Table 1. Summary of Recommended Measures

Description	SF-36	FACIT-Fatigue	PtGA
Type	Generic, health status measure	Fatigue and impact on function/daily activities	Measure of disease activity
Method of Administration	Self/Interviewer	Self/Interviewer	Self
# of Items	36	13	1
Recall period	Past 4 weeks/past week	Past 7 days	Varies
Scoring	8 domain scores; 2 component summary scores	Total score	Total score
Measurement properties	Measurement properties demonstrated in other rheumatologic diseases (SLE, RA, SpA, and chronic knee pain); construct validity demonstrated in a single study with GCA patients	Measurement properties demonstrated in rheumatologic populations including SLE, PsA, and RA	Select measurement properties have been demonstrated in GCA and other populations (RA, TAK, RP)
Concept coverage	GCA impacts (emotional, physical, and social functioning, and daily activities), bodily pain, fatigue	Fatigue	Overall assessment of disease activity
Administered in recent GCA studies	YES	YES	YES

SLE= systemic lupus erythematosus, SpA = spondyloarthritis, PsA=psoriatic arthritis, RA=rheumatoid arthritis, GCA= giant cell arteritis, TAK= Takayasu's arteritis, RP=relapsing polychondritis

Disclosure of Interests: Hilary H Colwell Consultant of: Janssen Pharmaceutical Companies of Johnson & Johnson, Susan Mathias Consultant of: Janssen Pharmaceutical Companies of Johnson & Johnson, Kelly Kato Employee of: Janssen Pharmaceutical Companies of Johnson & Johnson, Catherine E Najem Shareholder of: Johnson & Johnson, Employee of: Janssen Pharmaceutical Companies of Johnson & Johnson, Amy M Archer Employee of: Janssen Pharmaceutical Companies of Johnson & Johnson

DOI: 10.1136/annrheumdis-2021-eular.1199

AB0358 ABATACEPT AS A bDMARD IN STEROID-REFRACTORY POLYMYALGIA RHEUMATICA

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Background: polymyalgia rheumatica (PMR) is a chronic inflammatory disorder affecting elderly people. Glucocorticoids (GC) are the mainstay of therapy for PMR. Despite this treatment, the disease may relapse or GC dosage cannot be tapered. Methotrexate (MTX) may be helpful in these cases. Blockade of IL-6 has an effective steroid-sparing effect in patients with giant cell arteritis (GCA) and may improve the clinical symptoms of patients with PMR. The pathophysiology of PMR still remains uncertain but there are evidences for a participation of Th1 and Th17 lymphocytes. Recently, blockade of the costimulatory pathway by abatacept (ABA) gave favorable results in GCA.

Objectives: to report our experience (efficacy and safety) of ABA in the treatment of patients with isolated PMR who required longstanding GC and who failed to respond to MTX and/or IL-6 receptor blocking agents.

Methods: a call for observations of all cases of patient with PMR who received at least one dose (SC or IV) of ABA was sent to the members of the French specialist network "Club Rhumatismes & Inflammation" (CRI: www.cri-net.com)(rheumatologist and internal medicine). Patients must satisfy the EULAR/ACR criteria for PMR and have isolated PMR without associated GCA.

Results: 4 cases were declared during a 12 months period: 2 men and 2 women; age (median [range]): 60.7 [50-68] years; disease duration 30 [7-48] months; duration of GC treatment before starting ABA: 60 [10-72] months.

They all required prednisolone with a daily dosage of 14.5 mg [10-20]. Before ABA administration, they all received MTX while 2 patients were treated by IL-6R inhibitor (tocilizumab 6 months and sarilumab 3 months, respectively) without improvement. ABA was given as a monthly infusion (10 mg/kg) in one case and subcutaneously (every week) in 3 cases. ABA treatment duration ranged from 3 to 18 months. Two patients responded to the treatment with a progressive decline of DAS-PMR and GC dosage tapering during a 12 month follow-up, while there was no improvement for the 2 others (Table 1). CRP levels also decreased for one responder. The safety was excellent for all. ABA was still maintained in one responder. Among the non-responders, one patient was switched to tocilizumab and the second still received high GC dosage.

Conclusion: ABA may be effective in certain PMR patients who were unable to taper GC, with a good safety profile. A randomized controlled trial is required in order to determine its place in the treatment of PMR and to select the appropriate patients who could potentially benefit from this biological agent.

