CONDITION

Acantholysis bullosa

Learn about acantholysis bullosa, also called Hailey-Hailey disease, including its inherited cause, skin symptoms, diagnosis, treatment options, and when to seek urgent care.

WHAT IT IS
Inherited skin condition causing recurrent fragile skin

BODY SYSTEM
Skin

MAIN SYMPTOMS
Blisters, raw patches, cracks, crusting

WHAT CAUSES IT
Inherited ATP2C1 gene change

HOW IT IS DIAGNOSED
Skin biopsy

MAIN TREATMENT
Skin protection, trigger reduction, inflammation and infection control

CAN IT BE CURED?
No, inherited tendency remains in skin

IS IT AN EMERGENCY?
Sometimes, rapidly spreading redness with fever

LONG-TERM OUTLOOK
Long-term flares alternating with quieter periods

Overview

Acantholysis bullosa is an uncommon name for Hailey-Hailey disease , also called familial benign chronic pemphigus. It is an inherited skin condition that causes repeated episodes of fragile, irritated skin, especially in areas where skin rubs against skin.

The term acantholysis means that skin cells lose some of the connections that normally hold them together. As a result, the outer layer of skin can split easily. This produces small blisters, raw patches, cracks, and crusting.

Symptoms often affect the armpits, groin, neck, buttock crease, and skin beneath the breasts. Heat, sweating, friction, and skin infections can trigger or worsen a flare. The condition often begins in early to middle adulthood, although it can appear at other ages.

Acantholysis bullosa is not contagious. It is also different from autoimmune pemphigus, in which the immune system mistakenly attacks skin-cell connections. Despite the word "bullosa," it is not a form of epidermolysis bullosa, a separate group of inherited skin-fragility disorders.

Hailey-Hailey disease usually follows a long-term pattern of flares and quieter periods. It can be painful and may interfere with walking, exercise, sleep, work, or sexual activity. Treatment focuses on protecting the skin, reducing triggers, controlling inflammation, and treating infection when it occurs.

Symptoms

The condition usually causes tender or itchy patches in warm, moist skin folds. A flare may begin with small blisters or red bumps. Because the affected skin is fragile, the blisters often break quickly and may not be noticed.

The broken areas can join together to form moist, raw patches called erosions . Cracks may develop where the skin bends or stretches. The surface may become crusted, scaly, or thickened during a prolonged flare.

Commonly affected areas include:

Symptoms may include burning, itching, soreness, or pain. Moisture and bacterial growth can produce an unpleasant odor. Odor does not necessarily mean that hygiene is poor, but a new or worsening odor can be a reason to check for infection.

Symptoms are often similar on both sides of the body. Some people have mild changes limited to one or two folds, while others experience larger or more frequent flares. The mouth is usually unaffected, although uncommon mouth or genital changes can occur.

Fine white lines running lengthwise along the fingernails may develop. These lines do not usually damage the nails and may be easy to overlook.

- The armpits and groin
- The sides or back of the neck
- Beneath the breasts
- The crease between the buttocks
- Skin around the genitals or anus
- Other areas exposed to repeated rubbing

Causes and risk factors

Acantholysis bullosa is usually caused by a disease-related change in the ATP2C1 gene. This gene provides instructions for a protein that helps control calcium and manganese inside skin cells. Proper mineral balance is important for the structures that join neighboring cells together.

When these connections are weakened, heat, moisture, friction, or inflammation can make the upper skin layers separate. This explains why symptoms often occur in body folds.

The condition follows an autosomal dominant inheritance pattern. This means that a person can inherit the condition from one affected parent. Each child of an affected person has a possibility of inheriting the altered gene. However, the age of onset and severity can vary greatly, even among relatives with the same genetic change.

Some people have no known family history. This may happen because relatives had mild or unrecognized symptoms, or because the genetic change occurred for the first time in that person.

Factors that may trigger or aggravate symptoms include:

Having one of these triggers does not confirm the condition. Several common skin disorders can cause similar rashes in body folds.

- Hot weather, sweating, and high humidity
- Friction from clothing, exercise, or skin rubbing
- Excess moisture in skin folds
- Cuts, scratching, shaving, or other skin injury
- Bacterial, fungal, or viral skin infections
- Irritating soaps, deodorants, adhesives, or skin products
- Sunburn or other sources of heat

Diagnosis

A dermatologist may suspect Hailey-Hailey disease from the appearance, location, and repeated pattern of the rash. A family history can support the diagnosis, but it is not required.

Because the rash can resemble eczema, contact dermatitis, inverse psoriasis, fungal infection, or ordinary skin-fold irritation, testing may be needed. A clinician may collect a swab or scraping if bacterial, fungal, or viral infection is possible.

A skin biopsy is often used to confirm uncertain cases. After numbing the area, the clinician removes a small skin sample for examination under a microscope. Hailey-Hailey disease produces a characteristic loss of connections between skin cells. This appearance is sometimes described as a "dilapidated brick wall."

A test called direct immunofluorescence may be performed when autoimmune blistering disease is a concern. This test looks for immune proteins deposited in the skin. It is generally negative in Hailey-Hailey disease and can help distinguish it from autoimmune pemphigus.

Genetic testing for an ATP2C1 change is not necessary in every case. It may be considered when the diagnosis remains uncertain or when a person wants more information for family planning. A genetics professional can explain the test's possible benefits, limitations, and effects on relatives.

