Canine Megaesophagus
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Megaesophagus is a condition characterized by decreased or absent esophageal tone and motility usually resulting in diffuse dilation of the esophagus.

**Megaesophagus Etiology (ME):**

ME can be broken down into two major categories: Congenital and Acquired.

1. **Congenital Megaesophagus (CME)**

Congenital megaesophagus should be considered in puppies and young dogs that present for regurgitation. Important differential diagnoses in this pet population are vascular ring anomalies, dysmotility secondary to skeletal immaturity, congenital strictures, and esophageal foreign bodies.

This condition can occur in purebred dogs as well as mixed breed dogs. It is known to be inherited in the wire-haired fox terrier and the miniature schnauzer. Other breeds with an increased prevalence of CME include German shepherds, Great Danes, Labrador retrievers, Newfoundlands, Irish Setters and Shar-peis. It is not yet known, but thought that a hereditary basis for CME exists in these breeds.

The pathogenesis is unknown but suspected etiologies include esophageal hypomotility or defects in vagal afferent innervation of the esophagus. Some dogs have congenital myasthenia gravis causing their megaesophagus. Breeds predisposed to congenital myasthenia gravis are Parson Russell terriers, smooth fox terriers, dachshunds and springer spaniels.

Clinical signs usually begin around the time of weaning (~3 months of age) however they may not become evident until one year of age. Smooth fox terriers with congenital myasthenia gravis causing megaesophagus show clinical signs earlier, around 4-9 weeks of age.

2. **Acquired Megaesophagus (AME)**

Acquired ME occurs spontaneously and usually affects middle aged to older, large breed dogs, typically between 5-12 years of age but can occur at any age. Irish Setters, Great Danes, German shepherd, Labrador retrievers, and Newfoundlands have an increased prevalence for AME. In Newfoundlands, AME secondary to myasthenia gravis occurs at a much younger age (<2 years).

Acquired ME can be broken down into two major categories: acquired idiopathic ME and acquired secondary ME.
1. Acquired idiopathic ME is the most common form. Unfortunately, most adult dogs with AME will not have an underlying primary disease elucidated. The etiopathogenesis is unknown but thought to be neurogenic rather than myogenic.

2. Acquired secondary ME can be the consequence of many other primary diseases. The most common cause is myasthenia gravis. It accounts for about 25% of acquired secondary ME. It can affect any age, however it typically has a bi-modal age distribution of < 2-3 or > 9 years old. Focal myasthenia gravis predominately affects esophageal, pharyngeal, or facial muscles without generalized muscle weakness. Focal myasthenia gravis occurs in 36-43% of all canine cases of myasthenia gravis. Generalized forms of myasthenia gravis occur in 57-64% of all canine cases with 90% of them having ME. Other causes of secondary AME include esophagitis, hypoadrenocorticism, dysautonomia, thymoma, lead toxicity, organophosphate toxicity, polynuropathy, CNS disease, bilateral vagal damage, SLE, polymyositis, polymyopathies, botulism, pituitary dwarfism. Hypothyroidism has been suggested as a cause but a definitive association is yet to be proven.

Many obstructive esophageal diseases (neoplasia, granuloma, vascular ring anomaly, stricture, periesophageal masses and foreign bodies) can also lead to megaesophagus if they are chronic enough in duration.

**Clinical Signs:**

Regurgitation is the classic clinical sign seen with ME. Dysphagia, halitosis, ptyalism and vomiting may also be reported. Coughing, lethargy, and nasal discharge may also be seen as a result of aspiration pneumonia secondary to regurgitation. Some cases may present with clues to an underling disease, such as generalized muscle weakness associated with myasthenia gravis.

It is very important to differentiate vomiting from regurgitation through careful historical investigation. Regurgitation is a passive process (no active abdominal component) with lack of prodromal signs. It can occur minutes to hours after ingestion of food or water. It difficult to ascertain based on an owner’s description, videotaping the event could be helpful. Sometimes both vomiting and regurgitation are part of a patient's clinical picture, often in cases where vomiting has led to esophagitis and subsequent regurgitation. Typically both liquid and solid food passes poorly with motility disturbances of the esophagus, whereas liquids often pass easily with obstructive lesions.

**Physical examination:**

When performing a physical examination in patients with ME, close attention should be paid to the respiratory and neuromuscular systems.

Physical examination findings are dependent on the duration of the megaesophagus and the underlying disease causing the megaesophagus. Findings include poor body condition, fever, and harsh lung sounds or crackles if aspiration pneumonia is present.

