

Ichthyosis Focus



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Enlightened Therapy of the Disorders of Cornification

Peter M. Elias, M.D. and Mary L. Williams, M.D.



Peter M. Elias, M.D. and
Mary L. Williams, M.D.

Toward Enlightened Therapeutics for Ichthyosis

Treatment of the ichthyoses and related disorders, in which a thickened or otherwise abnormal stratum corneum (SC) (outermost, non-living skin layer) is a key component, has traditionally focused on the dual goals of: 1) removing scale to improve appearance; and 2) enhancing SC moisturization to improve pliability and comfort. For the most part, these treatments have been neither disease-specific nor based upon new knowledge about disease pathogenesis. For example, the same therapies often are used for both ichthyoses associated with increased scale retention, such as recessive X-linked ichthyosis (RXLI), and for hyperproliferative disorders, such as non-bullous congenital ichthyosiform erythroderma (CIE). In addition to variable effectiveness, in some instances these therapies can be counterproductive. For example, treatments that peel off the SC may "overshoot the mark" and result in decreased function (e.g., decreased frictional resistance (blisters) that can occur with retinoid treatment of epidermolytic hyperkeratosis (EHK)).

Treatment, in an era of "enlightened therapeutics," should attempt to correct the underlying defect. When this goal is neither possible nor practical, treatments can still

be deployed to address the underlying pathophysiology of the disorder. Unfortunately, the first principle (i.e., correction of the primary or underlying genetic defect) remains an unattained goal, because gene therapy is still in its infancy. Yet, it is possible to apply current understanding of disease pathogenesis towards disease-specific therapy that should be both more appropriate and more effective.

During the past two decades, the underlying genetic causes of the inherited disorders of cornification have been largely deciphered. The causative defects have been surprisingly broad, and include not only inborn errors of lipid (fat) metabolism, but also mutations that affect epidermal structural proteins, as well as defects in cellular communication, signaling or proliferation (Table, page 6). Yet, because a wide variety of defects lead to scaling, only a few, final common pathways account for disease pathogenesis. In other words, the epidermis can either: a) fall apart (as it does in epidermolysis bullosa simplex, EHK, and Netherton's syndrome); b) it can apoptose ('kill off a bad cell', as it does in Darier disease); and/or c) it can become hyperkeratotic (thickened and scaly, as it does by definition in not only the ichthyoses, but even psoriasis). Hyperkeratosis, in turn, can arise through either a delay in desquamation (shedding), and/or as a consequence of increased epidermal cell production (hyperplasia). However, only in a few instances is the hyperkeratosis due solely to a failure to desquamate (i.e., a retention hyperkeratosis, as in RXLI). Most commonly, there is a component of epidermal hyperproliferation, as well. Therefore, understanding the

biology of both desquamation and epidermal hyperplasia, as well as their causes and consequences, is critical in designing appropriate therapies for these disorders. Recent advances in knowledge of SC function and its metabolic regulation have provided insights into the pathogenesis of several of the ichthyoses, with potential new therapeutic implications.

Overview of Stratum Corneum (SC) Function and the Mechanism of Scaling

The SC is composed of multiple layers (normally 12-15 layers) of cells (corneocytes). Corneocytes are specialized to provide a tough, yet flexible barrier to mechanical injury. Since they are devoid of nuclei and other organelles they are considered 'dead.' These cells are filled with keratin filaments, as well as amino acids and other small molecules derived from the breakdown of filaggrin, a protein that surrounds the keratin filaments. A highly cross-linked

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**Are You a Member
in Good Standing?**

Members in
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are entitled to
important benefits.
See page 2 for details.

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Correspondence Corner

Dear Members of F.I.R.S.T.:

I am a research fellow working with Dr. Catherine Read, a nursing professor at Boston College. Dr. Read is conducting a survey entitled, "Psychological Response to Genetic Information." It would be very helpful to us to have participants who have had personal experience with genetic disorders. The survey asks questions about the feelings people have when they know they carry a disease-related gene.

Dr. Read's website is: <http://www2.bc.edu/~readca/>

Thank you.

Sincerely,

Donna Perry, RN, MS
Boston, MA

Dear Friends:

This is my reply to Deep Koshla's question about having lasik surgery with ichthyosis:

I have lamellar ichthyosis and suffer from dry eyes. I was told early on in my life, when I had to get glasses, that I would not be able to get contacts because of my eyes drying out too fast. I dealt with this for years. I just had my regular glasses and a pair of prescription sunglasses, and switched between the two. However, a little over two years ago, I lost my sunglasses on a roller coaster ride and decided I would get tested for the lasik eye surgery.

After several tests with my eye doctor and talking with my dermatologist, the doctors determined that I would be a candidate for the procedure. The biggest warning they gave me was to stay out of really windy or cold conditions without eye protection for about three months after the surgery and to **always** carry lots of moisturizing eye drops with me. It is key to keep your eyes very moist during the healing process.

I had the surgery a little over two years ago and have loved every minute since. My appointment was at 3:00 p.m. and I was home at 3:30 p.m. The next morning I had an appointment at 8:00 a.m. and my vision was 20/20. Not bad for the big "E" being blurry less than twenty-four hours before.

Between waking up in the morning and the clock being clear and getting to wear off-the-shelf sun-
glasses, I have been really happy.

Sincerely,

Martin Haas
Rochester, NY

Are You a Member in Good Standing?

The Foundation for Ichthyosis, like all non-profits, is very conscious of its budget. Keeping our expenses as low as possible is a constant concern and our responsibility. To maintain this responsibility, our staff continually updates our database in an effort to have the most current information possible for our mailings. As we've been examining each record in the database, it has come to our attention that there are many of you whose memberships have lapsed. Please remember that membership donations are required on an annual basis to remain a "member in good standing." As a "member in good standing," you receive all the benefits of membership, including the *Ichthyosis Focus* and skin care product samples. Increasing postage, administrative, and production costs make it difficult to provide our services without adequate yearly support from our members.

Our newsletter is consistently mailed every three months. To continue receiving this valuable resource, we need to hear from you. Our annual membership donation is now \$40.00. If you have let your membership lapse, don't worry. Simply contact us at 1-800-545-3286 to reactivate your membership. The Foundation can accept your donation by phone with a credit card (MasterCard, American Express, or Visa). If you cannot afford the annual membership donation, please contact us.

In Memory of Karen J. Jones

The Foundation lost one of its strongest advocates last year on November 12. Karen J. Jones lost her battle with cancer in her hometown of Wilcox, PA. Karen was the mother of Tracie Pretak and the grandmother of Bailey Jones, a young woman affected with lamellar ichthyosis. Karen is survived by her husband, Jerold, her four children, and her six grandchildren. Karen worked as a part-time secretary for most of her life, either for her church or local businesses. She was deeply loved.

