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Clinical Profile And Outcomes Of Atypical Haemolytic Uremic Syndrome in a resource limited setting: A Retrospective, Single-center Study

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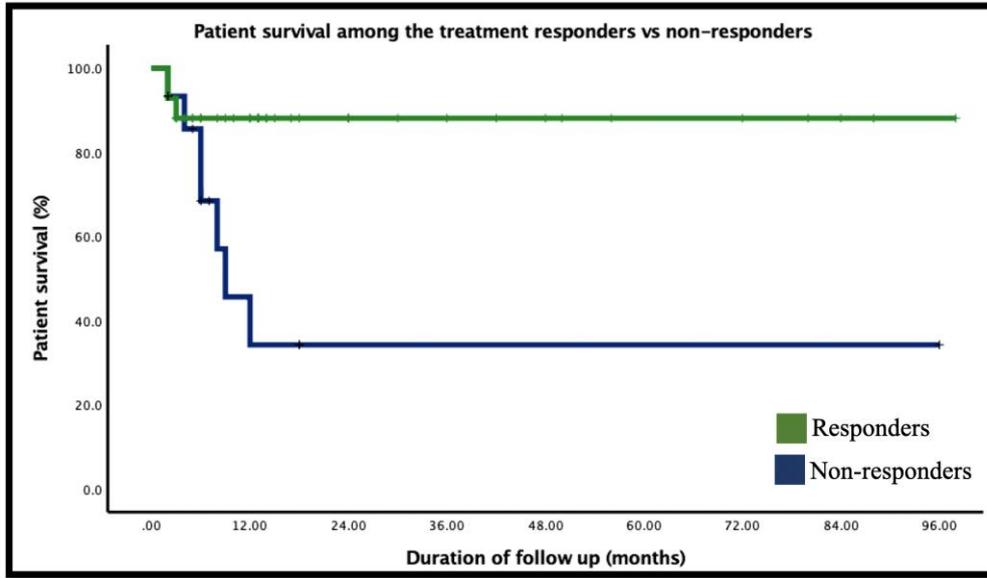[2]Department of Nephrology, Professor, India

Objectives : Atypical hemolytic uremic syndrome (aHUS) is a severe thrombotic microangiopathy predominantly affecting the kidneys, often associated with complement dysregulation. This study aimed to analyze the clinical characteristics, treatment outcomes, and long-term implications of aHUS, particularly focusing on anti-complement factor H(anti-CFH) antibodies, in a resource-limited setting.

Methods : A retrospective observational study was conducted at a single Indian institute between January 2016 and December 2022. We included all patients with aHUS, excluding secondary causes and renal transplant recipients. Demographic profiles, clinical features, laboratory parameters, treatment modalities (immunosuppression and plasma exchange), and outcomes were collected. Anti-CFH antibody, complement levels, and genetic mutation analysis were performed to ascertain etiological factors.

Results : Fifty-seven patients (mean age:12.5± 4.9 years; 63% males) were analyzed. Among them, eight presented post-partum, and 33(57.9%) tested positive for anti-CFH antibodies. Initial remission was achieved in 42 (73.6%) patients, with 13 (22.8%) partial and 29 (50.9%) complete remission. The median follow-up duration was 24 months (IQR 8.5-84); 12 (21%) patients died, with 2 deaths during the index admission, 6 among non-responders, and 4 among responders. On Kaplan-Meier analysis, the overall mean patient survival was 64.9 months (95%CI: 55.4-74.5). The mean patient survival in the responder group was 75.7 months (95%CI: 67.9-83.4), which was significantly higher as compared to patient survival in non-responders (31.7 months, 95% CI: 10.2-53.2; p<0.001). Dialysis-free renal survival was superior in anti-CFH seropositive patients (81.2%) compared to seronegative counterparts (55.9%), while patient survival was comparable. Elevated anti-CFH titers(>4000 AU/ml) correlated with suboptimal therapy responses. Additionally, age ≥16, female gender, and seizures predicted non-responsiveness.

Conclusions : This study emphasizes the significant impact of anti-CFH antibodies on aHUS in the Indian population. In resource limited settings, a combination of plasma exchange and immunosuppression showed promising results in the short and long term and may be a viable alternative in settings with non-availability of Eculizumab.

Figure#1. Patient survival.jpg

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Figure#1. Patient survival.jpg