AB0960 GROWTH AND SEXUAL MATURATION IN GIRLS WITH JUVENILE IDIOPATHIC ARTHRITIS

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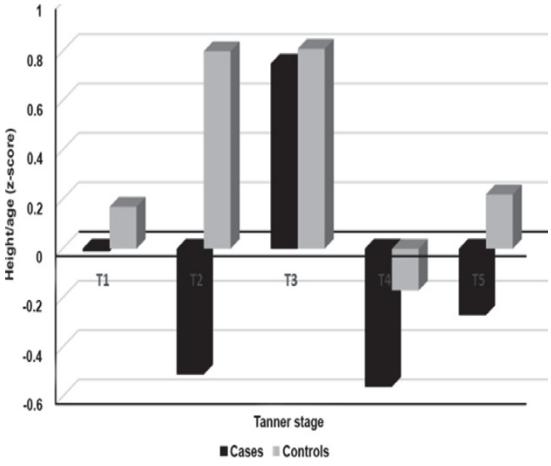
Background: Juvenile idiopathic arthritis (JIA) is a heterogeneous group of diseases with onset before the age of 16 years and joint inflammation as a main feature. Longitudinal growth is one of the main physical changes in childhood and adolescence. The etiology of delayed growth in children with JIA is multifactorial and strongly associated with prolonged inflammatory activity.
Objectives: To evaluate growth, sexual maturation, and the difference between final and expected height in girls with JIA and no glucocorticoid treatment for at least six months, as compared to a group of healthy girls.
Methods: This cross-sectional study involved 44 girls with JIA, diagnosed according to International League of Associations for Rheumatology (ILAR) criteria, and 59 healthy controls, aged between eight and 18 (incomplete) years with no comorbid chronic diseases. Demographic data were collected from all participants, and disease and treatment variables were compiled for the patient group. Anthropometric measurements were converted into z-scores based on WHO standards. Sexual maturation was classified according to Tanner stages.
Results: BMI and height z-scores were lower in girls with JIA as compared to control participants. These values differed significantly in Tanner stage II. Three (6.8%) girls with JIA had height-for-age z-scores <-2 (short stature). Girls with polyarticular JIA and higher cumulative glucocorticoid doses were significantly more likely to present with short stature. The percentage of prepubertal girls in the JIA group was significantly higher than that observed in the control group, (p=0.012). Age of menarche, adult height, and the difference between actual and expected height did not differ between groups.

Table 1. Comparison of pre- and postmenarcheal growth parameters between groups

Variables	Patients (n=44) Mean ± SD	Control participants (n=59) Mean ± SD	*p*
Menarche – n (%)	17 (38.6)	37 (62.7)	0.026
Age of menarche (years)	12.2±1.51	11.5±1.24	0,066
Menarche >2 years – n (%)	13/17 (76.5)	22/37 (59.5)	0.363
Δ Target height (father/mother)	-3.15±7.87 (n=8)	1.31±5.44 (n=16)	0.117
Nutritional data			
BMI z-score	-0.10±1.29	0.92±1.19	0.007
Height/age z-score	-0.14±1.24	0.27±1.17	0.253
Bone age z-score	-1.53±4.29	-1.42±2.17	0.928



Conclusions: These findings suggest that even six months after the suspension of glucocorticoid treatment, children with more severe forms of JIA and exposure to higher doses of glucocorticoids are still susceptible to growth impairment and delayed puberty.

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AB0961 A COHORT OF PATIENTS WITH AUTOINFLAMATORY DISEASES FOLLOWED-UP IN A UNIT OF PAEDIATRIC AND TRANSITIONAL RHEUMATOLOGY: A DESCRIPTIVE STUDY

