



Ichthyoses: 39 (Current Problems in Dermatology)

P.M. Elias, M.L. Williams, D. Crumrine, M. Schmuth

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The various manifestations of ichthyoses are classified either by their appearance or their molecular genetics. This volume focuses on generalized, inherited disorders of cornification, which constitute an ever-enlarging group of monogenic diseases caused by a large number of genes that affect a broad array of cellular functions. The authors' overview reflects their unique perspective that the clinical phenotype in the inherited ichthyoses mirrors a 'best attempt' by a metabolically compromised epidermis to maintain a barrier sufficiently impermeable for survival in a desiccating external environment. The basis for threats to survival is illuminated, and the systemic problems, including growth failure, also reflect a compromised barrier. A new consensus classification of these disorders is provided, and the distinguishing clinical features of each disorder are described. Further, the latest molecular genetic information is succinctly reviewed with up-to-date and comprehensive references. Yet, the major emphases of this volume are on disease pathogenesis and on the identification of key ultrastructural features. This publication will prove an invaluable aid to dermatologists, pediatric dermatologists and pediatricians dealing with patients with inherited ichthyoses. In addition, clinical geneticists and dermatopathologists will find it interesting reading.

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