

Inherited Respiratory Disease in Wolfhounds Identified as Primary Ciliary Dyskinesia

by Margret L. Casal, DVM, MS, PhD

Wolfhound rhinitis is a disease that mainly Wolfhound rhinitis was thought to be caused quently since then. However, we have recently become aware of an increased number of cases affected dogs, and waited until they were clinicollaborative effort between Dr. Iris Reichler at the University of Zürich in Switzerland and ourselves at the University of Pennsylvania, we have begun collecting data and blood samples from affected dogs, their healthy littermates, and their parents.

We have strong evidence that wolfhound rhinitis is due to a malfunction of the cilia, which are microscopically small hair-like structures that line the airway passages (nose, sinuses, trachea, and lungs) and beat in an organized manner. The cilia are responsible for the transport of particles such as dust, dirt, and microbes out of the lungs. If the cilia are absent or do not beat fected Irish Wolfhounds in Europe also had in a synchronized fashion, the particles become hydrocephalus. Hydrocephalus is the abnormal trapped in the airways. The result is chronic respiratory infections leading to bronchitis and pneumonia. The common clinical signs are sniffling, sneezing, clear to greenish nasal discharge (or rhinitis, hence the name 'wolfhound rhinitis'), and coughing. It is important to realize that rhinitis is just a symptom, and is not the cause of disease. Affected dogs may develop fevers when the infections get worse. Dogs may already be showing signs at birth (runny noses) or symptoms may not appear until later (six months to one year). In general, dogs and humans with this condition can live quite long if they are maintained on antibiotics. Their lungs need to be cared for by frequent inhalation of steam and coupage (patting the affected dog on the side of the chest for several minutes on each side) which help loosen any mucous that is stuck in the deeper airways.

For many years there has been a debate whether this is truly a disease of malfunction-Wolfhound rhinitis. The main reason why example.

affects the respiratory tract, i.e. the nasal pas- by an immune defect is because, in the past, the sages and the lungs. In the Irish Wolfhound, immune systems of affected dogs were always this syndrome was first described in the 1960's examined during phases of severe illness; the but has been mentioned in the literature infre- immune system is never normal in very sick or dying individuals. For our studies, we treated the both in the United States and in Europe. In a cally healthy to examine their immune systems. After an exhaustive examination and comparison of the immune systems of affected dogs and their normal littermates, we found no differences. Our results suggest that Wolfhound rhinitis is not caused by an immune system defect. We have also looked at the cilia of affected dogs under the microscope and have determined that the cilia do not move correctly. Further examination under the electron microscope revealed abnormal structure of the cilia. These findings suggest that Wolfhound rhinitis is caused by abnormal ciliary function.

> Interestingly, a small proportion of the afbuildup of cerebrospinal fluid in the ventricles (cavities) of the brain. The form of hydrocephalus seen in the Irish Wolfhound does not cause any clinical signs and does not impair the dog's quality of life. However, there are forms of PCD in humans that are also associated with hydrocephalus (the cilia are also responsible for moving the cerebrospinal fluid in the brain). This observation strengthens the hypothesis that we are truly dealing with PCD in the Irish Wolfhound and not, as previously suggested, an immune disease.

For the breeder, it is most important to know the mode of inheritance in order to avoid producing affected puppies. Careful analysis of the pedigrees from Irish Wolfhounds with PCD indicates that it is an autosomal recessive disease. This means that an affected dog must have two defective genes in order to show signs of disease, both male and females can be equally affected, and the parents can look completely normal. ing cilia, or if a defective immune system causes The figure shows the mode of inheritance as an

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Hydrotherapy Helps Heal

by Kathy Wilson

ydrotherapy is making a big splash in the new and growing field of canine sports medicine and rehabilitation. Water workouts are not just for poodles and water spaniels any more. Riding the wave of popularity that human sports medicine and rehab programs have enjoyed, swim centers and specialized sports medicine centers for canines are beginning to surface all over the country. Physical therapy is now accepted as an integral part of the recovery process following injury or surgery. Remember, not that long ago, days or weeks of rest, rather than rehab, was the prescription following orthopedic or neurologic surgery. The mindset has changed as knowledge of how the body recovers from trauma grows by leaps and bounds. Many cutting-edge veterinary specialty practices and veterinary teaching hospitals have added swimming in hydrotherapy pools and work on underwater treadmills to their arsenal of more conventional treatment regimens. Water workouts can provide more improvement in less time for wolfhounds recovering from trauma or surgery.

