

## INTRODUCTION

Congratulations on your new baby! You just joined a very select, very special club... and you didn't even have to apply! In fact, until a few days ago, you probably had never even heard of Ichthyosis. Don't worry, none of us had, either. You are probably feeling terrified right now, and overwhelmed, and worried that you aren't going to be able to care for a newborn, let alone one with special needs.

You are not alone.

Every one of us here has been through the same thing you are going through, which is why we put together this guide. It covers the “Frequently Asked Questions” from the Ichthyosis Forum, and is a very brief introduction in how to care for your child with Ichthyosis. This guide covers very general issues that pretty consistently apply to all types of Ichthyosis. Of course there are issues that are unique to specific types of the condition, and for those issues this Board is an amazing resource. The information contained in this guide represents the collected wisdom of caregivers and adults with the condition; in no way is it intended to take the place of your child’s doctors.

Please feel free to print a copy of this guide or bookmark it for quick reference.

## TABLE OF CONTENTS

### **PART I GENERAL CARE GUIDE**

A. CARING FOR A NEW BABY WITH ICHTHYOSIS.....	4
B. POST-COLLODION CARE.....	7
1. DRYNESS.....	8
2. ITCHING.....	9
3. FLAKING/SCALING.....	10

### **PART II SPECIAL ISSUES**

A. CRACKS.....	12
B. EARS.....	13
C. EYES.....	15
D. SCALP.....	16
E. REMOVING LOTION FROM ... EVERYTHING.....	17
F. ILLNESS/FEVER.....	24
G. TEMPERATURE ISSUES/SWEAT IMPAIRMENT.....	25
H. NUTRITION .....	27
I. DEALING WITH BLISTERS (EHK).....	29

### **PART III GLOSSARY OF PRODUCTS**

A. LOTIONS AND CREAMS.....	33
B. DESCALERS.....	36

C. EAR PRODUCTS..... 36

D. EYE PRODUCTS..... 37

E. CLEANSERS..... 37

F. SHAMPOO..... 38

G. ANTIBACTERIALS AND ANTIFUNGALS..... 39

H. SUNSCREEN..... 39

**PART IV**

RESOURCES..... 40

**PART I**  
**GENERAL CARE GUIDE**

**A. CARING FOR A NEW BABY WITH ICHTHYOSIS**

Your baby may have been born with a "collodion" shell at birth. The collodion membrane is a thin sheet of dry skin that covers the baby's entire body. In a Harlequin baby, the skin is much thicker, and the large scales are referred to as "plates." The collodion and plates can look frightening to a new parent, but don't worry -- it's just skin! When the baby was growing inside the mother, his skin was growing, too. In some forms of Ichthyosis, this skin grows too fast, and when the baby is born it has built up because it couldn't dry and flake off. The skin can build up so much that the baby is unable to close his mouth or his eyes, even turning the eyelids inside out. This can look scary to you, and may be painful to your child. In serious cases, the tight skin can prevent proper development and deform fingers, ears, and toes -- but this is not common.

When a collodion baby is finally born and is exposed to air for the first time, this built-up skin dries out and flakes off. Each hospital has a different protocol to care for your infant. Many hospitals will treat your child as a burn-patient, and you will have to follow very strict hygiene rules to prevent infection. They will do this because your baby is at risk of infection when his skin first cracks and peels off. You may not get to hold your infant for a few days, or even touch him. If your child is a newborn and you are at a hospital that has never dealt with a collodion or Harlequin birth before, **CALL THE ICHTHYOSIS FOUNDATION (F.I.R.S.T.)** in the U.S. (215.619.0670). They will put you and your doctors in touch with specialists who are familiar with caring for the special needs of an infant with Ichthyosis. F.I.R.S.T. will also direct you to other parents in your region for

emotional support. It is a fantastic organization that is devoted to helping you and your child.

Once your child is released from the hospital, you will be responsible for your baby's skin on top of general newborn care. This can be very overwhelming, especially if this is your first child. In addition to being sleep deprived and stressed, you may hear horrible comments from family, friends, and strangers that your baby "would

*We've All Been There*

*"I remember our first Christmas being so excited to send our first Christmas cards with our new baby (born with Harlequin). We spent all morning setting up that perfect shot. Our daughter was adorable in her little holiday outfit. When I went to pick up the pictures I was handed an empty envelope with a note: "Did not fill. Baby red."*

*I was crushed."*

have been better off if he had just died" or shrieks of "what is wrong with your baby?!" **There is nothing wrong with your baby.** The problem is with THOSE people, not you, and definitely not your child. Your child is beautiful, and will give you more joy (and more headaches) than you can ever imagine. **Most people** with Ichthyosis lead normal, happy, productive lives. They marry, have children, and contribute to society. It is normal to worry, but try not to project the worst that could happen to your child. If people in your life are making this stressful time even more stressful, you may need to cut contact with them for a while. Your child needs love, lotion, and lots of support. As a parent, your job is to keep the toxic people out of your child's life.

After you are discharged from the hospital you will have to figure out not only how to care for a new baby, but how to care for one with Ichthyosis. There is a learning curve, so do not feel discouraged. It can take a few weeks for the collodion to completely come off, and once it does your child's skin will still undergo normal fluctuations depending on the weather, humidity, and illness. One day your baby may look great, the next day dry, flaking, and bright red, or even brown and "dirty" from scale build-up. The constant changes in your child's appearance can be demoralizing and frustrating. You may find yourself questioning your ability to take care of your baby, and find yourself depressed. In that first year, try to focus on getting to know your baby and finding a skin-care routine that keeps your child comfortable and is manageable for you. **Your goal the first year shouldn't be perfect skin**, it should be getting your "sea legs," i.e. becoming comfortable on your new role as parent *and* skin care guru. It's ok to feel sad, and lonely, and angry. Find another caregiver in your area or on this Board to vent to.

Collodion care really isn't that different from post-collodion care (see the Section B, below). You are going to want to keep your child hydrated (either through extra breast-feeding sessions or extra bottles) and keep your child's skin moisturized. The main difference between collodion care and later skin care is the risk of infection -- a newborn infant naturally has greater risk of infection, and when you add in Ichthyosis it can be very hard on your baby's body. **Make hand washing an absolute rule whenever someone enters your home, and again before**

### **TIP:**

One mother used disposable rubber gloves each time she applied her child's lotion, changing gloves before she dipped back into the lotion tub. This helped dramatically reduce the risk of infection that first year.

