

# Off to a Bad Start

A 10 year-old boy presented with a one-week history of lower extremity edema. While on vacation, he and his parents noted indentations on his ankles from his socks.

His past medical history was significant for environmental allergies with nasal congestion and sneezing. He has asthma with 1-2 episodes of wheezing per year, generally in the fall season, sometimes in the context of a respiratory infection.

Physical examination showed a blood pressure of 116/54 mmHg, clear lungs, mild ascites and 2+ pretibial and pedal edema. No rashes were present.

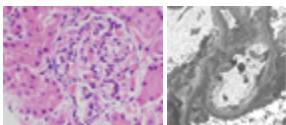
A urine dipstick shows a specific gravity of 1.030, pH 7, with no blood but 4+ protein. The urine protein/creatinine ratio was 6.5 (normal < 0.20, nephrotic range at this age > 2.0). His electrolytes were normal. Other serum studies included: creatinine 0.6 mg/dl, albumin 2.3 g/dl, calcium 8.1 mg/dl, C3 of 110 mg/dl (normal: 80-210), C4 of 22 mg/dl (normal: 15-45). ANA was negative.

1. What is the most likely diagnosis? (Select from either **List 2** or **List 3**).

2-3. Give two (2) reasons that MPGN is unlikely in this case.

The patient was started on prednisone 60 mg once daily. Four weeks later, his urinalysis showed negative protein. He was converted to alternate day prednisone, but 5 days later, he again had 4+ urine protein. A decision was made at that time to perform kidney biopsy.

## Pathology Report for Biopsy #1



Slide 1

Slide 2

The kidney biopsy shows mild mesangial hypercellularity with low-grade mesangial IgM and C3 deposition (no IgG, IgA or C1q). Podocytes show diffuse foot process fusion (**slide 2**). No glomerular, tubulointerstitial or vascular sclerosis was present.

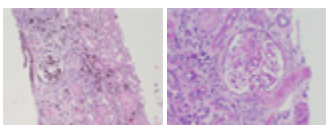
4. What is the diagnosis at this time? (Select from either **List 2** or **List 3**).

After the biopsy, the patient was started on cyclosporine and continued on prednisone. Urine protein became negative within 3 weeks. The patient was weaned off prednisone over the next 3 months and he was maintained on cyclosporine.

He continued in remission for a total of 11 months. However, coincident with a viral respiratory illness, he had a relapse with 4+ urine protein. Prednisone was re-started with remission within 3 weeks. Over the next several months, the patient proved steroid-dependent on several cycles of attempted weaning and restarting. Ultimately, he became steroid-resistant.

With a urine protein/creatinine ratio at 6.2, albumin at 2.3 g/dl and serum creatinine at 0.8 mg/dl, a second renal biopsy was done.

## Pathology Report for Biopsy #2



Slide 1

Slide 2

The biopsy shows areas of tubular atrophy with interstitial fibrosis and chronic inflammation. Occasional glomeruli show segmental areas of scarring (**slides 1-2**). Podocytes again show diffuse foot process effacement.

5. What is the diagnosis? (Select from either **List 2** or **List 3**).