Evaluation of the adult with acute weakness in the emergency department

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INTRODUCTION

Weakness is a common, nonspecific emergency department (ED) complaint that encompasses a broad differential diagnosis. Causes include neurologic ailments and a range of non-neurologic conditions. The diagnosis of potentially life-threatening neurologic and neuromuscular processes requires a systematic, anatomic approach based upon a careful history, physical examination, and in some cases, imaging studies.

In older adults, infection, cardiovascular disease, and dehydration must particularly be considered as possible causes of weakness. However, such conditions cause generalized malaise rather than true neuromuscular weakness and will not be discussed here, except to mention them as important considerations in the differential diagnosis.

The approach to the diagnosis and initial management of patients presenting to the ED with acute, nontraumatic neurologic and neuromuscular weakness will be reviewed here. Medical conditions characterized by general malaise or chronic weakness is discussed separately.

DIFFERENTIAL DIAGNOSIS OF ACUTE WEAKNESS

Although this topic reviews the approach to the patient with acute weakness from nontraumatic neurologic or neuromuscular disease, a broad differential diagnosis, including causes of generalized weakness (or malaise), is presented here to assist clinicians looking for additional information about these conditions.

Life-threatening central causes of unilateral weakness

- Ischemic stroke – Sudden loss of focal brain function is the core feature of the onset of ischemic stroke. This may manifest as acute, focal, unilateral weakness or paralysis in the face, upper extremity, or lower extremity, or as difficulty with coordination and gait. Other medical illness can mimic stroke (table 1), and symptoms of stroke can vary widely based upon the cause and the artery involved (table 2 and table 3). (See "Overview of the evaluation of stroke" and "Initial assessment and management of acute stroke".)

- Intracerebral hemorrhage – The neurologic symptoms and signs associated with intracerebral hemorrhage frequently worsen over minutes to hours, in contrast to ischemic stroke and subarachnoid hemorrhage. Headache, vomiting, seizures, and a decreased level of consciousness often occur. Unilateral weakness or paralysis may be present. (See "Spontaneous intracerebral hemorrhage: Pathogenesis, clinical features, and diagnosis".)

- Subarachnoid hemorrhage (SAH) – Sudden onset of severe headache is the most common presentation of SAH. This may be accompanied by loss of consciousness, seizure, nausea and vomiting, and meningismus. Lateralizing signs, such as unilateral weakness, are uncommon. (See "Aneurysmal subarachnoid hemorrhage: Clinical manifestations and diagnosis".)

Life-threatening and other serious causes of bilateral weakness

Brainstem stroke

- Brainstem stroke – Lesions in the brainstem may produce ipsilateral cranial nerve and contralateral body weakness. Clinical findings vary depending upon the areas involved but may include vertigo, depressed mental status, visual field deficits, oculomotor abnormalities, and bulbar findings. (See "Posterior circulation cerebrovascular syndromes".)

Spinal cord disease

- Spinal cord inflammation or compression – A spinal cord lesion may be suspected when there are bilateral motor and sensory signs or symptoms that do not involve the head.

Motor deficits involve weakness, muscle spasticity, and hyperreflexia (although hyporeflexia is often seen acutely); sensory findings involve a discrete level below which sensation is absent or reduced. Urinary bladder function may be affected resulting in either urinary retention or incontinence depending on the level of spinal cord injury. A wide range of pathologies can cause spinal cord disease, including trauma, infection (eg, epidural abscess), neoplasm, hemorrhage, inflammation (eg, transverse myelitis), and degenerative disorders. (See "Disorders affecting the spinal cord" and "Anatomy and localization of spinal cord disorders".)

Peripheral nerve disease

- Guillain-Barré syndrome (GBS) – The cardinal clinical features of GBS consist of progressive, fairly symmetric muscle weakness, accompanied by absent or depressed deep tendon reflexes [1,2]. Patients usually present a few days to a week after onset, with progressive weakness of the legs and arms (sometimes initially only in the legs). Patients frequently present to the ED with a complaint of numbness or paresthesias in the limbs [1,2]. Weakness can vary from mild difficulty with walking to nearly complete paralysis of all extremity, facial, respiratory, and bulbar muscles. Early diagnosis enables clinicians to initiate therapy as soon as possible to prevent further nerve damage [3]. (See "Guillain-Barré syndrome in adults: Pathogenesis, clinical features, and diagnosis".)

- Tick paralysis – Tick paralysis caused most commonly by Dermacentor ticks usually begins with paresthesias and a sense of fatigue and weakness. Fever is characteristically absent. Despite patients' reports of paresthesias, the sensory exam is typically normal. Most patients eventually develop an unsteady gait that progresses to an ascending complete paralysis. Deep tendon reflexes are characteristically absent. Respiratory paralysis and death can occur in severe cases. (See "Tick paralysis".)

Neuromuscular junction disease

- Myasthenia gravis (MG) – MG can produce weakness in any muscle group. Certain presentations are more common: ocular symptoms (eg, ptosis, diplopia) occur in 50 percent; bulbar symptoms (eg, dysarthria, dysphagia, fatigable chewing) occur in about 15 percent; and isolated limb weakness occurs in about 5 percent. Myasthenic crisis occurs when respiratory and/or bulbar muscle weakness produces acute respiratory distress. (See "Clinical manifestations of myasthenia gravis" and "Myasthenic crisis".)

- Organophosphate and carbamate poisoning – Acute toxicity from organophosphorus agents presents with manifestations of cholinergic excess. The dominant clinical features include bradycardia, miosis, lacrimation, salivation, bronchorrhea, bronchospasm,

urination, emesis, diarrhea, diaphoresis, and generalized weakness. (See "Organophosphate and carbamate poisoning".)

- Botulism – Patients with food-borne botulism may have a prodrome of vomiting, abdominal pain, diarrhea, and dry mouth. Symptoms of cranial nerve involvement then develop (eg, fixed pupillary dilation, diplopia, nystagmus, ptosis, dysphagia, dysarthria), followed by descending muscle weakness, which usually progresses from the trunk and upper extremities to the lower extremities. Smooth muscle paralysis leads to urinary retention; diaphragmatic paralysis can lead to respiratory distress requiring intubation. (See "Botulism".)

Muscle disease

- Alcoholic myopathy – This myopathy occurs in long-standing alcoholics, presents with muscle cramps, tenderness, and swelling, and is a major cause of nontraumatic rhabdomyolysis. (See "Drug-induced myopathies", section on 'Alcohol'.)

- Myositis – Both dermatomyositis and polymyositis usually present with symmetric proximal muscle weakness, which has often been worsening over several months. Muscle pain and tenderness is present in up to half of cases. (See "Clinical manifestations of dermatomyositis and polymyositis in adults".)

Life-threatening medical causes with focal findings — Metabolic conditions most often cause generalized weakness without focal findings. However, exceptions exist, most notably hypoglycemia and hypokalemic periodic paralysis.

- Hypoglycemia – Symptoms and signs of severe hypoglycemia are nonspecific and can include fatigue, dizziness, visual disturbances, drowsiness, dysarthria, and depressed mental status. Untreated, symptoms can progress to seizures or coma. (See "Hypoglycemia in adults without diabetes mellitus: Clinical manifestations, causes, and diagnosis".)

- Periodic paralysis – Severe electrolyte abnormalities can cause generalized or focal muscle weakness. Hypo- or hyperkalemia, hypo- or hypercalcemia, hypomagnesemia, or hypophosphatemia may be the cause, and are discussed separately:

  - Potassium disorders (see "Clinical manifestations and treatment of hypokalemia in adults" and "Myopathies of systemic disease", section on 'Hypokalemic myopathy' and "Clinical manifestations of hyperkalemia in adults" and "Hypokalemic periodic paralysis" and "Hyperkalemic periodic paralysis")

- Calcium disorders (see "Clinical manifestations of hypocalcemia" and "Clinical manifestations of hypercalcemia", section on 'Musculoskeletal')

- Magnesium and phosphate disorders (see "Hypomagnesemia: Clinical manifestations of magnesium depletion" and "Hypophosphatemia: Clinical manifestations of phosphate depletion")

Muscle weakness usually does not occur at potassium concentrations above 2.5 meq/L if hypokalemia develops slowly, but significant weakness may occur with sudden decreases. Weakness usually begins with the lower extremities, progresses to the trunk and upper extremities, and can worsen to the point of paralysis. Patients with renal dysfunction can develop hyperkalemia that manifests initially as weakness and may deteriorate into a life-threatening arrhythmia if untreated.

Endocrine abnormalities, such as thyrotoxicosis, may also cause periodic paralysis. (See "Thyrotoxic periodic paralysis".)

Life-threatening causes of generalized weakness — A brief list of several important causes of generalized weakness is provided here to assist with the differential diagnosis and to allow ready access to additional information.

- Sepsis – Among other symptoms, malaise and generalized weakness may be a manifestation of sepsis. (See "Evaluation and management of suspected sepsis and septic shock in adults".)

- Acute coronary syndrome (ACS) – A significant percentage of older adult patients with ACS complain only of generalized weakness. Diabetics and women may also complain of weakness rather than chest discomfort when experiencing an ACS. (See "Overview of the acute management of ST-elevation myocardial infarction" and "Overview of the acute management of non-ST-elevation acute coronary syndromes" and "Initial evaluation and management of suspected acute coronary syndrome (myocardial infarction, unstable angina) in the emergency department".)

- Carbon monoxide (CO) poisoning – The clinical findings of CO poisoning are highly variable and largely nonspecific. Moderately or mildly CO-intoxicated patients often present with constitutional symptoms, including headache (the most common presenting symptom), malaise, nausea, and dizziness, and may be misdiagnosed with acute viral syndromes. (See "Carbon monoxide poisoning".)

- Adrenal insufficiency – Patients with chronic, progressive adrenal insufficiency most often develop the following symptoms: chronic malaise, lassitude, fatigue that is worsened by exertion and improved with bed rest, weakness that is generalized (ie, not limited to particular muscle groups), anorexia, and weight loss. (See "Clinical manifestations of adrenal insufficiency in adults".)

Other neurologic causes of acute weakness

- Multiple sclerosis (MS) – There are no clinical findings that are unique to MS, but some are highly characteristic of the disease (table 4) while others are more common during initial presentation (table 5). The typical patient presents as a young adult with two or more clinically distinct episodes of CNS dysfunction with at least partial resolution.

- Hemiplegic migraine – This uncommon migraine variant is characterized by unilateral motor and sensory symptoms. (See "Pathophysiology, clinical manifestations, and diagnosis of migraine in adults".)

- Postictal (Todd's) paralysis – Generalized or complex partial seizures may be followed by a focal motor deficit that can persist for hours, but typically resolves within 30 to 60 minutes, and is often related to a structural abnormality of the brain. (See "Overview of the clinical features and diagnosis of brain tumors in adults".)

Other medical causes of generalized weakness

- Hypothyroidism – Many of the manifestations of hypothyroidism reflect one of two changes induced by lack of thyroid hormone (table 6). The first is a generalized slowing of metabolic processes. This can lead to fatigue, slow movement and slow speech, cold intolerance, constipation, delayed relaxation of deep tendon reflexes, and bradycardia. The second is accumulation of matrix glycosaminoglycans in tissues, which can lead to coarse hair and skin, puffy facies, enlargement of the tongue, and hoarseness. (See "Clinical manifestations of hypothyroidism".)

- Infection – Occult infection, particularly in older adult patients, may manifest as malaise and generalized weakness. (See "Medical care in skilled nursing facilities (SNFs) in the United States".)

- Anemia – The differential diagnosis of anemia is broad, but the presentation often includes progressive weakness and pallor. (See "Diagnostic approach to anemia in adults".)

- Dehydration or hypovolemia – Hypovolemia, often as a result of vomiting, diarrhea, or diuretics, can cause generalized weakness. (See "Etiology, clinical manifestations, and

diagnosis of volume depletion in adults".)

- Presyncope – Patients with acute episodes of weakness may describe a sensation of "nearly fainting." Determining the presence of a cardiac cause is of particular importance in the emergency department. (See "Approach to the patient with dizziness" and "Approach to the adult patient with syncope in the emergency department".)

- Medications – Many medications, in isolation or combination, and illicit drugs can cause weakness. Common culprits include beta blockers, diuretics, laxatives, chemotherapeutic agents, isoniazid, opioids, and alcohol. Some agents may have direct toxic effects on muscle, including glucocorticoids, statins, antimalarial drugs, antipsychotic drugs, colchicine, antiretrovirals, alcohol, and cocaine. (See "Drug-induced myopathies".)

- Rheumatologic disease – Weakness is a common feature of rheumatologic disease. As examples, fatigue is the most common complaint in patients with systemic lupus erythematosus, occurring in over 80 percent of patients, and a common complaint among those with rheumatoid arthritis. Polymyalgia rheumatica, which can be associated with the vision-threatening condition of temporal arteritis, can present with generalized weakness and myalgias. (See "Overview of the systemic and nonarticular manifestations of rheumatoid arthritis" and "Clinical manifestations and diagnosis of polymyalgia rheumatica" and "Clinical manifestations and diagnosis of systemic lupus erythematosus in adults", section on 'Mucocutaneous involvement'.)

