calcineurin inhibitors was 92% at 5th year. Serious infection occurred in four patients (19%) in the CI +group, and one of them had a fatal course.

Conclusions: Our retrospective analysis suggested that calcineurin inhibitors could be an additional option for AOSD and further prospective studies were desired.

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SAT0613 INCREASED SERUM LEVEL OF IL-36 RECEPTOR ANTAGONIST IS ASSOCIATED WITH ACTIVE DISEASE IN PATIENTS WITH ADULT-ONSET STILL'S DISEASE

H. Kim, J. Kang, B. Nam, J.S. Kim, D.-H. Yoo. *Division of Rheumatology, Hanyang University Medical Center, Seoul, Korea, Republic Of*

Background: Adult onset Still's disease (AOSD) is an inflammatory disorder which was associated with varying level of pro-inflammatory cytokines. However, the role of natural anti-inflammatory molecules has not been evaluated to date. Interleukin (IL) 1 cytokine family is closely related to clinical presentations, disease activity and thus is a target for treatment of AOSD. IL-36 receptor antagonist (Ra) is an anti-inflammatory molecule, but its clinical significance has not been studied in AOSD patients.

Objectives: To figure out the role of IL-36Ra in monitoring disease activity in patients with AOSD.

Methods: The number of 49 AOSD patients meeting Yamaguchi criteria were recruited. Each patient was serially monitored following clinical course of flare and remission, which presented at least 2 points of change in modified Pouchot's score. They were divided into two groups by clinical courses, which were predominant systemic symptoms or arthritis. We compared erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), ferritin as disease activity markers between flare and remission. Serum levels of inflammatory cytokines, IL-18, IL-37, and IL-36Ra were measured by enzyme-linked immunosorbent assay (ELISA) in each clinical state of AOSD patients.

Results: Forty-nine patients with AOSD were included in this study; 40 were women (81.6%) and 9 were men (18.3%), with mean age of 49.08 ± 14.17 years old. The mean duration of follow-up was 6.44 ± 3.87 years, and mean difference of modified Pouchot's score was 5.37 ± 1.98 between remission and flare. The number of 33 (67.4%) patients had presented systemic symptoms predominantly, while 16 (32.7%) presented arthritis more frequently in their clinical course. In flare state of AOSD, overall inflammatory markers were elevated, including cytokines

of IL-18 and IL-37. The serum level of IL-36Ra was 164.04 ± 169.03 pg/mL in active state, compared to 125.36 ± 452.0 in inactive state of AOSD patients ($p < 0.001$). IL-36Ra presented positive correlation with modified Pouchot's score and inflammatory markers, including CRP ($r = 0.286$, $p < 0.01$), ferritin ($r = 0.225$, $p < 0.05$) and IL-37 ($r = 0.353$, $p < 0.01$), but was not with level of IL-18 and ESR in active AOSD. Distribution of IL-36Ra level was analysed by each clinical course, however, there was no significant difference in level of IL-36Ra by clinical presentations stratified with predominance of arthritis.

Abstract SAT0613 – Table 1. Comparison of serum IL-36Ra levels in active AOSD patients with other markers for disease activity

	Modified Pouchot's score	ESR	CRP	Ferritin	IL-18	IL-37
IL-36Ra	0.295 ^a	0.086	0.286 ^b	0.225 ^a	0.095	0.353 ^a

Correlation efficient was presented by each inflammatory markers; ^a $p < 0.05$, ^b $p < 0.01$

Conclusions: Serum IL-36Ra level was significantly increased in active AOSD patients compared to inactive AOSD patients, presenting positive correlation with other inflammatory markers. In patients with AOSD, level of IL-36Ra might be another potential serologic marker to estimate disease activity, especially active state of the disease.

Disclosure of Interest: None declared

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SAT0614 NEW VARIANT IN THE IL1RN-GENE ASSOCIATED WITH LATE ONSET AND ATYPICAL PRESENTATION OF DIRA

J.B. Kuemmerle-Deschner¹, K. Hoernagel², S. Schlipf³, S. Hansmann⁴, A. Hospach⁵, X. Liu⁶, S.M. Benseler⁷, A.N. Weber⁸. ¹Department of Pediatrics, Division of Rheumatology, University Hospital Tuebingen; ²Praxis für Humangenetik Tuebingen, Tuebingen; ³Kinderarztpraxis Dr. Lakner, Schwäbisch Gmünd; ⁴Department of Pediatrics, Division of Pediatric Rheumatology, UNIVERSITY HOSPITAL TUEBINGEN, Tuebingen; ⁵Zentrum für Pädiatrische Rheumatologie, Klinikum Stuttgart, Olgahospital, Stuttgart; ⁶Department of Immunology, University of Tuebingen, Tuebingen, Germany; ⁷Rheumatology, Department of Paediatrics, Alberta Children's Hospital, University of Calgary, Alberta, Calgary, Alberta, Canada

Background: Deficiency of the interleukin-1 receptor antagonist (DIRA) is an autoinflammatory disease characterised by severe systemic inflammation with bone and skin involvement present in the first days of life.

