

Compendium

Megaesophagus

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Abstract: *Megaesophagus is a disorder of the esophagus characterized by diffuse dilation and decreased peristalsis. It is classified into congenital and acquired forms. Gastrointestinal, endocrine, immune-mediated, neuromuscular, paraneoplastic, and toxic disorders have been associated with acquired megaesophagus. Common clinical signs of megaesophagus are regurgitation, weight loss, coughing, and halitosis. Most cases of megaesophagus can be diagnosed using thoracic radiography; however, diagnosing the underlying cause requires a thorough history and additional diagnostics. The treatment, management, and prognosis of megaesophagus vary greatly depending on the underlying cause.*

Megaesophagus is defined as a disorder of the esophagus characterized by diffuse esophageal dilation and decreased peristalsis.¹ It may be congenital or acquired¹; acquired megaesophagus is subclassified into idiopathic and secondary forms. Congenital and idiopathic acquired megaesophagus disorders are suspected to be due to a combination of neurologic dysfunction within the afferent arm of the swallowing reflex, altered esophageal viscoelastic properties, and poor vagal responsiveness to intraluminal esophageal distention.^{1,2} Secondary acquired megaesophagus can be caused by any disease that inhibits esophageal peristalsis by disrupting central, efferent, or afferent nerve pathways or by any disease of the esophageal musculature, including immune-mediated, infectious, and preneoplastic etiologies.³

Clinical Signs

In uncomplicated cases of megaesophagus, patients may present with only regurgitation and weight loss. Other patients may present with additional clinical signs that hint at the underlying cause of megaesophagus.¹ The most common complication of megaesophagus is aspiration pneumonia; often, these patients present with a moist cough, dyspnea, or fever.¹

History

The index of suspicion for megaesophagus should be high when a patient presents for regurgitation. Regurgitation that occurs in a young patient at the time of weaning and conversion to solid food is likely due to congenital megaesophagus. In older patients, the frequency of regurgitation and timing may be more variable.

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In these cases, acquired megaesophagus should be suspected.¹ Other concerns owners may report are weight loss, coughing, and halitosis. Weight loss due to megaesophagus results from regurgitant loss of caloric intake.¹ Coughing occurs with aspiration pneumonia, and halitosis is a sequela of chronic retention and regurgitation of ingesta.

Signalment

Congenital megaesophagus is documented in Newfoundlands, Parson Russell terriers, Samoyeds, springer spaniels, smooth fox terriers, and shar-peis.^{1,2} These dogs typically present at the time of weaning with signs of regurgitation.¹ Smooth fox terriers that have megaesophagus secondary to congenital myasthenia gravis (MG) present between 4 and 9 weeks of age.⁴ Irish setters, Great Danes, German shepherds, Labrador retrievers, miniature schnauzers, and Newfoundlands have an increased prevalence for acquired megaesophagus.^{1,2,5} Dogs with acquired megaesophagus present from 7 to 15 years of age.¹ In Newfoundlands, acquired megaesophagus and MG occur at a much younger age (≤ 2 years). These dogs do not have a history of congenital megaesophagus.⁶ Congenital and acquired megaesophagus have been documented in cats, with a familial disposition for the congenital form in the Siamese breed.¹

Congenital Megaesophagus

The suspected etiology for congenital megaesophagus is esophageal hypomotility.⁷ In some patients, this hypomotility is due to delayed maturation of esophageal function that may or may not improve with age.⁷ Congenital MG is an inherited autosomal recessive

condition in Parson Russell terriers, springer spaniels, and smooth fox terriers that results in a deficiency or functional abnormality of acetylcholine receptors (AChRs) at the neuromuscular junction.⁸ The long-term prognosis for congenital MG is poor because of the mechanism of the condition, lack of a specific treatment, and high complication rate of aspiration pneumonia. Congenital MG patients usually present with generalized weakness in addition to megaesophagus. Congenital MG patients generally succumb within 1 year; however, there are reports of some cats and Parson Russell terriers that have survived for several years.⁸

Acquired Megaesophagus

The etiology for acquired idiopathic megaesophagus is unknown. The current theory is that a defect in the afferent neural pathway causes reduced responsiveness to esophageal distention.¹

The diffuse neuromuscular dysfunction of acquired secondary megaesophagus can be caused by a variety of neuromuscular, immune-mediated, endocrine, gastrointestinal, paraneoplastic, and toxic diseases.^{1,2,9-12}

