



ICHTHYOSIS FOCUS

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Published by The Foundation for Ichthyosis and Related Skin Types, Inc., (F.I.R.S.T.), PO Box 20921, Raleigh, North Carolina 27619-0921 (formerly The National Ichthyosis Foundation). Telephone 919-782-5728. F.I.R.S.T. is a non-profit corporation for the benefit and education of its members and the public regarding medical, psychological and social aspects of the ichthyosis, a family of primarily genetic skin disorders marked by excessive scaling. Letters, suggestions and contributions are welcomed.

Helpful drugs may be restricted

During the Williamsburg conference held this past July, Eileen Enny Leach, R.N., a speaker for the day, informed our members that the Food and Drug Administration (FDA) may seek to restrict the use of certain prescription drugs (i.e. Retin A, Accutane, etc...) which have been very helpful to ichthyosis patients.

The FDA is considering these restrictions due to misuse by certain individuals. These prescription drugs would be limited to us because they were not developed specifically for ichthyosis.

We urge you to write to the following addresses using the format below. If we do not act quickly, we may all suffer the consequences. Thank you so much for your prompt assistance in this area.

Write to BOTH addresses: Dr. David Kessler, MD, Commissioner of the Food and Drug Administration, 5600 Fishers Lane Room 1467, Rockville, MD 20857, phone: 1-301-443-1544.

And to Dr. Louis Sullivan, Secretary of Health and Human Services, 200 Independence Ave. SW,

Washington, DC, 20201, phone: 1-202-619-0257.

Dear Dr.

I am an Ichthyosis (patient, parent of a child/ adult) and have recently been informed that the Food and Drug Administration will possibly be restricting the use of drugs such as Retin A and Accutane, as well as others. Patients who suffer from Ichthyosis benefit greatly from the use of these prescriptions.

I am requesting that the Food and Drug Administration grant Ichthyosis patients an exemption from this possible restriction since this is an orphan disease. Patients who suffer with Ichthyosis have no other means of medical treatment available to them at this time. The use of these prescriptions increases the quality of life for many patients, and without the use of these drugs, these same patients will suffer. Aside from the physical discomforts, many patients would lose time on the job,



Eileen Enny Leach, B.S., R.N.

resulting in a loss of income.

I fully understand the position the Food and Drug Administration has concerning misuse of prescription drugs, but this generalization to an Ichthyosis patient would be a grave one. I thank you for your careful consideration into this situation and anxiously await your reply.

Sincerely,
(sign your name)

During the conference Eileen Leach reviewed the history of drug development and discussed how the current FDA came into being.

It's all in the genes

One of the highlights of the Williamsburg conference was Dr. Virginia Sybert's lecture entitled "Your Genetic Tune-up. An Overview of Ichthyosis Disorders and how they are inherited." Dr. Sybert is a dermatologist, a pediatrician and a geneticist in Seattle, WA. She told the group

attending the conference that physicians often learn more from their patients than the patients learn from the doctors.

"The single most important fact to remember about ichthyosis is that it is a genetic disorder and it runs in families. We are dealing not just

with the here and now, but with the future and with recurrence risks," she said.

She explained that physicians have an agenda when they first meet with new parents who have just given birth to a baby with ichthyosis. "Our . . . Continued on next page



Frances Ott, left, and Ellen Rowe, right, present Dr. Virginia Sybert with a plaque expressing our appreciation for her support of F.I.R.S.T. and help with the summer conference.

agenda includes making the correct diagnosis, telling you about the natural history of the disorder, explaining recurrence risks and causation, discussing prenatal testing, and offering referral for care. These are all issues for us as doctors. Your agenda is "What is it? And can you fix it?"

Dr. Sybert said that most parents just want to know if their baby is going to die. "When we tell you no, your baby is not going to die, you say, okay, then fix it. When we tell you we can't, that's all you hear." Dr. Sybert explained that people have needs at

different times for different information.

"So don't expect too much from your doctor. Don't expect too much from yourself. You can only absorb a little information at a time."

She encouraged people to tell their physicians what they are worrying about, even if it seems silly. "Ask questions. Be open with your health care workers. It's a communication process."

Dr. Sybert emphasized the importance of getting a correct diagnosis. "You need to know if you have a dominant or recessive trait."

