two drugs, or standard therapy. The term "rare" was defined by the European Union as a condition that occurs in no more than 1 in 2,000 individuals.
Two review authors independently assessed trial quality and extracted the data. We screened the search results and included studies if they met the selection criteria. If we identified two or more trials that investigated the same rare disease and used the same assessment tools we performed a meta-analysis.
Results: 135 studies were screened, of which 34 met the inclusion criteria. In total, we analysed data on 11 different orphan diseases, encompassing 2,324 participants. There was a high degree of statistical and clinical heterogeneity in these trials. Several sources of potential bias were identified in the included studies, for example, a lack of description of the blinding methods and allocation concealment, as well as the small size of the study populations. We included studies such as rituximab against cyclophosphamide in ANCA-associated vasculitis. These studies demonstrated a non-inferiority of rituximab. The meta-analysis resulted a combined odds ratio (OR) of 1.42 in favour of rituximab (95% CI). Further meta-analyses were possible for another 22 studies involving, among others, Behçet's disease, systemic sclerosis, cryopyrin-associated periodic syndromes, and giant cell arteritis. Compounds studied were immunosuppressants like corticosteroids, methotrexate and azathioprine, or biologicals such as rilonacept, infliximab, and canakinumab.
Conclusions: A high degree of evidence is hampered by the limited number of study participants in each trial. On the other hand, diseases such as systemic sclerosis, ANCA-associated vasculitides, or Behçet's disease had more high quality trials available. The amount of data for most other rare disease remains unsatisfactory.
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THU0577 TOCILIZUMAB MONOTHERAPY FOR ADULT ONSET STILL'S DISEASE – RESULTS OF 52-WEEK TREATMENT OF A PROSPECTIVE, SINGLE-CENTER, SINGLE-ARM, OPEN TRIAL

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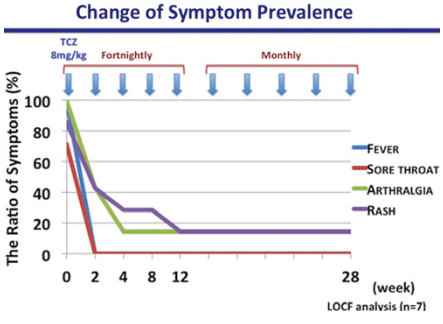
Background: Adult-onset Still's disease (AOSD) is a systemic inflammatory disease of unknown etiology. Corticosteroids still provide mainstay AOSD therapy despite various adverse effects. Recently, AOSD patients have been successfully treated with anti-cytokine therapies such as with TNF-α blocking agents, an IL-1 receptor antagonist, and an anti-IL-6 receptor monoclonal antibody. Among these case reports, TCZ seems to be highly effective for treating patients refractory to TNF antagonists and IL-1 antagonist.
Objectives: To assess the efficacy and safety of tocilizumab (TCZ) monotherapy for the induction therapy of adult onset Still's disease (AOSD) in a prospective single-arm, single-center, cohort, pilot study.
Methods: Seven AOSD patients (male 2, female 5) who had agreed with our prospective trial since April 2010 till May 2015 were enrolled. Our study protocol is that patients received 8 mg/kg of intravenous TCZ fortnightly for the first two months (five courses), then monthly for the next 5 months and after that they stop TCZ therapy and we monitor symptoms about AOSD relapses. In this report, we evaluated the efficacy and safety in 52 week. Efficacy was evaluated by serum markers (WBC, CRP and serum ferritin), clinical symptoms and ratio of patients who required additional therapy, and safety was evaluated by adverse events for 52 week.
Results: The mean age was 41.4. The ratio of fever, arthralgia, rash and sore throat are 100% (n=7/7), 100% (n=7/7), 85.7% (n=6/7) and 71.4% (n=5/7) respectively. LOCF analysis revealed that WBC, CRP and serum ferritin level decreased significantly from 14757±4667/μl to 6985±2903/μl, from 13.4±7.1mg/dl to 0.3±0.6mg/dl and from 6927±5376ng/ml to 2416±5589ng/ml at month 7 (TCZ final infusion) respectively (each, $P<0.01$). The improvement rate of fever, arthralgia and eruption were 100% (n=7/7), 85.7% (n=6/7) and 85.7% (n=6/7) respectively at month 7. One patient couldn't continue TCZ therapies because of lack of efficacy (14.3%, n=1) and required additional therapy (prednisolone) at week 2. Another patient also abandoned this trial due to adverse event (14.3%, n=1, urinary tract infection). The other 5 patients could complete the 7-month course of the study and had no symptoms at month 7. Four of five patients had no flare-up signs until month 5 after stopping TCZ. One patient had relapse symptoms such as rash and arthralgia and increased serum level of CRP and serum ferritin at month 2 after final TCZ infusion. There were no serious adverse events during the course of the trial.



