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**Literature search results**

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<tr>
<td>Search completed by:</td>
<td>Richard Bridgen</td>
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**Search details**

Reports on histiocytic variant of Sweet’s Syndrome and Anakinra

**Resources searched**

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

**Database search terms:** (sweet OR sweet's) adj0 syndrome*; ACUTE FEBRILE NEUTROPHILIC DERMATOSIS; “acute febrile neutrophilic dermatos*”; “neutrophilic dermatoses”; (histiocytic OR histiocytoid) adj3 variant*; (histiocytic OR histiocytoid) adj5 variant*; HISTIOCYTE; “tissue monocyte*”; anakinra; histiocyt*; macrophage*; anakinra; exp INTERLEUKIN 1 RECEPTOR ANTAGONIST PROTEIN; "interleukin-1 receptor antagonist*”; "interleukin (IL)-1 antagonist*”; “anti-interleukin-1 receptor antagonist*”; kineret; “recombinant interleukin 1 receptor antagonist*”; recombinant interleukin 1 receptor blocker*”; RECOMBINANT INTERLEUKIN 1 RECEPTOR BLOCKING AGENT

**Evidence search string(s):** (“sweet syndrome” OR "sweets syndrome" OR "sweet's syndrome" OR "acute febrile neutrophilic dermatos*” OR "neutrophilic dermatoses") (anakinra OR kineret OR "interleukin 1 receptor antagonist*” OR "interleukin-1 receptor antagonist*”)

**Google search string(s):** (~"sweet syndrome" OR ~"acute febrile neutrophilic dermatosis" OR ~"neutrophilic dermatosis") (anakinra OR kineret OR ~"interleukin 1 receptor antagonists")

~"histiocytic variant" (~"sweet syndrome" OR ~"acute febrile neutrophilic dermatosis" OR ~"neutrophilic dermatoses") (anakinra OR kineret OR ~"interleukin 1 receptor antagonists")
**Summary**

There is no research on anakrina and histiocytic variations of sweet’s syndrome. I have included research on interleukin-1 receptor antagonist and sweet’s syndrome in case you find them useful.

**Guidelines**

None found.

**Evidence-based reviews**

None found.

<table>
<thead>
<tr>
<th>Published research</th>
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| 1. **[Indications and modes of use for interleukin (IL)-1 antagonists in inflammatory dermatosis: a new therapeutic approach to immune-mediated inflammatory diseases].** [French] Indications et modalités d’utilisation des antagonistes de l’interleukine (IL)-1 dans les dermatooses inflammatoires.  
**Author(s)** Lipsker D, Lenormand C  
**Citation:** Annales de Dermatologie et de Venereologie, June 2012, vol./is. 139/6-7(459-67), 0151-9638;0151-9638 (2012 Jun)  
**Publication Date:** June 2012  
**Abstract:** BACKGROUND: IL-1 antagonists are used in the treatment of patients with rheumatoid arthritis and cryopyrinopathies. As yet anecdotal observations suggest that they may allow effective treatment of patients with different types of inflammatory skin disease. This review focuses on our current knowledge of the use of IL-1 antagonists in dermatology.PATIENTS AND METHODS: A Medline search was performed combining the keywords: “anakinra; canakinumab; rilonacept” AND “skin; neutrophilic dermatoses; Sweet syndrome; pyoderma gangrenosum; hidradenitis suppurativa; Schnitzler syndrome; Still disease”. The precise dermatological phenotype of patients with IL-1 antagonist-responsive auto-inflammatory disorders was analysed in order to compare it to related complex disorders.RESULTS: Double-blind randomized controlled trials have demonstrated the efficacy of these treatments in cryopyrinopathies with dermatological involvement including chronic infantile neurological cutaneous and articular (CINCA) syndrome, Muckle-Wells syndrome and familial cold urticaria. Anakinra is the only treatment for Schnitzler syndrome that is almost constantly efficacious, even in refractory disease, as attested by numerous case reports. It is also efficacious in the treatment of patients with adult-onset Still disease and systemic juvenile arthritis. Neutrophilic dermatoses constitute the cutaneous hallmark of IL-1-responsive auto-inflammatory disorders, and neutrophilic dermatoses could thus form an indication for this treatment. However, to date, only 9 reports have been published showing efficiency in patients with Sweet syndrome, in one case of neutrophilic panniculitis, and in two cases of pustular psoriasis. Anakinra appears less efficacious in patients with pyoderma gangrenosum.CONCLUSION: IL-1 antagonists are a first-line treatment in patients with Schnitzler syndrome and cryopyrinopathies. They could become important alternatives in patients with acute and febrile neutrophilic dermatoses either unresponsive to or with contraindications to conventional treatments, but this requires confirmation by further clinical trials. Copyright 2012 Elsevier Masson SAS. All rights reserved.  
**Source:** Medline |
| 2. **Efficacy of anakinra, an IL1 receptor antagonist, in refractory Sweet syndrome.**  
**Author(s)** Delluc A, Limal N, Puechal X, Frances C, Piette JC, Cacoub P  
**Citation:** Annals of the Rheumatic Diseases, February 2008, vol./is. 67/2(278-9), 0003- |
3. An overview of interleukin-1 receptor Antagonist, Anakinra, in the treatment of cutaneous diseases

