



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, as originally described by the Hailey brothers. It is a rare inherited skin condition in which red scaly areas that can be itchy and sore, can lead to superficial blisters and eroded (broken) areas of the skin 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 patients are more severely affected and experience more persistent painful raw areas of the skin with development of superficial blisters.

What causes Hailey-Hailey disease?

A small error in the DNA code of a gene on chromosome number 3 (one of the 46 chromosomes that we all have) has been shown to cause Hailey-Hailey disease. Normally this gene plays a part in ensuring that the cells in the outer layer of the skin (the epidermis) 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, especially in areas of skin prone to friction 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, or 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.

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 it 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 a patient might need a few days of rest. Severe flare-ups may lead to infection and discharge and may lead to an unpleasant smell from the affected areas.

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, in the skin folds under the arms, groin or under the breasts. The skin looks normal in between flare-ups, and does not scar. Some patients 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 examining a sample of the skin (a biopsy) under the microscope. A test called direct immunofluorescence is also performed on a skin biopsy to confirm the diagnosis.

Can Hailey-Hailey disease be cured?

No. The underlying genetic defect cannot be altered; however, treatment does help with varying periods of remission.

How can Hailey-Hailey disease be treated?

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

- 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.
- There is an increased chance of developing a skin infection if you have Hailey-Hailey disease. Infection with germs 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.
- More recently, tacrolimus ointment has been found useful in some patients, although this preparation does not have a product licence to treat Hailey-Hailey disease.
- Sometimes oral (taken by mouth) treatments that affect the immune system may need to be used (e.g. oral corticosteroids, ciclosporin, dapsone 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.
- 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. The treatment can be painful.
- Other possible approaches to treatment include the use of botulinum toxin (to reduce sweating), the removal of affected areas surgically (which may result in scar formation), and 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. 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 comfortable fitting, 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:

www.dermnetnz.org/dna.Hailey-Hailey/HHD.html

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

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

**BRITISH ASSOCIATION OF DERMATOLOGISTS
PATIENT INFORMATION LEAFLET
PRODUCED FEBRUARY 2008
UPDATED JANUARY 2011, JANUARY 2014,
DECEMBER 2016
REVIEW DATE DECEMBER 2019**

