Abstract AB1080 – Table 1. Demographic, clinical, electroencephalography and neuroimaging features of CAPS patients

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7	Patient 8	Patient 9
Age (years)	10	2	6	4	3	13	6	12	14
NLRP3 gene analysis result	Q703K	Negative	G569R	Q705K	Negative	T433I	Negative	T436A	A242A
Sensorineural hearing loss	No	No	Yes (Bilateral)	No	No	No	No	No	No
Uveitis or conjunctivitis	No	No	No	No	No	No	No	Yes	Yes
Papilledema	Yes	No	No	No	Yes	No	No	No	Yes
Optic atrophy	No	No	No	No	Yes	No	No	No	No
Headache	Yes	No	No	No	No	Yes	No	No	Yes
Seizure	Yes	No	Yes	No	Yes	No	No	No	Yes
Mental retardation	No	No	Yes	No	Yes	No	No	No	No
Aseptic meningitis	No	No	Yes	No	Yes	No	No	No	No
EEG	Normal	Normal	Focal epileptiform activity	Normal	Generalised fast rhythm and sharp-wave activity	Normal	Normal	Normal	Neuronal hyperexcitability in left temporal lobe
Cranial MRI	normal	normal	Diffuse parenchymal atrophy, hydrocephalus	normal	Global cortical atrophy, sequela lesions	normal	normal	normal	Normal

findings (2/9), ocular symptoms/signs (2/9), sensorineural hearing loss (1/9), optic atrophy (1/9) (table 1). MRI of the brain was abnormal in two patients.

Conclusions: Increased understanding and awareness of this rare but treatable syndrome among neurologists is essential, since the disease could manifest with neurologic manifestations such as seizure, papilledema, sensorineural deafness and aseptic meningitis. If remains untreated and recognised, this autoinflammatory syndrome could lead to significant morbidity and mortality. Hence, early treatment with anti-interleukin-1 therapy provides complete resolution of symptoms and, also prevent progression of neurologic findings when initiated in the late stage of the disease.

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AB1081 FIRST TRANSITION CLINIC OF ADOLESCENTS WITH RHEUMATIC DISEASES IN MÉXICO, A PILOT STUDY

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Background: The adolescence is a critical period when different process are made, as identity, cognitive changes, enhance of autonomy, etc. Besides that, an adolescent with rheumatic disease has to front facing the disease acceptance. Already, different Transition Programs are made around the world. In México does not exist this important project yet.

Objectives: Provide and uninterrupted, multi-disciplinary and coordinated, properly developed health system with a comprehensive and psychosocial background.

Methods: We included rheumatologist, rehabilitation specialists, psychiatrist, nutritionist, clinical psychologist, nurse and social services. Got Transition Questionnaire (GOT) in Spanish version was used to evaluated patients and parents to evaluated the knowledge in transition process. We implemented the beginning of the clinic last July. We included patients with established rheumatic diseases, older than 16 years old. We use descriptive statistics with measures of central tendency or frequencies depending on variable characteristics. We use Spearman's correlation test to evaluated concordance of answers in GOT between parents and patients.

Results: We made a systematic evaluation program of all patients. Nineteen patients had already seen at the clinic, most of them are female, the rest of clinical

variables are shown in Table 1. Even though we found a positive Spearman's coefficient (ρ), and significant difference in answers related with "Perception of capability of adult-centred health care", "Health knowledge", and "Use of health-care services knowledge", GOT results demonstrate weak of correlation between answers from patients and their parents. Correlation plots from GOT results are shown in figure 1.

Abstract AB1081 – Table 1. Clinical and demographic characteristics

Variables	n=19
Age, years, mean $\pm$ SD	18 $\pm$ 1.4
Female gender (%)	15 (79)
Rheumatic diagnosis (%)	JIA SLE Inflammatory Myopathy Scleroderma Episcleritis
BMI, median (min – max)	20.3 (15.7–41.5)
BMI based classification (%)	Underweight Normal weight Overweight Obese
Any limited joint mobility and other musculoskeletal problems (%)	18 (95)
Psychiatric assessment abnormal (%)	14 (74)

Abstract AB1081 – Figure 1. GOT Transition correlations between parents and patients.

Conclusions: In this pilot study, we shown a high prevalence of psychiatric, nutritional, and mobility problems among adolescents, adding the non-concordance of transition abilities perception in GOT answers between parents and patients. These results encourage the need of an organised, specialised, multidisciplinary, and integrated clinic in which the patient could adapt to adult care centres.

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AB1082

CHARACTERISATION OF A GROUP OF PATIENTS WITH IGG4-RELATED DISEASE: SINGLE CENTRE EXPERIENCE

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Background: Immunoglobulin G4-related disease (IgG4-RD) is a chronic systemic inflammatory condition with an unclear pathophysiology and IgG4-positive plasma cells infiltration of various organs and parts of the body. If untreated, the disease can lead to fibrosis and irreversible organ damage. IgG4-RD mostly has been described in adults, hence it is generally unknown among paediatricians.

Objectives: We conducted a retrospective analysis of clinical features and response to therapy of five patients (one female, four males, median age 13.6 years) with IgG4-related disease, treated in our Centre.

Methods: The diagnosis was confirmed by detection of lymphoplasmacytoid infiltration with >30% of cells expressing IgG4 in all, and elevated IgG4 serum concentration in 4 cases.

Results: Three patients had localised lesions (orbit, hip muscle, peripancreatic tissue, respectively), two – multi-organ disease with polylymphadenopathy, pulmonary, renal and hepatic foci, dacryoadenitis with oedema of the eyelids. Auto-immune thrombocytopenia ($70 \times 10^9/l$), neutropenia ($0.79 \times 10^9/l$) were present in one patient. Rituximab therapy was successful in 2 cases (one patient received monotherapy with rituximab, another one – Rituximab and Sirolimus). Two other patients received JAK inhibitor therapy (ruxolitinib) with good effect. No side effects were noted. One patient underwent surgery – the infiltration in the abdominal cavity was removed with positive effect without specific therapy.

