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 >425µmol/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: _______ / _______ / _______
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

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

| 3.11 Poor sucking | Yes | No | Unknown |
| 3.12 Arching of neck (retrocollis) | Yes | No | Unknown |
| 3.13 Arching of back (opisthotonus) | Yes | No | Unknown |
| 3.14 Hypertonia | Yes | No | Unknown |
| 3.15 Hypotonia | Yes | No | Unknown |
| 3.16 Seizures | Yes | No | Unknown |
| 3.17 Weak/absent Moro reflex | Yes | No | Unknown |
| 3.18 Lethargy | Yes | No | Unknown |
| 3.19 Shri! cry | Yes | No | Unknown |

**C) RISK FACTORS FOR NEONATE**

| 3.20 Evidence of hemolysis | Yes | No | Unknown |
| 3.21 Pronounced hematoma | Yes | No | Unknown |
| 3.22 Evidence of neonatal hemorrhage | Yes | No | Unknown |
| 3.23 Breast-feeding | Yes | No | Unknown | If yes, specify: exclusive __ breast/formula __ |
| 3.24 Feeding difficulties | Yes | No | Unknown |
| 3.25 Sepsis/infection | Yes | No | Unknown | 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
3.36 Hypothyroidism
3.37 Hypernatremic dehydration
3.38 Other

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

SECTION 4 – CRITERIA TO ESTABLISH DIAGNOSIS OF KERNICTERUS

A) Neurological symptoms  Yes  No  Unknown
4.1 Extrapyramidal symptoms
4.2 Disturbance of tone
4.3 Oral motor problems
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
4.11 CT of the brain
4.12 BERA/ABR
4.13 Audiogram
4.14 Autopsy
4.15 Others, specify:

*Please provide a copy of the MRI report removing name and identifying data and adding the CPSP report number on previous page.
SECTION 4 – CRITERIA TO ESTABLISH DIAGNOSIS OF KERNICTERUS (cont’d)

C) Diagnosis of kernicterus

4.16 Age of diagnosis: _____ /_____ /_______

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.