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## The National Ichthyosis Foundation

# ICHTHYOSIS FOCUS

THE NATIONAL ICHTHYOSIS  
FOUNDATION

710 Laurel Avenue, #B-8  
San Mateo, CA 94401

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### FROM A MEDICAL POINT OF VIEW

#### DIET AND GENETIC SKIN DISEASE

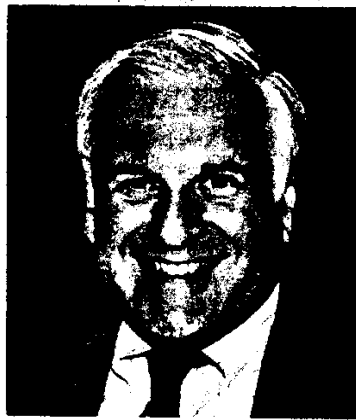
(More Than Skin Deep)

Dietary treatment for genetic disease began when the cause of phenylketonuria (PKU) was discovered in the 1930s. Dietary treatment is now possible for some genetic diseases of the skin and hair. We do not advise any special diet for most forms of ichthyosis, although a diet with more protein than normal may be necessary for those forms of ichthyosis (lamellar and epidermolytic hyperkeratosis) in which there is excessive shedding of scale, since the scale contains proteins which the body has synthesized.

My personal experience with inherited skin disease has profoundly changed my thinking about inherited disorders of keratinization and has broadened my awareness of the systemic nature and possible etiology of diseases which are limited to the epidermis.

The most dramatic instances of a disorder of keratinization occurred in families I studied in North Carolina. A young girl I treated had profound eye and skin disease which severely limited her lifestyle. A special diet has restored her to a normal way of life.

The child was from a rural county in which many of the inhabitants were related (often distantly) to each other. When she was five weeks of age, she had excess tearing and eye pain when exposed to light. These symptoms persisted and progressed until large hazy areas covered the cornea (the clear portion of the eye covering the pupil and iris), allowing her only to distinguish the brightest of light when I met her at the age of 13 months.



Our guest columnist is Dr. Lowell A. Goldsmith, the James H. Stierner Professor of Dermatology at the University of Rochester School of Medicine and Dentistry, Rochester, New York. He has been involved in diagnostic and therapeutic approaches for genetic disorders of keratinization for over a decade. He is chairman of our Medical Advisory Board.

Her skin problems began at the age of five months with painful erosions, which progressed to crusted and scaling lesions on the palms and soles and under some nails. Although she was seen by many specialists previously and had received many therapies, neither her skin nor eyes improved.

My experience with a similar patient in Boston a few years previously led me to believe that she had a defect in the breakdown of tyrosine. Tyrosine is a common amino acid present in most food, but it also can be made in the body from the amino acid phenylalanine.

Chemical tests of the child's blood and urine established a level of tyrosine 22 times normal. This high level of tyrosine was caused by a deficiency of a liver enzyme which breaks down tyrosine. The enzyme, tyrosine aminotransferase (TAT), has been studied for many years, but the consequences of its deficiency are only shown in patients like this child who have a genetic deficiency of the enzyme.

Therapy with a commercial low-phenylalanine, low-tyrosine diet caused the child's disposition to improve within two days and her eyes became less sensitive to light. Within three days, her skin improved and rapidly cleared. Her eyes were better within three days and were almost normal after six weeks. She was restored to normal health by dietary treatment and attends a regular school. She has been maintained on the diet and her blood tyrosine level monitored for almost seven years.

A distant cousin of the patient, a 55-year-old man, had persistent skin symptoms throughout his adult years. His feet were thick and so painful that he walked on his knees to avoid the pain caused by pressure on his feet. After a half century of suffering, the low-tyrosine diet restored him to normal health.

The disease these patients have, tyrosinemia II, has been reported over the past decade in all parts of the world, and all cases have responded to the special diet.

In addition to tyrosinemia, phenylketonuria and Refsum's disease (which is a mild form of ichthyosis) respond to diets which minimize certain nutrients. Other genetic diseases often respond to high doses of vitamins of other amino acids. Each therapy is specific for the individual disease and should be used only when the exact disorder is diagnosed and the patient can be carefully monitored.

The key lesson I learned from tyrosinemia is that a deficiency of a liver enzyme may have manifestations limited to the skin and eyes. This led me to speculate that many of the ichthyoses and other disorders of keratinization are systemic in nature and that the altered metabolism is expressed most markedly in the skin. Such a way of thinking broadens our horizons, leads to new hypotheses and encourages us to look deeper than the skin in our study of ichthyotic and other genetic disorders of the skin.

