Emergency Neurological Life Support Acute Non-traumatic Weakness Protocol Version 5.0

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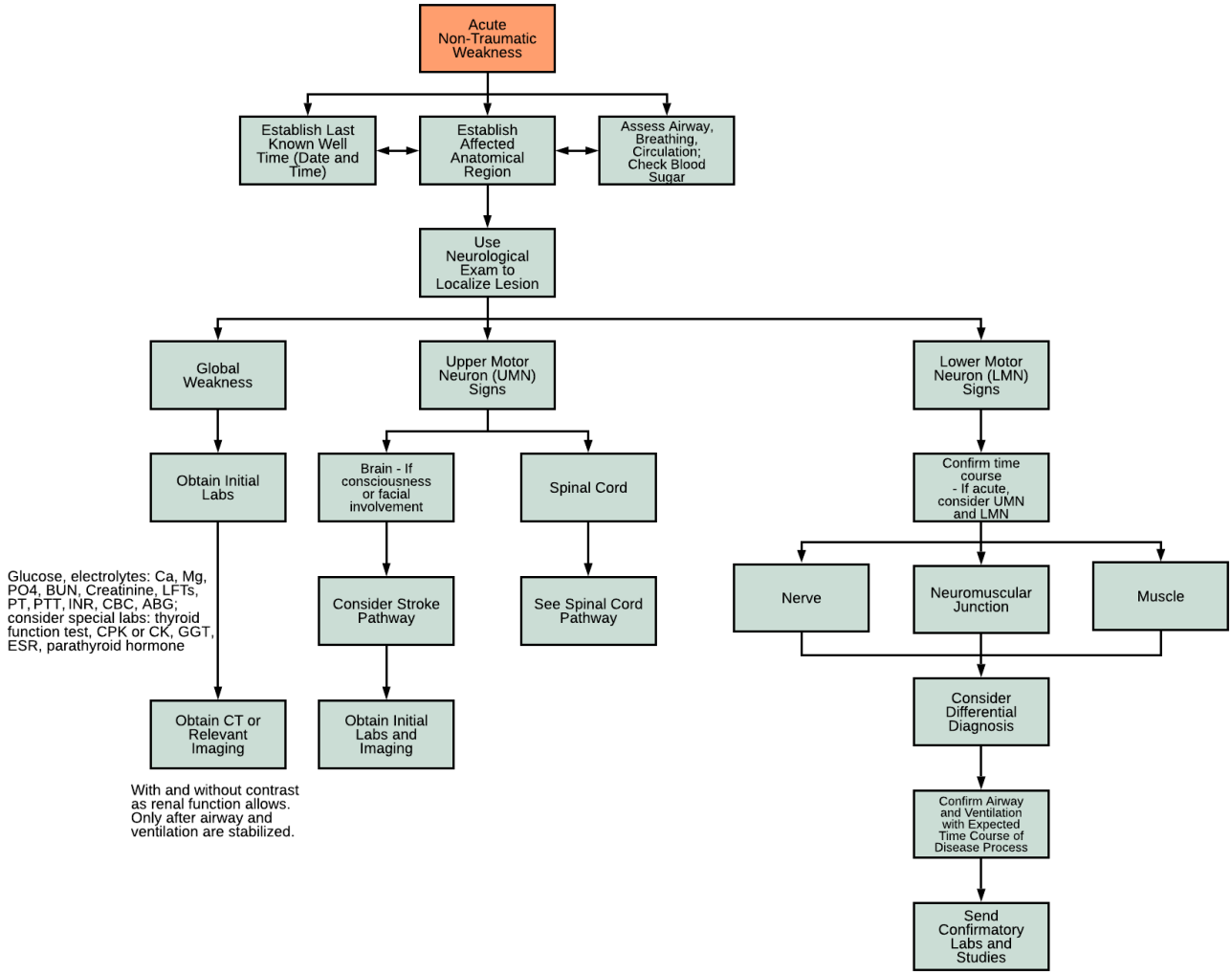
ENLS

