

Mrs Gill Key

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3<sup>rd</sup> February 2010

**F.A.O. Committee, Wirehaired Dachshund Club**

**Re: Lafora Disease in Miniature Wire Hair Dachshunds**

I am the owner of a 10 year old dog, Alfie, who is affected by Lafora Disease. You may have read my article in the Dachshund Club newsletter about the impact of the condition on the dog and the owners (appendix i). Since I wrote that piece, Alfie has had more fits and his poor eyesight and his dementia continues to get worse.

Mainly as a result of the response to my postings on [www.dachshundforum.co.uk](http://www.dachshundforum.co.uk) and more recently the publication of the article, I have been contacted by other owners of affected dogs, some of whom were extremely emotional about the whole business, all looking for help and advice. Dogs displayed a variety of symptoms, but many of them are at a similar level of intensity to those experienced by Alfie. Some individuals were also keen to raise the issue with various media organizations with a view to forcing a requirement for genetic testing before breeding.

I decided to set up a group (Lafora Dogs Support) with the key intention that this should be a positive force for those owners who need answers themselves, and are keen that other people and dogs will not suffer in the future. (I have summarized the main aims and how they are supported on the attached appendix ii.) With the minimum of publicity, we already have 15 members. Several members have more than one dog, some are concerned that they may have dogs that will be affected in the future and some no longer have any live affected dogs. I have begun to compile a list of owners who would be keen to get their dogs tested as soon as possible and again, with the minimum of publicity, I already have expressions of interest from owners of more than 30 dogs who would be keen to get their dogs tested, subject to this being affordable, and accessible.

The purpose of this letter is to:

- a) express my thanks for all the work so far to investigate the practicality of setting up a testing programme
- b) offer my support in any way I can as the instigator of the Lafora Dogs Support Group
- c) provide feedback on reactions from Lafora Dogs Support Group members and others
- d) and finally request, on behalf of the members of the Lafora Dogs Support Group, that a decision on the way forward for both tests is made in the very near future.

I have a broad understanding of the genetics behind inherited autosomal recessive conditions and I appreciate the importance of identifying affected, clear and carrier dogs to provide the most effective future breeding programme. I am aware of the two options available from Canada for affected/unaffected and clear/carrier tests and the requirement for the latter to be performed on jugular bloods rather than saliva samples. I understand that you are proposing therefore to offer testing at various Dachshund Shows, (the first towards the end of March?)

I would like to make several observations based on conversations with Lafora Dogs Support Group Members and ask for them to be considered as part of your discussions on Lafora testing at your February meeting.

- a) Not all of those who would like to get their dogs tested are either able or will find it convenient to visit a Dog Show (those who have expressed an interest live right across the UK, including one in Jersey)
- b) Many are keen to get moving with testing their dogs for affected/unaffected as soon as possible, rather than waiting for the setting up of the carrier/clear test, which may still take some time to organize/fund.
- c) Some owners are conscious their dog is due to come into season and would like to take the responsible action of checking whether their breeding pairs are affected before the dams come into season, in which case they will not breed until the carrier/clear test is available. (They do appreciate that an 'unaffected' dog may well still be a carrier but there will be a reduced probability of producing affected puppies).
- d) I understand that the 'saliva testing kits' provided by the Canadian Team are relatively easy to administer by an amateur. I have heard directly from them that there is no problem collecting the samples separately and sending them individually. An alternative option is to ask that all testers test their dogs on the same date and forward them to a central point from where they can be logged and then shipped in bulk by FEDEX or alternative. I believe that blood is still fine for testing up to a week from the time it is first drawn.
- e) Suggested 'reasonable costs' for the tests seem to vary between £25 and £50
- f) Where dogs are identified as 'affected', could consideration be made to put in place an arrangement for a central organization (Wire Club, Kennel Club or other) to write to the owner of their dam and sire to advise that they also be tested and consider avoiding breeding/breed responsibly with clear dogs only?
- g) Without exception, the members of the Lafora Dogs Support Group would not consider purchasing another Mini Wire Hair Dog unless the sire and dam had been tested, and subject to the outcome of those tests, the puppy concerned tested as well. As awareness of the condition inevitably grows, this requirement is likely to become more common, and as a result, those breeders brave enough to have their dogs tested may well command a premium price.
- h) A proposition has been put to me that any dogs tested should also be microchipped, chip checked or tattooed so that it is possible to guarantee that the correct dog is actually offered for testing, given that there may be a substantial financial benefit in recording a valuable champion as 'clear'. I have also had offers to microchip at the same time as the bulk testing at shows for the cost of £7.50 per dog. Because the carrier/clear test is new, it might be worth considering introducing such a requirement right at the beginning of the testing programme rather than retrospectively, after some dogs have already undergone the test. On the other hand, permanent identification by chipping or tattooing of all stock is something the KC may insist on eventually anyway.

I do hope that you find this letter useful. It is entirely my purpose to support your work and the continuing existence of this wonderful breed, healthy and happy into old age.

