following transplant. Additionally, this case highlights the need to determine the optimal treatment of recurrent MN in kidney transplant recipients, who are already on standard immunosuppressive regimens, to maintain kidney function and maximize long-term graft outcomes.

G-476. MINIMAL CHANGE DISEASE AND THYMOMA: A 14-YEAR STRUGGLE AND THE ROLE OF STEROID-SPARING AGENTS:

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Minimal Change Disease (MCD) is a podocytopathy commonly seen in children but accounts for up to 25% of adult nephrotic syndrome cases. While most cases are idiopathic, rare secondary associations with malignancies such as thymoma are documented. This report explores the 14-year longitudinal course of a patient with MCD and thymoma, emphasizing the challenges of managing frequent relapses and steroid dependency, and the potential of steroid-sparing agents.

A 30-year-old African American female presented with nephrotic syndrome characterized by severe edema, hypoalbuminemia, and nephrotic-range proteinuria. Renal biopsy confirmed MCD. Concurrently, an anterior mediastinal mass was diagnosed as WHO Grade AB thymoma, necessitating thymectomy. The patient's MCD course was marked by multiple relapses requiring high-dose corticosteroids, transitioning to steroid-sparing agents including azathioprine, cyclosporine, and tacrolimus, with limited success. Rituximab was eventually initiated in 2024, providing sustained remission. Periodic re-biopsies demonstrated persistent podocyte foot process effacement without evidence of progression to focal segmental glomerulosclerosis (FSGS).

The interplay between MCD and thymoma suggests a paraneoplastic mechanism involving T-cell dysregulation, underscoring the need for vigilant malignancy screening in adult-onset MCD. While corticosteroids remain the mainstay of therapy, prolonged use poses risks, highlighting the importance of exploring steroid-sparing alternatives. Recent evidence supports rituximab for steroid-dependent or frequently relapsing cases, as seen in this patient. However, the long-term efficacy and safety of B-cell depleting therapies warrant further study

This case highlights the complexities of managing secondary MCD in the setting of thymoma. Emerging therapies like rituximab may redefine the treatment paradigm, offering hope for achieving durable remission in refractory cases. Collaborative research is essential to optimize care for such challenging presentations.

G-477. LIVING WITH IGA NEPHROPATHY: A FOCUS GROUP STUDY ON THE PATIENT EXPERIENCE:

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In the United States, IgA nephropathy (IgAN), the most common form of glomerulonephritis, is a rare autoimmune disease that affects ~200,000 individuals. With IgAN, IgA antibodies accumulate and clump inside the glomeruli, causing inflammation that leads to chronic kidney disease (CKD) and often to kidney failure. There is no cure for IgAN, however treatments can help to delay progression to kidney failure. To help mitigate the burden of disease on individuals with IgAN, the American Kidney Fund (AKF) sought to gain a personalized understanding of the IgAN patient experience to inform the development of resources that empower patients to effectively manage their condition.

In October 2024, AKF conducted a virtual focus group with three individuals living with IgAN. Participants in the focus group are members of AKF's Rare Kidney Disease Action Network and were recruited via email applying convenience sampling varying in age, gender, race, education, and stage of CKD. Focus group responses were recorded, transcribed, and thematically analyzed.

Findings revealed three distinct themes and two thematic questions among participants: 1) IgAN was well understood, participants could define the condition and how it affects the kidneys. 2) Dietitians and nutrition plans impact how patients manage IgAN. 3) Support from caregivers, family, and support groups helped reduce emotional and care-related burdens. Participants also expressed questions about treating the disease (not just symptoms) and if IgAN can reoccur post-transplant.

While participants understood and could explain IgAN, they expressed uncertainty about treatment mechanisms and if IgAN affects a transplanted kidney. Although participants reported having found helpful resources for support in disease management, particularly related to diet, voids in patient-centered information about most recent treatments and resources that explain the trajectory of IgAN post-transplant remain. Leveraging these findings, AKF will develop more targeted materials for IgAN patients, emphasizing treatment options and post-transplant life. Future research should investigate the optimal dissemination strategies for these materials and assess their impact on patient understanding and engagement.

G-478. DNAJB9 FIBRILLARY GLOMERULONEPHRITIS IN A PATIENT WITH SLE:

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Fibrillary glomerulonephritis (FGN) is a rare glomerular disease characterized by deposits of randomly oriented fibrils composed of DNAJB9, a heat shock protein. Most cases are idiopathic, though associations with monoclonal gammopathy, autoimmune diseases, and infections have been reported. We present a patient with systemic lupus erythematosus (SLE) and nephrotic-range proteinuria diagnosed with FGN.