Neuromuscular and Immune-Mediated Causes

The most common neuromuscular disorders associated with megaesophagus include MG and generalized inflammatory myopathies such as polymyositis and those associated with infectious diseases.^{13,14} Less common neuromuscular disorders associated with megaesophagus include myopathies such as muscular dystrophies, dysautonomia, storage diseases, and neurogenic muscular atrophy.^{13,14} Because the canine esophagus is composed predominantly of striated muscle, any neuromuscular disease that affects limb muscles can affect the esophagus.⁸

Of all acquired megaesophagus cases, approximately 25% are secondary to MG.¹ Acquired MG is a disorder of neuromuscular transmission due to immune-mediated destruction of postsynaptic AChRs in skeletal muscle by AChR antibodies.² Acquired MG can present in focal, generalized, and acute fulminating forms.⁸ Focal MG can present with various degrees of esophageal, facial, laryngeal, or pharyngeal dysfunction.⁸ Ninety percent of dogs with generalized MG have megaesophagus.⁸ Although acquired MG can affect dogs of any age older than a couple of months, most affected dogs are between 2 and 3 years of age or older than 9 years.¹⁵ Acquired MG occurs most often in German shepherds and golden retrievers.³ Akitas and Scottish terriers have an increased relative risk for MG.¹⁶ Familial and breed-associated forms have been described in Newfoundlands and Great Danes.^{6,17} Affected feline breeds include the Abyssinian, Somali, and Siamese.^{1,18}

Approximately 14% of dogs with diagnosed generalized inflammatory myopathies present with megaesophagus.¹⁴ Patients presenting with generalized weakness, stiff gait, dysphagia, or diffuse muscle atrophy may have myositis.¹⁴ Serum creatine kinase (CK) activities may or may not be elevated, so a normal CK activity does not rule out myositis. *Generalized inflammatory myopathy* is a comprehensive term used to group causes of diffuse myositis. Generalized inflammatory myopathies can have immune-mediated (polymyositis), infectious, or preneoplastic etiologies.¹⁴ Protozoal,

rickettsial, spirochetal, and fungal infections can be associated with myositis. Preneoplastic syndromes differ from paraneoplastic syndromes in timing. Preneoplastic syndromes occur with occult cancer. Preneoplastic syndromes that can cause myositis include bronchogenic carcinoma, lymphoma, myeloid leukemia, and tonsillar carcinoma.¹⁴

Megaesophagus associated with distemper is due to demyelination.¹⁹ Neurologic signs can develop 1 to 3 weeks or even months after initial recovery. The nerve damage is due to an inflammatory response to the viral antigens in neurons and glial cells.²⁰ This results in gray matter damage and demyelination.²⁰ Dogs with clinical signs of nasal and footpad hyperkeratosis are more likely to develop central nervous system disease.²¹

Generalized tetanus is known to cause esophageal dysfunction in dogs and humans.²² Classic clinical signs in dogs include a stiff gait, an outstretched or dorsally curved tail, the “joker’s smile” (erect ears, drawn-back lips, wrinkled forehead), protrusion of the third eyelids, enophthalmos, trismus, increased salivation, and a strong response to stimuli.²²

Although dysautonomia is rare, megaesophagus is a common finding in these patients.²³ Dysautonomia is an idiopathic autonomic nerve disorder of cats and dogs that is suspected to be immune mediated. All ganglia and sympathetic and parasympathetic nerves are affected, with neuronal cell body damage and axonal degeneration.²⁴ Within 1 to 7 days, patients experience a fulminant loss of autonomic nervous system function, followed by constipation, dry mucous membranes, pupillary dilation, prolapsed nictitating membranes, diminished pupillary light response, bradycardia, areflexic anus, and bladder atony.²³

Glycogen storage diseases (GSDs) are inborn errors of glycogen metabolism.²⁵ Of the eight human GSD types, small animal equivalents have been published for GSD I A (glucose-6-phosphatase deficiency), GSD II (α -glucosidase deficiency), GSD III (debrancher enzyme amylo-1,6-glucosidase deficiency), GSD IV (branching enzyme α -1,4-D-glucan), and GSD VII (phosphofructokinase deficiency).²⁶ Only GSD II, which is documented in Swedish Lapland dogs, has been associated with megaesophagus.²⁵

A clinical presentation of megaesophagus associated with gait abnormalities and laryngeal paralysis is suggestive of laryngeal paralysis–polyneuropathy complex (LP-PNC).¹³ Megaesophagus is documented in most dogs that are affected with LP-PNC, which is due to neurogenic muscular atrophy.¹³ The intrinsic laryngeal and appendicular skeletal muscles are affected. LP-PNC is documented in Dalmatians, Leonbergers, Pyrenean mountain dogs, and rottweilers.¹³ Puppies usually present between 2 and 6

Key Points

- Incidental esophageal dilation is associated with excitement, aerophagia, general anesthesia, and vomiting.
- Measurement of acetylcholine receptor antibody titer is not a diagnostic tool for congenital MG.
- The best food consistency to minimize regurgitation varies with each patient.

