THU0532 PAEDIATRIC UVEITIS IN FRENCH REFERRAL OPHTHALMOLOGIC CENTERS: A DESCRIPTIVE ANALYSIS OF 74 CHILDREN

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Background: Uveitis in children is rare. Intensive interactions between ophthalmologists and paediatric rheumatologists are needed in order to choose the best therapeutic strategies for severe uveitis attacks.
Objectives: Describe a cohort of 74 patients with paediatric uveitis.
Methods: Retrospective analysis of children followed for uveitis before 18, by one paediatric rheumatologist (SGC) for systemic treatments' management and members of 3 ophthalmologic departments specialized in uveitis care in children (AR, CT, ML and BB) in Paris, during the 2006–16 period.
Results: There were 74 paediatric uveitis, 42 anterior (57%, group1), 16 intermediate (21%, gr2), 7 posterior (9%, gr3) and 9 pan-uveitis (12%, gr4). Gender was equal in gr2–4, but there were more females in gr1. At presentation, mean ages were 8.6±4.1, 9.8±3.9, 9.1±3.6 and 10±4.2 years old. Mean follow-up was 3.7±3.7 years. JIA was the leading cause of gr1 uveitis (45%); gr2–3 uveitis were idiopathic in 81% and 86%, respectively. In gr4, etiologies were found in 7 out of 9 patients (Behçet-3, JIA-2, BBS-1, TINU-1).

Table 1

Complications	I: Anterior uveitis (42) 51%	II: Intermediate uveitis (16) 71%	III: Posterior uveitis (7) 71%	IV: Panuveitis (9) 100%
Cataracts	25%	21%		44%
Papilledema	19%	21%		56%
HTP/Glaucoma	16%			
Macular edema			29%	33%
Vitreous hemorr.		21%		33%
Retinal detach.		29%		33%
Blindness	6% (n=2Uni)	7% (n=1Uni)	29% (n=2Bi)	33% (n=2Uni+1Bi)

Conclusions: Paediatric uveitis induce a very high-level burden in children, even when anterior and sometimes despite optimal therapeutic management in tertiary care centers. Their early recognition and tight control in specialized units are absolutely required in order to decrease the level of definitive complications.
Disclosure of Interest: None declared
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THU0533 DO JIA CORE OUTCOME VARIABLES AT BASELINE PREDICT CLINICALLY INACTIVE DISEASE STATES AT ONE YEAR?

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Background: Identifying predictors for early clinically inactive disease (CID) would allow stratified treatment decisions at diagnosis, minimising the burden of unnecessary therapies. JIA core outcome variables (COVs) are routinely collected and would therefore be convenient predictors. However, different groups of children are identified by current CID definitions (Wallace's preliminary criteria vs. clinical Juvenile Arthritis Disease Activity Score in 10 joints (cJADAS10)) and predictors may differ depending on which CID outcome is applied.
Objectives: To assess whether baseline COVs predict achievement of CID according to Wallace's preliminary criteria or the cJADAS10 cut-off in patients with JIA.
Methods: Children and young people enrolled to the Childhood Arthritis Prospective Study (CAPS), a UK multicentre inception cohort, before January 2011, were selected if diagnosed with oligoarticular, RF-negative or RF-positive polyarticular JIA.
At one year following initial presentation to paediatric rheumatology, children were classified as i) CID according to Wallace's preliminary criteria and ii) CID according to cJADAS10. Baseline COVs (active joint count, limited joint count, physician's global, parental global, functional ability (Childhood Health Assessment Questionnaire (CHAQ)) and ESR) were tested for predictive ability for these outcomes using univariate and forced-entry multivariate logistic regressions, adjusting for age and symptom duration at initial presentation, gender and ILAR subtype. Multiple imputation accounted for missing data.
Results: Of 829 children included, 70% were female and the majority had oligoarticular JIA (68%). At one year, 28% had achieved CID according to Wallace's preliminary criteria and 38% according to the cJADAS10 (21% CID on both).
In univariate analyses, increased baseline CHAQ and physician's global assessment score predicted lower odds of achieving both CID states. In addition, increased active joints (OR: 0.97, 95% CI0.94, 0.99), and patient/parent global assessment scores (OR: 0.87, 95% CI 0.81, 0.93) predicted lower odds of CID on the cJADAS10 only. In multivariate analyses, one increased CHAQ point at baseline independently predicted 28% lower odds of CID on the cJADAS10 (95% CI 0.53, 0.98). However, no baseline COVs predicted CID on Wallace's preliminary criteria. No demographic variables were significantly predictive in any model.
Conclusions: There were different predictors for CID on the cJADAS10 vs. Wallace's preliminary criteria. Children with poor functional ability at initial presentation are less likely to achieve CID on the cJADAS10. These children could be targeted with more aggressive treatment strategies to better control their disease.
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THU0534 BASELINE CHARACTERISTICS AND DESCRIPTIVE SAFETY DATA OF INTRAVENOUS ABATACEPT-TREATED PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS IN A US HEALTHCARE CLAIMS DATABASE

