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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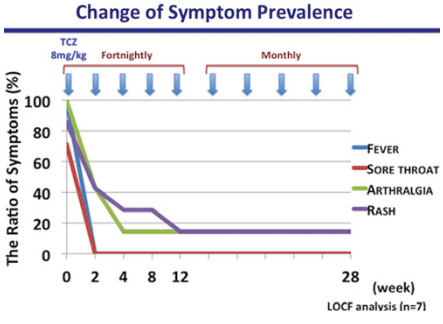
Background: Adult-onset Still's disease (AOSD) is a systemic inflammatory disease of unknown etiology. Corticosteroids still provide mainstay AOSD therapy despite various adverse effects. Recently, AOSD patients have been successfully treated with anti-cytokine therapies such as with TNF-α blocking agents, 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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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%).
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Disclosure of Interest: None declared
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THU0579 HYPOCOMPLEMENTEMIA IS RELATED TO ELEVATED SERUM LEVELS OF IGG SUBCLASSES OTHER THAN IGG4 IN IGG4-RELATED KIDNEY DISEASE

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Background: IgG4-related kidney disease (IgG4-RKD) is a comprehensive term for renal lesions associated with IgG4-related disease [1]. IgG4-RKD is frequently complicated by hypocomplementemia [1, 2, 3], but its clinical significance and mechanisms have not been clarified.
Objectives: This study aimed to investigate clinical features of IgG4-RKD patients with hypocomplementemia compared with those without it, leading to clarification of the clinical significance and mechanisms of hypocomplementemia.
Methods: We extracted 25 patients with IgG4-RKD between September 2005 and December 2016 in our hospital. Based on the presence/absence of hypocomplementemia at diagnosis, we divided them into a hypocomplementemia group (n=11) and normal complement group (n=14), and retrospectively analyzed various clinical features (age, sex, serum IgG levels, serum IgG4 levels, gaps between serum IgG and IgG4 level, ratio of serum IgG to serum IgG4, serum IgG subclasses, serum IgE levels, serum creatinine levels, urinary protein and urinary occult blood, urinary β2-microglobulin, urinary N-acetyl-β-D-glucosaminidase, initial dose of prednisolone, serum IL-2R levels, multiple organ lesion) during the clinical course in the two groups.
Results: The patients comprised 18 men and 7 women with an average age of 67.5 years (range, 44 to 81 years). Serum IgG levels (3971±729 mg/dL vs. 2157±598 mg/dL; $p<0.001$), gaps between serum IgG and IgG4 level (2992±770 mg/dL vs. 1482±444 mg/dL; $p<0.001$), serum IgG1 levels (2043±1025 mg/dL vs. 891±209 mg/dL; p=0.017), and the number of involved organs (4.1±1.1 vs 2.9±1.1; p=0.018) were significantly different between the two groups, while serum IgG4 levels (979±477 mg/dL vs. 791±575 mg/dL; p=0.298) and serum creatinine levels (1.96±1.89 mg/dL vs. 1.09±0.48 mg/dL; p=0.298) were not. At relapse of renal lesions, although both groups showed serum IgG4 re-elevation, the hypocomplementemia group showed exacerbation of hypocomplementemia and re-expansion of gaps between serum IgG and IgG4 level, while the normal complement group did not.
Conclusions: Hypocomplementemia may be associated with multiple organ involvement and elevation of IgG subclasses other than IgG4 including IgG1 in IgG4-RKD. In patients who initially show hypocomplementemia, a decline in serum complement levels implies renal lesion relapse.
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THURSDAY, 15 JUNE 2017

Education

THU0580 WHAT ARE THE PATIENTS' ISSUES AND NEEDS RELATED TO THEIR BIOLOGICS (BDMARDS) AND METHOTREXATE (MTX) TREATMENT IN DAILY LIFE: A QUANTITATIVE CROSS-SECTIONAL SURVEY AMONG 344 PATIENTS TO DEVELOP AN EDUCATIONAL SMARTPHONE APP

