

Tintinalli's Emergency Medicine: A Comprehensive Study Guide, 8e >

Chapter 172: Acute Peripheral Neurologic Disorders

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INTRODUCTION

Acute peripheral neurologic lesions are a diverse group of disorders. By definition, they involve injury or disease in sensory and motor fibers outside of the central nervous system (CNS) extending to the neuromuscular junction. The peripheral nervous system (PNS) serves sensory, motor, and autonomic functions. Thus, the patient with a peripheral nerve lesion may have deficits in any combination of these functions.

Exclude central processes, such as stroke or spinal cord injury, before considering an acute peripheral lesion.

DISTINGUISHING CENTRAL AND PERIPHERAL LESIONS

Use CNS and PNS neuroanatomy principles to distinguish lesions. Peripheral nerves contain varying amounts of motor, sensory, and autonomic fibers and follow well-described paths that make them prone to typical injuries. Thus, peripheral nerve lesions are more likely to be confined to one limb and to present with the involvement of multiple sensory modalities and motor symptoms. A typical example would be a nerve compression syndrome presenting with weakness, numbness, and tingling that developed after the arm was held in an unusual position for a prolonged period. However, weakness and numbness can be seen in both peripheral and central disorders. Hyporeflexia sometimes occurs with acute central lesions, but hyperreflexia and spasticity invariably develop with time. PNS disorders, like CNS diseases, can affect bulbar structures, resulting in diplopia, dysarthria, or dysphagia. Despite the overlap, CNS disorders have other features not seen in peripheral disease. For example, aphasia, apraxia, and vision loss are hallmarks of cortical disease. Most CNS lesions will result in upper motor neuron signs: hyperreflexia, hypertonia (spasticity), and extensor plantar (Babinski) reflexes. Perhaps the most important distinguishing component is the examination of deep tendon reflexes. Dorsiflexion of the great toe with fanning of remaining toes and flexion of the leg is a pathologic Babinski's sign, indicating a central disruption of the pyramidal tract. Although there can be many similarities between patients with CNS and PNS lesions, the distinctions are clear ([Table 172–1](#)). Lateralization of weakness, hyperreflexia, positive Babinski's sign, or any other CNS finding requires further investigation for a central rather than peripheral disorder.

Table 172–1

Differentiating Central Nervous System from Peripheral Nervous System Disorders

	Central	Peripheral
History	Cognitive changes Sudden weakness Nausea, vomiting Headache	Weakness confined to one limb Weakness with associated pain Posture- or movement-dependent pain Weakness after prolonged period in one position
Physical Examination		
Reflexes	Brisk reflexes (hyperreflexia) Babinski's sign Hoffman's sign	Hypoactive reflexes Areflexia
Motor	Asymmetric weakness of ipsilateral upper and lower extremity Facial droop Slurred speech	Symmetric proximal weakness
Sensory	Asymmetric sensory loss in ipsilateral upper and lower extremity	Reproduction of symptoms with movement (compressive neuropathy) All sensory modalities involved
Coordination	Discoordination without weakness	Loss of proprioception

LOCALIZING PERIPHERAL NERVE LESIONS

Patients with peripheral nerve disorders frequently require diagnostic evaluation unavailable during the initial ED encounter. A careful history and physical examination will exclude critical diagnoses and point toward the appropriate management.

HISTORY

The elements required to localize the process will usually include the following: symmetry; proximal versus distal symptoms; sensory, motor, or autonomic involvement; and mono- versus polyneuropathy. Sensory symptoms may include numbness, tingling, dysesthesias, pain, or ataxia. Motor symptoms manifest as weakness. Autonomic disability may present as orthostasis, bowel or bladder dysfunction, gastroparesis, or sexual dysfunction. Associated findings will frequently narrow the differential diagnosis or suggest a specific diagnosis. For example, in the setting of a recent viral infection or vaccination, consider Guillain-Barré syndrome. Is there a history of diabetes or subacute trauma? Does the patient with thrush or wasting and undiagnosed immunodeficiency have a human immunodeficiency virus (HIV)-associated neuropathy? Patients with an attached tick may have tick paralysis.

PHYSICAL EXAMINATION

Perform a comprehensive neurologic examination on patients with peripheral nerve disorders, paying particular attention to the specific area and distribution involved. Evaluate for hypotonia, muscle wasting, fasciculations, and hyporeflexia. Is there focal weakness? What is the perception of light touch, vibration, and position sense? If there are sensory deficits, what spinal nerve levels do they involve?

DIAGNOSIS

Many patients with peripheral nerve disorders will require electromyography and nerve conduction velocity studies. Electromyography is used to differentiate primary muscular or neuromuscular junction problems from peripheral nerve disorders. Nerve conduction velocity studies, which measure the speed of conduction by observing the response to nerve stimulation by distally placed electrodes, can differentiate between axonal loss and demyelination. Combined with the history and physical examination, these tests allow the consultant to better localize the lesion and make a firm diagnosis. Lumbar puncture and cerebrospinal fluid (CSF) analysis are frequently required to confirm the diagnosis in acute or subacute inflammatory and infectious processes. In certain cases, nerve biopsy may be required. In cases where biopsy is considered, MRI can direct the biopsy site to the area of greatest utility.

TREATMENT

Management of PNS disorders depends on the specific diagnosis. However, a few general principles apply. Initiate supportive care for severe, life-threatening neuromuscular diseases in the ED. Monitor patients with the potential for respiratory failure, aspiration, and cardiac dysrhythmias. **For patients at risk for diaphragmatic failure, measure baseline forced vital capacity or negative inspiratory pressure in the ED to assess whether there is an immediate need for respiratory support or admission to an intensive care unit.** Admit patients with acute peripheral neurologic conditions if there is potential respiratory or autonomic compromise, or with severe or rapidly progressing weakness. If a peripheral disorder is suspected and the patient does not require admission, arrange for neurologic follow-up within 7 to 10 days.

ACUTE PERIPHERAL NEUROPATHIES

GUILLAIN-BARRÉ SYNDROME

Guillain-Barré syndrome is an acute polyneuropathy characterized by immune-mediated peripheral nerve myelin sheath or axon destruction. The prevailing theory is that antibodies directed against myelin sheath and axons of peripheral nerves are formed in response to a preceding viral or bacterial illness. Symptoms are at their worst in 2 to 4 weeks, and recovery can vary from weeks to a year.

