FRI0574

CLINICAL RESPONSE TO HIGH-DOSE INTRAVENOUS METHYLPREDNISOLONE IN CHILDHOOD AUTOIMMUNE UVEITIS: A RETROSPECTIVE ANALYSIS

Anja Schnabel¹, Elisabeth Unger¹, Normi Bruck¹, Annette Holl-Wieden², Henner Morbach², Reinhard Berner¹, Anna Leszczynska³, Christian Hedrich⁴.
¹Technical University of Dresden Carl Gustav Carus, Children's Hospital, Dresden, Germany; ²University of Würzburg, Children's Hospital, Würzburg, Germany; ³Technical University of Dresden Carl Gustav Carus, Department of Ophthalmology, Dresden, Germany; ⁴Institute of Translational Medicine, University of Liverpool, Department of Women's and Children's Health, Liverpool, United Kingdom

Background: Intraocular inflammation accounts for up to 10-15% of total blindness cases [1]. The incidence of uveitis in children ranges around 4.9/100.000, and a significant number of patients develop chronic courses and irreparable complications [2]. An impressive 74% of children with juvenile idiopathic arthritis (JIA)-associated uveitis are legally blind at diagnosis [3]. This underscores the importance of timely diagnosis and effective anti-inflammatory treatment.

Objectives: To evaluate the clinical response to high-dose intravenous methylprednisolone (IVMP) in children and adolescents with autoimmune uveitis.

Methods: A retrospective chart review was conducted in two tertiary referral centers in Germany (TU Dresden and University of Würzburg) to investigate treatment responses to IVMP (10-30mg/kg/day on three successive days with a total of one to five IVMP at monthly intervals) in children and adolescents (<16 years) with autoimmune uveitis diagnosed between 2003 and 2016. Clinical features of uveitis, disease activity, outcomes, and concomitant anti-inflammatory treatment were documented at treatment initiation, after 3 and 6 months.

Results: Fifty-six patients (93 affected eyes) with a median age of 7.4 (range: 2.5-16.7) years were included. In 29% of patients uveitis was associated with JIA. Uveitis was predominately located in the anterior segment (43%), bilateral (66%) and recurrent (43%). Complications occurred in 77% of patients and included visual loss, synechiae, cataract and/or retinal lesions. Patients with active uveitis received between 1 and 5 IVMP. Visual acuity improved significantly (0.52 ± 0.33 to 0.69 ± 0.30 at 3 months ($p < 0.001$), 0.78 ± 0.31 at 6 months ($p < 0.001$)) independent of the number of IVMP. Furthermore, anterior chamber cells (45% to 18%, $p = 0.01$), synechiae (47% to 32%, $p < 0.005$), keratic precipitates (27% to 18%, $p < 0.001$), papillary edema (30% to 13%, $p < 0.001$) and/or macular edema (15% to 4%, $p = 0.01$) improved 3 months after IVMP. Over all, children treated with 3 or more IVMP ($n = 27$) (as compared to 1 IVMP ($n = 18$)) experienced fewer relapses (Median 1 [0-6] vs 3 [0-13], $p = 0.186$), developed fewer cataracts (7% vs 39%, $p < 0.02$) and less frequently required treatment with biologics (19% vs 39%, $p = 0.174$).

Conclusion: High-dose IVMP induces rapid improvement in children with autoimmune uveitis. Data suggest improved outcomes in children treated with three or more courses of IVMP when compared to one course (without reaching statistical significance). Prospective randomized trials in larger cohorts are required to confirm results.

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Disclosure of Interests: Anja Schnabel: None declared, Elisabeth Unger: None declared, Normi Bruck: None declared, Annette Holl-Wieden: None declared, Henner Morbach: None declared, Reinhard Berner Grant/research support from: Local Principal Investigator in a Clinical Trial on Canakinumab for sJIA supported by Novartis (Trial Registration: NCT00886769, NCT00889863, NCT00426218 and NCT00891046) Local Principal Investigator in a Clinical Trial DAP-PEDOST-11-03 supported by Merck (Trial Registration EUDRACT 2013-000864-28), Anna Leszczynska: None declared, Christian Hedrich Grant/research support from: Novartis Pharmaceuticals for Research study on effector T cells in psoriasis, Speakers bureau: In 2016: Roche Pharmaceuticals, Rheumatologisch, Dresden, Germany; Novartis, Advisory board > Travel costs.
DOI: 10.1136/annrheumdis-2019-eular.2495

