



# Treatment of systemic vasculitis

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## Purpose of review

This review will attempt to summarize the most potentially impactful new data on the treatment of systemic vasculitic conditions, including ANCA-associated vasculitis (AAV), giant cell arteritis, polymyalgia rheumatica and Takayasu arteritis.

## Recent findings

Rituximab, cyclophosphamide, upadacitinib, baricitinib, mepolizumab, benralizumab and tocilizumab have all had new clinical trials and observational data from real world registries showing their treatment benefit in various vasculitic conditions. The recently developed classification criteria for five different vasculitic conditions (AAV, giant cell arteritis, and Takayasu arteritis), very important for clinical trial recruitment, have serious methodological issues that continue to be present in the new criteria sets and these need to be addressed before they can be widely adopted.

## Summary

Important new data over the last several years for the treatment of systemic vasculitis have the potential to change how these conditions are managed. The remaining issues outlined in this review still need to be addressed to best serve vasculitis patients.

## Keywords

ANCA-associated vasculitis, classification criteria, giant cell arteritis, polymyalgia rheumatica, Takayasu arteritis, treatment guidelines

## INTRODUCTION

Treatment and long-term management of systemic vasculitic conditions, including ANCA-associated vasculitis (AAV), giant cell arteritis (GCA), polymyalgia rheumatica (PMR) and Takayasu arteritis (TA) have been an active area of research in recent years. This review will attempt to summarize the most potentially impactful new data and publications over the last 18–24 months and try to discuss some of the controversy surrounding these with the aim to improve patient care in systemic vasculitis.

## CLASSIFICATION CRITERIA

2022 saw publications for the new classification criteria for the five major vasculitic syndromes, granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), eosinophilic granulomatosis with polyangiitis (EGPA), GCA, and TA [1–5]. As previously discussed, an important methodological issue was that these criteria were not appropriately validated in independent validation sets of patients [6,7].

Tomelleri *et al.* undertook the validation of the TA criteria in their cohort [8]. They demonstrated that the new criteria set had decreased specificity

compared to the old criteria set, significantly different than what had been published in the original criteria paper [4], with the 2022 American College of Rheumatology (ACR)/European Alliance of Associations for Rheumatology (EULAR) criteria having better sensitivity (95.83% vs. 82.94%) and a negative predictive value, but a markedly decreased specificity (63.51% vs. 90.54%) and a positive predictive value, which further emphasize the issues with the new criteria development process. In rare conditions such as AAV, the specificity of classification criteria is more important than the sensitivity, especially when they are used for research purposes, as the increased likelihood of misdiagnosing someone with AAV when they have another condition is increased with a set of criteria that have lower specificity, which seems to be case with the new criteria. This has ethical implications, as well. In a research setting,

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**Curr Opin Rheumatol** 2026, 38:6–11

DOI:10.1097/BOR.0000000000001129

## KEY POINTS

- The 2022 AAV classification criteria need proper validation studies before they are widely adopted for clinical research.
- Baricitinib, upadacitinib, tocilizumab, mepolizumab, and benralizumab have new data suggesting they will become part of the treatment options for patients.
- As always, more data, especially longer term follow up, are needed on these medications before they are more widely used in the treatment of vasculitis.

patients may be exposed to drugs that have not yet been proven to help AAV patients, so being as certain as we can that these patients actually have AAV is critical [9]. The compliance with the important issue of independent validation cohorts has thus far not been vigilantly evaluated by ACR and EULAR in preparation of the AAV criteria. Revalidation of these criteria are urgently needed by both ACR and EULAR before they are widely incorporated into research projects.

## TREATMENT

### Granulomatosis with polyangiitis and microscopic polyangiitis

#### Rituximab

Use of reduced dose glucocorticoids (redGC) along with rituximab in the treatment of granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA) was studied by Nagle *et al.* [10<sup>¶</sup>]. This was a retrospective, multicenter descriptive study in 19 university and general hospitals in France and Luxembourg. They showed that the subgroup of patients treated with rituximab and redGC, had a higher risk of experiencing the primary study outcome (a composite outcome that included minor relapse, major relapse, progression before achieving remission, occurrence of end-stage kidney disease (ESKD) requiring dialysis for >12 weeks and/or kidney transplantation and death) with a hazard ratio (HR) of 2.36 [95% confidence interval (CI) 1.18–4.71]. In the original PEXIVAS study, where this redGC regimen was studied, rituximab with redGC showed a trend towards less efficacy as well, with a HR of 1.86 (95% CI 0.83–4.14) for meeting the primary outcome of death from any cause or ESKD [11]. Comparing the differences between the two patient populations offers clues as to why this was the case. The PEXIVAS