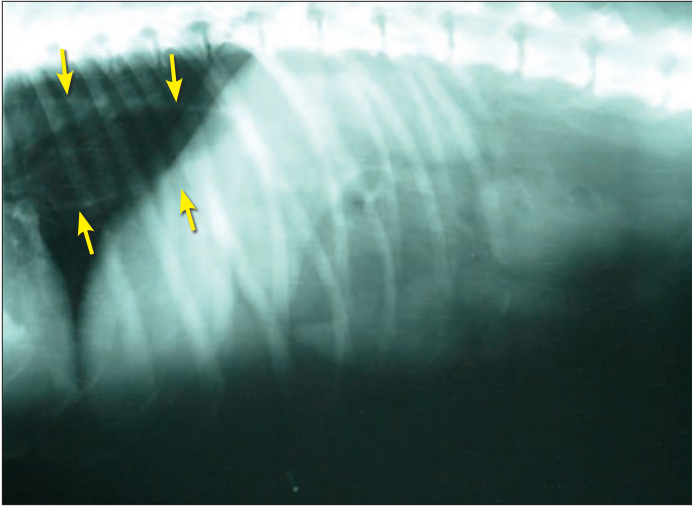


Figure 1. Radiographic appearance of a dilated esophagus (lateral abdominal view; arrows). Megaesophagus was an incidental finding. This patient was diagnosed with esophagitis associated with chronic inflammatory bowel disease and gastrointestinal reflux. *Courtesy of Dr. J. Kramer, Huntington Animal Hospital, Huntington Station, NY.*

months of age¹³; however, in Leonbergers, onset is delayed to 1 to 9 years of age.¹³ A demyelinating polyneuropathy has been reported in a family of miniature schnauzer dogs presenting with predominantly respiratory dysfunction associated with laryngeal paralysis and esophageal dilatation.²⁷

Endocrine Causes

Hypoadrenocorticism and hypothyroidism are associated with reversible megaesophagus. Patients with hypoadrenocorticism may have megaesophagus due to electrolyte imbalances and a cortisol deficiency. Electrolyte imbalances cause altered membrane potentials, which results in decreased neuromuscular function. In addition, muscle weakness is a consequence of deficient cortisol.²⁸

The association between megaesophagus and hypothyroidism has yet to be understood. Hypothyroidism is prevalent in some breeds that are predisposed to megaesophagus and laryngeal paralysis.^{1,29,30} Megaesophagus occurs in 3% of hypothyroid dogs.²⁹ Resolution of megaesophagus once the thyroid is regulated has been reported.³¹ Aspiration pneumonia may cause a sick euthyroid syndrome that may be misdiagnosed as hypothyroidism.²⁹

Gastrointestinal Causes

Gastrointestinal disorders associated with acquired megaesophagus include esophagitis, esophageal obstruction, gastric dilatation–volvulus, and hiatal hernia. In cats, acquired secondary megaesophagus is due to pyloric dysfunction.¹

Esophagitis is a common finding associated with megaesophagus.¹ It may or may not precede megaesophagus. In patients with esophagitis, secondary megaesophagus develops due to chemical or obstructive irritation. Gastric reflux contains gastric acid, pepsin, bile salts, and trypsin, all of which cause esophageal inflammation and ultimately decrease esophageal motility⁷ (FIGURE 1).

