**EMAPALUMAB, AN INTERFERON GAMMA (IFN-γ)-BLOCKING MONOCLONAL ANTIBODY, IN PATIENTS WITH MACROPHAGE ACTIVATION SYNDROME (MAS) COMPLICATING SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS (SJIA)**

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**Objective:** To collect retrospective data in patients with JIA and other diseases with respect to vaccination with live-attenuated vaccines in patients with rheumatic diseases on high-dose DMARDs, glucocorticoids or biological agents. Common practice is to withhold vaccination with live-attenuated vaccines in patients with rheumatic diseases on high-dose DMARDs, glucocorticoids or biological agents, due to limited safety data, and the (theoretical) risk of introducing an infectious disease to the patient. Evidence for this approach is limited. We collected data from pediatric rheumatologists who vaccinate these patients, to obtain additional safety data, which might update and revise this approach.

**Methods:** Data from 13 pediatric rheumatology centers in 10 countries were collected.

**Results:** 234 patients were reported; mean age 5±2.7, 70% girls. 206 had JIA; 46% oligoarticular, 36% polyarticular, 8% systemic, 5% SPA types, 5% JIA and uveitis. 48% of JIA patients were in remission on medication. Disease activity was low in 38%, high in 2%, moderate in 7%; 11 patients had juvenile dermatomyositis, 3 systemic and 2 localized scleroderma, 4 isolated idiopathic uveitis, 1 CINCA syndrome, 1 MKD, and 1 FMF.

**Conclusion:** Vaccination with live-attenuated vaccines in patients with rheumatic diseases is safe: multicenter, retrospective data collection.

**References:**


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**RESULTS:**

Patients 5 (females, median age 11 years, range: 2-25 years) received emapalumab according to the prescribed dose regimen. Treatment duration ranged from 3 (2 patients) to 4 weeks. Prior to emapalumab, all patients failed methylprednisolone pulses, in 2 patients plus cyclosporine A (CsA) and in 2 patients plus CsA and anakinra. The achieved emapalumab concentrations led to rapid neutralization of IFNγ as indicated by CXCL9 levels and subsequent deactivation of T cells as indicated by sIL2R levels. In all patients, complete response was achieved by week 8, in 4 patients by end of treatment (Table). Systemic glucocorticoids were weaned in all patients. Emapalumab infusions were well tolerated and none of the patients discontinued emapalumab. A CMV reactivation was reported by investigator as a serious event possibly related to emapalumab, but resolved completely with treatment.

**Conclusion:** Emapalumab administration with the tested dosing regimen led to rapid neutralization of IFNγ as shown by normalization of CXCL9, associated with evidence of decreased T cell activation. In all patients, emapalumab treatment was effective in controlling MAS with a favourable safety profile.

**REFERENCE:**