

Fibrillary Glomerulonephritis: Clinicopathologic Features and Outcomes

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Abstract

Background:

Fibrillary glomerulonephritis (FGN) is a rare immune-mediated glomerular disease characterized by glomerular fibrillar deposits and historically associated with poor kidney outcomes. Despite recent diagnostic advances, including *DNAJB9* immunohistochemistry, the clinical spectrum, prognostic factors, and optimal management of FGN remain incompletely defined.

Methods:

We conducted a retrospective cohort study of patients with biopsy-proven FGN identified through the Cleveland Clinic Kidney Biopsy Epidemiology Project between January 2015 and March 2023.

Results:

Thirty patients met inclusion criteria. The cohort was predominantly female (70%) and white (86%), with a median age of 64 years. At the time of kidney biopsy, all patients had hypertension and acute or chronic kidney disease. Additional features included hematuria (76%), proteinuria (60%), and nephrotic syndrome (48%). Monoclonal protein was detected in 20% of cases. During a mean follow-up of 24 months (range 0–70 months; n = 13), 23% achieved partial remission, 23% had persistent disease, and 53% progressed to end-stage kidney disease (ESKD). Higher serum creatinine at biopsy was the only variable associated with progression to ESKD.

Conclusion:

FGN remains a rare glomerular disease with substantial risk of progression to kidney failure. This single-center cohort highlights the heterogeneous clinical presentation and generally poor kidney outcomes of FGN in the modern diagnostic era. Larger, multicenter studies are needed to refine prognostic stratification and guide evidence-based management strategies.

Introduction

Fibrillary glomerulonephritis (FGN) is a rare glomerular disease characterized by randomly arranged, nonbranching fibrils measuring approximately 15–25 nm in diameter on electron microscopy.^{1,2} On immunofluorescence, the deposits typically stain for IgG, C3, and both κ and λ light chains, supporting an immune complex-mediated process.¹ FGN accounts for roughly 0.5–1% of native kidney biopsies, yet it carries a disproportionately high risk of progressive kidney failure, with 36–44% reaching end-stage kidney disease (ESKD) within a few years of diagnosis.^{3–5}

Historically, diagnosis relied solely on electron microscopy, where the diameter of the fibrils could help differentiate it from immunotactoid glomerulopathy.¹ The identification of *DNAJB9* as an immunohistochemical marker for FGN enabled reliable identification of cases across large biopsy cohorts.⁶ *DNAJB9* staining also revealed that FGN is far more homogeneous than previously appreciated, with a consistent molecular profile and strong correlation with classic ultrastructural findings.^{3,7}

FGN was initially considered an idiopathic condition; however, subsequent studies have demonstrated associations with autoimmune diseases, malignant neoplasms, and chronic infections such as hepatitis C.⁷ Clinically, fibrillary glomerulonephritis most commonly presents with proteinuria, often in the nephrotic range, hematuria, impaired kidney function, and hypertension. Despite early recognition with biopsies and advances in supportive therapies, including RAAS blockade and immunosuppression, outcomes remain poor.^{6,7} Nearly 50% of patients progress to end-stage kidney disease within four years of diagnosis.^{7–9}

While multicenter cohorts have expanded our understanding of FGN, data from large integrated health systems remain limited, and institution-specific patterns of presentation, pathology, and outcomes are not well characterized. This is particularly relevant in high-volume biopsy centers, where systematic evaluation may provide additional insight into disease heterogeneity and real-world management.^{3–5,8–10} In this study, we describe the clinical, pathological, and outcome characteristics of 30 biopsy-proven FGN cases evaluated within the Cleveland Clinic Health System, providing contemporary tertiary center data to further inform prognosis and clinical decision-making.

Methods

Forty-five (45) patients within the Cleveland Clinic health system with a diagnosis of FGN were identified by retrospective review of all native kidney biopsies evaluated in the Cleveland Clinic Kidney Biopsy Epidemiology Project from January 2015 to March 2023.¹¹ Fifteen patients were excluded from this study because of insufficient clinical data and/or incomplete biopsy information (Fig. 1). The remaining 30 patients that were included in this study had biopsy proven FGN.

