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## FAMILIAL BENIGN CHRONIC PEMPHIGUS (HAILEY-HAILEY DISEASE)

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### SUMMARY:

Familial benign chronic pemphigus (Hailey-Hailey disease) is a blistering dermatosis, which is inherited as an autosomal dominant trait and usually presents around the third and fourth decades. Painful erosions, vesico-pustules and scaly erythematous plaques appear at sites of friction such as the sides of the neck, the axillae, the groins and the perineum. A case of familial benign chronic pemphigus in a 54 year-old woman is presented. The disease started at the age of 35. The areas of predilection are the axillary and submammary folds and genital area. Erythematous, macerated plaques were found with multiple painful fissures, vesicles and crusts in its periphery. The diagnosis was established on the basis of physical examination and was confirmed by histological examination of a skin biopsy. A suprabasal cell separation (acantholysis) of the epidermis was found. A family history is present. An autosomal dominant mode of inheritance was confirmed by genealogical analysis. Thirteen members have been affected by the same disease. In the presented case topical and systemic antibiotics, antimycotics, corticosteroids and retinoids were applied with a transient result. The patient was treated with topical Pimecrolimus which showed a good result. Postlesional hyperpigmentations were found after this treatment.

**Key words:** Familial benign chronic pemphigus, Hailey-Hailey disease, acantholysis, Pimecrolimus.

### INTRODUCTION:

Familial benign chronic pemphigus (Hailey-Hailey disease) is a blistering dermatosis, which is inherited as an autosomal dominant trait and usually presents itself around the third or fourth decades. Painful erosions, vesico-pustules and scaly erythematous plaques appear on the sites of friction such as the neck, axillae, groins and perineum.

### Case report:

A 54-year-old woman with a nineteen-year history of flexural blistering eruptions was presented. The disease started at the age of 35 with recurrent erythema, vesicles and erosions in the intertriginous areas – axillary, submammary,

inguinal folds and the neck. Bullous and erosive lesions epithelised slowly without scars. There was no seasonal influence, however the skin eruptions appeared around menstruation. The patient's father and other 11 relatives in three consequent generations were affected by the same disease. Physical examination showed erythematous, macerated plaques with multiple fissures, peripheral vesicles and crusts in the axillary and submammary folds and the genital area (Fig.1). The routine blood count and urine analysis were within the normal limits. Microbiological examination revealed *Candida albicans* and *Staphylococcus aureus*. Histological examination of skin biopsy showed intraepidermal clefts of varying sizes both suprabasally and higher in the epidermis, as well as the characteristic incomplete acantholysis giving it the appearance of a "dilapidated brick wall" (Fig. 2). An autosomal dominant mode of inheritance was confirmed by genealogical analysis. Thirteen family members have been affected by the same disease (fig. 3). The patient's medical history revealed a negative direct immunofluorescence evaluation, which is consistent with a diagnosis of Hailey-Hailey disease (or chronic benign familial pemphigus). In the presented case systemic and topical antibiotics (Clindamycin caps., Fucidin cream), antimycotics (Itraconazole), topical corticosteroids and retinoids were applied with a transient result. Finally the patient was treated with topical pimecrolimus two times a day with a good result. All of the lesions healed within 2 weeks. Postlesional hyperpigmentations were found after this treatment (fig. 4). We observed no side effects like pruritus and burning sensation. The patient was followed up.

### DISCUSSION:

Hailey-Hailey disease, or chronic benign familial pemphigus, is a genodermatosis arising in adult age with recurrent vesicles and erosions primarily in the flexural areas. Along with the typical intertriginous localization of Hailey-Hailey disease, generalized cutaneous involvement may also occur (6, 10, 14). It is an autosomal dominant skin disorder characterized by abnormal keratinocyte adhesion in the suprabasal layers of the epidermis. The responsible defect has been identified in the gene named ATP2C1 on