International Symposium on Metabolic Disorders and Nutrition Related to the Skin, sponsored by the European Society for Dermatological Research, Utrecht, March 8-10, 1990

Metabolic Disorders and Nutrition Correlated with Skin

Volume Editors
B.J. Vermeer, Leiden
K.D. Wuepper, Leiden/Lake Oswego, Oreg.
W.A. van Vloten, Utrecht
H. Baart de fa Faille, Utrecht
J.C. van der Schroeff, Leiden

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VI. Protoporphyria
During a sabbatical experience at Leiden University Hospital in the summer of 1990, it was my pleasure to review in detail the 26 manuscripts printed in these Proceedings. For one who could not attend the ESDR Clinically Oriented Symposium on 'Nutrition and Metabolic Diseases' held in Utrecht, The Netherlands, March 8-10, 1990, the flavor of the meeting comes through well. Here one can discover many major unsolved problems in clinical dermatology with approaches to their solution. Examples are the role of foodstuffs or their additives in elicitation of contact urticaria, atopic dermatitis or systemic eczematous allergic contact dermatitis. Other articles provide useful solutions to the care and feeding of patients with gliaden-sensitive dermatitis herpetiformis.

A major section of this book is devoted to improved chemical or genetic understanding of the photosensitive porphyrias. The elegant delineation of two codon mutations in alleles of two patients with Gunther porphyria defines the precise structural defect and explains the decreased activity of the enzyme uroporphyrinogen III synthetase. The old 'leaky enzyme' hypothesis can now be put to rest. New diagnostic tools, allele-specific oligonucleotides, offer the potential for rapid and improved prenatal diagnosis. Additional papers in this section of the book point to several useful animal models of certain porphyrias that bear further investigation. Finally, improved biochemical analysis in variegate porphyria, porphyria cutanea tarda of the sporadic or familial types, and coproporphyria offer new insights into these diseases and regulation of porphyrin metabolism. Newer ultrastructural studies of erythropoietic protoporphyria point to 'silent' hepatic injury that was unsuspected by other methods.

The wayward expression of bioactive molecules by malignant cells continues to be discovered and good examples are found in the production of other pancreatic proteins in the glucagonoma syndrome and in parathyroidlike hormones elaborated by squamous cell carcinomas in some instances of humoral hypercalcemia of malignancy. The skin is a perpetrator or a target in a vast array of paraneoplastic syndromes! Current emphasis upon receptor-ligand interactions, intracellular signaling mechanisms and gene regulation has focused heavily upon receptor
molecules [see J Invest Dermatol 1990;94: 1-174S]. Yet defective ligand molecules are now getting attention. The review of patients with familial dysbeta lipoproteinemia emphasizes how single codon mutations in apolipoprotein E disturb the binding of this VLDL or IDL ligand to its receptor. Dermatologists can be on the watch for xanthoma- or xantho chromiapalmaris striata as clinical clues to the detection of these patients. This volume will have interest for dermatologists and for their colleagues in clinical nutrition, clinical chemistry or pathology, genetics and allergy. The old hack ‘You are what you eat’ is well modified here; one is not only what we eat but how our genetic make-up reacts to what we eat. These manuscripts complement, rather than duplicate, the 36th Annual Symposium on the Biology of Skin which was devoted to ‘Nutrition and the skin’. That symposium addressed disorders of vitamin and mineral metabolism, protein-calorie malnutrition and selected heritable disorders [Arch Dermatol1987;123:1375-1385a, 1535-1547a, 1674-1706a]. The organizers and editors of this Symposium have assembled a wealth of information in a well-defined sphere of dermatologic interest. A careful read will be well rewarded.

Leiden, The Netherlands Kirk D. Wuepper, MD