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Background: The autoinflammatory diseases (AD) are uncommon, most of them are presented as episodes of recurrent fever and may be accompanied by other inflammatory symptoms. This group of diseases includes polygenic entities (without a single known genetic mutation) such as Behçet's disease (BD), systemic-onset juvenile idiopathic arthritis (soJIA), Chronic recurrent multifocal osteomyelitis (CRMO) and PFAPA syndrome. On the other hand, we found the entities that present with specific monogenic mutations, such as Familial Mediterranean Fever (FMF), TNF receptor-associated periodic syndrome (TRAPS), hyper-IgD syndrome and periodic fever (HIDS), cryopyrinopathies (FCAS, MWS, CINCA), Blau's syndrome and PAPA. A group of patients who can not be classified into a specific diagnosis are clustered as recurrent fever without known genetic anomaly (RFW).
Objectives: To describe and compare the clinical features of monogenic and polygenic AD and RFW seen in a paediatric and transitional rheumatology unit of a Spanish tertiary care hospital.
Methods: We performed a retrospective study including 39 patients with AD followed-up in our center.
Results: The distribution of diagnoses was: soJIA 19 patients (48.72%), BD 5 (12.82%), PFAPA 6 (15.38%), CRMO 3 (7.69%), RFW 4 (10.26%), HIDS 1 (2.56%) and CINCA 1 (2.56%). Patients came from different regions of Spain, being 22 of them boys (56.41%) and 17 girls (43.59%). The genetic study was performed in 12 patients, being positive in 7 (17.95%). Mean age at onset of symptoms was 5±5.65 years in monogenic diseases, 7.96±4.84 years in polygenic disorders and 9.5±5.91 years RFW. Delay in diagnosis in monogenic diseases was higher than in polygenic diseases (67±69.29 months vs. 24.03±30.33 months, respectively). The clinical manifestations more frequently found were fever, followed by joint involvement, being more common in monogenic diseases than in polygenic disorders (table). Haemoglobin levels were lower in monogenic than in polygenic diseases 9.95 g/dL ± 0.63 vs. 11.69 g/dL ± 2, ESR and CRP was higher in monogenic diseases 106 mm/h ± 68.5 and 80.5 mg/L ± 84.14 vs. 56.1 mm/h ± 33.78 and 57.95 mg/L ± 57.95, unlike ferritin that was more elevated in polygenic disease 896 μg/dL ± 1788.34 than in monogenic diseases 183 μg/dL ± 195.7. During his follow up 84.62% of patients received corticosteroids, 51.8% methotrexate and 46.15% biological therapy.

	Monogenic	Polygenic	Recurrent fever
Fever	100%	81.25%	100%
Joint involvement	100%	62.5%	75%
Rash	100%	59.38%	0%
Lymphoadenopathy	50%	46.88%	25%
Splenomegaly	100%	12.5%	0%
Abdominal involvement	30%	21.88%	25%

Conclusions: soJIAs was the most frequent AD in our center. All the patients had a similar gender distribution. Delay in diagnosis was greater in monogenic diseases compared with polygenic disorders. Fever and joint involvement were the more common clinical manifestations, especially in monogenic diseases. Ferritin levels were higher in polygenic diseases, whereas CRP and ESR which were higher in monogenic diseases. During the follow-up most patients required treatment with corticosteroids and approximately half of them required biological therapy.

Disclosure of Interest: None declared
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AB0962 CLINICAL AND LABORATORY CHARACTERISTICS OF NON-BACTERIAL OSTEOMYELITIS: DATA ANALYSIS OF 91 PATIENTS

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Background: Non-bacterial Osteomyelitis (NBO) is a sterile inflammatory bone disorder of unknown etiology. It typically affects children and most commonly presents with bone pain and/or swelling.
Objectives: The aim of study is to evaluate clinical and laboratory features of non-bacterial osteomyelitis in children.
Methods: Our retrospective – prospective study was included 91 patients with NBO. A routine blood test (WBC, platelets, ESR, C-reactive protein (CRP) and hemoglobin levels), a radiological examination and a bone biopsy with evaluation bacteriological and morphological data were performed in all patients.
Results: The mean age of onset NBO was 7.3 years (2.5; 10.6). We did not reveal any gender peculiarities in our study. Family history of immune-mediated diseases is found in 5/75 (6.7%) in prospective group. Concomitant immune-mediated diseases were noted in 62/89 (68.1%). Diagnostic delay was 6.3 (2.0; 17.8)

months. The radiological examination was performed in the following ratio: X-rays - 91 (100.0%), CT - 79 (86.8%), MRI - 66 (72.5), including MRI "whole body" with 15 pts, bone scintigraphy - 54 (59.3%). Monofocal form was registered in 1/3 cases. 2/3 cases was presented as a typical multifocal process with predominant involvement femur - 37 (40.7%), bone of foot - 36 (39.6%), tibia - 33 (36.3%), spine – 29 (31.9%). The number of foci is 3.0 (1.0; 6.0). We did not revealed any significant differences in quantity of WBC, platelets, hemoglobin level, ESR, CRP). Evidence confirming NBO was a negative bone biopsy in 100.0% cases. However, morphological data were as non-specific, as granulomatous inflammation.