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Table 1. changes in DAS-PMR and CRP levels in 4 patients with PMR while receiving abatacept (M: months; CRP in mg/L).

Cases	DAS- PMR M0	DAS- PMR M3	DAS-PMR M6	DAS- PMR M12	CRP M0	CRP M3	CRP M6	CRP M12
1	20	21	21	NA	5	8.6	8.4	NA
2	31.1	30.5	NA	NA	21	25	NA	NA
3	10.7	8.35	5	13	37.2	22.7	22	22
4	18	22	17.2	12	1.2	3.3	3.5	3

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.1246

AB0359 CRANIAL INVOLVEMENT IN BEHÇET'S DISEASE

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Background: Behçet's disease (BD) is a vasculitic multisystem inflammatory disorder. It may also involve the skin, mucosa, eyes, blood vessels, joints, gastrointestinal system, and central nervous system (CNS).

Objectives: In this study, we aimed to present CNS involvement data in patients followed up with a diagnosis of BD.

Methods: The clinical, demographic, laboratory and medication data of 394 patients who were followed up with a diagnosis of BD in our rheumatology outpatient clinic between 2000 and 2020 were retrospectively evaluated.

Results: CNS involvement was detected in %5.6(24) patients who were followed up with a diagnosis of BD. 66.7% of the patients were male, and their mean diagnosis age was 25.1±8.2 years, mean CNS involvement age was 28.39±9.6 years. Neurological symptoms occurred in an average of 3.3 years after the diagnosis of BD. Parenchymal involvement was present in 54.2% of the patients with CNS involvement, 45.8 % had cerebral venous thrombosis. Oral aphthous ulcer was found in 91.7 % of the patients, genital ulcer in 70.8%, pathergy positivity in %45.8 papulopustular lesion in %37.5, erythema nodosum in %45.8. Patients with CNS involvement had headache, vision loss, diplopia, hemiparesis, epilepsy, proptosis, and ataxic gait and walking difficulty in 66.6%, 12.5%, 25%, 12.5%, 8.3%, 8.3% and 12.5%, respectively. Patients with and without CNS involvement were compared in terms of clinical findings and medications. A significant difference was observed in terms of cardiac involvement (Table 1). While one of the 2 patients with cardiac involvement had a thrombus in the right ventricle, the other had pericardial effusion. The patient with cardiac thrombus also had cerebral venous thrombus. Steroid, azathioprine, cyclophosphamide treatments have been used at a higher in patients with CNS involvement. In addition, the patients with parenchymal and cerebral venous involvement were compared in terms of clinical, demographics findings and medications. The presence of papulopustular lesions (9.1% vs 57.1%) and pathergy positivity (18.2% vs 71.4%) were higher in patients with parenchymal involvement (p:0.033, p: 0.020, respectively).

Table 1. Comparison of data of patients with and without CNS involvement

	CNS Involvement (+)	CNS Involvement (-)	
n (%)	n:24	n:370	P
Gender			0.396
Female	8(33.3)	164(44.3)	
Male	16(66.7)	206(55.7)	
Oral Aphthous Ulcer	22(91.7)	357(96.5)	0.230
Genital Ulcer	17(70.8)	239(64.6)	0.661
Papulopustular lesion	9(37.5)	120(32.4)	0.655
Erythema Nodosum	11(45.8)	139(37.6)	0.516
Pathergy	11(45.8)	174(47)	0.988
Uveitis	9(37.5)	129(34.9)	0.827
Retinal Vasculitis	1(4.2)	10(2.7)	0.504
Arthritis	3(12.5)	95(25.7)	0.221
Deep Venous Involvement	6(25)	59(15.9)	0.256
Artery Occlusion	1(4.2)	8(2.2)	0.436
Arterial Aneurysm	1(4.2)	14(3.8)	1
GIS involvement	2(8.3)	15(4.1)	0.277
Cardiac involvement	2(8.3)	4(1.1)	0.046

Conclusion: It has been reported that neurological symptoms occur in an average of 2.5 to 6.5 years after the diagnosis of BD. On the other hand, it has been stated that neurological complications may be the first involvement in 3-33% of the cases and this situation will cause diagnostic difficulties¹. These data are consistent with our study.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.1365

AB0360

A DECADE OF SUSPECTED GIANT CELL ARTERITIS: CLINICAL FEATURES OF A HIGHLY HETEROGENEOUS DISEASE

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Background: Giant cell arteritis is the most common chronic systemic vasculitis in older adults. Permanent visual loss is the most concerning complication can occur in up to 20% of patients (1). Diagnosis is based on a combination of clinical findings, laboratory evidence of inflammatory markers, and temporal artery biopsy (TAB) confirming histologic evidence of inflammation. There are no definitive markers of the disease when TAB is negative.

