

## Overview

### Useful For

Assisting in the diagnosis of hereditary nephritis (Alport syndrome)

### Special Instructions

- [Renal Biopsy Patient Information](#)
- [Renal Biopsy Procedure for Handling Tissue for Light Microscopy, Immunofluorescent Histology, and Electron Microscopy](#)

### Method Name

Direct Immunofluorescence

### NY State Available

Yes

## Specimen

### Specimen Type

Special

### Shipping Instructions

1. Advise shipping specimens in Styrofoam transportation coolers to avoid extreme hot or cold temperatures to ensure specimens are received at required specimen stability temperature.
2. Attach the green pathology address label included in the kit to the outside of the transport container.

### Necessary Information

**A pathology/diagnostic report is required.**

### Specimen Required

Preferred: Frozen tissue

Supplies: Renal Biopsy Kit (T231)

Specimen Type: Kidney tissue

Container/Tube: Transport medium (Michel's or Zeus media), frozen tissue

Specimen Volume: Entire specimen

Collection Instructions:

1. Collect specimens according to the instructions in [Renal Biopsy Procedure for Handling Tissue for Light Microscopy, Immunofluorescent Histology, and Electron Microscopy](#).
2. If standard immunoglobulin and complement immunofluorescence has already been performed, submit the residual frozen tissue (must contain glomeruli) on dry ice.

Acceptable: Frozen tissue

Slides: 2 Frozen tissue unstained positively charged glass slides (25-x75-x1-mm) per test ordered; sections 4-microns thick, submitted on dry ice

Specimen Type: Skin tissue

Container/Tube: Transport medium (Michel's or Zeus media)

Specimen Volume: Entire specimen

Collection Instructions: Submit punch biopsy in Zeus/Michel's media.

Forms

1. [Renal Biopsy Patient Information](#)
2. If not ordering electronically, complete, print, and send a [Renal Diagnostics Test Request](#) (T830) with the specimen.

Reject Due To

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

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Special	Frozen (preferred)		
	Ambient		
	Refrigerated		

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:

- 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.

**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 Jun;12(6):1238-1241
2. Hashimura Y, Nozu K, Nakanishi K, 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 Aug 8;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; Sep 29;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

**Performing Laboratory Location**

Rochester

**Fees & Codes****Fees**

# Test Definition: ALPRT

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

- 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

## 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