

Ichthyosis

Release the Butterfly

A Handbook for Parents & Caregivers
of Children with Ichthyosis



FIRST

Foundation for Ichthyosis & Related Skin Types™

Educate • Inspire • Connect

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RELEASE THE BUTTERFLY

*A Handbook for Parents & Caregivers
of Children with Ichthyosis*

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*This booklet is dedicated to all the children
and families affected by ichthyosis.*

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INTRODUCTION

It may be perhaps the most anguishing experience you will ever know. Your baby has a skin disorder called ichthyosis. The excitement you anticipated at the birth of your child has been tempered by fear and pain; the joy has been diluted by tears. It's okay to cry, but remember this ... life gets better.

Your child will laugh, dance, run, and play, and will have friends. He or she, should they desire, will play sports, will attend the homecoming dance, will find a mate, and with diligence will achieve his or her life goals. Your baby will have a normal childhood and will lead a normal life.

However, much depends on you. Ichthyosis will influence your child's development only as much as you let it. The Foundation for Ichthyosis and Related Skin Types, Inc.[™] (FIRST) is here to answer your questions and help you through the challenges ahead, as you guide your child to their special place in the world.

This book is for parents, by parents, and is dedicated to new parents of children with ichthyosis. We hope that it meets your needs during a strange and confusing time: caring for a child affected by a rare, genetic skin disease.

The original printing of this book, in 1994, was the result of many hours of hard work by a devoted group of parents of children with ichthyosis, and medical professionals. The parents set out to write something that would have been helpful when they were a new parent of a child with ichthyosis: a useful source of

information about the practical, day-to-day matters of caring for a child with ichthyosis.

This booklet has been edited several times since its original printing in 1994 to include the most up-to-date medical information and practical advice.



WHAT IS ICHTHYOSIS?

We had been told by a neonatologist that our baby would die of an infection and dehydration. Later, the dermatologist gave us an image to cling to in the first, horrible days. “Think of a caterpillar that breaks out of its cocoon and becomes a beautiful butterfly,” he said.

“Ichthyosis.” After hours, days, months, or even years of examinations, specialists, and testing, you finally have a diagnosis. It can be a relief to have a name, but you may feel strong emotions: *Did I cause it? I shouldn’t have ... while I was pregnant. I should have known.* These emotions are often compounded by confusion. Since all the ichthyoses are so rare, sometimes even the professionals will give you outdated or inaccurate information.

Your best defense in this situation is up-to-date and accurate information from a physician who has experience with ichthyosis. Getting a correct diagnosis of the type of ichthyosis that affects your child will help you to best understand his or her condition and predict his or her current and future needs and the impact on your family.

The term ichthyosis derives from *ichthys-*, the Greek root for “fish.” The fanciful reference is to the skin’s scales, which some consider fish-like. Rather than a single disease, ichthyosis identifies a family of about 30 related disorders. The ichthyoses (plural) are a group of rare genetic skin diseases whose primary characteristics include dry, thickened, and scaly skin. In some

forms, the skin blisters and peels, especially during infancy and childhood. Many people with ichthyosis have darkened, rough skin that can appear as scales separated by deep creases. Mild forms of the disorder, particularly mild forms of Ichthyosis Vulgaris, may go undiagnosed, misdiagnosed, or overlooked because a parent has it.

The characteristic thickened and scaling skin results from genetic defects in the body's ability to make and maintain the outermost layer of the skin, the *stratum corneum*. These genetic errors result in one of two biological outcomes, depending on the type of ichthyosis. Either the body manufactures the stratum corneum too rapidly (up to 300% faster than normal), or the stratum corneum fails to shed (desquamate) properly from the surface of the skin. Because skin constantly renews itself, either of these two interruptions in the balance between the making of the skin and its shedding results in the main signs of ichthyosis: thickened, scaly, red skin. In one form of ichthyosis, Netherton Syndrome, the stratum corneum is too thin, rather than too thick. Here the problem is one of too rapid shedding (desquamation).

Some infants with ichthyosis are born encased in a tight, shiny, cellophane-like membrane. This is called a "collodion" membrane, and it usually covers the entire body. Collodion is a descriptive term for the membrane, which is a sheet of stratum corneum cells that have failed to shed during development in the uterus. Therefore, infants must shed the membrane. They may do this within a few days, although the membrane may not completely disappear for several months.

The collodion membrane can vary tremendously in thickness, and the period during which it is shed may be a vulnerable time for the infant. As the membrane cracks, fissures may extend through layers of skin making the infant susceptible to infection. Tightness around the chest and trunk, as well as around the nostrils, may make breathing difficult. The tightness of the membrane may cause the fingers and toes to contract and may restrict blood flow. The tightness of the membrane around the

eyes may cause the eyelids to flip inside out (ectropion). This may also happen to the lips (eclabion). The effects of the tight membrane around the mouth may create difficulties with nursing. Infants with collodion membranes may have problems with fluid and electrolyte balance, and are particularly at risk for hyponatremia (elevated blood levels of sodium).

Babies with Epidermolytic Hyperkeratosis (EHK) have fragile skin, which may blister severely in infancy. They may lose large amounts of their outer layer of skin (the epidermis) during the birth process. Their skin may also blister or tear away with the slightest trauma. After a few weeks of living in a protective environment to avoid infection, these babies will appear less raw and their skin will be less fragile.

Babies with other forms of ichthyosis may be born with reddened skin, or thick, cheesy-like scales, or even looking normal or relatively normal, although their skin will gradually take on an abnormally rough, cracked and dry appearance. Most types of ichthyosis are diagnosed sometime within the first year of life, but in some cases, especially with Ichthyosis Vulgaris (the most common form of ichthyosis), the disorder may go undiagnosed indefinitely since it may be so mild that it is just treated as “dry skin,” or as a family trait.

Conclusive diagnosis of an ichthyosis is based on a combination of visible signs, family history, microscopic analysis of a biopsied section of skin, biochemical tests, and today, for many forms of ichthyosis, genetic analysis. The four most prevalent types of ichthyosis are Ichthyosis Vulgaris (IV), Lamellar Ichthyosis (LI)/Congenital Ichthyosiform Erythroderma (CIE), recessive X-Linked Ichthyosis (RXLI), and Epidermolytic Hyperkeratosis (EHK), which is sometimes called bullous Congenital Ichthyosiform Erythroderma (bullous CIE).

The ichthyoses are genetic conditions. They result from alterations or mutations in genes. This means that ichthyosis is not contagious. Frequently, parents who are not affected with ichthyosis bear a

child who is affected and this may cause confusion. FIRST publishes a booklet entitled *Ichthyosis: The Genetics of Its Inheritance*, which explains the patterns of inheritance for genetic disorders, and explains why parents do not necessarily need to have ichthyosis in order to have affected children. FIRST's booklet *Ichthyosis: An Overview* describes the most common forms of ichthyosis and answers frequently asked questions.

Knowledge is empowering! We urge you to study carefully FIRST's pamphlets that outline the hereditary patterns and the physical and psychological manifestations of ichthyosis. But this is only a starting place. FIRST can provide you with detailed information about your child's specific condition, treatment advice, and resources for dealing with issues common to ichthyosis. Ask for a connection to the Regional Support Network through which you can talk to or email other parents of children with ichthyosis. An experienced parent can talk you through your fears and concerns, and offer practical advice.

Searching the medical literature or the internet for information about ichthyosis can be useful. However, the medical literature is written for medical professionals and information offered in chat rooms may be inaccurate. When searching medical websites, keep in mind that the language may be very technical and the articles presume that you already know a great deal about skin biology and medicine. A good medical dictionary can help you learn the language so you can better understand ichthyosis and carry on meaningful discussions with medical professionals. Keeping a small notebook with all your questions and the answers from medical professionals can help you better understand what is happening to your child and enable you to ask intelligent questions when there is something you don't understand.

Your doctors may be nearly as unfamiliar with ichthyosis as you are. Your high level of interest, knowledge, and commitment can foster the same in him or her. You may need to rely on yourself at times so the more you know and understand; the better choices you can make for your child.

