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BSR Guideline

British Society for Rheumatology RHEUMATOLOGY OXFORD

BSR Guideline

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

Kathryn Biddle[1,‡], Judith Jade[2,‡], Harold Wilson-Morkeh[3,4,‡], Madura Adikari[5], Chadwan Al Yaghchi[6], Zoi Anastasa[7], Neil Basu[8], Paul Brogan[9,10], Dimitrios Chanouzas[11,12,13], Shouvik Dass[14], David D'Cruz[1], Marcos Martinez Del Pero[15], Emmandeep Dhillon[16], Georgina Ducker[17], Siân Griffin[18], Rosemary J Hollick[19,20], David Jackson[21], Catherine King[11,12,13], Matko Marlais[22,23], Alice Mason[24], Stephen McAdoo[25,26], Devesh Mewar[27], Janice Mooney[28], Eleana Ntatsaki[29], Fiona Pearce[30], Benjamin Rhodes[31], Hitasha Rupani[32], Alan Salama[33], Salman Siddiqui[4,34], Rona Smith[35], Lorraine Harper[11,12,*]; for the British Society for Rheumatology Guideline Steering Group[§]

[1]Department of Rheumatology, Guy's and St Thomas' NHS Foundation Trust, London, UK
[2]Department of Rheumatology, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK
[3]Department of Rheumatology, Imperial College Healthcare NHS Trust, London, UK
[4]National Heart and Lung Institute, Imperial College London, London, UK
[5]Department of Rheumatology, Royal Chesterfield NHS Foundation Trust, Chesterfield, UK
[6]ENT Department, Imperial College Healthcare NHS Trust, London, UK
[7]Expert by Experience, Plymouth, UK
[8]Department of Rheumatology, University of Glasgow, Glasgow, UK
[9]Department of Rheumatology, Great Ormond Street Hospital NHS Foundation Trust, London, UK
[10]Great Ormond Street Institute of Child Health, University College London, London, UK
[11]Department of Nephrology, University Hospitals Birmingham NHS Foundation Trust, Birmingham Health Partners, Birmingham, UK
[12]School of Health Sciences, College of Medicine and Health, University of Birmingham, Birmingham Health Partners, Birmingham, UK
[13]School of Infection, Inflammation and Immunology, College of Medicine and Health, University of Birmingham, Birmingham Health Partners, Birmingham, UK
[14]Department of Rheumatology, Leeds Teaching Hospitals NHS Trust, Leeds, UK
[15]ENT Department, Cambridge University Hospital, Cambridge, UK
[16]Department of Rheumatology, New Cross Hospital, Royal Wolverhampton NHS Trust, Wolverhampton, UK
[17]Department of Rheumatology, Norfolk and Norwich University Hospital, Norwich, UK
[18]Department of Nephrology and Transplantation, University Hospital of Wales, Cardiff, UK
[19]Aberdeen Centre for Arthritis and Musculoskeletal Health, University of Aberdeen, Aberdeen, UK
[20]Department of Rheumatology, NHS Grampian, Aberdeen, UK
[21]Department of Respiratory Medicine, Guy's and St Thomas' NHS Trust, London, UK
[22]Department of Nephrology, Great Ormond Street Hospital for Children, NHS Foundation Trust, London, UK
[23]Department of Nephrology, West Suffolk Hospital, Bury St Edmunds, UK
[24]Department of Rheumatology, University Hospital Southampton, Southampton, UK
[25]Immunology and Immunotherapy Department, Imperial College London, London, UK
[26]Renal and Transplant Unit, Imperial College Healthcare NHS Trust, London, UK
[27]Department of Rheumatology, Liverpool University Hospitals NHS Foundation Trust, Liverpool, UK
[28]School of Health, Education, Policing and Sciences, Staffordshire University, Stafford, UK
[29]Department of Rheumatology, East Suffolk and North Essex NHS Foundation Trust, Ipswich, UK
[30]Clinical Sciences Building, City Hospital Campus, University of Nottingham, Nottingham, UK
[31]Department of Rheumatology, University Hospitals Birmingham NHS Trust, Birmingham, UK
[32]Department of Respiratory Medicine, University Hospital Southampton, Southampton, UK
[33]University College London Centre for Kidney and Bladder Health, Royal Free Hospital, London, UK
[34]Respiratory Medicine Department, Royal Brompton and Hammersmith Hospitals, London, UK
[35]Renal Department, University of Cambridge, Cambridge, UK

