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Von Willebrand Disease

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BASIC INFORMATION

Description

Von Willebrand disease (VWD) is an inherited bleeding disorder of the platelets that occurs in dogs, cats, and people. Platelets are responsible for helping blood clot. Von Willebrand factor is a protein produced by platelets that helps platelets stick together and form a clot. When this factor is abnormal or deficient, platelets do not effectively stick together, and the result is prolonged or inappropriate bleeding.

Causes

Three types of VWD exist, depending on the type and amount of factor present. In type I, the factor is normal but is present in very low amounts, and the severity of bleeding varies. Type I is most common in the Doberman pinscher, German shepherd dog, Shetland sheepdog, and standard poodle. In type II, the factor proteins are abnormal and decreased, which results in more severe bleeding episodes than in type I. Type II disease is typically seen in German shorthaired and wirehaired pointers. In type III, the factor is not made at all, and this is the most severe form. Type III is most commonly identified in the Scottish terrier, Chesapeake Bay retriever, and Shetland sheepdog.

Clinical Signs

Excessive or abnormal bleeding is the primary clinical sign of VWD. Because this is an inherited condition, signs may be seen early in life. Excessive bleeding from the gums occurs when baby teeth fall out. Nose bleeds, blood in the urine, dark or tarry stools, and swollen, painful joints may also occur. Sometimes excessive bleeding is not noted until after trauma or during surgery. Events that further affect platelet function, such as vaccination or use of nonsteroidal anti-inflammatory medications, may cause temporary bleeding and mild bruising.

Diagnostic Tests

In general, dogs with a suspected bleeding problem initially undergo routine laboratory testing, which may include a complete blood count, platelet count, biochemistry profile, and screening coagulation tests. If VWD is suspected, a test may be done in the clinic to assess platelet function; this is called the *buccal mucosal bleeding time*. This test is not specific for VWD, but an abnormal result increases suspicion for the disease.

To diagnose the presence of VWD, the amount of von Willebrand factor in the blood must be measured. The results can be labeled as normal, borderline, or abnormal. Dogs with abnormal amounts are at risk for increased bleeding tendency. Borderline

results are sometimes rechecked, because the laboratory assay can be affected by many factors. The usual test for VWD does not identify the type (I, II, or III) present. If it is necessary to determine the type, further specialized tests must be run.

Some studies have shown a relationship between low thyroid hormone levels (hypothyroidism) and VWD, so your veterinarian may recommend thyroid tests to ensure that this is not a complicating factor.

TREATMENT AND FOLLOW-UP

Treatment Options

There is no cure for VWD. Treatment involves administration of normal von Willebrand factor during episodes of bleeding or when surgery is anticipated. Von Willebrand factor by itself cannot be purchased, but blood products containing the factor (such as plasma or cryoprecipitate) can be obtained from normal donors. Transfusions of these blood products are given during times of active bleeding and prior to surgery.

Injections of a drug called *DDAVP* (desmopressin acetate) may temporarily increase blood levels of von Willebrand factor for a few hours and is sometimes administered prior to surgery. There is some controversy as to whether this medication is effective. Dogs diagnosed with hypothyroidism are usually placed on a thyroid supplement.

Follow-up Care

Because there is no cure, prevention is aimed at eliminating affected dogs from the breeding pool. All dogs from breeds at risk for this disease and dogs with a history of VWD in their genetic line should be tested. Affected and carrier individuals should not be bred. Breeds routinely tested include the Doberman pinscher, golden retriever, Shetland sheepdog, Rottweiler, schnauzers, German shepherd dog, standard poodle, Scottish terriers, Pembroke Welsh corgi, German shorthaired and wirehaired pointers, and others.

Prognosis

Without treatment, severely affected dogs may bleed to death following injuries or surgeries not considered to be life-threatening. Dogs with borderline levels of von Willebrand factor may have increased bleeding tendencies that are not life-threatening but must be managed proactively with measures such as limiting the use of medications that affect platelet function, avoiding unnecessary invasive surgeries, and giving transfusions or DDAVP before planned surgeries.