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**Literature search results**

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**Search details**

Antibiotics, macrolides, erythromycin and clindamycin for the treatment of hailey-hailey disease of familial benign pemphigus.

**Resources searched**

NICE Evidence; TRIP Database; Cochrane Library; EMBASE; MEDLINE; Google Scholar

**Database search terms:** antibiotic*; ant-biotic*; exp ANTIBIOTICS; exp ANTI-BACTERIAL AGENTS; “anti-bacterial agent*”; “antibacterial agent*”; macrolides; MACROLIDES; erythromycin; ERYTHROMYCIN; clindamycin; CLINDAMYCIN; hailey-hailey; PEMPHIGUS, BENIGN FAMILIAL; “familial benign pemphigus”; pemphigus adj1 benign adj1 familial

**Evidence / Google Scholar search string(s):** ("hailey hailey" OR "hailey-hailey" OR "familial benign pemphigus") (antibiotics OR macrolides OR clindamycin OR erythromycin)

**Summary**

I wasn’t sure if you were looking at just the antibiotics: macrolides, erythromycin and clindamycin, or antibiotics as well as the three named drugs. For this reason I have included results for the three named drugs first, and then included additional research looking at antibiotics more generally.
### Guidelines and Policy
None found.

### Evidence Reviews
None found.

### Published Research – Databases

**Macrolides, erythromycin and clindamycin only**


**Author(s)** Usmani N., Wilson C.

**Citation:** Journal of the European Academy of Dermatology and Venereology, February 2007, vol./is. 21/2(264-266), 0926-9959;1468-3083 (February 2007)

**Publication Date:** February 2007

**Source:** EMBASE

Available in fulltext from *Journal of the European Academy of Dermatology & Venereology at EBSCOhost*

Available in fulltext from *Journal of the European Academy of Dermatology & Venereology at EBSCOhost*

2. A review on Hailey-Hailey disease

**Author(s)** Cheng T.S.

**Citation:** Hong Kong Journal of Dermatology and Venereology, March 2007, vol./is. 15/1(10-16), 1814-7453 (Spring 2007)

**Publication Date:** March 2007

**Abstract:** Hailey-Hailey disease (HHD) is a rare autosomal dominant blistering skin disease first described in 1939. It is characterised by vesicular or crusted erosions on the neck and in intertriginous areas. Though it is known that the gene responsible for HHD is located on 3q21-q24, it is only recently found that the disease is a result of the mutations in the Ca<sup>2+</sup> ATPase ATP2C1. A spectrum of mutations have been reported since the identification of the mutation. The disease not only causes itching, unpleasant smell, it might also lead to pain. Thus, the disease may cause significant impact on quality of life. A variety of modalities of treatment have been offered to the patients with various success. This paper reviews the literature to provide a current understanding of the disease.

**Source:** EMBASE

Available in fulltext from *Hong Kong Journal of Dermatology and Venereology at Free Access Content*


**Author(s)** Deng A, Lowitt M

**Citation:** SKINmed, July 2012, vol./is. 10/4(251-3), 1540-9740;1540-9740 (2012 Jul-Aug)

**Publication Date:** July 2012

**Abstract:** A 70-year-old woman urgently presented with severe eruptive skin dermatitis
associated with fever and malaise 7 days after taking clindamycin for an unknown skin eruption. She had a 40-year-long history of well-controlled Hailey-Hailey disease. Physical examination revealed erythrodermic skin changes covering more than 80% of the patient's body surface, with hundreds of nonfollicular pustules. Many of the pustules fused into large bullae, involving the intertriginous as well as the extensor areas, sparing the mucosa. Her body temperature was 103 degrees F. Laboratory workup was significant for neutrophilia with a white cell count > 10,000/mm3.

Source: Medline

4. Benign familial pemphigus (Hailey-Hailey disease) treated with oral cyclosporine A
[Italian] PEMFIGO FAMILIARE BENIGNO (MALATTIA DI HAILEY-HAILEY) TRATTATO CON CICLOSPORINA A ORALE

Author(s) Cecchi R., Bartoli L., Brunetti L., Pavesi M., Giomi A.

