improved but C-reactive protein (CRP) remained in the same levels. On demand anakinra prevented progression of prodromes to full-blown attacks which was demonstrated by decrease in the rate of attack/prodrome ratio ($p=0.002$). On demand anakinra can be continued in 10 subjects on long-term but continuous treatment was required in 5 subjects.

Abstract THU0620 – Table 1. Comparison of attack characteristics before and after on demand anakinra protocol

	Colchicine period	Colchicine plus On-Demand Anakinra period	p
Attack severity, VAS	10 (2)	6 (3)	0.002
Duration, days	3 (2)	1.5 (1)	0.001
Frequency*, number	4 (1.5)	1.5 (1.75)	0.001
CRP (mg/L)	5.1 (6.1)	4.1 (5.9)	0.58
AIDAI	18 (22.5)	4 (5)	0.001
Attack/prodrom ratio, (n=10)	1	0.6 (0.4)	0.002
Absenteeism, days	7 (8)	2 (2.5)	0.001
Presenteeism, days	9 (7.5)	2.5 (3)	0.002

Attack frequency and work productivity parameters are adjusted for 3 months intervals. VAS: visual analogue scale, CRP: C-reactive protein, AIDAI: autoinflammatory disease activity index

Conclusions: On demand anakinra significantly improved FMF attacks which suggest this approach would be of benefit in daily practice in selected patients.

Disclosure of Interest: None declared

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THU0621

PERSISTENT PRURITIC SKIN LESIONS WITH DYSKERATOTIC CELLS IN UPPER LAYER OF EPIDERMIS ARE SPECIFIC AND ASSOCIATED WITH HIGH LEVELS OF SERUM IL-18 IN ADULT-ONSET STILL'S DISEASE

H. Nishikawa¹, Y. Taniguchi¹, N. Maeda-Aoyama¹, K. Nakajima², S. Inotani¹, Y. Shimamura¹, K. Inoue¹, K. Arai³, S. Sano², Y. Terada¹. ¹Nephrology and Rheumatology, ²Dermatology, Kochi Medical School Hospital, Nankoku; ³Internal Medicine, Kochi Red Cross Hospital, Kochi, Japan

Background: Adult-onset Still's disease (AOSD) is an acute and systemic inflammatory disorder that is characterised by high spiking fever, evanescent rash, arthralgia/arthritis and hyperferritinemia. However, recent reports showed that not only typical evanescent salmon-coloured rash but also atypical skin lesions, persistent pruritic papules and plaques, could be associated with AOSD.

Objectives: To assess the clinical significance of dyskeratotic cells (DCs) in skin lesions of AOSD.

Methods: We assessed histology of skin lesions including persistent pruritic skin lesions in Japanese patients with AOSD (n=15). Moreover, we compared histology of AOSD with dermatomyositis (DM) (n=6), drug eruptions (DE) (n=7), and graft versus host disease (GVHD) (n=6).

Results: AOSD with persistent pruritic skin lesions (n=10) histologically showed DCs only in upper layer of epidermis and horny layer without inflammatory cells infiltrations, indicating dyskeratosis. AOSD with evanescent rash (n=5) histologically showed no DCs. DCs were positive by ssDNA staining, suggesting apoptotic cells. Serum IL-18 showed significantly higher in AOSD patients with dyskeratosis (n=10) than without dyskeratosis (n=5). In contrast to AOSD with DCs, the histology of DM, DE and GVHD demonstrated that DCs existed in all layers of epidermis with inflammatory cells infiltrations.

Conclusions: Persistent pruritic skin lesions in AOSD are specific by prominent epidermal apoptosis involving the upper layers of epidermis. Moreover, hyper IL-18 might be related with dyskeratosis.

Disclosure of Interest: None declared

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THU0622

HISTOPATHOLOGY AND EXPRESSIONS OF CHEMOKINES, CXCL10, CXCL13, AND CXCR3, AND AN ENDOGENOUS LIGAND S100A8/A9 IN LYMPH NODES OF PATIENTS WITH ADULT-ONSET STILL'S DISEASE

H.-A. Kim¹, J.H. Han². ¹Department of Rheumatology, ²Department of Pathology, Ajou University School of Medicine, Suwon, Korea, Republic of Ireland

Background: Adult-onset still's disease (AOSD) is a rare systemic inflammatory disease with several symptoms, such as a persistent high spiking fever, typical rash, and lymphadenopathy. Endogenous factors related to interleukin (IL)-1,

such as S100A8/A9 and several chemokines including CXCL10, CXCL13 and CXCR3, could play a potential role in the pathogenesis of AOSD.