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Background: Abatacept (ABA), the first selective co-stimulation modulator approved and used for the treatment of moderate-to-severe polyarticular juvenile idiopathic arthritis (JIA), has a mechanism of action that is different from other biologic (b)DMARDs.
Objectives: To describe the baseline (BL) characteristics and validated safety outcomes of patients (pts) with a diagnosis of JIA in a US healthcare claims database treated with IV ABA and those with a minimum of 12 months (M) of ABA treatment.
Methods: Pts aged <18 years (yrs), diagnosed with JIA and in the Truven Health MarketScan® database between 1 Jan 2006 and 30 Sep 2014 were eligible for inclusion. Pts were required to have ≥180 days (d) of continuous health plan enrolment prior to a diagnosis of JIA based on two International Classification of Diseases, Ninth Revision, Clinical Modification codes (714.3x) within 90 d. BL characteristics and prior bDMARD use were analysed for two IV ABA-treated groups: a total cohort and a subgroup of pts with ≥12M of treatment. Three categories evaluated prior bDMARD use claims: biologic holiday (other bDMARD in history but not within 180 d of BL), biologic switcher (other bDMARD within 180 d of BL) and true initiator (no history of other bDMARD). Incidence rates (IR; number of events/person yrs [p-y] of exposure) per 100 p-y of exposure with 95% CI were calcuated for infections, malignancies and uveitis.
Results: A total of 238 IV ABA-treated pts were identified with 89 pts having ≥12M of ABA treatment. The mean (SD) follow-up duration was 1.73 (1.28) yrs for the total IV ABA cohort and 2.28 (1.03) yrs for the ≥12-M subgroup. Most pts were female, and mean age was 12.4 yrs. Overall, the total IV ABA cohort was more likely to have a claim in the BL period for asthma and cardiovascular disease versus the ≥12-M subgroup; the ≥12-M subgroup was more likely to have uveitis (Table). The most frequent other bDMARD claim for the total IV ABA-treated pts in the biologic holiday group was etanercept (41.9%), and adalimumab for

BL Characteristics	Total IV ABA cohort (n=238)	IV ABA ≥12-M subgroup (n=89)
Female, n (%)	195 (81.9)	78 (87.6)
Age, mean (SD)	12.4 (3.2)	12.4 (3.2)
Uveitis, n (%)	22 (9.2)	11 (12.4)
Asthma, n (%)	21 (8.8)	6 (6.7)
Cardiovascular disease, n (%)	18 (7.6)	6 (6.7)
bDMARDs, n (%)	84 (35.3)	30 (33.7)
Non-biologic DMARDs, n (%)	105 (44.1)	37 (41.6)
Inpatient visits, mean (SD)	0.2 (0.8)	0.1 (0.3)
Outpatient visits, mean (SD)	11.6 (10.7)	10.0 (9.8)

Abstract THU0532 – Table 2

Treatments	I: Anterior uveitis (42)	II: Intermediate uveitis (16)	III: Posterior uveitis (7)	IV: Panuveitis (9)
High dose systemic steroid	45% (19)	80% (12)	71% (5)	100% (9)
Synth DMARDs	66% (27) (MTX-26, AZA-2)	60% (9) (MTX, AZA)	71% (5) (MTX, AZA)	78% (7) (MTX, AZA, COL)
SynthDMARDs + Biologics	34% (14) (IFX-4, ADA-10)	33% (5) (IFX-1, TCZ-1, IFN-4)	43% (3) (IFX-2, IFN-1)	33% (3) (IFX-3)
Surgery	18% (7) (cataracts – 5, glaucoma – 1, keratopathy – 1)	20% (3) (cataracts – 3, vitrectomy – 1)	0	63% (5) (cataracts – 2, glaucoma – 1, vitrectomy – 1, antiangio. inj.– 1)

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

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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

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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.

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

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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%),