EMERGENCY NEUROLOGICAL LIFE SUPPORT

Acute Non-Traumatic Weakness Algorithm



Checklist

☐ Assess and manage airway, breathing, and circulation
☐ Characterize the weakness by neurological exam
☐ Localize the lesion to create a differential diagnosis of the causes of weakness
☐ Initial labs: Glucose, electrolytes, Ca, Mg, PO4, BUN/Cr, LFTs, PT, PTT, CBC, and ABG
☐ Special Labs: TFTs, CPK or CK, ESR, parathyroid hormone, GGT
☐ Relevant MRI and CT imaging

Communication

☐ Salient history and exam findings
☐ Airway status and any respiratory issues
☐ Relevant labs and imaging (if done)
☐ Cause of weakness if known; differential diagnosis if not known
☐ Treatments provided
☐ Trajectory of disease process including last known well time.

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 patient's pattern of weakness, one can decide the degree of urgency for airway and ventilatory support as well as the need for administration of time-sensitive treatments such as intravenous thrombolytic. Determining the pattern of weakness and associated findings on clinical history and exam help to localize the anatomical lesion. Each anatomical lesion has different disease processes that affect that location.

Establish LKW Time (Date and Time)

If possible, asking the patient or a witness the timeline of onset, where the person is weak, if there are any sensory symptoms including numbness or tingling, and any associated symptoms is helpful. For children, a developmental and family history obtained from the parents may help in early evaluation for the first acute presentation of metabolic or neurodegenerative disorder with muscle weakness.

Establish Affected Anatomical Region

Perform a neurological examination on the patient that includes:

- Mental Status – Alterations in consciousness? Ability to speak and understand when spoken to?
- Cranial Nerves – note whether pupils and/or eye movements are normal, facial weakness, gag and cough reflexes (oropharyngeal weakness), or neck weakness.
- Motor
    - Strength testing of proximal and distal extremity muscles, and compare flexor versus extensor muscle strength, noting any asymmetry between sides
    - Watching a person's chest wall movement and use of accessory muscle can determine if there is any respiratory insufficiency. Counting from 1 to 20 on a single breath notes normal respiratory muscle strength.
- Sensory exam – determine the presence or absence of sensory signs.
- Deep tendon reflexes

Table: These are usual findings for five anatomical localizations of weakness

Localization	Sensory symptoms/signs	Reflexes	Pattern of weakness
Brain/spinal cord	sometimes	Acutely decreased then increased	distal > proximal extensors > flexors
Anterior Horn Cell	never	increased in ALS; decreased in polio	Proximal and distal; prominent atrophy and fasciculations
Peripheral nerve	nearly always	decreased	Distal > proximal, often length dependent
Neuromuscular junction	never	normal, decreased if muscle is paralyzed	proximal; first eye muscles, neck extensors, pharynx, diaphragm, followed by more generalized weakness
Muscle	never	normal, unless muscle severely weak	Proximal> distal

Assess Airway, Breathing, Circulation, Check Blood Sugar

Assess the patient's airway and potential need for assisted ventilation

Periodically assess airway and muscles of respiration, as the condition may change over time.

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)
- Failed single breath test
- Hypoxemia
- Vital capacity (VC) < 1 L or 20 ml/kg, or 50% decrease in VC in a day
- Maximum inspiratory pressure > -30 cm H_2O
- Maximum expiratory pressure < 40 cm H_2O
- Nocturnal desaturation
- Hypercarbia (a late finding)

Special Considerations for Intubation

- Rapid sequence induction/intubation is advised.
- Avoid use of succinylcholine if there is evidence of underlying progressive neuromuscular disease (precipitates acute hyperkalemia), chronic neuromuscular

weakness, or prolonged immobilization. Consider 1.0 – 1.4 mg/kg 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 non-depolarizing 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, while the diagnosis is established or with a known neuromuscular condition expected to have a rapid resolution (e.g., myasthenia gravis exacerbation).
- Prepare atropine/glycopyrrolate, fluids, and vasopressors if there is evidence of autonomic instability.

