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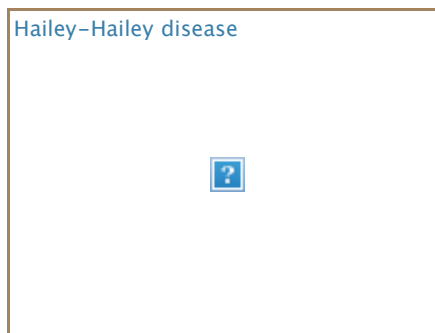
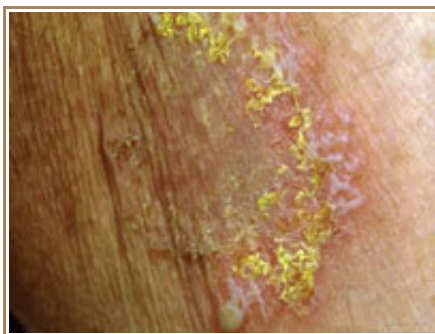
Hailey-Hailey disease

Hailey-Hailey disease, sometimes called 'familial benign chronic pemphigus' is a rare hereditary blistering skin disease. The Hailey brothers first described it in 1939.

Hailey-Hailey disease usually appears in the third or fourth decade, although it can occur at any age. It typically begins as a painful erosive skin rash in the skin folds. Common sites include the armpits, groins, and neck, under the breasts and between the buttocks. The lesions tend to come and go and leave no scars. As the lesions get bigger the centre clears leaving a typical ring shape. If the lesions are present for some time they may become thickened. The skin then tends to macerate leaving quite painful cracks. Secondary bacterial infection, which is not uncommon, can give rise to an unpleasant smell. White bands on the fingernails and pits in the palms can also occur.

Heat, sweating and friction often exacerbates the disease, and most patients have worse symptoms during the summer months.

Hailey-Hailey disease



What is the cause?

Hailey-Hailey disease is an inherited skin disorder, although occasionally sporadic cases arise without a family history. The defect responsible has now been identified on a gene called *ATP2C1* found on chromosome 3q21-24. This gene codes for the protein SPCA1 (Secretory Pathway Calcium/manganese-ATPase), a calcium and manganese pump. The skin cells (keratinocytes) stick together via structures called desmosomes and it seems the desmosomes do not assemble properly if there is insufficient calcium.

The genetic defect in Hailey-Hailey disease causes the skin cells to become unstuck from one another. Normally the cells are packed together tightly in much the same way as bricks and mortar. Patients with Hailey-Hailey disease have defective 'mortar' and the cells fall apart, like a dilapidated brick wall.

As yet there is no diagnostic test available to family members.

How is it diagnosed?

Usually Hailey-Hailey disease is diagnosed by its appearance and the family history, but it is often mistaken for other skin problems. [Impetigo](#), [thrush](#), [tinea](#) (jock itch) and other blistering conditions look similar.

Diagnosis may require a [skin biopsy](#). The histology is characteristic, with layers of detached skin cells ('acantholysis'). Unlike [pemphigus vulgaris](#), the immunofluorescence test for antibodies is negative.

How is it treated?

Unfortunately there is no cure for Hailey–Hailey disease. Treatment is aimed at reducing symptoms and preventing flares.

- Avoiding trigger factors such as [sunburn](#), sweating and friction helps to control the disease.
- Wear absorbant pads in underwear
- [Corticosteroid](#) (cortisone) creams are effective in treating lesions and preventing exacerbations.
- Combination corticosteroid/ antibiotic creams are helpful, as treating the secondary [infection](#) is important.
- Prolonged courses of oral antibiotics such as [tetracycline](#) may be useful.
- [Benzoyl peroxide](#) is another useful antibacterial cream.
- [Calcipotriol](#) (Daivonex) cream is useful in some patients.
- If [herpes](#) virus infection is a recurrent problem oral antivirals such as [aciclovir](#) may be useful.
- A number of other oral medications ([retinoids](#), [ciclosporin](#), [dapson](#)e, and [methotrexate](#)) have been reported in single cases as partially effective, but large trials have not been performed.
- Wet compresses to dry up oozing patches
- [Botulinum toxin](#) to reduce sweating
- [Phototherapy](#) (ultraviolet light) has also been used.
- [Lasers](#) have been reported to be useful in one study, vaporising the affected skin.
- In severe cases surgery can be performed to remove the affected skin. Skin grafts are usually necessary to repair the wounds.

Are there any complications?

For the vast majority of patients Hailey–Hailey disease is a nuisance rather than a serious problem. The pain and smell can often be the main problems. If the lesions get infected with herpes virus a sudden severe flare can occur, which often needs prompt treatment (see above).

Will it improve?

Many patients have long remissions and an improvement with age does occur.

Related Information

Other websites:

- [International Pemphigus Foundation](#)
- [The British Association of Dermatologists'](#) information for patients with Hailey–Hailey disease
- [Benign familial pemphigus](#) – emedicine dermatology, the online textbook

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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