

Overview

Useful For

Evaluating patients suspected of having autoimmune vasculitis, both Wegener granulomatosis and microscopic polyangiitis

Profile Information

Test Id	Reporting Name	Available Separately	Always Performed
MPO	Myeloperoxidase Ab, S	Yes	Yes
PR3	Proteinase 3 Ab (PR3), S	Yes	Yes

Reflex Tests

Test Id	Reporting Name	Available Separately	Always Performed
ANCA	Cytoplasmic Neutrophilic Ab, S	Yes	No

Testing Algorithm

If myeloperoxidase antibody or proteinase 3 antibody is > or =0.4 U, then cytoplasmic neutrophilic antibodies will be performed at an additional charge.

Method Name

Multiplex Flow Immunoassay

NY State Available

Yes

Specimen

Specimen Type

Serum

Ordering Guidance

For monitoring disease activity, we advise physicians to order ANCA / Cytoplasmic Neutrophil Antibodies, Serum or MPO / Myeloperoxidase Antibodies, IgG, Serum.

Specimen Required

Collection Container/Tube:

Preferred: Serum gel

**Acceptable:** Red top  
**Submission Container/Tube:** Plastic vial  
**Specimen Volume:** 1 mL  
**Collection Instructions:** Centrifuge and aliquot serum into a plastic vial.

Forms

If not ordering electronically, complete, print, and send 1 of the following forms with the specimen:  
[-General Request](#) (T239)  
[-Renal Diagnostics Test Request](#) (T830)

**Specimen Minimum Volume**  
0.5 mL

Reject Due To

Gross hemolysis	Reject
Gross lipemia	Reject
Gross icterus	OK

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Serum	Refrigerated (preferred)	21 days	
	Frozen	21 days	

Clinical & Interpretive

Clinical Information

Antineutrophil cytoplasmic antibodies (ANCA) occur in patients with autoimmune vasculitis including Wegener granulomatosis (WG), microscopic polyangiitis (MPA), or organ-limited variants thereof such as pauci-immune necrotizing glomerulonephritis.(1) ANCA react with enzymes in the cytoplasmic granules of human neutrophils including proteinase 3 (PR3), myeloperoxidase (MPO), elastase, and cathepsin G.

Autoantibodies to PR3 occur in patients with WG (both classical WG and WG with limited end-organ involvement) and produce a characteristic pattern of granular cytoplasmic fluorescence on ethanol-fixed neutrophils called the cANCA pattern. Antibodies to MPO occur predominately in patients with MPA and produce a pattern of perinuclear cytoplasmic fluorescence on ethanol-fixed neutrophils called the pANCA pattern.

Autoantibodies to PR3 and MPO can also be detected by EIA methods and are referred to as PR3 ANCA and MPO ANCA, respectively.

Reference Values

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**MYELOPEROXIDASE ANTIBODIES, IgG**

&lt;0.4 U (negative)

0.4-0.9 U (equivocal)

&gt; or =1.0 U (positive)

Reference values apply to all ages.

**PROTEINASE 3 ANTIBODIES, IgG**

&lt;0.4 U (negative)

0.4-0.9 U (equivocal)

&gt; or =1.0 U (positive)

Reference values apply to all ages.

**Interpretation**

Positive results for proteinase 3 (PR3) antineutrophil cytoplasmic antibodies (ANCA) and cANCA or pANCA are consistent with the diagnosis of Wegener granulomatosis (WG), either systemic WG with respiratory and renal involvement or limited WG with more restricted end-organ involvement.

Positive results for MPO ANCA and pANCA are consistent with the diagnosis of autoimmune vasculitis including microscopic polyangiitis (MPA) or pauci-immune necrotizing glomerulonephritis.

A positive result for PR3 ANCA or MPO ANCA has been shown to detect 89% of patients with active WG or MPA (with or without renal involvement) with fewer than 1% false-positive results in patients with other diseases.(1)

**Cautions**

Sequential measurements of titers of antineutrophil cytoplasmic antibodies (cANCA) are useful to monitor the response to treatment in patients with Wegener granulomatosis (WG). While titers often decrease following successful treatment, the results cannot be relied upon in all cases to determine the response to therapy. In individual patients, the titers of cANCA may not correlate well with response to treatment. The results of proteinase 3 (PR3) ANCA (an autoantibody with a cANCA pattern) by EIA have not been shown to be useful for monitoring disease activity. Accordingly, for monitoring disease activity we advise physicians to order ANCA / Cytoplasmic Neutrophil Antibodies, Serum or MPO / Myeloperoxidase Antibodies, IgG, Serum (the ANCA vasculitis test panel is not appropriate for monitoring disease activity).

The ANCA vasculitis test panel cannot be relied upon exclusively to establish the diagnosis of autoimmune vasculitis (WG or microscopic polyangiitis: MPA). Some patients with WG or MPA may not have a measurable titer of antibodies detected by this panel of tests. Some of these patients have antibodies to other neutrophil antigens not included in the panel (eg, neutrophil elastase).

**Clinical Reference**

1. Russell KA, Wiegert E, Schroeder DR, et al: Detection of anti-neutrophil cytoplasmic antibodies under actual clinical testing conditions. Clin Immunol 2002 May;103(2):196-203
2. Specks U, Homburger HA, DeRemee RA: Implications of cANCA testing for the classifications of Wegner's Granulomatosis: performance of different detection systems. Adv Exp Med Biol 1993;336:65-70

Performance

Method Description

See Individual Unit Codes

PDF Report

No

Day(s) Performed

Monday through Saturday

Report Available

3 to 5 days

Specimen Retention Time

See Individual Unit Codes

Performing Laboratory Location

Rochester

Fees & Codes

Fees

- Authorized users can sign in to [Test Prices](#) for detailed fee information.
- Clients without access to Test Prices can contact [Customer Service](#) 24 hours a day, seven days a week.
- Prospective clients should contact their account representative. For assistance, contact [Customer Service](#).

Test Classification

This test has been cleared, approved, or is exempt by the US Food and Drug Administration and is used per manufacturer's instructions. Performance characteristics were verified by Mayo Clinic in a manner consistent with CLIA requirements.

CPT Code Information

83516 x 2

86036 (if appropriate)

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
VASC	ANCA Panel for Vasculitis, S	90230-4

Result ID	Test Result Name	Result LOINC® Value
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Test Definition: VASC

Antineutrophil Cytoplasmic Antibodies  
Vasculitis Panel, Serum

MPO	Myeloperoxidase Ab, S	48404-8
PR3	Proteinase 3 Ab (PR3), S	74106-6