2022 EULAR Recommendations for ANCA-Associated Vasculitis

The European Alliance of Associations for Rheumatology (EULAR) has updated its recommendations for the management of ANCA-associated vasculitis (AAV). These evidence-based guidelines cover diagnosis, treatment, and monitoring of granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA). Key changes include new recommendations on ANCA testing, use of rituximab, glucocorticoid dosing, and management of EGPA. The guidelines aim to improve outcomes for patients with these rare but serious autoimmune diseases.

Overarching Principles

1 Shared Decision-Making

Patients with AAV should be offered best care based on shared decision-making between the patient and the physician, considering efficacy, safety, and costs.

2 Patient Education

Patients should have access to education focusing on the impact of AAV, its prognosis, key warning symptoms, and treatment, including treatment-related complications.

3 Periodic Screening

Patients with AAV should be periodically screened for treatment-related adverse effects and comorbidities. Prophylaxis and lifestyle advice are recommended to reduce these complications.

4 Multidisciplinary Management

AAV requires multidisciplinary management by centers with, or with ready access to, expertise in vasculitis.

Diagnosis of AAV

1. ## Clinical Assessment

   Structured clinical assessment is recommended to evaluate signs and symptoms of AAV.

2. ## ANCA Testing

   Testing for both PR3-ANCA and MPO-ANCA using high-quality antigen-specific assays is recommended for suspected AAV.

3. ## Biopsy

   Biopsy of affected organs is strongly recommended to confirm diagnosis and distinguish active disease from damage.

4. ## Imaging

   Imaging studies like CT, MRI and PET scans are recommended to detect organ involvement and identify potential biopsy sites.

Induction Therapy for GPA/MPA

Rituximab

Rituximab is recommended for induction of remission, especially for relapsing disease. It is preferred over cyclophosphamide for patients wishing to preserve fertility.

Cyclophosphamide

Cyclophosphamide remains an option for induction therapy, particularly for new-onset disease or severe renal involvement.

Glucocorticoids

Oral glucocorticoids are recommended at a starting dose of 50-75 mg prednisolone equivalent/day, with rapid tapering to 5 mg/day by 4-5 months.

Avacopan

Avacopan may be considered in combination with rituximab or cyclophosphamide to reduce glucocorticoid exposure.

Maintenance Therapy for GPA/MPA

1

Rituximab

Rituximab is recommended as first-line maintenance therapy after induction with either rituximab or cyclophosphamide.

2

Alternative Agents

Azathioprine or methotrexate may be considered as alternatives to rituximab for maintenance therapy.

3

Duration

Maintenance therapy is recommended for 24-48 months following induction of remission for new-onset disease.

4

Extended Treatment

Longer duration of therapy should be considered for relapsing patients or those at increased risk of relapse.

Management of EGPA

Severe EGPA

For organ-threatening or life-threatening EGPA, high-dose glucocorticoids plus cyclophosphamide or rituximab are recommended for induction of remission.

Non-severe EGPA

For non-organ-threatening EGPA, glucocorticoids alone are recommended for induction. Mepolizumab is recommended for relapsing or refractory non-severe disease.

Maintenance

Mepolizumab is recommended for maintenance of remission after non-severe relapse. For maintenance after severe disease, methotrexate, azathioprine, mepolizumab or rituximab should be considered.

Monitoring and Prophylaxis

1

Clinical Assessment

Structured clinical assessment, rather than ANCA or CD19+ B-cell testing alone, should guide treatment decisions.

2

Immunoglobulin Monitoring

Serum immunoglobulin concentrations should be measured before each rituximab course to detect secondary immunodeficiency.

3

Infection Prophylaxis

Trimethoprim-sulfamethoxazole is recommended as prophylaxis against Pneumocystis jirovecii pneumonia and other infections for patients receiving rituximab, cyclophosphamide, or high-dose glucocorticoids.

Special Considerations

Fertility Preservation

Rituximab is preferred over cyclophosphamide for patients wishing to preserve fertility.

Renal Disease

Plasma exchange may be considered for patients with serum creatinine >300 µmol/L due to active glomerulonephritis.

Pulmonary Hemorrhage

Routine use of plasma exchange is not recommended for alveolar hemorrhage in GPA and MPA.

Refractory Disease

Patients with refractory disease should be managed in conjunction with or referred to a vasculitis center of expertise.

Future Directions

1

Biomarker Research

Further studies are needed to identify predictive biomarkers of relapse in AAV.

2

Treatment Optimization

Research is ongoing to optimize glucocorticoid-sparing regimens and determine ideal duration of maintenance therapy.

3

Novel Therapies

Clinical trials are evaluating new targeted therapies and treatment approaches for AAV.

4

Personalized Medicine

Efforts are underway to develop more personalized treatment strategies based on individual patient characteristics and risk factors.