Joel L. Spitz, editor. Genodermatoses.
A Clinical Guide to Genetic Skin Disorders.

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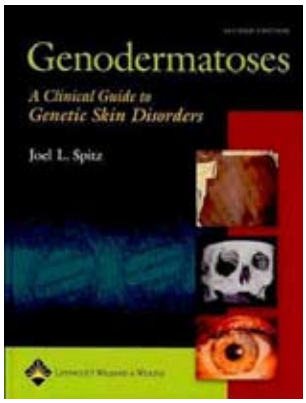

Although dermatologists frequently deal with genodermatoses in practice, this is one of the rare books in which this complex matter has been adjusted for use in daily routine. In the last decades, considerable progress has been made in the field of genetics and in the understanding of genetically determined disorders. The editor of the book gathered 12 authors, while Vaune J. Hatch made a great job by in enriching the contents of the book with more than 300 full-color illustrations. Each syndrome is described on one to two pages. When there are synonyms, they are listed at the beginning. Bulleted text summarizes the patterns of inheritance, prenatal diagnosis, incidence, age of presentation, pathogenesis, key features, differential diagnosis, laboratory data, management and prognosis. Boxed features highlight clinical pearls and breadth to the material. Full body diagrams and clinical photographs of each syndrome are included. At the end of each chapter, additional reading is suggested. In the first chapter, the disorders of cornification are described. Description of Mal de Meleda is found in this chapter. In chapter two, disorders of pigmentation are described, i.e. oculocutaneous albinism types I and II, then Hermansky-Pudlak syndrome, Chediak-Higashi syndrome, Griscelli syndrome, Waardenburg syndrome, piebaldism, incontinentia pigmenti, LEOPARD syndrome, Carney complex, neurofibromatosis I and II, McCune-Albright syndrome and tuberous sclerosis. In chapter three, among disorders of vascularization, description is included of the new PHACE syndrome (Posterior fossa brain malformations, large facial Hemangioma, Arterial anomalies, Cardiac anomalies and aortic coarctation, Eye abnormalities, sternal clefting and/or supraumbilical raphe). The importance of PHACE is that it should alert clinicians seeing infants with large segmental facial hemangiomas to be aware of the potential structural anomalies of the heart, eyes, central nervous system and arterial vasculature. The fourth chapter of the book is dedicated to connective tissue disorders. Genodermatoses with malignant potential, i.e. basal cell nevus syndrome, xeroderma pigmentosum, Muir-Torre syndrome, dyskeratosis congenita, Peutz-Jeghers syndrome, Cowden syndrome, Gardner syndrome, multiple endocrine neoplasia type IIb and new Birt-Hogg-Dube syndrome, are presented in a separate chapter. It is followed by description of a group of epidermolyses and porphyrin metabolism disorders, a group of genodermatoses associated with photosensitivity, and disorders associated with immunodeficiency. In the chapter on hair and nail disorders, two new syndromes are described: EEC (Ectrodactyly-Ectodermal Dysplasia-Cleft lip/palate syndrome) and AEC (Ankyloblepharon filiforme adenatum-Ectodermal Cleft palate syndrome). In chapter 12, the following disorders associated with chromosomal abnormalities are presented: Down syndrome, Turner syndrome, Noonan syndrome and Klinefelter syndrome. The book ends with a chapter on genodermatoses characterized by short stature, followed by highly useful lists of support groups, institutions and laboratories all over the world specialized in the study and management of particular genodermatoses.

This book is specially designed for dermatologists, pediatricians and family doctors as a guide to genetic skin disorders and an ideal resource for clinical practice.

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