**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 physical 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 1–18). The diagnoses included PFAPA in 15, mutation-negative FMF in 11, autoinflammatory disease with low-penetrance variants in nine (all NLRP3) 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 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 mild to moderate gastrointestinal side effects and in 50% of patients and appeared to be dose dependent.

**Conclusions:** Children and adults with unclassified autoinflammatory diseases may benefit significantly from 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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**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, 176 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:** 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±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 hepato-biliary 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:** 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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**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...