

**Title:** Treatments for TAFRO syndrome in Japan

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**Abstract:** TAFRO syndrome is a systemic inflammatory disorder of unknown etiology characterized by thrombocytopenia, anasarca, fever, reticulin myelofibrosis, renal dysfunction, and organomegaly, and frequent lymphadenopathy with Castleman disease-like histology. In this study, we explored optimal treatments for this syndrome using a patient cohort registered in a retrospective registry in Japan. Among 81 patients with TAFRO syndrome, 68 received corticosteroids as the first-line treatment, and as the second-line treatment, 21 received tocilizumab (Toc), 14 received cyclosporine A (CsA), and 8 received rituximab (Rit) in addition to corticosteroids. We compared these second-line treatment groups by setting the primary endpoint as time to next treatment or death (TTNT). Kaplan-Meier analysis showed that the median TTNT in the Toc, CsA, and Rit groups were 2.8 months, 9.2 months, and not reached, respectively. The TTNT of the Rit group was significantly longer than that of the Toc group. In contrast, there were no significant differences in overall survival between groups. Approximately 30% died within 1 year, and the major causes of death were infections. Further studies are warranted to establish the optimal treatment strategies for this syndrome.

During this poster session, a part of the results published in the [International Journal of Hematology 2021, 113\(1\):73-80](#) will be presented.