Hybrid protein deregulates complement in dense deposit disease

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Dense deposit disease is a rare congenital disorder that is associated with complement dysfunction and often results in end stage renal disease within 10 years of the initial diagnosis. A small percentage of dense deposit disease is associated with mutations in the genes encoding factor H or C3 and autoantibody production.

In this issue of the *Journal of Clinical Investigation*, Peter Zipfel and colleagues at the Leibniz Institute for Natural Products Research and Infection Biology, evaluated an index family that had 2 reported cases of dense deposit disease.

The authors identified a chromosomal deletion in the complement factor H–related (CFHR) gene cluster that resulted in production of a hybrid CFHR2/CFRH5, which stabilized C3 convertase.

Treatment with soluble C1 restored C3 convertase decay and may be a promising treatment for patients with a similar refractory form of dense despite disease.

More information: Complement factor H–related hybrid protein deregulates complement in dense deposit disease, *J Clin Invest*. DOI: 10.1172/JCI71866

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