Esophageal obstructions can be caused by esophageal foreign bodies, neoplasia, strictures, or vascular ring anomalies. Foreign bodies can cause a partial or complete mechanical obstruction. Peristaltic spasms over the retained foreign object cause tissue edema and mucosal abrasions. Although possible in any small dog, there seems to be a higher incidence of esophageal foreign bodies in young terriers. Because these terriers are young, this may be a condition of delayed esophageal maturation.⁷

Foreign bodies or chronic gastroesophageal reflux (GER) can cause esophageal strictures, which occur secondary to mucosal healing attempts.^{32,33} Esophageal damage that penetrates the submucosa and muscularis layers causes inflammation resulting in collagen deposition and fibrous connective tissue stricture.^{32,33}

Extraluminal esophageal obstruction is most commonly associated with vascular anomalies. In 95% of patients with secondary megaesophagus due to a vascular ring anomaly, the cause is a persistent right aortic arch.³⁴ Other vascular anomalies associated with secondary megaesophagus include persistent right or left subclavian arteries, double aortic arch, persistent right dorsal aorta, left aortic arch, right ligamentum arteriosum, aberrant intercostal arteries, and persistent left cranial vena cava.^{34,35}

Dogs with chronic or recurrent gastric dilatation with or without volvulus have an increased risk of developing megaesophagus.²⁸ In these dogs, megaesophagus is due to decreased lower esophageal sphincter (LES) tone caused by a combination of esophagitis from chronic GER or vomiting; chronic intermittent obstruction of the LES; increased intragastric and intraabdominal pressures; and delayed gastric emptying.^{7,28}

In patients with hiatal hernia, the esophagus is essentially obstructed. Four types of hiatal hernias have been described in humans.¹⁰ Two of these types are applicable to animals. Type I is the “sliding” hernia, defined as intermittent cranial displacement of the abdominal esophagus, LES, and gastric cardia through the hiatus.¹⁰ Type II is the paraesophageal hernia, in which the gastroesophageal junction remains in its normal anatomic position; however, the stomach and abdominal organs enter the caudal mediastinum through a defect adjacent to the esophageal hiatus.¹⁰

Paraneoplastic Causes

According to one study, megaesophagus was present in 40% of dogs with thymoma.³⁶ The incidence of thymoma in dogs with MG is 3%; in cats with MG, the incidence is 26%.^{16,18} In humans, thymomas have increased production of CD4⁺CD8⁺ T cells and lack antigen-presenting cells that function for negative selection. This combination results in autoimmune disease.³⁷ The prognosis for nonresectable thymoma in a dog with MG and megaesophagus is poor. However, complete thymic resection can result in resolution of megaesophagus and a decrease in AChR antibody titer.^{8,38}

Toxic Causes

Toxic substances that can cause megaesophagus include lead, organophosphates, and snake venom.^{3,28} Low-level lead exposure causes severe abdominal pain, vomiting, diarrhea, and megaesophagus.³⁹ Lead intoxication can occur from ingestion of batteries,

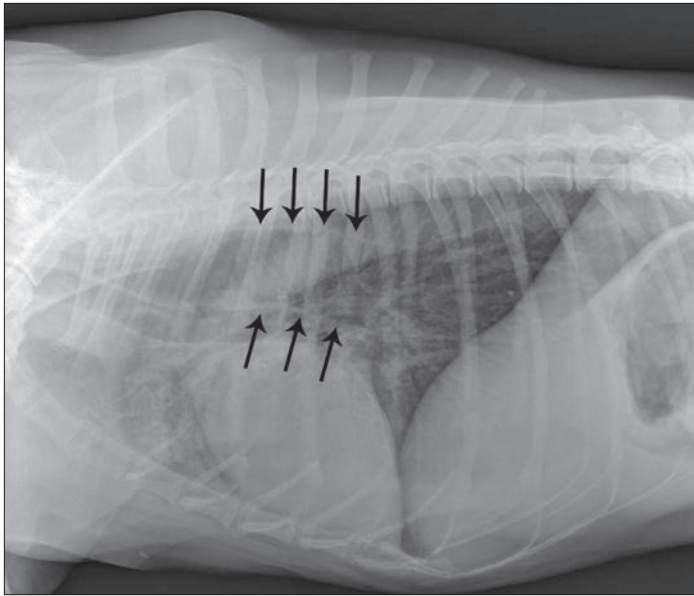


Figure 2. Radiographic appearance of a dilated esophagus (arrows). Note the radiopaque walls of the esophagus and luminal dilation with air on both sides of the diaphragmatic silhouette.

fishing line weights, lead-based paint, linoleum, and plumbing or solder supplies. Organophosphate toxicosis should be suspected if a patient presents with concurrent weakness and cerebellar signs.⁴⁰ Organophosphates exist in flea collars and insecticides. They irreversibly bind to acetylcholinesterase, causing a cholinergic crisis (salivation, lacrimation, urination, defecation). Australian tiger snake envenomation causes a rapidly progressing myopathy of skeletal muscle.³ If not lethal, Australian tiger snake envenomation has a 75% recovery rate for normal esophageal function.³

