was suspected and markers revealed serum ferritin 30.000 ng/ml, fibrinogen 250 mg/dl,Triglycerides 350 mg/dl and soluble CD25 12.000U/ml. Bone marrow aspiration confirmed the diagnosis.Cytomegalovirus (CMV) IgM and PCR were significantly high.With regard to all the previous clinical and laboratory features,a CMV-induced secondary MAS in JSLE was confirmed.Patient was managed according to HLH-2004 treatment protocol (pulsed methylprednisolone 30mg/kg/day,cyclosporine A 6mg/kg/day, IV etoposide 150mg/m²)and IVIG.Ganciclovir was also added (5mg/kg IV q12h x7 days). Patient showed marked clinical improvement together with significant diminution of the MAS laboratory markers to normal.Two weeks post discharge on oral dexamethasone and cyclosporine A, the patient showed a severe clinical and laboratory MAS relapse (serum ferritin 50.000 ng/ml). The previous treatment protocol was restarted together with adding Rituximab 375mg/m² x 3 doses two weeks apart.Patient showed marked clinical and laboratory improvement and was maintained on full dose oral prednisone and oral Cyclosporine A. Tight gradual withdrawal of oral steroids was done with close clinical and laboratory follow up.

Conclusion: JSLE is a major diagnostic conundrum in pediatrics owing to its extremely variable clinical manifestations which can mimic many common pediatric conditions(e.g.malignant disease,other auto-inflammatory conditions,malignancy and any specific organ associated disease).Compared to adults JSLE has a more severe disease presentation.MAS is a life threatening condition that requires high index of suspicion and prompt management.In our case, controlling JSLE activity using Rituximab was significantly beneficial in arresting HLH evolution.

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Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2019-eular.6857

AB0922 UPDATES ON THE EVIDENCE BASED INTERDISCIPLINARY GUIDELINES FOR HENOCH-SCHÖNLEIN PURPURA

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Background: Henoch-Schönlein purpura (HSP) is the most common childhood vasculitis, it is characterized by inflammation of small vessels leading to non-thrombocytopenic purpura, arthritis/arthralgia, GI hemorrhage and glomerulonephritis[1]. There is a very poor evidence base to guide the management of HSP, particularly for those with the severe forms of HSP nephritis, Up till now there are no EULAR or ACR recommendations for management of HSP.

Objectives: To set an updated interdisciplinary recommendations for the management of HSP

Methods: Interdisciplinary guidelines were developed with representative from PRINTO Egypt for pediatric rheumatology as well as patients' group. A systemic literature analysis in MEDLINE was performed, evidence and recommendations were graded, an algorithm for treatment and final statements were discussed in a consensus meeting (Nominal Group Technique). A preliminary draft was fine tuned and discussed thereafter by all participants (Delphi Procedure).

Results: Consensus was reached on recommendations, including a standardized treatment strategy according to the HSP severity state in the individual patient. In this updated interdisciplinary guideline for HSP; treatment was tailored to the patient's pathology and its severity (figure 1). Criteria for admission were identified as: severe arthritis or arthralgia limiting ability to weight bearing and mobilization, severe abdominal pain, GI hemorrhage, evidence of nephritis/nephritic syndrome or renal impairment, evidence of neurological symptoms. Criteria for early referral to pediatric nephrologist were also identified including: hypertension, abnormal renal function, macroscopic hematuria for 5-days, nephrotic syndrome, acute nephritic syndrome, as well as persistent proteinuria.

Conclusion: A total of seven evidence based interdisciplinary recommendations for management of HSP have been formulated, that provide an up-to-date guidance of HSP management.



Figure 1: recommendations for management of HSP

Figure 1

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Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2019-eular.713

AB0923 EFFECTS OF EXON 10 MUTATIONS VS NON-EXON 10 MUTATIONS ON FMF PHENOTYPE AND RESPONSE TO TREATMENT

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Background: Familial Mediterranean fever (FMF) is the most common monogenic periodic fever syndrome. The MEFV gene mutation encoding the pyrin protein results in an uncontrolled increase in interleukin-1. Today, more than 333 MEFV mutations have been identified; however, exon 10 mutations are still seem to be best correlated with clinical findings.

Objectives: In this study, we aim to investigate the role of exon 10 mutations vs non-exon 10 mutations on clinical features and response to treatment in patients with FMF.

Methods: Data charts of children (n=935) with FMF from Dokuz Eylul University childrens' hospital and Dr.B.Uz childrens' hospital were reviewed. Patients were divided into two groups with regard to having

Ann Rheum Dis: first published as 10.1136/annrheumdis-2019-eular.1589 on 27 June 2019. Downloaded from http://ard.bmj.com/ on September 2, 2021 by 193.140.151.30. Protected by copyright.

exon 10 mutation or non-exon 10 mutations. Genotype-phenotype features and response to treatment were compared.
Results: There were exon 10 mutations in 631 (67.5%) patients and non-exon 10 mutations in 304 (32.5) patients. The follow-up period was 50 (26-83.2) months. The age of symptoms onset was significantly lower in group with exon 10 positive than compared group with non-exon 10 mutation. There was no difference between the age of diagnosis and colchicine onset and the diagnosis delay time. The symptoms of fever, chest pain, and arthritis were significantly higher in the exon 10 mutation group than compared other group. Biological agent need was statiscally higher in exon 10 mutation group (4.8%) than group with non-exon 10 mutation (1.3%) (Table 1).
Conclusion: In our study, it was observed that cases with exon 10 mutation have early symptoms of disease. Fever, chest pain and joint findings were more prominent in cases with exon 10 mutation than cases with non-exon 10 mutation. Additionally, colchicine resistance should be kept in mind in cases with exon 10 mutation.

