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NEUROLOGY BOARD REVIEW MANUAL

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Neuromuscular Junction Abnormalities: Case Studies

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I. INTRODUCTION

Myasthenia gravis (MG) is an autoimmune disorder that affects the neuromuscular junction at the **post-synaptic** level. Although the cause of the disorder is unknown, the role of immune responses (circulating antibodies directed against the nicotinic acetylcholine receptor) in its pathogenesis is well established. The disorder is characterized by fluctuating, fatigable weakness of muscles under voluntary control; some patients may experience symptoms only late in the day or after exercise. Because this disorder is quite treatable, prompt recognition is crucial.

The prevalence of MG in the United States is roughly 14.2 cases per 1 million.¹ Although MG may present at any age, it has a bimodal peak of age at onset. In women, the onset usually occurs between 20 and 40 years of age; among men, the onset is usually at 40 to 60 years of age. Familial occurrence of MG is rare; however, first-degree relatives do have a higher incidence of other autoimmune diseases.²

During the past decade, our understanding of the disease has improved substantially, leading to new modalities of treatment with a substantial reduction in the mortality and morbidity. This review focuses on MG (clinical features, electrodiagnostic testing, and treatment) and also briefly discusses other disorders that

affect the neuromuscular junction (Lambert-Eaton myasthenic syndrome [LEMS], botulism). Two case patients are presented to illustrate different aspects of management of these diseases.

II. CASE PATIENT 1 PRESENTATION

Patient 1 is a 36-year-old woman with a 6-month history of intermittent horizontal diplopia and difficulty swallowing. She indicates that her symptoms worsen as the day progresses. She also has drooping of the left eyelid at times and also reports some headaches. She reports difficulty with swallowing liquids, including occasional gagging, and easily gets fatigued. She denies any sensory problems. Her examination reveals mild left eyelid ptosis that worsens with sustained upward gaze. Her neck flexors/extensors are normal in strength. The remainder of her neurologic examination is unremarkable.

- **Based on the clinical history and examination, the most common signs and symptoms of MG in patient 1 are all of the following EXCEPT:**

- A) Intermittent diplopia
- B) Difficulty swallowing
- C) Eyelid ptosis
- D) Headaches