characteristics of autoimmune diseases associated with sarcoidosis (sarcoidosis-overlap group) patients and isolated sarcoidosis (isolated sarcoidosis group) were analyzed and compared.

Results: Autoimmune disease was detected in 15 (11.5%) of 131 patients with sarcoidosis (1Sjögren syndrome, 3rheumatoid arthritis, 1Still disease, 1scleroderma, 4ankylosing spondylitis, 1familial Mediterranean fever, 1gut arthritis, 1immune trombocytopenic purpura, 1Hashimoto thyroiditis and 1Graves disease). Most of these diseases occurred before (such as RA, AS, Still, FMF) and others after sarcoidosis diagnosis. Among 15 sarcoidosis patients with autoimmune disease 10 were female and 5 were male, the mean age was 50.8 years and mean disease duration was 3 months (1–30 months). When compared with isolated sarcoidosis patients, more hand finger joint involvement, RF positivity, higher ESR and less NSAIDs usage were found in patients with sarcoidosis-overlap group (p=0.035, p=0.049, p=0.015, p=0.018 respectively). There was no statistically significant differences between the two groups when evaluated for demographic, clinical parameters and other treatment modalities.

Conclusions: Concomitant autoimmune diseases in patients with sarcoidosis may be often seen. This patients are characterized with more hand finger joint involvement, RF positivity, higher ESR and less NSAIDs usage. Therefore, in patients with a diagnosis of sarcoidosis, it is necessary for the physician to be careful and to make a wider differential diagnosis in terms of the presence of another underlying autoimmune disease. Multicenter, prospective studies involving large numbers of patients are needed to understand whether the association of sarcoidosis-autoimmune diseases is based only on coincidence or on a common etiopathogenesis.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.1376

THU0574 CLASSICAL IMMUNOSUPPRESSION AND DAMAGE PROGRESSION IN A GROUP OF PATIENTS DIAGNOSED WITH BEHCET'S DISEASE

S. Daia-Iliescu, C. Buzatu, A. Borangiu, I. Saulescu, L. Groseanu, V. Bojinca, A. Balanescu, D. Predeteanu, R. Ionescu, D. Opris-Belinski. *Internal Medicine and Rheumatology, Sf. Maria Clinical Hospital, Bucharest, Romania*

Background: Behcet's Disease is a rare type of vasculitis that involves both arterial and venous blood vessels of all sizes. The type of organ involvement and overall disease activity evaluated in the clinical practice determine the course of treatment and the decision to initiate immunosuppression. Activity scores such as Birmingham Vasculitis Activity score (BVASv3), Behcet's Disease Current Activity Form2006 (BDCAF), or damage indices like Vasculitis Damage Index (VDI) have been developed in this respect.

Objectives: To evaluate the ability of classical immunosuppressant therapy to prevent damage progression. To find the correlation between disease activity scores: BVASv3, BDCAF, long term treatment, immunosuppressant use and damage after remission, as calculated by VDI.

Methods: A study on a cohort of patients diagnosed with Behcet's Disease from an Internal Medicine and Rheumatology Clinic was performed. Activity and damage scores,BVASv3,BDCAF and VDI after obtained remission, were calculated. The documented cases were diagnosed according to the International Criteria for Behçet's Disease (ICBD).Windows Excel/SPSS20.0 (Spearman's correlation)were used to analyse the data.

Results: The study included 16 patients treated with long term cortisone and immunosuppressive therapy. The mean age at the time of the diagnosis was 32.3years with a male predominance 62% (10 patients). Severe systemic involvement was present in 10 cases (Ophthalmological involvement-6cases, recurrent venous thrombosis-6cases, pulmonary vasculitis-1 case, severe cardiac involvement-1case, central nervous system involvement-3cases) and all patients received classical immunosuppression (cyclophosphamide, azathioprine).The mean scores for BVASv3 and BDCAF at the time of the diagnosis were 9 and 4.12.A strong correlation was identified between BVASv3 and BDCAF (r=0.830, p<0,001). The use of immunosuppressive therapy due to severe organ involvement and long-term immunosuppression correlated stronger with BVASv3 (r=0.718) than with BDCAF (r=0.533). Vasculitis damage index (VDI) calculated after remission was obtained. There was an important correlation between disease activity scores and damage (BVASv3-VDI r=0,687,p<0,001, BDCAF-VDI r=0,676, p<0,001). Types of treatment were evaluated, a comparison was made between long-term cortisone therapy and immunosuppression.There was a stronger correlation between long term cortisone use and VDI (r=0,600) than between immunosuppression duration and damage (r=0,472).

Conclusions: Damage progression is influenced by disease activity, as calculated by activity scores (BVASv3 and BDCAF). Classical immunosuppression is used for severe organ involvement and for limiting new organ lesions once started. There was a stronger correlation between long-term cortisone use and VDI than between immunosuppression duration and damage. The damage index increased by irreversible organ damage due to disease activity and long term cortisone use, but not due to the immunosuppressive therapy.

References:

[1] DavatchiF,et al How to deal with Behcet's disease in daily practice. Int J Rheum Dis. 2010 May.

