



HAILEY-HAILEY DISEASE

What are the aims of this leaflet?

This leaflet has been written to help you understand more about Hailey-Hailey disease. It tells you what it is, what causes it, what can be done about it, and where you can find out more about it.

What is Hailey-Hailey disease?

Hailey-Hailey disease is also known as familial benign chronic pemphigus. It was originally described by the Hailey brothers. It is a rare inherited skin condition in which there are red scaly areas that can be itchy and sore, that can lead to superficial blisters and eroded (broken) areas of the skin. This usually affects the folds of the groin, armpits, neck and under the breasts. The condition flares intermittently and tends to come and go. Many patients are able to lead full and normal lives, with their condition being a nuisance rather than a serious problem. Some people are more severely affected and experience more persistent painful raw areas of the skin with development of superficial blisters.

What causes Hailey-Hailey disease?

The disease is caused by a change in the DNA of a gene on chromosome 3. Normally this gene helps the cells in the outer layer of the skin (the epidermis) to stick together. In Hailey-Hailey disease this adhesion of cells is weakened, resulting in separation of the cells in the surface layer of the skin. This happens especially in areas where the skin is rubbed such as the groin and under the arms. The altered gene runs in families and affects both men and women equally. Hailey-Hailey disease is not contagious and is not a result of allergies. There is a tendency for Hailey-Hailey disease to get worse with sweating and friction, as well as in hot weather.

Is Hailey-Hailey disease hereditary?

Yes. The condition is inherited in a pattern known as 'autosomal dominant inheritance', which means that there is a 1 in 2 [50:50] chance that each child of an affected parent will inherit the skin problem. The severity may vary between affected members of the same family. While it is hereditary not all patients have affected family members.

What are the symptoms of Hailey-Hailey disease?

During a flare-up, the affected skin may become uncomfortable, with burning and itching symptoms. This is aggravated if the skin surfaces are continually rubbing against each other, such as in the groin when walking. Flare-ups tend to settle down in a few days with appropriate treatment although sometimes they may improve without treatment.

Severe and painful flare-ups in an area such as the groin, genital area or in the crease of the buttocks may make walking and working so uncomfortable that an individual may need a few days of rest. Severe flare-ups may lead to infection and discharge, which may cause increased pain or a strong smell from the affected areas. The affected areas may become particularly problematic if secondarily infected. This may be due to bacterial and/or viral (particularly the cold sore virus) infection.

What does Hailey-Hailey disease look like?

The condition usually first appears between the ages of 15 and 40 years, but it may begin at any age. The severity of the condition varies widely and is unpredictable. Red, scaly areas, superficial blisters or raw areas appear at areas of friction, especially at the sides of the neck and in the skin folds (under the arms, groin or under the breasts). The skin looks normal in between flare ups, and does not scar. However some patients may find that there is a brown discolouration that can occur as a result of the inflammation (post-inflammatory hyperpigmentation), which can take months to fade. Some people notice that their condition improves as they get older.

How will Hailey-Hailey disease be diagnosed?

It is very common for the condition to be mistaken for other skin conditions such as eczema, psoriasis, contact dermatitis (due to an allergy), fungal infection or impetigo (a bacterial skin infection). The diagnosis of Hailey-Hailey disease can be confirmed by taking a sample of the skin (a biopsy) for

examination under the microscope. A second biopsy is often taken for direct immunofluorescence to confirm the diagnosis.

Can Hailey-Hailey disease be cured?

No. The underlying genetic defect cannot be altered; however, treatment does help with prevention of flare-ups and may increase the speed of healing.

How can Hailey-Hailey disease be treated?

There are several ways of helping patients with Hailey-Hailey disease:

Topical treatments:

- Corticosteroid anti-inflammatory creams can be very helpful and may be prescribed in combination with an antibiotic cream to treat skin infection. A pricking or itching sensation is usually present when a flare up is starting and the corticosteroid cream may prevent the rash from developing any further if the cream is applied immediately. Enough of the cream should be applied to cover the affected area. The cream only needs to be used once or twice a day on the affected skin. There is no need to put the steroid cream on unaffected skin.
- Careful washing is important with an antiseptic soap substitute or antiseptic bath oil to prevent infection occurring.
- Topical antibiotic e.g. clindamycin, mupirocin, or antifungal e.g. ketoconazole creams can be helpful for short periods of time such as a week or two. Long-term antibiotic creams are best avoided to reduce risks of germs becoming resistant.

Tablet treatments:

- There is an increased chance of developing a skin infection if you have Hailey-Hailey disease. Infection with bacteria such as *Staphylococcus aureus* may occur and require treatment with oral antibiotics prescribed by your doctor.
- The "cold sore" virus (herpes simplex) sometimes causes skin infection in patients with Hailey-Hailey disease. If the condition suddenly gets worse and is much more painful than usual this may be a sign of a cold sore virus infection. You should contact your GP or dermatologist immediately if this happens, as you may need oral anti-viral treatment.
- Sometimes oral (taken by mouth) treatments that affect the immune system may need to be used (e.g. oral corticosteroids, ciclosporin, acitretin or methotrexate). These drugs have potentially serious side effects and are only prescribed for patients who do not respond to first line topical treatment or oral antibiotic treatment.

- There is new evidence for the use of magnesium chloride oral solution. Naltrexone, which inhibits opioid pain-relieving receptors, may also be of help. It is not clear yet how these work.

Other treatments:

- Botox (botulinum toxin) is an injection which can help alongside topical treatments. It works by reducing sweating and keeping the area dry, and reduce the risk of infection.
- Topical photodynamic therapy, a type of light treatment, has been reported to help in some resistant cases. This involves application of a cream to the affected skin that sensitises the skin cells to light, followed by exposure to a special red lamp 3 hours later. Other possible approaches to treatment include the removal of affected areas surgically (which may result in scar formation), dermabrasion and certain forms of laser treatment.

Self-care (What can I do?)

Heat, sweating or friction may make the rash develop; it is sensible to try and avoid these where possible. For example, it may be helpful to wash a new shirt to soften a stiff collar before wearing it. It is advisable to wear cool loose fitting clothing, and keep your weight down to minimise friction. Avoid sweating and friction by doing exercises such as swimming, rather than activities such as cycling. Holidays in hot and humid countries may make the condition worse.

Where can I get more information about Hailey-Hailey disease?

Links to patient support groups:

Hailey-Hailey Disease Society

Web: www.haileyhailey.com

Web links to detailed leaflets:

<https://www.dermnetnz.org/topics/benign-familial-pemphigus>

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

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