(CVD) (53.92%), including hypertension (50.0%) and cardiosclerosis (34.31%), endocrine and metabolic disorders (66.67%), including hypercholesterolemia (65.70%) and obesity (32.35%), diseases of the digestive system (30.39%), dorsopathies (22.55%). The number of comorbidities was associated with age ($p=0.42$, $p<0.001$) and body mass index ($p=0.29$, $p<0.001$).

In patients with bilateral ($n=44$), compared with those affected by unilateral ANFH ($n=58$), prevalence of younger age (43.5 (36–51) vs. 54.4^{3–62} $p<0.01$) and male gender (odds ratio (OR) 2.99 (95% confidence interval (CI) 1.28–6.99), $p<0.05$) were detected. A history of (CVD) was more frequent in patients with unilateral ANFH (63.79% vs. 40.91%, OR 3.62 (95%CI 1.67–7.88), $p<0.05$), as well as hypertension (62.07% vs. 34.09%, OR 3.16 (95%CI 1.40–7.17, $p<0.01$). Patients with unilateral ANFH were more likely to have higher number of comorbidities (3.80^{1,17–6.43} vs. 2.89 (0.73–5.05)) and Charlson Comorbidity Index (0.72 (0.43–1.47) vs. 0.52 (0.43–1.87)).

Conclusions: ANFH is associated with high prevalence of comorbidities, especially CVD.

CVD, including hypertension, were more likely to be found in patients with unilateral ANFH, as well as higher comorbidity burden. It can be explained by the fact that patients with CVD received treatment according to national guidelines, including antihypertensive drugs, anticoagulants, statins, etc. This indicates that performing secondary prevention of CVD can be important in both CVD and ANFH, as such treatment can influence on intraosseous blood circulation in the contralateral joint.

REFERENCE:

- [1] Valderas JM, Starfield B, Sibbald B, Salisbury C, Roland M. Defining comorbidity: implications for understanding health and health services. *Ann Fam Med* 2009;7:357–363.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7405

SAT0618

RELATIONSHIP BETWEEN ARTERIAL STIFFNESS AND DISEASE DURATION IN BEHCET'S DISEASE

K. Ayar¹, B. Sensoy², M.E. Aslancı³, T. Teker⁴, S. Cekiç⁴. ¹Rheumatology, University of Health Sciences, Bursa Yüksek İhtisas Training and Research Hospital; ²Cardiology; ³Ophthalmology; ⁴Internal Medicine, University of Health Sciences, Bursa Yüksek İhtisas Training and Research Hospital, BURSA, Turkey

Background: There are few studies in the literature investigating arterial stiffness in Behcet's Disease (BD). The association of arterial stiffness with duration of disease is unknown.

Objectives: The aim of this study is to compare arterial stiffness by assessing pulse wave velocity (PWV) and augmentation index (AI) measurements in healthy controls (HC) and patients diagnosed as BD with low and high disease duration.

Methods: Seventy BD patients and 40 HC without diabetes mellitus, Hypertension or chronic renal failure were included in the study. Patients with BD who were diagnosed within 12 months were taken to the group with low disease duration (LDD) and the others to the group with high disease duration (HDD). Demographic data and lipid parameters of all participants were recorded and PWV and AI measurements were made from brachial artery. The clinical characteristics of the patients with BD were recorded and the Behcet's Syndrome Activity Scale (BSAS) questionnaire was filled in for all patients with BD. Findings were compared between LDD, HDD and HC groups.

Results: Age, gender and body mass indexes were not statistically different between groups ($p=0.134$, $p=0.948$, $p=0.929$ respectively). Augmentation index and PWV measurements were different between groups ($p=0.014$, $p=0.024$ respectively). In the HDD group, both PWV and AI were significantly higher than HC ($p=0.013$, $p=0.006$ respectively). In LDD group AI was significantly higher than HC however PWV was not ($p=0.047$, $p=0.919$ respectively). There was moderate correlation between PWV and disease duration (Rho:0.393; $p<0.001$) and there was no correlation between AI and disease duration ($p=0.287$)

Abstract SAT0618 – Table 1. Demographic data, parameters of arterial stiffness, lipid parameters in all participants and disease duration, BSAS scores in patients with BD.

