MPA and 1 with GPE (Patients characteristics are described in Table 1). From the 59 patients with GPA according to clinical judgment, 34 fullfilled the ACR 1990 criteria (57,63%) whereas 51 (86,44%) fullfilled the ACR/EULAR 2007 criteria. From the 33 patients diagnosed as having MPA 9 (27,27%) and and 3 (9.09%) fulfilled classification criteria for GPA according to ACR 1990 criteria and 2017 ACR/EULAR preliminary criteria, respectively.The patient with GPE did no classify as GPA by either set of criteria.The values for Sensibility, Espicifity, LR+ LR-, PPV and NPV in our population for both Criteria sets, are described in table 2.

Table 1. Patient characteristics

	N	Gender (F)	Age, Av (S.D.)	ACR 1990 + for GPA	ACR/ EULAR 2017 + for GPA
GPA	59 (63.44%)	30 (50.85%)	46.54 (13.32)	34 (57,63%)	51 (86,44%)
MPA	33 (35.48%)	23 (69.7%)	62,72 (2.11)	9 (27,27%)	3 (9,09%)
GPE	1 (1.08%)	1	62	0	0
Total	93	39 (41.94%)	52,45 (14.97)	43 (46,24 %)	54 (58,06 %)

Table 2.- Performance of ACR 1999 and ACR/EULAR preliminary Criteria for GPA in Chilean Patients

GPA Clasification Criteria	ACR 1990	ACR/EULAR 2017, preliminary Criteria
Sensitivity	57,6%	91,5%
Especificity	70,6%	91,2%
Positive Predictive Value	77,3%	94%
Negative Predictive Value	49%	86%
Positive Likelihood Ratio	1,96	10,3
Negative Likelihood Ratio	0,6	0,9
accuracy	0,62	0,91

Conclusions: In our population, provisional 2017 ACR/EULAR criteria for classificatión of GPA have better acurracy than ACR 1990 classification criteria.

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AB0591 EFFICACY AND SAFETY PROFILE OF INTRAVENOUS CYCLOPHOSPHAMIDE TREATMENT IN ELDERLY PATIENTS WITH SYSTEMIC VASCULITIS

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Background: Intravenous cyclophosphamide is mainstay of remission induction and dose reduction of glucocorticoid in patients with systemic vasculitis. However, little evidence has yet shown the safety profile of intravenous cyclophosphamide, especially in elderly patients.

Objectives: To evaluate efficacy and safety of patients diagnosed as systemic vasculitis and treated with intravenous cyclophosphamide.

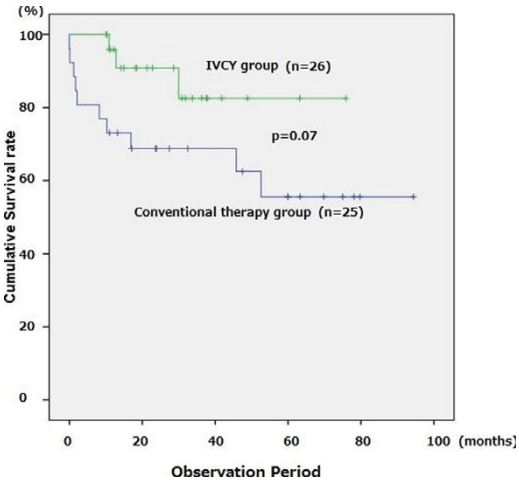
Methods: This retrospective study comprised the patients with active systemic vasculitis who were admitted to Kitami Red Cross Hospital and Obihiro-Kosei General Hospital from April 2009 to March 2016. These patients were treated with intravenous cyclophosphamide plus conventional therapy (IVCY group) or only with conventional therapy (glucocorticoid/azathioprine/tacrolimus and methotrexate) (conventional therapy group). The patients treated with oral cyclophosphamide or rituximab were excluded. Primary endpoint was defined as death or serious infections. Prognostic factors in IVCY group were analyzed by multivariate Cox regression methods.

