Immunopathogenesis of thrombotic thrombocytopenic purpura

- HLA-DQB1*03 and HLA-DRB1*11 are genetic factors involved in the loss of tolerance of the immune system towards ADAMTS13

- ADAMTS13 is endocytosed by APC and processed to peptides (FINVAPHAR and ASYILIRD) that are loaded on MHC-II molecules

- The complex MHC-II/peptide will get recognized by TCR, which will cause the activation of the CD4+ T-cell

- Such activated CD4+ T-cell will help autoreactive B-cell to produce ADAMTS13-specific auto-Abs

Hrdinová et al., Haematologica, 2018