Mucopolysaccharidoses Type IIIA, (MPS IIIA)

In brief

Mucopolysaccharidoses (MPS) are a group of rare inherited lysosomal storage disorders resulting from a deficiency in the enzymes required for degradation of glycosaminoglycans. Enzyme deficiency causes accumulation of metabolic by-products in cells disrupting their normal function. Several different forms of mucopolysaccharidoses with different clinical signs have been recognised in dogs. Type IIIA mucopolysaccharidosis (MPS IIIA) is encountered in Dachshunds and the New Zealand Huntaway. MPS IIIA is characterised by progressive neurological signs with little effect on bones and other organs as occurs in other forms of mucopolysaccharides. MPS IIIA is inherited in an autosomal recessive manner.

Clinical overview

Mucopolysaccharidosis type IIIA is characterised by progressive ataxia (uncoordinated movements). The first signs of ataxia can be seen in pelvic limbs progressing later to all four limbs. The clinical signs include dysmetric gait and loss of balance. For example, climbing stairs becomes increasingly difficult as the disease progresses. An affected dog may also sway while standing. First clinical signs are usually observed in the third year of life. The affected dogs are usually euthanised a few years following the onset of clinical signs.

References

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

Scientific articles

