Overview

Useful For
Evaluating patients with an underlying demyelinating neuropathy
Diagnosis of a neurofascin-155 IgG4 mediated neuropathy

Method Name
Flow Cytometry

NY State Available
Yes

Specimen

Specimen Type
Serum

Specimen Required
Collection Container/Tube:
Preferred: Red top
Acceptable: Serum gel

Specimen Volume: 2 mL

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

Reject Due To
- Gross hemolysis: Reject
- Gross lipemia: Reject
- Gross icterus: Reject

Specimen Minimum Volume
1 mL

Specimen Stability Information

<table>
<thead>
<tr>
<th>Specimen Type</th>
<th>Temperature</th>
<th>Time</th>
<th>Special Container</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum</td>
<td>Refrigerated (preferred)</td>
<td>28 days</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Frozen</td>
<td>28 days</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ambient</td>
<td></td>
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</tbody>
</table>

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 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**

**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**


**Performance**

**Method Description**

This is a cell-binding assay that 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

Specimen Retention Time
28 days

Performing Laboratory Location
Rochester

Fees & Codes

Test Classification
This test was developed, and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information
86255