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*Correspondence to: Lorraine Harper, Institute of Applied Health Research, College of Medicine and Health Sciences, University of Birmingham, Birmingham Health Partners, Birmingham B15 2TT, UK. E-mail: l.harper@bham.ac.uk

‡K.B., J.J. and H.W.-M. contributed equally.

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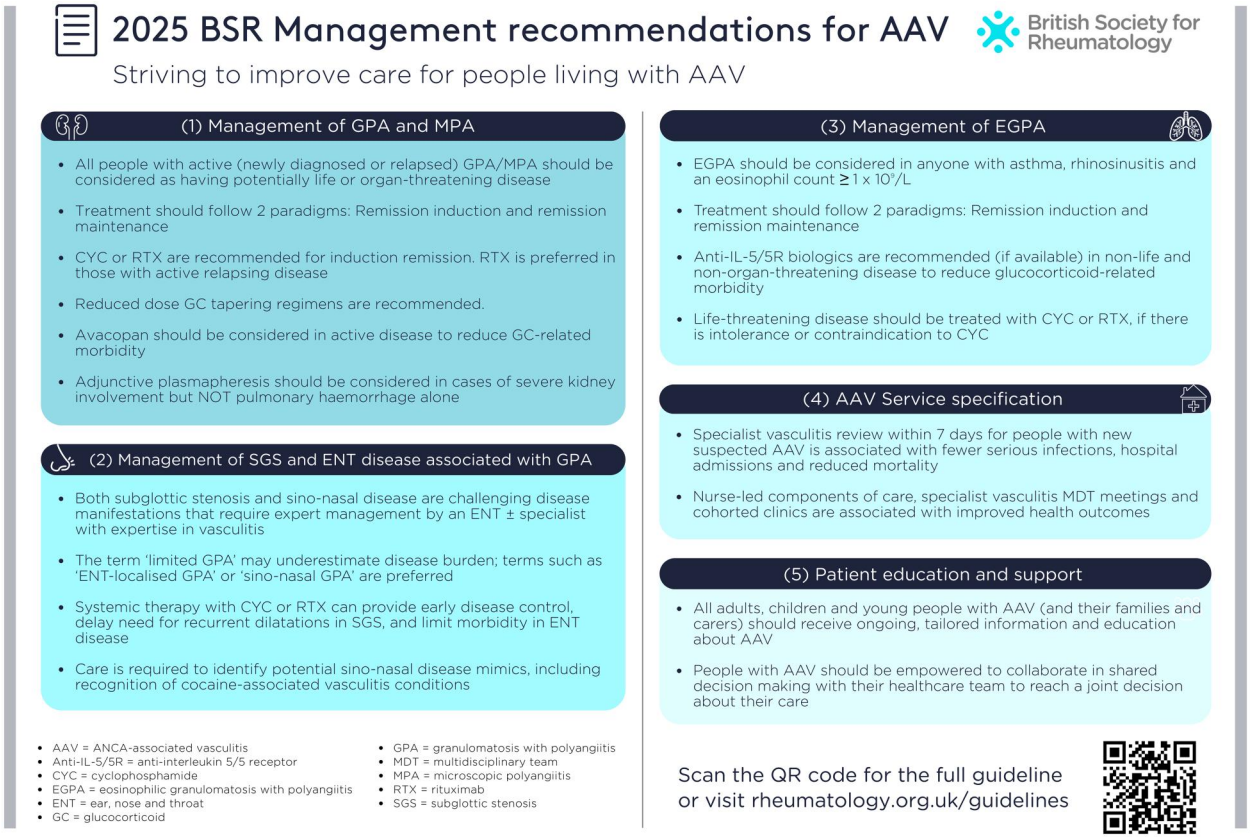
Graphical abstract



Background

Anti-neutrophil cytoplasm antibody (ANCA)-associated vasculitis (AAV) are heterogeneous, multisystem disorders characterized by inflammation and necrosis of small and medium-sized 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 ~25/million population and 200/million, respectively [1]. Despite significant advances in treatment, mortality rates remain elevated, 2.3 times 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.

