Conclusion: This study suggests that JIA patients treated with DMARDs continued their treatment during the pandemic. In contrast, parents' reluctance was a major obstacle for returning to school. Therefore, more solidified school reopening strategies should be developed.

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POS1315

USEFULNESS OF SYNOVIAL BIOPSY IN THE DIFFERENTIAL DIAGNOSIS AND AS POSSIBLE PREDICTOR OF RESPONSE TO TREATMENT IN JUVENILE IDIOPATHIC ARTHRITIS

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Background: While synovial biopsy is an invasive procedure and is not required for the diagnosis of juvenile idiopathic arthritis (JIA), it may be useful in doubtful cases.

Objectives: Aims of the study were i.to verify the role of synovial biopsy in the differential diagnosis of JIA and ii. to review the pathology slides in order to evaluate possible associations of a histologic pattern with response to treatment.

Methods: We reviewed data from medical records of patients under the age of 18 years who underwent a synovial biopsy requested by our Pediatric Rheumatology Unit over the last 10 years. We collected information on demographic, clinical, laboratory, radiological, histopathological characteristics, as well as treatment response (in particular, remission at the last visit and number of examination, number of biologic drugs used). Among variables in the histologic score, number of layers in the synovial lining and inflammatory infiltrate (0-5) were compared to clinical status at last visit. Potential differences in variables between responders and non responders were assessed by unpaired t-test or non-parametric Mann-Whitney test, as appropriate.

Results: We identified 64 patients (40F, 24M) with a median age at disease onset of 9 years (range 1-15) and a median follow-up time of 161 months (range 8-1160). We recognized two groups of interest: patients with a known JIA diagnosis (28/64) and patients with unknown diagnosis (36/64) at the moment of synovial biopsy. In the group with known JIA, most underwent the procedure during orthopedic surgery, and in all cases the histology was consistent with JIA. Among the unknown diagnosis group, in 19 cases results were consistent with a chronic synovitis, while among the other 17 histology could lead to a diagnosis of other conditions in 6 cases (foreign body and villonodular synovitis n=2 each, sarcoidosis and osteochondromatosis n=1 each). In the remaining 11 the final diagnoses were varied (mostly genetic forms eg skeletal dysplasia, CACP, Thiemann disease).

Between the two groups we identified 46 patients with a definite JIA diagnosis. At the last follow-up visit 29 of them were in clinical remission, albeit on medication. The remaining 17 had a severe course of disease, with persistent activity and use of at least two biologic drugs. In 26 cases we could evaluate the correlation between status at last visit and number of layers/inflammatory infiltrate, but no statistical significant correlation was found.

Conclusion: Despite its limited use nowadays, synovial biopsy may still be a useful tool in patients whose diagnosis is unclear. In our study, while it confirmed the suspicion in most cases, in other instances it allowed the diagnosis of rare

conditions that would have been otherwise missed. No association between disease course and histological features in a small JIA cohort was found. We are currently expanding the study with a larger series.

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POS1316

VISUAL FUNCTION AND QUALITY OF LIFE: PRELIMINARY RESULTS FROM A PIVOTAL CROSS-SECTIONAL STUDY ON ONE HUNDRED PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS-ASSOCIATED AND IDIOPATHIC UVEITIS

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Background: Juvenile idiopathic arthritis (JIA) is the main cause of chronic uveitis in childhood and JIA associated uveitis (JIA-U) is the most common extraarticular complication of JIA. Despite continuous improvement in its management, pediatric uveitis still represents a serious condition with potential sight-threatening complications and a significant impact on quality of life (QoL).

Objectives: To evaluate visual function (VF) and QoL in children with JIA-U and idiopathic uveitis

Methods: A cross-sectional study was conducted in two tertiary Pediatric Rheumatology Centres, enrolling all patients seen with JIA-U, JIA without uveitis and idiopathic uveitis. VF was assessed by a translated form of the available EYE-Q, adapted for cross-cultural feasibility into a 10-question tool, while QoL was evaluated by the Italian version of the Pediatric Rheumatology Quality of Life scale part of the Juvenile Arthritis Multidimensional Report (JAMAR), shortened for feasibility to a 8-question tool. JAMAR section on treatment compliance and school attendance was also included. Parents, and patients when appropriate, were asked to complete each patient/parent-reporting outcome measure, answering on a 4-point Likert scale, with a total score ranging from 0 to 72 (worst condition). Medical charts were reviewed regarding JIA and uveitis features and outcome. Quantitative and qualitative variables were compared by means of Mann-Whitney U test or chi-square/Fisher exact test, as appropriate; correlations among quantitative non-parametric variables were evaluated by Spearman's test.