Older adult patients — Occult infection, metabolic disorders, stroke, and medication related problems are common causes of weakness in older adults. While some medications can cause myopathy directly (eg, glucocorticoids, statins), a number of medications can create problems in older patients through adverse reactions or drug interactions (see the drug interactions program). Twenty percent of patients over 60 had symptoms attributable to prescription medications in one study of patients with a chief complaint of weakness or dizziness [4]. (See "Drug prescribing for older adults" and "Falls in older persons: Risk factors and patient evaluation" and "Frailty" and "Approach to abnormal gait in adults".)

HISTORY

Defining weakness — The initial challenge is to determine exactly what a patient means when they complain of "weakness." True weakness is the inability to perform a desired movement with normal force because of a reduction in muscle strength. While this may be due to a primary neuromuscular process, more often the complaint represents malaise associated with a

medical illness. Such an illness may include virtually the entire spectrum of medical pathology, including such serious conditions as sepsis, acute coronary syndrome, heart failure, dehydration, severe hypokalemia, adrenal insufficiency, or hypothyroidism. (See 'Differential diagnosis of acute weakness' above.)

Other important definitions include paresis, which indicates partial or complete paralysis, and plegia, which indicates total loss of muscle contraction.

Approach to the history — When a neurologic or neuromuscular problem is suspected, an algorithmic approach based upon a thorough history and examination facilitates neuroanatomic localization of the pathology (algorithm 1). Exclusion of potentially life-threatening and other serious illness at each neuroanatomic site is an important part of this approach (table 7). (See 'Assessment of life threatening illness' below.)

The history should include a description of the distribution of weakness (eg, bilateral hands) and its manifestations (eg, difficulty with fine motor tasks), the period over which symptoms developed, and clinical features associated with the weakness (eg, aphasia, diplopia). The important initial tasks are to determine whether the weakness is unilateral or bilateral and whether signs associated with central neurologic involvement are present.

Unilateral weakness — Important questions to consider when evaluating a patient with unilateral weakness include the following:

- Are cortical signs present, such as aphasia, neglect, agnosia, or apraxia?
- Is the face involved (eg, facial droop)?
- Is there a myotomal pattern to the distribution of weakness?
- Is the description of weakness consistent with a particular peripheral nerve?

Generally, unilateral facial weakness implies a lesion above the spinal cord, either in the brainstem or cortex. Other associated cortical features can help localize pathology in one of the two cerebral hemispheres (algorithm 1).

In the case of isolated extremity weakness without cortical signs, the clinician uses knowledge of common radicular and peripheral nerve entrapment syndromes to identify the problem. A localized process (eg, weakness and paresthesias limited to one or two fingers) suggests peripheral nerve entrapment, although distinguishing spinal from peripheral nerve entrapment can be challenging in some instances. Familiarity with cervical and lumbosacral dermatomes and myotomes helps with recognition of spinal nerve root compression, and may help to differentiate this from peripheral pathology (table 8 and figure 1 and table 9 and figure 2 and table 10).

Bilateral weakness — Important questions to consider when evaluating a patient with bilateral weakness include the following:

- Is mental status depressed?
- Which limbs are involved?
- Is there sensory involvement? If so, is a sensory level deficit suggested?
- Is there bladder involvement?
- Does weakness primarily involve proximal or distal muscles?
- Are there bulbar signs (involving tongue, jaw, face, or larynx)?
- Does the degree of weakness fluctuate?

A central nervous system (CNS) lesion causing bilateral weakness is usually accompanied by diminished mental status, unless the lesion lies in the spinal cord. The presence of bladder dysfunction or a sensory deficit below a discrete dermatomal level suggests a myelopathy.

The proximal motor weakness typically found early in the course of a myopathy is suggested by difficulty walking up stairs or getting up from a chair, if the lower limbs are involved, or difficulty with overhead activities (eg, combing hair), if the upper extremities are involved.

Symptoms associated with disease at the neuromuscular junction include visual symptoms, particularly ptosis and diplopia, and bulbar signs. Bulbar muscle weakness may manifest as nasal speech, coughing, or lingual dysarthria. A fatiguing pattern to weakness, suggested by worsening with repeated activity (eg, chewing), suggests myasthenia gravis.

Periodic paralysis due to imbalances in potassium regulation (or other electrolyte abnormalities) is suggested by acute attacks of weakness lasting a few hours to a couple of days that spontaneously resolve.

PHYSICAL EXAMINATION

A neurologic examination, with careful attention to strength testing and deep tendon reflexes, is necessary in all patients who complain primarily of acute weakness. A more detailed examination must be performed in patients with symptoms suggestive of a central process (eg, dysarthria, oculomotor or visual dysfunction, ataxia). The detailed examination should include careful assessment of cranial nerve and motor function. Performance of the neurologic examination is discussed in detail separately. (See "The detailed neurologic examination in adults".)

Motor neuron findings — Recognition and distinction between upper motor neuron (UMN) or lower motor neuron (LMN) signs is important when determining the nature of a weakness syndrome (table 11 and figure 3). How to make this distinction between UMN and LMN lesions and localize spinal cord lesions are discussed in detail separately, but a few highlights are described below. (See "Anatomy and localization of spinal cord disorders".)

Signs of UMN disease include spasticity (ie, increased muscle tone), hyperreflexia, and an extensor plantar response (Babinski reflex). These findings are absent in pure peripheral disease. However, hyporeflexia and flaccid paralysis may occur with acute central lesions, with hyperreflexia and spasticity developing later. An example of this clinical scenario is an emergency department (ED) patient who presents with acute spinal cord transection from trauma. (See "Acute traumatic spinal cord injury".)

Signs of LMN disease include decreased muscle tone and hyporeflexia. The Babinski reflex is absent. If weakness is bilateral and signs of LMN disease are present, the major disorders to consider are neuropathies, myopathies, and disorders of the neuromuscular junction (NMJ). The distinguishing features of these processes are listed in the table (table 12).

Neuropathies tend to involve distal muscle groups, while myopathies more often involve proximal muscles. Reflexes are decreased in neuropathies, but may be present, decreased, or absent with myopathies. Myopathies may have associated myalgias, but sensory symptoms are generally absent. The most distinguishing feature of neuromuscular junction disease is early involvement of the bulbar musculature (ie, tongue, jaw, face, and larynx).

Strength testing — Strength testing is central to the examination of the weak patient. Clinicians should employ the standardized motor grading scale agreed upon by the Medical Research Council (table 13) [5]. Using historical features as a guide, first try to identify if there is a pattern to the weakness. Hemiparesis suggests a hemispheric lesion; paraparesis suggests a spinal cord lesion. Peripheral nerve disease, notably Guillain-Barré syndrome, can also cause paraparesis.

If the history suggests a proximal pattern of weakness, such as difficulty walking up stairs or difficulty standing from a chair, seek to distinguish proximal from distal muscle weakness. Proximal weakness suggests a myopathic process rather than a neuropathy. Strength testing of specific muscle groups is useful when assessing isolated extremity weakness (figure 4 and figure 5). If pathology at the NMJ is suspected, oculomotor and bulbar testing are critical. Assess extraocular movements, eyelid strength, masseter muscle strength, facial expression, and palatal movement.

Reflex testing — Reflex testing aids in the diagnosis and the localization of nerve lesions (table 8 and table 14). Tendon jerks are graded using a standard scale, with zero representing absence and four representing hyperactivity with clonus (table 15).

Lesions in one cerebral hemisphere generally result in hyperreflexia of the affected side due to disruption of the UMN. Interruption of the corticospinal tracts bilaterally generally causes symmetrical hyperreflexia and extensor plantar responses (positive Babinski). A spinal cord lesion is the most common cause of such findings, but in rare instances, bilateral cerebral hemisphere damage or brainstem disease may be present.

Nerve root compression most often causes unilateral absence of a specific lower extremity reflex. As an example, unilateral loss of the ankle jerk suggests a lesion at the S1 nerve root.

Fatigability — Fatigability describes normal strength during initial testing that decreases significantly as testing continues. It is characteristic of myasthenia gravis. Maneuvers, such as the ice test or Tensilon test can be performed to diagnose NMJ pathology. (See "Diagnosis of myasthenia gravis", section on 'Clinical testing'.)

Sensation testing — While the presence, pattern, or absence of sensory symptoms and signs can be helpful in confirming the neuroanatomic level producing weakness, the subjective nature of sensory findings makes interpretation challenging. Classically, a cortical lesion causes relatively mild hemisensory loss, which affects touch and proprioception more than pain. The patient may simply describe this as their arm or leg "feeling funny." A spinal cord lesion often affects sensation bilaterally, with the upper level of the sensory loss defining the lesion level.

In contrast, cervical central cord lesions most commonly produce a "cape" sensory loss over the shoulders affecting pain and temperature sensation, but spares vibration and proprioception (so-called "dissociated" sensory loss). Lesions of the conus medullaris or cauda equina produce loss of sensation in the perineum. When the lateral half of the spinal cord is damaged (Brown-Sequard syndrome) proprioception and vibration are lost ipsilateral to the lesion, while pain and temperature sensation are lost on the contralateral side.

In the case of more localized sensory loss in a limb, familiarity with dermatomes and peripheral nerve sensory distributions helps to differentiate a spinal root lesion from that of a peripheral nerve (figure 1 and figure 2 and figure 6). The absence of sensory symptoms in such cases suggests the weakness stems from a myopathy or NMJ disease.

ANCILLARY STUDIES

General approach — The extent of diagnostic testing performed on a weak patient in the emergency department (ED) varies depending upon the differential diagnosis and the potential pace of deterioration of any neuromuscular process being considered. As examples, a potentially septic patient with no obvious source of infection undergoes extensive diagnostic testing, a patient with a possible acute ischemic stroke requires an emergent head computed tomography (CT), and most patients with a presumed peripheral nerve entrapment require no diagnostic testing in the ED.

For many patients with generalized weakness, it is reasonable to obtain a hemoglobin concentration and electrolyte panel testing. A hemoglobin measurement identifies anemia while a standard electrolyte panel enables assessment of serum glucose, potassium, sodium, and renal function. An electrocardiogram (ECG) is a reasonable screening tool in older adult patients with generalized weakness and in patients with an abnormal potassium or calcium level. Beyond these baseline tests, a focused differential diagnosis determines further testing.

Pulmonary function testing — Pulse oximetry provides a reasonable index of oxygenation but is insensitive for identifying patients with significant neuromuscular disease. Arterial blood gas measurement may not be helpful in establishing the likelihood of imminent respiratory failure, because the pCO_2 can rise precipitously as diaphragmatic and intercostal muscle weakness progresses with neuromuscular disease.

Patients with such diseases commonly lose tidal volume before upper airway weakness develops, resulting in an increased respiratory rate to maintain minute ventilation. In such cases, the pCO_2 may remain normal or low until the tidal volume becomes dangerously low.

For this reason, selected pulmonary function tests (PFTs) are needed to determine the degree of respiratory compromise and to aid in the decision to intubate patients with suspected neuromuscular disease, such as Guillain-Barré syndrome (GBS) or myasthenia gravis (MG). As always, patients determined to be in respiratory distress or in danger of imminent respiratory compromise are intubated based upon clinical assessment. (See "The decision to intubate".)

The specific tests that can be used to assess respiratory function are described separately. (See "Respiratory muscle weakness due to neuromuscular disease: Management", section on 'Chronic ventilatory support'.)

Neuroradiography — Neuroradiological diagnostic testing may be useful depending upon where a lesion is suspected. A head CT without contrast is performed when a cortical process must be assessed. CT identifies acute hemorrhage, mass lesions, and cerebral edema in the great majority of cases, but may appear normal in the early stages of ischemic stroke. A head CT with contrast is obtained if an intracranial tumor or certain infections (eg, toxoplasmic

encephalitis) are suspected. (See "Neuroimaging of acute stroke" and "Overview of the clinical features and diagnosis of brain tumors in adults" and "Approach to the patient with HIV and central nervous system lesions".)

Brainstem and cerebellar lesions are best seen with magnetic resonance imaging (MRI), since this portion of the brain is poorly visualized on CT due to bone artifact from the base of the skull.

Spinal MRI is the imaging modality of choice if a nerve root or spinal cord process is suspected. CT scanning with myelography can be useful for diagnosing many myelopathic processes if MRI is unavailable or there is a contraindication to its use. (See "Clinical features and diagnosis of cervical radiculopathy".)

Chest radiography — A plain chest radiograph may be helpful if a malignancy, or related complication, is suspected. Such circumstances might include looking for a Pancoast tumor in a patient with a brachial plexopathy or a small-cell lung cancer in a patient with Lambert-Eaton syndrome. (See "Approach to diagnosis and evaluation of acute decompensated heart failure in adults" and "Superior pulmonary sulcus (Pancoast) tumors" and "Lambert-Eaton myasthenic syndrome: Clinical features and diagnosis".)

Cerebral spinal fluid (CSF) analysis — CSF analysis is indicated when Guillain-Barré syndrome (GBS), myelitis, or demyelinating peripheral neuropathy is suspected. In GBS, the CSF often reveals an elevated protein with a normal white blood cell count. However, detection of the elevated protein concentration is highly dependent on the timing of the lumbar puncture. (See "Guillain-Barré syndrome in adults: Pathogenesis, clinical features, and diagnosis", section on 'Cerebrospinal fluid analysis'.)

