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BSR Guidelines

British Society for Rheumatology RHEUMATOLOGY OXFORD

BSR Guidelines

The 2025 British Society for Rheumatology management recommendations for ANCA-associated vasculitis

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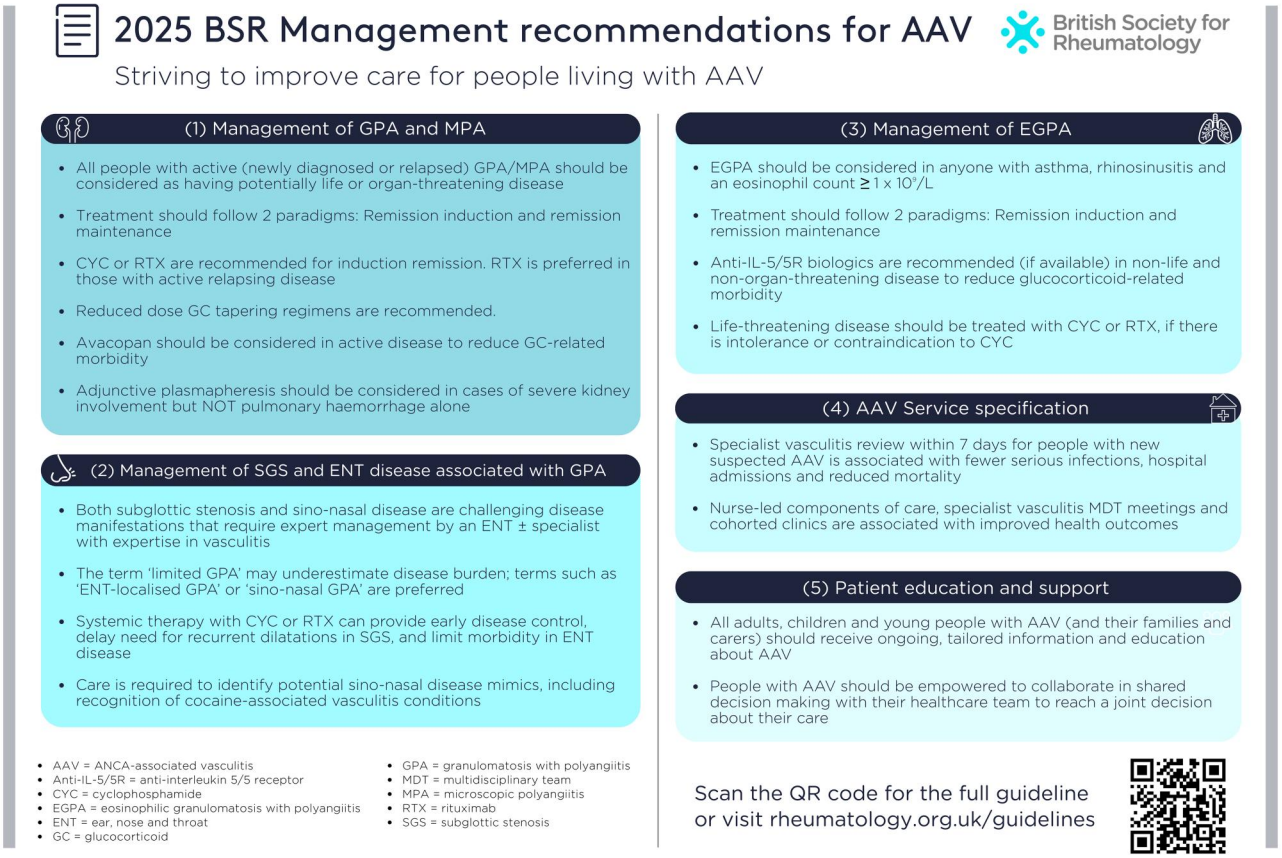
Abstract

ANCA-associated vasculitis (AAV) is comprised of three specific conditions: granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA). Since the publication of the last British Society for Rheumatology (BSR) and British Health Professionals in Rheumatology (BHPR) guideline for the management of adults with AAV in 2014, a plethora of randomized controlled trials, additional research and recommendations have provided novel insights into how the management of AAV can be optimized, thus improving patient quality of life. The BSR AAV Working Group (WG) reviewed published guidelines, undertook a systematic literature review and utilized expertise from specialist vasculitis centres across the UK and patient representatives to formulate a list of 26 recommendations with corresponding strength of agreement (SOA) scores. Recommendations were updated from the published 2014 BSR and BHPR guideline. The 26 recommendations encompassed five key domains: 1. Treatment for GPA and MPA; 2. Management of subglottic stenosis and ear, nose and throat (ENT) manifestations of AAV; 3. Management and treatment for EGPA; 4. Service specifications; 5. Patient education and support. These recommendations provide an update on care delivery of AAV based on current evidence and specialist opinion. In addition, we have provided research and audit recommendations to support equitable access to care and improve health outcomes. The lay summary that accompanies this abstract can be found in Supplementary Data S1, available at *Rheumatology* online.

Keywords: ANCA vasculitis; treatment.

Graphical abstract



Background and rationale for recommendations

Anti-neutrophil cytoplasm antibody (ANCA)-associated vasculitides (AAV) are heterogeneous, multisystem disorders characterized by inflammation and necrosis of small and medium blood vessels with unknown aetiology. Three distinct clinico-pathological syndromes have been identified: granulomatosis with polyangiitis (GPA), microscopic polyangiitis

(MPA) and eosinophilic granulomatosis with polyangiitis (EGPA) associated with autoantibodies directed against neutrophil granular proteins, proteinase 3 and myeloperoxidase. These conditions are uncommon with incidence and prevalence rates of about 25 per million population and 200 per million, respectively [1]. Despite significant advances in treatment, mortality rates remain 2.3 times worse than that of the general population [2]. Early diagnosis, instituting appropriate immunosuppression swiftly, and limiting toxicity from treatment is key to mitigating mortality and damage from AAV.

The current British Society for Rheumatology (BSR) guideline for management of AAV was published in 2014 [3] and was an important step forward in managing this complex disease. It provided a roadmap for best practice management aiming to harmonize treatment and investigation of AAV. Due to significant changes in the management of AAV, incorporating new trials and evidence-based therapies, the current BSR recommendations no longer reflect best practice and updates are required.

Objective

This document offers systematic and evidence-based recommendations to support UK clinicians in management of AAV across the whole life course.

Target audience

The primary audience consists of health professionals in the UK who are directly involved in the management of patients with AAV. The recommendations are designed to apply across clinical specialties and all branches of medicine and professional groups supporting people living with AAV and people living with AAV themselves.

Commentary scope

The scope of this document is broader than that of the 2014 BSR guideline and extends to management of children and adolescents as well as adults living with disease. Where differences between the management of adults and children are suggested, these are explicitly stated. Given the different management strategies used for EGPA compared with GPA and MPA, these clinical entities are considered separately.

The authors focused on five key recommendation domains: GPA and MPA treatment (domain 1), management of subglottic stenosis and ear, nose and throat (ENT) disease associated with GPA (domain 2), EGPA management and treatment (domain 3), service specification (domain 4) and patient education and support (domain 5).

Diagnosis and investigation of systemic GPA and MPA or the management of disease or treatment related chronic damage is not covered in these recommendations, as the guidance outlined in the BSR guideline 2014 remains current [3].

Despite there being considerable advances in the classification of AAV since the publication of the BSR guideline 2014, with the 2022 ACR/EULAR Classification Criteria for vasculitis being the most recent [4–6], classification criteria have been validated for research use only and not for diagnostic or treatment purposes and are therefore not discussed further in these management recommendations.

Stakeholder involvement

These recommendations were commissioned by the BSR Guidelines Steering Group (GSG). A working group (WG) was created, comprising a chair (L.H.) alongside representatives from relevant stakeholders (Table 1).

Involvement and affiliations of stakeholder groups

The WG consisted of adult and paediatric rheumatology and nephrology consultants and resident doctors, consultants in respiratory medicine and ENT surgery, clinical nurse specialists and representatives from the Vasculitis UK Charity, including an expert through experience (a person living with AAV). All members of the WG contributed to the development of all the recommendations.

Recommendation development

Literature search: scope and search strategy

Since 2014, there have been many updated international guidelines addressing the management of AAV published, including American College of Rheumatology (ACR) 2021, Kidney Disease Improving Global Outcome (KDIGO) 2021 (and update 2024), The European Alliance of Associations for Rheumatology (EULAR) 2022 and European EGPA Study Group (EESG) Nature Evidence Based Guideline 2024 [7–10], amongst others [11, 12]. Given that extensive literature searches were performed across these published guidelines, a full systematic review of the literature since 2014 was not repeated. Instead, we accepted existing literature as presented by the ACR 2021, KDIGO 2021 and update 2024, EULAR 2022 and EESG 2024 guidelines and focused on differences between them [7–10]. In addition, we undertook a systematic review of literature published after the EULAR guideline literature review was completed (Dec 2021) where topics or questions had already been considered. New topics, not covered in the BSR guideline 2014, or the published international guidelines literature search, the systematic reviews were from December 1990. Key terms were agreed among the members of the domain subgroup. Systematic literature searches, in Medline and limited to English language publications, were performed separately for each domain by the small working groups. Details of domain-specific literature searches are provided in Supplementary Data S2, available at *Rheumatology* online.

Rigour of development

Recommendations in this report were developed based on the BSR Creating Clinical Guidelines Protocol v5.4 (2023) using AGREEII (Appraisal of Guidelines for Research and Evaluation II) methodology. Following a virtual meeting of the full WG, the scope of the project was agreed and grouped into domains. Small working groups for each domain were formed and developed initial recommendations for discussion by the full WG. These initial recommendations were then adapted over a series of virtual meetings of the full WG. The guideline protocol was not followed fully, in that the scope of the work was not published prior to the final literature review. The reason for this was that, at the time this work was commenced, no significant new evidence had been highlighted since publication of the ACR 2021, KDIGO 2021 and update 2024, EULAR 2022 and EESG 2024 guidelines [7–10], and the intention was to provide a timely update of the BSR guideline 2014 relevant to the UK clinical context. It is for this reason that this manuscript is described as ‘Management Recommendations’ rather than a ‘Guideline’. Development was overseen by the BSR Guideline Steering Group throughout. Subgroups (memberships summarized in Table 1) developed the initial recommendations, which

Table 1. Working group members and roles

WG member	Role	Subgroup domain
Dr Madura Adikari	Consultant rheumatologist	5
Mr Chadwan Al Yaghchi	Consultant ENT surgeon	2,3
Ms Zoi Anastasa	Expert by experience and Director of Operations Vasculitis UK	5
Prof Neil Basu	Consultant rheumatologist	4
Dr Kathryn Biddle	Trainee rheumatologist	1
Prof Paul Brogan	Consultant paediatric rheumatologist	1,2,3,4,5
Dr Dimitrios Chanouzas	Consultant nephrologist	1,4
Dr Shouvik Dass	Consultant rheumatologist	4
Prof David D'Cruz	Consultant rheumatologist	2,3
Ms Emmandeep Dhillon	Specialist nurse	5
Ms Georgina Ducker	Specialist nurse	4,5
Prof Siân Griffin	Consultant nephrologist	1,4,5
Prof Lorraine Harper	Chair of working group & consultant nephrologist	1,2,3,4,5
Dr Rosemary Hollick	Consultant rheumatologist	4
Prof David Jackson	Consultant respiratory physician	2,3
Dr Judith Jade	Trainee rheumatologist	4
Dr Catherine King	Trainee nephrologist	1
Dr Matko Marlais	Consultant paediatric nephrologist	1,4,5
Mr Marcos Martinez Del Pero	Consultant ENT surgeon	2,3
Dr Alice Mason	Consultant rheumatologist	4
Dr Stephen McAdoo	Consultant nephrologist	2,3
Dr Devesh Mewar	Consultant rheumatologist	4
Dr Janice Mooney	Specialist nurse	5
Dr Eleana Ntatsaki	Consultant rheumatologist	1,4,5
Dr Fiona Pearce	Consultant rheumatologist	1
Dr Benjamin Rhodes	Consultant rheumatologist	4
Dr Hitasha Rupani	Consultant respiratory physician	2,3
Prof Alan Salama	Consultant nephrologist	1,2,3,4,5
Prof Salman Siddiqui	Consultant respiratory physician	2,3
Dr Pasupathy Sivasothy	Consultant respiratory physician	2,3
Dr Rona Smith	Consultant nephrologist	4
Dr Harold Wilson-Morkeh	Trainee rheumatologist	2,3

Composition of working group (WG): list of members, professional roles and involvement in subgroup domains. Domain 1: granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA) treatment; domain 2: ear, nose and throat (ENT) disease associated with GPA; domain 3: eosinophilic granulomatosis with polyangiitis (EGPA) treatment; domain 4: service specification; and domain 5: patient education and support.

were then adapted over a series of virtual meetings of the full WG. The final draft of the guidance was submitted to the BSR Guidelines Steering Group for stakeholder and internal review and feedback.

