



# Test Definition: PCDES

Pediatric Autoimmune Encephalopathy/CNS Disorder Evaluation, Serum

## Overview

### Useful For

Evaluating children with autoimmune central nervous system disorders using serum specimens

### Profile Information

| Test Id | Reporting Name                       | Available Separately | Always Performed |
|---------|--------------------------------------|----------------------|------------------|
| PCSI    | Peds Autoimmune CNS Interp, S        | No                   | Yes              |
| AMPCS   | AMPA-R Ab CBA, S                     | No                   | Yes              |
| ANN1S   | Anti-Neuronal Nuclear Ab, Type 1     | No                   | Yes              |
| CS2CS   | CASPR2-IgG CBA, S                    | No                   | Yes              |
| DPPCS   | DPPX Ab CBA, S                       | No                   | Yes              |
| GABCS   | GABA-B-R Ab CBA, S                   | No                   | Yes              |
| GD65S   | GAD65 Ab Assay, S                    | Yes                  | Yes              |
| GFAIS   | GFAP IFA, S                          | No                   | Yes              |
| LG1CS   | LGI1-IgG CBA, S                      | No                   | Yes              |
| GL1IS   | mGluR1 Ab IFA, S                     | No                   | Yes              |
| MOGFS   | MOG FACS, S                          | Yes                  | Yes              |
| NCDIS   | Neurochondrin IFA, S                 | No                   | Yes              |
| NMDCS   | NMDA-R Ab CBA, S                     | No                   | Yes              |
| NMOFS   | NMO/AQP4 FACS, S                     | Yes                  | Yes              |
| PCATR   | Purkinje Cell Cytoplasmic Ab Type Tr | No                   | Yes              |

### Reflex Tests

| Test Id | Reporting Name              | Available Separately | Always Performed |
|---------|-----------------------------|----------------------|------------------|
| AN1BS   | ANNA-1 Immunoblot, S        | No                   | No               |
| AN2BS   | ANNA-2 Immunoblot, S        | No                   | No               |
| DPPTS   | DPPX Ab IFA Titer, S        | No                   | No               |
| GFACS   | GFAP CBA, S                 | No                   | No               |
| GFATS   | GFAP IFA Titer, S           | No                   | No               |
| GL1CS   | mGluR1 Ab CBA, S            | No                   | No               |
| GL1TS   | mGluR1 Ab IFA Titer, S      | No                   | No               |
| MOGTS   | MOG FACS Titer, S           | No                   | No               |
| NMDIS   | NMDA-R Ab IF Titer Assay, S | No                   | No               |
| NMOTS   | NMO/AQP4 FACS Titer, S      | No                   | No               |

|       |                               |    |    |
|-------|-------------------------------|----|----|
| PCTBS | PCA-Tr Immunoblot, S          | No | No |
| AN1TS | ANNA-1 Titer, S               | No | No |
| GABIS | GABA-B-R Ab IF Titer Assay, S | No | No |
| NCDCS | Neurochondrin CBA, S          | No | No |
| NCDTS | Neurochondrin IFA Titer, S    | No | No |
| PCTTS | PCA-Tr Titer, S               | No | No |
| AMPIS | AMPA-R Ab IF Titer Assay, S   | No | No |

## Testing Algorithm

Results from this test can be entered into the [TRUE-MOGAD calculator](#), a practical, weighted, point-based tool that helps distinguish myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) among patients with positive MOG-IgG testing.

If the indirect immunofluorescence assay (IFA) pattern suggests antineuronal nuclear antibody type 1 (ANNA-1), then the ANNA-1 immunoblot, ANNA-1 IFA titer and ANNA-2 immunoblot will be performed at an additional charge.

If the IFA pattern suggests Purkinje cytoplasmic antibody (PCA)-Tr, then the PCA-Tr immunoblot and PCA-Tr IFA titer will be performed at an additional charge.

