**QUESTION**

with amyotrophic lateral sclerosis, the patient usually first complains of:

a) weakness  
b) difficulty breathing  
c) numbness  
d) paresthesia  

amyotrophic lateral sclerosis often begins with degeneration of:

a) muscle mass  
b) the motor unit  
c) the anterior horn  
d) the **pyramidal tracts**  

amyotrophic lateral sclerosis is usually associated with ______ deficits  

a) segmental  
b) **progressive**  
c) autonomic  
d) cognitive  

amyotrophic lateral sclerosis is best classified as a disease of what?  

a) myelin  
b) **motor neurons**  
c) alpha motor neurons  
d) the dorsal horns  

trouble sleeping in a patient with ALS is often because of:  

a) upper motor neuron lesions that cause muscle spasm  
b) upper motor neuron lesions that cause hyperreflexia  
c) numbness  
d) sympatheticotonia  

**ANSWER**

a) **amyotrophic lateral sclerosis**  
*myo – muscle, a-negative impact on muscle  
*amyotrophic = wasting and thinning of muscles  
*lateral refers to part of spinal cord degeneration  
*a subgroup is “primary lateral sclerosis” which is purely UMN disease at first, that may turn into LMN  
*UMNs of corticospinal tract may be affected first, then anterior horn cells  
*ALS is grouped into a larger category called “motor neuron disease”  
*GENERALIZED WEAKNESS OR PARALYSIS  

**symptoms**  
*first complaint of pt. is usually weakness  
some pts. will notice muscle atrophy  
painful muscle cramps usually at night, sleep disturbances  
fasciculations (pt. notices twitching of muscles)  
respiratory failure in later stages  

**signs**  
*neurological exam reveals **paresis** that may be symmetrical  
*weakness in multiple muscle groups  
fasciculations  
paresis may be present for a long time before muscle atrophy is evident  
spasticity, hyperreflexia, **pyramidal tract signs**  
spasticity may be canceled out by LMNL  
*spasticity may occur before ventral horn ganglion cells are affected  
disease may initially present as spastic spinal paralysis, or pseudobulbar palsy  
*progression of dz affects m. of respiration  
**bulbar signs**  
*slurred speech, dysphagia, flaccid facial paresis  
tongue atrophy  
tongue fasciculations  
enhancement of muscle reflexes of face: jaw jerk  
foot drop  
paresis of intercostal muscle and diaphragm  

**tests**  
*chronic denervation and subsequent renervation leads to increased size of motor unit. denervated fibers are reinnervated by the terminal sprouts of surviving motor neurons. this leads to EMG of very large motor units with abnormal spontaneous activity, with **large amplitude**, and **fasciculations**  

**what would a patient with ALS demonstrate?**  
radiculopathy: segmental SSx (this disease affects multiple levels, often starting with the cord)  
dermatome hypoesthesia  
myotome hypotonicity  
autonomic deficits  
paresthesia  

**normal tests, modalities**  
finger-to-nose  
heel-to-shin  
rapid alternating movements  
bladder function  

**what are all of the pyramidal signs that a patient with ALS could demonstrate?**  
light touch, pinprick, vibration, joint position sense  

**what would a patient with ALS NOT demonstrate?**  
pronator sign  
dysdiadochokinesia  
Romberg  
praxis deficit  

a) the glabellar reflex  
b) the grasp reflex  
c) the abdominal reflex  
d) the sucking reflex

**what are the signs of a corticospinal/pyramidal lesion?**
- pathological reflexes/primitive reflexes: babinski-extensor reflex, grasp reflex, sucking reflex, glabellar reflex  
- loss of superficial reflexes (**abdominal, anal, cremasteric, corneal**)

