

Megaesophagus

References:

Tams TR, Chapter 4, "Diseases of the Esophagus," in Todd R Tams *Small Animal Gastroenterology*, 2nd Edition.

I. Etiopathogenesis:

A. Anatomy and physiology.

1. Esophagus is divided into three parts:
 - a. Cervical (upper esophageal sphincter to the thoracic inlet).
 - 1) Cricopharyngeus mm + thyropharyngeus m = upper esophageal sphincter (UES).
 - 2) UES always closed except to let food in.
 - b. Thoracic (thoracic inlet to the diaphragm).
 - c. Abdominal (diaphragm to the lower esophageal sphincter).
 - 1) LES inner smooth m and outer striated m in the dog.
 - 2) LES all smooth m in the cat.
 - 3) Drugs that increase LES tone:
 - i. Metoclopramide.
 - ii. Cisapride.
 - iii. Bethanechol.
 - iv. Erythromycin.
 - v. Domperidone.
 - 4) Drugs that decrease LES tone:
 - i. Acepromazine.
 - ii. Atropine.
 - iii. Diazepam (dogs).
 - iv. Propofol.
 - v. Xylazine.
 - vi. Halothane.
 - vii. Isoflurane.
2. Layers of the esophagus.
 - a. Adventitia (no serosa).
 - b. Muscularis.
 - 1) Entirely striated in the dog.
 - 2) In cats, cranial 2/3 is striated and caudal 1/3 is smooth.
 - c. Submucosa.
 - 1) Contains glands, nerves and blood vessels.
 - 2) Greatest holding strength for sutures.
 - d. Mucosa.
3. Innervation:
 - a. Glossopharyngeal nerve (CN 9)
 - b. Vagus nerve (CN 10) – recurrent laryngeal branches.
 - c. Sympathetic supply.

4. The swallowing process has three phases:
 - a. Oropharyngeal.
 - 1) Triggers primary esophageal contractions.
 - 2) Solids produce stronger primary contractions and weaker secondary contractions.
 - b. Esophageal.
 - 1) Esophageal distension produces secondary esophageal contractions.
 - 2) These waves begin proximal to the bolus.
 - 3) Liquids produce stronger secondary contractions.
 - 4) Speed in dogs is 75-100 cm/sec (striated m contracts faster).
 - 5) Speed in cats is 1-2 cm/sec. ****IMPORTANT WHEN TUBE FEEDING****
 - c. Gastroesophageal.
5. The canine esophagus can produce 10x the contraction pressure of the human esophagus.

B. Etiologies of megaesophagus:

1. Localized – dilation of megaesophagus proximal to obstruction.
 - a. Stricture.
 - 1) Trauma.
 - 2) Caustic substance swallowed – doxycycline.
 - b. Foreign body.
 - c. Parasite – *Spirocerca lupi*.
 - d. Vascular ring anomaly.
 - 1) Persistent Right Aortic Arch (PRAA).
 - 2) Compression by left subclavian a and brachiocephalic trunk.
 - e. Neoplasia.
 - 1) Mediastinal mass.
 - i. LSA.
 - ii. Thymoma.
 - 2) Esophageal mass.
 - f. Congenital.
 - 1) English bulldog – deviation of esophagus at base of the heart.
2. Generalized – entire esophagus affected.
 Most common causes of acquired megaesophagus are:
 1 – idiopathic
 2 – myasthenia gravis
 - a. Megaesophagus only.
 - 1) Esophagitis.
 - i. Gastroesophageal reflux.
 - ii. Hiatal hernia.
 - iii. Chronic GDV.
 - 2) Myasthenia gravis.
 - 3) Congenital.
 - 4) Idiopathic.
 - b. Megaesophagus as part of generalized neuropathy, myopathy or junctionopathy.
 - 1) Hypothyroidism
 - 2) Hypoadrenocorticism.
 - i. Abnormal sodium and potassium concentration affect membrane potentials and neuromuscular function.
 - ii. Cortisol deficiency causes muscle weakness.

