## Vogt-Koyangi-Harada Disease (Human) UveoDermatological Syndrome (Canine)

UveoDermatological (UV) Syndrome or VKH-like syndrome is the canine equivalent of Volt-Koyanagi-Harada's Disease (VKH) found in humans. As the symptoms are very similar to the human version it is often referred to as VKH, although dogs do not appear to get ear or hearing problems as in the human version and unlike the human version it is more likely to be seen in MALES.

In canines ,mostly found in Nordic breeds, Samoyed, Siberian Husky, Alaskan Malamute, Chow Chow and Akita and their crosses but can occur in any breed.

In humans it is mostly found in Asian, Middle Eastern, Hispanic and Native American populations but skin colour alone is not a predisposing factor, VKH distinctly UNCOMMON in Africans it is rarely seen in Northern European individuals. In humans the condition occurs most frequently in FEMALES

Onset of the condition can be sudden with no obvious signs but may manifest with depigmentation and conjunctivitis in dogs this is likely to involve depigmentation and/or swelling of nose and eyelids followed by detached retina (very painful). The condition usually manifests in young adults, 20 – 50 in humans but has been seen in infant. Similarly in dogs, onset is generally around 13 months to 6yrs but occasionally seen in puppies.

The general characteristics are the same for humans and dogs affecting eyes and skin.

- Uveitisis\* is always present in UveoDermatological Syndrome (UVD)/VKH but is not necessarily symptomatic. There are numerous causes of Uveitis and it is commonly found condition in all breeds, however, in breeds such as Akitas, Siberian Husky, Alaskan Malamute and Samoyed where there is thought to be a predilection for VKH a close watch should be kept for other UVD symptoms. \*Uveitis inflammation of the middle layer of the eye, this is likely to be very painful and a serious threat to vision. If left untreated there is a risk of retinal detachment and glaucoma. Retinal discharge may be accompanied by smelly discharge. The changes to the eye is common to humans and dogs, the changes it produces in the eye of previously affected dogs can be seen ophthalmoscopically, eye examinations may identify the early emergence of the condition but cannot be used to predict if a dog is likely to develop the condition.
- Vitiligo (loss of skin pigment) common in humans and dogs, almost always present
- Poliosis (depigmentation loss of colour in coat in dogs, commonly seen around the eyes and nose) in humans depigmentation is also apparent along with disorders involving hair, skin, sweat and oil glands.
- In dogs mucous membrane area are often involved, eyelids, mouth, anus and vulva and sometimes pads on paws where crustiness or blisters may be seen.

The condition appears to have different categories, owners of UVD affected dogs have described symptoms as mild, affecting eyes mainly; moderate affecting eyes and skin pigment to a degree; severe where both skin and eyes are severely affected. This compares to the different categories of VKH in humans where the condition complete VKH where symptoms are severe involving eye changes, depigmentation in eyes and skin, neurological and auditory changes. Incomplete VKH

involves changes in the eyes, and <u>either</u> skin <u>OR</u> neurological and auditory signs but not both. Probable VKH only involves ocular disease.

## **Pathophysiology**

What causes the changes? Currently the focus is on the major Histocompatibility Complex involved in the recognition of body tissue. In UVD or VKH-like syndrome the body's melanin-containing cells are attacked by antibodies, when this happens the melanocytes of the eye and skin express an antigen that is seen as a foreign body by the dogs/human's immune system.

Dog Leukocyte Antigen (DLA) was present in UVD affected dogs but not in unaffected dogs.

It is felt by researchers that as the condition tends to occur in families and is associated with isolated populations that a genetic predisposition exists that is triggered by some sort of event such as infection or stress. UVD is not something that can be 'caught' like kennel cough.

In humans there is also uncertainty about the exact nature of the condition, however, the focus is also on the central mechanism involved involving the Human Leukocyte Antigen (HLA) currently VKH is considered to be a cell-mediated autoimmune disease directed against melanocytes.

The loss of melanocytes suggests an infectious or autoimmune basis for VKH; however, the association between VKH and certain racial and ethnic groups suggests an immunogenetic predisposition for the condition.

In essence this suggests that there is still uncertainty about the exact nature of the disease in humans but there are some strong indications that there is likely to be a genetic predisposition for the condition.

Studies carried out on dogs in 2005 found an association between DLA DQA1)00201 and UVD syndrome in both the American Akita and Japanese Akita populations in Asia and Europe also known to be affected by UVD. A similar condition has also been observed in Samoyeds, Irish Setters (19) and the **CHOW CHOW** (1). Despite the association of DQA1\*00201 with UVDin American Akitas is not predictive (diagnostic) for UVD, but it is seen as a suggesting dogs with this allele are at higher risk of developing the disease. However not all dogs with UVD carry this allele.

## What can we do as breeders?

Routine eye checks will detect any dog that has previously been affected by UVD, and may detect early signs of the condition. UVD is a recurrent condition and can be difficult to control, early diagnosis may minimise the damage done.

Vigilance, where chows are diagnosed with uveitis with no specific cause (trauma, infection etc) AND have other signs associated with UVD such as depigmentation it is strongly recommended that they have diagnostic skin biopsies of affected areas to confirm or eliminate UVD/VKH as a diagnosis. Whilst the eye condition is always present in UVD/VKH the condition is confirmed by skin biopsy as the histopathological changes to the tissues of the eye can only be shown following the removal of the eye.

Peter Bedford (eye specialists) believes that there is a genetic predisposition to the VKH-like syndrome (UVD), has diagnosed the condition in Akita's and Siberian Husky and believes it to be typically found in Spitz Type dogs, however he has not to date seen the condition in a CHOW

Similarly Rosario Cerundolo (Skin Specialist) has diagnosed the condition in various breeds, but has never seen UVD/VKH in CHOWS.

The Willows Veterinary Centre in Solihull that specialises in eye conditions has informed me that they have treated several chows with VKH-like syndrome in the past few years. The surgeon was understandably reluctant to give details because of client confidentiality.

## **CURRENT INFORMATION**

I am aware of one current case that is fully documented with a copy of the laboratory test results confirming the diagnosis.

I am also in communication with the owner of another confirmed case who is also responding well to treatment. The owner has supplied a letter from her veterinary surgeon confirming the diagnosis for our records.

Most importantly the Breed Council must be informed of any diagnosed cases to be able to evaluate the scale of the problem in the breed. The recommendation of the specialists is that animals that have produced affected offspring should be withdrawn from breeding programs. It is noted that this is not always the case in other breeds that have more experience of the condition; the view is that dogs that have produced affected offspring should not be mated back into the lines that produced the effected offspring but can be used as an outcross.

At this time there is no way of identifying potential sufferers or to identify carriers assuming that there is a genetic predisposition for the condition to develop. There is a strong recommendation that affected animals should not be breed from.