

Pemphigus

Basics

OVERVIEW

- A group of diseases in which the immune system attacks the skin (known as “auto-immune dermatoses”); auto-immune diseases are ones in which the body produces antibodies against its own tissue; an “antibody” is a protein that is produced by the immune system in response to a specific antigen (a substance that induces an immune response)—when the body is exposed to the antigen (in the case of pemphigus, the antigen is some part of the skin), the antibody responds, resulting in signs of disease
- The pemphigus group of diseases is characterized by varying degrees of loss of tissue on the surface of the skin, frequently with inflammation (known as “ulceration”); dried discharge on the surface of a skin lesion (known as a “crust”); and formation of small, raised skin lesions containing pus (known as “pustules”) and blisters or small, circumscribed elevation of the outer layer of the skin filled with clear fluid (known as “vesicles”)
- Affects the skin and sometimes the moist tissues of the body (known as “mucous membranes”)
- Forms identified in animals include pemphigus foliaceus, pemphigus erythematosus, pemphigus vulgaris, panepidermal pustular pemphigus/vegetans, canine benign familial chronic pemphigus (Hailey-Hailey disease), and paraneoplastic pemphigus; type of pemphigus based on location of skin lesions and microscopic appearance of skin lesions



GENETICS

- Benign familial chronic pemphigus (Hailey-Hailey disease)—may be a genetic disease

SIGNALMENT/DESCRIPTION OF PET

Species

- Pemphigus foliaceus, erythematosus, and vulgaris—dogs and cats
- Panepidermal pustular pemphigus—dogs

Breed Predispositions

- Pemphigus foliaceus—Akitas, bearded collies, chow chows, dachshunds, Doberman pinschers, Finnish spitzes, Newfoundlands, and schipperkes
- Pemphigus erythematosus—collies, German shepherd dogs, and Shetland sheepdogs

Mean Age and Range

- Usually middle-aged to old pets

SIGNS/OBSERVED CHANGES IN THE PET

Pemphigus Foliaceus

- Scales (accumulations of surface skin cells, such as seen in dandruff); crusts (dried discharge on the surface of skin lesions); pustules (raised skin lesions containing pus); superficial loss of skin tissue (known as “erosions”); reddened skin (known as “erythema”); hair loss (known as “alopecia”); circular patterns of hair loss (alopecia) bordered by scales or surface peeling of the skin (pattern is known as an “epidermal collarette”); and thickening of the skin (known as “hyperkeratosis”) of the footpads with furrows or slits (known as “fissures”)
- Occasional blisters (vesicles) are transient
- Common involvement—head, ears, and footpads; often becomes generalized
- Lesions involving the moist tissues of the body (mucous membranes) and areas where the moist tissues of the body contact the skin, such as the lips (areas known as “mucocutaneous junctions”) are uncommon
- Cats—nipple and nailbed involvement are common
- Sometimes enlarged lymph nodes (known as “lymphadenopathy”), fluid buildup in the skin (known as “edema”), depression, fever, and lameness (if footpads involved) may be present; however, pets are often in good health
- Variable pain and itchiness (known as “pruritus”)
- Secondary bacterial infection is possible

Pemphigus Erythematosus

- Same signs as for pemphigus foliaceus
- Lesions usually confined to head, face, and footpads
- Loss of pigment of the moist tissues (mucous membranes) and skin (known as “mucocutaneous depigmentation”) more common than with other forms of pemphigus; loss of pigment may precede crusting

Pemphigus Vulgaris

- Ulcers of the mouth are frequent, and may precede skin lesions
- Ulcerative lesions; superficial loss of skin tissue (erosions); circular patterns of hair loss (alopecia) bordered by scales or surface peeling of the skin (pattern is called epidermal collarettes), blisters, and crusts (dried discharge on the surface of skin lesions)
- More severe than pemphigus foliaceus and pemphigus erythematosus
- Affects moist tissues of the body (mucous membranes), areas where the moist tissues of the body contact the skin, such as the lips (mucocutaneous junctions), and skin; may become generalized
- Area under the front legs and between the rear legs (known as the “axillae and groin”) often involved
- Positive Nikolsky sign (new or extended erosive lesion created when lateral pressure is applied to the skin near an existing lesion)
- Variable itchiness (pruritus) and pain
- Lack of appetite (known as “anorexia”), depression, and fever
- Secondary bacterial infections are common

