

# Living with ichthyosis

A guide to the condition and its management

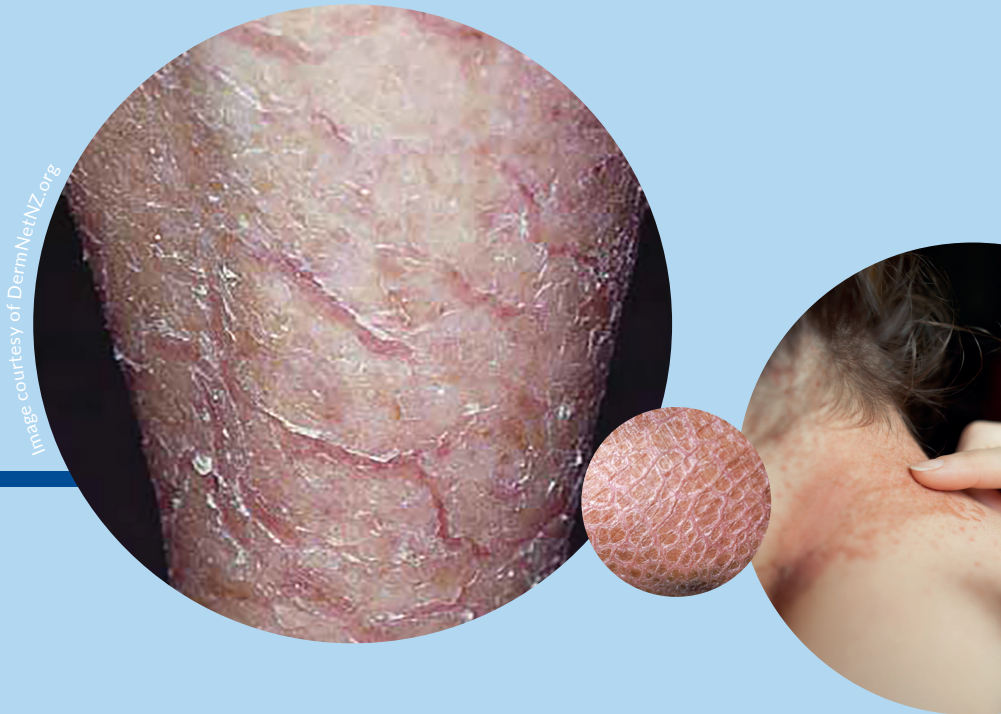


# Ichthyosis

Ichthyosis describes a group of long-term conditions in which the skin is dry and scaly. The word ichthyosis comes from the Greek words meaning 'fish' and 'disease'.

Unlike dry skin conditions such as eczema and psoriasis, the scaling found in ichthyosis is continuous and can affect the whole body. The skin may also be inflamed or thickened. The skin always peels but in some forms it may also blister.

There are many different types of ichthyosis and some are described here in more detail.



## What is ichthyosis?

Ichthyosis is rare and usually inherited but sometimes may be acquired as the result of another medical condition. Most types of ichthyosis are congenital, meaning they run in families and are present from birth. It is not contagious.

The inherited (genetic) types tend to persist throughout life – although some milder types may improve with age. The signs and symptoms of inherited ichthyosis usually appear at birth or within the first year of life.

Acquired ichthyosis may develop at any age and may be associated with other illnesses, but this is rare.

## What are the symptoms?


Symptoms vary considerably between different people and between different types of ichthyosis. Mild forms like Ichthyosis vulgaris may appear as dry skin affecting hands, feet and some areas of the skin whilst others affect the whole body. The skin may be reddish and flaky or brownish and scaly but in most cases the body will be covered in plate-like scales. Hair and nail growth may also be affected.

## What causes it?

The skin is constantly regenerating by shedding and replacing cells. This is controlled by genes. There may be a mistake or mutation in the genes which means the process of shedding and replacement of skin cells doesn't happen properly. Different types of ichthyosis are caused by mutations in different genes. In some mutations, the skin cells are produced too quickly and they pile up on the surface of the skin, which leads to thickened skin.

In other forms, the skin cells are produced at a normal rate but the top layers are not shed to make way for the new skin cells, so they build up in layers.

Whether old skin cells are shed too slowly or new skin cells are reproducing too quickly, the result is a build-up of the rough, scaly skin condition known as ichthyosis.



*"You should enjoy life as much as you can and don't let anything get you down."*

Nusrit, 32, has Harlequin Ichthyosis

## Types of ichthyosis

There are many types of ichthyosis. All types cause dry, scaly skin but it looks different depending on the type you or your child has.

