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Hemolytic Uremic Syndrome in
Pregnancy and Postpartum

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Abstract

Background Pregnancy is associated with various forms of thrombotic microangiopathy, including hemolytic uremic syndrome. A previous small French study suggested that pregnancy-associated hemolytic uremic syndrome was to be included in the spectrum of atypical hemolytic uremic syndrome linked to complement alternative pathway dysregulation.

Design, setting, participants, & measurements We sought to retrospectively analyze the presentation, outcome, and frequency of complement alternative pathway gene variants in a larger international (France, United Kingdom, Italy) cohort of patients with pregnancy-associated hemolytic uremic syndrome.

Results Eighty-seven patients with pregnancy-associated hemolytic uremic syndrome were included. Hemolytic uremic syndrome occurred mainly during the first pregnancy (58%) and in the postpartum period (76%). At diagnosis, 56 (71%) patients required dialysis. Fifty-six (78%) patients underwent plasma exchanges, 21 (41%) received plasma infusions, and four (5%) received eculizumab. During follow-up (mean duration of 7.2 years), 41 (53%) patients reached ESRD, 15 (19%) had CKD, and 18 (28%) patients experienced hemolytic uremic syndrome relapse. Twenty-four patients (27%) received a kidney transplant and a recurrence of hemolytic uremic syndrome occurred in 13 (54%) patients. Variants in complement genes were detected in 49 (56%) patients, mainly in the *CFH* (30%) and *CFI* genes (9%).

Conclusions Pregnancy-associated hemolytic uremic syndrome and atypical hemolytic uremic syndrome nonrelated to pregnancy have the same severity at onset and during follow-up and the same frequency of complement gene variants.

hemolytic uremic syndrome complement pregnancy
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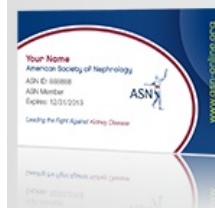
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