- 3) Myasthenia gravis (acquired) – most dogs with generalized myasthenia gravis have at least some degree of esophageal dysfunction.
 - i. Autoimmune disorder.
 - ii. Autoantibodies against nicotinic acetylcholine (Ach) receptors at neuromuscular junctions.
 - iii. Complement mediated destruction of junction folds.
 - iv. There are three kinds of acquired MG: generalized, focal (facial, larynx, pharynx and esophagus), acute fulminant.
 - v. Megaesophagus has been associated with all 3 types.
- 4) Muscular Dystrophy.
- 5) Dysautonomia – more common in cats.
- 6) Immune mediated disease – rare
 - i. Dermatomyositis.
 - ii. Systemic Lupus Erythematosus (SLE).
 - iii. Polymyositis.
- 7) Giant axonal neuropathy.
- 8) Congenital.
 - i. Myasthenia gravis.
 - ii. Hereditary myopathy of Labrador retrievers.
- 9) Toxicity
 - i. Lead – sources of lead toxicity include old paint, old toys, lubricants, hobby materials, automotive materials, plaster board, roofing materials, fishing sinkers, and improperly glazed dishes.
 - ii. Thallium.
 - iii. Organophosphate.

C. **Pathogenesis:** megaesophagus is the most common esophageal disorder in dogs.

1. Physiologic studies suggests that a defect exists in the afferent neural pathway.
2. Efferent neural pathways seem to be intact.

II. Epidemiology/Signalment

A. Breed.

1. Boston Terrier.
 - i. PRAA.
 - ii. Hiatal hernia.
2. Bouvier – hereditary muscular dystrophy.
3. Collie – familial dermatomyositis.
4. English Bulldog – entrapping L subclavian a and brachiocephalic trunk.
5. German shepherd.
 - i. Acquired myasthenia gravis (**most common).
 - ii. Congenital megaesophagus.
 - iii. PRAA.
 - iv. Idiopathic megaesophagus.
 - v. Giant axonal neuropathy.
6. Golden Retriever
 - i. Acquired myasthenia gravis (**most common).
 - ii. Congenital megaesophagus.
 - iii. Idiopathic megaesophagus.
7. Great Dane.
 - i. Congenital megaesophagus.
 - ii. PRAA.
 - iii. Idiopathic megaesophagus.

8. Greyhound.
 - i. Congenital megaesophagus.
 - ii. Idiopathic megaesophagus.
9. Irish Setter.
 - i. Congenital megaesophagus.
 - ii. PRAA.
 - iii. Idiopathic megaesophagus.
10. Jack Russell Terrier – congenital myasthenia gravis.
11. Labrador Retriever.
 - i. Congenital megaesophagus.
 - ii. Idiopathic megaesophagus.
 - iii. Hereditary myopathy.
12. Miniature Schnauzer.
 - i. Congenital megaesophagus.
 - ii. Idiopathic megaesophagus.
13. Newfoundland.
 - i. Congenital megaesophagus.
 - ii. Idiopathic megaesophagus.
14. Pug – hiatal hernia.
15. Rottweiler – spinal muscular atrophy.
16. Shar Pei.
 - i. Congenital megaesophagus.
 - ii. Idiopathic megaesophagus.
 - iii. Hiatal hernia.
17. Siamese.
 - i. Congenital megaesophagus.
 - ii. Idiopathic megaesophagus.
18. Smooth fox terrier – congenital myasthenia gravis.
19. Springer Spaniel – Polymyopathy.
20. Wire haired fox terrier.
 - i. Congenital megaesophagus.
 - ii. Idiopathic megaesophagus.
21. Congenital megaesophagus known to be inherited in:
 - i. Miniature schnauzer – simple autosomal recessive, or 60% entrance autosomal recessive.
 - ii. Wire haired fox terrier – simple autosomal recessive.

B. Age.

1. young.
 - i. Vascular ring anomaly.
 - ii. Congenital megaesophagus – weaning to 6 months of age.
2. young to middle aged.
 - i. Hypoadrenocorticism.
 - ii. Hypothyroidism.
 - iii. Myasthenia gravis.
3. geriatric.
 - i. Hypothyroidism.
 - ii. Idiopathic.
 - iii. Myasthenia gravis.

III. History.

A. **Regurgitation** is the most common clinical sign.

1. The degree of esophageal dysfunction does not always correlate with severity of clinical signs.
2. How do you tell vomiting from regurgitation (and coughing)?

