Methods: In January 2016, the Autoimmune Diseases Study Group (GEAS-SEMI) created a national registry (SARCOGEAS) of patients with sarcoidosis. Sarcoidosis was diagnosed in agreement with the criteria proposed by the ATS/ERS/WASOG 1999 statement, and extrathoracic disease was classified with the 2014 WASOG instrument.
Results: The cohort consisted of 1082 patients (82% biopsy-proven), including 618 (57%) women and 464 (43%) men, with a mean age at diagnosis of 47yrs; 140 (13%) patients were born outside Spain, 965 (89%) were White, 69 (6%) Hispanic, 30 (3%) Black/African American and 18 (2%) Asian. Thoracic involvement was present at diagnosis in 979 (90%) patients, including 437 (40%) patients with stage I, 374 (35%) with stage II, 123 (11%) with stage III and 26 (2%) with stage IV. The most frequently reported extrathoracic involvements at diagnosis were cutaneous in 385 (36%) patients, extrathoracic lymph nodes in 218 (20%), liver involvement in 151 (14%) and ocular involvement in 118 (11%). Potentially life-threatening WASOG involvements were reported in frequencies less than 10%, including neurological involvement in 77 (7%) patients, kidney involvement in 59 (5%) or cardiac involvement in 21 (2%). Therapeutic approaches at diagnosis included the use of oral glucocorticosteroids in 637 (59%) patients, immunosuppressive agents in 84 (8%, mainly methotrexate in 63 patients) and biological agents in 15 (1%, mainly infliximab in 10 cases).
Conclusions: In this large series of sarcoidosis from Southern Europe, clinical presentation is dominated by adenopathies (both thoracic and extrathoracic) and cutaneous involvement (erythema nodosum), with lower frequencies in the main extrathoracic involvements than that reported in US and Japanese series.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.4387

THU0540 EFFICACY AND SAFETY OF ADALIMUMAB IN BEHÇET'S DISEASE RELATED UVEITIS: A MULTICENTER RETROSPECTIVE OBSERVATIONAL STUDY

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Background: Current information on the use of adalimumab (ADA) in the treatment of ocular Behçet's disease (BD) is still based mainly on small series or case reports; nonetheless preliminary evidence is promising.
Objectives: The study aim was to evaluate the efficacy of ADA in a large series of BD-related uveitis.
Methods: We performed a multicenter retrospective observational study including 40 selected patients (66 eyes) receiving ADA. Clinical data were retrospectively analyzed at baseline, at 3 and 12 months of treatment. Primary end-point was: reduction of ocular inflammatory flares. Secondary end-points were: improvement of Best Corrected Visual Acuity (BCVA), reduction of macular thickness measured by optical coherence tomography (OCT), reduction in the occurrence of vasculitis assessed by fluorescein angiography (FA), evaluation of statistically significant differences between patients treated with ADA monotherapy and those undergoing ADA plus DMARDs and in patients firstly treated with ADA compared to patients previously administered with other biologics; ADA steroid sparing effect was also evaluated.
Results: During the first 12 months of ADA therapy the number of flares significantly decreased from 200 flares/100 patients/year to 8.5 flares/100 patients/year ($p<0.0001$). Similarly BCVA improved if compared to baseline (7.4±2.9 versus 8.5±2.1, $p=0.03$). OCT findings significantly improved showing a mean reduction of central macular thickness (CMT) of 27.27±42.8 microns at the end of follow up ($p<0.006$). FA identified retinal vasculitis in 22 cases at baseline (55%), 8 (20%) cases after 3 months and in only one (2.5%) case at 12-month follow-up. FA improvement was highly significant at 3 and 12-month follow-up if compared to baseline ($p<0.0001$ and $p=0.006$, respectively).
Conclusions: ADA is highly effective and safe for the treatment of BD-related uveitis, providing a long-term control of ocular inflammation.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.1026

THU0541 ROLE OF IL-1 INHIBITION IN ADULT ONSET STILL'S DISEASE (AOSD): A RETROSPECTIVE, OBSERVATIONAL MULTICENTRIC STUDY FROM THE ITALIAN SOCIETY OF RHEUMATOLOGY STUDY GROUP ON AUTOINFLAMMATORY DISEASES

