Incontinentia Pigmenti
Incontinentia pigmenti (IP) is an X-linked dominantly inherited disorder. Females with the abnormal gene on only one of their two X chromosomes are hemizygous for this condition and hence are so severely affected that they typically die in utero. This explains the predominance of female patients with this disorder. To date, over 40 male patients with IP have been reported. The male patients have had a single Y chromosome or a Y chromosome with a compensatory X chromosome, as in cases of 47,XXY or Klinefelter syndrome. The familial form of this disorder, IP2 (or 3/11...
The alterations in the second stage consist of acanthosis, irregular papillomatosis, and hyperkeratosis. Intraepidermal inflammatory infiltrate intermingled with melanophages. This infiltrate extends into the epidermis in many places. Classical incontinentia pigmentosa, is localized to the Xq28 region. It is due to a mutation in the IKK-gamma gene as part of the NEMO complex.
The disorder has four stages. The first stage, consisting of erythema and bullae arranged in lines, either...
In about 80% of the cases, IP is associated with various congenital abnormalities, particularly of the central nervous system, eyes, and teeth. Partial alopecia at the vertex is also often seen.

**Histopathology**

The vesicles seen during the first stage arise within the epidermis and are associated...
The areas of pigmentation seen in the third stage show extensive deposits of melanin within melanophages.
A different pattern has recently been described on the skin of the legs of an infant in whom the vesiculation...
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Pathogenesis

The fact that the first two stages of IP are seen predominantly on the extremities and the third stage mainly on the trunk has led to the assumption by some authors that the pigmentary changes of the third stage occur independently of the bullous and verrucous lesions of the first two stages. Even in the first stage, many keratinocytes and melanocytes show degenerative changes resulting in the presence of dyskeratotic keratinocytes in the epidermis during all three stages of the disease have been confirmed.
The presence of eosinophils in epidermal and dermal infiltrates can be explained by the presence in the early vesicular stage of basophils, which release eosinophil chemotactic factor.
of anaphylaxis. Eosinophil chemotactic activity has been demonstrated in patients with IP in the blister fluid and in eluates of crusted scales overlying the lesions.