

## A Review on Different Types of Ichthyosis and Management

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### ABSTRACT

Ichthyosis is one of the most devastating of the genodermatoses. Neonates usually die within the first few days of life from infection or dehydration related complications. Prenatal diagnosis remains difficult but may be possible in high risk pregnancies by performing a foetal skin biopsy or by three-dimensional ultrasonography. Ichthyosis may be either inherited or acquired. Inherited Ichthyosis is usually apparent during the first year of life, often at birth, and continues to affect a person throughout life. Acquired Ichthyosis may occur as the result of medical problems including hormonal, inflammatory or malignant disorders. Managing the signs and symptoms the treatment is targeted which includes creams, lotions, or ointments to relieve dryness.

**Keywords:** Ichthyosis, Genetic disorder, General Systemic Diseases

### DEFINITION

The word "Ichthy" comes from the Greek word for fish. This condition is called "ichthyosis" because the thickened skin sometimes has the appearance of fish scales. The ichthyoses are a family of genetic diseases characterized by dry, thickened, scaling skin. This condition is called "Ichthyosis" because the thickened skin sometimes has the appearance of fish scales. Because each form of Ichthyosis is rare and there is an overlap of clinical features among disease types. Ichthyosis may be either inherited or acquired. There are approximately 28 recognized forms of Ichthyosis and related skin types<sup>[1]</sup>.

### Signs and symptoms<sup>[2]</sup>

Specific signs and symptoms depend on the particular form. The most common signs include dry scaly skin, redness, blistering, or

excessive skin shedding. Symptoms include itching, overheating, and pain<sup>[2]</sup>.

### Causes of Ichthyosis<sup>[2] [3]</sup>

It is seen to be a Genetic disorder that is passed from parent to child or that occurs spontaneously. Certain soaps or lotions containing certain scents or perfumes may also trigger this.

Use of harsh soaps or detergents make the skin dry and are known to cause Ichthyosis.

Cold weather is also an important factor which increases dry skin and leads to Ichthyosis.






Acquired Ichthyosis is not inherited and occurs for the first time in adulthood. It is usually associated with some general systemic diseases, such as under active thyroid, sarcoidosis, lymphoma, generalized cancer or HIV.

It may be provoked by certain medications (such as kava, nicotinic acid and hydroxy-urea).

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### DIFFERENT TYPES OF ICHTHYOSIS

Types of Ichthyosis according to Foundation for Ichthyosis & Related Skin Types (FIRST)<sup>[1]</sup>

Type of Ichthyosis	Picture <sup>[4][5]</sup>
Epidermolytic Ichthyosis (previously called Epidermolytic hyperkeratosis). Characterized by thick, often spiny dark scales and skin that may blister easily following trauma.	
Ichthyosis vulgaris: Characterized by mild skin scaling and dryness. It is another form, recessive X-linked Ichthyosis, are relatively common and appear similar.	
Congenital ichthyosiform erythroderma: Characterized by red skin and fine scales.	
Localized Ichthyosis: Characterized by thick or scaly skin that is localized to particular regions such as the palms of the hands and soles of the feet.	
Lamellar Ichthyosis: Characterized by large, plate like scales and thickening of the skin.	

Ichthyosis Vulgaris <sup>[1]</sup>	<p>Ichthyosis vulgaris is one of the more commonly seen types of Ichthyosis. It appears in approximately 1 in 250 individuals. It often goes undiagnosed because people who have it think they have simple “dry skin” and never seek treatment. In Ichthyosis vulgaris, the skin cells are produced at a normal rate, but they do not separate normally at the surface of the stratum corneum, the scaling is usually limited to the forehead and cheeks. Ichthyosis vulgaris is treated topically with moisturizers, and keratolytics. It is not considered severe enough to warrant use of oral synthetic retinoids.</p>
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<p>Epidermolytic Ichthyosis (EI)<sup>[1]</sup></p>	<p>Epidermolytic Ichthyosis also called Epidermolytic hyperkeratosis or EHK) (bullous congenital ichthyosiform erythroderma) is rare, occurring in approximately 1 in 300,000 births. Scales tend to form in parallel rows of spines or ridges. The skin may be fragile and may blister easily following injury. Over time there is a gradual decrease in the blistering, but an increase in the severity of the thickness and scaling. A generalized redness of the skin (erythroderma) is present in some individuals. Skin infections and heat intolerance can be common problems. Treating EI can be a challenge. The medications that are used to help remove the excess thickened skin (topical keratolytics or oral retinoids) often remove too much scale, leaving very fragile underlayers exposed. Barrier repair creams, containing ceramides, cholesterol, petrolatum or lanolin, can help along with topical or systemic anti-bacterial agents. Keratolytics and oral retinoids should be used with caution.</p>
<p>X-Linked Ichthyosis</p>	<p>X-linked ichthyosis is one of the more commonly seen forms of ichthyosis. It occurs in 1 in approximately 6,000 births, can range from mild to severe, and occurs only in males. In X-linked ichthyosis, the skin cells are produced at a normal rate, but they do not separate normally at the surface of the stratum corneum.</p> <p>Typically the face, scalp, palms of the hands and soles of the feet are unaffected, while the back of the neck is almost always affected. X-linked ichthyosis frequently improves in the summer. X-linked ichthyosis is treated topically with moisturizers and keratolytics. Cholesterol containing ingredients may also improve scaling. X-linked ichthyosis is not considered severe enough to warrant use of oral synthetic retinoids.</p>