Objectives: We report a novel variant in the *IL1RN*-gene associated an atypical phenotype of DIRA.

Methods: A 3 year-old Caucasian boy presented with recurrent monthly episodes of fever and fatigue, associated with lymphadenopathy, pericarditis, pleuritis, pancreatitis, and arthritis involving sacroiliac, hip, knee and ankle joints in the absence of any skin involvement. Symptoms had started at age one and had progressed over time to life-threatening episodes requiring intensive care therapy. Throughout, inflammatory parameters including ESR, CRP, SAA, S100A8/9, leukocytes, and platelets were highly elevated. Treatment with colchicine and steroids improved symptoms, however did not prevent flares. Immune deficiencies were ruled out; genetic testing for FMF, CAPS, TRAPS, HIDS and DITRA did not reveal variants in the associated genes.

Results: Whole exome sequencing detected a novel homozygous stop variant c.62C>G; p.Ser21* in the *IL1RN* gene (NM_173842.2). Mother, father and brother are heterozygous for the same variant. In addition, three variants of unknown significance were identified in the patient's *PCGF5*, *CPA1* and *SPTA1* genes. Functional studies revealed only marginal secretion of IL-1RA in the patient's unstimulated leucocytes and after stimulation with IL-1 β and LPS, confirming the disease-causing nature of the variant. Subsequently, IL-1 inhibition with anakinra at 2 mg/kg/d was started resulting in complete resolution of clinical symptoms, normal inflammatory markers and dramatically improved energy levels. Intolerance to daily subcutaneous injections prompted a switch to canakinumab at 4 mg/kg/4 weeks. However patient's and mothers assessment of disease activity was inferior on canakinumab compared to anakinra. After four months a flare appeared and lead to return to anakinra.

Conclusions: This is the first report of the novel c.62C>G; p.Ser21* variant in the *IL1RN*-gene primarily causing severe serositis in a homozygous carrier, while heterozygous family members were completely symptom free. Skin disease, one of the most prominent features in other patients with DIRA was not observed in this patient, while IL-1 inhibition was likewise effective.

Pathogenic variants in all reported DIRA patients so far affect all 4 isoforms of IL-1RA.¹ The different phenotype in the patient reported here, may be due to the selective loss of secreted IL1RN.

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SAT0615 SYMPATHETIC JOINT EFFUSION IN AN URBAN HOSPITAL

J.L. Barlow¹, I.J. Tan². ¹Department of Internal Medicine; ²Department of Internal Medicine; Section of Rheumatology, Temple University Hospital, Philadelphia, USA

Background: Sympathetic joint effusion (SJE) or sympathetic synovial effusion (SSE) is a rheumatologic entity that has not been well defined in the medical literature. It is a non-inflammatory synovial fluid collection that is associated with infection or inflammation of an adjacent anatomic structure.¹ The epidemiology and clinical characteristics of SJE/SSE are largely unknown. This knowledge gap has led to a lack of recognition and misdiagnosis by clinicians.

Objectives: Our study aimed to determine incidence, demographic information, and describe the clinical characteristics and potential triggering conditions for this presumptive reactive phenomenon.

Methods: We conducted a study of patients >18 years of age hospitalised at Temple University Hospital (TUH) between January 31, 2010 and December 10, 2016 who underwent diagnostic arthrocentesis for painful effusions. Individuals with synovial fluid white blood cell count (WBC) in the normal range of 200 WBC/mm³ or less were included. Patients with both non-inflammatory and inflammatory range synovial fluid of greater than 200 WBC/mm³ were excluded to limit confounders. Demographic and clinical data of 72 patients were included for detailed analysis.

Results: SJE/SSE was seen in 80/882 hospitalised patients (incidence of 9%). Seventy-two patients fulfilled inclusion criteria for detailed chart review. Demographic information revealed: male 46/72 (64%), female 26/72 (36%), African-American 38/72 (53%), Caucasian 16/72 (22%), Hispanic 10/72 (14%), undefined and other 8/72 (11%). Onset was typically acute, with 45/72 (63%) of patients developing symptoms within six days of arthrocentesis. All patients (100%) with SJE/SSE presented with painful effusion, and a minority had physical findings of warmth 23/72 (32%) or erythema 12/72 (17%). Interestingly, nearly a third of patients 21/72 (29%) were misdiagnosed with crystal or septic arthritis based solely on clinical exam, and empiric treatment was often administered prior to arthrocentesis. The most commonly affected joint was the knee 61/72 (85%), followed by the elbow 5/72 (7%), shoulder 3/72 (4%) and hip 3/72 (4%). Identifiable pathology in the affected limb was found in 29/72 (40%) of patients. Infection was the most common etiology, found in 17/29 (59%) of patients, and included cellulitis, abscess, osteomyelitis, septic bursitis, myositis, and necrotizing fasciitis. The majority of cases of SJE/SSE 23/29 (79%) were associated with concomitant infection, DVT or intramuscular hematoma in the affected limb which required specific therapeutic interventions.

