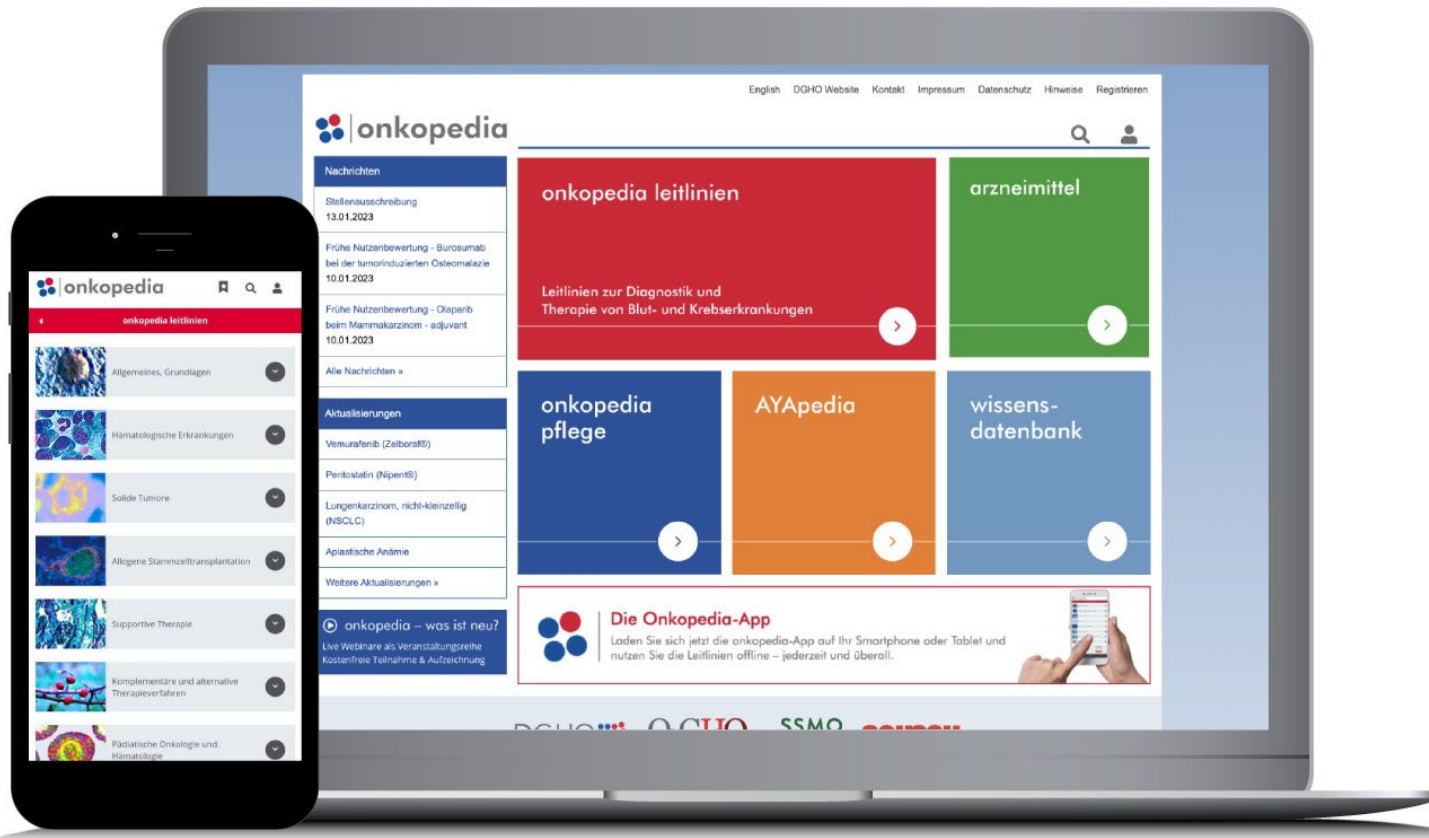


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HÄMATOLOGIE UND MEDIZINISCHE ONKOLOGIE

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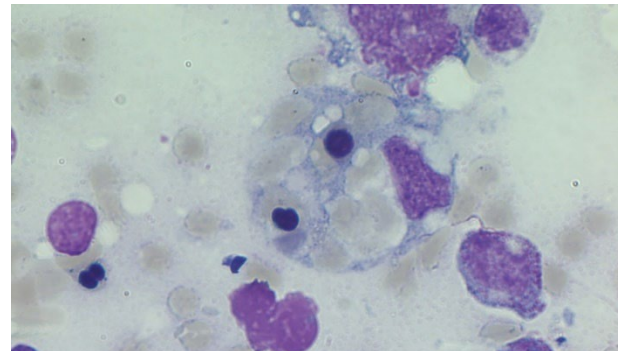
[www.dgho.de/mitglied-werden](http://www.dgho.de/mitglied-werden)

# Hämophagozytische Lymphohistiozytose (HLH)

**Paul Graf La Rosée**

**ONKOPEDIA – Online-Seminar**

**18.10.2024**



# Offenlegung Interessenskonflikte

## 1. Anstellungsverhältnis oder Führungsposition

- keine

## 2. Beratungs- bzw. Gutachtertätigkeit

- Novartis, SOBI,

## 3. Besitz von Geschäftsanteilen, Aktien oder Fonds

- keine

## 4. Patent, Urheberrecht, Verkaufslizenz

- keine

## 5. Honorare

- Novartis, SOBI

## 6. Finanzierung wissenschaftlicher Untersuchungen

- Novartis

## 7. Andere finanzielle Beziehungen

- keine

## 8. Immaterielle Interessenkonflikte

-keine

# Agenda

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Fallbeispiel

Epidemiologie

Biologie

Früherkennung / Prophylaxe

Klinisches Bild

Diagnostik

Therapie

Struktur (Algorithmus)

Stadien

Arzneimittel

Nachsorge und Rehabilitation

**Männlich, 19 Jahre**

13.1.22 FUO, B-Symptomatik, Einweisung Rheumatologie, DD Vasculitis?

18.1.22 FDG-PET-CT: Lymphadenopathie, Splenomegalie, Knochenläsionen

1 21.1.22 Knochenmark: Hämophagozytose, kein Hinweis auf Lymphom

24.1.22 500 mg Prednisolon i.v., Hydroxychloroquin, Ciclosporin

2 28.1.22 Lymphknoten-Biopsie: Unspezifische Lymphadenitis

31.1.22 IVIG (2 g/kg KG)

7.2.22 Verlegung Hämatologie SBK: **Ferritin 2744 ng/mL, sIL2R 11000 U/mL**

3 10.2.22 LK-Biopsie: Hämophagozytose

4 11.2.22 Knochenbiopsie: Hämophagozytose

5 18.2.22 LK-Biopsie: **T-NHL (ALCL),**

20.2.22 ICU, IVIG 20 g, Dexamethason 20 mg/d

21.2.22 **Ferritin 10361 ng/mL, sIL2R 22300 U/mL**, Etoposid 200 mg, IVIG 30 g

6 22.2.22 Hautbiopsie: unspezifische Inflammation; **Ferritin 10361 ng/mL, sIL2R 22300 U/mL**

**sCD25 : Ferritin**

**2 : 1**

**Lymphom**

6 x CHOEP

23.05.-13.06. BEAM, AutoPBSCT

**Lymphom-assoziierte HLH**

## Weiblich, 67Jahre

### Akute myeloische Leukämie IDH2+ (AML) ED 10/21

#### Therapie & Verlauf:

- 4.11.21 - Induktionschemotherapie + Enasidenib
- 18.11.21 - Neutropene Enterocolitis IV° in der Aplasie  
**-Sepsis**

			18.11.21
Ferritin	ng/ml	15-160	1782 +
Transferrinsättigung			
anorg. Phosphat	mmol/l	0.8-1.45	0.76
Magnesium	mmol/l	0.75-1.10	0.70 -
C-reaktives Protein	mg/l	<5	372.08 ++
Procalcitonin	ng/ml	< 0.5	33.41 +
Interleukin-6	pg/ml	<15	811.6 +

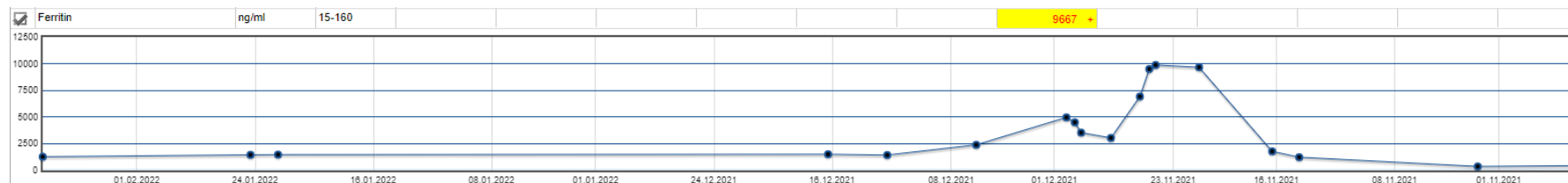
### 22.11.21 Sepsis mit assoziiertem Makrophagenaktivierungssyndrom (MAS-like Sepsis)

- Anakinra 23.11.-25.11.2021
- Dexamethason 23.11.- 27.11.2021
- Immunglobuline vom 23.11.-25.11.

		27.11.21	22.11.21
Löslicher Interleukin-2-Rezeptor	U/ml	158 - 623	3637 +
			7443 +

- 12/21-05/22 5 x Azacytidin 75mg/m2 d1 -5
- 9.6.22 - Allogene SZT
- 08/23 - Komplette Remission, Nachsorge

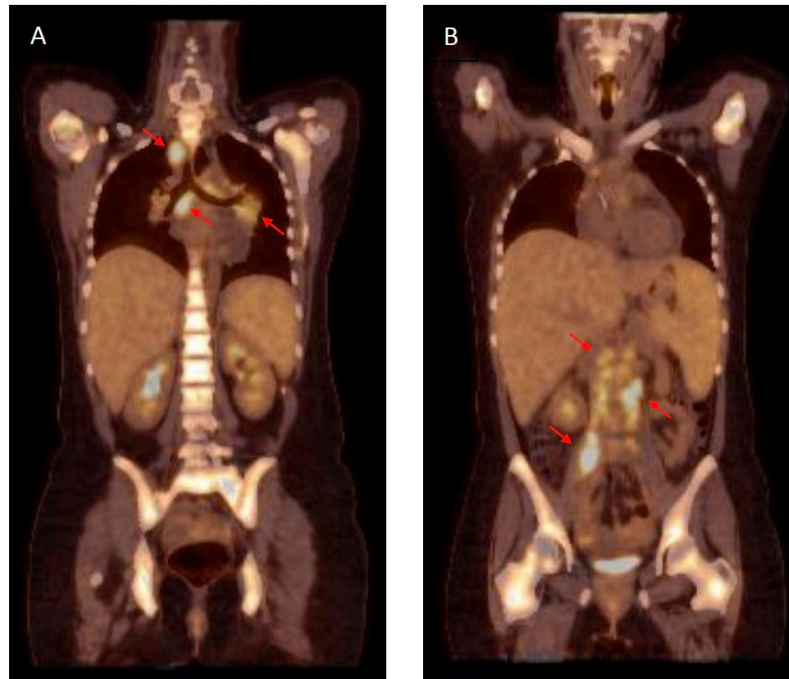
#### Ferritin Peak



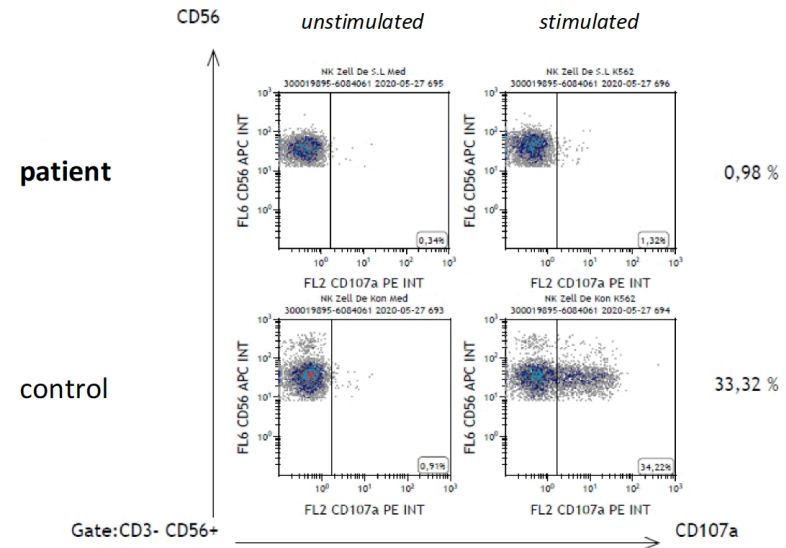
**Sepsis mit nachfolgender MAS-like Sepsis bei KM-Aplasie durch Induktionschemotherapie der AML**

Weiblich, 29 Jahre

# Mycobacterium avium, HLH



Allo-SCT 11/11 MUD



GATA2 c.177C>G; p.Tyr59\*

Infektions-getriggerte HLH bei Immundefektsyndrom



## Previously undetected lymphoma led to ESPN reporter's death, fiancée reveals



Edward Aschoff had stage 4 non-Hodgkin lymphoma in his lungs. It was found during his autopsy. (Source: University of Florida/CNN)

Updated: Thu 11:22 AM, Jan 16, 2020



(CNN) - It was a big shock for college football fans when Edward Aschoff, a reporter for ESPN, died on Christmas Eve, his 34th birthday.

