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.

### 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). It is 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 lymphohistocytosis syndrome (HLH) criteria from 2004 and provisional secondary MAS criteria for JIA proposed by Ravelli in 2016.

### 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 SLE and 37.5 years for SLE. Mean time from diagnosis of the baseline disease to MAS episode was 11.31 years, with 3 cases of sJIA and 5 cases of SLE. Mean time from diagnosis of the baseline disease to MAS episode was 11.31 years, with 3 cases of sJIA and 5 cases of SLE. Mean time from diagnosis of the baseline disease to MAS episode was 11.31 years, with 3 cases of sJIA and 5 cases of SLE. Mean time from diagnosis of the baseline disease to MAS episode was 11.31 years, with 3 cases of sJIA and 5 cases of SLE. Mean time from diagnosis of the baseline disease to MAS episode was 11.31 years, with 3 cases of sJIA and 5 cases of SLE.

### Analysis:

### 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.

### REFERENCES


### Disclosure of Interests:
None declared


**AB1010**

### MACROPHAGE ACTIVATION SYNDROME: A CASE SERIES OF 16 PATIENTS

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### Background:
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### REFERENCES


### Disclosure of Interests:
None declared


**AB1011**

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

**Mireia Lopez Corbeto, Estefania Moreno Ruzafa. Hospital Universitari Vall d’Hebron, Rheumatology, Barcelona, Spain**

### 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 AIDs 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 Translational 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 moment ADDI was applied. Variables related with disease duration, current treatment and accumulated corticosteroids treatment were assessed.

### Continuous variables are presented as mean and standard deviation (mean ± SD) and categorical variables are presented by percentages.

### Results:
A total of 41 patients with AIDs were included, 61% were male, 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
HENOCH-SCHÖNLEIN PURPURA AND UVEITIS, AN CHILDREN IN RISK OF LOW BONE MASS HAVE MORE


REFERENCES

Disclosure of Interests: None declared

# AB1012
HENOCH-SCHÖNLEIN PURPURA AND UVEITIS, AN UNUSUAL ASSOCIATION

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Background: Heno Schonlein purpura (HSP) is the most common childhood vasculitis characterized by cutaneous palpable purpura predominantly located in the lower limbs, arthralgia/arthrosis, 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 edema of the optic disc. Blood tests showed increased values of CRP (1.92 mg/dL) and ESR (26 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. 0-240 mg/dL) and IgM 72 mg/dL (n.v. 60-240 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 inflamed periventricular white matter lesion with restricted diffusion, suggestive of an acute or subacute encephalitis. Systemic therapy was started with iv. methylprednisolone (30 mg/kg/d for 3 consecutive days) and continued with oral prednisone (2 mg/kg/day), with a progressive improvement of the ocular, skin, nephological 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 nephological involvement. Uveitis was observed in the first week of vasculitis onset. Although rare, the presence of ocular involvement can occur during HSP.

REFERENCES

Disclosure of Interests: Ilaria Maccora: None declared, Francesca Tirelli: None declared, Gabriele Simonini Grant/research support from: Abbvie, Speakers bureau: Abbvie, Teresa Giani: None declared, Rolando Cimaz: None declared

# 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. Cacemia, 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 preschoolers (2-3 years old), 33 schoolchildren (4-9y), 55 teenagers (10-17y) and 6 young adults (18-20y).

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

<table>
<thead>
<tr>
<th>RF</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insufficient calcium intake in the diet</td>
<td>84.5</td>
</tr>
<tr>
<td>Association of a second chronic diagnosis</td>
<td>4.9</td>
</tr>
<tr>
<td>Hypovitaminosis D in blood (&lt;30 nmol/L)</td>
<td>9.1</td>
</tr>
<tr>
<td>Sedentary lifestyle (PAQ test &lt;2)</td>
<td>13.6</td>
</tr>
<tr>
<td>History of long bone or vertebral fractures</td>
<td>12.6</td>
</tr>
<tr>
<td>24-hour urine hypercalciuria</td>
<td>3.1</td>
</tr>
<tr>
<td>Proteinuria &gt; 0.20 g/L in 24-hour urine</td>
<td>17</td>
</tr>
<tr>
<td>Drugs with osteopenic potential (non-corticosteroid</td>
<td>31.1</td>
</tr>
<tr>
<td>immunosuppressants)</td>
<td>31.1</td>
</tr>
<tr>
<td>Corticosteroids at the time of inclusion</td>
<td>19.4</td>
</tr>
<tr>
<td>Corticosteroids prior to inclusion in the study</td>
<td>18.4</td>
</tr>
</tbody>
</table>

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. 43% of the sample had an isolated RF, 38% had 2 RF, 31% 3, 15% 4, and 12% 5 or more.

87% 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,