MAYO CLINIC LABORATORIES

Test Definition: PR3

Proteinase 3 Antibodies, IgG, Serum

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

Multiplex Flow Immunoassay

NY State Available

Yes

Specimen

Specimen Type

Serum

Specimen Required

Collection Container/Tube:
Preferred: Serum gel
Acceptable: Red top
Submission Container/Tube: Plastic vial
Specimen Volume: 0.5 mL
Collection Instructions: Centrifuge and aliquot serum into a plastic vial.

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

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

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). Alternately, 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 diagnosis and marker of disease activity in Wegener granulomatosis. Lancet 1985;1:425-429
2. Finkleman JD, Lee AS, Hummel AM, et al: ANCA are detectable in nearly all patients with active severe Wegener's granulomatosis. Am J Med 2007;120:643
3. Russel KA, Wiegert E, Schroeder DR, et al: Detection of anti-neutrophil cytoplasmic antibodies under actual clinical testing conditions. Clin Immunol 2002;103:196-203
4. Savige J, Gillis D, Benson E, et al: International consensus statement on testing and reporting of antineutrophil cytoplasmic antibodies (ANCA). Am J Clin Pathol 1999;111:507-513

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) Performed

Monday through Saturday

Report Available

Same day/1 to 3 days

Specimen Retention Time

14 days

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

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
PR3	Proteinase 3 Ab (PR3), S	74106-6

Result ID	Test Result Name	Result LOINC® Value
PR3	Proteinase 3 Ab (PR3), S	74106-6