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 epidermis, the surrounding skin structures look healthy.
Differentiation of familial benign pemphigus from Darier’s disease as a rule is not very difficult, because in the former the suprabasal region is more clearly affected and dyskeratosis is much less evident. In Darier’s disease, on the other hand, the suprabasal region is less involved and dyskeratosis is more prominent.

Pemphigus vulgaris often resembles familial benign pemphigus to a striking degree, and in some specimens, histologic examination may be of no help in distinguishing the one from the other. In such cases, immunofluorescence will decide the issue.
There used to be much discussion as to whether familial benign pemphigus represents a vesicular variant of Darier's disease with vesicular lesions or cases of familial benign pemphigus with the presence of corps ronds.
Evidence against a relationship is also shown by the fact that in affected families, always only one of the two diseases is due to a mutation in the ATP2A2 gene on chromosome 3q, 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 other cases, the cytoplasm may be quite vacuolated, resembling 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's disease and pemphigus vulgaris. In familial benign pemphigus, suprabasal separation of the epidermis caused by acantholysis and resulting in lacunae or bullae and villi formation.