Test Definition: NF4FS

Neurofascin-155 IgG4, Flow Cytometry, Serum
(Non-Orderable)

Overview

Useful For

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

Evaluating patients with an underlying demyelinating neuropathy

Diagnosis of a neurofascin-155 IgG4 mediated neuropathy

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

Flow Cytometry (FCM)

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

Specimen Minimum Volume

See Specimen Required

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

Neurofascin-155 IgG4 antibodies are present in approximately 4% to 18% of patients with chronic inflammatory demyelinating neuropathy (CIDP) and, more rarely, in those with more acute forms of demyelinating neuropathy. This biomarker aids in the identification of a subset of these patients who are more likely to present with distal weakness, gait disturbance, tremor, and dysarthria as compared to classic CIDP. Most patients who are seropositive for neurofascin-155 IgG4 have been reported to be refractory to intravenous immune globulin (IVIG) therapy and often require second line treatment that includes B-cell depleting therapies such as rituximab. Studies in animal models, as well as the disease pathology indicate neurofascin-155 IgG4 antibodies directly disrupt the paranodal structure ultimately leading to demyelination. The presence of these antibodies, when detected, using flow cytometry is highly specific to CIDP and has not been reported in other disease mimics such as hereditary neuropathies, distal acquired demyelinating symmetric neuropathy, and motor neuron disease. This is test is useful in diagnostic work up of patients being evaluated for CIDP and related demyelinating peripheral neuropathies. This test should only be utilized in the appropriate clinical context.

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

A positive result is consistent with a neurofascin-155 IgG4 mediated demyelinating neuropathy.

Cautions

A negative result does not exclude the presence of disease. The use of immunotherapy prior to sample collection may negatively impact the sensitivity of this assay.

Clinical Reference

1. 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
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. 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

Performance

Method Description

This cell-binding assay utilizes flow cytometry to detect neurofascin 155 (NF155) IgG4 antibodies in patient sera. Briefly, a stable HEK293 cell line expressing human NF155 on the cell surface is premixed with parental HEK293 cells that do not express human NF155. The 2 cell populations are distinguished using a green fluorescent protein marker, which is only expressed in NF155 expressing cells. The mixture of cells is incubated with diluted patient sera to allow antibodies present in the sample to bind target antigens. Next the cells are incubated with a human IgG4 specific secondary antibody conjugated to AlexaFluor 647 to detect cell bound human IgG4 antibodies. The AlexaFluor 647 signal intensity of the different cell populations is measured using a flow cytometer. The IBI (IgG binding index) is then calculated as the median fluorescent intensity (MFI) of AlexaFluor 647 of the NF155 expressing cells divided by the MFI of the parental non-NF155 expressing cells. When the IBI is greater than or equal to 2.0 the result is considered positive for NF155 IgG4 antibodies.(Unpublished Mayo method)

PDF Report

No

Day(s) Performed

Monday, Friday

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
NF4FS	Neurofascin-155 IgG4, S	100845-7

Result ID	Test Result Name	Result LOINC® Value
614591	Neurofascin-155 IgG4, S	100845-7