Test Definition: KLHCC

Kelch-Like Protein 11 Antibody, Cell Binding Assay, Spinal Fluid

Overview

Useful For

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

Reflex Tests

Test Id	Reporting Name	Available Separately	Always Performed
K11TC	KLHL11 Ab IFA Titer, CSF	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

CSF

Specimen Required

Patient Preparation: For optimal antibody detection, specimen collection is recommended before initiation of immunosuppressant medication, or corticosteroid or intravenous immunoglobulin (IVIg) treatment.
Container/Tube: Sterile vial
Preferred: Collection vial number 1
Acceptable: Any collection vial
Specimen Volume: 1 mL

Specimen Minimum Volume

1 mL

Reject Due To

Gross	Reject

hemolysis	
Gross lipemia	Reject
Gross icterus	Reject

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
CSF	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

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.

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(1):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):1420-1429

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Performance

Method Description

Patient specimen 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

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
KLHCC	KLHL11-IgG CBA, CSF	99073-9

Result ID	Test Result Name	Result LOINC® Value
622118	KLHL11-IgG CBA, CSF	99073-9