Amyotrophic Lateral Sclerosis

- Death of both upper and lower motor neurons in the motor cortex of the brain, the brain stem, and the spinal cord.
- Majority of ALS cases are sporadic (sALS)
- 15-20% are familial (fALS)

Upper motor neurons

- Motor neurons that originate either in the motor region of the cerebral cortex or in the brain stem and carry motor information down to the lower motor neurons.

- Primary motor cortex or precentral gyrus, is one of the most important areas in the frontal lobe.

- The precentral gyrus is the most posterior gyrus of the frontal lobe and it lies anterior to the central sulcus.

Upper motor neurons

- The pyramidal cells of the precentral gyrus are also called upper motor neurons.
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- The fibers of the upper motor neurons project out of the precentral gyrus ending in the brainstem, where they will decussate (intersect) within the lower medulla oblongata to form the lateral corticospinal tract on each side of the spinal cord.

- neurons connect the brain to the appropriate level in the spinal cord, from which point nerve signals continue to the muscles by means of the lower motor neurons.

Lower motor neuron

- Motor neurons located in either the anterior grey column, anterior nerve roots (spinal lower motor neurons) or the cranial nerve nuclei of the brainstem and cranial nerves with motor function (cranial nerve lower motor neurons).

- All voluntary movement relies on spinal lower motor neurons, which innervate skeletal musclefibers and act as a link between upper motor neurons and muscles.

Amyotrophic Lateral Sclerosis
Pathology

- Degeneration and death of motor nerves
  - Upper Motor Neuron
    - within brain/spinal cord
  - Lower Motor Neurons
    - leaves brain (stem)/spinal cord
- Relatively spared
  - Eye movements and bowel/bladder function

Amyotrophic Lateral Sclerosis
Clinical Presentation

- Lower motor neuron signs
  - Weakness, muscle wasting, hyporeflexia, muscle cramps, fasciculations
- Upper motor neuron signs
  - Spasticity, hyperreflexia, weakness