he combination of buoyancy and resistance which water provides offers more support for weakened joints and limbs. This support reduces the likelihood of injury, while allowing the dog to exercise with less pain. Dogs with neurological disease and severe weakness, who have been unable to achieve weight-bearing on land, can often begin to use their limbs. The water's support and the reduction in weight-bearing is a powerful combination, allowing them to progress more quickly toward normal function.

nderwater treadmills specifically designed for use in canine physical therapy allow the therapist to adjust the speed and angle at which the dog exercises, as well as the water level. Different levels of water change the weight-bearing balance between the front and rear legs. On land, distribution of weight is approximately 60%/40%, with most of the weight supported by the front limbs. By carefully choosing the water level to suit each patient, the physical therapist can alter this weight distribution to suit the limb or joint needing rehab. Although hydrotherapy is *not* a magic bullet, it can be a significant component in an overall treatment regimen designed for your wolfhound. Of course, the canine underwater

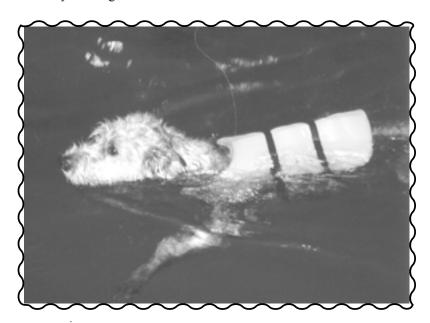
treadmill is expensive—so expensive that few facilities have acquired them. Finding a facility which utilizes the treadmill is much easier if you live near a major urban center.

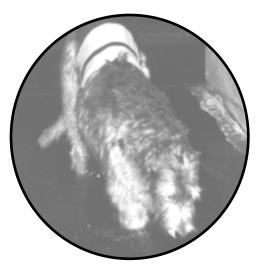
hen vou locate a canine sports medicine center, you'll want to check out more than the equipment. The University of Tennessee offers a certificate in Canine Rehabilitation for veterinary personnel (including veterinarians and veterinary technicians) and physical therapists. Auburn University's College

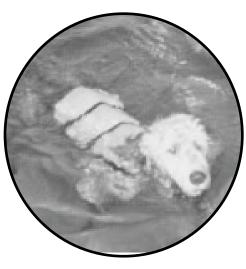
of Veterinary Medicine has a Veterinary Sports Medicine program devoted to canine and equine athletes and working animals. In addition, this specialty has been recognized by the American Physical Therapy Association as a special interest group under their Orthopedic Section. Within the next decade, it may become a field with standards for accreditation and formal training requirements. In the meantime, a referral from your veterinarian or veterinary specialist may be the best guarantee vou can hope for to ensure that

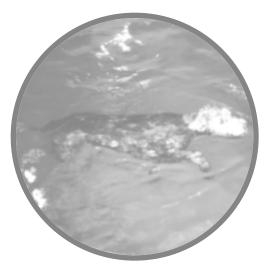
you've chosen a reputable facility for your dog's rehabilitation or exercise program. Some of the larger facilities devoted to canine rehabilitation and sports medicine complementary therapies such as acupuncture, chiropractic, and massage in addition to hydrotherapy.

hen used as part of a comprehensive program designed to address the specific needs of each individual wolfhound, aquatic therapy is more than just treading water. It could be a stroke of genius.









plash! Merlin Makes Waves

by Karen Malone

Merlin, like many Irish Wolfhounds, was not getting adequate exercise. As I'm not a runner or a biker, and I don't have any other dogs who will help exercise him by running and playing with him, I decided to take him for hydrotherapy to keep his body well-muscled,.

Now, as we all know, Irish Wolfhounds are not by nature water dogs. The swim center that I took him to was Interstate Equine and Canine Swim Center, which had over ten years of experience in hydrotherapy, but no experience with wolfhounds. At first, it was a bit of a challenge, but we made gradual progress. Merlin started out swimming three times a week for 6 minute intervals during a 20 minute session in his attire of a life vest and a tether held by a swim center staff member. His swimming time was gradually increased up to 20 minutes while still wearing the life vest. Eventually, he graduated to the next level—swimming without the life vest, which he now does with excellent results. I found that not only was Merlin exercised, but I got some exercise, too, as I walked around the perimeter of the pool to encourage him to swim to me!

Did this exercise program work? Swimming enabled Merlin to continue to develop physically into the handsome devil he is today! Merlin went to the National that May, and, guess what? He won Best Puppy! I guess his hard work paid off and we are all very proud of him.

Merlin continues to swim twice a week, just to stay fit and well-muscled. He now swims for 20 minutes at a time without the life vest. I'm looking for a tee shirt that says "Swim Mom" to put Merlin's picture on, so I can show him off everywhere we go!