[2] Raashid Ahmed Luqmani Disease assessment in systemic vasculitis. Nephrol Dial Transplant 2015.

Acknowledgements: The first two authors,Sinziana Daia-Iliescu and Casandra Buzatu contributed equally to this study.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.5530

THU0575 EFFICACY OF RITUXIMAB IN RESISTANT PALINDROMIC RHEUMATISM:FIRST REPORT IN LITERATURE

S. Sreenath[1], S. Cherian[1], G. Antony[1], U. Mony[2], P. Shenoy[1]. *[1]Centre for Arthritis and Rheumatism (CARE); [2]Molecular Medicine, AIMS, Cochin, India*

Background: Palindromic rheumatism (PR) although often considered as a benign disease can be severe and resistant to DMARDs in some patients. In these patients it can result in almost daily attacks, migrating from joint to joint resulting in poor quality of life. Rituximab has been proven to be effective in treatment of seropositive RA.

Objectives: To determine the efficacy and safety of Rituximab in patients with seropositive PR who had an inadequate response to CsDMARDs

Methods: PR was diagnosed based on criteria proposed by Hannonen P et al. Seropositive (ACPA±RF positivity) PR patients who had active disease despite being treated with two Cs DMARDs for >3 months, were treated with Rituximab. Active disease was defined as >4 attacks per month requiring intake of NSAIDS. All the patients were started on 500mg of rituximab after baseline work up. If complete control of palindromic attacks was not achieved and B cells were detectable in the peripheral blood by flow cytometry another 500 mg infusion was given after 2 weeks. Patients were continued on maximum tolerable dose of DMARDS. Patients were given repeat infusion of Rituximab once the patient developed clinical relapses as evidenced by recurrence of palindromic attacks.

Results: Twenty three patients with a mean age of 44.60±13.51 yrs and mean disease duration of 5.47±3.25 yrs were included. All patients were positive for ACPA while 17 patients were positive for RF. These patients were on a background of minimum of 2 DMARDs. Despite the maximum tolerable dose of DMARDs they had mean attack rate of 5.30±2.38 attacks per month. During a mean follow up of 14.17±8.62 months seven patients required two infusions and three patients required three infusions. Of the 33 infusions 500 mg was effective in controlling the attacks majority (88%) of the times. Seven patients required another 500 mg infusion after 2 weeks as initial 500 mg dose failed to achieve complete control of disease and B cell were not depleted in the peripheral blood. At one month follow up all patients achieved complete control of disease. Remission lasted for 10.33±5.75 months. When symptoms recurred patients were treated with rituximab again and all regained complete control of the symptoms. None of the patients evolved into RA during the study period. No serious adverse events were observed. Five patients experienced minor allergic reactions during infusion which were managed according to the standard protocol.

Conclusions: This case series indicates in patients of PR resistant to Cs DMARDs rituximab not only achieves disease control but also prevents progression to RA. To best of our knowledge this is the first report regarding efficacy of rituximab in PR. Although it needs to be proved in a larger blinded RCT this early data indicates that Rituximab may be a therapeutic option to prevent development of RA in seropositive patients.

References:

[1] Hannonen P, Möttönen T, Oka M. Palindromic rheumatism. A clinical surveyof sixty patients. Scand J Rheumatol. 1987;16(6):413–420.

[2] Sanmarti R, Cañete JD, Salvador G. Palindromic rheumatism and other relapsing arthritis. Best Pract Res ClinRheumatol. 2004;18(5):647–661.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.5723

THU0576 THERAPEUTIC OPTIONS FOR PATIENTS WITH RARE RHEUMATIC DISEASES: A SYSTEMATIC REVIEW AND META-ANALYSIS

T.T.A. Bender[1], M. Mücke[1,2,3], J. Leyens[1], C. Stieber[1,4], D. Kravchenko[5], M.F. Seidel[6,7]. *[1]Center for Rare Diseases Bonn (ZSEB); [2]Institute of General Practice and Family Medicine; [3]Department of Palliative Medicine; [4]Institute of Human Genetics; [5]University Hospital Bonn, Bonn, Germany; [6]Department of Oncology, Hematology and Rheumatology, University Hospital Bonn, Bonn, Germany; [7]Schmerzklinik Basel, Basel, Switzerland*

Background: Rare rheumatic diseases are challenging for both patients and clinicians. This problem is further propelled by the scarce number of approved treatment regimens. Therapy is often limited to immunosuppression with corticosteroids or off-label use of drugs for common rheumatic diseases.

Objectives: We have recently described a set of 82 classified rare diseases in rheumatology [1]. In this systematic review, we analysed the evidence for therapeutic regimens from randomized clinical trials of rare rheumatic diseases.

Methods: For this systematic review and meta-analysis, we searched the Cochrane Central Register of Controlled Trials (CENTRAL), MEDLINE, PUBMED and SCOPUS, up to 20th of October 2016. To validate the search strategy, we selected sentinel references.

We included randomized controlled trials regarding rare rheumatic diseases and put a focus on pharmacological treatment compared with placebo, application of