THE ICHTHYOSSES & RELATED SKIN TYPES

Acquired Ichthyosis

Autosomal Dominant Lamellar Ichthyosis

Chanarin-Dorfman Syndrome (neutral lipid storage disease)

CHILD Syndrome (unilateral hemidysplasia)

Conradi-Hünermann Happle Syndrome (X-linked
Chondrodysplasia Punctata syndrome)

Congenital Ichthyosiform Erythroderma (CIE, nonbullous CIE)

Darier Disease (keratosis follicularis)

Epidermal Nevus Syndrome

Epidermolytic Hyperkeratosis (EHK; bullous CIE)

Erythrokeratodermias (E. progressiva symmetrica,
E. variabilis & E. heimalis)

Giroux-Barbeau Syndrome

Harlequin Ichthyosis (harlequin fetus)

Hailey-Hailey Disease (familial pemphigus)

Ichthyosis en confetti (ichthyosis with white spots)

Ichthyosis Hystrix (Curth-Maklin type)

Ichthyosis Vulgaris

Keratosis Follicularis Spinulosa Decalvans

KID Syndrome (keratitis-ichthyosis-deafness)

Lamellar Ichthyosis (recessive)

Multiple Sulfatase Deficiency

Netherton Syndrome (ichthyosis linearis circumflexa)

Palmoplantar Keratoderma (various types)

Peeling Skin Syndrome

Pityriasis Rubra Pilaris

Recessive X-Linked Ichthyosis (steroid sulfatase deficiency)

Refsum Disease (phytanic acid storage type)

Rud Syndrome

Sjögren-Larsson Syndrome

Trichothiodystrophy (Tay Syndrome; IBIDS syndrome)



BEGINNINGS

One day, when we were having a particularly traumatic time in the pediatric intensive care unit, I found a phone and called my pediatrician. He was on the phone instantly and talked to me for fifteen minutes, interpreting for me the jargon being used by the specialists and giving me some guidelines about how to approach them with my concerns. With his help, I went back in feeling more confident, not like some intruder in a world I didn't understand.

DEALING WITH DOCTORS

If you have never spent a lot of time with doctors, now is the time to make up for it. Since ichthyosis is a rare and complex condition, many specialists will probably be brought in on your baby's behalf. Although the number of professionals may intimidate you, as you become familiar with each of them and their functions, you will begin seeing them as your baby's team. Remember, you and your physicians are on the same side; you both want what is best for your child.

Get a notebook that you can easily carry in the hospital and begin making a record of the professionals you meet. Note their departments, specialties, and how to reach each of them. Use your notebook to keep track of tests, medications, your baby's progress, and your doctors' observations. Write down unfamiliar words you encounter and don't be reluctant to ask a doctor or nurse to stop and spell a word and explain its meaning. Don't hesitate to ask questions or express concerns you might have to any of your child's caregivers. Early on you will discover that you

relate more to some doctors or nurses than others; these few can serve as helpful resources and friends in the neonatal intensive care unit's confusing environment of machines and medical terminology.

Find out right away who is in charge of your baby's care. This may be a neonatologist, a physician who specializes in the treatment of newborns. In the case of infants with ichthyosis, however, primary care may fall to a dermatologist or pediatric dermatologist. If your baby is at a teaching hospital, there may be several levels of authority and responsibility. This can be confusing, so make certain you get a clear picture of these lines of responsibility.

If your baby's doctor is on a tight schedule and doesn't offer you the time you feel you need, or if you want time to organize your thoughts and questions, set up an appointment to meet him or her in the privacy of an office. If, understandably, you are having trouble expressing yourself clearly or retaining information, consider bringing along a friend or family member with a clear head. If you can, also bring a tape recorder so you can replay the information later to refresh your memory.

It is easy to feel intimidated by doctors. Don't be! Most doctors appreciate parents who express a willingness to be informed about their child's condition. Remember that your doctor has other patients and parents, and respect his or her time by organizing and prioritizing your questions. But be assured that you have rights, too.

You and your partner are your child's most important advocates. You are entitled to any information you seek. If you don't understand something your doctor says, ask for a fuller explanation. If you don't understand why your child is receiving a particular treatment, ask. Don't be afraid that a question might make you look ignorant and don't be afraid to speak up! It is vital for you as a parent to feel in control of the well being of your child.

FEELING AT HOME IN A NEONATAL INTENSIVE CARE UNIT

Our daughter spent the first month of her life in a neonatal ward. My husband and I made her space as warm and personal as possible. We taped family pictures to her isolette and brought a small tape recorder and some soothing tapes from home. Our daughter particularly seemed to enjoy a tape of ocean sounds so her nurses, getting into the spirit of things, made her a colorful mobile with palm trees and sailboats. I think all of these special touches comforted my husband and me as much as it did our baby.

When you first walk into a neonatal intensive care ward with its high-tech equipment and imposing staff, it can be a frightening, intimidating experience. However, by the time your baby is discharged, it will probably feel like a second home.

Familiarize yourself with the ward, its rules, and staff. Get acquainted with the equipment and the function of each piece. Don't be afraid to ask questions. Though many neonatal wards are relatively crowded, don't feel as though you're in the way. This is your baby's temporary home, so pull up a chair and relax. Also, get to know your baby's nurses; this way, when you are at home, you can feel comfortable about who is caring for your baby. The nurses will be your first source of information about how your baby is doing, since they spend the most time with him or her. You can request that the same one or two nurses care for your baby every day. Having the same caregivers every day will allow the nurses to learn as much as possible about ichthyosis, its special circumstances, and how it affects your child. Remember, your baby is an infant with the need to sleep many hours. You have the right to limit the medical professionals who see your child to those who are immediately necessary. You have the right to ask a doctor or technician to come back at a time when the baby is not sleeping.

One family worked with the nursing staff to set up two cribs for the baby. One crib was decorated with comfort items and pleasant stimuli. This was her safe place; nothing painful could happen here. The other crib, on the other side of the room, had

only white sheets. This is where dressing changes, baths, blood work, and other procedures took place. The staff purchased a brightly colored apron that they all wore during the painful procedures. This was a trigger that the painful part was about to happen. The baby realized the difference and would recover quickly when placed back in her safe crib.

It is extremely important that you plug into your baby's care cycle. This will offer a valuable bonding opportunity as well as help you prepare for your baby when he or she comes home. Help the nurses give baths and put on lotions and ointments. Taking part in your baby's care will also build your self-confidence and increase your skill at managing your baby's special needs. It will provide the "hands on" experience you will need as you explore doctors and treatment options for the future. Despite many parents' initial instinct to stand on the sidelines and leave things to the "experts," now is the time to become active and learn everything you can about your baby and his or her skin. Remember, your baby is coming home soon, and then you will be in charge. You must have the skills by this time to take over his or her care.

Remember, too, that these first weeks are a time for a mother to rebuild her own strength after the rigors of delivery. You cannot spend all of your time with your baby at the hospital, so don't feel that you should. Don't feel uneasy or guilty about taking time for yourself and the rest of your family. Don't let yourself feel guilty when you are not at the hospital with your child. It helps no one if you allow yourself to become physically and emotionally drained.

BONDING WITH YOUR BABY

After we were finally allowed to hold baby Joshua in sterile blankets and a cap, ourselves outfitted in sterile gloves and gowns, we could not resist the joy of kissing his forehead when no one was looking.

A baby with ichthyosis can be a very different looking baby. In some cases, your child may look frightening. Even parents with "normal" babies often take weeks before they feel especially

close to their infants. It is only natural that you might feel detached and even afraid to hold your baby.

Take a deep breath, relax, and give yourself time. Initial rejection is not abnormal, and it can be worked through. You must allow yourself, your spouse, and your family time to grow close to your new baby. Often, before this can happen, you must cast aside your preconception of the “perfect” baby and get to know the baby you gave birth to. You must put your fear aside and look beyond your baby’s unusual skin. Remember that beneath the skin is a normal infant who needs to be loved. Hold your baby as soon as you can. Hugging, kissing, and stroking your baby will help you bond. Almost at once you will begin discovering your baby’s special qualities. All babies have them.

If for some reason you are told not to handle your baby (some types of ichthyosis prohibit immediate handling), or if you just don’t feel comfortable with the idea at first, do what you can to let your baby know you are there. Hold your baby’s hand, stroke his or her head, read a story or sing a song. Your baby is one of the few people who will never get tired of hearing your voice. In fact, your voice is so soothing to your baby, consider taping yourself reading or singing to comfort him or her while you are away.

BREAST FEEDING YOUR BABY

The first time I ever really felt maternal toward my baby was the day when I could finally turn my back on the bright lights of the neonatal ward, cuddle her on a mountain of pillows and nurse. At last, I felt like she needed me and I was giving her something neither the specialists nor the machines could provide.

Breast-feeding should not be ruled out because your child has ichthyosis or because he or she is in the hospital. Mother’s milk is the most nutritionally sound food for your baby. It promotes optimum health because it provides specific immunities against illness, and proteins that enhance development. Since children with ichthyosis are often susceptible to infection, breast-feeding should be seriously considered when evaluating your baby’s

feeding needs. Breast-feeding also prevents over feeding and offers emotional rewards to both you and your baby. It can help you overcome the feeling of separation you may feel while your baby is in the hospital.

