**THU0624**

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


Department of Pediatrics, Division of Rheumatology, University Hospital Tuen-bingen, Tuen-bingen, 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 center 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 NLPR3) 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.

Disclosure of Interest: None declared


**THU0625**

**IGG4-RELATED DISEASE MANIFESTATIONS BETWEEN ASIAN AND NON-ASIAN SUBJECTS**

Z.S. Wallace1, K. Okazaki2, C. Perugino3, H. Umehara3, T. Saeidi4, M. Kawano5, Y. Zen6, R. Naden7, J.H. Stone8, 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; 3Shinshu Nagahama Byoin, Nagahama; 4Nagaoka Red Cross Hospital, Nagaoka; 5Kanazawa University, Kanazawa; 6University of Kobe, Kobe, Japan; 7University of Hamilton, Hamilton, Canada

Background: 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 in this large cohort of IgG4-RD subjects submitted by an international group of investigators.

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

Methods: Methods: To validate the ACR/EULAR IgG4-RD Classification Criteria, we collected diagnostic data from North America, South America, Europe, and Asia and 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 age, sex, and female sex.

Results: None declared

Disclosure of Interest: None declared


**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, G.M. Toz1, 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 (BDCF), 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’s rho=-0.085, p=0.411), while a significant difference was found in the mean BDCF 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 biological 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


**THU0627**

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

K. Sheth1, G. Jayasahmugaranra2, J. Simard3, S. Shor1, 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...