

Myasthenia Gravis and Megaesophagus

Two and 1/2 years ago, my Scottie was diagnosed with the primary disease Myasthenia Gravis and a secondary megaesophagus. At that time she was basically given a very small chance of survival. I would like to share what I have learned since that time, in hopes that someone else may benefit if they one day find that their dog has this terrible disease. It used to be thought of as a rare disease, but in recent years it has been discovered to possibly be the most widespread autoimmune disease diagnosed in the dog. Also, I feel strongly that this list should be aware that in a recent published study, Scottish Terriers have been named 3rd highest at risk for this disease.

In October of 1997, my Scottie began to vomit foam or spit, or so I thought. She continued to do so for 3 weeks although I took her to the vet on numerous occasions, trying to find out what was wrong. At first the vet thought that she had a throat infection, and he put her on an antibiotic. One week later, her symptoms hadn't abated so my vet recommended a stronger antibiotic. During the third week she was definitely worse and I rushed her into the vet. At this point, she wasn't able to keep anything down and it was obvious that she was very sick. I was scared to death, and I wasn't even sure she would survive. A few hours later, I received a call, and my vet had a diagnosis. He had performed a barium test and discovered that she had what is known as Megaesophagus. It is a condition where the esophagus has become enlarged.

She was no longer able to get very much food or water into her stomach because her esophagus had lost all motility. Whatever she ingested, would actually just sit in her esophagus, which now resembled a saggy bag until her gag reflex would bring it up. That is when I learned that she hadn't been vomiting at all. She had been regurgitating. There is a big difference and it is important to know the difference. Regurgitation occurs almost spontaneously; often the dog is unaware that it is going to happen. It isn't uncommon that the dog is taken completely by surprise. On the other hand, when a dog is going to vomit, they will usually salivate and heave; sometimes walking around looking for just the right spot to do so. Also, regurgitation doesn't contain the stomach enzymes and is usually lacking the yellow color of bile. Knowing the difference between regurgitation and vomit is key in diagnosing megaesophagus.

She also had pneumonia. During episodes of regurgitation, tiny food particles are often aspirated into the lungs and aspiration pneumonia is a constant threat. Pneumonia is the biggest threat in this disease. In a portion of a special monthly feature on Neuromuscular Cases published on the Internet by Dr. Diane Shelton, she states that: "Unfortunately, the mortality rate in canine acquired MG is still unacceptably high with approximately 50% of the dogs diagnosed with acquired MG succumbing to aspiration pneumonia or respiratory paralysis."

Once we had the diagnosis of Megaesophagus, I learned that it was almost always secondary to a primary disease. If we could discover and treat her primary disease, the secondary megaesophagus could possibly respond. To discover what her primary disease might be, I saw a board certified internist who had experience with megaesophagus. There are several diseases that can cause megaesophagus, but in recent years, Myasthenia Gravis (MG) has been discovered to be very high on the list as a cause of megaesophagus. It is now recommended that a veterinarian test for MG if a dog presented is regurgitating. A blood sample is drawn and sent to the University of

California in San Diego. There they perform a blood test to detect circulating antibodies specifically against acetylcholine receptors. A positive titer means that the dog has created antibodies. This is the only place in the country where they do this test. If the test is positive, or the titers are high, MG is diagnosed. If upon subsequent testing, the titers are reduced, that usually indicates remission. Remission may or may not last. New information found (again, on Dr. Diane Shelton's web page) indicates that many dogs go into a permanent remission. Some do not, as in the case of my Scottie.

MG has many variations. It is an autoimmune neuromuscular disease where the connection from the brain to the muscle has been lost due to a very complicated process that involves autoantibodies. The brain can no longer get the required message to the muscle; hence the muscle loses motility or strength. My Scottie has what is known as Acquired Focal Myasthenia Gravis with secondary megaesophagus. The only muscle involved is her esophagus. She has no generalized weakness in her limbs or other muscles, which can be very common in this disease.

