

# Megaesophagus

## (Enlargement of the Esophagus)

### Basics

#### OVERVIEW

- The esophagus is part of the digestive tract; it is a muscular tube that runs from the throat to the stomach; food and liquids are moved through the esophagus by peristalsis (wave-like motion produced by contraction and relaxation of muscles) toward the stomach
- Megaesophagus is defined as a generalized, diffuse enlargement (dilation) of the esophagus, in which peristaltic movement is decreased to absent

#### GENETICS

- Congenital (present at birth)/inherited form—inherited in wirehaired fox terriers as an autosomal recessive trait and in miniature schnauzers as an autosomal dominant or 60% penetrance autosomal recessive trait
- Other breeds reported include dachshund, German shepherd dog, Great Dane, Irish setter, Labrador retriever, pug (one litter with four puppies affected), and Chinese shar-pei
- Myasthenia gravis (a disorder of neuromuscular transmission characterized by muscular weakness and excessive fatigue) may be congenital (present at birth) in Jack Russell terriers, Springer spaniels, smooth fox terriers, dachshunds and Samoyeds
- Acquired (present later in life/after birth) form—many diseases may have an association with megaesophagus, some have genetic susceptibilities



#### SIGNALMENT/DESCRIPTION OF PET

##### Species

- Cats
- Dogs are affected more commonly than cats

##### Breed Predilections

- Dog—wirehaired fox terrier, miniature schnauzer, dachshund, German shepherd dog, Great Dane, Irish setter, Labrador retriever, pug, Chinese shar-pei
- Cat—Siamese and Siamese-related cats

##### Mean Age and Range

- Congenital (present at birth) cases present soon after birth or at weaning from liquid diets to more solid foods
- Acquired (present later in life/after birth) cases may be seen at any age, depending on the cause of the megaesophagus

#### SIGNS/OBSERVED CHANGES IN THE PET

- Owners often report vomiting (forceful ejection of stomach contents up through the esophagus and mouth); the veterinarian must differentiate vomiting from regurgitation (return of food or other contents from the esophagus or stomach back up through the mouth—not forceful)
- Regurgitation (considered the hallmark sign of megaesophagus); difficulty swallowing (known as “dysphagia”); coughing, fever and nasal discharge with aspiration pneumonia (inflammation of the lungs, caused by accidentally inhaling food, vomit, or liquids); ravenous appetite or lack of appetite; weight loss or poor growth; and excessive drooling (known as “ptyalism”)
- Swelling of the neck, bad breath (known as “halitosis”); weight loss or poor growth; extreme weight loss with muscle wasting (known as “cachexia”); weakness
- Other signs such as a change in voice (known as “dysphonia”) depend upon underlying cause of megaesophagus
- Pets will be assessed carefully for other muscle and/or nervous system deficits that may indicate generalized disease

## CAUSES

### **Congenital (Present at Birth)**

- Unknown cause (so-called “idiopathic megaesophagus”)
- Myasthenia gravis (a disorder of neuromuscular transmission characterized by muscular weakness and excessive fatigue) is a rare cause

### **Acquired/Adult Onset**

- Unknown cause (so-called “idiopathic megaesophagus”) is most common type
- Neuromuscular disease: myasthenia gravis (25% of cases in dogs); systemic lupus erythematosus (an autoimmune disease that affects many organs and tissues of the body); inflammation of the muscles (known as “myositis”) or other muscle disease (known as “myopathic disease”); dysfunction of the autonomic nervous system (known as “dysautonomia”; more common in cats); tetanus or botulism; trauma to the vagus nerves (nerve that goes to the throat, voice box, windpipe, esophagus, gastrointestinal tract, and other organs); possible association between paralysis of the voice box or larynx (known as “laryngeal paralysis”) and megaesophagus has been identified
- Esophageal blockage or obstruction: congenital (present at birth) abnormal development of major arteries of the heart, resulting in the esophagus being trapped by the blood vessels causing obstruction (abnormal arteries known as a “vascular ring anomaly”); cancer (primary esophageal cancer or cancer involving tissues around the esophagus); narrowing or stricture of the esophagus; foreign body; granuloma
- Toxicity: lead, thallium, drugs that inhibit acetylcholinesterase, an enzyme in the central nervous system (drugs are known as “anticholinesterase drugs”); an example is acrylamide
- Endocrine disease: inadequate levels of steroids produced by the adrenal glands (known as “hypoadrenocorticism” or “Addison's disease”); inadequate levels of thyroid hormone (known as “hypothyroidism”)
- Miscellaneous: thymus tumor (known as “thymoma”) in 25% of cats; stomach dilates with gas and/or fluid (known as “gastric dilatation”), and subsequently rotates around its short axis (known as “volvulus”)—condition known as “gastric dilatation-volvulus” or “bloat”; hiatal hernia; folding of the stomach up into the esophagus (known as “gastroesophageal intussusception”); inflammation of the esophagus (known as “esophagitis”) due to parasites or reflux of stomach contents

## Treatment

### HEALTH CARE

- Therapy for underlying cause of megaesophagus (when applicable) should be instituted
- Most important aspects are meeting nutritional requirements and treating or preventing aspiration pneumonia (inflammation of the lungs, caused by accidentally inhaling food, vomit, or liquids)
- Aspiration pneumonia may require oxygen therapy, administration of medication in a fine spray (known as “nebulization”) and efforts to dislodge secretions in the lungs and to induce coughing (known as “coupage”), fluid therapy with balanced electrolyte solution

