AUTOIMMUNE HYPOGLYCEMIA EXPANDS THE BIOLOGICAL SPECTRUM OF HHV8+ MULTICENTRIC CASTLEMAN DISEASE.

D. Boutboul¹,8,¹⁰, P. Arnautou¹, M. Auclair², S. Fellahi³, C. Bouché⁴, C. Fieschi¹,⁸, E. Barrak⁵, V. Queyrel-Moranne⁶, L. Chaillous⁵, N. Blin⁵, J. Fadlallah¹,⁸, R. Bertinchamp¹,⁸, L. Gérard¹,⁸, D. Bengoufa⁷, L. Galicier¹,⁸, E. Oksenhendler¹,⁸, C. Vigouroux²,⁹

¹Clinical Immunology Department, Hôpital Saint Louis, Université de Paris, Paris, France
²Sorbonne Université, Inserm UMRS_938, Saint-Antoine Research Centre, IHU ICAN, Paris, France
³Biochemistry Laboratory, Hôpital Henri Mondor, Créteil, France
⁴Endocrinology Department, Hôpital Lariboisière, Université de Paris, Paris, France
⁵Endocrinology Department, CHU Nantes, Nantes, France
⁶Rheumatology Department, CHU Nice, Nice, France
⁷Immunology and Histocompatibility Laboratory, Hôpital Saint Louis, Université de Paris, Paris, France
⁸National Reference Center for Castleman disease, Hôpital Saint Louis, Université de Paris, Paris, France
⁹Assistance Publique-Hôpitaux de Paris, Saint Antoine Hospital, National Reference Centre for Rare Diseases of Insulin Secretion and Insulin Sensitivity (PRISIS), Department of Endocrinology, and Department of Molecular Biology and Genetics, Paris, France
¹⁰U976 HIPI, INSIGHT team, Hôpital Saint Louis, Université de Paris, Paris, France
Castleman disease (CD) is a rare non-clonal lymphoproliferative disorder. Diagnosis is based on histological features (1). Some cases of multicentric CD (MCD) are related to a polyclonal proliferation of HHV8-infected plasmablasts (called “HHV8+ MCD”), mostly encountered in immunocompromised hosts. The infected cells are uniformly IgM and λ-positive and locate in the mantle zone of the lymph node. These cells are associated with a marked polyclonal plasma cell infiltrate. Hemophagocytic lymphohistiocytosis (HLH) and other cytokine release complications make this condition life-threatening (2,3). Numerous auto-antibodies can develop during MCD flares, and autoimmune thrombocytopenia (ITP), autoimmune hemolytic anemia (AIHA) and Thrombotic Thrombocytopenic Purpura (TTP) (4) have been described.

We here report on autoimmune hypoglycemia as a new autoimmune complication occurring in 5 patients with active HHV8+ MCD. We demonstrate that hypoglycemia is caused by autoantibodies directed towards the insulin receptor and that rituximab is an effective and safe treatment of this condition.