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AB1067 THE IMPACT OF ACUTE RHEUMATIC FEVER IN A PROVINCE OF CENTRAL-NORTH ITALY

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Background: Acute rheumatic fever (ARF) is a multisystem complication of group A streptococcal (GAS) infection. The incidence of ARF varies greatly in different geographic areas, with the highest incidence in Asia, Eastern Europe, and Australia (10-350 out of 100 000 per year) (1) and lowest (0.5-3 out of 100 000) in the US and Western Europe.
In 2015 the latest version of Jones Criteria has been published (2), incidence less than 2 out of 100.000 per year was established to define low-risk (LR) population. Due to the lack of nationwide epidemiologic data in Italy, it is difficult to define the correct population Jones criteria's category to use.
Objectives: To estimate the incidence of ARF in a metropolitan area of Central-North Italy and to study the clinical characteristics of the disease in a developed country.
Methods: We retrospectively analyzed the data of all patients with ARF aged 5-14 years old, diagnosed according to the classical and now LR Jones criteria (2), referred to Sant'Orsola-Malpighi Hospital of Bologna from January 2012 to December 2017 living in the province of Bologna.
Results: We identified a total of 24 patients diagnosed by ARF. In the province of Bologna, the inhabitants between 5 and 14 years of age varied from 82.967 in 2012 to 89.699 in 2017. Every year the annual incidence of ARF was above 2 out of 100.000. The highest incidence was reported in 2013 with 8 new diagnosis (incidence of 9.3 out of 100 000 per year). Carditis was present in 20 patients (83.3% of cases), chorea in 8 children (33.3%). Only 3 patients presented polyarthritis at diagnosis while 9 had polyartralgia and 3 presented with monoarthritis. In 6 patients the diagnosis followed the onset of chorea and no joint involvement was recorded. In 5 patients the diagnosis of ARF was made after the detection of classical cardiac findings without other major criteria. Cardiac involvement was characterized by mitral regurgitation in 17 cases and aortic regurgitation in 10 (8 patients with mitral aortic involvement). Five patients presented with fever ($\geq$38.5°C), 11 children had elevated inflammatory markers as erythrocyte sedimentation rate (ESR) $\geq$ 60 mm/h and/or C-reactive protein (CRP) $\geq$3.0 mg/dL. Only 5 patients had positive swab for GAS. Twelve patients (50%) were treated with steroid. Fourteen patients received secondary prophylaxis every 28 days, two of them switched to every 21 days.
Conclusion: Our study confirms the previous published data of Breda *et al.* (3) and Licciardi *et al.* (4) in other Italian areas supporting the evidence for high risk (HR) in our region. Furthermore, this study shows that in our population only few patients had the classical polyarthritis presentation. It is important to consider the new HR criteria for arthritis in order not to miss diagnosis. A careful clinical history is needed because the demonstration of GAS infection is often difficult. These data may strengthen the hypothesis to consider our whole country as a HR area for ARF. A nationwide study is mandatory.

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AB1068 CLINICAL FEATURES IN TURKISH CHILDREN WITH CHRONIC NON-BACTERIAL OSTEOMYELITIS