Vomiting	Regurgitation	Coughing/Gagging
Minutes to hours after eating.	Minutes to hours after eating.	Not usually associated with eating, but can be associated with drinking water.
Expulsion process seems more active, with abdominal contractions	.Expulsion process seems more passive.	Expulsion process associated with gagging, but not usually with abdominal contractions, unless gagging causes retching.
Can be preceded by salivation, pacing, swallowing, anxiety and apparent discomfort.	Few premonitory signs except salivation. Sometimes associated with dysphagia (repeated swallowing, difficulty swallowing, or gagging up food).	May be preceded by coughing.
Consistency varies from hardly digested to liquid. Smell is variable.	Consistency varies from hardly digested to liquid. May smell sour and fermented.	White foamy material. Not usually foul smelling.
Less often contains mucus.	Often contains mucus (saliva). Foamy and slimy.	White foamy material, may contain mucus.
“Coffee grounds” digested blood, if present, suggests vomiting.	Blood is rare.	May be blood tinged if respiratory disease is severe.
Maybe contain bile.	Never bile stained.	Never bile stained.
pH is variable – unreliable indicator.	pH is variable – unreliable indicator.	pH usually near neutral.
Why we are so confused – sometimes regurgitation or coughing leads to gagging, and then vomiting.		

B. **Signs of aspiration pneumonia:**

1. Coughing – acute or chronic.
2. Fever.
3. Dyspnea.
4. Cyanosis.

C. **Coughing** can also be due to enlarged esophagus and its contents compressing on the airways and lungs.

1. Can have coughing without regurgitation.
2. ****SUSPECT MEGAESOPHAGUS IN AN OLDER DOG WHO IS BOTH COUGHNG AND “VOMITING.”****

D. **Oropharyngeal dysphagia**, if part of generalized myopathy or neuropathy.

1. Difficulty swallowing
2. Gagging up food while eating, or shortly thereafter.

E. **Weight loss** – can be severe.

F. **Poor appetite or salivation.**

G. **Signs of esophagitis:** most often anorexia, salivation and lethargy.

H. Neurologic deficits.

1. Generalized lower motor neuron disease with myopathies, neuropathies and junctionopathies.
2. Generalized muscle atrophy with muscular dystrophy, neuronal degeneration, polymyositis or even starvation.
3. Muscle pain with polymyositis.

I. Signs of myasthenia gravis:

1. Episodic weakness and weakness with exercise.
2. Difficulty barking.
3. Dysphagia.
4. Weak or absent palpebral reflex.

J. Signs of hypoadrenocorticism:

1. Vomiting, diarrhea (especially with blood).
2. Lethargy and weakness.
3. Shock if severe.

K. Signs of hypothyroidism:

1. Obesity.
2. Hair loss.
3. Pyoderma.

L. Signs of lead toxicity.

1. GI signs – vomiting, diarrhea, abdominal pain, anorexia.
2. Neurologic signs – seizures, dullness, behavior changes, apparent blindness.

M. Signs of immune mediated disease.

1. Muscle pain – polymyositis.
2. Joint pain and/or swelling – polyarthritis.
3. IMHA or ITP.
4. Skin or mucus membrane lesions.
5. Fever.

IV. Physical Exam

A. Loss of body condition – most common clinical sign.

B. If aspiration pneumonia.

1. Mucopurulent nasal discharge.
2. Fever.
3. Crackles in small airways.
4. Dull lung sounds if lung consolidation.

C. Valsalva maneuver causes puffing of the cervical esophagus.

1. Nose and mouth are occluded.
2. Thorax is compressed.
3. I don't usually do this until after thoracic rads.
4. Doing this on a dog with an esophagus filled with food could cause aspiration.

D. There may be saliva or food particles at the back of the throat.

V. Diagnosis – the majority of dogs have idiopathic disease. But the tests are worth doing, because without a treatable cause, long prognosis without permanent stomach tube is poor.

A. CBC.

1. Increased WBC with aspiration pneumonia, with or without degenerative left shift.
2. May have anemia of chronic disease – mild to moderate non-regenerative anemia.
3. IMHA would be rare.