**anyone handles your infant.**

Some children have trouble early on getting enough nutrients by mouth and have to be supplemented with a feeding tube. Your pediatrician should closely monitor your baby's growth and question you about your baby's feeding schedule. It is helpful to keep a log of how often your baby eats and how many ounces (use a scale before and after breastfeeding to note how many ounces if you are not using a bottle). Children with Ichthyosis tend to be small because their bodies spend so much of their calorie intake on skin growth. That said, your baby still should follow a growth curve that is normal for *him*. You may need to see a pediatric nutritionist if your child's growth does not follow a normal curve.

Many physicians are not familiar in caring for a child with Ichthyosis, so you will need to be very aggressive in your child's care. **DO NOT BE AFRAID TO ASK QUESTIONS.** Many parents are overwhelmed by their child's doctor, who is, after all, a medical expert. When it comes to a rare disease, though, most doctors are just as unfamiliar with the condition as you are. Find a doctor who is willing to learn about the condition with you, who isn't afraid to call the experts and ask questions. If your doctor doesn't seem willing to do the extra work to help care for your child, find a new doctor. Caring for your child should be a team effort.

## **B. POST-COLODION CARE**

The hardest part of caring for an infant with Ichthyosis is knowing what your child is trying to tell you. This can lead to over-lotioning or over-zealous scale removal. **Just take your time, learn what is normal for your child, and be willing to try new things.** What works for one child may not work for you. What worked last week may not work this week. The key to Ichthyosis care is **flexibility**.

The main symptoms of all types of Ichthyosis are (1) dryness, (2) flaking, (3) itching, and (4) skin build up (called “scaling”). We will cover typical care for each of these issues in turn.

## **1. DRYNESS**

Although there are many different presentations of Ichthyosis, all of them share in common an “impaired skin barrier.” That means the skin layer is broken -- think of it like a dry, cracked river bed. This “broken” layer leads to excess water loss and a feeling of dry skin. Dry skin causes the other main symptoms of Ichthyosis (flaking, itching, and scaling). Treating dryness is critical to help ease the other three problems.

There are three main ways to treat dryness: (1) increasing hydration (breast milk/formula for infants, while water or sugar free sports drinks are best for older children), (2) setting up humidifiers in the rooms where your child is most of the day (usually family room and bedroom) and (3) adjusting the number of baths and/or the number of lotion applications to keep your child comfortable.

Each child is unique, and some children need lotion applied more often than others. Most kids seem to be comfortable with two or three lotion applications a day. Apply a layer of thick lotion (Aquaphor/Eucerin) in the morning, at lunch, a touch-up mid-evening if needed, and a thick layer of lotion after bath. It takes time to learn what is best for your child -- too much lotion can encourage bacterial and fungal growth, while too little lotion leads to itching and increased flaking. Remember, be patient and experiment.

Generally you should bathe your child once a day unless you live in a dry climate or your child is scratching/complaining of itching. For some children, bathing too often can actually make them feel more dry than skipping a bath. **DO NOT BE AFRAID TO EXPERIMENT.**

You will not hurt your child. If you try something and see an increase in flaking/scaling, then switch and try something else. Even though it may feel like homework, it helps in the beginning to keep a log of what products you are using, how often you are using them, and what results you are seeing (less flaking, no change, etc.). This will help you remember later if you have tried a product and if it worked.

Play around and figure out what works for you. Even though all of our children have Ichthyosis, EVERY PERSON IS DIFFERENT and will respond to different products and a different routine. Sometimes what worked yesterday isn't working today.

It's ok to go into a closet and scream into a pillow at that point. We've all been there. Just make sure you calm down before you sit down, revisit your skin care routine, and start making adjustments.

## **2. ITCHING**

As a parent, itching is the hardest issue to deal with. Many forms of Ichthyosis itch all of the time, with no relief. To know that your child, who you love with all your heart, constantly feels as if he or she has been bitten by bug bites can tear out your heart.

The first thing to remember is that this is your child's reality. Your child has never known what "normal" skin feels like. As hard as this is to accept, Ichthyosis *is* your child's normal. Because some of the treatments for itching are not good for your child long-term, it is often best not to treat a symptom unless it causes your child distress.

Adding a cup of (unscented) bath salts to your child's bath can help with itching. Other children find relief in oatmeal baths and products such as Aveeno.

**TIP:** You can grind regular oatmeal purchased from the grocery in a food grinder, place it in an old hose stocking with the top tied off, and drop it into the bath. This makes an inexpensive oatmeal bath without clogging your bathtub. Be sure to dispose of it after bath to prevent bacteria from growing or spreading.

If your child scratches constantly and cannot seem to get relief, cries or is irritable, scratches in his/her sleep, or complains of itching, then it is time to move past hydration and lotion and look into medications.

Steroid creams are often the first step physicians will take to treat itching. Even over-the-counter hydrocortisone can help relieve the itching, but all steroids have drawbacks with long-term use. In a child, steroids can stunt growth, thin the skin, and lower the body's ability to fight infection. Talk with your pediatrician and your dermatologist to weigh the pros and cons of long-term steroid use.

For severe itching (child scratches off large sections of skin, scratches until he/she bleeds, or is in obvious distress), Retinoids can be a last-resort treatment. The Ichthyosis Forum has a whole section devoted to the pros and cons of Retinoids, which you can review before talking with your child's doctor.

### **3. FLAKING/SCALING**

Removing extra skin is helpful not just for cosmetic purposes, but for medical purposes as well. Scales can grow bacteria, so removing them will help prevent infections. That said, you can be **too** ambitious in scale removal -- not every single scale must be removed every single time. As long as you are keeping your child's skin clean, bacteria will usually not be an issue.

