NEURO INSTITUTE

Continuing Education for Rehabilitation Professionals

Upper Motor Neuron Syndrome

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NeuroRestorative's COVID-19 Response

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- We are committed to protecting the health and safety of the individuals we serve, our staff, and the community. Our services are considered essential, and we are taking precautions to minimize disruption to services and keep those in our care and our team members safe. In some programs, that has meant innovating our service delivery model through Interactive Telehealth Services. We provide Interactive Telehealth Services throughout the country as an alternative to in-person services. Through Interactive Telehealth Services, we deliver the same high-quality supports as we would in-person, but in an interactive, virtual format that is HIPAA compliant and recognized by most healthcare plans and carriers.

- You can learn more about our COVID-19 prevention and response plan at our Update Center by visiting **neurorestorative.com**.

Objectives

- Understand the types of deficits in upper motor neuron syndrome.
- Understand when or why to treat.
- Understand the medication vs. non medication treatment options.

Upper Motor Neuron Syndrome

- Lesion of the corticospinal tract
  - Negative signs-performance deficits
    - Loss of dexterity
    - Weakness
    - Loss of selective control over certain limb segments
  - Positive signs-abnormal or exaggerated behaviors
    - Spasticity
    - Clonus
    - Co-contraction
    - Flexor and Extensor spasms
    - Spastic dystonia
    - Associated reactions
    - Muscle stiffness and contracture

Muscle Overactivity

- Spasticity has often been used as a collective term for the positive signs, but muscle overactivity is a better term.
- Treatment is different for the various positive signs.
- Prolonged overactivity can lead to muscle stiffness and contracture after a prolonged period. Can lead to rheologic or viscolelastic/plastic changes.

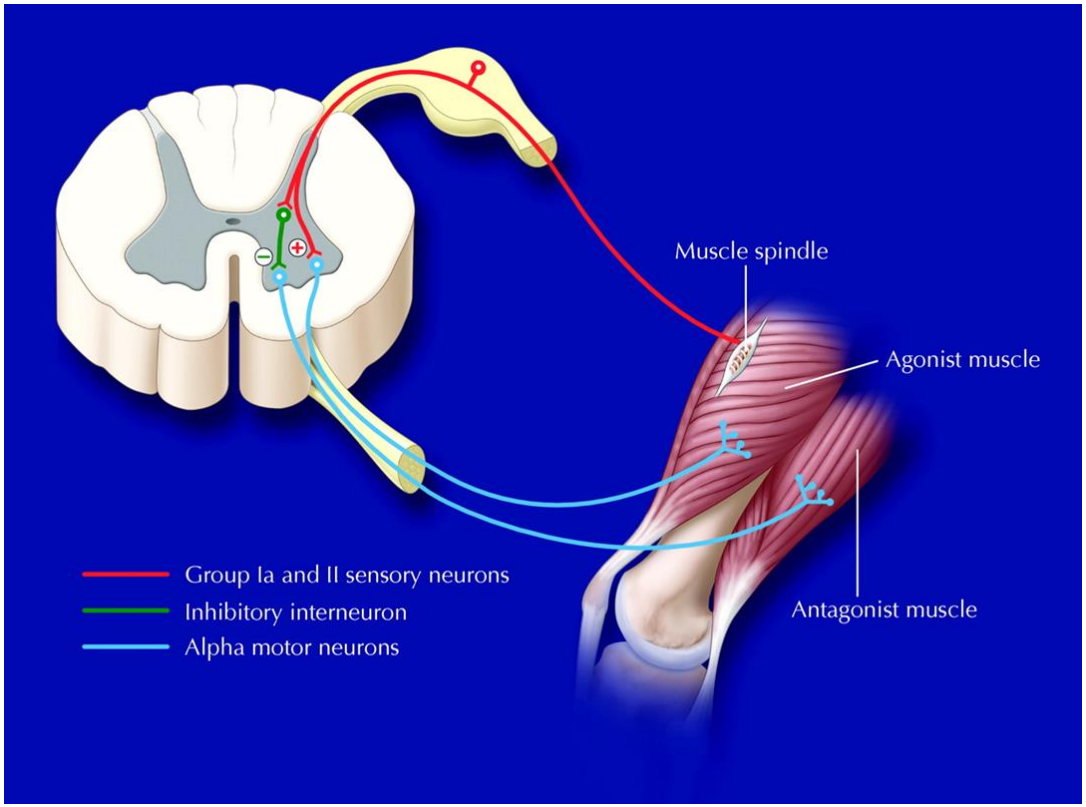
Negative Signs

Represent the broad issue of impaired voluntary control, and when control is absent, muscle overactivity creates unbalanced forces across joints that restrain rom and promote stiffness and contracture.

