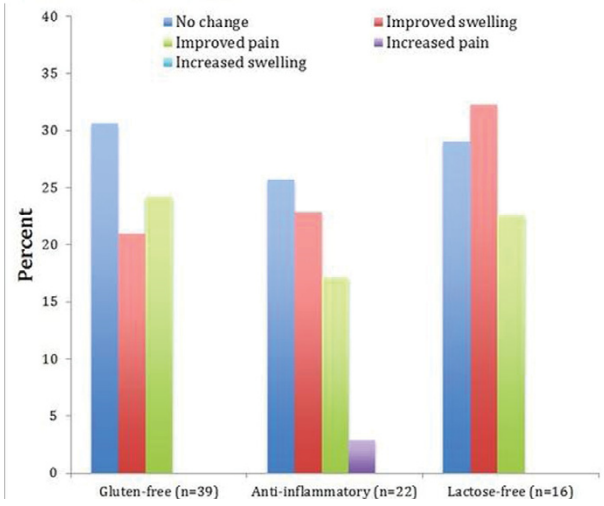
and a lactose-free diet (26%). There were similar clinical responses among the three diets (Figure 1). Twenty-five to thirty percent reported no change in joint symptoms whereas 20–30% reported improved pain or joint swelling. Sixty-one (34%) parents were willing to participate in a 3-month dietary intervention study and 78 (44%) parents answered "it depends".

Table 1

Population	Adult patients (n=49)	Pediatric patients (n=178)
Number of joints affected, n (%)		
≥5	43 (88)	125 (70)
<5	5 (10)	51 (29)
Treatment exposure, n, (%)		
Systemic glucocorticoid	40 (82)	108 (61)
DMARDs	44 (90)	146 (82)
Biologicals	35 (71)	114 (64)

Figure 1. Parental report of clinical responses to a gluten-free diet, an anti-inflammatory diet and a lactose-free diet in children



Conclusions: This is the first report of the family/patient perspective of the role of dietary intervention on JIA. Almost half of the affected patients attempted special diets, and many reported improvement in symptoms. Future interventional studies with objective outcome measurements are needed to validate these reports.

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Other orphan diseases

THU0538 EFFECTS OF GLUCOCORTICOIDS AND METHOTREXATE-BASED THERAPEUTIC REGIMENS ON B CELL SUBPOPULATIONS IN PATIENTS WITH IGG4-RELATED DISEASE

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Background: IgG4-related disease (IgG4-RD) is a systemic fibro- inflammatory disorder characterized by fibrotic lesions infiltrated by IgG4 positive plasma cells (1). The prompt clinical responses obtained after B cell depletion with rituximab in IgG4-RD patients suggest that B lymphocytes drive the pathogenesis of this condition and sustain disease activity (2). This conclusion, however, requires further confirmation because IgG4-RD responds also to non-B cell depleting therapies such as glucocorticoids and methotrexate

Objectives: To evaluate the effects of glucocorticoids and methotrexate-based therapeutic regimens on B lymphocyte subpopulations in patients with IgG4-RD.

Methods: Sixteen patients with active IgG4-RD were studied. FACS analysis was performed on peripheral blood in order to identify the following B cell subpopulations: total B cells (CD19+CD20- and CD19+CD20+ cells), circulating plasmablasts (CD19+CD20- CD27+CD38++ cells), naïve B cells (CD19+CD20+CD27-CD38+ cells), memory B cells (CD19+CD20-CD27+CD38- cells), circulating plasma cells (CD38+CD138+ cells). Disease activity was assessed by means of the IgG4-RD responder index (IgG4-RD RI). Flow cytometry was performed at baseline and after six months of immunosuppressive therapy with glucocorticoids (0.6–1mg/kg/day) and/or methotrexate (10–20mg/week). 16 sex and age matched healthy subjects were used as controls.

Results: At baseline, circulating plasmablasts were expanded in IgG4-RD patients (median 3780 cell/mL; range 330–9300) compared to controls (median 280 cell/mL; range 0–1000) ($p<0.05$); total B cells (median 133000 cell/mL; range 34000–569000) and naïve B cells (median 13080 cell/mL; range 1970–64270) were reduced in IgG4-RD patients compared to controls (median 280 cell/mL; range 194–330; and median 54020 cell/mL; range 21050–106780, respectively) ($p<0.05$). No circulating plasma cells were detected in healthy controls. No differences in memory B cells were observed ($p>0.05$). Circulating plasmablasts but not other B cell subsets positively correlated with serum IgG4 levels, number of organ involved, and IgG4-RD RI ($p<0.05$). At six months follow-up, the median IgG4-RD RI decreased from 9 to 2. Circulating plasmablasts, circulating plasma cells, and naïve B cells counts decreased in all patients together with disease improvement (p=0.0002, 0.0002 and 0.025 compared to baseline values, respectively); total B cells and memory B cells were unaffected by immunosuppressive therapy.

Pt	Age (years)	Sex	Organ involvement	Atopy	Eosinophil (cell/uL) (<400)	ESR (mm/hr) (<12)	CRP (mg/dL) (<6)	IgG4 (mg/dL) (<135)	PBL (cell/mL) (<650)	Naive B Cell (cell/mL)	Memory B Cell (cell/mL)
1	83	F	Pancreas - Biliary tree	-	100	13	46	1360	9000	8940	26940
2	73	F	Pancreas - Lymph nodes	+	300	4	Neg	534	7000	13030	15750
3	53	F	Aorta	-	500	21	21.3	185	6000	59800	107710
4	69	M	Pancreas - Lungs	-	300	21	46.3	343	5400	1970	6590
5	73	M	Pancreas – Biliary tree - Lungs	-	500	7	Neg	4900	9000	8370	9340
6	69	F	Aorta	-	300	38	14	420	1000	20830	55040
7	64	M	Pancreas	-	470	-	-	308	4000	13130	11060
8	57	M	Lacrimal glands – Salivary glands - Pancreas	+	300	<12	<6	1120	2020	30310	31210
9	60	M	Lymph nodes	-	200	15	10	313	5130	3370	48590
10	76	M	Biliary tree	-	200	-	<6	314	2150	56490	27760
11	66	M	Pancreas	-	600	59	<6	362	1760	43510	21600
12	54	M	Lymph nodes	+	1800	?	?	799	9300	18420	21880
13	70	M	Pancreas – Lymph nodes	-	300	20	<6	221	3560	11220	9980
14	79	M	Salivary glands- Aorta - Pancreas	-	200	19	<6	498	2520	64270	6860
15	67	M	Pancreas	-	200	-	-	586	940	9900	20070
16	61	M	Pancreas	-	100	<12	<6	137	330	12490	25970

Conclusions: Non-B cell depleting therapies based on glucocorticoids and/or methotrexate induce clinical improvement and deplete circulating plasmablasts, plasma cells and naïve B cells in patients with IgG4-RD; circulating total B cells and memory B cells are not affected by glucocorticoids and methotrexate. Our study, performed with non-B cell depleting agents, provides clinical evidences that circulating plasmablasts are likely linked to IgG4-RD pathogenesis and disease activity.

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THU0539 SARCOIDOSIS IN SPAIN: CLINICAL AND EPIDEMIOLOGICAL CHARACTERISTICS AT DIAGNOSIS IN 1082 PATIENTS

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Objectives: To characterize the main features at presentation of sarcoidosis in a large multicenter cohort from Southern Europe.

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
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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
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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%)