treatment of systemic-onset and polyarticular course juvenile idiopathic arthritis (JIA), and is used off-label in other treatment refractory inflammatory diseases. Real-world data regarding tolerance of TCZ in children are scarce.

Objectives: To assess the incidence rate and type of serious adverse events in children treated with TCZ.

Methods: A single-center retrospective review of all consecutive patients receiving TCZ in the pediatric rheumatology division from 01/2007 to 12/2018 was performed. Inclusion criteria were a diagnosis of inflammatory rheumatic disease and treatment with at least one dose of TCZ. Patients with missing data were excluded. Serious adverse events (SAE) were defined as a life-threatening event and/or an event requiring hospital admission, leading to permanent disability or treatment discontinuation.

Results: A total of 70 children (39 females) were included. Thirty children had systemic JIA, 25 polyarticular course JIA, 14 systemic inflammatory disorders such as autoinflammatory syndromes and one patient severe refractory idiopathic uveitis. Median age at diagnosis was 4.2 years (IQR 2.0 – 7.3). The majority of children had received prednisone (n=56), and/or at least one biologic agent (n=63) prior to TCZ initiation. At TCZ start, children had a median age of 8.9 years (IQR 4.9 – 12.3) and disease duration of 3.2 years (IQR 0.9 – 7.4). TCZ was initiated at a 2-weekly interval in 44 children, 26 children received TCZ every 4 weeks. Fifty-eight children received concomitantly another immunosuppressive agent (prednisone (n=47) and/or methotrexate (n=20) and/or various other drugs (n=5)). A median of 26 TCZ doses were given per child (IQR 7 – 48), TCZ exposure was 146.8 patient years. Twenty-two SAE were observed in 18 (26%) children (SAE 14.9/100 patient years): 8 severe infusion reactions, 7 serious infections, 3 cutaneous reactions (toxic reaction, cutaneous vasculitis, scleroderma-like skin thickening with fasciitis), two macrophage activation syndromes, and one abnormal hepatic function and severe dizziness each. Infusion reactions occurred most frequently during the first few infusions (median during 4th infusion, IQR 3 – 5). All patients with infusion reactions had either systemic JIA (n=4) or another autoinflammatory disease (n=4). At time of infusion reaction, all children (8/8) had active disease, 7/8 had a concomitant treatment with prednisone and 2/8 additionally with cyclosporine.

At last follow up, 35 (50%) children were still on TCZ, the other 35 had discontinued TCZ: 4 for remission, 13 because of inefficacy, 12 because of side effects, 3 because of both inefficacy and side effects, and 3 for other reasons. No death occurred during TCZ treatment; two patients with systemic inflammatory disease, in whom TCZ had been inefficient, died from uncontrollable disease 13 and 15 months after TCZ discontinuation, respectively.

Conclusion: In this cohort, serious adverse events were observed in a quarter of children with half of them having severe infusion reactions. Higher incidence rates of severe infusion reactions were observed in children with systemic JIA or autoinflammatory diseases. Ongoing careful monitoring of patients treated with TCZ, especially those with important systemic inflammation is required.

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SAT0487

THE OUTCOMES OF JUVENILE IDIOPATHIC ARTHRITIS (JIA) IN CHILDREN MANAGED WITH CURRENT TREATMENTS: THE SINGAPORE EXPERIENCE

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Background: JIA remains one of the most common rheumatic diseases afflicted children in SE Asia¹. Despite the improvement in our treatment, outcomes of children treated with current therapy are unknown.

Objectives: To describe the clinical outcomes of children with JIA in a large, single center cohort in Singapore

Methods: Children diagnosed with JIA with a minimum follow-up of 6 month duration were recruited from our registry, from 1997 to 2015, at KK Women's and Children's Hospital, Singapore. Kaplan-Meier survival analyses were used to estimate the probability of ever attaining inactive disease (ID), remission with (CRM) or off (CR) medication according to modified Wallace criteria². Revised Steinbrocker functional classification

was used to assess functional status of JIA patients at their last follow-up visit. Non-parametric descriptive statistics including median (IQR) were used to described data.

Results: In a cohort of 251 children (62% male and 71% Chinese; 35% ERA, 34% oligoarthritis and 12% RF-negative polyarthritis were among the most common JIA subtypes encountered locally), 43% used biologics and 65% used methotrexate. Median follow-up was 35 months (IQR 13 – 70). Of all subtypes, 89%, 71% and 21% achieved ID, CRM and CR at least once during the course of follow-up, respectively. Over 85 – 100% of all JIA children could experience ID, including RF-positive polyarthritis (Fig. 1, 2). The probabilities of attaining ID within 1 and 5 years were 30 – 81% and 80 – 100%. The probabilities of attaining remission within 1 and 5 years were 10-59% and 67-100% across JIA subtypes. All of our psoriatic arthritis and systemic onset JIA reached remission on medication within 3 and 5 years, respectively. Only 33.6% of children could totally stop their medication at 1 year, but 19.3 – 87.0% could do so at 5 years. However, none of the RF-positive polyarthritis and undifferentiated JIA children had CR during the course of their diseases. Functional classes were achieved in 96%, 4%, 0.4% and 0% for class I – IV, respectively.

Fig 1. Cumulative proportion of children attaining clinical inactive disease and clinical remission on medication

Fig 2. Proportion of children achieving at least one time of clinical inactive disease and remission in each JIA category during the follow-up period.

Conclusion: Majority of children with JIA managed by our current therapy attained inactive disease in 2 years. Despite significant improvement in JIA therapy, most of them remained on medications to maintain their inactive disease status. Only 20% were able to totally withdraw treatment. Almost all of our JIA children had no disability.

