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POS0077

PHYSICAL ACTIVITY AND SPORT PARTICIPATION: A COMPARISON BETWEEN CHILDREN AND ADOLESCENTS WITH JUVENILE IDIOPATHIC ARTHRITIS (JIA) AND THE GERMAN GENERAL POPULATION

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Background: Physical activity (PA), including sport participation is essential for children throughout their growth and maturation. It improves physiological and psychosocial health and limits the risk of developing metabolic disorders. The beneficial effect of PA specifically in patients with JIA has also been linked to its potential regulatory effect on the balance between pro- and anti-inflammatory responses [1].

Objectives: The study aimed i) to quantify the frequency of PA and participation in (organised) sports compared to the general population, ii) to determine self-reported reasons for not practicing sports, and iii) to identify clinical parameters associated with non-participation in sports.

Methods: Data from children and adolescents with JIA recorded in the National Paediatric Rheumatological Database (NPRD) in the year 2019 were considered for the analyses. In accordance with the methodology used in the general population survey (KIGGS) [2], achievement of the WHO recommendations on PA for at least 60 minutes per day as well as sport-related data were determined on the basis of self-reported outcomes in individuals aged 3 to 17 years. In order to compare PA-related data with the general population, a sex- and age-matched

sample was drawn. A logistic regression model was used to explore the association between non-participation in sports and patients' clinical outcomes.

Results: Data of 5.333 matched-pairs (mean age 11.0 ± 4.3 years, female 67%, patients' disease duration 4.8 ± 3.8 years, persistent oligoarthritis 43%) were available for evaluation. Almost 38% of patients aged 3 to 17 years met the recommended PA amount (76% aged 3 to 6; 48% aged 7 to 10; 30% aged 11 to 13; 15% aged 14 to 17). In matched controls, 21% fulfilled the WHO recommendations on PA (41% aged 3 to 6; 23% aged 7 to 10; 17% aged 11 to 13; 10% aged 14 to 17). Largest differences across JIA categories were found in persistent oligoarthritis (43%) and enthesitis-related arthritis (22%). 64% of patients and 74% of controls reported participating in sports, of which 72% of patients and 58% of controls stating to participate in a formally organised way. In both groups, boys indicated organised sports participation more often than girls. Among those who declared not participating in sports, "no interest" (patients 27% vs. controls 29%), "no suitable offer nearby" (patients 25% vs. controls 31%), "health restrictions" (patients 22% vs. controls 4%) and "no time" (patients 15% vs. controls 23%) were the most frequently mentioned reasons (multiple responses possible). CJADAS-10 (OR = 1.02, 95% CI = 1.00-1.04), CHAQ (OR = 1.79, 95% CI = 1.50-2.14), DMARD use (OR = 1.32, 95% CI = 1.15-1.53) and disease duration (OR = 0.97, 95% CI = 0.95-0.99) were significantly associated with non-participation in sports.

Conclusion: Based on self-reported data, children and adolescents with JIA meet the WHO recommendation on PA more often than general population controls. Patients are less frequently engaged in sports, but more often involved in formally organised forms. In order to bring joyful, interesting PA opportunities in line with WHO recommendations, further components (e.g. intensity), facilitators and barriers to PA and sports need to be addressed in the future while controlling for JADAS and CHAQ.

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POS0078

CROSS-SECTIONAL ANALYSIS OF INTERFERON SIGNATURE IN PEDIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: the role of interferon pathways in the pathogenesis of systemic lupus erythematosus (SLE) has been proven over the past years. Existing data suggest that interferon score (IFN I score) may serve as a useful marker of disease activity and patient clinical characteristics.

Objectives: to compare characteristics of pediatric SLE patients with high and normal IFN I score.

Methods: 40 SLE patients (33 girls, 7 boys) under 18 years old were included in the cross-sectional study. In all cases the diagnosis was made using Systemic Lupus International Collaborating Clinics (SLICC) classification criteria. The data on clinical manifestations, disease activity by SLEDAI and ECLAM, laboratory findings in the onset of the disease and at the moment of interferon signature assessment were evaluated. Interferon signature was assessed by real-time PCR quantitation of 5 IFN I-regulated transcripts; median expression of ≥ 2 was considered as a threshold. The patients were divided into 2 groups depending on the level of interferon score: high (group1, n=31) and normal (group2, n=9).

