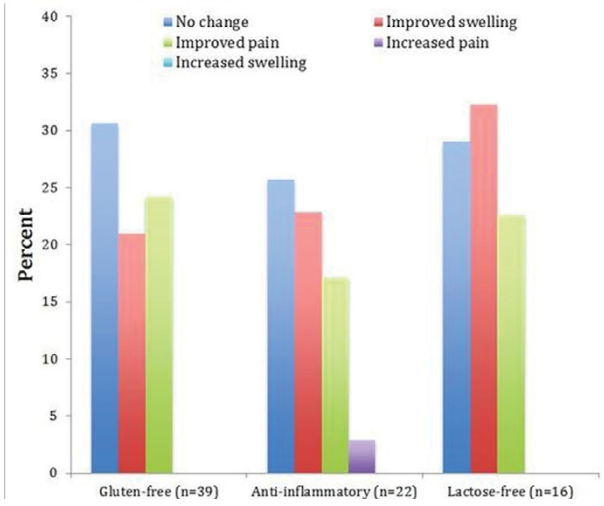
and a lactose-free diet (26%). There were similar clinical responses among the three diets (Figure 1). Twenty-five to thirty percent reported no change in joint symptoms whereas 20–30% reported improved pain or joint swelling. Sixty-one (34%) parents were willing to participate in a 3-month dietary intervention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