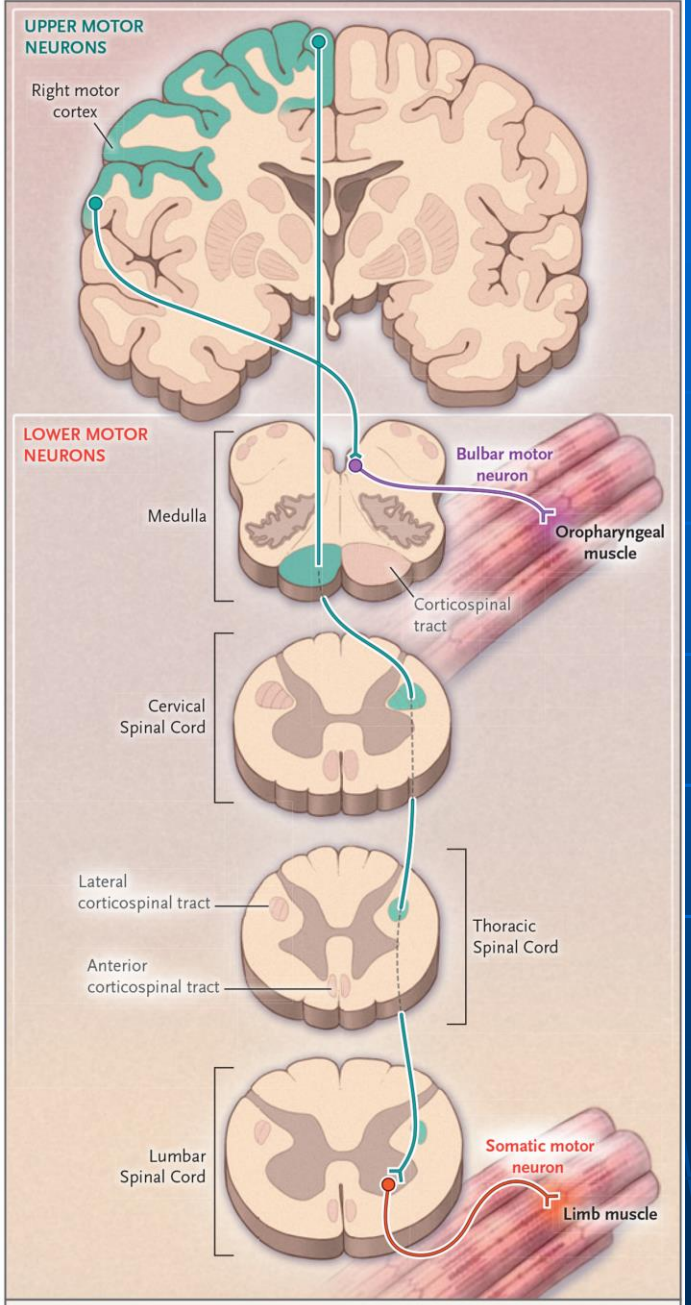
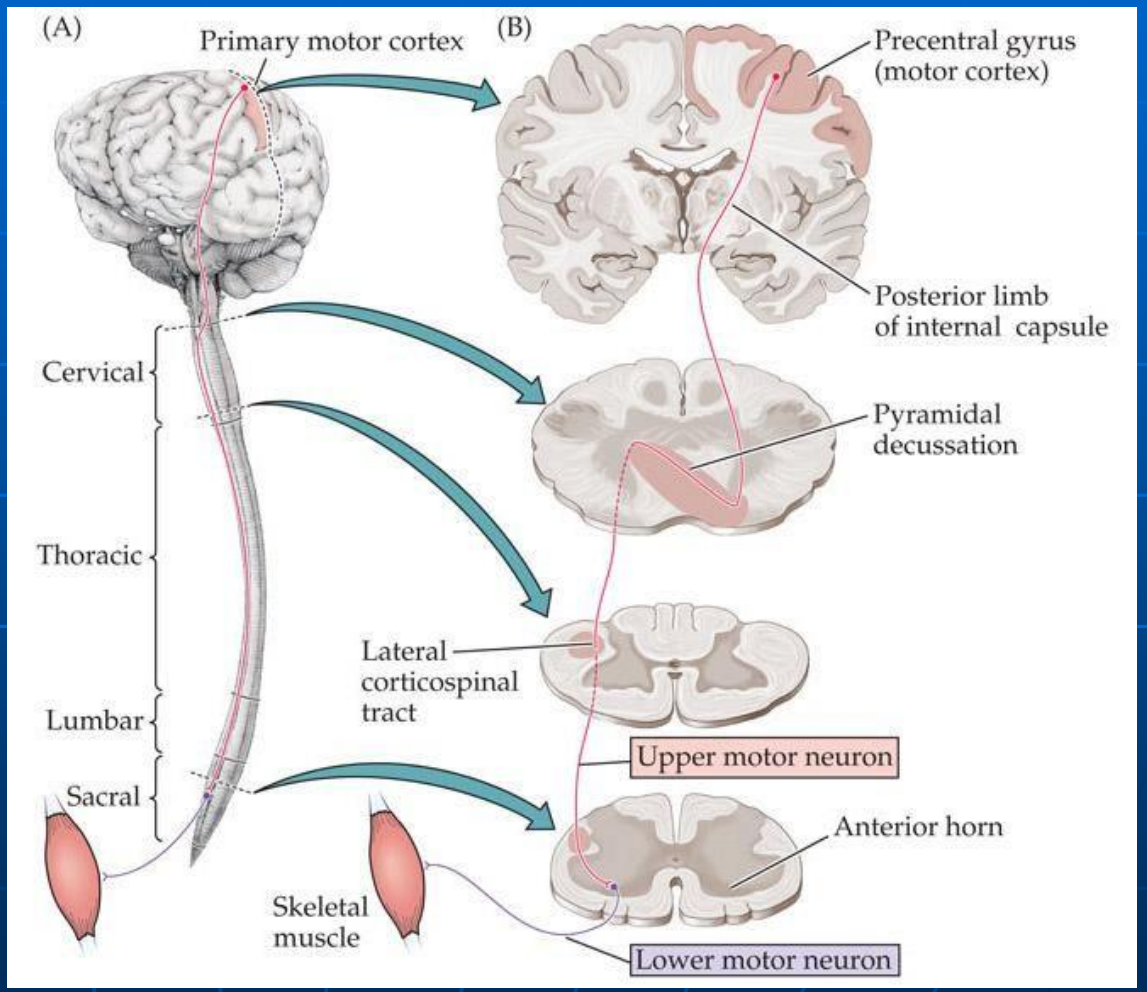
