inactive group. Antiphospholipid antibody was present in 3 (23.1%) in active disease activity group and 5 (25%) in inactive disease activity group. The mean serum concentrations of DNAase1 were 15.394 ng/ml in active disease group and 15.205 ng/ml in inactive disease group. There was no statistically significant difference in the serum DNAase1 concentrations between the two groups (p=0.943). There was also no significant difference in the mean serum concentration of DNAase1 in patients with or without nephritis (p= 0.080).
Conclusion: The present study could not established any correlation between serum DNAase1 levels and disease activity in pediatric-onset SLE. There was no association between serum DNAase1 levels and organ involvement such as nephritis in the enrolled patients

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FRI0567 ABATACEPT THERAPY EXPERIENCE IN THE CASE SERIES OF PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS ASSOCIATED WITH OVERLAP SYNDROME AND/OR SJOGREN'S SYNDROME

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Background: The choice of Biologics in patients with juvenile idiopathic arthritis (JIA) who have overlap syndrome with juvenile onset of systemic lupus erythematosus (JSLE), juvenile dermatomyositis (JDM), juvenile systemic sclerosis (jSSc) and juvenile Sjogren's syndrome (jSS) is difficult. TNF inhibitors are limited in this kind of disorders and abatacept (ABA) seems to be the favorable option for children, but there are not enough studies in pediatric overlap syndrome and jSS.
Objectives: The evaluation of the abatacept therapy in case series of patients with JIA associated with overlap syndrome and/or jSS.
Methods: Patients who fulfilled the criteria for JIA and the criteria of above mentioned rheumatic disorders and received abatacept therapy according to JIA indication in real clinical practice of our rheumatological clinic.
Results: 11 patients included in the retrospective study (8 girls, 3 boys). The average age of patients was 15.1 years (from 7 to 18 years). Mean duration of disease was 5.4 years. 9 patients were RF positive, 4 patients had ACCP. All of the patients had antinuclear antibody (from 1:320 to 1:2560), 3 patients had positive anti-Ro antibody. Sjogren's syndrome was the most common concomitant autoimmune disorder (**n** = 8), JDM (**n** = 2), jSSc (**n** = 2), JSLE (**n** = 1). 7 patients were treated by a low dose of prednisolone (median 5.7 mg/day). All patients received methotrexate in dose 10-15 mg/m^2/week. ABA was indicated as first line of biology therapy in 10 patients, in one patient after adalimumab withdrawn due to severe flare. Mean duration of abatacept treatment was 23.7 month (from 6 to 78). The most of the patients (82%) had excellent stable effect of ABA treatment, but two patients needed to be switched to rituximab due to secondary inefficacy. We observed wonderful result of treatment with full remission of arthritis, calcinosis regression and resolving of Sjogren's syndrome in the boy with overlap syndrome treated by ABA for 78 months. The seroconversion from initial levels of RF 32.9 IU/ml (>2N), antinuclear antibody 1:640, anti-Ro antibody 200 U/ml (>9N) to negative results was registered. The ABA was good tolerated in all patients. In one case it was canceled due to pregnancy.
Conclusion: ABA is efficacy and safety option in patients with pediatric overlap syndrome and jSS. ABA is good alternative to anti-B cell therapy and preferable choice among Biologics in such category of patients.
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FRI0568 THE USE OF NEXT GENERATION SEQUENCING PANEL IN UNDIFFERENTIATED AUTOINFLAMMATORY DISEASES IDENTIFY A SEPARATE SUBSET OF COLCHICINE-RESPONDER RECURRENT FEVERS DISTINCT FROM PFAPA SYNDROME