Dogs with focal myasthenia gravis as the underlying cause usually have weakness present in 1 or more muscle groups (ie esophageal, pharyngeal, laryngeal muscles) but generally do not
have any signs or thoracic or pelvic muscle weakness. Facial muscle weakness is usually manifested as a decremental blink reflex. Dogs with generalized myasthenia gravis can have a wide range of physical examination finding but often will have pelvic limb affected more than thoracic limbs, which can improve with rest and worsen with activity.

**Diagnosing Megaesophagus:**

ME is often easily diagnosed on plain thoracic radiographs on which an air or sometimes a fluid/ingesta filled esophagus is apparent. Conditions that cause aerophagia (excitement, nausea, vomiting, dyspnea, etc.) can result in air dilation of a normal esophagus. It is important to also evaluate thoracic radiographs for evidence of aspiration pneumonia and intrathoracic masses/neoplasia as well.

The degree of esophageal dilation has not been shown to be associated with survival time or etiology.

Occasionally, dogs with ME will need a contrast esophogram to demonstrate the outline of a dilated esophagus when plain radiographs are questionable. It should be noted that contrast studies are not without risk in this patient population as there is a risk of aspirating contrast material.

Esophagoscopy is rarely needed for a diagnosis of ME and is far less superior to plain radiographs and contrast studies. It can be helpful for suspected cases of esophagitis however rarely it is utilized for this cause due to risks of aspiration pneumonia in these dogs.

**Diagnostic work-up for patient with ME:**

Following a diagnosis of megaesophagus, further investigation is necessary to determine if ME is secondary to an underlying cause.

The following diagnostics should be performed in all patients:

- Complete blood count
- Chemistry panel (including creatinine kinase)
- Urinalysis
- Acetylcholine receptor antibody test
- Baseline cortisol +/- ACTH stimulation test
- Thyroid testing

Additional diagnostic that should be considered include:

- Abdominal ultrasound: search for underlying GI cause for esophagitis, search for neoplasia as trigger for immune mediated diseases (i.e. myasthenia gravis, polyneuropathy, etc.)
- Pilocarpine response test: Dysautonomia
- Blood cholinesterase: Organophosphate toxicity
- Esophagoscopy: Esophagitis, foreign body, mass, stricture
- Electromyography: Myasthenia gravis, polyneuropathy
- Muscle biopsy: Dermatomyositis, polymyositis, glycogen storage disease, congenital myasthenia gravis
- Nerve conduction/nerve biopsy: Polyneuropathy
- Skin biopsy: Dermatomyositis
• Tensilon test: Myasthenia gravis

Some cases of myasthenia gravis have negative titers on initial presentation and therefore repeating this test 2-3 months later should be performed in all cases deemed as idiopathic megaesophagus.

Congenital myasthenia gravis cannot be diagnosed with measurement of acetylcholine receptor (AchR) antibody titer as the disease is due to a deficiency or functional abnormality of AchRs, rather than an immune mediated destruction of AchRs. A muscle biopsy (intercostal muscle) is needed to diagnose congenital myasthenia gravis. If this is elected, contacting the comparative neuromuscular disease lab prior to obtaining the biopsy is recommended for instructions. http://vetneuromuscular.ucsd.edu/

**Treatment:**

If an underlying cause is found it should be treated as this allows for the best outcome. Myasthenia gravis is treated with long acting anticholinesterase drugs, pyridostigmine bromide 0.5-3mg/kg orally q8-12 or neostigmine bromide 0.1-0.25 mg/kg PO (total daily dose not to exceed 2mg/kg). Pyridostigmine bromide is preferred in most clinical situations because of its longer duration of action and fewer side effects. Ach receptor antibody concentrations should be monitored every 4-6 weeks, since spontaneous remission can occur. Treatments should continue until titers are within the normal range. If a confirmed case of MG is not responding to anticholinesterase drugs, immunosuppression can be considered. Prior to use of immunosuppressant agents pneumonia or other infections disease should be completely resolved.

Esophagitis treatment involves liquid/slurry sucralfate and a proton pump inhibitor, such as pantoprazole (IV) or omeprazole (PO). Many studies have shown that proton pump inhibitors are superior to H2 antagonists (Famotidine, etc.) and therefore should be used preferentially in esophagitis cases. The dose to effectively decrease gastric pH with omeprazole or pantoprazole is 1 mg/kg q12. Certain dogs will also benefit from promotility agents such as metoclopramide or cisapride.

