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 matrix 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.*

tuberculosis.¹ The inflammatory ocular patterns were: panuveitis (66), posterior uveitis (PU),³¹ intermediate uveitis (IU),⁵ anterior uveitis (AU),¹² AU and PU,² AU and IU,¹ IU and PU,¹ exudative retinal detachment,³ vasculitis,⁴ pseudotumor,¹ scleritis² and sclero-uveitis.¹ Bilateral ocular involvement was observed in 74 cases. Patients were assessed at basal visit and day 2–5, 7, 15 and 30 after IVMP. The main outcome variable was best corrected visual acuity (BCVA), the degree of inflammation of the anterior chamber and vitreous, and macular thickness (macular oedema defined by OCT >300 µm). The results are expressed as mean ±SD for normally distributed variables, or as median [interquartile range] when are not. Comparison of continuous variables was performed using the Wilcoxon test.

Results: We studied 74 ♀/55 ♂; mean age 42±14.3 years. IVMP dose ranged from 40–1000 mg/day for 3–5 consecutive days. Prior to IVMP, cycloplegic and corticosteroid eye drops were used in all cases. Improvement was faster among patients with inflammation in anterior chamber and vitritis than in BCVA, CME, retinitis and retinal vasculitis (**TABLE**). Total remission was achieved in 19.4% of the 129 patients after IVMP. In 126 of the patients continued with oral corticosteroids and 4 received an intraocular dose of steroids. These immunosuppressive drugs were added: Methotrexate (52), Cyclosporine A,⁴⁸ Azathioprine⁴⁰ and others.⁵ In a few cases biological therapy (64) was administered afterwards. In general, acute respiratory infection was the side-effect most frequent.

Abstract SAT0622 – Table 1

	Affected eyes	Affected eyes after high-dose IVMP (%)			
	baseline	Day 2-3	Day 7	Day 15	Day 30
BCVA, mean±SD	0.47±0.35	0.56±0.34*	0.61±0.33*	0.68±0.31*	0.75±0.3*
Anterior chamber cells [median (IQR)]	0.75 [0-4]	0 [0-4]*	0 [0-4]*	0 [0-3]*	0 [0-2]*
Vitritis [median (IQR)]	0 [0-5]	0 [0-5]*	0 [0-5]*	0 [0-3]*	0 [0-3]*
OCT (microns) mean±SD	405.36±189.1	365.49±159.7*	346.48±150.9*	308.2±120.06*	281.79±107.6*
Choroiditis/ Chorioretinitis n, (%)	59 (23%)	51 (20%)	33 (13%)	23 (9%)	15 (5%)
Retinitis n, (%)	86 (34%)	74 (29%)	70 (27%)	46 (18%)	21 (8%)
Retinal vasculitis n, (%)	66 (26%)	58 (23%)	52 (20%)	36 (14%)	16 (6%)
Synechiae n, (%)	39 (15%)	32 (12%)	30 (12%)	30 (12%)	33 (13%)
Cystoid macular edema (CME) n, (%)	83 (33%)	68 (27%)	57 (22%)	41 (16%)	20 (8%)

*p <0.05 compared with basal

Conclusions: High-dose IVMP pulse therapy is beneficial in the prompt control of severe uveitis and it is well tolerated.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3780

SAT0623

CLINICAL FEATURES AND TREATMENT OF IDIOPATHIC RECURRENT ACUTE PLEURO-PERICARDITIS

K. Dietz, H.-M. Lorenz, N. Blank. *Rheumatology, Internal Medicine 5, University Hospital Heidelberg, Heidelberg, Germany*

Background: Idiopathic recurrent acute pleuro-pericarditis (IRAP) is an increasingly recognised autoinflammatory disease comprising post-pericardiotomy-syndrome, recurrent pericarditis and post-myocardial-infarction-syndrome. Different autoimmune mechanisms were discussed in the past. Recently, IRAP is considered as an autoinflammatory disease. Therapeutic options comprise colchicine, prednisolone and interleukin (IL)–1beta blockingagents.

Objectives: to investigate whether idiopathic and post-interventional pleuro-pericarditis represent a clinical spectrum and to identify treatment options.

Methods: This study analyses demographic, clinical and laboratory features of post-interventional and idiopathic pleuro-pericarditis and adult onset Still's disease (AOSD) as a reference. Patients with infectious disease, connective tissue disease, chronic heart failure, renal failure and other non-exsudative effusions were excluded from this analysis. Patients with IRAP were treated with colchicine, prednisolone and IL1β blocking agents.