Quality of evidence

The quality of evidence was graded according to the GRADE approach. Evidence was graded as strong (A), moderate (B) or weak (C), reflecting confidence in the estimates of benefit or harm.

Strength of agreement

The content and wording of all recommendations were discussed until all members of the WG were satisfied that they could score at least 80 on a scale of 1 (no agreement) to 100 (full agreement). The strength of agreement for each recommendation is presented as the mean of the WG's individual ratings, expressed as a percentage. Anonymized votes are shown in Supplementary Data S3, available at *Rheumatology* online.

Strength of recommendation

The content and wording of all recommendations were discussed and strength of recommendation agreed upon with all members of the WG, assigned as strong (designated as 1) or weak (designated as 2).

Recommendations and rationale

Diagnosis and investigations

We endorse the diagnosis and investigation principles outlined in the BSR guideline 2014 (3) for MPA and GPA. The only new recommendation in this area relates to the routine screening for cocaine use in all patients presenting with sinonasal disease as standard of care [13] (recommendation 15). The unchanged recommendations from the BSR guideline 2014 on the management of AAV can be found in Supplementary Data S2, available at *Rheumatology* online. Given advances in diagnosis and treatment options for EGPA since publication of the last BSR recommendations, we have included a review of diagnosis, disease assessment and severity stratification in this edition.

Terminology and definitions

We advocate the EULAR terminology and distinction of AAV to organ- or life-threatening disease and AAV with non-organ- or life-threatening manifestations. Sinonasal disease is often viewed as non-organ-threatening; however, it is important to highlight that sinonasal disease can cause significant local destruction and may be considered organ-threatening. Examples of manifestations that are *not* ultimately organ- or life-threatening may include nasal and paranasal disease *without* bony involvement (erosion) or cartilage collapse or olfactory dysfunction or deafness; skin involvement *without* ulceration; myositis (skeletal muscle only);

non-cavitating pulmonary nodules; episcleritis. Assessment of disease severity in the individual patient may differ, and the treatment choice is ultimately a clinical judgment.

Recommendation 1	All people with lived experience of active (newly diagnosed or relapsed) AAV should be considered as having potentially life- or organ-threatening disease (GRADE 1C, SoA 98%).

We also endorse the EULAR definitions of disease activity states as shown in Table 2.

Table 2. EULAR consensus definitions of AAV disease activity [10]

Activity state	EULAR consensus definition
Active disease	Presence of typical signs, symptoms or other features (such as glomerulonephritis or pulmonary nodules) of active AAV.
Remission	Absence of typical signs, symptoms or other features of active AAV with or without immunosuppressive therapy.
Sustained remission	Absence of typical signs, symptoms or other features of active AAV over a defined time period with or without immunosuppressive therapy.
Response	≥50% reduction of disease activity score and absence of new manifestations.
Relapse	Recurrence of active AAV after a period of remission.
Refractory	Unchanged or increased signs, symptoms or other features of active AAV after a period of standard induction therapy. Damage, infections, side effects of treatment or comorbidities as potential causes of the persistent or worsened disease manifestations need to be ruled out.

Domain 1: GPA and MPA treatment

These recommendations apply to people with lived experience of GPA and MPA. A flowchart (Fig. 1) outlines the GPA and MPA treatment algorithm for quick reference.

Primary induction of remission for GPA and MPA

Recommendation 2a	All people with lived experience of active GPA or MPA should be assessed for induction of remission treatment with immunosuppressants combined with glucocorticoids (GC) or avacopan (GRADE 1A, SoA 99%).
Recommendation 2 b	The recommended options for immunosuppression for remission induction of newly diagnosed GPA or MPA are intravenous pulsed cyclophosphamide (CYC) or rituximab (RTX) (GRADE 1A, SoA 98%).
Recommendation 2c	For active relapsing disease, treatment with RTX is preferred (GRADE 1B, SoA 97%).
Recommendation 2d	A combination of both CYC and RTX can be considered for organ-threatening or life-threatening disease (GRADE 2C, SoA 98%).
Recommendation 2e	Certain individuals with active GPA or MPA, with no evidence of life- or organ-threatening disease, may be considered for alternative induction therapy with methotrexate (MTX) or mycophenolate mofetil (MMF) (GRADE 1A, SoA 96%).

These recommendations are in broad alignment with the 2014 BSR guideline but have been extended to recommend the use of RTX or CYC in all individuals living with active disease [3]. As RCTs demonstrated equivalent efficacy of CYC and RTX in new onset disease, we do not recommend one agent over the other [14, 15]. This agrees with the 2022 EULAR guideline [10], but is discordant with the ACR guideline [7] that conditionally recommends RTX over CYC for active severe disease due to a preferable toxicity profile, although this is not supported by the PEXIVAS trial (evidence level 1A) [16]. Controversy remains across the published guidelines about the preferred use of RTX in those with severe renal involvement [estimated glomerular filtration rate (eGFR) <30 mls/min] due to limited inclusion of such patients within trials. However, observational analyses suggest no difference in benefit between CYC and RTX [17, 18]. Both KDIGO and EULAR suggest use of either agent in this setting [8, 10]. Our BSR recommendations do not suggest an eGFR cut-off for use of RTX in patients with severe disease.

We also have preference for RTX and CYC over the use of MTX or MMF due to higher rates of sustained remission and lower GC exposure, in agreement with EULAR 2022 [10]. Although no RCTs have compared RTX against other agents in patients with non-organ-threatening GPA or MPA, induction of remission trials with RTX in AAV have included patients with non-organ-threatening disease and have shown non-inferior outcomes on efficacy and safety [14].

The clinical consensus for induction therapy RTX, as indicated in the BSR and BHPR guideline 2014, is that both the dose of two 1 g infusions two weeks apart and the 'lymphoma' regimen of four infusions at weekly intervals of 375 mg/m^2 appear equally effective. In a systematic review, the two regimens were compared and found to be of equal efficacy, with no difference in the duration of B-cell depletion or the therapeutic effect [19]. If the lower dose schedule is employed, there is a significant NHS cost saving in terms of reduced NHS activity (50%) and reduced drug costs (40%) compared with the higher licensed dose [20].

We advocate RTX in active relapsing disease, as cumulative doses of CYC increase risk of malignancy and other side-effects, and the RAVE trial [14] suggested better outcomes in those with relapsing disease. This agrees with other guidelines [7, 8, 10].

A combination of both CYC and RTX may be considered for organ-threatening or life-threatening disease, although data for this is limited to the small RITUXVAS trial [15] and uncontrolled cohort studies [21, 22]; however, potential toxicity must be considered. The RITUXIVAS trial dosing regimen was rituximab at a dose of 375 mg per square meter per week, for four consecutive weeks, and intravenous cyclophosphamide at a dose of 15 mg per kilogram with the first and third rituximab infusions [15]. This approach is not recommended by ACR due to concerns regarding toxicity [7]; it is recognized by KDIGO as an alternative to 3–6 months of CYC [8] and EULAR noted the use of combined CYC and RTX to reduce cumulative doses of CYC and the possibility of GC minimization but no recommendation on its use is provided [10]. More data from controlled studies are awaited (including ENDURRANCE trial NCT03942887).

For patients with refractory disease after treatment with RTX or CYC, consideration should be given to revisiting the

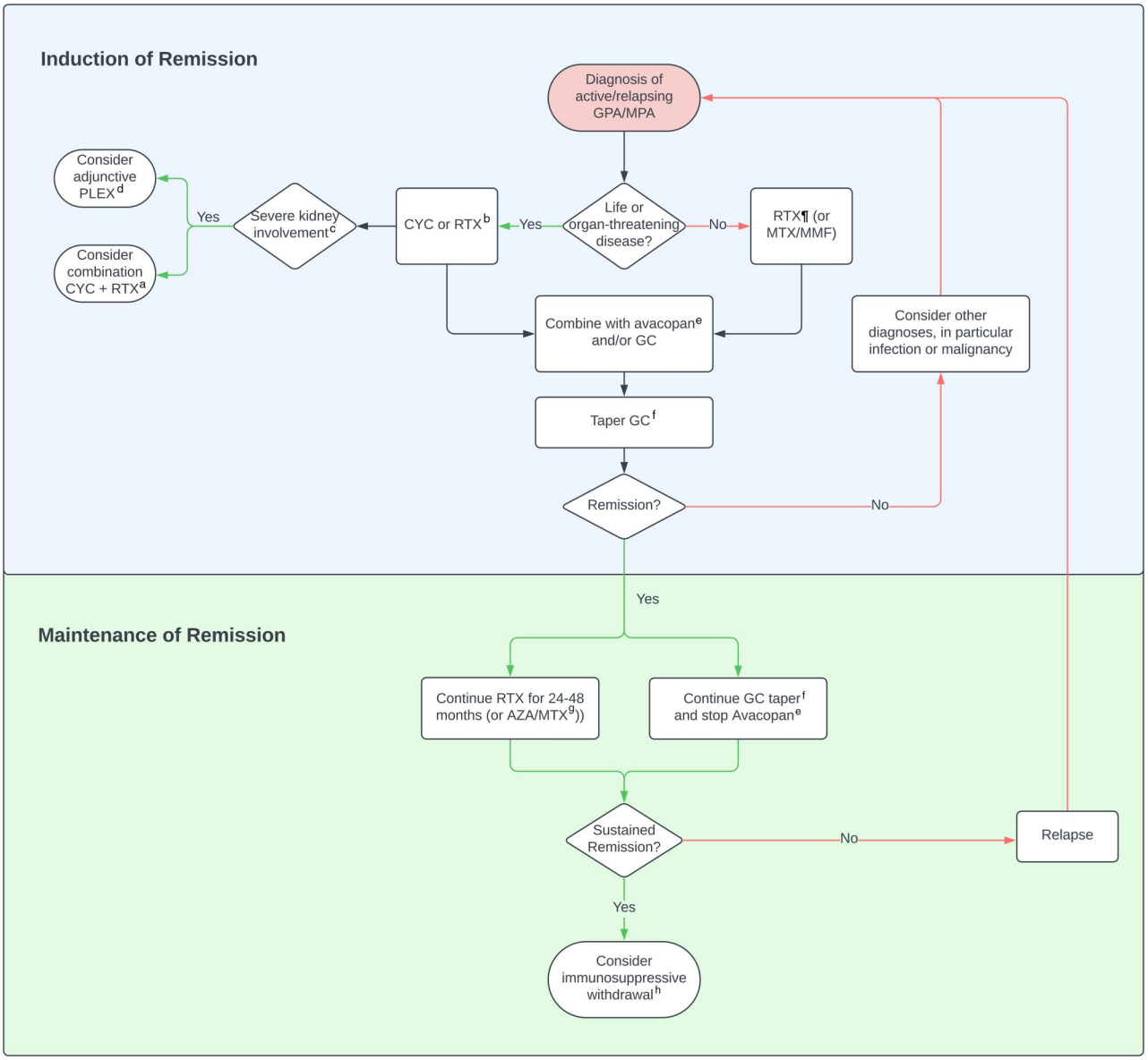


Figure 1. GPA and MPA treatment algorithm. [a]A combination of CYC and RTX can be considered in life-threatening or organ-threatening disease. [b]For patients with active relapsing disease, treatment with RTX is preferred (recommendation 2c). [c]Creatinine >300 μmol/l. [d]PLEX can be considered provided that the risk of adverse events has been weighed against the benefits. [e]The recommended duration of avacopan use is 12 months as there is no data on its use beyond this (recommendation 6). [f]The optimum duration of GC tapering is uncertain. A suggested tapering regime is outlined in Table 3 (recommendation 10). [g]MMF can be considered when there is contraindication or intolerance to RTX, AZA or MTX. [h]Decision making around the length of treatment should consider patient risk factors for relapse and infection as well as patient preferences. AZA: azathioprine; CYC: cyclophosphamide; GC: glucocorticoids; MMF: mycophenolate mofetil; MTX: methotrexate; PLEX: plasma exchange; RTX: rituximab

diagnosis and ensuring that potential underlying driving factors, such as infection, are ruled out. If these are excluded, treatment with the alternative induction agent not previously used (CYC or RTX respectively) is recommended.

