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TABLE 1. Main baseline features and follow-up of a series of 177 patients with refractory uveitis due to Behçet's disease undergoing infliximab (IFX) or adalimumab (ADA) therapy.

	IFX group (n=102)	ADA group (n=75)	p
Number of patients/affected eyes, (n)	103/165	74/131	
Age, mean (SD) years	40.4 (10.1)	38.7 (11.3)	0.29
Sex, men/women, n/n	55/48	39/35	0.92
HLA-B*51 positive, (%)	69.4	68.9	0.74
Duration of uveitis before anti-TNF- α , median (IQR) months	36 (12-72)	24 (12-60)	0.69
Ocular features at the time of anti-TNF- α onset			
AC cells, median (IQR)	1 (0-2)	1 (0-2)	0.25
Vitreitis, median (IQR)	1 (0-2)	1 (0-2)	0.12
BCVA, mean (SD)	0.50 (0.30)	0.56 (0.34)	0.08
Macular thickness, mean (SD)	331.11 (53.97)	345.37 (58.14)	0.49
Presence of retinal vasculitis, n (%)	114 (56)	78 (55)	0.51
Presence of choroiditis, n (%)	41 (21)	10 (7)	<0.01
Pattern of uveitis, n (%)			
Bilateral	82 (79.61)	57 (77.03)	0.68
Unilateral	21 (20.39)	17 (22.97)	0.68
Anterior	11 (10.68)	14 (18.92)	0.19
Posterior	28 (27.18)	14 (18.92)	0.19
Panuveitis	64 (62.14)	45 (60.81)	0.19
Intermediate	9 (9)	1 (1.35)	0.19
Treatment before anti-TNF- α onset, %			
Oral corticosteroids	95	88	0.08
Intravitreal pulses of MP	31	31	0.98
Csa	75	78	0.85
AZA	57	57	0.89
MTX	44	42	0.77
Other treatments	3.84	1.92	0.41
Prednisolone dose at anti-TNF- α onset, mean (SD), mg/d	54.35 (15.84)	53.37 (17.52)	0.37
Combined treatment, %			
AZA	76.5	76.3	
Csa	21.8	19.2	
MTX	41.1	55.7	
CFM	35.3	21.1	
MMF	1.3	0.0	
FK-506	1.3	0.0	0.35
Follow-up on anti-TNF- α therapy, mean (SD), months	31.52 (23.51)	26.48 (18.57)	0.13
Remission, n (%)	78 (76.47)	61 (82.43)	0.34
Relapses, mean (SD)	1.13 (2.62)	1.66 (8.62)	0.61
Drug withdrawal, n (%)	57 (55.33)	21 (28.37)	<0.01
-remission, n (%)	20 (19.41)	6 (8.45)	0.58
-inefficacy, n (%)	18 (17.47)	11 (14.88)	0.09
-severe side-effects/toxicity, n (%)	8 (7.78)	4 (5.38)	0.58
-others, n (%)	11 (10.68)	0 (0)	0.03
Serious side-effects, n (per 100 patients/year)	4 (1.48)	4 (2.46)	0.40

Abbreviations: ADA, adalimumab; AZA, azathioprine; CFM, cyclophosphamide; Csa, cyclosporine A; FK-506, tacrolimus; IFX, infliximab; IQR, interquartile range; MMF, mycophenolate mofetil; MTX, methotrexate; MP, methylprednisolone; SD, standard deviation; TNF- α , tumor necrosis factor alpha.

FRI0270

APREMILAST IN NON-ULCER MANIFESTATIONS IN BEHÇET'S DISEASE. MULTICENTER STUDY OF 32 CASES IN CLINICAL PRACTICE

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Background: Behçet's disease (BD) is a variable vessel vasculitis with a wide and heterogeneous set of signs and symptoms. The inhibitor of phosphodiesterase-4 Apremilast (APR) has demonstrated efficacy in the treatment of oral and/or genital ulcers.

Objectives: To assess the efficacy and safety of APR in BD patients with manifestations different from mucocutaneous ulcers.

Methods: National multicenter retrospective study on 32 BD patients treated with APR at maintained standard dose of 30 mg twice daily.

Results: From a cohort of 49 patients with oral and/or genital ulcers related to BD and refractory to conventional and/or biological treatment, we selected the cases with another clinical manifestation/s (n=32, 23 women/9 men), mean age of 46.35±15.05 years. Non-aphthous manifestations present at apremilast onset were: arthralgia/arthritis (15), folliculitis/pseudofolliculitis (12), asthenia (7), erythema nodosum (3), furunculosis (2), paradoxical psoriasis by TNFi (2), ileitis (2), deep venous thrombosis (2), erythematous and scaly skin lesions (1), fever (1), eating disorder (1), fibromyalgia (1), unilateral anterior uveitis (1) and neurobehçet (1). APR was used in monotherapy (n=3) or combined (n=29) with oral corticosteroids (20), colchicine (17), methotrexate (5), azathioprine (3), dapsone (1), tocilizumab (1), hydroxychloroquine (1) and/or mesalazine (1).