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Background: MTX and bDMARDs are the core treatments of chronic inflammatory arthritis (IA). We lack information on the patients' problems and needs in daily life, particularly on safety issues.
Objectives: 1-collect the most frequent issues 2-explore the patients' perceptions on a dedicated smartphone application (SP App) 3) determine the profile of the patients interested.
Methods: The survey was conducted on line. The questionnaire was designed by rheumatologists, methodologists, anthropologists, members of patients association (AP) and included 1-a non-exhaustive list of potential issues in daily life (fever, vaccines, ...) and practical aspects 2-a list of potential use of the App 3-free opinions 4- Two self-administrated questionnaire to test patients' knowledge on bDMARDs [1] and MTX [2].
Results: The survey was carried out from June to August 2016 on the websites of the PA. Non-associative patients were recruited by 3 rheumatologists who provided the Internet link. 344 patients responded, 331 analyzed, 83% female, 50% had rheumatoid arthritis, 40% had spondyloarthritis, mean age 53 years, 60% were AP; 67% were treated with MTX, 70% had bDMARDs, 34% had MTX-bDMARDs combotherapy.
66% of patients reported problems; 67% had needed help or advice. The main issues were infections (27%), vaccines (13%), surgery (10%), dental care (7%), self-administration (6%), conservation/travelling (9%) and skipped doses (5%).
Among the 76% patients who have a SP, 80% use Apps and 32% Apps for their health. Among users, 87% patients would find an App useful to manage their treatment (36% rather agree and 51% strongly agree), 82% for symptoms requiring to stop their treatment, 93% for situations related to safety, 80% as a reminder of their treatment, 80% to know what to do in case of a skipped dose, 77% to have a safety checklist before treatment administration, 66% to recall the modalities of self-injections. Patients interested in the App are younger ($p<0.05$) non-associative ($p<0.05$) and live in medium-sized cities ($p<0.01$). No correlation was found with other sociodemographic characteristics, level of education, type/duration of arthritis or knowledge.
Conclusions: Two-third of patients with arthritis face issues related to their treatment especially in case of infections, vaccination, surgery and travelling. A dedicated App is considered useful by 87% patients who already have a SP. The potential use of the App may improve safety, adherence and self-management in daily life.
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THU0581 WHAT WE SEE, WHAT WE LEARN, AND THE PREVALENCE OF RHEUMATIC DISEASES IN OUR POPULATION: A DIAGNOSIS CORRELATION STUDY

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Background: The postgraduate program in rheumatology aims learning of musculoskeletal and autoimmune disorders. In México, objectively-structured clinical examination (OSCE) is applied in postgraduate certification processes by the Mexican Board of Rheumatology annually [1]. Peláez-Ballestas et al. described an epidemiological study (COPCORD, Community Oriented Program for the Control of Rheumatic Diseases) of 19,213 individuals in 5 regions in our country where they found a prevalence of musculoskeletal pain in 25.5%, osteoarthritis in 10.5%, back pain in 5.8%, rheumatic regional pain syndromes in 3.8%, rheumatoid arthritis (RA) in 1.6%, and fibromyalgia in 0.7% [2].
Objectives: The aim of the study is to describe the student training in rheumatic diseases and correlate them with OSCE assessment and the prevalence of rheumatic diseases in our population.
Methods: An observational and analytical study was made between March 2014 to March 2015 in a single rheumatology training center at University Hospital. Student training was defined according to the times they evaluated patients with a determined diagnosis, this information was obtained by medical records. We categorize OSCE questions according to the rheumatic diagnosis. Finally, the two results were compared with prevalence of the rheumatic diagnosis according to COPCORD, which were registered according a score pain >4. We made descriptive statistics and a Spearman's Rho to evaluate the correlations of the diagnosis frequencies by each category.
Results: We reviewed 6279 medical records, 854 (13.6%) were of first-time evaluation. We had 5,400 (86.4%) women, with a mean age of 47.9 (SD 15.45) years.
Descriptive statistics are in Table 1 and Figure 1, which included: medical consultations, OSCE assessment and a column with rheumatologic diagnosis according to COPCORD.
The Spearman correlation coefficients of the 32 different diagnoses were: student training vs OSCE 0.492 (p=0.004), student training vs COPCORD 0.597 (p=0.01) and OSCE vs COPCORD 0.624 (p=0.01).
Conclusions: Although the most common musculoskeletal disease in our community did not obtain the frequency observed by students or evaluated in