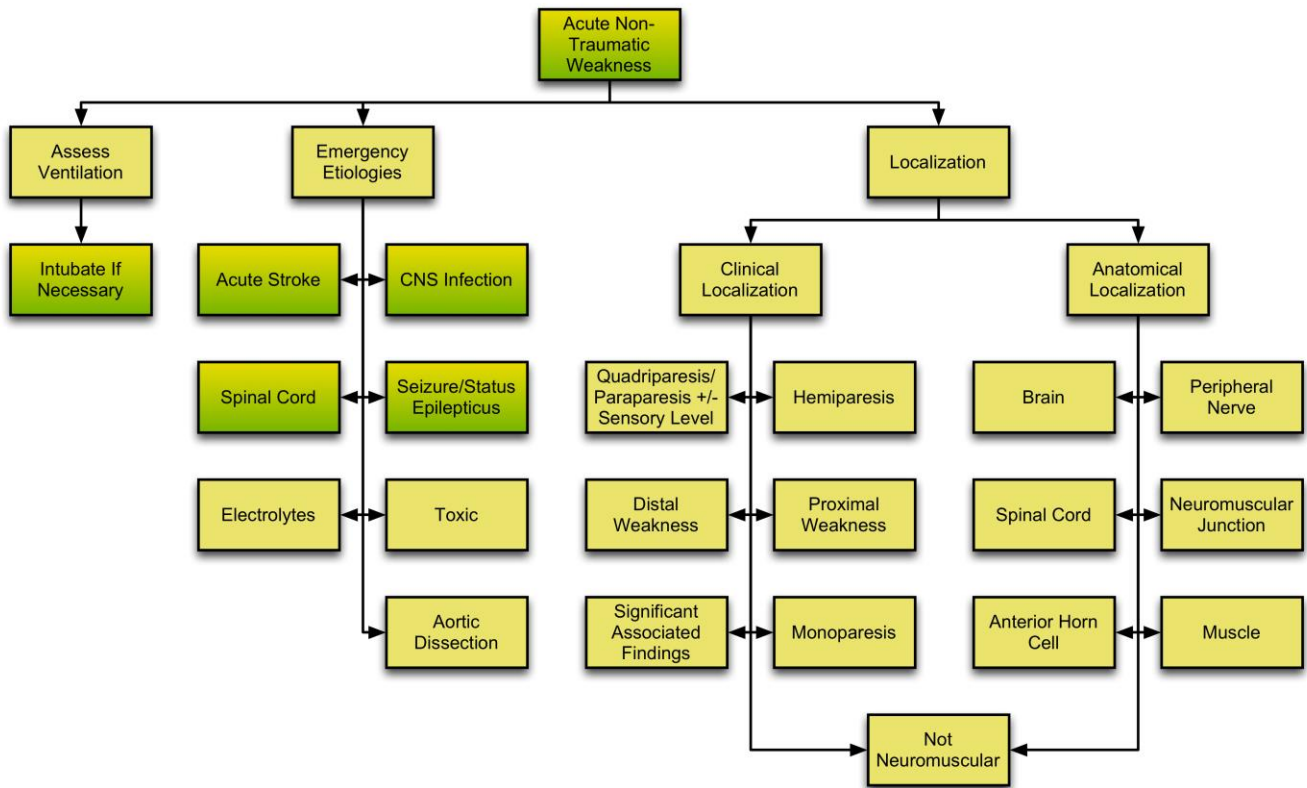


# Acute Non-Traumatic Weakness

## Emergency Neurological Life Support

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## Checklist

- Assess airway, breathing, and circulation
- Characterize the weakness by detailed exam
- Build an initial differential diagnosis of the cause of weakness
- Consider emergency causes
- Initial labs: Glucose, electrolytes, Ca, Mg, PO<sub>4</sub>, BUN/Cr, LFTs, PT, PTT, and CBC
- Special labs: TFTs, CPK, ESR
- Relevant imaging

## Communication

- Cause of weakness if known; differential diagnosis if not known
- Airway status and any respiratory issues
- Salient history and exam findings
- Relevant labs and imaging (if done)
- Treatments provided



## Acute Stroke

### Within the time window?

If the patient has signs and symptoms consistent with acute stroke, especially if the patient is within the time window for thrombolysis or endovascular therapy, see the emergency evaluation of [Acute Stroke](#).