FRI0575

PREVALENCE OF OVERWEIGHT AND OBESITY IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS

Meriem Sellami, Alia Fazaa, Saousen Miladi, Kmar Ouenniche, Leila Souabni, Selma Kassab, Selma Chekili, Leith Zakraoui, Kawther Ben Abdelghani, Ahmed Laatar. *Mongi Slim Hospital, Rheumatology, Tunis, Tunisia*

Background: Children with juvenile idiopathic arthritis (JIA) may have an increased risk for overweight and obesity, which could be an additional risk factor for inflammatory arthritis.

Objectives: The aims of this study were to determine the prevalence of overweight and obesity in children and adolescents with JIA, and to assess the association between overweight and disease parameters in this population.

Methods: We assessed the weight (kg) and height (cm) according to the standard deviation score (SDS) in a cross-sectional study of JIA children. The diagnosis of JIA was based on the International League of Association of Rheumatology (ILAR) criteria. Overweight and obesity were defined by the Body Mass Index (BMI) (weight/height^2) matched on age and sex and in reference to the French curves. Children were classified as obese if their BMI was ≥ 95 th percentile, overweight if their BMI was between the 85th and 94th percentile, and healthy weight if their BMI was between the 5th and the 84th percentiles. Functional disability was determined by the Childhood Health Assessment Questionnaire (CHAQ). Disease activity was assessed using a validated score, Juvenile Arthritis Disease Activity Score 27 (JADAS- 27).

Results: Fifty-five patients (38 boys and 17 girls) with JIA were enrolled in this study. The median age was 8.5 ± 4.12 years (range 6-14). Thirty-one patients (53%) had persistent oligoarticular JIA, 15 (27%) had polyarticular JIA, 5 patients (9%) had systemic JIA, and 4 (7%) had enthesitis-related arthritis. The median disease duration was 3.2 ± 2.8 years. Twelve patients had active disease at the time of the study with a mean JADAS 27 of 6.9 ± 2.7 . The mean CHAQ was 1.4 ± 0.5 . Nineteen children (52.7%) had received corticosteroids during an average period of 1.7 years [0.6-3] with a mean of 10mg/day of prednisone or equivalent. The mean BMI was $14.56 \pm 2.1 \text{ kg/m}^2$. Twenty-two patients (40%) were overweight, 15 (27%) were obese and 18 (33%) have normal weight. Patients with normal weight, overweight and obese represented respectively 60%, 20% and 20% of systemic forms, 53%, 27% and 20% of polyarticular form, 39%, 32% and 29% of persistent oligoarticular forms, and 50%, 25% and 25% of enthesitis-related arthritis forms. Obesity was more frequent in older patients ($p = 0.021$), with significant functional impairment ($p < 0.001$) and with active disease ($p < 0.001$). Only systemic JIA was more likely to be associated with overweight ($p = 0.031$) and obesity ($p = 0.024$). There was no relationships with other subtypes of JIA ($p = 0.428$) or with corticosteroid treatment ($p = 0.636$).

Conclusion: In our study, more than 60% of patients were overweight. Severe functional limitation, systemic JIA, and active disease were the most correlated parameters with obesity. Better management of the activity and functional status of the disease may reduce overweight in children with JIA.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.4922

FRI0576

PULSE WAVE VELOCITY EVALUATING ARTERIAL STIFFNESS IN FAMILIAL MEDITERRANEAN FEVER

Vassiliki Sgouropoulou, Stella Stabouli, Maria Trachana. *Aristotle University of Thessaloniki, 1st Department of Paediatrics, Hippokraton Hospital, Thessaloniki, Greece*

Background: Chronic inflammation leads to atherosclerosis, resulting in increased arterial stiffness. Familial Mediterranean Fever (FMF) patients, experiencing recurrent inflammatory process during the attacks, are at increased risk for atherosclerosis and cardiovascular disease. Carotid-femoral Pulse Wave Velocity (cf-PWV) is considered the gold standard for arterial stiffness evaluation. There are few studies investigating arterial stiffness with the use of PWV only in adult patients with FMF, but none in children.

