other health professionals working in the field of autoimmunization were asked to identify the variables that they consider as important, in the current clinical practice, for the diagnosis of patients with inherited periodic fever and PFAPA. This survey was open to not influence the experts.

Results: We sent the first survey to 129 experts. The overall rate of response was 102 (79%). 88 experts responded to be interested in the survey and 72 completed and confirmed it; 6 experts responded not to be interested. Up to date, we have analyzed 44 experts’ survey. Not all the participants completed the survey for each disease. The table shows the preliminary results of the analysis. There was any clinical variable chosen by all the participants for the five diseases. For a high rate of experts, a positive genetic test is a relevant element for the diagnosis of FMF, TRAPS and MKD. Conversely, for the PFAPA the exclusion of the inherited genetic periodic fever by genetic test is important for a little part of experts. The response to treatment was proposed as interesting variable for FMF, CAPS and PFAPA.

Conclusions: The preliminary results of the first Eurolever Delphi Survey show a high rate of response, underlying the interest of the scientific community in this topic. There is a wide heterogeneity in the experts’ response for each disease. At the end of the Delphi Survey rounds, we will obtain different set of clinical criteria and we will verify their performance in comparison to already existing criteria in a cohort of patients with inherited periodic fever and PFAPA enrolled in the Eurolever Registry. The final step will be a Consensus among experts (geneticists and clinicians) in order to define the best combination of clinical and genetic data for the definitive classification of patients with inherited periodic fevers and PFAPA.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2015-eular.3694

SAT0505 | SEVERE VITAMIN D DEFICIENCY IN PATIENTS WITH KAWASAKI DISEASE: ITS POSSIBLE ROLE IN THE RISK TO DEVELOP CORONARY ARTERY DAMAGE

F. Falcini1, S. Stagi2, G. Lepri1, E. Casalini3, D. Rigante3, M. Matucci-Cerinic1,2

1Department of Internal Medicine, Section of Rheumatology, Transition Clinic; 2Health Sciences Department, Anna Meyer Children’s University Hospital, University of Florence, Florence; 3Institute of Pediatrics, Università Cattolica del Sacro Cuore, Roma, Italy

Background: Kawasaki disease (KD) is an acute febrile systemic vasculitis childhood that is associated with inflammatory cytokines, in which the vascular inflammation results in damage to the coronary arteries. Vitamin D plays an important role in the regulation of immunity, and several studies demonstrate that its active form 25(OH)D exhibits anti-inflammatory activities and modulate the inflammatory response in the systemic vasculitis.

Objectives: 1. To assess serum levels of 25(OH)D in children with KD, and 2. to evaluate the possible relationship between 25(OH)D deficiency and coronary artery damage in these patients.

Methods: We evaluated 25(OH)D serum levels in 60 patients with KD (18 females, 42 males, mean age at KD diagnosis 23.8±15.4 years). These patients were compared with a sex- and age-matched control group. In all patients 25(OH)D serum levels were measured in the acute phase of the illness before IVIG treatment. All patients received timely the current treatment for KD, IVIG 2 g/kg and aspirin (30-50 mg/kg). Among the 60 pts 3 developed coronary artery aneurysms (diameter >5 mm <8 mm) and 2 coronary ectasia.

Results: KD patients showed significantly reduced 25(OH)D levels (8.9±2.24 mg/dl) in comparison to controls (21.70±4.35 mg/dl; p<0.0001). We did not detect significant difference among KD patients with less and more than 1 year of age (9.05±2.31 mg/dl vs 9.20±2.61 mg/dl). On the contrary, we observed significant differences in KD patients both with and without coronary aneurysms (7.48±2.33 mg/dl vs 9.90±2.81 mg/dl; p<0.05).

Conclusions: 25(OH)D level might have a role in the development of coronary damage. Yet, low 25(OH)D levels might contribute to the chronic course and severity of coronary aneurysms in the KD. Future larger studies are needed to explore the relationship between serum 25(OH)D levels and KD cardiac damage.

