Benign familial chronic pemphigus = اﻠﺴﻠﻴﻢ اﻠﻌاﺌﻠﻲ اﻠﻤزﻤﻦ اﻠﻔﻘاﻊ _ ﻫﻴﻠﻲ ﻫﻴﻠﻲ
Hailey-Hailey Disease
Familial benign pemphigus is inherited as an autosomal dominant trait, with a family history obtainable in about two thirds of cases. 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.
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 intercellular bridges, the keratinocytes remain attached to each other by desmosomes.
Differentiation of familial benign pemphigus from Darier's disease as a rule is not very difficult, because in ... 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 ...
There used to be much discussion as to whether familial benign pemphigus represents a vesicular variant...
Evidence against a relationship is also shown by the fact that in affected families, always only one of the
diseases is found to be present.
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 suprabasal layers, the acantholytic cells form larger plaques and intercellular bridges among themselves.

Differential Diagnosis:

Histologically, familial benign pemphigus shares certain features with both Darier's disease and pemphigus vulgaris. In both conditions, there is suprabasal separation of the epidermis caused by acantholysis and resulting in lacunae or bullae and villi formation.