 Flowchart

## Acute Weakness

### Patients presenting with any form of new weakness

This topic provides an organized approach to the patient with new weakness not associated with or caused by trauma. If the patient has experienced trauma follow the links to the ENLS protocols [Traumatic Brain Injury](#) and [Traumatic Spine Injury](#) as appropriate.

Based on the pattern of weakness, one can decide the degree of urgency for airway and ventilatory support and administration of time-sensitive treatments.

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## Anatomical Localization

### Based on the location of nervous system pathology

Understanding the cause of weakness can be aided by localizing anatomically, since diseases are often specific for each anatomic region. The neurological examination helps localize the etiology of the weakness. This protocol breaks down anatomic regions into brain, spinal cord, anterior horn cell (alpha motoneuron), peripheral nerve, neuromuscular junction (NMJ), and muscle.

Diseases of the brain and spinal cord (central nervous system) produce "upper motor neuron (UMN) weakness," meaning disruption of descending motor axons or cell bodies that innervate the lower motor neurons (LMN- the anterior horn cell, peripheral nerve, and NMJ). However, in the acute phase of new weakness, an UMN lesion may resemble a LMN lesion.

After performing a neurological examination, refer to the table below to ascribe the appropriate anatomical localization. Focus should center on the presence or the absence of sensory signs (loss of sensory modality) or symptoms (complaints of numbness or tingling). If sensory signs or symptoms are absent, a peripheral nerve process is eliminated, and a central nervous system process becomes less likely. Anterior horn cell causes are principally amyotrophic lateral sclerosis (ALS or "Lou-Gehrig's disease") and polio, neither of which have acute treatments. Examination of the reflexes is helpful in differentiating among the remaining causes. In general, lesions of the brain and spinal cord and the NMJ are the most emergent causes to consider, as there are specific treatments for some of these diseases (acute stroke and spinal cord compression) or public health concerns (botulism).

Table: These are usual findings for 5 localizations of weakness

Localization	Sensory symptoms/signs	Reflexes	Pattern of weakness
Brain/spinal cord	sometimes	increased	distal > proximal
Motoneuron	never	increased in ALS; decreased in polio	varies; prominent atrophy
Spinal nerve/peripheral nerve	nearly always	decreased	follows nerve innervation
Neuromuscular junction	never	unchanged	proximal
Muscle	never	unchanged	proximal

 Flowchart

## Anterior Horn Cell

### Alpha motoneuron

Pattern of Weakness:

- Proximal and distal weakness
- Fasciculations (involuntary contractions or twitching of muscle fibers) are prominent

Sensory loss: Absent

Reflexes:

- Normal, or decreased if muscle bulk is severely decreased
- Increased in ALS

Acute etiologies: polio, West Nile virus



## Brain

### Cerebral cortex, white matter or brainstem

Pattern of Weakness (UMN weakness):

- Distal weakness more than proximal
- Extensors weakness more than flexors (in upper extremity)
- Hemiparesis or single limb

Sensory Loss:

- May be present depending on whether cortex or sensory tracts are involved

Reflexes:

- Increased; however, may be decreased initially but later increase

Acute Etiologies:

- See ENLS protocols [Acute Stroke](#), [Subarachnoid Hemorrhage](#), and [Status Epilepticus](#)
- Hypertensive encephalopathy

## Clinical Localization

### Based on the presenting clinical symptoms and neurological examination

After performing the neurological examination, consider the pattern of weakness. Specifically, note whether all four limbs are weak (quadriparesis) and if there is a sensory level, or whether there is unilateral body weakness (hemiparesis), one limb weakness (monoparesis), distal extremity (hands, wrists, feet) weakness (distal weakness), or proximal muscle (axial muscles, deltoids, and hip flexors) weakness (proximal weakness)? Are there any significant associated findings?

Accurately defining the presenting complaint helps generate a focused differential diagnosis. A good clinical history is essential, as the examination may be difficult or unreliable in the obtunded or confused patient. However, it should be possible to elicit whether the deficit is unilateral or bilateral, which anatomical region is affected, and whether there is a sensory deficit. With a cooperative patient, it should also be possible to establish whether the weakness is symmetrical or asymmetrical, and whether it is affecting the proximal or distal extremities. It is important to differentiate between an upper motor neuron (UMN) and a lower motor neuron (LMN) lesion in the acute setting, although this may be difficult in some situations. In well-established UMN lesions (brain and spinal cord), hyperreflexia, increased extremity tone, and a positive Babinski sign (great toe extension with lateral plantar stimulation) are seen on exam. In comparison, LMN lesions (from the anterior horn cells to the muscles) cause a flaccid, areflexic weakness, and with time, atrophy and fasciculations. However, in the acute phase, UMN lesions may mimic a LMN lesion (flaccid paralysis, normal or reduced extremity tone), and with decreased reflexes. In acute UMN lesions there is not enough time for atrophy to be evident.



