related with the introduction of biologic Disease-Modifying Antirheumatic Drugs (bDMARDs) and the uninterrupted monitoring following the transition of young patients to adult rheumatology settings.

Objectives: To capture the contemporary ERA profile in Northern Greek patients by analyzing the characteristics and treatment outcome in the era of bDMARDs.

Methods: This retrospective cohort study included patients who had been monitored on a 3-month schedule for ≥ 12 months, from 2000 to 2017. The periodic metric assessment included the disease status and burden by applying contemporary tools in respect to activity, clinical remission (CR) and damage (cJADAS, JSpADA, Wallace criteria for CR and JADI, respectively).

Results: Forty-three patients, mainly male (60%) with a mean age at disease onset of 10.75 (SD:2.75) years were enrolled. The predominant joints were the hip, ankle and sacroiliac (56%, 49% and 46%, respectively). Median lag time from diagnosis to bDMARDs initiation was 8.5 months. Patients with sacroiliitis were more likely to receive bDMARDs (hazard ratio [HR]:3.26, 95% confidence interval [CI]: 1.35, 7.88). Thirty six patients (84%) achieved clinical remission (CR) on medication (CRONM), within a median time of 11 months and correlated with compliance (HR:3.62, 95%CI: 1.34, 9.76). Twenty patients (47%) experienced a flare following CR, mainly as a single episode (75%). The median flare-free survival following remission on and off medication (CROFFM) was 42 and 34 months, respectively. At the last evaluation, both median baseline cJADAS (8), and JSpADA (2) dropped to 0, while 13 patients (30%) were in CROFFM, 17 (40%) in CRONM, and 13 (30%) had persistent disease activity. The median percentage of CR per patient was 54% and no patient had JADI >0.

Conclusion: Early administration of bDMARDs and compliance to monitoring and treatment improved the long-term outcome in ERA. Axial involvement emerged as a negative prognostic factor with an increased need for bDMARDs and diminished rates of CR.

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AB0977

DISEASE COURSE AND TREATMENT RESPONSES IN JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS; A SINGLE CENTER EXPERIENCE

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Background: Systemic lupus erythematosus (SLE) is a chronic autoimmune inflammatory disease that may cause morbidity and mortality by affecting multiple systems. The 10-20% of patients have juvenile onset and this cluster have may more severe kidney, neuropsychiatric or hematological involvement.

Objectives: The aim of this study was to assess the clinical and laboratory characteristics, disease activity, and treatment response of patients with juvenile SLE (jSLE).

Methods: This is a retrospective study involving patients between 1 July 2016 and 1 January 2020. The data of patients diagnosed with jSLE and followed up for a minimum of 6 months, were collected. The SLEDAI-2K scores at initiation and at the follow-up (1st, 3rd, 6th, and 12th months of treatment) were examined. The SLEDAI-2K score was considered to be ≤ 4 , for disease remission status.

Results: A total of 49 children were included in to the study. The female/male ratio was 4.4/1 and the median age of the patients at the diagnosis was 13 (IQR: 11.1–15.2) years. The median follow-up of patients was 19 (IQR: 12–25) month. Four of the patients were diagnosed with monogenic SLE. Two siblings were diagnosed with c3 deficiency and two were diagnosed with familial chilblain lupus. The most common clinical findings were found musculoskeletal complaints (69.4%), malar rash (51%), oral ulcers (38.8%), and fever (30.6%), respectively in over all the group. The frequency of involvement of the system and organs was as follows; mucocutaneous 77.6%, musculoskeletal 69.4%, renal 44.9%, hematological 34.7%, serous membranes 16.3%, neuropsychiatric 12.2%, respectively. All patients had anti-nuclear antibody positivity, while 46.9% had anti-ds DNA, 14.3% had anti-Sm and 8.2% had antiphospholipid antibody positivity. While all patients received hydroxychloroquine treatment, 22.4% of the patients were received were mycophenolate mofetil, 22.4% were azathioprine, 14.3% cyclophosphamide, 12.2% methotrexate and 10.2% were rituximab. The median SLEDAI-2K score was 14 (IQR: 10–18.5) at admission, besides it was found to 6 (IQR: 4–12), 4 (IQR: 2–6), 2 (IQR: 0–6) in the 1st, 6th and 12th months of treatment, respectively. While 98% of the patients had active disease at admission, 67.3% at 1 months, 32.7% at 6 months and 22.4% at 12 months still had active disease (SLEDAI-2K >4). Patients with initially high SLEDAI-2K scores had significantly lower remission rates in the first month ($p=0.003$). It was observed that patients with high SLEDAI-2K scores in admission were more

resistant to conventional immunosuppressive treatments and the use of rituximab was more frequent in these patients. At least one major organ (renal, hematological, neurological) were affected in 57% of patients. The remission rate of these patients at 6 months was found significantly decreased compared to the others ($p < 0.005$). Renal biopsy was performed in 21 patients (42.9%). 12 of them had type 4 lupus nephritis (LN), 5 had type 2, 2 had type 3, and 1 had type 5. It was observed that patients with renal involvement were the group that reached remission latest.

