KERNICTERUS (K)

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 KERNICTERUS

Report any child up to six years of age with:

- a history of significant neonatal hyperbilirubinemia (peak bilirubin $>425\mu$mol/L or exchange transfusion)
  and
- two or more of the following symptoms:
  a) extrapyramidal disorders (e.g., dystonia, athetosis)
  b) other movement disorder (spasticity or hypotonia)
  c) gaze abnormalities
  d) sensorineural hearing loss
  e) intellectual deficits
  f) enamel dysplasia of the deciduous teeth

  OR
- abnormal MRI with bilateral lesions of basal ganglia/midbrain (globus pallidus + subthalamic nucleus) with a history of neonatal hyperbilirubinemia.

Exclusion criteria

- Born at less than 35 weeks gestational age.
- Metabolic condition with basal ganglia involvement (e.g., glutaric acidaemia type II, pyruvate dehydrogenase deficiency, Hallervorden-Spatz disease, neurofibromatosis type I, or children with carbon monoxide poisoning).

SECTION 1 – DEMOGRAPHIC INFORMATION

A) Infant/Child

1.1 Date of birth: _____ /_____ /_____
DD MM YYYY

1.2 Sex: Male ___ Female ___

1.3 Place of birth (country, province/territory): _____

1.4 Home birth: Yes ___ No ___

B) Mother

1.5 Place of residence (province/territory): _____

1.6 Age at delivery (years): _____

1.7 Ethnicity: Aboriginal _____ Black _____ Caucasian _____ Latin American _____ Middle Eastern _____
Asian _____ Other (specify): _____ Unknown _____

C) Father

1.8 Ethnicity: Aboriginal _____ Black _____ Caucasian _____ Latin American _____ Middle Eastern _____
Asian _____ Other (specify): _____ Unknown _____

SECTION 2 – FAMILY HISTORY

		Yes	No	Unknown	
2.1	Consanguinity	___	___	___	If yes, specify: _____
2.2	Early infant death	___	___	___	If yes, specify: _____
2.3	Previous sibling with neonatal jaundice	___	___	___	If yes, specify: _____
2.4	Family history of G6PD deficiency	___	___	___	If yes, specify: _____
2.5	Other neonatal hemolytic disease	___	___	___	If yes, specify: _____
2.6	Previous sibling with neurological disorder	___	___	___	If yes, specify: _____
2.7	Previous sibling with metabolic disorder	___	___	___	If yes, specify: _____

SECTION 3 – MEDICAL HISTORY

A) Neonatal history

3.1 Gestational age: _____ weeks completed

3.2 Birth weight: _____ grams

3.3 Apgar score at 1 minute _____ 5 minutes _____

3.4 Type of delivery: vaginal ____ forceps/vacuum ____ caesarian ____ unknown ____

3.5 Cord blood PH _____ PCO2 _____ Base excess _____

3.6 Neonatal peak bilirubin: _____ μmol/L Age: if < 24 hours _____ otherwise _____ days

3.7 Readmitted from home: Yes ___ No ___ Unknown ___ If yes, specify age: _____ days

3.8 Serum bilirubin before first discharge? Yes ___ No ___ Unknown ___
If yes, specify age: _____ days

3.9 Other neonatal medical conditions _____
_____
_____

3.10 Neonatal medications: Yes ___ No ___ Unknown ___

B) NEUROLOGICAL SYMPTOMS AS A NEONATE		Yes	No	Unknown
3.11	Poor sucking	___	___	___
3.12	Arching of neck (retrocollis)	___	___	___
3.13	Arching of back (opisthotonus)	___	___	___
3.14	Hypertonia	___	___	___
3.15	Hypotonia	___	___	___
3.16	Seizures	___	___	___
3.17	Weak/absent Moro reflex	___	___	___
3.18	Lethargy	___	___	___
3.19	Shrill cry	___	___	___

C) RISK FACTORS FOR NEONATE		Yes	No	Unknown	
3.20	Evidence of hemolysis	___	___	___	
3.21	Pronounced hematoma	___	___	___	
3.22	Evidence of neonatal hemorrhage	___	___	___	
3.23	Breast-feeding	___	___	___	If yes, specify: exclusive __ breast/formula __
3.24	Feeding difficulties	___	___	___	
3.25	Sepsis/infection	___	___	___	If yes, specify: _____

