Results: From December 2018 to November 2019, 230 AAV patients were recruited in 6 non-academic and 3 academic hospitals (120 vs 110 patients respectively). Differences in clinical diagnoses (GPA, MPA and eGPA) were observed between non-academic and academic centers ($p=0.05$), which was mainly caused by a higher number of MPA patients in non-academic centers. The year of diagnosis was comparable (median 2013 [2009-2016], $p=0.150$). The median follow up since diagnosis was 4.8 years [1.8-9.6] with a median in-hospital time-to-diagnosis of 13 days [2-50]. Patients were diagnosed at a mean age of 63 years (± 11.18) in non-academic centers and 53 years (± 16.92) in academic centers ($p<0.001$). Besides steroids, oral cyclophosphamide was the most preferred drug (54%) for induction therapy, whereas rituximab was given significantly more often as (part of the) induction therapy in patients treated in academic centers compared to patients in non-academic centers (27% vs 8%, $p<0.001$). In non-academic centers pneumocystis pneumonia (PCP) prophylaxis was prescribed significantly less (76% vs 91%, $p=0.003$). Also, screening for *Staphylococcus aureus* carriage was significantly less (17% vs 68%, $p<0.001$). With respect to mortality and co-morbidity, 23 patients (10%) died, 100 patients (44%) had at least one infection and 24 patients (10%) suffered from at least one malignancy. We observed no significant differences on these endpoints between academic and non-academic centers.

Conclusion: The present study highlights important practice variation in the management of AAV between academic and non-academic hospitals in the Netherlands. A high proportion of patients is treated with oral cyclophosphamide as induction therapy while rituximab is increasingly used in academic centers. Rates of mortality, infections and malignancies were not different. Altogether, this study raises awareness into the variation of management for AAV patients and allows the identification of areas for improvement of clinical care for Dutch AAV patients.

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THU0305

PREVALENCE AND CLINICAL OUTCOME OF INTERSTITIAL LUNG DISEASE IN ANCA ASSOCIATED VASCULITIS

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Background: Lung involvement is frequent in ANCA-associated vasculitis (AAV). Classical lung manifestations consist of capillaritis with lung haemorrhage, inflammatory infiltrates and nodules. Interstitial lung disease (ILD) is increasingly recognized among patients with AAV. However, little is known concerning risk factors and clinical course of these patients.

Objectives: The aim of our study was to characterize the prevalence and clinical course of ILD in patients with AAV.

Methods: We have performed a clinical retrospective single-centre observational analysis (1990-2019) of all patients with the diagnosis of microscopic polyangiitis (MPA) and granulomatosis with polyangiitis (GPA) diagnosed according to 2018 Draft Classification Criteria for GPA and MPA¹. Demographic, clinical and immunologic data were reviewed. Radiologic pattern of ILD were assessed by high-resolution-CT. Main outcome evaluated was overall survival.

Results: The study population consisted of 123 patients, 56% female, aged 59.3 $\pm$ 18.2 years old at the time of diagnosis. Clinical diagnosis was of MPA in 54% of patients and GPA in 46%. While 108 (88%) ANCA positive patients had PR3 (n=25) or MPO (n=83), 15 (12%) patients had negative or atypical ANCA. Any lung involvement was present in 82 (71%) and ILD was identified in 24 (20%) of all patients. ILD pattern was of usual interstitial pneumonia (UIP) in 12 patients, non-specified interstitial pneumonia (NSIP) in 9 and chronic organizing pneumonia (OP) in 3. There was an association between the presence of ILD and

ANCA specificity: MPO were present in 100% of patients with UIP and in 75% of patients with NSIP/OP ($p=0.017$). Bronchiectasis were more prevalent among patients with ILD (19/24; $p<0.001$). During the median follow-up time period of 68 (23-126) months, mortality was of 42% among patients with ILD-AAV compared with 11% in no ILD-AAV (log-rank $p=0.0001$). On the multivariate Cox regression model, ILD was an independent predictor of mortality HR 2.95 (95%CI 1.09-7.96; $p=0.033$).

Conclusion: ILD is a frequent manifestation of MPA and GPA patients. The presence of ILD, particularly UIP, is associated with ANCA-MPO and is a predictor of mortality. Therefore, a better management of fibrotic lung involvement in AAV is warranted.

References:

- Robson JC, Grayson PC, Ponte C, et al. Draft classification criteria for the ANCA associated vasculitides. *Ann Rheum Dis* 2018;77 (suppl 2):60-1.

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THU0306

ROLE OF 18-FDG PET/CT IN DIAGNOSIS AND FOLLOW UP OF LARGE VESSEL VASCULITIS

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Background: 18-FDG PET/CT is a functional imaging method which allows to identify inflammation of vessel walls. The use of PET in large vessels vasculitis (LVV) at disease onset and during follow up is still debate either to confirm clinical remission either to drive the therapy choice. American Society of Nuclear Cardiology (ASNC) recently advanced recommendations aimed to standardize the application of PET in LVV(1).

Objectives: The aim of our study was to assess the clinical role of PET performed in patients affected by LVV at the diagnosis and during the follow up.

Methods: We retrospectively evaluated PET/CT of 49 patients affected by clinically active LVV according to LVV visual grading (LVG, grading 0-3) and measured the standardized uptake value(SUV) of large vessels. 38 (77.6%) patients were affected by Giant Cells Arteritis and 11(22.4%) by Takayasu Arteritis. 32(65.3%) patients repeated the imaging after a mean follow-up of 11.5 $\pm$ 5.4 months.