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SAT0488 THE FRENCH PAEDIATRIC COHORT OF CASTLEMAN DISEASE

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Background: Castleman disease (CD) is a very rare non-malignant lymphoproliferation of undetermined origin. CD diagnosis is difficult and often delayed because of insidious onset, low awareness and clinical heterogeneity. Two major disease phenotypes can be distinguished: unicentric CD (UCD) and multicentric CD (MCD). Diagnosis confirmation is based on histopathological findings on an involved lymph node.

Objectives: We attempted to survey all cases of paediatric CD identified in France so far, in order to set up a national registry aimed to improve CD early recognition, treatment and follow-up, within the context of a new reference centre (<http://www.castleman.fr>).

Methods: In 2016, we e-mailed a questionnaire to members of the French paediatric immunohaematology society, the French paediatric rheumatology society and the French Reference Centre for Castleman Disease to retrospectively collect cases of paediatric CD (first symptoms before age 18 years). Anatomopathological confirmation was mandatory.

Results: We identified 23 patients (12 girls and 11 boys) diagnosed with UCD (n=17) and MCD (n=6) between 1994 and 2018 in 14 centres. The average age at first symptoms was 11.5 years for UCD and 8.3 years for MCD. The mean diagnosis delay was 8.2 months for UCD and 5.2 years for MCD. In UCD, the initial symptoms were: isolated lymph nodes (n = 10) or lymph node associated with other symptoms (n = 7), fever was present in 3 patients. Patients with MCD presented with fever (n=5), abdominal lymph nodes (n=5), failure to thrive (n=3), hepatomegaly and/or splenomegaly (n=3), arthralgia (n=2), abdominal pain (n=2), fatigue (n=2), facial oedema (n=1), isolated lymphadenopathy (n=1), trunk rash (n=1), vascular hepatopathy with oesophageal varicose veins (n=1), diarrhea (n=1) or cholestasis (n=1). One patient had a Duchenne muscular dystrophy. No patients had HIV or HHV8 infection.

In MCD cases, one patient with recurrent fevers, had a heterozygous mutation in *MEFV* gene and another had a homozygous mutation in *TNFRSF1A* gene (P75L). A UCD patient experienced fever episodes and pericarditis two years after surgery. Genetic tests revealed one heterozygous mutation in *IL10RA* gene (V406L) and one in *IL36RN* gene (S113L). Nevertheless, our patient did not have any psoriasis or inflammatory bowel disease.

Treatment of UCD was mainly surgical resection, steroids, and radiotherapy. Treatments of MCD were tocilizumab, rituximab, anakinra, steroids, chemotherapy and splenectomy. Overall survival after an average of 6 years of follow-up, was 100% for both unicentric and multicentric forms.

Conclusion: Paediatric CD seems still underdiagnosed with a significant diagnosis delay specially for the MCD but new international criteria will help in future. In UCD, surgery is the main treatment. New drugs are used specially for the MCD, but more studies need to be conducted in children. The global survey is better in children than adults.

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Milet: None declared, Sarah Jannier: None declared, Eric Jeziorski: None declared, Yves Perel: None declared, Francois Maurier: None declared, Christophe Piguet: None declared, Eric Oksenhendler: None declared, Isabelle Koné-Paut Grant/research support from: SOBI has supported drug product (anakinra) for the presented study, Consultant for: SOBI, Novartis, Pfizer, Abbvie, UCB, CHUGAI, ROCHE, Caroline Galeotti: None declared DOI: 10.1136/annrheumdis-2019-eular.4425

SAT0489 PREVALENCE OF AUTOIMMUNE DISEASES IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Juvenile idiopathic arthritis (JIA) is one of at least 80 different types of autoimmune diseases, and is the most common paediatric rheumatic disease. After cancer and heart disease, autoimmune diseases are the most common type of disease in the US. The estimated prevalence of autoimmune diseases is 3.2% and 5.9% in the US¹ and Europe², respectively, with a corresponding increase in mortality.³ Many autoimmune diseases share a common pathogenic mechanism. There are few published studies quantifying the occurrence of other autoimmune diseases in patients with JIA.

Objectives: To estimate and compare the prevalence of 38 co-existing autoimmune diseases in patients with JIA and in a general paediatric population.

Methods: This was a retrospective cohort study conducted using registry data from the Cincinnati Children's Hospital Medical Center (CCHMC; Cincinnati, OH, USA) from Jan 2010 to Oct 2018. Patients <21 years old who had at least one clinic visit at the CCHMC were included in the general patient (GP) cohort, excluding pts with JIA. The JIA cohort consisted of patients <21 years old who had one of the International Classification of Diseases (ICD)-9 or ICD-10 diagnosis codes for JIA. No exclusion criteria were applied. Cases for each of 38 autoimmune conditions (AICs) were identified using ICD9/10 codes.

Comparisons were made between the JIA and the GP cohorts for each of the 38 AICs in crude prevalence (reported as X cases per patient-year) for each year up to the end of the study period. A Bayesian Poisson regression model was used for estimating and comparing prevalence rates for the JIA and GP cohorts at the end of study period. The rate and corresponding 95% confidence limits of the two cohorts on their prevalence rates are reported.

Results: There were 2,026 and 41,572 patients included in the JIA and GP cohorts, respectively. Out of 38 AICs, 26 showed significantly higher prevalence in the JIA cohort compared with the GP cohort (Figure). Of the 38 ICD9/10 codes assessed, 14 (36.8%) showed a more than 20-fold greater prevalence in the JIA cohort than in the GP cohort. There

Figure. Significant results from Bayesian Poisson analyses on point prevalence comparisons: JIA cohort vs GP cohort

GP=general patient; JIA=juvenile idiopathic arthritis; NEC=not elsewhere classified