Conclusions: NBO is determined as a primarily chronic multifocal process without specific clinical and laboratory peculiarities, associated with immune-mediated diseases. Diagnose must be established on morphological and bacteriological data bone biopsy.

Disclosure of Interest: None declared

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AB0963 LIPID ABNORMALITIES IN CHILDREN AND ADOLESCENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Systemic lupus erythematosus is an autoimmune disease that leads to the progressive destruction of the vital organs and systems, promotes early disability, premature mortality. The main reasons for the latest adult population of patients have complications of atherosclerotic vascular lesions stuch as the myocardial infarction and the stroke. The greatest effectiveness of preventive measures of the progression of atherosclerotic process and its associated cardiovascular complications determine the maximum early to start immediately after diagnosis in childhood and adolescence.

Objectives: To evaluate the lipid spectrum of the blood of children and adolescent patients with SLE.

Methods: It was been examined 26 persons 7–18 years with systemic lupus erythematosus, mostly female (88.46%), which were determined by the concentration of total cholesterol (total cholesterol), HDL cholesterol (HDL cholesterol), low density lipoprotein cholesterol (LDL), triglycerides (TG), calculated coefficient of the atherogenicity (CA = (total cholesterol - HDL cholesterol)/HDL cholesterol). Distribution groups conducted depending on the duration of the disease: the first consisted of 10 patients (38.46%) with a term of 1–3 years of the disease, the second - 16 people (61,54%) with duration of more than 3 years. The control group consisted of 10 healthy peers.

Results: Average lipid spectrum of the blood of patients did not exceed the reference values and had no statistical differences in the patients allocated to groups. However, in comparison with the control group the children with SLE had significant differences that reflect the peculiarities of formation of their metabolic disorders. Thus, total cholesterol and TG levels in both the first and the second groups were significantly higher than the control value and accounted for (5,02±0,26) mmol /L (5,24±0,26) mmol/L vs (3,39±0,20) mmol/L ($p<0$, 05) and (1,64±0,54) mmol/L (1,45±0,19) mmol/L vs (0,72±0,08) mg /L ($p<0.05$), respectively. Similar changes had occurred with parameters of LDL cholesterol, which significantly increased among patients with disease duration of 1–3 years ((3,08±0,26) mmol/L vs (1,73±0,03) mg/dL in the control group; $p<0.05$) and reached biggest values in patients with SLE over the course of 3 years ((3,57±0,82) mmol/l, $p<0.05$). In parallel, the concentration of HDL cholesterol patients of the second group decreased ((1,38±0,06) mmol/L vs (1,51±0,08) mg/dL healthy subjects, $p<0.1$), the consequence was significant increase in their spacecraft ((2,79±0,69) conv. units. against (1,24±0,17) conv. units. in comparison group, $p<0.05$).

In addition, patients with SLE found a direct correlation of total cholesterol blood of disease activity, confirmed a direct correlation ($r = 0,632$; $p<0.05$).

Conclusions: Thus, changes of blood lipid profile in the SLE patients occur in the early stages of the disease in childhood and adolescence, have the atherogenic focus, which compounded with increasing the duration and disease activity.

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AB0964 SOME PECULARITIES OF THE COURSE OF JUVENILE IDIOPATHIC ARTHRITIS IN PATIENTS TREATED WITH TOCILIZUMAB

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Background: The introduction of biological therapy drugs contributes to changing the natural course of the disease in cases of severe JIA.

Objectives: To clarify impact of tocilizumab on the course of JIA.

Methods: Of 117 children with JIA being under observation for the last five years 35% receive biologic therapy (3 etanercept, 21 adalimumab, 11 tocilizumab) during 1–5 years. Retrospective analysis of the data from clinical, laboratory and instrumental studies in the dynamics of the TCZ treatment of patients with JIA was made.

