Table 1. Clinical manifestations

	Hb	Nt	Pla	Crea	Brb	AST	ALT	Fe	Tg	Fb	INR
sJIA	10.1	2.736	116930	0.85	2.77	141.8	257.9	9508.7	297.7	237.9	1.24
SLE	7.4	1.403	59983	1.53	9.77	274.3	133.2	5736	433	192.3	2.06

Table 2. Analytical measures. Hb=haemoglobin(g/dl), Nt=neutrophils ($10^3/\text{mm}^2$), Pla=platelets ($10^3/\text{mm}^2$), Cr=creatinine (mg/dl), Brb=bilirubin (mg/dl), AST (UI/L), ALT (UI/L), Fe=ferritin (ng/ml), Tg=triglycerides (mg/dl), Fb=fibrinogen (mg/dl), INR (International Normalized Ratio).

	Hypotension	Fever	Rash	Adenopathy	Splenomegaly	Hepatomegaly	Haemorrhage
sJIA	1 (10%)	7 (70%)	7 (70%)	3 (30%)	4 (40%)	1 (10%)	0
SLE	0	4 (66.7%)	1 (16.7%)	3 (50%)	5 (83.3%)	4 (66.7%)	3 (50%)

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AB1010 MACROPHAGE ACTIVATION SYNDROME: A CASE SERIES OF 16 PATIENTS

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Background: Macrophage activation syndrome (MAS) is a severe complication of several rheumatologic diseases, being of special relevance systemic lupus erythematosus (SLE) and systemic juvenile idiopathic arthritis (sJIA). Its characterized by an excessive activation of the immune system due to various mechanisms, including hyperactivation of macrophages and a failure in downregulation activity by NK and cytotoxic lymphocytes. There are various criteria for its diagnosis, highlighting secondary lymphohistiocytosis syndrome (HLH) criteria from 2004 and provisional secondary MAS criteria for JIA proposed by Ravelli in 2016¹.

Objectives: To describe a case series of patients with MAS.

Methods: This is a retrospective case series of 16 patients with MAS secondary to systemic autoimmune diseases diagnosed in Ramón y Cajal University Hospital between April 2009 and September 2018.

Results: The baseline pathology was sJIA in 8 patients (2 cases with 2 episodes) and SLE in the other 6 patients. Mean age at diagnosis was 17.44 years for sJIA and 37.5 years for SLE. Mean time from diagnosis of the baseline disease to MAS episode was 11.31 years, with 3 cases being the initial manifestation of their systemic disease. 43.8% of patients were treated with corticosteroids previously to MAS episode, and 50% were being treated with DMARDS/biologic agents (SLE: 3 patients with hydroxychloroquine and 1 patient with mycophenolate and hydroxychloroquine; sJIA: 2 patients with Anakinra, 1 patient with tocilizumab and 1 patient with etanercept). Clinical and analytical characteristics of the patients are presented in table 1 and table 2, respectively. In SLE group, only 2 patients (33.3%) had high anti-DNA titers during the MAS episode, 5 patients (83.3%) had increased C3 consumption and 4 patients (66.6%) had increased C4 consumption. As severe manifestations, 4 SLE patients presented neurologic abnormalities and 3 patients presented external hemorrhage. Infection was confirmed as a trigger in 3 patients with SLE (50%) and 4 patients with JIA (40%). Prednisone at high doses was prescribed to all patients, cyclosporine in 4 patients (66.6%) with SLE and 9 patients (90%) with sJIA. Additionally, anakinra was prescribed in 4 patients (40%) with sJIA. 4 patients (66.6%) with SLE met secondary HLH criteria and 9 patients (90%) with sJIA met secondary MAS criteria. Bone marrow biopsy was performed in all patients with SLE and in 9 patients with sJIA, demonstrating hemophagocytosis in 5 patients (83.3%) with LES and 5 (50%) patients with sJIA. Only 2 patients in the SLE group died because of MAS.

Abstract AB1010 Table 2. Analytical measures. Hb=haemoglobin(g/dl), Nt=neutrophils ($10^3/\text{mm}^2$), Pla=platelets ($10^3/\text{mm}^2$), Cr=creatinine (mg/dl), Brb=bilirubin (mg/dl), AST (UI/L), ALT (UI/L), Fe=ferritin (ng/ml), Tg=triglycerides (mg/dl), Fb=fibrinogen (mg/dl), INR (International Normalized Ratio).