When Tracie gave birth to Bailey, almost 18 years ago, she relied on her mother's strength and faith to guide her through the rough patches. Tracie was a young college student when Bailey was born. Karen took Bailey into her home and raised her for the first few years, so that Tracie could finish school and earn a degree. Karen taught Bailey, "God does not make mistakes. You are meant to be."

Karen was a passionate believer in educating the public about ichthyosis. Whenever an opportunity arose to tell someone about the disease, Karen took it. She would leave brochures in her hotel room or with a tip for a waitress. She never left home without an armful of information about ichthyosis and the Foundation. On one occasion, Karen took Bailey on vacation. As they approached the hotel pool to go for a swim, the manager told Karen that Bailey could not swim in the water because of her skin. Karen quickly pulled out brochures and handed them to all the guests lounging around the pool and asked if anyone objected to Bailey swimming. Every guest said "No," and Bailey proceeded to have fun with her Grandma all afternoon.

Karen taught Tracie to "do whatever you can to help other people" and Tracie has lived by those words. Tracie has given back to the Foundation in so many ways, especially by performing spiritual concerts with Bailey to educate others about ichthyosis and the Foundation. Even during this most difficult time, when Tracie and Bailey have lost their best friend, they are thinking of the Foundation. All donations sent in loving memory of Karen J. Jones have been designated to supporting the programs and services of the Foundation.

News & Notes

Pachyonychia Congenita Project

The Pachyonychia Congenita Project (PC Project) was launched in 2003 with the mission statement, "Find a cure for PC." The PC Project is enthusiastically supported by the Pachyonychia Congenita Fund, a charitable organization established by the Schwartz family of Salt Lake City, Utah. The PC Project will provide research grants and Career Development Awards, host research symposiums, sponsor a PC registry, foster a PC community, support genetic testing, and take other active steps towards achieving the mission statement.

Pachyonychia Congenita (PC) is a rare genetic skin disorder characterized by thick, discolored nails, as well as calluses and blisters on the hands and feet. Other symptoms include a whitish tongue, wart-like bumps on the body, cysts, and sometimes hair and laryngeal changes. PC is often listed with other scaly skin disorders and is also sometimes classified as an ichthyosis. Though often viewed as primarily a nail disorder, frequently the major challenge of PC is walking with constant pain. The nails become infected at times and are also very painful.

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Enlightened Therapy

(continued from page 1)

protein shell, the cornified envelope, surrounds the corneocyte, which together with keratin filaments accounts for both the flexibility and mechanical resilience of the SC. It is the presence of small, intracellular filaggrin-breakdown products that render the corneocyte capable of absorbing and binding water.

Surrounding the corneocytes are lipids (fats) organized into membrane stacks (lamellar bilayers). These membranes derive from lamellar bodies (LB). LB are small packages inside the outer epidermal cells that secrete their lipid contents into the intercellular spaces, between the corneocytes, just as the SC is forming. These water-repellant lipids waterproof the skin, preventing loss of excess body fluids into the dry, external atmosphere. They also block the entry of potentially toxic chemicals, environmental antigens, and microbes into the body.

In the lower SC, specialized protein structures, called corneodesmosomes (CD), span the intercellular spaces at regular intervals, forming links or welds between adjacent corneocytes. Normally, CD are progressively degraded as corneocytes move outward through the SC, allowing invisible shedding of corneocytes as single cells. The progressive loss of CD is achieved by one or more protein-digesting enzymes (serine proteases), which in turn, are regulated (kept in an inactive state) by a family of serine protease inhibitors. The desquamatory proteases are also packaged in lamellar bodies and co-delivered to the SC intercellular spaces along with membrane lipids (fats). Thus, SC retention can occur either when the serine proteases are excessively inhibited (as in X-linked ichthyosis), or when insufficient proteases are secreted (as in ichthyoses characterized by reduced secretion of lipids and enzymes, as in Harlequin ichthyosis and CIE). In contrast, absence of a key regulatory serine protease inhibitor (LEKTI 1), as occurs in Netherton's syndrome, can lead to premature shedding (thinning) of SC, with a potentially devastating loss of barrier function.

Desquamation normally results from the repeated, mild mechanical frictions of everyday life. It is enhanced during bathing, when corneocytes first hydrate and swell, followed by dehydration and shrinking. Both friction and swelling/shrinkage

weaken intercellular attachments in the SC. Therefore, SC retention can occur when corneocytes are unable to attract water osmotically (as occurs in ichthyosis vulgaris due to a deficiency in filaggrin breakdown products).

Although the SC is 'dead'; it nevertheless functions as an exquisite biosensor, sending signals to lower skin layers in response to changes in humidity or external trauma. Because permeability barrier function is so critical for survival, it is not surprising that the underlying skin layers are closely attuned to SC function. Any acute or sustained insult that results in barrier compromise, stimulates homeostatic repair responses, including both increased synthesis and secretion of lipids and a mitogenic stimulus to the underlying living cell layers of the epidermis ("Make more lipid! Make more cells!"). These repair responses are signaled by: 1) changes in epidermal ion gradients (i.e., calcium and other ions normally are concentrated in the upper epidermis; barrier perturbations flush away this gradient, thus signaling fat and enzyme secretion) and; 2) release/activation of molecules, called cytokines (e.g., IL-1a, IL-1b, TNFa), that, among other functions, recruit white blood cells that cause inflammation, initiating a downstream cytokine cascade that can lead to both epidermal hyperplasia and inflammation. Since the barrier is abnormal in several ichthyoses, these signals continue to be sent, resulting in the erythroderma (red skin) and hyperplasia (thickened skin).

Application of Enlightened Therapeutics to Specific Disorders of Cornification

Ichthyosis Vulgaris: While the genetic basis for this relatively common disorder remains to be elucidated, the net result is a marked deficiency in the protein, filaggrin. The decrease in filaggrin, in turn, leads to a marked reduction in the levels of water-holding, metabolites within the corneocyte, resulting in an inability to hydrate the SC normally. The flexural sparing that is so typical for this disorder reflects the increased hydration of these body regions, resulting in normalization of shedding. Although a specific replacement formulation is not available, use of humectant-containing moisturizers, such as urea and/or glycerol-containing creams, represent logical strategies in this disorder (Table, page 6). However, a limiting factor in the treatment of ichthyosis vulgaris can be the com-

mon co-occurrence of atopic dermatitis, which renders the patient more vulnerable to some potentially-irritating topical humectants (in such cases, therefore, alpha-hydroxy acids should be avoided).

