscles of the patient, the examiner must recognize that this is not an athletic match but rather a determination of whether the patient has normal strength. There is a significant range of normal, and a sense of that latitude can be gained only by examining multiple individuals.

Table 1-2 Grading System for Clinical Documentation of Degree of Weakness

Grade	Clinical Findings
0	No movement (complete paralysis)
1	Able to move a muscle but no movement of limb
2	Minor movement of limb but inability to overcome gravity
3	Moderate weakness; movement of limb against gravity
4	Mild weakness; some resistance against mild pressure
5	Normal; resistance against moderate pressure

Adapted from Brain. Aids to the Examination of the Peripheral Nervous System. *4th ed. Philadelphia: WB Saunders; 2000.*

Motor Lesions

CEREBRAL CORTEX

When evaluating patients with *focal weakness due to brain lesions*, one should document the evolution of symptoms and any associated changes in sensation or pain. Sudden onset of localized weakness, without preceding trauma or associated pain, suggests ischemic or hemorrhagic cerebral damage. CNS processes cause preferential weakness of the arm extensors and leg flexors. Pure motor weakness of the arm and leg, with slurring of speech, is the hallmark of a stroke in the posterior limb of the internal capsule. Strokes involving the brainstem typically have corticospinal weakness associated with cranial nerve findings. Language deficits usually point to a left hemispheric processes. Neglect of the affected arm or hand, in association with variable degrees of left-sided weakness, often occurs with pathologic processes in the right hemisphere. Visual field deficits may also develop, depending on whether there is concomitant involvement of the optic nerve, chiasm, tract, radiation, or optic cortex.

BRAINSTEM BULBAR WEAKNESS

Rarely, the weakness may be confined to the *brainstem bulbar musculature*, leading to difficulty speaking, chewing, swallowing, or even breathing. Posterior inferior cerebellar artery (PICA) *infarcts* often present with these symptoms accompanied by vertigo, and crossed body sensory loss. Lesions at the *motor neuron* levels such as bulbar amyotrophic lateral sclerosis (ALS), or hypoglossal nerve injury from carotid artery dissection, also require consideration in this setting. Similar symptoms are rarely presenting signs of peripheral *nerve* lesions, including Guillain–Barré and tick paralysis, the *neuromuscular junction*, such as myasthenia gravis and botulism, and rarely *inflammatory myopathies*.

Poliomyelitis and diphtheria are always suspected in the rare geographic areas these disorders are still endemic. Fortunately, these are now more of historical interest where modern immunization programs are successful.

MYELOPATHIES

It is necessary to differentiate weakness caused by *spinal cord* lesions from brain disorders. Primary lesions affecting the spinal cord include compressive lesions from progressive spondylosis (thickening of the bony spinal canal), metastases, trauma, demyelinating processes, particularly MS or transverse myelitis, and spinal epidural abscess. Depending on the location and temporal profile, spinal cord lesions often begin with subtle symptoms of gait disturbance, weakness, or both. Concomitantly, spinal cord lesions are usually associated with sensory findings and urinary bladder difficulties. Pain frequently accompanies acute spinal cord lesions; localized spine and or radicular pain from concomitant nerve root involvement is typical of metastatic cancer, epidural abscess, or transverse myelitis. These disorders can rapidly lead to paraplegia.

A very careful examination is crucial in order to define the presence of a sensory level; this is often best documented by using pin and temperature modalities. One must either sit the patient up or turn them on their side, carefully moving the sensory stimulus from the buttocks to the neck to see if there is a sudden change in degree of perception characteristic of a "sensory level." Failure to perform this evaluation may lead to missing a treatable spinal cord lesion. Detailed knowledge of the specific sensory territories of the nerve root dermatomes (Fig. 1-14) is very helpful when assessing potential spinal cord lesions. Looking for a sweat level is also sometimes helpful because the skin below the level of a significant spinal cord lesion will be noticeably drier from loss of autonomic sympathetic innervation. Acute lower extremity weakness is also seen with the Guillain–Barré syndrome or other acute generalized polyneuropathies. These disorders may mimic a primary spinal cord lesion.

