Wound care guidelines

Local skin and mucosal care in pemphigus and pemphigoid

AutoImmune Blistering Diseases (AIBDs)

This booklet is about the care needed by people with Pemphigus and Pemphigoid. It is the result of several years of exchanges, on the one hand, between the APPF and patients with autoimmune blistering diseases and their families; and, on the other, between the APPF and Professor Catherine Prost, dermatologist, co-coordinator of the National Expert Center for Blistering Toxic and Autoimmune Diseases.

The content of this booklet is informed by the questions most frequently asked by patients and their families, to which Professor Prost, Doctor Christelle Le Roux and Ms Yvonne Bautista (state registered nurse at Bobigny Avicenne Hospital, Dermatology Department) answered within the framework of an internship organized by the APPF and funded by the Groupama Foundation for Healthcare. Professor Prost, Doctor Christelle Le Roux and Ms Bautista agreed to proofread it carefully, correct the final text, and added photos to illustrate it.

Thierry Ségard, a professional photographer, and Brigitte and Christian Ducat, pharmacists, completed the illustrations. The layout and printing of this book were largely funded by the National Agency for Drug Safety (ANSM) - it should be noted that the project of the APPF was selected following a call for tender. Thank you very much to all those who made this achievement possible!

This document was highly anticipated by patients. We hope that these tips to ease everyday life will improve the well-being of patients and have a positive effect on their blistering diseases. We also hope that it will help the APPF volunteers who answer questions received by email, telephone, post, etc., as well as the relatives and caregivers of patients.

We are aware that there are other rare autoimmune blistering diseases (AIBDs) than Pemphigus and Pemphigoid. But these other AIBDs are not taken into account in this document because they are even more rare and, our financial means being limited, a choice had to be made. Furthermore, another booklet on the diagnosis and treatment of all AIBDs has been published and can be obtained from the APPF.

We wish to thank Laurence Gallu and Isabelle Nicot who translated the brochure into English and Pr Catherine Prost and Dr Christelle Le Roux who proofread it.

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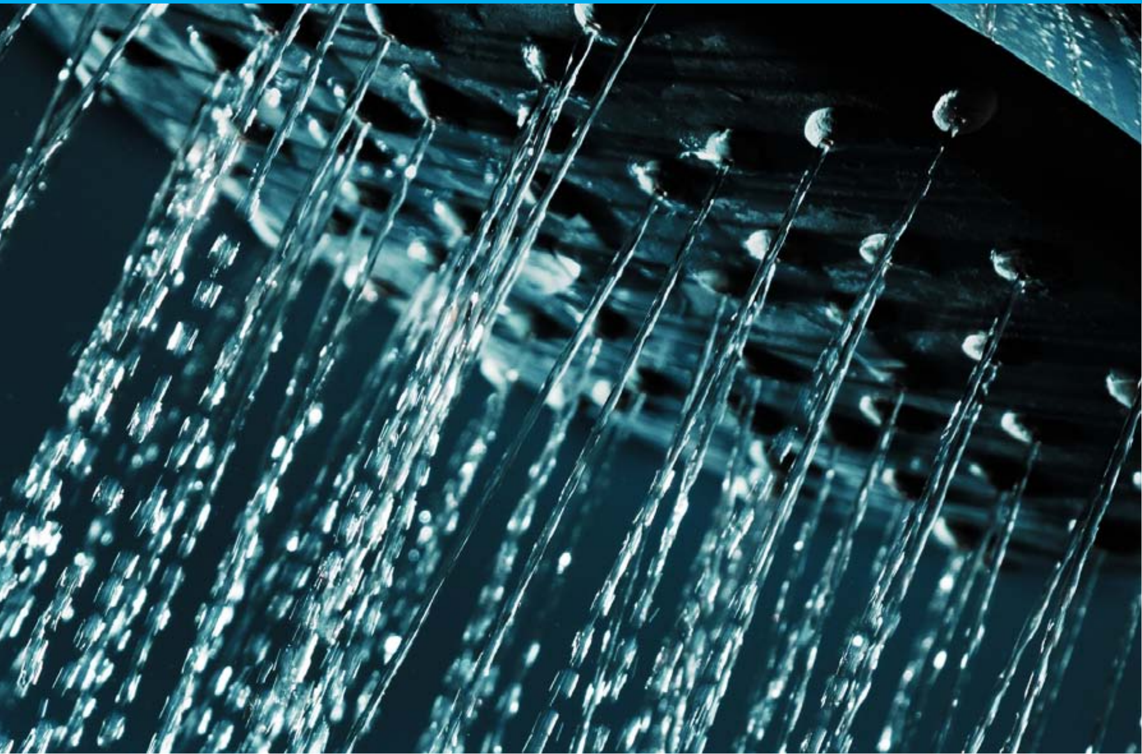


How to wash, avoid infection, dry, clean and take care of clothes and linen?

• Baths

Since gauze pads stick to skin lesions, it is difficult to remove them without causing some pain to patients. When there are numerous lesions, we recommend **a 10 to 15-minute bath so that the gauze pads can be easily taken off.**

Once the gauze pads are taken off, the best soap to use **is a soft one,** ie. a soap with a neutral pH, if possible a gentle one, like Dove, which is cheap and can be purchased as a bar or as a shower gel. Another possibility is to use Sanex shower gel, but it is less moisturizing than Dove because it is less oily. After washing, rinse with the hand shower, making sure the stream is not too strong in order to avoid causing pain.

After quick soaping and rinsing, once the skin is clean, patients can resort to **baths with starch** especially when the water is very "hard". Starch has an anti-itching effect: **it prevents the urge to scratch and leaves the skin very soft.** In practice, 2 to 3 tablespoons of starch bought in bulk should be diluted in a bowl of cold water and then poured at the bottom of the tub. This bath should last 10 to 15 minutes and patients must not rinse AFTER.

• Is there a risk in using tap water?

There are almost no germs in running water (ie. no microorganisms causing diseases), unlike stagnant water where germs do develop. Anyway, nobody lives in a sanitized environment. There is no risk of infection with bath water. The important thing is to soap and rinse well.

• Antiseptic products

They are **only used in cases of superinfections** (see the paragraph on superinfection below in the brochure). A chlorhexidine foaming solution (Septivon®, Hibiscrub® or Plurexid®...) is used directly in the bath water, not in an excessive amount – 2-3 caps (note: Hibiscrub® is not reimbursed). **Do not add any other soap.** Again, it is necessary to rinse well with water because this product dehydrates. When there are only a few blisters, an aqueous (alcohol-free) form of chlorhexidine (Diaseptyl® , for example) can be used directly on the lesion(s). Do not forget that chlorhexidine can cause skin allergies (allergic contact dermatitis) in some patients.