Recessive X-linked Ichthyosis (RXLI): This not-uncommon, X-linked trait is caused by a deficiency of the enzyme, steroid sulfatase, causing an accumulation of one of its substrates, cholesterol sulfate, and a deficiency of its product, cholesterol, in the extra-lamellar membranes of the SC. While this lipid (fat) imbalance leads to a minor abnormality in barrier function, the predominant clinical issue in RXLI is impaired shedding. SC retention in RXLI can be attributed to a direct inhibitory effect of the excess cholesterol sulfate on the serine protease enzymes that digest corneodesmosomes. This disorder responds relatively well to treatment with alpha-hydroxy acids, which accelerate shedding of outer SC. Cholesterol-containing emollients also improve the scaling abnormality. Hence, an ideal treatment might combine an alpha-hydroxy acid with a barrier repair formulation that is enriched in, or dominated by, cholesterol (Table, page 6).

The Autosomal Recessive Congenital Ichthyoses: This genetically heterogeneous (mixed) group is characterized by a congenital onset of generalized scaling. It includes patients with a deficiency of epidermal transglutaminase 1, resulting in classic lamellar ichthyosis, and patients with non-bullous congenital ichthyosiform erythroderma (CIE). Although the basis for CIE phenotypes differs, some patients display mutations in two enzymes of fatty acids metabolism (lipooxygenases). All these patients display a hyperproliferative epidermis with variable scaling and erythroderma (redness). The hyperproliferative state is secondary in most, if not all, instances to abnormalities in the quantity or quality of intercellular membrane lipids (fats), resulting in a barrier abnormality. The barrier defect, in turn, results in the release of signals that stimulate hyperproliferation and inflammation. In many instances, fats are not fully secreted, but remain entombed within corneocytes. Failure to secrete fats and enzymes leads not only to a deficiency in lamellar membranes, but also to decreased rates of corneodesmosome breakdown (decreased shedding). In Harlequin ichthyosis, the

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most severe entity in this group of diseases, these problems are present in their most extreme form, due to a virtual absence of both intercellular fats and enzymes. The ideal treatment, based upon pathogenic considerations, would include a barrier repair formulation, combined with a serine protease, and/or short-term therapy with an oral retinoid, since retinoids promote shedding of corneodesmosomes (Table, page 6).

Epidermolytic Hyperkeratosis (EHK):

EHK is caused by a mutation in one of the keratins (K1, K10 or K2e) that are normally expressed in the outer, nucleated cell layers of the epidermis. These are called dominant-negative disorders, meaning that the mutant keratin pairs with a normal keratin, thereby interfering with the normal function of the keratin network within the cell. Normally, these keratins form protein filaments (cables) that loop between the cell membrane and the nucleus. Disruption of these cables produces a cell that is poorly resistant to mechanical trauma, and therefore were susceptible to trauma (blistering). Moreover, the abnormal keratins also interfere with the delivery of fats and enzymes into the intercellular spaces of the SC. Hence, instead of secretion into the intercellular domains, the fats and enzymes largely remain entombed within corneocytes in the SC. Thus, the pathophysiology is similar to CIE phenotypes (i.e., defi-

ciency in both intercellular lipids and defective corneodesmosome proteolysis), but in the case of EHK, these features are combined with a increased fragility, and a tendency to blister. The challenge here is to devise a strategy that both restores barrier function, while stimulating sloughing of outer corneocytes, without promoting further fragility in the underlying epidermis (a common problem with retinoids). Although no currently available treatments meet all of these challenges, a combination of strategies is helpful in EHK (Table, page 6).

Netherton syndrome: Netherton syndrome presents the opposite problem of ichthyosis. Although clinically scaly, there is typically a reduced number of layers of SC, due to unopposed enzyme (serine protease) activity due to the absence of a serine protease inhibitor, LEKTI 1, resulting from mutations in the SPINK 5 gene. Agents that remove scale (e.g., keratolytics such as the alpha-hydroxy acids and oral retinoids) have no role to play in the management of this disorder, and may further aggravate this disease. An atopic-like dermatitis and anaphylactic reactions to foods are another feature of this recessive trait. But immunosuppressants such as tacrolimus are hazardous due to the increased risk for systemic absorption and toxicity across an impaired barrier. The ideal topical therapy, instead, would be a barrier repair formula-

tion, combined with a serine protease inhibitor, a combination that does not yet exist on the market (Table, page 6).

Conclusions

Treatment of the ichthyoses can be frustrating for the clinician, as well as for patients. We have attempted to provide a scheme for a rational (enlightened) therapeutic approach to some of the disorders of cornification, based upon increased understanding of disease pathogenesis. Our suggestions should be modified as new information about disease causes and pathogenesis becomes available, and as new products become available. Clearly, there is a gap currently between what we understand about these conditions and what we can offer patients as treatments. Yet, the approach outlined here can provide a model framework against which therapies can be devised and assessed. While we may not be able to eradicate the problem, we can instead focus efforts on those aspects of the disease that are most bothersome, and through a therapeutic partnership with your physician, it should be possible to help ichthyosis patients to achieve a comfortable, and more functional state.

Reprinted from Clinics in Dermatology, Volume 21, Mary L. Williams, MD, and Peter M. Elias, MD, "Enlightened Therapy of the Disorders of Cornification," pp. 269-273, with permission from Elsevier.

Foundation Resources

Skin Care Products List

The Foundation's skin care products list was recently updated. Our new list covers over 90 company listings representing over 100 products for skin, scalp, eye, and ear care, and sun protection. You, our members, recommended all of the products on our list. Contact the national office for your copy of the updated list. Call 1-800-545-3286 or email info@scalyskin.org. The product listing is available to "Members in Good Standing" only. (See page 2 for a definition of Member in Good Standing.)

Share Your Strength and Creativity

The Foundation is currently strengthening its Fund Raising Committee. We are

looking for volunteers who are willing to roll up their sleeves and get involved. The Fund Raising Committee is responsible for coordinating fund raising plans and helping to identify contacts, programs, and other creative fund raising avenues for the Foundation. If you are interested in joining this exciting and rewarding committee, please contact Jean Pickford at 1-800-545-3286 or email at jpickford@scalyskin.org.

Australia / New Zealand Support List

One of our members from Australia has graciously offered to coordinate a contact list for Australia and New Zealand. To date, we have had only one other person interested in being placed on a contact list, in spite of regular requests from individu-

als "down under" for support and other contacts in their area. The geography, climate, and medical environment in Australia and New Zealand make it difficult for people with ichthyosis to find good information, resources, and other individuals who share the same challenges. A support contact list would be a valuable resource for our members in these countries.

If you would like to be a resource person to someone else in Australia and New Zealand or would just like to share your experiences with someone, please contact Maureen in the Foundation office, 650 N. Cannon Ave., Suite 17, Lansdale, PA 19446, USA, or at info@scalyskin.org.

