Human factor H-related protein 2 (CFHR2) regulates complement activation.

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Abstract

Mutations and deletions within the human CFHR gene cluster on chromosome 1 are associated with diseases, such as dense deposit disease, CFHR nephropathy or age-related macular degeneration. Resulting mutant CFHR proteins can affect complement regulation. Here we identify human CFHR2 as a novel alternative pathway complement regulator that inhibits the C3 alternative pathway convertase and terminal pathway assembly. CFHR2 is composed of four short consensus repeat domains (SCRs). Two CFHR2 molecules form a dimer through their N-terminal SCRs, and each of the two C-terminal ends can bind C3b. C3b bound CFHR2 still allows C3 convertase formation but the CFHR2 bound convertases do not cleave the substrate C3. Interestingly CFHR2 hardly competes off factor H from C3b. Thus CFHR2 likely acts in concert with factor H, as CFHR2 inhibits convertases while simultaneously allowing factor H assisted degradation by factor I.

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