

## Guideline summary Renal vasculitis

### Recommendations\*

- If anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis (AAV) is suspected, the gold standard of tissue diagnosis should be used along with serum anti-neutrophil cytoplasmic antibody (ANCA) measurement for initial diagnosis.
- Measurement of ANCA by both ELISA and indirect immunofluorescence (IIF) in combination ensures optimal test sensitivity and specificity.
- The use of serial ANCA monitoring alone is insufficient to predict relapse or monitor disease activity.

Therapy for ANCA associated systemic vasculitis (AASV)	
Induction Therapy	Maintenance Therapy
Cyclophosphamide (IV or oral) and prednisolone	Azathioprine and lower doses of prednisolone to be used once remission has been established, to prevent a relapse
OR: Methotrexate and prednisolone for patients with serum creatinine <150 µmol/L	Prolonged cyclophosphamide is not recommended due to side effects. Etanercept is not recommended at all.
Plasma exchange is preferred over pulse methylprednisone as an adjunct to severe ANCA assoc. AAV causing necrotising glomerulonephritis and acute kidney failure (creatinine >500 µmol/L)	Methotrexate may be used in patients with well preserved kidney function. But it is not as effective.
	Prolonged oral co-trimoxazole can be effective in decreasing upper airways disease, but not relapse in other organs

### Suggestions for clinical care<sup>†</sup>

#### Diagnosis:

- Test performance is poor as a screening test in patients with few clinical features of AAV.
- Rapid ELISA test may be helpful as an adjunct to urgent therapeutic decisions, but should not supplant the need for histological confirmation of disease and ANCA IIF testing.
- Serial ANCA testing (ELISA and IIF) to monitor disease activity may be useful in some situations such as:
  - disappearance of ANCA is associated with disease remission and a lower risk of relapse,
  - reappearance or rising ANCA titre is of greater relevance in the setting of worsening clinical features, and
  - the persistence of anti-proteinase 3 (ant-PR3) antibodies is associated with a higher risk of relapse.

#### Maintenance Therapy:

- Optimal treatment length unknown. Recommended: continue for 12 after remission, or up to 24-36 months if at risk.
- Those at risk of relapse are: Patients with Wegner's granulomatosis (WG), ongoing ANCA positivity, particularly PR3-ANCA.
- Optimal steroid regimen unknown. Recommended: Steroids used with another agent with gradual dose reduction.
- Mycophenolate may be useful, but evidence is lacking so restrict use to those intolerant of other agents.

#### Disease relapse:

- Do not base relapse diagnosis on change in ANCA titre alone.
- Optimal treatment of relapse unknown. Recommend: Induction-like therapies
- Rituximab may be useful

#### Renal transplantation:

- ANCA assoc AAV is not a contraindication to renal transplantation
- Optimal timing for transplantation is unknown. Recommended: after clinical remission has been achieved.
- Decision to transplant and timing of transplant should not be on ANCA alone
- Standard transplantation of immunosuppression regimes for patients with AASV is appropriate
- Cyclophosphamide and high dose corticosteroids can be used to treat relapses for patients with AASV and transplant.

**Data Sources:** Medline; Cochrane CENTRAL Register of Controlled Trials (Cochrane Library)

**Study Selection and Assessment:** Guidelines were developed using high level (I – II) evidence (i.e. systematic reviews of RCTs or standard RCT studies) when available, otherwise observational studies (level III – IV evidence) such as cohort, case-control and case series studies were used.

**Full guideline recommendations:** [http://www.cari.org.au/ckd\\_renal\\_vasculitis\\_published.php](http://www.cari.org.au/ckd_renal_vasculitis_published.php)

Date of summary: February 2010

Date guideline published: August 2008

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\*Level I-II evidence, †-Level III-IV evidence

# Implementation of this guideline into practice

- Ensure there is an up to date policy and procedure document for your unit
- Evaluate regularly
- Below is a suggested clinical pathway, which can be adapted to suit the needs of your unit

