MAYO CLINIC LABORATORIES

Test Definition: MSFGN

Fibrillary GN Confirm, LC MS/MS

Overview

Useful For

Diagnosis of fibrillary glomerulonephritis

Testing Algorithm

A pathology consultation is typically not required. If the results of this test do not support the clinical findings, a PATHC / Pathology Consultation may be added if appropriate, upon client approval.

Method Name

Liquid Chromatography-Tandem Mass Spectrometry (LC-MS/MS)

NY State Available

Yes

Specimen

Specimen Type

AMYLOID

Necessary Information

1. Preliminary pathology report and history are required.

2. A brief explanatory note or consultative letter is also recommended.

Specimen Required

Supplies: Pathology Packaging Kit (T554)

Specimen Type: Formalin-fixed or B5-fixed, paraffin-embedded tissue block

Collection Instructions:

1. Do not send fixed tissue slides. Testing can only be done on paraffin-embedded tissue blocks.

2. Attach the green pathology address label included in the kit to the outside of the transport container.

Forms

If not ordering electronically, complete, print, and send a Renal Diagnostics Test Request (T830) with the specimen.

Reject Due To

Fixed tissue slides wet or frozen tissue Cytological smears Nonformalin fixed tissue Nonparaffin embedded tissue	Reject

Specimen Stability Information

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

Clinical & Interpretive

Clinical Information

Fibrillary glomerulonephritis (FGN) is a rare kidney disease with fibrillary deposits in the glomeruli that contain polyclonal IgG and complement, indicating immune complex deposition. Although usually Congo-red negative, recently cases with weak Congo-red positivity have been observed, making the distinction from amyloid more challenging. Liquid chromatography-tandem mass spectrometry (LC-MS/MS) performed on microdissected glomeruli from patients with FGN demonstrates a unique proteomic profile including the protein DNAJB9 (Mayo Clinic unpublished observations). The presence of DNAJB9 was found to be highly sensitive and specific for FGN, distinguishing it from other glomerular diseases including amyloid, immunotactoid glomerulopathy, and immune complex-mediated proliferative glomerulonephritis. The presence of DNAJB9, in the appropriate clinical and pathological context, can be useful to establish a diagnosis of FGN.

Interpretation

An interpretation will be provided.

Cautions

No significant cautionary statements

Clinical Reference

1. Said SM, Sethi S, Valeri AM, et al: Renal amyloidosis: origin and clinicopathologic correlations of 474 recent cases. Clin J Am Soc Nephrol 2013 Sep;8(9):1515-1523

2. Vrana JA, Gamez JD, Madden BJ, et al: Classification of amyloidosis by laser microdissection and mass spectrometry-based proteomic analysis in clinical biopsy specimens. Blood 2009;114(24):4957-4959

3. Rosenstock JL, Markowitz GS, Valeri AM, et al: Fibrillary and immunotactoid glomerulonephritis: Distinct entities with different clinical and pathologic features. Kidney Int 2003;63:1450-1461

4. Casanova S, Donini U, Zucchelli P, et al: Immunohistochemical distinction between amyloidosis and fibrillar glomerulopathy. Am J Clin Pathol 1992;97:787-795

5. Rosenmann E, Eliakim M: Nephrotic syndrome associated with amyloid-like glomerular deposits. Nephron 1977;18:301-308

6. Nasr SH, Vrana JA, Dasari S, et al: DNAJB9 is a Specific Immunohistochemical Marker for Fibrillary Glomerulonephritis. Kidney Int Rep 2017;3(1):56-64

7. Dasari S, Alexander MP, Vrana JA, et al: DnaJ Heat Shock Protein Family B Member 9 is a Novel Biomarker for Fibrillary GN. J Am Soc Nephrol 2018;(1):51-56

Performance

Method Description
Affected areas are removed from paraffin-embedded tissues by laser microdissection. Protein digestion is performed, followed by liquid chromatography-tandem mass spectrometry.(Unpublished Mayo method)

PDF Report
No

Specimen Retention Time
Until Reported

Performing Laboratory Location
Rochester

Fees & Codes

Test Classification
This test was developed, and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information
82542

88380

LOINC® Information

Test Definition: MSFGN

Fibrillary GN Confirm, LC MS/MS

Test ID	Test Order Name	Order LOINC Value
MSFGN	Fibrillary GN Confirm, LC MS/MS	In Process

Result ID	Reporting Name	LOINC®
BA0389	Interpretation	59465-5
BA0390	Participated in the Interpretation	No LOINC Needed
BA0391	Report electronically signed by	19139-5
BA0392	Material Received	81178-6
BA0393	Disclaimer	62364-5
BA0394	Case Number	80398-1