With a myelitis, CSF analysis is normal in approximately half of patients. In the other half, it may reveal an elevated protein or moderate lymphocytosis, but the glucose concentration remains normal. A process such as an inflammatory demyelinating peripheral neuropathy generally demonstrates elevated CSF protein. A lymphocytic pleocytosis is often present if disease is related to HIV. (See "Transverse myelitis: Etiology, clinical features, and diagnosis" and "Epidemiology, clinical manifestations, diagnosis, and treatment of HIV-associated distal symmetric polyneuropathy (HIV-DSPN)" and "Chronic inflammatory demyelinating polyneuropathy: Etiology, clinical features, and diagnosis", section on 'Lumbar puncture'.)

Edrophonium test — A more invasive and potentially hazardous test that may aid in the diagnosis of myasthenia gravis when available is the patient's response to the short-acting anticholinesterase agent edrophonium. Edrophonium blocks acetylcholine esterase, allowing sufficient acetylcholine to remain within the synaptic cleft to stimulate the decreased number of

postsynaptic binding sites, thereby improving symptoms. Edrophonium is not available in the United States, Canada, the United Kingdom, and many other countries. Performance of the test is discussed separately. (See "Diagnosis of myasthenia gravis", section on 'Pharmacologic testing'.)

False-positive responses to edrophonium chloride have been reported in Amyotrophic lateral sclerosis, Lambert-Eaton myasthenic syndrome, GBS, wound botulism, cavernous sinus lesions, polymyositis, and alcoholic myositis.

Other serologic testing — Serum creatine phosphokinase (CPK) is a sensitive marker for muscle damage and may be a useful screening test when an acute myopathy is considered.

ASSESSMENT OF LIFE THREATENING ILLNESS

The immediate life threats from acute neuromuscular weakness include inability to protect or maintain the airway, respiratory failure from thoracic and diaphragmatic muscle weakness, and circulatory collapse from autonomic instability.

Airway and breathing — The emergency clinician's first task is to identify patients at risk for acute respiratory failure. Signs of respiratory distress in patients with neuromuscular weakness can include [6]:

- Rapid, shallow breathing
- Poor respiratory effort; use of accessory muscles
- Difficulty swallowing; inability to handle secretions
- Inability to lift head off bed
- Weak, ineffective cough
- Weak or muffled voice
- Depressed mental status

Often, the first and most apparent sign is tachypnea. Patients with progressive generalized neuromuscular weakness commonly begin to lose tidal volume before upper airway weakness develops, resulting in an increased respiratory rate to maintain minute ventilation [7]. (See "Respiratory muscle weakness due to neuromuscular disease: Clinical manifestations and evaluation".)

The decision to secure the airway of patients with respiratory difficulty is made primarily on clinical grounds. However, measurement of simple bedside pulmonary tests, end-tidal carbon dioxide ($EtCO_2$), and arterial oxygen saturation (SpO_2) may provide insight into the patient's

respiratory status. Note that patients may be able to maintain CO_2 levels in the normal or even low range despite dangerously low tidal volumes. (See 'Pulmonary function testing' above.)

Airway assessment and management are discussed in detail separately. (See "The decision to intubate" and "Overview of advanced airway management in adults for emergency medicine and critical care".)

Airway management — When performing rapid sequence intubation (RSI), it is important in some settings to avoid depolarizing neuromuscular blocking agents (eg, succinylcholine) and instead to use a nondepolarizing neuromuscular blocking agent (eg, rocuronium) [8]. This avoids the risk of hyperkalemic respiratory arrest. (See "Neuromuscular blocking agents (NMBAs) for rapid sequence intubation in adults for emergency medicine and critical care".)

Evidence suggests that neurologic injury markedly exaggerates the potassium release associated with succinylcholine administration [9]. A conservative estimate is that the initial threat of such increased potassium release develops three days after complete denervation or seven days after partial denervation.

The risk of a hyperkalemic response is greatest among particular patients with neuromuscular weakness, including the following:

- Denervating injuries (eg, stroke, spinal cord injury) of greater than three days duration
- Denervating diseases (eg, Guillain-Barré syndrome with symptoms over three days, multiple sclerosis, amyotrophic lateral sclerosis, transverse myelitis with symptoms over three days)
- Inherited myopathies (eg, Duchenne's muscular dystrophy)
- Prolonged immobilization

Succinylcholine is safe in myasthenia gravis, but increased doses are needed. (See "Neuromuscular blocking agents (NMBAs) for rapid sequence intubation in adults for emergency medicine and critical care", section on 'Clinical use'.)

Circulation — Autonomic dysfunction accompanies some polyneuropathies and disorders of the neuromuscular junction, and can also occur in patients with generalized weakness due to systemic infection or acute coronary syndrome. Autonomic instability in the setting of neurologic weakness typically manifests initially as a hypersympathetic state, heralded by sinus tachycardia [7]. Thereafter, fluctuation in the heart rate and blood pressure occur. Treatment is largely supportive. Rarely, bradycardia can occur, which may require temporary pacing.

Critical diagnoses — With severely ill patients, the clinician must simultaneously assess the patient for life-threatening signs, begin resuscitative measures as indicated, develop a preliminary differential diagnosis, and initiate an appropriate work-up. An overview of the differential diagnosis for acute weakness is provided. (See 'Differential diagnosis of acute weakness' above.)

Important **neurologic** diagnoses to consider in the weak patient with respiratory distress or hemodynamic instability include:

- Ischemic or hemorrhagic stroke (see "Overview of the evaluation of stroke")
- CNS infection (see "Clinical features and diagnosis of acute bacterial meningitis in adults")
- Spinal cord injury (including compression) or inflammation (see "Disorders affecting the spinal cord")
- Guillain-Barré syndrome (see "Guillain-Barré syndrome in adults: Pathogenesis, clinical features, and diagnosis")
- Myasthenic crisis (see "Myasthenic crisis")

Important causes of **generalized weakness** to consider in the patient with respiratory distress or hemodynamic instability include:

- Sepsis (see "Evaluation and management of suspected sepsis and septic shock in adults")
- Acute cardiovascular disease (eg, coronary syndrome, decompensated heart failure)
- Intoxication or poisoning (see "General approach to drug poisoning in adults")
- Severe electrolyte abnormalities (eg, hypokalemia, hyperkalemia)
- Endocrine crisis (eg, diabetic ketoacidosis, hypoadrenal (Addisonian) crisis, myxedema coma, thyrotoxicosis)

DIAGNOSIS

Algorithmic approach — Once life-threatening problems have been addressed or ruled out, the clinician approaches the patient with objective weakness in a systematic manner, as outlined in the accompanying algorithm (algorithm 1). The first important step in this approach is to determine whether the weakness is unilateral (asymmetric) or bilateral (symmetric), and to look closely for signs of central neurologic involvement.

In the assessment of both unilateral and bilateral weakness, it is helpful to begin cephalad and centrally and then progress caudad and peripherally. This approach provides a reliable framework for neuroanatomic localization and accurate diagnosis.

If unilateral weakness is identified, look carefully for signs suggestive of cortical, subcortical (lacunar), or brainstem lesions. If these are absent, a peripheral process (radiculopathy, plexopathy, or peripheral nerve injury) most likely accounts for the patient's symptoms. Key questions for assessing unilateral weakness include:

- Are cortical signs present (eg, aphasia, neglect, agnosia, apraxia)? If so, pathology lies in the cerebral cortex.
- Is the face involved (eg, facial droop)? Unilateral facial weakness suggests a lesion above the spinal cord, either in the brainstem or cortex (or with Bell's palsy a peripheral nerve).
- Is there a myotomal pattern to the distribution of weakness? In such cases, familiarity with important cervical and lumbosacral myotomes helps to localize the lesion (figure 4 and figure 5 and table 8 and table 9 and table 14).
- Is the description of weakness consistent with a particular peripheral nerve? (See "Overview of upper extremity peripheral nerve syndromes" and "Overview of lower extremity peripheral nerve syndromes".)

If bilateral weakness is identified, consider the patient's mental status and look carefully for signs of upper or lower motor neuron lesions and associated abnormalities. The constellation of examination findings should allow approximate identification of the site of the lesion and determination of the need for imaging studies, specialist consultation, and treatment. Key questions for assessing bilateral weakness include:

- Is the patient's mental status depressed? Central nervous system (CNS) pathology that causes bilateral weakness is usually accompanied by diminished mental status, unless it is found in the spinal cord.
- Which limbs are involved? Lesions involving both the upper and lower extremities are located more proximally in the spinal cord (figure 1)
- Is there sensory involvement? A sensory deficit demarcated by a specific dermatome suggests spinal cord pathology (figure 1 and figure 7 and figure 2), while more diffuse sensory findings such as paresthesias may be an early sign of a polyneuropathy. (See "Guillain-Barré syndrome in adults: Pathogenesis, clinical features, and diagnosis".)
- Is there bladder involvement? Bladder dysfunction suggests a myelopathy.
- Does weakness primarily involve proximal or distal muscles? Involvement primarily of proximal muscles suggests a myopathy, while involvement of distal muscles suggests a

polyneuropathy.

- Is there eye muscle or “bulbar” weakness (involving the tongue, jaw, face, or larynx)? Diseases affecting the neuromuscular junction frequently present with ptosis, diplopia, or bulbar weakness. (See "Diagnosis of myasthenia gravis" and "Botulism".)

- Does the degree of weakness fluctuate? A fatiguing pattern to the weakness, suggested by worsening with repeated activity such as chewing or maintaining upward gaze, suggests a process involving the neuromuscular junction, such as myasthenia gravis; acute attacks of weakness lasting a few hours and then spontaneously resolving suggest periodic paralysis. (See "Diagnosis of myasthenia gravis" and "Botulism" and "Hypokalemic periodic paralysis" and "Hyperkalemic periodic paralysis".)

Unilateral weakness

Cortical findings — In a patient complaining of unilateral weakness, first inquire whether the limbs and lower face on the same side are involved. If so, a lesion exists in the contralateral cerebral hemisphere.

To further pinpoint the lesion's location, look closely for cortical signs, such as an aphasia, hemineglect, gaze preference, visual deficits, or apraxia. Aphasia usually corresponds to a left hemispheric stroke, since the left cerebral hemisphere controls language function in the majority of both right-handed and left-handed individuals. A nonfluent (Broca's) aphasia often accompanies a right hemiplegia of cortical origin, since the motor cortex is in close proximity to Broca's area. Several tables to assist in the identification of stroke are included (table 3 and figure 8 and table 16). Detailed discussions of the clinical manifestations and acute management of stroke are provided separately. (See "Clinical diagnosis of stroke subtypes" and "Initial assessment and management of acute stroke" and "Neuroimaging of acute stroke".)

Left-sided neglect is a cortically mediated deficit that usually occurs in right hemispheric strokes. The centers controlling conjugate gaze are located in each frontal lobe. If one center is damaged, the unopposed action of the other causes the eyes to deviate toward the side of the lesion and away from the hemiplegia.

Contralateral homonymous hemianopia is another important ocular finding that occurs in association with a lesion anywhere along the hemispheral visual pathways. Cortical sensory loss accompanies the hemiplegia if the sensory cortex, located across the Rolandic fissure, is involved. Rarely, a cortical infarction causes pure motor hemiparesis, with no cortical findings other than hemiparesis [10]. (See "Homonymous hemianopia".)

Lacunar syndromes and basal ganglia lesions — Lesions in the subcortical cerebral hemisphere, namely deep hemorrhages due to hypertension and ischemic lacunar strokes, can cause weakness. Importantly, cortical deficits are absent (table 17). (See "Lacunar infarcts".)

Lacunar infarcts are small, deep cerebral infarcts due almost exclusively to disease of the perforating arterioles. They account for about one quarter of strokes [11]. Each of the classically described lacunar syndromes has also been associated with nonischemic lesions, including hemorrhage and tumor.

Brainstem processes — Weakness syndromes referable to the brainstem may present with "crossed" findings (ipsilateral cranial nerve weakness and contralateral hemiparesis) due to lesions involving cranial nerve nuclei or their tracts and the corticospinal tract before its decussation. However, this is often not the case. As an example, a lacunar stroke in the pons can appear indistinguishable clinically (hemiparesis, ataxia) from a lacunar stroke affecting the internal capsule. (See "Posterior circulation cerebrovascular syndromes".)

Brown-Sequard syndrome — Most myelopathies present with bilateral weakness, even if the distribution is patchy. The main exception is Brown-Sequard syndrome. In this syndrome, involvement of either lateral hemisection of the spinal cord results in ipsilateral hemiplegia or monoplegia, ipsilateral loss of vibration and proprioception, and contralateral loss of pain and temperature below the level of the lesion. Brown-Sequard syndrome is typically caused by penetrating trauma, but idiopathic cases from preexisting dural defects have been reported [12-15]. (See "Anatomy and localization of spinal cord disorders", section on 'Brown-Sequard (hemicord) syndrome'.)

Radiculopathies — Spinal nerve roots mark the beginning of the peripheral nervous system. Radiculopathies are any disease or condition affecting the spinal nerve roots. These syndromes typically present as pain in a dermatomal distribution. Weakness in a myotomal distribution can occur but is less common [16]. Most spinal nerve compression syndromes usually involve either the cervical or lumbosacral region (table 8 and figure 1 and table 9 and figure 2 and table 10) [17]. (See "Clinical features and diagnosis of cervical radiculopathy" and "Acute lumbosacral radiculopathy: Etiology, clinical features, and diagnosis".)