BSR updates the 2014 recommendation such that MTX and MMF may be used for certain individuals with lived experience of GPA and MPA without organ- or life-threatening manifestations that need alternative induction agents to RTX and CYC. In contrast, the ACR guideline conditionally recommends MTX over CYC or RTX for non-severe disease due to preferable toxicity profile [7] despite higher long-term relapse rates. The NORAM trial used MTX as an alternative to CYC. In this study of patients with minimal or no renal disease (creatinine under 150 μmol/l), MTX was as effective as CYC at inducing remission over a year [23]. However long-term follow-up has indicated that these patients remain at higher risk of relapse [24]. EULAR does not recommend routine use of MTX as first-line treatment even in those with non-organ-threatening disease due to increased risk of relapse [10]. The MYCYC trial, an RCT excluding patients on dialysis or with life-threatening disease, showed that MMF was non-inferior to CYC for remission induction in new-onset GPA or MPA [25]. Although there was no safety benefit of MMF, and a higher relapse rate for PR3-positive patients, the trial provided evidence that MMF is a potential alternative to CYC for remission induction in non-life threatening AAV [25]. A recent

meta-analysis suggests there may be benefit in patients with MPO-ANCA disease with mild-moderate kidney disease [26]. However, controversy exists around the use of MMF in guidelines; ACR advise last-line use, EULAR suggests second-line use and KDIGO suggests use in non-organ-threatening disease with preferential use in MPO-AAV over PR3-AAV [7, 8, 10].

Plasmapheresis for GPA and MPA

Recommendation 3a	Active GPA or MPA and severe kidney involvement with creatinine >300 μmol/l should be considered for adjunctive plasmapheresis provided their risk of potential adverse events has been considered (GRADE 2B, SoA 96%).
Recommendation 3 b	For children living with active GPA or MPA, there is insufficient data to routinely recommend plasmapheresis for severe renal involvement; this therefore should only be considered on a case-by-case basis after discussion with an expert centre (GRADE 2C, SoA 96%).
Recommendation 3c	Adjunctive plasmapheresis is not routinely recommended for pulmonary haemorrhage without severe kidney involvement (GRADE 1A, SoA 96%).

The 2014 BSR guideline recommended plasmapheresis in patients with creatinine values >500 μmol/l or for other life-threatening situations such as pulmonary haemorrhage. Since then, the largest randomized controlled trial of plasmapheresis in AAV (the PEXIVAS trial) and a meta-analysis of plasmapheresis in AAV have been published [16, 27]. The PEXIVAS trial, recruiting patients with eGFR <50 mls/min and/or pulmonary haemorrhage, suggested no benefit of plasmapheresis on top of pulsed intravenous methylprednisolone (MP) and oral GC on a combined end point of end-stage kidney disease (ESKD) or death [16]. Subgroup analysis also showed no reduction in the development of ESKD with creatinine >500 μmol/l in those who received plasmapheresis [16]. This finding was at odds with the MEPEX study which demonstrated that plasmapheresis, compared with methylprednisolone, had a beneficial effect on renal survival at one year, in those with severe kidney disease (serum creatinine >500 μmol/l or requiring dialysis), but no impact on survival [28]. However, this early benefit appeared to be lost during long-term follow-up [29].

Meta-analysis of nine plasmapheresis studies has suggested an overall benefit of maintaining or gaining independent renal function one year following adjunctive plasmapheresis [27]. This may be particularly important in older AAV patients, as increased age correlates with negative dialysis outcomes [30]. The benefit on renal survival must be weighed against an increased risk of serious infection, estimated to increase by 20% with plasmapheresis in the meta-analysis [27]. Patients with the highest risk factors for infection were most at risk (absolute risk increase 13.5% at 12 months compared with 2.7% in the lowest risk group). Controversy exists around the conclusions of this meta-analysis [31].

No children were included in the studies included in the plasmapheresis meta-analysis. This, combined with the ongoing debate regarding the conclusions of the meta-analysis [27, 31] leads us to recommend that consideration of plasmapheresis in children should be on a case-by-case basis discussed with expert centres.

We have updated our recommendation on using adjunctive plasmapheresis in people living with severe renal involvement (creatinine >300 μmol/l) provided risk of adverse events are considered. This is in line with the EULAR 2022 guidance [10]. However, ACR suggest plasmapheresis may be considered for certain people living with active glomerulonephritis or who are critically ill and are not responding to treatment [7] and KDIGO suggest a practice point to consider PLEX in patients with creatinine >500 μmol/l or rapidly increasing creatinine despite treatment [8].

The PEXIVAS study does not support a treatment effect of plasmapheresis in patients with pulmonary haemorrhage, compared with those without [16]. Therefore, we do not routinely recommend plasmapheresis in patients with pulmonary haemorrhage without severe kidney involvement, in line with EULAR 2022 recommendations. The practice point by KDIGO to consider plasmapheresis in patients with diffuse alveolar haemorrhage and hypoxaemia is not well justified [8].

Glucocorticoid treatment for GPA and MPA in those not considered for avacopan use

Recommendation 4a	In organ- or life-threatening disease, we advocate treatment with oral GC at a starting dose of 50–75 mg or 1.0 mg/kg/day (dependent on weight with a maximum of 75 mg daily). Oral GC (prednisolone) should be tapered in accordance with the PEXIVAS tapering schedule, achieving a dose of 5 mg prednisolone equivalent per day by 4–5 months (see Table 3) (GRADE 1B, SoA 96%).
Recommendation 4 b	In non-organ- or non-life-threatening disease, lower GC-tapering regimens can be considered, at a starting dose of 0.5 mg/kg/day oral GC (prednisolone), with tapering in accordance with the LoVAS regimen (see Table 3) (GRADE 1B, SoA 97%).
Recommendation 4c	Whilst children were not included in PEXIVAS, this tapering regimen can be considered for adolescents. For younger children, the SHARE guideline for prednisolone tapering could also be considered [32] (GRADE 2C, SoA 96%).
Recommendation 4d	Despite commonplace use, there is a lack of supporting trial evidence for intravenous methylprednisolone (IV MP) pulses. Therefore, IV MP pulses are not routinely recommended but can be reserved as an option for the management of organ-threatening manifestations, including active renal disease and diffuse alveolar haemorrhage (GRADE 2C, SoA 97%).

High-dose GC therapy is associated with a significant adverse effect burden including infection, the leading cause of excess mortality in individuals with lived experience of GPA and MPA [33]. Accordingly, a treatment priority is the minimization of GC exposure, without compromise on the adequate treatment of AAV.

The PEXIVAS RCT compared two GC regimens in participants with organ- or life-threatening GPA and MPA, treated with CYC or RTX [16]. The reduced-dose GC regimen was

non-inferior with respect to death and the development of ESKD and resulted in a 40% reduction in oral GC exposure in the first 6 months and a significant reduction in serious infections in the first year [16]. In the LoVAS trial, participants with active AAV without severe renal involvement or pulmonary haemorrhage demonstrated non-inferiority of low-dose compared with high-dose GC regimens, regarding time to remission, death, relapse rate and ESKD [34, 35]. All participants also received RTX. A systematic review of the two studies found that reduced-dose GC regimens may reduce death and infection at 6 and 12 months, with no associated increase in the rate of ESKD [36]. Table 3 illustrates the PEXIVAS and LoVAS reduced-dose GC regimens.

In agreement with our recommendations, the EULAR and KDIGO guidelines advocate a reduced-dose GC tapering regimen for induction-remission in GPA and MPA [8, 10]. The ACR guideline conditionally recommends this approach [7].

Despite a lack of supporting clinical trial evidence, the use of IV MP pulses is commonplace for induction-remission in AAV in both real-life and trial settings, including in the PEXIVAS trial [16]. Observational data suggests that regimens with higher doses of GC, including pulsed IV MP, are associated with an increased risk of infection with no associated benefit in efficacy [37, 38]. A recent propensity-matched registry study, however, has suggested benefit on all-cause mortality for the use of methylprednisolone [39]. Further work is required in this area.

Use of complement inhibitor avacopan

Recommendation 5	Patients with active GPA or MPA should be considered for avacopan use as a steroid sparing agent, with or without a short course of glucocorticoids (tapering over four weeks) (GRADE 1A, SoA 96%).

Avacopan (usual dose 30 mg twice daily) has been introduced following the ADVOCATE trial as a potential GC avoidance strategy to circumvent the adverse effects associated with GC [40]. Efficacy, measured by sustained remission, was similar at 6 months but improved at 12 months in the avacopan-treated group. However, it must be noted that in ADVOCATE patients receiving induction RTX did not receive maintenance immunosuppression thereafter, which may have influenced the outcome at 12 months. Glucocorticoid-related adverse effects were significantly lower in the avacopan-treated group. The improvement in eGFR over 6 and 12 months was greatest in the avacopan-treated cohort [40, 41]. Participants with eGFR 15–20 mls/min at inclusion had the greatest renal function improvement [40] suggesting those with severe renal disease may benefit most from avacopan. In the trial, limited GC (tapered over 4 weeks) were given to the avacopan-treated group (Table 3). Guidance is the same for children, with post-pubescent individuals treated with the same dose as adults.

The new BSR recommendation on the use of avacopan is in line with EULAR 2022 [10] and both the National Institute for Health and Social Care Excellence (NICE) [42] and the Scottish Medicines Consortium advise [43] as an option use of avacopan in patients with severe active GPA and MPA in conjunction with CYC or RTX. There are no specific statements on avacopan use in the ACR guideline as it was not approved at time of writing. KDIGO consider avacopan use in induction therapy [8].

As there is no data on the use of avacopan beyond one year of therapy, the recommended duration of avacopan therapy is not currently well defined and dictated by local/regional policies in the UK.

Maintenance of remission treatment for GPA and MPA

Recommendation 6a	Following induction of remission with an RTX or CYC-based treatment regimen, we recommend maintenance of remission with RTX in preference to other agents (GRADE 1A, SoA 98%).
Recommendation 6 b	Maintenance rituximab should be administered at a dosing range of 500 mg to 1000 mg every 4–6 months (GRADE 1A, SoA 97%).
Recommendation 6c	Azathioprine (AZA) or MTX may be considered as alternative options (GRADE 1A, SoA 98%).
Recommendation 6d	MMF is an option only where there is intolerance, or a contraindication, to RTX, AZA or MTX (GRADE 2B, SoA 97%).