B. Serology:

1. Low albumin and/or elevated liver enzymes if septic due to aspiration pneumonia, or if immune mediated glomerulonephritis.
2. High cholesterol and/or triglycerides – may suggest hypothyroidism.
3. CK and AST – elevated with polymyositis.
4. Thyroid profile.
 - a. Some do T4 only first, with panel; follow up with TSH and freeT4 by equilibrium dialysis if T4 is low.
 - b. Others do TSH, T4 and freeT4 (ED or not) at the outset.
 - c. Confirmed hypothyroidism should not preclude testing for hypoadrenocorticism and myasthenia gravis. A significant number of dogs have both hypothyroidism and one of these other diseases.
5. Electrolytes – high potassium, low sodium in 90% of Addisonians.
6. Myasthenia titer – anticholinesterase antibody – to test for localized or generalized myasthenia gravis, in dogs and cats .
 - a. See Appendix 3 for lab information, submission instructions and forms .
 - b. All pets with myasthenia should be tested for hypothyroidism, as 20% of dogs with myasthenia also have hypothyroidism.
 - c. >0.6 nmol/L is diagnostic in dogs, and >0.3nmol/L in cats.
 - d. The test is diagnostic for all 3 types of MG.
7. ANA – of limited usefulness
 - a. Immune mediated diseases is a very rare cause of megaesophagus.
 - b. ANA is not terribly sensitive for immune mediated disease, though its specificity is reasonable when the titer is high.

C. **Urinalysis** – proteinuria if immune mediated disease.

D. Fecal examination.

1. Sugar and salt flotation solutions (S. G. = 1.22) will not float the typical embryonated eggs of *Spirocerca lupi*.
2. NaNO₃ (S.G. = 1.36) will float the eggs.
3. *Spirocerca* eggs can sometimes be seen on direct wet mount.

E. ACTH Stimulation Test

1. Protocol – see Appendix 4.

F. Toxicology.

1. Blood lead greater than 40 ug/dl suggests lead poisoning.
2. Level greater than 60 ug/dl is diagnostic for lead poisoning.

G. Radiographs.

1. Survey thoracic and cervical radiographs.
 - a. May be normal.
 - b. May see enlarged, air filled esophagus.
 - c. May need to do both right and left laterals to see air.
 - d. May see mass in mediastinum or esophagus, or foreign body.
 - e. Only part of the esophagus will be dilated when there is localized megaesophagus due to obstruction.
 - f. Severity of dilation due to obstruction can indicate prognosis.
2. Barium swallow and cervical/thoracic rads can confirm.
 - a. Give 10-20 c barium by mouth, and take films immediately.

- b. Use barium liquid, paste or barium coated food.
 - 1) Paste coats best, but can be dangerous if aspirated.
 - 2) ****WARN OWNER OF DANGER OF THIS PROCEDURE****
 - 3) Some motility disorders will not be detected by liquid barium alone – barium coated food may be necessary.
 - c. Take care to prevent aspiration of barium into the lungs.
 - d. Normal esophagram will show tiny lines only at the esophageal mucosal border.
 - e. Barium will pool in the esophagus with megaesophagus.
 - f. Can estimate wall thickness.
 - g. May see filling defect due to radiolucent foreign body, esophageal mass or ulcer.
3. Abdominal survey rads.
- a. Radiopaque foreign body in GI tract with heavy metal poisoning.
 - b. Chronic gastric volvulus.

H. Fluoroscopy .

- 1. The best way to evaluate mild to moderate esophageal hypomotility.
- 2. Liquid barium first.
- 3. Then liquid barium mixed with canned food.
- 4. Then liquid barium coated kibble.

I. Abdominal ultrasound – to look for neoplasia or chronic inflammation/infection as a cause for acquired myasthenia gravis.

J. Tensilon test.

- 1. Protocol – edrophonium 0.05-0.1 mg/lb IV.
 - a. Response within 1-2 minutes.
 - b. Response lasts several minutes.
- 2. Difficult to interpret results unless weakness is profound.
- 3. It can take 10-15 minutes of exercise to cause weakness in some cases.

K. Electrodiagnostics - EMG and nerve conduction velocity (NCV) – can differentiate between polymyositis, myopathy, neuropathy and junctionopathy.