Results: We herein describe results from the first 100 patients enrolled (76% female), with a median age at study time of 12.8 (9.0-17.6) years. Forty-nine had JIA-U, 37 JIA without uveitis and 14 idiopathic uveitis. Uveitis was active in 14/63 patients (22.2%), with a median of uveitis duration of 9.0 years (3.6-14.8). Almost all children with uveitis were on systemic treatment (58/63, 92%) at the time of interview; 54.0% of patients presented an ocular damage, with 8.0% having a best corrected visual acuity (BCVA) < 4/10. Total score, VF and QoL scores resulted significantly higher in JIA-U patients compared to JIA without uveitis, while no differences were noticed among children with uveitis with or without JIA (Table 1). School absence was reported more frequently in JIA-U compared to JIA only (32.7% vs 10.8%, p 0.0211). VF was significantly worse in patients with ocular damage and BCVA < 4/10 (p 0.0351 and 0.0123, respectively). In patients with uveitis, VF and QoL showed a significant correlation (r 0.50, p < 0.0001) especially in patients with idiopathic uveitis (r 0.74, p < 0.0001).

Conclusion: Visual function is a crucial component of QoL in children with uveitis and it correlates with ocular damage. Since eye involvement significantly affect QoL in patients with JIA, a specific tool widely validated and cross-cultural adapted is highly demanded in the clinical care of JIA-U patients.

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Table 1.

	JIA-U n = 49	Idiopathic uveitis n = 14	JIA n = 37	p-value*	p-value [#]
Total score	5.0 (3.0-11.0)	4.5(2-9.8)	2.0(0-4.0)	0.5739	<0.0001
VF score	2.0 (0-3.0)	1.0(0-3.0)	0(0-0)	0.9098	<0.0001
QoL score	3.0(2.0-6.0)	3.5(2.0-4.8)	2.0(0-4.0)	0.5611	0.0005

Numbers are medians (IQR). * JIA-U vs idiopathic uveitis; [#]JIA-U vs JIA

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POS1317

PREDICTIVE FACTORS FOR RESPONSE TO TREATMENT IN A LONG-TERM COHORT OF PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS-ASSOCIATED UVEITIS

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Background: Uveitis is the main extraarticular complication of juvenile idiopathic arthritis (JIA) with still a significant impact on JIA morbidity, despite continuous improvement in systemic treatment. Although antinuclear antibody positivity and early onset of JIA have been associated with a high risk of uveitis onset, so far no clinical features have been widely recognized as predictive factors for JIA-associated uveitis (JIA-U) response to treatment.

Objectives: To investigate clinical features associated with response to systemic treatment in a long-term cohort of patients with JIA-U

Methods: Clinical records of patients with JIA-U were retrospectively reviewed with regard to clinical features, therapeutic choices and outcome. Clinical and laboratory variables were compared by means of Mann-Whitney U test or chi-square/Fisher exact test, as appropriate.

Results: Data from 164 JIA-U patients were analysed (81.7% female), with a median follow up of 12.1 years (7.1-17.3). Median age at JIA and uveitis onset was 2.6 (1.6-4.8) and 4.8 (2.9 – 7.0) years, respectively. Monotherapy with a conventional disease-modifying antirheumatic drug (DMARD) was used in 25.0% of patients, while 111 patients (67.7%) received at least one biologic DMARD (bDMARDs). Compared to patients responsive to DMARDs, children requiring a bDMARDs for uveitis had a lower median age at both JIA (2.4 vs 4.3 years, p 0.0234) and uveitis onset (4.1 vs 6.2 years, p 0.0023). Despite no differences in ocular damage at onset and median disease duration, patients not responsive to conventional DMARDs showed a higher frequency of ocular damage at the last visit (66.2% vs 33.3%, p 0.011). Children requiring more than one bDMARD for uveitis presented a more frequent polyarticular course (87.0% vs 20.2%, p 0.0022), a longer disease duration (median follow-up: 14.2 vs 10.4 years, p 0.0397) and a higher frequency of visual loss (best corrected visual acuity < 4/10: 23.3% vs 6.3%, p 0.0069).