Patients with *painless asymmetric weakness*

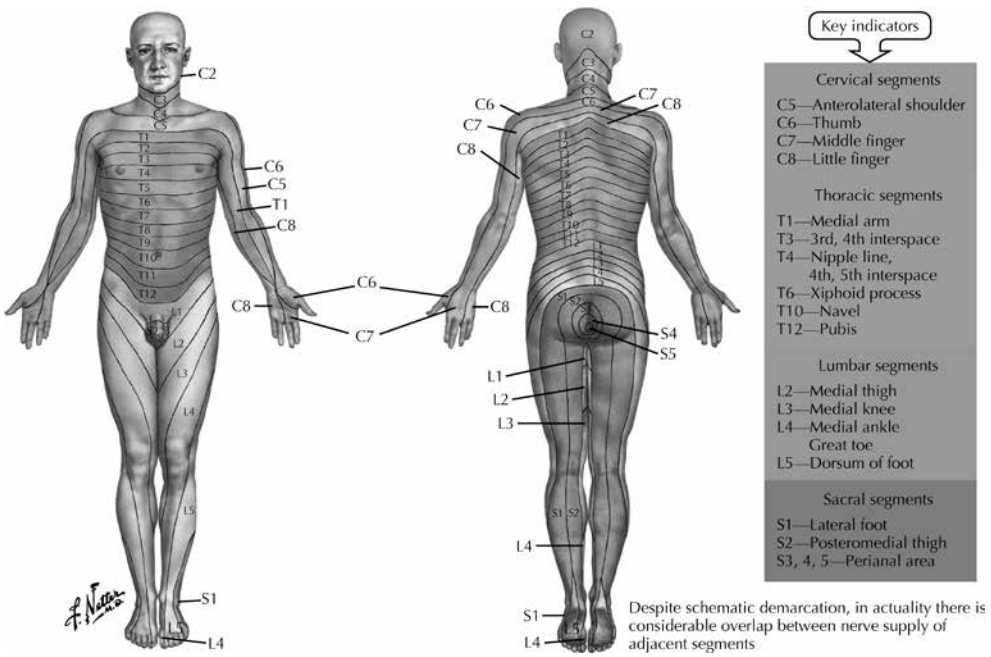


Figure 1-14 Dermatomal Levels.

typically have primary motor neuron or very occasionally motor nerve root, motor nerve level demyelinating lesions. Fasciculations, spontaneous firing of small groups of muscle fibers innervated by a single motor axon (a motor unit), commonly accompany lower motor neuron weakness. Although often perceived by the patient as twitching or jumping, fasciculations may not be easily seen with the naked eye. Sometimes it may be necessary to observe a specific muscle for several minutes to see these signs. Fasciculations are quite common and often benign; when present in isolation with no motor weakness or muscle atrophy, and the patient has a normal EMG, there

is little chance that the individual has primary motor neuron disease. Typically lower motor nerve lesions have a concomitant diminution of specific MSRs; however, with ALS the MSRs are exaggerated and often accompanied by Babinski signs.

NERVE ROOT, PLEXUS, OR PERIPHERAL NERVE

The presence of cervical or lumbosacral pain with concomitant focal extremity numbness or weakness is characteristic of a radiculopathy. Interspinal disc herniation and spinal stenosis are the most common processes affecting individual nerve roots. Because sensory examination is the most subjective part of the neurologic examination, occasionally it is difficult to clearly define. Sometimes the patient, per se, can provide the most accurate assessment by using his or her finger to outline the area of diminished sensation. It often then becomes clear that the pattern of sensory loss specifically fits the distribution of a particular peripheral nerve or nerve root dermatome. Knowledge of the cutaneous sensory supply of peripheral nerves is essential to perform an accurate and useful clinical sensory examination (Fig. 1-15).

