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 establish 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

REFERENCES:

- [1] Skiljevic D, Jeremic I, Nikolic M, Andrejevic S, Sefik-Bukilica M, Stojimirovic B, et al. Serum DNase I activity in systemic lupus erythematosus: correlation with immunoserological markers, the disease activity and organ involvement. Clin Chem Lab Med. 2013 May;51(5):1083–91.

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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/m²/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.

Disclosure of Interests: None declared

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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 from 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 (pharyngotonsillitis, 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.

REFERENCES:

- [1] Rusmini M, et al. Next-generation sequencing and its initial applications for molecular diagnosis of systemic auto-inflammatory diseases. Ann Rheum Dis. 2016 Aug;75(8):1550-7.

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. (369 patients)	P value
Mean duration of episodes (days; $\pm$ SD)	5.9 $\pm$ 1.4	NR	NR	NR	4.0 $\pm$ 3.1	<.0001
Median interval between episodes (weeks; $\pm$ SD)	3.0 $\pm$ 0.3	NR	NR	NR	3.3 $\pm$ 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