Aschoff's fiancée said after his autopsy, they found out he had stage 4 non-Hodgkin lymphoma in his lungs. It's an aggressive kind of cancer that can be undetectable until it's very advanced.

When Aschoff died, he'd been diagnosed with multi-focal pneumonia and hemophagocytic lymphohistiocytosis, or HLH, which affects the immune system.

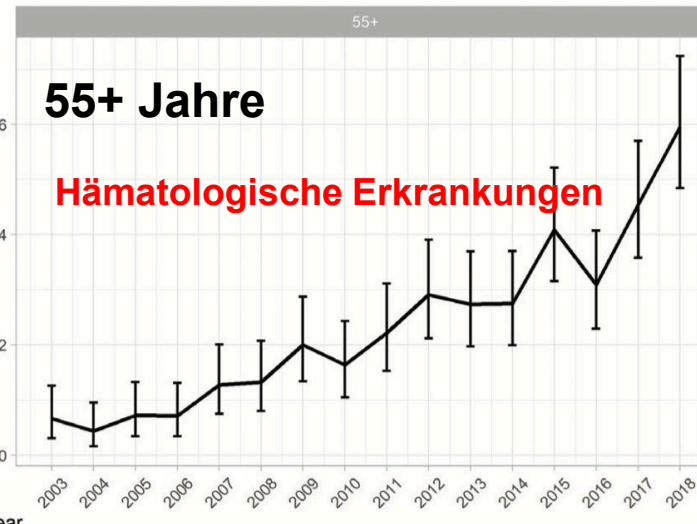
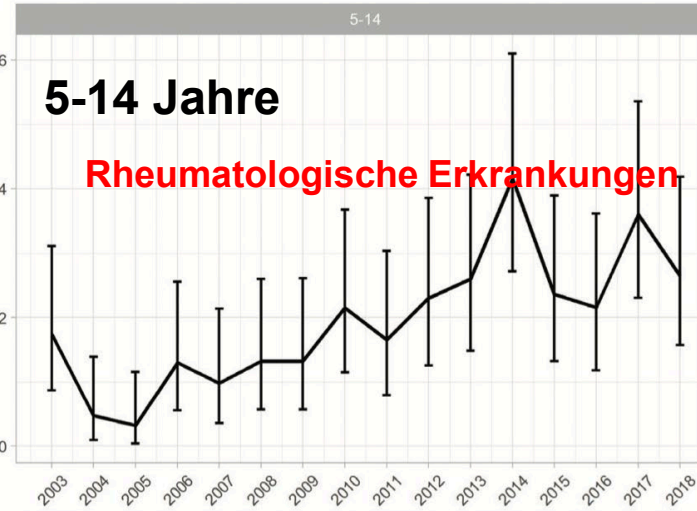
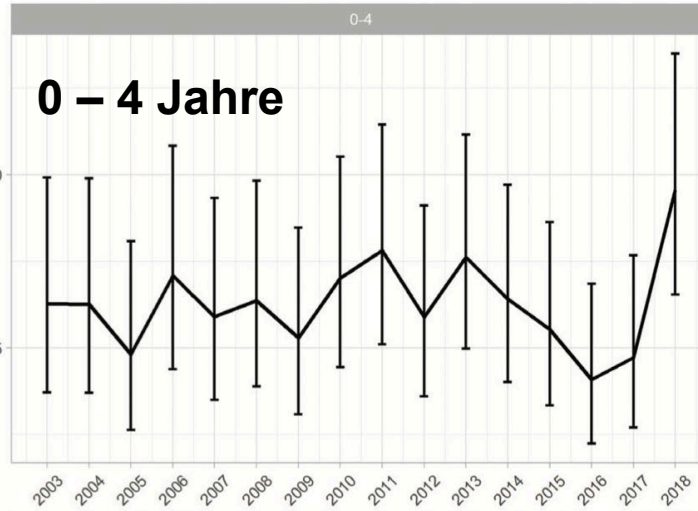
## Lymphom-assoziierte HLH (LA-HLH)



## AOSD und MAS-HLH

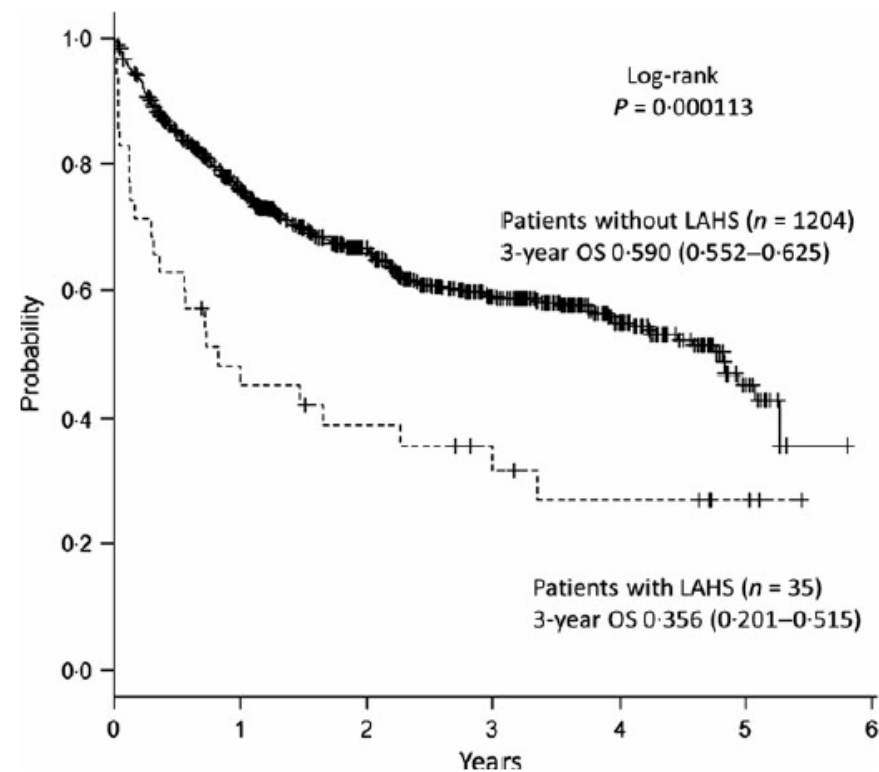


Inzidenzrate/Million Population



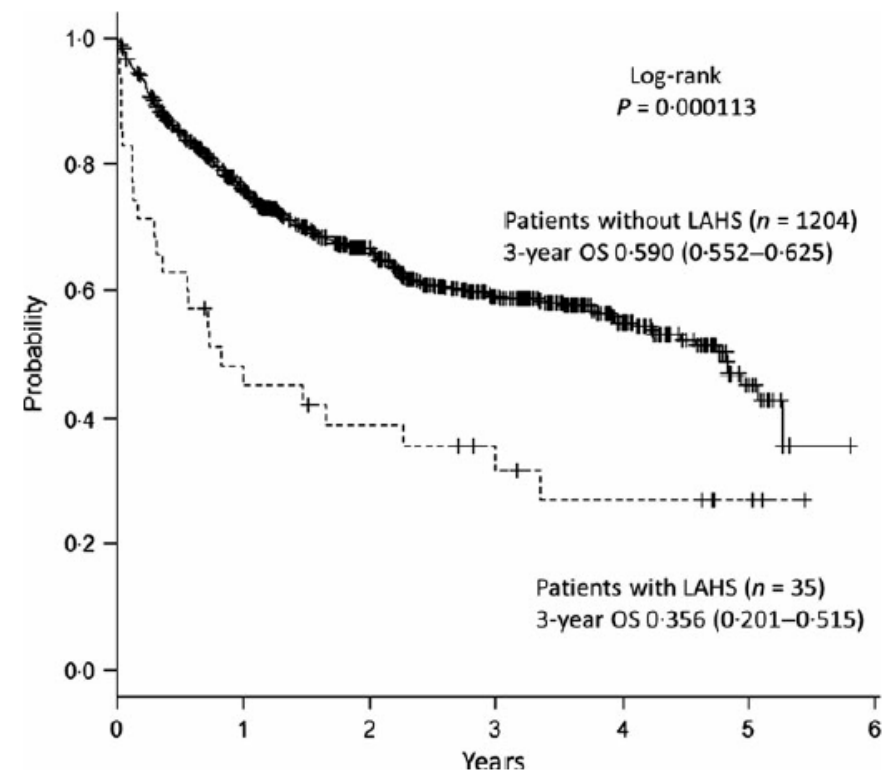
**12 Institutionen**  
**N = 1239 Lymphompat.**  
**LA-HLH: 35/1239 (2,8%)**

	LAHS (%)	non-LAHS
<b>A</b>		
DLBCL*	18 (2.7)	679
IVL	5 (38.5)	8
PCNSL	0	9
NMZBCL	1 (33.3)	2
FL	0	213
MALT lymphoma	0	32
MCL	0	31
BL	0	28
LPL	0	14
B-LBL	0	12
SMZL	0	8
SLL	0	3
HCL	0	1
LYG	0	1
<b>B</b>		
PTCL-NOS	6 (13.3)	39
ENKL	3 (15.8)	16
ANKL	3 (30.0)	7
AITL	2 (5.9)	32
ALCL	1 (4.8)	20
T-LBL	1 (14.3)	6
ATL	0	58
HSTCL	0	1
MF	0	1



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HSTCL	0	1
MF	0	1



## Hemophagocytic syndrome in patients with acute myeloid leukemia undergoing intensive chemotherapy

Karen Delavigne,<sup>1,6</sup> Emilie Bérard,<sup>2,3</sup> Sarah Bertoli,<sup>1</sup> Jill Corre,<sup>4</sup> Eliane Duchayne,<sup>4</sup> Cécile Demur,<sup>4</sup> Véronique Mansat-De Mas,<sup>4,6</sup> Cécile Borel,<sup>1,6</sup> Muriel Picard,<sup>1</sup> Muriel Alvarez,<sup>5</sup> Audrey Sarry,<sup>1</sup> Françoise Huguet,<sup>1</sup> and Christian Récher<sup>1,6</sup>