Citation: Giornale Italiano di Dermatologia e Venereologa, 1993, vol./is. 128/11(615-617), 0026-4741 (1993)

Publication Date: 1993

Abstract: Benign familial pemphigus (Hailey-Hailey disease) is a rare dominantly inherited dermatosis characterized by a not well defined breakdown of the desmosomkeratin filament complexes, resulting in a widespread suprabasal keratinocyte acantholysis. A variety of treatments have been recommended for recalcitrant cases, ie topical and systemic steroids, antibiotics, dapsone, calcitriol, PUVA or methotrexate, often with transient and unsatisfactory results. A 50-year-old woman, affected with benign familial pemphigus for 30 years, was successfully treated with oral cyclosporine A at an initial dose of 4 mg/kg/day for 6 months, during an exacerbation of the disease. The authors discuss a possible mode of action of cyclosporine in this condition, on the basis of literature data.

Source: EMBASE


Author(s) Persic-Vojinovic S, Milavec-Puretiv V, Dobric I, Rados J, Spoljar S

Citation: Acta Dermatovenerologica Croatica, 2006, vol./is. 14/4(253-7), 1330-027X;1330-027X (2006)

Publication Date: 2006

Abstract: Hailey-Hailey disease is a rare autosomal dominant skin disorder that typically affects the intertriginous areas. The responsible defect has been identified in the gene named ATP2C1 on chromosome 3q21-24. We present a 50-year-old man with a 16-year history of blistering eruptions and positive familial history where this disease had appeared through four generations. The diagnosis was confirmed by histopathologic studies and negative immunofluorescence findings. A combination of topical tacrolimus therapy and oral erythromycin seemed to play a considerable part in this case, in which all of the lesions healed within 2 weeks.

Source: Medline

6. Hailey-Hailey disease

Author(s) Griffiths C.E.M.

Citation: Journal of Dermatological Treatment, 2000, vol./is. 11/4(217), 0954-6634 (2000)

Publication Date: 2000

Source: EMBASE

Available in fulltext from Journal of Dermatological Treatment at EBSCOhost

7. Hailey-Hailey disease exacerbated by multiple pregnancies: Case report and review of
8. Hailey-Hailey disease: The clinical features, response to treatment and prognosis

Author(s) Burge S.M.

Citation: British Journal of Dermatology, 1992, vol./is. 126/3(275-282), 0007-0963 (1992)

Abstract: Fifty-eight individuals with Hailey-Hailey disease were studied to delineate the clinical features, response to treatment and prognosis. The disease generally presented between the second and fourth decades, but the morphology of lesions was varied and a delay in diagnosis was common. Nail changes have not been documented in previous studies of Hailey-Hailey disease, but asymptomatic longitudinal white bands were present in the fingernails in 71% of 38 patients examined and are a helpful physical sign. The disease is predominantly flexural. Friction and heat or sweating exacerbate the lesions and pain may limit physical activities. The prognosis was assessed in 27 patients with longstanding disease and the long-term outlook is generally good. Seventeen patients had improved and the disease was static in seven patients. Three patients deteriorated with age. Topical corticosteroids with or without added antibiotics were an effective treatment.

Source: EMBASE

Available in fulltext from British Journal of Dermatology at EBSCOhost

Available in fulltext from British Journal of Dermatology at Directory of Open Access Journals

9. Successful management of Hailey-Hailey disease with potent topical steroid ointment

Author(s) Ikeda S., Suga Y., Ogawa H.