## CNS Infection

Consider meningitis or encephalitis as a cause. See ENLS protocol [Meningitis and Encephalitis](#).

## Distal Weakness

- Vasculitic neuropathy
- Toxin-induced peripheral neuropathy
- Nerve compression syndromes

Distal weakness is weakness mainly affecting the hands, wrists, and feet. It is typically caused by peripheral neuropathies that often present along with sensory symptoms. Patients have decreased grip strength and drop objects or develop gait disturbances due to foot drop. The pattern of weakness and history are of great significance.

Of the many types of peripheral neuropathy, vasculitic and toxin-induced neuropathies are the most likely to produce acute weakness. It may also be produced by local nerve compression syndromes (e.g. carpal tunnel syndrome) that predominantly affects the distal extremities, causing both sensory and motor symptoms.



## Electrolyte Disturbance

### Glucose, potassium, phosphate, sodium

Acute hypoglycemia, hypokalemia, hypophosphatemia or other electrolyte disturbances suggesting other organ dysfunction should be considered next.



## Emergency Etiologies

### **Exclude these time-sensitive emergency causes first**

There are several time-sensitive causes of acute weakness that should be excluded quickly before moving on to a more comprehensive localization of the cause of weakness. Consider each of the causes before proceeding to localization.

## Hemiparesis

### Unilateral body weakness

- See ENLS protocols [Acute Stroke](#), [Intracerebral Hemorrhage](#), or [Subarachnoid Hemorrhage](#)
- Intracranial mass
- See ENLS protocol [Meningitis and Encephalitis](#)
- Hypoglycemia/hyperglycemia
- Postictal Todd's paresis
- Hemiplegic migraine
- Brown-Sequard syndrome

Hemiparesis is partial paralysis affecting only one side of the body. Acute hemiplegia is complete paralysis of one side of the body. While acute hemiplegia is most commonly due to an ischemic stroke, other differential diagnoses must be considered because management varies. The history and demographic of the patient will narrow the diagnosis, and examination findings provide further clues. A blood glucose level and a non-contrast head computed tomography (CT) scan are part of the initial workup.



## Localizing the Cause of Acute Weakness

### This will help determine the cause

Perform a neurological examination on the patient that includes:

- Deep tendon reflexes
- Strength testing of proximal and distal extremity muscles, and compare flexor versus extensor muscle strength, noting symmetry between sides
- Judge diaphragmatic and chest wall muscle strength to determine if there is any respiratory insufficiency (single breath count from 1 to 20 [external intercostal muscles] and maximal inspiratory pressure or negative inspiratory force [diaphragm].)

After performing this focused neurological examination, determine the pattern of weakness.



## Monoparesis

### Weakness of a single limb

- See ENLS protocol [Acute Stroke](#)
- Intracranial mass
- Postictal Todd's paresis
- Nerve compression syndromes
- Diabetic lumbosacral radiculoplexus neuropathy
- Acute poliomyelitis

Monoparesis refers to paralysis of a single muscle, muscle group, or limb. Acute paralysis involving a single limb may be caused by a central or a peripheral lesion. Historical and examination factors may help to localize the lesion. For example, sudden onset right arm weakness with an associated dysphasia is most likely to result from a central lesion, whereas wrist drop in the right hand, with hypoesthesia on the back of the hand following falling asleep with the arm over the back of a chair, results from a peripheral nerve compression syndrome. Poliomyelitis is rare, but can occur in the unvaccinated.



## Muscle

Pattern of Weakness:

- Proximal

Sensory Loss:

- Absent

Reflexes:

- Normal unless muscles severely weak

Acute etiologies:

- Rhabdomyolysis

## Need for Assisted Ventilation

### Do you need to intubate this patient?

Assess the patient's airway and potential need for assisted ventilation. If any of the following general, subjective or objective findings are present, consider intubation.

#### General:

- Increasing generalized muscle weakness?
- Dysphagia?
- Dysphonia?
- Dyspnea on exertion and at rest?

