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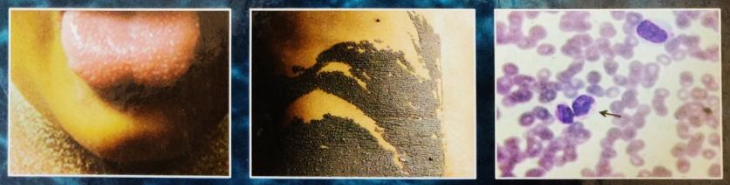

Foreword
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JAYPEE

32 CHAPTER Ichthyotic Disorders of Skin

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INTRODUCTION

Ichthyotic disorders are mostly congenital and manifest at or around birth. However, some systemic diseases and exogenous factors like drugs, may give rise to ichthyotic changes in skin.

These are designated as secondary ichthyotic disorders. The discussion in this chapter will primarily highlight the congenital ichthyotic disorders.

The term "ichthyosis" is descriptive of "fish-scale", derived from the Greek word "ichthys". Congenital ichthyoses are characterized by variable degree of generalized, persistent scaling, with a chronic course throughout the life.

Ichthyotic disorders are characterized by either increased stratum corneum production or abnormal corneocyte shedding along with altered skin barrier function. Clinical expression is highly variable ranging from mild ichthyosis vulgaris (IV) to severe, lethal harlequin ichthyosis (HI). Categorizing the ichthyosis subtypes is often difficult without genetic studies.

CONGENITAL ICHTHYOSIS

Classification

The current system being followed for classification of congenital ichthyoses has been derived from the "2009 Ichthyosis Consensus Conference on Terminology and Classification of Inherited Ichthyosis." The classification of ichthyosis has been presented in table 1.

TABLE 1: Classification of ichthyosis

Nonsyndromic forms		Syndromic forms	
Common ichthyosis	• Ichthyosis vulgaris (IV) • Recessive X-linked ichthyosis (RXLI), nonsyndromic	X-linked ichthyosis syndromes	• RXLI (Syndromic form) • IFAP syndrome • Conradi–Hünermann–Happle syndrome
Autosomal recessive congenital ichthyosis (ARCI)	Major types • Harlequin ichthyosis (HI) • Lamellar ichthyosis (LI) • Congenital ichthyosiform erythroderma (CIE) Minor variants • Self healing collodion baby (SHCB) • Acral SHCB • Bathing suit ichthyosis (BSI)	Autosomal ichthyosis syndromes	• *Prominent hair abnormalities*: ○ Netherton syndrome (NS) ○ Ichthyosis-hypotrichosis syndrome (IHS) ○ Ichthyosis-hypotrichosis-sclerosing cholangitis (IHSC) syndrome ○ Trichothiodystrophy (TTD) syndrome • *Prominent neurologic signs*: ○ Sjögren–Larsson syndrome (SLS) ○ Refsum syndrome ○ Mental retardation-Enteropathy-deafness-neuropathy-ichthyosis-keratoderma (MEDNIK) syndrome • *Fatal disease course*: ○ Gaucher syndrome type 2 ○ Multiple sulfatase deficiency (MSD) ○ Cerebral dysgenesis, neuropathy, ichthyosis, and palmoplantar keratoderma (CEDNIK) syndrome ○ Arthrogryposis-renal dysfunction-cholestasis (ARC) syndrome

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