

Infant Male with Severe Combined Immunodeficiency Treated with IVIG Human-slra 10% for Unexplained Post-Transplant Fever.



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LEARNING OBJECTIVES

- To recognize that IVIG products are pooled antibodies from a group of donors with different compositions. IVIG needs to be matched to patient characteristics for improved safety and efficacy (1).
- To appraise and select different IVIG products.

BACKGROUND

- IVIG-slra is a commercially available IVIG product with high respiratory viral titers approved for children two years of age with primary immunodeficiency(2).

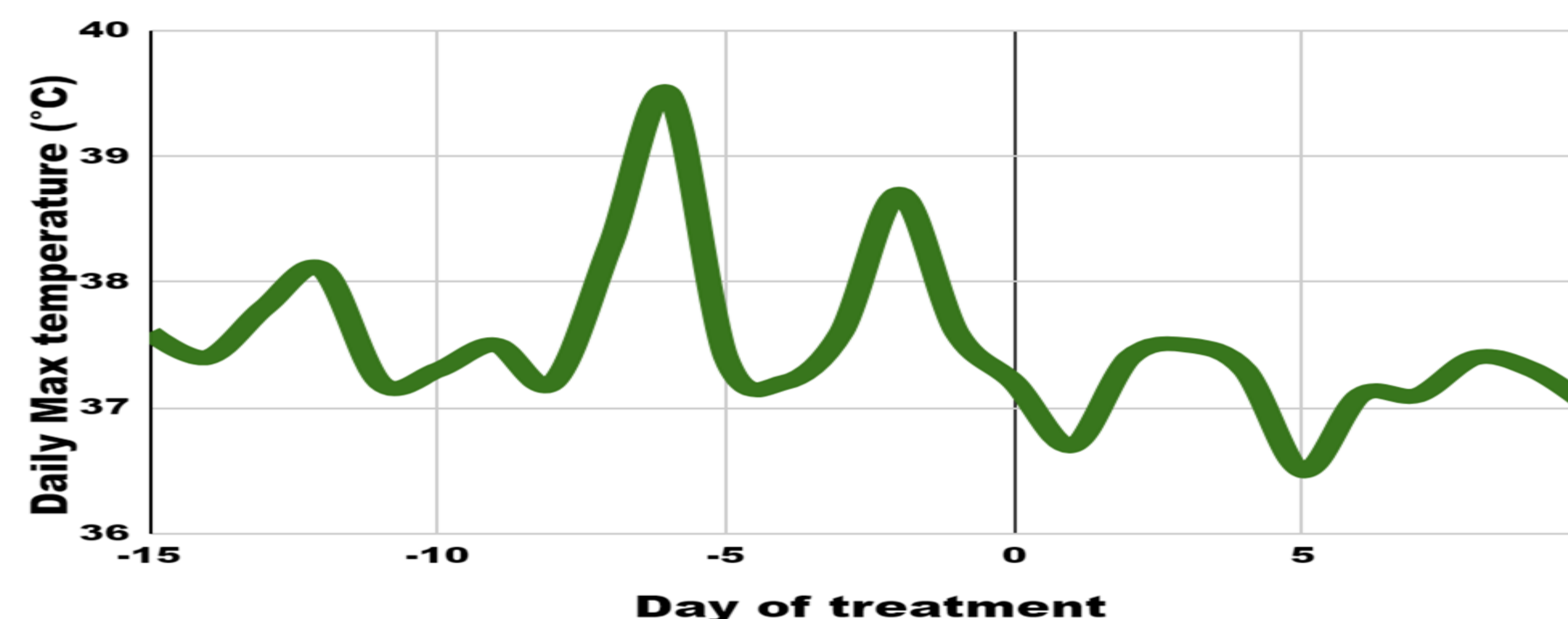
CASE PRESENTATION

- The patient is a 4-month-old male with X-linked T-B+NK- SCID (hemizygous pathogenic IL2RG c.676C>T p.Arg226Cys) diagnosed by newborn screen and family history of SCID.
- His pre-transplant course was notable for an upper respiratory infection with positive nasal PCR for rhinovirus/enterovirus which was persistently positive.
- He underwent matched unrelated donor bone marrow transplant with busulfan conditioning per CSIDE clinical trial protocol (NCT03619551).

