Abstract FRI0481 – Figure 1. Interactions between GCA, IBD and covariates

Conclusions: The probability that GCA patients may also suffer from IBD is increased in comparison with age- and gender-matched controls. Our findings indicate that this association is most prominent in younger patients (<70). Screening for IBD amongst GCA patients in this age group may be warranted.

REFERENCES:

- [1] Colombat M, et al. Giant cell arteritis localized to the colon associated with Crohn's disease. *Histopathology* 2001;38(1):21–24.
- [2] Sy, Alice, et al. Vasculitis in patients with inflammatory bowel diseases: A study of 32 patients and systematic review of the literature. *Seminars in arthritis and rheumatism* 2016;45(4);Elsevier.
- [3] Sanges, Sébastien, et al. Giant cell arteritis and ulcerative colitis: An unusual association. *Seminars in arthritis and rheumatism* 2016;46(1) Elsevier.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3976

FRI0482

CAUSES OF DEATH IN CONNECTIVE TISSUE DISEASE (CTD'S) AND VASCULITIDES; DATA FROM THE NORWEGIAN CONNECTIVE TISSUE DISEASES AND VASCULITIS REGISTRY (NOSVAR)

T. Garen, K. Lerang, H. Andersson, Ø. Midtvedt, K. Kilian, S. Reister, R. Gunnarsson, B. Gudbrandsson, Ø. Molberg, Ø. Palm. *Department of Rheumatology, Oslo University Hospital, Rikshospitalet, Oslo, Norway*

Background: Mortality as an outcome of disease's severity and causes of death can give extended insight into the nature of each specific diagnose and highlight distinct needs for monitoring. In connective tissue diseases (CTD) and systemic vasculitides, mortality and causes of death have been studied within most single diseases, but with heterogeneity of the studies and widely varying results. Studies applying similar methods across the diagnoses are lacking. Consequently, it is difficult to compare causes of death between the different diagnoses.¹

Objectives: To identify the causes of death within different CTDs and systemic vasculitides.

Methods: We performed a prospective, observational, controlled study between 1999 and 2017 of adult patients (at least 18 years of age) with diagnosis of CTD or vasculitides. All patients were included in the Norwegian connective tissue disease and vasculitis Registry (NOSVAR). In total, 2140 patients were diagnosed and followed up until death or study end by April 31st 2017. To avoid bias by selection, we included only incident cases, excluding patients with diagnoses set prior to 1999. Moreover, cases with a disease course not consistent with the initial diagnoses were excluded. Causes of death were identified by linking NOSVAR to the Norwegian Causes of Death Registry and by reviewing hospital charts. We divided the causes of death into the main groups of cardiovascular diseases (CVD), neoplasms, chronic respiratory disease (CRD), infections and other (gastrointestinal, renal insufficiency and trauma). To compare causes of death to the general population we used data from WHO Mortality Database, Causes of death.²

Results: During a mean (SD) follow-up time of 9.2 years (4.7), 279 patients (13%) deceased. The major causes of death were, in descending order of frequency; CVD (27%), neoplasms (25%), CRD (16%), infections (11%), gastrointestinal manifestations (4%), renal insufficiency (2%). Data from the general population, adjusted for age and gender, showed that deaths by CVD, CRD and infections were more prevalent among the patients.² The leading causes of death are shown in figure 1. In Takayasu arteritis and ISSc, CVD was the most frequent cause of death; (56%) and (41%), respectively. More than half of the patients (53%) with antisynthetase syndrome died of CRD. Those with dermatomyositis died most frequently of neoplasms (50%).

Conclusions: Compared to general population, patients with CTD and vasculitis died more often of CVD, CRD and infections. CVD as a cause of death was most prevalent in patients with Takayasu arteritis, giant cell arteritis and systemic sclerosis, while neoplasm was the major cause of death in dermatomyositis. In antisynthetase syndrome, both CRD was the major causes of death. The study gives the clinician valuable information on how to monitoring the different CTDs and vasculitides regarding serious outcome.