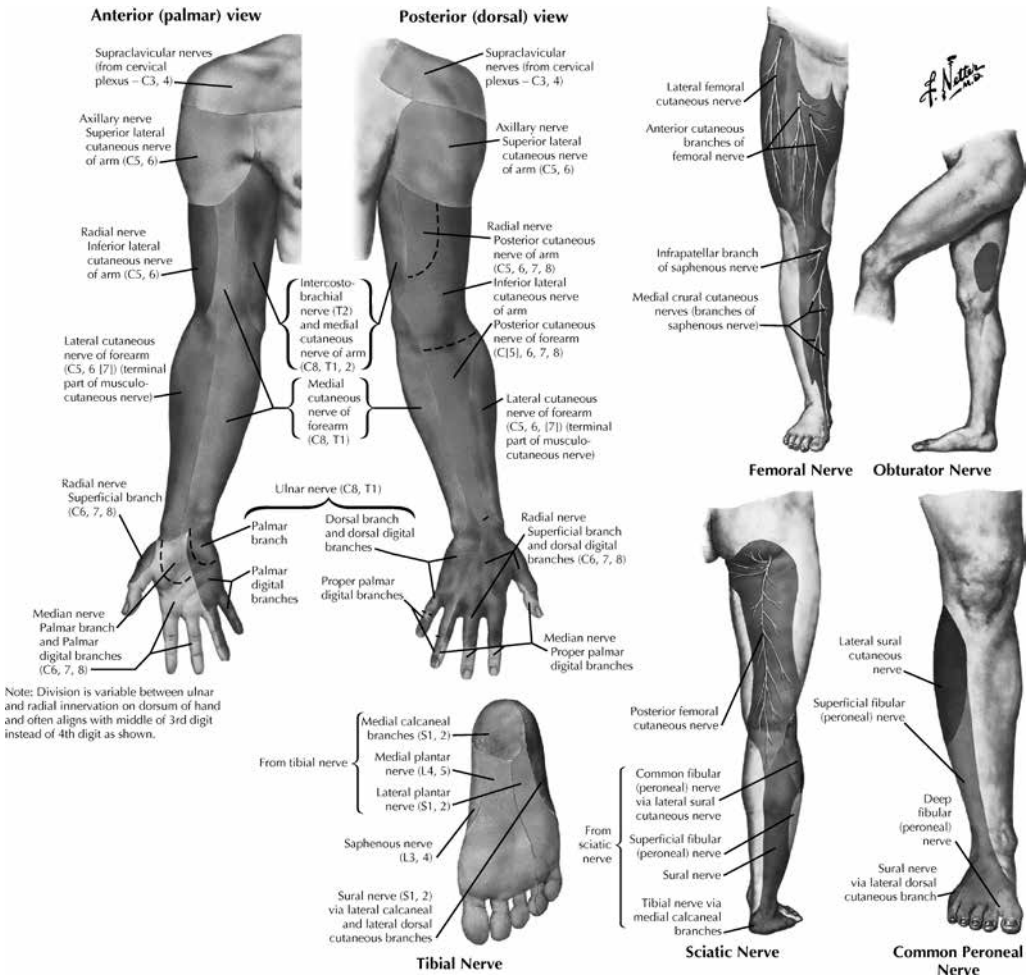


Figure 1-15 Cutaneous Innervations.

Some peripheral mononeuropathies, or rarely multifocal motor neuropathies, present with unilateral peripheral weakness; in particular, the wrist drop of radial nerve lesions and foot drop of peroneal nerve lesions are mistaken for processes above the foramen magnum, often mimicking a stroke. Understanding the motor distribution of the major peripheral nerves ultimately aids in the correct diagnosis. Although a peroneal nerve lesion causes a foot drop, similarly an L5 nerve root lesion also presents with a foot drop but usually with associated low back pain. Additionally, the L5 lesion also produces weakness of the posterior tibial muscle innervated by the tibial nerve; this provides

the means to make a clinical distinction from a common peroneal nerve lesion. Rarely, lesions as high as the parasagittal frontal lobe within the brain may also present with foot weakness.