Objectives: We aimed to find out typical histopathologic features, expressed pattern of chemokines in lymph nodes (LN) of AOSD patients.

Methods: Formalin-fixed paraffin-embedded excisional LN tissues from 48 AOSD patients and 6 nonspecific reactive hyperplasia were histologically reviewed. The immunohistochemical stain for CXCL10, CXCL13, CXCR3 and S100A8/A9 were done. The clinical and laboratory data of the patients who underwent LN biopsies were reviewed.

Results: The LN specimens were categorised according to four distinctive patterns: follicular (n=2, 4.2%), paracortical (n=19, 39.6%), diffuse (n=9, 18.8%) hyperplasia, or mixed pattern (n=18, 37.5%). The other examined histologic features were presence of necrosis, karyorrhexis, immunoblastic, histiocytic and vascular proliferation. Most of the cases were required to take into differential diagnosis such as dermatopathic lymphadenitis (n=16, 33.3%), lymphoma (n=11, 22.9%) and histiocytic necrotizing lymphadenitis (n=9, 18.8%). The expression of chemokines and S100A8/A9 were higher than that of nonspecific reactive hyperplasia. The expression of chemokines and S100A8/A9 were higher than that of nonspecific reactive hyperplasia.

Conclusions: Histopathologic findings of LN in AOSD patients are diverse enough to be included various differential diagnosis. Because the several chemokines and S100A8/A9 were more expressed in AOSD patients than those of reactive hyperplasia, they may serve as a pathogenesis of AOSD.

Disclosure of Interest: None declared

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THU0623

SERUM IGG4 LEVELS AT DIAGNOSIS CAN PREDICT THE OUTCOMES OF UNTREATED PATIENTS WITH IGG4-RELATED DISEASE: A RETROSPECTIVE STUDY

I. Mizushima, N. Suzuki, M. Yoshida, A. Takeji, T. Matsunaga, T. Zoshima, S. Hara, K. Ito, H. Fujii, K. Yamada, M. Kawano. Division of Rheumatology, Kanazawa University Hospital, Kanazawa, Kanazawa, Japan

Background: IgG4-related disease (IgG4-RD) is a recently recognised systemic fibro-inflammatory disorder that can affect many organs.¹ In IgG4-RD, spontaneous, or at least temporary, remissions without treatment have been reported, and watchful waiting may be appropriate in certain patients with asymptomatic and inactive disease.² However, the outcomes of patients with IgG4-RD who do not undergo treatment are still unclear.

Objectives: This study aimed to clarify the outcomes of untreated patients with IgG4-RD and the factors related to the outcomes.

Methods: We retrospectively reviewed the medical records of 107 patients with IgG4-RD, who were followed up for more than 6 months, at a single centre in Japan. Among them, 27 patients were followed up without treatment after the initial diagnosis. We compared the clinical features of these 27 patients with those of the 80 patients who underwent treatment. The outcomes of untreated patients were investigated, and logistic regression analysis was performed to assess factors related to the outcomes. Deterioration of IgG4-RD was defined as symptomatic, radiological, or functional exacerbation of the organ involved or new organ involvement.

Results: The patients comprised 73 men and 34 women (mean age 65.7 years). The follow-up periods were 7–252 (mean, 64.1) months, and the serum IgG4 levels at diagnosis were 10.7–3610 (mean, 706) mg/dL. The 27 untreated patients had significantly fewer affected organs (1.9 ± 1.2 vs 3.0 ± 1.6 , $p=0.001$), lower IgG4-RD responder index (10.8 ± 5.1 vs 13.8 ± 6.8 , $p=0.048$), and lower frequency of ophthalmic and renal parenchymal lesions (25.9% vs 53.8% , $p=0.015$, and 3.7% vs 26.3% , $p=0.012$, respectively) than did the 80 patients who underwent treatment. Of these 27 patients, 8 experienced deterioration of IgG4-RD 3–232 months (mean, 62.8) after the diagnosis. New organ involvement was observed in all 8 patients, 2 of whom concurrently suffered exacerbation of the organs involved. In age- and sex-adjusted logistic regression analysis, serum IgG4 elevation (per 100 mg/dL, odds ratio 1.194, 95% confidence interval 1.017–1.402, $p=0.030$) was the only significant factor related to deterioration of disease in untreated patients with IgG4-RD.

Conclusions: The present study suggests that serum IgG4 levels may be useful to predict the outcomes of untreated patients with IgG4-RD, who tend to have fewer affected organs and lower IgG4-RD responder index.

