

# Insights into EBV Infection in an Adult with X-Linked Lymphoproliferative Syndrome

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References



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## Background

- X-Linked Lymphoproliferative disease (XLP) is a rare Inborn Error of Immunity (IEI) characterized by immune dysregulation and increased susceptibility to Epstein-Barr Virus (EBV)<sup>1</sup>.
- Hemophagocytic lymphohistiocytosis (HLH), often associated with EBV<sup>2</sup>, is the leading cause of mortality in patients with XLP who forego haematopoietic stem cell transplantation (HSCT).
- Rituximab has been used in the treatment of EBV viremia in the setting of post-transplant lymphoproliferative disease<sup>3-4</sup>. In this case, it was ineffective.
- We present a patient with XLP1, and progressive EBV viremia, who responded well to one dose of nivolumab.

## Mechanism of Action

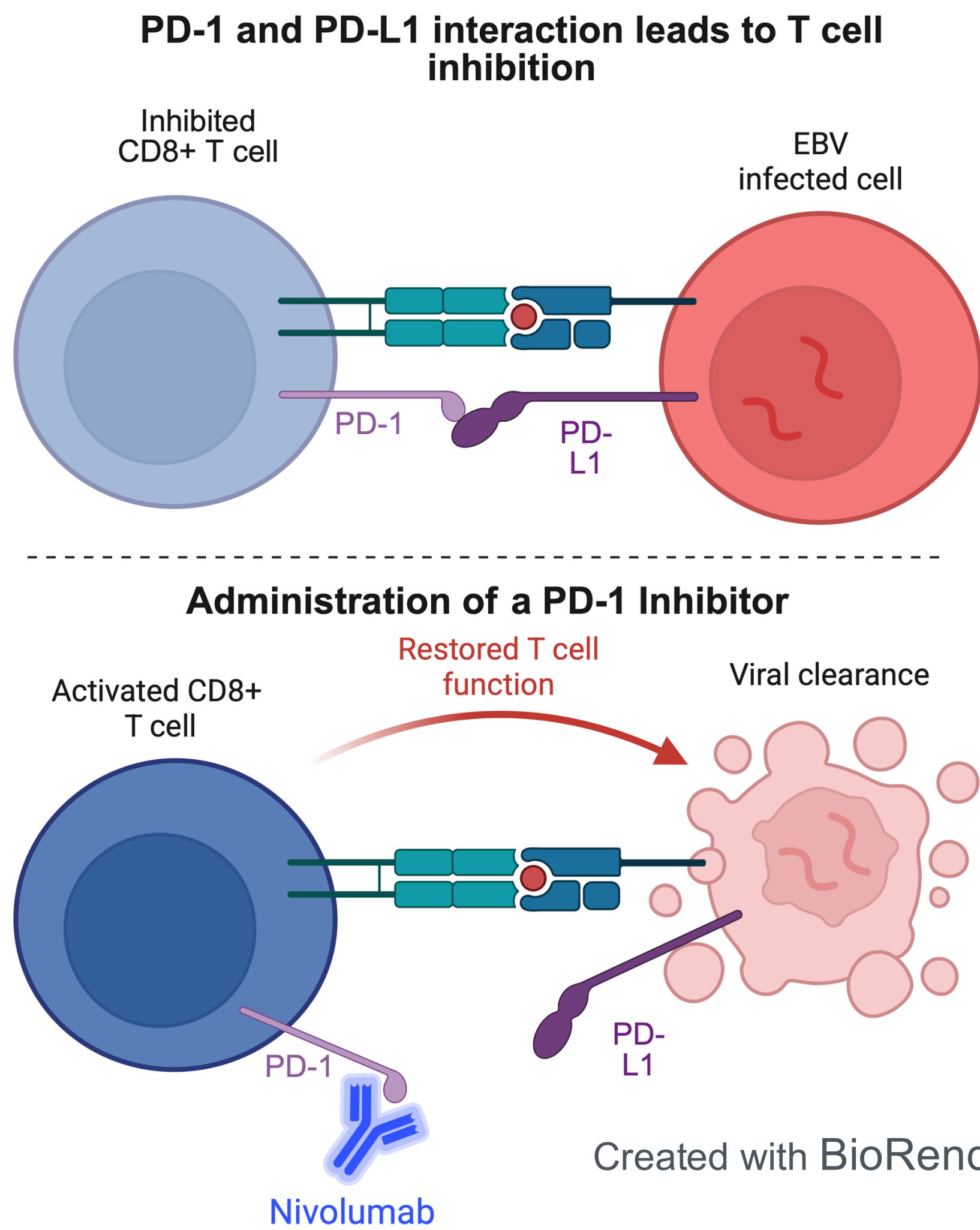
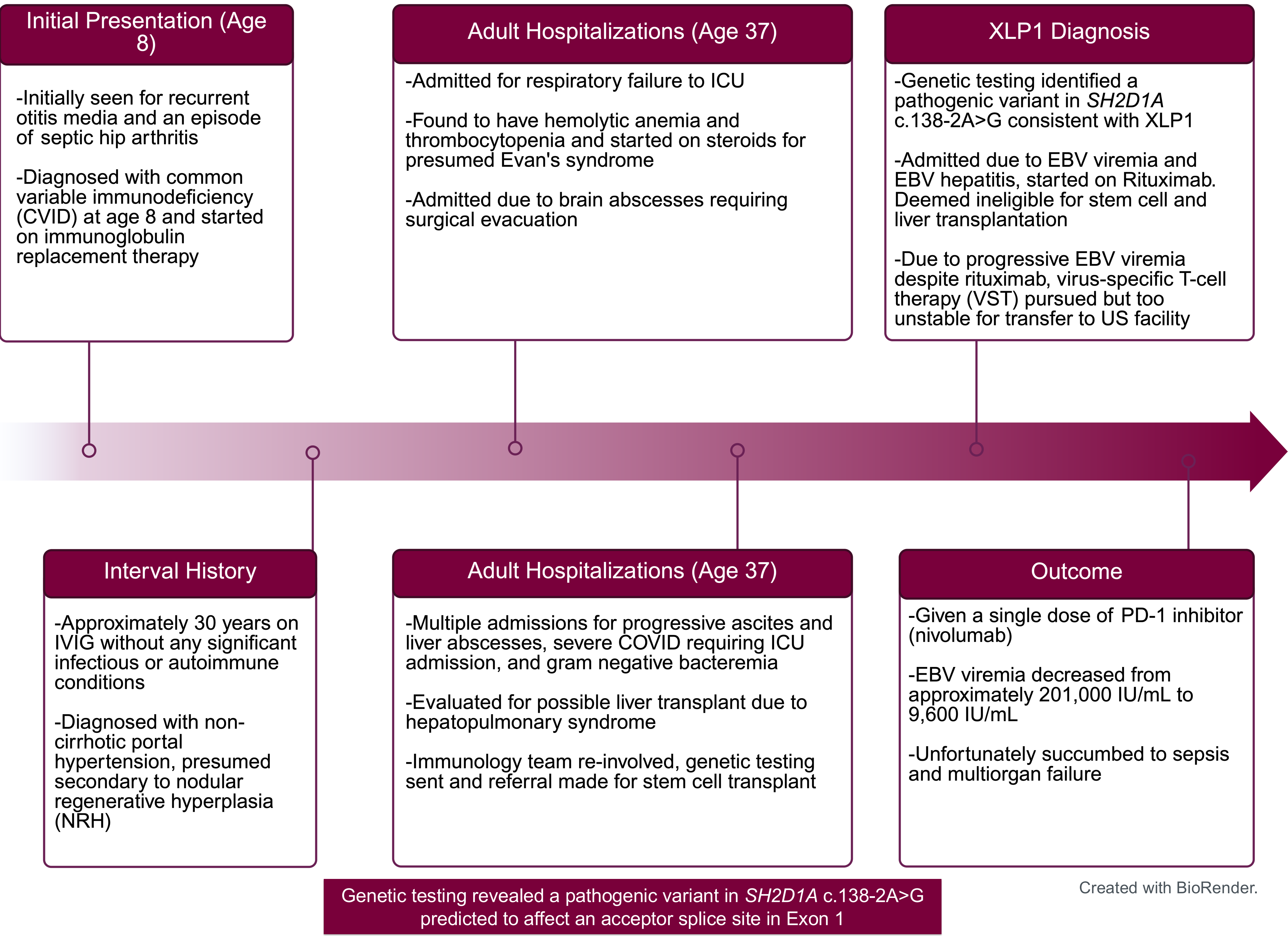


Figure 1. Mechanism of action of Nivolumab in EBV infection.