### Ichthyosis vulgaris (IV)

Ichthyosis vulgaris (IV) is the most common form of ichthyosis and affects between one in 100 to 250 people. It is usually quite mild and develops in early childhood with fine, light grey scales and roughness on the upper and lower limbs, but sparing the folds of the arms and legs.

It may be more widespread and more obvious in the winter time. It is sometimes associated with atopic or childhood allergic eczema and may cause an increased wrinkling of the palms and soles.



### X-linked (recessive) ichthyosis (XLI)

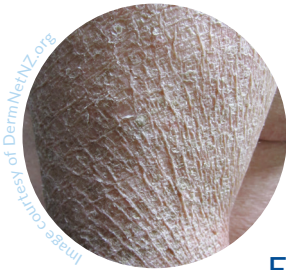
X-linked (recessive) ichthyosis (XLI) only affects boys (fewer than one in 2,000) and appears in the first few months after birth. Brownish, flat scales are seen particularly over the arms, legs and tummy. It may also affect the ears and face. It varies in severity and may improve in summer or sunny weather.

### Epidermolytic ichthyosis (EI)

Epidermolytic ichthyosis is also known as Bullous congenital ichthyosiform erythroderma (BCIE) or Epidermolytic hyperkeratosis (EHK). It appears at or soon after birth with blistering, fragile, red, shiny skin. This is later replaced by thick scaling especially around the joints. Although the scale lifts off quite easily the skin underneath may be painful and skin infections are quite common. It is a rare condition and occurs in 1 in 100,000 babies.

### Netherton syndrome (NS)

Netherton syndrome (NS) affects 1 in 200,000 babies. Red, inflamed, itchy, scaly skin is present from birth. It can vary in severity and affects all areas of the body. The hair may be sparse, brittle and spiky. Many people with NS develop food allergy and also may develop hay fever, asthma and atopic eczema.



## Autosomal recessive congenital ichthyosis (ARCI)

Autosomal recessive congenital ichthyosis (ARCI) is an umbrella term referring to a group of conditions that share a similar genetic pattern and a collodion membrane presentation at birth.

After shedding of the collodion membrane, the skin can show a variety of appearances. Although still rare, the most common types are Non-bullous ichthyosiform erythroderma, Lamellar ichthyosis and Harlequin ichthyosis.

ARCI are rare disorders estimated to occur in about 1 in 200,000 births.

### Non-bullous ichthyosiform erythroderma (NBIE)

Non-bullous ichthyosiform erythroderma (NBIE) is also known as Congenital ichthyosiform erythroderma (CIE).

It is generally seen at birth with the appearance of a collodion membrane on the newborn baby. This is a shiny film stretched across the skin, which dries out and is gradually shed during the first week of life.

The skin is red and inflamed without blisters but with fine white scales affecting the whole body including the scalp. It is a rare condition and affects 1 in 300,000 babies.

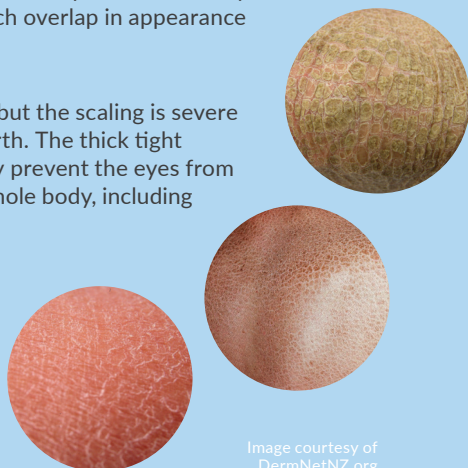
### Lamellar ichthyosis (LI)

Lamellar ichthyosis (LI) occurs in 1 in 100,000 babies and is a rare condition.

At birth, the baby will have collodion skin which is shed within a few days. This is followed by scaling all over the body and scalp, with particularly large brown scales on the limbs and torso. The skin may be itchy but is not always red. There are many subtypes of LI and CIE which overlap in appearance

### Harlequin ichthyosis (HI)

Harlequin ichthyosis (HI) is extremely rare, but the scaling is severe and requires intensive care, especially at birth. The thick tight skin can block the ears and nostrils and may prevent the eyes from shutting. Thick plates of scales affect the whole body, including the face.





## What treatments are available?

At present, there is no cure for ichthyosis, but a daily skincare routine usually keeps the symptoms manageable.

The main aim of treatment is to improve skin condition and to make it less dry and scaly. This will help the skin to feel more comfortable.