Plexopathies — The best way to diagnose a plexopathy (eg, brachial plexopathy, thoracic outlet syndrome, lumbar plexopathy) is to identify a motor and sensory deficit in a limb that involves more than one spinal or peripheral nerve. Lower motor neuron (LMN) signs are more prominent than sensory findings. Causes include trauma, radiation therapy, and malignancy [18,19]. (See "Brachial plexus syndromes" and "Overview of cancer pain syndromes", section on 'Plexopathies' and "Lumbosacral plexus syndromes".)

Peripheral nerve injuries — Peripheral nerve injuries typically involve neurapraxias, implying a temporary insult to the nerve that resolves when compression is relieved. The most common sites for such neurapraxias are narrow passageways in which the nerve moves during flexion and extension of the neighboring joint. Carpal tunnel syndrome is a classic example. (See "Overview of upper extremity peripheral nerve syndromes" and "Overview of lower extremity peripheral nerve syndromes".)

Bilateral weakness

Cortical or brainstem lesions — A lesion in the central nervous system causing bilateral weakness is usually accompanied by diminished mental status, unless the pathology resides in the spinal cord. The simultaneous occurrence of bilateral, symmetric lesions of the motor cortex is highly unlikely, but a lesion in the interhemispheric fissure, such as a parasagittal meningioma, could result in paraparesis simulating a spinal cord lesion. This is because the areas involved with lower extremity function lie on the medial sides of the motor strip of each cerebral cortex and face each other in the interhemispheric fissure.

A small lesion at the decussation of the pyramids (cruciate paralysis of Bell) can cause bilateral paralysis of the upper extremities, with no involvement of the lower extremities [20-22]. Such lesions are rare and generally result from trauma.

The "locked-in syndrome" is a devastating central process that causes quadriparesis and mutism, but leaves consciousness intact [23]. In this syndrome, a ventral pontine lesion causes quadriplegia, facial weakness, lateral gaze weakness, and dysarthria. Vertical gaze and eyelid opening are preserved. In the ED it is critical to distinguish between a comatose patient and one with locked-in syndrome, who may be a candidate for reperfusion therapy to recanalize a basilar artery occlusion [24,25]. (See "Locked-in syndrome" and "Stupor and coma in adults".)

Myelopathies — Nontraumatic myelopathies (diseases of the spinal cord) typically present with bilateral extremity weakness and sensory deficits. Leg weakness is the most common symptom, but arm weakness can occur when the cervical spinal cord is involved. Bowel and bladder symptoms may be present due to interruption of the descending upper motor neuron pathways, which control the urinary and rectal sphincters [26]. Impotence or priapism may occur. Disturbances in autonomic function, such as loss of sweating, trophic skin changes, loss of temperature control, and vasomotor instability, can occur below the level of the lesion. (See "Disorders affecting the spinal cord".)

The onset of a myelopathy can help to determine its cause. Without a history of trauma, acute disruptions suggest vascular pathology; subacute or chronic development of symptoms suggests an inflammatory lesion.

Myelopathies generally present with some signs of upper motor neuron (UMN) disease. However, the traumatic and vascular processes that cause myelopathy often produce a flaccid areflexic paralysis initially. Over several hours to days, the motor findings become characteristic of an UMN paralysis, with hyperreflexia and bilateral extensor plantar responses (positive Babinski test). Discrete dermatomal sensory loss may not be obvious on examination.

Myelopathy is generally diagnosed by history and physical examination, and confirmed by MRI. Emergent consultation with a spine surgeon is essential if an epidural compression syndrome is suspected, since prognosis depends upon the patient's neurologic function at the time of intervention. Specifically, among patients with epidural compression due to a malignancy, those who are ambulatory generally remain so, while only ten percent of patients who are paraplegic at the time of surgery regain ambulation [27,28].

Polyneuropathy — Polyneuropathies (eg, Guillain-Barré syndrome) manifest both motor and sensory symptoms, unlike myopathies and NMJ disorders. Weakness is due to the large number of nerves involved. Distal power is reduced most dramatically. Deep tendon reflexes are characteristically diminished and vibratory sense is invariably lost distally. Paresthesias often herald the weakness from a peripheral neuropathy [1,29]. (See "Overview of polyneuropathy" and "Guillain-Barré syndrome in adults: Pathogenesis, clinical features, and diagnosis".)

Neuromuscular junction processes — The most common presenting signs of disease at the neuromuscular junction (NMJ) are ocular (ptosis, diplopia) and bulbar abnormalities (dysarthria, dysphagia). Limb weakness, when it occurs, tends to involve the upper extremities [30]. (See "Overview of neuromuscular junction toxins".)

Myasthenia gravis (MG) is a common example and diagnosis is straightforward in a patient with ocular, bulbar, and limb weakness that fluctuates throughout the day and improves with rest. The patient with milder disease or disease limited to a specific muscle group can present a diagnostic dilemma. Provocative maneuvers can help make the diagnosis in such cases. (See "Diagnosis of myasthenia gravis".)

Myopathies — Myopathies result from disease within the myocyte (eg, muscular dystrophy) or as a manifestation of a systemic disorder (eg, metabolic or inflammatory polymyopathies). Sensory abnormalities are usually absent in patients with generalized myopathies, while myalgias may occur in those with inflammatory myopathies. Weakness typically begins in the proximal muscles and extends distally thereafter. Deep tendon reflexes are often maintained until severe weakness develops. Hence, hyporeflexia cannot be relied upon to distinguish myopathies from other neuromuscular processes. Although atrophy is a characteristic finding, it

is not apparent initially when large muscle groups are involved. (See "Myopathies of systemic disease".)

Myotonic muscular dystrophy type 1 presents with distal muscle involvement. Myotonic muscular dystrophy type 2 (ie, proximal myotonic dystrophy) most often presents with weakness in the muscles of the hip girdle.

DISPOSITION

Patients with a new, undiagnosed process causing weakness that is suspected to be neuromuscular or toxin mediated should be admitted. Consultation with neurology and toxicology services is obtained as needed. Any patient at risk of developing respiratory or cardiovascular instability should be admitted to a highly monitored setting, typically an intensive care unit.

Patients without a clear diagnosis but at risk for a polyneuropathy such as GBS should generally be admitted. Patients who insist upon discharge must be advised to return to the ED immediately if worsening weakness or difficulty breathing develops, and an adult capable of providing assistance should be at home with them.

Patients who appear well and will be discharged, but in whom potentially dangerous diagnoses cannot be definitively excluded, should be provided with written discharge instructions that include specific guidance about danger signs to look for and when and where to return should these develop. As an example, most patients with GBS are initially misdiagnosed and discharged home [1].

Most patients complaining of weakness do not have a neurologic emergency or rapidly progressive neuromuscular disease and can be safely discharged home after a thorough, systematic assessment has ruled out pathology requiring admission. Discharged patients should be referred to a specialist or their primary care provider for further evaluation.

SUMMARY AND RECOMMENDATIONS

- **Causes and assessment** – Weakness is a common, nonspecific emergency department (ED) complaint that encompasses a broad differential diagnosis including neurologic and non-neurologic diseases. In older adults, infection, cardiovascular disease, and dehydration must be considered in the differential diagnosis. (See 'Differential diagnosis of acute weakness' above.)

The diagnosis of potentially life-threatening neurologic and neuromuscular processes requires a systematic, anatomic approach based upon a careful history, physical examination, and in some cases, imaging studies. (See 'History' above and 'Physical examination' above and 'Ancillary studies' above.)

- **Airway, breathing, circulation** – The emergency clinician's first responsibility is to rule out life-threatening or permanently disabling causes of weakness that require urgent treatment. The immediate life threats from acute neuromuscular weakness include inability to protect or maintain the airway, respiratory failure from thoracic and diaphragmatic muscle weakness, and circulatory collapse from autonomic instability. (See 'Assessment of life threatening illness' above.)

- **Acute ischemic stroke** – Patients diagnosed with an acute ischemic stroke should be rapidly evaluated to determine appropriate treatment with systemic thrombolytic therapy or emergency endovascular revascularization. (See "Overview of the evaluation of stroke" and "Initial assessment and management of acute stroke".)

- **Algorithmic approach to diagnosis** – Once life-threatening problems have been addressed or ruled out, approach the patient with objective weakness in a systematic manner, as outlined in the accompanying algorithm (algorithm 1). The first step is to determine whether the weakness is unilateral (asymmetric) or bilateral (symmetric), and to look closely for signs of central neurologic involvement.

  When assessing acute weakness, it is helpful to **begin cephalad and centrally and then progress caudad and peripherally**. This approach provides a reliable framework for neuroanatomic localization and accurate diagnosis.

  - **Unilateral weakness** – If unilateral weakness is identified, look carefully for signs suggestive of cortical, subcortical (lacunar), or brainstem lesions. If these are absent, a peripheral process (radiculopathy, plexopathy, or peripheral nerve injury) most likely accounts for the patient's symptoms.

  - **Bilateral weakness** – If bilateral weakness is identified, consider the patient's mental status and look carefully for signs of upper or lower motor neuron lesions and associated abnormalities. The constellation of examination findings should allow approximate identification of the site of the lesion and determination of the need for imaging studies, specialist consultation, and treatment.

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Topic 294 Version 20.0

GRAPHICS

Acute stroke differential diagnosis

Migraine aura
Seizure with postictal paresis (Todd paralysis), aphasia, or neglect
Central nervous system tumor or abscess
Cerebral venous thrombosis
Functional deficit (conversion reaction)
Hypertensive encephalopathy
Head trauma
Mitochondrial disorder (eg, mitochondrial encephalopathy with lactic acidosis and stroke-like episodes or MELAS)
Multiple sclerosis
Posterior reversible encephalopathy syndrome (PRES)
Reversible cerebral vasoconstriction syndromes (RCVS)
Spinal cord disorder (eg, compressive myelopathy, spinal dural arteriovenous fistula)
Subdural hematoma
Syncope
Systemic infection
Toxic-metabolic disturbance (eg, hypoglycemia, exogenous drug intoxication)
Transient global amnesia
Viral encephalitis (eg, herpes simplex encephalitis)
Wernicke encephalopathy

Graphic 69869 Version 7.0

Characteristics of stroke subtypes

Stroke type	Clinical course	Risk factors	Other clues
Intracerebral hemorrhage	Gradual onset and progression during minutes or hours in most patients, but may present abruptly with maximal deficit at onset.	Hypertension, trauma, bleeding diatheses, illicit drugs (eg, amphetamines, cocaine), vascular malformations. More common in Black people and Asian people than in White people.	May be precipitated by sex or other physical activity. Patient may have reduced alertness
Subarachnoid hemorrhage	Abrupt onset of sudden, severe headache. Focal brain dysfunction less common than with other types.	Smoking, hypertension, moderate to heavy alcohol use, genetic susceptibility (eg, polycystic kidney disease, family history of subarachnoid hemorrhage) and sympathomimetic drugs (eg, cocaine)	May be precipitated by sex or other physical activity. Patient may have reduced alertness
Ischemic (thrombotic)	Stuttering progression with periods of improvement. Lacunes develop over hours or at most a few days; large artery ischemia may evolve over longer periods.	Atherosclerotic risk factors (age, smoking, diabetes mellitus, etc). Males affected more commonly than females. May have history of TIA.	May have neck bruit.
Ischemic (embolic)	Sudden onset with deficit maximal at onset. Clinical findings may improve quickly.	Atherosclerotic risk factors as listed above. Males affected more commonly than females. History of heart disease (valvular, atrial fibrillation, endocarditis).	Can be precipitated by getting up at night to urinate o sudden coughing or sneezing.

