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Commentary

A Brief Description on Canine Hereditary Ataxia

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DESCRIPTION

Degenerative cerebellar diseases have been described in a large number of dog breeds, but most of these reports are descriptions of individual animals. However, there are many cerebellar diseases that have been proven to be hereditary and pose a major problem for the ethnicity involved. These breeds include Gordon setter, Brittany Hound, Old English sheepdog, American Staffordshire terrier, and Scottish terrier. Clinically, this group of diseases can cause insidious ataxia episodes, which are characterized by distance measurement disorder (abnormal range of motion), intention tremor (head tremor when the animal is focused on something), trunk swing (the body moves from One side visibly shakes) to the other side walking), spontaneous nystagmus (uncontrolled abnormal eye movements), and loss of physiological nystagmus. The age of onset and the rate of symptom progression vary from species to species. Gordon Setters and Old English Sheepdogs first show signs when they are between 6 and 40 months of age, while Scottish Terriers show signs at 8 weeks. The American Staffordshire Terrier and Breton Spaniel signs appear later, usually between the ages of 3 and 8. The signs progress slowly, but develop at different rates depending on the affected species, eventually the animal cannot walk without falling and has difficulty eating and drinking due to intentional tremors. Among the breeds listed above, it is unusual for the Scottish terrier to be euthanized due to disease of the cerebellum. Although their gait is unusual, they are not disabled. In contrast, although the American Staffordshire terrier develops symptoms later, it is usually euthanized due to illness within 2 to 8 years of the onset of symptoms. Old English Shepherd and Golden Setter signs may stabilize, but some improve until they need to be euthanized.

The diagnosis of these ante-mortem diseases is based on the presence of signs of cerebellar dysfunction in the neurological examination of otherwise healthy dogs, a consistent history of chronic progressive symptoms (which may be related to the affected dog), and normal blood tests And the cerebellar atrophy magnetic field. An autopsy can make a clear diagnosis; cerebellar atrophy is visible to the naked eye, there is degeneration of Purkinje cells in histopathology, and then neurons in the molecular and granular layers of the cerebellum are lost. Histopathological changes are mainly limited to the cerebellar cortex of the species we are studying. To date, all autopsies of dogs suspected of having hereditary ataxia based on clinical signs and medical history have confirmed the disease, so consistent medical history and clinical signs seem to be a reliable means of diagnosing the disease.

The pedigree analysis of Gordon setter, Old English Sheepdog, American Staffordshire Terrier, and Scottish Terrier indicates that the disease is inherited in an autosomal recessive manner. This means that a dog must have two copies of the mutant allele to show signs of disease. Dogs with only one copy of the mutant allele (carrier) are clinically normal, and the breeder does not know that if they are crossed with another carrier, they may produce affected dogs. The recessive nature of the disease and the late appearance of symptoms in certain breeds of diseased dogs lead to involuntary full reproduction of diseased and carrier dogs, thereby spreading harmful alleles within the breed. This is especially true for the American Staffordshire Terrier. In this breed, symptoms usually do not appear until five years of age or older, and we estimate that the prevalence of harmful alleles in the pedigree we examined is 39%. In fact, this problem has been recognized in dogs from all geographic regions of the United States and Europe. We continue to recruit affected dogs at a rate of 1 dog per month for our research, and we have been solving this problem for nearly 5 years since the beginning. Obviously, genetic testing is indeed needed to identify carriers and affected states of all four dog breeds.

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