

Ichthyosis vulgaris and Acquired ichthyosis

What is ichthyosis vulgaris?

'Ichthyosis' is a group of skin conditions characterised by dry, scaly skin and ichthyosis vulgaris (IV) is the most common form of inherited ichthyosis. The severity of IV varies widely from very mild cases of dry skin to severe areas of thickened skin and scaling over the body, arms, legs and face. The condition runs in families, often along with eczema and/or asthma and/or hay fever.

IV usually presents in early childhood but an individual's skin will usually appear normal at birth. The lifelong condition may improve with age and often subside in the warmer summer months, while becoming more severe when the weather is cold and dry in winter.

What is acquired ichthyosis?

Acquired ichthyosis is a form of ichthyosis which generally appears in adults. It commonly results from natural dryness of the skin in our later years of life but it may rarely be associated with an underlying disease. There are a number of conditions associated with acquired ichthyosis including nutritional deficiencies, metabolic factors and side effects from some medications. A short list of examples is given below but this is by no means all-inclusive. The severity of acquired ichthyosis largely depends on the underlying cause but if the cause is removed or the condition is treated, the ichthyosis should improve.

Conditions associated with acquired ichthyosis

- Advancing age
- Environmental factors: excessive washing, excessive heat, cold windy weather, chlorine from swimming pool, air conditioning
- Metabolic factors eg thyroid disease, excessive weight loss
- Vitamin A deficiency
- Treatment with nicotinic acid eg to lower cholesterol
- Extremely rarely, other serious conditions

What are the signs?

The signs of IV and acquired ichthyosis are quite similar. The skin is seen to be dry and flaky with the build-up of fine, pale scales in certain areas, usually the outer parts of arms, lower legs and across the abdomen. In IV the palms and soles have more noticeable creases and wrinkles of the skin. Eczema may also develop in acquired ichthyosis and IV, making the skin itchy and red.

How are the conditions diagnosed?

Symptoms vary from person to person but there are a number of common signs that doctors will check to identify IV or acquired ichthyosis.

Unlike most ichthyoses, IV and acquired ichthyosis are not usually present at birth. IV typically develops in early childhood, while acquired ichthyosis develops later in childhood or adulthood.

Usually the diagnosis can be made by an experienced doctor simply looking at the skin.

If the diagnosis is not clear, sometimes a blood test or skin sample (biopsy) are needed. In IV, a skin biopsy can be done to check for a protein called profilaggrin which is reduced or absent in these patients. A skin biopsy is usually done by a dermatologist rather than a GP or paediatrician.

Blood tests may be needed to check for conditions associated with acquired ichthyosis.



People who care about ichthyosis

Why are ichthyosis vulgaris and acquired ichthyosis sometimes misdiagnosed?

Occasionally patients are not diagnosed for months or even years as symptoms may come and go and dry skin can be a symptom of other skin conditions, such as eczema or rare forms of immune deficiency. If the ichthyosis is mild and not troublesome for the patient, no diagnosis is requested. But if the ichthyosis is troublesome it is important to ensure an individual has additional tests if IV is suspected in order to rule out other conditions or else identify the underlying disease.

Managing the conditions

As yet, there is no cure for IV but the prognosis for the condition is generally very good. Symptoms tend to come and go but continual care with moisturisers should help keep the skin as healthy as possible. It may also be necessary to avoid environmental conditions that can worsen ichthyosis eg excessive washing, dry, cold and windy weather. Success of the treatments will depend on the individual and the severity of their condition. What works for one person may not work for another and patients will need to consider different options to find the best treatment for them. Treatment surrounding diseases which result in acquired ichthyosis should be tailored to that specific condition and are not covered in this leaflet. Individuals should consult their doctor for further guidance on treating the underlying condition, though they may find the following information useful in managing their ichthyosis symptoms.

Moisturising creams and skin treatment

It is very important that the skin is kept moisturised at all times and that moisturising creams or ointments are applied frequently. Moisturising agents should be perfume-free and without additives to avoid any allergic reactions. The use of bath oils (not bubble bath or soap) is helpful so that bathing does not cause drying or irritation of the skin and soaking in the bath tends to be more beneficial than taking a shower.

Emollients and keratolytic creams which contain urea, and/or lactic acid (see product's ingredients label for these) can be useful to remove scales. However these should be avoided if the skin is particularly itchy or red because they may irritate the skin.

In very severe cases antibiotics can be given to treat secondary infections.

More specific information on caring for the skin in IV and other forms of ichthyosis can be found on the Ichthyosis Support Group website at www.ichthyosis.org.uk.

Personal care

Scaling and peeling skin, particularly in the most visible areas such as the scalp or face, can be difficult for building self-confidence so individuals may need extra support. General practitioners need to understand the psychological impact of the condition and provide adequate support to the family. This may require the involvement of other healthcare professionals.



People who care about ichthyosis

How is ichthyosis vulgaris inherited?

IV is a genetic disorder so the condition runs in families. IV is an autosomal semi-dominant form of genetic disorder. This means that an affected child needs only one copy of the faulty filaggrin gene to have the condition, but individuals with two faulty copies have more severe IV than individuals with one faulty copy. Since each parent provides half their child's genes, there is a 50:50 chance that a parent with IV can pass the condition onto their children if their partner is not affected.

Filaggrin is the genetic code for a protein called profilaggrin. Profilaggrin is important in building the skin barrier, to protect the body from irritants and allergens in the outside world. It also holds moisture in the skin. In IV patients, faulty filaggrin produces less profilaggrin and that is why the skin is dry. IV patients may also develop allergic diseases (eczema, asthma, hay fever and some food allergies) because the skin barrier is faulty.

Acquired Ichthyosis cannot be passed down in families but the underlying condition behind the disorder may in some cases. Patients should consult their doctor for further information.

Further help

There are a number of online forums about Ichthyosis where individuals can share their experiences and detail their own treatment recommendations. Not everything will work for everyone but these forums are a good place both to receive and offer support.

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

To find out more about the ISG or become a member please get in touch in one of the following ways:

By Phone or Fax:

Tel: **0845 602 9202** Fax: **0560 343 8046** (on request)

By Email:

isg@ichthyosis.org.uk

By Post:

Ichthyosis Support Group

PO Box 1242, Yateley GU47 7FL

Facebook:

facebook.com/ichthyosissupportgroup

Twitter:

twitter.com/ISG_Charity