Clinical Features

The classic Guillain-Barré syndrome is a demyelinating disorder of the peripheral nerves. It is sometimes preceded by a viral or bacterial illness. Common infectious precipitants include *Campylobacter jejuni*, Zika virus, Cytomegalovirus, Epstein-Barr virus, or *Mycoplasma pneumoniae*. The cause is unknown but thought to be associated with antiganglioside antibodies. Signs and symptoms of the classic form include an **ascending symmetric weakness or paralysis and areflexia or hyporeflexia**. Paralysis may ascend to the diaphragm, compromising respiratory function and requiring mechanical ventilation. Autonomic dysfunction may be present as well. There are many variants of Guillain-Barré syndrome. The demyelinating form (lymphocytic infiltration of the myelin sheath of peripheral nerves) is common in the US and the axonal form (motor paralysis with sensory function intact) in Asia. The Miller-Fisher syndrome variant is characterized by ophthalmoplegia, ataxia, and areflexia. Severity of symptoms and duration of illness depend upon the form of the disease.

Diagnosis

The diagnosis is mostly historical, but lumbar puncture and electrodiagnostic information can improve confidence in the diagnosis. [Table 172–2](#) lists the specific diagnostic criteria.

Table 172–2

Diagnostic Criteria for Classic Guillain-Barré Syndrome**Required**

- Progressive weakness of more than one limb
- Areflexia

Suggestive

- Progression over days to weeks
- Recovery beginning 2–4 weeks after cessation of progression
- Relative symmetry of symptoms
- Mild sensory signs and symptoms
- Cranial nerve involvement (Bell's palsy, dysphagia, dysarthria, ophthalmoplegia)
- Autonomic dysfunction (tachycardia, bradycardia, dysrhythmias, wide variations in blood pressure, postural hypotension, urinary retention, constipation, facial flushing, anhidrosis, hypersalivation)
- Absence of fever at onset
- Cytoalbuminologic dissociation of cerebrospinal fluid (high protein and low white cell count)
- Typical findings on electromyogram and nerve conduction studies

CSF analysis shows high protein levels (>45 milligrams/dL) and WBC counts typically <10 cells/mm³, with predominantly mononuclear cells. When there are >100 cells/mm³, other considerations include HIV, Lyme disease, syphilis, sarcoidosis, tuberculous or bacterial meningitis, leukemic infiltration, or CNS vasculitis. Electrodiagnostic testing demonstrates demyelination. Nerve biopsy reveals a mononuclear inflammatory infiltrate. If MRI is performed to rule out alternative diagnoses, it will show enhancement of affected nerves.

Treatment

The first step in management is assessment of respiratory function. Airway protection in advance of respiratory compromise decreases the incidence of aspiration and other complications. A well-established monitoring parameter is vital capacity, with normal values ranging from 60 to 70 mL/kg. A simple bedside assessment of respiratory status is obtained by trending values reached when the patient counts from 1 to 25 with a

single breath. **Avoid depolarizing neuromuscular blockers like succinylcholine for intubation in Guillain-Barré syndrome due to the risk of a hyperkalemic response.**

Both IV immunoglobulin and plasmapheresis shorten the time to recovery. Neither has been shown to be superior to the other, nor are they more efficacious when used together. There are adverse effects seen with both modalities of treatment. IV immunoglobulin has been associated with thromboembolism and aseptic meningitis; plasmapheresis is associated with hemodynamic instability, but a lower rate of relapse. In general, IV immunoglobulin is more widely available and less cumbersome to administer. Corticosteroids are of no benefit and may be harmful.¹

Disposition

Admit patients with acute Guillain-Barré syndrome to a unit where cardiac, respiratory, and neurologic functions can be monitored. Even if a patient does not initially meet the criteria for intubation, intensive care unit admission may still be indicated in order to avoid sudden, unmonitored respiratory failure (**Table 172–3**).²

Table 172–3

Managing Respiratory Failure in Guillain-Barré Syndrome

Indications for intubation

Vital capacity <15 mL/kg

Declining one breath count

Pao₂<70 mm Hg on room air

Bulbar dysfunction (difficulty with breathing, swallowing, or speech)

Aspiration

Indications for admission to intensive care unit

Patients with 4 or more of the following findings: inability to stand, inability to lift the head, inability to lift the elbows, insufficient cough, time from symptom onset to hospital admission <7 days or elevated liver enzymes

Autonomic dysfunction

Bulbar dysfunction

Initial vital capacity <20 mL /kg

Initial negative inspiratory force less than –30 cm of water

Decrease of >30% of vital capacity or negative inspiratory force

Inability to ambulate

Treatment with plasmapheresis

Abbreviation: Pao₂ = partial pressure of arterial [oxygen](#).

BELL'S PALSY AND UNILATERAL FACIAL PARALYSIS

Bell's palsy or idiopathic facial nerve palsy is the most common cause of unilateral facial paralysis. There are 23 to 25 cases per 100,000 annually, affecting men and women equally. Cranial nerve VII, the facial nerve, supplies motor innervation to the muscles of expression of the face and scalp, the stapedius muscle, and taste to the anterior two thirds of the tongue. The cause of Bell's palsy is not clear. Herpes simplex virus DNA and

antigens have been discovered around the facial nerve of affected patients. However, antiviral medications are ineffective, casting doubt on the once predominant theory that herpes simplex virus is responsible for the disease.

