EM CASES SUMMARY

Episode 156 Approach to Acute Motor Weakness

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Recognition and management of respiratory failure associated with neuromuscular disease

Tachypnea is a sign of impending respiratory compromise in the patient with neuromuscular disease

Patients with neuromuscular disease are at particularly high risk of respiratory failure, given the propensity for altered mental status and diaphragmatic and/or accessory respiratory muscle weakness. Tachypnea often presents sooner than, and may herald other signs of, impending respiratory failure.
It is prudent for the ED physician to look for the following when assessing the airway status of patient with motor power loss:

- Abnormal or poor mentation
- Difficulty with speech or weak voice
- Drooling or other indication of difficulty handling secretions
- Inability or difficulty lifting their head off the stretcher
- Weak, rapid, or shallow breaths or use of accessory muscles

Pitfall: *A common pitfall is to assume that the cause of tachypnea in a patient with suspected neuromuscular disorder with a normal oxygen saturation is due to acidemia only. Tachypnea is often a sign of impending respiratory compromise in these patients due to neuromuscular compromise that may require a definitive airway.*

When to intubate the patient with suspected neuromuscular disease: Neck flexor weakness and the "20/30/40 rule"

In addition to tachypnea, impending respiratory compromise can be heralded by decreased neck flexion strength, as it has shared innervation with the diaphragm. This can be tested by resisting neck flexion with your hand against the patient's forehead and asking them to lift their head off the bed. Normally, the neck flexors are able to overcome the examiner's hand.

The "20/30/40 rule" for indications for a definitive airway in patients with neuromuscular disease

Objective measures that help guide the decision for intubation include:

- Vital capacity (VC) less than 20cc/kg,

- Maximal inspiratory pressure (MIP) less than 30 cmH2O, and
- Maximal expiratory pressure (MEP) less than 40 cmH2O.

Succinylcholine should generally be avoided in patients with neuromuscular disease and a lower dose of rocuronium may be prudent

For RSI in patients with neuromuscular disease it is probably best to avoid using depolarizing neuromuscular blockade, such as succinylcholine. This is because the upregulation of acetylcholine receptors with use of succinylcholine leads to leakage of potassium extracellularly and in-turn may result in hyperkalemia.
It is probably safer to use a non-depolarizing paralytic, such as rocuronium. In myasthenia gravis, it is recommended to use a lower dose of rocuronium since the pathophysiology of the illness includes destruction of the acetylcholine receptors at the neuromuscular junction.

A 5 step approach to acute motor weakness

1. Does the complaint of weakness represent a true loss of motor power?

Weakness can be described as fatigue, malaise, anhedonia or decreased strength. Patients may use a myriad of synonyms to describe their symptoms such as heaviness, "lack of energy" or even numbness. It is important for us to hone-in on what the patients' symptoms truly represent. The chief complaint of "weakness", much like "dizziness" is non-specific and has many etiologies such as hypoglycemia, polypharmacy, dehydration, anemia, stroke, spinal cord trauma, and chronic neurodegenerative diseases. In order to assess the patient with true loss of motor strength, the vague term of 'weakness' should be further characterized by an assessment of power and localization of pathology. It is better to use the word "*power*" when charting or communicating about a patient's symptoms to other care providers rather than "*weakness*". When eliciting the patient's history about neuromuscular strength, inquire about their functionality. Asking what he/she/they can no longer do, or do with new difficulty, is a good start at defining the problem.

2. The geography of weakness – patterns of motor power loss

Localization of pathology leading to motor power loss is important in understanding the etiology and building a differential diagnosis.

