Atypical hemolytic uremic syndrome (AHUS) is a rare disease characterized by complement over-activation and represents an estimated 5-10% of all hemolytic uremic syndrome cases. 

- Affects approximately 2 per million people in the United States. 
- Characterized by systemic thrombotic microangiopathy (TMA) which causes blood clots in small blood vessels. These blood clots can lead to stroke, heart attack, kidney failure, and other conditions. 
- AHUS can be of genetic origin or acquired and shows no variation by race or gender. 
- Although AHUS may develop at any age, it most commonly develops in children under the age of 5 years. 
- Prognosis is poor in patients diagnosed with AHUS with a 36% risk of death within five years after onset. 
- Treatment for patients with AHUS generally includes plasma exchange or infusion and administration of eculizumab. 

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Patient Demographics 

- The sample population included 6,571 visits representing 2,899 individual patients. 
- More than 70% of the hospital visits occurred in the outpatient setting. 
- Among the outpatient visits 57% had a primary diagnosis of hemolytic uremic syndrome and 43% had a diagnosis of chronic renal disease. The mean age of the sample was 23 years with 59.3% under the age of 18 and 61% female. 
- AHUS may be of genetic origin or acquired and shows no variation by race or gender. 
- These data represent only utilization within the hospital setting. 
- Significant drivers of increased length of stay are outlined in Table 2 and include transfusion (55.9%) followed by dialysis (34.1%). A small number of patients were treated with eculizumab (1.2%) and transplants (2.5%). 

Comorbidities 

- Charlson comorbidities for this population, outlined in Table 4, include renal disease (56.2%), congestive heart failure (11.1%), chronic pulmonary disease (10.0%), and diabetes without chronic complications (9.8%). 
- Other common comorbidities in the inpatient population included hypertension (12.0%), diabetes (11.4%), severe neurological disorders (7.3%), pancreatitis (6.4%), and thrombocytopenia (5.8%). 

Study Limitations 

- Among inpatient admissions, the average LOS was 16.02 days. 
- Significant drivers associated with increased length of stay are outlined in Figure 4 and include transfusion and the presence of a transplant. 

Readmissions 

- Some hospital readmissions occurred in 14.5% of the inpatient sample. 
- The most common diagnoses for readmissions are complications of surgical and medical care and diseases of the blood as shown in Figure 3. 
- Significant drivers of readmission including tumors and diabetes with chronic complications are shown in Figure 4. 

Procedures and Treatments 

- The most common inpatient procedures are shown in Figure 1. 
- In the inpatient setting the most common treatment was transfusion (55.9%) followed by dialysis (34.1%). 
- A small number of patients were treated with eculizumab (1.2%) and transplants (2.5%). 
- Eculizumab was the most prevalent treatment in the outpatient setting (7.5%). 
- Transfusions occurred in 4.5% of the patients. 

Table 1: Charlson Comorbidities 

Table 2: Patient Demographics 

Table 3: Charlson Comorbidities 

Table 4: Comorbidities 

Table 5: Charlson Comorbidities