All types of Ichthyosis, with the exception of EHK<sup>1</sup>, will benefit from gentle exfoliation. First, have your child soak in a warm (not hot!) tub of water for at least 10 minutes. Then gently rub your child's skin with your fingers, a wash cloth, exfoliating bath scrub, or a sea sponge, whichever your child seems to prefer. Generally you can remove more skin without soap than with it -- the oils in your fingers will mix with the residual lotion on your child's skin and help "roll" the excess skin off. Remove the loose scales gently, then wash with a gentle cleanser or soap to remove the dirt and bacteria on your child's body. Pat your child dry, then lotion over the scales that did not come off. **DO NOT PICK AT SCALES THAT SEEM STUCK.** They will come off when they are ready. Picking at scales can cause them to bleed and introduce bacteria into your child's skin.

For children with severe scale build-up, more intense exfoliation or descaling products may be required. See the sections below on scalp and foot treatments for advice on special issues.

If you choose to use a descaling product, follow the directions carefully. They often contain acids that loosen the scales by softening them. For some children, these products sting and cause more distress than the presence of scales. Some children are not bothered by descalers at all. See the product list below to understand the different creams preferred by different people.

So, to recap: **(1) LIQUIDS, (2) HUMIDITY, (3) EXFOLIATION, (4) LOCK IN WATER WITH LOTION, and (5) ALWAYS BE OPEN TO TRYING NEW THINGS.** This will be your routine until your child is old enough to care for himself/herself.

Trust me, you are going to get *very, very* tired of lotion.

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<sup>1</sup> See the section below on special issues in EHK

## PART II SPECIAL ISSUES

### A. CRACKS

Your child will experience cracks (also called “fissures”) on her hands and feet at some point in the year. Little hands tend to crack when the infant is teething and uses her hands to soothe herself, while feet tend to crack in summer months when skin is exposed (sandal wearing) or in winter when the weather is most dry. Hand



cracks usually occur around the nail bed, while foot cracks tend to be between the toes. Are these cracks painful? They certainly look that way. Some children are very bothered by cracks, while others do not seem to notice them at all.

The best way to treat cracks is to prevent them. For hands, this means keeping the hands well-moisturized, applying lotion frequently and *always* after hand washing. For infants, use a thick lotion such as Aquaphor, which will not come off as easily when fingers are sucked on. Don't worry about the child ingesting it -- a little lotion can actually help with constipation!

Every parent has their own recipe for foot care. Some parents file thick skin build up off with emery boards, others use special foot files, and one parent even swears by a Dremel sander! Removing

(exfoliating) the dead skin off is important to prevent cracks -- thick skin will eventually collapse on itself (the same thing that happens to a diabetic's foot) and will ulcerate if not attended to. In an infant rubbing the foot during bath time will be sufficient to remove build up. Once a child is walking, the skin will thicken, and stronger methods of exfoliation may be needed. Acidic lotions such as AmLactin can be used on very thick skin to remove foot scale, if this doesn't seem to sting or bother your child.

To use a decaling product on the feet, exfoliate the foot during bath, dry between the toes, rub on a decsaling product, seal with a thick layer of lotion, then cover in socks or footy pajamas. This routine is usually enough to prevent cracked feet. Many adults prefer to use plastic wrap over the lotion, covering the wrap with a sock to lock in the lotion. If your child will tolerate the plastic wrap, this can be a good way to lock in moisture, but is not advisable for small children. They can remove their socks in the night and the plastic wrap could pose a choking or suffocation hazard. If you choose to try the plastic wrap trick, do so while your child is awake and being still, such as during a favorite t.v. show or movie.

Once or twice a week check your child's toes for cracks. If a crack has developed, add a touch of antibiotic cream and then lotion and cover as usual. If the crack does not heal, smells bad, weeps and crusts, or seems to be really bothering your child, take your child to the doctor. He may have a fungal or bacterial infection unrelated to the Ichthyosis.

## **B. EARS**

Ear care, believe it or not, is the most controversial issue in the Ichthyosis Community! The outer canal of the ear is made of skin, just like the rest of the body, so it flakes and scales just like the rest of

your child's skin. The normal production of ear wax only makes a bad situation worse, and for some children the build-up can rapidly reduce their ability to hear. In an infant this is especially troublesome, because it can lead to permanent hearing loss. Build-up in the ear canal can also be a breeding ground for bacteria and can make ear exams almost impossible.

Regular ear cleanings are crucial for most types of Ichthyosis. On that, we all agree. It's *how* to clean the ears that heats up the debate!

For kids with Ichthyosis, ear specialists advise regular, routine ear cleanings by a trained professional. For many parents, though, this is not an option, either because of the cost involved or difficulty getting to a specialist. Many parents rely on at-home cleaning with regular check-ups by a primary care doctor.

If you are going to try at-home cleaning, be very, very careful. It is not advisable to **ever** put anything into your child's ear -- most people are not able to gauge the depth of the ear canal, and you can easily puncture your child's eardrum. There are some parents who use tweezers to clean scales from their children's ears, going against the general advice of ear specialists. A better method for most families is to use the following routine:

- Apply a few drops of olive oil into the ear canal at bedtime. The oil loosens and dissolves the wax, and helps moisturize the canal.
- The next day, mix a capful of hydrogen peroxide with a capful of warm water (50/50 solution).
- Using a dropper, fill your child's ear canal. The mixture will bubble up the loosened wax and scales, and you can wipe the build up off with a towel.

- Repeat until the ear is no longer bubbling up wax, and then do the other ear.

Each child is different, but you can usually get by doing this once or twice a week.

There are commercial products available to remove ear wax, such as Debrox, but the olive oil and peroxide trick is inexpensive, readily available, and effective if done regularly. **BE SURE TO USE HYDROGEN PEROXIDE AND NOT ALCOHOL.** Never use alcohol on a child with Ichthyosis. Alcohol stings and will cause your child to scream in pain.

There is a product available in the U.S. stores called an “Ototek loop ear wax remover” that has a built-in stopper to prevent going too far into the canal. Many children love this tool, which helps remove build-up and scratches the itch that their little fingers can’t quite reach. The ear specialist at the Chicago F.I.R.S.T. conference reviewed the product and admitted that it seemed safe to use, so if you are able to purchase this item it is a must-have for home ear care. As always, though, be very careful whenever you insert anything into your child’s ear, erring on the side of caution. Better not to get all of the wax than to puncture your child’s ear drum.