### Autosomal Recessive Congenital Ichthyosis - lamellar type<sup>[1]</sup>

Autosomal Recessive Congenital Ichthyosis (ARCI) – lamellar type (or classical lamellar Ichthyosis) is one of the more commonly seen types of Ichthyosis. It is one of the most severe forms, and it occurs in approximately 1 in 300,000 births. Recessive genes cause lamellar Ichthyosis, similar to blue eyes. In lamellar Ichthyosis, the skin cells are produced at a normal rate, but they do not separate normally at the surface of the stratum corneum and are not shed as quickly as they should be. The result is a build-up of scales. The entire body is covered with broad, dark, plate-like scales separated by deep cracks. People with lamellar Ichthyosis may experience a condition called ectropion (a turning out of the eyelids to expose the red inner lid). People with lamellar

Ichthyosis may also have thickened nails and hair loss due to the thickness of the scales on their scalp. They may also have reddened skin (erythroderma), thickened skin on the palms of the hands and soles of the feet, and decreased sweating with heat intolerance.

Lamellar Ichthyosis is present at birth. Many babies born with lamellar Ichthyosis are also “collodion babies” because a clear membrane (the collodion) may cover their bodies. The collodion is then shed within a few days to a few weeks. Sometimes described as having a shellacked appearance, these newborns have skin which is taut, dark and split. Often the eyelids and lips are forced open by the tightness of the skin, and there may be contractures around the fingers. Problems with temperature regulation, water loss, secondary infections, and systemic infection

can occur in the newborn with lamellar Ichthyosis. Lamellar Ichthyosis is typically treated topically with moisturizers and keratolytics. Creams with high concentrations of alpha-hydroxy acids are commonly used. Lamellar Ichthyosis may be treated systemically with oral synthetic retinoids (Accutane or Soriatane). Retinoids are used only in severe cases due to their known bone toxicity and other complications.

#### Netherton Syndrome<sup>[1]</sup>

Netherton syndrome is a rare syndrome characterized by red, scaly skin, short brittle hair and a predisposition to allergies, asthma and eczema. Newborns with Netherton syndrome have reddened skin (erythroderma) and, occasionally, a thick shell-like covering of skin (collodion membrane). They may also be born prematurely. Trouble gaining weight during infancy and childhood is common. Atopic dermatitis (red, itchy patches of skin) may be present, and a cradle cap-like scale and redness may appear on the face, scalp and eyebrows. Unlike many of the Ichthyosis, Netherton syndrome produces too few layers of the outer skin, instead of two many layers. Current treatment options are limited to use of mild moisturizers containing petrolatum or lanolin and/or a barrier repair formula containing ceramides or cholesterol.

#### **Autosomal Recessive Congenital Ichthyosis – CIE type**<sup>[1]</sup>

Autosomal Recessive Congenital Ichthyosis (ARCI) – CIE type (Nonbullous Congenital Ichthyosiform Erythroderma) is considered

one of the more commonly seen types of Ichthyosis. Like lamellar Ichthyosis, CIE is rare, occurring in 1 in 300,000 births. Recessive genes cause it. In CIE, there is an overproduction of skin cells in the epidermis. These cells reach the stratum corneum (the outermost layer of skin) in as few as four days, compared to the normal fourteen. New skin cells are made faster than old cells are shed and build up in the stratum corneum and underlying layers. The severity and scaling of CIE varies. The scales on the face, scalp and torso are usually fine and white, but the scales on the legs can be large and plate-like (like the scales of lamellar Ichthyosis). The skin is often quite reddish beneath the scales. CIE is present at birth. Many babies with CIE were born as “collodion babies,” so called because a clear membrane (the collodion) covers their bodies. The collodion is then shed within a few days to a few weeks.