References:
[1] Kudo K, Hasegawa S, Suzuki Y, Hirano R, Wakiguchi H, Kittaka S, Ishiyama T,1,2,25-Dihydroxyvitamin D(3) inhibits vascular cellular adhesion molecule-1 expression and interleukin-8 production in human coronary arterial endothelial cells. J Steroid Biochem Mol Biol 2012;132:290-4.
[2] Suzuki Y, Ichiyama T, Ohsaki A, Hasegawa S, Shiraiishi M, Furukawa S. Anti-inflammatory effect of 1alphap,25-dihydroxyvitamin D(3) in human coronary arterial endothelial cells: Implication for the treatment of Kawasaki disease. J Steroid Biochem Mol Biol. 2009;113:134-8.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2015-eular.5784

SAT0506 | TOWARDS THE DEVELOPMENT OF AN ULTRASOUND COMPOSITE DISEASE ACTIVITY SCORE FOR JUVENILE IDIOPATHIC ARTHRITIS

I. Borzani1, G. Di Landro1, A. Ravelli2, F. Corona1, G. Filocamo1, Fondazione IRCCS Ca’ Granda Ospedale Maggiore Policlinico, Milano; 2IRCCS G Gaslini and University of Genova, Genova, Italy

Background: Assessment of disease activity is a fundamental component of the clinical evaluation of children with Juvenile Idiopathic Arthritis (JIA) and the first composite disease activity score for JIA, the Juvenile Arthritis Disease Activity Score (JADAS), was reported in 2004. Ultrasound (US) is a powerful tool for the assessment of joint disease and has been shown to be more accurate than clinical examination in detecting synovitis. However, the validity of Gray Scale US (GSUS) and Power Doppler US (PDUS) techniques for evaluating inflammatory activity in affected joints remains unproved.

Objectives: To develop and to investigate the construct validity of a composite disease activity index for JIA that includes US findings of synovitis compared with cJADAS in assessing joint inflammation.

Methods: Before the study visit, parents of children with JIA were asked to complete the Juvenile Arthritis Multidimensional Assessment Report (JAMAR), which includes a standardized assessment of pain and WB on a 21-numbered circle visual analog scales (VAS), and several other parent centered JIA outcome measures. At study visit, a pediatric rheumatologist, who was unaware of parent’s reports, performed a formal joint assessment and scored the physician’s global assessment (PG) on a 11-point VAS. After the visit, the pediatric radiologist with almost 10 years of experience in US assessment in JIA, evaluated independently the presence or the absence of synovial hypertrophy/effusion (GSUS), and PDUS in elbows, wrists, metacarpophalangeal and interphalangeal joints, knees and ankles. The joint with hypertrophy/effusion at GSUS and positivity for PDUS was defined GSUS and PDUS active joints respectively. In each patient, the GSUS and the PDUS JADAS -10 were calculated as the sum of the scores of GSUS active joints and PDUS active joints to a maximum of 10 respectively, the parent WB, and the PG, which yields a global score between 0 to 30. The correlation between the number of active joints defined clinically, GSUS and PDUS active joints, the GSUS and PDUS JADAS-10 and the clinical JADAS-10 (2) was assessed using Spearman’s rank correlation. Correlations were considered high if 0.7, moderate if 0.4–0.7, and low if <0.4.

Results: The JAMAR was completed by parents of 28 unselected patients, 17 with persistent oligoarthritis, 4 with extended oligoarthritis, 6 with rheumatoid factor-negative polyarthritis, 1 with systemic arthritis; aged between 5 months and 21 years. The median (range) of cJADAS-10 (2) was 12.25 (0-26.5). The table shows the correlation between the the active joint count and the cJADAS 10 with the variables considered.

Active joint count (up to 10) cJADAS10

| cJADAS10 |
|-------|
| 0.71 |
| 0.66 |
| 0.65 |
| 0.56 |
| 0.51 |

Conclusions: The preliminary results of the construct validity of a composite disease activity index for JIA that encompasses ultrasound findings of synovitis, evidenced high correlation between the clinical JADAS 10 and US JADAS 10, both for GSUS and PDUS. The correlation between the active joint count defined clinically with the active joints evidenced by US was moderate.