Initially, after diagnosis, she was treated with a drug called Mestinon. ((Pyridostigmine Bromide). It is the treatment of choice, but occasionally if muscle strength doesn't improve, corticosteroid therapy is recommended. After a period of time on the Mestinon, I began to see a difference in the frequency of her regurgitation. She was still very ill with the pneumonia, and was treated with antibiotics for 3 weeks. She also seemed quite nauseous and I used Pepto Bismal and Tagamet as suggested by my vet, which seemed to help. Eventually, over time she completely stopped regurgitating.

Probably, the most important element of all in the treatment of MG with megaesophagus is elevating the food and water to let gravity carry the food or water to the dog's stomach. If this isn't done, the chances increase dramatically that the dog will regurgitate. Each time the dog regurgitates the chances of contracting aspiration pneumonia increase.

It has been found that frequent, smaller meals are also desirable. They seem to be able to manage small amounts of food, fed more often, over large meals fed once or twice a day. Many dogs have a more difficult time with water than they do with food. I soaked my dog's food in water until it was very soggy. That supplied most of the water she needed throughout the day. What works for one, may not work for another and it is possible that you may have to experiment with different foods and textures. I also found that by holding her up for at least 15 minutes after eating or drinking, she was less likely to regurgitate.

I would like to stress that taking care of a dog with this disease is very difficult. I lost many hours of sleep. I had to watch her carefully. Many times she would awake during the night and I would have to hold her upright to try to stop the regurgitation. She had to be fed upright and held up afterwards for 15 minutes 5 times a day. I was afraid to leave her alone and I honestly don't know what I would have done if I had to leave my home for an outside job. People have managed to do this though. I personally know a girl who holds a full time job and has successfully managed this disease in her Scottie. I was very relieved when my dog stopped regurgitating, but I was to soon find that our problems were far from being over.

Our first visit to the specialist, who as I mentioned was a board-certified internist, cost \$362.00. They performed a few tests, took radiographs, did a complete CBC and profile and drew blood to send to California. I had already spent approximately \$400.00 at my

own vet in the previous 3 weeks trying to obtain a diagnosis. Upon that first visit, I wasn't given very much hope for her survival. Understandably, veterinarians who deal with megaesophagus on a regular basis know that aspiration pneumonia is often fatal. It wasn't until two weeks later on a second visit that I knew we were dealing with MG and I was informed then that remission was a possibility. Surprisingly enough, often with remission, and early diagnosis, the esophagus can return to normal. My dog did go into remission, it took approximately 2 months, and a new radiograph showed a normal esophagus. Again, we had much more to face in the coming months.

My dog whelped a litter of four puppies 6 months previous to her showing symptoms of this disease. It turns out that Dr. Diane Shelton DVM, PhD at the University of California in San Diego is usually consulted on cases of Myasthenia Gravis. She is a leading expert in this field and has written many published studies on the subject. She told my doctor that new information indicated that pregnancy and hormone surges in a female dog might be a "trigger" for MG symptoms to occur. That and the fact that she was 3 years old, which is a classic age for symptoms to emerge. Now I was strongly advised to spay my dog to avoid a hormone surge with her upcoming heat. I did so, but unfortunately a week after she was spayed, she came out of remission, and everything started all over again. Stress may have been a factor in her relapse. There are many factors that can affect this disease. Once more, about 2 months later she went into remission again.

I was also advised to spay and neuter all 4 of the puppies. There are many references to hereditary factors in this disease and it makes sense anyway. My girl, the mother of those pups, has a diagnosed disease and I would not have been interested in reproducing that disease. Below is an abstract from a recent study that addresses that issue.

1: J Am Vet Med Assoc 1999 Oct 1;215(7):956-8, 946 Related Articles, Books Inherited predisposition to myasthenia gravis in Newfoundlands. Lipsitz D, Berry JL, Shelton GD Department of Medical Sciences, School of Veterinary Medicine, University of Wisconsin, Madison 53706, USA.

