Clinical Neurologic Evaluation

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H. Royden Jones, Jr., and Kinan Hreib

The neurologic sciences are the most intellectually challenging, unequivocally fascinating, and tremendously stimulating of the various clinical disciplines. Initially, the vast intricacies of basic neuroanatomy and neurophysiology often seem overwhelming to both medical student and neuroscience resident alike. However, eventually the various portions of this immense knowledge base come together in a discernible pattern, not unlike a Seurat canvas. Often one is expanding or revisiting our neurologic base as we are challenged by variations on the theme of our previous experiences. It is the keen observation and coding of these clinical experiences that leads the astute neurologic physician to solve new patient challenges.

One must first and foremost be an astute historian initially listening very carefully to the patient. Most often the intricacies, as well as the subtleties, of the neurologic history provide the essential foundation leading to a rational and structured neurologic examination as well as the appropriate diagnostic testing. Although it is easy to define the requisite methodology to examine the neurologic patient, it is much more challenging to similarly address the history acquisition other than making a few generalities. One of the most important elements of neurologic training is the opportunity for the student and the resident to observe senior neurologists evaluate a patient. As a resident, this was absolutely one of our most important learning experiences. Too often the student does not appreciate the elegance illustrated by a carefully derived neurologic clinical history. A major attribute of a skillful and successful neurologist is being an astute listener. This requires the neurologist to bring together various seemingly disparate and subtle data from the patient's various concerns and then focus on this information with specific questions to decide on its relevance to the issues at hand. Understanding the temporal profile of the patient's symptoms is crucial; were the symptoms' onset acute and stable or have they followed an ingravescent course? Very often, this information provides a most important perspective that is one of the very important keys to diagnosis.

Clinical Vignette

A 42-year-old woman with juvenile autoimmune diabetes mellitus came for further investigation of her extremely painful neuropathy initially presumed secondary to diabetes, or possibly to recent chemotherapy for breast cancer. However, her temporal profile was the final clue to her diagnosis. On careful review of the onset of her symptoms, it was found that she had never had the slightest hint of intolerable paresthesiae until awakening from her mastectomy. Her pain had begun precipitously in the recovery room. It was steady from its inception and totally incapacitating in this previously vigorous woman whose favorite pastime was backpacking in mountainous national forests. This temporal profile was in total contradistinction to any symmetric diabetic or antineoplastic chemotherapy-related polyneuropathy. These disorders always have a clinical course of a subtle onset and very gradual evolution.

With this information, we investigated what transpired at the time of her breast surgery when she awakened with this extremely limiting painful neuropathy. In fact, she had had a general anesthetic with nitrous oxide (N_2O) induction. This N_2O uncovered a second autoimmune disorder, namely vitamin B_{12} deficiency. The anesthetic had precipitously led to symptoms in this previously clinically silent process. Fortunately, vitamin B_{12} replacement led to total resolution of her symptoms.

Comment: In this instance, her initial physicians had let themselves be trapped by what was familiar to them because diabetes is the most common cause for a painful neuropathy. However, only rarely does it lead to a precipitous onset of symptoms. The fine-tuning of this patient's temporal profile, especially the abrupt onset of symptoms, led us to seek a more detailed history as to whether some toxic process was operative. Review of the operative records per se led to the diagnosis when the suspicion of nitrous oxide intoxication was confirmed.

Most neurologic disorders follow a well-defined clinical paradigm. However, it is their very broad clinical perspective that continually challenges the astute neurologic clinician to maintain a vigilant intellectual posture. When these specific clinical subtleties are appreciated, the clinician is rewarded with the knowledge of having done the very best for his or her patient as well as having the intellectual rewards for being on the cutting edge of the clinical neurosciences. The skillful clinician, taking a very careful history, is the one most able to recognize the attributes of something quite uncommon presenting in a fashion more easily confused with more mundane afflictions.

For example, numbness or tingling in a patient's hand most commonly represents entrapment of the median nerve at the wrist, reflecting the presence of a very common disorder known as the *carpal tunnel syndrome*. However symptoms of this type may occasionally represent early signs of a pathologic lesion at the level of the brachial plexus, nerve root, spinal cord, or brain per se. It is imperative for the clinician to always consider a broad anatomic perspective in each patient evaluation. When this approach is not carefully followed, less common, and potentially treatable disorders may not be diagnosed in a timely fashion. It is absolutely imperative that no compromise be made in obtaining a thorough and accurate history when first meeting the patient. This is the most important interchange the physician will have. It needs to be taken in a relaxed, hopefully noninterrupted setting allowing for privacy. Additionally, it is very important to invite the spouse, parent, or significant other into the room. Rarely will a patient object to same; having another close

observer of a patient's difficulties available can provide insight that may be essential to diagnosis. A thorough initial evaluation engenders a patient-family sense of trust in the physician as a detailed history, with a careful examination demonstrates a major commitment. Once developed, this clinical setting encourages the patient to communicate openly with their physician as they outline their diagnostic plans and eventually a treatment formulation. This chapter provides a foundation that will serve as an anchor for both the student and resident as they learn the art and science of the performance of detailed neurologic evaluations.

NEUROLOGIC HISTORY AND EXAMINATION

An accurate history requires paying attention to detail, often observing the patient's demeanor while reading the patient's body language, having the opportunity to witness the patient's difficulties, and interviewing family members. History taking is a special art and science in its own right. It is a skill that requires ongoing additions to one's own interviewing techniques. Listening to the patient is a most important part of this exercise; it is something that can be more time consuming than current clinical practice "time allowed guidelines" provide for within various patient settings. This approach provides the diagnostic keystone that often distinguishes an astute clinician's ability to find a diagnosis where others have failed.

A complete neurologic examination also requires carefully honed acquired skills. For example, the ability to decide whether the patient is truly weak and not giving way, or similarly does or does not have a Babinski sign present, often makes the difference between arriving at a correct diagnosis. The ability to define a sensory loss at a spinal cord level is another very crucial exercise.

One of the most challenging clinical scenarios occurs with the patient who has already seen another clinical neurologist and no diagnosis has been made. The patient is frustrated, as often was his or her prior neurologist. To be fair to the patient, as well as oneself, when evaluating such an individual seeking another neurologic opinion it is important to gain one's own initial and totally unbiased history and examination. Furthermore, in order to prevent unwelcome bias, the new neurologist should avoid reading other colleagues' notes or looking at previous neurologic images prior to gaining his or her own history and performing the examinations.

Although time-consuming, the history is the most important factor leading to accurate diagnoses. One of the essential attributes of a skillful neurologist is the ability to be a good listener so as not to miss crucial historic points. It is important to begin the initial meeting by asking patients why they have come; this offers them the opportunity to express concerns in their own words. If at all possible, the neurologist should not interrupt, thus providing the patient the opportunity to provide their primary concerns to the neurologist, emphasizing the symptoms of greatest importance. Rarely, anxious or compulsive patients may speak of their concerns at great length; with experience, physicians learn to make discreet interjections to maintain control of the evaluation and draw the patient back from extraneous tangents.

When the patient's primary concerns are established, specific issues can be explored. Additionally, making careful observations during the review of history allows better focus for subsequent questions. An accurate baseline assessment of mental status and language can be obtained from listening to the patient and observing responses to questions. It is through listening that the clinician gains insight into the patient's real concerns. For example, it is not unusual to see a patient referred to a neurologist for evaluation of headaches, which only became exacerbated with the recent discovery of a brain tumor in someone known to the patient.

Unfortunately, the economics of modern health care has forced primary care physicians and specialists to shorten visit times with patients and their families. One must be fastidious not to use diagnostic tools, such as magnetic resonance imaging (MRI), as substitutes for careful clinical history and examination. The current detailed medical information available on the Internet, in conjunction with a more sophisticated basic health education environment, has indeed enhanced patients' knowledge bases, although not always in a balanced format. Patient expectations sometimes affect the diagnostic approach of physicians. In this environment, it is not surprising that imaging techniques such as MRI and computed tomography (CT) have replaced or supplemented a significant portion of clinical judgment. However, even the most dramatic test findings may prove irrelevant without appropriate clinical correlation. To have patients unnecessarily undergo surgery because of MRI findings that have no relation to their complaints may lead to a tragic outcome. Therein lies the importance of gaining a complete understanding of the clinical issues.

Although neurology may seem in danger of being subsumed by overreliance on highly sophisticated diagnostic studies, this needs to be kept in perspective as many of these innovations have greatly improved our diagnostic skills and therapeutic capacities. For example, much knowledge regarding the early recognition, progression, and response to treatment of multiple sclerosis (MS) depends on careful MRI imaging.

It is essential to make patients feel comfortable in the office, particularly by fostering a positive interpersonal relationship. Taking time to ask about patients' lives, education, and social habits often provides useful clues. A careful set of questions providing a general review of systems may lead to the key diagnostic clue that focuses the evaluation. When the patient develops a sense of confidence and rapport with an empathetic physician, he or she is more willing to return for follow-up, even if a diagnosis is not made at the initial evaluation. Sometimes a careful second or third examination reveals a crucial historic or examination difference that leads to a specific diagnosis. Follow-up visits also allow the patient and physician to have another conversation regarding the symptoms and concerns. Some patients may come to their first office visit with an exhaustive list of concerns and symptoms, whereas others provide minimal information. Subsequent visits are therefore intended not only to discuss the results of tests but also to clarify the symptoms and or response to treatment. If patients feel rushed on their first visit, they may not return for follow-up, thus denying the neurologist a chance at crucial diagnostic observations. The physician–patient relationship must always be carefully nurtured and highly respected.

APPROACH TO THE NEUROLOGIC EVALUATION

Throughout training, examination skills are continually being amplified as the resident is exposed to an ever-evolving clinical experience. One of the most important is the opportunity to observe the varied skill sets demonstrated by academic neurologists as they approach different types of patients. One of the essentials for appropriate interpretation of the neurologic patient evaluation is learning how to elicit important, sometimes subtle, clues to diagnosis; an appreciation of what is "normal" at different ages is also important. A hasty history and examination can be misleading. For example, briskly preserved ankle reflexes in an elderly patient is not normal, whereas moderately diminished vibration sense is normal at the ankles. For example, a diagnosis of early MS may be missed by not asking about such things as previous problems with visual function, shooting electric paresthesiae when bending the neck (Lhermitte sign), or sphincter problems manifested by increasing urgency to urinate.

Even though carpal tunnel syndrome is the most common cause for a patient to experience a numb hand, one must always be fastidious not to overlook other potential pathoanatomic sites, such as within the peripheral nervous system at the level of the more proximal median nerve, the brachial plexus, or the cervical nerve root. In another instance, the failure to undress a patient whom one suspects to have a presumably benign cause for a numb hand, that is, carpal tunnel syndrome, may preclude the examining physician from recognizing the presence of an unexpected positive Babinski response indicative of a central nervous system (CNS) lesion. Similarly, identifying a sensory level is indicative of a myelopathy as the pathophysiologic explanation for the patient's numb hand. Lastly the finding that the sensory loss in the fingers primarily involves position sense and stereognosis becomes the entre to examine the cerebral cortex as the site for these complaints.

Another important outcome from performing a complete neurologic examination at the initial evaluation in almost every patient is that this not only establishes the patient's current status but will provide a baseline for future comparison. There are certain "normal" asymmetries in many individuals, often not previously appreciated by the patient per se or his or her relatives. These may include a patient's slightly asymmetric smile, somewhat irregular pupils, or hint of ptosis. However, at times such findings do take on significant meaning. As an example, a middle-aged woman was thought to have benign tension headaches. This was based on a "normal" neurologic exam elsewhere. However she had an asymmetric smile that previously had not been appreciated. Imaging studies identified a frontal lobe tumor contralateral to her weakness. Thus, the careful observation of seemingly subtle clinical findings may prove to have significant bearing on the issue at hand. Even when these findings are proven to be "normal variants," clear documentation may often be very helpful during the course of the patient's illness or later on when new concerns occur. In that setting, the prior definition of what proves to be a normal asymmetry will prevent erroneous conclusions from being developed.

Formulation

One of the most intellectually challenging aspects of neurology relates to the neurologist's ability to amalgamate the historical and physical findings into a unitary hypothesis. One needs to first consider the multiple neuroanatomic sites that can potentially explain the patient's clinical presentation. Subsequently, this is placed in the perspective of the clinical temporal profile of symptom occurrence. Did all of the patient's symptoms begin abruptly, as usually seen with a stroke but sometimes with a tumor or demyelinating process? Or was there an evolution of degree of clinical loss or did new features gradually get added to the patient's findings as is characteristic of certain neoplastic lesions and sometimes more diffuse vasculitides. Formulation can be hindered by the patient's inability to provide an accurate history or participate in the neurologic examination. One of the more subtle and difficult conditions to recognize is anosognosia to one's illness, as may occur in patients with right parietal brain injury. Under these circumstances, the patient may not have sensory, visual, or motor neglect, but unawareness of cognitive, emotional, and other functional limitations. Family interview is most important in this setting.

