Overview

Useful For
Evaluating patients suspected of having Wegener granulomatosis (WG)

Distinguishing between WG and other forms of vasculitis, in conjunction with:

- MPO / Myeloperoxidase Antibodies, IgG, Serum
- ANCA / Cytoplasmic Neutrophil Antibodies, Serum (may be obtained as VASC / Antineutrophil Cytoplasmic Antibodies Vasculitis Panel, Serum)

May be useful to follow treatment response or to monitor disease activity in patients with myeloperoxidase antibodies

Method Name
MultiplexFlowImmunoassay

NY State Available
Yes

Specimen

Specimen Type
Serum

Specimen Required

Container/Tube:

Preferred: Serum gel
Acceptable: Red top

Specimen Volume: 0.5 mL

Forms
If not ordering electronically, complete, print, and send a Renal Diagnostics Test Request (T830) with the specimen.

Specimen Minimum Volume
0.35 mL

Reject Due To

<table>
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<tr>
<th>Condition</th>
<th>Decision</th>
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<tbody>
<tr>
<td>Gross hemolysis</td>
<td>Reject</td>
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<tr>
<td>Gross lipemia</td>
<td>Reject</td>
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<tr>
<td>Gross icterus</td>
<td>OK</td>
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Specimen Stability Information
**Clinical and Interpretive**

**Clinical Information**
Proteinase 3 (PR3) antigen is a 29-kD serine protease that exists as a protein triplet in human neutrophils.

Wegener granulomatosis (WG) is an autoimmune vasculitis that affects the kidneys and lungs, as well as other organs. Patients with WG develop autoantibodies to the PR3 antigen of myeloid lysosomes (PR3 antineutrophil cytoplasmic antibodies [PR3 ANCA]).(1)

Since it is often impossible to distinguish between WG and other forms of vasculitis on the basis of clinical signs and symptoms, tests for PR3 ANCA should be employed with other serologic tests in the initial diagnostic evaluation of patients with clinical features of vasculitis (eg, VASC / Antineutrophil Cytoplasmic Antibodies Vasculitis Panel, Serum).

**Reference Values**

- <0.4 U (negative)
- 0.4-0.9 U (equivocal)
- > or =1.0 U (positive)

Reference values apply to all ages.

**Interpretation**
Proteinase 3 antineutrophil cytoplasmic antibodies (PR3 ANCA) are detectable in nearly all patients with severe active Wegener granulomatosis (WG).(2) The presence of PR3 ANCA is a specific diagnostic indicator of WG; less than 2% of positive results occur in patients who do not have the disease.(3,4)

A negative result for PR3 ANCA diminishes the likelihood that a patient has active WG; but, approximately 20% of patients with limited WG may test negative for PR3 ANCA.(3)

The levels of PR3 ANCA often decline following successful treatment of patients with WG. Nevertheless, follow-up testing for PR3 ANCA to evaluate clinical status in treated patients should be used with caution as the levels of antibodies may correlate poorly with clinical status in some patients.

**Cautions**
While the presence of proteinase 3 antineutrophil cytoplasmic antibodies (PR3 ANCA) is highly specific for Wegener granulomatosis (WG), it is recommended that positive test results obtained by immunoassay be confirmed by another testing method.(4) This is best accomplished by testing for cytoplasmic ANCA (cANCA) and perinuclear ANCA (pANCA) by indirect immunofluorescence microscopy (ANCA / Cytoplasmic Neutrophil Antibodies, Serum). Alternatively, VASC / Antineutrophil Cytoplasmic Antibodies Vasculitis Panel, Serum includes tests for PR3 ANCA, myeloperoxidase antibodies, and, if indicated, cANCA and pANCA. This panel is recommended for the initial diagnostic evaluation of patients clinically suspected of having systemic vasculitis. The simultaneous presence of PR3 ANCA and cANCA has a specificity greater than 99% for WG.(3)
Clinical Reference
1. van der Woude FJ, Rasmussen N, Lobatto S, et al: Autoantibodies against neutrophils and monocytes: tool for
2. Finkleman JD, Lee AS, Hummel AM, et al: ANCA are detectable in nearly all patients with active severe
clinical testing conditions. Clin Immunol 2002;103:196-203

Performance

Method Description
Proteinase 3 (PR3) antigen is covalently coupled to polystyrene microspheres that are impregnated with fluorescent
dyes to create a unique fluorescent signature. PR3 antibodies, if present in diluted serum, bind to the PR3 antigen on
the microspheres. The microspheres are washed to remove extraneous serum proteins. Phycoerythrin
(PE)-conjugated antihuman IgG antibody is then added to detect IgG anti-PR3 bound to the microspheres. The
microspheres are washed to remove unbound conjugate, and bound conjugate is detected by laser photometry. A
primary laser reveals the fluorescent signature of each microsphere to distinguish it from microspheres that are
labeled with other antigens. A secondary laser reveals the level of PE fluorescence associated with each
microsphere. Results are calculated by comparing the median fluorescence response for PR3 microspheres to a
4-point calibration curve.(Package insert: Bio-Plex 2200 Vasculitis. Bio-Rad Laboratories, Hercules, CA 4/2012)

PDF Report
No

Day(s) and Time(s) Test Performed
Monday through Saturday; 4 p.m.

Analytic Time
Same day/1 day

Maximum Laboratory Time
3 days

Specimen Retention Time
14 days

Performing Laboratory Location
Rochester

Fees and 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.
Test Definition: PR3
Proteinase 3 Ab (PR3), S

- Prospective clients should contact their Regional Manager. For assistance, contact Customer Service.

Test Classification
This test has been cleared, approved or is exempt by the U.S. 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

LOINC® Information

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<td>Proteinase 3 Ab (PR3), S</td>
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