Therapy for Ichthyoses Based Upon Disease Pathogenesis

Disease (genetic basis)	Pathogenesis	Consequences (severity)	Rational Therapy
Ichthyosis Vulgaris (unknown)	↓Filaggrin → ↓Corneocyte amino acids and their metabolites (e.g., urocanic acid)	Xerosis (moderate) Hyperkeratosis (mild)	Humectants (amino acids, small non-polar metabolites [e.g., urea], and/or glycerol)
Recessive X-Linked Ichthyosis (steroid sulfatase deficiency)	↑Corneodesmosomes due to ↓SCpH→↑SC cohesion and/or protease inhibition	Barrier abnormality (mild) Hyperkeratosis (mild-moderate)	1) Barrier repair-physiologic lipids (cholesterol-dominant) ¹ 2) α-hydroxy acid (? neutralpH-buffered)
Lamellar Ichthyosis (TGI Mutations)	Defective cornified envelope scaffold →Bilayer fragmentation	Barrier abnormality Epidermal hyperplasia Inflammation (mild) Hyperkeratosis (moderate-severe)	1) Barrier repair-physiologic lipids (ceramide ¹ or cholesterol ² dominant) 2) Barrier repair - non-physiologic (e.g., petrolatum, lanolin) 3) Keratolytic (e.g., α-hydroxy acid) 4) Systemic retinoid
Congenital Ichthyosiform Erythroderma (3- and 12-lipoxygenase mutations)	↓lamellar body secretion→ ↓lamellar bilayers and intercellular hydrolases	Barrier abnormality (variable) Erythroderma (variable) Hyperkeratosis (variable)	1) Barrier repair-physiologic lipids (ceramide or cholesterol dominant) 2) Barrier repair - non-physiologic (e.g., petrolatum, lanolin) 3) Mild keratolytic (e.g., α-hydroxy acid) 4) Systemic retinoid
Harlequin Ichthyosis (unknown)	↓lamellar bodies→ ↓lamellar bilayers and intercellular hydrolases	Barrier abnormality (severe) Erythroderma (severe) Hyperkeratosis (moderate-severe)	1) Barrier repair-physiologic lipids (ceramide or cholesterol dominant) 2) Barrier repair - non-physiologic (e.g., petrolatum, lanolin) 3) Keratolytics 4) Systemic retinoids
Epidermal Hyperkeratosis (K1/10 mutations)	Defective keratin cytoskeleton→ Impaired lamellar body secretion and cytosolic fragility	Barrier abnormality (severe) Blisters (variable) Epidermal hyperplasia (severe) Hyperkeratosis (severe) Inflammation Secondary infections	1) Barrier repair-physiologic lipids (ceramide or cholesterol dominant) plus 3) &4) 2) Barrier repair - non-physiologic (e.g., petrolatum, lanolin) plus 3) &4) 3) Antimicrobial - topical and/or systemic 4) Keratolytics and retinoids (with caution)
Netherton Syndrome (SPINK 5 mutations)	↓serine protease inhibitor (SPI) ↑serine protease activity→ ↓SC thickness, bilayer fragmentation	Barrier abnormality (severe) Inflammation (moderate) Erosions Secondary infections	1) Barrier repair-physiologic lipids (ceramide or cholesterol dominant) 2) Barrier repair - non-physiologic (e.g., petrolatum,lanolin) 3) Topical serine protease inhibitor (SPI)

¹e.g., TriCeram cream (Osmotics Corp.); and² Crème de l'Extrême (Osmotics Corp.) are available on the web (Osmotics.com), at Nordstroms, Saks, and at many Long's.

Seniors Speaking

Dear Maureen:

Your Editor's Note in the Fall 2003 issue of Ichthyosis Focus mentions that you need to know if "this column meets your needs" (re: the needs of seniors). I am 84 and have ichthyosis vulgaris. I look forward to each issue of the Focus and hope that you continue the "Seniors Speaking" column.

Yours truly,

C.L. Pittman
Maryville, TN

Dear Friends:

Many thanks to Ichthyosis Focus. I am so glad that I found a site to answer the questions that I have. I have had a lot of unanswered questions for years. I am considered a senior, within the criteria of Seniors Speaking, and would not like to see it go by the wayside.

I am a mere 53 years of age and have a mounting number of questions for anyone out there who will listen. I have

ichthyosis vulgaris and have had it since birth. My question is, "Do others experience dryness in other areas of the body besides the skin, particularly the mucous membranes (i.e., nose, eyes, mouth, and other more "intimate" areas)? I am well aware that at my age some dryness problems are to be expected, but this was a problem when I was as young as 23, and continues to be.

Beth Gaddy
Begadd@aol.com

What's New, What's Hot & What Works

Dear F.I.R.S.T.:

I do not have any members of my family that suffer from this horrible disease, but I was brought to tears Sunday after reading an article in the Herald Democrat in Sherman, TX. It was about a beautiful little three-year-old named Zoe who lives in Bells, TX. Until I read the article I wasn't familiar with ichthyosis. It breaks my heart that we can spend millions of tax dollars sending a probe device to Mars, but we can't come up with a lotion that doesn't hurt, or a cosmetic procedure that can give people like Zoe a quality life, or better than any of that ... a cure for this disease. I really think our world has got its priorities backwards.

I know that you all are probably familiar with every kind of lotion out there, but just in case you haven't heard of *Genes*, which is manufactured by Panco, LTD, I thought I would pass the information on. My mother, who is 87 years old, has cellulitis that causes her legs to swell and burst open. She discovered this cream about a month ago and it is already making a big difference in her skin. She also has lesions on her forehead that she has been applying this to and they are almost gone, some already are. She said it doesn't sting

at all. The phone number to the company is 800-253-3593. We found it at, of all places, Sam's Warehouse. It was demonstrated one weekend and everyone was so impressed with how it felt that they sold 65 cases of it.

I wish there was something I could do, like write letters and get the word out! It's just so sad that we get so caught up in our own little world of "entertain me," that things like this are going on around us and we don't even know it.

Thanks for your time!

Carolyn Craig
Sherman, TX

Editor's Note: Genes is a Vitamin E cream, 100,000 IU's. It is available at Sam's Club or BJ's wholesale warehouses. Customers can purchase three 16-ounce jars directly from the company for \$45.00 (tax included) by calling Panco, LTD. at 1-800-253-3593.

Dear F.I.R.S.T.:

My grandsons, who are 5 and 2, both have ichthyosis. Their scalps, legs, and trunks are the most affected. Recently, my daugh-

ter discovered a lotion, shampoo, and conditioner which have made a tremendous difference in their skin. I just wanted to share the results with you. Perhaps these products will be beneficial to other people with ichthyosis.

The company name is *Ahava*. My daughter uses the Advanced Body Lotion, Anti-Dandruff Shampoo, and Conditioner on both boys. The difference it has made in the condition of their scalps is amazing. I hope others will get the same benefits from these products as we have.

