Department of Dermatology, Venereology and Leprosy
Sree Balaji Medical College and Hospital, Chennai

Journal discussion (09/09/2025)

Title of journal: THERAPEUTIC AGENTS FOR HAILEY-HAILEY DISEASE: A NARRATIVE REVIEW

Presented by: Dr Madhumita R(Junior Resident)

Abstract: Hailey-Hailey disease (HHD) is a rare autosomal dominant blistering disorder caused by ATP2C1 mutations leading to impaired calcium homeostasis and keratinocyte adhesion. Management remains challenging, as most therapies target secondary inflammatory pathways or triggers rather than the primary genetic defect. This review analyzed available literature from 1984 to 2024, classifying therapies based on mechanism and level of evidence. Conventional measures include avoidance of triggers, topical corticosteroids, calcineurin inhibitors, antibiotics, and antifungals, while systemic options range from corticosteroids, retinoids, and immunosuppressants to newer agents like naltrexone, magnesium chloride, vitamin D, and biologics. Recent focus is on dupilumab and JAK inhibitors, which act via modulation of Th2 cytokines. Procedural and surgical interventions such as botulinum toxin, NB-UVB, dermabrasion, lasers, and grafting provide additional options in refractory cases.

Conclusion: Despite advances in understanding the pathogenesis of HHD, long-term remission remains difficult to achieve. Current therapies largely provide symptomatic control, and many patients require combination or sequential treatment. Emerging agents targeting cytokine pathways and calcium regulation, particularly dupilumab and JAK inhibitors, offer promising results but lack robust trial data. Until stronger evidence is available, management should focus on individualized, stepwise approaches—beginning with trigger avoidance and supportive care, escalating to systemic agents or biologics in resistant disease, and considering procedural interventions as a last resort.

Journal citation :Sardana K, Bansal A, Muddebihal A, Khurana A.

Indian J Dermatol Venereol Leprol. 2025;91:462–9.

DOI: 10.25259/IJDVL_906_2024