

# - French Langerhans cell histiocytosis registry

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## General

### Identification

|  |  |
|--|--|
| Detailed name  | French Langerhans cell histiocytosis registry      |
| CNIL registration number, number and date of CPP agreement, AFSSAPS (French Health Products Safety Agency) authorisation | CCTIRS : 99 087 (1999), CNIL: 99 80 71(15/07/1999) |

### General Aspects

|                     |   |
|---------------------|---|
| Medical area        | Hematology<br>Pediatrics<br>Pneumology<br>Rare diseases |
| Health determinants | Genetic   |
| Keywords            | Morbidity, Incidence, Prevalence, Mortality             |

### Scientific investigator(s) (Contact)

|                      |  |
|----------------------|--|
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| Unit                 | Service d'Héματο Oncologie Pédiatrique   |
| Organization         | Hopital Trousseau  |

### Collaborations

### Funding

|  |  |
|--|--|
| Funding status                                     | Mixed  |
| Details  | Invs, Inserm, Association Histiocytose France  |
| Governance of the database                         |  |
| Sponsor(s) or organisation(s) responsible          | Hopital Trousseau  |
| Organisation status                                | Public   |
| Additional contact                                 |  |
| Main features                                      |  |
| Type of database                                   |  |
| Type of database                                   | Morbidity registers  |
| Additional information regarding sample selection. | <p>The cases are recorded from patients' clinical records obtained from pediatric hematology or general and specialist pediatric departments. These are consulted by post, telephone or on-site monitoring.</p> <p>The national registry for hematological diseases in children is also consulted and a listing exchange is carried out annually with the on-site team on this registry.</p>   |
| Database objective                                 |  |
| Main objective                                     | <p>The public health objectives of this registry are to:</p> <ul style="list-style-type: none"> <li>- evaluate the incidence and prevalence of the disease</li> <li>- determine the risk factors of the disease manifesting and the possible prevention means</li> <li>- evaluate death rates in the population</li> <li>- The incidence and prevalence of long-term sequelae of this disease (pituitary affects ? sclerosing cholangitis ? respiratory failure ? neurological and psychiatric problems) and to evaluate prevention methods</li> <li>- Evaluate the impact of therapies on the long-term progression of the disease - particularly mortality - and long-term sequelae</li> <li>- Enable the implementation of basic biological research on broad samples of patients whose progressive profiles have been determined. Such studies have two objectives: The determination of factors for the disease manifesting and the improvement of determining factors of the disease's progression - particularly the sequelae.</li> </ul> |

|                    |  |
|--------------------|--|
| Inclusion criteria | <p>all cases of Langerhans cell histiocytosis in children under 15 years of age</p> <ul style="list-style-type: none"> <li>- proven by histology</li> <li>- defined by radio-clinical criteria if the diagnosis is validated by at least two physicians from the reference center on the basis of the following criteria:</li> </ul> <ul style="list-style-type: none"> <li>? Typical radiological lesions of the bone associated with diabetes insipidus</li> <li>? Typical radiological lesions of the bone if histology - although not providing a formal diagnosis of histiocytosis - excludes a malignant tumor, angiomas of the bones or an infection</li> <li>? Typical radiological lesions of the lung, demonstrating an association of cystic and nodular lesions</li> </ul> |
|--------------------|--|

## Population type

|     |   |
|-----|---|
| Age | <p>Newborns (birth to 28 days)</p> <p>Infant (28 days to 2 years)</p> <p>Early childhood (2 to 5 years)</p> <p>Childhood (6 to 13 years)</p> <p>Adolescence (13 to 18 years)</p> <p>Adulthood (19 to 24 years)</p> <p>Adulthood (25 to 44 years)</p> <p>Adulthood (45 to 64 years)</p> <p>Elderly (65 to 79 years)</p> <p>Great age (80 years and more)</p> |
|-----|---|

|                    |                 |
|--------------------|-----------------|
| Population covered | Sick population |
|--------------------|-----------------|

|        |                          |
|--------|--------------------------|
| Gender | <p>Male</p> <p>Woman</p> |
|--------|--------------------------|

|                |          |
|----------------|----------|
| Geography area | National |
|----------------|----------|

|                              |                         |
|------------------------------|-------------------------|
| Detail of the geography area | All of french territory |
|------------------------------|-------------------------|

