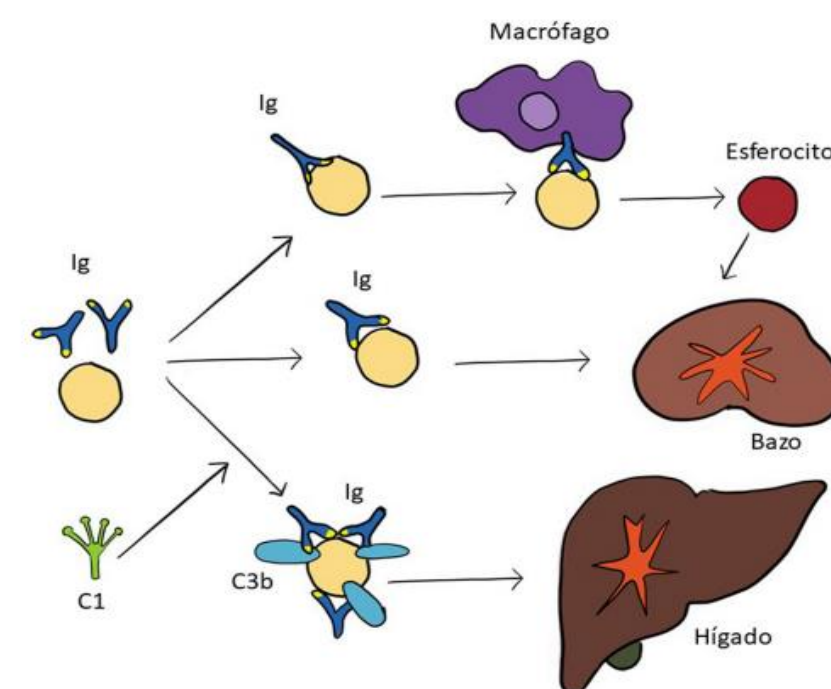


## BACKGROUND AND IMPORTANCE

Autoimmune hemolytic anemia (AIHA) due to warm antibodies is the most common type of autoimmune hemolytic anemia, usually IgG.

As first line treatment, after splenectomy in necessary cases, glucocorticoids with or without rituximab and intravenous immunoglobulins are used. In refractory or persistent cases, immunosuppressive agents and cytostatics such as mycophenolate mofetil, sirolimus or cyclophosphamide are used.



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## OBJETIVE

To evaluate the off-label treatment of abatacept for a patient with refractory warm antibody AIHA. The patient was diagnosed with autoimmune bicytopenia with anemia, plateletopenia, and splenomegaly. The direct Coombs' test was IgG positive, giving the diagnosis of warm antibody AIHA probably in the context of splenic lymphoproliferative syndrome. He received treatment with corticosteroids and rituximab and underwent splenectomy in 2019.

## METHODS AND MATERIAL

We describe treatment with abatacept, CTLA-4 agonist, in a 54-year-old patient with AIHA due to warm antibodies refractory to splenectomy and corticosteroids plus rituximab at 375 mg/m<sup>2</sup> weekly from February 2021, being retreated from February to April 2022. During all this time, he required numerous blood transfusions. The patient is a heterozygous carrier of the p.(Arg51+) variant in the CTLA4 gene, considered pathogenic and associated with the development of autoimmune lymphoproliferative syndrome, an increase in autoreactive B lymphocytes and the appearance of autoimmune cytopenias

## RESULTS

Haemoglobin values and transfusion requirements were analysed. Treatments with glucocorticoids plus rituximab had a mean haemoglobin of 11.3 g/dL (1.97;14.6-9.3) during the period February 2021 to March 2021 and 10.8 g/dL (1.4;12.9-9.7) during February to April 2022. Prior to these treatments, the patient required 7 blood transfusions and 3 transfusions during the treatment period. During abatacept treatment from August 2022 to June 2024 the mean haemoglobin was 12.5 g/dL (1.2;14.6-10.6) and no blood transfusions were required.

## CONCLUSIONS

Treatment with abatacept for patients with refractory warm antibody AIHA is poorly documented. The results obtained show a clear improvement in analytical parameters and a disappearance of haemolytic phenomena and transfusion needs. It is a valuable option for this type of patient.