REFERENCES:

- [1] Toledano E, Candelas G, Rosales Z, Martinez Prada C, Leon L, Abasolo L, et al. A meta-analysis of mortality in rheumatic diseases. *Reumatologia clinical* 2012;8(6):334–41.
- [2] University of Washington, Causes of Death (COD) visualization VizHub. [https://vizhub.healthdata.org/cod/University of Washington;2017](https://vizhub.healthdata.org/cod/University%20of%20Washington;2017) [updated 2017] cited 2017].

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.2031

FRI0483

SULFASALAZINE AS A POTENTIAL TREATMENT FOR IGA-VASCULITIS (HENOCH-SCHÖNLEIN PURPURA) IN ADULTS

S. Giliyev, A. Semenovykh. *I.M. Sechenov First Moscow State Medical University, Moscow, Russian Federation*

Background: Primary IgA-vasculitis and inflammatory bowel diseases (IBD) share many clinical, endoscopic, and radiological signs. It may suggest a common pathogenetic background for both pathological conditions. While sulfasalazine (SASP) is one of the well-known potential agents able to improve symptoms in IBD, the usefulness of SASP in treatment of the primary IgA-vasculitis remains unclear.

Objectives: Retrospective study to assess a therapeutic value of the SASP in primary IgA-vasculitis.

Methods: Totally 78 adult patients with primary IgA-vasculitis were enrolled in this study. Diagnosis was made on the basis of EULAR/PRES criteria¹ after thorough screening to exclude a secondary nature of the disease, including colonoscopy. Purpura/petechia was present in all patients being mandatory diagnostic criterion. Arthritis/arthralgia was seen in 48 patients (61.5%). There were no patients with abdominal syndrome just before enrollment, although 46 (59%) patients had transient abdominal pains in the history. Mild to moderate signs of renal involvement (hematuria and/or proteinuria) was seen in 35 (45%) patients without renal impairment. There were 20 (25%) drug-naïve patients, 40 (51%) patients after unsuccessful immunosuppressive treatment and 18 (23%) patients failed to respond to anticoagulants or antiplatelet agents. Initially SASP was prescribed in a daily dose 1 g followed with gradual titrating up to the 2 g/day depending on tolerability and clinical response. Most patients (96%) have been taking SASP longer than 6 month and about a half of the patients (56%) – longer than year. The longest treatment was 5 years.

Results: Complete clinical remission of the skin rash was achieved in 48 patients (58.9%). In 27 patients (35%), there was partial improvement of the skin eruptions, characterised with less quantity of the skin purpura or longer periods free of

symptoms (see table 1). Four patients failed to improve at all. Persistent remission of the arthritis/arthralgia was found in 41 patients (85%). One patient did not improve. Complete resolution of the urinary abnormalities were registered in 14 patients (40%), improvement – in 17 cases (48.5%), further deterioration in 4 patients (11.5%).

There were side effects in 13 patients (16%). In 4 (5.1%) cases there were headache and nausea, respectively. In 2 (2.5%) cases treatment was accompanied with skin itching and urticarial rash. Most severe complication presented with reversible cytotoxicity, characterised with increase of AST and ALT up to the 2–5 normal limits in 3 cases (2.5%).

Abstract FRI0483 – Table 1. Clinical response to SASP treatment in primary IgA-vasculitis (n=78)

Clinical features	Complete remission		Partial remission		No response		Worsening	
	n	%	n	%	n	%	n	%
Skin purpura (n=78)	46	58.9	27	35	4	5.1	-	-
arthritis or arthralgia (n=48)	41	85	6	13	1	1.2	-	-
hematuria and/or proteinuria (n=35)	14	40	17	48.5	-	-	4	11.5

Conclusions: To our knowledge, this is the first experience of SASP in treatment of primary IgA-vasculitis not reported before. Preliminary results look promising and worthy of further evaluation.

