

## Overview

### Useful For

Serological testing for Purkinje cell cytoplasmic antibody-Tr for patients with acquired cerebellar ataxia of undetermined etiology, particularly if the patient has a history of Hodgkin lymphoma

Reporting an end titer result from spinal fluid specimens

### Testing Algorithm

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

### Method Name

Only orderable as a reflex. For more information see:

DMC2 / Dementia, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

ENC2 / Encephalopathy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

EPC2 / Epilepsy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

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

PCDEC / Pediatric Autoimmune Encephalopathy/CNS Disorder 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:

-DMC2 / Dementia, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

-ENC2 / Encephalopathy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

-EPC2 / Epilepsy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

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

-PCDEC / Pediatric Autoimmune Encephalopathy/CNS Disorder Evaluation, Spinal Fluid

**Container/Tube:** Sterile vial

**Specimen Volume:** 4 mL

**Specimen Minimum Volume**

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

The earliest description of a Purkinje cell cytoplasmic antibody (PCA) was reported by Trotter et al in 1976 as a serological accompaniment of paraneoplastic cerebellar ataxia in a patient with Hodgkin lymphoma.(1) IgG of that specificity was recently characterized more fully by Graus et al,(2) who confirmed the association with Hodgkin lymphoma and named the antibody "anti-Tr" in recognition of Dr. John L. Trotter's original report.

To be consistent with a generic classification of neuronal nuclear and cytoplasmic autoantibodies,(3) we introduced the name PCA-Tr to distinguish this Purkinje cell cytoplasmic antibody from PCA-1 (a marker of ovarian or breast carcinoma) and PCA-2 (a marker of small-cell lung carcinoma),(4) which also are found in patients presenting with paraneoplastic neurological autoimmunity.

**Reference Values**

Only orderable as a reflex. For more information see:

- DMC2 / Dementia, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid
- ENC2 / Encephalopathy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid
- EPC2 / Epilepsy, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid
- MDC2 / Movement Disorder, Autoimmune/Paraneoplastic Evaluation, Spinal Fluid

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-PCDEC / Pediatric Autoimmune Encephalopathy/CNS Disorder Evaluation, Spinal Fluid

<1:2

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

**Interpretation**

A positive value (at 1:2 dilution or higher) is consistent with neurological autoimmunity and justifies a search for Hodgkin lymphoma. Purkinje cell cytoplasmic antibody-Tr (PCA-Tr) has not yet been identified in any other context.

Seropositive patients usually have Hodgkin lymphoma and present with subacute cerebellar ataxia.(1-3)

**Cautions**

Interference from coexisting autoantibodies may preclude interpretation of the immunofluorescence pattern.

A negative result does not exclude neurological autoimmunity or Hodgkin lymphoma.

The Purkinje cell cytoplasmic antibody-Tr (PCA-Tr) antigen has not been defined immunochemically. It has been reported, but not yet confirmed that autoantibodies against glutamate receptors may occur in this context.(4)

No Western blot characteristics have been defined for the PCA-Tr antigen.

**Clinical Reference**

1. Trotter JL, Hendin BA, Osterland CK: Cerebellar degeneration with Hodgkin disease. An immunological study. *Arch Neurol.* 1976 Sept;33(9):660-661
2. Graus F, Gultekin SH, Ferrer I, Reiriz J, Alberch J, Dalmau J: Localization of the neuronal antigen recognized by anti-Tr antibodies from patients with paraneoplastic cerebellar degeneration and Hodgkin's disease in the rat nervous system. *Acta Neuropathologica.* 1998 July;96(1):1-7
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. Graus F, Vincent A, Pozo-Rosich P, et al: Anti-glial nuclear antibody: marker of lung cancer-related paraneoplastic neurological syndromes. *J Neuroimmunol.* 2005 Aug;165(1-2):166-171
5. Klein CJ: Autoimmune-mediated peripheral neuropathies and autoimmune pain. In: Pittock SJ, Vincent A, eds. *Autoimmune Neurology.* Elsevier; 2016:417-446. Aminoff MJ, Boller F, Swaab DF, eds. *Handbook of Clinical Neurology;* vol 133

**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 |
|---------|-------------------|--------------------|
| PCTTC   | PCA-Tr Titer, CSF | 94362-1            |

| Result ID | Test Result Name  | Result LOINC® Value |
|-----------|-------------------|---------------------|
| 43448     | PCA-Tr Titer, CSF | 94362-1             |