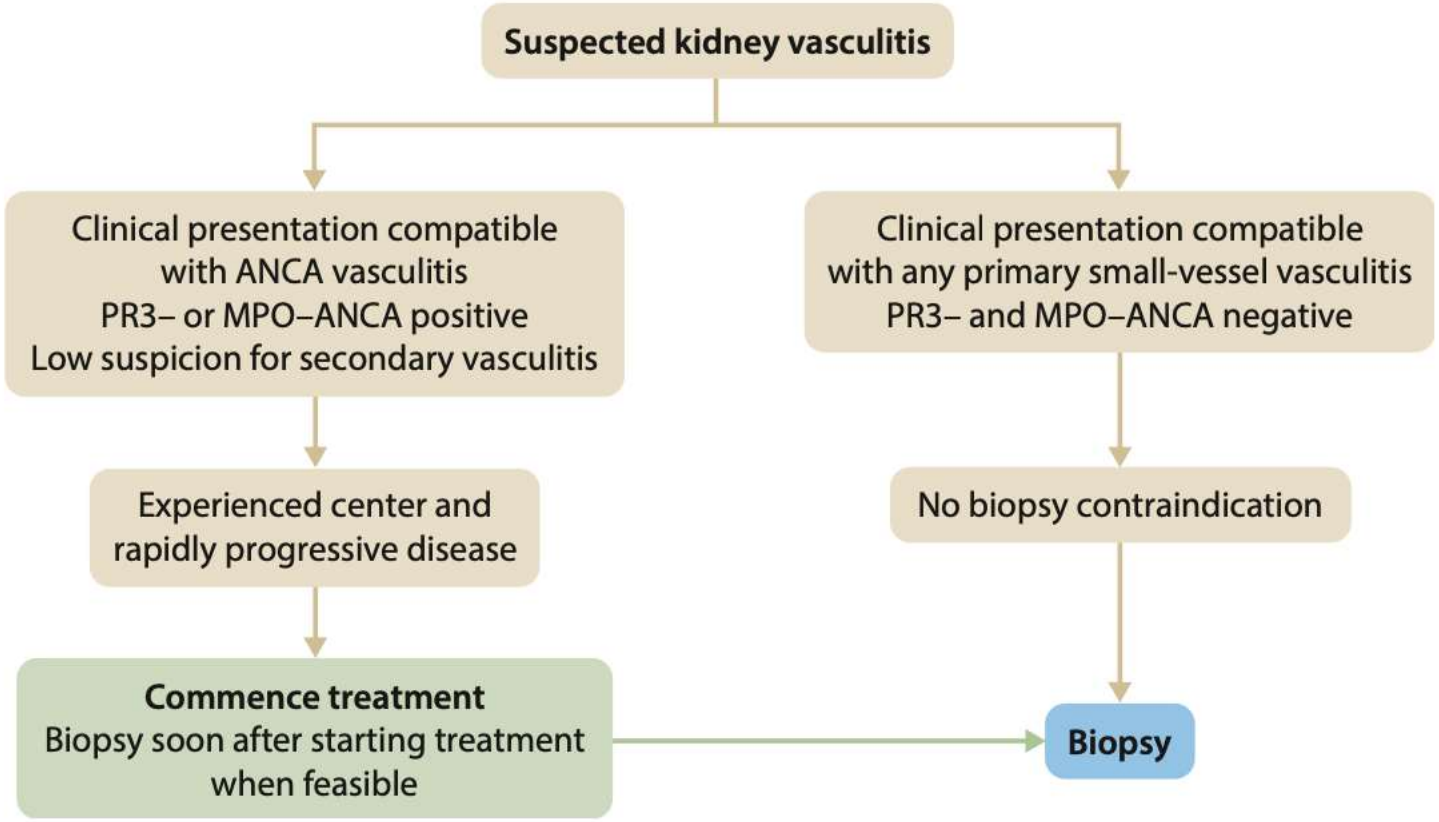
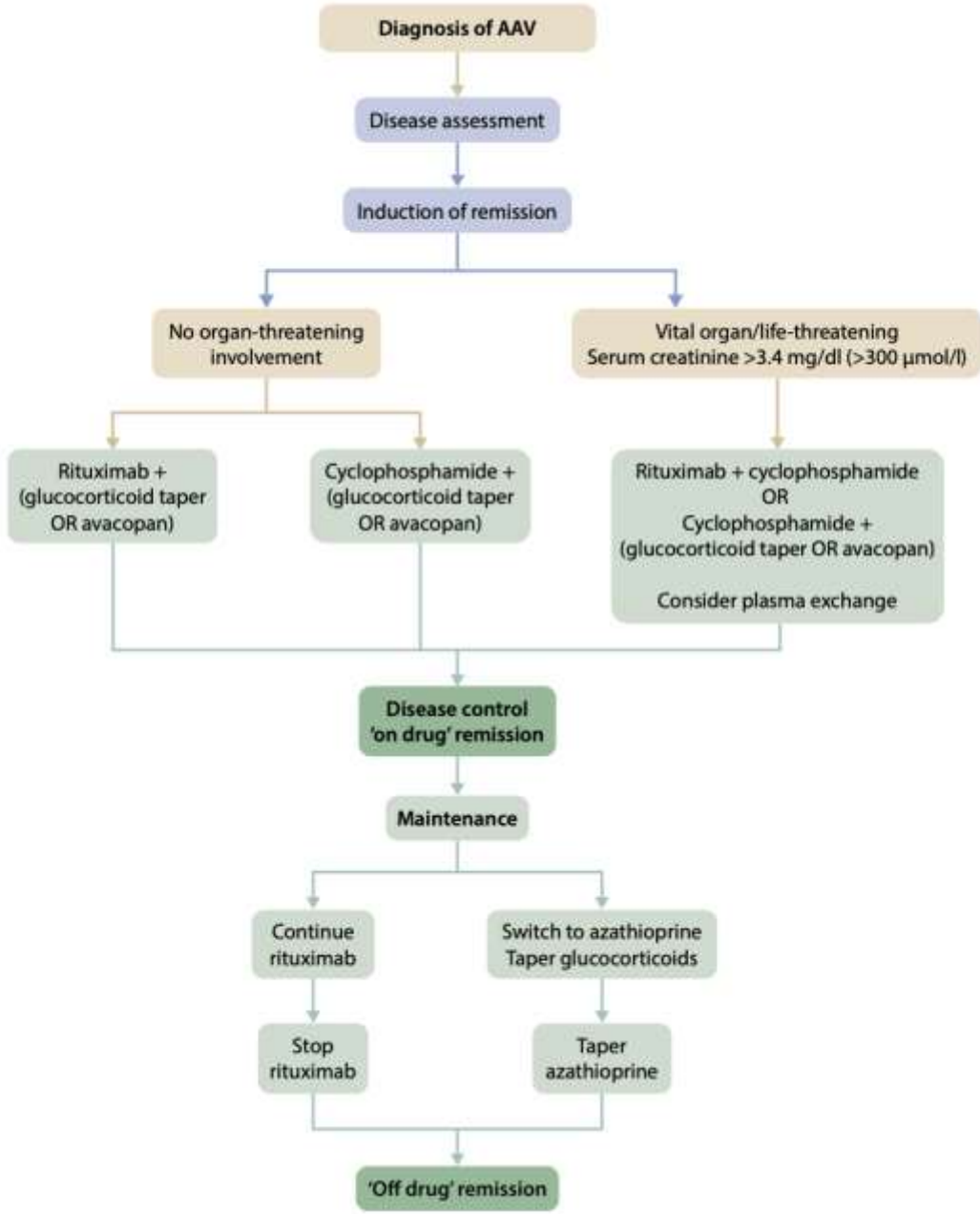
KDIGO ANCA VASCULITIS