Results: Among children treated with TCZ 63,6% were sJIA cases, 63,6% were female. Age of onset 6,1±4,7y (6m–10,5y), all had acute onset, hyperthermia, severe pain syndrome, exudative arthritis in all pJIA cases and half of sJIA cases. ESR 39,7±11,2 mm/h, CRP 53,8±16,1 mg/L, all patients had anemia, leukocytosis and were seronegative for RF and anti-CCP, 3 of pJIA patients revealed ANA (1:1200–2400), IL-6 19,78±6,7 pg/ml. Before biological therapy has begun, SJIA courses were continuously relapsing in all cases with 4,7±2,1 exacerbations per y; during the 1st y of illness 8 cases run with coxitis, 7-cervical spine involvement, 9-wrists damage. All patients received CS therapy before initiating TCZ, 45,4% of them with pulse therapy, all marked by the inability to minimize the CS dose, all received 2–4 DMARDs in high doses. 2 patients received adalimumab before TCZ treatment. Elapsed time from the onset to biological agent prescription was 5,37±5,1 years. At the start of biological therapy JADAS was 19,6±5,7, stunted growth -1,88±0,3 σ, according to densitometry, osteoporosis took place in every case (Z=-2,7±1,1). After 6 month JADAS was 1,8±1,1, ESR and CRP normalized, IL-6 rate remained high in 36% cases. After 1 year the severity of osteoporosis decreased (Z = -1,17±0,8), bone deficiency depended on duration of TCZ exposure (r = -0,72) and on the elapsed time from onset of JIA before the start of biological therapy (r = -0,84). The mean increase in height was 7.73 cm/patient-year (+1±0,8 σ). Stunted growth depended on the duration of the TCZ course (r=-0,81) and the elapsed time from the onset of the disease to the start of biological therapy (r=-0,72). After 1 y of TCZ all children had normal weight and BMI for age (19,89±1,9). After 1–5 ys of treatment JADI was evaluated 2,1±2,9 (0–7), the degree of joint damage didn't depend on the duration of the biological therapy (r=0,24) and correlated with the time elapsed from the onset of the disease before treatment (r =0,59). During the treatment, exacerbations were marked only in 1 case, adverse events in 3 cases (skin infections, leukopenia). TCZ therapy allowed to completely discontinue CS in 63,6% cases, minimize them to 4 mg/day in others, DMARDs are discontinued in 9% cases.

Conclusions: Administration of TCZ is rather effective towards the drug induced remission even in long-time JIA process, but the least joint damage, osteoporosis and stunted growth can be obtained with earlier tocilizumab prescription. Reclassification of patients according to specific clinical and immunological features leads to optimization of the selection of targeted therapy.

References:

[1] While identifying in onset of JIA polyarticular destruction and systemic features (hyperthermia, anemia, leukocytosis, high laboratory activity and severity of osteoporosis) one should consider the feasibility of early administration of tocilizumab.

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AB0965 EVALUATION OF BONE MINERAL DENSITY IN CHILDREN WITH CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS AT DIAGNOSIS

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Background: Chronic recurrent multifocal osteomyelitis (CRMO) is a rare autoinflammatory disorder that has sterile bone inflammation as a main phenotypic feature. Diagnosis of CRMO is based on typical clinical and radiologic findings and presence of some inflammatory markers. Plain radiographs, technetium bone scan and MRI may be used for diagnosis of children with suspected CRMO. Despite data for other imaging methods in chidren with CRMO, little information is available regarding the bone mineral density.

Objectives: The aim of the study was to evaluate bone mineral density (BMD) findings in children with CRMO at time of diagnosis.

Methods: The medical records of children with CRMO were reviewed retrospectively. Children who met Bristol diagnostic criteria were included in the study. Clinical and laboratory findings, bone scan and MRI features were analyzed. Bone mineral density was measured by DEXA technique at femoral neck and lumbar spine regions; a Z-score< -2 was considered as osteoporosis.

Results: A total of 6 patients, one girl and 5 boys, with a median age of 10 years (4 – 14 years) and a median follow-up period of 38.5 months (3 – 72 months) were included in the study. MRI and whole body technetium bone scan were performed at 6 and 4 children, respectively and detected at least two lesions MRI or bone scans. Osteoporosis was detected in 5 patients at diagnosis. Bone mineral density Z-scores were median -2.9 [-4.9±(-2,4)] and -2.9 [-4,1 – (-1,8)] at femoral neck and lumbar spine, respectively.

Conclusions: Osteoporosis is common in children with CRMO, it may be related with disease spectrum. Evaluation of bone mineral density abnormalities at time of diagnosis and during therapy may be a part of primary care in children with CRMO.

Disclosure of Interest: None declared

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