Congenital defects in the expression of the glycosylphosphatidylinositol-anchored complement regulatory proteins CD59 and decay-accelerating factor

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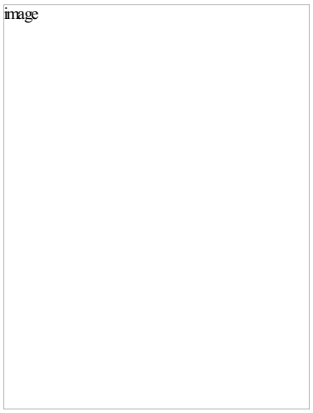

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Congenital defects in the expression of the glycosylphosphatidylinositol-anchored complement regulatory proteins CD59 and decay-accelerating factor

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Abstract

CD59 and decay-accelerating factor (DAF) are glycosylphosphatidylinositol (GPI)-anchored complement regulatory proteins critical for regulating complement activation on the host cell surface. Defective expressions of CD59 and/or DAF caused by mutations in the genes coding for these proteins or genes involved in the biosynthesis of GPI, such as PIGA, PIT and PIGM, are associated with various clinical symptoms that are mediated by dysregulated activation of complement, especially the C5 component. Eculizumab, an anti-C5 antibody, is effective in relieving the symptoms seen in patients with complement dysregulation.