

ANCA-Associated Vasculitis

Practical bedside approach • GPA / MPA with kidney involvement

01 RECOGNISE THE DISEASE

- AAV causes **necrotising small-vessel inflammation**: granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA).
- Kidney involvement typically causes **pauci-immune necrotising crescentic glomerulonephritis**: haematuria, proteinuria and rising creatinine; progression may be rapid.
- Look for sinus/ear disease, pulmonary nodules or haemorrhage, purpura, neuropathy and constitutional symptoms. Kidney-limited disease also occurs.

02 DIAGNOSTIC WORK-UP

- Check creatinine/eGFR, potassium, full blood count, CRP, urinalysis, sediment and quantified proteinuria. Dysmorphic red cells and red cell casts support glomerulonephritis.
- Test **PR3-ANCA** and **MPO-ANCA** with antigen-specific assays. PR3 often accompanies GPA and MPO often accompanies MPA; neither association is absolute.
- **Positive ANCA alone does not establish AAV; negative ANCA does not exclude it.** Exclude infection, drug-induced disease and other mimics.
- Arrange kidney biopsy when feasible. Test **anti-GBM antibodies** in suspected rapidly progressive GN/pulmonary-renal syndrome; assess lungs and other involved organs.
- **Do not delay treatment for biopsy** in a rapidly deteriorating patient with a compatible presentation and positive MPO-/PR3-ANCA.

03 INDUCE REMISSION

- Organ-threatening GPA/MPA: **rituximab or cyclophosphamide plus glucocorticoids**, usually using a reduced-dose taper when appropriate.
- Rituximab is often preferred for relapsing disease, PR3-AAV or fertility concerns. Severe kidney failure may favour cyclophosphamide or a combined specialist regimen.
- **Avacopan**, a C5a receptor antagonist, may replace much of the oral glucocorticoid regimen in selected patients; it accompanies rituximab/cyclophosphamide, not monotherapy.
- **Plasma exchange is not routine.** Consider with creatinine >3.4 mg/dL, dialysis requirement/rapid creatinine rise, or hypoxaemic alveolar haemorrhage; weigh infection risk.
- **Add plasma exchange for AAV with anti-GBM overlap.** Provide kidney replacement therapy for standard urgent indications.

04 MAINTAIN REMISSION AND PREVENT TOXICITY

- After remission, use **rituximab or azathioprine with low-dose glucocorticoids**. Individualise maintenance, generally **18 months to 4 years**, according to relapse and toxicity risks.
- Provide Pneumocystis prophylaxis during cyclophosphamide and ≥6 months after rituximab induction; extend if needed.
- Before rituximab, check hepatitis B and immunoglobulins. Review vaccines, infection risk and fertility.
- Monitor blood counts, kidney function and toxicity; liver tests with avacopan, immunoglobulins with rituximab.

05 MONITOR RESPONSE AND RECOGNISE RED FLAGS

- Follow symptoms, organ involvement, creatinine, urine sediment and proteinuria. Distinguish active vasculitis from infection and chronic damage.
- **ANCA titres alone should not determine treatment changes.** Persistent haematuria/proteinuria does not always mean active disease; consider repeat biopsy when uncertain.
- **Rapid kidney deterioration, oliguria, hypoxaemia or suspected alveolar haemorrhage** requires urgent hospital assessment; haemoptysis may be absent.

KEY MESSAGE

Recognise AAV early: kidney and pulmonary involvement can be life-threatening. Treat active disease promptly, then prevent relapse while limiting infection and treatment toxicity.

REFERENCE

KDIGO 2024 Clinical Practice Guideline for the Management of ANCA-Associated Vasculitis. Kidney Int. 2024;105(Suppl):S71-S116; corrected treatment algorithm, 2024. kdigo.org.