• Applying the soap

The soap is either put directly into the bath or applied to the lesions.
In the case of baths, the liquid soap is poured directly into the water. The soap being already in the bath, there is no need to put it on the skin and lesions again. This allows patients with numerous lesions **to avoid touching them.**
The soap can also be **applied to the lesions by hand.** The direct contact of the hands with the skin makes it possible to better detect if one is in the presence of a tense blister, a punctured one or a blister whose roof is gone. **A washcloth or a sponge** does not allow the same contact and **may tear the blister,** which is unbearable. But, this way of washing is efficient in the shower when a bath is not possible. Finally, you can also simply pour liquid soap on the skin. The correct method depends on each individual and is chosen on a case by case situation. Whatever the case may be, always rinse thoroughly, remembering that the shower stream should be neither too strong, nor too hot in order to avoid causing pain.

Avoid the following:

Do not prolong the bath beyond 10 to 15 minutes. The bath water softens the skin and beyond this amount of time, it fosters maceration.

Do not put bath salts in the water as they are irritating.

Do not put essential oils either because their exact composition is not known (even if they are listed as "Natural", there is a risk of skin allergy)... furthermore, they make the bathtub, slippery which is dangerous for the elderly as they risk getting a bone fracture.

• Drying

The patient should dry by **dabbing very gently** with a cloth, leaving the skin still a little wet. This way, the subsequent application of cream or ointment will be much easier. But, if this procedure is still too painful, the skin can be dried with a hair dryer kept far away from the skin and on low heat (not hot because heat exacerbates pain). Remember that in cases of intense pain, it is necessary to take pain medication (analgesics), about 30 minutes before any skin care takes place.

• Frequency

A bath once a day is enough, but not less. The patient can go up to two per day if needed.

• Should clothes and linen be washed with detergents?

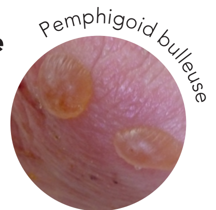
Cotton clothing is strongly recommended especially cotton underwear. **For the laundry, a conventional laundry detergent** is fine - **a liquid one** is preferable and the recommended dosage should be respected since liquid laundry detergent foams more than washing powder or tablets. **Washing at 60 °C,** which is possible with coton, is enough to destroy most germs. Boiling the laundry or using bleach is not necessary. On the other hand, washing laundry at a temperature of 30 °C, or even 40 °C (recommended for synthetics) is not sufficient to destroy germs and fungi. Fabric softeners are useless and can cause skin allergies (eczema). **Iron the laundry with steam to soften it.**

What is the appropriate local skin care before the application of corticosteroids?

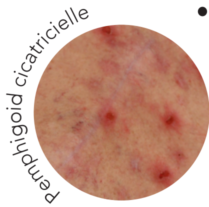
• Drying

Generally, local care is the same in all blistering diseases. However **the effect is a little different for each** disease depending on the nature of the lesions and their location (scalp, face, folds, elsewhere on the skin...). Primary lesions are always blisters filled with fluid. Subsequently, either the blisters are deliberately punctured or they burst on their own and the liquid they contain flows out.
When the blisters have burst and their roof is gone, they turn into post-blistering erosions (after blisters) that are often very painful. Finally, you should remember that care is not the same depending **if there is a superinfection or not.**

List of different lesions, according to the pathology (ie. the disease):

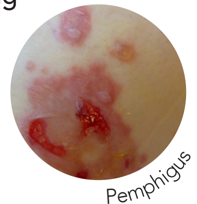


• in **Bullous Pemphigoid,** the lesions are mostly large, tight blisters, often numerous and need to be punctured; local care is therefore particularly lengthy.



• in **Cicatricial Pemphigoid,** lesions are also blisters; they are fewer in number, but break, leaving long-lasting erosions that take time to heal - therefore, they require care over a prolonged period.

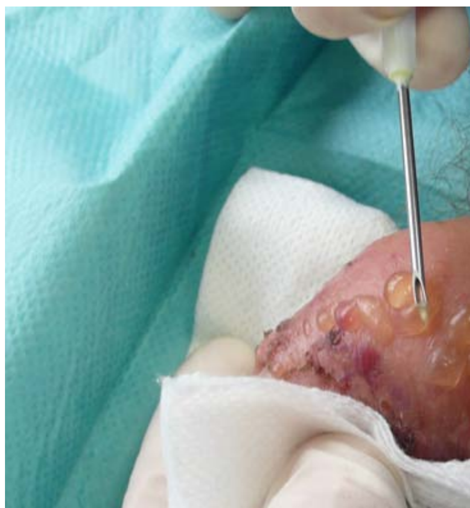
• in **Pemphigus,** the blisters are flaccid and often burst on their own because the roof of the blister is fragile.



• Bullous pemphigoid

In Bullous Pemphigoid, **the blisters are counted and then punctured as they form** and **are completely emptied of their content.**
Since blisters in bullous pemphigoid are often very numerous, puncturing them takes a long time - it is therefore imperative that patients be comfortable.
During a dermatology consultation where it is not possible to bathe or take a shower, **lesions are cleansed with 0.9% saline solution.** This is a neutral product. There are bottles of 250 mL and 500 mL. Bottles of this type should be used instead of the small archi-sterile 5 to 10 mL plastic ampoules, because when lesions are extensive, a whole bottle may be needed. First, it is necessary to place a single use "sterile drape" between the examination table and the undressed patient, so that everything that flows out of the blisters will be absorbed... saline solution and liquid.
At home, after the bath or the shower, patients settle in bed - absorbent towels should then be put between their bodies and the bed, because, in general, sheets cannot properly absorb all the liquids, especially if they are not 100% cotton.

The blisters are punctured with a large sterile needle or a scalpel blade; then, a small linear 2 to 3 mm incision is made on each side of the blister. This procedure is completely painless. On the contrary, patients often feel relief. The fluid of the blisters is then drained by pressing on them very gently with a gauze pad. After being emptied, that first blister continues to flow, while the next blister is being taken care of. Sometimes, **two minutes later, it fills up again** because **the hole** that was made **has clogged up.**



In this case, usually, when large blisters fill up very quickly, the content of the blister is drained with a needle mounted on a 10 mL syringe. This way the blister fills back up much less quickly. It is indeed frustrating to see that a blister that has already been taken care of fills up with more fluid. **The blister roof should always be left in place** to protect patients from pain (and superinfection). If, unfortunately, the roof is gone, patients experience burning sensations and / or very severe pain.

• Is the counting of blisters essential?

The counting of new blisters while puncturing them is **imperative** because it allows **to determine their rate of appearance.** For doctors, the number of **new blisters** appearing per day is a fundamental **indicator.** This is the best **criterion for monitoring the evolution of the disease.** The number of blisters appearing daily should decrease. Doctors can, then, adjust the treatment and modify it (by increasing it or by reducing it). Their outlook will not be the same if patients have several new blisters a day or only one a week. Since blisters are counted as they are punctured, they are counted only once. If blisters are not punctured, they risk being counted several times. This counting can prove to be difficult in the case of Bullous Pemphigoid when patients have 200 to 300 blisters a day - if such is the case, blisters are not counted so rigorously: a count is made before the beginning of the treatment and then at least every day. When patients are hospitalized, blisters are counted twice a day. It is easier to do this in the hospital than at home, when the nurse must pierce between 60 and 80 blisters on the same patient…

It is imperative that **the number of new blisters** counted per day be **recorded in writing,** day by day, by the nurse or the patient (on such day = so many new blisters), which means that it is necessary to keep a journal and write down the number of new blisters that appear daily.

