Lambert-Eaton Myasthenic Syndrome:
UAB (University of Alabama at Birmingham) experience.

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Lambert-Eaton myasthenic syndrome (LEMS) is an autoimmune disease clinically characterized by easy fatigability, proximal leg weakness, paucity of oculobulbar symptoms, and hyporeflexia. LEMS is a presynaptic disorder induced by the antibodies against voltage-gated calcium channels. This antibody was found in 70% of UAB LEMS patients. LEMS is often associated with small cell lung cancer, seen in 60% of our cases. The classic clinical triad of LEMS includes proximal leg weakness, hyporeflexia or areflexia, and cholinergic dysautonomia. A transient improvement in muscle strength and reflexes immediately after brief exercise, the pathognomonic finding of LEMS, was found in 43% of cases according to our experience.

LEMS is a disease of the elderly, with the most common age of onset of symptoms being about 60 years of age. The diagnosis of LEMS can only be confirmed by the repetitive nerve stimulation (RNS) test. It is characterized by the classical triad: (a) low compound muscle action potential, (b) decremental response at low rate stimulation, (c) marked incremental response at high rate stimulation or after brief exercise.
Our study showed that 10 seconds exercise should be the standard test and a more than 60% increment in post-exercise facilitation and at high rate stimulation is sufficient for the diagnosis of LEMS. Our study also showed that the RNS test is more typical of LEMS and more abnormal in the seropositive LEMS group.

Our studies showed that 3,4-diaminopyridine is the drug of choice for symptomatic treatment. Guanidine HCl is an alternative choice of drug, usually with pyridostigmine.