Diagnosis

Diagnostic Imaging

Thoracic radiography is diagnostic for most cases of megaeosophagus.^{1,3} Common findings include a prominent, dilated esophagus that can be filled with air or ingesta² (FIGURE 2). The degree of esophageal dilation has no diagnostic value in determining the etiology.² Underlying causes of megaeosophagus that may be revealed by radiography include neoplasia, foreign body, vascular ring anomaly, gastric dilatation–volvulus, and hiatal hernia. Normal midline tracheal location does not exclude a vascular ring anomaly; however, focal leftward deviation of the trachea near the cranial border of the heart on a dorsoventral or ventrodorsal view is a reliable sign of persistent right aortic arch in young dogs that regurgitate after eating solid food.^{34,35} Radiographic findings of megaeosophagus with concurrent aspiration pneumonia or a distended stomach, small bowel, or urinary bladder should raise suspicion for dysautonomia.²³ Incidental esophageal dilation does occur and is associated with excitement, aerophagia, general anesthesia, and vomiting.²

If thoracic radiographic findings of megaeosophagus are questionable, a barium contrast esophagram can confirm dilation and

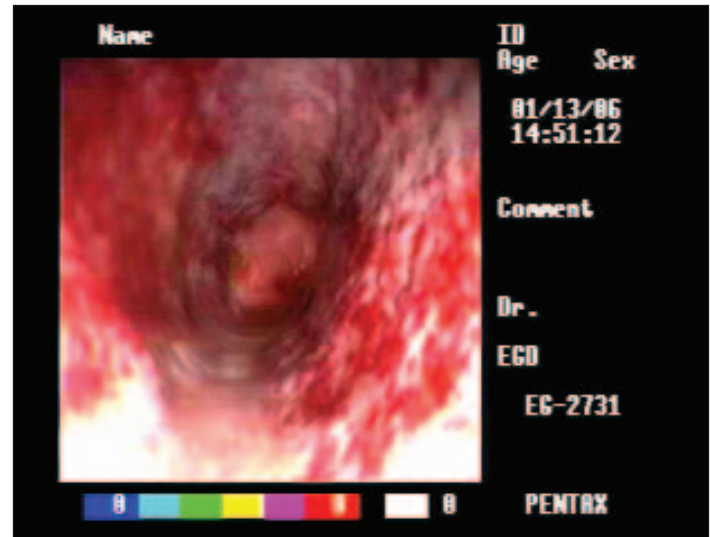


Figure 3. Endoscopic appearance of the esophagus following endoscopic retrieval of a rawhide that had been lodged in the distal esophagus. Note the severe multifocal mucosal abrasions. The patient (a terrier) was treated with liquid sucralfate and broad-spectrum antibiotics. Fortunately, follow-up endoscopy revealed that no stricture developed.

mechanical obstruction. Barium accumulates within the distended esophagus. Focal narrowing of the esophagus at the cardiac base is suggestive of a vascular ring anomaly.^{1,34} However, the diagnostic benefit of a contrast study should be weighed against the potential for aspiration of contrast agent.

Fluoroscopy evaluates pharyngeal motility and the presence and intensity of esophageal peristalsis. However, this diagnostic modality is not essential for diagnosis of megaeosophagus and may not be readily available. It can be helpful in cases of MG or esophagitis. MG can selectively affect only the pharyngeal and esophageal musculature without more overt clinical signs. Also, in cases of mild esophagitis, fluoroscopy may be of greater diagnostic value than a contrast esophagram in detecting hypomotility.

Esophagoscopy is rarely indicated for a diagnosis of megaeosophagus, but it can be helpful for suspected cases of obstructive disease or reflux esophagitis³⁴ (FIGURE 3). Esophagoscopy may identify an esophageal stricture due to a vascular ring anomaly, but it cannot differentiate the type of vascular ring anomaly.³⁴

Laboratory Testing

A complete blood count (CBC), serum chemistry panel that includes CK activity, and urinalysis should be performed for all regurgitating patients and those in which megaeosophagus is suspected. In addition, an AChR antibody titer test should be performed in all cases of acquired megaeosophagus. AChR antibody testing is performed by the Comparative Neuromuscular Laboratory in the School of Medicine at the University of California, San Diego. Information regarding sample submission can be obtained at <http://vetneuromuscular.ucsd.edu/>. Corticosteroid therapy at immunosuppressive dosages for longer than 7 to 10 days lowers AChR antibody levels, so a pre-corticosteroid serum sample is



Figure 4. This specially made feeding chair was designed by an owner of a megaesophagus patient. The chair aids in feeding and in maintaining a postprandial upright position.

recommended.⁴¹ Additional diagnostics are performed based on the history, physical examination, and preliminary laboratory findings.¹ The diagnostic objective is to determine whether the megaesophagus is associated with a potentially treatable disorder. For example, MG, hypothyroidism, hypoadrenocorticism, polymyositis, and lead poisoning all have specific treatments, whereas treatment for idiopathic megaesophagus is limited to supportive and symptomatic management.

