Objectives: To analyze whether benefit in SF-36 MCS was maintained in patients originally assigned to TCZ compared with those originally assigned to placebo (PBO) plus a 28- or 52-week prednisone taper among patients who achieved clinical remission at week 52 and maintained treatment-free clinical remission in the 2-year, long-term extension of GIACTA.

Methods: At the end of part 1, patients entered open-label part 2, in which GCA therapy (including initiation/termination of open-label TCZ and/or GCs) was given at the investigator’s discretion according to disease status. Change from baseline in SF-36 MCS score was compared for combined original TCZ (n = 33) and PBO (n = 17) patients who achieved clinical remission at week 52 and maintained treatment-free (no TCZ or GCs) clinical remission in part 2 using a repeated-measures model. The minimal clinically important difference (MCID) for SF-36 MCS is >2.5.7

Results: During treatment, SF-36 MCS scores in all 50 patients who maintained treatment-free clinical remission in part 2 had diverged between the TCZ and PBO groups as early as 36 weeks after baseline, with greater improvements evident in the TCZ group (Figure). The difference in least square means (LSM) change between TCZ and PBO was statistically significant at week 52 (p = 0.016) and maintained at weeks 100 (p = 0.023) and 156 (p = 0.002). The LSM difference (95% CI) between TCZ and PBO at weeks 52, 100, and 156 was 5.6 (1.1-10.2), 6.5 (0.9-12.1), and 7.4 (2.9-11.9), respectively, exceeding the MCID.

Conclusion: Among patients who maintained treatment-free clinical remission during part 2 of GIACTA, those originally assigned to receive TCZ prednisone taper plus a prednisone taper during part 1 maintained statistically significant and clinically meaningful improvements in SF-36 MCS up to week 156 compared with those originally assigned to receive PBO prednisone taper in part 1. This was true even though neither of the patient groups received TCZ or GC treatment after they achieved clinical remission at week 52.

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SAT0276 CARDIAC MAGNETIC RESONANCE IMAGING IN PATIENTS WITH BEHÇET’S DISEASE: A PILOT STUDY
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Background: Behçet’s disease is a multisystemic vasculitis characterized by mucocutaneous, ocular, arthritic, and vascular manifestations.[1] However, the incidence and nature of cardiac involvement in Behçet’s disease have not been clearly documented yet.

Objectives: This study aimed to assess the cardiac involvement in patients with Behçet’s disease using cardiac magnetic resonance imaging (MRI).

Methods: This cross-sectional descriptive study was carried out on thirty consecutive patients with Behçet’s disease (21 males, 9 females) with mean age 32.3±8.9 years and with no evidence of cardiac disease. They underwent cardiac MRI to determine morphological and functional changes of the heart and to detect areas of hyperenhancement after IV administration of gadolinium.

Results: At least one abnormality on cardiac MRI was observed in 20/30 patients (66.67%). Myocardial oedema was observed in 3 patients (10.0%) and late gadolinium enhancement in only 1 patient (3.3%). Pericardial effusion was found in 3 patients (10.0%), global hypokinesia in 6 patients (20.0%) and intra-cardiac thrombosis in only 1 patient (3.3%). Pulmonary artery was dilated in 4 patients (13.3%). Left ventricular and right ventricular end diastolic volume were altered in 4 patients (13.3%) and 7 patients (23.3%) respectively. Also, left ventricular and right ventricular end systolic volume were abnormal in 27 patients (23.3%) and 5 patients (16.7%) respectively. Moreover, there was aortic valve regurgite in 2 patients (6.7%), tricuspid valve regurgite in 9 patients (30%), and mitral valve regurgite in 9 patients (30%). Dilated left main coronary artery in 2 patients (6.7%) and arythmogenic right ventricular dysplasia in only one patient 1 patient (3.3%).

Conclusion: Behçet’s disease may cause cardiac abnormalities without clinical manifestations and cardiac MRI may represent a tool for early detection of these subtle abnormalities.

References:
[1] Geni, G., et al., Spectrum of cardiac lesions in Behçet disease: a series of 52 patients and review of the literature. Medicine, 2012. 91(1): p. 25-34.

