

## Hailey Hailey disease – A case report

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### I. Introduction

Hailey Hailey disease is a rare benign chronic genetic disorder characterised by recurrent painful and malodorous blisters and erosions typically in the skin folds like the armpit, groin and under the breast. It is caused by mutations affecting skin adhesion and symptoms are commonly triggered by heat, friction or sweat.

In 1939 Hailey brothers first described the disease. They were American dermatologists William Howard Hailey (1898-1967) and Hugh Edward Hailey (1909-1963). They are famous for discovering this disease.

Z Wu in 1925 cited one article reporting Hailey Hailey disease and was treated effectively.

W Turczym in 2026 concluded that Hailey Hailey disease remains as a therapeutic challenge due to its chronic relapsing nature.

### II. Background

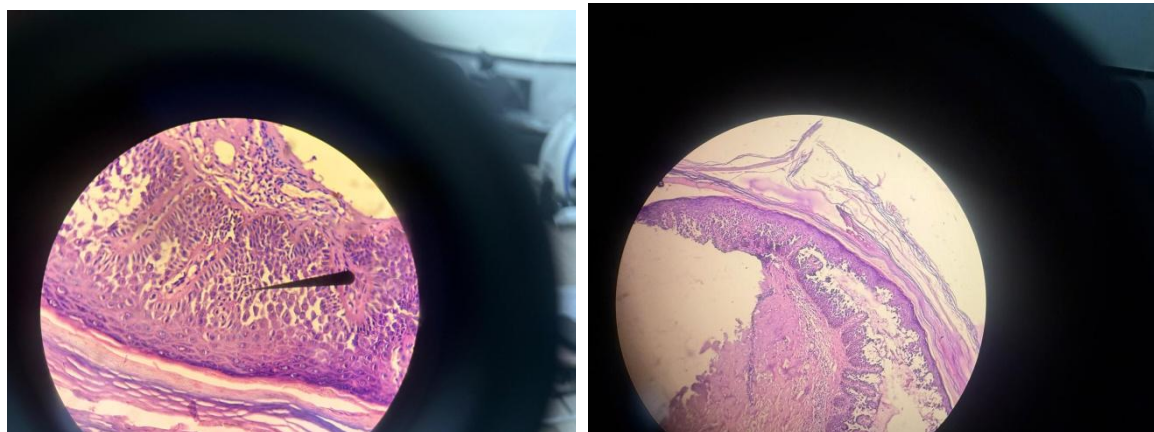
A 32y old female presented in the derma opd with blisters. The patient was examined and the soft tissue was taken for biopsy and sent for histopathological examination. The tissue specimen was processed and on microscopic examination showed stratified squamous epithelium with acantholysis at various levels of epidermis, predominantly at the supra basal level showing a dilapidated brick like patterns. Dermal papillae are elongated at places lined by single layer of basal cells. Occasional dyskeratotic scales are also noted. Sub epithelium shows a mild mixed inflammatory cell infiltrations.

### III. Methodology

This case report was prepared in Pragjyotishpur Medical College and Hospital, a tertiary care hospital established at Birubari Guwahati. This is a new setup and this case was amongst the first few cases of histopathology done in Pragjyotishpur Medical College. The processing of the tissue was done in our laboratory itself and was reported subsequently.

### IV. Results:

It is a benign condition and runs in families. It is genetically induced and mostly affects the middle-aged men and women. The result was confirmed by histopathology.



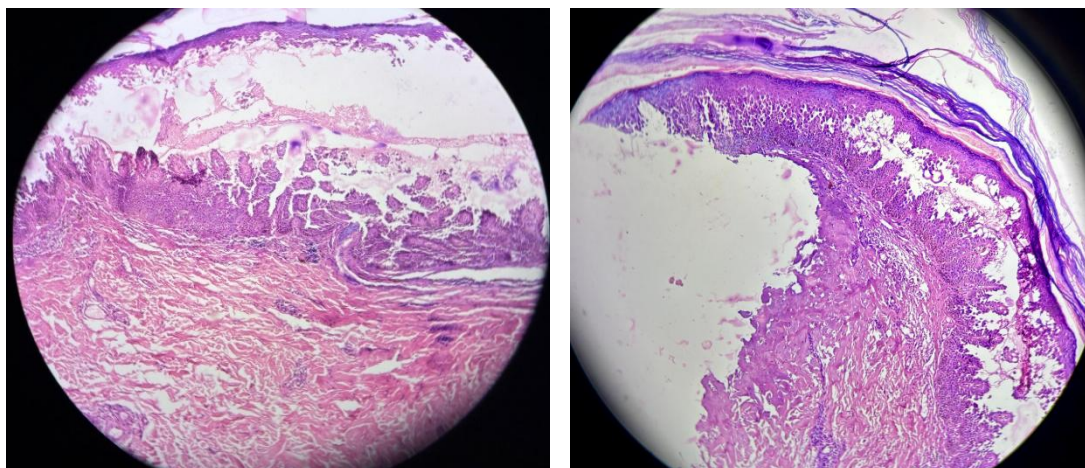


Fig – Photo Micrograph showing Hailey Hailey Disease

Genetic studies have localized the key mutations to the ATP2C1 gene on chromosome 3q, specifically 3q21-q24. Clinical variability is attributed to haplo insufficiency. This locus encodes the SPCA pumps that are responsible for ATP dependent calcium transport along with the SERCA – proteins that are mutated in Darier disease. The defective pump protein leads to decrease calcium content in the usually calcium rich basal keratinocytes. It is hypothesized that the calcium defect disrupts protein processing leading to failure to process desmosomal proteins and therefore loss of cell-cell adhesion. Keratin expression and keratinocyte differentiation may also be affected. Exacerbation by physical stress such as heat and friction may lead to an additive decrease in SPCA expression.

## V. Discussion

After discovering Hailey Hailey disease in 1939 it was reported worldwide.

N Kodali in 2025 mentioned Hailey-Hailey disease and said that it is an autosomal dominant genodermatosis caused by mutation of the ATP 2C1 gene.

VM Patel in 2019 cited – Hailey Hailey Disease (HHD) is an autosomal dominant genodermatosis characterised by crusted macerated erosions.

AM Porro in 2024 cited Hailey Hailey disease is a rare genodermatosis described in 1939.

PR Goswami in 2025 cited 1 – Hailey Hailey disease (HHD) commonly known as benign familial pemphigus.

It is a rare genetic disease occurs 1 in 50000 people.

## VI. Conclusion

Hailey Hailey disease is a rare genetic disorder which may occur from early adulthood to middle aged people. It is best treated by corticosteroids and topical ointments. It can occur in those people who are having a genetic preponderance for the disease. Our case is one of such.

Conflict of Interest – There is no conflict of interest.

Funding – No funding received from anywhere.

Permission from Head of the Institute was taken before hand.

## Reference

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