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,

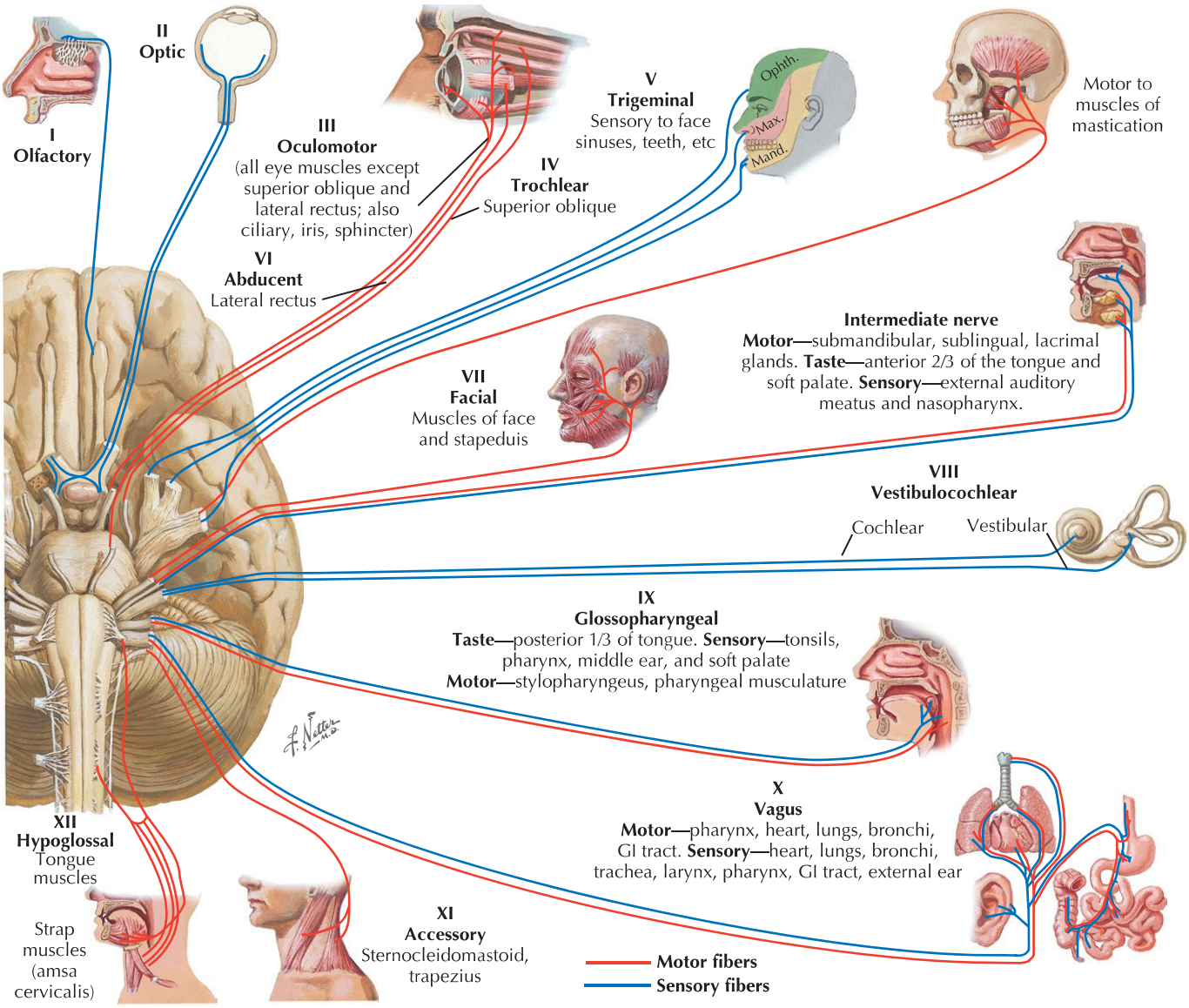


Figure 1-1 Cranial Nerves: Distribution of Motor and Sensory Fibers.

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.

CRANIAL NERVES: AN INTRODUCTION

The 12 CNs subserve multiple types of neurologic function (see 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

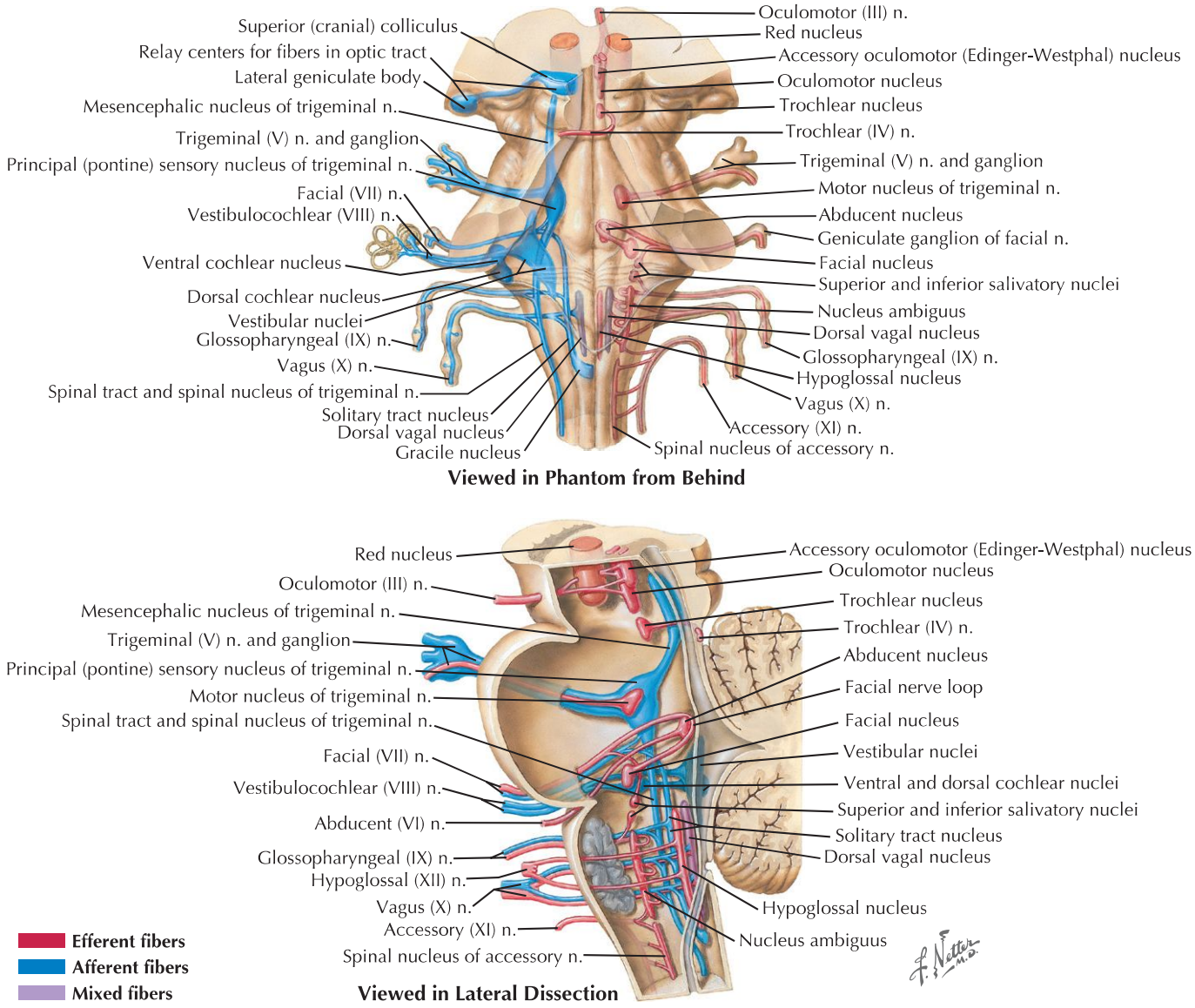


Figure 1-2 Cranial Nerves: Nerves and Nuclei.

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 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.

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

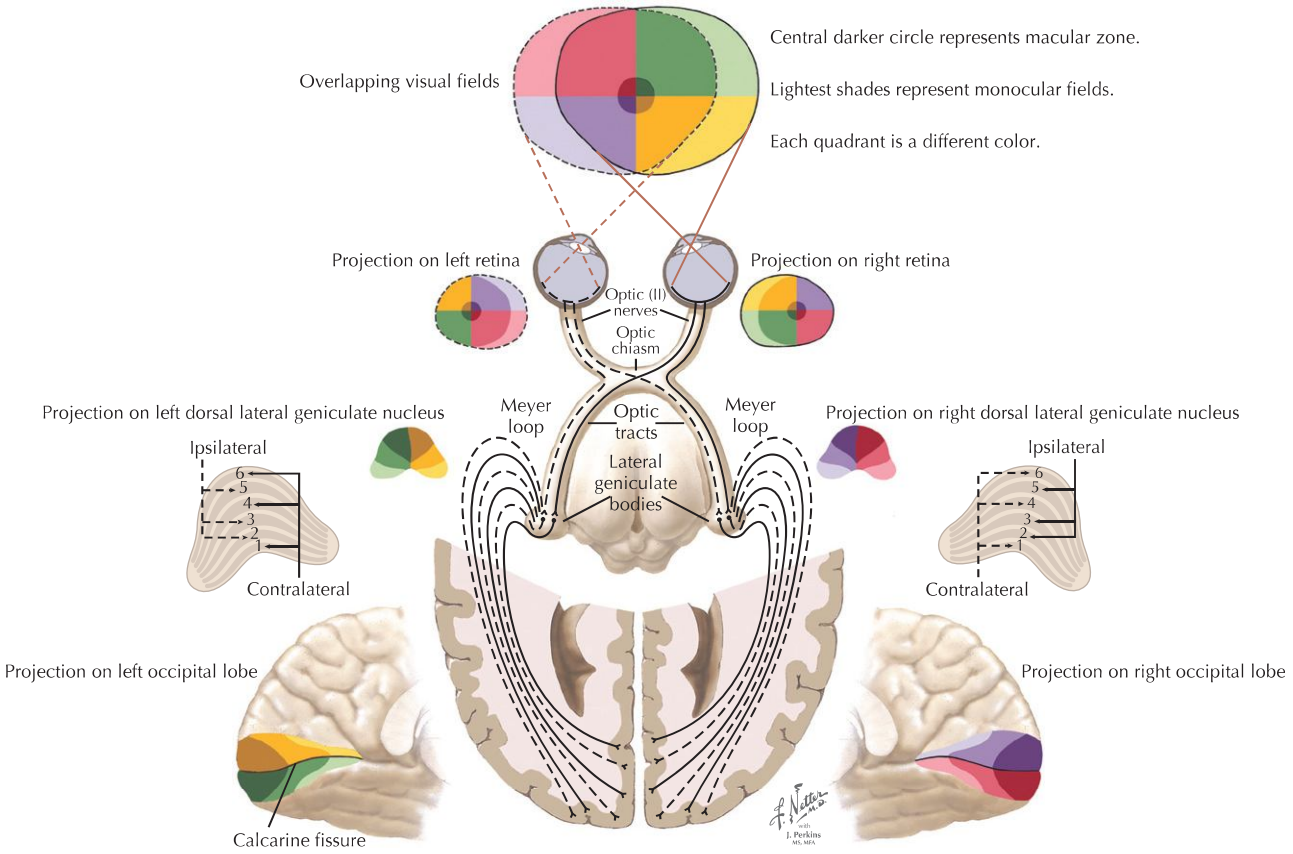


Figure 1-3 Visual Pathways: Retina to Occipital Cortex.

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.

In *kinetic perimetry*, a stimulus is moved from a non-seeing area (far periphery or physiologic blind spot) to a seeing area, with patients indicating at what point the stimulus is first noticed. Testing is repeated from different directions until a curve can be drawn connecting the points at which a given stimulus is seen from all directions. This curve is the isopter for that stimulus for that eye. The isopter plot has been likened to a contour map, showing "the island of vision in a sea of darkness." The Goldmann perimeter, a half-sphere onto which spot stimuli are projected, is the premiere device for this mapping. The normal visual field extends approximately 90° temporally, 45° superiorly, 55° nasally, and 65° inferiorly. Practically, this geographic shape mimics the oblique teardrop shape of aviator-style sunglass lenses.

In *static perimetry*, the test point is not moved, but turned on in a specific location. Typically automated, computer testing preselects locations within the central 30° of field. Stimuli are dimmed until they are detected only intermittently on repetitive presentation—this intensity level is called the threshold. The computer then generates a map of numeric values of the illumination level required at every test spot, or the inverse of this level, often called a sensitivity value. Values may also be displayed as a grayscale map, and statistical calculations can be performed—by comparing to adjacent spots or precalculated normal values or noting sudden changes in sensitivity—to detect abnormal areas.

Most visual field changes have localizing value: specific location of the loss, its shape, border sharpness (i.e., how quickly across the field the values change from abnormal to normal). Its concordance with the visual field of the other eye tends to implicate specific areas of the visual system. Localization is possible because details of anatomic organization at different levels predispose to particular types of loss (see Chapter 4).

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

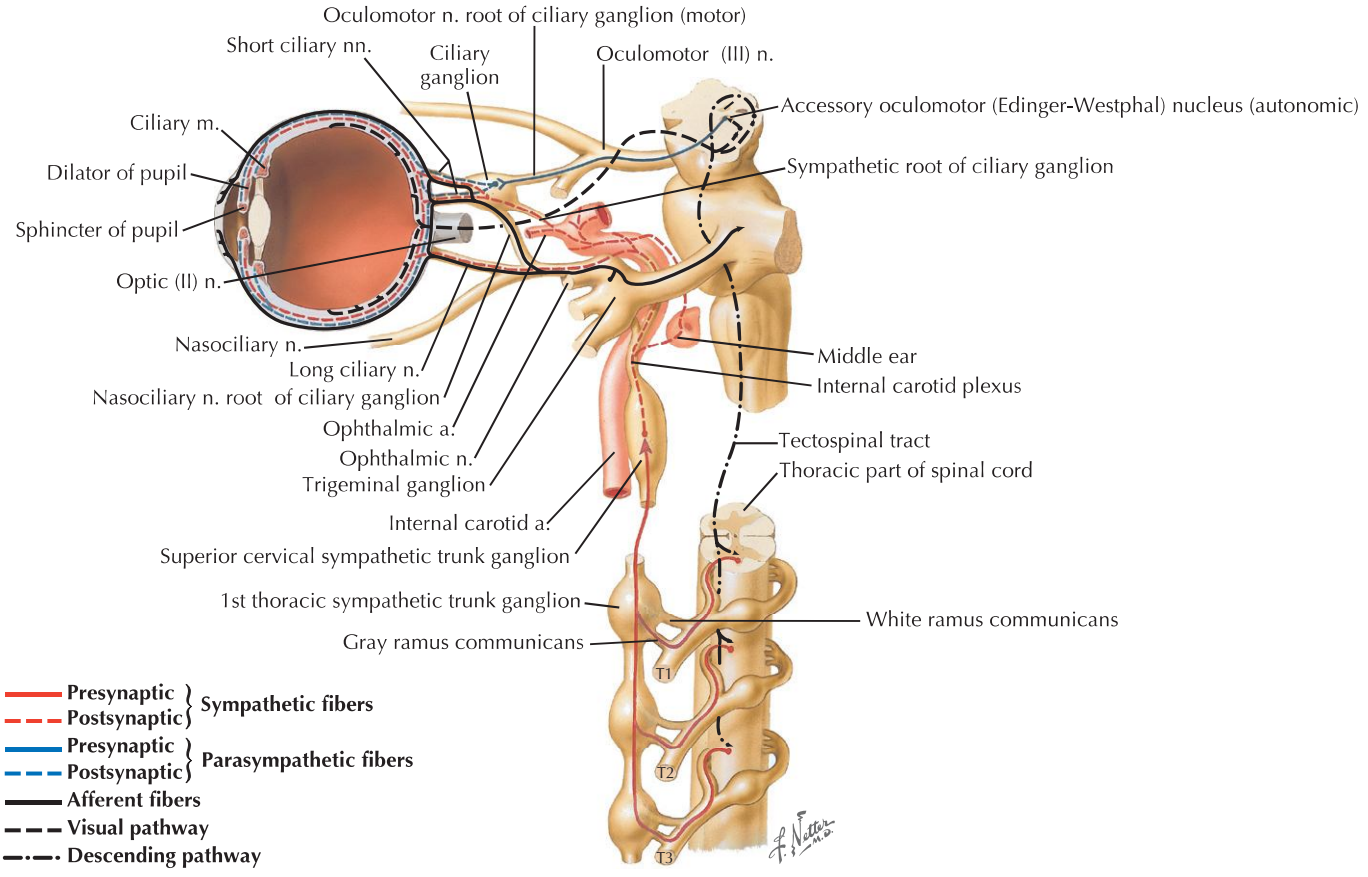


Figure 1-4 Autonomic Innervation of Eye.

