VETERINARY TECHNICAL DATASHEET

von Willebrand's Disease (vWD) Type 3; mutation originally found in Kooiker Hound

Mutation Found In :Kooikerhondje, Scottish Terrier, Shetland Sheepdog

Disorder Type

- Blood

Disease Severity

- Moderate

Background

The clotting pathway is a complex process. The vWF glycoprotein complexes with factor VIII and both are required for platelet adhesion and preventing the rapid clearance of factor VIII. Von Willebrand's disease (vWD) is a bleeding disorder affecting multiple breeds and several genetic variants have been characterized. Type 3 is the most severe form of vWD. The disease is inherited in an autosomal recessive manner.

Key Signs

- Bleeding following surgery or trauma
- Bleeding from the nose
- Bleeding from the gums
- Gastrointestinal bleeding
- Blood in the urine

Clinical Description

Type 3 von Willebrand's disease is a moderate to severe clotting disorder. Typical clinical signs include mucosal bleeding, such as nose bleeding, bleeding from the gums, gastrointestinal bleeding, and blood in the urine. Exceptionally excessive and prolonged bleeding may be observed after a trauma or surgery, and spontaneous bleeding may also be encountered. This condition can lead to death if untreated. Most affected dogs will have a normal PT/aPTT but have prolonged bleeding. When assayed, these dogs have no appreciable levels of vWF present; specific vWF factor can be assayed at a reference laboratory. Performing a functional test like a buccal-mucusal bleeding time prior to surgery is recommended.

Mode of Inheritance

- autosomal recessive

Gene Name

- VWF

Next Steps

Some dogs may respond to desmopressin acetate (DDAVP) therapy. In most cases, therapy is limited to supportive care (which can include transfusions if needed) and avoiding surgery. Performing a functional test like a buccal-mucusal bleeding time prior to surgery is recommended.

References

Pathak EJ. Type 3 von Willebrand's disease in a Shetland sheepdog. Can Vet J 45(8):685-687, 2004.

Rieger M, Schwarz HP, Turecek PL, Dorner F, van Mourik JA, Mannhalter C. Identification of mutations in the canine von Willebrand factor gene associated with type III von Willebrand disease. Thromb Haemost 80(2):332-337, 1998.

Venta PJ, Li J, Yuzbasiyan-Gurkan V, Brewer GJ, Schall WD. Mutation causing von Willebrand's disease in Scottish Terriers. J Vet Intern Med 14(1):10-9, 2000.

Ackerman L. The Genetic Connection. Lakewood: American Animal Hospital Association Press, 2011.