Clinical Features

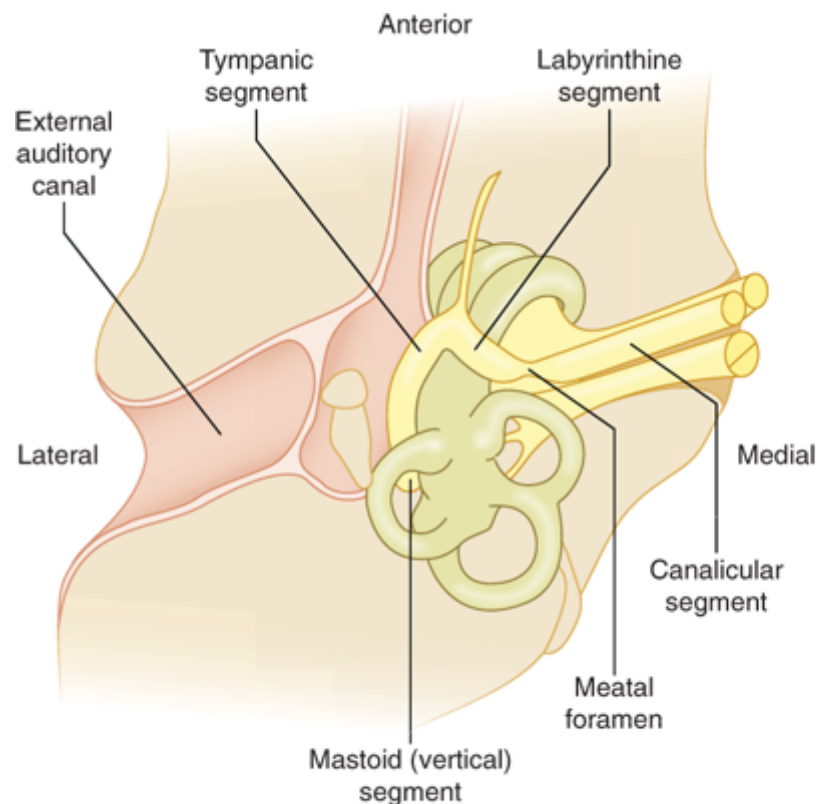
Idiopathic Bell's palsy may be preceded by pain around or behind the ear. Onset of facial paralysis is acute, with maximal symptoms in 2 to 3 days. Facial numbness or hyperesthesia can accompany paralysis. Subtle dysfunction of cranial nerves V, VIII, IX, and X may be associated; for example, patients may also complain of decreased taste or hyperacusis due to paralysis of the stapedius muscle. On exam, patients will have facial droop, effacement of wrinkles and forehead burrows, and inability to completely close the eye. Recurrent idiopathic Bell's palsy occurs in a small number of patients.

Diagnosis

Diagnosis of idiopathic Bell's palsy is based on the history and physical exam and is a diagnosis of exclusion of other conditions that can cause facial palsy. **The most important alternative diagnoses to exclude are ear infections and stroke.** Always perform an ear examination to identify otitis media and malignant otitis, and palpate the mastoids for tenderness, because infections of the ear and mastoids can affect the mastoid, tympanic, labyrinthine, or meatal segments of the cranial nerve VII ([Figure 172–1](#)).

FIGURE 172–1.

Relationships of cranial nerve VII to the inner and middle ear. [Adapted with permission from Lalwani AK (ed): *Current Diagnosis & Treatment in Otolaryngology-Head & Neck Surgery*, 3rd ed. McGraw-Hill, Inc., 2012. Sect XVI Facial Nerve [Chapter 70](#). [Figure 70–3](#), Part A.]



Source: J.E. Tintinalli, J.S. Stapczynski, O.J. Ma, D.M. Yealy, G.D. Meckler, D.M. Cline:
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When a central process causes facial paralysis, the forehead will be spared, as the forehead is supplied by cranial nerve VII arising near the pontomedullary junction, with crossed innervation. Peripheral facial nerve palsies will manifest as weakness throughout the facial nerve distribution, including the forehead. **Middle cerebral artery ischemia or stroke** consists of hemiparesis, facial plegia sparing the forehead, and sensory loss all contralateral to the affected cortex. **Rarely, a brainstem stroke may mimic Bell's palsy if the stroke affects the area where the facial nerve wraps around the abducens (cranial nerve VI) nucleus.** Brainstem stroke findings include peripheral facial nerve palsy and ipsilateral gaze palsy due to ischemia of the abducens nucleus. So, **test extraocular muscle function for all patients suspected of having Bell's palsy. Any patient with facial paralysis sparing the forehead or inability to abduct an eye should undergo neuroimaging to assess for stroke.** No imaging or laboratory testing is needed in patients with high suspicion of Bell's palsy unless further studies are required to exclude alternative diagnoses.

A number of viral infections and other disorders have been associated with Bell's palsy (e.g., mumps, Epstein-Barr virus, rubella, Lyme disease; see later discussion) but are suspected by associated signs and symptoms.

Treatment and Disposition

Treatment with corticosteroids increases the frequency of complete recovery.³ The dose of [prednisone](#) is 1 milligram/kg per day PO for 7 days. There is no benefit from antiviral medications, either alone or in addition to steroid therapy.⁴ There is insufficient evidence to support acute surgical decompression. The greatest risk to patients with facial paralysis is corneal abrasions and keratitis due to eyelid weakness and incomplete eye closure. Patch the affected eye if there is incomplete closure, and provide instructions for ophthalmology follow-up to determine when the patch can be removed. Make sure there is no lid movement under the eye patch. Provide patients with ocular lubricants, including a tear replacement for daytime and a more viscous topical lubricant for sleep.

Most patients begin to recover within 3 weeks, but about 15% will have permanent paralysis. Ensure follow-up within 7 days with a primary care physician or ear, nose, and throat specialist.

RAMSEY HUNT SYNDROME (HERPES ZOSTER OTICUS)

Ramsey Hunt syndrome is a herpes zoster infection of the geniculate ganglion. Signs and symptoms include unilateral facial nerve palsy, severe pain, and a vesicular eruption on the face. Ramsey Hunt syndrome may be indistinguishable from Bell's palsy if paralysis precedes the vesicular eruption. Cranial nerve VIII may also be involved with associated vertigo, nausea, and hearing loss. As opposed to classic Bell's palsy, when active herpes zoster is suspected, treatment is with both steroids ([prednisone](#) 1 milligram/kg per day PO for 7 days) and antivirals ([famciclovir](#) 500 mg PO three times a day for 7 days or [valacyclovir](#) 1 gram PO three times a day for 7 days).⁵

ACUTE NEUROPATHIES OF LYME DISEASE

Lyme disease may cause a broad range of nervous system disease, both peripheral and central, acute and chronic. Consider Lyme disease as the causative agent in patients who have facial palsy with erythema migrans, tick bite, or arthritis. Some patients will have bilateral facial palsy. Multifocal Lyme polyradiculopathy may occur in the acute phase of Lyme infection. Signs and symptoms include burning and painful neuropathy, plexopathy, and other mononeuropathies.