A baby born with a collodion membrane is often capable of breast-feeding. In fact, the area around your baby's mouth is often the first place the membrane begins to shed. If your baby is in an isolette, don't feel intimidated. Cream your baby up and put him or her to the breast. If you and your baby have difficulty getting a good latch, don't give up. Patience is important, for nursing comes naturally only to a lucky few; most moms and babies have to work at it for a while. Ask an experienced nurse for help, and if you continue to have trouble, consider seeking the help of a lactation specialist. Most hospitals will have one on staff who can help you, and if not, your doctor or nurse can recommend one. You can also contact your local LaLeche League.

Some babies, especially those with EHK, need to be handled gently because of the fragility of their skin. If the fragility of your baby's skin makes nursing difficult, electric breast pumps are available for collecting milk for later feeding. Regular pumping will also build up your milk supply for the day you are finally able to nurse. Sometimes the stress of having your baby in the hospital will prevent you from nursing effectively. In these cases, too, using a breast pump and bringing mother's milk to the hospital will help your baby enormously. Then, when your baby does come home, it may not be too late to begin nursing him or her.

If you can't or don't wish to nurse, commercial formulas provide perfectly fine nourishment for your baby. Millions of healthy babies have been raised with the bottle, and yours will do just fine. However, consult your physician about nutrition supplements, such as iron, that may be helpful.

DOCTORS FOR LONG-TERM CARE

When your baby was born, you acquired a group of doctors whom you did not choose. Contact with these physicians gave you a good chance to evaluate specialists that you might turn to for long-term care for your child. At the same time, you are not bound to return to anyone for follow-up visits if your experience with them in the hospital was negative. You may turn instead to other specialists in your community.

As you evaluate specialists, keep in mind that most types of ichthyosis are very rare. You want doctors who have not only an academic, scholarly interest in medicine, but also clinical experience and a special interest in treating ichthyosis. These are the physicians who monitor medical journals, textbooks, scientific meetings, and pharmaceutical data to ensure that your child is benefiting from the latest and best research. Most important of all, you must feel that you can build a long-term professional relationship with the physician, one in which you are working together to manage a common problem.

To find such a physician, contact FIRST and ask for a physician referral in your area. FIRST maintains a list of physicians who are knowledgeable about ichthyosis, are treating other patients with ichthyosis, or who have an interest in seeing patients with ichthyosis. Or, you may want to contact an accredited medical school in your area for a list of names of faculty members who have dermatology practices in your community. A pediatric dermatologist is often a good choice because these subspecialists are used to caring for infants and children with skin diseases. At the very least, find a dermatologist with an interest in ichthyosis. A doctor who might be fine for teenagers with acne, or one who is solely a skin cancer specialist, may or may not have the professional curiosity and motivation necessary to monitor a complex disease in an infant.

You will also need to choose a pediatrician very carefully. For many children with ichthyosis, additional time and attention to skin care will be the only medical issues that set them apart from

other children. However, other issues may be a problem for some children. Minor childhood diseases like chicken pox can be serious in a child with ichthyosis. Some children with ichthyosis do not sweat properly and can overheat dangerously. Some children with ichthyosis cannot wear adhesive bandages because removing them tears the skin. Nutrition and growth can also be a significant secondary concern for your pediatrician and dermatologist to watch carefully.

For most children with ichthyosis, physical, intellectual, and psychological development can be perfectly normal. Some forms of ichthyosis are associated with developmental delays, and in all forms of ichthyosis thickened skin blunts the touch sensation. Physical (motor and fine motor) and cognitive development, vision and eye control, and psychological well-being can all be affected by ichthyosis and require the support of a physician who is scholarly, yet practical, kind and compassionate without being maudlin, and accessible to you when you need advice. Federal and state funding is often available to help cover the costs of such things as delayed motor development, and a doctor can assist you in obtaining such aid. He or she can also be your advocate with insurance companies who are unfamiliar with the condition and therefore deny claims related to your child's disorder.

If you are in an HMO or PPO that requires you to go to a certain clinic or panel of doctors, it is worthwhile scheduling an appointment with an administrator who can help direct you to one physician who will be responsible for your child's overall care. This is to everyone's advantage, since it will avoid having to explain ichthyosis to each new physician you encounter.

It is a good idea to encourage professional relationships between physicians caring for your child. A good dermatologist/pediatrician team will work together on such issues as diet, skin infections, chicken pox, stitches, casts, and other medical concerns that cross specialties where ichthyosis is concerned.

Many parents have found it useful to seek a consultation with an expert in ichthyosis at least once (FIRST can provide you with the names and locations of many of these physicians), even if they have to travel some distance and pay for it out of pocket. This evaluation can serve as a way to make sure that your child's regular physicians are providing state-of-the-art care, and can provide someone to call for a second opinion when future medical decisions must be made.

As your child gets older, talk to him or her about the doctors you have selected. Do they like them? Do they feel comfortable with them? (One mother found that the doctor her three-year-old liked best was the one who provided a cotton child-sized examining gown so she did not feel on display during examinations.) Many teenagers with ichthyosis have expressed a desire that doctors talk to them and not their parents. If your child has a problem with a doctor, discuss it with the physician and consider changing to someone who will lessen your child's anxiety.

Interested doctors and nurses can offer tremendous support. Dermatology nurses, for example, can demonstrate effective ways to apply medications and dressings. Pediatric nurses often have great practical ideas for distracting a child during painful treatments. Nurses can interpret medical terms for you, help you get a doctor on the phone, and give you samples of medications they know you use. Seek them out as allies.

Once you have addressed your child's medical needs, you may want to consider choosing a mental health professional to help you through the traumatic early months of parenting a child with special needs, and to follow your family in the future. Depression can be a problem for people with ichthyosis, and may require intervention by a professional. But less serious psychological issues deserve airing, too. Some parents find it helpful, especially for pre-teens and teenagers, to have a child or adolescent psychologist, or social worker, work through the emotional aspects of what can be a disfiguring condition. It is useful for

adolescents to have this kind of support from someone who is not a family member. Seek references from a trusted physician, a children's hospital, a nearby university hospital, or a state social services agency.

THE FINANCES OF MAJOR MEDICAL EVENTS

When all of the doctor and hospital bills started rolling in, my first response was to panic. My wife and I had set aside money for an average delivery and that was all. I forced myself to push all my worries to the back of my mind and focus all my energy on our new baby. When we knew she was out of the woods, I started dealing with the hospital financial department and our insurance company. I was able, in most cases, to set up long term payment plans that fit into our budget.

Children born with ichthyosis usually spend some time in the hospital, anywhere from a few days to a month or more. Since hospitalization will likely be in a neonatal intensive care unit, the costs can be astronomical. Additionally, throughout infancy and childhood, your child will probably require at least occasional, if not frequent, specialized medical attention. In short, the economics of ichthyosis can become a significant factor in any parent's life.

The first rule of the economics of ichthyosis is *don't worry about it*. In the beginning, put financial worries on the back burner. Instead, focus on your baby and his or her condition. Focus on your family. Learn about ichthyosis and give your attention to getting your child home, where you can create a warm, secure environment.

If You Have Medical Insurance

If you have medical insurance, review your policy. If you don't have your most recent policy handbook, get one. Once you are familiar with your coverage and benefits, schedule an appointment with the person at your place of employment who oversees your company's health care insurance plan to discuss your situation. Be prepared to discuss ichthyosis, your child's prognosis, and the long-term care commitments you may be

facing. However, don't try to predict the future just to answer a question. Where unknowns exist (and there will be plenty of unknowns), make clear to policy administrators and insurance claims adjusters that some questions cannot be answered.

If possible, obtain a copy of your company's policy contract. The fact is, your health insurance carrier is probably unfamiliar with ichthyosis. In the course of filing claims, they will be evaluating unfamiliar medical circumstances to make reimbursement determinations. These determinations frequently will be made on speculative grounds.

You may find that your insurance company occasionally (and perhaps frequently) does not allow coverage or reimbursement for various procedures and/or treatments. You *always* have the right to appeal any and all determinations. Your first line of defense against denial of benefits is being fully informed. If necessary, enlist the assistance of your child's primary physician to support your appeal. Discuss problems with your health insurance administrator at work. If you must, see an attorney. Experience has shown that appeals work.

After your child is born, any change in employment that changes your insurance coverage may bring into play pre-existing condition exclusions. Some insurance companies deny coverage to those who need it most by refusing to insure anyone who has a pre-existing condition; that is, an illness or medical condition that existed prior to the application for coverage. This may apply to any genetic disorder, including ichthyosis. However, some states have made such practices illegal. Find out what the law is in your state. You can usually find the phone number for your state's insurance commission in the blue pages of your local phone book. Or you can access your state's government website by using [www.state.\(your state\).us](http://www.state.(your state).us), i.e. www.state.pa.us.

