different haematological and biochemical variables are significantly milder in patients on biological therapy, which may limit the utility of classification criteria in this group. Assessment of these findings in larger cohorts is needed.

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Other orphan diseases

THU0554 THE CLINICAL UTILITY OF TWO VASCULITIS ACTIVITY SCORES(BVAS AND BDCAF) IN BEHÇET'S SYNDROME: A PROSPECTIVE COHORT STUDY

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Background: Behçet's Disease(BD) is a rare chronic autoinflammatory condition that can lead to irreversible organ damage. The potential for multi-organ involvement and fluctuating activity highlights the need to perform a careful and systematic assessment of disease activity that is sensitive to change. Several disease activity tools have been used in both daily practice and clinical trials, yet there is no published data comparing the clinical utility of different tools in informing changes to therapy.

Objectives: To compare the utility of two major activity scores:BD Current Activity Form (BDCAF2006)¹ and Birmingham Vasculitis Activity Score (BVAS)² in predicting physician's decision to adjust treatment (step-up/step-down) in patients with BD.

Methods: A 6-month prospective observational study was performed in a cohort of patients meeting the International Criteria for Behçet's Disease (ICBD),at the National Centre for BD in Liverpool,UK. Participants were described for their demographics, clinical manifestations and treatment plan.BVAS and BDCAF2006 activity scores were completed for each patient at evaluation.The outcome of interest was treatment change which was classified as 'step-up' or 'step-down', reflecting escalation or de-escalation in treatment (dosage adjustment or adding new immunosuppressant), respectively. We assessed the association between BVAS and BDCAF scores and step-up/step-down treatment using Spearman rank correlation and multivariate logistic regressions, adjusting for gender, age and patient's perception of disease activity on visual analogue scale (VAS).Odds ratios(OR) and 95% confidence intervals were calculated. Data analysis was conducted in Microsoft Excel,SPSS 2.0 and STATA.

Results: Ninety-five patients met inclusion criteria: 25 males(26.3%) and 70females(73.7%) with a mean age at diagnosis of 32.7years(±11.3 SD). HLAB51 was positive in 11/51 cases(11.6%).The most frequent clinical manifestations were oral ulcerations(100%), genital ulcerations(94.7%) followed by papulo-pustular skin lesions(37.8%) arthralgia(31.6%)and headache(30.5%).

Mean BVAS score(range 0-6) was 2.14(±1.8 SD) and mean BDCAF score (range 0-8) was 3.04(±1.72 SD). Both BVAS and BDCAF correlated with decision to step-up treatment ($r=0.752$; $r=0.370$, respectively). Furthermore, BVAS was more strongly associated with decision to step-up treatment than BDCAF(OR 4.25 95%CI 2.37 to 7.61; 1.51 95%CI 1.15 to 2.00, respectively). Adjusting for gender, a stronger association was observed in male participants across BVAS and BDCAF scores(OR 5.89 95%CI 1.17 to 29.63; 3.48 95%CI 1.20 to10.09,respectively). Following adjustment for patient's perception of their disease (VAS), BVAS remained significantly associated with treatment step-up(OR 3.87 95%CI 2.08 to7.19) but not BDCAF(OR 1.30 95%CI 0.91 to1.84).

Regarding different clinical manifestations,the BVAS mucocutaneous and ocular activity showed a significant odds ratio for step-up therapy (OR=5.78, CI:1.49-22.15; and OR=4.2,CI: 2.26-7.83).

Conclusion: BVAS can be a useful tool to assess BD activity. In this study, BVAS correlated better with clinical treatment decisions than BDCAF, particularly in male participants. It also appears to be less influenced by patient's subjective perception of disease activity, and therefore may be a more objective measure of BD activity.

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THU0555 10 YEAR PROGNOSIS OF PATIENTS DIAGNOSED WITH FAMILIAL MEDITERRANEAN FEVER

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Background: In Familial Mediterranean Fever (FMF), other than amyloidosis factors affecting mortality are being debated. In our previous study, we did not observe any atherosclerotic plaque formation in carotid or femoral artery. We thought that the risk of atherosclerosis did not increase in patients diagnosed with FMF.

Objectives: The aim of this study was to assess the 10 year prognosis and comorbidity of patients diagnosed with FMF who have been treated in our rheumatology clinic.

Methods: The sample group is a subset of 2009 study. In 2009, the patients who already had myocardial infarction or cancer diagnosis were excluded. The patients were interviewed with polar questions of whether they were diagnosed with acute myocardial infarction (AMI), cerebrovascular events, cancer, diabetes, and hypertension.

Results: We studied 71 patients (37 males, 34 females; mean age: 49.66±6.91) with FMF, and 59 patients (24 males, 35 females) in healthy control (HC) group. The gender and age difference between two groups was not found significant.

During 10 year follow-up, 8% of FMF patients had either a cardiovascular or cerebrovascular event comparing to 5% in HC ($p>0.05$). 3% of FMF patients had a cancer diagnosis comparing to 3% in HC ($p>0.05$). Even though diabetes mellitus diagnosis rate was higher in FMF patients (15% to 10%), results were still not significant ($p>0.05$). Hypertension diagnosis was 5% higher in FMF group ($p<0.05$)

Table. Prognostic Factors of FMF patients compared with Healthy Controls

	FMF 2018, n (%)	HC 2018, n (%)	p value
Female	34 (47.89)	35 (59)	0,193
Age	49±6.91	51±5.59	0,076
AMI/Stroke	6 (8.45)	3 (5.08)	0,45
Cancer	2 (2.82)	2 (3.39)	0,85
DM	9 (14.86)	6 (10.17)	0,198
Hypertension	25 (33.78)	10 (16.95)	0,019*
Total	71	59	

Conclusion: Even though there was a significant increase in hypertension, increased diabetes, cancer, and AMI/Stroke ratio was not found significant when compared to the HCs. Therefore, any cardiovascular and malignancy related comorbidities are not associated with FMF.

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