Spasticity

- An increase in excitability of muscle stretch reflexes, both phasic and tonic, that is present in most patients with UMN lesion.
- Excessive resistance of muscle to passive stretch- resistance increases with the velocity of stretch.

Fig. 1: The stretch reflex arc.



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Stretch Reflex Arc

The stretch reflex arc. When a muscle is stretched, an impulse is generated in the muscle spindle and is transmitted via the sensory neuron to the grey matter of the spinal cord. Here the sensory neuron synapses with the motor neuron, and the transmitted impulse results in muscle contraction. While agonist muscles contract in response to stretching, antagonist muscles must relax. Their relaxation is brought about via an inhibitory neuron within the spinal cord.

Spasticity

- The cord is felt to be in an excitatory state which promotes exaggerated motor responses to ordinary cord input.
- Multiple theories of “signal mishandling”.
- Depends on muscle stretch and onset occurs after movement has begun after some stretch displacement has taken place.

Clonus

- Low frequency rhythmic oscillation in one or more limb segments.
- Generated by rapid stretch and hold of a muscle group.
- Can be triggered by a patient who unintentionally stretches a muscle group during limb positioning.
- Can be triggered during voluntary movement
- May be sustained or unsustained and can be stopped with repositioning at shorter lengths.
- Usually associated with other hyperexcitable phasic stretch reflexes.

Co- Contraction

- Simultaneous activation of agonist and antagonist muscles during voluntary movement.
- May represent an impairment of supraspinal control of reciprocal inhibition.
- May occur during isometric effort so that it is not necessarily related to muscle stretch.
- Simultaneous activation of flexors/extensors of the elbow.
- Can be a normal mechanism to provide joint stability, but can be unwanted
- Generated by simultaneous motor drive to agonist/antagonist during voluntary effort

Co-Contraction

- When a co-contracting antagonist undergoing stretch develops superimposed spasticity, the combination can be called spastic co-contraction.
- Slow effortful movements
- Speed of back and forth is often asymmetrical.
- May respond to a weakening strategy- chemodenervation or tendon lengthening. Tizanidine may help.

Flexor and Extensor Spasms

- The flexor reflex is elicited by afferent stimuli including cutaneous receptors responding to
  - Touch, temperature and pressure
  - Nociceptors responding to painful stimuli
- By typically recruiting flexor muscles across several joints, the flexor reflex is an example of an interjoint reflex that has tissue protective value such as enabling quick withdrawal from a noxious stimuli.
  - Can also be an extensor reflex.

Flexor/Extensor spasm

- After UMN lesions- esp spinal cord- release of inhibitory descending influences makes these reflexes more pronounced.
- Spasms may represent disinhibited flexor/extensor reflexes
- Overt stimuli can trigger flexor and extensor spasms or they may be set off by covert stimuli such as full bladder, distended bowel, other sensory sources.
- The threshold is reduced, the intensity of muscle contraction for the same stimulus input is increased and interjoint components of the reflex are often expanded.
- Baclofen/diazepam can inhibit the polysynaptic activity in the cord and reduce the frequency and intensity of spasms.

Spastic Dystonia

- Tonic muscle activity that maintains the limb in a fixed posture in the absence of phasic stretch or voluntary effort.
- Abnormal supraspinal drive- characterized by an inability to inhibit muscle activity despite efforts to do so.
- May need EMG to determine.
- Limb also can be held in fixed position due to tissue stiffness, contracture, HO

Associated Reactions

- Involuntary activity in one limb that is associated with a voluntary movement effort made by other limbs.
- May be due to disinhibited spread of voluntary motor activity into a limb affected by an UMN lesion.
- Felt to be supraspinal in origin
- Baclofen/diazepam would not be expected to work on this or dystonia
- Many movements done daily in these patients that can lead to associated reactions, that cause muscle stiffness and contracture as much as spasticity itself does.
- More studies are needed.

