2025 BSR Management recommendations for AAV

British Society for Rheumatology

Striving to improve care for people living with AAV

(1) Management of GPA and MPA

- All people with active (newly diagnosed or relapsed) GPA/MPA should be considered as having potentially life or organ-threatening disease
- Treatment should follow 2 paradigms: Remission induction and remission maintenance
- CYC or RTX are recommended for induction remission. RTX is preferred in those with active relapsing disease
- Reduced dose GC tapering regimens are recommended.
- Avacopan should be considered in active disease to reduce GC-related morbidity
- Adjunctive plasmapheresis should be considered in cases of severe kidney involvement but NOT pulmonary haemorrhage alone

(2) Management of SGS and ENT disease associated with GPA

- Both subglottic stenosis and sino-nasal disease are challenging disease manifestations that require expert management by an ENT ± specialist with expertise in vasculitis
- The term 'limited GPA' may underestimate disease burden; terms such as 'ENT-localised GPA' or 'sino-nasal GPA' are preferred
- Systemic therapy with CYC or RTX can provide early disease control, delay need for recurrent dilatations in SGS, and limit morbidity in ENT disease
- Care is required to identify potential sino-nasal disease mimics, including recognition of cocaine-associated vasculitis conditions

(3) Management of EGPA

- EGPA should be considered in anyone with asthma, rhinosinusitis and an eosinophil count $\geq 1 \times 10^9$/L
- Treatment should follow 2 paradigms: Remission induction and remission maintenance
- Anti-IL-5/5R biologics are recommended (if available) in non-life and non-organ-threatening disease to reduce glucocorticoid-related morbidity
- Life-threatening disease should be treated with CYC or RTX, if there is intolerance or contraindication to CYC

(4) AAV Service specification

- Specialist vasculitis review within 7 days for people with new suspected AAV is associated with fewer serious infections, hospital admissions and reduced mortality
- Nurse-led components of care, specialist vasculitis MDT meetings and cohorted clinics are associated with improved health outcomes

(5) Patient education and support

- All adults, children and young people with AAV (and their families and carers) should receive ongoing, tailored information and education about AAV
- People with AAV should be empowered to collaborate in shared decision making with their healthcare team to reach a joint decision about their care

- AAV = ANCA-associated vasculitis
- Anti-IL-5/5R = anti-interleukin 5/5 receptor
- CYC = cyclophosphamide
- EGPA = eosinophilic granulomatosis with polyangiitis
- ENT = ear, nose and throat
- GC = glucocorticoid
- GPA = granulomatosis with polyangiitis
- MDT = multidisciplinary team
- MPA = microscopic polyangiitis
- RTX = rituximab
- SGS = subglottic stenosis

Scan the QR code for the full guideline or visit rheumatology.org.uk/guidelines