The APPF published two booklets prepared by the AIBDs expert centers and these are kept for reference. One uses images to show how to provide care while the other provides a template to record daily the number of blisters and how many tubes of ointment were required. The APPF will send both these booklets to patients and caregivers on request.

• Is the location of blisters important?

The location of blisters is important only **for the diagnosis of the disease** (Bullous Pemphigoid, Cicatricial Pemphigoid or Pemphigus). Once the diagnosis is made, doctors no longer attach any importance to the location of blisters, because, it is the number of blisters that counts, not their location.

• What to do after puncturing bullous pemphigoid blisters?

Once the blisters are punctured and emptied of their fluid content, it is necessary **to try to "dry them out",** to prevent the formation of new liquid and its outflow. You can **use a 1% silver nitrate solution** to assist drying. It is sold in pharmacies in small tinted bottles to protect the content from light - otherwise, silver oxidation occurs. Silver nitrate solution **stains** clothes, floors, tables and even sinks if they are white, and it is very difficult to remove these black spots. Be careful when handling this product at home.

Pour the silver nitrate solution onto gauze pads to apply it. The pads, like any light surface stained with silver nitrate, will get blackish in color.

Gently dab the punctured blisters and the **lesions** with gauze pads soaked in silver nitrate.

Some patients say the silver nitrate solution burns, while others say that silver nitrate solution feels very good. The reaction varies from patient to patient, and also depends on the disease.

!

Eosin is another substance that dries broken blisters but **it should be** avoided because it reddens the skin and alters the area around the wound making it difficult to tell if superinfection has set in. It also indelibly stains clothes and linen... **Dermatologists** have never used eosin as a treatment.

• What to do after puncturing pemphigus blisters?

The care is slightly different. Pemphigus blisters often burst spontaneously. Counting new blisters therefore will not reflect the activity of the disease as much. Doctors will mainly **evaluate the surface of the lesions.** Lesions are much more painful in pemphigus than in bullous pemphigoid and it is imperative that the patient take analgesics (pain relief) before treatments.

Frequently, lesions **are covered with scabs that need to be removed.** A bath is not always enough to soften them. It is necessary to use an oily lubricant like vaseline to remove these scabs.

Finally, unlike bullous pemphigoid patients, pemphigus patients most often have difficulty **tolerating silver nitrate solution.** Be aware of the fact that silver nitrate can dry the skin too much, and result in further discomfort. In case of intolerance, do not insist on applying silver nitrate.

Local treatment

The local treatment of autoimmune blistering diseases is based on local corticosteroids.

How to choose between ointment, cream and gel ?

• Local application of a corticosteroid

Local application of corticosteroids on the skin **is always subject to medical prescription**. In no case can a nurse make the decision to apply local corticosteroids on the skin without a prescription from a doctor (a general practitioner or, preferably, a dermatologist). Local corticosteroids come in many forms - ointment, cream, gel and even shampoo.

> ! The doctor's prescription must clearly state what form should be applied. **Ointments and creams are not interchangeable.**

• Ointments

Ointments only **contain fatty substances** (vaseline in general) **in addition to corticosteroids.** Dermoval® (Clobetasol) does not exist as an ointment.

Their application feels good, better than creams, hence their benefit when being applied on lesions that are spread out and painful. On the other hand, **you cannot use them on oozing lesions** uch as post-blistering lesions, except in certain cases (see section "prevention of scabs and anal lesions"). Ointments form a kind of film, and then slowly penetrate the surface of the skin.

• Creams

Creams are **much less oily** than ointments because they contain water. They penetrate the skin faster than ointments and can be spread more easily. But be aware of the fact that you need a larger amount of cream than ointment to coat the same area. Creams can be applied on oozing lesions.

• Gels

Dermoval (Clobetasol) gel is a form of corticosteroid usually **used for scalp lesions.** It is widely used for psoriasis of the scalp. But it is not used as frequently in the particular case of blistering diseases, because **it contains alcohol and is generally poorly tolerated.** Indeed, during the acute phase, when patients have raw lesions, **applying this gel will sting.** It can nevertheless be useful because the alcohol it contains dries out the lesions more quickly. In order to choose between cream and gel, it is necessary to do a test by applying the gel on a very small area and the final choice will depend on each patient's tolerance of this application.

How to apply local corticosteroids?

Should local corticosteroids be applied to the whole body or only on red spots and lesions? What precautions should be taken when they are being applied on the face and in folds of skin?

• Local application

The local application of corticosteroids **is always done on clean skin** after the blisters have been punctured and dried, provided that there is no superinfection. In fact, **local corticosteroids** are not applied on infected lesions, generally old post-blistering lesions, because **they trigger** the proliferation of agents responsible for superinfection (see below, the section on **superinfection**). This is the only situation in which local corticosteroids should not be applied. Otherwise, local corticosteroids must be applied on all lesions, choosing between cream and ointment, depending on the type of lesions.

• Bullous Pemphigoid (PB)

In the case of Bullous Pemphigoid, **local corticosteroids** are applied almost everywhere on the skin, that is to say **all over the body, except the face** (where usually there are no lesions). Blisters and post-blistering lesions are treated, as well as the rest of the body, to prevent the appearance of new blisters. Some time ago, when patients were hospitalized, local corticosteroids were only applied on the skin in the morning; patients had no more blisters at night ... but had a big flare-up of blisters the following morning! Now, corticosteroids **are applied morning and evening** in order to prevent the morning flare-up of blisters.

• Pemphigus and pemphigoid (other than PB)

In Pemphigus and Pemphigoid other than Bullous Pemphigoid, **local corticosteroids are only applied on lesions** because they only serve **as a secondary treatment,** the main treatment being given orally (by mouth) at the same time.

• The face

Warning! In autoimmune blistering diseases other than bullous pemphigoid, there may be lesions on the face as well. **Very powerful corticosteroids** that are applied on the body such as Dermoval® (Clobetasol) or Diprosone® (fluorinated corticosteroids) **cannot be used on the face** because they create complications specific to the face. Only non-fluorinated corticosteroids such as Efficort® (which is almost as potent as Diprosone®) or lower-potency corticosteroids such as Tridesonit® or Locoid® can be applied to the face.

! Ointments should not be used in folds of the skin because sweating increases the risk of maceration - the skin softens under the ointment. So, where folds are concerned, a local corticosteroid cream should be used instead. However, you should be careful not to apply too much cream: since the folds create an obstruction, closing up, and retaining the cream, which boosts local corticosteroid permeation.

"Infected" or "superinfected" lesions?

How can you tell that lesions are superinfected? What is the difference between "infected" and "superinfected"?

