Hendrix's Legacy *Fighting for every Staffy*

Inherited neurological conditions in Staffies

A guide for Staffordshire Bull Terrier breeders

Summary

- Hendrix's Legacy began after Hendrix, a Staffordshire Bull Terrier, died in September 2026 from what we believe was a neurological condition. It has not been identified. It may be inherited, and it may turn out to be a form of NCL (neuronal ceroid lipofuscinosis), but nobody yet knows.
- You cannot DNA test for a condition that has not been identified. Finding the cause depends on cases being noticed, investigated and reported.
- Keep testing for the conditions that can be tested for. For the Staffordshire Bull Terrier, The Royal Kennel Club lists DNA tests for hereditary cataract (HC-HSF4) and L-2-HGA as best practice.[5,6]
- Breeders are often the first to hear when something goes wrong with a dog they bred. What you do with that information matters.

What we know, and what we do not

What is not known. What Hendrix died from. Whether it is inherited. Whether other Staffordshire Bull Terriers have the same condition, and if so how many.

What NCL is. The neuronal ceroid lipofuscinoses (NCLs) are a group of inherited, progressive diseases of the nervous system. In dogs, NCL has been documented in more than 20 breeds and in mixed-breed dogs, and a 2017 review listed 12 causal variants in 8 different genes.[1] Each form needs its own DNA test: a test for one form will not detect another.

Where NCL4A fits. NCL4A is one form of NCL, caused by a variant in the ARSG gene. It has been confirmed in peer-reviewed studies of the American Staffordshire Terrier.[2,3] We have found no published report of it in the Staffordshire Bull Terrier, in the UK or anywhere else.[3] When Hendrix died, NCL4A was thought the most likely explanation. He was never tested, and specialists have since pointed out that other conditions can look similar. It may not be the answer.

How recessive conditions are inherited

Many inherited conditions in dogs, including L-2-HGA in the Staffordshire Bull Terrier and NCL4A in the American Staffordshire Terrier, are autosomal recessive.[3,7] Every dog has two copies of each gene, one from each parent. A dog is only affected if it inherits a faulty copy from both parents. Where a DNA test exists, it shows how many faulty copies a dog has.

Result	Copies of the variant	What it means
Clear (normal/normal)	None	Highly unlikely to develop the condition. Passes a normal copy to every puppy.[5]
Carrier (normal/variant)	One	Highly unlikely to develop the condition, but each puppy has a 50% chance of inheriting the variant.[5]
Affected (variant/variant)	Two	Likely to develop the condition, and passes the variant to every puppy.[5]

What each pairing can produce[5]

Parent 1	Parent 2	Possible puppies	Affected puppies possible?
Clear	Clear	All clear	No
Clear	Carrier	On average half clear, half carrier	No
Clear	Affected	All carriers	No
Carrier	Carrier	On average 25% clear, 50% carrier, 25% affected	**Yes. Avoid.**
Carrier	Affected	On average 50% carrier, 50% affected	**Yes. Avoid.**
Affected	Affected	All affected	**Yes. Avoid.**

Percentages are averages across many litters. Any single litter can differ.

Carriers are not "bad" dogs. The Royal Kennel Club advises that breeding only from clear dogs can reduce genetic diversity, and that carriers can be an important part of a breeding plan when they are mated to clear dogs.[5]

Why an unidentified condition matters to breeders

A recessive condition can pass silently through generations of healthy-looking carriers. If the condition also appears only in adulthood, an affected dog may already have been bred from before anyone knows. That is the situation a DNA test exists to prevent, and it is exactly why an unidentified condition is a risk: until the cause is found, there is nothing to test for.

The breed has been here before. L-2-HGA, now one of the two DNA tests The Royal Kennel Club lists as best practice for the Staffordshire Bull Terrier, was traced to its gene through a study of 21 affected Staffordshire Bull Terriers, published in 2007.[4,6] A test became possible because affected dogs were recognised, investigated and studied.

