Langerhans cell histiocytosis (LCH)

CANADIAN PAEDIATRIC SURVEILLANCE PROGRAM
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REPORTING INFORMATION
(To be completed by the CPSP Senior Coordinator)

Report number:	
Month of reporting:	
Province:	
Today's date:	

Please complete the following sections for the case identified above.
Strict confidentiality of information will be assured.

CASE DEFINITION FOR LANGERHANS CELL HISTIOCYTOSIS

Report any new patient presenting from birth up to their 18th birthday with:

- Clinical LCH features that may include unexplained bone pain and soft tissue swelling, diabetes insipidus and hypothalamic-pituitary dysfunction, proptosis, recurrent otitis or otorrhoea, maculopapular rash or seborrhoeic or napkin dermatitis resistant to treatment, interstitial pneumonitis, sclerosing cholangitis.

AND
either a or b

a) Biopsy proven LCH, with lesional cells containing:
   - Birbeck granules demonstrated on electron microscopy and/or
   - CD1a positive cells and/or
   - Langerin-positive cells and/or
   - S100 positive cells with characteristic H&E histopathology

b) Lytic bony lesions or pituitary/hypothalamic lesions characteristic of LCH without biopsy where:
   - Risks of biopsy are considered too hazardous due to site of lesion
   - Lesion has shown characteristic spontaneous regression

Month first seen _____

SECTION 1 – DEMOGRAPHIC INFORMATION

1.1 Date of birth: _____ /_____ /_____ (DD / MM / YYYY) 1.2 Sex: Male ___ Female ___

1.3 Province/Territory of residence at birth: _____ at time of reporting: _____

1.4 Ethnicity (check all that apply):
First Nations ___ Innu ___ Inuit ___ Métis ___ South Asian ___ Black ___ Caucasian ___
Latin American ___ Middle Eastern ___ Other (specify) _____
Unknown ___

SECTION 2 – FAMILY HISTORY

2.1 Father's occupation (before pregnancy): _____

2.2 Mother's occupation (during pregnancy): _____

2.3 Family history of LCH: Yes ___ No ___ Unknown ___
If yes, specify family member affected: _____ age at diagnosis: _____

2.4 Maternal history of thyroid disease: Yes ___ No ___ Unknown ___
If yes, specify: _____

2.5 History of smoking/second-hand smoke: Yes ___ No ___ Unknown ___

2.6 Associated, or previous malignancy: Yes ___ No ___ Unknown ___
If yes, specify type of malignancy:_____ age at diagnosis: _____

SECTION 2 – FAMILY HISTORY (cont'd)

		Yes	No	Unknown
2.7	Maternal pregnancy (check all that apply)			
2.7.1	Alcohol use	___	___	___
2.7.2	Cigarette use	___	___	___
2.7.3	Radiographs	___	___	___
2.7.4	Fever/infection	___	___	___
2.7.5	Bleeding	___	___	___
2.7.6	Medications	___	___	___

If yes, specify: _____

SECTION 3 – DIAGNOSIS AND REFERRAL

3.1 Presenting symptoms: _____

3.2 Date of first symptoms: _____ /_____ /_____ or age: _____ years _____months
DD MM YYYY

3.3 Date of first visit: _____ /_____ /_____ or age: _____ years _____months
DD MM YYYY

Specify type of physician: Family physician ___ Paediatrician ___ Orthopedic surgeon ___
Neurosurgeon ___ Paediatric haematologist/oncologist ___ Dermatologist ___ ENT surgeon ___
Other _____

If seen first by a paediatric haematologist/oncologist, go to question 3.6

3.4 Date of referral to specialist: _____ /_____ /_____ or age: _____ years _____months
DD MM YYYY

Specify type of specialist: Paediatrician ___ Orthopedic surgeon ___ Neurosurgeon ___
Paediatric haematologist/oncologist ___ Dermatologist ___ ENT surgeon ___
Other _____

3.5 Date of referral to paediatric haematologist/oncologist: _____ /_____ /_____
DD MM YYYY

3.6 Date of biopsy or date of definite diagnosis if no biopsy

Biopsy: _____ /_____ /_____ Definite diagnosis: _____ /_____ /_____
DD MM YYYY DD MM YYYY

3.7 Interval between symptom onset and diagnosis: ____ weeks

SECTION 4 – INVESTIGATIONS

		Yes	No	Unknown
4.1	Skeletal survey:	___	___	___

If yes, specify date: ____ /_____ /_____ results: _____
DD MM YYYY

4.2	Bone scan:	___	___	___

If yes, specify date: ____ /_____ /_____ results: _____
DD MM YYYY

4.3	Chest computerized tomography:	___	___	___

If yes, specify date: ____ /_____ /_____ results: _____
DD MM YYYY

4.4	Magnetic resonance imaging of the brain:	___	___	___

If yes, specify date: ____ /_____ /_____ results: _____
DD MM YYYY

SECTION 4 – INVESTIGATIONS (cont'd)

4.5 PET or PET-CT scan: ___ ___ ___

If yes, specify date: ____ /_____ /_____ results: _____
DD MM YYYY

4.6 Early morning urine osmolality: ___ ___ ___

If yes, specify date: ____ /_____ /_____ results: _____
DD MM YYYY

4.7 Water deprivation test: ___ ___ ___

If yes, specify date: ____ /_____ /_____ results: _____
DD MM YYYY

SECTION 5 – MANAGEMENT

		Yes	No	Unknown
5.1	Observation only	___	___	___
5.2	Surgical curettage	___	___	___

If yes, specify who performed the curettage: Paediatrician ___ Paediatric orthopedic surgeon ___
Paediatric neurosurgeon ___ Paediatric surgeon ___ Paediatric haematologist/oncologist ___
Other _____

		Yes	No	Unknown
5.3	Medications:			
5.3.1	Steroids	___	___	___
5.3.2	Vinblastine	___	___	___
5.3.3	Steroids and vinblastine	___	___	___
5.3.4	Steroids and vinblastine and methotrexate	___	___	___
5.3.5	Cladribine (2CDA)	___	___	___
5.3.6	Other, please specify: _____			
5.4	Enrollment in clinical trial	___	___	___
	If yes, specify type of clinical trial: _____			
5.5	Positive response to treatment	___	___	___
5.6	Salvage treatment required	___	___	___
	If yes, specify treatment: _____			

SECTION 6 – REPORTING PHYSICIAN

First name_____ Surname_____

Address_____

City_____ Province_____ Postal code_____

Telephone number_____ Fax number_____

E-mail_____ Date completed_____

___ **I agree to be contacted by the research team for further information.**

___ **I do not wish to be contacted by the research team for further information.**

Thank you for completing this form.