

VETERINARY TECHNICAL DATASHEET

Neonatal Cerebellar Cortical Degeneration or Cerebellar Abiotrophy, (NCCD)



Mutation Found In :Beagle

Disorder Type

- Nervous system

Disease Severity

- Moderate/severe

Background

Neonatal cerebellar cortical degeneration or cerebellar abiotrophy (NCCD) is encountered in several dog breeds. Cerebellar abiotrophy is characterized by progressive degeneration of neurons in the cerebellar cortex. An affected dog suffers from ataxia (uncoordinated movements) and dysmetria (improper measuring of distance in muscular acts). The age of onset and the progression of clinical signs seem to vary in different breeds due to different causative mutations. This variant is found in the Beagle.

Key Signs

- Loss of coordination
- Loss of balance

Clinical Description

The typical signs of neonatal cerebellar cortical degeneration (NCCD) in Beagles can be seen when affected puppies start to move at three weeks of age. The clinical signs include cerebellar ataxia, wide-based stance, loss of balance, and dysmetric gait with inability to regulate rate and range of movement. The affected puppies have a normal state of alertness.

Mode of Inheritance

- autosomal recessive

Gene Name

- SPTBN2

Next Steps

Treatment is supportive care and assistance with daily activities as needed depending on the severity of the dog's clinical signs.

References

Forman OP, De Risio L, Stewart J, Mellersh CS, Beltran E. Genome-wide mRNA sequencing of a single canine cerebellar cortical degeneration case leads to the identification of a disease associated SPTBN2 mutation. BMC Genet 13:55, 2012.