

# **Langerhans Cell Histiocytosis in Adults**

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# Langerhans Cell Histiocytosis

## Orphan Disease

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- Limited resources for many (inter-) national children's cancer study groups in the 70s
- Research and treatment of many entities was lost, and LCH became an “orphan disease”.

# Langerhans Cell Histiocytosis Orphan Disease

- **HOWEVER**, this might have been the best thing that happened to the histiocytoses.
- Clinicians and basic scientists with great interest in the histiocyte and its disorders banded together to build their own home:  
**THE HISTIOCYTE SOCIETY.**

# LCH in Adults

## Orphan Disease under Orphan Diseases



ELSEVIER

European Journal of Cancer 40 (2004) 1467–1473

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European  
Journal of  
Cancer

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Review

Langerhans cell histiocytosis in adults: more questions  
than answers?

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# LCH in Adults

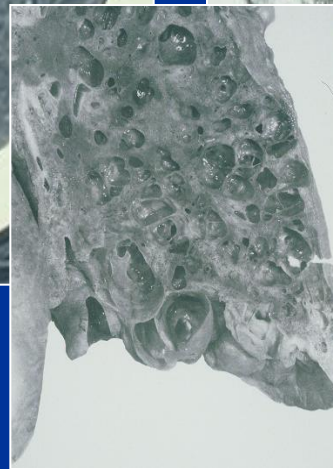
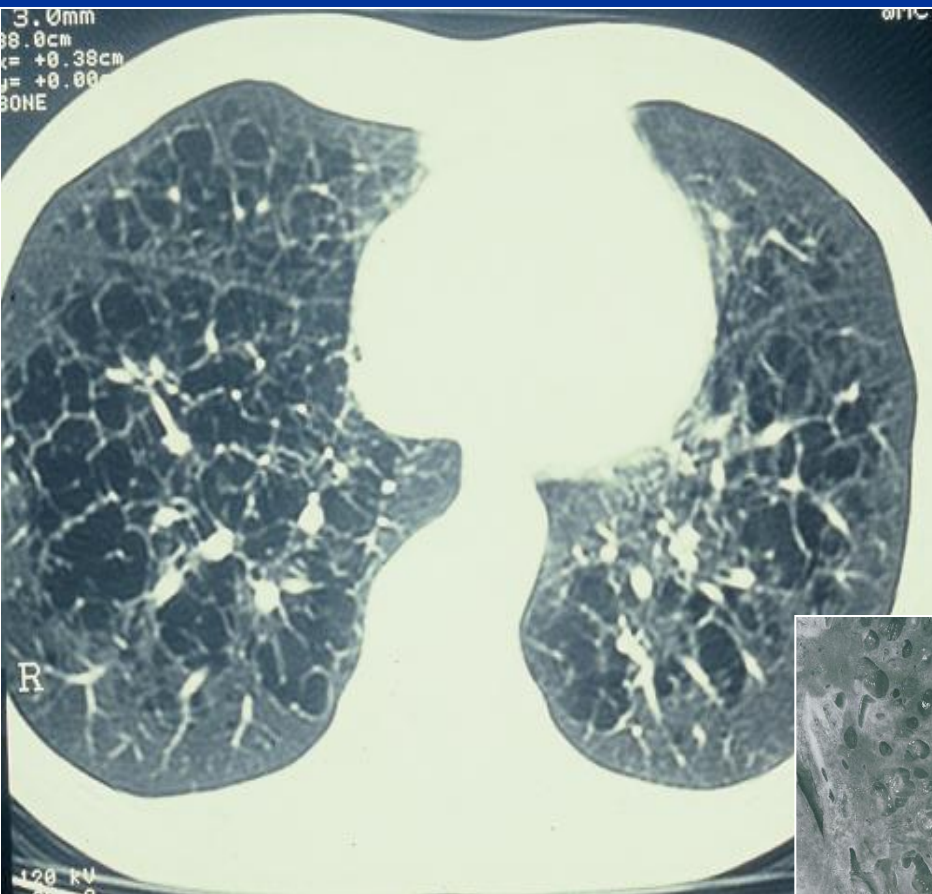
## Orphan Disease under Orphan Diseases

