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