• The "blister"

The blister is the **primary lesion** in autoimmune blistering diseases and **is never infected.** In all autoimmune blistering diseases, the newly formed blister **contains a clear, odorless liquid (plasma) which is characteristic of the absence of infection.**

• Pustules and skin infections

When the fluid collection is cloudy, usually this means the fluid is pus containing altered white blood cells and, in this case, they are not called blisters but pustules. Diseases yielding pustules are not autoimmune blistering diseases. Most often these are primary infectious skin diseases such as erysipelas, boils ... Fever blisters due to the herpes virus are also primary infectious lesions. The term **''skin infectio"** is the term used for these primary skin diseases.

• Superinfection

Primary lesions resulting from blistering diseases, i.e. **blisters** or more often **post-blistering erosions, can** in a second phase **be invaded by infectious agents** (bacteria, viruses); we then talk about superinfection. This will modify the aspect of primary lesions (see below). Most frequently, this superinfection is **caused by a staphylococcus aureus** but in pemphigus cases, it can also often be **a herpes superinfection.**

Superinfection can spread from lesion to lesion if patients scratch themselves with germ-carrying fingers; it can also spread from patient to patient (hence the importance of reinforcing hand hygiene).

• Odors

Another sign of superinfection, although rare, is **the occurrence of bad smells.** This can be unbearable for patients: they can no longer stand themselves, and wonder how their families, friends and colleagues can put up with this.

The liquid in the blister is ''normall" sterile and has no odor. The emergence of bad smells is a sign of **anaerobic superinfection** (i.e, due to germs growing without oxygen) especially when these occur in the mouth. Anaerobic germs are always suspected when a particularly bad smell occurs.

On the skin, superinfection with pseudomonas aeruginosa is also responsible for nasty smells.

In the mouth, foul smells can also be the consequence of **an oral hygiene problem;** patients have so much pain that they do not brush their teeth anymore ... and this is not taking into account possible dental cavities, abscesses and so on...

Some patients are in so much pain that they do not drink anymore and dry mouth aggravates these smells (healthy people notice this same dryness in the morning, even though they have brushed their teeth the night before).

• Pain

Pain experienced by patients is a sign of **a possible superinfection.** Therefore, it is necessary to listen attentively to patients when they say it is not like before. This might indicate that something unusual is happening. **This pain** can be very **severe** in herpetic superinfections of pemphigus lesions.

• Changing of appearance in lesions

Another sign that patients do not assess as easily as pain, **is the change of appearance in their lesions.** Some patients notice themselves that something weird has happened: **a dark red halo** (different from the itchy red patches in bullous pemphigoid) **appears around the wound.** Most often, nurses are the ones who notice this change when they are taking care of the bandages. Nurses and doctors exchange information on a daily basis: they discuss symptoms, what they find weird, what they do not understand ... In herpetic superinfection of pemphigus lesions, small vesicles appear on the periphery of the lesions. **This situation,** which is quite common, **can be mistaken for an aggravation of pemphigus** and wrongly results in a more forceful continuation of treatment. It is therefore necessary to be aware of this and remember it in the case of pemphigus. It is also necessary, if possible, to take virological samples and, most importantly, give an anti-herpetic treatment.

• Fever

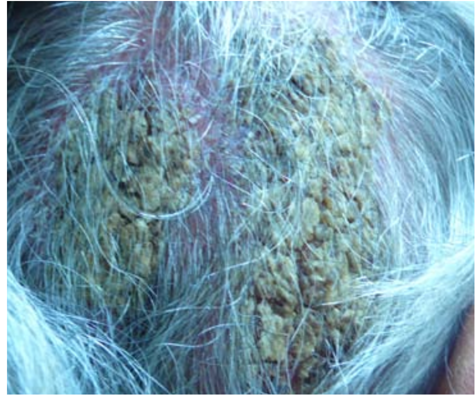
Finally, it is necessary to know how to use a thermometer, because **fever is a reliable sign of infection.** For patients taking corticosteroids, especially those with pemphigus, **the fever does not rise much.** However **a temperature of 38 °C is a sign of a little more serious infection or superinfection.** At the hospital, measuring patients' temperatures has become almost routine and is done in the ear as it is the most accurate method. The normal temperature in the ear is 37 °C. Mercury thermometers are now prohibited, even though they were probably more accurate.

• Samples

Bacteriological or virological samples may be necessary. When a doctor's recommendation for treatment of superinfected lesions have been followed but the patient's condition still doesn't rapidly improve, samples need to be taken to see if the origin is a virus or a multi-resistant germ, which would then require a specific treatment. For instance, pemphigus patients are often superinfected by herpes, but antibiotics are ineffective on the herpes virus. You have to administer an anti-herpes treatment. However, other blistering diseases are more often infected by staphylococcus aureus. There are relatively benign staphylococcus aureus, and others that are very nasty, which are multi-resistant. If a very nasty multi-resistant staphylococcus aureus is discovered, indications given previously about linen become even more important. Drastic measures must be taken to ensure good hygiene: rigorous cleaning and decontamination of all linen to eliminate germs as well as strict methods of personal hygiene both before and after treatment. Effective antibiotic treatments on these germs can generally only be administered intravenously.

What to do when there are scabs?

To minimize pain and bleeding, scabs **must be removed once they have been softened** either by a bath or by the use of an oily substance. It is essential **to avoid the formation of scabs,** because they foster the proliferation and multiplication of germs in the case of superinfections and slow down the wound healing process. It is commonly thought that «when there is a scab, it is better to leave it». A scab is a nest of germs and an obstacle to wound healing.



• Moist scabs

If the scabs are «wet», a bath is best for **softening them.**

• Dry scabs

If the scabs are dry, **they can be ''greased''** with Vaseline or Fucidin® or an ointment which is left on for 15 to 20 minutes, sometimes half an hour. These substances are very well tolerated by patients because they are very oily. You can also use Flamazine®, a silver sulfadiazine cream, which is a cream (and therefore less greasy than the Fucidin® ointment); it contains a silver salt which is a very powerful antiseptic. When the lesions are a bit painful, patients report quick relief after applying it. **Once softened,** the scabs yield quite easily, which allows the caregiver **to see what is happening underneath.** There are sometimes big surprises after the removal of scabs. After lifting them, **the presence of a yellow or green liquid,** sometimes smelly, **can indicate a superinfection.** This might have already been suspected if the patient had complained of pain. The most recent recommendations of the High Authority of Health (HAS) go a bit against the use of Fucidin®. This medication exists as a cream and as an ointment but it also exists in the form of tablets that are taken orally. However if patients develop an allergy to Fucidin® after local application of the cream or ointment and, therefore, decide to take the oral tablets later, there can be very serious accidents: in general, all substances (especially antibiotics) that may be allergenic and can be taken orally should not be used locally.

