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
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**
Pneumocystis carinii in patient with H.I.V.	1	1
Campylobacter yeyuni	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 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**
Pneumocystis carinii in patient with H.I.V.	1	1
Campylobacter yeyuni	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 (x10^9/L)	13 500	5 000-52 500	16 000	11 000-44 000	12 000	5 000-99 000	0,281
Pt ≤ 100 000	25	83,3%	13	100%	12	70,6%	**0,052**
Leu (x10^9/L)	1 250	238-3 153	1 300	150-3 940	1 400	200-3 340	0,457
Neu (x10^9/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 autoinflammatory disease characterized by recurrent, self-remitting attacks of fever, serositis, arthritis, and erysipelas-like erythema. Canakinumab is an Interleukin-1β 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