

VETERINARY TECHNICAL DATASHEET

Mucopolysaccharidosis Type IIIA, (MPS IIIA); mutation originally found in Dachshund



Mutation Found In :Dachshund (Shorthaired), Dachshund (Miniature Shorthaired), Dachshund (Wirehaired), Dachshund (Miniature Wirehaired), Dachshund (Longhaired), Dachshund (Miniature Longhaired)

Disorder Type

- Metabolic

Disease Severity

- Moderate/severe

Background

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 recognized in dogs. Type IIIA mucopolysaccharidosis (MPS IIIA) is encountered in Dachshunds and the New Zealand Huntaway. MPS IIIA is characterized by progressive neurological signs with little effect on bones and other organs as occurs in other forms of mucopolysaccharides.

Key Signs

- Pelvic limb ataxia
- Severe generalized spinocerebellar ataxia
- Hypermetria
- Exaggerated reflexes

Clinical Description

Mucopolysaccharidosis type IIIA is characterized by progressive ataxia (uncoordinated movements), and first clinical signs are usually observed in the third year of life. The signs of ataxia are initially seen in pelvic limbs progressing later to all four limbs. The clinical signs include dysmetric gait and loss of balance. An affected dog may also sway while standing.

Mode of Inheritance

- autosomal recessive

Gene Name

- SGSH

Next Steps

This disorder is progressive with no cure. Wellbeing of affected dogs should be monitored. Owners should be advised that as clinical signs progress, affected dogs may have difficulties on smooth surfaces and with climbing stairs. Euthanasia is often elected on welfare grounds when clinical signs become severe.

References

Aronovich EL, Carmichael KP, Morizono H, Koutlas IG, Deanching M, Hoganson G, Fischer A, Whitley CB. Canine heparan sulfate sulfamidase and the molecular pathology underlying Sanfilippo syndrome type A in Dachshunds. Genomics 68:80-84, 2000.