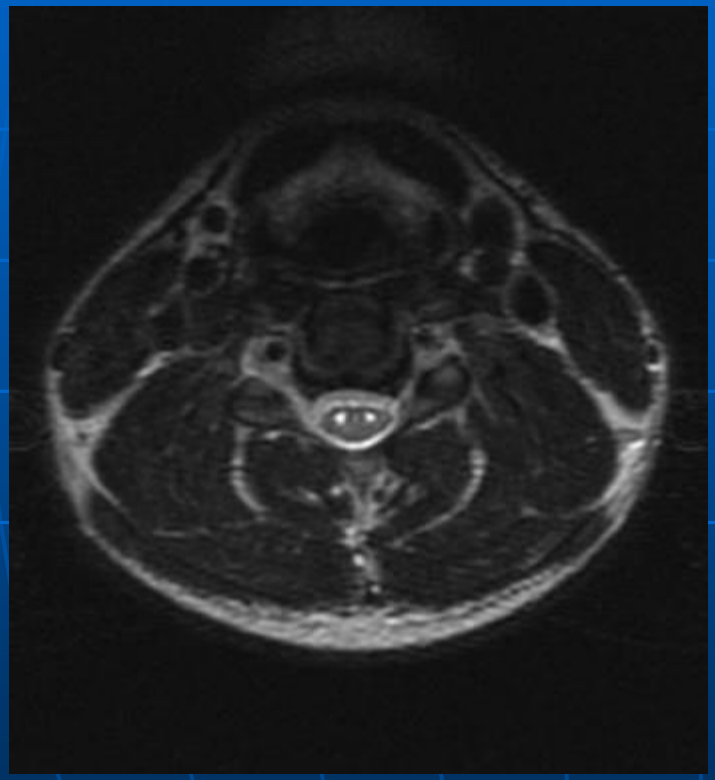