Results: The mean age of the disease onset was 12 (9.5; 14.0) years. The most common symptoms were skin lesions (85%), arthritis (67.5%), fever (55%), mucosa (45%), CNS (37.5%) and kidney (30%) involvement. Anemia, leukopenia and thrombocytopenia were observed in 62.5%, 27.5% and 50% of cases,

while 87.5% and 70% of patients had ANA positivity and dsDNA antibodies at the onset. The comparison between the groups with increased and normal IFN I-signature is presented in Table 1.

Table 1.

Parameters	Group 1 (High IFN-s)	Group 2 (Normal IFN-s)	p-value
Girls, n (%)	25 (80.7)	8 (88.9)	0.567
The onset age, years	12.0 (10.0; 14.0)	11.0 (9.0; 13.0)	0.353
Time to IFN-signature study, months from onset	18.3 (7.0; 26.5)	0.97 (0.87; 1.73)	0.987
Skin involvement, n (%)	12 (38.7)	4 (44.4)	0.837
CNS involvement, n(%)	8 (25.8)	1 (11.1)	0.353
Arthritis, n(%)	11 (35.5)	2 (22.2)	0.455
Anemia, n(%)	9 (29.0)	2 (22.2)	0.687
Leucopenia, n(%)	9 (29.0)	1 (11.1)	0.274
ANA-positivity, n (%)	27 (87.1)	5 (55.6)	0.037
anti dsDNA antibodies, n(%)	12 (38.7)	2 (22.2)	0.361
Rheumatoid factor, n (%)	11 (35.5)	0 (0.0)	0.036
Hypocomplementemia, n (%)	18/28 (64.3)	2/6 (33.3)	0.162
Ferritin level, mkg/l	112.0 (39.0; 271.0)	21.0 (5.3; 23.7)	0.0008
Hematuria, n (%)	10 (32.3)	0 (0.0)	0.049
Proteinuria, n (%)	11 (35.5)	0 (0.0)	0.036
SELENA-SLEDAI, points	9 (2; 15)	1 (0; 4)	0.073
ECLAM, points	3.0 (1.0; 6.0)	1.0 (0.0; 1.5)	0.048
Treatment with Rituximab or Cyclophosphamide, n (%)	22 (71.0)	3 (33.3)	0.040
GCS dose 0.2 mg/kg achievement for 6 months, n (%)	9/21 (42.9)	5/6 (83.3)	0.080

Conclusion: high IFN I-signature correlated with kidney involvement, ANA and RF-positivity, ferritinemia, proteinuria and hematuria. Patients with high IFN I-signature received more aggressive treatment and needed longer glucocorticosteroid (GCS) treatment. More meticulous dynamic evaluation of IFN-signature is needed to clarify its role as a predictive and prognostic marker.

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POS0079

PATIENTS WITH JUVENILE SYSTEMIC SCLEROSIS HAVE A DISTINCT PATTERN OF ORGAN INVOLVEMENT. RESULTS FROM THE JUVENILE SYSTEMIC SCLEROSIS INCEPTION COHORT. WWW.JUVENILE-SCLERODERMA.COM

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Background: Juvenile systemic sclerosis (jSSc) is a rare disease with a prevalence of around 3 in 1,000,000 children. To better capture the clinical manifestations of jSSc the juvenile systemic sclerosis inception cohort (jSScC) has been prospectively enrolling patients with predetermined clinical variables over the past 12 years. One of the goals is to study the demographic, clinical features, and physician and patient reported outcome differences between those with juvenile limited cutaneous (lc) compared to diffuse cutaneous (dc) disease subtypes, to determine if characteristics are similar or different between dc and lc jSSc.

Objectives: Evaluation of the baseline clinical characteristics of jSSc patients in the jSScC. Compare clinical phenotype between diffuse (dcjSSc) and limited cutaneous (lcjSSc) subtypes.

Methods: Demographic, physical examination, organ system evaluation, autoantibody profile, treatment, and patient and physician reported outcome variables were evaluated from the jSScC Inception cohort and summary statistics applied using chi-square test and Mann Whitney U-test comparing lcjSSc and dcjSSc subtypes.