Yours sincerely

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## Appendix i:

### Alfie's Story – Living with Lafora's

I was always keen to follow in the family tradition of having a smooth hair dachshund of my own when I grew up, and in 1999, married and with my own family, and working from home, I was finally in a position to make the family complete with our own small addition, but my husband wasn't quite convinced .....

Until, on holiday in South Devon, he, like the rest of us, fell in love. Out walking in the country around Salcombe, we came across a whole gaggle of Mini Wires. We were completely bowled over by their feistiness, cheerfulness, and most importantly friendliness – my childhood dogs had always been wary of strangers and it was appealing to have our overtures met by such a warm response.

We were hooked. I was all too aware of the back problems as it was this that caused the premature death of my parent's first dog, Pippa, so I checked the internet and spoke to various breeders into other potential problems specific to MWHD and was delighted to discover that the breed had no other reported serious health concerns.

After various abortive forays, we found exactly the dog we wanted – we met father and mother-to-be, both delightful, at home in Taunton. It was made clear to us that his sire was too big for showing, and there was a very good chance that the pups would be the same, but we didn't care..... all we wanted was a healthy dog from a family background with the sire's great temperament. As a precaution, we did check out his pedigree and were delighted and reassured to find several champion dogs including several from what, after my research, were familiar kennel names of top breeders.



Alfie was everything we'd hoped and more: intelligent, willing to try anything, including climbing to the top of rocky outcrops in Somerset (see picture) or making it to the top of Great Gable Mountain in the Lake District; ready to make up his own games ..... then persuading us to join in, such as bodyboard surfing 'chase the ball' in South Devon, (see: <http://www.youtube.com/watch?v=DCYPU0eDOwM>); on the touchline in his unofficial capacity as mascot of the mini Rugby side ..... yet just as happy curled up for a cuddle on our laps.

He had his major health problems – two back ops to sort out a total of 9 calcified discs, but he bounced back stronger each time; skin allergies needing frequent treatment with anti fungal shampoos; a hernia operation, but each time he came back good as new.

Until one day, when he was 6, we were out walking with friends in woods near Tarn Hows when suddenly, with no prior warning, Alf keeled over and had a full blown grand mal fit, foaming at the mouth, twisting and twitching. When he came round, he didn't recognize us and hid in terror behind a tree ..... until, like a switch being turned on he came back to us, greeting us with such relief to be back from whatever horrible place he had been.

Over the months to follow, every few weeks it happened again, and we began to recognize the triggers and the signs of an imminent fit – he'd never taken any notice of the TV before, but now, when the Rugby was on – he'd watch the screen transfixed until he fitted again. Out walking in woods with sun streaming through the leaves or playing an exciting game of hunt the ball in a cardboard box, he'd suddenly keel over as if polaxed. Our vet advised us to avoid putting him on a drug routine if possible and we learnt to avoid the triggers – he was banned from being in the room when we were watching the Rugby World Cup; we only walked in woods in dull weather and we invented new, less stimulating games.

So most of the time, we were managing to keep the grand mal seizures under control, but another distressing reaction was starting to happen. Every so often, if someone passed close by him; if we threw a ball in his direction; if there was a sudden noise; we'd see him suddenly jerk back as if badly shocked. It was distressing to see. He also seemed to stumble more when walking, and he began to have difficulty catching a ball.

The vet hadn't seen anything like it before but put it down to another manifestation of his epilepsy. I wanted to find out if there was anything we might be able to do to help lessen Alf's symptoms, so started to search on the internet for any articles about epilepsy in dogs – nothing useful until I changed the search and added in the words 'wire hair dachshund' – and there it was...

A vet in England (Clare Rusbridge) and another in Canada had identified a genetic condition that affected 5% of MWHD. The gene was similar to one found in human teenagers who were affected by a form of epilepsy called Laforas which ultimately would prove fatal. The article said Lafora wasn't fatal in dogs, but from its first appearance, typically when the dog was around 6-7 would "progress over many years and gradually other symptoms such as ataxia, blindness and dementia occur". There was also a short video on the site showing a MWHD reacting to sudden movements and noise just as Alf had done, and we learnt that the correct clinical name for the reaction is myoclonus. <http://www.veterinary-neurologist.co.uk/lafora.htm>

I was shocked. Why hadn't anybody told us about this before?

We showed our Vet, who confirmed he had never heard of this condition. The article explained that the symptoms appear to be due to a build up of a starch like material in certain cells in the body, as the result of a genetic mutation. We tried changing his diet (the web article recommended this) but to no avail, but by now we were 90% certain we'd identified what was wrong with Alfie.

In the early morning one day in February 2009 we heard horribly familiar noises from the room where Alfie slept and found him in the middle of another grand mal fit. We comforted him and hoped that was it, but 45 minutes later he had another ..... and didn't seem to come out of it. We rang the Vet's emergency help line and he told us to come straight down and he'd open up for us. Just as we got there, Alf had yet another fit.

