Table 1. Clinical Comparison of pNBD and BD without neurological involvement^a

Parameters	pNBD (n=42)	Controls ^b (n=84)	P Value	OR(95%CI)
Age (years, mean±SD)	36.3±11.6	35.2±10.9	0.601	1.01 (0.98-1.04)
Gender (F/M)	25/17	50/34	1.000	—
Clinical Features of BD				
Oral Ulceration	42 (100%)	84 (100%)	1.000	—
Genital Ulceration	27 (64.3%)	61 (72.6%)	0.337	0.68 (0.31-1.50)
Skin Lesions	19 (45.2%)	40 (47.6%)	0.801	0.91 (0.43-1.91)
Gastrointestinal lesions	3 (7.1%)	17 (20.2%)	0.058	0.30 (0.08-1.10)
Ocular involvement ^c	15 (35.7%)	16 (19.0%)	0.041*	2.36 (1.03-5.44)
Vascular involvement	6 (14.3%)	10 (11.9%)	0.705	1.23 (0.42-3.66)
Laboratory tests				
ESR(mm/h, median and range)	15(0-85)	11(1-80)	0.072	1.01(0.99-1.03)
CRP (mg/L, median and range)	3.0(0.14-124)	3.3(0.13-104)	0.995	1.00 (0.98-1.02)

OR= Odds Ratio; CI=Confidential Interval.

^aValues are the number (percentage) unless otherwise indicated.^bEighty-four age- and gender-matched BD patients without neurological involvement were served as control.^cOcular involvement includes uveitis, retinal vasculitis, scleritis.

*P<0.05.

Conclusions: The parenchymal neurological involvement is a rare complication of BD with a high mortality. Its most frequent location is brainstem. Ocular involvement is a risk factor for pNBD. Early identification and active treatment are vital for a good prognosis.

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FRI0493

PET/CT IN PATIENTS WITH GIANT CELL ARTERITIS AFTER 1 YEAR OF TREATMENT WITH TOCILIZUMAB PLUS PREDNISONE TAPER OR ONLY PREDNISONE TAPER

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Background: The arterial uptake of ¹⁸fluorine-2-deoxy-d-glucose (FDG) by positron emission tomography-computed tomography (PET/CT) has been used for the diagnosis of giant cell arteritis (GCA). The role for PET/CT in following disease activity and monitoring treatment effects, however, remains unclear

Objectives: To examine the degree of FDG uptake within the large arteries of GCA patients after 1 year of treatment

Methods: We studied a subgroup of patients enrolled in the GiACTA trial.¹ Patients received either tocilizumab (TCZ) given weekly or every other week plus a 26 week prednisone taper, or placebo (PBO) plus either a 26- or 52 week prednisone taper. PET/CTs were done according to site feasibility and not as part of the GiACTA protocol. For this analysis, the 2 TCZ and PBO groups, respectively, were combined. Patients had whole-body PET/CT imaging within 16 weeks of week 52 visit. The ascending aorta, carotid artery and subclavian artery territories were assessed by maximum standardised FDG uptake values (mSUV) measurement. For each territory, we calculated average, most diseased segment (MDS) and most diseased slide mSUVs and target background ratios (TBR). PET/CTs were read centrally by investigators blinded to the clinical and treatment data

Results: From 251 patients enrolled in GiACTA, 24 patients underwent PET/CT within 16 weeks of week 52 visit. Seventeen patients (71%) received TCZ plus a prednisone taper (TCZ group) and 7 patients received a prednisone taper alone (PBO group). There were no significant differences between groups in baseline characteristics (table 1). Mean (SD) cumulative prednisone dose at week 52 was 2.5 g (1.7 g) in the TCZ group and 3.2 g (1.2 g) in the PBO group (p=0.11). FDG

uptake was consistently numerically lower in all vascular territories in TCZ-treated patients compared to PBO-treated patients except in the ascending aorta. However, the between-group differences were statistically significant only in the left subclavian artery (table 2). Adjustment for selected confounders (new onset disease, baseline prednisone dose, prednisone dose at the time of PET/CT, cumulative prednisone dose, flare prior to PET/CT and clinical activity at the time of PET/CT) did not alter the conclusions derived from the univariate analyses

Table 1 Baseline characteristics: All results represent means (SD) except otherwise indicated. PBO=placebo, TCZ=tocilizumab