REFERENCE:

- [1] Ozen S, et al., EULAR/PRINTO/PRES criteria for Henoch-Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: Final classification criteria. *Ann Rheum Dis.* 2010 May;69(5):798–806.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.1809

FRI0484 OVERALL SURVIVAL AND MORTALITY RISK FACTORS IN TAKAYASU'S ARTERITIS: A MULTICENTER STUDY OF 318 PATIENTS

A. Mirose¹, C. Comarmond¹, L. Biard², M. Lambert³, A. Mékinian⁴, J.-E. Kahn⁵, E. Hachulla³, E. Messas⁶, M. Resche-Rigon², P. Cauboo, 1D. Saadoun¹ on behalf of French Takayasu network. ¹Service de médecine interne et immunologie clinique, Groupe hospitalier Pitié-Salpêtrière; ²SBIM, Hôpital Saint-Louis, Paris; ³Service de médecine interne, CHRU Lille, Lille; ⁴Service de médecine interne, Hôpital Saint-Antoine, Paris; ⁵Service de médecine interne, Hôpital Foch, Suresnes; ⁶Service de médecine vasculaire, Hôpital Européen Georges Pompidou, Paris, France

Background: Takayasu arteritis (TA) is a rare large vessel vasculitis of unknown origin. Main epidemiological data come from Asia. TA significantly increases morbidity and mortality and there is an unmet need to identify patients with higher complications and mortality risks.

Objectives: To report the long term mortality in Takayasu arteritis (TA) and to identify risk factors associated with mortality.

Methods: We analysed epidemiological data according to gender and ethnic origins, the causes of death, and the factors associated with mortality in a cohort of 318 patients [median age at diagnosis was 36^{25–47} years and 276 (86%) patients were women] fulfilling American College of Rheumatology and Ishikawa criteria of TA. Overall survival and factors associated with death and vascular complications were identified. A prognostic score for death and vascular complications was elaborated based on a multivariate model.

Results: In a cohort of 318 patients, men tended to be more frequently smokers (42% vs. 26%, p=0.07), had more frequently cardiac insufficiency (14% vs. 9%, p=0.034), but less frequently aneurysmal lesion (51% vs. 76%, p=0.005). Ethnic origins were Caucasian/White (n=87, 37%), North-Africa (n=73, 31%), Black (n=56, 18%), and other (n=17, 5%). White patients were more frequently smokers and diagnosis was more frequently made after 35 years. Black patients had less frequently a supra-aortic vessels involvement and more frequently a vessel wall thickening at diagnosis (78% vs. 62%, p=0.033).

Among 318 TA patients, 16 (5%) died after a median follow-up of 6.1 years. The mean (±SD) age at death was 38^{25–47} years with 88% of women. Main causes of death included mesenteric ischemia (n=4, 25%), aortic aneurysm rupture (n=4, 25%), sudden death (n=3, 19%), septic shock (n=3, 19%), pulmonary embolism (n=1, 6%), and cardiogenic shock (n=1, 6%). One, five, and ten years' overall survival were 98.9% [97.8–100], 98.1% [96.4–99.8], and 96.1% [93.4–98.9%], respectively. Factors associated with mortality were ethnic origin (White 9.5% vs. North Africa 7.1% vs. Black 0%, p=0.049), and tobacco smokers (10.8% vs. 4.2%, p=0.002).

We define high risk patients for death and vascular complications according to the presence of two of the following factors (i.e a progressive clinical course, thoracic aorta involvement and/or retinopathy) elaborated based on the multivariate model. The probability of death and complication free survival at five years was 78.4% (69.4–88.6) and 51.5% (38.3–69.2) (p=0.001) in the low risk and high risk group, respectively.

