MAYO CLINIC LABORATORIES

Test Definition: PC2TS

Purkinje Cell Cytoplasmic Antibody Type 2 (PCA-2) Titer, Serum

Overview

Useful For

Evaluating patients who present with a subacute neurological disorder of undetermined etiology and have risk factors for lung cancer

Reporting an end titer result from serum specimens

Testing Algorithm

If the indirect immunofluorescence pattern suggests Purkinje cell cytoplasmic antibody type 2 (PCA-2), then this test will be performed at an additional charge.

Method Name

Only orderable as a reflex. For more information see:
-PAVAL / Paraneoplastic, Autoantibody Evaluation, Serum
-DMS2 / Dementia, Autoimmune/Paraneoplastic Evaluation, Serum
-ENS2 / Encephalopathy, Autoimmune/Paraneoplastic Evaluation, Serum
-EPS2 / Epilepsy, Autoimmune/Paraneoplastic Evaluation, Serum
-MDS2 / Movement Disorder, Autoimmune/Paraneoplastic Evaluation, Serum
-MAS1 / Myelopathy, Autoimmune/Paraneoplastic Evaluation, Serum
-AIAES / Axonal Neuropathy, Autoimmune/Paraneoplastic Evaluation, Serum
-DYS2 / Dysautonomia, Autoimmune/Paraneoplastic Evaluation, Serum
-GID2 / Gastrointestinal Dysmotility, Autoimmune/Paraneoplastic Evaluation, Serum

Indirect Immunofluorescence Assay (IFA)

NY State Available

Yes

Specimen

Specimen Type

Serum

Specimen Required

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-DMS2 / Dementia, Autoimmune/Paraneoplastic Evaluation, Serum
-ENS2 / Encephalopathy, Autoimmune/Paraneoplastic Evaluation, Serum
-EPS2 / Epilepsy, Autoimmune/Paraneoplastic Evaluation, Serum
-MDS2 / Movement Disorder, Autoimmune/Paraneoplastic Evaluation, Serum

-MAS1 / Myelopathy, Autoimmune/Paraneoplastic Evaluation, Serum
-AIAES / Axonal Neuropathy, Autoimmune/Paraneoplastic Evaluation, Serum
-DYS2 / Dysautonomia, Autoimmune/Paraneoplastic Evaluation, Serum
-GID2 / Gastrointestinal Dysmotility, Autoimmune/Paraneoplastic Evaluation, Serum

Specimen Minimum Volume
0.6 mL

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Serum	Refrigerated (preferred)	28 days	
	Ambient	72 hours	
	Frozen	28 days	

Clinical & Interpretive

Clinical Information
Purkinje cell autoantibodies (PCA) are among the antineuronal autoantibodies (ANNA) recognized clinically as markers of a patient's immune response to specific cancers (paraneoplastic autoantibodies).

In 1976, a PCA, defined by indirect immunofluorescence, was described by Dr. John Trotter and colleagues as a serological accompaniment of cerebellar ataxia related to Hodgkin lymphoma. That autoantibody is now known as anti-Tr or PCA-Tr.

PCA-1 (or anti-Yo), first described in 1983, serves as a serological marker for a new or recurrent carcinoma of the ovary, other Mullerian tissue, or breast. PCA-1-positive patients are women in 99% of cases. They usually present with subacute cerebellar degeneration, but 10% have sensory or motor neuropathy.

In 2000, the Mayo Clinic Neuroimmunology Laboratory described and named PCA-2, a new IgG marker of an immune response to small-cell lung carcinoma (SCLC) in patients presenting with a subacute paraneoplastic neurologic disorder.

Other autoantibody markers of immune responses to SCLC include ANNA-1, ANNA-2, ANNA-3, amphiphysin, collapsin response-mediated protein-5 (CRMP-5)-IgG, anti-glial/neuronal nuclear antibody-type 1 (AGNA-1), neuronal calcium channel antibodies (N-type > P/Q-type), ganglionic acetylcholine receptor antibodies, muscle acetylcholine receptor antibodies, neuronal potassium channel antibodies, and striational antibodies.

