Keratosis linearis-ichthyosis congenita-sclerosing keratoderma syndrome

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Keratosis linearis-ichthyosis congenita-sclerosing keratoderma syndrome. ORPHA:281201

Keratosis linearis-ichthyosis congenita-sclerosing keratoderma syndrome is an inherited epidermal disorder characterized by palmoplantar keratoderma, linear hyperkeratotic papules on the flexural side of large joints (cord-like distribution around wrists, in antecubital and popliteal folds), hyperkeratotic plaques (on neck, axillae, elbows, wrists, and knees), mild ichthyosiform scaling, and sclerotic constrictions around fingers that present flexural deformities.