Results: This study comprised 90 patients with active systemic vasculitis (61 microscopic polyangiitis, 9 eosinophilic granulomatosis with polyangiitis, 10 granulomatosis with polyangiitis, and 10 polyarteritis nodosa). Fifty-one patients were over 70-year-old (26 patients in IVCY group). The mean observation period was 30.1 months. IVCY group had a trend for higher event-free survival rate as compared with conventional therapy group (p=0.19). IVCY group had significantly higher cumulative survival rate as well (p=0.04). Age (HR=1.07, 95% CI 1.01–1.14, p=0.03), five factor score (FFS, HR=3.39, 95% CI 1.36–8.44, p=0.01) and glucocorticoid dose at 24 weeks (GC24, HR=1.14, 95% CI 1.05–1.22, p=0.001) were identified as risk factors for events. In patients over 70-year-old, IVCY group had a trend for higher cumulative survival rate as well (p=0.07, Figure1). FFS (HR=3.30, 95% CI 1.19–9.15, p=0.02) and GC24 (HR=1.10, 95% CI 1.02–1.20, p=0.01) were identified as risk factors for events.

Table 1. Background of patients with IVCY group and conventional therapy group

	IVCY group (n=54)	conventional therapy group (n=36)	P value
MPA,n (%)	36 (67%)	25 (69%)	0.95
EGPA,n (%)	6 (11%)	3 (8%)	0.56
GPA,n (%)	8 (15%)	2 (6%)	0.36
PN,n (%)	4 (7%)	6 (17%)	0.19
age (y)	66.4±15.5	72.3±15.9	0.09
Male,n (%)	19 (35%)	13 (36%)	0.93
FFS (d=0)	0.6±0.7	0.7±0.8	0.40
BVAS (d=0)	10.9±5.5	10.5±6.4	0.71
PSL (mg, d=0)	50±11	38±13	0.39
PSL pulse,n (%)	18 (33%)	22 (61%)	0.01
AZA,n (%)	13 (24%)	14 (39%)	0.10
MTX,n (%)	2 (4%)	4 (11%)	0.17

Abbreviation: MPA (microscopic polyangiitis), EGPA (eosinophilic granulomatosis with polyangiitis), GPA (granulomatosis with polyangiitis), PN (polyarteritis nodosa), BVAS (Birmingham Vasculitis Activity Score), AZA (azathioprine), MTX (methotrexate), d = 0 means day 0; the start of the treatment.



Conclusions: Intravenous cyclophosphamide treatment had acceptable safety profile even in elderly patients.

Disclosure of Interest: None declared

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AB0592 PULMONARY ARTERY ANEURYSM IN BEHÇET'S DISEASE: RETROSPECTIVE MONOCENTRIC TUNISIAN STUDY

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Background: Behcet's disease (BD) is a chronic inflammatory disorder. Arterial inflammatory involvement includes predominantly aortic and pulmonary aneurysmal lesions, affects about 10% of patients with BD. They account for the severity of the disease and are a leading cause of death

Objectives: To investigate the frequency of Behcet's disease with pulmonary artery aneurysm (PAA). We aimed to review PAA and other systemic involvements associated with PAA in BD and to provide a review of diagnostic techniques, treatment and prognosis

Methods: 243 BD patients were recruited for this study (152 men, 91 women, mean age 31.7±7 years. Diagnosis of BD was made according to the international study group for Behçet's disease [International Study Group for Behçet's Disease, lancet 1990; 335: 1078–80]. All patients underwent full clinical examination, routine laboratory investigations. Chest X-rays and pulmonary CT angiography were performed on all patients with pulmonary involvement.

Results: Eight of the patients have pulmonary aneurysm, all of them are male, mean age 32.6±13, The mean disease duration until PAA appear was 2.8±3.5 years. The main pulmonary symptoms were as follows: dyspnea 87%, cough 50%, hemoptosis: 75%, fever 37%. Other systemic involvements associated PAA are as follows: buccal (100%) and genital (75%) ophthalmic 25%, neurological 50%, cardiac 25%. 3 patients presented with Hughes Stovin syndrome. The treatment includes corticosteroids, colchicine and immunosuppressant agents (Cyclophosphamid or azathioprine), only two patients reveive coil embolization. At follow up for a median of 4 years (1 to 25 years), three patients died because they stopped their medication.

