# Diagnosis of Amyotrophic Lateral Sclerosis

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In a recent article in *Annals* on "Diagnosis of Amyotrophic Lateral Sclerosis," Rowland and associates [1] recommend that "if only lower motor neurons are affected clinically, [use] the term . . . progressive spinal muscular atrophy" and that "the term 'ALS' [should be] reserved for patients who also have definite upper motor neuron signs." Their terminology implies there is a good correlation between upper motor neuron signs and lateral sclerosis ("*Amyotrophy* is another word for neurogenic atrophy of muscle. *Lateral sclerosis* refers to the hardness felt when the spinal cord is examined at autopsy") [2].

Brownell and colleagues [3] reported physical findings and neuropathological abnormalities in 36 patients who had "typical" motor neuron disease. From the patients' medical records, they noted whether the physician had found "upper motor neuron involvement (extensor plantar responses, pathological tendon reflexes)." They examined the spinal cord and brainstem for pyramidal tract degeneration (defined as "myelin pallor"). Eighteen patients had upper motor neuron signs; 16 had pyramidal tract degeneration, 2 did not. One patient with equivocal upper motor neuron signs had pyramidal tract degeneration. Definite or equivocal upper motor neuron signs falsely predicted pyramidal tract degeneration in 11% of patients. Seventeen patients had no upper motor neuron signs. Of these, 6 had no pyramidal tract degeneration and 11 did. For 65% of patients without upper motor neuron signs, the absence of these signs falsely predicted normal pyramidal tracts.

According to Brownell and colleagues' data, the presence or absence of upper motor neuron signs predicts pyramidal tract involvement inaccurately in patients with "typical" motor neuron disease. While it is probably useful clinically to divide patients with motor neuron disease into those with upper motor neuron signs and those without, it is also useful to know that the absence of upper motor neuron signs does not mean normal pyramidal tracts and that the presence of upper motor neuron signs does not always mean there is pyramidal tract degeneration.

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# Naltrexone and Tourette's Syndrome

Gerald Erenberg, MD, and Richard J. Lederman, MD

Kurlan and associates [1] have recently described their findings in patients with Tourette's syndrome (TS) treated with the opiate antagonist naltrexone. We have also used this form of treatment, although in an open study [2].

Our original report was based on the treatment of 7 TS patients with naltrexone at dosages of 100 to 150 mg daily. Since that time, we have treated 2 other TS patients with this drug. All 9 patients were males, and 7 were under the age of 17 years. In our open study, 8 of 9 patients did not report sufficient improvement to warrant continued treatment after a 3-month trial. One 16-year-old had improved dramatically and was maintained on chronic treatment. However, he reverted to his pretreatment level of tics after 3 years of naltrexone.

The patients treated in the study by Kurlan and associates responded with only a mild degree of improvement in tic severity. We would agree with the authors' conclusion that long-term use of this drug in the treatment of patients with TS does not seem warranted at this time. It is certainly hoped that agents that act at more specific opioid receptor sites may produce greater benefit.

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# Treatment of Guillain-Barré Syndrome with Protein-A Immunoadsorption: Report of Two Cases

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Protein-A immunoadsorption (PAIA) is a new therapeutic technique derived from plasma exchange (PE). PAIA allows selective removal of immunoglobulins from plasma of patients through the use of two staphylococcal protein-A–containing columns. The method takes advantage of the ability of protein-A to bind immunoglobulins selectively. This technique has been used in several autoimmune diseases [1, 2]. Guillain-Barré syndrome (GBS) seems to be an autoimmune disorder directed against peripheral nerve myelin. The

 