Overview

Useful For
Evaluating patients with paraneoplastic or autoimmune encephalitis (brainstem encephalitis or limbic encephalitis or cerebellar ataxia) using serum specimens

Testing Algorithm
If the cell binding antibody result is reactive, then the immunofluorescence titer assay will be performed at an additional charge.

Reflex Tests

<table>
<thead>
<tr>
<th>Test Id</th>
<th>Reporting Name</th>
<th>Available Separately</th>
<th>Always Performed</th>
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<tbody>
<tr>
<td>K11TS</td>
<td>KLHL11 Ab IFA Titer, S</td>
<td>No</td>
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Method Name
Cell-Binding Assay (CBA)

NY State Available
Yes

Specimen

Specimen Type
Serum

Specimen Required
Container/Tube:
Preferred: Red top
Acceptable: Serum gel
Specimen Volume: 1 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
0.5 mL

Specimen Stability Information
**Clinical & Interpretive**

**Clinical Information**
Kelch-like protein 11 (KLHL11 or Kelch-like family member 11) IgG is a biomarker of paraneoplastic encephalitis. KLHL11 encephalitis is a unique paraneoplastic syndrome commonly associated with testicular germ cell tumors mainly seminoma. Ataxia, diplopia, dysarthria, and vertigo are common presenting features of the rhombencephalitis phenotype. Hearing loss and tinnitus may precede other neurological signs and symptoms by weeks to months. A subset of patients also has clinical and magnetic resonance imaging (MRI) presentations consistent with limbic encephalitis. Most patients with this syndrome have inflammatory spinal fluid profiles, especially elevated oligoclonal bands. MRI brain demonstrates T2 fluid attenuated inversion recovery (T2/FLAIR) abnormalities involving the brainstem or limbic system. The accompanying neurological disorder is usually severe. Clinical improvement following treatment of cancer or immunotherapy has been reported.

**Reference Values**
Negative

**Interpretation**
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**Cautions**
A negative Kelch-like protein 11 (KLHL11) antibody test result does not exclude autoimmune neurological disease or cancer.

**Clinical Reference**

**Performance**

**Method Description**
Cell Binding Assay:
Methodology for detecting Kelch-like protein 11 (KLHL11)-IgG uses a cell binding assay (CBA) with confirmation by tissue immunofluorescence (IFA). The CBA utilizes HEK293 cells that are stably transfected with DNA encoding the KLHL11 protein that has been tagged with green fluorescent protein (GFP). Since KLHL11 is localized to cytoplasmic vesicles when ectopically expressed, cells will be fixed and permeabilized prior to exposure to patient sample. Patients that are positive for KLHL11-IgG will have human IgG bound to the transfected cells. Binding will colocalize with the GFP-tagged...
KLHL11 protein in cytoplasmic vesicles. Patient IgG will be detected using a tetramethylrhodamine (TRITC)-conjugated anti-human secondary antibody. The negative samples will not bind to KLHL11-GFP in transfected cells. Performed in a 96 well plate format, the plates are scanned and images saved using the ImageXpress Micro Confocal High-Content Imaging System (Molecular Devices). Images will be scored positive or negative. (Unpublished Mayo method)

Indirect Immunofluorescence Assay:
Tissue IFA utilizes mouse composite slides including brain, kidney, and stomach tissue sections, which are commercially purchased. After tissue sections are fixed and permeabilized, patient sample is added to the well. After washing with phosphate buffer saline, bound human IgG is detected with a fluorescent conjugated secondary antibody targeting human IgG. Slides are read under a fluorescent microscope for the unique tissue-specific staining pattern characteristic of KLHL11-IgG. (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
86256 (if appropriate)