SECTION 3 – MEDICAL HISTORY (cont'd)

3.26 Asphyxia	___	___	___
3.27 Dehydration	___	___	___

D) NEONATAL DIAGNOSIS FOR HYPERBILIRUBINEMIA	**Yes**	**No**	**Unknown**	
3.28 ABO incompatibility	___	___	___	
3.29 Other antibodies	___	___	___	
3.30 G6PD deficiency	___	___	___	
3.31 Pyruvate kinase deficiency	___	___	___	
3.32 Hereditary spherocytosis	___	___	___	
3.33 Unstable hemoglobin	___	___	___	
3.34 Urinary tract infection	___	___	___	
3.35 Sepsis	___	___	___	If yes, specify: _____
3.36 Hypothyroidism	___	___	___	
3.37 Hypernatremic dehydration	___	___	___	
3.38 Other	___	___	___	If yes, specify: _____

E) NEONATAL TREATMENT FOR HYPERBILIRUBINEMIA	**Yes**	**No**	**Unknown**	
3.39 Phototherapy	___	___	___	
3.40 Exchange transfusion	___	___	___	
3.41 Intravenous immunoglobulin administration	___	___	___	
3.42 Other blood products	___	___	___	If yes, specify: _____

SECTION 4 – CRITERIA TO ESTABLISH DIAGNOSIS OF KERNICTERUS

A) Neurological symptoms	**Yes**	**No**	**Unknown**	
4.1 Extrapyramidal symptoms	___	___	___	If yes: athetosis ___ chorea ___ dystonia ___
4.2 Disturbance of tone	___	___	___	If yes: hypotonia ___ hypertonia ___
4.3 Oral motor problems	___	___	___	If yes: dysarthia ___ chewing/swallowing difficulties ___
4.4 Paralysis of upward gaze	___	___	___	
4.5 Vision impairment	___	___	___	
4.6 Hearing loss	___	___	___	
4.7 Developmental delays	___	___	___	
4.7.1 Motor skills	___	___	___	
4.7.2 Intellectual	___	___	___	
4.7.3 Language	___	___	___	
4.8 Seizures	___	___	___	
4.9 Tooth/enamel dysplasia	___	___	___	

B) Investigations

4.10 MRI of the brain Yes ___ No ___

If yes, bilateral lesions of basal ganglia/midbrain: Yes ___ No ___

If yes, specify other specific MRI findings:* _____

_____

*Pease provide a copy of the MRI report removing name and identifying data and adding the CPSP report number on previous page.

4.11 CT of the brain	Yes ___	No ___	Findings _____
4.12 BERA/ABR	Yes ___	No ___	Findings _____
4.13 Audiogram	Yes ___	No ___	Findings _____
4.14 Autopsy	Yes ___	No ___	Findings _____
4.15 Others, specify: _____	Yes ___	No ___	Findings _____

SECTION 4 – CRITERIA TO ESTABLISH DIAGNOSIS OF KERNICTERUS (cont'd)

C) Diagnosis of kernicterus

4.16 Age of diagnosis: _____ /_____ /_____
DD MM YYYY

4.17 Diagnosis confirmed by: Family physician ___ Paediatrician ___ Neurologist ___

4.18 Was there another diagnosis confirmed?

4.18.1	Glutaric acidaemia type II	Yes ___	No ___	
4.18.2	Pyruvate dehydrogenase deficiency	Yes ___	No ___	
4.18.3	Hallervorden Spatz disease	Yes ___	No ___	
4.18.4	Neurofibromatosis type I	Yes ___	No ___	
4.18.5	Carbon monoxide poisoning	Yes ___	No ___	
4.18.6	Other metabolic disorder	Yes ___	No ___	If yes, specify_____
4.18.7	Other medical condition	Yes ___	No ___	If yes, specify_____

SECTION 5 – REPORTING PHYSICIAN

First name_____ Surname_____

Address_____

City_____ Province_____ Postal code_____

Telephone number_____ Fax number_____

E-mail_____ Date completed_____

SECTION 6 – FOLLOW-UP

6.1 Will you follow up this child? Yes ___ No ___

If yes, are you willing to be contacted for further information/cohort study? Yes ___ No ___

Thank you for completing this form.