References:
[1] Consolaro A et al. Development and validation of a composite disease activity score for juvenile idiopathic arthritis. Arthritis Rheum. 2009
[2] Consolaro A, et al. Arthritis Care Res 2014

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2015-eular.6101

SAT0507 | GROWTH AND WEIGHT GAIN IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS: RESULTS FROM THE REACH-OUT COHORT

J. Guzman1, T. Kerr1, L. Ward2, J. Ma2, K. Oen3, G. Boire4, B. Feldman5, R. Scuccimarri6, K. Houghton1, A. Bruns4, P. Dancey7, A. Rosenberg8, L. Tucker1 on behalf of ReACCh-Out Investigators.

1UBC, Vancouver; 2U. of Ottawa, Ottawa; 3U. of Manitoba, Winnipeg; 4U. de Sherbrooke, Sherbrooke; 5U. of Toronto, Toronto; 6McGill University, Montreal; 8Memorial University, St. John’s; 9U. of Saskatchewan, Saskatoon, Canada

Objectives: To determine 1) height, weight and BMI trajectories in children with juvenile idiopathic arthritis (JIA) in a contemporary cohort; 2) their risk of growth delay, short stature, weight gain and obesity; and 3) the impact of disease activity and corticosteroid use on growth and weight gain.

Methods: 1154 children newly diagnosed with JIA from 2005 to 2010 at 16 Centres in the Research in Arthritis in Canadian Children emphasizing Outcomes (ReACCh-Out) inception cohort were included. Height and weight measurements scheduled at 0, 6, 12, 18, 24, 36, 48 and 60 months after enrolment were used to calculate age and sex standardized Z-scores for height, weight and body mass

Conclusions: The preliminary results of the construct validity of a composite disease activity index for JIA that encompasses ultrasound findings of synovitis, evidenced high correlation between the clinical JADAS 10 and US JADAS 10, both for GSUS and PDUS. The correlation between the active joint count defined clinically with the active joints evidenced by US was moderate.
index (BMI). Locally weighted scatter plot smoothing was used to display mean Z-score trajectories. Growth delay meant a decrease of 1.0 height Z-scores or more from the child’s baseline. Short stature meant a height below the 2.5th percentile for age and sex. Weight gain meant an increase of 1.0 BMI Z-scores or more from the child’s baseline. Obesity meant a BMI above the 95th percentile for age and sex. The cumulative incidence of growth delay, short stature, weight gain and obesity was estimated with Kaplan Meier survival methods. The impact of disease activity and corticosteroid use on Z-scores was estimated with mixed effects models.

Results: Median age at diagnosis was 9.5 years (IQR 4, 13) and 64% were female. Median follow-up was 35.5 months (23, 49). Mean height Z-scores remained stable in children with enthesis related, oligoarthritis and undifferentiated JIA. They decreased markedly in systemic JIA (from +0.2 to -0.3). In other categories, they decreased mildly during the first year and recovered afterwards. Mean BMI Z-scores increased in children with systemic JIA and RF-positive polyarthritis, decreased slightly in psoriatic arthritis and remained stable in other categories. At enrolment, the prevalence of short stature was 2.5% and the prevalence of obesity was 16%. The cumulative incidences of new-onset growth delay, short stature, weight gain and obesity within 3 years of diagnosis were 8.9% (95% CI 7.1-11.0), 3.1% (2.1-4.4%), 10.4% (8.6-12.6) and 10.5% (8.6-12.9), respectively. These were higher in systemic JIA (Figure) at 24.4% (15.3-37.5), 10.5% (5.1-20.8), 28.2% (19.1-40.5) and 34.7% (23.2-49.9), respectively. Systemic corticosteroids were prescribed for 85.5% of children with systemic JIA. One mg/kg of prednisone corresponded to a mean height decrease of 0.48 Z-scores (0.29-0.68) and a mean BMI increase of 0.45 Z-scores (0.25-0.65). After adjusting for prednisone dose, an increase of 1 cm in the physicians global assessment of disease activity corresponded to a mean decrease of BMI of 0.023 Z-scores (0.006-0.039), with no effect on height (p=0.42).

