Results: A total of 164 patients were included. All 164 patients had S100A8/9 collected but only 151 had S100A12 samples. Patients with sJIA had higher levels of S100A8/9 and S100A12 compared to patients with non-systemic JIA (nsJIA) and periodic fever syndrome (PFS). CRP levels were not statistically different among diagnosis while S100 proteins were. Both S100A8/9 and S100A12 showed a good ability to differentiate sJIA from PFS (AUC=0.76 for both) compared to AUC=0.6 for CRP. At a cut-off value of 164 ng/ml, S100A12 was 71% sensitive and 73% specific in differentiating between sJIA and PFS (Figure 1). In JIA there was no correlation between S100 protein levels and AJC, either sJIA or nsJIA patients. S100A8/9 and S100A12 were significantly elevated in active sJIA patients compared to inactive ones while CRP levels were not different. S100 proteins levels were significantly higher during a flare of PFS compared to inactive disease, as well as CRP. There was no difference in S100 proteins and CRP in patients with or without tocilizumab treatment (small number on tocilizumab).

Conclusion: S100A8/9 and S100A12 are particularly elevated in sJIA compared to nsJIA and PFS. They are useful in distinguishing sJIA from other diagnoses with similar presentations, such as PFS. S100 proteins were significantly higher in active disease status of patients with sJIA and PFS compared to inactive status and their levels were closer associated with sJIA disease activity than CRP. The S100 proteins could provide valuable data in the work-up of patients with autoinflammatory conditions and monitoring disease activity in sJIA and PFS patients.

Figure 1. Panels (a) and (b) show serum levels of S100A8/9 (a) and S100A12 (b) across diagnoses. Values represent median and IQR (error bars). Comparison between diagnoses are done by Kruskal-Wallis test. Panels (c) and (d) show receiver operating characteristic (ROC) curves for S100A8/9 (c) and S100A12 (d) in differentiating systemic juvenile idiopathic arthritis (sJIA) vs. periodic fever syndrome (PFS). The area under the curve (AUC) for both S100A8/9 and S100A12 is 0.76. Sensitivities and specificities for the cut-off values are outlined.

* $p \leq 0.05$, ** $p \leq 0.01$, *** $p \leq 0.001$, **** $p \leq 0.0001$

Figure 1

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AB1066

DEFICIENCY OF ADENOSINE DEAMINASE 2: THE IMPORTANCE OF A DIAGNOSIS BEFORE IRREVERSIBLE DAMAGE

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Background: In 2014, two independent groups, Zhou et al.(1) and Navon-Elkan et al.(2), described the first 34 patients with disease-associated mutations in adenosine deaminase 2 (ADA2).

Objectives: To describe a new case of ADA2 deficiency (DADA2) in a 15 years old girl presenting with acute lower limb flaccid paralysis.

Methods:

Results: A 15 years old girl presented with acute lower limb flaccid paralysis. Neurologic exam showed peripheral deficit of the VII cranial nerve, absent motor activity at lower limbs with sensitive deficit. At physical examination she had marked and diffuse livedo reticularis. She underwent a brain and spinal magnetic resonance imaging (MRI) revealing multiple areas of signal abnormalities (T2 hyperintensity) in both thalamus and basal ganglia, small area in the spinal cord at C2-C3, at D10-D11 and L1 levels; these images were related to vascular damage. Blood exams showed high blood counts and inflammatory markers (C-reactive protein 16.76 mg/dl). Coagulation was normal, antiphospholipid and antinuclear antibodies were negative, immunological work-up showed normal immunoglobulins level, T and B phenotype. She received intravenous steroid and high-dose intravenous immunoglobulin. The constellation of findings led to clinical suspicion for DADA2, which was confirmed by low plasma ADA2 activity. For the presence of microhematuria, worsening proteinuria and hypertension, vascular ultrasound was performed with no evidence of renal artery stenosis. She underwent renal biopsy negative for glomerulonephritis, renal vasculitis and amyloidosis. Following the diagnosis of DADA2, she was initiated on etanercept 50 mg once weekly and she was able to taper steroid with normalization of inflammatory markers and improvement of skin rash.

The girl is the second of 3 children, born from consanguineous parents of Italian origin. By the age of 1 she started suffering from fever, arthritis, pericarditis with elevated inflammatory markers and was diagnosed with systemic juvenile idiopathic arthritis (sJIA) and chronic disease anemia. At that time, she already presented livedo reticularis. She was treated with short term oral steroid and methotrexate with remission of fever and articular symptoms. At the age of 2 she had a convulsive crisis, polysomnography and brain MRI were negative. One year later she presented a convulsive status epilepticus, brain MRI was repeated and resulted negative. She was started on topiramate with no further crisis, treatment was stopped 2 years later. When she was 9, she was admitted for pancytopenia and she was diagnosed of Leishmania infection. The two siblings of the girl were tested for plasma ADA2 activity and both resulted deficient. The boy is 12 years old with mild development delay and a previous diagnosis of sJIA when he was 5. He was treated with steroid and Methotrexate with remission. The older sister is now 19 years old and she had an episode of erythema nodosum. Genetic test of all family members is still pending.

Conclusion: Given the important morbidity, especially in consanguineous families, a high index of suspicion is needed for early diagnosis and precocious intervention, before persistent neurologic damage has been established. Our report describes for the first time a new neurological presentation as acute transverse myelitis in a patient with DADA2.

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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 polyarthralgia 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 ($\geq 38.5^{\circ}\text{C}$), 11 children had elevated inflammatory markers as erythrocyte sedimentation rate (ESR) ≥ 60 mm/h and/or C-reactive protein (CRP) ≥ 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 ESR 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
Clavícula	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