- 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 Pediatrics Pneumology Rare diseases
Health determinants	Genetic
Keywords	Morbidity, Incidence, Prevalence, Mortality

Scientific investigator(s) (Contact)

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Surname	Jean
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Unit	Service d'Hémato 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.	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. 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.

Database objective

Main objective	The public health objectives of this registry are to: - evaluate the incidence and prevalence of the disease - determine the risk factors of the disease manifesting and the possible prevention means - evaluate death rates in the population - 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 - Evaluate the impact of therapies on the long-term progression of the disease - particularly mortality - and long-term sequelae - 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.

Inclusion criteria	all cases of Langerhans cell histiocytosis in children under 15 years of age - proven by histology - 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: ? Typical radiological lesions of the bone associated with diabetes insipidus ? Typical radiological lesions of the bone if histology - although not providing a formal diagnosis of histiocytosis - excludes a malignant tumor, angiomatosis of the bones or an infection ? Typical radiological lesions of the lung, demonstrating an association of cystic and nodular lesions

Population type

Age	Newborns (birth to 28 days) Infant (28 days to 2 years) Early childhood (2 to 5 years) Childhood (6 to 13 years) Adolescence (13 to 18 years) Adulthood (19 to 24 years) Adulthood (25 to 44 years) Adulthood (45 to 64 years) Elderly (65 to 79 years) Great age (80 years and more)
Population covered	Sick population
Gender	Male Woman
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 Paraclinical data Biological data 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 Buccal cells DNA DNAc/RNAm
Details of biobank content	Cryopreserved tissueTissues included in paraffinBuccal cellsDNADNAc / RNAm
Health parameters studied	Health event/morbidity Health event/mortality Health care consumption and services
Care consumption (detail)	Medical/paramedical consultation 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	http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Expert
Link to the document	http://www.histiocytose.org
Link to the document	http://www.eurohistio.net
Link to the document	http://tinyurl.com/PUBMED-LCH
Description	Liste des publications dans Pubmed
Link to the document	http://tinyurl.com/HAL-LCH
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