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THU0624 COLCHICINE: AN EFFECTIVE TREATMENT OPTION FOR UNCLASSIFIED AUTOINFLAMMATORY DISEASES IN CHILDREN

J.B. Kuemmerle-Deschner¹, A.-L. Schock¹, S. Hansmann¹, S.M. Benseler².¹Department of Pediatrics, Division of Rheumatology, University Hospital Tuebingen, Tuebingen, Germany; ²Rheumatology, Department of Paediatrics, Alberta Children's Hospital, University of Calgary, Calgary, Alberta, Canada

Background: Children and adults with clinically and genetically defined autoinflammatory diseases (AID) including CAPS, TRAPS and HIDS can receive expensive Interleukin-1 (IL-1) inhibitors in many countries around the world. However, patients suffering from unclassified autoinflammatory conditions characterised by recurrent fevers and organ dysfunction and the absence of a known pathogenic mutation commonly have no access to these treatment options.

Objectives: The aim of this study was to explore the efficacy and safety of colchicine treatment in children and adults with autoinflammatory diseases without pathogenic mutations.

Methods: Consecutive children and adults with autoinflammatory diseases without pathogenic mutations treated with colchicine were included in this single centre study and observed for a median of 12.94 months (range 1.25–66.73). Clinical features, autoinflammatory disease activity indices (AIDAI), inflammatory markers ESR, CRP, SAA and S100, frequency and duration of flares and physician global assessment of disease activity (VAS) were recorded serially and compared at baseline and while receiving Colchicine therapy.

Results: A total of 39 patients were included in the study. These were 16 girls and 23 boys, median age at start of colchicine therapy was 4 years (range 1–54). The diagnoses included PFAPA in 15, mutation-negative FMF in 11, autoinflammation with low-penetrance variants in nine (all *NLRP3*) and other unclassified AID in four patients. Recurrent fever was the leading symptom, mostly associated with arthralgia and myalgia. The mean disease activity decreased from 4.4 at baseline to 2.2 on colchicine. Mean SAA-levels decreased from 159 to 63.3 mg/L, CRP levels from 6.4 to 2.3 mg/dl. Flare frequency was reduced in 72% and remained unchanged in 28% of patients. Flare duration was reduced in 82%, unchanged in 14% and increased in only 4% of patients. Most common adverse events were abdominal pain and nausea in 50% of patients and appeared to be dose dependent.

Conclusions: Children and adults with unclassified autoinflammatory diseases may benefit significantly for colchicine therapy. Control of clinical disease activity and improved inflammatory markers were documented in 59% of patients. Colchicine should be considered in patients with active inflammatory disease with no access to IL-1 inhibitors. Controlled trials are needed to further explore this approach.

Disclosure of Interest: None declared

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THU0625 IGG4-RELATED DISEASE MANIFESTATIONS DIFFER BETWEEN ASIAN AND NON-ASIAN SUBJECTS

Z.S. Wallace¹, K. Okazaki², C. Perugino¹, H. Umehara³, T. Saeki⁴, M. Kawano⁵, Y. Zen⁶, R. Naden⁷, J.H. Stone¹, on behalf of For the EULAR/ACR IgG4-Related Disease Classification Criteria Development Group. ¹Massachusetts Gen Hosp Rheumatol Unit, Harvard Med School, Boston, USA; ²Kansai Medical University, Osaka; ³Shiritsu Nagahama Byoin, Nagahama; ⁴Nagaoka Red Cross Hospital, Nagaoka; ⁵Kanazawa University, Kanazawa; ⁶University of Kobe, Kobe, Japan; ⁷University of Hamilton, Hamilton, Canada

Background: IgG4-related disease (IgG4-RD) is a multi-system immune-mediated condition that can affect nearly any organ. No study has evaluated differences in disease manifestations according to race. We evaluated this in a large cohort of IgG4-RD subjects submitted by an international group of investigators.

Objectives: To evaluate racial differences in manifestations of IgG4-RD.

Methods: To validate the ACR/EULAR IgG4-RD Classification Criteria, 76 investigators from North America, South America, Europe, and Asia submitted cases they considered to be IgG4-RD in either the preliminary phase or the validation phase. For each case, investigators included details related to diagnostic certainty, age at disease onset and diagnosis, race, organ involvement, biopsy findings, and laboratory results. Based on reported race, we dichotomized subjects into either Asian or non-Asian categories; subjects of South Asian (n=14) descent (e.g., India, Pakistan), all of whom resided in North America or Europe were grouped with non-Asian subjects. We compared the distribution of disease features according to race using t-tests, Wilcoxon tests, and Chi square tests, where appropriate, as well as in multivariable-adjusted models.