Results: At the time of data extraction, 175 jSSc patients were enrolled in the cohort, 81% were Caucasian and 81% female. Diffuse cutaneous jSSc subtype predominated (73%). Mean disease duration was 3.1 year (±2.7). Mean age at

Raynaud's was 10 years (+3.8) and mean age of first non-Raynaud's was 10.2 years (±3.8). Significant differences were found between dcjSSc versus lcjSSc, regarding several clinical characteristics. Patients with diffuse cutaneous subtype had significantly higher modified Rodnan skin score (p=0.001), presence of sclerodactyly (p=0.02), presence of Gottron's papules (p=0.003), presence of telangiectasia (p=0.001), history of digital tip ulceration (p=0.01), and frequency of elevated CK value (p=0.04). Cardiac involvement was significantly higher in limited cutaneous jSSc subtype (p=0.02). Diffuse cutaneous jSSc patients had significantly worse scores for Physician Global Assessment of disease activity (38 vs 25; p=0.002) and disease damage (34 vs 19; p=0.008).

Table 1. Comparison of demographic data and significant differences between dcjSSc and lcjSSc at time of inclusion

	Whole Cohort N=175	Diffuse Subtype N=128	Limited Subtype N=47	P value
Female to Male Ratio	4.3:1 (142/33)	4.1:1 (103/25)	4.8:1 (39/8)	0.829
Cutaneous subtype				
Diffuse subtype	73% (128)	128	0	
Limited subtype	27% (47)	0	47	
Mean Disease duration (years)	3.1 (± 2.7)	3.3 (± 2.9)	2.6 (± 2.2)	0.135
Mean age of onset of Raynaud's (years)	10.0 (± 3.8)	9.8 (± 3.6)	10.6 (± 4.3)	0.219
Mean age of onset of non-Raynaud's (years)	17 non-Raynaud 10.2 (± 3.9)	10 non-Raynaud 10.0 (± 3.7)	7 non-Raynaud 10.9 (± 4.3)	0.173
Disease modifying drugs	88% (154)	89% (114)	85% (40)	0.446
Cutaneous				
Mean modified Rodnan skin score	14.3 (0-51)	17.4 (0-51)	6.1 (0-24)	0.001
Gottron Papules	27% (46/171)	33% (41/124)	11% (5)	0.003
Sclerodactyly	78% (126/162)	82% (98/119)	65% (28/43)	0.020
Laboratory values				
Elevated CK	25% (30/122)	30% (26/88)	12% (4/34)	0.041
Vascular				
Telangiectasia	36% (56/154)	44% (49/111)	16% (7/43)	0.001
History of ulceration	53% (91/173)	61% (77/127)	30% (14/46)	0.001
Cardiac				
Cardiac Involvement	6% (10)	2% (3)	15% (7)	0.002
Patient Related Outcomes				
Physician global disease activity (0-100) min-max	35 (0-90) n=141	38 (0-90) n=108	25 (0-80) n=33	0.002
Physician global disease damage (0-100) min-max	31 (0-85) n=140	34 (0-85) n=108	19 (0-60) n=32	0.008

Conclusion: Results from this large international cohort of jSSc patients demonstrate significant differences between dcjSSc and lcjSSc patients. According to the general organ involvement and physician global scores, the dcjSSc patients had significantly more severe disease. These observations strengthen our previous findings of the unique organ pattern of pediatric patients.

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POS0080

TOFACITINIB TREATMENT IN CHILDREN WITH RHEUMATIC DISEASES: SINGLE-CENTER EXPERIENCE

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Background: While efficacy of tofacitinib (TOF) has been proven in many adult immune-mediated conditions, the information on its safety and efficacy in the pediatric population is limited.

Objectives: to evaluate the safety and efficacy of TOF in children with immune-mediated diseases.

Methods: from 23 children whom TOF has been initiated, 17 children with treatment duration of > 6 months were extracted including 16 girls and 1 boy with the following diagnosis: JIA (n=10), autoinflammatory diseases (AID) (n=5) and juvenile dermatomyositis (JDM) (n=2) due to impossibility to taper corticosteroids (CS) or previous biologic treatment failed. The treatment outcome was classified according to the opinion of the attending physicians as complete response (CR) i.e., the absence of disease activity, partial response (PR) – a significant