NCL Description for Dachshunds

The scientific literature contains two reports of NCL in Dachshunds. We have recently prepared a third report of Dachshund NCL and submitted it for publication. The first report, published in 1977 by Cummings and de Lahunta, described a Wirehaired Dachshund that began showing hind leg weakness at three years of age. As the disease progressed the dog developed a head tremor, trunkle ataxia and a hypermetric (goose-stepping) gait. The dog died at four and a half yeas of age and histopathologic examination of the brain suggested NCL. It has, however, been subsequently reported that Wirehaired Dachshunds with these same symptoms and histopathologic findings do not have NCL but instead have a related neurodegenerative disease know as mucopolysaccharidosis IIIA (Fischer et al., 1998). Mucopolysaccharidosis IIIA of Dachshunds is caused by a deletion in their heparin sulfate sulfamidase gene (Aronovich et al., 2000).

In 1980 Vandevelde and Fatzer described an adult onset NCL in Longhaired Dachshunds. At the University of Missouri, we have been studying a distinct, juvenile-onset form of NCL, which also occurs in Longhaired Dachshunds and causes neurodegeneration leading to death at approximately one year of age. We have identified and characterized the mutation responsible for this disease and we have devised a DNA test that distinguishes between normal Dachshunds, Dachshunds that are carriers of the disease who could produce affected puppies if bred to another carrier, and affected Dachshunds destined to endure neurodegeneration and early death (Awano et al., submitted for publication, 2006).

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Adult-Onset NCL of Dachshunds

Age of on-set of clinical signs: 4.5 - 6.5 years

Age of euthanasia: 4.5 - 7 years

Abnormalities often observed by the owner:
Mental changes: behavior changes, dullness, compulsive pacing
Changes in gait and posture: pacing, ataxia (uncoordinated movements), hypermetria (high-stepping or goose-stepping)
Visual abnormalities: unknown
Seizures/convulsions: present
Other changes: polyphagia (excessive hunger), polydypsia (excessive thirst)

Abnormalities observed upon clinical examinations:
Clinical neurologic changes: ataxia, hypermetria.
Clinical ophthalmic changes: none reported
Visual abnormalities: none reported

Retinal changes: none reported
Electroretinography (ERG): not assessed
Other clinical findings: ceroid lipofuscin in gastrointestinal tract

Histopathology
Brain: *Light microscopy* – intraneuronal accumulation of cytosomes with fluorescent and tinctorial properties of ceroid lipofuscin throughout the central nervious system. *Electron microscopy* – large numbers of intracytoplasmic membrane-bound cytosomes and lipofuscin granules with mostly membraneous content sometimes assuming fingerprint patterns.
Eyes: not assessed.
Other organs and structures: none reported

Mode of inheritance: unknown

Gene containing mutation: Unknown

References:

Vandevelde M, Fatzer. *Neuronal ceroid-lipofscinosis in older Dachshunds*. Vet Pathol 1980;17:686-692.

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Juvenile-Onset NCL of Dachshunds

Age of on-set of clinical signs: 9 months

Age of euthanasia: 12 months

Abnormalities often observed by the owner:
Mental changes: loss of housebreaking, dullness, compulsive circling, aggressiveness, unresponsiveness to previously learned commands
Changes in gait and posture: pacing, ataxia (uncoordinated movements), hypermetria (high-stepping or goose-stepping)
Visual abnormalities: present
Seizures/convulsions: present
Other changes: vomiting

Abnormalities observed upon clinical examinations:
Clinical neurologic changes: ataxia, hypermetria.
Clinical ophthalmic changes: none reported
Visual abnormalities: none reported
Retinal changes: none reported
Electroretinography (ERG): not assessed
Other clinical findings: none reported

Histopathology
Brain: *Light microscopy* -- Depletion of neurons in the cerebellar cortex; accumulation of cytoplasmic granules throughout the CNS. *Electron Microscopy* – Cytoplasmic granules containing curvilinear forms.
Eyes: not assessed.
Other organs and structures: none reported

Mode of inheritance: autosomal recessive

Gene containing mutation: to be announced

References:

Awano et al., submitted for publication, 2006

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Testing recommendations

There is currently no DNA test for adult-onset NCL in Dachshunds. If you own a Dachshund showing signs of adult-onset NCL, please contact us (email Liz Hansen at HansenL@missouri.edu or call 573-884-3712). We do have a DNA test for juvenile-onset NCL of Dachshunds. This test identifies affected dogs, non-symptomatic carriers, and normal dogs. The cost is $40 per test. Instructions and forms for testing are available in the "Sample Submission" section of the website - http://www.CanineGeneticDiseases.net/CL_site/mainCL.htm. We recommend testing for all Dachshunds under 18 months of age that exhibit the behavioral and gait changes listed above. Furthermore, we recommend the testing of close relatives of young Dachshunds that have exhibited these changes. We also recommend the testing of Dachshunds expected to be used extensively in breeding programs to determine if they are carriers. Because this is a serious recessive disease, carriers should only be bred to dogs tested “normal” to ensure that the mating will not produce affected puppies destined to develop this fatal neurodegenerative disease.

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