Conclusions: The impact on growth and weight gain was marked in children with systemic JIA and those who received systemic corticosteroids. For most other children, growth and weight gain were similar to what is seen in the general population.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2015-eular.2400

SAT0508 PREVALENCE OF UVEITIS AND RELATED SECONDARY COMPLICATIONS IN JUVENILE IDIOPATHIC ARTHRITIS

J. Klotsche 1, K. Minden 1,2, S. Schenck 1, M. Niewerth 1, T. Hospach 3, J-P. Haas 4, R. Berendes 5, G. Ganser 6, A. Heiligenhaus 7, C. Tappeiner 7,8,9.

1 Deutsches Rheumaforschungszentrum; 2Children’s University Hospital, Charite Universitätsmedizin Berlin, Berlin; 3Pediatric Rheumatology, Olga Hospital, Stuttgart; 4Deutsches Zentrum für Kinder- und Jugendrheumatologie, Garmisch-Partenkirchen; 5Zentrum für Kinder- und Jugendmedizin, Klinik für Kinder- und Jugendrheumatologie, Sendenhorst; 6Department of Ophthalmology at St. Franziskus Hospital, Muenster, Germany; 7Department of Ophthalmology, Inselspital, University of Bern, Bern, Switzerland; 8Deutsches Rheuma Forschungszentrum, Berlin, Germany

Background: Uveitis is one of the most threatening complications in juvenile idiopathic arthritis (JIA). It occurs in approximately 10% of all JIA patients. Whether the frequency of uveitis in JIA has decreased over time as a result of the widespread use of immunosuppressive substances, is still an open question. Few data on the occurrence of JIA-associated uveitis and related secondary complications are available from population-based studies.

Objectives: To determine the change in uveitis prevalence and related secondary complications in patients with JIA between 2002 and 2013.

Methods: Data source for this study was the National Paediatric Rheumatological Database (NPRD). Uveitis onset, disease characteristics and details on treatment were provided by rheumatologists once a year. Ophthalmologists reported about uveitis characteristics such as new-onset, treatment, uveitis activity and eye complications in detail in a specific uveitis add-on module. Data from the years 2002 to 2013 were used to determine the annual prevalence of uveitis and frequency of secondary complications. Two-level random effect models were used for investigating the change between 2002 and 2013.

Results: A total of 60 centers included 18,955 JIA patients, which were recorded in the NPRD between 2002 and 2013. The mean age of the patients was 11.2±4.6 years, their mean disease duration 4.4±3.7 years. 66.9% were female and 51.7% ANA positive. Patients’ mean age at arthritis onset was 6.8±4.5 years. In a multivariable regression analysis, the following risk factors for uveitis were identified: oligoarthritis (OR=4.21, p<0.001), and ANA positivity (OR=2.81, p<0.001), a higher disease activity measured by the cJADAS-10 (OR=1.02, p<0.001), whereas higher age at JIA onset was negatively associated with the onset of uveits (OR=0.90, p<0.001). Treatment rates with conventional and biological DMARDs increased during the observation period (cDMARD: 39% to 47.2%, bDMARD: 3.3% to 21.8%). Uveitis prevalence significantly decreased from 2002 to 2013 (13.0% to 11.6%, OR=0.98, p<0.015). The prevalence of secondary uveitis complications also significantly decreased between 2002 and 2013 (33.6% to 23.9%, OR=0.94, p<0.001). Among the ophthalmological complications, the most common were synchiae (2002: 40.6%, 2013: 46.7%), cataract (2002: 40.6%; 2013: 29.3%) and band keratopathy (2002: 42.2%; 2013: 13.3%). The percentage of patients with inactive uveitis significantly increased from 30.6% in 2002 to 65.3% in 2013 (OR=1.15, p<0.001). Visual acuity of the worse eye also improved over time, in fact cJHS LN 8±20/20 was found in 36.7% of uveitis cases in 2002 compared to 8.0% in 2013.