Summary of similarities and differences between adult and childhood LCH

Manifestation	Adult	Childhood
Bone disease	Frequent	Extremely frequent
Skin disease	Frequent	Frequent
Dental involvement	Frequent	Infrequent
Pulmonary disease	Very frequent	Infrequent
Pulmonary isolated disease	Very frequent	Exceptional
Genital involvement	Frequent	Exceptional
Diabetes insipidus	Frequent	Frequent

# LCH in Adults

## Orphan Disease under Orphan Diseases



# LCH in Adults

## Orphan Disease under Orphan Diseases

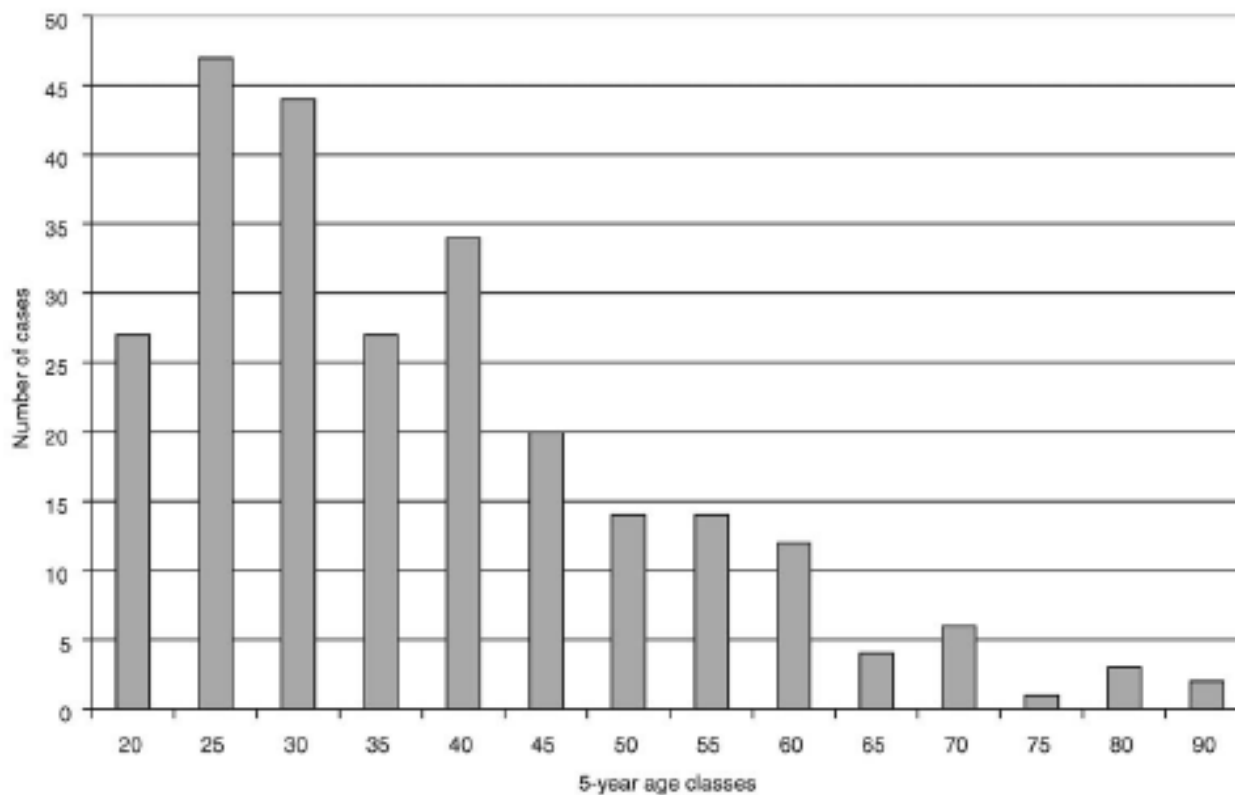


Fig. 3. Age distribution for patients with LCH diagnosed at 18 years or older.