Atrophy of muscles innervated by the involved nerve occurs when there is significant denervation. Measuring extremity circumference may document significant side-to-side asymmetries and, by inference, muscle atrophy secondary to anterior horn cell, nerve root, or peripheral nerve damage. It is most important also to carefully search for sensory loss, such as one finds with the ulnar nerve lesion often presenting with painless intrinsic hand muscle atrophy mimicking ALS or syringomyelia.

MUSCLE DISORDERS

Most *myopathic* processes lead to *symmetric proximal weakness*, although such can occur with other disorders, particularly chronic inflammatory demyelinating polyneuropathy or rare neuromuscular transmission defects, such as LEMS. Neck flexor and arm extensor weakness may provide early signs of a myopathic process, especially with myasthenia gravis and the inflammatory myopathies. At its most extreme, these patients may present with a floppy head. On rare occasions, primary myopathies have an asymmetric distribution, particularly inclusion body myositis, that mimics ALS or fascioscapulohumeral muscular dystrophy.

MOTOR TONE

The motor system depends on multiple inputs in order to provide precise, well-synchronized, and smooth muscle function. These include positive inputs from the cerebrum, basal ganglia, cerebellum, brainstem, and spinal cord through the corticospinal tracts. Projections from the pontine reticular formation and reticulospinal tract also have direct connections with motor neurons innervating the proximal and axial body musculature. These fibers also originate from the cerebrum and cerebellum and have a primary inhibitory function that serves to decrease motor tone. Subsequent to damage of structures above the pontine reticular formation, this circuit loses its inhibitory input from the cerebrum and cerebellum, leading to excessive stimulation of motor neurons, especially with antigravity muscles, including arm flexors and leg extensors leading to a flexed and pronated arm posture and an extended and adducted leg position. This increase in tone is referred to as spasticity. This has an interesting paradox in that at rest the spastic muscle has limited tone, but if there is a sudden attempt by the examiner to change the posture the limb is easily moved for a very short distance and then the degree of resistance immediately and rapidly increases up to a maximum and then dissipates. This resembles a "clasp knife," i.e., pocket knife, resistance/relaxation.

Four primary types of changes in tone are found in patients with primary CNS disease: hypotonia, spasticity, flaccidity, and rigidity. It is important to place these observed changes in motor tone within the context of the complete neurologic examination rather than in isolation. The patient's *body tone* is best evaluated when the individual is fully relaxed. Sometimes, it is useful to check tone more than once during the examination. *Tone* is described as the patient's primary level of muscular tension. To become comfortable with this part of the examination, it is important, as with other portions of the neurologic evaluation, to routinely check these parameters in healthy individuals to establish one's normal base of observations.

Hypotonia

This is occasionally demonstrable in patients with cerebellar hemispheric lesions. For example, the distal part of the ipsilateral extremity may not be able to perform rapid alternate movements (called dysdiadochokinesia) because of the inability to maintain a stable posture. Similarly, the smooth, straight pursuit seen when one elicits the knee MSR loses the out-and-back motion that typically has an inhibitory cerebellar check. Instead, on return, there is overshoot with no check, leading to a repetitive pendular response. This classic hypotonic cerebellar tone is a relatively uncommon finding.

A more generalized loss of normal tone is most commonly seen among infants with either central or peripheral motor unit disorders, classically spinal muscular atrophy (Werdnig–Hoffmann disease) or the various congenital myopathies. Although a similar example is seen in adults, rarely, a floppy head syndrome develops in an older patient.

Flaccidity

This is the term for a total loss of tone and is seen in various disease processes affecting the upper motor neurons. Most commonly, this occurs in acute settings such as with a recent stroke or a sudden spinal cord injury, that is, spinal shock. However, with both of these, the flaccidity is temporary

and tone increases later to present in the form of varying degrees of spasticity.