## **C . EYES**

Since the skin around the eyes can be pulled tight by dry skin, it can be difficult for your child to close her eyes or blink. This leads to dry eyes. Rapid skin production can also drop skin flakes into the eye, over time scarring the eye and causing blindness. Excessive skin production can also lead to bacterial infections, which can cause blindness.

To prevent eye damage, children with Ichthyosis should have yearly eye exams with a medically trained eye specialist (an Ophthalmologist, not an Optometrist). Follow the advice of your Ophthalmologist closely, and seek immediate help if you notice your child rubbing his eyes often, blinking rapidly as if he is uncomfortable, seems unable to close his eyes when he is asleep, has red eyes, or has developed discharge from his eyes. Good eye care is the best way to protect your child's sight.

There are several products available by prescription that "rewet" the eye and help with dry eye. See the product list below for suggestions. Follow the directions of your eye doctor carefully. Overuse of lubricating ointments can cause loss of vision in a child's developing eye.

#### **D. SCALP**

Because we are such a hair-obsessed society, the scalp is one of the hardest issues to master. The skin under your child's hair is no different than the skin anywhere else on her body, so it needs to be exfoliated and moisturized. If scales/flakes are not removed, permanent scarring can develop and your child may not be able to grow hair. Children with certain types of Ichthyosis have naturally brittle hair, which make scalp care even more difficult. The scalp, in other words, is a real pain.

The best way to avoid scalp problems is to develop a care routine and prevent problems **before** they start. Do the same things for your child's scalp that you do for her body when you bathe her. When your child is in the tub, always wet her head to loosen the scales, rub the scalp gently to remove flakes, wash her hair with a moisturizing, descaling, or dandruff shampoo (see product list below) and finish by rubbing a small amount of scalp moisturizer onto her head. Some

parents will also use a fine tooth comb, such as a lice comb, to help comb out flakes when they are soft after bathing. For some children, though, this irritates their scalp and hurts. Err on the side of being gentle with your child.

**TIP:** If your child gets lotion in his hair, you can use cornstarch to remove the oils. Use a pointed tip applicator, such as a plastic condiment bottle, to apply small amounts of cornstarch onto the roots of the hair and onto any sections that look oily. Gently rub the cornstarch into the scalp and sections, the brush through with a natural bristle brush until you no longer see the cornstarch. For dark hair, you can mix cinnamon, cocoa powder, or nutmeg into the cornstarch to darken it, if you don't mind smelling like dessert!

**TIP:** Avoid using hair dryers and styling products that contain alcohol. These dry out the scalp and make flaking problems worse. If you want to style long hair, try a wet/dry flat iron. This helps dry hair and style it at the same time in a safe way without affecting the scalp.

## **E. REMOVING LOTION FROM ... EVERYTHING**

### **IN GENERAL:**

Little children + lotion=big mess

You've worked so hard to get nice furniture, nice rugs, a house you can be proud of ... and then you had a baby. And not just any baby,

who would be messy enough, you have a baby who is coated in lotion head to toe, 24 hours a day.

Say goodbye to that nice couch and those nice rugs, and hello to Mr. Slipcover.

When you start baby proofing your home, you will also need to do “lotion” proofing. Look around your house and determine where your child will have access, and what type of fabrics are in those rooms. Do you have silk curtains or pillows? Put them away immediately. Persian rugs? If it pains you to see them stained, replace them with knock-offs that can be destroyed without worry. Do you have fabric or leather seats? Leather may be the better way to go if you don’t like the slip covered look. Lotion on leather will give it an “aged” look, but as a bonus it will also help keep it moisturized! If you prefer fabric furniture, choose a fabric with texture or a busy pattern, which will help hide lotion stains, or invest in removable slipcovers which can be washed.

Carpet on floors will suffer from lotioned feet, so be prepared to either clean them often or replace them with hardwood. Keeping socks on your child can help reduce the amount of lotion transferred to carpeting, but with some children getting them to keep socks on their feet is a struggle.

When doing your home inventory, you will also need to inventory your clothing and separate out the easily stained fabrics (synthetics and silks) and invest in a series of inexpensive, easily washable fabrics (mainly cotton) for every day wear. When your child is small, you will not only be covered by boogers and leftover spaghetti, you will also be a repository for lotion. If you don’t want to cry over your clothes, get some cheap cotton t-shirts that are easily laundered.

In order not to make your child self-conscious of the impact his lotion has on your clothing, it’s best to wear unimportant clothing around

him while he is small, wiggly, and likely to stain you. If you must wear “dressy” clothes, put them on after you have lotioned your child or cover them with a bath robe or “messy shirt.” Stress to your child that there are different types of clothes (pajamas, bathing suits, dressy clothes, play clothes) and that you want to keep your dress clothes nice. Tell your child he is always welcome to snuggle, just give you a moment to change or toss on a robe. **Stress as often as you can that your child is more important to you than things**, and that you do not care if lotion gets on things as long as it isn’t intentional.

Because at some point, it will be:



Avoid synthetic nightgowns on your child -- the lotion will make them stiff and uncomfortable. Polar fleece pajamas, however, hold in

lotion very well and stay soft. They hold up well and are a good investment for winter, as are flannel pajamas.

### **LAUNDRY:**

If you are washing clothes in a washing machine, then you will need to clean it regularly for both grease and bacterial build-up. Fill your washer with hot water and add a cup of bleach and a degreasing cleanser (such as Simple Green). After the mixture has soaked a bit, while it is still hot, drain the liquid and scrub the machine with a soft towel or scrub brush. Running the mixture through the machine's pipes will help clean the lotion out of the machine's system. Eventually your machine will die from lotion build-up, but this will help prolong your machine's life.

**TIP:** To get lotion out of clothing always wash with hot water. You can add a squirt of degreasing dishwashing liquid to the wash cycle if your machine will tolerate it. Use an unscented laundry detergent, and if your child has very sensitive skin, double rinse the clothes. One parent recommends using double the amount of recommended liquid washing detergent to get out lotion. If you do this, be sure to double rinse the clothes. Some adults with Ichthyosis have confided they prefer a little lotion in their clothes because it keeps them soft. How clean you get your clothes is a personal preference.

