Benign familial chronic pemphigus
Hailey-Hailey Disease
Familial benign pemphigus is inherited as an autosomal dominant trait, with a family history obtainable in about two-thirds of patients. Only very few instances of mucosal lesions have been reported, of the mouth, the labia majora, and the esophagus.
Although, as in Darier's disease, early lesions may show small suprabasal separations, so-called lacunae, fully developed lesions show large separations, which are elongated papillae lined by a single layer of basal cells, protrude upward into the bulla, and in some cases,
narrow strands of epidermal cells proliferate downward into the dermis. Many cells of the detached stratum malpighii show loss of their intercellular bridges; thus, acantholysis affects large portions of the epidermis.

Individual cells and groups of cells usually are seen in large numbers in the bulla cavity. Despite the extensive loss of epidermal bridges, the epidermis remains loosely attached to the dermis by a thin layer of basal cells.
Differentiation of familial benign pemphigus from Darier's disease as a rule is not very difficult, because in Darier's disease a bulla is easily formed since there is aclotulation of the cells in the suprabasal region; and dyskeratosis consisting of the formation of corps ronds and grains is much more evident.

Pemphigus vulgaris often resembles familial benign pemphigus to a striking degree, and in some specimens, histologic examination may be difficult. In case of doubt, immunofluorescence will decide the issue.
There used to be much discussion as to whether familial benign pemphigus represents a vesicular varia
Evidence against a relationship is also shown by the fact that in affected families, always only one of the two diseases 3q is mutated in Hailey-Hailey, while Darier's disease is due to a mutation in the ATP2A2 gene on chromosome 12.
Many of the cells of the stratum malpighii that have lost all or most of their intercellular bridges show a fairly normal cytoplasm and a normal nucleus in which mitotic activity has even been observed. Some of the acantholytic cells, however, have a homogenized cytoplasm, suggesting premature partial keratinization. In the stratum granulosum and lucidum, acantholytic cells or acantholytic cell bridges are present. The foamy cells resemble the grains of Darier’s disease. Occasionally, a few corps ronds are present in the granular layer.

**Differential Diagnosis**

Histologically, familial benign pemphigus shares certain features with both Darier...
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