<table>
<thead>
<tr>
<th>a patient with advanced ALS would not demonstrate:</th>
<th>c. <strong>Motor Unit</strong></th>
</tr>
</thead>
</table>
| a) extensor planter reflex  
| b) hyperreflexia  
| c) weakness  
| d) **cremasteric reflex**  |
| in an early form of ALS, in which only the corticospinal tract is affected, the patient would have a normal: |
| a) muscle tone  
| b) reflexes  
| c) motor unit  
| d) strength |
| in an advanced stage of ALS, what is a typical normal finding? |
| a) flexor planter reflex  
| b) astereognosis  
| c) normal peripheral nerve conduction velocity  
| d) normal EMG findings |
| if the patient has a form of ALS that begins with the degeneration of the UMN and will progress to degeneration of the LMN, hypotonicity will become evident as soon as the ________ is affected |
| a) corticobulbar tract  
| b) rubrospinal tract  
| c) nerve root  
| d) **lower motor neuron**  |
| if the patient has a form of ALS that begins with the degeneration of the UMN and will progress to degeneration of the LMN, hypotonicity will become evident as soon as the ________ is affected |
| a) **motor unit**  
| b) pyramidal tract  
| c) peripheral nerve conduction velocity  
| d) upper motor neuron |
| a patient with ALS will **not** display |
| a) **latency of ulnar nerve conduction**  
| b) neuropathy  
| c) recruitment of denervated muscle fibers  
| d) decentralization |
| a common finding in ALS is tongue fibrillation, which is most likely due to: |
| a) loss of extrapyramidals  
| b) **denervation potentials**  
| c) loss of upper motor neurons  
| d) myopathy |
The motor unit consists of one lower motor neuron (alpha) and all of the individual muscle fibers innervated by it. Lesions from the cell body to and including the myoneural junction are classified as neuropathies. In conditions where motor units recruit nearby denervated muscle fibers (decentralization) we see an increasing size of motor units although the overall population of motor units is decreasing. This can lead to the onset of “clumsiness” and reduction in ability to perform fine motor acts.

onset of muscle wasting, in ALS, begins when the ________ is affected

a) muscle strength
b) muscle tonicity
c) corticobulbar pathway
d) anterior horn cells

*autonomics are spared, bladder, GI function, sexual function
Clinical diagnosis of ALS requires findings of:

a) decreased NCV and EMG changes
b) unilateral motor deficits
c) respiratory paralysis
d) UMN and LMN lesions

d. and progressive spread of these sx.

Requirements for the Diagnosis of Amyotrophic Lateral Sclerosis

The diagnosis of amyotrophic lateral sclerosis (ALS) requires

A. The presence of

1. Evidence of lower motor neuron (LMN) degeneration by clinical, electrophysiological, or neuropathologic examination,
2. Evidence of upper motor neuron (UMN) degeneration by clinical examination, and
3. Progressive spread of symptoms or signs within a region or to other regions, as determined by history or examination,

and together with

B. The absence of

1. Electrophysiological and pathological evidence of other disease processes that might explain the signs of LMN and/or UMN degeneration, and
2. Neuroimaging evidence of other disease processes that might explain the observed clinical and electrophysiological signs.

A. El Escorial Criteria

Definite ALS: UMN and LMN signs in three regions.

Probable ALS: UMN and LMN signs in at least two regions with UMN signs rostral to (above) LMN signs.

Possible ALS: UMN and LMN signs in one region, UMN signs alone in two or more regions, or LMN signs above UMN signs.

Suspected ALS: LMN signs only in two or more regions.

B. Airlie House (modified) criteria

Clinically definite ALS: clinical evidence alone of UMN and LMN signs in three regions.

Clinically probable ALS: clinical evidence alone of UMN and LMN signs in at least two regions with some UMN signs rostral to (above) the LMN signs.

Clinically probable—laboratory-supported ALS: clinical signs of UMN and LMN dysfunction are in only one region, or UMN signs alone in one region with LMN signs defined by EMG criteria in at least two limbs, together with proper application of neuroimaging and clinical laboratory protocols to exclude other causes.

Possible ALS: clinical signs of UMN and LMN dysfunction in only one region, or UMN signs alone in two or more regions; or LMN signs rostral to UMN signs and the diagnosis of clinically probable—laboratory-supported ALS cannot be proven.

Suspected ALS: this category is deleted from the revised El Escorial Criteria.
an early neurological sign of ALS is:

a) DTR grade < 2  
b) EMG changes  
c) clonus  
d) final common pathway deficits

c. note that the picture below says “demyelination”... well this is kind of misleading. the whole neuron dies.

Amyotrophic lateral sclerosis (ALS, also known as Lou Gehrig's disease) is a severe condition characterized by progressive degeneration of motor neurons of the brainstem and spinal cord.

Amyotrophic refers to muscle atrophy. Lateral sclerosis refers to the hardening to palpation of the lateral columns of the spinal cord in autopsy specimens. Lateral sclerosis is caused by an increased number of astrocytes (astrocytic gliosis) following the degeneration and loss of motor neurons.

ALS is a familial motor neuron disease in 5% to 10% of cases. The others are assumed to be sporadic. Mutations in the gene encoding superoxide dismutase 1 (SOD1) account for 20% of the cases of familial ALS. The remaining 80% are caused by mutations of other genes.

