
Overview**Useful For**

Identifying female patients whose subacute cerebellar degeneration or peripheral neuropathy is due to a remote (autoimmune) effect of gynecologic or breast carcinoma

Reporting an end titer result from spinal fluid specimens

Testing Algorithm

If the indirect immunofluorescence pattern suggests Purkinje cell cytoplasmic antibody type 1 (PCA-1), then this test will be performed at an additional charge.

Method Name

Only orderable as a reflex. For more information see:

ENC2 / Encephalopathy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

MDC2 / Movement Disorder, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

MAC1 / Myelopathy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

Indirect Immunofluorescence Assay (IFA)

NY State Available

Yes

Specimen**Specimen Type**

CSF

Ordering Guidance

Serum is preferred. Spinal fluid testing is particularly useful when interfering antibodies are present in the serum.

Specimen Required

Only orderable as a reflex. For more information see:

ENC2 / Encephalopathy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

MDC2 / Movement Disorder, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

MAC1 / Myelopathy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

Container/Tube: Sterile vial

Specimen Volume: 4 mL

Specimen Minimum Volume

1 mL

Reject Due To

| | |
|-----------------|--------|
| Gross hemolysis | Reject |
| Gross lipemia | Reject |
| Gross icterus | Reject |

Specimen Stability Information

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

Clinical & Interpretive
Clinical Information

Purkinje cell cytoplasmic autoantibody type 1 (PCA-1), also known as anti-Yo, binds to Purkinje cell cytoplasm in a characteristic pattern by indirect immunofluorescence. It is found in the serum, and usually cerebrospinal fluid, of patients with paraneoplastic cerebellar degeneration associated with gynecological or breast carcinoma. It is also found in some patients with sensory, sensorimotor, or motor neuropathy with some gynecologic cancer. Almost all (99%) seropositive patients are women.

Reference Values

Only orderable as a reflex. For more information see:

ENC2 / Encephalopathy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

MDC2 / Movement Disorder, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

MAC1 / Myelopathy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

<1:2

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

Interpretation

Purkinje cell cytoplasmic autoantibody type 1 (PCA-1) has not been found in any healthy subject. It is rarely found in patients with neurologic diseases (including cerebellar disorders) without gynecologic or breast cancer. The ovarian cancers found in these patients are typically limited in metastatic spread and may not be detected by imaging procedures. If mammography is negative, exploratory laparotomy is advised (as a "second look" in management of ovarian carcinoma). Breast carcinoma may coexist with a Mullerian cancer. PCA-1 is rarely found in patients with gynecologic cancer without neurologic dysfunction (<2%). PCA-1 is readily distinguished from PCA-Tr (a marker of Hodgkin lymphoma) and PCA-2 (a marker of small-cell lung

carcinoma) by standardized staining criteria. PCA-1 rarely, if ever, has accompanying neuronal cytoplasmic or nuclear antibodies.

Cautions

Purkinje cell cytoplasmic antibody type 1 (PCA-1) is rarely found in male patients (1%, usually with intra-abdominal adenocarcinoma) and never in patients with cerebellar ataxia associated with lung cancer.

Seven different IgG autoantibodies are currently recognized as accompaniments of paraneoplastic neurologic disorders occurring with small-cell lung carcinoma (SCLC):

- Antineuronal nuclear antibody-type 1 (ANNA-1, sometimes called anti-Hu) is found most often with sensory, autonomic, and sensorimotor neuropathies, and encephalomyeloradiculopathies in the context of SCLC
- ANNA-2 (sometimes called anti-Ri) is found most often with midbrain/brainstem encephalitis, cerebellar ataxia, myelopathy associated with breast cancer, or SCLC; peripheral neuropathy may be a presenting sign
- ANNA-3 is found with multifocal autoimmune neurologic manifestations of aerodigestive carcinomas (usually SCLC)
- PCA-2 is found with multifocal autoimmune neurologic manifestations of SCLC
- PCA-Tr is found in patients with cerebellar ataxia related to Hodgkin lymphoma
- Collapsin response-mediator protein-5 neuronal (CRMP-5-IgG) is found in patients with multifocal autoimmune neurologic manifestations of SCLC or neuromuscular or encephalopathic manifestations of thymoma
- Antigliial neuronal antibody (AGNA-1) is found in patients with multifocal autoimmune neurologic manifestations of SCLC, but particularly with Lambert-Eaton syndrome, peripheral neuropathy, limbic encephalitis, and dysautonomia

Clinical Reference

1. Hetzel DJ, Stanhope CR, O'Neill BP, Lennon VA: Gynecologic cancer in patients with subacute cerebellar degeneration predicted by anti-Purkinje cell antibodies and limited in metastatic volume. *Mayo Clin Proc.* 1990 Dec;65(12):1558-1563
2. 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
3. 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
4. Yu Z, Kryzer TJ, Griesmann GE, et al: CRMP-5 neuronal autoantibody: marker of lung cancer and thymoma-related autoimmunity. *Ann Neurol.* 2001 Feb;49(2):146-154
5. Pittock SJ, Kryzer TJ, Lennon VA: Paraneoplastic antibodies coexist and predict cancer, not neurological syndrome. *Ann Neurol.* 2004 Nov;56(5):715-719
6. Horta ES, Lennon VA, Lachance DH, et al: Neural autoantibody clusters aid diagnosis of cancer. *Clin Cancer Res.* 2014 Jul;20[14]:3862-3869

Performance**Method Description**

The patient's sample is tested by a standardized immunofluorescence assay that uses a composite frozen section of mouse cerebellum, kidney, and gut tissues. After incubation with sample and washing, fluorescein-conjugated goat-antihuman IgG is applied. Neuron-specific autoantibodies are identified by their

characteristic fluorescence staining patterns. Samples that are scored positive for any neuronal nuclear or cytoplasmic autoantibody are titrated to an endpoint. 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 Jul 18;4(5):e385. doi: 10.1212/NXI.0000000000000385)

PDF Report

No

Day(s) Performed

Monday through Sunday

Report Available

6 to 8 days

Specimen Retention Time

28 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 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 |
|---------|------------------|--------------------|
| PC1TC | PCA-1 Titer, CSF | 94363-9 |

| Result ID | Test Result Name | Result LOINC® Value |
|-----------|------------------|---------------------|
| 43446 | PCA-1 Titer, CSF | 94363-9 |