

Upper Motor Neuron vs. Lower Motor Neuron Disease
Never Miss Again After This Video Case Presentation
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Introduction

The ability to examine a patient and determine where in the body the disease is located is critical to determining the cause, best therapy and prognosis. Weakness is a common presenting complaint and can result from spinal cord (upper motor neuron - UMN) or nerve and muscle disease (lower motor neuron-LMN). Understanding the function of the UMN and LMN system will enhance the accuracy of lesion localization and therefore determination of the diagnostic testing and prognosis. This talk will discuss the function of the UMN and LMN system and then how to assess these systems via examination of a patient's gait, postural responses, and reflex testing. Secondly we will discuss assessing the cutaneous trunci reflex and for focal pain to assist in lesion localization and honing the list of possible the spinal cord diseases. Lastly we will discuss evaluating the palpebral response and laryngeal / pharyngeal / esophageal function in LMN disease.

Tonic Gamma Loop Mechanism

Muscle tone refers to the intrinsic tension of the muscle when supporting the body against gravity while standing, shifting weight from limb to limb, jumping or performing stairs – but how does this occur? The tonic gamma loop mechanism (TGLM) is intrinsic to the LMN system and understanding TGLM physiology offers insight into how gait is generated and why UMN or LMN disease cause alterations in muscle tone and reflex. When we jump down from a height and our knees start to bend or buckle there is a sudden stretch of the quadriceps muscles and stimulation of stretch receptors (neuromuscular spindle) within these muscles. These receptors then stimulate the sensory portion of the femoral nerve which then directly synapse and stimulates the motor portion (alpha motor neuron) of the femoral nerve. This causes the quadriceps muscle (which is innervated by the femoral nerve) to contract and prevent your knees from buckling. When the patella tendon is artificially simulated with a reflex hammer it fools the body into thinking there is a sudden heavy load on the quadriceps muscle (like jumping down), the TGLM is stimulated and the knee jerks.

Gait Generation

As mentioned above, when information from the neuromuscular spindle of the TGLM returns to the spinal cord it directly synapses on the alpha motor neuron or motor portion of the same muscle in which the spindle is located. However, the information also stimulates inhibitory interneurons that then reduce activity or tone in the antagonistic muscle group. Therefore when there is contraction of the quadriceps there will be reduced tone in the hamstring or flexor group. The modulation of flexion is performed by the phasic gamma loop or flexor gammas loop. The UMN system acts on the tonic and then phasic gamma loop mechanisms to generate extension and then flexion of the leg by activating this intrinsic reflex mechanisms and therefore generate gait.

UMN Lesions Influence the TGLM

In our example above, gravity lengthen the quadriceps muscle and stretches the neuromuscular spindle, which then via a direct synaptic connection, stimulates the femoral nerve. This causes contraction of the quadriceps muscle which then causes more stretching of the neuromuscular spindle and more contraction of the quadriceps muscle. This system, if not modulated would lead to dramatic increases in muscle tone and reflex. The UMN system modulates or controls the TGLM and therefore controls muscle tone and reflex. Disease of the UMN lesion can cause increased tone and reflex. Examination of dogs with UMN spinal cord disease often reveals increased tone because there is resistance to flexion of the stifle. This stiffness can also manifest in the protraction phase of the gait and appear as swinging out of the limb (circumduction) or a long-strided gait. Furthermore, brainstem lesion (where the UMN tracts start) can lead to opisthotonus also known as decerebrate rigidity where the head, neck and limbs are held in rigid extension.

Reflex Testing

A reflex is something that occurs automatically or spontaneously without influence from the cerebrum whereas a reaction or response requires the unconscious participation of the cerebrum. In reflex testing there is a sensory stimulus that runs into the spinal cord or brainstem and then an immediate spinal cord or brainstem mediated response. For example, stretching the patella tendon with a pleximeter (reflex hammer) causes a sudden, intense stimulation of the stretch receptors within the femoral nerve, in essence simulating what would happen if we jumped down from a large height. Immediately the muscles innervated by the femoral nerve contract and the knee jerks. An absence of reflex often means there is a lesion of the motor or sensory portion of the femoral nerve or severe disease of the quadriceps muscle. If there is an increase in reflex (exaggerated, clonus) then there is a failure of UMN system to control this reflex.