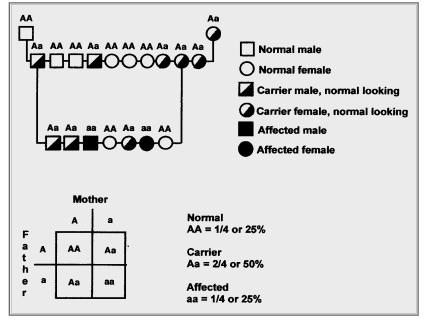
I think both the swim center and I have learned a few things from this experience that are worth passing on.

I would recommend that you immediately put a leash on each dog after he finishes his swim, even before the life vest is removed. Imagine my horror when, after Merlin's first swim session, as the center's owner and I were chatting at the side of the pool, Merlin proceeded to walk off the edge of the pool! All I could say was "Oh, my god!" as the owner of the center dove into the water (cell phone and all) and rescued Merlin! Merlin was none the worse for wear—he had not swallowed any water, nor was he panicked (unlike his owner!)— but it gave us quite a scare. And I don't think that cell phone responded to CPR.....

☐ Surprise! Wolfhounds don't fit in shower stalls designed for most dogs. The center installed a bigger shower stall to accommodate IW bathing. ☐ Even IW's can swim. As you can see from the photos, Merlin is a casual, relaxed swimmer—or dog paddler—exceptat the end of his swim, when the timer goes off.

☐ Swimming an IW is a great way to keep a wolfhound who doesn't get much routine exercise healthy and well-muscled. Merlin's brother, Logan, who lives nearby in Maryland, has taken the plunge, too. Logan started swimming shortly after Merlin began, and continues to swim regularly. The result? Both boys are staying well-muscled and fit. I recommend swimming to anyone who needs more exercise — dog or human!

This drawing shows a situation tions if both parents were "carriers". where an accidental brother-sister In autosomal recessive traits, such as mating occurred. All genes are PCD, carriers (Aa) "carry" the defecpresent in pairs in each individual, tive gene (a), but they also have a except for those genes on the X and normal one (A), which allows them to Y chromosome. Each parent passes be healthy. The checkerboard shows one of its two copies of a gene to that there is – statistically – a 25% an offspring and there is basically chance of having completely normal a 50:50 chance, which of the genes offspring, a 50% chance of having from each parent gets passed on. In carriers, and a 25% chance of having the example above, we will look at affected dogs. The top of the pedigree only one particular gene – let's say the above shows the first mating between PCD gene. "A" stands for the normal two unrelated parents, one being a copy of the gene and "a" stands for carrier and the other one normal. the defective or mutated copy of the Remember that both of these parents PCD gene. The checkerboard-square look normal and do not show signs demonstrates the possible combina- of disease (i.e. there was no way of



knowing the mother was a carrier). If you were to do the "checkerboard" math again, you would see that a mating between AA and Aa would result in 50% AA and 50% As offspring. The second breeding in this pedigree was a carrier-brother X carrier-sister mating resulting in normal, carrier, and affected puppies. While the affected pups will be easy to recognize, the carrier and normal dogs cannot be distinguished just by looking at them. This is demonstrated in the drawing by the open symbols and the AA or Aa above each symbol. It is important to remember that when assessing risk percentages that the influence of ancestors is cumulative.

Currently, there is no reasonable way to distinguish carriers from normal dogs, as they look the same. To eliminate PCD from the breeding population, a DNA-based test would be extremely valuable. At this time, the only way to know if potential parents are a carriers, is by having affected pups born (both parents are automatically carriers) or if a test mating was done by breeding the parent in question to a known carrier. If more than 11 normal pups are born, then the prospective parent is most likely not a carrier. However, this is a very crude method of "genetic testing." Currently, we are trying to find the gene for PCD and subsequently develop a DNA-based test for the disease. To perform these investigations, we are requesting blood samples from normal and affected dogs to extract DNA. Five to ten ml of EDTA blood (purple top tube) should be sufficient. The blood may be sent on ice packs or at room temperature, but not frozen. Please include basic pedigree information (parents and possibly grandparents) and remit the signed consent form (you will receive a copy). The samples should be sent to the address below. For risk analysis, please contact Anne Janis (iwstudy@earthlink.net). All information is handled confidentially. Thank you very much for your participation in this important study that will benefit all Irish Wolfhounds!