Conclusions: The prognosis of PAA is poorer than other lesions involved in BD, treatment (immunosuppressant agents, colchicine) seems to improve the prognosis. It is important to maintain the immunosuppressive therapy and a regular follow-up to prevent these complications

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AB0593 ANCA-ASSOCIATED VASCULITIS WITH BOTH MPO-ANCA AND PR3-ANCA SHARES CHARACTERISTICS OF ANCA-ASSOCIATED VASCULITIS WITH SINGLE ANCA

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Background: The anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides are heterogeneous group of necrotizing inflammation of small vessel and the presence of the ANCA. ANCAs are defined according to the target antigens, leukocyte proteinase 3 (PR3) and myeloperoxidase (MPO). Recently, the ANCA specificity could be better for classification of ANCA-associated vasculitides than the clinical diagnosis. A few patients have both MPO- and PR3 ANCA. However, the clinical characteristics of these patients were not known in detail.
Objectives: To analyze organ involvement of patients with ANCA-associated vasculitis according to ANCA type focusing both MPO- and PR3-ANCA (both-ANCA) positive vasculitis
Methods: The medical records of the patients with positive ANCA and clinical diagnosis or the patients with positive ANCA and vasculitis diagnosis confirmed by biopsy were reviewed at two regional tertiary hospitals. The age at diagnosis, sex, and the organ involvement of kidney, lung, upper airway (nose/sinus/ear), skin, peripheral nervous system, central nervous system, and gastrointestinal tract were collected. The clinical variables were analyzed by ANCA type.
Results: Total 82 patients with positive ANCA and clinical diagnosis or histologic diagnosis of vasculitis were searched. MPO-ANCA positive patients was 63 (76.8%), PR3-ANCA 9 (11.0%), and both MPO- and PR3-ANCA was 10 (12.2%). The age at diagnosis of patients with PR3-ANCA was younger than patients with MPO-ANCA or both-ANCA (PR3-ANCA, 49.6 vs. MPO-ANCA, 66.1 vs. both-ANCA, 62.1, $p<0.05$). Moreover, kidney involvement were MPO-ANCA was 77.8%, PR3-ANCA 22.2%, and both-ANCA 80% ($p<0.05$). Upper airway involvement was also significantly associated with ANCA type (PR3-ANCA, 66.7% vs. MPO-ANCA, 23.8% vs. both-ANCA, 50.0%, $p<0.05$). The involvement of skin, central or peripheral nervous system, gastrointestinal tract or the presence of lung fibrosis and lung nodule or mass did not differ according to ANCA type.
Conclusions: ANCA-associated vasculitis with both MPO-ANCA and PR3-ANCA has more kidney involvement than ANCA-associated vasculitis with PR3-ANCA and more upper airway involvement than ANCA-associated vasculitis with MPO-ANCA.
Disclosure of Interest: None declared
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AB0594 RELATION OF CERTAIN BLOOD PICTURE PARAMETERS AND VASCULAR ENDOTHELIAL GROWTH FACTOR TO CLINICAL MANIFESTATIONS AND DISEASE ACTIVITY IN BEHÇET'S DISEASE PATIENTS

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Background: Behçet's disease (BD) is a systemic inflammatory condition sharing the clinical features of both auto- inflammatory diseases and variable vessel vasculitis. Endothelial dysfunction (ED) plays an important role in the pathogenesis of BD. Several markers can be used to evaluate ED as mean platelet volume (MPV), red cell distribution width (RDW), neutrophil-lymphocyte ratio (NLR) and vascular endothelial growth factor (VEGF).
Objectives: The aim of the present study was to measure the MPV, RDW, NLR and serum level of VEGF in BD patients and to study their relation with disease manifestations and activity.
Methods: Ninety six BD patients and 60 matched controls were enrolled in this study. The MPV, NLR, RDW and serum VEGF level were measured. Disease activity was assessed by the BD Current Activity Form (BDCAF). The influence of an associated metabolic syndrome (MetS) was also considered.
Results: The mean age of the 96 patients was 34.9±10.1 years (18–73 years), male:female 4.7:1 and the disease duration was 9±7 years (0.6–40 years). MetS was present in 13.7%. Two patients were siblings and 5 had juvenile-onset BD. The RDW and NLR were significantly higher in patients (15.5±2% and 2.7±2.9) than controls (14.3±1.03% and 1.5±0.8) ($p<0.001$ each), while the MPV and VEGF were comparable. The MPV was significantly decreased in patients with vascular involvement (p=0.04) and increased in those with psychiatric disorders (p=0.02). The RDW was significantly higher in patients with vascular involvement (p=0.04) especially those with venous thrombosis and in those with neurological manifestations (p=0.03). The NLR was higher in males (p=0.01) and in those with retinal vasculitis (p=0.03) and vein occlusion (p=0.02). None of the parameters significantly correlated with the BDCAF. However, the NLR was the most valuable parameter to predict disease activity at a cut-off level of 1.69 ng/L (sensitivity 75%, specificity 55.6%). The MPV significantly correlated with the body mass index (BMI) (p=0.008), cholesterol (p=0.01) and low density lipoprotein (LDL) ($p<0.001$); the RDW correlated with the erythrocyte sedimentation rate (ESR) (p=0.003) and total leucocytic count (TLC) (p=0.04); the NLR with TLC (p=0.001) and blood urea (p=0.001) and VEGF with the TLC (p=0.048) and high density lipoprotein (HDL) (p=0.02). None of the parameters was significantly different according to the presence of MetS. Regarding the medications received, the RDW was significantly higher in patients who received cyclophosphamide and warfarin than those who did not (p=0.003 and $p<0.001$ respectively) and the level of VEGF tended to be lower in patients who received colchicine (p=0.06).
Conclusions: In BD, only the RDW and NLR were significantly increased raising the possibility of a potential role in the disease susceptibility and pathogenesis with no obvious relation to the disease activity or to an associated MetS. Together with the serum VEGF, they may all serve as useful markers to reveal the pattern of organ involvement while the NLR was the most valuable cost effective parameter to signify the disease activity. The influence of medications warrants further studies.
Disclosure of Interest: None declared
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AB0595 NOVEL APPROACHS BASED IN CURRENT EVIDENCE IN ANCA-ASSOCIATED VASCULITIS WITH RENAL INVOLVEMENT TREATMENT