- 1. Myasthenia gravis.
 - a. EMG and NCV are usually normal.
 - b. Decremental response to repeated nerve stimulation.
 - c. Decremental response eliminated by Tensilon test.
- 2. Polymyositis:
 - a. EMG sharp waves, fibrillation potentials, high frequency discharges.
 - b. No decremental response to repetitive nerve stimulation.

L. Muscle and nerve biopsies.

- 1. Indicated only when megaesophagus is a component of generalized weakness.
- 2. Can give definitive classification as myopathy or neuropathy, to guide further testing and therapy.
- 3. Can give definitive diagnosis of myopathies such as polymyositis, muscular dystrophy, and certain neuropathies.
- 4. Can support evidence of endocrine disease.
- 5. See Appendix 3 for sample handling instructions and submission forms, for the Comparative Neuromuscular Lab at UC-Davis. Choose Complete Muscle and Nerve Profiles.
- 6. REMEMBER – anesthesia can be very dangerous for patients with megaesophagus.
 - a. Large amounts of food and ingesta in the esophagus can be aspirated.
 - b. Many pets with ME also have dysphagia, and have abnormal swallowing reflexes, so they can't protect their airways well.

VI. Treatment.

A. Treat underlying cause.

1. Hypothyroidism – thyroxine, initial dose 0.1 mg/lb SID to BID. Recheck T4 in 4 weeks and adjust.
2. Hypoadrenocorticism.
 - a. Fludrocortisone acetate (Florinef) – 1.5-2 tablets (0.1 mg each) per 10 kg body weight PO SID or divided BID.
 - b. DOCP – desoxycorticosterone pivalate (Percorten) – 1 mg/lb SQ or IM every 25-30 days.
3. Myasthenia gravis.
 - a. Pyridostigmine (Mestinon) syrup or tablets.
 - 1) 0.25-1.5 mg/lb PO BID-TID.
 - b. Neostigmine (Prostigmin) 0.2 mg/lb IM q6hrs.
--use this if you are not sure if PO meds will get to the stomach.
 - c. Watch for signs of overdose:
 - 1) muscle weakness.
 - 2) salivation.
 - 3) miosis.
 - 4) vomiting.
 - 5) diarrhea.
4. Vascular ring anomaly, hiatal hernia, chronic GDV, neoplasia – surgery.
5. Esophageal stricture – balloon dilatation, bougienage (repeat every 2 weeks until resolved).
6. Lead toxicity – EDTA chelation, remove foreign body if present.

B. Elevated feedings.

1. Primary treatment if animal is to be fed by mouth.
2. Upper body should be elevated 45-90 degrees, for at least 10 minutes, to use gravity as an aid to move food from the mouth to the stomach.
3. Since the esophagus is rarely empty with megaesophagus, holding the patient in an upright position for 5-10 minutes several times daily can also be helpful.
 - a. This is especially important prior to periods of known prolonged lateral recumbency, when the patient is at risk for aspiration.
 - b. For example, this should be done before bedtime.
4. It is important that proper positioning be carefully demonstrated, so the owner understands clearly what to do.
5. Patients should be fed 2-4 times per day, and avoid feeding within several hours of bedtime. People who work full time can often feed breakfast and lunch.
6. Tools that can be used:
 - a. Small dogs and cats can be held in an upright position while they eat and after.
 - b. Shoulder feeding harness for small dogs.
 - c. Feeding pedestals only for dogs and cats who will not know them over, or if they can be well secured.
 - d. Specialized feeding chairs (“high chairs”) for small to medium dogs.
 - e. Step ladder, or stairs for large or giant dogs.
 - f. Long legged dogs can be put in a sitting position and fed with the head up.
 - g. Some owners build special platforms.
7. Experiment with type of food to find out what consistency is associated with the least regurgitation.
 - a. Liquid/gruel might be better if primary esophageal contractions are weak.
 - b. Meatballs might be better if secondary esophageal contractions are weak.
 - c. Sometimes dry kibble works best.