See ENLS protocols Airway, Ventilation and Sedation and Pharmacotherapy.

Blood sugar

Always assess a blood sugar level and treat hypoglycemia.

Use Neurological Exam to Localize Lesion

Assess if the patient has global weakness or an upper or lower motor neuron injury.

Exam component	Key Maneuvers and Findings
State of consciousness	Interaction with examiner Glasgow Coma Scale FOUR score
Language	Following verbal commands, naming, repeating, and reading
Cranial nerves	Follow the examiner's finger in an H pattern Ask the patient to smile Protrude the tongue Say "ah" while observing the movements of the palate and uvula In an intubated patient, in-line suction catheter and oral suction catheter can test cough and gag reflexes
Motor strength	Lift the arms and legs Pronation of the arms when held in an extended position Detailed motor strength testing of each muscle group Single breath count for muscles of respiration Neck flexion and extension
Tone	Passively move the neck, arms and legs: may be either increased or decreased (flaccid)
Reflexes	Rapidly tap on the tendon of a muscle
Sensory	Different modalities include pain/temperature, light touch, vibration, and proprioception Pain and light touch in emergencies
Coordination	Finger to nose to finger Heel to shin

Perform a neurological examination on the patient that includes:

- Mental Status
    - Alterations in consciousness: State of consciousness is easily assessed on every patient based on his or her ability to interact with the examiner. The Glasgow Coma Scale (GCS) and the FOUR score formalize these findings and add a quantitative measurement.
    - Speaking ability: Formal language testing may be limited in an emergency situation but may include the ability to follow verbal commands, naming, repeating, and reading when the situation is more stabilized.

- Cranial Nerves: The cranial nerves control the movements of the face.
    - Abnormal pupils and eye movements: Having the patient follow the examiner's finger in an H pattern tests extraocular eye movements.
    - Facial weakness: Ask the patient to smile, protrude the tongue, and say "ah" while observing the movements of the palate and uvula to test the remaining cranial nerves.
    - Poor gag and cough reflexes (oropharyngeal weakness): In an intubated patient, an in-line suction catheter and oral suction catheter can test cough and gag reflexes.
    - Neck weakness: test flexion and extension.
- Motor: Interpretation of the motor exam should account for the patient's age, capacity to understand commands, and degree of cooperation. In infants, the exam will rely primarily on observation.
    - 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) (maximal inspiratory pressure for external intercostal muscles or negative inspiratory force for diaphragm.)
- Sensory exam – determine the presence or absence of sensory signs, e.g. pain/temperature, light touch, vibration, and proprioception. In an emergency situation, light touch and pain are most useful and can be done quickly by running a soft material along the patient's skin or, if a greater degree of stimulation is required for testing, pinching the patient can be used. Care must be taken to not injure the skin by twisting or puncturing.
- Deep tendon reflexes - tested by rapidly tapping on the tendon of a muscle to watch for response.

Global Weakness

Acute generalized weakness may occur due to acute metabolic disorders including sepsis, electrolyte disturbances, anemia, or endocrine disorders.

- Marked hyperglycemia may rarely present with an acute hemiplegia, but the majority of electrolyte disorders result in symmetric involvement.
- Hypoglycemia must be excluded early, but it should be noted that most of these patients are confused or have a decreased level of consciousness.
- Other electrolyte causes that must be considered include hyponatremia and hypernatremia (Table 17), hypermagnesemia (Table 18), and hypophosphatemia (Table 19).
- Thyroid function studies may be helpful.
- Specific vitamin deficiencies can cause generalized weakness but are usually not emergent.
- Central nervous system infection (meningitis, encephalitis, encephalomyelitis) may cause weakness and is diagnosed with lumbar puncture. See ENLS Meningitis and Encephalitis module.
- A postictal patient or a patient in status epilepticus can also present with focal or generalized weakness. See ENLS Status Epilepticus module. There is typically little confusion about the diagnosis, but if there is no history available leading up to the presentation of a weak patient, the diagnosis may be more elusive.
- Acute weakness is also a prominent feature of certain organophosphate toxicity and envenomations, though the latter is exceedingly rare (see Tables 26 and 27).
- Specific drugs may cause acute weakness including slow clearance of neuromuscular blocker given during intubation.