If You Do Not Have Health Insurance

If you do not have health insurance, don't despair. Your baby will *not* be denied medical care. However, studies show that the

uninsured receive fewer medical services and generally lower quality of care than people who are insured. In order to ensure your child's long-term health-care needs, you may need to take advantage of one or more state and federal assistance programs. Because assistance programs, as well as criteria to qualify for them, vary greatly from state to state, we cannot offer a blanket summary of programs. However, a few things are essential.

Make an appointment with a social worker at the hospital in which your child is being treated (or in your state of residence, if it is different from the location of the hospital). If no social worker is available there, contact your county or state Department of Health and Human Services. The department will be able to tell you where to go to learn about programs available to you and your child. Regardless of other programs available in your state, one program is key: Supplemental Security Income (SSI), which is under the Social Security Administration (SSA). Federally funded and state managed, SSI provides for medically needy people of all ages who meet the criteria for medical and financial need. Perhaps the most important advantage of becoming eligible for SSI benefits is that in most states this will qualify your child for Medicaid, which is essentially state-paid medical insurance.

Experience has shown that SSI/Medicaid coverage for children with ichthyosis is often denied upon initial application. However, coverage is frequently granted upon appeal. The program allows for several levels of appeal. Because coverage, once granted, becomes retroactive to the date of your first application for the program, it is important to begin the process as soon as possible. The process is frequently long and frustrating, but the benefits of coverage far outweigh the difficulties of the application and appeal.



PRACTICAL MATTERS

GENERAL ADVICE ABOUT THE SKIN

In general, the goal in taking care of a child with ichthyosis is to hydrate (moisturize) the skin, hold in the moisture, and keep scale thickness to a minimum. But remember – *ichthyosis is different from person to person. Two people with the same type of ichthyosis may have very different expressions of the condition and their response to the similar treatments may be very different as well.* What works well for one person may not work well for you or your child. For instance, a lotion that works wonderfully for one child may be painful and even harmful for your child.

Your dermatologist and pediatrician are, of course, excellent sources of information. But don't forget a third professional - the pharmacist. A good pharmacist can make life much easier for a family dealing with ichthyosis. He or she can offer suggestions, keep you informed about new products, place special orders for you, and offer you discounts for products you buy in bulk.

In time, you will become the expert in the practical care of your child. You will develop your own regimen by consulting with doctors, talking to other parents of children with ichthyosis, and experimenting on your own. You can keep abreast of what other people are using by following the FIRST's quarterly newsletter, *Ichthyosis Focus*, and by interacting with other parents through the Regional Support Network. (*Always check with your doctor before changing your routine or trying a new product, even if another parent swears by it.*)

In the following sections, we have tried to offer some general guidelines on skin care. *These are only guidelines. You, your pediatrician, and your dermatologist should ultimately design your child's skin care regimen.*

BATHS

Baths are good; more baths are better. Despite popular belief, dry skin does not lack oil it, lacks water. Baths put the water back into the skin, soften scale so it can be scrubbed off without too much difficulty, and are also great splashy playtimes for you and your little one. Keep in mind that soaps rob skin of essential oils, so they should be used sparingly or avoided altogether. Speak to your dermatologist about how much soap you should use. Some types of ichthyosis respond better to the steam in a comfortably warm shower than to complete immersion in a bath. *Experiment to see what works best for your child.*

You might also consult your dermatologist about which bath additive might be best for your child's skin. Doctors often recommend oatmeal-based additives, which you can buy at the grocery store or drugstore, or make at home if you have a food processor to finely grind whole oats. For patients who are at risk for infection, doctors sometimes recommend adding a few drops of antibacterial soap or even bleach (e.g. Clorox®) to the bath. Some shampoos can help reduce scale on the scalp.

If your baby has large raw areas on his or her skin (as many babies with EHK do), put a dab of petroleum jelly on the raw areas before the bath. This helps alleviate the pain of the first contact with the bath water.

Once your child has had time to play and soak, scrub the loose skin off gently with your palms, a wash cloth, or a specially made product designed to loosen skin cells. (Nail salons often carry several kinds of nail filing blocks that are gentle to the skin.) You can also try a loofah or a pumice stone if your child has thick calluses. You will learn with experience how much to scrub. Moderation is the key. If too much skin is removed, there will be

tender (sometimes very red or even raw) skin beneath. It is especially important not to overdo scrubbing off the scale in babies. Some topical medications may be absorbed internally if the skin is overly abraded or raw. Excessive scrubbing can also cause blisters in children with EHK. And scrubbing of any type is not recommended in Netherton Syndrome.

After a bath, thoroughly dry the creases under the neck, under arms and in the diaper area to guard against maceration, which are sores resulting from over-moisturized skin in creases and folds. Only pat dry the rest of the body to keep the skin moist. Immediately apply cream or lotion to seal in that precious moisture.

How often should you give your child a bath? It depends on the condition of the skin and the weather. If you live in a climate with dry, windy, or cold weather, you may have to give your child more frequent baths. If your child is in air-conditioned environments, he or she may also need more frequent baths.

CREAMS, LOTIONS & OINTMENTS

You will want to set up a convenient, colorful, and well-lit area in your home to apply lotions and creams. At first, you may need to coat your baby at every feeding and/or diaper change. With time, though, you will learn by experience what your child's skin needs. Babies will often react better to warmed cream or lotion, as opposed to cold lotion. Try rubbing a scoop full between your hands before applying the cream or lotion on your baby. Or try putting the tightly closed container in the warm bath water while you bathe him or her; the lotion will be warm when the bath is done. Next, moisturize the rest of the child's body. Rub the lotion or cream in well, gently massaging your baby as you go. This can be a nice time for telling stories, singing songs, or just talking to your baby.

Some of the most effective agents to remove scale include alpha-hydroxy acids, such as lactic or glycolic acid. Some products containing these ingredients are available by prescription,

although many over-the-counter products contain them as well. Remember, though, that sometimes it is better not to try to remove the scale, but just to keep the skin supple and well lubricated. Glycerin, urea, and propylene glycol are other effective ingredients that attract water (called humectants) and work well on severely dry skin. Glycerin alone can be purchased from any pharmacy; you can add it to your favorite lotion to help the skin stay moist longer. Skin barrier repair formulas containing ceramides or cholesterol may also improve scaling.

Use your dermatologist's guidance, but at the same time begin learning the active ingredients in the creams and lotions you use. Although there is a dizzying array of topical products available for purchase over the counter, most, you will find, are composed of a relatively small number of key ingredients. At the same time, prices vary considerably, even among remarkably similar products.

Often, a heavier cream, which forms an occlusive barrier, is helpful to seal in moisture once you have applied a humectant. Again, these are most effective when applied immediately after a bath. Many occlusives contain a petrolatum or mineral oil base. Lotions, although easiest to apply, are often relatively ineffective at holding moisture in the skin.

Some products (particularly the alpha-hydroxy acid lotions and creams) may cause stinging or mild irritation especially in small children, so it is wise to discuss their use with your dermatologist. Avoid applying these to areas where the skin is cracked or raw. Furthermore, there is a theoretical risk of systemic (internal) lactic acidosis (accumulation of lactic acid in the blood above normal levels), particularly in infants. Again, work closely with your dermatologist in determining your child's treatment regimen.

DIAPERS AND CLOTHING

Your choice of diapers may depend on what type of ichthyosis your child has. Disposable diapers, particularly the super absorbent type, usually work well on children with ichthyosis

(except those with EHK). They pull most wetness away from direct contact with the skin, yet are occlusive, so they hold in moisture. Because of this, the skin in the diaper area may look better than on other parts of the body.

Babies with EHK often cannot wear disposable diapers because they are too rough on the skin. All-cotton diapers don't wick away moisture, but without consistent use of plastic pants, the moisture will evaporate and the cotton will be the least irritating. To counter the moisture loss, use petroleum jelly or similar occlusive often, and rub it in well. Thickly grease the diaper area to prevent urine or feces from contacting your infant's skin, especially if he or she has deep, open cracks or raw patches.

If your EHK baby has extremely fragile skin, he or she may not be able to wear diapers or clothing at all without blistering and peeling. In this case, buy or make oversized cotton receiving blankets. In the center of one, place a waterproof lap pad (available at baby stores) covered by layers of cotton diapers. Put your baby on top of the diapers without pinning them, and wrap him or her in a loose swaddle. (In the summertime, don't even bother to swaddle. Just place your baby on top "au natural.") If your baby still cannot wear diapers when he or she begins to roll over or crawl, make the lap pad and diaper layers bigger in the playpen. You will go through a lot of receiving blankets and diapers, but the mess will be minimal, and your baby will do better without the irritant of fastened clothing.