Although CBC, chemistry panel, and urinalysis results do not provide definitive information regard-

ing MG, they may exclude other causes of muscle weakness or help identify a concurrent disease.¹⁵ Elevated serum CK activity occurs with muscle damage associated with some myopathies (inflammatory, necrotizing, and dystrophic) and muscle trauma and may be mildly elevated in patients in extended recumbency or after intramuscular injections.⁴² Definitive diagnosis of acquired MG requires an AChR antibody titer test.⁸ However, this test is not useful in diagnosing congenital MG, which is a result of structural or functional AChR abnormalities and not immune-mediated damage. Therefore, congenitally affected dogs and cats do not have measurable circulating AChR antibodies.^{2,8} In the future, diagnosis of congenital MG will depend on mutational analysis of candidate genes.

Edrophonium chloride, a short-acting acetylcholinesterase drug, can be administered to support a presumptive diagnosis of congenital or acquired MG.⁸ Before administration of the edrophonium, the patient is exercised until fatigued. The appearance of MG fatigue can include weakness, stiff gait, collapse, inspiratory stridor, or a reduced palpebral reflex.⁴³ After fatigue is induced, edrophonium is administered IV (0.1 to 0.2 mg/kg).⁸ A positive response is characterized by improved muscle strength. This commonly occurs within 30 seconds of the edrophonium injection, and weakness returns within 5 minutes. Temporary improvement of generalized muscle weakness is suggestive of, but not definitive for, MG.⁸ The degree of megaesophagus is not affected by this test; however, improvement in motility may be observed if evaluated by fluoroscopy after administration of contrast agent.

Hypercholesterolemia, hypertriglyceridemia, and hyponatremia with or without the presence of a normochromic, normocytic, nonregenerative anemia is suggestive of hypothyroidism.⁴⁴ In most cases, low total T₄, elevated canine thyroid stimulating

hormone, and low free T₄ levels confirm the diagnosis of hypothyroidism.⁴⁴

Dogs with typical or atypical hypoadrenocorticism can present with megaesophagus. Hyperkalemia and hyponatremia are suggestive of typical hypoadrenocorticism. A low sodium:potassium ratio is not definitive for hypoadrenocorticism, even with studies that found a sodium:potassium ratio <15 to be more diagnostic for hypoadrenocorticism than a ratio of 27:1.⁴⁵ Typical and atypical hypoadrenocorticism are diagnosed with an ACTH stimulation test.⁴⁵

Nucleated erythrocytes without anemia or basophilic stippling of red blood cells is suggestive of lead poisoning. These abnormalities are caused by transportation of lead to bone marrow.⁴⁶ Serum blood lead tests are commercially available.

Organophosphate toxicosis can be excluded by measuring cholinesterase in a whole blood sample. A cholinesterase activity less than 25% to 50% of normal is suggestive of organophosphate toxicosis.⁴⁷

Additional diagnostic tests may include antibody titers for *Toxoplasma gondii*, *Neospora caninum*, *Borrelia burgdorferi*, *Ehrlichia canis*, and *Rickettsia rickettsii*; electromyography; measurement of nerve conduction velocity; and muscle and nerve biopsies to exclude myopathic and neuropathic disorders. Atropine, histamine, or pilocarpine tests can be performed to exclude dysautonomia.^{1,2,9,13,14,23,48} Atropine is administered IV (0.02 mg/kg) or SC (0.04 mg/kg); lack of an increase in heart rate is supportive of dysautonomia.²³ When the intradermal histamine test (0.01375 mg per dog) is used, absence of a wheal and flare within 15 minutes is supportive of dysautonomia.²³ The histamine test has limited value in cats, as there is no significant difference in the histamine response between dysautonomic and control cats.²⁴ If miosis does not occur after topical ophthalmic administration of pilocarpine 0.1%, a presumptive diagnosis of dysautonomia can be made.²³

Treatment

Treatment for idiopathic megaesophagus is largely supportive and symptomatic with periodic rechecks. Thoracic radiography is advised to monitor progress of esophageal dilation and aspiration pneumonia. Treatment for acquired secondary megaesophagus depends on managing the underlying specific disease process in addition to providing supportive and symptomatic care. Medications should be in liquid (not pill) form to enhance movement to the stomach and avoid accumulation within the esophagus, which can lead to esophageal irritation and nontherapeutic medication levels. If accumulated medication passes into the stomach, overdose may occur.