Although certain types of ichthyosis, such as CIE and Lamellar, have been categorized separately in the past, Dr. Sybert feels that these autosomal recessive diseases are really heterogeneous and that the labels can be used interchangeably.

"Differences in mutations may account for differences in patients. We will learn more as we learn to distinguish among them and learn more about the differences. There actually may be nine or ten different disorders in this group. I just don't get hung up

on the differences."

Dr. Sybert showed slides of various forms of ichthyosis and explained how similar types genetically can look very different visually. "But because skin is abnormal, doesn't mean the inside is abnormal," she stated. She described a five year old patient who is a bright, interactive, social kindergartner who excels in spite of her severe skin disease.

"Children who are affected will never experience the pain you feel as a parent, never. As parents, we feel helpless. But if you have ichthyosis, at some level you come to grips with yourself. You say, this is who I am. This is how I am. This is my life and I'm going to make something of it."

Dr. Sybert ended the day with a panel discussion with Dr. William Rizzo and Eileen Enny Leach, R.N. Excerpts from this panel will be printed in future issues of FOCUS.

Many thanks to volunteers

The success of the summer conference was due to the team effort of a group of dedicated volunteers. The event began with a pool party the night before the conference. A terrific thunderstorm encouraged us to move indoors where we were greeted by streamers and Hawaiian leis enthusiastically distributed by some of our children. Families had a chance to get to know each other during this informal gathering and to catch up with old friends from past conferences.

The next morning registration began bright and early. More than 100 people attended this year's conference. The children had a wonderful day of magic shows, juggling acts, colonial music and visits from members of Williamsburg's living history program. These events were sponsored by generous contributions from **Herald Pharmacal, Glaxo Dermatological, Sween Corp., Stiefel Laboratories, Neutrogena Dermatologics, and Owen/Galderma Laboratories, Inc.**

F.I.R.S.T. would like to thank everyone who helped make the conference such a huge success. Among

those active volunteers were: **Cynnle Bates, Frances McHugh, Lynne Alba, Georgie Betts, Ellen Rowe, Dave Baucom, Reba Benson, Nancy Ott, Randy La Barbera, and Jean O'Brien.**

There's always work to be done at the F.I.R.S.T. office. Several volunteer opportunities are available if you are interested. We need someone to translate F.I.R.S.T. booklets and other literature into Spanish and other foreign languages for use by people for whom English is a second language. We have already received a request for this from the state of California.

If you would like to distribute F.I.R.S.T. literature to your local dermatologists, hospital, health department, and library, contact us and we will send you brochures to hand out. Also, don't forget to call your regional representative and volunteer to be part of the support network.



Cynnle Bates, Reba Benson, Frances McHugh and Dave Baucom were among the many volunteers who made the Williamsburg, VA conference a big success.

And remember that your contributions make this newsletter possible. If you have enjoyed reading this issue, send a donation today. F.I.R.S.T. is **YOUR** foundation. Write to us. Call us. Let us know what you would like to see and read about in FOCUS.

If you were unable to attend the conference, you might want to order a copy of the video. Send a check for \$25 to: **Davenport Photography/Video, 4453 Powhatan Crossing, Williamsburg, VA 23188.**

Correspondence Corner

Lori and Mark Rogers are the proud parents of a new baby boy, Ben, born July 11th. Ben has ichthyosis. Lori and Mark would like to hear from other parents who have children with ichthyosis. Their address is 11712 S. Williamsburg Dr., Knoxville, TN, 37922. You can also call them at (615) 966-0936.

Janna and Steven Lloyd of Elverta, California also have a new baby. Their daughter, Kelsie, was born with ichthyosis on July 2. Janna says, "We are getting a lot of 'I don't knows' from the doctors. We have put out feelers all over trying to locate families in similar circumstances... We love our daughter so much and want to give her the best that we can. She is a beautiful little girl."

The Lloyds address is 8218 16th St., Elverta, CA 95626. Phone: (916) 991-5749.

Jacqueline Grless, age 13, would like a pen pal. She would like to correspond with other children who have ichthyosis. Jacqueline's address is 12th St., Wymore, NE, 68466.

Tatiana Hamor, age 11, would also like a pen pal. Tatiana lives in Spain, and says she enjoys playing the piano, saxophone and Nintendo! Tatiana has EH. Her address is PSC 60 Box 4515, APO AE 09641.

