ORIGINAL ARTICLE

Current therapeutic options for adult patients with urticarial vasculitis: A scoping review

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Background: Urticarial vasculitis (UV) is a rare form of small vessel vasculitis, and there are limited published data on its management.

Objective: This study aims to review the current therapeutic options for UV.

Methods: A PubMed search was conducted, selecting articles published from 2000 to January 2024.

Results: Of 305 identified articles, 21 were included. Mild cutaneous UV can be treated with antihistamines and nonsteroidal anti-inflammatory drugs. For intermittent cutaneous UV, short courses of systemic corticosteroids are recommended. Hydroxychloroquine, colchicine, and dapsone show comparable efficacy to corticosteroids and are often used for refractory and hypocomplementemic UV patients. In cases with persistent symptoms, first-line immunosuppressants such as azathioprine, cyclophosphamide, cyclosporine A, methotrexate, and mycophenolate mofetil may be considered. Some studies suggest the effectiveness of omalizumab, rituximab, canakinumab, anakinra, and plasmapheresis.

Limitations: Only noninterventional observational studies, which were mostly retrospective, were found and included in our scoping review. Furthermore, the study is limited by small sample sizes due to the nature of UV.

Conclusion: UV is a rare condition with insufficient treatment data. This scoping review outlines potential treatment options, highlighting the need for further research. (J Am Acad Dermatol https://doi.org/10.1016/j.jaad.2025.03.056.)

Key words: urticarial vasculitis; urticarial vasculitis syndrome; urticarial vasculitis treatment.

BACKGROUND

Urticarial vasculitis (UV) is a rare small vessel vasculitis characterized by urticarial wheals that persist more than 24 hours and a leukocytoclastic vasculitis on histopathology.[1] Angioedema, purpura, pain, and residual hyperpigmentation may be observed. UV may be divided into 2 categories: normocomplementemic UV and hypocomplementemic UV (HUV). The latter is subdivided into HUV and HUV syndrome. The HUV category is more often associated with systemic symptoms and an underlying disease, such as malignancy, infections, and autoimmune and autoinflammatory diseases.[1] However, many UV cases are idiopathic. Possible systemic symptoms include fever; asthenia; abdominal pain; arthralgia; lymphadenopathy; and ocular, renal, and lung manifestations. Rarely, UV may involve the ear, nose, pericardium, and central nervous system. UV predominantly affects females, and the median age of onset is 45 years. Its prevalence is

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unknown. The pathophysiology of UV remains unclear, but it is believed to be driven by immune-complex–mediated inflammation. There is a paucity of published studies on the management of UV. There are no approved therapies or official clinical guidelines for UV. Clinicians are faced with therapeutic uncertainties when treating these patients. The objective of this scoping review is to synthesize current treatment options in UV.

CAPSULE SUMMARY

- The manuscript reviews treatments for urticarial vasculitis, highlighting antihistamines, corticosteroids, immunosuppressants, and biologics.
- It provides a crucial therapeutic framework for dermatologists to navigate this rare condition, addressing the absence of guidelines and emphasizing tailored approaches to optimize outcomes and minimize medication side effects.

METHODS

The population, intervention, comparison, and outcome framework was applied to structure the research question: in adult patients with UV or UV syndrome, what treatments currently exist to induce clinical remission (or relapse free survival) while limiting adverse events.

We performed a PubMed search on December 29, 2023. The terms used for our literature search were "Urticaria" AND "Vasculitis" AND "Treatment". We included published randomized and nonrandomized trials, observational studies, case series, and systematic reviews. Case reports were excluded to reduce the risk of publication bias. We restricted our search to articles published in English or French, from 2000 to 2024.

When available, extracted data included study details (author, year, and country), patient characteristics (age, sex, UV type, extra-cutaneous involvement, underlying disease, and biopsy results), treatment parameters (options, duration, initial dose, and number of doses), outcomes (time to relapse/remission, treatment efficacy, and disease activity scores), and relapse occurrence. Clinical response was collected as defined by each study, or as complete (complete response) or partial resolution (partial response) of symptoms. Relapses were defined as the occurrence of active disease after a period of remission.

We did not plan "a priori" quantitative analyses with summary estimates since we expected important clinical heterogeneity within included records. Thus, data were summarized qualitatively in the form of text, tables, and figures.