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.

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 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.

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.

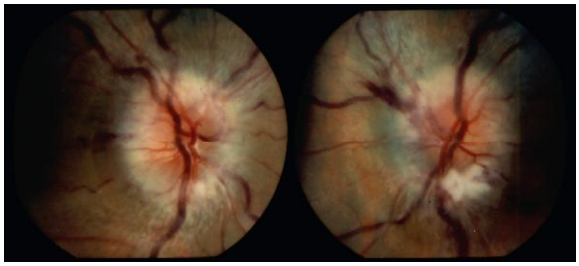
Interruption of the sympathetic fibers outside the brain causes ipsilateral ptosis, anhidrosis, and miosis without abnormal ocular mobility.

Figure 1-5 Right Horner Syndrome.

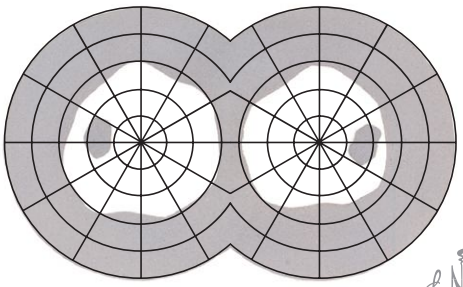
Sympathetic efferent fibers originate within the hypothalamus and traverse the brainstem and cervical spinal cord, then exit the upper thoracic levels and course rostrally to reach the superior cervical ganglia (see Fig. 1-4). Subsequently, these sympathetic fibers track with the carotid artery within 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).

OPTIC FUNDUS

The ability to peer into the patient's eye is a very unique and fascinating experience as it provides an opportunity to directly examine not only the initial portion of the optic nerve but also tiny arterioles and veins. This is the only portion of human anatomy that provides the physician with such an option. Here one may find signs of increased intracranial pressure or evidences of the effects of poorly controlled hypertension or diabetes mellitus. Today all of these various lesions are much less commonly observed because of much better treatment of systemic illnesses that affect the smaller blood vessels. Similarly the



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.

development of MRI and CT scanning makes it easier to identify intracerebral mass lesions at a much earlier stage of illness. Today as brain tumors no longer reach a critical size, obstructing cerebrospinal fluid flow, creating the increased intracranial pressure that leads to papilledema, this is now a relatively rare finding but one that still demands recognition.

A careful optic funduscopic examination is essential in the evaluation of very many neurologic disorders. This evaluation is best performed in a relatively dark environment that leads to both a reflex increase in pupillary size and improvement in contrast of the posterior chamber structures. Findings that should be documented include optic nerve margins, venous pulsations, and the presence of hemorrhages, exudates, or any obvious obstruction to flow by embolic material (such as cholesterol plaque in patients complaining of transient visual obscuration), and pallor of retinal fields that may reflect ischemia.

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.

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 (see Fig. 1-6) as well the reflex relation between the ocular and vestibular systems (Fig. 1-7).

The connection of the vestibular system to the medial longitudinal fasciculus can be tested by two different means. One is the doll's-eye maneuver. Here the patient's head is rotated side to side while the examiner watches for rotation of the eyes. Passive movement of the head to the left normally moves the eyes in the opposite direction, with the left eye adducting and the right eye abducting. The opposite occurs when the head is rotated to the right.

Ice-water caloric stimulation provides another option to study vestibular ocular MLF pathways. This is primarily used for the examination of comatose patients; on very rare occasions, it is extremely helpful for rousing a patient presenting with a suspected nonorganic, that is, feigned coma. Patients are placed at an elevation of approximately 45°. Next, the tympanic membranes are checked for intactness, and then 25–50 mL of ice water is gradually infused into each ear. A normal response in the awake patient, after left ear stimulation, is to observe slow deviation of the eyes to the left followed by rapid movement (nystagmus) to the right (see Fig. 1-10). In contrast, the comatose patient with an intact brainstem has a persistent ipsilateral deviation of the eyes to the site of stimulation with loss of the rapid eye movement component to the opposite side.

The center for vertical conjugate gaze and convergence is also located within the midbrain, although the underlying circuit is not well delineated. The vertical conjugate gaze centers can be tested by flexion of the neck while holding the eyelids open and watching the eye movements. When CNS processes affect conjugate gaze, such as with MS, a prominent nystagmus is often defined. The nystagmus is thought to result from an

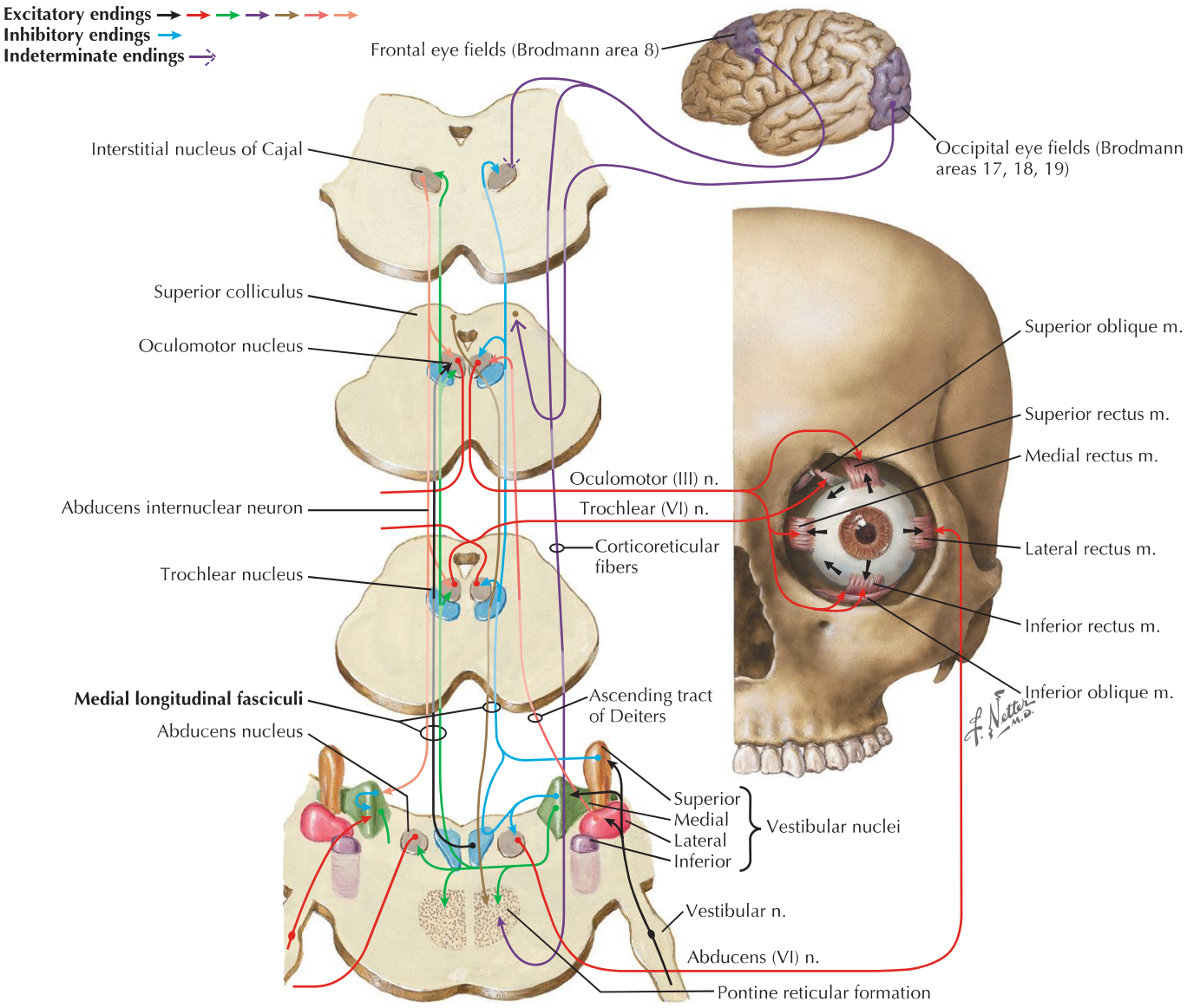


Figure 1-7 Control of Eye Movements.

attempt to maintain conjugate function of the eyes and minimize double images.

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; 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.

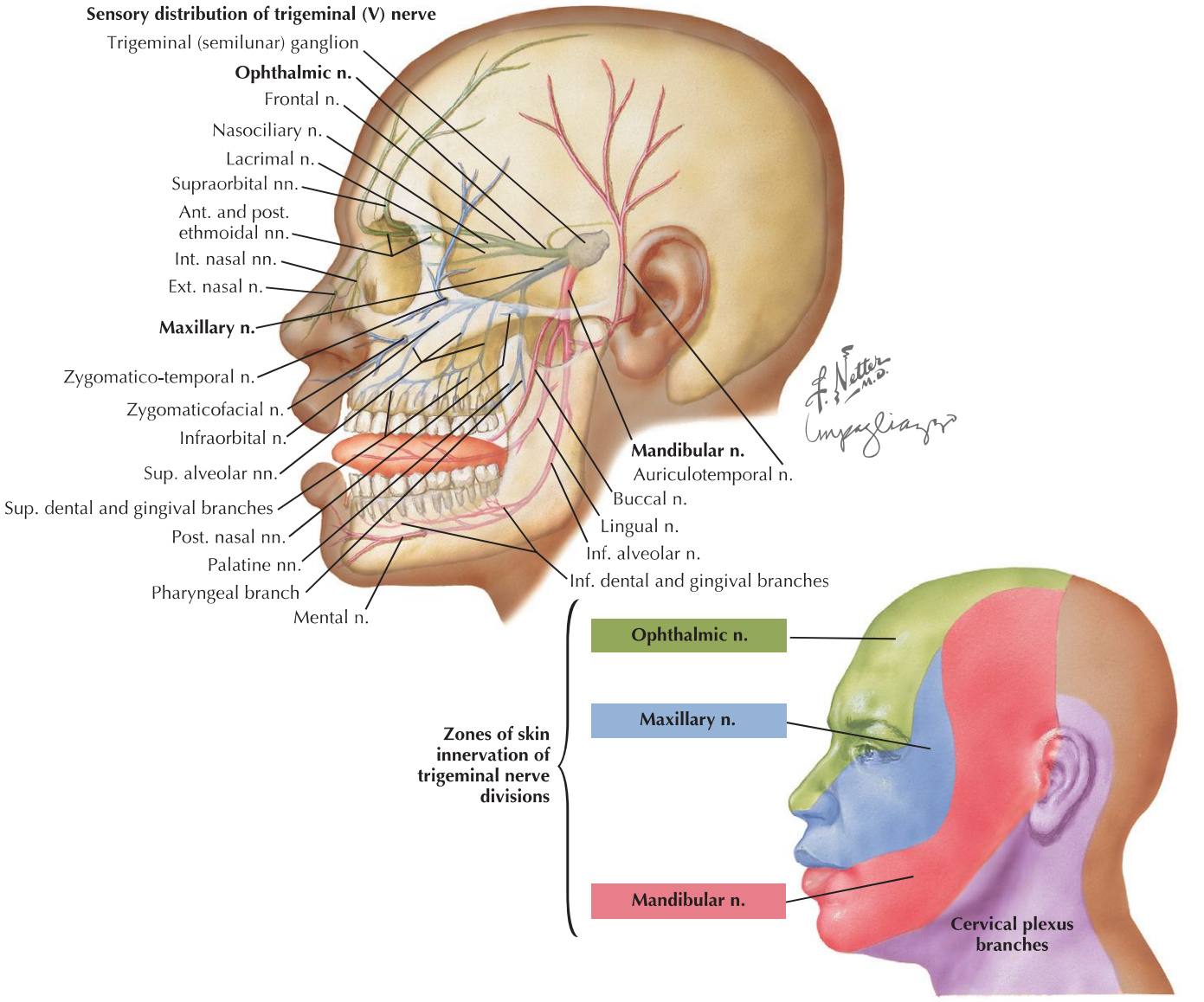


Figure 1-8 Trigeminal Nerve Neuralgia.