Obtain Initial Labs

Initial: Glucose, electrolytes, Ca, Mg, PO4, BUN/Cr, LFTs, PT, PTT, CBC, and ABG
Special: TFTs, CPK or CK, ESR, parathyroid hormone, GGT

Obtain CT or Relevant Imaging

MRI or CT imaging with and without contrast may need to be emergently obtained. Depending on the likely differential diagnosis and local healthcare systems, transfer to an appropriate facility with the necessary imaging modalities and/or relative medical expertise will need to be considered at various points in this initial assessment.

Upper Motor Neuron (UMN) Signs

In well-established UMN lesions of brain or spinal cord, hyperreflexia, increased extremity tone, and a positive Babinski sign (great toe extension with lateral plantar stimulation) are seen on examination.

Note: The anterior horn of the spinal cord forms the connection between the upper and lower motor neurons. Conditions that affect these cells can cause exam findings of both UMN and LMN and spare sensory neurons. There is a limited number of disease processes that affect this area: amyotrophic lateral sclerosis (ALS) or “Lou-Gehrig’s disease”, enterovirus D68, polio, West Nile virus, and in children, acute flaccid myelitis. These are rare conditions and require expert consultation where available. Only 29 cases of polio have been reported worldwide in 2018.

Brain – if consciousness or facial movement

Brain lesions or global metabolic processes such as toxins may affect consciousness.

If the patient has signs and symptoms consistent with acute stroke such as abrupt onset of hemiparesis and is within the time window for intravenous thrombolysis or endovascular therapy, see the emergency evaluation of ENLS: Acute Ischemic Stroke.

Consider Stroke Pathway

Within the time window?

If the patient has signs and symptoms consistent with acute stroke such as abrupt onset of hemiparesis and is within the time window for intravenous thrombolysis or endovascular therapy, see the emergency evaluation of ENLS: Acute Ischemic Stroke.

Obtain Initial Labs and Imaging

Initial labs: Glucose, electrolytes, Ca, Mg, PO4, BUN/Cr, LFTs, PT, PTT, CBC, and ABG
Special labs: TFTs, CPK or CK, ESR, parathyroid hormone, GGT

Imaging: MRI or CT imaging with and without contrast may need to be emergently obtained. Depending on the likely differential diagnosis and local healthcare systems, transfer to an appropriate facility with the necessary imaging modalities and/or relative medical expertise will need to be considered at various points in this initial assessment.

Spinal Cord

Quadriparesis/Paraparesis

- Transverse myelitis (Table 15)
- Spinal cord compression
- Acute West Nile virus associated paralysis
- Spinal cord infarction (Table 16)
- Syrinx
- Drug ingestion (nitrous oxide inhalation)
- Generalized weakness: electrolyte and glucose abnormalities (Tables 9, 10, 17, 18, 19)

Quadriparesis/plegia is symmetrical weakness of all four limbs. Paraparesis/plegia is a symmetrical weakness of both lower limbs. Often, this is related to a spinal cord dysfunction. See *ENLS: Spinal Cord Compression* module.

See Spinal Cord Pathway

Quadriparesis/Paraparesis

- Transverse myelitis (Table 15)
- Spinal cord compression
- Acute West Nile virus associated paralysis
- Spinal cord infarction (Table 16)
- Syrinx
- Drug ingestion (nitrous oxide inhalation)
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Quadriparesis/plegia is symmetrical weakness of all four limbs. Paraparesis/plegia is a symmetrical weakness of both lower limbs. Often, this is related to a spinal cord dysfunction. See *ENLS: Spinal Cord Compression* module.