Conclusions: Sympathetic joint effusion or sympathetic synovial effusion (SJE/SSE) is relatively common in hospitalised patients. SJE/SSE may be a sentinel sign for a more serious disorder affecting the same limb. Clinicians should maintain a heightened index of suspicion for SJE/SSE. A search for underlying infection, venous thrombosis, and intramuscular hematoma in the affected limb is warranted when encountering acute painful joint effusion with normal range synovial fluid WBC count.

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SAT0616 TEN-YEAR RETENTION RATE OF INFLIXIMAB IN PATIENTS WITH BEHÇET'S DISEASE-RELATED UVEITIS

J. Sota¹, C. Fabiani², A. Vitale¹, G. Emmi³, G. Lopalco⁴, V. Venerito⁴, I. Orlando¹, R. Franceschini¹, B. Frediani¹, M. Galeazzi¹, F. Iannone⁴, G.M. Tosi¹, L. Cantarini¹. ¹University of Siena, Siena; ²Humanitas Clinical and Research Center, Milan; ³University of Florence, Florence; ⁴University of Bari, Bari, Italy

Background: To date, a few studies have reported the long-term efficacy of Infliximab (IFX) in Behçet's disease (BD)-related uveitis.^{1,2} Nevertheless, it is known that TNF- α inhibitor drugs may lose their efficacy over time due to the formation of anti-drug antibodies, causing secondary failure and influencing the retention rate of these agents.³ In this regard no previous study has specifically investigated IFX retention-rate in BD-related uveitis.

Objectives: To evaluate the 10 year drug retention rate of IFX in BD-related uveitis, the effect of a concomitant use of disease modifying anti-rheumatic drugs (DMARDs) on drug survival and differences according to the lines of biologic treatment.

Methods: Cumulative survival rates were studied using the Kaplan-Meier plot, while the Log-rank (Mantel-Cox) test was used to compare survival curves.

Results: Forty patients (70 eyes) were eligible for analysis. The drug retention rates at 12-, 24-, 60- and 120 month follow-up were 89.03%, 86.16%, 75.66% and 47.11% respectively. No differences were identified according to the use of concomitant DMARDs ($p=0.20$), while a statistically significant difference was observed in relation to the different lines of IFX treatment ($p=0.014$). Visual acuity improved from baseline to the last follow-up visit ($p=0.047$) and corticosteroids-sparing effect was observed ($p<0.0001$).

Conclusions: IFX retention rate in BD-uveitis is excellent and is not affected by concomitant DMARDs

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SAT0617 COMORBIDITY IN PATIENTS AFFECTED BY AVASCULAR NECROSIS OF THE FEMORAL HEAD AND IMPACT OF COEXISTENT DISEASES ON CONTRALATERAL JOINT INVOLVEMENT

K. Gudkevich¹, N. Martusevich¹, A. Murzich². ¹Belarusian State Medical University; ²RSPC for Traumatology and Orthopedics, Minsk, Belarus

Background: Avascular necrosis (AN) represents a significant problem of modern rheumatology. The highest incidence of this disease is in able-bodied men. The risk of disability among patients with avascular necrosis of the femoral head (ANFH) is high, especially in case of bilateral process, as AN tends to progress steadily and results in osteoarthritis due to untimely diagnosis. Patients with AN can be affected by other coexistent diseases, that can influence the disease presentation and management.

Objectives: This study aimed to analyse the prevalence and pattern of comorbidities in patients affected by ANFH and to determine if there is a differential pattern of comorbidities in patients with bilateral and unilateral ANFH.

Methods: This was a cross-sectional study, comprising 102 adult patients, aged 48,^{38–58} with a diagnosis of ANFH according to Ficat and Arlet criteria. Sociodemographic, anthropometric, clinical parameters of interest were collected from medical records. In order to characterise the burden of coexisting diseases or conditions Charlson Comorbidity Index was calculated. The data obtained were processed with STATISTICA 10.0 using descriptive and nonparametric statistics.

Results: Among a total of 102 patients, a significant prevalence of comorbidities (86,27%) was found. 78,85% of all patients suffered from 2 or more coexistent conditions. The most prevalent comorbidities were: cardiovascular diseases