but again there may be side effects. Discuss the ins and outs of these therapies with your skin doctor before trying them.

Drugs by mouth – Synthetic vitamin A drugs (called retinoids e.g. acitretin or isotretinoin) can be very beneficial in decreasing the level of scaling but have little impact on the redness of the skin. They can have many side effects and the benefits and risks need to be discussed with your skin doctor. Close supervision of treatment and avoidance of pregnancy is vital if these drugs are used.

Hand splinting – If the fingers develop contractures and cannot be fully straightened, physiotherapy and night time splinting may help.

Psychological support/Counselling – Even the strongest individual will find severe ichthyosis difficult to cope with at times. Parental rejection, problems with schooling and making friends, and depression may all occur. Having a supportive GP and dermatologist is important. Professional counselling especially by those that deal with disfigurement can be very useful. Ask your doctor about locally available services. If depression is severe a course of medication can help one to get through a bad patch.

Lamellar ichthyosis is very rare and you may feel that you are on your own. Fortunately the **Ichthyosis Support Group** exists in the UK, who will provide you with psychological support and practical information about how best to deal with this condition. This in many ways is the best information as it comes from people who have been through exactly the same problems that you are currently experiencing.

* A note on classification of the subtypes of ichthyosis.
Historically the different types of ichthyosis were described or named according to the actual appearance of the skin. This system is still used in the UK but it has its limitations and in fact some European and American dermatologists use rather different names. You may for instance come across the term “ichthyosis congenita types 1-4”. We feel this latter naming system further confuses the issue for patients and parents. In the future the best and indeed only way to classify these conditions is by working out the abnormal genes which are after all the key underlying problem. Until research has uncovered these causes we prefer to stick to the names used here.

Ichthyosis Support Group
October 1998

Lamellar Ichthyosis

What is it?

This is an extremely rare skin disorder characterised by abnormal scaling and shedding of the skin. It is estimated to occur in 1/600,000 births. Most affected individuals will be born as a “collodion baby”.

What does it look like?

The skin appears to have large plate-like scales which often are rather dark or brownish in colour. The scaling often involves all of the skin although the face may show milder changes. The condition usually appears in the first few days of life, lasts lifelong and can be very severe.

Are there different types of Lamellar ichthyosis?

Yes there are two variations to the above appearance.

Limited lamellar ichthyosis: In this very rare condition there appear to be areas of normal looking skin and areas of skin with abnormal plate-like scaling. The amount of normal skin may vary at any one time.

Erythrodermic lamellar ichthyosis: The typical scaling of above may be accompanied by an underlying red or inflamed skin (called “erythroderma”).

What is it caused by?

It can be caused by a variety of different genetic or inherited abnormalities that affect normal skin shedding. To date only one gene has been discovered (called the “transglutaminase” gene). This gene makes an enzyme (chemical) in the very outer layer of the skin which is vital to allow normal shedding of skin cells. Abnormalities in this gene account for less than half of individuals with Lamellar ichthyosis so the other cases presumably have abnormalities in different genes, as yet unidentified.

How is it inherited?

Lamellar ichthyosis is usually inherited in an "autosomal recessive" fashion. This means that each parent has normal skin but is a carrier for one single abnormal gene. One in four children will inherit two abnormal genes (one from each parent) and therefore develop the condition. Very rarely Lamellar ichthyosis can be inherited in an "autosomal dominant" fashion. This means that one in two children will be affected as only one abnormal gene is needed to cause the condition. Normally one of the parents will be affected.

Is it catching or contagious?

No, absolutely not.

Can it be diagnosed pre-natally if I want to have another child?

Unfortunately most cases of Lamellar ichthyosis can not be diagnosed pre-natally. However in the future, research is likely to uncover the underlying genetic causes so pre-natal diagnosis may become a very real possibility, although it will only be available in one or two specialist centres.

What are the complications?

Overheating – Severe scaling of the skin prevents normal sweating so hot weather or vigorous exercise can cause problems.
Constriction bands – Very rarely in early childhood tight bands of skin form around the fingers or toes which can prevent normal blood supply to the finger tips and end of the toes.
Eye problems – Ectropion is the term used by doctors to describe when the eyelids are pulled outwards by the tightness of the skin. This makes the eyelids (usually just the lower one) look red and the eye can be more prone to drying and irritation.
Hand problems – Untreated, the skin of the palms may become thickened, tight and prevent normal bending and straightening of the fingers ("contractures").
Hair loss – Severe scaling of the skin of the scalp can lead to patchy loss of hair but this is rarely permanent.
Psychological – The severe plate-like scaling of the skin and the appearance of the eyes can cause severe psychological problems for both parents and the child.

Unfortunately society's reaction to the appearance of ichthyosis is often hostile and unsympathetic. Whilst this stems from ignorance it is a problem that cannot be underestimated. Two of the most difficult times are when a child starts school and during teenage years. Staring and teasing will occur. Having said that, there are a number of individuals with severe ichthyosis who have adjusted well and have managed to lead relatively normal lives, although this requires a strong personality and plenty of family support.

Are there any internal abnormalities we should keep an eye out for?

No, Lamellar ichthyosis is confined to the skin.

Is there a cure for Lamellar ichthyosis?

No, currently there is no available cure. However there are some treatments that can help.

Treatment:

Moisturisers (also called emollients) and bath oils are a vital part of therapy and will need to be used lifelong. They make the skin feel more comfortable, look less dry and flaky and prevent cracking. These creams vary in their greasiness and some contain urea or lactic acid. It is important that you try a number of these to see which works best for you and your child.
Particular attention should be paid to massaging cream regularly into the palms and under the eyes to prevent tightness of the skin here.
Prevent overheating (especially in children) – Avoid prolonged exposure to the sunshine, exercise in the middle of the day and wear cool loose fitting clothes. If overheating does occur the use of damp sponges will help.
Eye treatment – Use of moisturising eyedrops (artificial tears) can be useful if the eyes become dry. Surgery to the eyelids is not always the right answer for pulled down eyelids as it often will recur.
Constriction bands – These are fortunately very rare indeed but if they occur surgical release can be used.
Specific creams – Synthetic vitamin D creams and synthetic vitamin A-derived creams have been used in a different skin condition called psoriasis. Some patients with severe ichthyosis may find these helpful but they can have side effects especially if large areas of the skin are being treated. Creams containing salicylic acid have also been used