Diagnosis is based on history of tick bite and a physical examination that uncovers an attached tick or erythema chronicum migrans rash (see [Figure 251–18](#)). If CSF is obtained, it will show the mononuclear pleocytosis typical of Lyme infection. Treatment is with [doxycycline](#) 100 milligrams twice daily for 14 to 21 days. Facial nerve palsies in Lyme disease represent the secondary stage of illness and require 1 month of treatment with [doxycycline](#) (100 milligrams PO twice a day) or [amoxicillin](#), [cefuroxime](#), [ceftriaxone](#), or [azithromycin](#).

FOCAL MONONEUROPATHIES

Focal mononeuropathies are most likely due to focal nerve compression, although systemic processes may also lead to mononeuropathy. Diabetes is the most common systemic cause of noncompressive focal neuropathy. Focal mononeuropathy can occur at any point of compression along the course of a peripheral nerve, but most commonly occurs along the ulnar, median, radial, lateral femoral cutaneous, and peroneal nerves.

MEDIAN MONONEUROPATHY (CARPAL TUNNEL SYNDROME)

The most common form of any focal mononeuropathy is median mononeuropathy, or carpal tunnel syndrome. Carpal tunnel syndrome results from compression of the median nerve at the wrist where it traverses the carpal tunnel. The boundaries of the carpal tunnel are the carpal bones and the flexor retinaculum. The most common etiology is injury due to repetitive use, but other causes include diabetes mellitus, pregnancy, amyloidosis, obesity, renal failure, rheumatoid arthritis, hypothyroidism, trauma, and edema.

Clinical Features

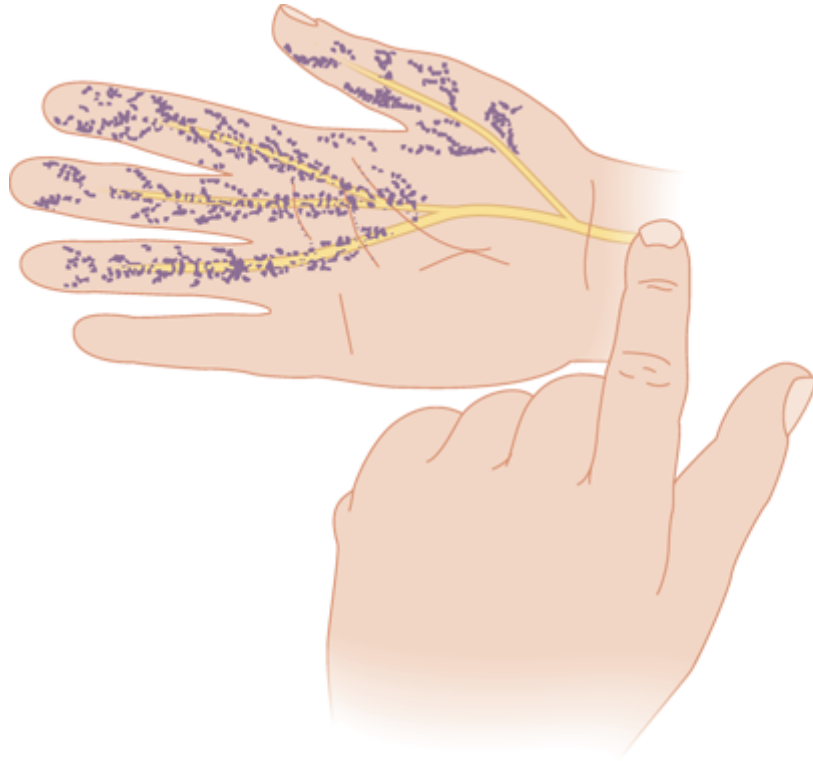
The classic signs of carpal tunnel syndrome are pain, paresthesias, and numbness in the distribution of the median nerve—the palmar aspect of the thumb, index, middle, and radial aspect of the fourth finger. Patients may report symptoms in the entire hand, but careful examination will reveal preserved sensation in the fifth and ulnar fourth digits. Symptoms are worsened by extension and flexion of the wrist and are worse at night. Weakness and wasting of the muscles of the thenar eminence may be noted on exam.

Diagnosis

Provocative testing, such as Tinel's sign and Phalen's maneuvers, can confirm the diagnosis of carpal tunnel syndrome ([Figure 172–2](#)). **Tinel's sign** detects irritated nerves by percussing over the nerve and eliciting tingling. Tapping on the palmar aspect of the wrist will result in an electric shock sensation shooting into the hand when there is compression of the median nerve. **Phalen's maneuver** is positive for carpal tunnel syndrome when holding the wrists in flexion for 60 seconds evokes or worsens symptoms. Electrodiagnostic testing demonstrates slowing of nerve conduction across the carpal tunnel. Testing is typically performed on an outpatient basis to confirm the diagnosis or to aid in the decision for operative repair.

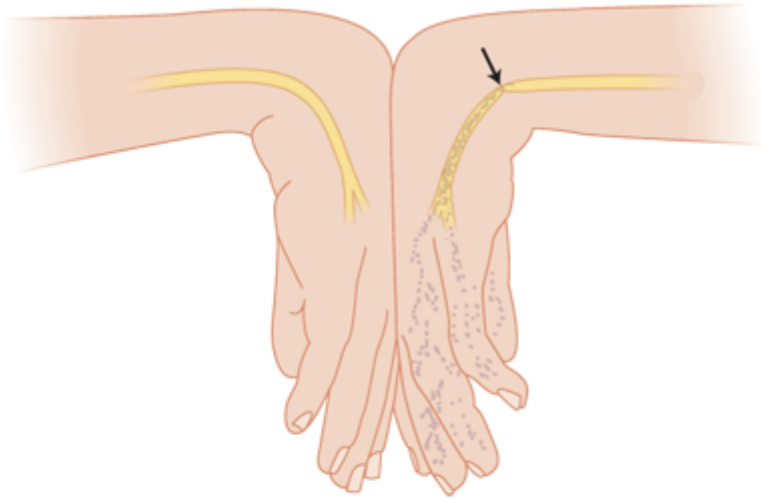
FIGURE 172–2.