Conclusions: TCZ monotherapy can be an alternative treatment strategy for AOSD in some patients.
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[1] Sakai R, et al.:Clin Rheumatol. 2012 Mar;31(3):569–74.
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Disclosure of Interest: T. Kondo: None declared, Y. Okada: None declared, A. Shibata: None declared, K. Chino: None declared, T. Kurasawa: None declared, A. Okuyama: None declared, H. Takei: None declared, K. Amano Grant/research support from: Chugai Pharmaceutical Co.Ltd.
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THU0578 SPECTRUM OF THE DISEASES WITH ORBITAL INVOLVEMENT IN RHEUMATOLOGY: SINGLE-CENTER STUDY

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Background: It is assumed that in ophthalmological clinical practice lymphoproliferative disorders in patients with affection of different orbital organs and tissues constitute 25–55% and 21% of patients have IgG4-related ophthalmic disease [1,2].
Objectives: To evaluate the most common conditions affecting the orbit in rheumatologic clinical practice.
Methods: During 2004 – 2016 years 138 patients (male – 33, female - 105) with eyelid edema and/or tumefactive lesions in the orbit were examined in Nasonova Research Institute of Rheumatology. In all patients full clinical, ophthalmological, serological (rheumatoid factor, C-reactive protein, IgG, IgG4, IgM, IgA, ANA, anti-Ro/La, C3/C4 complement) examination was carried. In all cases diagnosis was verified pathomorphologicaly with immunohystochemical staining (anti-CD 138, CD 68, IgG, IgG4, κ-chain, λ-chain). Diagnosis was established on the basis of pathomorphological examination of different tissues: orbit – 79 patients, parotid salivary gland – 40, submandibular salivary gland – 14, nasal – 4, lymph nodes – 3 and other – 6. In 79 cases (54%) the diagnosis was established on the results of orbital biopsy.
Results: Different non-neoplastic diseases were diagnosed in 108 patients (78.5%) and 30 had different conditions of hematological spectrum, including malignant conditions in 25 (18.2%) patients (see table 1). Some patients at baseline had simultaneous involvement of the major salivary glands (23 patients with IgG4-related disease, 35 with sarcoidosis, 12 with non-Hodgkin lymphomas and 2 with AL-amyloidosis). The most rare conditions affecting the orbit were Cogan's syndrome, relapsing polychondritis, Erdheim-Chester disease, NK/T-cell nasal lymphoma and calcifying aponeurotic fibroma.

Table 1. Spectrum of the diseases with orbital involvement in rheumatologic practice (n=138)

	N, pts	N, %		N, pts	N, %
Non-neoplastic conditions	**108**	**78.5**	**Hematologic conditions**	**30**	**21.5**
• IgG4-related disease	48	35.0	• non-Hodgkin lymphomas	20	14.5
• Granulematous lesions (sarcoidosis, granulematosis with polyangiitis, necrosing sarcoidal granulematosis)	41	29	• Erdheim-Chester disease	1	0.7
• Autoimmune dacryoadenitis (non-differentiated)	7	5	• AL-amyloidosis	3	2.1
• Idiopathic orbital inflammation	7	5	• NK/T-cell nasal lymphoma	1	0.7
• Endocrin ophthalmopathy	2	1.4	• Histiocytosis	4	2.9
• Cogan's syndrome	1	0.7	• Calcifying aponeurotic fibroma	1	0.7
• Relapsing polychondritis	1	0.7			

Conclusions: In rheumatologic practice in 78.5% of patients with orbital involvement different non-neoplastic conditions are diagnosed: IgG4-related ophthalmic disease (35.0%), granulematous lesions (29%). The most common hematological disorders in rheumatologic clinic are non-Hodgkin lymphomas (17.5%) and histiocytosis (3.5%).
References:
[1] Japanese study group of IgG4-related ophthalmic desease. A prevalence