Results: Between 2005 and 2017 66 cases of idiopathic and post-interventional pleuro-pericarditis were identified and compared to 83 cases of AOSD. Clinical and laboratory features suggest that idiopathic and post-interventional pleuro-pericarditis represent a clinical spectrum which is identical with IRAP.

Prednisolone was started with 25 mg to 125 mg and tapered to less than 7 mg or discontinued if not effective. 47 of 66 patients (71%) were treated with prednisolone and 10/47 (21%) were in remission with no need of any further therapeutically escalation.

Colchicine was given to 44/66 patients (67%) and 29/44 (66%) were in complete remission.

Four of 66 patients (6%) did not respond or had contraindications against colchicine or prednisolone and were treated with anakinra. Of these patients 4/4 (100%)

were in remission. During the follow-up period of 20 patient*months 2 of 4 patients maintained the remission with anakinra every 2nd day and two patients discontinued anakinra and remained in remission.

Conclusions: 1. post-pericardectomy-syndrome, post-myocardial-infarction-syndrome and idiopathic recurrent pericarditis represent a clinical spectrum of autoinflammatory diseases.

2. treatment options comprise colchicine as a first-line therapy, prednisolone and anti-IL1 blocking agents.

Acknowledgements: none.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3027

SAT0624

INCIDENCE AND BASELINE CHARACTERISTICS OF RELAPSE OR EXACERBATION IN PATIENTS WITH PULMONARY SARCOIDOSIS: A SINGLE CENTRE LONG-TERM OBSERVATIONAL COHORT STUDY

O. Murata^{1,2}, A. Kudo², K. Suzuki³. ¹Division of Pulmonary Medicine, Allergy, and Rheumatology, Department of Internal Medicine, School of Medicine, Iwate Medical University, Morioka; ²Department of Pulmonary Medicine, Hachinohe Red Cross Hospital, Hachinohe; ³Division of Rheumatology, Department of Internal Medicine, School of Medicine, Keio University, Tokyo, Japan

Background: Sarcoidosis is a systemic granulomatous disease that can affect multiple organs. Lung involvement is particularly common, occurring in more than 90% of patients.¹ However, spontaneous remission of pulmonary sarcoidosis is often observed within 2 years.² Factors reported to indicate a poor prognosis in pulmonary sarcoidosis include chest radiographic stage, skin involvement, age over 40 years and smoking history.^{3, 4} To date, however, no study has comprehensively described factors of relapse and exacerbation and characteristics of untreated patients with favourable outcomes over a long-term period of more than 2 years.

Objectives: The aim of study was to identify the incidence and baseline characteristics of relapse and exacerbation in patients with pulmonary sarcoidosis over a long-term period.

Methods: One hundred- and three patients who visited our division of Hachinohe Red Cross Hospital between January 2007 and December 2016 and were clinically diagnosed as having pulmonary sarcoidosis were enrolled. The incidence and characteristics of relapse or exacerbation were prospectively calculated and statistically analysed using clinical, laboratory and imaging data collected from medical records.

Results: Of 103 patients, 79% were women and mean age at diagnosis was 50.1 ±16.4 years. Mean observation period was 9.8±8.6 years. The number of patients with previously reported short-term prognostic factors were as follows: chest radiographic stage (Stage 0, n=7; Stage I, n=32; Stage II, n=36; Stage III, n=18; Stage IV, n=10), skin involvement (n=9), age over 40 years old (n=73), and smoking history (n=38). Overall relapse or exacerbation was 22.3% (n=23) and mean period to relapse or exacerbation was 8.7±8.3 years. We then analysed the 69 patients who were observed for more than 5 years, and identified relapse or exacerbation within 5 years in 9 patients. Comparison of characteristics at diagnosis between the relapse or exacerbation group and spontaneous group showed that the relapse or exacerbation group had a significantly elevated frequency of bilateral hilar lymphadenopathy, disease duration, ocular involvement, cardiac involvement, and oral glucocorticoid use at diagnosis (p=0.014, 0.027, 0.019, 0.035, and 0.0043, respectively). On stratification, the number of risk factors was positively and significantly associated with the cumulative ratio of relapse or exacerbation (p=0.048).

Conclusions: Our long-term observational cohort study newly identified the incidence and baseline characteristics of relapse or exacerbation in patients with pulmonary sarcoidosis over a long-term period. Scoring the number of the factors at baseline may facilitate the prediction of relapse or exacerbation.

REFERENCES:

- [1] Br. J. Dis. Chest 1976;70:206–210.
- [2] Dan. Bull. Med 1982;29:27–32.
- [3] Br. Med. J 1961;2:1165–1172.
- [4] Chest 2006;130:1851–6.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.2745