	PBO group (n=7)	TCZ group (n=17)	p-value
New onset disease, n (%)	6 (86)	9 (53)	0.19
Baseline prednisone dose (mg)	30 (30-50)	30 (20-40)	0.30
Cumulative prednisone dose (g) at week 52	3.2 (1.2)	2.5 (1.7)	0.11
Prednisone dose (mg) at PET time, median (IQR)	0 (0; 3)	0 (0; 7.5)	0.44
PET time point (weeks from baseline)	54.47 (5.75)	57.38 (8.33)	0.11
Primary outcome met, n (%)	4 (57)	11 (65)	1.00
Clinically active disease at the time of PET, n (%)	1 (14)	3 (18)	1.00
Disease relapse before PET, n (%)	3 (43)	8 (47)	0.61
TCZ treatment at the time of PET, n (%)	0 (100)	7 (41)	0.07

All results represent means (SD) except otherwise indicated. PBO = placebo, TCZ = tocilizumab

Table 2 Results: All results represent means (SD). mSUV=maximum standardised uptake value, MDS=most diseased segment, TBR=target to background ratio

	PBO group (n=7)	TCZ group (n=17)	p-value
Left subclavian artery			
Average mSUV	2.74 (0.77)	2.33 (0.42)	0.11
Average MDS mSUV	3.05 (0.90)	2.47 (0.46)	0.04
Hottest spot mSUV	3.23 (1.02)	2.56 (0.48)	0.04
TBR	2.94 (0.86)	2.47 (0.45)	0.09
Right subclavian artery			
Average mSUV	2.59 (0.86)	2.28 (0.43)	0.25
Average MDS mSUV	2.72 (0.88)	2.35 (0.42)	0.17
Hottest spot mSUV	2.82 (0.91)	2.49 (0.49)	0.26
TBR	2.76 (0.88)	2.41 (0.40)	0.19
Left carotid artery			
Average mSUV	2.51 (0.49)	2.36 (0.41)	0.50
Average MDS mSUV	2.64 (0.54)	2.52 (0.49)	0.66
Hottest spot mSUV	2.70 (0.54)	2.64 (0.54)	0.82
TBR	2.67 (0.52)	2.51 (0.43)	0.78
Right carotid artery			
Average mSUV	2.86 (0.77)	2.38 (0.44)	0.14
Average MDS mSUV	3.12 (0.95)	2.63 (0.65)	0.14
Hottest spot mSUV	3.23 (0.94)	2.76 (0.69)	0.16
TBR	2.98 (0.67)	2.54 (0.46)	0.31
Ascending aorta			
Average mSUV	2.85 (0.63)	2.86 (0.51)	0.93
Average MDS mSUV	2.99 (0.68)	2.98 (0.56)	0.97
Hottest spot mSUV	3.01 (0.71)	3.12 (0.61)	0.97
TBR	3.05 (0.65)	3.05 (0.61)	0.99
Hottest spot total score	13.40 (4.08)	12.62 (3.30)	0.62
Average MDS total score	12.84 (4.01)	12.04 (3.06)	0.60

All results represent means (SD). mSUV = maximum standardized uptake value, MDS = most diseased segment, TBR = target to background ratio

Conclusions: In this exploratory study, subclavian FDG-uptake was significantly reduced in GCA patients treated with TCZ plus prednisone versus prednisone monotherapy despite lower cumulative prednisone doses. A larger, carefully designed prospective study is required to fully understand the utility of PET/CT in the longitudinal evaluation of large-vessel vasculitis associated with GCA

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FRI0494

SPECTRUM OF VISUAL INVOLVEMENT IN GIANT CELL ARTERITIS

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Background: The dreaded complication of giant cell arteritis (GCA), irreversible sight loss (SL), is present in 15%–25% of patients at diagnosis. This is likely due to delayed presentation, recognition and treatment. Fast track pathways for urgent assessment of suspected GCA reduce SL with associated cost savings. Information regarding the burden of SL in GCA is required to guide health strategy and has been explored in this UK wide study.

Methods: This is a prospective observational multi-centre study of visual involvement attributable to GCA. Demographic, disease features and treatment information was collected on all patients with new or relapsing GCA with or without visual symptoms (VS) or SL. SL was defined as symptomatic loss of acuity, field of vision or diplopia ascribable to ischaemic complications of GCA. Patients with transient VS were not considered to have SL. Analyses were performed using chi-squared and rank sum tests.

