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

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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

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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

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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

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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

be increasing and there is little information about the clinical and epidemiological characteristics of this group of patients.

Objectives: To compare basic demographics of a cohort of deceased sarcoid patients with a non-deceased sarcoid population, and to describe the presenting organ involvement among the deceased sarcoid population.

Methods: Patients seen at the Stanford University Hospital and Clinics from 2009–2017 who were >18 years of age with at least one ICD-9 or 10 diagnosis of sarcoidosis and at least one clinical note with the term "sarcoidosis" were identified by EMR. After determining the demographics of this cohort, deceased patients were identified and an extensive chart review of these subset of patients was conducted. The project was approved by Stanford's Institutional Review Board.

Results: 1190 adult patients with an ICD code for sarcoidosis were identified. Demographic comparison between the non-deceased and deceased population is summarised in the table 1 below.

	Non-deceased sarcoid patients	Deceased sarcoid patients
Total patients	1086	104
Gender:	660 (60.8%)	59 (56.7%)
Females	426 (39.2%)	45 (43.3%)
Males		
Race:	67 (6.2%)	1 (0.9%)
Asian	197 (18.1%)	26 (25%)
Black	596 (54.9%)	45 (43.2%)
White	6 (0.6%)	0 (0%)
Native American	128 (11.8%)	18 (17.4%)
Others	91 (8.4%)	14 (13.5%)
Unknown		
Ethnicity	918 (84.5%)	80 (76.9%)
Non-Hispanic	67 (6.2%)	11 (10.5%)
Hispanic/Latino	101 (9.3%)	13 (12.6%)
Unknown		

In comparison with non-deceased patients with sarcoidosis, the proportion of African American population was higher in the deceased population, whereas the proportion of Asians and whites was lower in the deceased population. The percentage of male patients in the deceased subset was slightly higher than the non-deceased population. 65 (62.5%) of the 104 deceased patients were symptomatic at diagnosis, 6 (5.8%) were asymptomatic and information was not available for 33 (31.7%) patients. Of the 65 symptomatic patients, presenting organ involvement was either pulmonary (34, 52.3%), cardiac (11, 16.9%), cutaneous (8, 12.3%), neurological (4, 6.2%), ocular (3, 4.6%), constitutional (5, 7.7%), unknown (1, 1.5%), musculoskeletal (1, 1.5%), renal–calcium related (1, 1.5%), hepatic (2, 3%), laryngeal (1, 1.5%), nasal and sinus (2, 3%), parotid gland (1, 1.5%), gastrointestinal (1, 1.5%), or others (5, 7.6%).

Conclusions: This study compares the demographics of subset of deceased sarcoid patients with a population of non-deceased sarcoid patients, and describes the presenting organ involvement among the deceased sarcoid population. African American males had a higher mortality rate compared to Asians and Caucasians. Pulmonary and cardiac disease represented 69.2% of organ involvement in the deceased. Our study showed a mortality rate of 8.7% in the sarcoid population which is in keeping with the newer studies that mortality in patients with sarcoid might be on the rise. With further study of this population, we aim to compare the characteristics of non-deceased sarcoid patients with the deceased patients, including presenting organ involvement, diagnostic workup and treatment regimen and hope to gain a better understanding of the possible reasons for increasing mortality in patients with sarcoidosis.

Disclosure of Interest: None declared

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THU0628 DIFFERENCES IN CLINICAL MANIFESTATIONS OF BEHCET'S SYNDROME BY GENDER: CROSS-SECTIONAL ANALYSIS IN A UK COHORT

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Background: Geographical variations in genotype and phenotype of Behçet's syndrome (BS) are reported.^{1,2} Previous meta-analysis of international cohorts has highlighted differences in clinical manifestations of BS by gender.³ A comparison of BS clinical manifestations by gender has not been analysed previously in a large UK BS cohort.

Objectives: 1) To compare the clinical manifestations of BS in a UK cohort by gender and 2) to compare this to published international data.

Methods: A retrospective cross sectional analysis was performed using clinical databases at the Liverpool and Birmingham BS Centres of Excellence. Patients with a multi-disciplinary diagnosis of BS or International Study Group (ISG) diagnostic criteria were included. Clinical manifestations and HLA-B51 positivity were compared by gender. T-tests and chi-squared tests were used for continuous and categorical data respectively and a p value of 0.05 or less was considered statistically significant.

Results: 433 patients met inclusion criteria (140 males, 32.3%). Male patients were younger and had significantly higher rates of ocular and vascular involvement, papulopustular skin rash and HLA-B51 positivity. Female patients had significantly higher rates of genital aphthosis and arthralgia.

	Male (n=140)	Female (n=293)	p value
Age (years), mean (s.d.)*	41.2 (12.5)	44.1 (11.9)	0.02
Recurrent oral aphthosis, n (%)	139 (99.3)	291 (99.3)	0.97
Genital aphthosis, n (%)	111 (79.3)	278 (94.9)	<0.01
Papulopustular skin rash, n (%)	69 (49.3)	107 (36.5)	0.01
Erythema nodosum, n (%)	29 (20.7)	56 (19.1)	0.70
Skin aphthosis (n,%)	7 (5.0)	8 (2.7)	0.23
Uveitis, n (%)	81 (57.9)	99 (33.8)	<0.01
Retinal vasculitis, n (%)	39 (27.9)	36 (12.3)	<0.01
Central nervous system involvement, n (%)	16 (11.4)	20 (6.8)	0.11
Large vein thrombosis, n (%)	22 (15.7)	19 (6.5)	<0.01
Arterial thrombosis or aneurysm, n (%)	4 (2.9)	7 (2.4)	0.77
Thrombophlebitis, n (%)	10 (7.1)	5 (1.7)	<0.01
Gastro-intestinal involvement, n (%)	8 (5.7)	25 (8.5)	0.30
Arthralgia, n (%)	68 (48.6)	180 (61.4)	0.01
Ethnicity (British or White), n (%)	90 (64.3)	238 (85.9)	<0.01
	(n=126)	(n=277)	
HLA-B51 positive, n (%)	14 (58.3)	9 (20.0)	<0.01
	(n=27)	(n=45)	

*statistically significant

Conclusions: Similar to published international cohorts, male patients with BS in the UK have higher rates of ocular and venous involvement, papulopustular skin rash and HLA-B51 positivity and lower rates of genital aphthosis and arthralgia in comparison to female patients. However, the extent to which these differences are attributable to ethnicity is unclear.

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THU0629 A STUDY OF WBMRI IN ESTIMATING DISEASE ACTIVITY OF PM/DM AND THE FOLLOWING DISEASES DIAGNOSIS

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Background: IM is a group of disease characterised by chronic symmetrical myasthenia, muscle fatigue and monocyte infiltrating skeletal muscle. This disease usually associates with the muscle of trunk and proximal limbs, skin lesions accompanied or not. Dermatomyositis (DM) and polymyositis (PM) account for the major part of IM clinically. The diagnosis of these diseases mainly rely on clinical manifestation, biopsy, ECG and serum creatase. Whole-body magnetic resonance imaging (WBMRI) has been reported succeeding in diagnosing PM/DM through short tau inversion recovery (STIR) sequence, but the ability to estimate disease activity hasn't been reported.

Objectives: To evaluate the value in WBMRI through STIR sequence in evaluating the disease activity of polymyositis (PM) and dermatomyositis (DM) and screening interstitial lung disease (ILD) and osteonecrosis.