• Scabs in the hair

What to do with scabs in the hair? Scabs on the scalp are particularly **difficult to treat.** Again, it is recommended to apply a greasy substance (Vaseline, Diprosone® ointment, Fucidin® ointment) or Flamazine®, followed by **a mild shampoo** (with Clobex®, a corticosteroid shampoo), **if the patient tolerates it.** Otherwise ,scabs are carefully removed manually and with great care after having given the patient an analgesic treatment (against pain, see below).

!

Preventing the appearance of scabs

It is very important to prevent the appearance of scabs. These are formed when the erosions are exposed to the air, especially if there is a superinfection. To prevent scabs from appearing, it is imperative to cover the erosions with a bandage (see next page).

Which wound care dressing to use?

Once blisters have been punctured, drained and counted, and corticosteroids rubbed on the skin, **blisters and wounds** that have lost their outer membrane (the blister roof) **must be covered** in order to prevent scabs, but also to alleviate the pain, because wounds left to dry uncovered are painful and may "burn".

• Non-stick pads

The first step for nurses is to apply **a non-stick** pad that will not adhere to the wound.

There are two options:

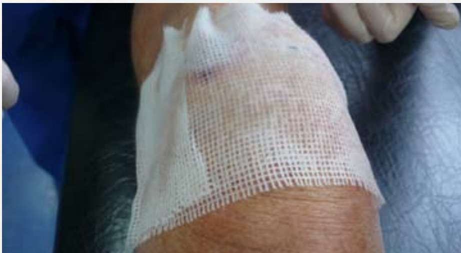
The first is to use **vaseline gauze dressings,** like Jelonet®. These dressings are very moist; they are made of open weave gauze impregnated with vaseline, which is an inert substance, unlike the ones used on tulle bandages, which are not sold anymore due to the allergic reactions they caused.
Jelonet® open weave gauze is non-occlusive and **allows the wound to "breathe",** preventing tissue maceration.
These pads come in 2 sizes (5 x 5 cm or 10 x 10 cm).
They are hard to cut but can be bent before being applied. They lose their properties over time and removing them can be painful. They are known as "conventional" (i.e traditional) dressings.

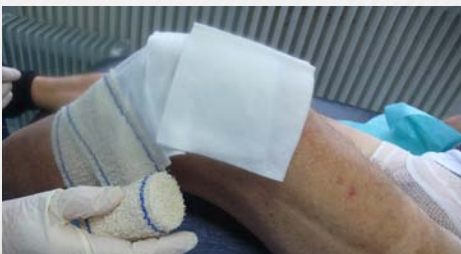
The second possibility is to choose **interface** (non-adherent) **dressings,** like Mepitel® or Urgotul®, Altreet® , Physiotulle® and Hydrotul®. They are known as advanced (or "modern") wound care dressings and are more expensive than traditional dressings. They are, however, specifically recommended by the French National Authority for Health (HAS) to care for wounds caused by blistering diseases (epidermolysis bullosa) (order dated 16 July 2010). They differ from simple tulle dressings on account of their low adherence, which does not increase over time and minimizes trauma and pain on removal. Mepitel® is available in a variety of sizes and can also be cut to fit any size wound. It should be emphasized that **bandages used for ulcers or pressure ulcers are absolutely not suitable for wound care in bullous diseases.**

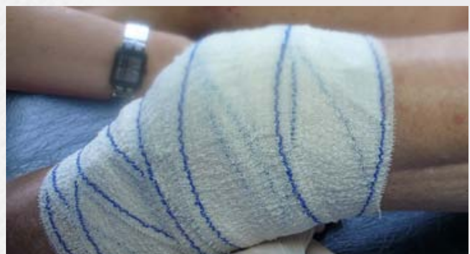
• Gauze pads and bandages

Nurses then apply **a sterile gauze pad** (reimbursed by the Social Security System) on top of the dressing **and then a bandage or an elastic tubular stretch net.**

It is recommended to use wrap bandages, even though they are not very glamorous. Patients treated with corticosteroids have very fragile skin that bruises easily. The wrap bandage provides cushioning: it protects the wounds and avoids trauma.







Elastic stretch netting exists in a variety of sizes. The gauze is applied first, then the elastic tubular netting to keep the gauze in place. On the legs, an elastic stretch netting can be added on top of the wrap bandage to prevent it from moving during the night. By doing this, the pad underneath is secured and does not move. Punctured blisters or wounds are already painful; a loose wrap bandage increases the pain. Conversely, a firmly wrapped bandage is far more comfortable.

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Warning: no adhesive dressings

All dressing products must be non-adherent. The following must be avoided: tape and transparent film dressings like Tegaderm®, and hydrocolloids dressings… Patients suffering from blistering diseases have extremely fragile skin that can be easily torn. When these types of dressings are being removed, they stick to the skin, causing additional wounds. Patients who have had bad experience with such dressings do not mind wrap bandages.

The difficulty of getting nursing home care

It is not easy to provide wound nursing care for nursing home residents in long-term care or for stay-at-home patients. In long-term hospitals or nursing home facilities, where most of very old patients with bullous pemphigoid stay, there is not enough staff on duty and they lack the time. The **relapse** of bullous pemphigoid is, therefore, almost automatic **because the cream or ointment has not been applied correctly,** i.e only on the blisters and not on the whole body… or, **even worse, it has been applied everywhere, except on the blisters.**

For stay-at-home patients, **the official rate for nursing care does not really take into account the time necessary to provide proper nursing care** to the patient. **A standard wound care procedure** (taking an estimated time of 10 minutes) **is registered under AMI2,** while **a delicate and complex wound care procedure is listed under AMI4;** when, in fact, the nurse might need an hour to do it. AMI means Acte Médico-Infirmier (Medical and Nursing Procedure). The rate fixed by the Health Insurance Authority has not changed since 2012. A nurse gets paid € 3.15 for an AMI. An AMI 2 is € 6.30... and an AMI 4 is € 12.60. That is the reason why visiting nurses sometimes refuse to take care of these patients. Patients who have leg ulcers have the same problems: they cannot get wound care from private nurses because the treatment time is too long
At the end of the day, those patients often end up in hospital in a terrible condition, with very strong wound odor resulting from superinfection.
To prevent this from happening to patients with autoimmune blistering diseases, hospitals tend not to discharge them if they still have many blisters (they are let to stay longer) and the prescriptions are customized to fit their lifestyle.

• How to adapt the prescription in order to make sure proper care is provided?

• In nursing home facilities

It is essential that nurses get help from a nursing assistant to turn and sit those elderly patients who no longer have the ability to move. For the well-being of the patient, the help of a second person is essential to carry out the care and to pass the material, the gauze pads, the ointment.

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This is why it is imperative to ask the prescribing doctor to specify "care by a nurse + a care assistant" on the prescription.

