WHAT ARE THE SIGNS?

L-2-HGA causes a variety of signs but not every dog will display the same ones

* Signs of the condition may start between 6 & 12 months, although some dogs may show signs shortly after birth and others are not diagnosed until after 3 years old
* Signs are not specific to L-2-HGA but may indicate that the condition is present
* Many affected dogs can live into their teens but some die or are euthanised at just a few months old due to severe symptoms, especially fits
* Common signs are:
  - *Seizures (fits, epilepsy)*
  - *Ataxia (wobbly gait, also known as 'dolly walking*
  - *Dementia and other behavioural changes*
  - *Tremors and cramps ('staffie cramps') especially in back legs*
  - *Other reported signs include loss of obedience training, disorientation, wanting t o be alone in a quiet place*
* Signs may start suddenly or gradually.

TESTING

On testing, the results will be shown as One of 3 options:

CLEAR: the dog has 2 copies of the normal gene and will neither develop L-2-HGA, nor pass a copy of the L-2-HGA gene to any of its offspring.

CARRIER: the dog has one copy of the normal gene and one copy of the mutant gene that causes L-2-HGA. It will not develop L-2-HGA but will pass on the L-2-HGA gene to 50% (on average) of its offspring.

AFFECTED: the dog has two copies of the L-2-HGA mutation and is affected with L-2-HGA. It will develop L-2-HGA at some stage during its lifetime, assuming it lives to an appropriate age.

Carriers can still be bred to clear dogs. On average, 50% of such a litter will be clear and 50% carriers; there can be no affecteds produced from such a mating. Pups which will be used for breeding can themselves be DNA tested to determine whether they are clear or carrier.

Testing can be done via the Kennel Club. Follow the link for more information

https://tinyurl.com/L2HGA-test

WHY TEST FOR L-2-HGA?

In the UK, there are two groups of Staffordshire Bull Terriers—those who are routinely tested prior to breeding and those who are not.
The Kennel Club holds information about KC registered dogs that ***are*** tested.

By testing both male and female before breeding, owners can be sure that their pups are 'hereditarily clear' and cannot have the mutation. If just the male or female is tested, whilst the pups may not have symptoms, they could still be carriers

L-2-HGA (L-2-hydroxyglutaric aciduria) in Staffordshire Bull Terriers is a neurometabolic disorder characterised by elevated levels of L-2-hydroxyglutaric acid in urine, plasma and cerebro-spinal fluid.

L-2-HGA affects the central nervous system, with clinical signs usually apparent between 6 months and one year (although they can appear later). Symptoms include epileptic seizures, "wobbly" gait, tremors, muscle stiffness as a result of exercise or excitement and altered behaviour.

The mutation, or change to the structure of the gene, probably occurred spontaneously in a single dog but once in the popula-tion has been inherited from generation to generation like any other gene. The disorder shows an autosomal recessive mode of inheritance: two copies of the defective gene (one inherited from each parent) have to be present for a dog to be affected by the disease. Individuals with one copy of the defective gene and one copy of the normal gene – called carriers – show no symptoms but can pass the defective gene onto their offspring. When two apparently healthy carriers are crossed, 25% (on average) of the offspring will be
affected by the disease, 25% will be clear and the
remaining 50% will themselves be carriers.

The mutation responsible for the disease has recently been identified at the Animal Health Trust. Using the information from this research, we have developed a DNA test for the disease. This test not only diagnoses dogs affected with this disease but can also detect those dogs which are carriers, displaying no symptoms of the disease but able to produce affected pups. Carriers could not be detected by the tests previously available, which involved either a blood or urine test detecting elevated levels of L-2-hydroxyglutarate or magnetic resonance imaging. Under most circumstances, there will be a much greater number of carriers than affected animals in a population. It is important to eliminate such carriers from a breeding population since they represent a hidden reservoir of the disease that can produce affected dogs at any time.

Logan

Logan was our ambassador for a while. Here's what his Mum Dee said about him

"I first met him at Battersea Old Windsor fun day and was told he needed a special home. I fell in love but already having 2 rescue dogs in the house, 1 of which can be reactive, it was something that had to be thought about, so off I went with my current dogs to meet 'Jesse' (now Logan) and they got on instantly! So just to be on the safe side I agreed to foster Jesse first in case things didn't work out. After a few weeks we knew that all would be well and he was most definitely going to stay!

