PURDUE UNIVERSITY

Animal Disease Diagnostic Laboratory

Purdue Canine Genetics Lab

Basset Hound Glycogen Storage Disease (BHGSD)

Disease: Glycogen Storage Disease

Breed: Basset Hounds

About the condition:

An inherited metabolic disease, specifically a Glycogen Storage Disease, has been reported in Basset Hounds (BHGSD). Cases have been confirmed by post-mortem examination by board certified veterinary pathologists.

This condition proceeds in a gradual, subtle way, but with harmful effects; affected dogs typically appear healthy until they suddenly are not, which can be anywhere between 3-6 years of age. This is particularly challenging because affected dogs may have been bred prior to showing any signs of disease. Clinical signs can be subtle to non-existent and can be very difficult to notice. The first sign something is wrong can be sudden death of the dog. If warning signs are observed, they can include lethargy, exercise intolerance, and laziness (although this is certainly a "tough call" in some Basset Hounds, who can be inclined toward sedentary lifestyles already), heavy/labored breathing and other signs of cardiac disease, including enlarged heart on imaging (radiographs/X-rays and/or ultrasound/echocardiogram). Some affected dogs had other disorders, including gastrointestinal problems, and inflammation of the central nervous system, however, the main cause of death is heart failure due to the accumulation of abnormal glycogen.

BHGSD is a metabolic disease where glycogen, an energy storage product in the body, accumulates abnormally and excessively in several different tissues, including the heart. Autopsy of deceased cases identified significantly elevated amounts of glycogen in the cells of the heart muscle, which ultimately results in the dog's heart failing.

Research carried out at Purdue University indicates that this disease is inherited in an autosomal recessive manner. We have identified a mutation within a disease-associated gene, and all affected dogs have two copies of this mutation. We are now offering a genetic test which allows owners to determine their dog's status for this mutation (clear, carrier, or affected) in order to diagnose affected dogs and to guide future breeding decisions. Carriers do not need to be removed from the breeding population; however, they should only be bred to clear mates to avoid producing affected offspring.

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