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Background: The number of monogenic innate immune system disorders classified as systemic autoinflammatory diseases (SAID) has increased during the recent years. However, more than 70% of patients with clinical manifestations of SAID not achieved a molecular diagnosis, thus getting into the so-called undifferentiated or undefined SAID (uSAID).
Objectives: The aim of the present study is to characterize a subgroup of patients with recurrent fever episodes distinct to PFAPA that turned out to be negative to a large 41-gene NGS panel.
Methods: We designed an NGS panel including 41 genes clustered in seven subpanels.
Results: Fifty patients were enrolled in the study. 34 patients (72%) displayed recurrent fevers and sixteen presented a chronic inflammatory disease course. A total of 100 gene variants were found (mean 2 per patient; range 0-6). Mutations with a sure or possible diagnostic impact were detected in five patients (10%). Differently from PFAPA syndrome (table), genetically negative patients with recurrent fevers presented episodes that lasted on average of six days (P<0.0001). Abdominal pain and limb pain were the most common symptoms. The classic PFAPA triad (pharingotonsillitis, aphthosis and cervical lymphadenopathy) was less frequently reported (P<0.0001) while skin rash and arthritis were more frequent (P<0.0001). Eighteen patients were exclusively treated with steroid on demand with a high response rate (94%). In 18 patients, colchicine treatment was used with an overall complete or partial response of 78%.
Conclusion: Even with a low molecular diagnostics rate, an NGS-based approach is able to provide a final diagnosis in a proportion of uSAID patients. It also allows the identification of a subgroup of genetically negative patients with recurrent fever responding to steroid on demand and colchicine.

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Table. Clinical features of patients with undifferentiated recurrent fevers compared to PFAPA syndrome. *Values are number of patient (%) when not specified. NR = not reported. NS = not significant. P values were assessed using Chi square test or T-test as appropriate. If significant interactions were determined, a post-hoc test for multiple comparison was performed.*

Clinical features	Our cohort (34 patients)	Thomas et al. (82 patients)	Hofer et al. (301 patients)	Batu et al. (131 patients)	Pehlivan et al. (359 patients)	p value
Mean duration of episodes (days; ±SD)	5.9 ± 1.4	NR	NR	NR	4.0 ± 3.1	<.0001
Median interval between episodes (weeks; ±SD)	3.0 ± 0.3	NR	NR	NR	3.3 ± 1.5	NS
Abdominal pain	17 (50)	40 (49)	136 (45)	60 (46)	102 (29)	NS
Arthritis	7 (21)	NR	8 (3)	NR	7 (0)	<.0001
Skin rash	11 (32)	7 (9)	38 (13)	7 (5)	NR	<.0001
Pharyngotonsillitis	13 (38)	59 (72)	271 (90)	126 (96)	359 (100)	<.0001
Cervical lymphadenopathy	6 (18)	72 (88)	236 (78)	70 (53)	215 (60)	<.0001
Oral aphthosis	13 (38)	57 (70)	171 (57)	56 (43)	317 (88)	<.0001
Response to colchicine	14/18 (78)	0/1 (0)	NR	5/11 (45)	24/45 (53)	NS

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FRI0569 WHAT DOES IT MEAN TO BECOME PREGNANT WITH JUVENILE IDIOPATHIC ARTHRITIS? A MONOCENTRIC EXPERIENCE IN A TERTIARY CENTRE OF MILAN DEDICATED TO YOUNG ADULTS AFFECTED BY JIA

