

Commonly asked questions about dermatomyositis (DM or FCD) in dogs

1) What is dermatomyositis?

Dermatomyositis (DM) is a devastating inherited inflammatory disease of the skin and/or muscle which most commonly afflicts Collies, Shetland Sheepdogs and their crosses. The skin lesions consist of hair loss with or without skin redness, scaling and crusting of the face, ears, legs and tail tip. One or more of these areas of the body may be affected. In addition, some dogs may have muscular involvement. Sometime this muscle involvement is so pronounced that it results in muscle atrophy. Other cases may suffer from megaesophagus (enlarged food tube in the body) with the end result of aspiration pneumonia. In milder cases the dogs may appear to be sloppy eaters, or have a strange high stepping gait. Shetland Sheepdogs are fortunate because muscle involvement does not seem to be as common as with the Collie. Most commonly dogs develop the skin lesions within the first few years of life but dogs as old as 8 years have been reported to develop this problem. It is thought that certain triggering factors such as stress may cause a dog to express the DM. These triggering factors are thought to include such circumstances as a bad viral infection (e.g. parvo) and hormonal fluctuations (heat cycles). In some cases no triggering factor has been identified. Intact females appear to be more subject to hormone related stresses than intact males. Stress of travel, moving, family upsets may also make the symptoms worse. Since this is a genetic problem, affected dogs should be spayed or neutered. This will also help the dogs respond better to treatment.

DM is not an itchy problem unless a secondary infection is present. So if your pet is itchy and it does not look like a secondary infection is present (bacteria or yeast) then your pet's problem is probably not DM. The only exception would be if you had a dog with DM and an underlying allergy. This can occur but is not common.

2) Preferred treatment options for DM:

- Pentoxifylline - This continues to be the best treatment for DM symptoms. IT IS IMPORTANT TO ONLY USE THE BRAND NAME TRENTAL® if you decide to use pentoxifylline as your treatment option. When generics have been used they have been found to be either ineffective (not helped the animal with the DM) or associated with side effects of vomiting and diarrhoea. These side effects have only been seen when the drug is not given with food, or a generic brand of pentoxifylline is used. The dose used in the study is 25 to 30 milligrams per kilogram of body weight every twelve hours given by mouth and always with food. These tablets have been successfully split when used in treating the DM study dogs.
- Dapsone was also studied and could be used as an alternative in those rare dogs who do not tolerate Trental. The Dapsone dose in the study was 1 milligram per kilogram of body weight every eight hours, given by mouth. However, several dogs did well after being on the Dapsone for a month or two at 1 milligram per kilogram of body weight every twelve hours, by mouth.

Other treatments with less successful results include:

- Steroids - Side effects include liver problems, suppression of the adrenal gland function, and secondary infections due to the immunosuppression.
- Azathioprine ("Imuran") - Azathioprine is the most common type of immunosuppressive drug that has been reported to be used to treat DM in dogs. The use of this drug in dogs has been associated with several different side effects. The main side effect that azathioprine causes is bone marrow suppression (decreased red blood cell count, decreased white blood cell count, decreased platelet count). Other side effects of azathioprine in dogs include vomiting, diarrhoea (most common side effect, may have blood in stool), hypersensitivity reactions (especially the liver), an inflamed pancreas, skin rashes and hair loss. These dogs need to be closely monitored by a vet and periodic bloodwork performed on them (initially every 2 weeks) to make sure that the dog is not having a problem with the azathioprine.
- Immunostimulant drugs- Immunoregulin is a product of a kind of killed bacteria (*Propionibacterium acnes*) which is injected in the vein during set periods of time (initially twice weekly, then weekly then monthly). Side effects occasionally occur after the injection and include lethargy, increased body temperature, chills and decreased appetite. Anaphylactic shock reactions have also been

reported. If the drug is given outside of the vein then local tissue inflammation (swelling) has been reported. long term toxicity studies have demonstrated inflammation of the liver (hepatitis), vomiting, diarrhoea, decreased appetite, malaise (feeling poorly), fever, increased water consumption and acidosis.

- Antioxidants - The most common antioxidant which has been used to treat DM is Vitamin E. Some have used it topically but most vets use it systemically (give a tablet or capsule by mouth). If used at the dose reported in the literature then no side effects have been reported. This treatment has not been evaluated scientifically in a large number of dogs. Contact your vet for recommendations on the appropriate dose of Vitamin E to use on your dog.