SOD1 is an enzyme that requires copper to catalyze the conversion of toxic superoxide radicals to hydrogen peroxide and oxygen. The toxic effects of mutant SOD1 results in the disorganization of intermediate filaments (NF-L, NF-M, and NF-H; see Figure 8-4), mitochondrial abnormalities, and apoptosis of motor neurons. Autophagy may have a role in the pathogenesis of ALS. Patients with sporadic ALS have antibodies against voltage-gated Ca2+ channels, which may interfere with the regulation of intracellular Ca2+, leading to the degeneration of motor neurons. However, immunotherapy has not been effective in patients with ALS.

The clinically apparent signs are overactive tendon reflexes, Hoffman's sign (digital reflex; flexion of the terminal phalanx of the thumb following nipping of the nail), Babinski sign (extension of the great toe and abdution of the other toes after plantar stimulation), and clonus (Greek klonos, a tumult; muscle contraction and relaxation of a muscle in rapid succession).

what would differentiate ALS from cauda equina syndrome?

a) Babinski sign  
b) leg muscle weakness bilaterally  
c) decreased leg muscle tone bilaterally  
d) fasciculations of leg muscles bilaterally

A 47-year-old man came for evaluation of a 2-month history of weakness and wasting in the right hand. Thinning, weakness, and atrophy were present in all of the muscles of the right hand and arm, and the reflexes were abnormally active. Both his mother and his maternal uncle had died of ALS.

the above patient is experiencing what signs?

a) upper motor neuron lesion  
b) lower motor neuron lesion  
c) upper and lower motor neuron lesion  
d) brain lesion

c. atrophy and twitchs (fascialculation) are seen in LMNL hyperreflexia is due to UMNL

<table>
<thead>
<tr>
<th>Upper motor neuron weakness (UMN)</th>
<th>Lower motor neuron weakness (LMN)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spasticity or very stiff</td>
<td>Flaccid or reduced stiffness</td>
</tr>
<tr>
<td>Increased tone</td>
<td>Decreased tone</td>
</tr>
<tr>
<td>Increased muscle stretch reflexes</td>
<td>Decreased muscle stretch reflexes</td>
</tr>
<tr>
<td>Minimal muscle atrophy</td>
<td>Profound muscle atrophy</td>
</tr>
<tr>
<td>Fasciculations absent</td>
<td>Fasciculations seen or twitchs</td>
</tr>
<tr>
<td>+/- sensory disturbances</td>
<td>+/- sensory disturbances</td>
</tr>
</tbody>
</table>

c. these contain the lower motor neuron cell bodies. when the LMN degenerates, the tongue undergoes denervation. fasciculation and fibrillation is a result of this due to increased ACh receptors.
in a patient with ALS, motor neurons degenerate in which locations?
a) cord only  
b) PNS only  
c) cord and PNS  
d) brain, cord, and PNS

**FIGURE 27-1**  
Amotrophic lateral sclerosis. Axial T2-weighted MRI scan through the lateral ventricles of the brain reveals abnormal high signal intensity within the corticospinal tracts (arrows). This MRI feature represents an increase in water content in myelin tracts undergoing Wallerian degeneration secondary to cortical motor neuronal loss. This finding is commonly present in ALS, but can also be seen in AIDS-related encephalopathy, infarction, or other disease processes that produce corticospinal neuronal loss in a symmetric fashion.
A 45-year-old man complains of slowly progressive muscle weakness involving the hands and lower limbs.

He initially noticed reduced finger dexterity and wrist drop (extremity weakness), and his family noticed occasional slurring of words and choking at meals (bulbar signs). He has had muscle wasting, weakness, rigidity, and slowness. He denies any incontinence or changes in his bowel habits. He has also noted difficulty walking.

VS: normal. PE: muscle atrophy, weakness, wasting, fasciculations, loss of stretch reflexes, and bradykinesia noted in upper extremities bilaterally (LMN signs); muscle rigidity, spasticity, clonus, and hyperactive DTRs noted in bilateral lower extremities (UMN signs); Babinski’s sign present (UMN sign); spastic gait; no sensory deficit; normal cognitive exam.

Muscle biopsy shows grouping of muscle fiber types (as nerves die, adjacent nerves send buds to reinnervate muscle and fibers switch types). LP: mildly elevated protein (50 mg/dL) in CSF EMG: fasciculations and evidence of denervation in upper extremities with normal nerve conduction velocities.