Conclusions: IgG4-RD symptoms can be diverse and sometimes atypical, so dealing with this pathology requires physician's awareness. Rituximab was effective in patients with multi-organ manifestations, and JAK inhibitor (Ruxolitinib) was effective in patients with mono-focal disease. Steroids are routinely used in IgG4-RD as a first line of treatment with significant side effects. We propose that alternative drugs could be used in IgG4-RD, especially in paediatric patients to achieve fast remission with significant morbidity.

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AB1083

CLINICAL AND DENSITOMETRIC CHARACTERISTICS IN PAEDIATRIC POPULATION WITH RISK FACTORS TO DEVELOP LOW BONE MASS/OSTEOPOROSIS

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Background: Low Bone Mass (LBM)/Paediatric Osteoporosis (Ped OP) is a disorder of unknown prevalence, partly due to the lack of diagnosis, associated with the absence of clinical manifestations of the disease until patients develop complications such as fractures, deformities or pain

Objectives: To describe the clinical and densitometric characteristics of the paediatric population with risk factors to develop LBM/Ped OP

Abstract AB1083 – Table 1

Age group	N,% age group	Average calcium intake (mg/d)±SD	Recommended Daily Amount (RDA) (mg/d)	% that reach RDA
Preschooler (2–3 y)	8, 8.9%	847±271	700	50%
Schooler age (4–9 y)	29, 32.2%	671±238	1000	27.6%
Teenagers (10–17y)	47, 52.2%	660±348	1300	8.7%
Young (18–20 y)	6, 6.7%	726±156	1100	0%

Methods: We collected prospectively demographic and clinical data of patients aged 2 to 20 years, referred for bone health assessment due to the presence of a risk factors for LBM/Ped OP, including: inflammatory diseases, immunosuppressants and/or corticosteroids, malabsorptive diseases and chronic systemic

disorders. We evaluated daily calcium intake and Bone Densitometry (DXA) was performed. We did also a vertebral morphometry

Results: Data were collected from 90 patients, with an average age of 9.9 years, 53% female, 82% Caucasian. The age distribution and calcium intake by age can be seen in table 1.

There was a significant decrease in the compliance of the RDA with calcium with increasing age ($p=0.01$). No differences were found in the average daily calcium intake between the different diagnostic groups.

The most frequent diagnoses were: Malabsorption:44.4%, JIA:20%, Nephropathies:17.8%, Haematological diseases:7.8% and Vasculitis: 4.4%.

18% of the sample had had a fracture (Fx), 44% of them had more than one, being the adolescents the group of greater prevalence. 3 cases met the criteria for fragility Fx (vertebral Fx).

20% of the patients were on systemic corticosteroids, with an average dose of 5.9 mg of prednisone (or equivalent)/day, and another 20% had previously received them. The total cumulative average corticosteroid dose in both groups was: 7 grams of prednisone, with an average exposure of 37 months. 29 patients (32%) received immunosuppressive treatment, of which 20% were methotrexate (alone or in combination with biological DMARD).

Only 7% had supplements with Calcium and 14% with Vitamin D.

100% had a normal calcium, 82% a normal phosphate (rest slightly increased) and 11% were deficient in Vitamin D

13% of the sample had a LBM for their age assessed by DXA. The densitometric results can be seen in Table 2.

Abstract AB1083 – Table 2

DXA results	Max Z score	Min Z score	% with Z score≤ -2
Vertebral Z score	-3	2	13.3%
Whole body Z score	-3	1.8	9.2%
Total femoral Z score	-4.8	2.3	10.7%
Femoral neck Z score	-4.8	3	12%

Conclusions: Calcium intake in children and young with at least 1 risk factor for LBM/Ped OP is lower than recommended, especially in the groups with the highest requirements.

Up to 13% of this population have a BMO for their age and a 3'3% meets Ped OP criteria.

Larger studies are needed to help us to identify paediatric patients who are candidates for bone health screening

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AB1084

PRELIMINARY RESULTS OF THE USE OF SERUM CALPROTECTIN (MPR8/MPR14) IN CLINICAL PRACTICE IN PAEDIATRIC RHEUMATOLOGY

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Background: Serum Calprotectin is a new biomarker of clinical activity in Rheumatology, especially in Juvenile Idiopathic Arthritis (JIA)

Objectives: To assess the use of serum Calprotectin in paediatric patients with inflammatory/Rheumatic diseases in clinical practice

Methods: We retrospectively collected Demographic and Clinical data from patients of our Paediatric Rheumatology Clinic, in which serum Calprotectin levels were determined.

The determination of serum Calprotectin was carried out using the ELISA technique.

Diagnostic and Inflammatory activity data were also collected: RCP, ESR and Clinical Assessment of the patients

Results: We present 28 patients, 61% females, with an average age of 11 (3–23 years)

The diagnoses were as follows: 16 JIA (57% of the total), of which 8 were of the Oligoarticular type, 3 were Polyarticular, 3 were Arthritis related to Enthesitis, 1 was Psoriatic and 1 Systemic. Other diagnoses were: Behçet, ¹Autoinflammatory Diseases: 4 (2 ADA2 Deficit, 1 Familial Mediterranean Fever, 1 PFAPA) and 7 patients had suspected rheumatological/inflammatory diseases in study

17 patients were considered clinically inactive, 6 with inflammatory activity and 3 doubtful at the time of blood test. The mean values of Calprotectin, RCP and ESR can be seen in Table 1.