Table 3. Glucocorticoid (GC) weaning regimens: reduced dose GC tapering regimen in the PEXIVAS trial [16], LOVAS trial [34] and an optional steroid regimen in patients on avacopan

	Prednisolone tapering regimen in PEXIVAS trial (mg/d)			Prednisolone tapering regimen in the LoVAS trial	Steroid regimen whilst on Avacopan Optional
Week	<50 kg	50–75 kg	>75 kg		
1	50	60	75	0.5 mg/kg/d	0.5 mg/kg/d
2	25	30	40	0.5 mg/kg/d	
3–4	20	25	30	0.25 mg/kg/d	Taper to 0 by week 4
5–6	15	20	25	7.5 mg/d	
7–8	12.5	15	20	5 mg/d	
9–10	10	12.5	15	4 mg/d	
11–12	7.5	10	12.5	3 mg/d	
13–14	6	7.5	10	2 mg/d	
15–16	5	5	7.5	2 mg/d	
17–18	5	5	7.5	1 mg/d	
19–20	5	5	5	0	
21–22	5	5	5	0	
23–52	5	5	5	0	
>52	Investigators local practice			0	

Following induction of remission, most individuals living with GPA or MPA should receive maintenance of remission therapy with an appropriate agent. Since the 2014 BSR guideline, the MAINRITSAN and RITAZAREM clinical trials have shown RTX to be superior to AZA for the prevention of relapses [44, 45]. Long-term follow-up of the MAINRITSAN participants further confirmed reduced relapse rates and reduced toxicity amongst those treated with RTX [46]. Following an RTX or CYC-based induction of remission treatment course, RTX is therefore the preferred agent for maintenance of remission. This is in accordance with the recently published EULAR and ACR guidelines [7, 10]. KDIGO suggest using rituximab or azathioprine [8].

There are several RTX dosing schedules used for maintenance of remission including fixed schedules (500 mg 6-monthly in the MAINRITSAN trials and 1 g 4-monthly in RITAZAREM) as well as tailored dosing based on serum ANCA level, and B-cell repopulation. A tailored dosing schedule was shown to be similar in efficacy compared with a fixed dosing schedule in the MAINRITSAN2 trial [47]. However, the recently published pooled analysis of the MAINRITSAN trials has shown that the rate of major relapse in the 18-month individually tailored RTX group was significantly higher than the 18-month fixed-dosing RTX group [48]. A further study has suggested that re-dosing RTX based on B-cell repopulation compared with re-dosing based on a rise in ANCA titre resulted in fewer clinical relapses; however, individuals had to have received at least 24 months scheduled 6-monthly rituximab treatment prior to inclusion in the trial [49]. The WG recommend a fixed dosing range of 500 mg to 1000 mg every 4–6 months in agreement with EULAR, ACR and KDIGO.

AZA and MTX are alternative maintenance of remission options where rituximab is not used. The WEGENT trial and the 10-year follow-up analysis have shown AZA and MTX to be equivalent in their efficacy in maintaining remission [50, 51].

MMF was associated with an increased risk of relapse when compared with AZA in a head-to-head RCT [52]. Therefore, the use of MMF as a maintenance of remission agent should be limited to those patients that are intolerant to, or have a contraindication to RTX, AZA and MTX.

Maintenance of remission for GPA and MPA: duration of immunosuppression

Recommendation 7a	Maintenance of remission should be continued for a period of 24–48 months (GRADE 1A, SoA 97%).
Recommendation 7 b	People living with severe renal involvement who remain dialysis dependent have a high risk of infection. Patients with renal limited disease who remain dialysis dependent may not require ongoing immunotherapy. Maintenance of remission therapy to prevent relapse should be balanced against the risks of immunosuppression (GRADE 2C, SoA 98%).

The optimum duration of maintenance therapy remains uncertain. The REMAIN trial compared 48 months *vs* 18–24 months of AZA maintenance treatment with improvement in relapse rate and renal survival but not mortality with longer treatment duration at the expense of increased severe adverse events [53].

The MAINRITSAN3 trial investigated the efficacy of prolonged RTX maintenance therapy in GPA and MPA [54]. In this small trial, the extension of 6-monthly RTX infusions for an additional 18 months following an initial 18-month maintenance of remission course reduced the risk of relapse at 28 months with no observed increase in toxicity [54]. However, the recently published pooled analysis of the MAINRITSAN maintenance of remission trials has shown that at a longer-term follow-up of 84 months, there was no improvement in overall and major relapse-free survival in the 36-month compared with the 18-month RTX group [48]. The authors of the pooled analysis suggested that 36-month RTX therapy should be reserved for patients at high risk of relapse [48].

We recommend that maintenance immunosuppressive therapy should be continued for 24–48 months. Decision making around the length of treatment should consider patient risk factors for relapse and infection as well as patient preferences. A longer duration may be considered in frequently relapsing disease, but this should be balanced against the risks of continued immunosuppression. This agrees with recommendations from EULAR 2022 and KDIGO update 2024 which suggest remission-maintenance treatment to continue for at least 18 months and consideration in those at higher risk of relapse to continue for longer [8, 10]. The ACR recommendation is ungraded and suggests duration of maintenance therapy to be guided by the patient's clinical condition, preferences and values [7]. KDIGO also suggests that patients with MPO-AAV who achieve remission after induction may not require maintenance therapy due to low rates of relapse [8]. The WG agreed there was insufficient evidence to endorse this statement and make no differentiation on duration of maintenance immunosuppressive therapy between MPA and GPA or ANCA types.

Retrospective cohort analyses have shown that dialysis-dependent patients have higher rates of infection and lower rates of relapse compared with the same patients pre-dialysis or patients with preserved kidney function [55–57]. Maintenance of remission may not be required in people with lived experience of renal limited disease who remain, or subsequently become, dialysis-dependent. In individuals with lived experience of dialysis dependency and prior evidence of extra-renal manifestations, consideration of continuation of treatment to prevent relapses should be balanced against the risks of immunosuppression. This agrees with KDIGO expert opinion [8].

Maintenance of remission for GPA and MPA: duration of GC treatment

Recommendation 8	The optimum length of treatment with GC during the maintenance phase is uncertain. Depending on concurrent immunosuppression, complete GC withdrawal may be possible within 6–12 months following induction of remission treatment (GRADE 2B, SoA 98%).

The optimum length of treatment with GC, where not used with avacopan, is uncertain. A meta-analysis published in 2010 showed that early GC withdrawal, within 12 months of

diagnosis, was associated with an increased relapse rate [58]. More recently, preliminary results from the TAPIR trial have suggested that complete withdrawal of GC at 6 months following induction of remission was not associated with major relapses [59], although there were more minor relapses noted in the withdrawal group. In patients who received RTX for induction of remission, there was no difference in total relapse rate. It is important to caveat that these preliminary results have been presented in abstract form but not yet published in full (TAPIR study, trial number NCT01940094) [59]. Nevertheless, these data suggest that depending on concurrent immunosuppression, complete GC withdrawal may be possible within 6 months following induction of remission treatment. EULAR and ACR guidelines note there is limited evidence to recommend duration of GC therapy during remission-maintenance therapy and should be personalized to patient requirements [7, 10].

Timing of kidney transplant in GPA and MPA

Recommendation 9	People living with GPA or MPA should be in stable clinical remission for at least 6–12 months prior to receiving a kidney transplant (GRADE 2C, SoA 98%).

Despite significant advances in the treatment of AAV, 20–25% of individuals living with GPA or MPA will reach ESKD within a few years of diagnosis [60]. Several reports have demonstrated that kidney transplantation confers a significant survival benefit for individuals living with AAV and ESKD [61–63]. In addition, graft survival in AAV is at least equivalent, and in some reports improved, when compared with graft survival in other non-diabetic ESKD diagnoses [63–65]. However, timing of transplantation should be considered. A report of outcomes amongst 107 kidney transplant recipients living with AAV-related ESKD showed that transplantation within 12 months of remission was associated with increased mortality [66]. However, a subsequent report did not find any association between the timing of transplantation following remission and mortality or graft survival [67]. There are conflicting reports on the association of ANCA seropositivity at the time of transplant and risk of relapse [66, 68, 69]. With current immunosuppression treatment regimens, relapse rates post-transplant are very low [67]. We recommend that people with lived experience of GPA or MPA should be in stable remission for at least 6–12 months before receiving a kidney transplant. The presence of circulating ANCA should not delay transplantation provided the disease is in stable remission. Recommendations are in line with KDIGO [8].

Domain 2: Subglottic stenosis and ear, nose and throat disease recommendations

Guidance for the management of specific features of AAV were not included in the 2014 BSR guidance and the ACR 2021, EULAR 2022 and KDIGO 2024 update guidelines do not provide detailed recommendations on specific disease manifestations beyond kidney disease [7, 8, 10]. Although there are no randomized controlled trials to guide therapy, the WG recognized that subglottic stenosis and sino-nasal disease are particular areas of challenge in disease management.

GPA-related subglottic stenosis diagnostic considerations

Recommendation 10	Airway symptoms (exertional dyspnoea, stridor) associated with GPA should be investigated by an Ear, Nose and Throat (ENT) and/or Respiratory specialist with expertise in vasculitis and airway stenosis (GRADE 1C, SoA 99%).

Subglottic stenosis (SGS) develops in 10% to 23% of people living with GPA, while endobronchial stenosis is reported in 6% [70, 71]. SGS is more commonly seen in women (70.5%) while systemic GPA has an equal gender distribution [70]. GPA-related airway stenosis can lead to life-threatening complications and require emergent airway management and occasionally a tracheostomy.

Idiopathic subglottic stenosis (iSGS) is a rare fibrotic disease of yet unknown aetiology causing stenosis of the subglottic and upper tracheal airway [72]. While a diagnosis of exclusion, this patient group has a homogeneous demographic, presentation phenotype and histopathological findings suggesting a unique disease entity. Individuals living with iSGS are almost exclusively Caucasian females presenting between the fourth and sixth decade of life [72].

While probably two different aetiologies, it is sometimes difficult to distinguish iSGS and GPA-related SGS as 7–30% of the latter are ANCA negative [70, 71] and ANCA status can change after many years of negative serology. GPA-SGS, unlike systemic GPA, can be frequently associated with MPO-ANCA positivity, while systemic GPA is more commonly associated with PR3-ANCA [73, 74]. Therefore, patients with iSGS with atypical demographic features, inflamed airway or regular dilations of <6 monthly intervals may benefit from referral to a vasculitis clinic for further investigations and consideration of a vasculitis diagnosis.

Systemic and surgical treatment options in subglottic stenosis

Recommendation 11	GC therapy can help reduce inflammation in GPA-sub-glottic stenosis but is not the preferred option for maintenance therapy. More significant disease requires induction and maintenance therapy following the recommendations for systemic GPA and MPA treatment (GRADE 1C, SoA 98%).

While there is a lack of specific evidence for the management of GPA-related airway stenosis, concurrent endoscopic surgical intervention and systemic immune suppression lead to improved outcomes with reduced frequency of surgical interventions and tracheostomy insertion [71].

Endoscopic dilation with concurrent intralesional long-acting GC injection is the mainstay of surgical management

of GPA-related SGS [70]. In-office serial intralesional steroid injection (SILSI) has been gaining traction in recent years for the management of SGS including GPA-related despite limited evidence [75, 76]. The potential benefit of adjuvant SILSI treatment should be balanced against side effects when making treatment decisions [75].

Medical management has not been specifically assessed in this group of people living with GPA-SGS, although patients with SGS were not excluded from clinical trials; in general people living with GPA and SGS should be treated with standard induction regimens. However, it is recognized that MPO-positive GPA-SGS disease may require less immunosuppression for disease control [74] and the course of SGS may occur independently of systemic disease control [77]. The majority with GPA-SGS have been treated with combinations of GC and/or immunosuppressants and surgical therapy [77]. High-dose GC therapy is associated with reduced likelihood of failure of endoscopic therapy [78]. However, medical therapy alone is generally insufficient, with local endoscopic therapy frequently required in the majority of patients despite use of GC and immunosuppressants [79]. It does seem that combination of immunosuppression and dilatation can reduce the overall incidence of subglottic stenosis dilatation [74]. Addition of sirolimus may help reduce GC requirements in some patients [80], while single agent RTX or CYC-based therapies are associated with a significant reduction in need for surgical re-intervention [81].

