

Owner Information Sheet – *Myasthenia gravis*

Background

Myasthenia gravis is a disease that affects the chemical junction between the peripheral nerves in the body (those outside the spinal cord and brain) and the skeletal muscles they innervate. This causes a blockage of the messages between the nerves and muscles, resulting in the muscles becoming weaker, particularly when an animal exercises. There are two forms of myasthenia gravis: a rare congenital form and a more common acquired form. Congenital myasthenia gravis occurs in very young animals and has been described in certain dog breeds, including English springer spaniels, Labrador retrievers, dachshunds, Jack Russell terriers and smooth fox terriers. It has also been described in certain cat breeds, including the Devon Rex and Sphynx. Acquired myasthenia gravis is typically seen in middle-aged or older animals and is caused by an inappropriate response of the animal's immune system (autoimmune disease). The trigger for this inappropriate immune response is typically unknown, but can be related to a concurrent disease, such as underlying cancer. Acquired myasthenia gravis can occur in any breed of dog and has also been reported in cats, with Abyssinian cats being overrepresented.

Cause

Animals with congenital myasthenia gravis are generally born without specific receptors on the muscle membrane. In healthy animals, these receptors initiate muscle contraction by binding a chemical neurotransmitter released from the nerves called acetylcholine. In the acquired form of the disease, the acetylcholine receptors become the target for *autoantibodies* (antibodies produced by an animal against its own body) due to malfunction of the immune system. As a result, transmission between the peripheral nerves and the skeletal muscles is decreased or completely blocked, the muscles cannot contract properly, and the animal becomes weaker and weaker. This weakness is particularly apparent at times of activity when the muscles cannot meet the increased demand of exercise.

Clinical signs (symptoms)

The most characteristic clinical sign is *exercise intolerance* and affected animals are not able to perform physical activities that may be effortless for a healthy individual. After a period of rest, the muscle function can go back to normal, therefore the generalised muscle weakness may be episodic and the neurological examination between episodes can be normal. The muscles of the face can also be affected, resulting in a lack of blinking in response to touching the face, and the muscles that power the oesophagus (the tube between the throat and stomach) are commonly affected in dogs. Weakness of the oesophageal muscles results in a condition called *megaesophagus*, in which the oesophagus becomes distended and an animal will show frequent passive regurgitation of undigested food and water. This can be particularly dangerous as it predisposes to the aspiration of food and water the lungs, resulting in aspiration pneumonia. The clinical signs can be more variable and difficult to recognise in cats, particularly as they are a more sedentary species that we do not take on walks. Typical signs in cats include generalised weakness, a stiff gait, reluctance to jump and a low head carriage.

Diagnosis

A clinical suspicion for myasthenia gravis can sometimes be made based on the characteristic clinical signs, patient breed, age and history, and by exclusion of other causes of generalised weakness, such as cardiovascular or metabolic diseases. To exclude an underlying tumour as a trigger for acquired myasthenia gravis, chest X-rays and an abdominal ultrasound scan are frequently performed. The chest X-rays are also essential to look for evidence of megaesophagus or aspiration pneumonia. In the acquired form, a blood test can be taken to detect an increased level of antibodies against the acetylcholine receptors. This test is not 100% sensitive but can usually confirm a diagnosis in approximately 98% of cases. Electrophysiological tests can also be performed under general anaesthesia to support the diagnosis. Drugs that temporarily increase the concentration of acetylcholine at the chemical junction between the nerves and skeletal muscles (neuromuscular junction) can also be given intravenously ('Tensilon test') and the animal's response monitored; a rapid but short-lived improvement directly after drug administration would support the diagnosis. However, there are some risks to consider when performing this test. A definitive diagnosis of congenital myasthenia gravis can only be made by analysis of a muscle biopsy or a genetic test (blood test).

Treatment and prognosis

The treatment of acquired myasthenia gravis in dogs primarily focuses on increasing the concentration of acetylcholine at the neuromuscular junction to stimulate the remaining receptors that are not bound by antibodies. Treatment is usually long term (months to years) and may include gradually increasing or decreasing the dose of medication, together with monitoring of the animal's clinical signs and the levels of antibodies in the blood. If this is not effective, then silencing the overstimulated immune system by the careful use of immunomodulatory drugs (such as corticosteroids) can also be performed. However, there are potential risks associated with the use of corticosteroids in these cases such as the exacerbation of weakness and an increased risk of pneumonia. Filtering the antibodies from the blood, as performed in humans with myasthenia gravis, is very expensive and is not widely available for animals at present. If possible, treatment of any underlying triggers for the disease is recommended (i.e. surgical resection of the tumour or chemotherapy). A vital part of treatment is the management of megaesophagus, if present, by careful feeding and prompt recognition and treatment of pneumonia. The presence of megaesophagus is a negative prognostic factor because of the increased risk of developing aspiration pneumonia. Therefore, the prognosis in the early stages of the disease can be guarded due to recurrent bouts of pneumonia, requiring repeated visits to the vet. However, in animals that respond well to the therapy and that do not develop complications, the overall prognosis can be more promising.

The prognosis of congenital myasthenia gravis depends upon the breed affected as sadly some forms of congenital myasthenia gravis cannot be successfully treated.

Your veterinary neurologist or primary care veterinarian will discuss in more detail with you the prognosis for recovery and the expectations of treatment on an individual basis.

If you have any concerns about your dog or cat, or their treatment, do not hesitate to contact your veterinarian.

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The Granary, Bunstead Barns, Poles Lane,
Hursley, Winchester, Hampshire, SO21 2LL.

Tel: 01962 767920 Fax: 01962 775909

Email: info@andersonmoores.com

www.andersonmoores.com

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