Treatment and management

There is no single treatment that prevents every flare. Management is usually adjusted according to the affected area, symptom severity, infection risk, and response to earlier treatments.

Reducing heat, sweat, and friction is an important part of care. Loose, breathable clothing may be more comfortable than close-fitting garments. Promptly changing out of sweaty clothing and gently drying skin folds can also help. Weight management may reduce friction for some people, but the condition is not caused by body weight.

Skin should be washed gently rather than scrubbed. Fragranced products or strong cleansers may increase irritation. A clinician may recommend a particular antiseptic wash when recurrent infection or bacterial overgrowth is a concern. Antiseptics should be used as directed because overly strong or frequent use can damage already fragile skin.

During a flare, nonstick dressings can protect raw areas from clothing and further rubbing. Plain barrier ointments may reduce friction, although some thick or fragranced products can trap heat or irritate sensitive skin. A dermatologist can help identify suitable products.

Topical corticosteroids are commonly used for short periods to reduce inflammation. Skin folds absorb medicine readily and are more vulnerable to steroid like prednisone or hydrocortisone side effects, such as thinning, stretch marks, and color changes. For that reason, the strength, location, and duration should be guided by a clinician.

Topical calcineurin inhibitors, such as tacrolimus or pimecrolimus, may sometimes be used to control inflammation without causing steroid-related skin thinning. These medicines can sting or burn when first applied, particularly on open skin. Their role should be discussed with the

treating clinician.

Antibiotic like amoxicillin or doxycycline , antifungal like fluconazole or terbinafine , or antiviral treatment may be needed when a specific infection is identified or strongly suspected. These medicines do not treat the underlying genetic condition, and routine antibiotic use can contribute to side effects and antibiotic resistance.

If heavy sweating regularly triggers symptoms, treatment aimed at reducing sweat may be considered. Options can include prescription antiperspirants, medicines that reduce sweating, or botulinum toxin injections. Each approach has possible side effects and is not suitable for everyone.

For persistent disease, dermatologists may discuss additional topical medicines, oral medicines, or procedures. Reported options include oral antibiotics with anti-inflammatory effects, retinoids, low-dose naltrexone, laser treatment, dermabrasion, and surgical removal of severely affected skin. Many of these uses are off-label , meaning the medicine or procedure has not been specifically approved for Hailey-Hailey disease. Evidence often comes from small studies or individual reports rather than large clinical trials.

Procedures are usually reserved for an area that continues to cause significant problems despite simpler measures. They may improve treated skin but do not change the inherited tendency in other areas. A specialist can explain expected recovery, scarring risk, and the chance of symptoms returning.

Complications

The damaged skin barrier makes secondary infection more likely. Bacteria or yeast can increase pain, crusting, drainage, or odor. The herpes simplex virus can also infect affected skin and may cause clusters of painful blisters or rapidly worsening erosions.

Long-lasting or repeated flares can interfere with movement and sleep. Genital or groin involvement may affect intimacy and urination. Visible lesions, odor, and drainage can also cause embarrassment, anxiety, or social withdrawal. These effects are valid reasons to discuss treatment adjustments and emotional support.

Scarring is not usually the main feature of Hailey-Hailey disease, but color changes or thickened skin can remain after inflammation. Procedures and severe infections can also leave scars.

Skin cancer arising in chronically affected skin has been reported but is unusual. A sore, lump, or thickened area that does not heal should be assessed rather than assumed to be another routine flare.

Follow up

Routine follow-up with a primary care clinician or dermatologist can help identify triggers, review skin care, and reduce medicine-related side effects. Follow-up is particularly useful when flares are frequent, the diagnosis is uncertain, or treatment is no longer controlling symptoms.

Arrange a prompt medical review for:

Seek emergency care for severe illness, confusion, fainting, difficulty breathing, or rapidly spreading redness with fever. These findings are not typical of a routine flare and may indicate a serious infection or another urgent problem.

People who are pregnant, planning pregnancy, or breastfeeding should review all medicines with their clinician. Those who want to understand the chance of passing the condition to a child may request genetic counseling.

Keeping a brief record of flares can support routine care. Useful details include the affected areas, heat or sweating exposure, new products, possible infection symptoms, and treatments used.

- Increasing pain, warmth, swelling, pus, or foul-smelling drainage
- A flare that spreads quickly or does not respond as expected
- Clusters of new, painful blisters that may represent herpes infection
- Fever or feeling unwell along with worsening skin symptoms
- A persistent ulcer, lump, or changing area of skin

Sources

- MedlinePlus Genetics. Hailey-Hailey disease . U.S. National Library of Medicine, National Institutes of Health.
- British Association of Dermatologists. Hailey-Hailey disease . Patient Information Leaflet.
- Ben Lagha I, Ashack K, Khachemoune A. Hailey-Hailey disease: an update review with a focus on treatment data. American Journal of Clinical Dermatology . 2020;21(1):49-68.
- Engin B, Kutlubay Z, Çelik U, Serdaro lu S, Tüzün Y. Hailey-Hailey disease: a fold (intertriginous) dermatosis. Clinics in Dermatology . 2015;33(4):452-455.



- Burge SM. Hailey-Hailey disease: the clinical features, response to treatment and prognosis. British Journal of Dermatology . 1992;126(3):275-282.
- Hu Z, Bonifas JM, Beech J, et al. Mutations in ATP2C1, encoding a calcium pump, cause Hailey-Hailey disease. Nature Genetics . 2000;24(1):61-65.