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 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

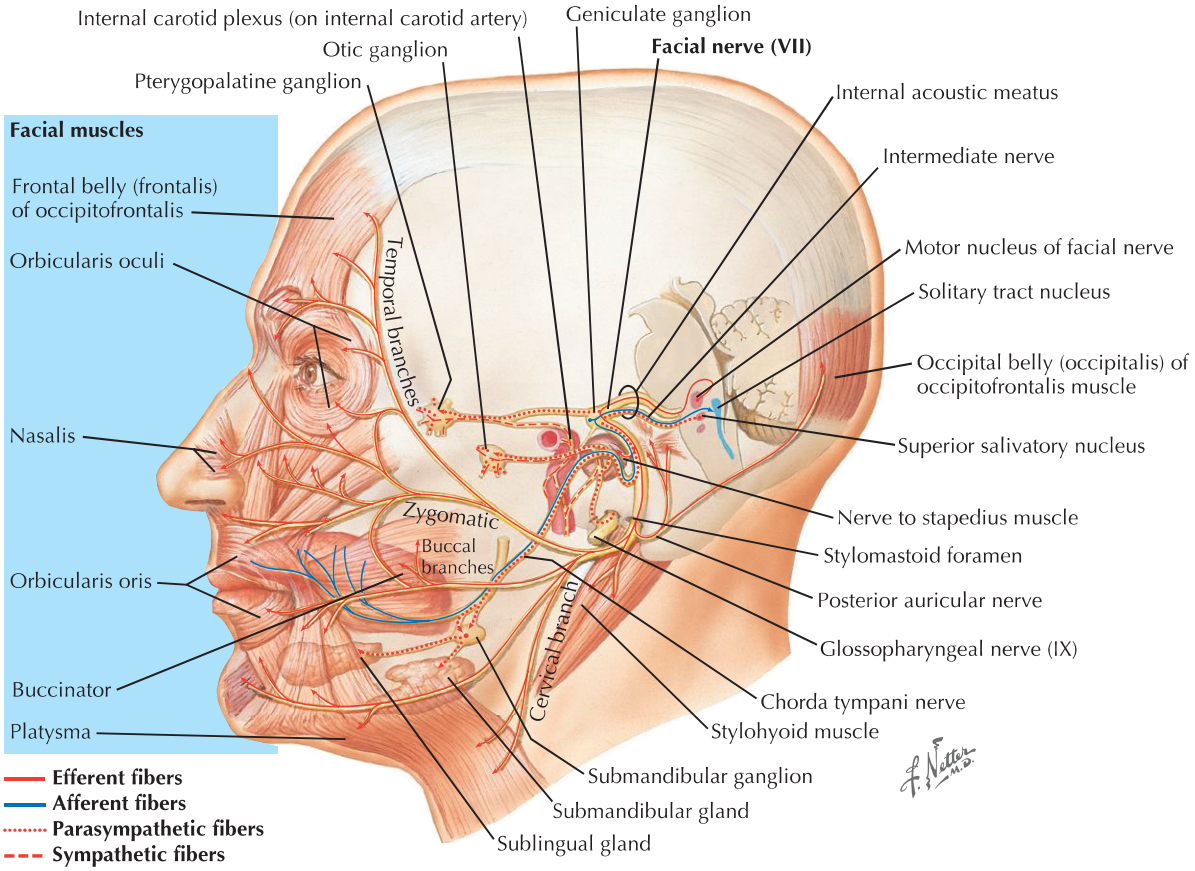


Figure 1-9 Facial Nerve With Its Muscle Innervation.

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 (Auditory [Cochlear] Nerve)

Many mornings some of us are blessed by a virtual ornithological symphony in our backyards. This always makes one pause and give thanks once again for this marvelous primary sensation. Here yet another cranial nerve, the cochlear, provides for the emotional highs that auditory sensations bring to the human brain. Whether it is the first cry of a newborn, the reassuring words of a loved one, or Beethoven's seventh symphony, this unique sensation of higher animal life is tracked through this one cranial nerve.

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. 1-11). 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, 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.

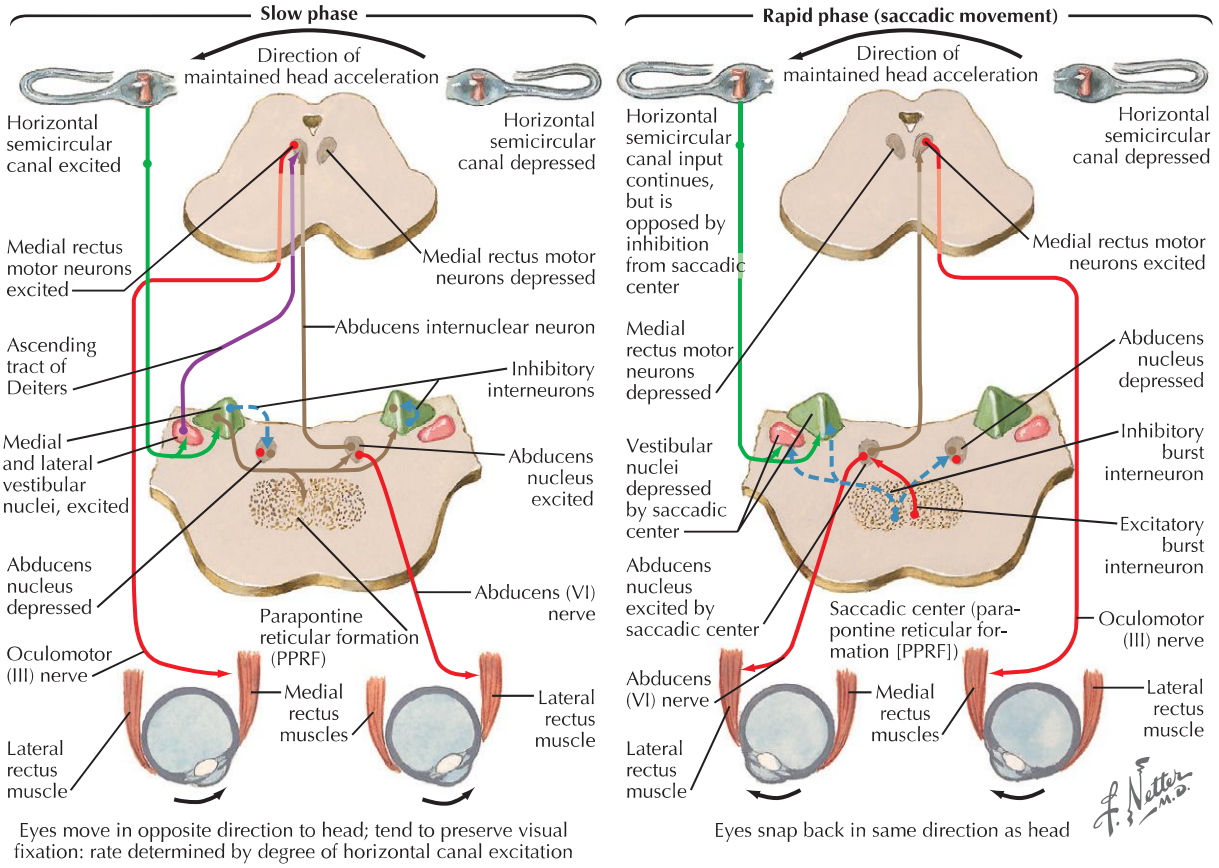


Figure 1-10 Vestibular Eighth Nerve Input to Horizontal Eye Movements and Nystagmus.

VESTIBULAR NERVE

The vestibular system can be tested indirectly by evaluating for nystagmus during testing of ocular movements or by positional techniques, such as the Barany maneuver, that induce nystagmus in cases of benign positional vertigo (BPV) where inner ear dysfunction is caused by otolith displacement into the semicircular canals (Fig. 1-12). Here the patient is seated on an examining table and the eyes are observed for the presence of spontaneous nystagmus. If none is present, the examiner rapidly lays the patient back down, with the head slightly extended and concomitantly turning the head laterally. If after a few seconds' delay, the patient develops the typical symptoms of vertigo with a characteristic delayed rotary, eventually fatiguing nystagmus, the study is positive.

Eye movements depend on two primary components, the induced voluntary frontal eye fields and the primary reflex-driven vestibular–ocular movement controlled by multiple connections (Fig. 1-10; see also Fig. 1-7). The ability to maintain conjugate eye movements and a visual perspective on the surrounding world is an important brainstem function. It requires inputs from receptors in muscles, joints, and the cupulae of the inner ear. Therefore, when the patient has dysfunction involving any portion of the vestibular-ocular or cerebellar axis, the maintenance of basic visual orientation is challenged. Nystagmus is a compensatory process that attempts to help maintain visual fixation.

Traditionally, when one describes nystagmus, the fast phase direction becomes the designated title (see Fig. 1-10). For example, left semicircular canal stimulation produces a slow nystagmus to the left, with a fast component to the right. As a result, the nystagmus is referred to as right beating nystagmus. Direct stimulation of the semicircular canals or its direct connections, that is, the vestibular nuclei, often induces a torsional nystagmus. This is described as clockwise or counterclockwise, according to the fast phase.

A few beats of horizontal nystagmus occurring with extreme horizontal gaze is normal in most individuals. The most common cause of bilateral horizontal nystagmus occurs secondary to toxic levels of alcohol ingestion or some medications, that is, phenytoin and barbiturates.

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. 1-13). Indirect mirror examination of the vocal cords may demonstrate paralysis of the ipsilateral cord. The traditional test for gag

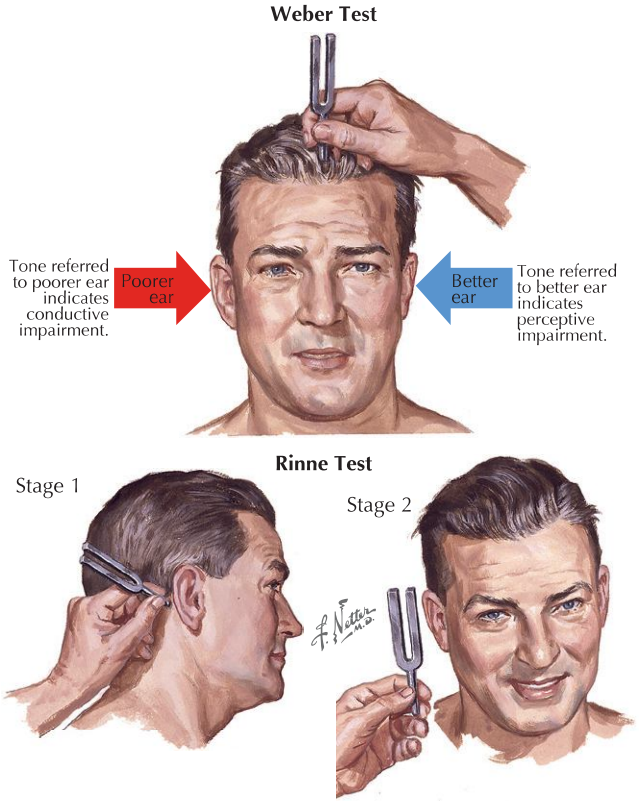


Figure 1-11 Auditory Nerve Testing: Weber and Rinne Testing.

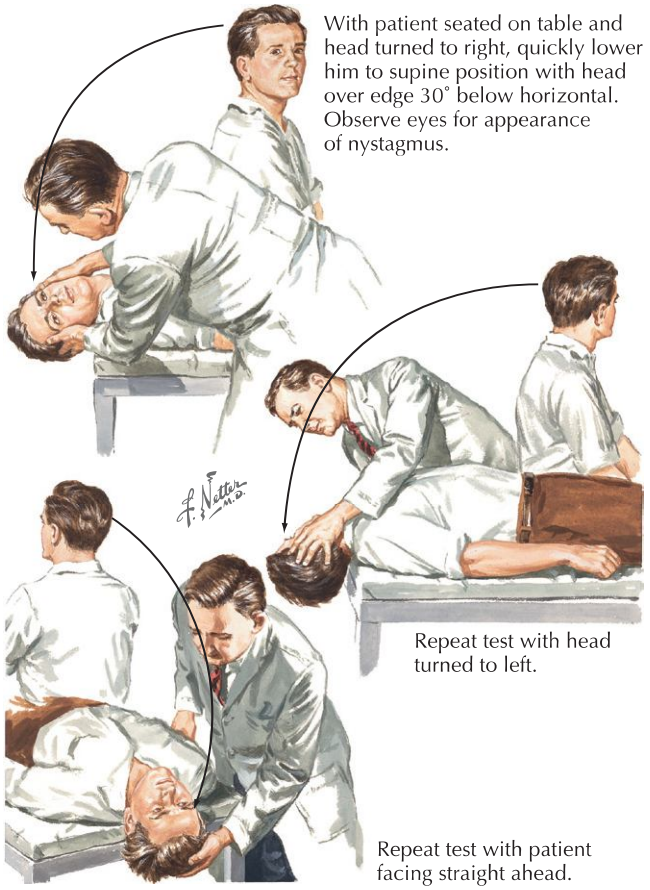


Figure 1-12 Test for Positional Vertigo.

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 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 (see Fig. 1-13). 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.

CRANIAL NEUROPATHIES AND SYSTEMIC DISEASE

When one evaluates a patient presenting with any cranial neuropathy, it is important to search for signs of other neurologic and systemic disorders. The patient with recently discovered anosmia may have early Parkinson disease. An acute painful, but pupil sparing, third nerve palsy may be a tip-off to the diagnosis of diabetes mellitus. When one meets an individual with unilateral or bilateral Bell palsies, Lyme disease, as well as sarcoidosis, always requires consideration in the differential diagnosis. When one evaluates patients having multiple cranial neuropathies, leptomeningeal infiltration from metastatic carcinoma or lymphoma, sarcoidosis or chronic infectious processes, such as tuberculosis, always require diagnostic consideration.

