F.I.R.S.T.
Foundation for Ichthyosis and Related Skin Types

ICHTHYOSIS FOCUS

Vol. 8, No. 2 FALL 1989

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 ichthyoses, a family of primarily genetic skin disorders marked by excessive scaling. Letters, suggestions and contributions are welcome.

Disneyworld extravaganza set for summer of 1990

Get ready for the incredible F.I.R.S.T. National Conference and Family Reunion coming up next summer, on Saturday, June 16, 1990, just four miles from Disneyworld in Kissimmee, Florida.

We are giving everyone advanced notice so that you can start planning next summer's vacation and be a part of the special events we have in store for you. The one-day conference and reunion will take place at the Hilton Inn Gateway, 7470 West U.S. 192, Kissimmee, Fla. 32741. Room rates will astonish you - only $55 per night for the first 25 rooms. Children under 18 stay free.

After the first 25 rooms have been booked, the rates jump to $61 per night. However, this is still a bargain when rates range from $80-$90 per night for summertime near Disneyworld. To book your rooms, call 1-800-327-9170. Be sure to say you are attending the F.I.R.S.T. conference and ask for the special rates.

The hotel has a playground, pool, patio, and game room. Babysitting will be available at an extra charge. Families may get these special rates for three nights if they wish to stay in the area for more fun in the sun. Our board member, Andrea Thorne, is inquiring about group rates into Disneyworld and other places of interest in the area. We will give you an update on this as the information comes in.

Conference registration fees should be sent directly to F.I.R.S.T., P.O. Box 20921, Raleigh, N.C. 27619. Fees are $35 per person or $50 per couple, and $15 per child. Registration fees include coffee and danish, lunch, and an afternoon snack break.

Some of the topics for workshops during the day will include genetics, with basic information on how ichthyosis is inherited, and some of the latest developments in genetic research. We will also conduct small discussions on various topics of interest to you.

Most important, we will have a chance to meet and share with others our own experience, strength and hope. Some people have been reluctant to attend a conference in the past for fear that it may be upsetting. Those of us who have gone have found quite the opposite to be true. We've made good friends. We have discovered we are not alone. Children have been delighted to play with others like themselves, and some not like themselves. It is truly a learning experience, and one filled with many good feelings and memories. You won't want to miss it!

You won't want to miss Tanya Creech in our new video on ichthyosis.

Meet the stars

Each of us is unique and special, and yet, we are no different from other people. We experience the joys and heartaches of life, just like everyone else. We have friends, families, and lovers. We work, play, rejoice and mourn.

Those of us with ichthyosis give birth and raise our own children. We become lawyers, teachers, accountants, salesmen. We participate in all aspects of life. In a new video about ichthyosis, you can meet some of us,

(continued on page 2.)

Kim Creech is one of the stars of the show.

Positive parenting helps children cope

Many parents call the F.I.R.S.T. office with concerns about their child's physical well-being. Their concern is understandable and legitimate. However, sometimes the emotional and psychological development of a child with ichthyosis is even more important.

What can you, as parents, do to help your child develop into a normal, well-adjusted, happy person? F.I.R.S.T. asked Dr. Wilmer C. Betts, a North Carolina psychiatrist, for some helpful tips for raising children with ichthyosis. Dr. Betts is the father of three children with ichthyosis and one child with normal skin.

The most important element, says Dr. Betts, is to "LOVE THEM. They are yours! All children thrive on love, so give them all you can."

Dr. Betts reminds parents that it is important to remember two things: "You didn't cause it, and you can't cure it. But you can do some very helpful things for your child. Realistically, one, you can relieve some of the discomfort and complications; two, you can help with developing a healthy self-image; and three, you can reduce the negative attitudes that the child may have towards him or herself."

It is helpful to let the child know that he is not bad and that he did not cause the ichthyosis. "If other family or normal marital difficulties occur, it's important for the child to understand these are not his fault and are not happening because he has ichthyosis," explains Dr. Betts.

"Allow for freedom and encourage independence. Help your child develop a sense of responsibility for himself, particularly in skin-care. Assist in establishing a routine, but don't push too hard."

Dr. Betts feels, "as a parent, your primary goal is to avoid letting your child develop bitterness, self-pity or an attitude of entitlement."