Sincerely,

Peggy Fogle
Phoenix, AZ

Editor's Note:

Ahava products are manufactured in Israel with minerals harvested from the Dead Sea. They can be ordered from www.AHAVAshop.com, or www.ahava-usa.com. Ahava-usa can also be reached by calling 1-877-337-6600, 9:30 a.m. to 5:30 p.m. Eastern Standard Time. Our international friends can order from the AHAVAshop website or write to Ahava, Dizingof 46, Tel Aviv, Israel.

Executive Director's Report



January 2004

Dear Friends and Members of the Foundation,

I hope your holiday season was a joyous one. On behalf of the Board of Directors and staff, I extend best wishes for a very Happy New Year! Our upcoming year promises to be an exciting one for the Foundation, and I look forward to continuing my relationship with you.

Thank you to everyone who regularly contributes to our programs and services, especially through our recent holiday appeal. Our Foundation is growing stronger every day, thanks to our generous and devoted members. The Foundation cannot survive without you.

This November, our Board of Directors met in Philadelphia for a face-to-face weekend retreat. The many issues discussed included; strengthening our board, future plans for fund raising, and the Ichthyosis Registry. Two new members were nominated to the Board of Directors. I am pleased to announce the election of Mark Klafter and Janet McCoy. Mark is the father of a little boy affected with EHK, and Janet is the mother of little girl affected with CIE. In addition to being parents of affected children, both come to the board with a wealth of knowledge in public and media relations, marketing, and non-profit experience. These retreats are always very productive meetings, and this one was no exception. The next board retreat will take place in Kansas City, Missouri, in July, immediately following the family conference.

Speaking of Kansas City, the staff and I are very excited to host the *Fountain of Knowledge* Family Conference in July. We have been working hard soliciting sponsorships, recruiting speakers, organizing schedules, and making hotel arrangements. I have started to receive inquiries about the conference from many of our members. Please see pages 12 and 13 for more detailed information. On a personal note, I truly encourage anyone who has the opportunity to attend this conference to do so. It is by far one of the most informative, enjoyable, and engaging opportunities you will have to learn about and meet others with ichthyosis. You will not be disappointed.

You may already know that raising money for research is a major focus of the Foundation. In 2003, our ultimate goal was to raise \$100,000. Although we were a little shy of our target, I am proud to report that we did receive almost \$50,000 and an additional \$45,000 in commitments. When the commitments are fulfilled, the Foundation will have raised \$95,000! Raising this money took a little longer than projected, but it is still a tremendous accomplishment. Now we must continue on this exciting path and raise another \$100,000 in 2004. Please continue to support our research campaign with your generous donations.

It is time once again, for the annual American Academy of Dermatology (AAD) meeting. This year's meeting will take place in Washington, DC, on February 6 - 11. I will be attending this five-day event, along with 17,000 dermatologists from all over the country and the world. It is an excellent opportunity to inform dermatologists about the Foundation and the services and programs that benefit their patients.

I am also representing the Foundation in the first annual Skin Disease Research Day on Capitol Hill, on February 5. This day is dedicated to advocating for increased federal funding for skin disease research and will be held in conjunction with the AAD's annual meeting. In addition to Skin Disease Research Day, I will also represent the Foundation at the traditional NIAMS Day event in March. NIAMS Day is an awareness day dedicated to informing Congress about the need for increased funding to the National Institutes of Health, particularly the branch for Arthritis, Musculoskeletal, and Skin Disease.

As always, please feel free to contact our staff or me if you have any questions, suggestions, concerns, or just want to talk. We are here to help you and enjoy hearing from our members.

Sincerely,

A handwritten signature in cursive script that reads "Jean".

Jean R. Pickford, Executive Director



Board Members Meet at Weekend Retreat.

In general, the goal in taking care of ichthyosis is to hydrate (moisturize) the skin, hold in the moisture, and keep scale thickness to a minimum.

**Foundation for Ichthyosis & Related Skin Types, <http://www.scalyskin.org>*



Aquaphor® Healing Ointment helps heal dry skin associated with ichthyosis. Its unique petrolatum-based formulation combines a moist environment with the benefits of a semi-occlusive barrier that allows skin to breathe and absorb fluids.

Aquaphor is ideal for daily use because it is hypoallergenic, non-comedogenic, fragrance and preservative-free. Aquaphor Healing Ointment is safe enough for even the most sensitive skin.

*Special Offer
For Readers of this F.I.R.S.T. Newsletter*

MANUFACTURER'S COUPON EXPIRES 06/30/2005

**Save \$2.00
on any
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Product**

(NO TRIAL SIZES)

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Consumer: Coupon good on the purchase of any Aquaphor® product, no trial sizes. Limit one coupon per item purchased. Any other use constitutes fraud. Not good with any other special offer. Consumer must pay sales tax.

Retailer: You are authorized to act as our agent to redeem this coupon and we shall reimburse you at face value plus 8¢ handling in accordance with our redemption policy. Copies available upon request. Offer void if copied and where prohibited, taxed or otherwise restricted. Cash value 1/100¢. Mail to: Beiersdorf Inc, PO Box 880504, El Paso, TX 88588-0504. Good only in USA.



2003 Ichthyosis Awareness Week Highlights

National Ichthyosis Awareness Week (IAW) took place this year October 5 to 11. The Foundation staff and many members helped to educate their local communities about ichthyosis. Once again, this special week was very successful. We have our devoted members to thank for this increased awareness.

Thank you to....

- The 110 members of F.I.R.S.T. who responded to our invitation to offer educational brochures to their family, friends, doctors, co-workers, etc. Over 600 brochures were distributed in many different parts of the country.
- NeoStrata, Inc. and Doak Dermatologics, who sent press releases announcing Ichthyosis Awareness Week to the national newswire service.
- Doak Dermatologics who donated the production and mailing of our new physician's brochure to over 4600 dermatologists nationwide.
- Bobbi DeLuca and her co-workers, who raised \$300 at Prospect Middle School in Pittsburgh, PA.
- Raj Dosanjh, who participated in a marathon and raised \$1600 in donations in honor of her niece who has ichthyosis.
- LouAnna Dugan, who participated in the sale of "Ichthyosis Butterflies" and raised \$200. LouAnna's children, Amelia and Justyn, also spoke to their third grade class about ichthyosis and how it affects their sister Mattingly.
- Lori Florian and Shelby Riggs, who organized a successful yard sale, raising over \$650.
- Daniel Hogan, a physician who was interviewed by the ABC affiliate in Shreveport, LA.
- Greg LiCalzi, who participated in the sale of "Ichthyosis Butterflies" at Union

College and raised \$200.