RESULTS

Of the 305 studies screened, 21 records were included in our review.

Mild isolated cutaneous urticarial vasculitis

Eleven studies focused on therapies for mild UV, which is characterized by mild isolated cutaneous symptoms. In these cases, symptomatic treatment with nonsteroidal anti-inflammatory drugs or antihistamines at standard doses has proven effective.[1-6] In a retrospective study involving 47 patients, 46 experienced partial or temporary relief with hydroxyzine or desloratadine after 6 months.[7] Another study involving 8 UV patients found that 7 achieved complete relief when antihistamines were combined with reserpine, with no significant side effects reported.[8]

Intermittent cutaneous urticarial vasculitis

Ten studies examined therapies for intermittent, relapsing UV. Relapses were defined as episodes of active cutaneous lesions occurring after at least 1 month of complete remission.[4] For intermittent disease, brief courses of systemic oral corticosteroids are commonly prescribed. The corticosteroid doses reported in the literature vary widely, ranging from 5 mg to 80 mg per day.[4,9] No studies have compared the safety and efficacy of different doses of systemic oral corticosteroids for UV. Corticosteroids are usually prescribed for 5 to 10 days for treatment of flares.

In contrast to mild UV, a retrospective study found that 39 of 47 patients with intermittent, relapsing UV were refractory to antihistamines.[7] These patients were subsequently treated with cinnarizine (not available in the United States or Canada) or a combination of prednisolone (0.2-0.5 mg/kg/day) and an antihistamine for 4-6 months. Among those treated with the combination of antihistamine and prednisolone, 13 of 20 patients achieved a complete response. Furthermore, those who attained a complete response maintained sustained remission during follow-up, with a median duration of 4 years.[7]

Refractory urticarial vasculitis

Refractory cases of UV are characterized by persistent symptoms despite treatment with corticosteroids. In such instances, treatment options include hydroxychloroquine, colchicine, or dapsone.[2,4,10,11] If symptoms persist, first-line immunosuppressants such as azathioprine, cyclophosphamide, cyclosporine A, or mycophenolate mofetil may be indicated (see

Abbreviations used:

anti-TNF:	anti-tumor necrosis factor
HUV:	hypocomplementemic urticarial vasculitis
IgE:	immunoglobulin E
IL:	interleukin
UV:	urticarial vasculitis

Table I for suggested doses).[4,5,11] A retrospective study involving 4 patients with corticosteroid-dependent (requiring prednisolone doses of 10-60 mg/day) normocomplementic UV assessed the efficacy of methotrexate.[12] In this study, 1 patient achieved complete remission, 2 experienced a partial response that allowed for a reduction in steroid dosage, and 1 showed no benefit from 25 mg of methotrexate weekly.[12] Additionally, rituximab has emerged as a promising alternative, with several case reports indicating its potential efficacy.[4,13]

Omalizumab is currently an established treatment for chronic idiopathic urticaria.[14] Chen et al conducted a prospective, single-center, open-label study involving 23 patients with normocomplementemic UV treated with 3 doses of omalizumab 300 mg subcutaneously.[15] Among these patients, 17 experienced an improvement in disease activity, resulting in a response rate of 73%. Notably, the baseline immunoglobulin E (IgE) levels of the 6 nonresponding patients were lower than those of the responders, and these patients exhibited more systemic manifestations. The treatment was well tolerated, with no significant side effects reported. However, a 100% relapse rate was observed upon discontinuation of the medication.[15] Additionally, co-treatment with methotrexate has demonstrated effectiveness.[16]

Interleukin (IL) 1 is thought to play a crucial role in UV and other causes of leukocytoclastic vasculitis.[10,17] An open-label study treated 10 patients with UV with canakinumab, an anti-IL-1 monoclonal antibody. Among these patients, 7 demonstrated a reduction in their Urticarial Vasculitis Activity Score after receiving a single dose, indicating a 50% improvement in disease activity.[18] A multicentric retrospective study in France evaluated the use of anti-IL-1 in refractory UV patients with exclusive cutaneous, articular, or gastrointestinal involvement. Among these patients, 5 of 6 patients achieved a complete clinical response.[13]

Currently, there are no studies assessing the efficacy of anti-IL-6 and anti tumor necrosis factor (anti-TNF) therapies, including etanercept, infliximab, and tocilizumab, although their use has been successfully reported in UV.

