

Pemphigus: The Body Under Attack

A rare immune system disorder in horses attacks the coronary band

By Derek Knottenbelt BVM&S, DVM&S, FACVSc

The body's immune system is designed to help recognize, and overcome, infections and foreign proteins that enter it, and every living creature relies heavily on this complicated mechanism for protection. The system seems to recognize "self" tissues, for example, those proteins and tissues that make up the body, and "know" they must reject foreign bodies unless they are of an identical tissue to the host.

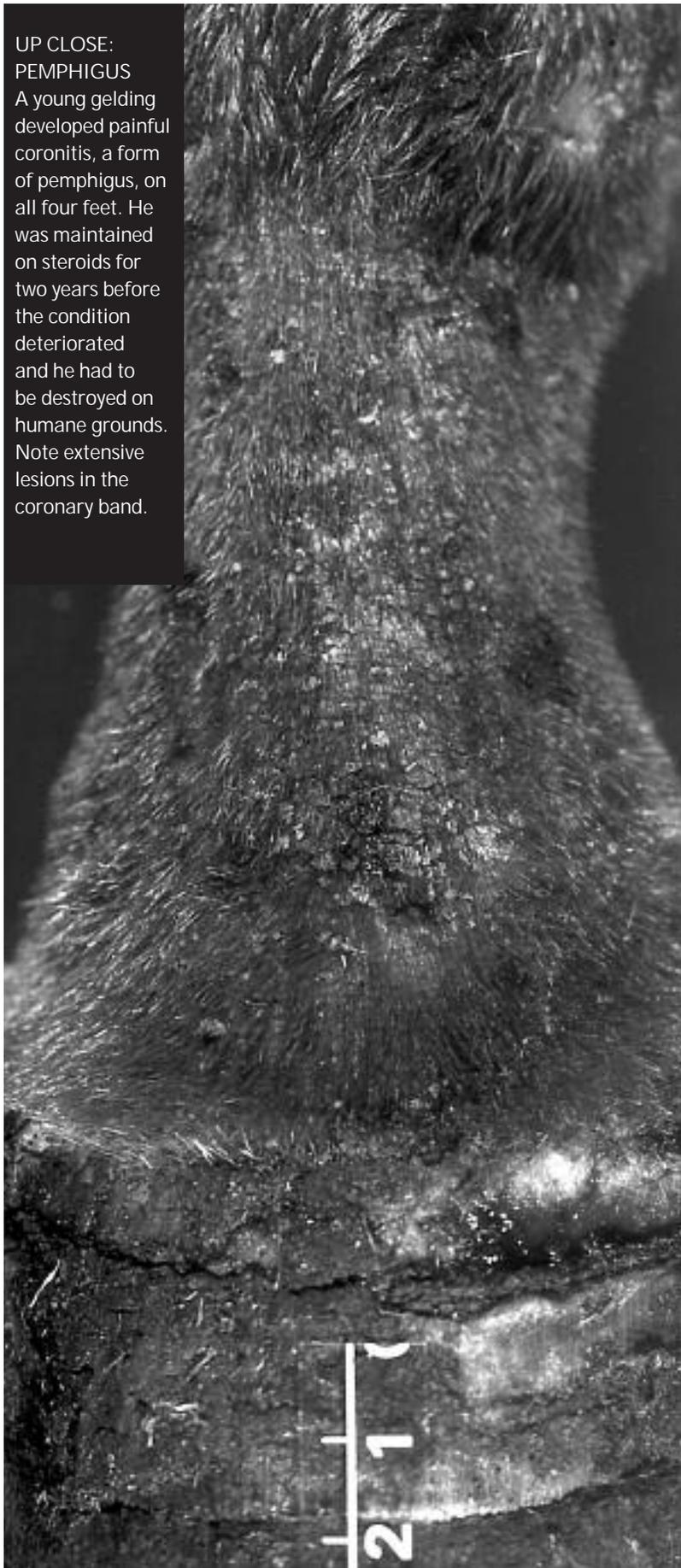
The immune system is very efficient at detecting foreign proteins but occasionally it can go wrong and the body rejects its own tissues. This malfunction can affect all tissues, but in the case of the equine disease

known as pemphigus, the rejection is targeted at the skin and the mucous membranes (superficial linings of the mouth, intestine, bladder and other tubular organs).