Muscle Stiffness and Contracture

- Physical shortening of muscle length and it is often accompanied by fixed shortening of other soft tissues such as fascia, nerves, blood vessels and skin.
- Development is promoted by a number of processes that start when an acute UMN lesion occurs.
  - Paresis impairs cycles of shortening and lengthening of agonist/antagonist muscles
  - Gravity generates positional effects
  - Muscle overactivity
- If all muscle contraction were blocked, physical shortening would still remain.

Muscle Stiffness and Contracture

- After UMN lesion, paralyzed muscles immobilized for long periods in a shortened position become shortened and stiffer .
- When muscle overactivity develops in the shortened muscles, tension is generated at shorter lengths.
- A lack of voluntary contraction in the antagonists prevents their natural reextension, leading to a continuation of the process.
- Physical/surgical treatments necessary.
- Typically only correct for about ½ of range due to the shortening of nerves/vessels/skin.

Evaluation

- Identify the clinical pattern of motor dysfunction
- Identify the patient's ability to control muscles involved in the clinical pattern
- Identify the role of muscle stiffness and contracture as it relates to functional problem.
- In TBI- the degree of voluntary function that remains can vary greatly from muscle to muscle even in the same limb segment

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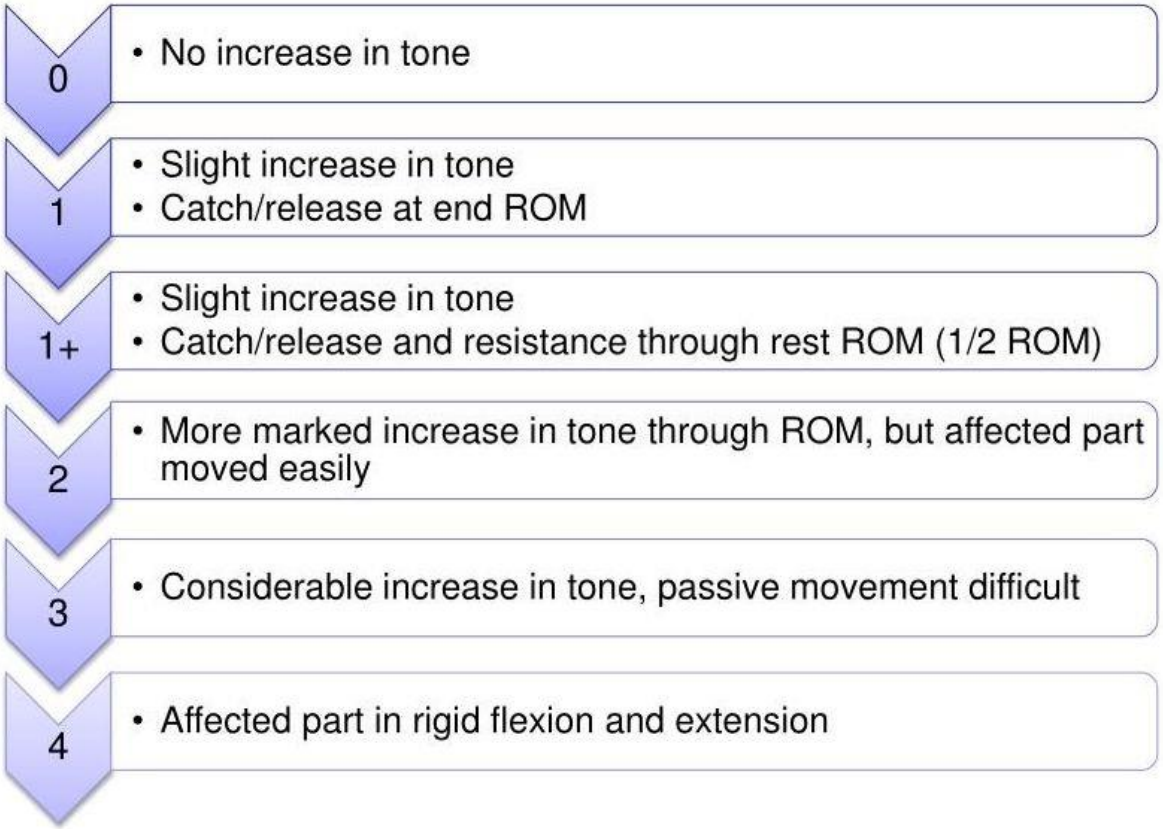
Modified Ashworth Scale



Reasons to treat

- Pain
- Function
- Hygiene
- Caregiver ease

Pharmacologic Treatment

- Often a trade off against side effects.