## Data collection

### Dates

|  |      |
|--|------|
| Date of first collection (YYYY or MM/YYYY) | 1994 |
|--|------|

## Size of the database

|  |                           |
|--|---------------------------|
| Size of the database (number of individuals) | [1000-10 000[ individuals |
|--|---------------------------|

|                                      |                |
|--------------------------------------|----------------|
| Details of the number of individuals | 1454 (02/2011) |
|--------------------------------------|----------------|

## Data

|                              |  |
|------------------------------|--|
| Database activity            | Current data collection  |
| Type of data collected       | Clinical data<br>Paraclinical data<br>Biological data<br>Administrative data             |
| Clinical data (detail)       | Direct physical measures   |
| Paraclinical data (detail)   | Imaging, Spirometry  |
| Biological data (detail)     | Hematology, biochemistry, genetic polymorphisms, immunology                              |
| Administrative data (detail) | identification data, sociodemographic data, family tree                                  |
| Presence of a biobank        | Yes  |
| Contents of biobank          | Tissues<br>Buccal cells<br>DNA<br>DNAc/RNAc  |
| Details of biobank content   | Cryopreserved tissueTissues included in paraffinBuccal cellsDNADNAc / RNAc               |
| Health parameters studied    | Health event/morbidity<br>Health event/mortality<br>Health care consumption and services |
| Care consumption (detail)    | Medical/paramedical consultation<br>Medicines consumption                                |

## Procedures

|                                       |  |
|---------------------------------------|--|
| Data collection method                | The data is recorded from patients' clinical records obtained from pediatric hematology or general and specialist pediatric departments. These are consulted by post, telephone or on-site monitoring. |
| Classifications used                  | D76.0 C960 C 961 D76.3   |
| Participant monitoring                | Yes  |
| Details on monitoring of participants | Participant follow-up is carried out from medical records and for an undetermined period of time   |
| Links to administrative sources       | No   |

## Promotion and access

### Promotion

|                      |   |
|----------------------|---|
| Link to the document | <a href="http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Expert">http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Expert</a> |
|----------------------|---|

|                      |   |
|----------------------|---|
| Link to the document | <a href="http://www.histiocytose.org">http://www.histiocytose.org</a> |
|----------------------|---|

|                      |   |
|----------------------|---|
| Link to the document | <a href="http://www.eurohistio.net">http://www.eurohistio.net</a> |
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|                      |   |
|----------------------|---|
| Link to the document | <a href="http://tinyurl.com/PUBMED-LCH">http://tinyurl.com/PUBMED-LCH</a> |
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|-------------|------------------------------------|
| Description | Liste des publications dans Pubmed |
|-------------|------------------------------------|

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|----------------------|---|
| Link to the document | <a href="http://tinyurl.com/HAL-LCH">http://tinyurl.com/HAL-LCH</a> |
|----------------------|---|

|             |                                 |
|-------------|---------------------------------|
| Description | Liste des publications dans HAL |
|-------------|---------------------------------|

### Access

|   |   |
|---|---|
| Terms of data access (charter for data provision, format of data, availability delay) | Publications. Presentation at the annual registry day and international congresses. |
|---|---|

|                           |                                 |
|---------------------------|---------------------------------|
| Access to aggregated data | Access on specific project only |
|---------------------------|---------------------------------|

|                           |                                 |
|---------------------------|---------------------------------|
| Access to individual data | Access on specific project only |
|---------------------------|---------------------------------|