C. Gastrostomy tube (see section on Feeding Tube Placement).

1. Temporary gastrostomy tube can help, until pet is stabilized and an effective therapeutic regimen is developed.
 - a. Often are required for 1-3 months.
 - b. Medications can be given by gastrostomy tube (giving PO meds to pets with megaesophagus is dicey at best).
 - c. NEVER GIVE DOXYCYCLINE PO TO A PATIENT WITH ME.
 - d. Can be extremely helpful in managing aspiration pneumonia.
2. Some dogs and cats can do very well with a low profile permanent gastrostomy tube.
 - a. Place a Pezzar tube first.
 - b. When stoma well healed, replace with low profile gastrostomy tube.
 - c. These may need to be replaced periodically.
3. Owners of pets with megaesophagus who can not be managed feeding by mouth have to be vigilant to prevent pets from eating anything on their own.

D. Prokinetics.

1. Cisapride (0.05-0.25 mg/lb PO TID).
 - a. Smooth muscle prokinetic.
 - b. Theoretically would be more effective in cats than dogs, as the feline esophagus has more smooth muscle than the canine esophagus.
 - c. However, some dogs do have fewer episodes of regurgitation when they take cisapride.
 - d. Response can vary from no response to dramatic positive response.
2. Metoclopramide (0.2-0.4 mg/kg PO SQ IV TID-QID).
 - a. Has no effect on the esophagus.
 - b. But may help gastroesophageal reflux by decreasing stomach emptying time.
 - c. I use only if lower esophagitis is a problem.

E. Treat aspiration pneumonia.

1. Broad spectrum antibiotics that are effective against gram negatives, gram positives and anaerobes.
2. Fluid therapy.
3. Coupage.
4. I don't tend to do nebulization, because I don't want to increase respiratory secretions which can be aspirated.
5. Consider a gastrostomy tube for feeding (See Section on Tube Feeding).
6. Give all meds by injection or by G-tube – not PO.
7. Long term antibiotics indicated in dogs who have constant mild pneumonia.
 - a. Choose 3 that work and rotate every 6-8 weeks.

F. Treat esophagitis. – acute or chronic.

1. Sucralfate (0.5-1g/15 lbs PO TID-QID).
2. H2 blockers.
 - a. Cimetidine (Tagamet) 2.5-5 mg/lb PO TID-QID.
 - b. Ranitidine (Zantac) 1 mg/lb (dog) and 1.5 mg/lb (cat) PO BID-TID (**prokinetic).
 - c. Famotidine (Pepcid) 0.25-0.5 mg/lb PO SID-BID.
 - d. Nizatidine (Axid) 1.25-2.5 mg/lb PO SID (**prokinetic).
3. Proton pump blockers – acid suppression more complete and long lasting than H2 blockers.
 - a. Omeprazole (Prilosec) - 0.3 mg/lb PO SID.
 - b. Lansoprazole (Prevacid) – 15 mg PO SID for small dogs; 30 mg PO SID for large dogs.

- c. Esomeprazole (Nexium)
 - 1. 0.7 mg/kg PO SID for dogs.
 - 2. Granules in capsule inactivated if sprinkled on food.
 - d. Pantoprazole (Protonix) – 10-140 mg PO SID; 1 mg/kg IV SID.
 - e. Rabeprazole (Aciphex) – 5-20 mg PO SID.
- 4. Prokinetics (see above), to decrease stomach emptying time;
 - 5. Therapy should continue for at least 3 weeks for acute cases, and longer or chronically if needed.

G. Nutrition

- 1. Low fat diet, to reduce stomach emptying time.
- 2. Low fiber diet, to reduce stomach emptying time.

H. Immunosuppression.

- 1. Might be indicated for megaesophagus related to:
 - a. Polymyositis.
 - b. SLE.
 - c. Myasthenia gravis.
- 2. Indicated only when immune mediated disease is confirmed, or as a last resort.
- 3. Immunosuppression is dangerous for pets who can develop aspiration pneumonia at any time.
- 4. Immunosuppression is contraindicated in pets with megaesophagus and aspiration pneumonia.
- 5. Some patients with myasthenia gravis acutely decompensate when treated with immunosuppressive drugs.
 - a. Prednisone can antagonize Ach, creating a neuromuscular junction blockage of the Ach receptor channel, which may then uncouple the excitatory processes.
 - b. The above can cause a fulminating MG crisis.
- 6. Drugs:
 - a. Prednisone 0.25 mg/lb/day divided BID, and gradually increased to 1 mg/lb/day over 1-2 weeks, if tolerated. If positive response, keep at this dose for 2 weeks, then wean slowly over 3-6 months.
 - b. Azathioprine 0.5 mg/kg PO SID x 14 days, then increase to 1 mg/kg/day. After in remission, can go to 1 mg/kg PO QOD. If that is well tolerated for 1-2 months, you can then try 0.5 mg/kg PO QOD until MG titer is normal.
 - c. These drugs can be used together when megaesophagus is a component of SLE.
 - d. Dogs who respond to immunosuppression can sometimes be weaned off of Mestinon.
- 7. Bottom line for immunosuppression is that patients can respond dramatically positively, not at all, or dramatically negatively.