A horse suffering from coronitis, a very frustrating strain of pemphigus as the chances of treating it are very low, and though the horse may be sound for a long as three years, the condition often results in lameness.

UP CLOSE:
PEMPHIGUS
A young gelding developed painful coronitis, a form of pemphigus, on all four feet. He was maintained on steroids for two years before the condition deteriorated and he had to be destroyed on humane grounds. Note extensive lesions in the coronary band.



Although rare in horses, when pemphigus does strike it presents a serious challenge to the owner and vet and, invariably, threatens the life of the horse.

Because one of its most common forms includes lesions of the coronary band, the disease is of interest to farriers, as well.

Problem signs

There are two main types of pemphigus in horses. While they have slightly different features, they are largely the same. How the pemphigus lesions look depends on where the auto-antibody response occurs:

If it happens between the upper and lower layer of the skin, the disease is usually called *Pemphigus foliaceus*.

If it involves the mucous membranes and areas around the mouth, eyelids, vulva or anus it is called *Pemphigus vulgaris*.

The former is by far the most common true pemphigus condition in horses. There is probably little or no breed or color susceptibility but for some reason Arabians and Appaloosas may be over-represented on the case numbers. Pemphigus affects horses of all ages.

It is hard to understand why the body suddenly begins to develop antibodies to its own tissue and in some cases it may occur concurrently with respiratory tract infections or internal cancers.

In order to develop the disease, the immune system must either fail to recognize that the tissues of the skin are "self" or the tissue must be altered in such a way that the immune system recognizes it as foreign. In either case, once the process is instigated, it is very difficult to stop.

Recognizing pemphigus

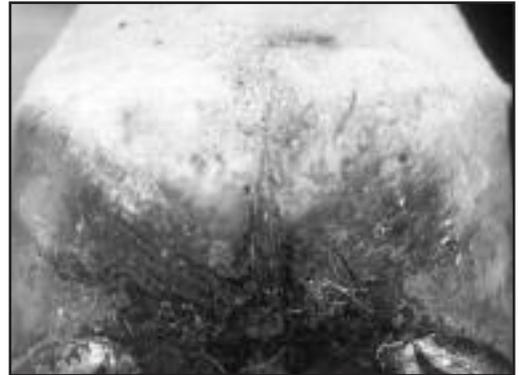
In pemphigus foliaceus, the skin develops tiny (often unapparent) blisters and then the superficial keratinised cells begin to peel off. This results in circular, or large, undefined areas of hair loss with gray, flaky skin.

Some serum may be exuded from the sites and while it normally starts over a limited area, this form will generally extend rapidly to larger areas. Interestingly, the legs may be less affected than the body, neck and head. The skin can look very inflamed but more commonly looks quieter, with extensive exfoliation of scale and crusts.

At the same time, the horse is often dull, lethargic and depressed. Weight loss can be obvious in a short time and appetite may be poor. In many severe cases, the scaling and crusting of the skin is accompanied by dramatic hair loss; even gentle stroking will often dislodge large amounts of hair. The horse may not show itchiness of the skin, as it can be the less hairy areas of the body, such as

C O R O N I T I S

The coronary band is often severely affected. Indeed, in forms of pemphigus called coronitis, the changes are limited to the coronary band and edges of the ergots and chestnuts. The coronitis form of pemphigus alters horn production and hoof quality becomes poor. The hoof wall loses its shiny periople and becomes increasingly flaky. In many cases, the legs also become filled with fluid, the skin may feel hot, and movement may be stiff and painful.



the groin, lips or face that show the severe signs first.