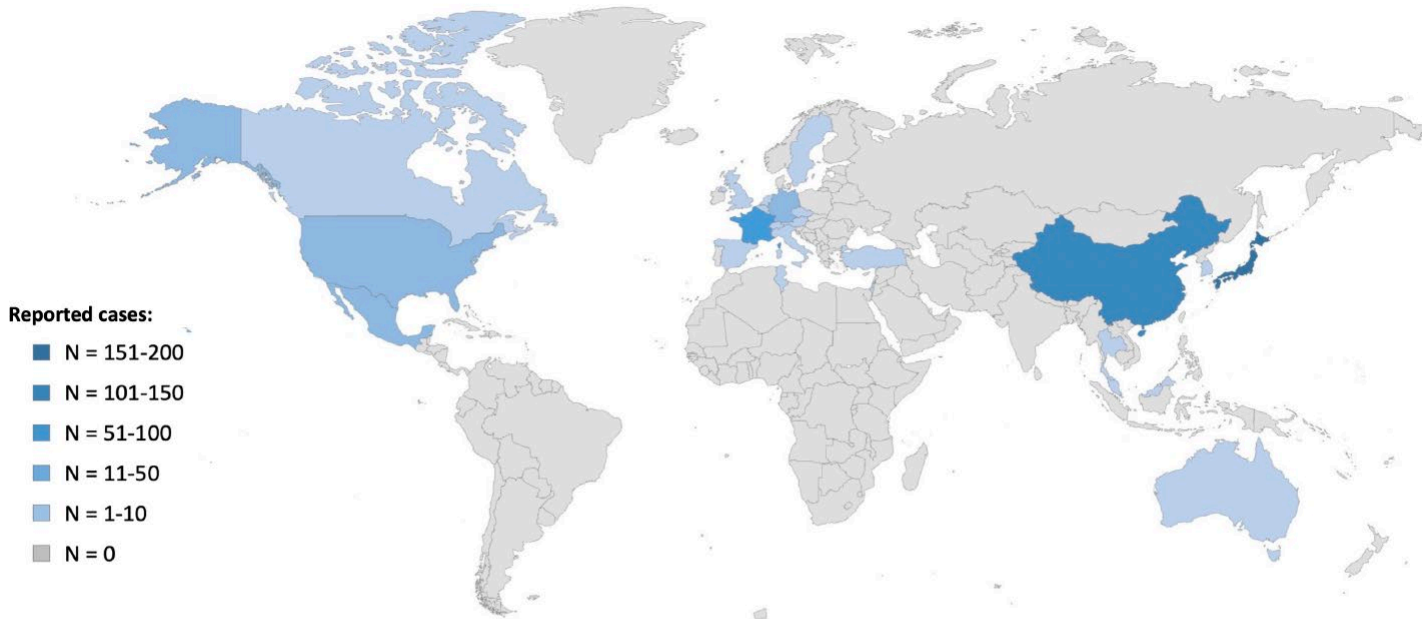
**AML, n = 343 (5 Jahre retrospektiv, 7 + 3 Induction)**

- Antibiotika-resistentes Fieber
- Rasch dynamischer Ferritin-Anstieg
- Hämophagozytose i. KM

**n = 32 (9,3%)**

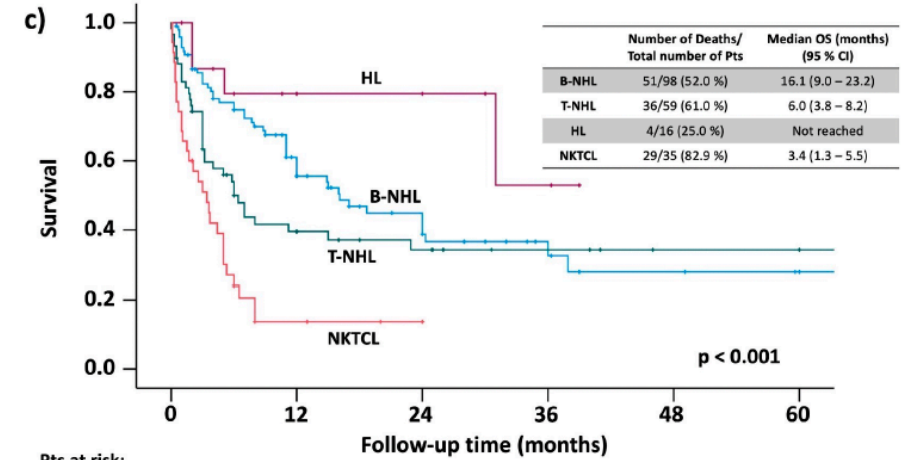
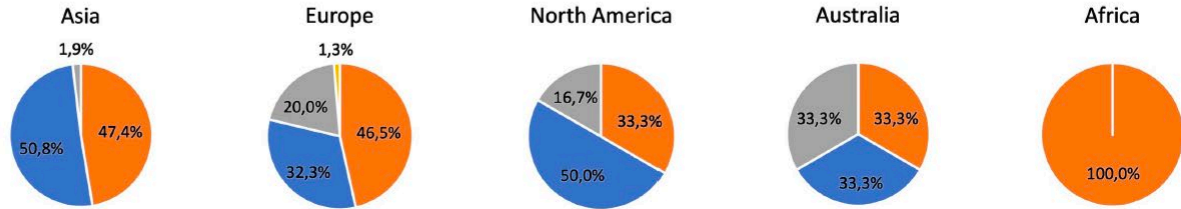
**Ferritin-Max (median):      HLH+      Hämophagozytose + / HLH-**

	<b>6953</b> (4425-15305)	<b>3373</b> (1808-5149)	<b>p 0.0071</b>
> 500, n. (%)	32 (100)	17 (94.4)	0.3600
> 3000, n. (%)	26 (81.3)	11 (61.1)	0.1797
→ > 8000, n. (%)	15 (46.9)	2 (11.1)	0.0104
> 10000, n. (%)	13 (40.6)	2 (11.1)	0.0287



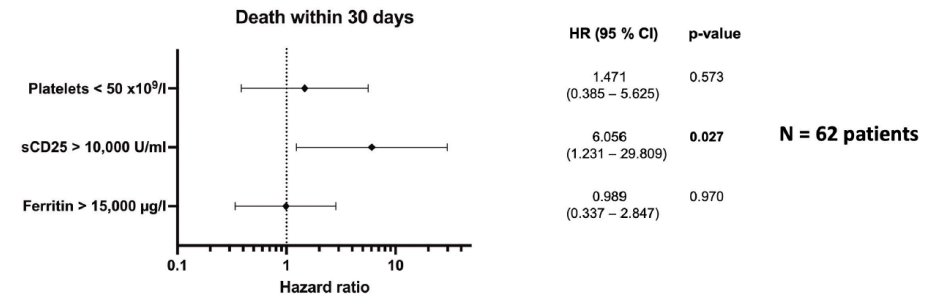
**Lymphoma subgroups:**

- B-NHL (N = 247)
- T-NHL (N = 245)
- HL (N = 48)
- Unspecified (N = 2)



**Pts at risk:**

	0	12	24	36	48	60
B-NHL	98	45	22	9	4	2
T-NHL	59	19	12	7	4	4
HL	16	7	5	2	0	0
NKTL	35	3	1	0	0	0



## Secondary HLH (apparently non-Mendelian HLH)

Infection-associated HLH
Virus-associated HLH
EBV-associated HLH
CMV-associated HLH
HLH associated with other defined herpes virus infections
HIV-associated HLH
Influenza-associated HLH
HLH associated with other defined virus infections
Bacteria-associated HLH*
Parasite-associated HLH*
Fungal-associated HLH*
Malignancy-associated HLH
Malignancy-triggered HLH (HLH at onset of malignancy)
Hematological malignancies
T-cell lymphoblastic lymphoma/leukemia
T-cell non-lymphoblastic lymphomas
B-cell leukemias
B-cell lymphomas (non-Hodgkin)
Hodgkin lymphomas
NK-cell lymphomas/leukemias
Myeloid neoplasia
Other hematological malignancies
Solid tumors
Unclassified malignancies
HLH occurring during chemotherapy (not associated with initial diagnosis of malignancy)*
HLH associated with a malignancy, but not further defined
HLH associated with defined rheumatologic conditions (MAS-HLH, or MAS-HLH*)
HLH associated with SoJIA
HLH associated with adult-onset Still disease
HLH associated with SLE
HLH associated with vasculitis
HLH associated with other defined autoimmune conditions
HLH associated with a not defined autoimmune condition
Transplant-related HLH*
HLH associated with iatrogenic immune activation*
HLH associated with iatrogenic immune suppression
HLH associated with other apparently non-Mendelian conditions
HLH of unknown/uncertain origin

## Erworbene HLH, Erwachsenenalter



## Hereditäre HLH (kindlich, jugendlich, selten late onset im Erwachsenenalter)

### Primary HLH: Mendelian inherited conditions leading to HLH

HLH associated with lymphocyte cytotoxic defects

FHL2 (*PRF1*)

FHL3 (*UNC13D*)

FHL4 (*STX11*)

FHL5 (*STXBP2*)

XLP1 (*SH2D1A*)

Griscelli Syndrome type 2 (*RAB27A*)

Chediak-Higashi Syndrome (*LYST*)

HLH associated with abnormalities of inflammasome activation

XLP2 (*BIRC4*)

*NLR4*

HLH associated with defined Mendelian disorders affecting inflammation

Lysinuric protein intolerance (*SLC7A7*)

*HMOX1*

Other defined Mendelian disorders affecting inflammation

Familial (apparently Mendelian) HLH of unknown origin

Bei positiver Familienanamnese, genetisch testen

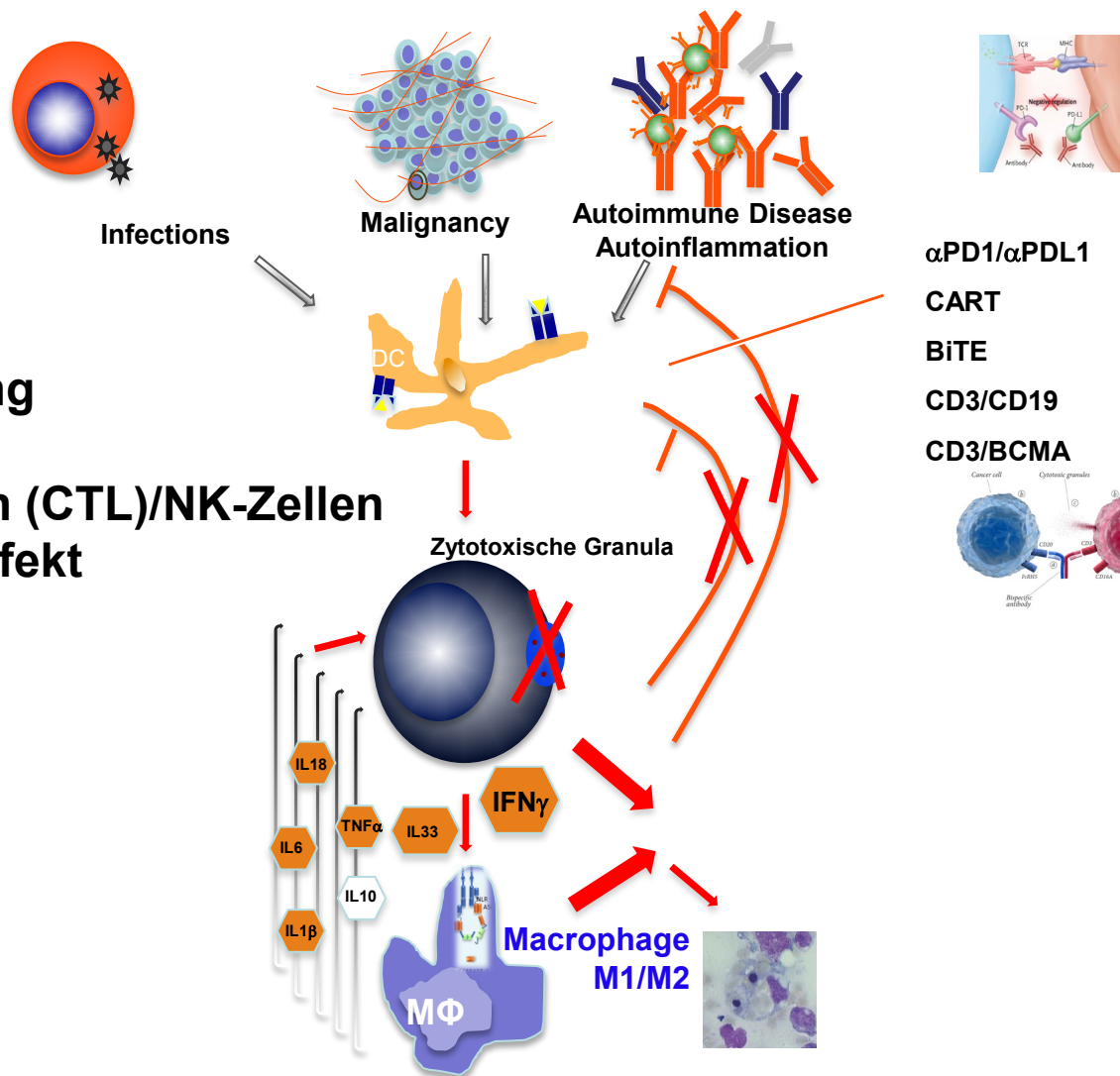
Bei Erwachsenen den optimierten HLH-Index nutzen (OHI)

Anamnese, Anamnese, Anamnese

Ans Bett!