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Abstract AB0923 Table 1. Comparison of patients with exon 10 and non-exon 10 mutations

	Exon 10 (+)	Exon 10 (-)	p
n (%) = 935 (100)	631 (67.5)	304 (32.5)	
Female/Male (n) =445/490	303/328	142/162	0.707
Median age of symptoms onset (months)	**48 (24-86.5)**	**60 (26-109)**	**0.009**
Median age of diagnosis (months)	80 (47-127)	87 (54-136)	0.106
Median diagnosis delay time (months)	17 (6-38)	15 (7-36)	0.484
Symptoms n (%)			
Fever	**442 (70.3)**	**193 (64.1)**	**0.050**
Abdominal pain	458 (72.8)	206 (68.7)	0.191
Chest pain	**65 (10.3)**	**18 (6)**	**0.029**
Erysipelas like rash	11 (1.7)	10 (3.3)	0.127
Arthralgia	231(36.8)	110 (36.8)	0.999
Arthritis	**137(21.8)**	**46 (15.3)**	**0.019**
Myalgia	78 (12.5)	26 (8.6)	0.083
Emesis	12 (2)	12 (4)	0.061
Use of biologic agent	**30 (4.8)**	**4 (1.3)**	**0.009**

Disclosure of Interests: Hatice Adiguzel Dundar: None declared, ozge altug gucenmez Speakers bureau: Novartis, Abbvie, Ceyhun Acari: None declared, Serkan Turkucar: None declared, Balahan Makay Speakers bureau: Enzyvant, Novartis, Roche, Abbvie, Erbil Unsal Grant/research support from: Novartis, AbbVie, Roche, Koçak Pharma, Speakers bureau: Novartis, AbbVie, Roche, Koçak Pharma
DOI: 10.1136/annrheumdis-2019-eular.1589

AB0924 EVALUATION OF PERIPHERAL NERVOUS SYSTEM INVOLVEMENT IN PATIENTS WITH JUVENILE SYSTEMIC SCLEROSIS AND JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Juvenile systemic sclerosis (JSS) and juvenile systemic lupus erythematosus (JSLE) are rare connective tissue disorder characterized by multisystemic involvement, including gastrointestinal, cardiovascular, respiratory and nervous system complications. According to data from literature, peripheric nervous system (PNS) involvement is seen in 10-86.7% of adult patients with systemic sclerosis. Frequency of PNS disorders is reported as 10-86.7% in adult patients. Data on PNS involvement in patients with JSS and JSLE are scarce.
Objectives: We aimed to evaluate PNS involvement in patients with JSS and JSLE. Consequently, we sought to detect patients with PNS disorders, in order to enable early diagnosis and timely intervention.
Methods: Patients with JSS and JSLE were included in a cross-sectional study. Demographic and clinical data of patients were recorded during clinical visits. All of patients were evaluated and examined for sings of PNS involvement. In order to examine mononeuropathy, polyneuropathy and trigemino-facial involvement, all patients underwent routine nerve conduction studies (NCS), blink reflex (BR) and sympathetic skin responses (SSR) evaluation.
Results: Twenty JSS (15 (75%) female) and 18 (15 (83%) female) JSLE patients were initially included. All of JSS and JSLE patients had normal neurologic examination. NCS was normal in all JSS (20/20) and JSLE (18/18) patients. SSR was not recorded in 1 (5%) JSS and in 3 (16.67%) JSLE patients. BR was recorded in all JSLE (18/18) patients and in majority of JSS patients (19/20, 95%). According to SSR, mean latency of hand and foot was similar in both patients' groups. Amplitude of foot response was lower in JSS patients, comparing to JSLE (Table 1). Among JSLE patients, amplitude of hand response was lower than amplitudes of foot response.

Abstract AB0924 Table 1. Sympathetic skin responses

SSR	JSS mean latency	JSS mean amplitude	JSLE mean latency	JSLE mean amplitude
HAND	1424,63	0,75	1353,26	0,9
FOOT	1935,26	0,55	2011,6	**2,6**

According to BR, R1, R2, R2K duration and latency were not different between right and left eyes in both patients' groups. R3 was absent in 2 (10%) bilaterally and in 1 (5%) JSS patients unilaterally (right). R3 was absent in 4 (22.2%) bilaterally and in 2 (11%) JSLE patients unilaterally. Absence of R3 was more prominenet in JSLE patients, comparing to JSS. (Table 2)

Abstract AB0924 Table 2. Blink reflex responses

	JSS-right	JSS-left	JSLE-right	JSLE-left
R1 duration	5,9	5,8	6,7	6,5
R1 latency	10,3	10,3	10,3	10,2
R2 duration	33,5	33,7	32,7	29,1
R2 latency	30,3	29,8	29,6	30,5
R2K duration	34,4	29,4	34,7	32,2
R2K latency	32,5	33,3	31,9	33,1
R3 presence	**3/19**	**2/19**	**4/18**	**6/18**

Conclusion: BR could be considered as a potential indicator of neuropathy in JSLE and JSS patients. Data on SSR need to be evaluated in studies with higher number of patients with juvenile-onset connective tissue disorders.

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Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2019-eular.6580

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