> Dr.. Margret L. Casal Section of Medical Genetics VHUP Room 4015 3900 Delancey Street Philadelphia, PA 19104-6010 casalml@mail.vet.upenn.edu

The Irish Wolfhound Foundation Launches Membership Campaign

As the Irish Wolfhound Foundation enters its third year, the need for a consistent, reliable, and predictable revenue source has become more and more apparent. Our educational programs have brought the best and brightest researchers and practitioners to speak at national and regional IW specialties. Our fledgling Foundation is sponsoring five major IW health studies and provides free annual health screening opportunities for hundreds of wolfhounds. Our nationally-recognized newsletter is our means of letting you know about the latest research. These programs cost money—lots of it—over \$40,000 in 2003 alone. This fiscal reality dictates the necessity for a membership program.

Until now, these expenses have been offset by generous donations from IW fanciers worldwide, a large bequest from the late Phillippa Crowe, and profits from the IWF Gift Shop. In the early days of the Foundation, it was important to get the word out about its existence and the important work it was undertaking. To help create this awareness, *Focus* has been mailed out to as wide an audience as possible. Now, as we mature as an organization, we must look for a more fiscally responsible way of providing this important information to those IW owners and fanciers who truly believe in, and wish to support, our ongoing work.

Beginning in January, 2005, annual membership in the Irish Wolfhound Foundation will cost \$20 per year. Membership includes a subscription to *Focus* and a 10% discount for the member on all items purchased in the Foundation Gift Shop. The greatest benefit, however, will be the satisfaction you feel from contributing to the health and welfare of our beloved breed through the work of the Foundation.

Membership forms and details will be provided in the next issue of the newsletter.

Reproduction To Be Topic at IWADV Specialty Oct. 3rd

Plan to join us at the IWADV Welcome Reception on Sunday evening, October 3, where veterinarian Dr. Robert S. Dove will discuss various aspects of reproduction important to IW breeders. Dr. Dove, a Scottish Deerhound breeder for over 20 years, specializes in reproduction. He will share all the latest information and insights he's gleaned from atteding the most recent conferences of the European Veterinary Society for Small Animal Reproduction, the European College of Veterinary Reproduction, and the American College of Theriogenology/Society for Theriogenology annual meeting, where he was a featured speaker.

Dr. Dove's presentation is jointly sponsored by the IW Foundation and IWADV. This is a 'must attend' for all breeders!

RESULTS OF THE 2004 IRISH WOLFHOUND CERF CLINIC

IWCA Specialty, Lima, Ohio May 21, 2004

by Anne Gemensky-Metzler, DVM, MS, DACVO

At this year's specialty, a very small number of Irish Wolfhounds were examined (21). Nine dogs were normal. The most common abnormality was iris or uveal cysts (5 dogs affected). Iris cysts appear to be common in the IW and are most often an incidental finding. However, in Golden Retrievers and Great Danes, iris cysts can be associated with inflammation and glaucoma. Therefore, the cysts should be monitored periodically for increases in size and/or number and intraocular pressures should be

Retinal folds were diagnosed in two dogs. Retinal dysplasia refers to abnormal development of the retina, often occurring in both eyes, resulting in a clinically apparent area of retinal folding or thinning or, in severe cases, retinal detachment. Retinal folds and geographic retinal dysplasia typically are not significant to vision and do not progress. Breeding dogs with retinal dysplasia is discouraged because the trait can occur in offspring in a more severe form (e.g., a dog with folds could have offspring with a large area of geographic dysplasia or, rarely, retinal detachment). However, in most breeds, including the IW, folds are "breeder option." The hereditary nature of retinal dysplasia has not been investigated in the IW, and the CERF genetics committee will monitor the condition in the breed and make appropriate breeding recommendations. The best recommendation for now would be to have a CERF examination annually and, if the dog is to be bred, it should be bred to a dog with a normal CERF exam and should be eliminated from the breeding program if retinal dysplasia is detected in the offspring. Litters should be examined between five to ten weeks of age to screen for retinal dysplasia, congenital cataracts, optic nerve hypoplasia, and other potentially heritable ocular disorders.

Lens abnormalities were diagnosed in 5 dogs. In 4 of these, the lens lesions were not considered hereditary and were marked in the comments box or were marked "significance unknown," meaning that they would still get a CERF number. One dog had focal posterior cortical cataracts that were typical of hereditary cataracts.

One dog each was affected with vitreous degeneration, persistent pupillary membranes (PPMs), and scrolled cartilage. Degeneration of the vitreous humor can be a normal aging change, but it is being monitored in some breeds that show early onset. PPMs are not problematic in IW's as long as they are the "iris to iris" type. Scrolled cartilage refers to kinking of the vertical cartilage of the third eyelid, resulting in cosmetic deformity and, sometimes, exposure of the gland of the third eyelid and inflammation of the cornea. This defect requires surgical correction when severe.

