Table 1. Laboratory results of the FMF and the control group

Variables	FMF patients (n=54)	Control (n=28)	P value
Omentin, pg/mL	108.05(19.97-343.22)	199.5(42.98-339.41)	0.03
SAA, ng/mL	3.7 (1.18-22.75)	1.31(0.95-3.16)	<0.001
ESR, mm/h	15(2-68)	12(7-17)	<0.001
CRP, mg/L	12(1-194)	2.5(1-8)	<0.001

Variables were given as median (IQR).

Calculated using Mann-Whitney U test for non-normal distribution.

FMF=Familial Mediterranean fever, SAA=serum amyloid A, ESR=erythrocyte sedimentation rate, CRP=C-reactive protein, IQR=interquartile range.

Table 2. Laboratory results of FMF patients with and without colchicine resistance

Variables	With resistance (n=16)	Without resistance (n=38)	P value
Omentin, pg/mL	76.64(19.77-224.33)	186.47(28.41-343.21)	0.006
SAA, ng/mL	3.69(1.18-22.75)	3.77(1.18-21.49)	0.784
ESR, mm/h	25.5(2-68)	15(2-60)	0.835
CRP, mg/L	11(1-67)	19(1-194)	0.111

Variables were given as median (IQR).

Calculated using Mann-Whitney U test for non-normal distribution.

FMF=Familial Mediterranean fever, SAA=serum amyloid A, ESR=erythrocyte sedimentation rate, CRP=C-reactive protein, IQR=interquartile range.FOM

Conclusion: FMF patients with colchicine resistance are associated with decreased omentin concentrations, probably mediated by inflammation-driven mechanisms.

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SAT0514 FATIGUE IN FAMILIAL MEDITERRANEAN FEVER

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Background: Fatigue is an important and common symptom in rheumatologic diseases. It causes disability and worsens patients quality of life. Familial Mediterranean Fever (FMF) is a hereditary autoinflammatory disease characterized by recurrent episodes of fever and serositis. Mutations in the MEFV gene that encodes pyrin protein are responsible for the disease. Most frequent mutation is M694V and FMF patients with M694V/M694V genotype have more severe disease.

Objectives: The aim of this study is to investigate fatigue and its impact on quality of life of FMF patients who are attack-free for more than one year.

Methods: Seventy-seven FMF patients and 70 age and sex matched healthy controls were enrolled in the study. Fatigue severity scale (FSS) was used to evaluate fatigue level. Disease severity was evaluated via FMF disease severity score. Short form-36 (SF-36) was used to evaluate the quality of life of the participants and Beck depression inventory (BDI) was used to evaluate depression.

Results: No statistically significant difference was found when FMF group and healthy controls were compared for demographic variables and laboratory markers (complete blood count, erythrocyte sedimentation rate, c-reactive protein levels, liver and kidney function tests). All of the subscale scores of SF-36 were lower in FMF group when compared with the control group. BDI and FSS scores were higher in the FMF group.(Table 1)

Table 2. Correlation of fatigue, depression, disease severity and duration with quality of life in FMF patients

	Physical functioning	Social functioning	Role Physical	Role emotional	Mental health	Vitality	Pain	General health
FMF disease severity scale	r= -0,223	r= -0,319**	r= -0,313**	r= -0,184	r= -0,124	r= -0,188	r= -0,191	r= -0,351**
Beck Depression Inventory	r= -0,418**	r= -0,554**	r= -0,461**	r= -0,470**	r= -0,660**	r= -0,594**	r= -0,308**	r= -0,576**
Fatigue severity scale	r= -0,391**	r= -0,426**	r= -0,475**	r= -0,324**	r= -0,650**	r= -0,739**	r= -0,410**	r= -0,521**
Disease duration	r= -0,40**	r= -0,218	r= -0,232*	r= -0,287*	r= -0,026	r= -0,211	r= -0,189	r= -0,109

** p<0.01

*P<0.05

Table 1. Comparison of FMF and control groups for fatigue, depression and quality of life

	FMF group (n: 77) median(min-max)	Control group (n: 70) Median (min-max)	p
Fatigue severity scale (FSS)	31(27-47)	28 (24-37)	p<0.05
Beck depression inventory (BDI)	15 (0-35)	12 (0-19)	p<0.05
SF-36 Physical functioning	80.00(25.00-100.00)	95.00 (50.00-100.00)	p<0.01
SF-36 Social functioning	55.50 (40.00-88.80)	77.70 (22.20-88.80)	p<0.01
SF-36 Role physical	25.00 (0.00-100.00)	100.00 (0.00-100.00)	p<0.01
SF-36 Role emotional	33.30 (0.00-100.00)	100.00 (0.00-100.00)	p<0.01
SF-36 Mental health	56.00 (24.00-88.00)	72.00 (24.00-100.00)	p<0.01
SF-36 Pain	55.50 (0.00-100.00)	77.70 (33.30-100.00)	p<0.01
SF-36 Vitality	45.00 (15.00-95.00)	65.00 (5.00-100.00)	p<0.01
Sf-36 General health	40.00 (5.00-92.00)	67.00 (20.00-100.00)	p<0.01