Reference Values
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-DMS2 / Dementia, Autoimmune/Paraneoplastic Evaluation, Serum
-ENS2 / Encephalopathy, Autoimmune/Paraneoplastic Evaluation, Serum
-EPS2 / Epilepsy, Autoimmune/Paraneoplastic Evaluation, Serum
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-AIAES / Axonal Neuropathy, Autoimmune/Paraneoplastic Evaluation, Serum
-DYS2 / Dysautonomia, Autoimmune/Paraneoplastic Evaluation, Serum
-GID2 / Gastrointestinal Dysmotility, Autoimmune/Paraneoplastic Evaluation, Serum

<1:240
Neuron-restricted patterns of IgG staining that do not fulfill criteria for Purkinje cell cytoplasmic antibody type 2 may be reported as "unclassified antineuronal IgG." Complex patterns that include non-neuronal elements may be reported as "uninterpretable."

Interpretation
A positive value (at 1:240 dilution or higher) is consistent with neurological autoimmunity and justifies a thorough search for a lung cancer, particularly small-cell carcinoma. The cancers are usually limited in metastasis. An extrapulmonary primary small-cell carcinoma (eg, skin, breast, larynx, cervix, prostate) should be considered.

Purkinje cell antibody type 2 is found in less than 2% of patients with uncomplicated small-cell lung carcinoma.

Cautions
Western blot with native neuronal proteins may be required to detect a positive result when interfering autoantibodies preclude interpretation of immunofluorescence pattern.

Supportive Data
Purkinje cell antibody type 2 (PCA-2) binds to the cytoplasm of cerebellar neurons in a characteristic pattern. Western blots of reduced/denatured cerebellar and small-cell lung carcinoma proteins reveal a common antigenic band, approximately 280 kDa.(1) Nine of 10 seropositive patients initially identified had a subacute neurological presentation (elements of encephalomyeloneuropathy), and 9 of 10 had lung cancer confirmed.(1) Similar neurological and oncological correlations have been observed in 104 subsequently identified seropositive patients.(VA Lennon, unpublished data)

Clinical Reference
1. Galanis E, Frytak S, Rowland KM, et al: Neuronal autoantibody titers in the course of small-cell lung carcinoma and platinum-associated neuropathy. Cancer Immunol Immunother. 1999 May-June;48(2-3):85-90
2. Vernino S, Lennon VA: New Purkinje cell antibody (PCA-2): marker of lung cancer-related neurological autoimmunity. Ann Neurol. 2000 Mar;47(3):297-305
3. McKeon A, Tracy JA, Pittock SJ, Parisi JE, Klein CJ, Lennon VA. Purkinje cell cytoplasmic autoantibody type 1 accompaniments: the cerebellum and beyond. Arch Neurol. 2011 Oct;68(10):1282-9. doi: 10.1001/archneurol.2011.128
4. Pittock SJ, Kryzer TJ, Lennon VA: Paraneoplastic antibodies coexist and predict cancer, not neurological syndrome. Ann Neurol. 2004 Nov;56(5):715-719

Performance

Method Description
The patient's specimen is tested by a standardized immunofluorescence assay that uses a composite frozen section of

mouse cerebellum, kidney, and gut tissues. After incubation with the specimen and washing, fluorescein-conjugated goat-antihuman IgG is applied. Neuron-specific autoantibodies are identified by their characteristic fluorescence staining patterns. Specimens that are scored positive for any neuronal nuclear or cytoplasmic autoantibody are titrated. Interference by coexisting non-neuron-specific autoantibodies can usually be eliminated by serologic absorption.(Honorat JA, Komorowski L, Josephs KA, et al. IgLON5 antibody: Neurological accompaniments and outcomes in 20 patients. Neurol Neuroimmunol Neuroinflamm. 2017;4[5]:e385. Published 2017 Jul 18. doi:10.1212/NXI.0000000000000385)

PDF Report
No

Day(s) Performed
Monday through Sunday

Report Available
5 to 8 days

Specimen Retention Time
2 days

Performing Laboratory Location
Mayo Clinic Laboratories - Rochester Main Campus

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 was developed and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. It has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information
86256

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
PC2TS	PCA-2 Titer, S	94351-4

Result ID	Test Result Name	Result LOINC® Value
43438	PCA-2 Titer, S	94351-4