

Pemphigus Complex

ABOUT THE DIAGNOSIS

Pemphigus is the name given to a group of autoimmune diseases that affect the skin. Normally, the function of the immune system is to protect the body by attacking foreign substances or organisms that try to invade the body. Autoimmune diseases occur when the immune system mistakenly perceives healthy tissues of the body as foreign and proceeds to attack that part of the body. In pemphigus, the immune system attacks important substances that attach the skin cells to one another. The result is a separation of the skin cells causing the spontaneous formation of blisters and ulcers. These blisters and ulcers occur even though there has been no external trauma to the skin; it is the weakness of the skin's tissue caused by the autoimmune reaction that causes these lesions. Crusts (scabs) and pustules (pimples) also occur commonly with pemphigus. The severity of the disease is related to the depth within the skin where the immune reaction occurs. In pemphigus vulgaris, the deeper skin layers are affected, making this the most serious of the pemphigus diseases. Other pemphigus complex conditions that involve layers of the skin that are more superficial include pemphigus foliaceus, pemphigus erythematosus, and pemphigus vegetans.

Pemphigus foliaceus is the most common form of pemphigus. In dogs, scales (flakes of dried skin), crusts (scabs), and pustules (pimples) occur on the face and the ears. The footpads may thicken and crack. Gradually, skin over other parts of the body becomes affected. In cats, the skin around the base of the nails and around the nipples is frequently involved.

Pemphigus vulgaris, since it occurs in a deeper layer of the skin, causes severe skin ulcers (open sores). Areas where the mucous membranes meet the skin are frequently involved, so ulcers and crusts may be seen around the mouth, eyelids, and anus. The skin of the armpits and the groin often can be affected. The ears and the skin around the nails can also be involved. Without treatment, this disease is usually fatal. With treatment and monitoring, pemphigus vulgaris may be put in remission, and a good quality of life is realistic.

Pemphigus vegetans causes thick crusts (scabs) that are attached to the skin and wart-like growths from groups of skin pustules. This is a relatively benign disease.

For all types of pemphigus, the diagnosis is made by excluding other diseases with similar appearance and from a pathologist's examination under the microscope of skin biopsy specimens. At the microscopic level, biopsy samples will show the separation of the skin cells (called acantholysis) that is characteristic of the condition. Skin biopsies are generally taken under heavy sedation or general anesthesia, but it may be possible to take skin biopsies under just local anesthesia if the condition is not too painful.

LIVING WITH THE DIAGNOSIS

Pemphigus foliaceus and pemphigus vulgaris usually require sustained and potentially intensive treatment, and that treatment may need to be lifelong. Periodic monitoring by your veterinarian is required because medications can have a potential for side effects. Pemphigus erythematosus and pemphigus vegetans are milder diseases and may only need intermittent treatment.

TREATMENT

Pemphigus vulgaris is treated with immunosuppressive therapy. This usually consists of high-dose corticosteroids (cortisone-type drugs) plus other immunosuppressive drugs given by mouth. Azathioprine or a combination of tetracycline and niacinamide is the treatment most often used in dogs; chlorambucil or cyclosporine is the usual choice for cats. The treatment for pemphigus foliaceus depends upon its severity and may range from topical treatments (treatments that are applied directly to the affected skin) to immunosuppressive therapy similar to that for pemphigus vulgaris. Pemphigus erythematosus and pemphigus vegetans are less severe diseases and often are controlled with corticosteroids alone. Treatment may only need to be given intermittently when the disease flares up. When the condition responds to treatment and symptoms improve, medications often can be slowly tapered to a low level and sometimes stopped (in consultation with your veterinarian).

DOs

- Keep your pet with pemphigus out of direct sunlight; UV radiation/light worsens the condition.
- Handle immunosuppressive medications with caution (e.g., wash your hands after handling tablets/pills) to avoid taking in even tiny amounts of the medications unnecessarily.

DON'Ts

- Don't stop medication or lower the dose without consulting your veterinarian. The benefit achieved with careful treatment and monitoring over days or weeks can be lost very quickly if you stop the medication prematurely.

WHEN TO CALL YOUR VETERINARIAN

- If your pet's symptoms worsen.
- If you notice any of the medication side effects listed under Signs to Watch For below.

SIGNS TO WATCH FOR

- Corticosteroids can cause a variety of side effects. Many are mild, expected reactions such as increased panting (dogs), increased appetite, and increased drinking and urination. These are not alarming, but you should be aware that they can occur and act accordingly (e.g., increase the number of a dog's walks per day for urinating if water consumption has increased).
- Other side effects of immunosuppressive therapy can result in vomiting, lack of appetite, inactivity, and infections. The occurrence of these symptoms should prompt you to call your veterinarian.

ROUTINE FOLLOW-UP

- Pets diagnosed with pemphigus should have frequent checkups at first to monitor the effectiveness of treatment and the course of the disease. After the disease is under control, less frequent rechecks are needed. If oral immunosuppressive treatment is needed, your veterinarian will use blood tests to periodically

monitor your pet for side effects of the medication, with the goal of detecting these before they cause problems. Immunosuppression lowers the resistance to infection, so tests may be run to look for infection, such as a urinalysis and urine culture to check for bladder infections. Immunosuppressive drugs such as azathioprine or chlorambucil may cause bone marrow suppression, which can be detected by blood tests.

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