Patterns of Motor Power Loss

Pattern	Definition	Etiology
Paraplegia	Motor loss of both legs or both arms	Lesion at the spinal cord or peripheral nerves
Hemiplegia	Motor loss on one side of the body	Lesion at the contralateral hemisphere of the brain
Quadriplegia	Motor loss of all four limbs	High spinal cord lesion or myopathy
Ascending	Progressive motor loss distal to proximal	Guillain-Barré syndrome, transverse myelitis, tic paralysis
Descending	Progressive motor loss proximal to distal	Myasthenia gravis, botulism, diphtheria
Diffuse Pattern	Sporadic motor loss involving different parts of the body	Hypo/hyperkalemia, thyrotoxicosis, polymyositis, dermatomyositis, toxins

3. Timing, course and fatigability of acute motor weakness

The differential diagnosis of decreased power can be further narrowed based on time it takes to occur or evolve, the course it takes, and whether or not it is fatigable.
Abrupt onset of power loss is a vascular cause until proven otherwise. However, lacunar/small vessel infarcts may present in a stuttering fashion over minutes or hours.

Onset over minutes to hours – consider metabolic disturbances, toxin related or a lacunar/small vessel stroke.
Onset over many hours – consider lacunar/small vessel stroke, Guillain-Barré syndrome, myasthenia gravis and tick paralysis.
Onset over many days – peripheral neuropathies and neuromuscular junction disease usually take a week or longer to develop

Pitfall: Not all strokes result in abrupt onset of motor power loss. A small vessel stroke or lacunar stroke can result in decrease neuromuscular power that takes hours or even days to progress. It is important to keep stroke on the differential in the subacute pattern of motor weakness.

The course of decreased motor strength can also hint at the etiology. Power loss that frequently relapses or fluctuates can be related to multiple sclerosis, periodic paralysis and myasthenia gravis. On the other hand, transient motor loss may be caused by peripheral nerve entrapment or hemiplegic migraine.
Fatigability. Diminished strength with repeated normal use or testing of the muscle is highly suspicious for a neuromuscular junction disease such as myasthenia gravis.

4. Associated findings

There are 5 key associated findings of motor power loss that can help localize the lesion and narrow the differential diagnosis.

Dysarthria or dysphasia	Left hemisphere
Neglect	Right hemisphere
Diplopia, dysarthria, dysphagia, incoordination	Brainstem
Bladder dysfunction	Spinal cord
Absence of reflexes	Peripheral nerve

Clinical Pearl: *Hemi-spacial neglect can be examined through observation and visual field testing. Hemi-sensory neglect can be examined with the double extinction test (see video blelow). Begin with tapping on the patient's forearm on one side and asking which side they felt, right or left. Then repeat the test on the other side. Then tap on both forearms simultaneously. The patient with hemi-sensory neglect will report sensation only on the non-neglected side.*

4. Distinguish upper versus lower motor neuron weakness by degree and speed of movement

The degree of loss of motor power in patients with lower motor neuron lesions are generally greater than in upper motor neuron lesions. For example, a wrist drop from a radial nerve palsy portends a greater loss of power than the loss of wrist extension power seen in a patient with a cerebral infarct.

Upper motor neuron loss of power also comes with slowness (*corticospinal tract slowness*). When a patient is asked to tap their foot or roll their forearms, the tempo is slow. In patients with lower motor neuron lesions, their tapping rolling speed would not be affected.

Clinical	Upper Motor Neuron	Lower Motor Neuron
Reflexes	Increased	Decreased
Muscle Tone	Increased (maybe delayed onset)	Flaccid/decreased
Babinski Sign	Present	Absent
Degree of weakness of affected side	Weak	Extremely weak
Speed (ie., foot tap, arm roll)	Slow	Normal

5. Differentiate the types of lower motor neuron lesions – peripheral neuropathy vs neuromuscular junction vs myopathy

Neuropathy is a process of axonal degeneration or demyelination of nerves. In neuropathic diseases, sensory involvement usually occurs before motor impairment with the longest nerves being affected first. An example is Guillain-Barre syndrome (GBS) where paresthesia occurs first, followed by motor weakness.

Neuromuscular junction disorders are usually systemic due to antibodies against acetylcholine receptor. Distinguishing features of neuromuscular junction processes include fluctuating

weakness along with **early involvement of the ocular** (diplopia, ptosis) and **bulbar** (tongue, jaw, face, larynx) musculature. **Myopathies and neuromuscular junctional pathologies usually involve purely motor deficits.** Myopathies are painful, usually symmetric, with proximal muscles involved.