study enrolled both GPA (40%) and MPA (60%) patients, defined by their ANCA antibodies, myeloperoxidase (MPO) and proteinase 3 (PR3). PR3 positivity is a known risk factor for more severe disease and more closely associates with a diagnosis of GPA. In the PEXIVAS study rituximab was used for remission induction in about 15% of patients. In the real-world study by Nagle *et al.* 60% of patients had GPA and 40% had MPA, and 53% were positive for PR3 while 45% were positive for MPO. Rituximab was the drug used for remission induction in 74% of the patients (78% for standard GC vs. 71% for redGC). Compared to the PEXIVAS study, there were more GPA patients in the real-world study, more patients were started on rituximab for remission induction, and the worst outcome was seen in the rituximab with redGC group. With fewer GPA (using PR3 as a proxy) patients and fewer patients using rituximab for remission induction in the PEXIVAS study, there was still a trend for worse outcome in those who used rituximab with redGC for remission induction.

These worse outcomes are likely due to several issues we currently have with AAV trials [12]. First, GPA and MPA patients probably should not be enrolled into the same trial; if they are, the results should be analyzed and reported separately. GPA and MPA are different conditions and GPA patients commonly have a more severe disease course. Diluting the severity of the pool patients in a trial by enrolling MPA patients along with GPA patients, would not unexpectedly favor rituximab as a remission induction agent. This has, it seems, to have led majority of patients with AAV to be treated with rituximab, as shown in the real-world report by Nagle *et al.* leading to worse outcomes for some patients who may have done better with cyclophosphamide as their remission induction medication. Evidence for this is also present in the PEXIVAS study itself, where administration of oral cyclophosphamide as induction therapy was associated with lower risks of relapse [13]. Additionally, the relentless push to decrease GC use in vasculitis treatment seems to have led to a potentially weaker option for sicker patients, rituximab, to be used also with reduced doses of GC, compounding the potential harm to patients, probably leading to worse outcomes. There needs to be a frank discussion of these accumulating data to potentially revise our approach to treating AAV and the guidelines that impact clinical practice.

#### Abatacept

Langford *et al.* compared the efficacy of abatacept to placebo for the treatment of relapsing, nonsevere GPA, as a potential safer option [14]. The primary end point was the rate of treatment failure, defined as

relapse, disease worsening, or failure to get to a Birmingham Vasculitis Activity Score (BVAS) score of 0 or 1 by 6 months. Sixty-five patients were studied, and no statistical difference in the treatment failure rate was found between the abatacept and placebo groups. Treatment with abatacept did not show any benefit in any of the secondary outcomes, either; there was no difference in the frequency or severity of adverse events between treatment arms. In conclusion, even though safety of abatacept was similar to the placebo group overall, abatacept failed to help control the disease activity and did not reduce the risk of relapse, severe worsening, or help achieve remission.

## Eosinophilic granulomatosis with polyangiitis

### Mepolizumab

Weschler *et al.* [15]. reported the open label extension study (OLE) of MIRRA, which had shown the efficacy of mepolizumab, a humanized monoclonal antibody that specifically targets interleukin (IL)-5 to reduce proliferation, activation and survival of eosinophils, and is approved for the treatment of eosinophilic granulomatosis with polyangiitis (EGPA) [16]. MIRRA study, a phase 3 placebo-controlled trial, showed that mepolizumab was associated with higher rates of remission and had significantly fewer relapses over 52 weeks of treatment, in addition to glucocorticoid sparing benefits, compared to placebo. The OLE study that enrolled patients from MIRRA who continued to require oral glucocorticoids  $\geq 5$  mg/day after the conclusion of MIRRA. All 100 patients who enrolled in the OLE received mepolizumab 300 mg subcutaneously every four weeks plus standard of care until mepolizumab was discontinued. No new safety signals were identified compared to the mother trial and the median glucocorticoid dose decreased from 10.0 mg/day to 5.0 mg/day at study exit. Furthermore, the percentage of patients using glucocorticoids  $>7.5$  mg/day decreased from 75% at baseline to 32% while 28% of the patients discontinued them. The OLE study showed that mepolizumab continued to be well tolerated and led to further decrease in glucocorticoid use.

