CEREBELLAR ABIOTROPHY

What is cerebellar abiotrophy?

The cerebellum is the part of the brain that regulates the control and coordination of movement. In this condition, cells in the cerebellum mature normally before birth, but then deteriorate prematurely causing clinical signs associated with poor coordination and lack of balance. The Purkinje cells in the cerebellum are primarily involved; cells in other areas of the brain may also be affected.

How is cerebellar abiotrophy inherited?

An autosomal recessive mode of inheritance has been confirmed or is strongly suspected for the abiotrophies listed below, with the exception of x-linked cerebellar ataxia in the English pointer, which has an x-linked mode of inheritance.

What breeds are affected by cerebellar abiotrophy?

Neonatal cerebellar abiotrophy (very rare) - Affected cells start to degenerate before birth, so that signs of cerebellar dysfunction are present at birth or when the pup first walks.

Beagle, samoyed

Postnatal cerebellar abiotrophy - Cells in the cerebellum are normal at birth and begin to degenerate at variable times thereafter.

Australian kelpie, border collie, Labrador retriever - Clinical signs are first seen at 6 to 12 weeks, and the condition worsens quickly (over a few weeks).

Airedale - There is early onset (12 weeks of age) and a slow progression of clinical signs.

Bern running dog, Bernese mountain dog, bull terrier, German shepherd - Signs are seen by 6 months of age.

Gordon setters - Clinical signs develop at 6 months to 2 years of age, and the progression is slow (months to years).

Brittany spaniels - The onset of clinical signs is late (average age 10 years), and the condition progresses slowly.

English pointers - Clinical signs develop around 8 to 12 weeks and become pronounced by 16 months of age.

Cerebellar and extrapyramidal nuclear abiotrophy - Cells in other regions of the brain deteriorate as well.

Kerry blue terrier - The first signs occur at 6 to 12 weeks and dogs are unable to stand by 1 year of age.

Rough-coated collie, miniature poodle - Clinical signs are seen at 4 to 12 weeks of age, and there is rapid deterioration over a few weeks to months.

For many breeds and many disorders, the studies to determine the mode of inheritance or the frequency in the breed have not been carried out, or are inconclusive. We have listed breeds for which there is a consensus among those investigating in this field and among veterinary practitioners, that the condition is significant in this breed.

What does cerebellar abiotrophy mean to your dog & you?

The cerebellum is the part of the brain that regulates the control and coordination of voluntary movement. The clinical signs of cerebellar dysfunction in affected dogs may include poor balance, a wide-based stance (feet planted far apart), stiff or high-stepping gait, apparent lack of awareness of where the feet are (standing or walking with a foot knuckled over), and head or body tremors. These signs worsen either quickly or slowly (see breed list above). Affected dogs may become unable to climb stairs or stand without support. They have normal mental alertness.

Where other regions of the brain are also affected, you may see signs such as behaviour change (loss of house training, aggression), confusion, blindness, and seizures.

How is cerebellar abiotrophy diagnosed?

This is a rare disorder. The clinical signs are suggestive of cerebellar disease, particularly if they are seen in a breed in which abiotrophy is known to occur. Your veterinarian will do tests to rule out other conditions that can cause similar signs.

For the veterinarian: Routine diagnostic tests are normal with this condition and a definitive diagnosis can only be made by brain biopsy or on post-mortem. MRI may be helpful in dogs in which there is gross cerebellar malformation; however generally with this condition, the cerebellum appears grossly normal. Histopathologic abnormalities are often minimal and do not seem to correlate with the severity of cerebellar signs.

How is cerebellar abiotrophy treated?

There is no treatment for this condition. Dogs do not recover from this disorder and usually at some point (depending on the rate of the progressive deterioration that occurs), euthanasia becomes the best option.

Breeding advice

Affected dogs, their parents (carriers of the trait), and their siblings (suspect carriers) should not be bred. With x-linked cerebellar ataxia, only male pups are affected, and the mother is the carrier of the trait.

FOR MORE INFORMATION ABOUT THIS DISORDER, PLEASE SEE YOUR VETERINARIAN.

Resources

March, P.A. 1996. Degenerative brain disease. Veterinary Clinics of North America: Small Animal Practice. 26(4): 945-971.

Coates, J.R. 1996. Weeble, wobble, roly, poly: a study of cerebellar disease. ACVIM-Proceedings of the 14th Annual Vet. Med. Forum. pp 684-687. **This reference provides a comprehensive breed list, with associated clinical and pathologic findings.**

O'Brien, D. 1993. Hereditary cerebellar ataxia. ACVIM-Proceedings of the 11th Annual Vet. Med. Forum. pp 546-549.

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