CONDITION

Familial lichen amyloidosis

Familial lichen amyloidosis is an inherited skin condition that causes very itchy clusters of thickened bumps, often on the shins.

WHAT IT IS
Inherited localized skin amyloid causing itchy, raised patches

BODY SYSTEM
Skin

MAIN SYMPTOMS
Intense itching, clustered bumps, thickened skin, darker patches

WHAT CAUSES IT
Inherited changes affecting skin cells and itch signalling

HOW IT IS DIAGNOSED
Skin biopsy; genetic testing may identify the inherited cause

MAIN TREATMENT
Topical itch control and other skin-directed treatments

CAN IT BE CURED?
No - treatments control symptoms, not the inherited tendency

IS IT AN EMERGENCY?
No - urgent care for spreading redness with fever

LONG-TERM OUTLOOK
Chronic but usually limited to the skin

Overview

Familial lichen amyloidosis is an inherited form of primary localized cutaneous amyloidosis. "Cutaneous" means involving the skin, while "localized" means that the abnormal material remains in the skin rather than affecting organs throughout the body.

Small amounts of a protein material called amyloid collect in the upper layer of the skin. The amyloid is thought to come from damaged keratinocytes, the cells that make up most of the outer skin. These deposits contribute to firm, closely grouped bumps and persistent itching.

The bumps most often develop on the shins but may also occur on the thighs, arms, or back. Scratching can make the skin thicker, darker, and itchier, creating an itch-scratch cycle.

Familial lichen amyloidosis is also called type IX amyloidosis. It differs from systemic

amyloidosis, in which amyloid deposits can damage internal organs. The lichen form is generally confined to the skin and is not expected to become systemic amyloidosis.

The itching can be intense and long-lasting. It may disturb sleep, affect concentration, and make clothing or physical activity uncomfortable. Treatment focuses on reducing itching, protecting the skin, and flattening thickened areas.

Symptoms

Symptoms may begin earlier in inherited cases than in non-inherited lichen amyloidosis, although the age of onset varies. Several relatives may have similar skin changes but different levels of severity.

Typical features include:

The bumps are usually small and closely packed. They may form rippled, ridged, or plaque-like areas. Some people have features of both lichen amyloidosis and macular amyloidosis, which causes flatter, darker, rippled patches.

Itching may be constant or occur in flares. Heat, dry skin, sweating, stress, friction, and scratching can make it worse. Broken skin, crusting, bleeding, or soreness usually results from rubbing and scratching rather than the amyloid itself. Damaged skin is also more vulnerable to infection.

- Very itchy, firm bumps, often on both shins
- Groups of brown, reddish-brown, or skin-coloured bumps
- Thickened, rough skin caused partly by repeated scratching
- Darker or lighter marks remaining after inflammation

Causes and risk factors

Familial lichen amyloidosis results from an inherited tendency to develop amyloid deposits in the skin. Changes in the OSMR or IL31RA genes have been identified in some affected families. These genes help control signals involved in skin inflammation, cell survival, and itching.

The condition is usually autosomal dominant, meaning one altered copy of a related gene can be enough to cause it. However, not every family has an identifiable gene change, and symptoms may vary among relatives with the same change.

Scratching does not cause the inherited condition, but it can damage skin cells and contribute

to amyloid formation, thickening, and continuing itch. Friction from rough fabrics or repeated rubbing may have a similar effect.

A close relative with lichen amyloidosis increases the likelihood of a familial form. A family history of unexplained, long-term itchy bumps or darkened patches may also be relevant.

Lichen amyloidosis can occasionally occur alongside other inherited conditions. For example, an itchy amyloid patch on the upper back has been reported in some families with multiple endocrine neoplasia type 2. This is a separate genetic situation and does not mean most people with lichen amyloidosis have an endocrine tumour syndrome.

Diagnosis

A dermatologist will examine the shape, colour, texture, and distribution of the skin changes. Your personal and family history can help distinguish familial lichen amyloidosis from other itchy skin conditions.

A skin biopsy is commonly used to confirm the diagnosis. After numbing the area, a clinician removes a small sample for examination under a microscope. Special staining, often Congo red viewed with polarized light, can show amyloid in the upper skin.

Biopsy can also rule out conditions with a similar appearance, including eczema, lichen simplex chronicus, prurigo nodularis, and hypertrophic lichen planus.

Genetic testing may identify a change in OSMR , IL31RA , or another relevant gene. It can support the diagnosis and help relatives understand the inheritance pattern. A negative result does not rule out familial lichen amyloidosis because current tests do not identify every genetic cause.

Blood tests and organ scans are not routinely needed solely because lichen amyloidosis is present. They may be considered if symptoms or examination findings suggest a separate health problem.

Treatment and management

No treatment reliably removes the inherited tendency or permanently prevents skin changes. Management depends on the severity of the itching, the extent and thickness of the bumps, and their effect on daily life.

Regular use of a fragrance-free moisturizer can reduce dryness and cracking. Cooling the skin, keeping nails short, and covering itchy areas at night may limit damage from unconscious

scratching.

