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. Mékianin⁴, 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.