- Marc, Denise, and Brian Merritt, who made chocolate lollipops and sold them at various businesses in their community. The Merritts raised over \$2600. Marc's employer matched \$1000 of this tremendous accomplishment.



Shelby Riggs and her cousin Brianna selling lemonade at the yard sale.



Denise, Marc and Brian smile proudly.

- Nancy Osentoski, who participated in the sale of "Ichthyosis Butterflies" and was featured in a newspaper article in the Detroit area.
- Lisa Short, who sold "Ichthyosis Butterflies" in her office cafeteria and raised \$250.
- Michelle Weckwerth, who organized a bake sale at her place of employment and raised \$134.

- Paula Wetterlund and family, who sent donations to the Foundation in memory of Paula's mother. Memorial donations totaled over \$300.

In addition to the efforts of our members, the Foundation staff emailed, faxed, and mailed our press release to over 70 major newspapers nationwide.

A hint of our success during IAW was the major increase in website activity in the week immediately following. While scalyskin.org averages about 1600 hits per week, during the week of October 12 to 20 our website received over 4400 hits. Congratulations to all on a job well done!

Raising funds for the Foundation does not have to take place just during Ichthyosis Awareness Week. Raising money to advance the mission and programs of the Foundation can happen any time during the year. For instance, one family is planning a benefit auction in their neighborhood for this spring. Our staff is always ready to provide guidance or help. *Please note*, if you are planning some kind of event to raise funds on behalf of the Foundation or you plan to use the Foundation's name, you are required to contact the national office prior to the event. The Foundation staff must be aware of activities taking place around the country that could affect its established reputation.

Thank you to everyone who works to advance the Foundation's mission and increase awareness of ichthyosis in his or her communities.



Camp Discovery Continues A Tradition of Fun in Caring Surroundings

Imagine missing the thrill of summer camp, the warm glow of a campfire, the serene beauty of a canoe ride, or the excitement of making new friends, because you have a skin condition that prevents you from participating in outdoor activities. The American Academy of Dermatology (AAD) couldn't imagine denying kids this unforgettable opportunity either, which is why this national dermatology association founded Camp Discovery.

Since its inception in 1993, approximately 200 children with serious dermatologic conditions have participated in the AAD's Camp Discovery each year. While summer camp often seems like an impossible dream for these children due to their medical needs, Camp Discovery gives them the opportunity to enhance their self-confidence, learn from other children who have similar dermatologic conditions, and enjoy a week's worth of fishing, boating, swimming, water skiing, and arts and crafts. Dermatologists and nurses volunteer their services to provide medical supervision for the campers.

"Camp Discovery is a perfect example of how volunteerism and a vision can enrich so

many lives," said dermatologist Raymond L. Cornelison, Jr., MD, President of the American Academy of Dermatology. "These children are so grateful for the opportunity to spend time at camp and meet other kids with similar life experiences. It gives them confidence at a time in their lives when looking different from other kids can be difficult."

Children ages 10 through 13 are invited to attend Camp Discovery at Crosslake, Minnesota, from July 10 to July 16, 2004. Teen Camp Discovery will also be held at this location for teenagers ages 14 through 16, from July 17 to July 21, 2004. An additional Camp Discovery, in Millville, Pennsylvania, will be held from August 15 to August 21, 2004, for children ages 8 through 13. Campers are assigned locations depending on age and space availability.

Camp Discovery is completely free to campers. All campers receive full tuition and transportation scholarships to attend Camp Discovery through the generous contributions of AAD members, corporations, individuals, and organizations.

Recently, the AAD established an endow-

ment fund for Camp Discovery to guarantee the continuation of this important program. In December it met its goal of \$2.5 million. The Academy has also established an endowment for Teen Camp Discovery to ensure that during the often challenging transition from teenager to adult, Camp Discovery remains available for teenagers with serious dermatologic conditions.

Applications for this year's Camp Discovery are currently being accepted and are due to the Academy by Friday, April 16, 2004. The 2004 application can be found on the Academy's website, www.aad.org. Potential campers must have a letter of recommendation from their dermatologist. Members of the Academy are encouraged to recommend candidates for Camp Discovery. For more information about Camp Discovery, or to make a contribution to Camp Discovery, call the AAD at 847-330-0230 or visit the Academy's web site at www.aad.org.

Excerpted from the American Academy of Dermatology press release, December 12, 2003.

Fight Scaly Skin with

Epilyt[®] Lotion Concentrate



- ◆ *Loosens and removes scales**
- ◆ *Softens and smooths rough, dry, scaly skin**
- ◆ *Non-greasy, clear, moisturizing lotion*
- ◆ *Controls severely dry skin*

Please bring this ad to your Pharmacist who will order Epilyt[®] (NDC 01-45-0624-04), for you through one of the following Pharmaceutical wholesalers.

Distributor	Item Number
AmeriSource	3433877
Bergen Brunswig	520-601
Cardinal	1412212
McKesson	1102151



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Epilyt[®] is also available for purchase online at www.webderm.com

Please visit us at www.stiefel.com

Paid Advertisement

Conference Chatter

2004 National Family Conference: A Fountain of Knowledge

July 2 – 4, 2004

The Westin Crown Center, Kansas City, Missouri

Registration – See pages 13 and 14 for the official registration form. The registration form can also be completed online at www.scalyskin.org.

Clinical Screening – Experienced, knowledgeable dermatologists will examine conference attendees with ichthyosis. If you are interested in a consultation with one of the doctors, be sure to complete Section III of the registration form so that an appointment can be made for you. There is no charge for the clinical screening.

Conference Program – The conference program will feature four tracks of programs to accommodate each age group. Age-appropriate material will be included in each track. There will be a strict policy that no exceptions will be made to any age group.

Track 1 (Adults, ages 18+)

- What's new in research?
- Breakout discussions on specific disease types and treatments.
- Breakout discussions for women, men, mothers, and fathers.
- Sibling talk (for parents) deals with concerns often experienced by brothers and sisters, such as peer issues, resentment, and concerns about the future.
- Specialist panel will talk about eyes, ears, and nutritional issues.
- Breakout discussions to share information and helpful hints.
- Breakout session addressing depression in children.
- The ABCs of advocacy.
- School, workplace, and insurance issues.

