

A 15-year-old boy comes to the physician because of hematuria and lower abdominal pain. This is his third episode of hematuria in the past 2 years. He has a family history of renal disease. His temperature is 37.1° C (98.9° F), blood pressure is 140/90 mm Hg, pulse is 80/min, and respirations are 14/min. Examination shows mild sensorineural deafness bilaterally. Urinalysis shows hematuria and proteinuria. Laboratory studies show BUN of 50 mg/dL and serum creatinine of 3.1 mg/dL; serum complement levels are normal. Renal biopsy shows foam cells, and immunofluorescence shows no immunoglobulins or complement. Electron microscopy shows alternating areas of thinned and thickened capillary loops with splitting of GBM. Which of the following is the most likely diagnosis?

- ☐ A. Alport's syndrome
- ☐ B. Acute interstitial nephritis
- ☐ C. Acute post infectious glomerulonephritis
- ☐ D. Anti-glomerular basement membrane disease
- ☐ E. Benign recurrent hematuria
- ☐ F. Goodpasture's syndrome
- ☐ G. Henoch-Schonlein purpura
- ☐ H. Idiopathic anti-GBM antibody mediated glomerulonephritis
- ☐ I. IgA nephropathy
- ☐ J. Mixed essential cryoglobulinemia
- ☐ K. Microscopic polyangiitis
- ☐ L. Systemic lupus erythematosus
- ☐ M. Thin basement membrane disease
- ☐ N. Wegener's granulomatosis

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[Proceed to Next Item](#)**Explanation:**User Id: [REDACTED]

The above vignette illustrated the classic presentation of Alport's syndrome. This is a familial disorder which usually presents in childhood as recurrent gross hematuria and proteinuria. Sensorineural deafness usually occurs. Electron microscopy findings include alternating areas of thinned and thickened capillary loops with splitting of the glomerular basement membrane (GBM).

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(Choice M) Thin basement membrane disease is also a familial disorder, but it presents in adulthood as microscopic hematuria without proteinuria. Renal biopsy reveals a markedly thinned basement membrane.

(Choice E) Benign recurrent hematuria is asymptomatic. Renal biopsy is normal in most cases. This condition has an excellent prognosis.

Educational Objective:

Suspect Alport's syndrome in patients with recurrent episodes of hematuria, sensorineural deafness and a family history of renal failure.

Time Spent: 2 seconds

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