Myasthenia gravis in the canine patient

Ilaria Vercesi BSc (Hons) RVN, RCH

Neurology & Rehabilitation Department, Queen Mother Hospital for Animals, Royal Veterinary College, London, UK

ABSTRACT: Myasthenia gravis is a relatively common neurological disease affecting the neuromuscular transmission in dogs. Patients with myasthenia gravis often develop secondary pathologies that hugely affect prognosis. Due to this, myasthenic patients require intensive supportive care. This article discusses the causes, diagnosis, treatment and nursing consideration of the myasthenic patient, in order to better patient outcomes.

Keywords: neurology nursing; myasthenia gravis; megaoesophagus; aspiration pneumonia; nursing care; rehabilitation

Ilaria Vercesi BSc(Hons) RVN, RCH

Ilaria qualified as a veterinary nurse from the Royal Veterinary College London in 2015. She holds a Level 3 Certificate in canine hydrotherapy. Ilaria currently works in the Neurology and Rehabilitation department at the Queen Mother Hospital for Animals.
Email: ivercesi2@rvc.ac.uk

Myasthenia gravis (MG) is a relatively common neurological disease affecting neuromuscular transmission in dogs, and less commonly cats (Platt & Shelton, 2013). The disruption of neuromuscular transmission occurs when acetylcholine (ACh) released from the presynaptic vesicles present at the axon terminal of the motor neuron is unable to bind to receptors on the endplate of the muscle fibre membrane. This causes progressive weakness and fatigue, characteristic of MG in dogs (Richmond, 2015). There are two forms of MG: congenital and acquired.

Congenital MG (CMG)

CMG presents as early as 6-8 weeks of age, and is rare in cats and dogs (Platt & Shelton, 2013). It occurs due to an abnormal deficiency of ACh receptors on the postsynaptic muscle fibre membrane. Patients with CMG present progressive weakness, exacerbated by exercise (Penderis & Martin-Vaquero, 2016). CMG has been described as a recessive genetic disease in Smooth Fox Terriers, Jack Russel Terriers and Springer Spaniels. Minature Dachshunds can have a reversible form of CMG, which resolves at 6 months of age (Platt & Shelton, 2013).

Acquired MG (AMG)

In its acquired form, MG is an autoimmune condition in which antibodies are produced against the postsynaptic ACh receptors of skeletal muscles, resulting in reduced numbers of receptors on the end plate. This reduction of ACh receptors is characterised by episodic, generalised muscle weakness, similarly to patients with CMG (Dewey et al., 1997). AMG has been reported in dogs 7 weeks – 15 years of age, of both genders. Golden Retrievers, German Shepherds, Dachshunds, Scottish Terriers, Akitas, German Shorthaired Pointers and Chihuahuas have been reported as the breeds most predisposed (Platt & Shelton, 2013). AMG is the most common neuromuscular disorder diagnosed in the dog, and presents with three main clinical forms (Platt & Garosi, 2012).

- Focal AMG is characterised by facial, pharyngeal, laryngeal and oesophageal muscle weakness. Subsequently, the patient may present with a reduced palpebral reflex, respiratory stridor, regurgitation, decreased gag reflex and/or megaoesophagus (Platt & Garosi, 2012; Richmond, 2015). Appendicular weakness is not present in this clinical form. Reported in 26-43% of all cases of MG (Platt & Shelton, 2013).
- Generalised AMG presents with severe exercise induced appendicular muscle weakness, with or without focal signs and megaoesophagus. This form affects up to 57% of dogs with MG (Platt & Shelton, 2013).
- Acute fulminating AMG is defined by a rapid onset of appendicular weakness, leading to non-ambulatory tetraparesis and impaired respiration. Varying degrees of focal signs may also be present. These patients often develop aspiration pneumonia due to a combination of respiratory muscle weakness, megaoesophagus and laryngeal/pharyngeal dysfunction (Platt & Garosi, 2012; Richmond, 2015).

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AMG can also be paraneoplastic, specifically associated with a thymoma. Complete removal of the mass has been associated with the resolution of clinical signs (Platt & Shelton, 2013).

