Conclusion: The present targeted NMR based metabolomics study demonstrated that the serum metabolic profiles of amino acid may help guide therapy and ratio of Val/Tyr can differentiate active from inactive patients. However, future studies on large patient cohorts are required to establish its clinical utility.

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SAT0226 INFLIXIMAB FOR THE TREATMENT OF REFRACTORY POLYARTERITIS NODOSA

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Background: Polyarteritis Nodosa (PAN) is a necrotizing vasculitis predominantly affecting medium and small size arteries (1). Cyclophosphamide, a drug with narrow therapeutic range and poor safety profile, constitutes the treatment of choice for PAN vasculitis with major organ involvement. (2)

Objectives: To describe our clinical experience in treating refractory PAN with infliximab (a TNF inhibitor), a drug with good tolerability and better safety profile than cyclophosphamide.

Methods: Twenty-six PAN patients were admitted to our rheumatology unit between 2006 and 2017, of whom 9 patients, with severe and refractory disease, were treated with infliximab after failure of standard treatment.

We describe herein the patients' characteristics, clinical manifestations, severity and response to infliximab treatment and review the current literature.

Results: Complete remission was defined as the absence of features of active disease and withdrawal of prednisone therapy. Significant improvement was defined as clinical improvement and prednisone dose reduction of at least 50% or a 50% reduction in immune modulatory medications other than prednisone.

After 4 months of treatment, 8/9 (89%) patients achieved significant improvement, with two of them achieving complete remission.

Conclusion: We suggest anti TNF agents, and in particular infliximab, are relatively safe and efficacious treatment options in refractory PAN. A randomized controlled trial should be done in order to objectively evaluate infliximab in PAN.

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SAT0227 MYOCARDIAL FIBROSIS IS ASSOCIATED WITH ANCA STATUS, ORGAN INVOLVEMENT AND DISEASE SEVERITY IN PATIENTS WITH GRANULOMATOSIS WITH POLYANGIITIS

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Background: Granulomatosis with polyangiitis (GPA) is a systemic vasculitis with an increased burden of cardiovascular (CV) events compared to

the general population, but the prevalence of primary cardiac involvement in this disease is unknown.

Objectives: The objective of this prospective study was to describe with cardiac magnetic resonance (CMR) the myocardial abnormalities associated with GPA and their correlations with disease characteristics.

Methods: Twenty-six patients with GPA and no prior CV disease (CVD) or diabetes mellitus underwent contrast-enhanced CMR, including late gadolinium-enhancement (LGE), T1 mapping for native T1 and extra-cellular volume (ECV) quantification for assessment of myocardial fibrosis and tissue tagging for assessment of left ventricular (LV) function. Characteristics of disease, treatments and CV risk factors were collected; disease relapse was defined as reappearance or worsening of vasculitis symptoms requiring an increase of the current treatment or introduction of additional immunosuppressive medication. Twenty-five healthy volunteers (HV) with comparable age, sex, BMI and arterial blood pressure served as controls.

Results: Patients with GPA (median age 58 years, disease duration 8 years, BVAS 2; females 46%, PR3 ANCA 58%) had similar cardiovascular risk profile to HV. A focal, non-ischemic LGE pattern of fibrosis was detected in 24% of patients and no controls (p=0.010; Table 1). Patients with myocardial LGE were more frequently PR3 ANCA negative (93% vs 7%, p=0.007), and they presented more frequently with involvement of the lower respiratory tract (75% vs 25%, p=0.097) and skin (63% vs 38%, p=0.087). Values of LGE-related fibrotic mass were higher in patients presenting with renal involvement (p=0.036). Native T1 and ECV were higher in patients with GPA than HV (Table 1); ECV was higher in those with relapsing disease, and native T1 was inversely associated with PR3 ANCA ($\beta=-0.664$, p=0.001). Peak systolic strain was slightly reduced in GPA than controls (Table 1); LV ejection function (LVEF) was inversely correlated with disease duration ($\beta=-0.454$, p=0.026).

Table 1. CMR measures†

LV volumes and mass			
LV end diastolic volume index, mL/m ²	69 (61, 84)	79 (70, 89)	0.105
LV end systolic volume index, mL/m ²	28 (23, 37)	32 (28, 39)	0.122
LV mass index, g/m ²	37 (42, 47)	50 (42, 56)	0.014
LV mass/LV end diastolic volume, g/mL	0.59 (0.48, 0.66)	0.59 (0.54, 0.66)	0.486
LV function			
LV ejection fraction, %	59 (56, 63)	58 (53, 62)	0.351
LV peak systolic strain			
Base	-0.19 (-0.21, -0.16)	-0.21 (-0.22, -0.19)	0.017
Mid	-0.21 (-0.23, -0.20)	-0.22 (-0.23, -0.20)	0.298
Apex	-0.21 (-0.23, -0.16)	-0.23 (-0.25, -0.20)	0.101
Torsion, deg	12.3 (9.2, 14.5)	13.8 (11.6, 16.0)	0.086
Peak twist	14.1 (9.0, 15.7)	14.1 (11.7, 17.7)	0.222
Myocardial tissue characterization			
Native T1 (ms)	1225 (1199, 1255)	1203 (1185, 1233)	0.006
Extracellular volume, %	25.0 (23.3, 27.2)	23.6 (20.5, 25.1)	0.009
LGE, n (%)	6/25 (24)	0/25 (0)	0.010
LGE scar tissue, g	1.25 (0.81, 2.63)	0 (0, 0)	NA

†Continuous data are presented as median (interquartile range).

Conclusion: Patients with GPA had significant myocardial abnormalities on CMR compared to HV. ANCA status, systemic organ involvement and disease severity were associated with CMR markers of myocardial fibrosis. CMR could be a useful tool for identification and future risk stratification of myocardial involvement of GPA.

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SAT0228 LEUKOCYTE DYNAMICS IN GIANT CELL ARTERITIS AND POLYMYALGIA RHEUMATICA PATIENTS BEFORE, DURING AND AFTER TREATMENT

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Background: Giant cell arteritis (GCA) and polymyalgia rheumatica (PMR) are inflammatory diseases requiring long-term treatment with