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Background: During the last seventeen years, biological and non-biological therapies have been used in our center in an open prospective study for the treatment of refractory JIA not only in paediatric age but also in young adults. The availability of more effective drugs for JIA has dramatically increased the number of patients willing to become pregnant. Data regarding the effect of JIA on pregnancy outcome are scant.
Objectives: The study aims to evaluate the capability of JIA patients to become pregnant, the effects of drugs on pregnancy outcome and the impact of gestation on the disease course.
Methods: JIA patients regularly followed up in our Transition Clinic of the Division of Rheumatology, Gaetano Pini Institute of Milan, were enrolled in the study at positive pregnancy test. During the whole pregnancy patients underwent monthly clinical examination and obstetric ultrasound. Data regarding disease activity and pregnancy course (fetal morphometric parameters, fetal heart rate, fetal growth, Doppler velocimetry) were collected. Gestational age, birth weight and APGAR score were also recorded.
Results: 29 pregnancies in 23 women affected by refractory JIA became pregnant during a 17 years follow up. All patients had a long lasting polyarticular disease (median duration of 23 years) not responsive to DMARDs, and became pregnant during biologic therapy. Patients were treated with Etanercept (11 patients), Golimumab (4 patients), Rituximab (3 patients), Adalimumab and Certolizumab (2 patients respectively) as monotherapy, and in most of the cases after multiple switches. One woman was treated with Etanercept during the first pregnancy and Adalimumab during her second pregnancy. Three patients decided for an elective termination and 3 experienced an early miscarriage; among 23 pregnancies resulting in live born infants, only 3 had premature births and 1 cleft palate. A pregnancy was complicated by gestosis, two by placental detachment. All the babies were followed up during the 17 years of observation and did not experience any major late complication. Eight patients were subjected to intra-articular infiltrations during pregnancy to switch off the disease, 18 patients resumed therapy shortly after childbirth and only 7 patients decided to breastfeed.
Conclusion: despite a large amount of studies demonstrating the safety of anti-TNF during pregnancy, data regarding the effects of biologics on pregnancy outcome in JIA are still lacking. The very low number of patients treated with traditional DMARDs achieving low disease activity underline the pivotal role of new biologic drugs in the management of aggressive long standing JIA, to improve the quality of life of these patients, including family planning. In our experience, no greater number of unexpected complications or side effects were observed in JIA patients during pregnancy compared to the other autoimmune diseases. Discontinuation of therapy has increased the risk of flare, requiring local therapy, confirming that EULAR recommendations for the use of biologics during pregnancy can be applied also in JIA.

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FRI0570 CLINICAL CHARACTERISTICS IN PATIENTS WITH PEDIATRIC SYSTEMIC LUPUS ERYTHEMATOSUSDIAGNOSED BEFORE 6 -YEARS OLD IN A THIRD-LEVEL HOSPITAL IN MEXICO

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Background: Pediatric-onset Systemic Erythematosus Lupus (pSLE) is known to be more aggressive than adult-onset SLE. The disease is extremely rare under the age of 5 years and with severe course.
Another big difference is the association of pSLE with monogenic primary immune deficiencies (PID). Some PIDs have been associated with SLE or lupus-like manifestations: deficiencies of the classical complement pathway, female carriers of X-linked chronic granulomatous disease, IgA deficiency (present in 5% of pSLE).
It is important to consider PIDs in patients with severe manifestations in pSLE, in our country there are no studies of this association.
Objectives: To describe the clinical manifestations, laboratory features, characteristics and therapeutics of patients with diagnosis of SLE before 6 years of age at National Institute of Pediatrics in Mexico City
Methods: This is a retrospective study with the review of the patient's records with diagnosis of SLE before 6 years old, from the Immunology department between January 2005 and December 2016 at National Institute of Pediatrics in Mexico City.
Results: Between 2005-2016 there were 295 newly patients diagnosed with pediatric Systemic Lupus Erythematosus (pSLE) at our hospital, from these patients 2.7% (8) were diagnosed with pSLE before 6 years of age. According to this, 7 (87.5%) were female. The youngest patient was 26 months at moment of diagnosis.
Every patient had at least 4 criteria of the ACR classification for SLE. Only 2 (25%) had malar rash at diagnosis, and 1 (12.5%) presented discoid rash. Seventy-five percent of patients presented glomerulonephritis at diagnosis and also 75% had an Hematological disorder.
In terms of serology, 37.5% had positive native DNA antibody, 37.5% antibody to Sm protein and 37.5% antiphospholipid antibodies. One hundred percent of patients had presence of antinuclear antibody (ANA) at moment of diagnosis.
Conclusion: During 2005-2016 we found 295 newly patients with diagnosis of pediatric Systemic Erythematosus Lupus (pSLE), from these, eight patients (2.7%) were diagnosed before 6 years of age. Seventy-five percent presented recurrent infections and 50% with severe infections, and one patient died due to septic shock.
Systemic erythematosus lupus with an early onset has a more severe phenotype and has been associated with monogenic primary immune deficiency, presenting with severe infections and higher mortality.
In our country, we do not have genetic testing to help us determine the endless monogenic defects associated with early-onset SLE, being necessary to determine etiology and specific treatment as needed.
We recommended that in every patient with severe pSLE who develops life-threatening, infections or due to unusual germs, associated with hypocomplementemia and/or consanguinity, to perform an approach to rule out any primary immune deficiency.

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