

## Overview

### Useful For

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

### Reflex Tests

| Test Id | Reporting Name         | Available Separately | Always Performed |
|---------|------------------------|----------------------|------------------|
| K11TS   | KLHL11 Ab IFA Titer, S | No                   | No               |

### Testing Algorithm

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

### Method Name

Cell-Binding Assay (CBA)

### NY State Available

Yes

## Specimen

### Specimen Type

Serum

### Specimen Required

#### Collection Container/Tube:

**Preferred:** Red top

**Acceptable:** Serum gel

**Submission Container/Tube:** Plastic vial

**Specimen Volume:** 1 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

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  |                   |
|               | Frozen                   | 28 days  |                   |
|               | Ambient                  | 72 hours |                   |

**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**

- Mandel-Brehm C, Dubey D, Kryzer TJ, et al: Kelch-like protein 11 antibodies in seminoma-associated paraneoplastic encephalitis. *N Engl J Med.* 2019;381:47-54
- Dubey D, Wilson MR, Clarkson B, et al: Expanded clinical phenotype, oncological associations, and immunopathologic

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insights of paraneoplastic Kelch-like protein-11 encephalitis. JAMA Neurol. 2020 Aug 3;77(11):1-10

## 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 the 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

### Day(s) Performed

Varies

### Report Available

3 to 5 days

### Specimen Retention Time

28 days

### Performing Laboratory Location

Rochester

## 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**

0432U

**LOINC® Information**

| Test ID | Test Order Name  | Order LOINC® Value |
|---------|------------------|--------------------|
| K11CS   | KLHL11 Ab CBA, S | 99072-1            |

| Result ID | Test Result Name | Result LOINC® Value |
|-----------|------------------|---------------------|
| 610581    | KLHL11 Ab CBA, S | 99072-1             |