Spasticity

Extremes of muscle tone that are maximal at the initiation of the physician's attempt to move the limb and then suddenly release partway through the movement (a clasp-knife, spastic release) are the typical findings seen with a spastic limb. Significant degrees of spasticity are easily elicited with any reasonable stimulation of muscles that induces the stretch reflex. More subtle spasticity may be obvious only with stretching the muscle in a specific direction and at a specific rate. Increased tone, such as may occur with stroke or spinal cord injury, evolves from a flaccid state to spasticity over a matter of days to weeks subsequent to the initial neurologic injury.

Decerebrate Rigidity

When there is total loss of a motor neuron inhibition, as may occur with an upper brainstem injury, the syndrome of decerebrate rigidity develops. Here, a simple noxious stimulus leads to bilateral extension in unison of all four extremities, with the arms pronated and the legs adducted (Fig. 1-16) rotated inward. Most commonly, one sees this in the setting of cardiac arrest or from shear injuries to the brainstem resulting from severe head injuries, most typically from automobile accidents or battlefield injuries. When these patients survive 1 to 3 months, and are otherwise totally unresponsive, they are said to be in a *persistent vegetative state*.

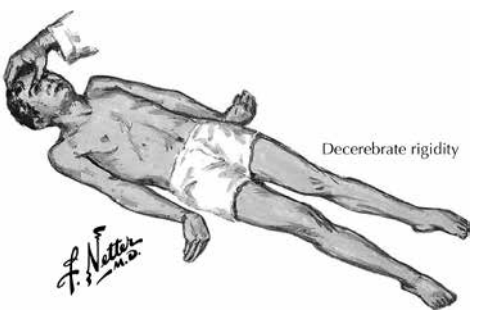


Figure 1-16 Motor Tone Abnormality.

Rigidity

Increasing tone from basal ganglia disorders, as may occur with Parkinson disease, is known as rigidity. Rigidity creates a continuous sense of tightness in the attempt to move the joint through a full excursion from extension to flexion.

MUSCLE STRETCH REFLEXES, CLONUS, AND THE BABINSKI SIGN

During the neurologic examination, MSRs (named for the specific muscle stretched) are usually readily elicited by tapping lightly over the muscle insertion tendon or while palpating the tendon and then percussing the palpating digit. Occasionally, it is difficult to obtain MSRs in healthy individuals. In this setting, it is sometimes useful to distract the patient or apply techniques that reinforce the reflex to potentiate the appearance of the MSRs. The most common method is the Jendrassik maneuver, wherein patients flex their fingers, interlocking one hand with the other and pulling on the count of 3 while the clinician percusses the appropriate tendon at the knee or ankle. For the upper extremities, the patient may be asked to clench the contralateral fist as the neurologist percusses over the arm tendons, activating the intrafusal muscle spindle.

When grading MSRs, the extremes are easy to appreciate and range from 0 to 4. A reflex grading of 0 is indicative of complete lack of MSR. A generalized loss of reflexes is pathologic and is known as areflexia; this typically occurs in Guillain–Barré syndrome. Briskly responding MSRs are graded as 4 and are typical of a prior stroke or spinal cord lesion. When the patient has brisk MSRs, a single Achilles tendon percussion sometimes elicits a repetitive series of dorsi and plantar movements in the foot. This is known as *clonus*. This does not commonly occur spontaneously, but clonus may be elicited by giving a quick snap to the dorsiflexed foot as it is held in the palm of the hand. This also occurs, rarely, at the quadriceps tendon. Here the reflex is graded as 4+. The remainder of the grading is very logical. A reflex of 1 is a mere contraction of the muscle; a 2 is a contraction.

The *Babinski sign* is an important pathologic reflex that is elicited at the lateral, plantar surface of the foot using subtle, very careful stroking with a tongue depressor or the base of a key. The great toe extends, and the remaining toes fan out (Fig. 1-17).

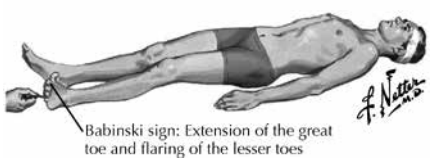


Figure 1-17 Elicitation of the Babinski Sign.