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

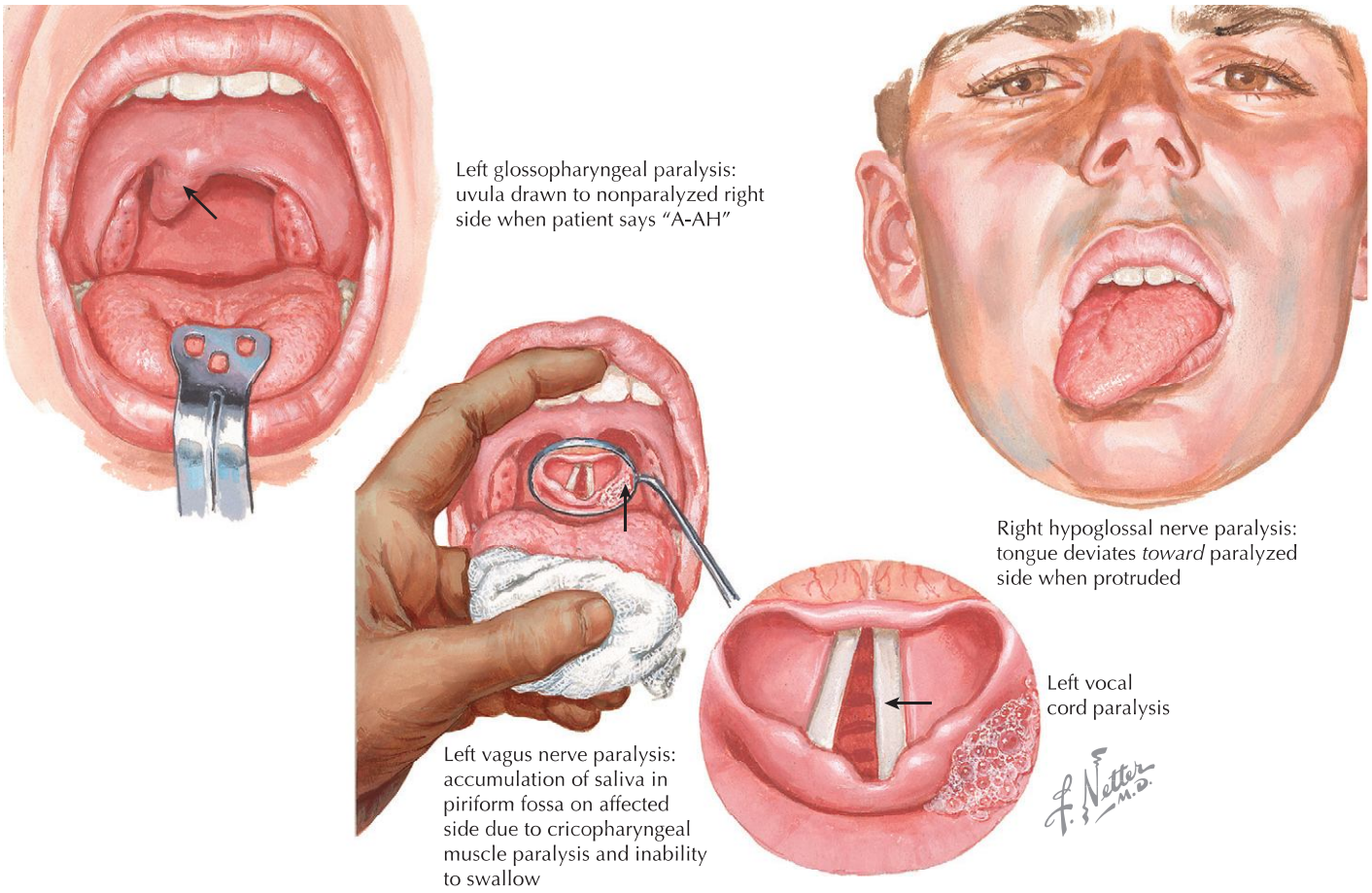


Figure 1-13 Uvula, Tongue, and Vocal Cord Weakness.

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

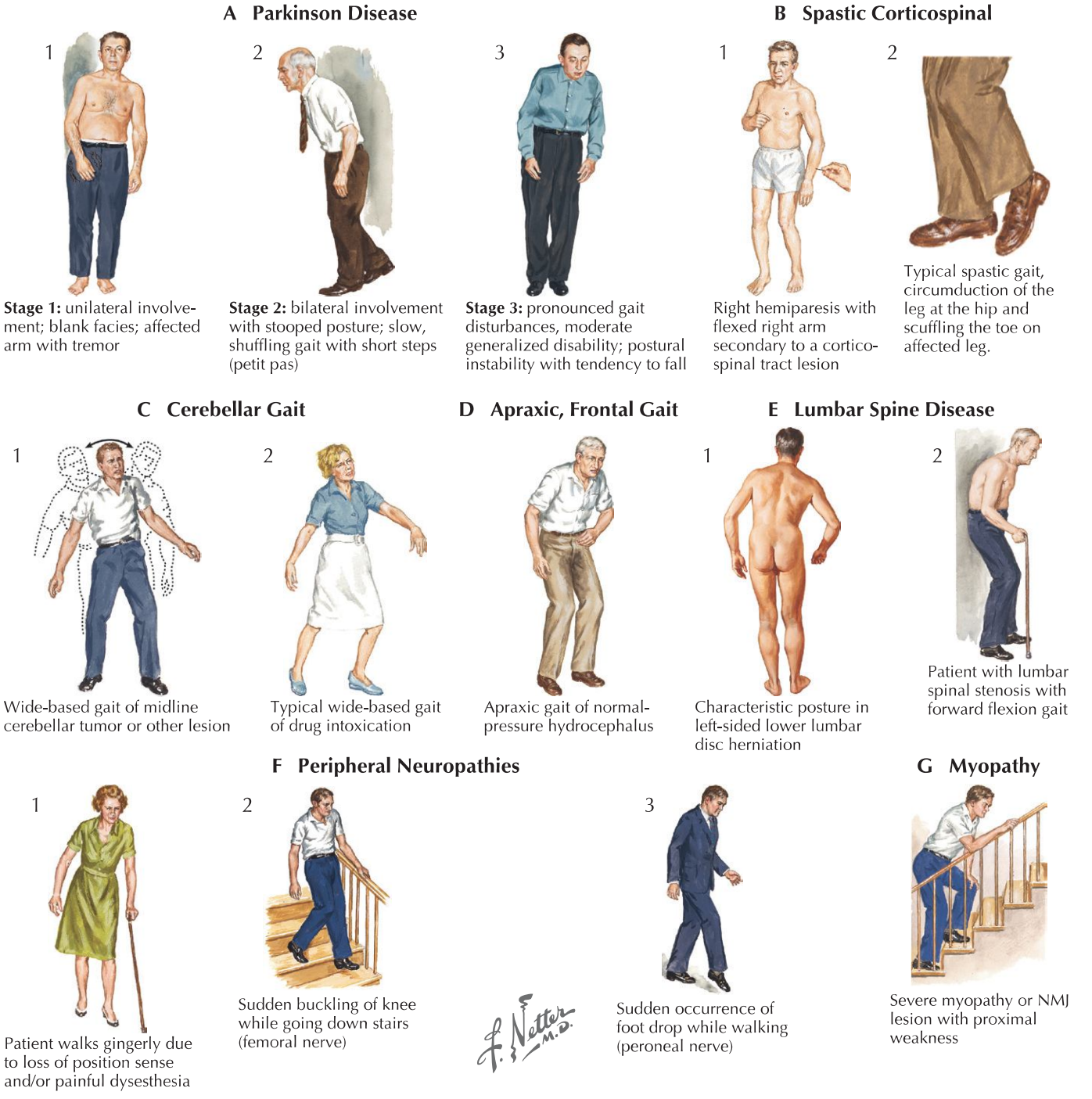


Figure 1-14 Gait Disorder Characteristics and Etiology.

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. 1-14).

Frontal lobe (Fig. 1-14,D) processes including 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. 1-15). Various *neurodegenerative* conditions, particularly those affecting the basal ganglia, such as *Parkinson disease* (Fig. 1-15,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,

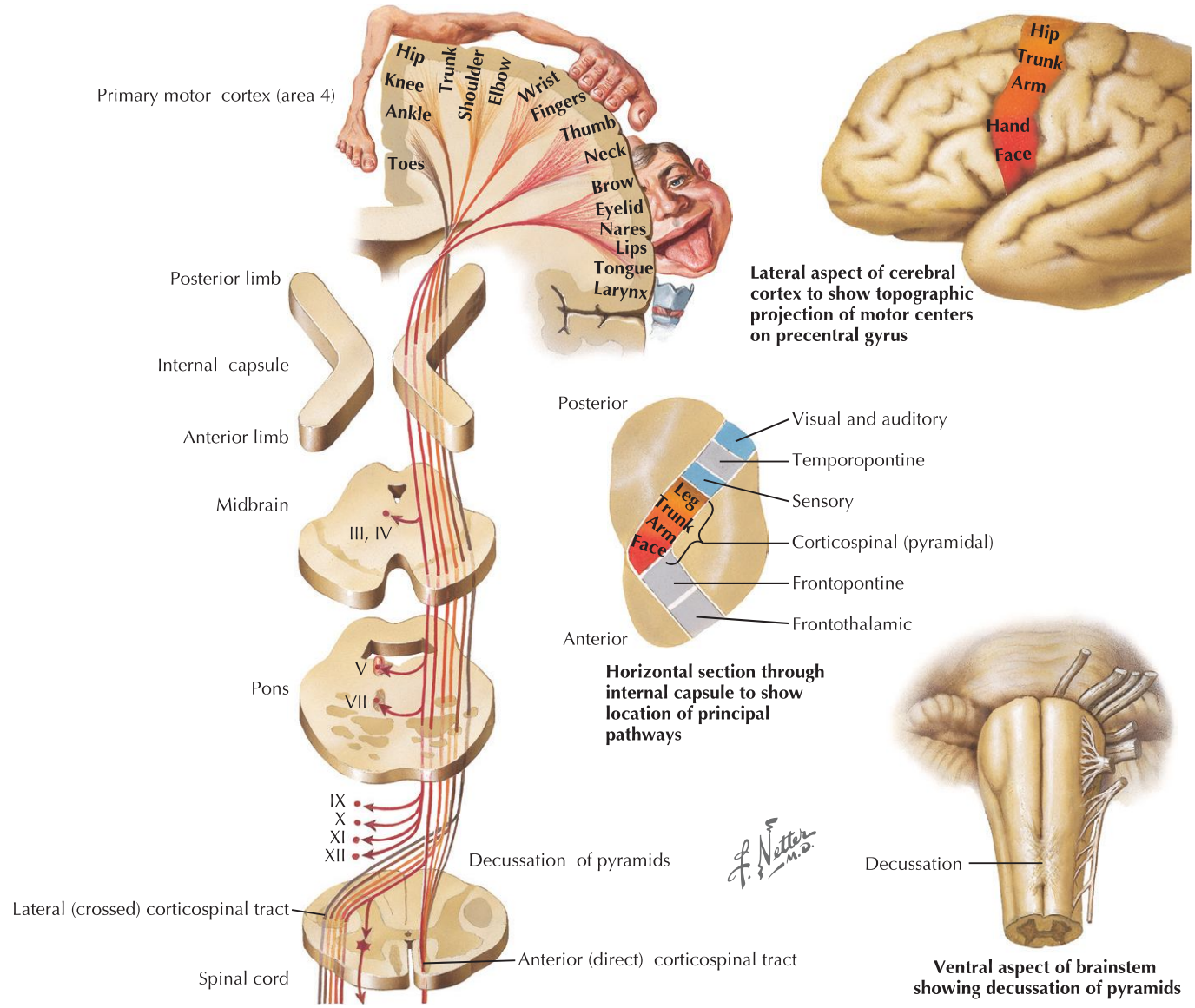


Figure 1-15 Pyramidal System, Corticospinal Tract. Gait Disorders Can Arise From Interruption of These Pathways at Any Level.

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 ataxia (Fig. 1-14,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. 1-14,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.

An often overlooked cause of gait difficulties is orthopedic and musculoskeletal problems. A perhaps simplistic perspective on the contribution of this system to gait is the analogy that the musculoskeletal system functions similar to an axle on a car, maintaining alignment and proper, symmetric rotation of the wheels. Our vertebral column is a sophisticated axle that with time loses some of its alignment. The attached muscles, to a misaligned chain of vertebrae, ultimately generates aberrant feedback loops to the spinal cord and the brain.

Many of our *senior citizens* gradually lose precise control of their gait, initially manifested by subtle changes on neurologic exam. Healthy older individuals often have limited ability to perform tandem gait. The very important message here is that this finding in isolation should not be considered abnormal per se among patients living into their eighth decade. Nevertheless, older patients become increasingly limited by a dwindling ability to walk independently.

Very often, in this setting there is not one specific mechanism either operative or identifiable. A number of patients have a multifaceted source related to the gradual *aging* (graying) of multiple neurologic systems. One source that always requires consideration is the possibility of *orthostatic hypotension*. Most commonly, this relates to medications; however, one of the neurodegenerative disorders, multiple system atrophy (see Chapter 34), may present in this fashion. Thus, it is important to carefully check blood pressures in the supine posture, when seated, then immediately on standing, and then every 30 seconds thereafter until the pressure is stabilized. A persistent drop in blood pressure of 20–30/15 mm Hg is usually regarded as significant in this setting.

It is important to ask about the circumstances accompanying the gait decline. Does the individual scuff a foot because of a spastic leg that interferes with a smooth alteration of individual legs? What settings lead to a fall? Does one catch one's toe on a rug as with subtle spasticity (Fig. 1-14,B1–2) or feel a leg give out going downstairs secondary to weakness of the quadriceps femoris muscle (Fig. 1-14,F2)? Having such information, the examiner can then easily try to reproduce the circumstances that lead to the falls.

Typically, gait function is tested under several conditions, including walking straight, walking at least 10 yards in open space, making turns, maneuvering through a tight corridor, attempting tandem gait, or in low light settings as well as on the stairs. The normal degree of foot separation (the base) is widened when proprioception or midline cerebellar vermis function is compromised. Occasionally, having the patient climb stairs reveals a subtle degree of iliopsoas weakness as found in various peripheral motor unit disorders (particularly myopathies) and, less commonly, neuromuscular junction or proximal peripheral neuropathies (Fig. 1-14,G). Finally, the appearance of spasticity may be enhanced by having the patient walk longer distances and even asking him or her to walk several blocks and return to the clinic. Rarely, this uncovers an unsuspected corticospinal tract lesion. Chapter 32 expands on the clinical evaluation of gait disorders.

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 VII.