Conclusion: In our case series rash and fever were more frequent among sJIA patients, the rest of the clinical manifestations were more common in SLE group. Analytical measures were more altered in SLE group except for ferritin and ALT. Mortality was 33.33% in SLE group vs 0% in sJIA group, probably due to early diagnosis and treatment in these patients.

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AB1011 APPLICATION OF AUTOINFLAMMATORY DISEASE DAMAGE INDEX (ADDI) TO AUTOINFLAMMATORY DISEASES IN A TERTIARY REFERRAL HOSPITAL

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Background: Autoinflammatory diseases (AIDs) cause chronic systemic inflammation that can damage multiple organs. Recently, the autoinflammatory disease damage index (ADDI) has been developed and validated in the four most common monogenic AIDs, Cryopyrin-associated Periodic Syndrome (CAPS), Familial Mediterranean Fever (FMF), Mevalonate Kinase Deficiency (MKD) and Tumor Necrosis Factor Receptor-associated Periodic Fever Syndrome (TRAPS). The use of ADDI index could also be of great value in other AIDs.

Objectives: The aim of this study is to assess the application of ADDI in patients with the four most common monogenic diseases and other AIDs. To accomplish this objective a detailed cohort of patients with different AIDs is presented.

Methods: All patients with AIDs followed in the Pediatric Rheumatology Unit comprising the Transitional Care and specialized AIDs outpatient clinics from Hospital Universitari Vall d'Hebron were identified. A cross-sectional, descriptive study was performed applying ADDI by two pediatric rheumatologists (EM, ML). Laboratory test including C-reactive protein (CRP) mg/dl, amyloid protein (AP) mg/L, erythrocyte sedimentation rate (ESR) mm/h and protein/creatinine rate (mg/g Cr) were performed at the moment ADDI was applied. Variables related with disease duration, current treatment and accumulated corticosteroids treatment were assessed. The continuous variables are presented as mean and standard deviation (mean $\pm$ SD) and categorical variables are presented by percentages.

Results: A total of 41 patients with AIDs were included, 61% were female, with a median age of 20 ± 11.9 years at inclusion. Disease duration was 11 ± 8.2 years. AIDs included were 11 patients with FMF (26.8%), TRAPS n=4 (9.8%), MKD n=3 (7.3%), CAPS n= 2 (4.9%), Blau

syndrome $n=7$ (17.1%), SAVI syndrome $n=3$ (7.3%), CRMO $n=4$ (9.8%), PFAPA $n=2$ (4.9%), APLAID $n=1$ (2.4%), Stickler syndrome $n=1$ (2.4%), and 3 unknown AIDs with genetic test negative $n=3$ (7.3%). Current treatment is variable among patients, 6 (15.8%) are taking disease-modifying antirheumatic drugs (DMARDs), 9 (23.7%) Colchicine, 8 (21.1%) Anakinra, 13 anti-TNF therapy (34.2%), 1 (2.6%) Ruxolitinib and 1 (2.6%) Abatacept. Only 6 patients were receiving corticoids with mean prednisone dose of 7.5 mg/day. The global ADDI mean score was 2.3 ± 2.2 . Regarding the eight different items included in the item, musculoskeletal domain shown the highest score with 1.02, followed by the ocular domain with 0.42. The patient with APLAID syndrome had the highest score of 6 followed by BLAU syndrome with 4.71. FMF has the lowest score with 0.83. Laboratory test results were mean ESR 27.2 ± 26.7 mm/h, CRP 0.7 ± 1.3 mg/dl, AP 13.9 ± 18.6 mg/L. Proteinuria was present in 2 patients with mean 286.5 ± 246.1 mg/g. EM and ML applied ADDI in 5-10 minutes average.

Conclusion: ADDI is a feasible index suitable to measure damage in a single patient. Despite it was performed to the four most common AIDs it could be applied to other diseases. In our cohort the mean ADDI index was low and musculoskeletal item has the highest score. This result could be explained by the tight control of the disease and successful targeted therapy. Laboratory tests also support this finding. Nevertheless, some organ systems are not assessed like respiratory, cardiovascular or cutaneous damage, important in some syndromes. Knowing the difficulties of applying an unified index for all diseases, ADDI may be supportive in other AIDs and longitudinal cohorts.

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AB1012 HENOC-SCHÖNLEIN PURPURA AND UVEITIS, AN UNUSUAL ASSOCIATION

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Background: Henoch-Schönlein purpura (HSP) is the most common childhood vasculitis characterized by cutaneous palpable purpura predominantly located in the lower limbs, arthralgia/arthritis, renal and bowel involvement. To our knowledge only 4 cases of HSP with ocular involvement have been reported so far.