### Benralizumab

In another study, benralizumab was compared to mepolizumab for the treatment of EGPA [17].

This was a randomized, double blind noninferiority trial to evaluate the efficacy and safety of benralizumab as compared with mepolizumab, with the primary endpoint of remission at weeks 36 and 48. One hundred forty patients were treated and the

percentage of patients with remission at weeks 36 and 48 was 58% in the benralizumab group and 56% in the mepolizumab group, showing noninferiority but not superiority of benralizumab over mepolizumab. The accrued duration of remission and the time to first relapse were similar in the two groups. Adverse events were also similar between the two drugs, along with the efficacy data, suggesting that benralizumab may potentially be a good treatment option for patients with EGPA.

### Rituximab

Another study looked at the comparison of rituximab to conventional therapy for remission induction in EGPA [18]. This randomized double-blind study enrolled 105 patients with active disease defined as a BVAS of 3 or greater and compared glucocorticoids plus rituximab with conventional therapy (glucocorticoids alone or in combination with cyclophosphamide in severe forms) for induction of remission, a BVAS of 0 and a prednisone dose of 7.5 mg/day or less at day 180. Thirty-three (63.5%) patients in the rituximab group achieved the primary end point compared with 32 (60.4%) in the control group (relative risk, 1.05 [95% CI, 0.78–1.42];  $P=0.75$ ); results were similar at day 360. There was no difference noted in duration of remission, either, between the two strategies ( $48.5 \pm 6.51$  weeks in the rituximab group and  $49.1 \pm 7.42$  weeks in the conventional strategy group,  $P=0.41$ ). All relapse and major relapse rates were similar between the two groups, as well as glucocorticoid use. In conclusion, rituximab was not superior to a conventional remission induction therapy in EGPA. Authors discuss that this study was not designed to test the two treatment strategies in severe EGPA and state that rituximab and cyclophosphamide cannot be considered equivalent in patients with severe disease. This assessment is consistent with the data, as rituximab was not even superior to glucocorticoids alone which was used by majority in the comparison group. An emulation trial by the same group has shown that addition of cyclophosphamide to glucocorticoids led to reduced risk of vasculitis flares [19]. The totality of these data suggest, as is the case with other AAV as will be discussed elsewhere in this paper, cyclophosphamide should be the preferred treatment option for severe cases.

## Giant cell arteritis

### Upadacitinib

One of the major developments in the treatment of giant cell arteritis (GCA) was the approval of upadacitinib recently by the FDA for this condition. This

approval came after the phase 3 trial of upadacitinib trial by Blockmans *et al.* [20<sup>¶</sup>]. Patients with new-onset or relapsing GCA were assigned to upadacitinib 15 mg ( $n=209$ ) or 7.5 mg ( $n=107$ ) with a 26 week glucocorticoid taper or placebo ( $n=112$ ) with a 52 week glucocorticoid taper. The primary end point was sustained remission between weeks 12 and 52. Upadacitinib 15 mg arm was statistically significantly better than placebo in meeting the primary end point (46.4% [95% CI, 39.6–53.2] vs. 29.0% [95% CI, 20.6–37.5];  $P=0.002$ ). It was also better for sustained complete remission, time to a disease flare, cumulative glucocorticoid exposure, and patient-reported outcomes. Interestingly, upadacitinib 7.5 mg dose was also better than placebo numerically with 41.1% vs. 29.0% of patients achieving sustained remission, but this was not statistically significant. However, when the actual response difference between the two doses of upadacitinib are compared, there is only about a 5% difference, 41.1% vs. 46.4%. There were no major differences in adverse events, except serious adverse events were lower in the upadacitinib 7.5 mg compared to upadacitinib 15 mg, 12.1% vs. 22.5%, along with 2 deaths in upadacitinib 15 mg and no deaths in upadacitinib 7.5 mg. Based on this study upadacitinib was approved for the treatment of GCA for the 15 mg dose, however, it is hard to argue that there was no benefit from the lower dose, upadacitinib 7.5 mg, as it was close to significance and in a larger patient population, which will be the case when it is used in the real world, it may have also reached significance. It would be good to have the option of using the lower dose, too, for the treatment of GCA, especially as it was also overall associated with fewer serious adverse events compared to upadacitinib 15 mg.