### **FLOORS:**

To remove lotion from linoleum or tile, use a general purpose cleanser such as 409 or a degreaser such as CitraSolv (an organic degreaser). Because many forms of Ichthyosis are also susceptible

to infection, it is a good idea to regularly use bleach or a disinfectant on your floors, tubs, and other areas where your child plays. For chemical free disinfecting, you can invest in an all-purpose steam cleaner, which uses a stream of hot water vapor to disinfect.

Removing lotion from carpet is a huge challenge. For oriental (wool) carpets, try a one part vinegar to 2 parts cold water solution. For synthetic carpet (wall-to-wall) you can try a solution of 1 tablespoon household ammonia, 1 quart cold water, and ¼ teaspoon liquid soap (14.79 ml ammonia, 1.6 pint cold water and 1.232 ml liquid soap).

- Begin by blotting with a clean cloth to remove as much of the grease as possible. It's very important to BLOT throughout this entire process. DO NOT SCRUB. Your goal is to "lift" the stain, not rub it into the fibers. Working from the outside of the stain towards the center will keep the stain from spreading.
- Spray stain with appropriate cleaning solution for your carpet type.
- Blot with a clean dry cloth until no more stain shows on the cloth. You will need to change your cloth often or use a clean patch of the towel each time as you are blotting to be sure you are not spreading the stain.
- Repeat steps 2 and 3 until the stain is no longer noticeable.
- Spray with clean water to rinse.
- Blot with clean cloth. Repeat as necessary to be sure all cleaning solution has been removed. Residue left from cleaning solutions can collect dirt more quickly and will create another situation to be dealt with.

- Cover stain with clean cloth or stack of paper towels and put heavy weight on top (heavy books, etc.)
  - Leave to set for several hours or overnight to absorb any remaining liquid.
  - Let area dry thoroughly.
  - Vacuum to return fibers to normal texture.
- ❖ Always use cold water to keep from setting the stain.
  - ❖ Do not use laundry detergent or dishwasher detergents as they may damage the fibers of your carpet.
  - ❖ Do not use bleach or ammonia on wool carpets as it will damage the fibers.
  - ❖ Do not use carpet spot cleaners on oriental rugs as it will damage the fibers.
  - ❖ Oriental rugs should be cleaned with caution. If your rug is an antique, silk, or part silk, contact a professional to remove the stain.
  - ❖ Spot Shot is a cleaner that is available at most U.S. stores and is well-known for removing tough stains.

#### FURNITURE:

For leather furniture, simply wipe the lotion off with a dry cloth. If you have fabric furniture and do not have washable slipcovers, then you need to use upholstery cleaner to remove stains from fabric. Follow the directions on the container carefully, especially taking the time to do a test spot in order to avoid ruining your entire piece of

furniture. If the stain does not come out or is deeply set, you may need to contact a professional upholstery cleaner in your area.

## **WALLS:**

To remove lotion stains from walls, you will need a grease fighting cleaner (examples in the U.S. are Dawn Power Dissolver (mix with water), Greased Lightening, Mean Green, or the universally available vinegar and water. You will also need soft cloths or a sponge, a bucket or bowl, and water.

### **Steps to Remove the Stains:**

- Begin by wiping the area with a clean cloth or paper towel to remove as much of the grease as possible.
- Next, apply the chosen grease-fighting cleaner to the area. If the cleaner is mixed with water, it is often more effective if the water is hot as it helps to break through the grease for easier removal.
- Use a sponge or soft cloth to gently scrub the area. If the paint is textured, avoid using any harsh scrubbers or too much pressure as it may remove the texture plaster. A soft brush may be helpful on these walls to get in between the grooves without damaging the surface.
- Once the stain is removed, rinse the area with a clean cloth moistened with water.
- Dry with a soft cloth and allow to air dry completely.

If the stain cannot be removed, it may be easiest to paint over it with a primer and then the original color. The primer will prevent the grease from coming back through the paint surface.

## F. ILLNESS/FEVER

If your child is sick, the first thing you need to know is that he/she will probably do a “full-body” peel during and after the illness. This means that **ALL** of the skin on your child’s body will begin to flake and peel off, scalp to toe. The skin underneath can either be very red and sore, or look soft and perfect, depending on how sick your child has been. In addition to the full-body peel during illness, many people with Ichthyosis do a full body (scalp to toe) peel during major shifts in the weather. Spring and Fall seem to be the worst times for peels. Treat the skin as you normally do, with maybe a bit of extra exfoliation and moisturizing to help with the peeling. Be aware that the skin can be extra sensitive during a full peel, so be very gentle and dress your child in very soft clothing that doesn't rub.



If your child has a fever and is sweat-impaired, give a fever reducer such as acetaminophen and help cool your child down by placing her in a luke-warm bathtub. Allow your child to play for as long as he or she is comfortable in the tub, using cool (not iced) rags on her neck to help bring her temperature down. **DO NOT PACK YOUR CHILD IN ICE**, as this can cause chills and actually increase the fever. During a fever or illness, increase liquid intake and apply extra lotion as seems necessary. **AVOID SALICYLIC ACID PRODUCTS DURING A**

**FEVER OR MAJOR ILLNESS**, because of the risk of Reye's Syndrome. Salicylic acid is another name for aspirin, and your child will absorb it into his skin. Reye's Syndrome is a potentially fatal condition caused by aspirin products given to a child when he or she has a fever.

Your child will likely tolerate being ill without major complications. During an illness, be very vigilant about infection control (use a capful of bleach in the bath water, always wash your hands before applying lotion, and use antibacterial ointments on any cracks or fissures that appear during a peel). Keep your child well-hydrated and comfortable. If your child is sweat-impaired and his fever does not reduce, gets as high as 105°F (40.5°C), or your child seems in distress in any way, get him to the emergency room immediately. At the doctor, explain that your child has Ichthyosis and cannot sweat, and that you need help immediately to prevent a febrile seizure. If your child can sweat but has a high fever that does not go down with fever reducers, then seek medical care for possible infection.

#### **G. TEMPERATURE ISSUES/SWEAT IMPAIRMENT**

Some kids with Ichthyosis cannot sweat. Why exactly is a mystery, and some children seem to sweat a little while others cannot sweat at all. Some children can sweat but cannot regulate their temperature, freezing on warm days. It can be very frustrating and scary not knowing how your child will react to temperature changes.

