biologic switchers (34.5%). For the total IV ABA cohort, six hospitalized infection claims were reported with an IR (95% CI) of 2.4/100 p-y (0.9, 5.3) and an IR of 2.8/100 p-y (1.2, 5.9) for new-onset uveitis. There were no validated cases of malignancies in the follow-up period.

Conclusions: Compared with an overall JIA population[1], abatacept pts are slightly older, more likely to use additional prior biologics, and have a history of asthma or cardiovascular disease. The rates of hospitalized infection and new onset of uveitis in this study are within published ranges[2,3] and are consistent with findings in the abatacept JIA registry.[4]

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THU0535 LONG-TERM FOLLOW-UP OF 12 CASES OF CORONARY GIANT ANEURYSM AFTER KAWASAKI DISEASE

T. Sato[1], J. Somura[1], S. Hoshino[1,2,3], O. Furukawa[1], N. Okamoto[2,4], Y. Maruo[1]. *[1]Department of Pediatrics, Shiga University of Medical Science; [2]Department of Pediatrics, Omihachiman Community Medical Center; [3]Department of Pediatrics, Nagahama Red Cross Hospital; [4]Department of Pediatrics, Okamoto Kids Clinic, Shiga, Japan*

Background: The incidence of Kawasaki disease has been increasing since it was first reported by Tomisaku Kawsasaki in 1967. Among complications of the condition, the formation of coronary artery aneurysms is the most important. In particular, giant aneurysms with diameters that exceed 8 mm are likely to not regress and result in serious complications, such as acute myocardial infarction.

Objectives: To understand the long-term course of patients with giant aneurysms and Kawasaki disease as well as to consider the cause of aneurysm formation and its appropriate treatment.

Methods: We retrospectively studied the long-term course of 12 cases of giant coronary artery aneurysms accompanied with Kawasaki disease, which were being followed at Shiga University of Medical Science Hospital, Omihachiman Community Medical Center, and Nagahama Red Cross Hospital. These are three major facilities in Shiga prefecture of Japan, whose population is 1.4 million, comprising 200,000 children.

Results: Ten male and two female patients were included. The average current age was 16.8 years, the median age was 14.3 (10.7–18.9) years, and 5 cases were of adults. The average age at the time of onset was 3.5 years, the median age was 3.7 years (1.8 - 4.4), and all experienced onset between 1 and 5 years of age. The mean period from onset to treatment start was 6.7 days (median 5.0 (4.3–8.3)), but the average period until fever declined was 16.0 days and only three patients' temperature was reduced in 10 days. Aneurysm formation occurred at 14.1 days on average (median 12 (10–17)). The average size of the maximum coronary artery aneurysm at onset was 11.3 mm, and the median size was 9.5 mm (8.8 mm – 13.8 mm).The average and median follow-up periods were 13.2 years and 11.7 years (5.4–13.4), respectively. The number of patients received steroid therapy was four, and all their onset was after 2006. None received infliximab or underwent plasmapheresis.

During the course of the condition, all patients underwent multiple centripetal echocardiography. Among all cases, 7 underwent coronary angiography CT, 10 underwent myocardial scintigraphy. All 12 patients underwent cardiac catheterization and the total number of underwent cardiac catheterization for them was thirty-one. Two adult patients had a history of acute myocardial infarction and had undergone cardiac bypass surgery. Through this survey, we found that 9 cases developed giant coronary artery aneurysm between 1983 and 2007, and 3 cases between 2012 and 2015 used prednisolone.

Conclusions: All patients are currently receiving anticoagulant therapy and undergoing diagnostic imaging. In our prefecture, the incidence of giant coronary artery aneurysm accompanied with Kawasaki disease has been decreasing gradually. From 2007 to 2012, in which high dose gamma globulin therapy (2g/kg) has become commonly underwent for Kawasaki disease in Japan, there were no giant aneurysm formation in our hospitals. Three patients in whom giant aneurysms developed between 2012 and 2015 were taking oral prednisolone, thereby suggesting a relationship between prednisolone and giant aneurysm formation.