A. Tinel's sign for median nerve compression. The Tinel test is performed by tapping the volar surface of the wrist over the median nerve. B. Phalen's maneuver. Phalen's maneuver is performed by compressing the opposing dorsal surfaces of the hands with the wrists flexed together as shown. This causes tingling over the median nerve distribution. [Reproduced with permission from Simon RR, Sherman SC, Koenigsknecht SJ: Wrist. In: *Emergency Orthopedics: The Extremities, 5th ed.* © 2007, McGraw-Hill Inc., New York.)



A

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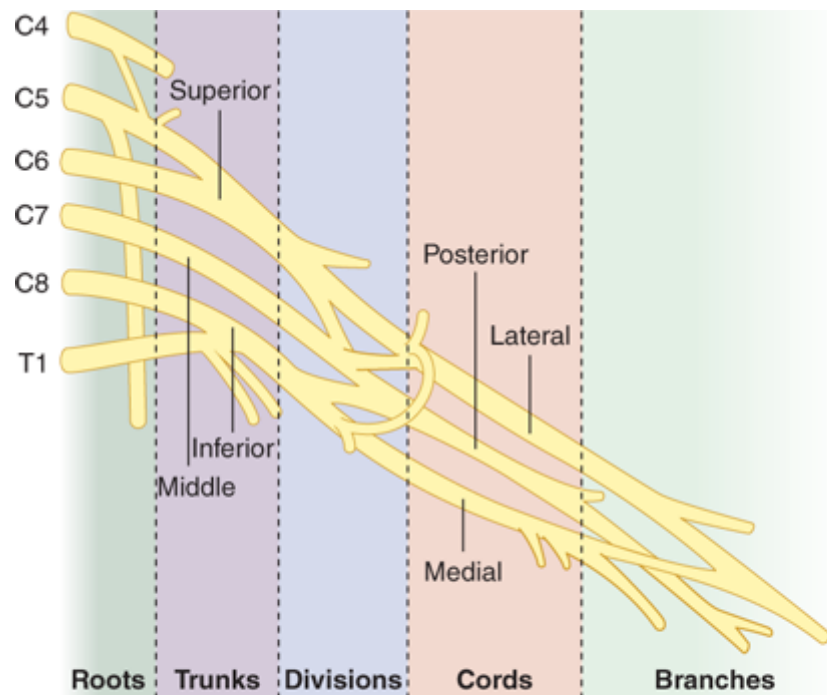
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Treatment and Disposition

The initial treatment of carpal tunnel syndrome is conservative. Encourage evaluation of workplace ergonomics. Provide a wrist splint, with the wrist in neutral position (**Figure 172-3**). Nonsteroidal anti-inflammatory drugs are often given. Refer to a hand surgeon if splinting is not effective, for steroid injection and consideration for surgical repair.

FIGURE 172-3.

The anatomy of the brachial plexus. [Reproduced with permission from Reichman EF, Tolson DR: Regional nerve blocks, in Reichman EF, Simon RR (eds): *Emergency Medicine Procedures*. New York, McGraw-Hill, 2004.]



Source: J.E. Tintinalli, J.S. Stapczynski, O.J. Ma, D.M. Yealy, G.D. Meckler, D.M. Cline:
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ULNAR MONONEUROPATHY (CUBITAL TUNNEL SYNDROME AND GUYON'S CANAL SYNDROME)

The ulnar nerve extends from the medial cord of the brachial plexus and courses through the cubital tunnel behind the medial epicondyle at the elbow before entering the forearm. At the wrist, it passes through Guyon's canal, bounded by the hamate and pisiform bones and the ligament connecting them. It supplies cutaneous innervation to the medial palm via the superficial terminal branch and to the fifth and medial fourth fingers via the deep terminal branch and innervates the palmaris brevis muscle. The ulnar nerve is most vulnerable to repetitive stress, inflammation, and trauma in the cubital tunnel and at Guyon's canal.

Cubital tunnel syndrome is the most common ulnar mononeuropathy and the second most common compressive mononeuropathy. It results from pressure on the ulnar nerve at the medial epicondyle.

Guyon's canal syndrome is caused by entrapment of the ulnar nerve in Guyon's canal of the wrist. It is also called "**handlebar palsy**," because it occurs in cyclists from prolonged compression of the wrist against the handlebars.

Clinical Features

The classic symptoms of ulnar mononeuropathy include tingling in the fifth and lateral fourth fingers. With time, numbness and weakness of the intrinsic muscles of the hand develop. In severe cases, paralysis and wasting of the intrinsic muscles of the hand may occur.

Diagnosis

Provocative testing is useful. Ulnar compression is possible if tapping on the cubital tunnel at the elbow provokes symptoms. A positive elbow flexion sign is seen when symptoms recur within 3 minutes of the elbow being held in flexion with the wrist in extension. **Froment's sign** is an inability of the thumb to oppose or put pressure against the index finger. To perform this test, ask the patient to hold a piece of paper between thumb and index finger. If you can pull the paper away or the thumb flexes at the interphalangeal joint to compensate for weakness of the adductor pollicis brevis, the test is positive. In the patient with suspected **cubital tunnel syndrome**, consider C8 entrapment and thoracic outlet syndrome. C8 entrapment can be differentiated by the presence of neck pain and worsening symptoms with neck flexion. Thoracic outlet syndrome worsens with shoulder abduction. Electrodiagnostic testing serves to precisely localize the lesion. In Guyon's canal syndrome, or handlebar palsy, compression of the ulnar nerve is more likely to spare sensory fibers that branch off before the canal. It presents as intrinsic weakness without sensory symptoms. Diagnosis of Guyon's canal syndrome is difficult, and MRI can sometimes aid in diagnosis.