Panepidermal Pustular Pemphigus

- Pustule (raised skin lesion containing pus) groups become masses that ooze
- Involvement of the mouth has not been seen
- No systemic illness

CAUSES

- Undetermined—genetics and a possible triggering event (such as a viral infection or medication)

RISK FACTORS

- Undetermined

Treatment

HEALTH CARE

- Initial inpatient supportive therapy for severely affected pets
- Outpatient treatment with initial frequent hospital visits (every 1–3 weeks); taper to every 1–3 months when remission is achieved and the pet is on a maintenance medical regimen

- Severely affected pets may require antibiotics and hydrotherapy/soaks

DIET

- Low-fat—to avoid inflammation of the pancreas (known as “pancreatitis”), which can be a side effect of steroids and (possibly) azathioprine therapy

SURGERY

- Surgical biopsy of the skin lesion and the skin surrounding the lesions

Medications

Medications presented in this section are intended to provide general information about possible treatment. The treatment for a particular condition may evolve as medical advances are made; therefore, the medications should not be considered as all inclusive

PEMPHIGUS FOLIACEUS AND PEMPHIGUS VULGARIS

- Steroids—prednisone or prednisolone
- Chemotherapeutic drugs and other drugs to decrease the immune response—more than half of pets require medications other than steroids to decrease the immune response; these drugs generally work in conjunction with prednisone, allowing reduction in dose and side effects of the steroid; examples include azathioprine, chlorambucil, cyclophosphamide, cyclosporine, and dapsone
- Gold-salt treatment or chrysotherapy—gold salts are used to decrease inflammation and the immune response; often used in conjunction with prednisone; such as auranofin

PEMPHIGUS ERYTHEMATOSUS AND PANEPIDERMAL PUSTULAR

PEMPHIGUS

- Steroids—prednisone or prednisolone administered by mouth
- Steroids administered by application directly to the skin (known as “topical steroids”) may be sufficient in mild cases

ALTERNATIVE STEROIDS

- Use instead of prednisone, if undesirable side effects to prednisone or poor response occur
- Methylprednisolone—for pets that tolerate prednisone poorly
- Triamcinolone
- Steroid pulse therapy—methylprednisolone sodium succinate administered intravenously for 3 consecutive days to induce remission; limited application

TOPICAL STEROIDS (ADMINISTERED TO THE SKIN DIRECTLY)

- Hydrocortisone cream
- More potent topical steroids—0.1% betamethasone, fluocinolone, or 0.1% triamcinonide

MISCELLANEOUS MEDICATIONS

- Tetracycline and niacinamide

Follow-Up Care

PATIENT MONITORING

- Monitor response to therapy
- Monitor for medication side effects—routine bloodwork (complete blood count [CBC] and serum biochemistry), especially pets on high doses of steroids, chemotherapeutic drugs, or gold-salt treatment; check every 1–3 weeks, then every 1–3 months when in remission

PREVENTIONS AND AVOIDANCE

- Pet should avoid the sun, because ultraviolet (UV) light may worsen the lesions

POSSIBLE COMPLICATIONS

- Depend on type of pemphigus

- Secondary infections
- Side effects of medications may affect quality of life
- Pemphigus foliaceus and pemphigus vulgaris may be fatal, if untreated (especially pemphigus vulgaris)

EXPECTED COURSE AND PROGNOSIS

Pemphigus Foliaceus and Pemphigus Vulgaris

- Treatment with steroids and chemotherapeutic drugs and medications to decrease the immune response is needed
- Pets may require medication for life
- Monitoring is necessary
- Side effects of medications may affect quality of life
- May be fatal, if untreated (especially pemphigus vulgaris)
- Secondary infections cause morbidity and possible mortality (especially pemphigus vulgaris)

Pemphigus Erythematosus and Panepidermal Pustular Pemphigus

- Relatively benign and self-limiting
- Steroids administered by mouth eventually may be tapered to low maintenance doses; may be stopped in some pets (as directed by your pet's veterinarian)
- Skin disorder (known as “dermatosis”) develops, if untreated; generalized (systemic) signs are rare
- Prognosis fair

Key Points

- A group of diseases in which the immune system attacks the skin (known as “auto-immune dermatoses”)
- Pet should avoid the sun, because ultraviolet (UV) light may worsen lesions