In the Canine Eye Registry Foundation (CERF) Eye Disease Report 1991-1999, only 386 IW were examined. Of these, 285 (74%) were normal, 15 (3.86%) were affected with micropapilla or optic nerve hypoplasia and the percentages of dogs affected with the other abnormalities were similar to those seen at this clinic. It is important to continue to examine Irish Wolfhounds prior to breeding to continue to screen for potentially hereditary ophthalmic conditions.

HOW CERF WORKS

The Canine Eye Registration Foundation collects data on *all dogs* examined by ACVO diplomates, regardless of the outcome of the eye exam. If the dog is found to be free of heritable eye disease, he may receive a CERF number and be registered in the CERF Registry, where this data can be easily accessed at the CERF website. Their database is searchable by the dog's name, AKC registration number, or CERF number...

The ophthalmologist files the research copy of the CERF form for every dog examined, whether or not the dog has any disease. Information on dogs who are found to have heritable eye disease is strictly confidential and the individual dog's identity will never be revealed. However, this information will be entered into the database for each breed.



2004 IW CERF CLINIC Total IWs Tested 21 Normal IWs 9 Abnormal IWs 12 Iris Cysts 5 2 Retinal Folds Lens Abnormalities 5 Vitreous Degeneration 1 PPM 1 Scrolled Cartilage

The Hounds Thank You!

For years, Marcella Gran and Sheila dous debt of gratitude for the thou-Miller have devoted their spare moments to raising money for Irish Wolfhound charitable causes. By any measure, they have been wildly successful. They've refined the art of offering for sale the perfect accessories and necessities for the well-healed wolfhound and his owner. While stocking the shelves of the Irish Wolfhound Foundation Gift Shop (formerly the IWCA Boutique) with the best books and finest clothing for the discerning IW aficionado, they've exhibited time and time again an uncanny ability to select just the right "objets de wolfhound" to adorn our homes and our hounds. These two __ tireless and tasteful women have raised thousands and thoufor Irish Wolfhound sands of dollars Rescue, Research, and Education over the past decade. shopkeepers Marcella Gran and Sheila Miller have handed over the keys to the

Irish Wolfhound Foundation Gift Shop to Carolyn Dean, who will now set about the daunting task of carrying on this tradition.

Beyond the years of enjoyment that they have given us with delightful IW merchandise, we owe Marcella and Sheila a tremen-

sands of dollars their efforts have brought us. The IWCA Rescue, Research, and Education funds were established from the monies raised through the old IWCA Boutique and Auction. The yearly income we came to expect enabled IW Research to establish a significant long-term research program, which continues to this day under the aegis of the Irish Wolfhound Foundation. We still rely on proceeds from the Gift Shop to help fund our breed-specific research and to make our findings available to IW caretakers and their veterinarians through this newsletter.

Of course, we'll miss that special "je ne sais quoi" which Sheila and Marcella brought to the IW Foundation Gift Shop, but we look forward to Carolyn Dean's new approach.

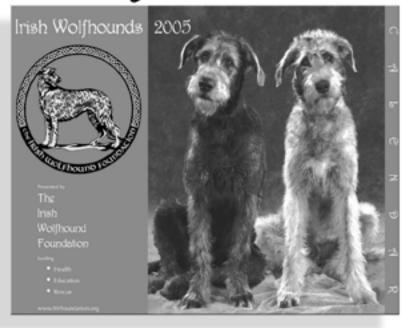
Sheila and Marcella, the hounds and their humans thank you more than mere words—or woofs—can



Marcella Gran E Sheila Miller

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 - U.S. Regional and Local Clubs
 - National Clubs world wide
- 12 Monthly Photos, plus many more

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Calendars will be available for purchase at matches this fall or online November 1st -www.IWFoundation.org-

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In memory of Phillippa Crowe

In memory of Sam Ewing & Phillippa Crowe

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My wonderful hounds

In memory of Rhonda Jane In memory of Miles, a very special hound

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Downloadable entry form at www.IWFoundation.org

Box lunches available each day at \$9 per person; Saturday evening Italian buffet dinner, \$20 per person.

Information packet and meal reservation details sent upon receipt of entry.

For more information, contact Marypat Corbett at 636 .273.6123 glendaloo@juno.com

Kathy Wilson 540.592.3705 glendorling@earthlink.net

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