Diagnostik wiederholen bei laufender Therapie





Dendritische Zellen  
Antigen Präsentation

Zytotoxische T-Zellen (CTL)/NK-Zellen  
mit zytolytischem Defekt

Zytokin Sturm

Myeloid derived  
cytokines

Inflammasome  
activation

### Immune Effector Cell-Associated HLH-Like Syndrome

$\alpha$ PD1/ $\alpha$ PDL1

CART

BiTE

CD3/CD19

CD3/BCMA

sIL2R (sCD25) ↑

Ferritin ↑

# HLH-2004

1. Fever
2. Splenomegaly
3. Cytopenia (2 or 3 lineage)  
Hemoglobin  $<9$  g/dl  
Platelets  $<100 \times 10^9/l$   
  
Neutrophils  $<1 \times 10^9/l$
4. Hyperferritinemia  $>500$  ng/ml
5. (a) Hypertriglyceridemia  $>265$  mg/dl  
or  
(b) Hypofibrinogenemia  $<150$  mg/dl
6. Hemophagocytosis
7. Low or absent NK-cell activity
8. Elevation of sCD25 (sIL-2 receptor)  
 $>2400$  U/ml

## H-Score

Known underlying immunodepression	No	▼
Maximal Temperature (C)	Between 38.4 and 39.4	▼
Hepatomegaly	Yes	▼
Splenomegaly	Yes	▼
Lower Hemoglobin level	less than or equal to 9.2 g/dl	▼
Lower Leucocytes count	less than or equal to 5000/mn	▼
Lower Platelets count	less than or equal to 110000/l	▼
Higher Ferritin level (ng/ml)	Strictly greater than 6000	▼
Higher Triglyceride level (mmol/l)	Strictly greater than 4	▼
Lower Fibrinogen level (g/l)	less than or equal to 2.5	▼
Higher SGOT/ASAT level (UI/L)	Greater than or equal to 30	▼
Hemophagocytosis features on bone marrow aspirate	Yes	▼
Calculate		
HScore		303
Probability of having HS (%)		99.97742823651381

<http://saintantoine.aphp.fr/score/>

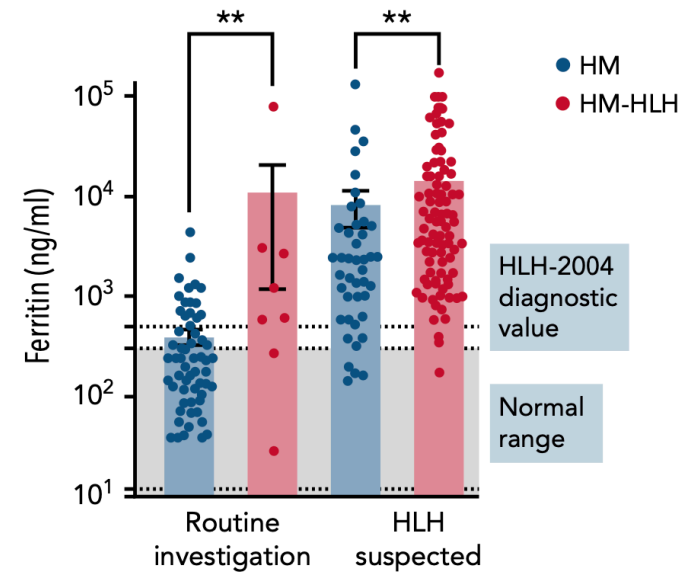
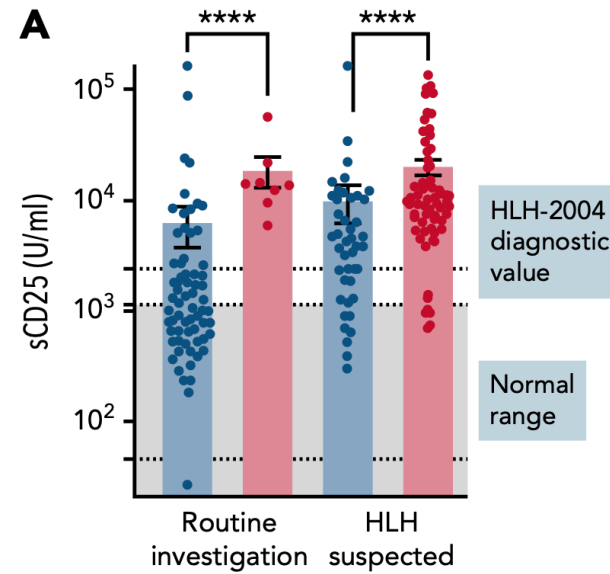
	HLH-1994 [3]	HLH-2004 [4] HLH 2016-2021 [5]	HLH-2024 [1]	OHI [8]
Clinical criteria	Fever Splenomegaly	Fever Splenomegaly	Fever Splenomegaly	Fever Splenomegaly
Laboratory criteria	Cytopenia in $\geq 2$ of 3 cell lines Hemoglobin $<90$ g/L Platelets $<100 \times 10^9$ /L Neutrophils $<1.0 \times 10^9$ /L Hypertriglyceridemia and/or Hypofibrinogenemia Fasting triglycerides $\geq 2$ mmol/L Fibrinogen $\leq 1.5$ g/L	Cytopenia in $\geq 2$ of 3 cell lines Hemoglobin $<90$ g/L Platelets $<100 \times 10^9$ /L Neutrophils $<1.0 \times 10^9$ /L Hypertriglyceridemia and/or Hypofibrinogenemia Fasting triglycerides $\geq 3$ mmol/L Fibrinogen $\leq 1.5$ g/L	Cytopenia in $\geq 2$ of 3 cell lines Hemoglobin $<90$ g/L Platelets $<100 \times 10^9$ /L Neutrophils $<1.0 \times 10^9$ /L Hypertriglyceridemia and/or Hypofibrinogenemia Fasting triglycerides $\geq 3$ mmol/L Fibrinogen $\leq 1.5$ g/L	Cytopenia in $\geq 2$ of 3 cell lines Hemoglobin $<90$ g/L Platelets $<100 \times 10^9$ /L Neutrophils $<1.0 \times 10^9$ /L Hypertriglyceridemia and/or Hypofibrinogenemia Fasting triglycerides $\geq 3$ mmol/L Fibrinogen $\leq 1.5$ g/L
Histopathologic criteria	Hemophagocytosis in spleen/bone marrow/lymph node no evidence of malignancy	Hemophagocytosis in spleen/bone marrow/lymph node no evidence of malignancy	Hemophagocytosis in spleen/bone marrow/lymph node no evidence of malignancy	Hemophagocytosis in spleen/bone marrow/lymph node
Inflammatory criteria		low or absent NK-cell activity  Ferritin $\geq 500$ $\mu\text{g/L}$ soluble CD25 $\geq 2400$ U/ml	Ferritin $\geq 500$ $\mu\text{g/L}$ soluble CD25 $\geq 2400$ U/ml	Ferritin $\geq 1000$ $\mu\text{g/L}$ soluble CD25 $\geq 3900$ U/ml
Overall survival	5y - 50%	2004: 5y - 59% 2016-2021: 3y - 79 %	n.a.	5y 10 – 30%*

CME Article

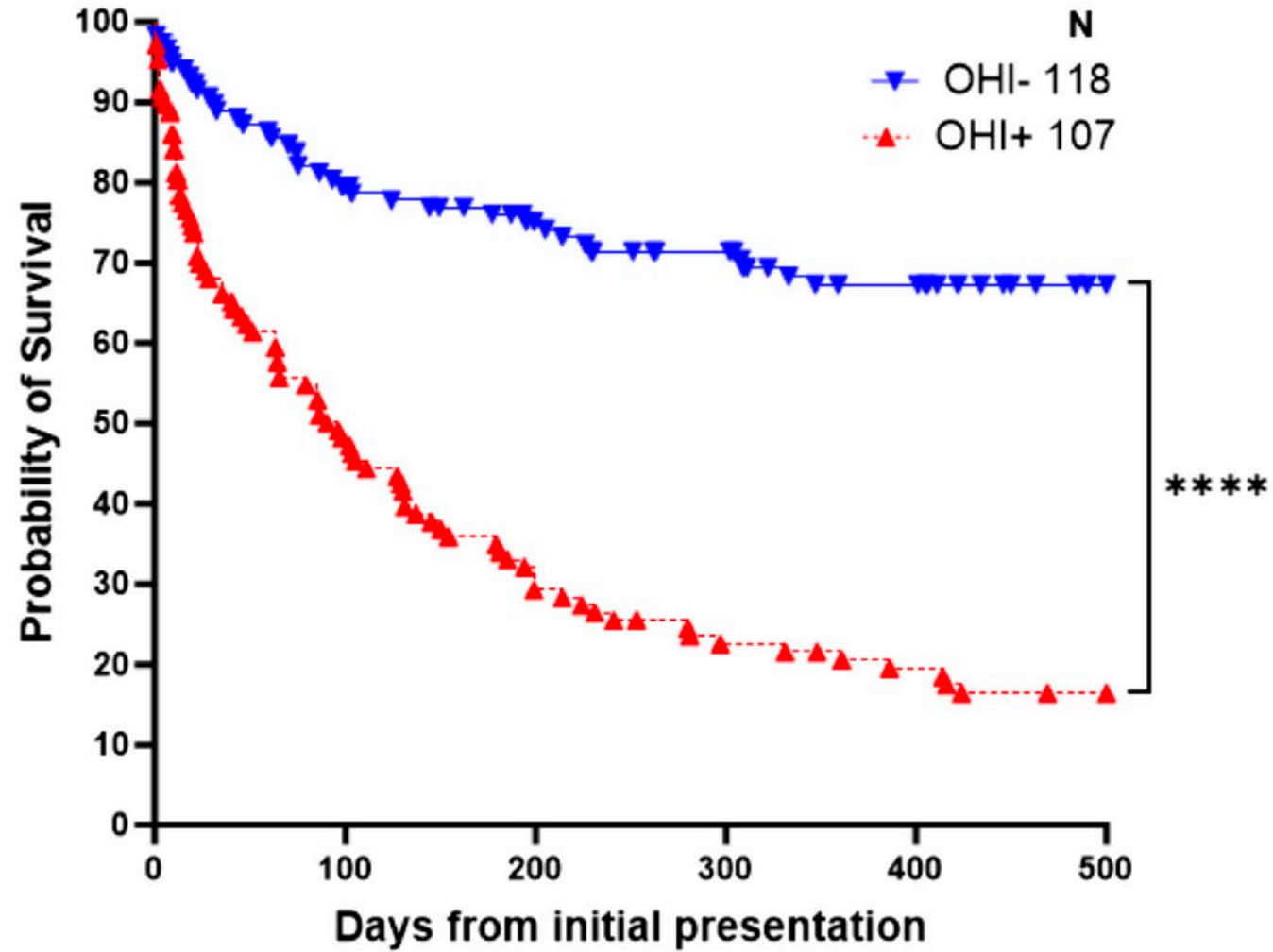
## An improved index for diagnosis and mortality prediction in malignancy-associated hemophagocytic lymphohistiocytosis