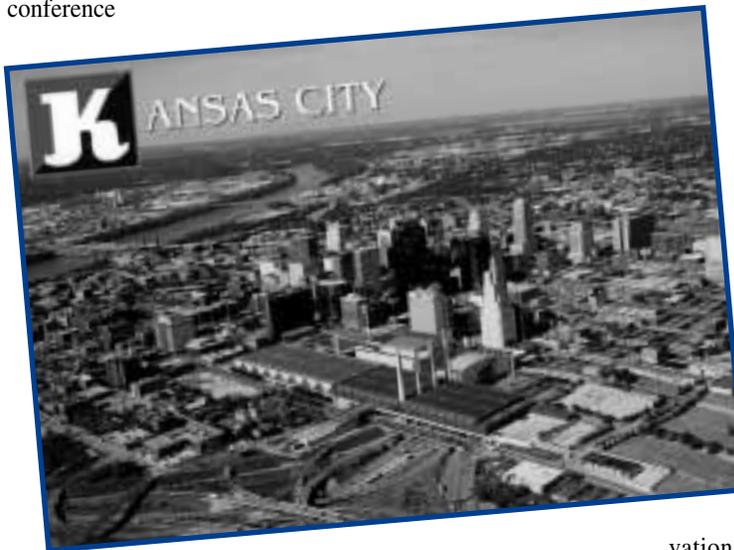
Track 2 (Teenagers, ages 13-18) & Track 3 (Pre-teens, ages 9-12).

- Self-defense techniques.

- Making healthy choices in nutrition (teens only).
- Stress relief techniques.
- Affected teens and pre-teens talk (moderated by a clinical psychologist).
- *Sibshop* for unaffected siblings (moderated by a specialist in sibling issues).
- Career panel discussion, preparing for the future (teens only).
- Creative expression using art (pre-teens only).
- Exercise, stretching, and health.
- Teen and Pre-teen Jeopardy!

restrictions apply. To obtain this discount, call US Airways' Group and Meeting Reservation Office toll-free at 1-877-874-7687, 8:00 am – 9:30 pm EST. Refer to Gold File Number 64162913.

Delta Airlines – Points of origin include US, Bermuda, Caribbean, Mexico, and Canada. Receive 5% off US Domestic published fare or 10% off published YO6/YR06 fare. Certain dates, rules, and restrictions apply. To obtain this discount, call Delta Airlines at 1-800-241-6760 and refer to Meeting Identifier Code DMN200484A.



National Patient Travel Center - You may be eligible for free transportation using the Angel Flight America Program. If you live within 1000 miles of Kansas City, Missouri, Angel Flight will fly a family in a 4-seater or 6-seater plane to and from the conference at no cost. Contact the National Patient Travel Center at 1-888-675-1405 and refer to the 2004 Foundation for Ichthyosis Family Conference Special Lift Program.

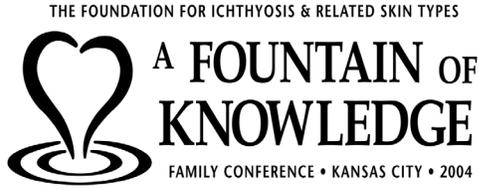
Track 4 (Children, ages 1-8)

- Program specifically designed for young children. Games, toys, art supplies, books, etc., will encourage children to interact. This track will be supervised by the licensed, trained, and caring professionals of A1 Tiny Tots, a professional childcare company.

Discounted Travel to Kansas City, Missouri -

US Airways – Receive 5% off lowest applicable published fares or 10% off with 60 days advance reservations/ticketing required from all points on US Airways' route system. Certain dates, rules, and

Accommodations – Attendees must make their own room reservations at the Westin Crown Center. Westin's discounted room rate is \$89.00 per room, per night, flat occupancy + applicable taxes. Call 1-888-627-8538 to reserve your room at the Westin Crown Center. The Reservations Center is open 24 hours a day. Be sure to name the "Foundation for Ichthyosis" to receive the discounted rate in the room block. If you are interested in extending your stay in Kansas City, this rate is available from June 28 through July 7, 2004. However, you must make your reservation by June 4, 2004 to guarantee this discounted rate. After June 4, 2004, the rooms will be available on a first-come, first-served basis.



**Westin Crown Center Hotel
Kansas City, Missouri
Friday, July 2 – Sunday, July 4, 2004**

Section I – Contact Information

Name: _____

Address: _____

City: _____ State: _____ Zip: _____

Phone: _____ Email: _____

Section II – Registrant's Information

This section must be completed with registrant's complete name, age, and appropriate track for each attendee. Please print clearly; nametags will be provided.

<u>Name</u>	<u>Age</u>	<u>Type of Ichthyosis</u>	<u>Adult</u> 18+	<u>Teen</u> 14-17	<u>PreTeen</u> 10-13	<u>Child</u> 1-9
_____	_____	_____	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
_____	_____	_____	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
_____	_____	_____	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
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_____	_____	_____	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
_____	_____	_____	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Section III – Clinical Screening Appointments

This is an opportunity to be seen by knowledgeable dermatologists to answer any questions or concerns that you may have. These appointments will take place throughout the day on Saturday, July 3. Your appointment will be assigned and posted at the registration table.

<u>Name of person for appointment</u>	<u>Age</u>	<u>Male/ Female</u>	<u>Type of Ichthyosis</u>	<u>Confirmed by a dermatologist?</u>	
_____	_____	_____	_____	Yes	No
_____	_____	_____	_____	Yes	No

Section IV – Other

- | | Yes | No |
|---|--------------------------|--------------------------|
| • Do we have your permission to take photos during the conference? | <input type="checkbox"/> | <input type="checkbox"/> |
| • Do we have your permission to place photos of you and your family on our website? | <input type="checkbox"/> | <input type="checkbox"/> |
| • Do we have your permission to include your contact information on a conference roster to be distributed to all registrants? | <input type="checkbox"/> | <input type="checkbox"/> |
| • Have you ever attended a conference before? | <input type="checkbox"/> | <input type="checkbox"/> |
| • Can you volunteer some time at the conference to help? | <input type="checkbox"/> | <input type="checkbox"/> |

Section V – Payment Information

The Foundation has received several sponsorships, which allows us to offer the discounted registration fees of \$95 per adult and \$50 per child. The actual cost per person at the conference is \$150.00. Conference fees help offset the cost of food, beverages, room rental, supplies, etc.

Number of adults attending? (18 or over) _____ x \$95.00 = \$ _____

Number of children attending? (17 or under) _____ x \$50.00 = \$ _____

Additional Donation (optional) _____ \$ _____

Grand Total \$ _____

My check is enclosed, made payable to FIRST.