All patients had their kidney biopsies performed and interpreted at our institution. Standard processing of the biopsies included light microscopy (LM), immunofluorescence (IF), and electron microscopy (EM). For LM, all cases were stained with hematoxylin and eosin, periodic acid-Schiff, trichrome, and Jones methenamine silver. Standard IF was done for IgG, IgM, IgA, C3, C1q, kappa, and lambda. Pronase digestion was used when necessary to uncover masked immunoglobulins. Staining for *DNAJB9* was done when available. Interstitial fibrosis and tubular atrophy were graded by their severity as absent (< 10%), mild (10–25%), moderate (26–50%), or severe (> 50%). Partial remission was defined as a > 50% reduction in proteinuria.

Statistical Analysis

Baseline demographic, clinical, laboratory, and pathologic characteristics were summarized using descriptive statistics. Continuous variables were reported as mean ± standard deviation or median

(interquartile range), as appropriate based on data distribution, while categorical variables were expressed as counts and percentages. Comparisons between groups were performed using the Student *t*-test for continuous variables and Fisher's exact test for categorical variables. Kidney outcomes, including progression to ESKD, were assessed descriptively given the limited sample size. All statistical tests were two-sided, and a *p*-value < 0.05 was considered statistically significant. Analyses were conducted using SPSS version 28.0.11 (IBM Corp., Armonk, NY).

Results

Clinical features

Among 4,126 native kidney biopsies performed during the study period, FGN was identified in 45 cases, representing 1.1% of all biopsies. Fifteen patients were excluded because of incomplete data, leaving 30 patients for the final analysis (Fig. 1, Fig. 2). The cohort was predominantly female (70%) and white (86.7%), with a mean age at biopsy of 65 ± 11 years. Hypertension was present in all patients, while diabetes mellitus was observed in 60%. Additional comorbidities included heart failure (33.3%), solid organ malignancy (10%), and liver cirrhosis (3.3%). More than half of patients (56.6%) were receiving renin-angiotensin-aldosterone system (RAAS) inhibitor therapy at the time of biopsy (Table 1).

Table 1

Demographic and Clinical Characteristics of 30 Patients with Biopsy-Proven Fibrillary Glomerulonephritis

Variable	Mean (SD) or No. (%) of patients
Demographics	
Age at Biopsy (years)	65 (11)
Sex (Female)	21 (70)
Race (White)	26 (86.7)
Tobacco use (yes)	19 (63.3)
Comorbidities	
Hypertension (yes)	30 (100)
Diabetes (yes)	18 (60)
Heart Failure (yes)	10 (33.3)
Liver Cirrhosis (yes)	1 (3.3)
Solid organ malignancy (yes)	3 (10)
Hepatitis C positive by PCR	4 (16)
Dialysis	
Need for dialysis before kidney biopsy	1 (3.3)
Need for dialysis after kidney biopsy	11 (36.7)
Time between biopsy and dialysis (months)	9 (20)
Proteinuria	
Mean 24-hour urine protein (g/day)	3.6 (2.8)
Serum Albumin at time of biopsy (g/dL)	3.3 (0.6)
Serum Creatinine at time of biopsy (mg/dL)	2.7 (2.0)
eGFR at time of biopsy (mL/min)	26.0 (16.0)
Hematuria	
Hematuria	22 (75.9)
Gross Hematuria	2 (9.1)
Normal Complement levels	21 (95.5)
eGFR: estimated glomerular filtration rate, MGUS: monoclonal gammopathy of undetermined significance, PCR: polymerase chain reaction, UPCR: Urine protein creatinine ratio	

Variable	Mean (SD) or No. (%) of patients
Demographics	
Dysproteinemia	
Underlying dysproteinemia	5 (16.7)
MGUS	4 (80)
Multiple Myeloma	1 (20)
eGFR: estimated glomerular filtration rate, MGUS: monoclonal gammopathy of undetermined significance, PCR: polymerase chain reaction, UPCR: Urine protein creatinine ratio	

At the time of kidney biopsy, all patients had evidence of acute or chronic kidney disease, with a mean serum creatinine of 2.7 ± 2.0 mg/dL and a mean estimated glomerular filtration rate (eGFR) of 26.0 ± 16.0 mL/min/1.73 m². Proteinuria was present in 60% of patients, with a mean 24-hour urine protein-to-creatinine ratio of 3.6 ± 2.8 g/g. Hypoalbuminemia was common, with a mean serum albumin of 3.3 ± 0.6 g/dL, and nearly half of the cohort (48%) met criteria for nephrotic syndrome (Table 1).

