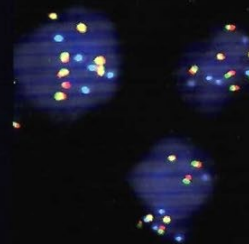
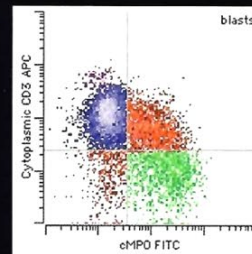
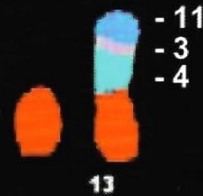
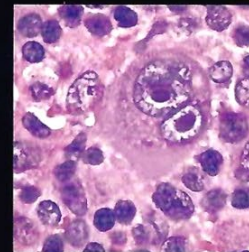
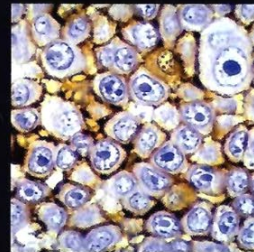
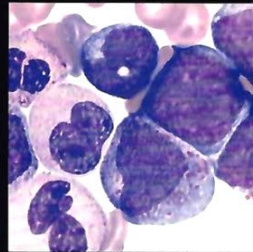
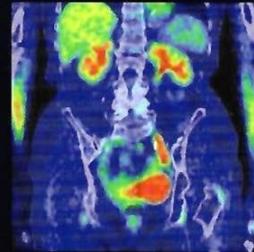


# WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues

Steven H. Swerdlow, Elias Campo, Nancy Lee Harris, Elaine S. Jaffe, Stefano A. Pileri, Harald Stein, Jürgen Thiele, Daniel A. Arber, Robert P. Hasserjian, Michelle M. Le Beau, Attilio Orazi, Reiner Siebert



# Schematic view on relation between lymphomas and leukemias

**solid neoplasm**

transitional

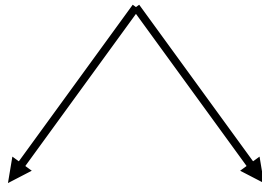
**bone marrow neoplasm**

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**LYMPHOMA**

features

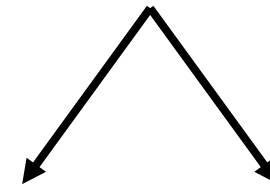
**LEUKEMIA**  
type: CLL, HCL, T and other



PB seeding

present

**absent**



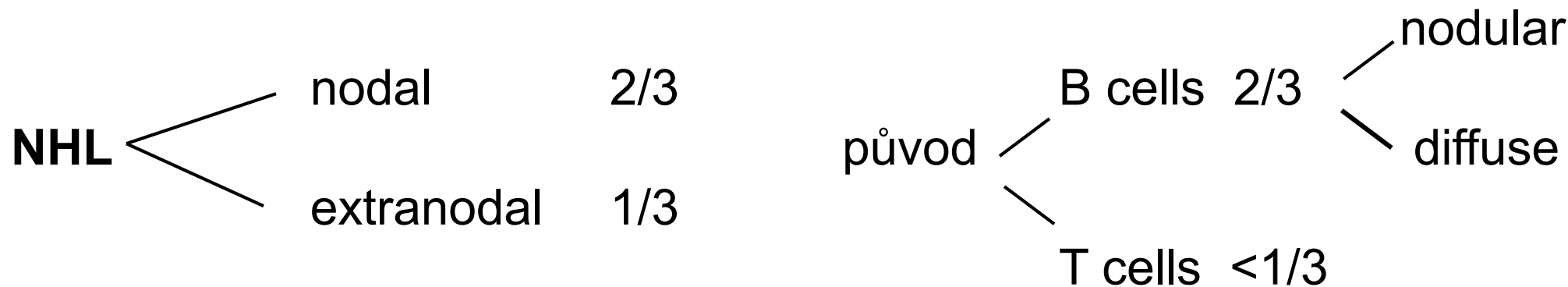
PB seeding

**present**

absent

# Neoplasms of the hemopoetic system - lymphomas

ALL, CLL, HCL



## Hodgkin lymphoma (HL – cHL, NLPHL)

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neoplasms of the reticular cells (rare)