If the N-methyl-D-aspartate receptor (NMDA-R) antibody cell binding assay (CBA) result is positive, then the NMDA-R IFA titer will be performed at an additional charge.

If the gamma-aminobutyric acid B receptor (GABA-B-R) antibody CBA result is positive, then the GABA-B-R IFA titer will be performed at an additional charge.

If the dipeptidyl-peptidase-like protein-6 (DPPX) antibody CBA result is positive, then the DPPX IFA titer will be performed at an additional charge.

If the IFA pattern suggests metabotropic glutamate receptor 1 (mGluR1) antibody, then the mGluR1 antibody CBA and mGluR1 IFA titer will be performed at an additional charge.

If the IFA pattern suggests glial fibrillary acidic protein (GFAP) antibody, then the GFAP antibody CBA and GFAP IFA titer will be performed at an additional charge.

If the neuromyelitis optica/aquaporin-4-IgG (NMO/AQP4-IgG) fluorescence-activated cell sorting (FACS) screen assay requires further investigation, then the NMO/AQP4-IgG FACS titration assay will be performed at an additional charge.

If the myelin oligodendrocyte glycoprotein (MOG) FACS screen assay requires further investigation, then the MOG FACS titration assay will be performed at an additional charge.

If IFA pattern suggests neurochondrin antibody, then the neurochondrin antibody CBA and neurochondrin IFA titer will

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be performed at an additional charge.

If alpha-amino-3-hydroxy-5 methyl-4-isoxazolepropionic acid (AMPA)-receptor antibody CBA result is positive, then AMPA-receptor antibody IFA titer assay will be performed at an additional charge.

For more information, see the following:

[Pediatric Autoimmune Encephalopathy/Central Nervous System Disorders Evaluation Algorithm-Serum](#)  
[Pediatric Autoimmune Central Nervous System Demyelinating Disease Diagnostic Algorithm](#)

**Special Instructions**

- [Pediatric Autoimmune Encephalopathy/CNS Disorders Evaluation Algorithm-Serum](#)
- [Pediatric Autoimmune Central Nervous System Demyelinating Disease Diagnostic Algorithm](#)

**Method Name**

AMPCS, CS2CS, DPPCS, GABCS, GFACS, LG1CS, GL1CS, NCDCS, NMDCS: Cell-Binding Assay (CBA)

MOGFS, MOGTS, NMOFS, NMOTS: Flow Cytometry

AMPIS, ANN1S, AN1TS, DPPTS, GABIS, GFAIS, GFATS, GL1IS, GL1TS, NCDIS, NCDTS, NMDIS, PCATR, PCTTS: Indirect Immunofluorescence (IFA)

GD65S: Radioimmunoassay (RIA)

AN1BS, AN2BS, PCTBS: Immunoblot (IB)

PCSI: Medical Interpretation

**NY State Available**

Yes

**Specimen****Specimen Type**

Serum

**Ordering Guidance**

Multiple neurological phenotype-specific autoimmune/paraneoplastic evaluations are available. For more information as well as phenotype-specific testing options, see [Autoimmune Neurology Test Ordering Guide](#).

When more than one evaluation is ordered on the same order number, the duplicate test will be canceled.

For a list of antibodies performed with each evaluation, see [Autoimmune Neurology Antibody Matrix](#).

This test **should not be requested** for patients who have recently received radioisotopes, therapeutically or diagnostically, because of potential assay interference. The specific waiting period before specimen collection will depend on the isotope administered, the dose given, and the clearance rate in the individual patient. Specimens will be screened for radioactivity prior to analysis. Radioactive specimens received in the laboratory will be held 1 week and assayed if sufficiently decayed or canceled if radioactivity remains.

### Necessary Information

Provide the following information:

- Relevant clinical information
- Ordering healthcare professional's name, phone number, mailing address, and email address

### Specimen Required

**Patient Preparation:** For optimal antibody detection, specimen collection is recommended before starting immunosuppressant medication or intravenous immunoglobulin (IVIg) treatment.