Conclusions: The overall mortality in our Takayasu cohort was 5% after a median follow-up of 6.1 years. Caucasian and tobacco smokers were associated with mortality in TA. We developed a simple and useful prognosis score to identify patients at risk for vascular complication and death.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4810

FRI0485 PHENOTYPE OF ANCA ASSOCIATED VASCULITIS ASSOCIATES WITH MAJOR CARDIOVASCULAR EVENTS: A RETROSPECTIVE OBSERVATIONAL STUDY FROM THE LEEDS VASCULITIS COHORT

A. Giolli^{1,2,3}, E.M.A. Hensor^{1,2}, J. Andrews², M.H. Buch^{1,2}, ¹Leeds Institute of Rheumatic and Musculoskeletal Medicine, University of Leeds; ²NIHR Leeds Biomedical Research Centre, Leeds, UK; ³Rheumatology Unit, Department of Medicine, University of Verona, Verona, Italy

Background: Patients with ANCA-associated vasculitides (AAV) are at increased risk of cardiovascular (CV) disease than the general population. The factors associated with CV involvement in AAV are poorly understood.

Objectives: To explore whether patient and disease-related characteristics associated with CV disease from a single-centre AAV cohort.

Methods: Medical records from 145 patients with AAV were reviewed and patients' diagnoses reclassified according to a validated algorithm.¹ CV events (CVE) were defined as myocardial infarction, cerebrovascular accident, thromboembolic event, heart failure, or death/hospitalisation due to CV cause. Time to first CVE was calculated; patients without CVE were censored at most recent clinical follow-up. Data on CV risk factors (hypertension, hyperlipidaemia, diabetes mellitus, chronic kidney disease stage III or higher) were collected. Putative predictors included age, sex, diagnosis (GPA, MPA, EGPA, unclassifiable AAV – UAAV), ANCA (absent, PR3, MPO), number of BVAS items, prior CV disease and number of CV risk factors. Cox proportional hazards regression was used to assess the relationship between CVE and predictors. In this exploratory analysis we used p<0.10 to indicate potential associations.

Results: 122 patients who had been followed up for between 6 months and 23 years were included in the analysis. 11/122 patients (10%) had prior history of CV disease. 17 patients (14%) experienced a CVE within median (IQR) 5.8 (1.4, 7.6) years of diagnosis. Univariable analyses indicated older age at diagnosis, MPA and prior CVD were associated with increased risk of CVE, and that those with UAAV were at decreased risk compared to GPA. With the exception of previous CVD an adjusted model confirmed the same associations (table 1).

Variable	No CVE n=105	CVE n=17	Unadjusted HR (90% CI)	Adjusted HR (90% CI)	P value
Age: mean (SD)	49 (17)	57 (13)	1.04 (1.01–1.07)	1.03 (1.00–1.06)	0.049
Female:% (n)	55% (58)	41% ⁷	0.76 (0.34–1.67)	0.69 (0.28–1.64)	0.488
Diagnosis:% (n)					
GPA	54% (57)	65% ⁷	Ref.	Ref.	0.370
MPA	13% ¹⁴	6% ¹	0.71 (0.12–2.44)	0.38 (0.05–2.21)	0.031
EGPA	10% ¹⁰	29% ⁵	3.37 (1.34–7.76)	4.81 (1.46–15.36)	0.071
UAAV	23% ²⁴	- (0)	0.15 (0.00–0.91)	0.09 (0.00–0.83)	
ANCA:% (n)					
Absent	25% ²⁶	12% ²	Ref.	Ref.	0.259
PR3	45% ⁴⁷	65% ¹¹	1.65 (0.58–6.25)	0.33 (0.08–1.79)	0.207
MPO	30% ³²	24% ⁴	1.62 (0.46–6.77)	0.27 (0.05–1.52)	
BVAS items: median (IQR)	4 (3–5)	4 (4–5)	1.26 (0.96–1.65)	1.35 (0.95–1.92)	0.156
Prior CVD:% (n)	10% ¹¹	18% ³	1.28 (1.17–9.23)	2.58 (0.72–8.03)	0.211
CV risk factors: median (IQR)	1 (0–2)	1 (0–2)	0.92 (0.59–1.39)	0.64 (0.36–1.04)	0.135

Conclusions: This exploratory analysis suggests that, among AAV, MPA may have a higher CV risk, whereas UAAV may be associated with a decreased risk. Along with ANCA status, AAV phenotype may inform future CV risk reduction interventions in AAV.