histiocytic diseases

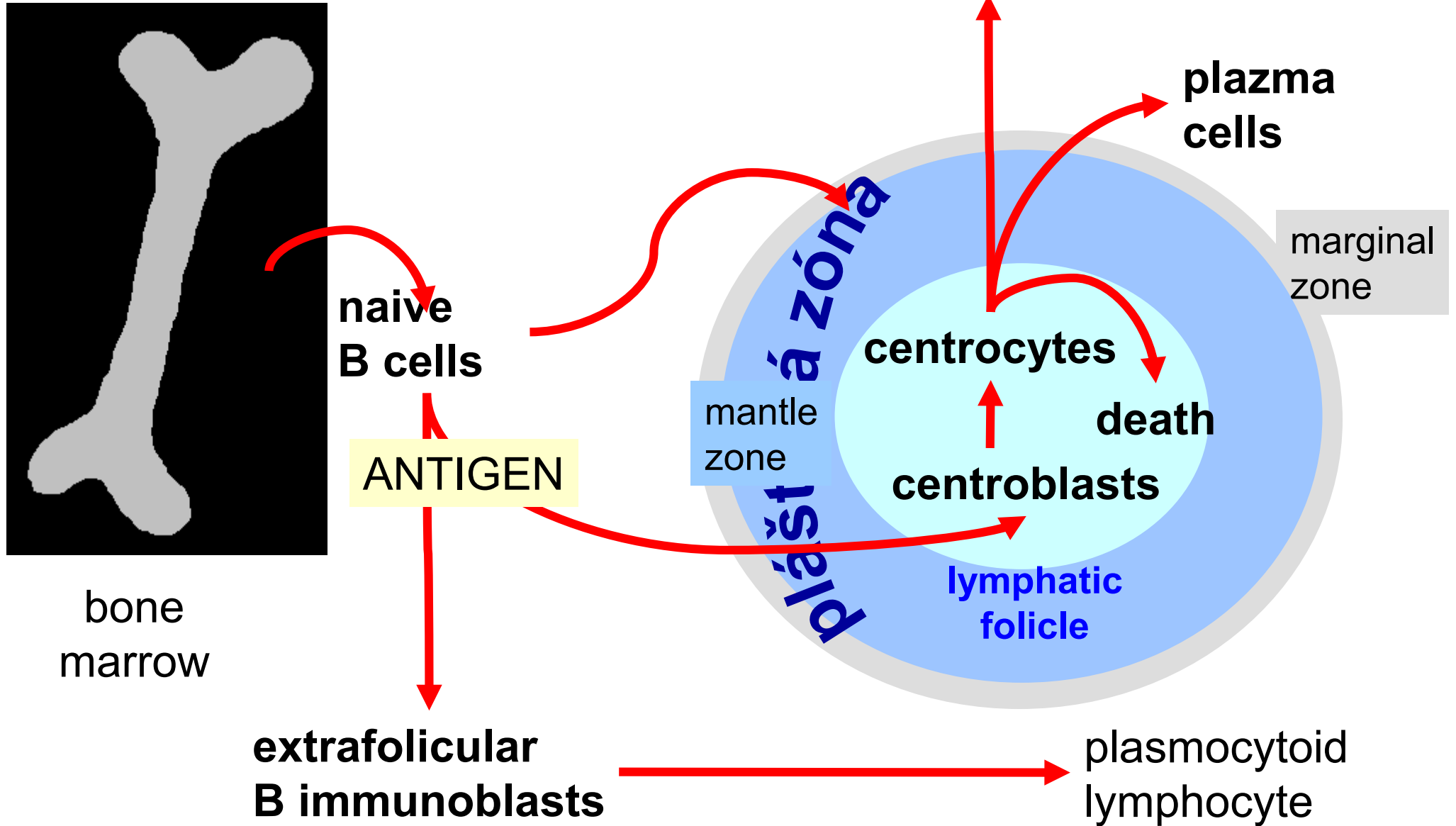
„common histiocytes“  
LCH

# Neoplasms of the hemopoetic system - lymphomas

non-Hodgkin lymphomas - NHL

about 4 % of all malignant neoplastic diseases

# Development of B cell lymphocytic line



# WHO Classification of the Lymphoid Neoplasms

*I. Precursor B-Cell Neoplasms*      CD19 +, CD10 +, TdT +

*II. Peripheral B-cell Neoplasms*      CD20 +

# WHO Classification of the Lymphoid Neoplasms

## *I. Precursor B-Cell Neoplasms*

precursor B lymphoblastic leukemia / lymphoma

## *II. Peripheral B-cell Neoplasms*

chronic lymphocytic leukemia / small lymphocytic lymphoma

lymphoplasmacytic lymphoma

mantle cell lymphoma

follicular lymphoma (grade I, II, III)