The recommendation for treating with immunosuppressive therapy over surgical dilatation with intralesional GC alone in GPA-SGS is consistent with the ACR 2021 recommendations [7].

Sino-nasal GPA recommendations

Sinonasal disease is common (more so in GPA than MPA) and results in significant morbidity and reduced quality of life [82, 83]. The presence of sinonasal disease may precede a formal diagnosis of AAV by months to years [84, 85], thus established damage is often evident at time of diagnosis. Sinonasal disease is associated with relapse and treatment refractory disease [86–88].

Nomenclature in sino-nasal GPA

Recommendation 12	The term 'limited GPA' may underestimate disease burden; terms such as ENT-localised or sino-nasal GPA are preferred (GRADE 1C, SoA 97%).

The term 'limited GPA' has been used to describe disease restricted to the upper airways. This does not reflect the significant symptom burden experienced by people living with GPA restricted to the upper airways, may underestimate disease severity and potentially risks undertreatment. People living with GPA first presenting with 'limited' features may progress to systemic disease and require monitoring for disease evolution to other organs [85, 89]. 'Limited GPA' should be avoided, and terminology that specifies the location of organ involvement and the degree of severity of the disease in each of these locations is preferred. We have used the term sino-nasal GPA throughout this document.

Integrated multi-disciplinary assessment in sino-nasal GPA

Recommendation 13	All people living with AAV affecting the sino-nasal tract should be offered multidisciplinary assessment that includes input from ENT surgeons and physicians experienced in the management of AAV (GRADE 1C, SoA 98%).

Persistent or 'refractory' sino-nasal symptoms are common; these may reflect disease activity, established damage and/or superadded infection. Careful multidisciplinary assessment is required to determine diagnosis, disease activity and extent, related damage, the role of immunosuppression, antibiotics, and topical treatments, and timing of surgical interventions. A number of mimics should be considered during assessment (Table 4, with suggested investigations Table 5). There is evidence of benefit for use of combined ENT-medical vasculitis clinics [90, 91] in managing people living with sino-nasal GPA.

Table 4. Sino-nasal disease mimics

Malignancy	NK-T-cell lymphoma
Infection	Invasive fungal infection Tuberculosis
Other autoimmune or inflammatory disease	Relapsing polychondritis Sarcoidosis IgG4-related disease
Drug-associated vasculitis	Cocaine-induced midline destructive lesion (CIMDL) [92] • Localized areas of inflammation and ischaemia due to the direct irritant and ischaemic effects of inhaled cocaine, leading to tissue necrosis and midline destruction of bone, cartilage and soft tissue • Anterior septum and nasal tip involvement predominates, often sparing the sinuses • Destruction of palate, columella and alar loss are more typical of CIMDL than GPA • Systemic features typically absent • ANCA may be present (often by IIF, with atypical or discordant patterns; NE-ANCA frequent); however, presence or serotype of ANCA does not appear to affect disease phenotype [93] Levamisole-induced vasculitis (LIV) [93, 94] • Systemic vasculitis induced by levamisole, anthelmintic cut with cocaine • Frequent cutaneous involvement • Organ-threatening disease may feature • Dual positivity for PR3-/MPO-ANCA suggestive but not pathognomonic

ANCA: anti-neutrophil cytoplasmic antibody; CIMDL: cocaine-induced midline destructive lesion; GPA: granulomatosis with polyangiitis; IgG4: immunoglobulin G4; IIF: indirect immunofluorescence; LIV: levamisole-induced vasculitis; MPO: myeloperoxidase; NE: neutrophil elastase; NK: natural killer; PR3: proteinase 3.

Table 5. Recommended investigations for suspected or persistent AAV with sino-nasal involvement

Microbiology	• Nasal swabs for detection of infection and carriage of Staph aureus • Enable directed antibiotic use in cases of active infection
ANCA Serology	• Sino-nasal GPA may be ANCA negative (20–30% [85]) • Dual MPO-/PR3-ANCA positivity is common in levamisole-induced vasculitis (LIV) • Neutrophil elastase (NE)-ANCA are a feature of cocaine-induced midline destructive lesion (CIMDL) and LIV but not routinely available
Toxicology	• Urine toxicology for cocaine and metabolites is widely available • Testing is sensitive within 72 hours of drug use (plasma half life is short), but may remain positive for up to 14 days depending on use • Hair or fingernail analysis may detect drug consumption within last 3 months, but are not widely accessible • Toxicology screening should be offered at diagnosis in all patients, and repeat testing considered in those with destructive nasal lesions and treatment-refractory disease
Imaging	• Sinus CT is frequently abnormal and may assist diagnosis though findings often non-specific (and shared with chronic rhinosinusitis) • Assessing the extent of sinus involvement may aid treatment decision-making • May assist in distinguishing EGPA (widespread mucosal thickening, polyposis) from GPA (thin bone erosion & osteitis more typical) • Erosion of hard palate is not typical of GPA and suggests alternative diagnosis (NK-T-cell lymphoma, CIDML) • Interval imaging has limited role in assessing response to treatment in GPA
Nasal biopsy	• May provide histological confirmation of disease (necrosis, vasculitis, granulomata) • Relatively non-invasive • Has poor positive predictive value (>50% non-diagnostic—non-specific chronic inflammation), may reflect suboptimal sampling [96–98] • May have a role in exclusion of malignancy and invasive fungal disease

ANCA: anti-neutrophil cytoplasmic antibody; CIMDL: cocaine-induced midline destructive lesion; CRS: chronic rhinosinusitis; CT: computerised tomography; EGPA: eosinophilic granulomatosis with polyangiitis; GPA: granulomatosis with polyangiitis; LIV: levamisole-induced vasculitis; MPO: myeloperoxidase; NE: neutrophil elastase; NK: natural killer; PR3: proteinase 3.

Recognition of cocaine-associated vasculitis conditions

Recommendation 14	History of previous and current cocaine use should be assessed (both drug history and toxicology) at baseline in all individuals suspected of having AAV and repeat toxicology screening considered in patients with persistent or refractory sino-nasal disease (GRADE 1B, SoA 99%).

Cocaine use is increasingly common in the UK; almost 3% of 16–59-year-olds use it [95]. There are two recognized cocaine-associated vasculitis syndromes (Table 4) although it should be appreciated that these may overlap, that cocaine use may serve as a trigger or potentiating factor for systemic vasculitis that is indistinguishable from 'idiopathic' GPA or MPA, and that incidental cocaine use may occur in those with primary forms of AAV. Abstinence from cocaine use is central to management. Professional support for abstaining from substance use should be offered. Glucocorticoid or immunosuppressive treatment may be required acutely (e.g. with evidence of organ-threatening disease) or in cases of persistent disease despite confirmed abstinence from cocaine. However, use of immunosuppression is often futile with persistent use of cocaine [13].

Treatment for sino-nasal disease

Recommendation 15a	Immunosuppression: first line treatment with RTX or CYC and GC or avacopan for organ-threatening ENT disease is recommended as this provides early disease control and limits accrual of damage (GRADE 1A, SoA 98%).
Recommendation 15 b	Topical and local symptomatic treatments should be offered to people living with sino-nasal disease (GRADE 1B, SoA 99%).
Recommendation 15c	Screening for bacterial carriage and infection, and antimicrobial treatment where indicated, should be offered to people living with sino-nasal disease (GRADE 1C, SoA 98%).

The BSR recommendation is in agreement with the EULAR 2022 guideline recommending first-line treatment with RTX or CYC and GC or avacopan for sino-nasal GPA, as this provides early disease control and limits accrual of damage [10]. The use of alternative agents such as MTX or MMF is associated with longer time to remission, higher relapse risk and greater GC burden [85].

All patients with sino-nasal disease should be offered topical and local symptomatic treatments [99]. Nasal irrigation with saline and application of glycerin- or oil-based lubricants may aid dissolution of crusts, providing symptomatic benefit and removing foci of infection in the nasal tract [99]. Use of intranasal glucocorticoids may reduce nasal inflammation and associated symptoms and can be continued long term if a 6-week course proves beneficial [99].

Chronic nasal *Staphylococcus aureus* carriage may identify GPA patients with high relapse rate [100], though causality is not clear. One randomized controlled trial demonstrated that *full-dose* treatment with co-trimoxazole was superior to placebo in preventing relapses in GPA patients in remission after therapy with CYC and GC and reduced infections [101]. However, a recent meta-analysis [102] and systematic review [103] have found that co-trimoxazole is not associated with a reduction in relapse risk. Co-trimoxazole is not recommended alone to prevent disease relapse but may be useful for infection-associated sinonasal disease activity. This treatment regimen is distinct from prophylactic co-trimoxazole for prevention of *Pneumocystis jirovecii* pneumonia (PJP) and other infections. Topical antibiotic treatment may be recommended to prevent or treat secondary infection in a rotating antibiotic application. Directed antibiotic therapy should

be used in suspected active sino-nasal infection, which may be evident by increase in sinus pain, crusting, purulent or blood-stained discharge, in association with positive microbiology (e.g. for Staph, Strep, Pseudomonas spp.).

Timing of sino-nasal reconstructive surgery

Recommendation 16	It is essential that disease is in remission for at least 12 months (and desirable that maintenance prednisolone dose is ≤5 mg) at time of reconstructive surgery, otherwise high failure and complication rates are frequently observed (GRADE 1C, SoA 97%).

Timing of nasal reconstructive surgery in patients with GPA depends on several factors including local and systemic disease activity, the severity of nasal deformities, and treatment. Active inflammation increases the risk of complications including poor wound healing, infection and progression of deformity post-operatively [104, 105]. Close collaboration between AAV-experienced physicians and ENT surgeons is therefore required to individualize assessment and balance risks and benefits of surgery. Based on experience and expert opinion, we recommend that disease should be in sustained remission for 12 months on a maintenance dose of prednisolone </=5 mg before performing reconstructive surgery. In patients receiving additional maintenance immunosuppression, this should generally be maintained rather than attempting treatment withdrawal in the peri-operative period given the risk of potential flare [106, 107].

Domain 3: EGPA management

EGPA, formerly known as Churg–Strauss syndrome, is a rare complex chronic disease characterized by asthma, sino-nasal disease, peripheral and tissue eosinophilia, granulomatous inflammation and necrotizing vasculitis of target organs. The WG largely support the latest existing recommendations on the diagnosis and management of EGPA produced by the ACR, EULAR and the European EGPA study group (EESG) [7, 9, 10].

Recommendation 17	A diagnosis of EGPA should be considered in any individual with a combination of asthma (especially of adult-onset), chronic rhinosinusitis (with or without nasal polyposis) and peripheral eosinophilia typically $\geq 1 \times 10^9$/L who develop end-organ involvement (GRADE 1C, SoA 99%).

Symptoms of the upper and lower respiratory tract predominate in people living with EGPA [4, 7, 9, 10, 108]. The onset of asthma is usually seen in adulthood and chronic rhinosinusitis, frequently with nasal polyps requiring surgery, and which typically predates the diagnosis by several years, although the evolution of the disease is highly variable. A peripheral blood eosinophilia (>10% of the circulating white cell count or $>1 \times 10^9$/l) is almost universally observed at the time of diagnosis or historically. Additional symptoms and signs (listed in Table 6 and categorized by organ/tissue type) should raise the suspicion of EGPA.

Table 6. Main clinical manifestations of EGPA

Lung	Asthma, lung infiltrates, pleural effusions
ENT	Chronic rhinosinusitis with or without nasal polyposis
Cardiac	Peri/myocarditis, cardiomyopathy, myocardial infarction
GI	Gastro-oesophageal reflux disease, gastritis, bowel vasculitis
Skin	Petechiae, purpura, urticaria
MSK	Non-erosive inflammatory arthritis, arthralgia, myalgia
Renal	Haematuria, proteinuria, eosinophilic tubulointerstitial nephritis, rapidly progressive glomerulonephritis
Neurological	Peripheral neuropathy, mononeuritis multiplex
Systemic	Malaise, weight loss, fever, sweats

ENT: ear, nose, and throat; GI: gastrointestinal; MSK: musculoskeletal.