Conclusion: JIA-U patients with a lack of response to conventional DMARDs were significantly younger both at JIA and uveitis onset. Severe JIA-U requiring more than one bDMARDs was associated with polyarticular JIA course and longer disease duration. Children resistant to conventional treatment need prompt recognition and additional strategies to improve long-term outcome.

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POS1318

A NATIONWIDE COHORT STUDY ON CLINICAL AND LABORATORY MANIFESTATIONS IN CHILDREN WITH MULTISYSTEM INFLAMMATORY SYNDROME (MIS-C)

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Background: Multisystem inflammatory syndrome in children (MIS-C) was recognized during the 2020 pandemic of SARS-CoV-2. Because of the relative rarity current knowledge is limited, especially in the European Caucasian population.

Objectives: To report the epidemiology, clinical and laboratory characteristics of patients with MIS-C in a nationwide cohort study in Slovenia.

Methods: This is a nationwide prospective cohort study of all consecutive patients with MIS-C, admitted from the beginning of epidemics to 31st December 2020 to University Medical Centre Ljubljana, Slovenia, the only tertiary care pediatric rheumatology center in the country. The inclusion criteria were meeting the CDC criteria for MIS-C. Infection with SARS-CoV-2 was confirmed in all patients by positive antibodies for SARS-CoV-2. Data were collected from the patients' medical records. Data on the COVID-19 epidemics in Slovenia were collected from National Institute of Public Health. Population data were provided by the Statistical Office of the Republic of Slovenia.

Results: Twenty-three patients with MIS-C were diagnosed nationwide in Slovenia, all of them in the second wave of epidemics from 14th September to 31st December 2020. All patients were Caucasian and the estimated prevalence of MIS-C was 5.8/100 000 persons younger than 19 years of age. Detailed analyses were available in 20 patients of which 14 were boys (70 %), median age was 12.4 years (4 months to 17.7 years). Two patients (10 %) were treated in the intensive care unit and none of the patients died. Troponin was elevated in 15/20 (75 %) patients during the disease course, and 7/15 (47 %) of these had normal troponin level at admission. The serum level of troponin closely followed the serum level of CRP. Six out of 20 (30 %) patients had elevated pancreatic enzymes in the second week of the disease after treatment was already given, and one patient developed asymptomatic acute pancreatitis with serum lipase level reaching the maximum of 25µkat/L. All patients had elevated levels of D-dimer with no signs of thrombosis. Five patients (5/20; 25 %) had pleural effusions and five patients (5/20, 25 %) had ascites. Half of the patients (10/20; 50 %) had hepatosplenomegaly and eight (8/20; 40 %) had mesenteric lymphadenopathy. Three patients (3/20; 15 %) had radiologic signs of cholecystitis. Two patients had thickened lung parenchyma. All patients received IVIG and systemic glucocorticosteroids. Because of resistant or organ threatening disease 4 patients (4/20, 20%) received high dose methylprednisolone pulse therapy. Biologic therapy with anakinra was started in 2 patients. Nineteen patients (19/20, 95%) received acetylsalicylic acid and prophylactic anticoagulation was prescribed in 15/20 (75%) of patients.

The mean follow up was 50 days (14 – 122). At the last follow-up visit all patients had normal laboratory parameters of inflammation, troponin, pro-BNP, d-dimer values and normal heart function.

Table 1. Clinical characteristics.

	n (%)
Fever n (%)	20 (100)
Fever duration (days) [min; max]	5.9 [4; 8]
Headache	8 (40)
Lymphadenopathy	15 (75)
Chest pain	6 (30)
Tachycardia	16 (80)
Gastrointestinal involvement	19 (95)
• Abdominal pain	16 (80)
• Vomiting	11 (55)
• Diarrhoea	9 (45)
• Loss of appetite	18 (90)
Cough	7 (35)
Skin and mucous involvement	14 (70)
• Rash	12 (60)
• Palmar/plantar oedema	3 (15)
• Lip and mouth changes	12 (60)
• Bilateral conjunctivitis	14 (70)
Myocarditis	19 (95)

Conclusion. A very high incidence of MIS-C, estimated 5.8/100 000 persons under the age of 19 with a predominantly cardiac involvement but very good outcome was noted in European Caucasian population in a nationwide cohort study in Slovenia. Attention to newly described pancreatic involvement should be raised.