marginal zone lymphoma

hairy cell leukemia

plasma cell myeloma

diffuse large B-cell lymphoma

Burkitt lymphoma

# non-Hodgkin lymphomas - NHL

**symptomatology** 14 days and more

**local** with regards of the tumor site

LN enlarged, non-tender !

gastric symptomatology

increased intracranial pressure

nondescript pains, e.g. in the abdominal cavity

tumor – tissue enlargement of a visible part of the body

**systemic**

fatigue

weight loss

fevers over 38 °C

night sweats

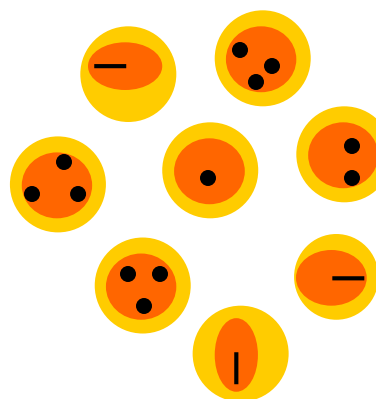
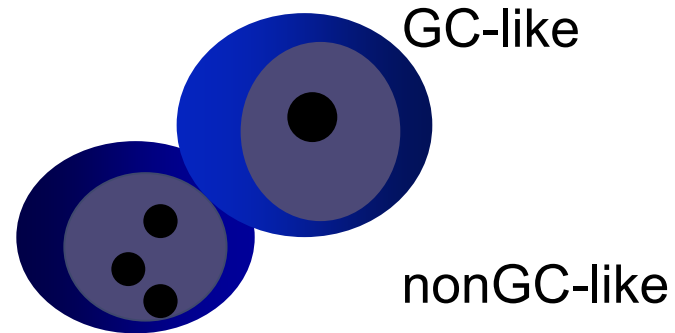