The outcome of the different clinical symptoms is shown in TABLE. The patient with neurobehçet kept stable (paresthesias) during the 6 months of follow-up. The 2 cases of deep venous thrombosis and the case of anterior uveitis resolved with anticoagulants and adjuvant topical treatment, respectively. Furunculosis, folliculitis/pseudofolliculitis and ileitis were the manifestations that improved completely and rapidly. The cases of arthritis experienced improvement, while those with arthromyalgias presented a torpid evolution.

Conclusion: Our data show an improvement of the cutaneous follicular and intestinal clinic with APR and a stability of the neurological clinic, while the musculoskeletal manifestations were mostly refractory.

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TABLE. Outcome of non-aphthous clinic with apremilast.

	1-2 Weeks	4 Weeks	3 Months	6 Months	12 Months	18 Months	24 Months
Non-aphthous manifestations at APR onset (n)							
Neurobehçet (NCS vasculitis) (1)	NC	NC	NC				
Unilateral anterior uveitis (1)	CR	CR					
Deep venous thrombosis (2)	CR (2)	CR (2)	CR (2)	CR (2)	CR (1)	CR (1)	CR (1)
Arthromyalgias (11)	CR (1)	CR (1)	PR (4)	PR (5)	PR (5)	NC (1)	NC (1)
	NC (10)	PR (2)	NC (1)	NC (1)			
Arthritis (4)	NC (3)	PR (2)	CR (1)	CR (1)	CR (1)		
	ND (1)	ND (1)	ND (1)	ND (1)			
Folliculitis/pseudofolliculitis (12)	CR (5)	CR (8)	CR (9)	CR (9)	CR (2)	CR (1)	
	PR (2)	PR (3)	PR (3)	NC (1)			
Furunculosis (2)	NC (5)	NC (1)	NC (1)				
	CR (2)	CR (2)	CR (1)	CR (1)			
Erythema nodosum (3)	PR (1)	PR (1)	CR (1)	NC (1)			
	ND (2)	NC (1)					
Psoriasis/erythematous-scaly skin lesions (3)	NC (2)	NC (2)	PR (3)	PR (2)	PR (2)	PR (1)	PR (1)
	ND (1)	ND (1)					
Ileitis (2)	CR (1)	CR (2)	CR (2)	CR (2)	CR (1)		
	PR (1)						
Asthenia (7)	CR (1)	CR (1)	CR (1)	NC (4)	NC (2)	NC (1)	NC (1)
	NC (6)	NC (6)	NC (6)				
Fever (1)	ND	ND					
Eating disorder (1)	ND	ND	ND				
Fibromyalgia (1)	NC	NC	NC				

Abbreviations: CR= complete response; (n)= number of cases; ND= non-data available; NC= no changes observed; PR = partial response.

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FRI0271

USE OF CONTRAST ENHANCED ULTRASOUND SONOGRAPHY (CEUS) IN LARGE VESSEL VASCULITIS (LVV)

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Background: C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) are important parameters in the monitoring of LVV. Since Tocilizumab is approved for treatment of LVV these cheap and easy repeatable parameters are worthless because of their normalisation by Tocilizumab. MRI and PET-CT as an alternative are not only much more expensive, they are also not arbitrarily repeatable and available. Thus, monitoring of LVV-Patients undergoing a Tocilizumab therapy remains unclear – especially upon showing a persisting thickened vessel wall.

Objectives: CEUS can increase the visibility of tissue perfusion, particularly if there is a very slow bloodflow, which cannot be detected by (power)-doppler sonography.

Methods: In this proof of concept study we investigated patients with active and inactive LVV (aLVV/iLVV) with CEUS. After injection of ultrasound contrast agent we measured the contrasted area of large vessels in a transverse section first if the lumen was completely contrasted and once again 4-8 seconds later. If the vessel wall incorporated the contrast agent the contrasted area increased (Fig 1). The increase of the contrasted area (CA) was correlated with CRP and ESR. Patients were only included if they were not treated with Tocilizumab and therefore ESR and CRP were usable to evaluate the disease activity.

