

SIGNIFICANCE STATEMENT

Activation of the complement alternative pathway plays a key role in the pathogenesis of IgA nephropathy (IgAN). Large, international, genome-wide association studies have shown that deletion of complement factor H-related genes 1 and 3 (*CFHR3,1Δ*) is associated with a reduced risk of developing IgAN. This study compares the renal outcomes of patients with IgAN according to their *CFHR3,1Δ* genotype. We demonstrate that *CFHR3,1Δ* is associated with a reduced level of glomerular immune deposits, but is not associated with renal prognosis in a cohort of 639 patients. Study of the regulation of the complement alternative pathway may help in understanding the pathogenesis of this disease. Large-scale sequencing of alternative pathway regulation genes is required to evaluate the cumulative effect of variants.