Printing costs for this issue of Ichthyosis Focus have been defrayed by a grant from the educational division of Hermal Pharmaceutical Laboratories, Inc., Oak Hill, N.Y. 12460.

## Michigan Chapter

Our first fund-raiser is over and we are proud to announce it was a great success! Total donations amounted to \$1,355.00.

On Sunday, May 2, 1982, a personal friend of ours, Denny Sullivan, donated his hair studio for the day. His hairstylists were asked to participate and all gladly accepted the invitation.

As another drawing card, we approached the Detroit Lion Football organization and asked them to donate a football autographed by the Lions to be raffled the day of the Cut-a-thon. Eddie Murray, All-Pro placekicker for the Detroit Lions offered to sign autographs.

To promote the Cut-a-thon we designed a flyer that was reproduced and widely distributed. Major newspapers and radio stations in the Detroit-Metropolitan area were contacted to help promote the event. We would like to single out one media at this time — WPON radio of Pontiac. The station manager interviewed Denny and I on the air for 25 uninterrupted minutes and following the broadcast, we received numerous inquiries about Ichthyosis and the Cut-a-thon.

We would like to extend our special thanks to the following individuals and businesses for their help in making the Cut-a-thon a success.

### BUSINESSES:

Denny's Hair Studio (hair studio)  
The Detroit Lions (autographed football)  
Future Reproductions, Inc. (raffle tickets)  
Laser Personnel, Inc. (flyers)  
WPON Radio, Pontiac (radio broadcast)

### INDIVIDUALS:

Denny & The Denny Hair Studio Staff  
Eddie Murray (All-Pro Detroit Lion)  
Avery Eliau (\$100 Personal Donation)  
Mick & Bonnie King (\$50 Personal Donation)

— Carol and Dick DeLoughary



Nadine: Working on one of her many customers.



Lori: Working as fast as she can before the baby arrives. Lori is due anyday!!!



Eddie Murray: Maybe Eddie is starting Denny's haircut a little low.



Sandy: Ready for the kickoff!!!



The raffle ticket box and football just prior to the drawing.



Carol: Ended up with the job no one volunteered for.



Quinn: Getting ready for the draw!!!



Quinn: And the winner is!!!

## CORRESPONDENCE CORNER

E. Roger Hamor

P.O. Box 6349  
Tampa, Florida 33608

Daughter, Tatiana, has E.H. We would like to hear from others in the Tampa Bay area.

Judy Finnigan

1245 W. 8th Ave.  
Vancouver, B.C. Canada  
Would like to hear from others in the Vancouver area.

A. Hastings Carey, Jr.

RFD 6 Box 18  
Hockessin, Delaware 19707  
Would like to hear from others in the Delaware area.

Pat Harper

147 East Fedora  
Fresno, CA 93704  
Is starting new chapter and would like to hear from interested people in the San Joaquin Valley area.

Love life and life  
will love you back.  
Love people and they  
will love you back.

## CHAPTER HAPPENINGS

by Susan Nye

Over the past few months we have had requests from many of you for information on how to organize and effectively maintain chapters in your communities. In this new section of your Newsletter, we will address these questions on a regular basis, and, hopefully, this will assist you in organizing your chapter.

Our information comes from the Southern San Joaquin Valley Chapter headquartered in Fresno, California. This chapter began in January, 1982, when two mothers, Mary Householder and Pat Harper, were put in contact with each other through the Foundation. They started working with Assistant Director, Susan Nye, to form the initial nucleus of the chapter. Their region covers a 26,000 square mile, predominantly rural, area of central California with a population of 1,470,700. Since they are a relatively new chapter we will have an opportunity to follow their progress along. Their first priority was setting up a sound organizational framework from which to work. Now that this is accomplished, they have begun membership and fundraising drives. (Over seven hundred dollars has been raised, twenty-five paid members have joined and fifteen families with Ichthyosis have been located.) The following is their recommended guide for putting together a chapter and avoiding fragmentation.

### The Executive Committee and Its Responsibilities

Start with five people who want to work with each other as an executive committee. That is, a group that wants to provide leadership, set the goals, plans and direction of the chapter. It is essential that your executive committee have at least two people on it who are not affected personally by ichthyosis but who are committed to the goals of the Foundation (for example, nurses, physicians, teachers, counselors, etc.) These people can provide an objective and realistic view of what can be accomplished in your community because they have not, as many have, been devastated by the disease — a reality that can interfere with the organizing process. In addition, it is helpful if several members of the committee have experience working with organizations; but this is not a requirement for success.

## MEDICAL ADVISORY BOARD MEETING

The Medical Advisory Board met at the recent Society of Investigative Dermatology meeting in Washington, D.C. The N.I.F. wishes to thank all the members who attended for their active support. The Ichthyosis Focus looks forward to publishing your articles. A special thank you to Dr. George Thorne for buying breakfast and to Dr. Lowell Goldsmith for making all the arrangements for the meeting.