Diagnosis

Diagnosing the MG case can be challenging as presentation and clinical signs may be similar to a variety of diseases including diseases of the neuromuscular junction, muscle and peripheral nervous system (Penderis & Martin-Vaquero, 2016).

Antibody titres

Antibody titres are the gold standard diagnostic tool for MG (Richardson, 2011). The diagnosis of MG can be confirmed by determining the presence of autoantibodies against muscle ACh receptors in a serum sample (Shelton, 2002). Reactive antibodies are present in approximately 98% of dogs with AMG (Platt & Shelton, 2013). Rarely, patients with recent onset clinical signs may test negative, therefore re-testing may be suggested (Platt & Shelton, 2013). A disadvantage of antibody titres is that they are costly and results may take up to a few weeks (Brooks, 2018). For this reason, other presumptive diagnostic tests may be performed.

Edrophonium challenge

A presumptive diagnosis of MG can be made with an edrophonium challenge (also referred to as the Tensilon test). A short acting anticholinesterase drug, such as edrophonium chloride (Tensilon, Mylan Institutional), can be administered intravenously to the patient with suspected MG (Richardson, 2011). This allows for acetylcholine to gather in the neuromuscular junction, thus improving muscle strength. The response to this test is rapid, and dogs can be observed standing, ambulating and often running. However this effect is short lived and the weakness reoccurs rapidly (Brooks, 2018). A negative test does not rule out MG, as some patients have an insufficient number of ACh receptors, and thus do not show a positive response to the edrophonium challenge. It is important to be aware that this test can cause a cholinergic crisis, where the patient develops bradycardia, hypersalivation, dyspnoea and cyanosis. This can be reversed with atropine (Atrocare, Animalcare) (Platt & Shelton, 2013).

Electromyography

Electromyography (EMG) may also be used to make a presumptive diagnosis of MG. A decrease in the amplitude of the compound muscle action potential in response to repetitive nerve stimulation can be observed in the patient with MG. A limitation of EMG as a diagnostic indicator is that it requires an often lengthy general anaesthetic, therefore it may be contraindicated in the critical patient (Richardson, 2011).

Treatment

Anticholinesterase agents (acetylcholinersterase inhibitors)

The use of anticholinesterase agents is the first line of treatment for MG (Richardson, 2011). They act by increasing the action of ACh in the neuromuscular junction, enhancing the neuromuscular transmission (Bexfield et al., 2006). The drugs available include pyridostigmine bromide (Mestinon, Bausch Health Companies) and neostigmine bromide (Prostigmin, Valeant Pharmaceuticals). They are started at the lowest possible therapeutic dose and gradually increased until the desired effect is reached (Richmond, 2015). Whilst trying to ascertain the correct dose, side effects of excessive levels of anticholinesterase agents may be noticed. The most common side effects include bradycardia, hypersalivation, retching, vomiting and diarrhoea (Richmond, 2015).

Immunosuppressive drugs

If muscle weakness is not adequately controlled by anticholinesterase agents, or if the side effects are intolerable, low dose corticosteroid treatment should be considered (Platt & Shelton, 2013). Immunosuppressive doses of corticosteroids such as prednisolone (Prednicortone, Dechra), azathioprine (Imuran, Aspen Pharmacare) and cyclosporine (Atopica, Novartis) have all been associated with a positive response in patients with MG (Bexfield et al., 2006). However, these drugs can initially exacerbate muscle weakness if used early in the disease (Dewey et al., 1999). The concurrent use of immunosuppressive drugs is contraindicated in MG patient where aspiration pneumonia is already present, as they are usually being treated with antibiotics (Platt & Shelton, 2013).

Antibiotics

The use of antibiotics is often warranted to treat aspiration pneumonia (Richardson, 2011). The antibiotic should be selected depending on the culture and sensitivity results obtained by broncho-alveolar lavage or trans-tracheal wash (Richmond, 2015).

Nursing considerations

Patients with MG present showing a diversity of clinical signs, therefore the nursing care provided to these patients may vary hugely, dependent on the patients clinical presentation, their response and financial implications (Richmond, 2015).

