Test Definition: K11CS

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

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	
	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 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 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

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
K11CS	KLHL11 Ab CBA, S	99072-1

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