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Pemphigus vulgaris

In this form of the condition the earliest signs are often detected in the mouth, eyelids and nasal cavity or the regions around the nose. However, signs also show in the skin itself.

These symptoms result in severe difficulty eating and drinking, and the affected horse becomes very dull and is clearly extremely ill and in pain. Fortunately, this is very rare.

Confirming the diagnosis

A skin biopsy is usually diagnostic although a veterinarian may need to take several biopsies from different parts of the body before a typical lesion will be found. The problems are that many of the older lesions just show inflamed skin and secondary bacterial infections. Once the auto-antibody is seen or if the typical intradermal blisters are present, the diagnosis can be made with confidence.

Possible Treatments

Unfortunately, the pemphigus complex is very difficult to treat. Drugs that suppress the immune mechanisms are usually used (e. g., corticosteroids) but they have potentially harmful side effects, especially at the extremely high doses that are required to control the disease.

Horses that develop the disease early in life (less than five years) have a better chance of successful treatment than older ones; but even then, many of the young animals don't respond well to treatment or relapse as soon as the drugs are withdrawn.

There are some interesting non-steroidal drugs that can manage the disease, including a gold salts injection, but again these are very expensive and have non-desirable side effects.

Management of the coronitis group of pemphigus-like conditions is difficult. There have been many suggestions about the use of vegetable oils and hoof oils but none is likely to cure the problem, although they may delay the onset of the severe stage.

Mild blistering of the coronary band has also been suggested as a treatment, but this seems illogical for an inflammatory disorder. New drugs are being tested that might have a delaying effect on this form of pemphigus.

There are many reports of homeopathic treatment of this disease and while they seem to offer some hope, each case has to be considered on its own merits and veterinary advice is very important.

The prognosis

The outlook varies because some cases have extreme forms of the disease that don't respond to treatment at all, while others are much milder. Where a good response is obtained in a young horse, the prognosis is reasonable but relapses are to be expected.

Horses that have developed pemphigus secondarily to serious internal disease have a correspondingly lower prognosis that reflects these complications.

The coronary band forms are perhaps the most frustrating because the condition is so trivial, yet the chances of curing it are very low. Although many early cases can be managed and remain sound for some time (between one and three years), the condition usually declines gradually until the hoof quality deteriorates and secondary infections result in lameness.

Currently, pemphigus cannot be prevented. It is rare, and therefore it's virtually impossible to predict which horses will get it. Many suggested causes have been made, though, including poor nutrition, overstimulation of the immune system, such as an abscess or a tumor, and poisonings from pollution. These factors are, as yet, unproven.

Only when we understand much more about the

mechanisms that change the immune responses will we be able to focus therapy on the cause of the disease. The use of corticosteroids or even gold salts is only a broad "blanket" treatment that manages the clinical effects by altering the whole immune process. Unfortunately, the side effects of these treatments may result in laminitis or immune suppression to the extent that a horse loses all his ability to resist infection.

The trick is to find a way of controlling the unwanted immunological effects without affecting the normal protective responses that the body relies on to prevent infections.

Pemphigus is one of the most difficult diseases to manage and once diagnosed the owner can expect a disappointing outcome. Many cases either fail to respond at all, or have relapses every time treatment is reduced or have unacceptable side effects from the treatment. Set against this, though, there are some horses who have recovered fully.

Derek Knottenbelt is professor of equine medicine and surgery at the University of Liverpool in England. He is author of Diseases and Disorders of the Horse, and a noted researcher and educator. He writes extensively about sarcoids, pemphigus and other troubling equine disorders.

Environmental Injuries: Insect Infestation and Stings

THE ENVIRONMENT ISN'T JUST an onslaught of elemental conditions; horses share their environment with other species, some of whom may have an adverse effect on hooves and even a horse's life.

