WHAT BREEDERS NEED TO KNOW ABOUT CONGENITAL MEGAESOPHAGUS

By Cynthia McFadden

Anyone who has dealt with a megaesophagus puppy knows what a heartbreaking disease it is. In the 2001 Rhodesian Ridgeback Health Survey, megaesophagus was ranked first among reported gastrointestinal disorders, with an 81 percent mortality rate. This disorder occurs in many lines, and though the mode of inheritance is not clearly understood, it is likely a genetic defect.

While megaesophagus can occur in adult dogs secondary to diseases such as myasthenia gravis, Ridgeback breeders will most commonly encounter it in young pups as they are weaned to solid food, at 4 weeks or thereafter. This congenital form of megaesophagus is characterized by a lack of muscular contractions in the esophagus, which creates a pouch effect in the pup’s chest. Food cannot enter the stomach normally, but instead simply sits in the dilated esophagus, which cannot push the food into the stomach. Because there is no way to restore elasticity to the esophagus by surgical means, there is no course of treatment for this form of megaesophagus.

Another reason for congenital megaesophagus is a vascular ring anomaly such as persistent right aortic arch. In this scenario, fetal blood vessels that should have disappeared at birth create a fibrous band that externally constricts the esophagus. This causes the area of esophagus above the constriction to expand when food cannot pass through the constricted area. If caught in time, the vascular ring can be cut, and the esophagus often returns to normal. Delaying surgery may cause irreparable esophageal damage.

Esophageal laxity/dilation and vascular ring anomalies are both believed to have a hereditary component, and affected dogs should not be bred.

Megaesophagus puppies may regurgitate after eating, sometimes 20 or more times a day, and nutritional deficits may quickly appear. Such puppies will likely fail to thrive, will lose weight, and may develop aspiration pneumonia from spontaneously inhaling regurgitated food. (Regurgitation is different than vomiting. Vomiting involves muscular contractions of the abdomen -- retching -- that propel stomach contents up the esophagus. Regurgitation, by contrast, is a passive process, usually brought on by gravity – i.e., the pup puts his head down and the food just comes back out. As the food never enters the stomach from the esophagus, retching does not occur. Regurgitation can occur very quickly and quietly, and littermates may “clean up” the results before the breeder discovers them). Severely affected pups are generally euthanized by the breeder.

But megaesophagus pups may not exhibit all of these symptoms, or even any of them to a significant degree. Sometimes the only clinical signs may be repeated episodes of aspiration pneumonia, or a wet cough that fails to clear up. Some pups grow out of the disorder, whether by resumption of normal esophageal function, or a loosening of the stricture that previously prohibited food from entering the stomach. It is not clear at the time of diagnosis which pups will grow out of it and go on to enjoy a normal quality of life, and which pups will continue to be significantly affected.

Breeders who decide not to euthanize megaesophagus puppies, regardless of the degree of severity, must realize they are making a long-term commitment: They may need to keep a pup for up to a year to determine how affected it will be, and might very well need to use special equipment, such as a “Bailey chair” (www.geocities.com/bailey_chair), to keep a puppy upright (in a “begging position”) while eating. This posture allows gravity to assist food in entering the stomach as much as the pup’s anatomy will allow. Pups can also be taught to sit with their front feet elevated, such as propped on a stool, for a period of time after feeding, again permitting a “gravity-assist”. Sometimes these methods are enough for a pup to receive adequate nourishment and maintain reasonable condition, but still represent a significant time and energy commitment on the breeder’s part.

Pups that gain weight, are able to self-regulate activity levels following feeding, and can avoid repeated episodes of pneumonia generally have a better long-term prognosis than pups that suffer severe symptoms from the outset.
The most definitive way to diagnose megaesophagus is via a barium-swallow X-ray study performed in a veterinarian's office. If possible, this should preferably occur under fluoroscopic guidance (which uses moving X-rays to visualize swallowing and esophageal motility, as opposed to static X-ray images, which typically use measurements of the diameter of the barium-filled esophagus for diagnosis). Fluoroscopic examinations are available at most veterinary teaching hospitals and some specialty clinics. In addition, survey chest/abdominal radiographs taken without barium may sometimes reveal megaesophagus, and can show evidence of aspiration pneumonia, foreign bodies, or other obstructions. Examination of the esophagus using a flexible scope, called esophagoscopy, is also diagnostic.

As congenital megaesophagus appears to demonstrate varying degrees of severity in Ridgebacks, symptoms will range along a continuum from barely affected to severely affected. A frightening situation arises at the “barely affected” end of the spectrum. A “barely affected” pup may not exhibit any of the obvious symptoms of megaesophagus, or only rarely regurgitate, yet **still be affected**. The situation may occur where the breeder, failing to recognize signs of the disease, or lacking personal experience with it, sells an affected pup as a show potential. Use of an affected dog in a breeding program then becomes a very real possibility. While the genetics of megaesophagus are still unclear, it should be apparent that using an animal affected with a genetic disease is a bad thing, especially if a test exists to diagnose the problem.

What does this mean for breeders? Do all litters need to be screened by barium swallow before placement to know for sure? Or should barium swallows only be performed on pups that show symptoms? Or should pups with symptoms be euthanized without a barium swallow? While situations vary, and no one answer is correct for every set of circumstances, breeders need to be aware that not all affected pups may show symptoms. It is not reasonable to expect breeders to screen all litters with barium swallows before placement. It is also not reasonable to euthanize a pup before a definitive diagnosis of megaesophagus is reached, as foreign bodies may also produce similar symptoms. However, when a breeder has reason to suspect that a pup may be affected, and that pup is confirmed through diagnostic testing, the ideal is for **all** pups in the litter to then be tested with barium swallows to identify normal, as well as potentially sub-clinical pups. While the barium swallow study itself is not entirely without risk (it can occasionally cause an aspiration pneumonia, even in a normal dog), it is important that affected, yet non-symptomatic (or barely symptomatic) pups be identified before they are inadvertently included in a breeding program. They can then be watched, and perhaps eventually placed as pets. In all cases, breeders should work closely with their veterinarian to make an informed decision.

The health of our breed depends on the actions of dedicated breeders to help eliminate genetic disease. Megaesophagus occurs in many Ridgeback lines, and it is important to take the necessary steps to reduce the occurrence of this heartbreaking disease. This includes using the testing available to expose it. If you encounter megaesophagus, or think a pup shows signs of it, please consider testing the **entire litter**. Don’t let an affected sub-clinical pup slip through the cracks.

### How You Can Help

Megaesophagus occurs in many breeds, including the Rhodesian Ridgeback. The prevalence is unclear, as it is presumed that many breeders simply euthanize affected puppies as soon as the condition is diagnosed, and so it goes unreported.

Before research into the mode of inheritance and possible genetic markers for megaesophagus is launched, more information about the disease needs to be known.

The RRCUS Health & Genetics Committee would like breeders who have megaesophagus in their litters to cooperate with confidential research. Robert Washabau, an internal medicine specialist at the University of Minnesota College of Veterinary Medicine, has agreed to examine a litter where megaesophagus is
present. This will not only allow him to describe how the disease manifests, but also to ascertain whether there are affected subclinical siblings -- that is, littermates who have a mild form of the disease that is not readily apparent without diagnostic testing.

Please help us to help our breed. If you have a litter with megaesophagus puppies -- or are planning one in which this might be a concern -- please contact one of the Health and Genetic Chairs: Denise Flaim at revodana@aol.com or Cynthia Roethel at houndscrest@earthlink.net. As with all research projects, all details are kept in the strictest confidence.