

**Harvard Medical School Department of
Continuing Education and the Renal Division
of Brigham and Women's Hospital**



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Antineutrophil Cytoplasmic Antibodies (ANCA) in the Long-Term Management of Wegener Disease: How Should We Use Them?

By Andreas Herrlich, MD, PhD

Objectives: After studying this review the reader should be able to:

- Understand the sensitivity and specificity of ANCA testing
- Be familiar with current approaches to induction and remission therapy in Wegener granulomatosis (WG)
- Recognize the value of plasmapheresis in induction therapy
- Understand the relationship between ANCA levels, disease activity, and relapse.

Questions: True or False

1. First-line treatment for remission induction includes oral prednisone (1 mg/kg) and cyclophosphamide (CP; 500 mg – 1 g/m²/day in the first week).
True False
2. Major items in the Birmingham Vasculitis Activity Score (BVAS) are generally those that would trigger the use of CP and include, for example, gangrene, scleritis, pulmonary hemorrhage, red blood cell casts, glomerular filtration rate reductions of >25% or a rise in creatinine of >30%, mononeuritis multiplex, and stroke.
True False
3. Mycophenolate mofetil may provide a less toxic alternative to CP in patients with limited disease and those who cannot be treated with CP.
True False
4. The reported sensitivity to ANCA for patients with WG and other ANCA-associated vasculitis syndromes varies widely, ranging from 50%–95%. Immunofluorescence (IF) has less sensitivity and specificity than enzyme-linked immunosorbent assay, but the combination improves diagnostic sensitivity and specificity.
True False
5. Plasmapheresis is used in the treatment of every WG relapse with renal involvement.
True False
6. Nearly all Wegener patients with severe disease are ANCA-positive, but as many as 20% of patients with limited disease are ANCA-negative.
True False
7. WG has a tendency to relapse in at least 50% of cases within 5 years, and continued immunosuppression during remission is usually needed to prevent relapse.
True False

8. ANCA levels fall or become negative in only 20% of patients within the first 1-3 months of induction treatment, and only a few patients have a renewed (or persistently) positive ANCA at the point of relapse.

True False

9. Falling ANCA titers are associated with a low risk of relapse. Increases in ANCA may be followed by a relapse in disease within 3-6 months.

True False

10. Current best practice for WG maintenance treatment includes incremental adjustments of immunosuppression based on a careful correlation with signs of active clinical disease and ANCA titer changes (increases or declines).

True False

11. This issue of *Nephrology Rounds* adequately addressed the topic, and the data and discussion were fair and balanced.

AGREE DISAGREE

12. Potential conflicts of interest disclosed by the author on the back page were properly expressed.

AGREE DISAGREE

13. The information presented in this issue of *Nephrology Rounds* will increase my clinical knowledge and improve the care of my patients.

AGREE DISAGREE

14. *Nephrology Rounds* from Brigham and Women's Hospital and Harvard Medical School is an effective CME program.

AGREE DISAGREE

Comments/Topic Suggestions: _____

To receive AMA category 1 credit, you must correctly answer 60% of questions 1-10, and answer 11-14.

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