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Disclosure of Interests: None declared

Figure 2: A. Long-term mortality in patients with TAK B. Relapse-free survival in patients with TAK at 7-year follow-up.

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AB0536

EFFECT OF HYDROXYCHLOROQUINE TREATMENT IN MUCOCUTANEOUS MANIFESTATIONS IN PATIENTS WITH BEHÇET'S SYNDROME

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Background: Behçet syndrome (BS) is a rare multisystemic vasculitis, most commonly seen in regions along the ancient Silk Road. It runs a relapsing and remitting course. Mucocutaneous disease, consisting of oral ulcers, genital ulcers and skin lesions is often reported. EULAR recommendations advise colchicine and topical agents for the treatment of these lesions.¹ Not all patients respond adequately, thus, it is important to explore alternative treatment options.

Objectives: To study the efficacy of hydroxychloroquine (HCQ) 400mg daily in patients with mucocutaneous BS.

Methods: Data on all patients who presented at the outpatient Behçet clinic in New York were recorded. Patients with a first prescription with HCQ and a follow-up of 3 months (range: 2.75-41.2 months) were included. Patient reported outcomes BSAS and RAPID3 were used to evaluate the effect of HCQ. Results of all patients and of International Study Group (ISG) positive patients were analyzed separately using Wilcoxon rank tests.

Results: We included 94 patients with a first prescription of HCQ. 72 patients (76.6%) fulfilled ISG criteria. Mean age was 36.1 years (SD 12.5), 76 patients (80.9%) were female and 11 patients (11.7%) were from Silk Road countries.

Mean duration until follow-up was 6.5 months (SD 5.7). Median BSAS scores in ISG+ patients at baseline did not differ significantly from ISG- patients, except for skin lesions (5.0 in ISG+ vs. 0.5 in ISG- $p=0.005$). BSAS scores at follow-up did not differ significantly (ISG+ vs. ISG-).

Median BSAS scores were significantly lower at follow-up compared to baseline for oral ulcers ($p=0.010$), skin lesions ($p=0.018$) and overall activity ($p=0.019$). Regarding genital ulcers there was no significant result, due to only 37 patients

Table 1. Median BSAS scores of patients treated with HCQ.

	Baseline (median, IQR)	Follow-up 3 months (median, IQR)	P-values
All patients (n=94)			
Oral ulcers	5.0 (2.00-7.88)	3.0 (1.00-6.00)	0.010
Genital ulcers	0.0 (0.00-3.88)	0.0 (0.00-3.00)	0.371
Skin lesions	5.0 (1.25-7.00)	2.5 (0.00-7.00)	0.018
Overall activity	5.5 (4.00-8.00)	5.0 (2.00-7.25)	0.019
ISG+ patients (n=72)			
Oral ulcers	5.25 (2.00-7.63)	3.25 (1.00-6.00)	0.007
Genital ulcers	0.5 (0.00-4.00)	0.0 (0.00-3.00)	0.684
Skin lesions	5.0 (2.00-7.13)	3.0 (0.00-7.00)	0.015
Overall activity	6.0 (4.00-8.00)	5.0 (2.00-7.50)	0.057

reporting complaints of genital ulcers. Performing these analyses in ISG+ patients only did not change these results, except for BSAS overall activity, which lost significance ($p=0.057$).

RAPID3 scores were not statistically different between baseline and follow-up (9.67 vs. 8.75, $p=0.145$), nor were its separate components function ($p=0.67$ vs. 0.67, 0.713), pain (4.0 vs. 4.0, $p=0.157$) and patient global (5.0 vs. 4.5, $p=0.095$).

The majority of patients used prednisone at baseline (58.5%) and at follow-up (57.4%). In 15 patients, prednisone was stopped at follow up, in 13 patients it was started.

Conclusion: HCQ improves median BSAS scores for oral ulcers, skin lesions and overall activity at 3 months follow-up compared to baseline. These results were similar in ISG+ patients (except for overall activity). Additional research is needed to assess the effect of HCQ in more patients and over multiple time points.

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AB0537

DIAGNOSTIC ACCURACY OF SYMPTOMS AND SIGNS FOR GIANT CELL ARTERITIS: SYSTEMATIC REVIEW AND META-ANALYSIS

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Background: Making a correct diagnosis of giant cell arteritis (GCA) is critical given the potential complications of the disease and its therapy. Estimation of the clinical probability of GCA is challenging. Prediction models might be helpful, but have methodological and practical drawbacks. One earlier meta-analysis described the diagnostic accuracy of symptoms and signs for a positive temporal artery biopsy [1]. In the latter study, the diagnostic accuracy of symptoms and signs might have been overestimated due to inclusion of case-control studies.

Objectives: To evaluate the diagnostic accuracy of symptoms and signs for GCA. **Methods:** PubMed, EMBASE and the Cochrane Database were searched for relevant studies. Studies were eligible if: all patients were suspected of having GCA; either a temporal artery biopsy (TAB), imaging test, or clinical diagnosis was used as reference standard for GCA; a 2x2 table was available for at least one symptom, physical sign or routine laboratory test (i.e. the index tests). Case reports and case-control studies were excluded. The screening, full text review, quality assessment with QUADAS-2 tool and data extraction were performed by two investigators. Hierarchical logistic regression modelling provided the pooled estimates of likelihood ratios with their 95% confidence intervals. Likelihood ratios <0.5 (i.e. making GCA less likely) or >2.0 (i.e. making GCA more likely) were considered diagnostically relevant.

Results: Out of 1359 reports screened, 59 studies were included in the study. These reports contained 13406 patients, including 3940 GCA patients. Most studies were retrospective, performed at tertiary centres and published after the prior meta-analysis [1]. TAB was the reference standard in 36 studies and a clinical diagnosis in 23 studies. Quality assessment revealed substantial risk of selection bias in the majority of studies. Studies using a clinical diagnosis were at risk of bias, as the reference standard might be influenced by the index tests (e.g. symptoms). Jaw claudication, limb claudication, temporal tenderness, temporal artery abnormalities, especially arterial thickening or loss of pulse, and anterior ischemic optic neuropathy provided a positive likelihood ratio >2.0. None of the 19 symptoms and 7 physical signs evaluated, provided a negative likelihood ratio <0.5. An ESR >60mm/hr and platelet count >400*10⁹/L provided a positive likelihood ratio of >2.0. Absence of an elevated CRP level or ESR >60mm/hr yielded a negative likelihood ratio of <0.5. Studies using a clinical diagnosis as reference standard reported higher positive likelihood ratios for jaw claudication, weight loss and PMR when compared to TAB studies. Absence of an elevated CRP provided a lower negative likelihood ratio in studies using the clinical diagnosis.

Conclusion: Few clinical findings may help to estimate the clinical probability of GCA. The presence or absence of any particular symptom or sign does not sufficiently rule out or rule in GCA. These findings highlight the need of additional tests (i.e. imaging, biopsy) in patients suspected of having GCA.