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, autoinflammatory diseases 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 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


CORRELATION AMONG SERUM AMYLOID A LEVELS, CLINICAL MANIFESTATIONS, TREATMENT AND DISEASE ACTIVITY IN PATIENTS WITH BEHCET’S DISEASE

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Background: Behcet’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 therapies 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 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.

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DEMographics and Presenting Organ INVOLVEMENT in a COrHort 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