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10th Annual World Congress on

Pediatrics, Pediatric Gastroenterology & Nutrition

March 23-25, 2017 Orlando, USA

Incontinentia pigmenti in a boy

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n 11 years boy is brought with complaints of abnormal movements, fever, passing mucous stools, vomitings and decreased vision A in left eye of about 10 days. Boy was delivered at term vaginally. Child had blisters with erythematous base on his trunk and upper and lower limbs, since the first few weeks of life. Later, hyperpigmentation developed. He received various treatments with topical medications, albeit without success. On clinical evaluation, the following clinical features of IP are observed: CNS: Seizures (GTCS), Dental: White opaque lines seen on all labial surfaces of the teeth with retained deciduous teeth and pegged lateral incisor, partial anodontia, Eye: Cotton wool spots, Skin: Hyper pigmented hyperkeratotic plaques over lines of blaschko over thighs, both legs and axilla, lower trunk and buttocks. Skin biopsy showed hyperkeratotic epidermis with mild acanthosis and basal cell degeneration, vacuolization and decreased pigment content. Melanophages extend into the epidermis and also into papillary dermis.

Biography

Sunil Kumar Gonuguntla has completed his MD Pediatrics from Dr. NTRUHS, Vijayawada, India. He is currently working as a Resident in SVRR GGH, Tirupati, India.

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