Please charge my credit card: MasterCard, American Express, Visa (circle one)

Credit Card Number _____

Expiration _____ Authorized Signature _____

Miscellaneous Information

- ✓ The conference will begin on Friday, July 2, at 2:00 pm CST and end on Sunday, July 4, at 12:00 pm CST.
- ✓ Breakfast, lunch, and dinner will be provided on Saturday, July 3, and breakfast will be provided on Sunday, July 4.
- ✓ On Friday evening, July 2, there will be free time to visit Kansas City. Transportation will be available from the hotel to deliver guests to the famed Country Club Plaza, home to a wide variety of shops, boutiques, restaurants, and theaters.
- ✓ Conference attendees are responsible for making their own room reservations at the Westin Crown Center. Call toll-free 1-888-627-8538, 24 hours a day.
- ✓ For those who are driving, there is a parking fee of \$12.50 per night, with a \$10.00 weekend max.
- ✓ For driving directions to the hotel, use www.mapquest.com or <http://maps.yahoo.com/>. The hotel address is 1 Pershing Road, Kansas City, MO 64108.
- ✓ KCI Shuttle provides transportation from the airport to the hotel. The cost for a one-way transport is \$14.00 or \$23.00 round trip. Simply report to the nearest KCI Shuttle ticket counter adjacent to most airline baggage claim areas.
- ✓ Mail this form to FIRST, 650 N. Cannon Ave., Suite 17, Lansdale, PA 19446. Registration closes June 11, 2004.

National Ichthyosis Registry

“The times, they are a changing ...”

Hello to the FIRST membership. The National Registry for Ichthyosis has come to a turning point. Our funding through the NIH expires late this year, and we will only be able to continue to enroll new people in the Registry through the end of March 2004.

What does this mean to FIRST, to those who are enrolled, or to you if you want to enroll? It means we have made a great start! The purpose of the Registry is to improve understanding of the diagnosis, causes, and treatment of the ichthyoses. We do this by gathering information from enrolled individuals and entering that information into a database. The information allows us to characterize the enrollee and to determine if he or she fits criteria for the diagnosis of a particular disorder. In some cases, biopsy slides are reviewed and DNA may be analyzed for mutations in genes underlying some of the ichthyoses. This information results in a large group of well characterized individuals. The information gathered can be useful

for future studies. In most cases, enrollees have asked to be informed of appropriate future studies.

The first phase of the Registry, identifying affected individuals and gathering the information, will end this year. The analysis of DNA samples is ongoing, as well as completion of the file review and data entry in the database. Over 600 people have enrolled. This is a GREAT beginning! We still look to investigators to utilize this resource and will lobby hard to make it available after open enrollment in the Registry is closed.

Our current plan for information in the database is to continue to make it available to researchers. We also plan to maintain the bank of DNA samples for future investigations. We are exploring ways to support these services.

The Foundation for Ichthyosis has been an excellent and always supportive partner with the Registry, and we praise and thank the staff and the membership for the unflagging enthusiasm you have all shown us through the years.

Personally, Dr. Fleckman, Kim Pineda, and I would all like to say a heartfelt thank you. You have helped make the Registry a reality. As anyone who has spoken with us has heard us say, we don't know specifically how we may benefit you, but we can't ever figure out ways to help with the ichthyoses until we know more, and that's what the Registry has been set up to help accomplish.

As more information is available regarding future Registry plans, we will share it with you through the Ichthyosis Focus. Additionally, we still encourage you who have not enrolled to do so while you still can, and for anyone who isn't sure please contact us and we will find out together where you stand.

Best wishes to us all in 2004! Remember, “Well begun is half done.”

Sincerely,

Geoff Hamill, RN
Registry Coordinator

Camp Wonder

By Christine Tenconi

Children! Come to a place where there are no secrets, a place where you can play with other kids who experience similar daily challenges, and where you do not have to hide your feelings about how you look. Camp Wonder, a medically staffed summer camp for children with skin diseases ages 5 to 16, is now accepting applications for Summer 2004.

Camp Wonder is held at Camp Arroyo, a facility built for children with special needs. It is situated on 138 acres adjacent to the Del Valle Reservoir in Livermore, California. Camp Wonder is a very special place where children with chronic diseases have the opportunity to build self-esteem, camaraderie, and long-lasting friendships while participating in activities such as swimming, fishing, boating, horseback riding, dances, talent shows, and much more.

Camp Wonder is free to all campers. Two one-week sessions are scheduled from June 20 to 25 and June 27 to July 3, 2004. For more information or to request an application please call 925-947-3825, or email ctenconi@hotmail.com.

News & Notes

continued from page 3

Current research indicates PC is caused by mutations in keratin genes, specifically K6a, K6b, K16, and K17. PC usually follows an autosomal dominant mode of inheritance.

For more information visit the PC Project website at www.pachyonychia.org, or write to PC Project at 2386 East Heritage Way, Suite B, Salt Lake City, UT 84109.

Tax Credit for Families Attending Medical Conferences

The Internal Revenue Service (IRS) issued a ruling on May 8, 2000, that allows parents of children with chronic illnesses to deduct some of the costs associated with attending medical conferences related to their children's condition. The rule allows parents to deduct the expenses of admission and transportation to a medical conference related to the chronic illness of the individual's dependent. It is explained in the Internal Revenue Bulletin 2000-19, which is located on the IRS website at <http://www.irs.gov/bus/info/bullet.html>.

IRS Publication 502 details allowable medical and dental expenses. Go online to www.irs.gov to print this publication or call your local IRS office and request a copy.

Drugs@FDA

The U.S. Food and Drug Administration (FDA) plans to roll out a new drug information Web site for consumers soon. It will be called **Drugs@FDA**, and consumers may visit an online pilot version of it now. Simply enter **Drugs@FDA** in the search box on the FDA home page (www.fda.gov).

The new site is part of FDA Commissioner Mark McClellan's consumer education initiative. It will include information from the FDA's drug information sheets, which are summarized versions of product labeling that the FDA currently posts on the Web site of its Center for Drug Evaluation & Research (CDER).

Consumers will be able to search by drug name and find out such things as whether a generic alternative of a product is available. The FDA intends for the site to become a clearinghouse for drug-related regulatory histories, product labels, and manufacturing information. The FDA is seeking consumer feedback through the online pilot version.

Household Products Database

The National Institutes of Health (NIH) recently unveiled an online consumer's guide

providing information on the potential health effects of more than 2,000 ingredients contained in more than 4,000 common household products. Known as the National Library of Medicine's (NLM) Household Products Database, it may be accessed at <http://householdproducts.nlm.nih.gov>.

Information in the database is derived from publicly available sources, including product labels and information provided by the manufacturers. The products in the database include those most commonly found under the kitchen sink, in the garage, in the bathroom, and on the laundry room shelf. They are grouped by category, such as "Auto Products," "Hobbies and Crafts," and "Landscape/Yard." The database is designed to answer such questions as:

What chemicals are contained in specific brands and in what percentage?

Which products contain specified chemicals?

Who manufactures a specific brand?

How can I contact the manufacturer?

What are the potential health effects of the chemical ingredients in a specific brand?

Reprinted from Orphan Disease Update, Winter 2003/2004, Volume XXII, Edition 1, a publication of the National Organization for Rare Disorders (NORD).

Have you moved recently or are planning on moving soon? Please let us know your change of address so you can continue to receive the Focus. Postage for the newsletter does not include forwarding.



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