thrombus in RV and showed bilateral multiple aneurysms along the pulmonary artery and its branches. According to ICB, the patient was diagnosed with BD due to having aphthous ulcers, pseudofolliculitis, and vascular involvement. IV MP (500 mg/day) for 3 days was followed by oral prednisolone 1 mg/kg/day, which was subsequently tapered. IV cyclophosphamide at a dose of 500 mg was also given every 3 weeks for a total of 6 cycles, followed by oral azathioprine (AZA). Concomitant subcutaneous IFN- α 2a was given two times per week for 6 months. Within two weeks, cough and fever disappeared, CRP values normalized. After 1 year the pulmonary artery aneurysm disappeared and cardiac thrombosis resolved. We have been following the patient with AZA for four years without recurrence.

Conclusion: We present two pediatric patients with pulmonary involvement of BD. PAI is a life-threatening condition and should be managed with more aggressive medical therapy. Early diagnosis and aggressive immunosuppressive treatment are very important in PAI. We strengthened our treatment with IFN- α 2a. There is no data in the literature regarding the use of IFN- α 2a in PAI treatment along with low dose cyclophosphamide. There were no mortality or recurrences within the 6 and 4 years follow up period. An aggressive immunosuppressive therapy leads to better prognosis in this most dreadful complication of BD.

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SAT0494

HOME MONITORING OF INACTIVE DISEASE IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS: PREDICTIVE VALUE OF EQ-5D-5L-Y

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Background: In recent years, juvenile idiopathic arthritis (JIA) research has shifted towards treat-to-target therapy based on clinical assessments and patient-reported outcomes (1). A well-known measurement of quality of life is the EQ-5D-5L (2). Herewith, we report preliminary results of a retrospective study using the child-friendly 'EQ-5D-5L-Y' with an E-health application (Reuma2GO) to monitor children with JIA in an outpatient setting.

Objectives: To assess the relationship between dimensions of the health-related quality of life 'EQ-5D-5L-Y' questionnaire and conventional assessments for children with JIA, including the Juvenile Arthritis Multidimensional Assessment Report (JAMAR) and active joint count (AJC), and to investigate the potential of the EQ-5D-5L-Y as instrument for outpatient management.

Methods: The study was designed as monocentric retrospective cohort study. Data from October 2017 to January 2019 were available for 70 patients with JIA. The relationships between individual dimensions of the EQ-5D-5L-Y, JAMAR and several clinical assessments were investigated. Furthermore, dimensions of the EQ-5D-5L-Y were investigated as possible predictors for binary disease activity using AJC > 0 as reference standard for active disease.

Results: Seventy patients with JIA completed 115 EQ-5D-5L-Y and JAMAR questionnaires within two weeks before a clinical visit. Moderate to high correlations were found between the EQ-5D-5L-Y and JAMAR. Moreover, the best possible EQ-5D-5L-Y score, with and without health-related visual analogue scale (EQ-VAS), demonstrated high sensitivity (81.1%) and negative predictive value (84.8%) for active disease (Table 1). The few patients who were incorrectly classified as having inactive disease (false-negatives) did not have their medication changed at the clinical visit and experienced little to no impact of disease activity on their quality of life, as indicated by the JAMAR questionnaire.

Table 1. Predictive value of EQ-5D-5L-Y dimensions for active disease using AJC > 0 as reference standard.

Cut-off for 'Active Disease'	Accuracy (% CI)	Sensitivity	Specificity	PPV	NPV
EQ-5D Mobility > 1	74.8% (65.8-82.4)	59.5%	82.1%	61.1%	81.0%
EQ-5D Self-Care > 1	76.5% (67.7-83.9)	35.1%	96.2%	81.2%	75.8%
EQ-5D Usual Activities > 1	67.8% (58.5-76.2)	73.0%	65.4%	50.0%	83.6%
EQ-5D Pain/Discomfort > 1	62.6% (53.1-71.5)	62.2%	62.8%	44.2%	77.8%
EQ-5D Anxiety/Depression > 1	73.9% (64.9-81.7)	51.4%	84.6%	61.3%	78.6%
EQ-5D EQ-VAS <85	60.0% (50.4-69.0)	64.9%	57.7%	42.1%	77.6%
EQ-5D Total Score > 5	60.0% (50.4-69.0)	81.1%	50.0%	43.5%	84.8%
EQ-5D Score > 5 & EQ-VAS <85	52.2% (42.7-61.6)	83.8%	37.2%	38.8%	82.9%

PPV: positive predictive value; NPV: negative predictive value.

