Incontinentia pigmenti

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Incontinentia pigmenti. ORPHA:464

Incontinentia pigmenti (IP) is a rare X-linked dominant multi-systemic ectodermal dysplasia usually lethal in males and presenting neonatally in females with a bullous rash along Blashko's lines (BL) followed by verrucous plaques evolving over time to hyperpigmented swirling patterns. It is further characterized by teeth abnormalities, alopecia, nail dystrophy and affects occasionally the retina and the central nervous system (CNS).