The executive committee meets regularly (at least once a month) and stays in contact by telephone. They discuss all the major issues affecting the chapter and arrive at a consensus before making decisions or proceeding. When they decide they want to organize a chapter, they review the Foundation by-laws and request chapter status (a simple procedure) from National Headquarters. Then, one of the committee members accepts responsibility for being the:

### Publicity Coordinator

This person will coordinate all the publicity and advertising regarding the activities of the chapter in their community. This is done primarily through public service announcements (no cost) in the newspaper and on television and radio. The first task is to notify all the dermatologists in the region and inform them of the chapter's plans (contact the Foundation office for sample notification letters). This is for public relations and information only. Your main membership response will not come from physicians. The primary response will come from the media coverage you receive through your public service announcements. Therefore, after you have notified the medical community, place your announcements in the newspaper and on television. Your announcement should direct potential members to telephone a member of the executive committee who is the:

### Self-Help Coordinator

This is a person who wants to assume responsibility for coordinating the support which provides encouragement and reduces the isolation of those suffering from ichthyosis. They insure that regular contact is maintained among persons and schedule support group meetings as necessary. However, this alone is not enough. If people merely meet among themselves your larger community will not learn about your needs. Eventually you will become disenchanted and you will fragment. Then it will be a much longer time indeed, before a cure is found. To be successful you need broad-based support. This means: public awareness, a large membership which reflects a cross section of your community and a strong financial base. Therefore, one member of your executive committee should be the:

### Fundraising Coordinator

This person's primary responsibility is to insure that money is raised. He or she

should be someone who enjoys fundraising. Their task is to plan and organize ongoing fundraising events for the chapter. (The Southern San Joaquin Valley Chapter has monthly rummage sales to raise operating funds for its larger projects — a wine/cheese tasting benefit and raffle scheduled for October 2 and a buffet fashion show next spring.) After approval from your executive committee, the Fundraising Coordinator submits the Chapter's annual budget to the Foundation for review and approval (another simple procedure).

This chapter reports they received more moral and financial support from their community than they had originally imagined. They have also discovered that people enjoy the social contact and sense of purpose they experience from raising money for the Foundation and supporting its goals.

Our goals cannot be reached without funds. It costs money to print literature, to maintain long distance contact by telephone, to purchase supplies and buy postage. More importantly, a stronger financial base will enable us to support ichthyosis research, which is the only sure way to find a cure and bring an end to the suffering.

The remaining two members of the executive committee work primarily on fundraising, membership and public relations. That is, they talk with community groups and service clubs about ichthyosis and the Foundation on a regular basis. In this way you become known to people who are active in your community and they will help spread the word.

Finally, you will need a treasurer to keep track of finances and expenditures (but this person does not have to be a member of the executive committee) and a secretary to keep notes of your meetings so that there will be a record. Accurate recording, following your budget, and keeping receipts of your expenditures are practices which all tax-exempt organizations follow. They must be audaciously adhered to if we are to maintain our tax-exempt status.

I hope these guidelines will help others get started. It is important to note that no one person had the full responsibility for starting this chapter. It was shared. As long as each individual task is accomplished you will be successful as a whole. The National office will assist you all we can but ultimately, you must do the work. It can be accomplished if you give just several hours a week on a consistent basis. If any of you are interested in organizing a regional chapter in your community, please contact us.

In the meantime, to boost your enthusiasm, anyone who raises ten thousand dollars for the Foundation will have their way paid as one of our representatives to the American Academy of Dermatology Meeting this December 5th through the 9th in New Orleans, Louisiana!



Clockwise from left: Robert A. Briggaman, M.D.; Gerald G. Kreuger, M.D.; Peter Elias, M.D.; Mary L. Williams, M.D.; Howard Baden, M.D.; Lowell Goldsmith, M.D.; Laurence Miller, M.D.; and George Thorne, M.D.

# ANGER

Jane S. Rome, M.D.

"Why me?" or "Why my child?" must be nearly universal thoughts associated with Ichthyosis. These thoughts are inevitably accompanied by strong feelings of anger. It is understandable that the affected person, child or adult, should have angry feelings. But when siblings or parents or other family members also experience anger, they often feel it is somehow wrong or unreasonable or unjustifiable. But it happens. In fact, it would be unusual if it did not happen, in some form, sooner or later. This anger may be directed toward the physician, the hospital, a husband or wife, unknown ancestors, or other children. Hardest of all to accept is anger toward the person with Ichthyosis. Certainly it is unfair to feel anger toward a person because he or she has a disease. Fair or unfair, right or wrong, justified or not, it happens.

