My Weimaraner puppy was recently diagnosed with Ehlers-Danlos Syndrome, what is it and is there a cure?

Ehlers-Danlos Syndrome is an extremely rare, inherited, congenital, connective tissue disorder that has been recognized in humans and animals for many years. So, you certainly have a very rare pet indeed.

The primary problem is abnormal production or degradation of collagen. Collagen is a compound important to the strength of connective tissues that are located in the skin, tendons, and ligaments. Other names for this disorder are cutaneous asthenia (weak skin) or dermatosparaxis (torn skin).

The cause is a biochemical defect in the production and/or degradation of collagen. Collagenase, an enzyme that breaks down collagen, has been found to have activity levels 2 ½ times above the normal level in affected animals. Several breeds of dogs, including the Weimaraner, cats, sheep, cattle, and mink have been reported with this disorder.

Due to the lack of collagen strength, the affected animal has skin that is very weak, stretches easily, is loosely attached to underlying tissues, and tears easily. So with minimal trauma, the skin tears resulting in large, gaping wounds.

The wounds usually heal readily but leave thin, highly visible “cigarette paper” scars. The skin can be stretched to extreme lengths and may hang loose in folds, especially on the legs and the throat. In addition to the weak skin, widening of the bridge of the nose, subcutaneous hematomas (bleeding under the skin), elbow hygromas (fluid and swelling around the elbow), joint instability, and rarely eye disorders may be seen.

An absolute diagnosis may require skin biopsies with biochemical and/or ultra structural studies (electron microscopy), although, using the skin extensibility index along with the above described signs will usually provide strong support of a diagnosis.

The skin extensibility index is performed by taking a fold of skin from over the back and extending it to its maximal distance above the back without causing pain and then measuring the vertical height of the skin. The distance from the back of the head to the base of the tail is also measured. The vertical height of the skin is divided by the distance from the back of the head to the base of the tail, and multiplied by 100. In affected dogs this percentage should be greater than 14.5 percent, and the cat, greater than 19 percent.

There is no known cure. Since it is inherited, the animal should not be used for breeding and the sire and dam that produced the affected dog should not be used for breeding either.
The following environmental modifications should be addressed to avoid injury: 1) make sure visitors don’t grab the dog, 2) sharp corners or rough surfaces in the house should be smoothed or padded, 3) resting and sleeping areas should be well-padded, 4) quick attention to skin diseases that might cause the pet to scratch is important, and 5) wounds should be sutured

promptly.

If all of the above are done, and the affected animal does not have joint instability, they can live long lives. Although not a proven treatment, some authors believe Vitamin C, 500mg twice a day, may lessen the skin fragility.

This column is provided by the faculty of the OSU Boren Veterinary Medical Teaching Hospital.

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