



Date: June 12, 2007

Program

**First World Conference on Ichthyosis
August 31 – September 2, 2007**

Münster, Germany

Organized by
Network for Ichthyoses and related keratinization disorders (NIRK)
together with
Selbsthilfe Ichthyose e.V.
and
EU-Coordination Action GENESKIN

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Location: Lecture Hall
 Department of Dermatology
 University Hospital
 Von Eschmarch-Str. 58
 48149 Münster
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Friday, August 31, 2007

8:30	Opening of the conference
	Welcome and greetings by Prof. E. Schlatter, Dean for Research Prof. T. Luger, Head of Department of Dermatology B. Kleinow, Selbsthilfe Ichthyose e.V. Prof. H. Traupe, Speaker of NIRK
	Workshop on clinical diversity and diagnostic standardization Chair: E. Sprecher and H. Traupe
9:00	D. Metze, Münster Histopathology of ichthyoses: Clues for diagnostic standardization
9:20	I. Hausser, Heidelberg Ultrastructural characterization of lamellar ichthyosis: A tool for diagnostic standardization
9:35	F. Ückert, Münster The data base behind the NIRK register: a secure tool for genotype/phenotype analysis
9:50	V. Oji, Münster Classification of congenital ichthyosis
10:10	M. Raghunath Congenital Ichthyosis in South East Asia
10.25	Panel discussion: How do we name autosomal recessive nonbullous congenital ichthyosis? E. Sprecher, P. Steijlen, H. Shimizu, A. Vahlquist, H. Traupe
10:40	Coffee break
	Keratinization disorders and keratins Chair: P. Steijlen, A. Vahlquist
11:15	I. Hausser, Heidelberg Ultrastructure of keratin disorders: What do they have in common?
11:25	M. Arin, Köln Recent advances in keratin disorders
11:45	E. Sprecher, Haifa Naegeli-Franceschetti-Jadassohn Syndrome: a Keratin Disease

12:05	P.M. Steijlen, Maastricht Epidermolytic palmoplantar keratoderma with “tono tubular” keratin
12:25	Panel discussion: Epidermolytic keratinization disorders: how should we call them? Panel: P. Steijlen, E. Sprecher, D. Metze, I. Haußer
12:40	Lunch break
	Molecular advances in epidermal differentiation
14:30	D. Kelsell, London Role of connexin isoforms for epidermal differentiation and wound healing
14:50	L. Bruckner-Tudermann, Freiburg Role of kindlin in human disease and keratinocyte motility
15:10	M. Guerrin, Toulouse Granular keratinocytes transcriptome: Identification and characterisation of new differentiation markers
15:30	K.H. Grzeschik: Molecular basis of focal dermal hypoplasia
15:50	Coffee break
17:15	Bus transfer for the reception and dinner GOP Restaurant and Vaudeville, Münster

Saturday, September 1, 2007

	Recent advances in gene mapping and in lipid genes Chairman: J. Fischer, H.C. Hennies
8:30	J. Fischer, Paris Mapping genes for nonbullous autosomal recessive congenital ichthyosis: What we know today
8:50	H.C. Hennies, Köln Functional understanding of mutations in congenital ichthyosis
9:10	P. Krieg, Heidelberg 12R – Lipoxygenase Deficiency impairs Skin Barrier Function
9:30	G. Schmitz, Regensburg Apolipoprotein E and lipid traffic within keratinocytes
9:50	H. Shimizu, Sapporo What can we learn from Harlequin ichthyosis?
10:10	Anna Thomas, London In vitro models for harlequin ichthyosis
10.25	Coffee break
10:50	R. Happle, Marburg The CHILD syndrome revisited: the clinical perspective
11:05	A. König, Marburg Functional understanding of NSDHL mutations
11:20	P. Elias, San Francisco Similarities in the Pathogenesis of the Ichthyosis in Two Lipid-Metabolic Disorders – RXLI and NLSDI
	European and international perspective
11:40	G. Zambruno, Rome EU coordination action GENESKIN Purpose, structure and achievements of GENESKIN
12:00	I. Zwoch, Bonn Orphan diseases and the European Union – what patients and scientists may expect
12:20	M. Williams, San Francisco Structure and aims of Foundation for Ichthyosis and Related Skin Types
12:40	Lunch break
	Joint workshop together with Selbsthilfe Ichthyose e.V. and European network of self support groups for ichthyosis

	Therapy of ichthyosis: a challenge in daily practice Chair: G. Wehr, M.Arin
14:00	A. Vahlquist, Uppsala Introduction to the topic: therapy of ichthyosis/general principles and substances
14:20	M.L. Preil, Bad Salzschlirf Management of ichthyosis: The TOMESA experience
14:40	A.M. van Steensel, Maastricht Our experience with RAMBAs in treatment of congenital ichthyosis
14:55	H. Traupe, Münster Results of an ongoing study with Liarozol for lamellar ichthyosis
	Topical treatment/the patient perspective
15:10	N.N. The experience from Germany
15:25	Jean de Witt, Belgium The experience from Belgium
15:40	Vladio Minelli, Italy The experience from Italy with special focus on the scalps
16:10	N.N. (Sveden) (2 persons) What can be done for palms and soles
16.25	Panel discussion and questions for experts
16:45	Coffee break
	Experimental therapies
17:15	M. Braun-Falco, Freiburg Gene therapy for keratinization disorders: what is the current state?
17:35	D. Roop, Denver Oligonucleotide therapy for keratin disorders
17:55	H. Traupe, Münster Enzyme replacement therapy of lamellar ichthyosis: the current state
18:15	J. A. McGrath, London Cell therapy approaches: the example of Epidermolysis bullosa
18:35	End of program for the day Evening at individual disposal

Sunday, September 2, 2007

	<p>Proteases and keratinization disorders Chairman: A. Taïeb und D. Hohl</p>
9:00	<p>P. Hachem, Brussels Importance of serine proteases for epidermal differentiation</p>
9:20	<p>A. Taïeb, Bordeaux Insights into Pathogenesis of Ichthyosis in Trichothiodystrophy Syndromes</p>
9:40	<p>A. Hovnanian, Toulouse Towards functional understanding of Netherton syndrome</p>
10:00	<p>A. Ishida-Yamamoto, Distinct intracellular transport for different epidermal lamellar body molecules</p>
10:20	<p>Coffee break</p>
	<p>Ichthyoses and the cornified envelope Chairman: W.H. I. McLean and M. Paulsson</p>
10:45	<p>W.H. I. McLean, Dundee Ichthyosis vulgaris and filaggrin: what we know today</p>
11:05	<p>S. Weidinger, München Genetics of epithelial barrier integrity in atopic diseases</p>
11:25	<p>M. Paulsson, Köln Transglutaminase-3 deficient mice: a subtle skin phenotype</p>
11:45	<p>WK Jacyk, Pretoria Bathing suit ichthyosis, the South African experience</p>
12:05	<p>K. Aufenvenne, Münster Towards functional understanding of bathing suit ichthyosis</p>
12:25	<p>B. Ahvazi, Bethesda Modelling of transglutaminase-1 and transglutaminase-3: what can we predict?</p>
12:45	<p>End of the conference</p>