# LCH in Adults

## Orphan Disease under Orphan Diseases



PERGAMON

European Journal of Cancer 39 (2003) 2341–2348

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European  
Journal of  
Cancer

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### Langerhans cell histiocytosis in adults

#### Report from the International Registry of the Histiocyte Society<sup>☆</sup>

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N. Grois<sup>f</sup>, J.-F. Emile<sup>g</sup>, E. Lukina<sup>h</sup>, E. De Juli<sup>i</sup>, C. Danesino<sup>j</sup>

# LCH ADULT REGISTRY

## Demographics

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- **Gender: M/F** 144/126 (53/47%)
- **Age at disease onset (n=201)** 33 yrs
- **Age at diagnosis (n=244)** 35 yrs

# LCH ADULT REGISTRY

## Family history

- **Consanguinity:** 2 (0.7%)
- **Pulmonary disease:** 16 (5.8%)
- **Thyroid disease** 10 (3.6%)
- **Diabetes insipidus** 8 (2.9%)
- **Skin disease** 6 (2.2%)
- **Neoplasia** 27 (9.8%)
- **LCH** 1 (0.4%)

# LCH ADULT REGISTRY

## Working status

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- **Unemployed** 34
- **Employed** 80
- **Unknown** 160

# LCH ADULT REGISTRY

## Associated conditions

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- **Thyroid diseases** 11 (4%)
- **Malignancy** 14 (5.1%)

# LCH ADULT REGISTRY

## List of malignancies

• Lymphoma	5*
• Skin	2
• CLL	1
• ALL	1
• Breast	1
• Prostate	1
• “Genital Ca”	1
• “malignant histiocytoma”	1
• Not defined	4

\*“mediastinal mass” n=1

# LCH ADULT REGISTRY

## Involved “sites”

• Bone	157	(57.3%)
• Skin	101	(36.9%)
• Lung	160	(58.4%)
• Lung only	44	(16.1%)
• Hepatosplenomegaly	45	(16.4%)
• Diabetes insipidus	81	(29.6%)
• Hypothyroidism	18	(6.6%)

# LCH ADULT REGISTRY

## Pattern of involvement

• Single system	29	(10.6%)
• Single system,multifocal	13	(4.7%)
• Pulmonary only	44	(16.1%)
• Multisystem	188	(68.6%)

# LCH ADULT REGISTRY

## Survival analysis

### (from disease onset)

- **Number** 197 patients
- **Median follow-up:** 54 months  
(IQR 26-115)
- **Number of events:** 15 deaths
- **Death rate:** 1.1 /100 person-yr  
(95%CI 0.7-1.8)
- **5-year survival:** 94.5%  
(95%CI 89.2-97.3)

# LCH ADULT REGISTRY

## Survival analysis (from diagnosis)

- Number: 236 patients
- Median follow-up: 28 months  
(IQR 10-67)
- Number of events: 15 deaths
- Death rate: 1.5/100 person-yr  
(95%CI 0.9-2.4)
- 5-year survival: 92.3%  
(95%CI 85.6-5.9)

# LCH ADULT REGISTRY

## Survival by disease extension

	<b>N pts (deaths)</b>	<b>Death Rate (95% CI)</b>	<b>5-year survival (95% CI)</b>
<b>SS/MF</b>	<b>37 (1)</b>	<b>0.8 (0.1-5.4)</b>	<b>100%</b>
<b>PULM</b>	<b>34 (4)</b>	<b>5.6 (2.1-15.0)</b>	<b>87.8% (54.9-97.2)</b>
<b>MS</b>	<b>46 (4)</b>	<b>1.5 (0.6-4.0)</b>	<b>87.5% (65.5-95.9)</b>
<b>MS+lung or liver</b>	<b>117 (6)</b>	<b>1.1 (0.5-2.4)</b>	<b>93.6% (84.7-97.4)</b>

# LCH in Adults

## Orphans with an Orphan Disease

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### ■ CONFERENCE REPORTS

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## Adults with LCH – orphans with an orphan disease

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