Consider, for instance, the mange mite, *Chorioptes bovis* (below left), which lives on the surface of the



Mange mite enlarged x70



skin under a draft or Friesian horse's "feathers". It lives on scale and "dander" from the skin. A 1987 study of horses in Holland found that 30 out of 71 horses examined were infected with mange mites, while only six showed signs of disease. Some horses may be mange mite carriers, and never be infected, but spread the mites to horses who may develop pastern sores (bottom photo) that look like common scratches. As Dr Ferraro suggests on page 25, treatment with "Frontline" for dog fleas is the suggested treatment once mange mites are diagnosed.

A bit further afield, Hoofcare correspondents who have traveled to Central America to work on horses have reported horrible conditions of hooves that locals attribute to spider bites on the coronet. While no one reports actually seeing a spider bite a horse, the aftermath of this alleged attack is a hoof-sloughing nightmare. Our latest report comes from the UK's Haydn Price, who traveled to El Salvador with the ILPH last year and called these cases "toxic trauma".

Haydn writes: "A local species of spider allegedly crawls up to the coronary band to feed on the hair and periople." Photo (top left) shows a hoof whose periople was believed to have dinner for a spider.

In some places in Central America, children are kept busy patrolling horse paddocks and killing spiders just to protect the horses' hooves. A variation of Haydn's experience in another locale is the belief that the urine of large spiders causes laminitis.

Generally, spiders may (and do) bite horses on the neck, chest, or even pastern but leave evidence of a bite with the appearance of an abscess. These Latin American spiders are still a mystery.

This is the story of Thor, a ten-year-old chestnut and white Appaloosa gelding. His story is interesting because it shows how difficult it can be to find the cause and solution to unusual problems in the horse.

My client Ruth bought Thor out of a mud lot in New York. Ordinarily Ruth would not buy a horse of any value without a purchase examination but she felt so bad for Thor in the mud lot that she wrote the owner a check and loaded him up.

When we looked at Thor shortly after purchase the most noticeable problem was thick crusts on all four coronary bands. Ruth had initially felt the crusting was due to irritation from the mud Thor was housed in, but it had not improved any since he had been moved. Thor would be visibly upset when Ruth or I pushed or pulled on the crusts. If the crusts were removed, an ulcer would be found under them. Samples of the crusts and skin were sent to the lab to check (histopathology) for any evidence of bacteria or fungi. Although bacteria were found, treatment with antibiotics did not bring any improvement in Thor's condition. Subsequently, sections of tissue were sent to Dr Danny Scott at



Cornell University; Dr. Scott authored the textbooks *Large Animal Dermatology* and *Equine Dermatology* and is a respected specialist in horse skin problems.

After microscopically examining thin sections of Thor's crust and skin, Dr. Scott felt that supplementing Thor's ration with zinc might help his problem. If the zinc did not help, he advised that we should try steroids or pentoxifyllin to see if that would help. These were all tried with only a small improvement.

Dr. John Perdrizet was called in to examine the crusts and offer a second opinion; he discovered a small fluid-filled bump in Thor's mouth. Drs. Scott and Perdrizet felt that Thor's problem could be an auto-immune disease, or a disease where the body mistakenly attacks itself at the cellular level.

Thor was started on dexamethasone which resulted in some improvement in his condition, although Ruth eventually discontinued the medication because of concern about side effects. Today, Thor is being used and doing well but still has crusting on all four coronary bands.

We suspect Thor has pemphigus foliaceus, an uncommon disease in horses. The disease seems to occur more frequently in Appaloosas and often results in crusting on the face and legs but can appear just on the coronary bands. About half the horses affected with pemphigus have signs of systemic illness, depression, weight loss or fever.

The most accurate test for this type of pemphigus is the microscopic examination of vesicles like the one Dr. Perdrizet found in Thor's mouth. Treatment for pemphigus includes dexamethasone and gold compound. Newer immunosuppressive drugs like cyclosporine may ultimately prove to be the most effective if they become affordable for use in horses. There is currently no cure for pemphigus foliaceus in the horse.

Case description provided by Kevin Sherman DVM of Brimfield, Massachusetts; photos by Dr Sherman.