Conclusion: The presence of high initial SLEDAI-2K scores and the major organ involvement have poor predictive value to achieve inactive disease.

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AB0978

EFFICACY OF ANAKINRA TREATMENT IN PEDIATRIC RHEUMATIC DISEASES; A SINGLE-CENTER EXPERIENCE

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Background: Anakinra, a recombinant IL-1 receptor antagonist, is a treatment option that acts by blocking the biological activity of IL-1 in autoinflammatory conditions. The diseases that the IL-1 was over expressed are the potential conditions for this treatment. Such as familial Mediterranean fever (FMF), cryopyrin-associated periodic syndrome (CAPS), and hyperimmunoglobulin D syndrome (HIDS) with monogenic inheritance, and systemic juvenile idiopathic arthritis (SoJIA) or idiopathic recurrent pericarditis as non-Mendelian polygenic diseases, can be listed as examples of these diseases.

Objectives: The aim of this study was to review the efficacy of anakinra treatment in children with rheumatic disease followed in our center.

Methods: The study group consisted of children with pediatric rheumatic diseases followed up in the Pediatric Rheumatology Department of University of Health Sciences and treated with anakinra (anti-IL 1) for at least one month, between 1 July 2016 and 1 January 2020. The data of these patients were collected retrospectively. The disease activity of the patients at 3rd month and 12th month after the treatment were assessed. We aim to report our experiences of pediatric rheumatic diseases treated with anakinra.

Results: There were 28 patients treated with anakinra for the different pediatric rheumatic diseases. The diagnoses of these patients were as follows; eight were macrophage activation syndrome (MAS) complicating SoJIA, six were HIDS, four were CAPS, four were FMF, four were idiopathic recurrent pericarditis, one was deficiency of interleukin-36 receptor antagonist (DITRA), and one was undefined systemic autoinflammatory disease. 46.4% of the patients were male and 53.6% were female. The median age of diagnosis of the patients was 6.5 (interquartile range (IQR): 4-12.7) years. The median follow-up duration of the patients was 14 (IQR: 3.7-28) months. The patients median anakinra treatment duration was 3 (IQR: 1-4) months. Fever reduced and C-reactive protein normalized within median 2 (IQR: 1-3) and 5 (IQR: 5-7) days, respectively. In the 3rd month after treatment; It was observed that 53.6% of patients achieved a complete remission (no attack was seen or MAS was improved). The frequency of attacks were decreased more than 50% in 35.7% of patients and less than 50% in 7.1%. 3.6% of patients were unresponsive to treatment. In the 12th month assessment after the initiation of treatment, it was observed that 28.6% of patients were still under anakinra treatment and in remission, 10.7% of them were in remission without anakinra treatment. In 60.7% of patients, anakinra was switch to other biological treatments for different reasons (35.7% partial response or unresponsiveness, 17.8% injection site reactions and 7.1% daily-injection difficulty). Biologic drug switch to canakinumab and tocilizumab was observed in 88.2% and 11.8% of patients, respectively. One patient developed recurrent MAS episodes when the anakinra dose was tapered, and one another patient was unresponsive to the anakinra and dead due to secondary to MAS.

Conclusion: Anakinra seems to be a successful treatment to achieve inactive disease in a significant portion of patients in the early period. The recurrence of disease attacks while drug tapering and injection site reactions were appears the main causes of treatment switch or discontinuation.

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AB0979

MEVALONATE KINASE DEFICIENCY AT TREATMENT WITH CANAKINUMAB: RARE BLEATED CUTANEOUS FEATURE

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Background: Mevalonate Kinase Deficiency (MKD) is an autosomal recessive autoinflammatory syndrome. Diagnostic criteria are based in clinical and genetic features (mutations in MVK gene) and its main treatment consists of blocking IL-1¹⁻⁵.

Objectives: To describe a case-series of a rare bleated cutaneous feature.

Methods: From January 2004 to September 2019, all cases diagnosed of MKD have been reviewed.