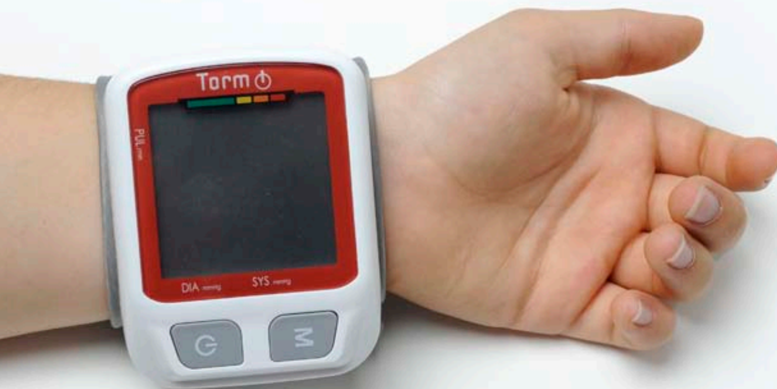
• At home care

The prescription must state: **«particular and complex wound dressing».** Being reimbursed on the same basis as burns on more than 5% of the body, **prior agreement from the social security system is required.** In case of emergency, wound care can be started before obtaining this agreement. **The nurse can apply an excess charge** in case of a particularly long procedure. The reimbursement of the excess charge **will depend on each voluntary insurance company's terms.**
See "Extract from the list of nursing procedures" on page 29.

Are ointments and creams containing corticosteroids absorbed into the bloodstream?

• The answer is yes

If local corticosteroids are used, the medication is always absorbed into the bloodstream. The higher the number of erosions, the greater the risk. The absorption of corticosteroids into the blood is higher where the erosions are located, the skin being no longer there to act as a barrier. This is why it is recommended to **monitor blood pressure and blood glucose** (i.e the blood sugar level that rises in case of diabetes) of patients who have a lot of lesions.



Nevertheless, the level of corticosteroids in the blood after using topical corticosteroids on normal or damaged skin is still much lower than with oral corticosteroid therapy. There is no comparison whatsoever. At the end of the day, the benefit of local corticosteroids is much higher than their potential risks.

• Salt-free diet?

Should patients treated with potent local **corticosteroids restrict their salt intake** in the same way as patients who take oral corticosteroids? The answer is no: the level of corticosteroids in the bloodstream is extremely low.

• Are local corticosteroids safe for the fetus?

Do patients with **pemphigoid gestationis** (autoimmune bullous dermatosis of pregnancy) who receive **corticosteroids** during their pregnancy **put their babies at risk?**

The answer is no, for two reasons. First, absorption into the bloodstream through the skin is very low. Second, to reach the baby's blood, corticosteroids would have to cross a second barrier, ie the placenta.

To sum it up, the amount of corticosteroids getting into the baby's blood after crossing the maternal skin barrier and the placental barrier is absolutely minimal.

• Should the ointment be applied wearing gloves?

The answer is yes. One **should always wear disposable gloves** when applying topical corticosteroids. Anyone who has to apply corticosteroid cream on patients must follow the same rule, because **repeated applications** can amount to significant levels of corticosteroids in the bloodstream of care-givers. **Slightly dampen and heat** the gloves before using them.

To do this, simply put on the gloves and run warm tap water over your hands, or soak them in a bowl of warm water. Doing this and applying local corticosteroids to the patient's skin immediately after a bath or a shower, when the skin is still damp, saves product and makes it easier to spread the ointment. It is also much more comfortable for the patient.

How long does the local treatment last?

It depends on the disease:

• Bullous pemphigoid

As for Bullous Pemphigoid, for which no treatment other than local corticosteroids is generally given, **the applications of local corticosteroids** must be **continued 15 days after the appearance of the last blisters.** They are then spaced (1 day out of 2, then 2 times a week, then 1 time per week) according to medical prescription. Only the doctor can decide to stop or continue the treatment. **If the treatment is stopped too early and too quickly, patients relapse quickly.** Whether methotrexate can help stop topical corticosteroids faster, is currently under evaluation.

• Cicatricial pemphigoid and Pemphigus

As for mucous membrane pemphigoid (cicatricial pemphigoid) and Pemphigus, **local corticosteroids are stopped when there are no more lesions,** since the systemic (i.e oral) treatment is given to prevent the appearance of new lesions.

Special cases

What to do when **the disease affects the mouth?**

Pemphigoid and pemphigus vulgaris often cause lesions in the mouth. Lesions in the oral cavity can appear on the gum tissue, inside the cheeks, the palate, the lips.
They can be painful and can sometimes prevent patients from eating normally.
Like blisters on the skin, those lesions require adequate treatment: an antiseptic mouthwash, corticosteroids applied locally, a change in eating habits until the systemic treatment is effective. Proper teeth and gum care is also essential.

• Mouthwashes

Antiseptic mouthwashes must not contain harsh ingredients. It is highly recommended that the mouthwash be prepared by the pharmacist **(compounded drug or customized preparation,** reimbursable, provided by the pharmacist for a particular patient and prescribed by a doctor). The preparation is a mixture of Eludril® + Mycostatin® + baking soda - Lidocain can be added to the preparation to decrease the pain. It must be kept refrigerated. If a customized preparation is not available, Alodont® can be used instead (not reimbursable).
The customized preparation contains Mycostatin®, **an antifungal agent,** in order **to avoid fungal infections** that can develop in the mouth following the use of corticosteroids. This is why doctors seldom have to prescribe an antifungal systemic drug.

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Warning! Resin dentures do not last forever. The material can become porous and bacteria and fungi can grow on the denture. Dentures should be checked by a dentist on a regular basis.

• Local corticosteroids

Applied in the mouth, they generally help **reduce pain** and accelerate the healing of lesions. It can be applied in two different ways, either using a mouthwash, or an adhesive paste.

The drug **Celestene®** is often used as a mouthwash, its benefit being that it is highly concentrated: 2mg of Celestene® are the equivalent of 20 mg of Cortancyl®. The prescription should state that the mouthwash is meant to be spat out, **not swallowed.**

The adhesive paste is a preparation consisting of Diprolene® ointment (a corticosteroid ointment as potent as Dermoval®, only available as a cream) in a 50:50 mix with Orabase®, which is a substance that adheres to the gums and the inside of cheeks for about half an hour (while a mouthwash will be effective for 5 minutes at most). This is particularly helpful for patients with fairly localized lesions.

Dermoval® can be applied **on the lips** once a day and the lips can be **moisturized throughout the day with Vaseline** or a fragrance-free lip balm.

• Change in eating habits

In case of mouth pain, patients should **adjust their diets:** eat warm, soft, low-acid or no-acid foods. In case of severe pain and on medical prescription only, patients can take an anaesthetic: either liquid Xylocaine® (Lidocain) mixed with mouthwash, or Xylocaine Viscous® (or Dynexan®) applied directly on the lesions. In that case, it is essential to eat very slowly because without sensation, food may be swallowed the wrong way.

• Brushing your teeth

Whenever possible, patients **should brush their teeth,** even if it causes bleeding. Again, local anesthetics can be applied before brushing (Dynexan® or Xylocaine viscous®). The use of **a surgical toothbrush or an Inava Sensitivity toothbrush** for example (with cone-shaped extra flexible bristles) is recommended. **Waxed dental floss** can be useful for removing dental plaque that forms between teeth. Air-powered water flossers are often too strong and can cause pain. Standard toothpastes are generally poorly tolerated. ''Mild'' toothpastes such as La Vie Claire Clay sage toothpaste, or Wéléda Ratanhia Toothpaste should be preferred. Good oral hygiene helps to prevent bad breath.