Treatment and Disposition

In most cases of ulnar nerve mononeuropathy, the goal of treatment in the ED is to initiate conservative treatment and to ensure proper diagnosis and appropriate follow-up, usually with an orthopedist. Pain is not a common component of this syndrome, but anti-inflammatory medication may alleviate symptoms. Reduce repetitive use at work and home, and eliminate habits that cause pressure on the elbow or wrist.

DEEP PERONEAL NERVE ENTRAPMENT

The deep peroneal nerve may become entrapped at three locations along its course: the fibular head, anterior to the ankle joint as it passes beneath the extensor retinaculum (anterior tarsal tunnel syndrome), and distal to this point. Compression of the deep peroneal nerve may result from proximal fibular fracture or habitual crossing of the legs. Patients develop foot drop or numbness of the web between the great and second toes. Conservative treatment is recommended initially—provide a splint or brace to maintain the foot at a right angle with the leg. Prompt follow-up with a neurologist is important because nerve conduction studies should be performed to differentiate this syndrome from lumbar root or motor neuron disease.

MERALGIA PARESTHETICA

Meralgia paresthetica is entrapment of the lateral femoral cutaneous nerve in the inguinal canal. Entrapment causes numbness and pain of the anterolateral thigh. The cause can be pelvic (pregnancy, enlarging mass, aneurysm), extrapelvic (trauma, tight garment or belt, obesity), or systemic (diabetes). On examination, the patient may complain of hyperesthesia in the area of pain, and Tinel's sign may be evident when percussing over the anterior superior iliac spine. The **pelvic compression test** also supports this diagnosis: turn the patient on his or her side, compress the pelvis, and if the patient's symptoms are relieved after 30 seconds of lateral compression of the pelvis, the diagnosis is confirmed.

Conservative management with nonsteroidal anti-inflammatory drugs, weight loss, relief from tight garments, and physiotherapy is usually successful. Local injection of **lidocaine** and corticosteroid near the insertion of the inguinal ligament into the anterior superior iliac spine provides relief. Surgical decompression or nerve resection may be required for relief of symptoms.

PLEXOPATHIES

The cervical, brachial, and lumbosacral plexuses are formed by the convergence of nerve roots, which then branch into peripheral nerves. In the case of the brachial plexus, the process of mixing and dividing is repeated in the stages of trunks and cords before peripheral nerves take off. Plexopathies share many of the same causes, the most common being trauma, surgery, neoplasm, and radiation therapy. Despite this similarity, plexopathies differ significantly in diagnosis and management.

BRACHIAL PLEXOPATHY

The brachial plexus, formed by the C5-T1 nerve roots, is the most common site of plexopathy. The brachial plexus is made complex by the nerve roots merging into upper middle and lower trunks, which split and merge into lateral posterior and medial cords that split again and form five peripheral nerves (see [Figure 172–3](#)). Injury at each of these anatomic locations will result in differing symptoms. Brachial plexopathies generally manifest as weakness first, but pain and paresthesias may also develop. On examination, the patient has weakness in various distributions of the brachial plexus. The upper trunk is the more common site of involvement, affecting strength of proximal arm and shoulder musculature. Infraclavicular plexopathy due to trauma is frequently associated with injury to the axillary vessels. Causes of brachial plexopathy include trauma (penetrating trauma, humeral neck fracture, severe traction injury, or dislocation), shoulder reduction, neoplasm (Pancoast tumor), radiation, or surgery. A **"burner"** is burning pain involving one upper extremity, sometimes with numbness, paresthesias, or weakness, occurring after trauma to the neck and shoulder. It can occur in football players and other athletes during sports. A burner can be a brachial plexopathy, caused by traction to the shoulder or an injury to the cervical nerve roots caused by neck flexion-hyperextension. Brachial plexopathies can also result from

cervical rib compression, midshaft clavicular fracture, or penetrating trauma. Assessment consists of careful neurologic examination, spine imaging, and consultation with neurology, neurosurgery, or orthopedics, depending on the specific injury identified.

LUMBOSACRAL PLEXOPATHY

The L1-S4 nerve roots form the lumbosacral plexus. Causes of lumbosacral plexopathy are less likely traumatic and more likely to include radiation, diabetic amyotrophy, aortic aneurysm, retroperitoneal hemorrhage, or compression from arteriovenous malformations. The differential diagnosis also includes the **cauda equina and conus medullaris syndromes**. Lesions affecting the lumbar portion of the plexus result in weakness of hip adduction and flexion and knee extension, decreased sensation at the top and inner thigh, and decreased patellar reflexes. Lesions affecting the sacral portion of the plexus result in inability to abduct the thigh at the hip joint, weakness of hip extension and knee flexion, and decreased sensation of the back of the thigh and below the knee. Plain radiographs of the lumbar spine are useful to screen for spine compression from degenerative or neoplastic disease. Perform MRI when cord injury or compression is considered. See [chapter 279](#), Neck and Back Pain, for detailed discussion of epidural compression syndromes, cauda equina syndrome, and conus medullaris syndrome. CT of the abdomen will exclude aortic aneurysm, psoas muscle masses, and retroperitoneal hemorrhage, which also present with asymmetric lower extremity weakness. Management of plexopathies is usually directed at the underlying cause.

CERVICAL PLEXOPATHY

The cervical plexus, formed by the C1-C4 nerve roots, is the least common plexopathy. Apart from trauma or neoplasm, the condition is occasionally seen postoperatively due to positioning during surgery. Patients may have few symptoms. Neoplastic lesions may cause a deep boring and constant pain. Due to anatomic parameters, electrodiagnostic assessment is difficult and unreliable. Order a CT or MRI scan if a neoplastic process is considered. The management of cervical plexopathy is usually nonoperative.