**Supplies:** Sarstedt Aliquot Tube, 5 mL (T914)

**Collection Container/Tube:**

**Preferred:** Red top

**Acceptable:** Serum gel

**Submission Container/Tube:** Plastic vial

**Specimen Volume:** 4 mL

**Collection Instructions:** Centrifuge and aliquot serum into a plastic vial.

### Forms

[If not ordering electronically, complete, print, and send a Neurology Specialty Testing Client Test Request \(T732\)](#) with the specimen.

### 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 |
|---------------|--------------------------|----------|-------------------|
| Serum         | Refrigerated (preferred) | 28 days  |                   |
|               | Ambient                  | 72 hours |                   |
|               | Frozen                   | 28 days  |                   |

## Clinical & Interpretive

### Clinical Information

Autoimmune encephalitis and myelitis is increasingly recognized as a cause of central nervous system disease in children and adolescents. N-methyl-D-aspartate receptor antibody (NMDA-R) encephalitis and myelin oligodendrocyte glycoprotein (MOG) autoimmunity are most common, though other entities, including aquaporin-4 autoimmunity, contactin-associated protein-like 2 (CASPR2) autoimmunity, autoimmune glial fibrillary acidic protein (GFAP) astrocytopathy, and paraneoplastic encephalomyelopathies, may also occur in children.

### Reference Values

| Test ID | Reporting name                       | Methodology*           | Reference value  |
|---------|--------------------------------------|------------------------|--|
| PCSI    | Peds Autoimmune CNS Interp, S        | Medical interpretation | Interpretive report                                      |
| AMPCS   | AMPA-R Ab CBA, S                     | CBA                    | Negative   |
| ANN1S   | Anti-Neuronal Nuclear Ab, Type 1     | IFA                    | Negative   |
| CS2CS   | CASPR2-IgG CBA, S                    | CBA                    | Negative   |
| DPPCS   | DPPX Ab CBA, S                       | CBA                    | Negative   |
| GABCS   | GABA-B-R Ab CBA, S                   | CBA                    | Negative   |
| GD65S   | GAD65 Ab Assay, S                    | RIA                    | < or =0.02 nmol/L<br>Reference values apply to all ages. |
| GFAIS   | GFAP IFA, S                          | IFA                    | Negative   |
| LG1CS   | LGI1-IgG CBA, S                      | CBA                    | Negative   |
| GL1IS   | mGluR1 Ab IFA, S                     | IFA                    | Negative   |
| NCDIS   | Neurochondrin IFA, S                 | IFA                    | Negative   |
| MOGFS   | MOG FACS, S                          | FACS                   | Negative   |
| NMDCS   | NMDA-R Ab CBA, S                     | CBA                    | Negative   |
| NMOFS   | NMO/AQP4 FACS, S                     | FACS                   | Negative   |
| PCATR   | Purkinje Cell Cytoplasmic Ab Type Tr | IFA                    | Negative   |

### Reflex Information:

| Test ID | Reporting name                | Methodology | Reference value |
|---------|-------------------------------|-------------|-----------------|
| AMPIS   | AMPA-R Ab IF Titer Assay, S   | IFA         | <1:240          |
| AN1BS   | ANNA-1 Immunoblot, S          | IB          | Negative        |
| AN1TS   | ANNA-1 Titer, S               | IFA         | <1:240          |
| AN2BS   | ANNA-2 Immunoblot, S          | IB          | Negative        |
| DPPTS   | DPPX Ab IFA Titer, S          | IFA         | <1:240          |
| GABIS   | GABA-B-R Ab IF Titer Assay, S | IFA         | <1:240          |
| GFACS   | GFAP CBA, S                   | CBA         | Negative        |
| GFATS   | GFAP IFA Titer, S             | IFA         | <1:240          |

