

Lagotto Storage Disease, (LSD)

In brief

Lagotto storage disease (LSD) is a progressive neurological disorder characterised by cerebellar ataxia. Histological examination reveals neuronal vacuolisation in the peripheral and central nervous systems. Aggregation of vacuoles can also be seen in several other tissues, but only vacuolisation of the nerve cells seems to be relevant. Lagotto storage disease affects the Lagotto Romagnolo breed. The disorder is inherited in an autosomal recessive manner.

Clinical overview

The onset of clinical signs and pattern of progression vary considerably between individuals. The first signs can be seen at the age of 4 months to 4 years. Typically the first observable sign of LSD is usually progressive ataxia (uncoordinated movements). The first signs of slowly progressing ataxia are not necessarily noticed by the owner. Some of the affected dogs also suffer from episodic nystagmus (involuntary eye movement). In some cases, nystagmus is the first sign noticed. Lagotto storage disease is a progressive condition leading to behavioural changes such as restlessness, depression, and aggression. The life expectancy of affected dogs depends on the progression of the disorder and the severity of the signs. Some dogs can live for several years with mild signs, but dogs with severe clinical signs are usually euthanised earlier.

References

Online database

Online Mendelian Inheritance in Animals, OMIA (<http://omia.angis.org.au/>). Faculty of Veterinary Science, University of Sydney.

Scientific articles

Kyöstilä K, Syrjä P, Jagannathan V, Chandrasekar G, Jokinen TS, Seppälä EH, becker D, Drögemuller M, Dietschi E, Drögemuller C, Lang J, Steffen F, Rohdin C, Jäderlund KH, Lappalainen AK, Hahn K, Wohlsein P, Baumgärtner W, Henke D, Oevermann A, Kere J, Lohi H, Leeb T. A missense change in the ATG4D gene links aberrant autophagy to a neurodegenerative vacuolar storage disease. PLoS Genet 2015 Apr 15;11(4):e1005169.

Disease severity

Severe

Clinical signs

- Ataxia
- Episodic nystagmus
- Changes in behaviour (restlessness, depression, aggression)

Mode of Inheritance

Autosomal Recessive

Results of the genetic test are reported as follows:

- Clear
-  Carrier
-  At risk

Mutation(s) found in:

Lagotto Romagnolo - Romagna Water Dog