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