Author(s) Pazyar N., Feily A., Yaghoobi R.

Citation: Current Clinical Pharmacology, 2012, vol./is. 7/4(271-275), 1574-8847 (2012)

Abstract: Interleukin (IL)-1 is a pivotal proinflammatory cytokine consisting of two molecular species, IL-1alpha and IL-1beta. Anakinra (Kineret), a recombinant human IL-1 receptor antagonist, is regarded as a biological agent which blocks the inflammatory effects of IL-1. The aim of this review was to search the literatures and summarizes in vivo, in vitro and human studies on anakinra uses in dermatological disorders. The results show that anakinra is currently used clinically for the treatment of a variety of skin conditions such as psoriasis, atopic dermatitis, photoaging, melanoma, Schnitzler syndrome, pyoderma gangraenosum, PAPA syndrome, hidradenitis suppurativa, lamellar ichthyosis, Sweet's syndrome, panniculitis, Muckle-Wells syndrome, familial Mediterranean fever, SAPHO syndrome and other disorders. Notably, anakinra is expensive to produce and administer. Injection is the route of therapy and allergic reaction is most possible. 2012 Bentham Science Publishers.

Source: EMBASE

4. Indications and modes of use for interleukin (IL)-1 antagonists in inflammatory dermatosis: A new therapeutic approach to immune-mediated inflammatory diseases [French] Indications et modalités d’utilisation des antagonistes de l’interleukine (IL)-1 dans les dermatoses inflammatoires

Author(s) Lipsker D., Lenormand C.

Citation: Annales de Dermatologie et de Venereologie, June 2012, vol./is. 139/6-7(459-467), 0151-9638 (June 2012)

Abstract: Background: IL-1 antagonists are used in the treatment of patients with rheumatoid arthritis and cryopyrinopathies. As yet anecdotal observations suggest that they may allow effective treatment of patients with different types of inflammatory skin disease. This review focuses on our current knowledge of the use of IL-1 antagonists in dermatology. Patients and methods: A Medline search was performed combining the keywords: “anakinra; canakinumab; rilonacept” AND “skin; neutrophilic dermatoses; Sweet syndrome; pyoderma gangrenosum; hidradenitis suppurativa; Schnitzler syndrome; Still disease”. The precise dermatological phenotype of patients with IL-1 antagonist-responsive auto-inflammatory disorders was analysed in order to compare it to related complex disorders. Results: Double-blind randomized controlled trials have demonstrated the efficacy of these treatments in cryopyrinopathies with dermatological involvement including chronic infantile neurological cutaneous and articular (CINCA) syndrome, Muckle-Wells syndrome and familial cold urticaria. Anakinra is the only treatment for Schnitzler syndrome that is almost constantly efficacious, even in refractory disease, as attested by numerous case reports. It is also efficacious in the treatment of patients with adult-onset Still disease and systemic juvenile arthritis. Neutrophilic dermatoses constitute the cutaneous hallmark of IL-1-responsive auto-inflammatory disorders, and neutrophilic dermatoses could thus form an indication for this treatment. However, to date, only 9 reports have been published showing efficiency in patients with Sweet syndrome, in one case of neutrophilic panniculitis,
and in two cases of pustular psoriasis. Anakinra appears less efficacious in patients with pyoderma gangrenosum. Conclusion: IL-1 antagonists are a first-line treatment in patients with Schnitzler syndrome and cryopyrinopathies. They could become important alternatives in patients with acute and febrile neutrophilic dermatoses either unresponsive to or with contraindications to conventional treatments, but this requires confirmation by further clinical trials. 2012 Elsevier Masson SAS. All rights reserved.