Citation: Journal of Dermatological Science, 1993, vol./is. 5/3(205-211), 0923-1811 (1993)

Abstract: In order to know the real effect of steroid for Hailey-Hailey disease, seven patients were treated with strong steroid ointments in conjunction with antibiotics and/or antmyotic ointments. All patients were infected by various kind of bacteria and fungus. All patients showed remission within 2-16 weeks. Four patients maintained remission from 1.5 to over 4 years when ointments were continuously applied. No serious side-effects except skin atrophy and contact dermatitis were observed. These findings suggest that strong steroid ointment was really effective for this disease when secondary infection was prevented by the application of antibiotics and/or antmyotics, and that this disease is controllable with strong steroid ointments.
10. Treatment of Hailey-Hailey disease with oral erythromycin

**Author(s)** Nasca M.R., De Pasquale R., Amodeo S., Fazio A., Tedeschi A., Micali G.

**Citation:** Journal of Dermatological Treatment, 2000, vol./is. 11/4(273-277), 0954-6634 (2000)

**Publication Date:** 2000

**Abstract:** Hailey-Hailey disease is a rare autosomal dominant acantholytic disorder characterized from late adolescence or adulthood by recurrent eruptions of vesicles and blisters usually located on the neck, axillae and groin. The clinical cases of four unrelated adult male patients with Hailey-Hailey disease are presented. In all patients, a relatively short-course treatment with oral erythromycin (3-4 weeks) induced a long-lasting remission (8 months). Since bacteriological investigations excluded local infection, other hypothetical pharmacological effects (anti-inflammatory action and inhibition of cytokine release), besides the antibacterial properties of erythromycin, should be considered to explain such clinical improvement. However, until precise information about the pathogenesis of Hailey-Hailey disease is available, the mechanism of action of erythromycin in this disease remains speculative. Further studies in larger series of patients are needed in order to assess the role of erythromycin, and perhaps other macrolides, as a standard therapeutic option for Hailey-Hailey disease. (J Dermatol Treat (2000) 11: 273-277).

**Source:** EMBASE

Available in fulltext from Journal of Dermatological Treatment at EBSCOhost

**Other antibiotics**

1. Hailey-Hailey disease with skin lesions at unusual sites and a good response to acitretin

**Author(s)** Vasudevan B., Verma R., Badwal S., Neema S., Mitra D., Sethumadhavan T.

**Citation:** Indian Journal of Dermatology, Venereology and Leprology, January 2015, vol./is. 81/1(88-91), 0378-6323;0973-3922 (01 Jan 2015)

**Publication Date:** January 2015

**Source:** EMBASE

Available in fulltext from Indian Journal of Dermatology, Venereology & Leprology at EBSCOhost

Available in fulltext from Indian Journal of Dermatology, Venereology & Leprology at EBSCOhost

Available in fulltext from Indian Journal of Dermatology, Venereology and Leprology at Free Access Content

Available in fulltext from Indian Journal of Dermatology, Venereology and Leprology at Directory of Open Access Journals

2. Familial benign chronic pemphigus and doxycycline: a review of 6 cases.

**Author(s)** Le Sache-de Peufeilhoux L, Raynaud E, Bouchardeau A, Fraitag S, Bodemer C

**Citation:** Journal of the European Academy of Dermatology & Venereology, March 2014, vol./is. 28/3(370-3), 0926-9959;1468-3083 (2014 Mar)

**Publication Date:** March 2014

**Source:** Medline

3. Hailey-hailey disease responding to thalidomide

**Author(s)** Nanda K., Saldanha C., Jacintha M., Kamath G.

**Citation:** Indian Journal of Dermatology, March 2014, vol./is. 59/2(190-192), 0019-

**Author(s)** Vanderbeck K.A., Giroux L., Murugan N.J., Karbowski L.M.

**Citation:** Dermatology Reports, 2014, vol./is. 6/1(21-23), 2036-7392;2036-7406 (2014)

**Publication Date:** 2014

**Source:** EMBASE

Available in fulltext from Dermatology Reports at National Library of Medicine
Available in fulltext from Dermatology Reports at National Library of Medicine
Available in fulltext from Dermatology Reports at Free Access Content
Available in fulltext from Dermatology Reports at Directory of Open Access Journals

5. Successful therapy of refractory Hailey-Hailey disease with oral alitretinoin

**Author(s)** Sardy M., Ruzicka T.

**Citation:** British Journal of Dermatology, January 2014, vol./is. 170/1(209-211), 0007-0963;1365-2133 (January 2014)

**Publication Date:** January 2014

**Source:** EMBASE

Available in fulltext from British Journal of Dermatology at EBSCOhost
Available in fulltext from British Journal of Dermatology at EBSCOhost