|       |                             |      |          |
|-------|-----------------------------|------|----------|
| GL1CS | mGluR1 Ab CBA, S            | CBA  | Negative |
| GL1TS | mGluR1 Ab IFA Titer, S      | IFA  | <1:240   |
| MOGTS | MOG FACS Titer, S           | FACS | <1:20    |
| NCDCS | Neurochondrin CBA, S        | CBA  | Negative |
| NCDTS | Neurochondrin IFA Titer, S  | IFA  | <1:240   |
| NMDIS | NMDA-R Ab IF Titer Assay, S | IFA  | <1:240   |
| NMOTS | NMO/AQP4 FACS Titer, S      | FACS | <1:5     |
| PCTTS | PCA-Tr Titer, S             | IFA  | <1:240   |
| PCTBS | PCA-Tr Immunoblot, S        | IB   | Negative |

**\*Methodology abbreviations:**

Immunofluorescence assay (IFA)

Cell-binding assay (CBA)

Fluorescence-activated cell sorting assay (FACS)

Radioimmunoassay (RIA)

Immunoblot (IB)

**\*\*Neuron-restricted patterns of IgG staining that do not fulfill criteria for ANNA-1, ANNA-2, or PCA-Tr may be reported as "unclassified anti-neuronal IgG." Complex patterns that include non-neuronal elements may be reported as "uninterpretable."**

### Interpretation

Autoantibodies targeting neuronal cell-surface or intracellular proteins expressed in the central nervous system (CNS) serve as important clinical markers for autoimmune encephalopathy and related CNS disorders in children. These antibodies are generally absent in healthy individuals and are commonly linked to a subacute onset of seizures, behavioral changes, movement disorders, cognitive issues, or other neurological symptoms. A positive result in this pediatric autoimmune encephalopathy panel indicates a possible immune-mediated cause and may suggest further evaluation for factors like post-infectious immune syndromes or occult neoplasia, alongside clinical assessment. However, negative results do not rule out autoimmune CNS disease.

A recent study evaluated which clinical, radiographic, and laboratory features best predict true-positive myelin oligodendrocyte glycoprotein (MOG) antibody results. The resulting TRUE-MOGAD score is a practical, weighted, point-based tool that helps distinguish MOG antibody-associated disease (MOGAD) among patients with positive MOG-IgG testing. It is particularly helpful in atypical or otherwise undifferentiated presentations and may support diagnostic and therapeutic decisions in clinical practice. However, it does not replace the 2023 MOGAD diagnostic criteria because it excludes key elements (eg, myelitis, brainstem/cerebellar syndromes, spinal fluid MOG-IgG); instead, it should be used as a complementary aid. To calculate your patient's TRUE MOGAD score, see the [TRUE-MOGAD calculator](#).

### Cautions

Negative results do not exclude a diagnosis of an autoimmune central nervous system disorder.

Intravenous immunoglobulin (IVIg) treatment prior to the serum collection may cause a false-positive result.