**A model of lymphoma pathogenesis  
from germinal centre cells**

**de novo DLBCL**

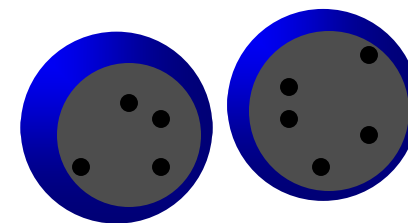
**germinal  
centre**

**CD10 a BCL6  
MUM1 (IRF4)**



**CD10 a C-MYC**

**Burkitt lymphoma**

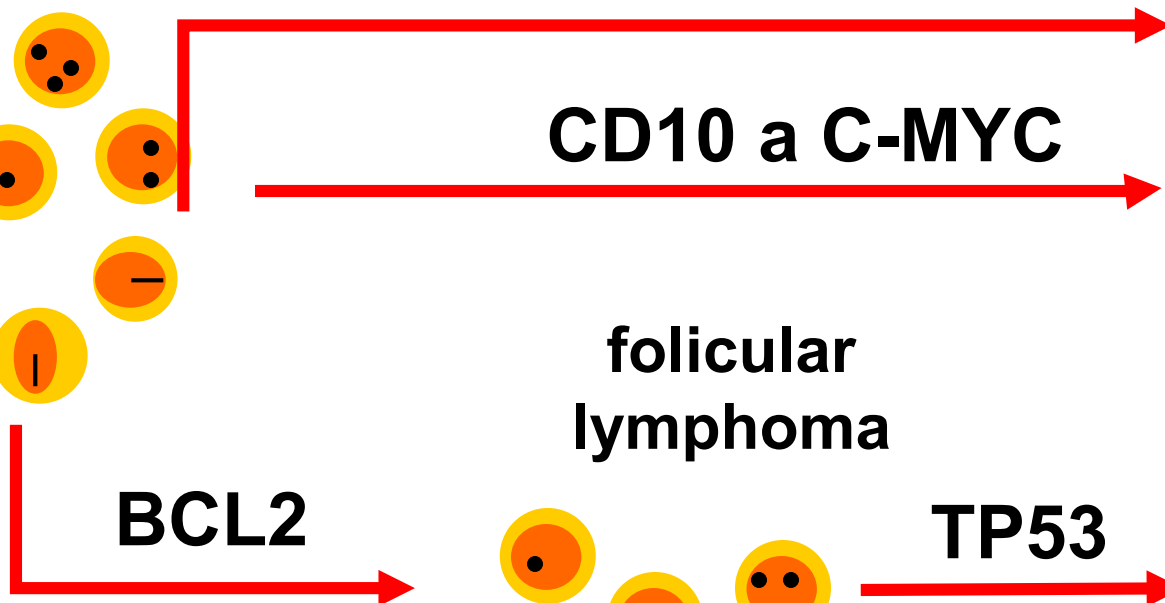
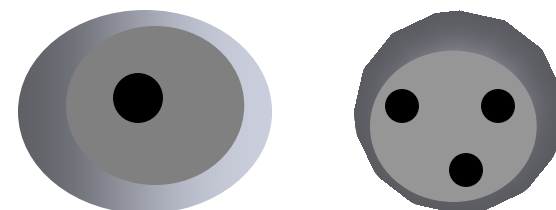
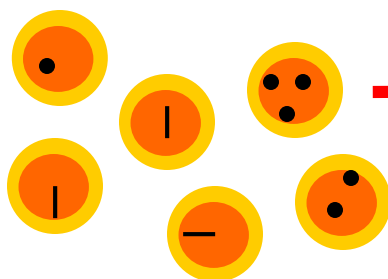


**BCL2**

**follicular  
lymphoma**

**TP53**

**transformed  
DLBCL**



diffuse large B cell lymphoma  
difúzní velkobuněčný B lymfom

**DLBCL**

**> 40 %**

ICD-O  
M 9680/3

# DLBCL

prognosis depends on

**stage of the disease = localised x generalised**

immunofenotype of the tumor

GC - like

non GC - like

double expressor

BCL-2

MYC

..

genotype

..

