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 (±11.8) 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 carrierstatus was significantly less (17% vs 68%, p<0.001). With respect to mortality and co-morbidity, 22 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 in the period 1990-2019. All baseline (T0) and follow up (T1) clinical data of disease activity were collected. Radiologic pattern of ILD were assessed by high-resolution-CT. Main outcome evaluated was over-all survival.

Results: The study population consisted of 123 patients, 56% female, aged 59±14.2 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). Bronchiolitis 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 GPA and MPA 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.

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THU0306 ROLE OF 18-FDG PET/CT IN DIAGNOSIS AND FOLLOW UP OF LARGE VESSELS 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±4,5 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

Graph 1. Survival analysis of patients with AAV according to the presence of ILD

Patients with ILD-AAV have decreased survival when compared with patients with AAV and no ILD. Log-rank test p=0.0001

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LVV, whereas in 24/49 (49.4%) 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 (4.2%) 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%).

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 to 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 to 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 underrate 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:

Disclosure of Interests: Laura Gigante: None declared, Dario Bruno: None declared, Vanessa Feudo: None declared, Silvia Laura Bosello Speakers bureau: Abbvie, Pfizer, Boehringer, Lucia Lecissotti: None declared, Alessia Musto: None declared, Laura Gigante: None declared, Dario Bruno: None declared. Laura Gigante has nothing to disclose.

TABLE:

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<th>Week 4</th>
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<th>Month 12</th>
<th>Month 24</th>
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<td>11 (36.7)</td>
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<td>Partial resolution</td>
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<td>5 (17.2)</td>
<td>3 (10.3)</td>
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<tr>
<td>No response</td>
<td>2 (6.7)</td>
<td>3 (10.3)</td>
<td>0 (0)</td>
<td>1 (2.0)</td>
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<td>0.8 (4.7)</td>
<td>0.7 (2.9)</td>
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Abbreviations: C= combined; M= monotherapy; n= available data.

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