Results: Investigated were 16 patients (13 female, 3 male), 8 with aLVV and 8 with iLVV, respectively. The mean CRP was 85 ± 69 (aLVV) vs. 4 ± 2 mg/l (iLVV) ($p < 0.0001$), the ESR 80 ± 28 (aLVV) vs. 7 ± 4 (iLVV) mm/h ($p < 0.0001$). The mean age was 74.6 ± 8.4 y (range 56-82). The increase of the CA was 66.6 ± 44.6 (aLVV) vs. 2.4 ± 6.6 (iLVV) ($p < 0.0001$). The increase correlated significantly with the CRP $r = 0.87$, $p < 0.0001$. An increase of CA of $\geq 20\%$ has a sensitivity of 92,3% and a specificity of 90% for active LVV.

Conclusion: The results of our proof of concept study demonstrate, that CEUS can detect aLVV with a good sensitivity and specificity. Including CEUS in clinical routine will be much easier repeatable, save, quicker and by far more cost-effective than MRI or PET-CT. CEUS might be a good method for monitoring disease activity in LVV treated with Tocilizumab. The limitation of our study is the small number of patients, the missing blinding of the investigator and the method intrinsic fact, that you can't investigate all involved vessels by ultrasound/CEUS.

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None

A: Vessel before contrast agent (1: lumen; 2: thickened vessel wall)
B: Vessel after lumen is contrasted (3: contrasted lumen)
C: Vessel after lumen and wall is contrasted (4: contrasted lumen and wall)

Fig.1

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FRI0272

RISK OF POTENTIAL GLUCOCORTICOID-RELATED ADVERSE EVENTS IN PATIENTS WITH GIANT CELL ARTERITIS: RESULTS FROM A US-BASED ELECTRONIC HEALTH RECORDS DATABASE

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Background: Oral glucocorticoids (OGC) have been the mainstay of treatment for giant cell arteritis (GCA). However, OGCs are associated with several adverse events (AEs).

Objectives: To estimate the risk of potential OGC-related AEs in patients with GCA.

Methods: This retrospective, observational cohort study utilized the 2008-2017 IBM Explorys Electronic Health Records database which includes lab values. Inclusion criteria included age ≥ 50 years with ≥ 2 GCA diagnoses ≥ 7 days apart, 1 OGC prescription within 6 months of the first GCA diagnosis (index date = date of first OGC prescription) followed by a second OGC prescription, no other autoimmune disease requiring high-dose OGCs, no exposure to anti-tumor necrosis factor or anti-interleukin-6 therapies, ≥ 1 C-reactive protein (CRP)/erythrocyte sedimentation rate (ESR) lab test and 12 months of data available pre- and post-index. Potential AEs assessed during the 12 months post-index were descriptively summarized across cohorts of patients based on quartiles (Q) of mean daily dose of OGCs measured over 6 months post-index among this patient sample (Q1: ≥ 1.00 to ≤ 13.75 mg; Q2: > 13.75 to ≤ 25.00 mg; Q3: > 25.00 to ≤ 40.00 mg; Q4: > 40.00 mg). Potential AEs included type 2 diabetes (T2D) diagnosis, hemoglobin A1c (HbA1c), blood glucose level, serious infections, cataracts, gastrointestinal bleeding or ulcer and increases in body mass index (BMI). Actual OGC use by patient could not be confirmed and is a limitation of this study.

Results: Mean age of the 785 eligible patients was 76 years (SD 9); 70% were female. Mean Deyo Charlson Comorbidity Index score at baseline was 1.57 (SD 2.01). The most common baseline comorbid conditions were cerebrovascular disease, diabetes, chronic pulmonary disease, and renal disease. Mean daily OGC dose was 28.9 mg during the first 6 months post-index. Mean (SD) CRP and ESR during the 12-month follow-up was 5.1 (13.6) and 26.5 (20.7), respectively. The proportion of patients with newly diagnosed T2D or with HbA1c ≥ 7.5 during the 12-month follow-up ranged from 7.5% to 24.5% from OGC daily dose Q1 to Q4 cohorts. The proportion of patients with glucose ≥ 200 mg/dL ranged from 7.5% to 15.0% from Q1 to Q4. Serious infections ranged from 16.8% to 24.8% from Q1 to Q4 and cataract ranged from 12.0% to 21.7% from Q1 to Q4. The proportions of patients with gastrointestinal bleed/ulcer ranged from 6.0% in Q1 to 11.8% in Q4. An increase in BMI of 5 ranged from 4.1% to 6.4% from Q1 to Q4.

Conclusion: In patients with GCA, potential OGC-related AEs increased with increased daily OGC dose. This highlights the need for effective therapies that reduce the exposure and potential risk of OGCs.

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