There are several problems to deal with simultaneously. One, acknowledging the feeling as a real (and therefore valid) feeling. Two, deciding how to handle the feeling within oneself and with the person who has Ichthyosis. Three, learning how to "get it out" or manage the feeling so that it doesn't get in the way. Different people do this in different ways. Some people talk directly; some talk to an uninvolved person. Some people are not talkers and deal with anger by some physical activity such as jogging or playing tennis. Sometimes it is helpful for children to be allowed to punch a pillow or other inanimate object. These are only a few of many good and useful ways to deal with anger. Sometimes, however, these approaches do not work. In many cases, families find professional counseling a useful way to learn to handle issues of anger associated with Ichthyosis and other long-term diseases.

Angry feelings also have the potential for being a positive force. They may be one way of asking for someone to listen, or for closeness or understanding. Some families learn to look at anger as a signal that talking needs to be done, or that someone needs a certain kind of attention.

Ichthyosis is a family problem. Anger can be a family problem, too. Learning to handle and even benefit from anger can be an enriching experience for a family.

Jane S. Rome, M.D., is Assistant Clinical Professor of Pediatrics at the University of California, San Francisco. A member of the Child Study Unit, at UCSF. Dr. Rome specializes in developmental pediatrics and family therapy.

## FUNDRAISING

The response to the need for money was rather disappointing, to say the least. Only a handful of people responded. And to those who did respond, I salute you. Your donations showed tremendous caring and sacrifice.

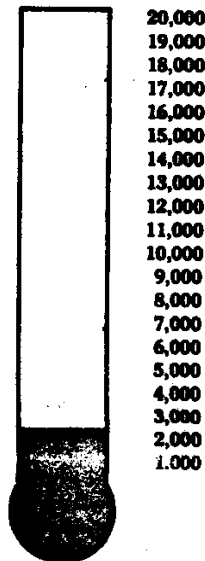
Hoffman-LaRoche Laboratories responded with a \$750.00 donation, for which we are most grateful.

Pat Mondt, of New Jersey, single-handedly raised \$300.00 from family and friends and has vowed to keep going.

The Michigan Chapter made over \$1,200 from their first major fundraiser and are now in the planning stages for their next one.

Mary Householder of the Fresno, California Chapter has single-handedly raised in the neighborhood of \$700.00 by talking to friends and holding garage sales. The Fresno Chapter is also in the process of planning their first major fundraiser.

\$20,000



WHAT CAN YOU AND YOUR CHAPTER DO TO HELP US REACH OUR GOAL???

by John Brennan  
Accountant and Member  
Board of Directors

I know many Chapters and individuals are planning fundraising efforts such as bake sales, bowl-a-thons, walk-a-thons, and other creative ideas. Along with the rest of the Board of Directors, I am grateful for your energy and enthusiasm.

I am also the Board member who looks after the Foundation's accounting and tax filings. Yes, even the N.I.F. must file tax returns every year. Most non-profit organizations do, even though they don't owe taxes.

The N.I.F. benefits from its non-profit tax status in several ways. It makes contributions deductible on the contributor's income tax return. Lower postal rates become available, too. Sending you this newsletter would be more expensive without these special postal rates.

How does this affect you as a fundraiser for N.I.F.? You must help in two ways —

1. A list of all cash must be kept or checks received, as they are collected. The amount of every expense you pay to help you raise money must also be recorded. Describe the expenses well. Get and keep receipts for them.

2. The Foundation's Bylaws say that the funds you raise must be sent to the N.I.F. National Office. Along with these proceeds, send a copy of the

records you kept and any expense receipts. Keep copies for yourself, too.

Your chapter may wish to keep money for local activities. If so, you must request the money back from the National Office after the results of your fundraising success have been received and recorded there. The Foundation's policy has always been to return to chapters some of the money they raise. Chapter needs in excess of this allocation should be discussed with the Chairman. Some funds must always be kept by the National Office for its costs (such as mailing this newsletter!) and for the national education and research programs that are so badly needed.

Good luck and happy fundraising!

## AMERICAN ACADEMY OF DERMATOLOGY MEETING

The 40th annual meeting of the American Academy of Dermatology was held in San Francisco, December 5-10. It was attended by 4000 physicians. The National Ichthyosis Foundation was represented by the following members: Judy Droste, Deatra Landwehr, Jim O'Brien, Susan Nye and Barbara Landwehr. This representation enabled us to distribute literature about our organization. It also provided the opportunity to discuss ichthyosis with various pharmaceutical companies who also exhibited at the meeting as well as with numerous physicians.

The next meeting of the Academy will be held in New Orleans, Louisiana in December, 1982.