Treatment for idiopathic megaesophagus is largely supportive and involves feeding modifications. Goals are to decrease the frequency of regurgitation, provide adequate nutrition, and treat secondary complications as they arise.

Dogs should be fed small meals of a calorically dense diet multiple times a day. The optimal food consistency varies from dog to dog, however canned meatballs seems to be one of the most well tolerated consistencies. Some dogs do best with a gruel or liquid consistency, therefore trial and error may be necessary to figure out what is best for the individual dog.

They should be fed upright with their spine perpendicular to the floor. A “Bailey chair” is a great way for owners to accomplish this task. After feeding the dog should remain upright for at least 10 minutes after each feeding. Simply raising the food bowl to head height is not effective.

Some dogs continue to regurgitate despite appropriate feeding techniques. In these patients, a gastrotomy tube is indicated as an effective means of providing nutrition, however this does not eliminate regurgitation as patients will likely still regurgitate their salivary secretions. Esophagostomy tubes as a means of providing nutrition should not be used. A recent cases series was published in which both a gastrotomy tube and esophagostomy tube were placed.
The esophagostomy tube was placed for intermittent suctioning of the esophagus and decreased the amount of regurgitation and aspiration pneumonia events in 3 out of 4 dogs.

The use of promotility agents, such as metoclopramide and cisapride, are generally not recommended in cases other than some esophagitis cases, as they are not effective in improving motility of the canine esophagus as they are effective only on smooth muscle and the canine esophagus is composed of striated muscle. In addition, these medications increase lower esophageal tone, which may make passage of food into the stomach more challenging. However, anecdotally, some patients do improve with these agents which may be because they have concurrent reflux esophagitis.

Secondary complications of ME include aspiration pneumonia and esophagitis. For aspiration pneumonia, broad spectrum antibiotics are recommended. An endotracheal wash can be performed however, this should be reserved for patients not responding to empirical therapy as there is increased risk of worsening aspiration in ME patients.

Note that medications should be in liquid (not pill) form to enhance movement into the stomach and to avoid esophageal irritation, if possible.

**Prognosis:**

The prognosis for ME patients varies and is difficult to predict. Even with diligent care, aspiration pneumonia is frequent and often a fatal complication. The most recent study (2011) evaluating factors associated with survival in dogs with ME, found that the overall prognosis is poor with a median survival time of 90 days, however 31% of dogs were alive 2 years after diagnosis. It is important to note that this study did not separate congenital, acquired idiopathic, and secondary acquired cases of ME when evaluating survival time.

Congenital megaesophagus generally has a guarded to poor prognosis. Reported recovery rates vary from 20%-46%, with Miniature schnauzers having the best likelihood of recovery as most typically return to normal by 6-12 months of age. The same study cited earlier found that dogs younger than 13 months old diagnosed with ME had a better survival than those diagnosed older than 13 months, which was thought to be due to skeletal immaturity at the time of diagnosis and improvement of esophageal motility with maturity. Congenital myasthenia gravis in Dachshunds may resolve spontaneously and potentially has a favorable prognosis.

The prognosis for acquired idiopathic megaesophagus in general is guarded to poor due to recurrence of aspiration pneumonia and malnutrition. Most die of aspiration pneumonia or are euthanized due to ongoing regurgitation or debilitation within 5 months of diagnosis. One study found that the presence of aspiration pneumonia at that time of diagnosis is a poor prognostic indicator (7.6 fold increase risk of dying before discharge and 2.2 times more likely to die at any given time point as compared to dogs without aspiration pneumonia).

The prognosis for secondary ME can be fair to good if the underlying disease can be treated successfully.

For acquired myasthenia gravis, the prognosis is guarded to fair as spontaneous resolution can occur within an average of 6 months (range 1mo to 1 year) in 50% of patients. However many patients will succumb to aspiration pneumonia prior to that time. ME appears to respond well in patients with hypoadrenocorticism.
Previous studies have shown that dogs with thymoma and ME had a poor prognosis, with a median survival time of 4 days. However a study in 2013, showed that surgical resection of thymomas in patients with megaesophagus or myasthenia gravis can have a good prognosis, however resolution of myasthenia gravis and ME is variable in these patients. The prognosis for ME secondary to dysautonomia is grave.

Dogs with ME secondary to polyradiculoneuritis, SLE, botulism and polymyositis, can recover esophageal function after successful treatment of the primary disease.