



ICHTHYOSIS FOCUS

Special Issue

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

Annual Report

Ichthyosis is a relatively rare disease with no cause or cure. Although at least one in every 100,000 people suffers from it, the disease has received little or no attention and has remained an enigma to doctors for thousands of years.

1991 marks the 10th anniversary of the Foundation for Ichthyosis and Related Skin Types, the only organization in the world which is dedicated specifically to helping people who suffer from this genetic problem. F.I.R.S.T. has had a busy year completing its first decade. Newsletters were published quarterly and response from donors was heart warming. Contributions ranging from \$5 to \$1,000 were sent by about 350 individuals and companies who are determined to see F.I.R.S.T. reach its goals.

Last summer 101 people attended the National Conference and Family Reunion in Orlando, Fla. Speakers included leading dermatologists Lawrence Schachner, M.D., Mary Spraker, M.D. and John Koo, M.D., all members of F.I.R.S.T.'s outstanding medical advisory board. Plans have already begun for a 1991 summer conference July 3rd in Williamsburg, Va.

"I am encouraged by the increase of support we have seen this year," says Ellen Rowe, president of the foundation. Mrs. Rowe, mother of two children ages 10 and 13, was re-elected as president for another term beginning Jan 1, 1991. "I have been touched by the compassion and energy shown by so many parents of children who have ichthyosis. I have ichthyosis myself, but my children do not. I have no concept of the tremendous dedication these parents have to their children. We also have grandparents who have become very actively involved in F.I.R.S.T. I would like for all of them to know how much we appreciate their continued support."

Total contributions for 1990 topped out at \$27,728.00. These funds were spent to rent an office, pay the phone bill, hire a part-time secretary, print newsletters and brochures, mail information to interested people, send representatives to Washington D.C., and sponsor a national conference. In order to accomplish those tasks, F.I.R.S.T. spent \$25,278.00 We are still in business, folks, and going strong.

Leonard Milstone, M.D. of Yale University remains as chair of the medical advisory board. Assisting Ellen Rowe as co-vice presidents for the coming year will be Frances McHugh of Delran, New Jersey, and Andrea Thorn of Jackson, Mississippi. Frances has a grandson with EH, and both Andrea and her daughter, Anna, have ichthyosis. Kathy Rogers of Austin, Texas will continue in the position of treasurer. She also has a daughter, Angie, with ichthyosis. "Bebe" Blades functions as secretary and office manager for the foundation. Other board members include Cynnle Bates of Tuscaloosa, Alabama; Hans Kummer of Melbourne, Florida, Lynne Alba of Norristown, PA and Linda Morgan of Ellicott City, MD. Cynnle, Linda and Lynne have children with ichthyosis, and Hans, a college student at F.I.T., has ichthyosis himself.

The office will be open from 1 p.m. to 4 p.m. An answer machine will take your message if you call during other hours. Contributions to the newsletter are welcomed. The board is also interested in your suggestions and comments for helping F.I.R.S.T. progress in the next decade.

The inside pages of this report contain information you may wish to share with friends and relatives, or even strangers who ask about ichthyosis. Feel free to make copies of this material for your own use. If you would like brochures or pamphlets to hand out in your community, please contact the F.I.R.S.T. office.

Goals and Objectives

The Foundation for Ichthyosis and Related Skin Types is a nonprofit 501(c)(3) corporation dedicated to helping individuals and families with the genetic skin diseases collectively called Ichthyoses, a family of primarily genetic skin disorders marked by excessive scaling. The means by which F.I.R.S.T. hopes to achieve this goal include:

- Publishing a quarterly newsletter which provides information regarding the medical, psychological and social aspects of this rare disorder for patients and their families.
- Advocating for scientific research to find better treatments and eventually a cure for Ichthyosis.
- Serving as a resource network to help relieve the physical and mental distress among persons suffering from Ichthyosis by providing practical advice, guidance and moral support.
- Creating and distributing educational material about Ichthyosis to the public and medical profession.
- Sponsoring workshops and conferences for patients and their families.
- Seeking opportunities for national exposure and recognition of this disease, thus creating greater public awareness.

The Foundation is a non-profit tax exempt organization incorporated in California on January 1, 1981 and now headquartered in Raleigh, NC.