Twenty-three of FMF patients had M694V/M694V genotype and those patients had higher disease severity scores (p<0.01), higher FSS (p<0.01) and higher BDI scores (p<0.05) when compared with other FMF patients. Regarding the quality of life, patients with M694V/M694V genotype had lower scores in social functioning, role physical, role emotional and pain subscales of SF-36. In the correlation analysis depression and fatigue were found to be the major determinants of quality of life in FMF. Disease severity or duration were not strongly correlated with the SF-36 scores (Table 2).

Conclusion: Despite being attack-free for more than one year, FMF patients had poor quality of life and fatigue when compared with the healthy controls. The quality of life in FMF patients, whose attacks are well controlled, is mainly determined by fatigue and depression rather than disease severity.

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SAT0515 COMPLEX HYPERMOBILITY EHLERS-DANLOS SYNDROME (HEDS): MAPPING THE PATIENT'S JOURNEY OVER 40 MONTHS IN A TERTIARY REFERRAL CENTRE

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Background: Ehlers-Danlos Syndromes are heritable connective tissue disorders. They are multisystemic and patients can present with several symptoms such as joint pain and instability, visceral and autonomic dysfunction, as well as significant psychosocial sequelae. Managing this cohort of young patients is usually challenging as many patients present late due to delayed diagnosis, often with several complications, problems with mobility and opioid use. Furthermore, there is often a prolonged lack of coordinated healthcare and access to social care services. A recent parliamentary debate in the U.K. highlighted that hEDS services are excluded from specialist Rheumatology commissioning services. In order to ascertain the relevance and utility of specialist services in this population, we conducted this study.

Objectives: The objective of this study was to map the patient experience following a referral to the specialist clinic in order to assess the need for an integrated, multidisciplinary approach to treating patients with hypermobility EDS.

Methods: We retrospectively reviewed the records of 50 patients with the diagnosis of hypermobility EDS who were seen in a specialist hypermobility clinic at University College Hospital UCLH between January 2016 and March 2016. Relevant data was collected regarding their medical care in our hospital up to October 2019.

Results: The median age was 37 (range 21-59). We had 10 males and 40 females. The diagnosis of hypermobility EDS was based on the 1997 criteria as these patients were seen prior to the 2017 classification. Overall, the study yielded 6 key themes: 1. All patients experienced chronic pain, with 36% reporting use of opioids for pain management. 2. Patients were referred to multiple

medical specialities within the same hospital trust, (22% patients were referred to ≥ 5 specialities). 3. Patients required a high number of follow up appointments (28% of patients required ≥ 20 follow-up visits). 4. Failed discharges were common; patients were often referred back to the Rheumatology Clinic despite being discharged to primary care. 5. Patients had a significant number of comorbidities, reflected by polypharmacy. (36% of patients were prescribed ≥ 5 medications). 6. Disability was high (20% of patients reported severe mobility problems).

Conclusion: This study shows that patients with hEDS referred to UCLH have significant levels of disability, opioids use and polypharmacy especially for a relatively young population of patients. They need a complex interdisciplinary approach in a timely manner. In order to minimise delays and allow earlier diagnosis and intervention, we have recently adopted a multidisciplinary team approach, including pain specialists, rheumatologists, psychologists, physiotherapists, nurse specialists, urogynaecologists and neurogastroenterologists. This allows more coordinated and efficient care and incorporates an EDS-specific pain management programme. Specialised services for complex hEDS cases should be established and adequately resourced. Moreover, it would be cost effective to commission a patient-centred "one-stop-shop" service, where patients, who often travel from long distances with severe disabilities, can be seen by multiple specialities in a single visit.

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SAT0516

CLINICAL CHARACTERISTICS AND PROGNOSTIC FACTORS IN PATIENTS WITH SECONDARY HEMOPHAGOCYTIC SYNDROME

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Background: The Hemophagocytic Syndrome (HPS) had a mortality rate between 20% and 90%. The mortality of HPS secondary to autoimmune diseases (AID) is lower than hemato-oncological diseases (HOD). In general, the HOD, thrombocytopenia, age, and a prolongation of prothrombin are considered to be an adverse prognostic factor.(1)

Objectives: To describe and identify differences between patients who survived and did not survive to HPS during hospital admission to a tertiary hospital between 2005 and 2019.