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Background: IL-1 pathway plays a major role in the pathogenesis of Adult onset Still's disease (AOSD). IL-1 inhibitors [Anakinra (ANK) and Canakinumab (CAN)] proved their efficacy in AOSD as demonstrated in several case-reports and small case-series. However, due to the disease rarity, large randomized control trials are still lacking.
Objectives: To retrospectively evaluate the efficacy and safety of the use of IL-1 inhibitors (ANK and CAN) in patients with AOSD.
Methods: Data from patients with diagnosis of AOSD (Yamaguchi criteria) referred by 18 different Italian centres were retrospectively collected. Clinical and serological features as well as concomitant treatments, were collected on a dedicated database at the beginning of anti-IL-1 treatment (baseline) and then after 3, 6 and 12 months of follow-up. The Pouchot's score was used to evaluate disease activity.
Results: Data from 140 patients were collected (Table). All were treated with ANK and 4 were subsequently switched to CAN after ANK failure. The systemic pattern vs the chronic-articular were present in ANK and CAN groups in 104/140 (74.2%) and 3/4 (75%), respectively, vs 48/140 (25.8%) and 1/4 (25%), respectively. MTX was the most commonly used DMARD before ANK or CAN [91/140 (75.8%), 2/4 (50%) respectively]. In ANK group, the most commonly used previous biologic drug (bDMARD) was etanercept (79.3%). ANK was adopted as second-line bDMARD in 29/140 (20.7%) patients. In most of the cases it was administered at 100 mg/day in 126/140 patients (90%). CAN was employed at 150 mg every 8 weeks. ANK was effective on all AOSD clinical (fever, rash, pneumonia, pericarditis, pleuritis, sorethroat, lymphadenopathy, hepatomegaly, myalgia, arthritis, MAS) and serological (increased liver enzymes, hyperferritinemia, leucocytosis) manifestations ($p<0.0001$). The Pouchot's score was significantly reduced at all time points ($p<0.0001$). In ANK group no difference in treatment response was identified stratifying patients according to age, sex, pattern of disease, monotherapy vs combination. ANK primary and secondary inefficacy after 12 months were 15/140 (10.7%) and 11/140 (7.8%) cases, respectively. Adverse events (AEs) [mainly represented by in situ (28/47, 59.5%) or diffused (12/47, 25.5%) skin reactions and infections (12/47, 12.7%)] were the main reason for ANK discontinuation. Similarly, in CAN group Pouchot's score and clinical and serological features significantly ameliorated at all time points

Clinical features	ANK group	CAN group
Sex (M/F)	47/93	0/4
Age at onset (mean±SD, years)	35.4 ± 17	34.7 ± 13.3
Age at diagnosis (mean±SD, years)	37.4 ± 16.1	34.2 ± 15.4
Disease duration before treatment (mean±SD, months)	50.33 ± 81.67	58.33 ± 48.4
Previous therapies:	137/140 (97.8%)	4/4 (100%)
- STEROIDS	120/140 (85.7%)	2/4 (50%)
- DMARDs	29/140 (20.7%)	2/4(50%)
- bDMARDs		
Monotherapy	34/140 (24.2%)	2/4 (50%)
Combination therapy (+DMARDs)	106/140 (75.8%)	2/4 (50%)
N° Patient baseline	140 (100%)	4/4 (100%)
N° patients 3 months	118 (84.2%)	4/4 (100%)
N° patients 6 months	109 (77.8%)	4/4 (100%)
N° patients 12 months	97 (69.2%)	3/4 (75%)
N° patients at the time of this study	69 (49.2%)	2/4 (50%)
Mean duration of therapy (mean±SD, months)	35.7 ± 36.1	22.1 ± 16.5
AEs	47/140 (33.5%)	0/4 (0%)
Reasons of discontinuation:		
-AEs	24/71 (33.8%)	0/4 (0%)
-Remission	20/71 (28.1%)	1/4 (25%)
-Primary inefficacy	16/71 (22.5%)	0/4 (0%)
-Loss of efficacy	11/71 (15.4%)	1/4 (25%)
Pouchot's score at baseline	5.5 ± 1.9	4.2±2.6
Pouchot's score at 3 months	1.1 ± 1.4	1.2±1.8
Pouchot's score at 6 months	0.6 ± 1.2	1.5±1.9
Pouchot's score at 12 months	0.4 ± 0.8	1±1.4
% patients with steroids (baseline)	137/140 (97.8%)	4/4 (100%)
% patients with steroids (time of the study)	22/69 (31.8%)	2/2 (100%)
% patients with DMARDs (baseline)	120/140 (85.7%)	2/4 (50%)
% patients with DMARDs (time of the study)	35/69 (50.7%)	0/2 (0%)