What breeders can do

1. **Keep testing for what can be tested.** HC-HSF4 and L-2-HGA, plus the eye and elbow schemes The Royal Kennel Club lists for the breed.[6]
2. **Stay in touch with your puppy owners.** Ask them to tell you about any health problem, including unexplained changes in behaviour, balance, movement or sight.
3. **Take unexplained neurological signs seriously.** If a dog you bred, or one of its relatives, develops them, encourage the owner to see their vet, and tell the breed's Lead Breed Health Co-ordinator, through the contact form on The Royal Kennel Club's Staffordshire Bull Terrier breed page.
4. **Talk to your vet before breeding** from any dog that has shown unexplained neurological signs.

5 **Ask your vet about ruling known conditions out.** Where a dog has signs, DNA tests for known conditions, including L-2-HGA and NCL4A, can rule those variants out. A clear result rules out that one variant only. Our guide "DNA testing for Staffies" explains what each test can and cannot tell you.

6 **Share what you know.** Results, including clear results, and health problems in your lines, shared with the Breed Health Co-ordinator, are how a picture of the breed is built.

Why this guide exists

Hendrix was a much-loved Staffordshire Bull Terrier who died in September 2026. **Hendrix was never diagnosed.** No laboratory test was carried out and no sample was kept. Sedatives given when he was young, for example for his neutering, worked as expected for his age and weight. From about two years of age that changed: the sedation his vet prescribed for visits (the "Chill Protocol") repeatedly failed, and this pattern is recorded over several years. In his final hours, a dose his vets estimated was enough for a 160kg dog did not properly sedate him; he weighed 25kg. The professionals attending him considered a neurological condition likely, and at the time NCL4A was thought the most likely explanation. Specialists have since pointed out that other conditions, including ones not yet identified, can look similar. We have reported his case to the breed's Lead Breed Health Co-ordinator. His story is the reason this campaign exists; it is not evidence about any disease.

Next steps

- Read our guide "DNA testing for Staffies: what the tests can and cannot tell you".
- If you are worried about a dog's health, talk to your vet.

This guide is for information only and is not veterinary advice. Information correct as at October 2026; check the sources for updates.

Sources

1. Canine neuronal ceroid lipofuscinoses: promising models for preclinical testing of therapeutic interventions (review). *Neurobiology of Disease*. 2017;108:277-287. doi:10.1016/j.nbd.2017.08.017. PubMed 28860089. https://pubmed.ncbi.nlm.nih.gov/28860089/
2. Abitbol M, Thibaud JL, Olby NJ, et al. A canine Arylsulfatase G (ARSG) mutation leading to a sulfatase deficiency is associated with neuronal ceroid lipofuscinosis. *Proceedings of the National Academy of Sciences USA*. 2010;107(33):14775-80. PMID 20679209. https://pubmed.ncbi.nlm.nih.gov/20679209/
3. OMIA (Online Mendelian Inheritance in Animals), University of Sydney. OMIA:001503-9615, Lysosomal storage disease, ARSG related, in dog. Last updated 3 December 2025. https://omia.org/OMIA001503/9615/
4. Penderis J, Calvin J, Abramson C, Jakobs C, Pettitt L, Binns M, Verhoeven N, O'Driscoll E, Platt S, Mellersh C. L-2-hydroxyglutaric aciduria: characterisation of the molecular defect in a spontaneous canine model. *Journal of Medical Genetics*. 2007;44:334-340. doi:10.1136/jmg.2006.042507. https://pmc.ncbi.nlm.nih.gov/articles/PMC2597990
5. The Royal Kennel Club. Understanding and using your dog's DNA test results. Undated. https://www.royalkennelclub.com/health-and-dog-care/health-dog-care/health/getting-started-with-health-testing-and-screening/dna-testing/breeding-advice-for-dna-test-results
6. The Kennel Club. Health Standard. January 2025. https://kc-media-production.azureedge.net/media/qvffqvwg/kc_health_standard.pdf
7. Wisdom Panel. L-2-Hydroxyglutaric aciduria (Discovered in the Staffordshire Bull Terrier). Undated. https://www.wisdompanel.com/en-us/dog-health-conditions/l2hga