Supportive and Symptomatic Care

Nutrition

Nutritional needs must be met and regurgitation minimized. This can be accomplished by frequent feeding of small, high-calorie meals with the patient in a cranially elevated position (FIGURE 4). This position uses gravity to help move the ingesta to the stomach.¹ The optimal food consistency to minimize regurgitation varies

with each patient, so experimentation is encouraged.¹ The use of enteral feeding may be needed in weak patients or in patients in which regurgitation cannot be controlled by other methods.⁸ In these cases, a gastrotomy tube should be placed.¹⁵ In our clinical experience, a percutaneous endoscopic gastrostomy tube or low-profile gastrotomy tube is effective. Nasoesophageal or esophageal tubes are not advised because they increase regurgitant volume, raising the risk of aspiration pneumonia.

Treatment of Secondary Complications

Aspiration pneumonia and esophagitis are the most common complications of megaesophagus.¹ For aspiration pneumonia, administration of a broad-spectrum antibiotic is advised.⁸ Culture and sensitivity testing of a transtracheal wash or bronchoalveolar lavage sample can be helpful in choosing an antibiotic; however, the risk of obtaining the sample should be considered.

Esophagitis can result in esophageal stricture within 1 to 3 weeks; therefore, eliminating further mucosal damage and allowing the mucosa to heal are important considerations.³³ Liquid sucralfate is advised because it binds to eroded mucosa, allowing it to heal.^{1,33} For megaesophagus secondary to esophagitis, addressing the underlying cause of GER is the key to improvement. Antacids (e.g., calcium carbonate), H₂-blockers, and proton-pump inhibitors are all used to lower gastric acidity and may prevent esophagitis due to GER.³³

Promotility drugs (e.g., metoclopramide, cisapride) have no current documented benefit in managing canine megaesophagus but may have a role in managing feline megaesophagus. This is due to the differences in feline and canine anatomy and the mechanism of action of these drugs. Metoclopramide and cisapride act on smooth muscle. They have no effect on striated muscle. The canine esophagus has striated muscle its entire length.^{1,33} Cats have smooth muscle within the distal esophagus, so cisapride may improve lower esophageal smooth muscle motility in cats.^{33,49} Metoclopramide and cisapride are not advised in canine megaesophagus patients because these drugs increase the LES tone, which slows esophageal emptying and contributes to further regurgitation.^{42,49–52} Bethanecol may be a better option for dogs as it is documented to stimulate esophageal propagating contractions in skeletal muscle by stimulating cholinergic receptors.¹

Acquired Secondary Megaesophagus

Treatment for acquired secondary megaesophagus is based on a definitive diagnosis. For example, the cornerstone of treatment for this form of MG is anticholinesterase drugs.⁸ If an animal is confirmed by positive AChR antibody titer as having MG and pyridostigmine alone does not control the clinical signs, then other agents are indicated. These can include either low-dose prednisone (not an immunosuppressive dosage, which can worsen the weakness) or immunosuppressants such as azathioprine, mycophenolate mofetil, or cyclosporine.⁵³ Although anticholinesterase therapy does not decrease the AChR antibody titer, many dogs with acquired MG (if they do not develop severe megaesophagus and aspiration pneumonia and expire) go into spontaneous

remission in the absence of immunosuppression.⁵⁴ Before immunosuppressive therapy is used, pneumonia or any other infectious disease should be completely resolved.