Figure 1. Cardiac MRI in 32 year-old female patient with Behçet’s disease for 3 years.

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SAT0277 HEAD AND NECK INVOLVEMENT OF IGA VASCULITIS: A CASE-CONTROL STUDY
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Background: IgA vasculitis (IgAV) is an immune-complex mediated, small-vessel vasculitis which predominantly involves the skin on the lower extremities. Head and neck involvement is rarely reported.

Objectives: To describe the presentation and outcome of a series of patients with head and/or neck involvement in comparison to patients with cutaneous findings isolated to the lower extremities.
Results: Baseline characteristics, laboratory parameters, treatments and outcome were compared between cases and controls. The cutaneous features were isolated to the waistline or distal lower extremities of IgAV (case) was matched to two age- and sex-matched control patients with IgAV. In this cohort, patients with documented head/neck (H/N) involvement were identified through direct medical chart review. Among this cohort, patients with documented H/N-IgAV involvement included facial (cheeks, forehead) (n=6), perioral/oral/lip (n=5), auricular (n=2), nasal (n=2), and neck (n=1). All patients in both groups had evidence of purpuric skin lesions. Patients with H/N-IgAV involvement more frequently had evidence of skin ulcerations (23% vs. 0%; p=0.01) [Figure 1]. Overall baseline renal involvement and microscopic hematuria were less commonly observed in patients with H/N-IgAV. Among H/N-IgAV cases, at last follow-up all had resolution of H/N lesions but 3 of 13 had persistent skin lesions on the lower extremities despite ongoing treatment. Long-term outcome between cases and controls did not identify any significant differences in the development of end-stage renal disease, time to resolution of hematuria or proteinuria, time to complete IgAV response, or time to first IgAV relapse.

Conclusion: This study reports the largest series of patients with head/neck involvement of IgA, a rarely reported entity. In this cohort, patients with H/N-IgAV had less frequent renal involvement compared to IgAV patients with lower extremity only skin lesions. Clinicians should be aware of atypical locations of IgAV involvement. Additional research is needed to further understand this clinical subset.

Table 1. Baseline characteristics of patients with head and neck involvement of IgA-vasculitis compared to those with lower extremity only

| Characteristic, n (%) | H/N-IgAV (N=13) | LE-IgAV (N=26) | p-value |
|-----------------------|-----------------|----------------|---------|
| Age at diagnosis, years* | 38 (24) | 38 (24) | 1.0 |
| Male                  | 7 (54%) | 16 (62%) | 0.65 |
| Caucasian             | 13 (100%) | 24 (96%) | 0.47 |
| Length of follow-up, years* | 2.5 (3.5) | 13 (2.0) | 0.28 |
| Body mass index, kg/m²* | 32 (17) | 28 (10) | 0.72 |
| Hypertension          | 2 (15%) | 8 (31%) | 0.30 |
| Infection within 4 weeks | 7 (54%) | 11 (42%) | 0.50 |
| Antibiotic exposure within 4 weeks | 4 (31%) | 6 (24%) | 0.65 |
| Abdominal ischemic symptoms | 2 (15%) | 3 (12%) | 0.74 |
| Palpable purpura       | 13 (100%) | 26 (100%) | --- |
| Skin ulceration        | 3 (23%) | 0 (0%) | 0.01 |
| Any renal involvement  | 5 (38%) | 19 (73%) | 0.04 |
| Microscopic hematuria  | 4 (31%) | 17 (65%) | 0.04 |
| Proteinuria            | 4 (31%) | 15 (58%) | 0.11 |
| C-reactive protein, mg/L* | 28 (25) | 22 (22) | 0.61 |
| Erythrocyte sedimentation rate, mm/hr* | 20 (21) | 27 (25) | 0.80 |
| eGFR (ml/min/1.73m²)*   | 87 (41) | 95 (42) | 0.88 |

*mean (±standard deviation); H/N, head and/or neck; LE, lower extremity only

Figure 1. A) H/N involvement in IgAV (perioral, nasal, cheeks and neck). B) Severe PP in lower extremities with bullous, ulcerations and skin necrosis.