

Megaesophagus and Esophagus Dysmotility

The esophagus plays a critical role that we rarely appreciate until it is not working properly. A poorly functioning esophagus is a serious problem because of the potential for aspiration of material into the lungs. Esophageal problems cause significant difficulty with swallowing and can include megaesophagus, in which the esophagus is dilated throughout with weak contraction, or esophageal dysmotility, in which the esophagus appears structurally normal but does not function properly.

Cause

Megaesophagus is a disorder with several potential causes; it can be associated with a global neuromuscular disorder called Myasthenia Gravis as well as some hormonal conditions such as hypothyroidism and Addison's disease. It can be seen after an episode of esophageal inflammation due to toxicity or trauma and is also associated with certain tumors. Megaesophagus can also arise without a known cause; this is referred to as idiopathic megaesophagus.

Esophageal dysmotility syndromes can also occur after esophageal inflammation or trauma, but, in many cases, a direct cause cannot be determined. Some patients are born with a poorly functioning esophagus, including structural anomalies such as strictures and diverticulum or odd, pouch-like structures.

Clinical Signs

Most patients regurgitate food and water soon after eating or drinking. Regurgitation differs from vomiting in that the patient does not always retch prior to food or water coming up, instead, the patient coughs or gags and food and water come out. Patients may get some food down, but swallowing movements are exaggerated and the patient is visibly distraught. Some appear to be trying to clear their throat repeatedly and make odd roaring sounds in an effort to get comfortable. Patients with long-standing difficulty will be thin and weak and may become dehydrated, requiring intravenous fluids and support.

Patients that regurgitate are at significant risk for aspiration, in which food, water, or saliva is drawn into the airway. This is potentially immediately life-threatening, and patients that seem to be struggling to breathe after regurgitating should be evaluated immediately to receive appropriate support.

Diagnosis

Megaesophagus and esophageal dysmotility syndromes are suspected in patients with characteristic clinical signs. Megaesophagus is generally visible on thoracic radiographs (XRAY) as a large, air or fluid-filled structure that extends through the chest. Patients with esophageal dysmotility syndromes may have visible fluid or air in the esophagus on radiographs, but are different in that the entire esophagus does not appear dilated.

Because other health conditions can be associated with megaesophagus and esophageal dysmotility, blood tests, including specific tests looking for neuromuscular or hormonal conditions will be recommended. Additional imaging, such as ultrasound or computed tomography (CT scan) may be necessary if tumors or other conditions are suspected.

In some patients, evaluation of the esophagus with an endoscopic exam is recommended in order to evaluate for structural abnormalities. Extra anesthetic precautions are necessary for these patients.

Oral contrast agents such as barium are never recommended for patients with megaesophagus and esophageal dysmotility syndromes, because of the risk of aspiration of the liquid contrast, which is potentially life-threatening.

Treatment

Therapy for a patient with a poorly functioning esophagus depends upon whether or not an underlying cause can be found. Patients that have a global neuromuscular or hormonal condition are started on disease-specific therapy in addition to changes in home management with the hope that esophageal function will improve or resolve.

Patients with a poorly functioning esophagus for which disease-specific therapy is not possible are managed by changing the way the patient receives nutrition and hydration. In some cases, this means the placement of a permanent feeding tube and bypassing the esophagus altogether. Doing so does not completely eliminate the possibility of aspiration pneumonia, but it

can reduce risk. This is often necessary for patients that present in poor physical condition, or for pediatric and congenitally affected patients.

Patients without feeding tubes must receive food and water in an upright position, and remain in an upright position for several minutes after eating or drinking. This allows gravity to help move food and water through the esophagus. This practice is generally combined with altering the consistency and type of food the patient is fed.

Medications to improve overall esophageal/intestinal motility are sometimes used in patients with poor esophageal function. Additionally, antacids and anti-gas medications often provide some symptomatic relief. Recently the use of Sildenafil has made a large improvement and we are seeing better outcomes in these patients. Long-term studies and prognosis with this newer treatment have not been performed to date but results seem promising compared to older treatments.

Prognosis

The prognosis for patients with megaesophagus and esophageal dysmotility has generally been guarded as there is no way to completely prevent aspiration pneumonia in patients with poor esophageal function. Some patients, particularly those with an underlying, treatable health condition or temporary injury enjoy a complete resolution with appropriate therapy.

Without resolution of the esophageal dysfunction, lifelong commitment to altered feeding protocols can sometimes allow patients to have a good quality of life and long term survival.

Roughly 80% of patients with esophageal dysfunction that succumb to the disease die due to the development of severe aspiration pneumonia. Some patients have repeated episodes of pneumonia over many years and develop pulmonary scarring that can affect the quality of life and survival, but others recover well with aggressive therapy.

Long Term Follow-Up

Patients with megaesophagus and esophageal dysmotility syndromes are generally diagnosed and treated by the internal medicine specialists at Veterinary Specialty Center. If the patient has an underlying health condition, such as Myasthenia Gravis or cancer, additional specialists may be involved in long term patient care. Patients on multiple medications, recurrent or severe aspiration pneumonia, or with permanent feeding tubes require frequent follow-ups for lab tests, radiographs, and adjustments to home therapy. Because decisions about changes in medication, diet, and feeding instructions are based on observations made during the physical exam in addition to other testing, our recommendation is that follow-up for this disease be done at Veterinary Specialty Center. All routine preventive care should continue with your primary care veterinarian, with special precautions if anesthesia or certain medications are recommended.