Cynnle Bates, one of our board members, would like to pass along the name of a company which sells dermatology products at discount prices through the mail. The firm is called Carriage Shed, and the address is 211 N. Atlanta Ave., Sheffield, AL 35660. The phone number is (205) 386-0797. Write or call for a list of products and prices.

Dennis Kaden, 1611 Caudor St., Leucadia, CA 92024, (619) 942-9228, tells us that "recently I have found some healthy skin care products that have made a positive impact on my life." He is willing to share this information with anyone who would like to get in touch with him.

Nancy Ott, 503 Pitt St, Mt. Pleasant, SC 29464 wants to pass along the name of a book which describes the different types of ichthyosis. The title is "The Ichthyoses, A Guide to Clinical Diagnosis, Genetic Counseling and

Therapy" by H. Traupe. Nancy says this book is available in the medical library at most teaching hospitals. It can be purchased from Springer Publishers, New York for \$129.00.

Betty McMasters, 1838 So. Muskogee, Sapulpa, OK, 74066, writes to say that she enjoys getting newsletters and often goes back to read her old ones. "When I read the many articles in these old newsletters, tears come to my eyes. I feel like these newsletters tie us all together as one big family who share a common factor." Betty's daughter, Katy, age 12, has ichthyosis.

Delora Bell, the mother of 9 year old Jacqueline, would like to learn more about how other parents cope with their children's ichthyosis. The Bells are a military family and have not met anyone else with this disease. Their daughter has been having severe ear problems requiring surgery. If anyone can help, please call them at (513) 252-0503 or write to them at 5022 Cobb Drive, Dayton, OH 45431.

Cindy Cook, age 30, called to let us know that she had severe reactions to antidepressant medications. She experienced intense itching and swelling from four different types of antidepressants. Cindy has EH. F.I.R.S.T. would like to hear from others who might have had similar reactions. Antidepressants are generally very helpful and have been taken successfully by other folks with ichthyosis. Cindy's phone number is (312) 975-7815 if anyone wants more information. She lives in Chicago, IL.

Peter Cole has a two year old child with ichthyosis. He would like to hear from other families. The Cole's address is 140 Gloaming Hill, Titahi Bay, Porirua, New Zealand, 04368930. Please write.

Robin Joyce, Route 1, Box 189-F, Sandy Ridge, NC 27046, has a six month old baby boy named Ethan. He was born with EH. "I feel that my family and I are so alone. My emotions are constantly on a roller coaster. I want to know how people cope with EH? I want to give Ethan the best life ever. I want him to always be proud and hold his head high," says Robin. She has many questions about diapering, clothing, etc... and would like

to communicate with other mothers who have babies or toddlers. Her telephone number is (919) 871-3277. Some adults with EH might also give her a call and offer support.

Janet Greusel, Box 184 G Plum Road, Mohrsville, PA 19541, says that she really enjoyed the summer conference in Williamsburg. "The only problem was the day passed too quickly." She wanted to pass along a tip to others who suffer from fungus infections on their feet. Janet found relief from a prescription drug called Naftin. Ask your dermatologist about it if you have this problem.

Mary Toth, 76 years young, recently wrote to say that she has had a rough summer with her skin. "It hasn't been this bad since I was a youngster. When I go out, and my hands are peeling, I feel like hiding them." Mary would like to hear from some other senior citizens. Her address is 137 Knowlton St., Stratford, CT, 06497. Mary attended the conference in New Haven several years ago.

Stephen Chin-Bow, 211 East 53rd St., Apt. 9H, New York, NY 10022, wants to start a support group in New York City for people with ichthyosis (all types) or their relatives and friends. The meetings will be held somewhere in Manhattan. Write to him for details and information. Support groups can offer encouragement and hope to those who feel alone with this disease. If you live within driving distance of New York City, this is a wonderful opportunity to meet others like yourself.

An anonymous tip was put in the HOT TIPS box at the summer conference. This person passed along that her 4-year-old daughter has good results from using Mary Kay's extra emollient nite cream on her face. She has CIE.

If you would like a pen pal, have a comment to make, or would like to be in touch with others who have ichthyosis, write to: Ichthyosis Focus, P.O. Box 20921, Raleigh, NC 27619, and we will include your name and address in Correspondence Corner. We do not release names and addresses without permission.