Conclusion: These results demonstrate the discriminatory value of the EQ-5D-5L-Y between active and inactive disease in our cohort of patients with JIA. High negative predictive value was found for the total EQ-5D-5L-Y score, with and without EQ-VAS. In conclusion, the EQ-5D-5L-Y could be a valuable instrument for monitoring children with JIA in an outpatient setting which could aid physicians with deciding whether a clinical visit is necessary or not.

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SAT0495

PHENOTYPE OF PATIENTS WITH JUVENILE DERMATOMYOSITIS ASSOCIATED WITH ANTI-FACTOR H AUTO-ANTIBODY: A SEVERITY FACTOR?

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Background: Juvenile Dermatomyositis (JDM) is a rare, autoimmune and highly heterogeneous paediatric-onset myopathy, characterized by skin and muscles inflammation. Development of vasculopathy is associated with the severe extra-muscular manifestations of JDM, and portends a poor prognosis. Impaired function of JDM vasculature includes immune complex and complement deposition.

Objectives: The aim of our study is to describe the clinical and paraclinical phenotype of patients with anti-factor H autoantibody (Ab) associated with JDM and overlap myositis.

Methods: Patients with a diagnosis of JDM or overlap myositis and presence of anti-FH auto-antibody, followed in two Parisian tertiary centers over the last twelve years, were retrospectively selected. Besides demographic data, clinical features (cutaneous lesions, severity of muscle involvement, extra-muscular manifestations), treatment options and their outcome were retrieved for each patient. Biological data and presence of myositis-specific autoantibodies were collected, muscle MRI were examined, and data from muscle biopsies were assessed using a validated score tool when available.

Results: Eleven female patients were included in the study, with a median age at diagnosis of 8.8 years [2.8 - 13.1], and a median follow-

up of 3 years [1 – 11.7]. Apart typical cutaneo-muscular presentation, seven developed calcinosis, and six presented with subcutaneous oedema (of which 5 were anti-NXP2 positive). Median CMAS was 24/52 [1 – 47], and median MMT was 52/80 [2 – 76]. Gastro-intestinal involvement was found in 10 patients (91%), 4 presented a pulmonary impairment, and 3 girls had psychiatric symptoms. Six patients had a severe form (54%), one of them led to death (after 2.8 years of evolution).

MSA were found in 9 patients (anti-NXP2 = 1, anti-fibrillarin = 2, anti-MDA5 = 1 and anti-TIF1gamma = 1). IFN-signature was positive in 9 patients. Complement exploration was normal in all patients, and median positive rate of Anti-Factor H Ab was 260 UA/L [115 – 2800]. Muscle biopsy was performed in all patients but one with a median total score severity of 20. All patients received corticosteroids, and one to six other lines of treatment to achieve remission, obtained in 8 patients, with a median time to remission of 2 years [0.5; 4.8]. Use of plasmapheresis/immunoadsorption and/or Ruxolitinib was necessary in 3 of them (27%).

Conclusion: Presence of anti-factor H auto-antibodies in JDM or overlap myositis patients seems associated with more frequent gastro-intestinal involvement, and a more severe phenotype, requiring more aggressive treatment.

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SAT0496 THE FREQUENCY OF JOINT HYPERMOBILITY IN TURKISH SCHOOLCHILDREN: EFFECTS TO PHYSICAL ACTIVITY, BALANCE, PAIN AND QUALITY OF LIFE

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Background: Recent studies have focused on the joint hypermobility (JH) to show the association with musculoskeletal pain, functional disability, motor development and psychological distress (1). In contrast, in some publications, the negative effects of JH in childhood were not observed (2).

Objectives: The aim of this study was mainly to determine the frequency of JH in Turkish schoolchildren and to investigate whether relationship between JH and pain, physical activity level and the balance. In addition, the study aimed whether JH has an impact on the quality of life.

Methods: This cross-sectional school-based study evaluated 737 children (52.5% girls) from 8 to 14 years of age, and the data were collected in 2018 in the city of Denizli, Turkey. Firstly, each of the participants was individual assessed by a clinician on the Flamingo Balance Test for stability and Beighton for the diagnosis of JH. According to Beighton, children who scored ≥ 5 were accepted as hypermobile. Secondly, all participants completed the self-reported measures for the screening of physical activity level and quality of life. The Physical Activity Questionnaire (PAQ) and the Pediatric Quality of Life Inventory (PedsQL) tests were used to determine the level of physical activity and the quality of life. Also, pain severity was quantified by the Visual Analogue Scale (VAS) that is ranging from no pain (score: 0 mm) to worst pain (score: 100 mm) in the last month.