Graphic 69907 Version 5.0

Acute ischemic stroke syndromes according to vascular territory

Artery involved	Syndrome
Anterior cerebral artery	Motor and/or sensory deficit (leg > face, arm) Grasp, sucking reflexes Abulia, paratonic rigidity, gait apraxia
Middle cerebral artery	Dominant hemisphere: aphasia, motor and sensory deficit (face, arm > le > foot), may be complete hemiplegia if internal capsule involved, homonymous hemianopia Non-dominant hemisphere: neglect, anosognosia, motor and sensory deficit (face, arm > leg > foot), homonymous hemianopia
Posterior cerebral artery	Homonymous hemianopia; alexia without agraphia (dominant hemisphere); visual hallucinations, visual perseverations (calcarine cortex); sensory loss, choreoathetosis, spontaneous pain (thalamus); III nerve palsy, paresis of vertical eye movement, motor deficit (cerebral peduncle, midbrain)
Penetrating vessels	Pure motor hemiparesis (classic lacunar syndromes) Pure sensory deficit Pure sensory-motor deficit Hemiparesis, homolateral ataxia Dysarthria/clumsy hand
Vertebrobasilar	Cranial nerve palsies Crossed sensory deficits Diplopia, dizziness, nausea, vomiting, dysarthria, dysphagia, hiccup Limb and gait ataxia Motor deficit Coma Bilateral signs suggest basilar artery disease
Internal carotid artery	Progressive or stuttering onset of MCA syndrome, occasionally ACA syndrome as well if insufficient collateral flow

Graphic 75487 Version 7.0

Suggestive and atypical features of multiple sclerosis

Features suggestive of multiple sclerosis
Relapses and remissions
Onset between ages 15 and 50 years
Optic neuritis
Lhermitte sign
Internuclear ophthalmoplegia
Fatigue
Heat sensitivity (Uhthoff phenomenon)
Features atypical for multiple sclerosis
Steady progression
Onset before age 10 or after age 50 years
Cortical deficits such as aphasia, apraxia, alexia, or neglect
Rigidity or sustained dystonia
Convulsions
Early dementia
Deficit developing within minutes

Graphic 63940 Version 6.0

Manifestations of multiple sclerosis

Symptoms and signs	Total (percent)
Sensory in limbs	31
Visual loss	16
Motor (subacute)	9
Diplopia	7
Gait disturbance	5
Motor (acute)	4
Balance problems	3
Sensory in face	3
Lhermitte sign (electric shock-like sensations that run down the back and/or limbs upon flexion of the neck)	2
Vertigo	2
Bladder problems	1
Limb ataxia	1
Acute transverse myelopathy	1
Pain	<1
Other	3
Polysymptomatic onset	14

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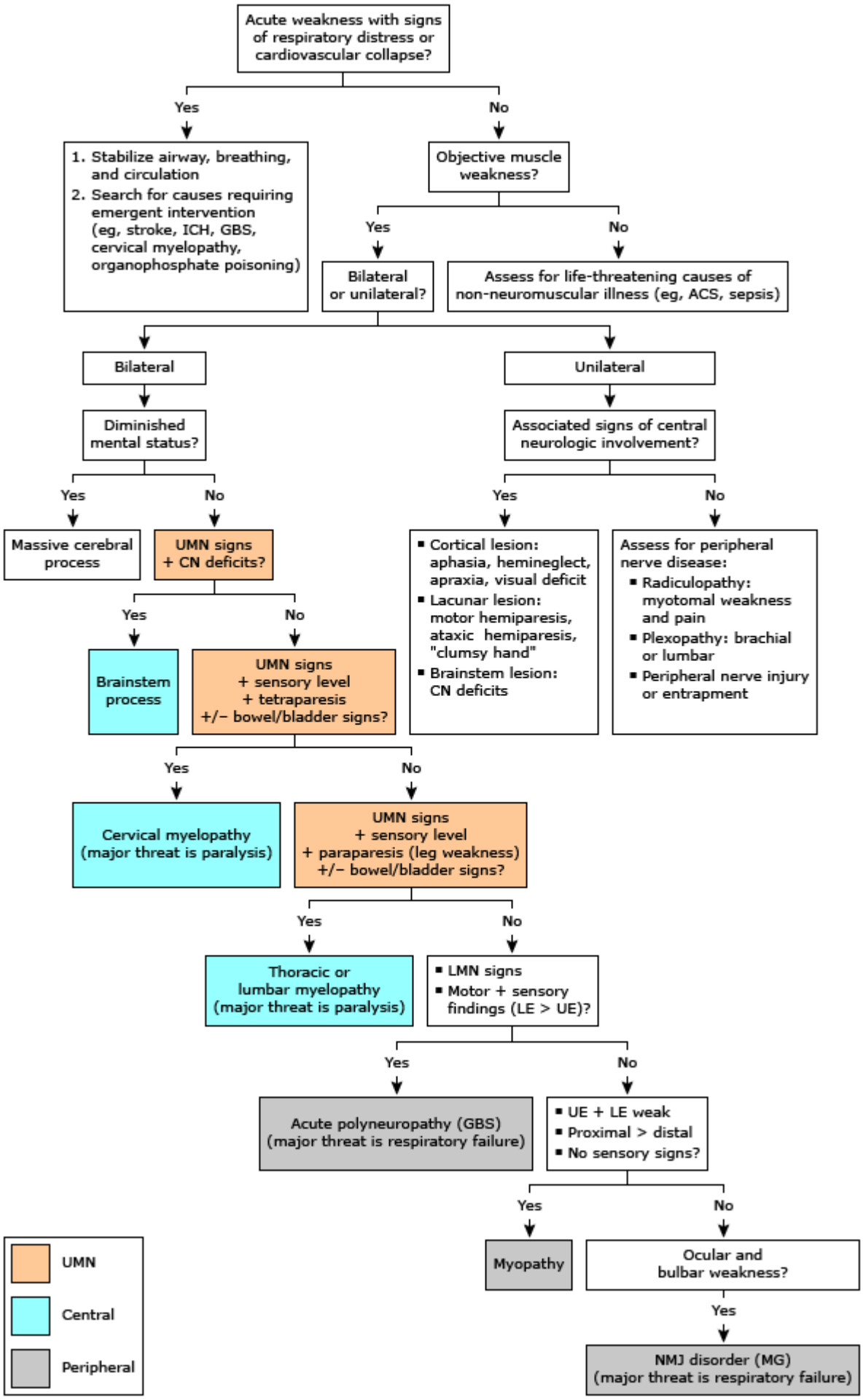
Graphic 61789 Version 5.0

Major symptoms and signs of hypothyroidism

Mechanism	Symptoms	Signs
Slowing of metabolic processes	Fatigue and weakness Cold intolerance Dyspnea on exertion Weight gain Cognitive dysfunction Intellectual disability (infantile onset) Constipation Growth failure	Slow movement and slow speech Delayed relaxation of tendon reflexes Bradycardia Carotenemia
Accumulation of matrix substances	Dry skin Hoarseness Edema	Coarse skin Puffy facies and loss of eyebrows Periorbital edema Enlargement of the tongue
Other	Decreased hearing Myalgia and paresthesia Depression Menorrhagia Arthralgia Pubertal delay	Diastolic hypertension Pleural and pericardial effusions Ascites Galactorrhea

Graphic 62676 Version 5.0

Approach to the adult with weakness in the emergency department


Acute weakness with signs of respiratory distress or cardiovascular collapse?
Yes
No
1. Stabilize airway, breathing, and circulation
2. Search for causes requiring emergent intervention (eg, stroke, ICH, GBS, cervical myelopathy, organophosphate poisoning)
Objective muscle weakness?
Yes
No
Bilateral or unilateral?
Assess for life-threatening causes of non-neuromuscular illness (eg, ACS, sepsis)
Bilateral
Unilateral
Diminished mental status?
Associated signs of central neurologic involvement?
Yes
No
Yes
No
Massive cerebral process
UMN signs + CN deficits?
▪ Cortical lesion: aphasia, hemineglect, apraxia, visual deficit
▪ Lacunar lesion: motor hemiparesis, ataxic hemiparesis, "clumsy hand"
▪ Brainstem lesion: CN deficits
Assess for peripheral nerve disease:
▪ Radiculopathy: myotomal weakness and pain
▪ Plexopathy: brachial or lumbar
▪ Peripheral nerve injury or entrapment
Yes
No
Brainstem process
UMN signs + sensory level + tetraparesis +/− bowel/bladder signs?
Yes
No
Cervical myelopathy (major threat is paralysis)
UMN signs + sensory level + paraparesis (leg weakness) +/− bowel/bladder signs?
Yes
No
Thoracic or lumbar myelopathy (major threat is paralysis)
▪ LMN signs
▪ Motor + sensory findings (LE > UE)?
Yes
No
Acute polyneuropathy (GBS) (major threat is respiratory failure)
▪ UE + LE weak
▪ Proximal > distal
▪ No sensory signs?
Yes
No
Myopathy
Ocular and bulbar weakness?
Yes
NMJ disorder (MG) (major threat is respiratory failure)
UMN
Central
Peripheral

ICH: intracerebral hemorrhage; GBS: Guillan-Barré syndrome; ACS: acute coronary syndrome; UMN: upper motor neuron; CN: cranial nerve; LMN: lower motor neuron; LE: lower extremities; UE: upper extremities; NMJ: neuromuscular junction; MG: myasthenia gravis.

Adapted from: Asimos AW. Weakness: A systematic approach to acute, non-traumatic, neurologic and neuromuscular causes. Emerg Med Pract 2002; 4:1.

Graphic 54645 Version 3.0

Differential diagnosis of acute weakness organized anatomically

Central	
Cerebrum	▪ ***Stroke***
	▪ **Space occupying/structural lesion**
	▪ *Postictal (Todd) paralysis*
	▪ *Multiple sclerosis*
	▪ Other inflammatory conditions
Subcortex/brainstem	▪ ***Stroke***
Spinal cord	▪ **Acute transverse myelitis**
	▪ **Spinal cord infarct**
	▪ **Spinal epidural or subdural hemorrhage**
	▪ **Central intervertebral disc herniation**
	▪ **Tumors (metastatic or primary)**
	▪ Multiple sclerosis
Peripheral	
Spinal nerve root	▪ *Intervertebral disc herniation*
	▪ **Epidural abscess**
	▪ Tumors
	▪ Leptomeningeal metastases
Polyneuropathies	▪ **Guillain-Barré syndrome**
	▪ *Diabetic*
	▪ Ciguatoxin (ciguatera poisoning)
	▪ Saxitoxin (paralytic shellfish poisoning)
	▪ Tetrodotoxin poisoning (pufferfish poisoning)
	▪ Porphyria
	▪ Lead or other heavy metal poisoning

	▪ Alcohol or drug induced
Plexopathies	▪ Brachial
	▪ Lumbar
Peripheral neuropathies	▪ **Nerve compression syndromes**
NMJ Disorder	▪ **Myasthenia gravis**
	▪ **Lambert-Eaton myasthenic syndrome**
	▪ **Botulism**
	▪ **Organophosphate poisoning**
	▪ **Tick paralysis**
Myopathy	▪ **Electrolyte induced**
	▪ Inflammatory (polymyositis)
	▪ Alcohol or drug induced
	▪ Muscular dystrophy
	▪ Endocrine related
Nonphysiologic/noncategorical	
▪ Conversion disorder	
▪ Chronic fatigue syndrome (CFS), also known as myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS)	
▪ Anxiety disorders	
▪ Fibromyalgia	
▪ Malingering	

The *italic* bulleted content indicates common conditions, where the **bold** bulleted content indicates serious conditions. ***Bold italic*** bulleted content indicates common serious conditions.

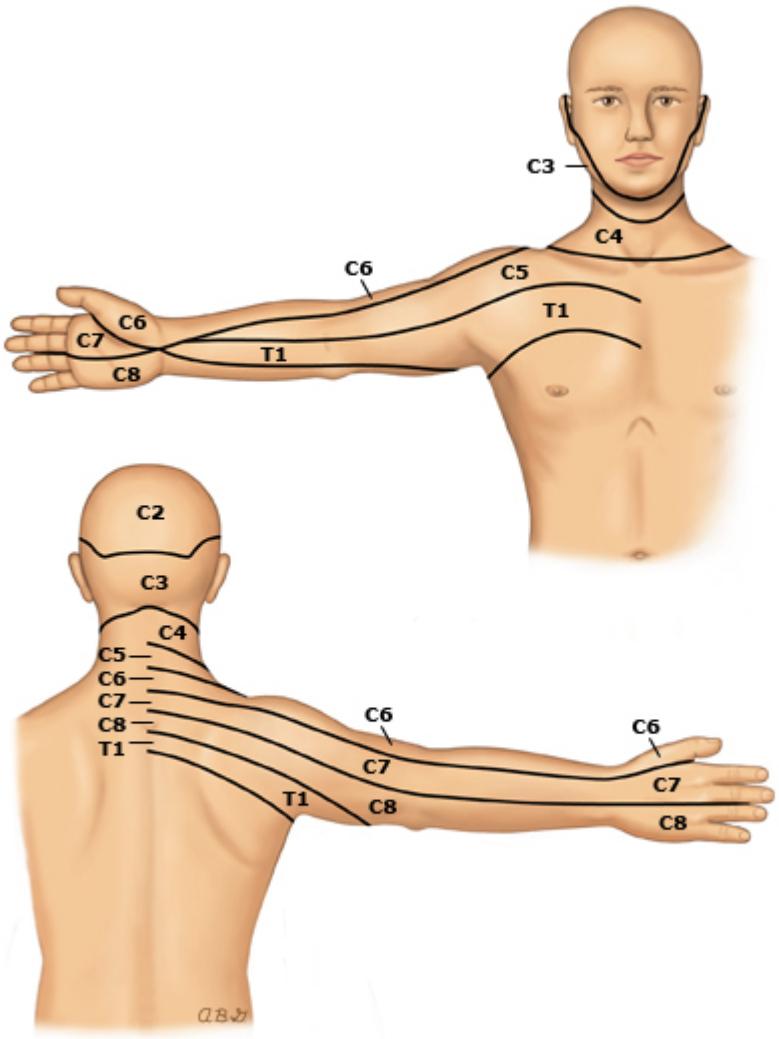
Graphic 61895 Version 6.0

Symptoms and signs of cervical root lesions

Root	Pain	Numbness	Weakness	Reflex affected
C5	Neck, shoulder, scapula	Lateral arm (in distribution of axillary nerve)	Shoulder abduction, external rotation, elbow flexion, forearm supination	Biceps, brachioradialis
C6	Neck, shoulder, scapula, lateral arm, lateral forearm, lateral hand	Lateral forearm, thumb and index finger	Shoulder abduction, external rotation, elbow flexion, forearm supination and pronation	Biceps, brachioradialis
C7	Neck, shoulder, middle finger, hand	Index and middle finger, palm	Elbow and wrist extension (radial), forearm pronation, wrist flexion	Triceps
C8	Neck, shoulder, medial forearm, fourth and fifth digits, medial hand	Medial forearm, medial hand, fourth and fifth digits	Finger extension, wrist extension (ulnar), distal finger flexion, extension, abduction and adduction, distal thumb flexion	None
T1	Neck, medial arm and forearm	Anterior arm and medial forearm	Thumb abduction, distal thumb flexion, finger abduction and adduction	None

Graphic 74632 Version 4.0

Cervical dermatomes



Schematic representation of the cervical and T1 dermatomes. There is no C1 dermatome. Patients with nerve root syndromes may have pain, paresthesias, and diminished sensation in the dermatome of the nerve that is involved.

