MAYO CLINIC LABORATORIES

Test Definition: MSFGN

Fibrillary Glomerulonephritis Confirmation, Mass Spectrometry, Paraffin Tissue

Overview

Useful For
Diagnosing fibrillary glomerulonephritis

Testing Algorithm
A pathology consultation is typically not required. If the results of this test do not support the clinical findings, 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 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 1 of the following forms with the specimen:
-Kidney Transplant Test Request
-Renal Diagnostics Test Request (T830)

Reject Due To

Fixed tissue slides wet or frozen	Reject

tissue Cytological smears Nonformalin fixed tissue Nonparaffin embedded tissue	

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glomeruli from patients with FGN demonstrates a unique proteomic profile including the protein DNAJB9 (Mayo Clinic unpublished observations).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;8(9):1515-1523

2. Vrana JA, Gamez JD, Madden BJ, Theis JD, Bergen HR 3rd, Dogan A. 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, Sacchi G, Appel GB, D'Agati VD. Fibrillary and immunotactoid glomerulonephritis: Distinct entities with different clinical and pathologic features. Kidney Int. 2003;63(4):1450-1461

4. Casanova S, Donini U, Zucchelli P, Mazzucco G, Monga G, Linke RP. Immunohistochemical distinction between

amyloidosis and fibrillar glomerulopathy. Am J Clin Pathol. 1992;97(6):787-795
5. Rosenmann E, Eliakim M. Nephrotic syndrome associated with amyloid-like glomerular deposits. Nephron. 1977;18(5):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;29(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

Day(s) Performed
Monday through Friday

Report Available
7 to 15 days

Specimen Retention Time
Until reported

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

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82542
88380

LOINC® Information

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

Result ID	Test Result Name	Result LOINC® Value
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