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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. Autoimmune thrombocytopenia (70×10^9/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.
Disclosure of Interest: None declared
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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
Preescholar (2–3 y)	8, 8.9%	847±271	700	50%
Scholar 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,[1] Autoinflammatory 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.