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## De Sanctis Cacchione Syndrome

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De Sanctis Cacchione syndrome is the most severe and extremely rare variant of xeroderma pigmentosum which is an autosomal recessive trait(1). The associated features of this syndroms are microcephaly, mental retardation, premature closure of sutures, retarded growth and sexual development, choreoathetosis, cerebellar ataxia, quadriparesis, sensorineural deafness, and shortening of Acheles tendon. The last three manifestations occur later in life(2).

### Case Report

An 8-month-old male child, born to second degree consanguinous parents, was admitted for evaluation of vomiting of 15 days duration. On examination, he had microcephaly with closed anterior fontanelle, developmental retardation, scaling erythema and freckling of exposed areas which worsened on exposure to sunlight. The laboratory investigations were normal, except for urinary tract infection with *E. coli*. His urine showed generalized aminoaciduria which was persistent after control of urinary tract infection. The serum aminoacidogram was normal. An assay of immunoglobulins showed raised levels of IgA-134 mg/dl (N=14-100), IgG-1616 mg/dl (N = 500-1200) and IgM 241 mg/dl (N=43-239). His vomiting was controlled with antiemetics and adequate treatment of urinary tract infection.

### Discussion

De Sanctis Cacchione syndrome was first described in 1932. Sporadic cases as in this report are occasionally observed.. The neurological abnormalities are progressive, and frequent infections are common in these children. Associated immunodeficiency is reported in some and the reported case has high IgE(3). Elevated levels of tryptophan, glycine and galactose in urine were reported in two cases(4). Generalized aminoaciduria and definite hydropic degenerative changes in the renal tubular epithelium were noted by Siegelman and Sutow at autopsy in two non-related patients(5). Ours is the first case reported in infancy with an association of hypergammaglobulinemia and generalized aminoaciduria. Difficulties in differentiating this syndrom from Hartnups disease may arise but the absence of microcephaly and neutral aminoaciduria aids in the diagnosis of the latter. Further studies are required to evaluate hypergammaglobulinemia as an association.

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Management includes protection from exposure to ultraviolet light, genetic counselling and follow-up for malignancies. Amniocentesis for cell culture and early interruption of pregnancy may aid in prevention.

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## Superficial Fungal Infections in Newborns


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Superficial fungal infections in the newborn period are extremely rare and include candidiasis, aspergillosis and dermatophytosis(1,2). Although dermatophytes are ubiquitous, most exposures do not result in clinical infections(3). Congenital cutaneous candidiasis, often extensive but benign form of neonatal infection, is an uncommon entity(3). We report three cases of neonatal superficial fungal infections seen among 16,000 babies over a 5-year period.

**Case Reports**

*Case 1:* A full term male baby, developed rashes on the 3rd day of life which became generalized discrete pustules on the 4th day. The lesions involved even the palms and soles (*Fig. 1*), but spared the scalp and genitalia. The baby was free of constitutional symptoms. A smear from the lesions revealed *Candida* and culture con-

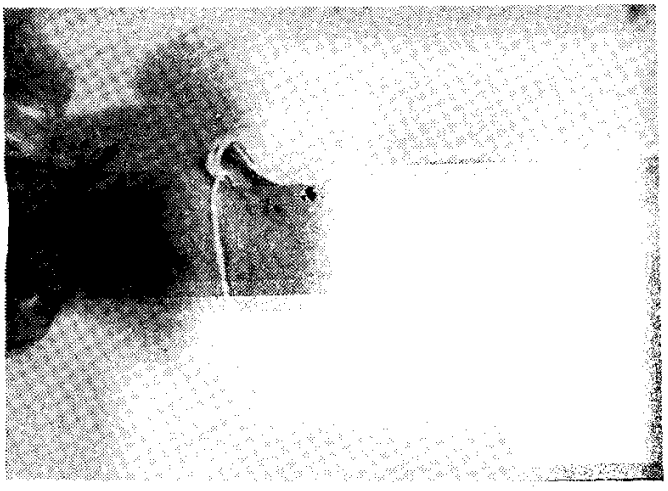

*Fig. 1. Close-up of upper limb showing the pustular lesions.*


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