Need for updated management recommendations

The current British Society for Rheumatology (BSR) Guideline for management of AAV was completed in 2014 and was an important step forward in the management of these complex disorders [3]. It provided a roadmap for best

practice management aiming to harmonize treatment and investigation of AAV.

Updating the guideline is now required to reflect important changes incorporating new studies and trials describing significant advances in treatment and new therapies. The 2014 guideline no longer reflects current best practice and does not reflect all the available high-quality evidence that underpins management of AAV.

Additionally, in line with other BSR guidelines and equality considerations, the updated recommendations now include consideration of people all of ages affected by AAV with specific consideration of the relevance to children and adolescents including transition into adult services.

Objectives

These recommendations were produced by comparing other international society recommendations and updating the review for UK practice. They offer systematic and evidence-based recommendations to support UK clinicians in the management of AAV across the whole life course.

Target audience

The target readership is all clinicians, including primary care, involved in management of people with AAV, and all people living with AAV.

Areas these recommendations do not cover

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.

Significant advances in classification have occurred since the 2014 BSR guideline was produced. However, the classification is not validated for diagnosis or clinical management and is not included in this current guideline [4].

Methods and rigour of development

These recommendations were commissioned by the BSR Guidelines Steering Group (GSG). A working group (WG) was created involving relevant stakeholders. Recommendations in this report were developed, where appropriate, using the BSR Creating Clinical Guidelines Protocol using AGREEII (Appraisal of Guidelines for Research and Evaluation II) methodology. 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 [5–8], 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.

Following a virtual meeting of the full Working Group (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. Each suggested recommendation in the final document was evaluated by all members and subjected to a vote relating to strength of agreement on a scale of 1 [no agreement] to 100% [complete agreement]. The wording of each recommendation was revised until all members were satisfied that they would score at least 80%.

In addition, and in accordance with the BSR protocol, accompanying each recommendation in parentheses is a statement reflecting the strength of recommendation and quality of supporting evidence. Assessment of supporting evidence quality in GRADE reflects confidence in the estimates of benefits, harms and burdens. These recommendations use three levels and a letter (A, B, C) to reflect high, moderate or low/very low quality of evidence. The content and wording of all recommendations were also discussed in order for strength of recommendation to be agreed upon with all members of the WG, assigned as strong (designated as 1) or weak (designated as 2).

The final draft of the recommendations was submitted to the BSR Guidelines Steering Group for stakeholder and internal review and feedback.

Literature search: scope and search strategy

Systematic literature searches were performed separately for each domain by the four subgroups. Where topics or questions had already been considered in previously published international guidelines, the literature search was from December 2021, the end of the European League Against Rheumatology (EULAR) 2022 guideline literature search. For new topics not covered in the BSR guideline 2014 or the published international guidelines [EULAR 2022, American College of Rheumatology (ACR) 2021, Kidney Disease Improving Global Outcomes (KDIGO) 2021 or European Eosinophilic Granulomatosis with Polyangiitis (eGPA) Study Group (EESG) Nature Evidence Based Guideline 2024] [5–8] the literature search was from December 1990. The evidence was drawn from Medline and limited to English language publications. Key terms were agreed within the members of the domain working group. The domain review groups prepared a summary of the quality of evidence following the GRADE approach (https://www.gradeworkinggroup.org/) that informed group review and discussion of the draft recommendations.

Key recommendations

Domain 1: GPA and MPA treatment

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%).

Use of immunosuppressants for GPA and MPA

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%).

2b. 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%).
2c. For active relapsing disease, treatment with RTX is preferred (GRADE 1B, SoA 97%).
2d. A combination of both CYC and RTX can be considered for organ-threatening or life-threatening disease (GRADE 2C, SoA 98%).
2e. Certain patients 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%).