Ask your dermatologist about the best method of cleaning your baby at diaper changes. Some recommend unscented diaper wipes without alcohol, cotton balls covered with oil, or just plain water. Another mom suggests placing a roll of toilet paper into a large plastic bag into which a small amount of baby oil has been added. The toilet paper will absorb the oil, and the plastic bag makes the whole ensemble easy to carry in a diaper bag.

Buy clothes large, so they do not rub. If your child's skin is especially sensitive, turn outfits inside out to avoid having the

seams rub. 100% or mostly cotton clothing is the least irritating. Avoid elastic. Avoid polyester, especially acrylic sweaters, since it can irritate the skin. Take your child with you while shopping and rub his or her hand on clothing you are uncertain about. If it catches, do not buy it. Babies and young children tend to scratch themselves until they are raw and bleeding. One piece footed pajamas, outfits, or “onesie” type shirts with overalls tend to prevent scratching to rawness or infection in children under age 3. Most babies, whether they have ichthyosis or not, need nothing on their feet, which can be especially sensitive. This will change once they begin walking, as walking barefoot dries out the feet and pressure from walking can cause cracks in thicker calloused areas such as the heel. Wearing shoes and socks helps keep the feet hydrated and reduces the depth and frequency of the cracks.

Using a heater *and* a humidifier may keep the room warm enough so clothes will not be needed. Also, putting your baby on a lambskin pad in the crib or playpen will help him or her maintain body temperature while providing extra soft padding.

INFECTIONS

People with ichthyosis have an impaired skin barrier function that leaves them more susceptible to infection. Thick moist scale will trap bacteria, yeasts, and fungi. The cracking and fissuring of the skin also leaves the body open to infection. Your dermatologist can help you watch for fungal, yeast and bacterial infections, and teach you how to distinguish between them. All infections require prompt medical attention and can usually be treated with topical prescription ointments and creams.

If your baby’s skin is open or raw, begin with applying the antibiotics suggested by your dermatologist to these areas. Infections may begin as raised white bumps, raw areas that are weeping, or areas covered with a yellowish crust. If your child seems particularly itchy and uncomfortable, or has a low fever without any other obvious symptoms, he or she may have an infection. If your child develops a high temperature, or if the

infection spreads, contact your doctor immediately. Treatment with either oral or parenteral (intramuscular or intravenous) antibiotics may be needed. This is particularly important in newborns, in whom infections can spread rapidly and can be serious.

NUTRITION & ICHTHYOSIS

Because the skin is the largest organ of the body and because it has a very large surface area in contact with the environment, what happens to the skin has a significant impact on the whole body. It is important to realize that the skin requires large amounts of iron, minerals, and particularly protein to continue functioning. Increased loss of skin from the surface puts an enormous strain on one's entire system.

Children with hyperproliferative types of ichthyosis, such as widespread Netherton Syndrome, EHK, and CIE, where there is rapid skin turnover, are at most risk for growth retardation. The nutritional demands are very high for these skin types and are even higher in children who are growing rapidly to begin with. Many children with ichthyosis consume enough calories and nutrition to support normal growth, but they do not consume enough calories to compensate for the increased demand of their ichthyosis. Infants with ichthyosis may need supplemental feedings and nutritional supplements to support their growth. Consulting with a nutritionist may be helpful.

Children with poor skin barriers need foods that are adequate or even "super-adequate" in their protein and iron content just to maintain "normal" growth. Some children with ichthyosis may be smaller than other siblings in the family, which may relate, at least in part, to their nutrition. Thus, a good diet and supplementary vitamins and minerals are important.

Infants with severe forms of ichthyosis may be born prematurely and are small for their gestational age, and when nursing they may suck poorly due to restriction of the inflexible skin around the mouth. The breast, of course, is flexible to the mouth of the infant and is therefore preferred. Failing this, a premie nipple

(one that is more flexible and doesn't require a strong suck to deliver) may be used with the holes slightly enlarged to better deliver milk. Look for nipples labeled "Neonatal," "Premature," or "Natural Flow." Again, breast milk is high in minerals, vitamins, and protein. It is important for mothers to maintain good nutrition so infants will get the highest quality milk. Of course, if formula is used it should be one enriched with iron.

Solid foods should probably be introduced at the normal time, at four to six months of age. Enriched baby cereal is an excellent first food because the protein in the cereal is the best absorbed of all suitable infant proteins. When your baby is ready for more texture, try whole grain cereals.

Many nutritionists feel that children should not be subjected to adult "diets" that are low in cholesterol and fats because children need the cholesterol and the nutrients in all dairy products, and in eggs, for normal body and brain growth. This is particularly true for children with ichthyosis who need whole milk, especially during the growing years.

Managing your child's diet on top of round-the-clock skin care is not easy. Your child's pediatrician or a nutritionist can help you develop a balanced diet plan that is nutritionally and developmentally appropriate for your child.

If you are not already aware of which foods are high in iron, protein, vitamins, and minerals, excellent books are available at many libraries and bookstores. Nutritional services may be available at agencies in your area.

While not all children with ichthyosis experience growth problems, a significant number do. If your child with ichthyosis begins to experience these problems, have your child evaluated by a pediatrician. Consider the following helpful hints:

- Try to keep a written record of everything your child consumes each day. Feed infant cereal until your child is at least 12 to 18

months, since it is the best absorbed of all infant-suitable proteins.

- Offer nutritious and interesting meals and snacks.
- Respect your child's ability to know when he or she has had enough to eat. Offer plenty of liquids, since water is lost through the skin. This is especially important when your child has diarrhea or is vomiting.
- Don't let meal times become burdensome or stressful. Keep feeding times pleasant and relaxed.
- Try planning a daily eating schedule, which might include three meals and three snacks. Keep plenty of healthy snack choices on hand.

OVERHEATING

Children with ichthyosis are more vulnerable than most children to temperature and the weather. Overheating can become a significant problem since the child's thickened skin does not allow the body to get rid of body heat through sweating. During hot weather and when your child is active at play, watch carefully for signs of overheating. The skin may redden, your child may appear cranky, and, when seriously overheated, your child may become listless and lethargic. Overheating is a potentially dangerous situation, and steps must be taken immediately to bring your child's core temperature down to normal levels. Take him or her into a cool environment and, if necessary, put him or her into a lukewarm bath. Applying wet, cool washcloths to the face and extremities may also be helpful if the overheating is not yet too serious. If you are seriously concerned about how overheated your child is, call 911.

The best treatment of overheating is prevention. Make ice water or juice available all the time. A spray bottle of cold water is useful for counteracting the effects of hot weather, for example when riding in a car. A game may be made of spraying with cool

water. For outside play during warm weather, one mom dresses her child in wet clothes. Available, too, are “cool vests,” which are pocketed smocks in which packs of frozen coolant gel are placed. In excessively hot weather, your child may need to stay inside.

If your child spends time in preschool or with any outside caregiver, the person must be aware of the signs of overheating, and must be able to respond to them at once.

When overheating is due to fever, try to keep the fever down with an aspirin substitute (for example, acetaminophen, or Tylenol); you may also use lukewarm baths to cool your child. During a fever, avoid thick creams that can hold in the heat. Many parents have noticed that after a high fever, their child’s skin takes a turn for the worse. This is a temporary setback, but may require extra attention for a few days.

To help avoid overheating due to warm weather, dress your child in layers that can be removed as needed. When out in the sun, use a hat with a wide brim to protect both eyes and skin. Ask your doctor about sunscreens.

FIRST publishes a resource sheet on overheating that details the signs and symptoms of overheating, what to do in a heat emergency, and resources for cooling products and sun protective clothing. Parents may request this resource sheet and copy it to all the individuals who help care for their child.

SYSTEMIC THERAPIES: THE RETINOIDS

Some severe cases of ichthyosis have improved when treated with oral retinoid therapy. The retinoids are a class of pharmaceutical product available only by prescription, and a physician who is experienced in their use *must* monitor their use closely.

The key retinoid drugs, Soriatane® (acetretnin) and Accutane® (isotretinoin), are derivatives of vitamin A, which in its natural form is toxic in high doses. Although these synthetic derivatives

of vitamin A substantially reduce the natural toxicity, they still remain potent and potentially dangerous drugs.

Because retinoids may cause significant effects on bone development, their use in children who are still in their growing years requires careful consideration. Regardless of age, periodic X-rays to monitor bone development are essential. Other side effects include fluctuations in body chemistries (for example, cholesterol, serum triglycerides, and liver function), so retinoid therapy requires a physician to closely monitor the patient, completing essential tests at regular intervals.