Objectives: In the present study we aimed to investigate indices of arterial stiffness, cf-PWV and aortic augmentation index (AIx75), measured with the SphygmoCor XCEL device, in patient with FMF -mostly of childhood- and their associations with disease-related factors and colchicine treatment.

Methods: A total of 85 individuals were enrolled in the study, 43 FMF patients, including 30 children, in attack free period, attending the outpatient clinic of a paediatric rheumatology referral center in Northern

Greece, between September 2015 and June 2018 and 42 healthy controls.

Results: Patients with FMF presented similar systolic Blood Pressure (sBP), central Systolic Pressure (cSP) and cf-PWV values to controls, but significantly higher Alx75 values (patients vs. controls, 19.76% and 9.96%, $p < 0.05$). Only one FMF patient had cf-PWV $> 95^{\text{th}}$ percentile. Statistical analysis in FMF patients showed that Alx75 adjusted for age and sex associated with complete response compared to partial response to treatment ($B = -17.78$ 95% CI -31.17 – -4.40 , $P < 0.05$) and the presence of M694V.M680I genotype ($B = -16.75$ 95% CI -33.81 – -0.30 , $P = 0.05$). Cf-PWV presented an inverse relationship with colchicine treatment duration ($B = -0.003$, 95% CI -0.006 – 0.00 , $P < 0.05$). Cf-PWV values adjusted for age and sBP associated with attacks frequency (for < 2 months vs. > 2 months, $B = 0.48$ 95% CI 0.01 – 0.96 , $P < 0.05$). Addition of colchicine treatment duration to the model, attenuated the association between cf-PWV and attack frequency, supporting the protective role of colchicine.

Conclusion: The normal values of cf-PWV in FMF patients may reflect the compliance to colchicine treatment, that seems to have a protective role against vessel inflammation. However, the increased values of Alx75 need further investigation.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7954

FRI0577

ANTIPHOSPHOLIPID SYNDROME SECONDARY TO PEDIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS. EXPERIENCE IN A THIRD-LEVEL HOSPITAL IN MEXICO CITY

Erika Sifuentes, Alejandra Ramirez, M. A. Yamazaki-Nakashimada, F. Rivas-Larrauri. *Instituto Nacional de Pediatría, Mexico City, Mexico*

Background: Antiphospholipid syndrome (APS) is a multisystemic autoimmune disease that is characterized by thromboembolic events, pregnancy morbidity and other manifestations in the presence of elevated titers of antiphospholipid antibodies.(1)

APS can be either primary, occurring as an isolated clinical entity, or secondary to other diseases including infections, malignancy or autoimmune, the latter associated in particular with systemic lupus erythematosus (SLE).(1)

Patients with SLE can produce a great variety of autoantibodies, including the so called antiphospholipid antibodies (aPLs), such as lupus anticoagulant (LA); anti-cardiolipin antibodies (aCL) or anti- β_2 Glycoprotein-I antibodies (anti- β_2 GPI).(2) These aPLs have been described in 20-40% of SLE patients.

Objectives: To report frequency of patients with antiphospholipid syndrome secondary to pediatric systemic lupus erythematosus (pSLE) at National Institute of Pediatrics in Mexico City from 2005-2016. In addition, describe their clinical manifestations and laboratory features.

Methods: Retrospective study that included all pediatric systemic lupus erythematosus (pSLE) patients diagnosed at National Institute of Pediatrics in Mexico City from 2005 to 2016. We then identified patients with positive antiphospholipid antibodies (aPLs) and/or clinical manifestation of antiphospholipid syndrome (APS). Demographic, clinical and laboratory features were extracted from their clinical records. Study approved by the local Ethics Committee.