**Author(s)** Higashimae K, Park K, Kabashima K, Tanabe H

**Citation:** European Journal of Dermatology, April 2013, vol./is. 23/2(265-6), 1167-1122:1952-4013 (2013 Apr 1)

**Publication Date:** April 2013

**Source:** Medline

Available in fulltext from European Journal of Dermatology at Free Access Content

7. Oral terbinafine as an alternative treatment in Hailey-Hailey disease
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12. Hailey-Hailey disease

Author(s)

Citation: Consultant, October 2004, vol./is. 44/12(1591), 0010-7069 (October 2004)
Publication Date: October 2004
Source: EMBASE

13. Hailey-Hailey Disease

Author(s)

Citation: Consultant, December 2003, vol./is. 43/14(1744), 0010-7069 (December 2003)
Publication Date: December 2003
Source: EMBASE


Author(s) Ikeda S, Suga Y, Ogawa H

Citation: Journal of Dermatological Science, June 1993, vol./is. 5/3(205-11), 0923-1811;0923-1811 (1993 Jun)
Publication Date: June 1993
Source: Medline


Author(s) Burge SM

Citation: British Journal of Dermatology, March 1992, vol./is. 126/3(275-82), 0007-0963;0007-0963 (1992 Mar)
Publication Date: March 1992
Source: Medline

16. Chronic benign familial pemphigus.

Author(s) Galimberti RL, Kowalczyk AM, Bianchi O, Bonino MV, Garcia Garcia A

Citation: International Journal of Dermatology, September 1988, vol./is. 27/7(495-500), 0011-9059:0011-9059 (1988 Sep)
Publication Date: September 1988
Source: Medline

Available in fulltext from International Journal of Dermatology at EBSCOhost
Available in fulltext from International Journal of Dermatology at EBSCOhost
Treatment of Hailey–Hailey disease with oral erythromycin


Hailey-Hailey disease is a rare autosomal dominant acantholytic disorder characterized from late adolescence or adulthood by recurrent eruptions of vesicles and blisters usually located on the neck, axillae and groin. The clinical cases of four unrelated adult male ...

Commentary: Hailey-Hailey Disease: Familial Benign Chronic Pemphigus

B Michel - Archives of Dermatology, 1982 - archderm.jamanetwork.com

... Topical tetracycline, erythromycin, and nystatin are also in widespread use. ... Tap water compresses used in conjunction with topical steroids and antibiotics can also be ... dapsone23 (diaminodiphenylsulfone) in other bullous diseases has prompted its use in Hailey-Hailey disease ...

Hailey-Hailey disease of the vulva

JS Wieselthier, SH Pincus - Archives of dermatology, 1993 - archderm.jamanetwork.com

... Disorder Clinical Presentation Pathognomonic Histopathologic Features Hailey-Hailey disease ... Positive family history of HH was elicited in all three women. Conservative treatment with topical antibiotics was effective in all of our cases. ...

Hailey-hailey disease and review of management.


... the most benefit with addition of oral antibiotics, excisional procedures and botulinum toxin A. Other therapies are described but with much less supporting evidence.

CONCLUSIONS: Herein we review the literature to identify successful treatments for Hailey-Hailey disease. ...

Darier’s Disease and Hailey-Hailey Disease

LA Goldsmith, E Epstein Jr - Principles of Molecular Medicine, 1998 - Springer

... below), MANAGEMENT/TREATMENT Standard therapy for Hailey-Hailey disease includes antibiotics-both systemic and local-and topical steroids. Responses are inconsistent, but fre-quent enough to warrant their use. Stronger ...

Familial benign pemphigus: a trichophytosis form

JM Chevallier - Journal of the European Academy of …, 1995 - ingentaconnect.com

... Abstract: Familial benign pemphigus is a rare genodermatosis. We report a case of atypical circinate aspect with erythematousquamous edges and tendancy to central cicatrisation. Our patient responded to treatment with tropical and systemic antibiotics. Document Type: Abstract. ...

Published Research – Database Search Strategy

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