### Tocilizumab

Tocilizumab had been approved for the treatment of GCA after the successful GiACTA trial [21]. Since then, there have been efforts to assess if lower doses and/or a shorter duration of glucocorticoids may be used in the first year along with tocilizumab to achieve remission.

Two groups have been trying to answer this question. The GUSTO trial [22<sup>¶</sup>] was an open label, small proof of concept trial that aimed to evaluate the efficacy and safety of tocilizumab monotherapy for 52 weeks and then stopping it, along with initial ultra-short-term glucocorticoid treatment at baseline only, in patients with new-onset GCA. It showed that 14/18 (78%) of the patients achieved remission within 24 weeks and 13 of 18 showed no relapses up to 52 weeks (72%) after an ultra-short pulse of glucocorticoids of 500 mg methylprednisolone intravenously (IV) for three consecutive days only.

The same group now looked at the role of MRI in disease activity monitoring in GCA as part of the GUSTO trial [23]. They examined vascular and musculoskeletal inflammation using MRI; cranial, thoracic and abdominal MRI exams were performed at baseline and at weeks 24, 52, and 104. Vasculitic vessels were still detectable in one in four cranial segments at week 24. These were resolved at weeks 52 and 104, however, large vessels, except for the ascending aorta, showed ongoing inflammatory activity over time, suggesting that while vasculitic manifestations in the cranial vessels normalized after 52 weeks of treatment, large vessel findings persisted despite the lasting full clinical remission. This, of course, questions the decision about stopping tocilizumab treatment once clinical remission is achieved at week 52. If there is ongoing inflammation in the large vessels, while the patient is clinically silent, it may mean further treatment is needed to prevent future disease manifestations and damage. A further question is if “true” remission needs to be defined as remission in signs and symptoms of GCA along with normal imaging, a concept that requires additional studies.

The TOPAZIO study also looked at the same issue [24]. The goal of the study was to assess the impact of tocilizumab monotherapy after ultra-short-pulse glucocorticoids on clinical manifestations, and vessel inflammation and damage in large vessel-GCA (LV-GCA). This was a prospective observational study, where 18 patients received 500 mg per day IV methylprednisolone for three consecutive days at baseline only and weekly tocilizumab injections from day 4 until week 52. PET/CT was performed on all patients at baseline and at weeks 24 and 52, with reduction in the PET vascular activity score (PETVAS) as the primary end point. Both at weeks 24 and 52, a significant reduction in PETVAS was seen, with about 56% (10/18) and 47% (8/17) of patients in relapse free remission at weeks 24 and 52, respectively. There were no patients who had new aortic dilation, however, 4 patients with aortic dilation at baseline showed increases in aortic dilation.

Next, Muratore *et al.* followed these patients off treatment to assess the maintenance of efficacy in the next 26 weeks out to week 78 for the 17 patients who were available for analysis [25]. PETVAS still showed a significant decrease compared to baseline at week 78, however, compared with week 52, PETVAS significantly increased, 6 months after tocilizumab discontinuation. Relapse-free clinical remission at week 78 was 65% (11/17), and age and sex-adjusted hazard ratio (95% CI) for each unit increase of PETVAS indicating subsequent relapse was 1.36 (0.92–2.00).

Both of these studies suggest that while ultrashort glucocorticoids at baseline along with tocilizumab for



a year and then discontinuation was able to provide very good clinical remission. There seems to be, at least in some patients, ongoing large vessel inflammation, initially subclinical but potentially leading to future relapses. This needs to be considered before stopping tocilizumab after a year of treatment even if the patients are in clinical remission and tocilizumab may need to be continued; how much longer remains unknown.

