ICHTHYOSIS

WHAT IS IT?

Ichthyosis is continual and widespread scaling of the skin.

There are several forms of inherited ichthyosis. Some are severe rare conditions, while others are more common and milder.

There is an acquired form linked to other medical problems such as kidney disease.

ICHTHYOSIS VULGARIS

This is the most common form of inherited ichthyosis.

It is usually quite mild and develops in early childhood with fine, light-grey scales and roughness on the arms and legs.

It affects one in every 250 people.

It is inherited by autosomal dominant transmission, with a risk for each child of 50 per cent.

X-LINKED RECESSIVE ICHTHYOSIS

This form occurs only in males and varies in its severity.

It develops in infancy, with tan or grey scales on the limbs and across the trunk. It may affect the ears and face and results in scales that appear to be stuck on like stamps.

It is inherited by X-linked recessive transmission.

NON-BULLOUS ICHTHYOSIFORM ERYTHRODERMA

This is one of two congenital conditions where a shiny, yellow film called collodion membrane stretches across the skin of a newborn baby.

The collodion membrane dries out and sheds within the first week of life and the skin remains red and has fine, white scales affecting all of the skin surface.

It affects one in every 300,000 births.

In severely affected children, the eyelids may be pulled outwards and there may be some mild scalp hair loss and tightness of the fingers.

It is an autosomal recessive condition.

LAMELLAR ICHTHYOSIS

This is the less common form of congenital ichthyosiform erythroderma.

The skin is less red but the scaling is larger, perhaps darker and more adherent, or stuck down.

It is autosomal recessive.

BULLOUS ICHTHYOSIFORM ERYTHRODERMA

This rare form is also known as bullous ichthyosis.

At birth the skin seems to be fragile and may be blistered.

Scaling and thickening becomes more obvious through childhood.

Warty skin changes develop around the creases of the joints. Skin infections are quite common and can lead to a characteristic odour.

This is an autosomal dominant disorder but in at least half of cases neither parent is affected, therefore the child has developed a new genetic fault.

HARLEQUIN ICHTHYOSIS

Harlequin ichthyosis is a very severe but extremely rare form.

Incidence is approximately three cases per year in the UK.

It is evident at birth because the newborn baby looks like a harlequin costume, with very thick scaling all over.

NETHERTON’S SYNDROME

The incidence of this condition is not known but it is probably in the region of one in every 200,000 births in the UK.

The newborn child is very red and has scaly or peeling skin.

It is autosomal recessive.

REFERENCES