After the membrane is shed, dry red skin is revealed. Often the eyelids and lips are already forced open by the tightness of the skin, and there may be contractures around the fingers. CIE is treated topically with moisturizers and keratolytics. Creams with high concentrations of alpha hydroxy acids are commonly used. CIE can be treated systemically with oral synthetic retinoids (Accutane, Soriatane). Retinoids are only used in severe cases due to their known bone toxicity and other complications.

**Diagnosis**<sup>[2]</sup>:The diagnosis is based on the results of a medical history and physical exam. In some cases, doctors use blood tests or skin biopsies to diagnose the condition or identify the particular form.

#### **Complications**<sup>[3]</sup>:

<b>Overheating</b>	Severe scaling of the skin prevents normal sweating so hot weather or vigorous exercise can cause problems.
<b>Constriction bands</b>	Very rarely in early childhood tight bands of skin forms around the fingers or toes which can prevent normal blood supply to the finger tips and end of the toes.
<b>Eye problems</b>	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”
Hairloss	Hair loss is 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.

#### **GUIDELINES FOR INDIVIDUALIZED THERAPY<sup>[6]</sup>**

##### **Congenital Ichthyosis (neonatal intensive care)**

- Monitor fluid balance, electrolytes; watch for skin infections
- Incubator with high humidity but somewhat lower temperature; remember infants may have trouble with sweating;
- monitor temperature
- Never bathe without relubricating; apply nonmedicated cream thinly 6-8 times a day
- Ectropion management by ophthalmology
- Watch for flexural contractures
- Check vision and hearing; get neurologic evaluation
- Put family in touch with Ichthyosis self-help group

##### **Newborns and infants**

- Bathe twice daily with sodium bicarbonate
- Rub lightly with soft washcloth or microfiber towel
- Apply nonmedicated cream 2-3 times daily
- No ura or salicylic acid
- Ectropion management by ophthalmology; cleaning of external ear by otorhinolaryngology
- Physical therapy to avoid flexural contractures

##### **Children**

- Bathe twice daily with sodium bicarbonate
- Rub with soft washcloth, microfiber towel, or pumice stone
- Apply 5% urea cream twice daily
- Treat scalp with stronger urea cream 7%-10%
- Ectropion management by ophthalmology; cleaning of external ear by otorhinolaryngology
- On warm summer days, measure temperature, force fluids, cool environment, no occlusive ointments; play outside earlier or later, not in midday
- No vacations in warm climates

##### **TREATMENT<sup>[3]</sup>:**

Because there is no cure for Ichthyosis, treatment is targeted at managing the signs and symptoms. Treatment may include creams, lotions, or ointments to relieve dryness. Lengthy bathing in salt water or preparations containing salicylic acid (aspirin) or urea may also ease scaling. For more severe cases, doctors may prescribe vitamin A derivatives called retinoids<sup>[3]</sup>.

<b>Moisturisers and bath oils</b>	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,
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	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.
<b>Prevent overheating</b>	Prevent overheating used 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.
<b>Eye treatment</b>	Eye treatment Use of moisturising eye drops (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.
<b>Specific creams</b>	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 but again there may be side effects. Discuss the ins and outs of these therapies with your skin doctor before trying them.
<b>Drugs by mouth and Hand splinting</b>	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/Counseling/patient counseling<sup>[3]</sup>:**

- 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 counseling 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.

### **CONCLUSION**

Ichthyosis is a mutilating disease, having numerous social and psychological effects. The

more severe forms of ichthyosis can be able to cause many other complicated diseased conditions. When the skin loses moisture, it becomes dry, tight, and rigid. This rigidity makes moving uncomfortable as it make the skin to crack and break open. Extreme thickening on the skin on the soles of the feet makes walking difficult for many individuals, and cracking around the fingers can make even simple tasks difficult and painful. In some types of ichthyosis the skin is very fragile and will rub off with the slightest scrapes. These cracks and scrapes may lead the skin open to infections which further make the disease more complicated.

Few patients with ichthyosis have trouble closing their eyes completely. This is due to the surrounding skin becomes so tight. This condition, called ectropion, causes the eyelids



to turn outward, exposing the red inner lid and causing continuing irritation. If it is left untreated, it leads the damage to the cornea which further leads to impaired vision.

Although it is a non-curable disease it can be controlled by various treatment procedures like moisturisers and bath oils for preventing overheating. Skin treatment with specific

creams like synthetic vitamin D and derivatives of vitamin A can be used. Eye treatment with moisturising eye drops like artificial tears can be useful when the eyes become dry. Drugs by oral route like retinoids such as acitretin or isotretinoin can benefit in decreasing the level of scaling and others like hand splinting with physiotherapy may reduce contractures.

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