Upper Motor Neuron System

The UMN system primarily starts in the brainstem. The axons from this collection of neurons run within the white matter of the spinal cord and synapse in the ventral horn of the spinal cord to activate the peripheral nerve (LMN). This system activates the LMN to generate gait and modulates or controls tone and reflex by influencing the tonic gamma loop mechanism. A lesion of the descending or motor component of the UMN system results in paresis (weakness), paralysis, increased reflex and increased muscle tone. A lesion of the ascending or sensory system causes a disordered gait and postural deficits (see below).

Lower Motor Neuron System

The LMN system starts within the spinal cord where the cell bodies are grouped in the grey matter of the spinal cord within the ventral horn at the intumescence (swelling) located at spinal cord segments C6-T2 and L3-S3. The numbered nerves then run to the brachial or lumbar plexus and then exit as named nerves that will then innervate specific muscles. The LMN generates muscle tone and with a lesion there is weakness, paralysis and loss of muscle tone and reflex. The LMN system also carries sensory information from receptors in the joints and skin into to the dorsal horn of the spinal cord and this is eventually relayed via the UMN system to the cerebellum and somatosensory cortex via the spinocerebellar and spinothalamic tract, respectively.

Spinal Cord Ataxia and Postural Reactions

A complete lesion of the UMN system causes no movement or paralysis and an increase in muscle tone (spastic paralysis). A partial lesion will cause only weakness or paresis but the movement will be ataxic. Ataxia means disorder. The absence of ascending information reaching the brain can result in a loss of self-reception (proprioception) and consequently spinal cord or proprioceptive ataxia and slow postural reactions. Spinal cord ataxia can take the form of a long-strided gait, the limbs can circumduct, cross midline, and interfere with each other - occasionally causing the patient to trip or fall. In addition the patient might stand on the dorsal surface of the paw or stand with limbs too close, too far apart or with limbs crossed. Besides observation of the gait, testing of the postural reactions (paw flip test, hopping, tactile placing) also assesses the function of the UMN system. The postural reactions will be delayed to absent with an UMN lesion.

LMN Lesions

A complete lesion of the LMN system causes paralysis with an absence of muscle tone (flaccid paralysis). An incomplete lesion causes weakness and the patient will have a short-strided or choppy gait as though they are walking on egg shells. Importantly, incomplete LMN lesions do not cause significant disruption of the sensory system. Therefore LMN lesions do not cause ataxia. Furthermore, if the patient's weight is properly supported the postural reactions will be normal. Please see Table 1.

C6-T2 Spinal Cord

A lesion that involves the white matter of the spinal cord at C6-T2 will cause UMN signs to the pelvic limbs. The pelvic limbs will have increased tone and reflex, reduced postural reactions, weakness and ataxia. A lesion of the grey matter in this area will generate LMN signs to the thoracic limbs manifested as a short-strided gait, preserved postural reactions and no ataxia, reduced reflex, and neurogenic muscle

atrophy. The long-strided, stiff and ataxic gait in the pelvic limbs is much different than the short-strided gait of the thoracic limbs and sometimes referred to as a two engine gait.

T3-L3 Spinal Cord and the Cutaneous Trunci Reflex

Disease between the two intumescences is called T3-L3 spinal cord disease and results in upper motor neuron disease to the pelvic limbs. The presence of a cut-off or cessation of the cutaneous trunci reflex can indicate the level of the spinal cord lesion. The input for the reflex is stimulation of dorsolateral cutaneous receptors. Once a stimulus is registered the information then ascends in the spinal cord where it synapses motor neurons at the level of spinal cord segment C8 -T2. These nerves form the lateral thoracic nerve that causes contraction of the cutaneous trunci muscle. Functionally a pinch of the skin with hemostats should stimulate contraction of the entire cutaneous trunci muscle along the entire flank of the patient. With a thoracolumbar spinal cord lesion, pinching of the skin behind the lesion will not result in twitching of the skin and thus there appears to be a cut-off of this reflex. A cut-off in the cutaneous trunci reflex indicates the lesion is about 2 vertebral bodies cranial to the cut-off. Furthermore, following surgery movement of the cut-off caudally predicts recovery while movement cranially predicts myelomalacia.

