Long-term Rituximab Use Benefits Patients with Stiff Person Syndrome
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Objective:
To describe the effects of chronic Rituximab therapy in Stiff Person Syndrome (SPS).

Background:
Rituximab is effective for various neuroimmunologic conditions. Multiple reports suggested efficacy in SPS but a recently published clinical trial was negative. This trial had several limitations including small sample size (12 on active arm), lack of sensitive outcome measures, and importantly it was short in duration.

Design/Methods:
Data was obtained from more than 150 SPS patients followed at the Johns Hopkins Hospital. Demographics, SPS symptoms/signs, pertinent laboratory tests, and timed 25-foot walk (T25FW) were recorded. Data was managed using REDCap and analyzed using Stata15.

Results: Forty-one patients with SPS were exposed to Rituximab in our cohort. Twenty-three had the minimum information required for analysis. The majority were female(78%) and Caucasian(61%). Average age at first Rituximab infusion was 49 years (range:20-76 years). Mean follow up was 24 months (range:3-60 months). Twenty one patients were antiGAD65 antibody positive SPS, 1 seronegative SPS, and 1 antiGAD-associated pure cerebellar ataxia. Seventeen patients (74%) experienced improvement in SPS symptoms and physical findings. Five had no obvious efficacy and one experienced worsening. Eleven patients (68%) reported improvement in back spasms, 9(70%) in leg spasms and 11 (64%) in axial rigidity. Examination showed resolution of axial rigidity in 4 patients (22%), paraspinal muscle spasm in 6 patients (27%) while 6 (25%) patients had an objective improvement in gait. Sufficient T25FW data was available for 12 patients. Mean T25FW at baseline was 9.7 seconds (SD:±7.3) which decreased in last visit to 7.4 seconds (± 4.2). Five patients (42%) had significant improvement in walking speed defined as >20%change from baseline.

Conclusions:
Long-term Rituximab therapy appears to benefit many patients with SPS. This observed improvement may be related to frequency and/or consistency of treatment. Further studies are needed to better understand what factors are associated with Rituximab treatment responders versus non-responders.