A 42-year-old female with SLE, hypertension, and type 2 diabetes presented with microscopic hematuria and proteinuria. In the past year, she was treated with azathioprine and hydroxychloroquine for extrarenal lupus symptoms, including malar rash and arthralgias. Workup revealed normal serum creatinine (0.8 mg/dL), hypoalbuminemia (3.2 g/dL), and urine total protein of 5.7 g/g. ANA pattern was homogeneous with a titer of 1:2560. Anti-dsDNA was 129 IU/mL, anti-RNP, SS-A, anti-chromatin, and anti-citrulline antibodies were all strongly positive, and antiphospholipid antibodies were present in high titer. C3 and C4 were normal. Serum immunofixation was negative, free light chain ratio was normal, hepatitis and HIV serologies were negative. Kidney ultrasound was unremarkable. A kidney biopsy was performed.

Light microscopy showed mesangial expansion, hypercellularity and segmental basement membrane thickening. Immunofluorescence revealed smudgy mesangial and capillary wall staining, prominent for IgG, kappa and lambda light chains, and less intense staining for C1q, C3, IgA, and IgM. Electron microscopy revealed diffuse podocyte effacement and randomly oriented fibrils 15-20 nm in thickness. Congo Red staining was negative. Immunohistochemical staining for DNAJB9 was strongly positive in glomeruli, confirming FGN.

The patient's immunosuppression was switched to mycophenolate mofetil (MMF) 2 grams daily and prednisone with continuation of hydroxychloroquine. After 1 year, proteinuria decreased to 2 g/g, and serologic parameters improved.

FGN does not fit within the traditional classification schema for lupus nephritis (LN). Currently, there is no standard management for FGN. It is recommended to screen for associations and treat the cause in secondary cases. In our case, treatment for proliferative LN with MMF and steroids led to great improvements in laboratory and clinical parameters.

G-479. A MECHANISTIC BIOPSY STUDY TO EVALUATE STRUCTURAL AND FUNCTIONAL CHANGES IN KIDNEYS OF PATIENTS WITH IGA NEPHROPATHY (IGAN) RECEIVING IPTACOPAN ON TOP OF SUPPORTIVE CARE:

Dana V Rizk[1], Bart Maes[2], Hong Zhang[3], Clint W Abner[4], Marie-Anne Valentin[5], Vipin N[6], Maria Fernanda Di Tata[7], Jonathan Barratt[8]. [1]The University of Alabama at Birmingham; [2]Delta General Hospital; [3]Peking University First Hospital; [4]Novartis Pharmaceuticals; [5]Novartis Pharma AG; [6]Novartis Healthcare Ltd; [7]Novartis Farmacéutica SA; [8]University of Leicester & Leicester General Hospital

Overactivation of the alternative complement pathway (AP) is a key driver of IgAN, leading to kidney inflammation and injury. This mechanistic study aims to evaluate the effects of iptacopan, a selective complement factor B inhibitor, on the underlying immunopathology in patients with IgAN.

This multicenter, single-arm, open-label, repeat-biopsy study will enroll up to 20 adult patients with biopsy-proven IgAN, eGFR ≥30 mL/min/1.73m^2, proteinuria ≥0.8 g/g or 1 g/day, and receiving a maximally tolerated and/or stable dose of supportive care treatment for ≥90 days before baseline. Patients will receive iptacopan 200 mg bid for 9 months, with kidney biopsies at baseline and study end (Figure). The primary objective is to quantify changes in mesangial complement C3c-containing fragment deposition, an indicator of AP activation, from baseline to 9 months, assessed by immunofluorescence staining. Other objectives will evaluate changes in biomarkers related to the AP, kidney function and structure, and disease progression from baseline to 9 months.

This study will explore iptacopan's impact on IgAN immunopathology by assessing glomerular complement activation together with renal histopathology, kidney function, and key biomarkers. The findings will enhance understanding of iptacopan's mechanistic effects on IgAN and potential kidney-protective benefits.

bid, twice a day; D, day; EOS, end of study; n, number of participants.
[1]Eligible participants may enroll in the roll-over extension study, contingent upon local regulations.

G-480. LUPUS VASCULOPATHY: A CALL TO ACTION FOR ENHANCED AWARENESS AND RESEARCH:

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Lupus vasculopathy (LV) is a rare form of vascular involvement in systemic lupus erythematosus (SLE), defined by necrosis and accumulation of immunoglobulins and complements in the walls of arterioles and small arteries leading to luminal narrowing. Here we present a case of a young female with SLE admitted with immunologically active disease and found to have LV.

18-year-old African American female with known SLE class IV (Biopsy proven 2021) medication nonadherent for one year, who presented with nausea and body aches. She was found to have worsening kidney function (Cr 7.7 mg/dL). On admission she had elevated dsDNA, low complement levels, and SLE disease activity index of 24. She was found to have pancytopenia including hemolytic anemia with multiple schistocytes in peripheral smear. She was treated with pulse dose steroids and received urgent plasmapheresis. ADAMTS13 and anti-phospholipid antibodies were negative. Renal biopsy was consistent with diffuse proliferative lupus class IV with focal necrotizing arteritis, RBC fragmentation and mucoid intimal edema consistent with TMA; interstitial fibrosis and tubular atrophy (25-30%); and moderate arteriosclerosis. Patient remained dialysis-dependent on discharge pending genetic testing. Discharge regimen included oral steroids, belimumab, hydroxychloroquine, and mycophenolate.