Results: Patients were predominantly female (274/388) and Caucasian; 3 non-Caucasian (Asian British, Black/Afro-Caribbean and Turkish). VS were present in 135/388 patients (35%) from 19 UK sites. Of patients with VS, these were transient or intermittent SL (n=9,7%), diplopia (n=33,24%), blurred vision (n=17,13%), flashing lights (n=4,3%) and eye pain (n=7,5%). Transient or permanent loss of acuity occurred in 92 patients (48 anterior ischaemic optic neuropathy [AION], 6 central retinal artery occlusion [CRAO], 1 posterior ischaemic optic neuropathy, all attributed to GCA); 1 hemispheric retinal vein occlusion, 1 branch retinal artery occlusion, 1 peri-papillary haemorrhage, 1 vitelliform dystrophy macula. 8 had oculomotor nerve palsies (NP) (1 bilateral 3rd, 1 bilateral 6th and 6 unilateral 3rd or 6th NP). Patients with VS were older than those without (mean $\pm$ SD age 75.9 $\pm$ 9.3 and 73.5 $\pm$ 8.5 respectively, $p<0.001$). Patients with VS had less headaches ($p=0.012$) and scalp tenderness ($p=0.05$) but more frequently tongue claudication ($p=0.014$), shorter symptom duration (3.9 $\pm$ 5.5 days, $p=0.001$), lower CRP although not statistically significant ($p=0.059$), were less likely to meet ACR criteria and to improve than those without VS ($p<0.001$). No association was found with comorbidities or deprivation index. SL in 54 (40%) patients was due to AION or CRAO. Transient CRAO or AION was described in 17 and 30/47 (64%) had permanent SL (data missing from 7 patients). Patients with AION/CRAO were older (80.9 $\pm$ 6.7 years) than those without AION/CRAO (73.3 $\pm$ 8.7 years, $p<0.001$). They had shorter symptom duration (4.2 $\pm$ 7.0 days, $p=0.024$), less likely to improve than those without AION/CRAO ($p<0.001$), less likely to have headaches ($p<0.001$) or polymyalgic symptoms ($p=0.002$), and more likely to have jaw claudication ($p=0.043$) and hypertension ($p=0.002$). Only 7 had diplopia on presentation. 5 patients experienced AION as manifestation of recrudescing disease.

Conclusions: VS often leading to SL in GCA requires urgent management. Patients with VS were older, without 'typical symptoms' such as headache and polymyalgia but more likely to have ischaemic symptoms such as jaw and tongue pain and hypertension. Recognition of VS associated features should be embodied in public and professional awareness programs to prevent permanent SL in GCA.

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FRI0495

BEHÇET'S SYNDROME IN NEW YORK AND AMSTERDAM: EVOLUTION FROM PROBABLE BEHÇET'S TO ISG CRITERIA POSITIVE BEHÇET'S

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Background: Behçet's syndrome (BS) is an auto-inflammatory vasculitis, most common in countries along the ancient Silk Road. Classification of BS most often is done with the International Study Group criteria (ISG criteria)¹. ISG criteria positivity for BS is reached when a patient has recurrent oral ulceration and any two of the following symptoms: recurrent genital ulceration, uveitis, skin lesions and pathergy positivity. However, many other manifestations are reported¹. Differences in clinical presentation can complicate classification of the diagnosis, especially in areas where the disease is low in prevalence. Thus, some patients are classified as "probable BS". In some of these cases patients developed additional symptoms over time and met the ISG criteria in a later stage.

Objectives: To describe characteristics of patients presenting with a probable diagnosis of BS in Amsterdam and New York and to study if patients who eventually met the ISG criteria differ from those who did not.

Methods: We included consecutive patients classified as possible BS to our outpatient clinics in New York and Amsterdam. Patients fulfilling ISG criteria at enrollment were excluded, as well as patients in whom an alternative diagnosis was made at enrollment. Baseline data were evaluated retrospectively and patients were divided into two groups: those developing ISG positive (ISG + or "true") BS during follow up and those who did not meet ISG criteria after follow-up (ISG-). Turkey, Asia, Middle and Far Eastern countries, Arabic countries and Northern Africa were considered endemic areas; Italian, Greek, Hispanic, Portuguese, African-American and Caucasian patients were considered not from endemic areas. Statistical analysis was performed using SPSS, with Chi-square tests or Fisher's exact tests for categorical data and independent sample t-tests for numerical data.

Results: 189 patients were included, of whom 20 were from Amsterdam. During follow up, 71 patients (37.6%) could be classified as "true" Behçet's syndrome after a mean period of 9.4 years ($\pm$ 8.3 years) after onset of symptoms. Age,

gender, ethnicity, duration of symptoms at enrollment, duration of follow up as well as RAPID3 and almost all clinical manifestations at baseline were comparable for both groups. Labial ulcers and skin manifestations at enrollment were more frequently reported. Genital ulcers as a group was not significantly associated with developing "true" Behçet's, nor were specific skin manifestations such as erythema nodosum. We also considered HLA-B*51, pathergy, erythrocyte sedimentation rate and C-reactive protein, but due to a large amount of missing data, we were unable to draw any significant conclusions for these variables.