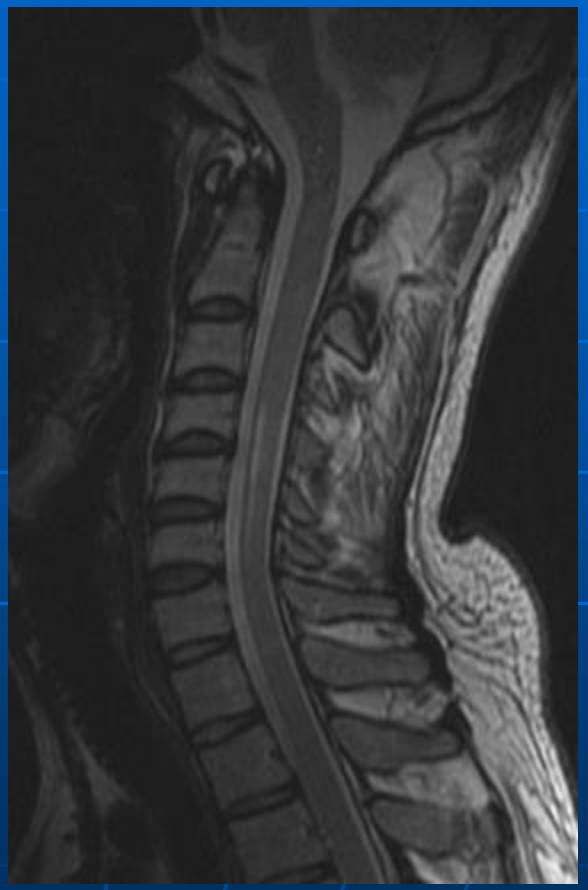
Figure 1. The Motor System.

The motor system is composed of corticospinal (upper) motor neurons in the motor cortex and bulbar and spinal (lower) motor neurons, which innervate skeletal muscle.



