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## Vogt-Koyanagi-Harada Syndrome

### Issue Description

*In UDS the autoimmune reaction is directed against the melanocytes (the pigment-producing cells) of both skin and eyes (iris). The disease causes severe uveitis but early diagnosis and therapy may prevent serious vision loss. In the skin it most commonly causes depigmentation (Vitiligo) of the periocular region, lips and nose.*

### Other Names

*UDS, VKH Syndrome, Uveodermatologic Syndrome*

### Causes

Vogt-Koyanagi-Harada-like syndrome (VKH) is a rare, idiopathic and autoimmune disease in dogs. The exact cause of this syndrome is still unknown both in people and dogs.

As in other autoimmune diseases there are several causes that may trigger the syndrome (e.g. stress). It is also speculated that the immune-reaction is initially triggered by a virus, but there is no certainty yet.

### Breeds Affected

Although the condition has been reported in many different breeds of dogs, UDS is predisposed in following breeds and their crosses:

- Akitas
- Siberian Husky
- Alaskan Malamute
- Samoyed
- Chow Chow

The disease is more common in male dogs than female dogs. The age of onset ranges from 13 months to 6 years.

### Symptoms

Often the first noticeable sign of UDS is uveitis. The skin and hair changes typically follow within three to six months after the eye disease has begun. The changes in hair colour are seen in some 90% and depigmentation of skin is seen in some 50% of the affected dogs.

Skin and hair abnormalities are caused by vitiligo, a depigmentation of skin areas, which seems to be present in almost every affected dog. The most typical areas of vitiligo are periocular region, nose and lips. Sometimes also palate, footpads, vulva, scrotum and anus are affected.

The hair changes are whitening of patches of hair and hair loss. The skin and hair changes are mainly a cosmetic concern but sometimes erythema, ulceration, and crusting of skin is present. Pruritus may be a feature and lymphadenopathy is common.

Dogs with UDS have various degrees of uveitis involving iris, ciliary body and choroids. The uveitis may be very painful and the changes may appear as bloodshot eyes, constricted pupils, cloudy eyes or changes in eye color. Conjunctivitis will often be followed by a detached retina, which shows as a milky blue surface on the eye ball. Unlike in the human disease, deafness and meningitis are not a feature in dogs.

## Diagnosis

The best way to confirm this diagnosis is by a skin biopsy (the lip is said to be the best location). Treatment, however, is focused on the eye disease as this has the most serious outcome – blindness - while the skin disease is generally cosmetic.

## Treatment

Because the eye disease has the most serious outcome (blindness) and the skin disease is typically only a cosmetic concern, treatment then focuses on the eye disease. As in other autoimmune diseases, the primary goal of therapy is to suppress the body's immune response with large doses of systemic glucocorticosteroids such as prednisone. To prevent blindness, on-going immune suppression is needed and more potent drugs like Cytoxan, Azathioprine or Imuran are used if steroids fail. Continued topical treatment is also needed, usually with steroid-containing eye drops or steroid injection. Controlling eye pain may also be required.

In cases where vitiligo has occurred, protecting the affected areas e.g. with sunblocks may be necessary to prevent the sunburn and squamous cell carcinoma that may follow.

## Prognosis

Prognosis is poor overall. The uveitis tends to recur and may result in permanent blindness due to cataract and retinal degeneration after long term separation or inflammation. Even vigorous therapy may not control the situation. With aggressive treatment some dogs are able to regain some vision but, in general, vision cannot be preserved and a more realistic goal is to control the eye pain.

In patients in whom inflammation is controlled, useful vision may be retained and melanosis of the skin may recur.

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