Most children with sweat impairment are comfortable up to around 78°F (25.5°C), as long as they are not active. Once the temperature gets warmer than that, their bodies begin to have trouble, and they will often begin to overheat. For outdoor activity, temperatures that high can often be too much. Each child is different, so you will just have to monitor your child to learn what his body can tolerate.

Signs of overheating are:

1. **Bright red face**
2. **Muscle cramps**
3. **Tiredness and weakness**
4. **Dizziness or fainting**
5. **Headache**
6. **Nausea or vomiting**
7. **Confusion**
8. **Loss of consciousness**
9. **In small children, irritability can also be a sign of overheating**

If you see your child turning red, **immediately** get her to a cool, shady place and give a cold drink. Apply cool, wet washcloths to her neck and loosen or even remove clothing. **If your child vomits or loses consciousness, this is a medical emergency and you should seek immediate medical care.**

There are many products available to help with sweat impairment. For older children there are medical assistance devices available, called “cooling vests,” which use removable ice packs to help lower your child’s body temperature. These devices can be rather heavy, though, and are not practical for small children. For toddlers, use a spray bottle with ice water to mimic sweating. Wet or even frozen t-shirts can help small children stay cool. What is most important is finding a system that works with your child, remembering to carry the products with you on outings, and watching your child closely for signs of overheating. As your child gets older, this will get easier, especially around the age of five when he can tell you he is too hot. Until then, you will have to be vigilant to guard against heat stroke.

## H. NUTRITION

The following is excerpted from a lecture presented by Dr. Mary Williams, a pediatrician who is an expert in the nutritional needs of children with Ichthyosis:

Good nutrition is important for everybody, but it is especially important during growth. Some infants and children with more severe forms of Ichthyosis have special nutritional needs. With a few rare exceptions, poor growth in children with Ichthyosis is not part of the syndrome (it is not genetic). Instead, it is a consequence of “skin failure.” Severe forms of Ichthyosis include:

- Harlequin Ichthyosis
- Netherton syndrome
- LI/CIE (some)
- EHK (some)
- I. en confettis

Growth failure in Ichthyosis usually begins soon after birth, when the infant feeds well but does not gain weight. The infant falls to a low percentile, sometimes “off the chart” (less than 3<sup>rd</sup> or 5<sup>th</sup> centile). Even when consuming enough calories to support growth in a normal child of the same age, the infant does not catch up. Usually the GI tract is functioning normally, and there are no nutritional deficiencies.

Growth failure can be attributed to the failure of the skin barrier. The function of the skin is to provide protective barriers from the external environment -- keep the outside out and the inside in. When the barrier is impaired, as in Ichthyosis, the child’s skin loses too much water. The average child loses a cup of water a day through water evaporation. **A child with Ichthyosis loses 3 to 5 cups of water a day.** As water evaporates it takes with it heat, which means it is taking with it calories. A normal child burns 40-130

calories a day from water loss. A child with severe Ichthyosis loses 80-1000 calories a ***day just in water loss***. This is in addition to the calories it takes just to run your child's body.

What does this mean as a caregiver? It means that children with Ichthyosis need more fluids (water) and food (calories). Talk with your pediatrician about any growth concerns you have. Is your child "off the chart" or "off the curve"? Is he much smaller than his siblings were at the same age? If so, your child may benefit from a consult with a nutritionist. You may need to increase the caloric intake of your child through higher calorie foods, nighttime NG tube feedings, or even surgical insertion of a "G-tube." Usually, though, you can add a high calorie drink such as Pediasure/Kindercal once a day, and push fluids to your child throughout the day.

A bottle or "sippy cup" of water should always be near your child, even in bed with her at night. Older children can be given sports drinks during the day as well, since electrolyte loss is also a concern. Keep water available for your older child at night as well.

**TIP:** Since Pediasure and Kindercal are really just sweetened milk with added vitamins, you can make a cheaper version at home. Mix whole milk (for the extra calorie boost) with flavored drink syrup (chocolate, vanilla, or strawberry are usually widely available). Add vitamin drops, or wait and after your child has finished the drink give her chewable vitamins.

**TIP:** Make your own powdered electrolyte solution:

- 2 quarts water
- 1 teaspoon baking soda
- 7 teaspoons sugar
- 1 (1/4 ounce) packet unsweetened Kool-Aid powdered drink mix (optional)
- 1/2 teaspoon salt (optional)

Mix all together well and store in the fridge. Make ice cubes or popsicles with this.

(metric)

- 1.89 liter water
- 4.93 ml baking soda
- 34.50 ml sugar
- 1 (1/4 ounce) packet unsweetened Kool-Aid powdered drink mix (optional)
- 4.93 ml salt

## **I. DEALING WITH BLISTERS (EHK)**

Children with EHK are usually born with blisters or began blistering shortly after birth. If your baby was born in the hospital the staff whisked your child away to the NICU to be treated and diagnosed. Best case scenario, your child was placed in a bassinet and her blisters treated like E.B. (Epidermolysis Bullosa) blisters. The nurses probably covered your child with petroleum and then gauze bandages. In the hospital your child's dressings probably were changed every 8 hours. The nurses should have taught you how to wash your child with sterile water, cover the open sores with petroleum and then cover with various types of gauze. The best gauze to use is petrolatum gauze. This helps keep the petroleum moist and allows the sores to heal. After the petrolatum gauze

generally cotton gauze is wrapped around your child's limbs. **DO NOT USE ADHESIVES** on your child's skin. This will only cause the skin to tear even more. To secure the bandages, wrap the gauze tape so that it sticks to itself or put tube gauze around the cotton gauze.

In EHK, the diaper area is always the worst. Avoid using tight diapers and cut out elastics if you use disposable diapers. This will cause messes but your child will be more comfortable, and you can avoid hurting your child's skin even more. Soft cotton cloth (prefold) diapers are best, if messy.