Disclosure of Interest: None declared

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THU0536 CHILDHOOD ONSET OF BEHÇET DISEASE (BD) SYMPTOMS IN AN ADULT COHORT OF BD PATIENTS

V. Paisal[1], E. Al-Abadi[1], S. Protheroe[2], D. Carruthers[3], D. Situnayake[3], S. Powell[4], D. Mitton[4], T. Southwood[1]. *[1]Paediatric Rheumatology; [2]Paediatric Gastroenterology, Birmingham Children's Hospital; [3]Behçet Syndrome National Centre of Excellence, Birmingham and Midland Eye Centre; [4]Behçet Syndrome National Centre of Excellence, Birmingham and Midland Eye Centre, Birmingham, United Kingdom*

Background: Behçet Disease (BD) is rarely reported in children. It a systemic inflammatory condition characterised by autoinflammatory and vasculitic clinical features including recurrent oral aphthosis, genital ulceration, skin, eye, neurological and vascular inflammation, which is most commonly diagnosed between 20–40 years of age. It is unclear how many adult patients with BD have the onset of symptoms during childhood.

Objectives: The aim of our study was to investigate the age of symptom onset in a large cohort of adult patients with BD.

Methods: Since 2012, the Behçet Syndrome National Centre of Excellence, Birmingham and Midland Eye Centre, has used a multidisciplinary approach aiming to shorten time to diagnosis of BD, reduce blindness and morbidity, improve knowledge and ensure equity in access to biological therapies. Patient demographic and disease information, including patient recalled age at symptom onset, symptom sequence, documented clinical signs, treatment and outcome data are maintained in a secure web-based clinical data archive. For this study, the database was interrogated to determine patient age at recalled onset of the first BD symptoms, the sequence in which symptoms appeared and whether the ISG classification criteria for the diagnosis of BD were fulfilled before the age of 16 years.

Results: To June 2016, 478 patients aged between 11–68 years had data recorded in the BD database. 60 patients (12.7%, 49 females) who fulfilled International Study Group criteria for the diagnosis of BD, reported that the onset of their first symptoms was before the age of 16 years (range 2–15 years). All 60 patients reported recurrent oral ulceration and 55 reported genital ulceration but with a reported time interval between the onset of these 2 features of 0–44 years. Of the 60 patients, 26 patients reported sufficient symptoms to have fulfilled ISG criteria before their 16th birthday, including the coincident onset of recurrent oral and genital ulceration in 15/26 patients. Eye disease was only reported in 1 patient and a positive family history of BD in 5/26 patients. 11/26 patients reported a delay between the onset of oral and genital ulcers of a mean of 5.45 years (range 1–11 years).

Conclusions: Many adults with BD recall the onset of their first symptoms before their 16th birthday, usually with recurrent oral and genital ulceration. We speculate that greater awareness of BD in childhood may reduce the delay in diagnosis of this chronic condition.

Disclosure of Interest: None declared

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THU0537 FAMILY AND PATIENT'S PERCEPTION OF DIETARY INTERVENTION IN JUVENILE IDIOPATHIC ARTHRITIS (JIA)

E.M. Little[1], S. Grevich[2], J.L. Huber[3], D.L. Suskind[4,5], M.C. Bradford[4], A.M. Stevens[2,6], Y. Zhao[2,4]. *[1]Department of Pediatrics, Alaska Native Medical Center, Anchorage; [2]Pediatric Rheumatology, Seattle Children's Hospital, Department of Pediatrics, University of Washington; [3]Division of Nutrition, Seattle Children's Hospital; [4]Center for Clinical and Translational Research, Seattle Children's Research Institute; [5]Gastroenterology, Seattle Children's Hospital, Department of Pediatrics, University of Washington; [6]Center for Immunity and Immunotherapies, Seattle Children's Research Institute, Seattle, United States*

Background: Juvenile idiopathic arthritis (JIA) is the most common pediatric rheumatologic illness and can lead to significant disability. Complementary and alternative treatments are commonly practiced by families of patients with JIA, and >40% of patients with chronic arthritis seek dietary changes after their diagnosis. Dietary intervention studies in adults with rheumatoid arthritis showed moderate improvement in joint symptoms. Dietary supplements of omega-3 fatty acids have been tested in children with chronic arthritis and found to be associated with less NSAID use and lower serum IL-1 and TNF levels. There is an increasing need to understand if there is a role for dietary therapy in chronic arthritis.

Objectives: We aimed to evaluate the prevalence of special diets and the perception of the effectiveness of these diets on arthritis in JIA. We also assessed the interest of dietary interventions and perceived barriers.

Methods: An online survey was designed through a REDCap database capturing demographic information, self- or parent-initiated special dietary interventions and self- or parent-observing effects on joint symptoms, willingness to participate in a dietary intervention study. The survey link was posted on social media websites and distributed by the Arthritis Foundation. Descriptive statistical analyses were performed.