MUSCLE STRENGTH EVALUATION

Weakness is one of the most common complaints of patients seeking neurologic care. The motor pathways encompass multiple anatomic areas within the CNS, including the cerebral cortex and important subcortical structures such as the basal ganglia, the brainstem, the cerebellum, the spinal cord, and the peripheral motor unit (Fig. 1-16). Although complaints of generalized weakness, fatigue, or both often are not caused by a specific neurologic condition, the possibility of multiple sclerosis in younger individuals and Parkinson disease in older patients always needs to be considered. When the patient is significantly overweight or has a neuromuscular disorder, sleep apnea needs consideration as a cause of fatigue or a feeling of "weakness." Peripheral motor unit disorders are important considerations for the differential diagnosis of a patient with generalized weakness. These include processes affecting the anterior horn cell (i.e., amyotrophic lateral sclerosis), peripheral nerve (i.e., Guillain–Barré syndrome or chronic inflammatory demyelinating disorders), neuromuscular junction (including Lambert–Eaton myasthenic syndrome [LEMS]), or muscle cells (various myopathies).

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.

Focal weakness often has a subtle character that frequently is not recognized by the patient as loss of motor strength. Dropping objects or clumsy handwriting may represent a single peripheral nerve lesion such as a radial neuropathy leading to a wrist drop. Tripping on rugs or steps may be the expression of a peroneal nerve lesion causing a foot drop (Fig. 1-14,F3). In

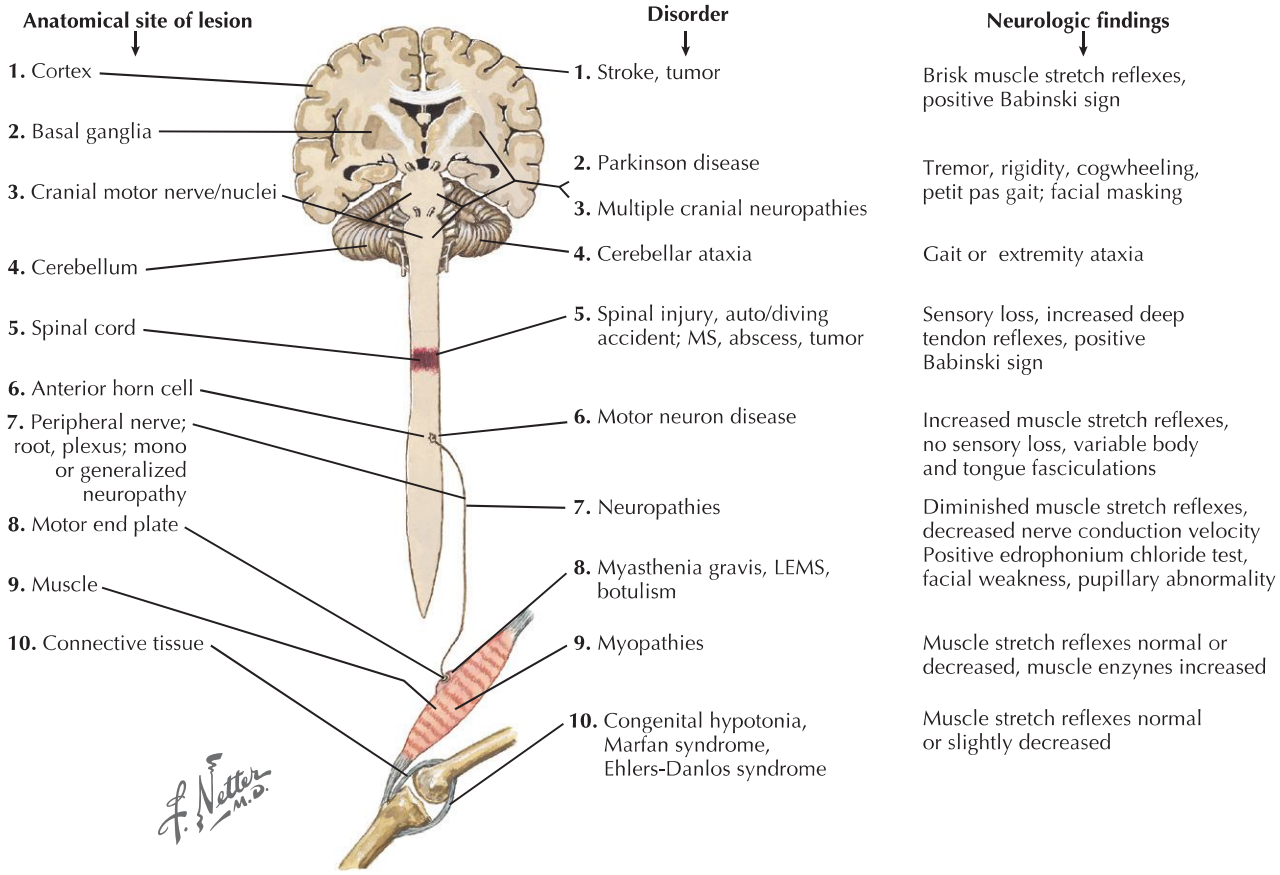


Figure 1-16 Primary Sites of Motor Disorders.

contrast, dramatic whole limb weakness is obvious and of greater patient concern, often leading to immediate medical attention as occurs with a stroke. Bilateral motor loss without cognitive or visual difficulties is most commonly due to lesions affecting the spinal cord or the peripheral nervous system and muscle.

When analyzing the complaint of weakness, the physician must consider the presence or absence of associated neurologic complaints or difficulties, such as language, speech, and visual changes; gait dysfunction; difficulty with rising from chairs and associated movements; and alteration in sensation. The neurologist testing for strength must search for evidence of atrophy and fasciculations, or spasticity. Equally important is the need to note the degree of patient effort and cooperation, as well as to consider associated problems that may compromise the testing, such as pain and skin or orthopedic lesions. Formal strength testing must be conducted in a systematic manner evaluating successive areas of the motor unit beginning at the brain and proceeding distally to the individual muscles per se (see Fig. 1-16). Here one places an initial focus on the major muscle groups, such as the flexors and extensors, to seek out any areas of weakness. More specific muscle testing is particularly useful when distinguishing between lesions of the nerve root, plexus, or mononeuropathies (Table 1-2).

When individual muscle testing does not demonstrate specific weakness, other techniques sometimes uncover more less-obvious functional loss. If the patient is instructed to extend the arms with the palms up and the eyes closed, subtle arm weakness may manifest as a pronating downward or lateral drift of the affected extremity. Similarly, moving the fingers as if playing piano or rapidly tapping may demonstrate a subtle in-coordination. Subtle proximal lower extremity weakness may not be appreciated with individual muscle testing. Watching the patient rise from a chair may demonstrate use of furniture arms to "push off" and is a good means to identify early proximal leg weakness. One particularly effective means to uncover proximal leg weakness is to observe the patient climb stairs or squat and attempt to rise without using their arms. Also asking the patient to walk on the heels or the tips of the toes is helpful in uncovering distal leg weakness.

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 1-3). 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

Table 1-2 Muscle Testing in a Routine Neurologic Examination

Muscle	Action	Nerve	Root
Infraspinatus	External rotation of arm	Suprascapular	C5
Biceps	Flexion of forearm	Musculocutaneous	C5-6
Deltoid	Abduction of arm	Axillary	C5
Triceps	Extension of forearm	Radial	C7
Extensor digitorum	Extension of fingers	Posterior interosseous of radial	C7
Flexor digitorum	Grip	Median	C7-8
APB* and opponens pollicis	Abducting thumb	Median	T1
Dorsal interossei	Spread fingers apart	Ulnar	C8
Iliopsoas	Flexion of thigh	Femoral	L2-3
Quadriceps	Extension of leg	Femoral	L3-4
Hamstring	Flexion of knee	Sciatic	S1
Gluteus medius	Abduction of thigh	Superior gluteal	L5
Gluteus maximus	Extension of thigh	Inferior gluteal	S1
Tibialis anterior	Dorsiflexion of foot	Deep peroneal	L5
Tibialis posterior	Inversion of foot	Tibial	L5
Peroneus longus	Eversion of foot	Superficial peroneal	L5, S1
Gastrocnemius	Plantar foot flexion	Tibial	S1-2

APB, Abductor pollicis brevis.

Table 1-3 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.*

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.

The examiner assesses the symmetry of function and coexisting changes in tone to formulate appropriate conclusions regarding the significance of subtle changes. The patient's degree of effort also needs to be assessed to distinguish organic disorders from feigned weakness in those with somatoform disorders or individuals with potential for secondary gain, as may occur with workers' compensation or other litigation. One of the most useful methods here is to ask the patient to very briefly put all of his or her effort into just one muscle. Most patients with various emotional nonorganic causes for "weakness" will not move the limb at all or produce very inconsistent (consistently inconsistent) efforts in contrast to a normal person's very firm, persistent motor output. The individual with nonorganic weakness classically "*gives way*," after just a very brief effort.

In the setting of possible "give way" weakness, one also needs to consider whether there is evidence of posttetanic facilitation, where the patient's initial effort suggests weakness but on a few more tries seemingly normal motor strength is achieved. This is the classic feature of a presynaptic defect in neuromuscular transmission as seen in Lambert-Eaton myasthenic syndrome (LEMS). Occasionally, one sees something like this with early Guillain–Barré syndrome or multiple sclerosis. This is an important and occasionally difficult differentiation. One must always listen carefully to the patient; when one is uncertain, the best study is sometime a careful reexamination of the patient. Today the findings of normal neuroimaging and neurophysiologic modalities are reassuring when considering diagnosis of a functional nonorganic disorder. It is very important to recognize that this is a diagnosis of exclusion. Furthermore, there is no urgency to make such a psychological-based diagnosis. Repeated, careful evaluations may uncover definitive findings leading to an organic diagnosis, or when normal, reassure both physician and patient alike that there is less concern about a serious illness.

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. 1-17) 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

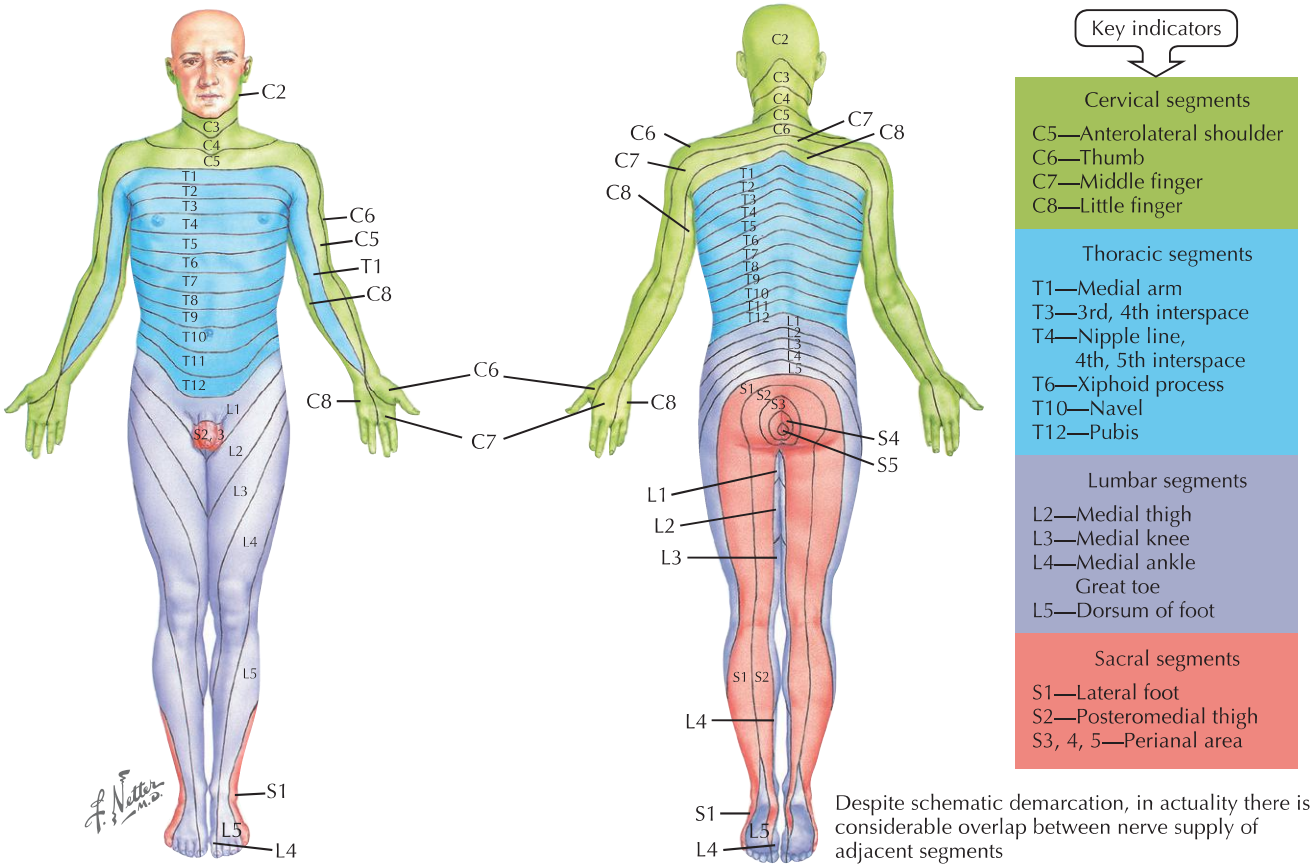


Figure 1-17 Dermatomal Levels.

polyneuropathies. These disorders may mimic a primary spinal cord lesion.

Patients with *painless asymmetric weakness* 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. 1-18).