Graphic 53006 Version 3.0

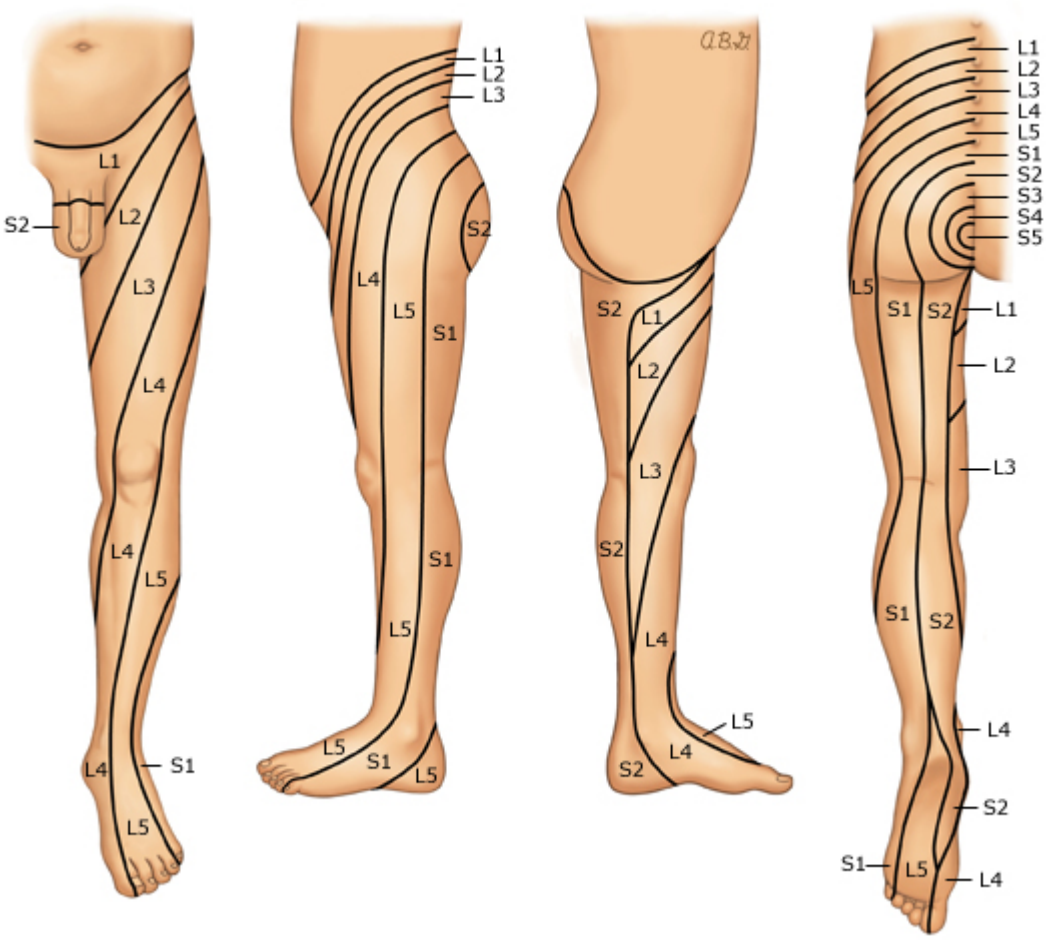
Lumbosacral myotomes

L1
Femoral nerve
Iliopsoas (hip flexion)
L2
Femoral nerve
Iliopsoas (hip flexion)
Quadriceps (knee extension)
Obturator nerve
Hip adductors
L3
Femoral nerve
Iliopsoas (hip flexion)
Quadriceps (knee extension)
Obturator nerve
Hip adductors
L4
Femoral nerve
Iliopsoas (hip flexion)
Quadriceps (knee extension)
Obturator nerve
Hip adductors
L5
Peroneal nerve
Ankle dorsiflexion (tibialis anterior)
Ankle eversion (peroneus muscles)
Tibial nerve
Ankle inversion (tibialis posterior)
Superior gluteal nerve
Hip abduction (gluteus medius)

Leg internal rotation (tensor fascia latae)
S1
Inferior gluteal nerve
Hip extension (gluteus maximus)
Sciatic nerve
Knee flexion (hamstrings)
Tibial nerve
Ankle plantar flexion
S2
Inferior gluteal nerve
Hip extension (Gluteus maximus)
Sciatic nerve
Knee flexion (hamstrings)
Tibial nerve
Ankle plantar flexion

Graphic 64209 Version 2.0

Lumbosacral dermatomes



Schematic representation of the lumbosacral dermatomes. Patients with sciatica may have pain, paresthesias, and diminished sensation in the dermatome of the nerve root that is involved.

Graphic 50419 Version 2.0

Symptoms and signs of lumbosacral radiculopathies

Nerve	Symptoms	Signs
L5	Back pain radiating down the lateral leg to foot	Decreased foot dorsiflexion, toe extension, foot inversion and eversion; mild weakness of leg abduction in severe cases
S1	Pain radiating down the posterior leg to foot; leg pain greater than back pain	Decreased leg extension, foot inversion, plantar flexion, an toe flexion; decreased sensation in the posterior leg and lateral foot; loss of ankle jerk
L2-4	Acute back pain radiating around the anterior leg into the knee and possibly foot	Decreased hip flexion, knee extension, leg abduction; decreased sensation in the anterior thigh down the medial aspect of the shin; diminished knee jerk in severe cases
S2-4	Sacral or buttock pain radiating down the posterior leg or into the perineum	Minimal weakness; bowel and bladder dysfunction

Graphic 75974 Version 3.0

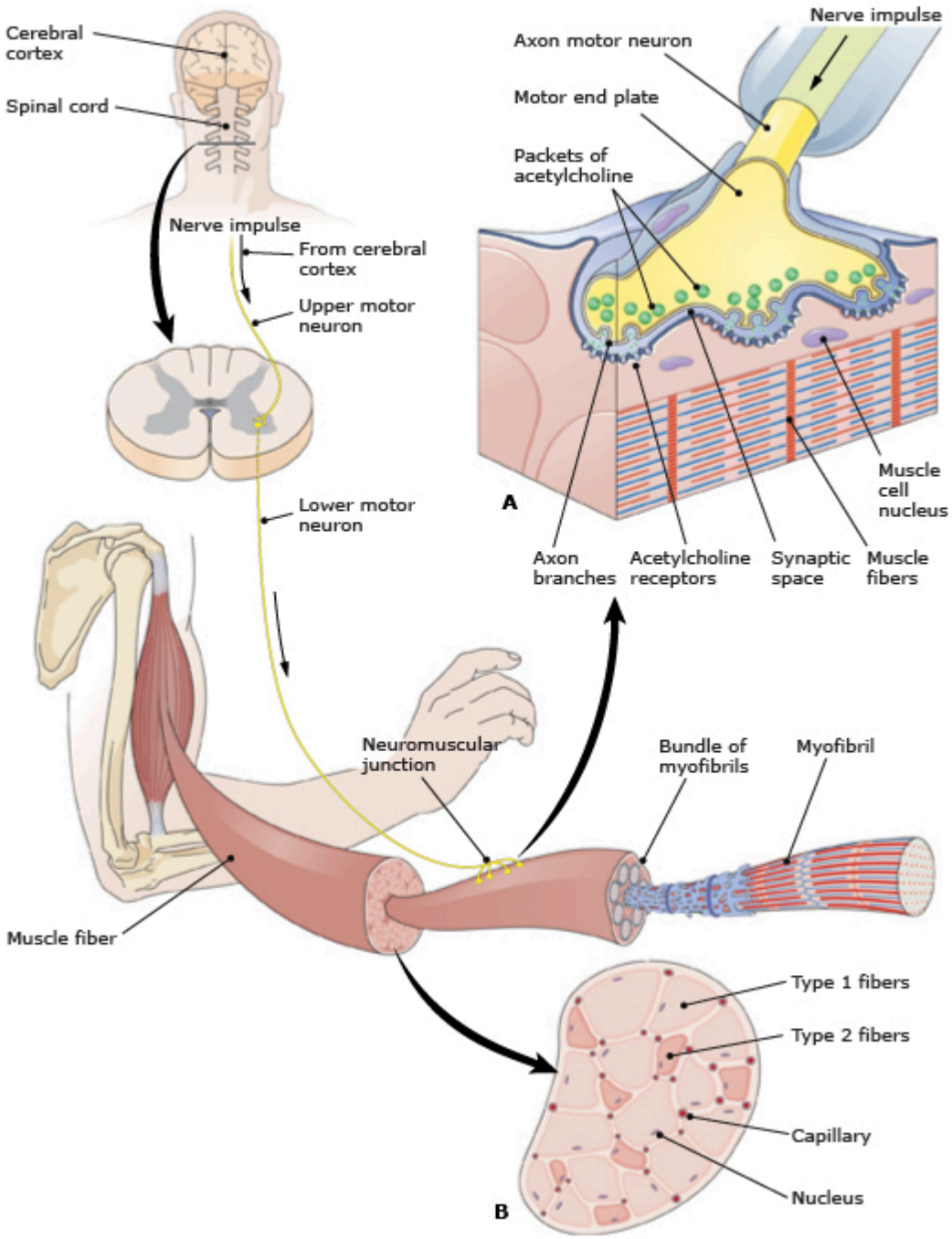
Signs of upper versus lower motor neuron disease

Clinical	UMN	LMN
Muscle tone	Increased	Decreased
Reflexes	Hyperreflexia	Hyporeflexia
Babinski sign	Present	Absent

UMN: upper motor neuron; LMN: lower motor neuron.

Graphic 56458 Version 2.0

Schematic diagram of neuromuscular motor function



The motor unit (neuromuscular apparatus). Voluntary muscle control begins with a nerve impulse from the brain, which is relayed by an upper motor neuron to muscle by a lower motor neuron. A) Detail of the motor end plate, where nerve meets muscle. A nerve impulse causes discharge of acetylcholine into the synaptic space, where it attaches to receptors on muscle cells and causes muscle fiber contraction. B) Detail of skeletal muscle. In this microscopic depiction, type 1 (slow twitch) fibers are pale and type 2 (fast twitch) are dark.

Reproduced with permission from: McConnell, TH. The nature of disease pathology for the health professions, Philadelphia:

Graphic 62641 Version 2.0

Comparison of findings in neuropathy, myopathy, and NMJ disorders

	Neuropathy	Myopathy	Neuromuscular junction
Typical distribution	Distal > proximal	Proximal > distal	Diffuse (oculomotor and bulbar early)
Reflexes	Decreased	Normal to decreased	Normal
Sensory involvement	Present	Absent	Absent

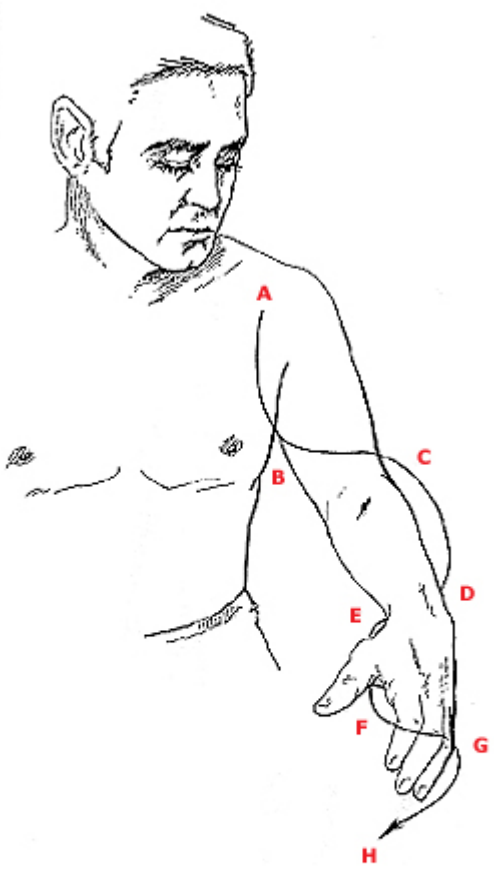
Graphic 75434 Version 2.0

Motor function score

Score	Response
0	No contraction
1	Flicker or trace of contraction
2	Active movement with gravity eliminated
3	Active movement against gravity
4	Active movement against gravity and resistance
5	Normal power

Graphic 77388 Version 2.0

Nerve roots and peripheral nerves corresponding to the principal movements of the upper extremity



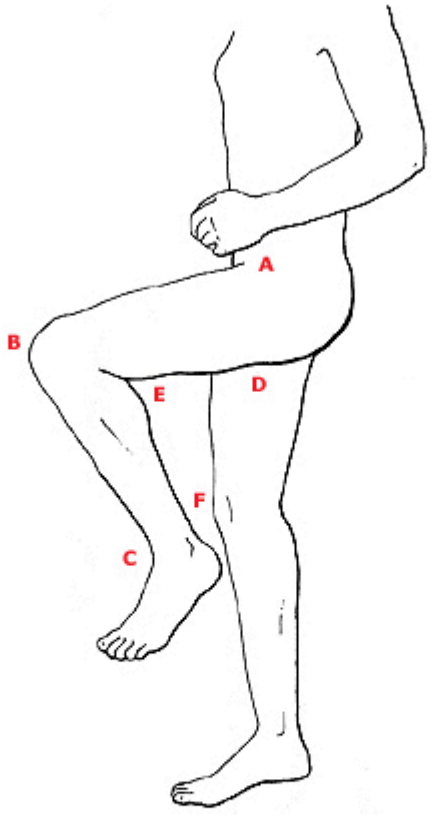
Movement	Nerve root	Peripheral nerve
A. Shoulder abduction	C5	Axillary
B. Elbow flexion	C5-6	Musculocutaneous
C. Elbow extension	C6-7	Radial
D. Wrist extension	C6-7	Radial
E. Wrist flexion	C7-8	Median
F. Finger flexion	C8	Median
G. Finger extension	C8	Radial
H. Finger abduction	T1	Ulnar

The letters labeling the movements form a spiral down the extremity. The nerve roots and peripheral nerves corresponding to each movement are listed below. Figure redrawn with permission from Gelb, DJ. The Neurologic Examination.