Recommendation 18	The diagnosis of EGPA can be challenging due to the heterogeneous clinical phenotype and requires a specialized multi-disciplinary approach to exclude alternative eosinophilic syndromes – MDT discussion and consensus are encouraged when ratifying the diagnosis of EGPA (GRADE 1C, SoA 99%).

People living with suspected EGPA should be evaluated in a systematic, integrated, multi-disciplinary team (MDT) environment involving clinicians with expertise in AAV. Involvement of main target organs, including but not limited to the upper and lower respiratory tract, heart, kidneys, skin and neurological system should be actively screened with a detailed history, blood tests, imaging and histopathology recommended in most cases. A list of 'essential' and 'desirable' investigations is outlined in Table 7, adapted from the European EGPA Study Group recommendations [9, 109].

Hypereosinophilic syndromes (HES) share overlapping clinical features with EGPA and should be actively excluded as part of the diagnostic evaluation as the management can be quite different. The involvement of haematologists in individuals who do not respond to a diagnostic trial of oral GC should be considered. We strongly advocate molecular investigations and a bone marrow assessment as directed by established guidelines in conjunction with haematology in these individuals [9, 109].

New classification criteria for EGPA were developed and validated in 2022 [4]. These were not created for the purpose of making a diagnosis of EGPA and thus should not be used as such. It should be noted that ANCA are only positive in ~40% of EGPA cases, and more often in individuals with renal and neurological manifestations [110].

Recommendation 19	Management of EGPA should be stratified according to clinical manifestations and disease severity (GRADE 1C, SoA 99%).

Table 7. Proposed algorithm for investigation of suspected EGPA

Essential investigations		**Desirable investigations guided by clinical context**	
Investigation(s)	**Rationale**	**Clinical context**	**Investigation(s)**
Routine phlebotomy		Lung infiltrates	Sputum MC&S BAL
FBC, blood film	General haematological screening, assess for dysplastic eosinophils ± blasts	ENT abnormalities, e.g. sinusitis, nasal polyps, hearing loss etc.	Audiometry Nasal endoscopy CT sinuses
U&Es, LFTs	End-organ damage screening	GI symptoms, e.g. reflux, dysphagia, bleeding etc.	Endoscopy Faecal calprotectin
ESR, CRP	Systemic inflammation screening	Kidney abnormalities, e.g. renal impairment, active urinary sediment etc.	Renal ultrasound Renal biopsy
Troponin, BNP	Cardiac involvement screening	Skin abnormalities, e.g. petechiae, purpura, urticaria, rash etc.	Skin punch biopsy
LDH, Tryptase, Vitamin B12	Myeloproliferative disorders screening	Cardiac abnormalities, e.g. arrhythmia, ischaemic changes on ECG, signs of heart failure, elevated cardiac biomarkers etc.	24-h Holter Cardiac MRI (preferred to echocardiography if available) CT coronary arteries (with calcium scoring) Endomyocardial biopsy
Immunology		Neurological abnormalities, e.g. peripheral neuropathy, mononeuritis multiplex, focal neurological deficit	Nerve conduction studies MRI brain ± spine Lumbar puncture
ANCA, anti-MPO, anti-PR3	AAV immune biomarkers	Haematological abnormalities, e.g. cytopenia, eosinophilia $>1.5 \times 10^9$/L not suppressed by corticosteroids, organomegaly, elevated LDH, tryptase, vitamin B12	Bone marrow aspirate and trephine Lymphocyte phenotyping T-cell immunophenotyping TCR rearrangement studies FIP1L1-PDGFRA, JAK2, BCR-ABL, KIT D816V mutations
		Travel history to areas of high helminthic parasite prevalence/ strong clinical suspicion of helminthic infection	Serology against schistosomiasis, filiriasis, strongyloides, fasciola, hydatid, toxocara, anisakis & cover with ivermectin with doses according to the BNF
IgG, IgA, IgM, IgE, IgG4	Related immune markers		
Serology			
HBsAg, HBcAb, HCAb, HIV 1&2	Blood-borne virus screening		
IgE and IgG to A. fumigatus	Fungal screening		
Urinalysis			
Urine dipstick uPCR/uACR	Renal involvement screening		
Faeces			
Stool cultures for ova, cysts and parasites (e.g. *Strongyloides stercoralis*)	Parasitology screening		
Imaging			
CXR, HRCT	Lung involvement screening		
CT sinuses	ENT involvement screening		
Echocardiography	Cardiac involvement screening		
CT abdomen & pelvis	General assessment, solid tumour and organomegaly screening		
Other			
12-lead ECG	Cardiac involvement screening		
Lung function testing FeNO	Lung involvement, asthma screening		

Adapted from Emmi *et al.* and Wardlaw *et al.* with permission [9, 110].
AAV: ANCA-associated vasculitis; ANCA: anti-neutrophil cytoplasmic antibody; anti-MPO: antibodies against myeloperoxidase; anti-PR3: antibodies against proteinase 3; BAL: bronchoalveolar lavage; BMAT: bone marrow aspirate and trephine; BNF: British National Formulary; BNP: brain natriuretic peptide; CRP: C-reactive protein; CT: computed tomography; CXR: chest X-ray; ECG: electrocardiogram; EMG: electromyography; ENT: ear: nose and throat; ESR: erythrocyte sedimentation rate; FBC: full blood count; FeNO: fractional exhaled nitric oxide; FESS: functional endoscopic sinus surgery; HBcAb: hepatitis B core antibody; HBsAg: hepatitis B surface antigen; HCAb: hepatitis C antibody; HIV 1&2: human immunodeficiency virus 1&2; HRCT: high-resolution computed tomography; Ig: immunoglobulin; LDH: lactate dehydrogenase; LFTs: liver function tests; LP: lumbar puncture; MC&S: microscopy, culture and sensitivity; MRI: magnetic resonance imaging; NCS: nerve conduction studies; TCR: T-cell receptor; uACR: urine albumin: creatinine ratio; uPCR: urine protein: creatinine ratio; U&Es: urea and electrolytes.

Table 8. Prognostication and stratification of disease severity in EGPA

Revised 2011 Five-Factor Score [111][a]	Life/organ-threatening manifestations[b]	Non-life/organ-threatening manifestations[c]
Age >65 years	Alveolar haemorrhage	Chronic rhinosinusitis without bony involvement
Cardiac insufficiency	Glomerulonephritis	Asthma
Renal insufficiency	Central nervous system vasculitis	Mild systemic symptoms (malaise, fever, weight loss etc.)
[stabilised peak serum creatinine >150 µmol/L (1.7 mg/dL)]	Mononeuritis multiplex	Uncomplicated cutaneous disease (i.e. without ulceration)
Gastrointestinal involvement	Cardiac involvement	Mild inflammatory arthritis
Absence of ear, nose and throat manifestations (presence is associated with better prognosis)	Mesenteric ischaemia	Myositis (skeletal muscle only)
	Limb/digit ischaemia	Non-cavitating pulmonary nodules

[a] Each factor is weighted as one point. Possible FFS scores range from 0–2:0 = No factors present; 1 = 1 factor present; 2 = ≥2 factors present.
[b] It is not possible for the authors to list every potential manifestation of EGPA, thus clinical judgement is also required.

It is widely acknowledged that treatment of EGPA, like MPA/GPA, should follow the paradigm of remission-induction and maintenance [9, 10]. The intensity of remission-induction treatment should be directed by disease severity. Current disease severity scores in EGPA include the revised prognostic Five-Factor Score (FFS) which stratifies prognosis by age and clinical manifestations at diagnosis as in Table 8 [111].

Disease severity in EGPA can also be defined by the presence or lack of life- and/or organ-threatening manifestations as laid out in Table 8 [7]. The use of these criteria, which include broader disease manifestations than the FFS for severity stratification (e.g. alveolar haemorrhage), are consistent with the ACR 2021, EULAR 2022 and EESG 2022 recommendations [7, 9, 10].

Induction of remission for EGPA

Recommendation 20a	All people living with active (newly diagnosed or relapsed) EGPA should be considered as having potentially life- or organ-threatening disease (GRADE 1C, SoA 99%).
Recommendation 20 b	All people living with active EGPA should be assessed for induction of remission treatment with GC combined with other immunomodulatory agents (GRADE 1C, SoA 99%).
Recommendation 20c	The recommended immunomodulatory options for people with life- or organ-threatening EGPA are intravenous pulse CYC as first line OR RTX if CYC is either contraindicated or not acceptable to the patient (GRADE 1C, SoA 98%).
Recommendation 20d	Anti-IL-5/IL-5R directed therapies (both ligand and receptor) have demonstrated broad efficacy in EGPA and are recommended (if available for any of the licensed indications) for remission induction in non-life or non-organ-threatening disease (GRADE 1A, SoA 98%).
Recommendation 20e	In non-life- or organ-threatening active EGPA, alternative induction therapy with MTX, MMF or AZA may be considered when anti-IL-5/IL-5R is not available or as adjunctive therapy depending on disease phenotype (GRADE 2C, SoA 98%).

The WG support the use of CYC in the presence of life- or organ-threatening active EGPA. Its use is well established in the treatment of EGPA and low-dose regimens have continued to demonstrate good efficacy and safety in this context [112]. To date, the use of RTX in EGPA has only been investigated in one randomized controlled trial, REOVAS, which has thus far been published only in abstract form [113]. The study concludes that RTX induction given as 1 g twice separated by 2 weeks with GC was not superior to conventional remission-induction therapy based on FFS (i.e. GC alone or in combination with CYC). Observational data have demonstrated some efficacy and safety of RTX in EGPA, but frequent relapses still occurred in disease domains closely linked to type 2 inflammation (asthma and ENT disease) [114]. The use of CYC in life- or organ-threatening EGPA is therefore favoured over RTX, although the latter can be used in contexts where CYC is less preferable such as people with childbearing potential, people with a large burden of cumulative CYC and/or strong individual choice. These recommendations are in keeping with those of ACR 2021, EULAR 2022 and the EESG 2022, which recognize the greater body of evidence and experience with CYC use in severe disease.

IL-5 pathway blockade in EGPA was tested in the MIRRA trial, a randomized controlled trial comparing mepolizumab 300 mg every 4 weeks to placebo in non-life and non-organ-threatening disease, and MANDARA, a non-inferiority randomized controlled trial assessing the efficacy and safety of benralizumab (30 mg), a monoclonal antibody directed against the IL-5 receptor (IL-5R) compared with mepolizumab (300 mg), both given 4 weekly [115, 116]. A recent registry study reported no difference in efficacy between mepolizumab at either 100 mg or 300 mg every four weeks, suggesting that the UK-licensed severe asthma dose of 100 mg may be sufficient in EGPA [117]. Currently, in the UK, access to IL-5/IL-5R therapies for EGPA is predominantly via severe asthma pathways in specialist centres, although we anticipate this may change in light of evolving randomized controlled trial and observational data, demonstrating broad efficacy of these agents for attaining remission and GC tapering in EGPA. We therefore recommend use of anti-IL5/IL-5R therapy, where it is available, for remission-induction therapy for non-life or non-organ-threatening active EGPA. This guidance also includes children living with active EGPA as defined by the medicines for children NHS specialized commissioning rules. This is consistent with ACR 2021 and pragmatically aligned with EULAR 2022 and European EWG guidelines

which recommend anti-IL-5/IL-5R therapy for relapsing non-severe disease [7, 9, 10].