Prescription topical corticosteroids can reduce inflammation and itching. They are generally used for limited periods because prolonged or inappropriate use can thin the skin and cause other local effects. A clinician may recommend placing a dressing over the medicine to increase absorption, but this also raises the risk of side effects.

Other options include topical calcineurin inhibitors, which reduce inflammation without being steroids. Creams containing urea or salicylic acid may soften thick scale and rough areas, although they can sting or irritate broken skin.

Antihistamines like cetirizine or loratadine do not treat the amyloid deposits. A sedating antihistamine may sometimes help with sleep when itching is severe, but it can cause daytime drowsiness and is not suitable for everyone.

Phototherapy exposes affected skin to controlled ultraviolet light. Narrowband ultraviolet B or psoralen plus ultraviolet A therapy may improve itching and appearance. It requires repeated visits and carries risks including burning, pigment changes, and cumulative ultraviolet exposure.

For severe or widespread symptoms, a dermatologist may discuss systemic medicines such as a retinoid. These require careful monitoring and may cause important adverse effects, including serious pregnancy-related risks. Evidence for systemic treatment is limited, and improvement is not guaranteed.

Laser treatment, dermabrasion, or other procedures may flatten selected areas. Recurrence, scarring, and skin-colour changes are possible. Treatment often combines itch control, protection from scratching, and methods to soften or flatten the bumps.

Complications

Familial lichen amyloidosis does not usually damage internal organs or shorten life. Its main complications result from persistent itching, scratching, and treatment side effects.

Scratched skin can become cracked, crusted, or infected. Signs of infection include increasing pain, warmth, swelling, pus, or redness spreading beyond the original patch.

Long-term inflammation may leave darker or lighter skin, even after the bumps and itching improve. Repeated rubbing can also cause marked thickening.

Active itching may disrupt sleep and affect mood, concentration, school, or work. Visible lesions may cause embarrassment or distress.

Treatment complications can include skin thinning from topical steroids like hydrocortisone or

betamethasone , irritation from keratolytic creams, burns from phototherapy, or scarring after procedures. Regular review helps balance symptom relief against these risks.

Prevention

The inherited tendency to familial lichen amyloidosis cannot currently be prevented. Lifestyle choices do not cause the gene change, and the condition is not the result of poor skin care.

Reducing scratching and friction may limit further damage and prevent patches from becoming thicker. Regular moisturizing and early treatment of itching may make flares easier to manage, but they cannot guarantee that new areas will not appear.

Genetic counselling can explain the likely inheritance pattern, the limits of testing, and what results may mean for relatives or future pregnancies. It does not change the condition in someone who already has it.

Living with it and what to expect

Familial lichen amyloidosis is usually long-term. Individual bumps or patches may remain for months or years. Symptoms can improve, return, or spread slowly to other areas.

Itching often responds sooner than the visible skin changes. Dark or light marks may take much longer to fade and can remain after inflammation is controlled.

Some people manage the condition with moisturizers and occasional prescription treatment. Others need continuing dermatology care, especially if itching disrupts sleep or large areas are affected. Improvement is often partial, and symptoms may return after treatment stops.

Routine follow-up helps clinicians adjust treatment, check for side effects, and assess changes in the lesions. A new type of lump, an ulcer that does not heal, or a major change in appearance should be examined rather than assumed to be part of lichen amyloidosis.

Most people can continue their usual activities, although practical changes such as wearing softer clothing or keeping the sleeping environment cool may help during active flares.

When to get medical help

Arrange a routine medical appointment for persistent itchy bumps, especially if close relatives have similar skin changes. Seek assessment if a known patch spreads, itching regularly disrupts sleep, or treatment no longer controls symptoms.

Contact the clinician managing your treatment if you notice skin thinning, easy bruising, severe irritation, or other possible side effects. New or changing lesions may need examination or another biopsy.

Seek prompt medical care for rapidly spreading redness, increasing warmth or pain, pus, red streaks, or fever. These may indicate an infection where scratching has broken the skin.

Call emergency services for trouble breathing, swelling of the face or tongue, faintness, or a widespread blistering or peeling rash after using a medicine. These may be signs of a serious treatment reaction rather than familial lichen amyloidosis itself.

Common questions

Is familial lichen amyloidosis contagious?

No. It is related to an inherited tendency and cannot spread through touching, sharing towels, or close contact.

Does lichen amyloidosis affect internal organs?

The primary localized lichen form is generally confined to the skin. It differs from systemic amyloidosis, which can involve organs such as the heart or kidneys.

Will the bumps go away on their own?

They may become less noticeable, but they often persist without treatment. Skin-colour changes can remain after the bumps flatten.

Why does it itch so much?

Inherited changes may affect itch signalling. Amyloid deposits, inflammation, dryness, and scratching can then reinforce an itch-scratch cycle.

Can I pass it on to my children?

Familial lichen amyloidosis is commonly inherited in an autosomal dominant pattern, so an affected parent can pass it to a child. A genetic counsellor can explain the pattern in your family and discuss testing.

Is there a permanent cure?

There is currently no established permanent cure. Treatment may reduce itching, flatten bumps, and improve comfort, but the inherited tendency remains and symptoms can return.

Sources

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