Hematuria was observed in 76% of patients, although gross hematuria was uncommon (9.1%). Complement levels were normal in the majority of patients, with low C3 levels identified in only one case. Underlying dysproteinemia was identified in 16.7% of patients, most commonly monoclonal gammopathy of undetermined significance (MGUS). Hepatitis C virus infection was present in 16% of those tested. One patient (3.3%) required dialysis prior to kidney biopsy, whereas 11 patients (36.7%) initiated dialysis after biopsy, with a median time of 9 months between biopsy and dialysis initiation (Table 1).

Pathologic characteristics

On light microscopy, the predominant glomerular pattern was mesangial proliferation, observed in 93% of cases, while endocapillary hypercellularity was seen in only 7%, and crescents were identified in 10% (1 fibrous, 1 cellular and 1 unspecified). Arteriosclerosis and arteriolar hyalinosis were present in 67% of patients. Interstitial fibrosis and tubular atrophy were moderate to severe in 76% of cases.

Immunofluorescence studies demonstrated IgG deposition (mean intensity 2.5+), with frequent co-deposition of C3 (93%) and C1q (48%). IgM and IgA staining were less common (each 13%). Both kappa and lambda light chains stained positively in 96% of patients, consistent with polyclonal immune complex deposition. IgG staining characteristically exhibited a smudgy appearance without clear granular or linear patterns.

Electron microscopy revealed randomly arranged fibrillary deposits, most commonly located in the mesangium (86%) and along the glomerular basement membrane (82%). Subepithelial deposits were rare, identified in only one patient. DNAJB9 immunostaining was performed in two cases and was positive in

both, confirming the diagnosis of FGN, reflecting the fact that *DNAJB9* testing was introduced during the later years of the study (Table 2).

Table 2
Pathologic characteristics of our cohort with 30 patients with biopsy proven fibrillary glomerulonephritis

Variable	N	Mean (SD) or No. (%) of patients
Light microscopy	30	
Proliferation pattern		
Endocapillary proliferation		2 (6.7)
Mesangial proliferation		28 (93.3)
Presence of Crescents		3 (10)
Arteriosclerosis and Arteriolar Hyalinosis		
Mild		5 (16.7)
Moderate		9 (30)
Severe		6 (20)
Interstitial Fibrosis and Tubular Atrophy		
Mild		7 (23.3)
Moderate		12 (40)
Severe		11 (36.7)
Immunofluorescence	29	
<i>DNAJB9</i> Positive		2 out of 2 tested
IgG (Mean Intensity 2.5+)		29 (100)
IgM		4 (13.8)
IgA		4 (13.8)
C3 (Mean Intensity 2+)		27 (93.1)
C1q		14 (48.3)
Kappa		28 (96.6)
Lambda		29 (100)
Electron microscopy	22	
Glomerular Deposits		
Mesangial		19 (86.4)
Subepithelial		1 (4.5)
GBM: glomerular basement membrane		

Variable	N	Mean (SD) or No. (%) of patients
Mixed		2 (9.1)
GBM Thickening		18 (81.8)
GBM: glomerular basement membrane		

Clinical Outcomes

Seven patients (23%) received immunosuppressive therapy, including five treated with rituximab. Among rituximab-treated patients, three achieved partial remission, one had persistent disease, and one progressed to ESKD. Follow-up data were available for 13 patients, with a mean follow-up duration of 24 months (range, 0–70 months) (Table 3). During follow-up, three patients (23%) achieved partial remission, three (23%) had persistent disease, and seven (53%) progressed to ESKD.