## Clinical Case Presentation



## EBV DNA and Nivolumab

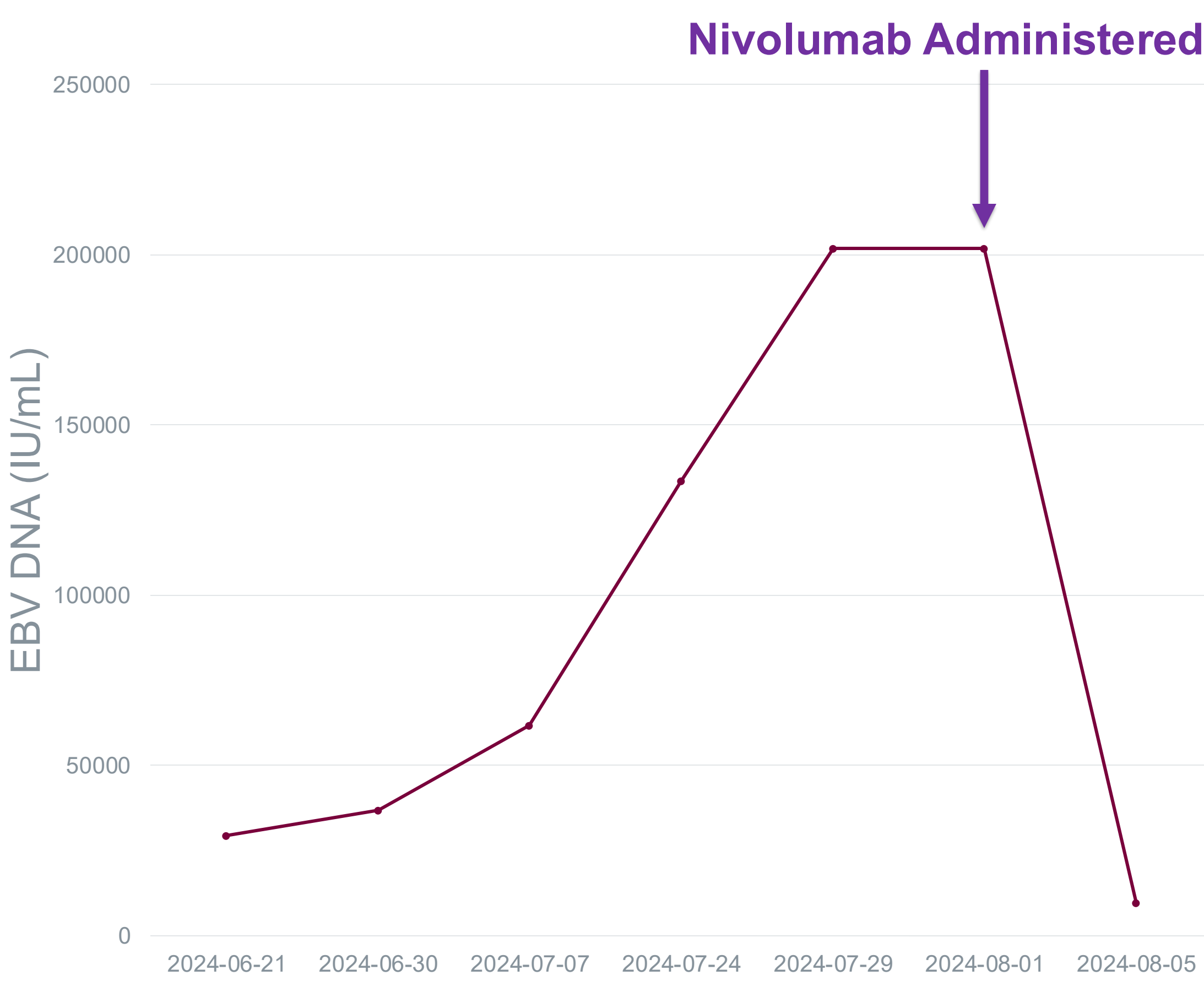


Figure 2. EBV DNA levels in relation to Nivolumab administration.

## Outcome and Recommendations

- Our patient ultimately succumbed to sepsis and respiratory failure.
- Despite limited literature, PD-1 inhibitors may play a significant role in the treatment of certain EBV-associated conditions, particularly in patients with IEIs.
- We recommend considering PD-1 inhibition for any patient with overwhelming EBV infection.
- Severe EBV disease in a patient with a longstanding CVID diagnosis emphasizes the need to genotype for potentially pivotal molecular diagnosis.
- Treatment options for EBV susceptibility disorders need further exploration and accessibility.