NEUROMUSCULAR JUNCTION DISORDERS

BOTULISM

Botulism is a toxin-mediated neuromuscular junction disorder that causes acute weakness leading to respiratory failure. In 2010, the Centers for Disease Control and Prevention reported 112 cases of botulism. Of these, 76% were infantile botulism, 15% were wound botulism, and 8% were food borne.⁶ Botulism affects infants between the ages of 1 week and 11 months and has been implicated as a cause of sudden infant death syndrome. The causative organism, *Clostridium botulinum*, is an anaerobic spore-forming bacterium found in soil and honey. In suitable

environments, these spores germinate and release toxin that irreversibly binds the presynaptic membrane of peripheral and cranial nerves, inhibiting the release of **acetylcholine** at the peripheral nerve synapse. Three of the eight known toxins, types A, B, and E, produced by *C. botulinum* cause human disease. The most commonly implicated is toxin A, which is also used cosmetically in injection form. Most cases of botulism are isolated events associated with improperly preserved canned foods. In adults, botulism results from ingestion of preformed toxin. Infantile botulism occurs due to ingestion of spores that germinate in the higher pH of the infant GI tract. Botulism can result from wound infections, mostly occurring in injection drug users. In 2010, every reported case of wound botulism but one was due to injection drug use.⁷ Toxin type E is associated with preserved or fermented fish and marine mammals, and the latter are the most important sources of botulism in Alaska, Japan, Russia, and Scandinavia. *C. botulinum* is classified as a category A bioweapon by the Centers for Disease Control and Prevention, among the most lethal.⁷

Clinical Features

Onset of symptoms occurs 6 to 48 hours after the poisoning with toxin. Patients experience nausea, vomiting, abdominal cramps, and diarrhea or constipation, which may easily be misdiagnosed as an acute gastroenteritis. **Classically, botulism produces a descending, symmetric paralysis.** Since the disorder is at the neuromuscular junction, there is no sensory deficit and no pain. The muscles first affected are the cranial nerves and bulbar muscles. The patient presents with diplopia, dysarthria, and dysphagia and may report blurred vision. Deep tendon reflexes are normal or diminished. Toxin-mediated decrease in cholinergic output causes anticholinergic symptoms, such as constipation, urinary retention, dry skin and eyes, and hyperthermia. **Pupils are often dilated and nonreactive to light.** Pupillary findings provide an important point of differentiation from myasthenia gravis, which does not affect the pupil. Infantile botulism presents as constipation, poor feeding, lethargy, and weak cry; consequently, this diagnosis must be in the differential of the "floppy" infant.

Diagnosis

The diagnosis of botulism is made based on clinical findings and exclusion of other processes. The toxin can be identified in both serum and stool, but the assay is not commonly available. If the suspected food source is available, it should be tested for toxin.

Treatment and Disposition

Initial treatment is supportive. Secure the airway if there is respiratory compromise. In several retrospective cohort studies, botulinum equine antitoxin was associated with decreased number of days on the ventilator and in the hospital, and it is most effective when used early.⁷ The Centers for Disease Control and Prevention recommend giving one 10-mL vial of trivalent equine antitoxin as soon as the diagnosis is made.

Human botulism immunoglobulin has been shown to decrease mechanical ventilation requirements and length of intensive care unit and hospital stays.⁹

Patients diagnosed with botulism require admission to the intensive care unit and intubation for respiratory failure. Report suspected cases to the local health department so that further exposures can be prevented.

TICK PARALYSIS

Tick paralysis, or tick toxicosis, is an uncommon disease caused by a poorly defined neurotoxin secreted in the saliva of several tick species. It is most common in spring and summer and in the Pacific Northwest, western, and southeastern states. Most cases are in children.

Clinical Features

Initially, 4 to 7 days after tick attachment, patients experience nonspecific influenza-like symptoms including malaise and weakness. These symptoms progress to ataxia and then an **ascending weakness and paralysis** without sensory involvement. Infants and children may be listless or irritable. When the bulbar nerves and diaphragm become involved, respiratory failure ensues.

Diagnosis

Tick paralysis is diagnosed based on typical symptoms and when the culprit tick is located. **Perform a complete body search for ticks before initiating treatment for other possible diagnoses.** The most common tick attachment site is the scalp. If no tick is found, the differential diagnosis of acute ataxia and ascending flaccid paralysis without sensory involvement includes Guillain-Barré syndrome, botulism, spinal cord tumor, and poliomyelitis.

Misdiagnosis of tick paralysis can lead to failure to find and remove the tick and unnecessary therapies such as central venous access, plasma exchange, and IV immunoglobulin. Tick paralysis and Guillain-Barré syndrome are indistinguishable even with nerve conduction studies.¹⁰

Treatment and Disposition

Treatment involves complete removal of the attached tick. Remove ticks by taking hold of the very head of the tick with tweezers and applying gentle steady traction.

Admit patients with respiratory distress to the intensive care unit, and admit all patients to the hospital to ensure symptoms do not progress. Local wound care and supportive care (including intubation and mechanical ventilation if necessary) result in excellent recovery within hours to days.¹⁰

INFLAMMATORY MYOPATHIES

Although not acute peripheral neurologic lesions, polymyositis and dermatomyositis present with similar symptoms to many peripheral neuropathies. These diseases are rare inflammatory myopathies causing symmetric weakness that progresses over weeks to months.

Clinical Features

Patients develop weakness of the proximal limbs, trunk, and neck. Some experience dysphagia, myalgias, and rarely dyspnea when the respiratory muscles become involved. Sensation and reflexes are normal, and the ocular muscles are usually spared. Occasionally cardiac muscle may be involved with cardiomyopathy, heart failure, and conduction disturbances. The findings of dermatomyositis are similar with the addition of a diffuse violaceous rash often on the face and trunk.

Diagnosis

Diagnosis is mainly clinical, but certain laboratory findings are characteristic such as elevated erythrocyte sedimentation rate, elevated creatinine kinase, and leukocytosis. The differential diagnosis includes Lambert-Eaton syndrome, endocrinopathies, toxic myopathies, and infectious myopathies.

Treatment and Disposition

To prevent irreversible muscle damage, both polymyositis and dermatomyositis require treatment with systemic corticosteroids or other immunosuppressants.

Admit patients with rhabdomyolysis, respiratory distress, or profound weakness. Otherwise, patients may be discharged with neurology or rheumatology follow-up within 7 days.

SUBACUTE AND ACUTE ON CHRONIC PERIPHERAL NERVE LESIONS

Neuropathies can also be associated with an underlying medical disorder, such as HIV and diabetes.

