till EOS. The number of pts with absent autoinflammatory disease improved from 23.5% (4 pts) to 64.7% (11 pts). This improvement was also observed in the assessment of skin rash; proportion of patients with absent skin disease increased from 29.4% (5 pts) to 94.1% (16 pts). The mean decrease in CRP and SAA levels from baseline were -10.4mg/L and -54.36/L respectively. Overall, 10 (58.8%) pts had AEs suspected to be related to study drug; the most common were diarrhoea, pneumonia, rhinitis and cough (3 pts each). Eight (47.1%) pts experienced at least 1 Serious AE (4 MWS and 4 NOMID pts); with pneumonia being the most common 2 (11.8%). No deaths were reported during the study.

Conclusions: CAN effectively maintained clinical and serological efficacy in CAPS pts. No new safety findings were observed and the safety profile of CAN was consistent with previous studies, which corroborates the long-term use of CAN in the treatment of CAPS patients.

References:

[1] Kuemmerle-Deschner JB, et al. Arthritis Res Ther. 2011;13(1):R34.
[2] ILARIS (Package Insert). Novartis Pharmaceuticals, Inc. 2014

Disclosure of Interest: P. Brogan Grant/research support from: Novartis Roche and SOBI, Consultant for: Novartis Roche and SOBI, M. Hofer Consultant for: Novartis, J. Kuemmerle-Deschner Grant/research support from: Novartis, Consultant for: Novartis, SOBI, Baxalta, B. Lauwerys: None declared, A. Speziale Employee of: Novartis, K. Leon Employee of: Novartis, X. Wei Employee of: Novartis, R. Laxer Grant/research support from: Database funding from Novartis

DOI: 10.1136/annrheumdis-2016-eular.4865

FRI0504 PROGNOSTIC FACTORS OF ADULT ONSET STILL'S DISEASE: ANALYSIS OF 100 CASES IN 3 TERTIARY REFERRAL CENTERS

P. Ruscitti[1], P. Cipriani[1], F. Ciccia[2], F. Masedu[3], D. Iacono[4], V. Liakouli[1], G. Guggino[2], F. Carubbi[1], O. Berardicurti[1], P. Di Benedetto[1], M. Valenti[3], G. Triolo[2], G. Valentini[4], R. Giacomelli[1]. *[1]Division of Rheumatology, University of L'Aquila, L'Aquila; [2]Division of Rheumatology, University of Palermo, Palermo; [3]Division of Medical Statistic Unit, University of L'Aquila, L'Aquila; [4]Division of Rheumatology, Second University of Naples, Naples, Italy*

Background: Adult onset Still's disease (AOSD) is rare inflammatory disease [1]. Three different patterns of AOSD have been identified: i. monocyclic pattern, characterized by a systemic single episode; ii. polycyclic pattern, associated with multiple flares, separated by remissions; iii. chronic pattern, related to a persistently active disease with associated polyarthritis [2]. Until now, the treatment of AOSD remains largely empirical, lacking controlled clinical trials [1].

Objectives: We aimed to investigate clinical data of AOSD patients and any possible correlation among these features and the outcome of patients.

Methods: Clinical data of 100 AOSD patients were recorded. All patients fulfilled the Yamaguchi AOSD diagnostic [3]. Gender, age, clinical features, disease activity Pouchot's score [4], complications, serum ferritin, erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), treatments, comorbidities, patterns and AOSD-associated death, were reported. Furthermore, we evaluated any possible correlation among these clinical features and the outcome of patients, by using linear regression analysis.

Results: One hundred consecutive AOSD patients (66 men, 34 women), whose age at diagnosis was 45.35±16.23 (mean±SD), were enrolled. All patients experienced fever, 86 patients showed joints involvement, 79 patients showed splenomegaly, skin rash was present in 78 patients, 62 patients showed hepatic involvement, 57 patients showed enlargement of lymph nodes. The Pouchot's score, 6.11±2.02 (mean±SD), confirmed the activity of the disease. Sixteen patients experienced different complications, mainly the macrophage activation syndrome. A strong increase of inflammatory markers (mean±SD) was observed: serum ferritin was 2560.07±3726.64 ng/mL, ESR was 67.28±26.65 mm/hr, CRP was 78.35±72.76. All the patients received steroids at different dosages, and 55 patients were treated with steroids and immunosuppressive drugs, mainly methotrexate and ciclosporin. Thirty-two patients were treated with biologic agents: 5 with anakinra, 8 with tocilizumab, and 19 with TNF inhibitors. Twenty-nine patients showed the monocyclic pattern, 22 patients showed the polycyclic pattern, 33 patients showed the chronic pattern. AOSD-associated death occurred in 16 patients. Regression analysis showed that Pouchot's score ($p=0.011$), the presence of any complication ($p<0.0001$) and associated comorbidities ($p<0.0001$) correlated with the unfavorable outcome.

Conclusions: This study provides information on the clinical, laboratory, therapeutic, and prognostic features in a large cohort of AOSD patients. The Pouchot's score, the presence of complications and comorbidities were significantly associated with the AOSD-associated death.