Source: EMBASE

5. Efficacy of anti-interleukin-1 receptor antagonist anakinra (Kineret) in a case of refractory sweet's syndrome

Author(s) Kluger N., Gil-Bistes D., Guillot B., Bessis D.

Citation: Dermatology, May 2011, vol./is. 222/2(123-127), 1018-8665 (May 2011)

Publication Date: May 2011

Abstract: Sweet's syndrome is a neutrophilic dermatosis characterized by fever, an elevated neutrophil count, and painful erythematous cutaneous lesions. Histopathological analysis reveals a neutrophilic dermal infiltrate. Systemic corticosteroid therapy remains the mainstay of treatment. We report the case of a 66-year-old male patient who had a 5-year history of Sweet's syndrome refractory to various conventional treatments. Anti-interleukin-1 receptor antagonist anakinra was initiated and this resulted in a dramatic clinical and biological improvement. Anakinra is a promising treatment for neutrophilic dermatoses and sheds light on the interleukin-1/inflammasome pathway as central in the pathophysiology of neutrophilic dermatosis. Copyright 2011 S. Karger AG, Basel.

Source: EMBASE

Available in full text from Dermatology at EBSCOhost


Author(s) Cohen P.R.

Citation: American Journal of Clinical Dermatology, 2009, vol./is. 10/5(301-312), 1175-0561;1179-1888 (2009)

Publication Date: 2009

Abstract: Sweet syndrome, pyoderma gangrenosum, and subcorneal pustular dermatosis are neutrophilic dermatoses conditions that have an inflammatory infiltrate consisting of mature polymorphonuclear leukocytes. The neutrophils are usually located within the dermis in Sweet syndrome and pyoderma gangrenosum; however, in subcorneal pustular dermatosis, they are found in the upper layers of the epidermis. Sweet syndrome, also referred to as acute febrile neutrophilic dermatosis, is characterized by pyrexia, elevated neutrophil count, painful erythematous cutaneous lesions that have an infiltrate of mature neutrophils typically located in the upper dermis, and prompt clinical improvement following the initiation of systemic corticosteroid therapy. Classical, malignancy-associated, and drug-induced variants of Sweet syndrome exist. Pyoderma gangrenosum is characterized by painful, enlarging necrotic ulcers with bluish undermined borders surrounded by advancing zones of erythema; its clinical variants include: ulcerative or classic, pustular, bullous or atypical, vegetative, peristomal, and drug-induced. Subcorneal pustular dermatosis is an uncommon relapsing symmetric pustular eruption that involves flexural and intertriginous areas; it can be idiopathic or associated with cancer, infections, medications, and systemic diseases. Since Sweet syndrome, pyoderma gangrenosum, and subcorneal pustular dermatosis share not only the same inflammatory cell but also similar associated systemic diseases, it is not surprising that the concurrent or sequential development of these neutrophilic dermatoses has been observed in the same individual. Also, it is not unexpected that several of the effective therapeutic interventions including systemic drugs, topical agents, and other treatment modalities for the management of these dermatoses are the same. The treatment of choice for Sweet syndrome and idiopathic pyoderma gangrenosum is systemic corticosteroids; however, for subcorneal pustular dermatosis, dapsone is the drug of choice. Yet, tumor necrosis factor-alpha antagonists are becoming the preferred choice when pyoderma gangrenosum is accompanied by
inflammatory bowel disease or rheumatoid arthritis. Potassium iodide and colchicine are alternative first-line therapies for Sweet syndrome and indomethacin (indometacin), clofazimine, cyclosporine (ciclosporin), and dapsone are second-line treatments. Cyclosporine is effective in the acute management of pyoderma gangrenosum; however, when tapering the drug, additional systemic agents are necessary for maintaining the clinical response. In some patients with subcorneal pustular dermatosis, systemic corticosteroids may be effective; yet, systemic retinoids (such as etretinate and acitretin) have effectively been used for treating this neutrophilic dermatosis either as monotherapy or in combination with dapsone or as a component of phototherapy with psoralen and UVA radiation. Topical agents can have an adjuvant role in the management of these neutrophilic dermatoses; however, high-potency topical corticosteroids may successfully treat localized manifestations of Sweet syndrome, pyoderma gangrenosum, and subcorneal pustular dermatosis. Intralesional corticosteroid therapy for patients with Sweet syndrome and pyoderma gangrenosum, hyperbaric oxygen and plasmapheresis for patients with pyoderma grangrenosum, and phototherapy for patients with subcorneal pustular dermatosis are other modalities that have been used effectively for treating individuals with these neutrophilic dermatoses.

