was shorter at relapsing patients (44 vs 64 months, $p=0.001$). Smoking rate was higher at male patients but no association was observed with vascular relapses.

Abstract FRI0488 – Table 1. Clinic and demographic features of Behçet's Disease patients

		Female (n=22)	Male (n=102)	p
Mean age at first vascular event		37±12.6	31.5±8.5	0.019
Mean age at BD diagnosis		30.5±9.5	29.1±7.5	0.467
Smoking	Yes	4 (%33.3)	32 (%68.1)	0.018
	No	8 (%66.7)	15 (%31.9)	
Pteryg	Positive	12 (%66.7)	54 (%62.8)	0.99
	Negative	6 (%33.3)	32 (%37.2)	
Number of vascular event/s	1	14 (%63.6)	55 (%54.5)	
	2	6 (%27.3)	39 (%38.5)	
	3 or more	2 (%9.1)	7 (%7)	

Abstract FRI0488 – Table 2. Type and characteristics of vascular involvement

Venous n=134 (%73.2)	Ekxtremities	106 (%) 79.1)	Arteries n=23 (%23.5)	Pulmoner Trombus	36 (%) 81.8)
	Cerebral	16 (%) 12)		Aneurysme (2 coronary, 2 pulmoner, 2 aorta)	6 (%) 13.6)
	Others (SVC, IVC, renal, retinal)	12 (8.9)		PAT+PAA	2 (%) 4.6)

IVC: inferior vena cava, PAA: pulmoner arter aneurysmei, PAT: pulmoner arter trombus, SVC: superior vena cava

Conclusions: Our results show that female BD patients have a vascular event at a later age compared to males, but the course of vascular disease is not influenced with gender. Early termination of immunosuppressive treatments seems to be the most important cause of vascular relapses

Disclosure of Interest: None declared

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FRI0489 UTILITY OF APREMILAST IN REFRACTORY ORAL AND/OR GENITAL ULCERS IN BEHÇET'S DISEASE

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Background: Behçet's disease (BD) is characterised by recurrent oral and/or genital ulcers accompanied by ocular, cutaneous, articular, gastrointestinal, and/or neurologic manifestations. Oral and/or genital aphthous ulcers are often refractory to conventional treatment. Apremilast is an orally-active small molecule which inhibits phosphodiesterase-4 (PDE-4) that modulates some inflammatory pathways.

Objectives: Our aim was to assess the efficacy of apremilast in BD patients with oral and/or genital ulcers refractory to conventional treatment.

Methods: Retrospective national multicenter open-label study on 19 BD patients treated with apremilast at standard dose of 30 mg twice daily. The main outcome was achievement of oral ulcers remission.

Results: We included 19 patients (14 women and 5 men) with a mean age of 43.6 ±14.8 years. Before apremilast, all patients had also received several systemic conventional drugs: oral corticosteroids (n=18), colchicine (n=19), NSAIDs (n=10), methotrexate (n=10), azathioprine (n=10), cyclosporine (n=6), infliximab (n=3), adalimumab (n=5), dapsone (n=3), etanercept (n=1), mycophenolate mofetil (n=1), tocilizumab (n=1). The main clinical symptoms for starting apremilast were oral aphthous ulcers (n=19) and genital ulcers (n=14). Other manifestations present at apremilast onset were arthralgia/arthritis (n=6), folliculitis/pseudofolliculitis (n=6), asthenia (n=5), furunculosis (n=1), erythema nodosum (n=1), erythematous and scaly skin lesions (n=1), psoriasis (n=1), deep venous thrombosis (n=2) and ileitis (n=1). Table 1 shows the evolution of the patients. After a median follow-up of 6 [interquartile range, 5–10] months, most of the patients experienced clinical improvement. In this period of time, 11 patients developed any side-effect: dyspepsia (n=5), nausea (n=4), diarrhoea (n=4), abdominal pain (n=4), headache (n=3), loss of appetite (n=3), weight loss (n=1) and halitosis (n=1). Three patients had to reduce the dose to 30 mg/day. Apremilast was discontinued in 4 patients: because of not obtaining the expected improvement (n=2), due to desire of pregnancy (n=1) and due to development of neurological involvement (n=1).

Abstract FRI0489 – Table 1

	Baseline n=19	Week 1-2 n=19	Week 4 n=19	Month 3 n=18	Month 6 n=11
Resolution of main symptom, oral and/or genital ulcers n, (%)					
Complete		7/19 (36.8)	12/19 (63.1)	13/18 (72.2)	7/11 (63.6)
Partial		8/19 (42.1)	4/19 (21.0)	1/18 (5.5)	3/11 (27.3)
Resolution of other symptoms n, (%)					
Complete		5/13 (38.5)	6/13 (46.1)	8/12 (66.6)	5/7 (71.4)
Partial		1/13 (7.7)	2/13 (15.4)	4/12 (33.3)	2/7 (28.6)
Dose of prednisone (mg/day), median [IQR]	7.5 [5-12.5]	6.2 [5-10]	6.2 [5-10]	3.7 [0-5]	2.5 [0-4]

Conclusions: Apremilast leads to a rapid and maintained improvement in many patients with refractory mucocutaneous ulcers of BD. Even in patients refractory to several systemic drugs including biologic therapy. However, the development of adverse digestive effects is frequent.

Disclosure of Interest: None declared

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FRI0490 MORTALITY AND EARLY SEVERE INFECTION IN PATIENTS WITH ANCA-ASSOCIATED VASCULITIS

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Background: The introduction of treatment regimens comprising of cyclophosphamide or rituximab combined with corticosteroids has brought about dramatic improvements in the prognosis of ANCA-associated vasculitis.¹ Severe infectious events, especially in the early phase of treatment, associated with risk of death have been reported in the past several studies.^{2,3}

Objectives: We retrospectively investigated the association between mortality and early severe infection in patients with antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis (AAV), and we also attempted to identify the potential predictors for early severe infection.

Methods: We recruited 182 consecutive hospitalised patients newly diagnosed with AAV at our hospital, from January 2000 to June 2017, in this retrospective cohort study. All cause mortality, relapse, and severe infections within six months after starting treatment (early severe infection) were analysed.

Results: The mean age was 70 years at diagnosis, and the classification according to the Chapel Hill Conference definition were microscopic polyangiitis (MPA) in 82 patients (45.1%), granulomatosis with polyangiitis (GPA) in 36 patients (19.8%), eosinophilic granulomatosis with polyangiitis (EGPA) in 24 patients (13.2%), and renal-limited vasculitis in 32 patients (17.6%).⁴ Median follow up was 158 weeks (range 0–182 w). Using Cox regression analysis, elderly onset (age ≥75 years) AAV ($p=0.027$) and early severe infection ($p<0.001$) were independent predictors of all cause mortality (table 1).

Early severe infection tended to increase among patients who received immunosuppressive therapy of a corticosteroid combined with cyclophosphamide or rituximab (conventional treatment), and this trend was significant in non-severe (BVAS <20) AAV patients ($p=0.030$) (table 2). Treatment response rate ($p=0.058$)