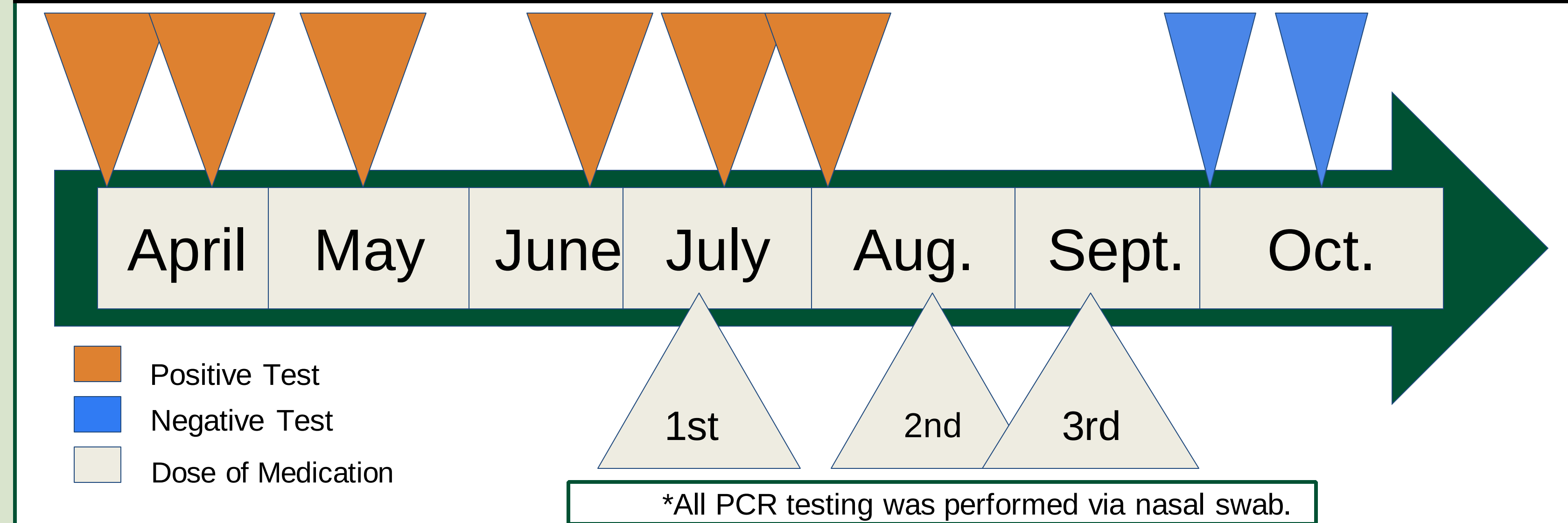
CLINICAL COURSE

- His post-transplant course was complicated by fluid overload requiring dialysis, hepatic veno-occlusive disease requiring defibrotide, Staphylococcus Hominis CLABSI, and respiratory failure requiring intubation.
- Approximately one-month post-transplant, he was febrile with negative infectious evaluation except for rhinovirus/enterovirus nasal PCR despite broad-spectrum antibiotics, antifungals, antivirals, and standard IVIG.
- The patient was administered IVIG-slra 570 mg/kg.
- His fevers ceased within days of infusion and he tolerated the infusion without any immediate adverse events.
- Patient was eventually rhinovirus/enterovirus negative by PCR after 2 additional infusions of IVIG-slra
- Patient was infection free but unfortunately suffered a catastrophic brain hemorrhage believed to be secondary to coagulopathy from defibrotide.

Fever Curve



Rhino/ Entero PCR testing



CONCLUSION

- Our case describes the off label use of IVIG Human-slra for an infant critically ill with severe combined immunodeficiency (SCID) and suspected respiratory viral infection.
- Continuous viral RNA detection for months suggest active replication and persistent infection(3).
- Although, we suspect that this is a viral fever. The patient was taking many medications and we cant rule out drug fever as a cause.
- Viral infections can be deadly in this vulnerable population(4,5). Administration of IVIG-slra can be considered when a respiratory viral infection is suspected(2).

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