In non-severe active EGPA, there are limited data to support the use of alternative immunosuppressive agents for remission-induction. A prospective controlled trial did not show benefit of AZA in addition to GC in non-severe EGPA [118]. No randomized controlled trials are available on the use of MTX or MMF; evidence is limited to small retrospective cohorts [119, 120]. Recognizing that anti-IL-5 therapy may not be available in all cases, we conditionally recommend the use of conventional immunosuppressants alongside GC for non-severe EGPA depending on clinical phenotype (e.g. relapsing or refractory disease). This is aligned with ACR 2021, though contrasts with EULAR 2022 and EESG 2023, which do not recommend use of traditional immunosuppressants for remission induction in non-severe disease [7, 9, 10]. However, the latter guidelines do not make provision for settings where anti-IL-5 therapy is not routinely available for all patients.

Maintenance of remission for EGPA

Recommendation 21a	Anti-IL-5/IL-5R directed therapies are recommended (if available for any of the licensed indications) for maintenance of remission and to aid tapering of GC (GRADE 1A, SoA 99%).
Recommendation 21 b	RTX, MTX, MMF or AZA may be considered as alternative options when anti-IL-5/IL-5R is not available, or as adjunctive maintenance therapies depending on disease phenotype (GRADE 2C, SoA 98%).
Recommendation 21c	GC should be tapered to the lowest possible effective dose whilst maintaining disease remission and considering patient-specific disease manifestations, comorbidities and preferences (GRADE 1A, SoA 99%).

As discussed, two recent randomized controlled trials support the use of anti-IL-5/IL-5R therapy during the remission-maintenance phase in EGPA, where their use was associated with greater time in remission and reduced GC burden [115, 116]. Whilst these trials were conducted in people with non-life- or non-organ-threatening disease, the benefit of anti-IL-5/IL-5R therapy is likely to extend to all patients with eosinophilic respiratory manifestations (asthma, rhinosinusitis); thus we have recommended their use for remission maintenance where available in the UK.

In people with life- or organ-threatening EGPA who have achieved remission after a CYC- or RTX-based induction regimen, we recommend additional immunosuppressive therapy (with or without anti-IL-5/IL-5R) to minimize relapse risk. Whilst there is a lack of high-quality data to guide treatment choice, a similar recommendation is made by ACR 2021, EULAR 2022 and EESG 2023 [7, 9, 10]. Rituximab may be preferable in people who have achieved remission following RTX-based induction [121, 122]. In people living with non-life or non-organ-threatening disease who have attained remission after treatment with GC alongside MMF, MTX or AZA, we suggest continuing immunosuppressant treatment and tapering steroids to the lowest possible effective dose.

The WG agree with current EULAR recommendations for the management of EGPA and propose the following adapted management algorithm, illustrated in Fig. 2 [10].

Domain 4: AAV service specification

The AAV service specification recommendations provide evidence-based service standards to improve quality of care. The 2014 BSR guideline identified several key quality standards around service specification and holistic delivery of care for patients with AAV, based on expert opinion [3], which are also reflected by EULAR 2022 recommendations on overarching principles of care [10]. Since then, evidence is provided by one study that has systematically explored the role of specific service components on patient outcomes. The mixed methods study, VOICES, used survey data regarding service specifications across the UK and Ireland, and qualitative and population-based outcome data from the UK, to provide high-quality evidence describing key care components in AAV, which associated with improved health outcomes [123]. VOICES provides the foundation for the service specification recommendations, as it is the most robust evidence on this topic to date. Recognizing that some service components are interdependent and often co-exist together within care delivery systems, this guidance should be considered as a holistic package to improve the whole care pathway. This includes support for national, regional and local planning and implementation of key service components, acknowledging the different approaches to service commissioning and set-up across different geographical areas.

Timely access to services

Recommendation 22a	Waiting times for individuals with new symptoms and with a high index of clinical suspicion for active AAV to be reviewed by a vasculitis expert should not exceed 7 days from initial referral (GRADE 1B, SoA 97%).
Recommendation 22 b	Children living with suspected AAV should be discussed acutely with tertiary paediatric sub-specialty teams (GRADE 1C, SoA 99%).
Recommendation 22c	Services offering AAV care should be enabled to access intravenous therapy for initial treatment or relapse within 7 days (GRADE 1C, SoA 98%).

Timely response to illness is one of the aspects of healthcare particularly valued by people living with AAV [123]. Waiting times of <7 days for individuals with newly diagnosed disease to be reviewed by a vasculitis expert are associated with fewer serious infections, fewer emergency hospital admissions and reduced mortality [123]. Evidence also suggests that individuals seen in a timely manner have lower cancer and cardiovascular disease rates [123].

This should form part of a fast-track referral pathway that includes efforts to raise awareness amongst colleagues in primary care as what to look for, who to contact if AAV is suspected, and in secondary care, the ability to identify red flags and gaps in pertinent clinical information and undertake appropriate triaging and review. We define a vasculitis expert as someone with up-to-date knowledge in managing a range of people living with vasculitis, as opposed to where someone is physically based.

Children living with AAV should initially be managed under tertiary sub-specialty teams, including paediatric rheumatologists, paediatric nephrologists and other allied sub-specialties

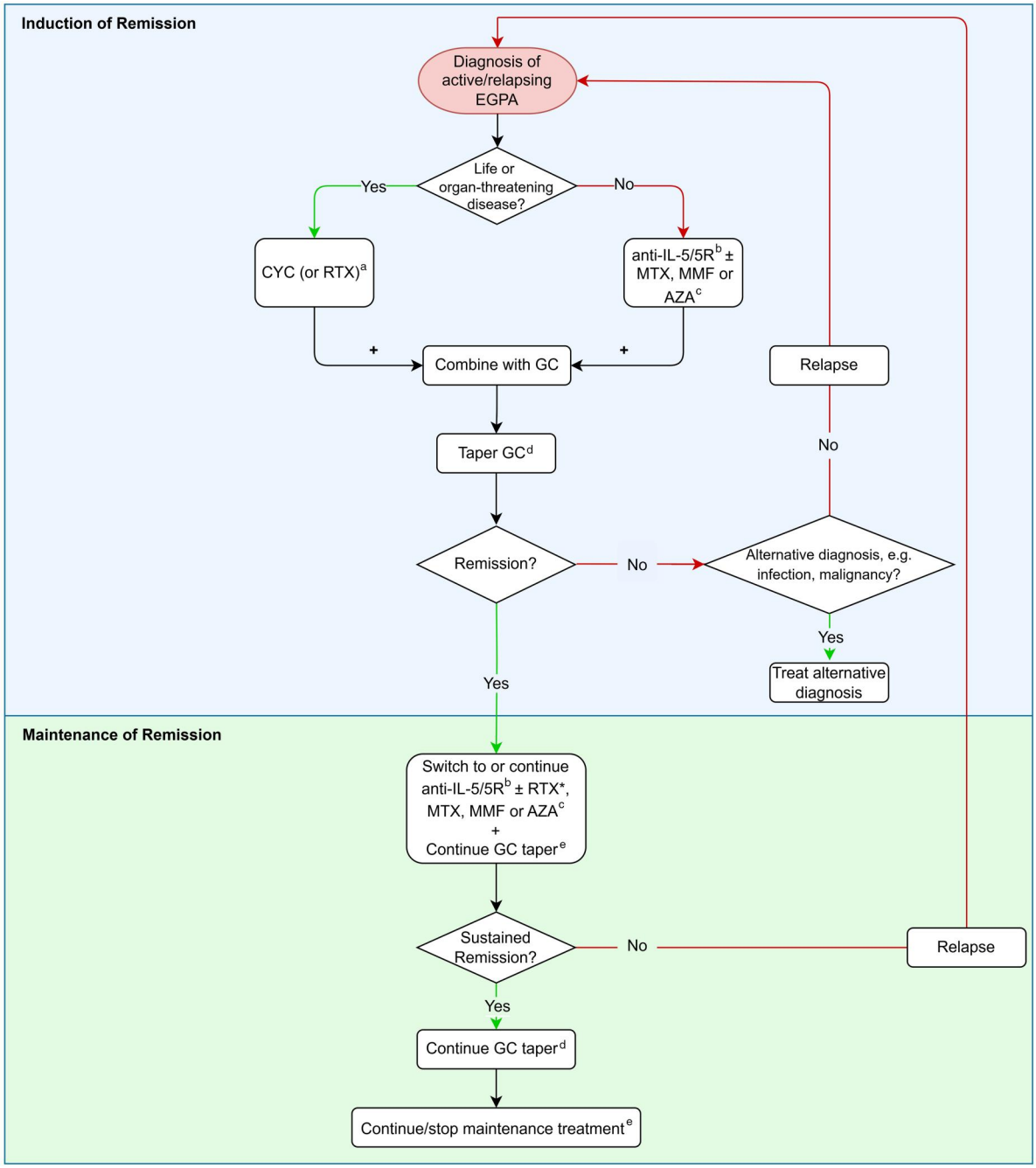


Figure 2. Proposed EGPA management algorithm (adapted from latest EULAR recommendations with permission [10]). [a]RTX is only conditionally recommended in cases of confirmed vasculitic complications (as opposed to type-2 mediated inflammation) when CYC is less preferable (e.g., patients with childbearing potential, previous exposure to a large burden of cumulative CYC and/or strong patient preference). [b]anti-IL-5/5R therapy is recommended if patient meets NICE TA criteria for any of the currently licensed clinical indications (e.g. severe eosinophilic asthma). [c]There is limited data to support the benefit of AZA in EGPA, whilst no RCTs are available on the use of MTX or MMF. [d]GC should be tapered to the lowest possible effective dose whilst maintaining disease remission and considering patient-specific disease manifestations, comorbidities and preferences. A slow gradual taper below physiological doses (approximately prednisolone 3 mg daily) should be attempted in the majority of cases to allow adrenal recovery and avoid long-term adrenal insufficiency. [e]Dependent on specialist assessment of individual disease manifestation(s), comorbidities and patient preferences. Anti-IL-5/5R: anti-interleukin-5/anti-interleukin-5 receptor biologics; AZA: azathioprine; GC: glucocorticoids; CYC: cyclophosphamide; EGPA: eosinophilic granulomatosis with polyangiitis; HES: hypereosinophilic syndrome; MMF: mycophenolate mofetil; MTX: methotrexate; RTX: rituximab

dependent on clinical presentation and organ involvement. Paediatric AAV is rare, therefore expert opinion asserts that initial new presentations should be discussed acutely with relevant teams.

AAV is a potentially organ- and life-threatening disease and, as such, access to urgent intravenous (IV) treatment is necessary. Across the UK, there is significant variation in how services deliver IV treatments [123]. Average wait time for urgent IV treatment was 2.67 days (range 1–10 days), with 18% of services having wait times over 7 days [124]. Specific challenges included the availability of slots and staff, particularly in shared day case units, due to competition with other services and limited awareness of the importance of timely treatment for AAV. This could impact on timing and

treatment decisions (e.g. prolonged GC prescriptions). Access to a dedicated day-case facility was associated with a reduced risk of serious infection, albeit with some uncertainty around the effect estimate [122]. Services offering AAV care should be enabled to access IV therapy for initial treatment or relapse within 7 days to minimize organ damage, especially renal function [124]. Residual renal function is highly predictive of longer-term outcomes [125].

Integrated care

Recommendation 23a	People living with AAV should be cared for in cohorted* rather than general clinics (*where people living with AAV are grouped together and seen in a dedicated clinic) (GRADE 1B, SoA 98%).
Recommendation 23 b	Services for people living with AAV should be coordinated across specialties to deliver timely and effective care. Multi-specialty clinics should form the gold standard (GRADE 1C, SoA 98%).
Recommendation 23c	AAV services should have access to a nurse with specialist knowledge of vasculitis to support care coordination and holistic care. Nurse-led clinics should be implemented, complementing care delivered through cohorted clinics (GRADE 1B, SoA 98%).
Recommendation 23d	Transition from paediatric to adult care should be supported by multi-disciplinary teams with dedicated clinics. Established NICE guidelines on healthcare transition should be followed (GRADE 1C, SoA 99%).