So, how can parents do this? Dr. Betts suggests using the technique of minimizing. "In other words, use healthy denial. Everyone has something wrong. No one is perfect. And remind your child that there is always somebody worse off."

Dr. Betts cautions parents to try to avoid allowing their own feelings of disappointment to carry over to your child. "Work through these feelings with another adult or with a professional. See your child as different, and yet not different. After all, it is only skin. I always tell my children that it's what is on the inside that counts," he smiles.

Dr. Betts strongly believes that socialization should be encouraged early in a child's life. "Never use ichthyosis as an excuse yourself, and don't let your child use it as an excuse to avoid contact with others or to avoid activities that might be fun," he says.

"Consistency and stability help. People who are familiar and accept ichthyosis provide reassurance to the child that, 'Hey, I am o.k.' Try to provide this stability in school, church and your neighborhood."

Dr. Betts has found that it helps to give children some pat answers which he or she can use to protect himself from unkind remarks or prying questions. "Perhaps you might tell your child to say he just has dry skin, or maybe just ignore rude people. Explain that some people do not have manners and don't know it is impolite to stare," he comments.

"But, don't always shield your child. We can grow from pain. No pain. No gain. Your children will learn to have empathy for others less fortunate."

Dr. Betts has discovered that there are strengths that children can obtain from having ichthyosis. Take a look at the flip side. What are your child's positive attributes? Is he or she more sensitive to other people's feelings? Is your child more willing than most to accept people who are different? Share some of these strengths that you see with your child.

They will grow and thrive, with your help. Many, many adults with ichthyosis live normal lives, raise families, work and play, just like the rest of the world. Your child will, too!

Correspondence Corner

Parker and Debra Piercey have a 2½ year old daughter with CIE. They are interested in meeting others who have children of the same age with ichthyosis. Their address is Rt. 3, Box 478, Ogdensburg, NY 13669. Phone: (315) 393-6699.

Brian Miller, age 30, would like to hear from others who have Ichthyosis Hystrix. His address is 1326 Pennelwood, Toledo, OH 43614. Phone: (419) 382-9302

Patricia Swierczek is working at the American Consulate in Melbourne, Australia and would welcome any letters from Australians with ichthyosis. Her address is: 11/261 Domain Road, South Yarra, Victoria, Australia 3141 Her daughter, Lynn, has lamellar ichthyosis.

Lisa Jane Pullen, age 26, would like to hear from others who have lamellar ichthyosis. Her address is: 21 Allendale Road, Brampton, Ontario, Canada L6W 2Y7. Her phone number is: (416) 459-0304.

If you would like a pen pal, or would like to be in contact with others who have your type of ichthyosis, write to Ichthyosis Focus, and we will include your name and address in Correspondence Corner. We do not release names and addresses without permission.

Meet the stars

(continued from page 1.)

find out who we are and what we can do, and do well. Yes, we are beautiful people with a true zest for life. See for yourself!

To order your copy of the video on ichthyosis, contact Dave Beaver, 168 Fairview Ave., Johnsonburg, PA 15845. Phone: (814) 965-5195. The video costs $22.95. A portion of the proceeds will be donated to F.I.R.S.T. to support its programs.

Many thanks to Dave Beaver for producing this wonderful video, and to all the "stars" who shared a part of themselves to make this documentary a reality.

Ichthyosis Focus has been mailed to you with funds provided through the generosity of Herald Pharmacal, Inc. of Richmond, VA, manufacturers of Aqua Glycolic Lotion and other products for skin care. To order products, call 1-804-745-3400.

Laugh and the world laughs with you

Having a sense of humor can help people with ichthyosis get through some of those rough moments. Board Member Valerie Lutters in Southbury, Connecticut, always has a smile on her face, and if you've ever met her at a conference or talked with her on the phone at the F.I.R.S.T. office in California while she was our administrative assistant, you know she has a laugh that puts you right at ease.

Here's a little quiz Valerie concocted for all the "flaky" people out there. Hope you have as much fun reading it as Valerie did writing it. Thanks, Val!

You know you have ichthyosis if...