2024 UPDATE

Diagnostic Approach



Practice Point 9.3.1.1: A practical treatment algorithm for AAV with kidney involvement is given in Figure 6.



Recommended dosing

Oral cyclophosphamide	Intravenous cyclophosphamide	Rituximab	Rituximab and i.v. cyclophosphamide	MMF	Avacopan
2 mg/kg/d for 3 months, continue for ongoing activity to a maximum of 6 months	15 mg/kg at weeks 0, 2, 4, 7, 10, 13 (16, 19, 21, 24 if required)	375 mg/m^2/week × 4 weeks OR 1 g at weeks 0 and 2	Rituximab 375 mg/m^2/week × 4 weeks, with i.v. cyclophosphamide 15 mg/kg at weeks 0 and 2 OR Rituximab 1 g at 0 and 2 weeks with i.v. cyclophosphamide 500 mg/2 weeks × 6	2000 mg/d (divided doses), may be increased to 3000 mg/d for poor treatment response	30 mg twice daily as alternative to glucocorticoids, in combination with rituximab or cyclophosphamide induction
Reduction for age: • 60 yr, 1.5 mg/kg/d • 70 yr, 1.0 mg/kg/d Reduce by 0.5 mg/kg/day for GFR <30 ml/min/1.73 m^2	Reduction for age: • 60 yr 12.5 mg/kg • 70 yr, 10 mg/kg Reduce by 2.5 mg/kg for GFR <30 ml/min/1.73 m^2				

Figure 10 | Immunosuppressive drug dosing for AAV. AAV, ANCA-associated vasculitis; ANCA, antineutrophil cytoplasmic antibody; GFR, glomerular filtration rate; i.v., intravenous; MMF, mycophenolate mofetil.

When to prefer what

Rituximab preferred	Cyclophosphamide preferred
• Children and adolescents • Premenopausal women and men concerned about their fertility • Frail older adults • Glucocorticoid-sparing especially important • Relapsing disease • PR3–ANCA disease	• Rituximab difficult to access • Severe GN (SCr >4 mg/dl [354 µmol/l]), combination of 2 intravenous pulses of cyclophosphamide with rituximab can be considered

Intravenous cyclophosphamide	Oral cyclophosphamide
• Patients who already have a moderate cumulative dose of cyclophosphamide • Patients with lower white blood cell counts • Ready access to an infusion center • Adherence may be an issue	• Cost is an important factor • Access to an infusion center difficult • Adherence is not an issue

Plasma Exchange Frequency

ANCA vasculitis with severe kidney disease	Vasculitis with diffuse pulmonary hemorrhage	Vasculitis in association with anti-GBM antibodies
Seven treatments over a maximum of 14 days, 60 ml/kg volume replacement, albumin substitution	Daily until bleeding stops, replace albumin with fresh, frozen plasma	Daily for 14 days or until anti-GBM antibodies are undetectable

Figure 11 | Plasma exchange dosing and frequency for AAV. If a patient is at risk of bleeding, volume replacement should be with fresh, frozen plasma. AAV, ANCA-associated vasculitis; ANCA, antineutrophil cytoplasmic antibody; GBM, glomerular basement membrane.

Key points

- The KDIGO : update to the 2021 version

- Key changes include the

  - Use of lower-dose corticosteroids

  - Incorporation of Avacopan as an alternative to traditional glucocorticoids,

  - Updated recommendations on plasma exchange for patients with advanced kidney disease