Table 3
Treatment characteristics and renal outcomes in patients with FGN during follow-up

Patient	First line chemotherapy after FGN diagnosis	Treatment outcome	Time to renal outcome from kidney biopsy in months	Presence of a clone
1	-	ESKD	5	-
2	Rituximab	Partial Remission	18	-
3	-	ESKD	1	-
4	Rituximab, cyclophosphamide	Persistent disease	36	MGUS
5	Cytosan/Prednisone	ESKD	28	-
6	-	Persistent disease	12	-
7	Rituximab	ESKD	70	-
8	Bortezomib	Persistent disease	48	MM
9	Rituximab	Partial Remission	5	-
10	-	ESKD	1	-
11	Rituximab	Partial Remission	6	-
12	-	ESKD	3	-
13	-	ESKD	1	-
ESKD: end-stage kidney disease, FGN: Fibrillary glomerulonephritis, MGUS: monoclonal gammopathy of unknown significance, MM: multiple Myeloma				

Exploratory analyses using Fisher's exact test and univariate comparisons were performed to identify predictors of progression. Given the limited sample size, no statistically significant associations were identified between clinical or pathological variables and kidney outcomes, with the exception of higher serum creatinine at the time of kidney biopsy, which was associated with progression to ESKD. Multivariable analysis was not performed due to limited event numbers.

Discussion

Clinical Presentation

FGN is a rare kidney disorder characterized by the deposition fibrils in the glomerular mesangium and along the glomerular basement membrane.^{12–15} This study provides a detailed description of biopsy-

proven FGN within a large integrated healthcare system. In our cohort, patients most commonly presented with proteinuria, microscopic hematuria, and impaired kidney function, findings that are consistent with prior reports..^{8,16} Similar to earlier studies, disease presentation was heterogeneous, ranging from urinary abnormalities with preserved kidney function to rapid progression to ESKD.^{17,18}

Compared with previously published cohorts from Mayo Clinic, Columbia University, and the University of North Carolina (UNC), our patients exhibited a higher prevalence of diabetes mellitus (60%) and autoimmune disease (23%), alongside a lower overall degree of proteinuria (mean 3.6 g/day) (Table 4).^{4,5,19,20} Differences in proteinuria assessment methods, predominantly urine protein-to-creatinine ratio in our cohort versus 24-hour urine collections in others, may partially account for these findings. Nonetheless, nephrotic-range proteinuria remained common regardless of measurement approaches.

Table 4
Clinical characteristics of patients with fibrillary glomerulonephritis in the five largest cohorts

Variable	Multi-institutional Cohort ²⁰ (n = 266)	Columbia ¹⁹ (n = 61)	Mayo Clinic ⁴ (n = 64)	UNC ⁵ (n = 42)	Cleveland Clinic (n = 30)
	Mean (SD) or No. (%) of patients	Mean (SD) or No. (%) of patients	Mean (SD) or No. (%) of patients	Mean (SD) or No. (%) of patients	Mean (SD) or No. (%) of patients
Age (years)	61	57	59	54	65
Race (White)	NA	92	NA	71	86
Sex (Female)	65	61	74	60	70
Creatinine (mg/dL)	2.1	3.1	2.5	3.2	2.7
Proteinuria (g/d)	NA	6.4	5.1	5.7	3.6
Nephrotic Syndrome	15	52	25	NA	43
Hematuria	39	60	90	97	75
Hepatitis C	16	17	7	27	13
Autoimmune disease	9	5	14	13	23
Diabetes Mellitus	22	20	24	28	60
Malignancy	7	7	10	12	10
Dysproteinemia	8	15	4	42	16
Characteristic pathologic findings in fibrillary glomerulonephritis. Glomeruli become infiltrated by fibrillary deposits and can display a wide range of patterns by light microscopy. Panel A (Jones methenamine silver stain, 400x) shows mesangial hypercellularity as well as replacement of the typically black mesangial matrix and glomerular basement membranes with a grey-pink color due to the diffuse infiltration in this glomerulus. Immunofluorescence staining typically shows smudgy appearing deposits within glomeruli that stain for IgG (pictured in panel B, 400x), C3, kappa and lambda. Electron microscopy (panel C at 30,000x magnification) shows deposits composed of fibrils (typically in the 15–25 nm range) often admixed with more granular electron dense material. Immunohistochemical staining with DNAJB9 can confirm the diagnosis (panel D, 200x).					