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Background: Chronic non-bacterial osteomyelitis (CNO) is a rare bone disease of autoinflammatory origin. CNO patients present with bone, joint pain and swelling. Other autoinflammatory disorders such as palmoplantar pustulosis, psoriasis and inflammatory bowel disease may accompany CNO.
Objectives: Our aim is to determine demographic, clinical and radiological characteristics of Turkish children with CNO.
Methods: Clinical, laboratory features and imaging findings were retrospectively analyzed in 28 CNO Turkish patients from three pediatric rheumatology centers. Clinical findings, radiological data, treatments and outcome were evaluated.
Results: Of the 28 CNO patients, 10 (35.7%) female and 18 (64.3%) were male. Mean age at diagnosis was 9.54 (3-16.5) year. The lag time between symptoms and diagnosis was 6.5 (0-60) months. The median follow-up time was 18.5 (1-107) months. At the time of admission, symptoms were arthralgia (75.0%), bone pain (64.3%), limping (32.1%), swelling (28.6%), weakness (10.7%), and fever (3.6%). Two patients had inflammatory bowel disease (IBD) and six had familial Mediterranean fever (FMF). The bone lesions were mostly multifocal (%96.4),%3.6 was unifocal. The most frequently involved bones were femur (67.9%), tibia (53.6%) and pelvic bones (32.2%). (Table I) The patients had mildly elevated CRP and ESH levels. HLA B27 association was 25%. ANA was positive in 5%. All patients were RF negative. Six patients had mutations in the MEFV gene, all of them had M694V allele. The most common finding in Histopathological examination (n=9) demonstrated sterile mixed cell infiltration composed of lymphocyte and neutrophils. MRI results were mostly defined as bone marrow edema, and bone scan findings showed increased osteoblastic activity. All of patients received NSAIDs. Other drugs were methotrexate (46.4%) sulfasalazine (39%), steroids (25%), anti-TNF drugs (32%), and pamidronate (25%). Response to treatment was partial in 32.1%, whereas 39.3% patients had complete remission. There was no response in 8 patients (28.6%).
Conclusion: CNO was first described by Giedion in 1972 as multifocal bone lesions of inflammatory origin.[1] There are 500 CNO patients reported in the literature as individual or case series.[2] The laboratory findings are nonspecific and the diagnosis is mostly clinical and radiological. MRI and bone scan are the most commonly used imaging modalities. Histopathological results are non-specific, and may be necessary to exclude infection and malignancy. NSAIDs, sulfasalazine and methotrexate are mostly used in the treatment. Steroids, biological drugs and pamidronate are other options. There is still need for further treatment options, because there are significant number of resistant patients.

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Affected bones	***n***	***%***
Femur	19	67.9
Tibia	15	53.6
Pelvic bones	9	32.1
Calcaneus-talus	6	21.4
Clavicula	4	14.3
Mandible	3	10.7
Vertebra	2	7.1
Metatars	2	7.1
Sacrum	1	3.6
Cranium	1	3.6
Localisation	***n***	***%***
Metaphysis	17	60.7
Metaphysis-epiphysis	6	21.4
Metaphysis-diaphysis	4	14.3
Diaphysis	1	3.6

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AB1069 HYPERZINCAEMIA AND HYPERCALPROTECTINEMIA SYNDROME: MORE THAN JUST AUTOINFLAMMATION?

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Background: Hyperzincaemia and hypercalprotectinemia (HandH) syndrome has been described as a new rare entity characterized by recurrent infections, dermatological involvement, increased inflammatory markers, hepatosplenomegalia and anemia. Little is known about its heterogeneous presentation, pathophysiology and treatment.
Objectives: To describe three cases with HandH syndrome
Methods: Serum calprotectin (MRP8/14) was measured according to Buehlmann assay (ELISA) and plasmatic zinc by atomic absorption spectrometry
Results: Three patients were referred to our centre because an history characterized by recurrent episodes of skin rash, severe oral aphtosis and increased level of serum amyloid A (SAA). Patient 1 presented, since the age of ten years, with recurrent episodes of fever and rash; skin biopsy showed a picture consistent with a lymphocytic lichenoid vasculitis resembling erythema multiforme. Patient 2 presented at birth, with hemolitic anemia and thrombocytopenia. At the age of 5 she was admitted to another hospital due to EBV related hemophagocytic limphohystiocytosis (HLH). At the age of 8, she was first seen at our center because of a persistent desquamant erythematous rash with recurrent abdominal pain and recurrent arthritis. Intestinal biopsy showed small intestine inflammation (erosions in the digiunum). Patient 3 presented with recurrent episodes of fever, rash, two episodes of transient hip synovitis and muscoloskeletal pain. A bone scintigraphy was performed resulting normal. Patients 1 and 2 suffered from recurrent infections (pneumonia, otitis, skin abscesses). Immunological studies revealed in patient 2 a reduction of memory B cells and a reduced response to Toll like receptor 9 agonist. Of note, all the patients presented in their medical history at least one episode of vasculitis: patients 1 and 3 suffered from Schonlein-Henoch's purpura at the age of 11 and 3 respectively and patient 2 had at the age of 2 years an undefined vasculitis (evaluated elsewhere). Laboratory tests showed in all patients elevated inflammatory markers, zinchemia and serum calprotectin (table)