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Background: ANCA-associated vasculits (AAV) are a group of multi-system autoinmune diseases characterized by inflamation and necrosis in small and medium vessels.AAV could respond to diferent therapeutics protocols depends on diverse levels of clinical severity and early treatment could improve the outcome of the disease.In spite of recognized efficay of regimens consisting of cyclophosphamide and high-dose corticosteroids to control the AAV,efforts to minimize drugs-related toxicity led to consider targeted therapies.
Objectives: Considering the currently quality evidence in therapies novely proposed to AAV and the severity of renal disease presentation,we suggest new rational approaches emphasizing targeting B-cells therapy and preventing disease relapse
Methods: We identified the latest quality evidence using methodological search filters,assessed the evidence quality with Cochrane Renal Goup check list and determinated the strenght of recommendations by Levels of Evidence (Oxford Centre for Evidence-based Medicine)
Results: Rituximab (RTX), a monoclonal anti-CD 20antibody, has emerged as the biologic agent more using in AAV patients in current publications and unlike latest Guides and Recommendations published,RTX would be recommended in induction and maintenance AAV with renal involvement treatment (Table 1).

Table 1

AAV Induction Therapy	Early systemic (GF>60 ml'):MTX + GC (Ib,B) Severe generalised (Cr>5.68 mg/dl):CFM IV/PO + MPS (Ia,A) Generalised with contraindication to CFM:RTX + GC (Ib,B) Prophylaxis against *Pneumocystis jirovecii* (in CFM or RTX therapy):Cotrimoxazol PO (IIb,B) Severe with RPGN: Plasma Exchange-adjuvant therapy (Ia, B)
AAV Maintenance Therapy	Low-dose GC + AZA up 18 months (IIb,B) Low-dose GC + LF (less safer) (Ib,B) Low-dose GC + MTX with GF >60 ml/'(Ib,B) Avoid use CFM long-term (higher relapse risk) (Ia,A) In GPA:RTX + GC low-dose each 6 months up18 months (Ib, B)
AAV Relapse	Minor Relapse:increase GC dose (IIb,C) Major Relapse:RTX + GC (Ia,A) Major Relapse with CFM cumulative dose <36 gr:CFM + GC (Ib,B) Plasma Exchange and/or MPS (IIb,C)
VAA Refractaria	RTX + GC,specially patients whose never recived RTX (II,B) Plasma Exchange in RPGN and/or dialysis-dependen (Ia,B)

GF: glomerular filtrate, MTX:metotrexate, GV: glucocorticoids, Cr: creatinine, CFM: ciclophosphamide, IV: intravenous, PO: oral, MPS: metilprednisolone, RTX: rituximab, RPGN: rapidly progressive glomerulonephritis, AZA: azathioprine, LF: leflunomide, GPA: granulomatosis with polyangiitis.

Conclusions: Current therapeutical protocols for AVV with renal involvement show that emerging therapies like RTX could improve rates of relapses and treatment-related toxicity. Further studies would provide target-therapeutical options
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