Tell Me Doctor

by Melodie Buxman, M.D.

Q. My aunt has just been diagnosed as having Darier's disease. Is this a form of ichthyosis and what are the chances I or my children will be affected with this condition (I am 30 years old)?

A. Darier's disease is often classified as a type of ichthyosis because it is an inherited abnormality of skin formation which results in thickened (hyperkeratotic) skin. Although it is inherited as a "dominant" (directly from parent to child) disease, there are many who do not give a positive family history. Thus, there must be those who carry the trait without expressing any skin abnormality. It generally begins in adolescence, although a few adult onset cases have been reported.

Areas of skin affected by Darier's are rough, thickened and brown, and may have a "cobblestone" appearance. In these areas the skin cells are actually falling apart and the skin tries to heal itself by heaping up over the damaged cells.

The palms and soles may be very thick as well, and the nails may show red and white streaks and break easily. Some of the same complications experienced by ichthyosis patients may affect Darier's as well, such as frequent skin infections, cracking, fissuring, and drying.

In addition, sun exposure may trigger development of visible Darier's disease in otherwise normal appearing skin. Thus, flares of the condition occur regularly in summer. Some of the same treatments useful for ichthyosis, such as antibiotics, soaks, emollients and Retinoids (synthetic vitamin A treatments) may help if used with supervision by a doctor.

Q. What is Netherton's syndrome? I understand it is a rare

form of ichthyosis.

A. You are correct. There are three generally accepted features of Netherton's syndrome (incidentally, the word "syndrome" has an interesting derivation — it comes from "syn" the Greek word for "together" and "dromein" meaning "to run").

In Netherton's syndrome three main features tend to "run together," namely, hair shaft abnormalities, such as "bamboo hairs" where the shaft seems to telescope on itself producing a "ball in socket" appearance; eczema, or dry red itchy patches of skin; and ichthyosis, which may be widespread, scaling and reddish, or present as waves of double-edged scaling ridges over the skin.

Males are apparently more frequently affected than females, and signs of the disorder are usually present at or near time of birth. Netherton's syndrome may improve with oral synthetic vitamin A treatment.

Melodie M. Buxman, M.D. is a dermatologist on the F.I.R.S.T. Medical Advisory Board. She will be happy to answer your questions. Please send your questions to the F.I.R.S.T. office and we will forward them to her.

Matching gifts help F.I.R.S.T.

You may be affiliated with a company that will match your generous gift to F.I.R.S.T. Your company's matching gift program may double or even triple your donation dollars. The following list includes some of the many companies that currently have matching gift programs. If you are employed by one of these companies, please obtain the proper matching form from your personnel or public affairs office and enclose the form with your gift. We will be glad to complete any information needed for the matching funds.

Your contribution will then

have an even greater impact on spreading the word about ichthyosis.

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The Washington Post Company
Westinghouse Electric Fund
The Xerox Foundation

Regional Support Network

An exciting project is underway at the F.I.R.S.T. office. Board member Cynn timer Bates of Lexington, KY has established a system of regional representatives who will serve as contact persons for eight different areas of the United States.

These representatives met via telephonic conference call on Sept. 22 to discuss the goals and objectives of the Regional Support Network. They are working on defining specific responsibilities of the regional representatives, and you will be hearing from them in the coming months.

The purpose of the support network is to facilitate communication between families and people affected by ichthyosis. Parents can exchange practical tips and child care information. Adults can support each other in job hunting, making new friends, dealing with depression, and other social coping strategies.

Years ago F.I.R.S.T. tried to develop regional representatives for each state, but we did not have a system for putting these reps in touch with you. We are working on this project and hope to organize some regional get-togethers next year, rather than one large national conference.

If you are interested in becoming a support person for others in your area, please contact the representative for your region. When new babies are born, it is comforting to have other families near by visit or call the new parents. Also, you may just want to find out if someone else with your type of ichthyosis lives close to you so that you can exchange information.

If you were previously a regional rep and would like to volunteer again for your state, please contact the person who is coordinating your area.