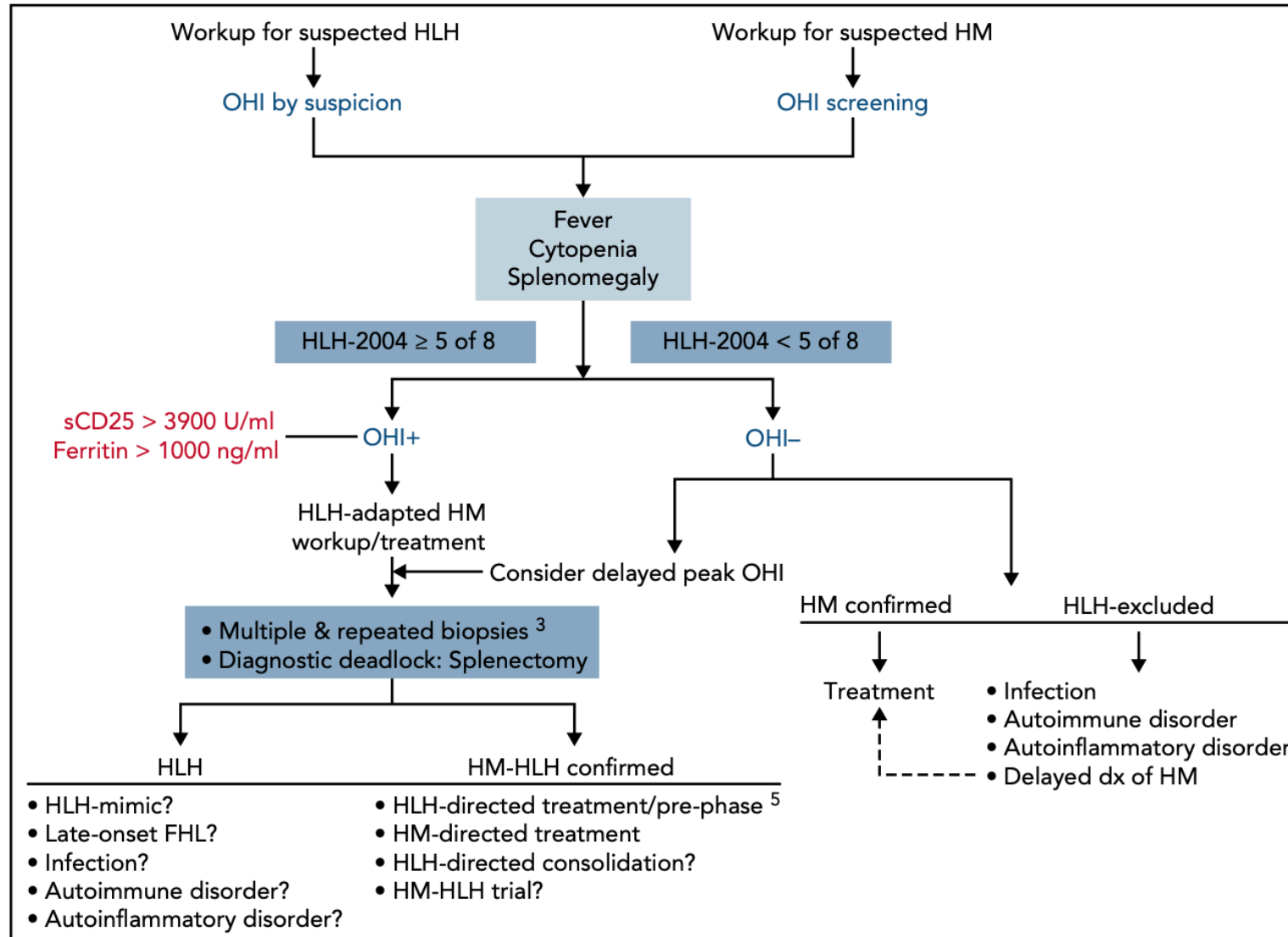
Adi Zoref-Lorenz,<sup>1-3</sup> Jun Murakami,<sup>4</sup> Liron Hofstetter,<sup>3,5</sup> Swaminathan Iyer,<sup>6</sup> Ahmad S. Alotaibi,<sup>7,8</sup> Shehab Fareed Mohamed,<sup>7</sup> Peter G. Miller,<sup>9-11</sup> Elad Guber,<sup>3,12</sup> Shiri Weinstein,<sup>3,13</sup> Joanne Yacobovich,<sup>3,14</sup> Sarah Nikiforow,<sup>9</sup> Benjamin L. Ebert,<sup>9,11,15</sup> Adam Lane,<sup>16</sup> Oren Pasvolsky,<sup>3,5</sup> Pia Raanani,<sup>3,5</sup> Arnon Nagler,<sup>3,17</sup> Nancy Berliner,<sup>10</sup> Naval Daver,<sup>7</sup> Martin Ellis,<sup>1,3,\*</sup> and Michael B. Jordan<sup>2,18,\*</sup>

sIL2-R



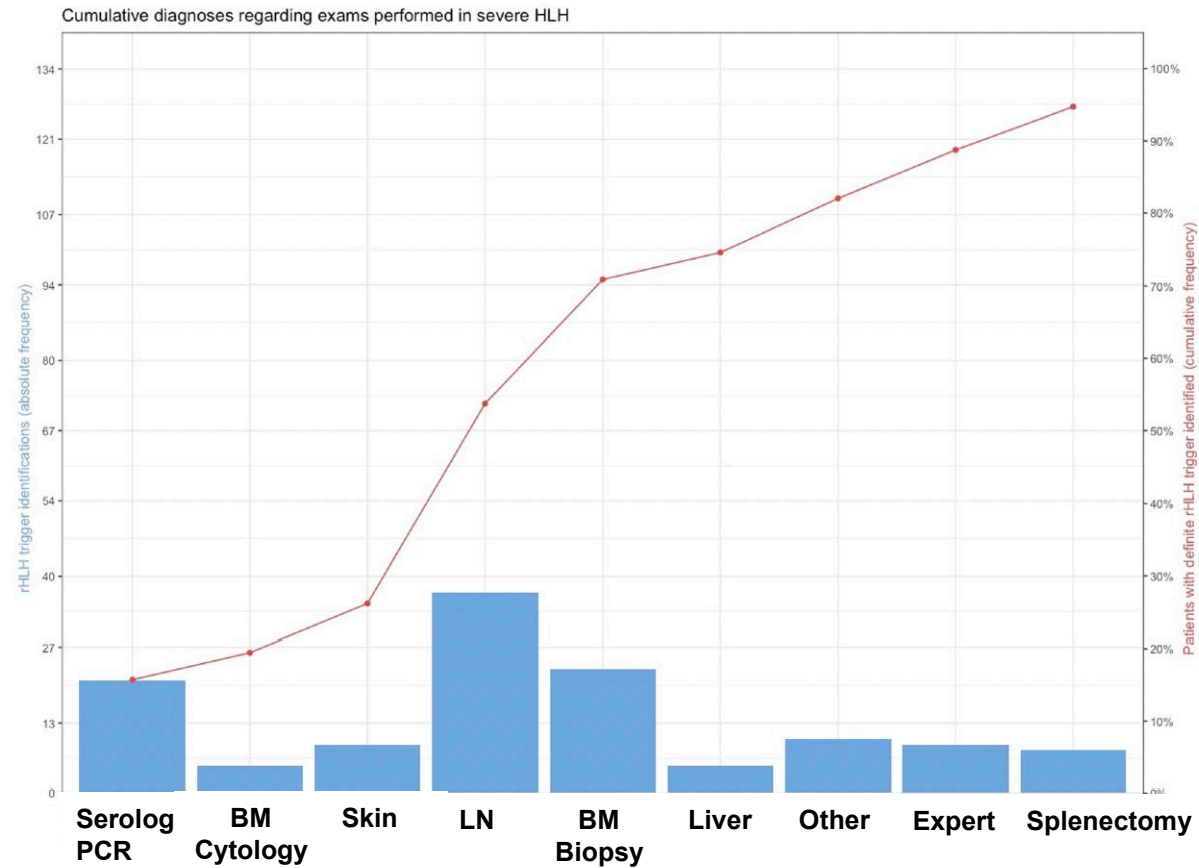
Ferritin







# Okkulte hämatologische Neoplasie durch aggressive Biopsiestrategie detektieren



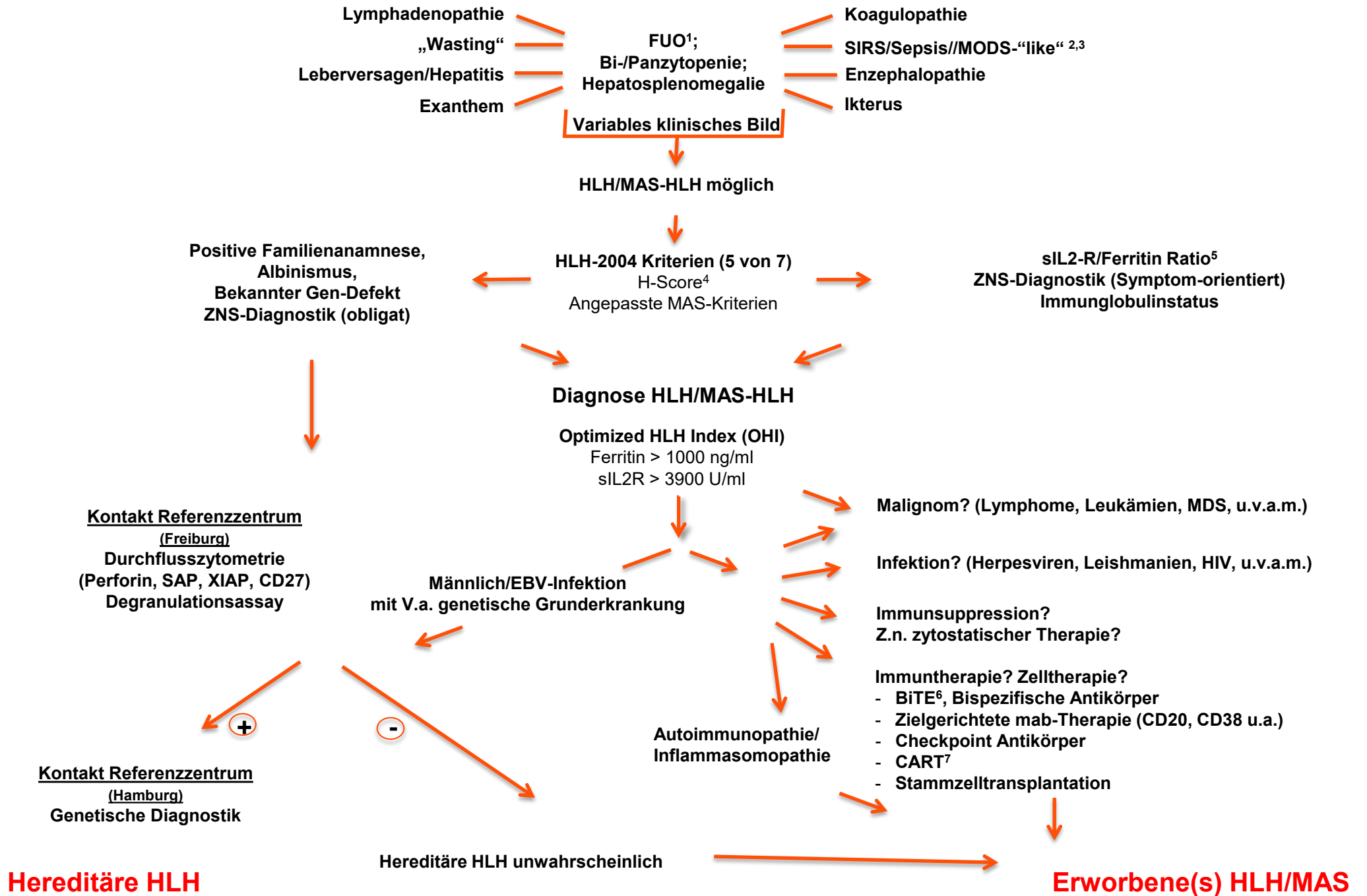
## Diagnostische Scores, HLH, MAS, MAS-ähnlich

	Fever	Ferritin	sIL2R	Hepatho- pathy	Organo- megaly	Cytopenia	Coagulo- pathy	Haemo- phago- cytosis	Hyper- triglyze- demia	Renal Failure	Resp Failure
<b>HLH</b>											
<b>HLH-2004<sup>1</sup></b>	✓	500	2400	✓	✓	✓	✓	✓	✓		
<b>H-Score<sup>2</sup></b>	✓	6000 2000-6000 2000		✓	✓	✓	✓	✓	✓		
<b>AML-HLH<sup>3</sup></b>	✓	8000						✓			
<b>OHI<sup>4</sup></b>	✓	1000	3900	✓	✓	✓	✓	✓	✓		
<b>CARTOX<sup>5</sup></b>		10000		✓				✓		✓	✓
<b>MAS<sup>6</sup></b>	✓	684		✓		✓	✓		✓		
<b>MAS-like Sepsis<sup>7</sup></b>				✓			✓				
<b>Covid (CIS)<sup>8</sup></b>	✓			✓			✓		✓		✓