In all, he had 15 fits in 12 hours. The vet dosed him up with valium, to no effect, and then anesthetic, but he was still fitting. Mid afternoon, in a desperate last ditch attempt, he gave him a dose of anesthetic that should have knocked a grown man out for 48 hours..... and finally, Alfie calmed down and slept peacefully. An hour later, he started to come round and take an interest in his surroundings, and by the time the veterinary nurses were doing their rounds with evening meals for the other patients he was ravenously demanded to be fed and wagging his tail.

At first, when we were allowed in to take him home, he didn't appear to recognize us, but gradually, light dawned and he gave us a lovely welcome. Gradually, over days, he re-learnt the orientation of the house, the garden, his favourite walks.

So where are we today?

Thanks in a huge part to the care and persistence of our Vet, Dr David Holmes of the Golden Valley Veterinary Practice for operating successfully on his back, sorting out his hernia and of course bringing him around from the Status Epilepticus episode against the odds, Alfie is still here. Alf is now 10 and on a daily dose of phenobarbitone similar to that which many human epileptics need to control their condition (according to the packet, he isn't allowed to drive or operate machinery now, which is a shame!). He hasn't had any grand mal fits since but the myoclonus continues and his vision is gradually getting worse (though there's nothing physically wrong with his eyes). We've invested in a pair of 'Doggles' for him, which seems to help him walking in bright sunlight.

He's also gradually and gently beginning to show signs of losing his marbles – and more importantly his personality. He is still our Alf and we love him just the same – but he's not the same dog, and it can only get worse.

As you'll have gathered from the above, it's a horrible condition. Having used the wonders of email and forums, I'm now in touch with a few other owners of affected dogs and it seems that the actual symptoms do vary from animal to animal, which makes it more difficult to positively identify. I just wonder how many other owners out there are struggling with dogs with some of these symptoms who haven't been able to track down the potential cause, particularly if it isn't widely known within the veterinary profession.

I'm so delighted to hear reports that the Wirehaired Dachshund Club has just agreed unanimously to work on offering mass testing at breed shows, which will help find out just how widespread this genetic condition is in the Miniature Wirehaired Dachshund population – how many have symptoms, and how many are carriers?

It may be too late to help Alfie, and I'd be very cautious about having another Mini Wire, but if there's anything I can do to help towards the eventual eradication of this condition from the mini wire dachshund population, I'd be delighted to help in any way I can, – hence this article, which I hope will help with publicizing the testing programme amongst other pet owners like me, as well as breeders and others who regularly show their dogs.

Watch out for more news on Dachshund Forum in the months to come. Meanwhile, if any other owners have dogs who they think might be affected and would like to talk or read more about it, do get in touch via the 'lafores in mini wire hair dachshunds' topic on the forum.

Gill Key

## Appendix ii

### Aims of the Lafora Dogs Support Group

Aims	How?
<p>providing a central hub for information for people who either think their dog might be suffering, has just been diagnosed or are concerned that they be in the future or etc. This includes details of what to expect, advice on management and the possible impact of dietary adjustments.</p>	<p>Via our:</p> <ul style="list-style-type: none"> <li>• <a href="mailto:laforadogs@btinternet.com">laforadogs@btinternet.com</a> email address,</li> <li>• the new <a href="http://www.laforadogs.org">www.laforadogs.org</a> website</li> <li>• email newsletter</li> <li>• <a href="http://www.dachshundforum.co.uk">www.dachshundforum.co.uk</a> Lafora topic and others</li> <li>• Links to and from other relevant sites, e.g. various websites focusing on Canine Epilepsy, etc.</li> <li>• Via telephone conversations</li> </ul>
<p>Collecting details of Lafora dogs and their owners, in order to provide a database of dogs which should be of beneficial for any further research.</p>	<p>We ask for address and contact details, dogs name, age, sex, date of onset, date of birth and death, details of the condition symptoms and medications.</p> <p>We <u>do not pass</u> on any personal information without asking permission first. I started off by asking for pedigrees too, but aware of the sensitivities, this is no longer a requirement, unless the owner wants to send it to me. I do not share information about any dogs that have appeared more than once in any pedigrees I've seen.</p>
<p>Offering support where necessary for emotionally traumatized owners, who feel the need to be in touch with others who have faced the same challenges</p>	<p>Providing a confidential 'matching service' – using the database and knowledge of the owner gleaned through email and phone conversations, where owners are willing to provide informal counseling advice ,on the understanding that this is an informal basis and does not constitute professional advice.</p>
<p>Raising awareness of the condition in a positive context, whilst seeking to reduce the potential for inaccurate press reports and maintaining the popularity of the breed.</p>	<ul style="list-style-type: none"> <li>• Providing news via the email newsletter to members and provide proof that 'something positive is happening'</li> <li>• Informed and informative articles on the website and in pdf format to help people understand the background and complexities faced, e.g. explanations of the genetics involved, template letters for owners to use to write to puppy owners where their dam or sire has been diagnosed with Lafora</li> </ul>