Hypocomplementemic urticarial vasculitis

In patients with HUV, a median of 3 treatment attempts is typically required before achieving a response.[9] These patients often have systemic symptoms and may have an underlying systemic disease.[5,9,19] Effective therapies for this population include systemic corticosteroids, hydroxychloroquine, colchicine, immunosuppressants, and rituximab. Corticosteroid doses reported in the literature vary significantly, ranging from 5 mg to 80 mg per day.[4,9] Notably, the efficacy of both high (0.5-1 mg/kg/day) and low doses (less than 0.5 mg/kg/day) of corticosteroids has been found to be comparable.[9]

In a study involving 57 patients with HUV, treatments included prednisone (57%), hydroxychloroquine (46%), and colchicine (14%).[9] The response rates were comparable across the treatments: hydroxychloroquine showed a 50% cutaneous response and a 48% immunologic response, colchicine exhibited a 43% cutaneous response and a 40% immunologic response, and prednisone had a 53% cutaneous response and a 32% immunologic response. Additionally, the study highlighted the efficacy of dapsone, which demonstrated a 100% cutaneous response rate but a 0% immunologic response.[9] The immunologic response is defined as the normalization of complement levels during treatment. It was also noted that anti-C1q antibodies do not influence treatment outcomes, and the time to treatment failure was not prolonged when using a steroid-sparing immunosuppressive agent.[9]

If patients do not show improvement with prednisone, hydroxychloroquine, colchicine, or dapsone, immunosuppressants are the next course of action. Effective immunosuppressive therapies include azathioprine, cyclophosphamide, cyclosporine A, methotrexate, and mycophenolate mofetil. In the study of 57 patients with HUV mentioned previously, azathioprine, cyclophosphamide, and mycophenolate mofetil exhibited similar times to treatment failure.[19] Another study involving 7 patients found that cyclophosphamide achieved complete cutaneous remission in 5 patients, a partial cutaneous response in 1, and no response in another.[9] However, due to its potential toxicity and side effects, cyclophosphamide is less frequently prescribed.[16] Cyclosporine A has been shown to be effective in treating HUV, including cases with lung involvement.[6,10,11] Additionally, patients with HUV who are refractory to cyclophosphamide may respond to cyclosporine A.[11] Notably, approximately one-third of patients experience relapse upon discontinuation of cyclosporine A.[6]

Rituximab successfully treated a patient with HUV and biopsy-proven kidney involvement.[20] In a study of 8 patients treated with rituximab, only 6 achieved

Table I. Classification of treatments for UV

Agents	Classes	Dosage
Indomethacin	Nonsteroidal anti-inflammatory drug	25 mg po tid
Hydroxyzine	Histamine H1 antagonist	25 mg po die
Desloratadine	Histamine H1 antagonist	5 mg po die
Prednisolone	Systemic corticosteroid	5-80 mg po die
Prednisone	Systemic corticosteroid	10-80 mg po die
Hydroxychloroquine	Antimalarial	400 mg po tid*
Colchicine	Antigout agent	0.6 mg po bid
Dapsone	Antibiotic	50-150 mg po die
Azathioprine	Immunosuppressant	Depends on TPMT levels*
Cyclophosphamide	Immunosuppressant	1-2 mg/kg/d po or 750-1000 mg/m^2 q mo IV
Cyclosporine A	Immunosuppressant	5 mg/kg/d po
Mycophenolate Mofetil	Immunosuppressant	2 g/d po*
Methotrexate	Immunosuppressant	7.5-25 mg q wk
Omalizumab	Monoclonal anti-IgE antibody	300 mg subcutaneous (wk 0, 4, and 8)
Rituximab	Monoclonal anti-CD-20 antibody	4 injections at 375 mg/m^2 or 2 injections of 1 g at 15 d interval
Canakinumab	Monoclonal anti-IL-1β antibody	300 mg subcutaneous
Anakinra	IL-1 receptor antagonist	10-30 mg subcutaneous die
Plasmapheresis	-	-

IgE, Immunoglobulin E; *IL*, interleukin; *IV*, intravenous; *PO Bie*, per os twice a day; *PO Die*, per so once daily; *PO Tid*, per os three times a day; *TPMT*, thiopurine methyl transferase; *UV*, urticarial vasculitis.
*Dose prescribed for cutaneous vasculitides.