Nutrition

The development of megaoesophagus is common in patients with MG, therefore supportive treatment is essential (Figure 1).

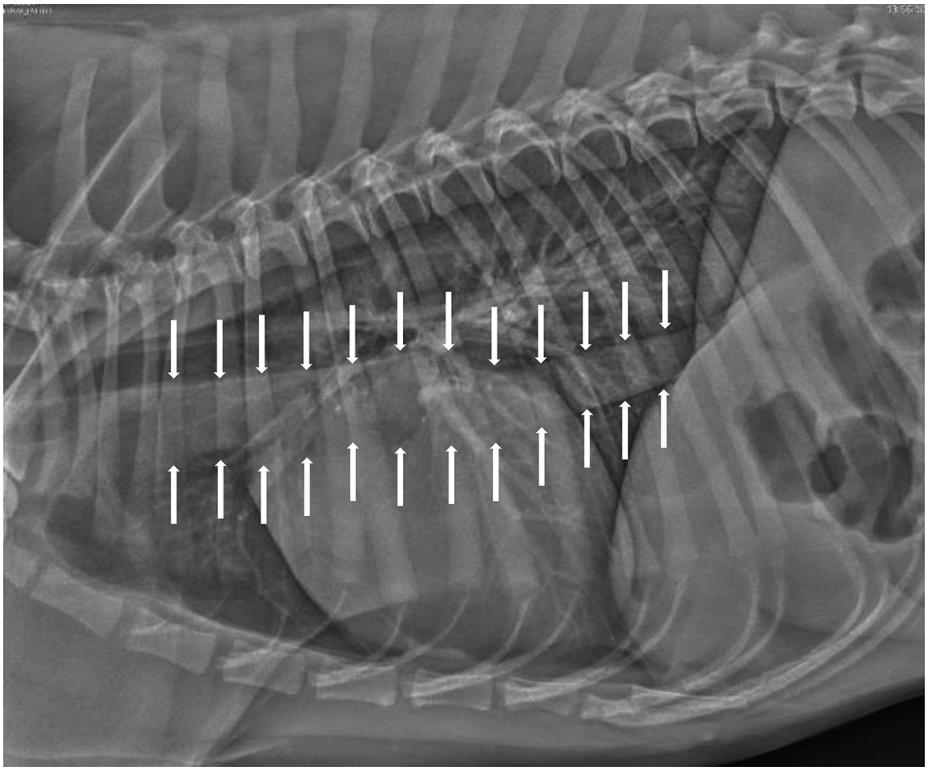

Figure 1. Radiograph of a 9 month old cross breed with unconfirmed AMG. The radiograph confirmed megaoesophagus.

These patients often suffer with regurgitation and hypersalivation, putting them at higher risk of developing aspiration pneumonia. Because of this, managing their dietary intake is of huge importance.

Elevated feeding should be implemented in the patients that maintain some degree of ambulation to aid their limited oesophageal function. In the canine patient, a feeding chair such as the 'Bailey chair' can be used to keep the patient in an upright position for feeding and watering (Figure 2). The patient should be fed small balls of soft food, ensuring that they are swallowing each ball before offering the next. The patient should be kept in the chair for 15 minutes following the feed, to allow the food to move into the stomach. This method of feeding is usually well tolerated, with patients quickly associating the chair with a positive experience.

In the patients where regurgitation and hypersalivation become unmanageable, the use of a feeding tube should be considered. Due to poor oesophageal function, regurgitation may still occur with a nasogastric or oesophageal tube, therefore a gastrostomy tube, specifically a percutaneous endoscopic gastrotomy (PEG) tube, would be best suited to the MG patient. The advantages of a PEG tube are many, including the reduced risk of aspiration pneumonia. Furthermore, the tube helps ensure that the patient is receiving its entire resting energy requirement, and permits direct delivery of oral medication into the stomach (Otte et al., 2003). However, the placement of a PEG tube requires a short anaesthetic, which may be contraindicated in the critical myasthenic patient (Penderis & Martin-Vaquero, 2016). As PEG tubes can remain in situ for up to six months, patients are often discharged with these (Collins, 2012). It is therefore the nursing staff's responsibility to advise and teach owners, on discharge of the patient, the correct feeding technique and the importance of maintaining the stoma site. Regular communication with the client can help ensure they feel supported throughout the recovery period. It is imperative that the PEG tube not be removed for at least seven to ten days, to allow for adhesions to form between the stomach and abdominal wall, reducing the risk of peritonitis (Collins, 2012).

