Complement factor H related proteins (CFHRs).

Skerka C, Chen Q, Fremeaux-Bacchi V, Roumenina LT (2013) Complement factor H related proteins (CFHRs). *Mol Immunol* 56(3), 170-180.

Details

PubMed

Abstract

Factor H related proteins comprise a group of five plasma proteins: CFHR1, CFHR2, CFHR3, CFHR4 and CFHR5, and each member of this group binds to the central complement component C3b. Mutations, genetic deletions, duplications or rearrangements in the individual CFHR genes are associated with a number of diseases including atypical hemolytic uremic syndrome (aHUS), C3 glomerulopathies (C3 glomerulonephritis (C3GN), dense deposit disease (DDD) and CFHR5 nephropathy), IgA nephropathy, age related macular degeneration (AMD) and systemic lupus erythematosus (SLE). Although complement regulatory functions were attributed to most of the members of the CFHR protein family, the precise role of each CFHR protein in complement activation and the exact contribution to disease pathology is still unclear. Recent publications show that CFHR proteins form homo- as well as heterodimers. Genetic abnormalities within the CFHR gene locus can result in hybrid proteins with affected dimerization or recognition domains which cause defective functions. Here we summarize the recent data about CFHR genes and proteins in order to better understand the role of CFHR proteins in complement activation and in complement associated diseases.

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doi: 10.1016/j.molimm.2013.06.001

PMID: 23830046