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**Clinical Reference**

1. Dubey D, Pittock SJ, Krecke KN, et al. Clinical, radiologic, and prognostic features of myelitis associated with myelin oligodendrocyte glycoprotein autoantibody. *JAMA Neurol.* 2019;76(3):301-309. doi:10.1001/jamaneurol.2018.4053
2. McKeon A, Lennon VA, Lotze T, et al. CNS aquaporin-4 autoimmunity in children. *Neurology.* 2008;71(2):93-100
3. Dubey D, Hinson SR, Jolliffe EA, et al. Autoimmune GFAP astrocytopathy: Prospective evaluation of 90 patients in 1 year. *J Neuroimmunol.* 2018;321:157-163. doi:10.1016/j.jneuroim.2018.04.016
4. Philipps G, Alisanski SB, Pranzatelli M, Clardy SL, Lennon VA, McKeon A. Purkinje cell cytoplasmic antibody type 1 (anti-Yo) autoimmunity in a child with Down syndrome. *JAMA Neurol.* 2014;71(3):347-349
5. Lopez-Chiriboga AS, Klein C, Zekeridou A, et al. LGI1 and CASPR2 neurological autoimmunity in children. *Ann Neurol.* 2018;84(3):473-480. doi:10.1002/ana.25310
6. Lopez-Chiriboga AS, Majed M, Fryer J, et al. Association of MOG-IgG serostatus with relapse after acute disseminated encephalomyelitis and proposed diagnostic criteria for MOG-IgG-associated disorders. *JAMA Neurol.* 2018;75(11):1355-1363. doi:10.1001/jamaneurol.2018.1814
7. Clardy SL, Lennon VA, Dalmau J, Childhood onset of stiff-man syndrome. *JAMA Neurol.* 2013;70(12):1531-1536. doi:10.1001/jamaneurol.2013.4442
8. Banwell B, Tenenbaum S, Lennon VA, et al. Neuromyelitis optica-IgG in childhood inflammatory demyelinating CNS disorders. *Neurology.* 2008;70(5):344-352. doi:10.1212/01.wnl.0000284600.80782.d5

**Performance****Method Description****Cell-Binding Assay:**

Patient sample is applied to a composite slide containing transfected and nontransfected EU90 cells. After incubation and washing, fluorescein-conjugated goat-antihuman IgG is applied to detect the presence of patient IgG binding. (Package insert: IIFT: Neurology Mosaics, Instructions for the indirect immunofluorescence test. EUROIMMUN; FA\_112d-1\_A\_UK\_C13, 02/25/2019)

**Fluorescence-Activated Cell Sorting Assay:**

Human embryonic kidney cells (HEK 293) are transfected transiently with a plasmid (pIRES2-*Aequorea coerulea* green fluorescent protein [AcGFP]) encoding both green fluorescent protein (AcGFP) and AQP4-M1. After 36 hours, a mixed population of cells (transfected expressing AQP4 or MOG on the surface and AcGFP in the cytoplasm and nontransfected lacking AQP4 or MOG and AcGFP) are lifted and resuspended in live cell-binding buffer. Cells are incubated with patient sample and an AlexaFluor 647-labeled secondary antibody is added. Two populations are gated on the basis of AcGFP expression: positive (high AQP4 or MOG expression) and negative (low or no AQP4 or MOG expression). Positivity is based on the ratio (positive >2.0) of the average median fluorescence intensity (MFI) of each cell population (MFI GFP positive:MFI GFP negative). (Unpublished Mayo method)

**Indirect Immunofluorescence Assay:**

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.

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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)

**Radioimmunoassay:**

(125)I-labeled recombinant human antigens or labeled receptors are incubated with patient sample. After incubation, anti-human IgG is added to form an immunoprecipitate. The amount of (125)I-labeled antigen in the immunoprecipitate is measured using a gamma-counter. The amount of gamma emission in the precipitate is proportional to the amount of antigen-specific IgG in the sample. Results are reported as units of precipitated antigen (nmol) per liter of patient sample. (Griesmann GE, Kryzer TJ, Lennon VA. Autoantibody profiles of myasthenia gravis and Lambert-Eaton myasthenic syndrome. In: Rose NR, Hamilton RG, et al, eds. *Manual of Clinical and Laboratory Immunology*. 6th ed ASM Press; 2002:1005-1012; Walikonis JE, Lennon VA. Radioimmunoassay for glutamic acid decarboxylase [GAD65] autoantibodies as a diagnostic aid for stiff-man syndrome and a correlate of susceptibility to type 1 diabetes mellitus. *Mayo Clin Proc*. 1998;73[12]:1161-1166; Jones AL, Flanagan EP, Pittock SJ, et al. Responses to and outcomes of treatment of autoimmune cerebellar ataxia in adults. *JAMA Neurol*. 2015;72[11]:1304-1312. doi:10.1001/jamaneurol.2015.2378)

