

Soluble interleukin-2 receptor is a sensitive diagnostic test in adult HLH: Supplemental Data

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Table 1. Characteristics of Non- HLH patients (N = 40)

Disease	No. patients
Autoimmune disorder	8
Sepsis	6
Unknown	5
Histiocyte disorder	4
Lymphoma	3
Acute hepatitis	3
Acute leukemia	2
Other malignancy	1
IgG4 related disease	1
Myocarditis	1
Hemolytic anemia	1
TTP	1
Hb SC disease	1
Immunodeficiency	1
CAPS	1
Sarcoidosis	1
No. HLH criteria	No. patients
7/7	0
6/7	1
5/7	4
4/7	5
3/7	6
2/7	10
1/7	6
0/7	8

Table 2. Characteristics of HLH patients (N=38)

HLH subtype		No. patients
	MAHS	16
	IAHS	14
	Unknown	5
	MAS	3
No. HLH criteria*		No. patients
	7/7	7
	6/7	8
	5/7	21
	4/7	1**
	3/7	1**
	2/7	0
	1/7	0
	0/7	0

MAHS – malignancy associated hemophagocytic syndrome, IAHS – infection associated hemophagocytic syndrome, MAS – macrophage activation syndrome

*Because NK cell function is not available in our center most patients were evaluated on the basis of the seven available HLH 2004 criteria.

**Two patients were deemed by their treating Hematologist to have HPS/HLH despite having fewer than 5/7 HLH 2004 criteria. The patient with 4/7 criteria, EH21, was deemed to have HPS/HLH after presenting with fever, severe hepatitis, ferritin 10466 µg/L, sIL-2r 3784 U/mL, HScore 218, hemophagocytosis and metastatic adenocarcinoma (lung origin) in bone marrow. The patient with 3/7 criteria, H61, had disseminated tuberculosis in the context of inflammatory bowel disease, with hepatitis, ferritin 10466 µg/L, sIL-2r 3784 U/mL, HScore 200, which improved with steroids, IVIg and treatment of the tuberculosis.

Figure 1. Trends of sIL-2r and ferritin for HLH patients with multiple Measurements

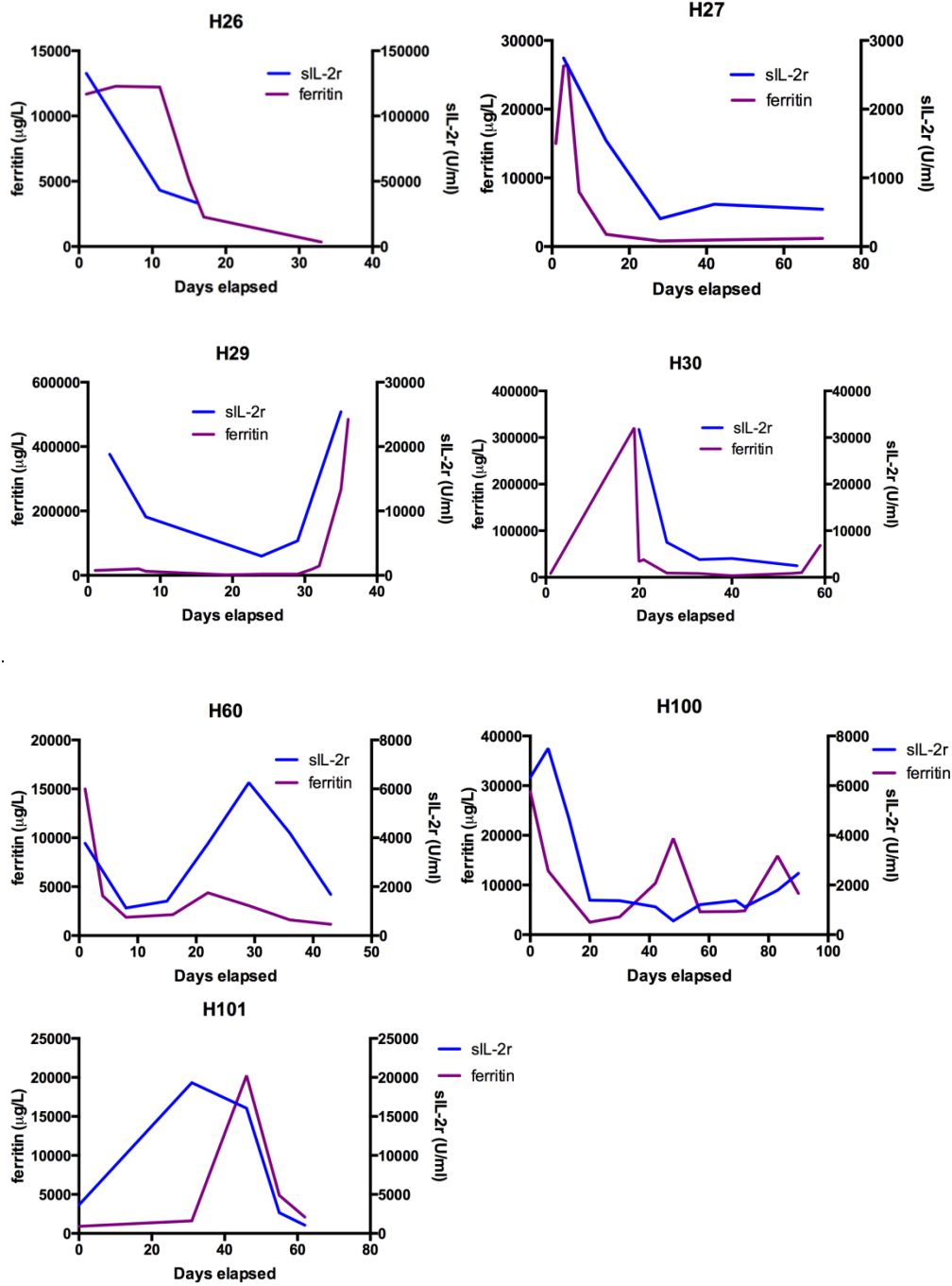


Table 3: HLH subtypes breakdown

	N	%	sIL2r mean	SD	ferritin mean	ferritin SD	sIL2r/ferritin ratio mean
IAHS	14	37%	9720.6	9081.9	78154.9	143048.7	0.50
MAHS	16	42%	20241.6	16991.8	58715.8	84460.9	1.41
MAS	3	8%	5007.7	1692.9	32728.7	1692.9	0.21
Unknown	5	13%	8788.2	3889.2	28982.0	27628.0	0.59