Characteristics	All patients (n=189)	ISG + (n=71)	ISG - (n=118)	P-value
Age years (SD)	34.3 (13.7)	34.1 (13.4)	34.5 (14.0)	ns
Male n (%)	46 (24.3)	13 (18.3)	33 (28.0)	ns
Duration of follow up years (SD)	3.2 (2.4)	3.3 (2.5)	3.1 (2.4)	ns
Ethnicity: endemic area n (%)	43 (22.8)	14 (19.7)	29 (24.6)	ns
RAPID3 mean score (SD)	9.8 (6.8)	9.6 (6.9)	9.9 (6.7)	ns
Duration of symptoms years (SD)	7.3 (7.8)	7.7 (7.9)	7.0 (7.8)	ns
Oral ulcers n (%)	174 (92.1)	66 (93.0)	108 (91.5)	ns
Genital ulcers n (%)	108 (57.1)	47 (66.2)	61 (51.7)	ns
- Labia	63 (33.3)	31 (43.7)	32 (27.1)	0.019
Skin disease n (%)	58 (30.7)	31 (43.7)	27 (22.9)	0.003
Eye disease n (%)	33 (17.5)	17 (23.9)	15 (12.7)	ns
Deep venous thrombosis n (%)	4 (2.1)	2 (2.8)	2 (1.7)	ns
Thrombophlebitis n (%)	5 (2.6)	2 (2.8)	3 (2.5)	ns
Central nervous system n (%)	22 (11.6)	6 (8.5)	16 (13.6)	ns
Vascular disease n (%)	7 (3.7)	1 (1.4)	6 (5.1)	ns
Pulmonary disease n (%)	3 (1.6)	0 (0)	3 (2.5)	ns
Arthritis n (%)	35 (18.6)	16 (22.5)	19 (16.1)	ns
Gastrointestinal disease n (%)	40 (21.2)	14 (19.7)	26 (22.0)	ns
Headache n (%)	12 (6.3)	5 (7.0)	7 (5.9)	ns
Epididymitis n (% of males)	3 (6.5)	0 (0)	3 (9.1)	ns

ISG +: patients who did fulfill ISG criteria after follow-up; ISG -: patients who did not fulfill ISG criteria after follow-up; SD: standard deviation; ns: not significant

Figure 1 Baseline data from all patients who were classified as probable Behçet's syndrome at enrollment

Conclusions: About a third of patients classified as probable Behçet's syndrome at enrollment develop new manifestations over time. Thus, they can be reclassified as ISG positive ("true") BS. Based on our data, presence of skin manifestations and labial ulcers at enrollment was significantly higher in the group eventually developing a ISG criteria positive BS.

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FRI0496

POLYARTERITIS NODOSA: OVER 20 YEARS' CLINICAL EXPERIENCE

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Background: Polyarteritis nodosa (PAN) is a necrotizing vasculitis of predominantly medium size vessels.

Objectives: The present study aimed to summarise the characteristics of PAN patients, and also analyse the trend of decreasing PAN frequency in the last 25 years, in Turkey

Methods: Paediatric and adult PAN patients followed up in Hacettepe University and Istanbul University Cerrahpaşa Faculty of Medicine between 1990 and 2015, were included. The demographics, clinical findings and outcomes were evaluated retrospectively.

Results: One hundred thirty-three patients, including 66 children, were enrolled in the study. The mean follow-up duration was 13²⁻²⁷ years. Among 133 patients, 86 (64.7%) had fever, 108 (81.2%) had skin involvement, 54 (40.6%) had renal involvement, 43 (32.3%) had neurological involvement, 32 (24.1%) had gastrointestinal involvement, 10 (7.5%) had cardiac involvement, 6 (4.5%) had pulmonary involvement. The median (minimum-maximum) leukocyte count, erythrocyte sedimentation rate and C-reactive protein levels at the time of diagnosis were 10400 (6100-32000)/mm³, 58(2-132) mm/h and 5.22 (0-46) mg/dL, respectively. All patients were ANCA negative. Hepatitis serology was analysed in 121 patients and found positive in 13 of them. *MEFV* mutations were screened among 65 patients, 24 of them had mutations in at least one allele. Myalgia and skin involvement were significantly more frequent in children whereas neurologic involvement