A more exaggerated response, known as *triple flexion*, includes flexion of the hip, knee, and foot, often with a Babinski response. Because this reflex primarily depends on sensory stimulation of the foot, a kind, gentle, nonirritating stimulus is best to obtain an accurate response. It absolutely does not require excessive or painful pressure. With sensitive or ticklish patients, appropriate responses can usually be obtained from a careful stimulation of the lateral outside, not plantar, surface of the foot. However, some patients have a withdrawal response wherein the foot and entire set of toes dorsiflex. This is often overcome by separately pulling down on the middle toe while carefully stimulating the sole in traditional fashion.

The clinical circumstance where there is a combination of brisk MSRs, clonus, and a Babinski sign indicates an *upper motor neuron lesion*. These abnormalities result from various pathophysiologic mechanisms originating in the brain or spinal cord. The many possibilities include destructive cerebral lesions, such as stroke, tumor, encephalitis, and spinal cord trauma, or demyelinating disorders such as MS affecting the spinal cord, the brain, or both. Additionally, signs of upper motor neuron lesions are sometimes observed in patients during the postictal period after a seizure or in patients who have toxic or metabolic encephalopathies. Therefore, although brisk MSRs and a Babinski sign are nonspecific regarding the anatomic setting of the CNS abnormality, their presence provides unequivocal evidence of anatomic persistent upper motor neuron pathology, with the exception of the postictal or encephalopathic setting.

SENSORY EXAMINATION

In most clinical settings, it is best to separate the sensory examination into two major categories, that is, those derived from superficial skin receptors or deeper mechanoreceptors. The former are small, unmyelinated, slowly conducting type C fibers or larger, slightly myelinated, somewhat more rapidly conducting type A-delta fibers. These small fibers primarily subserve *pain and temperature* (respectively tested using a pin point or a cold object such as the handle of a tuning fork) and gross touch modalities. The large, well-myelinated type A-alpha and A-beta fibers carry the kinesthetic modalities of *position sense* studied by the examiner's passively moving the finger or toe in the vertical plane and asking the patient which direction the digit was moved, either up or down.

Fine tactile discrimination is evaluated by using a pair of calipers to check their ability to recognize whether one or two points are applied to the digit. *Vibratory sensation* depends on both deep afferent and cutaneous sensory modalities subserved by type A-alpha fibers. It is best tested by a 128-Hz tuning fork that typically has a low frequency rate and longer duration of action. This modality is the one that most commonly diminishes in sensitivity with aging.

Classic Syndromes of Peripheral Sensory Dysfunction

Generalized polyneuropathies typically present with symptoms of numbness and tingling at the tips of the toes and, later, fingers, that is, a stocking-glove distribution (Fig. 1-18). Eventually, this loss will gradually spread proximally past the ankles and wrists into the legs and forearms but usually not above the knees and elbows. On examination with a cold object, a pin (for small fiber function), a tuning fork, and position sense (if large fibers are also involved), the examiner notes a distal loss that is maximum in the periphery and gradually reaches normal at a more proximal site.

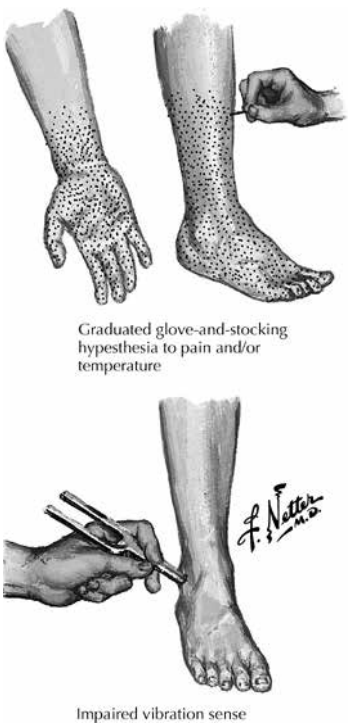


Figure 1-18 Documentation of Various Types of Sensory Modalities in a Peripheral Neuropathy.