MAYO CLINIC LABORATORIES

Test Definition: ALPRT

Alport (Collagen IV Alpha 5 and Alpha 2) Immunofluorescent Stain, Renal Biopsy

Overview

Useful For

Assisting in the diagnosis of hereditary nephritis (Alport syndrome)

Special Instructions

- Renal Biopsy Patient Information
- Renal Biopsy Preparation Instructions

Method Name

Direct Immunofluorescence

NY State Available

Yes

Specimen

Specimen Type

Special

Shipping Instructions

1. Advise shipping frozen specimens (unstained slides or tissue block) in Styrofoam transportation coolers filled with dry ice to ensure specimens are received at required specimen stability temperature.
2. Attach the green "Attention Pathology" address label (T498) to the outside of the transport container before putting into the courier mailer.

Necessary Information

A pathology/diagnostic report is required.

Specimen Required

Submit only 1 of the following specimens:

Preferred:
Specimen Type: Unstained slides (unfixed)
Source: Kidney tissue or skin tissue
Slides: 1 Slide
Collection Instructions: Submit 1 frozen tissue unstained positively charged glass slide (25- x 75- x 1-mm) per test ordered; sections should be 4-microns thick, **centered on the slide,** and shipped on dry ice.
Additional Information: Broken or cracked unstained slides will be rejected. Stained slides that were broken and repaired (ie, double mounted) at the client site will not be rejected, as, in most cases, they can still be reviewed under

the microscope. However, due to the double mounting, digital scanning cannot be performed.

Acceptable:
Specimen Type: Unfixed tissue block (O.C.T)
Source: Kidney tissue or skin tissue
Specimen Volume: Entire specimen
Collection Instructions:
1. Embed in O.C.T. compound.
2. Freeze specimen and ship on dry ice.

Acceptable:
Specimen Type: Wet tissue
Source: Kidney tissue or skin tissue
Supplies: Renal Biopsy Kit (T231)
Container/Tube: Renal Biopsy Kit, Zeus/Michel's
Specimen Volume: Entire specimen
Collection Instructions:
1. Collect specimens according to the instructions in Renal Biopsy Preparation Instructions.
2. If standard immunoglobulin and complement immunofluorescence has already been performed, ship the residual frozen tissue (must contain glomeruli) on dry ice.

Forms

1. Renal Biopsy Patient Information
2. If not ordering electronically, complete, print, and send 1 of the following forms with the specimen:
-Kidney Transplant Test Request
-Renal Diagnostics Test Request (T830)

Specimen Minimum Volume

See Specimen Required

Reject Due To

Broken or cracked unstained slides	Reject
Broken or cracked stained slides that cannot be repaired	Reject

All specimens will be evaluated at Mayo Clinic Laboratories for test suitability

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Special	Frozen		

Clinical & Interpretive

Clinical Information

Alport syndrome is a hereditary disease of basement membrane collagen type IV. Variants in collagen IV alpha genes cause characteristic abnormal immunofluorescence staining patterns within the glomerular basement membrane. Alport syndrome is characterized by hematuria, proteinuria, progressive kidney failure, and high-tone sensorineural hearing loss.

Reference Values

An interpretive report will be provided.

Interpretation

This test (when not accompanied by a pathology consultation request) will be reported as one of the following:
1) Normal pattern
2) Consistent with X-linked hereditary nephritis
3) Consistent with autosomal hereditary nephritis

If additional interpretation or analysis is needed, request PATHC / Pathology Consultation along with this test and send the corresponding renal pathology light microscopy and immunofluorescence (IF) slides (or IF images on a CD), electron microscopy images (prints or CD), and the pathology report.

Cautions

Approximately one-third of patients with established hereditary nephritis based on typical ultrastructural findings and family history show loss of glomerular basement membrane or epidermal basement membrane staining for the alpha 5 chain of type IV collagen. Therefore, a normal staining pattern does not exclude the diagnosis of hereditary nephritis.

Because alpha 3 and alpha 4 chains of type IV collagen are not expressed in the epidermal basement membranes, patients with autosomal hereditary nephritis have preserved staining for alpha 5 on epidermal basement membranes and, therefore, skin biopsy cannot exclude autosomal hereditary nephritis.

Broken or cracked unstained slides are not compatible with the instrumentation used in our performing laboratories and will be rejected. Additional material may be requested.

Attempts will be made to repair broken or cracked stained slides; however, if they are unable to be repaired (ie, shattered in transit), they will be rejected and discarded due to safety concerns.

Clinical Reference

1. Kagawa M, Kishiro Y, Naito I, et al. Epitope-defined monoclonal antibodies against type-IV collagen for diagnosis of Alport's syndrome. Nephrol Dial Transplant. 1997;12(6):1238-1241
2. Hashimura Y, Nozu K, Kaito H, et al. Milder clinical aspects of X-linked Alport syndrome in men positive for the

collagen IV alpha 5 chain. Kidney Int. 2014;85(5):1208-1213
3. Kamiyoshi N, Nozu K, Fu XJ, et al. Genetic, clinical, and pathologic backgrounds of patients with autosomal dominant Alport syndrome. Clin J Am Soc Nephrol. 2016;11(8):1441-1449
4. Said SM, Fidler ME, Valeri AM, et al. Negative staining for COL4A5 correlates with worse prognosis and more severe ultrastructural alterations in males with Alport syndrome. Kidney Int Rep. 2016;2(1):44-52

Performance

Method Description
Direct immunofluorescence staining on sections of fresh/frozen tissue.(Unpublished Mayo method)

PDF Report
No

Day(s) Performed
Monday through Friday

Report Available
1 to 2 days

Specimen Retention Time
Unstained slides: 2 weeks after results are reported; Stained slides: digital images are obtained for all slides used in testing and kept indefinitely; Unfixed tissue blocks: 5 years

Performing Laboratory Location
Mayo Clinical 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
88346

Test Definition: ALPRT

Alport (Collagen IV Alpha 5 and Alpha 2) Immunofluorescent Stain, Renal Biopsy

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
ALPRT	ALPORT Immunofluorescence	In Process

Result ID	Test Result Name	Result LOINC® Value
71285	Interpretation	50595-8
71268	Participated in the Interpretation	No LOINC Needed
71272	Material Received	81178-6
71271	Gross Description	22634-0
71269	Report electronically signed by	19139-5
71270	Addendum	35265-8
71619	Disclaimer	62364-5
71848	Case Number	80398-1