

### 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. In patients with markedly reduced or rapidly declining kidney function, cyclophosphamide is preferred because of limited experience with rituximab (Figure 2).

### 3

#### Rituximab as initial treatment

Rituximab is the preferred initial treatment in children and adolescents, pre-menopausal women and men concerned about their fertility, frail older adults, patients with relapsing disease, patients with PR3-ANCA disease and in patients in whom glucocorticoid-sparing is especially important.

### 4

#### Plasma exchange

Plasma exchange should be considered for patients with SCr >5.7 mg/dl (500 µmol/l) requiring dialysis or with rapidly increasing SCr, and in patients with diffuse alveolar hemorrhage who have hypoxemia (Figure 2). Plasma exchange should be added to initial treatment for patients with an overlap syndrome of ANCA vasculitis and anti-GBM.

### 5

#### Tapering of glucocorticoids

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 2). 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.

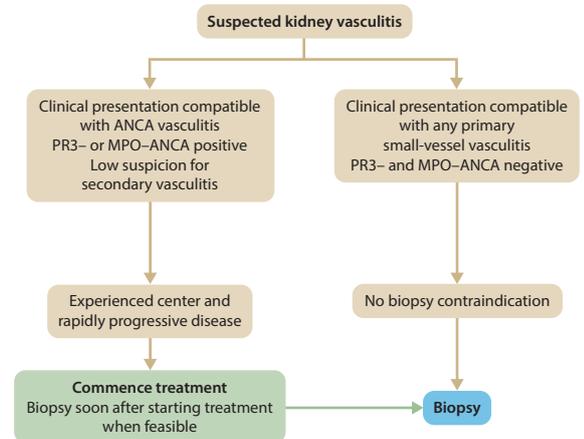


Figure 1

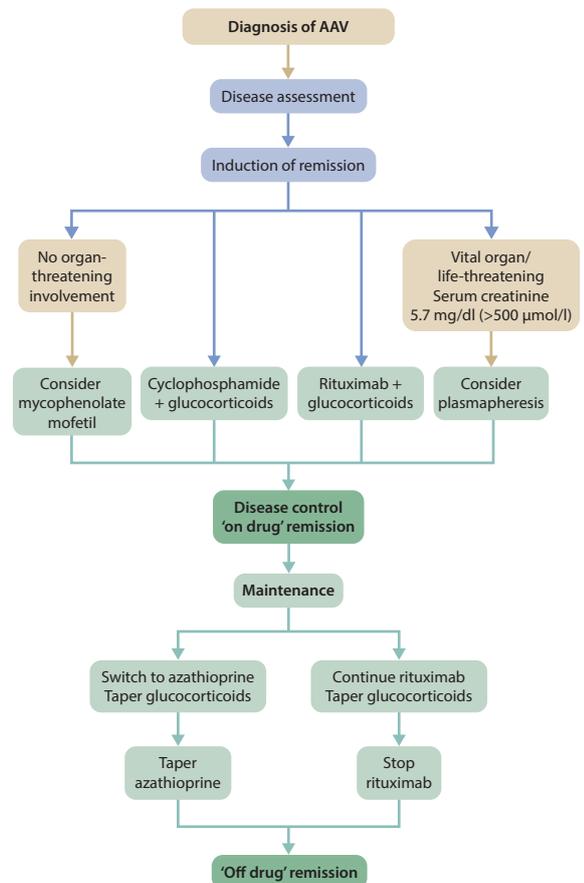


Figure 2