Unfortunately I was unaware of L2 and we always put this down his poor start in life. Then in April this year, Logan was out for a walk with his friends as usual and his legs began to shake, I thought he had been running about too much and took him home, I then popped out and returned 20 mins later to find him having a full seizure . Lucky for us I have a very good vet and after some assessment he came to the conclusion that this was L2, so I sent off the DNA and it came back positive. Since then Logan has had a couple of tremors but not any full seizures. We now know that this is a progressive disorder so we are enjoying life with him as normal but keeping a very close eye out.

So he lives with our dogs Millie and William and 5 rescue Guinea pigs who he loves!
His humans are myself (Dee), his dad Nigel and his human brother and sister Jess and Harry.

I'm currently teaching him simple tricks and we really hope to promote L2-HGA

He is a very happy chap who just loves life "

Sadly, Dee lost Logan to L-2-HGA in March 2017

@willowstaffieL2HGA

Who is Willow?

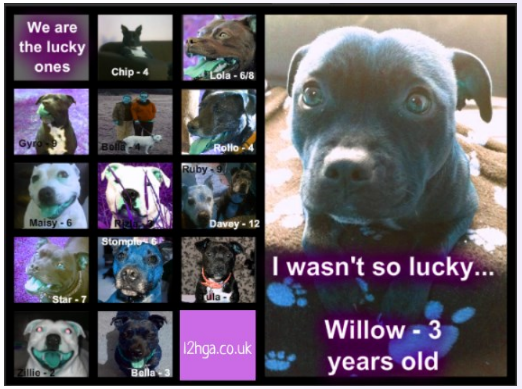
"Willow was a Staffordshire Bull Terrier. We got her in 2010 as a companion for our older Staffie, Megan and in all honesty, Meg chose Willow. We'd taken her with us to see the pups and she showed a clear preference for Willow so that was it. Decision made.

Over time, her owner noticed that while out walking, Willow's gait would change, her back legs seemed to become stiff and she seemed uneasy. A trip to the vet later and a diagnosis of a pulled muscle put her owner's mind at rest for a while but then it happened again but this time was captured on video.

Once again the vet said it was a pulled muscle but Willow's owner wasn't happy with this explanation and sought advice from another vet. The vet there suggested it may be L-2-HGA and organised tests. Sadly, they confirmed that Willow was suffering from this debilitating disease. Although only showing mild symptoms, it was distressing to watch but now there was a reason for it.

Willow was a happy loving soul for the most part but as time went on, she would withdraw and take herself off to a quiet room to be on her own.

Sadly, Willow took a turn for the worst one day and showed major aggression towards her sister. Her family had always said that once Willow's quality of life was threatened, they would do the hardest but kindest thing for her and on the vet's advice, took the decision. Willow was only 3 years old. Needless to say, everyone who knew Willow was devastated. It was hard to face up to the fact that we wouldn't see that cheeky little face any more or get the cuddles she loved so much."



Gemma is Willow's 'Mum' and grew up around dogs with various dog rescue
charities in her 'spare time'. She is married with 2 dogs, both SBT cross.es having lost Megan through old age. Willow was diagnosed with L2 and at that time Gemma realised how little information was widely available about the condition. Since then she has wanted to help spread information about L2HGA to vets, owners & breeders

Andrea is Willow's 'Grandma' and also grew up around dogs. Married with a son as well as a daughter and 3 dogs (all SBT crosses), she runs Deed Not Breed, is a founder member and trustee of Wags & Bones Staffie Rescue Support fundraising and has attended seminars run by Trevor Cooper of DogLaw about BSL. She has previously worked as a volunteer for the RSPCA.

Both are regular Crufts attendees, promoting awareness of L2 -HGA.

Dee joined us when Logan was diagnosed with L2. She grew up with greyhounds and started doing dog
training when she was 8 with her first dog, a SBT x JRT called Sonny and carried on doing training with all my further dogs. When she left school she worked in a pet shop for 3 years and I has 3 NVQs in small animal care. She got Millie and moved into behaviour work. Dee is a Trick Trainer and currently works with reactive dogs. She also does obedience and heel work to music. Since losing Logan, Dee got Hector, her SBT pocket rocket who she does Canicross with