

# VETERINARY TECHNICAL DATASHEET

Alaskan Husky Encephalopathy, (AHE)



Mutation Found In :Alaskan Husky

## Disorder Type

- Nervous system

## Disease Severity

- Severe

## Background

The underlying cause of AHE is a mutation in the gene encoding a transporter protein required for thiamine transport into the cells of the central nervous system. Due to the severity of this disorder, affected dogs are usually euthanized within 2 to 7 months from the onset of clinical signs.

## Key Signs

- Seizures
- Altered mentation
- Behavioral abnormalities
- Dysphagia
- Central blindness
- Hypermetria
- Ataxia
- Tetraparesis

## Clinical Description

The first signs of AHE are usually observed at the age of 6 months to 3 years of age. Affected dogs may have a sudden onset of clinical signs or a chronic history with slowly progressing signs. The characteristic signs of AHE include generalized seizures, altered mentation, behavioral changes, dysphagia (eating difficulties), loss of vision, hypermetria (overreaching movements), ataxia (uncoordinated movements), and tetraparesis (weakness in voluntary movement of all four limbs).

## Mode of Inheritance

- autosomal recessive

## Gene Name

- SLC19A3

## Next Steps

For affected dogs, the progression of clinical signs should be carefully monitored, ensuring the dog is as comfortable as possible. Affected dogs are usually euthanized within 2 to 7 months from the onset of clinical signs on welfare grounds, although some dogs may live for months to years before the signs progress.

## References

Vernau KM, Runstadler JA, Brown EA, Cameron JM, Huson HJ, Higgins RJ, Ackerley C, Sturges BK, Dickinson PJ, Puschner B, Giulivi C, Shelton GD, Robinson BH, DiMauro S, Bollen AW, Bannasch DL. Genome-wide association analysis identifies a mutation in the thiamine transporter 2 (SLC19A3) gene associated with Alaskan Husky encephalopathy. PLoS One 8:e57195, 2013.