#### Subjective:

- Rapid shallow breathing
- Tachycardia
- Weak cough
- Interrupted or staccato speech (gasping for air)
- Use of accessory muscles
- Abdominal paradoxical breathing
- Orthopnea (difficult or painful breathing except when erect)
- Weakness of trapezius and neck muscles: inability to lift head
- Inability to perform a single-breath count: count from 1 to 20 in single exhalation (Forced vital capacity 1.0 L is roughly equal to counting from 1 to 10)
- Cough after swallowing

#### Objective:

- Decreased level of consciousness (have a lower threshold to control the airway if the patient requires transfer or movement to unmonitored areas)
- Hypoxemia
- Vital capacity (VC) < 1 L or 20 ml/kg, or 50% drop in VC in a day
- Maximum inspiratory pressure > -30 cm H<sub>2</sub>O
- Maximum expiratory pressure < 40 cm H<sub>2</sub>O
- Nocturnal desaturation
- Hypercarbia (a late finding)



## Neuromuscular Junction

### Pattern of Weakness:

- First in eye muscles, neck extensors, pharynx, and diaphragm
- Followed by more generalized weakness

Sensory Loss: Absent

### Reflexes:

- Normal
- Decreased if muscle is paralyzed

### Acute Etiologies:

- Botulism
- Tick bite
- Organophosphate toxicity
- Myasthenic crisis

## Not Neuromuscular Weakness

### Consider psychiatric cause

Some disease states may produce symptoms of generalized weakness or fatigue that do not have a neuromuscular basis. These may be medical emergencies in their own right, meriting urgent specific treatment.

Consider:

- Any severe medical illness can have weakness as a symptom, but generally these will become clinically obvious during the patient's evaluation

Diagnoses of exclusion:

- Malingering
- Conversion disorder
- Anxiety disorders
- Fibromyalgia
- Chronic fatigue syndrome



## Other Urgent Causes

Consider:

- Shock
- Myocardial infarct
- Addisonian crisis



## Paraplegia from Aortic Dissection

### Spinal cord infarct

Acute aortic dissection can close the artery of Adamkiewicz that supplies the anterior spinal artery to the mid thoracic and lumbar spinal cord. The patient will have an anterior spinal artery syndrome (paraplegia with loss of pain and temperature sensation below the lesion but preservation of proprioception and light touch). Assess distal lower extremity pulses and consider CT Angiogram (CTA), ultrasound or other techniques to rule out aortic dissection if the patient has an acute anterior spinal artery syndrome.



## Peripheral Nerve

### Pattern of Weakness:

- In the distribution of the nerve, or diffusely present as stocking/glove weakness

### Sensory Loss:

- Present

### Reflexes:

- Decreased or absent

### Acute Etiologies:

- Guillain-Barré syndrome
- Vasculitis



## Proximal Weakness

- Acute myopathy
- Guillain-Barré syndrome
- Acute diabetic lumbosacral radiculoplexus neuropathy (DLRN)
- Myasthenia gravis
- Acute West Nile virus-associated paralysis
- Lambert-Eaton myasthenic syndrome (LEMS)

Proximal weakness is weakness predominantly affecting the axial muscles, deltoids, and hip flexors. Acute proximal weakness classically presents with difficulty rising from a chair or brushing hair. Patients may have difficulties flexing and extending their neck. The most common cause is myopathy. Less common causes include LEMS and myasthenia gravis. DLRN may be the presenting feature of diabetes mellitus. While poliomyelitis is very rare in western countries, it remains endemic elsewhere. West Nile virus, with similar semiology as acute poliomyelitis, is more common in the United States and Europe.

## Quadripareisis or Paraparesis with or without Sensory Level

### Suggests spinal cord lesion

Quadripareisis/paraparesis  $\pm$  sensory level

- See ENLS protocol [Spinal Cord Compression](#)
- Spinal cord infarction
- Transverse myelitis
- Generalized weakness: electrolyte and glucose abnormalities

Quadripareisis/paraparesis is symmetrical weakness of either all four limbs (quadripareisis) or legs (paraparesis), characteristically with a sensory level. Often it is related to spinal cord injury. Non-traumatic spinal cord injury may occur from compression (e.g. epidural abscess, hematoma, expanding tumor, prolapsed intervertebral disc), ischemia (spinal cord infarction), or inflammation (transverse myelitis).

In the acute phase, a flaccid paralysis below the level of cord injury and areflexia is typically seen, with an accompanying corresponding sensory level, although there is considerable variation. Neurological examination should localize the lesion in patients with acute paraparesis/plegia to the thoracic or lumbar region and quadripareisis/plegia to the cervical region. Sensory abnormalities localize in the vertical plane (cervical (C), thoracic (T), lumbar (L), or sacral). Key sensory levels to remember are T4 and T10, which correspond to the nipple and naval, respectively.