Conclusions: Uveitis prevalence and the frequency of secondary complications significantly decreased between 2002 and 2013. Both were correlated with a more frequent use of DMARDs.

Acknowledgements: The study was supported by a grant from Pfizer Pharma GmbH Germany (Forschungsförderung Rheumatologie). The national pediatric database is financially supported by the Children’s Arthritis Foundation (Kinder-RheumaStiftung).

Disclosure of Interest: J. Klotsche: None declared, K. Minden: None declared, S. Schenck: None declared, M. Niewerth: None declared, T. Hospach Speakers bureau: Pfizer, Abbvie, J-P. Haas: None declared, R. Berendes: None declared, G. Ganser Grant/research support from: Abbott, Actelion, Pfizer, A. Heiligenhaus Grant/research support from: AbbVie, Pfizer, Novartis, and Deutsche Forschungsgemeinschaft, and has received study fees from AbVie, Alimera Sciences, Allergan, Santen, and XOMA, C. Tappeiner Grant/research support from: Swiss Foundation for Grants in Biology and Medicine (SFBGM), Swiss National Science Foundation (SNSF) and Novartis.

DOI: 10.1136/annrheumdis-2015-eular.5115

SAT0509 CLINICAL AND SEROLOGICAL PROFILE OF CHILDREN WITH POSITIVE SSA-RO/SSB-LA ANTIBODIES

J.G. Ovalle-Benítez 1,2, J.C. Nieto 1,3, J. Martínez-Barrio 1, J.F. López-Longo 1, I. Bautista 1, R. Nardeo 1, C.A. Echeverría 1, E. Naredo 1, C.M. Hinojosa 1, N. Bello 1, B. Serrano 1, C. Mata-Martinez 1, R. Gonzalez 1, C. Saenz 1, I. Montaegudo 1, D. Hernandez 1, L. Valor 1, L. Carreño 1, 2, J. Oudar 1, 2, 3, Rheumatology; 4Pediatric Rheumatology, Hospital General Universitario Gregorio Marañón, Madrid, Spain

Background: Several studies have shown the relationship between anti-SSA-Ro/SSB-La antibodies and Systemic Lupus Erythematosus (SLE), Sjögren Syndrome (SS) and other autoimmune diseases in adult population. However, the expression of these autoantibodies and clinical correlation in juvenile patients is poorly described.

Objectives: To characterize the clinical and serological profile and primary rheumatic diseases in pediatric patients with positive anti-SSA-Ro and/or anti-SSB-La antibodies.

Methods: The data was obtained from a long term prospective cohort of patients under age 18 diagnosed with rheumatic diseases in a tertiary hospital in Spain. Demographic, clinical, and laboratory data were collected from 1986 to 2010. Patients were divided into 2 groups: anti-SSA-Ro/SSB-La positive and anti-SSA-Ro/SSB-La negative.

Results: A total of 187 patients were tested for anti Extractable Nuclear Antigens (ENA), with a following mean time of 11 years. Mean age at disease onset was 12.6 years and 77% were female. Fifty-four (28.9%) anti-SSA-Ro/SSB-La positive subjects were compared against 133 (71.1%) anti-SSA-Ro/SSB-La negative subjects. Among positive cases, 13 (24.1%) patients were double-positive for anti-SSA-Ro and anti-SSB-La, 51 (94.4%) were positive for anti-SSA-Ro and 3 (5.5%) were single-positive for anti-SSB-La. The anti-SSA-Ro/SSB-La antibodies were found less frequently (p<0.003) in the overlapping syndromes, and more frequently in SLE (p=0.007). In addition rheumatoid factor (p=0.001), anti-SS-A (p<0.001) and anti-RNP (p<0.001) were frequently co-expressed with anti-SSA-Ro/SSB-La antibodies. Finally the anti-SSA-Ro/SSB-La positive group presented more hematological and skin manifestations than the negative group (p<0.05).

Conclusions: Similarly to adults, we observed a relationship between anti-SSA-