## Polymyalgia rheumatica

### Baricitinib

Saraux *et al.* looked at baricitinib in the treatment of polymyalgia rheumatica (PMR), which has been traditionally treated with glucocorticoids and there remains an unmet need for treatments that can limit or end the use of glucocorticoids, which are associated with adverse events [26]

BACHELOR was a randomized, double-blind, placebo-controlled, parallel-group trial at six centers in France. All patients ( $n=34$ ) had new onset PMR and were randomized to 4 mg baricitinib orally or placebo (with oral glucocorticoids as rescue treatment) for 12 weeks, which was later followed by 2 mg baricitinib or placebo for another 12 weeks. The primary endpoint (CRP PMR-AS of 10 or less) was reached at week 12 by 14 of 18 (78%) participants in the baricitinib group and 2 of 15 (13%) participants in the placebo group (relative risk 5.8, 95% CI 3.2–10.6; adjusted  $P < 0.0001$ ). There were no deaths and no major adverse cardiovascular events in either group.

This small study suggests that, compared with placebo, patients with PMR receiving 4 mg baricitinib are less likely to need oral glucocorticoids to have low disease activity and without any new safety signals.

### Tocilizumab

Efficacy of tocilizumab had been previously shown for the treatment of PMR [27].

Assaraf *et al.* [28]. conducted a multicenter retrospective analysis of use of tocilizumab in PMR patients requiring glucocorticoid-sparing treatment. They had 53 patients; 31 had active disease despite conventional synthetic DMARD treatment. Glucocorticoid dose was down to less than or equal to 5 mg/day in 77% of the patients at 6 months, and in 97% of the patients at 12 months. Proportions of glucocorticoid-free patients 6 and 12 months after the first tocilizumab infusion were 22.5% and 58.3%, respectively. In addition, tocilizumab infusion spacing rather than discontinuations seemed to be a better tapering strategy.

## Guidelines

British Society for Rheumatology published their guidelines for the management of AAV in 2025 [29]. Recommendations were updated from the published 2014 guideline mainly in treatment for GPA and MPA, management of subglottic stenosis and ear, nose, and throat (ENT) manifestations of AAV, and management and treatment for EGPA. For GPA and MPA, both rituximab and cyclophosphamide were recommended for remission induction, with a preference for rituximab in active relapsing disease. In cases of no evidence of life-threatening organ involvement, methotrexate or mycophenolate mofetil are also suggested. For maintenance of remission, rituximab was preferred, with azathioprine and methotrexate as alternatives in select cases, at least for 24–48 months. Avacopan was suggested as an additional option to help decrease glucocorticoid related issues. Similar recommendations, in addition to teamwork with ENT specialists are noted for GPA with ENT involvement. For EGPA, anti-IL-5/5R biologics were recommended for nonlife threatening disease, with rituximab and cyclophosphamide reserved for more serious involvement. Overall, these recommendations mirror similar ones from other rheumatology societies and also reflect what the current practice is. As discussed elsewhere here, there is still some controversy about the role of rituximab and cyclophosphamide in severe GPA and this was not addressed in this guideline.

## CONCLUSION

There continues to be issues with the 2022 vasculitis classification criteria that need to be addressed before more and more clinical trials continue to use them for selecting patients. A frank discussion of the methodological issue by the societies which sponsored the development of these criteria, ACR and EULAR, is urgently needed.

We now have new options in upadacitinib, for the treatment of GCA, baricitinib for PMR and mepolizumab and benralizumab for EGPA. Better understanding the role of rituximab cyclophosphamide in GPA and MPA is also provided with new data, suggesting that cyclophosphamide may be the preferred agent for severe cases, and potentially the first agent for GPA, which tends to have worse activity and outcomes compared to MPA. Tocilizumab continues to be a good agent for GCA, with potentially allowing for only limited glucocorticoid use, however, longer term large vessel involvement needs to be studied further after discontinuation of tocilizumab after a year. Tocilizumab also seems to be beneficial for PMR management.

Rituximab for EGPA and abatacept for nonsevere GPA did not show benefit over current treatment with glucocorticoids.

As always, totality of the data needs to be taken into account when we apply the new information to our individual patients. In this line, the new classification criteria need to be properly validated before they are fully adopted, the new treatment recommendation communicated widely and the newly approved drugs for the treatment of vasculitic conditions used in the appropriate patients, providing new options for better outcomes.

## Acknowledgements

None.

## Financial support and sponsorship

None.

## Conflicts of interest

Research support: Amgen, BMS, Genentech, Consulting: Amgen, Chugai, Otsuka, Soligenix, argenx.

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- of special interest
- of outstanding interest

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