- Pets may be recumbent and require soft bedding and should be maintained on their chests (known as “sternal recumbency”) or turned to alternate down side every 4 hours

## ACTIVITY

- Depending on cause of megaesophagus, restricted activity may not be necessary

## DIET

- Nutritional requirements should be calculated precisely; your pet's veterinarian can provide information on the amount and type of food needed
- Experimentation with different food consistencies is essential—liquid gruel, small meatballs, slurries made using a blender, and others may be used
- Some affected pets benefit from tube feedings
- Food and water bowls should be elevated (45–90 degrees from floor)
- The pet should be maintained in an upright position 10–15 minutes after eating or drinking; a special “Bailey” chair may help with keeping upright

## SURGERY

- Surgical intervention is indicated for congenital (present at birth) abnormal development of major arteries of the heart, (a vascular ring anomaly); opening between a bronchus (large airway) and the esophagus (known as “bronchoesophageal fistula”); some foreign bodies and other obstructive lesions; or surgical removal of the thymus (known as “thymectomy”)
- Balloon dilation (a mechanical means of stretching the tissue) is indicated for cases of narrowing of the esophagus (known as “esophageal stricture”)
- Surgical “correction” of megaesophagus is not recommended

## Medications

Medications presented in this section are intended to provide general information about possible treatment. The treatment for a particular condition may evolve as medical advances are made; therefore, the medications should not be considered as all inclusive

- Antibiotics for aspiration pneumonia (broad-spectrum antibiotics or antibiotic selection ideally based on culture and sensitivity from transtracheal wash [TTW] or bronchoalveolar lavage [BAL]—TTW and BAL are techniques used to obtain samples from the airways for microscopic evaluation and bacterial culture and sensitivity)
- Specific therapy for underlying cause, if indicated: examples include drugs to decrease the immune response (known as “immunosuppressive drugs”)—should be used with caution if pneumonia present—for immune-mediated disease; pyridostigmine for myasthenia gravis (a disorder of neuromuscular transmission characterized by muscular weakness and excessive fatigue); prednisone for Addison’s disease
- H<sub>2</sub> blockers for treatment of inflammation of the esophagus (esophagitis)—decrease stomach acid; include ranitidine, famotidine; proton pump inhibitors can be used in severe cases, such as omeprazole, pantoprazole
- Drugs that improve the propulsion of contents through the stomach and intestines (known as “gastrointestinal prokinetic agents,” such as metoclopramide, cisapride)—may increase tone of the muscle between the stomach and esophagus (gastroesophageal sphincter); may increase risk of aspiration pneumonia
- Metoclopramide increases lower esophageal sphincter tone, increases stomach motility and may increase esophageal motility
- Cisapride is effective for backward or reverse flow of stomach contents into the esophagus (known as “esophageal reflux”) but not megaesophagus; it may slow movement of food and liquids through the esophagus (known as “slowed esophageal transit time”); may be more helpful in cats due to smooth muscle in the lower third of the esophagus
- Other motility agents (such as nizatidine) have not been evaluated for esophageal motility

## Follow-Up Care

### PATIENT MONITORING

- Chest x-rays (radiographs) will be repeated when aspiration pneumonia is suspected
- Pets with pneumonia may require complete blood count, blood-gas analysis to evaluate oxygen and carbon dioxide levels in the blood, and bronchoalveolar lavage
- Repeat chest X-rays to monitor congenital megaesophagus since spontaneous resolution may occur
- Pets should be assessed and weighed regularly to evaluate disease progression and ensure adequate nutritional intake

## **PREVENTIONS AND AVOIDANCE**

- If an esophageal foreign body is identified, it should be removed as quickly as possible

## **POSSIBLE COMPLICATIONS**

- Aspiration pneumonia (inflammation of the lungs, caused by accidentally inhaling food, vomit, or liquids)
- Other complications depend on underlying cause of megaesophagus

## **EXPECTED COURSE AND PROGNOSIS**

- Congenital (present at birth) megaesophagus cases have a guarded prognosis (20–46% recovery)
- Miniature schnauzers may have better prognosis
- Prognosis may be improved with identification and treatment of specific cause of megaesophagus (for example, inadequate levels of steroids produced by the adrenal glands [hypoadrenocorticism, “Addison’s”], congenital [present at birth] abnormal development of major arteries of the heart, resulting in the esophagus being trapped by the blood vessels causing obstruction [vascular ring anomaly])
- Roughly 50% of pets with myasthenia gravis (a disorder of neuromuscular transmission characterized by muscular weakness and excessive fatigue) respond to therapy
- Prognosis for megaesophagus of unknown cause (idiopathic megaesophagus) as an adult-onset disease is poor
- Owner dedication is crucial to prognosis

## **Key Points**

- Most cases of megaesophagus will require lifelong therapy, even if an underlying cause is found; client dedication is important for long-term management
- Most pets with adult onset megaesophagus will succumb to aspiration pneumonia (inflammation of the lungs, caused by accidentally inhaling food, vomit, or liquids) or progression of underlying disease