CT/MR, brain and spinal cord: normal.

the clonus that this patient experiences is a sign of:

a) denervation supersensitivity
b) upper motor neuron degeneration
c) fibrillation and fasciculation
d) peripheral neuropathy

Amyotrophic lateral sclerosis (ALS), a.k.a. Lou Gehrig’s disease, causes progressive muscle weakness and atrophy that typically begins distally and proceeds proximally. The cause is unknown, but glutamate toxicity, mitochondrial dysfunction, and autoimmunity may play a role. ALS affects both anterior horn cells in the spinal cord and UMN in the corticospinal tract, resulting in both UMN and LMN deficits, which may be asymmetric.

Males are more likely to be affected than females. Incidence rises after age 40 and continues to increase until about 80. Occasionally associated with dementia and parkinsonism. A familial form of the disease with autosomal-dominant inheritance has been identified.

No specific treatment. Riluzole, which reduces the presynaptic release of glutamate, may slow progression. Symptomatic management is indicated, including anticholinergics to prevent drooling and braces and physical therapy to assist mobility and prevent contractures.

Dysphagia, respiratory compromise, and aspiration; death often within 5 years of symptom onset.

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**a. amyotrophic lateral sclerosis, and a form of it: progressive bulbar palsy**

---

**a. wasted, fasciculating, slow moving tongue in a patient with amyotrophic lateral sclerosis, indicates what?**

a) denervation supersensitivity
b) nerve conduction velocity is latent
c) a myopathy is occurring, and there is no evidence of a neuropathy
d) the motor unit is intact, and the lesion lies in the corticospinal tract

---

**a. wasted, fasciculating, slow moving tongue in a patient with amyotrophic lateral sclerosis, indicates what?**

a) the disease only involves upper motor neurons
b) the corticobulbar tract is degenerating
c) the body is in a state of sympatheticonia
d) the alpha motor neuron has been affected
when upper motor neurons are affected, but not lower motor neurons, the patient with ALS will exhibit what feature on EMG?
a) fasciculation and fibrillation
b) increased amplitude of MUAP
c) recruitment of denervated muscle fibers
d) normal motor and sensory conduction

when both upper and lower motor neurons are affected in a patient with ALS, what is a typical finding on EMG?

a) **large amplitude of MUAPs**
b) increased number of MUAPs
c) normal MUAPs

a muscle biopsy of a victim with advanced ALS will show:
a) small motor units
b) **denervation atrophy**
c) myopathy

a major difference between brown-sequard and ALS is:
a) combined upper and motor neuron lesion
b) reflex changes
c) weakness
d) sensory findings

what do patients with syringomyelia, brown-sequard syndrome, and ALS have in common?
a) peripheral neuropathy
b) loss of tactile discrimination
c) **absence of segmental findings**
d) radicular deficits

respiratory paresis in a patient with advanced ALS is due to what?
- lower motor neuron lesion, denervation of intercostal muscles and diaphragm

---

**Diagnostic Procedures**

1. Electromyography (EMG) as an extension of the clinical examination, is the single most useful test in evaluating disease of the lower motor neurons. The EMG will demonstrate abnormalities in upper and lower extremities and thoracic paraspinal muscles and the presence of normal nerve conduction velocities.
2. Muscle biopsy shows the typical appearance of denervation atrophy with atrophic fascicles coexisting with normal fascicles.
3. Muscle enzymes such as creatinine phosphokinase (CK) may be elevated in rapidly progressive cases.
4. The CSF is normal.
5. There are no MRI, CT, or myelographic abnormalities.

*chronic denervation and subsequent reinnervation* leads to increased size of motor unit. denervated fibers are reinnervated by the terminal sprouts of surviving motor neurons. this leads to **EMG of very large motor units** with abnormal spontaneous activity, with **large amplitude**, and **fasciculations**
A fasciculating tongue in a patient with amyotrophic lateral sclerosis, indicates what?
- a) corticobulbar neurons have died
- b) there must be no atrophy
- c) the motor unit must still be intact
- d) increased ACh receptors on the tongue muscle

D. When the patient experiences a neuropathy, often the distal axon will degenerate (Wallerian), thus "denervating" the muscle fibers. During the first 2-4 weeks of nerve degeneration, the muscle fibers become extremely "sensitive" to acetylcholine. In fact, cholinergic (nicotinic) receptors increase on the surface of the denervated muscle fibers and become available to any circulating acetylcholine. Attachment to these receptors by ACh will cause the muscle fiber to twitch. The patients can often sense this occurrence, and there may be a visible "rippling" effect under the skin. Clinicians can place mild finger pressure over the area and palpate the contracting fibers. These potentials are known as "fibrillation" or "denervation" potentials. They are always looked for after needle insertion while the muscle is at rest. It is important to indicate that upon insertion of the needle, there are always a quick series of "insertional fibrillation potentials" due to the obvious irritation of the penetrating needle. However, these quickly dissipate (within seconds), and the muscle returns to its resting state. Pathologic fibrillation potentials (at rest) are generally seen following the acute first 2-4 weeks of nerve injury and warrant quick intervention.