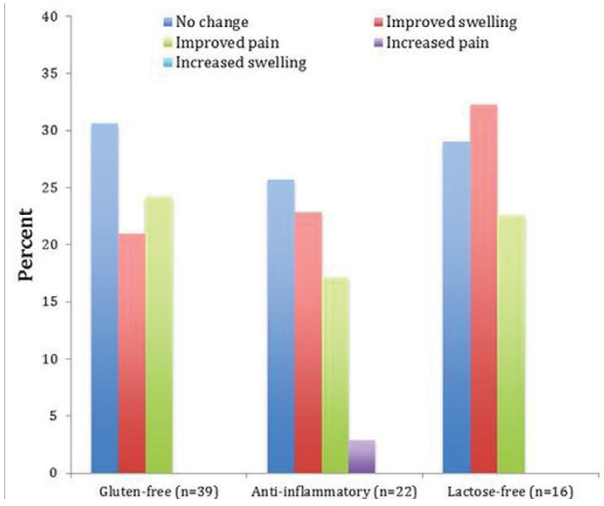
Results: A total of 265 responses were received from adult patients who had JIA and parents of children with JIA. We excluded 14 patients with inflammatory bowel disease or celiac disease-related arthritis and 24 responders with incomplete answers. Demographic and JIA characteristics of adult and pediatric patients are listed in Table 1. Ninety patients (63 children with JIA, 27 adults with history of JIA) had tried a special diet for arthritis. The top three special diets reported by parents included a gluten-free diet (62%), an anti-inflammatory diet (53%),

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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THURSDAY, 15 JUNE 2017

Other orphan diseases

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

M. Lanzillotta[1], E. Della Torre[1], R. Milani[2], E. Bozzalla[1], L. Rovati[1], E. Bozzolo[1], M. Falconi[3,4], L. Dagna[1,4]. *[1]Internal Medicine, Allergy and Clinical Immunology; [2]Unit of Immuno-hematology and Bone Marrow Transplant; [3]Pancreatic Surgery Unit, San raffaele Scientific Institute; [4]Università Vita-Salute San Raffaele, Milan, Italy*

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

R. Pérez-Alvarez[1], B. Kostov[2], A. González García[3], R. Gómez De La Torre[4], M. Lopez Dupla[5], B. De Escalante[6], A. Alguacil[7], J. Chara[8], J. Velilla[9], J. Rascón[10], J.S. Garcia Morillo[11], C. Feijoo[12], E. Fonseca[13], M. Bonet[14], J.L. Callejas[15], G. de la Red[16], E. Calvo[17], C. Soler[18], E. Peral[19], J.F. Gómez Cerezo[20], G. Cruz-Caparrós[21], P. Perez[22], S. Rodríguez Fernández[23], B. Pinilla[24], A. Gato[25], M. Akasbi[26], A. Robles[27], I. Ojeda[28], M.J. Vives[29], C. Morcillo[30], M. Penadés[31], M. De Vicente[32], M. Ramos-Casals[33], L. Pallarés[10], P. Brito-Zerón[30,33] on behalf of the SARCOGEAS-SEMI Registry. *[1]Hosp Alvaro Cunqueiro, Vigo; [2]IDIBAPS, Barcelona; [3]Hosp Ramón y Cajal, Madrid; [4]HUCA, Oviedo; [5]Hosp Joan XXIII, Tarragona; [6]Hosp Clínico, Zaragoza; [7]Hosp Virgen de la Salud, Toledo; [8]Hosp Josep Trueta, Girona; [9]Hosp Miguel Servet, Zaragoza; [10]Hosp Son Espases, Palma de Mallorca; [11]Hosp Virgen del Rocio, Sevilla; [12]Hosp Parc Taulí, Sabadell; [13]Hosp de Cabueñes, Gijón; [14]Althaia, Manresa; [15]Hosp San Cecilio, Granada; [16]Hosp Esperit Sant, Santa Coloma; [17]Hosp General San Jorge, Huesca; [18]Hosp de Sta Caterina, Girona; [19]Hosp Virgen Macarena, Sevilla; [20]Hosp Infanta Sofía, Madrid; [21]Hosp de Poniente, Almería; [22]Hosp Puerta del Mar, Cádiz; [23]Hosp da Barbanza, A Coruña; [24]Hosp Gregorio Marañón, Madrid; [25]CH, Albacete; [26]Hosp Infanta Leonor; [27]Hosp la Paz, Madrid; [28]Hosp Valle del Guadiato, Córdoba; [29]San Joan de Déu, San Boi; [30]Hosp CIMA-Sanitas, Barcelona; [31]Hosp de Manises, Valencia; [32]Hosp Nuestra Señora del Prado, Talavera; [33]Hosp Clínic, Barcelona, Spain*

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