

VETERINARY TECHNICAL DATASHEET

von Willebrand's Disease (vWD) type 1; mutation originally found in Doberman Pinscher



Mutation Found In :Bernese Mountain Dog, Coton de Tulear, Doberman Pinscher, Drentse Patrijshond, German Pinscher, Kerry Blue Terrier, Manchester Terrier, Papillon, Pembroke Welsh Corgi, Poodle (Standard), Stabyhoun

Disorder Type

- Blood

Disease Severity

- Mild/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 1 is the mildest form of vWD in which the level of von Willebrand's factor is reduced though all the multimers are present. Many cases are subclinical but can be associated with an increased bleeding tendency after surgery or trauma. The disease is inherited in an autosomal recessive manner, though some carriers can have clinical signs.

Key Signs

- Bleeding following surgery or trauma

Clinical Description

Type 1 von Willebrand's disease is the mildest form of vWD and while many dogs may be subclinical, some dogs may exhibit more severe clinical signs. Excessive bleeding may be observed after a trauma or surgery. Not all affected pups will exhibit the same severity of clinical signs as these are related to the amount of vWF present, which vary between affected individuals. An affected dog will have a normal PT/aPTT but have prolonged bleeding. When assayed, these dogs usually have low levels of vWF; specific vWF factor can be assayed at a reference laboratory. Performing a functional test like a buccal-mucosal bleeding time prior to surgery is recommended. Some carriers may show clinical signs though dogs with two copies of the mutation tend to be more severely affected. Medications known to interfere with clotting should be avoided. Some dogs may exhibit some improvement when treated with desmopressin acetate.

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-mucosal bleeding time prior to surgery is recommended.

References

Brooks MB, Erb HN, Foureman PA, Ray K. von Willebrand disease phenotype and von Willebrand factor marker genotype 1n Doberman Pinschers. Am J Vet Res. 62(3):364-369, 2001.

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