Demographically, our cohort was predominantly white (86%) with a female predominance (70%), consistent with earlier reports.^{21,22} However, Black patients represented 13% of our cohort, which is notably lower than the estimated local Black population in our cohort.²³ Prior work, particularly from UNC, has demonstrated a strong association between HCV infection and FGN among Black patients.⁵ Although

HCV infection was present in 13% of our cohort, we did not observe race-specific associations, likely reflecting limited sample size and regional referral patterns. In addition, FGN has been linked to paraproteinemia and monoclonal gammopathies in a subset of patients, suggesting that underlying clonal or immune dysregulation may contribute to disease pathogenesis across diverse populations. This observation has important clinical implications, as the identification of a circulating monoclonal protein may indicate an underlying plasma cell or B-cell clone and may influence both diagnostic evaluation and therapeutic strategies. In such cases, targeted therapies directed at the pathogenic clone may represent a potential treatment approach rather than conventional immunosuppression alone. In our cohort, 20% of patients had evidence of a detectable clone, supporting the concept that clonal immune processes may play a role in a subset of FGN cases.

Histopathologic Features

Histopathologic evaluation in our cohort confirmed the characteristic features of FGN, including randomly arranged non-amyloid fibrils predominantly involving the mesangium and glomerular basement membrane.^{12–15} Light microscopy most commonly demonstrated mesangial proliferation, while a substantial proportion of patients (76.5%) exhibited moderate to severe interstitial fibrosis and tubular atrophy, findings that likely contribute to poor kidney outcomes.

Immunofluorescence consistently revealed strong IgG and C3 staining with polyclonal kappa and lambda light chain expression, supporting the immune complex-mediated nature of FGN. The “smudgy” appearance of IgG staining observed in most cases further aligns with classic descriptions of the disease. Electron microscopy confirmed fibrillar deposits in the majority of cases, most commonly within the mesangium, with limited subepithelial involvement.^{8,9,21,24}

Importantly, immunohistochemical staining for *DNAJB9* was positive in both patients tested, supporting its growing role as a reliable biomarker for FGN. Although *DNAJB9* testing was not universally available throughout the study period, its incorporation into diagnostic workflows may reduce diagnostic uncertainty in future cohorts.^{3,10,25,26}

Clinical Outcomes and Management

Clinical outcomes in our cohort reflect the generally poor prognosis traditionally associated with FGN. Over a mean follow-up of 24 months, more than half of patients with available outcome data progressed to ESKD, highlighting the progressive nature of the disease. Higher serum creatinine at the time of biopsy was the only variable associated with progression to ESKD, consistent with prior studies identifying baseline kidney dysfunction as a key prognostic factor. In addition, the degree of chronic damage at the time of biopsy, including interstitial fibrosis and tubular atrophy, has been shown in prior cohorts to correlate with worse kidney outcomes and likely contributes to the progressive course observed in many patients.

Therapeutic strategies for FGN remain poorly defined. The polyclonal immune complex deposition observed in FGN has supported the use of immunosuppressive therapies, particularly B-cell-directed

agents.^{4,20,27–29} In our cohort, rituximab was the most commonly used immunosuppressive therapy, with partial remission achieved in a subset of treated patients. Although outcomes were variable, these findings align with prior reports suggesting potential benefit of rituximab in select patients.

A subset of patients with FGN have detectable monoclonal gammopathies, raising the possibility that clonal immune dysregulation contributes to disease pathogenesis. Further studies are needed to clarify the role of clonal processes and targeted therapies in FGN.

Limitations of our study include the retrospective design, modest sample size inherent to this rare disease, and incomplete longitudinal data for a subset of patients. Additionally, *DNAJB9* testing was not uniformly available throughout the study period, limiting its application across the entire cohort. Future studies where *DNAJB9* staining is universally applied may provide additional disease insights. Despite these constraints, the consistency of our findings with prior cohorts reinforces their validity.

Conclusion

In summary, this tertiary center cohort highlights the characteristic clinical and pathologic features of FGN, which most commonly presents with proteinuria, hematuria, and impaired kidney function and is frequently associated with progressive kidney function decline. Given the heterogeneity in presentation, treatment response, and outcomes, larger collaborative studies are needed to better define prognostic factors and guide evidence-based management strategies for this rare glomerular disease.