In the first few weeks of life your life will revolve around the dressing changes. The point of the petroleum and gauze is to keep the blisters from getting infected. **Once you notice the gauzes creating more blisters than they are helping STOP using them.**

Petroleum works well as a moisture barrier to keep in the moisture from your child's bath and will be sufficient for daily use. If you notice your child's skin tearing easily (and this is common, especially for the first 3-4 months after birth) you may want to put your child's clothing on wrong side out so that the seams do not touch and irritate the skin. Onesies work well for this. If you are using cloth diapers cut off the bottom half of the onesie and use as a shirt.

Your child will blister extremely easily for the first 3-4 months of life, but the blistering will continue for the rest of her life. Most of the "blisters" at birth are really abrasions. When your child begins to get real blisters they will look like the blisters you get on your feet when your shoes are too tight. They can be teeny tiny or huge. The longer you leave the blister alone the larger it will become, so the

best thing to do for the blister is to pop it.<sup>2</sup> An un-popped blister can be very uncomfortable for your child, as the skin is pulled around it creating pressure. To pop the blister take a needle that has been sterilized by passing it through a flame and wiped with an alcohol swab, then pierce the blister at the base and dab with a cloth to absorb the liquid. Cover the newly popped blister with petroleum to avoid infection and gauze if you so choose. Leaving the blister uncovered allows it to dry out and the skin will flake off.

You will eventually begin to notice the triggers of the blisters. Some common irritants to avoid are:

1. Excessive heat (which may or may not be too hot for you, but too hot for your baby)
2. Skin that is too moist (from an extremely long bath or with too much lotion/cream)
3. Synthetic materials (100% cotton clothing is best for EHK babies) and
4. Friction (from clothing, diapers, car seats, etc.)

Your child will probably not start building up scales until after the blistering slows. When the skin begins to build up it will look like tiny warts on top of each other, usually in skin creases like the neck, elbows, knees, ankles and wrists. Treat the skin buildup with exfoliation and moisturizers, but keep in mind your child's skin may

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<sup>2</sup> Popping blisters is discouraged by physicians as it can introduce infection to the skin. Leaving a blister alone on an EHK child can cause discomfort and allow infection when it inevitably ruptures. This is a judgment call on the part of the caregiver -- discuss the risks and benefits of popping blisters with your physician.

be sensitive to exfoliation and blister. Be very gentle with your child's skin.

Your EHK baby's skin will evolve over time. As a caregiver you need to pay attention to these changes and notice how sensitive your child's skin is and treat it accordingly.

If your child has palmer plantar EHK that means her hands and feet are affected with blisters and build-up. Treat the blisters on her feet the same way you would any other. There is no need to open up your baby's fist to check for blisters. When the skin on her feet begins to tear DO NOT pull on skin tags. If the skin tag is long you can cut it off with round tip scissors or fingernail clippers. It is also a good idea to do some manual manipulation of your baby's feet and hands if you notice the skin tightening to the point that your child has difficulty moving her fingers and toes. Most EHK babies have trouble or delay in learning to crawl and walk. You may need to work with your pediatrician and a physical therapist to aid your child in learning to walk.

Treat dry skin, scalp, and itching as detailed in Section I.

**TIP:** Allow your baby to play without a diaper to help clear up diaper rash or diaper area blisters. To avoid a mess, layer the crib or playpen in the following manner:

1. a layer of newspaper
2. a layer of plastic garbage bags, rubber backed crib protectors, or flannel blankets, and
3. top it off with a crib sheet.

Repeat this process for several layers, then place your child in the playpen without a diaper. As your child urinates or messes the sheet, simply remove that down to the next crib sheet. This is also helpful for when your child has diarrhea or is vomiting in his crib.

## PART III

### GLOSSARY OF PRODUCTS

**NOTE:** In the members only section of the F.I.R.S.T. website there is an extensive, detailed spreadsheet covering almost all of the products used by people with Ichthyosis. The following list is a shorter version of the staples recommended by caregivers of kids with Ichthyosis. We have also attempted to include products available in the U.K., Canada, and Australia.

#### A. LOTIONS AND CREAMS

<b>Product</b>	<b>Availability</b>	<b>Description</b>
Aquaphor	US	Gold standard for Ichthyosis treatment in the US. Thick, sticky petroleum based product, it lasts the longest of all creams. Ichthyosis patients in the US are eligible for a free case of Aquaphor every 3 months. Have your doctor fax a letter to 203-563-5940. The letter should include your child's diagnosis and treatment, and mention that Aquaphor is necessary for your child's care. After you receive your first shipment, keep the packing letter and call customer service every 3 months to renew. That number is: 800-227-4703.

Aveeno	US, Canada, Korea and AU	Gentle lotion containing oatmeal extracts. Good for soothing itchy skin.
CeraVe	US	Barrier repair cream available in US drugstores. No prescription needed.
Cetaphil	US, CA and AU	Gentle unscented cream, good for layering under thicker lotions or for touch-ups and face.
Cetraben	UK	Cream that soaks in easier than ointment and doesn't leave greasy residue while maintaining moisture levels.
Coconut Oil	Universal	Natural product derived from coconuts. Can be melted and used on the scalp or as a general body moisturizer, or can be added to other lotions to boost staying power.
Eucerin	US and UK	Product similar to Aquaphor, made by the same company.
Eucerin Dry Skin Relief Soothing Spray	US and UK	Light spray on moisturizer that contains polidocanol which helps to relieve itching caused by dry skin.
Eucerine Wools Alcohol	AU	Australian equivalent of Aquaphor, very thick petroleum based cream. Contains lanolin, which for some people can be an allergen.
Epaderm	UK	a mixture of yellow soft

		paraffin, liquid paraffin and emulsifying wax that produces a greasy moisturizer. It works by providing a layer of oil on the surface of the skin to prevent water evaporating from the skin surface.
Glycerin	Universal	Vegetable based moisturizer used to boost the staying power of other lotions. Also nice as a moisturizer on its own.
Kenkay	AU	Creams and emulsifying ointments used for compounding; Soothes and protects dry, cracked skin. Preservative free, contains no animal ingredients (BSE free), no added colours or fragrances
Mineral Oil	Universal	Used to supplement other lotions (boosts staying power). On it's own it can be used as a moisturizer.
Nutraplus	AU	Similar to Cetaphil, made by Galderma (same manufacturer)
Olive Oil	Universal	Emollient used to moisturize skin. Can be used as a moisturizer when other products are unavailable. Also good for ear and scalp care.
Petroleum jelly	Universal	Refined petroleum jelly is often used to mimic the missing proteins

		that cause the skin barrier in Ichthyotic skin to flake.
Shea Butter	Universal	Natural cream derived from the shea nut; used as a moisturizer and to boost staying power of other creams.
Vanicream	US	Thick white cream used as a light-weight moisturizer, often on the face and hands.