This case emphasizes several different renal manifestations of LV. Other potential diagnoses include thrombotic thrombocytopenic purpura, antiphospholipid syndrome-related TMA, atypical hemolytic uremic syndrome, and complement mediated TMA. Renal vascular complications of lupus include arterial sclerosis, thrombotic microangiopathy, and deposition of immune complexes.

It is important to consider the different forms of LV as it entails a poor prognosis, but there are no established guidelines for management. Previous case studies have shown possible effectiveness of plasma exchange with IV cyclophosphamide +/- eculizumab. Antiphospholipid antibodies might be involved in the pathogenesis of lupus vasculopathy. It has been suggested that direct complement activation due to SLE itself is responsible for LV and TMA in a majority of cases. More research is needed to understand this disease entity and define better therapeutic management pathways.

G-481. EXPLORING THE RISK OF THROMBOTIC MICROANGIOPATHY (TMA) IN ANTI-VEGF THERAPY: A GLOBAL MULTICENTRIC RETROSPECTIVE ANALYSIS:

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Anti-VEGF therapies have revolutionized cancer treatment by inhibiting angiogenesis, yet are associated with significant risks, including thrombotic microangiopathy (TMA). TMA is characterized by endothelial injury, microvascular thrombosis, and renal dysfunction, potentially leading to acute renal failure, dialysis dependence, and increased mortality. Understanding the impact of TMA in patients receiving anti-VEGF therapy is crucial for optimizing treatment outcomes.

We aim to assess the risk of TMA in patients treated with anti-VEGF therapy, focusing on dialysis dependence and survival outcomes. A retrospective analysis was conducted using the TriNetX database. Among patients treated with anti-VEGF agents, 228 who developed TMA were compared to those who did not. Primary outcomes included the incidence of dialysis dependence, overall survival, and post-dialysis survival. Statistical analyses included relative risk (RR) and hazard ratios (HR) with 95% confidence intervals (CI).

TMA Incidence**:** 7% of anti-VEGF-treated patients developed TMA, compared to 36.1% of controls (RR 1.321; 95% CI 1.151–1.516). Dialysis Dependence**:** 6% of TMA patients required dialysis, compared to 5% of controls (RR 5.08; 95% CI 4.081–6.33). Survival Probability**:** 9% of patients with VEGF-induced TMA survived compared to 20.3% in controls (HR 1.165; 95% CI 0.963–1.41, p = 0.42). Post-Dialysis Survival: 3% of TMA patients who required dialysis survived, compared to 86.5% of controls (HR 5.3; 95% CI 4.1–6.8, p = 0.601).

Anti-VEGF therapy is associated with an increased risk of TMA, leading to higher rates of dialysis dependence and poorer survival outcomes, particularly post-dialysis. While survival differences were not statistically significant, these findings underscore the serious renal complications associated with anti-VEGF therapy. Early detection of TMA and proactive renal monitoring are essential to mitigate these risks. Further research is needed to develop effective preventive and therapeutic strategies for managing TMA in this high-risk patient population.

G-482. MEMBRANOUS NEPHROPATHY AS AN EXTRAINTESTINAL MANIFESTATION OF ULCERATIVE COLITIS:

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Glomerulonephritis (GN) is a rare complication of Inflammatory bowel disease (IBD). Only a few cases of Membranous Nephropathy (MN) associated with IBD have been reported. We present a patient with Ulcerative colitis (UC) who developed proteinuria due to MN-proven biopsy and responsed to UC treatment.

66-year-old male with a 12-year history of UC, on infliximab infusion every two weeks, presented to hospital with bloody diarrhea and abdominal pain due to a UC exacerbation. His symptoms were associated with frothy urine and limb swelling. Physical examination revealed peripheral edema. Laboratory work-up showed acute kidney injury, creatinine peaked at 3.8 mg/dL and proteinuria of 4.6 g/g. A workup including tests for hepatitis B and C, HIV, ANA, C3, C4, free light chains, and serum protein electrophoresis, was negative. During treatment of the UC flare, proteinuria decreased to 1.6 g/g, and kidney function improved. A kidney biopsy was performed, revealing membranous glomerulonephritis.

Renal complications associated with chronic IBD are commonly due to amyloidosis, cystitis, and nephrolithiasis. The coexistence of IBD and GN is exceptionally rare. A review of the literature reveals only a few reported cases of GN associated with IBD, mostly IgA nephropathy, membranoproliferative glomerulonephritis, and focal segmental glomerulosclerosis. MN associated with UC has been documented in only a handful of cases. The underlying etiology remains unclear. Theories regarding the pathogenesis of renal complications in IBD suggest an immunological response directly linked to intestinal activity. The release of intestinal antigens secondary to bowel inflammation may trigger antibody production and immune complex formation, leading to glomerular injury. Our patient presented with a UC flare-up and was found to have nephrotic-range proteinuria, which improved significantly following treatment of the UC exacerbation without requiring specific