	Patient 1	Patient 2	Patient 3
Age at 1st evaluation (years)	14.3	7.8	4.9
Sex	F	F	M
Clinical features	Recurrent fever, recurrent infection, vasculitis, skin rash, musculoskeletal pain	Hemolitic anemia, thrombocytopenia, HLH, vasculitis, recurrent infections, recurrent fever, arthritis, skin rash, gastrointestinal involvement	Recurrent fever, skin rash, vasculitis, arthritis, musculoskeletal pain
SAA	13-23	27,5-76,4	20
Zinchemia (mcg/dl) (n.v. 80-125)	132-143	102-233	147
Serum MRP8/14 (ng/ml) (n.v. < 2900)	22431	10383	10363

Figure 1

Conclusion: We report three patients with high serum levels of calprotectinemia and zinch presenting with a clinical phenotype consistent with previously reported cases. The presence of vasculitis in all of the patients suggest that it may represent the first symptom of this condition. Vasculitis could lead to an increase of serum calprotectin already proposed as a marker of vascular impairment. Moreover, considering the immunological defect detected in one of our patient, we speculate that recurrent infections described in this syndrome may underline an immune-dysregulation process in which the role of zinc metabolism needs to be assessed
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AB1070 MICROVASCULAR NAILFOLD ALTERATIONS BY VIDEOCAPILLAROSCOPY IN PATIENTS WITH RHEUMATOLOGICAL SYMPTOMS WITH JUVENILE INFLAMMATORY BOWEL DISEASE

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Background: Juvenile Inflammatory Bowel Disease (jIBD) is a chronic relapsing inflammatory condition of the gastrointestinal system that includes Crohn's disease (CD) and Ulcerative Colitis (UC) and develops during childhood or adolescence in up to 25% of patients and affects the patients and parent s quality of life. Endothelial dysfunction is considered one of the etiological factors of jIBD. Nailfold videocapillaroscopy is one of the best and safest diagnostic non-invasive imaging techniques to analyze microvascular abnormalities. Previous studies describe the involvement of the microvasculature in the pathogenesis of this diseases, suggested that chronic mesenteric vasculitis is a pathogenetic mechanism in CD. Nailfold abnormalities founded are similar to those observed in some systemic vasculitidies. Subpapillary venous plexus dropout and low vessel density were previously reported.
Objectives: In this abstract we describe nailfold videocapillaroscopy findings in patients with jIBD and their correlation with disease activity.
Methods: This is a prospective and analytical study thad recruited pediatric patients between 2 and 18 years with jIBD. A single cross-sectional 8-finger nailfold videocapillaroscopy was performed using a 200x optical probe videocapillaroscope. The images were collected, encoded and stored using the Optipix® software. Qualitative, quantitative and semi-quantitative assessment for architecture were scored following the international definitions for the capillary abnormalities Blood chemistry, C-reactive Protein, erythrocyte sedimentation rate, antineutrophil cytoplasmic antibody, and calprotectin were performed. Sociodemographic data, clinical eva- luation, confirmation of IBD criteria, disease history, and activity evaluation were collected from patients clinical records. Statistics were performed using Spearman test to evaluate the correlation coefficient between the variables under study.
Results: 10 patients with jIBD were included 60% male, 50% Ulcerative Colitis and 50% Crohn s Disease. 20% had severe disease according the Pediatric Ulcerative Colitis Activity index, Pediatric Crohon s Disease activity index showed mild activity in just one patient. Rheumatological manifestations were found in 40%,4 had arthritis, systemic vasculitidies in 2 and 10% hematological manifestations. Abnormal endoscopy in 60% showing pancolitis. Normal microvasculature pattern were only found in 3 patients (25%), microangiopathy was found in 75%, edema of the nailfold bed was present in n=6 patients, a low capillary density was found in 100% of the patients with disease activity. A statistical negative correlation between the number of capillarities per millimeter and disease activity was found (coefficient -0.936, p=0.001).
Conclusion: Previous data showed that low capillary density was find in patients with jIBD, the data in this pilot study are consistent with those findings. The assessment of the microvasculature through the use of videcapillaroscopy could be useful in this diseases.

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