Figure 2. A Bailey chair used to feed a dog with megaoesophagus secondary to MG.

Respiration

Aspiration pneumonia is associated with increased morbidity and mortality, therefore prevention and treatment, through intensive nursing care, is essential to the patients prognosis (Penderis & Martin-Vaquero, 2016). Recumbent patients should be turned frequently (every 2-4 hours) to prevent hypostatic lung oedema relating to the pneumonia. Turning the patient will also help prevent the development of decubital ulcers. Ideally, the patient should be maintained in a sternal position, with their head elevated and nose dropped, to allow for the drainage of oesophageal reflux, reducing the risk of aspiration (Richmond, 2015).

Oxygen therapy may also be required in the patient with aspiration pneumonia. The delivery of oxygen should be as well tolerated as possible for the patient, to minimise stress (Penderis & Martin-Vaquero, 2016). Oxygen delivered through the use of nasal oxygen prongs is generally well tolerated in the recumbent patient (Figure 3). Alternatively, the use of an oxygen kennel or tent may be considered.

In addition to oxygen therapy, the use of nebulisation using a sterile saline solution and a human nebuliser may be required. This assists with the breakdown of secretions within the lungs and helps keep the airways moist (Richmond, 2015). For the patient presenting with severe weakness, or reduced oxygenation, the use of positive pressure ventilation may be required, however this may not be readily available due to the cost implications and the specialist equipment required (Penderis & Martin-Vaquero, 2016).

Coupage can be beneficial for the patient with aspiration pneumonia, as it helps expel pulmonary and bronchial secretions

Figure 3. Oxygen delivery through the use of nasal oxygen prongs.

by stimulating the cough reflex (Penderis & Martin-Vaquero, 2016). For patients with MG, suction should be used to rapidly clear secretions from the oral cavity, as due to the reduced oesophageal and pharyngeal function, the risk of airway obstruction is substantial (Richmond, 2015).

Further nursing considerations

Should the myasthenic patient be recumbent, urination should be monitored. Many patients become agitated, or even refuse to urinate/defaecate in their beds. Because of this, the size of their bladder should be monitored regularly (every 4-6 hours) to ensure it is not becoming too large and overdistended. This can be done either by palpation, or by the use of an ultrasound scanner. Their bed should be regularly checked for urine, as to avoid urine scalds. This can be performed every 2-4 hours, at the same time as turning, as to allow for the patient to rest uninterrupted in between. The placement of an indwelling urinary catheter could also be considered should bladder management become difficult.

Should the patient be excessively hypersalivating, cleaning of the ventral aspect of the mandible should be considered to avoid sores developing. If washing is required regularly due to hypersalivation or urination, the patient should be thoroughly dried, to ensure they do not become hypothermic due to their inability to ambulate.

The recumbent myasthenic patient may also benefit from physiotherapy, to minimise muscle wastage and maintain range of motion of the joints. Depending on how critical the patient is, different levels of physiotherapy may be adopted. Efflourage, petrissage and passive range of motion would be a good starting point for all recumbent patients, to ensure the muscles remain supple, and avoid the development of muscle contractures.

Prognosis

The aim of the treatment for patients with AMG is to provide clinical and immunological remission. Unfortunately MG can recur even once remission has been achieved; therefore, prognosis is guarded. The one year mortality rate for dogs with AMG has been reported to be as high as 60%, however this may be improved through early diagnosis and prompt initiation of therapy (Garosi, 2013).

Disclosure statement

No potential conflict of interest was reported by the author.

ORCID

Ilaria Vercesi http://orcid.org/0000-0002-0735-6590

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