Guidance for F.I.R.S.T. is provided by a volunteer board of trustees consisting of professional and lay people, both with and without Ichthyosis. The Foundation is also supported by an outstanding Medical Advisory Board, composed of physicians and scientists who have a special interest in the field of Ichthyosis.



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ICHTHYOSIS FACT SHEET

ICHTHYOSIS ('ik-thee-q-sis') is the general name for a group of almost forty inherited and acquired diseases that include severe scaling of the skin as a primary symptom. Some are recessive, some are dominant. All are part of the American gene pool and are passed on from generation to generation. Their cause and cure are unknown. Ichthyosis sufferers have little or no hope for themselves or their children.

The ichthyoses are disorders of cornification, i.e., formation of the top skin layer, the stratum corneum. In some forms, the skin grows too fast, while in others, the skin grows at a normal rate. In both forms, however, the individual microscopic skin cells don't release from each other and the skin surface quickly enough. Apparently, something is wrong with the "glue" holding the cells together. They form a rigid, thick surface layer of scales that discolor and crack.

In the most severe forms of the disease, babies die shortly after birth from dehydration and infection. Other forms of ichthyosis are associated with mental retardation, immune deficiencies, deafness, baldness, bone deformities, vision impairment, infected blisters, impaired mobility, and lack of body temperature control, as well as the psychological devastation that accompanies bodily disfigurement.

Over a million Americans suffer from the most common type of ichthyosis, while rarer forms affect about 100,000 people. Every year, almost 17,000 people are born in the U.S. with one of these diseases, facing a life of limited productivity and social interaction because of their disability and disfigurement.

There is no known cure for any of these diseases. There is very little research done to understand them. There is only one prescription drug approved specifically for ichthyosis, and it is effective only in some cases. Daily treatment costs, not to mention loss of productivity costs, can reach tens of thousands of dollars each year.

Research into the causes of the diseases can shed light on many other, more common maladies. Understanding these disease models can help our understanding of immune systems, cell growth patterns, and the basic genetic blue print of human life. Most important, research offers hope.



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ICHTHYOSIS CONGENITA

U.S. Population

TYPE	PREVALENCE	PER MILLION	TOTAL PEOPLE	BIRTHS YEARLY
Darier's	1 : 50,000	20/million	5,000	75
CIE	1 : 100,000	10/million	2,500	37
Lamellar	1 : 200,000	5/million	1,250	19
X-linked	1 : 2,000	500/million	62,500	1,865
EH	1 : 100,000	10/million	2,500	37
Harlequin	1 : 500,000	2/million	500	7
Other	1 : 10,000	100/million	25,000	373
Vulgaris	1 : 250	4,000/million	1,000,000	14,924

Total U.S. Population in 1990 - estimated 250,000,000

Total U.S. Births in 1990 - estimated 3,800,000

Every year almost 17,000 people are born in the U.S. with one of these rare diseases, facing a life of limited productivity and social interaction because of their disability and disfigurements.

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

Please put me on the mailing list:

NAME _____

MAILING ADDRESS _____

CITY _____ STATE _____ ZIP _____

Donor Category:

Single \$20/year Family \$35/year Sustaining \$50/year

Sponsor \$100/year Patron \$250/year

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

I can't afford to contribute now, but please keep me on the mailing list. (Please check)

Ichthyosis type in my family _____ # Persons _____

Mail to: P.O. Box 20921, Raleigh, N.C. 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.

Medical Advisory Board

The Foundation for Ichthyosis and Related Skin Types is supported by an outstanding Medical Advisory Board composed of physicians and scientists who have a special interest in Ichthyosis, a genetic skin disease characterized by excessive scaling. The means by which the Medical Advisory Board offers guidance and support to the Foundation include:

- Providing patients with information about the services available from F.I.R.S.T.
- Advocating for F.I.R.S.T. at professional meetings.
- Increasing public awareness of the disease by serving as consultants for the media and providing accurate source material to journalists.
- Editing a medical column for the newsletter which is published quarterly.
- Contributing information about new treatments and research for press releases and the newsletter.
- Lecturing at national conferences which are attended by patients and family members.
- Designating a recipient each year for the F.I.R.S.T. Travel Award.
- Participating in a regional referral system for patients.

The Foundation for Ichthyosis and Related Skin Types is a nonprofit 501(c)(3) corporation dedicated to helping individuals and families with the genetic skin diseases collectively called Ichthyoses cope with the medical, psychological and social aspects of this rare disorder.

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