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-fibrillarine = 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 hipermobile and non-hipermobile 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. Demografic findings of 737 Turkish Schoolchildren

Age		11.47 ± 1.304 (8-14)
Sex	**Female**	387 (52.5%)
	Male	350 (47,5%)
Beighton Score		2.75 ± 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 ± 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 ± 1,35	11,55 ± 1,28	**0.000**
Weight (kg)	42,42 ± 11,85	45,58 ± 12,3	**0.004**
Height (cm)	148,8 ± 10,7	151,76 ± 10,19	**0.002**
Beighton Score	6,2 ± 1,14	1,9 ± 1,42	**0.000**
Pain severity (During Motion)	1,04 ± 1,8	1,35 ± 2,07	0.148
Pain severity (During Rest)	0,91 ± 1,86	1,16 ± 1,9	0.063
Balance	28,13 ± 9,44	30,04 ± 9,45	0.062
PAQ Score	27,13 ± 7	26,18 ± 7,4	0.102
PedsQL 4.0			
Physical functioning	85 ± 12,74	96,21 ± 331,71	0.068
Emotional functioning	77,05 ± 18,18	75,37 ± 20,04	0.533
Social functioning	91,67 ± 13,1	90,44 ± 29,14	**0.049**
School functioning	83,72 ± 13,71	80,32 ± 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 hyperzincaemia/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

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