• Dentists

Dentists often **refuse to treat patients** with a blistering disease in the mouth because they are not familiar with those diseases. However, **regular dental scaling and dental care is important.** The general dentist should talk to the patient's dermatologist or stomatologist to know more about the patient's disease.

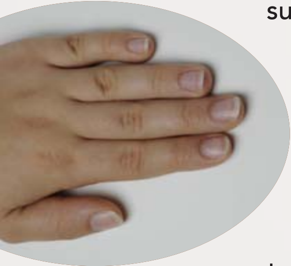
• Dental implants

Oral lesions caused by an autoimmune blistering disease **do not contraindicate, per se, dental implants.** However, **precautions should be taken** if the patient takes or has taken bi-phosphonates (Actonel®, Fosamax® ... prescribed to fix calcium while taking oral corticosteroids).

What to do when other areas are affected: nails, nose, genitals, anus...?

Blistering diseases rarely affect the nails, nose, external genitalia or anus.

• What to do if the nails are affected?



Patients should **soak fingers in an antiseptic solution** to prevent superinfection. Soak for 10 to 15 minutes and rinse well afterwards. **Do not touch the nail and apply topical corticosteroids** covered with a large occlusive dressing. Do not rub, because it is extremely painful. Do not hesitate to ask your doctor for analgesics if necessary.

Sometimes the nails fall off. Re-growth depends on the matrix of the nail. In general, it is not affected in pemphigus and a normal nail regrows. On the other hand, it is frequently affected in epidermolysis bullosa acquired (another AIBD) and the nail may not grow back.

• What to do in case of nose bleed and crusting?

All blistering diseases affecting mucous membranes **make the nasal membrane very thin** and bleed at the slightest touch (atrophic rhinitis). Patients often experience nasal discharge of bloody mucous followed by a sore that scabs over and forms a crust. This might be 'a scar" of the disease and not necessarily a sign that the disease is still active. **Vascularization of the nose is high.** That explains why it bleeds easily.

The ENT (Ears, Nose and Throat) **specialist** will direct the treatment. As for blisters elsewhere on the body caused by bullous diseases, it is necessary to disinfect, **apply local corticosteroids and moisturize** in case of scabs.

Nasal irrigation is very useful, especially when the nose is clogged. Sea-based products are marketed, but nose washes can be done simply with a saline solution, possibly supplemented with hydrogen peroxide. Nasal washing requires a large volume of fluid to be effective. The procedure is not painful, but tedious, for the patients. They must lay on their backs, inject the water into the nose through the nostrils, lean the head back, let the serum sink down the throat and then spit it out. The procedure has to be repeated several times. When the nose washing is administered by another person, the patient may swallow the wrong way. Local **corticosteroids** such as **Rhinocort®** are usually used in the nose for active lesions. They can be used as a long-term therapy and prevent crusting.

In case of nose scabs, **Balsamorhinol®** may be prescribed **for a short period of time.** It cannot be used long term as it can lead to lung disease. Another possibility is to put a little **vaseline** in the nostrils after washing with a saline solution, at bedtime. But the nose function is not only to detect smells. It also purifies the air we breathe (eliminating dust, allergens) and warms it. When the nose is clogged with vaseline and we breathe through the mouth, we inhale cold air that arrives directly into the throat without having been filtered and warmed by the nose. Many sore throats are caused by breathing through the mouth when the nose is clogged. It is therefore **not possible to sleep with Vaseline in the nose, on a regular basis.**

• Lesions on the anal mucosa (on the anus)

Very often people think that the discomfort they feel in the anus area is related to hemorrhoids and they do not talk to the doctor. Yet, the doctor must look because there may be hemorrhoids... **as well as pemphigus lesions on the inside of these hemorrhoids, or post-bullous erosions without hemorrhoids.** The auto-antibodies can be deposited there because this area's structure is the same as skin's. It is estimated that 20% of patients have lesions there. The anus is the last area to heal because it is constantly traumatized by passing stool. Proctologists are the specialists who examine the anus and the anal canal.

Local corticosteroids ointments (like Diprosone® for instance) are used to help the passage of stool. The application is done with one finger covered with a lubricated glove. You must insert a phalanx (2-3 cm) covered with ointment in the anal canal.

It is necessary to **avoid constipation** and to keep stools soft to minimize trauma to the tissues which aggravate the lesions. It is therefore recommended to drink a lot, to eat a high-fiber diet, or even to take an osmotic laxative like Forlax®, or a ballast and lubricant laxative, like Parapsyllium.

• Genital lesions

External genitals (penis, vulva) should be thoroughly washed in the bath or in the shower, especially in between skin folds. The **local corticosteroids** used for these areas are **creams** (not ointments), to be applied **in a thin layer** and in all the folds. Women who receive local corticosteroids on the vulva can suffer from vaginal mycosis and cystitis (urinary tract infections) caused by the treatment.

Very disturbing symptoms: Itching and Pain

• Itching

What to do about the unbearable itching (pruritus) of Bullous Pemphigoid and Superficial Pemphigus?

Bullous pemphigoid and sometimes superficial pemphigus can itch unbearably before blisters appear. Some patients cannot sleep. Others scratch themselves to the point of bleeding.

There is no effective local treatment for pruritus other than the local corticosteroids prescribed by the doctor.

Once the local corticosteroids are started, the itching disappears within a few days and patients do not scratch themselves anymore. When the itching reappears, it announces a relapse.

While waiting for the local corticosteroids to work, **ice** may anaesthetize the pathways to this itching sensation (use ice cubes: they should not be applied directly on the skin, but wrapped in a cloth).

Some doctors prescribe sedative **antihistamines** (they have a soothing effect), such as Atarax®, to reduce the itching.

Finally, when patients have severe itching that prevents them from sleeping, the doctor can resort to **a sedative**, like Stilnox® (fast-acting and effective for a short period of time).

• Pain

Pain management is a priority for public authorities. It must be implemented without waiting for specific disease treatments to be effective. Of all bullous diseases, Pemphigus is the one that causes the most severe pain, with patients complaining mainly of burning sensations. This is caused by the stimulation of the small nerves in the epidermis. Pain entails lots of negative feelings for patients: low morale and difficulties improving their conditions. Patients with blisters all over their bodies see their self-esteem deteriorate. Pain should be acknowledged and taken care of.

When patients are hospitalized, nurses must ask them everyday to describe the pain they feel and offer increasingly stronger analgesics, if necessary. The treatment should always start with simple analgesics, paracetamol or acetaminophen. If they are not strong enough and do not work, they are replaced by more potent medications (Topalgic®, tramadol,...). Sometimes the doctor may need to prescribe morphine, on an ad hoc basis.

