**Hailey-Hailey disease**

**What is Hailey-Hailey disease?**

Hailey-Hailey disease is also known as familial benign chronic pemphigus. It is a rare inherited skin condition, in which red scaly areas or small blisters appear at sites of friction.

**What causes Hailey-Hailey disease?**

A small error in the DNA code for 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 properly. In Hailey-Hailey disease, this adhesion of cells is impaired, resulting in separation of the cells in the epidermis, 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. 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 – see above. The condition is inherited in a pattern known as '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?**

Affected skin may become uncomfortable and itchy if the skin surfaces are continually rubbing against each other, such as in the groin when walking. Outbreaks tend to settle down in a few days with appropriate treatment (see below), and may improve without treatment. Very rarely, a severe flare-up 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 rest.

**What does Hailey-Hailey disease look like?**

The first signs of the condition usually appear between the ages of 15 and 40 years, but it may begin outside this age range. The severity varies enormously and is unpredictable. Red, scaly areas or small blisters appear at areas of friction, especially at the sides of the neck or in the skin folds under the arms, the groin or under the breasts. The skin looks entirely normal inbetween active episodes, and does not scar. Many 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 eczema, contact dermatitis (due to an allergy), a sweat rash, a fungal infection or impetigo (a bacterial skin infection). If required, the diagnosis of Hailey-Hailey disease can be confirmed by examining a sample of the skin (a biopsy) under the microscope.

**Can Hailey-Hailey disease be cured?**

No. The underlying genetic defect cannot be altered; however, treatment does help and long remissions are common.

For further information about Hailey-Hailey disease please see[this page](http://www.bad.org.uk/site/822/default.aspx) on the website of the British Association of Dermatologists