## Significant Associated Findings

### Other finding that may make the diagnosis clear

Certain constellations of symptoms and signs can make specific, often unusual diagnoses more likely. Below is a list of diagnoses and significant associated findings. Stroke syndromes also have characteristic patterns which are too numerous and varied to discuss here. However, findings such as aphasia, agnosia, apraxia, and neglect with acute weakness or sensory signs should prompt consideration of acute stroke.

Locked-in syndrome (usually due to basilar artery thrombosis; also consider residual neuromuscular blockade)

- Acute tetraplegia
- Facial muscles paralyzed except eyes
- Clear sensorium

Myasthenia gravis

- Fatigable weakness in eyelids and extra-ocular muscles
- Variable weakness elsewhere
- No sensory symptoms

Envenomation

- History of animal bite
- Descending paralysis
- Possible coagulopathy
- Rhabdomyolysis
- Shock

Hypertensive encephalopathy

- Severe, refractory hypertension
- Headache
- Transient, migratory neurological non-focal deficits

Guillain- Barré syndrome

- Ascending paralysis following upper respiratory viral illness/infection

Botulism

- Descending symmetrical paralysis
- No fever
- Clear sensorium

Organophosphate toxicity

- Weakness with prominent cholinergic signs and symptoms

Heavy metal toxicity

- Heavy metal exposure
- Prominent gastrointestinal symptoms
- Multi-organ failure

Periodic paralysis

- Episodic proximal weakness
- Family history of the same disorder

Dermatomyositis

- Heliotrope rash
- Proximal weakness

Acute intermittent porphyria

- Abdominal pain
- Proximal weakness
- Psychiatric symptoms
- Red urine

Tick paralysis

- Tick bite
- Followed by ascending paralysis

## Special Considerations for Intubation

### Consider

Special consideration for intubation:

- Rapid sequence induction/intubation is advised.
- Avoid use of succinylcholine if there is evidence of underlying progressive neuromuscular disease (precipitates acute hyperkalemia) such as Guillain-Barré, chronic neuromuscular weakness, or prolonged immobilization. Consider rocuronium as an alternative. Succinylcholine will be relatively ineffective to achieve muscle relaxation in myasthenia gravis, unless a higher dose is used (~2.5 times the standard dose). Conversely it is recommended to use half-dose of a nondepolarizing agent (rocuronium 0.5-0.6 mg/kg) in such patients because they may be more sensitive to nondepolarizing neuromuscular junction blockers.
- Consider non-invasive assisted ventilation as a temporizing measure in a neurologically stable patient expected to have a rapid resolution (e.g., myasthenia gravis).
- Prepare atropine/glycopyrolate, fluids, and vasopressors if there is evidence of autonomic instability.

See ENLS protocols [Airway, Ventilation and Sedation](#), and [Pharmacotherapy](#).



## Spinal Cord

### Non-traumatic cause

#### Pattern of Weakness:

- Acute flaccid quadriparesis/paraparesis (rarely hemiparesis) below the level of spinal cord compression
- Distal weakness more than proximal
- Extensors weaker than flexors

#### Sensory Loss:

- May be present depending on whether sensory tracts are involved; bilateral loss of sensation below a certain spinal level is diagnostic

#### Reflexes:

Elevated during acute brain insult; however, reflexes may be decreased initially and later increase

#### Acute etiologies:

- Epidural abscess, tumor

Acute spinal cord compression of non-traumatic cause should be suspected if the patient has weakness of both legs or both arms and legs with intact mental status and cranial nerves, especially if they have a history of cancer or complaint of back or neck pain. See ENLS protocol [Spinal Cord Compression](#). If there is any sign of trauma, see ENLS protocol [Traumatic Spine Injury](#).

 Flowchart

## Status Epilepticus or Seizure

### Postictal or non-convulsive status or Todd's paresis

Patients who are comatose, or encephalopathic, may be postictal. A patient with focal neurological findings (typically hemiparesis) may have a Todd's paralysis caused by a generalized seizure in a brain with a prior injury. Also, a patient may be having non-convulsive status epilepticus. See the ENLS protocol [Status Epilepticus](#).



## Toxic Cause

### **Any toxin exposure?**

Consider organophosphate or carbon monoxide exposure among others.