I. Surgery – not indicated in dogs.

- 1. Cardiac myotomy used for achalasia in people (failure of LES to relax, with ineffective esophageal peristalsis).
- 2. LES achalasia has not been documented in animals.
- 3. Esophageal tone is normal, not increased, in dogs with megaesophagus.

VII. Monitoring

- A. Whenever there is decompensation, evaluate and treat for esophagitis and aspiration pneumonia. If pneumonia is ruled out by chest rads, treat for esophagitis.
- B. Hypothyroidism – ensure adequate T4 levels.

- C. Myasthenia gravis – monitor myasthenia titer as an indication of resolution of disease, and cessation of treatment. Myasthenia titer ideally should be checked once monthly, until weaned off of all drugs and still normal.
- D. Dogs taking Imuran should have
 - 1. CBC:
 - i. 3 weeks after starting therapy.
 - ii. 6 weeks after starting therapy.
 - iii. Every 2 months thereafter.
 - iv. Imuran should be reduced if WBC falls below 2,500/ul, and stopped if it falls below 2,000/ul.
 - 2. Liver enzyme panel every 3 months.

VIII. Sequella/Prognosis

A. Sequellae.

- 1. *Spirocerca* granulomas can undergo malignant transformation to fibrosarcoma.
- 2. Nutritional wasting.
- 3. Aspiration pneumonia (potentially fatal).
- 4. ****GENERAL ANESTHESIA SHOULD BE DONE WITH GREAT CARE TO AVOID ASPIRATION IN PETS WITH MEGAESOPHAGUS****
 - a. Induction should occur quickly, so that the patient can be intubated ASAP, and the airway protected.
 - b. Drugs that inhibit LES tone should be avoided if possible.
 - 1. Acepromazine.
 - 2. Atropine.
 - 3. Diazepam (dogs).
 - 4. Propofol.
 - 5. Xylazine.
 - 6. Halothane.
 - 7. Isoflurane.
 - c. Contents of esophagus should be suctioned immediately after intubation, as needed during the procedure, and immediately prior to recovery.
 - d. The endotracheal tube should be pulled slightly inflated if possible.
 - e. The airway should be suctioned if needed, after extubation.

B. Prognosis.

- 1. *Spirocerca* granulomas are usually too large to be effectively treated prior to diagnosis.
- 2. Severe dilation secondary to obstruction often carries a poor prognosis for recovery of esophageal motility.
- 3. If no treatable underlying cause of megaesophagus, long term prognosis for idiopathic megaesophagus variable, depending on:
 - 1. Use of permanent gastrostomy tube.
 - 2. Response to cisapride.
 - 3. Tendency to develop aspiration pneumonia.
- 4. Prognosis for congenital megaesophagus is guarded in general; rarely, a puppy will have spontaneous resolution of megaesophagus between 6-12 months of age.
- 5. All patients with megaesophagus are at risk for sudden death due to aspiration and respiratory obstruction.
- 6. Prognosis for megaesophagus in hypothyroid dogs is guarded, as many do not respond to thyroid supplementation.
- 7. Prognosis for megaesophagus caused by hypoadrenocorticism is potentially good if aspiration pneumonia can be managed, as these dogs usually respond to therapy.
- 8. Long term prognosis for pets with myasthenia gravis is potentially good, as most cases spontaneously resolve within 6-9 months. However, short term prognosis is guarded, as more

than half of patients with myasthenia gravis die of aspiration pneumonia within 6-9 months of diagnosis.

IX. **Public Health Significance** – none.