AAN SUMMER CONFERENCE ABSTRACT

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EMBARGOED FOR RELEASE UNTIL 4 P.M. ET, WEDNESDAY, JULY 6, 2022

Abstract Title: Therapeutic Plasma Exchange in the Management of Stiff Person Syndrome Spectrum Disorders: A Case Series and Review of the Literature

Press Release Title: Preliminary Study: Blood Treatment is Safe, May Help Treat Stiff Person Syndrome

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Objective: To describe the safety, tolerability, and response to TPE in patients with SPSD.

Background: Stiff person syndrome spectrum disorders (SPSD) are a rare group of disabling neuroimmunological disorders. SPSD often require immune therapies especially in the setting of inadequate response to symptomatic treatments. The safety and efficacy of therapeutic plasma exchange (TPE) in SPSD is unclear.

Design/Methods: A retrospective review of medical records for patients with SPSD seen at Johns Hopkins Hospital was performed. Patient characteristics, exam findings, diagnostic studies, treatment response and TPE-related complications were recorded. We also conducted linear regression models to assess for possible predictive factors for TPE response. Literature review of SPSD and TPE was also conducted.

Results: Thirty-six SPSD patients were treated with TPE; mean age was 48 years, 81% female, and average anti-GAD65 antibody titer was 42352 U/mL (range, 0-309,902). Twenty-two patients had classic SPS, 10 had SPS-plus, and 4 had other phenotypes. Thirty-three patients were treated for acute exacerbations, and 3 were on maintenance TPE. There were 4 (11.1%) TPE-related complications (catheter infection, catheter thrombosis, hemorrhage), but no deaths or anaphylaxis. Twenty patients (55.6%) reported improvement in symptoms after TPE, 13 reported no change, and 3 reported worsening of symptoms. Of the 36 total patients who received TPE, 21 received TPE at Johns Hopkins Hospital for an acute exacerbation of their condition, with 12 requiring fewer anti-spasmodic medications 3 months after TPE treatment. There were no predictive factors in a positive treatment response to TPE. Literature review identified 42 more patients; 69% of these patients reported a temporary improvement in their condition.

Conclusions: We describe the safety and tolerability of TPE in patients with SPSD and show that TPE-related complications are uncommon and manageable. Additionally, many patients with SPSD derived improvement with TPE. Further studies could help inform clinicians when to use TPE in SPSD.