should be performed in a monitored setting with atropine available for potential administration. It is 95% sensitive for generalized myasthenia and 86% sensitive in cases of ocular myasthenia.[379]
The presence of the antiacetylcholine receptor antibody (AChR-Ab-binding, blocking, or modulating) can confirm the diagnosis; however, about 20% of patients with generalized myasthenia and about half of those with ocular myasthenia are seronegative. About one-third of these seronegative patients will be seropositive for muscle-specific kinase (anti MuSKAb). Lipoprotein-related protein 4 (LRP4) has been associated with generalized and ocular myasthenia gravis as well.[380]

Repetitive nerve stimulation testing (positive in only one-third of patients with ocular myasthenia) and the far more sensitive single-fiber electromyography (positive in over 90% of patients with ocular myasthenia) may also assist in diagnosis.[381] In many centers, single-fiber electromyography is considered the gold standard for diagnosis.

MANAGEMENT

Pyridostigmine bromide administered orally two to four times a day is the first line of treatment for myasthenia gravis, but about half of patients with strabismus-associated myasthenia show minimal response. In contrast, about 66% to 85% of patients show a positive response to corticosteroids.[382] For some patients, various forms of immunosuppressive therapy with azathioprine, which is known to be effective, and other agents under current investigation such as efgartigimod alfa-fcab may be offered by treating neurologists. Efgartigimod alfa-fcab has recently been FDA approved for patients who test positive for the anti-acetylcholine receptor. Thymectomy is indicated in some cases, always in the presence of thymoma, and may substantially reduce symptoms for certain subpopulations with myasthenia gravis.[367]

Diplopia and strabismus are highly variable and not readily remedied with prisms. Remission or stabilization of the disease is often possible after 2 to 3 years of treatment,[383] and at that point surgical intervention for strabismus may be considered if desired or if prism use is insufficient. Care is indicated in the use of anesthetic agents given any evidence of associated weakness of the respiratory muscles. Surgical management, with and without the use of adjustable sutures, has met with modest success in cases where there is a stabilized primary deviation, sometimes exacerbated by fatigue.[384-389] More than one strabismus surgery may prove necessary.

PROVIDER AND SETTING

Diagnosis and management of myasthenia gravis require the training and clinical judgment of an experienced ophthalmologist typically working in concert with a treating neurologist.

COUNSELING AND REFERRAL

Counseling and referral to a neurologist or neuro-ophthalmologist, and sometimes a general surgeon, is often indicated in the management of myasthenia. The ophthalmologist should discuss the findings, explain the disorder, provide a diagnosis, and discuss management options with the patient and any caregivers and be aware of any comorbidities such as respiratory distress that might present with generalization of the disease.