	Healthy controls (n=40)	The time since the diagnosis of BD		P
		LDD ¹ (12 months <=) (n=32)	HDD ² (12 months <) (n=38)	
Age (year)	38.35±8.99	37.16±8.89	41.37±9.38	0.134
Gender (f/m)	23/17	20/12	19/19	0.948
BMI ³ (kg/m ²)	26.55±3.65	26.33±3.68	26.22±4.38	0.929
BSAS ⁴ (0-100)		42.97±21.59	42.87±22.83	0.985
Disease duration (month)		0 (0-6.7)	120 (36-186)	<0.001
Augmentation index (%)	22.53±7.80	26.63±9.72	28.87±11.24	0.014
Pulse wave velocity (m/s)	5.75 (5.3-6.4)	5.8 (5.3-6.5)	6.6 (5.6-7.0)	0.024
LDL (mg/dl)	124.25±29.99	114.89±29.41	115.80±27.78	0.593
T. Cholesterol (mg/dl)	204.25±36.80	191.82±42.66	188.80±29.62	0.426
Triglycerides (mg/dl)	105.62±35.46	127.82±77.22	125.40±58.38	0.507
HDL (mg/dl)	54.75±10.34	52.35±13.62	47.65±10.71	0.182

LDD: Low disease duration, HDD: High disease duration, BMI: Body mass index, BSAS: Behcet Syndrome Activity Scale, BD: Behcet Disease

Conclusions: The augmentation index is higher in patients with BD than HC regardless of the duration of the disease. Pulse wave velocity does not increase in the early period of the disease in patients with BD but increases with prolonged disease duration.

REFERENCES:

- [1] Yilmaz S1, Celik G, Esmen SE. Assessment of arterial stiffness in patients with inactive and active Behcet's disease. *Scand J Rheumatol*. 2014;43 (1):63–9. doi:10.3109/03009742.2013.809787. Epub 2013 Sep 9
- [2] Yıldırım A1, Karakaş MS2, Kılınc AY3, Altın RE3, Yalçınkaya AS3. Evaluation of arterial stiffness and subclinical atherosclerosis in patients with Behcet's disease without cardiovascular involvement. *Türk Kardiyol Dern Ars* 2016;Oct;44(7):575–581. doi:10.5543/tkda.2016.06944

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.2823

SAT0619

ASSOCIATIONS BETWEEN CLINICAL MANIFESTATIONS OF BEHCET'S SYNDROME AND WORK OUTCOMES: RESULTS FROM A UK CROSS-SECTIONAL ANALYSIS

L. Chadwick^{1,2}, N.J. Goodson^{1,2}, R.J. Moots^{1,2}. ¹Department of Rheumatology, Aintree University Hospital; ²Institute of Ageing and Chronic Disease, University of Liverpool, Liverpool, UK

Background: Behcet's syndrome (BS) is a multi-systemic auto-inflammatory disease which exhibits significant heterogeneity in clinical manifestations, including orogenital ulceration, skin rash, arthralgia and ocular, venous, gastro-intestinal and neurological involvement. BS affects adults of working age, usually presenting between 20–40 years,¹ and can therefore have a significant impact on work. The association between individual and cumulative clinical manifestations of BS on employment status has not previously been assessed.

Objectives: To 1) describe demographics, clinical manifestations, work outcomes and use of unemployment benefits in a UK BS cohort and 2) to explore the relationship between individual and cumulative manifestations of BS and work outcomes.

Methods: A cross sectional analysis was performed using the Liverpool Behcet's Centre of Excellence clinical database. Inclusion criteria were clinical characteristics meeting International Criteria for Behcet's Disease (ICBD) diagnostic criteria and recorded employment data. Patients meeting ICBD but thought unlikely to have BS on clinical review by our multi-disciplinary team were excluded. Demographics, clinical manifestations, work outcomes and use of employment related benefits were described. A binomial variable 'Out of work' was generated and defined as unemployment, sickness absence, or patients marked as retired but of working age, a carer or a homemaker if the patient had to stop work due to BS. Associations between Out of work and: 1) individual clinical manifestations of BS, and 2) number of clinical BS manifestations, were explored using multivariate logistic regression adjusted for age, gender, mean EQ5D score and socio-economic status. Odds ratios (OR) and 95% confidence intervals were calculated.

Results: 120 patients met inclusion criteria. Mean age was 41.1 years and 33 patients (28%) were male. The minimum number of clinical manifestations were two in order to meet ICBD; the frequencies were 100% oral ulceration, 94% genital ulceration, 71% arthralgia, 43% skin rash and 20% ocular, 7% neurological, 6%

vascular and 5% gastro-intestinal involvement. 37 patients (31%) were out of work with 44 patients (37%) claiming employment related benefits. With regard to individual clinical manifestations, ocular disease had a statistically significant increased risk for being out of work when assessed alone with OR 2.84 (95% CI 1.13, 7.13) but lost statistical significance when analysed in the multivariate model: OR 2.45 (95% CI 0.70, 8.60). With regard to cumulative clinical manifestations, patients with four or more clinical manifestations of BS had a statistically significant increased risk of being out of work with OR 5.57 (95% CI 1.33, 23.27) in comparison to patients with two manifestations in the multivariate model.