Use of plasmapheresis

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%).
3b. 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%).
3c. Adjunctive plasmapheresis is not routinely recommended for pulmonary haemorrhage without severe kidney involvement (GRADE 1A, SoA 96%).

Glucocorticoid treatment in those not considered for avacopan use

4a. In patients with 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 (GRADE 1B, SoA 96%).
4b. In patients with 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 (GRADE 1B, SoA 97%).
4c. Whilst paediatric patients were not included in PEXIVAS, this tapering regimen can be considered for adolescents. For younger children, the SHARE guidelines for prednisolone tapering could also be considered (GRADE 2C, SoA 96%).
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%).

Use of complement inhibitor avacopan

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 GC (tapering over four weeks) (GRADE 1A, SoA 96%).

Maintenance of remission treatment

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

Maintenance of remission: duration of immunosuppression

7a. Maintenance of remission treatment should be continued for a period of 24–48 months (GRADE 1A, SoA 97%).
7b. 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 relapses should be balanced against the risks of immunosuppression (GRADE 2C, SoA 98%).

Maintenance of remission: duration of GC treatment

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%).

Timing of kidney transplant in GPA and MPA

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

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

GPA-related subglottic stenosis diagnostic considerations

10. GPA patients with airway symptoms (exertional dyspnoea, stridor) 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%).

Systemic and surgical treatment options in subglottic stenosis

11. Glucocorticoid 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%).

Nomenclature in sino-nasal GPA

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

Integrated multi-disciplinary assessment in sino-nasal GPA

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

Recognition of cocaine-associated vasculitis conditions

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%).

Treatment of sino-nasal disease

15a. Immunosuppression – first line treatment with RTX or cyclophosphamide 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%).

15b. Topical and local symptomatic treatments should be offered to people living with sino-nasal disease (GRADE 1B, SoA 99%).

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%).

Surgery for sino-nasal disease

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

Domain 3: EGPA management

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

Induction of remission for EGPA

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%).

20b. 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%).

20c. The recommended immunomodulatory options for patients 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%).

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%).

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%).

Maintenance of remission for EGPA

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%).

21b. RTX, MTX, MMF or AZA may be considered as alternative options when anti-IL-5/IL-5R therapies are not available, or as adjunctive maintenance therapies depending on disease phenotype (GRADE 2C, SoA 98%).

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%).

Domain 4: AAV service specification

Timely access to services

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%) [9].

22b. Children living with suspected AAV should be discussed acutely with tertiary paediatric sub-specialty teams (GRADE 1C, SoA 99%).

22c. Services offering AAV care should be enabled to access intravenous therapy for initial treatment or relapse within 7 days (GRADE 1C, SoA 98%).

Integrated care

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%).

23b. 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%).

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%).

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%).

Access to expertise

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%).

24b. All services looking after patients with AAV should have access to regular specialist MDT meetings (GRADE 1B, SoA 99%).

24c. There should be protected time and administrative support for leadership and attendance at MDT meetings and recording of outcomes (GRADE 1C, SoA 98%).

Additional recommendations for specialist centres

25a. A specialist centre should provide an overall MDT meeting for the surrounding region (GRADE 1C, SoA 98%).

25b. 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%).

25c. The regional MDT meeting should have provision 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%).

25d. There should be resource in job planning and administrative support for leading and supporting regional MDTs and meetings (GRADE 1C, SoA 99%).

Specialist centres are defined by NHS England [10] but not elsewhere; 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.

Domain 5: patient education and support

26a. All adults, children and young people with AAV (and their families and carers) should receive ongoing, 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%).

26b. AAV impacts on patients' quality of life; psychological support and self-management help should be provided for all patients (GRADE 1C, SoA 99%).

Approaches to further research and audit of the recommendations

Potential audit approaches and standards are included in the full Management Recommendations publication.

Supplementary material

Supplementary material is available at *Rheumatology* online.

Data availability

Data can be found in the full management recommendations publication and its Supplementary Material.

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