

# Dandy-Walker Syndrome in Toy Fox Terriers

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We have recently seen several Toy Fox Terrier pups who exhibited cerebellar ataxia from the time they began to walk. Post-mortem examination of the brain of affected pups showed a congenital malformation of the brain that is called the Dandy-Walker Syndrome in children. Talking to breeders, we have gotten reports of similar signs in other pups. Whether this is an isolated problem in one family or an indication of a wider problem in the breed remains to be seen. The purpose of this article is to provide information about the condition so that breeders can be alert to any future cases and help us find answers.

## What is cerebellar ataxia?

The term cerebellum in Latin means “the little brain”. The cerebellum is the part of the brain responsible for coordinating movements. Ataxia comes from a Greek term meaning “without order”. When the cerebellum does not function properly, the dog can move, but the movement is poorly coordinated. They are not weak, in fact, often the movements a dog with ataxia makes are too strong. They have a goose-stepping gait and when excited or running, their legs may appear to be going every which-way. Sometimes they have problems with their balance and will fall frequently.

## What is the Dandy-Walker malformation?

Making a brain is hard work. As a puppy's brain is forming in its mother's uterus, a variety of signals ensure that each portion of the brain develops at the right time. In the Dandy-Walker malformation, a portion of the cerebellum (the part of the brain responsible for coordination) doesn't get the message to develop like it should. The result is a shriveled up cerebellum that doesn't function properly. Affected pups may have other abnormalities, such as hydrocephalus (water on the brain) or blindness.

## What else can look like Dandy-Walker Syndrome?

Any disease that damages the cerebellum can cause ataxia that looks like Dandy-Walker Syndrome. Infectious diseases like canine distemper, some toxins, or other hereditary disease can all affect the cerebellum. Sometimes clues can be identified by a veterinarian that will point toward some other cause. The Dandy-Walker malformation can be seen readily on an MRI brain scan or if the pup must be euthanized, a post-mortem examination of the brain may provide a definitive answer.

## Is this a hereditary disease?

In children, Dandy-Walker Syndrome can either be hereditary, or caused from another disease such as an infection that affects the fetus during the wrong time in development. Though it is too soon to say conclusively, Dandy-Walker Syndrome appears to be hereditary in the Toy Fox Terrier and is most likely a recessive trait. In a recessive disease, both parents of an affected pup can appear normal. All animals have two copies of each gene, one that is inherited from the mother and one that is inherited from the father. A dog that has one normal gene and one gene that causes the disease is a carrier of the trait. They show no symptoms because the one good gene is enough for their brain to develop normally, but they will pass that bad gene on to about half of their offspring. If a carrier dog is bred to another carrier, then some of the pups (one quarter of them on average) will get a bad gene from each parent. Without one good gene to carry the day, the cerebellum cannot develop normally and the unlucky pup has cerebellar ataxia.

## How do we find the gene responsible?

The goal in dealing with hereditary diseases is to identify the gene responsible. Then we can develop a DNA test that will aid breeders in avoiding the disease in the future. Genes contain the genetic code that programs everything about an animal from the color of their coat to how their brains develop. Genetic disease occurs when a mutation interferes with the ability of a gene to function normally. Each dog has an estimated 20,000 individual genes, any one of which could contain a mutation that causes Dandy Walker Syndrome. We now have the tools, however, to find the mutation responsible for such diseases.

## How can I help?

If you have a litter with a pup you believe might be affected, please see our website, [www.caninegeneticdiseases.net](http://www.caninegeneticdiseases.net) or contact us directly. We can help your veterinarian in determining whether or not this is the problem in your pup. In return, we would ask your help in collecting the samples and information necessary to continue searching for the gene responsible for this disease. Your continuing support will be necessary to achieve our goal.

## **Any information provided to us will be kept strictly confidential**

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*Ataxia refers to poor coordination of movements. A dog with ataxia has exaggerated movements which produce a “goose stepping” gait and can cause frequent falls.*