Most significant, perhaps, is that the retinoids are highly teratogenic, that is, their presence in the system of a pregnant woman has a high likelihood of causing major birth defects. Even one pill of an oral retinoid can cause birth defects. Women taking oral retinoids must discontinue use of the drug for several months to several years, depending on which retinoids they were using, before attempting to become pregnant.

The beneficial effects of retinoid therapy on the skin will last only while the drug is being taken. When therapy is discontinued, the skin will revert to its previous condition. Therefore, the retinoids are considered long term therapy. Given the dangers associated with their use, one must weigh carefully the advantages and disadvantages. This is particularly true for women of childbearing age and children.

Anyone using retinoid therapy must have a close working relationship with a physician who has sound experience in using these drugs with ichthyosis, and with whom the patient has both trust and confidence.

SPECIAL TIPS

Dressing wounds on babies with EHK

When EHK babies have wounds that must be dressed, a non-stick bandage pad can be held in place by the soft cotton wrap used under casts, or gauze wrapping available at most drug

stores. Wrap the gauze around the entire extremity. Regular stick-on bandages do not stick to EHK skin. If they do, they often rip off healthy skin when they are removed. If your child is hospitalized, oxygen monitors and IVs cannot be held down directly with surgical tape. Have nursing staff wrap the arm with gauze, and then tape the IV to the gauze, not directly to your child's skin.

Watching for infection

Your dermatologist can help you watch for fungal, yeast, and bacterial infections, and teach you how to distinguish between them. All require prompt medical attention and can usually be treated with topical prescription ointments and creams.

Itching

Itching can be treated with oatmeal and oil baths and plenty of cream or lotion. If the itching persists, your doctor can prescribe oral and/or topical medications.

Scalp Scale

To remove difficult scales on the scalp, try applying your favorite lotion liberally to the scalp, then put on a shower cap or other occlusive garment for the night. In the morning, shampoo, massage, and comb out.

Scratching and Fingernails

Infants with ichthyosis often have fast-growing nails. Take care to keep them trimmed below the fingertips. You might want to try adult cuticle nippers instead of baby nail trimmers. Babies tend to scratch the upper arch of the ear, behind the ears, and their noses. You can put socks on the hands of your sleeping infant, but they learn to take them off after a few months. Leaving your baby's hands covered all the time could delay development of hand-eye coordination. If your baby seems to be scratching a lot, he or she may need an extra bath and lotion to feel comfortable, or they may need to be evaluated by your doctor for a skin infection.

Ear Wax and Scale

Just as the skin sloughs off on the areas you can see, it also sheds inside the ear where it can combine with wax and form a solid material that can smell bad and temporarily impair your child's hearing. Remember that infants must hear well to learn to speak well. Have your child's hearing checked regularly and discuss with your pediatrician safe ways to keep your child's ears cleaned of debris from exfoliating skin. Your physician may recommend an earwax remover, sold over the counter. Never stick anything in your baby's ears; this can puncture the eardrum and cause hearing loss. Some ichthyosis patients get relief from pure lanolin. Lanolin is easier to spread when mixed with water.

You can safely clean the arches of the external ear and also behind the ear with a cotton swab. Keeping skin growth under control in these areas will reduce itching and resulting open sores. If your baby seems to constantly rub at or pull at his or her ears, he or she may have a build up of scale in the ears that is causing itching.

It takes time

Give any new cream or other treatment a decent trial before deciding it doesn't work, and don't expect miracles. Some products need to be applied for at least two or three weeks before results become noticeable. If you are comparing treatments, use the regular treatment on most of the body, and apply the new treatment to a small area such as the thigh or forearm only. This way, you can see a visible difference between the two. If the new treatment is ineffective or causes the skin to blister, itch, peel, or dry out, the problem will be limited to the small area.

Body odor

Many people with ichthyosis have a distinctive, sometimes unpleasant, body odor. The problem is usually caused by bacteria trapped in thickened skin or in cracks or creases between thickened scales. It is usually most noticeable in the creases in a baby's neck or in the clenched hands. Try bathing with an antiseptic skin cleanser, or adding two teaspoons of chlorine

bleach to each gallon of bath water. *Consult with your dermatologist for specific recommendations and amounts to use.*

Ectropion

Infants and children with ectropion (eversion or flipping out of the eyelids) may have difficulty with dry, irritated eyes. Many children with ichthyosis sleep with their eyes open. Watch out for a child who rubs his or her eyes, or has frequent scratches around the eyes. Pay attention to pink, puffy eyelids or bloodshot eyes even when the child is not tired or sick. See an ophthalmologist regularly in order to guard against corneal damage that may result from dryness. Your physician may recommend drops or ointments to keep your child's eyes moist.



WHERE TO TURN

I had always wanted a baby girl, and I had a closet full of beautiful baby cloths: pink rompers and dresses with lace. Rachel couldn't wear clothes for almost a year, and I had a very hard time getting rid of the things I had saved for my "dream baby." Eventually I did it, though, when a friend who had been infertile for years had a healthy baby girl, and my joy for her overcame my sense of loss.

COPING

The experts recognize that parents of children with ichthyosis hurt more than the children themselves. The hurt you may be feeling now is undeniable, and many parents before you have experienced it. Here are some tips from other experienced parents that may help you cope with your situation.

Share your feelings with your spouse. Don't blame one another; strengthen each other. Share responsibility for learning about ichthyosis and become active about your child's special needs. If you don't have a spouse, develop a support system among family and friends.

Turn to those who have strengthened you in the past - friends, relatives, counselors, or clergy. If you find that someone you would normally turn to does not offer the support you need, look elsewhere. You may be surprised at which people end up becoming your strongest sources of support.

Remember, you are not alone. Parents who are experienced with ichthyosis may be your best source of information for the

practical day-to-day questions about skin care and other care management concerns. These parents can offer you support, understanding, and encouragement. We encourage you to become active in FIRST's Regional Support Network so you can interact with other parents in circumstances similar to yours. Look into the Internet community as well. Try out the links provided at the end of this booklet.

Take one day at a time. Worrying about negative possibilities will only drain your energy. Keep your attitude about your child's prognosis positive and open. On bad days, visualize an older version of your child being a successful swimmer, a fine musician, a happy and sensitive adult. Your child can only benefit from your good attitude.

You may find yourself thinking about ichthyosis almost constantly at first. Over time, though, you will think about it less and less. As you become accustomed to your child's skin, the lotions, baths, and other parts of the skin-care regimen, the ichthyosis will become a normal part of your life. In fact, you may find that it shrinks in importance beside the normal demands of parenting.

Although relatives and friends can offer wonderful support for you and your child, they may not support you in ways you need because they have difficulty understanding what you are dealing with, or they may find it too painful or overwhelming. Most will be concerned for you and your child, but their lack of knowledge about ichthyosis may cause them frustration and embarrassment. They will not know what to say, so they may be silent; or they may say things that make them feel better, but which leave you empty. Their words might minimize or invalidate your feelings ("It could be worse..." "At least you have one normal child..." "It's only his skin.")

Of course, you can (when you have the energy!) take time to educate others about what you need. Reacting to every thoughtless comment will exhaust you over time and isolate you from needed support. For example, you can say, "It doesn't help

me when you say that, but it would help me if you could just listen and offer support.” Involve family members in giving your child a bath or creaming. The more they see, the more they will understand. Share links to informative websites so they can get the information themselves as well as asking you.

Because you cannot change some people’s feelings, it may be easier to listen to the insensitive ones politely and then move on. Find other friends and supporters who will accept your child as he or she is, and who will support you in the ways that you (not they) truly need.

When you are out in public with your child you will sometimes encounter rude questions, remarks, or stares from strangers. Some of these people are sincerely curious, some are just plain nosy, but all of them are ignorant about ichthyosis.

You can choose to educate these people, or you can ignore them. Tell the stranger as little or as much as you feel comfortable discussing. Depending on how the stranger approaches you (curious, concerned, rude, intrusive, etc.), you might say your child has “very dry skin,” or that “my child was born with a genetic skin disorder.” Some find that using the word “disorder” rather than “disease” lets strangers know quickly that ichthyosis is not contagious. Many will compare their own experiences with eczema or psoriasis and suggest their favorite lotions or treatments. Sometimes it is easier to simply thank them for the suggestion and end the conversation. To rude or pushy strangers, you might respond strongly, saying something like “I’m sure you don’t mean to be cruel, but I am upset by your intrusion.” Older children are often self-conscious of their skin and prefer that you ask them for permission to discuss their skin. Experiment to find which responses work best for you in different situations.

GRANDPARENTS

The most devastating thing my parents did was offer to raise our baby for us. It signified a complete lack of faith in our abilities, and made us feel that we could never turn to them for support in difficult times. It

took years to heal our relationship with them, and the irony is, they were just trying to help.