ALS



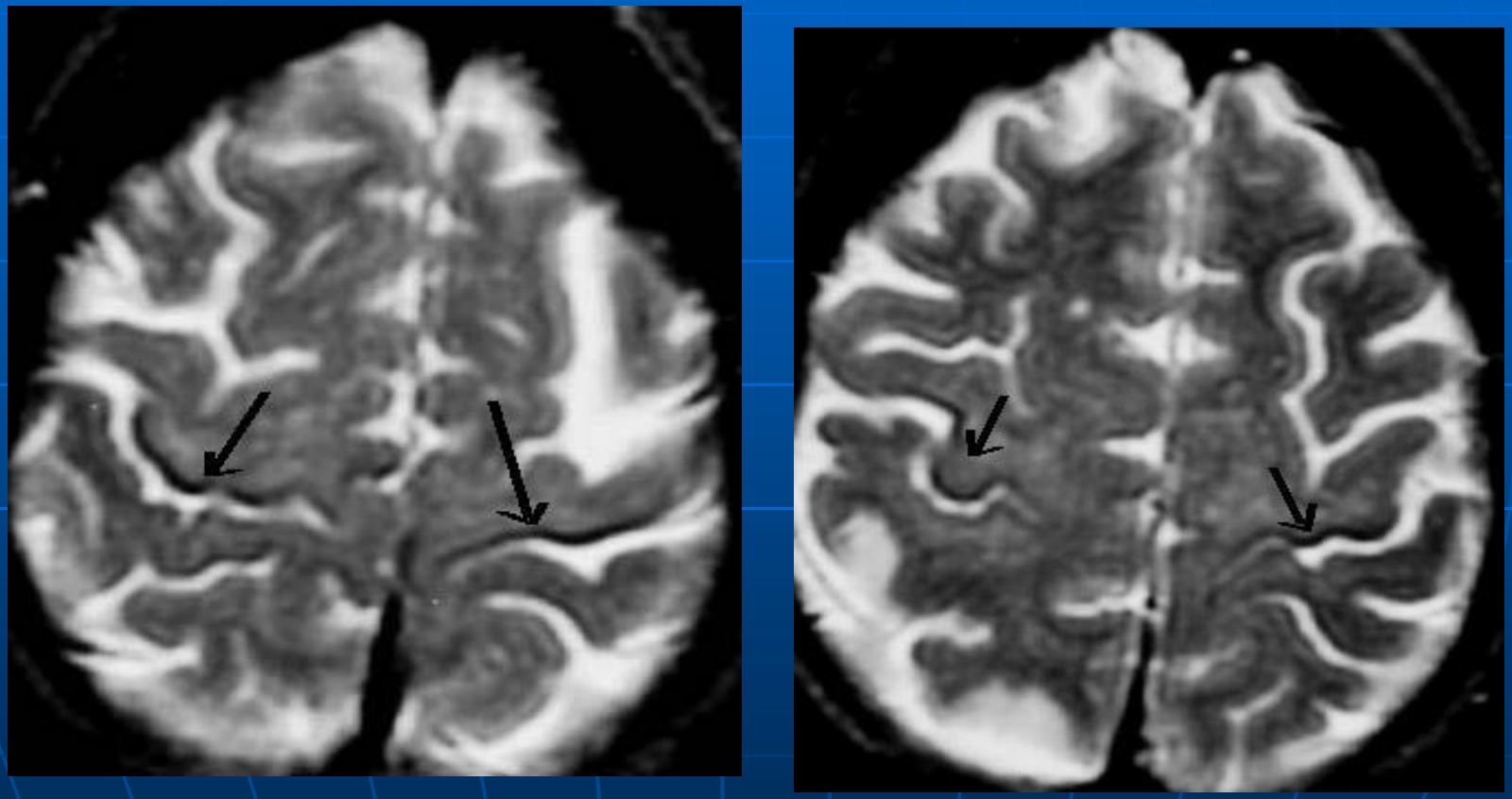


(Snake eyes appearance) = abnormal signal in anterior horn

ALS

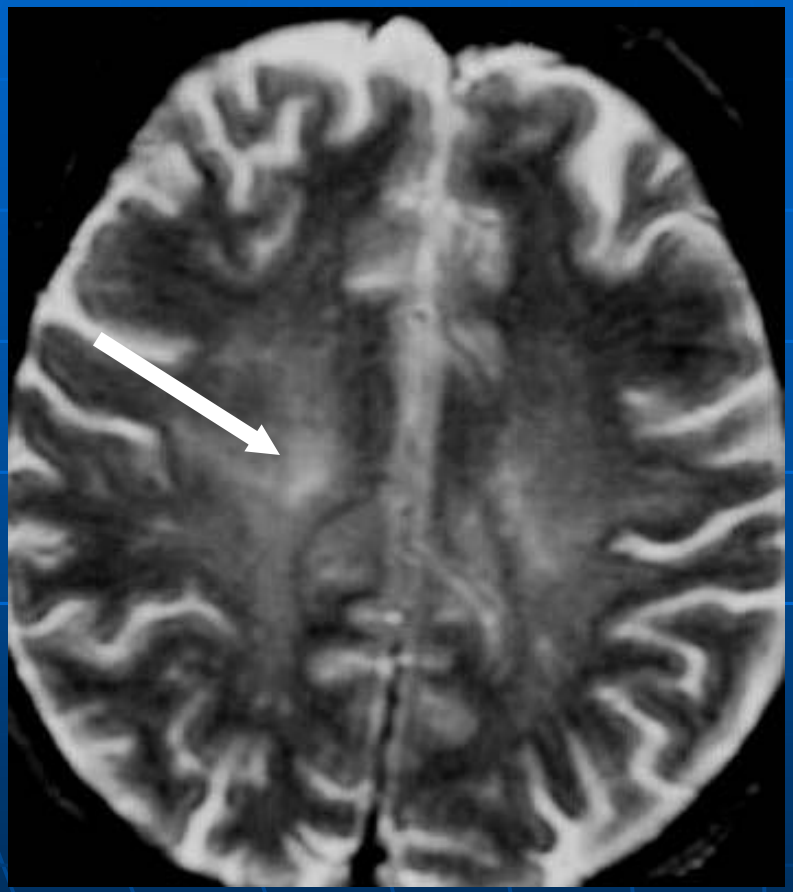
- Bilateral symmetric hyperintense foci along the course of the corticospinal tract from precentral gyrus to the level of the cord, representing myelin loss and gliosis.
- Hypointense signal intensity, typically uniform and laminar, localized in precentral gyrus of the motor cortex on T2 weighted sequences, due to iron or other mineral deposition associated with neuronal degeneration.
- Some other diseases may simulate symmetrical signal changes observed in ALS, but are usually limited to the level of internal capsule, such as progressive multifocal leukoencephalopathy, Wallerian degeneration.
- Adult form of Krabbe disease also simulates ALS by the involvement of pyramidal tracts.