Treatment involves moisturising, and in some conditions, exfoliating the skin every day to prevent dryness, scaling, cracking and the build-up of skin cells. Some of the more common forms of ichthyosis can be mild and may improve in the summertime.

Your dermatologist, nurse or GP will prescribe or recommend suitable emollients to use for washing and moisturising by applying directly to the skin to reduce water loss and cover it with a protective film.

A range of treatments may be prescribed. If you pay prescription charges, do purchase a pre-payment prescription certificate, this will make treatment more cost effective.

Apply for a Pre-Payment Certificate (NHS) at [www.gov.uk/get-a-ppc](http://www.gov.uk/get-a-ppc)



## Emollients

Emollients may take the form of a cream, ointment, lotion or bath oil. The best emollient is the one that suits you or your child's skin condition – your healthcare professional should give you the opportunity to try a variety of emollients.

They are available in many sizes and types of container, but the most important thing is to ensure you have a sufficiently large quantity of your preferred emollient.

Your healthcare professional may prescribe an emollient or you can buy one over the counter at a pharmacy.

If you or your children need to use an emollient regularly, it's worth keeping a good supply at home, school or work. Emollients can be applied as often as you like to keep the skin well moisturised. Ideally, this should be at least 3 or 4 times a day.

Emollients should be used as soap substitutes. Soap, bubble baths and cosmetic washes should be avoided as they will dry the skin and affect the skin barrier.

### Types of emollient

**Bath Oils** leave a fine film of moisturiser all over the body. This helps to hydrate and soften the skin.

**Ointments** are greasy emollients which may be useful for dry, scaly areas of skin. They have a softening and protective action on the skin, but are not suitable for wet or weeping areas.

**Creams/lotions** are a mixture of oil and water. They are soothing to dry skin and unlike ointments are easy to wash off.

## Other treatments

### Exfoliating cream

In addition to emollients, you or your child might be prescribed an exfoliating cream or keratolytics, which are used to break-down and reduce thicker scales.

This may contain urea, lactic acid and other alpha hydroxy acids to help exfoliate the skin. They may irritate the skin and should only be used under close medical supervision.

### Medicated shampoo

Older children and adults may need a special medicated shampoo designed to loosen scaly skin on the scalp.

### Retinoid tablets

Retinoid tablets (vitamin A) may be prescribed to help lessen the scaling in some severe forms of ichthyosis.

Pregnancy must be avoided during (and for a few months or years after) treatment with retinoids depending on the type of retinoid prescribed.

### Sunscreens

Always use a high factor sunscreen product, at least factor 25. Those formulated for children and for sensitive skin are more suited to 'fragile' skin. You should always do a patch test first, ask your pharmacist for samples to try before purchasing a sunscreen product. Some brands are available on prescription.

## Handy hints

You may find some of the following advice useful:

- Find an emollient you or your child likes for washing and regularly moisturising the skin.
- Apply emollients to damp skin to trap in the moisture – ideally a few minutes after having a bath or shower.
- Moisturise using long, smooth, soothing strokes (in the direction of hair growth).
- Take a pot of moisturiser to work, college or school and moisturise dry, scaly skin during the day.
- If scales are very thick, apply your emollient on the affected area in a circular motion to loosen scales. Thick scales can be removed with a pumice stone but it is better to prevent buildup with a regular emollient routine.
- Moisturise scalp and carefully comb hair to help remove scales.
- Contact the ISG to talk to other people living with ichthyosis to share more hints and tips.



## Problems obtaining your prescription?

If you are experiencing problems obtaining your prescribed skin care treatment, the Ichthyosis Support Group has letters you can print and take to your healthcare professional:

[www.ichthyosis.org.uk/leaflets](http://www.ichthyosis.org.uk/leaflets)

## Where can I find out more about ichthyosis?

Contact the Ichthyosis Support Group for information, advice, details on useful products, and to connect with other people to share experiences.

Ichthyosis Support Group

PO Box 1242

Yateley GU47 7FL

Email: [isg@ichthyosis.org.uk](mailto:isg@ichthyosis.org.uk)

Web: [www.ichthyosis.org.uk](http://www.ichthyosis.org.uk)

Phone: 0845 602 9202

There are a number of online forums about ichthyosis where experiences can be shared:

 [facebook.com/ichthyosissupportgroup](https://facebook.com/ichthyosissupportgroup)

 [twitter.com/ISG\\_Charity](https://twitter.com/ISG_Charity)

Ichthyosis information accredited by:



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