In a victim of advanced ALS, the grasp reflex is present and the anal reflex is absent.
- a) this is possible
- b) this is impossible

What could differentiate ALS from a herniated disc?
- a) negative orthopedic tests
- b) reflex changes
- c) weakness in a limb
- d) change in muscle tone

What could easily differentiate ALS from a herniated disc?
a) weakness of the legs bilaterally  
b) **absence of sensory deficits**  
c) EMG findings of spiked MUAPs  
d) atrophy of the intrinsic hand muscles  

What could easily differentiate ALS from a herniated disc?  
a) abnormal patellar reflex  
b) absence of the abdominal reflex  
c) absence of the cremasteric reflex  
d) **primitive reflexes**  

A 34-year-old white man complained of difficulty buttoning his clothes and holding onto things for the past 3 months.  

**Neurologic examination** disclosed weakness of flexion, extension, adduction, and abduction of the fingers of both the patient’s hands, but the weakness was more marked in his right hand. Atrophy and fasciculations were noted in the hypothenar and interosseous muscles bilaterally, but these also were more marked in his right hand. Patellar and Achilles reflexes were hyperactive.  

**Treatable Diseases to Be Ruled Out**  
Spinal cord tumor  
Herniated disc  
Cervical spondylosis  
Tuberculosis of the spine  
Nutritional neuropathy  
Neurosyphilis  

**Comment:** Note the involvement of the anterior horn cells, leading to weakness, atrophy, and fasciculations, and of the pyramidal tracts, causing weakness and hyperactive reflexes in the lower extremities. Amyotrophic lateral sclerosis usually spares the extraocular muscles, sensory nerves, and sphincters.  

**Synopsis:** Amyotrophic lateral sclerosis is a devastating degenerative disease of the motor horn cells and pyramidal tracts in the brainstem or spinal cord. The etiology is unknown, except in the small number of familial cases. Cases in which the patient presents with purely lower motor neuron disease are labeled **progressive muscular atrophy**, whereas those in which the patient presents with purely upper motor neuron disease are labeled **primary lateral sclerosis**. Whether these two types of cases are distinct entities is debatable. Amyotrophic lateral sclerosis is slowly progressive, with death from respiratory failure in 3 to 4 years in most cases. There is no known care, but riluzole (100 mg daily) may slow the progression of the disease and reduce mortality. Otherwise, treatment is supportive (see Appendix C). Diagnosis is by exclusion of other disorders with nerve conduction velocity studies, electromyography, and MRI of the cervical spine.  

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<table>
<thead>
<tr>
<th>site of lesion</th>
<th>superficial reflexes</th>
<th>pathological reflexes</th>
</tr>
</thead>
<tbody>
<tr>
<td>muscle</td>
<td>normal</td>
<td>absent</td>
</tr>
<tr>
<td>neuromuscular junction</td>
<td>normal</td>
<td>absent</td>
</tr>
<tr>
<td>peripheral nerve or</td>
<td>decreased or absent</td>
<td>absent</td>
</tr>
<tr>
<td>anterior horn cell</td>
<td></td>
<td></td>
</tr>
<tr>
<td>pyramidal tract</td>
<td>decreased or absent</td>
<td>present</td>
</tr>
<tr>
<td>extrapyramidal tract</td>
<td>normal</td>
<td>absent</td>
</tr>
<tr>
<td>cerebellum</td>
<td>normal</td>
<td>absent</td>
</tr>
</tbody>
</table>

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da. they would still have normal amplitude, however before the LMN is affected, the EMG is nearly normal  

denervation would give larger motor units, fibrillation, and a large amplitude, less MUAPs. ↓#MUAP’s with ↑amplitude = neuropathy  