**Immunoblot:**

All steps are performed at ambient temperature (18-28 degrees C) utilizing the EUROBlot One instrument. Diluted patient sample (1:101) is added to test strips (strips containing recombinant antigen manufactured and purified using biochemical methods) in individual channels and incubated for 30 minutes. Positive samples will bind to the purified recombinant antigen and negative samples will not bind. Strips are washed to remove unbound antibodies and then incubated with antihuman IgG antibodies (alkaline phosphatase-labeled) for 30 minutes. The strips are again washed to remove unbound antihuman IgG antibodies and nitroblue tetrazolium chloride/5-bromo-4-chloro-3-indolyl phosphate (NBT/BCIP) substrate is added. Alkaline phosphatase enzyme converts the soluble substrate into a colored insoluble product on the membrane to produce a black band. Strips are digitized via picture capture on the EUROBlot One instrument and evaluated with the EUROLineScan software. (O'Connor K, Waters P, Komorowski L, et al. GABAA receptor autoimmunity: A multicenter experience. *Neurol Neuroimmunol Neuroinflamm*. 2019;6[3]:e552 doi:10.1212/NXI.0000000000000552)

**PDF Report**

No

**Day(s) Performed**

Profile tests: Monday through Sunday; Reflex tests: Varies

**Report Available**

8 to 12 days

**Specimen Retention Time**

28 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

- 86341
- 86363
- 86053
- 86255 x 11
- 86256 AMPIS (if appropriate)
- 84182 AN1BS (if appropriate)
- 86256 AN1TS (if appropriate)
- 84182 AN2BS (if appropriate)
- 86256 DPPTS (if appropriate)
- 86256 GABIS (if appropriate)
- 86255 GFACS (if appropriate)
- 86256 GFATS (if appropriate)
- 86255 GL1CS (if appropriate)
- 86256 GL1TS (if appropriate)
- 86363 MOGTS (if appropriate)
- 86255 NCDCS (if appropriate)
- 86256 NCDTS (if appropriate)
- 86256 NMDIS (if appropriate)
- 86053 NMOTS (if appropriate)
- 84182 PCTBS (if appropriate)
- 86256 PCTTS (if appropriate)

### LOINC® Information

| Test ID | Test Order Name            | Order LOINC® Value |
|---------|----------------------------|--------------------|
| PCDES   | Peds Autoimm Enceph CNS, S | 101417-4           |

| Result ID | Test Result Name  | Result LOINC® Value |
|-----------|-------------------|---------------------|
| 80150     | ANNA-1, S         | 33615-6             |
| 81596     | GAD65 Ab Assay, S | 94345-6             |
| 83076     | PCA-Tr, S         | 84926-5             |

## Test Definition: PCDES

Pediatric Autoimmune Encephalopathy/CNS  
 Disorder Evaluation, Serum

|        |                               |          |
|--------|-------------------------------|----------|
| 61516  | NMDA-R Ab CBA, S              | 93503-1  |
| 61518  | AMPA-R Ab CBA, S              | 93489-3  |
| 61519  | GABA-B-R Ab CBA, S            | 93428-1  |
| 38324  | NMO/AQP4 FACS, S              | 43638-6  |
| 64279  | LGI1-IgG CBA, S               | 94287-0  |
| 64281  | CASPR2-IgG CBA, S             | 94285-4  |
| 65563  | MOG FACS, S                   | 90248-6  |
| 64933  | DPPX Ab CBA, S                | 94676-4  |
| 64928  | mGluR1 Ab IFA, S              | 94347-2  |
| 605155 | GFAP IFA, S                   | 94346-4  |
| 605131 | Peds Autoimmune CNS Interp, S | 69048-7  |
| 618907 | IFA Notes                     | 48767-8  |
| 615867 | Neurochondrin IFA, S          | 101452-1 |