a complete cutaneous response.[9] This study indicated that rituximab could sustain remission for longer periods compared to systemic corticosteroids and conventional immunosuppressants.[9,19] Several other studies have demonstrated rituximab's effectiveness in achieving complete remission of cutaneous symptoms, allowing for corticosteroid discontinuation.[17] In a French multicentric retrospective study, the use of IL-1β inhibitors was explored in 6 patients who were refractory to conventional immunosuppressive agents, rituximab, or omalizumab. However, patients with HUV did not achieve normalization of complement levels during treatment and experienced symptom relapses within days after discontinuation.[13]

Plasmapheresis may provide rapid but temporary symptom relief by removing immune complexes from the bloodstream.[4,11] It can be considered for patients who are refractory to all other therapies.[11] Successful outcomes with plasmapheresis have been reported in 2 HUV patients with biopsy-proven kidney involvement who did not respond to other treatments. Unfortunately, both patients eventually required kidney transplants due to progression to end-stage renal disease.[20] Currently, there are no studies supporting the use of intravenous immunoglobulin.

DISCUSSION

Our article reviews treatment options for UV and fills a critical gap in the literature by providing a structured therapeutic approach for physicians. In the absence of clinical guidelines, this stepwise framework can assist clinicians in optimizing UV treatment (Fig 1). Management should be individualized based on disease severity and clinical and biological presentation. The goal is to lessen chronic organ damage while minimizing medication side effects.[9,11,21] For patients with HUV, treatment can be customized according to complement levels. A rise in these levels often indicates an improvement in skin symptoms. Unfortunately, no serologic marker evaluates the response of other types of UV, making their treatment outcomes unpredictable.

If an underlying condition is present and contributes to the UV, the selected treatment should target that underlying disease.[16] For patients with underlying systemic lupus erythematosus, hydroxychloroquine and dapsone are viable treatment options.[11,16] Patients with hepatitis C should receive antiviral treatment based on the latest guidelines.

There is consensus between studies to try treatment with antihistamines[2,3] or nonsteroidal anti-inflammatory drugs for mild skin-limited diseases.[2] However, physicians must be aware that this is purely a symptomatic approach and does not address the underlying pathogenesis.[9,16,19]

Systemic corticosteroids are a primary treatment for UV and should be used to manage intermittent flare-ups of both cutaneous and extra-cutaneous disease. Studies show a wide range of doses, from 5 to 80 mg per day, with lower doses appearing

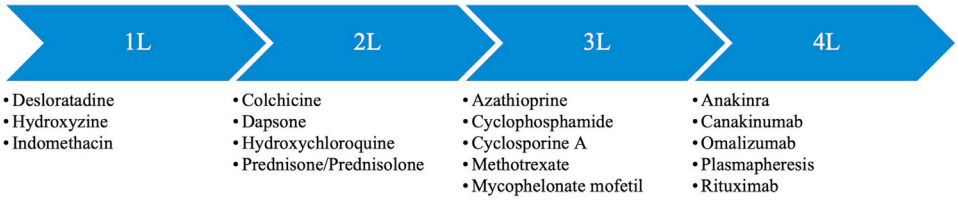

Fig 1. Suggested therapeutic ladder for treatment of UV; medications listed alphabetically. *UV*, Urticarial vasculitis.

equally effective for HUV.[9] It can be suggested that this principle may also apply to normocomplementemic UV. Flare-ups should be treated for 5 to 10 days. Risk of relapse upon corticosteroid withdrawal is independent of corticosteroid dose.[17] However, long-term use of systemic corticosteroids above supra-physiologic doses (dose greater than 7.5 mg/day) is at highest risk of adverse effects, including adrenal insufficiency, metabolic complications, infections, and fractures.[17,22] Thus, an initial dose ≤0.5 mg/kg/day is recommended.[9] The optimal dose and duration for corticosteroid treatment remain unclear.