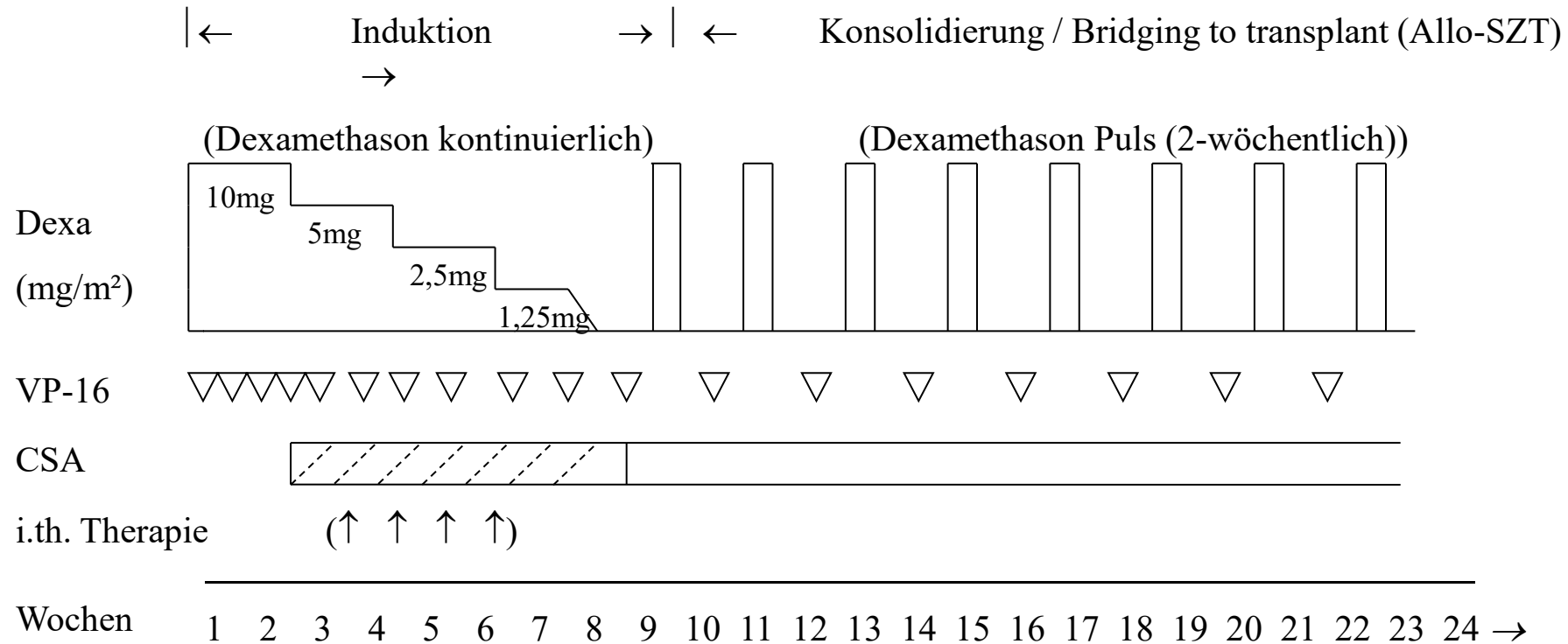
<sup>1</sup> Henter et al., *Pediatr Blood Cancer* 2007; <sup>2</sup> Fardet et al., *Arthritis & Rheumatology* 2014; <sup>3</sup> Delavigne et al., *Haematologica* 2014;

<sup>4</sup> Zoref-Lorenz et al., *Blood* 2022; <sup>5</sup> Neelapu S et al. *Nat Rev Clin Oncology* 2017; <sup>6</sup> Ravelli et al., *Arthritis & Rheumatology*, 2015;

<sup>7</sup> Shakoory et al., *Critical Care Medicine* 2016; <sup>8</sup> La Rosée F et al., *Leukemia* 2020



## Die kanonischen HLH-2004 Säulen



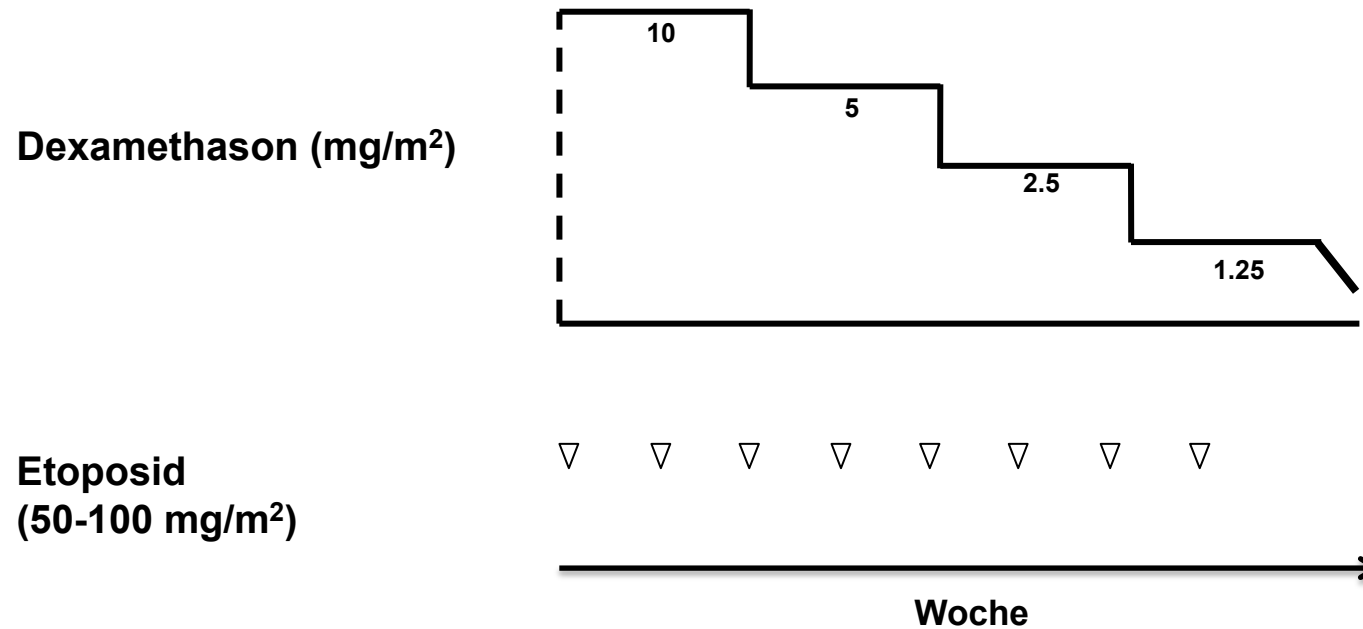
**Achtung: pädiatrisches Protokoll !!!**

- Dexa = Dexamethason Induktion (10 mg/m<sup>2</sup> Woche 1-2, 5 mg/m<sup>2</sup> Woche 3-4, 2,5 mg/m<sup>2</sup> Woche 5-6, 1,25 mg/m<sup>2</sup> Woche 7, Woche 8 ausschleichen. Bei nicht familiärer HLH ggf. Therapieende bzw. individuelle Therapieführung. Konsolidierungstherapie bei familiärer HLH mit Dexa-Pulsen Tag 1 – 3 zweiwöchentlich (10 mg/m<sup>2</sup>).
- VP-16 = Etoposid 150 mg/m<sup>2</sup> iv.
- CSA = Cyclosporin A Zielspiegel 200 µg/L (Monoklonaler Assay, Talspiegel). Start Woche 9, ggf. früher, cave Toxizität in Woche 1& 2 (Ehl. et. al, 2017)
- i.th. = Methotrexat intrathekal: <1 J 6 mg, 1-2 J 8 mg, 2-3 J 10 mg, >3 J 12 mg.  
Nur bei progredienten neurologischen Symptomen oder bei abnormalem Lumballiquor.

## Erfolgsgesichte der pädiatrischen HLH

	HLH94 <sup>11,12</sup>	HLH2004 <sup>10</sup>	Current study (active disease)	Current study (asymptomatic)
pHLH definition	affected sibling	genetics (94%) affected sibling (6%)	genetics (96%) function (4%)	genetics 100%
Patient number	60	168 FHL 29 non-FHL (15%)	56 FHL 20 non-FHL (26%)	7 FHL 5 non-FHL (42%)
Median age at onset	2 months	3.4 months	5.6 months	n.a.
Treatment (first-line)	HLH94	HLH2004	no major drug 10 (13%) etoposide 57 (75%) alemtuzumab 7 (9%) emapalumab 2 (3%)	4 Cyclosporin/IVIg 8 none
Median time diagnosis to SCT	183 days	148 days	88 days	246 days (birth to SCT)
Survival before SCT	44/60 (73%)	140/168 (83%)	69/76 (91%) etoposide 52/57 (91%)	12/12 (100%)
Survival after SCT	29/44 (66%)	94/135 (70%)	56/65 (86%) etoposide 44/52 (85%)	10/10 (100%) <sup>#</sup>
<b>Overall Survival [CI]</b>	<b>50% [± 13%] (5y)</b>	<b>59% [52-67%] (5y)</b>	<b>79% [68-86%] (3y) etoposide 77% [64-86%]</b>	<b>100% (3y)</b>

# Behandlung des drohenden Organversagens „Vorphase-Therapie“ der erworbenen HLH



Individualisiert:

Polyvalentes Immunglobulin 1 – 1,6g/kg verteilt über 3 Tage

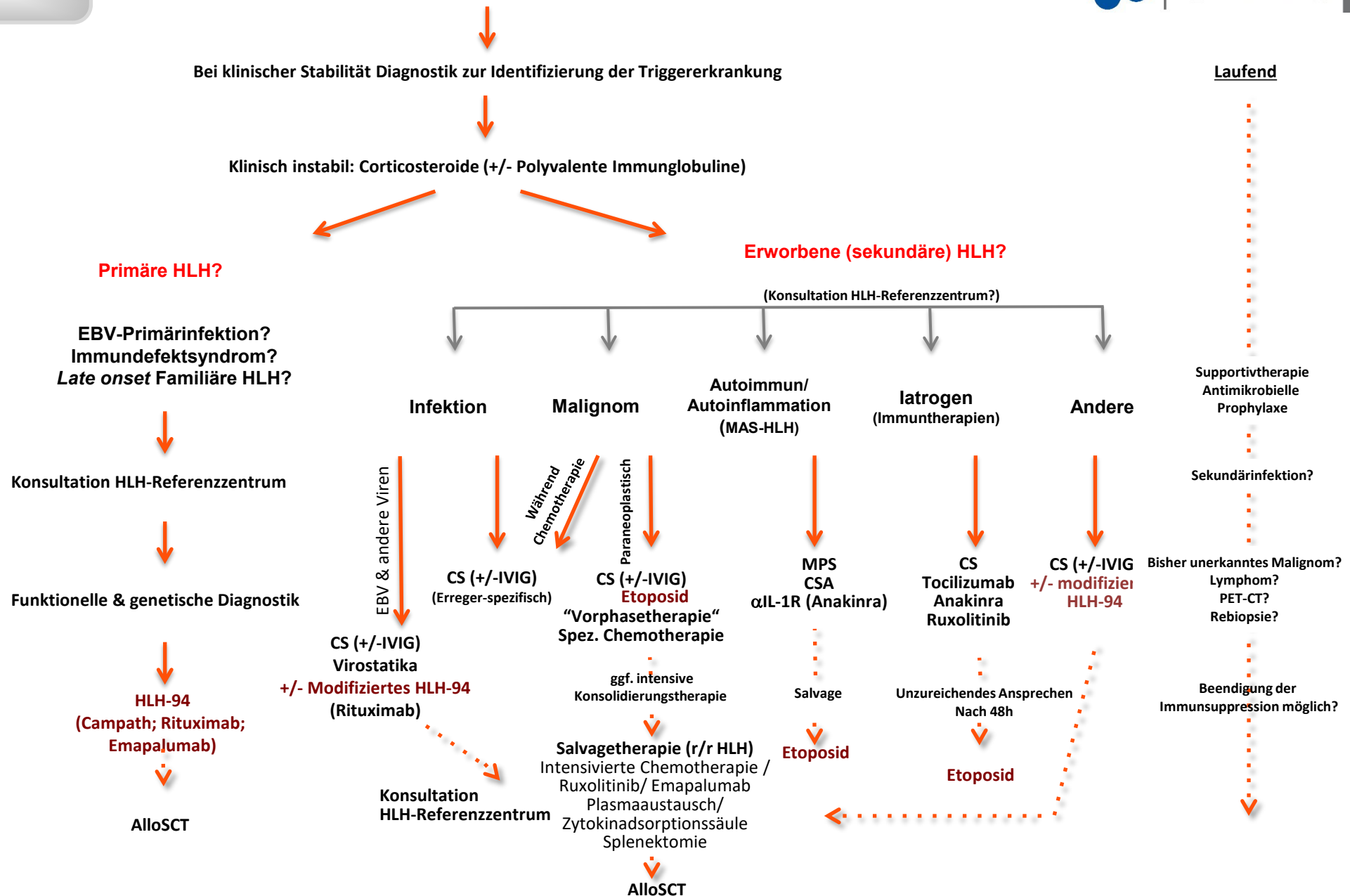
Erhaltungstherapie:

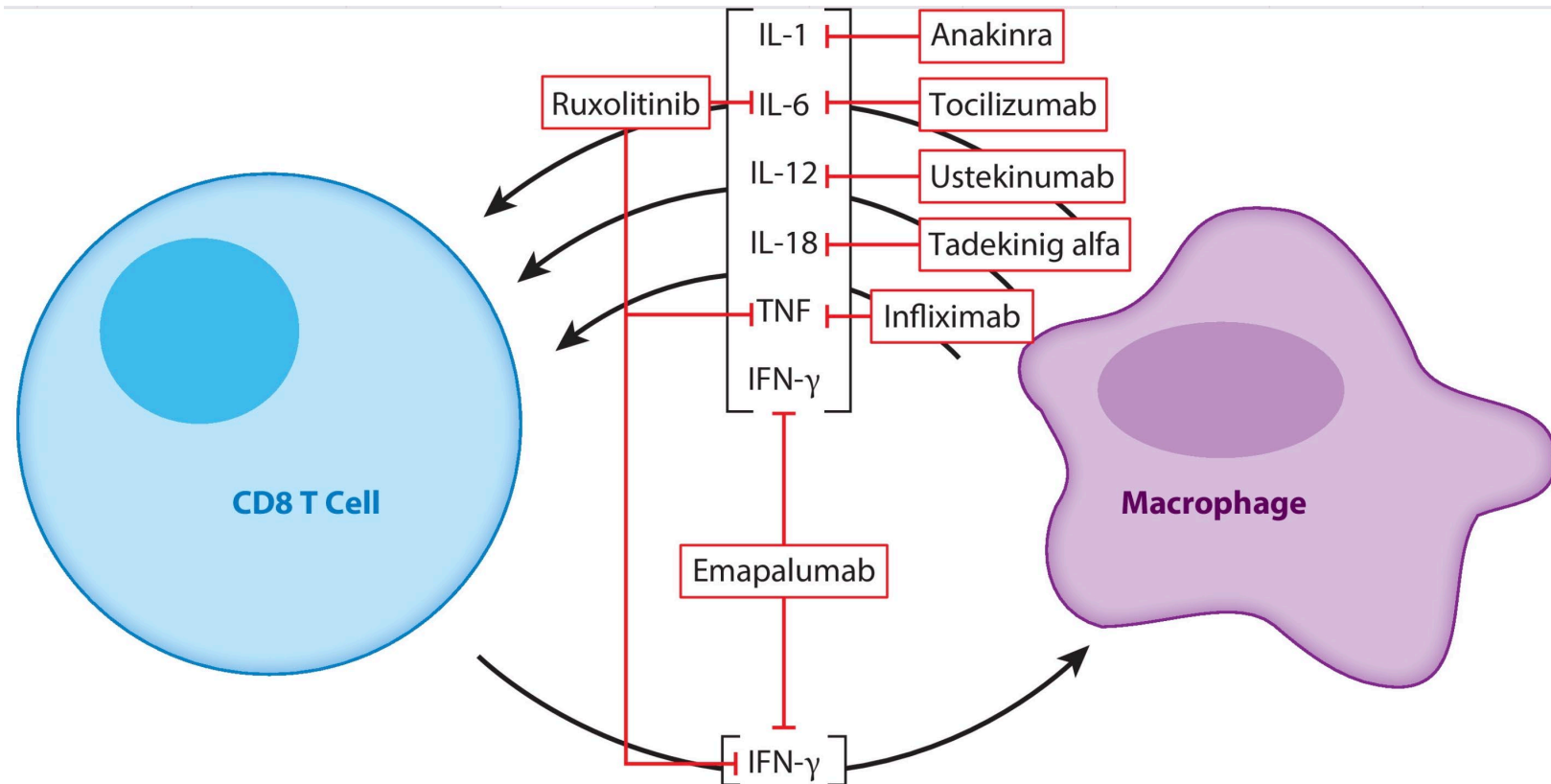
Ciclosporin A (150 – 200 µg/l Talspiegel)

oder

Tacrolimus ca. 0,2mg/kg/Tag (ca. 10ng/ml Talspiegel)







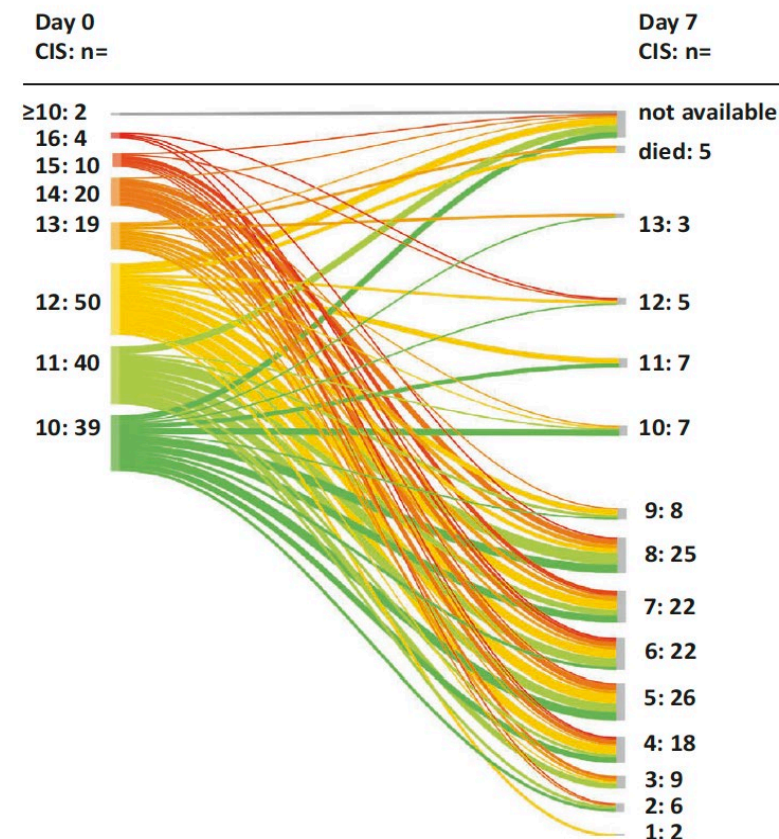
# Covid-19 CRS: RuxCoFlam

**Table 1** COVID hyperinflammation score ( $\geq 10$  of 16 threshold for inclusion).

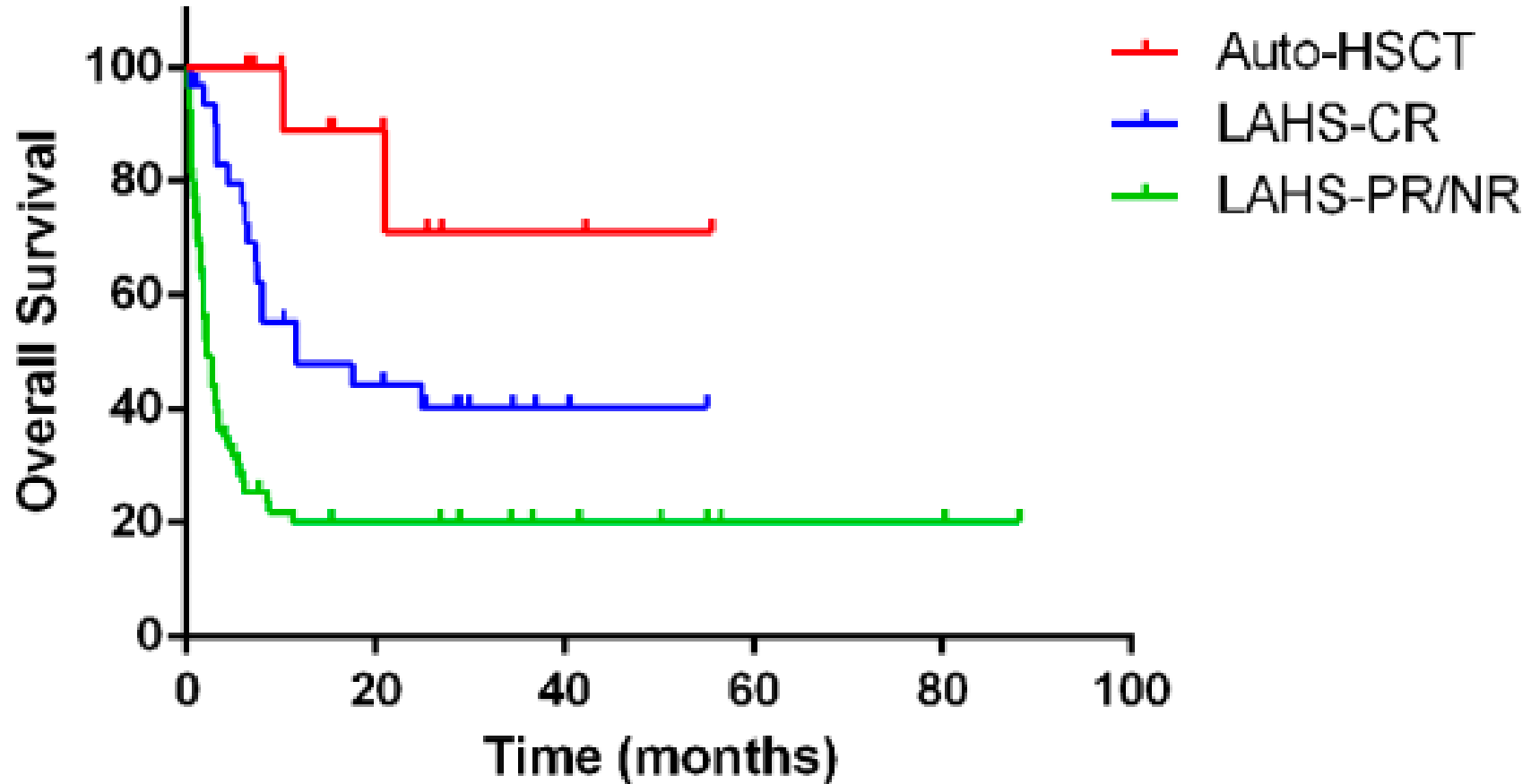
Points	
Chest-X-ray/Chest-CT consistent w/ hypersensitivity pneumonitis	3
CRP > 20 × ULN	2
Ferritin > 2 × ULN	2
Triglycerides > 1.5 × ULN	1
IL-6 > 3 × ULN	1
Fibrinogen > ULN	1
Leukocytes > ULN	1
Lymphopenia < 1.1/nL	2
Fever > 38.5 °C	2
Coagulation disorder	1
- DIC (D-Dimer > ULN)	
- PTT > ULN	

*ULN* Upper limit of normal, *DIC* Disseminated Intravascular Coagulation, *PTT* Partial thromboplastin time.