Human Ethics and Consent to Participate Declaration

IRB approval was obtained with waiver of informed consent and HIPPA authorization for retrospective data collection (IRB # 22-1093)

Declarations

Consent to Publish Declaration:

Not applicable

Competing Interest Declaration:

SB has research grants from Otsuka Pharmaceuticals. LH reports having consultancy with ChemoCentryx and reports having an advisory or leadership role at ASN Kidney 360 editorial board

Funding:

There was no funding for this project

Author Contribution

HS and SB conceptualized the project OO and HS did chart review and data collection OO and AB wrote the main manuscript texts and subsequent edits TG completed data analysis HS LH AM SC SG and SB assisted with editing and conceptualization All authors reviewed the manuscript

Data Availability

Retrospective data collection on patients done from EMR to RedCap. Data can be made available following IRB request

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Figures

Table 3. Clinical Characteristics	
	Mean Value, std. deviation (Values obtained)
Mean 24-hour urine protein (UPCR)	3.6g/g +/- 2.8 (17/30)
Serum Albumin at time of biopsy	3.3g/dL +/- 0.6 (28/30)
Serum Creatinine at time of biopsy	2.7mg/dL +/- 2 (28/30)
eGFR at time of biopsy	26 mL/min +/- 16 (28/30)
Need for dialysis before kidney biopsy	1/30
Need for dialysis after kidney biopsy	11/30
Time (months) between biopsy and dialysis	9 +/- 20
Hematuria	22/29
Gross Hematuria	2/22
Normal Complement levels	21/22
Hepatitis C positive by PCR	4/25
Underlying dysproteinemia	5/30
MGUS	4/5
Multiple Myeloma	5/5

Figure 1

Study flow diagram of patients with biopsy-proven fibrillary glomerulonephritis

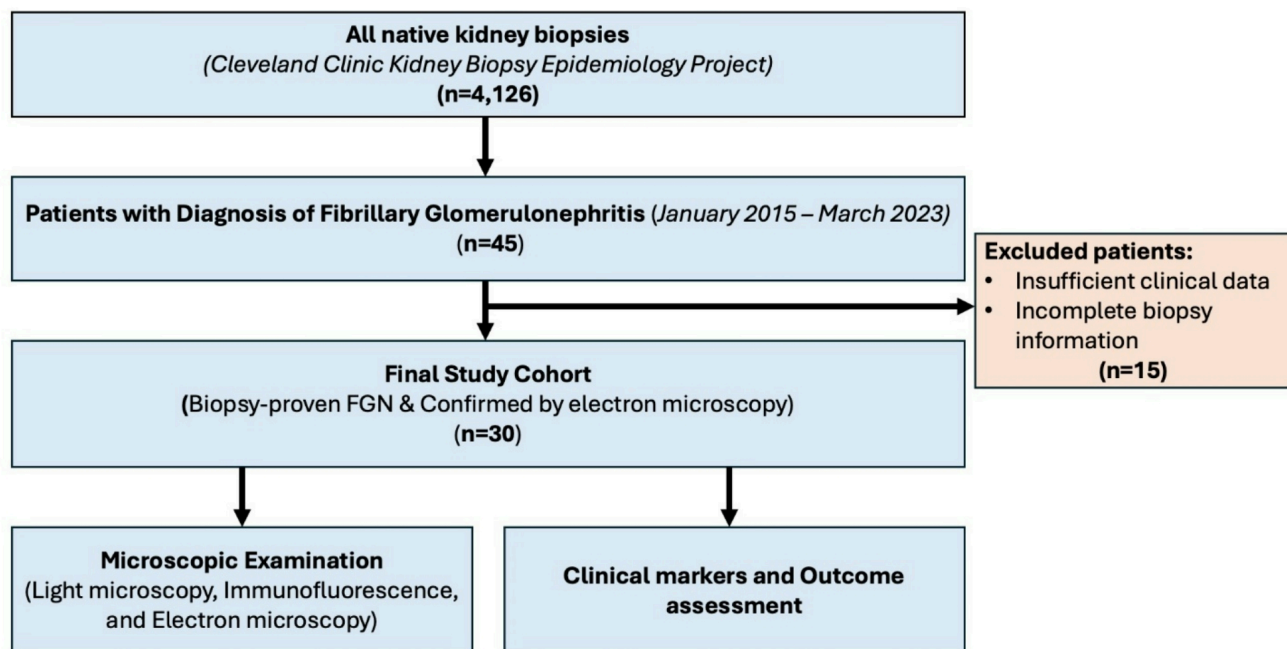


Figure 2

Kidney histopathologic features of patients with fibrillary glomerulonephritis.