In: Introduction to Clinical Neurology. Woburn, MA, Butterworth-Heinemann 2000.

Graphic 58656 Version 1.0

Nerve roots and peripheral nerves corresponding to the principal movements of the lower extremity



Movement	Nerve roots	Peripheral nerve
A. Hip flexion	L2-3	Femoral ("nerve to iliopsoas")
B. Knee extension	L3-4	Femoral
C. Ankle dorsiflexion	L4-5	Peroneal
D. Hip extension	L4-5	Gluteal
E. Knee flexion	L5-S1	Sciatic
F. Ankle plantar flexion	S1-2	Tibial

The letters labeling the movements proceed in order from proximal to distal down the front of the limb, and then repeat from proximal to distal down the back of the limb. The nerve roots and peripheral nerves corresponding to each movement are listed below.

Figure redrawn with permission from Gelb DJ. The Neurologic Examination. In: Introduction to Clinical Neurology. Woburn, MA, Butterworth-Heinemann 2000.

Graphic 64884 Version 2.0

Solitary nerve root lesions of the lumbosacral spine

Root	Pain	Sensory loss	Weakness	Stretch reflex loss
L1	Inguinal region	Inguinal region	Rarely hip flexion	None
L2-L3-L4	Back, radiating into anterior thigh, and at times medial lower leg	Anterior thigh, occasionally medial lower leg	Hip flexion, hip adduction, knee extension	Patellar tendon
L5	Back, radiating into buttock, lateral thigh, lateral calf and dorsum foot, great toe	Lateral calf, dorsum foot, web space between first and second toe	Hip abduction, knee flexion, foot dorsiflexion, toe extension and flexion, foot inversion and eversion	Semitendinosus/semimembranosus (internal hamstrings) tendon
S1	Back, radiating into buttock, lateral or posterior thigh, posterior calf, lateral or plantar foot	Posterior calf, lateral or plantar aspect of foot	Hip extension, knee flexion, plantar flexion of the foot	Achilles tendon
S2-S3-S4	Sacral or buttock pain radiating into the posterior aspect of the leg or the perineum	Medial buttock, perineal, and perianal regions	Weakness may be minimal, with urinary and fecal incontinence as well as sexual dysfunction	Bulbocavernosus, anal wink

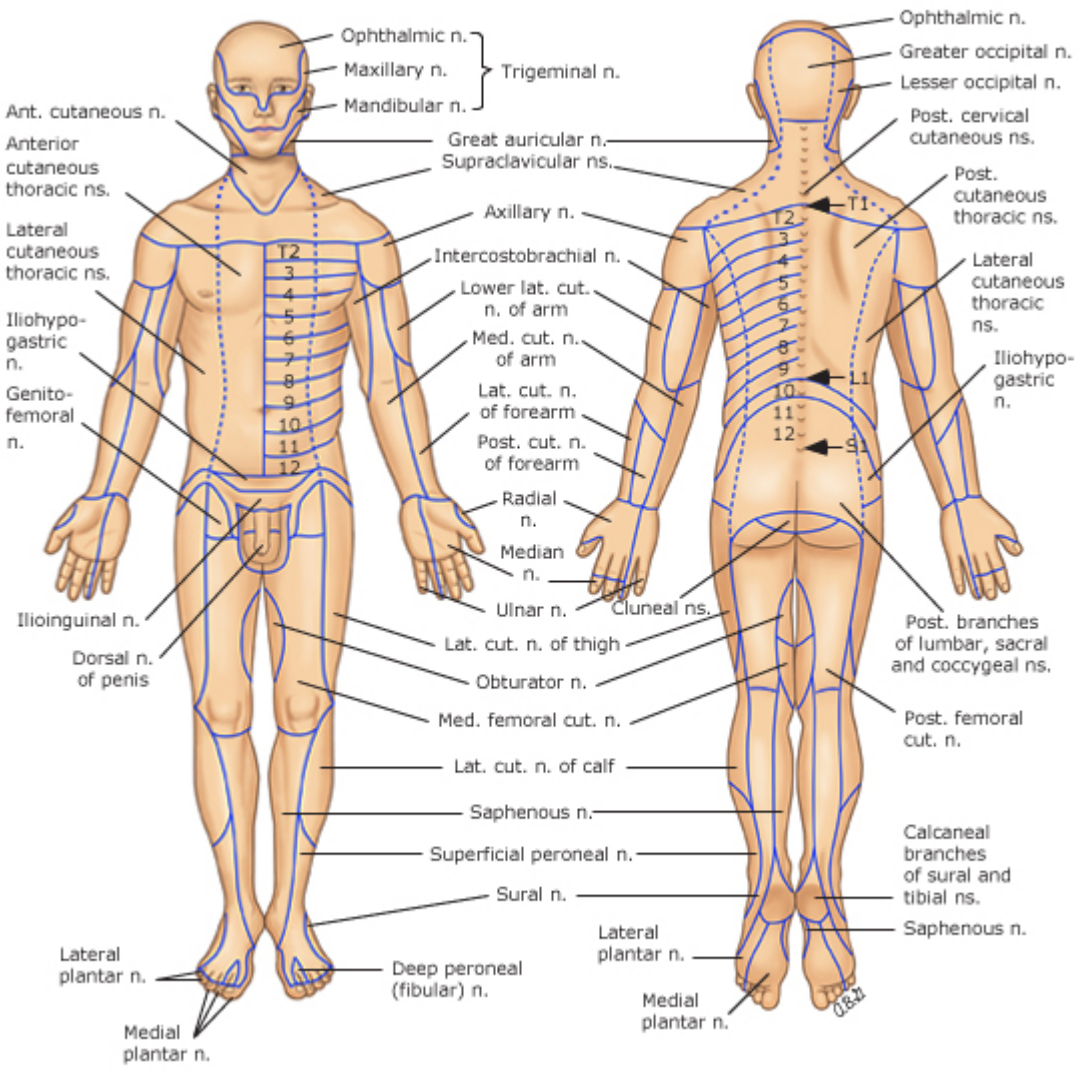
Graphic 75645 Version 4.0

Reflex examination score

Score	Description
0	Reflex absent
1	Reflex slight, less than normal (includes a trace response or a response brought out only by reinforcement)
2	Reflex in lower half of normal range
3	Reflex in upper half of normal range
4	Reflex enhanced, more than normal (includes clonus if present)

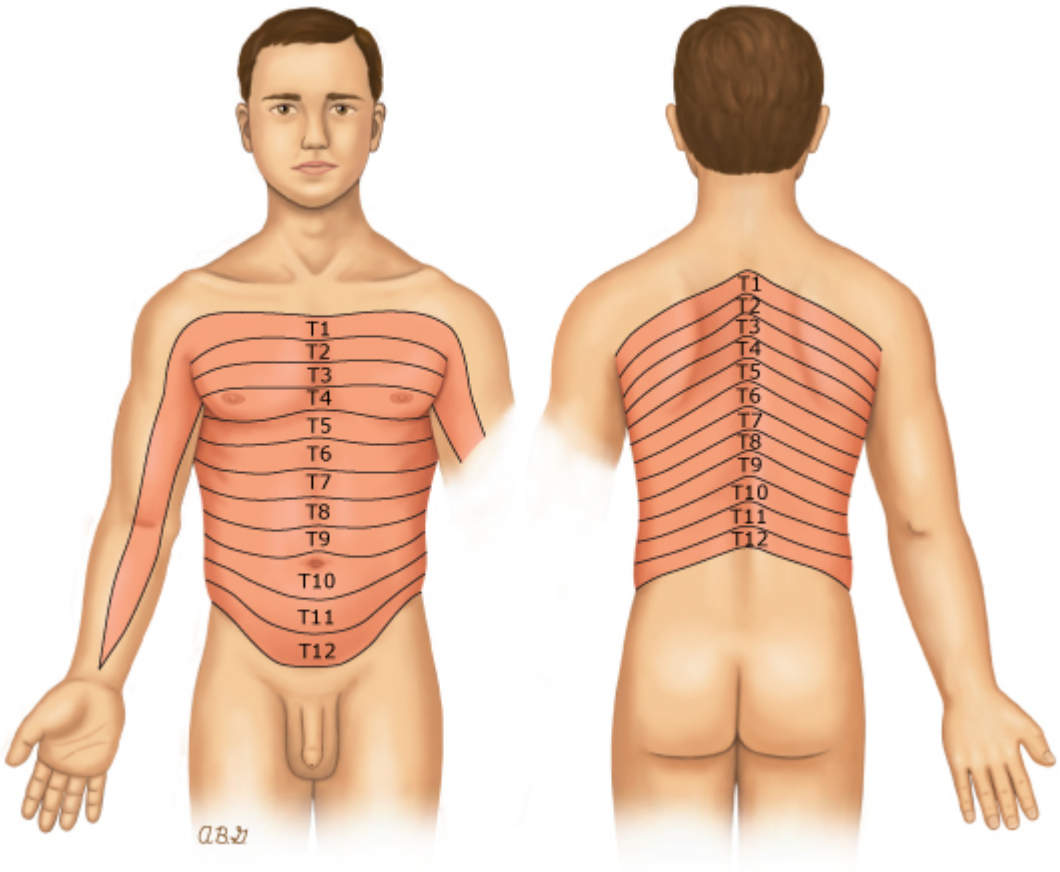
Graphic 82522 Version 12.0

Pattern of innervation of skin by peripheral nerves



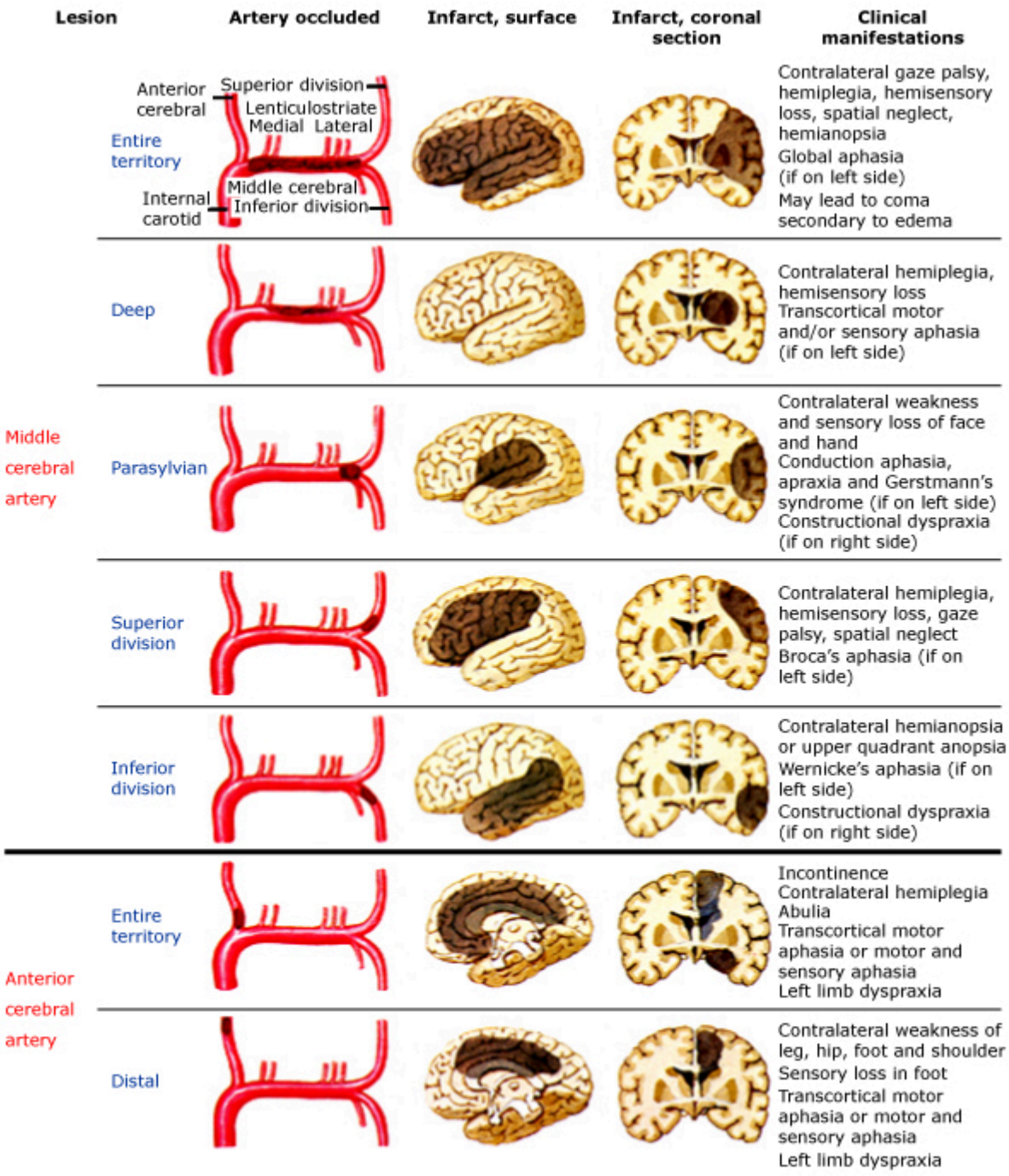
Graphic 54042 Version 5.0

Thoracic dermatomes



Graphic 64754 Version 2.0

Occlusion of middle and anterior cerebral arteries



Graphic 69630 Version 1.0

National Institutes of Health Stroke Scale (NIHSS)

Administer stroke scale items in the order listed. Record performance in each category after each subscale exam. Do not go back and change scores. Follow directions provided for each exam technique. Scores should reflect what the patient does, not what the clinician thinks the patient can do. The clinician should record answers while administering the exam and work quickly. Except where indicated, the patient should not be coached (ie, repeated requests to patient to make a special effort).

