

Subcutaneous Immunoglobulin (SCig) for Maintenance Therapy in Severe POTS: A Case Report.

Tahrira Ferdous, MD^{1*}

¹Division of Cardiovascular Medicine, Department of Medicine, The University of Toledo, Toledo, OH 43614

*Corresponding author: Tahrira.Ferdous@utoledo.edu

Published: 14 December 2023

Introduction: Introduction: Postural Orthostatic Tachycardia Syndrome (POTS) is a multisystem disorder involving the nervous and cardiovascular systems. POTS diagnosis requires an increase in heart rate of at least 30 beats per minute upon standing, in the absence of orthostatic hypotension (1).

There have been multiple theories suggesting a possible autoimmune pathogenesis of POTS, as multiple autoantibodies have been identified in patients with POTS such as ganglionic acetylcholine receptor (gAChR) and voltage-gated potassium channel complex, (Watari et al., 2018). Moreover patient with POTS tend to have high prevalence of ANA antibodies and co-occurrence of other autoimmune conditions such as systemic lupus (Blitshteyn, 2015).

The use of immunomodulator therapy such IVIG has been reported in multiple case reports with encouraging results, in this case we report significant improvement POTS symptoms with SCig in a patient with debilitating symptoms.

Case Presentation: A 42-year-old female with a past medical history significant for Ehlers-Danlos Syndrome complicated by multiple vertebral and disc issues was evaluated in the rheumatology clinic for severe debilitating POTS that was refractory to standardized treatment. The patient had repeated hospital admissions for syncopal episodes. The decision was made to start intravenous immunoglobulin and following an initial positive response, she developed aseptic meningitis. Intravenous therapy was discontinued, and the patient was started on subcutaneous immunoglobulin. At the time of this case report, the patient had been on subcutaneous Ig for more than 3 years. She has reported significant improvement in her symptoms with less hospitalization. The patient used an apple watch to measure and record her daily heart rate, summary of average HR reported over 3 years is summarized in the graph below.

Conclusion: This case highlights a case of debilitating POTS that showed a clinical subjective and objective response to subcutaneous immunoglobulin. which suggests a possible autoimmune pathway of disease. Further studies are needed to assess the efficacy and safety of Scig in the treatment of refractory POTS.

References

1. Sheldon, R.S., Grubb, B.P. II, Olshansky, B., Shen, W.K., Calkins, H., Brignole, M., Raj, S.R., Krahn, A.D., Morillo, C.A., Steward, J.M., Sutton, R., Sandroni, P., Friday, K.J., Hachul, D.T., Cohen, M.I., Lau, D.H., Maygua, K.A., Moak, J.P., Sandhu, R.K., Kanjwal, K. *2015 Heart Rhythm Society expert consensus statement on the diagnosis and treatment of postural tachycardia syndrome, inappropriate sinus tachycardia, and vasovagal syncope*. Heart Rhythm, 2015 Jun. **12**(6): pp. e41-63. doi: 10.1016/j.hrthm.2015.03.029
2. Ballow, M. *Safety of IGIV therapy and infusion-related adverse events*. Immunol Res, 2007. **38**(1-3): pp. 122-32. doi:10.1007/s12026-007-0003-5
3. Ruzieh, M., Batizy, L., Dasa, O., Oostra, C., Grubb, B. *The role of autoantibodies in the syndromes of orthostatic intolerance: a systematic review*. Scandinavian Cardiovascular Journal, 2017. **51**(5): pp. 243-247, doi:10.1080/14017431.2017.1355068