HUMAN IMMUNODEFICIENCY VIRUS–ASSOCIATED PERIPHERAL NEUROLOGIC DISEASE

HIV infection, its complications, and its pharmacologic treatments are associated with a number of peripheral neurologic disorders. The most common of these, HIV neuropathy and antiretroviral drug–induced neuropathy, are chronic processes that do not cause sudden disability or symptoms. HIV-infected patients have a high rate of mononeuritis multiplex and an inflammatory myopathy resembling polymyositis. In late-stage acquired immunodeficiency syndrome, almost all patients manifest neuropathy. Patients in the early stages of HIV infection have greater susceptibility to Guillain-Barré syndrome. The presentation is similar to that of the non-HIV-infected patient. Treatment is the same as in non-HIV Guillain-Barré syndrome, with an emphasis on identifying patients who are likely to need intubation and mechanical ventilation early in the ED stay followed by close monitoring of respiratory function.

CYTOMEGALOVIRUS RADICULITIS

In the latter stages of acquired immunodeficiency syndrome, cytomegalovirus may acutely infect the lumbosacral nerve roots, causing a polyradiculopathy or cauda equina syndrome. Evidence of systemic cytomegalovirus infection, including retinitis, is almost always present. As opposed to the more common chronic neuropathy of HIV, patients with cytomegalovirus radiculitis become acutely weak, with primarily lower extremity involvement, and may have variable degrees of bowel and bladder dysfunction. The examination shows primarily lower extremity weakness and hyporeflexia, with decreased sensation in the lower extremities and groin. Rectal tone may be impaired. Lumbar puncture reveals a pleocytosis with predominantly polymorphonuclear cells and modestly increased protein. Viral DNA is detected by polymerase chain reaction in most patients and is highly specific. MRI of the lumbosacral spine demonstrates swelling and clumping of the cauda equina. Imaging is recommended to exclude mass lesions of the lower spine or nerve roots. Early identification is important, as antiviral treatment is effective. The treatment of cytomegalovirus radiculitis is IV [ganciclovir](#), started at 5 milligrams/kg every 12 hours for 3 to 6 weeks. Treatment can be initiated before definitive diagnosis.

DIABETIC PERIPHERAL NEUROPATHY

Diabetes mellitus remains the most common cause of noncompressive focal neuropathy. A variety of mechanisms contribute to the pathophysiology of diabetic neuropathy. Chronic hyperglycemia and glycemic variability lead to oxidative stress and neuroinflammatory processes that result in a variety of neuropathic syndromes.

Diabetic peripheral neuropathy is the term used for any neuropathy in a type 1 or type 2 diabetic patient. The most common manifestation of diabetic peripheral neuropathy is a distal symmetric polyneuropathy, but it may also lead to focal neuropathies and mononeuropathy multiplex. Diabetic peripheral neuropathy is an example of a distal axonopathy resulting in length-dependent, centripetal "dying back" of the affected nerves. This phenomenon produces the typical stocking and glove distribution of diabetic neuropathy.

Hyperglycemic neuropathy is seen in newly diagnosed diabetics and normally improves with glycemic control. **Insulin** neuritis is a reversible condition characterized by acute limb pain and paresthesias associated with rapid glycemic control with **insulin**. Painful diabetic neuropathy is usually intermittent and worse at night. It can be experienced variably as pins and needles, throbbing, burning, achy, or cramping. Duration is less than 6 months. After 6 months, the same process is termed chronic painful diabetic neuropathy. Diabetic neuropathic cachexia is a rare syndrome causing weight loss and painful dysesthesias of the limbs and trunk.

Diabetic amyotrophy is a lumbosacral plexopathy seen in patients with a long-standing history of diabetes and presents with back pain followed by weakness. Patients report the acute onset of ipsilateral back pain, followed within days by progressive leg weakness. Sensory findings are absent. The examination reveals decreased leg power in a variety of patterns reflecting impairment of plexus function with relatively symmetric sensation. There may be muscle wasting in affected limbs in long-standing disease. Deep tendon reflexes may be diminished on the affected side. Bowel and bladder functions are not affected.

There is a clear relationship between glycemic control and neuropathy. The Diabetes Control and Complications Trial demonstrated a 60% reduction in risk of developing neuropathy with tight glycemic control, which persisted for 8 years.¹¹ Emphasize strict foot care to the diabetic who has already developed neuropathy. Neuropathy-associated anesthesia may result in inadvertent trauma and subsequent development of ulcers, cellulitis, fasciitis, osteomyelitis, and eventual amputation. Symptomatic management is an ED patient's most immediate concern. Nonsteroidal anti-inflammatory drugs are ineffective in treating neuropathic pain and are relatively contraindicated in the diabetic patient due to their renal and cardiac effects. Narcotics carry addictive potential. Tricyclic antidepressants, anticonvulsants, and topical capsaicin all have proven beneficial (**Table 172–4**). Best evidence supports the use of **duloxetine**, **venlafaxine**, **amitriptyline**, **pregabalin**, **gabapentin**, **valproate**, **tramadol**, or **oxycodone**.¹²

Table 172–4

Medications Used in the Treatment of Painful Diabetic Neuropathy

Class	Starting Dose	Notes
Anticonvulsants	Pregabalin , 50 milligrams three times daily Gabapentin , 300 milligrams at bedtime	May cause somnolence, dizziness, weight gain Similar adverse effects to pregabalin
Antidepressants	Duloxetine , 30 milligrams daily Desipramine , 10–25 milligrams before bedtime	Consider with concomitant depression Tricyclic antidepressant with fewest side effects
Topical therapy	Capsaicin, 0.075% up to four times daily Lidocaine , 5% two to four patches daily	Apply using gloved hand Cut in half to save cost
Opioids	Oxycodone CR ®, 10 milligrams twice daily Tramadol , 50 milligrams twice daily	May cause constipation, somnolence Avoid in patients with seizure or on selective serotonin reuptake inhibitor

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USEFUL WEB RESOURCES

13. Nerve Whiz iOS app : <http://itunes.apple.com/us/app/nerve-whiz/id380714187?mt?8>

14. National Institute of Neurological Disorders and Stroke peripheral neuropathy information page — <http://www.ninds.nih.gov/une.idm.oclc.org/disorders/peripheralneuropathy/peripheralneuropathy.htm>

15. Washington University Neuromuscular Disease Center — <http://neuromuscular.wustl.edu/>

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