Results: Over the 12-year study period, we collected 295 patients with a new diagnosis of pediatric systemic lupus erythematosus (pSLE). Eighty patients (27.11%) had at least a positive antiphospholipid antibody (aPL) or a clinical manifestation of antiphospholipid syndrome (APS). Figure 1 shows our study population. Of these 80 patients, 75 (93.75%) had a positive aPL at moment of pSLE. With respect to the remaining 5 cases, 3 of them developed a positive aPL during follow-up, while the remaining

2 cases presented a clinical feature of APS simultaneously to pSLE diagnosis and their serology persisted negative during follow-up.

Twenty (25%) patients had a clinical manifestation associated to APS. Concerning these patients, 11 (55%) patients presented clinical features of APS at time of pSLE diagnosis, 6 (30%) patients developed it during follow-up and the remaining 3 (15%) patients had previous history of a clinical manifestation associated to APS prior to pSLE diagnosis. Nevertheless, 2 of these 20 patients never reported positive antiphospholipid antibodies.

According to clinical manifestations 11 (55%) patients had venous thrombosis, 7 (35%) patients with arterial thrombosis, one patient identified with chorea and one showed evidence of thrombosis in a skin biopsy.

Characteristics of the patients are shown in Table 1.

Conclusion: Antiphospholipid syndrome (APS) secondary to pediatric systemic lupus erythematosus (pSLE) is a very common entity and is increasingly diagnosed. This study allowed us to report frequency and describe clinical manifestations of pSLE patients presenting with secondary APS from Mexico City.

Figure 1. Patients with pediatric systemic lupus erythematosus at National Institute of Pediatrics from 2005-2016.

Table 1. Characteristics of patients with clinical features of APS secondary to pSLE.

Case	Sex	Age at pSLE diagnosis	Clinical manifestation of APS according to pSLE diagnosis	APS clinical manifestation	Anti- β_2 GPI-IgG/IgM	Anti-aCL-IgG/IgM	LA
1	F	12 y 4 mo	Follow-up	Venous thrombosis of LL	+/+	+/+	+
2	F	5 y 9 mo	At diagnosis	Skin biopsy with thrombosis	-/-	+/+	-
3	F	9 y 10 mo	Follow-up	Venous thrombosis of LL	-/-	-/-	+
4	F	10 y 0 mo	Follow-up	Stroke	-/-	-/-	-
5	F	8 y 2 mo	At diagnosis	Stroke	-/-	-/-	+
6	F	12 y 5 mo	At diagnosis	Venous thrombosis of LL	+/+	+/+	+
7	M	14 y 11 mo	At diagnosis	Splenic vein thrombosis	-/-	-/-	-
8	F	5 y 6 mo	Previous history	Stroke	-/-	-/-	-
9	F	7 y 10 mo	Follow-up	Chorea	+/+	+/+	+
10	F	13 y 5 mo	At diagnosis	Digital ischemia	+/+	+/+	+
11	F	14 y 2 mo	At diagnosis	Venous thrombosis of LL	ND	+/ND	+
12	F	13 y 5 mo	Follow-up	Pulmonary thromboembolism	-/-	+/+	+
13	F	16 y 5 mo	At diagnosis	Venous thrombosis of LL	+/+	+/+	+
14	M	10 y 6 mo	At diagnosis	Digital ischemia	+/+	+/+	+
15	F	14 y 9 mo	Follow-up	Venous thrombosis of UL	-/-	-/-	+
16	F	15 y 8 mo	Previous history	Arterial pancreatic thrombosis	-/-	-/-	-
17	F	13 y 7 mo	Previous history	Stroke	+/+	-/-	ND
18	F	13 y 9 mo	At diagnosis	Stroke	ND	-/-	ND
19	M	16 y 2 mo	At diagnosis	Venous thrombosis of LL	-/-	-/-	-/-
20	F	15 y 1 mo	At diagnosis	Venous thrombosis of LL	-/-	-/-	-/-

Anti-aCL: anticardiolipin antibodies; Anti- β_2 GPI: anti- β_2 glycoprotein-I antibodies; F: female; LL: lupus anticoagulant; LL: lower limb; UL: upper limb; ND: no data; y: years

Antiphospholipid antibodies (aPLs) were positive in 27% of cases with pSLE. And more than 90% of cases had positive aPLs at moment of pSLE diagnosis, which is why we strongly agree with international recommendations of performing aPLs screening simultaneously to pSLE diagnosis.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.8239