

## Neurology/Neurosurgery – Myasthenia Gravis

<b>Definition</b>	Myasthenia Gravis (MG) is a neuromuscular disease leading to fluctuating muscle weakness and fatigability. It is an autoimmune disorder, in which weakness is caused by circulating antibodies that block acetylcholine receptors at the post-synaptic neuromuscular junction, inhibiting the stimulative effect of the neurotransmitter acetylcholine. Two forms of MG are recognized, congenital and acquired. Acquired MG is a disease seen secondary to the activation of antibodies against the post-synaptic acetylcholine receptors of skeletal muscles. The action of these antibodies results in impairment of action potential transmission from nerve to muscle
<b>Signalment and Clinical Signs</b>	A variety of clinical forms of acquired MG have been identified in the last few decades. The classic clinical presentation of a dog with acquired MG is episodic, generalized muscle weakness that is worsened by activity and ameliorated with rest. The pelvic limbs are preferentially affected in the majority of dogs with MG and megaesophagus is a common finding. Varying degrees of weakness of esophageal, facial, pharyngeal and laryngeal muscles with or without involvement of appendicular muscles may be seen in dogs with MG. Pharyngeal and laryngeal muscle dysfunction with or without megaesophagus may lead to aspiration pneumonia. Although acquired MG in dogs is a disorder with a wide spectrum of clinical forms, we tend to classify MG in three distinct categories: 1) the “classic” form with exercise-associated weakness of the appendicular muscles, 2) the focal form with megaesophagus and possible laryngeal and pharyngeal dysfunction, and 3) the fulminant form with rapid development of lower motor neuron type weakness. The clinician must be aware that the fulminant form of acquired MG may mimic the clinical signs seen in tick paralysis, polyradiculoneuritis, and botulism.
<b>Diagnosis</b>	<p>Edrophonium chloride (Tensilon, Enlon), a short acting anticholinesterase drug, is given intravenously at a dose of 0.11 to 0.22 mg/kg for diagnostic purposes. This drug usually produces a dramatic improvement in an animal collapsed in myasthenic crisis. Within one to two minutes the animal is up on its feet walking around with renewed strength. The strength only lasts one to three minutes and weakness returns. Megaesophagus may be seen on chest radiographs.</p> <ul style="list-style-type: none"> <li>• Contrast radiographic studies may be required for the identification of megaesophagus.</li> <li>• The electromyogram and peripheral nerve conduction studies are usually normal. On repetitive nerve stimulations, a decremental response is seen that is most characteristic of myasthenia gravis. The decremental response tends to disappear when edrophonium chloride is given.</li> <li>• The gold standard for the diagnosis of acquired MG however is an elevated serum acetylcholine receptor antibody titer.</li> </ul>
<b>Treatment</b>	Medical therapy for acquired MG is usually an oral long-acting anticholinesterase such as pyridostigmine (0.5-3.0 mg/kg PO every 8 to 12 hours). Immunosuppressive doses of glucocorticosteroids (or other agents) may be indicated in severe cases. Caution should be taken with administration of immunosuppressive treatment as this may exacerbate the development of aspiration pneumonia. Once the animal is stabilized, the anticholinesterase may be slowly decreased. Thymomas may be associated with acquired MG. Thymectomy may be an advocated surgical therapy in the presence of this tumor.
<b>Prognosis</b>	A number of dogs with acquired MG will achieve remission from the disease. Checking the levels of antibodies to anticholinesterase receptors prior to treatment and at regular intervals during treatment is important to monitor effectiveness of treatment and impending recurrence throughout the lifespan of affected dogs. Dogs that acquire MG later in life usually have a fairly good prognosis with treatment. Dogs can be maintained well in remission for a long time with this disease. The more attention paid to prevention of aspiration pneumonia, the better the long term prognosis. Many medications interfere with the actions of the anticholinesterase drugs, so careful thought must be given to the choice of medications to treat other problems in affected dogs.
<b>Questions? Neurology/Neurosurgery Expert: Peter Gordon, DVM, DACVIM (Neurology)</b>	The Veterinary specialty Center of Tucson has a board-certified neurologist available for questions and consultations during the weekdays. Dr. Gordon is on-call 24/7 to provide consultations to VSCT emergency doctors and to perform emergency diagnostics and surgery for patients seen by the VSCT emergency service. Board-certified neurologists have four additional years of training and are certified by the American College of Veterinary Internal Medicine to assure competency in neurology/neurosurgery.