Acquired myasthenia gravis was diagnosed in 6 Newfoundlands from 2 distinct lineages Three dogs, including 1 pair of littermates, from each lineage were affected. History and clinical signs did not differ from that reported for other dogs with acquired myasthenia gravis. Although a genetic predisposition for development of certain autoimmune diseases in several species has been identified, the immunopathogenesis of myasthenia gravis is multifactorial and includes hormonal, environmental, and infectious disease factors. The high incidence of myasthenia gravis in these 2 distinct lineages of Newfoundlands, a breed with a low relative risk for development of this disease, suggests an underlying genetic mechanism. However, mode of inheritance could not be determined from this small number of dogs. Regardless, breeders should be alerted to this disorder, and they should consider removing Newfoundlands with acquired myasthenia gravis from breeding programs. PMID: 10511859, UI: 99441530

If a dog is diagnosed with MG, one is strongly advised to have the blood sent out and titers checked every three months. I drive my dog to the specialist two hours away, every three months and will continue to do so for the rest of her life. I have spent close to \$3000.00 on her since the symptoms began. Needless to say, I would never have bred my

girl if I had known what the future held. Even now, I will never know for sure if her disease would have become active had I not bred her. She seemed perfectly healthy beforehand and was certainly vet checked before she was bred. I would have never guessed that this disease might have been lurking. My puppies are just a few short months from their 3rd birthday and I am holding my breath, hoping they won't be affected. This disease is devastating, and I would be loath to see it become common in a breed that I absolutely cherish. Unfortunately, a recent study has shown that Scottish Terriers are listed 3rd at risk for having this disease. Below is a portion of that study:

Risk factors for acquired myasthenia gravis in dogs: 1,154 cases (1991-1995)
G. Diane Shelton, DVM, PhD; Alan Schule, BS; Philip H. Kass, DVM, PhD
In comparison with mixed breed dogs, dogs with the highest risk of acquired MG were Akitas, Terrier Group, Scottish Terriers, German Shorthaired Pointers, and Chihuahuas.

My dog was in remission this last time for 15 months. On October 18, 1999 she once again had a relapse. Although I am now experienced with this disease, she still managed to get pneumonia. That is the very dark side of this disease. Fortunately she recovered and is now in her 3rd remission. Three months before her remission ended, I had her vaccinated for Bordetella.

That may have been a factor this time in her relapse. There are many unknowns in this disease, but we are playing it safe and she'll never receive another vaccination again, by my veterinarian's recommendation. Just before Christmas she began to feel better and look brighter. On January 11th 2000, her latest radiograph showed a perfectly normal esophagus. I won't have her blood test results back for another week, but at this time she is doing very well. I will never be able to relax and take for granted that she will totally recover from this disease. Often I become a nervous wreck when I hear a little cough or gag. I may have to fight this battle for the rest of her life, and will do so if I have to. There has been a lot of research done on MG and it is very well understood. At the same time, there are many, many unanswered questions. My hope is for a breakthrough or a cure, soon. If you are interested in more information on this disease, or on other neuromuscular diseases, Dr. Shelton has a very informative web site. The address is:

http://medicine.ucsd.edu/vet_neuromuscular/case.html

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DEBBIE SMITH POSTS THIS AFTERTHOUGHT:

Please know that although 3 years of age is a high risk age, Megaesophagus can occur at any time, (including soon after birth although this is known as Congenital Megaesophagus, which is not an autoimmune form). My information is a small part of all that exists about this disease and although Myasthenia Gravis is very high on the list, there are many other reasons why a dog can end up with Megaesophagus.

I wrote my article about my personal experience with my dog. Many factors can and do vary. There are dogs that breeze right through this disease without any treatment whatsoever, except supportive feedings, and there are also those that do not stand a chance, no matter what a dedicated owner may spend. That is what I think is one of the scariest things about this disease.

You may be lucky, or you may not. My first vet, that I no longer see, told me flat out that he had 4 cases of Megaesophagus in his career and they all had to be euthanized. He himself had no hope. Unfortunately he most likely still has this attitude. There is more available information out there now, and if regurgitation is suspected, MG is a possibility that should be explored. Hypothyroidism, Hypoadrenalcorticism, Vascular ring anomaly, and many other diseases can cause Megaesophagus. There is even Idiopathic Megaesophagus where no primary disease can be discovered...

Source: <http://www.scottiemed.com>