Objectives: We describe a 6-year-old female patient who presented HSP and panuveitis.

Methods: Describe a case report.

Results: A 6-year-old female, previously in good health, received the diagnosis of HSP for cutaneous palpable purpura on her lower limbs, bilateral ankle arthritis and haematuria. Two weeks later the girl was admitted to our Emergency Department due to monolateral ocular pain with red eye. At ophthalmological assessment there was evidence of anterior uveitis with and edema of the optic disc. Blood tests showed increased values of CRP (1.92 mg/dl) and ESR (69 mm/h) with normal values of complement levels and complete blood count. Urinalysis displayed mild hematuria and proteinuria. Immunoglobulin levels were slightly increased: IgG (1850 mg/dl, n.v. 540-1330 mg/dl), IgA 244 mg/dl (n.v. 50-240 mg/dl) and IgM 72 mg/dl (n. v. 50-180 mg/dl) An extensive infectious work-up for viral, bacterial and parasitic infections was negative. ANA, ANCA, ASCA, LAC, HLA B27 and B51 were all negative. Chest X ray, abdominal ultrasound and echocardiography were negative. A brain MRI with contrast revealed an iris inflammation and excluded CNS involvement. Systemic therapy was started with i.v. methylprednisolone (30mg/kg/d for three consecutive days) and continued with oral prednisone (2 mg/kg/day), with a progressive improvement of the ocular, skin, nephrological and articular involvement. Prednisone was then gradually tapered over 2 months without disease recurrence.

Conclusion: In our literature search we were able to find an association between HSP and uveitis only in two adults and in an 11-year-old child. All these patients showed cutaneous and articular involvement and 3/4 nephrological involvement. Uveitis was observed in the first week of

vasculitis onset. Although rare, the presence of ocular involvement can occur during HSP.

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AB1013 CHILDREN IN RISK OF LOW BONE MASS HAVE MORE THAN 2 RISK FACTORS

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Background: Low Bone Mass (LBM)/Infantile Osteoporosis (IOP) require an active evaluation for its diagnosis and prevention. Therefore, its incidence is unknown and could be undertreated. The systematic collection of risk factors associated with LBM/IOP could help identify the population at risk of presenting it

Objectives: To assess the prevalence and number of risk factors (RF) in the pediatric population at risk of developing LBM/IOP. Assess its influence on Bone Mineral Density

Methods: Demographic and clinical data were prospectively collected from patients from 2 to 20 years of age, who had at least one risk factor for LBM/IOP, among them: chronic diseases, treatment with immunosuppressants and/or corticosteroids and insufficient calcium intake. Calcemia, calciuria, and Vitamin D were determined in blood samples, and whole body and lumbar DXA were performed. The calcium intake, the number of previous fractures and other RF were collected

Results: Data were collected from 103 patients, with an average age of 9'8 years, 52'4% women, and 80% Caucasians. Of these, 9 were pre-schoolers (2-3 years old), 33 schoolchildren (4-9y), 55 teenagers (10-17th) and 6 young people (18-20th)

The most frequent diagnoses were: Malabsorption/Food allergies: 46.6%, JIA: 17.5%, Nephropathies: 17.8%, Hematological diseases: 6.8%, and Vasculitis and connective tissue diseases: 3.9% each

The frequency of RFs can be observed in Table 1

	%
Insufficient calcium intake in the diet	84.5
Association of a second chronic diagnosis	4.9
Hypovitaminosis D in blood (<30 nmol/L)	8.1
Sedentary lifestyle (PAQ test <2)	13.6
History of long bone or vertebral fractures	12.6
24-hour urine hypercalciuria	3.1
Proteinuria ≥ 0.20 g/L in 24-hour urine	17
Drugs with osteopening potential (non-corticosteroid immunosuppressants)	31.1
Corticosteroids at the time of inclusion	19.4
Corticosteroids prior to inclusion in the study	18.4

The average dose of current corticoids was 0.21 mg/kg/day of prednisone with a total cumulative average dose of: 7 gr, with an exposure of 1 to 144 months

4'3% of the sample had an isolated RF, 38% had 2 RF, 31% 3, 15% 4, and 12% 5 or more

8'7% of the sample presented a LBM and 4.8% met criteria for Opi for vertebral fractures, 3 of them asymptomatic and discovered by morphometry

In the multiple linear regression analysis: age, latin ethnicity, gender, and hypovitaminosis D were the main RFs related to lumbar BMD. Likewise,