Conclusions: This study has highlighted the significant burden of BS on work outcomes in this UK cohort. In particular, four or more cumulative BS manifestations were independently associated with being out of work in this young cohort. Further work is required to identify whether education or intervention in the workplace can help prevent disease related job loss in BS.

REFERENCE:

- [1] Ambrose NL, Haskard DO. Differential diagnosis and management of Behçet syndrome. *Nat Rev Rheumatol* 2013;9:79–89. doi:10.1038/nrrheum.2012.156

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4791

SAT0620

FURTHER EVIDENCES OF SECONDARY AMYLOIDOSIS IN ALKAPTONURIA

B. Frediani¹, L. Millucci², G. Bernardini², D. Braconi², M. Bardelli¹, B. Marzocchi², O. Spiga², M. Orlandini², S. Sestini³, M. Galeazzi¹, A. Santucci². ¹Departement of Medical Sciences, Surgery and Neurosciences, A.O.U Siena, Departement of Medical Sciences, Surgery and Neurosciences, A.O.U Siena; ²Department of Biotechnology, chemistry and pharmacy, Siena, Department of Biotechnology, chemistry and pharmacy, Siena; ³Italian Association Alkaptonuria Patients, AIMAKU, Italian Association Alkaptonuria Patients, AIMAKU, Siena, Italy

Background: Alkaptonuria (AKU) is an ultra-rare inborn error of metabolism due to a deficient activity of homogentisate 1,2-dioxygenase. Patients suffer from a severe arthropathy. Evidence was provided on the presence of a secondary serum amyloid A (SAA)-based amyloidosis[¹Here a complete microscopic and ultrastructural analysis of different AKU tissues, taken from six differently aged patients, is presented.

Objectives: SAA amyloidosis is a complication in AKU[² making the detection of amyloid deposits at an early phase, important for treatment[³ We present a study of tissues from patients of different age and relevance of symptoms, providing a detailed overview of AKU amyloidosis.

Methods: Different tissues were obtained from a cohort of 6 AKU patients: 4 M (63,68,42,44 y) and 2 F (66,71 y) with different severity of symptoms. Histology and amyloid were investigated. A complete microscopic and ultrastructural analysis is presented and patient features as radiological examination, mild-to-severe degenerative changes as joint space narrowing, cartilage irregularities, sub-chondral sclerosis or peripheral osteophytes and linear intervertebral disk calcifications were reported. SAAserum levels and other serological markers were measured too. Specimens were analysed by Congo Red, Immunofluorescence, Transmission Electron Microscopy.

Results: The analysis of all AKU specimens confirmed the massive presence of amyloid fibrils even in young patients. Alterations in collagen composition, strictly associated to amyloid bundles deposition, were observed especially in labial salivary gland, cartilage, tendons and infrapatellar fat pad. Histological analysis showed depletion of glycosaminoglycans in young patients, whereas, at light microscopy, calcification and fibrillation were observed only in elderly patients. Immunofluorescence assessed undoubtedly the presence of SAA in amyloid deposits in AKU, and we reported for the first time the finding of amyloid deposition in young AKU patients and in less common regions.

Conclusions: We provide the first detailed overview of amyloidosis in AKU. Overall, our findings depicted a novel biological framework underlining the pathological role of amyloidosis in several AKU tissues. Furthermore, we found that degradation of extra-cellular matrixin AKU is not limited to elderly. The clinical burden of AKU may notably increase, since amyloidosis was found even in young AKU patients, whereas degeneration of cartilage and tendons was limited to older subjects.

REFERENCES:

- [1] Millucci L, et al. *J Inher Metab Dis* 2015 Sep;38(5):797–805.
[2] Braconi D, et al. *Free Radic Biol Med* 2015 Nov;88(Pt A):70–80.
[3] Millucci L, et al. *Diagn Pathol*. 2014 Sep 26;9:185.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4645

SAT0621

CLINICAL SIGNIFICANCE OF INTERLEUKIN-18 AND INTERLEUKIN -6 ON DISEASE COURSE OF ADULT-ONSET STILL'S DISEASE

M.G. Ahmed. *Rheumatology, Alazhar University, Cairo, Egypt*

Background: Innate pro inflammatory cytokines interleukin (IL)–6 and IL-18 are critical for perpetuating the inflammatory processes in adult-onset Still's disease (AOSD),¹ and macrophage activation syndrome (MAS).²

Objectives: To assess the role of IL-6 and IL-18 in the pathogenesis of (AOSD) and MAS, and to investigate the clinical significance of serum cytokine profile with IL-6 and IL-18 in (AOSD) and MAS,

Methods: we analysed the serum IL-6 and IL-18 in patients with (AOSD) and compared them with the clinical features of (AOSD) 51 patients with (AOSD) including 17 patients with MAS were analysed. Levels of IL-6 and IL-18 were quantified in serum by enzyme linked immune sorbent assay. Results were compared with clinical features of (AOSD).