## B. DESCALERS

AmLactin	US	12% lactic acid neutralized with ammonium hydroxide to provide a lotion pH of 4.5-5.5. Lactic acid, an alpha-hydroxy acid, is a naturally occurring humectant for the skin. AmLactin moisturizes and softens rough, dry skin.
Calmurid	AU	10% urea, 5% lactic acid
Dermadrate	AU	thin cream with 10% urea and lactic acid
Lachydrin	US	Alpha Hydroxy Acid Lotion.

## C. EAR PRODUCTS

Debrox	US	Debrox Solution is a form of peroxide. It works by softening and removing earwax
Similasan	US, CA, Europe, UK,	Homeopathic remedy

	South Africa	for ear wax removal
Sweet Oil/Olive Oil	Universal	Natural product that actually melts the ear wax

#### D. EYE PRODUCTS

Cellufresh	AU	Daily drops to keep eyes moist
Celluvisic	AU	Thinner drops than Cellufresh, also used to keep eyes moist
Hypo Tears	US	used to relieve burning, irritation, and discomfort of the eyes due to dryness.
Lacrilube	US and AU	Thick moisturizing ointment for nighttime use.

#### E. CLEANSERS

Aquaphor Baby Wash	US	Non-soap cleanser formulated for infants and sensitive skin
Aveeno Bath Wash	US	Cleanser made with Colloidal oatmeal and oils
Cetaphil Cleanser	US	Non-soap cleanser, very gentle
Soap	Universal	Sometimes gentle enough to use without excessive dryness. For some people, too drying. Useful for removing bacteria and preventing infection.

## F. SHAMPOO

Carmol Antibacterial Shampoo	US	10% urea; helps remove scalp scales while also killing bacterial growth
Feeling Flakey by Free and Clear	US	Non-medicated hair care products that are gentle to both hair and scalp. Specially formulated to avoid common chemical irritants found in most shampoos and conditioners.
Head and Shoulders	US	Anti-dandruff shampoo containing pyrithione zinc
Paul Mitchell Awapuhi Moisture Mist	US	Leave in moisturizing conditioner. Can help with scalp moisture.
Paul Mitchell Tea Tree Shampoo	US	Tea Tree helps prevent infections, but can sting. Use sparingly on small children, never use on a freshly exfoliated scalp.
QV bath oil	AU	good moisturizer, add 1 capful to baby bath. Great for itchy skin
Salex	US	Prescription-strength formulation of 6% salicylic acid removes scales and reduces plaque. Can sting, use sparingly on small children.
T-Gel	US	Coal tar shampoo. Helps with flaking, but because of the coal tar use sparingly on small children
T-Sal	US	Contains salicylic acid, can sting.

## G. ANTIBACTERIALS AND ANTIFUNGALS

Bactroban	US	Bactroban is an antibiotic that treats or prevents infection that is caused by bacteria. Often used for MRSA.
Phisohex	AU	with 1% triclosan- great for MRSA infections but consult dermatologist first.
Ketoconazole	US	Cream for yeast infections
Nystatin	US	Cream for yeast infections

## H. SUNSCREEN

Sunsense toddler milk	AU	
Vanicream SPF 50	US	
Waterbabies SPRAY sunscreen	US	doesn't sting and dry out skin like other sunscreens.
Waterbabies STICK sunscreen	US	feels similar to Aquaphor, rather thick and sticky. Many people with Ichthyosis prefer the feel of this stick to any other type of sunscreen.

\*Sun protective clothing is also a good choice for children with the condition. They are easily purchased in stores and online, and have the added bonus of not drying out the child's skin.

## **I. COOLING PRODUCTS**

StaCool Vests: Cooling vest product preferred by most parents of children with Ichthyosis. Expensive, but can qualify for state assistance as a medical device. Contact your local disability office to see if your state/country offers coverage.

<http://www.stacoolvest.com> 866-STA-COOL (866-782-2665).

Outside the U.S.Call 352-597-1820.)

## **PART VI**

### **RESOURCES:**

There are many organizations that are devoted to helping families in your situation. You may be eligible for state assistance if you live in the U.S., or an allowance if you are in the U.K., Canada, or Australia.

The best thing you can do is find a mentor (and friend) through either the F.I.R.S.T. network or through the Ichthyosis Board. Having another caregiver who understands your child's unique issues will be an amazing foundation and help you get through the toughest times.

You will want to bookmark the contact information for the Ichthyosis Foundation, or F.I.R.S.T. Even though they are a U.S. based non-profit group, they work with Ichthyosis on an international scale. Their website is: [www.scalyskin.org](http://www.scalyskin.org)

F.I.R.S.T. is located at:

1364 Welsh Road G2  
North Wales, PA 19454  
Phone 215.619.0670  
Fax 215.619.0780

F.I.R.S.T. has a comprehensive care guide available for new parents, as well guides for specific issues related to the various types of Ichthyosis.

Madison's Foundation (contact information below) is a support group for parents of children with rare diseases. You can also find support through the F.I.R.S.T. network or through the Ichthyosis board.

Madison's Foundation  
PO Box 241956  
Los Angeles CA 90024  
Phone #: 310-264-0826

e-mail: [getinfo@madisonsfoundation.org](mailto:getinfo@madisonsfoundation.org)

Home page: <http://www.madisonsfoundation.org>

In the U.K., there is an Ichthyosis Support Group. Their website is:

<http://www.ichthyosis.org.uk/>

Ichthyosis Support Group,  
PO Box 7913,  
Reading.RG6 4ZQ.  
UK

Telephone No: 0845 602 9202

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

\* Household Cleaning tips taken from “How To Clean Stuff.net”  
found at <http://www.howtocleanstuff.net>