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

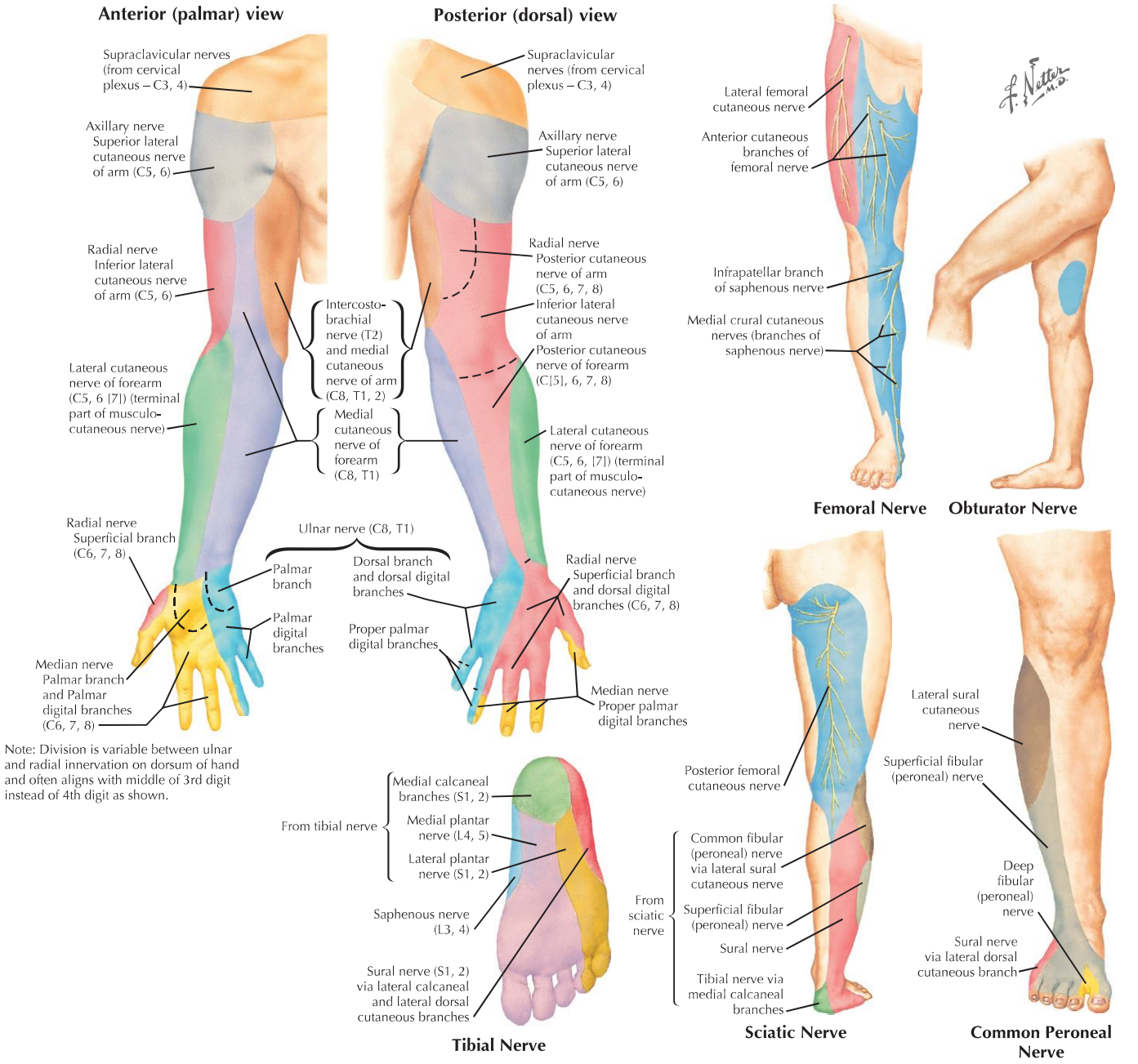


Figure 1-18 Cutaneous Innervations.

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

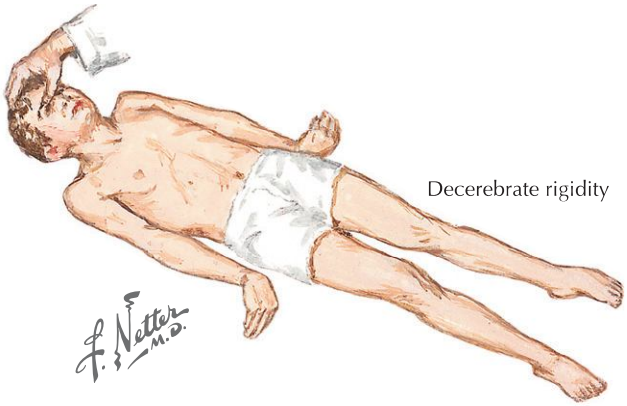


Figure 1-19 Motor Tone Abnormality.

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. 1-19) 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*.

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

Both Ia and Ib peripheral sensory nerve afferents join the posterior columns of the spinal cord, entering through the dorsal root ganglia. Their primary function is to convey information from touch and pressure receptors. Therefore, although the muscle spindles and Golgi tendon organs cannot be specifically tested, some of their spinal cord connections can be clinically evaluated by testing position and vibration sensory modalities. Additionally, the Ia and Ib afferents convey similar information to the cerebellum via the posterior spinocerebellar tract that travels into the cerebellum through the inferior cerebellar peduncle (Fig. 1-20). In isolation, it is difficult to assess the contribution of each tract specifically to motor control.

With simple passive stretching, such as occurs with tapping the patellar tendon at the knee, the intrafusal muscle spindle is activated, leading to a direct stimulus to the large alpha motor neurons. These in turn stimulate the extrafusal skeletal muscle fibers, leading to the clinically observed muscle contraction (Fig. 1-21). If the afferent sensory or efferent motor limb of this nerve supply is damaged, the muscle stretch reflex (MSR) is affected and may be diminished or lost, as occurs with many peripheral neuropathies. These reflexes are sometimes inappropriately referred to as deep tendon reflexes (DTRs) when in fact their physiologic basis primarily depends on the intrafusal muscle spindle fibers, not the Golgi tendon organs. MSR is a more accurate term.

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. 1-22). 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

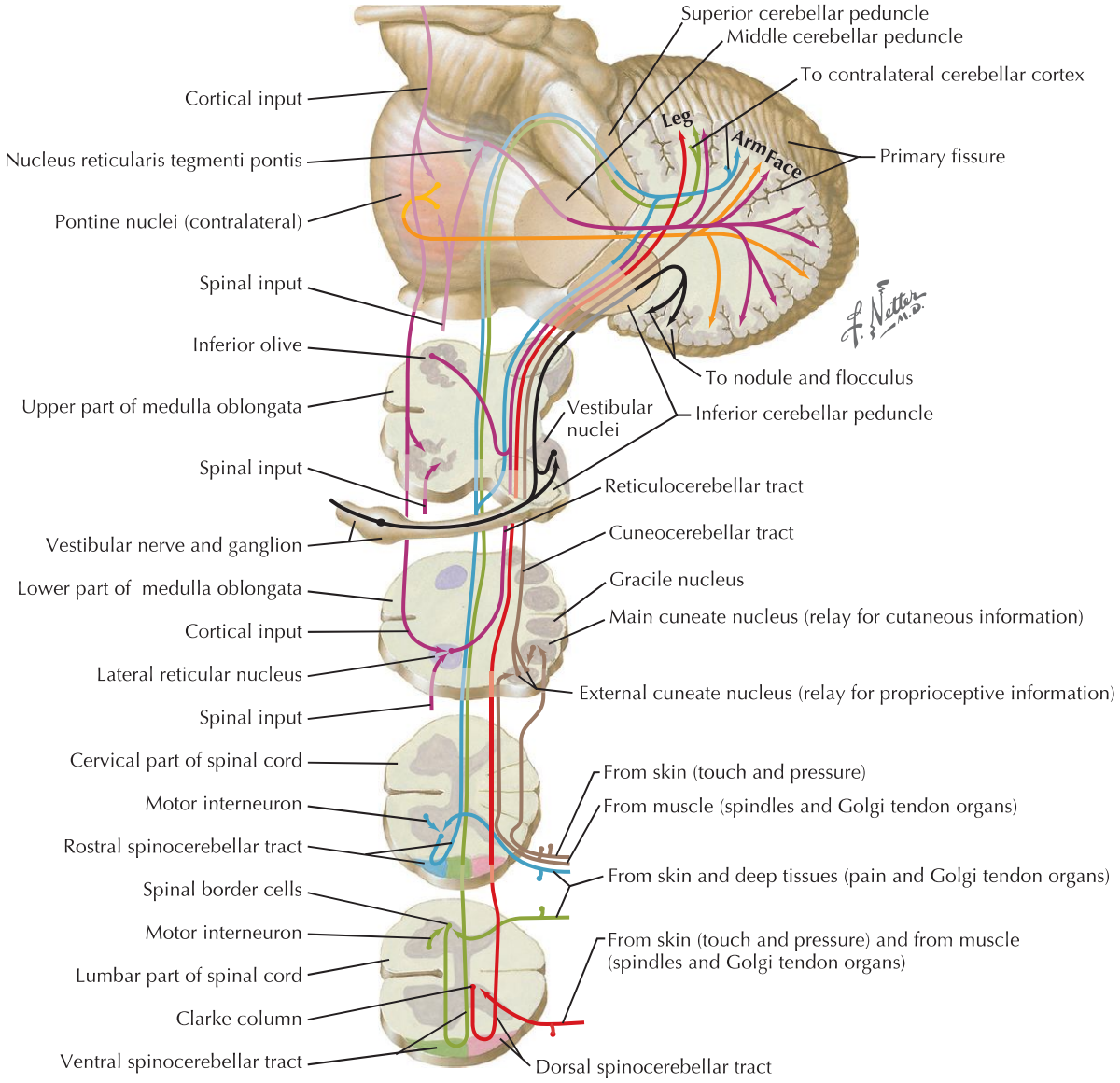


Figure 1-20 Cerebellar Afferent Pathways.

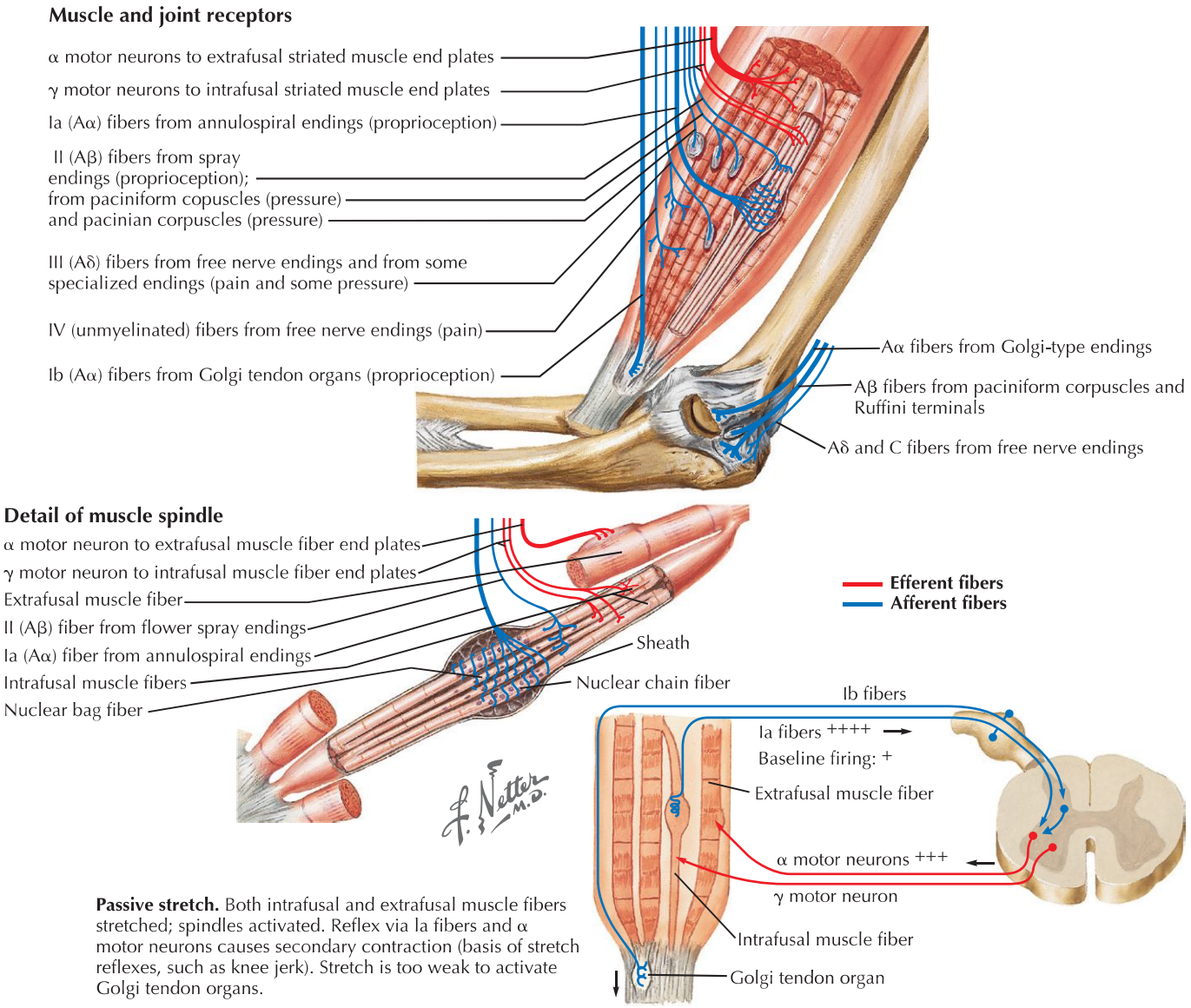
foot and entire set of toes dorsiflex. This is often overcome by separately pulling down on the middle toe while carefully stimulating the sole in traditional fashion.

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

A carefully designed sensory system evaluation is essential to define the presence or absence of normal sensation and, if abnormal, to define the specific anatomic patterns of loss for the affected modalities. Because part of the sensory examination is fairly subjective, the examiner should analyze the consistency of responses. Additionally, the relevance of sensory changes to the patient's complaints and other findings needs to be carefully evaluated. Initially, the examination needs to focus on defining the presence or loss of sensation. One must avoid having the patient be overly zealous trying to define the most subtle differences in sensory appreciation. This often leads to an exhausted patient and a frustrated clinician.

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



Passive stretch. Both intrafusal and extrafusal muscle fibers stretched; spindles activated. Reflex via Ia fibers and α motor neurons causes secondary contraction (basis of stretch reflexes, such as knee jerk). Stretch is too weak to activate Golgi tendon organs.

Figure 1-21 Muscle and Joint Receptors and Muscle Spindles.

