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Atypical hemolytic uremic syndrome (aHUS) clinical characteristics and treatment patterns associated with mortality during index hospitalization

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Background

aHUS is caused by dysregulation of the alternative complement pathway and is associated with significant morbidity and mortality. C5 inhibitors (C5i) improve thrombotic microangiopathy (TMA) response and renal recovery. This study described clinical characteristics and treatment patterns associated with mortality using real-world evidence from one of the largest, most diverse incident aHUS cohorts in the US during the C5i era.

Methods

This was a retrospective cohort study of 634 incident hospitalized adult aHUS patients, derived from the US Premier Healthcare Database, which contains ~25% of all US hospitalizations (1/2011–6/2021). aHUS was defined as the presence of a diagnostic code for TMA/HUS *and* a treatment code for a C5i in the absence of a diagnostic code for secondary causes of TMA/ HUS. Demographic and clinical characteristics were analyzed using t-test, Wilcoxon rank test, Fisher's exact test or Chi-squared test as appropriate.

Results

Overall, 78 patients (12.3%) died during hospitalization. Of 285 patients with mortality data post-discharge, 30- and 365-day mortality rates were 4% and 9%, respectively. In-hospital aHUS mortality was associated with older median age (63 vs 49 years, $p<0.001$), diabetes (30% vs 16%, $p=0.003$), longer median intensive care unit stay (14 vs 5 days, $p<0.001$) and hospital stay (31 vs 20 days, $p<0.001$), but not race, ethnicity, gender, history of hypertension, or CKD. In-hospital mortality was associated with a longer median delay between admission and therapeutic plasmapheresis (TPE) (5 vs 2 days, $p<0.001$) and C5i (15 vs 8 days, $p<0.001$). Although not associated with exposure or time until corticosteroids (CS), mortality was associated with duration of CS use (27 vs 16 days, $p<0.001$) and time from C5i initiation to CS discontinuation (14 vs 6 days, $p=0.02$). Mortality was associated with increased median time to renal replacement therapy (RRT) (5 vs 3, $p<0.001$) but not RRT requirement.

Conclusion

Delayed TPE, RRT, and C5i treatment and prolonged CS exposure are associated with increased mortality in aHUS. Future efforts should be made to reduce any treatment delays and develop more targeted, steroid-sparing treatments.

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Category

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Keywords (3 maximum)

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