Your child's birth may profoundly affect the grandparents, who may have to deal with many of the same issues you dealt with, but without the benefit of having control over the circumstances. They, too, imagined the birth of a perfect grandchild whom they could boast about, display pictures of, and spoil with clothes, toys, and unbounded love.

Some grandparents may experience guilt if the ichthyosis is genetically traced to them. Others may react by focusing too much attention on the condition, becoming overbearing; or the opposite, by pulling away from the grandchild to avoid dealing with emotional issues. However, just as most parents do, they will probably overcome their initial reactions and become tremendous supporters and advocates over time.

In times of trouble, we tend to turn to our spouses first, and then to our parents - no matter how long it has been since we lived under their roof. We hope that our parents will respond to our needs calmly, and that they will be pillars of support. And sometimes they are. If your parents fall into this category, be grateful and let them help, always being careful that they do not exert undue control, which is rightly yours.

If your parents do not immediately offer the help you need, don't despair. If they are overbearing, talk to them. Explain that you appreciate their help, but that you are capable of parenting your child. When you ask them for help, be specific about what you want or need. Generalized requests for help might inadvertently be construed as an invitation to take over.

Here are some suggestions from parents on "How to Be a Terrific Grandparent:"

- Listen. Take cues from your children about what they need. Bolster their self-confidence and take note of situations they

handle well. It may be difficult for you to know how to deal with your child's depression and bad days. During these times, avoid invalidating comments like, "Look on the bright side!" or "Don't think that way."

- Express faith and admiration in your daughter-in-law or son-in-law as well as in your own child.
- If you feel guilt, despair, hopelessness, or anger about your grandchild, express it, but to a friend, counselor, or clergyman. Don't burden your child with your feelings, even though your feelings are completely normal.
- Get to know your special grandchild beyond the skin condition. You can play a key role in bolstering your grandchild's self-esteem.
- Occasionally chip in to help buy medications or lotions. The expense of these can be a terrible burden for a young family.
- When buying clothes, first get advice about materials, styles, and sizes that will be comfortable.
- Avoid favoritism to a certain grandchild, either those with normal skin or with ichthyosis.
- Offer attention and time to siblings during the times when they get lost in the shuffle as their parents care for the special needs child.
- Once in a while, spoil your child and their spouse. Give them theater tickets and offer to baby sit.
- If you can, learn your grandchild's skin regimen and relieve the parents of their duties once in a while.
- Learn about ichthyosis, and make contacts that your children may not have time for. Subscribe to the *Ichthyosis Focus*

newsletter. If you come upon a new treatment, medication or research project, *offer* to share the information with the parents. Don't insist they follow a certain course, or voice doubts about their abilities to care for their child.

SIBLINGS AND THEIR SPECIAL NEEDS

I don't have ichthyosis, but my brothers do. Whenever we went out in public, like to the swimming pool, people would ask me what happened to my brothers. I always felt very protective and wondered why people bothered asking. What is the big deal? I have to admit; they sometimes did have a smell, but not a bad one. My little brother used to ask me to grease up his back with lotion. I used to hate doing it, but later I felt proud. My siblings have turned out to be such normal people with happy, productive lives.

Unaffected siblings may sometimes feel lost because their brother or sister with ichthyosis gets more of mom and dad's time. Parents have to be sensitive to the troubling emotions that siblings of children with ichthyosis sometimes experience.

Children know more than we give them credit for. It is a mistake to assume that children can't understand the unknown. Parents need to make sure siblings understand ichthyosis so it does not become frightening and mysterious. Explain the disease in words siblings can understand, and reinforce that ichthyosis is not their fault. When they are old enough, they must understand the genetics of ichthyosis long before they plan to marry and have children of their own.

Siblings may sometimes feel anger and resentment towards their brother or sister with ichthyosis. They may feel jealous, neglected, or rejected as they watch most of their parents' attention, energy and psychological support flow to the child with special needs. Parents should acknowledge these natural feelings and reassure siblings of their importance in the family. Special time should be set aside for siblings in order to help build their sometimes-fragile self-esteem.

On the other hand, the experience of a sibling may be a positive, enriching one that teaches them to accept other people as they are. Some become deeply involved in helping parents care for the child with ichthyosis, sometimes assuming responsibility beyond their years. However, the roles should be clear, and a sibling should never be made to feel responsible for care giving duties.

It is not uncommon for siblings to become ardent protectors and supporters of their brother or sister with ichthyosis, or to experience feelings of great joy in watching him or her achieve gains. Increased maturity, responsibility, altruism and tolerance, humanitarian concerns, a sense of closeness in the family, self-confidence, and independence are among the other positive effects you may observe in siblings.

Ultimately, ichthyosis is a family matter, regardless of how many in the family are affected with the disorder. Everyone in the family will have issues to deal with, will have good days and bad, and everyone's well-being is important.

DEALING WITH GRIEF

When my daughter was born with EHK, I started to feel sorry for her and for myself. I knew in my heart when she was only three weeks old that she would never go to the prom or get married. Finally, my dermatologist told me to stop trying to predict the future and that my daughter would do exactly as she pleased. She said she couldn't wait to dance at my daughter's wedding.

You have just spent an entire pregnancy expecting and hoping for a healthy, beautiful, perfect baby. Giving birth to a child who does not meet these normal expectations can be traumatic. Under these extraordinary circumstances, it is normal to feel powerful emotions; anger, guilt, fear and hopelessness, to name just a few. However, as you become more familiar with ichthyosis and the care giving routine, initial fears will give way to relief, and eventually optimism. Look ahead and visualize happy times, like witnessing your baby's first toothy grin or first messy meal.

Many parents' reaction is to blame themselves for everything that is wrong with their child, even their genetic coding. Some mothers consider themselves inadequate or defective because they didn't deliver the "perfect" baby, despite the fact that the origin of ichthyosis is an act of nature determined at the point of conception. Allow yourself to coast in and out of conflicting emotions without berating yourself for feeling differently than you assume (or someone else assumes) you should. If you feel incapable of handling your emotions, talk to someone close to you. Lean on a social worker, pastor, family member, or best friend. Many parents have found that getting in touch with other parents of children with ichthyosis provides an invaluable source of support. You can draw on FIRST and its Regional Support Network to connect with other parents who have been in your shoes. Consider attending a national family conference sponsored by FIRST. Family conferences offer the opportunity to meet with medical experts and other families affected by ichthyosis.



LOOKING AHEAD

Once I stopped blaming myself and came to terms with it, it seemed to just flow into Ben's life, and he became a happy, generous little boy, so different from the child who used to be lazy, moaning, and overwhelmed by the sense that he was different.

RAISING A HAPPY, WELL-ADJUSTED CHILD

All parents of children with ichthyosis hope and wish for improvements in their child's physical condition. This is only natural. Wouldn't it be nice if she could wear normal shoes, or if he could walk into a store without drawing stares? Yet, these are dreams that rely on the superficial assumption that "beauty is only skin deep." Beauty is the inner person we want our children to become: a person who is bright and kind, loving and humorous, thoughtful and generous.

Dr. Wilmer Betts*, a psychiatrist and father of four children, three of whom have ichthyosis, knows for a fact that children with ichthyosis can develop into successful, self-assured people. He urges parents to put more energy into accepting ichthyosis than in trying to find a cure for it; more energy into raising an emotionally well-adjusted child who copes well with the disorder than into elaborate physical treatments and endless searches for the "right" doctor. It is crucial, he says, for parents to accept that they didn't cause the disease, nor can they cure it. A parent's job is to accept it as a chronic disorder, relieve discomfort (both physical and emotional), reduce cosmetic problems, and help their child develop a healthy ego and a strong personality.

*Deceased

Parents should strive to develop an objective, unemotional attitude about the ichthyosis and its treatment, and separate this one characteristic from the child himself. If you need to give a painful treatment, explain it (don't lie and say it won't hurt), and then do it, preferably while distracting your child with some fun game, song, or activity. Don't let yourself get delayed by crying or excuses. This just prolongs the bad moment for both of you and sets you up for a more elaborate tactic the next time. Imaginative games during treatments work wonders for some children. Be Goldilocks during one treatment, a low-voiced bear the next. It will relieve the stress for everyone if you are not wincing, your baby screaming, and the treatment lasting seemingly forever.

Parents have a legal right to be with their minor children during any medical visits. Calm your child during these times. Carry puppets, toys, or other diversions. Again, don't lie to your child about treatments. Explain what will happen, why they need it, and how long it will take. Remind them that the quicker they get through it, the sooner they will be able to do that special activity you have already described: feeding the ducks, reading a special story, whatever.