Results: The prevalence of JH in schoolchildren was 19.7% in Turkish population (Table 1). The mean pain severity was 1.29 ± 2.024 in all children. Significant differences were found between hypermobile and non-hypermobile groups in social and school functioning ($p < 0.05$), but no significant differences were found in pain, physical activity level ($p > 0.05$) (Table 2). Beighton score was not significant correlated with pain severity, physical activity, quality of life and balance in childhood ($p = 0.515$, $p = 0.986$, $p = 0.512$, $p = 0.362$ respectively).

Conclusion: As a result, the existence of hypermobility in children had an impact on school and social functions. However, it has been observed that hypermobility does not have a negative effect on these children's pain, balance, physical activity level, and physical and emotional functions.

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Table 1. Demographic findings of 737 Turkish Schoolchildren

Age		11.47 $\pm$ 1.304 (8-14)
Sex	Female	387 (52.5%)
	Male	350 (47.5%)
Beighton Score		2.75 $\pm$ 2.191 (0-9)
BS = 5		46 (6.3%)
BS = 6		53 (7.2%)
BS = 7		24 (3.3%)
BS = 8		15 (2.0%)
BS = 9		7 (1.0%)
Hypermobility	Positive	145 (19.7%)
	Negative	592 (80.2%)
Pain severity		1.29 $\pm$ 2.024 (0-10)

Table 2. Comparison of the results of children with and without hypermobility

	With Hypermobility (n= 145)	Without Hypermobility (n=592)	P
Age (year)	11,13 $\pm$ 1,35	11,55 $\pm$ 1,28	0.000
Weight (kg)	42,42 $\pm$ 11,85	45,58 $\pm$ 12,3	0.004
Height (cm)	148,8 $\pm$ 10,7	151,76 $\pm$ 10,19	0.002
Beighton Score	6,2 $\pm$ 1,14	1,9 $\pm$ 1,42	0.000
Pain severity (During Motion)	1,04 $\pm$ 1,8	1,35 $\pm$ 2,07	0.148
Pain severity (During Rest)	0,91 $\pm$ 1,86	1,16 $\pm$ 1,9	0.063
Balance	28,13 $\pm$ 9,44	30,04 $\pm$ 9,45	0.062
PAQ Score	27,13 $\pm$ 7	26,18 $\pm$ 7,4	0.102
PedsQL 4.0			
Physical functioning	85 $\pm$ 12,74	96,21 $\pm$ 331,71	0.068
Emotional functioning	77,05 $\pm$ 18,18	75,37 $\pm$ 20,04	0.533
Social functioning	91,67 $\pm$ 13,1	90,44 $\pm$ 29,14	0.049
School functioning	83,72 $\pm$ 13,71	80,32 $\pm$ 16,09	0.030

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SAT0497 CLINICAL PICTURE OF 7 PAPA PATIENTS FOLLOWED IN A SINGLE PEDIATRIC RHEUMATOLOGIC CENTER

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Background: Pyogenic sterile arthritis, pyoderma and acne (PAPA) syndrome is an autosomal dominant inflammatory disorder caused by mutations in the *PSTPIP1* gene primarily affecting joints and skin. The E250K mutation cause the hyperzincemia/hypercalprotectinemia syndrome termed *PSTPIP1*-associated-related proteinemia inflammatory (PAMI) syndrome in which a bone marrow involvement is reported

Objectives: To describe the clinical presentation of 7 PAPA patients followed at a single pediatric rheumatology center

Methods: For each patient clinical and laboratory data were collected from medical charts. *PSTPIP1* was sequenced through Sanger Sequencing or targeted resequencing using a customized panel, and analyzed with the NextSeq[®] platform (Illumina)

Results: We describe 7 patients from 4 unrelated families with the E250K mutation in a mother and 2 siblings, the A230T variant in a father and his son and the R405C and D266N respectively in the last 2 unrelated patients. Disease onset occurred within the 7th year of life in all patients. Patients 3 and 4 (table) presented since the 1st year of life recurrent episodes of fever without any cutaneous or articular symptoms. In both patients inflammatory markers were elevated during fever episodes, but persistently negative during wellbeing not requiring any therapy. The variants described in these patients were not previously reported. However their pathogenic role is supported by the detection of markedly high serum calprotectin levels (>10.000 microg/ml). The predominant feature of patients 1 and 2 was articular involvement with recurrent episodes of arthritis associated to acne. Patient 1 was initially treated