Myopathy would give a low amplitude and increase # of MUAPs  

---  

b. in a form of amyotrophic lateral sclerosis that has only affected the UMN, what would be the findings on EMG?  
a) fibrillations  
b) a larger motor unit  
c) lower amplitude of MUAPs  
d) fewer MUAPs upon resistance  

c. in a form of amyotrophic lateral sclerosis that has only affected the UMN, what would be the findings on EMG?  
a) denervation supersensitivity  
b) **normal amplitude of MUAPs**  
c) decentralization  
d) decreased number of MUAPs  

d. in an early form of ALS, in which only the corticospinal tract is affected, the patient would have a normal:  
a. nerve conduction stays normal basically the whole time. neurons die, but the neurons that are left still have myelin. ALS is not a de-myelinating dz.
<table>
<thead>
<tr>
<th>Question</th>
<th>Option</th>
</tr>
</thead>
</table>
| a) nerve conduction  
b) strength  
c) muscle tone  
d) deep tendon reflexes |        |
| hyperreflexia, seen in ALS, is most likely due to:  
a) myopathy  
b) loss of alpha motor neurons  
c) loss of extrapyramidalis  
d) degeneration of the motor unit | c      |
| in an early form of ALS, in which only the corticospinal tract is affected, the patient would have normal:  
a) reflexes  
b) muscle tone  
c) strength  
d) alpha motor neuron function | d      |
| fibrillations and denervation potentials become evident on the surface of the skin as muscle twitching, and on EMG, in a patient with ALS, when the ______ is affected  
a) motor unit  
b) corticobulbar pathway  
c) patellar reflex  
d) dorsal nerve root | a. When the patient experiences a neuropathy, often the distal axon will degenerate (Wallerian), thus “denervating” the muscle fibers. During the first 2-4 weeks of nerve degeneration, the muscle fibers become extremely “sensitive” to acetylcholine. In fact, cholinergic (nicotinic) receptors increase on the surface of the denervated muscle fibers and become available to any circulating acetylcholine. Attachment to these receptors by ACh will cause the muscle fiber to twitch. The patients can often sense this occurrence, and there may be a visible “rippling” effect under the skin. Clinicians can place mild finger pressure over the area and palpate the contracting fibers. These potentials are known as “fibrillation” or “denervation” potentials. They are always looked for after needle insertion while the muscle is at rest. It is important to indicate that upon insertion of the needle, there are always a quick series of “insertional fibrillation potentials” due to the obvious irritation of the penetrating needle. However, these quickly dissipate (within seconds), and the muscle returns to its resting state. Pathologic fibrillation potentials (at rest) are generally seen following the acute first 2-4 weeks of nerve injury and warrant quick intervention. |
| fibrillation and fasciculation, seen in ALS, is most likely due to:  
a) increased stimulation of the alpha motor neuron by the muscle spindle neuron  
b) increased ACh receptors in the case of denervation  
c) upper motor neuron loss  
d) syringomyelia | b      |
| in the earliest stages of ALS, one may expect to find ______, and in the later stages of ALS, one may expect to find ______, regarding deep tendon reflex grading  
a) <2, <2  
b) >2, >2  
c) <2, >2  
d) >2, <2 | d      |
| hyperreflexia, then hyporeflexia  
>2..........................<2  
UMNL.......................LMNL |        |
| what causes muscle cramps, in a victim of ALS?  
a) NCV of zero  
b) upper motor neuron degeneration  
c) corticobulbar lesion  
d) hypersensitivity of the denervated muscle | d      |
| in ALS, disinhibition of the lower motor neurons is a result of:  
a) CNS descending tract lesion  
b) PNS lesion  
c) loss of the dorsal columns  
d) cerebellar lesion | a      |
| spasticity is due to ______ and clonus is due to ______ | a      |
| a) UMNL, UMNL | d |
| b) LMNL, LMNL | |
| c) LMNL, UMNL | |
| d) UMNL, LMNL | |

**Hoffman reflex is due to _____ and fasciculation is due to _______**

| a) UMNL, UMNL | d |
| b) LMNL, LMNL | |
| c) LMNL, UMNL | |
| d) UMNL, LMNL | |

**A person with amyotrophic lateral sclerosis will demonstrate:**

| a) ataxia | b |
| b) apraxia | |
| c) agnosia | |
| d) aphasia | |

**Both ALS and cauda equina may demonstrate:**

| a) nonspecific hypotonicity of the lower extremities | a |
| b) radiating segmental paresthesiae | |
| c) +Babinski | |
| d) disinhibition of the DTR | |

**A patient with ALS demonstrates upper motor neuron signs, but not lower motor neuron lesion. A lesion of the corticobulbar tract in this patient demonstrates:**

| a) spastic paralysis of the facial muscles | a |
| b) loss of sensation from the face | |