All baseline (T0) and follow up (T1) clinical data of disease activity were collected. Patients were treated according to EULAR LVV management recommendations(2). T0 PET/CT study was performed in patients with a clinically active disease defined by suggestive symptoms/signs and/or high inflammatory markers. The mean disease duration before T1 PET/CT examination was 4 months. T0 PET was performed in 25/49 patients(52%) at the diagnosis of

LVV, whereas in 24/49(48%) patients with already diagnosed but active LVV disease.

Results: Baseline PET was positive in 21 patients(42.9%). According to ASNC recommendations, 19 patients (38.8%) presented a LVG=3, 2(4.0%) a LVG=2, 6(12.2%) LVG=1 and 22 (44.9%) LVG=0. Patients performing PET at disease onset(75%) had higher LVG score than patients performing PET during the disease course (25%), $p=0.002$. At T0, aortic, carotid, axillary and subclavian SUV did not correlate with inflammatory markers.

Follow up PET/CT studies were performed in 32 patients, 13 (40.6%) with a clinically active disease despite therapy, while 19(59.4%) in clinical remission. Follow up PET was still positive in 8 patients (25%) with a LVG=3, 10 (31.2%) patients presented LVG=1 and 14 (43.8%) LVG=0. T1 PET/CT study showed a significant reduction of SUV values in descending aorta, left and right subclavian arteries, and left and right axillary arteries when compared with first PET/CT study. According to LVG, 12 patients with active PET/CT study at T0 (19 pts) presented a reduction of LVG from score 2 and 3 to grade 1 or 0 (64.2%) at second PET/CT study. Only 3 patients presented an increased LVG score at T1, while in the other 17 patients T1 PET confirmed the previous score. No significant difference was found between LVG scores according with clinical characteristics, but among 8 patients presenting an active T1 PET, 4(50%) were in clinical remission.

Conclusion: The use of ASNC recommendations for FDG PET/CT in LVV enables to confirm a metabolically active disease in 40% of patients and in 75% of patients at disease onset, suggesting that post-posing the exam could lead to underestimate the real extension of disease. Our data, even if limited, suggest that PET/CT could be crucial in management of patients in clinical remission, detecting patients with still metabolically active LVV. Further prospective studies are necessary to evaluate the role of PET/CT in driving therapeutic strategies.

References:

[1] Slart R et al - Eur J Nucl Med Mol Imaging, 2018

[2] Hellmich et al - Ann Rheum Dis 2018

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THU0307

RESPONSE OF BEHCET'S REFRACTORY ORAL AND/OR GENITAL ULCERS TO APREMILAST IN COMBINATION VS MONOTHERAPY. NATIONAL MULTICENTER STUDY OF 51 CASES OF CLINICAL PRACTICE

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Background: Apremilast (APR) has demonstrated efficacy in the treatment of oral and/or genital aphthous ulcers in Behçet's disease (BD). Combination of APR to other disease-modifying anti-rheumatic drugs (DMARDs) has not been assessed.

Objectives: To compare the efficacy and safety of APR in monotherapy or combined with DMARDs in refractory BD.

Methods: National multicenter open-label study on 51 BD patients with oral and/or genital ulcers refractory to conventional treatment.

Results: We included 51 patients (35 women/16 men), mean age 44.7±13.2 years. Before APR, all patients had received several systemic conventional drugs. The main clinical symptoms for starting APR were oral (n=19) and genital (2) aphthous ulcers or both (30).

Excluding corticosteroids, colchicine or NSAIDs, APR was given at standard dose of 30mg twice daily in monotherapy (n=31), or combined with conventional DMARDs in 16 cases (6 azathioprine, 5 methotrexate, 4 hydroxychloroquine, 4 sulfasalazine, 1 dapsone) or with biologic DMARDs in 4 (2 tocilizumab, 1 adalimumab, 1 infliximab). There were not found statistically significant differences in demographic features, previous therapy, clinical manifestations or reported adverse effects.

After a median follow-up of 6 [3-12] months, most of the patients experienced improvement of the orogenital ulcers in both groups (89.8% in the first 2 weeks), without statistically significant differences. (TABLE)

Conclusion: APR leads to a rapid and maintained improvement in most patients with refractory BD orogenital ulcers. APR seems as effective and safe in monotherapy as combined.

TABLE:

	Week 1-2		Week 4		Month 6		Month 12		Month 24	
Outcome of oral and/or genital ulcers n, (%)	C n=19	M n=30	C n=19	M n=26	C n=12	M n=17	C n=7	M n=6	C n=1	M n=1
Complete resolution	8 (42.1)	11 (36.7)	12 (63.2)	20 (77)	7 (58.4)	14 (82.4)	3 (42.8)	3 (50)	1 (100)	1 (100)
Partial resolution	9 (47.4)	16 (53.4)	7 (36.8)	3 (11.5)	5 (41.6)	2 (11.7)	4 (57.2)	3 (50)	0	0
No response	2 (10.5)	3 (9.9)	0	3 (11.5)	0	1 (5.9)	0	0	0	0
p value	0.9		0.1		0.1		0.8		0.7	

Abbreviations: C= combined; M= monotherapy; n= available data.

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THU0308

COMPARISON OF CHILDHOOD-ONSET VERSUS ADULT-ONSET TAKAYASU ARTERITIS: A STUDY OF 141 PATIENTS FROM TURKEY

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