

# Itching

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People with ARCI-lamellar and other forms of ichthyosis are unable to sweat normally and often develop itching, or “pruritus,” in hot, humid weather or when otherwise overheated by exercise, sunlight, or by overdressing in warm clothes. The reason for this inability to sweat normally is that the excessive skin scale clogs the pores of the sweat glands and traps the sweat. The trapped sweat irritates the skin, leading to itchiness, redness, and occasionally small blisters.

Scaling conditions that show skin redness or inflammation often have associated itching. These conditions include: congenital ichthyosiform erythroderma (CIE), erythrokeratoderma variabilis (EKV), Hailey-Hailey disease, Darier disease, pityriasis rubra pilaris (PRP), and others. Sjögren-Larsson syndrome, although not characterized by inflamed skin, also tends to be itchy. In this case, the metabolic defect results in accumulation of molecules (leukotrienes) that can stimulate itching. Use of a leukotriene synthesis inhibitor, such as zileuton, may relieve the itching in Sjögren-Larsson syndrome.

People with Netherton syndrome and ichthyosis vulgaris may also experience atopic dermatitis. Atopic dermatitis is a chronic inflammatory skin condition that is characterized by extreme itching, leading to scratching and rubbing that, in turn, creates the lesions of eczema. Another characteristic of Netherton syndrome is a predisposition to allergies, such as asthma or food allergies.

Cold winter air, with its associated low humidity, can worsen the dry skin of persons with all forms of ichthyosis, which may also result in itchy skin. Itching can also be a symptom of secondary infection with bacteria, fungi, or infestation with scabies or lice. Itching is often worse at night and can be made worse by tension and anxiety.

## **General treatment strategies for the itchy skin of ichthyosis are to:**

1. Consult with your family doctor or dermatologist to rule out a bacterial or fungal infection. Infections should be treated with the appropriate antibiotic or anti-fungal agent.
2. Keep the skin as moist as possible by using moisturizers and keratolytics. (Keratolytics, products containing alpha-hydroxy acids, or urea, that help exfoliate the skin may be useful, but, because they are sometimes irritating, can also increase itching in some patients). Skin care products which contain oatmeal, oatmilk, or colloidal oatmeal may help to soothe itchy skin.
3. Try to resist scratching! Scratching can cause a thickening of the skin and can lead to infection.

## **For temporary relief of itching:**

- Apply cool compresses to the affected areas.
- Try a lukewarm baking soda or oatmeal bath.
- Stay away from heat and humidity, and wear loose cotton clothing.
- Apply over-the-counter preparations for minor skin irritations, such as those containing pramosone or menthol. Calamine lotion can soothe itchiness, but tends to be quite drying.
- Do not use products with “caine” in them, such as Benzocaine® or Lanacane® products, or products with diphendryamine (Benadryl) directly on the skin. These preparations can effectively act as a local anesthetic by numbing the sensory nerve endings, but are known to be “contact sensitizers,” which can lead to skin allergies and a worsening of inflammation and itchiness in some individuals.

## **For prolonged, chronic, or problem itching, your doctor may prescribe:**

- Oral antihistamines. Antihistamines prevent the release of histamines deep in the cells of the skin. Histamines are chemicals that may aggravate itch nerves. Certain antihistamines are often used to induce drowsiness and promote sleep, if the itching is disrupting sleep.

Doxepine and hydroxyzine are sometimes prescribed to relieve itching. These compounds have a sedating effect and are also used to treat depression and anxiety. Other antihistamines do not have this affect, so speak to your doctor about which formulas are right for you.

- Topical steroids or a steroid-free topical immunomodulator. Topical steroids, from over-the-counter hydrocortisone to prescription strength preparations, decrease the production and action of chemical mediators of inflammation, and may help to reduce itching. The new steroid-free immunomodulators are also effective in treating inflammatory skin diseases like atopic dermatitis; however, many people with ichthyosis have skin that more easily allows the absorption of applied medications. Steroids or the non-steroidal medications could be absorbed through the skin and into the bloodstream. These medications should not be applied to large areas of the body surface by ichthyosis patients for this reason. Check with your doctor to find out which formulas may be right for you.
- Oral steroids. Cortisone and its derivatives are among the most effective anti-inflammatory drugs known. Long-term use of steroids can cause side effects such as changes in physical appearance, weight gain, irritability or depression. Treatment with steroids should always be kept at the lowest possible effective dose and should never be stopped suddenly. Oral steroid use must be monitored by your doctor.
- Systemic retinoids. Systemic retinoids, which are derivatives of Vitamin A in a pill form, help to normalize the skin and reduce inflammation throughout the whole body. Systemic retinoids do not treat the itch specifically, but help to improve the ichthyosis that is causing the itch. Systemic retinoid therapy almost always causes side effects; most are usually mild and easily controlled. However retinoids can also cause severe birth defects if taken by pregnant women. Retinoids should never be taken except under the direction of a doctor, and under strict guidelines, as outlined by the FDA and the drug company. Under no circumstances should retinoids be taken by anyone other than for whom the drug was prescribed.
- Anti-depressant agents. Tricyclic antidepressants have an antihistamine effect, induce sleep, and reduce itching. If itching is so severe that it causes sleep deprivation, irritability, and stress, and does not respond to other treatments, antidepressants can help to reduce the itch, scratch, itch cycle. Doxepin and hydroxyzine are sometimes prescribed for itching. These compounds have a sedating effect and are also used for treating anxiety and depression. These medications need to be used under a doctor's supervision.

The itching associated with various forms of ichthyosis can cause much physical and emotional distress. The scratch, itch, scratch cycle can lead to serious skin complications, including infection, and can interfere with daily activities, relaxation, and sleep. It is important to work with your doctor to identify the underlying cause of the itching and to determine the most effective, least potent treatment. Treating the problem on your own with hit-or-miss strategies may make the problem worse and lengthen the amount of time needed to get the itching under control. See your doctor early to get the best possible treatment advice.

*This information is provided as a service to patients and parents of patients who have ichthyosis. It is not intended to supplement appropriate medical care, but instead to complement that care with guidance in practical issues facing patients and parents. Neither FIRST, its Board of Directors, Medical & Scientific Advisory Board, Board of Medical Editors, nor Foundation staff and officials endorse any treatments or products reported here. All issues pertaining to the care of patients with ichthyosis should be discussed with a dermatologist experienced in the treatment of their skin disorder.*



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***Improve lives and seek cures for those affected by ichthyosis and related skin types.***

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