Results: 15 patients had MKD diagnosis (11 pathogenic mutations, homozygosis or double heterozygosis). Most common symptoms were oral and genital aphthous, abdominal pain, adenopathies and recurrent fever. Mean age of diagnosis was 10,67 years old (8,67 years later from the beginning of symptoms). Mean time of follow-up was 10 years. 3 patients developed this rare bleated cutaneous feature: suppurative hydrosadenitis. When the first injury appeared, all were at treatment with Canakinumab (mean time of treatment 4 years) and had 7,67 years of MKD course. This comorbidity began as repeating abscesses in folds with apocrine glands and hair follicles (armpits, inguinal, anal and genital folds). 2 patients continued Canakinumab and 1 switched to Adalimumab because of severity of cutaneous involvement.

Conclusion: This is the first case-series showing suppurative hydrosadenitis associated to MKD that has not been described in literature. Both theories have been found: as immune-mediated disease partnering this autoinflammatory syndrome (MKD) or as adverse event of treatment (anti-IL-1, less presumable).

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AB0980

CASE-REVIEW ON FAMILIAL MEDITERRANEAN FEVER IN A SPECIALIZED CENTRE

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Background: Familial Mediterranean Fever (FMF) is a genetic autoinflammatory disorder caused mostly by mutations in MEFV gene. Its inheritance is autosomal recessive and is the most frequent periodic fever syndrome. First-line treatment is based in colchicine use, so biologics (anti-IL-1) are reserved for refractory cases^{1,2}.

Objectives: To account for clinic and treatment features of patients with FMF in a specialized center as opposed to non-referent centers.

Methods: This study was developed in the Pediatric Rheumatology Service in Hospital Universitario y Politécnico La Fe de Valencia. Demographic, clinic and treatment data were collected from patients diagnosed of FMF since January 2004 to September 2019.

Results: 106 patients met last FMF criteria³. 55% had a pathogenic mutation in genetic analysis. 52% were female. Before 10 years old, 71% of patients had the diagnosis (51% before 4 years old). Arthralgia/myalgia (73%), periodic fever (62%) and abdominal pain (54%) were the most common symptoms. Systemic

Juvenile Idiopathic Arthritis (JIA, 6), other forms of JIA (9) and vasculitis (10) were the most prevalent comorbidities. When talking about treatment, 76,4% received Colchicine (60,5% with good response), 22,6% needed a classical disease modifying antirheumatic drug (mostly Methotrexate) and 22 patients got biologic treatment (73% anti-IL-1).

Conclusion: When analyzing this case-review, JIA has a strong association with our patients, so it could explain severe disease activity and more articular involvement. This could be an illustration to the higher use of Methotrexate. Also, the most relevant symptom was arthralgia while fever is the most frequent in literature. Likewise, age of diagnosis has been earlier than other case-series (this would be more frequent in other autoinflammatory syndromes, as literature relates)^{1,2,4}.

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AB0981

VALIDATION OF THE CAREGIVERS QUESTIONNAIRE (IMPACT OF PEDIATRIC RHEUMATIC DISEASES ON CAREGIVERS MULTIASSESSMENT QUESTIONNAIRE) IN PEDIATRIC RHEUMATIC DISEASES

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Background: The CAREGIVERS questionnaire is used as a multidimension screening instrument for burden of caregivers, it allows to examine the impact of juvenile idiopathic arthritis (JIA) on several areas of the caregiver's life, looking for specific concerns and needs in which treatment could be required. This instrument has shown a good understanding among participants, reliability and consistency to measure the psychosocial and economic impact on primary caregivers of patients with JIA.

Objectives: The aim of this study is to validate CAREGIVERS questionnaire with the most common pediatric rheumatic diseases, such as JIA, systemic lupus erythematosus (SLE) and juvenile dermatomyositis (JDM).

Methods: The external validation phase of the constructed questionnaire will be carried out following Feinstein's clinimetric and psychometrics methodology, to measure the impact of the disease on the caregiver and create a risk profile for treatment detachment.

Results: A total of 200 questionnaires are applied to caregivers of pediatric patients with rheumatic diseases: 109 caregivers of patients with JIA, 28 caregivers of patients with DMJ and 63 caregivers of patients with SLE. When analyzing all the questions together, a general Cronbach's alpha of 0.6751 was obtained (Table 1).

Table 1. External validation of the Questionnaire

Dimension	Numero de ítems	Alfa de Cronbach
I. Impacto Emocional	6	0.5669
II. Impacto Social	3	0.47
IIIA. Impacto Financiero	4	0.6736
IIIB. Impacto Laboral	3	0.3151
IV. Impacto Familiar	5	0.4948
V. Impacto en la Relación entre Cuidador-paciente	1	NA
VI. Impacto en la relación de pareja	1	NA
VII. Impacto en la Religión/Creencias/Espiritualidad	1	NA
VIII. Impacto de las Redes Sociales	4	0.6642
Total	28	0.6751