Prognosis

The prognosis for megaesophagus varies with the underlying etiology and presence of secondary complications. Aspiration pneumonia, dehydration, and malnutrition can significantly worsen the prognosis. Congenital megaesophagus has a guarded to poor prognosis; however, there is potential for improvement of esophageal motility with maturity up to 1 year of age.^{7,10} The prognosis for congenital MG is poor due to the mechanism of the condition, lack of a specific treatment, and high complication rate of aspiration pneumonia. Acquired idiopathic megaesophagus in general has a guarded to poor prognosis due to the common occurrence of aspiration pneumonia and malnutrition. Morbidity and mortality depend on the degree and nature of the underlying disease and client compliance.¹ In the absence of severe aspiration pneumonia or thymoma, the success rate for acquired MG can be good with early diagnosis and appropriate management.⁵³ Spontaneous remission of acquired MG with resolution of megaesophagus can also occur within an average of 6 months.⁵⁴ However, many myasthenic dogs die of aspiration pneumonia during the first month after diagnosis, so the overall prognosis is still guarded. With the exception of the acute fulminating form of myasthenia,⁵⁵ there is no association between the severity of MG and the possibility of remission.⁵⁴

In one study, 39% of dogs with immune-mediated polymyositis had clinical improvement of their megaesophagus with continued medical management.¹⁴ However, early diagnosis and initiation of appropriate therapy are key to a good clinical outcome. Evaluation of muscle biopsy samples early in the course of the disease to establish a diagnosis is critical. The prognosis for pre- and paraneoplastic myositis is poor due to the underlying cancer.¹⁴ Dysautonomia is progressive, with a survival rate of <25% in cats over 18 months.²⁴ Prognostic indicators include showing response to therapy (e.g., maintenance of body weight with oral feedings, fecal and urinary continence) within 7 to 10 days.²⁴

Conclusion

Megaesophagus is common in dogs and less common in cats. Regurgitation is the most common clinical sign of megaesophagus at presentation. Diagnosis of megaesophagus is made radiographically, and the primary cause should be evaluated with appropriate diagnostic testing. Idiopathic megaesophagus is a diagnosis of exclusion. Management of megaesophagus is supportive unless an underlying cause is identified. The prognosis for megaesophagus depends on the presence of aspiration pneumonia and the underlying condition.

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1. _____ is not a clinical sign of megaesophagus.

- a. Regurgitation
- b. Halitosis
- c. Weight gain
- d. Coughing

2. Which condition is matched with the wrong clinical signs?

- a. congenital megaesophagus: regurgitation at time of weaning
- b. pneumonia: moist cough, dyspnea, and fever
- c. myositis: generalized weakness, stiff gait, and dysphagia
- d. organophosphate toxicosis: stiff gait, erect ears, and strong response to stimuli

3. In which breed is congenital megaesophagus not commonly documented?

- a. Parson Russell terrier
- b. Malinois
- c. Newfoundland
- d. Samoyed

4. Which statement regarding feline megaesophagus is false?

- a. Congenital and acquired megaesophagus occur in cats.
- b. Abyssinian, Somali, and Siamese cats may have a familial predisposition for megaesophagus.
- c. In cats, acquired secondary megaesophagus is not due to pyloric dysfunction.
- d. Metoclopramide may have a role in managing feline megaesophagus.

5. Which statement is true regarding diagnosing megaesophagus?

- a. Thoracic radiography is diagnostic for most cases of megaesophagus.
- b. Radiography is never useful in revealing underlying causes of megaesophagus.
- c. Normal midline tracheal location excludes a vascular ring anomaly.
- d. Incidental esophageal dilation does not occur.

6. Which statement regarding MG is true?

- a. All dogs with generalized MG have megaesophagus.
- b. Acquired MG occurs more often in German shepherds and golden retrievers than in other canine breeds.
- c. Definitive diagnosis of congenital MG requires an AChR antibody test.
- d. Dogs with acquired MG do not have measurable circulating anti-AChR antibodies.

7. Which CBC/serum chemistry result is not correctly matched with an appropriate etiology?

- a. hypercholesterolemia and hyponatremia: hypothyroidism
- b. hyperkalemia and hyponatremia: hyperadrenocorticism
- c. normochromic, normocytic, nonregenerative anemia: hypothyroidism
- d. nucleated erythrocytes without anemia: lead poisoning

8. Which therapy is not recommended as supportive care for megaesophagus?

- a. enteral feeding with an esophageal tube
- b. broad-spectrum antibiotics
- c. liquid sucralate
- d. antacids and H₂ blockers

9. Which statement is true regarding medication administration for a patient with diagnosed megaesophagus?

- a. Formulation (liquid or pill) does not matter.
- b. Pills cannot result in subtherapeutic medication levels.
- c. Pills cannot result in overdose.
- d. Pills can lead to esophageal irritation.

10. Which statement regarding megaesophagus prognosis is true?

- a. The prognosis for congenital MG is good.
- b. Aspiration pneumonia contributes to a poor prognosis.
- c. Malnutrition does not contribute to the prognosis.
- d. Congenital idiopathic megaesophagus has a good prognosis.