For patients with refractory UV, corticosteroid-sparing treatments such as colchicine, hydroxychloroquine, or dapsone may be considered.[9,19] These medications are also regarded as first-line treatments for patients with HUV, who often experience more severe symptoms, systemic involvement, and have underlying conditions. Colchicine is an alkaloid that inhibits neutrophil chemotaxis, blocks lysosomal formation, and stabilizes lysosomal membranes.[11] Similarly, hydroxychloroquine prevents the release of lysosomal enzymes and IL-1.[11] The mechanism of action for dapsone, however, remains unclear.[11]

In patients with relapsing UV, treatment with an immunosuppressive agent is recommended. Initial therapy should involve systemic corticosteroids to achieve rapid symptom relief. Standard medications to consider include azathioprine, cyclophosphamide, cyclosporine A, methotrexate, and mycophenolate mofetil.[10] For relapsing HUV, immunosuppressants like mycophenolate mofetil are indicated.[11] While cyclophosphamide is less commonly used due to its side effect profile, it can still be considered. Cases with ocular and pulmonary manifestations are often the most difficult to treat, and cyclosporine A is a promising option.[9-11] Risk of relapse is significant upon discontinuing an immunosuppressant, and the optimal duration of treatment is not known.

Omalizumab, typically used for chronic idiopathic urticaria, may be an interesting option, particularly for normocomplementemic UV and refractory HUV. In some patients, autoreactive IgE could play a role in the pathogenesis of normocomplementemic UV, similar to chronic idiopathic urticaria. Consequently, patients with higher IgE levels may respond better to this treatment. However, the duration of treatment with omalizumab remains uncertain, as studies indicate that relapses can occur after stopping the medication. There are no studies assessing the long-term effects of omalizumab in this patient population.

There are few studies focused on patients who are refractory to standard immunosuppressive agents. However, rituximab, an anti-CD-20 monoclonal antibody, has shown success as a third-line treatment.[9] Patients with HUV tend to respond better to rituximab than to second-line therapies and experience a longer time until treatment failure. While the exact mechanism of action is not fully understood, rituximab has been effective in treating other immune complex vasculitides. The optimal treatment regimen is still unclear, as various protocols are reported in the literature, and the long-term effects have not been thoroughly studied. Anti-IL-1 antibody therapies, such as canakinumab and anakinra, are promising options, particularly for patients with cutaneous and joint manifestations. However, these medications often provide only partial relief since IL-1 is not the sole mechanism underlying UV. The duration of treatment is also uncertain, as studies indicate that relapses can occur soon after discontinuation. Plasmapheresis may also be considered, as it can offer temporary symptom relief. Other therapies mentioned in the literature, such as intravenous immunoglobulin, anti-IL-6 medications, and anti-TNF agents, require further research to determine their efficacy.

Our review has several strengths. It provides a comprehensive overview of UV and HUV, synthesizing current knowledge and offering a structured therapeutic framework to assist clinicians in managing these complex conditions. Additionally, it highlights critical gaps in research, emphasizing the need for further studies to establish treatment guidelines and optimal therapy durations. However, the review also has limitations. Due to the rarity of this condition, there is a scarcity of large-scale studies, which may limit the generalizability of our findings. Furthermore, the review reflects

diverse treatment protocols reported in the literature, leading to variability in clinical practice. Finally, there are insufficient long-term data on the efficacy and safety of treatments, posing challenges for formulating definitive guidelines. Overall, while our review provides valuable insights and a treatment framework, it underscores the necessity for more extensive research to inform clinical practice.

CONCLUSION

UV and HUV are rare forms of vasculitis, and their exact pathophysiology is not yet fully understood. Additional research is necessary to establish specific treatment guidelines and to determine the optimal duration of therapies. This review highlights these gaps in knowledge and aims to provide a stepwise approach to treatment, helping to guide clinicians in managing these complex conditions effectively.

Conflicts of interest

Dr Groleau has no conflicts of interest. Dr Mereniuk has received consultation or presentation fees from AbbVie, Amgen, Arcutis, AstraZeneca, Bausch, Janssen, Leo Pharma, Lilly, Medexus, Pfizer, and Sun Pharma outside the submitted work. Dr Makhzoum has received consultation or presentation fees from Roche, Otsuka, GlaxoSmithKline, AstraZeneca, and Sanofi and is an investigator in clinical trials funded by Health Canada, the Vasculitis Clinical Research Consortium (VCRC), Janssen, Novartis, and AbbVie outside the submitted work.

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