Instructions	Scale definition	Score
1a. Level of consciousness: The investigator must choose a response if a full evaluation is prevented by such obstacles as an endotracheal tube, language barrier, orotracheal trauma/bandages. A 3 is scored only if the patient makes no movement (other than reflexive posturing) in response to noxious stimulation.	0 = **Alert;** keenly responsive. 1 = **Not alert;** but arousable by minor stimulation to obey, answer, or respond. 2 = **Not alert;** requires repeated stimulation to attend, or is obtunded and requires strong or painful stimulation to make movements (not stereotyped). 3 = Responds only with reflex motor or autonomic effects or totally unresponsive, flaccid, and areflexic.	____
1b. Level of consciousness questions: The patient is asked the month and his/her age. The answer must be correct - there is no partial credit for being close. Aphasic and stuporous patients who do not comprehend the questions will score 2. Patients unable to speak because of endotracheal intubation, orotracheal trauma, severe dysarthria from any cause, language barrier, or any other problem not secondary to aphasia are given a 1. It is important that only the initial answer be graded and that the examiner not "help" the patient with verbal or non-verbal cues.	0 = **Answers** both questions correctly. 1 = **Answers** one question correctly. 2 = **Answers** neither question correctly.	____
1c. Level of consciousness commands: The patient is asked to open and close the eyes and then to grip and release the non-paretic hand. Substitute another one step command if the hands cannot be used. Credit is given if an unequivocal attempt is made but not completed due to weakness. If the patient does not respond to command, the task should be demonstrated to him or her (pantomime), and the result scored (ie, follows none, one or two commands). Patients with	0 = **Performs** both tasks correctly. 1 = **Performs** one task correctly. 2 = **Performs** neither task correctly.	____

trauma, amputation, or other physical impediments should be given suitable one-step commands. Only the first attempt is scored.		
2. Best gaze: Only horizontal eye movements will be tested. Voluntary or reflexive (oculocephalic) eye movements will be scored, but caloric testing is not done. If the patient has a conjugate deviation of the eyes that can be overcome by voluntary or reflexive activity, the score will be 1. If a patient has an isolated peripheral nerve paresis (cranial nerves III, IV or VI), score a 1. Gaze is testable in all aphasic patients. Patients with ocular trauma, bandages, pre-existing blindness, or other disorder of visual acuity or fields should be tested with reflexive movements, and a choice made by the investigator. Establishing eye contact and then moving about the patient from side to side will occasionally clarify the presence of a partial gaze palsy.	0 = **Normal.** 1 = **Partial gaze palsy;** gaze is abnormal in one or both eyes, but forced deviation or total gaze paresis is not present. 2 = **Forced deviation,** or total gaze paresis not overcome by the oculocephalic maneuver.	____
3. Visual: Visual fields (upper and lower quadrants) are tested by confrontation, using finger counting or visual threat, as appropriate. Patients may be encouraged, but if they look at the side of the moving fingers appropriately, this can be scored as normal. If there is unilateral blindness or enucleation, visual fields in the remaining eye are scored. Score 1 only if a clear-cut asymmetry, including quadrantanopia, is found. If patient is blind from any cause, score 3. Double simultaneous stimulation is performed at this point. If there is extinction, patient receives a 1, and the results are used to respond to item 11.	0 = **No visual loss.** 1 = **Partial hemianopia.** 2 = **Complete hemianopia.** 3 = **Bilateral hemianopia** (blind including cortical blindness).	____
4. Facial palsy: Ask - or use pantomime to encourage - the patient to show teeth or raise eyebrows and close eyes. Score symmetry of grimace in response to noxious stimuli in the poorly responsive or non-comprehending patient. If facial trauma/bandages, orotracheal tube, tape or other physical barriers obscure the face, these should be removed to the extent possible.	0 = **Normal** symmetrical movements. 1 = **Minor paralysis** (flattened nasolabial fold, asymmetry on smiling). 2 = **Partial paralysis** (total or near-total paralysis of lower face). 3 = **Complete paralysis** of one or both sides (absence of facial movement in the upper and lower face).	____

5. Motor arm: The limb is placed in the appropriate position: extend the arms (palms down) 90 degrees (if sitting) or 45 degrees (if supine). Drift is scored if the arm falls before 10 seconds. The aphasic patient is encouraged using urgency in the voice and pantomime, but not noxious stimulation. Each limb is tested in turn, beginning with the non-paretic arm. Only in the case of amputation or joint fusion at the shoulder, the examiner should record the score as untestable (UN), and clearly write the explanation for this choice.	0 = **No drift;** limb holds 90 (or 45) degrees for full 10 seconds. 1 = **Drift;** limb holds 90 (or 45) degrees, but drifts down before full 10 seconds; does not hit bed or other support. 2 = **Some effort against gravity;** limb cannot get to or maintain (if cued) 90 (or 45) degrees, drifts down to bed, but has some effort against gravity. 3 = **No effort against gravity;** limb falls. 4 = **No movement.** UN = **Amputation** or joint fusion, explain:_____ **5a. Left arm** **5b. Right arm**	____
6. Motor leg: The limb is placed in the appropriate position: hold the leg at 30 degrees (always tested supine). Drift is scored if the leg falls before 5 seconds. The aphasic patient is encouraged using urgency in the voice and pantomime, but not noxious stimulation. Each limb is tested in turn, beginning with the non-paretic leg. Only in the case of amputation or joint fusion at the hip, the examiner should record the score as untestable (UN), and clearly write the explanation for this choice.	0 = **No drift;** leg holds 30-degree position for full 5 seconds. 1 = **Drift;** leg falls by the end of the 5-second period but does not hit bed. 2 = **Some effort against gravity;** leg falls to bed by 5 seconds, but has some effort against gravity. 3 = **No effort against gravity;** leg falls to bed immediately. 4 = **No movement.** UN = **Amputation** or joint fusion, explain:_____ **6a. Left leg** **6b. Right leg**	____
7. Limb ataxia: This item is aimed at finding evidence of a unilateral cerebellar lesion. Test with eyes open. In case of visual defect, ensure testing is done in intact visual field. The finger-nose-finger and heel-shin tests are performed on both sides, and ataxia is scored only if present out of proportion to weakness. Ataxia is absent in the patient who cannot understand or is paralyzed. Only in the case of amputation or joint fusion, the examiner should record the score as untestable (UN),	0 = **Absent.** 1 = **Present in one limb.** 2 = **Present in two limbs.** UN = **Amputation** or joint fusion, explain:_____	____

and clearly write the explanation for this choice. In case of blindness, test by having the patient touch nose from extended arm position.		
8. Sensory: Sensation or grimace to pinprick when tested, or withdrawal from noxious stimulus in the obtunded or aphasic patient. Only sensory loss attributed to stroke is scored as abnormal and the examiner should test as many body areas (arms [not hands], legs, trunk, face) as needed to accurately check for hemisensory loss. A score of 2, "severe or total sensory loss," should only be given when a severe or total loss of sensation can be clearly demonstrated. Stuporous and aphasic patients will, therefore, probably score 1 or 0. The patient with brainstem stroke who has bilateral loss of sensation is scored 2. If the patient does not respond and is quadriplegic, score 2. Patients in a coma (item 1a=3) are automatically given a 2 on this item.	0 = **Normal;** no sensory loss. 1 = **Mild-to-moderate sensory loss;** patient feels pinprick is less sharp or is dull on the affected side; or there is a loss of superficial pain with pinprick, but patient is aware of being touched. 2 = **Severe to total sensory loss;** patient is not aware of being touched in the face, arm, and leg.	____
9. Best language: A great deal of information about comprehension will be obtained during the preceding sections of the examination. For this scale item, the patient is asked to describe what is happening in the attached picture, to name the items on the attached naming sheet and to read from the attached list of sentences. Comprehension is judged from responses here, as well as to all of the commands in the preceding general neurological exam. If visual loss interferes with the tests, ask the patient to identify objects placed in the hand, repeat, and produce speech. The intubated patient should be asked to write. The patient in a coma (item 1a=3) will automatically score 3 on this item. The examiner must choose a score for the patient with stupor or limited cooperation, but a score of 3 should be used only if the patient is mute and follows no one-step commands.	0 = **No aphasia;** normal. 1 = **Mild-to-moderate aphasia;** some obvious loss of fluency or facility of comprehension, without significant limitation on ideas expressed or form of expression. Reduction of speech and/or comprehension, however, makes conversation about provided materials difficult or impossible. For example, in conversation about provided materials, examiner can identify picture or naming card content from patient's response. 2 = **Severe aphasia;** all communication is through fragmentary expression; great need for inference, questioning, and guessing by the listener. Range of information that can be exchanged is limited; listener carries burden of communication. Examiner cannot identify materials provided from patient response. 3 = **Mute, global aphasia;** no usable speech or auditory comprehension.	____
10. Dysarthria: If patient is thought to be normal, an adequate sample of speech must be obtained by asking patient to read or	0 = **Normal.**	____

repeat words from the attached list. If the patient has severe aphasia, the clarity of articulation of spontaneous speech can be rated. Only if the patient is intubated or has other physical barriers to producing speech, the examiner should record the score as untestable (UN), and clearly write an explanation for this choice. Do not tell the patient why he or she is being tested.	1 = **Mild-to-moderate dysarthria;** patient slurs at least some words and, at worst, can be understood with some difficulty. 2 = **Severe dysarthria;** patient's speech is so slurred as to be unintelligible in the absence of or out of proportion to any dysphasia, or is mute/anarthric. UN = **Intubated** or other physical barrier, explain:_____	
11. Extinction and inattention (formerly neglect): Sufficient information to identify neglect may be obtained during the prior testing. If the patient has a severe visual loss preventing visual double simultaneous stimulation, and the cutaneous stimuli are normal, the score is normal. If the patient has aphasia but does appear to attend to both sides, the score is normal. The presence of visual spatial neglect or anosognosia may also be taken as evidence of abnormality. Since the abnormality is scored only if present, the item is never untestable.	0 = **No abnormality.** 1 = **Visual, tactile, auditory, spatial, or personal inattention** or extinction to bilateral simultaneous stimulation in one of the sensory modalities. 2 = **Profound hemi-inattention or extinction to more than one modality;** does not recognize own hand or orients to only one side of space.	____
		____

Adapted from: Goldstein LB, Samsa GP. Reliability of the National Institutes of Health Stroke Scale. Extension to non-neurologists in the context of a clinical trial. Stroke 1997; 28:307.

Graphic 61698 Version 8.0

Classic lacunar stroke syndromes

Lacunar syndrome	Location	Clinical findings	Positive predictive value
Pure motor hemiparesis	Internal capsule, corona radiata, basal pons, medial medulla	Unilateral paralysis of face, arm and leg, no sensory signs, dysarthria and dysphagia may be present	52-85 percent
Pure sensory syndrome	Thalamus, pontine tegmentum, corona radiata	Unilateral numbness of face, arm and leg without motor deficit	95-100 percent
Ataxic hemiparesis	Internal capsule-corona radiata, basal pons, thalamus	Unilateral weakness and limb ataxia	59-95 percent
Sensorimotor syndrome	Thalamocapsular, maybe basal pons or lateral medulla	Hemiparesis or hemiplegia of face, arm and leg with ipsilateral sensory impairment	51-87 percent
Dysarthria-clumsy hand syndrome	Basal pons, internal capsule, corona radiata	Unilateral facial weakness, dysarthria and dysphagia, with mild hand weakness and clumsiness	About 96 percent

Graphic 67341 Version 4.0