Exfoliative ichthyosis

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Exfoliative ichthyosis. ORPHA:289586

Exfoliative ichthyosis is an inherited, non-syndromic, congenital ichthyosis disorder characterized by the infancy-onset of palmoplantar peeling of the skin (aggravated by exposure to water and by occlusion) associated with dry, scaly skin over most of the body. Pruritus and hypohidrosis may also be associated. Well-demarcated areas of denuded skin appear in moist and traumatized regions and skin biopsies reveal reduced cell-cell adhesion in the basal and suprabasal layers, prominent intercellular edema, numerous aggregates of keratin filaments in basal keratinocytes, attenuated cornified cell envelopes, and epidermal barrier impairment.