# Test Definition: CONCS

Contactin-1 IgG Cell Binding Assay, Serum

## Overview

### Useful For

Evaluating for chronic inflammatory demyelinating polyradiculoneuropathy and related demyelinating peripheral neuropathies

Determining contactin-1 IgG results as a part of a profile

### Method Name

Only orderable as part of a profile. For more information see:
-CIDP / Chronic Inflammatory Demyelinating Polyradiculoneuropathy/Nodopathy Evaluation, Serum
-DMNES / Peripheral Nervous System Demyelinating Neuropathy, Autoimmune Evaluation, Serum

Cell Binding Assay (CBA)

### NY State Available

Yes

## Specimen

### Specimen Type

Serum

### Specimen Required

Only orderable as part of a profile. For more information see:
-CIDP / Chronic Inflammatory Demyelinating Polyradiculoneuropathy/Nodopathy Evaluation, Serum
-DMNES / Peripheral Nervous System Demyelinating Neuropathy, Autoimmune Evaluation, Serum

**Patient Preparation:** For optimal antibody detection, specimen collection is recommended to occur prior to starting immunosuppressant medication or intravenous immunoglobulin treatment (IVIg).
**Supplies:** Sarstedt Aliquot Tube, 5 mL (T914)
**Collection Container/Tube:**
**Preferred:** Red top
**Acceptable:** Serum gel
**Submission Container/Tube:** Plastic vial
**Specimen Volume:** 3 mL
**Collection Instructions:** Centrifuge and aliquot serum into a plastic vial

### Specimen Minimum Volume

0.5 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

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an acquired, immune-mediated condition effecting peripheral nerves and nerve roots and is characterized by electrodiagnostic features of demyelination with a chronic onset that leads to significant disability. The prevalence of CIDP is estimated at approximately 2 to 4 cases per 100,000 persons. Although a rarer cause of polyneuropathy, it is important to recognize as it is treatable with the appropriate use of immunomodulating therapies. Although the exact immunological trigger of CIDP remains unclear, a subset of patients with suspected CIDP have been identified with autoantibodies targeting nodal-paranodal proteins. These patients share common immunopathological mechanisms of disease, clinical features, and treatment responses that are distinct from classic CIDP. A common target of these autoantibodies is the neurofascin-155 (NF155)-contactin-1 (CNTN1) complex. NF155 is expressed at the paranodal loops of Schwann cells where it interacts with CNTN1 expressed on adjacent axons. This interaction stabilizes and allows the proper organization of the paranodal axoglial junction. Antibody-mediated disruption of this interaction in animal models recapitulates the pathophysiology observed in humans.

Contactin-1 IgG antibodies are present in approximately 2% of patients with CIDP-like presentations. CNTN1 IgG-positive cases are more likely to present with neuropathic pain, sensory ataxia, and subacute progressive demyelinating polyradiculoneuropathy or polyradiculopathy. The majority of seropositive patients have been reported to be refractory to treatment with intravenous immune globulin (IVIg). However, some of these patients respond well to B-cell depleting therapies such as rituximab. Up to half of CNTN1 IgG-positive patients with CIDP or CIDP-like presentations have been reported to develop membranous nephropathy and, thus, screening for proteinuria may be warranted.

### Reference Values

Only orderable as part of a profile. For more information see:
-CIDP / Chronic Inflammatory Demyelinating Polyradiculoneuropathy/Nodopathy Evaluation, Serum
-DMNES / Peripheral Nervous System Demyelinating Neuropathy, Autoimmune Evaluation, Serum

Negative

### Interpretation

Seropositivity for contactin-1 IgG is consistent with an immune-mediated demyelinating polyradiculoneuropathy/polyradiculopathy.

### Cautions

A negative result does not rule out an immune-mediated demyelinating disease.

This test should only be utilized in the appropriate clinical context.

The use of immunotherapy prior to sample collection may negatively impact the sensitivity of these assays.

### Clinical Reference

1. Dubey D, Honorat JA, Shelly S, et al. Contactin-1 autoimmunity: Serologic, neurologic, and pathologic correlates. Neurol Neuroimmunol Neuroinflamm. 2020;7(4):e771
2. Cortese A, Lombardi R, Briani C, et al. Antibodies to neurofascin, contactin-1, and contactin-associated protein 1 in CIDP: Clinical relevance of IgG isotype. Neurol Neuroimmunol Neuroinflamm. 2020;7(1):e639
3. Manso C, Querol L, Mekaouche M, Illa I, Devaux JJ. Contactin-1 IgG4 antibodies cause paranode dismantling and conduction defects. Brain. 2016;139(Pt 6):1700-1712
4. Le Quintrec M, Teisseyre M, Bec N, et al. Contactin-1 is a novel target antigen in membranous nephropathy associated with chronic inflammatory demyelinating polyneuropathy. Kidney Int. 2021;100(6):1240-1249
5. Ogata H, Yamasaki R, Hiwatashi A, et al. Characterization of IgG4 anti-neurofascin 155 antibody-positive polyneuropathy. Ann Clin Transl Neurol. 2015;2(10):960-971
6. Cortese A, Lombardi R, Briani C, et al. Antibodies to neurofascin, contactin-1, and contactin-associated protein 1 in CIDP: Clinical relevance of IgG isotype. Neurol Neuroimmunol Neuroinflamm. 2020;7(1):e639
7. Querol L, Nogales-Gadea G, Rojas-Garcia R, et al. Neurofascin IgG4 antibodies in CIDP associate with disabling tremor and poor response to IVIg. Neurology. 2014;82(10):879-886
8. Shelly S, Klein CJ, Dyck PJB, et al. Neurofascin-155 immunoglobulin subtypes: Clinicopathologic associations and neurologic outcomes. Neurology. 2021;97(24):e2392-e2403

## Performance

### Method Description

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)

### PDF Report

No

### Day(s) Performed

Monday through Thursday, 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

86255

### LOINC® Information

| Test ID | Test Order Name | Order LOINC® Value |
|---|---|---|
| CONCS | Contactin-1 IgG CBA, S | 101448-9 |

| Result ID | Test Result Name | Result LOINC® Value |
|---|---|---|
| 616442 | Contactin-1 IgG CBA, S | 101448-9 |