Lumbar Intumescence and Nerve Root Disease (Lumbosacral Syndrome)

Disease of the spinal column or spinal cord/nerve roots from the L5 to S1 vertebrae can generate LMN signs to the pelvic limbs, fecal and urinary incontinence as well as paralysis of the tail. These signs can overlap and be mistaken for osteoarthritis of the hip or stifle. A sciatic lesion can be the cause of an increased patella reflex as a consequence of losing strength and tone to the antagonist of stifle extension, this is called a pseudo-hyperpatella reflex and should not be mistaken for an UMN reflex. A reduction of the patella reflex can help localize lesion to L3-L4 vertebral bodies and would not be expected with disease from L5 – S1 vertebrae. The patella reflex can be absent in otherwise healthy middle-age and older dogs, presumably from degeneration of the sensory portion of the femoral nerve.

Pain Assessment

Diseases of the nerve and muscle (LMN disease) are typically not painful, however many spinal cord diseases are associated with pain. Determining the patient is painful at a specific location can direct diagnostic testing and also hone the list of possible causes of disease – for instance intervertebral disk disease, neoplasia, and diskospondylitis are typically painful whereas ischemic myelopathy (fibrocartilaginous emboli) and acute, non-compressive nucleus pulposus extrusions are often non-painful, especially after the first 24 hours. Neck pain is often suspected when patient spontaneously yelps out but there is no gait or posture deficits, intermittent thoracic limb lameness (root signature), or stiff neck or decreased range of motion is noted. Palpating muscle spasm laterally at level of transverse process, pain with manipulation or ventral process of C6, or resistance to range of motion can also indicate neck pain. Mid-back pain is often suspected with kyphosis, stiffness and when slow to sit or rise. Palpating and applying pressure to dorsal processes while putting pressure / palpating the ventrum and palpating muscle / rib heads at level of transverse process often allow for detection of back pain. Lumbosacral pain is suspected with abnormal tail carriage and when patient is slow to sit and rise. Pain can often be detected with rectal palpation of the lumbosacral junction (or spondylosis at L7-S1), tail extension or by applying pressure to muscle between dorsal process of L7 and S1. Hip extension will not differentiate back from hip pain. However, hip pain can be discerned by slowly elevating the femoral head about 3-5 mm from acetabulum by lifting up on the medial surface of the femur while the patient is in lateral recumbency.

Cranial Nerve Exam in LMN Disease

LMN disease can affect cranial nerves when there is a polymyositis, polyneuropathy, or disease of the neuromuscular junction (Myasthenia gravis). When LMN disease is suspected then a few physical examination maneuvers can be helpful. Firstly listen to the patient's breathing – a respiratory stridor can indicate weakness of neuromuscular system that abducts the vocal folds. Gagging can indicate pharyngeal weakness or incoordination and misdirection of saliva into the airway. Pneumonia may be present from laryngeal or pharyngeal dysfunction or from megasophagus – listen for a soft, moist cough and carefully auscultation the lungs. Thoracic radiographs are indicated in dogs with suspected LMN

disease to assess for megesophagus, aspiration pneumonia and other pathology. Secondly, assess temporalis muscle mass because marked atrophy can indicate a lesion of the mandibular nerve. Lastly, repeated stimulation of the medical canthus of the eye should provoke a prompt and complete blink response – incomplete blinking or an absent blink indicates there is neuromuscular disease.

Table 1. Distinguishing Characteristics of UMN and LMN Disease

| | UMN | LMN |
|---------------------|--------------|---------------|
| Gait Characteristic | Long strides | Short strides |
| Ataxia | Yes | No |
| Postural Deficit | Yes | No |
| Tone & Reflex | Increased | Decreased |
| Atrophy | No | Yes |
| Spinal Pain | Often | Seldom |

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