So you read a little bit about ichthyosis, perhaps in the F.I.R.S.T. brochures, or maybe you saw a talk show on television. You do have that pesky dry skin and you wonder if you have ichthyosis.

You know you could visit a doctor, get a biopsy and all that, but you'd like a better grasp of the issue before you go and see someone. Well, this questionaire may help.

Read each statement, then check the *one* answer that best describes your response.

1. When you use a public restroom, you:

A. Always wipe the toilet seat before you use it
B. Read everything on the walls and copy some of the choicer bon mots into a notebook you carry for this purpose
C. Always wipe the toilet seat before and after you use it
D. Spray the seat with Lysol and then cover it with the paper sani-guards you keep in your wallet
E. Sing at least one verse of The Star Spangled Banner

2. The OPEC countries have just announced another oil shortage which is expected to last for months. Everyone in the United States is lining up to buy and horde necessities. You have storage space for only two essentials. You line up for:

A. Gasoline and oil for the car and lawn mower
B. Rubber gloves and KY jelly
C. Bath oil and Vaseline
D. Fuel for the blow torches and plastic explosives
E. Olive oil and Gumby dolls

3. This energy crisis is affecting the availability of electricity, and the government has restricted each person to only two electrical appliances. You choose to continue using:

A. The stove and refrigerator
B. The recharging unit for your vibrator and your Tingling Fingers mattress
C. Your vacuum cleaner and your other vacuum cleaner
D. The flood lamps around the house and the electric surveillance system
E. Your HO train set and the blender

4. The last time you went to the seashore:

A. You had a picnic and played volleyball
B. You locked the kids in the car and went to the nude beach
C. Was sometime around the turn of the century when everyone used those charming umbrellas and men's and women's swim suits had tops
D. You left early because the CIA crept in, transparently disguised as Coast Guard officers and life guards
E. You stepped on a jellyfish and had a religious experience

5. Every so often you like to get a glass of wine and a good paperback novel and relax for an hour in a nice hot tub full of:

A. Bubbles
B. Musk
C. Salt, oatmeal, liquid bleach, cooking oil and an industrial strength loofah
D. You never take a bath; the government is putting chemicals in our water
E. Green Jello and feathers

If you answered mostly A's, you don't have ichthyosis but you may be June and Ward Cleaver.

If you answered mostly B's, there's a very short lady named Ruth who would love to talk with you.

If you answered mostly D's, remember: Just because you're paranoid doesn't mean they aren't talking about you.

If you answered mostly E's, we'd like to know what planet you come from and if they have any good treatments for ichthyosis there.

If you answered mostly C's, you may, indeed, have ichthyosis. You should see your dermatologist for a specific diagnosis.

We can offer you friendship and support, news of the latest ichthyosis treatments, and perhaps even a chance to laugh at the absurd problems that are concomitant with living with this disease.

Fears are often worse than reality

By Virginia P. Sybert, M.D.

Genetic counseling provides information about the correct diagnosis and natural history of inherited disorders, their treatment, recurrence risk and prevention, and referral for long term care. Genetic counseling requires communication of all this information so that it is understood and useful, and may require more than one visit.

As a rule, counseling is non-directive, i.e., the health care professional's function is to provide information which allows patients and families to arrive at their own decision based on their needs and beliefs. It is not the function of the counselor to impose his/her own value system on the patient.

While genetic counseling is usually given by a physician trained in medical genetics or by a genetics associate (an individual with a master's degree in Medical or Human Genetics), other health care providers may participate in the process.

Diagnosis

Accurate diagnosis is a prerequisite for genetic counseling. Disorders with *similar manifestations* may have different causes which may be genetic or non-genetic (phenocopy). If a disorder is genetic, mutations of different genes or different mutations of the same gene may produce similar clinical appearances (genetic heterogeneity).

Autosomal dominant, autosomal recessive and X-linked genes, each of which has a different recurrence risk, can cause disorders that cannot be distinguished on clinical grounds alone.

The natural history, treatment and recurrence risks may be different for each of apparently similar disorders. For this reason, a physician expert in the disorder in question should be involved in the evaluation.

