Top 10 Takeaways for Clinicians from the KDIGO 2021 Clinical Practice Guideline for the Management of ANCA-Associated Vasculitis (AAV)

1 Diagnosis of ANCA-associated vasculitis

Diagnosis of AAV must be made as early as possible to decrease the risk of permanent loss of kidney function and life-threatening complications. In case of clinical presentation compatible with small-vessel vasculitis in combination with positive myeloperoxidase (MPO)- or proteinase 3 (PR3)-ANCA serology, waiting for kidney biopsy should not delay starting immunosuppressive treatment, especially in patients who are rapidly deteriorating (Figure 1).

2 Initial treatment

Initial treatment of AAV is glucocorticoids in combination with cyclophosphamide or rituximab (Figure 2). In patients with markedly reduced or rapidly declining kidney function, both cyclophosphamide and glucocorticoids, and the combination of rituximab and cyclophosphamide can be considered in this setting.

3 Avacopan

Avacopan may be used as an alternative to glucocorticoids. Patients with an increased risk of glucocorticoid toxicity are likely to receive the most benefit from avacopan. Patients with lower GFR may benefit from greater GFR recovery.

4 Plasma exchange

Plasma exchange should be considered for patients with SCr >3.4 mg/dl (300 µmol/l) requiring dialysis or with rapidly increasing SCr, and in patients with diffuse alveolar hemorrhage who have hypoxemia (Figure 3). Plasma exchange should be added to initial treatment for patients with an overlap syndrome of ANCA vasculitis and anti-GBM.

5 Glucocorticoid use

Although high-dose glucocorticoids have traditionally been given during the initial treatment of AAV, recent data demonstrated that lower doses are equally effective but with fewer short- and long-term toxicities.

6 Maintenance treatment

Maintenance therapy with either rituximab, or azathioprine and low dose glucocorticoids is recommended after induction of remission (Figure 3). Optimal duration of the maintenance treatment is not known, but should be between 18 months and 4 years.

7 Preferred maintenance treatment

Rituximab as maintenance treatment is preferred in patients with relapsing disease, PR3-ANCA disease, frail older adults, azathioprine allergy, or when glucocorticoid-sparing is especially important.

8 Withdrawal of maintenance therapy

When considering withdrawal of maintenance therapy, the risk of relapse should be factored in, and patients should be informed of the need for prompt attention if symptoms recur.

9 Relapsing disease

Patients with relapsing disease (life- or organ-threatening) should be re-induced, preferably with rituximab.

10 Refractory disease

Patients with refractory disease can be treated by an increase in glucocorticoids (intravenous or oral), by the addition of rituximab if cyclophosphamide induction had been used previously, or vice versa. Plasma exchange can be considered.

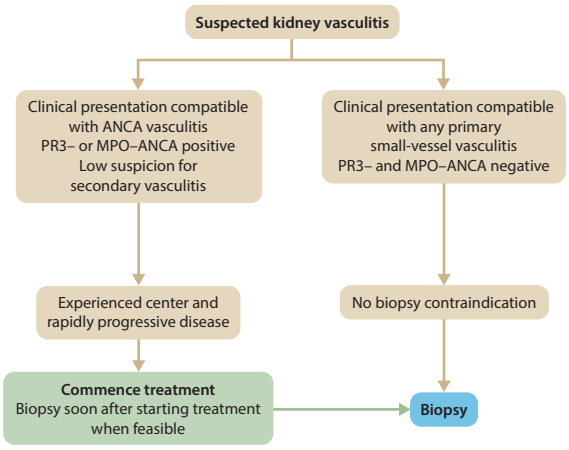

Figure 1

Rituximab preferred	Cyclophosphamide preferred
• Children and adolescents • Pre-menopausal 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 two intravenous pulses of cyclophosphamide with rituximab can be considered

Figure 2

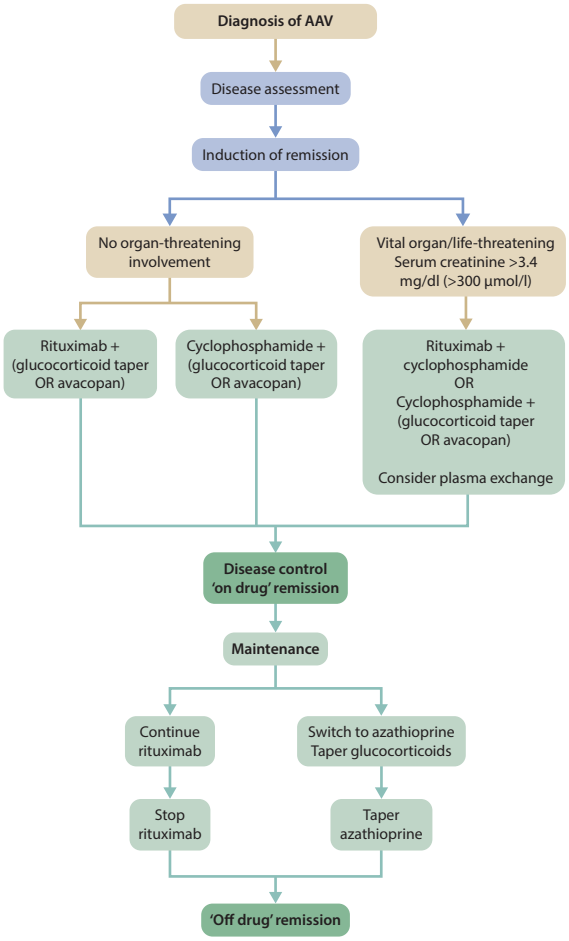

Figure 3