SIGNIFICANCE STATEMENT

Dysregulation of the alternative pathway of complement activation is associated with atypical hemolytic uremic syndrome (aHUS) and C3 glomerulopathy (C3G). The role of properdin, a positive complement regulator, in aHUS and C3G remains uncertain. The authors previously showed that properdin inhibition exacerbated a murine model of C3G. In striking contrast, these studies show that blocking properdin in a murine model of aHUS prevented renal disease and systemic thrombophilia. These results suggest that properdin contributes critically to renal disease in aHUS and that its inhibition in this disorder and other renal disorders may have therapeutic benefit.