

Myasthenia Gravis - When the Muscles & Nerves Don't Talk

Nerves send vital signals to muscles throughout the body. When these chemical signals don't interact with muscles normally, one possible result is profound muscle weakness. A unique disease caused by this very scenario is called myasthenia gravis, and has been documented in dogs, cats, and humans. This week I share some helpful information about this interesting ailment. Happy reading!



Myasthenia Gravis - What is it?

Muscles are controlled by nerves, but nerves don't directly connect to the muscles. There is a small gap between them - this is called a neuromuscular junction. Electrical signals travel through nerves until they reach the neuromuscular junction, and somehow the signal must jump the gap from nerve to muscle. This jump is facilitated by a chemical messenger called acetylcholine. This chemical is released from the nerve, flows across the neuromuscular

junction, and attaches to unique receptors on the muscle like a key fitting into a lock. In patients with myasthenia gravis, the communication between nerve and muscle is abnormal. To view a high-level review of the the neuromuscular junction, please watch the video below.

There are two forms of myasthenia gravis:

- Congenital - Patients aren't born with enough acetylcholine receptors. To use the lock & key analogy mentioned above, these patients are born with too few locks
- Acquired - The immune system produces special proteins called antibodies that destroy acetylcholine receptors. The locks are destroyed by the immune system

Myasthenia Gravis - What does it look like?

Patients with the congenital myasthenia gravis generally show clinical signs by 6-8 weeks of age, particularly generalized weakness and occasionally muscle tremors. This weakness often progresses and may ultimately lead to death. Certain breeds are hereditarily predisposed to the congenital form, including:

- Dachshunds
- Smooth fox terriers
- Springer spaniels
- Jack Russell Terriers

The acquired form is documented later in life, typically between 2-4 years of age and 9-13 years of age. Clinical signs may be as non-specific as generalized weakness with affected pets showing intolerance to exercise that improves with rest. Severely affected patients may too weak to lift their heads. The video below shows a dog who hindlimb weakness especially after taking a few steps.

Sometimes only a single muscle (or group of muscles) is affected, most commonly muscle groups in the mouth and throat. These patients may drool excessively, have difficulty swallowing, experience labored breathing, have a voice change, and/or regurgitate food and water. When the esophagus (the tube that connects the mouth to the stomach) is affected, then it becomes markedly dilated. This

condition is called [megaesophagus](#).

Myasthenia Gravis - How is it diagnosed?

Your family veterinarian will recommend some screening blood and urine tests after reviewing your pet's complete medical history and performing a thorough physical examination. These tests are:

- Complete blood count - non-invasive blood test that measure red blood cells, white blood cells, and platelets
- Biochemical profile - non-invasive blood test to evaluate kidney & liver function, as well as to measure electrolytes (i.e.: sodium, potassium) and certain gastrointestinal enzymes
- Urinalysis - non-invasive urine test to help evaluate kidney function and look for infection
- Chest radiographs / x-rays - a non-invasive imaging test that looks for evidence of cancer, pneumonia, and dilation of the esophagus

These screening tests are profoundly important to look for other diseases that could cause similar clinical signs. Ultimately the best test to diagnose myasthenia gravis in dogs and cats is the acetylcholine receptor antibody assay. This is a non-invasive blood test that measures the quantity of antibodies that may be attached the acetylcholine receptor (the lock). Depending on the result, additional testing may be recommended to support a diagnosis of myasthenia gravis:

- Edrophonium/Tensilon® challenge - Edrophonium chloride is a very short-acting antidote for myasthenia gravis. Upon injection into a vein, affected patients temporarily regain their strength. Watch the video below to a dog with myasthenia gravis improve after an injection of edrophonium
- Electromyogram - This is a minimally invasive test that delivers a small electrical signal to muscles and then evaluates how the muscles respond.
- Muscle biopsy - Evaluating a muscle sample for acetylcholine receptors is important in patients with congenital myasthenia gravis

Pet parents may find it uniquely helpful to partner with a board-certified veterinary internal medicine and neurologist to develop a logical and cost-

effective diagnostic plan.

Myasthenia Gravis - How is it treated?

There are many potential therapies to improve the quality of life of patients with myasthenia gravis. Some of these address the disease itself, while other address complications associated with it. Affected patients are classified into one of three groups:

- Group 1 - focal or mild generalized
- Group 2 - moderate generalized
- Group 3 - severe generalized

Patients in groups 1 and 2 are frequently treated with medicines that make acetylcholine stay in the neuromuscular junction longer and prevent the immune system from forming antibodies against the acetylcholine receptor. The former drugs are called anticholinesterase medications and the latter are immunomodulatory agents, of which there are several types. Pyridostigmine bromide/Mestinon® and neostigmine bromide/Prostigmin® are classic anticholinesterase drugs. Prednisone is the prototypical immunomodulatory.



Image courtesy of
<http://www.westward.com>

Patients in group 3 are quite challenging to treat, and are best managed in an intensive care unit. Affected patients typically need help breathing, and are placed on a device called a mechanical ventilator. Some referral hospitals offer therapeutic plasma exchange. During this treatment, a patient's plasma that contains the dangerous antibodies against the acetylcholine receptor is removed and then replaced with donor plasma.

Various supportive therapies, including fluid and nutritional support, as well as medications to treat aspiration pneumonia and support the gastrointestinal tract, may be needed in certain patients. Partnering with a board-certified veterinary internal medicine specialist or emergency and critical care specialist will be helpful to ensure your fur baby receives the most appropriate care.

The take-away message about myasthenia gravis...

Myasthenia gravis is an important cause of exercise intolerance and weakness in dogs and some cats. A common complication of this condition is megaesophagus with subsequent aspiration pneumonia. Early identification and treatment are essential to maximize the likelihood of an affected pet being able to lead a high quality of life.

To find a board-certified veterinary internal medicine specialist and neurologist, please visit the [American College of Veterinary Internal Medicine](#).

To find a board-certified veterinary emergency and critical care specialist, please visit the [American College of Veterinary Emergency and Critical Care](#).

Wishing you wet-nosed kisses,

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