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Atypical hemolytic uremic syndrome (aHUS) clinical characteristics and outcomes during index hospitalization diagnosis in the era of C5 inhibitor therapy (C5i)

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Background

C5i therapy has significantly improved clinical outcomes for aHUS. Due to its rarity, there are few previous large aHUS retrospective cohort studies and these are limited to patients from outside the US, children, and/or prevalent patients from before the era of C5i. This study sought to describe the clinical characteristics and outcomes in one of the largest and most diverse incident adult US aHUS cohorts treated with C5i to date.

Methods

This was a retrospective cohort study of adult aHUS patients in the US diagnosed during hospitalization, 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 thrombotic microangiopathy (TMA) or HUS and a treatment code for C5i in the absence of a diagnostic code for secondary causes of TMA or HUS. Prespecified clinical and outcome data were collected and analyzed using descriptive statistics.

Results

The cohort comprised 634 patients with a median age of 51 years (interquartile range [IQR] 32–64). The majority were female (67.2%), White (61.5%), and non-Hispanic (76.3%). Baseline comorbidities included hypertension (70.3%), chronic kidney disease (30.4%), and diabetes (18%). Median intensive care unit (ICU) and hospital stays (IQR) were 6 (3–11) and 20 days (14–32), respectively. The median time from admission to C5i was 9 days (5–15). For patients receiving corticosteroids (CS) (78.5%) and therapeutic plasmapheresis (TPE) (69.1%), the median time from admission to treatment was 2 days (1–5) and 3 days (2–5), respectively. Median treatment durations for CS and TPE were 17 days (8–28) and 5 days (3–12). Renal replacement therapy (RRT) was initiated in 77.3% of patients and discontinued prior to discharge in 51% of these patients. In-hospital mortality was 12.3%, and 22.7% of patients required follow-up in skilled care facilities.

Conclusion

Despite the substantial clinical benefits from C5i, aHUS remains a morbid disease, as shown by hospital and ICU stay duration, RRT incidence, discharge care requirements, and mortality described in this US cohort. Future efforts should focus on improved time to treatment and developing novel therapies.

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Category

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