Methods: This is a retrospective observational study. All patients who met the diagnostic criteria for LHH were included, or who presented haemophagocytic cells in the bone marrow biopsy, or who had diagnosis of HPS in the hospital discharge report.(2) Demographic, clinical, analytical, etiological, underlying disorder and prognosis variables were collected. Continuous variables are described with the mean or median according to the degree of normality. Kruskal Wallis, Fisher test and Mann-Whitney U test were used for the bivariate analysis, and also a multivariate logistic regression analysis was performed.

Results: Thirty patients with HPS were included. They were distributed in 5 subgroups (Table 1). Overall mortality was 43.3%, statistically significant higher in the HOD [8 patients (66.7%); $p = 0.029$]. Also, they were divided into 2 groups (survivor vs. non-survivor; Table 2). In the multivariate model the age and INR prolongation were confirmed to be independently associated with the outcome of mortality.

Table 1. Etiology of HPS

Etiology	n = 30	Mortality
AID	10	n = 1
Systemic lupus erythematosus	5	1
Adult Still's Disease	3	No
Rheumatoid arthritis	1	No
Sclerosing Disease IgG4	1	No
HOD	12	n = 8*
Non-Hodgkin's lymphoma	3	1

Table 1. Etiology of HPS

Etiology	n = 30	Mortality
Myelodysplastic syndrome	3	2
Acute leukemia	3	3
Extranodal NK cell lymphoma	1	1
Multiple Myeloma	1	No
Probable lymphoproliferative process	1	1
Infectious diseases	2	n = 1
<i>Pneumocystis carinii</i> in patient with H.I.V.	1	1
<i>Campylobacter jejuni</i>	1	No
Glyoblastoma multiforme with temozolomida	1	n = 0
HPS without defined aetiology	5	3

HIV: Human Immunodeficiency Virus, NK: Natural Killer. * $p = 0,029$

Table 2. Characteristics and differences between survivor and non-survivor groups

	Total	Non-survivor	survivor		
n	30	13	17	p<0,05	
Age	55,5 ±18,3	68	58,2-74,5	40	34-57 0,043
Women	16 53,3%	7	61,5%	9	47,1% 1,00
Comorbidities (≥ 2)	5 16,7%	2	15,4%	3	17,6% 1,000
Hospital stay	35,5 20-60,8	29	15,5-39	13	8-17 0,563
Splenomegaly	16 53,3%	7	53,8%	9	52,9% 1,000
Hepatomegaly	10 33,3%	5	38,5%	5	29,4% 0,705
Hb (g/dL)	7,1 6,4-7,9	710%	6,2-7,8	7,1	6,6-7,8 0,094
Pt ($\times 10^9/L$)	13 500 5 000-52 500	16 000	11 000-44 000	12 000	5 000-99 000 0,281
Pt $\leq 100 000$	25 83,3%	13	100%	12	70,6% 0,052
Leu ($\times 10^9/L$)	1 250 238-3 153	1 300	150-3 940	1 400	200-3 340 0,457
Neu ($\times 10^9/L$)	615 0-1 550	1 290	20-3 300	650	0-1 400 0,805
Fb (mg/dL) (n=24)	171 111-358	167,00	106-253	169,00	103-451 0,796
Fer (ng/mL) (n=28)	15 330 5 434-38 284	29 063	5 728-74 604	13 225	8 287-28 729 0,108
Tg (mmol/L)	341 226-438	254,00	184-382	471,00	341-604 0,053
GOT (U/L)	139 77,5-406,3	133,00	101-513	179,00	101-512,5 0,483
GPT (U/L)	162 46-389	109,00	41-333	199,00	99-298 0,198
INR (n=29)	1,5 1,1-1,9	2,1	1,2-3,7	1,5	1,1-1,6 0,028

Hb: Hemoglobin, Pt: platelets, Leu: leukocytes, Neu: neutrophils, Fb: fibrinogen, Fer: ferritin, Tg: triglycerides, GOT: aspartate aminotransferase, GPT: alanine aminotransferase

Conclusion: The HOD presented higher mortality. The non-survivor group presented a longer INR prolongation and a higher age at the time of diagnosis.

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SAT0518

CANAKINUMAB TREATMENT IN ADULT PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER: A SINGLE-CENTER STUDY

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Background: Familial Mediterranean Fever (FMF) is the most common auto-inflammatory disease characterized by recurrent, self-limiting attacks of fever, serositis, arthritis, and erysipelas-like erythema. Canakinumab is an Interleukin-1 β inhibitor that is shown to be effective and safe in treating colchicine resistant FMF patients.

Objectives: The main objective of this study is to present the single tertiary center experience of adult FMF patients who received Canakinumab.

Methods: The study is a retrospective analysis conducted at a tertiary rheumatology center experienced in FMF. The patients who had a clinical diagnosis of FMF and who were treated with at least a single subcutaneous injection of canakinumab were