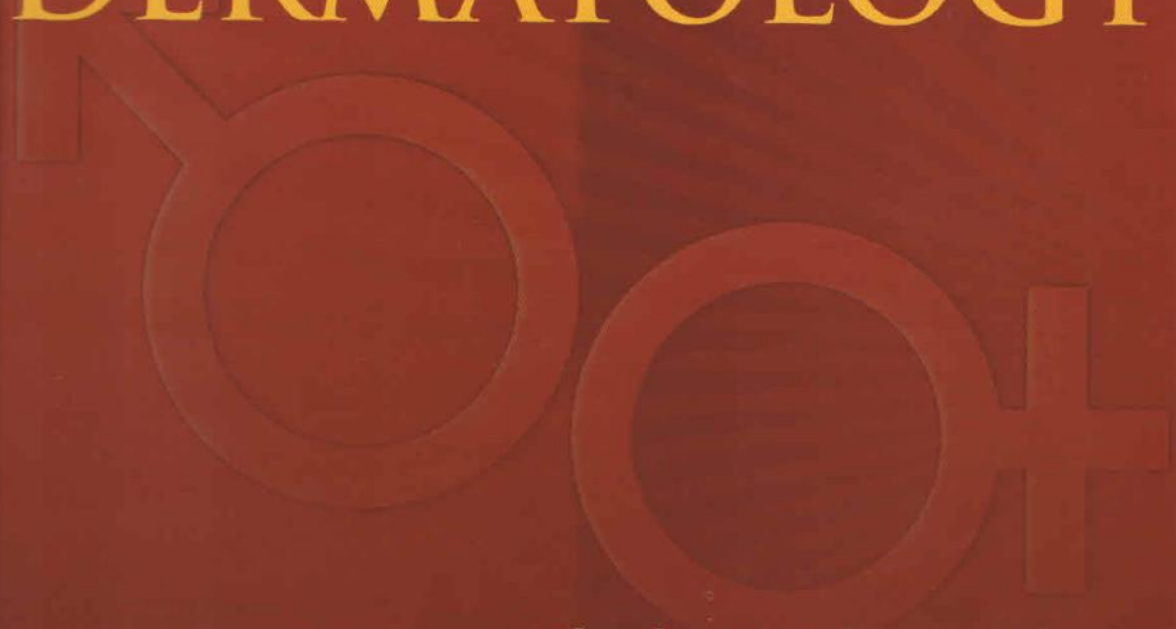


Manual of **GENDER**
DERMATOLOGY



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C O N T E N T S

Contributors	v
Preface	ix
Part I. Primer of Gender Dermatology	1
Chapter 1. Gender Dermatology Defined	3
Chapter 2. Principles of Gender-Specific Medicine	5
Chapter 3. Genetics and Molecular Biology of the Sexes	15
Chapter 4. Gender, Sex Hormones, and the Skin	25
Chapter 5. Gender and Immunology	31
Chapter 6. The Many Faces of Gender: Gender Psychology	37
Part II. Life Cycle	41
Chapter 7. Infancy	43
Chapter 8. Childhood	55
Chapter 9. Young Adulthood	67
Chapter 10. Mature Adulthood	73
Part III. Heredity and Environment	81
Chapter 11. Genodermatoses	83
Chapter 12. Environmental Effects	97
Part IV. Diseases of Skin Structure—Non-Hereditary	105
Chapter 13. Diseases of the Sebaceous, Apocrine, and Eccrine Glands	107
Chapter 14. Nail Disorders	117
Chapter 15. Hair Disorders	121

Genodermatoses

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Overview

Sex hormones vary in their effects on the development of the immune system and in their function in men and women, resulting in gender differences in the etiology, incidence, and course of inflammatory and autoimmune diseases in the field of dermatology. Opposed to these hormonal effects is the group of genodermatoses, where different genetic defects are significant.

Genodermatoses are manifested by the entire absence or insufficient production of specific proteins (enzymes or structural proteins). In most instances, these genetic defects are inherited by autosomal chromosomes that are the same in both sexes, not by the sex chromosomes X and Y, according to Mendel's principles of inheritance (autosomal-dominantly or autosomal-recessively). In these instances, both sexes have the same probability of expressing the traits, and significant differences cannot be expected in the incidence and clinical course of the diseases in the men, as compared to women.

X-linked inheritance is an exception. This is in contrast to the inheritance of traits with autosomal chromosomes. Due to the existence in humans of many more genes on the X chromosome than on the Y chromosome, there are many more X-linked traits than Y-linked traits. Genes that are presented on the X or Y chromosome are called sex-linked genes. In humans, the female is the homogametic sex, having two X chromosomes (XX), while the male is heterogametic, having one X and one Y chromosome (XY). Because men have only one X chromosome, they will express any recessive characters.

This chapter on genodermatoses will focus on those conditions with an X-linked inheritance, in view of the fact that these are gender-specific.

Genodermatoses with X-Linked Recessive Inheritance

If a given mutation is sufficient to cause disease in a man (XY), then we speak of X-linked recessive inheritance. Women typically are not affected. They are heterozygous carriers with one normal and one abnormal X gene. Because of female carriers, skipped generations are the rule, rather than the exception.

X-Linked Ichthyosis

Synonyms: Recessive X-linked ichthyosis, steroid sulfatase deficiency disease.

Definition: X-linked ichthyosis is an inherited genetic disorder of the skin that results from steroid sulfatase (STS) deficiency. The disease affects boys and men with generalized scaling and usually begins soon after birth (third to sixth month of life). It has an incidence of 1 in 6000 in boys and men.

Clinical manifestations: Dark discrete medium-sized scales, especially visible on the extensor surface are the hallmark of the disease (Figures 11-1a and b). All surfaces are involved. At the site of major folds, the skin appears lighter in contrast to the nigricant aspect of the surrounding skin. In severe cases, the face shows dark, fine desquamation with underlying erythema. The scalp is covered by