

HOSPITAL PHYSICIAN®

NEUROLOGY BOARD REVIEW MANUAL

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The *Hospital Physician Neurology Board Review Manual* is a peer-reviewed study guide for residents and practicing physicians preparing for board examinations in neurology. Each quarterly manual reviews a topic essential to the current practice of neurology.

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Neurodegenerative Disorders: Amyotrophic Lateral Sclerosis and Inclusion Body Myositis

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Neurodegenerative Disorders: Amyotrophic Lateral Sclerosis and Inclusion Body Myositis

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INTRODUCTION

Volume 8 of the *Hospital Physician Neurology Board Review Manual* focuses on sporadic degenerative conditions that affect the nervous system. This review addresses 2 conditions: amyotrophic lateral sclerosis (ALS) and inclusion body myositis (IBM).

ALS is a neurodegenerative condition affecting the motor neurons of the motor cortex, brainstem, and spinal cord (**Figure 1**). It is a fatal and relentlessly progressive disorder with eventual severe paresis of all limbs and bulbar muscles in most patients. Average disease duration from onset to death is approximately 3 to 4 years, with approximately 10% of patients surviving more than 10 years.¹ Older age at onset and bulbar or severe respiratory involvement at onset are typically associated with poor prognosis.² ALS is relatively easy to diagnose in advanced stages, when the typical combination of clinical signs develops. Clinical presentation in the early stages of the disease, however, may be non-specific and the diagnosis of ALS may be difficult.

IBM has traditionally been classified as an inflammatory myopathy and in many textbooks and review articles is discussed together with polymyositis and dermatomyositis. Although the cause of IBM remains unknown, IBM has been recognized as clinically and pathologically distinct from polymyositis and dermatomyositis. IBM has features consistent with a degenerative process affecting the skeletal muscle, and the inflammatory reaction in IBM may be an epiphenomenon of the myodegenerative process.^{3,4} As is the case with many degenerative conditions, there is no effective treatment for IBM.

The purpose of discussing ALS and IBM in a single review is that IBM may be misdiagnosed as ALS.⁵ There are several reasons for this:

- Both ALS and IBM are most prevalent in the elderly population.
- ALS and IBM share many clinical similarities, with muscle weakness, atrophy, and dysphagia common

in both conditions (caused in ALS by motor neuron degeneration and in IBM by muscle degeneration).

- Creatine kinase (CK) may be normal or mildly elevated in both ALS and IBM.
- The pattern of electromyographic abnormalities in IBM may be misinterpreted for a primary neurogenic condition such as ALS.^{5,6}
- Patients with IBM may exhibit incidental upper motor neuron (UMN) signs (eg, signs of corticospinal tract involvement caused by cervical spinal stenosis, or ischemic brain changes in elderly patients), which increase the risk of misdiagnosis of ALS.

TERMINOLOGY

The term *amyotrophic lateral sclerosis* was coined in 1874 by Charcot to emphasize the lateral corticospinal tract involvement in this disease.⁷ Many clinicians also use the term *motor neuron disease*, which is essentially synonymous with ALS. The term *motor neuron disease* was introduced into the neurologic literature by Brain in 1933⁸ to indicate the spectrum of clinical presentations of patients with degeneration of UMNs and lower motor neurons (LMNs). This spectrum includes purely LMN involvement (progressive muscular atrophy), typical ALS (“Charcot” ALS), purely UMN dysfunction (primary lateral sclerosis), and purely bulbar involvement (progressive bulbar palsy). In the United States, the term *amyotrophic lateral sclerosis* traditionally has been used to refer to the entire spectrum of patients (those who have ALS, progressive muscular atrophy, primary lateral sclerosis, or progressive bulbar palsy),⁹ whereas the term *motor neuron disease* is used commonly in the British neurologic nomenclature. However, many physicians in the United States also may use the term *motor neuron disease*.¹⁰

An important distinction in terminology must be made between the terms *motor neuron disease* and *motor neuron diseases*. The term *motor neuron diseases*¹¹ is used frequently as an all-encompassing term to apply to a broad category of conditions of various etiologies