Objectives: The objective of this study was to illustrate the clinical features of subjects suspected with GCA evaluated at the Dallas Veterans Affairs Medical Center (VAMC) and identify those features that would differentiate between TAB positive GCA, TAB negative GCA and non-GCA patients.

Methods: This retrospective study searched the Dallas VAMC database for subjects between January of 2010 until December 2019 with ICD-9 and ICD-10 entry code for GCA. Based on the 1990 ACR clinical classification criteria for GCA, suspected patients were classified as TAB positive GCA, TAB negative GCA and non-GCA. Subjects with incomplete data needed for classification were excluded. Group comparisons were performed with Fisher's exact test for categorical variables and the Mann-Whitney test for continuous variables.

Results: One-hundred and sixty-nine subjects had ICD-9/10 entry code for GCA. Ophthalmology was the specialty who did the initial evaluation in 46% of the

suspected cases. Seventy-one patients were excluded due to incomplete data. Of the remaining 98 patients, 42 (42.9%) were diagnosed with GCA out of which 10 patients had a positive TAB. Seventy-six percent of patients diagnosed with GCA had negative temporal artery biopsies. In 56 (57.1%) patients GCA was ruled out (Figure 1). New-onset headache was the predominant symptom suggestive of GCA affecting 88.9% TAB positive and 100% of TAB negative cases compared to 56.6% of non-GCA patients ($p < 0.001$), followed by scalp tenderness in 33.3%, 58.1% and 10.9%, respectively ($p < 0.001$), jaw claudication in 57.1%, 34.5% and 14.9% ($p = 0.021$), and elevated sedimentation rate in 33%, 65.6% and 33% ($p = 0.013$) (Table 1). Polymyalgia rheumatica was present in 28.6% and 21.7% of patients diagnosed with GCA compared to zero percent in non-GCA cases ($p = 0.017$). More than 30% of patients with GCA had normal acute phase reactants in this cohort.

Table 1. Comparison of demographics and clinical features of patients with suspected GCA

	GCA TAB Positive (n=10)		GCA - TAB Negative (n=32)		non-GCA (n=56)		
	n	%	n	%	n	%	p
Age (mean/sd)	71.3 (7.8)		67.3 (8.1)		67.3 (8.9)		0.367
Male	9	90	30/32	93.8	51/56	91.1	0.885
Clinical findings							
New onset headache	8/9	88.9	32/32	100	30/53	56.6	<0.001
Visual symptoms	9/10	90	22/32	68.8	42/54	77.8	0.351
Jaw claudication	4/7	57.1	10/29	34.5	7/47	14.9	0.021
PMR	2/7	28.6	5/23	21.7	0/30	0	0.017
Scalp tenderness	2/6	33.3	18/31	58.1	5/46	10.9	<0.001
Fever	0/5	0	5/23	21.7	2/35	5.7	0.117
ESR > 50	1/3	33.3	21/32	65.6	18/54	33.3	0.013
CRP > 10	1/3	33.3	5/30	16.7	6/54	11.1	0.473
Anemia (hemoglobin < 12 g/ dl)	2/7	28.6	15/32	46.9	21/52	40.4	0.642
Thrombocytosis (platelets >400)	2/7	28.6	3/32	9.4	4/51	7.8	0.228

Conclusion: There is clinical heterogeneity within the patients diagnosed with GCA regardless of TAB. A high index of clinical suspicion needs to be the cornerstone of diagnosis. There is need for new classification criteria to include patients with negative TAB.

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Disclosure of Interests: Adela Castro: None declared, Jiby Mathew: None declared, Ola Azzouqah: None declared, Jesus Diaz: None declared, Andreas Reimold Consultant of: Lilly, Grant/research support from: Abbvie Pfizer

Gilead, Kyawt Shwin: None declared

DOI: 10.1136/annrheumdis-2021-eular.1461