3) How long will my DM dog need to be treated?

This is a grey area. We do not know for sure. Some dogs need 3 to 6 months of treatment and do fine whereas other dogs need life long therapy. The length of treatment varies according to the individual dog and the severity of the disease.

4) How is DM genetically expressed or passed on?

We do not know but we have seen this problem in certain breeding lines so we know that it is an inherited problem. The exact mode of inheritance is not known. The current status of DM research in the collie and the Shetland sheepdog is as follows:

- Linkage analyses in the Shetland sheepdog suggest possible linkage to canine chromosome 35.
- Gene expression profiles were generated for affected and normal skins using a canine-specific oligonucleotide array having 49,929 probe sets. Two-hundred and eight-five genes, many of which are involved in immune function, were found to be differentially regulated in these tissues. Real-time quantitative pcr was carried out for three genes differentially regulated on the array and confirmed trends observed on the array.
- Immunological studies show that staining patterns for normal and affected samples are quite similar, suggesting that canine DM is a disease that may be immune-mediated but does not likely include the production of disease-specific autoantibodies.
- Linkage analyses in the collie are underway.
- Keratinocytes are currently being cultured from affected and normal dogs in an effort to better understand the disease at the cellular level.
- A new tool, the canine snp array, has recently become available. DNA from affected and normal collies and Shetland sheepdogs will be used to probe the snp array in hopes of identifying linkage with the gene causative for DM

5) Does my dog have dermatomyositis?

This is the information that your vet needs to have in order to be able to answer this question.

- If you have a Shetland Sheepdog, Collie or their cross with any of the clinical signs described in the first paragraph then you should have your dog biopsied.
- Affected areas of the skin and/or muscle are the locations where the biopsy should be taken.
- A minimum of 3 biopsies should be taken.
- The diameter of each biopsy should be at least 6 mm in size. If you choose to use a biopsy punch to take your sample then you will need to use scissors or a scalpel to collect some of the muscle below your biopsy site. Remember, the muscle may be involved with this disease so it is very important to obtain muscle tissue along with your skin sample.
- Each biopsy sample should be placed in an individual container with 10% neutral buffered formalin. The owner's name, pet's name and date should be written on each container with a permanent marker.
- Biopsy samples should be submitted to:

The Dermatopathology Specialty Service
ATTN: Dr. Joanne Mansell
PO Drawer 3040

College Station, TX 77841-3040
Phone 979-845-3413 or 888-646-5423

A special submission form may be obtained by contacting Sherry Lindsey RN BSN (830-620-6661), Shalainetx@aol.com).

6) What other samples are needed to study DM? Blood samples (preferred) or cheek swabs for DNA material and 5-7 generation pedigree information are needed from DM dogs.

7) What kind of blood samples are needed?

- Whole blood in EDTA (2.5 ml minimum)- collected in a lavender top vacutainer tube and kept cold but not frozen. The owner's name, pet's name, DM status and collection date should be clearly labeled on the tube using a permanent marker. Blood samples should be sent to:

Dr. Leigh Anne Clark
Clemson University
51 New Cherry Road
319 BRC building
Clemson, SC 29634
phone: 864-656-4696

- Blood samples should be sent via overnight shipping with cold packs included. It is best to ship blood samples at the beginning of the week. Shipping costs are the responsibility of the shipper. Dr. Clark will isolate the DNA and store it for use in future studies.

8) What type of pedigree information is needed? Even if your dog does not have DM, a 5-7 generation pedigree would provide valuable information for our DM research. We need to know which dogs are normal and which ones are abnormal in a pedigree so we can determine the possible mode of inheritance for DM.

9) What is the charge? We will perform the DNA and pedigree analysis at no charge. We will charge the usual rate for the interpretation of the biopsy sample which is \$50.

8) Questions?

Sherry Lindsey RN BSN is a longtime sheltie breeder who assisted Dr. Rees in all of the DM studies and housed and cared for the DM study dogs. She is happy to answer any questions concerning DM and may be reached at shalainetx@aol.com. A website with DM information, including photos of DM affected dogs, may be found at www.shalaine.com/DM/DM.html

Clinical signs of FCD and some cases before and after treatment