References:

[1] Gerfaud-Valentin M, et al. Adult-onset Still's disease, Autoimmun Rev 2014;13,708–722.
[2] Cush JJ, et al. Adult-onset Still's disease. Clinical course and outcome. Arthritis Rheum 1987;30:186–9425.
[3] Yamaguchi M, et al. Preliminary criteria for classification of adult Still's disease. J Rheumatol 1992;19:424–30.
[4] Pouchot J, et al. Adult Still's disease: manifestations, disease course, and outcome in 62 patients. Medicine (Baltimore) 1991;70:118–36.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2016-eular.3955

FRI0505 UNDIFFERENTIATED CONNECTIVE TISSUE DISEASE: A 121 PATIENTS AUDIT FOCUSING ON INITIAL DIAGNOSIS AND CHANGES OVER TIME

R. Ionescu[1,2], M. Leandro[1,3]. *[1]Rheumatology, University College London Hospitals, London, United Kingdom; [2]Rheumatology and Internal Medicine, Sfanta Maria Hospital, Bucharest, Romania; [3]Rheumatology, University College London, London, United Kingdom*

Background: The diagnosis of Undifferentiated Connective Tissue Disease (UCTD) has raised controversy over the years regarding making the diagnosis, evolution and prognosis. Le Roy et al[1] in 1980 and most recently Mosca et al[2] in 2014 proposed classification criteria for UCTD, but confusion still exists regarding making the diagnosis of UCTD, evolution and prognosis of the syndrome and how the patient should be followed up over time.

Objectives: Evaluate disease evolution, treatment and follow up plan in patients with UCTD diagnosis. Assess whether there are any factors that may predict disease evolution and should require regular follow up.

Methods: This was a retrospective study of patients followed up in a specialist clinic in a tertiary referral centre (University College London Hospitals) with a diagnosis of UCTD, focusing on clinical and serological features, treatment, follow up and disease evolution over time.

Results: A total of 121 patients were included in the study: 93% were females; the mean age at disease onset was 39 years (range 20 to 80); and the patients were followed up for at least 1 year (mean 12 years, range 1 to 40). 78% of these patients had a stable diagnosis of UCTD, while in 22% diagnosis changed over time: 9% evolved into a specific connective tissue disease (CTD) - 4 Systemic Lupus Erythematosus, 3 Sjogren's Syndrome, 1 Rheumatoid Arthritis, 1 Systemic sclerosis and 1 Anti-synthetase Syndrome -, 7% had a change of diagnosis from a specific CTD to UCTD, 4% evolved into an overlap syndrome and 2% of the patients were no longer diagnosed as having any CTD at the end of the study. The most prevalent manifestations were joint pain (arthralgia/arthritis) in 89% of the patients, fatigue 80%, Raynaud's phenomenon 63%, skin rashes 49% and sicca symptoms 45%. Lung involvement was observed in 11% of the patients within which 8% had a nonspecific interstitial pneumonia (NSIP) and 3% had a usual interstitial pneumonia (UIP) pattern. 36% of the patients were noted to have associated gastro-oesophageal reflux disease. Serological features included positive ANA in 98%, anti-RNP 33%, anti-Ro 32%, Rheumatoid Factor 20% and hypocomplementemia was observed in 19% of the patients. 7% of the patients did not require any treatment for their UCTD, but the majority of patients were treated with Hydroxychloroquine only (75%), and the rest with other immunosupressants/immunomodulators.

Conclusions: In our study the majority of patients initially diagnosed with UCTD kept this diagnosis over time but 13% of patients evolved to a defined CTD or an overlap syndrome and in 2% of patients, symptoms and serological features eventually resolved. Although, UCTD is often mild, significant major organ involvement such as interstitial lung disease can occur[3], as well as evolution to a defined CTD or overlap syndrome. This should guide follow up of these patients in clinic.

References:

[1] LeRoy EC et al. Undifferentiated connective tissue syndromes. Arthritis Rheum.1980; 23 (3):341 - 343.
[2] M. Mosca et al The diagnosis and classification of undifferentiated connective tissue diseases Journal of Autoimmunity 48–49 (2014) 50e52.
[3] Fischer A, Interstitial lung disease in undifferentiated forms of connective tissue disease. Arthritis Care Res (Hoboken). 015;67(1):4–11.

Disclosure of Interest: R. Ionescu Grant/research support from: EULAR scientific training bursary, M. Leandro: None declared

DOI: 10.1136/annrheumdis-2016-eular.5472

FRI0506 PREGNANCY IN PATIENTS WITH UNDIFFERENTIATED CONNECTIVE TISSUE DISEASE: CLINICAL OUTCOMES IN 78 CASES

R. Vagelli[1], C. Tani[1], L. Carli[1,2], D. Totti[1], M. Tristano[1], F. Strigini[1], M. Mosca[1]. *[1]Clinical and Experimental Medicine, University of Pisa, Pisa; [2]GenOMeC PhD, University of Siena, Siena, Italy*

Background: Pregnancy is considered to be an important factor that may affect the course of autoimmune diseases; few data are currently available in the literature on pregnant patients with diagnosis of undifferentiated connective tissue disease (UCTD).

Objectives: To assess the outcome of pregnancy and evaluate the percentage of disease flare in a cohort of patients with UCTD.

Methods: We conducted a prospective study on 78 pregnancies in 64 UCTD patients, regularly monitored from the positive pregnancy test until the postpartum period by a multidisciplinary team including rheumatologists and obstetricians at our unit from 2000 to 2015.

Results: The mean age of patients at pregnancy was 34.1 (range 21–43) and the mean disease duration was 7.26 years. Only 3 patients presented hematologic disease activity at the beginning of pregnancy and none of the patients had experienced previous renal involvement. 20.8% of UCTD patients presented antiphospholipid (aPL)-antibodies positivity (only 2 cases of triple positivity of LAC, anti-β2GPI and ACLA) and the same percentage had anti-Ro positivity.