Touch your child often, and frequently express your love and pride. If others make hurtful comments, teach them by example how to deal with rudeness and cruelty in a dignified way. Later, let them know that those people should be pitied for their ignorant, mean attitudes. In this way, you teach sensitivity and remove the onus from your child. The fact is, although your child has a problem with their skin's metabolism, this is a far less serious problem than the handicap of a person who lacks sensitivity and tolerance.

Avoid talking about your child as if he or she were invisible. Never ask your child to pull down her sock and show your neighbor how nice the ankles are looking this week. Your child is not a specimen. At a very early age children become sensitive to how you treat them. Be sensitive to your child's privacy and, most of all, dignity.

As your child grows, do not let their skin become an excuse to shelter themselves and avoid activities and people. Furthermore, a child with ichthyosis should be held to the same standards of good behavior that you would expect of any other child. It is not okay for your child to be rude or selfish. It is not okay for your child to hit you, or scream vicious things at you, even if you are administering a painful treatment. Set firm and specific rules in your household and be consistent in their application. Inconsistency can result in behavioral problems for your child with ichthyosis, and can breed resentment in their siblings.

The sooner you allow your child to take over the care and responsibility for their skin, the better. Every child needs to exert independence. Children as young as three can begin putting on lotions or scrubbing skin with a pumice stone. (At Foundation conferences, young peoples' most frequent comment is how they wish their parents would let them do more for themselves, and this includes decision-making.) A 12-year-old should be able to help weigh the benefits and risks of playing soccer, for instance.

Within reason, let your child pick out school and play clothes, making sure there are plenty of things in the closet that do not cause irritation. Soon they will learn to dress for the weather and skin comfort. Over time, you will no longer get into battles at the closet, and your child will become more self-confident.

Encourage your child to have friends. While they should know you are always there for them, early on they will begin needing support from others beyond the immediate family. Suggest to your child that they contact FIRST and ask to be connected with another child close to them, or with the same type of ichthyosis. Closer to home, adolescents may need an adult other than mom or dad to speak with. This may be a special aunt, a counselor, clergyman, or a teacher.

You should resist your instinct to overprotect your child and shield them from outside contact, even if it means a few hard knocks.

CHILD CARE & PRESCHOOL

On the first day of school, Ryan's kindergarten teacher sat down with the children in a circle and they had a talk about skin. What does our skin do for us? Are there different kinds of skin? Oh yes, there is brown skin and white skin and tan skin. There is freckled skin and skin with wrinkles and dry skin. All of the children were praised for their uniqueness and encouraged to touch each other's arms to experience the different colors and textures. Ryan's skin was just as fine and different as everyone else's, more interesting, perhaps, than Alice's, but not really as fascinating as Nathan's hundreds of freckles and two skinned knees.

When parents of children with ichthyosis talk with one another, they quickly learn there are many ways of parenting these special children. One mother keeps her child inside and only sends him to school in gloves, to protect his hands. One dad decides to let his daughter play intramural basketball, even though he knows the sport will take its toll on her skin. Another mother has to return to work six weeks after giving birth, even though it means training a babysitter in complex medical regimens. As you make decisions about your family's future, keep in mind that children are different, the severity of their condition is different, and parents are different, too.

Some families can afford to have a parent stay at home full time with their child. This may put the couple at ease knowing that their child is getting the most loving, competent care there is. For others, this would be a major financial or emotional strain. If you need childcare during infancy, the ideal caregiver may be a family member.

You may find that a caregiver who dealt wonderfully with your first child (without ichthyosis) cannot cope with a special needs baby. If this is so, don't feel guilty about finding a new caregiver. In fact, your decision might be met with relief by a person who feels obligated to stay, but knows she cannot cope with the many challenges of caring for a child with ichthyosis.

If your baby's ichthyosis is painful, seek a caregiver who is

compassionate, but able to demonstrate a nurse's detachment during treatments. A caregiver for such a baby must be intelligent, flexible, a quick thinker, reliable, and a lover of children. You do not want someone who is competent, but cold. You can check the availability of licensed practical nurses in your area, or a nanny or caregiver with an interest or experience in medical care.

Such people do exist. Many of us have found wonderful devoted caregivers whose loving care helped us realize we are not the only people who can cherish and care for our children.

This is just as important for families where a parent stays home to care for the children. "Burn-out" can flourish when a parent rarely gets a break from the hard work of raising a family and keeping a home on top of the responsibility of caring for a special needs child. Find a friend, relative, or mature high school or college student and train them to care for your child so you can get away occasionally from your family responsibilities. In many areas, state or local social service agencies provide access to respite care, a program providing temporary in-home care of children with special needs so that primary caregivers can have much needed breaks.

Eventually, most parents will want to explore outside childcare. The time will come when your child will need to begin interacting with other children in a play group, preschool, or school setting, if only for a few hours a week.

Most experts do not recommend home schooling for children with special needs unless their physical condition would be compromised by contact with others. Children whose conditions are "disfiguring" are likely to do best if they interact with other children at the earliest possible age. Two and three year olds take people "as they come" better than seven year olds do.

Day-to-day contact and play with peers is a normal part of childhood development. Beginning these experiences early will

offer your child essential social skills, and will give your child friends to rely on later. A child who has spent the first five years of life isolated at home will enter school not knowing how to share, how to cooperate, or how to play affectionately with other children. Such children may have problems adjusting to school, not because of ichthyosis, but because they lived in a cloistered world.

Check out preschools and schools as soon as you can. By law, public schools must accommodate all children. Most private ones will too. But seek a school that will embrace uniqueness gladly and willingly. Schools that welcome people of many cultures and backgrounds are likely to welcome children with special needs. (Some even state in their brochures that they seek applications from children with disabilities and/or a special need because they feel this kind of interaction is valuable for all children.)

After you have visited several schools to compare their philosophies, environments, and teaching styles, it may be time to meet with the administrators of the ones you like best. In an objective way, explain ichthyosis and describe your own child's condition and needs. Be careful to neither exaggerate nor downplay the severity of the condition. Ask whether the administrator believes the school can benefit your child. You might want to give the administrator some published information on ichthyosis, along with a letter from your child's doctor stating that your child's ichthyosis is not contagious and that he or she would likely fare well in child care.

Once your child is accepted into a program, offer to do an in-service with the center's teachers. If your child would like to participate, let your child help demonstrate how to apply medications or lotions. Explain what an infection looks like and how to treat it. Explain what to do for a scrape or very dry skin areas, and be sure to explain the warning signs of overheating.

Encourage teachers to treat your child just like all of the other children. Preschool should be a place to play, get dirty, be silly, and make messy art projects.

Dress your child in comfortable clothes that he or she can get dirty. Keep extra changes of clothes at school and plenty of necessary medications, lotions, and sunscreen. Water play should be fine if lotion is used afterward. On hot days, put a spray bottle of water in your child's cubby and remind your child to spritz to prevent overheating. Make sure the teacher knows why your child must do this. Even the sandbox may be fine for most children, although on days when your child's skin is raw or open, you might ask the teacher to encourage alternative activities.

This said, you might be thinking "why should I let my kid get dirty and sweaty, not to mention possibly scraped up?" The answer is, because it's part of being a kid. An occasional extra blister or scrape and some extra baths are a small price to pay for a childhood full of fun, games, and learning-by-doing.

ADDING TO YOUR FAMILY

The occurrence of ichthyosis is unexpected in most families. The more you know about ichthyosis and how it came to be in your family, the better prepared you will be to make decisions about adding more children to your family. The hospital where you delivered your child with ichthyosis can refer you to geneticists and genetic counselors who can discuss the genetics of ichthyosis with you and inform you about the probability of future children having ichthyosis.

Prenatal tests can diagnose some forms of ichthyosis before a child is born. Depending on the type of ichthyosis involved, these tests may include amniocentesis, chorionic villus sampling (which provides the material for genetic sequence analysis), biochemical tests, or microscopic examination of a fetal skin biopsy. Genetic testing may be done to obtain information about the baby that might be helpful for delivery, or for making decisions about continuing the pregnancy.

You should know that prenatal tests are not always one hundred percent accurate. Further, the tests can be expensive and your health insurance may not cover them. Some may also pose a risk

to the fetus. Prenatal testing should be discussed with your physician, and you should have a clear understanding of the risks, benefits, and reliability of any available testing.

Pre-implantation diagnosis is a relatively new technique in which embryos are created outside the body through in vitro fertilization. Genetic testing is performed on the embryos. Only the embryos that did not inherit the mutation are transferred into the woman's womb to create a pregnancy. This method is performed at only a few specialized centers in the world. Because this procedure is invasive and has benefits and limitations, it is best to have a thorough discussion with your physician and the specialists who perform the procedure before you consider pre-implantation diagnosis.



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