Source: EMBASE

Available in fulltext from American Journal of Clinical Dermatology at EBSCOhost

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**Google Scholar**

*From 1st 50 results…*

**Phenotype, genotype, and sustained response to anakinra in 22 patients with autoinflammatory disease associated with CIAS-1/NALP3 mutations**
KS Leslie, HJ Lachmann, E Bruning… - Archives of ..., 2006 - Am Med Assoc
... The development of Sweet syndrome in 1 patient and the development of pyoderma ... data, February 2006) places these conditions in the spectrum of neutrophilic dermatoses, in which there ... complete, and sustained response to treatment with small dosages of anakinra and the ... Cited by 90 Related articles BL Direct All 13 versions Cite More

**Acitretin-and tumor necrosis factor inhibitor-resistant acrodermatitis continua of Hallopeau responsive to the interleukin 1 receptor antagonist anakinra**
V Lutz, D Lipsker - Archives of dermatology, 2012 - Am Med Assoc
... An interesting fact is that neutrophilic dermatoses are present in most patients affected by one of those disorders.2,8 ... 4. Kluger N, Gil-Bistes D, Guillot B, Bessis D. Efficacy of anti-interleukin-1 receptor antagonist anakinra (Kineret) in a case of refractory Sweet's syndrome. ... Cited by 7 Related articles All 13 versions Cite More

**Acute febrile neutrophilic dermatosis (Sweet's syndrome)**
CL Anzalone, PR Cohen - Current opinion in hematology, 2013 - journals.lww.com
... Skip Navigation Links Home > Current Issue > Acute febrile neutrophilic dermatosis (Sweet's syndrome) Related articles All 3 versions Cite

**Neutrophilic Dermatoses**
PR Cohen - American journal of clinical dermatology, 2009 - Springer
... women as an acute febrile neutrophilic dermatosis in 1964.[3] The descriptive nomenclature for the condition emphasized its salient features ... Systemic treatments for neutrophilic dermatoses Agent Sweet syndrome Pyoderma gangrenosum Subcorneal pustular dermatosis ... Cited by 46 Related articles All 7 versions Cite

**Management of neutrophilic dermatoses**
CR Schadt, JP Callen - Dermatologic Therapy, 2012 - Wiley Online Library
... It was originally named acute febrile neutrophilic dermatosis by Sweet in 1964 in his
sentinel description of ... myeloperoxidase and chemotaxis and has demonstrated some efficacy in the treatment of **neutrophilic dermatoses**. ... Table 1. Anecdotal treatments of **Sweet’s syndrome**. ...

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**Interleukin-1, inflammasomes, autoinflammation and the skin**

E Contassot, HD Beer, LE French - Swiss Med Wkly, 2012 - smw.ch


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**The emerging role of interleukin-1β in autoinflammatory diseases**

HJ Lachmann, P Quartier, A So... - Arthritis & ..., 2011 - Wiley Online Library

... Rapid and sustained responses to **anakinra** have been reported for small series or single patients with Schnitzler syndrome, **Sweet’s syndrome**, Behçet’s disease, or relapsing polychondritis, many of whom had disease refractory to immunosuppressants, including steroids (see ... 

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**Acute febrile neutrophilic dermatosis (Sweet syndrome)**

PR Cohen, H Honigsmann... - … in general medicine. ..., 2008 - fitzpatricksdermatology.com


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