Dantrolene

- Effect is directly on the skeletal muscle fibers.
- Inhibits release of calcium from the sarcoplasmic reticulum
- Calcium initiates cross-bridging of myofilaments and build up of contractile tension.
- Reduces the force of muscle contraction and can reduce tension in overactive muscles.
- Good for clonus or brief spasms.
- Can be sedating, but less so than the others.
- Hepatotoxocity- monitor liver functions.

Diazepam

- Centrally acting and highly sedating
- Increases the central inhibitory effects of GABA(gamma amino-butyric acid).
- Appears to bind to receptors located at GABA-ergic synapses and increases GABA inhibition at those sites.
- The helpful effect in muscle overactivity appears to arise from the inhibitory effect on the alpha motor neuron in the spinal cord.
- Helpful at night for spasms with tolerable sedation.

Baclofen

- Derivative of GABA
- Appears to act as a GABA agonist inhibiting transmission at specific synapses within the spinal cord.
- Inhibitory effect on alpha motor neuron (lower motor neuron).
- Very good for SCI/MS- especially with spasms.
- Sedating side effects
- Not studied much in cerebral causes.

Baclofen

Intrathecal delivery

- Can reduce the sedating effects
- Much smaller doses are used and delivered into the subarachnoid space
- Has to be adjusted and refilled frequently
- Overdose or withdrawal can be life threatening.
- Overdose- respiratory depression/coma
- Withdrawal- increase spasticity, itching, hallucinations, seizures
- Technical problems
- Family must be compliant and reliable

Tizanidine

Acts at alpha 2 adrenergic receptor sites both spinally and supraspinally

Reduces muscle response to passive stretch in both the spinal and cerebral forms of muscle overactivity.

Side effects- hypotension, sedation, fatigue, dry mouth, hepatotoxicity

Good for co-contraction

Botulinum Toxin

- Focal action without significant side effects
- Injected directly into the affected muscle groups
- EMG/ultrasound guidance
- Causes reversible, dose dependent muscle relaxation by blocking acetylcholine at the neuromuscular junction.
- Can use BoNt A or B
- Benefits 3-7 days after injection
- Duration is about 3 months
- Adverse effects- excessive weakness, pain at injection site, headache, fatigue, flu-like symptoms

Nerve Blocks

- Diagnostic with lidocaine/bupivicaine- short acting to evaluate passive rom, muscle stiffness, changes in active rom when dynamic resistance is blocked, and enhanced motor control
- Phenol-when injected in or near a nerve bundle, it denatures protein in the myelin sheath or cell membrane of the axons.
- Can be administered directly into the nerve bundle in the OR
- Inject desired motor nerves, avoid mixed nerves with sensory and motor fibers to avoid dysesthesias.
- Duration about 6 months

Physical modalities

- TENS
- Dynamic splinting
- Serial casting
- All used with a combination of active therapy

Surgery

- If deformity is severe, may warrant early surgery to prevent a fixed contracture
- Releasing a contracted limb may allow improved ADLs even if the limb itself does not improve.
- Consider cost of not correcting limb deformities

    Consider dynamic EMG prior to surgery to avoid a procedure that worsens a deformity.

    Will treating a focal problem help just as much as a

    systemic agent

Treatment

- All of these treatments require ongoing therapy and stretching
- Patient and family education
- Nursing/staff education to prevent contractures early on.
- Range of motion and splints

References

- Brain Injury Medicine. Zasler, Katz, Zafonte. 2007
- Rehabilitation Medicine 3: Management of Adult Spasticity. Satkunam. Canadian Medical Association Journal.
- November 2003.
- Transcutaneous Electrical Nerve Stimulation for Management of Limb Spasticity-A Systematic Review. Mills, Patricia Branco MD, MHSc, FRCPC; Dossa, Farhana BASc, MSc. American Journal of Physical Medicine & Rehabilitation: April 2016 - Volume 95 - Issue 4 - p 309-318