FVII Deficiency in Alaskan Klee Kai Updated Information as of March 15, 2006

Several hereditary bleeding disorders have been identified in many different canine breeds and involve clotting (coagulation) factor deficiencies, platelet disorders, and von Willebrand disease. Coagulation factor VII (FVII) deficiency has been known to occur in Beagles for decades, and there are a few reports of FVII deficiency in the Alaskan Malamute, Bulldog, and a mixed breed dog. Very recently hereditary FVII deficiency was identified in a bleeding Alaskan Klee Kai dog and its family, as well as unrelated asymptomatic Alaskan Klee Kai dogs. A DNA test to identify the mutation responsible for FVII deficiency in Alaskan Klee Kai dogs has been developed at the University of Pennsylvania.

Dogs with hereditary FVII deficiency may exhibit an increased bleeding tendency following trauma or surgery or rarely appear to develop spontaneous bleeding. There are few reports of severe bleeding requiring blood transfusions, and some FVII-deficient dogs may remain unrecognized. As this is an autosomal recessive disorder, the diseased/mutant gene (allele) may be unknowingly passed on through generations not only via asymptomatic carriers but also affected dogs, as they may not show obvious signs. Carriers have one mutant and one normal gene and appear clinically normal, but they can pass the defective gene to their offspring. Only a small number of Alaskan Klee Kai dogs have been tested thus far, and hence the frequency and bleeding tendency remain to be elucidated.

Screening Alaskan Klee Kai dogs with a clotting test (PT assay) may suggest FVII deficiency, and measurement of plasma FVII coagulant activity could confirm a diagnosis of FVII deficiency. Results of the PT assay are normal for carriers, and measurement of plasma FVII coagulant activity will not accurately identify carriers, as there is overlap of FVII activities between carrier and normal dogs.

A mutation-based DNA test to screen Alaskan Klee Kai dogs (and Beagles) for FVII deficiency has been developed (Dr. Beth Callan, principal investigator) and has then been established at the Josephine Deubler Genetic Disease Testing Laboratory of the University of Pennsylvania School of Veterinary Medicine (Dr. Urs Giger, director; Mr. Adam Seng, Research Specialist). This test can clearly identify affected, carrier, and normal (also known as clear) Alaskan Klee Kai dogs. We recommend testing of any Alaskan Klee Kai dog with signs of bleeding, as well as its relatives. Furthermore, it is advisable to screen any Alaskan Klee Kai dog, particularly popular sires, prior to breeding to limit the spread of this bleeding disorder. Carriers could still be used in future breeding programs. Knowing which dog is a carrier or normal (clear) will allow the targeted breeding of carriers with desirable traits to normal dogs without ever producing affected dogs, as long as the offspring are also tested and only clear dogs used thereafter. With a breed of this size you cannot afford to neuter all animals with a mutant gene, as you want to preserve their desirable traits and the gene pool.

The Josephine Deubler Genetic Testing Laboratory at the University of Pennsylvania is offering screening for FVII deficiency in Alaskan Klee Kai dogs at a reduced introductory rate of $50 per dog through May 31, 2006, provided that a pedigree and adequate clinical information on the submission form are included ($75 per sample without pedigree). Samples suitable for this DNA test include 1-2 mLs EDTA-anticoagulated blood (preferable) or 2-3 cheek swabs (obtained with special cytology brushes). Cytology brushes and test submission forms are available through the United Alaskan Klee Kai Association (http://www.uakka.com) and the Alaskan Klee Kai Association of America (http://akkaoa.org). Test submission forms may also be downloaded from our website http://www.vet.upenn.edu/penngen. We are also offering test kits when requested with a self-addressed stamped envelop. For more information on canine FVII deficiency, please contact Dr. Beth Callan (215-898-3999; callan@vet.upenn.edu) or Dr. Urs Giger (215-898-8830; penngen@vet.upenn.edu). Blood and dried cheek swab samples, along with test submission forms and pedigrees, may be submitted in a ziploc bag to Dr. Urs Giger/FVII Rm 4006, Ryan Veterinary Hospital, University of Pennsylvania, 3900 Spruce Street, Philadelphia, PA, 19104-6010. Test results are generally available within 3 weeks of receipt of samples and are sent only to submitter of samples. All information is kept strictly confidential.