People living with AAV often require support from across the health and social care system and improving care coordination is a key priority of the UK Rare Diseases Framework [126]. Cohorted clinics (where people living with AAV are grouped together and seen in a dedicated clinic) and nurse-led clinics for people living with AAV are both associated with fewer serious infections and emergency admissions to hospital [123]. Joint or parallel clinics with other specialties are associated with fewer serious infections [123]. Although the evidence is less clear, cohorted, parallel or joint clinics may be associated with reduced mortality [123]. Multi-disciplinary care has also been shown to improve outcomes in EGPA [127].

These service components are characterized by their ability to overcome professional tensions between specialties, support continuity of care and timely access to expertise, and for people living with vasculitis, foster a sense of feeling safe [123]. The WG regard multi-speciality clinics as the gold standard in AAV care.

Nurses with specialist AAV knowledge have key skills that are valued by people living with AAV, supporting continuity of care, integration of care across different specialties, and a holistic approach to care. Nurse-led components of care are associated with fewer emergency admissions and reduced risk of mortality [123].

Ongoing care may be shared between secondary and tertiary care paediatric teams as appropriate for the child and their family or carers living with AAV. Transition from paediatric to adult care should follow current established NICE guidelines on healthcare transition, including multi-disciplinary support from specialists across the range of organ involvement [128].

Access to expertise

Recommendation 24a	AAV services should offer access to a nurse advice line to offer patient support and rapid access to advice in between clinic appointments (GRADE 1B, SoA 99%).
Recommendation 24 b	All services looking after patients with AAV should have access to regular specialist MDT meetings (GRADE 1B, SoA 99%).
Recommendation 24c	There should be protected time and administrative support for leadership and attendance at MDT meetings and recording of outcomes (GRADE 1C, SoA 98%).

Patient and provider interviews in VOICES revealed the vital role of specialist nurses in improving access to expertise via advice lines (which provide a single point of access for rapid advice within a supportive service infrastructure), care organization and escalation [123]. Patient access to nurse-led advice lines in AAV is associated with fewer serious infections and fewer emergency hospital admissions [123].

The timeliness, cost of care and adherence to national guidelines is improved for cancer patients managed within an MDT meeting [129] and this approach has been adopted by medical specialties to support care of complex medical patients. In VOICES, a top component of care prioritized by all specialties was access to specialist vasculitis MDT meetings [123]. Individuals looked after in services with access to vasculitis MDT meetings had fewer serious infections and emergency hospital admissions [123].

MDT meetings vary greatly in their structure and function. The WG consider an MDT meeting to be about 'getting the right people in a room together talking' [123]. Certain characteristics are needed with respect to the team, infrastructure, organization and logistics and centring the patient in discussions [123]. Appropriate technology, administrative support, adequate attendance and an assigned chairperson are some of the key factors for an effective cancer MDT meeting [130, 131]. To conduct effective MDT meetings that improve AAV patient care, it is vital that health professionals have the appropriate protected time and administrative support.

Additional recommendations for specialist centres

Recommendation 25a	A specialist centre should provide an overall MDT meeting for the surrounding region (GRADE 1C, SoA 98%).
Recommendation 25 b	A specialist centre should hold an MDT or MDT(s) meetings with a range of appropriate specialties, with identified leads for each specialty (GRADE 1C, SoA 97%).
Recommendation 25c	The regional MDT meeting should have arrangements for specialist centre approval of high-cost drugs with arrangements for local prescribing and administration, where agreed between centre and regional hospitals (GRADE 1C, SoA 98%).
Recommendation 25d	There should be resource in job planning and administrative support for leading and supporting regional MDTs and meetings (GRADE 1C, SoA 99%).

Recognizing that deliverability of all key service components may not be possible in smaller units, the WG agreed it was important to include specific recommendations for specialist centres regarding their role in supporting smaller services, and the resources required to enable them to do so. Specialist centres are defined by NHS England but not elsewhere [132]; however, we would expect specialist centres to provide expert advice and holistic care in an MDT fashion for at least 50 patients with AAV in cohorted clinics, be involved in research and provide support for adoption of new therapeutics.

Regional network delivery is important to ensure equity of access to specialist care for all people living with AAV, improve outcomes and support smaller units across wide geographical areas. Several UK national bodies and initiatives aim to develop specialist regional networks for patients with rare autoimmune conditions, including AAV [133–136]. This includes the provision of MDT meetings, practical support with high-tariff drug prescribing, patient review if needed, and recruitment into clinical studies.

There is limited evidence regarding the impact of regional clinical networks, due to network heterogeneity and challenges in measuring impact. However, fragmented care at more than one organization is associated with increased risk of severe infection, cardiovascular disease, end-stage kidney disease and stroke in rheumatology patients with systemic lupus erythematosus [137]. Observational and quasi-experimental studies indicate that clinical networks can lead to improvement in clinical care [138–141], adherence to guidelines [142, 143], as well as coordination of regional-based initiatives [140, 144, 145], such as educational activities [145], audit and care pathway redesign [142, 144] across a range of conditions. Qualitative studies indicate that availability of sufficient resources, strong leadership, a designated coordinator, a culture of inclusivity and widespread engagement and participation are recurring features of successful networks [145–148].

Domain 5: patient education and support

AAV patient education and support

Recommendation 26a	All adults, children and young people with AAV (and their families and carers) should receive ongoing and tailored information and education about their disease, treatment and side effects, including relapse, support systems, as well as diet and exercise from an appropriately qualified individual or organization (GRADE 1C, SoA 98%).
Recommendation 26 b	AAV impacts on patients' quality of life; psychological support and self-management help should be provided for all patients (GRADE 1D, SoA 99%).

The impact of a diagnosis of AAV is significant and patients need ongoing tailored information and support to help them manage their disease. Patient education is an essential component of the management of AAV. A UK survey found that 60% of people with AAV and autoimmune rheumatic diseases struggle to cope with their disease and 40% felt that they did not have enough information and support from the hospital [149]. There is a clear need expressed by patients for information and education regarding disease process, treatment and side effects, relapse, emotional impact, support systems, health promotion, diet and exercise [149–152]. This should be provided both verbally and as written text [150–152]. Structured educational programmes have been shown to increase patients' knowledge [151]. Provision of education should be tailored to the individual, considering issues such as learning difficulties, disabilities and English language ability [150, 151].

Written information is vital for many people with chronic health conditions who can forget between 40–80% of verbal content during consultations [153, 154]. Written information needs to use clear, easily accessible language. Health professionals should signpost to additional resources for optimum information and support access. This should focus on reputable information sources including online patient groups (e.g. Vasculitis UK, Lauren Currie Foundation, Vasculitis Ireland Awareness), online leaflets and other written information produced by charities (e.g. Versus Arthritis).

Patients need psychological support at diagnosis. Signposting to support groups such as Vasculitis UK, the Lauren Currie Foundation and Vasculitis Ireland Awareness can help with psychological support and empower self-management [155].

AAV shared decision making

Shared decision making is a collaboration that involves a person and their healthcare team working together to reach a joint decision about care and is central to high quality care of patients with AAV. NICE and the Scottish Systemic Vasculitis Managed Clinical Network provide support to improve shared decision making at an organizational and individual level [156, 157].

Research recommendations

AAV patient participation in research and service improvement

Patients in research-active healthcare settings have better outcomes and receive better care, with benefits extending to patients beyond those actively involved in research [158]. Making participation easier through online patient-facing platforms and remote provision of biobank samples is an aspiration for future studies.

Domain 1: GPA and MPA treatment research recommendations

There are several important research gaps in the management of GPA and MPA. Our recommendations highlight a lack of reliable biomarkers to guide treatment decisions and the paucity of clinical trials in children and adolescents living with GPA and MPA. Future work is needed to address these gaps in knowledge.

Further key areas for future research include (i) evaluation of the efficacy and safety of induction regimes using RTX in addition to CYC; (ii) investigation of the optimal dosing and duration of GC therapy, including the effectiveness and safety of intravenous GC; (iii) clarification of the optimal dosing and duration of avacopan therapy, the concomitant use of corticosteroids, and its safety with extended use; (iv) evaluation of re-dosing schedules of RTX in maintenance of

remission regimes and optimal duration; and (v) investigation of immunomodulatory therapies with novel modes of action in AAV (e.g. janus kinase inhibitors).

Domain 2: Subglottic stenosis and ENT disease management research recommendations

Further research into specific therapies aimed at understanding the pathogenesis and treatment of SGS and sino-nasal GPA are required to reduce cumulative morbidity and improve quality of life. Other than ANCA, there is a lack of reliable biomarkers to differentiate GPA-SGS from the idiopathic variant, and as such future work should be directed in this avenue. Additional potential research areas include the investigation of airway stenosis utilizing novel imaging approaches and modalities, defining the optimal dosing and duration intervals of intralesional GC and endoscopic dilation, and evaluation of novel surgical techniques for people living with SGS. There is a need to investigate the efficacy and safety of novel immunomodulatory therapies in both SGS and sino-nasal GPA.

Domain 3: EGPA management research recommendations

There are significant unmet needs in the diagnosis and management of EGPA, and addressing these gaps is crucial. We strongly advocate for the inclusion of every EGPA patient in clinical trials. There is a need to align existing registry data for individuals living with EGPA such as the UK and Ireland Vasculitis Rare Disease Group (UKIVAS) and the National Registry of Rare Kidney Diseases (RaDaR) with stored samples in relevant bioresource facilities. This would facilitate further research on novel biomarkers to aid diagnosis and disease activity assessment, targeted treatments to reduce steroid toxicity and ultimately improve patient-related and clinical outcomes.

Domain 4: Service specification research recommendations

Further research is needed to understand how the key components of care are best delivered in different populations and across different geographical areas, to ensure equitable access to care for all people living with AAV.

Whilst there is evidence of improved outcomes with vasculitis MDT meetings, it remains unclear which individual components are important, and how they interact with other key components of service. There has been limited evaluation of the impact of regional clinical networks. Specifically, it is important to understand:

- What is the relationship between specific clinical network and MDT functions and key patient reported outcomes?
- What individual components of MDT meetings and regional networks are important?
- How best to ensure people living with AAV have their views considered at MDT meetings?

Incorporating key service and experience outcomes into national administrative health datasets, registries and AAV cohorts will provide a framework for rigorous and timely evaluation of different service models across different healthcare contexts and populations and identify optimal combinations of service components.

Domain 5: Patient education and support research recommendations

Further research is needed on the most effective method of delivering patient education in AAV, particularly for patients with low health literacy skills.

Despite the increasing recognition of the role that self-management can play in chronic disease, more must be done to relate what we know of the experiences and needs of patients living with AAV in developing the tailored content of effective self-management programmes.

Shared decision making is acknowledged as an overarching principle benefitting patient care; however, there is limited evidence to support this principle, acknowledged in the EULAR guideline [10]. Small studies are contradictory on impact on clinical outcomes and patient experience.

Audit tool to support equitable access to care and improve health outcomes

The service specification recommendations provide evidence-based service standards associated with improved health outcomes. However, a key finding from the VOICES study was a lack of accurate and timely data to support service planning and monitor improvement efforts across healthcare systems [123]. National collection of key service level metrics across the main specialties providing vasculitis care (rheumatology and nephrology) will provide data to examine local, regional, and national care variation and measure implementation of key service components associated with good quality care for people with AAV.

The cross-specialty nature of AAV presents challenges in achieving this as care does not necessarily follow a single specialty pathway. It is important to complement and avoid duplication of data collected elsewhere. An annual service audit undertaken by regional networks consisting of five key audit metrics is proposed (see Supplementary Data S4, available at *Rheumatology* online).

Conclusion

AAV are complex multi-organ diseases requiring cross-specialty working. These recommendations, in five key domains, provide a framework for all healthcare professionals involved in caring for patients with AAV to provide evidence-based, good-quality care across the life course.

Supplementary material

Supplementary material is available at *Rheumatology* online.

Data availability

Data are provided in the article and its online supplementary materials.

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