# Test Definition: KLHCS

Kelch-Like Protein 11 Antibody, Cell Binding Assay, Serum

## Overview

### Useful For

Positivity for Kelch-like protein 11 (KLHL11)-IgG is indicative of a paraneoplastic neurological syndrome. Positivity indicates a high likelihood of finding a testicular cancer. A rigorous search for cancer should be initiated after KLHL11 autoimmunity is confirmed.

### 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

**Patient Preparation:** For optimal antibody detection, specimen collection is recommended before starting immunosuppressant medication or intravenous immunoglobulin (IVIg) treatment.
**Supplies**: Sarstedt Aliquot Tube, 5 mL (T914)
**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.

### 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

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 of the 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

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

### Cautions

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

### Clinical Reference

1. 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

2. Dubey D, Wilson MR, Clarkson B, et al. Expanded clinical phenotype, oncological associations, and immunopathologic

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

## 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: EUROIMMUN; FA_110a-50_A_UK_B01, 12/2023)

### PDF Report
No

### Day(s) Performed
Monday through Sunday

### Report Available
3 to 5 days

### Specimen Retention Time
28 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
0432U

### LOINC® Information

| Test ID | Test Order Name | Order LOINC® Value |
|---|---|---|

| | | |
|---|---|---|
| KLHCS | KLHL11-IgG CBA, S | 99072-1 |

| Result ID | Test Result Name | Result LOINC® Value |
|---|---|---|
| 622117 | KLHL11-IgG CBA, S | 99072-1 |