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.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5533

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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Disclosure of Interest: F. Kerstens Grant/research support from: Stichting Jan van Breemen Grant, F. Turkstra: None declared, C. Swearingen: None declared, Y. Yazici: None declared

DOI: 10.1136/annrheumdis-2018-eular.7069

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

was much more common among adults (table 1). The number of PAN patients declined significantly after 2010 (figure 1). 9 patients were re-categorised as DADA2 after 2014 and no patient were diagnosed with FMF+. PAN after 2008

Abstract FRI0496 – Table 1. Characteristics of paediatric and adult PAN patients

Features, n (%)	Patients (n=133)	Paediatric patients (n=66)	Adult patients (n=67)	P value
Gender, female	55 (41.4)	35 (53)	20 (29.8)	0.07
Fever	86 (64.7)	50 (75.7)	36 (52.9)	0.06
Myalgia	79 (59.4)	48 (72.7)	31 (46.2)	0.01
Abdominal pain	65 (48.9)	40 (60.6)	25 (37.3)	0.05
Weight loss	56 (42.1)	20 (30.3)	36 (53.7)	0.06
Arthralgia	87 (65.4)	48 (72.7)	39 (58.2)	0.06
Arthritis	23 (17.3)	11 (16.6)	12 (17.9)	0.58
Skin involvement	108 (81.2)	63 (95.5)	45 (67.2)	<0.001
GI ^a involvement	32 (24.1)	17 (25.7)	15 (22.3)	0.64
Renal involvement	54 (40.6)	22 (33.3)	32 (47.7)	0.09
Cardiac involvement	10 (7.5)	8 (12)	2 (2.9)	0.04
Neurologic involvement	43 (32.3)	14 (21.2)	29 (43.2)	0.007
WBC ^a , x10 ³ /mm ³	10400 (4200–32000)	11800 (4300–32000)	9850 (4200–21800)	0.02
PLT ^a , x10 ³ /mm ³	365000 (259000–726000)	456000 (259000–696000)	300000 (268000–726000)	<0.001
CRP ^a , mg/dl	5.22 (0–46)	4.6 (0.1–46)	5.8 (2–29.8)	0.88
Exitus	6 (4.5)	0 (0)	6 (8.9)	0.007

^aCRP, C reactive protein; GI, gastrointestinal; PLT, platelet; WBC, white blood cell

Abstract FRI0496 – Figure 1. Distribution of the PAN patients by years

Conclusions: Our results suggest a decrease in PAN in our country which may be due to improved healthcare and dissecting mimicking diseases. Further prospective studies with prolonged follow-up could help us to better understand the disease characteristics.

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Acknowledgements: None

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5533

FRI0497 CLINICAL CHARACTERISTICS AND TREATMENT OUTCOMES OF PATIENTS WITH BEHÇET'S DISEASE AND VASCULAR INVOLVEMENT

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Background: The international Chapel Hill Consensus Conference (CHCC) categorised Behçet's disease (BD) as variable vessel vasculitis, which was defined as vasculitis that can affect vessels of any size and type. Vascular manifestations

have been reported in up to 50% of patients with BD, most commonly as venous thrombosis.

Objectives: The aim of this study is to investigate characteristics and treatment outcomes of patients with vascular BD, particularly with large vessel involvement.

Methods: A retrospective review was performed on 2496 patients with ICD-10 code of Behçet's disease and/or aortic disease who visited Samsung Medical Centre between 2004 and 2014. Patients who were suspected to have BD and vascular involvement were enrolled.

Results: Eighty-three patients with clinical features of BD and vascular involvement were identified. Although less than half satisfied the classic International Study Group (ISG) criteria, greater proportion fulfilled the International Criteria for BD (ICBD), and all of the patients satisfied at least "suspected BD" according to the Japanese criteria. Fifty patients had arterial lesions including 34 with aorta involvement. Another 33 patients had venous thrombosis without arterial lesions. Patients showed a male predominance (87%) and a predilection of young age with the median of 42 years old. The most prominent type of arterial lesions was aneurysm (80%) with a high frequency of pseudoaneurysms. Among the aorta, the thoracic aorta was most commonly involved and 18 patients had aortic valve regurgitation. Seventy-five (90%) patients received glucocorticoids with a median initial dose of prednisolone of 30 mg per day and 68 (82%) received immunosuppressive treatment. Half of the patients had more than one relapse after stabilisation of the first vascular event. During the course, 44 patients underwent surgery and/or endovascular treatment and 60% of these patients had repeated treatment for the relapse. These included 31 aortic valve replacements on 13 patients.

Conclusions: Patients with vascular BD showed a predilection of young male and 60% of patients had arterial involvement. The aorta was affected in more than half of the patients who had arterial involvement. Aneurysmal change was frequent finding and relapse rate was high during disease course. Further studies into a practical and specialised diagnostic tool for vascular BD and optimal treatment strategies are required.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3617

FRI0498 COMPARATIVE STUDY OF THE TREATMENT OF REFRACTORY CYSTOID MACULAR OEDEMA TO CONVENTIONAL IMMUNOSUPPRESSIVE THERAPY: TOCILIZUMAB VS ANTI-TNF. MULTICENTER STUDY OF 59 PATIENTS

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Background: Cystoid macular oedema (CME) is the most serious complication of uveitis. This potentially severe complication may lead to irreversible visual loss.^{1–2}

Objectives: To compare efficacy and safety of Tocilizumab (TCZ) vs. Anti-TNF-α drugs in patients with refractory CME to conventional immunosuppressant (IS).

Methods: Multicenter study of patients with refractory CME to treatment with glucocorticoids and at least 1 conventional IS. The main objective was the improvement of macular thickness. Secondary objectives were the enhancement in best corrected visual acuity (BCVA) and the degree of ocular inflammatory activity.

Results: 59 patients/112 affected eyes. Causes of uveitis were: Behçet's disease (n=41), Birdshot's retinochoroidopathy (n=4), Juvenile Idiopathic Arthritis (n=9), Sarcoidosis (n=1) and idiopathic (n=4). No significant differences were observed at baseline in both groups (TCZ vs Anti-TNF-α) in sex (♂/♀; 8/17 vs. 15/19), mean age (35.6±18.9 vs 40.0±9.1), BCVA (0.40±0.31 vs. 0.48±0.31), Tyndall (1 [0–1] vs. 1 [0–2]), vitritis (1 [0–2] vs. 1 [0–3]) and macular thickness (431.8±165.9 vs. 397.1±138.1) (table 1).

25 patients were treated with TCZ as follows: 8 mg/kg/4 weeks (n=24) and 162 mg sc/2 weeks (n=1). Anti-TNF-α therapy was used in the remaining 34 as