Results: In the validation phase, there were 493 cases of IgG4-RD submitted by 23 investigators who practice in Asia and 29 investigators who practice in North America or Europe. There was no significant difference in the distribution of specialists (e.g., rheumatology, gastroenterology) between Asian and non-Asian investigators (p=0.3). The majority of IgG4-RD subjects, both Asian (n=208) and non-Asian (n=285), were male (61% and 69%, respectively). Asian subjects were significantly older both at symptom onset and diagnosis (61.2±13.2 years and 62.6±12.8 years, respectively) compared to non-Asian subjects (55.1±14.9 years and 57.2±14.4 years, respectively, p<0.0001 for both comparisons). There was a significantly shorter diagnostic delay among Asian subjects compared to non-Asian subjects (1.4±2.7 years vs 2.2±3.7 years, p=0.01). Head/neck involvement was more common in Asians (52% vs 27%, p<0.0001) whereas hepatobiliary involvement was more common in non-Asians (52% vs. 42%, p=0.04). Asian subjects had a significantly higher median serum IgG4 concentration (666 mg/dL, IQR 320.5–1230 vs 240.5, IQR 100–505, p<0.0001) and were more likely to have a serum IgG4 concentration greater than five times the upper limit of normal (48% vs 20%, p<0.0001). In multivariable-adjusted models, differences in age and serum IgG4 concentration according to race remained strongly significant (p<0.001 for both comparisons).

Conclusions: Asian and non-Asian subjects differed regarding the age of disease onset and diagnosis, the distribution of organ involvement, and baseline serum IgG4 concentrations. There was a significantly shorter diagnostic delay among Asian subjects compared to non-Asian subjects. The etiology(ies) of these observed differences in the respective presentation of IgG4-RD in Asian and non-Asian subjects requires further investigation, but could include differences in diagnostic approach, environmental factors, and genetic predisposition.

Disclosure of Interest: None declared

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THU0626 CORRELATION AMONG SERUM AMYLOID A LEVELS, CLINICAL MANIFESTATIONS, TREATMENT AND DISEASE ACTIVITY IN PATIENTS WITH BEHÇET'S DISEASE

J. Sota¹, A. Vitale¹, O.M. Lucherini¹, R. Franceschini¹, B. Frediani¹, I. Orlando¹, M. Galeazzi¹, C. Fabiani², G.M. Tosi¹, L. Cantarini¹. ¹University of Siena, Siena; ²Humanitas Clinical and Research Center, Milan, Italy

Background: Behçet's disease (BD) is an inflammatory disorder potentially leading to life- and sight-threatening complications: no laboratory marker correlating with disease activity or predicting the occurrence of disease manifestations is currently available in the clinical practice.

Objectives: To search for a correlation between serum amyloid-A (SAA) levels and disease activity evaluated via BD current activity form (BDCAF), to assess disease activity in relationship with different SAA thresholds, to examine the association between single organ involvements and the overall major organ involvement with different SAA thresholds, and to assess the influence of biologic therapy on SAA levels.

Methods: Ninety-five serum samples were collected from 64 BD patients, and their related demographic, clinical and therapeutic data were retrospectively collected.

Results: No correlation was identified between SAA levels and BD disease activity (Spearman ρ =0.085; p=0.411), while a significant difference was found in the mean BDCAF score between patients presenting SAA levels<200 mg/L and those with SAA levels>200 mg/L (p=0.027). SAA levels higher than 200 mg/L were significantly associated with major organ involvement (p=0.008). A significant association was found between SAA levels>150 mg/dl and ocular (p=0.008), skin (p=0.002) and mucosal manifestations (p=0.012). Patients undergoing biologic therapies were significantly associated with SAA levels<200 mg/L compared with patients who were not treated with biologics (p=0.012).

Conclusions: SAA level does not represent *per se* a biomarker of disease activity, but might be useful as a predictor of major organ involvement and ocular disease relapse at certain thresholds in patients with BD.

Disclosure of Interest: None declared

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THU0627 DEMOGRAPHICS AND PRESENTING ORGAN INVOLVEMENT IN A COHORT OF PATIENTS WITH SARCOIDOSIS

K. Sheth¹, G. Jeyashanmugaraja², J. Simard¹, S. Shoor¹. ¹Stanford University, Palo Alto, USA; ²Stanley Medical College, Chennai, India

Background: Sarcoidosis is a multisystem disorder of unknown etiology characterised pathologically by non-caseating granulomas in involved organs. Although mortality is reported in only 1%–5% of patients, there is data suggesting it might