

Benign Familial Juvenile Epilepsy or Remitting Focal Epilepsy

In brief

Benign familial juvenile epilepsy (BFJE) is a neurological disorder encountered in the Lagotto Romagnolo breed. Affected dogs suffer from focal-onset epileptic seizures in puppyhood. The seizures invariably resolve spontaneously by four months of age. In some cases, carriers might also present epileptic signs. BFJE is inherited in an autosomal recessive manner.

Clinical overview

Epileptic seizures are caused by a disruption of the normal electrochemical activity of the brain. Onset of focal epileptic seizures in BFJE is at 5-9 weeks of age. The seizures consist of whole-body tremors, ataxia (uncoordinated movements), and stiffness. The epileptic signs can sometimes be associated with alterations of consciousness. Frequency of seizures can vary between individuals: there can be several seizures a day or sporadic seizures a few times a week. Usually an affected puppy seems completely normal between the seizures. However, in severe cases there can be neurological signs, such as generalised ataxia between the seizures. BFJE form of epilepsy is relatively benign since the seizures typically end after 4 months of age.

References

Online database

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

Scientific articles

Seppälä EH, Jokinen TS, Fukata M, Fukata Y, Webster MT, Karlsson EK, Kilpinen SK, Steffen F, Dietschi E, Leeb T, Eklund R, Zhao X, Rilstone JJ, Lindblad-Toh K, Minassian BA, Lohi H. LGI2 Truncation Causes a Remitting Focal Epilepsy in Dogs. *PLoS Genet* 7(7): e1002194, 2011.

Jokinen TS, Metsähonkala L, Bergamasco L, Viitmaa R, Syrjä P, Lohi H, Snellman M, Jeserevics J, Cizinauskas S. Benign Familial Juvenile Epilepsy in Lagotto Romagnolo Dogs. *J Vet Intern Med* 2007;21:464-471.

Disease severity

Mild

Clinical signs

- Focal-onset epileptic seizures

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