DOI: 10.1136/annrheumdis-2018-eular.4040

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

DOI: 10.1136/annrheumdis-2018-eular.7473

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

Z.S. Wallace¹, K. Okazaki², C. Perugini¹, 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

DOI: 10.1136/annrheumdis-2018-eular.5245

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

DOI: 10.1136/annrheumdis-2018-eular.1178

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