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Canine von Willebrand's disease *von Willebrand's disease*

Affected Animals:

Dogs of many breeds can be affected with von Willebrand's disease. Some breeds have a higher incidence than others, including Doberman pinschers, Scottish terriers, Airedales, basset hounds, dachshunds, golden retrievers, German shepherds, Manchester terriers, and Shetland sheepdogs. The condition has been identified in more than 50 breeds of dogs.

Overview:

Occurring because of abnormalities involving a protein called von Willebrand's factor, von Willebrand's disease is an inherited bleeding disorder of dogs. A large number of breeds can be affected with the disease, but the breeds most commonly diagnosed include Doberman pinschers, Scottish terriers, Shetland sheepdogs, and Airedale terriers. Dogs with the illness will many times have no symptoms at all, but when symptoms do occur, abnormal bleeding results. Both spontaneous bleeding and excessive hemorrhage in response to a stimulus such as trauma or routine surgery have been observed.

Since the exact genetic manner in which the disease is transmitted is often unknown, and since reliable tests to identify carriers of the disorder are not always available, von Willebrand's disease is a very difficult condition to eradicate. When excessive hemorrhage occurs, it is best treated by careful use of physical techniques that stop bleeding, such as suturing and cautery. The administration of blood or blood components that help to replace deficient concentrations of von Willebrand's factor will also be important. In some animals, pre-treatment with a substance called DDAVP may allow for temporary increases in von Willebrand's factor concentration that will protect them during periods when they are at risk for bleeding.

Clinical Signs:

Many dogs with von Willebrand's disease never have clinical signs. Others will bleed inappropriately during surgeries, other stressful episodes, or when they are injured. Less commonly, affected dogs will bleed spontaneously, often from the nasal cavity or the urinary tract.

Symptoms:

See clinical signs.

Description:

Von Willebrand's disease refers to a bleeding disorder that results from reduced concentrations of a specific protein called von Willebrand's factor. This substance plays a key role in the adherence of platelets, the cells that normally help blood clot, to collagen,

a protein exposed by injury to blood vessels. Dogs with von Willebrand's disease may bleed spontaneously, most frequently from the nose, the mouth, and the urinary tract. More frequently, affected dogs bleed only when there is a stimulus for bleeding, such as an injury or surgery. Von Willebrand's disease can be a frustrating condition to diagnose, manage, and prevent because not all dogs with low concentrations of von Willebrand's factor have symptoms related to the disease.

Von Willebrand's disease is often classified according to the type and presence of different components of the von Willebrand's factor, called multimers. In type I von Willebrand's disease, all the different multimers are present, but in concentrations that are below normal. This type is the most common form of the disease and is characterized by bleeding from the mouth, nose, and urinary tract. Although spontaneous bleeding may not occur, excessive hemorrhage may be seen during elective procedures such as dentistry, ear cropping or tail docking, spaying, and castration. Breeds affected with Type I von Willebrand's disease include the Doberman pinscher, Airedale, Shetland sheepdog, and Pembroke Welsh corgi.

With type II von Willebrand's disease, smaller multimers are present, and there is a lack of the larger multimers, which are more important in the clotting process. This is a relatively uncommon form of the disease, but can result in severe bleeding episodes. German shorthaired pointers are most commonly affected.

Type III von Willebrand's disease is the most severe form. Animals with this type of the illness lack any measurable von Willebrand's factor and as a result, they can experience severe blood loss. The breeds most commonly diagnosed with this form of the disease are Scottish terriers and Chesapeake Bay retrievers.

Diagnosis:

The easiest test for inappropriate bleeding due to von Willebrand's disease is called the oral mucosal bleeding test. This is performed by making a standardized incision on the inside of the lip. This test is not specific for von Willebrand's disease since other conditions can cause prolonged mucosal bleeding times.

The most commonly used test that is more specific for the diagnosis is the measurement of von Willebrand's factor antigen on a plasma sample. Other diseases can cause this result to be artificially increased, however.

Prognosis:

The prognosis for von Willebrand's disease depends on whether signs of excessive hemorrhage are present, and whether the hemorrhage is controlled before critical amounts of blood are lost. For those dogs that are hemorrhaging but have no access to blood or blood component therapy, the outlook can be very grim.

Transmission or Cause:

The cause of von Willebrand's disease is deficiency of a protein called von Willebrand factor. This substance plays a key role in the adherence of platelets, the cells that normally help blood clot, to collagen, a protein exposed by injury to blood vessels. Von Willebrand factor may also play a role in the adhesion of platelets to other platelets during blood clot formation. Deficiencies in von Willebrand's factor are inherited.

Treatment:

The primary treatment for von Willebrand's disease is the administration of blood or blood products to patients with active or anticipated bleeding episodes. A blood product called

cryoprecipitate contains large amounts of von Willebrand factor, but it is seldom available. More commonly, the veterinarian will use fresh plasma, or plasma that was frozen immediately after collection and then thawed. Whole fresh blood may be used if hemorrhage has been severe.

The administration of a drug called DDAVP may be helpful in preventing hemorrhage in some affected animals if it is given prior to the time that bleeding occurs. This drug increases levels of von Willebrand factor available for the clotting process. DDAVP can also be given to dogs donating blood prior to blood collection, so that samples with high von Willebrand factor activity can be obtained. Not all dogs respond to DDAVP.

Prevention:

Prevention of von Willebrand's disease is a very difficult issue. Since the disease is hereditary, the ideal way to eliminate it would be to avoid breeding affected dogs. However, not all dogs with low von Willebrand's factor concentrations have significant bleeding. The mere fact that the concentration is low does not always mean that clinically significant problems will occur, even in breeds in which a significantly high number of dogs have reduced von Willebrand's factor. If all dogs that tested low for the factor were eliminated from breeding, then breeding programs would be quite restricted. Certainly, though, it makes sense not to breed dogs that have had clinically significant episodes of bleeding due to von Willebrand's disease, no matter what their breed.

In an affected dog, problems may be avoided prior to elective surgeries by remembering to screen for this condition, especially if previous bleeding episodes have occurred or if there is a familial history of bleeding. Pre-treatment with DDAVP may help avert disastrous consequences, as will having blood products on hand in case they are needed.