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2022 UPDATE TO EULAR RECOMMENDATIONS FOR AAV MANAGEMENT: FOCUS ON GPA/MPA

HIGHLIGHTS

- Clear guidance on **GC tapering**, including target dose and duration
- **RTX** a first line option for remission induction in GPA/MPA, **irrespective of disease manifestations** and preferred in relapsing disease
- Inclusion of **avacopan** as strategy to **reduce exposure to GCs** in GPA/MPA
- Recommendation to continue GPA/MPA **remission maintenance** therapies* for **24 to 48 months**
- Role of **patient** education, patient preference and shared decision-making in an **individualised treatment journey**

Diagnosis and assessment

Testing	In addition to biopsies, test for both PR3- and MPO-ANCA† if signs and/or symptoms suspicious of AAV

Remission induction in GPA/MPA

New-onset or relapsing organ/ life-threatening disease	Treat with combination of oral GCs (starting dose: 50–75 mg prednisolone equivalent/day, depending on body weight‡) and RTX§ or CYC
	Consider avacopan in combination with RTX or CYC, as part of a strategy to substantially reduce GC exposure
Non-organ/ non-life-threatening disease	Treat with combination of oral GCs (starting dose: 50–75 mg prednisolone equivalent/day, depending on body weight¶) and RTX (alternatives: MTX or MMF)
	Consider avacopan in combination with RTX, as part of a strategy to substantially reduce GC exposure
RPGN with serum creatinine >300 µmol/L**	Consider plasma exchange (routine use not recommended for alveolar haemorrhage)
GC tapering	Reduce GCs in stepwise fashion to a daily dose of 5 mg prednisolone equivalent/day by 4-5 months, with continued tapering into the maintenance phase
Refractory disease	Thoroughly reassess disease status and comorbidities; consider options for additional or different treatment; manage patients in close conjunction with, or refer to, a centre with vasculitis expertise

*RTX, AZA or MTX (see EULAR recommendations for information on duration of GC and avacopan therapy). †Using high-quality antigen-specific assay. ‡Use of intravenous methylprednisolone at a cumulative dose of 1–3 g on days 1–3 can be considered in patients with severely active disease. §Preferred in relapsing disease. ¶Lowering the starting dose to 0.5 mg/kg/day can be considered in individual patients without organ-threatening or life-threatening manifestations. **Due to active glomerulonephritis. Always refer to the product SmPC for approved indication before prescribing. AAV, ANCA-associated vasculitis; ANCA, anti-neutrophil cytoplasmic antibody; CYC, cyclophosphamide; EULAR, European League Against Rheumatism; GC, glucocorticoid; GPA, granulomatosis with polyangiitis; MMF, mycophenolate mofetil; MPA, microscopic polyangiitis; MPO, myeloperoxidase; MTX, methotrexate; PR3, proteinase 3; RPGN, rapidly progressive glomerulonephritis; RTX, rituximab.

Hellmich B et al. Ann Rheum Dis 2023. doi:10.1136/ard-2022-223764 (Epub ahead of print).

Veeva reference: HQ-AVA-2300047

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Remission maintenance in GPA/MPA

Therapies	Treatment with RTX is recommended (alternatives: AZA or MTX)*
Duration of therapy	In new onset disease, continue RTX (or AZA, or MTX) therapy for 24–48 months; in relapsing disease, or in individuals at increased risk of relapse, consider longer treatment duration, balancing this against patient preference and risk of continuing immunosuppression. Decide to continue or stop maintenance treatment following assessment of individual relapse risk, comorbidities and patient preferences.

Prophylaxis and monitoring

Infection prophylaxis	Use trimethoprim-sulfamethoxazole to prevent *pneumocystis jirovecii* pneumonia and other infections in patients receiving RTX, CYC and/or high-dose GCs
Assessing the need for treatment changes	Base decisions to change treatment strategy on structured clinical assessment (not on ANCA and/or CD19+ B cell testing alone)
Secondary immunodeficiency	Check for secondary immunodeficiency in patients receiving RTX by measuring serum immunoglobulin concentrations prior to initiation of each RTX course

Overarching principles

Patients should be offered best care based on **shared patient–physician decision making** considering efficacy, safety and costs

Patients should be periodically **screened** for treatment-related AEs and comorbidities and provided with **prophylaxis and lifestyle advice** to reduce these

Patients should have **access to education** focussing on the impact of AAV and its prognosis, key warning symptoms and treatment (including treatment-related complications)

AAV requires **multidisciplinary management** by centres with, or with ready access to, specific vasculitis expertise

*Data from the ADVOCATE trial, which included up to 52 weeks of avacopan exposure, suggested that avacopan may have efficacy for maintenance of remission; however, longer-term use cannot be not recommended due to lack of data beyond 1 year; decisions around GC dosage and duration should be based on the individual patient's disease course, risk for/presence of GC-related comorbidities, and preferences. Always refer to the product SmPC for approved indication before prescribing. AAV, ANCA-associated vasculitis; AE, adverse event; ANCA, anti-neutrophil cytoplasmic antibody; AZA, azathioprine; EULAR, European League Against Rheumatism; GC, glucocorticoid; GPA, granulomatosis with polyangiitis; MMF, mycophenolate mofetil; MPA, microscopic polyangiitis; MTX, methotrexate; RTX, rituximab.

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