	Neuropathy	Myopathy	Neuromuscular Junction Disorder
Pattern	Sensory affected before motor	Usually purely motor and no sensory involvement	Usually purely motor and no sensory involvement
Muscle Groups	Distal	Proximal*, usually painful and symmetrical	Multiple groups involved (Early involvement of ocular and bulbar musculatures)
Examples of diseases	Guillain-Barre	Statin-induced myopathy	Myasthenia Gravis

* The proximal motor weakness typically found early in the course of a myopathy may be suggested by *difficulty walking upstairs* or *getting out of a chair* if the lower limbs are involved, or *difficulty combing or brushing hair* with arm involvement.

Clinical Pearl: *If you suspect a myopathy, a review of current medications can help to identify the potential culprit sucha s statins, amiodarone, steroids, and alcohol.*

Dr. Baskind's quick screen motor exam for patients presenting with acute motor weakness

This exam takes about 3 minutes to perform and, when normal, generally obviates the need for a detailed segmental motor exam, unless a specific spinal cord level lesion is in question.

	Maneuvers	Abnormality
Facial symmetry and power testing	Actively close eyes tight, open eyes wide, squeeze lips tight, smile with all teeth showing, open mouth	Any asymmetry or weakness
Pronator drift test of the arm	With eyes closed, both elbows extended, fingers together, and hold position for **30 seconds**	Cupping of hand, flexion of fingers, fifth finger drifting away, and hand pronating
Forearm roll test	Patient makes a fist in both arms and rolls their forearms around each other quickly for 10 seconds and then reverses direction for 10 seconds.	The affected arm with the upper motor neuron lesion will be slow and the non-affected arm will rotate around the slower arm
Index finger roll test	Patient rolls their index fingers quickly around each other for 10 seconds and then reverses direction for 10 seconds.	The affected side will be slow, and the normal finger will rotate around the slow finger.
The drift test of the leg* (aka Mingazzini test):	Position patient supine with hips and knees flexed and ankle dorsiflexed. Have patient hold this position for at least 30 seconds.	The side with the proximal weakness will start to drift down.
Foot tapping*	While sitting, patient should be tapping their feet on the floor quickly and loudly.	Look at the speed of foot tapping with slow side being the weaker side
Reflexes	Deep tendon reflexes and plantar reflexes	Any abnormality in the reflexes is a red flag

*** Pronator drift test of the lower leg and foot tapping can be used to distinguish proximal versus distal muscle weakness respectively**

Take home points on approach to acute motor weakness

- Consider a 5-step approach to acute motor weakness: 1. Does the complaint of weakness represent a true loss of motor power? 2. The geography of weakness – patterns of motor power loss 3. Timing, course and fatigability 4.

Distinguish upper vs lower motor neuron weakness by degree and speed of movement along with reflexes 5. Differentiate the types of lower motor neuron lesions – peripheral neuropathy vs neuromuscular junction vs myopathy based on characteristic findings

- The term "weakness" is vague, and it is better to use "power" or "neuromuscular strength"
- Recognizing not only the patterns of motor power loss but the timing and evolution of muscle weakness helps construct the differential diagnosis
- The five associated findings of motor power loss that should be assessed are language dysfunction, neglect, cerebellar dysfunction, bladder dysfunction and reflexes
- Tachypnea is a sign of impending respiratory compromise in patients suspected of, or know to have, neuromuscular disease
- Avoid using depolarizing neuromuscular blockade, such as succinylcholine, in patient with neuromuscular disease
- The degree of muscle weakness in lower motor neuron lesions is generally greater than upper motor neuron lesions, whereas the speed of repetitive movement (ie., foot tapping) is slower with upper motor neuron lesions
- Sensory impairment usually occurs first before motor impairment in peripheral neuropathies
- Distinguishing features of neuromuscular junction disease include fluctuating weakness along with early involvement of the ocular and bulbar musculature

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