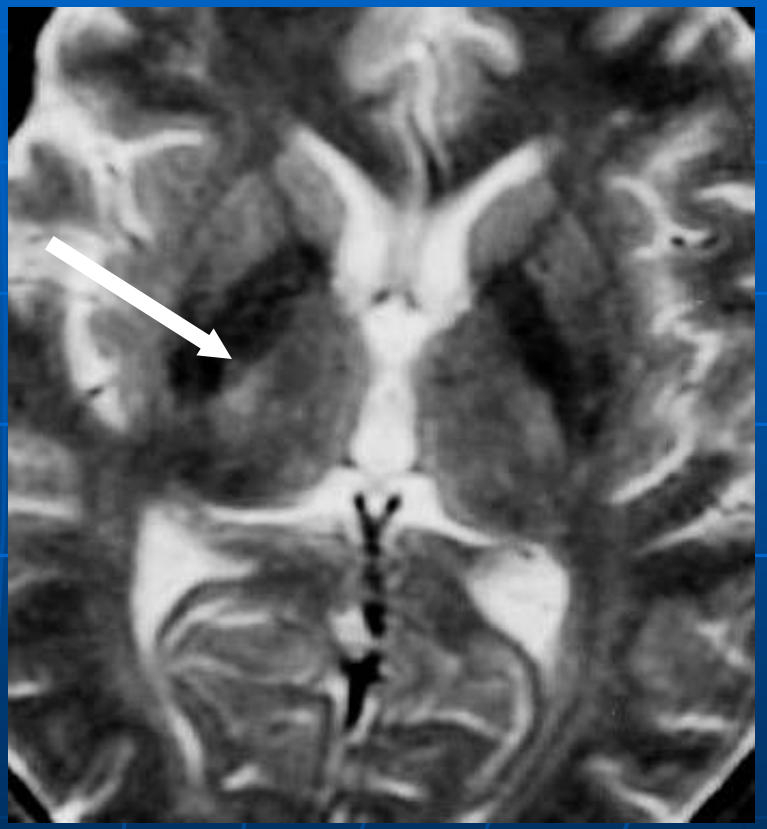
Amyotrophic lateral sclerosis



precentral gyrus reveals hyperintense foci extending along corticospinal tract, as well as hypointense precentral gyri (arrows)

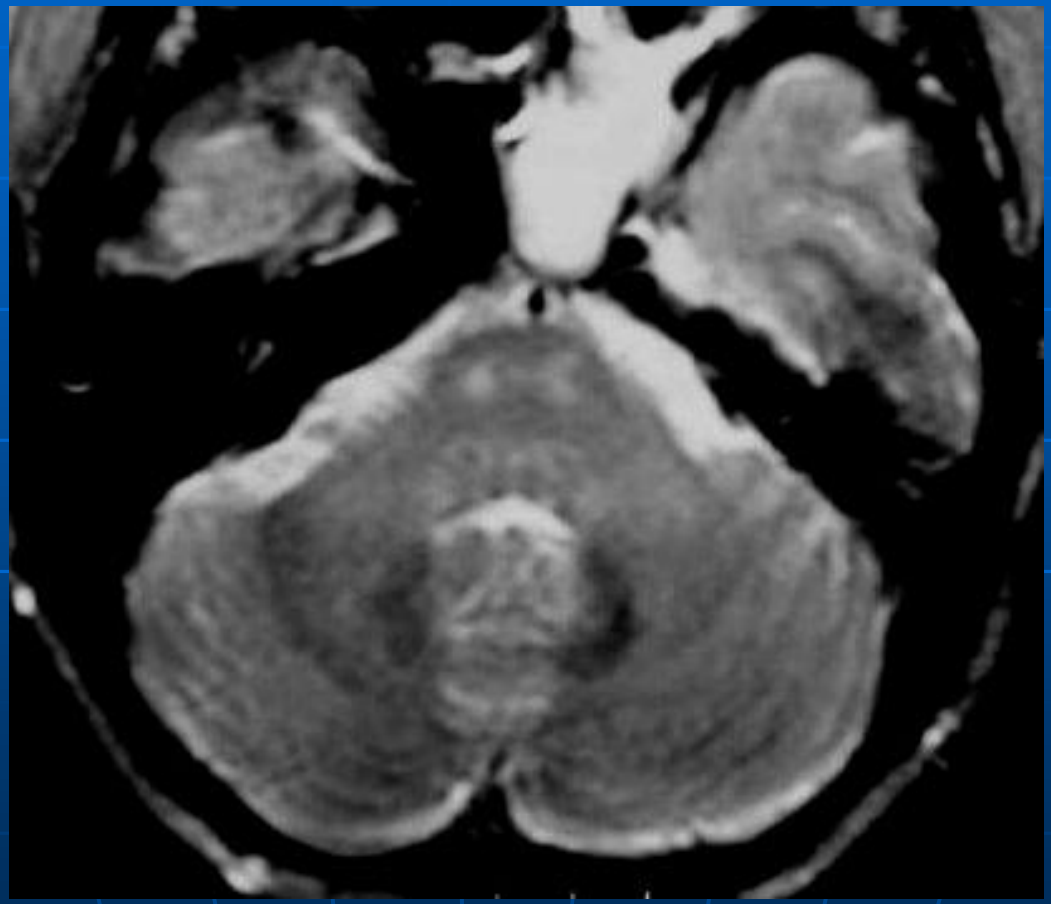
Amyotrophic lateral sclerosis





Increased signal in posterior limbs of internal capsules and corona radiata

Amyotrophic lateral sclerosis



reveals bilateral symmetric hyperintense foci of corticospinal tracts anteriorly.