Lower Motor Neuron (LMN) Signs

LMN lesions (from the anterior horn cells in the spinal cord to the muscles) cause a flaccid, areflexic weakness and, with time, atrophy and fasciculations (involuntary contractions or twitching of muscle fibers).

Confirm time course – if acute, consider UMN and LMN

In the acute phase, UMN lesions may mimic a LMN lesion: flaccid paralysis, normal or reduced tone, and unreliable reflexes. There is often not enough time for atrophy to be evident, and fasciculations are rarely seen.

Nerve

Peripheral nerve syndromes can cause acute weakness. Compression including compartment syndrome is a common cause. Knowledge of the innervation of each nerve is paramount to making the diagnosis. When more than one peripheral nerve has been affected, this is called mononeuropathy multiplex and is more commonly a vasculitic immune mediated process. These conditions are rarely neurological emergencies.

Localization	Pattern of Weakness	Sensory Loss	Reflexes	Acute Etiologies
Peripheral nerve	In the distribution of the nerve, or diffusely present as stocking/glove weakness	Variable	Absent or Decreased	Guillain-Barre syndrome Vasculitic neuropathy Toxin-induced nerve compression syndromes Acute diabetic lumbosacral radiculoplexus neuropathy

Neuromuscular Junction

Localization	Pattern of Weakness	Sensory Loss	Reflexes	Acute Etiologies
Neuromuscular junction	First in eye muscles, neck extensors or flexors, pharynx, diaphragm, followed by more generalized weakness	Absent	Normal, decreased if muscle is paralyzed	Myasthenia gravis, Lambert-Eaton myesthenic syndrome, Botulism, tick bite, organophosphate toxicity

Muscle

Localization	Pattern of Weakness	Sensory Loss	Reflexes	Acute Etiologies
Muscle	Proximal	Absent	Normal unless muscle severely weak	Acute myopathy, Rhabdomyolysis, Myositis

Consider Differential Diagnosis

A detailed history and comprehensive neurological exam are important but may not be practical in the prehospital and immediate resuscitation period, and may be completed when the patient's airway and ventilation have been stabilized. However, at a minimum, obtain the patient's last known well time and perform a brief neurological exam to form a workable and realistic list of differential diagnoses. If possible, asking the patient or a witness the timeline of onset, where the person is weak, if there are any sensory symptoms including numbness or tingling, and any associated symptoms is helpful. For children, a developmental and family history obtained from the parents may help in early evaluation for the first acute presentation of metabolic or neurodegenerative disorder with muscle weakness.

Confirm Airway and Ventilation with Expected Time Course of Disease Process

Assess the patient's airway and potential need for assisted ventilation

Periodically assess airway and muscles of respiration, as the condition may change over time.

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
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- 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% decrease in VC in a day
- Maximum inspiratory pressure > -30 cm H_2O
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Special Considerations 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 1.0 – 1.4 mg/kg 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 non-depolarizing 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, while the diagnosis is established or with a known neuromuscular condition expected to have a rapid resolution (e.g., myasthenia gravis exacerbation).
- Prepare atropine/glycopyrrolate, fluids, and vasopressors if there is evidence of autonomic instability.

See ENLS protocols Airway, Ventilation and Sedation and Pharmacotherapy.

Send Confirmatory Labs and Studies

Send all relevant information.

Initial labs: Glucose, electrolytes, Ca, Mg, PO4, BUN/Cr, LFTs, PT, PTT, CBC, and ABG
Special labs: TFTs, CPK or CK, ESR, parathyroid hormone, GGT

Imaging: MRI or CT imaging with and without contrast as renal function allows may need to be emergently obtained. Depending on the likely differential diagnosis and local healthcare systems, transfer to an appropriate facility with the necessary imaging modalities and/or relative medical expertise will need to be considered at various points in this initial assessment.