Results: Two distinct Adult Still's disease patient subsets based on their serum IL-6 and IL-18 levels were identified: an IL-18 dominant (IL-18/IL-6 1000) and an IL-6 dominant (IL-18/IL-6 1000). The IL-6 dominant subset had a significantly greater number of joints with active disease and higher serum levels of matrix metalloproteinase-3, whereas the IL-18-dominant subset was more likely to develop MAS. The cut off value of serum IL-18 to predict the development of MAS was 52000 pg/ml with 87.3% of sensitivity and 79.3% of specificity. The patients with IL-18 dominant pattern were likely to have monophasic or polycyclic disease course, whereas the patients with IL-6 dominant pattern were likely to have persistent disease course. Serum IL-18 levels in patients achieved remission decreased to the levels <1,000 pg/mL in inactive phase and normalised in remission phase. In contrast, serum IL-18 levels in patients experienced relapse during withdrawal of steroid within 12 months after disease onset demonstrated a sustained elevation of serum IL-18 levels >1,000 pg/mL during the inactive phase

Conclusions: Two subsets of patients with (AOSD), one which is prone for arthritis and another with prone for MAS, can be identified on the basis of their serum IL-6 and IL-18 levels. These two subsets appear to be characterised by certain distinct clinical features. Monitoring the cytokine profile with IL-18 and IL-6 might be useful to predict disease course. Furthermore, serum IL-18 levels reflect the biological activities of the immune system in (AOSD) and may predict the development of MAS and the prognosis of Adult Still's disease

REFERENCES:

- [1] Choi JH, Suh CH, Lee YM, Suh YJ, Lee SK, Kim SS, et al. Serum cytokine profiles in patients with adult onset Still's disease. *J. Rheumatol* 2003;30:2422–2427.
[2] Schuler GS, Grom AA. Macrophage activation syndrome and cytokine-directed therapies. *Best Practice & Research: Clinical Rheumatology* 2014;28:277–292.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6951

SAT0622

HIGH DOSE INTRAVENOUS METHYLPREDNISOLONE INDUCES RAPID IMPROVEMENT IN SEVERE UVEITIS: A MULTICENTER STUDY OF 129 PATIENTS

N. Vegas-Revenga¹, V. Calvo-Rio¹, I. González-Mazón¹, L. Sánchez-Bilbao¹, E. Beltrán², A. Follónosa³, O. Maiz⁴, A. Blanco⁴, M. Cordero⁵, N. Ortego⁶, I. Torre⁷, F. Hernández⁸, S. Muñoz⁹, M. Esteban⁹, M. Díaz-Llopis¹⁰, J. Cañal¹, J. A. Ventosa¹, R. Demetrio¹, M. Agudo¹, L. Domínguez-Casas¹, M.A. González-Gay¹, R. Blanco¹. ¹HUMV, Santander; ²H. Peset, Valencia; ³H. Cruces, Barakaldo; ⁴H. Donostia, Donostia; ⁵H. de León, León; ⁶H. San Cecilio, Granada; ⁷H. Basurto, Bilbao; ⁸H. Negrín, Gran Canaria; ⁹H. Infanta Sofía, Madrid; ¹⁰H. La Fe, Valencia, Spain

Background: In uveitis rapid and effective remission-inducing therapy is mandatory to avoid irreversible damage.

Objectives: To evaluate the efficacy and safety of high-dose intravenous methylprednisolone (IVMP) pulse therapy in uveitis.

Methods: Multicentre study of 129 patients (203 eyes) with uveitis who received IVMP. The underlying diseases were: Idiopathic (n=30), Vogt Koyanagi Harada,²⁹ Behçet disease,¹⁹ Sarcoidosis,⁶ Multifocal Choroidopathy,⁴ Birdshot chorioretinopathy,² Acute posterior multifocal placoid pigment epitheliopathy,¹ Granulomatosis with polyangiitis,² Aortitis,¹ Immune thrombocytopenic purpura,¹ Rheumatoid arthritis,² Axial Spondylitis,⁶ Psoriatic arthritis,² Juvenile Idiopathic Arthritis,¹ Thrombophilia synd.,¹ Eales Disease,² Sympathetic Ophthalmia,² Multiple Sclerosis,² Relapsing Polychondritis,¹ Cogan's synd.,¹ Sjögren synd.,² Crohn's disease,¹ Reactive arthritis,¹ Toxic oil synd. and brain ischemia,¹ Raynaud's Disease and brain ischemia,¹ Pseudotumor,¹ Cataract surgery,¹ Herpes Simplex,¹ Varicella Zoster-associated acute retinal necrosis,¹ *T. pallidum*³ and *M.*