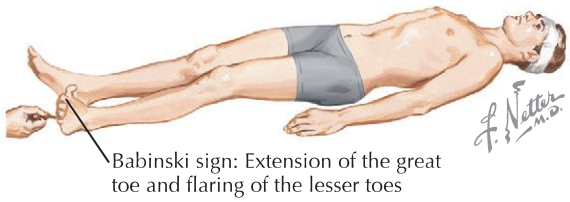


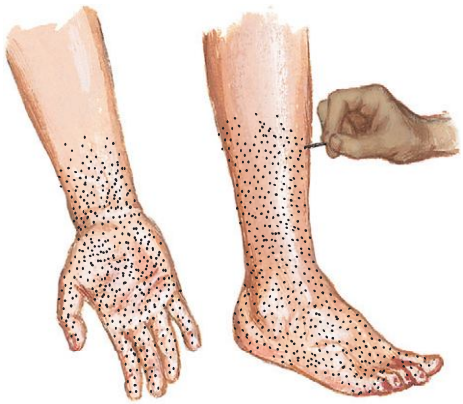
Figure 1-22 Elicitation of the Babinski Sign.

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.

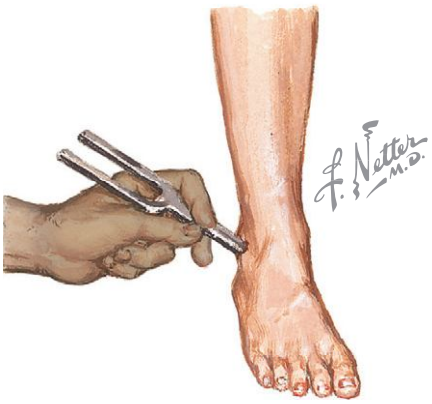
Fine tactile discrimination is evaluated by using a pair of calipers to check their ability to recognize whether one or two points are applied to the digit. *Vibratory sensation* depends on both deep afferent and cutaneous sensory modalities subserved by type A-alpha fibers. It is best tested by a 128-Hz 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. 1-23). Eventually, this loss will gradually spread proximally past the ankles and wrists into the legs and forearms but usually not above the



Graduated glove-and-stocking hypesthesia to pain and/or temperature



Impaired vibration sense

Figure 1-23 Documentation of Various Types of Sensory Modalities in a Peripheral Neuropathy.

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.

Individual mononeuropathies are typified by symptoms and findings specific to a single peripheral nerve (see Fig. 1-18). For example, the patient notes numbness in the thumb, index, middle fingers, and adjacent lateral aspect of the fourth finger if the median nerve is involved. In carpal tunnel syndrome with entrapment of the median nerve at the wrist, the examination results are often subtly abnormal, only with loss of fine discriminatory function with subtle diminution of two-point discrimination. Sometimes, one can employ a reflex hammer to percuss directly over the entrapment site. If there is a focal area of peripheral nerve injury, this action commonly elicits brief paresthesiae distal to the percussion site and within the specific distribution of the sensory fibers of that nerve, in this case the median. This maneuver is known as the Tinel sign; the name applies to instances wherein this simple provocative test defines the lesion site for any mononeuropathy.

Plexopathies are usually unilateral in distribution, affecting the brachial or lumbosacral groups of nerves. Typically, these are characterized by a combined motor sensory loss involving multiple peripheral nerves within the affected limbs. These lesions have a broader distribution of motor and sensory loss than do single nerve root or mononeuropathy lesions. Therefore, when a clinical examination demonstrates findings not exclusively defined by one specific peripheral nerve or nerve root, a plexus lesion is likely to be present.

Radiculopathies frequently are characterized by more subjective, often intermittent but sometimes persistent, symptoms confined to the dermatomal patterns of one specific nerve root (see Fig. 1-17). Pain is the most common symptom, starting in the neck, shoulder, and low back, often radiating down along the limb in a specific dermatomal distribution. The most common and classic example in the cervical region is at the C7 nerve root where there are paresthesiae primarily involving the index and middle fingers. Often there is a concomitant diminution in triceps muscle strength as well as loss of the triceps reflex. In the low back, the L5 nerve root is the classic example, with numbness in the first and second toes and the lateral calf and accompanying weakness of both the tibialis anterior and tibialis posterior muscles. However as the knee jerk relates to the L4 nerve root, and the ankle jerk to the S1 root, the examiner has to test a less commonly utilized reflex, namely the internal hamstring that has an L5 root innervation. When there is sensory involvement along the lateral aspect of the foot and the small toe with absence of the Achilles reflex, an S1 root lesion is most likely.

SPINAL CORD SYNDROMES

Transverse Complete

The site of a spinal cord lesion is defined by identifying the exact distribution of specific motor and sensory deficits of the various sensory modalities (Fig. 1-24). A *complete lesion* of the spinal cord leads to total loss of function distal to the site of the abnormality. A distinct level of sensory loss can be discerned with tests for loss of pain and/or temperature sensations, associated with loss of sweating below the lesion level. Concomitantly, all muscles subserved by anterior horn cells distal to the site of the lesion experience paralysis. Distinct partial cord syndromes are briefly described below and discussed further in Section VII (see also Fig. 44-13).

Brown–Séquard

A lesion in the anterior lateral aspect of the spinal cord causes contralateral loss of pain and temperature sensation. If the lesion is more extensive, leading to damage of the anterior and posterior aspects of the cord on one side, the *Brown–Séquard syndrome* occurs; it is characterized by contralateral loss of pain and temperature sensation, ipsilateral loss of position and vibration sensation, and ipsilateral upper motor neuron weakness.

Central Cord

Syringomyelia or a central hemorrhage leads to another anatomically specific lesion referred to as the *central cord* syndrome. The pathology occurs at the center of the cord, destroying fibers

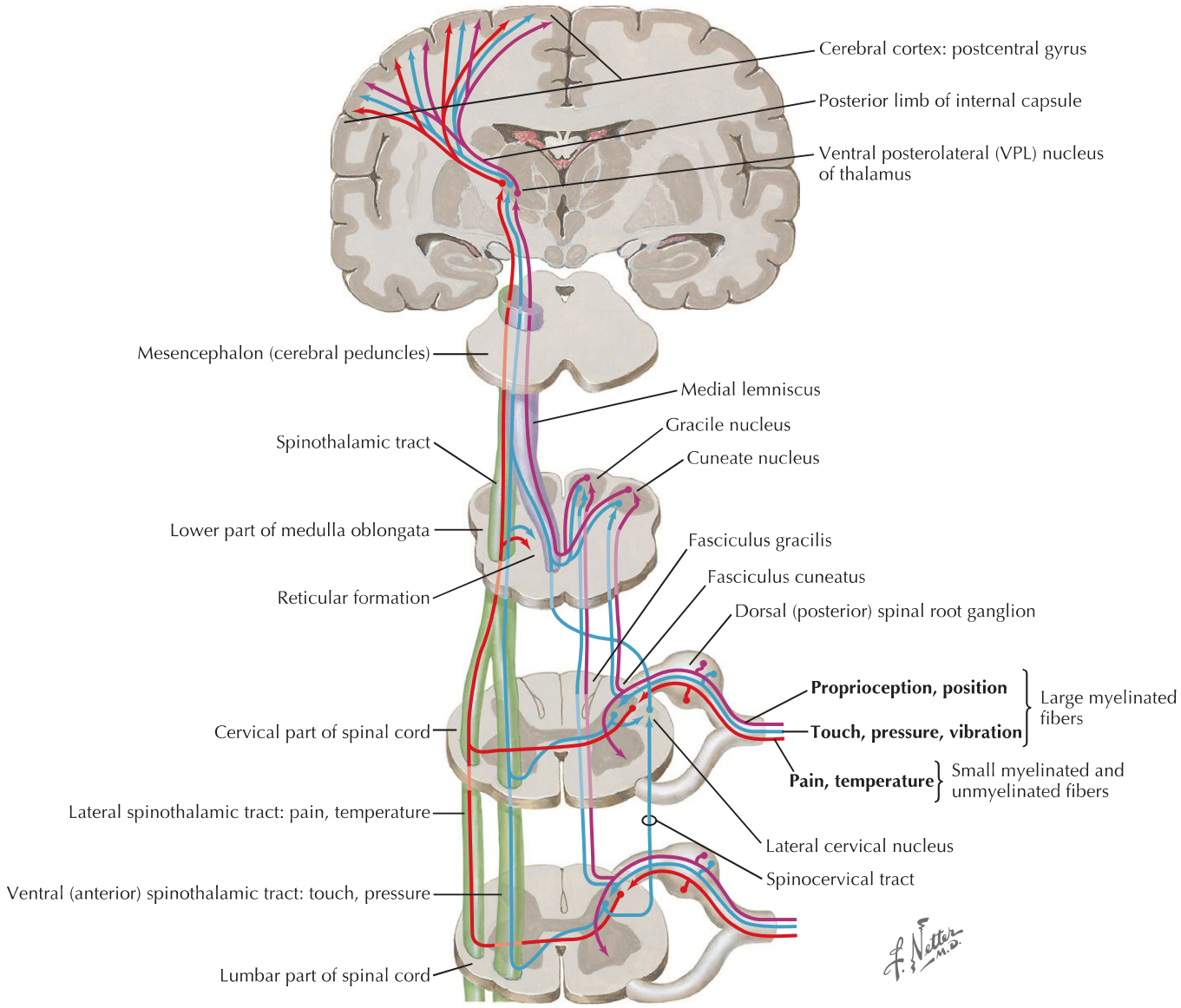


Figure 1-24 Somesthetic System: Body.

carrying pain and temperature sensation from both sides as they cross in the anterior commissure. Because the fibers carrying vibration and position sense do not initially cross at their entry level into the spinal cord, as do the spinothalamic tracts, these ascend within the posterior columns. Therefore a small centrally placed lesion within the cord spares those pathways. This leads to a dissociated sensory loss with isolated loss of pain and temperature sensation, usually in a "cape" distribution, while concomitantly, position sense is preserved.

Anterior Spinal Artery

A patient with an infarction within the territory of this essential artery presents another classic sensory picture. This is related to the inherent territory of supply of the anterior spinal artery; namely, it supplies the anterior two thirds of the cord. Here, there is bilateral damage to the spinothalamic and corticospinal tracts while the posterior columns are spared because of their dependence on the posterior spinal artery system. Although the patient is paralyzed and has total loss of pain and temperature sensation, position and often vibratory sensory modalities are preserved.

THALAMIC INVOLVEMENT

The ventral posterior lateral and ventral posterior medial thalamic nuclei are the two major sensory relay nuclei (Fig. 1-25). Lesions in these areas can cause loss of sensation to all modalities involving the entire contralateral half of the body. This most commonly occurs in patients with lacunar or hemorrhagic infarcts. Initially presenting with a relatively tolerable numbness, eventually the damage incurred from the stroke may produce an unpleasant, sometimes disabling, hyperpathic sensory alteration known as the thalamic pain syndrome. Rarely,

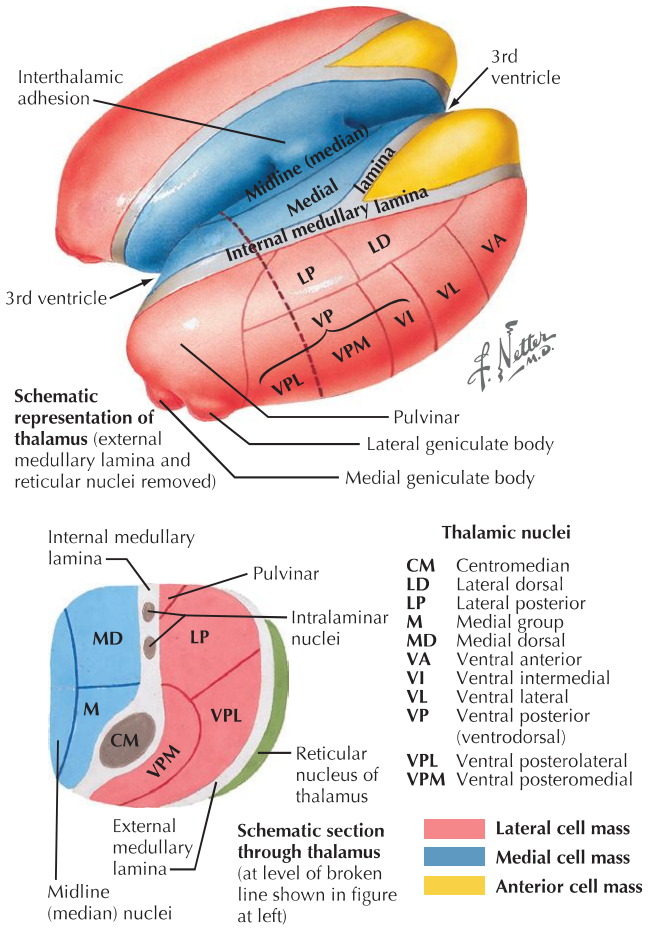


Figure 1-25 Thalamus and Its Multiple Nuclei.

this loss of sensation can lead to a limb deafferentiation sensory choreoathetosis. Lesions within the corona radiata, undercutting the parietal cortex, can cause similar, although often less extensive, findings.

CORTICAL SENSORY INVOLVEMENT

The parietal lobe receives topographically organized sensory inputs from the thalamic nuclei, brainstem, spinal cord, and peripheral nerves (see Fig. 1-24). An important function of the parietal lobe is the integration of this information with other sensory and motor information to formulate body awareness. In the purest form of cortical sensory dysfunctions, patients are unable to differentiate the location of their toes or fingers in space, make a distinction between one and two points, or employ stereognostic discrimination to allow differentiation of various objects placed in their hands, such as differing coin sizes. In addition, these individuals are unable to recognize numbers traced on the palm (graphesthesia).

Many other sensory abnormalities occur, including "neglect," wherein the patient with a right, nondominant, parietal lesion is unaware of paralysis or sensory loss of the contralateral limbs. These are especially obvious with double simultaneous stimulation (extinction). Here one or two sides of the body are variably stimulated, and the patient is asked to identify the stimulus location. Individuals with a more subtle parietal sensory loss cannot identify the contralateral stimulus when bilateral stimuli are applied. At the extreme a patient sustaining very large hemisphere and subcortical stroke presents with a complete loss of sensation of the contralateral body.

ADDITIONAL RESOURCES

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