The most useful tool for the diagnosis of genetic disease is the family history. To ask the appropriate questions, the counselor must be familiar with the major and minor manifestations of the suspected disorder and be able to ask about them in plain English. Any one person affected with the disorder may have one, some or all of its manifestations, mildly, moderately or severely (variable expression).

In most instances, information about first degree (parents, sibs, children), second degree (aunts, uncles, grandparents, grandchildren) and third degree (cousins) relatives is sufficient. Ethnic origins are usually asked about, as some disorders occur more frequently in one ethnic group than in others.

Some of the most useful questions in obtaining the family history are: "Is there anyone with similar problems in the family?" "Did anyone die young or unexpectedly?" "Did anyone have problems becoming pregnant, or in carrying a pregnancy to term?" "Were there any stillbirths or malformed infants in the family?" "Is there anything traveling in the family, such as the 'Smith ear' or the 'Jones foot'?"

It is important to remember that someone may not recognize a specific problem as part of the spectrum of a given disorder. Frequently, medical records of individuals suspected to be affected need to be obtained.

Family photographs are often invaluable in disorders in which there are specific physical features. An attempt to examine possibly affected relatives should be made.

A disorder may still be genetic in the absence of a positive family history. New dominant mutations, recessive disorders in families with few children and spontaneous X-linked recessive mutations may all give a negative family history and the erroneous impression that a disorder is non-genetic. Variable expression and delay of onset with age may also mask the genetic nature of a disorder.

Multifactorial disorders (those believed to result from an interaction of several genes with specific or non-specific environmental factors) may also give a negative family history.

A negative family history does not rule out a genetic component to a disease or an increased recurrence risk to relatives.

Once the diagnosis is established the counselor needs to explain the natural history or course of the disorder in order to provide a reasoned plan for health care maintenance and to allow for a rational assessment of the burden of a given disorder when decisions for reproduction have to be made.

Decisions about reproduction are often based more on the severity of the disorder than on the risk of recurrence.

Treatment

Treatment for genetic diseases may be specific for the given disorder, or non-specific for the symptoms of the disorder. Awareness of the usual course and common complications of a disorder allow for the practice of preventive medicine. Anticipation of problems may circumvent them.

Recurrence risks

Recurrence risks entail the actual likelihood of an infant inheriting the gene(s) for the disorder, the likelihood of showing the disorder and the likelihood of severity or expression of a given genentic disorder.

The genetic counselor is obliged to inform patients and families of the recurrence risks in a meaningful way and to help the family put these risks into perspective by giving them a framework within which to judge them.

It is not the function of the genetic counselor to make reproductive decisions for the family or to offer value judgements about the family's choices.

(To be continued in the next issue.)

Dr. Sybert is a member of the F.I.R.S.T. Medical Advisory Board. She is associate professor of pediatrics in the Medical Genetics Department and adjunct associate professor in the Dermatology Department at the University of Washington School of Medicine in Seattle, Washington. She practices medicine at Children's Hospital and Medical Center in Seattle, Washington.

Family and friends help Lisa find out how to enjoy life

By Lisa Jane Pullen

For the most part having ichthyosis has been difficult, but my family and friends are very supportive

However, when growing up to say it was rough is an understatement. The school I was attending was very sports oriented. Those not on a sports team were kind of left on the sidelines. In gym class, everyone had to do a 12-minute-run around the football field, including me.

Being unable to sweat made this quite hard. Once the run was completed, usually I was unable to complete the rest of the class because I was too hot. The students felt I was getting off easy and the teacher deducted marks because I was unable to participate in class.

The students came from basically a middle to upper class family so there were certain social groups who led the school; and because of my ichthyosis I was considered to be "different" therefore not allowed to participate.

Some of my teachers used me as a guinea pig. For example, health class was the worst because I was called before the class to explain my condition and answer questions I had no answers to. This made the students notice my ichthyosis even more and made me extremely uncomfortable and unhappy.

Turning sixteen was the absolute worst time. School dances were a nightmare because even though I love to dance I was unable to for any length of time; because my skin was very dry and flaky none of the boys would ask me to dance. Presto, one very unhappy wallflower.

To make matters worse, the first boy who asked me out did so on a dare, I found out after we had seen each other a couple of times and I had come to care for him very much.