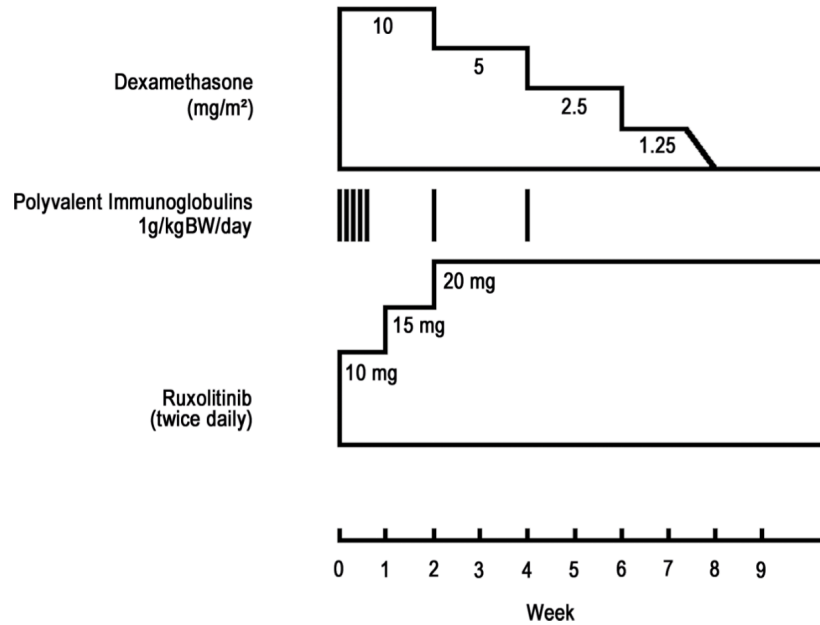
## 10mg Ruxolitinib bid



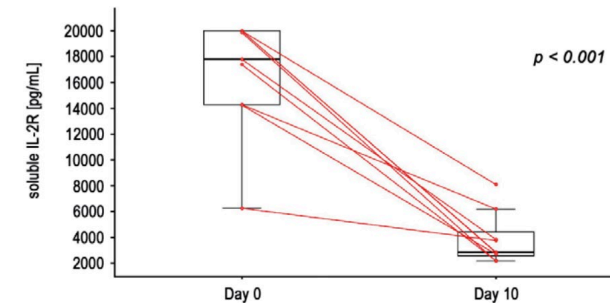
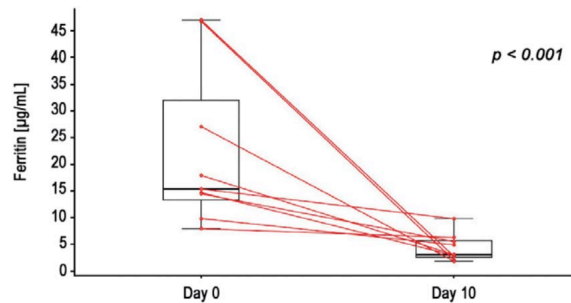
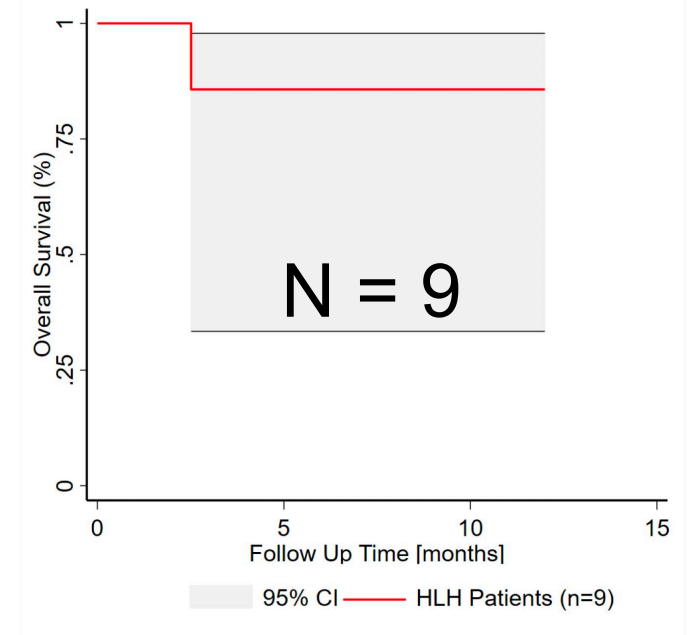
## LA-HLH: Einzelfallentscheidung zur Auto-PBSZT



## Chemotherapie-frei: Dex-Rux



Female	3 (33%)
Age	43 [32-54]
BMI	24.6 [22.6-26.9]
Underlying cause of HLH	
----EBV	3 (33%)
----Hodgkin's lymphoma	1 (11%)
----CAEBV/ Hodgkin's lymphoma	1 (11%)
----HIV	1 (11%)
----Legionella pneumophilia	1 (11%)
----PUUV	1 (11%)
----unknown/idiopathic	1 (11%)
Time to HLH diagnosis[days]	0 [0-15]
SOFA	9 [7-12]
---respiratory	2 [1-3]
---coagulation	2 [1-3]
---liver	2 [1-2]
---renal	0 [0-1]
---CNS	1 [0-4]
---hemodynamic	0 [0-4]
Ventilation	
----vvECMO	1(11%)
----IV	3 (33%)
----NIV	2 (22%)
----O <sub>2</sub> -support	4 (44%)
Vasopressor	4 (44%)
Renal replacement therapy	2 (22%)



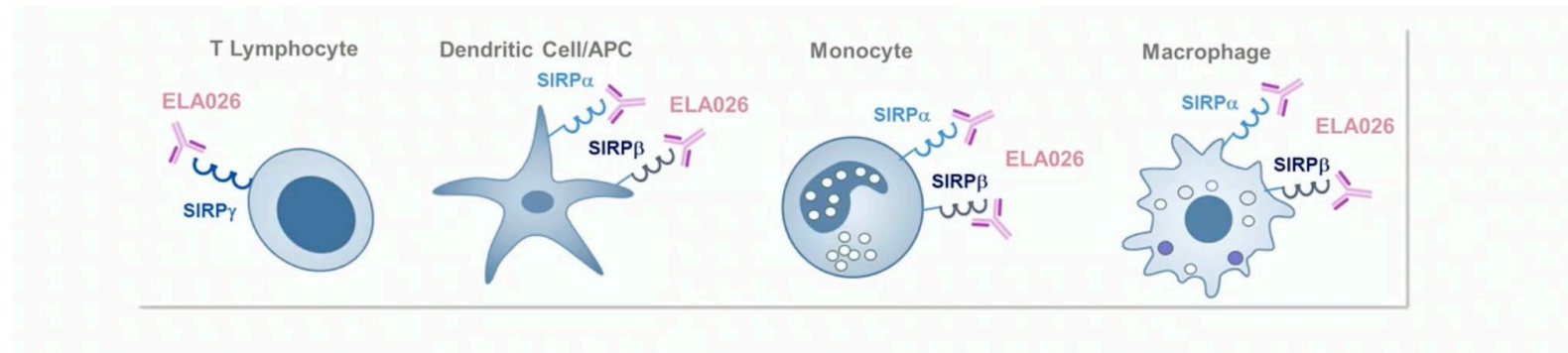
# In Entwicklung

## ELA026 TARGETING OF SIRP(+) IMMUNE CELLS RESULTS IN A HIGH RESPONSE RATE AND IMPROVED 2-MONTH SURVIVAL OF TREATMENT-NAÏVE MALIGNANCY-ASSOCIATED HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS IN A PHASE 1 STUDY

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June 16, 2024  
EHA2024 Hybrid Congress  
Late-Breaking Oral Session  
Abstract LB3442

\*Co-Principal Investigators





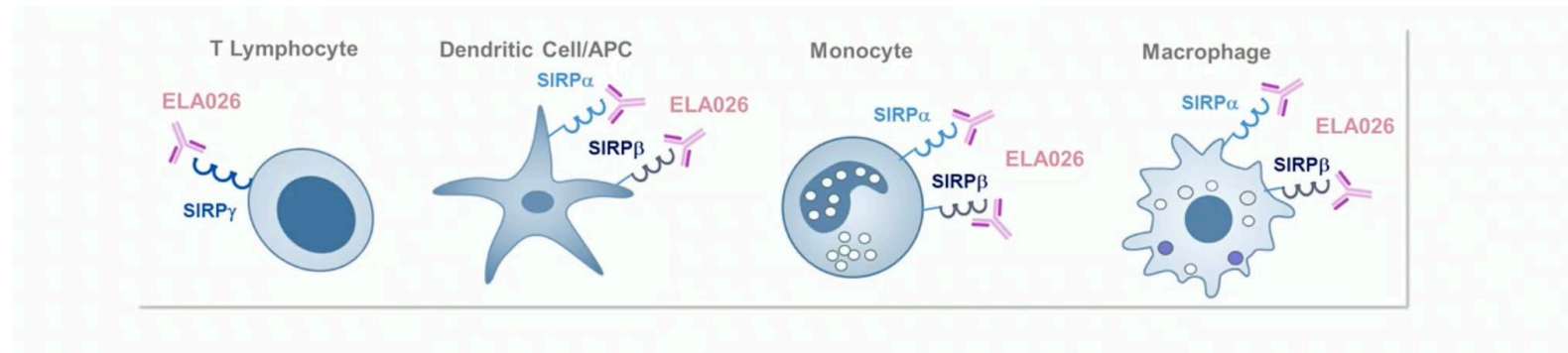
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Klassische Lymphom/Malignomnachsorge

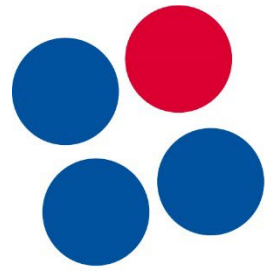
Immun-Rekonstitution, PjP-Prophylaxe

Infektionsmanagement, Re-Flare DD HLH vs Infektion

## Was ist neu?

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- **Die steigende Erkrankungsrate ist nicht nur HLH-Hype/Awareness**
- **Der optimierte HLH-Index (OHI) liefert Schwellenwerte für Ferritin & sCD25, die aggressive Diagnostik und Therapie rechtfertigen**
- **Immune Effector Cell-Associated HLH-Like Syndrome**
- **Die zunehmende Bedeutung von Zytokin-gerichteter, Chemotherapie-freier Therapie (Anakinra, Ruxolitinib, Emapalumab)**
- **Deutlich verbesserte OS-Daten für pädiatrische HLH und modifizierte HLH-2004 Kriterien (5 von 7, ohne NK-Zell-Aktivität)**

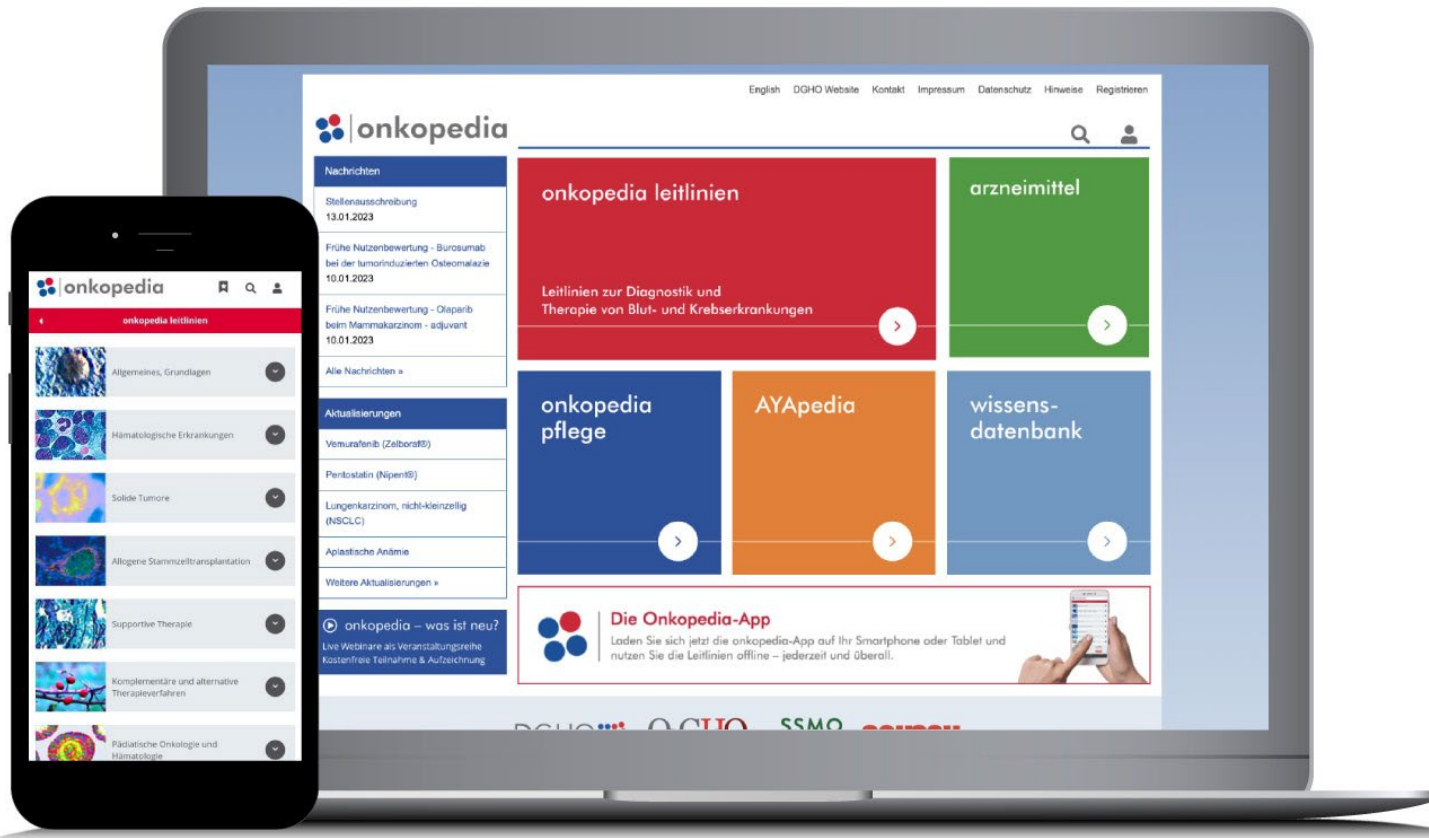


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