double hit DLBCL

gen *BCL-2* rearrangement

gen *MYC* rearrangement

primary mediastinal (thymic) large B cell lymphoma

primární mediastinální velkobuněčný B lymfom

## **PMBCL**

**~ 2 - 3 %**

median 35 years

females : males / 2 : 1

ICD-O

M 9679/3

Burkitt's lymphoma

Burkittův lymfom

**BL**

**> 1 %**

ICD-O  
M 9687/3

# Burkittův lymfom

endemic

# Burkittův lymfom

sporadic

follicular lymphoma  
folikulární lymfom

**FL**

**> 25 %**

ICD-0  
M 9690/3



mantle cell lymphoma

lymfom z buněk pláště

**MCL**

**> 7 %**

ICD-O  
M 9673/3

marginal zone lymphoma  
lymfom z buněk marginální zóny

*extranodal – MALT lymfom*

*splenic*

*nodal*

**MZL**

**10 %**

ICD-O  
M 9699/3

chronic lymphocytic leukemia / small lymphocytic lymphoma

chronická lymfocytická leukémie / malobuněčný lymfocytický lymfom

**CLL / SLL**

**> 10 %**

ICD-O

M 9823/3

M 9670/3

**plasma cell myeloma**

**PCM**

*multiple myeloma*

*mnohotný myelom*

15% hematological malignancies

ICD-O

M9732/3

**precursor**

**MGUS**

ICD-O

M 9765/1

# WHO Classification of the Lymphoid Neoplasms

## ***III. Precursor T-Cell Neoplasms***

precursor T lymphoblastic leukemia / lymphoma

## ***IV. peripheral T-Cell and Natural Killer Cell Neoplasms***

T-cell chronic lymphocytic leukemia

Large granular lymphocytic leukemia

Mycosis fungoides and Sézary syndrome

Peripheral T-cell lymphoma, unspecified

Angioimmunoblastic T-cell lymphoma

NK / T-cell lymphoma

Intestinal T-cell lymphoma

Adult T-cell leukemia / lymphoma

Anaplastic large cell lymphoma

Mycosis fungoides

**MF**

peripheral T cell lymphoma, unspecified

periferní T lymfom, blíže neurčený

**PTCL**

> 25 %

enteropathy associated T-cell lymphoma

enteropatický T lymfom

**EATCL**

< 5 %



anaplastic large cell lymphoma

anaplastický velkobuněčný lymfom

**ALCL**

**CD30+ ALK1-**

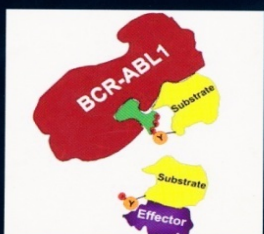
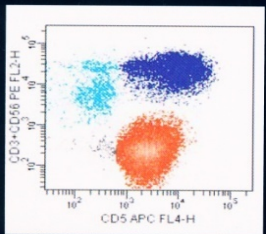
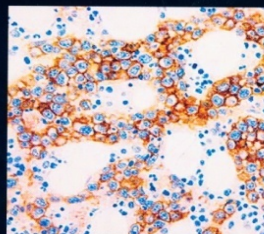
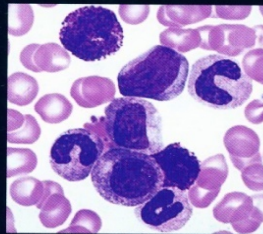
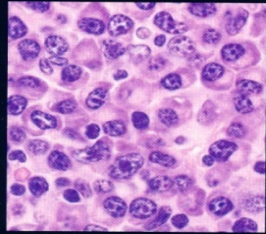
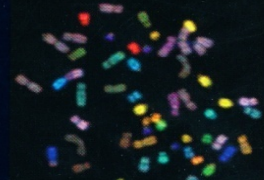
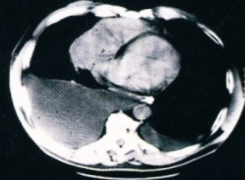
> 5 %

**CD30+ ALK1+**

7 %

# WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues

Edited by Steven H. Swerdlow, Elias Campo, Nancy Lee Harris, Elaine S. Jaffe, Stefano A. Pileri, Harald Stein, Jürgen Thiele, James W. Vardiman



## Hodgkin lymphoma

- classic HL (c-HL)
- nodular lymphocyte predominant Hodgkin lymphoma (NLPHL)

## people

- Hodgkin
- Reed
- Sternberg

## cells

- Hodgkin
- Reed-Sternberg H/RS

### their modifications:

- lacunar
- mumified
- popcorn (L&H / LP) cells

# Hodgkin lymphoma

- **neoplastic cells < inflammatory cells**
- **mononuclear cells + multinucleated cells (H/ RS)**
- **lymph node / coalescent**
- **young adults**
- **30 – 40 % lymphomas**

# Classic Hodgkin lymphoma, c-HL

## Clinical:

- coalescent LN
- neck 75%
- mediastinum
- fever, night sweats, weight loss (40%)

## Stage:

I: 1 LU / spleen / thymus

II: 2 LU a more / diaphragm

III: both sides of the diaphragm

IV: + extranodal

# Classic Hodgkin lymphoma, c-HL

## Classification

1. Nodular sclerosis 65-70%
2. Mixed cellularity 25-30%
3. Rich in lymphocytes 5%
4. Lymphocyte depletion <1%

# **NLPHL** Nodular Lymphocyte Predominant Hodgkin Lymphoma

**5% of all HL**

**favourable prognosis**

**commonly 1 LN only (neck, axillary, inguinal) = stage 1**

# **Langerhans cell histiocytosis, LCH**

- **Langerhans cells of the epidermis, formerly HX antigen presenting cells:**

**specific organelles - Birbeck granules**

- **Langerin<sup>+</sup>, S100<sup>+</sup>, CD1a<sup>+</sup>**

**involvement of the skin, bones and other tissues**



# Langerhans cell histiocytosis, LCH

- **unisystemic unifocal / multifocal**  
eosinophilic granuloma (*bones most frequently*)
- **multisystemic, chronic**  
Hand-Schüller-Christian disease
- **multisystemic, disseminated, acute**  
Abt-Letterer-Siweho disease