After this I guess I went a little nuts. I was pretty much a loner and was even more so after that. During this time, emotionally I was very depressed and had even gone so far as to think about committing suicide.

I guess I was giving off clues as to how very depressed I was because my mother confronted me and brought everything into the open. With my family's help I was able to sort everything out.

As I got older I came to realize that although this limited me in certain ways and caused problems in others, I was still extremely lucky, it could have been a lot worse.

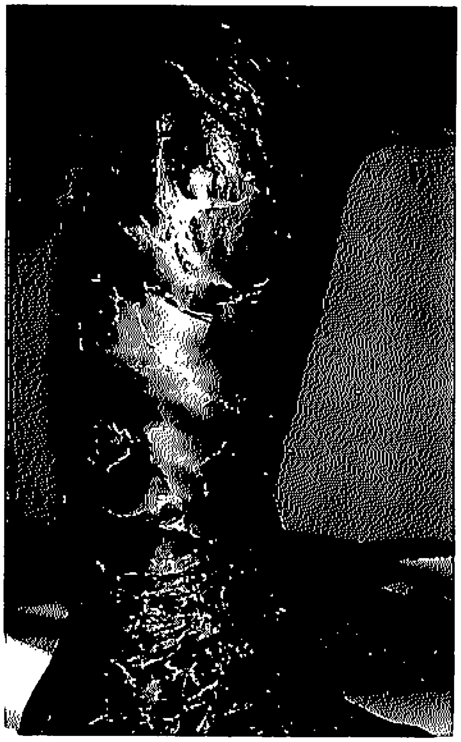
The biggest hurdle to overcome was finding a place to belong. My peers considered me different and the kids (handicapped) in the special class considered me "normal" in that I had sight, hearing and the use of both arms and legs. This made me feel lost.

Now, I am not so lost. I have a great family and a group of very special friends. My job is interesting and I am comfortable with me. You can't ask for more than that!

New products may help some with ichthyosis

Several members have reported that they have had good results using a product named Atractain by Sween. To contact Sween Corporation, phone toll free 1-800-533-0464 (except in MN). The address is: Sween Corporation, Sween Building, P.O. Box 980, Lake Crystal, MN 56055.

Someone wrote and asked if there are any discount pharmacies which sell products for ichthyosis. One reliable company is The Rx Allstates Pharmacy, Order By Mail Service, 360 W. Superior St., Chicago, Illinois 60610-9895. For fast service on credit card orders only, call toll free, 1-800-332-8244, ext. 48 during business hours Monday through Friday.



What type of ichthyosis do you have? The only way to get a confirmed diagnosis is to have a skin biopsy, a relatively painless procedure. Talk to your dermatologist for more information.

Milstone accepts chairmanship

Dr. Leonard Milstone of the Yale University Medical School Department of Dermatology has graciously accepted the position as chair of the F.I.R.S.T. Medical Advisory Board, succeeding Dr. Mary Williams, who has done an admirable job for the foundation.

Dr. Milstone was unanimously selected by your board of directors, and we are looking forward to working with him. He was instrumental in organizing our conference last year in New Haven, Conn. The conference was a huge success as those who attended will well remember.

We are pleased to have him as chair of the Medical Advisory Board. On another note, the Medical Advisory Board will meet on Saturday, Dec. 2 in the Pacific A Room, 4th floor, San Francisco Marriott, 785 Market Street in San Francisco, CA.

F.I.R.S.T. Financial Report

Statement of Cash Receipts and Disbursements
For the year ended December 31, 1988

Receipts:		
Donations	$5,996	
Memberships/newsletter fees	5,366	
Meeting/registration fees	1,382	
Research fund	853	
		$13,597
Disbursements:		
Office supplies	586	
Telephone	798	
Postage	766	
Printing	2,811	
License/permits	52	
Memberships	50	
Transportation/travel	1,403	
Workshop expense	25	
Meetings	125	
Miscellaneous	292	
Management/executive director	5,500	
Public relations	485	
Conference expense	1,001	
Grants	1,500	
Depreciation	720	
		16,114
Excess (deficit) of receipts over expenses		(2,517)
Beginning fund balance at January 1, 1988		12,452
Ending fund balance at December 31, 1988		$9,935

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