Region One: NY, PA, VT, NJ, CT, RI, MA, ME

Harry and Lynne Alba
917 Pierce Road
East Norristown, PA 19403
(215) 584-6366

Region Two: DE, MD, WV, VA, NC, SC

Trice Ovbey
803 Westmont Drive
Fayetteville, NC 28305
(919) 433-2777

Region Three: TN, GA, FL, AL, MS, LA, AR

Cathy Sipper
P.O. Box 127
Rutledge, AL 36071
(205) 335-6827

Region Four: KY, OH, IN, IL, MO, MI

Cynn timer Bates
1651 Tazwell Drive
Lexington, KY 40504
(606) 254-9080

Region Five: WI, IA, MN, ND, SD, NB

Marlene Huffman
1326 DeSoto Avenue
Glencoe, MN 55336
(612) 864-4508

Region Six: TX, OK, NM, CO, KS

Marge Boyd
7427 East 31st Place
Tulsa, OK 74145
(918) 622-9480

Region Seven: WA, OR, ID, MT, WY

Heather Gattuccio
3604 N.E. 50th
Portland, OR
(503) 284-8946

Region Eight: CA, NV, UT, AZ

Mark and Claudia MacNaughtan
610 East 800 N.
Genola, UT 84655
(801) 754-3064

United Way campaigns begin

United Way annual fall campaigns begin in most states in September. We want to remind you that F.I.R.S.T. is a 501(c) (3) nonprofit organization authorized by the IRS to accept tax deductible contributions, and therefore meets the requirements of United Way of America and its member organizations.

Each United Way, however, has requirements specific to its local community, but many honor donor designated programs and special interest organizations. F.I.R.S.T. currently receives donations designated by our members and their families and friends from United Way of Los Angeles, CA; United Way of Southeastern Pennsylvania, PA; United Way of San Francisco/Bay Area, CA; and United Way of the Greater Dayton Area, OH.

Call your local United Way and ask about donor designations. We will be happy to cooperate by submitting any required written data or forms.

I want to help F.I.R.S.T. in the fight against Ichthyosis.

Please put me on the mailing list:

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I don't wish to join now, but please accept my donation of \$ _____

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Ichthyosis type in my family _____ # Persons _____

Mail to: P.O. Box 20291, Raleigh, NC 27619-0921

All contributions to F.I.R.S.T. are fully tax deductible to the extent allowed by state and federal law. F.I.R.S.T. is a 501(c)(3) charitable organization supported entirely by private donations. A copy of the F.I.R.S.T. Annual Report is available to anyone upon request.

Research is well underway

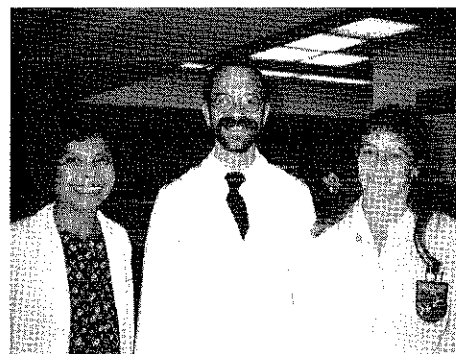
F.I.R.S.T. families participated in NIH research all summer in Bethesda, MD. Research geneticists and dermatologists are off to a great start, thanks to our help in locating ichthyosis patients. This research can only be a success if we continue to support it with our presence.

The purpose of the study is to try to locate the gene that causes ichthyosis.

Dr. Bale says that she receives as many as 30 calls a week from ichthyosis patients. Approximately one out of four calls results in an eligible participant. Dr. Bale and her staff flew to Iowa to study one family with 35 members and went to

Vermont to see a family with 16 members. It was easier to go to the families than to fly that many people to Bethesda. Since July 1, about 50 patients have been seen, and more than 60 are scheduled for visits this fall.

Certain types of ichthyosis are being looked at first. In some cases, all members of a family must participate. The good news is that the project has been funded to provide participants with expense-paid visits to the clinic which is located near Washington, DC. This is a great opportunity for a vacation to our nation's capital and to help with this important research.



Dr. Maria Turner, Dr. Jon DiGiovanna, and Dr. Sherri Bale have been amazed at the overwhelming response from ichthyosis patients to their research project at NIH.

Blood samples and small skin biopsies are needed. If you can volunteer, call Dr. Sherri Bale collect at (301) 496-4375.

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