Analgesics should also be administered routinely **half an hour before wound care,** following the same procedure. All stages of care can be painful, including the use of local corticosteroids, which can increase the burning sensation in lesions. It should be noted that Dermoval® gel, which is often prescribed for scalp lesions, contains alcohol and is particularly not suitable in case of erosions.

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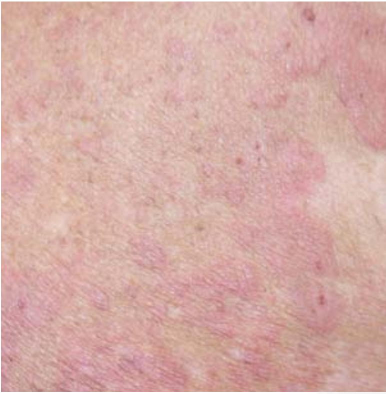
Warning: the use of EMLA®, must be avoided. Emla® is a local anesthetic, available as a patch, widely used with children before procedures like vaccination or taking blood samples. It is also used before cleaning leg ulcers and performing biopsies.

Microscopy of biopsies performed after application of EMLA® showed that it causes lesions, "holes" in the tissues, which make biopsies impossible to interpret. Erroneous diagnoses occurred (in children at Necker Hospital) until it was understood that the diagnostic errors were due to the application of EMLA®. It can, therefore, be assumed that applying EMLA® to a wound before wound care is likely to delay the healing process.

We have seen previously that good local care is the best way to reduce patients' pain with long lasting effects. It should be remembered that after a bath or a shower the scabs should be gently removed and the lesions quickly covered with a moist dressing or an interface dressing because contact with the air or clothing makes them painful.

Wound healing

• Repair of the epidermis



The wound healing process in blistering diseases is more repair than regeneration of the epidermis. Local corticosteroids slow down the healing process a little, mainly because they affect the dermis cells; as a consequence, those cells produce in lesser quantities the substances needed to heal. In the particular case of blistering diseases, wound healing can only happen if the inflammatory process is over -- i.e. only if corticosteroids are applied. The way post-bullous erosions heal has nothing to do with the way burns or other wounds do. The management of accidental injuries and wounds caused by bullous diseases is very different.
The only contra-indication for corticosteroids is obvious superinfection. In the absence of superinfection, corticosteroids should be rubbed onto the wounds.

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Scabs keep the wounds from healing.
Ideally, they should be moistened and then removed. If scabs are left in place, erosions will not heal.

• What to apply to help the healing process?

No cream or ointment has demonstrated its effectiveness to speed up healing. Doctors can prescribe moisturizing creams to moisturize the skin of the elderly which is often dry. A large range of moisturizers is available in pharmacies or drugstores. As a matter of fact, patients choose the moisturizers they like best, preferably a neutral one (fragrance-free), to avoid any risks of allergies.

• Is it safe to swim in a pool while wounds are healing?

You'd rather not, out of consideration for other pool users. Interestingly, there are few cases of superinfection of bullous wounds due to swimming pool fungi.

• Is it safe to swim in seawater?

The short answer is no. Saltwater in a wound can be a little painful and can damage tissues. Wounds will take longer to heal... but the choice is yours. If a teenager enjoys a swim in the sea and if, therefore, it takes him 15 more days to heal, it is no catastrophe. Just make sure that you rinse off thoroughly after swimming.

• Is sun exposure safe?

It is preferable to avoid the sun because it makes brown or black pigmented scars even darker, which is not very pretty. In addition, pemphigus and bullous pemphigoid are sometimes worsened by exposure to the sun.

• What to do about stretch marks, bruises...?

These are the side effects of local corticosteroids about which, unfortunately, little can be done. To minimize stretch marks, it is necessary to moisturize the skin with creams. For bruises and wounds, it is essential to avoid skin trauma (shocks, blows) which can cause wounds. Do not hesitate to protect the skin with thick and padded clothes. Remember that no adhesive bandage should be applied in direct contact with the skin, ever, because there is a risk of tearing the skin when removing it to change the dressing.

Extract from the list of nursing care procedures

Conventional values of key-letters as of 27-05-2012

AMI (Acte Médico-Infirmier: Medical and Nursing Procedure) = 3.15 € for Metropolitan France and 3.30 € for Overseas France.
IFD (Indemnité Forfaitaire de Déplacement, flat rate travel reimbursement) = 2.50 € for Metropolitan France and 2.70 € for Overseas France.

Article 2: standard wound care procedure AMI2
Removal of sutures or staples, ten or less, including the wound care associated if need be = AMI2
Removal of sutures or staples, more than ten, including the wound care associated if need be = AMI4

Article 3: Long and complex wound care procedures
Long and complex wound care procedures requiring strict aseptic techniques: wound care of large burns, chemical or thermal wounds, on more than 5% of the body surface area = AMI4

Excess charge: The nurse can apply an excess charge in the two following situations:
1 - Special circumstances of time or place due to a particular requirement of the patient (DE)
2 - When the visit has not been prescribed (DD).
The nurse states the reason for the excess charge on the medical claim form (feuille de soins) (DE or DD) and informs the affiliated patient at the beginning of the treatment. When applicable (see above), nurses set a reasonable fee and state the amount paid for on the medical claim form.

Reimbursement "by assimilation": When a patient with an unusual pathology needs a procedure not included in the nomenclature of procedures, the exceptional procedure can be assimilated to an existing one of the same importance listed in the nomenclature and, consequently, the same coefficient applies. The reimbursement of this procedure is subject to the approval of the social security doctor who performs the medical examination of the patient, and to the fulfillment of the formalities of the prior agreement, as indicated in Article 7. However, the absence of a response from the social security system within three weeks must be considered as a tacit refusal of the request for assimilation.

Article 7: Prior agreement. The social security will only contribute to the costs of certain procedures if, after authorization from their medical adviser, they have previously agreed to reimbursement, provided that the insured patient fulfills the legal requirements for benefits. (...) When prior approval is necessary, patients must send the social security doctor a request for prior approval duly filled in and signed by the practitioner who will perform the procedure. If the procedure is to be performed by a medical auxiliary, the request must include the medical prescription that prescribed the procedure, or a copy of that prescription. (...)

The postmark date certifies the date the request for prior approval was sent. The Social Security must reply to the patient or to the practitioner, if applicable, no later than ten days after the form has been sent. If there is no answer by that time, it is assumed the answer is positive. In that case, the insurance medical officer can always decide for or against the continuation of the treatment or procedures. Where there is an obvious urgency, the practitioner carries out the procedure but nevertheless complies with the formality indicated above, writing the words ''act of urgency". (...)

You will find the complete nomenclature on the website of private nurses: www.l-idel.fr

(Généralités / pratiques professionnelles / la nomenclature des actes - General / professional practices / nomenclature of procedures).

ASSOCIATION PEMPHIGUS PEMPHIGOÏDE FRANCE
22 rue des Boulangers 75005 PARIS
www.pemphigus.asso.fr
pemphigus.asso.77@laposte.fr

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